THE ASSOCIATION BETWEEN PAIN AND CHALLENGING BEHAVIOUR IN PEOPLE WITH INTELLECTUAL DISABILITY

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ABSTRACT

Pain is difficult to identify in individuals with intellectual disability. In addition to alleviating discomfort, it is important to recognise pain in these individuals as it may be associated with challenging behaviour.

A number of studies were conducted, which demonstrate a positive association between pain and challenging behaviour in people with intellectual disability. Gastro-oesophageal distress was associated with increased rates of challenging behaviour. Individuals with Tuberous Sclerosis Complex, a genetic syndrome associated with painful tumours, were shown to engage in higher rates (although not statistically significant) of challenging behaviour compared to individuals with Down syndrome, a low-risk pain group. Several psychometric properties of observational pain measures; the FLACC, NCCPC-R and direct pain behavioural codes, were appraised. The most robust pain measures were used to further demonstrate the association between pain and challenging behaviour and to examine the difference in pain scores between individuals with environmentally functional compared to non-functional challenging behaviour. The temporal relationship between behavioural indicators of pain and self injury was also assessed. Results indicated that pain may precede self injury in some individuals.

The results reported in this thesis demonstrate the importance of addressing the influence of pain and environmental factors when assessing and treating challenging behaviour.

DEDICATION

For my Mom

For being proud of me no matter what I do.

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TABLE OF ACRONYMS

ASD	Autism Spectrum Disorder
CBI	Challenging Behaviour Interview
CBQ	Challenging Behaviour Questionnaire
CDC	Cri du Chat syndrome
FACS	Facial action coding system
FAU	Facial action units
FLACC	Face, Legs, Activity, Cry, Consolability
GD	Gastro-oesophageal Distress
GD+	individuals who scored above the assigned cut off point on five or more items on the GDQ
GD-	Individuals with fewer than five clinically significant items on the GDQ
GDQ	Gastro-oesophageal Distress Questionnaire
GORD	Gastro-oesophageal Reflux Disorder
ID	Intellectual disability
MIPQ	Mood, Interest and Pleasure Questionnaire
NCCPC-R	Non-Communicating Children’s Pain Checklist- Revised
QABF	Questionnaire about Behavioural Function
SCQ	Social Communication Questionnaire
SD	Standard deviation
SIB	Self Injurious Behaviour
TAQ	The Activity Questionnaire
TSC	Tuberous Sclerosis Complex

CHAPTER 1

INTRODUCTION: BIOLOGICAL AND OPERANT THEORIES OF CHALLENGING BEHAVIOUR IN PEOPLE WITH INTELLECTUAL DISABILITY

1.1 Overview and rationale

Intellectual disability is defined as

> ‘significant limitations both in intellectual functioning and in adaptive behaviour as expressed in conceptual, social, and practical adaptive skills. This disability originates before age 18.’ (Luckasson, Borthwick-Duffy, Buntinx, Coulter, Craig, Reeve et al., 2002, p. 1).

Prevalence of intellectual disability is estimated to lie between 1% and 3% (Roeleveld, Zielhuis & Gabreels, 1997). Intellectual disability is associated with a number of problems, which may impact on the individual’s quality of life. Emerson, Cullen and Hatton (1996), for example, reported that the quality of life of people with intellectual disability tended to be compromised according to a number of criteria, such as participation in the community, developing skills and independence, and being given choice and control over their lives. Additionally, people with intellectual disability are likely to experience more health problems compared to the general population (Jansen, Krol, Groothoff & Post, 2004; van Schrojenstein, Lantman-de Valk, Henny, Metsemakers, Job, Haveman et al., 2000; van Schrojenstein Lantman-de Valk, Linehan, Kerr & Noonan-Walsh, 2007). Finally, a high proportion of people with intellectual disability engage in challenging behaviours, which have a detrimental impact on the individuals' lives and those of their carers (Emerson, 2003; Lowe*,* Allen, Jones, Brophy, Moore & James*,* 2007; Irazabal, Marsa, Garcia, Gutierrez-Recacha, Martorell, Salvador-Carulla et al., 2012; Peters-Scheffer, Didden & Korzilius, 2012). The possible relationship between the latter two problems experienced by people with intellectual disability, health problems and challenging behaviour, are the focus of this thesis.

Challenging behaviour is defined as:

> 'Culturally abnormal behaviour(s) of such an 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, page 3).

Challenging behaviour has also been defined as:

'Behaviour of such intensity, frequency or duration as to threaten the quality of like and/or the physical safety of the individual or others and is likely to lead to responses that are restrictive, aversive or result in exclusion' (Royal College of Psychiatrists, British Psychological Society and the Royal College of Speech and Language Therapists, 2007).

Challenging behaviour of this nature occurs in a significant minority of people with intellectual disability, with prevalence estimates ranging from 10% to 15% (Emerson, 2003; Lowe et al., 2007). Estimates of challenging behaviour are even higher in individuals with profound intellectual disability (McClintock, Hall & Oliver. 2003). For example, Poppes, van der Putten and Vlaskamp (2010) found self injury and aggression prevalence rates of 82% and 45% respectively in a group of 181 individuals with profound intellectual and multiple disabilities. Challenging behaviour has negative emotional costs for families and carers (Irazabal et al., 2012; Peters-Scheffer et al., 2012) and is often a cause of 'out of area' residential placements, resulting in high financial costs (Allen, Lowe, Moore & Brophy, 2007). For the purpose of this thesis, a slight variation of the definition of challenging behaviour quoted above will be employed. This is to recognise the potential as well as actual problems that are associated with such behaviour. Within this thesis, the term ‘challenging behaviour’ will refer to:

> Behaviour of such intensity, frequency or duration that the physical safety of the person or others is placed in, *or has the potential to be placed in,* serious jeopardy or behaviour which is likely to, *or has the potential to,* seriously limit or deny access to the use of ordinary community facilities.’

Using this definition, which includes low level behaviours with the *potential* to cause injury allows the examination of behaviours, which may otherwise be missed. Although these behaviours may not currently cause injury, they could develop into more severe challenging behaviours in the future. Therefore, understanding the cause of these low-level behaviours, and the factors that influence their maintenance and development is important for developing strategies to prevent or reduce clinically significant higher-level challenging behaviour. However, caution is advised when generalising or comparing the research findings from the following studies included in this thesis, which use this definition, as they may not be representative of research findings when a more restrictive definition of challenging behaviour is employed.

Given the prevalence of challenging behaviour in people with intellectual disability, successful intervention could have numerous beneficial implications for a large number of people. For example, the injuries caused to the individual and to others through engaging in self injury and aggression could be limited, fewer individuals could be placed in out of area residential housing which would reduce the cost of care (Allen et al., 2007) and the stress caused to families through coping with challenging behaviour could be mitigated (Maes, Broekman, Došen & Nauts, 2003).

A number of different strategies can be adopted for treating challenging behaviour. Interventions based on operant learning theory have dominated the research literature (Day, Horner & O'Neil, 1994; Durand & Carr, 1991; Harding, Wacker, Berg, Lee & Dolezal, 2009; Tiger, Hanley & Bruzek, 2008). This theory proposes that challenging behaviour is reinforced by sensory or environmental outcomes (Carr & Durand, 1985: Iwata, Dorsey, Slifer, Bauman, & Richman, 1994; Symons, 2011) (see section 1.3.1). Interventions are aimed at altering these associations or by introducing functionally equivalent behaviours that can replace challenging behaviour (Lalli, Casey & Kates, 1995; Hagopian, Fisher, Sullivan, Acquisto & LeBlanc, 1998). For operant learning interventions to be successful, treatment must be based on prior identification of the 'function' of behaviour. Treatment is then targeted to address each function of challenging behaviour (see section 1.3.2). However, an analysis of 152 single-subject experimental functional assessments of self injurious behaviour has revealed that 25.7% of behaviours were reported to be 'automatically reinforced', often assumed from the absence of a social function, and a further 4.6% were associated with an 'undetermined' function. Therefore, over 30% of self injurious behaviour was not associated with an environmental function (Iwata, Pace, Dorsey, Zarcone, Vollmer, Smith et al., 1994). Similarly, Toogood and Timlin (1996) failed to find an environmental function associated with 50% of challenging behaviours assessed with experimental functional analysis. In these cases, interventions based on contemporary applications of operant learning theory are unlikely to be successful.

Outside of the operant literature the possible association between pain and challenging behaviour is gaining increasing interest (Symons, 2011) (see section 1.3.6). Pain could be caused by underlying health conditions which, if recognised, could be treated effectively (Bosch, Van Dyke, Milligan Smith, Poulton, 1997; Carr & Blakeley-Smith, 2006). Remedying these health conditions could potentially provide a quick and effective strategy for treating challenging behaviour which would not be responsive to operant learning theory based interventions that focused on modifying environmental contingencies or introducing new behaviours.

Currently, there are few reported studies of interventions used to treat challenging behaviour that are targeted at underlying health problems. This could be due to two reasons. Either, appropriate interventions may not be available or it is not possible to determine when medical intervention would be appropriate to treat challenging behaviour. Given the availability of effective interventions for painful health problems, such as tooth decay, gastro-oesophageal reflux disorder and constipation, which are common health complaints in people with intellectual disability (Bohmer, Niezen-de Boer, Klinkenberg-Knol, Deville & Nadorp, Meuwissen, 1999; Böhmer, Taminiau, Klinkenberg-Knol & Meuwissen, 2001), it is unlikely that lack of availability of effective intervention can explain the limited number of health and pain focused interventions. Therefore, it follows that increasing the ability to identify pain and/or health problems in people with intellectual disability, may lead to an increase in successful interventions for challenging behaviour that is related to pain.

Developing strategies for identifying health problems in people with intellectual disability is not only important because of the potential improvement in challenging behaviour, but also because increasing knowledge in this area may lead to better health care and help to address the inequalities in health faced by people with intellectual disability (Emerson & Baines, 2010). Compared to the general population, people with intellectual disability have a shorter life expectancy (Hollins, Attard, van Fraunhofer, McGuigan & Sedgwick, 1998; McGuigan, Hollins & Attard, 1995), higher rates of health problems such as respiratory disease (Puri, Lekh, Langa, Zaman & Singh, 1995), dental problems (Barr, Gilgunn, Kane & Moore, 1999; Tiller, Wilson, Gallagher, 2001), sensory impairments (Carvill, 2001) and epilepsy (Branford, Bhaumik & Duncan, 1998; Matthews, Weston, Baxter, Felce & Kerr, 2008) and are more likely to have poor general health (Emerson & Hatton, 2007a; Emerson & Hatton, 2007b). Investigating the association between pain and challenging behaviour, and in doing so, improve the ability to identify and diagnose pain and health problems in people with intellectual disability, has the potential to reduce these inequalities in health.

In summary, challenging behaviour is a major problem for a significant number of people with intellectual disability. The operant learning based treatments currently available may not be appropriate for a proportion of people showing this behaviour. Research is needed to investigate other possible causes of challenging behaviour and this thesis will focus on the association between pain and challenging behaviour. In comparison to the well established operant learning theory of challenging behaviour, this is a relatively under-researched topic. There is a strong possibility that increasing the understanding of the association between pain and challenging behaviour could have a significant impact on the challenging behaviour shown by some people with intellectual disability as effective interventions may be readily available, and these medical based treatments could improve the health and quality of life of the individual, in addition to treating the challenging behaviour.

There is increasing evidence of a number of person characteristics that are associated with challenging behaviour in people with intellectual disability. These differences between individuals with and without challenging behaviour further signify that internal factors, such as pain, may influence some cases of challenging behaviour.

1.2 Person characteristics associated with challenging behaviour

There are several person characteristics that are associated with increased risk of challenging behaviour (McClintock et al., 2003). These include profound intellectual disability (McClintock et al., 2003; Poppes et al., 2010), a diagnosis of Autism Spectrum Disorder (ASD) (Baghdadli, Pascal, Grisi & Aussilloux, 2003; McClintock et al., 2003; Schopler, Reichler, De Vellis & Daly, 1980), poor communication skills (Deb, Thomas and Bright, 2001; McClintock et al., 2003), high levels of repetitive and stereotyped behaviours (Arron, Oliver, Berg, Moss & Burbidge, 2011; Petty & Oliver, 2005) high levels of activity and impulsivity (Arron et al., 2011; Hyman, Oliver & Hall, 2002; Oliver, Sloneem, Hall & Arron, 2009) and low affect (Ross and Oliver, 2002; Smiley & Cooper, 2003). Also, many genetic syndromes are associated with increased risk of challenging behaviour (Anderson & Ernst, 1994; Arron et al., 2011; Christie, Bay, Kaufman, Bakay, Borden & Nyhan, 1982; Clarke & Dykens, 1997; Rojahn, 1986).

1.2.1 Autism Spectrum Disorder

A diagnosis of ASD is associated with increased risk of challenging behaviour. Baghdadli et al., (2003) found that 50% of individuals with ASD engage in self injurious behaviour. They also found that higher severity of Autistic symptoms, as assessed by CARS (Schopler et al., 1980), is associated with increased risk of challenging behaviour. McClintock et al. (2003), in a meta-analysis of literature to identify risk markers for challenging behaviour, reported increased risk of self injury, aggression and disruption to the environment in people with ASD.

1.2.2 Poor communication skills

Poor communication skills are associated with self injury (Deb et al., 2001). Individuals with restrictive expressive communication are more than twice as likely to have severe self injury that persists over a seven year follow-up compared to individuals without expressive language difficulties (Emerson, Kiernan, Alborz, Reeves, Mason, Swarbrick et al., 2001b). In a meta-analysis, McClintock et al., (2003) demonstrated a significant association between poor expressive and receptive communication and self injury (p<.001). However, no significant association was observed between expressive communication and aggression.

1.2.3 Repetitive behaviours

Repetitive behaviour is an umbrella term referring to a set of behaviours, including stereotyped behaviour, that are highly repetitive, invariant and inappropriate in either goal or motor pattern (Turner, 1997). An association between self injurious behaviour and repetitive behaviour has been reported in people with intellectual disability (Petty & Oliver, 2005). Similarly, Arron et al. (2011) investigated the differences in person characteristics, including repetitive behaviours, between individuals with and without self injurious and aggressive behaviour in seven syndrome groups. Significantly higher rates of repetitive behaviours were observed in individuals with self injury compared to those without in four of the six groups tested (insufficient data for one group to be tested). Also, significantly higher rates of repetitive behaviours were also recorded in individuals with aggression compared to no aggression in two of the seven groups.

1.2.4 Hyperactivity

Hyperactivity is common in individuals with intellectual disability. Prevalence rates of hyperactivity in children with intellectual disability range from 15.7% to 50% (Epstein, Cullinan & Gadow, 1986; Handen, McAuliffe, Janosky, Feldman & Breaux, 1994) and an estimate of 20% has been reported in adults with intellectual disability (Fox & Wade, 1998). Hyperactivity was shown to be associated with self injury in people with Cornelia de Lange, Fragile X, Prader-Willi and Lowe syndromes (Arron et al., 2011). Hyperactivity was also shown to be associated with aggression in the Cornelia de Lange, Fragile X, Prader-Willi, Angelman and Smith-Magenis syndrome (Arron et al., 2011).

1.2.5 Impulsivity

Impulsive behaviours are those over which the individual seems to have very little control and are associated with a "quick, spur of the moment" action (Eysenk, Easting & Pearson, 1984). Compulsive behaviours, similarly characterised by a lack of inhibition in some neuropsychological models, are associated with self injurious behaviour in individuals with Cornelia de Lange syndrome (Arron et al., 2011; Hyman et al., 2002; Oliver et al., 2009), Fragile X, Prader Willi and Lowe syndrome (Arron et al., 2011). Individuals with aggression have also been shown to have higher levels of impulsivity compared to individuals without aggression in Cornelia de Lange, Fragile X, Prader Willi, Lowe, Cri du Chat and Angelman syndromes (Arron et al., 2011).

1.2.6 Low mood

In a literature review of behavioural correlates of depressive episodes in people with intellectual disability, Smiley and Cooper (2003) found multiple examples of a positive association between depression and self injury and aggression. Some researchers suggest that challenging behaviour is an atypical symptom of depression in people with severe intellectual disability (Marston, Perry, & Roy, 1997; Meins, 1995). This assertion is in part based on tautological reasoning whereby a group of individuals diagnosed with depression on the basis of atypical symptoms such as challenging behaviour, exhibit higher levels of challenging behaviour compared to 'non-depressed' individuals. It is equally plausible that challenging behaviour and low mood, in combination, might be indicative of pain or discomfort. To investigate the association between low mood and challenging behaviour, without the confound of a previous diagnosis of depression, Ross and Oliver (2002b) used the 'Mood, Interest and Pleasure Questionnaire' (MIPQ- Short version; Ross, Arron & Oliver, 2008) to measure the two key constructs of depression in people with intellectual disability (low mood and a loss of interest and pleasure). Results showed that individuals who engaged in at least one form of challenging behaviour over the last month scored significantly lower on both the 'mood' and 'interest and pleasure' subscales.

1.2.7 Genetic syndromes

Many genetic syndromes are associated with increased risk of challenging behaviour which contributes to the purported behavioural phenotype associated with the disorder (Arron et al., 2011). A behavioural phenotype is defined as an increased likelihood that people with a genetic syndrome will exhibit behavioural or developmental features when compared to individuals without a syndrome (Dykens, 1995). For example, self injury has been reported in 61% to 92% of individuals with Cri du Chat syndrome and aggression in 70% to 82% (Clarke & Dykens, 1997; Rojahn 1986). In Lesch-Nyhan Syndrome, self injury is shown by almost 100% of individuals (Anderson & Ernst, 1994; Christie et al., 1982) and this tends to be biting targeted around the lip, cheek and fingers (Anderson & Ernst, 1994; Christie et al., 1982). Individuals with Cornelia de Lange syndrome have also been shown to be at increased risk of self injury, with typical prevalence estimates ranging from 16% to 66% (Beck, 1987; Berney, Ireland & Burn, 1999; Oliver et al., 2009; Hyman et al., 2002).

1.2.8 Mobility

Individuals with severe intellectual disability who are ambulant have also been shown to engage in a higher number of challenging behaviours compared to non-ambulant children. For example, in a study including 114 children aged four to eleven years, Chadwick et al., (2000) demonstrated that destructive behaviours and physical aggression were significantly higher in ambulant children compared to non-ambulant children. However, this difference was not observed when looking at self injurious behaviour. These findings may reflect the fact that non-ambulant children have fewer opportunities to engage in acts of destruction or aggression whereas this is not the case with self injurious behaviour.

1.2.9 Summary of person characteristics associated with challenging behaviour

A number of person characteristics associated with challenging behaviour in people with intellectual disability have been identified. These findings demonstrate the influence of 'internal', rather than environmental, factors on the development of behaviours such as self injury and aggression. Investigation of these person characteristics could be used in clinical and research practice to help identify individuals at risk of challenging behaviour. Examination of the underlying mechanisms linking these person characteristics and aberrant behaviours could also help to inform theories which seek to explain the causes of challenging behaviour. There is also the possibility that some of these characteristics are associated with a third variable of pain and discomfort or health problems. For example, some syndromes are associated with health conditions known to be painful and some behaviours such as low mood and hyperactivity might be indicative of pain. Clearly, research in this area must acknowledge these possibilities. These associations also allude to strategies for research that might help progress understanding of the pain and challenging behaviour association.

1.3 Theories of challenging behaviour

There are several commonly cited theories of challenging behaviour that can be divided broadly into 'internal/biological' causes of challenging behaviour or an environmental\operant learning theory of challenging behaviour. These theories are not mutually exclusive or competing at a population level and it is entirely possible that different theories can account for similar and different forms of challenging behaviour both between and within individuals and over time.

1.3.1 Operant learning theory

The operant learning theory states that behaviours are inadvertently shaped through social or automatic reinforcement (Carr & Durand, 1985: Iwata et al., 1994a; Iwata et al., 1994b; Symons, 2011). Before being subject to social operant conditioning, behaviours may occur as accidental movements, stereotyped or pain-related behaviours. These actions can be subject to positive reinforcement through social attention (Carr, 1977; Hanley, Iwata & McCord, 2003; Iwata et al., 1994a; Iwata et al. 1994b; Lovaas, Freitag, Gold & Kassorla, 1965), negative reinforcement such as the removal of demands (Carr & Blakely-Smith, 2006) or automatic reinforcement such as sensory stimulation (Carr, 1977) and develop into more challenging behaviours. If the consequence remains reinforcing, the likelihood of the behaviour occurring in the presence of the antecedent is increased. For example, if challenging behaviour is reinforced with social attention, the challenging behaviour will become more likely to occur when social attention is not being provided. If the behaviour is not reinforced, it is, ultimately, likely to be extinguished from the behavioural repertoire (Oliver, Hall & Murphy, 2005).

1.3.2 Evidence for the operant learning theory of challenging behaviour

Evidence for the operant learning theory of challenging behaviour comes from a number of sources. Firstly, the impact of operant learning on challenging behaviour can be observed through experimental functional analysis, arguably the most robust way to assess the influence of environmental factors on challenging behaviour. Iwata et al., (1982) were the first to formulate a structured protocol to assess the environmental influences on challenging behaviour. In this pivotal study, the impact of systematically manipulating an individual’s social environment on self-injurious behaviour was assessed. Results clearly indicated a strong association between antecedent environmental settings and the expression of challenging behaviour. Since its initial use, variations of this protocol have been employed, producing a strong experimental literature in support of the operant learning theory of challenging behaviour (Day et al., 1994; Derby, Wacker, Peck, Sasso, Deraad, Berg et al., 1994; Hanley et al., 2003; Iwata et al., 1994a; Smith & Iwata, 1997).

Secondly, interventions based on the operant learning theory have been successful in reducing challenging behaviour in people with intellectual disability (Durand & Carr, 1991; Harding et al, 2009; Tiger, Hanley & Bruzek, 2008). For example, Day et al. (1994) demonstrated the effective use of functional communication training for reducing the rates of self injury and aggression in three individuals. During functional communication training, an individual is taught an alternative behaviour that elicits the rewarding response associated with challenging behaviour (Chandler & Dahlquist, 2006). This new communication then becomes functionally equivalent to the challenging behaviour but is associated with a lesser cost for the individual (i.e. less effort, no pain associated with self injurious behaviour) and thus, replaces challenging behaviour in the behavioural repertoire.

Finally, evidence for the operant learning theory of challenging behaviour can also be attained from natural observations, which identify antecedents and consequences to challenging behaviour occurring in the natural environment (Iwata, Pace, Kalsher, Cowdery, & Cataldo, 1990). This method of investigation is less restrictive than experimental functional assessments as the antecedent and consequent variables are not limited to those manipulated by the researcher (Mace & Lalli, 1991). However, function of behaviour cannot be inferred from natural observations as the association between the variables can only be described in terms of a correlation (Mace, Lalli, & Shea, 1992).

1.3.3 Limitations of the operant learning theory

The operant learning theory explains the function of challenging behaviour but does not account for the form. It does not explain why some topographies of challenging behaviour are associated with specific genetic syndromes (Arron et al., 2011), such as nail pulling in Smith-Magenis syndrome (Bouras, Dykens & Smith 2003), skin picking in Prader-Willi syndrome (Hellings & Warnock, 1994) and lip, tongue and finger biting in Lesch- Nyhan syndrome (Pellicer, Buendia-Roldan & Pallares-Trujillo, 1998). The operant learning theory also fails to explain why challenging behaviour is associated with any person characteristics (see sections 1.2.1 to 1.2.6). If challenging behaviour was determined by environmental factors *only*, it would be expected that the prevalence of challenging behaviour would be the same across all individuals, irrespective of personal characteristics. Finally, experimental functional assessments fail to ascribe a function to all challenging behaviours (Iwata et al., 1994b; Toogod & Timlin, 1996). Therefore, the operant learning theory of challenging behaviour is insufficient as an explanation of all challenging behaviour in people with intellectual disability.

1.3.4 Internal/Biological theories

Biological theories of challenging behaviour suggest that internal factors, such as abnormalities in neurotransmitter systems or underlying pain, influence the development of challenging behaviour. Biological theories provide a possible explanation for the difference in form of challenging behaviour observed across different genetic syndromes and could also explain the cause of challenging behaviour not associated with an environmental function.

1.3.5 The influence of neurotransmitter systems on challenging behaviour

Atypicality in neurotransmitter systems may influence the development of self injurious behaviour (Symons, 2011). For example, serotonergic and dopaminergic systems have been found to be abnormal in individuals with Lesch-Nyhan syndrome (Breese, Criswell, Duncan, Moy, Johnson, Wong et al., 1995; Cataldo & Harris 1982; Symons 2011). A review of animal models also indicates an influence of these systems in the expression of self injury (Winchel & Stanley, 1991). There has also been some success in treating self injurious behaviour with the use of atypical antipsychotic drugs, which alter the functioning of these neurotransmitter systems (Breese, Knapp, Criswell, Moy, Papadeas & Blake, 2005; Hammock, Levine & Schroeder, 2001; Dinca, Paul & Spencer, 2005). However, any medication that influences candidate neurotransmitter dysfunction that might be related to challenging behaviour has not been reliably identified.

1.3.6 Pain and Challenging Behaviour

There is a growing literature reporting on the association between pain and challenging behaviour in people with intellectual disability (Symons, 2011) (see section 2.2.5). This relationship has generally been demonstrated by reductions in challenging behaviour following treatment for painful health conditions (Bosch et al., 1997; Carr & Blakeley-Smith, 2006) and via the use of observational measures of pain, which show significantly higher numbers of pain indicators in individuals with challenging behaviour compared to those without the behaviour (Hartman, Gilles, McComas, Davon & Symons, 2008; Symons & Danov, 2005; Symons, Harper, McGrath, Breau & Bodfish, 2009a).

1.3.6.1 Investigating pain in people with intellectual disabilities

Pain is a subjective and personal experience. Therefore, reliance is often placed on self report to identify its presence and quantify severity (Van Schrojenstein Lantman-de Valk et al., 2007). Self report of pain has been used successfully with some individuals with intellectual disability (Fanurik, Koh, Harrison, Conrad & Tomerlin, 1998). For example Voepel-Lewis et al., (2002) demonstrated that a group of children with mild intellectual disability were able to self report level of pain using simple facial scales and number scales, or with the use of words such as ‘a lot’. However, self report is not possible in many individuals with intellectual disability, who have insufficient expressive communication skills (Adams & Oliver, 2011). In these individuals, pain is inferred from the observation of behavioural indicators. Some observation measures rely on fine grained coding of facial expressions (Facial Action Coding System: Ekman & Friesen, 1978; Symons, Harper, Shinde, Clary & Bodfish, 2010) whereas other methods focus on broader behaviour patterns such as body movements, negative affect and changes to physiological patterns such as eating and sleeping routines (FLACC: Malviya, Voepel-Lewis, Burke, Merkel & Tait, 2006; NCCPC-R: Breau, McGrath, Finley & Camfield, 2004).

1.3.6.2 The opioid hypothesis

Endogenous opioids are released as a natural analgesic in response to pain. This can produce a ‘natural-high’, which may then become associated with self injury. This may increase the likelihood that the self injurious behaviour will persist (Rojahn, Schroeder & Hoch, 2008; Sandman, 1990/1991). Significant improvements in self injury following treatment with naltrexone, an opioid receptor antagonist, provide evidence in support of this theory (Symons, 2011; Symons, Thompson & Rodriguez, 2004). However, a beneficial effect of naltrexone is only observed for some forms of self injury (Symons, 2011), suggesting the influence of opioids may be limited to certain forms of self injury.

1.3.6.3 The ‘reduced pain perception’ hypothesis

Reduced perception of pain could eliminate the negative cost typically associated with self injury (Rojahn et al. 2008; Sandman 1990/1991). Response efficiency is generally defined as the difference between the gain (reinforcing consequence) and cost associated with a behaviour (Oliver, 1995). The cost typically incurred through self injury is physical effort and pain. Reduction or removal of pain will improve the response efficiency of the behaviour and therefore increase the likelihood of its survival in the behavioural repertoire. Once established, the behaviour can then be moulded through the process of social reinforcement described above (see section 1.3.1). Research findings involving individuals with genetic syndromes associated with high levels of self injury support this theory. For example, an absence of key proteins in the perception of pain was found in Rett syndrome (Peebles & Price, 2012). Similarly, a protein involved in pain amplification is absent in individuals with Fragile X syndrome (Peebles & Price, 2012). Pain amplification refers to the decrease in pain threshold around the site of injury. This would typically protect the tissue from further damage. Absence of, or reduced pain amplification would mean that the area of self injury would be less sensitive to future insults, reducing the aversive nature of repeated self injury. Again, this would increase the efficiency of self injury as a functional behaviour.

1.3.6.4 The gate control theory

The gate control theory (Melzack & Wall 1965/1982) suggests that stimulation of non-pain receptors inhibits the synaptic transmission of nerve impulses travelling from pain receptors. This modulates the resulting pain experience (Melzack & Wall 1965/1982; Woolf & Slater 2000). Individuals with intellectual disability may learn an association between stimulation around the site of pain and the subsequent reduction of discomfort. These non self-injurious actions originally associated with reducing the perceived level of pain could then be shaped into self injurious behaviours through the process of operant learning. Although the gate control theory seems to be valuable for explaining how self injury could initially be introduced, recent research has questioned its validity. Electronic stimulation of non-pain nerve fibres demonstrated no reduction in the transmission of nerve impulses along C-fibres (those relaying pain signals from pain receptors) (Nathan & Rudge, 2012). Alternatively, rather than self injury stimulating non-pain receptors, as is proposed by the gate control model, it could be that self injury moderates the perception of underlying pain in a different way. In a typically functioning nervous system, noxious stimuli below detection levels would increase the threshold at which pain is perceived. In individuals with intellectual disability that have pre-existing pain, more intense stimulation (such as self injury), may be required to decrease the perception of pain (Peebles & Price, 2012).

1.3.7 Limitations of the internal/biological theory of challenging behaviour

Variations in challenging behaviours within genetic syndromes and the influence of environmental factors on challenging behaviour show that genetic/biological determinants alone are insufficient for explaining all challenging behaviour in people with intellectual disability (Tunnicliffe & Oliver, 2011). This reinforces the value of using both internal/biological theories and the operant learning theory to produce a more comprehensive account of challenging behaviour, which can be applicable across different forms of challenging behaviour and their development.

1.3.8 Interactions between operant and biological theories of challenging behaviour

Studies addressing the influence of operant learning on challenging behaviour within genetic syndromes provide an opportunity to combine the biological and operant theories of challenging behaviour. Tunnicliffe and Oliver (2011) conducted a review of the literature in this area and found that phenotypic problem behaviours can be shaped through operant processes. This interaction can take on different forms within different genetic syndromes. For example, in some cases, a particular genetic syndrome may increase the risk of the challenging behaviour being introduced into the behavioural repertoire, which is then subject to operant reinforcement. For example, a case study of a child with Rett syndrome demonstrated how mouth flicks and mouth hits, both phenotypic behaviours, were more likely to occur in particular environmental settings. The mouth flicks appeared to provide sensory stimulation and were observed in higher frequencies during times of no environmental stimulation. The mouth hits seemed to be associated with a function of social avoidance (Oliver, Murphy, Crayton & Corbett, 1993). A second way biologically determined behaviours may be influenced by environmental factors was also identified. Phenotypic characteristics of a genetic syndrome may increase the motivational drive for particular environmental outcomes. This would then increase the likelihood of the individual engaging in challenging behaviours that were previously paired with the reinforcing consequence, and thus, impact on the operant learning process (Tunnicliffe & Oliver, 2011).

Underlying pain could also interact with operant learning processes to influence challenging behaviour. Firstly, self injury introduced to moderate the perception of underlying pain could then be influenced by operant processes (see section 1.3.6.4). For example, Carr and McDowell (1980) demonstrated the successful treatment of self injurious scratching with operant theory based behavioural intervention. The self injury was initiated in response to contact dermatitis, which is inflammation of the skin sometimes associated with a burning or stinging sensation (Malten, 1981). Scratching is a good example of behaviour that may moderate the perception of pain through 'pain gating' (Melzack & Wall 1965/1982; Woolf & Slater, 2000). Secondly, if the perception of pain is reduced in an individual, there would be a reduced 'cost' associated with self injury and therefore, it would be more 'efficient', in terms of operant learning, at evoking an associated rewarding response (see section 1.3.6.4). Finally, pain could act as an 'establishing operation'/'setting event' (Carr & Smith, 1995; Carr & Blakeley-Smith, 2006; Carr, Smith, Giacin, Whelan, & Pancari, 2003; McGill, 1999). An establishing operation is a contextual variable that influences the pre-existing relationship between antecedents, behaviours and reinforcing consequences. For example, the presence of pain could increase the aversive nature of a task demand, and therefore make the removal of the demand more rewarding. Challenging behaviour, previously reinforced by the removal of demanding tasks, would then be more likely to occur when pain was present (Carr & Smith, 1995; Carr & Blakeley-Smith, 2006; Carr, Smith, Giacin, Whelan, & Pancari, 2003).

1.4 Chapter summary

This chapter has provided an overview of different theoretical accounts of causes of challenging behaviour in people with intellectual disability. Challenging behaviour has a negative impact on the lives of many individuals within this population as well as the family and carers who support them. Understanding how challenging behaviour is introduced into the behavioural repertoire and then develops or is maintained over time can help inform intervention strategies to treat or prevent challenging behaviour. The potential improvement to the lives of these individuals testifies to the importance of conducting research in this area.

