CHALLENGING BEHAVIOUR, PARENTAL EMOTIONAL DISTRESS, CHILD QUALITY OF LIFE AND SERVICE PROVISION IN CHILDREN WITH INTELLECTUAL DISABILITIES

by

EFTHALIA KARAKATSANI

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School of Psychology
College of Life and Environmental Sciences
University of Birmingham
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ABSTRACT

There are two areas of research into challenging behaviour that are as yet underexplored but important and likely to promote beneficial outcomes. First, there are very few studies of the persistence of challenging behaviour and of predictors of persistence. In this thesis this question is addressed by studying challenging behaviour in people having the same cause for their intellectual disability, Fragile X syndrome (FXS). Second, the interrelationships between the quality of life of children with intellectual disabilities who show challenging behaviour, parental wellbeing and service use has not yet been explored. The relationships between these factors are explored in a large-scale survey.

Challenging behaviour was found to be highly persistent in FXS and predicted by the presence of autism spectrum disorder. For the second question posed, impulsivity, a behavioural correlate of challenging behaviour, but not challenging behaviour alone, was predictive of parental emotional wellbeing. Behavioural correlates of challenging behaviour and parental emotional wellbeing were found to be related to the quality of life of children with intellectual disabilities. Finally, parental anxiety and the child's age were strongly associated with access to mental health and social services.

The results have important implications for the targeting of individualised early intervention strategies in relation to children at high risk, and which can effectively support children with intellectual disabilities and challenging behaviour in order to enable families to experience a

DEDICATION

For my cousin, John, for inspiring me to further my knowledge of the difficulties faced by people with intellectual disabilities and their families.

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TABLE OF CONTENTS

LIST OF TABLES

LIST OF FIGURES

CHAPTER 1

Challenging behaviour in children with intellectual disability

1.1. Preface

The present chapter provides an overview of the concepts, models and research that are immediately relevant to the empirical work described in this thesis. The chapter also includes an introduction to challenging behaviour in children with intellectual disabilities. Empirical research delineating the epidemiology, associated personal characteristics and causes of challenging behaviour in individuals with intellectual disabilities is then summarised. An evaluation of the previous research relevant to the present thesis, key findings and areas for further investigation are highlighted, providing a rationale for the ensuing empirical work reported in this thesis.

1.2. Introduction

People with intellectual disabilities (ID) comprise about 2% of the U.K population. A prevalence study conducted by Emerson (Emerson, 2010) found that 1,198,000 people in the U.K had ID. This population included 298,000 children (188,000 boys and 110,000 girls aged 0-17 years old) and 900,000 adults aged 18 and over (526,000 men and 374,000 women). People with ID face numerous difficulties throughout the lifespan. ID has been found to be associated with physical/health problems (van Schrojenstein Lantman-de & Walsh, 2008), with psychological and mental health problems (Cooper et al., 2007) and behavioural problems (Dekker et al., 2002; Eisenhower, Baker & Blacher, 2005). The following sections expand on these different problem areas in relation to individuals with ID.

1.2.1. Health problems in individuals with ID

It has been previously documented that people with ID can present health problems 1.7 times more frequently than individuals without ID (Straetmans, van Schrojenstein Lantman-de, Schellevis & Dinant, 2007). These problems are principally epilepsy, dermatological problems and mental health problems. It has also been reported that people with ID receive prescriptions more frequently, particularly psychiatric medication and anticonvulsants, compared to people without ID (Straetmans, van Schrojenstein Lantman-de, Schellevis & Dinant, 2007).

Furthermore, there is evidence to show that children with intellectual disability in the United Kingdom are likely to experience health inequalities in a number of areas, especially in relation to health conditions that may increase the risk of serious ill health in later life (e.g. obesity). More specifically, carers of children with ID reported that their children had poor versus good general health status 2.5 to 4.5 times more frequently, compared to carers of children without ID. In line with this finding, children with intellectual disability have been reported to be more likely to be obese than their non-intellectually disabled peers (Ells, 2006; Emerson, 2009; Emerson et al., 2011a). In addition, data from the Millennium Cohort Study also showed that children with intellectual disability were 3.7 times more likely never to exercise or participate in sports (Emerson et al., 2011a). Taken overall, these findings have also shown that ID may reduce the quality of life, and act as a barrier to social inclusion (Allerton et al., 2011).

Moreover, people with ID may also experience additional co-morbid medical conditions, which may affect their overall health status. Among the most frequently reported health problems in individuals with ID is epilepsy. The prevalence of epilepsy in the UK population is between 0.5 and 1 percent; among those with moderate intellectual disability this prevalence rises to 15 percent; and among those with severe and profound disability the rate increases to 30 percent (Chapman et al., 2010).

In addition to medical issues, individuals with ID have also been noted to have a high prevalence of mental health problems, which may also contribute to limiting their overall daily functioning. The following section goes on to discuss the importance of understanding the occurrence of mental health problems in individuals with ID.

1.2.2. Mental health problems in individuals with ID

There is evidence to show that in a cohort of adults with ID more than a third (40.9%) had mental health problems (Cooper, Smiley, Morrison, Williamson & Allan, 2007). In the same study, it was found that the occurrence of mental health problems was associated with a range of individual characteristics, such as gender, the degree of intellectual disability, the number of traumatic life events, the extent of service use, and the presence of other health problems among adults with ID. Interestingly, in a systematic review examining the relationship between mental health problems and intellectual disability, it was found that the proportions of children with psychopathology who had mild, moderate, profound and severe ID were 21%, 40%, 49% and 42%, respectively (Molteno, Molteno, Finchilescu & Dawes, 2001). Similar findings were derived from an empirical study which reported that children with ID were seven times more likely to have a diagnosis of a psychiatric disorder than children of the same age without ID (Emerson, 2003). It was shown that conduct disorders, anxiety, attention deficit and hyperactivity, as well as pervasive developmental disorders, are higher among children with ID than among children without ID. Conversely, the same study did not find significant differences between children with and without ID in regard to depressive and eating disorders, as well as psychosis (Emerson, 2003). To this end, further research is needed in relation to mental health problems in individuals with ID, to delineate the putative associations between particular types of psychiatric disorder and ID.

In addition to issues of health and mental health there is a theme from research exploring the close relationship between the occurrence of behavioural problems and intellectual disability. The following section discusses this area.

1.2.3. Behavioural problems in individuals with ID

Emerging literature has suggested the existence of a strong association between behavioural and sleep problems in relation to intellectual disability. It has been found that children with ID are more likely to experience abnormal sleep patterns, which may also have an effect on their daytime behaviour (Wiggs & Stores, 1996). Among the most critical behavioural problems that have been associated with sleep problems in children with ID are stereotypical behaviours, impulsivity, self-injurious behaviour, aggression, screaming, temper tantrums and non-compliance (Wiggs & Stores, 1996). Interestingly, the degree of intellectual disability has been shown to act as a moderating variable for the degree of the association between sleeping problems and the above behavioural manifestations.

Furthermore, findings in the field suggest the existence of strong associations between specific types of behavioural problems and ID. Self-injury has been shown to be more common amongst children with severe ID. Moreover, self-injury has also been found to be strongly associated with autism diagnoses and communication deficits in children with ID (McClintock, Hall & Oliver, 2003). Similarly, aggression has been shown to be more prevalent amongst individuals with autism, and with communication deficits, as well as varying by gender, with males found to be more likely to develop aggression compared to females. In addition, stereotypical behaviours have been found to be more common amongst individuals with severe ID (McClintock, Hall & Oliver, 2003). This finding is consistent with previous evidence to suggest an association between self-injury, aggression and stereotypical behaviours, and the degree of intellectual disability (Wiggs & Stores, 1996; Emerson, 2001; Rojahn & Esbensen, 2002).

In addition to the associations between behavioural problems and ID, there is a separate line of research showing an effect of children's behavioural problems on parental emotional distress, when other variables such as health problems and intellectual disability have been controlled for (Hastings, 2002). To this end, when investigating the role of behavioural problems in intellectual disability, the broader effects of such problems need to be taken into account, as these may have an effect upon another individual’s daily life (i.e. a parent’s), as well as on family functioning. This area of inquiry is a major concern of the present thesis and is discussed in detail in the following sections.

Understanding that the occurrence of health problems, mental health issues and behaviour problems among individuals with ID are interrelated may further help delineate the overall quality of life of individuals with ID, as well as help in assessing the relevant needs. To this end, the following section explores the impact of challenging behaviour on both the individual’s and their family’s daily functioning. In addition, the prevalence, persistence and risk markers of challenging behaviour in individuals with ID are also reviewed.

1.3. The definition of 'challenging behaviour'

It has been argued that the term 'challenging behaviour' has emerged from a desire to shift the understanding of behaviours such as aggression, self-injury and property destruction away from a medical model, and towards a conceptualization which acknowledges the roles of others as well as the wider environment (Oliver, in press). Challenging behaviour is defined as:

> "A*ny culturally abnormal behaviour of such intensity, frequency or duration that the physical safety of the person or others is likely to be placed in serious jeopardy, or behaviour which is likely to seriously limit use of, or result in the person being denied access to, ordinary community facilities*" (Emerson, 1995, p.24).

The term 'challenging behaviour' should be viewed as a social construct (Emerson, 1995; Morris & Midgley, 1990). Social rules, the ability of the person to provide a plausible explanation for their behaviour, others' beliefs about the nature of intellectual disability, the causes of the person's challenging behaviour, and the capacity of the service provided to manage an individual's behaviour are all aspects of behavioural manifestation that can be classified as challenging (Emerson, 1995).

The social construct of challenging behaviour encompasses a variety of different behaviours, including aggressive behaviour, anxiety, antisocial behaviour, hyperactivity, social withdrawal, and stereotyped behaviour, which often includes self-injury. There is a range of behavioural measures available in the literature, which have been developed to measure aspects of challenging behaviour. More specifically, the Aberrant Behavior Checklist (ABC: Aman, Singh, Stewart & Field, 1985), the Child Behavior Checklist (CBCL: Achenbach & Edelbrock, 1983) and the Developmental Behavior Checklist (DBC: Einfeld & Tonge, 1995) are some of the tools most widely used to assess challenging behaviour. These measures have been applied in a range of clinical and research settings. However, there is a critical need when measuring challenging behaviour to delineate the particular constructs of such behaviours. More specifically, stereotypical behaviours have been found previously to account for behavioural correlates of challenging behaviour, as opposed to challenging behaviour itself (Sloneem, Arron, Hall &Oliver, 2009). Moreover, individual and demographic characteristics of individuals with ID, such as the degree of intellectual disability, the individual's demographic variables, and pain, may have specific associations with these behavioural correlates. (Sloneem, Arron, Hall & Oliver, 2009). For instance, the most widely used measures of challenging behaviour are using total scores that may mask associations between such variables with their individual forms of behaviour, that may result in problems in identifying the underlying mechanisms of such behaviours. To date, although the available behavioural measures assess challenging behaviour as a single dimension of behaviour, there is an increasing consensus (National Institute for Health and Care Excellence, 2015) which proposes that challenging behaviour encompasses a variety of different behaviours including, but not limited to, aggressive behaviour, anxiety, antisocial behaviour, hyperactivity, social withdrawal, and stereotyped behaviour (sometimes including self-injury). Therefore there is clear merit in using more precise definitions, but also more targeted measures, in relation to the behaviours of interest.

A particular concern of the present thesis relates to the investigation of both self-injurious and aggressive behaviour as the most critical forms of challenging behaviour among individuals with ID. Self-injurious behaviour (SIB) is characterised by chronicity and may have a major impact on an individual's life. There is evidence to show that SIB has resulted in physical trauma, medical complications, including permanent tissue damage, bone fractures, dismemberments, and in the most severe cases, death (Yang, 2003). Moreover, SIB is frequently a life-long behaviour that persists from childhood through adulthood (Schroeder, 1996). It is also common for SIB to be associated with other behavioural problems, such as physical aggression, stereotypies, sexually inappropriate behaviours, tantrums and property destruction (Matson, Cooper, Malone & Moskow, 2008).

However, the impact of self-injurious behaviour is not limited to the individual's life but may also affect others' functioning in the individual's close environment. Most notably, self-injurious behaviour has been shown to have negative psychological effects and to be associated with stress in care staff, teachers, and professionals, as well as in other family members (Mossman, Hastings, & Brown, 2002). SIB may have a critical implication for both the individual with intellectual disability and their family's functioning. To this end, it is vital to identify the risk markers that may be associated with the manifestation of SIB, and that may inform daily practice and service provision. In the following section there is a discussion of the nature and causes of SIB, as well as the risk markers and persistence of such behaviours in individuals with ID.

1.4. Self-injurious behaviour and aggression in children with ID

The particular concern of the present work relates to the investigation of aggressive and self-injurious behaviour in individuals with ID. Self-injurious behaviour has been defined as:

> *"Any non-accidental behaviour, initiated by the individual, which directly results in physical harm to that individual. Physical harm (includes) bruising, lacerations, bleeding, bone fractures and breakages, and other tissue damage"* (Murphy & Wilson, 1985, p. 15).

Moreover, aggression has been defined as:

> *"Any behaviour directed toward another individual that is carried out with the immediate intent to cause harm"* (Bushman, 2001).

As indicated earlier in this chapter, there is research that has highlighted the existence of strong associations between SIB and ID. It is imperative to understand the exact nature of these associations, that may involve the prevalence of self-injury and aggression, and the personal characteristics which are associated with the presence of self-injurious and aggressive behaviour in individuals with ID. The following section explores these areas.

1.5. The prevalence of challenging behaviour

1.5.1. Prevalence of self-injurious behaviour, aggression and property destruction among individuals with ID

There is a line of research investigating the prevalence of challenging behaviour in individuals with ID. Most notably, it was found that 10-15% of people with intellectual disability exhibited challenging behaviour, while 5-10% of the same population exhibited more severe forms of challenging behaviour (Emerson et al., 2001). In the same investigation, it was found that the most common forms of challenging behaviour were: aggression, 7%, destructive behaviour, 4-5%, self-injurious behaviour, 4%, and other behaviour, 9-12%. The term “other” behaviours can be justified by the fact most people identified as showing challenging behaviour showed two or more of these main forms of challenging behaviour (Murphy et al., 1993). The co-occurrence of multiple forms of challenging behaviour has also been reported within such broad categories. Thus, for example, Oliver et al., (1987) reported that 54% of the people identified with self-injurious behaviour engaged in more than one self-injurious topography. Indeed, 3% (20 of the 596) engaged in five or more different forms of self-injury. Interestingly, the vast majority of the population studied was found to show two or more of the above forms of challenging behaviour.

A total population study (Emerson et al., 2001) was initially conducted in 1988 and a follow up study was conducted in 1995. Seven District Health Authorities in the UK participated in a total population survey of the extent and nature of challenging behaviours shown by people with ID. Two of these seven initial areas agreed to take part in the follow up study in 1995. Proxy data from 264 people with ID were collected and analysed regarding the prevalence of challenging behaviour. Challenging behaviour was investigated through the administration of the Behaviour Problems Inventory (Rojahn, Polster, Mulick & Wisniewski, 1989). Informants were asked whether individuals displayed any of the following behaviours: aggressive, self-injurious, destructive and ‘other’ difficult, disruptive or socially unacceptable behaviour. Informants were instructed to complete these sections if the person showed that form of challenging behaviour to the extent that it was considered by them to constitute a serious management problem or would do were it not for specific controlling measures undertaken in the person's current setting. Each section of the inventory collected information on the specific form of the challenging behaviour, its frequency, circumstances and the level of intervention required to deal with incidents; Although longitudinal investigation would be needed to address the prevalence and the persistence of challenging behaviour, there were some other limitations regarding this study. First of all, all the data collected came from key informants and as such, were limited by their knowledge of the person's abilities, situation and behaviour. In addition, the sample size fell in the follow up stage, which affects any regression analysis conducted to describe potential predictors of challenging behaviour. Consequently, the results are perhaps more useful for exemplifying broad themes than for detailed analyses of predictors of specific behaviours.

In the first comprehensive total population study of self-injury in individuals with an intellectual disability, it was found that 12% of people with intellectual disability engaged in self-injurious behaviour (Oliver, Murphy & Corbett, 1987). More recent prevalence studies have broadly supported this pattern of findings, with the prevalence rates of SIB found to vary between 4% (Cohen et al., 2010; Cooper et al., 2009; Holden & Gitlesen, 2006) and 17% (Collacott, Cooper, Branford & McGrother, 1998). Lower prevalence figures are typically associated with more stringent definitions of self-injury e.g. actual physical harm, rather than potential physical harm (Cooper et al., 2009). Likewise, less conservative prevalence figures include less severe and less frequent occurrences of self-injury within the prevalence data (e.g., Collacott et al., 1998). Generally, prevalence estimates of self-injury for individuals with intellectual disability are estimated to lie between 4 and 10%. These studies described the prevalence of self-injurious behaviour among individuals with ID. However, some methodological limitations have been observed in these studies. First, some of them (Cohen et al., 2010; Cooper et al., 2009) did not conduct an independent assessment of ASD and relied upon previous clinical diagnoses for their inclusion criteria. As ASD is diagnosed solely through behavioural markers, it is essential that studies reporting prevalence data ensure that all individuals within the study meet diagnostic criteria for ASD. Failure to do this threatens the external validity of the study, and renders the reported prevalence rates unusable for generalisation. A further limitation was the absence of follow up studies (Cohen et al., 2010) where authors were not able to know through a longitudinal study the extent to which any age-related changes would actually take place. These changes are relevant to a threat to internal validity because of maturation factors, namely the processes within subjects which act as a function in relation to the passage of time.

In addition to the prevalence of SIB in ID, there is also evidence to show the prevalence rates of aggression in ID. Most notably, it has been reported that 51.3% of individuals with profound or severe ID displayed aggressive behaviour (Crocker et al., 2006). However, the term 'aggression', as conceptualized in these observations, may classify behavioural manifestations, such as property destruction and self-injurious behaviour, as topographies of aggressive behaviour, rather than as separate forms of challenging behaviour. Additional observations from that study included the finding that physical aggression had a prevalence of 24.3% among individuals with ID, while property destruction was found to have a rate of 25.7% (Crocker et al., 2006). Limitations were that the data were collected from educators, who were limited by their knowledge and recollection of the person’s behaviour. Furthermore, no inter-rater reliability tests on ratings of aggressive behaviour were carried out. To minimize the resulting threat the authors administered a standardised instrument that has a history of good inter-rater reliability and with ratings that have clear behavioural anchors.The prevalence rates were related in a linear way with the ID level, where individuals with mild and moderate ID were more likely to show verbal aggression, compared to those with severe or profound intellectual disabilities. Conversely, individuals with profound and severe ID were more likely to show physical aggression and property destruction compared to those with mild or moderate ID.

Additional studies have also identified an association between the presence of aggressive behaviour and particular genetic syndromes. It was found that while the percentage of aggressive behaviour associated with individuals with ID was 46.3%, with individuals with Smith Magenis syndrome, Angelman and Cri du Chat syndrome in the same age group (mean age 16.46 years old) it was reported to be 70% (Arron et al*.,* 2011). In addition, prevalence rates for aggressive behaviour varied between 64.8% and 40.2% for individuals with Lowe, fragile X, Prader-Willi and Cornelia de Lange syndromes. 797 carers of individuals with Angelman (AS), Cri du Chat (CDCS), Prader Willi (PWS), Cornelia de Lange (CdLS), Lowe (LS), fragile X (FXS) completed questionnaires regarding individuals' behavioural manifestations. The Challenging Behaviour Questionnaire (CBQ; Hyman, Oliver & Hall, 2002) was used to investigate the presence or absence of self-injury, physical aggression, verbal aggression, property destruction and inappropriate vocalisations over the previous month. In this study, therefore, items regarding self-injury and physical aggression were used. Although in the study of Arron et al., (2011) direct observations were not used, their use of standardised measures strengthens the validity of the findings.

In summary, these prevalence indicators suggest that individuals with genetic disorders are more likely to show aggressive behaviour compared with individuals with ID. In this study, impulsivity and overactivity were established as correlates of self-injury and aggression in these syndrome groups. Given the variety of personal characteristics across and within these genetic syndromes an exclusive operant theory cannot explain these differences in the prevalence of aggression. However, there is evidence that operant theory is applicable to these behaviours seen in people with these syndromes (Hall, Oliver and Murphy, 2001; Arron et al., 2006; Taylor and Oliver, 2008, Strachan et al., 2009). It is clear therefore that more complete causal models of self injury and aggression must encompass both personal characteristics (including those of genetic origin) and environmental factors (Oliver, 1993; 1995).

In addition to the evidence associating particular genetic syndromes with the presence of self-injurious behaviour, there are also data illustrating an association between genetic syndromes and particular topographies of self-injurious behaviour. More specifically, the most common forms of self-injury have been found to be head banging, hitting other parts of the body, hitting the head against objects, and scratching (Lowe et al*.,* 2007; Symons et al*.,* 2004). In individuals with Lesch-Nyhan syndrome in particular, it was found that the most common sites of self-injury are the fingers, lips and tongue (Symons et al*.,* 2003).

Additionally, the most common topographies of self-injurious behaviour in individuals with Smith Magenis syndrome are self-hitting or self-biting (Arron et al*.,* 2011), while some individuals have been reported to remove their fingernails or insert objects into bodily orifices (Symons et al., 2003). Furthermore, individuals with Prader-Willi syndrome are more likely to pick their skin over wide areas of their body (Symons et al., 2003), whereas individuals with Cri du Chat syndrome or Cornelia de Lange syndrome are more likely to pull hair, scratch or rub skin. Finally, it was found that biting is a common self-injurious behaviour form in individuals with FXS (Symons et al*.,* 2003; Arron et al*.,* 2011). Taking these together, it is evident that a variation on the topography of SIB is observed in relation to each syndrome. This variation may be dependent upon syndrome-specific biological factors or environmental influences that may contribute to such behavioural manifestations (Symons et al., 2003).

There is also evidence to show a difference between the prevalence rates of aggressive, self-injurious and property destruction behaviour between children and adults with ID (Lowe et al., 2007). Children with ID were found to have a prevalence rate of 65% for serious or controlled aggressive behaviour, 42% for property destruction, as well as 35% for self- injurious behaviour. The prevalence rates for adults were found to be 51%, 29% and 35% for aggression, property destruction and self-injury respectively. However, other studies have not provided evidence for an association between age and self-injurious behaviour (Lowe et al., 2007). There is evidence to suggest that the prevalence of different types of challenging behaviour varies amongst individuals with ID. There are specific individual characteristics that have been shown to be able to predict the presence of such behavioural manifestations in individuals with ID. These characteristics, that range from environmental to behavioural variables, have been previously described as risk markers for challenging behaviour. It is critical at this point to review all these characteristics and their importance in delineating the nature of challenging behaviour amongst individuals with ID.

1.5.2. Personal characteristics associated with challenging behaviour

Research has identified a range of characteristics that may be associated with the presence of challenging behaviour in individuals with ID. The available literature has distinguished between individual and behavioural characteristics that have been shown to be primarily associated with the manifestation of a range of types of challenging behaviour. In the following section the associations between challenging behaviour and these two distinct types of risk markers for challenging behaviour are discussed.

1.5.3. Personal characteristics specifically associated with challenging behaviour

There are several individual characteristics that have been described in the literature as potential risk markers for challenging behaviour (McClintock et al*.,* 2003; Murphy et al., 1999). Demographic markers include a diagnosis of Autism Spectrum Disorder (ASD) or Attention Deficit Hyperactivity Disorder (ADHD), or a genetic condition, as well as the degree of intellectual disability, which have been shown to be involved in the presence of challenging behaviour. The following sections expand on these risk markers.

1.5.4. Demographic risk markers

Demographic characteristics, such as age and gender, have been found to be associated with the presence of challenging behaviour in individuals with ID (Davies & Oliver, 2013; McClintock et al., 2003). More specifically, in terms of age, there is evidence to show that the prevalence of aggressive behaviour might decrease in adulthood (Deb et al*.,* 2001; Tyrer et al*.,* 2006; Davies & Oliver, 2013). Emerging evidence has shown that while the prevalence of self-injurious behaviour increases to mid-adulthood (e.g. 40 years old), after the age of 50 it decreases significantly (Davies & Oliver, 2013). The same study also reported similar trajectories across the life span for aggressive behaviour.

Furthermore, there is evidence to show gender associations with the presence of aggressive behaviour. Males were found to be more likely to show aggressive behaviour compared to females (McClintock et al., 2003). However, another study did not report significant associations between gender and challenging behaviour in general (Holden & Gitlesen, 2006). However, in this study the sample size has been observed as a threat to its external validity. Most notably, a population validity threat has been observed through considering the small sample sizes in several syndrome groups except Down's syndrome. Considering the various types of behaviours which are incorporated in the term ‘challenging behaviour’, it is critical for future research to investigate the extant association between gender and the presence of different types of challenging behaviour in individuals with ID**.**

1.5.5. ASD and ADHD as risk markers for challenging behaviour

There is research reporting high prevalence rates of types of challenging behaviour in populations with a diagnosis of ASD or ADHD. More specifically, high prevalence rates of aggressive (Cooper et al., 2009) and self-injurious behaviour, have been reported in individuals with ASD, with at least 50% of them reported to show self-injury (Baghdadli et al., 2003; Richards et al., 2012; Medeiros et al., 2012); and over two thirds to manifest aggressive or destructive behaviour (Medeiros et al., 2012). A meta-analysis study found that individuals diagnosed with autism were more likely to manifest self-injurious behaviour, aggression and property destruction compared to individuals without an autism diagnosis (McClintock, Hall & Oliver, 2003). Evidence illustrating associations between ASD and challenging behaviour in general also exists, showing that in a study population of individuals with ASD, 35% displayed challenging behaviour (Holden & Gitlesen, 2006). In addition, with the evidence showing an association between ASD and challenging behaviour, there is also evidence to illustrate an association between aggression and the presence of ADHD (Cooper et al., 2009).

Overall, research has showed that the prevalence rates of different types of challenging behaviour are higher among individuals with ASD and ADHD, suggesting an association between these strands. However, it may be argued that the above types of behaviour may be part of disorder-specific symptomatology and not separate manifestations associated with the presence of ASD or

ADHD disorder respectively. Further research would be essential to delineate the nature of such associations and their role in the broader behavioural profile of individuals with ID.

1.5.6. Level of intellectual disability as a risk marker for challenging behaviour

A separate line of research has focused on examining the associations between the degree of intellectual disability and the manifestation of challenging behaviour (Emerson et al., 2001). More specifically, it has been estimated that the prevalence rates of people with ID displaying challenging behaviour vary from 30% to 60% (Lowe et al., 2007). In addition, self-injurious behaviour has been found to be related to lower levels of adaptive skills, such as in the communication, socialization and the daily living skills domains (Baghdadli, Pascal, Grisi & Aussilloux, 2003).

However, the absence of a standardised measure of self-injurious behaviour limits the identified association between self-injurious behaviour and lower adaptive skills because there is no evident consistency as to which behaviours proxy participants identified as self-injurious behaviour incidents. This is therefore a threat to the internal validity of using the selected instruments.

A significant proportion of individuals who display self-injurious behaviour may also have co-morbid conditions. Most notably, it has been found that there is a high prevalence of autistic spectrum phenomenology amongst individuals with Cornelia De Lange and FXS (Oliver et al., 2011). Moreover, self-injury appears to be more common amongst individuals with a severe or profound degree of intellectual disability, a diagnosis of autism and deficits in receptive and/or expressive communication (McClintock, Hall & Oliver, 2003).

1.5.7. Genetic conditions as risk markers for challenging behaviour

In addition to the level of ID, there is evidence to show that individuals with genetic syndromes associated with ID are more likely to develop challenging behaviour, compared to individuals with ID with heterogeneous aetiology. More specifically, prevalence studies have reported that the rates of challenging behaviour among individuals with ID vary between 10% and 15% (Emerson et al., 2001), when the corresponding rate for challenging behaviour among individuals with genetic syndromes was found to be significantly higher (Arron et al*.,* 2011). In fact, it was reported that 93% of individuals diagnosed with Smith–Magenis syndrome displayed self-injurious behaviour, whereas the prevalence rates of self-injurious behaviour were found to vary between 65% and 76% in individuals with Lowe, Cornelia de Lange and Cri du Chat syndromes. Similarly, the 50% of individuals with FXS and Prader–Willi syndrome showed self-injurious behaviour (Arron et al*.,* 2011). Also, it was found that individuals with Smith–Magenis syndrome, as a group, were at least 6.32 times more likely to manifest self-injury, compared to individuals with Cornelia de Lange, Cri du Chat, fragile X, Lowe and Prader–Willi syndromes. Similar findings about the high prevalence of self-injurious behaviour in individuals with particular genetic disorders were reported in Oliver et al., (2009), where it was found that 55.6% of the individuals with Cornelia de Lange showed self-injurious behaviour.

Furthermore, studies have shown that aggressive behaviour is more prevalent amongst individuals with genetic syndromes than in individuals with ID heterogeneous aetiology. It was found that 88% of individuals with Cri du Chat syndrome exhibited aggressive behaviour (Collins & Cornish, 2002). In addition, there is evidence that 80% of individuals with Smith Magenis syndrome show aggressive behaviour (Sloneem et al., 2011) whilst the corresponding percentage for aggressive behaviour amongst individuals with ID heterogeneous aetiology is 46.3 % (Arron et al., 2011).

Overall, the evidence presented in this section highlights the contribution of individual characteristics to the manifestation of different types of challenging behaviour. Taking these characteristics into account is critical to a better understanding of the individual differences in challenging behaviour associated with ID. In the following section, the behavioural risk markers associated with the presence of challenging behaviour are explored.

1.5.8. Behavioural risk markers

It has been suggested above that there are behavioural risk markers associated with the presence of challenging behaviour among individuals with ID. More specifically, restricted, repetitive and stereotyped behaviours, as well as over-activity and impulsivity, are behavioural correlates that have been previously defined as potential behavioural contributors to the manifestation of different types of challenging behaviour. The following sections explore the evidence on these behaviours.

1.5.9. Challenging behaviour and restricted, repetitive and stereotyped behaviours

Evidence is accumulating in support of an association between challenging behaviour and the presence of restricted, repetitive and stereotyped behaviours. It was found that insistence on sameness was significantly associated with the presence of challenging behaviour, and more specifically self-injurious behaviour, in children with ASD (Duerden et al., 2012). In addition, repetitive behaviours were also found to be associated with the presence of challenging behaviour (Oliver et al., 2012). Furthermore, there is evidence to highlight associations between restricted behaviour in individuals with ASD and self-injurious behaviour (Richards et al., 2012). Moreover, other research has highlighted associations between self-injury and stereotyped, compulsive behaviours and hyperactivity in individuals with Cornelia de Lange syndrome (Sloneem, Arron, Hall & Oliver, 2009; Rojahn, Wilkins, Matson & Boisjoli, 2010).

In addition to the evidence that reveals associations between stereotyped behaviours and challenging behaviour, there is also evidence to illustrate an association between repetitive or ritualistic behaviours and challenging behaviour, among children with severe ID (Oliver et al., 2012; Kanne & Mazurek, 2011). Further, research has also highlighted the associations between challenging behaviour and other behavioural correlates of challenging behaviour, such as over-activity and impulsivity, as is now discussed.

1.5.10. Challenging behaviour, over-activity and impulsivity

Impulsivity and over-activity are prevalent behaviours amongst individuals with ASD, with evidence to show an association between these behavioural correlates and the manifestation of self-injurious behaviour in these populations (Richards et al*.,* 2012; Richman et al*.,* 2013; Cooper et al., 2009). Interestingly, similar associations have been also documented in individuals with genetic syndromes. Most notably, in a multi-syndrome study, it was reported that over-activity and impulsivity were significantly associated with the presence of self-injurious and aggressive behaviour among individuals with genetic syndromes, such as FXS (Arron et al., 2011). The main hypothesis has been that an underlying executive dysfunction, evidenced by impulsivity, overactivity and compulsive behaviours, mediates the relationship between repetitive/stereotypic behaviours and self-injury (Bodfish *et al.,* 1995). In this study the relationship between repetitive/stereotyped behaviours and self-injury was examined genetically, but the absence of consideration of environmental determinants which might explain this relationship limits the generalisability of the findings. Given the fact that environmental factors were not assessed this might represent a threat to the study's ecological validity. Further studies need to be undertaken which take into account the environmental factors which might affect this relationship.

This model is supported by evidence that self-injury is associated with overactivity in adults with intellectual disability (Collacott *et al.,* 1998) and in individuals with Cornelia de Lange syndrome (Oliver, Sloneem, Hall & Arron, 2009). Hyperactivity has been found to be more common in individuals who display self-injury than in a matched comparison group who do not display self-injury (Schneider *et al.,* 1996). In addition, many individuals who engage in self-injurious behaviour also actively seek out and engage in self-restraint behaviours (Forman, Hall & Oliver, 2002; Fovel, Lash, Barron, & Roberts, 1989; Oliver, Murphy, Hall, Arron & Leggett, 2003). This may suggest that self-injury is not fully under the individual's control, perhaps due to a compromised ability to inhibit the behaviour. Compromised inhibition may influence self-injury in one or more ways (Barkley, 1997). It is plausible that impaired inhibition drives the individual to engage in self-injury as a prepatent response to triggering stimuli. Additionally, compromised inhibition may result in individuals being unable to terminate self-injury and thus self-restraint behaviours may reflect an attempt to suppress self-injury.

The relationship between self-injury and self-restraint was investigated in a group of individuals with CdLS (Hyman, Oliver & Hall, 2002). However, because there were no matched controls conclusions can only be drawn about this sample, rather than in relation to CdLS generally. This might represent a threat to the external validity of the finding. Hyman et al., (2002) found a significant association between self-injury and self-restraint. Importantly they also found that those individuals with CdLS who engaged in both self-injury and self-restraint displayed significantly more impulsive repetitive behaviours than those who did not display self-injurious or self-restraint behaviour.

The present section has highlighted the view that children with particular genetic syndromes and those who show ASD-types of behaviours may be at higher risk of displaying challenging behaviour compared to individuals with ID heterogeneous aetiology, and those who do not show repetitive, restricted and stereotyped behaviours. It is critical to understand the nature and contribution of such behavioural correlates to the manifestation of challenging behaviour in ID, which may also explain the individual differences observed in terms of the persistence of such behaviours. The following section expands on the persistent nature of challenging behaviour in individuals with ID.

1.6. The persistence of challenging behaviour

There is evidence to show that self-injurious behaviour tends to be persistent amongst individuals with ID. In a study examining the persistence and remission of challenging behaviour over the seven-year period from 1988 to 1995, it was found that 71% of individuals continued to display self-injury, and 29% of individuals were reported either to no longer show self-injury, or to show self-injury at a level which did not constitute a management problem (Emerson et al., 2001). Moreover, the same study also found that alongside self-injurious behaviour, 57% of participants also exhibited aggressive behaviour over the seven-year period, while 37% of participants also showed persistent property destruction (Emerson et al., 2001).

Further longitudinal evidence on the persistence of challenging behaviour has shown that 50% of adults with ID and challenging behaviour continued to display challenging behaviour over a two-year period (Totsika & Hastings, 2009). In addition, longitudinal data, examining the persistence of challenging behaviour over 20 years, showed that self-injurious behaviour was persistent in 84% of people with ID over a 20-year period (Taylor, Oliver & Murphy, 2011). However, the large variability in the scales used to assess challenging behaviours, and the subsequent definitions of persistence and change used in the studies included in the present review, suggest some need for caution when attempting to compare across the findings.

The prevalence, the characteristics associated with the presence, and the persistence of challenging behaviour were discussed in the relevant sections above. It has been shown that the individual and their behavioural characteristics are important for consideration when aiming to delineate the nature of the different types of challenging behaviour, and these characteristics may also explain some of the variation shown in the persistence over time of such behaviours. Although the association between personal characteristics and the presence of challenging behaviour has been explored, there are also data illustrating an association between genetic syndromes and the presence and severity of self-injury. Self-injurious behaviour has been identified to be more common in individuals with Lesch-Nyhan, fragile-X, Cornelia de Lange, Cri du Chat and Smith-Magenis syndromes (Arron et al., 2011). Moreover, the elevated prevalence rates of self-injury in relation to particular genetic syndromes, such as Lesch-Nyhan, fragile-X, Cornelia de Lange, Cri du Chat and Smith-Magenis syndromes, have been indicated previously (Arron, Oliver, Moss, Berg & Burbidge, 2011). In this connection, it would be useful now to review the existing causal models of self-injurious behaviour, as well as the potential causes of challenging behaviour that have been suggested in the extant literature. Specifically, in the following section, operant theories on the presence of self-injurious behaviour amongst individuals with ID will be explored. In addition, evidence highlighting an association between pain and self-injury is also reviewed.

1.7. Causes of challenging behaviour

The higher prevalence of self-injury associated with various genetic syndromes has given an opportunity to evaluate the causal models for self-injury. Firstly, the biological model has been developed to explore the biological basis of self-injurious behaviour in relation to particular genetic syndromes (e.g., Lesch-Nyhan syndrome). (Baumeister, Frye & Schroeder, 1985). The biological model for self-injurious behaviour proposes that differences in neurotransmitters and neuromodulators, and dopamine, serotonin and opioid peptides, in individuals with various genetic syndromes, such as Lesch-Nyhan syndrome, are associated with the presence of self-injurious behaviour (Schroeder et al., 2001; Kolevzon et al., 2010). Recently, May et al., 2009, found a known functional polymorphism in the Monoamine oxidase A (MAOA) promoter gene and problem behaviour in adult males with intellectual/developmental disabilities. More specifically, short alleles low expression MAOA was found in the 43% of participants who displayed challenging behaviour. In contrast only the 20% of participants without behavioural problems were found to have short alleles low expression MAOA. Although this finding underlines the biological cause of challenging behaviour in individuals with intellectual disabilities, in this study the behavioural data came from direct observations which were conducted by the residential staff. These findings could be replicated in a larger sample that would include measures of various topographical categories (e.g., aggression, property destruction, self-injury) and operant functions of problem behaviour (e.g., negative social reinforcement, positive social reinforcement, non-social reinforcement) to assess whether the MAOA promoter gene polymorphism differentially affects topographical and/or functional properties of problem behaviour in people with intellectual/developmental disabilities. In addition, this line of research warrants attention as it may, for example, offer a plausible account of within-syndrome variability regarding self-injury.

However, the most convincing models draw upon a phenotype and environment interaction to account for the greater prevalence and severity of self-injury (Langthorne & McGill, 2008; Oliver, 1993; Tunnicliffe & Oliver, 2011). The model that proposes an interaction between phenotype and environment noted that theories about genetic predisposition and about operant reinforcement remain quite distinct; neither theory type on its own is sufficient to explain challenging behaviour in genetic syndromes, and an integrated approach is required (Tunnicliffe & Oliver, 2011).Therefore, the causes of challenging behaviour are best studied using a bio-psychosocial model, to examine the behavioural and biological factors which may be involved in the development and maintenance of those behaviours (Xenidis, Russell & Murphy, 2001). In the following section, the operant theories on the presence of self-injurious behaviour in individuals with ID are explored.

1.7.1. Operant theories of challenging behaviour

There is evidence to show an association between environmental influences and the manifestation of self-injurious behaviour amongst individuals with ID. Operant theories suggest that self-injury is a functional, learned behaviour, which is broadly 'adaptive' (Emerson, 1998). The fundamental basis of the behavioural model of self-injurious and aggressive behaviour has been drawn from operant learning theory. Operant theories of self-injury have been supported by experimental studies that employ manipulations of antecedents and consequences, aiming to assess the individual's behaviour (Iwata, Dorsey, Slifer, Bauman & Richman, 1994). Operant theories suggest that self-injury and aggression are learned and maintained through either positive or negative reinforcement, which is delivered contingently on the presentation of the behaviour. For instance, positive social reinforcement has been documented when the adult–child interaction profile was found to be consistent with socially positive reinforcement of self-injurious behaviour (Oliver et al., 2005). In addition, an example of negative reinforcement might include the removal of aversive stimuli, such as tasks to be undertaken or demands that are placed upon the individual (Carr, 1977). The function of mutual social reinforcement suggests that self-injury and aggression can be rewarded by others' responses; and the rewarding responses of others themselves can be negatively reinforced by the termination of self-injury (Oliver, 1995; Oliver, Hall & Murphy, 2005).

In addition, there is evidence to suggest that self-injurious behaviour might be maintained by social reinforcement. In a study looking at the self-injurious behaviour of 152 individuals with intellectual disability, it was found that 38% of them evidenced self-injury maintained by social negative reinforcement, 26% by social positive reinforcement, 21% by positive automatic reinforcement, 5% by multiple controlled reinforcement (positive and negative) and only 10% showed undifferentiated patterns (Iwata et al., 1994). The results of this study underline the effectiveness of functional analysis in identifying the environmental causes of self-injurious or aggressive behaviour among individuals with ID. Functional analysis includes a series of paradigms, which involve contrasting the frequency of target behaviours across different experimental conditions, within which antecedents (e.g. low levels of adult attention, requirement to perform an academic task) and/or consequences (e.g. delivery of attention, removal of task) of behaviour are varied systematically. Such paradigms have formed the basis of a body of literature identifying operant functions for behaviours such as aggression and self-injury (Hanley, Iwata & McCord, 2003).

In summary, operant theories contribute to an understanding of how self-injury may be maintained and shaped by environmental contingencies. It will be useful to consider principles from operant theory in relation to the development and maintenance of other forms of challenging behaviour, except self-injurious behaviour, in individuals with ID. It has been suggested already that the causes of challenging behaviour should be examined using the bio-psychosocial model (Xenidis, Russell & Murphy, 2001). Therefore, alongside operant theories, which explain the maintenance of challenging behaviour among individuals with ID, pain theories have been proposed to describe the maintenance of challenging behaviour amongst individuals with ID. In the following section, the association between the presence of challenging behaviour and pain is explored.

