

**CHARGE SYNDROME IS A MEDICAL DIAGNOSIS.
CAN IT ALSO BE AN EDUCATIONAL DIAGNOSIS?**

by

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ABSTRACT

CHARGE syndrome is a common cause of congenital deafblindness, but it has been contended individuals with CHARGE form a distinct group within the broader deafblind population. This thesis explores the education of learners with CHARGE and what the similarities and differences between these two groups might be.

A review of literature identifies reported anomalies that may impact upon learning and development, and establishes very limited research-based evidence is available with regard to educational practice with this group of learners.

Cycle 1 of this investigation involved document analysis of educational reports, revealing internal factors considered to influence learning and development, and external factors including assessment, support from external professionals and teaching strategies. These were explored further in Cycle 2 involving a questionnaire to teachers of a child with CHARGE and interviews of practitioners in an overseas educational establishment.

Commonalities and distinctions between learners with CHARGE and the wider deafblind population were found, and also that established educational deafblind practice is applicable to learners with CHARGE, but that strategies may be alternatively employed and additional strategies also required. A variation in educational provision was also found according to the type of placement attended. In conclusion it was considered that, in a broad sense, educationally there is something unique and distinct in learners with CHARGE.

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CONTENTS

CHAPTER 1	INTRODUCTION	
1.1	CHARGE syndrome	1
1.2	My experience of working with children and young people with CHARGE	5
1.3	Personal statement	8
1.4	The development of research questions	10
1.5	The research framework	12
1.6	Terminology and definitions	
	1.6.1 CHARGE syndrome	14
	1.6.2 Deafblindness/Multi-Sensory Impairment	14
	1.6.3 The educational environment	17
1.7	Summary and conclusion	17
CHAPTER 2	CHARGE: A REVIEW OF LITERATURE	
2.1	Introduction	19
2.2	Aspects of CHARGE that may affect learning	23
	2.2.1 Vision	24
	2.2.2 Hearing	26
	2.2.3 Co-occurring visual and hearing impairments	29
	2.2.4 Vestibular issues	29
	2.2.5 Proprioception	31
	2.2.6 Low tone	32
	2.2.7 Touch	33
	2.2.8 Sensory integration dysfunction	35
	2.2.9 Neurology	40
	2.2.10 Cranial nerve anomalies	44

	2.2.11 Sleep disturbances	46
	2.2.12 Medical intervention, hospitalisation and pain	47
	2.2.13 Later onset features	49
	2.2.14 Summary	50
2.3	Development and function in individuals with CHARGE	51
	2.3.1 Gross motor development	52
	2.3.2 Communication and social interaction	55
	2.3.3 Cognitive development and attainment	61
	2.3.4 Behaviour	66
	2.3.5 Self-regulation	69
	2.3.6 Fatigue	70
	2.3.7 Summary	70
2.4	Assessment	71
2.5	CHARGE and deafblindness	74
2.6	Educating learners with CHARGE	76
	2.6.1 Meeting the educational needs of learners with CHARGE	82
	2.6.2 Educational placement	86
	2.6.3 Teacher strategies for learners with CHARGE	87
	2.6.3a Focusing on the child	88
	2.6.3b Sensory needs	90
	2.6.3c Communication and social interactions	93
	2.6.3d Functional life skills	95
	2.6.3e Behaviour management	95
	2.6.3f Delivery of the curriculum	96
	2.6.4 Summary	99
2.7	Final summary and conclusions	101

CHAPTER 3	DEVELOPING A RESEARCH FRAMEWORK: METHODOLOGY, METHODS AND ETHICAL ISSUES	
3.1	Introduction	107
3.2	Choice of methodology	108
3.3	Choice of methods	112
	3.3.1 Use of documentation	113
	3.3.2 Surveys	113
	3.3.2a Postal, self-completion questionnaire	114
	3.3.2b Semi-structured interviews	117
3.4	A summary of the research process	118
3.5	Data analysis	119
	3.5.1 Data analysis in Cycle 1	121
	3.5.2 Data analysis in Cycle 2	123
	3.5.3 Use of computer software for data analysis	123
3.6	Cycle 1	125
	3.6.1 Data collection	125
	3.6.2 Identification of subjects and educational reports	126
	3.6.3 Obtaining consent	127
	3.6.4 Authorship of the reports	128
	3.6.5 Limitations of the reports	129
	3.6.6 Extracting the data	130
	3.6.7 Data analysis	130
	3.6.7a Qualitative and quantitative analysis	131
	3.6.7b Developing the coding categories	132
	3.6.7c Undertaking a check of validity and reliability	133
	3.6.7d The final codings	136
3.7	Cycle 2: Questionnaire	138
	3.7.1 Questionnaire design	138

	3.7.2 Unit of analysis	139
	3.7.3 Content and structure	139
	3.7.4 Question format	141
	3.7.5 Pre-test of questions	144
	3.7.6 Pilot of the questionnaire	145
	3.7.7 Data collection	145
	3.7.7a Identification of potential respondents	145
	3.7.7b Obtaining informed consent	147
	3.7.7c Confidentiality and anonymity	148
	3.7.7d Securing a positive response rate	148
	3.7.7e Safe and appropriate storage and handling of data	150
	3.7.8 Data collation and analysis	150
	3.7.9 Reliability check	151
3.8	Cycle 2: Interviews	153
	3.8.1 Background	153
	3.8.2 The interview questions	154
	3.8.3 Data collection	155
	3.8.3a Identification of participants	155
	3.8.3b Gaining informed consent, anonymity and confidentiality	155
	3.8.3c The interview process	156
	3.8.3d Recording the interviews	156
	3.8.4 Data collation and analysis	156
3.9	Establishing trustworthiness	157
	3.9.1 Content validity	157
	3.9.2 Seeking appropriate methodology and methods	158
	3.9.3 The lone researcher	158
	3.9.4 Quality of evidence	159
	3.9.5 Multiple sources of evidence	160
	3.9.6 Reliability	161
	3.9.7 Generalisation	163

3.10	Ethical considerations	163
	3.10.1 The best interests of the children	163
	3.10.2 Respect for all involved individuals	165
	3.10.3 Gaining informed consent	165
	3.10.4 Confidentiality and anonymity	166
	3.10.5 Safe and appropriate storage and handling of data	167
	3.10.6 Costs and benefits	167
3.11	Summary and conclusions	168
CHAPTER 4	CYCLE 1: SUMMARY OF DATA GATHERED, ANALYSIS AND DISCUSSION	
4.1	Introduction	171
4.2	Quantitative information	172
4.3	Findings: The coding categories	174
	4.3.1 Internal factors	176
	4.3.2 External factors	178
	4.3.3 The provision of specialist support to teachers and the use of assessment processes	181
4.4	Discussion	183
4.5	Addressing the research questions	186
4.6	Initial insights and theory	188
4.7	Developing the next cycle of the research	190
4.8	Summary and conclusions	193

CHAPTER 5 CYCLE 2: QUESTIONNAIRE AND INTERVIEWS SUMMARY OF ANALYSIS AND INITIAL DISCUSSIONS

5.1	Introduction	196
5.2	Presentation of data and initial discussion	196
5.3	Questionnaire responses	199
	5.3.1 Initial checks	200
	5.3.2 A check for consistency of responses	200
5.4	Demographics	201
	5.4.1 Age range and current educational stages	201
	5.4.2 Type of educational provision	202
5.5	Child's primary need	204
5.6	The child	205
	5.6.1 Characteristics and sensory responses	205
	5.6.2 Communication	209
	5.6.2a Child's preferred communication modes	212
	5.6.2b Communication modes	215
	5.6.2c Communicative exchanges with a sensitive communication partner	219
	5.6.3 Social and emotional	223
	5.6.4 Conceptual ability	227
	5.6.5 Responses to routines and structure, and understanding of time and space	231
	5.6.6 Ownership of learning	236
	5.6.7 Orientation and mobility, and fine motor skills	238
	5.6.7a Orientation and mobility	238
	5.6.7b Fine motor skills	243
	5.6.8 General comment	245

5.7	Strategies	248
	5.7.1 Sensory responses	249
	5.7.2 Communication	252
	5.7.3 Social and emotional	256
	5.7.4 Conceptual ability	259
	5.7.5 Responses to routines and structure, and understanding of time and space	262
	5.7.6 Ownership of learning	265
	5.7.7 Orientation and mobility, and fine motor skills	268
	5.7.7a Orientation and mobility	268
	5.7.7b Fine motor skills	270
	5.7.8 General comment	271
5.8	Assessment	278
	5.8.1 Environmental audits	284
5.9	Professional support	285
5.10	Interviews	288
	5.10.1 Question 1a Do you feel that individuals with CHARGE are different to the broader deafblind population?	289
	5.10.2 Question 1b: If yes, in what way?	289
	5.10.3 Question 2: What teaching strategies do you find most helpful (or unhelpful)?	291
	5.10.4 Question 3: Do you consider their sensory impairments to be their primary need?	292
	5.10.5 Q4: What, if any, support have you had in relation to CHARGE syndrome?	295
5.11	Final summary and conclusions	296

CHAPTER 6	FINAL DISCUSSIONS AND CONCLUSION	
6.1	Introduction	301
6.2	The main findings	302
	6.2.1 What are the factors within the child likely to affect learning?	302
	6.2.2 What aspects of CHARGE syndrome might be distinct from the more general deafblind/MSI population?	310
	6.2.2a The classification of Special Educational Needs debate	314
	6.2.3 What strategies are likely to be effective in supporting the learning of children with CHARGE syndrome?	316
	6.2.4 CHARGE syndrome is a medical diagnosis. Can it also be considered an educational diagnosis?	322
	6.2.5 Learners with CHARGE in different types of educational settings	325
6.3	A review of the investigative process	329
6.4	Conclusions	337

LIST OF FIGURES

FIGURE A	The sensory integration process	36
FIGURE B	The research design framework for this study	118

LIST OF TABLES

Table 1	The medical diagnostic criteria for CHARGE syndrome according to Sanlaville and Verloes (2007)	3
Table 2	A summary of the skills involved in executive function	42
Table 3	The Cranial Nerves included in the diagnostic criteria for CHARGE	45
Table 4	A CHARGE behavioural phenotype	67
Table 5	A summary of recommendations with regard to the CHARGE behavioural phenotype	68
Table 6	Types of educational provision of children with CHARGE reported in two studies	87
Table 7	Number of reports contributed to by each author	128
Table 8	A summary of the initial codings generated during the process of open and axial coding and the revised codings developed following the check of validity and reliability of the coding process	134
Table 9	Cycle 1-Total number of different statements per category and number of children for whom those statements were made	173
Table 10	Those Internal factors from the coding categories, not identified within the sample of literature relating to the broad deafblind/MSI population	178
Table 11	Those external factors identified from the coding categories, not identified within the sample of literature relating to the broad deafblind/MSI population	179
Table 12	The number of children from the study in each key stage	201
Table 13	Types of educational provision where children were	202
Table 14	Presence and distribution of characteristics among types of provision and educational phases	207
Table 15	Factors within the child considered to effect the child's communication and distribution among types of provision and educational phases	211
Table 16	Child's preferred communication modes to receive communication and express themselves and distribution among types of provision and educational phases	214
Table 17	Communication modes used to support communication and distribution among types of provision and educational phases	216

Table 18	Number of communication modes used with a child to support communication, and distribution among types of provision	217
Table 19	A summary of the average scores obtained from the Likert Scales relating to 'The Child'	220
Table 20	Comparison of the number of peer friendships in 5 children for whom more than 10 secure relationships with adults were reported	225
Table 21	Score of children in KS3/4 for ease or difficulty in establishing abstract concepts	228
Table 22	Types of concrete cues used to support routine and structure and understanding of time and space	234
Table 23	Number of concrete cues reported to be used per child to support routine and structure and understanding of time and space	235
Table 24	Factors, other than sensory impairments, considered to affect child's orientation and mobility	240
Table 25	Presence of behaviours indicative of adaptive responses to vestibular and proprioceptive difficulties	242
Table 26	Responses to questions relating to fine motor skills	244
Table 27	Numbers of respondents who considered given strategies to be helpful with regard to the child's sensory needs	250
Table 28	Numbers of respondents who considered given strategies to be helpful with regard to communication	253
Table 29	Numbers of respondents who considered given strategies to be helpful with regard to social and emotional development	257
Table 30	Numbers of respondents who considered given strategies helpful with regard to conceptual ability	260
Table 31	Numbers of respondents who considered given strategies to be helpful with regard to response to routine and structure, and understanding of time and space	263
Table 32	Numbers of respondents who considered given strategies to be helpful with regard to ownership of learning	266
Table 33	Numbers of respondents who considered given strategies helpful with regard to orientation and mobility and fine motor skills	269
Table 34	Strategies more often perceived as helpful when employed in specialist settings than in a mainstream setting	273
Table 35	Strategies more often perceived as helpful when employed in an SLD/PMLD setting than other specialist settings or a mainstream setting	274

Table 36	Those strategies from inventories reported to be helpful by less than 45% of respondents	276
Table 37	Frequency of types of assessment undertaken or made available and level of effectiveness reported	278
Table 38	Types of environmental audits and the frequency where reported to be considered helpful and distribution among types of provision	284
Table 39	Potential characteristics in a child or young person with CHARGE that may influence learning and development	308
Table 40	Strategies included within the questionnaire and number of respondents reporting them to be helpful, and identification of whether each strategy is named by interviewees and in literature	317

APPENDICES

Appendix 1	Information booklet for parents	342
Appendix 2	Table (i) : A check of reliability and validity to discover the level of agreement between the assignment of statements to different coding categories- Pilot	344
	Table (ii): A second check of reliability and validity to discover the level of agreement between the assignment of statements to different coding categories	345
Appendix 3	Questionnaire	346
Appendix 4	Information booklet for participants	360
Appendix 5	Using Spearman's rho (rank correlation coefficient) to ascertain the consistency in responses given to questions C2 and S2	362
Appendix 6	Interview information letter/consent form	364
Appendix 7	Table (iii): A summary of the internal factors identified for each coding category	365
	Table (iv): A summary of the external factors identified for each coding category	368
Appendix 8	Table (v): Likert Scale scores for questions on communicative exchanges with a sensitive communication partner and distribution among types of provision and educational phases	372
	Table (vi): Number of adults with whom the child is reported to have a trusting and secure relationship/ number of peers with whom the child is reported to have a genuine friendship in school	373
	Table (vii): Likert scale score for questions relating to the development of relationships and emotional development	374
	Table (viii): Likert scale scores for questions relating to the child's conceptual ability Part 1	375
	Table (ix): Likert scale scores for questions relating to the child's conceptual ability Part 2	376
	Table (x): Likert scale scores for questions relating to child's responses to routines and structure, and understanding of space and time Part 1	377
	Table (xi): Likert scale scores for questions relating to child's responses to routines and structure, and understanding of space and time Part 2	378

Appendix 8 (contd.)	Table (xii): Likert scale scores for questions relating to ownership of learning	379
	Table (xiii): Factors considered to adversely affect the child's readiness and ability to engage and learn	380
	Table (xiv): Rank order of Likert scale scorings for how easy or difficult children were reported to find identified skills	381
Appendix 9	Table (xv): Communication modes identified as being used as part of a Total Communication approach	382
	Table (xvi): Range and average number of communication modes per child as part of a Total Communication approach	383
	Table (xvii): The distribution of responses across types of provision and educational phases where use of concrete tools was considered helpful to support concept development	383
	Table (xviii): Strategies regarded as helpful by more than 50% of respondents, set out in rank order	384
Appendix 10	Professional support	387
Appendix 11	Table (xix): Ways in which interviewees considered students with CHARGE to be different to other deafblind learners	388
	Table (xx): Strategies identified as helpful by interviewees	389
	Table (xxi): Support received by interviewees in relation to students with CHARGE	390
Appendix 12	Table (xxii): Rate of reporting of behaviours indicative of vestibular and proprioceptive difficulties	391
	Table (xxiii): Presence of potential indicators of sensory integration dysfunction	392
	Table (xxiv): Characteristics that may be present in a child or young person with CHARGE and influence learning and development, with related examples found in literature on deafblind education	392

CHAPTER 1- INTRODUCTION

I was introduced to my first child with CHARGE syndrome over twenty years ago, working as a teacher in the field of deafblindness. I have since been privileged to work with a number of children and young people with this condition. These individuals have continually led me to consider whether they are distinct from the wider deafblind population, and I have regularly been challenged and required to think in a new way when working to support them. Questions have continued to be raised during my work with these individuals with CHARGE syndrome and ultimately led to the studies that form this thesis, focusing on the child and the support provided within the educational setting.

1.1 CHARGE syndrome

CHARGE syndrome is a highly complex condition first described in literature in 1979 independently by Hall and also by Hittner et al. The original acronym 'CHARGE' presented by Pagon et al. in 1981 was felt to represent the primary characteristics of the condition:

C- Coloboma

H- Heart

A- Atresia of the choanae

R- Retardation of growth and development

G- Genitalia anomalies

E- Ear anomalies and hearing loss

An example of the importance of ongoing research informing the growth of knowledge is illustrated in the fact that as more became known about this condition, it has led to these initial diagnostic criteria being superseded by revised diagnostic criteria (Blake et al. 1998, and later by Sanlaville and Verloes, 2007), but the use of 'CHARGE' as an acronym has remained, possibly as a reminder of the origins of the name. The current diagnostic criteria used to make a medical diagnosis of CHARGE syndrome (Sanlaville and Verloes, 2007) are shown in Table 1 on the following page. The prevalence of these anomalies according to Hartshorne et al. (2011) is also shown. Previously an identified number of major and minor characteristics in combination was required for a diagnosis to be made, but there is now less clarity with regard to this; although Sanlaville and Verloes (2007) suggest the presence of three major or two major and two minor characteristics, no degree of affectedness is established, and as Hartshorne et al. (2011) explain, within each affected individual the characteristics range from being present to absent and from mild to severe in nature.

Initially CHARGE was considered to be an 'association' as it was felt this group of anomalies were not merely coming together by chance. Over time, supported by ongoing medical research indicating a distinct non-random pattern of anomalies presented, there was a growing opinion this condition should be designated as a 'syndrome'. With the discovery of a mutation of the CHD7 gene in a significant number of individuals with CHARGE in 2004 (Vissers et al.), the condition was then universally accepted as a syndrome and consequently now recognised as 'CHARGE syndrome' (Hartshorne et al., 2007). This stresses the uniqueness of CHARGE syndrome as a

TABLE 1: THE MEDICAL DIAGNOSTIC CRITERIA FOR CHARGE SYNDROME ACCORDING TO SANLAVILLE AND VERLOES (2007)

MAJOR diagnostic characteristics	Frequency of occurrence according to Hartshorne et al. (2011)
Coloboma of the eye	80-90%
Choanal atresia or stenosis (Blockage or narrowing of the airways at the back of the nose)	50-60%
Cranial nerve dysfunction or anomaly:	
○ I – hyposmia or anosmia (reduced or lacking sense of smell)	>90%
○ VII – facial palsy	40%
○ VIII hypoplastic auditory nerve	>80%
○ IX/ X swallowing difficulties with aspiration	70-90%
Characteristic CHARGE outer ear (shape)	>50%
Characteristic CHARGE middle/inner ear:	
○ Ossicular malformations	>80%
○ Mondini defect of the cochlea	>80%
○ Absent or hypoplastic semicircular canals)	>90%
MINOR diagnostic characteristics	
Genital hypoplasia	
○ Under-developed sexual parts	50%
○ Delayed puberty	>50%
	(but affecting 90% of males)
Cardiovascular malformation	75-85%
Growth deficiency:	
○ Short stature	70%
○ Growth hormone deficiency	15%
Cleft lip and/or palate	15-20%
Tracheoesophageal (T-E) fistula	15-20%
Renal anomalies	30-40%
Distinctive facial features	70-80%
Palmer crease	50%

distinct medical condition, and also raises the possibility of the separateness of this group of children from other learners who are deafblind. In both literature and conversation, this condition is often now simply referred to as 'CHARGE' and for ease of reference this term will be adopted for the remainder of this document.

Prior to the discovery of the CHD7 gene mutation in many individuals with CHARGE Jones & Dunne (1988) had reported:

The specific defects seen in this disorder can for the most part be attributed to arrest in various aspects of normal embryonic development (p.37).

This relates to the later identification of anomalies in the CHD7 gene by a team at The University Centre, St. Radboud, Netherlands (Vissers et al., 2004). CHD7 belongs to a family of regulation genes that potentially affect a large number of the developmental pathways. A defect in this process is thought to occur during early embryological development between the fourth and ninth week of gestation (Salem-Hartshorne and Jacob 2004; Sanlaville and Verloes 2007) and explains why so many organ systems are involved with CHARGE. Whilst mutations of the CHD7 gene have been found in a significant number of individuals with this condition, in testing of some individuals with CHARGE this mutation or deletion may not be revealed. It has been stressed this does not mean these individuals do not have CHARGE, but rather that there is another cause, possibly another gene that has yet to be identified (Kirk, 2005). As a consequence of this, although mutations are found in significant numbers of individuals with CHARGE, diagnosis currently continues to use the medical diagnostic criteria (Blake et al., 2008). When undertaking research involving the study of individuals with CHARGE, an essential criterion would therefore need to be that a clinical diagnosis had been received, rather than identification of a CHD7 mutation.

From the incidence rates provided by Hartshorne et al. (2011) it is possible to note that ear and eye anomalies are among the most commonly occurring and Hartshorne et al. (2011) report that approximately 90% of individuals with CHARGE have combined visual and hearing impairments and should therefore be recognised as part of the deafblind/multi-sensory impaired population. CHARGE has become recognised in recent years as a common cause of congenital deafblindness (Hartshorne, 2005) and particularly as a leading cause of congenital deafblindness among genetic conditions (Hartshorne et al. 2011).

Hartshorne et al. (2011) stated that CHARGE may be the only disorder that can cause “deficits of all the senses” (p.xiii) and as a consequence of this they may be regarded as “the most truly ‘multi-sensory impaired’ individuals you will ever meet” (Brown 2003b. p1). When studying CHARGE, it becomes evident there are issues in addition to deafblindness that will impact on the affected individual and create additional challenges. Herein lies an issue; are children with CHARGE to be viewed as part of the general deafblind population or should they be regarded as a distinct group? If they are distinct does this require a different response from those working to support these children and young people in the educational setting?

1.2 My experience of working with children and young people with CHARGE

When working as a peripatetic teacher for deafblind/multi-sensory impaired children over twenty years ago I received a referral for a little girl, Sarah¹, with CHARGE. The information made available confirmed to me Sarah was deafblind, and although I had

¹ Pseudonym used to protect anonymity

not heard of or supported a child with this condition previously, this was not an unusual situation to find myself in. I arranged to meet Sarah and her family in the home setting. Even during this initial visit I felt this child was very different from other deafblind children I was working with. On subsequent visits I was pushed even further out of my comfort zone and realised I was being challenged and required to think in a new way if I was to be able to support this child effectively.

Time passed and I received referrals for a few other children with CHARGE. I began to experience again what I had encountered previously with Sarah as I was forced to think and respond differently. I began to consider if there were specific elements particular to those children with CHARGE:

- Why did these children need to spend so much time lying on their backs?
- Why was it so challenging for these children to be in a supported upright position?
- Why was there such an overwhelming need for strict routine and structure?
- Why could a child recognise a word on one page but not be able to recall it when seen on the next page?
- Why do I need to develop or change my strategies that have worked with other deafblind children?
- Do these children think 'differently'?

The questions were endless. Additionally, the establishment of CHARGE as a syndrome in 2004 emphasised the uniqueness of this condition medically, but also fuelled my questioning of whether the CHARGE population is also distinct in other ways, including educationally. It was difficult finding other educationalists with

experience of working with children with CHARGE, and those I did communicate with also seemed to have more questions than answers. Within literature focused on CHARGE the emphasis was on medical reporting and research; however, some articles containing anecdotal reporting began to emerge that made reference to the education of children with CHARGE. Jones and Dunne (1988) identified a few implications for teachers working with a child with CHARGE based on their experiences; Lewis and Lowther (2001) considered the social behaviour of children with CHARGE; and Griffin et al. (2004) very briefly looked at how educational needs should be addressed. These articles provided some useful information to support me in my work but I felt a note of caution was required since these articles provided only anecdotes and opinion. Furthermore, the information available needed to be considered in light of subsequent additional medical knowledge continuing to emerge about this condition.

My working practice continued to be informed most by the children with CHARGE with whom I was involved. At times my thoughts would be developed further and occasionally confirmed by new medical research. For example, I had felt these children appeared to process information differently, this often being reflected in their responses and necessitating things being presented in another way. Then in 2004 Vissers et al. identified the involvement of the CHD7 gene. This works in early embryonic development and affects the development of the major organs (including the brain). A study by Raqbi, et al. a year later (2005) found evidence that the formation of the brain may be altered in a significant number of individuals with CHARGE. Blake et al. (2008) subsequently reported that as a consequence of the

altered neural crest formation, the twelve pairs of cranial nerves (that facilitate sensory functioning and control some muscles of the sense organs) are often affected. All this supported my thoughts that these children may process information differently to other deafblind children.

As mentioned previously, Brown (2003a) suggested children with CHARGE are truly multi-sensory impaired and at a conference he further suggested (2003b) that the issues present in CHARGE, in addition to the deafblindness, make the population difficult for those professionals working to support them and, he argued, different within the whole deafblind population. If this proposition is correct it leads to the question of whether children with CHARGE learn differently and whether they need a different educational approach to other deafblind learners. Certain aspects of functioning of individuals with CHARGE have now been addressed through research reported in literature, for example: behaviour (e.g. Hartshorne et al., 2005a); executive function difficulties (e.g. Nicholas, 2005); and communication (e.g. Thelin and Fussner, 2005). There has, however, to date been no published work on research relating to the educational philosophy and pedagogy of working with children with CHARGE.

1.3 Personal statement

During the period of time in which these studies have been undertaken I have worked as a multi-sensory impairment (MSI) teacher and then as principal MSI consultant with a voluntary organisation involved in supporting deafblind children and adults. Through my work I continue to have contact with a number of different children with CHARGE

and also have opportunities to share my thoughts and work with other practitioners. I have also been involved in other research in relation to CHARGE (that will be referred to later in this work) and have been fortunate to present at a number of national and international conferences within this field.

My initial degree was in the 'Education and Psychology of Mentally handicapped Children'. I subsequently qualified as a teacher of the deaf and then undertook a Masters in Education for MSI. During my initial studies the primary emphasis was on the use of a behaviourist approach. I acknowledge the benefits of this in supporting children to develop certain skills (such as putting on and fastening a coat), but have felt it to be lacking in supporting the development of deeper knowledge and secure concept formation. Whilst studying to become a teacher of the deaf I was encouraged and supported to become more flexible in my thinking and practice, and more focused on the child allowing myself to be child-led in my approach. During this time I was extremely fortunate to spend some time at the Rafael School in St. Michelsgestel (Netherlands), working with and observing a teacher at the centre, Dr. Jan van Dijk. He influenced my view of working with and educating children who are deafblind then, and has continued to do so over the years. I believe a child-centred approach is of great importance, identifying a child's strengths and needs and tailoring an individualised educational programme in response to those strengths and needs. Consequently, being a practitioner first and foremost, this would have an impact on the focus of any research undertaken, and this would be driven by a desire to improve educational practice with children with CHARGE.

1.4 The development of research questions

Petre and Rugg (2012) discuss the need to identify where potential gaps are in a given knowledge base as a way of informing the development of research questions. They explain the aim is to “reduce the problem space” (p.112). At the beginning of this process I was able to quickly identify that the “problem space” in this instance was the lack of research-based evidence and knowledge in relation to the pedagogy of learners with CHARGE. The creation of a clearly defined research question was a more complicated and quite convoluted process that involved a good deal of time and thought.

The United Nations (UN) Covenant on Economic, Social and Cultural Rights (1966) clearly states in Article 13 that everyone has right to an education that will support the full development of the individual. This is reinforced in the Convention on the Rights of the Child (United Nations, 1989) where in Articles 28 and 29 it emphasises that children and young people should be supported to achieve the highest level of education they are capable of. This clearly applies to learners with CHARGE, presenting the educationalist with the challenge of how to deliver an effective education that will foster such development and learning in affected children.

In undertaking this study the intention would be to increase understanding of this phenomenon, identifying what is already known and to provide further insights (thus reducing the “problem space”), and to ultimately provide evidence to support the improvement of educational practice with learners with CHARGE. One definition of education might be considered as the receiving of systematic instruction delivered

within a formal school setting. In broader terms, however, it might be regarded as any formative process influencing the individual child or young person in the development of their thinking, feeling and how they act. It was decided that for the purposes of this investigation the focus would be on the education of children with CHARGE in the established educational setting of the school, although it was recognised that findings might also apply to the broader environment the child encounters throughout their day.

Eventually the overarching research question identified was:

CHARGE syndrome is a medical diagnosis.
Can it also be considered as an educational diagnosis?

By making a medical diagnosis of CHARGE the medical profession is classifying a condition into the distinct category of this particular syndrome (see 1.1 above), and by doing so allowing for decisions to be made about the response needed. This research question encourages an investigation into whether there can also be an educational diagnosis for children identified with CHARGE, and if a particular (and possibly distinct) response is required. There has been some discussion as to whether the CHARGE populace are distinct from the wider deafblind population (See 2.5) and therefore form a separate group. The profile of CHARGE as a distinct group has been suggested by a number of practitioners and also raised and promoted by the CHARGE Syndrome Foundation in the USA and other active CHARGE family support groups. This illustrates a point raised by Hollenweger (2008) who identified a number of possible factors influencing the development of any category, including the advocacy of parent, professional and other groups. This investigation would help clearly identify if, and in what ways, learners with CHARGE might be different to other deafblind learners that may be challenging to the class teacher, and what might

practically be undertaken in the classroom to support learning. To make the exploration into this question more manageable it was necessary to identify a series of smaller sub-questions. These sub-questions were honed during the literature review process by repeatedly asking what it was that needed to be known. Eventually these thoughts were refined further to develop the following sub-questions:

- What are the factors within the child likely to affect learning?
- What strategies are likely to be effective in supporting the learning of children with CHARGE?
- What aspects of CHARGE might be distinct from the more general deafblind/MSI population?
- What specialist support is provided from other professionals and through assessments, to teachers working with a child with CHARGE?

1.5 The research framework

Having established the research questions it was necessary to identify the research methodology and techniques that would provide the framework for this study. An extensive literature review had revealed little evidence-based research in relation to the education of children with CHARGE (with the exception of Lieberman et al., 2012, who investigated physical education in learners with CHARGE), and a consequent heavy reliance on anecdotal reporting. It was recognised this research would need to be exploratory and descriptive in nature with an emphasis on discovery, with a focus on accumulating knowledge and strengthening understanding. According to Cohen et al. (2003), educational research as a whole is a relatively new field and currently consists of a largely descriptive element. How much more will this be required when

the educational research is focused on a condition such as CHARGE syndrome, a rare condition only recently identified?

An interpretive approach would be adopted and it was felt more beneficial to work inductively with theory and insights emerging from the research itself, rather than proposing a pre-defined theory to try and prove or disprove through the investigation (Cohen et al. 2003). A case study framework was employed that supported the use of both multiple methods and sources of evidence. A range of sources of data were used to gather evidence for this investigation, including analysis of documentation, and surveys in the form of a questionnaire and interviews.

The nature of the population being explored (with CHARGE being a low incidence condition) meant this would be a small-scale study (Cohen et al. 2003). There are concerns raised about the limitations of small-scale study and whether findings can be widely generalised (Robson 2011), and this research project would need to be undertaken systematically and rigorously (Mason, 2006). Through systematic investigation it was hoped to obtain new evidence which would increase knowledge that can be applied within the field. It is also a belief that theory grounded in real data obtained from the field is more likely to be accepted by those working at 'grass roots level' with the children.

1.6 Terminology and definitions

1.6.1 CHARGE syndrome

As mentioned previously, CHARGE syndrome is a highly complex medical condition first identified in 1979 by Hall and also Hittner et al. The acronym CHARGE was coined in 1981 (Pagon et al.) to represent the anomalies then felt to be the primary characteristics of the condition and to support diagnosis. The medical diagnostic criteria have since been revised in 1998 (Blake et al.) and 2007 (Sanlaville and Verloes) as more has become known about the condition. Previously assigned an 'association', it is now classified as a syndrome. A summary of the major and minor diagnostic characteristics according to Sanlaville and Verloes can be found in Table 1.

The prevalence of CHARGE has yet to be accurately established but there has been some consensus in the estimates that appear in literature. For example, Blake et al. (1998) estimated 1:10,000 – 1:15,000 live births; Vissers et al. (2004) estimated 1:12,000 birth incidence; and Blake & Prasad (2006) estimated 0.1-0.2: 10,000 live births. All these make it clear that CHARGE syndrome is a rare condition.

In literature CHARGE syndrome is often referred to simply as 'CHARGE'. As explained earlier this phraseology will be adopted making reference to this condition as CHARGE, whilst recognising that the full title would be 'CHARGE syndrome'.

1.6.2 Deafblindness/ Multi-Sensory Impairment

Despite ongoing debate, there is a lack of a universally agreed definition of deafblindness. It is generally agreed that this term is used when there exists a

combined visual impairment and hearing impairment, but there is no consensus on the degree of loss and nature of the impairments that comprise deafblindness. The two terms most frequently used in relation to this distinct disability in the UK are 'deafblindness' and 'multi-sensory impairment' (MSI). These two terms are often used interchangeably amongst practitioners involved in educating those children who have combined visual and hearing impairments. This includes those children defined by the Department for Education and Skills (2003) as:

Pupils with multi-sensory impairment have a combination of visual and hearing difficulties. They are sometimes referred to as deafblind but may have some residual sight and/or hearing. Many also have additional disabilities but their complex needs mean that it may be difficult to ascertain their intellectual abilities....

Pupils with multi-sensory impairment have much greater difficulties in accessing the curriculum and the environment than those with a single sensory impairment. They have difficulties in perception, communication and in the acquisition of information. Incidental learning is limited. The combination can result in high anxiety and multi-sensory deprivation. (p.7)

According to a Sense publication in 2012 there are an estimated 31 deafblind children per 100,000 of the population. This illustrates that deafblindness is in itself a low incidence disability, but has a higher prevalence than CHARGE. **Within the deafblind population there is a distinction between those children who are congenitally deafblind and those who have acquired deafblindness. As CHARGE is a condition that causes congenital deafblindness the assumption will be that comparison of learners with CHARGE is to be made to the wider congenital deafblind population unless otherwise stated.**

From the 1950s deafblind education developed alongside other areas of special education. Then in 1989 the Department for Education and Science released a

policy statement identifying deafblind learners as a distinct group that required specialist educational provision. The growth of a deafblind educational philosophy (in other words, identifying its purpose, the role of the educator, what is to be taught and the strategies adopted) has been influenced both by the development of thinking and experience within the field and wider influences (such as a changing population of deafblind individuals and the current inclusion model). Rødbroe and Souriau (1999) describe the early subscription to a behaviourist approach where the children were primarily 'acted upon' to establish a range of skills. Subsequently (since the 1980s) there has been a move away from this to a model where the focus is centred upon a reciprocal relationship with the educator following the child's lead, building positive relationships and facilitating 'active learning' (e.g. Nelson et al. 2010). Rødbroe and Janssen (2006) describe a resolution agreed at a Nordic conference in 1986 that outlined key areas where problems are experienced by deafblind individuals, these being accessing information, communication, and orientation. These are perhaps the key areas addressed by educators for every learner with deafblindness and have subsequently been reflected in the writing of other practitioners in the field (e.g. Aitken et al., 2000) and the development of specialist curricula (e.g. Murdoch et al., 2009). Strategies have developed over time that have become an established part of the deafblind philosophy (e.g. the use of augmentative communication systems), but there also continues to be recognition of the need to continue questioning and develop new knowledge (evidenced through research) to further improve practice.

1.6.3 The educational environment

Ware (2003) describes the learning environment as one that surrounds all normal teaching activities. This reflects a broad context that could include the home, the school and the wider community. For the purposes of this investigation, the aspect of the learning environment to be focused upon is the environment found within the more formal educational setting of the school that delivers a given curriculum. For clarification, within this research study, this aspect to be addressed will be defined as the 'educational environment'.

1.7 Summary and conclusion

In this introduction the experiences and thoughts that have led the researcher to undertaking this research project have been described together with her current involvement with children with CHARGE. The initial thought processes have been described and the development of the research questions outlined briefly that in turn led to the establishment of the research framework. As CHARGE is a rare condition people are often not familiar with, a definition has been provided for clarification together with information on the rate of occurrence within the general population. The majority of individuals with CHARGE will be identified as deafblind/multi-sensory impaired (MSI). An explanation of this terminology has been supplied and its use defined within the context of this research project. Clarification has also been provided for the context that will be focused upon within this investigation, namely the 'educational environment'.

In the remaining chapters the progress of this investigation will be presented in the pursuit of addressing the research question outlined with the intention of ultimately increasing understanding of this phenomenon and improving the practice of educators supporting learners with CHARGE.

CHAPTER 2- CHARGE: A REVIEW OF LITERATURE

2.1 Introduction

As part of this investigation a review of literature was undertaken to gather information and identify previous research undertaken that would help in responding to the overarching research question:

CHARGE syndrome is a medical diagnosis.
Can it also be considered an educational diagnosis?

It was intended this examination of literature would support the development of sub-questions to assist in answering the main research question and provide further structure and clarity to the investigation. Within this review of literature there would be a particular focus on:

- Identifying aspects of this condition that might affect learning
- CHARGE and deafblindness
- The development and functioning of learners with CHARGE
- Educational issues, including assessment

During the process of this review it was found much work has been undertaken in relation to the medical and physical aspects of CHARGE, with clinical research reported in peer-reviewed journals that has supported the increase of the knowledge base about the physical aspects of this condition. Conversely this literature review showed that at present there is a lack of published research in the area of education and learning in individuals with CHARGE with a very limited number of research based papers available. Some related areas, however, have been researched in some

depth (e.g. behaviour, motor development, communication and social interaction) and been published in both peer-reviewed journals and professional journals.

When reviewing the findings of research undertaken in relation to CHARGE it is important to recognise that since it is a rare, low-incidence condition (Blake & Prasad, 2006, quote 0.1-0.2:10,000 live births), many of the studies would be restricted in the number of cases they had access to. This can be put into perspective when considering Blake et al. (2008) who obtained information from the Canadian Paediatric Surveillance Program undertaken over a three year period (2001-2004) and yet only had access to a total of 99 individuals identified with CHARGE across Canada. Any generalisation by researchers of their findings (including this study) would need to be regarded with caution and with consideration to the limited number of cases available to the researcher. In relation to generalisation, it would also be necessary to assess whether the sample used in any research studies was representative of the wider population of individuals with CHARGE; Hartshorne et al. (2007) recognised the heterogeneous nature of this condition and this is also identified again by Hartshorne working with different co-authors in 2011, describing CHARGE as “highly complex” and “highly variable” (p.xiv). This is also likely to make any generalisation between children more difficult and as Hartshorne et al. (2011) explain, there is no ‘typical’ or ‘average’ child with CHARGE. They do, however, also express the view that ‘many similarities can be seen’ (p.xi) suggesting there may be commonalities that can be considered amongst the wider CHARGE population.

The research methodology and methods employed in previous research studies were also considered. Scientific investigation involving clinical tests has been undertaken with regard to some medical aspects of this condition (e.g. Collins and Buchman, 2002; Satar et al., 2003), that provides clinical evidence, particularly in relation to the different medical anomalies. Some use has been made of single and multiple case studies (e.g. Nicholas, 2005; Bernstein and Denno, 2005). Other research has involved document and data analysis (e.g. Strömland et al., 2005; Dammeyer 2012). More predominantly, surveys in the form of questionnaires have been undertaken (e.g. Salem-Hartshorne and Jacob, 2004; Lieberman et al. 2012). This use of questionnaires is likely to be partly a consequence of the low incidence of this condition and the geographical spread creating a practical difficulty in working directly with larger groups of individuals with CHARGE; use of this method is a practical tool that allows for studies of larger numbers of children with CHARGE. Additionally, children with CHARGE are often complex and difficult to assess (e.g. Salem-Hartshorne 2011) and this may be another reason why so many research studies related to the functioning of individuals with CHARGE have used surveys as a method, seeking responses from parents and practitioners. It is important to recognise that understanding of the question will be filtered through the knowledge and experience of the respondent and the responses received from these surveys will often be a record of the person's perceptions. It is very possible the response to a question may vary dependent upon whether the respondent is a parent or a professional.

It is acknowledged primary sources that set out the original presentation of data or evidence may be given more emphasis in a literature review, providing data and thinking in their original form that is likely to be the most reliable presentation of the material. Primary sources were therefore sought out during this review of literature, with many of those having been published in peer reviewed journals and therefore 'judged' (Thomas, 2013, p.60) by peers within the field (e.g. Izzekutz et al., 2005; Lieberman et al., 2012). Secondary sources, however, were also considered as relevant and important, often providing a useful analysis and discussion of the original data and thinking (e.g. Lewis and Lowther, 2004; Hartshorne et al., 2011). It was recognised that interpretation of research in secondary sources may be biased by the writer's viewpoint and there was a risk of data being wrongly or incompletely reported or interpreted; thus the information used would need to be verified by checking the primary source under discussion. For example, in a primary source Peltokorpi and Hettunen (2008) reported:

Their frequency [*of intentional communicative acts*] was rather low compared to the total number of communicative acts (p.39)..... Almost half of the child's communicative expressions could be classified as initiations indicating that the children were actively involved in the interaction session. (p.40)

Whereas, King Miller et al. (2011) reporting on Peltokorpi and Hettunen's (2008) study stated:

That these children initiated communication less than half the time and that their rate of intentional communication was very low. (p.299)

Chapters in edited books were also examined where the editors were experts in the field of CHARGE (e.g. Horsch and Scheele, 2011; Hartshorne et al., 2011). Both types of sources were used since:

There is no automatic correlation between the quality of a source and its 'primary-ness' or 'secondary-ness'. (Thomas, 2013, p.59)

In the eventuality, many of the sources examined provided a combination of both primary and secondary sources (e.g. Brown, 2005a; Lanson et al., 2007). Some review articles were also included as they were considered to provide a helpful summary and potentially be of value in identifying possible primary sources for examination (e.g. Blake et al., 1998).

Overall, it was found necessary to also review articles from periodicals, websites, webcasts and conference proceedings that might be defined as 'grey literature' (Schöpfel and Farace, 2010). These articles do not form part of the commercial publication process and may rely on anecdotal reporting and opinion. Although less rigorous academically, these were considered to also provide valuable information and insights that contribute to the exploration of the issue examined, particularly as many were produced by experienced educational practitioners in the field. Finally, due to the scarcity of materials it would also be necessary to examine older materials that may provide additional information.

2.2 Aspects of CHARGE that may affect learning

According to Hartshorne (2006), interest in CHARGE has increased greatly during recent years. The features of CHARGE identified as major characteristics by Sanlavielle & Verloes (2007), set out in Table 1 in themselves illustrate an extremely complex medical condition which is further complicated by a number of other medical and physical characteristics identified that are also manifest in affected individuals, these being well-documented in the literature available (a succinct summary can be found in Hartshorne et al., 2011). This explains why much research and literature has

focused on the medical aspects of CHARGE. For the purposes of this study, however, the focus will be on those aspects of CHARGE considered likely to affect daily functioning and learning in the educational environment, including sensory issues, low tone, neurological issues, sleep disturbances, medical intervention and later onset features.

2.2.1 Vision

According to Bowman et al. (2001) there are many causes of visual difficulties that will impact upon a child's vision in different ways. Visual issues in CHARGE are reported to predominantly relate to colobomas that often occur on the retina (but may also occur on the iris) and are thought to affect between 80-90% of individuals with CHARGE (Blake & Prasad, 2006; Lalani, 2007; Sanlaville & Verloes, 2007). Other eye conditions also seen in CHARGE include squint, nystagmus, microphthalmic (small) eye, and refractive errors (Hyvärinen, 2011; Russell-Eggitt, 2013).

Bowman et al. (2001) report that visual impairments in children generally affect acuity (how clearly a child is able to see), the child's visual field and/or the processing of visual information in the brain known as Cerebral Visual Impairment (CVI). Hyvärinen (2011) and Russell-Eggitt (2013) explain how the visual anomalies associated with CHARGE can cause visual difficulties with visual field loss and/or poor acuity in one or both eyes, poor contrast sensitivity, and also difficulty with binocular vision. Additionally Russell-Eggitt explains there may be delayed visual maturation with use of vision subsequently improving during the first year of life. There appears to be no other mention in literature of individuals with CHARGE experiencing difficulty with CVI

although there is some discussion of potential difficulty integrating sensory information as a whole that can have an impact on the child's ability to use their residual vision effectively (e.g. Brown, 2010; Hyvärinen, 2011). There was some conflicting opinion about the effectiveness of the correction of refractive errors through the use of glasses provided by two medical paediatric ophthalmic specialists, both experienced in working with children with CHARGE. Russell-Eggitt (2013) suggests that glasses provide only limited benefit and are often challenging to both prescribe and fit; whilst Hyvärinen (2011) describes using corrective lenses to support the child in using their vision more effectively. Brown (2010), an educational practitioner experienced in working with children with CHARGE, supports the latter opinion reporting that children with refractive errors are likely to benefit from glasses being prescribed. In a survey by Deuce et al. (2012) 57% of 44 respondents reported a child to have glasses prescribed, although it was not asked whether the glasses were felt to be beneficial.

Clearly there will be implications arising from the presence of any visual impairment upon the child's learning and development. Sugden (2010) explains the presence of a visual impairment in a child can affect the development of cognition and language, fine and gross motor skills, social and emotional skills and also self-help/independence skills. Bowman et al. (2001) explain that any two children with the same visual condition and acuity levels may each be affected very differently in their ability to function; this suggests it is important for the educationalist to recognise it is not only the physical condition of the eyes that can affect the child's functional visual skills. Bowman et al. (2001) describe factors that may affect a child's functional use of vision, these being the child's motivation, environmental access, effective differentiation of

materials and provision of IT equipment, and finally whether the child is supported to learn to use their residual vision effectively. No evidence was found of research undertaken to explore the impact of the visual difficulties experienced by children with CHARGE, but there is reporting of opinion that identifies additional factors affecting the ability to use their residual vision functionally. Brown (2010) refers to poor muscle tone and postural control (to be discussed in 2.2.6), difficulties with arousal levels (discussed in 2.3.5), and the impact of the child's other sensory impairments. Given the senses are designed to work to support each other (Ayres, 1979) each sensory impairment will have an impact on the use of the other senses. For example, vision is used to help maintain balance, but the vestibular (balance) system (vestibular issues are common in CHARGE- see 2.2.4) also supports the use of vision (Brown 2007b). For the child with CHARGE where most if not all sensory modalities are likely to be affected (Hartshorne et al., 2011), the impact of other sensory impairments, plus additional factors (such as poor physical tone), upon the child's functional use of vision must be recognised and responded to. Hyvärinen (2011) provides a clear example, explaining it is important to determine the impact of different physical positions since the child's posture can support or inhibit effective use of vision and will therefore have significance in the educational setting.

2.2.2 Hearing

There is a wide range of ear anomalies associated with CHARGE and according to Thelin (2011) every auditory structure can be affected. The characteristic 'CHARGE ear' refers to the unusual external ear shape, where the helix may be malformed, with small lobes and the ears often low-set (Lanson et al., 2007); Hartshorne et al. (2011)

report this can be seen in up to 50% of affected individuals. There may be middle ear anomalies including malformation of the ossicular chain and/or persistent otitis media, the latter often being a long term implication of choanal atresia or stenosis (Hall, 2002). Middle ear anomalies are thought to occur in up to 80% of individuals (Hartshorne et al., 2011). The inner ear is also often affected causing a permanent sensori-neural hearing loss in 80-90% according to Thelin and Fussner (2005), Blake & Prasad (2006), and Hartshorne et al. (2011). There can also be anomalies relating to the auditory nerve that may be hypoplastic or absent (Collins and Buckman, 2002). This also means that any assessment with a view to cochlear implantation must begin with the undertaking of an MRI scan to ascertain whether such surgery is viable following careful assessment of the cochlea and auditory nerve (Wheeler, 2013).

Literature explains children with CHARGE can be difficult to assess clinically, for example, Thelin (2011) and Wheeler (2013), who both state the need to adapt the test situation according to the child's other sensory difficulties, current developmental level, communication skills and so forth. This is perhaps a strategy that should be adopted for every aspect of clinical assessment (and again more widely in the educational setting, as advocated by Hartshorne N, 2002). Due to the challenge of obtaining accurate responses in behavioural testing Wheeler (2013) suggests professionals may rely on objective testing where the child is not required to give a response. This however, must also be undertaken with caution since neurological anomalies (as can be present in CHARGE- to be discussed in 2.2.9) can cause abnormal results in Auditory Brainstem Response (ABR) testing (Kraus et al., 1984). Where a child has residual hearing identified there is often a requirement to focus on making best use of

that residual hearing through appropriate amplification (e.g. Thelin, 2002 and 2013). Amplification may be regarded as important to support greater access to auditory information and the curriculum, but it is necessary to recognise that the fitting of post-aural hearing aids and achieving well-fitting earmoulds can be affected by the physical ear anomalies described previously, that in turn create difficulties in providing consistent good amplification for the child.

For the educator it is important that recognition is given to the impact a hearing impairment can have upon the child's learning and engagement in the educational setting. Gregory et al. (2002) explain that hearing impairment affects the development of receptive and expressive language, cognition and social-emotional development. These in turn can affect the child's communicative abilities and academic and social success (Marschark and Hauser, 2012).

It was noted in 2.2.1 that within literature there is recognition a child's use of vision may be affected by factors other than the physical eye condition. There is no apparent similar statement made with regard to the child's functional auditory skills but it would seem highly likely the same line of argument would apply to the child's functional use of hearing; that their other sensory impairments and additional factors (e.g. neurological anomalies) are also likely to impact on the child's ability to use their residual hearing to good effect. Therefore information on the physiological aspects of the child's hearing and an identification of the hearing loss alone will not provide the educator with a full understanding of how the child is likely to be able to access auditory information within the educational setting.

2.2.3 Co-occurring visual and hearing impairments

As explained previously, Hartshorne et al. (2011) reported over 90% of individuals with CHARGE will have both visual and hearing impairments. It is not clear if this is based on evidence arising from research or from an assumption resulting from the identified rate of occurrence of both separate sensory impairments. The fact that both these anomalies occur so often in affected individuals does indicate a large number are likely to have both a visual impairment and hearing impairment and will therefore be considered as deafblind/ multi-sensory impaired.

The presence of combined visual and hearing impairments will have clear implications for everyday functioning and gaining access to the curriculum as for other deafblind learners. According to Aitken (2000a) they will create challenges in accessing information, communicating with others and moving around their environment, and there may also be difficulty processing the information received. Thus there is a potential significant impact upon learning for the child who has co-occurring visual and hearing impairments that needs to be recognised and responded to. For the child with CHARGE there are also likely other potential factors creating additional challenges to learning and everyday life that will also need to be acknowledged and responded to appropriately.

2.2.4 Vestibular issues

A number of studies undertaken have identified the presence of structural vestibular anomalies (absent or under-developed semi-circular canals) in the majority of individuals with CHARGE (e.g. Collins and Buchman, 2002; Satar et al., 2003). The

fact that research has shown vestibular anomalies to be present in over 90% of individuals with CHARGE (as stated by Hartshorne et al., 2011) supports the view that vestibular dysfunction is regarded as largely inherent to CHARGE (e.g. Brown, 2003b). One identified consequence is a general delay in the development of gross motor skills. A study by Travis and Thelin (2007) found the average age for independent walking in their cohort with CHARGE was 42 months (compared with the typical age of development being 15 months according to Sheridan et al., 2008), deemed to be a direct consequence of vestibular difficulties. A potential further impact of the delay in the development of gross motor skills was found in an earlier study by Thelin and Fussner (2005) where they were surprised to note a relationship between the acquisition of symbolic communication and the child's ability to walk independently.

Thelin et al., (2011) recognised that a growing understanding of the impact of vestibular difficulties has come from observation of behaviour rather than objective testing. One practitioner who has provided a number of articles on this subject is Brown (e.g. 2003b, 2009), building on the work of Ayres (1979) who explained:

The vestibular system is the unifying system. (p.37)

Ayres (1979) described the vestibular system as underpinning information received through all the other sensory modalities, and considered difficulties with the vestibular sense would affect the whole nervous system. Research outlined above has identified the impact vestibular difficulties are likely to have on the acquisition of gross motor skills (and potentially the development of symbolic communication), but observations by Brown (e.g. 2007b, 2011b) outline the potential for it to greatly affect all aspects of

daily functioning since vestibular difficulties are likely to have some bearing on not only postural control and mobility but also effective use of vision and hearing and daily functioning. Brown (2011b) goes further to assert that in the future it is possible some CHARGE behavioural features of this condition that are currently “unexplained” (p.53) are likely to be linked to these vestibular difficulties. Given that vestibular function is so important for underpinning the reception and understanding of all other sensory information (Ayres, 1987) it is very possible the full implications of vestibular dysfunction for the individual with CHARGE have yet to be identified.

2.2.5 Proprioception

According to Brown (2005a) the word ‘proprioception’ derives from a combination of two Latin words and means an awareness or a feeling of one’s own self, with Davenport and Hefner (2011) believing the term was coined by a CS Sherrington (1907) to refer to the sensations received from the tendons, muscles and joint movement. Whatever the origin of this word, it is a term now widely accepted to describe the information received from receptors in the tendons, joints and muscles that make a person aware of their body position in space and where the body parts are in relation to each other (Bogdashina 2003).

Although no research could be found that had been undertaken in relation to this aspect, observations have been reported that imply poor proprioceptive abilities in individuals with CHARGE, particularly by Brown (2005a) who noted behaviours including a difficulty maintaining posture when in an upright position with a need to seek additional support (e.g. by leaning against furniture, propping the head on a

hand), uncoordinated movement, foot stamping when walking, or conversely walking on tiptoes, and seeking input from strong, firm pressure. This sensory input is important to provide good body awareness and help coordinate body movements (Ayres, 1979), and a poorly developed proprioceptive sense such as has been suggested in individuals with CHARGE (e.g. Brown, 2005a; Davenport and Hefner, 2011) will have a clear impact on the individual's ability to function effectively. Ayres (1979) stressed the importance of a well-developed proprioceptive sense to enable the child to know where their separate body parts are and how they are moving; without this Deuce (2002) considered:

They are unlikely to be able to effectively use their body to interact with, or act upon, their environment and learn. (p.10)

2.2.6 Low tone

Hypotonia, and poor tone in the upper body in particular, is also a common feature highlighted (e.g. Williams, 2011). This is identified in medical papers relating to CHARGE where, for example, Thelin & Fussner (2005) found 81% of individuals in their study were affected. Williams (2011) explains hypotonia will affect posture and motor development, making it more difficult to maintain a good upright position (e.g. sitting or standing) and delaying independent walking. Although hypotonia is reported in many individuals with CHARGE the potential cause has not been identified through research, although Williams (2011) does stress it is not a result of an abnormality in the muscles themselves but rather suggests it may be a result of an irregularity in the central nervous system. Brown (2003b), an educationalist, suggests hypotonia is partly a function of severe balance problems and is associated with low vision and “generally reduced sensory systems” that cause “reduced perceptual awareness”

(p.2). Brown (2003b) is one of the few educationalists to consider the impact of this in relation to daily functioning following his involvement with a number of different individuals with CHARGE; from his experience he reports he has found it can reduce perceptual awareness and may compound a lack of motivation to move caused by both vestibular and visual difficulties. Lieberman et al. (2012), who investigated physical education in learners with CHARGE, reported that low tone can result in difficulty maintaining contractions and in shifting body position. This is likely to have an impact on the child's ability to maintain a good working position, and it is possible their efforts may be taken up in maintaining their body position (these difficulties will be compounded if there is also vestibular dysfunction) so they are therefore not released to concentrate on other learning. This may be an issue that lies behind Williams' (2001) suggestion that adaptive (supportive) seating can be beneficial.

2.2.7 Touch

According to Davenport and Hefner (2011) touch becomes an important and often primary sensory channel for receiving information for the child with CHARGE, who may also be:

Particularly sensitive to textures and may have preferences for or aversions to particular sensory experiences. (p.8)

It is not clear what evidence this statement is based upon and practitioners need to beware of focusing primarily on providing sensory information through the tactual sense or assuming the presence of any tactual sensitivity without acknowledging other potential influencing factors. It is important consideration is given to the possible impact of other sensory issues and anomalies associated with CHARGE that may adversely affect the child's ability to use their sense of touch effectively. For example,

the importance of vision in supporting the development of a child's haptic perception (that is the active use of touch to explore and gain tactual information) is explained by McLinden and McCall (2002) who state where there is a significant visual impairment:

The information that young children receive through touch may be fragmentary and difficult to assimilate. (p.55)

This is not an aspect addressed in literature with particular reference to children with CHARGE but needs to be acknowledged given that visual impairment is reported in 80-90% of individuals with this condition (Hartshorne et al. 2011). Touch is also reported to be vitally important to individuals who are deafblind where the focus is often placed upon the child's hands. Miles (2003) explains the child's hands, in addition to being a tool and a sensory organ, often need to become the child's voice; she stresses the importance of therefore being very respectful when touching the hands of any deafblind child.

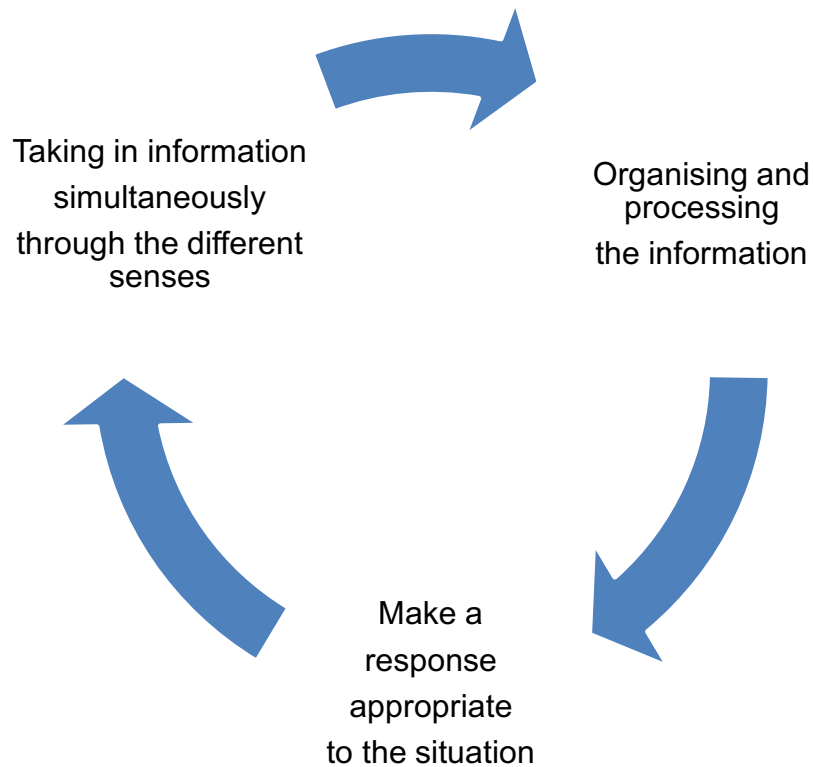
In addition to haptic perception, also referred to as 'active' touch, there is also 'passive' touch where the child receives tactual experiences through their skin coming into contact with a person or object, but without any active engagement on their part (McLinden and McCall, 2002). It is important to recognise that skin is the largest human organ and a child will not only receive tactual information through their hands, feet, mouth and other body parts intentionally used to seek tactual information, but will also receive a good deal of tactual information through passive touch over their whole body. Davenport & Hefner (2011) explain that although it is not known whether individuals with CHARGE have an altered perception of touch, they often demonstrate behaviours that suggest a preference or aversion to some sensory experiences. They also report these children often prefer to receive touch through firm, deep pressure,

this observation being shared by Brown (2005a) who reports many children with CHARGE prefer to be touched in a firm manner rather than light touch, the latter being thought to be uncomfortable or painful for the child. Brown (2005a) relates this to the child's proprioceptive sense being under-developed, resulting in the child needing firmer and deeper pressure to receive enough tactual sensory information in a way that is not irritating or painful. These examples relate to the child receiving information through passive touch, but Deuce (2002) and Brown (2005a) also consider that as a result of a poorly developed proprioceptive system, a child may also use additional pressure when touching, grasping etc. to gain tactual information at a level of intensity that is meaningful to them. In addition to the obvious implications, this can affect social relationships if the child accidentally inflicts pain (e.g. by holding on too tight etc.) without realising what the other child might be experiencing (Deuce, 2002).

2.2.8 Sensory integration dysfunction

Sensory integration dysfunction is considered by some to be inherent in CHARGE (e.g. Brown 2003b). Both 'sensory integration' and 'sensory processing' are terms used in literature to describe the theory proposed by Ayres (1979), a neuroscientist and occupational therapist. Sensory integration is described as the process of organising sensory input received from the different sensory modalities to enable the individual to operate on their environment (Ayres, 1979). This process is vital in enabling each one of us to develop awareness of our self and our environment and allowing us to engage effectively with our world and can be illustrated in the diagram shown in the following Figure A (devised by this researcher).

FIGURE A: THE SENSORY INTEGRATION PROCESS



Sensory integration dysfunction (or sensory processing difficulties) are thought to occur when there is difficulty receiving accurate sensory information through any of the senses that affects the child's ability to process it and make appropriate responses (Dunn, 1999). Given that potentially all seven sensory systems can be impaired in learners with CHARGE (Hartshorne et al., 2011), this may be a reason why so many individuals with this condition are thought to be affected as they receive mainly fragmented and distorted sensory information. Hampson (2013) explains children with CHARGE often have difficulty using their senses together in a coordinated way and therefore experience sensory integration dysfunction. There have been a number of articles written over time that discuss sensory integration difficulties in relation to individuals with CHARGE (e.g. Brown 2003a, 2003b; Thelin & Fussner 2005; Lauger 2007; Hampson, 2013), but no clear research undertaken.