Previous research has shown how operant learning processes and biological/internal factors can shape the development of challenging behaviour. Although each theory may be valuable for explaining some cases of challenging behaviour, neither theory alone is sufficient for explaining all challenging behaviour. Combining the biological and environmental theories can provide a more comprehensive and robust explanation for challenging behaviour. Possible ways in which these influential factors can interact to cause or maintain challenging behaviour have been discussed.

The possibility that pain causes challenging behaviour is important for several reasons. Inability to self report pain may contribute to the increased risk of challenging behaviour observed in people with profound intellectual disability and those with communication difficulties. Also, people with many genetic syndromes with behavioural phenotypes including challenging behaviours may also be more likely to suffer health conditions associated with pain (Berg, Arron, Burbidge, Moss & Oliver, 2007; Bull, Fitzgerald, Heifetz

& Brei, 1993; Luzzani, Macchini, Valade, Milani & Selicorni, 2003; Sommer, 1993; Van Schrojenstein Lantman-de Valk et al., 1997). This again may contribute towards the increased risk of challenging behaviour in some genetic syndromes. Also, if pain is shown to cause challenging behaviour, then it would not only be ineffective to treat the behaviour using non-pain based interventions but also unethical. Failure to effectively investigate and treat pain could lead to health conditions remaining undiagnosed and the individual experiencing unnecessary distress and discomfort. Finally, if pain does influence challenging behaviour, then greater awareness of this relationship may lead to the identification and diagnosis of previously unrecognised pain.

There is clear justification for investigating the possible influence of pain on challenging behaviour. However, the subjective and personal nature of pain means that it is difficult to investigate in people with limited expressive communication skills. The overarching aim of this thesis is to use a range of methodological approaches to indirectly measure pain, appraise the psychometric properties of measures of pain and then assess the association between pain behaviours and challenging behaviour in people with intellectual disability.

1.5 Thesis outline

It is important to evaluate the status of a purported association between pain and challenging behaviour in order to inform appropriate research strategies and the focus of research. The first step in achieving this is to review the available literature. Therefore, a systematic literature review will be conducted to summarise key findings relating to the association between pain and challenging behaviour.

As noted above, a positive association has been found between health conditions typically associated with pain, and challenging behaviour in people with intellectual disability (Bosch, et al., 1997; Carr & Blakeley-Smith, 2006). Also, many individuals with intellectual disability have a compromised ability to self report. This makes the identification of pain in this population difficult. One way to tackle this problem is to quantify and examine behaviour in the presence of health problems where pain in likely. In chapter 3, challenging behaviour will be compared between individuals with behavioural indicators of gastro-oesophageal distress to those without. This will provide an indication of how the prevalence and severity of challenging behaviour can be influenced by painful health conditions. Other correlates of challenging behaviour will also be investigated in chapter 3, such as low affect and high activity and impulsivity levels. This will add to the literature on person characteristics associated with challenging behaviour and whether they might be pain related.

Certain genetic syndromes are associated with increased risk of challenging behaviour (Arron et al., 2011). This may be due to the influence of pain and health problems associated with the syndrome. Investigating challenging behaviour in individuals with a genetic syndrome associated with painful health problems provides a second indirect strategy for examining the association between pain and challenging behaviour. In chapter 4, the challenging behaviour of individuals with Tuberous Sclerosis Complex (TSC) will be compared to that seen in Down Syndrome and individuals with other syndromes. This method allows comparisons to be made between several groups, all with well documented health and behavioural characteristics. TSC is selected as the group of interest as it is characterised by growths in all organs of the body, which often cause pain.

As discussed above there are methodological difficulties associated with identifying pain in individuals with intellectual disability who are unable to communicate (Adams & Oliver, 2011). To overcome this problem, research has focussed on the development of observational pain measures (Malviya et al., 2006; Breau, et al., 2000). The study described in chapter 5 investigates the validity and reliability of observational measures of pain. The findings of this chapter inform the research methodology of chapters 6 and 7 and also have wider reaching implications for research in general.

As noted in sections 1.3.1 and 1.3.6.1 to 1.3.6.4, many theories of challenging behaviour have been developed. The operant learning theory explains challenging behaviour in terms of environmental influences (Carr & Durand, 1985: Iwata et al., 1982/1994; Symons, 2011). The biological models of challenging behaviour propose that internal factors such as pain or differences in neurotransmitter systems influence challenging behaviour (Peebles & Price, 2012; Rojahn et al., 2008; Sandman, 1990/1991; Symons, 2011; Woolf & Slater 2000). Independently, each of these theories is insufficient to fully explain challenging behaviour in all individuals. By combining these theories, a more comprehensive explanation for challenging behaviour can be formed (Carr & Blakeley-Smith, 2006; Tunnicliffe & Oliver, 2011). This also highlights the importance of investigating both environmental and internal factors when assessing challenging behaviour in people with intellectual disabilities. In chapter 6, an assessment protocol is trialled that investigates the environmental and pain influences on challenging behaviour in a group of children with intellectual disability at high risk of challenging behaviour. Experimental functional assessments are used to identify individuals with challenging behaviour associated with an environmental function and observations of behavioural indicators of pain are used to infer the presence of pain. As in chapters 3 and 4, person characteristics associated with challenging behaviour are assessed. This will produce a comprehensive description of challenging behaviour in people with intellectual disability in terms of associated person characteristics, the proportion of people with challenging behaviour associated with an environmental function and the difference in pain behaviours between individuals with functional verses non-functional challenging behaviour.

There are many theories of how pain can influence challenging behaviour, and in particular self injury (the opioid hypothesis: Rojahn et al. 2008; Sandman, 1990/1991; 'reduced pain perception': Peebles & Price 2012; Rojahn et al. 2008; Sandman 1990/1991; gate control theory: Melzack & Wall 1965/1982; Woolf & Slater 2000) (see sections 1.3.6.2 to 1.3.6.4). Many of these theories propose that pain causes self injury (Melzack & Wall 1965/1982; Peebles & Price 2012; Woolf & Slater, 2000). However, it is equally plausible that self injury causes pain. One way to investigate which of these interpretations is accurate is to explore the local, temporal sequence of pain behaviours and self injurious behaviours. In chapter 7, the temporal association between behavioural indicators of pain and episodes of self injury are investigated in three participants. The results from chapter 7 are used to demonstrate how the nature of the association between pain and self injury can vary between individuals.

Finally, in chapter 8, key research findings from the five empirical chapters are discussed and implications are considered. The methodological and theoretical constraints of conducting research in this area are explored and future research directions identified.

1.6 Conclusion

Challenging behaviour is a major problem in the lives of people with intellectual disability. There is a well established literature demonstrating the role of operant learning processes on challenging behaviour but this theory is insufficient to account for all cases of challenging behaviour and does not explain how the behaviour first enters the behavioural repertoire. It is necessary to develop an understanding of other possible causes of challenging behaviour. A number of theories suggest that pain may be associated with challenging behaviour and there is a growing literature which supports this possibility. Understanding the role of pain may provide a more complete picture of what causes challenging behaviour in people with intellectual disability, which could complement the current understanding provided by operant learning theory. This could lead to the development of assessment and treatment protocols that are ethical, appropriate and effective for challenging behaviour.

The first step to understanding the role of pain in challenging behaviour is to review current research. Secondly, methodological strategies need to be identified or developed to overcome the difficulties of assessing pain in non-verbal individuals. These methods then need to be used to provide further evidence of the association between pain and challenging behaviour and to describe the nature of the relationship between these variables.

CHAPTER 2

PAIN AND CHALLENGING BEHAVIOUR IN PEOPLE WITH INTELLECTUAL DISABILITY; A SYSTEMATIC REVIEW OF CURRENT LITERATURE

2.1 Preface

Challenging behaviour negatively impacts on the lives of many individuals with intellectual disability. It is important to investigate all possible causes of challenging behaviour in order to develop comprehensive assessment and treatment protocols, which can address the appropriate cause of challenging behaviour for each individual. Underlying pain and discomfort has been identified as a possible cause of challenging behaviour in some people, although this is relatively under-researched compared to other causes of challenging behaviour, such as operant learning processes. Investigating the association between pain and challenging behaviour in people with intellectual disability is inherently difficult because, without the option of self report, pain cannot be directly assessed. Alternative methods for identifying pain which do not rely on an individual's ability to communicate must be employed.

In order to gain an understanding of the association between pain and challenging behaviour, and to document how previous research has tackled the methodological difficulty of assessing pain in people with intellectual disability, a review of relevant literature is required. This chapter will report the results from the first systematic review of literature on the influence of pain on challenging behaviour in people with intellectual disability. Key findings from the literature will be identified and implications for future research will be discussed.

2.2 Introduction

Challenging behaviour is a significant problem for a relatively high proportion of individuals with intellectual disability (Emerson et al., 2001b; Lowe et al., 2007; McClintock et al., 2003; Poppes, et al., 2010) (see section 1.1). Once established, challenging behaviour often becomes more severe and persists over long periods of time (Chadwick, Kusel, Cuddy & Taylor, 2005; Emerson, Kiernan, Alborz, Reeves, Mason, Swarbrick et al., 2001a; Taylor, Oliver & Murphy 2011; Totsika, Toogood, Hastings & Lewis, 2008). There is a growing literature on multiple causes of challenging behaviour and it is important to ensure that all possible causes of challenging behaviour are considered when developing assessment and intervention protocols. Information acquired through the exploration of possible causes and influential factors of challenging behaviour may lead to the development of more effective intervention strategies.

There is a growing literature reporting the association between pain and challenging behaviour in people with intellectual disability (Symons, 2011). However, compared to other theories of challenging behaviour, such as the operant learning theory, this is a relatively under-researched topic. Several different theories have been proposed, which seek to explain the association between pain and challenging behaviour (see sections 1.3.6.1 to 1.3.6.4). Firstly, the opioid hypothesis suggests that the release of endogenous opioids in response to pain may be rewarding. This may then influence the persistence of self injurious behaviour in people with intellectual disability (Rojahn et al. 2008; Sandman, 1990/1991; Symons, 2011; Symons, et al., 2004). Alternatively, it has been suggested that individuals with self injury may have a higher threshold at which pain is perceived. Therefore, self injury could be shaped through operant reinforcement without the typically adverse deterrent of pain (Rojahn et al. 2008; Sandman, 1990/1991; Peebles & Price 2012). Finally, self injury may cause the perception of pain from another source to be moderated, providing relief from pain and discomfort (Melzack & Wall, 1965/1982; Woolf & Slater, 2000).

These theories all potentially explain the association between pain and self injury. However, research has also shown an association between pain and other forms of challenging behaviour such as aggression and destruction of property (Carr & Owen-DeSchryver, 2007; Embregts, Didden, Schreuder, Huitink & van Nieuwenhuijzen, 2009; Kennedy, Juarez, Becker, Greenslade, Harvey, Sullivan & Tally, 2007). Therefore, an understanding of the association between pain and challenging behaviour could have clinical implications for a range of challenging behaviours beyond self injury.

Pain is a subjective and personal experience, with emphasis typically placed on self report for the purpose of description and assessment (Van Schrojenstein Lantman-de Valk et al., 1997). Often, this is not possible in individuals with severe or profound intellectual disability or those with communication difficulties. These person characteristics are associated with challenging behaviour (Deb et al., 2001; McClintock, et al., 2003, see sections 1.2 & 1.2.2). This highlights the importance of understanding the association between pain and challenging behaviour in people with intellectual disability who are unable to communicate their discomfort.

In summary, gaining a better understanding of the multiple causes of challenging behaviour could lead to the development of more effective assessment and treatment protocols. Increasing interest has focussed on the possible influence of pain. Compared to research on other causes of challenging behaviour, such as operant learning processes, the role of pain in challenging behaviour is relatively under-researched. One step to furthering knowledge regarding the association between pain and challenging behaviour is to attain an up to date understanding of previous research in this area. Therefore, in this chapter, a review of the available literature was conducted.

2.3 Aims of literature review

The aims of this systematic literature review are to:

1. Provide a summary of research investigating the association between pain and challenging behaviour.
2. Document and evaluate the methodology employed.
3. Identify key findings.
4. Identify areas for future research.

2.4 Method: procedure

Web of Science was used to conduct a literature search for all peer reviewed articles, published in ‘all years’, detailing the association between pain and challenging behaviour in people with intellectual disability. Table 2.1 lists the search terms that were employed.

Table 2.1. Search terms for articles reporting the association between pain and challenging behaviour in people with intellectual disability

Search term	**Variations**
Aggression	Aggressive behavio*,
Self injury	Self-injurio*, self-destruc*, self-mutilat* auto-mutilat*
Pica	
Destruction property	disruption property, damage property
Anal Poking	
Self-induced vomiting	self-induced regurgit*
Challenging behaviour	Challenging behavio*, maladaptive behavio*, abnormal behavio*, problem behavio*, aberrant behavio*, behavio* disorder, externalising behavio*
Intellectual disability	intellectual disab*, learning disab*, mentally retarded, metal retard*, mental retardation, mentally handicap*, mental handicap*, developmentally disab*, developmental difficult*, intellectual impairment, intellectually impaired cognitive impair*, cognitively impaired
Pain	Painful, Discomfort, Health, health problem, health difficult*

A search initially conducted in June 2009, and then updated on August 6th 2012 yielded 2,564 results. A search of all article titles was completed and irrelevant papers excluded. The abstracts of the remaining articles were read and further exclusions were made, resulting in 51 papers of interest. These articles were read in full. Final exclusions of all irrelevant papers were made, resulting in a total of 19 papers that were included in the literature review. Table 2.2 shows the main focus of the papers excluded from this review.

Table 2.2 Article topics excluded from the literature review

Main focus of the article	Variations of this focus
Health and wellbeing of carers	Parent and carer stress linked to challenging behaviour, staff and family coping.
Service needs of people with intellectual disability.	
Non-behaviour based difficulties	Mental health problems, sleep problems, problem solving skills, language problems, sexual abuse
Challenging behaviour in populations not of interest	Typically developing individuals, individuals with Alzheimer's disease and Parkinson's disease. Individuals with anxiety, obsessive compulsive disorders, brain injury, spinal cord injuries or depression. Individuals with congenital/hereditary insensitivity to pain. Non humans,
Risk factors for intellectual disability	
Behaviour not of interest	Accidental injuries
Risk factors for challenging behaviour not of interest	Gender, level of intellectual disability, home life.
Treatment of challenging behaviour (not relating to pain)	
Description of challenging behaviour	Challenging behaviour and age, prevalence of challenging behaviour, description of behavioural phenotypes.
Assessing and measuring pain	Pain management
Only Challenging behaviour	Not linked to pain, not in people with intellectual disability
Only health problems	Not in people with intellectual disability, not liked to challenging behaviour
Type of paper	Literature review, meeting abstracts,

2.5 Results and Discussion

2.5.1 Overview of papers identified

A search for research papers investigating the association between pain and challenging behaviour was conducted. A total of nineteen papers were identified. The association between pain/health problems and challenging behaviour was the primary focus of nine papers. The influence of pain on challenging behaviour was a secondary focus, or one of many variables of interest, for the remaining ten papers. Self injury was the most widely investigated form of challenging behaviour. Eighteen of the nineteen papers reported findings relating to self injury and eleven focussed exclusively on this form of challenging behaviour.

Due to the paucity of papers identified that directly assessed the association between pain and challenging behaviour, research was also included if the focus was on painful health problems and challenging behaviour. Additionally, papers were included if the construct validity was questionable but the focus was still in the area of interest, for example, if no objective measure of pain or challenging behaviour was employed. These papers have been tabulated separately.

2.5.2 Papers which directly assess the association between pain and challenging behaviour

Table 2.3 describes eleven studies, for which a clear research aim was to investigate the association between pain and challenging behaviour and in which objective measures of pain and challenging behaviour were used.

2.5.3 Papers which assess the association between painful health problems and challenging behaviour

Table 2.4 describes four papers with a focus on the association between painful health conditions and challenging behaviour. These research studies do not measure pain. Instead, pain is inferred from the presence of the health condition.

2.5.4 Papers which do not use an objective measure of pain

Table 2.5 lists four papers relating to pain and challenging behaviour, which do not use objective measures of pain and/or challenging behaviour.

Table 2.3. Papers identified that investigate the association between pain and challenging behaviour. All papers use objective measures of pain and challenging behaviour.

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Breau, Camfield, Symons, Bodfish, MacKay, Finley & McGrath (2003)	**N= 101** **Recruitment** Recruited through a tertiary paediatric centre. **Gender** 55% male **Level of ID** 9% moderate 65% severe 18% profound 8% not available <u>SIB group</u> **Age** 10.1 <u>Non-SIB group</u> **Age** 10.3	Caregivers completed a range of measures which documented their children's challenging behaviour and pain levels.	Behaviour Problems Inventory use to rate frequency and topographies of self injury. Self injury and Self restraint checklist used to rate the frequency of self injury over the last three months. Self Injury Grid to record the location of the self injury.	Non-Communicating Children's Pain Checklist-Revised (NCCPC-R) completed twice; once retrospectively to determine how frequently the children show the behaviours listed in the NCCPC-R during an episode of pain and once following a discrete pain episode. Pain intensity rating scale ranging from no pain at all to worst pain possible for the pain episode. Pain diary.	Self injurious behaviour (SIB) was **not** related to the children's physical, mental or medical characteristics. Caregivers perceived a relationship between their child's self injury and pain. **No** significant difference between SIB group and non-SIB group in severity of pain. Children's pain behaviour did not differ significantly between children with SIB and those without. No evidence for reduced sensitivity or expression of pain in children with challenging behaviour. There was no association between pain and the frequency or severity of self injury. If chronic pain is present, SIB is likely to be near the site of chronic pain.

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Carr and Owen-DeSchryver (2007)	**N**: 11 **Recruitment** Carers nominated individuals whose challenging behaviour worsened during illness. **Age** Mean:14.5, Range: 4-20 **Gender** 82% male **Level of ID** Mild-moderate & pervasive developmental disorder: 1 Moderate-severe & autism: 5 Moderate-severe & pervasive developmental disorder: 1 Severe & autism:2 Profound: 2	Phase 1) Retrospective screening questionnaire to rate frequency and intensity of challenging behaviour on sick days and well days and also to document how ill health was identified. Individuals excluded if the challenging behaviour on sick days was not significantly higher than on well days Phase 2) Prospective questionnaire completed to rate challenging behaviour, health and pain indicators on sick days and well days.	Self Injury, aggression, property destruction, stereotypic behaviour and temper outbursts. No objective definition of challenging behaviour used.	Informants provided with a list of verbal and motor indicators of pain. Frequency of behaviours reported before medical treatment on a 7 point Likert scale ranging from 'not at all' to 'all the time'.	Scores were significantly higher (P< .001) on sick days compared to well days for vocal and motor behaviour and frequency and intensity of challenging behaviour. The composite pain index was significantly higher on sick days compared to well days (P<.001). The higher the composite pain index, the greater the frequency (P<.05) and intensity (P<.01) of problem behaviour.
Carr, Smith, Giacin, Whelan & Pancari (2003).	**N**: 4 **Recruitment** Care staff identified individuals with challenging behaviour that worsened during menses and also when they were asked to complete a demand. This change in behaviour was verified with the use of community residence logs and menstruation charts. **Age-** 26-31. **Level of ID** Moderate- severe.	Menses was defined as the two days prior to menstrual flow and including the last day of menstrual flow. Functional assessment of challenging behaviour in response to demands. ABAB design during times of menses and non-menses. Implementation of multi-component intervention strategy. Medical and behavioural strategies.	Aggression, self injury and 'tantrums' operationally defined and strong inter-observer reliability obtained.	Staff reports taken of visible signs of pain during menses e.g. individual holding their abdomen or saying 'my stomach hurts'.	Challenging behaviour during demands was higher during menses compared to non-menses, suggesting pain may act as a setting event. Multi-component intervention using non-drug treatment (e.g. heat patches) and psycho-social intervention reduced challenging behaviour during demand. Not possible to determine which aspect of the intervention was responsible for the improvement.

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Courtemanche, Schroeder, Sheldon, Sherman & Fowler (2012)	**N: 4** **Recruitment** All identified as having chronic self injury through community based programme.. **Bobby-** age 28, male. Profound ID. **Brad-** age 29, male. Profound ID. **Jack-** age 46, male, profound ID. **Mike-** age 49, male, profound ID.	Video footage collected during a time when the individual was likely to engage in self injury. Self injury and behavioural indicators of pain coded. Time based sequential lag analysis used to assess the temporal relationship between pain behaviours and self injury.	Chronic self injury- defined as occurred at least once a day, on most days for at least a year. Specific topographies of challenging behaviour listed for each participant.	Direct observations of behavioural indicators of pain. Items on the NCCPC were operationally defined and coded from video footage.	Results were mixed for different participants and different episodes of self injury. Some behavioural indicators of pain were observed prior to self injury, some after, some not at all and some both before and after the episode of self injury.
Embregts, Didden, Schreuder, Huitink, van Nieuwenhuijzen (2009)	**N**: 87 **Recruitment** 1) moderate or borderline range of ID 2) lived in the facility for at least three months 3) regularly severe aggressive behaviour 4) at least three incidents of severe aggressive behaviour during the last three months. **Age** Mean:36, Range:13-76 **Gender** 71.3 % male **Level of ID** Borderline: 27% Mild: 53% Moderate: 20%	Direct care staff completed the Questionnaire About Behavioural Function (QABF) relating to their resident's aggressive behaviour.	Aggression with clear definition.	QABF used to rate frequency of target behaviours. One sub-scale refers to behaviours which indicate physical discomfort is a function of the individual's challenging behaviour.	19.5% of individuals scored higher on the 'physical discomfort subscale of the QABF compared to the other four subscales. Physical discomfort more strongly associated with aggression in females. Frequency of aggressive incidents positively correlated with mean scores on all QABF subscales **apart** from the physical discomfort subscale.

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Hartman, Gilles, McComas, Davon & Symons (2008).	**N**: 1 **Recruitment** Child receiving hospital care for hydrocephalus **Age** 12 years **Gender** Female **Level of ID** Profound	Case report Child's self injury recorded during times of scalp protrusion and times when cranial defect was flush with the skull. (ABA design). Pain ratings also made during this time.	Hitting her head, pulling her hair, hitting her led/foot against self, objects or bed rail. No objective definition given.	Five minute video footage taken during functional assessment of self injury were rated using the NCCPC-R.	Self injury significantly higher during times of scalp protrusion (P<.01). Highest total score on the NCCPC-R associated with scalp protrusion condition. Challenging behaviour anecdotally vastly improved after revision made to the individual's ventriculoperitoneal shunt.
Kennedy, Juarez, Becker, Greenslade, Harvey, Sullivan & Tally, (2007).	**Recruitment-** recruited from two local education agencies. <u>Challenging behaviour group-</u> Individuals with behaviour disorders with sufficient intensity to warrant systematic intervention. Challenging behaviour group and no-challenging behaviour group matched on sex and ethnicity. Similar ages and level of ID. In CB group, level of ID: Moderate:56% Severe:28%, Profound:17%	Questionnaires, direct observations and health checks were carried out. Data collected measured participant characteristics, behavioural disorders and health status. Analyses were conducted to determine whether there were any differences in characteristics or health status between individuals with challenging behaviour and those without.	Aggression, self injury and destruction of property reported. Aggression measured on the Achenbach Child Behaviour Checklist/Teacher report Form 6-18 years. Direct observations of aggression, self injury and property destruction. Objective definitions used and strong inter-observer reliability obtained.	Child Health Questionnaire used to measure health problems. One subscale is on 'bodily pain'.	Children with challenging behaviour had higher levels of overall health problems. Children with challenging behaviour had higher levels of bodily pain/discomfort.

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Symons & Danov (2005)	Case study of a child with midbrain gross total tumour resection. **N:**1 **Age:** 6 years **Gender:** Male	Maternal ratings of pain and self injury taken for a period of nine days. NCCPC-R scores were then compared between times with and without self injury.	Lip biting, tongue biting/chewing and eye poking. Self injurious behaviour rated using the Self Injury Trauma Scale, which measures frequency and severity self injury.	NCCPC-R scored on two hour observations that took place three times a day for a period of nine days.	Elevated ratings of self injury were associated with elevated pain ratings (P<.05).

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Symons, Harper, McGrath, Breau & Bodfish (2009a)	**N**: 70 **Recruitment** No information **Age** *SIB group*- Mean:37.8, Range:29-51 *Control group*- Mean:35.6 Range:23-48 **Gender** 57% male **Level of ID** Moderate: 8.6% Severe: 8.6% Profound: 82.8%	Primary care staff completed an adapted (minor word changes for use with adults) Non-Communicating Children Pain Checklist for individuals with and without self injurious behaviour. Individuals with known acute medical conditions were not included.	Inclusion criteria for SIB group; 1) weekly SIB according the Behaviour Problems Inventory 2) Tissue damaging SIB using the Self-Injury Trauma Scale 3) SIB lasted at least 12 months 4) SIB requires formal behavioural/medical or psychiatric intervention Non- SIB group- 1) No mention of SIB as a focus of treatment on their charts 2) confirmed by the Behaviour Problems Inventory (SIB scale)	NCCPC-R. Ratings based on behaviour over the last week. Any mention of pain was removed from the NCCPC-R to keep observers naïve to the purpose of the questionnaire. 'Activity' items did not correlate with mean NCCPC-R score so these items were removed from analyses.	SIB group had significantly higher modified NCCPC total scores than the non-SIB group (p= .004). All subscales scores were higher in the SIB group compared to the non-SIB group. This difference was significant in the vocal subscale (p= .009) eating/sleeping subscale (p= .026) and the social/personality subscale (p= .0005). For the SIB group, there was a significant positive association between NCCPC total score and overall severity of SIB (Behaviour Problems Inventory SIB subscale score) (p<.05).

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Symons, Harper, Shinde, Clary & Bodfish (2010)	**N**: 44. **Recruitment** Convenience sample recruited from a regional residential care facility. **Age** Mean: 46, Range: 28-67, SD: 9.87 **Gender** 52% male **Level of ID** Moderate: 13.64% Severe: 6.82 % Profound: 79.55%.	Five different modalities of sensory stimuli (including pin prick for pain) applied to the backs of people with ID. Facial Action Coding System (FACS) used to rate the facial indicators of pain in people with self injurious (N= 29) behaviour and those without self injurious behaviour (N= 15)	Presence of SIB confirmed by; 1) Behaviour Problems Inventory 2) tissue damage, as determined by the Self Injury Trauma Scale, 3) SIB occurred for at least the last 12 months, 4) SIB requires behavioural/medical or psychiatric intervention	16 Facial Action Units (FAU) were used from the FACS to rate response to pain episodes. Coders were blind to the condition (pain, other sensory stimuli, no sensory stimuli). Reliability between three FACS coders evaluated (76%).	Overall main effect of SIB on overall frequency of Facial Action Units during active trials, i.e. during sensory stimulation including pain, (P<.05). M=1.24 (no SIB) M= 2.1 (SIB). Increased sensory sensitivity and expression, including pain, in people with SIB compared to those without.
Symons, Wendelschafer-Crabb, Kennedy, Heeth & Bodfish (2009b).	**Recruitment** Recruited from a residential population at a tertiary care facility. **Level of ID** profound mental retardation: 80%. <u>*SIB group-*</u> **N**: 16 **Mean age**:41, **Gender:** 65% male <u>*Control group-*</u> **N**: 9 **Mean age**:38 **Gender**67% male	Typical sensory testing procedure was carried out using five different modalities of sensory stimuli (pin-prick, warm, cool, deep pressure and light touch). Coders used specific pain FAUs from FACS to measure the response of participants during sensory testing. Responses were compared between individuals with and without SIB.	Self injury; 1) Occurred at least daily (based on staff report, behaviour ratings scales and corroborated though direct observation- no specific details given) 2) Has been present for at least 12 months 3)was being treated	FAUs selected from prior work on pain expression in non-verbal individuals with intellectual disability were selected and these were used to code the expression of individuals during sensory testing. Good inter observer reliability.	Response (measured with FAUs related to pain expression) to tactile stimulation was higher in individuals who have SIB compared to individuals who do not (P<= .003).

Table 2.4. Papers identified that investigate the association between pain and challenging behaviour. Studies where pain is inferred from health problems typically associated with pain and discomfort.

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Bosch, Van Dyke, Milligan Smith, Poulton (1997).	Two part study. Part One: Description of 25 individuals referred to bio-behavioural service. Part two: Three case studies. **JA-** Female, age 8. Sever-profound ID. **JD-** Female, age 35, Severe-profound ID. **DW-** Male, age 9, severe ID.	**JA-** experimental functional assessment of challenging behaviour before and after medication change. **JD-** Medication introduced in an ABA design. Phone interviews regarding challenging behavoiur with parents. **DW-** Behaviour recorded before and after medication.	**JA**- self injury (topographies stated) **JD**- self injury and aggression (topographies stated) **DW**- self injury (topographies stated).	**JA**- previously undiagnosed ulceration and inflammation of the oesophagus, stomach and duodenum. **JD**- previously undiagnosed hiatal hernia with gastro-oesophageal reflux, delayed gastric emptying and a duodenal ulcer. **DW**- previously undiagnosed with constipation.	Part one: 28% individuals had at least one untreated medical condition. 6/7 improved SIB after treatment. Part two: **JA**- SIB occurred 25% prior to treatment and 1% of free play following treatment. **JD-** challenging behaviour reduced with treatment and then increased when treatment removed. **DW**- challenging behaviour reduced with treatment.

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Carr & Blakeley-Smith (2006)	**Recruitment** Children identified by teachers and parents if challenging behaviour worsened when ill. **Medical intervention group** **N** 11 **Age** Mean:7 Range: 5-10 **Gender:**82% male **Medical and behavioural intervention group** **N**= 10 **Age** Mean:7.7 Range: 3-11 **Gender:**90% male	Two groups; medical intervention only or medical plus behavioural intervention. Medical intervention- visit school nurse. Treatment such as heat patches, aspirin etc. All children received this care. Behavioural intervention- psychological strategies; embedding, increased choice, access to reinforcers and escape extinction plus prompts. CB ratings made on sick days for all participants.	Most serious challenging behaviour- aggression, self injury and property destruction. Less serious challenging behaviour- irritability, whining, stomping feet, defiance, uncooperative behaviours, yelling and disturbing others. Irritability subscale of the Aberrant Behavior Checklist-community. A questionnaire designed this study describing frequency of CB. No known psychometric properties.	Illness checklist. Designed for this study. Observers record symptoms and severity of illness. No known psychometric properties.	All challenging behaviour significantly lower in medical plus behavioural intervention group compared to the medical only (p<.001). Significantly more academic tasks completed by medical plus behavioural intervention group compared to medical only (P<.05). This evidence supports the idea that ill health can act as a setting event.

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Hall, Arron, Sloneem & Oliver (2008)	Questionnaire data: **N: 153** Observation data: **N: 54** **Recruitment** Teachers and key workers identified individuals suitable for the study. **Cornelia de Lange syndrome (CdLS) group** **Age** Mean:13.88 , SD: 8.58 **Gender** 46% male **Level of ID** (measures with the Wessex) Able: 13% Partly able: 41% Not able: 46% **Comparison Group** **Matched for age, gender and level of disability.**	Questionnaire data were collected. Some participants were visited by researchers at their usual day care settings. The groups were compared in terms of their health and sleep problems and their challenging behaviour.	Challenging Behaviour Interview used to rate self injury.	Health questionnaire measured presence and severity of 15 health problems. The tenth Revision of the International Statistical Classification of Disease and Related Health Problems was used to classify health problems. Good psychometric properties of the Health Questionnaire. Health problems recorded; Eye, ear, dental, gastrointestinal, bowel, heart, genitalia, hernia, lung, liver and skin. Cleft palates, hernias, epilepsy and diabetes.	CdLS group; -Individuals with skin problems more likely to show SI compared to individuals without skin problems and SI was significantly more severe. -Individuals who did not have eye problems were significantly more likely to show self injury compared to people who did Comparison group; No significant differences were observed in SI between those with and without skin problems.
Taylor, Rush, Hetrick & Sandman (1993)	**N** 9 **Recruitment** Women identified as having varying levels of SIB. **Age** 16-38 **Gender** 100% female **Level of ID** 7 profound, 1 severe, 1 moderate.	Daily records of frequency of SIB episodes recorded. Catamenial records also collected.	SIB episodes reported by primary caregivers. An episode did not vary depending on number or behaviours or duration. Inter-observer reliability for SI episodes was good.	6/9 participants previously diagnosed as having pain during menses and 5 regularly received analgesics during this time.	SIB elevated during early and late follicular phases of menses, which is typically associated with pain and when analgesics are typically given.

Table 2.5. Papers identified that investigate the association between pain and challenging behaviour. No objective measure of pain and/or challenging behaviour.