1.7.2. Pain associated with challenging behaviour

The association between pain and challenging behaviour has been documented in a range of behavioural studies (Carr & Owen-DeSchryver, 2007; O'Reilly, 1997; Davies, 2010; Richards, 2011). Moreover, there are studies showing that individuals diagnosed with particular genetic syndromes associated with intellectual disability have been reported to experience a high prevalence of painful health problems (Berg, Arron, Burbidge, Moss & Oliver, 2007). In a study that was conducted by Selicorni et al., (2005) it was found that kidney dysfunctions and urinary tract pain were present in the 40% of participants with Cornelia de Lange syndrome. There is evidence about the association between pain and self-injurious behaviour (Luzzani, Macchini, Valade, Milani & Selicorni, 2003; Breau, Finley, McGrath & Camfield, 2002). Most notably, Luzzani et al., (2003) found that gastro-oesophageal reflux issues were strongly associated with the presence of self-injurious behaviour in children and in young adults with Cornelia de Lange syndrome. Moreover, it has been reported that visual and tactile problems were found to be associated with the presence of self-injurious, stereotypical and challenging behaviour in general (Poppes, van der Putten & Vlaskamp, 2010).

Most notably, a single case study using a validated measure of pain-related behaviour (Non-Communicating Children's Pain Checklist-Revised, NCCPC-R; Breau, Finley, McGrath & Camfield, 2002) demonstrated that severe self-injurious behaviour was significantly associated over time with higher levels of pain-related behaviour (Symons & Danov, 2005). Similarly, O' Reilly (1997) and Christensen et al. (2009) reported case studies of children with ID who displayed self-injury associated with otitis media and constipation. Cohort studies have also reported associations between self-injury and pain problems. It has been found that data associating physical illness (e.g., coughs, colds, constipation, cuts, ear infections etc.) and menstrual discomfort were associated with the presence of self-injurious behaviour amongst individuals with ID (Carr & Owen-Deschryver, 2007).

Furthermore, the association between health problems and self-injury was evidenced in a study screening for risk markers of challenging behaviour, where the presence of one or more health problems was found to be significantly correlated with the presence and severity of self-injury in children aged between 2 and 12 (Davies, 2010). In addition to the studies investigating the association between the presence of challenging behaviour and pain, there is also evidence to show that self-injury was reduced when the medical problems that were found to be associated with the presence of behavioural problems were treated (Bosch et al*.,* 1997).

However, although there is an increasing consensus on the relationship between self-injury and pain, there is no research to date that has investigated the relationship between pain and aggressive behaviour in individuals with ID (Davies, 2010; Poppes, van der Putten & Vlaskamp, 2010). A major challenge in this area of inquiry may relate to the difficulties that individuals with profound ID can experience in effectively reporting their pain. Therefore, there is very limited evidence from investigating pain and its association with challenging behaviour in these populations. More specifically, the relationship between pain and self-injury, using an observational assessment of pain in non-communicating children, has not been evident (Breau et al., 2003). However, Symons et al., (2009), by using the same observational approach, reported a significant positive relationship between observable pain behaviour and self-injurious behaviour in an adult population. In a recent review, Devine and Symons (2013) report that the available evidence indicates that increased rather than decreased sensitivity to pain may be more influential in the expression of self-injurious behaviour. Overall, whilst a number of studies support an association between self-injury and pain amongst individuals with ID, further work to support this evidence is required. However, further research on the relationship between pain and other forms of challenging behaviour, such as aggressive behaviour, is yet to be conducted.

In summary, the causes of challenging behaviour and the association between pain and self-injury have been described from the literature. It has been suggested that challenging behaviour should be approached using a bio-psychosocial model to determine the causes of challenging behaviour in individuals with ID. Despite the contrasts in causal explanations for the presence and development of challenging behaviour, research findings are broadly consistent when detailing the consequences of challenging behaviour upon the individual who displays the behaviour, and upon those caring for the individual. In the following section, the relationship between challenging behaviour, parental emotional distress, child quality of life and service use is explored.

1.8. The Impact of challenging behaviour

1.8.1. Challenging behaviour and parental well-being

The association between challenging behaviour and parental emotional well-being has been documented in the literature. There is emerging evidence to show that parents of children with ID are at a greater risk of experiencing psychological distress. More specifically, it has been found that a child's challenging behaviour is strongly associated with maternal anxiety and/or depression (Hastings, 2003; Hastings & Brown, 2002; Baxter, Cummins & Yiolitis, 2000). Moreover, a model in the literature highlights the interrelationship between parental emotional distress and challenging behaviour (Hastings, 2002), where child behaviour problems are assumed to influence parental stress, and vice versa.

Group designs have been employed that demonstrate that stress in parents of children with developmental disability who also have significant behaviour problems is similar in intensity to the stress reported by parents of children without disabilities, but who also have significant behaviour disorder (Donenberg & Baker,1993; Dumas, Wolf, Fisman & Culligan, 1991; Floyd & Gallagher,1997). Thus, even when adaptive and intellectual disability varies between groups, parental stress is elevated in the face of significant child behavioural problems. Correlational studies have also shown that child behavioural problems predict parental stress when other family variables (e.g. socio-economic status, family size and social support) have been controlled (Konstantareas and Homatidis, 1989; Sloper et al., 1991). However, studies have shown that notwithstanding these variables other important variables, such as parental coping strategies, may mediate the impact of child behavioural problems on parental stress (Sloper et al., 1991). Although, there is evidence suggesting an interrelationship between parental emotional wellbeing and challenging behaviour (Hastings, 2003) authors have counted, under the umbrella term of challenging behaviour, impulsivity and overactivity as behavioural problems rather as behavioural correlates of challenging behaviour. Although, this study describes the relationship between parental mental health and children's behavioural problems there are some limitations which might threaten the external validity of the study. First, the sample was small and, therefore, lacked statistical power. Second, the homogeneity of the sample at the level of aetiology limits the generalisability of the results. Finally, by assessing child behaviour problems via teacher ratings, a further potential limitation is introduced. Behaviour problems are often expressed differently depending on the context, and the dimensions of behaviour reported by teachers might not be those that affect parental well-being.

It is essential that the interrelationship between challenging behaviour itself, behavioural correlates of challenging behaviour and parental emotional stress be further investigated. In addition to the association of challenging behaviour with parental wellbeing, there is evidence to suggest that challenging behaviour might also be associated with the quality of life of children with ID. In the following section, the association between challenging behaviour itself and children's quality of life is explored.

1.8.2. Challenging behaviour and the child's quality of life

Children with ID face a range of difficulties in their daily lives that can relate to the individual characteristics that can affect their quality of life. More specifically, there is evidence from parental reports to show that child characteristics, such as age, gender, severity of disease, and pain might be associated with the quality of life of children with cerebral palsy (Arnaud et al.,2010; Varni et al., 2005). In addition, there is evidence that parents of children with autism more frequently report that their children are experiencing a diminished quality of life, compared to parents of children with ADHD or children without disabilities (Lee, Harrington, Louie &

Newschaffer, 2008). More specifically, parents of children with ASD, who also exhibit self-injurious behaviour, reported that their children experienced a diminished health-related quality of life, and were also found to be 20% more likely to be hospitalised due to injuries in the hospital setting compared to children without self-injurious behaviour (McDermott, Zhou & Mann, 2008).

Furthermore, it has been reported that individuals with challenging behaviour may experience a diminished quality of life, compared to those who do not engage in challenging behaviour (Beadle-Brown, Murphy and DiTerlizzi, 2009). However, in this study parents of adults with mild/moderate intellectual disabilities responded regarding individuals' quality of life and adaptive skills. In addition, there was no specification about challenging behaviour and what behaviours were counted under the challenging behaviour umbrella term. It is necessary, therefore, that the quality of life of children with intellectual disabilities and challenging behaviour should be further investigated.

In summary, there is increasing evidence to highlight the role of challenging behaviour in affecting the quality of life of children with ID. Further investigation of the exact nature of such associations could have a critical impact on service provision that aims to help families to effectively manage challenging behaviour. In the following section, this area of inquiry is discussed.

1.8.3. Challenging behaviour and service provision

Families of children with challenging behaviour will often seek access to particular services to manage behavioural problems. However, it was found that individuals with aggressive behaviour or engaging in property destruction were at higher risk for out-of-borough placement as a consequence of their behaviour (Joyce, Ditchfield & Harris, 2001). Similarly, it was reported that a quarter of all study participants exhibiting challenging behaviour required significant support with behaviour intervention and medical issues, whilst more than half were rated as needing significant support with daily life skills, communication and education (Ruddick, Bacarese-Hamilton, Davies & Oliver, 2015). However, in relation to this present study several methodological issues have been identified. Most notably, intellectual disability was not clearly defined, which might represent a threat to the internal validity of the study regarding the constructs that are being assessed. Also data regarding visits to relevant professionals were not verified with professionals but relied on the primary carers' responses, which might misrepresent the exact number of visits.

Moreover, the presence of self-injury is known to increase the likelihood of reactive physical interventions and emergency medication use (Allen, Lowe, Brophy & Moore, 2009). It is apparent that individuals with challenging behaviour may seek service support much more often than the general public, and the absence of the immediate availability of support might have an impact on the manifestation of challenging behaviour itself.

There is some evidence to suggest that there are particular demographic characteristics, such as age, gender, ethnicity (Leslie et al*.,* 2000), parental mental health (Cornelius et al*.,* 2001) and socio-economic status (Zahner & Daskalakis, 1997) which may influence families' use of particular services. However, to date, there is not a clear consensus about the exact nature of these factors and the degree of their impact upon challenging behaviour. Further investigation in this area would be valuable.

1.9. Summary

The research reviewed in this chapter has highlighted the implication of a number of personal characteristics in relation to the manifestation of challenging behaviour in populations with intellectual disability of heterogeneous aetiology. More specifically, a range of behavioural and health conditions has been found to be associated with the presence of challenging behaviour. Moreover, the persistence that normally accompanies challenging behaviour in individuals with ID might suggest that challenging behaviour could be best described within a single genetic syndrome. Therefore it would be essential for the persistence of challenging behaviour in individuals with FXS to be investigated by employing longitudinal approaches. Similarly, noting the high prevalence rate of ASD amongst individuals with FXS (Hall, Lightbody & Reiss, 2008; Richards et al., 2012), a longitudinal study of the persistence of challenging behaviour in this particular syndrome group would provide further understanding of whether ASD diagnosis or ASD-types of behaviours are associated with the presence and persistence of challenging behaviour. In addition, the association between the behavioural correlates of challenging behaviour and their persistence alongside the presence of challenging behaviour remains to be further investigated.

Furthermore, the present chapter has reported the interconnection between challenging behaviour, parental emotional distress, child quality of life, and service use. Although the association between challenging behaviour and parental emotional distress has been investigated previously, it is essential to further delineate whether challenging behaviour itself, or behavioural correlates of challenging behaviour, might be associated with parental emotional distress. Despite the fact that previous research found high levels of challenging behaviour in individuals with particular genetic syndromes associated with ID, there has been only limited research investigating the potential association between challenging behaviour, parental anxiety and/or depression and the child's quality of life. This area of inquiry requires further delineation.

A complementary issue raised in the present study is the investigation of the individual personal and the family characteristics that might determine service use for families of children with ID and additional behavioural problems. This area of inquiry has been particularly under-researched, with only limited research evidence available to show the factors that determine service use for families of children with ID and challenging behaviour. An improved understanding of risk markers and behavioural correlates of challenging behaviour in children with ID might help lead to a comprehensive explanation of the relationship between parental mental health and the child's quality of life, in relation to challenging behaviour.

1.10. Conclusion, and aims of this thesis

The research reviewed in the present chapter has critically investigated the prevalence, persistence, the associated behavioural correlates of challenging behaviour, and the theoretical causes of challenging behaviour in individuals with ID. Additionally, the association between challenging behaviour, parental emotional distress, child quality of life and service use was also reviewed. This thesis consequently had a number of specific aims prompted by the reviewed research. These aims are presented below, together with a discussion of their rationales.

- The prevalence and the persistence of challenging behaviour in individuals with ID have been presented (Section 1.5 and 1.6). It has been suggested that challenging behaviour tends to be prevalent among people with particular genetic syndromes associated with ID. In addition, it has been found that there are particular behavioural correlates of challenging behaviour, such as impulsivity, over-activity, and repetitive, restricted and stereotyped behaviours, which are associated with the presence of challenging behaviour and are also characterised as ASD-types of behaviours. In addition, pain problems are also found to be associated with the prevalence of challenging behaviour among individuals with ID. It is advantageous to examine the persistent nature of challenging behaviour and the associated characteristics in relation to genetic syndromes, in which there is evidence from the literature about the high prevalence of self-injurious and aggressive behaviour. Taking note of the relationship between ASD-types of behaviours and challenging behaviour, it is essential for these behaviours to be prevalent in this syndrome group in order to evaluate whether ASD diagnosis or ASD-types of behaviours are associated with the persistence of challenging behaviour. Thus Chapter 2 presents a longitudinal analysis, describing the prevalence and the persistence of challenging behaviour in individuals with FXS.

Analysing existing data is a procedure with several pros and cons. The main advantage of analysing existing data is the low cost. Also, the data posted online are usually cleaned by professional staff members who often provide detailed documentation about the data collection and data cleaning process. Moreover, teams conducting large-scale population-based surveys whose results are made available to others usually employ statisticians to generate ready-to-use survey weights and design variables – something that most users of the data are unable to do – so this helps data users make necessary adjustments to their estimates. The increasing availability of such data encourages the creative use and cross-linking of information from different data sources. For example, experts in using hierarchical models can combine data from individual surveys with aggregate data from different administrative levels of a community (e.g., village, township, county, province, etc.) to examine the factors associated with health- related outcomes at each level.

The availability of such databases also provides statisticians with real-life data to test new statistical models. Such analyses could identify potential new interventions for existing problems that can subsequently be tested in prospective studies. However, the are several disadvantages in analysing existing data. The main disadvantage is that the available data are not collected to address the particular research question or to test the particular hypothesis. It is not uncommon that some important third variables were not available for the analysis. Similarly, the data may not be collected for all population subgroups of interest or for all geographic regions of interest. Another major limitation of the analysis of existing data is that the researchers who analyse the data are not usually the same individuals as those involved in the data collection process. Therefore they are probably unaware of study-specific nuances or glitches in the data collection process that may be important in the interpretation of specific variables in the dataset. Despite such limitations, this study's longitudinal analysis aims to investigate the persistent nature of challenging behaviour in relation to individuals with FXS.

- In the present study, the previously identified association between challenging behaviour, parental emotional distress and children's quality of life is investigated. It was found previously that although challenging behaviour is associated with either parental anxiety or depression, it is not yet clear whether challenging behaviour itself or behavioural correlates of challenging behaviour are associated with parental emotional distress. In addition, it is judged essential that for the first time the quality of life of children with ID and challenging behaviour is to be explored by investigating the association between challenging behaviour itself and behavioural correlates of challenging behaviour, and children's quality of life. Finally, it is necessary for challenging behaviour, parental emotional distress and children's quality of life, as three distinct strands, to be placed together in investigating whether there is an interrelationship between these strands. Therefore Chapter 3 presents a further investigation of the interrelationship between challenging behaviour, parental emotional wellbeing and the quality of life of children with ID.
- The earlier delineation of the interrelationship between challenging behaviour, parental emotional distress and the quality of life of children with challenging behaviour, was influential

in the exploration of service use for families of children with ID and challenging behaviour. Section 1.8.3 reviewed the relationship between challenging behaviour and service use among families of children with ID. There is a need for parental satisfaction to be explored, additionally in terms of the usefulness and effectiveness of the accessed services. Therefore, Chapter 4 provides an extensive exploration of service use by families of children with challenging behaviour.

- Finally, the exploration of service use for families of children with ID and challenging behaviour, as set out in Chapter 4, is essential for the investigation of the putative factors that are involved in service use for these particular families. Section 1.8.3 reviewed the association between particular demographic characteristics of children, such as age, gender, ability level and service use among families of children with ID. In addition, parental socio-economic status is also associated with service use (see Section 1.8.3). Moreover, it is judged as essential that service use in respect of different forms of challenging behaviour is now investigated. Therefore, Chapter 5 aims to explore whether there are any differences in the forms of challenging behaviour between those children who do and do not access services, and then to explore if any of the factors proposed in the literature about service use for families of children with ID can also predict service use for families of children with challenging behaviour and ID.

CHAPTER 2

The persistence of self-injurious and aggressive behaviour in children and adults with FXS

2.1. Preface

In the previous chapter, research was summarised which investigated the prevalence, personal characteristics associated with challenging behaviour, and the persistence of both self-injurious and aggressive behaviour. A number of areas for future research was identified. A pressing need was identified to describe the persistence of both self-injurious behaviour and aggression, using robust measures. Given the documented heterogeneity of prevalence figures across genetic syndromes, the study of one high risk syndrome in a study of persistence affords the opportunity to reduce such heterogeneity. To this end, the persistence of challenging behaviour, and characteristics related to persistence, in children and adults with FXS, is the focus of this empirical study.

2.2. Introduction

Two forms of challenging behaviour, self-injury and aggression, have been found to be particularly problematic for individuals with intellectual disability, and their families and carers, as well as for clinical services (see Section 1.5). The following section explores the characteristics that might predict the persistence of self-injurious and aggressive behaviour among individuals with ID.

2.2.1. Predicting the persistence of self-injurious and aggressive behaviour

Prevalence rates for aggressive and self-injurious behaviour vary between 4% (Cohen et al., 2010; Cooper et al., 2009; Holden & Gitlesen, 2006) and 17% for self-injury, and 30 to 40% for aggressive behaviour amongst individuals with ID (Collacott, Cooper, Branford & McGrother, 1998). The relationship between age and self-injurious and aggressive behaviour has been explored in the literature using a number of strategies (Collacott et al., 1998; Cooper et al., 2009; Holden & Gitlesen, 2006). In a recent review aggressive behaviour and self-injury were associated with age (Davies & Oliver, 2013). Most notably, aggressive behaviour increases during childhood, adolescence and young adulthood, and decreases after the age of 50; and people aged between 32 and 61 were at significantly less risk of self-injurious behaviour compared to those aged between 12 and 31. Moreover, it has been found that age is associated with the severity of self-injurious behaviour, with older people reported to have less severe self-injury (Davies & Oliver, 2013).

In addition to the evidence highlighting the association between age and challenging behaviour among individuals with ID, there is also a line of research illustrating the relationship between the persistence of self-injury and particular topographies of self-injury.

Most notably, the presence of head banging/hitting topographies of self-injurious behaviour is associated with the persistence of self-injurious behaviour (Emerson et al*.,* 2001; Taylor, Oliver & Murphy, 2010). This may support operant theories, which suggest that head banging can influence the behaviour of others and thus can predict the persistence of self-injurious behaviour. It has been hypothesised that the response efficiency of head hitting/banging may be greater than the response efficiency for other forms of self-injurious behaviour (Emerson et al., 2001).

There is additional evidence of the relationship between particular topographies of self-injury, and the persistence of self-injurious behaviour among individuals with ID. Most notably, head banging/hitting might be related to pain in individuals with ID (Luzzani, Macchini, Valade, Milani & Selicorni, (2003); see Section 1.7.2). However, the particular mechanisms underlying the relationship between topography and persistence need to be further investigated. Moreover, there are data illustrating a relationship between the persistence of self-injury and stereotyped behaviours (Berkson, Tupa & Sherman, 2001). In addition, there is evidence to show that resistance to sameness, and impulsivity, which are common behaviours in individuals with ASD, as well as with an additional diagnosis of autism, are strongly associated with the persistence of challenging behaviour in individuals with intellectual disability. (Murphy et al*.,* 2005). Finally, there is evidence to illustrate an association between the persistence of aggressive behaviour and a diagnosis of ADHD (Cooper et al*.,* 2009).

This line of research indicates that the persistence of challenging behaviour can be associated with age, particular topographies of self-injurious behaviour, and the presence of stereotyped, restricted behaviours, impulsivity and over-activity. Interestingly, these correlates of challenging behaviour are common behaviours among individuals with autism and/or ADHD. Although there is evidence describing the association between challenging behaviour and genetic syndromes (Arron et al., 2011), a longitudinal analysis of the persistence of challenging behaviour in individuals within syndromes, such as FXS, could help evaluate whether particular behaviours, such as stereotyped, compulsive behaviours, impulsivity and over-activity, which are common in individuals with FXS, can predict the persistence of challenging behaviour. In the following section, the behavioural phenotype of FXS is summarised.

2.2.2. The behavioural phenotype of FXS

FXS is caused by an expansion of CCG repeats on the X chromosome and it has been previously estimated that about 1 in 4,000 males and 1 in 8,000 females (Crawford, Acuna, & Sherman, 2001) are born with FXS. Gender differences on IQ scores have also been observed, showing that females with the full mutation were less intellectually affected compared to males. Most notably, it has been found that 50% to 70% of females with the full mutation (where the FMR-1 gene defect has >200 repeats) scored approximately 85 in IQ tests; and scores for males have been found to decline more rapidly during childhood, compared to females, who show a greater stability with maturation (Wright-Talamante et al., 1996; Fisch et al., 1999). Additionally, evidence has shown that receptive language skills are better in females with FXS than in males with FXS (Abbeduto et al. 2003). The behavioural phenotype among children with FXS differs between males and females, where males show more behavioural problems compared to females (Smith, Barker, Seltzer, Abbeduto & Greenberg, 2012). Finally, there are data illustrating an association between ASD and FXS. The association between ASD phenomena and FXS is summarised in the following section.

2.2.3. FXS and ASD

The association between FXS and ASD has been observed in several studies in which individuals with FXS score highly on diagnostic measures for Autism Spectrum Disorders (Demark, Feldman, & Holden, 2003). Moreover, cognitive deficits in individuals with FXS have been found to be associated with autistic behaviours in individuals with FXS (Loesch et al., 2007). Most notably, it has been reported that individuals with FXS experience social problems and exhibit stereotypic motor behaviours (Hatton et al. 2002). Interestingly, there is also evidence to illustrate that individuals with ASD and FXS show similar behavioural phenotypes (Smith et al., 2012). It has also been found that hyperactivity, stereotyped behaviours, impulsivity, and repetitive speech, are common behaviours in individuals with FXS and ASD (Baumgardner, Reiss, Freund & Abrams, 1995) and that hyperactivity and attention deficits are the most common behavioural problems amongst individuals with FXS at younger ages (Munir et al., 2000; Farzin et al., 2006). Social escape behaviour, such as face hiding and eye-rubbing, have also been found to be common in individuals with FXS (Hall, Bernardis & Reiss, 2006). Face hiding was reported to occur in 74 % of male participants and in 54% of female participants, indicating that this behaviour is a major component of social escape behaviour in individuals with FXS.

In addition to the above associations, there are data illustrating an association between ASD-associated behaviours and the presence of challenging behaviour within FXS. Most generally, the association between ASD phenomena and self-injurious behaviour has been explored in many studies (Bhaumik et al., 1997; Collacott et al., 1998). There is evidence to show that ASD- related behaviours are associated with the presence of self-injurious behaviour (Lowe et al., 2007; Murphy et al., 2005). Autistic traits, such as sociocommunication impairments, stereotyped movements, and poor eye contact, occur in the majority of individuals with FXS (Moss et al., 2013). The results from this study suggest that ASD phenomena may be useful putative risk markers for self-injurious behaviour within multiple populations. Supporting this assertion, there is evidence to show that ASD phenomena in individuals with various genetic syndromes are strongly associated with the presence of self-injury. Most notably, the Social Communication Questionnaire (SCQ, Rutter et al., 2003) has been utilised with individuals with Cornelia de Lange, fragile X, Prader-Willi and Lowe syndromes to identify associations between specific areas of the triad of impairments of ASD (i.e., repetitive behaviour, impulsivity and over-activity) and self-injurious behaviour (Arron et al., 2011). Taken together, this research evidence demonstrates that it is the presence of ASD phenomena rather than a diagnosis of idiopathic autism, that is associated with the presence of self-injury. Exploring ASD- related behaviours in individuals with FXS will enable the persistence of self-injurious behaviour in this group of people to be investigated. The following section expands on this area of inquiry.

2.2.4. Challenging behaviour in individuals with FXS

Challenging behaviour is part of the behavioural phenotype of individuals with FXS, with 58% of individuals with FXS reported to self-injure at some point during their lifetime (Symons, Clark, Hatton, Skinner & Bailey, 2003). Additionally, it has been found that 51% of individuals with FXS exhibited self-injurious behaviour within the previous three months (Arron et al., 2011). Similar evidence of the high prevalence of self-injurious behaviour in this syndrome group was presented in Richards et al., (2012), who found that 54.4% of individuals with FXS displayed self-injurious behaviour.

As noted above, there is overlap between the behavioural phenotypes of individuals with FXS and individuals with idiopathic autism. Similar prevalence rates of self-injurious behaviour are reported for both individuals with autism and those with FXS (Richards, Oliver, Nelson & Moss, 2012; Arron et al., 2011). In both of these studies approximately 50% of individuals with FXS displayed self-injurious behaviour. This line of research highlights the strong relationship between ASD- related behaviours in FXS and self-injurious behaviour (see Section 2.1.1).

Taking into account the high prevalence of ASD symptomatology amongst individuals with FXS and the occurrence of ASD- related behaviours alongside over-activity and impulsivity, that are associated with the presence of challenging behaviour in individuals with FXS, it is important to examine the developmental trajectories of these associations. The delineation of the developmental trajectories of such associations may help in identifying potential early risk markers of challenging behaviour in young populations with FXS. In the following section the developmental changes and their possible relationship with the persistence of self-injury and aggression amongst individuals will FXS will be explored.

2.2.5. Phenotypic developmental changes in FXS

There is a line of research investigating the developmental trajectories associated with the behavioural phenotype of individuals with FXS. In a longitudinal study covering the age span of 1 to 14 years, small but significant age-related increases in the symptom severity of autism for a sample of 116 children with FXS are reported (Hatton et al., 2006). However, although the Hatton et al, (2006) cohort extended into early adolescence, most participants were quite young, with the mean age of the participants being under five years at the initial visit. Additional evidence of the relationship between ASD- related behaviours and age in children is provided by Bailey, Hatton, Skinner & Mesibov, (2001). In this later study males with FXS, who also scored high for ASD characteristics, were found to have significantly lower developmental levels and slower trajectories of growth than did males with FXS only. In addition, there is evidence to show a long- term association between ASD and age in individuals with FXS, with the symptoms of autism reported to remain stable during adulthood (Sabaratnam, Murthy, Wijeratne, Payne, & Buckingham, 2003). However, the mean age of participants in this study was over 46 years old, which limits conclusions about the broader association between age and ASD, due to the exclusion of younger ages. Taking these together, it is likely that the associations between ASD symptomatology and FXS can be investigated through the exploration of the differences between ASD-only groups of individuals and groups of individuals having both ASD and FXS. In line with this theme, there is evidence to show that individuals with FXS and an additional autism diagnosis, and those with FXS only, differ significantly in the communication and restricted interests/repetitive behaviours domains, but not in the reciprocal social interaction domain (McDuffie et al*.,* 2010). Most notably, in that study the FXS+ASD and FXS - only groups differ significantly in terms of stereotypic utterances, delayed echolalia, pointing to express interest and non-verbal IQ. In terms of behaviour the groups differ significantly in repetitive object use, circumscribed interests and verbal rituals. Overall, the severity of autism symptoms improved with age for all participants, with the least improvement noted for restricted interests and repetitive behaviours domains.

In addition to the relationship between autism, FXS and age, the relationship between age and cognitive abilities among individuals with FXS has been reported in the literature (see Section 1.5). Cognitive abilities appear to be associated with the persistence of challenging behaviour among individuals with FXS (McDuffie et al*.,* 2010). Interestingly, specific deficits in the cognitive abilities of individuals with FXS have been reported in the literature. Most notably, problems in attention switching and control have been reported as fundamental deficits in the fragile X full mutation from infancy, through childhood and into adulthood (Cornish et al., 2008). Similar findings on the deficits in cognitive abilities among children with FXS have been also reported (Fisch, Simensen & Schroer, 2002). Young verbal children with autism initially exhibit lower intellectual abilities compared to children of the same ages diagnosed with both FXS and autism. In a similar vein, declines in IQ scores were reported for children with FXS and autism, whereas children with autism remained level on IQ scores (Fisch, Simensen & Schroer, 2002). In summary, there is evidence that individuals with FXS who score high on autism assessments, and on the associated behaviours, continue to display these behaviours over time, whereas a decline in cognitive abilities is also reported.

In addition to the above associations, it is helpful for the putative associations between self-injurious behaviour (SIB), aggression and FXS to be reviewed. In particular, it is important to delineate the persistence of self-injurious behaviour and aggression in individuals with FXS, as well as the associated characteristics of challenging behaviour in individuals with FXS. Such investigation helps to understand and distinguish the putative risk markers of challenging behaviour amongst individuals with FXS, and also to inform services in the interests of providing early and targeted interventions that aim to increase the overall quality of life. The following section expands the description of this area of inquiry.

2.3. The relationship between FXS, SIB and aggression

There is evidence to show that a high percentage of individuals with FXS engage in SIB (Arron et al., 2011; Richards et al., 2012; see also Section 1.5.3). In addition, there is evidence on the phenomena of self-injurious behaviour in this particular population, with hand biting commonly reported (Symons et al., 2003). In the same study, it was reported that change in the individual's routine and the presentation of difficult demands were the most common causes of self-injurious behaviour in individuals with FXS. Hand-biting occurred in 26% of male participants and in 15% of female participants. Richards et al., (2012) reported that 54.5 % of individuals with FXS exhibited self-injurious behaviour. Individuals with FXS were almost three times more likely than individuals with Down's syndrome to exhibit self-injurious behaviour. Interestingly, no significant difference was evident in the prevalence of SIB between individuals with autism and individuals with FXS. Self-biting was the most common form of self-injurious behaviour for individuals with FXS (Richards et al., 2012). In a similar study self-hitting was shown by 50% of the sample with 30% self-biting (Hessl et al., 2007).

In addition to the prevalence and the expression of SIB in individuals with FXS, there are also data that describe the prevalence and phenomena of aggression. Approximately, 50% of participants with FXS showed aggression (Arron et al., 2011). The most common aggressive behaviours were hitting others (49% of the sample) and kicking others (30%) Hessl et al., 2007).

Self-injury and aggression have been found to be associated with particular personal characteristics. Most notably, aggressive behaviour is more prevalent at younger ages (Arron et al., 2011). In addition, autistic-like behaviours, such as repetitive behaviour, over-activity and impulsivity, are associated with the presence of self-injurious behaviour in individuals with FXS. Over-activity has been also found to be associated with the presence of aggressive behaviour (Arron et al., 2011). Moreover, low scores on measures registering interest and pleasure and high scores on autistic- like social interaction are associated with the presence of self-injurious behaviour among individuals with FXS (Arron et al., 2011). In summary, taking into account the high prevalence of ASD- related behaviours in this syndrome group and the strong association between ASD- related behaviours and challenging behaviour, it is essential that the prevalence of challenging behaviour and the characteristics associated with challenging behaviour are investigated longitudinally.

In summary, in this section, available information on challenging behaviour in FXS has been summarised by exploring the prevalence data for self-injurious and aggressive behaviour in this syndrome group. Most notably, ASD- related behaviours and impulsivity and over-activity, which are prevalent amongst individuals with FXS, are found to be associated with the presence of challenging behaviour. Therefore, the persistence of self-injurious and aggressive behaviour among individuals with FXS, and the associated behavioural characteristics, can be investigated to elucidate the relationship.

2.3.1. Exploring the persistence of self-injurious and aggressive behaviour in FXS

There is evidence that self-injury tends to be persistent over time in individuals with FXS, with 81% of participants who manifested SIB reported to continue engaging in self-injurious behaviour within one month from the original report (Symons, Clark, Hatton, Skinner & Bailey, 2003). The mean age of participants who took part in the Symons et al. (2003) study was 80 months. However, to assess the persistence of self-injurious behaviour a longer time period may need to be considered. Self-injurious behaviour has been shown to increase during childhood and adolescence but to decrease during adulthood (see also Section 1.5). In a similar vein, challenging behaviour in FXS is persistent in this syndrome group (Hatton et al., 2002). Taking into account the relationship between age and self-injurious behaviour, it is important that the persistence of challenging behaviour is investigated across a wide age range.

2.3.2. Characteristics which predict the persistence of self-injurious and aggressive behaviour in FXS

There is evidence to show that the persistence of self-injury and aggression in FXS can be predicted by particular characteristics (Hatton et al., 2002). Most notably, autism- related behaviours and impulsivity and over-activity, can predict the presence of behavioural problems in children with FXS. Although there is some evidence showing that both self-injury and aggression tend to be persistent over time in children with FXS (Symons, Clark, Hatton, Skinner & Bailey, 2003; Skinner, Sullivan & Wheeler, 2002), there is currently no empirical evidence on the persistence of challenging behaviour during adolescence and adulthood.

2.4. Aims

The current study will fill an existing gap in the literature by delineating the persistence of both self-injurious behaviour and aggression in individuals with FXS, across childhood, adolescence and early adulthood, and investigating the characteristics that might be associated with the persistence of challenging behaviour in this syndrome group. By employing a longitudinal analysis, the study will explore the persistence and associated behavioural characteristics of self-injury and aggression in individuals with FXS. Through analysing challenging behaviour data from different time points [i.e., time 1 (2003), time 2 (2006) and time 3 (2011)] for participants with FXS, two key areas will be investigated:

i) The persistence of self-injury and aggression in FXS will be investigated longitudinally across the three time points.

ii) The association between ASD- related behaviours, and the persistence of self-injury and aggression among individuals with FXS will be investigated. Consistent with previous research, it is predicted that the persistence of self-injury and aggression will be related to higher levels of autistic-related behaviour in relation to individuals with FXS.

2.5. Methods

2.5.1. Recruitment

Data on the challenging behaviour of participants with FXS were extracted from existing databases from studies conducted within the Cerebra Centre for Neurodevelopmental Disorders at the University of Birmingham, U.K. School of Psychology, from 2003 to 2011. The studies were conducted during this time with the broader aim of examining the behavioural phenotypes of syndromes. This present longitudinal investigation extracted behavioural data collected at different time points from the same male participants diagnosed with FXS. Participants with missing data at any of the time points were excluded from participation.

2.5.2. Participants

Behavioural data from three different time points from eighty-one male participants with FXS were available for the present study. Two of the participants with missing data were excluded from analysis; therefore, the total sample size of the study was 79 individuals. Demographic characteristics of the participants are presented in Table 2.1. Descriptive analysis showed that the majority of the participants had acquired verbal skills, had a normal range of hearing and vision abilities, were mobile, and their carers reported that they are able with regard to self-help, based on the Wessex scale (Kushlick et al*.,* 1973) described in Section 2.4.3.1.

Table.2.1. Participant Descriptive Table

		Baseline (2003- 2004)	Follow up 3 (2011)
N		79	79
Age	Mean	17.03	24.25
	(SD)	9.11	8.90
	Range	6-47	14-54
Gender	% male	100	100
Self-Help	% partly able/ able	85.2	91.1
Mobility	% mobile	98.7	98.7
Speech	% verbal	87.3	92.4
SCQ total score	Mean	8.61	6.8
	(SD)	3.35	2.44
	Range	1-17	2-13
SCQ score (ASD above cut off)	% score above 15	**84.5**	**71.6**
Wessex Vision Categorised	% normal	93.6	89.9
Wessex Hearing score categorised	% normal	96.1	97.5
TAQ total score	Mean	38.32	33.82
	(SD)	19.55	19.64
	Range	1-72	0-71
MIPQ total score	Mean	38.23	37.85
	(SD)	5.20	5.52
	Range	25.5-48	20-48
RBQ total score	Mean	28.30	28.22
	(SD)	16.45	17.74
	Range	0-74	0-70

2.5.3. Measures

The behavioural data analysed in the present study have been collected by utilising a number of standardised measures that have been developed to assess the psychological and behavioural constructs associated with challenging behaviour. Information on the characteristics of each of the measures employed is provided below.

2.5.3.1. The Wessex Scale

The Wessex scale (Kushlick et al*.,* 1973) was developed to assess the adaptive ability of children and adults with ID. It comprises five subscales, including: continence, mobility, self-help skills, speech and literacy. For the purpose of the present study, the self-help subscale was used as an estimate of the degree of ability, and responses to items on mobility, speech, reading, writing and counting were used to further describe the sample. Reliability for the overall score on the SPI subscale is reported at 65%, and reliability for the overall score on the SSL scales is reported at 76%, indicating good inter-rater reliability at subscale level for both children and adults (Palmer & Jenkins, 1982).

2.5.3.2. The Mood, Interest and Pleasure Questionnaire

The Mood, Interest and Pleasure Questionnaire – Short form (MIPQ-S, Ross & Oliver, 2003) was included to assess affect in individuals with severe and profound ID and contains two subscales; the ‘mood’ subscale and the ‘interest and pleasure’ subscale, based on the two main constructs of depression listed in the DSM-IV. Twelve items form two subscales: Mood, Interest and Pleasure. The measure has good internal consistency (Cronbach’s alpha coefficients: total = .88, Mood = .79, Interest and Pleasure = .87), test-retest (.97) and inter-rater reliability (.85; Ross & Oliver, 2003).

2.5.3.3. The Activity Questionnaire

The Activity Questionnaire (TAQ, Burbidge et al*.,* 2010) was included to assess behaviours indicative of over-activity and impulsivity. The measure has eighteen items which form three subscales of over-activity, impulsivity and impulsive speech. Item level inter-rater reliability ranges from .31 to .75 (mean .56) and test-retest reliability ranges from .60 to .90 (mean .75). Inter-rater and test-retest reliability indices for subscales and total score exceed .70.

2.5.3.4. The Social Communication Questionnaire

The Social Communication Questionnaire (Rutter et al., 2003) is a 40 item questionnaire designed to assess communication skills and social functioning. Each item on the questionnaire describes a specific social, communicative or repetitive behaviour and requires a Yes/No response to indicate the presence or absence of each behaviour. A score of 22 or more constitutes the cut-off score for possible autism, with a score of 15 or more indicating possible ASD. In terms of convergent validity, highly significant correlations have been observed between the Autism Screening Questionnaire and the Autism Diagnostic Interview (ADI: Le Courteur et al*.,* 1989). Sensitivity and specificity of .85 and .85, respectively, are reported for the cut-off for possible ASD, and values of .75 and .60 are reported for the cut-off for autism. Finally, the correlation between individual item scores and the total score ranges from .26 to .73, with the majority exceeding .50 (Berument et al*.,* 1999).

2.5.3.5. The Repetitive Behaviour Questionnaire

The Repetitive Behaviour Questionnaire (RBQ, Moss, Oliver, Arron, Burbidge & Berg, 2009) comprises five subscales: stereotyped behaviour, compulsive behaviour, insistence on sameness, restricted preferences and repetitive speech. Previous examination of the psychometric properties of the RBQ (Moss et al*.*, 2009) reveals that it has good inter-rater reliability coefficients (range .46 - .80) and test-retest reliability (range .61 - .93; Moss et al*.*, 2009).

2.5.3.6. The Challenging Behaviour Questionnaire

The Challenging Behaviour Questionnaire (CBQ, Hyman et al*.*, 2002) has been developed to evaluate the presence of self-injury, physical aggression, destruction of property and stereotyped behaviour in the last month. The measure also examines eight topographies of self-injury that were adapted from Bodfish et al. (1995). For the present study items evaluating self-injury and aggression were used. Previous examination of the psychometric properties of the questionnaire has demonstrated good inter-rater reliability with reliability coefficients ranging from .61 to .89 (Hyman et al*.*, 2002).

2.6. Data analysis

Behavioural data from 79 participants have been analysed for the purposes of the present study. Data were tested for normality using Kolmogorov–Smirnov tests. Where data did not meet criteria for a normal distribution ($p < .05$) non-parametric tests were employed.

To investigate the persistence of self-injurious behaviour and aggression in individuals with FXS, descriptive analyses of self-injurious and aggressive behaviour were conducted at three different time points: 2003, 2006 and 2011. Participants were divided into three groups: those who show persistent self-injurious behaviour ($n = 30$, 38%) in 2003, 2006 and 2011, those who show transient self-injurious behaviour, including those who show either remission or incidence of self-injurious behaviour during the eight years ($n = 18$, 22.8%), and those who never showed self-injurious behaviour from 2003 to 2011 ($n = 31$, 39.2%). Similarly, participants were divided into three categories, based on the presence of aggressive behaviour: those who show persistent aggressive behaviour ($n = 22$, 27.8%), those with an absence of aggressive behaviour ($n = 40$, 50.6%), and those with remission of incidence of aggression ($N = 17$, 21.5%). In order to investigate the putative differences between the defined groups in terms of the behavioural correlates of self-injurious and aggressive behaviour, such as over-activity, impulsivity, repetitive restricted and stereotyped behaviours, a series of Kruskal- Wallis tests was conducted.

Binary logistic regressions were then undertaken to investigate whether particular correlates of challenging behaviour predicted the presence and the persistence of self-injurious behaviour and/or aggression in individuals with FXS. Binary logistic regression is used to predict a categorical (usually dichotomous) variable from a set of predictor variables

(Mertler & Vannatta, 2002). Similar analysis was conducted by Eden et al, (2014) to investigate the risk markers of self-injurious behaviour in individuals with Tuberous Sclerosis Complex.

In addition, Kruskal Wallis tests were conducted to investigate the differences between those individuals who never display self-injurious behaviour, those who displayed persistent self-injurious behaviour between 2003 and 2011 and those who displayed transient self-injurious behaviour, across the three time points. In addition, the Kruskal Wallis test was conducted to investigate the differences between those who display persistent aggressive behaviour, those who show transient aggression and those who never show aggressive behaviour. Finally, ANOVA Tamhane T2 post-hoc one way tests were performed to further assess the differences between these groups.