Parham and Mailloux (2010) and Kandel et al. (2000) agree a number of functional impairments can result from experiencing sensory integration difficulties which may have an adverse effect on the child's learning, these being:

- Poor social skills (including play)
- Poor self-confidence and self-esteem
- Poor fine and gross motor skills
- Lack of the development and use of adaptive responses
- Poor daily life skills

This examination of literature found sensory integration dysfunction is becoming more widely acknowledged in the medical world as illustrated by the policy statement on sensory integration presented in 'Pediatrics', the official journal of the American Academy of Pediatrics (2012). There does however, appear to be some contention as to whether sensory integration difficulties can be regarded to be a distinct condition or whether they only co-occur with other conditions (Ahn et al., 2004; American Academy of Pediatrics, 2012). Although sensory integration difficulties may be regarded as inherent in individuals with CHARGE it is not clear what the rate of incidence is. This is also by no means the sole group of children where these difficulties might be found; within the population of typically developing children Ben-Sasson et al. (2009) found sensory integration difficulties in almost 1:6 6-11 year olds, whilst Ahn et al. (2004) reported sensory integration difficulties in 1:20 of 5-6 year olds, although they extrapolated a figure of approximately 1:7 if the assumption was made that:

Non-respondents' rates were equivalent to respondents' rates. (p.291)

These studies used different assessment tools for measurement that make it difficult to compare the results, but both suggest it is a pertinent issue to be addressed within

the typical school population. This is even more so for children with special needs if we accept the view of Ahn et al. (2004) who state:

Estimated rates of sensory processing disorders for children with various disabilities have been derived from reliable and valid survey results and are reported to be as high as 40-88%. (p.287)

It is unfortunate that all five studies then cited are focused on the population of children with autism, although this does reflect the large amount of interest shown in sensory integration dysfunction in this particular population (e.g. Bogdashina, 2003). Some research in the field of sensory integration dysfunction has been undertaken with different conditions, such as that by Miller et al. (1999) who identified the presence of sensory integration difficulties in a sample of individuals with Fragile X syndrome. This indicates it should be possible to undertake a similar investigation to ascertain the rate of incidence in a population of children with CHARGE that might support (or disprove) the anecdotal reporting by practitioners working with learners with CHARGE.

In the work by Bogdashina (2003) she argued children with autism may experience different sensory experiences arising from their autism. Perhaps this interpretation can be transferred to children with CHARGE; that their sensory integration difficulties are in part rather than they have different sensory experiences, this being more marked because of the possibility of most, if not all, of the sensory modalities being affected. This is likely to have a significant impact upon the way each individual is able to perceive, and engage with, the world around them and this in turn affects how they modulate their own sensory systems and self-regulate, an aspect of sensory integration dysfunction that seems particularly prevalent in the population of children

with CHARGE according to literature (e.g. Brown 2003a, 2003b; Thelin & Fussner 2005; Lauger, 2007; Hampson, 2013).

The wider acceptance of sensory integration dysfunction as a condition may result from the fact that, although Ayres theory appears not be based on scientific data, there is increasing research that indicates evidence of a relationship between physiological differences and the presence of sensory integration dysfunction. McIntosh et al. (1999) undertook a construct validity test of their 'Short Sensory Profile' and demonstrated an association between the functional symptoms of sensory integration dysfunction measured through this profile, and physiological changes in the form of electrodermal responses (a change in the electrical properties of the skin as a response to anxiety or stress).

Within literature there is some disagreement about the effectiveness of the therapy treatments involving activities designed to stimulate the sensory systems and support the integration of a child's sensory, motor, cognitive and perceptual skills, focusing primarily on the vestibular, proprioceptive and tactile sensory systems (Dunn, 1999), rather than with the theory itself. For example, Goldstein (2000) disputes the evidence provided from some research indicating that this therapy is effective. His argument illustrates the long-standing debate about the recognition of the validity of research methodology employed within the social sciences rather than the more traditional scientific approach to research (Robson, 2011). Edelson et al. (2003) refute Goldstein's argument, defending their own research which found sensory integration interventions to be effective. Their response is, however, somewhat undermined by a

lack of information regarding their research process (e.g. no outline of participant selection and no clear description of the research methods used or tools used for measurement). There is, however, also other research such as that presented by Fertel-Day et al. (2001) where positive effects of sensory integration therapies were found; in this study the use of a weighted jacket was found to result in the child becoming less distractible with fewer self-stimulatory behaviours observed.

No specific research has been undertaken that focuses on which aspects (if not all) of the therapy approach would be most beneficial for learners with CHARGE and the impact of their use. Any such research would require the involvement of a physiotherapist or occupational therapist with expertise in the field of sensory integration dysfunction and qualified to undertake a sensory integration assessment (Hampson, 2013).

2.2.9 Neurology

There is some evidence the formation of the brain may be altered in a significant number of individuals and a clinical study by Raqbi et al. (2005) found 17/21 children studied had cerebral imaging that was atypical. Lauger (2007), drawing on work by Elliot (1999), explains that disruption to the sensory systems in utero (as occurs in CHARGE) also affects the neural pathways which connect the sensory systems to the brain. Neurological anomalies may have been anticipated once the mutation of the CHD7 gene had been identified (as explained earlier in 1.1) since this influences the development of all major organs in the body. Several neurologic conditions have been found in CHARGE relating to abnormalities in the embryonic development, including

epilepsy, cranial nerve dysfunction (discussed in more detail in 2.2.10), sleep disorders, cognitive impairment and global developmental delay (Gilles, 2011).

In addition, executive function deficits have been identified (Nicholas, 2005; Hartshorne et al., 2007) that can potentially impact upon an individual's ability to function, learn and develop. The term executive function (also known as 'cognitive control'), is a cognitive process that may be broadly defined as the individual's ability to "control cognitive actions" (Richland and Burchinal, 2013, p.87). As the name suggests, they are higher-level cognitive abilities which control and regulate other abilities and behaviours and are necessary for goal-directed behaviour (Nicholas 2005). Research undertaken by Nicholas (2005) involved a single case study making it difficult to generalise any findings to the wider CHARGE population, but later research by Hartshorne et al. (2007) studied 98 individuals with CHARGE. The results of this latter study were presented to show the mean scores for different executive function tasks measured within the scales employed, with the researchers reporting that over half "received clinically significant scores" (p.333). From the data presented it is not possible to gain a more specific figure, but these findings suggest executive dysfunction may be present in a significant number of individuals with CHARGE. In typical foetal development the brainstem and midbrain responsible for autonomic (bodily) functions are the first to form and are well-developed at birth. In contrast the higher regions, including the limbic system and cerebral cortex, develop later and are still quite primitive at birth (Linderkamp et al., 2009). It is the frontal lobe (one of the four major lobes in the cerebral cortex) that is responsible for executive function (Miyake et al., 2000), and it is possible this development is affected by the faulty

regulation of development caused by the CHD7 gene anomalies described in 1.1, explaining why executive function difficulties are highlighted in a significant number of individuals with CHARGE. Cooper-Kahn and Dietzel (2008) provide a clear and detailed description of the complex skills involved in executive function that are summarised in Table 2 below:

TABLE 2: A SUMMARY OF THE SKILLS INVOLVED IN EXECUTIVE FUNCTION

- Inhibition: the ability to control one's own behaviour and stop actions/ thoughts, with the opposite being impulsivity
- Shift: moving with ease from one activity or situation to another, and to work flexibly
- Emotional control: modulating one's emotional responses
- Initiation: the ability to start an activity, generate ideas and engage in problem- solving
- Working memory: the ability to retain information to support the completion of a task
- Planning and organisation on oneself: to be able to manage a task currently engaged in and to also plan ahead
- Organising materials: imposing order on work and play activities
- Self-monitoring/ self-regulation: monitoring one's behaviour and performance with a recognition of what is needed or might be expected

Cooper-Kahn and Dietzel (2008)

It is interesting to note that when comparing these aspects to the results from the study by Hartshorne et al. (2007), the sample examined achieved a score that reflected difficulties in all these areas except 'organising materials' where a clinically significant score was not obtained. One additional point worthy of consideration is that according to Nicholas (2005) and Hartshorne et al. (2007), the abilities to form concepts and think abstractly are often considered components of executive function. During this literature review it was found this subject was not readily addressed with

regard to the broader deafblind population and this may be an indicator it is more prevalent in learners with CHARGE. Understanding these difficulties result from neurological anomalies (and the development of the frontal lobe) implies, however, that other groups of individuals (including deafblind) may also be affected despite the lack of evidence in literature. One article written by Nicholas (2000) (who also identified executive dysfunction in individuals with CHARGE) was found which supports this; this relates to neurological functioning in individuals with Congenital Rubella syndrome (CRS) who may also exhibit impaired executive functions. In contrast to individuals with CHARGE, however, Nicholas felt that in the CRS population this is a later onset and a consequence of the damage to the central nervous system caused by the Rubella virus that:

Has no physiological or psychological effect until teen or adult age. (p.20)

This damage to a developed system in CRS is thus different to the likely cause of executive dysfunction in CHARGE which is due to anomalies with the formation of the brain; thus it is possible each separate group may be affected differently as a consequence. In literature on CHARGE it is suggested executive dysfunction has an impact on the child much earlier with the signs of difficulties emerging in the child as they begin to experience abstract concepts and become more independent in their learning (Hartshorne et al., 2007). Consideration of the skills involved in executive function can highlight the potential impact executive dysfunction may have on learning and development for the learner with CHARGE. If a child is experiencing executive function difficulties they may demonstrate difficulties in their social and emotional development; in monitoring and regulating their own behaviour; managing transitions between activities, different environments and people; working independently and

initiating activities and interactions; completing a task independently; organising oneself and the environment; maintaining a well-regulated sensory state (that may be further impacted by the presence of sensory integration difficulties discussed in 2.2.8); and forming clear concepts, especially when they become more abstract.

2.2.10 Cranial nerve anomalies

There are twelve pairs of cranial nerves, these being those nerves that emerge directly from the brain (in contrast to the spinal nerves). Certain cranial nerves have been identified as being affected in CHARGE and are included in the diagnostic criteria, these being listed in Table 3 together with a description of their function.

According to Gilles (2011) the presence of cranial nerve anomalies create:

Some of the greatest disease burdens in CHARGE. (p.141)

The potential severity of the implication of these anomalies on a child's learning and development becomes apparent when recognising these nerves facilitate sensory functioning and/or muscle control of different senses in the body. If a proposal by Blake et al. (2008) is accepted who, following an examination of clinical data gathered through a questionnaire to physicians for 99 individuals with CHARGE, suggested the other cranial nerves could also potentially be affected, this would have implications for vision (CNII, CNIII, CNIV and CN VI), touch and pain sensations in the face and head (CNV), and head movement (CNXI). All of these may affect the way the child with CHARGE functions and is able to engage with their world, and ultimately therefore affect learning. There is likely to be a difficulty in accurately identifying the presence of some of these cranial nerve anomalies through clinical testing that can be difficult to implement with this particular population, and reflects an issue likely to be frequently

encountered across the spectrum of research undertaken with individuals with CHARGE.

TABLE 3: THE CRANIAL NERVES INCLUDED IN THE DIAGNOSTIC CRITERIA FOR CHARGE

Cranial nerve (CN)	Function
CNI	To transmit smell
CNVII	Innervates the facial muscles (and therefore facial expressions), is involved in the sense of taste and controls the stapedius muscle in the ear that protects the ear from sudden loud sounds
CNVIII	Responsible for both hearing and balance
CNXI	Involved in swallowing and tasting
CNX	Involved in taste and controls the voice muscles as well as controlling the heart rate and digestion (Ref: Shipley and McAfee, 2008)

For example, a clinical study by Bergman et al. (2011) identified a relationship between anosmia (reduced or lack of sense of smell), as a consequence of anomalies with CNI, and hypogonadotropic hypogonadism (lack of production of the sex hormone). Although their findings may not be regarded as pertinent to education, it was interesting to note that as a consequence of using a smell test to identify hyposmia or anosmia as part of their study, individuals who were below the age of 5 years or with a low developmental age were excluded because of the difficulty

implementing the smell test. This raises the question of whether their sample could be considered truly representative of the whole CHARGE population, but is perhaps more helpful in demonstrating the difficulty that can be encountered in the research process involving individuals with CHARGE. According to Bergman and Ravenswaaij (2007) smell is probably one of the most under-rated senses. It is important that educators acknowledge the impact this may have for the child for, in addition to providing an alarm function, it is important for social interactions and is closely linked to emotion (Brown, 2007a) so will have a clear impact on the development of individuals with CHARGE and the formation of relationships. Similarly, CNVII anomalies can result in facial palsy (reported by parents to be present in 50% of the cohort studied by Deuce et al., 2012), and whilst this can affect speech and feeding, there are wider implications since it is likely to have an impact on social and emotional development when the child has difficulty smiling and showing facial expression.

2.2.11 Sleep disturbances

Hartshorne et al. (2008) undertook research that involved the collection of information on 87 children with CHARGE (a relatively large cohort for research undertaken within the CHARGE field). Using a sleep measurement score they found 57.5% of the cohort gained sleep disturbance scores that fell within in the clinical range. Heussler (2011) identified three possible reasons for a disturbed sleep pattern experienced by individuals with CHARGE, namely obstructive sleep apnea, a visual impairment affecting the Circadian (sleep) rhythm, and difficulty with sleep initiation and maintenance. Whatever the reason, Heussler (2013) stated that:

Sleep problems in any child have strong relationships to behaviour and function. (p.1)

This relates to the findings of Hartshorne et al. (2008) which showed a relationship between sleep disorder and “challenging behaviour” (p.6), particularly in the form of self-absorbed behaviour, that will need to be managed in the educational setting. Hartshorne et al. (2008) make the insightful suggestion that rather than the primary focus being on management of more difficult behaviours it may be more beneficial to address any sleep issues first.

Other research shows such sleep disturbance is not unique to CHARGE, and sleep disturbance scores in the clinical range have also been reported with regard to other syndromes (e.g. Cotton and Richdale, 2006). Although Hartshorne et al. (2008) discuss the impact of sleep disorder on the parents of an affected child this should also be considered with regard to the impact on the child’s time in school. A disturbed sleep pattern may adversely affect school attendance and the child’s ability to function in the educational environment as a result of fatigue and irritability, and may be manifested through the child’s behaviour.

2.2.12 Medical intervention, hospitalisation and pain

Due to the large number of serious anomalies related to this condition the early weeks and years of a child born with CHARGE can be medically extremely difficult (Dammeyer, 2012). Souriau et al. (2005) found 80% of the cohort studied (57 children) had been in hospital at least once during the first three years of their lives, with the average cumulative length of stay for these children being 194 days and the average number of surgical interventions being 5.2. Deuce, et al. (2012) also explored the period of hospitalisation immediately following the child’s birth for 44 children and

found the length of stay for these children ranged from one week to three years with the average length of stay being 91 days. Overall these studies illustrate the difficult and potentially traumatic start to life for many of these children that may well have:

Significant consequences to their later development.
(Souriau et al., 2005, p.279)

A related issue is the experience of pain. Davenport and Hefner (2011) report perception of pain could be altered in children with CHARGE, this statement possibly resulting from earlier interviews with parents (e.g. Davenport, 2002a) where parents reported their children with CHARGE appeared to have a very high pain threshold. Davenport and Hefner (2011) raise the point that children with CHARGE experience frequent pain (from surgeries, other medical intervention and repeated illnesses) and it is not clear whether these children actually have a higher pain threshold or whether they have:

Simply accommodated to the experience of pain. (p.9)

Nicholas (2011) presents a different view. He also explains individuals with CHARGE often experience chronic pain, but suggests it may be the child is not able to communicate effectively when they are in pain, taking longer to process information and therefore responding more slowly to a painful experience, but also having difficulty locating accurately where pain is being felt. Nicholas (2011) feels the challenge is for those supporting the child with CHARGE to:

Be able to accurately identify and recognise pain signals. (p.347)

This is an important point since if a child is experiencing chronic pain that is not recognised or treated it is likely to have an impact not only upon their behaviour, as noted by Hartshorne (2011), but also on their ability to attend to external stimuli and interact with their environment. Stratton (2013) reported on a research study

concerning the development of a non-verbal assessment tool of pain in individuals with CHARGE involving observations of behaviour, which may be of benefit to practitioners and parents. This assessment tool is an example of how a tool might be developed that can be used across the broad population of children with CHARGE and has the advantage of being standardised for this specific population.

2.2.13 Later onset features

Following the identification of CHARGE in 1979, more recently the first large group of identified children diagnosed with this condition are now reaching adolescence and adulthood. As a result more information is becoming available about individuals in these age-groups with longer-term manifestations of the condition beginning to emerge. Whilst children with CHARGE experience many medical challenges during their early years of life there is evidence emerging that as they age these individuals face an evolving set of medical issues (e.g. Searle et al., 2005; Blake et al., 2005; Blake and Prasad, 2006). These include scoliosis, pubertal delay, migraines (particularly abdominal migraines), epilepsy, endocrine deficiencies leading to issues with osteoporosis and obesity, sleep apnea, cataracts, retinal detachments, progressive hearing loss and renal failure (also see Searle et al., 2005; Issekutz et al., 2005; Hartshorne 2006). The presence of later onset features implies it is necessary to monitor the individual carefully to identify any developments, not only to ensure the individual remains in good health but also because these can be factors that influence an individual's behaviour (Blake and Salem-Hartshorne, 2011) and ability to function in the educational environment.

2.2.14 Summary

The literature examined clearly shows CHARGE to be an extremely complex condition where all sensory modalities are likely to be affected in addition to many other anomalies. This is reinforced by the more recent identification of anomalies with the CHD7 gene, belonging to a family of regulation genes which potentially affect a large number of the developmental pathways, explaining why so many organ systems are involved with CHARGE (Sanlaville & Verloes, 2007). When exploring the different anomalies associated with this condition it becomes clear some might also be seen in the wider deafblind population and beyond, for example, the presence of sensory integration issues, vestibular difficulties and hypotonia, in addition to the obvious co-occurring hearing and visual impairments. There are, however, other anomalies that appear to be more specific to CHARGE, including cranial nerve anomalies and neurological anomalies creating executive function difficulties, and it may be this is an element that creates a distinction between individuals with CHARGE and the rest of the deafblind population. According to Davenport (2002a) it is important to understand the interaction of the multiple anomalies present in this condition. The interaction and 'knock on' effect of each of these features in CHARGE can be clearly demonstrated, for example, when considering difficulties with the vestibular sense that affects effective use of residual vision and hearing (Brown 2003a; Hampson, 2013); and the development of motor skills which in turn affects the development of communication and symbolic language (Thelin and Fussner 2005).

All of the above, combined with the way these anomalies impact upon each other, can create barriers to learning and cognitive development and are an indicator that

individuals with CHARGE are contending with a wide number of issues. For example, receiving limited and distorted information; difficulties in organising their body effectively to act on their environment and engage in active learning; in communicating effectively with others; in gaining experiences important for concept development, and difficulty with the development of more abstract concepts. These are challenges recognised for the deafblind learner (Aitken, 2000a), but with all the sensory modalities potentially affected (and if not all, then many) the impact of these on the development and learning of the child with CHARGE may be intensified and more complex.

The combination of anomalies seen will be distinctive in each affected child, creating a unique set of needs. The challenges to development and learning the presence of these anomalies creates must be considered with regard to the educational needs of a child with CHARGE and it was therefore considered appropriate to continue to explore literature to examine research and thinking in relation to the development and functioning of affected individuals.

2.3 Development and functioning in individuals with CHARGE

Whilst there was no research identified within this review of literature that clearly focuses on the overall educational needs of individuals with CHARGE, certain aspects of development and functioning and the curriculum have been addressed that may usefully inform those involved in supporting these children and young people educationally.

2.3.1 Gross motor development

The delay in the development of gross motor skills in children with CHARGE is considered to be “almost universal” (Gilles, 2011, p.140) in relation to the main motor milestones usually established before the age of four years. For example, research by Thelin and Fussner (2005), Graham et al. (2005), Hartshorne et al. (2007), and Sanlavielle and Verloes (2007) all found independent walking to occur between 35-57 months. Given the presence of visual impairment in a large percentage of the population with CHARGE it is also pertinent to compare this with the achievement of this motor milestone in blind/visually impaired children, reported to occur at between 19-20 months (e.g. Levtzion-Korach et al., 2000). This suggests the delay in independent walking in children with CHARGE is caused not only by the presence of a visual impairment but also additional factors such as vestibular difficulties and hypotonia also cited in literature for this population (e.g. Graham Jr. et al., 2005; Sanlavielle and Verloes, 2007; Lieberman et al., 2012).

It is interesting to note that in earlier literature relating to CHARGE, Harvey et al. (1991) felt early gross motor delay was not necessarily indicative of overall poor development. Other studies, however, by Blake and Brown (1993) and Blake et al. (1998) identified motor difficulties as a factor likely to affect a child’s overall development (combined with sensory and other physical impairments). This was expanded upon in a study by Salem-Hartshorne and Jacob (2004) who obtained data from 100 questionnaires using an adaptive behaviour scale to gather information about the child’s developmental functioning. When analysing the data they attempted to identify factors that might be associated with the child’s developmental progress

and found that age of walking showed the strongest relationship with overall development. Then in 2007, Wachtel et al. undertook research involving 87 participants in a survey; they too found age of walking might be an indicator of potential developmental difficulties (i.e. the later a child with CHARGE achieves independent walking the more likely they will experience greater developmental delay); thus concluding:

The age at which the child first walks has been found to be a marker of how well the child with CHARGE develops. (Wachtel et al., 2007, p.479)

Unfortunately there is no discussion of what underlies this relationship, although Salem-Hartshorne and Jacob (2004) suggest it is necessary to develop activities to implement in the home and school to support the development of gross (and fine) motor skills. A study by Lieberman et al. (2012) explored the delivery of physical education for learners with CHARGE, and whilst the focus of the study was on teaching strategies (that will be discussed later in 2.6.2) they clearly regarded motor delay (occurring as a result of sensory impairments, poor tone and ligamentous laxity) as a key issue to be addressed in the educational setting to maximise a child's learning outcomes. It is possible this viewpoint was reached from the evidence arising from the research studies outlined above. These findings may be useful to the educationalist but a note of caution is raised by Davenport and Hefner (2011); they report there is a risk that where the developmental milestones (including motor development) are delayed, there is often an assumption made that intellectual disability is also present, although in their opinion Davenport and Hefner (2011) feel:

Usually, this is a wrong assumption. (p.6)

The latter is perhaps rather a blanket statement for such a heterogeneous population but raises an important point, this being that educationalists need to be wary of making assumptions that may limit expectation of what a child is able to achieve.

Thelin and Fussner (2005) identified the potential importance of independent mobility for the development of communication in a survey of parents of 29 individuals with CHARGE who were asked to judge the impact of different factors relating to communication development, specifically symbolic language. They found 16/17 participants who had developed symbolic language were able to walk independently, but only 3/11 participants who had not developed symbolic language were able to do so. As a result of this they reported:

The ability to walk independently was related significantly to the development of symbolic language (p.288)

Petroff (2013) reached a similar conclusion in a study of 97 deafblind individuals, also reporting a relationship between independent walking and the development of symbolic communication, although not identifying any potential causal link. Other research in the field of CHARGE found later age of walking was related to challenging behaviour (Hartshorne and Cypher, 2004); greater executive dysfunction (Hartshorne et al., 2007), an increase in 'autistic-like' behaviour (Hartshorne et al., 2005a), a difficulty developing adaptive skills (Salem-Hartshorne and Jacob, 2005) and sleep difficulties (Hartshorne et al., 2008), indicating that:

Age of walking is a marker for more serious developmental complications of CHARGE. (Salem-Hartshorne and Hartshorne, 2011, p.232)

Whilst identifying a link between independent walking and aspects of development that will have implications for learning, on the whole these researchers do not identify

any causal link with the exception of Thelin and Fussner (2005) who hypothesised that independent mobility was necessary to help a child with CHARGE overcome the hearing and visual impairments associated with the condition, enabling them to achieve the most effective position or location to receive communication. This may appear to be a reasonable assumption but, although a link has been identified between independent walking and symbolic communication, there is no evidence from other studies to support this initial hypothesis. A more measured view is taken by Salem-Hartshorne and Hartshorne (2011) who reflect it may be that independent walking facilitates increased opportunities for social interaction and communication as well as enabling a more accurate perception of the world (from an upright position), but that equally a child may not achieve independent walking so readily as a consequence of the impact of other anomalies and it may be these that also affect the child's development (including communication). In other words, although a link has been established it is not yet clear what causal relationship there might be.

2.3.2 Communication and social interaction

The development of successful communication and social interactions would generally be regarded as a vital requirement for a child to learn effectively. Early literature identified a wide variation in the development of communication skills in children with CHARGE (e.g. Goldson et al., 1986) and it is now recognised these children will very often demonstrate delay in developing both expressive and receptive communication skills (e.g. Brown, 2005b; Peltokorpi and Huttenen, 2007), with King Miller et al. (2011) going as far as to say that:

Expressive communication skills in all of these children are delayed.
(p.295)

Peltokorpi and Huttenen (2008) stated that hardly any reporting of communication in children with CHARGE had been made. Further investigation, however, reveals it may have been more accurate to explain there had been little formal investigation in this area at that time (e.g. Thelin and Fussner, 2005). It is a topic that had, however, been addressed within articles by a number of different authors (e.g. Lewis and Lowther, 2001; Brown 2003b; Hartshorne et al. 2005b; Thelin and Swanson, 2006) resulting in the circulation of a range of opinion but with little research-based evidence to substantiate these views. King Miller et al. (2011) acknowledged the difficulty in assessing the communication abilities of a child with CHARGE because of the significant communication delay, but also that the child might use idiosyncratic forms of communication that may be more difficult to recognise and interpret. This is likely to be one reason why there is a lack of published formal research in this area that has resulted in a reliance on anecdotal reporting and more personal observations. Swanson (2011) supports consideration of the reporting of such observations, explaining this is currently where the best information on this subject is likely to be found. It may be this that led Peltokorpi and Huttenen (2008) to undertake a multiple case study (of 3 children with CHARGE) as an initial attempt to begin gathering more evidence with regard to communication. Although it is not possible to generalise this to the wider population with CHARGE it identified some characteristics in the communication of the 3 children studied (e.g. the most frequent use of gesture as a communication mode) that may provide the foundation for future research.

Thelin (2002) stated that many children with CHARGE pass through critical periods in the development of speech and language in a state of sensory deprivation, not only due to visual and hearing impairments but also impairments of other sensory

modalities. There are few studies that focus specifically on the development of communication but research by Thelin and Fussner (2005) provided evidence for the view that poor communication ability is common in CHARGE. Their survey of parents of 28 children with CHARGE identified three factors relating to the acquisition of symbolic language, these being successful use of amplification (to provide greater access to speech where there is a significant hearing impairment); the ability to walk independently (where it was suggested this might enable the child to position her/himself within her/his 'communication bubble' to receive optimal visual and auditory communication); and the initiation of communication training by three years of age. The term 'communication bubble', previously used by Davenport (2002a), is a useful term for the educationalist to be aware of to ensure the provision of an optimal communication environment and represents the area around the child within which a person must be to enable the child to communicate comfortably. The degree of proximity needed will be dependent on the child's sensory impairments and ability to use their residual senses to gain visual, auditory and tactual information.

Whilst Thelin and Fussner (2005) relied on a questionnaire, Peltokorpi and Huttenen (2008) undertook a multiple case study to examine the communicative abilities of 3 children with CHARGE during interactions with their mothers. The children involved in this investigation all had a confirmed medical diagnosis of CHARGE and were at the pre-verbal language stage, but ranged in age with the youngest 1 year 4 months, the next 3 years 9 months, and the oldest being 8 years 4 months. All 3 children were found to be at the pre-symbolic level and most frequently used gesture and vocalisation rather than sign to express themselves. All 3 were found to have initiated

communication almost half of the time when interacting with their mothers, although the rate of intentional communication was low in comparison to the total number of communicative acts observed. Peltokorpi and Huttunen (2008) concluded that although the clinical profiles (including, for example, degree of visual impairment) for these children were very different, they all showed similarities in the form and purpose of their communication. This conclusion may not be so surprising when recalling that one of the criteria for participating in this study was quite specific in that the children needed to be at the pre-verbal language stage. Little acknowledgement was made of the variation in ages that would suggest the older child is progressing at a slower rate than the 2 younger children in the study, and perhaps reflects the heterogeneous nature of this condition. One major weakness of the study is a lack of attention to the impact of the children's hearing impairments. Profiles of each of the children involved in the study showed they all had a hearing impairment; two of these had unaided hearing levels reported to be 40dB and had been consistent hearing aid users for 3 years 3 months and 6 years each. The third child was reported to have a greater degree of hearing loss (40-80dB unaided level), was not using amplification and:

Cannot hear speech spoken with normal loudness. (p.30)

Despite the fact the impact of visual impairment on the child's interactions is discussed at length there is no recognition or discussion of these children's hearing impairments and how they might affect the child's interactions or the results of the investigation. They do, however, recognise it is important to provide positive auditory experiences and appropriate amplification.

The need for caution in making any attempt to generalise the findings of a study of only 3 children with CHARGE becomes clearer when considering work undertaken at the University of Tennessee involving a series of studies focused on the expressive communication skills of children with CHARGE, the entire study being presented by King (2009). The study was similar to that undertaken by Peltokorpi and Huttunen (2008), but involved a larger number of children (21) in a multiple case study, including children who had a wider range of communicative abilities, and only focusing on intentional communication. The children were categorised according to their communicative abilities but, as with Peltokorpi and Huttunen (2008) it is not clear whether they considered the degree and impact of any hearing impairment. Their findings illustrate and emphasise the diverse range of communication skills and modes used by children with CHARGE. They did, however, find that communication development in children with CHARGE is nearly always considerably delayed, and the rate of progress will be slower than typical development. This perhaps is the more significant finding of these studies for the educationalist supporting a learner with CHARGE, although the research reported by King (2009) also resulted in the development of an assessment tool for analysing the communicative abilities of these children. This may help to address some of the challenges in assessing the child with CHARGE as identified by King Miller et al. (2011).

Although Peltokorpi and Huttunen (2008) focus on the communicative abilities of the three children studied, this investigation could possibly also have explored further the social development of the children within these dyads, an aspect of a child's development described by Hartshorne and Salem-Hartshorne (2011) as the child's

ability to develop close and secure relationships and to be able to interact productively with others. Reda and Hartshorne (2008) explored the issue of early attachment and bonding of young children with CHARGE with their parents, and also the impact this had on parental stress. This is another study that employed a parental questionnaire, this time from parents of 25 children with CHARGE. Their findings showed almost half of the children in their cohort were reported to have experienced difficulty in becoming securely attached to their parents. It is interesting to note they found the longer it took for the child to show signs of attachment, the longer it also took the parents to feel they had bonded with their child. There are a number of anomalies associated with CHARGE that could potentially affect the early bonding process (e.g. visual and hearing impairments, facial palsy preventing smiling), together with the early medical difficulties and consequent frequent hospitalisations that may involve surgical intervention and prevent the parents being 'hands on' with their child. Although Reda and Hartshorne (2008) stated that in many cases these children do eventually become securely attached and the parents bond positively, they also found that nearly half the parents had total scores in the 'significant stress' range. As a result of their research they suggest:

Having a child with CHARGE syndrome is likely to be a challenging experience for parents. Early interventionalists should be alert to parenting stress and problems with attachment and bonding as they provide support for these families. (p.9)

Souriau et al. (2005) also explored the area of social development, although focusing more on social interactions. They collated the results of a parental survey for 71 individuals (with an average age of 8.71 years) and found that in social interactive exchanges only 4% were able to wait their turn to speak, whilst 38% had difficulty understanding and using social rules. Despite this, in a study by Hartshorne et al.

(2005a) it was found the children with CHARGE studied were less likely to try and avoid eye contact or touch, or to 'look through someone' than the norms for deafblind or autistic groups they were compared with through the use of a standardised checklist. This demonstrates that although children with CHARGE may be socially inept, they are nonetheless often very socially interested. This implies it is important for the educationalist to actively seek to identify strategies that will support the child with CHARGE to socially interact with their peers in a positive and successful way, particularly in light of the comment by Hartshorne and Hartshorne (2011) which states:

It is apparent that many children with CHARGE struggle to behave in a manner that promotes social relationships. (p.208)

In examining social relationships, Souriau et al. (2005) found that 63% preferred one-to-one relationships and 42% preferred adult rather than child company. This is supported by Hartshorne and Cypher (2004) who previously reported difficulty establishing same-age relationships was a primary concern for parents. Souriau et al. (2005) further found that 42% of their sample needed reference points whether they are temporal, visual or tactile, feeling this over-rode the respect for social rules. It was also noted that many individuals with good levels of language experienced some difficulty with language recall in the abstract forms of spoken and sign language. Although not established at the time of this investigation, it is possible this is linked to the executive function difficulties examined by Nicholas (2005) and Hartshorne et al. (2007) discussed previously in 2.2.9.

2.3.3 Cognitive development and attainment

The 'R' in the original CHARGE acronym represented retardation of growth and development, and early literature (e.g. Goldson et al. 1986) supported the view that

'mental retardation' was generally felt to be characteristic of this condition. Subsequent observations (Blake et al., 1990; Davenport, 1993), however, suggested about 50% of the population demonstrated good intellectual outcomes whilst approximately 25% had very poor intellectual outcomes. More recent research to ascertain the academic achievements of individuals with CHARGE reflects the medical heterogeneous nature of the condition and suggests these outcomes in the population of individuals with CHARGE are also very varied. Raqbi et al. (2003), Salem-Hartshorne and Jacob (2004 and 2005), Hartshorne et al. (2005b) and Nicholas (2005) all reach a similar conclusion that is encapsulated in a statement from Nicholas (2005):

Heterogeneity appears to be as great in the cognitive domain as it is in the medical symptoms of the disorder. (p.302)

The difficulty in generalising about the cognitive ability in children with CHARGE may be demonstrated when comparing the results of studies by Raqbi et al. (2003) and Dammeyer (2012). Raqbi et al. (2003) reported that approximately 50% of their cohort were without 'intellectual disability' whilst Dammeyer (2012) reported approximately 71%. However, Dammeyer gathered his data from medical records, whilst Raqbi et al. evaluated the intellectual level of their cohort by combining medical information, school performance, the type of school attended and their educational support requirements. Thus it can be seen these two research projects used different criteria and measurement tools and so it is not possible to make any true comparison or generalisation. Raqbi et al. (2003) attempted to ascertain whether there were any indicators that could predict the intellectual outcome for a person with CHARGE; in conclusion they felt that extensive colobomas resulting in low vision, microcephaly and brain malformation were the only three parameters predictive of poor intellectual

outcome. As mentioned, in this study the child's level of intellectual functioning was evaluated according to school performance, the type of educational provision the child was in and their "rehabilitation requirements" (p.484); this makes an assumption that the educational placement and the amount of support needed from therapists etc. is indicative of the child's cognitive abilities. The findings of this study might be explained in part if the presence of microcephaly and brain malformations are such that they impair the child's ability to learn. Additionally, the presence of more extensive colobomas may adversely affect the child's ability to use any residual vision effectively to help overcome any vestibular difficulties which may have led to Raqbi et al. (2003) asserting that a child with both an impaired vestibular system and extensive colobomas will have a poorer intellectual outcome than the child with a similar impaired vestibular system but better vision.

One additional factor that has not been discussed specifically in literature on CHARGE is the possible relationship between executive dysfunction (see 2.3.3) and learning disability. The Department of Health (2001) defines learning disability as impaired intelligence and social functioning that will have a permanent impact on development. The presence of executive function difficulties will most likely affect how far a child is able to perform to their cognitive potential since they contribute to all cognitive abilities and how they are utilised. What this illustrates is the difficulty in accurately assessing the intellectual abilities of many children with CHARGE (this will be discussed in 2.4), and the range of influencing factors that may need consideration. Overall, researchers of the studies outlined above do reach a point of consensus, although the evidence upon which this assertion is based is not clear, this being that:

Intellectual ability in individuals with CHARGE syndrome is underestimated. (Salem-Hartshorne, 2011, p.201)

Since major developmental milestones are delayed significantly in children with CHARGE there is a danger that cognitive impairment may also be presumed; this could be a false assumption according to Davenport & Hefner (2011), and echoes the earlier note of caution raised by Blake et al. (1990) who acknowledged cognitive impairment may be present:

But should only be diagnosed when the extent of the sensory deficit is known and when the child has been in an adequate educational programme. (p.223)

This is an important consideration for educationalists, especially those involved in early intervention, and implies children with CHARGE need to be given additional time to reach developmental milestones. There also needs to be an avoidance of making assumptions about a child's intellectual ability based on a comparison of when early developmental milestones are 'typically' achieved, using measures not appropriate for this population of children. Blake et al. (1998) reported many children with CHARGE show significant early developmental delay and explain there is often a discrepancy between results of early intellectual assessments (which they describe as focusing on gross and fine motor skill development and speech) and later cognitive assessments undertaken once they are more mobile and develop effective communication. Although this is the opinion of the authors expressed in this review paper, it does reflect the findings of Thelin & Fussner (2005) and Wachtel et al. (2007) who all suggest a relationship between the development of independent walking, communication skills and symbolic language that have a significant impact on learning (see 2.3.1).

An important factor in the development and learning of a child must be the way the child perceives their world and everyday experiences which affect the development of concepts. There appears to have been no research in this area although Davenport (2002b) feels the complex physical nature of the condition will have a 'profound effect' on how the child perceives the world and develops. Davenport & Hefner (2011) further state that many individuals with CHARGE can function within the normal intellectual range provided appropriate accommodations are made. Assuming that by 'accommodations' they are referring to altering the environment, curriculum format, or equipment to enable the child to gain greater access to the learning situation, these have not been readily researched, although Bernstein and Denno (2005) explored some accommodations in response to repetitive behaviours and Lieberman et al. (2012) examined accommodations to enable greater access to a physical education programme (to be discussed in 2.6 below). There is currently a reliance on anecdotal reporting and the sharing of opinion, and a dependency on the educationalist making an informed guess as to how best accommodate and respond effectively to the issues that have been raised.

An additional interesting point noted is that individuals with CHARGE are reported to often show a mixture of high potential features and conflicting low potential features in different areas of learning, perhaps being adept at some skills, but also engaging in basic stimulatory behaviour such as rocking or spinning (Hartshorne et al., 2005a; Lauger 2007). Possible causes for this are not identified but Hartshorne et al. (2005a) go further to describe it as:

The presence of "special abilities" in one area of development. (p.260)

This can have possible implications for how a child is perceived by those supporting him or her and may result in the child's learning abilities being greatly underestimated. This mixture of different levels of functioning could also result in the child developing 'splinter skills', otherwise called 'islands of good functioning' (e.g. Fein et al., 1981). Although this is an area that has been explored in relation to children on the autistic spectrum disorder it may be appropriate to also undertake investigations specifically in relation to children with CHARGE to help identify whether there are particular areas of learning where children with CHARGE experience greater strengths or difficulty and also what additional support is needed. This is important since the variation in functioning within different areas of learning means there is a potential danger of the child with CHARGE not developing a broad and secure foundation for later skills.

2.3.4 Behaviour

Within literature there is evidence that some genetic syndromes have a distinct but consistent pattern of behaviour that would be regarded as a behavioural phenotype (e.g. Cassidy and Morris, 2002). Hartshorne, a psychologist, has undertaken research and observations over several years to investigate the behaviour in individuals with CHARGE (often in collaboration with other researchers), and in 2008 Hartshorne et al. stated that moderate to severe behaviour difficulties frequently develop in this population. Within literature there appears to be a general consensus that these differ from behaviours seen in other syndromes and the rest of the deafblind population (eg. Bernstein and Denno, 2005; Hartshorne et al., 2005b). A review article by Blake and Prasad (2006) suggested the emergence of a behavioural phenotype, subsequently

developed further by Hartshorne (2011) building on his earlier research (Hartshorne and Cypher, 2004; Hartshorne et al., 2005a; Hartshorne et al., 2007). It is worth studying the elements of this CHARGE behavioural phenotype outlined by Hartshorne (2011), set out in Table 4, since this provides valuable insight for the educationalist and can support and influence the response made.

**TABLE 4: A CHARGE BEHAVIOURAL PHENOTYPE
(HARTSHORNE, 2011, p. 319)**

- Low normal cognitive functioning
- Very goal directed and persistent with a sense of humour
- Socially interested but immature
- Repetitive behaviours that increase under stress
- High levels of sensation seeking
- Under conditions of stress and sensory overload, find it difficult to self-regulate and easily lose behavioural control
- Difficulty with shifting attention and transitioning to new activities; easily lost in own thoughts.

This becomes more important when recognising behaviour in a child with CHARGE can be disruptive in the home and school settings, and may be so challenging that both the child and the people around them are put at risk (Hartshorne, 2011). The theory that children with CHARGE demonstrate distinct behavioural features is also supported by Bernstein and Denno (2005). They are some of the only researchers considering CHARGE behaviours to undertake their research in an educational setting where their focus was on 29 students assessed using a behavioural checklist. They also found the students with CHARGE displayed behaviours different to other deafblind learners in that they demonstrated many more repetitive behaviours than other students of a similar age, sensory impairments and ability levels. Hartshorne et

al. (2007) suggested potential causes for these behaviours that Hartshorne (2011) then later expanded upon, identifying four general sources of challenging behaviour, these being sensory and physical difficulties; negative life experiences; environmental factors; and behaviours relating directly to the genetic condition but independent of the sensory and physical difficulties experienced. This researcher's own experience suggests that teachers often cite management of what may be perceived as challenging behaviour as a major concern. If one is able to acknowledge the different elements of the behavioural phenotype and can identify why these behaviours may be occurring, the strategies developed in response are likely to be more informed and therefore more successful in reducing any negative behaviours occurring in the individual child. This knowledge can also potentially alter the teacher's perception as evidence is offered as to why the behaviour may occur, rather than the child simply being perceived as 'naughty' or 'disruptive'.

TABLE 5: A SUMMARY OF RECOMMENDATIONS WITH REGARD TO THE CHARGE BEHAVIOURAL PHENOTYPE (HARTSHORNE, 2011, pp. 324-325)

- To recognise that although a significant level of support may be needed, the child with CHARGE has the potential to succeed at school
- To accept that it may not always be easy to redirect a child's goals
- To recognise that social support from the child's friends/peers may be lacking
- That there is a need to identify potential sources of anxiety and stress and work to minimise these where possible with the likelihood that repetitive behaviours may then be reduced
- To accept the child is likely to need high levels of sensory input and stimulation
- To watch for signs of emotional and behavioural 'meltdown' and allow the child time to 'regroup'
- To provide schedules and routines that are regular and predictable

This is the approach adopted by Hartshorne (2011) and is one of the rare occasions where a researcher provides practical suggestions as an outcome of the research and theories generated, that can be implemented within the educational setting, as summarised in Table 5. A prudent step would be to test these suggestions and undertake further research to investigate the strategies employed in response to these insights that might be most effective when implemented with learners with CHARGE.

2.3.5 Self-regulation

One of the elements of the behavioural phenotype proposed by Hartshorne (2011) is a difficulty with self-regulation that enables a child to maintain a well-modulated physical and emotional state. He explains that individuals with CHARGE can fluctuate between being in an under-stimulated to an over-stimulated state, rapidly shifting between two extreme ends of this spectrum. This is a view shared by Brown (2011) who suggests this can prevent the child's readiness and ability to learn, and clearly implies there is a need for the educationalist to acknowledge this potential difficulty that can have an impact on the child's active engagement in the educational setting.

The literature examined suggests this is in part a consequence of sensory integration dysfunction (e.g. Brown 2003a, 2003b; Thelin & Fussner 2005; Lauger 2007, Hampson, 2013) as explained in 2.2.8. According to Hartshorne (2011), both parents and educators have observed that a child's difficulties with self-regulation are not entirely due to the sensory challenges the child encounters. With reference to earlier investigations by Nicholas (2005) and Hartshorne et al. (2007), Hartshorne (2011) suggests executive function difficulties are also an influencing factor. Brown (2011)

reflecting on his years of working as an educator with children with CHARGE states that in his opinion, executive function difficulties and self-regulation issues are:

The most challenging and least understood long-term aspect of this condition for the children themselves, their families and educators. (p.6)

There is a growing awareness and interest in this area, particularly with the self-regulation of emotion (e.g. Deuce, 2013b; Kennert et al. *in press*), and research-based investigations are likely to be of great benefit in developing knowledge and understanding in this area.

2.3.6 Fatigue

The issue of fatigue is raised frequently for children with CHARGE (e.g. Brown, 2005b and 2011a; Majors and Stelzer, 2008; Davenport and Hefner, 2011). Davenport and Hefner (2011) explain that deafblindness is “universally a source of fatigue” (p.11) but for the child with CHARGE there are also many other factors including the challenges created by other sensory impairments, but also chronic health conditions, multiple surgeries, and so forth that will make further demands on the child’s energy, resulting in potentially even higher levels of fatigue.

2.3.7 Summary

The heterogeneous nature of this condition has been identified by a number of different authors and it is likely that in every individual with CHARGE, the way in which the different anomalies come together (and the severity of each one), will be unique. Yet within literature authors also appear to suggest there are commonalities across the syndrome and the issues described above may be considered as affecting the development and functioning of each child with CHARGE to a greater or lesser

degree. Indeed, within the literature examined, the findings and discussion of research undertaken with a larger sample group often talk collectively of 'children with CHARGE', which perhaps implicitly supports the opinion that there are commonalities to be recognised and addressed. This is also expressed within the articles outlining opinion and providing anecdotal reporting presented by practitioners working in the field of education with learners who have CHARGE.

Considering the sheer number of anomalies involved with this condition it is perhaps not surprising that a child's overall development is likely to be affected. The potential impact of the issues on the development and functioning of a child with CHARGE outlined in section 2.3 become very evident from the information obtained in this exploration of literature. It is perhaps the cumulative effect that is most challenging for these children; for example, where vestibular difficulties, visual impairment and poor tone create issues with motor skills, which in turn are felt to affect the communication and development of symbolic language. It is incumbent upon educators to identify if and how a child is affected by each of these aspects (and others) with regard to overall learning and development, and to plan an effective response. The challenge of effective assessment of the development and functioning in a child with CHARGE that can inform this process has emerged from a number of the sources examined (e.g. Salem-Hartshorne and Jacob, 2004) and this will also benefit from closer exploration.

2.4 Assessment

Assessment is an important tool with the outcomes used to inform the development of an effective educational programme. This can include gathering information about the

child's sensory strengths and needs, identifying individual learning styles, obtaining a baseline of skills to support the educational plan, to enable a measure of progress, and so forth (Eyre, 2000). Developmental and cognitive assessment of individuals with CHARGE is referred to by a number of authors and there is a general consensus that ideal tools are not available for these assessments (e.g. Smith et al., 2005). As highlighted by Salem-Hartshorne and Jacob (2005) researchers reporting on development in children with CHARGE:

Often failed to report...how developmental progress was assessed. (p.262)

A number of issues have been raised relating to difficulties in assessing persons with CHARGE. Graham et al. (2005) state that early intellectual assessments tend to focus on gross motor and fine motor development as well as speech skills, all of which are likely to be more delayed in children with CHARGE than in typical development. As a consequence it is considered these developmental milestones are not likely to be accurate indicators of a child's underlying ability and cannot be relied upon to predict intellectual capacity. Hartshorne N (2002) and Salem-Hartshorne & Jacob (2004) reported that traditional IQ tests are also likely to under-estimate the true abilities of a child with CHARGE (as for the wider deafblind population, e.g. Hodges, 2004). There may be a number of issues that make it difficult to judge a child's responses and true capabilities, including confusing poor expressive language skills with mental ability, and lower scores reflecting difficulties with processing or output rather than cognitive impairments. As a result it is generally agreed that universal IQ tests need to be complemented by other tests more appropriate for these children (Raqbi et al., 2003; Hartshorne N 2002; Salem-Hartshorne & Jacob 2004). Other standardised assessment tests may be considered but this literature review has found the validity of

this approach does not yet appear to be established. Salem-Hartshorne (2011) considers that unless the assessor can be sure the child being tested has clearly understood what is involved within the test process, and also that the assessor can be confident of understanding the responses given, the results of these tests:

Should not be used for decision-making purposes. (p.201)

Where there are no standardised tests that can accurately measure a child's cognitive abilities and development there is a risk this may result in subjective and inaccurate judgements being made, and a cautious approach needs to be adopted. For example, in an earlier study by Blake and Brown (1993) it was stated a number of the children within the cohort had "normal intelligence" (p.400) although the authors found a number of children in the cohort had not received a "relevant assessment" (p.400) and it is unclear as to how this conclusion was reached. This research was based on the responses of a parental questionnaire and further evidence to validate this data is needed before the findings can be confidently accepted.

As a consequence of their multi-sensory impairments, the majority of children with CHARGE may be described as being 'input impaired' (Davenport & Hefner 2011) and it therefore follows that measuring output may not enable the educator to estimate true cognitive ability (Davenport 2002a). This all suggests that the educator and other involved professionals need to consider how accurate information can be obtained about how the child with CHARGE is developing and their true cognitive abilities, using processes that are reliable but do not depend on assessment tests not standardised for this group. An example of this can be found in research undertaken by Salem-Hartshorne & Jacob (2004 and 2005) who looked at developmental ability

using a measurement of adaptive behaviour. Sattler (2002) explains that measurement of adaptive behaviour focuses on how far the child is achieving age-appropriate levels of personal independence and also social skills. It is also stressed this measure can be completed by someone who is very familiar with the child and has observed the behaviours highlighted within this measure. Hartshorne et al. (2005b) suggest the level of adaptive behaviour often illustrates underlying intelligence, supporting the opinion that this would appear to be a more valid assessment tool. Nonetheless, it still needs to be recognised that the adaptive behaviour measure used by Salem-Hartshorne and Jacob (2005) for their investigation was one standardised on a population of typically developing children (as recognised by the researchers themselves) and some items included were likely to be inappropriate; for example, questions about eating skills, given that swallowing difficulties with aspiration being considered as one of the main diagnostic criteria for this condition. Use of standard test procedures alone may reflect a deficit model, focusing on what the child is not able to do rather than their individual achievements and strengths when compared to their hearing-sighted peers. According to Salem-Hartshorne (2011) it is important to employ a combination of formal and informal assessment, consultation, interview and observation to ascertain each individual child's strengths and needs.

2.5 CHARGE and deafblindness

As explained in 2.2.3 the majority of children with CHARGE will be considered part of the deafblind population, with this condition now regarded as the most common genetic cause of congenital deafblindness (Hartshorne et al., 2011). According to

Chandran & Shah (2011) hearing loss occurs in 85-100% of the CHARGE population and is the most commonly occurring anomaly in CHARGE. With eye anomalies also occurring in 80-90% of the CHARGE population (Hartshorne et al., 2011) it is evident that although not every child with CHARGE will have both visual and hearing impairments, the large majority (over 90% according to Hartshorne et al., 2011) will experience a significant impairment of the two distance senses of sight and hearing.

The question of whether they form a distinct sub-group within the deafblind population needs to be raised and, if this is accepted, whether it then follows that they require additional or alternative teaching approaches that have yet to be clarified and established through research. When studying CHARGE it quickly becomes evident there are issues in addition to the deafblindness that Brown (2003a) feels makes this population challenging and, he contends, different within the whole deafblind population. This view is supported in the findings of research by Hartshorne et al. (2005a) who found their cohort of 160 individuals with CHARGE were very different not only when compared to other children who were autistic, (in that they were more socially engaging and had better language and communication skills), but also other children who were deafblind. When compared to the wider deafblind norms established with the checklist employed, it was shown these children engaged in more sensory related behaviours and demonstrated a different behaviour profile. This led the researchers to conclude:

The behaviour of children with CHARGE cannot be attributed solely to deafblindness. (p.260)

A smaller research study by Bernstein and Denno (2005) found their cohort of 29 students with CHARGE engaged in more frequent repetitive behaviours than other

deafblind individuals with similar sensory impairments and ability in their educational setting, and concluded these behaviours were a characteristic of their diagnosis of CHARGE and not a function of their multi-sensory impairment or functional abilities.

Some of the issues, in addition to behaviour, that perhaps identify individuals with CHARGE as distinct from other deafblind persons become apparent when considering the number and type of anomalies related to this condition. These have been described previously, as have areas relating to development and learning that may also support this distinction. It has more recently been stated by Hartshorne et al. (2011) that CHARGE may be the only disorder which results in “deficits of all the senses” (p. xiii). This extreme multi-sensory impairment combined with other anomalies associated with CHARGE would indicate they may form a distinct group. This view is supported by Hartshorne et al. (2011) who report the impact of the sensory issues combined with the distinctive collection of medical and physiological disorders result in a “unique course of individual development” (p.xiv). This indicates it is therefore very possible this population may require a particular and distinct response and new thinking when planning intervention programmes.

2.6 Educating learners with CHARGE

In an extensive search of literature relating to CHARGE little evidence was found of research focusing directly on educational intervention and the provision of an optimal learning environment for these learners. Those that were available focused on a particular aspect, with Bernstein and Denno (2005) examining the repetitive behaviours and Lieberman et al. (2012) exploring physical education. Consequently

there is only limited information within these studies to inform the overall educational support provided to this particular population of learners.

The medical complexity of this condition has led to a focus on medical research that has proved invaluable in securing an appropriate medical response, and more children with CHARGE are now surviving than in the past (Hartshorne, 2006). This increasing knowledge base also provides important information about the affected individual's ability to function and learn. What is lacking is the research-based evidence on the education, teaching strategies and approaches that can effectively support children with CHARGE and enable them to achieve. Medical professionals Smith et al. (2010), expressed their concern over the lack of literature regarding school-based strategies for children with CHARGE, commenting this is problematic for both parents and teachers who are in need of support to facilitate the learning and development of these children. Every child and young person with CHARGE will face daily challenges and barriers to learning as a result of the impact of the different anomalies present. However, there are also external barriers and one of those is:

An educational system that is ill-prepared to meet their diverse needs.
(Hartshorne et al., 2011, p.xi)

It is likely that this is, at least in part, due to a lack of research-based evidence to inform educators when planning and implementing educational programmes for learners with CHARGE. Whilst it was not possible to identify evidence-based research papers focusing on pedagogy and learners with CHARGE, one research-based paper was published by Lieberman et al. in 2012 on the physical education of children with CHARGE. A number of articles and book chapters were also located that provide anecdotal information and report on the experience of practitioners directly involved in

supporting learners with CHARGE. It is recognised these sources provide a personal view of the subject being discussed not grounded in research, and were also not subjected to moderation by peers, but nonetheless it was felt these sources should be considered since the views of practitioners experienced in teaching learners with CHARGE potentially provide valuable insights; this in addition to little other material being available.

An early article by Jones and Dunne (1988) identified a few implications for teachers of pupils with CHARGE, although not research based and presumably arising from their own experiences. It was written prior to much of the more recently acquired knowledge of CHARGE, yet their suggestions might be considered as relevant in light of aspects of this condition more recently identified; for example, they suggest teaching functional skills in real situations which may be an effective response to a child's executive function difficulties (Cooper-Kahn and Dietzel, 2008). Lewis and Lowther (2001) provided a review of the social behaviour of children with CHARGE and factors to consider when planning educational intervention, with a focus on the impact of 'life events' that may put the child at risk from social and emotional problems which could manifest in behavioural issues. This paper draws on current knowledge at the time of writing and focuses on the initial diagnostic criteria (Pagon et al., 1981) that were subsequently refined as knowledge continued to increase. This illustrates the limitation of this paper in light of the subsequent increase in knowledge of CHARGE, and Lewis and Lowther (2001) would not have been able to consider the impact of other anomalies more recently identified, such as neurological impairments and executive function difficulties that may also adversely affect a child's social behaviour.

Nonetheless, more recent materials provide some evidence to support their suggestions of what might be needed where, for example, an appropriate response to the social interaction difficulties identified by Souriau et al. (2005) as set out in 2.3.2, might well be to encourage and support the child to develop their relationships with adults and their own peers and also encourage positive behaviour (e.g. in turn-taking in play) as advocated by Lewis and Lowther. In 2004, Griffin et al. very briefly considered the educational needs of learners with CHARGE in their article although the focus of their writing was the use of assistive technology. They were of the opinion there was a need to encourage these children to explore and interact with the environment and ultimately develop greater independence, advocating the use of technology to support this.

Majors and Stelzer are both educationalists who have worked for some time with a number of learners with CHARGE in a particular educational setting. In 2008 they provided a contribution to the CHARGE Syndrome Foundation's 'Professionals Packet' that addressed the educational needs of children with CHARGE, presumably drawing on their direct experience, and many of these points were refined and clarified in a later chapter by Majors (2011a). There is value in such reporting by experienced teachers, but it is important to recognise their experiences are of a cohort of students in a particular educational setting and it might be argued that entry criteria for placement within this provision could exclude some learners with CHARGE (for example, if they are not deafblind, or if they have profound and multiple learning difficulties). Consequently it is important to ask whether their reporting is relevant for

the full range of learners with CHARGE or only pertinent for the students within this particular educational setting.

Having expressed their concern over the lack of literature for educators, Smith et al. (2010) produced a paper ('An Educator's Primer') to provide information to support understanding and the development of education programmes for learners with CHARGE within the context of inclusive education. They provide a summary of some of the medical characteristics and a basic summary of development and behaviour. One section of this paper is largely a review of the writing of Jones and Dunne (1998), Lewis and Lowther (2001) and Griffen et al. (2004) and summarises some of the practical suggestions offered by these authors. One of the criticisms of literature on educating children with CHARGE voiced by Smith et al. (2010) is that:

Suggestions tend to be brief, general and not to address the underlying principles of intervention. (p.298)

Yet this is a criticism that could also potentially be made of this paper although, as a 'primer', the intention may well have been simply to provide basic information. Additionally, as medical professionals they are likely to lack the depth of knowledge and experience with regard to education and learning possessed by the teachers they intended the paper to reach. Smith et al. (2010) recognise the literature currently available on educating children with CHARGE is opinion rather than evidence-based, and that it frequently relates to interventions most often recommended for learners with a visual impairment. As a result of a lack of literature specifically on the education of learners with CHARGE they suggest turning to examine literature on children with multiple disabilities. Although this may be a helpful approach to take it is disappointing that they do not identify the need for research to provide a secure evidence base to

inform the development and implementation of intervention strategies specifically for learners with CHARGE.

The first book published on CHARGE, edited by Hartshorne et al. (2011) has been welcomed by practitioners working in the field. It is emphasised this book was written primarily for health professionals, although families and educators are also identified as part of the potential audience. Educational matters are mentioned in some chapters but there is no chapter specifically focused on educating children with CHARGE. Horsh and Scheele (2011) also edited a book on CHARGE, published initially in German and then subsequently translated into English. Within this book is a chapter by Majors (2011a) that addresses educational considerations for learners with CHARGE. As mentioned previously, Majors is an experienced practitioner in an educational provision that has been involved over the last 20 years in developing appropriate educational programmes for learners with CHARGE. Whilst this writing is not research-based it remains a valuable source to examine, drawing on the knowledge and reporting by an experienced practitioner, although recognising that this is limited to one educational setting. Similarly Brown (2011a) wrote an article outlining some of his thoughts on educating these children, specifically in relation to self-regulation, that also needs consideration as it is likely to provide valuable insights into the practical issues involved although recognising this is also only one person's perceptions, albeit from an educational practitioner experienced in working with learners with CHARGE.

A more recent development is the sharing of information through the medium of a webcast. Again whilst this is not research-based, the webcasts are presented by educational practitioners working with learners with CHARGE. Majors (2011b) shares her knowledge of the impact of CHARGE on communication and learning, whilst Stelzer (2011) identifies some teaching strategies for use with this group of children. As previously identified, there is a need to view these with a note of caution since both practitioners are working in the same educational setting and their opinions are likely to be influenced by the ethos of the school, and the group of learners with CHARGE who attend that particular provision.

2.6.1 Meeting the educational needs of learners with CHARGE

When one considers the complexity and range of anomalies associated with this condition it is not surprising a child with CHARGE will face many challenges that might interfere with their educational success (Smith et al., 2010). It is not enough, however, to simply recognise the features and implications of the possible anomalies that are a part of CHARGE. Educators also need to recognise the demands subsequently being placed upon the child (Brown, 2011a) and how this is likely to impact on their learning and overall development.

It can be argued that as a consequence of the heterogeneous nature of CHARGE it is likely an educational programme will need to be highly individualised for each child since the combination of the anomalies seen will be specific in each affected child, creating a unique set of needs. Despite the heterogeneous nature of CHARGE, as discussed in 2.3.7, it is suggested there are commonalities that should be considered

when developing an appropriate educational response to these needs. It is therefore possible there will be common elements in an educational programme and successful teaching strategies that may be applied to a range of individuals with CHARGE. This view is supported by Majors (2011a) who explains learners do present with similar personalities and learning characteristics, including facing multiple challenges, experiencing high levels of anxiety (and a consequent perseveration), a need to be in charge requiring the class teacher to develop the art of negotiation, an overwhelming need for routine and structure, and experiencing difficulties with transitions. This indicates it may be possible to generalise about the educational requirements of learners with CHARGE whilst maintaining respect for the individuality of each child.

There is recognition amongst educators that meeting the educational needs of a child with CHARGE will be challenging (e.g. Griffin et al., 2004), but as Majors (2011b) explained:

If we can figure out the right strategies, these children can learn amazing content.

The difficulty lies in establishing what strategies are appropriate. Smith et al. (2010) state teaching methodology should be evidence-based, but this is difficult when educators are reliant on direct reporting and anecdotal evidence not supported by research. One exception identified is the paper by Lieberman et al. (2012); although referring to the teaching of physical education there are some strategies the educationalist might be able to generalise to the broader educational setting (these will be discussed shortly). Looking at the anecdotal reporting and sharing of opinion, according to Brown (2011a) the first step must be to identify the issues present and then to formulate an effective response with one initial step being to consider the

medical needs of the child and the impact of their early medical history (also raised by Stelzer, 2011; Majors, 2011b).

The development and delivery of an appropriate curriculum with the necessary adaptations is fundamental to the teaching of any child. Smith et al. (2010) stress the need for a developmental curriculum although they, and Brown (2011a), recognise this can be more difficult to deliver in the later stages of education. Majors (2011b) suggests that as far as possible a 'typical' school curriculum should be offered to the child with CHARGE. Brown (2011a) explains it is also important to think about the way the curriculum is being delivered, with Majors (2011a) advocating a cross-curricular setting. These requirements are realised in "A curriculum for multi-sensory impaired children" (Murdoch et al., 2009) and implies this is a curriculum that might be appropriate for the child with CHARGE, as for other deafblind learners. It is, however, a curriculum designed for learners with special educational needs working towards Level 1 of the National Curriculum who are attaining at P levels P1-P8 (Qualifications and Curriculum Authority, 2009). Given a full range of ability noted in the population of children with CHARGE is acknowledged in literature (as discussed in 2.3.3), this curriculum is unlikely to be appropriate for those learners working at a higher academic level. Furthermore, there has been a small amount of research to support the view that the CHARGE population is distinct from the larger deafblind population (see 2.5). If this is so, it is possible there are elements that may not be adequately addressed in a curriculum designed for deafblind learners.