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Donovan (2002)	**N**: 8 **Recruitment** Snowball sample of learning disability nurses.	Unstructured interviews were conducted with learning disability nurses. They were asked 'can you tell me about a time that you recognized that a client was in pain?'	Self injury and aggression as described by nurse reports. Nurse reports.	Nurses reported behaviour that indicated a client was in pain. Behavioural indicators of pain included forms of self injury and aggression.	Nurses described that they may use self injury and/or aggression as behavioural indicators that a client is in pain.
Symons, Sutton & Bodfish (2001)	**N**: 4 **Recruitment** Recruited from a large public residential facility for individuals with severe and profound mental retardation. **Age** 27, 41, 44 and 45. **Level of ID** 100% profound	10 week randomized ABAB double blind, placebo-controlled experimental trial with naltrexone hydrochloride. Temperature of the primary and secondary self injury site and non-self injury site was taken during each condition Temperature may be indicative of nerve injury or disease and therefore be associated with pain or discomfort.	Self injury. Different topographies stated but no objective definition.	Temperature of the primary self injury site was taken during each condition along with the temperature of secondary self injury sites and non-self injury sites.	Temperature of primary self injury site was reduced during naltrexone. Secondary self injury sites and non-self injury sites were not associated with reduced skin temperature. SI reduced by 51%, 54%, 18% and 33% for the participants during naltrexone.

Authors	Method		Measures of assessment		Key findings
	Participants	Procedure	Challenging behaviour	Pain	
Fisher, Bowman, Thompson, Contrucci (1998)	**N**: 1 **Recruitment** Recruited following admittance to an inpatient unit specialising in the assessment and treatment of challenging behaviour **Age** 25 **Gender** male **Level of ID** Profound.	Case study looking at the change in self injurious behaviour following transcutaneous electrical nerve stimulation.	Self injury resulting in 'contusions, lacerations and auricular hematomas'.	Not measured. However, transcutaneous electrical nerve stimulation (TENS) is a treatment offered for pain relief. TENS could provide sensory stimulation or cause the release of endogenous opiates.	Self injury markedly decreased during transcutaneous electrical nerve stimulation compared to non- transcutaneous electrical nerve stimulation control conditions.
Poppes, van der Putten & Vlaskamp. (2010)	N: 181 **Age** mean: 35; SD 19. **Gender** 56% male **Level of ID** Profound	Questionnaires completed by direct care professionals.	Self injury, aggression and stereotyped behaviours defined by the BPI-01.	A questionnaire was given to direct care professionals. Items included chronic pain. The name or psychometric properties of the questionnaire not provided.	No significant difference in chronic pain scores between individuals with and without challenging behaviour.

2.5.5 Key findings

The majority of papers (seventeen out of nineteen) identified support the notion that pain can be positively associated with challenging behaviour and, in particular, self injury. It was also noted that pain could influence qualitative features of self injury. For example, individuals in pain may have more severe (more frequent and a higher intensity) self injury than individuals not in pain (Carr & Owen-DeSchryver, 2007) or the self injury may be targeted at locations associated with chronic pain (Breau et al., 2003).

A second finding was that pain-related facial expressions of individuals with self injury, in response to painful stimuli, were not different compared to pain-related facial expressions of individuals who did not self injure (Symons et al., 2009b; Symons et al., 2010). This indicates that some individuals who engage in self injury do not have reduced pain perception and therefore signifies that self injury cannot be explained by a reduced sensitivity to pain in these individuals (Sandman 1990/1991; Rojahn et al. 2008).

The findings from these papers also demonstrate that although pain is associated with challenging behaviour, it was not the only influential variable (Embregts et al., 2009). Intervention strategies for reducing challenging behaviour were shown to have the most success if they incorporated both medical and behavioural strategies (Carr et al, 2003; Carr & Blakeley-Smith 2006). The role of pain as a setting event/ establishing operator was also reported (Carr et al, 2003; Carr & Blakeley-Smith 2006; Taylor et al., 1993). A setting event alters a pre-existing behaviour-outcome relationship. For example, an individual may have learned, through operant reinforcement, an association between challenging behaviour and the removal of demands. During times of pain, the motivation to have demands removed may be higher. Therefore, the presence of pain, in combination with an environmental setting such as a demanding task, may make challenging behaviour more likely to occur.

2.5.6 Methods identified for investigating the association between pain and challenging behaviour; strengths and limitations

Individuals with intellectual disability are often unable to self report pain. Therefore, other methodological approaches must be employed that negate the need for self report. The research identified in this systematic literature search has provided examples of effective methodological approaches for investigating pain in people with intellectual disability, which can inform future research practice. Three main methods were identified. Firstly, direct observations of behavioural indicators of pain were used (Breau et al., 2003; Carr & Owen-DeSchryver, 2007; Courtemanche et al., 2012; Hartman et al., 2008; Symons & Danov, 2005; Symons et al., 2009a; Symons et al., 2009b; Symons et al., 2010). Secondly, pain was inferred from the observation of typically painful health problems (Bosch et al., 1997; Hartman et al., 2008; Taylor et al., 1993). Finally, the impact of alleviating pain on challenging behaviour was investigated (Bosch et al., 1997; Fisher et al., 1998).

The most widely employed observational measure of pain was the Non-Communicating Children's Pain Checklist- Revised (NCCPC-R) (Breau et al., 2003; Courtemanche et al., 2012; Hartman et al., 2008; Symons & Danov, 2005; Symons et al., 2009a), which was used by five of the nineteen studies. The NCCPC-R is a 30 item questionnaire, which asks informants to rate the frequency of behavioural indicators of pain over a two-hour observation window. The NCCPC-R was used in a number of different ways. Scores were compared between individuals with and without self injury (Breau et al., 2003) or at times when self injury had occurred compared to not (Symons & Danov, 2005; Symons et al., 2009a). Additionally, the NCCPC-R was used to provide an objective measure of pain during times of ill health (Hartman et al., 2008). In one study, items on the NCCPC-R were operationally defined to provide a list of behaviours that could be coded during live observations (Courtemanche et al., 2012). This provided a second-by-second account of the expression of behavioural indicators of pain and enabled researchers to explore the temporal association between pain and self injurious behaviour. Originally, the NCCPC-R was designed to be based on observations lasting two hours, although the research identified in this literature search demonstrated that the NCCPC-R can be used reliably when adapted for use in live coding (Courtemanche et al., 2012) or when ratings are based on observations made over a one-week time period (Symons et al., 2009a). Pain was also inferred from the coding of Facial Action Units (Symons et al., 2010; Symons et al., 2009b). The Facial Action Coding system was used to investigate the pain response in individuals with self injury compared to those without self injury. In these studies, pain was induced with a pin-prick.

The use of direct observational measures, which rate the frequency or presence of behavioural indicators of pain, such as the NCCPC-R and the FLACC have been shown to be a valid and reliable method for identifying pain (Breau et al., 2002; Breau, McGrath, Camfield, Rosmus, & Finley, 2000; Malviya et al., 2006; Voepel-Lewis, Merkel, Tait, Trzcinka & Malviya, 2002) (see also sections 4.3.3.7 & 5.3.3.1). The FLACC consists of five subscales (Face, Legs, Activity, Cry, Consolability). Each subscale describes a collection of behaviours which are indicative of pain. A score of zero on each of the subscales refers to no pain-related behaviour (e.g. face subscale includes 'no particular expression or smile') and a score of two on each subscale, the maximum score, includes behaviours which are indicative of high levels of pain (e.g. Face subscale includes 'constant quivering of the chin, clenched jaw). The score on each of the subscales are summed to give a total between zero and ten, zero indicating no or low level pain and a higher score representing a higher number of behavioural indicators of pain. In one study identified in this literature search, the reliability of the observer ratings such as those discussed here was questionable as assurances were not given that ratings were conducted independently. In addition to this, observers were not blind to the research hypothesis, increasing the potential for observer bias (Carr et al, 2003). Therefore, it is important for future research to control for these confounding variables to ensure the integrity of the research.

The second method for investigating the association between pain and challenging behaviour was by identifying health problems that are typically associated with pain. There were several variations of this method. Challenging behaviour was assessed on 'sick days' compared to 'well days' (Carr & Owen-DeSchryver, 2007; Carr et al, 2003; Taylor et al., 1993) or compared between individuals with and without health problems (Bosch et al., 1997; Hall et al., 2008; Kennedy et al., 2007). Reversal designs were also used, which demonstrated how challenging behaviour fluctuated with the improvement and deterioration of health (Hartman et al., 2008).

When inferring pain from health problems, researchers must assume that participants have a typically functioning pain response and therefore experience the health complaints in a typical way. In an effort to validate this assumption, Carr and Owen-DeSchryver (2007) incorporated the use of the NCCPC-R alongside the diagnosis of a health condition, which provided an objective measure of pain. In this study, Carr and Owen-DeSchryver (2007) demonstrated higher ratings of behavioural indicators of pain on 'sick days', which were defined as days with observable (e.g. diarrhoea) or measurable (e.g. a high temperature) signs of ill health. This indicates the validity of using health to signify the possibility of pain. Also, given the difficulties inherent in identifying pain in people with intellectual disability, the inference of pain through identification of health problems, although problematic, is a valuable methodological tool that should not be overlooked.

Finally, the impact of treating pain on challenging behaviour was assessed. In some studies, treatment was targeted directly at pain reduction with the use of transcutaneous electrical nerve stimulation (Fisher et al., 1998). In other studies, treatment was provided for ill health and therefore any impact on pain would be indirect and assumed (Bosch et al., 1997; Carr et al, 2003; Carr & Blakeley-Smith, 2006). A number of limitations were identified when investigating the association between pain and challenging behaviour by treating pain and/or health problems. Without an objective measurement of pain before and after treatment, it is not possible to conclude with certainty that the medical interventions impacted on the pain, or, even if it did, if this was responsible for the change in challenging behaviour. The improvements observed in challenging behaviour when 'poor health' was treated could be influenced by confounding variables. For example, sensory stimulation provided by heat packs (Carr et al, 2003; Carr & Blakeley-Smith, 2006) or social attention provided by visiting the school nurse (Carr & Blakeley-Smith, 2006) could also potentially influence challenging behaviour. In addition to this issue, many of the research studies identified in the literature search implemented multiple intervention strategies simultaneously, often combining medical and behavioural treatments (Carr et al, 2003; Carr & Blakeley-Smith, 2006). In these circumstances, it is not possible to infer which intervention, pain or non-pain related, was responsible for the change in challenging behaviour.

In several of the research papers identified, participants were specifically selected based on prior information that their challenging behaviour was influenced by pain or discomfort (Carr & Blakeley-Smith, 2006; Carr & Owen-DeSchryver, 2007; Carr et al, 2003), which means it is not possible to generalise the findings to the wider community of individuals with intellectual disability. Although these studies do provide a strong proof of principle, it is not possible to infer to what extent pain influences challenging behaviour in the wider population of individuals with intellectual disability. However, it is not assumed that pain influences all challenging behaviour in all people. The important message from these studies is that pain is associated with challenging behaviour in some individuals, and using this 'proof of principle' methodology is sufficient to uphold this assumption.

In a number of studies identified in this literature search, a lack of objective definitions for challenging behaviour (Carr & Owen-DeSchryver, 2007; Hartman et al., 2008; Symons et al., 2001) and/or pain limited the impact of the research findings. It is important for future research to clearly define what is meant by challenging behaviour and to provide an account of how pain was measured and defined. This is necessary for the research to be replicable, which would allow the reliability of the findings to be examined.

2.5.7 Implications for research

Health problems can provide an indirect indicator of underlying pain (Bosch et al., 1997; Hartman et al., 2008; Taylor et al., 1993). Some health problems are reliably reported to be associated with pain in people without intellectual disability (e.g. otitis media, gastro-oesophageal reflux). These medically identifiable health problems could provide a proxy measure for pain in people with intellectual disability. In chapters 3 and 4, groups will be identified in which pain is inferred from health conditions. In chapter 3, individuals with behavioural indicators of Gastro-oesophageal Reflux Disorder will be identified with the use of an observational questionnaire. Gastro-oesophageal reflux is typically associated with a painful 'burning' sensation in the throat and chest. In chapter 4, individuals with Tuberous Sclerosis Complex (TSC) will be recruited to represent a high risk group for pain. The physical phenotype of TSC includes benign growths in all organs of the body (Crino, Nathanson & Henske, 2006), which can lead to a number of painful health complaints. Challenging behaviour will be assessed in these groups at high risk of pain compared to more low risk populations.

Results from this literature search also demonstrate the value of using observational measures such as the NCCPC-R as a valid and reliable way to identify pain in people with intellectual disability (Breau et al., 2003; Courtemanche et al., 2012; Hartman et al., 2008; Symons & Danov, 2005; Symons et al., 2009a). In chapters 4 and 6, pain measures, which employ the observation of behavioural indicators of pain, will be used to examine the difference in pain between individuals with and without challenging behaviour.

Finally, research findings show that live coding of behavioural indicators of pain and challenging behaviour allow the temporal association of these two factors to be investigated (Courtemanche et al., 2012). In chapter 7, the temporal relationship between pain and self injury in a group of individuals with challenging behaviour regarded as ‘non-functional’ according to experimental functional analysis results will be investigated.

Therefore, this literature search identified key methodological approaches for investigating the association between pain and challenging behaviour. These findings will guide the design of studies described in this thesis, which aim to add to the currently limited understanding of the association between pain and challenging behaviour. A greater understanding of the possible influence of pain could provide an explanation for challenging behaviour that cannot currently be explained by other processes such as operant learning. Converging new information regarding the influence of pain, with the established literature on operant causes of challenging behaviour, could provide a more comprehensive approach for assessing and treating challenging behaviour in all people with intellectual disability.

CHAPTER 3

THE ASSOCIATION BETWEEN GASTRO-OESOPHAGEAL DISTRESS AND CHALLENGING BEHAVIOUR IN PEOPLE WITH INTELLECTUAL DISABILITY

3.1 Preface

Challenging behaviour in people with intellectual disability is potentially associated with pain and person characteristics such as low mood and hyperactivity (see sections 1.2.4 & 1.2.6). Understanding the relationship between these variables has important implications for intervention. The personal and subjective nature of pain means that there are inherent difficulties involved with assessing pain in people with poor expressive communication skills. A review of previous research identified a number of strategies for overcoming this problem (see section 2.5.6). Pain can be inferred from the presence of health conditions or behavioural indicators can be used to signify pain. In this chapter, behavioural indicators of Gastro-oesophageal Distress (GD), linked to Gastro-oesophageal Reflux Disease will be used to identify individuals likely to be experiencing pain. The association between GD, challenging behaviour, low mood and hyperactivity will also be assessed. Results are discussed with reference to different theories of challenging behaviour, in particular the view that challenging behaviour could be an atypical symptom of depression.

3.2 Introduction

The proportion of individuals experiencing health problems is higher in people with intellectual disability compared to the general population (Jansen et al., 2004; van Schrojenstein Lantman-de Valk, Metsemakers, Haveman et al., 2000; van Schrojenstein Lantman-de Valk et al., 2007). These health issues often go unrecognized and untreated (Howells, 1986). The 'Six lives: the provision of public services' report by the Parliamentary and Health Service Ombudsman (2009, page 29) commented on 'unnecessary distress and suffering' of individuals with intellectual disability in the health service and stated that

> 'the understanding of the issues and the focus on the quality of care [for people with intellectual disabilities] is at best patchy and at worst an indictment of our society' (page 30).

An independent inquiry launched in response to the 'Death by Indifference' report by Mencap found evidence of 'a significant level of avoidable suffering due to untreated ill-health' in people with intellectual disability (Michael & Richardson, 2008). This neglect within the Health Service is purported to be the consequence of poor training and a lack of valid and reliable health and pain assessment tools (Malviya, Voepel-Lewis, Merkel, & Tait, 2005). Researchers are addressing this latter problem by investigating behavioural correlates of health problems and pain in people with intellectual disability. These behaviours can then be used in observational health and pain assessment tools (Breau, Camfield, McGrath, Rosmus & Finley 2001; Breau et al., 2000; Craig, Hadjistavropoulos, Grunau & Whitfield 1994; Feldt, 2000) (see sections 2.5.6, 3.3.3.1, 4.3.3.7 & 5.3.3.1).

One health condition experienced by many people with intellectual disability is Gastro-Oesophageal Reflux Disease (GORD). GORD is the recurrent return of stomach acid into the oesophagus, which results in discomfort and is typically associated with a burning pain in the throat and chest and can be of such intensity that it is sometimes confused for a heart attack

(Fass & Dickman, 2006). The prevalence rate of GORD is approximately 50% in individuals with profound intellectual disability compared to 10% to 20% in the general population (Bohmer et al., 1999). In people with ASD, prevalence rates of GORD are reportedly as high as 70% (Horvath, Papadimitrion, Rabsztyn, Drachenberg & Tildon, 1999), although other research has found that individuals with ASD are no more likely to experience gastro-oesophageal problems than any other population group (Horvath & Perman, 2002). This discrepancy could potentially be accounted for by differences in recruitment strategies in these two studies. For example, Horvath et al., (1999) selected children with pre-existing symptoms including abdominal pain, diarrhoea and bloating whereas the estimates produced by Horvath and Perman (2002) are based on a population study. High prevalence rates of GORD have also been reported in genetic syndromes associated with intellectual disability. For example, Cri du Chat syndrome (CDC) (Wilkins, Brown, Nance & Wolf, 1983) and Angelman syndrome (Clayton & Smith, 1993) are both commonly associated with GORD and over 90% of individuals with Cornelia de Lange Syndrome have GORD (Kline, Krantz, Sommer, Kliewer, Jackson, FitzPatrick, et al., 2007; Luzzani et al., 2003).

In the majority of studies relating to GORD in people with intellectual disability, researchers have relied upon the use of invasive medical procedures such as endoscopies in order to diagnose GORD (Bohmer et al., 1999; Horvath et al., 1999; Luzzani et al., 2003). Although the use of this methodology ensures good construct validity, the procedure requires the expertise of medical professionals, involves local or general anaesthetic and can also result in side effects such as abnormal reaction to sedatives or accidental puncture of the upper gastro oesophageal tract (Sieg, Hachmoeller-Elsenbach & Elsenbach, 2001). Due to the human cost of these procedures, they are only performed when GORD is already suspected. Therefore it is possible that the prevalence rates of GORD are underestimated. In general medical practice, this reliance on invasive procedures to identify GORD may result in many cases of GORD going unrecognized and untreated in people with intellectual disability who have not been tested for gastro-oesophageal abnormalities. It is clear that there is the need for non-invasive GORD assessment tools that can be available for all people with intellectual disability to help identify previously unrecognised cases of GORD. This would help identify those at risk for GORD and thus make the human cost of an invasive procedure justifiable.

The Gastro-intestinal Distress Questionnaire (GDQ; Oliver & Wilkie, 2005) assesses the frequency of a number of behaviours which are potentially indicative of discomfort in the oesophagus and stomach. The GDQ has been validated for use with children and adults with Cornelia de Lange syndrome; a syndrome known to be strongly associated with gastro-oesophageal reflux (Richards & Oliver, in preparation). In this study, Richards and Oliver (in preparation), assigned a cut-off to describe a behaviour as being 'clinically significant'. Five or more 'clinically significant' behaviours were used to indicate if an individual currently had, or previously had, GORD. When used in this way with individuals with Cornelia de Lange syndrome, the GDQ was shown to have discriminative validity for identifying individuals who currently had or have ever had GORD (Richards & Oliver, in preparation). Parental reports were used to verify the presence of current or past episodes of GORD.

Non-invasive assessment tools of health and pain generally rely on the observation of behaviour (Breau, McGrath, Finley, & Camfield, 2004; Merkel, Voepel-Lewis, Shayevitz & Malviya, 1997). Specifically, two areas of behaviour have been shown to be strongly associated with health and pain: low mood and levels of activity (Berg et al., 2007, Merkel et al., 1997; Voepel-Lewis et al., 2002). Berg et al. (2007) found that individuals with intellectual disability who had health problems scored significantly lower on both 'mood' and 'interest and pleasure' subscales on the Mood, Interest and Pleasure Questionnaire - Short version (MIPQ-S) compared to individuals without health problems. Low mood has been incorporated into pain assessment tools. The NCCPC-R (Breau et al., 2004) uses ‘unhappy and cranky’ as a behavioural indicator of pain and the FLACC (Merkel et al., 1997) lists ‘withdrawn and sad’ as facial expressions indicative of pain.

High activity levels have also been linked to health problems. Luzzani et al. (2003) identified ‘Hyperactivity’ in 85% of the children with Cornelia de Lange Syndrome with GORD, which was significantly higher than the occurrence of ‘typical’ symptoms such as vomiting or regurgitation. High levels of activity are also included in validated pain assessment tools. ‘Fidgety, agitated and jumping around’ is listed as a behavioural indicator of pain in the Non-Communicating Pain Checklist- Revised (NCCPC-R) (Breau et al., 2004) and the Face, Legs, *Activity,* Cry, Consolability (FLACC) (Merkel et al., 1997) lists numerous behaviours linked to high activity levels such as ‘squirming’ and ‘severe agitation’.

In addition to high levels of activity and low mood included in pain assessments, challenging behaviour can also be a behavioural correlate of poor health. Carr and Owen-DeSchryver (2007), for example, demonstrated that the level of challenging behaviour (self injury, aggression, property destruction, stereotypic behaviour and tantrums) in a group of children with intellectual disability was significantly higher on ‘sick days’ rather than ‘well days’. The sick days were defined by either an observable (e.g. diarrhoea) or measurable (e.g. temperature) indication of poor health.

This summary of relevant research indicates that challenging behaviour, low mood and hyperactivity are all associated with pain and it follows that a painful condition such as GORD could also be associated with these behaviours. Consequently, individuals engaging in behaviours indicative of GORD would be expected to show higher levels of challenging behaviour and activity and lower mood as found in these previous studies. In this study it is predicted that challenging behaviour, hyperactivity and low mood will all be positively associated with behavioural indicators of GORD, identified through use of the GDQ.

In the last decade there has been increasing interest in the association between low mood and challenging behaviour (Davis, Judd & Herman, 1997; Hayes, McGuire, O'Neill, Oliver & Morrison, 2011; Ross & Oliver 2002b; Tsiouris, 2001) (see section1.2.6). One explanation for this association is that challenging behaviour is an atypical symptom of depression in people with severe intellectual disability (Marston et al., 1997; Meins, 1995). However, both low mood and challenging behaviour have been linked independently to pain and discomfort. Therefore, it is possible that the presence of pain/discomfort, rather than depression, could account for the association between low mood and challenging behaviour. For example, research within the general population has shown that high levels of pain are linked to low mood and even suicidal behaviour (Affleck, Tennen, Urrows, Higgins, 1992; Feldman, Downey, Schaffer-Neitz 1999; Walters, 1990). Several case studies document low mood and depression in individuals with intellectual disability suffering with chronic pain (Davis et al., 1997). Finally, research has repeatedly shown an association between health problems/discomfort and challenging behaviour (Carr & Owen-DeSchryver, 2007; Horvath & Perman, 2002).

Within this study, there is the opportunity to explore whether individuals with low mood and challenging behaviour engage in other behavioural indicators of pain, such as high levels of activity. If so, this would suggest individuals with low mood and challenging behaviour are more likely to be in pain compared to individuals with more positive mood or those not engaging in challenging behaviour. Therefore, underlying pain and discomfort would explain the association between low mood and challenging behaviour in these people. Alternatively, if individuals with low mood and challenging behaviour do not engage in other behavioural indicators of pain and therefore, the association between low mood and challenging behaviour cannot be explained by the influence of pain, it would support the alternative assumption that challenging behaviour may be a symptom of depression in people with intellectual disability. Given the evidence reviewed in the literature review in chapter 2, which reported an association between pain and challenging behaviour, it is predicted that individuals with low mood and challenging behaviour will be more likely to have higher levels of activity compared to individuals not engaging in challenging behaviour or evidencing low mood, which would imply that low mood and challenging behaviour, as for high activity levels, are behavioural indicators of pain.

In summary, previous research has demonstrated high prevalence rates of GORD in people with severe intellectual disability. There is a need for an assessment tool to identify likely GORD without the use of invasive medical procedures. This study will use an observational assessment tool, the GDQ, to identify behavioural indicators of GORD in two high risk groups, Cri du Chat syndrome and ASD. Previous research has shown that GORD is associated with hyperactivity and challenging behaviour and that poor health is linked to low mood. This study will assess whether these known associations are evident when individuals are identified as showing behavioural indicators of GORD. Using a cut-off of five or more clinically significant behavioural indicators of GORD, as measured on the GDQ, has been shown to be an effective way of identifying individuals with GORD (Richards & Oliver, in preparation). It is predicted that individuals identified as showing five or more behavioural indicators of GORD will have higher activity and challenging behaviour levels and lower mood compared to individuals showing fewer behavioural indicators of GORD.

Finally, previous research has shown an association between low mood and challenging behaviour. There is debate regarding the interpretation of this finding. One assumption is that challenging behaviour is an atypical symptom of depression (Marston et al., 1997; Meins, 1995). Conversely it is possible that the presence of pain might account for the association between low mood and challenging behaviour. In this study the latter is hypothesised. Therefore, it is predicted that individuals with both low mood and challenging behaviour will exhibit other behavioural indicators of pain, in this case high levels of activity.

3.2.1 Aims and hypotheses

In this study, the prevalence of gastro-oesophageal distress (GD) in two high risk groups; individuals with ASD and Cri du Chat syndrome will be assessed. The demographic characteristics of individuals with a clinically significant number of behavioural indicators of GORD (five or more) will be compared to individuals with fewer behavioural indicators of pain.

It is predicted that:

1. Individuals showing five or more behavioural indicators of GORD will have significantly higher levels of challenging behaviour compared to individuals showing fewer than five behavioural indicators of GORD. (Self injury severity will also be assessed between individuals with five or more clinically significant behavioural indicators of GORD compared to people with fewer than five clinically significant behavioural indicators of GORD).
2. Individuals showing five or more behavioural indicators of GORD will have significantly higher levels of activity and lower mood compared to individuals showing less than five behavioural indicators of GORD.
3. Individuals with low mood and challenging behaviour will also have high levels of activity.

3.3 Method

3.3.1 Recruitment and participants:

Data were collected as part of an ongoing questionnaire study at the Cerebra Centre for Neurodevelopmental Disorders (Arron et al., 2011; Moss, Oliver, Arron, Burbidge, & Berg, 2009; Oliver, Berg, Burbidge, Arron &. Moss, 2011b). Participants were aged three years and upwards with a diagnosis of ASD or Cri du Chat syndrome. Individuals with ASD were recruited through National Autistic Society branches and were screened to ensure that they scored past the cut-off (15) for ASD on the Social Communication Questionnaire (Rutter, Bailey & Lord, 2003). Diagnosis of Cri du Chat syndrome had been confirmed by a paediatrician or clinical geneticist. Participants with Cri du Chat syndrome were recruited through family support groups in the UK and Ireland or were already known to the Cerebra Centre for Neurodevelopmental Disorders and had provided consent to be contacted for future research purposes. All questionnaires were completed by parents and carers on behalf of the participants. Table 3.1 provides a description of the participants.

Table 3.1. Mean age (standard deviation) and range, percentage of males, percentage of partly able/able, fully mobile, normal vision and normal hearing and partly verbal/verbal participants.

		CDC	**ASD**
N		46	245
Age*	Mean	20.01	11.50
	(SD)	(12.29)	(5.97)
	Range	5-47	3-45
Gender	N	15	214
	Male (%)	32.6	86.3
Self ^{A} Help	Partly able/able^{B} (%)	54.3	88.7
Mobility^{A}	Fully mobile^{C} (%)	63.0	94.4
Vision^{A}	Normal (%)	89.1	95.2
Hearing^{A}	Normal (%)	87.0	96.4
Speech^{A}	Partly verbal/verbal (%)	76.1	92.3

*In years
^{A}Data derived from Wessex questionnaire (Kushlick et al., 1972; see section 3.3.3.3)
^{B}Those who score six or above on the total score of the self help subscale (items g to i)
^{C}Those who score six on the total score of the mobility subscale (items e & f)

3.3.2 Procedure

Parents and carers received a questionnaire pack, covering letter (appendix A), information sheet (appendix B) and consent forms (appendix C). Participants' parents and carers were asked to complete the questionnaire pack and consent forms and return them in a pre-paid envelope. Included in the questionnaire pack were the following measures;

3.3.3 Measures:

3.3.3.1 Gastro-oesophageal Distress Questionnaire (GDQ; Oliver & Wilkie, 2005, appendix D1)

The Gastro-oesophageal Distress Questionnaire is a seventeen-item informant based questionnaire. It is designed to rate the frequency of behaviours indicative of pain in the oesophagus and stomach in people with intellectual disability. In order to produce the GDQ, semi-structured interviews were conducted with approximately 100 parents of children with Cornelia de Lange syndrome, a genetic syndrome known to be associated with high rates of GORD (Bull et al., 1993; Luzzani et al., 2003; Sommer, 1993). Behaviours repeatedly endorsed by parents as being associated with their child's GORD related discomfort were collated to form the GDQ. When used with people with Cornelia de Lange Syndrome, the GDQ has good item level inter-rater reliability (Richards & Oliver, in preparation). Also, when used with this group, a clinically significant score on five or more items of the GDQ was shown to have discriminative validity for identifying individuals who have and had reflux since the age of three compared to those who have not ($\chi^2 = 11.01$, $p<.001$), who have ever had reflux ($\chi^2 = 8.04$, $p<.01$) and if they have reflux now ($\chi^2 = 6.20$, $p<.05$) (Richards & Oliver, in preparation). A clinically significant score for each item on the GDQ is defined as a behaviour occurring at a frequency of at least once a day, as this is likely to impact on everyday life.

For the purpose of this study, individuals who have scored at a clinically significant level on five or more items of the GDQ were referred to as being in the ***GD+*** group (gastro-oesophageal distress positive) and individuals who scored below this point were categorised into the **GD-** group (gastro-oesophageal distress negative).

There are two items on the GDQ that could confound some analyses in this study; item eight is 'Scratch, hit, press or rub around the upper chest or throat', which could be classified as a form of self injury. Item four on the GDQ is 'Fidget, wriggle or move their body a great deal', refers to level of activity. When exploring the association between gastro-oesophageal distress and self injury, item eight was excluded from the analyses and when examining the association between gastro-oesophageal distress and activity level, item four was removed. Therefore, for these analyses individuals were classified as being in the GD+ group if they scored past the assigned clinical cut-off point on five or more items on the GDQ, not including the confounding items.

3.3.3.2 Challenging Behaviour Questionnaire (CBQ; Hyman, Oliver and Hall, 2002, appendix D2)

The CBQ is a brief informant based questionnaire evaluating the presence of self injury, physical aggression, destruction of property and stereotyped behaviours over the last month. The questionnaire has been shown to have good inter-rater reliability with reliability coefficients ranging from .61 to .89 (Hyman et al., 2002). An overall self injury severity score can be calculated by summing the scores on items three, four and five on the CBQ. These items assess the duration of the longest outburst of self injury, the form of restraint required and the frequency of self injury.

Examples of challenging behaviour are provided to guide the informant to make a decision. The question referring to self injury reads;

> 'Has the person shown self-injurious behaviour in the last month? (e.g. head banging, head-punching or slapping, removing hair, self-scratching, body hitting, eye poking or pressing)'

The question referring to aggression reads;

> 'Has the person shown physical aggression in the last month? (e.g. punching, pushing, kicking, pulling hair, grabbing other's clothing).'

The question referring to destruction of property reads;

> 'Has the person shown disruption and destruction of property or the environment in the last month? (e.g. tearing or chewing own clothing, tearing newspapers, breaking windows or furniture, slamming doors, spoiling a meal).'

3.3.3.3 Wessex Questionnaire (Kushlick, Blunden & Cox, 1973, Appendix D3)

The Wessex questionnaire is a proxy measure of degree of intellectual disability. The Wessex is an informant-based questionnaire that comprises two subscales including; the Social and Physical Incapacity (SPI) scale and the Speech, Self-help and Literacy (SSL) subscale, although only the second subscale is used in this study. Reliability of this measure has been reported as percentage agreement on responses. Reliability for the overall score on the SPI scale is reported at 65%, reliability for the overall score on the SSL scales is reported at 76% (Palmer & Jenkins, 1982).

3.3.3.4 The Activity Questionnaire (Burbidge & Oliver 2008, appendix D4)

The Activity Questionnaire (TAQ) is an informant based questionnaire for use with children and adults with intellectual disability and is suitable for verbal and non-verbal individuals. The psychometric properties of the TAQ have been tested (Burbidge & Oliver; 2008; Burbidge, Oliver**,** Moss, Arron, Berg, Hill et al., 2010). Internal consistency for both the full scale and subscales was shown to be good. Each questionnaire was completed by two parents/carers of the participants for the purpose of assessing inter-rater reliability (N=125). Test-retest reliability of scores was assessed by administering the TAQ to the same informants on two occasions two weeks apart (N=103). Results show that all correlations are at .70 or above and provide evidence that at subscale and full-scale level, both inter-rater and test-re-test reliability of the TAQ is robust.

3.3.3.5 Mood, Interest and Pleasure Questionnaire- short version (Ross et al., 2008, appendix D5)

The Mood, Interest and Pleasure Questionnaire-Short version (MIPQ) examines emotional constructs in adults with severe and profound learning disability. The questionnaire is completed by the participant's carer, based on observations during the previous two-week period. The questionnaire contains two subscales; the mood subscale and the interest and pleasure subscale, based on two main constructs of depression listed in the DSM-IV. Therefore, this questionnaire uses ratings of behaviours that correlate with an individual's affective state to provide a good measure of depression in individuals who are unable to self report their emotional state. Examination of the psychometrics of the MIPQ has demonstrated good test-retest and inter-rater reliability scores with Kappa values of 0.87 and 0.76 respectively (Ross & Oliver, 2003). Internal consistency was 0.94. Evidence to support the construct validity was obtained by correlating scores with the Lethargy and Social Withdrawal scale on the Aberrant Behavior Checklist (Aman & Singh, 1986).