2.7. Results

2.7.1. Prevalence and persistence of self-injurious behaviour in FXS.

The first aim of the present study was to investigate the persistence of self-injurious behaviour and aggression in individuals with FXS. Based on the descriptive analyses, 49.4% of individuals with FXS displayed self-injurious behaviour in 2003. In 2011, 49.4% displayed self-injurious behaviour, suggesting that the prevalence of self-injurious behaviour remained stable over eight years.

The persistence, remission, incidence and absence of self-injurious and aggressive behaviour over the 8year period are illustrated in Table 2.2. The findings show that 36.3% of individuals who engaged in SIB in 2003 continued to self-injure in 2011. The respective percentage for aggression is 20.3%. In addition, the 32.4% of individuals with FXS showed either remission or incidence of self-injurious behaviour. The respective percentage for aggressive behaviour was found to be 35.4%.

Table 2.2 Number of participants and percentages (in brackets) in absence, persistence, remission and incidence groups between the baseline study in 2003 and the last follow up study in 2011.

Challenging Behaviour	**No Behaviour at either stage**	**Persistence**	**Remission**	**Incidence**
Aggression (%)	35(44.3)	16(20.3)	15(19.0)	13(16.4)
Self-Injury (%)	25 (31.3)	29(36.3)	16(20.0)	9(12.4)

With regard to the prevalence and remission of self-injurious behaviour, such behaviour across different age groups has been investigated. Participants were divided into three groups; under 11 years old, 12 to 18 years old and over 19 years old in 2003. The descriptive findings show that 39.5% (15) of the participants aged under 11 years old displayed self-injurious behaviour. Moreover, the analysis showed that 2.7% (1) of the children aged between 12 and 18 years old were found to show a decline in self-injurious behaviour and 15.8% (5) of individuals aged over 19 years old showed a decline in self-injurious behaviour. However, 12.4% (9) showed incidence of self-injurious behaviour.

In addition, the prevalence and the decline of aggressive behaviour across different age groups were examined. The descriptive findings show that 55.6 % (13) of children aged up to 11 years old displayed aggressive behaviour, 12.1 % (5) of individuals aged between 12 and 18 years showed a decline in aggressive behaviour, and 31.5 % (9) of individuals aged 19 years old and over showed a decline in aggressive behaviour. However, 16.4 (13) of the participants showed incidence of aggressive behaviour.

2.7.2. Behavioural correlates of the persistence of SIB in individuals with FXS

A second aim of the study was to investigate the behavioural correlates of the persistence of SIB and aggression in those individuals with FXS who show persistent, transient and absent SIB and/or aggression between 2003 and 2011. Kruskal-Wallis tests were conducted to investigate the differences between those individuals who never displayed self-injurious behaviour, those who displayed persistent self-injurious behaviour between 2003 and 2011 and those who displayed transient self-injurious behaviour across the three time points (see Table 2.3). First, the differences between individuals who showed either persistent, transient or absent self-injurious behaviour were investigated (see Table 2.4) with post hoc tests performed to further assess the differences between these groups.

Table 2.3 Demographic and behavioural data of the persistent, transient and absent SIB group of children with FXS

		Persistent	Transient group	Absent group	Kruskal Wallis	df	*p* value	*Post hoc analyses (ANOVA/Tamhane T2)*
N		27	27	25				
Age	Median *(IQ Range)*	13.00 *(9.00 19.00)*	17.00 *(11.00-22.00)*	16.00 *(11.00 – 22.50)*	-	-	-	
TAQ Impulsivity	Median *(IQ Range)*	21.00 *(14.00-24.00)*	17.00 *(10.00-23.00)*	14.00 *(5.00 - 20.00)*	5.794	2	.055	
TAQ Overactivity	Median *(IQ Range)*	23.00 *(8.00 - 30.00)*	18.00 *(9.00-26.00)*	16.00 *(4.00 - 24.00)*	3.768	2	.152	
SCQ Communication	Median *(IQ Range)*	7.00 *(6.00 -8.00)*	7.00 *(5.00 -8.00)*	7.00 *(5.00 -8.00)*	.703	2	.704	
SCQ Social Interaction	Median *(IQ Range)*	9.00 *(7.25-11.68)*	9.00 *(5.68-10.50)*	7.00 *(6.00-11.25)*	2.910	2	.233	
SCQ RRSB	Median *(IQ Range)*	5.00 *(4.00 - 6.00)*	5.00 *(3.00-6.00)*	4.00 *(1.00 -5.00)*	8.396	2	**.015**	**Persistent>Absent**

The results in Table 2.3 show that children who never displayed self-injurious behaviour, children who showed persistent self-injurious behaviour and children who showed transient self-injurious behaviour differed significantly in terms of restrictive, repetitive and stereotyped behaviours (Kruskal-Wallis χ^2 (2) = .015).

Similarly, to investigate the behavioural correlates of aggressive behaviour among individuals with FXS, a Kruskal-Wallis test was performed (see Table 2.4). Participants were divided into the persistent aggressive behaviour group (n= 22, 27.8%), the transient group (n= 17, 21.6%) and the absent group (n= 40, 50.6%). The results of this analysis suggest that children who show either persistent, absent or transient aggressive behaviour differ significantly in impulsivity and over-activity scores [Kruskal-Wallis χ^2 (2) = p <.001] and in over-activity [Kruskal-Wallis χ^2 (2) =.018].

Table. 2.4 Demographic and Behavioural data of the persistent, transient and absent aggression group of children with FXS

		Persistent	Transient	Absent group	Kruskal-Wallis	Df	*p* value	***Post hoc analyses (ANOVA/Tamhane T2)***
N		22	17	40				
Age	Median *(IQ Range)*	10.00 *(8.00 13.00)*	14.00 *(11.00-22.00)*	17.00 *(12.00 -23.75)*	-	-	-	
SCQ RRSB	Median *(IQ Range)*	4.50 *(3.00 - 6.75)*	5.00 *(4.00-6.00)*	4.00 *(2.00 -5.75)*	4.370	2	.112	
TAQ Impulsivity	Median *(IQ Range)*	22.50 *(17.25- 24.00)*	20.00 *(14.00-23.00)*	14.00 *(4.00 -19.00)*	**18.012**	**2**	**<.001**	Absent<Transient< Persistent
TAQ Over-activity	Median *(IQ Range)*	24.50 *(13.50 -32.00)*	19.00 *(11.00-28.00)*	11.50 *(4.00 -22.75)*	**8.083**	**2**	**.018**	Absent<Persistent
SCQ Communication	Median *(IQ Range)*	7.00 *(4.00 -7.50)*	7.00 *(6.00 -8.25)*	7.00 *(5.60 -8.00)*	1.315	2	.518	
SCQ Social Interaction	**Median** ***(IQ Range)***	**7.50** ***(6.00-9.43)***	**9.50** ***(6.75-11.00)***	**8.00** ***(6.00-11.00)***	**1.183**	**2**	**.554**	

However, it was essential to explore further these significant differences by employing post hoc tests. A post hoc test ANOVA one-way, with Tamhane T2 post-hoc test, was performed to further assess the significant differences which were revealed from the Kruskal- Wallis tests, and to confirm these significant differences between the absent, persistent and transient group of children, who showed either self-injurious behaviour or aggression. There was a statistically significant difference between groups, as determined by one-way ANOVA (*F* (2, 78) = 5.701, *p* = .005). A Tamhane T2 post-hoc test revealed that there were significant differences in repetitive, restricted and stereotyped behaviours between individuals who did not show self-injurious behaviour in 2003, 2006 and 2011 and those who showed persistent self-injurious behaviour across the three time points (p=.009).

Post hoc tests were performed to identify differences in impulsivity and over-activity between the persistent, transient and absent aggression group. There was a statistically significant difference between groups as determined by one-way ANOVA (*F* (2, 78) = 12.354, *p* <.001). A Tamhane T2 post-hoc test revealed that significant differences in impulsivity between the absent and persistent aggressive behaviour group were evident (p <.001) and significant differences in impulsivity between the absent and transient group (p <.001) were also evident. Additionally, a one-way ANOVA revealed that there were significant differences between the groups in over-activity [*F* (2, 78) = 4.187, *p* = .019]. A Tamhane T2 post-hoc test revealed that significant differences in over-activity between the absent and persistent groups with aggressive behaviour were evident (*p* = .030). Figures 2.1 and 2.2 illustrate the differences between the absent, persistent and transient group of children with self-injurious behaviour or aggression, for the behavioural characteristics significantly associated with challenging behaviour.

Figure 2.1 Mean, Standard Deviation and Standard Error of TAQ and SCQ subscale for absent, transient and persistent self-injury groups of children with FXS.

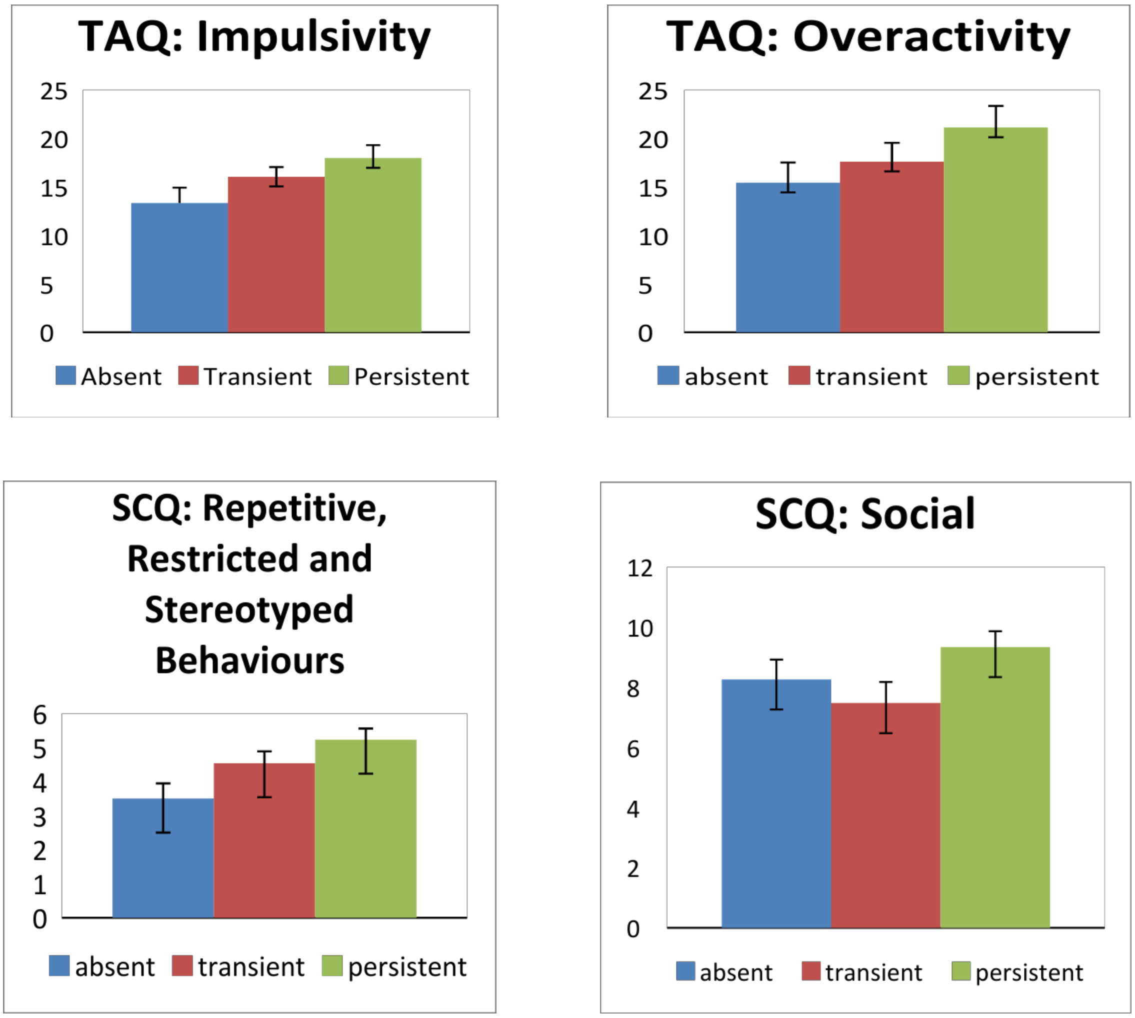
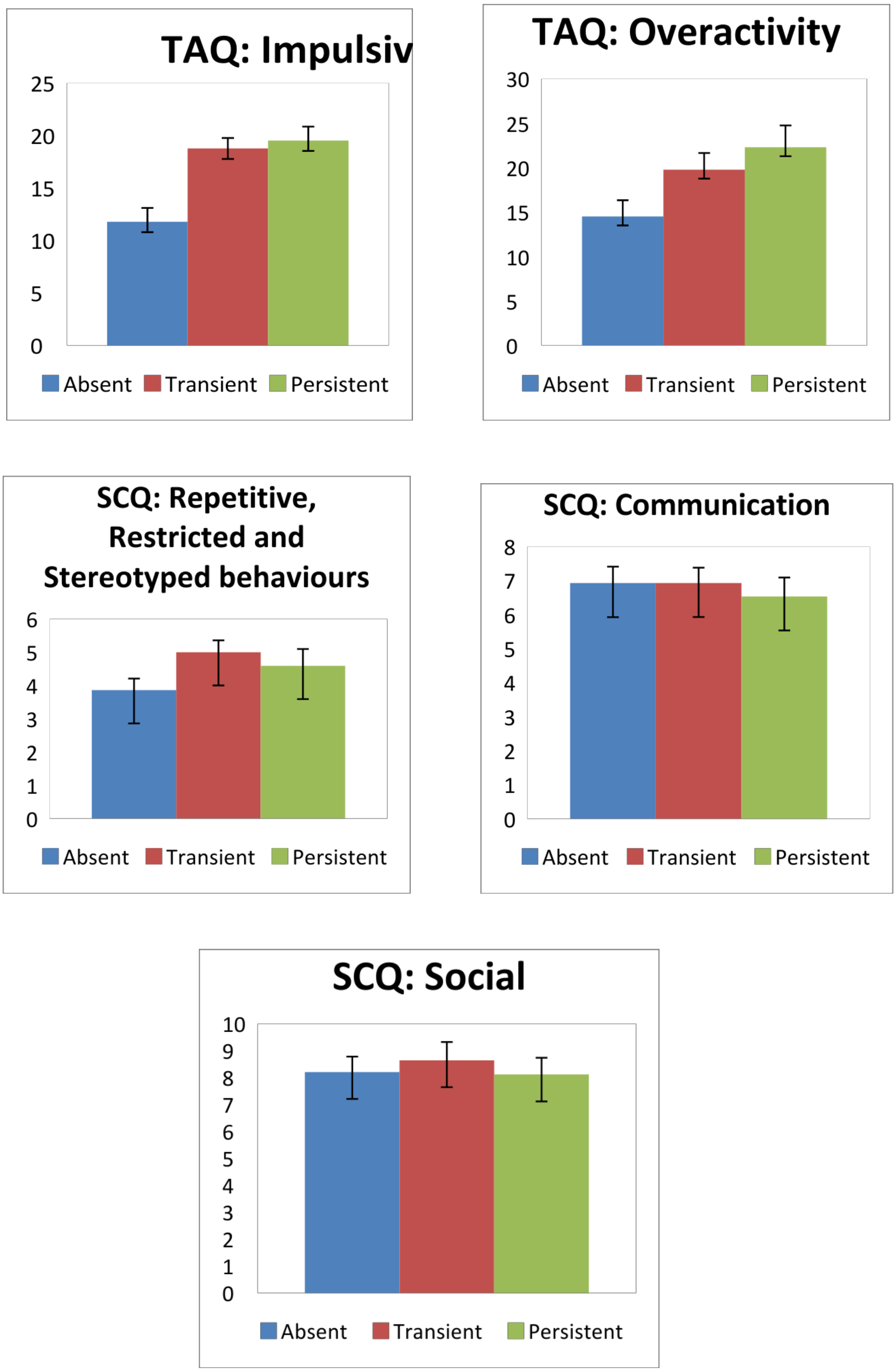


Figure 2.2 Mean, Standard Deviation and Standard Error of TAQ and SCQ subscale for absent, transient and persistent aggression groups of children with FXS.


TAQ: Impulsiv

TAQ: Overactivity

SCQ: Repetitive, Restricted and Stereotyped behaviours

SCQ: Communication

SCQ: Social

The third aim of the study was to evaluate which behavioural correlates of aggressive behaviour predict the persistence of aggression in individuals with FXS. Binary Logistic Regression was performed to evaluate whether the behavioural characteristics which have been previously identified may be involved in the presence of aggressive behaviour (see Table 2.5).

Table 2.5 Binary Logistic Regression predicting the likelihood of the persistence of aggressive behaviour

	B	**S.E**	**Wald**	***Df***	***p***	**Odds Ratio**	**95.0% C.I. for Odds Ratio**	
							Lower	**Upper**
TAQ Over-activity	-.005	.041	.013	1	.909	.995	.918	1.08
TAQ Impulsivity	.159	.070	5.140	1	**.023**	**1.17**	**1.02**	**1.34**

As shown in Table 2.5 the Impulsivity subscale of the Total Activity Questionnaire was found to be a significant predictor of the presence of aggressive behaviour, between individuals who showed aggressive behaviour at the three time points, from 2003 to 2011, and those who did not, again from 2003 to 2011. The full model containing TAQ Impulsivity as a predictor was statistically significant, [χ^2 (5) =13.242, p =.021], indicating that the model was able to distinguish between participants who are displaying persistent aggressive behaviour and those who are not. The model as a whole explained between 17 % (Cox and Snell R square) and 22.9% (Nagelkerke R squared) of the variance in aggressive behaviour, and correctly classified 66.2 % of cases. The odds ratio was 1.18, suggesting that individuals with high scores on TAQ Impulsivity are 1.18 times more likely to show aggressive behaviours.

2.8. Discussion

This longitudinal study aimed to investigate the persistence of self-injurious behaviour and aggression in individuals with FXS in an 8 year period. Moreover, the behavioural correlates which might predict the presence and the persistence of self-injury and aggression were investigated. The results extend the previous literature by examining both the prevalence and the persistent nature of self-injury and aggression in individuals with FXS. The study adds to the literature in terms of both the novelty analysis of longitudinal data about challenging behaviour in individuals with FXS, and the separate analysis that investigated the predictors of the presence of self-injurious behaviour and aggression in this syndrome group. Most notably, it was found that individuals who show either persistent or absent self-injurious and/or aggressive behaviour differ significantly between particular behavioural correlates of challenging behaviour, such as impulsivity, over-activity, repetitive restricted and stereotyped behaviours, communication and social interaction; and indications emerged as to which of those behavioural correlates might predict the persistence of self-injurious and aggressive behaviour in individuals with FXS.

2.8.1. The presence and persistence of self-injurious and aggressive behaviour in individuals with FXS

Descriptive statistics revealed that 49.4% of individuals with FXS displayed self-injurious behaviour. In addition, the 36.3% of the participants who showed self-injurious behaviour in 2003 continued to show self-injurious behaviour eight years later in 2011. Similarly, 12.4% of participants reported incidence of self-injurious behaviour at any of the three time points. However, 20% of individuals showed a decline in self-injurious behaviour. Additionally, 20.3 % of the participants were found to display aggressive behaviour eight years after the initial baseline study which was conducted in 2003. In addition, 16.4% of the participants were associated with incidence of aggressive behaviour at any of the three time points. However, 19.0% of the participants showed remission of aggressive behaviour. Most notably, it was found that 50.6% of individuals with FXS exhibited self-injurious behaviour in 2003. Additionally, 50% of them continued to display self-injurious behaviour 8 years later in 2011. This prevalence rate is similar to previous reports (Arron et al. 2011; Richards et al. 2012) for individuals with FXS. The demographic characteristics of participants with FXS who took part in Richards' et al, (2012) study are similar to the demographic characteristics of the participants in this current study, because overlapping samples were explored longitudinally. Similarly, participants in this current study were similar in age and abilities to the participants with FXS who took part in Arron's et al*,* (2011) study. In the current study it was found that 50.6% of participants with FXS displayed self-injurious behaviour and 40.5% showed aggressive behaviour. This finding is consistent with the findings of both Richards et al, (2012) and Arron et al, (2011). In addition, in this study it was found that individuals with FXS have autistic traits or dual autism diagnosis, as the majority of individuals scored high on the SCQ scale. These findings are consistent with a previous report (Moss et al*.,* 2013).

The prevalence of self-injurious behaviour and/or aggression among individuals with FXS, according to their age, is also described in the present study. The prevalence of self-injurious and aggressive behaviour was greater in childhood and adolescence compared to adulthood. However, this finding is derived from descriptive statistics and further statistical analyses have been employed to analyse the relationship between the prevalence of both self-injurious and aggressive behaviour. The relationship between age and the prevalence of both self-injurious and aggressive behaviour among individuals with intellectual disability (ID) was explored earlier in a study conducted by Baghdadli et al*,* (2008). Most notably, it was found that the prevalence of self-injurious behaviour declines in individuals with ID.

Based on the current study, investigating the persistent nature of self-injurious and aggressive behaviour in individuals with FXS, it was found that 36.3% and 20.3% of individuals show persistent self-injurious and aggressive behaviour respectively. The persistent nature of both self-injurious and aggressive behaviour among individuals with ID has been previously examined by Emerson et al.*,* (2001b), where it was demonstrated that over a seven year period 71% of individuals continued to display self-injurious behaviour. Similarly, Taylor, Oliver and Murphy (2011) demonstrated that self-injurious behaviour was persistent in 84% of people with ID over a 20 year period. However, participants who took part in Emerson et al.'s (2001b) follow- up study were young adults with ID. Although the findings of the longitudinal study in this thesis highlight the persistent nature of self-injury and aggression, no direct comparisons are possible because of the different samples employed in (Emerson et al., 2001b; Taylor, Oliver & Murphy, 2011).

With regard to the persistence of aggressive behaviour there is evidence of the persistent nature of aggressive behaviour among individuals with ID. Totsika, Toogood, Hastings and Lewis, (2008) found that 70% of their participants with ID continued to exhibit aggressive behaviour at the 11 years follow up. However, the persistence of both self-injurious and aggressive behaviour has been described in the literature among individuals with ID, rather than the persistence of challenging behaviour being explored among individuals within a single syndrome group. To this end, the present study found that although there is a small decline in self-injurious and aggressive behaviour over the years, as children get older and the maturation effects of cognitive and emotional development occur, behavioural problems tend to remain persistent. The persistent nature of self-injurious behaviour and aggression underlines the need to develop early interventions that target this group of individuals who are at higher risk for the development of self-injurious and aggressive behaviour.

2.8.2. Behavioural correlates of the persistence of self-injurious and aggressive behaviour

The second aim of the current study was to identify whether the behavioural characteristics which have been found to be related to self-injurious behaviour and aggression in individuals with ID may also be related to the persistent nature of such behaviours. Most notably, the study found that the group of individuals who show persistent self-injurious behaviour differ significantly from the group of individuals without self-injury, in relation to repetitive, restricted and stereotyped behaviours. This finding suggests that individuals with persistent self-injury tend to score higher in repetitive, restricted and stereotyped behaviours. Moreover, individuals with persistent aggressive behaviour differ significantly between the absent and transient group of individuals, in relation to impulsivity scores, suggesting that individuals with persistent aggression show higher impulsivity scores. In addition, a significant difference between persistent and absent aggressive behaviour groups in respect of over-activity scores indicates that individuals with persistent aggression show higher over-activity scores, compared to individuals without aggressive behaviour. Interestingly, Richards et al, (2012) found that individuals with FXS who engaged in self-injurious behaviour scored significantly higher on all SCQ subscales, compared to those who did not engage in self-injurious behaviour. Similarly, in the present study, individuals with FXS who showed persistent self-injury scored significantly higher in repetitive, restricted and stereotyped behaviours, compared to those who showed an absence of self-injurious behaviour, indicating that the persistence of self-injurious behaviour is associated with the presence of repetitive, restricted and stereotyped behaviours. Similarly, Arron et al, (2011) found that individuals with FXS at a younger age were more likely to show aggressive behaviour compared to those of older ages. In this study, the percentage of people who displayed self-injury and/or aggression differed in terms of age in this syndrome group. However, it is essential to investigate further the frequency of self-injurious behaviour and aggression among individuals with FXS in different age groups, rather than simply employing statistical tests. Such investigations were beyond the scope of the present study.

In Arron's et al., (2011) study it was also found that individuals with FXS, Cornelia de Lange, Prader–Willi and Lowe syndrome with self-injury displayed higher scores on measures of repetitive behaviour, over-activity and impulsivity. In addition, individuals with the above genetic syndromes who displayed aggressive behaviour scored higher in impulsivity and over-activity. These findings are along the same lines as the findings of the current study, where individuals with FXS who show persistent aggression scored significantly higher on impulsivity and over-activity, compared to those where aggressive behaviour was absent during the eight year period. However, Arron et al., (2011) aimed to describe the prevalence and the phenomenology of challenging behaviour in genetic syndromes rather than to describe the persistence of both self-injurious and aggressive behaviour among individuals with various genetic syndromes. In summary, these findings suggest an association in individuals with FXS between particular behavioural correlates of challenging behaviour, such as impulsivity, over-activity, repetitive, restricted, and stereotyped behaviours, and the persistence of challenging behaviour. However, although these findings suggest an association between behavioural correlates of challenging behaviour and the persistent nature of challenging behaviour, further investigation is needed on the extent to which these behavioural correlates can predict the persistence of challenging behaviour.

Further, the predictors of the persistent nature of aggressive behaviour were investigated in the present study. It was found that repetitive, restricted and stereotyped behaviours were strongly associated with the presence and the persistent nature of self-injurious behaviour; and impulsivity predicted the presence and the persistence of aggression in individuals with FXS. These results are in line with Baghdadli's et al*,* (2008) findings. However, Baghdadli et al., (2008) found that lower verbal ability and the severity of autism symptomatology were linked with the persistence of self-injurious behaviour. Such relationships were not examined in the current study.

In addition, in this present longitudinal study, repetitive, restricted and stereotyped behaviours were found to be significantly related to the persistence of self-injurious behaviour. This finding is consistent with Davies' & Oliver's (2016) study where it was found that individuals with self-injurious behaviour scored higher in measures regarding repetitive and restricted behaviours, and individuals with aggression scored higher in over-activity and impulsivity measures, suggesting that repetitive, restricted and stereotyped behaviours can predict self-injury in individuals with ID ; and over-activity and impulsivity were revealed as plausible predictors for aggressive behaviour among individuals with ID.

Impulsivity was revealed in the present study to be significantly related to and also to predict the presence of and the persistence of aggressive behaviour. This finding is consistent with Arron's et al., (2011) findings. Most notably, it has been found in the current study that individuals with high scores in relation to over-activity and impulsivity are more likely to display physical aggression. Similar findings, which suggest these two behavioural correlates of challenging behaviour as putative predictors for the presence of self-injurious and aggressive behaviour, have been described in several studies (Oliver & Richards, 2010; Moss et al., 2009; Hyman, Oliver & Hall, 2002; Symons, Clark, Hatton, Skinner & Bailey, 2003). The novelty of this longitudinal study is that it investigated the relationship between the persistence of self-injurious and aggressive behaviour in individuals, and impulsivity and repetitive, restricted and stereotyped behaviours.

In summary, through this current longitudinal investigation it has been found that there are particular behavioural correlates of challenging behaviour, such as impulsivity, over-activity, repetitive, and restricted and stereotyped behaviours, which are associated in individuals with FXS with the persistence of challenging behaviour. Investigating the persistence of challenging behaviour in individuals with FXS is critical because of the high prevalence of challenging behaviour in this syndrome group, and the high prevalence of ASD-types of behaviours amongst individuals with FXS. Moreover, this longitudinal investigation is of value for service providers and practitioners who might need to identify people who score high in these ASD-types of behaviours and then provide early interventions in order to reduce or eliminate the presence of challenging behaviour.

2.9. Strengths and limitations of this study

The present study aimed to describe the persistence of self-injurious and aggressive behaviour in individuals with FXS over a period of 8 years. The validity and reliability of the initial measures, which were employed in the first study in 2003, strengthens the validity and reliability of the results. Moreover, conducting a longitudinal investigation allowed the delineation of the differences in particular behavioural correlates of challenging behaviour between those individuals with FXS who show either persistent, absent or transient self-injurious and aggressive behaviour. This was through fine-grained design and robust statistical methods conducted to investigate longitudinally the persistence of challenging behaviour in individuals with FXS. Finally, the study replicated previous findings that highlighted the existence of predictors for the presence of self-injurious behaviour and aggression.

A limitation of the present study is that it can be considered that the sample size might limit the generalisability of the findings that relate to the persistence of self-injurious and aggressive behaviour. However, the sample size employed in this longitudinal investigation was equivalent to that of another study (Richards et al., 2012). Nevertheless, to further investigate the persistence of self-injurious behaviour in individuals with FXS, a larger sample size is needed. Moreover, it is essential that the persistence of self-injurious behaviour into adulthood is investigated by conducting follow-up studies in the same group of individuals in their later adult life. Although associations between behavioural correlates of challenging behaviour and the persistence of challenging behaviour have been uncovered, utilisation of such knowledge in clinical practice might be required to confirm such associations, and then gauge their putative significance in informing early intervention and practice for the most vulnerable.

The presence and the persistence of challenging behaviour in FXS has been investigated longitudinally by analysing existing data which have been collected and analysed previously within the Centre for Neurodevelopmental Disorders at the University of Birmingham. Although standardised measures have been used at both time points, and the demographic characteristics of the sample were measured respectively at both time points, there are some putative limitations. The limitations of analysing existing datasets have been described in Section 1.10 in Chapter 1 of this thesis. However, to overcome those limitations, it was ensured that the research questions of the previous studies conducted were in line with the current study's aims. In addition, the data collected and analysed were of interest only in relation to the FXS syndrome group because of the absence of a comparison syndrome group. However, due to the limited research investigating the presence and the persistence of challenging behaviour focusing on this particular syndrome group, this longitudinal investigation focusing on a single syndrome group was essential for the prevalence and persistence of challenging behaviour to be investigated. Another major limitation of the analysis of existing data is that the researchers analysing the data are not usually the same individuals as those involved in the data collection process. Therefore, they are probably unaware of study-specific nuances or glitches in the data collection process that may be important to the interpretation of specific variables in the dataset.

CHAPTER 3

Examining the relationship between challenging behaviour, parental emotional wellbeing and quality of life (QoL) in children with ID

3.1. Preface

The study conducted in Chapter 2 explored the persistent nature of self-injurious behaviour and aggression in individuals with FXS, along with the characteristics that might be associated with the persistence of self-injury in individuals with FXS. The relationship between behavioural correlates of challenging behaviour and challenging behaviour (see Section 1.5.2), the association between behavioural correlates of challenging behaviour and the persistence of self-injurious and aggressive behaviour among individuals with FXS (see Section 2.7.2) and the relationship between challenging behaviour and parental emotional wellbeing (see Section 1.8.1) have already been reviewed in this thesis. However, this chapter aims to investigate for the first time the relationship between challenging behaviour itself, behavioural correlates of challenging behaviour, parental emotional distress and the quality of life (QoL) of children with ID and/or challenging behaviour. Using a standardised online survey methodology, the present chapter will extend the findings from Chapter 2. It aims to describe the characteristics that are associated with challenging behaviour in individuals with a range of genetic syndromes, who are at high risk for challenging behaviour, to investigate whether challenging behaviour itself or behavioural correlates of challenging behaviour might predict parental emotional distress and the child's wellbeing; and also it aims to examine the associations between challenging behaviour, children’s QoL and carer wellbeing.

3.2. Introduction

The relationship between challenging behaviour and parental emotional distress has been previously highlighted in the literature (see Section 1.8.1). Most notably, it has been previously reported that parents of children with challenging behaviour experience greater parental stress and/or depression compared to parents of children without challenging behaviour. Interestingly, there is evidence showing that challenging behaviour is associated with parental emotional distress and compromised QoL for children manifesting the behaviour (Hastings, 2002). However, by exploring the association between behavioural correlates of challenging behaviour and challenging behaviour itself (see Sections 1.5.2 and 2.7.2) the proposition may be tested that parental emotional distress might not only be associated with challenging behaviour per se, but might also be associated with other behaviours that might be challenging for carers. In the following sections the relationship between parental emotional distress, challenging behaviour itself and behavioural correlates of challenging behaviour will be investigated.

3.2.1. Challenging behaviour and parental wellbeing

Evidence has accumulated suggesting that challenging behaviour might be related to poorer parental well-being (see Section 1.6). In a study conducted in Australia by Davis and Gavidia Payne (2009) it was found that challenging behaviour and child's ID status were correlated with parental emotional wellbeing. However, in the same study, parental emotional wellbeing was found to be associated with the type and amount of professional support received. Most notably, it was found that family income, professional support and the child's behavioural problem predicted family QoL. In summary, in the same study, parental emotional distress was found to be related to other factors, such as the child's self-help ability, family income and professional support, alongside dealing with challenging behaviour. In addition, professional support was found to be related to the extent of behavioural problems. Therefore, it is essential that the association between parental emotional distress and particular forms of challenging behaviour should be investigated.

In another study it was found that self-efficacy can act as a mediator of the relationship between the child's behaviour problems and the mother's anxiety and depression, but not the father's anxiety and depression (Hastings & Brown, 2002). Most notably, it was found that fathers with high self-efficacy scores were less anxious than were those with low self-efficacy scores when their child exhibited high rates of behavioural problems. In addition, it was found that mothers of children with ASD were at higher risk of developing mental health problems. Although in this study the relationship between the child's behavioural problems and parental emotional distress was investigated, the findings are not representative of families of children with ID heterogeneous aetiology, as only parents of children with autism took part in the study. In addition, there are likely to be variables other than the child's behaviour problems that affect parents' feelings of efficacy in dealing with difficult behaviours. Further research is needed, where an independent assessment of parental mental health problems is carried out, and a longitudinal analysis is essential on the causal role of self-efficacy in this population.

Moreover, recruiting a large size sample of parents of children with ID, rather than focusing only on ASD, is essential for the relationship between challenging behaviour and parental emotional distress to be further evaluated. There is evidence to show that parents of children with ID experience significantly higher stress compared to parents of children without disabilities (Baker et al., 2003). However, findings suggest that higher stress scores were associated with the child's behavioural problems rather than with their status. The authors of the study suggest the existence of an interrelationship between parental emotional wellbeing and the child's behavioural problems. However, this relationship may change depending upon other factors such as social support and the family's social economic status. Therefore, further investigation is needed to assess the direction and extent of this association.

Furthermore, there is evidence on the relationship between parental emotional distress and particular topographies of challenging behaviour. Drysdale, Jahoda and Campbell (2009), found that SIB is a highly emotive and stressful issue for mothers of children with ID. By employing a qualitative design, they found that mothers' attributions of children's self-injurious behaviour were consistent with dimensions of locus of cause, stability and controllability. In addition, in the same study, maternal control was found to be associated with maternal stress and a sense of responsibility when there was inadequate support. Overall, it may be assumed that the extent of particular forms of challenging behaviour will be related in a linear way with greater parental stress or depression. However, it is not clear whether professional support might be related to both maternal stress and self-injurious behaviour. Although, the qualitative description of the relationship between self-injury and maternal emotional wellbeing has been investigated, further research is required to examine the separate contribution to parental psychological distress of specific forms of children's challenging behaviour, such as aggression, self-injury and related behaviours. This type of investigation would be required to include the contribution of the frequency and type of self-injurious behaviour to maternal emotional distress. In summary, it is not yet clear whether parental emotional wellbeing is directly associated with challenging behaviour itself, or is related with other child behaviours, such as impulsivity, overactivity, repetitive, restricted and stereotyped behaviours, that link to the presence of challenging behaviour (see Section 1.5.3).

Furthermore, it has been found elsewhere that parental emotional distress is also associated with a range of other behaviours related to challenging behaviour itself. More specifically, it has been previously observed that mothers of children with autism reported the greatest stress rates when their children were more irritable, lethargic/socially withdrawn, hyperactive/non-compliant, unable to take care of themselves, and unable to communicate or interact with others (Tomanik, Harris, & Hawkins, 2004). In addition, in the same study, it was found that children who scored lower in self-care domains were more likely to present behavioural problems, and parents of those children reported greater emotional distress. Therefore, parental emotional distress might be associated with a range of other behaviours, which are also common among children with ID and challenging behaviour. In the following sections additional factors will be reviewed that might be associated with parental emotional distress, independently of the presence of challenging behaviour.

3.2.2. The child's pain and parental emotional distress

There is evidence to show that in the general population parental emotional wellbeing is associated with children's health problems (Casey et al. 2004). However, the relationship between child health problems and parental emotional distress among individuals with ID has not been clearly described. Most notably, Mitchell and Hauser-Cram (2008) highlighted a relationship between child health problems and maternal stress in the mothers of children receiving special education services. However, no significant associations with mothers' depression rates were evident, and also the investigation did not account for anxiety rates. Further investigation of the potential relationship between parent anxiety and depression and child health problems is essential. In addition to this evidence, there is a line of research on the relationship between parental emotional wellbeing and the presence of an ASD diagnosis on the child. The following section expands on this area of inquiry.

3.2.3. ASD and parental emotional wellbeing

There is a line of research that focuses on understanding the difference in emotional distress between parents of children with an ID, with and without an additional diagnosis of ASD. Generally, studies have reported higher levels of stress (Griffith et al., 2010) and depression (Bailey et al., 2007) in parents of children with an additional ASD diagnosis. However, there is limited evidence on the relationship between anxiety and ASD. Totsika et al., (2011) reported higher emotional distress scores for mothers of children with ASD. In addition, Griffith et al, (2010) found that mothers of children with autism were experiencing higher stress levels compared to mothers of children with Down's syndrome or mothers of children with ID heterogeneous aetiology. Moreover, the same study found that mothers of children with autism scored significantly lower on positive perceptions of their child, compared to mothers of children with other diagnostic criteria. In line with Totsika et al, (2011), Estes et al, (2009) found higher maternal distress scores in mothers of pre-school children with ASD; however, the presence of challenging behaviour might count as a co-variable in this relationship.

It is well known that parents of children with ID are at higher risk of developing mental health issues. However, despite the evidence for the impact of challenging behaviour on parental emotional wellbeing, few studies have looked at the influence of other related behaviours on parental emotional wellbeing. Most notably, the presence of challenging behaviour itself might lead to parental stress (Hastings, 2002). Moreover, other behavioural manifestations, such as impulsivity, over-activity, repetitive, restricted and stereotyped behaviours, have been found to be significantly correlated with challenging behaviour (Oliver et al. 2012; Oliver et al., 2009; Richards et al., 2012; see also Section 2.7.2). In the following section the association between behavioural correlates of challenging behaviour and parental emotional distress will be reviewed.

3.2.4. Behavioural correlates of challenging behaviour and parental emotional distress

There are several individual and environmental characteristics that appear to predate the development of more severe challenging behaviour and might therefore be considered to be risk markers. These include a greater degree of intellectual disability, the presence of social impairment or specific genetic syndromes, associations between social contact and behaviour that are consistent with operant theories of the maintenance of behaviour, and the presence of repetitive behaviour (Arron, Oliver, Berg, Moss & Burbidge, 2011; Baghdadli, Pascal, Grisi, & Aussilloux,2003; McClintock, Hall, & Oliver, 2003; Murphy et al., 2005; Murphy, Healy & Leader, 2009; Saloviita, 2000). There is very little research directly investigating the relationship between repetitive, restricted and stereotyped behaviours, and emotional wellbeing in parents of children with ID. Moreover, based on previous literature, it is well known that behaviours such as impulsivity, over-activity and health problems, might predict the presence of challenging behaviour among people with ID (Arron et al., 2011). This study used the Challenging Behaviour Questionnaire (CBQ, Hyman, Oliver and Hall, 2002), which is a brief informant-based questionnaire evaluating the presence or absence of self-injury, physical aggression, verbal aggression, destruction of property and inappropriate vocalisations over the last month. Items regarding self-injury and physical aggression were used in this study. Separate standardised measures about impulsivity, overactivity and mood were utilised in order to delineate the association between the presence of challenging behaviour and behavioural risk markers of challenging behaviour.

It has been suggested in this section that impulsivity and overactivity can be identified as behavioural correlates of challenging behaviour. This study used the Activity Questionnaire (TAQ, Burbidge & Oliver, 2008) which is an information based questionnaire designed to evaluate overactivity and impulsivity. The questionnaire comprises 18 items grouped into three subscales: overactivity, impulsivity and impulsive speech.

Most notably, there is evidence to show that repetitive, restricted, stereotyped behaviours, over-activity and impulsivity are strongly associated with the presence of challenging behaviour and parental stress (Oliver et al., 2012; Gupta et al., 2007).). Repetitive, restricted and stereotyped behaviours have been identified as behavioural correlates of challenging behaviour. Such behaviours can be measured through the Repetitive Behaviour Questionnaire (RBQ, Moss, Oliver, Arron, Burbidge & Berg, 2009). This measure comprises five subscales: Stereotyped Behaviour, Compulsive Behaviour, Insistence on Sameness, Restricted Preferences and Repetitive Speech.