Smith et al. (2010) encourage educators to devise strategies based on an understanding of the strengths and needs of each individual child. This would also be regarded as good practice when educating other deafblind learners (e.g. Eyre, 2000), stressing the importance of an individualised approach. When identifying the strengths and needs of the learner with CHARGE it is important to recognise that for much of the school day they are likely to be operating at the very thresholds of their sensory abilities and will not always be fully available for learning due to the complexity and “multi-layered set of issues” they experience (Brown, 2011a, p.6); this is therefore likely to be an important underlying factor influencing what the child’s needs might be. When educating all children there is a desire to support the achievement of developmental milestones; to increase each child’s knowledge and understanding; and to prepare them for adulthood (socially, vocationally and academically). Within the literature available some specific aspects are addressed as requiring particular focus by educators. At a fundamental, underpinning level is the need to encourage the child to explore and interact with their environment to provide the foundation for more advanced skills and increasing independence (Griffin et al., 2004). This approach is, however, not unique to learners with CHARGE and is recognised as important in underpinning the learning of other deafblind children. For example, Murdoch et al. (2009) describe their MSI curriculum as being:

Rooted in a transactional model of development emphasising interaction between the individual and the environment. (p.12)

The development of communication skills is identified by Griffin et al. (2004), Smith et al. (2010) and Majors (2011a), that links with the development of social interactions and social behaviours with both peers and adults (Lewis & Lowther, 2001). Additionally the need to support the development of self-organisational skills is raised

both by Stelzer (2011) and Majors (2011a); the importance of concept development is also highlighted by Majors (2011a) and Lewis & Lowther (2001), identifying what associations a child is able to make and what their understanding is.

Throughout the literature available the importance of ensuring that cognitive ability is not under-estimated is stated (e.g. Lewis & Lowther, 2001), with Brown (2011a) expressing the view that cognitive difficulties are less of an issue for the child with CHARGE than the impact of their sensory and neurological difficulties. So the question remains, what educational strategies should be adopted to enable each child with CHARGE to work effectively towards reaching their full cognitive potential? Strategies suggested in relation to these aspects as identified within the sources explored are set out below, and a comparison made with strategies that form a part of the education model for deafblind learners described within literature.

2.6.2 Educational placement

A study by Blake & Brown (1993) included the types of educational provision of the 30 children with CHARGE in their study who were in educational placements in the UK. A later study by Deuce et al. (in press) also gathered data on the educational placement of 44 children with CHARGE (also in the UK). Both studies gathered data using a questionnaire sent out to parents, with a comparison of the findings relating to educational placement set out in Table 6. This data may suggest a move towards greater inclusion of children with special educational needs in the mainstream setting, but there could be a range of other influencing factors that need consideration (such

as the closure of some specialist schools; the move to keep children within their local authority area etc.).

TABLE 6: TYPES OF EDUCATIONAL PROVISION OF CHILDREN WITH CHARGE REPORTED IN TWO STUDIES

Type of school	Percentage of children from study in the provision	
	Blake and Brown (1993)	Deuce et al. (in press)
Hearing impaired/ visually impaired	40%	27.5%
Physical impairment	3%	7%
Severe learning difficulties	33%	27.5%
Deaf blind provision	3%	-
Mainstream	14%	31%
Independent/ non-maintained	-	7%

What the findings of both studies do show is a range of different educational provisions where these children were placed. This is a further indication of the heterogeneous nature of CHARGE whilst also being a reflection of what is available to a child in the area where they live.

2.6.3 Teaching strategies for learners with CHARGE

Since most children with CHARGE will be considered part of the deafblind population it seems appropriate to utilise the educational philosophy for deafblind children and the identified strategies that support development and learning of this whole population. According to Brown (2011a) this offers the “best match”, but this phrase also suggests something else is required and the CHARGE population is separate and distinct in some way. This is indicated by Brown (2012) who proposes that in addition to deafblind education, there is a need to recognise ‘CHARGE-specific considerations’

(arising from impairment of multiple sensory modalities, medical issues and other anomalies). Majors (2011a), perhaps sums this up most succinctly, recognising that learners with CHARGE will have significant challenges as a result of their dual-sensory impairment, but also that:

The learner with CHARGE syndrome, additionally must cope with multiple challenges along with limited access to both vision and hearing. (p.209)

This implies there may be a need for a combination of teaching strategies identified for deafblind children as a whole, and also some strategies more specific to learners with CHARGE. In the literature focusing on the education of children with CHARGE (outlined earlier) some strategies are presented for consideration by educators involved with these learners, although sparse in number, these being discussed below.

2.6.3a Focusing on the child

It is vital educators spend time observing the child with CHARGE (Stelzer, 2011) and Lewis and Lowther (2001) raise some useful observational questions including asking what strategies does the child employ when exploring, and for what period of time is the child able to engage? Majors (2011a) and Jones & Dunne (1988) also identify the need to observe and identify the child's strengths and to then focus on these in the development of an individualised programme tailored to the child's strengths and needs. Majors (2011a) also stresses the importance of discovering what is motivational for the child and the need to follow the child's leads and preferences. All of these are well-established strategies used to support the learning of deafblind children; focusing on the child and building on identified strengths is intrinsic in the many of the chapters in Aitken et al. (2000), a resource for those teaching deafblind

children. This approach has underpinned much of the work developed in relation to communication and congenitally deafblind individuals (e.g. Janssen et al., 2003; Rødbroe and Janssen, 2006). It is also expressed by Murdoch et al. (2009) in their curriculum for deafblind learners, and by Nelson et al. (2010), in their assessment tool for use with this population. Similarly, following the child's lead, responding to their preferences, and identifying what is motivational to the child are highlighted in a range of resources for use with deafblind learners (e.g. Sense, 2002; Murdoch et al., 2009; Nelson et al., 2010; Aitken, 2000a; Pease, 2000).

Children with CHARGE can be observed to adopt a 'burst-pause' sequence to their learning; within the literature on CHARGE this term is used to describe a behaviour or learning style observed within the child, where the child demonstrates 'bursts' of activity, interspersed with periods of little obvious engagement ('pauses'), before re-engaging with another 'burst' of activity. Jones and Dunne (1998), Brown (2011a) and Majors (2011a) highlight the need to work at the child's pace and build in pauses during activities to match the child's own burst-pause sequence. Hodges (2000) writing about supporting deafblind learners, talks of the 'burst-pause rhythm' with regard to the activity the child is engaged in, rather than the 'burst-pause' element being observed directly in the child's behaviour, and Murdoch et al. (2009) describe the activity of deafblind learners at an early developmental stage as having "bursts and pauses" (p.28), illustrating that this learning style is not unique to children with CHARGE and may be observed in the wider deafblind population.

2.6.3b Sensory needs

As Smith et al. (2010) identified, the visual and auditory needs of children with CHARGE are often addressed in literature (e.g. Blake & Brown, 1993; Salem-Hartshorne & Jacob, 2004; Majors 2011a) including the need to attend to the use of equipment, together with the adaptation and differentiation of the environment, activities and materials. The importance of addressing the visual and auditory needs of deafblind learners as a whole is recognised within literature (e.g. Hodges, 2000; Pease, 2000) and is reflected in the “Quality Standards in Education Support Services for Children and Young People who are Deafblind/Multi-Sensory Impaired” (Sense 2002) where the need for input from a qualified teacher for the visually-impaired and a qualified teacher for the hearing-impaired are both highlighted in addition to support from a qualified teacher for deafblindness/MSI.

Brown (2011a) and Smith et al. (2010) amongst others, identify the need to consider vestibular issues and provide support for sensory integration difficulties. Brown (2011b) considers that vestibular dysfunction has “perhaps the most far-reaching...implications” (p.51) for children with CHARGE and may be why a range of investigations and articles have been written about the impact of vestibular dysfunction in CHARGE (see 2.2.4). Despite this being a factor that has significant impact on a child’s daily functioning and learning there is very little written offering practical strategies that may be helpful in responding to these difficulties. Brown (2007b) is one practitioner who has explored strategies that may be helpful; although this article is written with reference to all deafblind learners he does explain that damage to the vestibular system occurs “especially in children with CHARGE

syndrome” (p.18). Apart from this one paper, reference to vestibular difficulties is not raised or supportive strategies identified in the wider literature relating to the deafblind population. This suggests this is an area not widely considered but that it may also be regarded as a more significant issue for children with CHARGE than other deafblind learners.

Sensory integration difficulties are also rarely mentioned in literature relating to educating learners who are deafblind; it is Brown (2009) again who highlights the issue of sensory integration difficulties in the deafblind population, and although Murdoch et al. (2009) state the importance of recognising a child’s sensory integration difficulties, they do not expand upon this or suggest what response should be made. Other references to sensory integration difficulties with regard to the wider deafblind population were not found, and no evidence of potential strategies except in the article by Brown (2009). Brown (2005b) and Hampson (2013) discuss sensory integration difficulties in relation to children with CHARGE and suggest some strategies that Brown reiterates in his article of 2009, focusing on the deafblind population as a whole. Hampson (2013), an occupational therapist, also puts forward similar strategies as being potentially beneficial for the child with CHARGE who is experiencing sensory integration difficulties, like Brown (2009), this centres on the development and implementation of a ‘sensory diet’. Hampson (2013) makes a statement that perhaps explains in part why educationalists have not put forward suggestions for strategies to implement in response to a child’s sensory integration difficulties, this being that before activities to address the child’s difficulties can be put together (and form the sensory diet) there is a requirement for:

A sensory integration assessment by a suitably qualified and experienced paediatric Occupational Therapist (p.2)

This implies the need for close collaborative working (as advocated by Thomas and Deuce, 2013), and a helpful strategy should include securing an assessment from such a suitably qualified and experienced occupational therapist to inform the development of a response. Brown (2011a), writing of strategies for the learner with CHARGE, additionally suggests regular opportunities to move around and engage in large motor movements should be provided in response to the child's sensory integration and vestibular difficulties throughout the school day. Although this is not identified with regard to other deafblind learners in response to sensory integration difficulties, wider literature identifies this strategy as a requirement to support gross motor development (e.g. Aitken, 2000a; Murdoch et al., 2009). This raises an interesting point that the same or similar strategies may be employed with both learners with CHARGE and the wider deafblind population, but the reason for employing these strategies may differ between the two identified groups of learners.

In relation to the child's sensory difficulties, which together with other implications of CHARGE create daily challenges for learning and everyday living, are the issues of stress and anxiety, the emergence of Obsessive-Compulsive Disorder (OCD) behaviours and fatigue (Brown, 2003b; Graham, et al. 2005; Wachtel et al., 2007 and Smith et al., 2010). The presence of high levels of stress and anxiety is recognised in individuals with learning disability (e.g. Huntington and Bender, 1993; Svetaz et al., 2000) and is also addressed within the broad deafblind population (e.g. Pease, 2000; Department for Education and Skills, 2003) where Pease (2000) suggests some strategies to help minimise stress likely to arise from adult demands being placed

upon the child. These strategies match those recommended for teachers of learners with CHARGE, being to work at the child's pace, providing activities the child is felt to enjoy, reviewing the balance between familiar and new more challenging activities, and ensuring the child receives positive feedback (p.48). The use of clear structure and consistency of approach are also well-established strategies for both learners with CHARGE and other deafblind children (e.g. Brown 2011a; Murdoch et al., 2009), again providing an example to support the argument against children with CHARGE being distinct from other deafblind learners. In addition to reading and responding to stress and arousal levels, the issue of fatigue is reported and discussed more often in relation to children with CHARGE (see 2.3.6), which may indicate a distinction between learners with CHARGE and other deafblind learners. It is interesting to note that within the literature review, strategies to reduce the impact of fatigue were only located in sources directly relating to CHARGE; here Brown (2011a) suggests the need to respond by simplifying activities and ensuring the "teaching point is clear and accessible" (p.5) whilst also monitoring the learning environment to reduce other sensory demands, and Majors (2011a) suggests ensuring the use of 'sensory breaks' and access to a 'rest' space when required.

2.6.3c Communication and social interactions

The development of effective communication and social interactions, seen as an essential pre-requisite for effective learning, is likely to be a major challenge for any child with visual and hearing impairments. The development of an appropriate communication programme, including use of a Total Communication approach and other appropriate communication modes, is well-documented for learners with

CHARGE (e.g. Lewis and Lowther, 2001; Smith et al., 2010; Majors, 2011a&b; Brown, 2011), but is also often advocated for other deafblind learners (e.g. Pease, 2000; Murdoch et al., 2009) and children with hearing impairment and additional needs (e.g. National Deaf Children's Society, 2013). That a communication programme for deafblind children should include regular choice-making opportunities is expressed by many including Aitken, (2000a), Murdoch et al., (2009) and Nelson et al., (2010). This is also stated for learners with CHARGE (e.g. Smith et al., 2010, Majors 2011a) with the explanation that this in turn will support the development of negotiation skills and empower the child, supporting them to take greater ownership of their own learning (Majors 2011 a&b), this latter being an aspect of learning for deafblind learners addressed specifically by Murdoch et al. (2009). Although this again demonstrates the appropriateness of the same approach and strategies for both learners with CHARGE and other deafblind children, an element additionally addressed that may be more specific to CHARGE is the use of concrete components (Brown, 2011a) to address additional communication issues created by the presence of executive dysfunction (see 2.2.9).

The need to support the development of social interactions is also reflected in identified strategies for all deafblind learners (e.g. Murdoch et al., 2009). With regard to learners with CHARGE, Lewis and Lowther (2001) and Majors (2011 a&b) also express the need for strategies to support the development of positive social behaviour and also to overcome potential difficulties resulting from both executive function and sensory integration difficulties in addition to the child's sensory impairments, for example, learning to initiate and maintain interactive sequences.

2.6.3d Functional life skills

A number of different authors (e.g. Jones and Dunne, 1988; Graham et al., 2005; Nicholas, 2005; Smith et al., 2010 and Brown, 2011) stress the need for an educational programme that ensures the learner with CHARGE can engage in activities to support functional life skills through the use of meaningful tasks undertaken in real settings. This is also raised by Stelzer (2011) in her webcast. Research by Blake et al. (2005) involving 30 adolescents with CHARGE found their cohort were more independent in self-care skills than the more complex independent living skills, and they suggest this might in part be due to a lack of exposure to learning situations focused on developing these skills. If this is so, the importance of embedding this in an educational programme will need to be accepted and extend beyond the focus of supporting the development of self-help skills. Strategies to support the development of self-organisational skills (made more difficult as a consequence of executive function difficulties according to Nicholas, 2005) is also seen as important, and Majors (2011a) suggests the use of checklists and a visual calendar to help keep the child organised.

2.6.3e Behaviour management

Aitken (2000b) addresses the issue of 'challenging behaviour' in deafblind individuals at some length, explaining it is more prevalent in this population frequently as a result of communication difficulties, potential brain damage, but also as a consequence of the actions of others (being too controlling etc.). In this chapter he offers a range of strategies that might be helpful in responding to difficult behaviours. These may well be strategies also appropriate for learners with CHARGE, but it will be important to

question whether there are other strategies additionally needed given the development of the behavioural phenotype by Hartshorne (2011) which suggests the behavioural characteristics come together in a unique way as a part of this condition. Smith et al. (2010) and Majors (2011a) encourage educators to recognise the particular behavioural characteristics of CHARGE and to respond by employing a 'Positive Behavioural Support' approach as described by Hartshorne (2003). Strategies established to manage a child's behaviour must not only consider the child but also the learning since, as Brown (2011) states, a child's behaviour is often:

Compounded by inappropriate responses from the educational world itself. (p.1)

This is also recognised by Hartshorne et al. (2007) who identify environmental factors as influencing the child's behaviour, including how those around respond to the child in addition to the physical environmental features (e.g. distractions, physical barriers etc.). Majors (2011a) also implies it is the behaviour of the educationalists around the child that requires recognition and for the teacher to respond to the child's learning traits, for example, negotiating and providing ownership of learning as a response to the child's need to be 'in charge'. Earlier in 2.3.4 insights were outlined that Hartshorne (2011) highlighted following his investigation into behaviour in individuals with CHARGE and the educationalist is required to consider how these can be developed further and lead to practical strategies that might be beneficial, supported by thorough investigative research in this area.

2.6.3f Delivery of the curriculum

For a curriculum to be productive and effective it must be delivered successfully. Whilst a wide range of specific strategies were not outlined in great detail in the

literature reviewed on the education of children with CHARGE, more attention has been given to how the curriculum should be delivered in broader terms. Lieberman et al. (2012) suggest combining a 'top down' approach (where the end goal is the focus) with a 'bottom up' approach (focusing on the process rather than the end goal). Although this study only examined physical education it may be appropriate for these findings to be applied to the overall curriculum. The use of both a 'top down' and 'bottom up' approach was also identified in a research study examining curriculum access for deafblind pupils (Porter et al., 1997), where it was found teachers delivering the curriculum for deafblind learners used a combination of both these approaches. This is a method that can perhaps best provide the flexibility and individualisation Majors and Stelzer (2008) and Majors (2011a) feel is required when educating children with CHARGE.

Both Majors (2011a) and Jones and Dunne (1988) suggest the use of task analysis to break a task down into clear structured steps, as also advocated for deafblind learners (e.g. Hodges, 2000). This is supported by others (e.g. Brown, 2003b; Smith et al., 2010) who identify the requirement for a clear structure to activities with a defined beginning, middle and end. Brown (2011a) further states it is essential that the teaching point is clear and accessible to the child. Breaking a task down into steps provides opportunities for short breaks if required by the child and may help match the 'burst-pause' learning style discussed in 2.6.2a. Lieberman et al. (2012) expressed the view that clear breaks need to be included when undertaking an activity which requires continuous movement (e.g. swimming and running), but that they may also more naturally occur in some other activities such as kicking and jumping. Related to

this is the need to identify what the task itself means to the child and ensuring it is presented in a way that is meaningful to him or her (Brown, 2003b). The use of structure and clear routines is also seen as vital in the educational programme of a learner with CHARGE, supported through the consistent use of a visual calendar system (Graham et al., 2005, van Dijk and de Kort, 2005; Smith et al., 2010; Majors 2011a).

Majors (2011a) identifies the need for flexibility to enable an immediate response to the child's fluctuating needs and ability to actively engage. She further asserts the need to alternate between preferred and less preferred tasks, together with access to favourite characters and activities to help reduce stress and anxiety levels. Monitoring the demands made of the learner with CHARGE and reducing them when necessary is important and Majors (2011a) suggests giving the child time to observe and accepting 'partial participation' initially, then supporting an increase in the level of involvement by the child. This relates to the use of modelling as an effective strategy to support learning, which allows the child to observe initially before engaging. This approach to delivering the curriculum would also be advocated by educators of deafblind learners as a whole (e.g. Murdoch et al., 2009; Nelson et al., 2010). A further element for the learner with CHARGE, identified by Stelzer (2011), is the importance of prioritising the creation of opportunities for the child to do things concretely. The use of concrete aids is also advocated for learners with severe and profound learning difficulties (e.g. Farrell, 2006), but there appears to be an additional or different reason for using this strategy with learners who have CHARGE; Brown (2011a) explains the use of concrete tools can help to overcome executive function

difficulties, and Majors (2011a) recognises this can also support the learner when they are distracted or upset which may prevent them from processing spoken/signed information clearly.

Although adapting the environment in response to a child's visual and auditory needs has already been identified, there is also a need to consider adaptations more broadly, considering the physical environment, the people within it and the activities being presented, that would be deemed good practice for all deafblind learners (e.g. Hodges, 2000; Eyre, 2000; Murdoch et al., 2009). For learners with CHARGE this may necessitate setting the environment up so the individual is free to 'perform differently' according to their unique individual learning style and preferences (Smith et al., 2010; Majors 2011a; Stelzer, 2011; Brown, 2012).

2.6.4 Summary

This section considers the information available in literature with regard to the education of children with CHARGE. It illustrates a lack of evidence-based research in this area and the current reliance on anecdotal reporting and sharing of opinion based on direct experience that whilst valuable, is limiting because it lacks a foundation in sound research.

Within the sources examined it is suggested it will be challenging for educators to meet the educational needs of learners with CHARGE. The heterogeneous nature of this condition, reflected in the range of ability and needs, means it is possible that one defined curriculum will not be appropriate for all learners with CHARGE. Although use

of the deafblind educational model is identified as beneficial and appropriate in many aspects, if there is a distinction between learners with CHARGE and other deafblind learners, it is possible that there are additional and/or different elements to be addressed. Through comparing the strategies outlined for learners with CHARGE with established teaching practice for deafblind learners, it is clear there is support for Brown's (2011a) view that this may be the "best match", but that there are also elements that require a response to those "CHARGE-specific considerations" (Brown 2012). This review of literature suggests these may be a greater impact of vestibular and sensory integration issues, and also of fatigue. It is also possible there are additional aspects such as the use of strategies to help overcome executive function difficulties that can adversely affect learning that require a specific response. Given the theory of a CHARGE behavioural phenotype (Hartshorne 2011), that by definition is specific to CHARGE, it is possible this too would require a specific response. Finally, it is also possible that some of the strategies used with deafblind learners may also be used to support learners with CHARGE, but for a different reason; for example, large movement may be used with deafblind learners to support the development of gross motor skills (e.g. Aitken, 2000a; Murdoch et al., 2009), but also used with learners with CHARGE in response to their vestibular and sensory integration difficulties (Brown 2011a).

This literature review has demonstrated a need for more research to explore this area; Griffin et al. (2004) contend that as researchers discover more about the needs of learners with CHARGE so this will enable a broadening of the range of assistive technology interventions available to them. If we consider this in the broader context,

information obtained through evidence-based research is needed to develop theory and increase the knowledge base and understanding of the learner's overall needs and underpin practice as it informs the development of more effective intervention strategies across the curriculum and learning environment. Whilst it is important to consider the reporting and anecdotal evidence of practitioners experienced in the field of educating learners with CHARGE, as Smith et al. (2010) state, "teaching methodology should be evidence-based" (p.299) with a foundation on research-based intervention strategies.

2.7 Final summary and conclusions

In this chapter an exploration of literature was presented to discover what is known about individuals with CHARGE with regard to their learning, development and education. A range of research studies relating to the medical aspects of CHARGE were found that have led to an increase in knowledge and understanding of this as a medical condition. Some investigations were also discovered regarding specific aspects of development and functioning in individuals with CHARGE, but research undertaken that focused on the education of this particular population proved to be extremely limited. As a consequence it was necessary to look at anecdotal reporting and the sharing of opinion concerning educating learners with CHARGE although it was recognised these articles were not so rigorous academically and were not founded upon evidence obtained through research. This situation did however, help further with the identification of the "problem space" (Petre and Rugg, 2012) mentioned in 1.4, and thus informed the direction of this research project.

Some scientific investigation involving clinical tests with regard to medical aspects of this condition was reported and other research studies that employed single or multiple case study or document and data analysis. The majority of research studies examined, however, employed a survey method through the use of a questionnaire. This would likely provide easier access to a wider sample but also be utilised because children with CHARGE were found to be more difficult to accurately assess or obtain information directly from. It would therefore be more realistic to focus upon obtaining information from parents and practitioners, despite this often involving gathering perceptions from participants that would be influenced by their own experiences, world view and knowledge base. It was felt it would be useful to follow this example and include the use of a survey within this research project to obtain data relating to a wider sample of individuals with CHARGE.

The heterogeneous nature of CHARGE was described but also the view that commonalities can be found between affected individuals, as expressed in literature. It was shown that potentially all the sensory modalities can be affected, leading to these children being described as 'truly multi-sensory impaired' (Brown, 2003b). Other aspects were also identified, all of which demonstrate that more than sensory issues will very likely have an impact on the child's learning and need to be recognised and responded to by educationalists. Literature was also explored to ascertain whether these anomalies might also be observed within the wider deafblind population (or children with single sensory impairment or autism). Some similarities were noted (for example, the presence of sensory integration dysfunction), but it was found likely that others are more specific to CHARGE (for example, the cranial nerve anomalies, and

also neurological anomalies causing executive function difficulties). It was also suggested that in addition to those anomalies which might be regarded as more particular to CHARGE, it may be the combination and severity of impact of these anomalies that is more unique to CHARGE and more intensified and complex. Thus a potential cumulative effect was noted where the interplay between these different issues was a factor, with the aspects of development discussed likely to be affected to a greater or lesser extent in different children with this condition. Although considered a part of the deafblind population, the generally held view in the literature examined is that individuals with CHARGE form a distinct group, with affected individuals having many other issues in addition to deafblindness. The opinion shared is that CHARGE demonstrates a unique combination of medical and physiological conditions resulting in a developmental course that differs from other deafblind learners (Hartshorne et al., 2011).

Sources were examined to identify what is known about the development and functioning in children with CHARGE and the different aspects likely to be affected were reported. With the large number of anomalies involved with this condition it was perhaps not surprising to find that learning and development would be affected in this group of children. It was discovered that some key aspects of development and learning have been identified as being affected. Understanding of the potential impact on learning and development of visual impairment, hearing impairment and multi-sensory impairment is well-developed and will need to be applied to learners with CHARGE. The impact of other anomalies associated with this condition also need to be considered and when combined, may have a greater influence on the child. For

example, the average age of independent walking in children with CHARGE was reported to be 35-57 months as compared to 15 months in typically developing children, or 19-20 months for children with visual impairment/blindness.

A range of cognitive ability has been identified with this condition, and it was recognised that learning potential may be under-estimated or masked by the challenges and barriers to learning created by the overall impact of the different anomalies. The educational assessment of individuals with CHARGE has been addressed in literature and this was also examined. It was found it is agreed that standard assessment tools are likely to be inappropriate and unhelpful for this group of children, with a difficulty obtaining accurate results using assessment tools not standardised for the CHARGE population. Although an alternative approach was reported to be needed it was recognised it can be difficult to obtain precise information that is not biased by subjective judgement. It was noted that educational assessment of learners with CHARGE is likely to require a combination of formal and informal assessment, interviews, observations etc.

Finally, information relating to the education of learners with CHARGE was addressed. It was recognised this was drawn from anecdotal reporting and opinion since very little evidence-based research was found relating to educational intervention. Some reference was made to a developmental and cross-curricular curriculum model that may be most appropriate for learners with CHARGE. A deafblind educational model was considered to provide the 'best fit', but that there may be a need for a different emphasis and additional elements. Those strategies identified within the sources

examined with regard to educating learners with CHARGE were compared with strategies outlined in literature focused on educating deafblind learners as a whole. A clear overlap was noted, with a number of the same strategies identified for both groups. It was, however, also noted that some elements may require a greater emphasis (e.g. responding to the child's vestibular and proprioceptive difficulties), and that some additional strategies may need to be implemented in response to those more "CHARGE-specific considerations" (Brown, 2012). It was further suggested that some strategies may be employed with both groups of learners, but possibly for different reasons. It was recognised there is currently a lack of research that has investigated the education and teaching strategies to support learning and development in children with CHARGE, and that more research is required to explore this aspect so as to increase the knowledge base and inform and underpin practice.

This exploration and review of literature provided a wider understanding of what is currently known or thought, that would help begin to address the over-arching research question of whether CHARGE syndrome can be considered as an educational diagnosis. As much of this information, particularly in relation to the educational setting (and to a lesser extent, a child's functioning) came from anecdotal reporting and opinion, there is a need to investigate further and establish a clearer evidence base. As a result of this review of literature a series of sub-questions were developed to assist in addressing the over-arching research question, these being:

- What are the factors within the child likely to affect learning?
- What strategies are likely to be effective in supporting the learning of children with CHARGE?

- What aspects of CHARGE might be distinct from the more general deafblind/MSI population?
- What specialist support is provided from other professionals and through assessments, to teachers working with a child with CHARGE?

In the next chapter a description will be given of how the research framework was developed to generate the evidence needed to address these research questions, including the identification of appropriate methodology and methods, and considering how trustworthiness could be established.

CHAPTER 3 – DEVELOPING A RESEARCH FRAMEWORK: METHODOLOGY, METHODS AND ETHICAL ISSUES

3.1 Introduction

Following the review of literature the next significant step in this investigation was to establish the research framework, identifying appropriate methodology and methods that would generate the evidence needed to address the research questions and build upon the examination of literature. The exploration of literature revealed a lack of knowledge arising from evidence-based research as to what enables a child with CHARGE to learn effectively, and that presently educational professionals rely largely on anecdotal evidence and the application of strategies that have proved successful with other congenitally deafblind learners. It was recognised this research project would need to focus on adding to this limited pool of knowledge.

The nature of the population being explored (with CHARGE being a low incidence condition) and the potential difficulties this presented to the researcher in accessing a wide sample, meant this would be a small-scale study (Cohen et al., 2003), these factors influencing the choice of a methodological approach appropriate for this research project. There are concerns raised about the limitations of small-scale study and whether findings can be widely generalised (Robson 2011) and it was considered this research may in reality need to focus on providing deeper 'insights' if it was not possible to obtain a truly representative sample of the population of individuals with CHARGE that would allow for generalisation of the findings. It was intended the research framework would be developed to support the gathering of

evidence to respond to the sub-questions that arose from the literature review (as set out in 2.7) that in turn would help address the overarching research question of:

CHARGE syndrome is a medical diagnosis.
Can it also be considered as an educational diagnosis?

In this chapter the following will be presented:

- An outline of the choice of methodology
- A broad outline of the choice of methods employed, including data analysis processes
- A diagram to illustrate the research process undertaken
- A detailed description of the processes undertaken within Cycle 1 and Cycle 2 of this investigation, including the collection, collation and analysis of data, and validity and reliability checks
- A description of the actions taken to ensure the trustworthiness of this study
- An explanation of the ethical issues that were addressed

3.2 Choice of methodology

The establishment of the research questions this study would seek to address employed a common social science process (Creswell, 2013) of:

- Establishing a central question (that identifies the central phenomenon of the investigation)
- Developing associated sub-questions (that help direct the focus of the investigation)

Once the research questions were identified it was necessary to establish the research design that would define the structure of this investigation. It was recognised this research would be both exploratory and descriptive in nature (rather

than causal or relational) with an emphasis on discovery, since the early stages of investigation in a subject area will need to be dominated by research focused on accumulating knowledge and strengthening understanding. According to Cohen et al. (2003), educational research as a whole is a relatively new field and currently consists of a largely descriptive element. This will be particularly true when the educational research is focused on a condition such as CHARGE, a rare condition only relatively recently identified (see 1.1)?

There have been attempts to separate and clearly distinguish between 'scientific' and 'naturalistic' research, although more recently an easing of the tension between the two has been recognised, with both being regarded as valid (e.g. Robson, 2011). This accepts they do not necessarily need to be regarded as being in direct opposition and the various methodologies can:

Nest into and envelop each other. (Thomas, 2011, p.36)

The choice of framework adopted was influenced by the exploratory nature of the investigation and also the particular research questions posed. It was considered it would not be appropriate to design and undertake a controlled experiment as in the traditional 'scientific' approach (as described by Robson, 2011) since, in addition to being difficult to implement within this research setting, it would not lend itself to addressing the questions posed and would be unlikely to reflect the real world (Mason, 2006).

Since the questions were not seeking to discover any patterns, developments or changes over time, a longitudinal design (e.g. Cohen et al, 2003) was not considered

to be appropriate. Similarly the questions did not focus on the bringing about, and impact of, change and therefore the use of action research (e.g. Robson, 2011) was also dismissed. What the questions (and therefore investigation) sought to explore was the possible distinctiveness of learners with CHARGE compared to the wider deafblind population, and to gather detailed information about this phenomenon. Thus for the purposes of this investigation, a case study design frame was adopted that would support “in-depth research” (Thomas, 2013, p.150) of a ‘real life’ happening and a gathering of evidence to explore the phenomenon being examined. Whilst the case study may not be regarded as traditionally scientific, some (such as Robson, 2011; Thomas, 2011) argue it may be regarded as broadly scientific if the case study is used to ensure an effective response to the questions posed and demonstrates high quality evidence and reasoning.

As explained, case study involves in-depth exploration where the emphasis is upon explanation, remaining focused on the subject being explored (Verma and Mallick, 1999; Thomas, 2011). It allows for a detailed examination of the ‘case’ from different angles (Yin, 2008), and is:

Empirical in the sense of relying on the collection of evidence about what is going on. (Robson, 2011, p.179)

Thomas (2011) explains there are no procedures that are exclusive to a case study. For this investigation the intention was to work inductively where the insights and theory would arise and develop from the research itself, rather than working deductively and proposing a pre-defined hypothesis to try and prove or disprove through the investigation (Cohen et al., 2003).

The case study provides a framework within which the researcher is able to work flexibly (Robson, 2011) and enables the processes employed to emerge as the investigation proceeds. The degree of flexibility offered by this is also regarded as suitable for the exploration of new topics and ideas such as the topic of this investigation (Robson, 2011). A key characteristic of the case study is the use of multiple methods (to be discussed below) that Yin (2008) identifies as one of its strengths that provides a 'multi-faceted' view (Thomas, 2011, p.5) and supports triangulation (see 3.9).

The 'case' might be an individual, group, event, institution etc. (Gillham, 2000) that forms a 'characteristic unit' (Thomas, 2013, p.150). According to Thomas (2011) the case will be bounded (the subject) and framed (this providing the context of the investigation). With this in mind, the 'case' for this investigation would be CHARGE as represented by a selection of learners (with a confirmed medical diagnosis of the condition) and the views of practitioners.

Thomas (2011) argues there is no requirement or intention (and indeed it may not be possible) to generalise the findings of a case study. Whilst this is not likely to be possible using the case study of an individual, Yin (2008) argues it may be possible to achieve broad generalisations by studying a case that involves a larger selection, this argument being supported by Verma and Mallick (1999) who feel that if the case study is undertaken rigorously and systematically its findings may be generalised. There are inherent difficulties when research is focused on a low incidence condition that would need to be acknowledged. Nonetheless when analysing and discussing

the findings of this investigation it would be explored whether the cohort of learners with CHARGE reported on might be similar to the wider CHARGE population and thus allow for a tentative generalisation of the findings to other learners with CHARGE (the issue of generalisation will be discussed further in 3.9.7).

3.3 Choice of methods

It has been explained that within the case study framework, multiple methods may be employed that enrich the investigation and also support triangulation, and tools were chosen according to their relative strengths to generate the data and analysis that would secure the best responses to the research question and sub-questions. It was intended that both qualitative and quantitative data would be sought (as advocated by Robson, 2011), this being beneficial to the enquiry with both complementing each other (Thomas, 2013) and supporting the achievement of a deeper answer to the research questions.

Some methods adopted in the course of this research may be associated more readily with fixed design, such as a survey (Oppenheim, 2000), whilst other methods employed (document analysis and semi-structured interviews) are perhaps more often regarded as ethnographic, involving the collection of data to gather people's perceptions in seeking to investigate and explain this particular context. It was considered these methods could be suitably employed on a smaller scale as identified by Robson (2011) within the case study.

3.3.1 Use of documentation

For this research project, initially raw data (more qualitative in nature) would be collected through analysis of documentation in the form of educational reports provided for a number of children with CHARGE. Edwards and Talbot (1999) explain the potential value in examining documents as part of a research investigation should not be under-estimated. Thomas (2013) refers to this method as “document interrogation” (p.204) and it was considered this would be useful for providing a background and the initial exploration, whilst also being unobtrusive and non-reactive, that would help minimise any potential researcher effects (Robson, 2011). Although of benefit in that it could provide valuable information with regard to the subject being studied, it would also be important to recognise any potential weaknesses and limitations that may exist. For example, the data extracted would be dependent on the authors accurately reporting observations and may only be representative of the authors’ perspectives that may not be regarded by others more widely. The process used to analyse the data extracted from these documents is outlined in 3.3.3a below.

3.3.2 Surveys

Surveys would be employed in the form of both a questionnaire and interviews to obtain information from two different samples of the population of educators involved in directly supporting a child with CHARGE.

3.3.2a Postal, self-completion questionnaire

A questionnaire-based survey was chosen as a relatively simple and straightforward way of gathering a large amount of information from a wider number of participants (Robson 2011). The questionnaire was intended to be circulated to educators supporting a child (with a confirmed medical diagnosis of CHARGE) across the key stages from Early Years through to Key Stage 4 and to include children in a range of educational settings, with the intention of securing a broad representation. It was considered a questionnaire survey was a tool that would enable relatively easy gathering of data at low cost, and provide the respondent with a level of anonymity that might encourage more open and frank responses (Cohen et al., 2003; Robson, 2011). It was considered a postal, self-completion questionnaire would allow respondents time to access their personal records and complete the questionnaire at their own pace and convenience. Before finally deciding to make use of a postal, self-completion questionnaire other alternatives were considered, these being use of a face to face or telephone questionnaire, or use of an online survey. Implementing a questionnaire either face to face or via the telephone is considered likely to have a better response rate than a postal self-completion survey and would give the researcher the opportunity to provide any clarification needed, and also monitor the quality and ensure good recorded responses (Cohen et al., 2003). This type of survey is, however, labour intensive and the sheer practicality of meeting every respondent or talking to them individually over the telephone meant the survey would be more limited in its sample size or need to be more clustered geographically for face to face interviews to be undertaken (this would prove difficult given the low incidence of this condition). This would also not enable responses to be given

anonymously and this, combined with the likelihood that the researcher's own characteristics could also more greatly influence the responses, may create a biased response.

Use of the internet is now widespread and readily available to many people, and an online survey could offer advantages over more traditional surveys in providing access to individuals in distant locations and give the convenience of automated data collection that would reduce the demands on the researcher's time and effort. It could also provide the facility to move from a paper format to an electronic medium (Thomas, 2013). There are, however, also potential disadvantages some of which, according to Wright (2005), are more unique to the computer medium. It would be more difficult to monitor the participants to ensure that respondents were within the targeted sample group (i.e. practitioners supporting a child with a medical diagnosis of CHARGE) or to insure against multiple responses. Building in a process to overcome this issue, such as requiring participants to contact the researcher to obtain a code, was felt likely to reduce the response rate. It would also be difficult to track and ascertain the true response rate which in turn would create difficulty in following up on potential participants who did not respond initially. There was also a concern that the design, implementation and evaluation of an online survey would be limited by what each online survey package offered; having identified a wide number of potential sites it was found to be difficult to ascertain which would be the most suitable/effective for this particular research goal. Additionally, although most companies offered free customer support there were generally set limitations and additional support would require a further fee. From the researcher's experience of

using NVIVO for analysis in Cycle 1 of this research (described in 3.3.3c) there was a reluctance to rely on a tool that could require advice and support in this way since this had been time consuming and, on occasions, difficult to obtain when needed.

In using a postal, self-completion questionnaire it was recognised a number of aspects would need to be considered and addressed in relation to the design and validity. Firstly, whether the respondents completed the questionnaire accurately; this would be addressed by testing the wording of the questions used to promote a shared and common meaning and then piloting the questionnaire itself before the final version was circulated (see 3.6). A check for consistency of responses would also be built into the questionnaire (as explained in 3.6.9) as an indicator of the reliability of the responses given. Secondly, the issue of potential non-response would need to be addressed to help secure a positive response rate. It is often stated that postal self-completion surveys tend to have a low response rate (e.g. Robson, 2011), although Cohen et al. (2003) explain this does not have to be the case, and every effort would be taken to overcome this potential weakness of this type of questionnaire. There was a risk respondents would not give the questionnaire serious consideration so it would be necessary to reinforce the importance of the study and the respondent's involvement in the covering letter provided, whilst also highlighting the potential benefits to the respondent at the completion of the investigation. The questionnaire would also rely on the quality of its presentation and question structure to encourage people to respond (both of these issues are addressed in more detail in 3.6). It would also be necessary to establish a clear time line and actively seek to secure responses within the time frame established to

reduce the risk of there otherwise being a long data collection period that could slow the whole research process down.

3.3.2b Semi-structured interviews

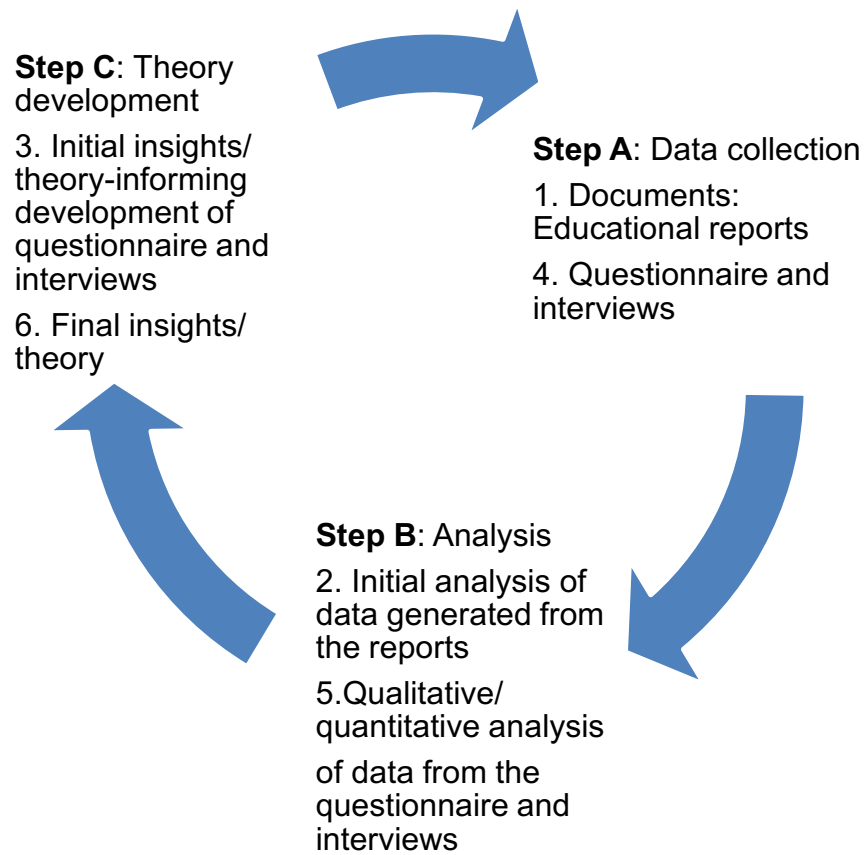
The interviews were to be undertaken with practitioners working with a number of pupils with CHARGE in an educational setting in the USA, to gain the views and opinions from practitioners in a different educational system, and consider other possible explanations (Kolb, 2012).

Thomas (2013) outlines three basic types of interview that were considered in turn when preparing to gather information using this method. 'Structured' interviews where the question and answer categories are set in advance (Cohen et al., 2003) would potentially limit the interviewees' responses and therefore the information gained. 'Unstructured' interviews with little or no structure provided from predetermined questions would most likely generate a wide range of responses on different topics that would be difficult to both organise and analyse (Robson, 2011). It was therefore decided to employ a 'Semi-structured' interview with a list of initial questions that would be presented to each interviewee that would enable greater comparison of the responses given (Cohen et al., 2003), but also provide the interviewee with more freedom to express their perceptions and opinions. The development of questions for this interview would also arise out of the analysis of data obtained from the previous document analysis.

3.4 A summary of the research process

Within the case study framework, the process which emerged and methods employed are set out in Figure B illustrating the two clear Cycles identified for this investigation:

FIGURE B: THE RESEARCH DESIGN FRAMEWORK FOR THIS STUDY



Cycle 1 involved gathering data through document analysis, using documents in the form of educational reports, these being analysed using the constant comparative method as a process for the analysis (described in 3.3.3a). The initial insights and theory arising from Cycle 1 would then be used to inform Cycle 2 where the views of teachers involved in supporting a child with CHARGE in their class would be sought

through the use of a questionnaire. Additionally, semi-structured interviews of practitioners involved in educating learners with CHARGE in an educational provision in the United States of America would also be undertaken. The constant comparative method would again be used to analyse the data generated, with the findings supporting the development of the final insights and theory generated by the investigation to address the research questions.

3.5 Data analysis

The opinion that theory (or ideas) is intended to emerge from the research situation rather than beginning with more set ideas to prove or disprove through the research process, (i.e. working inductively rather than deductively) is at the heart of interpretive research (Thomas, 2013), and a part of this approach is the requirement to analyse the data generated by the research act. A method often linked to interpretive inquiry is the constant comparative method and this would be employed as a process to analyse the data obtained within this investigation. This method was originally developed to be used in grounded theory methodology originated by Glaser and Strauss (1967), and provides the researcher with a clear structure for analysing the data obtained. Grounded theory as a research methodology would require the researcher to work inductively and interpretively, building theory by gaining explanations driven through data, and is considered useful where there is little or no pre-existing theory to build upon. According to Corbin & Holt (2004) possibly the most valuable aspect of grounded theory is its ability to generate basic concepts to provide the foundation and first steps towards developing a new body of knowledge. This methodology may be considered particularly appropriate for research undertaken

within an educational setting as it enables a focus on practice where the explanations are grounded in reality. As a research methodology, however, it also has many critics (e.g. Layder, 1993; Robrecht, 1995; Thomas and James, 2006). Although Thomas and James (2006) recognise that use of grounded theory is highly regarded within educational research they express the concern that adhering to the procedures can undermine the researcher's "interpretation, narrative and reflection" (p.767). This is reinforced by Robrecht (1995) and Layder (1992) who identify the need to ensure there is not an over-emphasis on the structure, techniques and processes, but rather a primary focus on the data generated, ensuring the researcher is guided by the data generated, rather than being limited by it. Therefore, although a grounded theory methodology would not be adhered to for this research due to the risk of the research process and the findings being limited (choosing instead to use a flexible design as described previously in 3.2), the constant comparative method was chosen as the method to analyse the data obtained, a process which Kolb (2012) states enables theories to be formed, enhanced, confirmed or discounted. Further support for this decision may be found in the work of Thomas (2013) who states:

Constant comparison is the kernel of grounded theory worth preserving.
(p.239)

Use of the constant comparison method would involve breaking the data down into discrete units that would be compared to other data obtained, allocating codings and creating categories, then developing connections between those categories with the intention of building up insights and supporting the formation of theory. Boeije (2002) feels "the term 'constant' might be a slight exaggeration", (p.406) but it was intended that regular comparison of the data obtained would be undertaken.

3.5.1 Data analysis in Cycle 1

Using the constant comparative method in Cycle 1, analysis of the data obtained from the reports was supported by the development of coding categories. In the first stage of the process, often referred to as 'Open coding', the aim was to begin organising the data extracted from the reports and developing the initial categories of information about what was being studied. At this stage a code was applied as a label to each piece of data in terms of its content. Within this research for each piece of data drawn from the educational reports the question was asked "what is this piece of data an example of?", and a label or 'code' then applied. This label was provisional in the initial stages and was open to change or further refinement as the research progressed. It was also accepted that one piece of data may have several codes and could fall into more than one conceptual category.

With the codes taking shape and being identified during the process of open coding comparison was then undertaken to begin looking for links/associations, thus beginning the process of 'Axial coding'. At this stage the analysis shifted towards identifying key (axial) components, looking for commonalities and how these concepts might come together, and it was recognised there was a need to have several main categories containing further sub-categories. The originators of grounded theory had diverging views about the process used to create these axial codings; Glaser (1992) felt the form the axial codings take would emerge from the data and not from being put into a pre-determined format, whereas Strauss and Corbin (2007) advocated the use of external tools and resources to help structure and interpret further the labels obtained through the process of open coding. Within

this study, during the coding process it was felt it would be helpful to adopt this latter strategy using the domains identified in a curriculum for learners who are multi-sensory impaired by Murdoch et al. (2009) as a structure. These categories would ultimately then be refined further and used to structure the questionnaire to be sent out to teachers supporting a child with CHARGE.

Although a distinction is made between open coding and axial coding it was recognised they were occurring almost simultaneously since whilst applying an initial open code to a piece of data, possible connections to other pieces of data and concepts were also being identified. The number of reports used within this document analysis enabled the point to be reached where the categories were eventually becoming 'saturated' and any continuing analysis was only likely to produce diminishing returns (Kolb, 2012). A further level of coding ('selective coding') could then be pursued that would involve concentrating attention on a core category, selecting that which was felt to be most representative and significant with the intention that this would then become the central focus of the analysis (Robson, 2011). However, for the purposes of this investigation it was felt there was a risk of the research process and the findings being limited if the direction taken was towards concentrating the ongoing study on a 'core category' as this could result in the exclusion of other potential important aspects. Thus the analysis of the data extracted from the reports was halted at this stage, and all the coding categories/sub-categories established then used to support the development of the subsequent questionnaire and interviews, where each of these aspects could be explored further.

3.5.2 Data analysis in Cycle 2

The development of the questionnaire was informed by the findings and the coding categories and sub-categories that emerged within Cycle 1 together with the information gathered from the literature review. The qualitative and quantitative data generated from the questionnaire was analysed through the use of statistics and by continuing the constant comparative method begun in Cycle 1 and described above. Similarly, the responses provided by the interviewees were recorded and categorised according to the questions that had initiated this information, and further comparison was then made between the interview responses obtained and also by comparing these responses to the findings of the questionnaire.

3.5.3 Use of Computer software for data analysis

Robson (2011) identified that investigations undertaken in the social sciences can generate a lot of qualitative raw data, and it was recognised this would need to be managed efficiently to facilitate effective use of the constant comparative process. Denscombe (2003) recommends the use of computer-aided analysis when using this process and following this advice possible software was identified that would support the data analysis and coding process, including Atlas.ti, SPSS and NVIVO. Eventually a computer software package called NVIVO (produced by QSR International) was chosen since it specifically claimed it could be used with the constant comparative method with the computer program developed based upon this process. Following discussions with another researcher who had experience of using NVIVO, it was considered the program would be flexible enough to be adapted to meet the needs of this study and would not restrict the analysis by forcing codings

into a hierarchical tree structure, but rather enable the undertaking of open coding and axial coding through use of 'free nodes'. NVIVO proved to be a useful tool in recording and supporting the coding of the statements obtained from the reports, however, an ongoing difficulty was encountered in that the system 'crashed' on several occasions and this resulted in halting proceedings, until assistance had been obtained from the support team (in Australia!). This was not an aspect that had been considered when choosing the software to support the analysis, but would need to be recognised when identifying computer software that might need to be utilised in the future.

For Cycle 2 of this investigation it was recognised it would not be possible to collate or analyse the quantitative element of the data obtained using the NVIVO program, and this, combined with the difficulties experienced when this program, supported the decision to consider other computer software packages. Consequently at this stage the collation and analysis of data from the questionnaire was supported through use of Microsoft Excel, a spreadsheet application that would also enable analysis of the quantitative data in addition to the qualitative data obtained. It also had the added advantage of being a software package the researcher was very familiar with. An initial framework for the recording and analysis of both the qualitative and quantitative data generated by the questionnaire was established and designed using Excel to develop a spreadsheet. This was structured so as to enable statistical analysis that included establishing formulae to calculate frequency of occurrence, mean and median (mode was easily identified from frequency figures) and scorings where a Likert scale was used. The spreadsheet was also structured

and coded to enable cross-referencing that would support the ongoing use of the constant comparative method.

3.6 Cycle 1

Within this first Cycle the steps were identified as Step 1 – Data collection using educational reports; Step 2 – Analysis of the data obtained, developing codings of statements extracted and the initial analysis (what the reports said); Step 3 – the development of initial insights and theory that would inform the development of the questionnaire and interviews in Cycle 2 of this investigation.

3.6.1 Data collection

Working as a teacher for multi-sensory impaired children for a national specialist organisation who support and campaign for deafblind people and their families and carers, provided the researcher with access to numbers of children with CHARGE. Whilst acknowledging that not every child with CHARGE will be deafblind, according to literature the vast majority do have combined visual and hearing difficulties and would therefore be considered as part of the deafblind population. (Hartshorne et al., 2011, reported this to be over 90%). This implied that in the first instance approximately 10% of individuals with CHARGE would potentially be excluded from the study. It was also recognised that focusing on children with CHARGE known to this specialist organisation would not provide access to children with CHARGE not supported by the organisation and this would be a limitation of the study at this point. Data for the later phase of this study would, however, be sought from a wider number of potential sources to provide access to a larger cohort of children.

The researcher had an obligation to her employers to ensure they were kept fully informed of the processes that would constitute this research study. As it was intended to contact families where the child received support from the organisation it was ensured consent was obtained from the nominated director before progressing any further.

3.6.2 Identification of subjects and educational reports

Having obtained consent from the organisation by whom the researcher was employed the organisation's database was then accessed to identify those children supported who had a confirmed diagnosis of CHARGE.

Thirty-eight children with CHARGE were identified with whom the organisation had been involved until the point this research project began. Of these children a number were excluded for the following reasons:

- 6 children aged 16 years or older, due to potential implications relating to the Mental Capacity Act (2005) with regard to obtaining informed consent
- 1 child who was very young and not yet placed in any educational provision according to the definition set for the study
- 4 children who had either moved and there was no current address, or the family had moved out of the country
- 1 child who was a 'Looked After Child', where there were likely to be issues relating to obtaining consent and ensuring complete anonymity

The intention was to extract data from reports provided within the context of each child's educational placement that reflected the definition set by the researcher for the 'educational environment' (see 1.6.3) that set the context for the research project itself. These reports included:

- Reports following a home visit (that related to the educational environment)
- Reports following a school visit
- Assessment reports
- Contribution towards Educational Advice for the child's Statement of Special Educational Needs

For the 26 children considered eligible for this research project, reports were identified that fulfilled these criteria. Only reports written between 2002 and September 2008 were considered; earlier reports could not be accessed through the central database available to the researcher, but it was also felt important to only use reports written prior to the start of this research project (September 2008), as it was felt the knowledge that reports might be used as part of the research project could influence the report content. It was also recognised that knowledge relating to CHARGE had increased over the previous few years and was likely to be reflected in the reports generated.

3.6.3 Obtaining consent

A request for consent to use identified reports was sent out through the post to the family of each child provided in the form of a letter (naming specific reports) and an information booklet (Appendix 1), together with a form to sign and return in the

envelope provided. The information booklet provided clear details of the purpose and content of the study, how the research would be used, and to whom it would be reported. It also addressed issues of confidentiality and anonymity, together with an assurance that consent could be withdrawn at any time. Consent was received from 17 families with no response from 9 families, providing a response rate of 65%. This resulted in access to a total of 58 reports.

3.6.4 Authorship of the reports

Eight different authors were involved in contributing to these reports (all being qualified multi-sensory impairment teachers), with the number of reports contributed to by each author shown in Table 7 below.

TABLE 7: NUMBER OF REPORTS CONTRIBUTED TO BY EACH AUTHOR²

Authors:	A	B	C	D	E	F	G	H
Number of reports contributed to:	33	9	1	5	6	8	2	1

Some reports had been co-written by more than one author and it also became clear the researcher (Author A) had contributed to a far greater number of reports than other authors. Although the reports had been written without the realisation that they would be used as part of this research study it was important to acknowledge the researcher's particular interest in CHARGE and personal viewpoint could potentially influence the report content. It would therefore be necessary to identify whether these opinions were also expressed by other authors. Consequently, a record was kept of which authors had expressed each statement extracted from the reports overall that

² Reports may have been written by a single author, whilst others were co-authored by 2 authors

would reveal which statements were authored solely by the researcher, or where the researcher was involved as an author of a statement on over 50% of occasions where the statement was noted. These statements would then be examined further and the views of other educators sought in relation to these as a part of the questionnaire and interviews in Cycle 2 of the investigation.

3.6.5 Limitations of the reports

Consideration was given to the limitations in the use of these reports. It has already been identified that reports would only be available about children supported by the organisation the researcher was employed by. This would be addressed later in the investigation by ensuring a wider pool of potential participants was sought in Cycle 2. It was accepted the reports may not provide straightforward factual evidence and were likely to have been constructed to convey the view of the authors and an interpretation of their observations. Additionally, the content of the reports would draw on the knowledge and experience of each author and may reflect any particular specialisms but also gaps in their current knowledge and understanding of CHARGE. It would also not be possible to identify clearly whether statements were the authors' opinion in relation to the specific child or they were sharing information extracted from literature. All authors, however, had obtained a specialist qualification in the field of multi-sensory impairment/ deafblindness and, on balance, it was considered that as these reports were written by a number of MSI specialist teachers with experience over a number of years of working with children with CHARGE they would enable the generation of data needed for initial analysis and interpretation.

3.6.6 Extracting the data

The 58 reports were numbered and names removed to protect anonymity as far as possible and all recommendations and statements relating to the child's learning and educational placement then extracted and numbered for easy reference. For example, 21:6 represented report 21, statement 6. Each child was used as a unit of analysis rather than separate reports since the number of reports written for each child varied. Cross-referencing between all reports for a particular child was then undertaken to remove any duplication of statements for any one child and avoid multiple recording of data. Having collated the statements extracted from the reports it was then possible to begin the process of analysing the data.

3.6.7 Data analysis

The constant comparative process can be used to extract both qualitative and quantitative information (Glaser and Strauss, 1967; Robson 2011) and it was intended both types would be generated through this analysis. When beginning to analyse the information extracted from the reports it was important to consider whether the statements would be interpreted in a literal, interpretive or reflexive way (Mason 2006). Literal reading would enable the researcher to extract content directly from the reports, but a wholly literal approach was prevented by these reports already reflecting an interpretation of a situation and the child's needs, and providing an opinion. It was also necessary to undertake an element of interpretive reading, to construct and document a version of the meaning intended for each extract that would enable more effective coding. This, in turn, was likely to include an element of reflexive reading since the researcher would be an informed reader whose personal

experience and knowledge would have an impact on the process of generation and interpretation of the reports. Thus, it was recognised that a combination of these different levels of interpretation would be employed.

3.6.7a Qualitative and quantitative analysis

Within Cycle 1 the main focus of this process was on qualitative analysis that would give consideration to all statements extracted and include those not frequently made but which may be relevant and raise an important point. Quantitative data was gathered to identify the frequency of statements and the number of children within the study for whom they were made. It was recognised some caution was needed in analysing this data quantitatively to ensure the results were not skewed due to:

- Differing numbers of reports being available for each child with varying numbers of statements extracted from them. Therefore it would not be possible to compare the overall number of statements provided for each child.
- Each coding content being likely to have different breadth and weighting (with some categories broader than others, and some broken down into sub-categories where felt necessary) so it would again be difficult to compare the number of statements provided for each category.

Although it was felt it might be beneficial to look at the number of different children for whom statements were made within each category, this would only provide limited information. If there was a heavy reliance on the quantitative analysis it was felt possible that other statements of potential importance to the study (but not frequently raised), could be missed. Therefore, although the quantitative information gathered

may be helpful it was considered this would only be used to support and supplement the qualitative data generated in the pursuit of addressing the research questions.

3.6.7b Developing the coding categories

Initial attempts to establish some form of structure for the coding process proved to be very difficult due to the diversity of statements extracted from the reports. Thus, as advocated by Strauss and Corbin (2007), an external tool was identified to help structure the codings, this being 'A curriculum for multi-sensory-impaired children: from Victoria School Birmingham' (Murdoch et al., 2009). This was chosen as it is a curriculum specifically designed for learners who are multi-sensory impaired and was well-trialled in a range of settings with children of differing needs. In addition it identifies key factors and strategies in supporting pupils' learning. Within this document, different areas of the curriculum are set out in eight 'domains' and a 'crib' sheet containing a summary of each domain (and additional categories) was composed for reference during the coding process. Each of the eight domains were used as the main categories for coding and as the 'open coding' process progressed these categories were then examined to identify potential relationships both within them and across different categories (the process of 'axial coding'). During this process, within each of these domains further sub-groups were established where the researcher felt refinement of the category was needed. For example, within the domain 'Orientation, Movement and Mobility', further sub-groups of 'gross and fine motor' and 'physical positioning' were added that might reflect the vestibular and proprioceptive difficulties, hypotonia etc raised by different authors. Three other main

categories were also added to support the coding process and facilitate the identification of relationships between the different statements, these being:

- CHARGE syndrome (for specific statements concerning CHARGE (e.g. “regular consultancy sessions for the whole staff team from a teacher qualified and in multi-sensory impairment who also has knowledge of CHARGE syndrome”))
- Support for learning (with additional sub-groups)
- Other (to include health, ASD etc.)

Statements could be assigned to more than one coding category if the content of the statement related to more than one aspect addressed. The initial coding categories established during the process of open and axial coding can be seen in the first column of Table 8 overleaf.

3.6.7c Undertaking a check of validity and reliability

The coding process, using NVIVO, was piloted with 9 reports (16% of the total reports). At this point a check of validity and reliability was introduced where a colleague (MSI teacher) was given the same collation of statements from the 9 reports and requested to code the statements provided according to the list of coding categories provided. A copy of the ‘crib’ sheet containing a summary of the content of each category was also given. The level of agreement/disagreement between the allocation of statements to particular codings would reveal the degree to which the codings might be consistently used by different researchers with similar results obtained, and would identify any potential overlap between the categories.

TABLE 8: A SUMMARY OF THE INITIAL CODINGS GENERATED DURING THE PROCESS OF OPEN AND AXIAL CODING AND THE REVISED CODINGS DEVELOPED FOLLOWING THE CHECK OF VALIDITY AND RELIABILITY OF THE CODING PROCESS

Initial codings	Revised codings
CHARGE	CHARGE
Communication	Communication
Conceptual development	Conceptual development
Orientation, Movement and Mobility Sub-groups: gross & fine motor physical positioning	Orientation, Movement and Mobility Sub-groups: fine motor physical positioning
Ownership of learning Sub-group: individual learning style	Ownership of learning Sub-group: individual learning style
Responses to routines and changes	Responses to routines and changes
Sensory responses Sub-groups: hearing and vision multi-sensory impairment other senses sensory integration vestibular & proprioceptive	Sensory responses Sub-groups: hearing and vision multi-sensory impairment other senses sensory integration vestibular & proprioceptive
Social relationships and emotional development	Social relationships and emotional development
Support for Learning Sub-groups: assessment & planning educational placement ICT professional support resources	Support for Learning Sub-groups: assessment & planning physical environment educational placement resources (include ICT) professional support
Understanding of space and time	Understanding of space and time
Other Sub-group: ASD	Other Sub-group: ASD/OCD/ADHD

It was also intended to support the development of the main categories and sub-groups, discarding or refining any that were weaker and including any additional ones needed, and to 'firm up' the category boundaries.