3.3.4 Data Analyses

Data were analysed using SPSS 18.0 software. All data were tested for normality using Shapiro-Wilk test and non-parametric tests were used where necessary. Throughout the analyses, individuals with five or more behavioural indicators of GORD (measured on the GDQ) will be categorised into the GD+ group and those with fewer than five behavioural indicators of GORD will be classified as GD-. In order to assess the prevalence of likely gastro-oesophageal distress in individuals with ASD and Cri du Chat syndrome, the percentage of individuals falling into the GD+ and GD- groups was calculated. Chi squared analyses were conducted to determine whether the proportion of challenging behaviour was significantly different between the GD+ and GD- groups. Mann-Whitney U tests were used to determine whether self injury severity differed between individuals in the GD+ and GD- groups. Mann-Whitney U tests were also used to determine whether level of mood, interest and pleasure, activity and impulsivity differed between individuals in the GD+ and GD- groups. The confounding impact of level of intellectual ability on each of these associations was assessed.

Finally, Kruskall Wallis and post-hoc Mann-Whitney U tests were conducted to determine whether individuals with a combination of low mood and challenging behaviour were likely to engage in higher levels of activity compared to individuals with just challenging behaviour, just low mood or neither of these characteristics.

3.4 Results

3.4.1 Prevalence of probable gastro-oesophageal distress in ASD and Cri du Chat syndrome

An aim of this study was to identify the prevalence of individuals likely to be experiencing gastro-oesophageal distress in two groups; people with ASD and people with Cri du Chat syndrome. The demographic variables of individuals in the GD+ group will then be compared to individuals in the GD- group. Table 3.2 shows the percentage of individuals categorised as GD+ and GD- in people with ASD and Cri du Chat syndrome. A number of demographic variables are compared between the GD+ and GD- groups.

Table 3.2. Mean age (standard deviation), percentage of males, percentage of partly able/able, fully mobile, normal vision and normal hearing and partly verbal/verbal participants with Cri du Chat syndrome (CdC) and Autism Spectrum Disorder (ASD).

		CdC				**ASD**			
		GD+	**GD-**	**t/χ^2**	***p* value**	**GD+**	**GD-**	**t/χ^2**	***p* value**
N		19	27			99	146		
(% of syndrome group)		(41.30)	(58.70)			(40.41)	(59.59)		
Age*	Mean	15.79	22.98	2.02	.05	10.21	12.46	5.18	**<.01**
	(SD)	(10.78)	(12.60)			(4.63)	(6.63)		
Gender	% Male	26.3	37.0	.58	.45	88.9	84.2	1.06	.30
Self [1]Help	% Partly able/able[2]	50.0	59.3	.38	.54	82.8	93.2	6.41	**<.05**
Mobility[1]	% Fully mobile[3]	44.4	77.8	5.24	**<.05**	92.9	95.2	.58	.45
Vision[1]	% Normal	88.9	92.6	.18	.67	93.9	95.9	.48	.49
Hearing[1]	% Normal	100	81.5	3.75	.05	93.9	97.9	2.68	.10
Speech[1]	% Partly verbal/verbal	77.8	77.8	.00	1.0	87.9	95.8	5.41	**<.05**

Significant differences between the GD+ and GD- groups for individuals with ASD and CdC are shown in **bold.**

Prevalence rates of likely gastro-oesophageal distress, indicated by five or more behavioural indicators of GORD on the GDQ occurring at a clinically significant frequency, were 41.3% in Cri du Chat syndrome and 40.41% in ASD. In the Cri du Chat group, individuals had significantly higher motor ability skills in the GD- group compared to the GD+ group. In people with ASD, participants in the GD- group were significantly more able, were older and had significantly higher speech skills compared to individuals in the GD+ group. It is important to ensure that any results are not confounded by difference in ability level. Therefore, the potential confounding effect of level of intellectual disability was investigated for each analysis (section 3.4.5).

3.4.2 Challenging behaviour and probable gastro-oesophageal distress

The first hypothesis was that the proportion of individuals engaging in challenging behaviour would be higher in individuals in the GD+ group compared to the GD- group. In order to test this hypothesis, Chi Squared (χ^2) analyses were conducted to compare the proportion of participants engaging in challenging behaviour between the GD+ and GD- groups. Four forms of challenging behaviour were assessed: self injury, aggression, stereotyped behaviour and property destruction. Relative risks were also calculated to compare the likelihood of individuals in the GD+ group engaging in challenging behaviour compared to individuals in the GD- group. Table 3.3 shows the results from this analysis.

Item 8 on the GDQ refers to scratching around the throat and chest, which could be interpreted as self injury. Therefore, this item was removed from this analysis so as to not confound the results. Therefore, the participants classified in the GD+ and GD- groups varied slightly from those described in table 3.2 (see appendix E for descriptive data on the participants included in this analysis).

Table 3.3. X^2 results comparing the proportion of challenging behaviour between the GD+ and GD- groups within syndrome groups. Relative risk of challenging behaviour in the GD+ group compared to the GD- group is also stated.

Behaviour	**Group**	**% of people showing behaviour GD+ Group (N)**	**% of people showing behaviour GD- Group (N)**	χ^2	***p* value**	**Relative Risk of Challenging Behaviour; GD+ compared to GD** ***(99% Confidence Intervals)***
Proportion of challenging behaviour in each group						
Aggression	CdC	73.7	55.6	1.57	.21	1.33
		(14)	(15)			*(.75-2.34)*
	ASD	79.4	48.9	22.38	**< .001**	**1.55**
		(77)	(68)			**(1.21-2.00)**
Property Destruction	CdC	57.9	33.3	2.74	.10	1.74
		(11)	(9)			(.73-4.12)
	ASD	74.2	33.6	37.88	**< .001**	**2.21**
		(72)	(47)			**(1.57-3.12)**
Stereotyped Behaviour	CdC	57.9	44.4	0.81	.37	1.30
		(11)	(12)			(.62-2.76)
	ASD	79.4	53.6	16.59	**< .001**	**1.48**
		(77)	(75)			**(1.16-1.89)**
Self Injury	CdC	100	75.0	5.31[A]	**< .05**	**1.33**
		(18)	(21)			**(1.01 – 1.77)**
	ADS	65.0	25.7	38.00	**< .001**	**2.93**
		(65)	(38)			**(1.93-4.43)**

[A] Fisher's Exact test used if expected cell count is less than five

X^2 test results revealed that, for the individuals with ASD, the proportion of participants who engaged in self injury, aggression, destruction of property and stereotyped behaviour was significantly higher in the GD+ group compared to the GD- group. The proportion of individuals with Cri du Chat syndrome who engaged in self injurious behaviour was significantly higher in the GD+ group compared to the GD- group. The relative risk calculations demonstrated that within the ASD group, there was a higher risk of all forms of challenging behaviour associated with membership of the GD+ group. There was a significant increase in risk of self injury associated with membership of the GD+ group for individuals with Cri du Chat syndrome. These results show that hypothesis one was upheld within the ASD group, as engaging in five or more behavioural indicators of GORD was associated with a higher proportion of individuals engaging in self injury, aggression, destruction of property and stereotyped behaviour. Within the Cri du Chat Syndrome group, hypothesis one was partially supported as engaging in five or more behavioural indicators of GORD was associated with a higher proportion of individuals engaging in self injurious behaviour.

3.4.3 Severity of self injury in people with probable gastro-oesophageal distress

The severity of self injurious behaviour was compared between the GD+ and GD- groups. Self injury severity considers the duration of the longest outburst of self injury, the form of restraint required and the frequency of self injury (see section 3.3.3.2). For the initial analysis, if self injurious behaviour did not occur the severity score was deemed to be zero. Thereafter, individuals who did not engage in self injury were excluded and the self injury severity was compared between the GD+ and GD- groups only in individuals who had engaged in self injurious behaviour in the past month. In addition to analysis of the overall self injury severity score, the contributing items were also analysed separately to see what was driving the results. Table 3.4 shows the results from these analyses.

Table 3.4. Mann-Whitney U tests assessing difference in self injurious severity between the GD+ and GD- groups. Overall self injurious severity was assessed as well as the individual items that were combined to give the overall self injury severity score.

	Group	**Median (inter-quartile range)**		**U**	***p*-value**	**Effect size (r)**
		GD+	**GD-**			
Severity of Self Injury -A score of zero is given to anyone who does not show self injury.						
Severity of self injury	CdC	6.0 (4-8.25)	3.0 (.38-4)	107.0	**<.01**	-.48 *medium- large*
	ASD	4.0 (0-7.75)	0.0 (0-3)	4332.0	**< .001**	-.39 *medium- large*
Severity of Self Injury - given that self injury has occurred. Breakdown of self injury severity components.						
Overall self injury severity	CdC	6.0 (4.0-8.25)	3.0 (3.0-5.0)	107.0	**< .05**	-.37 *medium*
	ASD	6.0 (4.0-8.75)	5 (3.5-6.0)	909.5	.05	-.19 *Small*
Level of restraint required	CdC	1.00 (0-3.0)	0.00 (0-1.0)	131.50	.08	-.28 *Small- medium*
	ASD	1.00 (0.0-2.8)	1.00 (0.0-2.0)	957.5	.10	-.16 *Small*
Duration of longest episode of Self injury	CdC	2.00 (1.0-3.0)	1.00 (1.0-2.0)	126.00	.06	-.30 *medium*
	ASD	2.00 (1.0-4.0)	2.00 (1.0-3.0)	1009.5	.20	-.13 *small*
Frequency of self injury	CdC	2.00 (1.75-3.0)	2.00 (1.0-3.0)	140.00	.15	-.23 *Small-medium*
	ASD	2.00 (2.0-3.0)	2.00 (1.0-3.0)	963.0	.11	-.16 *Small*

Bold signifies a significant difference between the GD+ and GD- groups**.**

Mann-Whitney U test results demonstrated that, when all individuals were included, overall self injury severity scores were significantly higher in the GD+ group compared to the GD- group in people with ASD and Cri du Chat syndrome. These differences were significant with a medium to large effect size in both the ASD and Cri du Chat syndrome groups. When individuals who had not engaged in self injurious behaviour in the past month were excluded from the analysis, self injury severity was significantly higher in the GD+ group compared to the GD- group in people with Cri du Chat syndrome but not the ASD group. This difference was associated with a medium effect size. No significant differences were found between scores on the individual items contributing to the overall self injury severity score between the GD+ and GD- groups in either the ASD or Cri du Chat syndrome groups.

3.4.4 Activity level and mood and probable gastro-oesophageal distress

The second hypothesis was that individuals with five or more behavioural indicators of GORD (GD+ group) would have higher levels of activity and lower mood compared to individuals in with fewer behavioural indicators of GORD (GD- group). In order to test this hypothesis, Mann Whitney U tests were conducted. The subscale scores on both the Mood Interest and Pleasure Questionnaire and The Activity Questionnaire were compared between the GD+ and GD- groups. Table 3.5 shows the results from these analyses.

When investigating activity level, item four was excluded from the GDQ as this item refers to high levels of activity. Therefore, the participants classified in the GD+ and GD- groups varied slightly from the participants described in table 3.2 (see appendix E for descriptive data on the participants included in the GD+ and GD- groups for this analysis).

Table 3.5. Mann-Whitney U test results showing difference in activity levels and mood between the GD+ and GD- groups. Effect sizes are also stated.

Subscale score	**Group**	**Median (inter-quartile range)**		**U**	***p*-value**	**Effect size (r)**
		GD+	**GD-**			
Activity level						
Impulsivity	CdC	20.0 (18.5-24.0)	15.0 (9.0-20.0)	119.0	**.02**	-.36 *Medium*
	ASD	20.0 (18.0-23.0)	15.0 (9.0-19.0)	3058.0	**<.001**	-.44 *Medium*
Hyperactivity	CdC	23.0 (12.0-30.5)	9.0 (5.0-18.0)	108.5	**<.01**	-.40 *Medium*
	ASD	28.0 (18.0-31.0)	12.5 (7.0-21.0)	2487.0	**<.001**	-.51 *Large*
Mood and interest and pleasure						
Mood	CdC	20.0 (17.0-22.0)	23.0 (20.0-24.0)	139.0	**<.01**	-.39 *Medium*
	ASD	17.2 (16.0-20.0)	20.0 (12.0-18.0)	4180.5	**<.001**	.37 *Medium*
Interest and Pleasure	CdC	18.0 (15.0-20.0)	20.0 (17.0-33.0)	182.5	.10	-.24 *Small-medium*
	ASD	13.0 (9.0-15.0)	15.0 (12.0-18.0)	5185.5	**<.001**	-.25 *Small-medium*

Bold signifies significant differences between the GD+ and GD- groups.

Individuals in the GD+ group scored significantly higher on impulsivity and hyperactivity subscales compared to individuals in the GD- group in people with ASD and Cri du Chat syndrome. All findings were associated with effect sizes ranging from medium to large. Therefore the second hypothesis that activity levels in the GD+ group would be higher than in the GD- group was upheld.

Individuals in the GD+ group had a significantly lower mood score on the MIPQ compared to individuals in the GD- group in individuals with Cri du Chat syndrome and ASD. Individuals with ASD in the GD+ group also had significantly lower interest and pleasure scores compared to individuals in the GD- group. This association was not significant in the Cri du Chat group although there was a small-medium effect size. Therefore, the findings from the ASD group were consistent with the hypothesised results that mood and interest and pleasure would be lower in individuals in the GD+ group compared to the GD- group. This hypothesis was partially upheld in the Cri du Chat syndrome group, although only when using the ‘mood’ subscale on the MIPQ.

3.4.5 Assessing the confounding influence of level of intellectual disability

Several analyses were conducted in order to test whether the associations between challenging behaviour, low mood, activity and gastro-oesophageal distress were an artefact of increased levels of intellectual disability within the GD+ group. A forced entry binary logistic regression was used to test whether the high proportion of self injurious behaviour, aggression, destruction of property and stereotyped behaviour in the GD+ group was an artefact of intellectual disability. Two independent variables were entered into this model; level of intellectual disability and gastro-oesophageal distress status (GD+ or GD-).

In order to investigate whether or not the observed association between gastro-oesophageal distress and levels of activity and mood were an artefact of increased level of intellectual disability in the GD+ group, partial correlations were conducted. The correlation between level of gastro-oesophageal distress (total number of clinically significant items of GD on the GDQ) and activity score or mood score was assessed before and after controlling for the effects of level of intellectual disability. Table 3.6 shows these results.

Table 3.6. Results from the binary logistic regression and the partial correlations assessing the association between behavioural indicators of gastro oesophageal reflux disease (GORD) with level of challenging behaviour, activity and mood when controlling for level of intellectual disability.

		Binary Logistic Regression. Level of ID (Wald)	*p* Value	Binary Logistic Regression GD status (GD+ or GD-) (Wald)	*p* Value
Self Injury present/absent	CdC	.13	.72	.00	1.0
	ASD	7.96	**<.01**	23.67	**<.001**
Aggressive present/absent	CdC	2.92	.09	.30	.59
	ASD	.00	.99	18.42	**<.001**
Stereotyped behaviour present/absent	CdC	1.17	.28	.20	.65
	ASD	3.26	.07	10.74	**<.01**
Destruction of property present/absent	CdC	.73	.39	1.55	.21
	ASD	.06	.81	31.10	**<.001**
		Correlation coefficient (r)	P Value	Partial correlation (controlling for level of intellectual disability) (r)	P Value
Self injury severity x number of behavioural indicators of GORD	CdC	.45	<.01	.42	**.01**
	ASD	.39	<.001	.28	**<.001**
Impulsivity Subscale Score x number of behavioural indicators of GORD	CdC	.37	<.05	.34	**<.05**
	ASD	.57	<.001	.50	**<.001**
Hyperactivity Subscale Score x number of behavioural indicators of GORD	CdC	.56	<.001	.53	**<.001**
	ASD	.65	<.001	.58	**<.001**
MIP; mood subscale score x number of behavioural indicators of GORD	CdC	.47	.001	.41	**<.01**
	ASD	.36	<.001	.34	**<.001**
MIP; Interest and Pleasure subscale score x number of behavioural indicators of GORD	CdC	.34	<.05	.26	.09
	ASD	.21	.001	.23	**<.001**

Bold text in the binary logistic regression, signifies which factors (level of ID or GD status) significantly increase the predictive value of the model for predicting the presence of challenging behaviour.
Bole text in the partial correlations signifies correlations that remain significant after the influence of level of intellectual disability has been removed.

Before the possible confound of intellectual ability was accounted for, it was shown that self injury was significantly more likely and more severe in people in the GD+ group compared to the GD- group in individuals with Cri du Chat syndrome and ASD. Aggression, destruction of property and stereotyped behaviour was more likely in the GD+ group compared to the GD- in people with ASD. It was also shown that levels of activity and impulsivity were higher in the GD+ group compared to the GD- group for people with both Cri du Chat syndrome and people with ASD. Also, in people with ASD and Cri du Chat syndrome, levels of mood were lower in the GD+ group compared to the GD- group. Finally, for individuals with ASD, levels of interest and pleasure were lower in the GD+ group compared to the GD- group. A binary logistic regression assessed the predictive value of GD status (GD+ or GD-), independent of the predictive value of level of intellectual disability for categorising people as having or not having self injury, aggression, destruction of property and stereotyped behaviour. For people with ASD, GD status independently predicted whether individuals engaged in all of these behaviours, above and beyond the predictive value of level of intellectual disability. In the Cri du Chat group, the binary logistic regression demonstrated that there was a non-significant predictive value of GD status for self injury, after the influence of level of ID had been removed. In the Cri du Chat syndrome group, GD status was shown to not independently contribute to the predictive model of whether or not an individual would engage in aggression, destruction of property and stereotyped behaviour. This was expected due to the non-significant findings reported in table 3.3. In both the ASD and Cri du Chat syndrome groups, level of intellectual disability was shown not to independently predict whether an individual would engage in aggression, destruction of property or stereotyped behaviour. In the ASD group, level of intellectual disability was a significant predictor of self injurious behaviour.

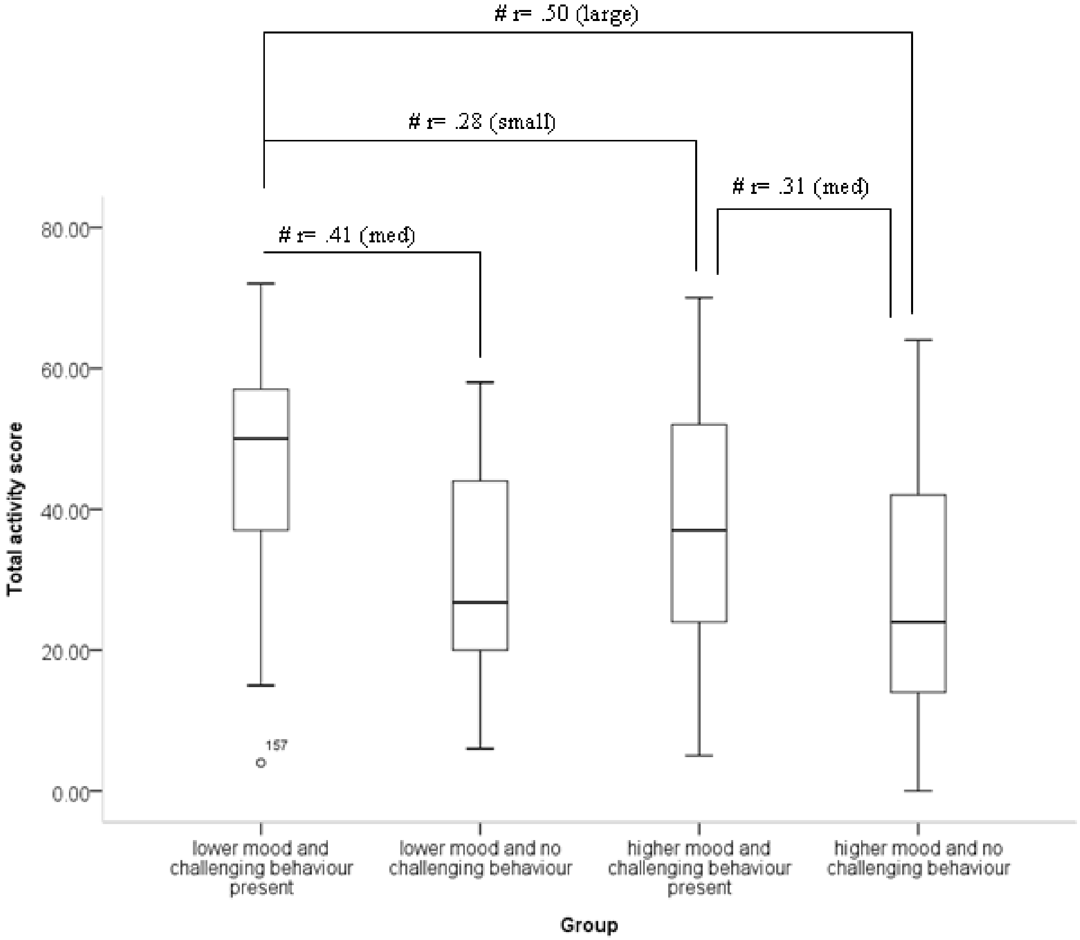
The results from the partial correlation analysis demonstrated that number of behavioural indicators of GORD were significantly associated with self injury severity, level of activity and impulsivity and level of mood and interest and pleasure, independent of the association between level of intellectual disability and these variables. This was the case for people with ASD and people with Cri du Chat syndrome. The association between interest and pleasure and gastro-oesophageal status in the ASD group, but not the Cri du Chat syndrome group was significant once level of intellectual disability was partialled out.

3.4.6 Is challenging behaviour a symptom of depression or does pain account for the association between challenging behaviour and low mood?

The final analysis tested the hypothesis that individuals with low mood and challenging behaviour would also have high activity levels. This prediction is based on the assumption that challenging behaviour is associated with low mood because both of these factors are caused by underlying pain and discomfort. Therefore, it was hypothesised that individuals engaging in challenging behaviour, who have low mood, would also have other behavioural indicators of pain, in this case, high activity. An alternative theory used to explain the association between low mood and challenging behaviour is that challenging behaviour is an atypical symptom of depression. If this theory is accurate, it would not be expected for individuals with challenging behaviour and low mood to also engage in other behavioural indicators of pain, such as high activity levels.

In order to test whether individuals with low mood and challenging behaviour are also more likely to engage in high levels of activity, the following analysis was completed. Participants were split into groups depending on their total mood score on the Mood Interest and Pleasure Questionnaire and whether or not they had shown aggressive or self injurious behaviour in the last month. Lack of power in the Cri du Chat syndrome group, evidenced by the small sample size, meant that data could only be analysed for the ASD group. Individuals who scored above the median total mood score were categorised as having 'higher mood' and individuals who scored below the median mood score were categorised as having 'lower mood'. Individuals who scored at median were allocated randomly to either the higher or lower mood group. A Kruskal- Wallis test and post-hoc Mann-Whitney U tests used to compare the activity levels between the following groups; lower mood and challenging behaviour present, lower mood and no challenging behaviour, higher mood and challenging behaviour present, higher mood and no challenging behaviour. The graph below (figure 3.1) shows the distribution of activity scores, as measured on The Activity Questionnaire, across the groups.

Fig. 3.1. Distribution of total activity scores across the following groups; lower mood and challenging behaviour present, lower mood and no challenging behaviour, higher mood and challenging behaviour present, higher mood and no challenging behaviour. Total activity scores are shown for the ASD group only. Post hoc Mann Whitney U tests used to show significant differences between the groups. Effect size (r) is stated.



Mann Whitney U test =< .001. Significant after bonferonni corrections

Results show a significant difference in activity scores between the different mood and challenging behaviour groups (*H*(3)= 48.12, p<.001). Post-hoc Mann-Whitney U tests, with bonferonni corrections, showed that the lower mood and challenging behaviour group had significantly higher levels of activity than all of the other groups. The higher mood and challenging behaviour group also had significantly higher levels of activity compared to the higher mood but no challenging behaviour group. This suggests that individuals who engage in challenging behaviour and have low mood have significantly higher levels of activity. All three variables; high activity, low mood and challenging behaviour, have been shown to be associated with pain. Therefore, the results from this analysis suggest that the association between low mood and challenging behaviour is potentially explained by the confounding impact of pain. These findings support the hypothesis that low mood and challenging behaviour, in combination, are associated with high activity levels, and thus, associated with pain.

3.5 Discussion

3.5.1 Summary of research findings

This study used an informant based questionnaire; the Gastro-oesophageal Distress Questionnaire (GDQ), to identify individuals with intellectual disability who were likely to be experiencing gastro-oesophageal distress. The participants had a confirmed diagnosis of either ASD (and selected by SCQ cut-off scores) or Cri du Chat syndrome. A range of questionnaire measures were used to assess the association between behavioural indicators of GORD, identified with use of the GDQ, and challenging behaviour, activity levels and mood. Analyses were also conducted to explore whether individuals with low mood and challenging behaviour also had high activity levels; a behavioural indicator of pain. This result would indicate that pain, as opposed to depression, can better explain the co-occurrence of low mood and challenging behaviour.

Based on previous literature, individuals who scored above the assigned cut off point on five or more items on the GDQ were identified as more likely to suffer with GORD (Richards &

Oliver, in preparation) and classified as GD+. Individuals with fewer than five clinically significant behavioural indicators of GORD were classified as GD-. This is the first time the GDQ had been used with a population group other than individuals with Cornelia de Lange syndrome, for whom the questionnaire was designed. In this sample, 41.30% of individuals with Cri du Chat syndrome and 40.41% of individuals with ASD were identified as having five or more behavioural indicators of GORD (GD+). This prevalence rate of GORD for ASD falls below the 70% identified by Horvath, et al. (1999) but above the 15.5% identified by Horvath and Perman (2002). A prevalence of 37.4% identifies ASD as a group at high risk of gastro-oesophageal distress compared to the general population where prevalence falls between 10 and 20% (Bohmer et al., 1999). These findings also confirm that Cri du Chat syndrome is strongly associated with gastro-oesophageal distress (Wilkins et al., 1983).

It was hypothesised that those individuals in the GD+ group would show high levels of behaviours previously shown to correlate with GORD which had been identified using invasive procedures (e.g.endoscopies), such as challenging behaviour, high activity levels and low mood (Luzzani et al., 2003). With regard to challenging behaviour, the results showed that levels of aggression, property destruction, stereotyped behaviour and self injurious behaviour were higher in individuals classified as GD+ compared to individuals classified as GD- in people with ASD. This supports previous findings in the literature that poor health is associated with challenging behaviour (Carr & Owen-DeSchryver, 2007; Carr et al., 2003; Kennedy & O'Reilly, 2006). In the Cri du Chat syndrome group, a significant association was evident between self injurious behaviour and behavioural indicators of GORD. Trends in the Cri du Chat syndrome data suggested that individuals in the GD+ group did engage in higher levels of aggression, property destruction and stereotyped behaviour although these associations did not reach statistical significance. Investigation into the effect size of the associations suggests that the lack of significance in the Cri du Chat group could be accounted for by the small sample size. Given that gastro-oesophageal problems are associated with high levels of pain, the findings from this study reflect those found in previous research which show pain to be associated with aggression, property destruction and stereotyped behaviour in individuals with intellectual disability and specifically those with genetic syndromes or ASD (Carr et al., 2003; Carr & Owen-DeSchryver, 2007; Oberlander & Symons, 2006). In addition to the association between the presence of challenging behaviour and likely GORD, when attributing a severity score of zero to individuals who did not engage in self injury, the severity of self injury was significantly higher in individuals in the GD+ group compared to individuals in the GD- group in individuals with ASD and Cri du Chat syndrome. When only including individuals who engaged in self injury, self injury severity was significantly higher in individuals in the GD+ group compared to the GD- in people with Cri du Chat syndrome but not people with ASD. This suggests that the severity of self injury in people with intellectual disability may be exacerbated when pain is present.

The second hypothesis was that activity levels would be higher in individuals in the GD+ group compared to the GD- group. This was the case in both the ASD and Cri du Chat syndrome groups. These results support previous research findings that show high activity levels are associated with the presence of GORD (Luzzani et al., 2003). As GORD is known to be a painful health condition, this finding may have implications for identifying pain in individuals with intellectual disability. Hyperactivity might alert carers to the possibility of pain and discomfort. This has already been implemented in several reliable and validated pain measurement tools such as the FLACC (Merkel et al., 1997) and NCCPC (Breau et al., 2004), which use 'agitated and restless activity' as a behavioural indicator for the presence of pain.

It was also hypothesised that individuals with five or more behavioural indicators of GORD would have lower mood and interest and pleasure (two main constructs of depression) compared to individuals with fewer indicators of GORD. This hypothesis was upheld. Participants with ASD in the GD+ group obtained significantly lower scores than participants in the GD- group on both the mood and the interest and pleasure subscales of the Mood Interest and Pleasure Questionnaire. Participants with Cri du Chat syndrome in the GD+ group scored significantly lower on the mood subscale compared to people in the GD- group. In both the ASD and Cri Du Chat syndrome group, the association between low mood and behavioural indicators of GORD was stronger than the association between low interest and pleasure and behavioural indicators of GORD. Low mood, like challenging behaviour and high activity levels, has been shown to be associated with health related pain (Berg et al., 2007). Therefore scores indicative of low mood in the MIPQ might indicate the need to investigate the possibility of pain and discomfort. Consequently, low mood may be a more valid indicator of pain linked to GORD than low interest and pleasure.

As noted in the literature and in the results of this study, there is a strong association between low mood and challenging behaviour (Ross & Oliver, 2002b). It is possible that the association between challenging behaviour and low mood could be accounted for by pain as a common causal factor (Ross & Oliver 2003). However, other researchers interpret the association between low mood and challenging behaviour differently, suggesting that self injurious behaviour may be an atypical symptom of depression (Marston et al., 1997; Meins, 1995). The final hypothesis was that pain accounts for the association between low mood and challenging behaviour. Results from this study demonstrated that individuals with both low mood and challenging behaviour also displayed another behavioural indicator of pain, in this case, high activity levels (Breau et al., 2004; Luzzani et al. 2003; Merkel et al., 1997). This result would not be expected if low mood was an atypical symptom of depression in all people but would be expected if both low mood and challenging behaviour were influenced by pain. Therefore, the findings of this study suggest that low mood is not necessarily an atypical symptom of depression but instead, the association between low mood and challenging behaviour can potentially be explained by the influence of a confounding variable, pain. This finding also highlights an important and exciting possibility. As yet, there is no consensus on how to distinguish between challenging behaviour that is associated with pain and challenging behaviour that is associated with depression. This finding suggests that perhaps activity level could be used to indicate which interpretation is more likely to be accurate for each case of challenging behaviour. This finding suggests that perhaps, when low mood and challenging behaviour are both present, *high* activity levels may indicate that pain is more likely whereas *low* activity levels may suggest that depression is more likely, which is in line with the presentation of depression in the general population (DSM-IV). Future research should further investigate the accuracy of using activity level to distinguish causes of challenging behaviour in this way.

In summary, individuals in the GD+ group had a higher frequency of different forms of challenging behaviour, more severe self injury, higher levels of activity and lower mood, interest and pleasure compared to individuals in the GD- group. These findings are consistent with hypotheses based on previous research, which used invasive methods to identify GORD (Luzzani et al., 2003). A high score on the GDQ could be used to alert health care professionals to likely cases of GORD which warrant further investigation. This could lead to the identification of previously unrecognised cases of GORD and mean that the human cost of using invasive methodology to confirm a diagnosis of GORD is more justifiable. The use of these behavioural indicators of likely GORD could be used more broadly in more generic pain assessment tools and these tools could be made available to all people with intellectual disability, again alerting health care professionals and carers to the possibility of pain and discomfort.

The results from this study support the notion that pain and discomfort may account for the association between low mood/depression and challenging behaviour. Therefore, the role of pain or discomfort needs to be considered in future research investigating the association between low mood and challenging behaviour. This again could lead to the identification of previously unrecognised or unresolved health conditions related to pain and discomfort.

3.5.2 Limitations

A limitation of this study was that there were relatively few participants in the Cri du Chat syndrome group (N= 46). The forced entry binary logistic regression failed to find an association between level of intellectual disability and health problems and/or self injurious behaviour in the Cri du Chat syndrome group and level of intellectual disability and aggression, destruction of property and stereotyped behaviour in people with ASD and Cri du Chat syndrome. These are all robust associations reliably found throughout the research literature (Emerson et al., 2001b; Jansen et al., 2004) so failure to replicate these findings suggest that there may have been insufficient power for some analyses. This may be the reason why a significant result was not found relating to challenging behaviour and low mood, which was expected in the Cri du Chat group. This is also supported by the various effect size calculations which revealed larger effect sizes on associations where statistical tests had suggested lower levels of significance.