More specifically, a small body of literature has reported a significant relationship between children’s restricted and/or repetitive behaviour and parental stress in parents of children with ASD (Beckman, 2002; Gabriels et al., 2005; Tomanik, Harris & Hawkins, 2004). However, stereotyped behaviours were found not to be related to parenting stress. Overall it is currently uncertain whether these behaviours can predict parental stress alongside challenging behaviour. Most notably, impulsivity, over-activity, repetitive, restricted and stereotyped behaviours, which have been found to be associated with the presence of challenging behaviour, (see Section 1.5.2) and the persistence of self-injurious and aggressive behaviour among individuals with FXS, (see Section 2.7.2) are also related to parental emotional distress. Hence evidence for such an association in parents of young children with ID is extremely limited, and further investigation is needed. Therefore the present study aims to investigate whether parental stress is related to challenging behaviour itself or to other behavioural manifestations.

In a review study, conducted by Oliver and Richards (2010), it was proposed that repetitive behaviour and impulsivity are indicators of behavioural self-regulation. Most notably, high scores on repetitive behaviour and impulsivity were found to be associated with the presence and severity of self-injurious behaviour amongst individuals with ID. Moreover, these behaviours were described as behaviours which are initiated without inhibition, and continue when an intervention for those behaviours is not taking place. Cross-sectional research has established a relationship between parental emotional distress and the presence of over-activity and/or impulsivity. Gupta (2007) found that parents of children with ADHD and developmental disabilities reported higher total parenting stress levels than parents of typically developing children. Interestingly, child-related stress was highest among parents of children with ADHD. Similarly, Lecavalier et al, (2006), found that challenging behaviour is related to parenting stress; but following a multiple regression analysis this relationship was not significant when other child behaviours were added, which illustrated that child hyperactivity is significantly related to parental stress. In addition, Tomanik et al, (2004), found a significant relationship between hyperactivity and parental stress across the group of children with Pervasive Developmental Disorder. To date, although a possible relationship between behavioural correlates of challenging behaviour and parental emotional wellbeing has been previously suggested, further investigation is needed to delineate whether parental emotional distress can be predicted from those behaviours alongside challenging behaviour.

In summary, there is evidence that impulsivity and over-activity are related to the presence of challenging behaviour in children with ID. Additionally, there are some well-structured studies confirming the relationship between parental emotional distress and particular child behaviours, other than challenging behaviour. However, it is currently unclear whether parental stress may be associated with particular correlates of challenging behaviour, such as impulsivity and overactivity, or whether challenging behaviour itself influences parental emotional distress.

In summary, the present study aims to address whether particular forms of challenging behaviour, such as self-injurious behaviour, and aggression or other related behaviours, are associated with parental emotional distress; and also to investigate whether challenging behaviour itself, and whether behavioural correlates of challenging behaviour, such as impulsivity, over-activity, repetitive, restricted and stereotyped behaviours, and child pain problems, are also related to parental emotional distress in relation to children with ID.

3.3. Measuring parental emotional distress

The emotional wellbeing of parents of children with ID and/or behavioural problems has been investigated in the literature by adopting either a qualitative or quantitative approach. In terms of quantitative approaches there are several measures that have been developed to assess parental stress and depression. For instance, the General Health Questionnaire (GHQ, Goldberg & Williams, 1988) measure can provide a generic psychological health assessment, and also can reliably discriminate between positive and negative mental health states (Green et al. 2005; Hu, Stewart-Brown, Twigg & Weich, 2007; see also Totsika et al., 2011). Moreover, the Questionnaire on Resources and Stress (QRS, Konstantareas, Homatidis & Plowright, 1992; see also Estes et al., 2009) has been developed to measure parental stress among parents of children with ID, and parents can rate their agreement or disagreement with questions that tap parental feelings about their child. Although both of these measures have adequate psychometric properties they cannot provide an accurate indicator of whether parents are experiencing either higher stress levels or higher depressive symptoms. It would be essential to use a measure which can evaluate both anxiety and depression symptoms in order to be employed for parents of children with ID.

Taking into account the need to develop a generic measure of parental emotional distress, Griffith et al, (2010) employed the Hospital Anxiety and Depression Scales (HADS) (Zigmond & Snaith, 1983). This measure has been reported to have excellent psychometric properties (Hastings et al., 2005) and has been widely used with parents of children with ID. This questionnaire-based measure provides a good indicator of the anxiety or depressive symptoms that parents are experiencing. The identification of the specific child behaviours associated with parental anxiety and depression symptoms is important in developing an effective community early intervention pathway, and in identifying those parents who may require the most support. By using such a measure a prevalence rate of parental anxiety and depression among the sample of children with ID can be calculated, and the relationship between parental stress and/or depression and challenging behaviour or other child behaviours can be further delineated. In the following section a further justification for the use of HADS as a measure of parental anxiety and/or depression will be provided.

3.3.1. Measuring parental emotional distress using HADS

The Hospital Anxiety and Depression Scale (HADS, Zigmond & Snaith, 1983) has been used in a number of studies to measure parental anxiety and depression symptoms (Hastings & Brown, 2002; Gallagher et al., 2008). HADS includes seven anxiety and seven depression items and has been reported to have excellent psychometric properties (i.e., Hastings et al., 2005). There are many studies that have employed HADS in studies of children with autism (Hastings & Brown, 2002; Griffith et al., 2010), or children with particular genetic disorders (Gallagher et al., 2008). Moreover, there are more than 200 published studies that have utilised HADS, mainly conducted in medical settings worldwide, highlighting the wider use of this measure (Herrmann, 1997). Subjects who have been studied over time include psychiatric, oncological, cardiological, neurological, and fibromyalgia patients, as well as other patients with various chronic health problems, in groups of people with general health complaints, but also with healthy people as control groups (Hastings et al., 2005).

The present section proposes that there is an interrelationship between challenging behaviour itself, behavioural correlates of challenging behaviour, and parental emotional distress. Moreover, although challenging behaviour might have a critical effect on parental emotional distress, it is also likely that children with challenging behaviour also experience diminished QoL. The following section expands this area of inquiry.

3.4. QoL of children with severe ID

QoL has emerged as an important concept in research into the wellbeing of people with ID. However, to date, there is little known about the QoL of children with ID, especially those with more severe ID.

There is a significant proportion of research describing the QoL of children with disabilities, along with the potential factors that may influence children's wellbeing. More specifically, evidence exists on the QoL of children with physical disabilities and/or chronic health conditions, reporting that child characteristics such as age, gender, severity of disease, and degree of pain can be associated with the QoL of children with cerebral palsy (Arnaud et al., 2008; Varni et al., 2005). Based on these studies, pain and the severity of disease have been found to be negatively correlated with the QoL of children. In addition, similar findings have been reported when studying children with cerebral palsy (Houlihan et al., 2004) and children with epilepsy (Williams et al., 2003). Most notably, findings from the latter study suggested that the families who are most vulnerable to a reduced QoL are those whose child has severe epilepsy problems, co-morbid disabilities and increased parental emotional distress. Although evidence exists on the relationship between physical disabilities and QoL in children, it is essential for the relationship between ID and QoL in children to be further investigated.

There is a line of research on the QoL of children with an ASD diagnosis. Most notably, Lee et al, (2008), found that parents of children with autism reported more frequently that their children had poorer QoL (i.e., emotional and social wellbeing) compared to children with ADHD. Similarly, Kuhlthau et al, (2010) found that children with ASD were experiencing poorer health wellbeing compared to typically developing children. However, in the same study it was found that, irrespective of the health domain, children with ASD were reported to experience poorer psychosocial health, social and emotional functioning, compared to both typically developing children and children with chronic health conditions. Unsurprisingly, in this study it was also found that the occurrence of repetitive behaviour, impulsive, and over-active behaviours were negatively associated with psychosocial, social and emotional wellbeing, highlighting the association between ASD type of behaviours and the QoL of children with ASD.

However, although evidence exists on the associations between particular behaviours associated with ASD and health related QoL in children with ASD, the possible causality of the relationship between these constructs is currently unknown. In addition, rather than focusing on children with particular physical disabilities or ASD, it is essential for the QoL of children with ID to be investigated. Therefore, a comprehensive study of the QoL of children with ID, with and without challenging behaviour, would be required. Moreover, considering the limited number of studies on the QoL of children with ID, it would be essential for a review of the existing measures of QoL to be conducted. The following section starts with a review in this area.

3.4.1. Measuring QoL in children with ID

There is a concern in the field that there is an insufficient number of QoL measures that have been developed for children with ID and challenging behaviour. In an attempt to delineate this area of inquiry, King et al., (2005), reviewed 33 QoL measures. Originally, 107 generic and disease-specific measures were identified, but 74 measures were later excluded as either focused primarily on health status, developed for adults, or with outdated concepts. Interestingly, it was found that 4 of the 33 reviewed measures of QoL could be administered to children with chronic health conditions and/ or ID, and only these four measures had proxy measures available (see Table 3. 1.).

Table 3.1 Quality of life measures

Measure	Psychometric Properties	Overview	Pros	Cons
Caregiver Priority and Child Health Index of Life with Disabilities (CP-CHILD)(*Narayanan, Weir and Fehlings, 2013*)	Overall reliability was excellent, with total score of 0.85. CPCHILD is a valid measure of the caregiver's perception of the HRQL of children with severe CP	The CPCHILD Questionnaire measures the caregiver's perspective about the child's health status, comfort, wellbeing, functional abilities and ease of care giving. It is a useful proxy measure of health related QoL of children with severe disabilities.	It is valid and reliable It is administered to children with cerebral palsy It can be completed by both children and parents in cases that parents need to complete on behalf of their children	It is administered to children whose disability affects mobility
Health-related QoL (HRQL) (Ronen, Streiner, Rosenbaum, 2003)	The evaluation with Cronbach's coefficient demonstrated acceptable internal consistencies of 0.7. There is good evidence of construct and discriminative validity.	Health-related QoL (HRQL) is an essential component of outcomes in clinical medicine because it reflects the patients' perceptions of their health and well-being and their functioning	A major aspect of HRQL measures is the power to discriminate.	It focuses only on children with epilepsy
The Paediatric Cancer QoL Inventory-32(PCQL-32) (Varni et al.1999)	This measure has good psychometric properties.	The Paediatric Cancer QoL Inventory-32(PCQL-32) has been developed to be a standardised assessment instrument to assess systematically paediatric cancer patient's health-related QoL (HRQOL) outcomes.	There is a proxy measure available The overall utility is good	This measure is primarily to assess the QoL of children with chronic health issues and not disability
Kiddy-KINDL (Ravens-Sieberer and Bullinger, 2000. KINDL^R Questionnaire for Measuring Health-Related QoL in Children and Adolescents Revised Version Manual)	Cronbach´s alpha as a measure of internal consistency reached values of around a =.70.	A screening tool for child's QoL.	There is a proxy measure available The overall utility is excellent	This measure has been administered to children with health problems but is a generic QoL measure, and there are 20 additional items which can be used for children with ID

Although there are validated and reliable measures available that have been developed to investigate the QoL of children with ID, they are mainly intended for children with chronic health issues or physical disabilities without ID. There is only one standardised measure available, the Kiddy-KINDL, that has additional items for children with ID. The original Kiddy-KINDL parent version consists of 24 Likert-scaled items associated with six dimensions: physical well-being, emotional well-being, self-esteem, family, friends and everyday functioning (school or nursery school/kindergarten). The parent version includes 22 additional items which can be treated as a sub-scale in their own right or can be added to the main categories.

As part of the present study, a modified Kiddy-KINDL version was used, tailored to individuals with ID. This modified Kiddy-KINDL parent version consisted of 36 Likert-scaled items, associated with four dimensions: physical wellbeing, emotional wellbeing, family wellbeing and social wellbeing. The self-esteem dimension was excluded, as it was not relevant to the study population. In addition, the sections that related to friends and everyday functioning were excluded, and a new dimension named social wellbeing was inserted. The additional 22 items were added in different categories according to the nature of the question (see Appendix E).

It is apparent that it is crucial for the association to be investigated between behavioural correlates of challenging behaviour, such as impulsivity, over-activity, repetitive, restricted and stereotyped behaviours, and parental emotional wellbeing. In addition, the relationship between behavioural correlates of challenging behaviour and the QoL of children with ID is also important for investigation. Differences between children with and without challenging behaviour, in terms of QoL, are also important to be delineated. Finally, the interrelationships between child QoL, parental emotional wellbeing and challenging behaviour need to be identified for the first time. In the following section, the association between behavioural correlates of challenging behaviour and the QoL of children with ID is reviewed.

3.4.2. Behavioural correlates of challenging behaviour and the QoL of children

The present study aims to investigate the behavioural correlates of parental emotional distress and children's QoL, alongside challenging behaviour. The association has already been described between particular behavioural correlates of challenging behaviour and the presence and the persistence of challenging behaviour (see Sections 1.5.2; 2.7.2). In addition, evidence has been highlighted showing that challenging behaviour can be associated with child QoL (see Section 1.8.2). Considering the relationship between behavioural correlates of challenging behaviour and parental emotional distress, the present investigation aims to delineate whether the behavioural correlates of challenging behaviour may also be associated with child QoL. The importance of this investigation lies in the fact that for the first time this relationship will be investigated, so as to provide an insight into the QoL of children with ID and challenging behaviour, and to inform service providers on the putative need to deliver early intervention programmes to help the most vulnerable.

3.5. The present study

In the present study the associations between the correlates of challenging behaviour, challenging behaviour itself, parental emotional wellbeing and child QoL will be evaluated. More specifically, by analysing data from Angelman Syndrome, Smith-Magenis Syndrome, Prader-Willi Syndrome, Cri du Chat, Cornelia de Lange, Sotos, 1p36, 8p23 deletion syndrome groups, three key areas will be investigated:

1 The relationship between particular behavioural correlates of challenging behaviour, such as over-activity/impulsivity, repetitive, restricted and stereotyped behaviours, and the presence of health problems and aggressive or self-injurious behaviour will be described.

2 The association between behavioural correlates of challenging behaviour and parental emotional distress will also be investigated. Consistent with previous research, a strong interrelationship is presented between challenging behaviour and parental emotional distress (Hastings, 2002). Moreover, the present study aims to investigate the relationship between particular behavioural correlates of challenging behaviour and parental emotional distress.

3. An additional purpose of the present study is to investigate the relationship between particular behavioural correlates of challenging behaviour, such as impulsivity, over-activity, repetitive, restricted and stereotyped behaviour, and the child's QoL. The challenging behaviour itself and the child's QoL, and also the association between parental emotional distress and the child's QoL are investigated.

3.6. Aims

1 This study aims to investigate the QoL of children with ID and/or challenging behaviour

2 This study aims to investigate the relationship between QoL, challenging behaviour and parental emotional wellbeing.

3.7. Hypotheses

1 It is hypothesised that impulsivity/over-activity, health problems, restricted, and repetitive and stereotyped behaviours are associated with challenging behaviour

2 It is hypothesised that challenging behaviour is associated with parental emotional wellbeing. Most notably, it is hypothesised that the extent of challenging behaviour is associated with greater parental anxiety and/or depression.

3.8. Methods

3.8.1. Recruitment

Invitation letters and emails were sent to 387 parents of children with Angelman Syndrome, Smith-Magenis Syndrome, Prader-Willi Syndrome, Cri du Chat, Cornelia de Lange, Sotos, 1p36, 8p23 deletion, 9q34 deletion (also known as Kleefstra syndrome), Lowe and Phelan-McDermid syndrome. Participants were recruited through the participant database available at the Cerebra Centre for Neurodevelopmental Disorders at the University of Birmingham, School of Psychology. Inclusion criteria included the age of the person they cared for being less than 16 years and having a confirmed diagnosis of a syndrome made by a General Practitioner, Clinical Geneticist or Paediatrician. If a large proportion of the data (i.e., 25% or more of items across questionnaires) was incomplete, the participant was excluded from further analysis. Questionnaires were returned by 120 parents (31%) (AS: n = 34, 28.3%), Cornelia de Lange (CdLS: n = 10, 8.3%), Cri du Chat (CdC: n = 13, 10.8%), Lowe (LS: n = 3, 2.5%), Prader-Willi (PWS: n = 17, 14.2%), Smith Magenis (SMS: n = 10, 8.3%), Soto Syndrome (Soto: n=6, 5%), 1p36 (1p36: n=14, 11.7%), 9q34 (9q34: n=2, 1.7%), 8p23 (8p23: n=6, 5%) and Phelan McDermid Syndrome (PMS: n=5, 4.2%).

3.8.2. Participants

The sample of the study consisted of 119 children (mean age: 7.95 years; range = 1.00 – 15.00). 51.3% were male (n=61) and 48.7% female (n=58). Demographic variables from the Wessex Behaviour Scale (Kushlick, Blunden & Cox, 1973) indicated that 60.2% were mobile, 65.3% had normal vision, 76.8% had normal hearing, 67% were partially verbal or verbal, and 48% were able or partially able (see Table 3.2). Although the sample consisted of 119 families of children with ID and/or challenging behaviour, the reported sample size for some particular outcome measures was reduced to 108, due to missing data.

As an initial confirmation of the validity of the collected data an assessment of the agreement between the present and previous reports was essential. More specifically, the behavioural data on challenging behaviour that had been collected within the Centre 6 months prior to the present study have been compared with the current challenging behaviour data. The previous set of data included data on challenging behaviour, such as forms, frequency and intensity of challenging behaviour, for each of the individuals who participated in the online survey for the present study; and these data were analysed in this present study. To test the agreement between past parental reports about children's challenging behaviour and current reports Kappa analysis was conducted. Specifically, these were samples and data coming from the last study, which was completed in 2012, six months prior to the present study, by Dr Mary Heald, Dr Leah Bull and Dr Lucy Wild within the Cerebra Centre for Neurodevelopmental Disorders at the University of Birmingham. Researchers in the earlier study had utilised the Challenging Behaviour Questionnaire (CBQ, Hyman, Oliver & Hall, 2002) to assess challenging behaviour in participants who took part both in the initial observations in 2012 and in the current study. In this present study, a question about challenging behaviour was added to the online survey in which parents could report whether their children display challenging behaviour or not.

Table. 3.2. Mean age (standard deviation) and range in years, percentage of males, percentage of participants who were able, mobile and verbal, mean SCQ total score (standard deviation) and range for between-group analyses.

N		119
Age	*Mean*	7.95
	(SD)	(*3.91*)
	Range	1-15
Gender	*% male*	51.3
Self Help	*% partly able/able*	48
Mobility	*% mobile*	60.2
Speech	*% verbal*	67
Hearing	*% normal*	76.8
SCQ cut off scores	*Above 15*	61.5
	Below 15	38.5
TAQ Impulsivity	*Median* *(IQ Range)*	17.50 (.00-24.00)
Over-activity	*Median* *(IQ Range)*	15.00 (.00-36.00)
SCQ Repetitive Behaviour	*Median* *(IQ Range)*	4.00 (.00-8.00)
Health Problems	0-1Health problems	68.1 (80)
	2-3 Health problems	26.9 (33)
	4-5 Health problems	5.0 (6)

Typically, Kappa analysis is a measure of agreement, standardised to lie on a -1 to 1 scale, where 1 is perfect agreement, and 0 is exactly what would be expected by chance, and negative values indicate agreement less than by chance. An almost perfect agreement level has been found, kappa= 1.00 indicating that the majority of individuals who displayed challenging behaviour in the last study, which was completed within the Cerebra Centre for Neurodevelopmental Disorders at the University of Birmingham, continue to display challenging behaviour in this study.

3.8.3. Procedure

Invitation letters offered participation in an online questionnaire study exploring parental/ caregiver views of children's genetic syndromes, their feelings, experiences and support from services. A web link, password and ID number were included so that parents could access the consent forms and questionnaire via ‘Lime survey’. Participants were notified that they could request a paper copy, and ten participants opted to return a paper copy. Four to six weeks after invitation letters were sent, a reminder procedure was followed, by asking parents who were contacted via telephone to enquire whether they had questions or difficulties accessing the survey.

At the beginning of the survey carers were asked to indicate whether their children had exhibited challenging behaviour in the last month. Participants completed a series of standardised questionnaires on their child’s behavioural profile, based on the last 6 months. Data about challenging behaviour, impulsivity, over-activity, repetitive behaviour, ASD, ability level and health problems had been collected within the Cerebra Centre for Neurodevelopmental Disorders six months prior to this study, and therefore were already available for analysis. In addition, in the present study, standardised measures about parental emotional distress and children's quality of life were utilised. Participants completed the Hospital Anxiety and Depression Scale first (Zigmond & Snaith, 1983); following this measure they were asked to indicate their children's QoL for children with and without challenging behaviour, and the degree of service provision for families of children who displayed challenging behaviour. Participants completed the measures in the order described above. Ethical approval for the study was obtained from the Ethical Review Committee at the University of Birmingham (see Appendix A). The approximate time for the completion of the survey was 45 minutes.

3.8.4. Measures

3.8.4.1. Measures used prior to this study

The TAQ, SCQ, RBQ, Health Questionnaire and CBQ were administered to the participants in the previous cross-syndrome study six month*s* prior to the present study (Appendix J; Appendix K; Appendix L; Appendix M; Appendix O and Appendix P).

The Activity Questionnaire

The Activity Questionnaire has been described in Section 2.5.3.3.

Social Communication Questionnaire (SCQ)

The Social Communication Questionnaire has been described in Section 2.5.3.4.

The Repetitive Behaviour Questionnaire (RBQ)

The Repetitive Behaviour Questionnaire (RBQ, Moss, Oliver, Arron, Burbidge & Berg, 2009) is an informant-based questionnaire for use with children and adults with a range of intellectual abilities. It is applicable for use with verbal and non-verbal individuals and for individuals with ID and\or autism spectrum disorder. The Repetitive Behaviour Questionnaire consists of nineteen items that comprise five subscales: stereotyped behaviour, compulsive behaviour, insistence on sameness, restricted preferences and repetitive speech. All items are based on operationally defined features of behaviours. Informants rate the frequency of behaviour over the preceding month. The response format consists of a five-point Likert-type rating scale ranging from ‘never’ to ‘more than once a day’.

The Health Questionnaire (HQ)

The Health Questionnaire (HQ, Hall et al., 2008) was designed for measuring the presence and severity of health problems and to classify these problems. Informants are asked to rate the presence and severity (0 = never occurred, to 3 = severe problem) of problems ever occurring in the person’s life and over the last month. Scores are summed to produce an Overall Health Score, indicating severity of health problems for the previous month and during the person’s life. The total numbers of health problems during the person’s life and the previous month were also calculated.

3.8.4.2. Measures used in the present study

Hospital Anxiety and Depression Scale (HADS)

The questionnaire consists of 14 items and includes two main subscales: for anxiety and depression (Zigmond & Snaith, 1983). Participants' responses are rated on a 4-point Likert Scale, from 'never' to 'always'. HADS has adequate psychometric properties and has been translated into thirteen languages. This instrument aims to identify parental anxiety and depression, with a cut-off point of 8/21 for the anxiety subscale and a cut-off point of 8/21 for the depression subscale (Crawford et al., 2001).

Kiddy-KINDL

Kiddy-KINDL (Ravens & Bullinger, 2000) is designed to assess child wellbeing. Kiddy-KINDL includes proxy measures for the assessment of children's and adolescents' QoL. The original questionnaire consists of 24 Likert-scaled items associated with six dimensions: physical, emotional, self-esteem, family, friends and everyday functioning (school or nursery school). The Kiddy-KINDL parent version includes 22 additional items which can be used as additional subscales, or each of the items can be added into the original six subscales. Kiddy-KINDL includes an additional subscale, the 'disease subscale', which can be used in relation to children with chronic health issues. Each of the responses is rated on a 3-point Likert scale from 'never' to 'very often'. A modified version of Kiddy- KINDL was used in this study, where two of the original subscales, self-esteem and friends, were excluded because these subscales were very unlikely to be applicable for children with severe ID and challenging behaviour (see Appendix E). The modified version of Kiddy-KINDL has not been validated and has been utilised as an exploratory quality of life measure for individuals with intellectual disabilities. Moreover, a threat to internal validity has been generated regarding the instrument because items from the additional sub scale in the parent version have been adapted. However, using the measure in the original form would have threatened ecological validity in that parents would have needed to complete non-relevant questions.

In addition, 20 of the 22 additional items have been adapted in the modified version. These were items which were relevant to the physical wellbeing subscales and were adapted into the physical wellbeing subscale. Similar adaptation procedures were followed for each of the additional items. The modified version includes 36 items which are associated with the four dimensions of wellbeing: physical, emotional, family and social. The social subscale comprises items of the original everyday functioning subscale and additional items on the child's social behaviour. Physical wellbeing comprises 9 items, emotional wellbeing 13 items; family wellbeing includes 7 items and social wellbeing comprises 7 items. In the original measure, each subscale had been standardised and the removal of subscales hasn't affected the psychometric properties of the measure. However, the additional 22 items which were included in the parent-version measure have been standardised as an additional subscale.

As a result, adapting the 20 items from the original and into new generated subscales affected the original psychometric properties of the scale. Total scores of all the dimensions of Kiddy KINDL are transformed to a range of 0 to 100. Given that only 4 QoL measures were tailored to individuals with ID these modifications were essential even though the psychometric properties have been affected. Reviewing the existing four main QoL measures three of them were found to be tailored either to individuals with mobility problems or to individuals with chronic health conditions. Although Kiddy-KINDL has been administered primarily in relation to typical developing children there are 22 items which can be administered in children with ID. The purpose of this modified measure is therefore to investigate QoL among individuals with ID.

Challenging Behaviour Questionnaire (CBQ)

The Challenging Behaviour Questionnaire is a questionnaire (CBQ, Hyman, Oliver & Hall, 2002) designed to assess the presence of self-injurious and aggressive behaviour during the previous month. The two items relating to the presence of self-injurious and aggressive behaviour were used in the present study. The authors report good interrater reliability for items relating to self-injurious behaviour (.95) and aggressive behaviour (.85). Appendix 3.2 provides detailed information on the standardised measures used in the present study.

3.9. Data Analysis

A total CBQ score was calculated, combining self-injury, aggression and property destruction, with the aim of exploring the impact of challenging behaviour on both parental and child wellbeing. Participants' data on self-injury, aggression and property destruction were combined in one category, the total challenging behaviour score. Data were tested for normality using Kolmogorov–Smirnov tests. Where data were not normally distributed ($p<.05$), non-parametric techniques were employed.

The analysis of the data was tailored to the hypotheses and the aims of this study. Firstly, it has been hypothesised that impulsivity and/or over-activity, pain problems, repetitive, restricted and stereotyped behaviours might predict challenging behaviour. Hence a further analysis was conducted for the identification of the children's characteristics in relation to challenging behaviour. A t-test for independent samples was conducted to describe the difference between participants who display challenging behaviour and these who do not. Chi-square tests were employed for categorical variables. Binary logistic regression was undertaken to identify which child behavioural and demographic characteristics might predict challenging behaviour.

The second aim of the study was to investigate the association between the child's behavioural characteristics, challenging behaviour, and parental stress or depression. Pearson correlations were employed in order to describe the relationship between the child's characteristics and parental stress and depression. A t-test for independent samples was conducted to delineate the differences in parenting stress and depression between parents of children with and without challenging behaviour. The second part of the aim was to delineate the predictors of parenting stress and depression. Binary logistic regression was undertaken.

Spearman's Rho correlation was undertaken for the association between children's QoL, parental emotional wellbeing and child's behavioural characteristics. Mann-Whitney U tests were employed to describe the differences in children's QoL and in parental emotional wellbeing. Finally, a 2x2 ANOVA was conducted that examined the effect of parental emotional distress and impulsivity on child QoL. A series of Mann-Whitney U tests was conducted to confirm ANOVA findings because of the non-normally distributed sample in each main condition. Mann-Whitney U tests were conducted to confirm the effects of parental emotional distress and impulsivity on child QoL.

3.10. Results

3.10.1. Behavioural correlates of challenging behaviour

The first aim of the present study was to explore whether impulsivity, over-activity, repetitive, restricted and stereotyped behaviours, as well as pain, are associated with the presence of and can also predict the occurrence of self-injurious behaviour and/or aggression among individuals with genetic syndromes and ID. First of all, descriptive analysis revealed that 76 people (70.4%) exhibited challenging behaviour and 32 people (29.6%) were not showing challenging behaviour. Participants who engaged in challenging behaviour and those who did not did not differ in terms of gender. A chi-square test of independence was performed to examine whether there is an association between gender and challenging behaviour, and showed no significant results [X2(1, n= 108) = .030, p=.864]. However, participants differed significantly in terms of age. Participants without challenging behaviour had a mean age of 7.96 (SD= 3.91) and were significantly younger than participants with challenging behaviour, who had a mean age of 8.66 years, t (84) =2.34, p= 0.022. To control this observed effect, age has been added in binary logistic regression to test whether it can predict challenging behaviour. (see Table 3.4).

Table 3.3. T-test, independent samples for challenging behaviour

Measures		**Participants with CB** (N=76)	**Participants without CB** (N=32)	**t-test**	**P value**
Self Help Total Score[1]	**Mean**	*5.65*	*5.11*	1.32	.191
	(SD)	*(1.71)*	*(1.93)*		
TAQ Impulsivity[2]	**Mean**	*18.11*	*9.48*	**6.016**	**<.001**
	(SD)	*(5.99)*	*(6.25)*		
TAQ Over-activity[4]	**Mean**	*18.28*	*13.23*	**2.186**	**.031**
	(SD)	*(10.57)*	*(8.1)*		

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[1] $_{3\text{-}9}$Total n differs between subscales because of missing data.

[2]

Table 3.3. T-test, independent samples for challenging behaviour (continued)

Measures		Participants with CB	Participants without CB	t-test	P value
		(N=76)	(N=32)		
RBQ Compulsive behaviours[6]	**Mean**	*4.20*	*0.41*	**5.72**	**<.001**
	(SD)	*(5.02)*	*(1.05)*		
RBQ Insistence on sameness [7]	**Mean**	*2.47*	*0.85*	**3.14**	**.003**
	(SD)	*(2.58)*	*(2.01)*		
SCQ Communication subscale [8]	**Mean**	*7.26*	*8.15*	-.927	.357
	(SD)	*(3.68)*	*(3.69)*		
SCQ Reciprocal Social Interaction [9]	**Mean**	*6.48*	*6.00*	.486	.628
	(SD)	*(3.99)*	*(4.01)*		
Health Total Score	**Mean**	*.474*	*.219*	**2.44**	**.017**
	(SD)	*(.642)*	*(.420)*		

As is shown in Table 3.3, impulsivity, over-activity, age, health problems, stereotyped behaviours, compulsive behaviours and insistence on sameness differ significantly between participants with and without challenging behaviour. To further investigate the above observed associations and the possible utilisation of these variables as predictors of the presence of challenging behaviour Binary Logistic Regression was performed (Table 3.4).

Table 3.4. Binary Logistic Regression predicting likelihood of displaying challenging behaviour

	B	**S.E**	**Wald**	***Df***	***p***	**Odds Ratio**	**95.0% C.I. for Odds Ratio**	
							Lower	Upper
Age	-.103	.126	.667	1	.414	.902	.704	1.16
Impulsivity	.290	.110	.6.972	1	**.008**	**1.34**	**1.1**	**1.7**
Over-activity	-.113	.080	1.999	1	.157	.893	.764	1.05
Stereotyped behaviours	.085	.092	.849	1	.357	1.1	.909	1.3
Compulsive behaviours	.400	.236	2.87	1	.091	1.5	.939	2.37
Insistence on sameness	.003	.216	.000	1	.987	1.00	.657	1.53
Health Problems	2.18	1.01	4.673	1	**.031**	**8.86**	**1.23**	**64.1**

The model contained seven independent variables (age, health problems, stereotyped behaviours, and compulsive behaviours, insistence on sameness, impulsivity and over-activity). The full model containing all predictors was statistically significant, ($\chi 2$ (7, N = 79) = 40.380, p < .001), indicating that the model was able to distinguish between participants displaying challenging behaviour and participants not displaying challenging behaviour. The model as a whole explained between 40% (Cox and Snell R square) and 57.1% (Nagelkerke R squared) of the variance in challenging behaviour, and correctly classified 83.5% of cases. As shown in Table 3.4, only two of the independent variables made a unique statistically significant contribution to the model. Impulsivity and health problems were significant predictors of challenging behaviour. Results suggest that individuals with high levels of impulsivity and health problems are 1.34 and 8.8 times more likely to show challenging behaviour, controlling for other factors in the model. The above findings confirm the first hypothesis of the present study, in which it was proposed that there are particular behavioural correlates of challenging behaviour which predict the presence of challenging behaviour. In this case impulsivity and health problems predict the presence of challenging behaviour.

3.10.1. Challenging behaviour and parental emotional wellbeing

The second aim of the study was to delineate the association between the child's behavioural characteristics, challenging behaviour and parental stress or depression. First, an independent samples t-test was conducted to compare parental anxiety or depression in relation to child's gender. There was not a significant difference in the scores for parental stress in male children (M=9.35, SD=4.55) and female children (M=9.98, SD=4.93); t (116) =-.725, p =.470. Similarly, there was no significant difference in the scores for parental depression in either male children (M=6.47, SD=3.62) or female children (M=6.59, SD=4.38); t (116) =-162, p =.872. A Pearson correlation analysis was conducted to examine the relationship between children's behavioural characteristics and parental anxiety and/or depression. High scores on impulsivity and over-activity were significantly positively correlated with parental anxiety and/or depression (see Table 3.5).

Table 3.5. Pearson correlation analysis of parental anxiety/depression and child’s characteristics

Measure Subscale	HADS_Anxiety	HADS_Depression	Emotional Distress
Age	-.15	-.05	-.01
Self Help Total Score	-.02	.00	.13
TAQ Impulsivity	**.35***	**.41***	**.34***
TAQ Over-activity	**.34***	**.41***	**.22***
RBQ Stereotyped behaviours	**.25***	**.31***	.19
RBQ Compulsive behaviours	**.23***	**.24***	.15

Table 3.5. Pearson correlation analysis of parental anxiety/depression and child's characteristics (continued)

	HADS_Anxiety	HADS_Depression	Emotional Distress
Measure Subscale			
SCQ Reciprocal Social Interaction	.10	.18	.03
Health Quality of Life Total Score	.13	.07	.11

*p< 0.05 level

3.10.2. Predictors of parenting anxiety and/or depression

It was hypothesised that challenging behaviour differed significantly between those parents who met the cut off criteria for anxiety, depression and emotional distress, compared to parents of children who did not (see Table 3.6). In addition, Spearman's correlation test was performed to test whether age was also correlated with parental emotional wellbeing. No significant results were revealed from this test.

Table 3.6. Number/percentage of parents who met the cut off criteria for anxiety, depression and emotional distress and whose children either display challenging behaviour or do not. Significant differences are highlighted in bold (p <.05; two tailed).

Subscales		Participants with challenging behaviour	Participants without challenging **behaviour**	Chi-square	*p* value
N		76	31		
Cut off for Anxiety	(n)	58 (76%)	12 (38.7%)	**13.76**	<0.001
Cut off for Depression	(n)	33 (44%)	5 (16.1%)	**7.78**	.005
Emotional Distress	**(n)**	**58 (76%)**	**31 (38.7%)**	13.76	<0.001

It was hypothesised that challenging behaviour is associated with parental anxiety and/or depression. A chi-square test was conducted to test the difference between challenging behaviour, parental stress, depression and emotional distress. It was found that challenging behaviour differed significantly between the parents who met the cut off criteria for anxiety, depression and emotional wellbeing and those who did not (see Table 3.6).

The next part of the association between challenging behaviour and parental emotional wellbeing, as described, was to investigate which child behavioural characteristics and challenging behaviour may predict parental anxiety and/or depression (see Table 3.7). Binary logistic regression was performed to assess the likelihood of particular behavioural characteristics predicting parental emotional distress.

Impulsivity was a significant predictor of parental emotional distress. The model contained three independent variables (impulsivity, health problems and challenging behaviour). The full model containing all predictors was statistically significant, (χ^2 (3, n = 87) = 18.075, $p < .001$), indicating that the model was able to distinguish between parents who met the cut off criteria for anxiety and those who did not. The model as a whole explained between 18.8% (Cox and Snell R square) and 26.1% (Nagelkerke R squared) of the variance in parenting stress, and correctly classified 73.6 % of cases. Summarising the above findings, it was found that although challenging behaviour is associated with parental emotional distress, only impulsivity can significantly predict parental anxiety and/or depression.

3.10.3. Quality of life of children with ID and challenging behaviour

This study aimed to explore the child's quality of life and to investigate which child and parent characteristics are associated with children's wellbeing. Spearman's Rho correlation coefficient was used to examine the relationship between age, health problems, stereotyped behaviours, and compulsive behaviours, insistence on sameness, impulsivity and over-activity, parental anxiety and depression and the child's quality of life dimensions (see Table 3.8).

Table 3.7 Logistic regression predicting likelihood of parental emotional distress

	B	**S.E**	**Wald**	***Df***	***P***	**Odds Ratio**	**95.0% C.I. for Odds Ratio**	
							Lower	Upper
Impulsivity	**.096**	**.042**	**5.339**	**1**	**.021**	**1.1**	**1.02**	**1.2**
Health Problems	.348	.455	.585	1	.444	1.42	.581	3.455
Challenging Behaviour	.924	.623	2.199	1	.138	2.52	.743	8.534

Table 3.8. Spearman's Rho correlation coefficients for associations between children's and parents' characteristics and children's quality of life. Bold text indicates a significant difference

Measures	**Kindl Total**	**Physical Wellbeing**	**Emotional Wellbeing**	**Social Wellbeing**	**Family Wellbeing**
Age	-.04	.11	-.02	-.17	-.06
Self-Help Total score	**-.23***	-.07	**-.28***	-.13	**-.37***
TAQ Impulsivity	**-.37***	.09	**-.45***	**-.29***	**-.36***
TAQ Over-activity	-.16	**.22***	**-.25***	-.16	-.13
RBQ Stereotyped behaviours	-.15	.01	**-.21***	-.16	-.02
RBQ Compulsive behaviours	**-.35***	-.09	**-.36***	**-.27***	**-.30***
RBQ Insistence on sameness	**-.38***	-.11	**-.38***	**-.34***	**-.24***
SCQ Communication	.08	-.05	.15	-.07	**.34***
HADS Anxiety	**-.49***	**-.22***	**-.51***	**-.40***	**-.20***
HADS Depression	**-.44***	-.11	**-.51***	**-.35***	**-.23***
Emotional Distress	**-.43***	-.12	**-.47***	**-.35***	**-.25***
Health Quality of Life Total Score	**-.28***	**-.21***	**-.21***	**-.25***	-.16

*Correlation is significant at the 0.05 level (2-tailed).

Findings in Table 3.8 show that high scores on impulsivity, over-activity, stereotyped, restricted and repetitive behaviours, compulsive behaviours, insistence on sameness, parental emotional distress and health problems were found to be negatively correlated with the child's quality of life. Additionally, to investigate whether gender is associated with the quality of life, the Mann Whitney U test was conducted because data were not normally distributed. More specifically, Mann Whitney U tests were performed to identify any difference in quality of life total score between male and female individuals, U =1378.0, p =.212, r = .12. These groups did not differ significantly in either total quality of life scores or on any other wellbeing dimension. Mann Whitney U tests were also utilised to test the difference between the child's quality of life dimensions and challenging behaviour (see Table 3.9).

Table 3.9 Median and inter-quartile range, KINDL total score, plus the physical, emotional, social and family wellbeing subscales for participants with and without challenging behaviour. Significant differences are highlighted in bold (p <.05; two tailed).

Subscales		**Participants with challenging behaviour**	**Participants without challenging behaviour**	**Mann Whitney U**	***p* value**
n		76	32		
KINDL Total score	Median	94.02	102.17	**577.00**	**.<0.001**
	(IQ Range)	(89.67-100)	(97.83-105.97)		
Physical Wellbeing	Median	119.44	119.44	1176.00	.787
	(IQ Range)	(111.11-125)	(111.11-127.78)		
Emotional Wellbeing	Median	215.28	236.11	**616.00**	**.<0.001**
	(IQ Range)	(200.69-230.56)	(225-243.75)		
Social Wellbeing	Median	62.50	73.61	**652.00**	**.<0.001**
	(IQ Range)	(52.78-69.44)	(63.89-80.56)		
Family Wellbeing	Median	90.28	100	**626.00**	**.<0.001**
	(IQ Range)	(80.56-97.22)	(91.67-105.56)		

Findings presented in Table 3.9 show that emotional, social and family wellbeing subscales differed significantly between children with and without challenging behaviour. There was no difference between children with and without challenging behaviour in terms of physical wellbeing.

3.10.4. Examining the interrelationship between parental emotional distress, challenging behaviour and children's quality of life

This study aimed to examine the relationship between parental emotional wellbeing, challenging behaviour and children's QoL. Findings so far suggest that although parental emotional distress is associated with challenging behaviour, in a regression analysis it was found that impulsivity (a behaviour correlate of challenging behaviour) predicts parental emotional distress. Impulsivity was revealed as a significant predictor of the presence of challenging behaviour. Additionally, impulsivity was revealed to be associated with the quality of life of children with and without challenging behaviour. To examine the relationship between quality of life, parental emotional distress and impulsivity in children with and without challenging behaviour a 2 x 2 ANOVA was undertaken (see Table 3.10). Specifically, a two-way ANOVA was conducted in order to compare the mean differences between child wellbeing dimensions that have been split on two independent variables differences (factors), the impulsivity scores and parental emotional distress scores, with each of the independent variable having two levels.

Table 3.10 ANOVA 2 x 2 measuring the interaction of impulsivity and parental emotional distress and the effects on child emotional wellbeing.

Variable	**Mean Square**	**DF**	**Error Df**	**F**	**P value**
Impulsivity	**2310.832**	**1**	**93**	**5.863**	**.017**
Parental Emotional Distress	**4685.542**	**1**	**93**	**11.887**	**.001**
Impulsivity*emotional distress (Interaction)	.104	1	93	.000	.987

Table 3.11 ANOVA 2 x 2 measuring the interaction of impulsivity and parental emotional distress and the effects on child family wellbeing.