Appendix 2 Table (i), records the levels of agreement/disagreement between the assignment of statements to different coding categories found during this initial check of reliability and validity. Statements could be assigned more than one coding and it was interesting to note that where there was an initial disagreement, there was subsequently a higher level of agreement as to a second assignment of a coding. When checking for reliability the level of disagreement was found to be higher in the areas of conceptual development; ownership of learning and the sub-group: individual learning style; sensory responses sub-group: MSI; and understanding of space and time.

Overall a level of 72% of total agreement was obtained. The areas of disagreement provided some enlightenment about the need to adjust some coding categories and ensure clarification through clear use of language to ensure two different people would code in the same way. This revision of the codings was informed by careful examination of the areas where higher levels of disagreement were found and discussions between the researcher and the colleague involved. For example, under the main category of 'Orientation, Movement and Mobility' a sub-group was redefined as 'fine motor', with statements about gross motor being included within the main category. A sub-category of 'physical environment' was also added in the main category of 'Support for Learning'. Clearer descriptors were also developed of what

was to be included in each category as this was also felt to be a cause of some disagreement. The revised codings are also listed in Table 8 above.

A further check for reliability and validity was then undertaken with a different colleague, using the same process again and the statements from the 9 reports used previously together with the revised codings and descriptors. The same set of statements was employed as any used in this check would need to be discarded for the final coding process and it was important to ensure that as many statements as possible following these revisions. The results of this second check are set out in Appendix 2, Table (ii); on this occasion the level of overall agreement achieved was 96%, where the lowest was 77% for the category 'Social relationships and emotional development'. This check for reliability supported the use of the revised main coding categories (with sub-groups where appropriate) and the set of descriptors established without further revision.

3.6.7d The final codings

Following this all the remaining statements extracted from the reports were then coded within these categories. As explained previously, to undertake this process, all the statements were interpreted using an element of literal, interpretative and reflexive thought to assign each statement to one or more codings depending on the content of each statement. During this process more standardised phrases were developed to encapsulate a common meaning where the information was phrased differently. For example, the following are statements extracted from reports for two different children:

Ongoing observations to develop a clearer understanding of how xxxx might best be able to use their vision and support the development of a visual profile,

and

It will be important to continue to collate observations of xxxx's use of vision to develop a functional visual profile to inform the planning and presentation of activities.

During the coding process these two statements were both assigned to the coded category of sensory responses within the statement:

The development of a visual and auditory profile will help to inform the planning and presentation of activities.

One of the key elements of the axial coding process that emerged was identifying whether the statements related to internal factors or to external factors. To support this level of coding the following definitions were created:

- Internal: existing, occurring or found within the child or pertaining to the child.
In other words, internal influences
- External: existing, occurring or found outside the child. In other words, external influences

This would provide information to address the two particular sub-questions "What are the factors within the child likely to affect learning?" (internal factors) and "What strategies are effective in supporting the learning of children with CHARGE syndrome?" (External factors).

Consideration was also given as to how the statements related to the content of the domains outlined within the curriculum document used to support the structure of the codings (Murdoch et al., 2009), and also with other identified literature on deafblindness/MSI. The intention of this was to identify what aspects may be more

specific to CHARGE and distinct from the general deafblind/MSI population, and what aspects may be considered as being common to both populations. This would support the researcher in addressing the research sub-question “What aspects of CHARGE syndrome might be distinct from the more general deafblind/MSI population?”

The coding category of ‘Support for Learning’ was established to support the identification of statements that would provide qualitative information to help address the remaining sub-question of “What specialist support is provided from other professionals and through assessments, to teachers working with a child with CHARGE syndrome?”

3.7 Cycle 2: Questionnaire

For the second phase of this research a survey in the form of a questionnaire (sent out to teachers supporting a child with CHARGE in an educational setting in England) was employed to collect data to examine further and build upon the particular features identified in Cycle 1 and through the literature review. Within this Cycle a semi-structured interview was also undertaken (see 3.3.2b).

3.7.1 Questionnaire design

The questionnaire would need to be designed to help answer the research questions and achieve the goal of this investigation (Robson, 2011; Thomas, 2013). It would also need to gain the cooperation of the respondents and elicit accurate and valid information. The task would therefore be to design a questionnaire that linked the

questions in the survey to the research questions and ensure they were presented in a clear and unambiguous way. The wording would be very important since if the questions were too difficult or unclear it could potentially lead to poor quality answers and a low response rate. As has been outlined previously, it was intended the questionnaire would provide both qualitative and quantitative data which Robson (2011) felt was more likely to facilitate a more 'accurate' answer to the research questions.

3.7.2 Unit of analysis

For the purpose of the questionnaire the individual child was identified as the unit of analysis, since it was information in relation to each child that was sought. Consequently it was explained to potential respondents that each questionnaire was to be completed in relation to one child only. Through obtaining the child's initials and date of birth it was possible for a check to be made to ensure more than one questionnaire had not been completed for a single child. The risk of this was further reduced since, after the researcher had identified where a child with CHARGE was attending school, a questionnaire was then sent out for that particular child with a covering note that again explained only one questionnaire was to be completed per child.

3.7.3 Content and structure

Following the emergent flexible design approach, the findings and initial insights and theory arising from Cycle 1 were used to inform the development of this second

Cycle of the investigation. After careful reflection it was decided that information needed to be generated with regard to:

- The internal factors in relation to the child. These would include sensory impairments and reference to other anomalies that form part of this condition and might have an impact on the child's learning, but would also consider different skills and ascertain where particular strengths and difficulties might lie
- The external factors surrounding the child. It was felt important to also gather evidence from teachers as to what they considered were helpful strategies to employ when supporting a child with CHARGE in the educational environment, whether they considered the implementation of particular assessments to be helpful, and what additional professional support was made available to them

The questionnaire was split into different sections to support the data collection and analysis in a manner that could be linked to the research questions. These sections were:

- A front sheet that reminded the respondent their completion of the questionnaire would be regarded as their giving consent and having an understanding of the purpose and focus of the research investigation. A check was included to ensure the child for whom the questionnaire was completed had a confirmed medical diagnosis of CHARGE, and the child's initials and date of birth requested to ensure against duplication of returns
- Background details (including presence of anomalies related to CHARGE, type of educational placement, etc.) - to obtain demographic information

- The child - this would focus on the internal factors and was divided up into categories that matched the domains set out in 'A Curriculum for multi-sensory-impaired children' (Murdoch et al., 2009). These were:
 - Sensory
 - Communication
 - Social and emotional
 - Conceptual ability
 - Response to routine and structure, and understanding of time and space
 - Ownership of learning
 - Orientation and mobility and motor skills
- A short section on assessment to ascertain what assessments were undertaken and whether educators had found them helpful
- Strategies- this would focus on the strategies that may be effective in supporting the child with CHARGE in the educational setting. These were set out within the same categories used to gather data on the child, so that clear comparison between the child and strategies could be made
- A short section in relation to professional support provided for the respondent in relation to the child with CHARGE they were teaching

3.7.4 Question format

Oppenheim (2000) is of the opinion that data collected through a survey questionnaire can be affected by the characteristics of each respondent (e.g. their knowledge base, and understanding of terminology and the language used). It was

recognised it would therefore be important to ensure the questions used within the questionnaire were clear and likely to mean the same thing to different respondents. This was of further importance since the researcher would not be available to the respondent to provide clarification.

It was decided use would be made of trichotomous questions that required a simple 'yes/no/don't know' response to gather demographic information, although this would be supplemented by some direct questions to obtain the additional information (e.g. type of school the child was attending). Further limited use of such questions was made where it was considered a simple 'yes/no/don't know' response would provide the information needed (e.g. 'Does the child demonstrate poor pencil skills/hand-writing skills?'). Although this closed question form limits the respondents' answers in a questionnaire they are useful to generate data that can be easily analysed, including statistical analysis of frequencies of response (Cohen et al., 2003). Additionally, limited use was made of some open question forms to generate more qualitative information, for example, 'What do you consider to be the main factors within the child that affect his/her communication?' The use of open-ended questions can lead to problems with data handling due to the nature of the variability in the responses given but they also encourage a greater depth of response and contain pieces of information that may not be caught through the use of more closed question forms (Robson 2011).

Within the section focusing on the child, use was made of a summated rating scale (a Likert Scale) that would allow for some measurement of the degree and intensity of a

response whilst still generating numbers. It enabled some movement away from closed questions and required the respondent to give a clear conviction in their response. Originally developed in the 1930s by Likert, this is a widely used scale and one it was felt respondents would be familiar with. As importantly, it is one Robson (2011) considers people enjoy completing. For the purposes of this questionnaire it was decided to phrase the questions by asking how easy/difficult a child was perceived to find an identified skill (e.g. 'how difficult is it for the child to organise him/herself?'). Respondents were asked to circle the number from 1-5 that most applied where 1 was 'very easy' and 5 was 'very difficult; these numbers would then be used to directly score the responses, so that a low scale score would mean a skill was regarded as comparatively easy, and conversely a high scale score would mean a skill was considered more difficult. Oppenheim (2000) reports respondents often avoid scoring the two extreme scores and this restricts the choice available to them. This led to the decision to provide this range for scoring (1-5) so a wide element of variation was available (in the eventuality respondents showed themselves very willing to use the full range of scorings made available).

Use was also made of inventories, that is, a list of options respondents were asked to tick if it was felt they applied to the child the questionnaire was being completed for. These were used primarily to ascertain strategies that might be employed and respondents were asked to tick those listed which they considered to be helpful in supporting a child with CHARGE in the educational setting. Whilst it was recognised that this could potentially 'lead' a respondent to give particular answers, Oppenheim (2000) stated this approach often encourages participants to respond more readily

than if they were presented with an open question. This information was supplemented by an additional box that encouraged respondents to add any additional strategies which were considered helpful with regard to the category being focused upon. This would allow for additional information to be obtained that may otherwise be overlooked.

3.7.5 Pre-test of questions

Since the wording of the survey questions would be so crucial, it was necessary to undertake a pre-test (protocol analysis) to ensure the questions were clear and understandable and would elicit the information sought (Robson, 2011). Initially the questions were tested informally on a number of different people from the field of education and other professions. This was undertaken on an individual basis where the researcher listened to the respondents' thoughts when each question was read out. This proved to be a useful tool to help improve the wording since it enabled the researcher to ascertain the meaning of the questions to the respondents and also how they arrived at their response. Throughout this process the question for the researcher was: "Are the questions clear, simple and unambiguous?" As a consequence of the responses obtained, some questions were rephrased and an example presented where it was felt further clarification was needed. The revised questions were then shown to 14 educators for comment (who were not working with a child with CHARGE and would therefore not be involved in the final survey) and also the researcher's supervisors before final revisions were made. This use of protocol analysis proved to be a valuable tool to identify any potential ambiguities or misunderstandings and the questions could then be altered accordingly.

3.7.6 Pilot of the questionnaire

According to Cohen et al. (2003) this type of data collection requires a pre-test (pilot) to support the development of an effective questionnaire. The primary function of this pilot would be to increase reliability and validity and ensure it could be practicably implemented (Oppenheim, 2000). Once the questions had been tested, this pilot was undertaken on a small number of respondents (as advocated by Robson 2011), involving the questionnaire, covering materials developed to be sent out with the final survey, and the data collection and recording processes (all of which will be described below).

Four respondents were asked to complete the pilot of this questionnaire and to mark any aspects that were unclear. They were also asked to record the length of time taken to complete the survey so this information could be given to the respondents of the final version. Following the return of the completed pilot questionnaires with accompanying comments, together with a further discussion with the researcher's supervisors, some further minor amendments were made and it was then considered the final questionnaire was ready for circulation (See Appendix 3).

3.7.7 Data collection

3.7.7a Identification of potential respondents

In Cycle 1 of this investigation the reports analysed were written in relation to a sample of children with CHARGE supported by the organisation the researcher was employed by. For Cycle 2 of this investigation it was important to try and secure data about a wider sample of the CHARGE population. It was hoped this would allow for a

greater degree of generalisation (Cohen et al., 2003). As in Cycle 1, only children with a confirmed medical diagnosis of CHARGE were considered, and the questionnaire would be circulated to teachers involved in supporting these children. The decision was made to include children across the educational key stages from Early Years to Key Stage 4, and attending a range of formal educational settings to widen the pool of potential children with CHARGE whose teachers might be involved in the investigation. In the information booklet accompanying the questionnaire (see Appendix 4) it was suggested that where the questionnaire was received by a sensory support teacher (for visual or hearing impairment, and/or multi-sensory impairment) the questionnaire was completed jointly with the class teacher.

All children who met these criteria and were supported by the employer's organisation were identified. To widen the cohort further and allow for the inclusion of children not known to this organisation, the following groups were also approached to support the identification of potential participants; the CHARGE Family Support Group with a request being placed in their newsletter, the CHARGE closed group on Facebook, MSI teacher networks, and the BECTA internet forum for MSI teachers. By approaching different MSI teacher groupings there was an awareness that they were less likely to know those children with CHARGE who do not have co-occurring visual and hearing impairments (approximately 10% of the who CHARGE population according to Hartshorne et al., 2011), however, these individuals might be more accessible through the CHARGE Family Support group and Facebook group.

All initial information provided was followed up to obtain the name of the educational setting where the child was placed, and establish a contact name where possible so that more direct contact could then be made.

3.7.7b Obtaining informed consent

Once some basic contact information had been obtained an introductory email was sent out explaining about the investigation and asking if the contact would be willing to be involved. Where a willingness to be involved was confirmed, a questionnaire was then sent out, together with the information booklet. This information booklet used the same format of the leaflet from Cycle 1, since feedback obtained through conversation with parents indicated they had found the first booklet easy to read and felt it contained all the information needed. The initial version of this booklet to be sent out with the questionnaire was redrafted after discussion with the researcher's supervisors and two past PhD candidates, and then used as part of the pilot for the questionnaire.

At the beginning of the questionnaire it was clearly stated that completion would be regarded as the teacher giving consent for the content to be used within the research study and that they understood:

- The nature of the information being collected
- The purpose of the research
- That the information provided within the questionnaire would be securely stored and only accessed by the researcher and other authorised persons

3.7.7c Confidentiality and Anonymity

It was important to ensure confidentiality and anonymity as far as was realistically possible, partly as a matter of integrity, but also to help elicit a more truthful and frank response (Robson, 2011). It was also recognised that an inability to assure potential respondents of a reasonable level of anonymity and confidentiality was likely to reduce people's willingness to engage (Cohen et al., 2003). Within the information booklet it was stressed involvement in this research was completely voluntary and every effort would be made to ensure confidentiality and anonymity. A reference coding was allocated to each questionnaire sent out and on its return, an independent person logged the reference number and also made a note of whether the child had a confirmed medical diagnosis of CHARGE and the child's initials and date of birth. This person then removed and stored the front sheet before the questionnaire was passed on to the researcher for collation and analysis to reduce the likelihood of the researcher identifying the respondent when collating the data from each completed questionnaire. Assurance was also given that there would be no use of participants' names when sharing and publishing the information obtained. Similarly, the child's date of birth and initials were requested solely to ensure only one questionnaire was processed per child and these details would not be used to identify the child.

3.7.7d Securing a positive response rate

Non-response is potentially a serious issue for postal self-completion surveys (Robson, 2011) and there were a number of aspects that needed careful consideration.

Design and layout: The appearance of the questionnaire is all-important according to Cohen et al. (2003) and would need to encourage the respondent to engage. The design would need to be clear and look easy to complete, with adequate spacing for the questions and answers. There would also need to be clarity of the instructions and overall wording. Following the pilot, it was also possible to provide an estimation of the time it was likely to take to complete the questionnaire which would be important as the questionnaire itself was quite big, and the researcher was able to stress that many of the responses required simple responses and the ticking of boxes.

Initial contact and mailing: Prior to the initial mail out the researcher made contact with potential respondents via email that enabled the establishment of an important initial contact, but also alerted potential respondents to the imminent arrival of the questionnaire. A copy of the questionnaire was then sent out together with a stamped addressed envelope as it was considered this was more likely to encourage a response. It was also decided to use stamps rather than the envelope being franked to make it less likely that it would be discarded as 'junk' mail. A handwritten covering note (as encouraged by Cohen et al., 2003) was attached to each questionnaire in which the receiver was named personally, provided with a reminder of the reason for the investigation, and an appreciation for their involvement. This note was written on paper containing the logo of the organisation the researcher was employed to add further weight. The advice of Robson (2011) was followed, who suggested questionnaires to be sent to organisations are best posted out on a Monday or

Tuesday. In response to the initial mail out 43 completed questionnaires were returned (a 64% response rate).

Follow up email: According to Edwards and Talbot (1999) one of the most productive ways of securing a more positive response rate is the addition of a follow up letter (or in this case, email) to those people who did not respond to the initial mail out. This email reminded the person of the questionnaire and explained another copy and stamped addressed envelope would be sent out. As a consequence of this action a further 11 questionnaires were returned, increasing the overall response rate to 81%.

3.7.7e Safe and appropriate storage and handling of data

As for Cycle 1 of this investigation, participants were assured data would be stored safely by the researcher. As part of the informed consent process permission was obtained for the data to be shared with the researcher's academic supervisors, and it was also explained that once the study was completed it would be presented to the academic body as a written thesis, and that ultimately the findings might be used for other publications, training days and for presentations of the research itself.

3.7.8 Data collation and analysis

A careful record was kept of what had been sent out and returns received with the researcher supported in this by an independent person who opened all the returned questionnaires, logged the details onto an Excel spreadsheet, and removed the initial front cover before passing the questionnaires on to the researcher. Each questionnaire was scanned so an electronic copy could be kept, and the original

questionnaire then stored safely in a secure lock box. From the initial Excel sheet the researcher was able to look at the unique reference code applied to each questionnaire and identify non-returns so a further copy could be sent out. An initial editing process was undertaken including checking the child had a confirmed medical diagnosis of CHARGE, that only one questionnaire was completed for any given child (by comparing initial letters of names and dates of birth), and that the child met the other criteria for inclusion. As a result of this two questionnaires were excluded; one because the child was too young and one because it was completed for an adult and neither was in an 'educational environment' as identified for the purposes of this investigation.

As explained in 3.5.3, an initial framework for the data collation and analysis was established and designed using Microsoft Excel to record both the quantitative and qualitative information gathered and provide a tool for statistical analysis. The responses received were put into the Excel sheet and then checked for accuracy by an independent person. The qualitative and quantitative data was then analysed using the constant comparative method as described previously.

3.7.9 Reliability check

A check of the consistency of responses given by participants was built into the questionnaire using Spearman's rho or rank correlation coefficient (See Clegg, 1993; Oppenheim, 2000) as a tool to show the measure of association between two sets of responses; in other words, to show the level of consistency in the responses given by a respondent to particular questions. Spearman's rho was chosen because it can be

used with two sets of discrete variables to show how close the association is (Oppenheim, 2000). In exploring correlation the researcher is required to deal with paired scores and for these purposes, the responses to the following questions were paired:

- C2: Child: what communication modes are used to support communication?
- S2: Strategies: To record the modes included in the use of a Total Communication approach

The process undertaken is set out in Appendix 5, with the level of significance found to be 0.94. These results demonstrated a close association between the results obtained for each of these sets of responses demonstrating that, although there was a slight degree of variation, there was a high level of reliability shown in the responses given by respondents for these two questions. In addition to undertaking this statistical analysis, a simple comparison of the responses given by each individual participant within this survey showed:

- 26/52 respondents gave exactly the same responses in each of these two questions
- 16/52 showed one variation in their responses
- 6/52 showed two variations
- 3/52 showed 3 variations
- 1/52 showed 4 variations

It was not possible to calculate the correlation for each individual respondent since this would need to include further 'repetitions' of questions thus making the questionnaire even longer as well as potentially causing respondents to become frustrated if they felt they were being asked to repeat the information they were

providing. These figures do illustrate that although there was some variation in responses, there was a high level of consistency shown in the responses of individual respondents for these particular questions. It is not possible to generalise this level of consistency of response to other questions, but is an indicator that respondents were likely to give consistent answers, assuming the questions were clear and likely to gain the information intended.

3.8 Cycle 2: Interviews

In Cycle 2 use was also made of a semi-structured interview process to obtain the viewpoint of practitioners involved in supporting learners with CHARGE in an educational setting in the USA.

3.8.1 Background

Perkins School for the Blind was founded almost two centuries ago. Historically it is well known within the deafblind 'world' with Laura Bridgman becoming a pupil at the school in 1837 followed by Helen Keller in 1888, and now has a well-established deafblind programme. Perkins School does not promote itself as a leading establishment for working with pupils with CHARGE but, according to the Assistant Educational Director, this is sometimes a misconception amongst parents. It does however have a high profile in the 'CHARGE world', with staff presenting at CHARGE conferences, developing and sharing materials and presenting webcasts. During the time of the researcher's visit, 27 of the 57 pupils on role had a confirmed medical diagnosis of CHARGE.

It was recognised that within the UK it would be unusual to find so many individuals with CHARGE in one educational setting, due in part to CHARGE being a rare condition, but also the move in the UK to educate all children within their local community wherever possible. These interviews would seek to obtain the opinions of some very experienced practitioners working within this provision at Perkins and more widely within the field for many years, enabling them to develop their knowledge and direct experience of supporting a number of learners with CHARGE. It was considered beneficial to obtain an international (and potentially different) perspective on the education of children with CHARGE that may provide further insight and support the development of the findings of this study. It was recognised, however, that this would obtain the opinions of staff working in only one educational setting, and therefore, whilst personal viewpoints would be conveyed these were likely to reflect the school ethos, culture and perspective that would be an influencing factor in the development of these opinions.

3.8.2 The interview questions

A list of initial questions was drawn up to provide a framework for the interviews and ensure each of the interviewees were asked the same questions that would support comparison of their responses during the analysis process, but also provide the interviewees with scope to expand upon their answers. The questions set were intended to gather information to provide further insights and also data that could be compared with the responses obtained within the questionnaire.

The interview questions asked of each interviewee were:

1a Do you feel that individuals with CHARGE are different to the broader deafblind population?

1b If yes, in what way?

2 What teaching strategies do you find most helpful? (Or unhelpful?)

3 Do you consider their sensory impairments to be their primary need?

4 What, if any, input/support have you had in relation to CHARGE?

3.8.3 Data collection

3.8.3a Identification of participants

Prior to the visit to the school, information about this research study was shared with the management team of the deafblind programme and their consent obtained to undertake these interviews. It was agreed all practitioners working in the deafblind programme could be approached to be interviewed, and an initial meeting was held to introduce the researcher to the staff to make them aware they were likely to be approached and encourage them to be involved. During the course of the researcher's visit 11 members of the team within the deafblind programme were interviewed. Of these, 9 were teachers, 1 was a speech and language pathologist (therapist), and 1 a developmental specialist (behaviour). All were actively involved in supporting students with CHARGE.

3.8.3b Gaining informed consent, anonymity and confidentiality

A summary of information was provided to each interviewee that outlined the nature of the study and the interview process (See Appendix 6). It also addressed the issue of confidentiality and anonymity, and explained all involvement was purely voluntary.

Storage and handling of the data was also addressed. Each interviewee was then requested to sign a consent form expressing their willingness to participate and for the information to be used for the purpose of this research study.

3.8.3c The interview process

As some of the potential interviewees were quite nervous it was agreed that interviews could be undertaken in pairs to increase their confidence, whilst recognising this may influence some of the responses given.

3.8.3d Recording the interviews

It was intended that an audio record of the interviews would be made to support the researcher's notes and provide a reliability check of the record of responses.

Unfortunately the audio equipment failed so a colleague agreed to also scribe responses which could then be compared with the researcher's own notes. Any discrepancies were later checked with the colleague to see if an agreed understanding of what was said could be reached.

3.8.4 Data collation and analysis

The two sets of notes taken by the researcher and colleague during the interviews were compared to check for any potential discrepancies and lack of clarity and responses then recorded on an Excel sheet. The data generated was mainly qualitative in nature, but it was possible to generate some basic quantitative data (e.g. the number of interviewees to express a particular opinion) that could easily be extracted from the Excel sheet. The responses obtained were then coded as during

the process in Cycle 1, continuing to use the constant comparative method (see 3.3.3b) to identify any groupings in the responses given, and compare them with the findings arising from the questionnaire.

3.9 Establishing trustworthiness

Robson (2011) identifies the need to establish a level of trustworthiness if the findings of any research are to be worthy of consideration. If an investigation can be shown to be valid and reliable it follows that it can also be considered trustworthy. Validity and reliability may be considered a main concern for the researcher; if research is not reliable it will not be valid (Cohen et al., 2003), and if it becomes invalid it is worthless (Petre and Rugg, 2012). Therefore the degree of validity and reliability obtained will determine the impact of the findings. Robson (2011) identifies different forms of validity that would be addressed with regard to this investigation, including content validity, internal validity (identifying potential flaws within the study itself), and external validity (the extent to which these findings can be generalised more widely).

3.9.1 Content validity

Content validity, whereby the research addresses the areas it pertains to cover (Cohen et al., 2003) was sought by the establishment not only of an over-arching research question, but also identifying more specific sub-questions that defined in more detail what was to be examined, and supported the direction of focus of this study.

3.9.2 Seeking appropriate methodology and methods

The methodology used as a guiding principle for this investigation has already been described with an explanation of why it was considered appropriate for this study (see 3.2). Use of a flexible design would support the examination of the particular area identified through an evolving, emergent design and would encourage the use of clear multiple methods to gather the data.

3.9.3 The lone researcher

According to Kolb (2012) researcher bias can be a “major threat to validity” (p.85), and working as a lone researcher it would be vital to recognise and acknowledge those assumptions and preconceptions arising from past experience, current knowledge, world values etc. that could potentially affect how the investigation was undertaken and the data interpreted (Thomas and James, 2006). The potential threat to validity this presented was partly reduced by the researcher being aware and making a conscious effort to maintain an open mind, working reflexively (Mason, 2006) to reflect, examine, and explore the researcher’s own position throughout all the stages of this investigation. Additionally this threat was further reduced by:

- Undertaking a reliability check to ascertain the level of agreement (involving a second person) to ensure a high level of consistency in the interpretation when coding statements extracted from the educational reports
- Using reports written by a number of different authors
- Seeking the opinions of a wide number of teachers working with children with CHARGE in a range of different educational settings

- Comparison of notes scribed by the researcher and a colleague during the interview process

3.9.4 Quality of evidence

Ensuring good quality evidence was sought firstly through the clear identification of the research sub-questions to ensure the appropriate data was generated. Only data relating to children with a confirmed medical diagnosis of CHARGE was included in this investigation.

Reports used for the document analysis were authored by specialist teachers from the field of deafblindness/ multi-sensory impairment with experience of working with children with CHARGE and the interviews undertaken involved participants who were also specialist practitioners with similar experience. The data obtained from these sources would be balanced by data obtained from the questionnaire that was directed at class teachers, the majority of whom were unlikely to have additional specialist qualifications in the field of deafblindness or a wider experience of working with children with CHARGE. (It is acknowledged this might be so for some respondents but this information was not requested as the researcher was concerned it might enable the identification of some respondents). Efforts were also made to secure a good response rate from the questionnaire.

For the questionnaire an initial pilot of the questions was undertaken to ensure there was a shared understanding of the questions. Clear use of language was also considered important with minimal use of jargon. To increase the circulation of the

questionnaire and reduce the risk of bias potential respondents were sought, not only through the children known to the organisation by whom the researcher was employed, but also through the CHARGE Family Support Group, a CHARGE Facebook group, networks of MSI teachers, and also an internet forum for MSI teachers.

3.9.5 Multiple sources of evidence

Different data sources were used in this investigation, these being document analysis and surveys in the form of a questionnaire and interviews. Using multiple methods to gather the data would enhance the validity and the trustworthiness of this investigation through this process of triangulation. Mason (2006) states that use of triangulation (that is, a number of different research methods to explore one set of research questions), enables the researcher to build up a more accurate 'reading' of the phenomenon being studied. This would allow for:

- 'Methodological' triangulation (Mason, 2006) through using different methods to obtain information
- Seeking the opinions of others (in the form of report authors, questionnaire respondents and interviewees)
- 'Data' triangulation (Oppenheim, 2000) through obtaining different types of evidence in relation to a number of children

According to Robson (2011), the generation of both qualitative and quantitative data would also further support triangulation.

3.9.6 Reliability

Reliability is closely related to validity, with the level of reliability indicating how accurately the research can be replicated (Robson, 2011) and that the tools used are consistent (Thomas, 2013). The most obvious step to take would be to ensure clear detailed descriptions of the processes undertaken.

As noted already, an inter-rater reliability check to ascertain the level of agreement was undertaken when coding the statements extracted from the educational reports (see 3.6.7c). The initial check to discover the level of agreement in coding the reports revealed an overall level of agreement of 72%. Those areas where the level of disagreement was found to be higher were reviewed after which a further check was then with the overall rate of agreement rising to 96%.

A check for the consistency of responses given by respondents was also built into the questionnaire and this was analysed using Spearman's rank correlation coefficient (see 3.7.9) and revealed a level of significance of 0.94 for the answers provided to two questions within the questionnaire. Although it is not possible to generalise this to state categorically that the respondents were therefore being truthful and accurate in responses given to all questions, according to Oppenheim (2000) it is possible to assume there is then a relatively stable "true' attitude" (p.147) in how the respondent has approached the questionnaire.

3.9.7 Generalisation

External validity is always likely to be a potential issue when research is undertaken in relation to a low incidence condition such as CHARGE despite the assurances in

literature that there are commonalities to be seen (e.g. Majors 2011a). It was considered this research would provide new insight that according to Thomas (2013) has integrity in its own right, although efforts were also made to generate findings that might tentatively be considered in relation to the wider CHARGE population. It was recognised there is a danger of trying to generalise where there is a lack of evidence and the greater the sample size the more strength would be given to generalising any principle derived from this investigation (Robson, 2011). In Cycle 1, reports were used from a limited number of children with CHARGE and it was possible these might be individuals more severely affected by the impact of this condition, by nature of the fact they had been referred to the organisation. To overcome some of the issues of potential selection bias that might result in the sample not being truly representative of the wider CHARGE population, a wider sample was sought in Cycle 2. This included children with a confirmed medical diagnosis of CHARGE from Early Years through to Key Stage 4 to cover a wide range of ages; and children in a range of educational settings, both of which were intended to increase the likelihood of securing a more representative cohort. It was hoped this broad entry criterion, together with seeking to obtain potential participants more widely through a range of different organisations and groups, would also result in a more representative sample that would reflect:

In important ways the characteristics of the actual population.
(Thomas, 2013, p.136)

It is difficult to ascertain what is a large enough sub-set and how this can be obtained when investigating a low-incidence disability, but it had been noted from the literature review, that research studies gaining a wider sample had all employed a survey, usually in the form of a questionnaire (e.g. Salem-Hartshorne and Jacob, 2005;

Hartshorne et al., 2007). Thus, although it is not possible to categorically state that the sample obtained is a full representation of the entire population, it would be hoped that a wide enough sample was obtained to provide some valuable insights that could begin to be generalised more widely, particularly if then supported by further research.

3.10 Ethical considerations

When undertaking any investigation there is an obligation on the researcher to address the ethical issues surrounding the study. Within this investigation the main ethical considerations were:

- The best interests of the children
- Respect for all involved individuals
- Gaining informed consent
- Confidentiality and anonymity
- Safe and appropriate storage and handling of data
- Costs and benefits

These were addressed following advice in literature, particularly the guidelines provided by the British Educational Research Association (2004), and were subsequently approved both the researcher's supervisors and the ethics committee of the University of Birmingham.

3.10.1 The best interests of the children

Petre and Rugg (2012) state the need to consider the impact any research might have on those being studied. Individuals with CHARGE are often very vulnerable and

the primary concern was to protect the best interests of the children to whom the data obtained related. By not directly involving the children in this investigation some of the potential costs to them would be removed, for example, the disruption of their daily routine, the anxiety caused that literature identifies as being challenging for many of these individuals (e.g. Brown, 2003b; van Dijk and de Kort, 2005; Smith et al., 2010). As a result of the children not being directly involved they were considered to be at low risk of being adversely affected by this research. It was therefore decided that consent would not be sought directly from them but rather from parents as suggested by Cohen et al. (2003). This was in part because the children would not be at significant risk of danger or harm, and it was also not clear what the benefits would be of sharing information about this research project with them (how meaningful it would be; would it cause additional anxiety and worry?). As this is a low incidence group, potentially there is a risk of children being identified and this was addressed at some length to minimise this risk as far as possible (see 3.10.4 below).

Having reviewed the potential costs to the children about whom information was obtained, it was possible to balance this against the potential benefits of this study for children with CHARGE. It has been explained that little evidence-based research has yet been undertaken in relation to the education of learners with CHARGE, and the possible benefits of this research to inform and improve practice could be of substantial potential benefit.

3.10.2 Respect for all involved individuals

Aspects of this research had the potential to cause disruption and intrusion on the time of those involved in responding to the questionnaire and engaging in the interview process. Every effort was made to minimise this potential cost and any disruptions or increased workload being placed upon those involved.

It was important to invest time in developing positive relationships, for example, by having a cup of tea with interviewees and later visiting them in their classrooms, and providing a hand-written covering letter with every questionnaire. Part of the information provided to participants made them aware of their right to withdraw with or without a given reason at any time as advocated by Bell (2010). Participants were also made aware of the potential benefits to them, in that it was intended a summary of the findings (and outlining practical strategies) would be prepared to share with educational practitioners that would be made available via the internet. It was hoped this would inform and support the development of practice amongst those working with a child with CHARGE.

Recognition was also given of the obligation towards the researcher's employer who was covering the tuition fees and facilitating access to certain data and participants. In response to this, the organisation remained informed of the progress of the investigation and the researcher adhered to the organisation's protocol.

3.10.3 Gaining informed consent

According to Petre and Rugg (2012), gaining informed consent entails ensuring that participants are fully and clearly informed of the intention of the investigation, what

demands are likely to be made of them, and what the potential risks and benefits are. As explained previously, in Cycle 1 an information leaflet was sent out to parents in which the researcher introduced herself, and explained the purpose of the investigation and what would happen. The fact that involvement was voluntary with the right to withdraw at any time was clearly stated, and the issues of confidentiality and anonymity addressed. Finally parents were informed of how the information was to be used and reported and then asked to sign an enclosed letter giving consent for access to identified reports. In Cycle 2 a similar information leaflet (Appendix 4) was prepared for teachers to whom the questionnaire was sent out, but on this occasion it was also stated that completion of the questionnaire would be regarded as consent being given. Similarly, an information letter/consent form (Appendix 6) was circulated to interviewees before the interview process was undertaken.

3.10.4 Confidentiality and anonymity

Cohen et al. (2003) are amongst those who recognise it is extremely difficult to maintain complete anonymity and confidentiality when engaging in research in the social sciences. This would be compounded in this investigation by the low incidence of this condition and the possibility of recognition of the children and participants. In light of this it was recognised that full guarantee would not be realistic although every effort would be made to maintain confidentiality and anonymity.

To reduce the risks a number of steps were taken:

- Names and other identifying matter would be removed from the educational reports

- Parents were informed this would be done before any information was shared for the purpose of checking reliability
- There would be no use of children's names, staff or school names when publishing the information obtained

Questionnaires were coded on a front page which was removed and logged by an independent person when the questionnaire was returned. This was to reduce the likelihood of the researcher identifying the respondent when collating the data.

Similarly, the child's date of birth and initials were requested solely to ensure that only one questionnaire was processed per child. These details were also contained on the front sheet of the questionnaire removed by the independent person once they had checked for any duplications.

3.10.5 Safe and appropriate storage and handling of data

Parents and participants were assured that data would be safely stored by the researcher in secure locked boxes and on an encrypted data stick. Permission was obtained (as part of informed consent) for the data to be shared with identified colleagues for the process of undertaking a reliability check and also with the researcher's academic supervisors.

3.10.6 Costs and benefits

Cohen et al. (2003) provide an outline of a costs/benefits rationale, adapted from Frankfort-Nachmias and Nachmias (1992). This encourages the researcher to maintain a balance between the potential costs and benefits to those involved in the study (whether directly or indirectly). The potential costs (risks) to the children

indirectly involved and the participants have been addressed above with a description of the steps taken to minimise the potential impact, likewise the potential benefits to the child, parents and participants have also been raised. This investigation was undertaken to reduce the 'problem-space' described by Petre and Rugg (2012) and provide research-based evidence that would hopefully increase the knowledge base in relation to the education of individuals with CHARGE that would better inform practice. It has been stated the intention was also to provide a summary of the findings (and outlining practical strategies) that could be shared with teachers (and other involved practitioners), being made available via the internet.

3.11 Summary and conclusions

In this chapter the methodology and choice of methods was explained, together with reasons for these choices. It was recognised the investigation would be exploratory and descriptive with an emphasis on discovery, adopting an interpretive approach with the intention that final insights/theory would emerge from the research itself.

To generate the evidence needed to help address the main research question and sub-questions it was decided that within the case study framework a flexible approach would be adopted that emerged and developed as the investigation progressed, employing mixed methods and multiple sources of evidence. The methods used to gather data were document analysis, surveys in the form of a questionnaire and semi-structured interviews, with the data being analysed using the constant comparative method. Use of multiple methods was shown to be a way of providing different forms of triangulation to help build up a more detailed and

accurate reading of the subject under investigation (Mason, 2006). It was intended these sources would generate both qualitative and quantitative data.

The research process was set out diagrammatically in Figure B, a diagram clearly showing the course of action that would be systematically undertaken during the course of this investigation involving two clear cycles. These cycles were then referred to again at regular points as each of the stages outlined was described, analysed and discussed. The lengthy process of establishing the final codings in Cycle 1, including an inter-rater reliability check, was described in detail. These codings were then used to inform the remainder of the investigation (together with the results of the examination of literature), supporting the structure and content of the following questionnaire and the development of questions for the interviews, and was therefore considered to be an important, if somewhat time-consuming, stage of this study.

Establishing the final questionnaire included a pre-test of the questions and a pilot of the questionnaire itself and also involved issues around acquiring informed consent, anonymity and confidentiality, and securing a positive response rate. When addressing these issues the focus was on ensuring that higher quality responses were obtained that would directly address the research questions set. Additionally, it was considered vital to ensure the trustworthiness of this investigation, since if a research study is not deemed to be trustworthy it follows that it will be regarded as worthless. It was therefore important to maintain integrity in the research so that its findings would be considered worthy of consideration. A range of steps were

described that were taken to ensure a high rate of validity and reliability with the intention of reducing the potential flaws within the study. It was recognised that some of the limitations of Cycle 1 would restrict the ability to begin to generalise any findings to the broader population of learners with CHARGE, and this was addressed by broadening the sample in Cycle 2 to gain the perceptions of teachers working with children with CHARGE across different educational key stages, educational settings and countries; the intention of this being to increase the potential for these findings to be generalised more widely amongst the population of individuals with CHARGE.

Finally, a summary of the researcher's response to the important potential ethical issues was also addressed. Children with CHARGE are potentially vulnerable individuals and it is for this reason, together with the need to safeguard the direct participants, that careful consideration was given to securing a balance between potential costs and benefits both to the child whom is indirectly involved, and all the participants.

The processes described above were considered the most appropriate, and likely to facilitate the gathering of evidence needed to address the research questions established. This would support the identification of both internal and external factors, assessment tools and professional support for teachers, and also allow for a comparison of the findings with literature on the broader deafblind population. In the following two chapters a summary of the data obtained in Cycles 1 and 2 will be presented, together with the analysis of that data and discussion that would then be used for a final address of the questions.

CHAPTER 4- CYCLE 1: SUMMARY OF DATA GATHERED, ANALYSIS AND DISCUSSION

4.1 Introduction

The exploration of literature described in chapter 2 led to the establishment of a number of sub-questions to help address the main research question, and also informed the choice of methodology and methods as set out in the previous chapter. The findings of the literature review, combined with the researcher's own experience as a practitioner may have been regarded as adequate to enable the composition of a questionnaire for this investigation, however, it was considered this could potentially weaken this research due to increased risk of researcher bias (see 3.9.3) and a strong reliance on those 'grey materials' which although valuable, also had limitations (see 2.1). Thus, Cycle 1 would be important to increase the rigour of this investigation through obtaining evidence from an additional source that might not only support the points raised through the review of literature, but raise additional elements to pursue through the questionnaire. Although the researcher authored a number of the reports used, other authors were also involved and this would enable the acquisition of the observations and perceptions of other practitioners in addition to those of the author (see 3.6.4). As importantly this cycle would also facilitate the developments of initial insight and theory and inform the development of the questionnaire and interviews to be undertaken within Cycle 2.

Within this cycle there was an examination of 58 educational reports written for 17 children with a confirmed medical diagnosis of CHARGE, with the methods and

processes undertaken described in detail in 3.6. This chapter sets out the analysis of the initial data generated, supported by discussion and the presentation of the initial insights and theory emerging.

4.2 Quantitative information

It has been explained the main focus of this analysis would be on the qualitative data obtained and the content of the statements allocated to each of the coding categories established, supported with quantitative data where appropriate and possible. The amount of quantitative data generated was limited, and although of some benefit, recognition of the potential weaknesses (see 3.6.7a) meant use of this data was restricted. For example, the total number of statements allocated to each category and sub-category, together with the number of children for whom statements were made, were recorded to ascertain whether they might all be regarded as relevant for the child with CHARGE. These findings (set out in Table 9) showed statements extracted were connected to each of the domains from 'A curriculum for multi-sensory impaired children' (Murdoch et al, 2009) and in relation to a number of different children. This demonstrates that despite the heterogeneous nature of CHARGE, authors of these reports highlighted the same issues for a number of learners with this condition thus providing an initial indicator there may be common themes as suggested in literature (e.g. Majors 2011a). A comparison of the number of statements allocated to each category was not undertaken since some (such as 'Fine motor') were more specific than other broader categories/sub-categories (e.g. 'Communication') that would not allow for a valid comparison to be made.

TABLE 9: CYCLE 1- TOTAL NUMBER OF DIFFERENT STATEMENTS PER CATEGORY AND NUMBER OF CHILDREN FOR WHOM THESE STATEMENTS WERE MADE

Coding categories	Total number of different statements in category	Number of Children for whom statements made in each category (N= 17)
CHARGE	8	13
Sensory responses	8	5
General		
Sub-groups:		
Hearing and vision	16	16
MSI	9	12
Other senses	5	5
Sensory integration	15	12
Vestibular/ proprioceptive	9	10
Communication	40	14
Conceptual development	23	13
Orientation, Movement and Mobility		
General	30	16
Sub-groups:		
Fine motor	6	10
Physical positioning	9	15
Ownership of learning		
General	15	13
Sub-group:		
Individual learning style	25	16
Responses to routines and changes	18	11
Social relationships & emotional development	26	15
Understanding of space and time	13	11
Support for Learning		
Sub-groups:		
Assessment & planning	30	16
Physical environment	9	16
Educational placement	14	6
Resources (include ICT)	27	10
Professional support	39	16
Other	5	3

The number of children for whom particular statements were identified was noted and whether these statements had been presented by different authors. This made it possible to identify those statements raised most frequently and widely although this was interpreted with a degree of caution for the reasons outlined in 3.6.7a. Overall, it was regarded as important to view all the statements made equally rather than focusing solely on those appearing in the reports with greater frequency; those statements raised more frequently may indicate what is more commonly regarded as important, but 'exceptional' points raised only once or twice in reports (originating from different authors), were also viewed as equally important as they may provide new insights, and raise issues not previously widely noted or considered.

4.3 Findings: The coding categories

Completion of the coding process showed a number of statements were assigned to each and every category and sub-category indicating the coding framework established following the pilot was appropriate. Although some statements extracted arose from reports for only one child (being equally acknowledged as explained above), the majority of statements were made for more than one child and presented by different authors. Where statements were made in relation to a larger number of children it might be considered these reflect opinions more commonly held by the authors, but these may also be issues that more regularly need addressing when supporting a child with CHARGE. In addition to data relating to the provision of specialist support for teachers and the use of assessment processes (to be discussed shortly), during axial coding it became evident the statements within each category could be further divided into 'internal' and external' factors (defined in 3.6.7d), in other

words, statements written in response to factors related to the child him or herself, and statements addressing issues arising from the learning environment (including people) around the child.

Using the results of the review of literature presented in Chapter 2, a comparison would be possible of the data obtained in this Cycle of the investigation to other information presented elsewhere relating to learners with CHARGE. As part of the analysis of this data an additional comparison would be made with a sample of literature relating to the wider deafblind/MSI field. The literature chosen for this comparison was:

- Aitken et al. (2000) 'Teaching children who are deafblind'. A book written by practitioners focusing on the education of children and young people who are deafblind, where the target audience was teachers, residential staff and students.
- Murdoch et al. (2009) 'A curriculum for multi-sensory-impaired children: from MSI Unit Victoria School Birmingham. Although the title describes this as a curriculum, it can be used as an assessment tool and also includes teaching approaches that can be employed in the delivery of the curriculum. This document was also used to support the development of the coding categories used in the initial analysis.
- Sense (2002) 'Quality Standards in Education Support Services for Children and Young People who are Deafblind / Multi-Sensory Impaired'. These standards were developed as guidance to support services working with both schools and families. They could also be used as a tool to evaluate qualitative

and quantitative aspects of the service delivered to children and young people who are deafblind/MSI.

- Nelson et al. (2010) 'Child-guided strategies: the van Dijk approach to assessment. For understanding children and youths with sensory impairments and multiple disabilities'. This is an assessment tool but also provides a guide to intervention.

These materials were chosen as they were considered to contain information relating to different aspects involved in the education of children who are deafblind/MSI, including pedagogy, assessment, curriculum, teaching strategies and good practice.

It was recognised there is an array of other literature available that could perhaps also have been considered, but it was important this comparison remained manageable, and so a sample was selected to represent a clear cross-section and address different aspects of the education of children who are deafblind/MSI. It was recognised the content of the more recent literature chosen (Murdoch et al., 2009 and Nelson et al., 2010) may be influenced by a greater awareness of learners with CHARGE (with contributors gaining experience of working with these children and reading the contributions of others), even though the sources used only refer to the deafblind population in generic terms.

4.3.1 Internal factors

A wide range of internal factors was extracted from the reports analysed involving sensory aspects, communication and social skills, motor skills and mobility, learning

processes and learning style, these being set out in Appendix 7, Table (iii). A comparison with the sample of literature relating to the wider deafblind/MSI population showed many of these internal factors were also identified as being potentially found in other individuals who are deafblind/MSI. For example, Aitken (2000a) identified three themes he regarded as common to many individuals with deafblindness. These themes indicate individuals who are deafblind/MSI will experience difficulty in:

Finding out information; communication with others; moving around the environment. (p.3)

All of these themes were raised within the factors extracted from the reports and show there are connections between individuals with CHARGE and the wider deafblind/MSI population. There were, however, a number of features included in the list of internal factors where no specific reference was found in this sample of literature (although it is accepted some might be evident in other bodies of work) these being presented in Table 10. Those statements extracted solely from reports the researcher was involved in authoring are highlighted in yellow (this will be addressed in 4.4). A number of these factors relate directly to the anomalies that form part of the diagnostic criteria for CHARGE, or the impact of these anomalies. For example, vestibular difficulties arising from the absence or under-development of the semi-circular canal that, together with hypotonia and proprioceptive difficulties, cause the child to experience difficulty in working against gravity and also create a need within the child to develop compensatory strategies. There were also a number not relating to the diagnostic criteria, but related to other anomalies thought to be associated with this condition, such as executive function difficulties.

TABLE 10: THOSE INTERNAL FACTORS FROM THE CODING CATEGORIES NOT IDENTIFIED WITHIN THE SAMPLE OF LITERATURE RELATING TO THE BROAD DEAFBLIND/MSI POPULATION

- True and extreme MSI- all senses likely to be affected
- Unique combination of anomalies in each child
- Sensory integration issues
- Sensory impairments- requiring stronger stimulation
- Vestibular and proprioceptive difficulties
- Difficulty working against gravity
- Reduced sense of smell and taste
- Executive function difficulties
- Conflicting demands- putting skills on hold whilst learning/practising physical skills
- Level of fine motor skill development
- Poor pencil skills/ writing skills
- Use of compensatory strategies, including adopting different positions
- Engaging in physical movement to support concentration
- Impact of changes to routine on the child's sensory system
- Facial palsy- awareness of social implications

(Those highlighted in yellow were extracted solely from reports researcher contributed to)

4.3.2 External factors

A large number of external factors were also identified most being a response to the internal factors raised and supporting the creation of a positive educational environment, these being set out in Appendix 7, Table (iv). It was found many of these external factors were also referenced in the sample of literature used for comparison, suggesting that the deafblind educational philosophy and approach (as defined in 1.6.2) could also be applied to learners with CHARGE. As with the internal factors, there were a number of external factors where no specific reference was found in this sample of literature, these being presented in Table 11.

TABLE 11: THOSE EXTERNAL FACTORS FROM THE CODING CATEGORIES NOT IDENTIFIED WITHIN THE SAMPLE OF LITERATURE RELATING TO THE BROAD DEAFBLIND/MSI POPULATION

- Raising awareness and knowledge of CHARGE, possible implications and the impact on learning/development and everyday living
- Training on the nature and implications of CHARGE
- Input from an MSI teacher with knowledge/experience of CHARGE
- Recognition of vestibular and proprioceptive difficulties and impact on learning
- Activities planned/implemented in response to vestibular and proprioceptive issues
- Opportunities to adopt a horizontal position
- Support to overcome issues relating to facial palsy
- Not placing too many demands on child when a 'difficult' day
- Recognition of/strategies to manage impact of fatigue
- Development of a sensory integration programme, including a sensory diet
- Access to a specialist SI OT/ Physio
- Appropriate behaviour management with recognition of impact of SI difficulties
- Ensuring against sensory 'overload' and developing appropriate management strategies
- Providing small 'fiddles' for use when sitting still
- An appropriate environment to reduce sensory overload, conflicting demands etc.
- Access to a safe rest area
- Recognition of executive function difficulties
- Use of a concrete component to overcome executive function difficulties
- Activities to support fine motor skill development and coordination
- Assessment of hand function
- Provision of activities to support development of pencil/writing skills
- Provision of alternative recording methods

(Those highlighted in yellow were extracted solely from reports researcher contributed to)

Some of these external factors also relate to specific anomalies that form part of the condition of CHARGE, for example, strategies to respond to the child's vestibular difficulties and support to respond to issues resulting from the child's facial palsy.

Other external factors identified in the reports analysed but not referenced in the sample of literature can also be considered a response to some issues specifically raised in relation to CHARGE that do not form part of the diagnostic criteria. For example, a response to executive function difficulties likely to relate to the neurological anomalies linked to CHARGE (Gilles, 2011), and a response to the poor fine motor skills likely to arise from the difficulty with developing gross motor skills as a consequence of impaired visual and vestibular senses (e.g. Lieberman et al., 2012) and low tone (e.g. Hartshorne et al., 2007). It was interesting to note there appeared to be some overlap, but also some differences, between what was felt to be required when supporting a deafblind/MSI child generally, and when supporting a child with CHARGE. For example, reference was found to sensory integration difficulties in the sample of literature used for comparison, where Murdoch et al. (2009) reported the need to be aware the deafblind child may experience difficulty with:

The processing and integration of information from residual hearing, vision and other senses. (p.11)

No reference was, however, then found in the literature sample that compared to the opinions of the report authors who advocated for a response to this through the development of a sensory integration programme.

There was also no obvious recognition these difficulties needed to be considered in the development of a behaviour management programme (i.e. that the sensory integration difficulties could be causing a child to act in a certain way and therefore needed addressing in relation to the child's behaviour). It is possible sensory integration difficulties have a greater impact upon the child with CHARGE since, not only is the child likely to have a visual impairment and a hearing impairment but all

the other senses can potentially be affected. Perhaps then, what should be considered is whether it is the degree of impact that differs, which in turn will influence how this is responded to through the development of specific planned intervention.

4.3.3 The provision of specialist support to teachers and the use of assessment processes.

Within the statements extracted from the reports, a frequently raised opinion was that teachers and other staff members supporting a child with CHARGE require specialist training to develop their knowledge and understanding of deafblindness/MSI but also to develop an understanding of the nature and implications of CHARGE. This was found in statements written independently by 4 different authors, and also in reports of which the researcher was a co-author, and is a view also supported within the Sense (2002) document where the need for ongoing support and training in 'specialist aspects' (p.14) is clearly identified.

Input from specialist teachers for the visually impaired and hearing impaired, and also from a specialist teacher for the multi-sensory impaired was advocated, again by different authors, as was input from a range of therapists (physiotherapist, an occupational therapist and a speech and language therapist). The need for other specialist input from a paediatric habilitation officer and educational psychologist was also highlighted. Involvement from these specialists was identified with regard to the planning and delivery of an educational programme, communication, sensory integration issues, motor development, positioning and seating, orientation and

mobility, and eating and drinking. In light of the number of different professionals who may become involved, 4 authors expressed the need for a multi-agency approach with close collaborative working.

Within the reports 7 of the 8 authors recommended that functional assessments be undertaken to consider the impact of different factors on the child's overall functioning. The need for an assessment of functional use of vision and hearing was specifically identified for a 10/17 children, together with the development of a visual and auditory profile to inform planning and the implementation of activities. Two report authors (1 of whom was the researcher) independently also considered it necessary to undertake assessment to help recognise a child's learning potential, although no particular formal assessment tools were identified. Additionally the view was expressed by different authors that environmental audits were required in relation to communication, sensory needs, social development, learning and orientation and mobility skills. The purpose of an environmental audit is to identify which aspects of the environment are supporting or adversely affecting the child's ability to learn and respond within that given environment and ultimately this can lead to making appropriate adaptations in response to the findings of the audit. Although environmental audits were not specifically identified within the literature examined (either in relation to CHARGE or deafblindness/MSI), the importance of managing the environment for all these children to provide an optimal learning situation is clearly recognised (e.g. Murdoch et al., 2009; Hodges, 2002; Brown, 2011; Majors, 2011a).

4.4 Discussion

The document analysis undertaken within Cycle 1 generated a wealth of data both qualitative and quantitative in nature, although the emphasis was on the qualitative data obtained the findings were supported and enriched by quantitative information where appropriate. It was recognised the statements extracted and analysed reflect the opinions and perceptions of the authors of the reports, and the individual characteristics of each author could potentially influence the data obtained. For example, since the authors of these reports were all employed by an organisation supporting children who are deafblind/MSI it is perhaps not surprising the areas of vision and hearing were addressed at some length for all but one child. It also, however, may relate to that fact that over 90% of individuals with CHARGE are reported in literature to have co-occurring visual and hearing impairments (Hartshorne et al., 2011), and also reflect a comment by Smith et al. (2010) who considered the visual and auditory needs of children with CHARGE are often addressed in literature. From some of the statements analysed there was some difficulty ascertaining whether the content of the statement was an expression of opinion in relation to the specific child or simply the sharing of information found in literature. For example, it was stated that high levels of stress are inherent in CHARGE and often the trigger for certain behaviours; this reflecting literature (e.g. Brown, 2003b; Graham et al., 2005) and providing an illustration of how the author's knowledge of existing literature of CHARGE may have been an influencing factor. A pragmatic view was taken of this, accepting that where this was included in a report the information would have been considered pertinent for that particular child.

A comparison of the data generated with literature on CHARGE previously examined in Chapter 2, identified internal and external factors reflected in the anecdotal reporting and opinion expressed by other practitioners supporting learners with CHARGE. This illustrates how such materials may influence the thoughts and actions of other practitioners, highlighting the need to expand and strengthen the acquisition of knowledge grounded and evidenced in research. Additionally and importantly, since the reports used were written in the period between 2002-2008, many of the statements extracted from these reports have also been highlighted in articles, webcasts etc. presented subsequently to this period. This reinforces and supports the perceptions of the authors of these reports, and suggests the reports used for this document analysis were a valid source of evidence. For example, the need to consider the impact of a child's sensory integration issues in the management of a child's behaviour would later be raised by Hartshorne (2011); the use of a concrete component to support recall, initiation, choice making and concept development would be confirmed by Stelzer (2011) and Brown (2011a); the need to pace activities and provide additional time for the child to process information would also be advocated by Majors (2011a) and Brown (2011a).

As explained in 3.6.4 it was acknowledged the researcher of this investigation was involved in authoring or co-authoring a large number of the reports used for this document analysis and, therefore, those statements emanating solely from reports the researcher wrote or co-wrote were identified. A closer examination showed most of these statements were also to be found in the sample of literature on the general deafblind population used for comparison (e.g. a need for recognition of the fatigue

resulting from the daily challenges created by MSI- raised in different chapters in the book edited by Aitken et al., 2000), indicating these opinions did not emanate solely from the researcher. There were additionally a small number of these statements that were not referenced in this sample of literature or literature on CHARGE, relating to both internal and external factors. These are highlighted in Tables 10 and 11 and would be explored further in Cycle 2 of this investigation to ascertain the views of other educators with regard to these points.

It was noted some of these statements also related directly to literature available at the time the reports were written and were possibly a reflection of the researcher's awareness of these articles (e.g. Brown, 2003a, reporting on the potential impairment of all the senses in a child with CHARGE resulting in 'true' multi-sensory impairment). Although other authors of these reports did not directly refer to this, they did provide other statements that were closely related (e.g. the importance of recognising challenges for the child created by their MSI). Similarly, in one report the researcher expressed the need for contingency plans to manage fatigue; whilst other authors did not identify this as a planning requirement, they did refer to fatigue as an issue for children for whom the reports were written.

A few statements although only expressed in reports authored by the researcher, can be regarded as quite general, and it might be assumed they could easily have been generated by any of the other authors (e.g. the need for an IT assessment to identify the equipment needed). With regard to the remaining statements, they were found to relate either directly to sensory integration difficulties (discussed in 2.2.8) or

executive function difficulties (discussed in 2.2.9). All authors of the reports used addressed the matter of sensory integration difficulties to some extent, but these additional statements identified other ways this might potentially impact on the child (e.g. putting skills 'on hold' whilst learning new skills) and specific strategies that may need to be implemented (e.g. not placing too many demands on the child when it is a 'difficult' day). The researcher also directly referred to the potential impact of executive function difficulties within statements assigned to different categories (e.g. as potentially affecting a child's communication, ownership of learning and learning style). It was noted that whilst other authors very rarely made direct reference to executive function difficulties, they did identify issues which may arise from executive dysfunction such as poor self-organisational skills and self-help skills, and the need for use of a concrete component to support learning (as identified in 2.2.9).

4.5 Addressing the research questions

This cycle of the investigation provided some initial findings to begin to address the research questions set out in 2.7. The process of open and axial coding identified a large number of internal aspects considered to be factors within the child likely to affect learning, whilst the external factors include strategies that may be advocated in supporting the learning of children with CHARGE. Likewise recommendations were made for input from a number of different professionals including specialist teachers to support the class teacher. These would all be examined further in Cycle 2 of this study, as will be explained shortly.

The comparison with the sample of literature used relating to the wider deafblind population showed many of the statements extracted from the reports were also referenced in this literature, suggesting there are similarities to be found between both the CHARGE and broader deafblind populations, and that the deafblind educational philosophy can also be applied to learners with CHARGE. However, this comparison also showed a number of statements that were not found in the sample of literature used for comparison (set out in Tables 10 and 11); it was recognised these statements may be raised in wider literature, but this also provides an initial indicator of what may be distinct about children with CHARGE and the support for learning required.

Other initial indicators that CHARGE may be distinct from the broader deafblind population were noted during the course of Cycle 1. As suggested by Strauss and Corbin (2007), an external tool was used to help structure the codings to be used in the document analysis, this being a curriculum designed for children who are MSI (Murdoch et al., 2009). During the coding process when the categories were developed and later finalised some additional categories and sub-categories were created to ensure greater clarity for the coding process and include aspects not fully addressed within the domains of the curriculum itself. This was partly due to broader issues than the curriculum being addressed within the reports (such as training for staff etc.), but may also suggest the educational needs of a child with CHARGE might not be met solely within a programme employing well-established strategies to support learners from the general deafblind/MSI population. Secondly, excluding those statements only expressed in reports either solely or jointly authored by the

researcher, a number of statements regarding CHARGE were raised. Authors were of the opinion there was a need to develop knowledge and understanding of the impact and implications of CHARGE and multi-sensory impairment, that the MSI teacher required additional knowledge and experience of CHARGE, that the class teacher would need to have an awareness of the challenges raised by the child's multi-sensory impairment but also in relation to the child's diagnosis of CHARGE, and that training need to be provided in relation to multi-sensory impairment and CHARGE. These statements imply it was considered there is something distinct or additional that requires the teachers (and other staff working with the child) to develop knowledge and understanding of multi-sensory impairment, but also additionally of CHARGE.

4.6 Initial insights and theory

As explained in Fig. B in 3.4, the data gathered and analysed in Cycle 1 of this investigation would inform the development of the surveys to be undertaken within Cycle 2 that would test and explore these findings and initial theory/insights generated. At this point in the investigation there is not enough evidence to yet be clear whether, if a child has a medical diagnosis of CHARGE, it is possible to make definite assumptions about the philosophy and strategies to employ in supporting their learning and development. This cycle of the investigation has, however, provided some data to begin to address the sub-questions and formation of initial theory and insights. These are important and significant in beginning to form a more accurate and deeper understanding of the phenomenon being studied that will ultimately help progress towards finding a possible answer to the main question

underpinning this research. Here then are the initial insights and theory arising from the first cycle of this investigation.

That there are common features to be found in children with CHARGE and other deafblind/MSI learners, but that there are also additional factors, all of which might have the potential to impact upon their learning and engagement in the educational environment. It may not only be sensory aspects, but also the presence and impact of other anomalies, which can affect learning and development.

That potentially all the sensory modalities can be affected, together with many other anomalies, making the degree of impact more severe and having an exponential effect. For example, the three common 'themes' identified by Aitken (2000a, p.3) highlight the issues he considers to be experienced by the majority, if not all, individuals who are deafblind/MSI as a result of their co-occurring visual and hearing impairments. If we consider each of these in turn, using examples from the data generated thus far, it is clear these may be further impacted by the presence of any additional executive function difficulties. So:

- Difficulty 'finding out information'- will be made more demanding by the difficulties experienced in initiating activities, remaining on task and completing a task independently, attending to what is relevant and ignoring distractions, responding appropriately in unfamiliar and novel situations, forming clear concepts, especially when they become more abstract, etc.