3.5.3 Clinical and research implications

This study has demonstrated an association between the presence of behavioural indicators of gastro-oesophageal distress, which is linked to high levels of pain and discomfort, high levels of challenging behaviour, high activity levels and low mood and interest and pleasure. There are important implications of these findings. Firstly, there is the possibility that challenging behaviour, activity levels and mood could be used as indicators for the presence of pain, providing indirect indicators that the individuals may be experiencing pain. This could lead to identification and treatment of unidentified health problems and also more accurate estimates of pain prevalence rates in people with intellectual disability. Secondly, this information can be used to target intervention strategies for challenging behaviour, which may be associated with the presence of pain. If these potential indicators for pain (hyperactivity and low mood in combination) are identified early, interventions can be implemented to reduce or prevent the development of pain related challenging behaviour. These interventions can take into consideration the impact that pain may have on the individual's behaviour. The findings from this study also highlight the importance of investigating the possibility of pain when an individual presents with challenging behaviour and low mood, and suggest that caution should be taken not to interpret these behaviours as 'symptoms of depression' without other substantial evidence for this diagnosis.

CHAPTER 4

SELF INJURY, AGGRESSION AND DESTRUCTION OF PROPERTY IN TUBEROUS SCLEROSIS COMPLEX

4.1 Preface

There are inherent difficulties with assessing pain in people with intellectual disabilities, which make investigating the association between pain and challenging behaviour problematic. In chapter 3, the difference in relevant behavioural and health characteristics between individuals with and without challenging behaviour was established. In this chapter, a different approach for investigating the association between pain and challenging behaviour will be employed. Challenging behaviour will be assessed in Tuberous Sclerosis Complex (TSC), a genetic syndrome characterised by painful heath conditions. Rates of challenging behaviour in TSC will be compared to rates of challenging behaviour in Down syndrome, a low-risk group for pain. The differences in person characteristics between individuals with and without challenging behaviour in TSC will also be examined to further support the findings reported in chapter 3.

4.2 Introduction

Tuberous Sclerosis Complex (TSC) is a genetic disorder resulting from a mutation of either the *TSC1* gene on chromosome 9q34 (van Slegtenhorst, Hoogt, Hermans, Nellist, Janssen, Verhoef, et al., 1997) or the *TSC2* gene on chromosome 16p13 (Nellist, Janessen, Brook-Carter, Hesseling-Janssen, Maheshwar, Verhoef et al., 1993). Estimated prevalence rates range from 1/6800 to 1/11400 (O'Callaghan, Shiell, Osborne & Martyn, 1998; O'Callaghan 1999; Ahlsen, Gillberg, Lindblom & Gillberg 1994) and birth incidence estimates range from 1/6,000 to 1/15,000 (Webb & Osbourne, 1992). TSC is characterised by abnormal growths in multiple organs including the central nervous system, skin, kidneys and heart (Crino et al. 2006). Intellectual ability shows a bimodal distribution, with ~30% of individuals showing profound intellectual disability while the remaining 70% fall on a normal distribution of ability (de Vries & Prather, 2007). Best estimates from epidemiological populations suggest that about 45% of individuals with TSC have intellectual disability (Joinson, O'Callaghan, Osborne, Martyn, Harris & Bolton, 2003). In addition to the well documented physical phenotype associated with TSC, there is an associated behavioural phenotype that includes high rates of aggression (Kopp et al., 2008), self injury (Staley, Montenegro, Major, Muzykewicz, Halpern, Kopp et al., 2008) and autistic-like behaviours (Baker, Piven, & Sato, 1998; Critchley & Earl, 1932; Gutierrez, Smalley &Tanguay, 1998; Smalley, 1998).

The behavioural problems associated with TSC can be of major concern for parents and carers (Hunt, 1983). This study focuses on three types of challenging behaviours; self injury, aggression and destruction of property. Hunt (1983) reported that 16% of individuals with TSC engaged in 'destructive behaviours'. However, there have been no further descriptions of destructive behaviours since Hunt's original report. Currently, there is a lack of consensus regarding rates of self injury and aggression. Relatively low rates of self injury between 10%

and 13% have been reported in some studies (Hunt, 1983; Hunt, 1997; Staley et al., 2008). These rates are similar to those reported in the total population of people with intellectual disability (Collacott, Cooper, Branford & McGrother, 1998; Crocker, Mercier, Lachapelle, Brunet, Morin & Roy, 2006). In contrast, de Vries, Hunt and Bolton (2007) reported higher rates of self injury (41%) in TSC. These discrepancies are also evident in the rates of aggressive behaviour with prevalence estimates ranging from 13.3% to 58% (Hunt, 1983; Hunt, 1997; Kopp, Muzykewicz, Staley, Thiele & Pulsifer, 2008; de Vries et al., 2007).

Such discrepancies in prevalence may be attributed to variations in the way in which self injury and aggression are defined across different studies or differences in the sampling methodology. For example, Staley et al. (2008) reviewed behavioural charts for records of self injury. It may be the case that self injury was only documented in these medical records/clinical records when an episode of self injury was unusual for the individual or particularly severe. Hunt (1997) used semi-structured interviews with parents of people with TSC while de Vries et al. (2007) used informant postal questionnaire surveys. The conceptualisation and definition of self injury used by researchers during an interview setting might have been narrow compared to that used by parents when providing answers in the postal survey. These methodological differences were also apparent in the studies that assessed aggressive behaviour (de Vries et al., 2007, Kopp et al., 2008) and may yield under- or overestimates of challenging behaviour. In order to compare rates of challenging behaviour between different syndrome groups, it is important that comparable methodological approaches are employed so that the risk of challenging behaviour in each syndrome group can be appraised accurately.

It is clear from the above review of the literature available on challenging behaviour in TSC that further research into the prevalence of self injury, aggression and destruction of property in children and adults with TSC is needed. Understanding how the rates of challenging behaviour in TSC compares to other syndrome groups in which the prevalence of challenging behaviour is well understood is important and would allow the risk of challenging behaviour in TSC to be appraised relative to other genetic syndromes.

The relationship between self injury and aggression in genetic disorders associated with intellectual disability is attracting increasing research interest. Arron et al., (2011) examined the rates and phenomenology of self injury and aggressive behaviours across seven different syndrome groups (Angelman, Cornelia de Lange, Cri du Chat, Fragile x, Lowe, Prader-WIlli and Smith-Magenis syndromes) compared to individuals with intellectual disability of heterogeneous cause and found that challenging behaviour was significantly more prevalent in particular syndrome groups including: Cri du Chat, Cornelia de Lange, Fragile X, Prader-Willi, Lowe and Smith-Magenis syndromes. Aggression was significantly higher in people with Angelman and Smith-Magenis syndromes. Arron et al. (2011) also showed that some topographies of self injury were more prevalent in people with certain syndromes. For example, 'hitting self with body' and 'pulling self' were significantly more likely to occur in Cornelia de Lange syndrome while 'biting self' was more likely in Fragile X syndrome. There are no studies of TSC that describe the rates of these manifestations in comparison to other syndromes.

In this study, the rates and topographies of self injury, aggression and destruction of property is compared primarily between individuals with TSC and Down syndrome. The rates of challenging behaviour will also be reported for individuals with ASD, Fragile X, Cornelia de

Lange and Angelman syndromes. The justification for including each of these groups is discussed below.

Down syndrome has been selected as the main comparison group because it has a well documented behavioural phenotype (Chapman & Hesketh, 2000; Collacott et al., 1998; Dykens, Shah, Sagun, Beck & King, 2002; Fidler, 2005; Gath & Gumley, 1986). Prevalence rates of self injury, aggression and destruction of property have been shown to be similar in Down syndrome compared to individuals with intellectual disability of heterogeneous aetiology, so Down syndrome will act as a homogenous, well documented challenging behaviour comparison group (Crocker et al., 2006, Collacott et al., 1998; Cooper, Smiley, Allan, Jackson, Finlayson, Mantry et al., 2009; Dykens et al., 2002, Gath & Gumley, 1986; Matson, Minshawi, Gonzalez & Mayville, 2006).

The rates of self injury, aggression and destruction of property will also be reported for individuals with ASD because TSC is associated with ASD (Baker et al., 1998; Smalley, 1998). In the most comprehensive study of ASD in TSC, Joinson et al (2003) showed that approximately 25% of individuals with TSC meet criteria for autism, and a further 25% for ASD. ASD has been identified as a risk marker for self injury (Emerson et al., 2001b; Baghdadli et al., 2003; McClintock et al., 2003) (see section 1.2.1). Reporting the rates of challenging behaviour in ASD will allow the risk of challenging behaviour associated with a diagnosis of TSC, above and beyond the association with ASD, to be evaluated.

The rates of self injury, aggression and destruction of property will also be reported for Fragile X, Cornelia de Lange and Angelman syndromes, using the same measures and methodology in the TSC, Down syndrome and ASD groups. This will enable meaningful comparisons to be made across the groups. Rates of challenging behaviour have been documented previously for these groups. Prevalence rates of self injury in Cornelia de Lange are reported to range from 16% to 64% and aggression is reported to range from 10% to 43% (Oliver, Arron, Powis & Tunnicliffe, 2011a). Destruction of property in Cornelia de Lange is reported to range from 10% to 53% (Oliver et al., 2011a). Symons, Clark, Hatton, Skinner and Bailey (2003) found that 58% of boys with Fragile X syndrome engaged in self injurious behaviour. Also, aggressive behaviour has been reported in 83% of children with Angelman syndrome (Strachan, Shaw, Burrow, Horsler, Allen & Oliver, 2009). If the prevalence rates of challenging behaviour in Cornelia de Lange, Fragile X and Angelman syndromes reported in this study reflect previous findings, there can be a greater degree of confidence in the validity of the results reported for the TSC group.

Research into the characteristics of individuals who show self injury and aggression within syndrome groups has also proven informative. Arron et al. (2011) assessed differences in person characteristics between individuals with and without challenging behaviour in different syndrome groups. Factors such as negative affect, high levels of repetitive and compulsive behaviours and increased levels of activity were shown to be predictive of challenging behaviour in a range of syndrome groups. For example, levels of activity and impulsivity were significantly higher in individuals with Cornelia de Lange syndrome who engaged in self injury and aggression compared to those who did not. Similar findings were identified in Fragile X and Prader-Willi syndromes while individuals with Angelman syndrome who engaged in self injury had significantly lower mood compared to those who did not. Also, more communication and socialisation deficits associated with ASD were demonstrated in individuals with Prader-Willi, Fragile X and Cornelia de Lange syndromes who engaged in self injury compared to individuals in those syndrome groups who did not engage in self injury. This replicates and extends previous research findings. For example, McClintock et al., (2003) conducted a meta-analysis of previous research and found that self injury, aggression and destruction of property were associated with a diagnosis of ASD. Furthermore, hyperactivity, impulsivity and repetitive behaviours have been shown to be associated with self injurious behaviour (Bodfish, Crawford, Powell, Parker, Golden & Lewis, 1995; Petty & Oliver, 2005; Cooper et al., 2009; Oliver et al., 2009) and low mood has been shown to be associated with the frequency and severity of challenging behaviour in people with severe and profound intellectual disability (Hayes et al., 2011; Ross & Oliver; 2002b). It is possible that these person characteristics, which vary across syndrome groups, may help explain differences in the prevalence of self injury and aggression between groups.

TSC is associated with a number of these characteristic risk markers for self injury and aggression. For example, hyperactivity has been found in 35% to 54% of children with TSC (de Vries et al., 2007; de Vries, Gardiner , Bolton, 2009; Hunt, 1983; Kopp et al., 2008). De Vries et al., (2007) also reported impulsivity in 52% of individuals and depressed mood in 23% of people with TSC. To date, research has yet to assess whether these characteristics, reported in a substantial proportion of people with TSC, are associated with self injury, aggression and destruction of property within the syndrome. It is predicted that these person characteristics, which have been shown to be associated with challenging behaviour in other genetic syndromes, will differ between people with and without challenging behaviour in TSC.

In addition to these risk markers, a growing literature provides evidence for a possible association between challenging behaviour and the presence of pain or distress, often related to health conditions (Carr & Owen-DeSchryver, 2007; Carr et al., 2003; Luzzani et al., 2003) (see sections 1.3.6 & 2.5.5). Given the complexity of health problems associated with TSC (for example SEGA brain tumours leading to raised intracranial pressure, headaches and photophobia (Ramesh, 2003); renal angiomyolipoma causing flank pain, bleeding, renal failure (Rakowski, Winterkorn, Steele, Halpern & Thiele, 2006); cardiac rhabdomyoma leading to chest discomfort (Choi, Bae, Noh, Yoon & Hwang, 2005)), it is likely that individuals with this syndrome may experience pain and discomfort at some point (Clarke, Hancock, Kingswood & Osborne, 1994; Leung & Robson, 2007; Curatolo, Bombardieri & Jozwiak, 2008). To date, there is no research exploring whether pain and discomfort play a role in the occurrence of challenging behaviour in people with TSC. This is of particular importance given that a significant proportion of individuals with TSC are likely to be non-verbal and have limited functional communication. Challenging behaviour may therefore be an indication of pain/discomfort.

Identification of pain and/or health problems in people with intellectual disability can be problematic as the individual is often unable to communicate the discomfort that they may be feeling (Malviya et al., 2005). Several observational measures have been used to overcome this problem (Malviya et al., 2006; Brea et al., 2000). In this study, two informant measures; the NCCPC-R, designed to assess general pain and the GDQ, designed to assess discomfort linked to gastro-oesophageal reflux (see section 3.3.3.1) are used. Although there are no reports of TSC being specifically associated with gastro-oesophageal reflux, the GDQ will be used as a secondary means for identifying individuals with TSC who may be likely to be in discomfort. It is predicted that pain and discomfort will be associated with self injury and aggression in TSC, as has been demonstrated previously with individuals with intellectual disability (Carr & Owen-DeSchryver, 2007; Carr, et al., 2003).

In summary, previous research has shown that it is revealing to compare between syndrome groups to determine which groups are at the highest risk of challenging behaviour. There is a paucity of research documenting the rates of self injury, aggression and destruction of property and topographies of self injury in TSC and as yet, there has been no previous research that compares TSC with other genetic groups in which the behavioural phenotype is well described. Similarly, there is no research documenting how the severity of self injury in TSC compares to other genetic syndromes. These questions will be addressed in this study.

It is also important to look within groups to examine person characteristics that differ between individuals with and without self injury and aggression; such as affect, activity levels, autistic-like behaviours and pain. It is predicted that characteristics which differ between people with and without challenging behaviour in other syndrome groups such as Cornelia de Lange, Fragile X and Angelman syndromes, will also differ between individuals with and without self injury and aggression in the TSC group.

4.2.1 Aims and hypotheses

The aims of this study are to;

1. Document the odds ratios of self injury, aggression and destruction of property in people with TSC compared to Down syndrome. The odds ratio between these behaviours occurring in Angelman syndrome, Fragile X syndrome and Cornelia de Lange syndrome compared to Down syndrome will also be reported to provide points of comparison.
2. Document the odds ratios of different topographies of self injury in TSC compared to Down syndrome. The odds ratios of topographies of self injury will also be reported for Angelman syndrome, Fragile X syndrome and Cornelia de Lange syndrome compared to Down syndrome to provide further points of comparison.

In this study it is hypothesised that:

1. Individuals with TSC with self injury and aggression will have higher levels of negative affect, autism spectrum behaviours, hyperactivity and repetitive and impulsive behaviours compared to individuals with TSC without these behaviours.

2. Individuals with self injury, aggression and destruction of property in TSC will have higher levels of behavioural indicators of pain and discomfort compared to individuals with TSC without self injury, aggression and destruction of property.

4.3 Method

4.3.1 Recruitment and participants:

Data regarding participants with TSC were collected as part of an ongoing survey at the Cerebra Centre for Neurodevelopmental Disorders (Arron et al., 2011; Moss et al., 2009; Oliver et al., 2011b). Carers were invited to complete the questionnaires on behalf of their child or person they cared for with TSC. All children under the age of 16 with TSC and adults with TSC who also had an intellectual disability were invited to participate in the study. Adults without an intellectual disability were not invited to participate because the questionnaires used in this study were all informant based and were not appropriate for self-report. Due to the difference in inclusion criteria between individuals under the age of 16 compared to those aged 16 or older, participants will be split into two groups for the purpose of analysis. Therefore, participants will be divided into ‘under 16’ and ’16 and over’ groups. Parents/carers were asked about various aspects of their child’s behaviour and also about the impact of caring for their child. These questionnaires were inappropriate for parents of adults with TSC who did not have an intellectual disability as these individuals would be able to live independent lives.

Data relating to individuals with other genetic syndromes were accessed from a database at the Cerebra Centre for Neurodevelopmental Disorders, University of Birmingham. The other syndrome groups included in this study were selected because they were broadly similar to the TSC group in terms of ability level (measured by the Wessex; Kushlick et al., 1973; see method section below). Analysis was conducted according to age groups. Cornelia de Lange syndrome, Fragile X syndrome, Down syndrome and ASD were the groups included in the under 16 category and Angelman syndrome, Cornelia de Lange syndrome, Down syndrome and ASD were the groups included in the '16 and over' category.

Children younger than four years were excluded as one of the measures required informants to rate behaviour when the child was four to five years old. Therefore, in the under 16 group, participants were aged between 4 and 15 years 11 months and participants in the adult group were aged between 16 and 62 years (inclusive). Any individuals with a maximum score on the 'self help' subscale of the Wessex Questionnaire were excluded from the adult group to guarantee that all participants in the adult group had an intellectual disability. This was the case with adults in all genetic syndrome groups.

Individuals within the ASD group were screened to ensure that they scored above the cut-off for ASD on the Social Communication Questionnaire. Individuals with TSC, Cornelia de Lange, Fragile X, Down and Angelman syndromes were included if their diagnosis had been confirmed by a paediatrician or clinical geneticist. All questionnaires in this study were completed by parents and carers on behalf of the participants. Table 4.1 provides a description of the participants.

Table 4.1. Demographic characteristics of the participants. Mean age (standard deviation), percentage of participants who were males, fully mobile had normal vision and hearing, were partly verbal/verbal and partly able/able in each syndrome group.

	N	Mean age^{B} (SD)	% male (N)	% verbal/ partly verbal (N)	%mobile (N)	%able/partly able (N)	% normal vision (N)	% normal hearing (N)
Under 16s								
TSC	37	10.08	51.4	89.2	81.1	78.4	**89.2**	**97.2**^{A}
		(3.09)	(19)	(33)	(30)	(29)	(33)	(35)
Down syndrome (comparison group)	43	9.00	41.9	95.2^{A}	83.7	90.7	55.8	53.5
		(3.31)	(18)	(40)	(36)	(39)	(24)	(23)
Cornelia de Lange syndrome	61	10.10	44.3	**60.7**^{A}	**49.2**^{A}	**44.3**	65.6^{A}	57.4
		(3.25)	(27)	(37)	(30)	(27)	(40)	(35)
Fragile X syndrome	112	**10.88**	**100**	95.5	68.8^{A}	89.3	**87.5**	**97.3**
		(2.58)	(112)	(107)	(77)	(100)	(98)	(108)
Autism Spectrum Disorder	188	9.37	**85.6**	93.0^{A}	**94.1**^{A}	87.2	**96.8**	**97.3**
		(3.14)	(161)	(174)	(177)	(164)	(182)	(183)
16 and over								
TSC	30	27.63	60.0	**44.8**^{A}	63.3	**36.7**	**83.3**	**100**^{A}
		(8.74)	(18)	(13)	(19)	(11)	(25)	(29)
Down syndrome (comparison group)	21	30.48	38.1	85.7	85.7	76.2	47.6	57.1
		(12.08)	(8)	(18)	(18)	(16)	(10)	(12)
Angelman syndrome	31	**21.81**	54.8	**32.3**	**45.2**	**32.3**	**93.5**	**100**
		(6.20)	(17)	(10)	(14)	(10)	(29)	(31)
Cornelia de Lange syndrome	49	**23.92**	38.8	**46.9**	62.5^{A}	51.0	63.3	64.6^{A}
		(6.72)	(19)	(23)	(30)	(25)	(31)	(31)
Autism Spectrum Disorder	18	**19.33**	**88.9**	66.7	83.3	94.4	**94.4**	**94.4**
		(5.43)	(16)	(12)	(15)	(17)	(17)	(17)

^{A}Data missing from one participant

^{B}In years

Bold indicates if the value is significantly different compared to the Down syndrome group (p<.05)

4.3.2 Procedure

Covering letters, information sheets, consent forms and questionnaire packs suitable for under 16 and over 16 year old participants were prepared and sent out by the Tuberous Sclerosis Association to their membership (see appendices F, G, H & I). Parents or carers of individuals with TSC were invited to take part in the study if they cared for a child under the age of 16 with TSC or an adult aged 16 or over with TSC who also had intellectual disability. Data were combined with data stored at the Cerebra Centre for Neurodevelopmental Disorders, University of Birmingham regarding children and adults with other genetic syndromes and idiopathic ASD.

4.3.3 Measures:

All participants completed the same questionnaire measures, with the exception of the NCCPC-R, which was only completed by parents and carers of individuals with TSC. The measures evaluated levels of activity and impulsivity, affect and interest and pleasure, deficits in communication and socialisation, levels of repetitive language, insistence on sameness, restricted preferences, and compulsive and stereotyped behaviours. The following measures are all informant based questionnaires and were completed by all participants.

4.3.3.1 Mood, Interest and Pleasure Questionnaire- Short version (Ross et al., 2008; appendix D5)

The Mood, Interest and Pleasure Questionnaire- Short version (MIPQ) assesses two key constructs of depression in people with intellectual disabilities (see section 3.3.3.5)

4.3.3.2 Challenging Behaviour Questionnaire (Hyman, Oliver & Hall, 2002; appendix D2)

The Challenging Behaviour Questionnaire (CBQ) rates the presence of physical aggression, destruction of property and specific topographies of self injury over the last month (see section 3.3.3.2)

4.3.3.3 Wessex Questionnaire (Kushlick, Blunden & Cox, 1973; appendix D3)

The Wessex questionnaire provides a proxy measure of degree of intellectual disability (see section 3.3.3.3).

4.3.3.4 The Activity Questionnaire (Burbidge & Oliver 2008; appendix D4)

The Activity Questionnaire (TAQ) provides a measure of hyperactivity, impulsivity and impulsive speech for children and adults with intellectual disability (see section 3.3.3.4).

4.3.3.5 Gastro-oesophageal Distress Questionnaire (Oliver & Wilkie, 2005; appendix D1)

The Gastro-oesophageal Distress Questionnaire (GDQ) rates the frequency of behaviours indicative of pain in the oesophagus and stomach in people with intellectual disability (see section 3.3.3.1).

4. 3.3.6 Social Communication Questionnaire (Berument et al., 1999; Rutter et al., 2003; appendix D7)

When using the Social Communication Questionnaire (SCQ), informants indicate the presence of behaviours associated with ASD. Nineteen out of 40 items rate the child's current behaviour and the rest ask questions relating to behaviour when aged four to five years. A clinical cut off of 15 or more on the SCQ is suggestive of ASD and a cut off of 22 is suggestive of Autism (Berument et al., 1999; Eaves, Wingert, Ho & Mickelson, 2006).

4.3.3.7 The Non-Communicating Children's Pain Checklist-Revised (NCCPC-R) (Breau, et al., 2004; appendix D6)

The NCCPC-R asks informants to rate the frequency of behaviours related to pain in children with intellectual disabilities. The NCCPC-R has been shown to have good internal validity when used retrospectively (Breau et al., 2000). The checklist has been shown to have high inter-episode consistency between two separate episodes of pain and consistent behaviour ratings when no pain is present (Breau et al., 2002). For the purpose of this study the administration of the NCCPC-R was amended. Respondents were asked to rate the frequency of behaviour over a two week time period rather than over two hours. This modification was made in order to identify individuals likely to be experiencing chronic health conditions and pain as opposed to episodes of acute pain. This modification has been used previously in order to measure 'typical' pain behaviour (Breau et al., 2003; Symons et al., 2009a). The NCCPC-R had not been included in the data collection conducted with the other genetic syndrome groups included in this study and was therefore only completed by parents and carers of individuals with TSC.

4.3.4 Data Analysis

Data were analysed using SPSS 18.0 software. All data were tested for normality using Shapiro-Wilk test and non-parametric tests were used where necessary. Analysis was conducted according to age groups meaning participants were divided into 'under 16' and '16 and over' groups. Comparison of rates of challenging behaviour and topographies of self injury was conducted using odds ratios, which can be used to indicate if the odds of challenging behaviour occurring in one group are significantly different compared to another group. An odds ratio is deemed significant if the confidence intervals do not cross one. When using 95% or 99% confidence intervals, confidence intervals that do not exceed one would represent a difference in odds at a significance level of *p*<.05 and *p*<.01 respectively. Kruskal-Wallis analysis was used to compare the severity of self injurious behaviour between TSC, Down, Angelman, Fragile X, and Cornelia de Lange syndromes.

4.4 Results

4.4.1 Prevalence of self injury, aggression, destruction of property and topographies of self injury in TSC

The primary aim of this study was to identify the rates of self injury, aggression and destruction of property in individuals with TSC and to determine whether the likelihood of these behaviours occurring in people with TSC was significantly different compared to people with Down syndrome. The odds of self injury, aggression and destruction of property was also compared in individuals with Fragile X, Angelman and Cornelia de Lange syndromes to people with Down syndrome in order to rank the risk of challenging behaviour across syndrome groups. Table 4.2 shows the percentage of individuals showing self injury, aggression and property destruction in each syndrome group. The results from the odds ratios analysis are also described which show the likelihood of these challenging behaviours occurring in each syndrome group compared to people with Down syndrome.

Table 4.2. Percentage of individuals engaging in self injury, aggression and property destruction in each group. Odds ratios and 99% confidence intervals show the likelihood of individuals in each syndrome group showing self injurious behaviour, aggression and destruction of property compared to individuals with Down syndrome.

	Self injurious behaviour		**Aggression**		**Destruction of property**	
	%	**Odds ratio (99% CIs)**	**%**	**Odds ratio (99% CIs)**	**%**	**Odds ratio (99% CIs)**
Syndrome Group		**Between syndrome groups**		**Between syndrome groups**		**Between syndrome groups**
Under 16s						
TSC	27.00	2.74 (.58-13.00)	50.00 ^{a}	1.47 (.45-4.80)	33.30 ^{a}	2.43 (.59-9.91)
Down syndrome (comparison group)	11.90^{a}	-	40.50 ^{a}	-	17.10 ^{b}	-
Cornelia de Lange syndrome	63.90	**13.12** **(3.21-53.66)**	44.10^{b}	1.16 (.40-3.33)	55.90 ^{b}	**6.16** **(1.74-21.88)**
Fragile X syndrome	54.50	**8.85** **(2.36-33.24)**	60.90 ^{b}	2.29 (.88-5.95)	54.50 ^{b}	**5.83** **(1.79-18.96)**
Autism Spectrum Disorder	41.70^{a}	**5.30** **(1.46-19.19)**	66.70^{c}	**2.94** **(1.19-7.29)**	56.90^{d}	**6.41** **(2.05-20.03)**
16 and over						
TSC	31.00 ^{a}	1.91 (.33-11.21)	37.90 ^{a}	2.60 (.46-14.81)	17.20 ^{a}	1.25 (.16-9.70)
Down syndrome (comparison group)	19.00	-	19.00	-	14.30	-
Angelman syndrome	44.80 ^{b}	3.45 (.61-19.43)	72.40 ^{b}	**11.16** **(1.86-66.83**)	41.40 ^{b}	4.24 (.65-27.76)
Cornelia de Lange syndrome	83.30 ^{a}	**21.25** **(3.70-121.99)**	45.80 ^{a}	3.60 (.71-18.11)	55.30 ^{b}	**7.43** **(1.26-43.97)**
Autism Spectrum Disorder	66.70	**8.50** **(1.24-58.48)**	38.90	2.70 (.40-18.09)	27.80	2.31 (.28-18.94)

^{a} data missing for one participant, ^{b} data missing for two participants, ^{c} data missing for 8 participants, ^{d} data missing for 7 participants

Bold text indicates significantly greater risk of challenging behaviour in test group compared to Down syndrome comparison group ($p<.01$)

The results show that although the rates of self injury, aggression and destruction of property in TSC were high in the under 16s group and the 16 and over group (self injury; 27.00% and 31.00%, aggression; 50.00% and 37.90% and destruction of property; 33.30% and 17.20% respectively), the risk of these behaviours occurring in people with TSC was not significantly different to the risk of the behaviours occurring in people with Down syndrome. Results from other genetic syndromes are reported to provide further comparisons. In the under 16s group, the risk of self injury and destruction of property was significantly greater in people with Cornelia de Lange syndrome and Fragile X syndrome compared to people with Down syndrome. The risk of self injury, aggression and destruction of property was significantly greater in people with ASD compared to people with Down syndrome. In the over 16 group, when compared to people with Down syndrome, the risk of self injury was higher in people with Cornelia de Lange syndrome and ASD, the risk of aggression was higher in people with Angelman syndrome and the risk of destruction of property was higher in people with Cornelia de Lange syndrome.

A further aim of the study was to identify whether there was a greater risk of different topographies of self injury in individuals with TSC compared to individuals with Down syndrome. Comparisons between ASD, Fragile X, Angelman, Cornelia de Lange syndromes and Down syndrome were also evaluated to demonstrate where the TSC group falls relative to these groups. Table 4.3 presents the results from the odds ratio analysis, which was used to compare the risk of topographies of self injury in TSC, ASD, Fragile X, Angelman and Cornelia de Lange syndromes to the risk of these behaviours in people with Down syndrome.

Table 4.3. Odds ratios and 99% confidence intervals demonstrating the likelihood of individuals in each syndrome group showing topographies of self injury compared to Down syndrome, which acts as the comparison group.

Syndrome Group	Hits self with body	Hits self against object	Hits self with object	Bites self	Pulls self	Rub/ scratch self	Inserts
Under 16s							
TSC	1.56 (.08-29.79)	1.00 (.06-17.90)	A	1.50 (.09-26.33)	.64 (.03-12.31)	1.00 (.06-17.90)	.29 (.01-5.47)
Cornelia de Lange syndrome	.96 (.08-11.68)	1.29 (.11-15.61)	A	1.58 (.13-19.16)	1.09 (.09-13.33)	.59 (.05-7.38)	.33 (.03-4.11)
Fragile X syndrome	.60 (.05-6.98)	.53 (.05-6.32)	A	5.54 (.46-66.77)	.45 (.04-5.35)	.37 (.03-4.46)	.10 (.01-1.29)
Autism Spectrum Disorder	.89 (.08-10.09)	.68 (.06-7.79)	A	1.01 (.09-11.49)	.60 (.05-6.91)	.91 (.08-10.32)	.10 (.01-1.23)
16 and over							
TSC	1.91 (.33-11.21)	.50 (.02-11.78)	A	6.00 (.18-197.37)	A	.33 (.02-6.56)	B
Angelman syndrome	3.45 (.61-19.43)	.018 (.01-4.71)	A	2.57 (.09-70.20)	A	.12 (.01-2.63)	.44 (.02-9.02)
Cornelia de Lange syndrome	**21.25** **(3.70-121.99)**	.58 (.04-8.69)	A	3.15 (.14-68.97)	A	.77 (.06-9.31)	.37 (.02-5.65)
Autism Spectrum Disorder	**21.25** **(1.82-248.62)**	.02 (.01-5.23)	A	3.00 (.11-83.88)	A	.33 (.02-5.68)	B

[A] Unable to calculate because there are no individuals showing that topography of challenging behaviour in the comparison/ Down syndrome group.

[B] Unable to calculate because there are no individuals showing that topography of challenging behaviour in the named syndrome group.

Bold text indicates significantly greater risk of topography of self injury in test group compared to Down syndrome comparison group ($p<.01$)

There was no significant difference in the risk of any topography of self injury in people with TSC compared to people with Down syndrome. A similar pattern of findings was identified across Fragile X, Angelman and Cornelia de Lange syndromes. The only exceptions were ‘hitting self with body’ in Cornelia de Lange syndrome and ASD in individuals aged 16 and over, for which there was a significantly greater risk of these behaviours occurring compared to the risk in people with Down syndrome.

4.4.2 Differences in person characteristics between people with and without challenging behaviour in TSC

It was hypothesised that factors previously shown to differ between people with and without self injury, aggression and destruction of property in individuals with other genetic syndromes (Arron et al., 2011) would also differ between individuals with and without these challenging behaviours in TSC. In order to test this hypothesis, t-tests were conducted to assess the difference in age between people with and without challenging behaviour in individuals with TSC. Chi square analysis was used to determine whether the proportion of individuals engaging in challenging behaviour was different between people with hearing and vision problems to those without, between individuals with good compared to poor mobility and self help skills and between individuals with and without the ability to speak full words. Table 4.4 shows the results from this analysis.

Table 4.4. Chi square analysis showing the association between participant demographic variables and challenging behaviour in individuals with TSC.