Variable	**Mean Square**	**DF**	**Error Df**	**F**	**P value**
Impulsivity	**2310.832**	**1**	**93**	**5.863**	**.017**
Parental Emotional Distress	**4685.542**	**1**	**93**	**11.887**	**.001**
Impulsivity*emotional distress (Interaction)	.104	1	93	.000	.987

Table 3.12 ANOVA 2 x 2 measuring the effects and the interaction of impulsivity and parental emotional distress on child social wellbeing.

Variable	**Mean Square**	**DF**	**Error Df**	**F**	**P value**
Impulsivity	387.576	1	93	2.626	.109
Parental Emotional Distress	**870.594**	**1**	**93**	**5.900**	**.017**
Impulsivity*emotional distress (Interaction)	13.444	1	93	.91	.763

A 2 x 2 ANOVA was conducted to examine the association between impulsivity, parental anxiety and/or depression and the quality of life of children. Simple main effects analysis showed that children who met the cut off criteria for impulsivity were experiencing poorer emotional wellbeing F (1, 93) =5.863, p = .017, and family wellbeing F (1, 93) =6.554, p =.012 (see Figure 3.1). Additionally, main effects analysis showed that children whose parents met the anxiety and depression cut off criteria were experiencing poorer emotional wellbeing F (1, 93) =11.887, p = .001 and social wellbeing F (1, 93) =5.900, p =.017. Figure 3.1 shows the association of impulsivity with emotional and family wellbeing. It shows the association between parental emotional distress and emotional and social wellbeing. However, these figures show the main effects and no interaction effects are revealed in this particular analysis.

ANOVA was conducted to investigate the association between impulsivity, parental emotional wellbeing and children's quality of life. However, as the groups with and without impulsivity, and with and without parental emotional distress were not normally distributed, it was crucial for the Mann-Whitney U test to be performed in order for the significant main effects from ANOVA to be confirmed (see Tables 3.10 to 3.12.). The Mann-Whitney U test revealed that there was a significant difference between the group with impulsivity and emotional distress and the group without impulsivity and emotional distress: U=513.50, p=.001 in relation to children's emotional wellbeing, U=704.00, p=.012 in relation to children's social wellbeing, and U=620.50, p=.001 in relation to children's family wellbeing. Therefore, the Mann-Whitney U test confirmed the ANOVA's significant main effects about impulsivity, parental emotional distress and children's quality of life dimensions. Summarising the ANOVA findings suggests that impulsivity, parental anxiety, and/or depression are associated with children's emotional, social and family wellbeing.

Figure 3.1. The association of impulsivity and parental emotional distress with child emotional, family and social wellbeing

Figure 3.1.1. Effects of parental emotional distress and impulsivity on emotional wellbeing.

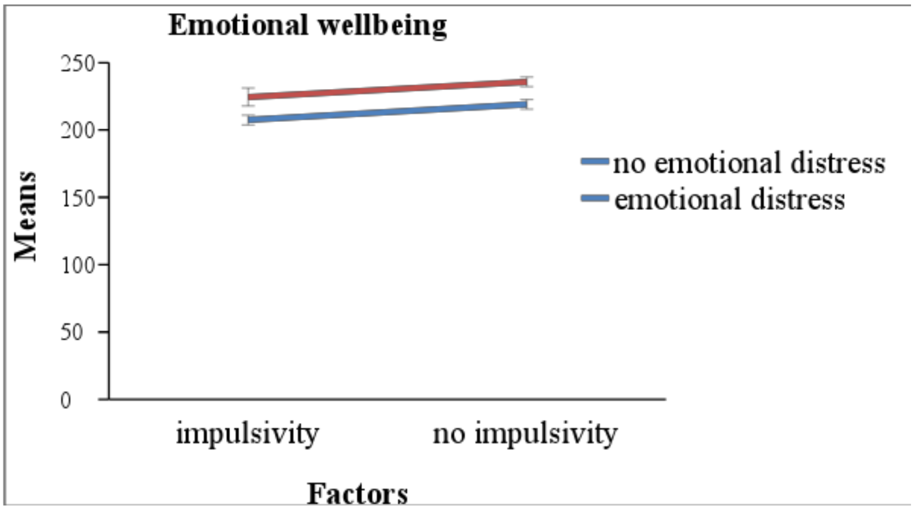


Figure 3.1.2. Effects of parental emotional distress and impulsivity on social wellbeing.

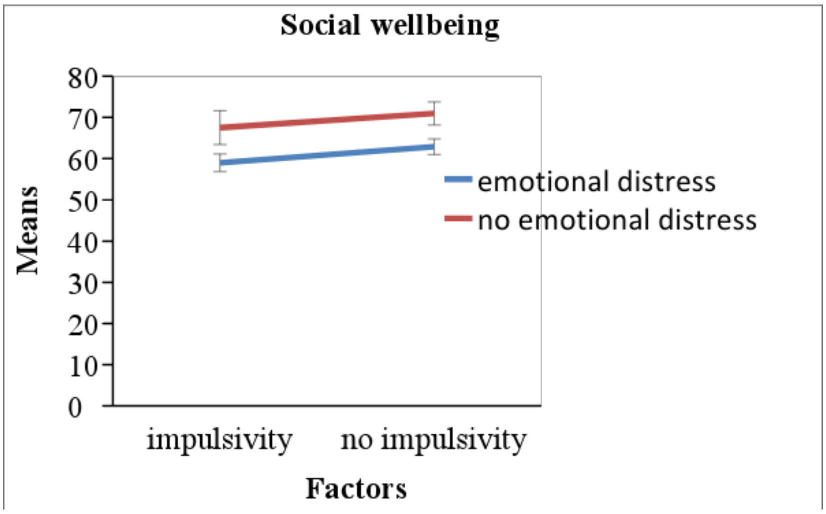
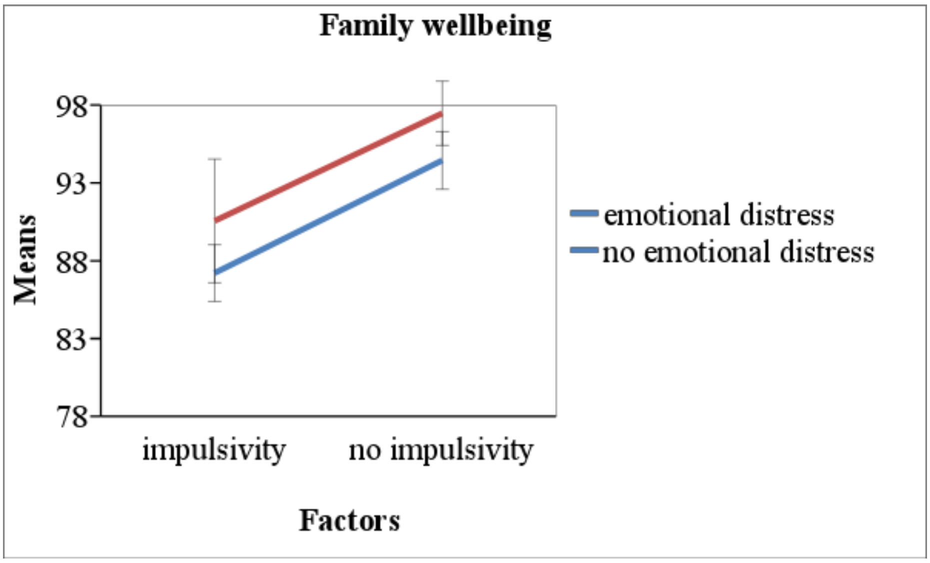


Figure 3.1.3. Effects of parental emotional distress and impulsivity on family wellbeing.



3.11. Discussion

The present study aimed to investigate the quality of life of children with ID and/or challenging behaviour, and also to investigate the relationship between quality of life, challenging behaviour and parental emotional wellbeing. Firstly, it was hypothesised that the presence of challenging behaviour can be predicted by impulsivity/over-activity, health problems, and restricted, repetitive and stereotyped behaviours. Secondly, it was hypothesised that the extent of challenging behaviour will be associated with greater parental anxiety and/or depression. In summary, the study reported that impulsivity, over-activity, repetitive, restricted and stereotyped behaviours and child health problems are associated with the presence of challenging behaviour. Most notably, participants who show challenging behaviour differ significantly in the above domains compared to individuals without challenging behaviour. However, regression analysis revealed that only impulsivity and health problems can predict the presence of challenging behaviour in individuals with ID.

To examine whether challenging behaviour itself, or behavioural correlates of challenging behaviour are associated with parental anxiety and/or depression, a further analysis was performed. It was found, as was expected, that challenging behaviour is strongly associated with parental emotional distress. In addition, behavioural correlates of challenging behaviour were found also to be significantly associated with parental emotional distress. Therefore a regression analysis was conducted to investigate whether challenging behaviour itself or behavioural correlates of challenging behaviour are associated with parental emotional distress. It was found that only impulsivity can predict parental emotional distress, showing that although challenging behaviour is related to parental emotional distress, impulsivity, a behavioural correlate of challenging behaviour, is strongly related to, and can predict parental emotional wellbeing.

The final aim of the study was to investigate the interrelationship between challenging behaviour, parental emotional distress and children's quality of life. Firstly, it was found that children with challenging behaviour differ significantly in terms of the social, emotional and family wellbeing that they experience, compared to children without challenging behaviour. In addition, high scores on impulsivity, over-activity, stereotyped, restricted and repetitive behaviours, compulsive behaviours, insistence on sameness, parental emotional distress and health problems were found to be negatively correlated with the child's quality of life. To further explore this association a series of ANOVA tests was conducted. It was found that parental emotional distress and the child's impulsivity were associated with the emotional wellbeing that children were experiencing. Parental emotional distress was also associated with diminished social wellbeing where impulsivity was related to diminished family wellbeing.

3.11.1. Predicting the presence of challenging behaviour

The current study investigated which of the behavioural correlates of challenging behaviour might predict the presence of challenging behaviour in a sample of children with ID, using an online survey to obtain a large cohort of families of children with a wide range of ID and/or challenging behaviour. Most notably, individuals with genetic syndromes are at high risk of developing challenging behaviour (see Section 1.5.4). This thesis has previously described the association between challenging behaviour and particular behavioural correlates of challenging behaviour such as impulsivity, over-activity, repetitive, restricted and stereotyped behaviours (see Section 1.5.2), as well as the association between challenging behaviour and age, and the relationship between challenging behaviour and pain (see Section 1.7.2). This study aimed to further assess these documented associations. This study's strength is that it employed a robust measure of child wellbeing, and separated measures about behavioural dysregulation characteristics from self-injury, aggression and property destruction, so that specific associations could be explored.

The study found that high scores on impulsivity and health problems may predict the likelihood of challenging behaviour becoming manifest. There are studies which have found that specific behavioural characteristics may predict challenging behaviour (Oliver and Richards, 2010; Arron et al., 2011; Davies, in press). The findings of the current study are in line with the previous findings which suggest that impulsivity and health problems can predict challenging behaviour (Carr & Owen-Deschryver, 2007; Carr et al., 2003; Luzzani et al., 2003; Oliver et al., 2012; Richards et al., 2012; Collacott et al., 1998; Arron et al., 2011). Interestingly, although impulsivity, over-activity, compulsive and stereotyped behaviours, insistence on sameness and health problems are strongly related with challenging behaviour, only impulsivity and health problems can actually predict the presence of challenging behaviour in this cohort of children with ID. Impulsivity is an ASD-type of behaviour, which is common among individuals with ASD and in individuals who are at risk for self-injury and aggression. Therefore it is assumed that the relationship between impulsivity and challenging behaviour further explains the association between ASD and challenging behaviour, confirming that not ASD diagnosis but ASD-types of behaviours are associated with the presence of challenging behaviour.

3.11.2. Challenging behaviour and parental emotional distress

The second hypothesis suggests that challenging behaviour is associated with parental stress. It is well-described in the literature that parents of children with ID and/or challenging behaviour experience heightened anxiety and depression symptoms, (Hastings & Brown, 2002; White & Hastings, 2004; Hastings, 2002). However, parental emotional distress has been found to be related to other behaviours, except challenging behaviour (Oliver et al. 2012; Oliver, Sloneem, et al., 2009; Richards et al., 2012; Beckman, 1983; Gabriels, Cuccaro, Hill, Ivers & Goldson, 2005). Additionally, ASD was found to be related to parental anxiety and/or depression (Griffith et al., 2010). In the current study, parental emotional distress was found to be related to impulsivity, over-activity, stereotyped, compulsive behaviours and insistence on sameness, when controlling for the challenging behaviour variable. These results are in line with previous findings (Beckman, 1983; Gabriels et al., 2005) which suggest that parental emotional distress is associated with other behaviours, other than with challenging behaviour itself. However, the previously documented association between autism and parental emotional distress was not confirmed in the present study. Interestingly, Adams et al, (2014) found that parents of children with Smith Magenis Syndrome reported significantly higher levels of stress compared to parents of children with Down's syndrome. However, parents of children with Down's syndrome reported significantly lower levels, which may suggest that stress associated with parenting a child with a rare genetic condition varies between syndrome groups. It has also been found that parental stress among children with genetic syndromes was greater compared to children with ASD. These behaviours, such as impulsivity, over-activity, repetitive and stereotyped behaviours, were found to be associated with parental stress and depression. Interestingly, the predictors of parental emotional distress were also evaluated in this study.

Hastings, (2002) found that parents of children with challenging behaviour were experiencing higher anxiety and depression levels than parents of children without challenging behaviour. However, Hastings, (2002) incorporates in one category behavioural and emotional problems as 'challenging behaviour'. This present study described the association between challenging behaviour, including behavioural correlates of challenging behaviour and parental stress, rather focusing on how self-injury, aggression and property destruction are related to parental emotional distress. Taking into account the broader definition of behavioural problems which was adopted in Hastings, (2002) study, the findings of the current study are in line with Hastings, (2002) findings. Parental emotional distress was associated with challenging behaviour and specific child behaviours, such as impulsivity, over-activity, repetitive and restricted behaviours. However, impulsivity was a strong predictor of emotional distress. Most notably, high levels of impulsivity were related to lower parental emotional wellbeing.

3.11.3. Quality of life of children with ID

The first aim of this study was to describe the quality of life of children with challenging behaviour. It is reported in the literature that children with ID are experiencing a poor quality of life (Lee et al. 2007; Kuhlthau et al. 2009). Both studies found that children with ID were experiencing poorer social, emotional and psychosocial wellbeing, compared to children without ID or chronic health problems. Similarly, in this study findings indicated that high impulsivity scores are associated with a lower child quality of life total score, and lower emotional, social and family wellbeing. Higher over-activity scores are associated with a lower level of child emotional wellbeing. There was no difference in the experience of physical wellbeing between participants with and without challenging behaviour. However, greater numbers of health problems were negatively associated with the total quality of life, the physical, the emotional and the social wellbeing domains.

In contrast, stereotyped, restricted and repetitive behaviours on the Repetitive Behaviour Questionnaire were found to be associated with a lower quality of life, and particularly with lower emotional and social wellbeing. It was essential that the association between challenging behaviour and the child's quality of life subscales was investigated. Results indicated that the quality of life differs significantly between participants with and without challenging behaviour. However, there was no significant difference in physical wellbeing between children with and without challenging behaviour. This might be explained because the physical wellbeing domain was mainly focused on the child's health condition and how health problems affect the child's physical wellbeing, in terms of sleeping, being in pain and having a healthy appetite. Although health problems are strongly related to the presence of challenging behaviour, in this study the relationship between challenging behaviour or health problems and physical wellbeing was investigated separately.

3.11.4. Quality of life, parental emotional distress and other behaviours

The last part of the analysis aimed to delineate the association between impulsivity, parental emotional distress and quality of life. High impulsivity scores and diminished parental emotional distress were associated with diminished child emotional wellbeing. Most notably, high impulsivity scores and higher parental emotional distress were associated with a child's poorer level of emotional wellbeing. Additionally, it was found that poorer social wellbeing was associated with greater parental emotional distress. Finally, high impulsivity scores were associated with poorer family wellbeing. Interestingly, no interactions between parental emotional distress, impulsivity and child wellbeing were found. It was found that each of the three wellbeing subscales was associated either with impulsivity or parental emotional distress, or with both variables. Item analysis was conducted to examine these differences further. Interestingly, it was found that each wellbeing subscale included different items which, because of the nature of the questions, were related to either parental emotional distress or impulsivity or to both.

The use of a variety of well-standardised measures with very good psychometric properties strengthens the validity of the results. Additionally, in this study, the individuals who took part had a range of genetic syndromes, which increases the generalisability of the results in a wide range of individuals with ID. However, there are some limitations of this online study. In this survey individuals with rare genetic syndromes took part, thus some syndrome groups consisted only of 1 to 3 individuals, and comparisons between these syndrome groups were not applicable.

The study could be improved by increasing sample size, as it would be interesting to explore whether these behavioural patterns are representative of all syndrome groups. Additional limitations relate to the measures used in this study and the adaptations made to them for use with this population. Interestingly, there was no sufficient pre-existing quality of life measure for children with ID. In this study, an additional quality of life measure was therefore utilised, which was used originally for children with chronic health problems. Adaptations of the suggested additional items having been made might well then change their psychometric properties; and further work is needed to validate these measures in ID populations. However, the modified subscales of Kiddy-KINDL appear particularly promising measures after the adaptations.

Existing data about self-help skills, and behaviour have been analysed in this study. The pros and the cons of analysing existing data have been described in detail in chapter 1 (Section 1.10) in this thesis. However, to overcome limitations it was established that the research questions from previous studies conducted within the Cerebra centre for Neurodevelopmental Disorders at the University of Birmingham were in line with the research questions of this current study. The questionnaires which were administered in the previous studies have well known psychometric properties. In addition, respondents' demographic data were cross checked across the syndrome dataset, ensuring that the same participants had taken part in both the previous and the current study. Moreover, a question regarding challenging behaviour was inserted into the online study to investigate whether individuals continue to display challenging behaviour at the time that this study was conducted, ensuring the validity of the behavioural data. However, it will be essential for future studies that standardised measures about self-help skills are repeated to investigate further changes in the ability level of individuals over the years.

Finally, the results are likely to be generalisable to a wide range of children with ID, as families of children diagnosed with a wide range of genetic syndromes took part in this study. Overall, these results highlight the importance of continuing to study factors associated with behavioural dysregulation separately from self-injury, aggression and property destruction, and in relation to parents and carers of people with genetic syndromes and ID. In addition, they point towards utilising a quality of life measure to gauge the importance of describing the wellbeing of children with challenging behaviour as well as the impact on the quality of life that these children are experiencing of challenging behaviour, parental emotional wellbeing and particular child behavioural characteristics.

CHAPTER 4

Service use for families of children with intellectual disabilities and challenging behaviour

4.1. Preface

Chapter 3 explored the characteristics associated with challenging behaviour in individuals with a range of genetic syndromes, as well as the association between challenging behaviour, children's quality of life and carer wellbeing. In summary, it was found that impulsivity was strongly associated with both challenging behaviour and parenting stress. This was the first time that the interrelationships between challenging behaviour, parental emotional wellbeing and the quality of life of children with intellectual disabilities and challenging behaviour had been investigated simultaneously. This chapter of the study will complement the findings in Chapter 3 by describing the services used by families and children with rare genetic syndromes associated with intellectual disabilities and challenging behaviour.

4.2. Introduction

People with intellectual disabilities face a number of difficulties across the lifespan. The presence of an intellectual disability is associated with a higher prevalence of co-morbid health problems, such as epilepsy (Morgan, Baxter and Kerr, 2003), hearing and visual impairments, and gastrointestinal problems (Van Schojenstein Lantman-de Valk et al., 1997). People with intellectual disabilities are also at higher risk of mental health problems and challenging behaviour (see Section 1.1 in Chapter 1). The association between challenging behaviour and intellectual disability was reviewed in this thesis (see Section 1.4.1.3 in Chapter 1). The association between challenging behaviour and particular genetic disorders has also been described (see Section 1.4.1.4 in Chapter 1). Alongside the emerging observations of an association between challenging behaviour and child behavioural and demographic characteristics there is growing evidence of a relationship between challenging behaviour and pain (Carr et al., 2003; O'Reilly, 1997; Carr & Owen-DeSchryver 2007; Luzzani, Macchini, Valadè, Milano & Selicorni, 2003; Leung & Robson 2007).

4.2.1. The impact of challenging behaviour on an individual's life

The impact of challenging behaviour on both the individual with an intellectual disability and their family has been explored in Chapter 3. Challenging behaviour is associated with risks to the physical safety of the individual and those around them, as well as restricted engagement in the community (Emerson, 1995). Challenging behaviour is also associated with greater emotional distress in parents (see Chapter 3; Hastings, 2002; Baker and Blacher, 2005). Furthermore, the cost of services for adults with intellectual disability and challenging behaviour is greater than for those who do not display challenging behaviour (Knapp, et al., 2005; Felce, et al., 2003) and these individuals also require a higher staff input (Felce et al., 2003). Children and adults displaying challenging behaviour are also at an increased risk of out of area placement (Allen, et al., 2007). Although challenging behaviour has serious effects on both the lives of individuals and their parents it is also essential for the impact of challenging behaviour on service use to be investigated.

4.2.2. The impact of intellectual disability and challenging behaviour on service use

Challenging behaviour can have a significant impact upon family functioning and on parental emotional wellbeing. Unsurprisingly, families of children with ID seek professional support in order to help manage and minimise the impact of such behaviours. Bromley et al. (2004) found that 75% of mothers of children with ASD were receiving help from speech therapists, and 45% have seen community paediatricians or school doctors. In the current study, the vast majority of children met the ASD criteria, and parents reported a range of disruptive behaviours that their children exhibited. Families of children who scored lower on language development reported seeing speech therapists more often. However, the involvement of speech therapists was not found to be associated with greater behavioural or emotional problems. In addition, mothers who reported that their children were more self-absorbed, or had greater language impairment and greater developmental delay, were more likely to be accessing health professionals. Although this study has earlier described the relationship between maternal emotional distress, children’s behavioural problems and informal support within the family, the relationship between using professional support and challenging behaviour has also been investigated.

A longitudinal study about health service provision, was conducted in the USA, by Newacheck, Inkelas and Kim (2004). They found that children with a range of physical disabilities and ID accessed higher levels of inpatient hospital services than children without disabilities. Specifically, children with disabilities were significantly more likely to be hospitalised. Additionally, children with disabilities used higher levels of physician services, psychologists, and social workers, and had approximately twice as many emergency department visits as children without disabilities. In addition, they were more likely to receive prescribed medications than children without disabilities were. However, no significant differences were found for use of dental services. In this study the association between ID and accessing medical professionals was investigated. In this context it is essential to review whether the presence of behavioural problems might be associated with increased use of the services of medical professionals.

Strydom et al., (2010) found that older adults with ID, mental health issues and physical disability were more likely to access medical professionals. However, in this study, the sample was of older adults, and the link between challenging behaviour and service use was not investigated. Similar findings were reported in Toms, Totsika, Hastings & Healy (2015), who reported that children with intellectual disability and mental health issues were more likely to access mental health services, compared to children with ID but no additional mental health problems. In addition, it was found that the rates of accessing mental health services were similar for children with ID and mental health issues and for children with mental health problems alone.

A study which was conducted by Knapp, Romeo &Beecham, (2007), found that children aged 4 to 17 with ID were more likely to use health services, social care services and education settings compared to children with an autism diagnosis but without ID. Although in this study the sample were children with ID, again the relationship between challenging behaviour and service use was not investigated. The relationships between intellectual disability, challenging behaviour and service use were investigated in Felce et al., (1998), where it was found that adults with ID and challenging behaviour were accessing more health professionals, including mental health services and social services. In the study which was conducted by Knapp et al., (2005), it was found that children with ID and challenging behaviour were using the services of health professionals, mental health services, education professionals and social services, according to the type of home residence. Most notably, people who lived in smaller homes/settings were more likely to use educational services compared to people living in private/voluntary homes, who were more likely to use health services or allied services and alternative services. Findings suggest that challenging behaviour influences the use of mental health services and of the services of dieticians.

There has been limited research investigating the aetiology of an intellectual disability and subsequent service use. Liptak et al., 2006, found that children with ASD were more likely to have outpatient and physician visits than children with heterogeneous intellectual disability. In a US study, children with ASD were associated with a greater number of medications and their families incurred significantly higher annual expenses for their total health care. Additionally, Mandell et al., (2005), found that school-aged children with ASD were more likely to receive educational and school based interventions than children with non-ASD diagnoses.

In summary, families of children with ID and/or challenging behaviour might access or use a wide range of professionals' services in order to help their children, and also to increase their own emotional wellbeing. Depending on the child's developmental level, behavioural problems, language skills and parental emotional distress, families of children with ID will seek the most appropriate services to provide targeted interventions for their children. In the following section, parental satisfaction will be described with regard to the services and the professionals used, for families of children with ID and challenging behaviour.

4.2.3. Parental satisfaction with service provision for children with ASD

Families of children with ID and challenging behaviour seek a range of professionals' support, tailored to the child and family's needs (Mandell et al*.,* 2005; Liptak et al*.,* 2006). However, little is known about parental satisfaction with the service provision. King et al., (1996) found that increased parental satisfaction levels were associated with information exchange, respectful and supportive care and partnership/enabling.

A large proportion of the research relating to ID and service satisfaction is focused upon families of children with an ASD. The studies focusing upon educational provision report that only 13% of parents of children with ASD reported a high level of satisfaction with the school's ability to address and meet their child's needs (Spann et al., 2003). Additionally, Kasari et al*.,* (1999), found that 50% of the parents who took part in their study wanted their child moved to another school.

The studies focusing upon parental perceptions of health services for ASD note that families of children with ASD were less positive than families of children with other disabilities about the ability of doctors to understand the impact of the disability on family functioning (Liptak et al., 2006). In comparison to parents of children with Down's syndrome and parents of children with a range of disabilities, parents of children with ASD were significantly less satisfied with the support they had received since their child was diagnosed (Bitterman et al*.,* 2008; Siklos & Kerns, 2006). This suggests that there is variability in the perception of services received, and highlights the need for more research in this area.

In addition, Kohler (1999) found that families of children with ASD received an average of 6.44 different services, in which some focused on the child's needs by providing preschool or school placements, therapeutic support staff, or speech and occupational therapy. Moreover, families accessed services, which focused on their own support by providing them with respite care, parent information classes and mobile services. However, 64% of parents who took part in this study reported at least one service as ineffective in addressing its intended purpose. Although this study provides a comprehensive description of families' service use regarding children with ASD, the small sample size (n=25 families), and the fact that this study is restricted to one geographical area limits the generalisability of the findings. In a recent study examining parental satisfaction with the services that they used for their children with ASD, it was found that 75% of parents reported overall satisfaction with the services that they used (Bayat, 2007). However, in this study information on the aspects that parents reported themselves satisfied with in the services was not provided. In summary, there is a need for the access to and use of services for children with ASD to be investigated.

In a study, which was conducted by Renty & Roeyers (2005), examining parental satisfaction with services among families of children with ASD, it was found that generally parents reported themselves satisfied with the educational support and other services that they used for their children. Most notably, the authors found that parental satisfaction was associated with the age of their children at ASD diagnosis. Similarly, Bromley, Hare, Davison & Emerson, (2004), found that parental satisfaction with the services used was associated with the child's age, the occurrence of severe developmental delays in the area of language development, independent functioning and socialization. In the same study, mothers were questioned about their satisfaction with the educational provision received. It was found that satisfaction was related to the suitability of their home for bringing up a child with ASD, the support received overall, the support received from the other family members, the amount of formal support, the number of children living at home, and the ability of the child to attend a school outside the LEA boundaries. Mothers whose children attended special schools rather than mainstream schools reported higher satisfaction levels with the educational services provided. However, no significant associations were reported between satisfaction with the educational services provided and gender, age, ethnicity of the child, household composition, household income, level of emotional disturbance or developmental delay of the child. Therefore, it appears that the factors that are associated with parental satisfaction with the services accessed or used are under-researched.

Conversely, Montes, Halterman & Magyar (2009) found that parents in the USA of children with ASD reported less access to and more dissatisfaction with school and community health services, compared to parents of children with special health care needs.

In summary, studies on parental satisfaction with the services used for children with ASD overall show that parents were less satisfied with the services used. However, further investigation is needed because even when the parents reported themselves satisfied, the factors associated with service satisfaction were not investigated. In the following section, parental satisfaction with the services used for children with ID will be reviewed.

4.2.4. Parental satisfaction with service provision for children with ID

The evidence about the satisfaction with service provision for families of children with disabilities other than ASD is less developed. The available studies have tended to focus upon what parents would like more of, rather than what they feel about current provision. For example, Fidler, Hodapp & Dykens, (2002), found high percentages of parents of children with Down's syndrome expressed a need for additional speech therapy. Similarly, parents of children with William syndrome expressed their desire for more one to one support in the classroom. Hatton et al., (2000), found that more speech and occupational therapy, as well as improved communication with the involved professionals, were listed as the desirable improvements by parents of children with FXS.

In terms of describing accessing services for children with ID Mitchell & Hauser-Cram, (2008), found that 73% of parents of children with ID considered the adolescent's primary care physician to be most important. A large number of parents responded either ‘‘do not know’’ or ‘‘not applicable’’ to three of the original items: communication with school (36%), communication with other services (52%), and communication with health insurance (53%). Examining the factors which are associated with parental satisfaction in this group of children authors found that providing reassurance and support to the parent, spending enough time with the adolescent during a visit, overall quality of care, showing respect for the family's culture, being available to advise over the telephone, explaining health needs in an understandable way, being easy to reach in an emergency, and respecting the adolescent all influence overall parental satisfaction with the services that they use .

In summary, challenging behaviour and intellectual disability may seriously influence children's quality of life and parental wellbeing. Therefore, given that children with rare genetic syndromes experience elevated health problems, parent stress and challenging behaviour, it is important that services provide input in these areas, but that the services also meet the physical and psychological needs of the child and their family members. In order to extend the literature on service use and perception to groups other than children with ASD, this study focused on a group of children who are likely to be using high levels of services, due to having both a rare genetic syndrome and challenging behaviour: and it documents both the services used and parental perceptions and satisfaction with such services.

4.3. Aims

1. To document the services used by children with rare genetic syndromes associated with an ID and challenging behaviour

2. To explore parental ratings of satisfaction with and effectiveness of the services provided

4.4. Methods

4.4.1. Participants

The parents or carers of 76 participants under 16 years old, with Angelman, Smith-Magenis, Prader-Willi, Cri Du Chat, Cornelia de Lange, Sotos, Lowe, Phelan McDermid syndrome, or 1p36, 8p23 and 9q34 deletion syndrome took part in this study. Participants were included based upon inclusion criteria set out in Chapter 3 (Section 3.6.1). Participants, as per the inclusion criteria, all exhibited challenging behaviour. To reduce the burden, questionnaire results on ability level were taken from informants' previous responses, collected six months prior to this study. The primary characteristics of the sample are presented in Table 4.1. High rates of mobility, verbal ability, hearing and vision were reported from most of the participants, using the Wessex scale to assess self-help abilities (see 4.4.2). Carers of children with Angelman syndrome, Cri du Chat, Cornelia de Lange syndrome, Phelan McDermid syndrome, and 1p36 and 8p23 syndrome reported low ability levels.

Table.4.1. Demographic characteristics of the sample

		Angelman	**Smith-Magenis**	**Prader-Willi**	**Cri Du Chat**	**Cornelia de Lange**	**Sotos**	**Phelan McDermid syndrome**	**1p36**	**8p23**	**9q34**	**Total Number**
N		22	8	10	12	6	4	3	6	3	1	76
Age	Mean	7.00	9.50	7.44	10.67	8.50	10.8	6.00	9.18	6.53	3.00	8.63
	SD	2.65	4.69	2.80	4.38	2.26	1.90	1	4.67	1.53	N/A	3.62
	Range	4-11	3-14	4-12	2-15	5-11	8-12	5-7	5-15	5-8	N/A	2-15
Gender	% male (N)	59.1 (13)	75 (6)	80 (8)	16.7 (2)	50 (3)	75 (3)	33.3 (1)	16.7 (1)	33.3 (1)	0	50.7 (39)
Ability	Able/Partly able % (N)	20 (3)	60 (6)	75 (12)	46.2 (6)	30 (3)	100 (6)	40 (2)	35.7 (5)	33.3 (2)	N/A	50 (45)
	Not able % (N)	80 (12)	40 (4)	25 (4)	53.7 (7)	70 (7)	0	60 (3)	64.3 (9)	66.7 (4)	100 (1)	50 (51)

Table.4.1. Demographic characteristics of the sample (continued)

		Angelman	**Smith-Magenis**	**Prader-Willi**	**Cri Du Chat**	**Cornelia de Lange**	**Sotos**	**Phelan McDermid syndrome**	**1p36**	**8p23**	**9q34**	**Total Number**
Vision	Poor/Blind%(N)	91.7 (11)	22.2 (2)	20 (3)	38.5 (5)	40 (4)	33.3 (2)	20 (1)	57.1 (8)	50 (3)	100	34.5 (39)
Hearing	Poor/Deaf%(N)	0	40 (4)	0	15.4 (2)	60 (6)	16.7 (1)	40 (2)	42.9 (6)	0	100	34.5 (21)
Mobility	Mobile%(N)	71.4 (10)	70 (7)	31.3 (5)	61.5 (8)	60 (6)	100 (6)	80 (4)	46.2 (6)	16.7 (1)	N/A	82.5 (53)
Speech	Verbal%(N)	33.3 (5)	80 (8)	93.8 (15)	69.2 (9)	50 (5)	100 (6)	60 (3)	42.9 (6)	100 (6)	N/A	67.7 (62)

4.4.2. Recruitment

The recruitment strategy was as described in the previous chapter (see Section 3.5.1 in Chapter 3). This study only focuses upon the data from the participants from the sample described in Chapter 3 who reported having experienced challenging behaviour in the last month. Three inclusion criteria were established.

(1) They were the primary carer of a person aged up to 16 years old with a diagnosis of one of the eleven syndromes listed above;

(2) The diagnosis of the genetic syndrome had been made by a professional, and

(3) The person they cared for had exhibited challenging behaviour within the last month.

Sixty-six participants completed the questionnaire online and 10 completed the paper copies they received.

4.4.3. Procedure

Parents were contacted either by email or letter, inviting them either to participate in an online questionnaire study, or to complete a paper copy of the study focusing upon the services accessed by children with rare genetic syndromes with challenging behaviour. A web link, password and ID number were emailed direct to parents, so that they could access the consent forms and questionnaire via an online questionnaire hosting site, ‘Lime survey’. Participants were notified that they could request a paper copy and 10 participants opted to return a paper copy. Four to six weeks after invitation letters had been sent, parents who had not responded were contacted via telephone to enquire whether they had questions or difficulties accessing the survey. The approximate time that they needed for the completion of the survey was 45 minutes.

4.4.4. Measures

4.4.4.1.Client Service Receipt Inventory (CSRI)

The CSRI is a measure of service provision, effectiveness and satisfaction (Knapp et. al., 2005). It can be adapted to suit different population groups etc. This measure is suitable for families of children with ID. It has been widely employed and has a multitude of variants. This tool enquires about the participant's access to, and use of, a range of services and of professionals who may support families of children with challenging behaviour. It asks for parental perceptions of the effectiveness of the services provided, and whether participants are satisfied with the provision. Most notably, participants could indicate the effectiveness and satisfaction of the services on a 5 point Likert scale, where scores 1 to 2 were the low rates and 4-5 the higher rates. A score of three was the baseline. The CSRI includes domains about the formal and informal support that families of children with ID receive, and parental perceived satisfaction and rating of the effectiveness of these services in helping to manage their child's challenging behaviour. CSRI is a descriptive tool for service provision, so frequency statistics about service provision can be produced. The aim of this tool is to investigate what kinds of services people are receiving, so validity and reliability measures are not applicable for this inventory. The number of participants who rated the usefulness and the effectiveness of the services differ in each category, as these two questions were not in mandatory fields in Lime Survey (see Table 4.6; 4.7).

4.4.4.2. Wessex Behaviour Scale (Kushlick, Blunden and Cox, 1973)

The Wessex Scale questionnaire is a short informant-based measure that assesses physical and social ability, developed by Kushlick et al., (1973). Domains covered include mobility, vision, literacy, hearing, continence, speech and self-help[3]. The questionnaire has been used in a wide range of published studies with people with ID (Moss et al., 2000; Oliver et al., 2008). The scale has modest reliability (Kappa value = .62 for overall scale and mean of .54 for item level). Higher scores on this measure indicate greater ability.

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[3] The self-help subscale is calculated by summing three items regarding independent washing, dressing and feeding ability. Each item is scored on a three point Likert scale, ranging from one (not at all) to three (without help), resulting in total scores ranging from three to nine.

4.5. Data Analysis

In order to address the first aim of the study, on what services children with CB and genetic syndromes access and use, descriptive statistics are presented about the professionals that children with challenging behaviour and genetics syndromes have seen in the last six months. These are initially presented for each profession, before the list of the professionals is grouped into six main categories: medical, psychology/mental health, advocate-social, learning, alternative therapists and allied health professionals (see Table 4.2).

Table 4.2 Categories of Professionals

Categories	**Professionals**
Medical	General Practitioner
	Learning Disability nurse
	Other community nurse
	Community Paediatrician
	Hospital based services
Psychology/mental health	Community Psychiatrist
	Clinical Psychologist
	Community Psychiatric nurse
	Community Mental Health Team Member
	CAMHS services and Psychologists
Advocate-social	Social workers
	Home help/ support workers
	Advocate/ counsellor
Educational	Educational Psychologists
	Speech therapists
	Special Educational Needs Co-ordinator (SENCO)

Table 4.2 Categories of Professionals (continued)

Categories	Professionals
Allied health professionals	Physiotherapists Podiatrists Audiologists Opticians Dentists Nutritionist/Dieticians Occupational Therapists Dysphagia services

For the second aim, to investigate how useful and effective parents find each of these services, descriptive statistics are presented on how effective the provided services are, based on parental reports. Both the usefulness and effectiveness of the services were rated on a 5 point Likert scale, where lower usefulness and lower effectiveness scores were rated with 1 or 2, and 3 was the baseline about usefulness and effectiveness. Scores 4 and 5 both indicate high levels of usefulness and effectiveness of the services. However, each service category includes particular professionals: thus both the usefulness and the effectiveness of each category were computed by calculating the mean of usefulness and effectiveness of all the professionals who comprise each category. To investigate the usefulness of the services, again descriptive statistics are presented.

4.6. Results

As each syndrome group includes a limited number of people, the results are presented as a collective sample of children with rare genetic syndromes. Descriptive statistics are presented in Table 4.2 by syndrome group and the total sample. Due to the small sample sizes of some syndrome groups, descriptive statistical analysis of the different syndrome groups cannot be undertaken.

In summary, 50% (31 participants) reported their children as of low ability levels. Most notably, those scoring between zero and eight had a clearly identified level of lower ability, based on the Wessex scale, which was used as a proxy estimate of level of ability. This was done in order to increase control over the ability levels of the groups for between-group analyses. The descriptive statistics about the formal service provision are presented in Table 4.3. This shows that the three professionals seen most commonly within the six months prior to completing the questionnaire were Dentists (71.1%), Speech Therapists (69.8%) and General Practitioners (61.8%). Of concern is that this also means that 28.9% of children involved in this study had not seen their dentist in the previous six months.

Table 4.3. Percentage of participants who have seen the service specialists in the last six months

Specialists	**% seen this professional in the last six months**
Dentist	71.1
Speech therapist	69.7
General Practitioner	61.8
Occupational therapist	57.9
Community Paediatrician	53.9
Social worker	53.9
Physiotherapist	50.0
Optician	47.4
Home help/support worker	38.2
Audiologist	32.9
SENCO	31.6
Educational Psychologist	27.6
Learning disability nurse	27.6
Nutritionist/dietician	26.3
Art/drama/music therapist	23.7
CAMHS services	21.1
Other community nurse	19.7
Clinical Psychologist	18.4
Hospital based services	17.1

Table 4.3. Percentage of participants who have seen the service specialists in the last six months (continued)

Specialists	**% seen this professional in the last six months**
Community Psychiatrist	11.8
Psychologists	11.8
Advocate/counsellor	10.5
Play therapist	9.2
Community mental health team member	7.9
Alternative therapist	7.9
Community Psychiatric Nurse	6.6
None of the Above	2.6
Dysphagia service	1.3

The range of professionals was then categorised into six broader professional categories: medical, psychology/mental health, advocate/social, learning, alternative therapists and allied health professionals, as per Table 4.3. Descriptive statistics about professionals seen within these broader categories are described in Table 4.4.

Table 4.4. Percentage of participants accessing services (by group) in the last six months

Categories	**% who have seen this profession in last six months**
Allied Health Professionals	69 (92%)
Medics	60 (80%)
Educational	55 (73.3%)
Advocate/Social	49 (65.3%)
Psychology/Mental Health	26 (34.7%)
Alternative Therapists	24 (32%)

Table 4.4 shows that when the professionals are combined into category groups, the allied health professionals category is listed as the one most accessed by participants, with 92 % of the sample (69 participants) reporting to have seen one or more allied health professional in the last 6 months. In contrast, only 32% of carers indicated that they had seen alternative therapists in the last six months.

4.6.1. Parental perceptions of the usefulness and effectiveness of the accessed services in helping to manage their child's challenging behaviour

The second aim of the study was to describe how useful and the effective parents found the services they accessed in helping to manage their child's challenging behaviour. The ratings for each professional category are provided in Tables 4.5 and 4.6 below. Not all parents rated all the professions for their effectiveness or satisfaction, so the numbers providing ratings for each professional category are documented in the data tables.

Table.4.5 Parent's rating of the usefulness of professional categories in relation to managing their child's challenging behaviour.