- Difficulty 'communicating with others'- will become further complicated by the difficulty in initiating social interactions, monitoring and regulating their own behaviour, attending to what is relevant and ignoring distractions, and so forth
- Difficulty 'moving around the environment'- becomes more testing as a consequence of difficulty monitoring and regulating their self-organisational skills; in responding appropriately in unfamiliar and novel situations; and in predicting what might happen next when it is outside their familiar routines and direct experience

That the educational philosophy and pedagogy underpinning the education of deafblind/MSI learners can be applied to children with CHARGE, but that additional strategies and a different emphasis may be required. This may include a greater emphasis on the impact of regulation and sensory integration difficulties, vestibular and proprioceptive difficulties, neurological anomalies and executive function issues, and fine motor skills.

4.7 Developing the next cycle of the research

In addition to the formation of the initial insights and theory, the final part of Cycle 1 of this investigation was also to inform the development of the questionnaire and interviews that would provide the tools for generating data in Cycle 2 to provide further data and analysis to address the over-arching research question and sub-questions of this study. This was necessary to build upon the evidence gathered thus far and test the initial insights and theory generated from Cycle 1, and to overcome the limitations of the reports used in Cycle 1 (see 3.6.5), seeking to obtain through

different sources further data from a broader representation of practitioners engaged in supporting learners with CHARGE.

The findings in Cycle 1 arose from the analysis of reports for 17 children that reflected the opinions and perspectives of the authors of the reports (including the researcher), as much as providing information in relation to the child's learning and educational placement. It was not always possible to identify whether statements were the authors' opinion in relation to the specific child or simply the sharing of information found in literature. It was intended that through Cycle 2 of this study, data would be gathered through the use of both a questionnaire and interviews to gather more evidence of the internal and external factors involved in the learning and development, and an effective educational approach for children with CHARGE. Although these survey forms would also rely on teacher perceptions, the material would be gathered from a wider range of educationalists directly involved in supporting a child with CHARGE in the educational environment.

It was decided the questionnaire would be comprised of two parts; building on the internal/external factor structure which emerged from the open and axial coding undertaken in Cycle 1. It had also been found that the same statements were often assigned to both the categories of 'Understanding of Space and Time' and 'Responses to Routines and Changes' and these two categories would be combined for the purposes of the questionnaire. The internal factors identified through Cycle 1 were likely to have been offered by authors in response to information being made available to them, conversations with the family and professionals, and their direct

observations of the child. The first part of the questionnaire would focus on the child and seek information to identify the presence of anomalies associated with this condition in the child, and to build on the material already obtained to gather perceptions of the impact of these internal factors, together with a focus on identifying possible strengths and needs, according to those skills perceived to be easier or more difficult for the learner with CHARGE. The external factors identified from the reports analysed in Cycle 1 included recommendations for practice but there was no evidence as to whether these were found to be beneficial by the practitioners to whom they were offered. Thus, the questionnaire would seek to gather opinions from teachers as to what strategies they felt were most helpful or effective in supporting the learning and development of children with CHARGE. Overall the data generated within Cycle 2 would be important to build up evidence to address the over-arching research question and sub-questions more fully. It would provide the opportunity to investigate further what these practitioners considered were the factors present within the child, the strategies considered to be helpful, to learn more about types of assessment undertaken and their usefulness, and to explore further the support provided to the teachers in regard to CHARGE.

The findings of Cycle 1 also suggested there are other issues in addition to the child's sensory impairments that may impact upon the child's learning and development. It was therefore considered beneficial to ascertain whether teachers considered the child's sensory needs constituted their primary need and this question would be asked in both the questionnaire and interview. Schools are required to complete the The School Census, previously known as the Pupil Level Annual

School Census (PLASC), on an annual basis where they are required to identify “the primary and secondary need of the pupil” (Department for Education, 2015, p.47). The information on The School Census states the data generated will support planning and policy development which implies this includes planning and choice of educational provision and support provided for a learner with CHARGE. These needs should also be reflected in the content of the Statement of Special Educational Needs or Education, Health and Care plan that replaces the Statement of Educational Needs with the implementation of the Children and Families Act (2014).

The interviews of practitioners working with pupils with CHARGE in an educational provision in the USA would additionally seek to gather further opinion of whether they consider individuals with CHARGE to differ from the broader deafblind population; what teaching strategies were considered most helpful and the support given to the practitioners themselves.

4.8 Summary and conclusions

The document analysis undertaken in Cycle 1 generated data that was examined and analysed to begin addressing the research questions. It was important to support the formation of initial insights and theory and to inform the development of the second cycle of this investigation. A sample of literature relating to the learning and education of the wider deafblind/MSI population was used as a means of comparison to examine the similarities and differences to the education of learners within the wider deafblind/MSI population.

A large number of internal and external factors were extracted from the reports analysed, involving sensory aspects, communication and social skills, motor skills and mobility, learning processes and learning style. The internal factors identified supported evidence from literature that all the sensory modalities are likely to be affected. Furthermore, in addition to sensory issues, other features were also identified that are likely to impact on learning and development. In light of this it has been suggested the impact of these impairments and other anomalies may be exponential and children with CHARGE therefore likely to be more severely affected than other deafblind/MSI learners. With regard to the external factors identified by authors of the reports, the findings suggested the educational philosophy for educating deafblind/MSI children may be appropriately employed by educators working with children with CHARGE, but there is also possibly a need for additional strategies and a greater or different emphasis on certain aspects.

Throughout this process of analysing the data extracted from the reports there was an awareness of potential researcher influence on the findings as a consequence of the researcher being one of the authors involved in the creation of these reports. It was therefore noted where statements extracted emanated only from reports solely written or co-authored by the researcher. From this it was clear some statements were similarly expressed by other authors of the reports (e.g. fatigue); many related to sensory integration difficulties, an aspect also highlighted by other authors although not in the same depth; and direct reference to executive function difficulties. Analysis of the statements from other authors noted they had raised some aspects that could potentially be linked to executive dysfunction although these were not

explicitly identified as executive function difficulties. This led to the conclusion that these aspects would need to be explored further within Cycle 2 of this investigation to ascertain whether a wider sample of teachers supporting learners with CHARGE considered these to be factors affecting the learning and development of this group of individuals. These initial findings would be developed and examined further in Cycle 2, seeking the opinions of a larger number of teachers in different educational settings, supporting a learner with CHARGE. In the following chapter the data generated during Cycle 2 will be presented, together with an initial discussion of the findings.

CHAPTER 5- CYCLE 2: QUESTIONNAIRE AND INTERVIEWS. SUMMARY OF ANALYSIS AND INITIAL DISCUSSION

5.1 Introduction

In the previous chapter, an analysis and discussion of the data obtained in Cycle 1 of this investigation was presented, that would inform the development of Cycle 2. This second cycle would explore further the initial insights and theory generated in Cycle 1, employing a questionnaire (see Appendix 3) to gather information on those factors within the child that may impact on learning and development and identify possible strengths and needs. It also addressed external factors, exploring teaching strategies, assessment and support provided to the teacher. Within this cycle interviews were also undertaken with staff at a specialist deafblind provision in the United States of America (see 3.7).

In this chapter the data obtained in Cycle 2 will be presented together with an initial discussion. The findings and discussion generated will then be used to return to, and address, the research questions of this investigation in the final chapter.

5.2 Presentation of data and initial discussion

Details of the construction of the questionnaire and how the data would be collated and analysed are set out in 3.6. The questionnaire generated large amounts of data to be analysed and each section of the questionnaire was examined as follows:

- An initial exploration of the overall pattern for each given question
- A comparison between questions both within a given section (e.g. 'The Child') and across different sections of the questionnaire that may help identify possible relationships (e.g. between the relative ease/difficulty perceived to be experienced for a given skill and the degree of helpfulness of a particular strategy)
- A comparison within questions, including across the different educational key stages and types of educational provision (discussed further below)

Within this survey use was made of closed questions requiring a simple yes/no/don't know answer that would provide data which was easy to analyse and elicit frequency of response information. These were supplemented by limited use of open questions that would provide additional, but what were likely to be more variable, responses. It is recognised these different question forms would generate different types of information and therefore, where the data was obtained through the use of open questions this will be highlighted. A Likert Scale was used in 'The Child' section to provide a measure of the intensity of response with regard to how easy or difficult a given skill was perceived to be for the child in question (1 being very easy and 5 very difficult). Inventories were also used, mainly in the 'Strategies' section); it has been acknowledged there was the risk of 'leading' the respondent (3.6.4) but that this was outweighed by the likelihood of eliciting a greater response than if open questions were used. In the following presentation and discussion it will be assumed the strategies named were included within the given inventories, unless otherwise identified.

As explained the analysis included a comparison of the data obtained for children across the different educational phases of Early Years/KS1, KS2, KS3 and KS4 with the intention of supporting the identification of whether there was any link between responses and the age of the child. Responses were also analysed to examine and compare scores obtained for children in different types of educational provision, these being categorised as SLD/PMLD provision, mainstream and other specialist provision. This may provide an indicator of the potential impact of learning disability, with the view that the most able children were more likely to be placed in mainstream provision and children with the greatest learning difficulties and complexity of need were more likely to attend an SLD/PMLD setting. It was however, acknowledged that with the current inclusive model, the presence of a learning difficulty to some degree could not be excluded in children attending these other settings. Thus it was considered the findings arising from the comparison of these sub-groups would need to be regarded with greater caution. This is further emphasised when recognising this data analysis might also provide a reflection of differing educational ethos within these different categories of provision, but also perceptions and expectations, availability of particular strategies and the knowledge and experience of teachers in the different settings. Additionally it would be important to acknowledge that the comparisons made within questions (i.e. between small sub-groups of different people) must be treated with additional caution because of the very small numbers of participants involved.

This presentation of data and discussion will focus on those points considered to be of primary importance and interest to this investigation. Where the findings of this

investigation closely relate to previously published findings and 'grey literature' this will be noted. The issue of the similarities/differences between the internal and external factors for learners with CHARGE and the wider deafblind population will be addressed in the following chapter when the research questions will be returned to.

5.3 Questionnaire responses

After initial enquiries, following the procedures set out in 3.6, 67 questionnaires were sent out to teachers working with a child with CHARGE in the educational setting with 54 completed questionnaires finally returned giving a response rate of 81%. Of these, 1 child was very young and not yet in a formal educational setting and 1 response was returned in relation to an adult who was beyond Key Stage 4. Both of these questionnaires were therefore discarded as they did not fall within the criteria as defined in 3.6.7a, giving a total of 52 questionnaires to be collated and analysed within Cycle 2 of this investigation.

It was acknowledged the questionnaire would elicit responses based on teachers' perceptions and were likely to be influenced by each respondent's current knowledge base, past experience and any additional professional development and specialism (for example, respondents working in a school for the deaf may be qualified or training to be teachers of the deaf). A detailed discussion of the question format used and how this might influence responses given is provided in 3.6.4. It would be important to recognise how the type of question form and the phrasing used might affect the responses (e.g. whether a closed or open question is used, understanding of the language used, etc.).

The data arising from the questionnaire is presented below in sections that mirror the categories used to structure the questionnaire and includes both analysis and an initial discussion.

5.3.1 Initial checks

An initial check of information was made to ensure that each child for whom a questionnaire was completed had a confirmed medical diagnosis of CHARGE syndrome, was within Early Years to Key Stage 4 of the educational stages, and that they were attending an educational environment as defined in 1.6.3. To ensure there were no duplications (i.e. only one form was returned in relation to a single child) teachers were asked to provide the initials and date of birth for the child so a check could be made. Where questionnaires were completed for different children within one school setting, a quick comparison of hand-writing showed the questionnaires had been completed by 52 different respondents. This was felt to be important since this questionnaire would be seeking the opinions of the respondents and if several questionnaires had been completed by the same person (although for separate children) this may have influenced the range of overall responses obtained.

5.3.2 A check for consistency of responses

As set out in detail in 3.6.9, a check of the consistency of overall responses was undertaken using Spearman's rho (rank correlation coefficient). The results showed that although there was a slight degree of variation, overall there was a high level of consistency in the responses given.

5.4 Demographics

5.4.1 Age range and current educational stages

The data showed a range of ages from 3-15 years. The current Key Stage (KS) each child was in was also identified, this information being set out in Table 12 below.

TABLE 12: THE NUMBER OF CHILDREN FROM THE STUDY IN EACH KEY STAGE

Phase	Years	Ages of children in key stage	Number of children from study in each Key Stage N=52
Early Years Foundation (EYFS)	Early Years	0 to 5 ³	4 (8%)
Key Stage1 (KS1)	1-2	5 to 7	9 (17%)
Key Stage 2 (KS2)	3-6	7 to 11	14 (27%)
Key Stage 3 (KS3)	7-9	11 to 14	12 (23%)
Key stage 4 (KS4)	10-11	14 to 16	13 (25%)

Overall these figures showed a spread of children across the educational phases although with fewer in the Early Years Foundation Stage (EYFS). Since the questionnaire was sent to teachers of children in more formal education settings (according to the definition of an 'educational environment' established for the purposes of this investigation -see 1.6.3), it is recognised this did not include children under the age of 5 who are not yet placed in a formal educational setting. This information would allow for some comparison of data across different educational

³ This study did not generally include 0-3's and only included 4-5 year olds placed in a formal educational setting.

phases that might highlight the possibility of different issues occurring over time which may relate to the age and development of the child, and the format and delivery of the curriculum at the different stages.

5.4.2 Type of educational provision

The data obtained showed the children sampled were placed in a range of different educational provision as presented in Table 13.

TABLE 13: TYPES OF EDUCATIONAL PROVISION WHERE CHILDREN WERE PLACED

Type of school	Total number of children N=52
Severe Learning Difficulties/Profound and Multiple Learning Difficulties (of which 4 are in a sensory/MSI resource)	21 (40%)
Specialist school for the deaf	12 (23%)
Specialist school for speech & language impairment	2 (4%)
Specialist school for physical disabilities/Moderate Learning Difficulties	1 (2%)
Mainstream primary school (including 2 in a Hearing Impairment resource)	10 (19%)
Mainstream secondary school (including 2 in a Hearing Impairment resource)	6 (12%)

This showed 16 (31%) children were placed in a mainstream setting (with 4 having access to a hearing impairment resource base on site). The remaining 36 (69%) were all in special school provision, although of different types and not all indicative

of cognitive impairment. From this information it is not possible to firmly establish a relationship between the type of placement and degree of learning difficulties experienced by the child but, as mentioned previously, it may be assumed that those 21 children placed in settings for pupils with Severe Learning Difficulties (SLD) /Profound and Multiple Learning Difficulties (PMLD) are likely to experience more significant cognitive impairment than those children in mainstream settings or, to a lesser extent, other specialist settings. It is important to recognise the range of different educational placements attended by children within this study may be due to a number of other factors apart from the child's perceived cognitive ability, including the child's sensory needs, what is available within the local authority, advice given regarding placement and parental choice.

From the data arising from this questionnaire it is not possible to identify why a child was placed in a particular educational setting and further research may be helpful to identify possible influencing factors.

When considering the educational placements of children in the different educational phases it was found that:

- 27 children were in Early Years/Key Stage 1/Key Stage 2, of whom:
 - 10 were in a mainstream setting
 - 9 were in an SLD/PMLD setting
 - The remaining 8 children were in other specialist school provisions
- 25 children were in Key Stage 3/Key Stage 4, of whom:
 - 6 were in a mainstream setting

- 9 were in an SLD/PMLD setting
- The 10 remaining children were in other specialist school provisions (7 being in residential placements)

This data shows a division between different types of placement but does not allow for any further accurate comparison. It does raise the question of whether a learner with CHARGE continues in the same type of provision as they progress through the education system; a question that supports the undertaking of a longitudinal study.

5.5 Child's primary need

In 4.7 it is explained there is a requirement for schools to complete the annual School Census (Department for Education, 2015)), a strategic resource containing data of pupil characteristics that includes the identification of a child's primary need. Responses showed that 46 (88%) of respondents considered the child's sensory impairments to be their primary need (see 4.7). The remaining 6 respondents who considered the child's sensory impairments were not their primary need reported they identified it was rather:

- Quadriplegic cerebral palsy (1)
- Speech and language difficulties (1)
- Distractibility (2)
- Coordination (1)
- Learning difficulties (1)

According to Brown (2006; 2007b; 2009) distractibility and poor coordination can result from a poorly developed proprioceptive and/or vestibular system and the sensory integration difficulties that are often felt to be inherent in CHARGE (Brown, 2003a). Therefore, although not identified as a sensory issue by the respondents, these may result from the child's sensory impairments, although may be a consequence of other issues within the child such as low body tone, or external factors such as the learning environment. Of those who felt sensory impairment was not the child's primary need, only one felt learning difficulties were the primary need. This is interesting since 17 (33%) of this study were in a provision for children with SLD/PMLD without a sensory/MSI resource, and only 20 (38%) were in a specialist provision for sensory impairments. The issue of whether 'primary need' is a relevant concept when educating learners with CHARGE will be reviewed at the end of this chapter (5.11) in light of other evidence arising from the exploration of data gathered.

5.6 The child

It was intended that this section of the questionnaire would gather demographic information but also focus particularly on gathering evidence to address the research questions relating to factors within the child that might affect learning (including areas of strength and need) that would also allow for comparison with the broader deafblind population.

5.6.1 Characteristics and sensory responses: The Child

Respondents were asked if a number of anomalies and characteristics were present in the child with CHARGE they were supporting, with the responses summarised in

Table 14 overleaf. The data gathered showed the rate of occurrence of visual impairment, hearing impairment and vestibular difficulties in this cohort were very similar to other prevalence rates previously identified (e.g. Hartshorne et al., 2011). This data is important as it suggests a likeness in the presence of these anomalies between this cohort of children with CHARGE and the wider CHARGE population, an indicator that the sample used for this investigation is consistent with the wider CHARGE population. It was also found that 87% of this cohort was reported to have combined visual and hearing impairments; this provides an evidence-based figure and relates to figures quoted in literature (e.g. Hartshorne et al., 2011), but may be the first time this information has been evidenced through research. These figures provide further supporting evidence that a significant proportion of individuals with CHARGE will be deafblind/MSI although recognising this also implies a number will not have combined visual and hearing impairments. The data also shows other senses are likely to be affected, so the child with CHARGE is therefore not likely to be solely single sensory impaired but also not 'just' deafblind, as indicated by the number of other characteristics and anomalies associated with this condition and found within this cohort.

The presence of the other characteristics reported to be present in a large number of children within this cohort also reflect what is presented in literature, with the exception of touch sensitivity. Only 35% of respondents perceived this ("a reluctance to use touch to explore or to touch certain textures") to be a difficulty; it is possible this may not be as significant for learners with CHARGE as previously suggested in

TABLE 14: PRESENCE AND DISTRIBUTION OF CHARACTERISTICS AMONG TYPES OF PROVISION AND EDUCATIONAL PHASES

	Yes	No	Don't know	SLD/PMLD provision	Mainstream provision	Other specialist provision	EY/ KS1	KS2	KS3	KS4
	(N=52)			(N=21)	(N=16)	(N=15)	(N=13)	(N=14)	(N=12)	(N=13)
Visual impairment	48 (93%)	4 (7%)		21	13	14	11	14	12	11
Hearing impairment	49 (94%)	3 (6%)		19	15	15	13	12	12	12
Combined VI/HI	45 (87%)			19	12	14	11	12	12	10
Vestibular difficulties	48 (93%)	2 (3.5%)	2 (3.5%)	20	16	12	13	12	12	11
Poor body awareness/ coordination difficulties	43 (83%)	7(13.5%)	2 (3.5%)	17	16	10	11	9	12	11
Touch sensitivity	18 (35%)	29 (56%)	5 (9%)	8	6	4	8	3	3	4
Difficulty using senses in a coordinated way	36 (69%)	11 (21%)	5 (9%)	16	11	9	11	11	9	5
Difficulty with distractibility/ remaining on task	39 (75%)	13 (25%)		19	11	9	7	12	11	9
Difficulty with sensory overload/ under-stimulation	39 (75%)	9 (18%)	4 (7%)	18	10	11	9	12	9	9
High anxiety or stress levels	31 (60%)	18 (34%)	3 (6%)	14	10	7	6	11	8	6
High levels of fatigue	27 (52%)	21 (41%)	4 (7%)	12	9	6	7	10	5	6

literature (e.g. Davenport and Hefner, 2011), or at least not present for many in this cohort of children.

The presence of behaviours that may be linked to sensory integration difficulties (see 2.2.8) were reported for large numbers of children within different age groups and educational placements, these being:

- Poor body awareness and coordination difficulties (43/83%)
- Difficulty using their senses together in a coordinated way (36/69%)
- Difficulty with sensory overload or under-stimulation (39/75%)
- Being distractible and finding it difficult to remain on task (39/75%)

The data obtained showed 31(60%) children were considered to experience high anxiety or stress levels. No research figures were found to allow comparison with the prevalence of this in the wider deafblind/MSI population. This is, however, an issue also identified within this group of learners as reflected in the definition given with the PLASC (Department for Education and Skills, 2003) where it is explained that multi-sensory impairment:

Can result in high anxiety and multi-sensory deprivation. (p.7)

Increased levels of anxiety have also been identified in other groups of learners with special educational needs. For example Kim et al. (2000), found the cohort of children with autism studied demonstrated higher levels of both anxiety and depression than children without learning difficulties, although no specific figures were offered. A survey undertaken by Meltzer et al. (1999) found mental disorders occurring in 6% of children without special educational needs in contrast to 40% of

children with a Statement of Special Educational Needs. Thus it can be shown increased levels of stress and anxiety are not unique to learners with CHARGE, nonetheless, the figures from this study suggest these levels may be greater than those experienced by other children with special educational needs. It is possible some of the characteristics included might be influenced by age or cognitive ability (e.g. distractibility, difficulty with sensory overload, high levels of fatigue), and a comparison of data was therefore undertaken looking across the educational phases and types of provision as also set out in Table 14. This analysis revealed the presence of these characteristics, although not reported for every child, were identified across both the age-ranges and different types of educational provision, and may therefore need recognition throughout a child's education and for learners in different educational settings.

5.6.2 Communication: The Child

Initially within this section respondents were asked what factors within the child they considered affected the child's communication. Although this open question was posed in relation to the child's communication, on reflection it may have been more beneficial to have set this question with regard to the child's overall learning. Nonetheless some interesting information was obtained from the responses given in relation to the child's communication. The results obtained are summarised in Table 15 including the distribution of these factors across age and type of educational provision.

A combination of factors was reported for 42 children, an indicator of the potential complexity of this condition. Sensory impairments were the most commonly reported factor affecting communication in 42 (81%) children. Although these responses are likely to be a reflection of the rate of occurrence of sensory anomalies in this condition it is interesting to note that this is less than the number of children with reported hearing and visual difficulties, suggesting teachers did not always perceive the child's sensory issues to affect their communication. Additional sensory elements were also identified (sensory processing and sensory integration difficulties), and overall these figures reflect a recognition of the impact sensory impairments can have on this area of a child's development. This relates to Aitken's (2000) suggestion that two of the major challenges faced by individuals who are deafblind/multi-sensory impaired are finding out information, and communicating with others as a consequence of their sensory impairments. Other physiological factors were also named and additional medical needs. Learning difficulties were considered to be a factor for 29% of the cohort showing that although learning difficulties were not reported to be a child's primary need (for all but one child studied), they may impact on a child's functioning. The relationship between a child's communication abilities and learning difficulties may be a reciprocal one with one impacting upon the other. It was interesting to note that whilst 9/15 children where this was reported were placed in SLD/PMLD provision, 3 were in mainstream settings and a further 3 in other specialist settings. Although it is recognised that many people with learning disability will experience communication difficulties (Jones, 2002, quotes between 50%-90% of people with learning disability experiencing such difficulties) this relationship in individuals with CHARGE may not be quite so clear given the range of

TABLE 15: FACTORS WITHIN THE CHILD CONSIDERED TO AFFECT THE CHILD'S COMMUNICATION, AND DISTRIBUTION AMONG TYPES OF PROVISION AND EDUCATIONAL PHASES

	Total where factor reported (N=52)	SLD/PMLD provision (N=21)	Mainstream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Sensory impairments	42 (81%)	20	11	11	9	11	9	13
Learning difficulties	15 (29%)	9	3	3	3	4	4	4
Distractibility	10 (19%)	4	4	2	1	3	4	2
Sensory processing/SI difficulties/time needed to process information	7 (13%)	3	1	3	1	2	3	1
Speech/articulation difficulties (cleft palate/ facial palsy)	5 (10%)	2	2	1	1	2	-	2
Medical needs	5 (10%)	4	1	-	3	-	1	1
Fatigue	3 (6%)	1	2	-	1	1	1	-
Behaviour	3 (6%)	-	2	1	-	2	1	-
Stress and anxiety	2 (4%)	2	-	-	-	-	1	1
Executive function difficulties	1 (2%)	-	1	-	1	-	-	-
Poor social skills	1 (2%)	1	-	-	-	-	1	-
Understanding concepts	1 (2%)	-	-	1	1	-	-	-

intellectual ability found in CHARGE (see 2.2.3); in a study by Dammeyer (2012), 71% of the cohort of 17 learners with CHARGE were found to be without 'intellectual disability' whilst 83% were reported to have 'moderate or severe language delay' (p.1294). Only 1 respondent directly identified executive function difficulties as a factor within the child considered to affect communication. Other respondents, however, also noted other factors that the examination of literature showed may be consequential to executive dysfunction (see 2.2.9), these being distractibility (noted by 10 respondents), poor behaviour (3), poor social skills (1) and the formation of clear concepts (1). This suggests it is possible that although teachers are recognising the impact of potential executive dysfunction they do not have either a specific concept of executive function nor direct knowledge of it, and may not be aware of the presence of executive dysfunction itself within the child. It is also important to recognise there may be a reciprocal element involved in the factors identified where, for example, behaviour was identified by 3 respondents as a factor but it may also be viewed conversely, that communication difficulties can adversely affect a child's behaviour, for example as a consequence of frustration.

5.6.2a Child's preferred communication modes

In this section respondents identified what they considered the child's preferred communication modes were for both receptive and expressive communication. On this occasion the respondents were not provided with an inventory, although one was provided for the following question set out on the same page that could provide a prompt for the respondent when answering. The data obtained (see Table 16) showed a range and combination of communication modes reported as being

preferred by children within this cohort and that different modes (and combinations) were used by children across the age-range. However, the findings also showed:

- Children in SLD/PMLD settings were those predominantly perceived to show a preference for the use of concrete (solid and tangible) cues
- Children in mainstream and other specialist settings were predominantly perceived to prefer using speech and/or sign

This may be a reflection of the needs of the children in these different settings arising from cognitive impairment and other anomalies, but may also be influenced by other factors, such as these communication modes being made available to the child and the skills of the supporting adults. Overall, from the data provided it was calculated the number of modes preferred by each child within this cohort for receptive communication ranged from 1-3 with an average of 1.6, and for expressive communication a range of 1-2 with an average of 1.4 modes. A comparison of the consistency in the use of modes preferred by each child for receptive and expressive communication was also undertaken which found 24 (46%) of children studied were perceived to prefer to use the same modes for both expressive and receptive communication. For the remaining 28 children it was noted that:

- 7 (13%) children showing a preference for using some form of concrete cue to express themselves, whereas 17 (33%) were perceived to prefer to make use of concrete cues to receive communication (only one of these was in a mainstream setting)
- Children in a mainstream setting who used a combination of speech and sign to receive communication were reported to use only speech to express themselves

TABLE 16: CHILD'S PREFERRED COMMUNICATION MODES TO RECEIVE COMMUNICATION AND EXPRESS THEMSELVES, AND DISTRIBUTION AMONG TYPES OF PROVISION AND EDUCATIONAL PHASES

	Communication modes	Total where factor reported (N=52)	SLD/PMLD provision (N=21)	Mainstream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Receptive	Speech/vocalising	16	3	9	4	5	3	3	5
	Sign/ gesture	9	1	2	6	1	3	3	2
	Speech & sign	10	4	4	2	3	2	-	5
	Speech, sign and concrete cues	2	2	-	-	-	1	1	-
	Vocalising & gesture	-	-	-	-	-	-	-	-
	Concrete cues (e.g. objects, photos, symbols)	6	3	-	3	1	3	1	1
	Speech & concrete cues	5	4	1	-	2	1	1	1
	Sign/ gesture & concrete cues	4	-	-	-	-	1	2	1
	Behaviour	-	-	-	-	-	-	-	-
	Behaviour & concrete cues	-	-	-	-	-	-	-	-
Expressive	Speech/ vocalising	19	2	13	4	6	4	3	6
	Sign/ gesture	14	7	2	5	2	4	3	5
	Speech & sign	5	2	-	3	1	2	-	2
	Speech, sign and concrete cues	-	-	-	-	-	-	-	-
	Vocalising & gesture	2	2	-	-	-	2	-	-
	Concrete cues (e.g. objects, photos, symbols)	1	1	-	-	-	1	-	-
	Speech & concrete cues	2	2	-	-	-	-	1	1
	Sign/ gesture & concrete cues	3	3	-	-	1	-	1	1
	Behaviour	4	2	1	1	2	1	1	-
	Behaviour & concrete cues	2	2	-	-	1	-	1	-

This demonstrates the need for provision of a range of different modes to be made available to the learner with CHARGE without making assumptions about particular modes that should be employed. It also recognises that for a number of children with CHARGE there may be differences in the preferred channels for both communicative reception and expression. Some children may also have a greater reliance on the use of concrete modes to receive communication than to express themselves. This could result from the executive function difficulties felt to be associated with CHARGE (see 2.2.9), with concrete cues helping to overcome the difficulty of understanding abstract concepts (and the symbolic representation of sign and speech), but may also be a consequence of the additional processing time needed (given the transient nature of speech and sign and the difficulty receiving whole information). It would be interesting to undertake future research to ascertain whether learners with CHARGE are more dependent than other deafblind learners on the use of concrete cues to support their communication.

5.6.2b Communication modes

Within the questionnaire respondents were provided with an inventory and asked to identify the different modes used with the child to support communication, the results being set out in Table 17. This showed a range of different modes were used to support the child and reflects the use of a combination of communication modes that is recognised in literature on deafblindness (e.g. Pease, 2000; Murdoch et al., 2009). Interestingly, in addition to speech/sign (both communication modes more transient in nature), teachers across the different types of educational settings identified the use of concrete communication modes, even though previously only 1 child in a

TABLE 17: COMMUNICATION MODES USED TO SUPPORT COMMUNICATION AND DISTRIBUTION AMONG TYPES OF PROVISION AND EDUCATIONAL PHASES

Communication modes	SLD/ PMLD provision (N=21)	Main- stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Speech	17	16	12	13	13	8	11
Sign	18	7	12	11	10	8	8
Photographs	17	8	10	10	8	9	8
Symbols & drawings	15	7	10	6	11	9	6
Reading child's behaviour	18	6	6	4	9	9	8
Object cues/ objects of reference	15	8	5	6	10	7	5
Printed word	6	12	9	8	7	5	7
Touch cues	8	2	-	4	3	1	2
Switch use	1	-	-	1	-	-	-
iPad	1	-	-	1	-	-	-
PECS	1	-	-	-	1	-	-

mainstream setting and 3 children in specialist settings other than SLD/PMLD provision had earlier been reported to have a preference for using a concrete mode for receptive communication.

The number of communication modes used with each child was then analysed (see Table 18) and it was found children within this cohort were being exposed to a far greater number of communication modes being used by those around them in the learning environment to support communication than the child's own use of preferred modes. The possible implications of this can be viewed in different ways. For example, it might be argued it is necessary to model use of additional communication modes to those currently used by the child, or conversely that the child is being overwhelmed by the presentation of so many different communication modes.

TABLE 18: NUMBER OF COMMUNICATION MODES USED WITH A CHILD TO SUPPORT COMMUNICATION AND DISTRIBUTION AMONG TYPES OF PROVISION

No. of modes	No. of children (n=52)	SLD/PMLD provision (N=21)	Mainstream provision (N=16)	Other specialist provision (N=15)
1	4	0	3	1
2	4	0	2	2
3	3	1	1	1
4	10	4	2	4
5	10	5	4	2
6	12	5	2	4
7	4	4	0	1
8	2	0	2	0
9	2	2	0	0
Average	4.7	5.7	4.1	4.3

Other analysis of data showed little variation in the average number of communication modes used across the different age-ranges. The data does however show that children in an SLD/PMLD setting were exposed to the use of a larger number of communication modes than children in other types of provision. This is of interest since the nature of children attending an SLD/PMLD school indicates a significant cognitive impairment and yet they are exposed to a large number of different communication modes. The high number of different modes being used leads one to question how capable the children would be of working with this number of systems and/or languages. There are some possible reasons for this that could include:

- More use being made of a combination of a wider number of different modes as staff in special schools may have greater awareness and experience of using these different forms to support communication
- It is more difficult to communicate effectively with these children and so a wider number of modes are employed to try and support this
- The children benefit from a greater number of communication modes being used

This is another example of the potential influence of the perceptions and knowledge and specialisms of the teacher together with the underlying ethos of the educational placement. From this research it is not possible to draw any conclusions, but this is an area that would benefit from further exploration and investigation in future studies.

5.6.2c Communicative exchanges with a sensitive communication partner

At this point a Likert scale was first introduced (with clear instructions for use) that would then be used at intervals in the section about the child. A summary of the average scores obtained for each of the questions measured using a Likert Scale is set out in Table 19.

Participants recorded how easy or difficult they perceived a given skill to be for the child in question with individual scores ranging from 1 (being very easy) to 5 (being very difficult). The total scores could range from 52 (if every child was reported to find it very easy) to 260 (if every child was perceived to find it very difficult) with a midway score of 156 (neither easy nor difficult). Therefore, the higher the score, the more difficult a skill was perceived to be for the cohort overall. More detailed data is presented in Appendix 8 and will be referred to where appropriate as each aspect is discussed.

With regard to communication, participants were asked their opinion of how easy or difficult it was for the child to initiate and maintain communicative exchanges with a sensitive communication partner. The data obtained (detailed analysis in Appendix 8, Table (v)) revealed 29 (56%) of children reported on in the study were considered to find it easy or very easy to initiate a communicative exchange, given a sensitive communication partner, but that overall maintaining a communicative exchange was regarded as more difficult. The figures showed a similar pattern of distribution of scores across the different types of educational settings and phases and suggest

TABLE 19: A SUMMARY OF THE AVERAGE SCORES OBTAINED FROM THE LIKERT SCALES RELATING TO 'THE CHILD'

	SLD/ PMLD provision (N=21)	Main- stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)	Total scores	Average (mean) of total scores
Communication									
How easy/difficult is it for the child to <u>INITIATE</u> communicative exchanges with a sensitive communication partner?	2.4	2.7	2.5	2.6	2.6	2.5	2.3	131	2.5
How easy/difficult is it for the child to <u>MAINTAIN</u> communicative exchanges with a sensitive communication partner?	2.9	3.3	2.9	3.1	2.9	3.3	2.7	156	3
Social and emotional									
How easy/difficult to develop trust and confidence with adults in school setting?	2.4	2.3	2.6	2.2	2.8	2.3	2.5	126	2.4
How easy/difficult to develop genuine friendships with peers in school setting?	4.6	3.9	3.7	3.6	4.3	4.3	4.2	213	4.1
How easy/difficult to understand their own emotions & express their own emotional state?	4.2	3.9	3.4	3.9	4.1	4.5	3.7	205	3.9
How easy/difficult to empathise with his/her peers when they are hurt, angry, upset etc.?	4.7	3.4	4.0	4.0	4.2	4.6	3.6	213	4.1

	SLD/ PMLD provision (N=21)	Main- stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)	Total scores	Average (mean) of total scores
Conceptual ability									
How easy/difficult is it for the child to establish real, concrete concepts?	2.6	2.0	2.1	2.8	1.9	2.5	2.0	119	2.3
How easy/difficult is it for the child to establish abstract concepts?	4.2	3.1	3.3	3.7	3.1	4.3	3.4	188	3.6
How easy/difficult is it for the child to identify solutions to simple problems?	3.2	3.3	3.1	3.2	3.2	3.5	2.8	166	3.2
How easy/difficult is it for the child to transfer and generalise skills and knowledge and apply it to different situations?	3.8	3.5	3.5	3.7	3.7	3.7	3.4	188	3.6
How easy/difficult is it for the child to recall a sequence of events?	3.3	2.8	3.1	3.5	3.3	3.4	2.6	171	3.3
How easy/difficult is it for the child to correctly order parts of an activity or story sequence?	4.1	2.8	2.6	2.2	3.4	4.0	2.6	171	3.3
Response to routine and structure, and understanding of time and space									
How easy/difficult is it for the child to cope with the unexpected & changes in their routine?	3.6	3.2	3.7	3.7	3.4	3.4	3.5	183	3.5
How easy/difficult is it for the child to cope <u>without</u> regular routines & structure?	4.0	3.9	4.0	4.2	3.8	4.0	4.0	208	4
How easy/difficult is it for the child to anticipate what is to happen next when appropriate cues provided?	2.1	2.5	2.4	2.2	2.4	2.3	2.3	120	2.3

	SLD/ PMLD provision (N=21)	Main- stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)	Total scores	Average (mean) of total scores
Response to routine and structure, and understanding of time and space (contd)									
How easy/difficult is it for the child to predict what is to happen tomorrow or at the weekend?	4.0	3.1	3.1	3.5	4.2	3.3	2.5	181	3.5
How easy/difficult is it for the child to recall past events?	3.8	2.8	2.8	3.8	2.9	3.5	2.6	166	3.2
Ownership of learning									
How easy/difficult is it for the child to learn and play independently?	3.3	3.6	3.4	3.5	3.6	3.6	3.2	179	3.4
How easy/difficult is it for the child to organise him/herself?	4.0	3.8	3.4	4.2	3.9	4.0	3.2	197	3.8
How easy/difficult it for the child to initiate activities?	3.5	3.8	3.3	4.2	3.7	3.5	3.7	184	3.5
How easy/difficult it for the child to undertake self-help skills and functional life skills (appropriate for age)?	3.7	3.8	3.0	3.6	3.8	3.5	3.2	184	3.5

these skills are attainable for a large proportion of the cohort when provided with a sensitive communication partner.

Considering comments in literature relating to CHARGE where, for example, Thelin and Fussner (2005) found poor communication ability was common in CHARGE, it would seem a key factor is the availability of a sensitive communication partner to facilitate children in undertaking and maintaining communicative exchanges (35/67% of respondents would later report the provision of a sensitive communication partner was a helpful strategy to support communication - See 5.7.2). This may also provide some insight into why children with CHARGE have been found to prefer adult rather than child company (Souriau et al., 2005), and are reported by parents to have difficulty establishing same-age relationships (Hartshorne & Cypher, 2004), since it is likely that peers (especially younger children or peers with learning difficulties) are less likely to have the skills to be a sensitive communication partner than supporting adults, or to have the degree of motivation to undertake this role.

5.6.3 Social and emotional: The Child

In this section respondents were asked to identify the number of adults with whom the child was felt to have a secure and trusting relationship and the number of peers with whom respondents felt the child had a genuine friendship in school, the data being presented in Appendix 8, Table (vi). On average, children in an SLD/PMLD setting were noted to have slightly fewer relationships with adults than learners in other settings. This may be a reflection of the impact of a learning disability but may

also involve other factors (such as the ethos of the school where there is the provision of a few consistent key workers, and the number of staff involved in delivery of the curriculum). An increasing average through the educational phases was also noted that may be suggestive of maturation and developing skills within the child, although again other factors may be involved, such as increasing the numbers of adults working with a child in the educational setting at the later key stages thus increasing opportunity to engage with a larger number of people. A similar pattern was found in the average number of genuine friendships the children were perceived to have according to the type of educational placement the child attended (i.e. lower for children in an SLD/PMLD setting) although with less variation in the averages obtained. In this instance age appeared not to be a factor. These figures show children within the cohort were considered to have formed fewer friendships with peers in the school setting and 25 (48%) children were felt to have no genuine friendships with their peers (14 in SLD/PMLD provision, 5 in mainstream and 6 in other specialist settings). The figures indicate children in SLD/PMLD provision were had the fewest peer friendships and children in mainstream provision had formed the most. This may be a reflection of the impact of learning difficulties but may also be dependent on the availability of an appropriate peer group. Overall, within the cohort children had fewer relationships with peers than adults.

The datum on the 5 individuals who were considered to have more than 10 secure relationships with adults in the learning environment was examined further to examine whether these children were also more successful in forming peer relationships. The findings of this comparison, set out in Table 20 overleaf, show of

these 5 children, 2 were felt to have no genuine peer friendships and each of the others had far fewer friendships with peers than relationships with adults, thus reflecting the pattern found for the overall cohort.

TABLE 20: COMPARISON OF THE NUMBER OF PEER FRIENDSHIPS IN 5 CHILDREN FOR WHOM MORE THAN 10 SECURE RELATIONSHIPS WITH ADULTS WERE REPORTED

Child	A	B	C	D	E	Average number of relationships/friendships
Secure adult relationships	12	14	15	16	20	15.4
Genuine peer friendships	4	6	0	0	4	2.8

The data presented above was then compared to that obtained through the use of the following Likert Scale that recorded how easy or difficult it was considered to be for the child to develop relationships with adults or peers in the school setting (See Table 19). This comparison showed that although children in mainstream and specialist settings other than an SLD/PMLD school were reported on average to have established more relationships with adults and peers, the degree of difficulty children were perceived to have in establishing relationships was similar across the different types of educational provision and age range.

Other questions within this particular section related to the ability to understand or express their own emotions or to empathise with their peers. Examination of the responses given (see summary - Table 19 and detail in Appendix 8, Table (vii)) showed a large percentage of children across the types of provision and educational phases found it difficult or very difficult to understand and express their own

emotional state (71%) or to empathise with their peers (69%), although it must be noted that this was again perceived to be most difficult for learners in SLD/PMLD settings. Further comparison of the data obtained found that, with only two exceptions, those children who were perceived to find it difficult or very difficult to understand or express their own emotions also had difficulty empathising and forming relationships with their peers. Poor social and emotional skills in learners with CHARGE are well-documented in literature (see 2.3.2) with Hartshorne et al. (2007) reporting these children tend to have “limited social success” (p.340). What defines ‘social success’ is not clearly established but, if this relates to the forming of relationships with others of the same age, this would reflect the figures from this investigation and the findings of the study by Hartshorne and Cypher (2004). Given the view, however, that one of the major challenges faced by all deafblind individuals is a difficulty in communicating with others (e.g. Aitken, 2000), it is clear this is not a difficulty likely to be uniquely experienced by children with CHARGE. Other research has also shown learners with a single sensory impairment can also experience difficulties with peer relationships where, for example, Nunes et al. (2001) found hearing impaired pupils in mainstream settings were:

More likely to be neglected by their peers and less likely to have a friend in class. (p.123)

Poor communication skills within the child are likely to be a factor (e.g. how does a child who is dependent on using concrete cues to communicate express an abstract emotional state or empathy?), but there may also be additional factors such as the adults and peers themselves, other demands being placed on the child, and whether strategies are implemented to support the development of peer relationships. Hodges (2000), for example, explains that individuals who are deafblind will need support to

learn to recognise and understand the emotions of themselves and other people since they miss all the verbal and auditory cues that may convey this information to a hearing-sighted person, and this will include individuals with CHARGE. What is not clear is whether there is a greater degree of difficulty experienced by individuals with CHARGE and what other factors might be and this would require further investigation to examine what elements support/inhibit the development of relationships with both adults and peers. There is a growing interest in the social and emotional development in individuals with CHARGE as reflected in articles such as Kennert et al. (in press) and the content of the Deafblind International (DbI) CHARGE network pre-conference meeting (2013). It would be beneficial to support this discussion with evidence obtained through further research undertaken in this area, potentially through the use of case studies and an identification of effective supportive strategies.

5.6.4 Conceptual ability: The Child

Within this section respondents were asked to share their perceptions of the child's development of aspects of conceptual ability (See Table 19 and detailed data in Appendix 8, Tables (viii) and (ix)). Only 9 children overall were reported to find establishing real concrete concepts difficult or very difficult (6 in SLD/PMLD provision, 2 in mainstream and 1 in another specialist setting), suggesting that learning may need to be grounded in real life experiences supported by the use of concrete tools to build on this strength. In contrast, the establishment of abstract concepts was perceived to be one of the most difficult areas for the children reported on for this study. Difficulty in developing abstract concepts would be anticipated in learners with

learning disabilities (Geary et al., 2000) but it is interesting to note that although proportionally more children in an SLD/PMLD setting were reported to find this skill difficult or very difficult, a number of children in the other types of settings were also similarly reported (6/16 children in mainstream and 7/15 children in other specialist settings). It was recognised the cohort included children not of an age where abstract concepts would be typically developing, and therefore the ages of the children and relationship between the responses obtained would need to be analysed further. According to Piaget & Inhelder (2000) the ‘Concrete Operational’ stage where children think logically but are very concrete in their thinking occurs typically between the ages of 7-11 years, and the ‘Formal Operational’ stage where children develop abstract reasoning typically occurring between 11-16 years. Thus one would not expect children in Early Years and Key Stages 1 and 2 to be typically establishing abstract concepts so it was necessary to focus on the scores of the 25 children in this cohort in Key Stages 3 and 4, this data being set out in Table 21.

TABLE 21: SCORE OF CHILDREN IN KS3/4 FOR EASE OR DIFFICULTY OF ESTABLISHING ABSTRACT CONCEPTS

N=25		SLD/PMLD provision	Mainstream provision	Other specialist provision
1=very easy 5=very difficult	1	-	1	-
	2	1	2	2
	3	3	1	1
	4	3	1	1
	5	4	1	4

The impact of a child’s cognitive impairment on concept development is again reflected in these figures with most learners in an SLD/PMLD setting finding the

establishment of abstract concepts either difficult or very difficult. However, a number of children in other educational settings were also perceived to experience a similar level of difficulty which may indicate it is important to recognise this as a potential issue for some more able learners with CHARGE.

The four questions addressed pertaining to conceptual skills perceived to be the most difficult all required the development of abstract concept formation and abstract thinking; that is, not related to a specific instance or object and removed from the facts of here and now. This was noted to be most difficult for children in an SLD/PMLD setting (most likely linked to cognitive impairments) but was also observed in children across the age range and educational settings. Concept formation is widely recognised as an issue for those with learning disability and all learners who are deafblind. It is addressed at some length by Miles and McLetchie (2008) who explain concepts:

Are the ideas that give meaning to our world. (p.1)

These will develop according to each child's individual experiences and, in common with all deafblind children, the child's sensory impairments must be recognised as a factor that will influence their concept development. It would be interesting to explore whether, for the child with CHARGE, the potential for all sensory modalities to be affected alters their experiences further, thus impacting differently on this concept formation than for other deafblind children. In addition to the impact of the child's sensory impairments, the presence of learning difficulties, and the child's age, an additional implicating factor may be executive dysfunction since the abilities to form

concepts and think abstractly are often considered components of executive function (see 2.2.9).

When considering the identification of solutions to problems, on average children were scored at the midway point (it was considered neither easy nor difficult) across both the different types of educational provision and phases, so although only 12 (23%) children were reported to find it either easy or very easy, conversely only 19 (37%) were felt to find it either difficult or very difficult. It is interesting to view this finding in light of the opinion of Hartshorne et al. (2007) who identified problem-solving as being challenging for learners with CHARGE as a result of executive dysfunction; this data suggests that whilst this is a difficulty for some learners with CHARGE it is less so for many others and may be regarded as a relative strength. Children with CHARGE are recognised as developing adaptive behaviours that some authors consider are suggestive of underlying intelligence (e.g. Salem-Hartshorne and Jacob, 2004, 2005; Hartshorne et al., 2005b). If considered more broadly, these adaptive behaviours may result from the child's efforts to make the environment 'work for them'; for example, as a result of vestibular difficulties the child can experience issues maintaining an upright posture, and addresses this problem by learning to compensate through increased movement or gaining additional support through propping and leaning (Brown 2007). This might therefore be considered as simple problem-solving and as such a relative strength as a consequence of the child needing to make the world around him a manageable place. The question is whether the teachers involved in this study (and more widely) recognise these behaviours as

problem-solving in addition to other ways that the child might demonstrate developing solutions to simple problems, such as using a step to reach a light switch.

5.6.5 Responses to routines and structure, and understanding of time and space: The Child

Respondents were asked to consider this aspect of learning and development, again recording their perceptions through the use of a Likert scale. A summary of the results is shown in Table 19 with further details in Appendix 8, Tables (x) and (xi). Of the children within this cohort, across the different educational settings and phases only 7 (13%) were reported to find it easy to cope without regular routine and structure, this being reflected in the total score and the averages obtained. Although this cohort was perceived to have less difficulty coping with unexpected changes to routine the results showed this was still an area of relative difficulty. The need for the use of structure and clear routines when educating children with CHARGE was identified within the literature review (see 2.6.1) but is also addressed in literature relating to the education of children who are deafblind (e.g. Murdoch et al., 2009; Pease 2000; and Nelson et al., 2009) and for children with other special educational needs, including autism (e.g. Jolliffe, 1992 cited in Howlin, 2004, p.137), and learning disabilities (e.g. Troia and Graham, 2002; Davis and Florian, 2004). It was noted that although slightly more children in the mainstream settings were reported to find it easy/very easy to cope with these unexpected changes or lack of routine, most were not perceived to find it so, indicating this is not just a consequence of learning difficulty and other factors are likely to be involved (e.g. the impact of sensory impairments, executive function difficulties). It would be interesting if future research

was able to measure and compare the degree/type of impact caused by changes to routine for deafblind learners in general and learners with CHARGE.

In contrast the figures showed relative ease in anticipating what is to happen next with the provision of appropriate cues, again for children across the educational settings and age range with only 7 (13%) children perceived to find this difficult, and none very difficult. The data also showed that within this section, with the exception of anticipating what is to happen next with the provision of appropriate cues, the mode was higher than the mean for each of the other questions, with more children reported to find it 'very difficult' than any other level of response. The total scores obtained suggest the children in this study overall were considered to find it slightly easier to recall past events than to predict the near future, however, the figures also reveal children placed in an SLD/PMLD setting found these skills more difficult than the children in other types of educational provision indicating that learning difficulties may be a contributory factor. The findings also show that whilst those in the primary phases (Early Years/KS1 and KS2) were perceived to experience greater difficulty in predicting a future event than children in KS3 and KS4, there was little difference across the age range with regard to recalling a past event. This may link to the development of concepts (discussed above in 5.6.4) and also that the child has had direct and immediate experience suggesting the benefit of learning occurring through the use of meaningful tasks undertaken in real settings.

Respondents were then provided with a list of five types of tangible cues and asked to identify those used to support routine and structure and an understanding of time

and space, with the data generated set out in Tables 22 and 23. Respondents were also provided with the opportunity to list other cues although very few additional cues were named. This provides an example of how the use of more structure within the questioning may elicit a greater response than the open, unstructured elements, This raises the question of whether higher levels of other cues may have been reported if they were included in the list provided, or conversely, less frequent reporting of the use of the given cues if the question was left open with no suggestions provided. This pattern in responses would be observed in the responses to the inventories set out in the 'Strategies' section later in the questionnaire and supports Oppenheim's (2000) statement (See 3.6.4, where the format of questions is discussed at some length) that this approach is likely to encourage participants to respond more readily than if presented with an open question.

The data obtained showed a range of tangible cues were employed, with an overall average of 3.3 types of concrete cues being used per child. Greater use was made of these cues with children in specialist provision than for those in a mainstream placement, with greatest use in the SLD/PMLD schools, nonetheless, there were only 2 children overall with whom such concrete cues were not used. A higher average number of cues was reported to be used with the younger children, suggesting there may also be a link to the developmental stage of the children involved and again their concept development (as discussed previously in 5.5.4). Research has shown that memory recall is linked to learning disability, but the author of one particular research paper that reached this conclusion (Swanson, 1993) made an interesting hypothesis suggesting that difficulty recalling events may be a consequence of executive

TABLE 22: TYPES OF CONCRETE CUES USED TO SUPPORT ROUTINE AND STRUCTURE AND UNDERSTANDING OF SPACE AND TIME

Cues	Total number of children where used	SLD/ PMLD provision (N=21)	Main-stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Visual calendar/ timetable	45 (87%)	18	14	13	12	14	9	10
Photographs	38 (73%)	20	7	11	9	10	10	9
Symbols and drawings	33 (63%)	16	6	11	7	11	7	8
Printed word	28 (54%)	9	11	8	8	7	4	9
Object cues	26 (50%)	14	4	8	7	9	4	6
Switches	1	1			1			
Sandtimers	1		1		1			
iPad	1	1			1			

TABLE 23: NUMBER OF CONCRETE CUES REPORTED TO BE USED PER CHILD TO SUPPORT ROUTINE AND STRUCTURE

Number of concrete cues reported to be used with a child:	Total number of children where reported (N=52)	SLD/ PMLD provision		Main-stream provision		Other specialist provision	
		EY/ KS1/ KS2	KS3/KS4	EY/ KS1/ KS2	KS3/KS4	EY/ KS1/ KS2	KS3/KS4
0	2	-	-	-	2	-	-
1	6	-	1	2	-	1	2
2	6	-	2	2	2	-	-
3	10	2	2	2	1	2	1
4	16	4	5	2	-	3	2
5	11	4	-	2	1	2	2
6	1	1	-	-	-	-	-
Average number of cues		4.4	3.1	3.0	2.0	3.6	3.3

function difficulties. If this is so, it could explain why such tangible cues were used with so many children in this cohort across the settings, given the evidence raised in this study that executive dysfunction can affect learning in individuals with CHARGE (see 2.2.9).

5.6.6 Ownership of learning: The Child

Within this section information was again gathered through the use of a Likert Scale, with a summary of the data generated to be found in table 19, with more detailed information set out in Appendix 8, Table (xii). Responses showed that overall the skill of learning and playing independently was felt to be the easiest of the aspects identified, although only 12 (23%) children studied were reported to find this easy or very easy. This may relate to the fact that only 9 (17%) children were perceived to find it easy/very easy to initiate activities that may in turn inhibit independent play and learning. The scores obtained showed that children were perceived to experience similar levels of ease/difficulty across the educational settings, and also across the age-ranges, with the exception of those learners in KS4 who were perceived overall to find it slightly easier to learn and play independently than children in the other phases, suggesting there may be a link to a child's overall development. A similar pattern was found for the skill area addressed where greatest difficulty was reported; that of developing and implementing self-organisational skills that may have implications in adult life in executing more complex daily living skills.

When examining the data relating to the undertaking of self-help skills and functional life skills (appropriate for their age) it was noted that although there was little variation

across the age range, those learners in 'other specialist provision' were perceived to find this skill easier overall than learners in both mainstream and SLD/PMLD settings, remembering that the structuring of scores in the Likert Scale used meant the lower the score, the easier the skill was perceived to be. A comparison was then made between the learners in this category who were in residential and day placements. It was found the average score given for the 7 learners in 4 different residential placements was even lower at 2.4 in contrast to the 3.5 obtained for those learners in day placements (this in fact being similar to those average scores obtained for the children in the other types of provision). This data may indicate a potential benefit of a learner attending a residential placement, where there is the possibility for more focus on supporting the development of self-help and functional life skills outside of the 'normal' school day (what residential schools often describe as a 'waking day curriculum'). This may reflect a paper by Blake et al. (2005) who considered adolescent and adult issues in individuals with CHARGE. They identified a difficulty in higher order, more complex daily living skills (that include self-help skills and functional life skills) and suggest this can be the result of a lack of exposure to learning activities to support the development of these skills earlier in life.

Respondents were then asked to list factors considered to adversely affect the child's readiness and ability to engage and learn. Those identified were a combination of internal and external factors as set out in Appendix 8 Table (xiii). It is important to acknowledge the 2 factors most frequently identified (environmental distractions and fatigue) were given as examples of what might be included in the response, this providing an illustration of how the phrasing of a question can act as a prompt and potentially 'lead' a respondent. Nonetheless, it was felt the inclusion of such

examples was necessary to support a shared understanding of the question that overall would elicit a more accurate response from the participants (as discussed in 3.6.4). The wide range of factors identified highlights the interplay between internal and external factors that may impact upon the child's engagement and learning. This suggests educators need to have an awareness of what is within the child that may need to be recognised and responded to, but that the challenge is also in creating an optimal learning environment and identifying those strategies that are most helpful or inhibiting in the educational setting. The majority of factors reported were noted for children across the different types of provision and age ranges. Those factors raised for pupils in an SLD/PMLD setting or other specialist setting, but not for pupils in mainstream, indicated a need for consistent adults, a reliance on routine and structure with support for transitions and behavioural support. Conversely, stress and anxiety and difficulty with managing the pace of the delivery of the curriculum were noted only for children in mainstream or other specialist provision, and it may be that in SLD/PMLD settings it is more feasible to match the pace of the curriculum to that of the pupil.

5.6.7 Orientation and mobility, and fine motor skills: The Child

5.6.7a Orientation and mobility

In this section respondents were given a list of factors and asked whether they felt they affected the child's orientation and mobility skills (see Table 24). It was perhaps to be expected that sensory impairments were regarded as affecting orientation and mobility for all but one child in this cohort. Through comparison it was found that those sensory impairments most commonly considered to be a factor were:

- Visual impairment for 41/48 children with a reported visual impairment
- Hearing impairment for 39/49 children with a reported hearing impairment

In addition to a child's visual impairment and hearing impairment, vestibular difficulties were also reported to be a factor for 47 (90%) children, this being reported for 47/48 children where vestibular difficulties were previously noted. Whether these are recognised as influencing factors by respondents would be in part dependent on staff knowledge, understanding and experience. It is also likely to be dependent on the degree of impairment and also how each impacts upon the use of the other senses.

This data indicates that within this cohort where a vestibular difficulty is identified, it will most likely to affect orientation and mobility. It is recognised the mobility of many deafblind learners will be affected by their visual and hearing impairments (e.g. Clark, 2000), whereas little recognition appears to be given to the impact of other sensory issues, particularly vestibular difficulties (other than Brown, 2007b). Greater awareness of this is found in literature on CHARGE (see 2.2.4). That might be because this is a more significant issue for individuals with CHARGE as indicated by the results of this study, which suggests these difficulties to be one of the most significant sensory issues that can impact on orientation and mobility. In addition to a child's sensory impairments, respondents also perceived the other aspects included in the question to affect the child's orientation and mobility these being body awareness (50%), coordination and organisation of the body (75%), and fatigue (69%). Overall, issues with body awareness and coordination and organisation of their body were felt to be a factor for 46% children.

TABLE 24: FACTORS OTHER THAN SENSORY IMPAIRMENTS CONSIDRED TO AFFECT THE CHILD'S ORIENTATION AND MOBILITY

	N=52	SLD/PMLD provision (N=21)	Mainstream provision (N=16)	Other specialist provision (N=15)	EY/KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Body awareness	Yes 26 (50%)	12	10	4	8	6	6	6
	No 15 (29%)	6	3	6	4	4	3	4
	Don't know 11 (21%)	3	3	5	1	4	3	3
Coordination and organising body	Yes 39 (75%)	15	15	9	11	9	9	10
	No 8 (15%)	4	-	4	2	3	2	1
	Don't know 5 (10%)	2	1	2	-	2	1	2
Fatigue	Yes 36 (69%)	13	11	12	11	9	9	7
	No 9 (17%)	4	3	2	1	2	2	4
	Don't know 7 (13%)	4	2	1	1	3	1	2

Two further questions were asked relating directly to behaviours that might be observed as adaptive behaviours to vestibular and proprioceptive difficulties (e.g. Brown 2007). Analysis of the data found these behaviours were observed in 37 (71%) children across the educational stages, with 28 needing to adopt a horizontal position during the school day, 34 children seeking additional support when sitting or standing, and 12 children demonstrating both behaviours. Although the data set out in Table 25 shows these behaviours were noted in children in the different educational settings, they were reported for a higher percentage of children in SLD/PMLD provision. It is possible there may be a link to learning disability, but it could also be that in the other types of provision it is not so easy for a child have the opportunity to adopt this position, whether it is due to availability of space or the perception that this would not be an appropriate behaviour for the child to engage in. Greater numbers were reported to seek additional support, and although again it was reported in a higher proportion of pupils in an SLD/PMLD setting (18/21 children), this was also reported in 7/15 children in other specialist settings and 9/16 learners in a mainstream setting. Importantly these figures show that amongst the children in this study, a number with identified vestibular difficulties were observed to engage in these adaptive behaviours regardless of age. Examination of the data also found the presence of both behaviours was reported in 25 children in this study, and only one of these behaviours in a further 12 children, so overall, 37 (71%) children were reported to display at least one of these adaptive strategies, suggesting this is an important area to be considered when working with the child with CHARGE.

TABLE 25: PRESENCE OF BEHAVIOURS INDICATIVE OF ADAPTIVE RESPONSES TO VESTIBULAR AND PRORIOCEPTIVE DIFFICULTIES

	N=52	SLD/PMLD provision (N=21)	Mainstream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Does the child need to adopt a horizontal position at any time during the school day?	Yes 28 (54%)	16	5	7	8	7	7	6
	No 22 (42%)	5	9	8	4	7	4	7
	Don't know 2 (4%)	-	2	-	1	-	1	-
Does the child seek additional support for their position when standing or sitting?	Yes 34 (65%)	18	9	7	9	12	9	4
	No 17 (33%)	3	7	7	3	2	3	9
	Don't know 1 (2%)	-	-	-	1	-	-	-

5.6.7b Fine motor skills

The data obtained in response to the two questions on fine motor skills (see Table 26) showed fine motor development was an area where many of the cohort were reported to experience difficulty across the educational settings and also the educational phases, although for fewer children in KS4. The data shows all children in a mainstream setting were perceived to experience fine motor difficulties, and 11/15 in other specialist settings, in contrast to 12/21 children in an SLD/PMLD placement. This suggests these difficulties may be more noticeable where learners with CHARGE are educated alongside typically developing children, whilst learners in an SLD/PMLD provision may be working at a developmental level where difficulties with fine motor skills may not be so obviously apparent. These results were compared to the participants' earlier responses in the questionnaire where they were asked if they felt the child they were reporting on had poor body awareness and coordination difficulties. It was noted that 39/43 children reported to experience difficulties in this area were subsequently also reported to experience either fine motor difficulties or poor pencil skills/hand-writing skills, or both. Respondents were not asked to comment on whether they felt the child experienced poor or delayed gross motor skills (since it was felt teachers of older children within this cohort may not be aware of the presence of delayed gross motor skills earlier in the child's development). This is however an aspect of development where a delay is often reported in literature (see 2.3.1) and a factor that can potentially cause difficulties with fine motor skills which are dependent on well-developed gross motor skills (Sylva and Lunt, 2002). Importantly it is noted this is an aspect not addressed in literature relating to the CHARGE population or indeed the broader deafblind

TABLE 26: RESPONSES TO QUESTIONS RELATING TO FINE MOTOR SKILLS

		SLD/PMLD provision (N=21)	Mainstream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Does the child experience fine motor difficulties?	Yes 39 (75%)	12	16	11	11	12	9	7
	No 11 (21%)	7	-	4	1	1	3	6
	Don't know 2 (4%)	2	-	-	1	1	-	-
Does the child demonstrate poor pencil skills/ hand- writing skills?	Yes 40 (77%)	18	11	11	10	12	11	7
	No 10 (19%)	2	4	4	1	2	1	6
	Don't know 2 (4%)	1	1	-	2	-	-	-

population and may be a hitherto unidentified aspect specific to learners with CHARGE.