		Self injury				**Aggression**				**Destruction of property**			
		present	**absent**	**t/χ^2**	***p***	**present**	**absent**	**t/χ^2**	***p***	**present**	**absent**	**t/χ^2**	***p***
Under 16													
Age*	Mean	8.00	10.85	2.70	**<.05**	9.17	10.89	1.70	.10	10.08	10.00	.07	.94
Gender	% Male	30.00	59.26	2.50	.15	38.89	61.11	1.78	.32	50.00	50.00	.00	1.00
Self Help	% Partly able/Able^2	50.00	88.89	6.51	**<.05**	72.22	83.33	.64	.69	75.00	79.17	.08	1.00
Mobility^1	% Fully mobile^3	80.00	81.48	.01	1.00	83.33	77.78	.18	1.00	91.67	75.00	1.42	.38
Vision^1	% Normal	80.00	92.59	1.20	.29	88.89	94.44	.36	1.00	91.67	91.67	.00	1.00
Hearing^1	% Normal	100.00	96.15	. 40	1.00	100.00	94.12	1.09	.49	100.00	95.65	.54	1.00
Speech^1	% Partly verbal/Verbal	80.00	92.59	1.20	.29	94.44	83.33	1.13	.60	100.00	83.33	2.25	.28
16 and over													
Age*	Mean Rank	17.56	13.85	67.00	.28	13.77	15.75	85.50	.54	18.40	14.29	43.00	.33
Gender	% Male	66.67	60.00	.12	1.00	54.55	66.67	.43	.67	80.00	58.33	.83	.62
Self ^{1}Help	%Partly able/Able^2	11.11	50.00	3.99	.10	36.36	38.89	.02	1.00	60.00	33.33	1.25	.34
Mobility^1	% Fully mobile^3	55.56	70.00	.57	.68	63.64	66.67	.03	1.00	80.00	62.50	.56	.63
Vision^1	% Normal	88.89	80.00	.34	1.00	90.91	77.78	.83	.62	80.00	83.33	.03	1.00
Hearing^1	% Normal	100.00	100.00	#	#	100.00	100.00	#	#	100.00	100.00	#	#
Speech^1	% Partly verbal/Verbal	33.33	52.63	.91	.44	50.00	44.44	.08	1.00	60.00	43.48	.45	.64

Bold text indicates if there is a significant difference in the demographic variables between individuals with and without challenging behaviour.

Results show that in the ‘under 16s’ TSC group, individuals who engaged in self injury were significantly younger and less able than those who did not self injure. No other significant differences were found.

In addition to demographic variables, it was hypothesised that other person characteristics including low mood, socialisation and communication difficulties, hyperactivity and impulsivity and compulsive, stereotyped and repetitive behaviours would differ between individuals with and without self injury and aggression (Arron et al., 2011; Hyman et al., 2002; Oliver et al., 2009). In order to test this hypothesis, Mann-Whitney U tests (and independent samples t tests where appropriate) were conducted to compare scores between individuals with and without self injury, aggression and destruction of property in people with TSC in the ‘under 16s’ and’16 and over’ groups. Tables 4.5 and 4.6 show these results.

Table 4.5. Differences in affect, autism spectrum behaviours, hyperactivity and repetitive behaviours between individual with and without self injury and aggression in the 'under 16s' TSC group.

		Median (Interquartile range)				
Measure	**Sub-Scale**	**Challenging behaviour present**	**Challenging behaviour absent**	**U Score**	***p* value**	**Effect size**
Under 16						
Self Injury						
MIPQ	Mood	16.00 (13.00-17.25)	21.00 (18.00-24.00)	32.00	**<.001**	-.59 *(large)*
	Interest and Pleasure	15.00 (8.00-17.50)	19.00 (13.50-21.25)	70.50	**<.05**	-.37 *(medium)*
SCQ	Communication	9.00 (5.88-10.75)	5.00 (4.00-9.00)	92.50	.14	-.24 *(small-medium)*
	Socialisation	8.50 (4.50-13.00)	5.00 (2.00-10.00)	92.00	.14	-.25 *(small-medium)*
	Repetitive behaviour	4.50 (3.25-6.25)	3.50 (1.00-6.00)	99.50	.22	-.21 *(small-medium)*
TAQ	Over-activity	21.00 (16.50-29.75)	5.50 (2.00-16.00)	62.00	**<.05**	-.42 *(medium-large)*
	Impulsivity	22.50 (15.75-24.00)	8.50 (2.75-23.00)	50.00	**<.01**	-.48 *(medium-large)*
RBQ	Compulsive behaviour	.00 (.00-17.50)	2.00 (.00-5.25)	117.00	.52	-.11 *(small)*
	Stereotyped behaviour	9.00 (2.75-12.00)	.00 (.00-4.25)	55.50	**.01**	-.47 *(medium-large)*
	Insistence on sameness	3.50 (2.75-5.50)	2.00 (0.00-4.00)	77.50	**<.05**	*-.34* *(medium)*
	Repetitive use of language*	9.00 (5.00-11.00)	3.50 (.00-5.75)	30.50	**.01**	*-.46* *(medium- large)*
	Restricted preferences*	6.00 (4.00-8.00)	4.00 (.00-7.00)	57.00	.19	*-.23* *(small-medium)*

Bold text indicates if there is a significant difference in the person characteristics between individuals with and without challenging behaviour.

Table 4.5 continued. Differences in affect, autism spectrum behaviours, hyperactivity and repetitive behaviours between individual with and without self injury and aggression in the 'under 16s' TSC group.

		Median (Interquartile range)				
Measure	**Sub-Scale**	**Challenging behaviour present**	**Challenging behaviour absent**	**U Score**	***p* value**	**Effect size**
Under 16						
Aggression						
MIPQ	Mood	17.50 (15.00-21.00)	22.00 (17.50-24.00)	79.50	**.01**	-.43 *(medium-large)*
	Interest and Pleasure	17.50 (11.75-20.00)	19.00 (12.00-21.50)	140.00	.48	-.12 *(small)*
SCQ	Communication	6.75 (4.00-9.00)	5.00 (3.50-10.69)	158.50	.91	-.02 *(small)*
	Socialisation	6.50 (4.75-10.00)	3.00 (2.00-11.50)	132.50	.35	-.16 *(small)*
	Repetitive behaviour	4.50 (3.25-6.25)	3.00 (1.00-4.50)	97.50	**<.05**	-.16 *(small)*
TAQ	Over-activity	19.50 (8.00-28.25)	5.00 (1.00-7.00)	67.50	**<.01**	-.38 *(medium)*
	Impulsivity	19.50 (10.00-23.00)	6.00 (1.00-16.50)	70.00	**.01**	-.46 *(medium-large)*
RBQ	Compulsive behaviour	2.00 (.00-15.50)	1.00 (.00-3.50)	132.00	.32	-.16 *(small)*
	Stereotyped behaviour	5.50 (.00-9.25)	.00 (.00-2.00)	91.00	**<.05**	-.39 *(medium)*
	Insistence on sameness	3.00 (1.50-5.50)	1.50 (0.00-3.25)	104.00	.06	*-.31* *(medium)*
	Repetitive use of language*	7.00 (2.75-10.00)	2.00 (.00-5.00)	54.50	**.01**	*-.47* *(medium-large)*
	Restricted preferences*	5.00 (4.00-7.75)	1.00 (.00-5.00)	74.50	.06	*-.33* *(medium)*

Bold text indicates if there is a significant difference in the person characteristics between individuals with and without challenging behaviour.

Table 4.6. Differences in affect, autism spectrum behaviours, hyperactivity and repetitive behaviours between individual with and without self injury and aggression in the '16 and over' TSC group.

		Median (Interquartile range)				
Measure	**Sub-Scale**	**Challenging behaviour present**	**Challenging behaviour absent**	**U Score**	***p value***	**Effect size**
16 and over						
Self Injury						
MIPQ	Mood	18.50 (15.00-20.84)	20.50 (16.25-21.75)	65.00	.24	-.22 *(small-medium)*
	Interest and Pleasure	10.50 (7.25-14.86)	11.00 (6.00-15.50)	81.50	.69	-.07 *(small)*
SCQ	Communication	13.00 (12.25-13.00)	11.19 (7.25-13.00)	43.00	**<.05**	-.43 *(medium-large)*
	Socialisation	14.50 (10.25-15.00)	11.00 (7.50-13.00)	45.00	**<.05**	-.40 *(medium-large)*
	Repetitive behaviour	4.50 (3.25-6.00)	3.50 (2.00-5.00)	70.50	.35	-.17 *(small)*
TAQ	Over-activity	19.50 (6.25-20.19)	9.00 (1.25-15.00)	60.50	.16	-.26 *(small-medium)*
	Impulsivity	21.25 (16.00-23.00)	13.50 (5.50-19.00)	49.00	.05	-.36 *(medium)*
RBQ	Compulsive behaviour	1.00 (.00-7.25)	.00 (.00-2.21)	75.00	.42	-.15 *(small)*
	Stereotyped behaviour	8.50 (4.00-11.50)	5.00 (.75-8.00)	54.00	.18	-.25 *(small-medium)*
	Insistence on sameness	2.00 (.00-4.50)	.00 (.00-4.00)	78.50	.55	*-.11* *(small)*

Bold text indicates if there is a significant difference in the person characteristics between individuals with and without challenging behaviour.

Table 4.6 continued. Differences in affect, autism spectrum behaviours, hyperactivity and repetitive behaviours between individual with and without self injury and aggression in the '16 and over' TSC group.

		Mean (SD)				
Measure	**Sub-Scale**	**Challenging behaviour present**	**Challenging behaviour absent**	***t***	***p*-value**	**Effect size**
Self injury						
	Repetitive use of language*	7.50	3.78	1.30	.23	*-.56*
		(.71)	(3.87)			*(large)*
	Restricted preferences*	8.00	5.67	1.51	.17	*-.34*
		(.00)	(4.64)			*(medium)*
Aggression		**Median (Interquartile range)**		**U Score**	***p value***	**Effect size**
MIPQ	Mood	19.00	20.00	91.50	.74	-.06
		(14.00-22.00)	(17.50-21.23)			*(small)*
	Interest and Pleasure	10.00	11.45	91.50	.74	-.06
		(6.00-16.00)	(8.00-15.00)			*(small)*
SCQ	Communication	13.00	11.38	83.00	.45	-.14
		(8.00-13.00)	(7.25-13.00)			*(small)*
	Socialisation	11.00	12.00	93.00	.79	-.05
		(8.00-14.00)	(9.50-14.50)			*(small)*
	Repetitive behaviour	4.00	4.00	91.50	.73	-.06
		(2.00-5.00)	(2.00-5.50)			*(small)*
TAQ	Over-activity	15.00	9.00	70.00	.19	-.24
		(5.00-20.25)	(1.50-16.00)			*(small-medium)*
	Impulsivity	20.00	13.00	48.50	**<.05**	-.42
		(15.00-23.00)	(4.50-18.00)			*(medium-large)*
RBQ	Compulsive behaviour	2.00	0.00	53.00	**<.05**	-.44
		(.00-6.00)	(.00-.50)			*(medium-large)*
	Stereotyped behaviour	7.00	6.00	80.00	.52	-.12
		(3.00-11.00)	(1.50-8.00)			*(small)*
	Insistence on sameness	3.00	.00	70.50	.16	*-.26*
		(.00-4.00)	(.00-4.00)			*(small-medium)*
	Repetitive use of language*	4.25	4.57	.13	.90	*-.04*
		(3.86)	(4.04)			*(small)*
	Restricted preferences*	6.25	6.00	.09	.93	*-.03*
		(4.19)	(4.62)			*(small)*

Bold text indicates if there is a significant difference in the person characteristics between individuals with and without challenging behaviour.

In the under 16s TSC group, affect and levels of interest and pleasure were significantly lower in individuals who engaged in self injury compared to those who did not. Levels of activity, impulsivity, insistence on sameness, repetitive language and stereotyped behaviours were significantly higher in individuals who engaged in self injury compared to those who did not. Similarly, individuals who engaged in aggressive behaviour had significantly lower affect and higher levels of activity, repetitive language and repetitive, impulsive and stereotyped behaviours compared to individuals who did not engage in aggression. Each of these differences was associated with a medium-large effect size.

In the 16 and over TSC group, individuals who showed self injury had significantly greater communication and socialisation difficulties and higher levels of impulsivity compared to individuals who did not show self injury. Individuals who engaged in aggression had higher levels of impulsivity and compulsivity compared to individuals who did not engage in aggressive behaviour. Again, each of these significant differences between individuals with and without self injury and aggression was associated with a medium-large effect size.

The final hypothesis proposed that pain and discomfort would differ between individuals with and without self injury, aggression and destruction of property in the TSC group. In order to test this hypothesis Mann-Whitney U tests were conducted on GDQ and NCCPC-R scores between individuals with and without challenging behaviour. When assessing self injurious behaviour, item 8 was excluded from the GDQ as this refers to self injury, which could confound the results. Table 4.7 shows the results from this analysis.

Table 4.7. Differences in pain and gastro-oesophageal distress between individual with and without self injury and aggression in the TSC group.

Measure	**Median (Interquartile range)**		**U score**	***p* value**	**Effect size (r)**
	Challenging behaviour present	**Challenging behaviour absent**			
Under 16					
Self injury					
GDQ score	16.50 (11.75-21.88)	4.00 (2.50-10.50)	48.00	**<.01**	-.50 *(large)*
NCCPC-R	26.00 (18.50-39.63)	9.00 (4.75-14.75)	27.00	**<.001**	-.62 *(large)*
Aggression					
GDQ score	12.00 (6.25-17.25)	3.00 (2.00-8.00)	91.00	**<.05**	-.38 *(medium)*
NCCPC-R	20.00 (13.00-35.00)	7.00 (3.00-10.00)	39.00	**<.001**	-.65 *(large)*
Destruction of property					
GDQ score	12.00 (7.25-19.50)	4.00 (3.00-12.50)	92.50	.08	-.29 *(small-medium)*
NCCPC-R	20.50 (11.50-42.50)	9.00 (4.25-14.00)	61.50	**<.01**	-.46 *(medium-large)*
16 and over					
Self injury					
GDQ score	11.00 (4.00-14.00)	10.50 (2.25-13.75)	53.00	.08	-.32 *(medium)*
NCCPC-R	12.00 (5.50-18.50)	12.00 (5.00-19.00)	57.50	.13	-.28 *(small-medium)*
Aggression					
GDQ score	7.00 (5.00-18.00)	11.00 (4.00-14.00)	95.50	.88	- .03 *(small)*
NCCPC-R	20.00 (10.00-36.00)	12.00 (5.50-18.50)	57.50	.06	-.35 *(medium)*
Destruction of property					
GDQ score	14.00 (7.00-19.50)	10.50 (3.50-14.00)	36.50	.17	-.25 *(small-medium)*
NCCPC-R	25.00 (14.00-40.50)	12.00 (5.25-18.75)	22.50	**<.05**	-.40 *(medium-large)*

Bold signifies significant differences between the challenging behaviour present and challenging behaviour absent groups

The pattern of results was different for the under 16s and the 16 and over groups. In the under 16s, individuals with TSC who engaged in self injury, aggression and destruction of property had significantly higher scores on the NCCPC-R, which suggests they had higher levels of overall pain. In the under 16s group, gastro-oesophageal distress scores were significantly higher in individuals who engaged in self injury and aggression compared to those who did not. No differences were found in gastro-oesophageal distress scores between individuals who engaged in destruction of property and those who did not. In the 16 and over group, scores on the NCCPC-R were higher in individuals who engaged in destruction of property compared to those who did not but no differences were observed in individuals who engaged in self injury or aggression compared to those who did not. Gastro-oesophageal distress scores were not significantly different between individuals who engaged in self injury, aggression or destruction of property compared to those who did not, although medium effect sizes were found in comparisons of gastro-oesophageal distress scores between individuals who engaged in self injury and aggression and those who did not.

4.4.3 Assessing the potentially confounding influence of level of intellectual disability

As shown in table 4.5, under 16's with TSC who showed self injurious behaviour were significantly younger and less able than those who did not. In order to evaluate whether these differences in age and intellectual disability confounded the differences found in factors such as mood, activity and pain between individuals who engaged in self injury compared to those who did not, a binary logistic regression analysis was conducted. In this test, self injury (present or absent) was the dependent variable. The predictor variables of interest were those shown to significantly differ between individuals who did and did not engage in self injury in the under 16s TSC group. The predictive value of these variables was assessed after factoring out the influence of age and level of intellectual disability. Table 4.8 shows the results from this analysis.

Table 4.8. Results from a binary logistic regression showing the association between mood, interest and pleasure, impulsivity, activity, stereotyped behaviour, pain and gastro-oesophageal distress and self injury when controlling for level of intellectual disability and age. Participants were individuals with TSC under the age of 16.

	B	***SE* β**	**Wald's χ^2**	***df***	***p***	***E* $^{\beta}$ (odds ratio)**
Controlling for level of intellectual disability						
Self injury Present x MIP; mood subscale	-.51	.21	5.99	1	**<.05**	.60
Self injury Present x MIP; Interest and Pleasure subscale score	-.12	.09	1.82	1	.18	.89
Self injury Present x TAQ; impulsivity subscale	.15	.07	5.30	1	**<.05**	1.16
Self injury Present x TAQ; Over activity subscale	.08	.04	3.83	1	.05	1.08
Self injury Present x RBQ; Stereotyped behaviour subscale	.22	.10	4.65	1	**<.05**	1.25
Self injury Present x RBQ; insistence on sameness	.31	.18	3.08	1	.08	1.36
Self injury Present x RBQ; repetitive use of language^{A}	.34	.16	4.75	1	**<.05**	1.40
Self injury Present x GDQ total	.11	.07	2.80	1	.09	1.12
Self injury Present x NCCPC total	.13	.06	5.02	1	**<.05**	1.14

	B	***SE* β**	**Wald's χ^2**	***df***	***p***	***E* $^{\beta}$ (odds ratio)**
Controlling for age						
Self injury Present x MIP; mood subscale	-.70	.29	5.84	1	**<.05**	.50
Self injury Present x MIP; Interest and Pleasure subscale score	-.25	.10	6.00	1	**<05**	.78
Self injury Present x TAQ; impulsivity subscale	.15	.07	4.84	1	**<.05**	1.17
Self injury Present x TAQ; Over activity subscale	.08	.04	4.14	1	**<.05**	1.08
Self injury Present x RBQ; Stereotyped behaviour subscale	.24	.10	5.38	1	**<.05**	1.27
Self injury Present x RBQ; insistence on sameness	.58	.25	5.21	1	**<.05**	1.78
Self injury Present x RBQ; repetitive use of language^{A}	.34	.16	4.88	1	**<.05**	1.41
Self injury Present x GDQ total	.11	.05	4.40	1	**<.05**	1.12
Self injury Present x NCCPC total	.12	.05	7.07	1	**<.01**	1.13

^{A}Verbal participants only

Bold text indicates significant associations between self injury and person characteristics that remain significant after the influence of level of intellectual disability is controlled.

The results show that, when controlling for age, all of the variables shown to differ between individuals with and without self injury in the under 16 TSC group were significantly associated with self injury once the influence of age was factored out. When controlling for level of intellectual disability, interest and pleasure, gastro-oesophageal distress, insistence on sameness and impulsivity were no longer significantly associated with the presence of self injury.

4.5 **Discussion**

This was the first study to investigate the rates of challenging behaviour in TSC relative to other genetic syndromes and the first study to describe the person characteristics associated with challenging behaviour in individuals with TSC. These characteristics included mood, activity, behaviours associated with ASD, repetitive and compulsive behaviours and behavioural indicators of pain.

In comparison to individuals with Down syndrome, the rates of self injury, aggression and destruction of property were higher in TSC but the differences were not statistically significant. The prevalence of self injury was 27% for individuals under the age of 16 with TSC and 31% for individuals over the age of 16. These results are consistent with previous research in individuals with TSC, although the prevalence of self injurious behaviour reported in this study is at the higher end of estimates (Hunt, 1983; Staley et al., 2008; de Vries et al., 2007). Prevalence rates for aggression of 50% and 37.% were reported in the under 16s and 16 and over groups respectively, which again fall within the range of estimates reported in other studies (Hunt, 1983; Staley et al., 2008; Kopp et al., 2008; de Vries et al., 2007). It is perhaps not surprising that the rates found in this study most closely reflect those found by deVries et al., (2007) as participants in both studies were recruited from parent/carer organisations.

Based on the previous literature regarding other genetic syndromes (Arron et al., 2011), it was hypothesised that certain characteristics would be different in people with self injury and aggression compared to people without these challenging behaviours. Consistent with this hypothesis, low mood, high levels of activity and impulsivity, stereotyped behaviours and repetitive use of language were significantly associated with self injurious and aggressive behaviour in individuals with TSC. In addition, lower levels of interest and pleasure, and increased repetitive and stereotyped behaviours were associated with self injury. Compulsive behaviour was significantly different between individuals with and without aggression. These findings show that there are differences in the personal characteristics of people with and without challenging behaviour. If operant learning processes were the only explanation for challenging behaviour, these differences in person characteristics would not be expected. Therefore, these findings, in addition to the different rates of challenging behaviour between syndrome groups, support the notion that operant learning processes cannot be the only explanation for all cases of challenging behaviour. Therefore, in some cases at least, internal factors, such as pain and discomfort, may contribute towards the development of challenging behaviour. There may be value in using these person characteristics or indicators of pain and discomfort to identify individuals with intellectual disability who are likely to engage in challenging behaviour.

The frequency of behavioural indicators of pain was also shown to be significantly different between individuals with and without self injury, aggression and destruction of property in people with TSC. It is not clear whether pain is the cause or the result of self injury in individuals with TSC. Previous literature suggests that self injury could moderate the perception of pain caused by ongoing health problems (Melzack & Wall 1965/1982; Woolf & Slater 2000, see section 1.3.6.4). Alternatively, the injuries sustained through self injury could cause an increase in pain. The association between pain and aggressive behaviour or destruction of property could be explained by the theory that pain acts as a setting event (Carr & Smith, 1995; Carr & Blakeley-Smith, 2006; Carr et al., 2003; McGill, 1999) (see section 1.3.8). This theory suggests that pain is likely to increase the chance of challenging behaviour occurring during situations that are typically associated with challenging behaviour. For example, challenging behaviour that is associated with the removal of demands may be more likely to occur when pain is present (Carr & Blakeley-Smith, 2006).

When the difference in pain scores were compared between individuals with and without self injury, aggression and destruction of property, it was noted that the influence of pain on challenging behaviour appeared to be more pronounced in the under 16s group compared to the 16 and over group. In the under 16s group, scores on the GDQ and the NCCPC-R were significantly higher in individuals with all three forms of challenging behaviour compared to individuals who did not engage in challenging behaviour. However, in the '16 and over' group, only the NCCPC-R scores were significantly higher in individuals who engaged in destruction of property compared to those who did not. This could indicate that pain influences the development rather than maintenance of challenging behaviour in individuals with TSC. Once established, challenging behaviour may become influenced by operant learning processes (see section 1.3.1), which supersede the influence of pain. After a behaviour becomes associated with environmental factors, it may then be expressed, irrespective of the level of pain experienced by the individual.

Another finding of this study was that the level of intellectual disability was associated with self injury in the under 16s group, which is similar to previous findings which reported self injury prevalence rates of 69%, 34% and 17% in people with severe/profound, mild and no intellectual disability respectively (de Vries, 2010). When intellectual ability was controlled for, significant associations between self injury and activity level, gastro-oesophageal distress and levels of interest and pleasure were no longer significant. These characteristics are all indicators of pain (Berg et al., 2007; Breau et al., 2004, Merkel et al., 1997; Luzzani et al., 2003) (see section 3.4.3). It might be suggested that individuals with higher levels of intellectual disability also have more clinical features of TSC, and are therefore more likely to suffer pain linked to health problems (de Vries et al., 2007). Thus, by controlling for intellectual disability, important differences in health and pain could also be lost. This issue would also emerge in research with other genetic syndromes where multiple systems within the body are affected such as Cornelia de Lange syndrome (Bhuiyan, Klein, Hammond, van Haeringen, Mannens, Berckelaer-Onnes, et al., 2006), Williams syndrome (Bruno, Rossi, Thüer, Córdoba & Alday, 2003), Charge syndrome (Sanlaville & Verloes, 2007) and DiGeorge syndrome (Baldini, 2004). Therefore it is important at this stage to not regard with certainty apparent associations between pain and challenging behaviour as being an artefact of increased level of intellectual disability.

4.5.1 <u>Limitations and directions for future research</u>

Participants were recruited to this study through a parent/carer support group. These organisations may attract a particular demographic of members, for example, membership may be biased towards parents of children with higher levels of challenging behaviour. This could mean that the rates of challenging behaviour reported in this study do not accurately represent the population of people with TSC as a whole. However, as a way of assessing the validity of the measures used in this study, the rates of challenging behaviour in other genetic syndromes, with well documented behavioural phenotypes, were assessed using the same methodology. The rates of challenging behaviour reported for Fragile X syndrome and

Cornelia de Lange in this study reflect those documented in previous research (Hyman et al., 2002; Symons et al., 2003), supporting the validity of the methodology employed in this study.

A methodological limitation of this study meant that it was not possible to accurately measure each participant's level of intellectual disability. The only measure of ability was the Wessex Questionnaire, in which informants are asked to rate their child's ability to independently complete self help tasks. During early childhood years, a child would not typically be able to perform these tasks without help. Therefore, it is not possible to ascertain from the Wessex data alone whether the young children had a developmental disability or whether they were developing typically and had just not reached this developmental stage. Previous research has employed the same methodology of grouping all individuals with TSC together regardless of ability level, suggesting that this is a difficulty that repeatedly occurs (Smalley, Burger & Smith, 2011, Staley et al., 2008; Lewis, Thomas, Murphy & Sampson, 2004; Baker et al., 1998). This is problematic as there is evidence to suggest that rates of challenging behaviour are different between different ability groups in TSC (de Vries et al., 2007). This highlights the need for future research to explore the rates of challenging behaviour and associated person characteristics in groups with well defined ability levels. This is important for two reasons. Firstly, these characteristics could potentially be used to identify individuals at risk of challenging behaviour, which might be different in people of different ability levels. Secondly, understanding the characteristics associated with challenging behaviour may help to inform causal models of challenging behaviour across different syndrome groups, which in turn, could have implications for intervention.

In this study, it was not possible to compare the challenging behaviour between children and adults with TSC as the inclusion criteria for each age group was different. Only adults with intellectual disability were included whereas children of any ability level were included. Thus, any differences observed between children and adults could have been confounded by differences in ability level. Given the large range of ability levels in TSC (Joinson et al., 2003; Shepherd & Stephenson, 1992; Webb et al., 1991) and the association between level of intellectual disability and challenging behaviour (Emerson et al., 2001b), it would be valuable for future research to explore the rates of challenging behaviour in different age groups that are matched on ability level. This information could be used to describe how the profile of challenging behaviour in people with TSC changes with age, which could have implications for intervention. It would also be interesting to study this for a second reason. Health problems associated with pain persist into adulthood in TSC (Smalley, et al., 1994). If challenging behaviour is associated with pain in TSC, it would be expected for rates of challenging behaviour to remain high into adulthood. This is different to previous research in individuals with intellectual disability for which an overall reduction in challenging behaviour with age has been reported (Holden & Gitlesen, 2006).

A final limitation of this study is that some of the measures used have only been validated for use with either adults or children although they have been used with all participants in this study. For example, the Mood, Interest and Pleasure questionnaire has only been validated with adults (Ross & Oliver 2003; Aman & Singh, 1986) and the Non-Communicating Children's Pain Checklist has only been validated for use with children (Breau et al., 2000; Breau et al., 2002). However, the Mood, Interest and Pleasure Questionnaire has been used extensively with children in a range of syndrome groups and has good face validity (i.e. low mood in Cornelia de Lange syndrome and high mood in Angelman syndrome) (Arron et al., 2011). Also, the Non-Communicating Adults Pain Checklist, which is adapted from the NCCPC-R and has 14 of the original items on the NCCPC-R, has been shown to be sensitive to detecting pain in adults with intellectual disability (Lotan, Ljunggren, Johnsen, Defrin, Pick

& Strand. 2009). Also, 19 of the 27 items on the NCCPC-R were shown to be significantly higher in adults during a pain episode (during a vaccination) compared to a non-pain episode, suggesting the NCCPC-R may be valid for use in adults (Lotan et al., 2009). Furthermore, when used with an adult population, scores on the NCCPC-R were significantly higher in individuals with chronic self injury compared to those without (Symons et al., 2009a). Further research would be useful to confirm the validity of using these measures in both child and adult populations.

4.5.2 Conclusion

In conclusion, self injury, aggression and destruction of property have been shown to be high in people with TSC, a high risk group for pain, although the risk of these behaviours occurring in people with TSC is not significantly greater than the risk of these behaviours in people with Down syndrome, a low risk group for pain. However, the number of behavioural indicators of pain (rated on the NCCPC-R and GDQ) was shown to be significantly higher in individuals with TSC who engaged in self injury, aggression and destruction of property compared to those who do not engage in these behaviours. Therefore, these findings suggest that diagnosis of a genetic syndrome associated with painful health conditions may not necessarily mean that challenging behaviour is more likely to occur in that population. However, it may be the case that diagnosis of a genetic syndrome such as TSC, which is associated with painful health conditions, may be a useful indicator that, if challenging behaviour is present, pain may be an influential factor.

CHAPTER 5

PAIN ASSESSMENT TOOLS; ASSESSING THE PSYCHOMETRIC PROPERTIES OF THE FLACC, NCCPC-R AND DIRECT OBSERVATIONS OF BEHAVIOURAL INDICATORS OF PAIN

5.1 Preface

The results of the previous studies have demonstrated that challenging behaviour can be associated with pain and also a number of person characteristics in people with intellectual disability. As pain is subjective and personal, it is difficult to identify in this population. The next study considers the use of observational pain measures as a method for identifying pain in people with intellectual disability. Whilst these are the most widely used methodology, their psychometric properties have, as yet, only been tested when pain is likely to be evident. When used to identify previously unrecognised pain, the reliability and validity of the assessments when pain is not established as likely to be present should be appraised. In this chapter, several psychometric properties of the FLACC, NCCPC-R and directly observable behavioural indicators of pain are assessed during times not previously defined as probably painful or pain free. The findings of this chapter will inform the methodology employed in chapters 6 and 7.

5.2 Introduction

Individuals with intellectual disability are at higher risk for developing health conditions than the general population (Allerton, Welch & Emerson, 2011; Van Schrojenstein Lantman-de Valk et al., 2000). Many of these health problems are associated with pain and discomfort, which would typically be identified through self report (Van Schrojenstein Lantman-de Valk et al., 1997). Self-report is compromised for individuals with intellectual disability who have insufficient expressive communicative skills to report their internal states (Adams & Oliver, 2011). This inherent difficulty with identifying and quantifying pain and poor health in people with disabilities is likely to contribute to the under-diagnosis of medical conditions within this population (Howells, 1986; Malviya et al., 2005). Apart from the importance of identifying pain for treatment of medical problems, there are broader implications as pain can be associated with challenging behaviour in people with intellectual disability (Carr & Owen-DeSchryver, 2007; Hartman et al., 2011). Therefore, identifying and treating pain is increasingly recognised as an essential component of assessment protocols for challenging behaviour.

In an attempt to overcome the problem of identifying pain in people with intellectual disability, research has focussed on the development of observational tools designed to quantify behavioural indicators of pain. Courtemanche et al. (2012) produced operationally defined behaviours based on the 30 items of the NCCPC-R (Breau et al., 2004), which were coded to produce a second-by-second account of the expression of pain behaviours. The inter-observer reliability of coding behaviour in this way was shown to be good to excellent (Fleiss, 1981). This is the only example of research using observations of behavioural indicators of pain on a continuous time sampling basis. Typically, ratings are made on the frequency of behaviours after a pre-defined observation time, such as two hours (Breau et al., 2003; Hartman et al., 2008; Symons & Danov, 2005; Symons et al., 2009a).

The most commonly employed observational rating scale, which requires the rater to record the presence of behavioural indicators of pain over a set time period, is the Non-Communicating Children's Pain Checklist-Revised (NCCPC-R). This lists 30 behaviours indicative of pain and discomfort (Breau et al., 2004). These include facial expressions, body movements, physiological changes, such as disruptions to eating and sleeping patterns, negative vocalisations and changes in social behaviour such as becoming withdrawn. NCCPC-R ratings are typically based on a two-hour observation. The Face, Legs, Activity, Cry, Consolability (FLACC) (Merkel et al., 1997) is another observational pain measure. This tool has only five subscales, which allows it to be administered quickly and easily by an observer. The five subscales are scored from zero to two; behaviours assigned to a score of zero indicate behaviour of an individual with no pain, a score of two lists behaviours associated with high levels of distress such as 'crying steadily' and 'screams and sobs' in the 'cry' subscale. A score of one lists behaviours that are indicative of less severe distress. The observer chooses the behaviour, and therefore the score, which best describes the child's behaviour throughout the observation.

Some of the psychometric properties of these measures have been assessed. Caregivers have completed the NCCPC on separate occasions, clearly identifiable as painful events (e.g. bee sting), distressful but not painful events (e.g. participant denied access to watching television) and calm events (e.g. listening to a story). The NCCPC differentiated between distressful and painful events, indicating that items on the NCCPC are specific to pain and not distress linked to other causes (Breau et al., 2000). The revised NCCPC (NCCPC-R) asks respondents to rate the frequency of pain behaviours rather than making present/absent judgements. The NCCPC-R was shown to have good inter-episode consistency across pain events and non-pain events. This suggests that pain behaviours and non-pain/baseline behaviours remain consistent over time (Breau et al., 2002). The Non-Communicating Children's Pain Checklist-

Post operative Version (NCCPC-PV) has also been shown to have good inter-rater reliability between primary caregivers and researchers (Breau et al., 2002), highlighting the possibility that the NCCPC-PV could be used to quantify pain reliably in children with intellectual disabilities by observers who are unfamiliar with the child.