Categories	Number ratings provided	Not very useful 1	2	3	4	Very useful 5
Allied Health Professionals	63	0	1 (1.5%)	11 (17.2%)	33 (51.6%)	19 (29.7%)
Alternative Therapists	24	1 (4.2%)	2 (8.3%)	2 (8.3%)	9 (37.5%)	10 (41.7%)
Advocate/Social	44	1 (2.3%)	6 (13.5%)	5 (11.4%)	16 (36.4%)	16 (36.4%)
Educational	51	3 (5.8%)	4 (7.6%)	8 (15.4%)	16 (30.8%)	21 (40.4%)
Medics	56	0	8 (14.3%)	13 (23.2%)	21 (37.5%)	14 (25%)
Psychology/Mental Health	24	1 (4.2%)	2 (8.3%)	6 (25%)	7 (29.2%)	8 (33.3%)

Table. 4.6 The usefulness of the provided services based on parental reports.Descriptive statistics about the usefulness and effectiveness of professional categories were provided (see Table 4.6 and Table 4.7).

Categories	**Number of ratings from**	**Less Useful**	**More Useful**
Allied Health Professionals	63	1 (1.5%)	52 (51.6%)
Alternative Therapists	24	3 (12.5 %)	19 (79.2 %)
Advocate/Social	44	7 (15.8 %)	32 (72.8 %)
Educational	51	7 (13.4 %)	37 (71.2 %)
Medics	56	8 (14.3%)	35 (62.5%)
Psychology/Mental Health	24	3 (12.5 %)	15 (62.5 %)

The table shows the variability in parental perceptions of effectiveness. Despite being the least accessed professional category, alternative therapists received the highest proportion of “Very Useful” ratings from parents. Only 6 parents rated any professionals as “Not Very Useful” in relation to managing their child’s challenging behaviour.

Descriptive results in Table 4.6 revealed that the 79.2% of parents reporting alternative therapists as useful was the highest proportion. Similar high rates about usefulness were reported for advocacy/social services (72.8 %) and learning (71.2 %). However, 14.3% of parents rated medical professionals as less useful in helping them to manage their child’s challenging behaviour. Parents were also asked to rate the effectiveness of the support provided. The data are provided in Table 4.7.

Table 4.7 Parent's ratings of the effectiveness of professional categories in relation to managing their child's challenging behaviour

Categories	**Number of ratings**	**Not effective** **1**	**2**	**3**	**4**	**Very effective** **5**
Alternative Therapists	22	1 (4.5%)	0	4 (18.2%)	9 (40.9%)	8 (36.4%)
Allied Health Professionals	61	0	2 (3.2%)	15 (24.2)	29(46.8%)	16 (25.8%)
Advocate/Social	42	1 (2.4%)	6(14.3%)	6 (14.3%)	14(33.3%)	15 (35.7%)
Medics	53	0	7(13.3%)	14(26.4%)	19(35.8%)	13 (24.5%)
Psychology/Mental Health	20	2 (10%)	1 (5%)	6 (30%)	4 (20%)	7 (35%)
Educational	49	3 (6%)	8 (16%)	12 (24%)	19 (38%)	8 (16%)

Similar to the results for the usefulness ratings, 36.% of parents who had seen alternative therapists gave them the highest rating of being "Very effective" in helping to manage their child's challenging behaviour, although advocate/social professionals and educational professionals received a very similar level of endorsement from parents, at 35.7% and 35% respectively. To refine the effectiveness rating from a descriptive approach based on parental reports effectiveness rates of 1 and 2 were combined into one effectiveness category ('less effective') and ratings of 4 and 5 the 'more effective' category (see Table 4.8).

Table 4.8 Describing the effectiveness of the accessed services

Categories	**Number of ratings**	**Less Effective**	**More effective**
Alternative Therapists	22	1 (4.5%)	17 (77.3%)
Allied Health Professionals	61	2 (3.2%)	45 (72.6%)
Advocate/Social	42	7 (16.7%)	29 (69%)
Medics	53	7(13.3%)	32 (60.3%)
Psychology/Mental Health	20	3 (15%)	11 (55%)
Educational	49	11 (22%)	27 (54%)

When the ratings of 4 (useful) and 5 (very useful) are combined, alternative therapists remain the most endorsed professional group, with 77.3% of participants who had seen an alternative therapist in the last 6 months rating them as "more effective" at managing their child's challenging behaviour. 22% of parents reported that educational professionals were less effective at helping to manage their child's challenging behaviour. Only 3.2% of parents reported allied health professionals as being "less effective".

4.7. Discussion

This study explored the services accessed by children with rare genetic syndromes associated with ID who are showing challenging behaviour. Additionally, the perceived effectiveness and the satisfaction with the provided services in relation to managing the child's challenging behaviour were described. Importantly, the recruitment of a sample of children with rare genetic syndromes and challenging behaviour ensures a robust exploration of service use in this population. Most notably, recruiting participants with various genetic disorders associated with ID enhances the generalisability of the descriptive findings on children with ID. The results of the study revealed that challenging behaviour influences what services families are using for their children. Additionally, it was found that challenging behaviour itself influences which services parents evaluated as effective and useful, based on responding to children's needs.

The first aim of the study was to describe the services that families of children with ID and challenging behaviour are using. The results of this study revealed that the group of professionals most frequently accessed by families of children with challenging behaviour are allied health professionals and medical professionals. These findings are in line with the findings reported in Felce et al., (1998); Knapp et al., (2005) that people with ID were more likely to use health services. However, the first study cited analysed data from adult individuals with ID. The second study, conducted by Knapp et al. (2005), employed a sample of children with ID. A plausible explanation of why families of children with ID and challenging behaviour used health services is revealed through the link between pain and challenging behaviour (see Section 1.6.3 in Chapter 1; Section 3.1.3 in Chapter 3). Although in this current study families of children with intellectual disability and challenging behaviour used mental health services, the relationship between challenging behaviour and service use is more fully investigated in the next chapter.

The second aim of this study was to describe the overall effectiveness and usefulness of the services used, based on parental reports. Parents rated the usefulness and effectiveness of the services used in the last six months. When interpreting the data, it must be remembered that only the parents who had used these services within the last six months were asked to rate their effectiveness and usefulness. Therefore, parents who had disengaged from the services more than six months ago, due to finding them ineffective, were not able to document their experiences. Descriptive results in this study revealed that allied health professionals and social and alternative therapists were rated highly by parents of children with ID and challenging behaviour. Conversely, lower ratings of usefulness were reported for mental health professionals. Similar descriptive results were reported about the effectiveness of the services used. Similar findings about the usefulness and effectiveness of the services used were reported in the literature. Most notably, Hare, Part, Burton, Bromley and Emerson (2004), found that medical professionals and day services were rated as very useful by parents of adults with autism. However, this sample was of adults with intellectual disability and therefore it was essential that the usefulness and effectiveness of accessed services in younger populations with intellectual disability and additional behavioural issues should be investigated. In another study, conducted by Bromley et al*.,* (2004), it was found that alternative therapists and social services were rated by the majority of the sample as accessible, appropriate and sufficient service provision for their children. Additionally, lower effectiveness rates about psychiatrists were reported by parents of children with autism. These findings are in line with the descriptive results of this current study. Except for studies which focused on parental satisfaction with the accessed services regarding their children diagnosed with ASD, the findings of this current study are in line with Mitchell's and

Hauser-Cram's (2006) findings, where parents of children with ID were satisfied with the medical professionals. A similar finding about parental satisfaction with alternative therapists was provided by Hatton et al. (2000), where it was found that more occupational therapy and speech therapy, as well as improved communication with the involved professionals, were listed as the desirable improvements by parents of children with FXS. However, in this current study educational professionals, including speech therapists, were not rated as highly as in Hatton's et al. (2000) study. Moreover, Hatton's et al. (2000) study focused on children with FXS, whereas this current study has investigated a sample of participants with various genetic syndromes but not with FXS.

These results, however, must be interpreted with caution. Although this sample includes a wide range of genetic syndromes, making the findings suitable to be generalised for people with ID, there is no control group for challenging behaviour. Additionally, the recruited families may differ in socioeconomic status and therefore service use may not be representative of the broader population with ID and challenging behaviour. Most notably, Knapp et al*.,* (2005) found the type of home residence influences service use. However, Bromley et al., (2004) found that neither gender, age nor ethnicity of the child, household composition or household income, level of emotional disturbance or developmental delay of the child were associated with service effectiveness or usefulness.

The descriptive results in this study indicate that families of children with challenging behaviour are more likely to access support from medical and allied health professionals. Secondly, the effectiveness and the usefulness of the services were evaluated based on parental reports, which were in relation to the services received commonly in the previous six months. As discussed in Chapter 3 there is an association between challenging behaviour, parental emotional distress and children's quality of life. The findings of that analysis lend strength to the suggestion that challenging behaviour influences service provision. Therefore there is a pressing need to investigate further the potential relationship between service provision and challenging behaviour.

The present study is limited by the small sample size recruited. The small sample size limits generalisation of the results. An additional limitation of the study was that there was no control group for challenging behaviour. Therefore it is not possible to compare service use by families of children with and without challenging behaviour.

Another limitation which needs to be taken into account is about analysing existing data. Participants' demographic data alongside data on self-help skills and behaviour have been collected and analysed in previous studies conducted within the Cerebra Centre for Neurodevelopmental Disorders at the University of Birmingham. The advantages and disadvantages of analysing existing data have been described in depth in Chapter 1 (Section 1.10). However, to overcome these limitations in the current study a question regarding challenging behaviour was introduced in the online survey to investigate whether challenging behaviour had fallen over the years and to establish the validity and reliability of the findings. In addition, standardised questionnaires about self-help skills had also been used in previous studies, which strengthens the validity of the study data. Finally, respondent's demographic data were extracted from the last study conducted, which was no more than six months prior to the implementation of the current study, to minimise major changes in the socio-economic background of the participants.

In summary, the results revealed that families of children with ID and challenging behaviour are using the services of health professionals, learning services and social services. Additionally, families highly rated the usefulness and effectiveness of alternative therapists, health professionals and social services. Lower ratings were reported for mental health services by parents of children with ID and challenging behaviour. These findings support the necessity and desirability of exploring further the factors that influence what services families of children with challenging behaviour will use.

CHAPTER 5

Factors which are associated with service use for families of children with intellectual disabilities and challenging behaviour

5.1. Preface

The study reported in Chapter 4 had two main aims; it explored the services accessed by families of children with rare genetic syndromes associated with intellectual disabilities and challenging behaviour, and it reported parents' satisfaction with these services. More specifically, regarding the first aim of the study, it was found that families of children with intellectual disabilities and challenging behaviour were using allied health and medical services more frequently and alternative therapists less frequently. Parental satisfaction with the services used was described by assessing the usefulness and the effectiveness of the services according to parental reports. Most notably, it was found that allied health professionals, social services and alternative therapists were rated highly in terms of both usefulness and effectiveness by parents of children with intellectual disabilities and challenging behaviour. Conversely, lower rates on usefulness and effectiveness were reported for mental health professionals. This study will extend the findings of Chapter 4 to explore the factors which are associated with services that families of children with challenging behaviour have used.

5.2. Introduction

There are many factors which may influence a family's decision to seek help from professionals. Due to the limited quantity of research specifically on children with intellectual disabilities, this introduction will first report on the literature pertaining to children without intellectual disabilities in order to identify factors that may influence service use, before discussing the intellectual disability- focused literature. The factors identified in these general studies will then be contrasted and compared to those factors found in studies about children with intellectual disabilities.

5.2.1. Factors influencing service use in relation to children without intellectual disabilities

There is evidence in the literature about the factors which are associated with service use in children with either mental health issues or chronic ill-health, or children of low-income families. Most notably, the factors which have been associated with use of mental health services are socio-demographic, drawing on information such as age and ethnicity. Leslie et al.'s (2000) study aimed to identify factors influencing outpatient mental health service use by children in foster care, by using data from 480 children aged between 2 and 16 years old in the USA. They found that the total number of outpatient mental health visits increased with age, and males were more likely to visit mental health professionals than females. Additionally, ethnicity was a significant predictor for mental health service use, with Asian-American and Latino youth under-represented in existing public mental health facilities. Finally, children with severe behavioural problems were more likely to attend more mental health appointments. Cornelius et al. (2001) investigated parental mental health issues, and the number of siblings, as possible factors that might influence mental health service use. They found that parental psychopathology, parental substance use, the number of siblings, and a diagnosis of ADHD and ODD could significantly predict mental health service use by male adolescents. However, the modest sample size, and the recruitment procedure, where participants were self-selected, limit the generalisability of these findings.

Zahner and Daskalakis, (1997) compared factors associating service use in mental health departments, general health departments and school settings. They found that socio-economic status was significantly associated with mental health and school support, while health problems, age, parental emotional distress and aggressive behaviour were found to significantly predict service use in mental health, general health and school settings. Most notably, it was found that health problems were associated with more use of all services, not only health services. This highlights the importance of considering health problems as possible factors for analysing service use in future studies.

There is evidence in the literature that there are particular factors which are associated with access in health services. Most notably, Janicke et al., (2001) found that information about a child’s current and past health status, psychosocial variables and maternal psychological distress predicted the amount of health care visits. However, the generalisability of the findings is restricted by the recruitment procedure, where the selected participants belong to middle socioeconomic classes. The association between socio-economic status and access in health services was confirmed in another study by Newacheck, et al. (1998), who found that age, gender, ethnicity, parental educational level and socio-economic status significantly predict access in health services for children with special health care needs.

In summary, there is evidence in the literature about the factors that might influence service use in relation to children without intellectual disabilities, specifically those who experience mental health issues, chronic ill-health or are children of low-income families. Factors identified were socio-economic status, child's mental health and general health status, parental emotional distress, child's age and gender, which were found to predict help-seeking behaviour, or the amount of health service visits.

5.2.2. Factors influencing service use in relation to children with intellectual disabilities

The increased use of services in families of children with intellectual disabilities is evidenced in many studies (Boulet, Boyle and Schieve, 2009; Nachshen and Minnes, 2005; Knapp et al., 2005; Barron, Molosankwe, Romeo and Hassiotis, 2013). However, research is lacking specifically focusing upon services used and exploring predictive factors.

Many studies combine findings relating to children with intellectual and physical disabilities. Newacheck, Inkelas and Kim, (2004) surveyed 13,792 children, of whom 7.3% had physical or intellectual disabilities, and were aged less than 18 years old. They found that age, gender, ethnicity and socio-economic status significantly predict the amount of health service use and the expenditures for these services.

In Chapter 1 the lifelong problems of people with intellectual disabilities were reviewed extensively (see Section 1.1 in Chapter 1). The potential under-diagnosis of chronic health conditions that require ongoing management was implicated in several recent studies (Janicki et al., 2002; Lewis et al., 2002; Merrick et al., 2004). Most notably, a small sample study conducted in the United Kingdom by Hensel, Rose, Kroese, and Banks-Smith (2002), found that individuals with intellectual disabilities reported that they had received more health checks than the controls over the previous year, that is compared to people without intellectual disabilities (the control group). The influence of health problems on service utilisation for individuals with intellectual disabilities has been reviewed in Morgan, Baxter and Kerr's (2003) study. Most notably, the authors found that individuals with intellectual disabilities who also had epilepsy used in-hospital services 2.5 times more frequently than those who did not have epilepsy.

In addition, the association between health problems and service use for individuals with intellectual disabilities has been reviewed in Lin, Wu and Lee (2003; 2004) studies. Most notably, in both studies it was found that having an additional health problem, being younger, and needing rehabilitative care predict health service utilisation. Additionally, Pruchno and McMullen (2004) found that there are different variables/factors which predict different types of services that people with intellectual disabilities accessed. Most notably, young adults with intellectual disabilities were more likely to receive psychology services if they were black, violent, and noncompliant. Predictors for unmet dental needs included being black, mother having difficulty in paying bills, living in a community with low to moderate levels of spending on disability services, being nonviolent, and being noncompliant. The relationship between a child's challenging behaviour and social support access has been described briefly in White and Hastings' (2004) study, in which it was found that accessing professional and service support appeared to be more strongly associated with the child's needs rather than with parents' needs. However, social support services included a range of professionals such as medical doctors and psychologists. In this particular online survey medical doctors and psychologists were placed in different service categories.

In Chapter 1 it was found that people with intellectual disabilities are more likely to show behavioural problems or to develop mental health problems, compared to typically developed individuals (see Section 1.1). The association between behavioural problems and mental health services was reviewed in Halstead et al's (2000) study. It was found that severe disability and behavioural problems predicted frequent primary care contact. Whitt, Kasper and Riley, (2003) aimed to examine the use of mental health services and the correlates of receiving services among children with physical and intellectual disabilities, aged between 6 and 17 years old. Data from 4,939 children with disabilities were analysed in this longitudinal study. It was found that age, ethnicity, financial problems and child's health status influenced the amount of mental health support received. The study also showed that the involvement of health professionals in care coordination was associated with greater access to mental health care for children with disabilities.

Floyd and Gallagher (1997) investigated levels of parental stress, the use of health services in children with physical or intellectual disabilities, some of whom had challenging behaviour, and the association between these variables. They collected data from 231 parents of children with intellectual disabilities or chronic illness. Results suggested that the presence of severe behavioural problems was related more to parental stress than to disability type. Additionally, the presence of severe behavioural problems was related to mental health services use. Floyd and Gallagher, (1997) found that parents of children with intellectual disabilities were more stressed about the continuation of ongoing care into adulthood. Interestingly, although single mothers were not more stressed than mothers from two-parent families, they did use more services.

A method of documenting services used that is frequently used in intellectual disability research is through cost data. Doran et al., (2012) investigated the cost of services accessed in a sample of individuals with intellectual disability in Australia. They found that the cost of the service used increased according to the severity of intellectual disabilities. In another Australian study, Einfeld et al., (2011) found that behavioural and emotional problems were associated with increased service costs among children with intellectual disabilities.

Barron et al., (2013) also found that the degree of intellectual disability was strongly associated with the cost of care, especially in relation to educational costs. Similar findings were reported in a study about service use and costs in children with intellectual disabilities (Barrett et al., 2015). Most notably, the authors found that the degree of intellectual disability and age were related to the educational service use. However, families of children with intellectual disabilities and challenging behaviour reported having less access in relation to community nurses trained in intellectual disability.

Knapp et al., (2005) conducted a study focusing on the association between intellectual disability and service utilisation in a group of people with intellectual disabilities living in care in England. They investigated the impact of challenging behaviour on service provision or in relation to service use for people with challenging behaviour. Their sample of 930 people over 18 years old with intellectual disabilities and/or challenging behaviour completed a series of standardised measures in order for this association to be explored. It was found that the degree of intellectual disability and the extent of challenging behaviour were both associated with high service costs. Additionally, it was found that the presence of challenging behaviour was associated with the kind of service that people were more likely to seek. Most notably, individuals with challenging behaviour were receiving more mental health services, such as those of psychiatrists and psychologists, than those without challenging behaviour. This study revealed that access to and the use of services by people with intellectual disabilities were not always linked to perceived or actual needs. Unsurprisingly, sector and the type of home residence also influenced service use and cost in various ways.

Focusing upon cost data is helpful for service planning and for making an economic argument, but it does not provide detail on the range of professionals accessed, and whether the children accessed a few expensive professionals or had regular contact with less expensive professionals. However, these studies highlight the importance of investigating the associated factors of service use for families of children with intellectual disabilities and behavioural problems.

In summary, demographic variables, such as age and gender, parental socio-economic status, health problems and parental mental health issues, were found to predict service use for individuals with intellectual disabilities. Additionally, health problems, the degree of intellectual disability, and parental mental health issues were found to predict service use for families of individuals with challenging behaviour. Whilst the research has identified those factors associated with increased service use, to date nobody has investigated the use of services in specific sub- population groups, such as individuals with genetic syndromes associated with intellectual disabilities. There are also limited data describing the actual professionals accessed rather than the outcome measure of cost.

5.3. Aims

1. To identify factors associated with increased service use in relation to children with rare genetic syndromes showing challenging behaviour
2. To explore whether the topography of challenging behaviour is associated with different factors predicting service use.

5.4. Methods

5.4.1. Participants

The parents or carers of 65 participants under 16 years old, who live in the United Kingdom, with Angelman, Smith-Magenis, Prader-Willi, Cri Du Chat, Cornelia de Lange, Sotos, Lowe, Phelan McDermid syndrome, and 1p36, 8p23 and 9q34 deletion syndrome took part in this study. The primary characteristics of the sample are presented in Table 4. 2 (see Chapter 4).

5.4.2. Recruitment

The recruitment strategy has been described in detail in the previous chapter. This study only focuses upon the data from the participants drawn in this study from the sample described in Chapter 3 who reported having challenging behaviour. Three inclusion criteria were established.

(1) Participants were the primary carer of a person aged up to 16 years old with a diagnosis of one of the eleven syndromes listed above;

(2) The diagnosis of the genetic syndrome had been made by a professional, and

(3) The person they cared for exhibited challenging behaviour.

5.4.3. Measures

5.4.3.1. Wessex Scale

The Wessex Scale (Kushlick et al., 1973) was used to assess adaptive ability in children and adults with intellectual disabilities. It comprises five subscales including: continence, mobility, self-help skills, speech and literacy. For the purpose of this study, the self-help subscale was used as an estimate of degree of ability, and responses to items on mobility, speech, reading, writing and counting were used to further describe the groups. The Wessex Scale has good inter-rater reliability at subscale level for both children and adults (Kushlick et al., 1973; Palmer & Jenkins, 1982).

5.4.3.2. Demographic Questionnaire

A demographic questionnaire that required information on date of birth, gender, diagnosis, parental educational level, parental annual income, post code, and number of siblings had been administered 6 months prior to this online survey by researchers within the Cerebra Centre for Neurodevelopmental Disorders, recruiting the same participants who took part in this online survey. The socio-economic status of the participants was calculated by extracting information about the post code, the educational level of parents and family annual income.

5.4.3.3. Client Service Receipt Inventory (CSRI)

This measure has been described in Chapter 4 (see Section 4.1.4).

5.4.3.4. The Health Questionnaire (HQ)

This measure has been described in Chapter 3 (see Section 3.1.4).

5.4.3.5. Hospital Anxiety and Depression Scale (HADS)

This measure has been described in Chapter 3 (see Section 3.1.4)

5.4.3.6. Kiddy- KINDL

This measure has been described in Chapter 3 (see Section 3.1.4)

5.4.3.7. Challenging Behaviour Questionnaire (CBQ)

This measure has been described in Chapter 3 (see Section 3.1.4).

5.5. Data analysis

Due to small numbers, predominantly categorical data and differences in sample size, non-parametric tests were employed. First, Mann-Whitney U analysis was undertaken to explore the differences in terms of self-help, quality of life, anxiety, depression, age, health problems and socioeconomic status between those children who received either medical, mental health, social, learning, alternative and allied health support and those who did not. As the analyses were exploratory, alpha was kept at .05.

The Bonferroni correction is an adjustment made to P values when several dependent or independent statistical tests are being performed simultaneously on a single data set. To perform a Bonferroni correction, one divides the critical P value (α) by the number of comparisons being made. The statistical power of the study is then calculated based on this modified P value. The Bonferroni correction is used to reduce the chances of obtaining false-positive results (type I errors) when multiple pairwise tests are performed on a single set of data. However, research has shown that adjusting statistical significance for the number of tests that have been performed on study data, the Bonferroni method, creates more problems than it solves. In addition, it has been shown that the Bonferroni method is concerned with the general null hypothesis (that all null hypotheses are true simultaneously), which is rarely of interest or use to researchers. The main weakness, however, is that the interpretation of a finding depends on the number of other tests performed. Moreover, the likelihood of type II errors is also increased, so that truly important differences are deemed non-significant. Therefore, in this study, binary logistic regression was used to investigate which of the differences that have been found significant in Mann-Whitney U tests might predict service use for families of children with intellectual disabilities and challenging behaviour.

Socio-economic status was calculated by adopting a cut-off point of 10% on deprivation scores, as this has been selected in other studies. Two new variables in SPSS were created. The deprivation code is the percentage of people who live in deprived areas, and the socio-economic code is the cut off 10%, where 0 refers to the least deprived areas and 1 are the most deprived areas.

Overall this study aimed to investigate the factors predicting service use for families of children with genetic disorders and challenging behaviour. A series of binary logistic regression calculations using the enter method was undertaken.

5.6. Results

In order to explore the first aim, to examine whether there was any difference in the factors identified in the introduction above as predictive of services accessed, differing between those who did and did not access services, a series of Mann-Whitney U tests was conducted. These had self-help, quality of life, parental anxiety and depression, child's age, health problems and socio-economic status as the dependent variable, and accessing or not accessing the services as the independent variable.

The factors explored were child quality of life (as measured by the raw score on the Kiddy-KINDL scale), parental anxiety and depression (measured by the raw score on the Hospital and Depression Scale), participant age, participant ability (measured by the Wessex Self-Help score), health problems (as measured by the number of current health problems reported on the Health Questionnaire) and socio-economic status (calculating the deprivation 10% cut off).

Tables of results are presented in Appendix H. The only significant differences were (1) that parental anxiety scores were higher in families who used mental health services for their children (U =289, 0, p =.028), and (2) children who had used social/support services were significantly older than children who had not (U=211, 0, p=.021). No significant differences were found between those who had used the services of learning services, alternative services and allied health professionals (see Appendix H).

5.6.1. Factors predicting service use for families of children with intellectual disabilities and challenging behaviour

The last aim of the study was to explore the factors which predict service use. Binary logistic regression analyses were performed to assess whether the use of particular services is predicted by the age of children, parental anxiety or depression, socio-economic status, self-help and child gender (see Appendix I). Information about parental anxiety and depression was collected via HADS, and the cut-off scores for anxiety and depression were analysed accordingly and separately. Cut-off scores for anxiety and depression discriminate between parents of children who met the cut-off criteria for anxiety and/or depression and those who did not. The employment of cut-off scores was necessary in the binary logistic regression, where the possible predictive variables needed to have only two levels.

In brief, the results reflect the results of the Mann Whitney-U analysis above and show that parental anxiety can predict access to mental health services. The model contained six independent variables (self-help, age, gender, SES, anxiety and depression). The full model containing all predictors was not statistically significant, ($\chi 2$ (6, n = 46) = 7.296, p =.276), indicating that the model was not able to distinguish between participants using mental health services and those who do not. The model as a whole explained between 14.6% (Cox and Snell R square) and 21.9% (Nagelkerke R squared) of the variance in mental health service use, and correctly classified 76.1% of cases. However, it was found that parental anxiety predicts using mental health services. The Wald criterion demonstrated that only parental anxiety made a significant contribution to prediction (p = .048). The Exp (B) value indicated that parents were1.37 times more likely to use mental health services. Parental depression, child's gender, socio-economic status and self-help were not found as significant predictors about the use by families of particular services (see Appendix 5.1). Binary logistic regression was undertaken to assess the impact of self-help, age, gender, socio-economic status, anxiety and depression on the likelihood of participants’ use of social/support services. The model contained six independent variables (self-help, age, gender, SES, anxiety and depression). The full model containing all predictors was statistically significant,

($\chi2$ (6, n = 46) =12.847, p =.048), indicating that the model was able to distinguish between participants using social/support services and those who did not. The model as a whole explained between 24.4% (Cox and Snell R square) and 34.4% (Nagelkerke R squared) of the variance in social/support service use, and correctly classified 71.7% of cases. The Wald criterion demonstrated that only child's age made a significant contribution to prediction (p = .034). The Exp (B) value indicated that older children were1.32 times likely to access social/support services (see Appendix 5.1).

5.7. Discussion

This study had two basic aims. First, to explore whether the different factors are associated with service use and to identify factors associated with increased service use in children with rare genetic syndromes showing challenging behaviour.

The first aim of the study was to investigate plausible factors that are associated with service use among families of children with intellectual disabilities and challenging behaviour. Potential factors were child's age, self-help ability, socio-economic status, anxiety, depression, and child's gender. These factors were evidenced in the literature to be associated with or to predict service use for people with disabilities and/or behavioural problems (Knapp et al., 2005; Floyd and Gallagher, 1997; Barron et al., 2013; Barrett et al., 2014 and Doran et al., 2012). In this study, it was found that parents using mental health services for their children had higher anxiety scores than parents not using such services. Similarly, Floyd and Gallagher, (1997) found that parental stress was strongly associated with the receipt of child mental health services. It was also found that parental anxiety was strongly related to the child's behavioural problems. The relationship between service use, parental emotional distress and parental quality of life was investigated in White and Hastings, (2004). The authors found that parental emotional distress was associated with the child's characteristics, including behavioural problems, parental wellbeing and mental health support. The association between challenging behaviour and parental emotional distress was investigated in Chapter 3 of the present study.

In this study, although the socio-economic status of families of children with intellectual disabilities and challenging behaviour might have been associated with possible variations in using particular services it did not predict use of the services of medical, learning, alternative and allied health professionals based simply on the socio-economic status of families of children with intellectual disabilities and challenging behaviour. Although socio-economic status as a putative factor associated with service use has been reviewed extensively in the introduction of this chapter, citing Newacheck, Inkelas and Kim, (2004), in this study it was not revealed as an associated factor. However, a larger sample size and/or a greater variation of socio-economic status among the participants might generate this as a factor associated with service use among families of children with intellectual disabilities and challenging behaviour.

The association between the level of intellectual disability and service use was evident in Doran et al's., (2012) study, in which the authors found that service use increases according to the severity of intellectual disabilities. In this study, it has been investigated whether service use can be predicted by the rated self-help ability which is associated with the level of intellectual disability. In this study, self-help was not a significant predictor for any of the following service categories: medical, mental health, social/support, learning, alternative and allied health professionals. However, because of the small sample size, variations after regression analysis were not enough to strongly justify the above finding. Moreover, to test whether the level of intellectual disability is associated with service use it is essential that a control group without intellectual disabilities be added.

Data about self-help skills, behaviour and demographic data were collected and analysed from an existing dataset. The pros and the cons of analysing existing data have been described in depth in chapter 1 (Section 1.10). To overcome these limitations standardised questionnaires that had been used in previous studies conducted in Cerebra Centre for Neurodevelopmental Disorders at University of Birmingham were used, an approach which strengthens the validity of the findings. In addition, a question regarding behavioural problems was inserted in the online survey to investigate whether previous findings on challenging behaviour were consistent with the findings of the current study.

Age was found to be a significant predictor for using social and support services for families of children with intellectual disabilities and challenging behaviour. When looking at the results, it could be inferred that this might be associated with increased access to services when preparing for transition to secondary school, as the mean ages for the group accessing social/support services was 10 years old, compared to 6 years for the group not accessing these services. Similar findings were reported in Barrett et al., (2014); Floyd and Gallagher, (1997); and Pruchno and McMullen (2004). The last named authors found that the child's demographic characteristics, including age, were able to predict use of social services. However, ethnicity and violent behaviour were also associated with access to social services. The findings of this study are in line with Pruchno and McMullen's (2004) findings except about ethnicity, a variable not investigated in this study because it was not relevant to the aims of this chapter. In line with previous findings, Barron et al., (2013) found that most of the adolescents with intellectual disabilities and challenging behaviour have increased levels of visits in relation to social workers. Additionally, less frequent visit levels were observed in relation to mental health professionals and alternative services. This finding is consistent with Barrett et al., (2014) and with Floyd and Gallagher, (1997). In both studies age was found to be associated with service receipt. An additional explanation of the association between age and social support services is that families of children with intellectual disabilities and challenging behaviour are concerned about the effective transition of their child to secondary school and the additional support that can be offered. Challenging behaviour is strongly associated with age (Baghdadli, Pascal, Grisi and Aussillux, (2003); Baghdadli et al., (2008); Hatton, Hooper, Bailey, Skinner, Sullivan and Wheeler, (2002);

Taylor, Oliver and Murphy, (2011)). Most notably, it has been found that challenging behaviour increases gradually from the age of 10 to the age of 19 years (see Results section 2.2.1 in Chapter 2). Therefore it is to be assumed that families of children with intellectual disabilities and challenging behaviour will use social/support services more frequently as their children grow into adolescence, because challenging behaviour will increase gradually during that period.

CHAPTER 6

General Discussion

6.1. Preface

Chapter 5 described the factors that are associated with service use for families of children with intellectual disabilities and challenging behaviour. More specifically, it highlighted the point that access to advocacy services differs for individuals who engage in aggression or present property destruction behaviour. Importantly, the study also emphasised that diminished parental emotional wellbeing status was associated with access to mental health services for families of children with intellectual disabilities and challenging behaviour, suggesting the existence of an interrelationship between challenging behaviour, parental wellbeing and service use. In this chapter, the results from the empirical studies of the thesis will be discussed and synthesised with existing literature, with a view to developing a broader understanding of the links between challenging behaviour, parental emotional wellbeing, child quality of life and service use.

6.2. Introduction

People with intellectual disabilities comprise about 2% of the UK population (Emerson et al., 2010). Individuals with intellectual disabilities face numerous difficulties across their lifespan. More specifically, health, mental health and behavioural problems are prevalent among individuals with intellectual disabilities (Straetmans, van Schrojenstein Lantman-de, Schellevis & Dinant, 2007; Allerton, Welch & Emerson, 2011; Cooper et al., 2007; Emerson, 2000; Rojahn and Esbensen, 2002; see also Section 1.2).

There is a line of research showing a high prevalence of challenging behaviour in individuals with intellectual disabilities. The evidence presented in Chapter 1 aimed to delineate the area of the manifestation of behavioural problems among individuals with intellectual disabilities, and of challenging behaviour in particular. More specifically, it was shown that 10-15% of individuals with ID exhibit challenging behaviour (Emerson et al., 2001; see also Section 1.2). In particular, self-injury, as the most common form of challenging behaviour, along with aggression, has been shown to be displayed by between 4 and 12% of individuals with intellectual disability of heterogeneous aetiology (Cohen et al*.,* 2010; Cooper et al*.,* 2009; Emerson et al*.,* 1997; Holden & Gitlesen, 2006; Lowe et al*.,* 2007; Oliver, Murphy & Corbett, 1987). Interestingly, the prevalence of aggressive behaviour among individuals with genetic disorders has been found to be 50% in individuals with particular genetic syndromes such as Smith-Magenis syndrome.

Furthermore, as reviewed in Chapter 1, there is evidence supporting the existence of a number of individual characteristics that have been described as risk markers for challenging behaviour (McClintock et al., 2003). More specifically, the associations between particular demographic characteristics, child behaviours, additional diagnosis of a genetic syndrome or of ASD and challenging behaviour have been shown (see Section 1.5.4). In addition, the relationship between self-injurious behaviour and pain has been previously described (Carr & Owen-Deschryver, 2007; Carr, Smith, Giacin, Whelan & Pancari, 2003; Christensen et al., 2009; O’ Reilly, 1997; see also Section 1.5). The consequences of challenging behaviour are known to be pervasive and damaging, influencing the quality of life (Beadle-Brown, Murphy &DiTerlizzi, 2009), care provision and practices (Allen, Lowe, Brophy & Moore, 2009; McGill et al., 2009), as well as the quality of life of carers and families (Hastings, 2003; Totsika, Hastings, Emerson, Lancaster & Berridge, 2011; Seltzer, et al.,2010). Detailed investigation in this area of inquiry may shed extra light on the parameters that are involved in the manifestation of challenging behaviour and the broader impact of such behaviour on the individual and family.

Furthermore, the literature review conducted in Chapter 1 has further delineated areas that relate to the prevalence of challenging behaviour, and the evidence that suggests associations between challenging behaviour and personal characteristics, such as age, gender, ability level, and behavioural correlates of different types of challenging behaviour (see Section 1.5). Taken overall, the investigation of these areas may be critical in understanding the nature and manifestation and the widespread impact of different types of challenging behaviour in these populations, and then to inform policy and practice.

In line with this view, the broader impact of challenging behaviour in individual and family functioning has been investigated in Chapter 1 (see Sections 1.6 and 1.8). Part of this analysis has shown the persistence of challenging behaviour among individuals with intellectual disabilities, as well as the association between challenging behaviour and parental emotional wellbeing. The aggregate of this evidence highlights the broader critical impact of challenging behaviour and stresses the implementation of efforts based on such evidence to provide targeted support to the individuals and families affected.

However, as highlighted in Chapter 1, there is very limited research investigating the persistence of challenging behaviour in individuals with genetic syndromes who are at high risk of developing self-injury or aggression behaviours. Moreover, although the link between challenging behaviour and parental emotional distress has been investigated previously, it is not yet clear whether challenging behaviour itself, or behavioural correlates of challenging behaviour, can be associated with parental emotional distress. In addition, although there is evidence about the diminished quality of life of individuals with intellectual disabilities, compared to those without disabilities, the literature review conducted in Chapter 1 highlighted that there is currently limited evidence about the relationship between challenging behaviour, parental emotional wellbeing and children's quality of life.

In the light of the review of the above areas of inquiry, the broad aim of this thesis was to utilise a multi-method approach to detail the relationship between challenging behaviour, associated personal characteristics, the quality of life of parents and children with intellectual disabilities, and service use in multiple samples of individuals with intellectual disabilities. The results of these studies would then contribute towards a more comprehensive model describing the interrelationship between challenging behaviour, parental emotional distress, children's quality of life and service use. To this end, four empirical studies were conducted. Chapters 3, 4 and 5 reported on survey methodologies employed to assess comparatively large populations of individuals with intellectual disabilities. These studies generated novel evidence on the relationship between challenging behaviour, parental emotional distress, children's quality of life, and service use in individuals with intellectual disabilities. In addition, in Chapter 2 a longitudinal analysis was used to investigate the persistence of self-injurious and aggressive behaviour among individuals with FXS, and also to identify which of the behavioural correlates of challenging behaviour which are associated with the presence of challenging behaviour can also be related to the persistence of challenging behaviour. The findings, strengths, limitations and clinical implications of this research will be discussed below, with reference to the existing literature.

6.3. Main Findings

Given the broad aims of this thesis and the diverse range of methods employed, the key results and implications can be considered most usefully within four domains:

(1) The investigation of the persistence of self-injurious and aggressive behaviour and the associated behavioural correlates of challenging behaviour

(2) The delineation of whether challenging behaviour or behavioural correlates of challenging behaviour may be associated with parental emotional distress

(3) The investigation of the interrelationship between challenging behaviour, parental emotional distress and the child's quality of life and

(4) The investigation of service use and the factors that are associated with service use for families of children with intellectual disabilities and challenging behaviour.

6.3.1. Persistence of self-injurious and aggressive behaviour among individuals with FXS

6.3.1.1. Prevalence and persistence of self-injurious and aggressive behaviour in FXS

A key aim of this thesis was to generate a robust and reliable estimate of the prevalence of self-injurious and aggressive behaviour in individuals with FXS. Previous research about the prevalence of self-injurious and aggressive behaviour among individuals with FXS found that almost 50% of individuals with FXS showed self-injurious behaviour (Arron et al., 2011; Richards et al., 2012). Similar prevalence rates have been reported for aggressive behaviour (Arron et al., 2011). Consistently with previous findings, the results of the study presented in Chapter 2 found that 49.4% of individuals with FXS displayed self-injurious behaviour and 40.5% of individuals with FXS exhibited aggressive behaviour. Moreover, the longitudinal study reported in Chapter 2 investigated the behavioural correlates of challenging behaviour and identified strong associations between behavioural correlates and the persistence of self-injurious and aggressive behaviour among individuals with FXS. More specifically, it has already been found that ASD-types of behaviours were strongly associated with the presence of challenging behaviour rather than an ASD diagnosis (Baumgardner, Reiss, Freund & Abrams, 1995). It was important to investigate the persistence of self-injurious and aggressive behaviour in a single genetic syndrome group, in which self-injury and aggression is highly prevalent, as the effect of the heterogeneity of challenging behaviour prevalence in varied genetic syndromes is controlled. In addition, the documented high prevalence of ASD-types of behaviours among individuals with FXS allowed the investigation of whether behavioural correlates of challenging behaviour can be associated with the persistence of self-injurious and aggressive behaviour among individuals with FXS.

The findings of the study that relate to the persistence of self-injurious and aggressive behaviour among individuals with FXS revealed that self-injury and aggression tended to be persistent over 8 years later than the initial assessment (i.e. 2003 to 2011). Moreover, the study reported that repetitive, restricted and stereotyped behaviour was associated with the persistence of self-injurious behaviour in individuals with FXS. Furthermore, persistent aggressive behaviour among individuals with FXS was found to be associated with impulsivity and over-activity. This pattern of findings is consistent with previous evidence suggesting that it is ASD-types of behaviour rather than an ASD diagnosis which is associated with challenging behaviour (Collacott et al., 1998; Lowe et al., 2007). Lowe et al., (2007) and Murphy et al., (2005) conducted total population surveys and found that self-injury was significantly associated with the triad of impairments of ASD. The very large populations from which these samples were obtained (total populations of 1.2 million and almost 35 thousand respectively) strengthen the findings and indicate a robust association between the presence of ASD phenomena and self-injury. However, given the sample sizes assessed in these studies, it was not possible to conduct individual assessments in which ASD diagnosis was confirmed. Consequently, many of the studies relied upon informant reports using measures such as the Disability Assessment Schedule (DAS; e.g., Collacott et al.*,* 1998; Lowe et al.*,* 2007). Thus, the results imply that the presence of ASD-type behaviours, rather than a clinical diagnosis of ASD, is associated with self-injury.

The results suggest that ASD phenomenology may be a useful putative risk marker for self-injurious behaviour within multiple populations. Supporting this assertion, Arron et al. (2011) used the Social Communication Questionnaire to demonstrate that for individuals with Cornelia de Lange, fragile X, Prader-Willi and Lowe syndromes, specific areas of the triad of impairments were associated with self-injury. Taken together, this research evidence demonstrates that it is the presence of ASD phenomenology, rather than a diagnosis of idiopathic autism per se, that is associated with the presence of self-injury.