5.6.8 General comment: The Child

Within this section a range of anomalies and characteristics were identified that potentially can impact upon development and learning, as well highlighting some areas of potential strength and greater need. Overall, every set of scores obtained for each skill described using a Likert scale covered a range of scores and is an illustration of the variation that may be found in different individuals with CHARGE. Thus it is important to acknowledge this whilst continuing with further analysis. A summary of the data obtained through use of a Likert scale is set out in rank order according to the total score obtained in Appendix 8, Table (xiv). The results show that for the cohort as a whole, most skills scored over the midway point (156), but only the top 4 total scores indicated this was a skill found difficult or very difficult overall. These figures are closely reflected in the overall mean average scores obtained for the responses given for each skill described. In contrast to the mean average, the mode average scores showed the most common response given was that the child found it difficult (a score of 4) or very difficult (a score of 5) for all skills that obtained a total score above the midway point (16/21 skills examined), with the exception of identifying solutions to simple problems and being able to learn and play independently. For the 5 lower scores the mode more closely matched the mean average with the exception of establishing real, concrete concepts, where the most common response was that the child was perceived to find this very easy.

Placing the total scores in rank order provides important insights into the learning of children with CHARGE; it shows that of the skills set out in the questionnaire, children in this cohort were perceived to have the most difficulty with skills related to social and emotional development with the exception of establishing relationships with adults in the school setting. These rankings also show the variation in developing a set of skills, for example, certain skills related to conceptual ability were perceived to be some of the most difficult, whilst another (establishing real, concrete concepts) was regarded as one of the easiest for these children. This provides evidence for the teacher who may need to recognise that learners with CHARGE are indeed likely to demonstrate a mixture of both strengths and needs within each developmental area (as discussed in 2.3.3), and there is consequently a risk of the development of 'splinter skills' as suggested by an author in a report analysed in Cycle 1 of the investigation.

The data showed there was some variation according to the type of educational setting the child was placed in. Certain skills were perceived to be more difficult for children in an SLD/PMLD setting, for example, in developing relationships and empathising with their peers, recalling past events and predicting future events, understanding abstract concepts, and greater use of concrete communication modes, and it is necessary to acknowledge cognitive ability may be an influencing factor in this. What was surprising was the number of skill areas where little difference was noted in the degree of difficulty a child was perceived to experience across the different types of educational settings, including the transference and generalisation of skills, coping without a regular routine/unexpected changes,

independent learning and play, self-help functional life skills and fine motor difficulties. Although it was recognised there is a clear link between cognitive impairment and the development of some skills, this data also suggests that in addition to the child's sensory impairments, other factors are also likely to be involved. These may be internal factors (such as executive dysfunction) but furthermore external factors are likely to be involved, such as the type of educational provision itself and its educational philosophy and curriculum, but also the skills and knowledge of staff in each setting, as well as each individual respondent's understanding of the questionnaire. For example, children in an SLD/PMLD setting were perceived overall to find it easier to anticipate what is to happen next when appropriate cues are provided, but it might be argued that in such a setting there would be greater emphasis on ensuring such cues are made available as part of the general school ethos. Again this is an important finding of this study; although children across the different educational settings were perceived to experience similar levels of difficulty with the majority of skills examined, the educational response is likely to differ dependent on the type of provision the child attends.

It was also noted that there was often little variation across the different educational phases, so that although a few skills were perceived to be easier for older children (e.g. recalling past events and predicting future events, organising oneself, use of problem-solving), most data did not vary greatly according to age. This is important, since it suggests that although the individual needs of each learner are likely to alter over time, it must not be assumed that particular issues encountered will be age-related.

It has been acknowledged the information gathered through the use of a survey such as this are the perceptions of respondents that will be influenced by each person's past experience, knowledge base, life view and so forth and likely to affect the answers given. So, for example, it was noted that overall children in EY/KS1 were perceived to find it easier to develop genuine friendships with peers than children in the other educational phases. This may be a reflection of the perception of what a friendship is for younger and older children, and relationships between older peers may be expected to be more complex in nature, and therefore more difficult to achieve.

5.7 Strategies

Within this section of the questionnaire responses were gathered on how helpful given strategies, presented as an inventory, were perceived to be in supporting the child for whom the questionnaire was completed, with respondents additionally asked to list others also identified during their work. The data was collated to show the number of participants who considered each of the given strategies to be helpful in supporting the child, across both the different educational phases and types of provision.

The structure of the questions within this section meant that where a given strategy was not perceived to be helpful by the respondent, it was not possible to identify whether it had been employed but was regarded as unhelpful or whether the strategy had not been implemented. On reflection, it is recognised a fuller picture could have

been obtained if respondents had been asked whether they had attempted to implement the given strategies, in addition to whether they found them helpful (in a similar way to the question form used to gain information on assessment (see 5.8). Again it was also acknowledged that responses given would be influenced by the respondents' own perceptions of the question, what a strategy entails, and also what is 'helpful' in supporting the child with CHARGE.

5.7.1 Sensory responses: Strategies

The data obtained within this section is presented in Table 27. It shows the strategies most often reported to be helpful involved management of the environment in response to the child's sensory needs through the use of visual and auditory aids, and also differentiating materials and activities. These were both considered helpful by 88% of respondents overall, and for similar percentages of children in the different types of educational provision and phases. Fewer respondents (67%) found the development of profiles of functional use of vision and hearing helpful, despite the possibility of the information made available through these being used to make these former two strategies more accurate and effective. The development of functional visual/auditory profiles that could be used to effectively inform the differentiation and delivery of activities is advocated for deafblind children (e.g. Eyre, 2000), and also for children with an impairment of only one of these senses, this being reflected in tools developed to support this process (e.g. Bell, 2013; Reed-Beadle et al., 2011). It was noted that although 35 (67%) of respondents overall found this strategy helpful, this was reported for only 6/16 (38%) children in a mainstream setting in contrast to 15/21 (71%) in an SLD/PMLD setting and 14/15 (93%) in other specialist settings.

TABLE 27: NUMBERS OF RESPONDENTS WHO CONSIDERED GIVEN STRATEGIES TO BE HELPFUL WITH REGARD TO THE CHILD'S SENSORY NEEDS

Strategy	Number reporting strategy as helpful (N=52)	SLD/ PMLD provision (N=21)	Main-stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Differentiating materials/activities in relation to child's sensory needs	46 (88%)	19	13	14	10	13	11	12
Use of visual/auditory aids	46 (88%)	17	15	14	12	11	10	13
Activities planned/implemented in relation to child's sensory integration difficulties	37 (71%)	17	9	11	8	12	9	8
Developing profiles of functional use of vision and hearing	35 (67%)	15	6	14	10	8	8	9
Appropriate behaviour management with recognition of impact of sensory integration difficulties	31 (60%)	16	5	10	6	9	9	7
Access to a safe rest area	25 (48%)	16	5	4	5	9	5	6
Undertaking an environmental audit in relation to child's sensory needs	23 (44%)	10	5	8	7	5	5	6
Activities to support the development of haptic perception (active use of touch)	13 (25%)	8	2	3	3	3	3	4

A similar pattern was also noted for a number of other strategies listed within this section, including the use of activities planned and implemented in relation to the child's sensory integration difficulties. It is possible the children in mainstream provision were less in need of these types of support but this is contradicted by the data presented in 5.6.1 which shows teachers perceived different behaviours that may indicate sensory integration difficulties to be present in most if not all of those children in a mainstream setting. It is therefore likely this is an example of how factors outside the child might also influence whether a strategy is employed or deemed to be helpful or otherwise, including the ethos of the educational setting, the curriculum, the knowledge and experience of staff, together with the practicalities of implementing certain strategies in a particular setting. This may also be reflected in the strategy whereby provision is made for access to a safe rest area, regarded as being helpful for 16/21 children in SLD/PMLD placements in contrast to 5/16 children in a mainstream setting and 4/15 children in other specialist settings. Further comparison of the data showed this was made available for all but one of the children in an SLD/PMLD setting where high levels of fatigue were reported and 4/6 children in other specialist settings, but only 4/9 children in a mainstream setting. This reinforces the question of whether this is linked to factors within the child (e.g. Are sensory breaks and a rest area required more often for children more likely to attend a SLD/PMLD setting?), or whether it is a need more readily identified, or a facility more easily and freely made available in the SLD/PMLD setting than mainstream provision.

Structured support for the development of haptic perception (active use of touch) was the strategy fewest respondents considered helpful in relation to a child's sensory needs, reflecting the views of 34 (65%) respondents who considered the child reported on did not show a reluctance to use touch to explore or to touch certain textures (see 5.5.1). This is a further indication this is not perceived to be such an issue for this cohort of children as suggested in literature on CHARGE (e.g. Davenport and Hefner, 2011).

Within this section, and subsequently for communication, social and emotional development, and orientation and mobility, participants were asked whether use of an environmental audit was considered helpful. This will be discussed later in 5.8.1.

5.7.2 Communication: Strategies

Of the strategies listed in the inventory for this section (see Table 28) the strategy reported to be most helpful was the use of a Total Communication approach (by 48 (92%) respondents). 'Total Communication' is a term coined by Holcomb in 1967 originally in relation to the education of deaf children. As a philosophy it considers the involvement of a combination of different modes of communication depending on the particular needs of the child with the expectation that teachers would use the communication methods most appropriate for a particular child at a particular stage of development (Solit et al., 1992). It was noted that the 4 children for whom this was not reported to be helpful were all in mainstream provision (but different educational phases). It may be this strategy is less helpful for these learners but it may also be

TABLE 28: NUMBERS OF RESPONDENTS WHO CONSIDERED GIVEN STRATEGIES HELPFUL WITH REGARD TO COMMUNICATION

Strategy	Total where strategy reported as helpful (N=52)	SLD/ PMLD provision (N=21)	Main-stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Use of a Total Communication approach	48 (92%)	21	12	15	13	13	10	12
Ensuring access to a peer group with an appropriate level of communication skills	42 (81%)	16	15	11	11	11	11	9
Providing a sensitive communication partner who is able to recognise and respond to the child's attempts to communicate	35 (67%)	18	7	10	9	9	9	7
Development and implementation of an individual communication programme	26 (50%)	14	3	9	6	5	7	7
Undertaking an environmental audit in relation to the child's communication needs	22 (42%)	13	2	7	6	6	7	5
Other:								
Use of task plans (as part of Total Communication)	1		1				1	
Within the child's IEP communication identified as a top priority	1			1			1	
Motivational activities	1	1				1		
Use of an iPad & communication apps	1	1			1			

that teachers in the mainstream setting are less aware of this approach, or insufficiently resourced to implement it. Where a Total Communication approach was felt to be helpful participants were asked to identify what this included (the responses to this question also being used to check the consistency of responses as set out in 3.6.9).

The results (See Appendix 9, Table (xv)) showed little variation according to age but clear differences according to the type of educational placement attended by the child. For example, speech and printed word were those most frequently used for learners in a mainstream setting whilst greatest use was made of touch cues, object cues and objects of reference, symbols/drawings and also photographs for children in an SLD/PMLD setting. The data illustrates the difficulty of achieving shared understanding between respondents, for example, with touch cues; within the deafblind and learning disability fields these would be described as specific movements or touch made on a child's body to communicate something about people, an activity, or a place (e.g. Demchak et al., 2002; Bruce, 2014). It would therefore perhaps be anticipated that most children where such use was reported would be in an SLD/PMLD setting where teachers were more likely to have knowledge and understanding of this approach. The use of touch cues reported for 3 learners in a mainstream setting might be less anticipated, and whilst it is possible touch cues were being used as described within the deafblind and learning disability fields, it is also possible that a different interpretation of the term has been made to include touch being used in a less ordered or formal way to support the child's communication or to gain their attention. Further analysis showed the number of

modes used per child within a Total Communication approach ranged from 2-7 with an overall average of 4.75 per child (Table 10, Appendix (xvi)). It was, however, evident that on average a higher number of communication modes was used with children in an SLD/PMLD setting (5.0) compared to children in other specialist settings (4.7) and mainstream settings (4.3), although there was little variation across the age ranges. This again illustrates the large number of different communication modes being used on average with the children within this cohort as discussed previously in 5.6.2b; in the future it may be prudent to examine more carefully the reasons for choosing to employ particular communication modes and the potential benefits or negative impact of using such a large number of different modes with each child in the educational setting.

Ensuring the child has access to a peer group with appropriate levels of communication was considered helpful by 42/81% of respondents, more so than the provision of a sensitive communication partner (35/67%). Comparison of the data showed that, whilst it was considered helpful to provide a peer group with appropriate communication skills for a similar percentage of children in each of the types of educational settings, the provision of a sensitive communication partner was deemed helpful for a lower percentage of children in a mainstream setting. This could equate to the findings in 5.6.3 where children in an SLD/PMLD setting were perceived to experience greater difficulty developing genuine friendships with peers.

It was noted an individualised communication programme was perceived to be helpful for similar numbers of children across the age range and with 14 children in

an SLD/PMLD setting, 9 in other specialist settings, but only 3 in a mainstream setting. This may be interpreted in a number of ways; it may be that the children supported by other respondents had an individual communication programme but that it was not felt to be helpful or, that such a programme was not in place. It may also be an indicator that there is less requirement for such a programme for those children in a mainstream setting, but could again be a reflection of the skills and knowledge of staff, together with the ethos of the school.

5.7.3 Social and emotional: Strategies

Recognising it was considered helpful to have access to a peer group with appropriate communication skills has an additional implication when considered in relation to the responses obtained in this section (see Table 29) where it was found 39 (75%) respondents considered the use of a structured programme/activities a helpful strategy in supporting interactions with their peers (see 5.6.3). This implies that in addition to ensuring an appropriate peer group it is important to provide an additional level of structure and support to facilitate communicative exchanges and social interactions between the child with CHARGE and their peers. These findings can be compared to research undertaken in relation to deafblind learners, where Correa-Torres (2008) for example, reported three deafblind pupils studied were all more likely to have closer relationships with adults and less likely to interact socially with their peers, explaining that opportunities for social interaction are not necessarily readily available and there is a need for structured intervention to support the development of peer relationships. In addition to barriers created by the child's

TABLE 29: NUMBERS OF RESPONDENTS WHO CONSIDERED GIVEN STRATEGIES TO BE HELPFUL WITH REGARD TO SOCIAL AND EMOTIONAL DEVELOPMENT

Strategy	Total where strategy reported as helpful (N=52)	SLD/ PMLD provision (N=21)	Main-stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Key people identified to develop secure relationships with child	47 (90%)	19	13	15	11	12	11	13
Use of a structured programme/activities to support interactions with peers	39 (75%)	17	10	12	8	11	10	10
Support to learn how to initiate/sustain social interaction sequences	33 (63%)	13	8	12	8	7	9	9
Implementation of a structured programme to develop the child's understanding of emotions	31 (60%)	13	10	8	7	8	7	9
Use of a defined programme to manage stress and anxiety levels	20 (38%)	14	1	5	3	6	8	3
Undertaking an environmental audit in relation to the child's social and emotional needs	17 (33%)	12	-	5	3	6	5	3
Other:								
Consistent calm approach	1			1				1
Awareness of potential triggers	1			1				1

sensory impairments, 33 (63%) reported they found it helpful to intentionally provide support to the child to learn how to initiate and sustain social interactions; a strategy that may help address some of the difficulties arising from executive dysfunction which include difficulty initiating actions (Cooper-Kahn and Dietzel (2008). The findings also showed over half of respondents (31/60%) considered it helpful to focus on supporting the understanding of emotions. A comparison of data showed that whilst this was considered helpful for 20/37 children reported earlier to understand or express their own emotional state (see 5.6.3) it was also perceived to be a helpful strategy for 8/17 children earlier reported not to have difficulty in this area and it is therefore possible the implementation of such a programme was supporting these 8 children in finding it easier to understand or express their own emotions. Further study would be beneficial to explore the content of such a programme.

The data shows that again the employment of the given strategies was spread across the educational phases. This was also apparent across the different types of educational provision with the exception of the use of an environmental audit (to be discussed later in 5.8.1), and the use of a defined programme to manage stress and anxiety levels. The latter was considered to be a helpful strategy predominantly for children in an SLD/PMLD setting and for only 1 child in a mainstream setting. When compared to those children reported earlier in the questionnaire to experience high anxiety or stress levels (See 5.6.1) it showed such a programme was reported helpful for:

- 12/14 children in an SLD/PMLD setting
- 3/7 children in other specialist settings

- 1/10 children in a mainstream setting

This can be interpreted in different ways: it may suggest such a programme is found to be more helpful for pupils most likely to attend an SLD/PMLD provision, but may also indicate such a programme is less likely to be made available in other settings (and particularly mainstream). This may be a consequence again of the individual characteristics of the children and also staff working in the different settings together with the ethos of the school, but may also be a direct result of the practicality of delivering such a programme in a mainstream setting.

5.7.4 Conceptual ability: Strategies

The responses obtained within this section are presented in Table 30 and show the most helpful strategies identified from the inventory with regard to conceptual ability involved the use of real (concrete) cues and direct experiences within the learning context itself. This may be a response to the perceived difficulty in developing abstract concepts reported earlier in the questionnaire (see 5.6.4) and also suggests an understanding amongst the respondents that the development of concepts is dependent upon each child's experiences. As mentioned earlier, Miles and Mcletchie (2008) describe concepts as those ideas which give meaning to our world. Although they do not specifically state that use of concrete cues is important in supporting the formation of concepts in deafblind learners, they do explain the need for direct experience and participation. An example given is supporting a child to develop the concept of 'on and off' through direct experience of a child getting on and off a swing (p.5) and this is an illustration of how a concrete element can support a deafblind

TABLE 30: NUMBERS OF RESPONDENTS WHO CONSIDERED GIVEN STRATEGIES HELPFUL WITH REGARD TO CONCEPTUAL ABILITY

Strategy	Total where strategy reported as helpful (N=52)	SLD/ PMLD provision (N=21)	Main-stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Use of concrete tools (e.g. real objects, photographs, symbols etc.)	46 (88%)	20	11	15	11	13	11	11
Provision of real experience provided within context	44 (85%)	19	11	14	8	14	11	11
Providing a balance between new and familiar activities	41 (79%)	19	10	12	10	10	10	11
Use of identified strategies to support the development of problem-solving skills	30 (58%)	15	7	8	7	9	7	7
Use of structured fading and pre-learning/post-learning to support learning	28 (54%)	11	10	7	5	6	8	9
Monitoring to identify the risk of the development of 'splinter' skills and responding	7 (13%)	3	1	3	-	1	3	3
Other:								
Use of role play	1		1				1	
Use of ICT	1		1				1	
Use of child's learning style	1	1					1	

child experiencing difficulties with developing abstract concepts through the use of concrete cues and real experiences. The fact that a large number of respondents in this investigation felt the use of real experiences were helpful in contributing to the concept development of this cohort of children with CHARGE suggests that this approach is perceived as important for learners with CHARGE as reflected in literature (See 2.6.3f).

Although it was noted the use of concrete tools was regarded as helpful for similar numbers of children across the age ranges, it was possible the developmental stage of the child might also be a factor, recognising that some older learners (e.g. with severe learning difficulties) might be functioning at an earlier developmental level. Thus further analysis was undertaken to explore the age ranges of the children within the different types of educational settings where this strategy was perceived to be helpful. The results set out in Appendix 9, Table (xvii), show the use of concrete tools was considered helpful for some children across the age ranges in each of the different types of setting. This may be linked to an earlier finding of this study (see 5.6.4) where the establishment of abstract skills was felt by teachers to be one of the most difficult skills identified in relation to conceptual ability, again for children across the age ranges and settings (although most difficult for children in an SLD/PMLD setting) and may be a reason why so many respondents felt the use of concrete cues and providing real experiences were helpful strategies.

Just over half of participants (28/54%) in this study reported they found the use of structured fading and pre-learning/post-learning to support learning a helpful

strategy. This strategy was identified within Cycle 1 of this investigation (hence being included in the questionnaire) but was not noted in the review of literature on CHARGE and is infrequently identified in literature relating to the wider deafblind population (e.g. it is mentioned by Murdoch et al., 2009). These findings indicate participants' responses were not based solely on knowledge that may have been obtained from literature, but were also derived from a willingness to share their own observations and experiences. If this argument is applied to the datum extracted which shows only 7 (13%) respondents felt it was of benefit to monitor and identify the risk of the development of splinter skills (discussed in 2.3.3) it suggests this may be an area where difficulties have not been observed, although contradicting the literature which reports individuals with CHARGE often demonstrate a combination of high and low potential features in different areas of their learning. Another argument may therefore be that there is a lack of awareness of this as an issue amongst teachers; for example, it is not discussed in the literature on the education of deafblind learners although Clark (2000) does caution teachers against teaching "splintered or fragmented skills" (p.91). Thus it is not possible to ascertain without further exploration in the future, whether this potential difficulty is not being recognised, effective strategies have not been identified, or this issue is not widely present within this cohort and thus a response not required.

5.7.5 Responses to routines and structure, and understanding of time and space: Strategies

In relation to this aspect, the findings (set out in Table 31) suggest that overall it was regarded as more helpful to consider how the overall day and activities were

TABLE 31: NUMBERS OF RESPONDENTS WHO CONSIDERED GIVEN STRATEGIES TO BE HELPFUL WITH REGARD TO RESPONSE TO ROUTINE AND STRUCTURE AND UNDERSTANDING OF TIME AND SPACE

Strategy	Total where strategy reported as helpful (N=52)	SLD/ PMLD provision (N=21)	Main-stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Use of a daily routine and structure implemented consistently throughout the day	48 (92%)	21	13	14	12	13	11	12
Structured support for transitions	42 (81%)	20	9	13	9	11	10	12
Ensuring activities have a clear beginning, middle and end	40 (77%)	18	11	11	9	10	10	11
Use of mini routines	33 (63%)	13	12	8	7	9	10	7
Individualised pacing (may include a reduced timetable)	31 (60%)	13	9	9	5	8	10	8
Use of concrete tools to support sequencing of an activity, transitions and daily routine	30 (58%)	17	3	10	6	7	10	7
Other: Preparing for possible changes ahead of time	1			1				1

structured and consistent routines implemented, than the use of concrete cues themselves. This reflects the data presented in 5.6.5 where the children were considered to find it easier to understand time and space, and to experience greater difficulty when routine and structures were either disrupted, or not established. The data presented in 5.6.5 also showed a very similar degree of difficulty was perceived to be experienced when there was no regular routine and structure, or where there were unexpected changes to those routines, by children across the different educational settings and phases. Despite this, a comparison of data showed it was reported to be helpful to provide structured support for transitions for a lower percentage of children in a mainstream setting (56%) than children in both an SLD/PMLD setting (95%) and other specialist settings (87%). In 5.6.5 substantial use of concrete cues was reported for many of the children within this cohort (3-6 concrete cues being used with 38 (73%) children) and, additionally, 46 (88%) respondents had also reported finding the use of object cues helpful to support concept development (see 5.7.4). It was therefore interesting to note that in comparison, only 30 (58%) respondents found it helpful to use concrete tools to support sequencing of an activity, transitions and daily routine (including use of a visual timetable, task plans and first/next symbols). The data showed these were reported to be helpfully employed for more children in an SLD/PMLD setting or other specialist setting than children in a mainstream setting which maybe a reflection of varying knowledge and skills of teachers in these different settings. These findings also lead to the question of what features of these cues are most significant to the child; it may be the tangible quality of the cue, but it might also be that the child benefits from a cue which is less transient in nature than speech or sign. Thus is it

possible such cues were less often reported helpful for pupils in mainstream settings where other alternatives (such as writing things down) may be as helpful.

5.7.6 Ownership of learning: Strategies

Following the analysis of the earlier responses given within this questionnaire (set out in 5.6.6) it was suggested that taking ownership of learning is often felt not to be easy for the children within this cohort; this is reflected in the high number of participants who reported the helpfulness of (thereby implying the use of) a number of strategies identified within this section (see Table 32). Identifying what is motivational for the child and employing a flexible approach that reacts to the child's changing needs and level of responsiveness were both regarded as helpful in encouraging ownership of learning by 44 (85%) respondents. These, together with the strategy of using the child's individual learning style to inform the development of activities (reported helpful by 41/79% of respondents), showed how helpful overall teachers found it to be led by the individual child. The importance of enabling the child to share control was reflected by 39 (75%) of respondents finding it beneficial to provide the child with regular opportunities to make choices.

A structured response to manage the impact of fatigue was reported to be a helpful strategy by 29 (56%) of respondents. These responses were compared to those given earlier (5.6.1) where 27 respondents had reported they felt the child showed high levels of fatigue; of these 27 teachers, 22 subsequently reported they felt a structured response to manage the impact of fatigue was helpful. Where teachers had earlier reported an issue with fatigue but did not then report a structured course

TABLE 32: NUMBERS OF RESPONDENTS WHO CONSIDERED GIVEN STRATEGIES HELPFUL WITH REGARD TO OWNERSHIP OF LEARNING

Strategy	Total where strategy reported as helpful (N=52)	SLD/ PMLD provision (N=21)	Main-stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Clear identification of what is motivational for the child	44 (85%)	20	12	12	10	12	11	11
Use of a flexible approach, responding to the child's changing needs and levels of responsiveness	44 (85%)	18	11	15	10	12	12	10
Clear identification of the child's individual learning style informing the delivery of activities	41 (79%)	18	10	13	9	10	11	11
Providing frequent choice-making opportunities	39 (75%)	19	10	10	9	9	12	9
Implementation of a structured programme to support self-help and self-organisational skills	37 (71%)	18	9	10	9	8	9	11
A structured response to manage the impact of fatigue	29 (56%)	13	8	8	10	7	7	5
Responding to the child's executive function difficulties	21 (40%)	14	3	4	2	6	9	4

of action to be helpful, it is not possible to ascertain whether either of the responses given were inaccurate, or a structured response had been tried but was found not to be helpful. Although such a programme was implemented with a higher percentage of children in an SLD/PMLD placement, it was noted as considered helpful for half of the children in both the other settings.

Within this section the term 'executive function' was used within a question, this being the only occasion it was directly used in the questionnaire. It is interesting to note fewer respondents (21/40%) reported it to be helpful to respond 'to the child's executive function difficulties' and this raises the importance of striving to ensure a shared understanding of the language being used and whether, if the question had been worded differently, the response rate would have altered. Nonetheless the level of response suggests there was some understanding of the term used. It was noted that most of the respondents reporting this to be a helpful strategy were commenting on a child within an SLD/PMLD provision which may indicate a greater awareness of executive dysfunction amongst teachers working in this setting since it impacts upon a child's cognitive performance (see 2.2.9 and 2.3.3). Additionally, this was only regarded as a helpful strategy for 2 children within EY/KS1, and although it is not possible to be certain, this may be linked to the development of executive function in the individual; executive functions develop from birth through to adolescence and beyond (Juric et al., 2013), but within this there are periods of acceleration in the development, one typically occurring between the ages of 5-8 years (De Luca et al., 2003). As children with CHARGE often demonstrate a delay in development (as discussed in Chapter 2, including 2.2.9 and 2.3.3) it is possible this period of

acceleration would also be delayed and therefore the potential impact of executive function difficulties may not be so evident within the early years.

5.7.7 Orientation and mobility, and fine motor skills: Strategies

5.7.7a Orientation and mobility

Responses within this section set out in Table 33 show the strategies most often reported to be helpful relate to gross motor skill development and the provision of regular opportunities to engage in large movements. The implementation of such strategies may reflect the perception that 39 (75%) children in this cohort experienced difficulty coordinating and organising their body (see 5.6.7a), with a comparison of the data revealing these strategies were considered helpful for 40/43 children earlier reported to have poor body awareness and coordination difficulties. Further examination showed these strategies were felt to be helpful by teachers working with a child with CHARGE across the key stages and within the different types of provision; it was noted, however, that whilst similar numbers of children in specialist school settings were also reported to benefit from the use of large movement play activities, the helpfulness of this strategy was reported for only 4 children in a mainstream setting and also 4 children in KS4. The data suggests learners with CHARGE may need access to such opportunities in the educational environment regardless of their age, but how this is provided may be influenced by the type of educational setting the child attends. This has implications for the delivery of the curriculum since as a child progresses through the key stages they are required to spend much more time seated, making it more difficult for the child to

TABLE 33: NUMBERS OF RESPONDENTS WHO CONSIDERED GIVEN STRATEGIES TO BE HELPFUL WITH REGARD TO ORIENTATION AND MOBILITY

Strategy	Total where strategy reported as helpful (N=52)	SLD/ PMLD provision (N=21)	Main-stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Providing regular opportunities to engage in large movements	42 (81%)	17	15	10	11	11	11	9
Structured activities to support the development of gross motor skills and body awareness	39 (75%)	18	11	10	11	10	8	10
Use of specialist/supportive seating	32 (62%)	12	10	10	13	9	5	5
Implementation of a fine motor skills programme	31 (60%)	15	7	9	9	10	8	4
Provision of alternative recording methods (e.g. scribe, use of IT, etc.)	29 (56%)	9	9	11	6	8	7	8
Use of large movement play activities	28 (54%)	15	4	9	7	7	10	4
Implementation of a pencil skills programme	21 (40%)	10	7	4	6	7	6	2
Undertaking an environmental audit in relation to child's orientation and mobility needs	21(40%)	10	1	10	7	4	5	5
Other:								
Touch typing	1		1			1		
Long cane training	1			1	1			
Learning sighted guide techniques	1			1	1			
Regular physiotherapy input	1	1			1			
1:1 mobility sessions	1			1				1

engage in regular large movements that so many of the respondents found to be helpful. Although use of specialist/supportive seating was regarded as being helpful by 32 (62%) respondents, it was noted that 22 of these children were in the earlier educational phases. This is of interest in light of Brown's argument (2003a&b) that as individuals with CHARGE grow older the impact of their impaired sensory systems (particularly the vestibular sense) can become less apparent, but that they continue to experience difficulties standing or sitting unsupported and will continue to benefit from supportive seating.

Some additional strategies identified (long cane training and use of sighted guide techniques) indicate likely specialist input from a habilitation (mobility) officer. It is recognised more detailed or insightful information might have been obtained regarding the child's orientation and mobility if responses had been sought from other professionals such as an occupational therapist or a habilitation officer working to support a child in this area. This will be discussed further in 5.9 where the data will show input from a habilitation officer was provided for only 10/52 children within this cohort.

5.7.7b Fine motor skills

Although 31 (60%) respondents reported they considered the implementation of a fine motor skills programme to be helpful, when compared to previous findings (5.6.7b) it was noted this was considered helpful for only 23/39 children perceived to experience difficulty with fine motor skills. Whilst 21 (40%) respondents felt a pencil

skills programme had been helpful, this included only 19/40 children earlier reported to have poor fine motor/pencil skills. This may indicate a lack of response to this need or a difficulty in developing a suitable programme but may also be related to the 56% of respondents who found it helpful to provide alternative recording methods (with one respondent specifically identifying the introduction of touch typing), thus the provision of alternative recording methods reducing the need for a pencil skills programme if the child is recording their work in an alternative way.

5.7.8 General comment: Strategies

The strategies considered most helpful in supporting the learner with CHARGE often reflected a response to the internal factors identified previously in 5.6.1 and those skills perceived to be more difficult as identified in the section on the child (5.6). This suggests teachers are generally acting upon those difficulties identified and employing particular strategies regarded as a helpful response. An exception was found where 39 (75%) children were reported to have difficulty with fine motor skills (5.6.7b), but respondents subsequently reported a fine motor skills programme as being helpful for only 23 of these 39 children; similarly a pencil skills programme was deemed helpful for only 19/40 children where a difficulty with pencil/writing skills was previously identified (again 5.6.7b). There are a number of possible reasons for this, one being that the respondent may simply not have ticked the appropriate boxes when responding. It is also possible, however, that a programme had been implemented for other children but was found not to be helpful, that a pencil skills programme was not regarded as beneficial and other approaches were adopted, or

also that the issue was not identified as significant and therefore did not require a response.

It was found that the implementation and helpfulness of strategies did not vary greatly across the ages of the children, and only small variations were noted across educational phases. The main exceptions were in the provision of a defined programme to manage stress and anxiety levels, and also in the area of orientation and mobility where the use of specialist/supportive seating, large movement play activities, a fine motor skills programme and pencil skills programme were reported to be helpful for a relatively smaller number of children in the later educational phases. It is possible these strategies are not so necessary for the older learner with CHARGE but this would contradict the high numbers of children in KS3 and KS4 where behaviours likely to require a response were noted (e.g. needing to seek additional support when standing or sitting) or who were reported to experience fine motor difficulties and demonstrate poor pencil/handwriting skills. Therefore the opinion of whether specific strategies implemented in response to these issues would be regarded as helpful is likely to also be influenced by other factors. One factor that must be recognised is the impact of teacher perceptions and understanding what assumptions or decisions might be made in consequence of these, for example, that there is not a need to focus on the development of gross and fine motor skills in older children, or conversely that a younger child needs additional time to develop their fine motor skills without direct intervention. Importantly, the findings overall suggest potential strategies should not be either implemented or dismissed on the grounds of a child's age, but rather according to their needs.

In contrast, it was found there was some variation in the data according to the type of educational provision the child was placed in. An examination was undertaken of the percentage of children within each type of educational provision where each strategy was perceived to be helpful; it was then noted where there was a difference of more than 25% across the different categories (SLD/PMLD setting, other specialist settings and mainstream settings). This showed a number of strategies more often perceived to be helpful when employed in the specialist settings than in a mainstream setting, these being set out in Table 34.

TABLE 34: STRATEGIES MORE OFTEN PERCEIVED AS HELPFUL BY RESPONDENTS WORKING IN A SPECIALIST SETTING THAN RESPONDENTS IN A MAINSTREAM SETTING

- Activities planned/implemented in relation to child's sensory integration difficulties
- Developing profiles of functional use of vision and hearing
- Appropriate behaviour management with recognition of impact of sensory integration difficulties
- Providing a sensitive communication partner who is able to recognise and respond to the child's attempts to communicate
- Development and implementation of an individual communication programme
- Use of concrete tools (e.g. real objects, photographs, symbols etc.)
- Provision of real experience provided within context
- Structured support for transitions
- Use of concrete tools to support sequencing of an activity, transitions and daily routine
- Implementation of a fine motor skills programme
- Use of large movement play activities

An additional number of strategies were more often perceived to be helpful when employed in an SLD/PMLD setting than the other specialist settings or a mainstream setting, as set out in Table 35.

TABLE 35: STRATEGIES MORE OFTEN PERCEIVED AS HELPFUL BY RESPONDENTS WORKING IN AN SLD/PMLD SETTING THAN RESPONDENTS IN OTHER SPECIALIST SETTINGS OR A MAINSTREAM SETTING

- Access to a safe rest area
- Activities to support the development of haptic perception (active use of touch)
- Use of a defined programme to manage stress and anxiety levels
- Use of identified strategies to support the development of problem-solving skills
- Providing frequent choice-making opportunities
- Responding to the child's executive function difficulties

It may be argued that the implementation of given strategies might be more necessary for children placed in specialist settings for as explained in 5.2, it was anticipated the most able children were more likely to be found within a mainstream setting, and conversely those children with the greatest learning difficulties would be more likely to attend an SLD/PMLD provision (although this could not be guaranteed given the current inclusive model of education), and thus it is possible a child's learning disability influences the choice of strategies to be used. Overall, the similarity between a higher rate of occurrence of perceived difficulties a child experiences and the number of respondents reporting the helpfulness of strategies likely to be employed in response to those difficulties, indicate the use of strategies are influenced in part by the child's internal factors and strengths and needs. Significantly though, these findings show that although a similar degree of difficulty experienced with particular skills was reported for children across different educational placements, the use of some corresponding strategies were not regarded as so helpful for those children in a mainstream setting. For example, within the section on 'Routine and Structure', similar difficulties were reported in coping

without regular routines and structure, and on average children in mainstream and specialist settings other than an SLD/PMLD placement, were perceived to experience greater difficulty anticipating what was to happen next when appropriate cues were not provided; yet the provision of structured support for transitions was viewed as far less helpful for children in mainstream settings and even less so, the use of concrete tools to support these transitions.

As explained previously, the identification of a child's strengths and needs, and the strategies implemented in response will be dependent upon teacher knowledge, experience, skills and perceptions and, these findings suggest, the type of educational placement the child attends will also be an influencing factor on the strategies implemented. This suggests the knowledge and skill set of staff, the ethos of school, the perceptions of what might be appropriate for the child and the practicalities of how a given strategy can be delivered within the school day and curriculum are also all likely to influence which strategies might be regarded as helpful and therefore implemented.

Some of the strategies named within the inventories provided were quite specific, for example 'Providing regular opportunities to engage in large movements' whilst others were less so, for example, 'A structured response to manage the impact of fatigue'. Future study to examine in more detail the elements of such a strategy that are found to be helpful in supporting the learner with CHARGE are likely to be more informative for the educational practitioner.

The inclusion of strategies in the inventories provided was informed by the knowledge gained from the literature review and findings from Cycle 1 of this investigation. Those strategies regarded as helpful by over 50% of respondents are set out in rank order in Appendix 9, Table (xviii). These strategies address different areas of learning and development and suggest the importance and need to maintain a holistic overview when planning and implementing an educational programme for the learner with CHARGE and to ensure against a narrow focus on any given area. It is important to also consider those strategies less often reported as helpful by respondents. There was a relatively small number regarded as being helpful by 45% or less of respondents, these being set out in Table 36.

TABLE 36: THOSE STRATEGIES FROM INVENTORIES REPORTED TO BE HELPFUL BY LESS THAN 45% OF RESPONDENTS

Strategy	Number where reported to be helpful (n=52)
Undertaking an environmental audit in relation to child's sensory needs	23 (44%)
Undertaking an environmental audit in relation to the child's communication needs	22 (42%)
Responding to the child's executive function difficulties	21 (40%)
Implementation of a pencil skills programme	21 (40%)
Undertaking an environmental audit in relation to child's orientation and mobility needs	21(40%)
Use of a defined programme to manage stress and anxiety levels	20 (38%)
Undertaking an environmental audit in relation to the child's social and emotional needs	17 (33%)
Activities to support the development of haptic perception (active use of touch)	13 (25%)
Monitoring to identify the risk of the development of 'splinter' skills and responding	7 (13%)

It is interesting to note that, excluding the use of environmental assessments (to be discussed in 5.8.1 below), for each of these strategies more children were perceived to experience a difficulty in the area each strategy may address than the number where it was reported to be a helpful strategy. For example, 40 (77%) of respondents considered the child demonstrated poor pencil/handwriting skills, but only 21 (40%) of respondents felt the potential corresponding strategy (implementation of a pencil skills programme) was helpful. It may be these strategies have been tried and found not to be of benefit, but there is also a possibility that some may not have been implemented, perhaps because the teacher has recognised the difficulty but does not consider such action would be the most effective response, as a consequence of them not being deemed necessary, or a lack of knowledge and understanding of the impact CHARGE may have on learning and development. For example, 88% of respondents found it useful to differentiate materials/activities and make use of visual and auditory aids, both of which are well-known and well-established strategies in the education of the wider deafblind population and children with single sensory impairments, but other aspects may not be so well-known or require a different emphasis (such as addressing the presence and impact of executive dysfunction). It is also possible (or even likely) that appropriate strategies have not yet been identified to effectively address some of the difficulties encountered by children within this cohort and potentially the wider CHARGE population. For this reason it is also important to acknowledge those additional strategies identified by one or few respondents, as these could be further strategies that future investigation may discover would be beneficial to employ more widely in the education of learners with CHARGE.

5.8 Assessment

As discussed in 2.4, assessment is important to support the gathering of information about a child's strengths and needs, learning style etc., these being aspects focused upon by some of the research questions. Therefore assessment was also addressed within the questionnaire with respondents asked to identify those types of assessments undertaken or made available to the school in relation to the child's education, and to then record how helpful they perceived these assessments to be (with the responses set out in Table 37).

TABLE 37: FREQUENCY OF TYPES OF ASSESSMENT UNDERTAKEN OR MADE AVAILABLE AND LEVEL OF EFFECTIVENESS REPORTED

Type of assessment	Number of children where assessment was provided (N=52)	Effectiveness of assessments as reported by respondents		
		Very helpful/helpful	Neither helpful/unhelpful	Unhelpful / very unhelpful
Functional use of hearing	46 (88%)	41	4	1
Functional use of vision	41 (79%)	38	3	-
Communication	41 (79%)	36	4	1
Seating	34 (65%)	29	5	-
Sensory integration	32 (62%)	29	3	-
Orientation and mobility	27 (52%)	24	2	1
Eating and drinking	24 (46%)	19	4	1
ICT	22 (42%)	15	5	2
Hand function	14 (27%)	9	4	1
Other:				
MSI Curriculum	2	2	-	-
Spoken/sign language	2	2	-	-
Cleft palate	1	1	-	-

Comparison of data showed an assessment of functional use of vision had been undertaken for 39/48 children where a visual impairment was identified and an assessment of functional use of hearing undertaken for 44/49 children where a hearing impairment was identified. It was also interesting to note a number of children had received an assessment of functional use of vision or hearing where these sensory impairments had not been identified. This may suggest an element of checking whether there were issues irrespective of the clinical information made available and/or recognition of the interplay of the senses and how other sensory impairments can have an impact on the child's ability to use their vision or hearing effectively.

The importance of undertaking assessments of functional use of vision and hearing is well-documented for deafblind learners to discover how a child is using their residual vision and hearing, how they use their different senses together, and the impact of different environments; all of this helping to build up a detailed profile of functional use of vision and hearing that can be used to inform the planning and implementation of the curriculum (e.g. Eyre, 2000; Sense, 2002; Murdoch et al., 2009; Nelson et al., 2010). The need to undertake functional visual and hearing assessments is also advocated by practitioners working with children with these single sensory impairments and the range of tools developed to support this is an indicator of the importance placed upon them (e.g. functional vision: Southwell, 2003 and Bell, 2013; functional hearing: Reed-Beadle et al., 2011 and NDCS 2013).

Within this study the majority of respondents who reported such assessments had been undertaken had perceived them to be helpful or very helpful. Comparison of this data, however, with that obtained earlier in the questionnaire (5.6.1) showed fewer respondents (35/67%) had found the development of profiles of functional use helpful, suggesting there are cases where the information from these assessments was either not used or was not beneficial in supporting the development of a profile of functional use of vision and hearing. Further examination of what aspects were most helpful would be beneficial and also the reasons why some respondents had reported they felt these assessments were unhelpful. Additionally it would be interesting to explore how successful functional assessments are undertaken that are able to effectively inform the planning and implementation of an educational programme. Communication assessments, as widely advocated in the education of deafblind learners (e.g. Miles and Riggio, 1999; Pease, 2000; Janssen and Rødbroe, 2007; Nelson et al., 2010), were also reported to have been undertaken for a large number children within this study (79%) and a further 3 additional assessments specifically focused on language (either spoken language or sign language) were also reported.

Assessments in relation to sensory integration and seating, reported as having been undertaken for 34 (65%) and 32 (62%) children respectively, are aspects not often specifically addressed in literature relating to the education of deafblind learners in general, although Hodges (2000) identified the need for provision of supportive seating to ensure physical stability that presumably would necessitate a seating assessment. When comparing this data to earlier responses presented in 5.7.1, it

becomes clear that although 26/37 children in this study where a planned sensory integration programme was considered helpful had received a sensory integration assessment, this was not reported for the remaining 11 children. If it can be assumed respondents completed the questionnaire reliably (supported by the check for reliability outlined in 3.7.9), these figures suggest that although the programme was felt to be helpful, a number of children had a sensory integration programme established without information from a sensory integration assessment. Whilst practitioners may recognise and acknowledge the impact of sensory integration difficulties on a child's learning, only those therapists who have the training and expertise in sensory integration dysfunction (sensory processing disorder) are qualified to undertake such an assessment and develop a remediation programme (Hampson, 2013). This raises an important point about the need for collaborative working, drawing on expertise from different practitioners, and ensuring access to the appropriate specialists.

Eating and drinking assessments had been undertaken for just under half of this cohort (24/46%) although 5 respondents had not found them to be helpful. Given that one of the major diagnostic characteristics for CHARGE involves the IX/X cranial nerve pairings likely to result in swallowing problems with including aspiration, this is an aspect that is likely to need attention in the educational setting around mealtimes. This is also supported by the research of Dobbelsteyn et al. (2008) who reported eating and drinking difficulties in >80% of individuals with CHARGE, and Deuce et al. (2012) where 84% of the investigation cohort (a total of 44 children) was reported to experience eating and drinking difficulties.

An overall average of 5.5 assessments was reported to have been undertaken per child, although it was noted the average for children in SLD/PMLD schools was higher at 5.7, and similar in other specialist provision (5.6), but slightly lower for children in a mainstream setting (4.8). Only 2 children were reported not to have had any assessments undertaken within the educational setting, both within a mainstream setting. The average number of assessments taken for the remaining children in a mainstream setting was 5.4, thus being very similar for children in other placements. This may be an indicator of the complexity of need of the children included in this survey and that teachers are likely to need additional information from a range of different perspectives to inform their planning and delivery of the curriculum. It also further illustrates the need for collaborative working as many of these assessments are likely to have involved practitioners with a particular specialism (e.g. specialist teachers, therapists, etc.).

Although a large proportion of assessments undertaken were felt to be helpful or very helpful, there was a notable number for each type of assessment where it was felt to be at best neither helpful nor unhelpful, but also considered unhelpful by some respondents. This suggests that for some teachers the outcomes of the assessments were perceived not to support and inform the planning and delivery of the curriculum for the child with CHARGE with whom they were working. This may reflect the difficulties in accurately assessing children with CHARGE as noted in literature (see 2.4) that might result in assessments being unsatisfactorily completed, possibly as a consequence of the assessor having a lack of experience in assessing a child with

CHARGE. It may also be that the type of assessment undertaken was not appropriate for the child and the teaching situation.

These figures showed overall a range of assessments had been carried out for children within this study. It was interesting to note that only a very small number of respondents reported an assessment in relation to the child's overall learning and the curriculum. It is possible this was under-reported as it was not an aspect specifically identified within the questionnaire. However, it is to be assumed that as an essential part of good practice, this type of assessment would be undertaken as part of the teaching process (recording a baseline, monitoring progress etc.). It may, however, again reflect the difficulty in assessing a child's cognitive abilities and progress effectively, an issue also raised in relation to the general deafblind population. Eyre (2000) explains that the use of standardised tests for deafblind learners is often unhelpful as:

The uniqueness of each individual deafblind child's learning difficulties does not bear comparison with any other child or group of children.
(p.120)

It is possible this distinction is even more marked for the individuals with CHARGE due to the truly multi-sensory nature of their impairment and the complex combination of anomalies associated with this condition. Further research is needed to develop the work of those who have explored the cognitive assessment of learners with CHARGE to examine the practical implications of this and the development of practical research tools.

5.8.1 Environmental audits

Within the 'Strategies' section of the questionnaire respondents were asked if the use of an environmental audit was helpful with regard to different aspects, with the data set out in Table 38 below. The format of the question does not support the identification of whether an environmental audit was undertaken unless the respondent reported that it was found helpful. The employment of environmental audits is well-documented in literature relating to education deafblind learners for sensory needs, communication, social and emotional, and orientation and mobility (e.g. Eyre, 2000; Pease, 2000; Murdoch et al., 2009; Nelson et al., 2010) as an important tool to inform the presentation of the physical environment and the delivery of the curriculum.

TABLE 38: TYPES OF ENVIRONMENTAL AUDITS AND THE FREQUENCY WHERE REPORTED TO BE CONSIDERED HELPFUL, AND DISTRIBUTION AMONG TYPES OF PROVISION

Type of environmental audit	Number where reported to be helpful (N=52)	SLD/PMLD provision	Mainstream provision	Other specialist provision
Sensory needs	23(44%)	10	5	8
Communication	22 (42%)	13	2	7
Social and emotional	17 (33%)	12	-	5
Orientation and mobility	21 (40%)	10	1	10

In literature relating to CHARGE the need to address the use of equipment and monitor the adaptation and differentiation of the environment, activities and materials in relation to the child's visual and auditory needs is also well-documented (see

2.6.3). It is reasonable to expect there also needs to be consideration of adaptations more broadly, considering the physical environment, the people within it and the activities being presented. The data obtained from this questionnaire shows that environmental audits were considered helpful by fewer than half the cohort for any of the areas being addressed, and for very few children in a mainstream setting. Since information gathered from an effective environmental audit can be used to inform all of these areas as part of the overall response, it may be beneficial to encourage and equip teachers to undertake such audits more often and to good effect.

5.9 Professional support

In the final section of the questionnaire respondents were asked to record those professionals in contact with the child with CHARGE in the educational setting as set out in the inventory provided and to list any others. Good practice suggests all professionals should work collaboratively (e.g. Thomas and Deuce, 2013) and ensure others have a clear understanding of their role, and it was recognised the accuracy of responses given would be dependent upon teachers knowing the role of each of those professionals. The responses obtained, set out in Appendix 10, were compared to information obtained earlier in the questionnaire (5.5.1) and found that:

- 22/48 children reported to have a visual impairment received input from a specialist teacher for visual impairment
- 38/49 children reported to have a hearing impairment received input from a specialist teacher for hearing impairment
- 38/45 children reported to have combined visual and hearing impairments received input from a specialist teacher for multi-sensory impairment. (2

additional children who did not have combined visual and hearing impairments also received input from this specialist teacher)

According to the 'Quality Standards for Children and Young People who are Deafblind/MSI' (Sense, 2002) there should be evidence of not only access to a qualified MSI teacher, but also support from specialist teachers for both visual impairment and hearing impairment. Input from appropriate specialist sensory impairment teachers is also set out within the more recent Code of Practice (Department for Education and Department of Health, 2014, 6.61), but these findings show this input is reported to be lacking for some of the children within this cohort.

Speech and language therapy input with regard to communication was the most reported input from an external professional being provided for 41 (79%) children in this cohort. In contrast input from a speech and language therapist with regard to eating and drinking difficulties was reported for only 18 (35%) children. When compared to the 24 (46%) children earlier reported to have had an eating and drinking assessment this suggests, that if both sets of figures are correct, some children had an eating and drinking assessment undertaken without the support of a specialist speech and language therapist. It is possible, that since a description was not provided of what an eating and drinking assessment comprised, that respondents included aspects that might not require input from a speech and language therapist (e.g. assessing food preferences). Given the number of studies that suggest over 80% of children with CHARGE experience 'feeding' difficulties (as cited in 5.8 above) this is an area that would benefit from further examination since it is not clear

whether this particular cohort are less affected or whether eating and drinking difficulties are being under-identified.

Only 10 children in this study were reported to receive input from a habilitation (mobility) officer. One of these children was reported not to have a visual impairment and one might therefore surmise that 39 children from this cohort, although having a visual impairment reported, did not receive input from a qualified habilitation worker. Input was noted as being provided for children where respondents had reported that 1:1 mobility sessions, long cane training and teaching sighted guide techniques were helpful (5.7.7a). According to Miller et al. (2011) the need for specialist medical and educational input is universally accepted for children and young people with a visual impairment and the same should apply to habilitation provision, with recognition that:

Assessment of the implication of sensory impairment in mobility and independence within the school should always be carried out by a Qualified Habilitation Worker. (p.20)

Since a large number of respondents felt that orientation and mobility of children in this cohort was affected by visual impairment, hearing impairment and vestibular difficulties (see 5.6.7a) it is likely to be even more important that appropriate habilitation support is provided. It would be interesting to explore whether a similar proportion of the wider deafblind population receive input from a qualified habilitation worker. One possible reason might be that there is a need for the habilitation worker to be able to communicate effectively with the child or young person (Miller et al., 2011), and this may be a barrier to provision of this input since one of the challenges faced by deafblind learners is a difficulty communicating with others (Aitken, 2000). This may be further compounded for children with CHARGE as a consequence of the reported difficulties in social interaction (see 2.3.2).

Thomas and Deuce (2013) identified potentially more than 50 professionals and agencies that could be involved in supporting the child with CHARGE and their family across health, education and social care, highlighting the need for collaborative working. Within this study the overall average number of professionals involved in providing support (in addition to the class teacher and any individual support worker) was 5.9, thus indicating the need for close collaborative working within the educational setting. There is a range of legislation and guidance that encourages working in partnership for all children with special needs and imposes duties upon the child's local authority to promote multi-agency working and collaboration (e.g. Department for Education, 2012). This is to help ensure the best possible support for the child, and the findings of this study suggest this needs to be recognised as a beneficial approach within the educational setting for learners with CHARGE.

5.10 Interviews

The interviews undertaken at the deafblind programme within Perkins School for the Blind obtained responses from 11 members of staff directly involved in working with students with CHARGE (as described in 3.7). The interviews sought the opinions of practitioners with experience of supporting a number of different students with CHARGE whilst recognising the information obtained was likely to reflect the particular ethos of this particular educational setting. It was anticipated the data gathered through the interview process would add depth to the overall findings of the study (See 3.8.1).

Questionnaire participants had been asked to complete the questionnaire with regard to one specific child whilst these interviewees would be asked to respond with regard to the group of learners with CHARGE they were involved with and also to draw on previous experience. This was made possible due to the number of students at the school who were or had been on the school roll (See 3.8.1), and it was considered this would provide data on the broader CHARGE population that could then be compared to the findings from the questionnaire where the individual child was the unit of analysis. The responses gained to each of the questions asked are as follows.

5.10.1 Question 1a: Do you feel that individuals with CHARGE are different to the broader deafblind population?

The general consensus was that learners with CHARGE are different to the wider deafblind population, with 10/11 interviewees giving an immediate and direct response. The remaining interviewee felt they were different, but also that there were similarities and so gave a yes/no response. These responses reflect the opinions expressed by a number of different authors and practitioners in the field as discussed in 2.5.

5.10.2 Question 1b: If yes, in what way?

Interviewees were encouraged to expand upon their response to the initial question, and shared the opinion that there were some similarities with the more general deafblind population in addition to any differences. The overall view expressed was that most individuals with CHARGE are part of the deafblind 'world' as a consequence of their combined visual and hearing impairments but that they are also distinct. This is perhaps best summarised by the comment that:

CHARGE syndrome is linked to the deafblind world but deafblindness does not equal CHARGE syndrome.

One interviewee explained:

The more you look at it, the more differences there are.

Three others drew attention to the view that there are:

So many other needs involved in CHARGE.

Responses obtained from interviewees showed they considered that philosophies and strategies well-established in the education of deafblind students can also be effectively employed with pupils with CHARGE. It was, however, additionally felt that a different emphasis may be needed on how these strategies are implemented, and different teaching methods will also be needed. The differences identified by interviewees between the students with CHARGE and the other learners in the deafblind provision are set out in Appendix 11, Table (xix), and relate to anomalies associated with this condition, the impact this has on the child's overall functioning, and also the strategies employed with this group of learners.

Many of these statements reflect elements of the CHARGE behavioural phenotype proposed by Hartshorne (2011) (see 2.3.4). Although interviewees gave general comments expressing a difference between students with CHARGE and other deafblind learners, it can be argued the particular examples given might also be demonstrated in other deafblind individuals. This will be discussed in detail in the following chapter, however, the language used by interviewees (e.g. 'elevated', 'greater', 'higher', 'more paramount' etc.) does indicate the opinion that the impact of CHARGE is more marked.

5.10.3 Question 2: What teaching strategies do you find most helpful (or unhelpful)?

The importance of giving recognition to, and employing, techniques and strategies established within the deafblind field was stressed by a number of interviewees. Three interviewees explained some deafblind strategies and techniques can be very valuable, but it is necessary to recognise some things need to be implemented differently and additional strategies developed when supporting a pupil with CHARGE. One person explained this, saying:

You need to know about deafblindness, but you need more too.

Another explained that some of the more standard deafblind strategies do not work in the same way, so need to be taken and individualised further. The implication of this is that teachers supporting the child with CHARGE will need to employ teaching strategies included in the 'toolkit' employed by teachers working with the broader deafblind population, but also other teaching strategies may be needed that could be regarded as more specific to learners with CHARGE. This view appears to be supported by most (if not all) of those interviewed at Perkins School.

Specific strategies identified by the interviewees are set out in Appendix 11, Table (xx). A comparison was made where these strategies identified were also addressed within the questionnaire which found that all had been reported to be helpful by a large percentage of questionnaire respondents, showing a level of consistency amongst practitioners as to some of the strategies regarded as beneficial when teaching learners with CHARGE. Three strategies (marked '▲' in Table (xx)) were not raised within the questionnaire but are well-documented in deafblind literature. A small number were not referred to in the questionnaire or noted in literature on the

education of learners with CHARGE or the wider deafblind population (these being marked ‘*’ in Table (xx)). Of these, sharing control may be regarded as an aspect of the development of ownership of learning but, in addition to empowering the student, may also be a strategy to reduce some of the stress and anxiety and the learner’s need to be ‘in charge’ as described by Majors (2011a). The 3 other strategies marked ‘*’ all relate to how information, instructions and expectations are conveyed to the learner.

5.10.4 Question 3: Do you consider their sensory impairments to be their primary need?

From the interviews it was interesting to note that whereas 10/11 interviewees were very clear learners with CHARGE are different to the wider deafblind population, there was greater difficulty in stating categorically whether the child’s sensory needs constituted the primary need overall. This demonstrates some benefits of face to face interviews that enable firstly, the interviewee to give a less precise response when not restricted by a Likert scale, tick boxes etc. (but also creating data responses that are more difficult to analyse statistically); and secondly, the researcher to judge the degree of conviction and certainty in an interviewee’s response that is not possible through the use of a survey.

Whilst 2 interviewees clearly stated they considered sensory impairments were the primary need for individuals with CHARGE, and 2 clearly stated they were not, the remaining 7 all gave a mixed yes/no response. This is in contrast to the responses of participants in the questionnaire for this research study (see 5.5) where 46 (88%)

respondents felt the child's sensory impairments were their primary need. The variation in these responses may be due to a number of factors:

- In the questionnaire participants were asked to respond to the question in relation to one specific child, whilst the interviewees at Perkins School related the question to a wider group of students with CHARGE
- The need to include these children as a part of the deafblind population to secure provision
- A variation in the educational philosophies (of special educational needs, inclusion, etc.) underpinning practice in the UK and at Perkins School

Both interviewees who responded 'yes' to this question felt it was the range of sensory impairments that made this the primary need. One explained about the impact of proprioceptive and vestibular difficulties on students, making it difficult for them to engage in a learning activity as it:

Takes all their attention to sit and hold their attention.

It was also explained that consequently all interventions need to be sensory based. Although perceiving sensory issues as creating the primary need, both gave recognition to other aspects of CHARGE that also have a significant impact on the students and need to be addressed including pain issues, and difficulty with initiation and shifting between activities.

Of those who responded 'no', one felt communication and the development of underlying concepts and cognition was the primary need, although it was recognised that the child's sensory impairments would affect the way materials were presented.

The other interviewee felt social and emotional learning difficulties to be the primary need. Amongst interviewees who gave a mixed 'yes/no' response it was felt that at times the student's sensory impairments could be regarded as the primary need, but in different situations other needs become more important with 1 interviewee describing it as:

A see-saw between sensory needs and other needs.

These interviewees all felt it was important to recognise the impact of the child's sensory impairments, but that it was not enough to address these issues alone. Other aspects identified as needing recognition were stress and anxiety, behaviour, planning and organisational skills, executive function difficulties, and social and emotional difficulties. The information provided by those who gave this mixed response illustrates the range of needs students with CHARGE may demonstrate and the difficulty in clearly identifying what the primary need is.

One interviewee raised the importance of highlighting sensory impairment as a primary need since this secures funding within the federal process and access to services only available where deafblindness/multi-sensory impairment is considered to be the primary need. Similarly, head teachers of special schools in the UK are required to participate in the School Census (Department for Education, 2015) that includes identifying and recording what each child's primary need is, which in turn will support planning and policy development, educational placement and resources to be made available to the child.

5.10.5 Q4: What, if any, support have you had in relation to CHARGE syndrome?

The comments obtained in response to this question (See Appendix 11, Table (xxi)) show that in addition to external sources, interviewees indicated there are clear benefits of working alongside colleagues who are experienced and knowledgeable about working with students with CHARGE, and having access to a range of different students with this condition.

Working in an educational establishment that has a number of pupils with CHARGE clearly lends itself to this type of support and professional development, but the challenge is how to provide this type of support to teachers in different settings attended by only one student with CHARGE (as is the situation for most teachers who completed the questionnaire) given this was the most frequently identified support mechanism. In addition to having experienced colleagues to hand, having such a large number of students with CHARGE in one educational setting also makes it more feasible to invest in a training programme on CHARGE, and also supports making links with a local college and being involved in research with students with CHARGE. It was noted, however, that only one interviewee reported direct input from external professionals in the field and is perhaps an illustration of how an institution such as this needs to safeguard against becoming insular and to continue seeking the knowledge, opinion and experience of colleagues further afield that may enrich and inform the future development of practice within this setting.

5.11 Final Summary and Conclusions

Cycle 2 of this investigation involved a questionnaire circulated to teachers supporting a learner with CHARGE and also interviews of practitioners in the deafblind program at Perkins School for the Blind. These surveys sought to gain information to test the initial theory and insights generated from Cycle 1 of the investigation (See 4.6) and build evidence to address the research questions.