The FLACC has also been shown to have good psychometric properties (Malviya et al., 2006) (see section 5.3.3.1). The FLACC had excellent inter-rater reliability when completed by two nurses following observations of children with intellectual disabilities who had undergone elective surgery (interclass correlation coefficient, ranging from .76 to 90). Scores on the FLACC correlated significantly with scores on a pain measure completed by parents and self report ratings made by the 23% of the children deemed able to self-report, suggesting good criterion validity. The FLACC scores also decreased after the administration of analgesics, suggesting good construct validity. Additionally, two nurses were asked to re-rate a selection of videos after four weeks. Results confirmed the FLACC had good test-retest reliability.

Although both the FLACC and NCCPC-R have strong psychometric properties, neither has been used with a population of people for which a painful episode has not already been identified. Thus, although these pain tools overcome some of the difficulties involved in measuring pain in children with intellectual disability, their psychometric properties in samples in which pain is not established as likely to be present have not been assessed.

In summary, people with intellectual disability are at high risk of health conditions that are associated with pain and discomfort. Current prevalence estimates of pain related health problems are likely to be underestimated as many health conditions may not be identified within this group. There is increased interest in the development of observational pain assessment tools that can be used to quantify pain in non-verbal individuals. Identifying and treating pain in this group may have positive implications for the management of challenging behaviour in people with intellectual disabilities. A number of observational measures have been shown to have good psychometric properties when pain is likely to be present. However, the psychometric properties of these tools to identify likely cases of pain in children who have no known health problems have not yet been tested. The first stage in being able to determine if this is possible, is to investigate whether these measures can be used reliably during times that are not previously defined as 'pain events'. Additionally, it is useful to assess the relationship between scores on these assessments to inform future research and clinical assessments. For example, the FLACC or NCCPC-R measures shown to be associated with the most robust reliability will be used in chapter six to compare pain scores in individuals with functional verses non-functional challenging behaviour. Additionally, the behavioural indicators of pain, coded during experimental functional assessments, that correlate most strongly with FLACC scores will be used in chapter 7 and the temporal relationship between the selected behavioural codes and challenging behaviour will be assessed.

5.2.1 Aims

The aim of this study is to assess the psychometric properties of the NCCPC-R, FLACC and direct observations of behavioural indicators of pain when observations are not restricted to previously defined pain or non-pain conditions. The following evaluations will be undertaken:

1. The inter-rater reliability of the FLACC will be assessed. This will include the inter-rater reliability between parent and teacher, primary and secondary researchers, and the primary researcher with a teacher.

2. The stability over time of the modified NCCPC-R and FLACC. FLACC ratings will be compared over five days and at three separate occasions on the same day. Ratings on the modified NCCPC-R will be compared within a two month time window.
3. The concurrent validity of the FLACC, modified NCCPC-R and direct observation of pain indicators will be assessed.

5.2.2 Expected findings

Given the transient nature of many forms of pain, the stability of pain scores is expected to be moderate. The strength of the correlation between ratings conducted at the same time is expected to be good, whereas the reliability between ratings conducted on the same day but at different times is expected to be moderate to low. Given the different target behaviours on the FLACC, NCCPC-R and direct observations of behavioural indicators of pain, concurrent validity between subscale items on these measures is expected to be fair. However, the 'total' scores should all reflect the overall pain experienced by the participants. Consequently, the correlation between the total FLACC, NCCPC-R scores and total behavioural indicators of pain are expected to be good.

5.3 Method

5.3.1 Recruitment and participants:

Children diagnosed with a genetic syndrome associated with neurodevelopmental disability or ASD aged 4-15 (inclusive) were included. Participants were identified from a database held at the Cerebra Centre for Neurodevelopmental Disorders, University of Birmingham. Parents were sent information sheets (appendix K), detailing the study aims and protocol, and consent forms (appendix L). Individuals interested in participating were then provided with research materials. A subgroup of individuals were visited by the researcher and completed experimental functional assessments in addition to the standard questionnaires, interviews and parent and teacher observations. This visit also allowed researchers to conduct direct observations of behavioural indicators of pain. The subset of participants who took part in this extended protocol are referred to as the 'direct observation group'. Table 5.1 provides a description of the participants included in this study.

Table 5.1. Mean age, mean age equivalent scores for expressive communication, receptive language, personal daily living skills and motor skills, percentage of males and percentage normal vision and normal hearing.

	Direct Observation group	All participants
N	29	40
Mean Age (years)	10.79	10.72
(SD)	(2.92)	(2.97)
Male N	12	19
(%)	(41.4)	(47.5)
Mean expressive communication age equiv.^{A}	10.79	15.78
(SD)	(2.92)	(11.82)
Mean receptive language age equiv.^{A}	15.48	25.53
(SD)	(9.89)	(33.77)
Mean gross motor skills age equiv. ^{A}	18.97	24.83
(SD)	(11.26)	(16.40)
Mean personal daily living skills age equiv. ^{A}	16.83	46.40
(SD)	(8.25)	(155.09)
% Normal vision	75.9	79.5
% Normal hearing	79.3	84.6

^{A}Age equivalent scores taken from the Vineland Adaptive Behavior Scale. Measured in months.

Table 5.1 describes the participants. Participants within the 'direct observation sample' all have poor expressive communication skills (see section 5.3.2). The 'direct observation sample' has a lower mean expressive and receptive language skills as well as lower mean ability with gross motor and daily living skills.

5.3.2 Procedure

All parents were invited to complete the questionnaires and observations of their children. A subgroup of 29 participants who scored below 19 on the expressive communication subscale of the Vineland Adaptive Behavior Scale (VABS) (raw score, see appendix Z), formed the "direct observation group". These participants were visited by two researchers who completed an experimental functional assessment (see section 6.3.3.3.2) and conducted direct observations of behavioural indicators of pain. The same primary researcher was present at all visits and conducted the experimental functional assessments. The secondary researcher was not consistent across visits and had the role of filming the assessments.

In order to assess the inter-rater reliability of the FLACC, observations were compared between parents and teachers, teachers and researchers, and the primary and secondary researchers. For the purpose of assessing the inter-observer reliability between parent and teacher ratings, parents and teachers completed five FLACC observations on the same days as each other but at different times. To assess the inter-observer reliability between researchers, who were both unfamiliar with the participant, the primary and secondary researchers simultaneously but independently completed three FLACC observations with participants in the ‘direct observation’ group. During one of these observations, a teacher simultaneously completed a FLACC observation to enable comparisons to be made between teacher ratings and primary researcher ratings.

The parent and teacher FLACC ratings completed over five separate days were used to assess the stability of the FLACC over time. This was also assessed using the three separate researcher FLACC ratings. Parents completed two modified NCCPC-R questionnaires (for explanation of the modification see section 5.3.3.2). The first modified NCCPC-R was completed during the same week as the FLACC assessments and the second was completed within two months of the first. This allowed the stability of the modified NCCPC-R to be assessed.

To assess the concurrent validity of the FLACC, NCCPC-R and direct observations of pain behaviours, parent and teacher FLACC scores were summed across the five observations and researcher FLACC scores were summed across the three observations to provide 'total' scores. Parent FLACC totals were compared to the modified NCCPC-R ratings completed within the same week as the FLACC observations. Teacher FLACC totals were compared to the teacher NCCPC-R. Experimental functional assessments were conducted with the 'direct observation' group. Video footage of the high attention conditions was coded for behaviours that are associated with pain, such as negative vocalisations or agitated body movement. These codes were based on behaviours listed in the FLACC. A list of these codes can be found in appendix M. The percentage of time that the participants engaged in these behaviours was compared to the primary researcher FLACC total.

5.3.3 Measures

5.3.3.1 **Face Legs Activity Cry Consolability (FLACC)** (Merkel et al., 1997**;** appendix N)

The FLACC is a five item observational tool used to rate the pain behaviour of a child occurring in approximately the last five minutes. The FLACC was shown to have good criterion validity (Spearman's *p*= 0.65–0.87; P < 0.001) and can be used reliably by care staff unknown to the child as well as familiar carers (interclass correlation coefficients ranging from 0.76 to 0.90, Kappa statistic 0.44–0.57) (Malviya et al., 2006; Voepel-Lewis et al. 2002). Pain scores on the FLACC decreased after analgesics had been administered, demonstrating the strong construct validity (n= 20; 6.1 ± 2.5 vs 2.2 ± 2.4; P < 0.001) (Malviya et al., 2006; Voepel-Lewis et al. 2002).

5.3.3.2 **Non-Communicating Children's Pain Checklist- Revised (NCCPC-R)** (Breau et al., 2004; appendix O & P)

The Non-communicating Children's Pain Checklist was shown to have good internal consistency when using retrospective parental ratings and was also shown to be reliable over two pain events when ratings were made by the same observer (α=0.66) (Breau et al., 2000).

Excellent internal consistency during two separate pain episodes was also reported for the NCCPC-R (repeated measure ANOVA, main effect of episode; F(1, 54) = .001, *P* = .978) (Breau et al., 2002) showing that the revised version of the NCCPC is also reliable over time. For this study a modification of the NCCPC-R was employed for the parent observations only (not the teacher NCCPC-R). This modification was that behaviours were rated over a one week period rather than a two hour time window. This modification has been made in previous studies (Breau et al., 2003; Symons et al., 2009) although the reliability or validity of using the measure in this way has not been reported. In this study, the modification was made in order to identify chronic pain behaviour rather than behaviour indicative of acute pain episodes.

5.3.4 Data Analysis

Data were analysed using SPSS 19.0 software. All data were tested for normality using Shapiro-Wilk test and non-parametric tests were used where necessary. Due to the small data set and the large number of tied ranks, Kendall's Tau was used when assessing the correlation between subscale scores on the various measures. Spearman's Rho was used when assessing the correlation between total scores. For all correlation analyses, a result of .10-.30 will represent a low/poor correlation, .30-.50 will indicate a 'moderate/fair' association, .50-.70 will be 'large/high' and a correlation above .70 will represent a very high correlation (Hopkins, 1997. Cited in Kotrlik & Williams, 2003).

5.4 **Results**

5.4.1 Inter-rater reliability of the FLACC

The inter-rater reliability of the FLACC was tested using the parent, teacher and researcher ratings. The results of this correlational analysis are shown in table 5.2.

Table 5.2. Inter-rater reliability of the FLACC. Correlations comparing; teacher and parent observations on the same day at different times, primary and secondary researcher observations at the same time and the primary researcher with teacher observations at the same time. Correlations are calculated with the total FLACC scores and each subscale score.

		Day 1		Day 2		Day 3		Day 4		Day 5	
		N	Correlation coefficient	N	Correlation coefficient	N	Correlation coefficient	N	Correlation coefficient	N	Correlation coefficient.
Parent X teacher	Total FLACC[A]	31	.31	31	.21	29	.26	24	.42	10	.16
	Face subscale	29	.20	29	.15	27	-.17	23	.19	9	.36
	Legs subscale	31	.12	31	.06	30	.26	24	.09	10	.38
	Activity subscale	31	.04	31	.10	30	.19	22	.41	10	-.33
	Cry subscale	30	.06	30	.09	29	-.05	23	.45	10	-.22
	Consolability subscale	31	.27	31	.10	30	.20	24	.13	10	[B]

		Observation 1		Observation 2		Observation 3	
		N	Correlation coefficient	N	Correlation coefficient	N	Correlation coefficient
Primary Researcher X Secondary Researcher	Total FLACC[A]	29	**.73**	29	**.68**	28	**.73**
	Face subscale	29	.07	29	.22	28	**.53**
	Legs subscale	29	.41	28	.42	28	**.69**
	Activity subscale	29	.66	29	.21	28	**.69**
	Cry subscale	29	.26	29	**.60**	28	**.74**
	Consolability subscale	29	[B]	29	[B]	27	-.04
Primary Researcher X Teacher (same time)	Total FLACC[A]	29	**.57**				
	Face subscale	29	**.54**				
	Legs subscale	29	.47				
	Activity subscale	28	**.73**				
	Cry subscale	29	**.79**				
	Consolability subscale	28	**.68**				

[A] A total FLACC score was calculated for each observation by summing the subscales of the FLACC. Correlation was calculated between the parent and teacher observations that occurred on the same days. Any observations that took place on different days were excluded.

[B] Correlation analyses could not be completed due to lack of variance in the data.

Bold and underlined: very high correlation, **Bold:** high correlation, Underlined; moderate correlation

Table 5.2 continued. Inter-rater reliability of the FLACC. Correlations comparing teacher and primary researcher observations on the same day at different times. Correlations are calculated with the total FLACC scores and each subscale score.

		N	Correlation coefficient
Primary Researcher X Teacher (same day, different time)	Total FLACC[A]	15	.15
	Face subscale	15	.19
	Legs subscale	15	[B]
	Activity subscale	15	**.61**
	Cry subscale	15	-.30
	Consolability subscale	15	-.07

[A] A total FLACC score was calculated for each observation by summing the subscales of the FLACC. Correlation was calculated between the parent and teacher observations that occurred on the same days. Any observations that took place on different days were excluded.
[B] Correlation analyses could not be completed due to lack of variance in the data.
Bold and underlined: very high correlation, **Bold:** high correlation, Underlined: moderate correlation

As expected, reliability was strongest when observations were conducted simultaneously. For example, between the primary and secondary researchers, the correlation between total FLACC scores across the three observations was high to very high (between .63 and .78). It was not possible to calculate the correlation between primary and secondary researcher ‘consolability’ subscale scores because of a lack of variance in the data, with too many tied ranks. This was due to a high proportion of zero scores on the ‘consolability’ subscale recorded by both researchers. Therefore, reliability was high for absence of behavioural indicators of pain when using this scale. Also, when observations were conducted simultaneously between the primary researcher and the teacher, the correlation for total FLACC score was high (.57). Unsurprisingly, the correlation was low between observations that were conducted at different times, even if on the same day. For example, the correlation between primary researcher and teacher total FLACC ratings was .15.

As mentioned earlier (see section 6.3.3.2.1), the teacher NCCPC-R ratings were completed following a two hour observation, whereas the parent NCCPC-R ratings were made based on behaviour over a one-week time frame. Because of this, the inter-rater reliability on the

NCCPC-R was not assessed as part of this study but the results from this analysis can be found in appendix Q.

5.4.2 Stability over time of the FLACC and modified NCCPC-R

In order to test whether these measures were stable over time, correlation analyses were completed between total and subscale scores across the separate FLACC and modified NCCPC-R ratings. It would be expected that many behaviours associated with chronic pain would remain fairly stable, thus the two week-long ratings on the modified NCCPC-R would be associated with moderate-high correlation. However, it was expected that observations conducted over shorter time periods, such as FLACC five minute observations, would be sensitive to fluctuation of pain behaviours associated with the possible natural ebb and flow of pain. Therefore, a low-moderate correlation was expected between the FLACC ratings across different days. As expected, on the NCCPC-R, the association between the two ratings was ‘moderate’ for the total score (r= .49) and all subscale scores (.33 to .46) apart from the ‘body’ subscale, which was ‘poorly’ correlated (.27).

The table below (table 5.3) displays the correlation between the five days of FLACC ratings provided by the teacher and parent.

Table 5.3. A correlation matrix of parent total FLACC ratings across the five days of observations and the teacher total FLACC ratings across the five days of observations.

	Parent FLACC				
	Kendall's tau (N)	Day 2	Day 3	Day 4	Day 5
Parent FLACC	Day 1	.34 (39)	.33 (39)	.44 (37)	.33 (36)
	Day 2	-------	**.61** (39)	**.52** (37)	**.60** (36)
	Day 3	-------	-------	**.52** (37)	.42 (36)
	Day 4	-------	-------	-------	**.68** (35)

	Teacher FLACC				
	Kendall's tau (N)	Day 2	Day 3	Day 4	Day 5
Teacher FLACC	Day 1	.36 (37)	.37 (36)	.30 (33)	.21 (24)
	Day 2	-------	.09 (36)	.22 (33)	.46 (24)
	Day 3	-------	-------	.22 (33)	.24 (23)
	Day 4	-------	-------	-------	.48 (22)

Bold and underlined: very high correlation, **Bold:** high correlation, Underlined: moderate correlation

Given the transient nature of pain, day to day scores on the FLACC would be expected to be 'poorly' correlated. Contrary to expectations, the parent FLACC scores were reasonably stable across the five days of observations, with all correlation coefficients in the 'moderate to high' range. However, this was not the case with the teacher FLACC ratings, for which half of the correlation outcomes were 'low' between the two ratings, which is in line with the predicted outcome.

Kendall's Tau correlation analyses were also used to compare the three FLACC ratings completed by each of the researchers. Results demonstrated 'moderate' correlation coefficients between .30 and .39. This means that the FLACC ratings were relatively unstable over time suggesting that the number of behavioural indicators of pain fluctuated on the day of the research visit (see appendix S for correlation matrix). Again, this would be expected given the unstable nature of pain.

5.4.3 Concurrent validity of the FLACC, modified NCCPC-R/ NCCPC-R and behavioural indicators of pain

To test the concurrent validity of the FLACC and modified NCCPC-R, the scores on each of these measures were compared and also compared to real time coding of pain related behaviours. Tables 5.4 and 5.5 display the results from these concurrent reliability assessments.

The behaviours listed on the FLACC and NCCPC-R subscales are not the same. Therefore, it was expected that the subscale scores would be associated with a ‘low-moderate’ correlation only. However, ‘total score’ on both the FLACC and NCCPC-R should be assessing overall pain and would therefore be expected to be moderate to highly correlated. The results displayed in table 5.4 show that, as expected, the subscale FLACC and NCCPC-R scores were associated with a poor correlation when completed by parents. However, surprisingly the total scores on these measures were also associated with a poor correlation when completed by parents. Conversely, when completed by the teacher, the total NCCPC-R and FLACC scores were moderately correlated along with several subscale scores.

Table 5.4. A correlation matrix comparing the parent FLACC and the modified parent NCCPC-R and the teacher FLACC with the teacher NCCPC-R. Ratings were made in the same week. Total scores and subscale scores were compared.

	Parent FLACC						
	Correlation coefficient (N)	Total FLACC	Face subscale	Legs subscale	Activity subscale	Cry subscale	Consolability subscale
Parent modified NCCPC-R	Total NCCPC	.28 (37)	.20 (36)	.17 (37)	.15 (37)	.18 (36)	.34 (37)
	Vocal subscale	.20 (36)	.22 (35)	.18 (36)	.15 (36)	.14 (35)	.25 (36)
	Social subscale	.12 (37)	.12 (36)	.03 (37)	.05 (37)	.17 (36)	.27 (37)
	Facial subscale	.20 (37)	.25 (36)	.05 (37)	.09 (37)	.19 (36)	.36 (37)
	Activity subscale	.02 (37)	-.08 (36)	-.01 (37)	.16 (37)	-.04 (36)	.04 (37)
	Body subscale	.17 (37)	.19 (36)	.23 (37)	.06 (37)	.14 (36)	.24 (37)
	Physiological subscale	.18 (37)	.11 (36)	.14 (37)	.13 (37)	.17 (36)	.34 (37)
	Eat and sleep subscale	.07 (37)	.05 (36)	.05 (37)	.03 (37)	.05 (36)	.27 (37)

	Teacher FLACC						
	Correlation coefficient (N)	Total FLACC	Face subscale	Legs subscale	Activity subscale	Cry subscale	Consolability subscale
Teacher NCCPC-R	Total NCCPC	.39 (33)	-.01 (33)	.24 (33)	.41 (33)	.20 (33)	.35 (33)
	Vocal subscale	.46 (32)	.20 (32)	.30 (32)	.42 (32)	.40 (32)	.44 (32)
	Social subscale	.33 (32)	.05 (32)	.22 (32)	.28 (32)	.26 (32)	.35 (32)
	Facial subscale	.14 (33)	-.01 (33)	.02 (33)	.06 (33)	.15 (33)	.25 (33)
	Activity subscale	.10 (30)	-.12 (30)	.35 (30)	.28 (30)	.01 (30)	.24 (30)
	Body subscale	.22 (32)	-.03 (32)	.26 (32)	.44 (32)	.21 (32)	.37 (32)
	Physiological subscale	.31 (32)	-.16 (32)	.20 (32)	**.59** (32)	.16 (32)	.48 (32)
	Eat and sleep subscale	.28 (25)	.02 (25)	.44 (25)	.29 (25)	.13 (25)	.35 (25)

Bold and underlined: very high correlation, **Bold:** high correlation, Underlined: moderate correlation

Table 5.5. A correlation matrix comparing the researcher FLACC ratings and ratings of direct observations of behavioural indicators of pain.

		Researcher 1 FLACC					
	Kendall's tau (N)	Total FLACC	Face subscale	Legs subscale	Activity subscale	Cry subscale	Consolability subscale
Direct observations	Negative vocalisations	.42 (28)	.06 (28)	.07 (28)	**.53** (28)	.32 (28)	-.04 (28)
	Negative facial expressions	.23 (28)	.08 (28)	.12 (28)	.15 (28)	.27 (28)	-.09 (28)
	Gesturing to body part	-.09 (28)	-.16 (28)	-.08 (28)	.05 (28)	-.11 (28)	-.05 (28)
	Non-goal directed activity	-.03 (28)	-.21 (28)	-.11 (28)	.10 (28)	.07 (28)	-.08 (28)
	Cry	.07 (28)	-.16 (28)	-.09 (28)	.20 (28)	.15 (28)	-.06 (28)
	Rigid posture	.22 (28)	**.91** (28)	.14 (28)	.24 (28)	.20 (28)	-.09 (28)
	Restless legs	.29 (28)	-.09 (28)	-.03 (28)	**.69** (28)	-.03(28)	-.08 (28)
	Total pain indicators	.37 (28)	.01 (28)	.09 (28)	**.53** (28)	.25 (28)	-.09 (28)

Bold and underlined: very high correlation, **Bold:** high correlation, Underlined: moderate correlation

The target behaviours in the direct observations were based largely on the items listed in the FLACC subscale, for example the direct observation 'cry' corresponded with the 'cry' subscale on the FLACC and 'rigid posture' and 'non-goal directed activity' were adapted from behaviours described on the 'activity' subscale of the FLACC. In these cases, the correlation between the FLACC subscales and the direct observations was expected to be moderate to high. This was not the case as the association between these observed behaviours and FLACC subscales were poor. 'Total pain indicators' and total FLACC scores should both be measuring overall pain and therefore, scores were expected to be moderate-largely correlated. The total FLACC and 'total pain indicators' had a correlation of .37, which is associated with a moderate correlation.

5.5 Discussion

This is the first study to test the psychometric properties of the FLACC and NCCPC-R when observations are not restricted to previously defined pain or non-pain conditions. Testing the psychometric properties of measures used in this way is an essential step towards being able to identify pain in individuals where there is no a-priori knowledge of underlying pain or health problems. The inter-rater reliability of the FLACC, stability over time of the FLACC and modified NCCPC-R and the concurrent validity of the FLACC, NCCPC-R and direct observations of behavioural indicators of pain were assessed.

Consistent with previous research, when observations were conducted at the same time, the inter-rater reliability of the FLACC between the primary research and teacher was strong (Voepel-Lewis et al. 2002). This was the first study to test the consistency of FLACC ratings over time. Correlation between ratings made at different time points were expected to be low-moderate due to the transient nature of pain. This was the case with teacher and researcher ratings whereas the parent ratings remained relatively stable. The reliability and sensitivity of the teacher and researcher FLACC to detect fluctuations in pain behaviours, suggests that it is a reasonably robust measure for identifying behavioural indicators of pain times not previously defined as 'pain events' and is therefore recommended for future research.

Concurrent validity was also assessed. Despite moderate correlations between teacher ratings on the FLACC and NCCPC-R, correlations between the parent FLACC and modified parent NCCPC-R were poor. Behaviours listed on the FLACC subscales were used as a guide to create codes for direct observations of behavioural indicators of pain. Therefore, a high correlation was expected between these two measures. However, subscale FLACC scores and proportion of time spent engaging in directly observable pain behaviours were poorly correlated. Conversely, the association between the 'total pain indicator' variable, created by combining all behavioural codes, and total FLACC score was moderate. As the FLACC has previously been shown to have good construct validity, these findings suggest that using highly specific codes for direct observations may not be robust enough to identify pain whereas combining the behavioural codes to form a 'total pain indicator' variable appears valid and should therefore be applied to future research.

The pattern of results in this study suggests that pain behaviours were stable in the home setting whereas they fluctuated across observations at school. This may reflect variability in pain, and hence pain behaviour, or reveal systematic differences in how parents and teachers make ratings of pain behaviour. Differences between parent and teacher ratings of children's behaviour have been demonstrated repeatedly in previous research (Greenbaum, Dedrick, Prange & Friedman. 1994; Grietens, Onghena, Prinzie, Gadeyne, van Assche, Ghesquiere, et al., 2004; Satake, Yoshida, Yamashita, Kinukawa & Takagishi, 2003; Verhulst & Akkerhuis, 1989). It would be beneficial for future studies to assess the inter-rater reliability of the FLACC between parents and teachers, with observations being conducted concurrently. This would provide a clearer indication of whether it is the behaviour or the approach taken by observers that varies.

It is most likely that the frequency of pain behaviours does vary over time. This is supported by findings that both teacher and researcher FLACC ratings fluctuate and that the inter-rater reliability between these observers is high when observations are conducted at the same time. Assuming that the frequencies of behavioural indicators of pain vary, it is important to note that this was not found in the parent ratings when using the FLACC. This implies that parents are less likely to identify or report changes in child behaviour than teachers. This may be because parent ratings are influenced by factors other than their child's behaviour. For example, they may expect their child to exhibit a high number of pain behaviours and therefore report these even when they are absent. Therefore, it may be worthwhile to complement parent ratings of pain with staff or teachers ratings to obtain a more comprehensive measure of pain behaviours across different environmental settings.

In this study, a modified version of the NCCPC-R was used. Observations were made over a one-week period rather than two hours. The rationale was to describe typical pain behaviour that may be indicative of chronic or recurrent acute pain as opposed to transient and acute episodes of pain. This strategy has been used in previous studies (Breau et al., 2003; Symons et al., 2009a). Results from this study demonstrate that correlation was moderate between two separate week-long NCCPC-R observations, indicating that 'typical pain' ratings remain fairly stable over time. However, taken in conjunction with other findings of this study, caution is advised when interpreting the results in this way. It may be the case that, even though parent modified NCCPC-R ratings correlated between two observations, this would not occur if ratings were made by other individuals, such as teachers.

The results of this study indicate that the FLACC has good inter-observer reliability when observations are conducted simultaneously. Additionally, the total FLACC and total NCCPC-R scores have good concurrent validity when observations are conducted by teachers. Therefore, the teacher FLACC and NCCPC-R are recommended as valid measures to be used in future research. Results also suggested that 'total pain indicators', which is a combination of all directly observable pain indicators, is the most valid variable to use when identifying pain by direct observations. Therefore, for a valid measure of pain when using direct observations, it is recommended to use a composite code such as 'overall pain indicators' rather than coding more specific fine grained behaviours.

CHAPTER 6

THE ASSOCIATION BETWEEN PAIN AND ENVIRONMENTAL FUNCTION OF CHALLENGING BEHAVIOUR IN PEOPLE WITH INTELLECTUAL DISABILITY

6.1 Preface

In previous chapters, the potential association between pain and challenging behaviour in people with intellectual disability has been investigated using different strategies and the psychometric properties of pain assessment tools suitable for this population have been appraised. In combination, the results point to the importance of the assessment of pain and a method for assessing pain alongside other well established causes of challenging behaviour such as operant learning. A next step is to implement both operant and pain assessments in practice to evaluate their relationship and trial their use. In this chapter, an assessment protocol is trialled which assesses challenging behaviour in non-verbal individuals with intellectual disability who are at high risk for challenging behaviour and examines the differences in pain and person characteristics in people with and without challenging behaviour associated with an environmental function.

6.2 Introduction

Challenging behaviour is a significant problem for many people with intellectual disabilities (Emerson et al., 2001b). It not only affects the health and wellbeing of the individual but also has emotional and financial implications for relatives and carers (Quine & Pahl, 1989; Qureshi, 1995). Researchers have become increasingly interested in the diverse possible causes of challenging behaviour in an effort to produce targeted and effective interventions. Two dominant theories that are emerging indicate that both pain and operant learning might be causal.

A number of research findings demonstrate that challenging behaviour in people with intellectual disability is associated with low mood (Hayes et al., 2011; Ross & Oliver, 2002b) and commonly occurs in individuals with a diagnosis of depression (Davis et al., 1997; Marston et al., 1997; Reiss & Rojahn 1993). This has lead to the claim that challenging behaviour may be an atypical symptom of depression in people with intellectual disability and therefore should be included in the diagnostic criteria for depression within this group. However, limitations in research methodology compromise the validity of this assertion (Davies, in review). For example, research has failed to address the possibility that a confounding variable may account for the co-occurrence of both low affect and the presence of challenging behaviour. Given the strong evidence that pain is independently associated with both low mood and challenging behaviour (Berg et al., 2007; Davis et al., 1997; Hayes et al., 2011; Ross & Oliver 2002b; Tsiouris, 2001), it is a reasonable assertion that pain could be a confounding variable that acts in this way (Davies, in review). Furthermore, the findings presented in chapter 3 demonstrate that individuals with low mood and challenging behaviour also engage in higher levels of activity, a behavioural indicator of pain, compared to individuals who do not engage in challenging behaviour or who have higher levels of mood. This finding supported the assertion that low mood, in combination with challenging behaviour, may be indicative of underlying pain in people with intellectual disabilities (see section 3.4.6).

There are a number of theories that attempt to explain the association between pain and challenging behaviour. First, the gate control theory suggests that stimulation of non-pain related neurones inhibits the transmission of signals originating from pain receptors in the body, thus moderating the perception of pain (Melzack & Wall, 1965/1982). More intense stimulation, such as self injury, may be required to have this moderating effect on pain in people with intellectual disabilities (Peebles & Price, 2012) (see section 1.3.6.4). Alternatively, some research has shown that self injury results in the release of endorphins (Rojahn et al., 2008), which could have an analgesic effect on the body, thus reducing the level of pain that is experienced due to underlying health problems (see section 1.3.6.2).

Research has also demonstrated an association between painful health conditions and challenging behaviour. This relationship was demonstrated in chapter 3 (see section 3.4.2). Individuals with ASD who engaged in a clinically significant number of behavioural indicators of gastro-oesophageal distress were significantly more likely to engage in self injury, aggression, destruction of property and stereotyped behaviours. Also, individuals with Cri du Chat syndrome who engaged in a clinically significant number of behavioural indicators of gastro-oesophageal distress were more likely to engage in self injurious behaviour. These findings build upon previous research, which has demonstrated the association between painful health conditions and challenging behaviour. For example Hartman et al. (2011) investigated self injury in a child with Congenital Hydrocephalus. Self injury directed around the participant's head (e.g. pulling hair, hitting head with other body parts or objects) became worse when her scalp was protruding compared to when it was flush with the skull table. Scalp protrusion would result in high levels of pain. Therefore, this case study demonstrates an increase of challenging behaviour targeted around the site of pain, during times of high pain. Self injury has also been shown to be more severe during times of menses when the women are likely to be experiencing abdominal discomfort (Carr et al., 2003).

Therefore, there is now sufficient evidence to suggest that underlying pain and discomfort may contribute to the development and severity of self injury. In this study the association between self injury, aggression and destruction and property with pain will be examined to extend the evidence generated in sections 3.4.2 and 4.4.1. Pain will be measured indirectly with the use of the FLACC (Merkel et al., 1997) and the NCCPC-R (Breau et al., 2004). These are observational pain measures in which respondents rate the presence of behavioural indicators of pain. Both of these pain measures have been shown to have good psychometric properties (Voepel-Lewis et al., 2002; Breau et al., 2000). In chapter 5, several psychometric properties of these measures were assessed during times which were not identified as being likely to be painful or pain free. The measures shown to have the most robust psychometric properties when used in this way are employed in this present study (see chapter 5, sections 5.4.1, 5.4.2, 5.4.3 & 5.5).

Although there is emerging evidence to support the pain theory of self-injury, this theory alone cannot account for the wealth of evidence suggesting that environmental factors influence challenging behaviour. Carr and Durand (1985) and Iwata et al. (1982/1994) set out a method for investigating the theory that challenging behaviour is shaped by reinforcing environmental consequences (see section 1.3.1). The environmental/ operant learning theory of challenging behaviour postulates that, over time, an association may be formed between challenging behaviour and rewarding consequences, thus making the behaviour previously associated with the rewarding consequence more likely to occur.