Moreover, it has been revealed that ASD-types of behaviour are associated not only with the presence of self-injurious and aggressive behaviour among individuals with FXS, but are also associated with the persistence of those behaviours in this syndrome group. Most notably, it has been found that impulsivity and over-activity, which are ASD-types of behaviours, are strongly related to the presence of challenging behaviour amongst individuals with FXS. This finding is consistent with (Collacott et al., 1998; Lowe et al., 2007) outcomes about ASD-types of behaviour. In summary, the significance of this finding lies in the notion that not ASD diagnosis itself but ASD-types of behaviour are associated with the presence of challenging behaviour.

Moreover, the study found that impulsivity and over-activity scores differ significantly between individuals with FXS who show persistent aggressive behaviour and those who show an absence of aggressive behaviour. Individuals with FXS who show persistent self-injurious behaviour differ significantly in repetitive, restricted and stereotyped behaviours from those who show an absence of self-injurious behaviour.

The findings of the first empirical study of the present thesis support the view that the behavioural correlates of challenging behaviour, which have been previously found to be associated with the presence of challenging behaviour in individuals with intellectual disabilities, are also associated with the persistence of self-injurious and aggressive behaviour in individuals who are at high risk of developing self-injurious or aggressive behaviour. Moreover, these findings further support the view that ASD-types of behaviour rather than ASD diagnoses are associated with challenging behaviour. Given the high prevalence of ASD diagnosis among study participants with FXS, these findings extend the concept of ASD diagnosis as a risk marker for self-injury and aggression, and suggest that it is the cumulative presence of ASD behaviours, rather than idiopathic autism per se, that is associated with self-injury and aggression in individuals with FXS. These findings can be interpreted in the light of operant theories regarding the communicative function of self-injury and the development of self-injury from repetitive behaviours (Guess & Carr, 1991). An increased risk of self-injury, associated with an increase in ASD-types of repetitive behaviours, has face validity if self-injury is seen to develop from repetitive and stereotyped behaviour (Guess & Carr, 1991; Petty, Oliver & Allen, 2009). In summary, examining the association between ASD-types of behaviour and challenging behaviour in individuals with

FXS informs intellectual disability research, practice and policy. Most notably, the longitudinal investigation of the relationship between ASD-types of behaviours and challenging behaviour in relation to various genetic syndromes will provide an insight as to whether ASD-types of behaviour can predict the persistence of such behaviours amongst individuals with ID. In addition, the evidenced relationship between ASD-types of behaviours and challenging behaviour can inform practice further, to design and deliver interventions which will aim to reduce ASD-types of behaviours in individuals with ID, because this will affect the presence of challenging behaviour. The early identification of the relationship between ASD-types of behaviours and challenging behaviour itself, in individuals with ID, will enhance the provision of early and effective interventions in this population.

6.3.1.2. Summary of the persistence and behavioural correlates of challenging behaviour

In summary, the findings of the longitudinal study presented in Chapter 2 suggest that self-injurious and aggressive behaviours tend to be persistent over the years among individuals who are at high risk for displaying challenging behaviour. In addition, the study provides evidence showing that the persistence of self-injurious and aggressive behaviour is associated with ASD-types of behaviours. Given the persistent nature of challenging behaviour in individuals with genetic syndromes who are at high risk for self-injury and/or aggression, it is possible that these individuals will experience a diminished quality of life compared to individuals not displaying challenging behaviour; and their parents will be at higher risk for mental health problems compared to carers of children not displaying challenging behaviour. Therefore, a study investigating this interrelationship was conducted and reported in Chapter 3.

6.3.2. Challenging behaviour, parental emotional distress and children's quality of life in individuals with intellectual disabilities.

A second key aim of this thesis was to investigate the relationship between challenging behaviour, parental emotional distress and children's quality of life in children with intellectual disabilities. The investigation in this area of inquiry has further delineated the behavioural correlates of challenging behaviour that are associated with the presence of challenging behaviour, as well as the relationship between challenging behaviour itself, or the behavioural correlates of challenging behaviour, and parental emotional wellbeing. In addition, the study has contributed to further exploring the quality of life of children with and without challenging behaviour and the interrelationship between challenging behaviour, parental emotional distress and children's quality of life.

6.3.3. Behavioural correlates of challenging behaviour

In the study presented in Chapter 3 it was found that individuals with challenging behaviour and those without challenging behaviour differ significantly in repetitive, restricted and stereotyped behaviours, impulsivity, over-activity and pain. Importantly, the results from the regression analysis showed that only impulsivity and pain are strongly associated with the presence of challenging behaviour. These findings are consistent with previous reports suggesting that ASD-types of behaviour are associated with the presence of challenging behaviour (Richards et al., 2012; Richman et al., 2013). In addition, previous research found pain to be strongly associated with the presence of challenging behaviour (Berg et al., 2007; Luzzani et al., 2003; Breau et al., 2002). Most notably, results from Berg, Arron, Burbidge, Moss and Oliver, (2007) suggest that the correlation between low mood and self-injury may be accounted for by health problems, which are undetected or unresolved. In this case, both self-injury and low mood are a result of pain experiences and, consequently, co-occur. Consistently with this line of research, the results of the studies presented in Chapters 2 and 3 demonstrated that behavioural correlates of challenging behaviour are associated with challenging behaviour. Moreover, the evidence presented confirmed that it is not ASD diagnosis but ASD-types of behaviours that are associated with challenging behaviour. Importantly, a strong association was evident between challenging behaviour and parental emotional distress. In the light of this evidence, it was deemed appropriate to investigate whether challenging behaviour itself or the behavioural correlates of challenging behaviour, were associated with parental anxiety and/or depression.

6.3.4. Parental emotional distress and challenging behaviour

The relationship between challenging behaviour and parental emotional distress has been extensively described in Chapter 1 (see Section 1.8.1). Most notably, there is evidence supporting the view that parents of children with challenging behaviour are at a higher risk of developing mental health issues compared to parents of children without challenging behaviour (Hastings, 2003; Hastings & Brown, 2002; Baxter, Cummins &Yiolitis, 2000). A further explanation about this relationship has emerged through the proposed model of the relationship between challenging behaviour and parental emotional wellbeing in Hastings, (2002). Most notably, it has been proposed that children's behaviour problems are assumed to influence parental stress, which in turn influences parental behaviour, which then influences child behaviour. However, in Hastings, (2002) the term describing challenging behaviour which was employed was focused on a range of behaviours which might be described as challenging for their parents. Thus, this term was not focused only on self-injurious and aggressive behaviour, but also included other behaviours, such as impulsivity, over-activity, and repetitive, restricted and stereotyped behaviours, which are behavioural correlates of challenging behaviour.

The findings of that study support the view that parents of children with and without challenging behaviour differ significantly in anxiety, depression and emotional distress. Emotional distress was an additional variable describing parents who met the cut-off criteria either for anxiety or depression. Most notably, this characteristic was in line with previous findings suggesting that parental emotional distress is associated with challenging behaviour. The percentage of parents who met the cut-off criteria for anxiety, depression and emotional distress was significantly higher in parents of children with challenging behaviour compared to parents of children without challenging behaviour. This finding supports further Hastings, (2002) findings that challenging behaviour rather than intellectual disability is associated with parental emotional wellbeing. In addition, it has been found that high scores on impulsivity, overactivity, repetitive, restricted and stereotyped behaviours and pain were significantly associated with parental emotional wellbeing. To test whether parental emotional distress and challenging behaviour or behavioural correlates of challenging behaviour are associated further analysis had to be conducted. It was found that impulsivity rather challenging behaviour itself is strongly associated with parental emotional distress. This finding supports the view that it is not challenging behaviour itself but impulsivity, a behavioural correlate of challenging behaviour, that is associated with parental emotional wellbeing. Although Hastings, (2002) proposed that challenging behaviour is associated with parental emotional wellbeing, this finding is controversial. However, one needs to take into account the term 'challenging behaviour' which, as employed in Hastings, (2002), includes impulsivity as challenging behaviour. The relationship between parental emotional distress and behavioural correlates of challenging behaviour was reviewed in (Beckman, 1983; Gabriels, Cuccaro, Hill, Iversand, and Goldson, 2005). However, in these studies the relationship between behavioural correlates and parental emotional distress was investigated in parents of children with ASD. In the current study, parents of children with intellectual disabilities, whose children scored high on impulsivity, experienced greater anxiety, depression and emotional distress compared to parents of children who did not score high on impulsivity.

In sum, the association between behavioural correlates of challenging behaviour, parental emotional distress and challenging behaviour has been explored. Most notably, ASD-types of behaviours are not only strongly associated with the presence of challenging behaviour in individuals with genetic disorders and ID, but are also associated with increased parental emotional distress. Moreover, for the first time it has been found that ASD-types of behaviour (impulsivity) can predict parental emotional distress rather than challenging behaviour being the predictor. This finding is significant in terms of both practice and research. It is essential to achieve early interventions which will aim to reduce ASD-types of behaviours in order to reduce challenging behaviour and to enable parental mental health to be stable. In addition, a further examination of the interrelationship between parental mental health, challenging behaviour and ASD-types of behaviour can provide research evidence as to whether by reducing ASD-types of behaviour the risk of children with ID developing challenging behaviour can be eliminated; and also whether this can prevent parents experiencing emotional distress. In the following section, the quality of life of children with

ID and challenging behaviour as investigated is described, as well as whether these elements are associated with child wellbeing.

6.3.5. Exploring the quality of life of children with intellectual disabilities and challenging behaviour

The study presented in Chapter 4 aimed to investigate the quality of life of children with intellectual disabilities and challenging behaviour. Previous evidence has highlighted the point that children with intellectual disabilities experience a diminished quality of life compared to children without intellectual disabilities (Arnaud et al., 2010; Varni et al., 2005). In these studies it has been suggested that personal characteristics, such as the severity of intellectual disabilities, and pain, are related to the quality of life of children (Williams et al., 2003). However, there was previously no evidence on whether there is a close relationship between challenging behaviour and the child's quality of life. The study conducted and reported in Chapter 4 found that children's quality of life is not only associated with challenging behaviour but is also associated with behavioural correlates of challenging behaviour and with parental emotional distress. Taking this evidence overall, it is proposed that an interrelationship exists between parental emotional wellbeing, children's quality of life and challenging behaviour. Given the relationship between impulsivity and parental emotional distress, further analysis was conducted to investigate the relationship between parental emotional wellbeing, impulsivity and children's quality of life. Findings suggest that high impulsivity scores are associated with emotional distress and diminished family wellbeing. In addition, parental emotional distress was found to be associated with emotional and social wellbeing. However, no interaction between parental emotional distress, impulsivity and children's quality of life was observed. The interrelationship between challenging behaviour, parental emotional distress, children's quality of life and behavioural correlates of challenging behaviour has been investigated for the first time.

These findings can therefore inform further research and practice. Most notably, further research, examining the relationship between parental emotional distress, challenging behaviour itself and behavioural correlates of challenging behaviour, should be conducted in order to address whether challenging behaviour itself or the behaviours correlated with challenging behaviour are associated with increased parental anxiety and/or depression. Furthermore, the findings about the quality of life of children with challenging behaviour highlight a strong relationship between parental anxiety, behavioural correlates of challenging behaviour and a diminished quality of life of children. Investigating this interrelationship is significant for practice because it highlights the importance of early and targeted interventions and service provision to children who are at high risk of developing challenging behaviour and to their families, in order to enhance the quality of life of children with ID.

6.3.6. Description of service use for families of children with intellectual disabilities and challenging behaviour

Previous studies have highlighted the point that the presence of challenging behaviour may be associated with increased access to health services (Joyce, Ditchfield & Harris, 2001; Ruddick, Bacarese-Hamilton, Davies and Oliver, 2015; Allen, Lowe, Brophy and Moore, 2009). In the last empirical study conducted in the present thesis (Chapter 5), a detailed investigation was conducted to describe the service use of families of children with intellectual disabilities and challenging behaviour. The study's results suggested that allied health professionals and medical services are the services most used among families of children with challenging behaviour. In addition, consistently with previous reports, alternative therapists were the least accessed services among families of children with challenging behaviour (Felce et al., 1998; Knapp et al., 2005). A plausible explanation of why families of children with intellectual disabilities and challenging behaviour accessed health services was revealed from the link between pain and challenging behaviour (see Section 1.6.3 in Chapter 1; Section 3.1.3 in Chapter 3). Challenging behaviour is strongly associated with high impulsivity scores and health problems. In addition, a relationship between child health problems and maternal stress in mothers of children receiving special education services was found by Mitchell and Hauser-Cram, (2008). However, no significant relationship between parental emotional distress and the number of health problems was observed in the Mitchell and Hauser-Cram, (2008) study. However, the inability of individuals with profound disabilities to communicate their pain and the easy access to medical professionals by parents of children with ID might explain why parental emotional distress and children's health problems are not related. Moreover, the present study has investigated whether the number of health problems is related to challenging behaviour and parental stress rather than to pain problems. Given the relationship between pain and challenging behaviour, it is assumed that families of children with challenging behaviour are more likely to have increased access to medical services.

6.3.7. Exploring the effectiveness and the usefulness of the services used

Taking into account the evidence from the earlier part of the study, describing the high percentage of medical services used by families of children with challenging behaviour, it was deemed necessary to evaluate the usefulness and the effectiveness of such services, according to parental reports. The study utilised parent reports on the effectiveness of services, and found that allied health professionals, and social and alternative therapists were rated highly in terms of their usefulness by parents of children with intellectual disabilities and challenging behaviour. Conversely, lower rates on the usefulness of services were reported for mental health professionals. These findings were in line with previous findings where parental satisfaction with the usefulness and the effectiveness of the services used has been investigated (Hare, Part, Burton, Bromley & Emerson, 2004; Bromley et al., 2004; Mitchell and Hauser-Cram, 2006; Hatton et al., 2000). In these studies, the usefulness of the medical services used, based on parental reports, was investigated in relation to individuals with intellectual disabilities. Higher rates of satisfaction about the usefulness and the effectiveness of the alternative therapists and social services were reported in the above studies. Lower rates about the usefulness and the effectiveness of mental health services, and particularly of psychiatrists, were reported in the above studies. Most notably, findings about the usefulness and the effectiveness of the alternative therapists, which were reported in Hatton et al., (2000), were consistent with the finding in the present study that occupational therapy and speech therapy, as well as improved communication with the involved professionals, were listed as desirable improvements by parents of children with FXS. It is notable that in the current study families of children with ID and challenging behaviour took part, enabling an assessment of the effectiveness and the usefulness of the services provided. It has been found that challenging behaviour is strongly associated with parental emotional distress and children's quality of life. Therefore, parents might have tried to increase their use of mental health services and of health professionals to help them to eliminate challenging behaviour.

In summary, in this study the usefulness and the effectiveness of the accessed services were investigated, based on parental reports. However, to describe the service use for families of children with intellectual disabilities and challenging behaviour it was necessary to investigate the factors which are associated with service use in families of children with challenging behaviour. Investigating the factors which are associated with service use for families of children with challenging behaviour will be informative for policy, practitioners and research. Most notably, investigating the factors that are associated with service use will enable policy providers to increase the use made by families of particular services, and to assess the value of services which are used more often by families of children with challenging behaviour. In addition, researchers can investigate further whether the appropriate service provision is related to parental emotional distress and children's quality of life.

6.3.8. Differences in terms of service use between individuals who show self-injury, aggression and property destruction

The final part of the present study aimed to assess the relationship between particular factors, such as the child's age, gender, quality of life, parental anxiety, depression, family socio-economic status and the child's health problems and service use. However, to investigate this relationship the differences were analysed in respect of service use between individuals who showed self-injury, aggression and property destruction and those who did not. It was found that individuals with and without self-injurious behaviour did not differ in service use. However, individuals exhibiting aggressive behaviour or property destruction differed significantly from those not exhibiting aggressive behaviour or property destruction in terms of using advocacy services. However, advocacy services in this study include social and support workers, who are involved in the child's placement, or can mediate for particular intervention programmes to be delivered by the appropriate professionals to those who ask for their help. Similar findings were reported in Pruchno and McMullen, (2004) and in White and Hastings, (2004). These studies found that children's aggressive behaviour was associated with increased access to psychology services. These findings suggest that aggressive behaviour affects the safety of others, and carers need additional and immediate support to effectively manage children's aggressive behaviour. In addition, self-injurious behaviour impacts significantly on an individual's quality of life, but might not place others in danger for their physical safety. Therefore, significant differences in service use for families of children with self-injurious behaviour were not observed. No significant differences were observed in using the services of educational, medical, alternative and allied health professionals between the families of children with and without self-injury, or displaying aggression and property destruction.

6.3.9. Factors which are associated with service use among families of children with challenging behaviour

The last aim of the present study was to investigate the relationship between the child's age, gender, parental anxiety, depression, socio-economic status, the child's health problems and the child's quality of life and service use. There is evidence in the literature describing the association between the above factors and increased access in various services (Knapp et al., 2005; Floyd and Gallagher, 1997; Barron et al., 2013; Barrett et al., 2014 and Doran et al., 2012). In this study, it was found that parents who met the cut-off criteria for anxiety differed significantly from those who did not, in use of mental health services. In addition, families of children with challenging behaviour who used social services were significantly older compared to families of children who did not use social services. No significant differences were reported when comparing families of children with challenging behaviour who had used educational, medical, alternative and allied health services with those who did not.

To investigate which of the above factors i.e. child's health status, self-help ability, age, parental emotional wellbeing, family socio-economic status and child's quality of life were associated with service use further analysis was conducted. It was found that parental anxiety can predict access to mental health services, and child's age could predict access to social services. No significant associations were observed between the child's quality of life, self-help, parental depression, socio-economic status or the child's health problems and service use. The findings which suggested that parental anxiety was strongly associated with use of mental health services were reported in Floyd and Gallagher, (1997) and in White and Hastings, (2004). This is related to the findings in Chapter 3, which suggest that challenging behaviour and the child's behavioural correlates of challenging behaviour are associated with parental emotional distress. In addition, there is evidence supporting the association between the individual's age and use of social services (Barrett et al., 2014; Floyd and Gallagher, 1997 and Pruchno and McMullen, 2004). In the present study it was found that the child’s demographic characteristics, including age, were predictive of use of social services. This finding suggests that both the frequency and the intensity of challenging behaviour are associated with the child’s age; and therefore as their children become older parents will make increased use of social services to support them, in terms of placement transition and intervention programme delivery. In addition, this finding is consistent with the findings reported in Chapter 2, where the persistent nature of self-injurious and aggressive behaviour among individuals with FXS was investigated. It was found that the highest percentage of individuals with FXS who engaged in both self-injury and aggression were aged up to 19 years old. The relationship between age and challenging behaviour was reviewed in many studies (Baghdadli, Pascal, Grisi and Aussillux, 2003; Baghdadli et al., 2008; Hatton, Hooper, Bailey, Skinner, Sullivan and Wheeler, 2002; Taylor, Oliver and Murphy, 2011). However, no significant relationships between the remaining proposed factors and service use were observed in this study. In summary, in this study it was found that parental anxiety is associated with use of mental health services among families of children with challenging behaviour, and the child's age is strongly associated with the use of social services.

6.4. Limitations and Strengths of the Research

Whilst the findings in this thesis are novel, a number of limitations to the conclusions and hypothesised model must be acknowledged. First, Chapter 2 reported where the persistence of self-injurious and aggressive behaviour among individuals with FXS was investigated. In this longitudinal study data about participants' behaviour, individual characteristics and self-help skills came from previous studies which have been taken place within the Cerebra Centre for Neurodevelopmental Disorders at University of Birmingham, and undertaken by my colleagues rather than directly by me. Although the measures used are standardised participants had not completed the measures at the time point when the present study took place. The use of existing data was acknowledged in all the empirical studies in which data about individuals' behaviour, family socio-economic status and individual self-help skills were analysed and discussed. The advantages and disadvantages of using existing datasets have been described in depth in chapter 1 (Section 1.10). The main advantage of longitudinal studies involves the use and collection of data covering long periods of time, which can determine overall patterns efficiently. Secondly, longitudinal studies, with clear research objectives and methods, allow the researchers to investigate particular patterns over time, helping to ensure the validity of these studies. The validity of the results can also be assisted through the examination of a particular sample which is very effective in undertaking research on developmental trends. Finally, such studies are highly flexible and can provide high degrees of accuracy when observing changes. The main disadvantages include the amount of time which researchers need to devote to selecting and analysing longitudinal data. Another limitation is the risk of gathering data which are not reliable for the time period that the longitudinal analysis is taking place in. Finally, an adequate sample size is needed to perform rigorous longitudinal analyses of the data.

Because of the small number of participants, predictions about the persistent nature of both self-injurious and aggressive behaviour could not be investigated: rather, associations between behavioural correlates of challenging behaviour and the persistence of challenging behaviour in individuals with FXS were assessed. In addition, the association between age and challenging behaviour was explored descriptively rather than by conducting further analysis. Therefore, the results were suggestive, that higher numbers of participants who engaged in both self-injury and aggression were aged up to 19 years old, rather than this relationship being investigated and confirmed statistically.

Second, in Chapter 3, although robust measures about the child's behavioural profile and parental emotional wellbeing were employed, a modified version was employed of the quality of life measure of children with intellectual disabilities, with and without challenging behaviour . Although the quality of life of children with disabilities has been explored in many studies (see Section 1.8.2) there are limited numbers of standardised measures of the quality of life of children with disabilities. The most appropriate measure was the Kiddy-KINDL, but this measure had sub-domains, such as self-esteem, which were not applicable for the children with profound disabilities who took part in this present study. Therefore, this sub-domain was removed from the measure and a scale, with additional items which were proposed in the initial measure was adopted, measuring social wellbeing. This scale was therefore an exploratory tool about the quality of life of children with intellectual disabilities and challenging behaviour, and further investigation is necessary in order to confirm the above findings.

Third, in Chapter 4, service use for families of children with intellectual disabilities and challenging behaviour is described. In that part of the present study, the Client Service Receipt Inventory was employed to describe the services actually used rather than investigating the services that families would like to have used. Therefore, these results are in relation to the services that families have used and do not necessarily reflect services that they might have been interested in using. Moreover, although the descriptive percentages varied between the highly used services and the less used services among parents of children with challenging behaviour, because of the absence of a control group of families whose children did not exhibit challenging behaviour it was not possible to investigate whether the extent of challenging behaviour is associated with the variation in relation to services used.

Fourth, in Chapter 5, factors associated with service use among families of children with challenging behaviour were investigated. However, further investigations of this association couldn't be carried out because of the absence of a control group of families of children with challenging behaviour. In addition, the measure about service use which has been utilised in this study has also been utilised in other studies measuring the economic aspects of service use. However, in this study information about the economic perspective of the services that they were using have not been collected. In addition, data about challenging behaviour had been collected in the past before this study took place. This might be a limitation, because parents were not completing measures about the behavioural profile of their children at the time that this study was taking place. Another limitation in this study might be regarding the socio-economic status of families who participated in the study. Families who completed the online survey had a similar socio-economic status and there was no great variation to allow investigation of possible differences in service use.

Throughout the research for this thesis, a sample of families of children with various genetic syndromes was studied, in order to increase the generalisability of the findings among individuals with intellectual disabilities. Moreover, various standardised measures were used in order to investigate whether challenging behaviour itself or behavioural correlates of challenging behaviour were associated with parental stress. In addition, novel findings about the relationship between challenging behaviour and parental emotional distress were supported in that part of the study, suggesting that rather than challenging behaviour itself, ASD-types of behaviour are strongly related to parental emotional wellbeing. Moreover, in this thesis and for the first time the interrelationship between challenging behaviour, parental emotional distress and children's quality of life was explored, suggesting that these three strands are related.

6.5. Future directions

As a result of the research in this thesis a number of key areas for future research can be identified. Firstly, further longitudinal analysis of the persistence of self-injurious and aggressive behaviour needs to be undertaken. This would be to assess which personal characteristics are associated with the persistence of self-injurious and aggressive behaviour in individuals with various genetic syndromes who are at high risk for self-injury and aggression. This would include individuals have scored high in ASD-types of behaviour, in order to evaluate such behaviours as risk markers for the presence and the persistence of challenging behaviour in individuals with intellectual disabilities. In addition, it is important that future research is conducted clinically, to investigate the association between particular behavioural correlates and parental emotional wellbeing. It is important to evaluate the relationships between challenging behaviour, behavioural correlates of challenging behaviour and parental anxiety or depression. Moreover, as in this thesis the quality of life of children with intellectual disabilities and challenging behaviour has been investigated, so it is essential for a standardised measure to be developed, which will be applicable to and appropriate for individuals with profound intellectual disabilities. It would then be necessary for this to be employed in future studies to investigate the quality of life of children with intellectual disabilities and challenging behaviour. Finally, a further study, is necessary which will examine the association between socio-economic status, parental mental health, and the child's challenging behaviour, but a study which will also employ a control group of families of children without challenging behaviour. This is necessary to investigate the factors that are associated with higher levels of service use, comparing families of children with and without challenging behaviour. In addition, further studies are necessary to investigate not only the services that are used but also the services that families want to use , in order to evaluate the unmet needs of these families.

6.6. Closing summary

"Research is to see what everybody else has seen, and to think what nobody else has thought". (Albert Szent-Gyorgyi).

As this quote suggests, research has a tendency to create novel and interesting questions exponentially. However, whilst this thesis has generated many new research ideas, it has also given findings providing suggestive answers to some critical questions about the persistence of self-injurious and aggressive behaviour among individuals with FXS, the association between particular behavioural correlates of challenging behaviour and parental emotional wellbeing, and has also investigated the interrelationship between challenging behaviour, parental emotional distress and children's quality of life in children with intellectual disabilities, with and without challenging behaviour. These findings highlight the importance of assessing challenging behaviour as a social construct which is associated with parental emotional wellbeing and challenging behaviour. Moreover, tailored interventions are necessary, focusing both on the individual's needs and on parental emotional wellbeing. Further, it is important for early clinical interventions to be provided

addressing the persistence of challenging behaviour in individuals with various genetic disorders, who are at high risk for displaying challenging behaviour, or score highly in ASD-types of behaviour, which were found in this thesis to be strongly associated with the presence of and the persistence of challenging behaviour.

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APPENDICES

APPENDIX A

Initial letter to parents

The Cerebra Centre for
Neurodevelopmental Disorders

UNIVERSITY OF
BIRMINGHAM

[redacted]

Dear <parent/carer>,

We are writing to you from the Cerebra Centre for Neurodevelopmental disorders at the University of Birmingham to let you know about a new questionnaire study that we will be carrying out and we would like to give you the opportunity to participate in.

The project is called ‘Parental Illness Representations of their Childs Genetic Syndrome, Challenging Behaviour and its Impact upon Parental and Child Wellbeing and Service Provision’. This broad study aims to investigate parent’s perceptions of the physical, cognitive and behavioural presentation of their child’s genetic syndrome and how these perceptions affect outcomes for both themselves and the individuals for whom they care. Moreover, this study aims to examine the impact of challenging behaviour on child’s wellbeing, on family functioning and the effectiveness of service provision for those behavioural problems.

We are interested in working with children with neurodevelopmental disorders because previous research has suggested that challenging behaviour set great boundaries in the daily life for both children with neurodevelopmental disabilities and their families. It’s important for us to find out more about this so we can begin to understand the impact of those behaviours on both family and individual’s wellbeing, to estimate the effectiveness and satisfaction of the provided services, to describe parent’s perceptions about child’s abilities and disabilities and finally to investigate how these perceptions affect parents and individuals quality of life.

If you think that you might be interested in taking part in this study, please visit <URL link> to fill out an online version of the questionnaire. The password to access the questionnaires is

(.....). To fill out a paper copy please return the expression of interest form in the stamped addressed envelope provided. [redacted]

Thank you very much for your time,

Kind regards,

Professor Chris Oliver Efthalia Karakatsani Natalie Jackson (Clinical Psychologist) (Research Investigator) (Research Investigator)

Expression of interest in the project

If you think that you and the child you care for may be interested in taking part in the project ‘Parental Illness Representations of their Childs Genetic Syndrome, Challenging Behaviour and its Impact upon Parental and Child Wellbeing and Service Provision’ and would like a paper copy of the questionnaire, complete the form below and return it to us in the stamped addressed envelope provided.

Your name:

The name of the child you care for:

Your address:

Your telephone number:

Day:

Evening:

Your email address:

APPENDIX B

Information sheet

Parental Illness Representations of their Childs Genetic Syndrome, Challenging Behaviour and its Impact Upon Parental and Child Wellbeing and Service Provision:

Information Sheet

Please read this information carefully before deciding whether you wish to take part in the study. [redacted]

If you have any medical/ other problems which make it difficult for you to read this information, please contact with Natalie Jackson or Efthalia Karakatsani for a verbal explanation of the research.

When you are happy that you have all of the information you need to be able to decide whether or not you and the person you care for would like to take part in the study, please complete the enclosed consent form and questionnaire pack return them to us in the prepaid envelope provided

Background

We would like to invite you to take part in a questionnaire study being conducted at the Centre for Neurodevelopmental Disorders, University of Birmingham. This research work, which is led by Professor Chris Oliver, looks at parental illness representations of their child's Genetic Syndrome, Challenging Behaviour and its Impact upon parental and child wellbeing and service provision. We hope that this information will enable us to define the association of challenging behaviour with parental perspectives of child's disability and family wellbeing with service provision.

The more people that take part in this research, the more meaningful the results will be. A good response will provide new and valuable information about challenging behaviour and family wellbeing. In the future we hope to follow up the progress of the people who take part in this study. However, participation in this stage of the project will not mean that you are obliged to participate in further surveys in the future.

Aims of the study

To understand how parent's perceptions of the physical, cognitive and behavioural presentation of their child's genetic syndrome impact upon parental wellbeing, quality of life and coping style.

To understand the impact of challenging behaviour on parental stress and child's wellbeing.

To estimate the effectiveness and satisfaction of provided services for challenging behaviour according to parental reports.

What will happen if you and your child/the person you care for decide(s) to participate?

Where will the research take place?

The research will involve completing the enclosed questionnaire pack. This can be completed by you in your own time at your home.

Who will be involved in collecting the data?

Members of the research team at the Cerebra Centre for Neurodevelopmental disorders including Natalie Jackson and Efthalia Karakatsani.

How long will participation in the study take?

The questionnaire pack will take approximately 45 minutes to complete.

In the future you may be asked if you would like to complete the questionnaire again so that we can start to understand what happens to people with challenging behaviour across their lifetime. We will only contact you with this invitation if you have previously agreed to be contacted by the research team at the University of Birmingham with information about research studies conducted by the team.

Sometimes after you have completed the questionnaire, we may need to contact you again in order to clarify any information that you have provided or to ask you for further information regarding the diagnosis of the person you care for. This helps us to ensure that our data is as useful and as accurate as possible. If this happens then we would contact you again within 6 months of receiving your questionnaire pack to ask whether or not you would be willing to provide us with the extra information.

What will participants be required to do during the study?

We will ask parents and caregivers to complete the enclosed questionnaire pack and return it to us alongside the consent form in the pre-paid envelope provided.

Are there any risks that individuals taking part in the study might face?

There will not be any risks associated with participation in this study.

What are the potential benefits for participants from taking part?

You will receive a personalised feedback regarding your child/ the person you care for. This study will help us to find out more about the lives of people with challenging behaviour and the difficulties that these people face. The results might help us to improve things for people with challenging behaviour in the future.

Where will data be stored?

The data collected will be kept in locked or password protected storage at the University of Birmingham. Only members of the research team at the University of Birmingham will have access to information that we collect about you. Information will be treated as strictly confidential and handled in accordance with the provisions of the Data Protection Act 1998.

If you/ the person you care for decide(s) to participate, what will happen after that participation?

You and your child/ person you care for will receive an individual feedback report describing the results of all of the assessments that were carried out during the study. If requested, this feedback report will be circulated to other interested individuals. Descriptions of research findings will be published in newsletters of the relevant family support groups and educational institutions involved. Any request for advice concerning the person you care for will be referred to Professor Chris Oliver, Clinical Psychologist.

The researchers will publish the findings from the study in scientific journals and will present the results at relevant conferences.

What will happen to the data afterwards?

The information that you provide will be locked in a filing cabinet at the University of Birmingham or held on a password protected database. Participants will be identified by a unique number so that the information you provide us with cannot be traced to your personal details. You will be able to decide whether or not you want to make your research data available to any professionals or clinicians working with you and the person you care for should they wish to see it. This is optional and will not affect your participation in the current study. If you agree to this, then your research data will only be made available to relevant clinicians or professionals should they contact us directly and request to see it. If you do not agree to this, then research data will not be made available to anyone other than the research team at the University of Birmingham.

After 6 months of receiving your questionnaire pack, your personal details will be ***destroyed unless you tell us otherwise.*** This means that we would no longer be able to trace the results of your assessments back to you. ***The section below on 'The Regular Participant Database Information'*** gives information about a database that we use to store the personal details of some participants. Please read this section in order to decide if you would like to join that database.

Regular Participant Database Information:

What is the regular participant database?

We have a database that we keep in the Cerebra Centre where we store the names and contact details of some previous participants. If you would like us to, we can add your details to this database. We would use this information for two things:

We will contact you with information about future research work to find out whether or not you would like to participate.

It is often important to find out how things change over time. By keeping your details we would be able to trace the results of the previous assessments that you have done with us back to you. This means that if you take part in other studies with us we would be able to look at how things have changed over time.

Who would have access to my details?

Only approved members of our research team would have access to your details. We would not share your details with anyone outside the research team.

When would I be contacted?

You would only be contacted by an approved member of the research team when we are starting another study or phase of a study that we think you might like to participate in or when we need to clarify some information that you have provided us with from participation in a research study.

What happens if I decide that I want my details to be added to the database but then I change my mind?

[redacted]

Consent

After having read all of the information and having received appropriate responses to any questions that you may have about the study you and the person you care for will be asked to give you and your child’s/ person you care for’s consent to participate in the study if you decide that you do wish to participate. The section below on ***’Giving consent’*** will explain this process. We need to receive consent from/ on behalf of potential participants in order for them to participate.

Withdrawal

Even after consent has been granted, participants can request to be withdrawn from the study at any time, without giving a reason. Even after participation has taken place, consent can be withdrawn and any data collected will be destroyed. This will not restrict the access of you/ the person you care for to other services and will not affect their right to treatment.

What if there is a problem?

[redacted]

Confidentiality

The confidentiality of participants will be ensured. If published, information on the participant will be presented without reference to their name or any other identifying information. All personal details will be kept separately from the information collected so that it will only be possible to connect results to individuals via a special code. This will ensure that results are kept anonymous. In the unlikely event of any evidence of abuse being identified, this information will be disclosed by the research workers.

Review

The study has been approved by the Ethical Review Committee

Further information

[redacted]

Giving consent

Now it is up to you whether you decide that you and your child/the person you care for would like to participate. The decision about whether or not to take part in the study must be 'informed'. This means that anyone making the decision must understand exactly what is involved in the study, what will be required from participants and why.

IMPORTANT:

You need to decide whether your child/the person you care for is able to understand enough about the study to make an 'informed' decision independently about whether or not they would like to participate and to communicate this decision to you. If you are unsure whether or not your child/person you care for is able to understand enough to make a decision independently then we can provide you with some guidelines to help you to assess this A symbol information sheet can also be made available to you if this would be of help.

[redacted]

Please choose from one of the following options:

My child/ the person I care for is able to understand what is involved in the study and what will be required from them if they participate and has communicated their decision to me:

If you think that the person is **able** to understand enough about the study in order to make an 'informed' decision and they decide that they would like to participate then please ensure that they complete **Section 1 of Consent Form A coloured YELLOW** enclosed, or that you complete it with them, on their behalf. A parent/carer will need to complete **Section 2 of Consent Form A coloured YELLOW** in order to indicate that they also agree to participate in the study. *A symbol information sheet can be made available in order to support your child/person you care for in making this decision if it would be of help.* Please contact the research team if you would like a copy of the symbol consent form or if you need us to adapt this information further, in order to suit your child's needs. Please return the consent form along with the questionnaire pack to us in the prepaid envelope provided.

My child/ the person I care for is unable to understand what is involved in the study and what will be required from them if they participate (either because they are too young to understand or because they are unable to understand) and cannot communicate their decision to me:

If you are reading this information on behalf of someone you care for who is under the age of 16 years and you decide that the person ***is not*** able to make an 'informed' and independent decision about whether or not they would like to participate, then we would like to ask you to decide whether or not you think that it is in your child's best interests for them to participate in the study and whether you would like to provide your consent to participation on their behalf. If you would like your child/person you care for to participate in this study, please complete **Consent Form B coloured PURPLE** enclosed. Please return the consent form along with the questionnaire pack to us in the prepaid envelope provided.

APPENDIX C

Consent form A

UNIVERSITY OF BIRMINGHAM

The Cerebra Centre for Neurodevelopmental Disorders

Consent Form A : **For individuals who are able to provide consent to participate in the study**

Parental Illness Representations of their Childs Genetic Syndrome, Challenging Behaviour and its Impact Upon Parental and Child Wellbeing and Service Provision

Study Director: Professor Chris Oliver

SECTION 1: Please complete this section if you are a person with challenging behaviour:

Has somebody else explained the project to you?	YES/NO
Do you understand what the project is about?	YES/NO
Have you asked all of the questions you want?	YES/NO
Have you had your questions answered in a way you understand?	YES/NO
Do you understand it is OK to stop taking part at any time?	YES/NO
Are you happy to take part?	YES/NO

If any answers are 'no' or you do not want to take part, do not sign your name!

If you do want to take part, you can write your name below

You can also choose if you want to say 'yes' to these questions:

If your Dr asks to see your results from this project is that OK?	YES/NO
Are you happy for us to contact you again in the future?	YES/NO

Your name:____

Date:______________________

The person who explained this project to you needs to sign too. If you are under the age of 16, this should be your parent/guardian.

Print name:___________________________ Sign:_________________________
Date:__________________

<u>SECTION 2:</u> Please complete this section if you are a parent/carer/guardian of a person with challenging behaviour who has provided their consent to particip study. Please initial box…

PTO…continued overleaf

I confirm that I have read and understood the information sheet date for the above study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily. ☐

I understand that my participation and that of my child/person I care for is voluntary and that I am free to withdraw at any time without giving any reason, without my or that of my child's/person I care for's medical care or legal rights being affected. ☐

I understand that relevant sections of my child's/person I care for's GP medical notes or records confirming genetic diagnosis and health status may be looked at by members of the Cerebra Centre for Neurodevelopmental Disorders research team at the University of Birmingham, where it is relevant to this research project. I give permission for these individuals to have access to these records. ☐

I agree to my child's/person I care for's GP being informed of my participation and that of my child/person I care for's in the study, where access to my child's/person I care for's medical records is required. ☐

I agree to take part in the above study. ☐

Optional clause: The statement below is <u>optional</u>:

I agree to the University of Birmingham research team sharing my research data with any professionals or clinicians working with me and the person I care for should they request to see them. ☐

Print Name: ___________________________________

Telephone number: ______________________________

Address: ____

Email: ____________________Relationship to participant:________________

Signature: ________________________Date: _____________________

SECTION 3: This is optional and allows you to provide consent for us to keep your personal details on the Regular Participant Database. See section titled 'R Participant Database' in the information sheet.

PTO…continued overleaf

Please initial box…

1. I have read and understood the section titled 'Regular Participant Database' and I would like my personal details to be added to the database. ☐

2. I understand that my name and contact details will be kept by the research team at the University of Birmingham in accordance with the provisions of the Data Protection Act 1998 and I will be contacted by an approved member of the team with information about future research that I and the person I care for may like to participate in. ☐

3. I understand that if my details are held on the database it will be possible for the research team to trace the results of the assessments that I complete in this project back to me and my child/person I care for so that they can look at changes over time if I take part in future projects. ☐

4. I understand that even after I have agreed for my details to be added to the database, I can request that they be removed by contacting Chris Oliver on 0121 414 7206 or at c.oliver@bham.ac.uk or by post at the School of Psychology, University of Birmingham, Edgbaston, B15 2TT. ☐

5. I understand the Professor Chris Oliver holds ultimate responsibility for the database. ☐

Print Name: ________________________

Signature: _________________Date:____________________________

APPENDIX D

Consent Form B

The Cerebra Centre for
Neurodevelopmental Disorders

UNIVERSITY OF
BIRMINGHAM

Consent Form B: For Children under the age of 16 who are not able to provide consent.

Parental Illness Representations of their Childs Genetic Syndrome, Challenging Behaviour and its Impact Upon Parental and Child Wellbeing and Service Provision

Study Director: Professor Chris Oliver

SECTION 1: Please complete this section if you are a parent/ guardian of a child (under 16 years) with challenging behaviour who is not able to provide consent.

Please initial box…

I confirm that I have read and understood the information sheet for the study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily. ☐

I understand that my participation and that of my child/person I care for is voluntary and tha I am free to withdraw at any time without giving any reason, without my or that of my child's/person I care for's medical care or legal rights being affected. ☐

I understand that relevant sections of my child's/person I care for's GP medical notes or records confirming genetic diagnosis and health status may be looked at by members of the Cerebra Centre for Neurodevelopmental Disorders research team at the University of Birmingham, where it is relevant to this research project. I give permission for these individuals to have access to these records. ☐

I agree to my child's/person I care for's GP being informed of my participation and that of my child/person I care for's in the study, where access to my child's/person I care for's medical records is required. ☐

I agree to take part in the study. ☐

Optional clause: The statement below is *<u>optional</u>:*

I agree to the University of Birmingham research team sharing my research data with any professionals or clinicians working with me and the person I care for should they request to see them. ☐

Print Name: ________________________________

Name of person you care for: ______________________________

Address:____

Email: ______________________________

Telephone number:___________________________

Relationship to participant: ________________________________

Signature: ________________________Date: __________________

<u>SECTION 2:</u> This is <u>optional</u> and allows keep your personal details on the Regular Participant Database. See section titled 'Regular Participant Database' in the information sheet.