The questionnaire received responses from 52 participants, with most being from different educational settings. This broadened the educational context thereby reducing the likelihood of answers being unduly biased by the prevailing educational philosophy or approach in any given educational establishment. For the purpose of the questionnaire the child was identified as the unit of analysis and the demographic data obtained showed there was an even spread across the different educational key phases (and therefore ages of the cohort) and different types of educational provision attended. This is important since the complex nature of CHARGE means it would not be possible to identify a 'typical' case, but by obtaining a cohort of children with CHARGE with a spread of ages and types of educational placements attended, it could be argued a sample that was likely to be more representative of the wider CHARGE population was achieved, that in turn would allow for some generalisation of the findings to the broader population of learners with CHARGE. This is further supported by the data that showed the presence and prevalence of anomalies and characteristics of CHARGE very similar to other data presented on the CHARGE population (See 1.1). This is an additional good indication that the cohort studied within this investigation may be considered as a fair sample and it is therefore

possible for other findings of this study to be applied to the wider CHARGE population. There does however continue to be a dichotomy expressed in literature between the belief that there are commonalities (e.g. Hartshorne et al., 2011) and the heterogeneous nature of CHARGE and it is therefore important to guard against making sweeping generalisations or assumptions.

The majority of respondents to the questionnaire (46/88%) felt the child's sensory impairments constituted the primary need in contrast to the more mixed response obtained from interviewees at Perkins School. By its nature, CHARGE is multi-faceted and although the child's sensory impairments are likely to be a substantial factor affecting learning and development, this research has shown other factors are also likely to be involved (including cognitive ability, executive function difficulties, etc.). A requirement is placed upon schools to identify a child's primary need in the annual School Census undertaken by the Department for Education (2015), and it is stated the information gathered in the census:

Supports the drive to raise standards, provides accurate targeting of funding, and assists the monitoring and development of policy. (p.3)

Although it may therefore be necessary to identify 'primary need' as a means to secure funding and appropriate provision, the evidence from this investigation indicates it must be questioned whether 'primary need' is a relevant concept in the education of learners with CHARGE as it carries the risk of a focus on the child's sensory needs to the detriment (and possibly lack of identification) of other needs. Educators will need to ensure a holistic overview is maintained; it is unlikely to be solely sensory impairments or 'true multi-sensory impairment', nor cognitive impairment or executive dysfunction that have an impact upon learning. Any or all

may be influencing factors, and all will need to be recognised and acknowledged with an individualised response tailored to the child. This is reflected in the most recent 'Code of Practice' (Department for Education and Department of Health, 2014) which states:

A detailed assessment of need should ensure that the full range of an individual's needs is identified, not simply the primary need. (p.86)

The mixed interview responses obtained highlight the need to maintain a holistic overview, with recognition that a child's needs are likely to vary over time and whilst sensory impairments will always create a need (that will necessitate a response) at any given time this may not be the primary and most pressing need at that point in time. Although it may be necessary to identify a child's MSI as their primary need to facilitate the release of appropriate and necessary resources, it is questionable whether it is most effective to attempt to identify the child's primary need with regard to their learning and development, and whether this identification could narrow the point of focus (rather than maintaining a holistic overview) and ultimately limit the recognition and response to other issues and resources made available to the child. Educators supporting a learner with CHARGE need to be aware of the many different aspects that can affect learning and development, as demonstrated in the findings of this investigation, that would need to be included in any educational 'diagnosis' of CHARGE, and not just a part of their multi-faceted needs.

The majority of the questionnaire cohort (45/87%) was reported to have combined visual and hearing impairments, thus being regarded as deafblind/ MSI, with the remaining all reported to have either a visual or hearing impairment. All the children were reported to experience difficulty with other senses (e.g. vestibular,

proprioceptive) and although not all senses were addressed in the questionnaire it is likely that some children within this cohort would experience impairments of these other senses given the rate of occurrence identified elsewhere in research. This is reflected in statements obtained during the interviews where learners were perceived to have 'more extreme sensory needs' arising in part from the combination of impairments that may be present. The internal factors likely to affect learning and development and the relative strengths and needs have been highlighted in the findings of the questionnaire, and these together with the information obtained from the interviews will be used to address the research questions in detail in the following chapter. Similarly, the degree to which questionnaire correspondents considered given teaching strategies to be helpful in supporting the learner with CHARGE and the identification of additional strategies was ascertained through analysis of the data gathered, with some of these strategies (and others) also being raised by interviewees. These will be compared to those described in literature on deafblindness and also literature more specific to CHARGE in the next chapter.

The data obtained allowed for some analysis to be undertaken with regard to the different educational phases and settings. The findings showed that across the age ranges there was little variation in the perceived level of ease/difficulty children were experiencing with regard to the development of identified skills, with only a few identified as being easier for those children in KS3/KS4 that may in part be a reflection of development and maturation. It was also found that although a few skills were perceived to be more difficult for children in an SLD/PMLD setting, overall there was little difference found across pupils in the different educational settings (see

5.6.8). The area where greatest variation was found across the different types of educational provision was in the reported level of helpfulness of the different strategies as discussed in (5.7.8). As discussed earlier, whether and how these are implemented (in addition to being a response to a child's identified needs) is likely to be influenced by teacher perceptions, their skills and knowledge, the school ethos, and the practical implications for the implementation of a given strategy. This study shows that what is made available to support the learner with CHARGE is likely to be influenced by the type of educational placement. It is therefore important to ask how different educational settings can ensure they are best placed to meet all the needs of these children.

Having gathered and analysed the data generated within Cycle 2 of this investigation, the final part of this cycle was to return to the research questions and develop final insights and theory. This will be set out in the following final chapter, together with a review of the entire research process undertaken.

CHAPTER 6- FINAL DISCUSSIONS AND CONCLUSION

6.1 Introduction

The intention of this research study was to explore how to contribute to reducing the 'problem space' (Petre and Rugg, 2012) that is a lack of research-based evidence and knowledge relating to the processes and practice of teaching learners with CHARGE. In 1.4 it was stated the challenge for the educationalist is how to deliver an effective education to foster development and learning in individuals with this condition.

The Code of Practice (Department for Education and Department of Health, 2014) states it is important to:

Ensure that all approaches used are based on the best possible evidence. (1.25)

When limited research-based evidence is available there will be a reliance on anecdotal reporting and opinion that, although helpful, can be subjective and limiting. Thus it is important to strengthen and build upon existing knowledge and acquire new evidence that can be drawn upon to support and inform the development of best practice. It was recognised this investigation would be exploratory in nature with the intention of providing some preliminary evidence to inform the education of learners with CHARGE, with a focus on children with a confirmed medical diagnosis of CHARGE in an educational environment (as defined in 1.6.3).

In chapter 1 it was explained that one purpose of making a medical diagnosis (and thereby classifying a condition into a specific category) is to inform and enable decisions to be made about the response that might be needed. This study explored whether it is possible that CHARGE could also be an educational diagnosis whereby, if a child was identified with this condition, it would follow that a particular educational approach was likely to be required. This led to the formation of the over-arching research question for this investigation: “CHARGE syndrome is a medical diagnosis. Can it also be considered as an educational diagnosis?”

6.2 The main findings

6.2.1 What are the factors within the child likely to affect learning?

In exploring the possibility of whether CHARGE can be considered as an educational diagnosis it was necessary to identify what the learning characteristics of an affected child might be.

CHARGE is a complex condition and there are many physiological features including the major and minor characteristics in the diagnostic criteria (explained in 1.1) that have the potential to impact on an affected child’s learning and development, as discussed in 2.2. The incidence rate of some of these anomalies in the CHARGE population, generally with regard to medical aspects, has been well-established through research. There is, however, less research-based evidence for the rate of occurrence of some other aspects (e.g. proprioceptive difficulties) and a greater reliance on anecdotal reporting by practitioners involved in supporting children and young people with this condition. In addition to noting the level of consistency (or

otherwise) when comparing the data arising from this study with established figures, it was also possible to explore to what extent the findings might support or contradict anecdotal reporting and opinion expressed in literature. The data also allowed for some examination of which of those factors explored may be perceived to impact most on a child with regard to their education.

Questionnaire and interview participants all considered the impact of the child's sensory impairments to be a significant factor affecting learning. The incidence rate of visual and hearing impairments and vestibular difficulties is well-evidenced in literature and medical reporting, and figures reported for this cohort of children closely matched other data (see Table 1 in 1.1). The likely impact of these impairments (including combined visual and hearing impairments resulting in deafblindness/MSI) is also well-documented in literature (discussed in chapter 2). Conversely there are no research findings that have accurately reported the prevalence of proprioceptive difficulties although different practitioners are of the opinion that individuals with CHARGE are affected and this subsequently impacts upon their learning (see 2.2.5). This investigation provided evidence that the majority of this cohort was perceived to demonstrate behaviours likely to be indicative of vestibular and proprioceptive difficulties which can impact upon their learning and development (see Table 25). Similarly opinion has been expressed that sensory integration difficulties are inherent in individuals with CHARGE (see 2.2.8) but again there has been no research to provide the evidence for this. Although a specific sensory integration assessment was not undertaken within this study, the data shows the presence of indicative behaviours within a large proportion of this cohort,

(summarised in Appendix 12, Tables (xxii) and (xxiii)). This suggests teachers are observing the presence of behaviours that can indicate sensory integration difficulties although it was noted only 7 (13%) respondents specifically named sensory integration dysfunction as a factor affecting communication and also the child's readiness and ability to engage and learn, and only 15 (29%) were reported to have input from a sensory integration specialist. Thus it may be a further raising of awareness and understanding of the potential presence of sensory integration dysfunction, together with an understanding of the impact this can have upon the child's learning and development as documented in literature (see 2.2.8) and the response required.

According to literature it is likely the development of haptic perception will be affected by a child's sensory impairments and particularly the presence of a visual impairment (see 2.2.7), a major characteristic of this condition. The findings of this study showed however that one particular aspect, a preference or aversion to some tactual experiences, may not be as significant for many learners with CHARGE as has been suggested previously (see 2.2.7) with only 18 (35%) children in this study perceived to experience a difficulty in this area.

In addition to the impact of all the child's separate sensory impairments and the interplay between them, within this investigation areas found to be of particular note in affecting learning and development of individuals with CHARGE were:

- High levels of fatigue: these were reported to affect children overall but also particular areas of development. Whilst only 3 (6%) respondents considered

fatigue to affect communication, 23 (44%) perceived it to affect the child's readiness and ability to learn, and 36 (69%) the child's orientation and mobility. Additionally, high levels of stress and anxiety (as reported by Wachtel et al., 2007) were reported in both the questionnaire (31/66%) and highlighted by interviewees

- Executive function difficulties: Appendix 8, Table (xiv) provides a summary of scores obtained with regard to how easy/difficult respondents considered a child to find a given skill. When comparing the skills listed in the top half of the table (those skills considered most difficult) and points raised by interviewees (see Appendix 11, Table (xix)), with the summary of the complex skills involved in executive function provided by Cooper-Kahn and Dietzel (2008) (see Table 2) it becomes apparent that executive dysfunction may be linked to those skills found most difficult overall by the children reported on in this study
- Sensory integration, sensory processing and sensory self-regulation affecting not only the child's sensory responses but other areas of development such as communication and social and emotional
- Neurological anomalies may also cause cognitive impairment that can result in learning difficulties (Gilles, 2011) and although sensory impairments may be regarded as a child's area of primary need, learning difficulties might also be a factor affecting the child in the educational setting as illustrated in 15 (29%) respondents identifying a child's learning difficulties as affecting communication.

Although gross motor development is often reported to be delayed in individuals with CHARGE, issues with the development of fine motor skills is an area not previously addressed. This investigation found fine motor skills were reported to be a difficulty for 39 (75%) of the cohort studied, and 40 (77%) were reported to demonstrate poor pencil/handwriting skills. The evidence from this investigation therefore strongly suggests this is an area that must be recognised and responded to appropriately.

The findings of this study identified factors reported to be present in large numbers of the children within this cohort. Since CHARGE is a congenital condition it would be expected that many of the physical anomalies associated with it (e.g. visual impairment, hearing impairment, vestibular difficulties etc.) would be present in children of all ages within this study. Other issues that might be regarded as potentially age-related, such as high levels of fatigue and poor body awareness etc., were however also perceived to be present in children across the different educational phases. Additionally, although a few of the skills examined were perceived to be easier for older children within the cohort, overall there was again little variation amongst the different age groups. This suggests that when supporting a learner with CHARGE it would be prudent to examine whether any or all of these factors are present so they might be addressed, and to recognise potential difficulties may continue to be experienced over time (supporting the need for a longitudinal study to explore this further). The heterogeneous nature of CHARGE means not every individual with CHARGE will be affected in exactly the same way, or to the same degree, by the anomalies associated with this condition. This is demonstrated in the variation in responses given (ranging from very easy to very difficult) for each

skill examined, but overall the data shows respondents reported very little that was considered overall to be very easy, and much that was perceived to be difficult or very difficult. The results set out in Appendix 8 Table (xiv), (discussed in 5.6.8), showed the greatest degree of difficulty perceived to be experienced by children within this cohort related to social and emotional development but also that difficulties were felt to be experienced within each developmental area. The findings also demonstrated a variation within a set of skills so that, within each developmental area, there was likely to be both skills regarded as some of the most difficult and conversely others reported to be easier in comparison.

Drawing on the findings of this investigation it is possible to propose some characteristics that may be present in the child or young person with CHARGE influencing their learning and development. These are set out in Table 39 on the following page that also records which element(s) of this investigation the evidence arose from. This reveals these findings were generated from different sources and demonstrate a positive level of agreement indicating the robustness of the investigation as a whole. That this level of agreement in the data generated from different sources was achieved also validates each individual source and strengthens the credibility of those issues raised solely in a particular source, including the original findings of this investigation. Since any of the many anomalies associated with this condition will come together in any affected child in a unique way it is important not to make assumptions that these characteristics will all be present in every individual with CHARGE. However, the rate of incidence within this cohort studied, together with the opinions of other practitioners experienced in supporting

TABLE 39: POTENTIAL CHARACTERISTICS IN A CHILD OR YOUNG PERSON WITH CHARGE THAT MAY INFLUENCE LEARNING AND DEVELOPMENT

Characteristic	Cycle 1	Cycle 2	
		Questionnaire	Interviews
A combination of sensory impairments- potentially all sensory modalities may be affected	✓	✓	✓
Sensory integration difficulties that include: Difficulty using their senses in a coordinated way Difficulty with distractibility/remaining on task	✓ ✓	✓ ✓	
Ongoing health issues and past medical experiences	✓		✓
High levels of fatigue	✓	✓	
High levels of stress and anxiety	✓	✓	✓
A preference for using different communication modes for both receptive and expressive communication that may include a greater preference: For using speech or sign/gesture to express themselves For concrete cues to support receptive communication	✓	✓ ✓	
Easier and more successful in forming relationships with adults than peers	✓	✓	
Difficulty developing same-age relationships		✓	
Difficulty understanding and expressing their own emotional state, and empathising with their peers	✓	✓	
Additional time to process information	✓	✓	✓
The ability to develop real, concrete concepts may be a strength		✓	
Establishing abstract concepts can be much more difficult	✓	✓	✓

Problem-solving may also be a relative strength if this is regarded as including the development of adaptive behaviours in response to difficulties or problems encountered		✓	
Needing a high level of routine and structure and experiencing great difficulty in coping when these are not established	✓	✓	✓
Benefitting from a use of concrete cues to assist anticipation of what is to happen next		✓	✓
Easier to recall past events than to predict events in the near future		✓	
Executive function difficulties that may be manifested as: Lack of flexibility and difficulty coping with unexpected changes Difficulty initiating activities and engaging in independent play and learning Difficulty developing and implementing self-organisational skills Difficulty coping with transitions and shifting attention from one activity to another Poor memory Difficulty transferring and generalising skills	✓ ✓ ✓ ✓ ✓	✓ ✓ ✓ ✓ ✓	✓ ✓ ✓
Easily affected by environmental distractions	✓	✓	
A need to be in control and a requirement for a level of negotiation			✓
Engaging in behaviours to address needs arising from poorly developed or under-stimulated vestibular and proprioceptive systems that may include: A need to adopt a horizontal position Seeking additional support for position when standing/sitting	✓ ✓	✓ ✓	
Poor body awareness and difficulty organising and coordinating their body		✓	
Experiencing fine motor difficulties with poor pencil/handwriting skills	✓	✓	

learners with CHARGE, including anecdotal reporting in literature, suggest due consideration should be given to them and where the presence of any of these characteristics is identified, this should then inform the development and implementation of any teaching programme and strategies employed.

6.2.2 What aspects of CHARGE syndrome might be distinct from the more general deafblind/MSI population?

It has been explained the majority of individuals with CHARGE will be considered as a part of the overall deafblind population, given that most will have combined visual and hearing impairments. Practitioners have, however, suggested that those individuals with CHARGE are educationally distinct from the broader deafblind population, despite extremely limited research-based evidence to support this (see 2.5). In considering whether CHARGE can be an educational diagnosis, it is helpful to also explore any similarities and differences that might be found when compared to the wider deafblind population, and identify characteristics that may be regarded as more specific to learners with CHARGE.

The diagnostic criteria of CHARGE (described in 1.1) clearly show the possibility for all sensory modalities to be affected; this combination of sensory impairments was not found to be reported elsewhere in deafblind literature, and is an initial clear distinction between individuals with CHARGE and the wider deafblind population. At the end of Cycle 1 of this investigation it was proposed that there are additional factors resulting not only from a child's sensory impairments but also other anomalies that can have an exponential impact on learning and development, this then being reinforced through the findings of Cycle 2. Having identified the potential

characteristics which might affect learning and development in individuals with CHARGE, literature on the education of deafblind learners was examined for related examples (a summary is provided in Appendix 12, Table (xxiv)). The initial comparison showed the majority of these characteristics were also addressed with regard to the broader deafblind population, suggesting similarities between learners with CHARGE and other deafblind individuals. Closer examination, however, shows the issues related to each aspect and how they impact upon the child may be different for learners with CHARGE than other deafblind learners. For example:

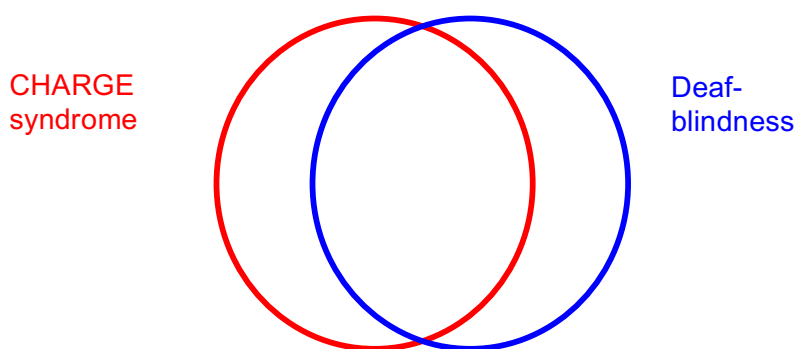
- Children in the cohort studied through the questionnaire were considered to experience difficulty engaging in independent play and learning. Hodges (2000) also explains there is a risk deafblind learners in general may “lapse into passivity” (p.188) when left to engage independently in an activity without direct adult support, suggesting this may be a consequence of the deafblind child having inadequate skills to engage purposefully with their environment in an independent way. This may well also apply to the child with CHARGE, but literature suggests a possible additional factor, that being executive dysfunction, that can also affect independent exploration as a consequence of a difficulty initiating activity and organising oneself.
- It is explained in deafblind literature that there is a need to employ a combination of communication modes (e.g. Pease, 2000; Murdoch et al., 2009) but it does not address the issue identified in this study, that a child may choose to use different modes for receptive and expressive communication and may have a greater preference for using concrete cues to support receptive communication than for expressing themselves.

- Clark (2000) and Murdoch et al. (2009) stated mobility in deafblind learners is likely to be affected because of poor body image, requiring input that focuses on developing body awareness. Poor body awareness was also identified as a factor affecting mobility amongst children reported on within the questionnaire, but additionally coordination and organisation of the body, fatigue and vestibular difficulties were also raised.

It has been explained it was found there were very few individual aspects that might be regarded as being distinct to the CHARGE population, although some were noted, namely high levels of fatigue and also stress and anxiety; executive dysfunction; the impact of neurological anomalies; sensory integration, sensory processing and self-regulation issues; poor fine motor development and pencil/handwriting skills. It is perhaps these aspects, in addition to the potential for true multi-sensory impairment, that have greatest impact on the learning and development of the child with CHARGE, and the combination of which makes them distinct from other deafblind learners. Furthermore, it would seem the anomalies associated with this condition may impact upon the child in a different way, creating different strengths and needs to other deafblind learners. The different way these anomalies come together and the varying degree of severity of each and the consequent impact upon learning and development will be different for every child, as shown in the range of scores obtained when a Likert Scale was employed to measure the perceived ease/difficulty experienced with regard to a given skill. This provides a reminder that not every child will experience the same degree of difficulty in developing skills and assumptions must not be made with regard to this simply because a child receives a diagnosis of

CHARGE. Nonetheless, the findings showed that for 15/21 skills explored, the most common response overall was that the child found them difficult/very difficult. McInnes and Treffry (1982) explain it is necessary to multiply the impact of each single visual impairment or hearing impairment when they come together to begin to understand the effect of these impairments being combined for the deafblind learner; building upon this description and considering the multiplicative effect of all the possible anomalies that can come together in CHARGE, then the resultant impact will be far greater again, likely to have an exponential effect as was proposed in 4.5.3 at the end of Cycle 1. This provides further support for the argument that an educational diagnosis of CHARGE might be possible.

This study has found there are both similarities and distinctions between learners with CHARGE and the broader deafblind population that might best be illustrated in the following diagram:



This shows an overlap between the features of the wider deafblind and CHARGE populations, but that there are also elements that are more particular to CHARGE which are likely to require a distinct response. It also suggests there may be aspects more particular to deafblindness, although not explored in this investigation (which

focused upon the intersection between CHARGE and deafblindness and what might be unique to CHARGE) which require further empirical examination.

6.2.2a The classification of Special Educational Needs debate

These findings can also be considered with regard to the wider debate on the categorisation of Special Educational Needs (SEN). The categories currently used (Department for Education and Skills, 2003):

Reflect administrative, placement and resource allocation decision-making...*[but]* not necessarily categories of learner characteristics that have pedagogic relevance. (Norwich and Lewis, 2005, p.4)

The potential dangers of categorising individuals experiencing difficulty learning in this way are raised in chapters throughout the publication by Florian and McLaughlin (2008), but potential benefits are also recognised (e.g. in the chapter by Terzi), and this process can support the identification of a child's strengths and needs and ensure an effective response is made.

Norwich and Lewis (2005) present a conceptual framework, focusing in particular on two positions that reflect how individuals with SEN's are responded to. The 'Unique Difference' position accepts pedagogy would be informed both by needs recognised as common to all learners, but also needs unique to the individual. Norwich (2008) advocates this as the 'default' position and being accepted for those broad areas of SEN (e.g. moderate learning difficulties, severe learning difficulties, etc.). Given that the unique differences position in addition to assuming that all learners have the same needs in general, also recognises and acknowledges needs unique to the individual, it may be argued this would be applicable to the CHARGE population

given the heterogeneous nature of this condition. This research, however, showed many similar perceived strengths and needs for children across the cohort and led to the formation of potential learning characteristics to be considered for all individuals with CHARGE, and also identified there is something distinct about this population. It is therefore argued it would be more appropriate to adopt the 'General Difference' position with regard to learners with CHARGE that, in addition to accepting the pedagogy would be informed by both needs common to all learners and unique to individuals, also recognises:

Needs that are specific or distinctive to a group that shares common characteristics. (Norwich and Lewis, 2005, p.3)

Indeed this is supported by the work of Norwich and Lewis (2005) who reported that where it was found a group is "more specifically defined" (Lewis and Norwich, 2005, p.215), such as Autistic Spectrum Disorder, visual impairment or hearing impairment, it is more likely the general differences position will be adopted. It has been argued in 5.11 that the concept of primary need, with regard to the Department for Education and Skills (2003) classification system, may not be a relevant concept for learners with CHARGE. This research has shown that 'CHARGE syndrome' as a grouping may be more appropriate in supporting the identification of potential learning characteristics and strategies to support an effective pedagogy for learners with CHARGE. It is important to understand that adopting the general difference position for this specific group:

Avoids simple stereotyping in terms of group-based pedagogic needs...because it takes account of common and individual needs. (Norwich, 2008, p.145)

It does not automatically imply a specialist pedagogy encompassing all learners with CHARGE to be administered in a particular way, but suggests specific aspects to be

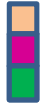
considered and applied differentially, acknowledging the importance of focusing on the individual child, whilst also considering the broader contextual framework.

6.2.3 What strategies are likely to be effective in supporting the learning of children with CHARGE syndrome?

One purpose for making a diagnosis is to enable decisions to be made about the response likely to be required. Accepting the argument that learners with CHARGE do form a distinct group it is then necessary to consider whether it is possible to 'prescribe' what is likely to be required educationally and thus examine the educational philosophy and strategies implemented. According to the Code of Practice (2014), high quality teaching includes differentiation and personalisation so as to effectively meet the needs of the individual child. This highlights a need for educators to focus initially on identifying both the strengths and needs of each child with CHARGE when planning and delivering an educational programme which then leads to the question of whether, given the need to make an individual response, it is possible to identify strategies that may be regarded as beneficial for the broader CHARGE population. The results of the questionnaire provide some evidence that this is possible given that, although the data did not allow for the identification of the total number of children with whom each strategy was employed, 35/46 strategies included within the questionnaire were reported to be helpful by more than 50% of respondents.

Table 40 provides a list of all those strategies included in the questionnaire and records those also raised by interviewees and whether they are identified in

TABLE 40: STRATEGIES INCLUDED WITHIN THE QUESTIONNAIRE AND NUMBER OF RESPONDENTS REPORTING THEM TO BE HELPFUL, AND IDENTIFICATION OF WHETHER EACH STRATEGY IS NAMED BY INTERVIEWEES AND ALSO IN LITERATURE.

Strategy  Strategy noted in deafblind but not CHARGE literature Strategy noted in CHARGE but not deafblind literature Strategy not identified in either deafblind or CHARGE literature	Questionnaire: Number of responses (n=52)	Interview responses	CHARGE literature	Deafblind literature
Use of a Total Communication approach	48 (92%)		✓	✓
Use of a daily routine and structure implemented consistently throughout the day	48 (92%)	✓	✓	✓
Key people identified to develop secure relationships with child	47 (90%)		✓	✓
Differentiating materials/activities in relation to child's sensory needs	46 (88%)	✓	✓	✓
Use of visual/auditory aids	46 (88%)		✓	✓
Use of concrete tools (e.g. real objects, photographs, symbols etc.)	46 (88%)	✓	✓	✓
Provision of real experience provided within context	44 (85%)		✓	✓
Clear identification of what is motivational for the child	44 (85%)	✓	✓	✓
Use of a flexible approach, responding to the child's changing needs and levels of responsiveness	44 (85%)	✓	✓	✓
Ensuring access to a peer group with an appropriate level of communication skills	42 (81%)			✓
Structured support for transitions	42 (81%)		✓	✓
Providing regular opportunities to engage in large movements	42 (81%)		✓	
Providing a balance between new and familiar activities	41 (79%)		✓	✓

Clear identification of the child's individual learning style informing the delivery of activities	41 (79%)		✓	✓
Ensuring activities have a clear beginning, middle and end	40 (77%)		✓	✓
Use of a structured programme/activities to support interactions with peers	39 (75%)	✓	✓	✓
Providing frequent choice-making opportunities	39 (75%)		✓	✓
Structured activities to support the development of gross motor skills and body awareness	39 (75%)		✓	✓
Activities planned/implemented in relation to child's sensory integration difficulties	37 (71%)		✓	
Implementation of a structured programme to support self-help and self-organisational skills	37 (71%)		✓	✓
Developing profiles of functional use of vision and hearing	35 (67%)		✓	✓
Providing a sensitive communication partner who is able to recognise and respond to the child's attempts to communicate	35 (67%)		✓	✓
Support to learn how to initiate/sustain social interaction sequences	33 (63%)	✓	✓	
Use of mini routines	33 (63%)		✓	✓
Use of specialist/supportive seating	32 (62%)		✓	
Appropriate behaviour management with recognition of impact of sensory integration difficulties	31 (60%)		✓	
Implementation of a fine motor skills programme	31 (60%)			
Implementation of a structured programme to develop the child's understanding of emotions	31 (60%)		✓	✓
Individualised pacing (may include a reduced timetable)	31 (60%)		✓	✓

Use of identified strategies to support the development of problem-solving skills	30 (58%)			✓
Use of concrete tools to support sequencing of an activity, transitions and daily routine	30 (58%)	✓	✓	✓
A structured response to manage the impact of fatigue	29 (56%)		✓	
Provision of alternative recording methods (e.g. scribe, use of IT, etc.)	29 (56%)			
Use of structured fading and pre-learning/post-learning to support learning	28 (54%)			✓
Use of large movement play activities	28 (54%)		✓	
Development and implementation of an individual communication programme	26 (50%)		✓	✓
Access to a safe rest area	25 (48%)		✓	
Undertaking an environmental audit in relation to child's sensory needs	23 (44%)			✓
Undertaking an environmental audit in relation to the child's communication needs	22 (42%)			✓
Responding to the child's executive function difficulties	21 (40%)		✓	
Implementation of a pencil skills programme	21 (40%)			
Undertaking an environmental audit in relation to child's orientation and mobility needs	21 (40%)			✓
Use of a defined programme to manage stress and anxiety levels	20 (38%)		✓	
Undertaking an environmental audit in relation to the child's social and emotional needs	17 (33%)			✓
Activities to support the development of haptic perception (active use of touch)	13 (25%)			
Monitoring to identify the risk of the development of 'splinter' skills and responding	7 (13%)		✓	✓

literature both on the education of learners with CHARGE and the broader deafblind population. This shows most of these strategies have also been identified within the literature on CHARGE examined during the literature review (noting that many of these are 'grey materials'), and a number also reported by interviewees.

The findings of this research study also show most of the strategies reported to be helpful by larger number of questionnaire respondents are included in established practice with deafblind learners, supporting the opinion of Brown (2011a) who stated that deafblind education philosophy and practice provides the 'best fit' for learners with CHARGE. The data arising from the questionnaire did not allow for analysis of how the strategies were used, and thus whether the same strategy may serve a different purpose when employed with a learner with CHARGE than other deafblind pupils. This was, however, a point raised by interviewees who explained a strategy established for use with deafblind learners may be employed in an alternative way or for a different purpose (see 5.10.3). For example, visual calendar systems likely to be employed with a deafblind learner to help the child order the day, support communication and social interactions and concept development (e.g. Engleman et al., 1998; Pease, 2000), were described by an interviewee as being employed with the learner with CHARGE to support self-organisational skills and help reduce and manage perseverative behaviour. A small number of strategies contained within the questionnaire were found to be noted within deafblind literature, but not mentioned in literature on CHARGE (coloured beige for easy reference in Table 40). With the exception of the provision of a peer group with appropriate communication skills, the remainder concerned the use of an environmental audit with regard to different

needs within the child. These were reported to be helpful by 44% or less of respondents, and whilst it may be these had been used but were found to be unhelpful, it may also be a reflection of the lack of identification of the use of such environmental audits in literature on CHARGE resulting in a lack of awareness of the potential usefulness of these as a tool to support the delivery of the curriculum by the teachers involved in this investigation.

As discussed in 2.6.3, there is some suggestion that the philosophy of deafblind education, although beneficial for learners with CHARGE, will not in itself fully meet the needs of this group of learners. Within Table 40, 10 strategies were noted found not to be reported in deafblind literature but that were expressed in literature on CHARGE (coloured pink). With most reported to be helpful by over 50% of respondents this provides some evidence additional strategies may be beneficial that might be regarded as more specific to the education of learners with CHARGE. Within Table 40 there are also highlighted a small number of strategies (coloured green) that have not been identified elsewhere in literature with regard to learners with CHARGE or the wider deafblind population. Structured activities to support the development of haptic perception was a strategy regarded as helpful by only 13 (25%) of respondents and, as discussed in 5.7.1 may be a reflection of little difficulty perceived to be experienced by children in this cohort (see 5.6.1) in contrast to what is stated in literature. This may also have been found by other practitioners working with children with CHARGE and thus a reason why this strategy has not been highlighted elsewhere. It will be important to acknowledge, however, this may be a helpful strategy for some learners with CHARGE, as illustrated by the findings of this

study. The three other strategies identified all relate to fine motor/pencil skills and the usefulness of providing alternative ways for the child to record their work. Whilst this might not be an issue widely identified within the deafblind population it could be argued this may also be present in other groups such as children with Down syndrome. Within this latter group, however, poor hand function is regarded as resulting from the physiology of the hand, poor motor coordination and a poor tactile sense (e.g. Jobling and Virji-Babul, 2004) whereas for learners with CHARGE, hand function difficulties seem less likely to be a consequence of a poor tactual sense according to the findings of this study, but may perhaps be more affected by the poor development of gross motor skills, poor proprioception and cranial nerve anomalies. This is an aspect that will benefit from further exploration and research.

It has also been identified that there may additionally be a need to employ established deafblind practice in an alternative way or for a different purpose. This provides further evidence that learners with CHARGE will require a supplementary approach that again might be considered as distinct from other groups of learners, thereby also supporting the argument that an educational diagnosis of CHARGE may be appropriate and beneficial.

6.2.4 CHARGE syndrome is a medical diagnosis. Can it also be considered an educational diagnosis?

CHARGE is medically classified as a syndrome which means there must be something distinct and unique about this condition since by definition the term 'syndrome' is assigned to a group of anomalies that consistently come together in a unique way. Medically it will be regarded as separate to other syndromes (that in

turn will all be distinct from each other), including those that may also result in congenital deafblindness (e.g. Congenital Rubella syndrome, Noonan's syndrome. etc.). The deafblind population is one of many parts, including the majority of individuals with CHARGE as demonstrated in this study. The issue is whether educationally learners with CHARGE are also distinct and unique from other parts of the deafblind population and if there is a distinct educational 'prescription' that is required in response to those educational needs.

The findings of this investigation have led to the proposal that there are identifiable potential characteristics of the learner with CHARGE that arise from both the anomalies associated with this condition themselves and also the impact they may have upon the child. Whilst there was considered to be a clear overlap between the features of learners with CHARGE and the broader deafblind population, and also the educational philosophy and pedagogy employed with each, the findings of this investigation have provided evidence that in addition to the medical uniqueness and distinctness of CHARGE, there is also educationally something distinct and unique about learners with CHARGE.

The appropriateness of classifying a condition in such a manner to form a distinct category for educational purposes does, however, require some further exploration. When a diagnosis is presented there is a danger of focusing on the condition rather than the individual, and making assumptions about how the condition will be manifested in any individual. It will always be important to recognise the

heterogeneous nature of CHARGE and ensure an individualised, child-centred approach where:

The main driving force must be clear identification of the child's educational needs and how these are to be met. (Deuce, 2013a, p.1)

The use of an educational diagnosis of CHARGE can potentially lend itself to the application of a medical model, as described by Booth and Ainscow (2011), where the focus is on the learner's educational difficulties seen as a consequence of impairments and/or deficits within the child. This study has shown however, that in addition to the internal factors there are also external factors that require consideration, this lending itself more to the social model (also described by Booth and Ainscow, 2011) as one where potential barriers to learning are to be found in the educational context and learning environment. Thus it becomes apparent that although an educational diagnosis of CHARGE can provide a potential 'checklist' of characteristics within the individual for consideration, it will also need to include attention to the learning environment itself. This is reflected in a study of the inclusion of deafblind young people in mainstream schools by Kamenopoulou (2012), where the inclusion process was found to be influenced by both internal and external factors.

This study has found that in addition to the similarities found between learners with CHARGE and the wider deafblind population and the pedagogy most effectively employed, there is also evidence that there are distinct elements. Thus it may be both possible and helpful to use CHARGE as an educational diagnosis which may then progress to the educationalist being able to prescribe what might be needed to help inform the response made. This investigation provides some initial indicators

that there may be a need for a specific programme to be implemented when educating learners with CHARGE. This is not a new notion with specific approaches clearly argued (with evidence through research to support them) for other specific groups of learners with special needs, including learners with autistic spectrum disorder (e.g. Mesibov et al., 2004) and Down syndrome (e.g. All Party Parliamentary Group on Down Syndrome (APPGDS),2012; Duff et al., 2012).

6.2.5 Learners with CHARGE in different types of educational settings

It was interesting to note the findings of this study showed some variation according to the type of educational provision the child was placed in. For example, it was found children reported on in a residential setting were perceived to find the undertaking of self-help skills and functional life-skills easier, and it was suggested this may indicate a potential benefit of a learner attending a residential placement (see 5.6.6).

The view has been expressed that on the whole special schools will be attended by pupils with more complex learning difficulties (e.g. Department for Education, Northern Ireland, 2006), this being supported by figures presented by the Department for Education (2014) but overall, the findings of this study (reported in 5.6) showed that whilst a small number of skills were perceived to be more difficult for children placed in an SLD/PMLD setting, similar levels of difficulty were perceived to be experienced across a number of skill areas for children in all the types of educational settings.

One unexpected but important finding of this investigation relates to those learners reported on in the questionnaire in mainstream provision (31% of the total cohort) who overall were reported to experience a similar degree of difficulty with regard to a number of skills examined as those children in specialist school settings (including SLD/PMLD schools). From this it might have been anticipated that similar responsive strategies may perhaps also be considered beneficial for children across the different educational settings. The findings of this investigation, however, showed many of the strategies examined were reported to be helpful less often for those children in a mainstream setting; Tables 34 and 35 set out those strategies more frequently reported to be helpful by teachers working in specialist settings but conversely, there were very few strategies reported to be helpful by individual teachers in mainstream settings that were not raised by teachers in specialist settings (e.g. the use of task plans and role play). One possible inference from these findings is that children attending mainstream provision may be supported differently (despite similar difficulties being perceived across the cohort).

It is very likely that the type of strategies implemented will be influenced not only by the child's strengths and needs but also by teacher and school characteristics that will include the different knowledge base and skill set of teachers in the separate settings. Thus it is suggested it would be beneficial to explore further the relative benefits of the different skills and knowledge base from teachers in all the educational settings and how these might be more widely shared to ensure learners with CHARGE have appropriate access to the curriculum and support for their learning and development needs to enable them to "achieve to the full" (Department

for Education and Skills, 2004, p.9), regardless of their educational setting. Government guidance (e.g. Department for Education and Skills, 2001 and 2004) advocates special schools undertaking an important role in supporting inclusion, and one possible inference from these findings is that staff in these settings may have a greater awareness of particular strategies that might be helpfully employed for the child with CHARGE. It would however also be important to ensure a reciprocal arrangement with teachers from mainstream provision also sharing their teaching experiences and useful different strategies they have developed.

Both Evans and Lunt (2002) and Möller and Danermark (2007), (the latter exploring the inclusion of postlingually deafblind students), reported difficulties in achieving effective inclusion as emanating from internal factors within the child, but also limitations within the school setting with the physical environment, poor delivery of the curriculum and inflexibility, poor differentiation and modification of resources, and a lack of professional development for staff. Following this investigation it is suggested that, when considering the education of learners with CHARGE, these issues may need to be generalised beyond the inclusive mainstream setting to all types of educational provision; for example, Dyson et al. (2004) found some evidence that children with special educational needs can make good progress academically and socially in an inclusive setting but also highlighted indicators that these children are at risk of isolation and low self-esteem. The findings of this study, however, suggest that many individuals with CHARGE are at risk of social isolation irrespective of the type of educational provision attended (see 5.6.3).

Providing adequate training and support for staff must be another key factor (as identified from this study- see 5.9 and 5.10.5). Booth and Ainscow (2002) claim:

There is a wealth of knowledge within a school about what impedes the learning and participation of students (p.5)

In reality, CHARGE is a low incidence condition and the majority of teachers working with a child with CHARGE in their class are unlikely to have developed a secure and broad knowledge base or have past experiences to draw upon. Providing an effective education for the majority of learners with CHARGE is likely to be a challenging task in any educational setting due to the complexity of the condition and this study has illustrated the number of specialists likely to be required when supporting learners with CHARGE in any educational setting (see 5.9). Giangreco et al. (1999) make an opening statement in their paper, that deafblind students require specialist support to experience successful education, a point reinforced by the findings of this study for learners with CHARGE. A study by Frederickson et al. (2007) showed the benefits of input from a specialist team to support effective inclusion. This is an example of the importance of the need for appropriate specialist support to ensure success in an educational setting not specialising in supporting particular areas of special needs. Thus, given the complexity of CHARGE and the distinct aspects identified within this study, it is possible that input from a 'specialist team' should be considered in every educational setting attended by a child with CHARGE, although acknowledging Evans and Lunt's (2002) explanation that the delivery of such specialist services is more difficult to provide outside a specialist setting. The findings of this study show an important contributor to this specialist team must be a specialist in the field of deafblindness who additionally has

knowledge and experience of supporting learners with CHARGE. Given that Möller and Danesmark (2007) stress the importance of knowledge about a young person's diagnosis of deafblindness to support successful education, how many more elements does the teacher of a child with CHARGE need to be made aware of and respond appropriately to?

Sixty-nine percent of children included in this cohort were placed in specialist school settings. It has been suggested it is possible the remaining 31% placed in mainstream school are supported differently. This may be for a number of reasons, for example, those strategies identified within the questionnaire had been tried but were found to be unhelpful, it was not necessary to implement these strategies, alternative strategies have been identified and employed, or teachers are unaware of these strategies. Kamenpoulou (2012) states:

The needs of deafblind pupils in mainstream schools are a significantly under-researched area (p.143)

This is equally valid for learners with CHARGE in the mainstream setting, and also warrants further investigation.

6.3 A review of the investigative process

From the outset it was intended this study would be exploratory in nature with an emphasis on discovery. It is recognised future research will be needed to reduce the research space further and to confirm or disprove the findings of this investigation. To date this is the only research study to explore the pedagogy for learners with CHARGE (although Lieberman et al., 2012 have explored the area of physical education for 26 children with CHARGE).

It was decided to adopt a case study methodology and two clear cycles were identified that would be undertaken during the process of this investigation as set out in 3.4. Cycle 1 involved gathering data through the analysis of educational reports to generate some initial insights and theory that would inform the development of Cycle 2 of the study. During the process of establishing the coding categories it was decided to adopt the use of an external tool in the form of a curriculum designed for MSI learners (see 3.5.7b). This helped provide a clear framework for the coding process and also had the additional advantage of providing an initial indicator of some potential differences between learners with CHARGE and the broader deafblind population (see 4.5.1). In Cycle 2 a survey was undertaken of teachers involved in supporting a child with CHARGE in their class through the use of a questionnaire. The structure of the questionnaire employed for the survey was designed in such a way that would encourage a response but also elicit accurate and valid information. Following on from Cycle 1 it was decided that the questionnaire would examine both internal and external factors that may be involved. It had been decided to use mainly closed questions and the use of a Likert Scale and inventories as a way of gathering responses, with the addition of a few open question forms. Although there is potential for limiting the way in which a response can be given, it was demonstrated this generated a greater level of response than might otherwise have been achieved, this being shown in the very limited number of responses provided when the use of a request to list 'others' was made. Using this structure to elicit information also made it easier to analyse the data and make comparisons and provided a way of observing patterns and regularities. The use of a Likert Scale further provided some measure of intensity of response that also proved to be

beneficial; it was interesting to note that the full range of the scale (5 points) was used by respondents despite Oppenheim (2000) cautioning that respondents often avoid scoring at the two extreme ends of the scale.

Some use was also made of open question forms to gather more qualitative information. The responses obtained through the use of a questionnaire will be dependent on the question forms used and how a sentence is phrased, but also affected by each respondent's views, knowledge and experience. When examining the responses given it is possible the examples provided may have influenced the respondents' answers given that, for example, the two most frequent answers to the question 'What factors do you feel adversely affect the child's readiness and ability to engage and learn (e.g. environmental distractions, fatigue etc.), were the two factors presented as examples. Whilst the examples could have been excluded from the questionnaire, they were provided to help ensure a shared understanding of the question. Other efforts were made to ensure shared understanding (see 3.6.4 and 3.6.5) but if, for example, the question on whether the child demonstrated touch sensitivity ('e.g. a reluctance to use touch to explore or to touch certain textures') had been phrased differently, would it have obtained a higher response more reflective of what is expressed in literature (not evidence-based) or are these findings a more accurate representation? The difficulty in ensuring shared understanding is also demonstrated in the fact that higher levels of response were obtained to issues related to executive dysfunction (see 6.2.1 above) than the responses obtained to the one question that specifically used the term 'executive function difficulties'. It was suggested one factor might be a lack of knowledge and understanding of the

terminology (a higher rate of response was obtained from teachers working in SLD/PMLD settings that may indicate a consequence of specialist knowledge- see 5.7.6), and demonstrates the importance of teachers clearly understanding all the potential implications for a child with CHARGE, including executive function difficulties.

On reflection a different phrasing of some questions would have elicited different or additional information that might have generated further useful data. For example, respondents were asked to identify factors within the child affecting their communication. It was recognised it could have been beneficial to ask this question in relation to the child's overall learning and development rather than restricting the question to communication. From the data generated from the 'Strategies' section it was not possible to ascertain whether a strategy had been employed but was then not found to be helpful. In hindsight it would have been useful to adopt the format used in the following section on 'Assessment' to ask whether a strategy had been employed, and then to establish whether it was found to be helpful.

Throughout this investigation every effort was made to ensure it was both reliable and valid. The purpose and significance of this study were clearly defined, with the intention of building on existing knowledge and providing research-based evidence to help reduce the 'problem-space'. The review of literature explored and identified prominent themes which were then used to inform the conceptual framework of this study including the establishment of the research questions. The chosen methodology and methods enabled the gathering of data that was then used

effectively to address the research questions. The process undertaken has been clearly outlined that would allow for this study to be repeated if required. This included checks for reliability in both cycles and also addressed the risk of potential researcher bias. Multiple sources and types of data were also employed with the level of agreement identified (and also supported in literature) demonstrating a robustness in the findings that also gave credence to the original findings of this study. Although the data gathered all reflected teacher perceptions, a wide range of opinions were obtained from practitioners in different educational settings, including teachers experienced in working with a number of learners with CHARGE and those for whom this may be the first time they have taught a child with this condition.

As with many other research studies that involve the use of surveys, the responses obtained will be a reflection of the respondents' perceptions, in this case the teachers involved in supporting the children. It would be difficult to obtain the information sought through other processes (for example, there are not formal assessment tools to elicit this information that are standardised for learners with CHARGE- see 2.4), and although researchers are reliant on perceptions reflecting what is real, teacher perceptions are an important and valid source of information as demonstrated in the reliance upon these for the scoring in profiles of attainment and development of individual teaching programmes (e.g. Assessment Reform Group, 2006) which are all dependent to some extent upon teacher perceptions. During the course of the academic year teachers would have the opportunity to get to know the children in their class well and build up direct knowledge to draw upon when responding to the questionnaire. Thus, teachers had been given some time to get to know the child with

CHARGE since the questionnaire was sent out during the spring term. Determining and addressing what influences those perceptions is necessary with consideration given to the teaching context, teacher experience and biography (Beijaard et al., 2000; Avramidis et al., 2000). The teaching context with regard to the type of provision, the culture of the school, shared values etc. may all have influenced the teacher's perceptions of the child. For example, when deciding on how easy or difficult a given skill is were the teachers comparing the child to the peers in their setting, which is likely to involve different peer groups for children in a mainstream rather than an SLD/PMLD setting? For all teachers, practice over time enables the development of a knowledge base that draws on past experience and is also likely to influence their perceptions. Hastings et al. (1996) found those:

With higher levels of previous contact with children with severe learning difficulties were generally more positive than those with little or no previous experience (p.139)

Considering CHARGE in light of this statement, it is likely to be a significant factor that for many teachers it will be a new experience working with a child with CHARGE due to the low incidence of the condition and the policy of inclusion, unless they are working in one of the few provisions that has more than one pupil with CHARGE. This also validates the use of interviews with practitioners at a school attended by a number of pupils with CHARGE, where there has been the opportunity for staff to build up their experience and knowledge base. Other experiences would also influence perceptions where, for example, teachers working in specialist settings may have more specialised knowledge that might be specific to that domain and would also influence the responses given. Rivard et al. (2007) consider that teacher perception has a critical role in the identification and management of a child's strengths and needs. The questionnaire findings noted that overall most of the skill

areas addressed were considered to be difficult or very difficult for the learner with CHARGE, that according to Rouse (2008) can lead on to the belief that:

When children find learning difficult, it is because there is something wrong with them [*the child*]. (p.6)

When a child is experiencing difficulty learning there is a danger this can result in a deficit model where teachers focus on the child's disabilities and difficulties, rather than seeking to identify their relative strengths to build upon. So, whilst most individuals may find at least some aspect of learning and development difficult, it is possible that teachers are not clearly identifying areas of strength that may be present in the child with CHARGE which may include them 'doing things differently'. As an example, identifying solutions to problems was scored at the midway point (see 5.6.4), so teachers overall perceived this to be neither particularly easy nor difficult. It was argued that if teachers are able to recognise adaptive behaviours as a problem-solving skill this might be recognised as a greater strength within the child (and one that Hartshorne et al. (2005b) felt often illustrates underlying intelligence. See 2.4). Understanding what may be involved with CHARGE and the impact this has on the child's functioning, together with the provision of the necessary specialist support to ensure teachers feel equipped to respond, will have the potential to influence teacher perceptions and ameliorate the educational environment through clearer identification of how the child learns and what is needed to support their learning.

Möller and Danermark (2007) explain that the deafblind population is small and it will therefore be difficult to obtain a large sample when engaging in research. This must be even more so when researching a low incidence condition such as CHARGE. It

was decided that use of a survey provided a realistic way of potentially reaching greater numbers for inclusion in the study, a method frequently employed by other researchers within the field of CHARGE (as discussed in 2.1). Although a sample was used chosen according to specific criteria, to try and secure data from a wide sample of the population these were kept as broad as possible (see 3.6.7a). It is not possible to state categorically that a truly representative sample was obtained but steps were taken to widen the pool as far as possible through a range of different sources (see 3.6.7a). The data from the sample obtained, ultimately showed a good cross-section had been achieved across both the educational phases and types of educational provision (see 5.4.1 and 5.4.2), and also included a number of children with CHARGE who did not have combined visual and hearing impairments.

Kamenopoulou (2012) recognises the heterogeneous nature of deafblindness that creates difficulty in generalising conclusions from research, but nonetheless was able to identify some emergent patterns from different studies examined. A similar argument could be presented for this investigation; although CHARGE is a diverse condition, some patterns have emerged that suggest commonalities amongst the learners with CHARGE included in the cohort. Similar rates of occurrence of anomalies that form a part of diagnostic criteria were noted within this sample that matched data from other research, together with the level of agreement obtained from a number of sources and through the employment of different tools, supporting the view this sample is likely to be similar to other individuals with CHARGE and it would therefore be possible to consider these findings in relation to the broader CHARGE population.

6.4 Conclusions

At the closing of this investigation some conclusions have been drawn in response to the research questions posed. There are many separate aspects found to be similar for both learners with CHARGE and the wider deafblind population. It is perhaps rather the way all the many anomalies come together within the affected individual having an exponential effect, together with the small number of aspects that might be regarded as more specific to this condition, that create a distinction between children with CHARGE and other deafblind learners. A similar conclusion has been reached regarding the pedagogy for learners with CHARGE; whilst much from the deafblind field may be considered helpful and appears to provide the 'best fit' suggested by Brown (2011a), it has also been suggested there is a need to implement strategies in an alternative way or for a different purpose, and again that something supplementary may be required. Thus it has been proposed that educationally there is something distinct and unique about CHARGE, with potential learning characteristics identified and strategies that may be beneficial when included in an educational programme.

Evidence has been gathered that suggests it may be possible for CHARGE syndrome to be used as an educational diagnosis in a broad sense. A cautionary note has been raised against taking a purely medical approach that focuses solely on the internal aspects and can lead to teachers adopting a 'deficit' model in their approach to teaching the child which can adversely affect attitude and expectations. This study has shown it is also important to consider the external factors of the learning environment and it has been suggested there is a need for any 'educational

diagnosis' to combine both medical and social models. Providing an educational diagnosis supports awareness of potential characteristics that may influence learning and development and also supports the educationalist in identifying what should be considered for inclusion in the response made. There is an argument that the heterogeneous nature of CHARGE requires a highly individualised educational programme whilst others present the case for commonalities within the CHARGE population (see 2.6.1). This investigation provides an illustration that there is a place for both sides of this argument. There may be a need for highly individualised programmes but the development of these should be informed by the common themes that have emerged with regard to learners with CHARGE.

The question of what is the best way to educate learners with CHARGE will need to be addressed in more depth. Support from a number of different specialists is likely to be required to support the implementation of an effective educational programme. It has been found that the input provided is likely to be influenced by the type of placement attended by the child. This study has shown a range of strategies are likely to be more widely employed by teachers in specialist settings than teachers in a mainstream setting and this raises the question of what appropriate strategies are being implemented in the mainstream setting. It will be important to identify the potential benefits of both the specialist and inclusive mainstream settings and how the best of each can be made available to every child. This should include considering how outreach support from specialist schools might be provided within the mainstream setting and vice versa. Given that a deafblind approach is likely to be

the 'best fit' it also follows that input should be provided from a specialist deafblind/MSI teacher.

This study has been exploratory in nature to build upon the small amount of evidence available from research and the large amount of anecdotal reporting and opinion previously expressed in literature. Sackett et al. (1996) identified the necessity for best medical practice to draw upon both a professional's experiential knowledge and the best external evidence obtained through systematic research. Within the field of the education of learners with CHARGE the challenge must be to consider how to integrate the two and seek to obtain a more even balance between them both by further increasing the amount of evidence available arising from systematic research.

This investigation has also highlighted some areas that would benefit from further research. For example, the aspects found to be of particular note in affecting learning and development of learners with CHARGE including executive dysfunction, fine motor skills and pencil/handwriting skills, the rate of occurrence of sensory integration difficulties (possibly exploring the research methods employed by Miller et al, 1999, with individuals with Fragile X syndrome), and the education of learners with CHARGE in an inclusive mainstream setting. The use of a survey has proved a popular method for generating data about individuals with CHARGE but this is often reliant on participants providing clear and accurate information. It will be important to explore other ways of gathering information, including directly from the affected individuals themselves.

For myself as a researcher, there have been a number of benefits arising from the undertaking of this investigation. It arose from an attempt to begin to find answers for the questions generated through my work with children with CHARGE and to reduce the 'problem space' within this field. It has provided the opportunity for me to discover more and supported the development of my knowledge and understanding of CHARGE. It has also expanded my academic experience and increased my recognition of the importance of systematic research and how this might be undertaken. Undertaking the process has also increased my confidence in exploring research techniques and enabled me to become more analytical in my evaluation of the research of others. Ultimately it has also resulted in an enthusiasm to undertake further research in the future to continue to increase our knowledge and understanding of CHARGE through evidence obtained through systematic research that ultimately may further improve the educational practice with this very special group of people.

APPENDICES

Will my child's part in this research be kept confidential?

Every effort will be made to ensure confidentiality. This will include no use of children's names, staff or school names when publishing the information obtained.

A colleague from the FEAS team will also need to look at the reports and the information I have obtained to ensure that this is a true and accurate picture. I intend to remove any names from the report before it is shared to help maintain some level of confidentiality and anonymity.

How will the information be used?

The information from these reports will be used to support the planning and development of the next stages of the project. At the end of the project this work will form part of the research study I am undertaking. It is likely to be some time before the final report is published, but ongoing work may be submitted for publication before this time.

All data will be stored safely by myself.

Who will the findings be reported to?

Once the whole project is completed it will be presented to the academic body of the University of Birmingham as a written thesis. The findings may also be used for other publications, training days and for presentations of the research itself. I will also write to all the parents who have kindly allowed their children to be a part of the research project, to give a summary of the evidence I have found.

Thank you for sparing the time to read this leaflet. If you have any questions please do contact me.

If you agree to me using your child's reports please remember to sign the enclosed consent form and return it to me in the envelope provided.

Information for parents

The learning environment for children with CHARGE syndrome

Gail Deuce
 Consultant teacher
 Family Education and Advisory Service
Organisation name

Address

Contact email
 Mobile number:

November 2008

Company logo

Dear Parent

My name is Gail Deuce and I work for (*organisation name*) in the Family, Education and Advisory Service (FEAS) as a consultant teacher for deafblind children, with a special interest in children with CHARGE syndrome.

At some time your child will have received input from myself or my colleagues, at school or for an assessment at our centre in xxx (or both!) which will have included written reports.

I am currently carrying out a research project looking at the learning environment for children with CHARGE syndrome as part of a study course I am undertaking with the University of Birmingham. In the first phase of the study I will need to look at educational reports, such as those written for your child, and will be looking for any factors which have helped, or have been recommended, to support learning for children with CHARGE.

I would be grateful if you would allow me to use reports written for your child for this stage of the research. Before you make a decision, please read the rest of this leaflet that will give more details about what this would mean for you and your child. If you are then happy for me to use your child's report(s) please could you complete the enclosed consent form and post it to me in the envelope provided.

I would be more than happy to discuss the project with you or answer any questions you might have and you can contact me either through email, telephone or the address on the front page.

Yours faithfully,
Gail Deuce
Researcher

What is the purpose of the study?

Many children with CHARGE syndrome experience extreme multi-sensory impairment and need to work hard to overcome the daily challenges they face. Through this research I hope to identify what factors are important to these children to support their learning and what is needed to create an optimal learning environment for them.

What will happen?

I will look at reports written by myself and colleagues from the FEAS to identify different aspects of the learning environment that have been recognised as successful or recommended to support more effective learning.

These results will then be used to support the planning and development of the next stages of this research project.

Do I have to take part?

Involvement in this research is completely voluntary. Everyone has the right to choose whether or not to allow reports to be used for this research. If you agree to me using your child's report(s) you are asked to sign a consent form. If, however, you then change your mind, you can ask for the report(s) to be withdrawn from the research at any time and without giving a reason.

Choosing not to be involved, or later choosing to withdraw, will not affect the work undertaken by (*organisation name*) with your child, either now or at any time in the future.

TABLE (i): A CHECK OF RELIABILITY AND VALIDITY TO DISCOVER THE LEVEL OF AGREEMENT BETWEEN THE ASSIGNMENT OF STATEMENTS TO DIFFERENT CODING CATEGORIES- PILOT

Coding categories:	Total number allocated:	Total number of agreements:	Total number of disagreements:	Level of agreement (%) :	Disagreement but agreed in another category:	Expressed as a %	No of total disagreement
CHARGE	3	3	0	100%			
Communication	35	24	11	69%	9/11	82%	2/11
Conceptual development	22	12	10	54%	9/10	90%	1/10
Orientation & mobility	11	9	2	82%	2/2	100%	0
- gross/fine motor	11	9	2	82%	2/2	100%	0
- physical positioning	6	5	1	83%	1/1	100%	0
Ownership of learning	33	19	14	56%	4/14	30%	10/14
- individual learning style	28	21	7	75%	2/7	29%	5/7
Responses to changes & routines	16	11	5	69%	1/5	20%	4/5
Sensory responses							
- hearing & vision	32	22	10	69%	8/10	80%	2/10
- MSI	19	8	11	42%	7/11	64%	4/11
- vestibular & balance	4	4	0	100%			
- other senses	2	2	0	100%			
- SI	22	20	2	91%	1/2	50%	1/2
Social relationships & emotional develop	19	13	6	68%	5/6	83%	1/6
Support for learning							
- Ax & planning	37	29	8	78%	4/4	100%	0
- educational placement	0	0	0				
- ICT	1	1	0	100%			
-prof support	35	30	5	86%	5/5	100%	0
-resources	8	8	0	100%			
Understanding of space & time	20	12	8	60%	5/8	63%	3/8
Total number:	364	262	102	72%	65/98	66%	33/98

**TABLE (ii): A SECOND CHECK OF RELIABILITY AND VALIDITY TO DISCOVER THE LEVEL OF AGREEMENT
BETWEEN THE ASSIGNMENT OF STATEMENTS TO DIFFERENT CODING CATEGORIES**

Coding categories	Total number allocated	Total number of agreements	Total number of disagreements	Level of agreement (%):	Disagreement but agreed in another category:	Expressed as a %	No of total disagreement
CHARGE	4	4	0	100%			
Communication	18	17	1	94%	1/1	100%	
Conceptual development	12	11	1	92%	1/1	100%	
Orientation & mobility	11	11	0	100%			
-Fine motor	1	1	0	100%			
-Physical positioning	7	7	0	100%			
Ownership of learning	13	11	2	85%	1/2	50%	1/2
-Individual learning style	18	17	1	94%	1/2	100%	
Responses to changes & routines	14	12	2	86%	2/2	100%	
Sensory responses	5	5	0	100%			
-Hearing & vision	25	25	0	100%			
-Vestibular & proprioceptive	4	4	0	100%			
-Other senses	2	2	0	100%			
-MSI	2	2	0	100%			
-Sensory integration	16	15	1	94%	1/1	100%	
Social relationships & emotional development	13	10	3	77%	2/3	67%	1/3
Support for learning							
-Ax & planning	28	27	1	96%	1/1	100%	
-physical environment	14	14	0	100%			
-educational placement	0	0	0	100%			
-resources (including ICT)	10	10	0	100%			
-prof support	31	28	3	90%	3/3	100%	
Understanding of space & time	6	6	0	100%			

APPENDIX 3: QUESTIONNAIRE

The learning environment for children with CHARGE syndrome

QUESTIONNAIRE

Before you begin to complete this questionnaire please read the accompanying leaflet entitled "Information for participants: The learning environment for children with CHARGE syndrome."

Please complete and return the questionnaire by **Friday 1 March 2013**.

If you have any queries or need help to complete the questionnaire you can contact Gail at:

Address

Contact email

Mobile number:

Consent

Completion of this form will be regarded as your consent for the content to be used within this research study and that you understand:

The nature of the information to be collected

The purpose of the research

That the information provided within this questionnaire will be securely stored and only accessed by the researcher and other authorised persons

Each questionnaire is to be completed in relation to **one** child with CHARGE syndrome whom you know and have taught/ are teaching (Foundation stage to KS4).

Is a sensory impairment teacher (for visual impairment, hearing impairment, and/or multi-sensory impairment) involved? If so, it would be helpful if the class teacher could complete the questionnaire jointly with the sensory impairment teacher.