PTO…continued overleaf

Please initial box…

I have read and understood the section titled ‘Regular Participant Database’ and I would like my personal details to be added to the database. ☐

I understand that my name and contact details will be kept by the research team at the University of Birmingham in accordance with the provisions of the Data Protection Act 1998 and I will be contacted by an approved member of the team with information about future research that I and the person I care for may like to participate in. ☐

I understand that if my details are held on the database it will be possible for the research team to trace the results of the assessments that I complete in this project back to me and my child/person I care for so that they can look at changes over time if I take part in future projects. ☐

I understand that even after I have agreed for my details to be added to the database, I can request that they be removed by contacting [redacted] ☐

I understand the Professor Chris Oliver holds ultimate responsibility for the database. ☐

Print Name: ___________________________Signature: ____________________________Date: ______________

APPENDIX E

The Kiddy-KINDL Questionnaire

KiddY-KINDL

Child's Wellbeing Questionnaire:

APPENDIX F

CSRI Questionnaire

Questionnaire about Parents' Experiences and Perceptions about Service Input

In the last 6 months, has your child seen any of the following community based professionals/services **for any reason?**					
Specialist	**Have they ever seen this professional** Y/N	**Total number of contacts** (during last 6 months)	**Average duration of contact (minutes)** during the last six months	**How useful was this service?** Please rate using the scale below	**How effective was this service?** Please rate using the scale below
				Least Useful (1) 1 2 3 4 5 Most Useful (5)	Least Useful (1) 1 2 3 4 5 Most Useful (5)
Community Psychiatrist					
Clinical Psychologist					
Educational Psychologist					
General Practitioner					
Community Psychiatric Nurse					
Learning disability nurse					
Other community nurse (e.g. district					

nurse/health visitor)					
Community mental health team member					
Speech therapist					
SENCO					
Physiotherapist					
Community Paediatrician					
CAMHS services					
Podiatrist					
Audiologist					
Optician					
Dentist					
Play therapist					
Nutritionist/dietician					
Occupational therapist					
Art/drama/music therapist					
Alternative therapist (eg reflexologist)					
Social worker					
Home help/support worker					
Advocate/counsellor					
Dysphagia service					

Hospital based services? (e.g. speciality inpatient ward).					
Other specialist? Specify.					

Prior to the last 6 months, has your child seen any of the following community based professionals/services **for any reason?**					
Specialist	**Have they ever seen this professional** Y/N	**Total number of contacts** (during last 6 months)	**Average duration of contact (minutes)** during the last six months	**How useful was this service?** Please rate using the scale below	**How effective was this service?** Please rate using the scale below
				Least Useful (1) 1 2 3 4 5 Most Useful (5)	Least Useful (1) 1 2 3 4 5 Most Useful (5)
Community Psychiatrist					
Clinical Psychologist					
Educational Psychologist					
General Practitioner					
Community Psychiatric Nurse					

Learning disability nurse					
Other community nurse (e.g. district nurse/health visitor)					
Community mental health team member					
Speech therapist					
SENCO					
Physiotherapist					
Community Paediatrician					
CAMHS services					
Podiatrist					
Audiologist					
Optician					
Dentist					
Play therapist					
Nutritionist/dietician					
Occupational therapist					
Art/drama/music therapist					
Alternative therapist (eg. reflexologist)					
Social worker					
Home help/support					

worker					
Advocate/counsellor					
Dysphagia service					
Hospital based services? (e.g. speciality inpatient ward).					
Other specialist? Specify.					

Interventions for Challenging Behaviour

"Have you had input from any professionals about the challenging behaviour [child's name] engages in?"

In the last 6 months, has your child seen any of the following professionals/services for **challenging behaviour?**					
Specialist	**Have they ever seen** Y/N	**Total number of contacts** (during last 6 months)	**Average duration of contact** (minutes) during the last six months	**Is the behaviour better or worse than before involvement, or has it remained the same?** 1= Behaviour much worse 2= Behaviour a bit worse 3= Behaviour about the same 4= Behaviour is better 5= Behaviour is much better	**How much did the input you received contribute to that change?** 1 = No contribution 2 = A small contribution 3= A moderate amount of contribution 4 =A large amount of contribution

Community Psychiatrist					
Psychologist					
General Practitioner					
Community Psychiatric Nurse					
Learning disability nurse					
Other community nurse (e.g. district nurse/health visitor)					
Community mental health team member					
Speech therapist					
Physiotherapist					
Occupational therapist					
Art/drama/ music therapist					
Alternative therapist (eg. reflexologist)					
Social worker					

Home help/support worker					
Advocate/counsellor					
Dysphagia service					
Hospital based services? (e.g. speciality inpatient ward).					
Other specialist? Specify					

Prior to the last 6 months, has your child seen any of the following professionals or services for **challenging behaviour?**				
Specialist	**Have they ever seen** Y/N	**Timeframe of input** How many appointments? Length of appointments?	**Is the behaviour better or worse than before involvement, or has it remained the same?** 1= Behaviour much worse 2= Behaviour a bit worse 3= Behaviour about the same 4= Behaviour is better 5= Behaviour is much better	**How much did the input you received contribute to that change?** 1 = No contribution 2 = A small contribution 3= A moderate amount of contribution 4 =A large amount of contribution
Community Psychiatrist				

Psychologist				
General Practitioner				
Community Psychiatric Nurse				
Learning disability nurse				
Other community nurse (e.g. district nurse/health visitor)				
Community mental health team member				
Speech therapist				
Physiotherapist				
Occupational therapist				
Art/drama/music therapist				
Alternative therapist (eg.Reflexologist)				
Social worker				
Home help/support worker				
Advocate/counsellor				
Dysphagia service				
Hospital based services? (e.g. speciality inpatient ward).				
Other specialist? Specify				

	Overall				
Specialist	**How much did the input you received contribute to that change?** 1 = No contribution 2 = A small contribution 3 = A moderate amount of contribution 4 =A large amount of contribution	**What were the perceived benefits of the input you received?**	**What were the perceived costs of the input you received?**	**What barriers did you encounter accessing support?**	**Would you like/or consider help from these specialists for challenging behaviour in the future?** If yes, rate on scale. 1 = least likely 2 = small likelihood 3 = quite likely 4 = very likely.
Community Psychiatrist					
Psychologist					
General Practitioner					
Community Psychiatric Nurse					
Learning disability nurse					
Other community nurse (e.g. district nurse/health visitor)					
Community mental health team member					
Speech therapist					
Physiotherapist					

Occupational therapist					
Art/drama/music therapist					
Alternative therapist (eg. reflexologist)					
Social worker					
Home help/support worker					
Advocate/counsel-lor					
Dysphagia service					
Hospital based services? (e.g. speciality inpatient ward).					
Other specialist? Specify					

<u>Questions Regarding Specific Interventions</u>

"You described above how you [name of person] suggested you try [type of intervention]. I am now going to ask you a little more about that intervention and about any other methods you may have tried to manage challenging behaviour".

Potential Intervention	**Y/ N**	**Timescale**	**Is the behaviour better or**	**How much did the input**	**Perceived benefits and costs**	**If the intervention**	**Is this something you**

		Is the intervention ongoing? If Yes: When did it start/finish?	**worse than before involvement, or has it remained the same?** 1= Behaviour much worse 2= Behaviour a bit worse 3= Behaviour about the same 4= Behaviour is better 5= Behaviour is much better	**you received contribute to that change?** 1 = No contribution 2 = A small contribution 3 = A moderate amount of contribution 4 =A large amount of contribution	**in relation to intervention** E.g. Ease of implementation effectiveness, Parental control, Consistency across settings	**terminated, why was this?**	**would consider trying in the future?** If yes rate on scale. 1 = least likely 2 = small likelihood 3 = quite likely 4= very likely.
Education about challenging behaviour (general) with staff/carers							
Education about challenging behaviour (specific) with staff/carers							
Structured reinforcement programme							
Use of restraints							
Restructuring the environment							

Communication training/Functional Communication Strategies							
Sensory integration							
One to one therapy with person you care for (CBT/Psychodynamic/ other)							
Family therapy							
Psychoactive medication							
Treatment of underlying health problem (operation/physical treatment)							
Treatment of underlying health problem (medication							
Special diet							
Vitamin supplements							
Music therapy							
Others							

Additional sources of support/information

"What additional support/information have you had on challenging behaviour apart from that received from specialists?"

	Timeframe of input (past/present)	**Is the behaviour better or worse than before involvement, or has it remained the same?** 1= Behaviour much worse 2= Behaviour a bit worse 3= Behaviour about the same 4= Behaviour is better 5= Behaviour is much better	**How much did the input you received contribute to that change?** 1 = No contribution 2 = A small contribution 3= A moderate amount of contribution 4 =A large amount of contribution	**Nature of the input you received?**	**Perceived benefits and costs**	**What barriers did you encounter accessing support?**	**Would you like/or consider help from these specialists in the future?** If yes what is the likelihood of seeking help 1 = least likely 2 = small likelihood 3 = quite likely 4= very likely.
Parents							
Family							
Friends							
Religion							
Associations							
Teacher							
Proxy							
Written Information / Resources							
Electronic Information							

/ Resources							
Events (e.g. training/ conferences / conventions / support groups)							

UNMET NEEDS

Are there any services you think your child would benefit from that s/he is not receiving at present?

***Yes** or* **No** *(please circle)*

If yes: Please give details ____

Finally, are there any other services – of any kind - not already mentioned that your child has been using in the last 6 months? *(e.g. voluntary services, home help, etc.).* If so, please give details below:

Service ___________________________________ No. of visits / contacts ___________________________

Service ___________________________________ No. of visits / contacts ___________________________

Q. "Do you have any additional comments regarding your experiences of input for challenging behaviour?"

APPENDIX G

Hospital Anxiety and Depression Scale

Hospital Anxiety and Depression Scale (HADS)

APPENDIX H

Median and Mann-Whitney U scores investigating the differences between participants who have access in different type of services and those who do not

Table.5.4 Median scores and Mann-Whitney statistics for measures of self-help, quality of life, anxiety, depression, age, health problems and socioeconomic status for participants with and without medical service use. Bold text indicates a significant difference (p<.01, one tailed).

	Accessed medical professionals	**Not seen medical professionals**	**Mann Whitney U / χ^2**	***P* value**
N	52	12		
Quality of Life Total Score	92.39 (86.55-98.91)	95.92 (92.26-101.10)	233.00	.174
Health problems	2.00 (1.00-4.00)	2.00 (1.00-3.00)	233.50	.768
Anxiety	10.50 (8.00-13.75)	9.00 (7.25-13.25)	243.50	.237
Depression	7.00 (5.25-10.00)	6.50 (4.00-12.00)	294.00	.756
Self-help	5.00 (4.00-7.00)	5.00 (4.00-7.00)	228.00	.415
Socioeconomic status	9.98 (4.93-16.26)	*13.83* *(9.30-21.70)*	149.00	.184
Age	8.50 (5.00-12.00)	*8.00* *(6.00-12.00)*	221.50	.855

Table.5.5. Median scores and Mann-Whitney statistics for measures of self-help, quality of life, anxiety, depression, age, health problems and socioeconomic status for participants with and without mental health service use. Bold text indicates a significant difference (p<.01, one tailed).

	Access in mental health professionals	**Not seen mental health professionals**	**Mann Whitney U / χ^2**	***p*value**
N	20	44		
Self Help	6.00 (4.00-7.00)	5.00 (4.00-7.00)	380.00	.877
Socioeconomic Status	12.25 (4.99-8.63)	10.11 (5.67-14.87)	229.00	.397
Quality of Life	91.85 (84.38-91.23)	93.75 (89.67-99.86)	365.50	.280
Anxiety score	**12.50** **(9.00-16.00)**	**10.00** **(8.00-12.00)**	**289.00**	**.028**
Depression score	9.00 (6.00-11.50)	7.00 (4.00-10.00)	352.50	.203
Health problems score	*3.00* *(1.25-3.00)*	*2.00* *(1.00-3.00)*	279.00	.447
Age	10.00 (6.00-12.00)	*8.00* *(5.00-12.00)*	262.50	.403

Table.5.6. Median scores and Mann-Whitney statistics for measures of self-help, quality of life, anxiety, depression, age, health problems and socioeconomic status for participants with and without social/support service use. Bold text indicates a significant difference (p<.01, one tailed).

	Access in social/support professionals	**Not seen social/support professionals**	**Mann Whitney U / χ^2**	***p*value**
N	43	21		
Self Help	5.00 (4.00-7.00)	6.00 (5.00-7.00)	300.50	.146
Socioeconomic Status	11.34 (6.65-19.76)	9.54 (4.80-14.46)	231.00	.420
Quality of Life	91.85 (85.87-98.91)	94.022 (89.95-101.90)	367.50	.229
Anxiety score	11.00 (8.00-14.00)	9.00 (6.00-11.50)	334.00	.092
Depression score	7.00 (6.00-12.00)	7.00 (4.00-9.00)	407.00	.522
Health problems score	*2.00* *(2.00-4.00)*	*2.00* *(1.00-3.00)*	277.50	.247
Age	10.00 (6.00-12.00)	*5.50* *(4.00-11.25)*	**211.00**	**.021**

Table.5.7. Median scores and Mann-Whitney statistics for measures of self-help, quality of life, anxiety, depression, age, health problems and socioeconomic status for participants with and without learning service use. Bold text indicates a significant difference (p<.01, one tailed).

	Access in learning professionals	**Not seen learning professionals**	**Mann Whitney U / χ^2**	***p*value**
N	46	18		
Self Help	6.00 (4.00-7.00)	4.00 (4.00-7.50)	333.50	.590
Socioeconomic Status	9.89 (4.89-15.26)	14.61 (826-31.45)	168.00	.088
Quality of Life	93.48 (86.68-99.59)	94.02 (90.08-99.59)	378.50	.591
Anxiety score	10.00 (8.00-14.00)	10.00 (8.00-12.50)	404.00	.881
Depression score	7.00 (4.00-10.00)	7.50 (4.25-12.25)	403.50	.875
Health problems score	*2.50* *(1.25-4.00)*	*2.00* *(1.00-2.75)*	249.50	.191
Age	7.50 (5.00-11.75)	*10.50* *(6.50-12.75)*	225.50	.085

Table.5.8. Median scores and Mann-Whitney statistics for measures of self-help, quality of life, anxiety, depression, age, health problems and socioeconomic status for participants with and without alternative therapist service use. Bold text indicates a significant difference (p<.01, one tailed).

	Access in alternative professionals	**Not seen alternative professionals**	**Mann Whitney U / χ^2**	***P* value**
N	46	18		
Self Help	4.00 (4.00-6.50)	6.00 (4.00-7.00)	141.50	.101
Socioeconomic Status	11.34 (5.32-15.94)	10.24 (5.37-19.78)	161.00	.832
Quality of Life	93.21 (88.72-99.18)	93.48 (87.36-99.73)	227.50	.907
Anxiety score	10.50 (8.00-13.50)	10.00 (8.00-14.00)	215.00	.681
Depression score	7.00 (4.00-12.00)	7.00 (4.75-10.00)	227.50	.906
Health problems score	*2.00* *(2.00-3.00)*	*2.00* *(1.00-4.00)*	167.00	.803
Age	7.00 (6.00-10.00)	*10.00* *(5.00-12.00)*	130.00	.260

Table.5.9. Median scores and Mann-Whitney statistics for measures of self-help, quality of life, anxiety, depression, age, health problems and socioeconomic status for participants with and without allied health professionals' access. Bold text indicates a significant difference (p<.01, one tailed).

	Access in allied professionals	**Not seen allied professionals**	**Mann Whitney U / χ^2**	***P* value**
N	58	6		
Self Help	5.00 (4.00-7.00)	7.50 (4.75-9.00)	87.00	.058
Socioeconomic Status	11.58 (6.56-19.69)	6.67 (3.06-12.54)	56.00	.183
Quality of Life	93.48 (87.36-100.00)	93.21 (90.08-95.52)	161.00	.764
Anxiety score	10.00 (8.00-14.00)	10.00 (8.25-11.25)	154.00	.644
Depression score	7.00 (4.00-10.50)	8.50 (4.00-9.75)	168.50	.899
Health problems score	*2.00* *(2.00-3.00)*	*1.00* *(.500-3.50)*	70.50	.094
Age	8.00 (6.00-12.00)	*5.00* *(5.00-10.00)*	87.50	.248

APPENDIX I

Predictors of using different types of services

Table.5.10 Logistic regression predicting likelihood of medical service use

	B	**S.E**	**Wald**	***Df***	***p***	**Odds Ratio**	**95.0% C.I. for Odds Ratio**	
							Lower	Upper
Anxiety	**.314**	**.859**	**3.90**	**1**	**.048**	**1.37**	**1.00**	**1.90**
Depression	-.120	.130	.850	1	.356	.887	.687	1.15
Age	-.020	.131	.023	1	.879	.980	.759	1.30
Gender	-.776	.912	.723	1	.395	.460	.077	2.80
SES	.025	.029	.712	1	.399	1.03	.968	1.09

Table.5.11 Logistic regression predicting likelihood of mental health service use

	B	**S.E**	**Wald**	***Df***	***p***	**Odds Ratio**	**95.0% C.I. for Odds Ratio**	
							Lower	Upper
Anxiety	-.037	.235	.024	1	.876	.964	.609	1.53
Depression	-.019	.209	.009	1	.926	.981	.651	1.48
Age	.185	.209	.787	1	.375	1.20	.800	1.81
Gender	.913	1.89	.233	1	.630	2.49	.061	101.80
SES	.108	.126	.738	1	.390	1.11	.871	1.43
Self-Help score	-.936	.573	2.67	1	.102	.392	.128	1.21

Table.5.12 Logistic regression predicting likelihood of social/support service use

	B	**S.E**	**Wald**	***Df***	***p***	**Odds Ratio**	**95.0% C.I. for Odds Ratio**	
							Lower	Upper
Anxiety	.092	.139	.439	1	.508	1.10	.835	1.44
Depression	-.068	.130	.278	1	.598	.934	.724	1.21
Age	-.229	.132	3.02	1	.082	.795	.614	1.03
Gender	-.710	.849	.699	1	.403	.492	.093	2.60
SES	-.029	.031	.872	1	.350	.971	.913	1.03
Self-Help score	.313	.282	1.23	1	.267	1.37	.787	2.38

Table.5.13 Logistic regression predicting likelihood of learning service use

	B	**S.E**	**Wald**	***Df***	***p***	**Odds Ratio**	**95.0% C.I. for Odds Ratio**	
							Lower	Upper
Anxiety	-.036	.131	.076	1	.782	.964	.746	1.25
Depression	-.078	.115	.461	1	.497	.925	.738	1.16
Age	-.245	.127	3.73	1	.054	.782	.610	1.00
Gender	1.52	.896	2.90	1	.089	.218	.038	1.26
SES	.020	.031	.403	1	.526	1.02	.960	1.08
Self-Help score	.085	.273	.096	1	.756	1.10	.636	1.86

Table.5.14 Logistic regression predicting likelihood of alternative service use

	B	**S.E**	**Wald**	***Df***	*p*	**Odds Ratio**	**95.0% C.I. for Odds Ratio**	
							Lower	Upper
Anxiety	-.037	.235	.024	1	.876	.964	.609	1.53
Depression	-.019	.209	.009	1	.926	.981	.651	1.48
Age	.185	.209	.787	1	.375	1.20	.800	1.81
Gender	.913	1.89	.233	1	.630	2.49	.061	101.80
SES	.108	.126	.738	1	.390	1.11	.871	1.43
Self-Help score	-.936	.573	2.67	1	.102	.392	.128	1.21

Table.5.15 Logistic regression predicting likelihood of allied health service use

	B	**S.E**	**Wald**	***Df***	*p*	**Odds Ratio**	**95.0% C.I. for Odds Ratio**	
							Lower	Upper
Anxiety	-.036	.131	.076	1	.782	.964	.746	1.25
Depression	-.078	.115	.461	1	.497	.925	.738	1.16
Age	-.245	.127	3.73	1	.054	.782	.610	1.00
Gender	1.52	.896	2.90	1	.089	.218	.038	1.26
SES	.020	.031	.403	1	.526	1.02	.960	1.08
Self-Help score	.085	.273	.096	1	.756	1.10	.636	1.86

APPENDIX J

The Activity Questionnaire

APPENDIX K

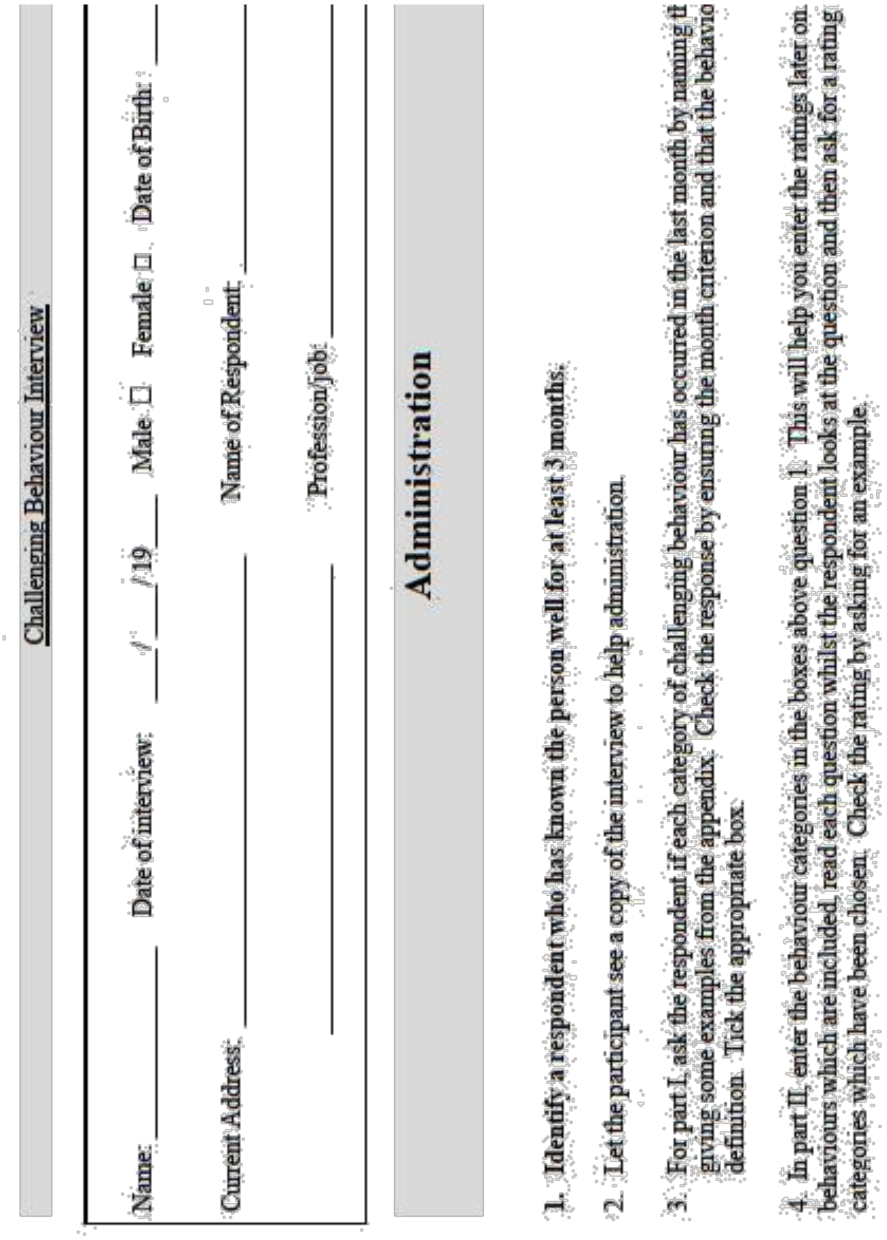
The Social Communication Questionnaire

APPENDIX L

CHALLENGING

BEHAVIOUR

INTERVIEW



Challenging Behaviour Interview

Name: ______________ Date of interview: ____/____/19____ Male ☐ Female ☐ Date of Birth: ____

Current Address: ______________________________ Name of Respondent: ______________

______________________________ Profession/job: ______________

Administration

1. **Identify a respondent who has known the person well for at least 3 months.**
2. Let the participant see a copy of the interview to help administration.
3. For part I, ask the respondent if each category of challenging behaviour has occurred in the last month by naming th giving some examples from the appendix. Check the response by ensuring the month criterion and that the behavio definition. Tick the appropriate box.
4. In part II, enter the behaviour categories in the boxes above question 1. This will help you enter the ratings later on. behaviours which are included, read each question whilst the respondent looks at the question and then ask for a rating categories which have been chosen. Check the rating by asking for an example.

CHALLENGING BEHAVIOUR INTERVIEW (PART I)

1. Which of the following behaviours have occurred in the last month? (See appendix for definitions and example

Challenging Behaviour Category	**List behaviours from examples in appendix**
☐ Self –Injury (SIB)	
☐ Physical aggression (PAG)	
☐ Verbal aggression (VAG)	
☐ Disruption and destruction of property or the environment (DST)	
☐ Anal poking (AP)	
☐ Stereotyped behaviours (STB)	
☐ Inappropriate vocalisations (IV)	
☐ Inappropriate removal of clothing (IRC)	
☐ Pica (PIC)	
☐ Inappropriate or unacceptable sexual behaviour (ISB)	
☐ Smearing (SMR)	
☐ Stealing (STL)	
☐ Self-induced vomiting and regurgitation (SIV)	

CHALLENGING BEHAVIOUR INTERVIEW (PART II)

In each box, enter the category of challenging behaviour that is being considered

1. **Think about how often this behaviour occurred in the last month. If there was no change and you watched thi you *definitely* see the behaviour:**

⑤	④	③	②	①
In the next 15 minutes	In the next hour	By this time tomorrow	By this time next week	By this time next month

2. **In the last month, for how long did the *longest* episode or burst of this behaviour last?**

①	②	③	④	⑤
Less than a minute	Less than 5 minutes	Less than 15 minutes	Less than an hour	More than an hour

3. **In the last month, for how long have episodes or bursts of this behaviour *typically* lasted or lasted on average?**

①	②	③	④	⑤
Less than a minute	Less than 5 minutes	Less than 15 minutes	Less than an hour	More than an hour

4. **For the worst episode of behaviour in the last month, what response was necessary[1]?**

⓪	①	②	③	④
Nothing	Verbal discouragement or reminder	**Informal physical intervention by one member of staff e.g. blocking, holding an arm briefly, taking objects from an individual** Removal to a safe environment Removal of staff or others from immediate environments	Informal physical intervention by more than one member of staff Formal restraint procedure Protective or restrictive devices employed	Seclusion PRN medication Legal involvement or legal advice has been sought Section of MHA invoked

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[1] To score, identify any items which have occurred and take highest scoring item.

5. **In the last month, what has been the worst effect of this behaviour on the individual's *physical health*?**

⓪	①	②	③
No effect at all	**Minor, temporary injury,** such as reddening of the skin, but *no* bruising or tissue damage	**Moderate injury,** such as bruising , cuts or abrasions **or illness lasting less than a day,** e.g. brief stomach upset, a single episode of vomiting	**Significant injury** e.g. fractured bones, sutures required, minor or major operation required **or illness lasting more than a day**

6. **In the last month, what has been the worst *direct* effect of this behaviour on the *physical health* of staff or carers**

⓪	①	②	③
No effect at all	**Minor, temporary injury,** such as reddening of the skin, but *no* bruising or tissue damage	**Moderate injury,** such as bruising , cuts or abrasions **or illness lasting less than a day,** e.g. brief stomach upset, a single episode of vomiting	**Significant injury** e.g. fractured bones, sutures required, minor or major operation required **or illness lasting more than a day**

7. In the last month, what has been the worst *direct* effect of this behaviour on the *physical health* of other servic

⓪	①	②	③
No effect at all	**Minor, temporary injury, such as** reddening of the skin, but *no* bruising or tissue damage	**Moderate injury,** such as bruising, cuts or abrasions **or illness lasting less than a day,** e.g. brief stomach upset, a single episode of vomiting	**Significant injury** e.g. fractured bones, sutures required, minor or major operation required **or illness lasting more than a day**

8. Throughout the whole of the last month, has the behaviour had any negative effects on the well-being of other disruption to planned activities, service users are frightened or upset, belongings or clothing are damaged or

⓪	①	②	③	④
No effect at all on the well-being of other service users	Effect on the well-being of other service users **about once in the last month**	Effect on the well-being of other service **users about once a week**	Effect on the well-being of other service users **about once every 3 days**	Effect on the well-being of other service users **nearly every day**

9. In the last month, what has been the ***direct*** effect of this behaviour on the environment in which the individua

⓪	①	②	③	④
No damage or loss at all	Disruption or mild damage to property or the living areas e.g., objects thrown, furniture tipped, doors slammed, meals spoiled, paint scratched. Item does not require repair or replacement.	Moderate damage to property or living areas e.g. curtains torn, furniture partly broken. Item requires repair but can be used.	Significant damage to property and living areas. Item requires repair and cannot be used.	Extreme damage to property or living areas. Item requires replacement and cannot be used or repaired e.g. windows broken, furniture unusable.

10. In the last month, as a result of this behaviour, have restrictive or protective devices (e.g. arm splints, helmet
clothing (e.g. all-in-one suit) been worn by the individual?

⓪	①	②	③	④
Never	Some of the time	About half the time	Most of the time	Almost continuously

(If so was it: Arm splint(s) ☐, Helmet or headgear ☐, Gloves/mittens/other items on hands ☐, Specially design
(please specify) ________________________________)

11. **Has the environment in which the individual currently lives been modified because of this behaviour (example in the box below)?**

⓪	①	②	③
No modifications	**Modifications to the person's possessions but not elsewhere e.g. padding on a wheel chair, clothing which is strengthened**	**Modifications have been made to the environment but are not noticeable unless pointed out e.g. curtains on Velcro, window locks**	**Modifications have been made to the environment and are noticeable**

Examples of modification to the environment: windows are not made of glass, TV is in a protective cabinet or out of r
cupboard door is secured, a door is secured, curtains are absent (because they will be torn down), pictures are out of reacl
padded, service users are always visible, a room is out of bounds, cutlery is plastic, furniture is deliberately heavy, door c
slamming, wallpaper is washable in rooms apart from kitchen and bathroom, fridge is secured, ornaments are out of reacl
been removed, furniture is chosen because it has particular qualities e.g. no sharp edges etc.

12. **In the last month, as a result of this behaviour, has a verbal response by staff or carers been necessary e.g. dis distraction to another activity, reminder, reprimand?**

⓪	①	②	③	④
Never	**At least once a month**	**At least once a week**	**At least once a day**	**At least once an hour**

13. In the last month, as a result of this behaviour, has physical contact or prevention or restraint by staff or care
blocking, taking objects from an individual, temporary restraint of an arm?

⓪	①	②	③	④
Never	At least once a month	At least once a week	At least once a day	At least once an hour

(If so was it a written procedure ☐ or an informal procedure ☐, please tick.)

14. In the last month, for this behaviour, was it necessary for more than one member of staff to respond when the

⓪	①	②	③	④
Never	At least once a month	At least once a week	At least once a day	At least once an hour

15. In the last month, has there been any contact with any of the following regarding this behaviour?

	Name and Contact Number
☐ Clinical Psychologist or Psychology Assistant working with a Clinical Psychologist	
☐ Psychiatrist	
☐ General Practitioner	
☐ Challenging Behaviour specialist or team	
☐ Speech and language therapist	
☐ Legal advisor	
☐ Other	
☐ Other	
☐ Other	

Summary of Scores

	Behaviours				
Qu.					
1					
2					
3					
4					
5					
6					
7					
8					
9					
10					
11					
12					
13					
14					
Total					

NOTES:

APPENDIX TO PART I: OPERATIONAL DEFINITIONS AND EXAMPLES OF CATEGORIES OF CHALLENGING

Behaviour	Description	Examples	
Self-Injury (SIB)	Non-accidental behaviours which producing temporary marks or reddening of the skin or cause bruising, bleeding or other temporary or permanent tissue damage.	Self-biting, head banging, head-punching or slapping, removing hair, self-scratching, body hitting, eye poking or pressing.	Do not inclu do include p orifices. Any behaviours w criteria shou not under ste behaviours.
Pica (PIC)	Eating inedible substances.	Eating, paper, leaves, discarded food etc.	Must include substances. I cigarette end
Physical aggression (PAG)	A non-accidental, physical act involving physical contact with another person likely to result in pain or distress.	Punching, pushing, kicking, tripping, pulling hair, scratching, throwing objects, using objects as weapons, grabbing clothing.	Do not inclu such as threa
Verbal aggression (VAG)	Verbal statements which are likely to induce fear or distress.	Threats, insults etc.	Exclude inap vocalisations
Disruption and destruction of property or the environment (DST)	A non-accidental physical act which results in disruption or superficial or substantial damage to any property or the environment.	Tearing or chewing own clothing, tearing newspapers, notes, books, breaking windows or furniture, throwing or tipping furniture, slamming doors, pulling pictures from walls, spoiling a meal, throwing objects, knocking objects to the floor.	This include disruption of for example furniture.
Anal poking (AP)	Insertion of objects, fingers or hand into the anus.		

Stereotyped behaviours (STB)	Apparently meaningless, repetitive movements, executed in an almost identical way each time.	Rocking, hand regard, twiddling objects, strobing, patting or tapping part of the body, spinning, head weaving or rolling, constant hand movements, hand sucking and eye pressing (when there is no tissue damage).	Some stereot may result in hair loss fron when in a cha coded under s
Inappropriate vocalisations (IV)		Shouting, screaming, repetitive groaning or moaning, growling.	
Inappropriate removal of clothing (IRC)	Removing clothing to the point at which there is an unacceptable degree of exposure given the social setting.	Stripping when others are present but not for washing, bathing, use of the toilet, changing clothing or going to bed.	
Inappropriate or unacceptable sexual behaviour (ISB)	A sexual act which is inappropriate to the social setting and/or the relationship between the participants.	Masturbation when others are present. Inappropriate kissing and touching. Any physical, sexual contact which is rejected or unwanted .	
Smearing (SMR)	Non-accidental, inappropriate and repeated wiping of faeces, saliva or mucus with the hand.		
Stealing (STL)	Taking items or possessions which do not belong to the individual.		Include takin someone else
Self-induced vomiting and regurgitation (SIV)	1. Vomiting which is induced by inserting the fingers or hand into the mouth or throat. 2. Regurgitating and reswallowing vomit.		Ensure that th not have a me hiatus hernia.

APPENDIX M

BACKGROUND INFORMATION

BACKGROUND INFORMATION

Please tick or write your response to these questions concerning background details:

1. **Today's date:** ________________________

2. **Gender:** Male ☐ Female ☐

3. **Date of Birth**: ___/___/____ **Age:**______________

4. **Is the person you care for verbal?** (i.e. more than 30 signs/words in their vocabulary)

   Yes/No (delete as appropriate)

5. **Is the person you care for able to walk unaided?**

   Yes/No (delete as appropriate)

6. Has the person you care for been diagnosed with a syndrome? Yes/No (delete as appropriate)

If yes, please indicate which syndrome in 5a. and answer questions 6 to 8. If no, please move on to question 9

6.a

Cornelia de Lange syndrome ☐
Prader-Willi syndrome ☐
Fragile X syndrome ☐
Lowe syndrome ☐

Cri du Chat syndrome ☐
Rubinstein Taybi syndrome ☐
Down syndrome ☐
Soto Syndrome ☐

Rubinstein-Taybi syndrome ☐
8p23deletion ☐

9q34 deletion ☐
Tuberous Sclerosis ☐

Other ______________________________

7. What is the genetic mechanism causing the syndrome in the person you care for?

Uni-parental disomy ☐
Deletion ☐
Unknown ☐

Sequence repetition ☐
Translocation ☐
Other ___________________________________

8. When was the person you care for diagnosed? _____________________________________

9. Who diagnosed the person you care for?

Paediatrician ☐
GP ☐

Clinical Geneticist ☐
Other _____________________________

10. Has the person you care for had any medical/health difficulties in the last six months? If yes, please give details:

____

_

____

In the information sheet and consent form we informed you that we may need to contact your child's/person you care for's GP in order to clarify any information regarding your child's health and diagnostic status (see consent form and information sheet for more information). <u>If you have already indicated on the consent form that you are happy for us to do this</u>, please complete the relevant details below:

11. Name of your child's/person you care for's GP____

GP Address____

____GP Telephone number____________________________

The following questions ask for background information <u>about you and your family</u>. Please tick the appropriate boxes or write in the spaces provided.

1. Are you male or female? Male ☐ Female ☐

2. What was your age in years on your last birthday? _____________years

3. Please tick the highest level of your educational qualifications.

No formal educational qualifications... ☐

Fewer than 5 GCSE's or O Level's (grades A-C), NVQ 1, or BTEC First Diploma……. …. ☐

5 or more GCSE's or O Level's (grades A-C), NVQ 2, or equivalent…………………..…….. ☐

3 or more 'A' Levels, NVQ 3, BTEC National, or equivalent... ☐

Polytechnic/University degree, NVQ 4, or equivalent.. ☐

Masters/Doctoral degree, NVQ 5, or equivalent……….. ☐

4. What is your relationship to your child with a genetic syndrome (e.g. mother, father, stepmother, grandmother, adoptive parent)? ______________________________

5. In total how many people currently live in your home? _________ Adults _______ Children

6. Does your child with a genetic syndrome normally live with you? ***Yes*** ☐
No ☐

If no, then where do they live? ____

7. What is your current marital status?

Married, and living with spouse.. ☐

Living with partner... ☐

Divorced/Separated/Widowed/Single and NOT living with a partner......... ☐

If living with partner/spouse, please answer the following questions, if not, please go to question 12.

8. Is your partner male or female? Male ☐ Female ☐

9. What was their age in years on their last birthday? _____________ years

10. Please tick the highest level of your partner/spouse's educational qualifications.

No formal educational qualifications... ☐

Fewer than 5 GCSE or O Level (grades A-C), NVQ 1, or BTEC First Diploma…………… ☐

5 or more GCSE or O Level (grades A-C), NVQ 2, or equivalent………………………..… ☐

3 or more 'A' Levels, NVQ 3, BTEC National, or equivalent.. ☐

Polytechnic/University degree, NVQ 4, or equivalent... ☐

Masters/Doctoral degree, NVQ 5, or equivalent……….. ☐

11. What is your partner/spouse's relationship to your child with a genetic syndrome (e.g., mother, father, stepmother, adoptive parent)? ______________________________

12. Recent data from research with families of children with special needs has shown that a family's financial resources are important in understanding family member's views and experiences. With this in mind, we would be very grateful if you could answer the additional question below. We are not interested in exactly what your family income is, but we would like to be able to look at whether those with high versus lower levels of financial resources have different experiences.

What is your current total annual family income? Please include a rough estimate of total salaries and other income (including benefits) before tax and national insurance/pensions.

Please tick one box only:

Less than £15,000……………………………………………………………..…………..… ☐

£15,001 to £25,000…………………………………………………………...………….. ☐

£25,001 to £35,000……………………………………………………...…..……….. ☐

£35,001 to £45,000………………………………………………………..…...………… ☐

£45,001 to £55,000……………………………………………………...………………..…. ☐

£55,001 to £65,000……………………………………………………..……………….….. ☐

£65,001 or more………………………..……………………………..…………………. ☐

APPENDIX M

WESSEX QUESTIONNAIRE

APPENDIX N
THE MOOD INTEREST AND PLEASURE QUESTIONNAIRE

APPENDIX O

THE RBQ

THE RBQ

APPENDIX P

GENERAL HEALTH DEVELOPMENT

General Health and Development

1. a) Has any professional (eg. doctor, clinical geneticist, paediatrician) said that the child (please tick all that apply)**:**

Is autistic... ☐

Has autistic like traits .... ☐

Has cerebral palsy or muscular dystrophy . ☐

Has a genetic syndrome:............................ ☐

Has an autistic spectrum disorder........................... ☐

Has features of autism... ☐

b) If you answered yes to genetic syndrome, please specify:
……………………………………...……….

2. Please name any prescribed medications the child is currently taking whilst at school: ....

....

3. Please indicate the number of days that the child did not attend school in the last full term:

0-5 ☐ 5-10 ☐ 10-15 ☐ 20+ ☐ N/A ☐

4. Please circle the appropriate response regarding the child's general development

Walk without help	1…Not at all	2…Not upstairs	3…Upstairs and elsewhere
Feed self	1…Not at all	2…With help	3…Without help

Wash self	1…Not at all	2…With help	3…Without help
Dress self	1…Not at all	2…With help	3…Without help
Wetting (days)	1…Frequently	2…Occasionally	3…Never
Soiling (days)	1…Frequently	2…Occasionally	3…Never
Reads	1…Nothing	2…A little	3…Newspapers and/or books
Writes	1…Nothing	2…A little	3…Own correspondence
Counts	1…Nothing	2…A little	3…Understands money values
Speech	1…Never a word	2…Odd words only	3…Sentences and normal
Vision	1…Blind or almost	2…Poor	3…Normal
Hearing	1…Deaf or almost	2…Poor	3…Normal

5. To what extent have the following health problems affected the child in the last month?

	Never	**Mild**	**Moderate**	**Severe**
Eye problems (eg. infections).................................	0	1	2	3
Ear problems (eg. infections)	0	1	2	3
Dental problems (eg. cavities/gum problems)...........	0	1	2	3
Digestive problems (eg. reflux/stomach problems)	0	1	2	3
Skin problems (eg. eczema/dry skin)	0	1	2	3
Any other health or painful condition	0	1	2	3

(please specify) ....