Diagnosis

Does the child have a clinical diagnosis of CHARGE syndrome? **(Please circle)**

Yes - Please continue with this questionnaire

No - Please do not continue- this questionnaire is to be completed only for a child with a known clinical diagnosis of CHARGE syndrome

Date questionnaire completed: _____

Child's initials: _____

Child's date of birth: _____

Background information

B1 Type of school child attending: _____

Does the child have:

(Please circle)

B2i Visual impairment?	Yes	No	Don't know
B2ii Hearing impairment?	Yes	No	Don't know
B2iii Vestibular (balance) difficulties?	Yes	No	Don't know
B2iv Poor body awareness/coordination difficulties	Yes	No	Don't know
B2v Touch sensitivity (e.g. a reluctance to use touch to explore or to touch certain textures)	Yes	No	Don't know
B2vi Difficulty using his/her senses together in a coordinated way (e.g. looking and listening at the same time, looking and exploring through touch together)	Yes	No	Don't know
B2vii Difficulty with distractibility and remaining on task	Yes	No	Don't know
B2viii Difficulty with sensory overload/ under-stimulation	Yes	No	Don't know
B2ix High anxiety or stress levels	Yes	No	Don't know
B2x High levels of fatigue	Yes	No	Don't know

B3i Do you consider the child's sensory impairments to be a primary need? (Please circle)

Yes

No

B3ii If 'no', what do you consider to be the child's primary need? _____

The child

Please provide information as requested

Communication

C1 What do you consider to be the main factors within the child that affects his/her communication (e.g. visual impairment, hearing impairment, learning difficulties, distractibility)?

Please list:

C2 What communication modes are used to support communication?

Please tick all that apply:

- Speech
- Sign
- Object cues/ objects of reference
- Touch cues
- Reading child's behaviour
- Photographs
- Symbols and drawings
- Printed word
- Other

Please name: _____

C3 What is the child's preferred communication mode to:

i Receive communication? _____

ii Express him/herself? _____

Within this section some parts include a scale of 1-5 for you to record your answer where 1 = very easy and 5 = very difficult. Please circle the number that most applies.

	Very Easy		Very Difficult		
C4 How easy/difficult is it for the child to <u>initiate</u> communicative exchanges with a sensitive communication partner?	1	2	3	4	5
C5 How easy/difficult is it for the child to <u>maintain</u> communicative exchanges with a sensitive communication partner?	1	2	3	4	5

Social and emotional

C6 With approximately how many adults does the child have a secure and trusting relationship in the school setting? _____

C7 With approximately how many peers does the child have a genuine friendship with in the school setting? _____

Please circle the number that most applies.

	Very Easy					Very Difficult
C8 How easy/difficult is it for the child to develop trust and feel confident with <u>adults</u> in the school setting?	1	2	3	4	5	
C9 How easy/difficult is it for the child to develop genuine friendships with <u>peers</u> in the school setting?	1	2	3	4	5	
C10 How easy/difficult is it for the child to understand their own emotions and express their own emotional state?	1	2	3	4	5	
C11 How easy/difficult is it for the child to empathise with his/her peers when they are hurt, happy, upset, etc.?	1	2	3	4	5	

Conceptual ability

Please circle the number that most applies.

	Very Easy					Very Difficult
C12 How easy/difficult is it for the child to establish real, concrete concepts (e.g. naming real objects, sorting real objects)?	1	2	3	4	5	
C13 How easy/difficult is it for the child to establish abstract concepts (e.g. light/heavy; happy/sad, real/pretend)?	1	2	3	4	5	
C14 How easy/difficult is it for the child to identify solutions to simple problems?	1	2	3	4	5	
C15 How easy/difficult is it for the child to transfer and generalise skills and knowledge and apply it to different situations?	1	2	3	4	5	
C16 How easy/difficult is it for the child to recall a sequence of events (e.g. what happened this morning)?	1	2	3	4	5	
C17 How easy/difficult is it for the child to correctly order parts of an activity or a story sequence?	1	2	3	4	5	

Response to routines and structure, and understanding of time and space
Please circle the number that most applies.

	Very Easy				Very Difficult
C18 How easy/difficult is it for the child to cope with the unexpected and changes to their routines?	1	2	3	4	5
C19 How easy/difficult is it for the child to cope <u>without</u> regular routines and structure?	1	2	3	4	5
C20 How easy/difficult is it for the child to anticipate what is to happen next when appropriate cues are provided?	1	2	3	4	5
C21 How easy/difficult is it for the child to predict what is to happen tomorrow or at the weekend?	1	2	3	4	5
C22 How easy/difficult is it for the child to recall past events?	1	2	3	4	5

C23 Do you use any of the following concrete cues to support the above?
Please tick all that apply:

- Object cues
- Photographs
- Symbols and drawings
- Printed word
- Visual calendar/timetable
- Other

Please name: _____

Ownership of learning
Please circle the number that most applies.

	Very Easy				Very Difficult
C24 How easy/difficult is it for the child to learn and play independently?	1	2	3	4	5
C25 How easy/difficult is it for the child to organise him/herself?	1	2	3	4	5
C26 How easy/difficult is it for the child to initiate activities?	1	2	3	4	5
C27 How easy/difficult is it for the child to undertake self-help skills and functional life skills (appropriate to their age)?	1	2	3	4	5

C28 What factors do you feel adversely affect the child's readiness and ability to engage and learn (e.g. environmental distractions, fatigue, etc.)?

Please list:

Orientation and mobility, and motor skills

C29 Which of the following are factors that you feel affect the child's orientation and mobility?
(Please circle)

Sensory impairments	Yes	No	Don't know
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If yes, please tick those that apply:

- Visual impairment
- Hearing impairment
- Touch
- Vestibular (balance) difficulties
- Other Please name: _____

Body awareness	Yes	No	Don't know
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Coordination and organising body (e.g. changes in position, moving location)	Yes	No	Don't know
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Fatigue	Yes	No	Don't know
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Other	Please name: _____		
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C30 Does the child need to adopt a horizontal position at any time during the school day (e.g. lying down, resting head on table)?

	Yes	No	Don't know
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C31 Does the child seek additional support for their position when standing or sitting (e.g. leaning against the wall, furniture or other people; propping his/her head on his/her hand)?

	Yes	No	Don't know
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C32 Does the child experience fine motor difficulties (e.g. manipulating small objects, using scissors etc.)?

	Yes	No	Don't know
--	-----	----	------------

C33 Does the child demonstrate poor pencil skills/hand-writing skills?

	Yes	No	Don't know
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Assessment

A1 Please tick those types of assessments (and list any others) undertaken or made available to the school in relation to the child's education. Then indicate how helpful you have felt them to be in informing the educational planning for the child with CHARGE syndrome **by circling the number that most applies**.

	(Please circle)				
	Very helpful				Very unhelpful
<input type="checkbox"/> Functional use of vision	1	2	3	4	5
<input type="checkbox"/> Functional use of hearing	1	2	3	4	5
<input type="checkbox"/> Communication	1	2	3	4	5
<input type="checkbox"/> Sensory integration	1	2	3	4	5
<input type="checkbox"/> Hand function	1	2	3	4	5
<input type="checkbox"/> Orientation and mobility	1	2	3	4	5
<input type="checkbox"/> Eating and drinking	1	2	3	4	5
<input type="checkbox"/> Seating	1	2	3	4	5
<input type="checkbox"/> ICT	1	2	3	4	5
<input type="checkbox"/> Other. Please list:					
<input type="checkbox"/>	1	2	3	4	5
<input type="checkbox"/>	1	2	3	4	5
<input type="checkbox"/>	1	2	3	4	5
<input type="checkbox"/>	1	2	3	4	5
<input type="checkbox"/>	1	2	3	4	5

Strategies

Under each section **please tick** those strategies (**and list any others**) you have found helpful in supporting the child with CHARGE syndrome.

S1 Sensory

- Developing profiles of functional use of vision and hearing
- Use of visual/auditory aids
- Differentiating materials/activities in relation to child's sensory needs
- Activities planned/implemented in relation to child's sensory integration difficulties
- Appropriate behaviour management with recognition of impact of sensory integration difficulties
- Activities to support the development of haptic perception (active use of touch)
- Undertaking an environmental audit in relation to child's sensory needs
- Access to a safe rest area
- Other

Please list:

S2 Communication

- Development and implementation of an individual communication programme
- Use of a Total Communication approach that includes:
 - Touch cues
 - Spoken word
 - Printed word
 - Object cues/ objects of reference
 - Other
- Photographs
- Sign
- Symbols and drawings
- Providing a sensitive communication partner who is able to recognise and respond to the child's attempts to communicate
- Ensuring access to a peer group with an appropriate level of communication skills
- Undertaking an environmental audit in relation to child's communication needs
- Other

Please list:

Please tick those strategies (and list any others) you have found helpful in supporting the child with CHARGE syndrome.

S3 Social and emotional

- Key people identified to develop secure relationships with the child
- Implementation of a structured programme to develop the child's understanding of emotions
- Use of a structured programme/activities to support interactions with peers
- Support to learn how to initiate/sustain social interaction sequences
- Use of a defined programme to manage stress and anxiety levels
- Undertaking an environmental audit in relation to child's social and emotional needs
- Other

Please list:

S4 Conceptual ability

- Use of structured fading and pre-learning/ post-learning to support learning
- Monitoring to identify the risk of the development of 'splinter' skills and responding
- Provision of real experience provided within context
- Providing a balance between new and familiar activities
- Use of identified strategies to support the development of problem-solving skills
- Use of concrete tools (e.g. real objects, photographs, symbols etc.)
- Other

Please list:

Please tick those strategies (and list any others) you have found helpful in supporting the child with CHARGE syndrome.

S5 Response to routines and structure and understanding of time and space

- Use of a daily routine and structure implemented consistently throughout the day
- Use of mini routines
- Use of concrete tools to support sequencing an activity, transitions and daily routine
- Individualised pacing (may include a reduced timetable)
- Ensuring activities have a clear beginning, middle and end
- Structured support for transitions
- Other

Please list:

S6 Ownership of learning

- Clear identification of what is motivational for the child
- Providing frequent choice-making opportunities
- Implementation of a structured programme to support self-help and self-organisational skills
- Clear identification of child's individual learning style informing the delivery of activities
- Use of a flexible approach, responding to child's changing needs and levels of responsiveness
- A structured response to manage the impact of fatigue
- Responding to child's executive function difficulties
- Other

Please list:

Please tick those strategies (**and list any others**) you have found helpful in supporting the child with CHARGE syndrome.

S7 Orientation and mobility and motor skills

- Structured activities to support the development of gross motor skills and body awareness
- Providing regular opportunities to engage in large movements
- Use of large movement play activities
- Implementation of a fine motor skills programme
- Implementation of a pencil skills programme
- Provision of alternative recording methods (e.g. scribe, use of IT, etc.)
- Use of specialist/supportive seating
- Undertaking an environmental audit in relation to child's orientation and mobility needs
- Other

Please list:

Professional support

P1 Please tick those professionals (and list any others) who have contact with the child with CHARGE syndrome in the educational setting.

- Specialist multi-sensory impairment teacher
- Specialist visual impairment teacher
- Specialist hearing impairment teacher
- Intervenor
- Occupational therapist
- Physiotherapist
- Sensory Integration specialist
- SaLT- communication
- SaLT- dysphagia (eating and drinking difficulties)
- Mobility rehabilitation officer
- ICT specialist
- Educational psychologist
- Other

Please list:

P2 What input/support have you had regarding CHARGE syndrome? (E.g. websites, MSI input, training, etc.).

P3 What else do you feel might be needed?

Thank you for taking the time to complete this questionnaire.

Please return in the envelope provided to:

**Gail Deuce
Principal MSI Consultant
*Address***

<p>How will the information be used? The information from this questionnaire will be used to establish and identify factors that are most likely to support learning for children with CHARGE syndrome. It is likely to be some time before the final report is published, but ongoing work may be published before this time. All data will be stored safely by myself.</p> <p>Who will the findings be reported to? Once the project is completed it will be presented to the academic body of the University of Birmingham as a written thesis. The findings may also be used for other publications, training days and for presentations of the research itself.</p> <p>I also intend to prepare a summary of the findings of this study to share with educational practitioners and parents that will be made available in due course on the (<i>organisation name</i>) website.</p> <p>How long will it take to complete the questionnaire? The average time taken to complete the pilot of this questionnaire was 30 minutes.</p> <p>Although there are a number of pages, answering the majority of the questions involves a tick list, choosing a number on a scale or circling a yes/no/don't know response.</p> <p>Thank you for sparing the time to read this leaflet. If you have any questions please do contact me.</p> <p>I am very grateful for your time and willingness to share your experience and knowledge of teaching a child with CHARGE syndrome– your involvement will help make a difference and improve the support we can give.</p>	<p style="text-align: center;">Information for participants</p> <p style="text-align: center;">The learning environment for children with CHARGE syndrome</p> <p>Gail Deuce Principal MSI Consultant Children's Specialist Services</p> <p><i>Address</i></p> <p><i>Contact email</i> Mobile number:</p> <p>January 2013 <i>Company logo</i></p>
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Introduction

My name is Gail Deuce and I work for (*organisation name*) in the Children's Specialist Services as a Principal MSI Consultant teacher, with a special interest in children with CHARGE syndrome. I am currently undertaking a PhD at the University of Birmingham and carrying out a research project looking at the learning environment for children with CHARGE syndrome. During the first phase of the study I gathered and analysed data looking for any factors that have been identified as helpful or have been recommended to support learning for children with CHARGE.

In this second phase I have developed a questionnaire as a result of the findings of Phase 1. I am now approaching teachers who are teaching, or have recently taught, a child with a medical diagnosis of CHARGE syndrome (Foundation stage to Key Stage 4), to ask if they would be willing to complete this questionnaire.

You have been identified as a teacher who may be teaching, or have recently taught (within the last 5 years), a child with CHARGE, and I would be grateful if you would consider completing a questionnaire. If you are a sensory impairment teacher (for visual impairment, hearing impairment, and/or multi-sensory impairment) involved it would be helpful if you could complete the questionnaire jointly with the class teacher. Before you make a decision, please read the rest of this leaflet. If you are then happy to complete the accompanying questionnaire please could you do so and post it to me in the envelope provided. I would be more than happy to discuss the project with you or answer any questions you might have and you can contact me either through email, telephone or the address on the front page.

Yours faithfully,
Gail Deuce (Researcher)

What is the purpose of the study?

Many children with CHARGE syndrome experience extreme multi-sensory impairment and need to work hard to overcome the daily challenges they face. Through this research I hope to identify what factors are important to these children to support their learning and what is needed to create an optimal learning environment for them.

What will happen?

I will collate and analysis the information obtained from the completed questionnaires. It is hoped that the findings will identify how educational practitioners might most effectively support the learning for a child or young person with CHARGE syndrome.

The longer term aim is to provide teachers with knowledge and a resource that is of practical use when supporting learners with CHARGE syndrome.

Do I have to take part?

Involvement in this research is completely voluntary. Completion of a questionnaire will be considered as your consent for the information obtained to be used within this research study.

Every effort will be made to ensure confidentiality and anonymity. Questionnaires will be coded on the front page which will be removed and logged by an independent person when the questionnaire is returned to reduce the likelihood of the researcher identifying the respondent when collating the data from each completed questionnaire. There will be no use of participants' names when sharing and publishing the information obtained. Similarly, the child's date of birth and initials are requested solely to ensure that only one questionnaire is processed per child. These details will not be used to identify the child.

**APPENDIX 5: USING SPEARMAN'S RHO (RANK CORRELATION COEFFICIENT)
TO ASCERTAIN THE CONSISTENCY IN RESPONSES GIVEN TO QUESTIONS
C2 AND S2**

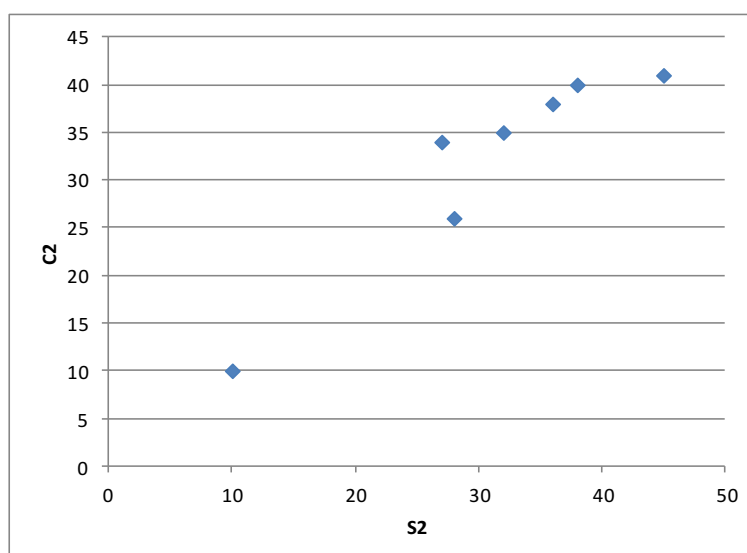
The total scores for each of the communication modes listed under each question are set out in the table below.

The total score for communication modes listed in
questions C2 and S2

	C2	S2
Touch	10	10
Printed word	27	34
Object cues/ objects of reference	28	26
Symbols/ drawings	32	35
Photographs	36	38
Sign	38	40
Speech	45	41

When plotted on a scattergram graph the results suggested a positive correlation (i.e. there is a relationship between the two sets of responses obtained) as shown in the following figure.

The results of Table x plotted as a scattergram of the two sets of responses
obtained to questions C2 and S2



Following this the data was then cast into a table as shown below in the following Table, considering the total scores for each mode identified and their rankings overall within the answers for each question.

The total scores obtained for modes listed in responses to questions C2 and S2, with rankings

	A: C2	A: S2	Rank A	Rank B	Difference	D ²
Touch	10	10	1	1	0	0
Printed word	27	34	2	4	2	4
Object cues/ objects of reference	28	26	3	2	1	1
Symbols/drawings	32	35	4	3	1	1
Photographs	36	38	5	5	0	0
Sign	38	40	6	6	0	0
Speech	45	41	7	7	0	0

Spearman's rho was then calculated as follows:

N= number of paired scores. i.e: 7

Multiply N by its own value twice. Then subtract its own value.

$$(7 \times 7 \times 7) - 7 = 343 - 7 = 336$$

Total of values in D²

$$0 + 4 + 1 + 1 + 0 + 0 + 0 = 6$$

Multiply the sum of D² by 6, then divide by the score from step 2 (i.e. 336)

$$(6 \times 6) \div 336 = 36 \div 336 = 0.107$$

$$\text{Rho} = 1 - 0.107 = +0.893$$

$$\text{Reliability} = \frac{2r}{1+r} = \frac{2(0.893)}{1+0.893} = \frac{1.786}{1.893} = 0.94$$

Overall, the level of significance was therefore found to be 0.94.

APPENDIX 6- INTERVIEW INFORMATION LETTER/CONSENT FORM

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My name is Gail Deuce. I work for (organisation name) in the Children's Specialist Services in the UK as a Principal MSI Consultant teacher for deafblind children, with a special interest in children with CHARGE syndrome.

I am currently undertaking a PhD at the University of Birmingham and carrying out a research project looking at the learning environment for children with CHARGE syndrome. I am hoping to gain a better understanding and identification of what is needed to support effective learning for children and young people with CHARGE that will be of benefit and practical use for educational practitioners working in this field.

I would like to gain a more international perspective on my research and tap in to the knowledge and experience that already exists at Perkins School. I would be very grateful if you would be willing to be interviewed for this purpose. I intend to record the discussions to enable me to check the content of my notes from the interviews are accurate. These recordings will not be made available to anyone else and will be stored safely for the duration of my research study. Every effort will be made to ensure confidentiality and anonymity. This will include no use of participants' names when sharing and publishing the information obtained.

Involvement is completely voluntary. If after the interview you feel you would like to withdraw any statements made please contact me and I will ensure that this information is removed from the data obtained.

I do hope you will feel able to take part in this study. If you are happy to be involved please could you sign the consent form below and return it to myself.

With many thanks,

Gail

Email address

PhD study: The learning environment for children with CHARGE syndrome
Researcher: Gail Deuce

I am willing to participate in interviews and give consent for the information obtained to be used for the purpose of this research study.

Signed:

Date:

APPENDIX 7: THE CHILD

TABLE (iii): A SUMMARY OF THE INTERNAL FACTORS IDENTIFIED FOR EACH CODING CATEGORY

CHARGE	
True and extreme MSI Unique combination of anomalies in each child Sensory integration issues	Speech difficulties Feeding difficulties Stress and anxiety Behavioural difficulties
SENSORY RESPONSES: General	
'Input impaired' Fatigue	Sensory impairments- requiring stronger stimulation
SENSORY RESPONSES: MSI	
True and extreme MSI- all senses likely to be affected MSI- primary need Fatigue	MSI- affecting ability to learn Needing additional time to take in information through the senses
SENSORY RESPONSES: Hearing and Vision	
Combined VI/HI Functional use of vision and hearing Time to acquire visual/auditory information	Fatigue Impact of other factors on ability to use residual vision/hearing
SENSORY RESPONSES: Vestibular and Proprioceptive	
Vestibular difficulties	Proprioceptive difficulties
SENSORY RESPONSES: Other senses	
Use of touch/haptic perception to explore tactually Reduced sense of smell and taste	Time to acquire information through use of touch
SENSORY RESPONSES: Sensory Integration	
Sensory Integration difficulties affecting learning and behaviour Sensory overload and anxiety	Fatigue Difficulty with self-regulation and remaining well- modulated
COMMUNICATION	
Difficulty communicating with others Visual/auditory limitations Time to receive and process information communicated	Fatigue Impact of other factors Current level of communication skills Preferred communication modes

CONCEPTUAL DEVELOPMENT	
'Construction'/ understanding of the world Current concept development Consolidation of understanding and ability to transfer knowledge and skills	Time to take in, process and formulate a response Executive function difficulties Distractability Developing 'splinter' skills Problem solving skills
ORIENTATION, MOBILITY AND MOVEMENT: General	
Gross motor skills Vestibular/proprioceptive difficulties Difficulty moving around the environment	Conflicting demands- putting skills on hold whilst learning/practising physical skills Body awareness
ORIENTATION, MOBILITY AND MOVEMENT: Fine Motor	
Other sensory impairments/difficulties Poor proprioception	Level of fine motor skill development Poor pencil skills/ writing skills
ORIENTATION, MOBILITY AND MOVEMENT: Physical positioning	
Vestibular, proprioceptive and sensory integration difficulties Difficulty working against gravity	Use of compensatory strategies, including adopting different positions
OWNERSHIP OF LEARNING: General	
Active learner? Level of confidence Motivation Control of pace of engagement in an activity	Able to make needs known? Involvement in decision making/ planning own programme Level of self-organisational/self-help skills
OWNERSHIP OF LEARNING: Individual learning style	
Burst-pause sequence to learning Additional time to take in and process information Taking time before ready to engage in a new /unfamiliar environment Engaging in physical movement to support concentration	Other factors affecting ability to use their senses (conflicting demands, familiarity of activity, additional cues, fatigue, working position) Executive function difficulties Fatigue
RESPONSES TO ROUTINES AND CHANGES	
Is the child experiencing conflicting sensory demands? Response to different environments/ people	Impact of changes to routine on the child's sensory system

SOCIAL RELATIONSHIPS AND EMOTIONAL DEVELOPMENT	
Bonding and attachment: Forming trusting relationships with intervenor and other key people Development of early social skills: Eye contact Shared attention Turn-taking Ability to initiate/sustain social interactions with peers Understanding of emotions	Self-awareness and self-esteem Experiencing success- building confidence Facial palsy- awareness of social implications Anxiety levels Early life experiences Psychological impact of CHARGE /MSI
UNDERSTANDING OF TIME AND SPACE	
Burst-pause learning style Understanding gained from routine/ structure	Ability to anticipate what is to happen next

TABLE (iv): A SUMMARY OF THE EXTERNAL FACTORS IDENTIFIED FOR EACH CODING CATEGORY

CHARGE	
<ul style="list-style-type: none"> ○ Individualised response ○ Raising awareness and knowledge of CHARGE, possible implications and the impact on learning/development and everyday living 	<ul style="list-style-type: none"> ○ Training on the nature and implications of CHARGE. ○ Input from an MSI teacher with knowledge/experience of CHARGE
SENSORY RESPONSES: General	
<ul style="list-style-type: none"> ○ Placing a priority on sensory needs ○ Ensuring against sensory 'overload' 	<ul style="list-style-type: none"> ○ Working at child's pace ○ An environmental audit
SENSORY RESPONSES: MSI	
<ul style="list-style-type: none"> ○ Recognition of challenges created by MSI ○ Input from qualified MSI teacher ○ Provision of information for parents about MSI 	<ul style="list-style-type: none"> ○ Training on the nature and implications of deafblindness/MSI ○ Educational programme to address needs as MSI learner
SENSORY RESPONSES: Hearing and Vision	
<ul style="list-style-type: none"> ○ Use of visual/auditory aids ○ Profiles of functional vision and hearing ○ To inform planning/development of educational programme 	<ul style="list-style-type: none"> ○ An appropriate visual/auditory environment ○ Differentiating activities/materials in respect of child's VI/HI ○ Input from qualified teachers for VI/HI
SENSORY RESPONSES: Vestibular and Proprioceptive	
<ul style="list-style-type: none"> ○ Recognition of vestibular and proprioceptive difficulties and impact on learning ○ Providing small 'fiddles' for use when sitting still 	<ul style="list-style-type: none"> ○ Activities planned/implemented in response to vestibular and proprioceptive issues
SENSORY RESPONSES: Other senses	
<ul style="list-style-type: none"> ○ Activities to support development of haptic perception 	
SENSORY RESPONSES: Sensory Integration	
<ul style="list-style-type: none"> ○ Recognition of sensory integration difficulties and impact on learning, and an appropriate response ○ Appropriate behaviour management with recognition of impact of SI difficulties ○ Access to a specialist SI OT/ Physio 	<ul style="list-style-type: none"> ○ Development of a sensory integration programme, including a sensory diet ○ Development of appropriate management strategies ○ An appropriate environment to reduce sensory overload, conflicting demands etc. ○ Access to a safe rest area

COMMUNICATION	
<ul style="list-style-type: none"> ○ A sensitive communication partner ○ Contingent responses to communication attempts ○ Providing support for understanding and self expression ○ Individual communication programme ○ Identification and use of appropriate strategies and resources ○ Use of a concrete component to overcome executive function difficulties ○ Recognition of child's preferred communication modes 	<ul style="list-style-type: none"> ○ An environmental audit to ensure a rich communication environment ○ A Total Communication environment ○ A signing environment. ○ A peer group with appropriate communication skills. ○ Access to sign language and appropriately skilled adults. ○ Consistency between home and school. ○ SALT advice and support ○ MSI teacher advice and support
CONCEPTUAL DEVELOPMENT	
<ul style="list-style-type: none"> ○ Provision of relevant and meaningful experiences provided within context ○ Balance between new/familiar activities ○ Opportunities to explore ○ Activities to support concept development, problem solving and transference of skills/ knowledge 	<ul style="list-style-type: none"> ○ Monitoring to protect against development of 'splinter' skills ○ Use of identified strategies including structured fading, pre/over learning, following the child's interest ○ Use of strategies to keep child on task ○ Use of concrete components
ORIENTATION, MOBILITY AND MOVEMENT: General	
<ul style="list-style-type: none"> ○ Recognition of vestibular and proprioceptive difficulties ○ Activities to support development of gross motor skills and body awareness ○ Regular opportunities to engage in large movements ○ Large movement play activities 	<ul style="list-style-type: none"> ○ Input from physio/ OT ○ Support for independent movement ○ Time to explore and become familiar with new environments ○ Environmental audit- orientation and mobility ○ Mobility rehabilitation officer ○ Check whether is able to cross midline effectively
ORIENTATION, MOBILITY AND MOVEMENT: Fine Motor	
<ul style="list-style-type: none"> ○ Activities to support fine motor skill development and coordination ○ Assessment of hand function 	<ul style="list-style-type: none"> ○ Provision of activities to support development of pencil/writing skills ○ Provision of alternative recording methods ○ Advice and support from physio/OT
ORIENTATION, MOBILITY AND MOVEMENT: Physical positioning	
<ul style="list-style-type: none"> ○ Opportunities to adopt a horizontal position 	<ul style="list-style-type: none"> ○ Active seating/ supportive seating ○ Provision of a secure physical base/

<ul style="list-style-type: none"> ○ Provision for adopting different working positions 	<ul style="list-style-type: none"> ○ secure base for a child's feet ○ Advice and support from an OT
OWNERSHIP OF LEARNING: General	
<ul style="list-style-type: none"> ○ Sharing control with the child/ supporting active involvement ○ Choice-making opportunities. ○ Identification of what is motivational ○ Provision of activities that ensure success/ build confidence ○ Careful time management 	<ul style="list-style-type: none"> ○ Individualised pacing- at child's pace ○ Strategies to enable the child to make needs known ○ Involving child in decision making/ planning ○ Support to develop self-organisational and self-help skills
OWNERSHIP OF LEARNING: Individual learning style	
<ul style="list-style-type: none"> ○ Recognition of child's individual learning style leading to an appropriate response/ strategies ○ Pace child-led ○ Following child's lead and preferences ○ Flexible approach responding to child's changing needs/ recognition of levels of responsiveness ○ Not overloading with information and time given to assimilate it ○ Not placing too many demands on child when a 'difficult' day 	<ul style="list-style-type: none"> ○ Balance of familiar/unfamiliar activities ○ Opportunities for repletion/to return to an activity ○ Awareness of impact of different working positions ○ Recognition of/strategies to manage impact of fatigue ○ Recognition of executive function difficulties ○ Use of modelling to support learning
RESPONSES TO ROUTINES AND CHANGES	
<ul style="list-style-type: none"> ○ Awareness of impact of changes to routine on child ○ Awareness of child's response to different environments/people ○ Provision of additional time for new environments/people ○ Provision of structure and routines, implemented consistently throughout the school day ○ Use of mini routines 	<ul style="list-style-type: none"> ○ Routines used consistently in different settings ○ Regular contact between home/school to ensure continuity of approach ○ Use of concrete cues to support sequencing an activity, transitions, and daily routine ○ A balance between new and familiar activities
SOCIAL RELATIONSHIPS AND EMOTIONAL DEVELOPMENT	
<ul style="list-style-type: none"> ○ Intervenor support ○ Key people with whom strong, trusting relationships are formed ○ Intervenor to support development of early social skills ○ Structured programme/activities to support peer interactions ○ Support to learn how to 	<ul style="list-style-type: none"> ○ similar sensory impairments / communication needs ○ Support to develop understanding of emotions ○ Activities to ensure success and therefore build confidence ○ Support to overcome issues relating to facial palsy

<p>initiate/sustain interactions</p> <ul style="list-style-type: none"> ○ Access to a peer group with appropriate communication skills ○ Opportunities to meet others with 	<ul style="list-style-type: none"> ○ Support for staff- recognition of impact of early life experiences; high levels of anxiety; psychological impact of CHARGE/MSI
UNDERSTANDING OF TIME AND SPACE	
<ul style="list-style-type: none"> ○ Activities proceeding at child's pace ○ Careful time management and individualised pacing (may include reduced timetable) ○ Clear structure and routines ○ Tasks to have an identifiable structure with clear beginnings/ endings 	<ul style="list-style-type: none"> ○ Use of a visual communication timetable ○ Concrete tools to prepare for transitions ○ Continuity between home/school and sharing of information to support conversations

TABLE (v): LIKERT SCALE SCORES FOR QUESTIONS ON COMMUNCIATIVE EXCHANGES WITH A SENSITIVE COMMUNICATION PARTNER AND DISTRIBUTION AMONG TYPES OF PROVISION AND EDUCATIONAL PHASES

			SLD/ PMLD (N=21)	Main- stream (N=16)	Other specialist (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)	Total scores	Average (mean) of total scores
How easy/difficult is it for the child to <u>INITIATE</u> communicative exchanges with a sensitive communication partner?	5= very difficult	1	4	3	5	3	3	3	3	131	2.5
		2	9	5	3	4	4	3	6		
		3	3	3	4	2	3	4	2		
		4	5	4	1	3	4	1	1		
		5	-	1	2	1	-	1	1		
	Average		2.4	2.7	2.5	2.6	2.6	2.5	2.3		
How easy/difficult is it for the child to <u>MAINTAIN</u> communicative exchanges with a sensitive communication partner?	1= very easy	1	2	2	4	2	3	1	2	156	3
		2	7	3	2	4	1	3	4		
		3	4	4	4	1	6	2	4		
		4	7	3	2	3	2	4	2		
		5	1	4	3	3	2	2	1		
	Average		2.9	3.3	2.9	3.1	2.9	3.3	2.7		

TABLE (vi): NUMBER OF ADULTS WITH WHOM THE CHILD IS REPORTED TO HAVE A TRUSTING AND SECURE RELATIONSHIP/ NUMBER OF PEERS WITH WHOM THE CHILD IS REPORTED TO HAVE A GENUINE FRIENDSHIP IN SCHOOL

		Total number of children (N=52)	SLD/PMLD (N=21)	Main-stream (N=16)	Other specialist (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Number of adults with whom child is felt to have a trusting and secure relationship in school	0	1	1	-	-	-	1	-	-
	1	3	1	1	1	2	1	-	-
	2	5	3	1	1	2	2	1	-
	3	8	3	3	2	3	2	1	2
	4	8	6	1	1	2	3	2	1
	5	3	1	1	1	1	-	1	1
	6	11	4	2	5	2	2	4	3
	7	1	-	1	-	-	-	1	-
	8	2	-	1	1	-	-	1	1
	9	2	1	-	1	1	1	-	-
	10	3	-	2	1	-	-	1	2
>10	5	1 (16)	3 (12,14,15)	1 (20)	-	2 (14,16)	-	3 (12,15,20)	
Average		5.73	4.5	6.8	6.3	3.8	5.3	5.58	8.3
Number of peers with whom child is felt to have a genuine friendship in school	0	25	14	5	6	4	9	5	7
	1	5	3	2	1	1	1	3	-
	2	7	4	2	2	3	1	1	2
	3	5	3	1	2	2	1	-	2
	4	2	-	1	1	-	-	-	2
	5	4	-	2	2	1	1	1	1
	6	3	-	3	-	1	1	1	-
	7	0	-	-	-	-	-	-	-
	8	0	-	-	-	-	-	-	-
	9	0	-	-	-	-	-	-	-
	10	1	-	-	1	1	-	-	-
	>10	0	-	-	-	-	-	-	-
Average		1.73	1.0	2.6	1.7	1.5	1.2	1.3	1.8

TABLE (vii): LIKERT SCLAE SCORE FOR QUESTIONS RELATING TO THE DEVELOPMENT OF RELATIONSHIPS AND EMOTIONAL DEVELOPMENT

		SLD/ PMLD (N=21)	Main- stream (N=16)	Other specialist (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)	Total scores	Average (mean) of total scores	
How easy/difficult to develop trust and confidence with adults in school setting?	5= very difficult	1	6	4	4	4	3	3	126	2.4	
		2	5	6	2	4	2	4			
		3	6	4	5	3	6	4			
		4	3	2	4	2	3	-			4
		5	1	-	-	-	-	1			-
Average		2.4	2.3	2.6	2.2	2.8	2.3	2.5			
How easy/difficult to develop genuine friendships with peers in school setting?	5= very difficult	1	-	-	1	-	1	-	213	4.1	
		2	-	1	3	1	-	1			2
		3	1	5	3	5	1	2			1
		4	7	5	1	5	4	1			3
		5	13	5	7	2	8	8			7
Average		4.6	3.9	3.7	3.6	4.3	4.3	4.2			
How easy/difficult to understand their own emotions & express their own emotional state?	1= very easy	1	-	-	1	-	-	-	205	3.9	
		2	2	1	1	-	1	-			3
		3	2	5	3	4	3	2			1
		4	7	5	1	6	3	2			2
		5	10	5	7	3	7	8			6
Average		4.2	3.9	3.4	3.9	4.1	4.5	3.7			
How easy/difficult to empathise with his/her peers when they are hurt, angry, upset etc.?	1= very easy	1	-	1	1	-	-	-	213	4.1	
		2	1	1	1	-	1	-			2
		3	2	5	4	5	3	2			1
		4	-	8	-	3	2	1			2
		5	18	1	9	5	8	9			6
Average		4.7	3.4	4.0	4.0	4.2	4.6	3.6			

TABLE (viii): LIKERT SCALE SCORES FOR QUESTIONS RELATING TO THE CHILD'S CONCEPTUAL ABILITY PART 1

		SLD/ PMLD provision (N=21)	Main- stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)	Total scores	Average (mean) of total scores	
How easy/difficult is it for the child to establish real, concrete concepts?	5 = very difficult	1	5	8	6	3	8	2	6	119	2.3
		2	7	3	2	3	2	4	3		
		3	3	3	6	3	1	5	3		
		4	3	1	1	2	3	-	-		
		5	3	1	-	2	-	1	1		
Average		2.6	2.0	2.1	2.8	1.9	2.5	2.0			
How easy/difficult is it for the child to establish abstract concepts?	5 = very difficult	1	-	-	1	-	1	-	-	188	3.6
		2	2	7	4	3	5	1	4		
		3	4	3	3	3	2	1	4		
		4	3	3	3	2	3	3	1		
		5	12	3	4	5	3	7	4		
Average		4.2	3.1	3.3	3.7	3.1	4.3	3.4			
How easy/difficult is it for the child to identify solutions to simple problems?	1 = very easy	1	1	1	2	-	-	1	3	166	3.2
		2	5	1	2	4	2	1	1		
		3	8	7	6	3	7	5	6		
		4	3	6	3	5	4	1	2		
		5	4	1	2	1	1	4	1		
Average		3.2	3.3	3.1	3.2	3.2	3.5	2.8			

TABLE (ix): LIKERT SCALE SCORES FOR QUESTIONS RELATING TO THE CHILD'S CONCEPTUAL ABILITY PART 2

		SLD/ PMLD provision (N=21)	Main- stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)	Total scores	Average (mean) of total scores	
How easy/difficult is it for the child to transfer and generalise skills and knowledge and apply it to different situations?	5= very difficult	1	-	1	-	-	-	2	188	3.6	
		2	2	2	1	1	1	1			
		3	6	6	4	5	5	4			2
		4	9	6	5	4	5	5			6
		5	5	2	3	3	3	2			2
	Average		3.8	3.5	3.5	3.7	3.7	3.7	3.4		
How easy/difficult is it for the child to recall a sequence of events?	5= very easy	1	2	3	3	2	1	1	171	3.3	
		2	2	4	4	2	4	1			3
		3	4	3	1	1	3	3			2
		4	4	5	2	3	2	3			2
		5	9	1	5	5	4	4			2
	Average		3.3	2.8	3.1	3.5	3.3	3.4	2.6		
How easy/difficult is it for the child to correctly order parts of an activity or story sequence?	1= very easy	1	1	2	3	1	-	1	171	3.3	
		2	1	5	6	6	4	1			1
		3	2	4	3	-	4	1			5
		4	7	4	-	2	3	3			2
		5	10	1	3	4	3	6			1
	Average		4.1	2.8	2.6	2.2	3.4	4.0	2.6		

TABLE (x): LIKERT SCALE SCORES FOR QUESTIONS RELATING TO CHILD'S REPOSSES TO ROUTINES AND STRUCTURE, AND UNDERSTANDING OF SPACE AND TIME PART 1

			SLD/ PMLD provision (N=21)	Main- stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)	Total scores	Average (mean) of total scores
How easy/difficult is it for the child to cope with the unexpected & changes in their routine?	5= very difficult	1	1	1	2	1	-	1	2	183	3.5
		2	3	5	1	-	5	3	1		
		3	5	4	3	6	2	2	2		
		4	6	2	2	1	3	2	4		
		5	6	4	7	5	4	4	4		
Average			3.6	3.2	3.7	3.7	3.4	3.4	3.5		
How easy/difficult is it for the child to cope <u>without</u> regular routines & structure?	5= very difficult	1	-	-	-	-	-	-	-	208	4
		2	3	2	2	1	3	1	2		
		3	2	4	2	1	3	3	1		
		4	7	3	5	5	2	3	5		
		5	9	7	6	6	6	5	5		
Average			4.0	3.9	4.0	4.2	3.8	4.0	4.0		
How easy/difficult is it for child to anticipate what is to happen next when appropriate cues provided?	1= very easy	1	5	1	2	3	1	2	2	120	2.3
		2	11	9	7	6	8	5	8		
		3	3	3	4	2	3	5	-		
		4	2	3	2	2	2	-	3		
		5	-	-	-	-	-	-	-		
Average			2.1	2.5	2.4	2.2	2.4	2.3	2.3		

TABLE (xi): LIKERT SCALE SCORES FOR QUESTIONS RELATING TO CHILD'S REPOSSES TO ROUTINES AND STRUCTURE, AND UNDERSTANDING OF SPACE AND TIME PART 2

		SLD/ PMLD provision (N=21)	Main- stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)	Total scores	Average (mean) of total scores	
How easy/difficult is it for the child to predict what is to happen tomorrow or at the weekend?	5 = very difficult	1	-	2	2	1	-	1	2	181	3.5
		2	2	3	3	3	1	1	3		
		3	7	5	4	3	3	6	4		
		4	1	3	3	1	2	1	3		
		5	11	3	3	5	8	3			
Average		4.0	3.1	3.1	3.5	4.2	3.3	2.5			
How easy/difficult is it for the child to recall past events?	1 = very easy	1	1	1	5	1	2	-	4	166	3.2
		2	2	9	2	3	4	3	3		
		3	6	2	2	1	4	3	2		
		4	3	1	3	1	1	3	2		
		5	9	3	3	7	3	3	2		
Average		3.8	2.8	2.8	3.8	2.9	3.5	2.6			

TABLE (xii): LIKERT SCALE SCORES FOR QUESTIONS RELATING TO OWNERSHIP OF LEARNING

379

		SLD/ PMLD provision (N=21)	Main- stream provision (N=16)	Other specialist provision (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)	Total scores	Average (mean) of total scores	
How easy/difficult is it for the child to learn and play independently?	5 = very difficult	1	1	1	1	1	-	1	179	3.4	
		2	2	4	3	3	2	2			
		3	9	2	4	5	3	3			4
		4	7	2	3	-	4	5			3
		5	2	7	4	4	4	2			3
Average		3.3	3.6	3.4	3.5	3.6	3.6	3.2			
How easy/difficult is it for the child to organise him/herself?	5 = very difficult	1	-	-	1	-	-	-	1	197	3.8
		2	1	2	2	-	1	-	4		
		3	3	4	5	3	3	4	2		
		4	11	5	4	5	7	4	4		
		5	6	5	3	5	3	4	2		
Average		4.0	3.8	3.4	4.2	3.9	4.0	3.2			
How easy/difficult it for the child to initiate activities?	1 = very easy	1	-	1	1	-	-	1	1	184	3.5
		2	2	-	5	2	3	1	1		
		3	10	4	1	6	2	5	2		
		4	5	8	4	5	5	1	6		
		5	4	3	4	-	4	4	3		
Average		3.5	3.8	3.3	4.2	3.7	3.5	3.7			
How easy/difficult it for the child to undertake self-help skills and functional life skills (appropriate for age)?	1 = very easy	1	-	-	2	-	-	-	2	184	3.5
		2	3	1	2	2	-	2	2		
		3	6	5	5	3	4	5	4		
		4	7	6	5	6	9	2	1		
		5	5	4	1	2	1	3	4		
Average		3.7	3.8	3.0	3.6	3.8	3.5	3.2			

TABLE (xiii): FACTORS CONSIDERED TO ADVERSELY AFFECT THE CHILD'S READINESS AND ABILITY TO ENGAGE AND LEARN

Factors	Number where reported (N=52)	SLD/ PMLD (N=21)	Main-stream (N=16)	Other specialist (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Environmental distractions	24	9	8	7	7	5	6	6
Fatigue	22	9	8	5	6	8	3	5
Motivation, interest & concentration	14	7	3	4	3	7	4	-
Communication difficulties/lack of understanding	9	3	3	3	2	3	3	1
Sensory impairments & needs	8	2	4	2	4	3	-	1
Mood/Attitude/Emotional state	7	4	1	2	2	3	-	2
Physical state- including health issue, hunger, pain, sleep	6	3	2	1	2	2	1	1
Lack of routine/ changes to routines/transitions	6	4	-	2	1	1	2	2
Lack of familiar or consistent adults	4	2	-	2	-	1	1	2
Behaviour/OCD's	4	4	-	-	1	2	1	-
Sensory integration difficulties/ regulation	4	-	2	2	-	2	2	-
Pace of delivery of curriculum, time to process information	4	-	3	1	-	1	2	1
Stress & anxiety	3	-	1	2	1	-	2	-
Difficulty with access to materials	3	1	1	1	1	-	-	2
Learning difficulties	2	1	1	-	-	-	1	1
Lack of understanding of social rules	1	-	1	-	-	-	1	-
Poor incidental learning	1	-	1	-	-	-	1	-
Poor seating	1	-	-	1	1	-	-	-
Poor memory	1	-	1	-	-	-	-	1
Journey to school	1	-	1	-	-	-	-	1

TABLE (xiv): RANK ORDER OF LIKERT SCALE SCORINGS FOR HOW EASY OR DIFFICULT CHILDREN WERE REPORTED TO FIND IDENTIFIED SKILLS

		Total scores	Mean	Mode
Social & Emotional	How easy/difficult to empathise with his/her peers when they are hurt, angry, upset etc.?	213	4.1	5
Social & Emotional	How easy/difficult to develop genuine friendships with peers in school setting?	213	4.1	5
Responses to routines & structure/ understanding of time & space	How easy/difficult to cope <u>without</u> regular routines & structure?	208	4.0	5
Social & Emotional	How easy/difficult to understand their own emotions & express their own emotional state?	205	3.9	5
Ownership of learning	How easy/difficult to organise him/herself?	197	3.8	4
Conceptual ability	How easy/difficult to transfer & generalise skills & knowledge & apply to different situations?	188	3.6	4
Conceptual ability	How easy/difficult to establish abstract concepts?	188	3.6	5
Ownership of learning	How easy/difficult to initiate activities?	184	3.5	4
Ownership of learning	Undertake self-help skills and functional life skills(appropriate for age)?	184	3.5	4
Responses to routines & structure/ understanding of time & space	How easy/difficult to cope with the unexpected & changes in their routine?	183	3.5	5
Responses to routines & structure/ understanding of time & space	How easy/difficult to predict what is to happen tomorrow or at the weekend?	181	3.5	5
Ownership of learning	How easy/difficult to learn and play independently?	189	3.4	3
Conceptual ability	How easy/difficult to correctly order parts of an activity or story sequence?	171	3.3	5
Conceptual ability	How easy/difficult to recall a sequence of events?	171	3.3	5
Conceptual ability	How easy/difficult to identify solutions to simple problems?	166	3.2	3
Responses to routines & structure/ understanding of time & space	How easy/difficult to recall past events?	166	3.2	5
Communication	How easy/difficult is it for the child to <u>MAINTAIN</u> communicative exchanges with a sensitive communication partner?	156	3	2,3,4
Communication	How easy/difficult is it for the child to <u>INITIATE</u> communicative exchanges with a sensitive communication partner?	131	2.5	2
Social & Emotional	How easy/difficult to develop trust and confidence with adults in school setting?	126	2.4	3
Responses to routines & structure/ understanding of time & space	How easy/difficult to anticipate what is to happen next when appropriate cues provided?	120	2.3	2
Conceptual ability	How easy/difficult to establish real, concrete concepts?	119	2.3	1

TABLE (xv): COMMUNICATION MODES IDENTIFIED AS BEING USED AS PART OF A TOTAL COMMUNICATION APPROACH

Communication mode	Total where mode reported to be used (N=52)	SLD/ PMLD (N=21)	Main-stream (N=16)	Other specialist (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Speech	41	19	11	11	11	11	8	11
Sign	40	19	7	14	11	12	8	10
Photographs	38	16	7	15	10	9	10	9
Symbols/ drawings	35	16	6	13	9	9	8	9
Printed word	34	11	12	11	8	10	6	10
Object cues/ objects of reference	26	15	5	6	6	7	7	6
Touch cues	13	9	3	1	5	4	2	2

TABLE (xvi): RANGE AND AVERAGE NUMBER OF COMMUNICATION MODES USED PER CHILD AS PART OF A TOTAL COMMUNICATION APPROACH

	SLD/ PMLD	Main- stream	Other specialist	EY/ KS1	KS2	KS3	KS4
Range of modes used per child	2-7	2-7	3-7	3-7	2-7	3-7	2-7
Average number of modes used	5.0	4.3	4.7	4.7	4.6	4.9	4.8

TABLE (xvii): THE DISTRIBUTION OF RESPONSES ACROSS TYPES OF PROVISION AND EDUCATIONAL PHASES WHERE USE OF CONCRETE TOOLS WAS CONSIDERED HELPFUL TO SUPPORT CONCEPT DEVELOPMENT

Type of provision	Total number where strategy reported helpful	EY/KSI	KS2	KS3	KS4
SLD/PMLD provision	20	3	7	6	4
Mainstream provision	11	4	3	1	3
Other specialist provision	15	4	3	4	4

TABLE (xviii): STRATEGIES REGARDED AS HELPFUL BY MORE THAN 50% RESPONDENTS, SET OUT IN RANK ORDER

Category:	Strategy:	Number where strategy reported helpful (N=52)
Communication	Use of a Total Communication approach	48 (92%)
Responses to routines & structure and understanding of time and space	Use of a daily routine and structure implemented consistently throughout the day	48 (92%)
Social and Emotional	Key people identified to develop secure relationships with child	47 (90%)
Sensory	Differentiating materials/activities in relation to child's sensory needs	46 (88%)
Sensory	Use of visual/auditory aids	46 (88%)
Conceptual ability	Use of concrete tools (e.g. real objects, photographs, symbols etc.)	46 (88%)
Conceptual ability	Provision of real experience provided within context	44 (85%)
Ownership of learning	Clear identification of what is motivational for the child	44 (85%)
Ownership of learning	Use of a flexible approach, responding to the child's changing needs and levels of responsiveness	44 (85%)
Communication	Ensuring access to a peer group with an appropriate level of communication skills	42 (81%)
Responses to routines & structure and understanding of time and space	Structured support for transitions	42 (81%)
Orientation & mobility and fine motor skills	Providing regular opportunities to engage in large movements	42 (81%)
Conceptual ability	Providing a balance between new and familiar activities	41 (79%)
Ownership of learning	Clear identification of the child's individual learning style informing the delivery of activities	41 (79%)
Ownership of learning	Ensuring activities have a clear beginning, middle and end	40 (77%)
Social and Emotional	Use of a structured programme/activities to support interactions with peers	39 (75%)

Category:	Strategy:	Number where strategy reported helpful (N=52)
Ownership of learning	Providing frequent choice-making opportunities	39 (75%)
Orientation & mobility and fine motor skills	Structured activities to support the development of gross motor skills and body awareness	39 (75%)
Sensory	Activities planned/implemented in relation to child's sensory integration difficulties	37 (71%)
Ownership of learning	Implementation of a structured programme to support self-help and self-organisational skills	37 (71%)
Sensory	Developing profiles of functional use of vision and hearing	35 (67%)
Communication	Providing a sensitive communication partner who is able to recognise and respond to the child's attempts to communicate	35 (67%)
Social and Emotional	Support to learn how to initiate/sustain social interaction sequences	33 (63%)
Responses to routines & structure and understanding of time and space	Use of mini routines	33 (63%)
Orientation & mobility and fine motor skills	Use of specialist/supportive seating	32 (62%)
Sensory	Appropriate behaviour management with recognition of impact of sensory integration difficulties	31 (60%)
Responses to routines & structure and understanding of time and space	Individualised pacing (may include a reduced timetable)	31 (60%)
Social and Emotional	Implementation of a structured programme to develop the child's understanding of emotions	31 (60%)
Orientation & mobility and fine motor skills	Implementation of a fine motor skills programme	31 (60%)
Conceptual ability	Use of identified strategies to support the development of problem-solving skills	30 (58%)
Responses to routines & structure and understanding of time and space	Use of concrete tools to support sequencing of an activity, transitions and daily routine	30 (58%)

Category:	Strategy:	Number where strategy reported helpful (N=52)
Ownership of learning	A structured response to manage the impact of fatigue	29 (56%)
Orientation & mobility and fine motor skills	Provision of alternative recording methods (e.g. scribe, use of IT, etc.)	29 (56%)
Conceptual ability	Use of structured fading and pre-learning/post-learning to support learning	28 (54%)
Orientation & mobility and fine motor skills	Use of large movement play activities	28 (54%)

Type of professional	Number of children where involved (N=52)	SLD/ PMLD (N=21)	Main-stream (N=16)	Other specialist (N=15)	EY/ KS1 (N=13)	KS2 (N=14)	KS3 (N=12)	KS4 (N=13)
Specialist multi-sensory impairment teacher	40 (77%)	17	11	12	10	10	10	10
Specialist visual impairment teacher	22 (42%)	9	5	8	6	6	5	5
Specialist hearing impairment teacher	38 (73%)	12	12	14	12	9	7	10
Intervenor	17 (33%)	8	2	7	5	5	4	3
Occupational therapist	38 (73%)	14	11	13	10	12	9	7
Physiotherapist	26 (50%)	11	8	7	8	5	6	7
Sensory integration specialist	15 (29%)	7	-	8	5	6	2	2
SaLT- communication	41 (79%)	18	11	12	10	12	9	10
SaLT- dysphagia (eating & drinking difficulties)	18 (35%)	10	3	5	5	4	6	3
Mobility rehabilitation officer	10 (19%)	4	3	3	3	2	1	4
ICT specialist	13 (25%)	4	4	5	1	4	2	6
Educational psychologist	22 (42%)	10	8	4	5	6	6	5
Learning support specialist	1		1			1		
Yoga instructor	1			1				1
Orthotics team	1	1			1			
School nurse	1	1			1			

APPENDIX 11: INTERVIEWS

TABLE (xix): WAYS IN WHICH INTERVIEWEES CONSIDERED STUDENTS WITH CHARGE TO BE DIFFERENT TO OTHER DEAFBLIND LEARNERS

- More extreme sensory needs
- Neurological differences in the brain, meaning they process information differently
- Elevated rate of anxiety and related repetitive/OCD behaviour resulting in a greater need for the environment and daily routine to be very regimented. One interviewee identified this as the biggest factor needing consideration when teaching a child with CHARGE
- Less flexible and unable to cope with miniscule changes in routines and the environment that other deafblind learners could tolerate, so that any change could “throw them”. One interviewee described this as being “unique to CHARGE”
- Greater reliance on use of routines and schedules with a higher level of structure and more schedule oriented
- A higher level of negotiation needed throughout the day
- A greater need to be in control, and very driven by their own topics
- Additional time for the development of self-organisational skills
- More socially aware and have an awareness of social connections. Very interested in families and babies
- Socially over-vigilant, accumulating a lot of social information that causes them to over-react in social family situations
- Social and conversational needs more paramount. Need additional teaching on social nuances
- Chronic health issues that need to be addressed
- That these children are deafblind as a consequence of CHARGE rather than any other condition was also highlighted

TABLE (xx): STRATEGIES IDENTIFIED AS HELPFUL BY INTERVIEWEES

Strategies:	% of questionnaire respondents reporting strategy to be helpful
Use of routine and structure, highlighted by 7 interviewees, with one explaining that this needed to be highly defined “so that the pupil does not get lost in the activity”	92%
Use of calendar and schedule systems noted by different interviewees who explained why it was important, including providing a concrete cue for the pupil to refer back to; containing additional descriptive information about an activity; helping reduce anxiety by letting them know what is to happen next; providing an element of control	58%
Providing a degree of flexibility with recognition of what must be done and what can be left	85%
The need for negotiation reported by 4 interviewees, with one describing it as being “the number one strategy” and one interviewee feeling this was a specific skill that teachers needed to learn	*
Sharing control was considered a helpful strategy by 3 interviewees who explained that pupils with CHARGE need to ‘hold the reins’ and giving controlled choices was empowering for the pupil	*
Being extremely clear and direct about expectations was felt to be very important by 4 interviewees. It was also explained the need to “take yourself out of the power struggle” and not to become personally invested	*
Giving clear information and providing a clear answer (e.g. “If asked what colour shirt you are going to wear, respond with ‘blue’ rather than I haven’t decided or wait and see”). Also, the need to avoid getting into in-depth dialogue (that could ‘feed’ perseverative behaviours), explaining “the further you get in the harder it is to get out!”	*
Checking basic concepts, and being as concrete as possible when developing new concepts	▲
Use of concrete aids	88%
Emphasising social interactions and encouraging humour, turntaking, and developing personal relationships. Making use of social scripts	63% and 75%
Offering motivating activities and spending time getting to know the student and what their interests are	85%
Building confidence through backward chaining, task analysis, and ending on a positive note	▲
Additional time to process information	▲
Ensuring interventions are sensory based	88%

TABLE (xxi): SUPPORT RECEIVED BY INTERVIEWEES IN RELATION TO STUDENTS WITH CHARGE

Type of input /support	Number of interviewees
Experienced colleagues in school who support. Undertaking observations and discussions	9
Annual training from experts in the field	7
The programme- Perkins has so many students with CHARGE that it is a rich resource to learn from, and the chance to work with different pupils with CHARGE over time	6
Internet/ websites/ webcasts	4
Financial support to attend external training/conferences	3
The student's family	2
Links with a local college and the opportunity to engage in research	2
Direct input and contact from external professionals within the field	1

APPENDIX 12

TABLE (xxii): RATE OF REPORTING OF BEHAVIOURS INDICATIVE OF VESTIBULAR AND PROPRIOCEPTIVE DIFFICULTIES

Behaviour	% where reported
Vestibular (balance) difficulties	48 (93%)
Poor body awareness/ coordination difficulties	43 (83%)
Does the child need to adopt a horizontal position at any time during the school day?	28 (54%)
Does the child seek additional support for their position when standing or sitting?	34 (65%)

TABLE (xxiii): PRESENCE OF POTENTIAL INDICATORS OF SENSORY INTEGRATION DYSFUNCTION

Behaviour	% where reported
Difficulty using senses together in a coordinated way	36 (69%)
Difficulty with distractibility/ remaining on task	39 (75%)
Difficulty with sensory overload/ under-stimulation	39 (75%)

TABLE (xxiv): CHARACTERISTICS THAT MAY BE PRESENT IN A CHILD OR YOUNG PERSON WITH CHARGE AND INFLUENCE LEARNING AND DEVELOPMENT, WITH RELATED EXAMPLES FOUND IN LITERATURE ON DEAFBLIND EDUCATION

Characteristics	Related examples found in literature on deafblind education
A combination of sensory impairments- potentially all sensory modalities may be affected	
Sensory integration difficulties that include: <ul style="list-style-type: none"> ○ Difficulty using their senses in a coordinated way ○ Difficulty with distractibility/remaining on task 	Every individual with deafblindness would benefit from a sensory integration assessment: Brown, 2009 There may be a difficulty with how the senses are integrated: Clark, 2000
Ongoing health issues and past medical experiences	Many will need ongoing access to arrange of healthcare services: Aitken, 2000
High levels of fatigue	The impact of fatigue on a child’s readiness to engage and learn: Pease, 2000; Murdoch et al, 2009
High levels of stress and anxiety	Included in the definition of MSI in the PLASC, Department for Education and Skills, 2003; Pease, 2000
A preference for using different communication modes for both receptive and expressive communication that may include a greater preference: <ul style="list-style-type: none"> ○ For using speech or sign/gesture to express themselves ○ For concrete cues to support receptive communication 	Difficulty in communicating with others: Aitken, 2000 A need for the use of a combination of communication modes: Pease, 2000; Murdoch et al, 2009
Easier and more successful in forming relationships with adults than peers	Social interactions will vary from child to child; some will only interact with adults who know them well: Rowland, 2009 May have a preference in relating to adults rather than peers: Eyre, 2000
Difficulty developing same-age relationships	Support to form and nurture peer relationships: Mar and Sall, 1995
Difficulty understanding and expressing their own emotional state, and empathising with their peers	Will need support to learn to recognise and understand their emotions and those of others since they miss verbal and auditory cues that may convey this information: Hodges, 2000

Characteristics	Related examples found in literature on deafblind education
Additional time to process information	Pease, 2000; Nelson et al, 2010; Murdoch et al, 2009
The ability to develop real, concrete concepts may be a strength	Concept formation is an issue for all deafblind learners: Miles and McLetchie, 2008
Establishing abstract concepts can be much more difficult	
Problem-solving may also be a relative strength if this is regarded as including the development of adaptive behaviours in response to difficulties or problems encountered	Use of direct experience to develop problem-solving skills: Rowland and Schweigert (1997)
Needing a high level of routine and structure and experiencing great difficulty in coping when these are not established	A need for clear routine and structure to support understanding: Pease, 2000; Nelson et al, 2009; Murdoch et al, 2009
<p>Executive function difficulties that may be manifested as:</p> <ul style="list-style-type: none"> ○ Lack of flexibility and difficulty coping with unexpected changes ○ Difficulty initiating activities and engaging in independent play and learning ○ Difficulty developing and implementing self-organisational skills ○ Difficulty coping with transitions and shifting attention from one activity to another ○ Poor memory ○ Difficulty transferring and generalising skills 	<p>Too easy for a child to “lapse into passivity” (p.188) when not receiving direct adult support: Hodges, 2000</p> <p>Importance of teaching self-help skills: Clark, 2000</p> <p>Physical transitions between activities may need support: Murdoch et al, 2009</p> <p>Issues applying skills and knowledge to different situations: Aitken, 2000; Murdoch et al, 2009</p>
Benefitting from a use of concrete cues to assist anticipation of what is to happen next.	Use of concrete cues to support sequencing an activity, transitions, and daily routine: Hodges, 2000; Murdoch et al, 2009; Nelson et al, 2009

Characteristics	Related examples found in literature on deafblind education
Easier to recall past events than to predict events in the near future.	Memory is supported through frequent repetition: Pease, 2000
Easily affected by environmental distractions.	Need to create a distraction- free environment: Murdoch et al, 2009
A need to be in control and a requirement for a level of negotiation.	
Engaging in behaviours to address needs arising from poorly developed or under-stimulated vestibular and proprioceptive systems that may include: <ul style="list-style-type: none"> ○ A need to adopt a horizontal position ○ Seeking additional support for position when standing/sitting. 	Use of proprioception mentioned: Eyre, 2000
Poor body awareness and difficulty organising and coordinating their body.	Difficulty “moving around the environment” (p.3) as a consequence of their visual and hearing impairments: Aitken, 2000 A need to focus on body awareness: Clark, 2000; Murdoch et al, 2009
Experiencing fine motor difficulties with poor pencil/handwriting skills.	

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