The association between challenging behaviour and environmental factors has been demonstrated repeatedly through the use of experimental functional analysis (Hanley et al., 2003) (see section 1.3.2). During experimental functional analysis, the social environment is systematically manipulated by repeatedly presenting test conditions, such as high attention and low attention. Differentiated levels of challenging behaviour across experimental conditions signify a possible relationship between challenging behaviour and the presence of particular environmental conditions. In a review of experimental functional analysis, Hanley et al., (2003) found that 80.10% of challenging behaviour assessed was associated with an environmental function with the most commonly identified function being negative reinforcement through the removal of demands. This estimate is likely to be inflated due to publishing biases, such as failure to publish studies which do not identify environmental functions (Easterbrook, Berlin, Gopalan & Matthews, 1991). It is, therefore, not possible to accurately predict the proportion of challenging behaviour associated with an environmental function in individuals with an intellectual disability. Despite this, there remains strong evidence in support of the environmental influence on challenging behaviour.

Although the empirical evidence for the operant learning theory of challenging behaviour is extremely strong, not all challenging behaviour can be explained in this way using ‘standard’ functional analysis conditions. It may be that most frequently used experimental designs fail to address all possible environmental functions. For example, Richards (2012) demonstrated that typical experimental functional analysis procedures, such as those employed by Carr and Durand (1985), failed to identify an environmental function for self-injury in children with ASD. The experimental procedure was then adapted to incorporate individualised ASD social environmental triggers for self injury identified by carers. These triggers, such as noises or preventing the child from completing an established routine or ritual, were incorporated into the functional analysis procedure. These adapted experimental functional analyses, were able to identify a function associated with 67% of self injury whereas traditional experimental functional analysis procedures were unable to attribute a function for any cases of self injury (Richards, 2012).

In summary, strong evidence suggests that environmental influences can explain some challenging behaviour. However, not all challenging behaviour can be explained by this theory as in some cases, no environmental function can be identified. Also, the operant learning theory of challenging behaviour is insufficient for explaining the differences in person characteristics observed between people with and without challenging behaviour.

The operant and pain\discomfort theories can be combined to produce a more comprehensive explanation of challenging behaviour. Carr and Smith (1995) introduced the notion of pain as a ‘setting event’. In this model, pain or ill health can act as an ‘establishing operation’ or motivational state that increases the rewarding nature of reinforcing consequence and therefore increases the likelihood of the behaviour previously associated with the rewarding consequence being performed (see section 1.3.8). To evaluate this principle, Carr and Blakeley-Smith (2006) studied the effectiveness of interventions for challenging behaviour in a group of school children with intellectual disability. They found that interventions which addressed physical ill health and reduced the aversive nature of the environment were the most effective at reducing challenging behaviour compared to responding only to the underlying physical discomfort.

Similarly, work completed by Applegate, Matson and Cherry (1999) using the Questions About Behaviour Function (QABF), an informant-based functional assessment questionnaire, shows that an individual’s challenging behaviour may be influenced by both environmental and biological factors. In addition to the ‘attention maintained’ and ‘demand escape’ functions addressed by typical experimental functional assessment and analysis, the QABF also addresses functions such as access to tangibles, self stimulation (non-social) and physical health problems.

In summary, previous research has shown that both environmental and biological factors are important in explaining challenging behaviour in people with intellectual disability. These factors may interact to influence challenging behaviour as suggested by the 'setting event' literature or pain and environmental factors may influence challenging behaviour at different times such as during its development and/or maintenance. For example, the gate control theory may explain how self injurious behaviours are initially introduced into the behavioural repertoire and then operant learning processes explain how challenging behaviours are maintained.

As yet, there is no established protocol for assessing challenging behaviour that addresses the influence of both pain and environmental factors. The implementation of a standard assessment procedure that addresses the possible influences of both pain and environmental factors on challenging behaviour would promote the use of effective and targeted interventions. It is also important to have an understanding of the proportion of challenging behaviour that can be explained by either underlying pain or environmental factors as this can help direct future research attention and funding to where it may have the greatest impact.

In addition to the pain and operant learning theoretical accounts, there is evidence for individual characteristics, such as low mood and high levels of activity, impulsivity and repetitive behaviours being associated with high rates of challenging behaviour (Bodfish et al., 1995; Oliver et al., 2011b; Oliver, Murphy, Hall, Arron & Leggett, 2003; Hayes et al., 2010; Ross & Oliver 2002b; Perry & Roy, 1997; Reiss & Rojahn 1993) (see sections 1.2 to 1.2.8). This finding has also been replicated in chapter 4. In people with Tuberous Sclerosis Complex, challenging behaviour was associated with high levels of activity, impulsivity, insistence on sameness, repetitive language and stereotyped behaviours. Challenging behaviour in this group was also associated with lower mood and lower levels of interest and pleasure. However, as yet, these characteristics have not been assessed in the context of functional and non-functional challenging behaviour to determine whether these characteristics are more or less strongly associated with challenging behaviour that is environmentally influenced.

6.2.1 Aims and hypotheses

The aims of this study are to:

1. Trial an assessment procedure and report the proportion of self injury, aggression and destruction of property maintained by social environmental factors. Positive and negative reinforcement hypotheses will be tested through demand/social escape and access to social attention experimental functional analysis conditions.

2. Compare pain scores in people with challenging behaviour maintained by social environmental factors compared to those presenting challenging behaviour with no identified environmental function.

3. Compare the levels of repetitive behaviours, mood, activity and impulsivity in individuals with and without environmentally influenced challenging behaviour.

It is predicted that:

1. Individuals engaging in self injury or aggression will score higher on pain measures than those not showing these behaviours.

2. Challenging behaviour will be associated with person characteristics including low affect and high levels of activity, impulsivity and repetitive behaviours.

6.3 Method

6.3.1 Recruitment and participants

Participants were recruited from a database held at the Cerebra Centre for Neurodevelopmental Disorders, University of Birmingham (see section 5.3.1). Participants in this study had to be under the age of 16 and have an intellectual disability. A subset of participants participated in experimental functional assessments in addition to the standard protocol, which included questionnaires, interviews and observations (see section 5.3.2). This subgroup of participants will be referred to as the 'direct observation' group. Participants were selected to be in the 'direct observation' group if they scored below 19 (raw score) on the expressive communication subscale of the Vineland Adaptive Behavior Scale (appendix Z).

6.3.2 Procedure

Initially, parents were invited to complete a postal questionnaire pack, which included measures relating to their child's behaviour. Following the return of the questionnaire pack, participants' teachers were asked to complete the NCCPC-R and FLACC observation measures and act as informants during the Challenging Behaviour Interview. Researchers also completed FLACC assessments for the direct observation group only. Challenging Behaviour Interviews occurred on the day of the research visit for the direct observation participants or over the phone at a time convenient to the teacher for all other participants.

Researchers conducted experimental functional analyses of challenging behaviour for participants in the direct observation group. These assessments were conducted at school in a private room with minimal distractions for the participant over a period of one school day and ensuring that the child maintained their typical school routine in terms of play and food breaks. Additional breaks from the functional analysis procedure were also programmed to ensure that the participant did not become tired or distressed. During these breaks, the researcher engaged with the participant in a way that was preferable to the child.

6.3.3 Measures

Detailed descriptions of each of these measures can be found in previous chapters. Therefore only brief summaries are provided below. The sections where detailed descriptions can be found are noted for each measure.

6.3.3.1 Questionnaire Measures

6.3.3.1.1 Mood, Interest and Pleasure Questionnaire- Short version (Ross et al., 2008) (see section 3.3.3.5, appendix D5).

Items on this questionnaire measure behaviours indicative of two key constructs of depression; low mood and lack of interest and pleasure.

6.3.3.1.2 Wessex Questionnaire (Kushlick et al., 1973). (see section 3.3.3.3, appendix D3).

The Wessex Questionnaire provides a proxy measure of intellectual disability.

6.3.3.1.3 The Activity Questionnaire (Burbidge & Oliver 2008). (see section 3.3.3.4, appendix D4).

Items on this questionnaire ask informants to rate activity and impulsivity in the people they care for.

6.3.3.1.4 Social Communication Questionnaire (SCQ: Rutter et al., 2003) (see section 4.3.3.6, appendix D7).

Items on the SCQ measure behaviours associated with a diagnosis of ASD.

6.3.3.2 Pain observation Measures

6.3.3.2.1 Non-Communicating Children's Pain Checklist-Revised (NCCPC-R: Breau et al., 2004) (see section 4.3.3.7, appendix P).

This is a 30 item questionnaire that rates the frequency of behavioural indicators of pain observed over a two hour window.

6.3.3.2.2 Face Legs Activity Cry Consolability (FLACC: Merkel et al., 1997) (see section 5.3.3.1, appendix N).

The FLACC consists of five subscales (face, legs, activity, cry and consolability). In this study, teachers were invited to complete one FLACC observation for five consecutive school days. The five daily subscale scores were summed to provide a total score for each subscale. The five daily FLACC scores (all subscales scores combined) were summed to provide the overall total score.

6.3.3.3 Assessments of challenging behaviour

6.3.3.3.1 Challenging Behaviour Interview (CBI: Oliver, McClintock, Hall, Smith, Dagnan & Stenfert-Kroese, 2003a) (appendix T & U).

The Challenging behaviour Interview (CBI) was used to assess the occurrence and severity of self injury and aggression. Part one of the interview identifies whether the challenging behaviour has occurred over the last month and part two assesses the severity of the behaviour. Severity is assessed by recording the frequency and duration of behaviour, the level of injury caused by the behaviour and the actions that were necessary when responding. Research has shown that the CBI has good inter-rater reliability (Pearson correlation .67 for part one and .48 for part two) and test-retest reliability (Pearson correlation .86 for part one and .76 for part two) (Oliver et al., 2003a). Comparisons with the Aberrant Behavior Checklist also show the CBI has good construct validity (Oliver et al., 2003a).

When conducting this interview, respondents were asked if the person they cared for had engaged in self injury that either has caused, or had the potential to cause, an injury. A description of self injury was provided to guide the informants' response;

> 'Non-accidental behaviours which producing temporary marks or reddening of the skin or cause bruising, bleeding or other temporary or permanent tissue damage.'

The same question was asked regarding aggression. Respondents were asked if the person they cared for had engaged in aggression that had either caused injury to another person or had the potential to cause injury to another person. The description for aggression was;

> 'A non-accidental, physical act involving physical contact with another person likely to result in pain or distress.'

6.3.3.3.2 Experimental Functional Analysis

An experimental functional analysis of self injurious behaviour, aggressive behaviour and destruction of property was conducted. Behaviour was defined as challenging when actual damage occurred or when there was the potential for damage to occur. Damage refers to any mark on the body resulting from self injury or aggression and in the case of destruction of property, any mark or impact on an object that was not already present.

The experimental functional analysis followed the structure first formulated by Carr and Durand (1985) and was designed to assess the influence of social reinforcement on challenging behaviour. The functional analyses consisted of alternating between the following five minute experimental conditions in an ABAC design:

- *Condition A- High attention/control condition;* The researcher maintained high levels of verbal and physical attention, whilst placing no demands on the participant. There were no planned consequences for challenging behaviour.

- *Condition B- Low Attention;* The researcher remained in close proximity to the participant but removed all attention. No demands were placed on the participant. Researchers responded to self injury with a standard statement of 'Don't do that X, you'll hurt yourself' and aggression with 'Don't do that, that hurts'. If an episode of property destruction occurred, the researcher responded with a statement of 'Don't do that, you'll break it'

- *Condition C- Demand:* The researcher prompted the child through a tabletop task that had been identified by a teacher as being challenging for the child. Prompts followed three stages: (1) verbal prompt only, (2) verbal prompt with a gesture/model and (3) verbal and physical prompt. For each trial, the researcher began with phase one of the hierarchy and if the participant did not respond or responded incorrectly, the researcher continued to the next phase of the prompt hierarchy. Social praise was provided when the task was completed (independently or prompted) and, after 3 to 5 seconds, the prompt hierarchy began again. If challenging behaviour occurred during this time, the researcher responded with a standard response of 'OK, we don't have to do that now' and the task was removed for a period of 10 seconds, or if challenging behaviour occurred during this time, after an additional 5 seconds of no challenging behaviour.

The different conditions of the experimental functional analysis allowed for different hypotheses to be tested. Condition A (high attention) allowed a baseline to be taken for comparison with the other two conditions. This condition allowed for a negative reinforcement through social escape hypothesis to be tested. No consequences for social escape were provided but if the child engaged in higher levels of challenging behaviour at a time of constant social contact compared to times of low social interaction (condition B, low attention) results would indicate an association between challenging behaviour and the removal of social attention. Condition B allowed a positive reinforcement through attention delivery hypothesis to be tested. If the child engaged in higher levels of challenging behaviour during times of low social interaction compared to times when social contact was not limited, it would suggest an association between challenging behaviour and the delivery of social contact as a reinforcing consequence. Finally, condition C (demand) allowed a negative reinforcement through escape from demands hypothesis to be tested. If the child engaged in higher levels of challenging behaviour during times of high demand compared to no demands, this would suggest an association between challenging behaviour and the removal of demanding tasks. If challenging behaviour was observed at a similar level across conditions, this was considered to be undifferentiated and suggests that challenging behaviour is not functional or maintained by social environmental variable.

6.3.4 Data Analysis

Data were analysed using SPSS 19.0 software. All data were tested for normality using Shapiro-Wilk test and non-parametric tests were used where necessary.

Participants were split into a challenging behaviour and a no-challenging behaviour group depending on the reports of teachers in the Challenging Behaviour Interview. Teacher reports were used rather than parental reports because they were more likely to accurately reflect the participants’ behaviour when at school where the experimental functional assessments took place. Also, in chapter 5, psychometric properties of the teacher FLACC and NCCPC-R ratings were shown have the strongest psychometric properties compared to parent pain ratings (see sections 5.4 to 5.4.3). Based on this finding, teacher pain ratings were selected for use in this study rather than parent pain ratings. Therefore, when comparing teacher pain ratings between individuals with and without challenging behaviour, it was deemed most appropriate to use teacher reports of challenging behaviour in order to categorise participants into ‘challenging behaviour’ and ‘no challenging behaviour’ groups. Participants who engaged in either self injury or aggression were categorised into the challenging behaviour group. This was based on the first item of the Challenging Behaviour Interview in which teachers were asked to report any self injury or aggression that they had observed in the previous month.

For the direct observation participants from the challenging behaviour group, D-Stat (Cliff, 1993) calculations were performed to compare the proportion of time spent engaging in challenging behaviour in the test conditions (low attention or demand) compared to the control condition (high attention) during the experimental functional analysis assessments. A D-stat of .50 indicates that challenging behaviour occurred for a higher proportion of time in half of the experimental conditions compared to the control condition. A D-stat of .75 demonstrates that the proportion of time spent engaging in challenging behaviour was higher in three out of four test conditions compared to the control condition. Moderate function is associated with a D-stat value of .5 or above and strong function is .75 or above. D-stat calculations were performed separately for self injury, aggression and destruction of property. In addition to this, all occurrences of self injury, aggression and destruction of property were combined in to one variable; overall challenging behaviour. D-stat calculations were also calculated for this ‘overall challenging behaviour’ variable.

6.4 Results

The proportion of children with intellectual disability reported to engage in self injury or aggression, with use of the teacher Challenging Behaviour Interview, was calculated. Table 6.1 shows the number of children reported to engage in self injury or aggression over the preceding month according to teacher responses on the Challenging Behaviour Interview.

Table 6.1. The number of participants reported to engage in self injury and aggression according to the Teacher Challenging Behaviour Interview. Descriptive information regarding the mode severity of the challenging behaviour is also reported.

			No Challenging Behaviour	Self Injury Only	Aggression Only	Self Injury and aggression
Direct observation group		N	5	4	7	12
		(%)	(17.86)	(14.29)	(25.00)	(42.86)
	SIB	Mode response to 'how long until you definitely see the behaviour again?'	--	In the next 15 minutes	--	By this time tomorrow*
		Mode duration of worst episode	--	Less than 5 minutes	--	Less than a minute*
		Mode duration of typical episode	--	Less than 5 minutes	--	Less than a minute*
	Agg.	Mode response to 'how long until you definitely see the behaviour again?'	--	--	By this time next month	By this time next week*
		Mode duration of worst episode	--	--	Less than a minute	Less than a minute
		Mode duration of typical episode	--	--	Less than a minute	Less than a minute
All participants		N	9	6	9	14
	SIB	Mode response to 'how long until you definitely see the behaviour again?'	--	In the next 15 minutes	--	In the next 15 minutes
		Mode duration of worst episode	--	Less than a minute	--	Less than a minute*
		Mode duration of typical episode	--	Less than a minute	--	Less than a minute
	Agg.	Mode response to 'how long until you definitely see the behaviour again?'	--	--	By this time next month	By this time next month
		Mode duration of worst episode	--	--	Less than a minute	Less than a minute
		Mode duration of typical episode	--	--	Less than a minute	Less than a minute

*Multiple modes exist. The least severe description of challenging behaviour is shown.

6.4.1 The association between pain and challenging behaviour

It was predicted that pain scores would be higher in individuals reported to engage in either self injurious behaviour or aggressive behaviour than those not showing these behaviours. Pain scores are the total scores on the teacher NCCPC-R, the teacher FLACC totals summed across the five days of observations and the researcher FLACC totals summed across the three observations. Table 6.2 show the results from this analysis.

Table 6.2. Mann-Whitney U test results. Comparisons between challenging behaviour and no-challenging behaviour groups using the teacher FLACC and NCCPC-R pain measures.

	Median (Inter-quartile Range)		U score	*p* value (1 tailed)	Effect size (r)
	No challenging behaviour	Challenging behaviour			
Researcher FLACC total summed over three observations	0.00 (.00-1.00)	0.00 (.00-2.00)	45.50	.24	.16
Teacher FLACC totals summed over five days	0.00 (.00-2.50)	6.00 (4.00-9.00)	31.50	**<.001**	.55
Teacher NCCPC total	2.00 (.00-7.00)	6.00 (2.25-10.00)	69.00	.06	.28

The results reported in table 6.2 show that significantly higher teacher FLACC total scores were observed in the challenging behaviour group compared to the no-challenging behaviour group. There was no significant difference observed between in the teacher NCCPC-R totals or researcher FLACC totals between the challenging behaviour and no challenging behaviour groups.

6.4.2 Person characteristics and challenging behaviour

It was hypothesised that challenging behaviour would be associated with low mood, high activity levels and high rates of repetitive behaviours and impulsivity. Table 6.3 show the results from Mann-Whitney U tests used to assess the difference in pain scores and other person characteristics between participants with and without challenging behaviour.

In table 6.3, the result show that although not significant, a lower MIPQ total score was observed in the challenging behaviour group, indicating a trend in the data between challenging behaviour and negative affect. No other significant differences were observed between the challenging behaviour and no-challenging behaviour groups.

6.4.3 The function of challenging behaviour

Tables 6.4 and 6.5 show the proportion of challenging behaviour associated with undifferentiated, attention maintained, demand escape, social escape and access to tangibles functions. All children who participated in experimental functional analysis assessments are included in the description below (tables 6.4 and 6.5), regardless of whether or not they were reported to engage in challenging behaviour on the Teacher CBI. Table 6.4 shows the proportion of individuals engaging in challenging behaviour associated with each type of function. In some cases, challenging behaviour was found to be associated with more than one environmental function. Table 6.5 shows the proportion of challenging behaviour that was associated with each type of function, including when challenging behaviour was associated with multiple functions. A D-stat value of .50 was used in table 6.5 (Cliff, 1993).

Table 6.3. Mann-Whitney U test results. Comparisons between challenging behaviour and no-challenging behaviour groups using The Activity Questionnaire, Mood Interest and Pleasure Questionnaire and the Repetitive Behaviour Questionnaire.

		Median (Inter-quartile Range)		**U score**	**P value (1 tailed)**	**Effect size (r)**
		No challenging behaviour	**Challenging behaviour**			
Mood interest and pleasure						
MIPQ	Total	39.00 (37.00-42.00)	37.00 (32.25-39.00)	79.50	.05	.27
	Mood subscale	22.00 (18.00-23.00)	19.50 (18.00-22.00)	91.50	.13	.20
	Interest and pleasure subscale	19.00 (14.50-21.00)	17.00 (15.00-19.00)	93.00	.13	.28
Activity levels						
TAQ	Total	26.00 (13.50-43.50)	35.69 (27.63-48.75)	87.00	.09	.22
	Impulsivity subscale	14.00 (9.00-21.00)	20.25 (12.25-23.00)	88.00	.10	.21
	Hyperactivity subscale	12.00 (7.50-23.00)	20.50 (11.25-26.00)	90.50	.11	.23
Repetitive and ritualistic behaviour						
RBQ	Total	22.00 (9.00-36.50)	13.00 (7.00-20.00)	92.50	.15	.18

Table 6.4. The proportion of challenging behaviour associated with an environmental function. Moderate function is associated with a D-stat value of .5 or above and strong function is .75 or above.

Topography	Undifferentiated	Moderate attention maintained	Strong attention maintained	Moderate demand escape	Strong demand escape	Moderate social escape	Strong social escape	Moderate access to tangibles	Strong access to tangibles
Self injury N (%)	14/19 (73.68)	2/19 (10.53)	2/19 (10.53)	2/19 (10.53)	2/19 (10.53)	1/19 (5.26)	0	0	0
Aggression N (%)	9/17 (52.94)	4 /17 (23.53)	3/17 (17.65)	3/17 (17.65)	1/17 (5.89)	0	0	1/17 (5.89)	0
Destruction of Property N (%)	10/18 (55.56)	1/18 (5.56)	0	6/18 (33.33)	4/18 (22.22)	1/18 (5.56)	0	1/18 (5.56)	0
Overall challenging behaviour N (%)*	13/28 (46.43)	5/28 (17.86)	4/28 (14.29)	8/28 (28.57)	4/28 (14.29)	1/28 (3.57)	0	2/28 (7.14)	1/28 (3.57)

* Self injurious behaviour, aggression and destruction of property combined to form one variable (overall challenging behaviour)

Table 6.5. Proportion of challenging behaviour associated with each type of environmental function.

	No distinguishable function	Attention maintained only	Demand escape only	Social escape only	Access to tangibles only	Attention maintained and demand escape	Social escape and access to tangibles
Self Injury N (%)	14/19 (73.68)	2/19 (10.53)	2/19 (10.53)	1/19 (5.26)	0/19 (0)	0/19	0/19
Aggression N (%)	9/17 (52.94)	4/17 (23.53)	3/17 (17.65)	0/17	1/17 (5.88)	0/17	0/17
Destruction of Property N (%)	10/18 (55.56)	1/18 (5.56)	6/18 (33.33)	0/18	0/18	0/18	1/18 (5.56)
Overall challenging behaviour* N (%)	13/28 (46.43)	4/28 (14.29)	7/28 (25.00)	1/28 (3.57)	2/28 (7.14)	1/28 (3.57)	0/28

*All challenging behaviours combined to form one variable (overall challenging behaviour)

Tables 6.4 and 6.5 show that, of the 19 individuals who engaged in self injurious behaviour during the experimental functional analyses, only five individuals engaged in self injury for which an environmental function can be identified. Conversely, approximately half of all observations of aggression and destruction of property were associated with an environmental function.

6.4.4 The association between pain and environmental function of challenging behaviour

An aim of the present study was to investigate if pain scores differ between individuals with challenging behaviour that is influenced by environmental factors compared to behaviour that is not environmentally influenced. In order to investigate whether this is the case, Kendall's Tau correlation analyses were conducted to examine the association between D-stat values and pain scores. Given the use of teacher reported pain behaviour (FLACC observations), only the participants who were reported to engage in challenging behaviour in the teacher CBI were included in this analysis. Table 6.6 shows the results from this analysis.

Table 6.6. Kendall's Tau analyses demonstrating the association between the highest absolute D-stat value for each topography of challenging behaviour and pain scores reported by the researcher and teacher.

	Researcher	Teacher totals calculated over five days						
Highest absolute D-stat value	FLACC total	FLACC total	FLACC face	FLACC legs	FLACC activity	FLACC cry	FLACC consolability	NCCPC total
Overall challenging behaviour T	.08	-.24	.06	-.41**	-.26	-.14	-.39*	.19
(N)	(23)	(22)	(22)	(22)	(22)	(22)	(22)	18
Self injurious behaviour T	.06	-.12	-.29	.12	-.17	-.05	-.06	.06
(N)	(16)	(16)	(16)	(16)	(16)	(16)	(16)	(16)
Aggressive behaviour T	.39*	-.05	.21	-.02	-.15	-.11	-.26	.22
(N)	(16)	(16)	(16)	(16)	(16)	(16)	(16)	(12)
Destruction of property T	-.01	.06	.32	-.30	.05	.17	-.22	.24
(N)	(15)	(15)	(14)	(14)	(14)	(14)	(14)	(11)
Highest d-stat value from self injury, aggression, destruction of property or overall challenging behaviour T	.15	-.34*	-.01	-.34*	-.23	-.26	-.46**	.16
(N)	(23)	(22)	(22)	(22)	(22)	(22)	(22)	(18)

*P<**.05, ** P<.01, ***P<.001**

Table 6.6 shows that function of challenging behaviour (D-stat value) was not significantly correlated with the majority of pain scores. However, in the cases where there was a significant association between D-stat value and pain scores, the majority of correlations were negative. This indicates that, where challenging behaviour is not associated with an environmental function, it is more likely to be influenced by pain. However, there was a significant positive correlation between researcher FLACC total and D-stat value for aggression. This indicates that when cases of aggression are associated with an environmental function, pain may also influence the behaviour.

To further investigate whether challenging behaviour not shown to be associated with an environmental function is more likely to be associated with pain, the participants were split into functional and non-functional challenging behaviour groups. Participants with a D-stat score of .50 or higher for self injury, aggression, destruction of property or overall challenging behaviour were categorised as functional. Mann-Whitney U tests were then used to test the difference in pain scores between these groups. Table 6.7 shows the results from this analysis. See appendix W for analysis of subscale scores.

Table 6.7. Mann Whitney U results. Comparisons of the functional behaviour and non-functional behaviour groups using the FLACC and NCCPC-R pain measures.

	Median (Inter-quartile Range)		U score	*p* value	Effect size (r)
	Non-functional challenging behaviour	Functional challenging behaviour			
Researcher FLACC total over three observations	0.00 (0.00-1.00)	0.00 (0.00-3.00)	55.00	.51	.12
Teacher FLACC totals over five days	9.50 (5.00-13.50)	6.00 (1.75-7.25)	27.50	.05	.42
Teacher NCCPC total	6.00 (2.50-14.50)	5.00 (3.50-9.50)	31.50	.92	.02

The results reported in table 6.7 indicate that there were no significant differences in pain scores between the functional and non-functional groups.

6.4.5 Person characteristics and function of challenging behaviour

Person characteristics shown to be associated with challenging behaviour were also compared between individuals with and without challenging behaviour associated with moderate environmental function. The results from this analysis are displayed in table 6.8.

Table 6.8 reports results showing that the difference in impulsivity score on the TAQ between the non-functional behaviour group compared to the functional behaviour group was approaching significance. No other significant differences between the groups were observed.

Table 6.8. Mann Whitney U results. Comparisons were made between the functional behaviour and non-functional behaviour groups using the Mood Interest and Pleasure Questionnaire, The Activity Questionnaire and the Repetitive Behaviour Questionnaire.

		Median (Inter-quartile Range)		U score	P value	Effect size (r)
		Non-functional challenging behaviour	Functional challenging behaviour			
Mood interest and pleasure						
	Total	33.00 (29.50-37.50)	37.00 (35.25-39.50)	44.00	.23	.25
MIPQ	Mood subscale	19.00 (16.50-20.50)	20.50 (18.75-22.00)	42.50	.19	.24
	Interest and pleasure subscale	15.00 (11.50-18.50)	17.00 (15.07-19.25)	45.50	.27	.23
Activity levels						
	Total	48.00 (38.00-53.50)	35.00 (27.86-44.00)	34.00	.07	.25
TAQ	Impulsivity subscale	25.00 (18.00-27.50)	18.50 (11.00-25.25)	26.50	<.05	.48
	Hyperactivity subscale	23.00 (20.00-24.00)	17.50 (10.50-21.25)	43.50	.22	.26
Repetitive and ritualistic behaviour						
RBQ	Total	12.00 (7.00-18.50)	12.00 (7.00-16.00)	50.50	.60	.29

6.5 Discussion

This study trialled an assessment procedure designed to identify the possible influences of operant conditioning and pain on challenging behaviour in people with intellectual disability. Experimental functional analyses were used to identify challenging behaviour associated with negative reinforcement through demand or social escape and positive reinforcement through the delivery of social attention. Observational pain measures were used to investigate whether there was an association between the total teacher and researcher FLACC and teacher NCCPC-R pain scores and the presence of self injury and aggression. Pain scores were also compared between children with challenging behaviour associated with environmental influences and children for whom an environmental function was not identified. Other person characteristics were assessed in terms of their association with self injury, aggression and destruction of property and to determine whether these characteristics differed between individuals with environmentally-functional verses non-functional challenging behaviour. This was the first study of its kind to investigate the influence of pain and environmental factors on challenging behaviour in people with intellectual disability. This is also the first attempt to produce an assessment protocol that incorporates the assessment of both of these variables on challenging behaviour.

This study found that, when using teacher reports 82.14% of participants were found to engage in challenging behaviour. This included self injurious and aggressive behaviour which either caused, or had the *potential* to cause injury. It is important to note that these findings cannot be generalised to challenging behaviour which is defined in a more conservative manner to only include behaviours which cause an immediate observable impact or injury. Using low-level behaviour which does not currently cause injury may result in an inflated estimated rate of challenging behaviour and also have further implications that are discussed later.

As hypothesised, significantly higher pain scores were observed in children reported to engage in self injury or aggression by their teachers compared to children who were reported not to engage in these behaviours. This supports the theory that pain can be associated with challenging behaviour. This reaffirms the importance of investigating possible health problems when challenging behaviours develop. However, person characteristics associated with pain such as hyperactivity and low affect did not differ significantly between children with and without challenging behaviour. This could be due to the fact that it is not each of these person characteristics in isolation that indicate the presence of pain, but more the combination of these characteristics that form the behavioural pattern exhibited by an individual who is in discomfort. This demonstrates the importance of using valid and sensitive measures of pain, which utilise all known behavioural indicators of pain.

An environmental function was identified for one quarter of participants who engaged in self injury and just under half the participants who engaged in aggression and destruction of property. Across all types of behaviour, a greater proportion of individuals engaged in behaviour with no discernable environmental function compared to the proportion of individuals who engaged in challenging behaviour for which an environmental function could be identified. This differs from the results reported in previous literature (Hanley et al., 2003), which indicate that a greater proportion of challenging behaviour is associated with environmental function than not. This discrepancy may highlight a publishing bias in the experimental functional analysis literature. Clinical studies with statistically significant results have been shown to be more likely to be published than those with non-significant findings and this bias is greatest in observational and laboratory based experimental studies (Easterbrook et al., 1991). Additionally, within this study, brief functional assessments were conducted over the course of one day. It may be the case that this method failed to identify a function for behaviour that is only evident during times of high pain (i.e. when pain acts as an establishing operation/setting event) or during other relevant establishing operations that were not present on the assessment day. This may lead to an underestimation of the proportion of functional cases observed in the present study. Furthermore, it may be that a smaller proportion of challenging behaviour was associated with an environmental function in this study compared to other research because this study included challenging behaviour that had the potential to cause injury as well as higher level, more severe behaviour. This low-level challenging behaviour may be less likely to be associated with an environmental function compared to more clinically significant challenging behaviour reported by other studies. Also, experimental functional assessments may have failed to identify function for the challenging behaviour observed in this study because the right environmental triggers were not tested. For example, when looking at the results of experimental functional analyses assessments with 152 participants, Iwata et al., (1994) demonstrated many different environmental functions that were not assessed within this study including positive reinforcement with food or materials only, negative reinforcement through the removal of social interaction, physical examinations and ambient stimulation such as noise. Furthermore, it may also be the case that the behaviours were associated with a specific learning history, involving a particular person or location, which was not assessed during these experimental functional assessments (Smith & Iwata, 1997).

Although it was not tested statistically, findings from the present study indicate that a greater proportion of self injury was classified as 'undifferentiated' compared to aggression and destruction of property. This could be the result of a proportion of self injury being associated with self stimulation, which would be unlikely to be a function of other forms of challenging behaviour. This difference is also consistent with a pain theory of challenging behaviour, such as the gate control theory (see sections 1.3.6-1 to 3.6.4), which suggest self injury may result from underlying pain and discomfort. This theory is unable to account for the emergence of other types of challenging behaviour such as aggression or destruction of property. Therefore, the difference observed between the proportion of self injury classified as 'undifferentiated function' compared to other forms of challenging behaviour may be evidence for a more significant role for pain in the development of self injury compared to other forms of challenging behaviour. This theory was also supported by the finding that D-stat for aggression was positively associated with pain scores whereas this was not the case for self injury. This indicates that pain may interact with environmental factors to influence aggression whereas pain may play more of a direct role in influencing some cases of self injury.

In summary, this study has demonstrated the need for a two pronged approach when investigating challenging behaviour, which incorporates the assessment of both pain and environmental influences on challenging behaviour. No significant differences were observed between pain scores in children with and without functional behaviour, although results from a correlation analysis suggested that level of environmental function was negatively associated with pain, when total FLACC scores were compared to the highest D-stat value for each participant, not specific to a particular form of challenging behaviour. This suggests that challenging behaviour does not fall into two distinct groups; either pain related or influenced by environmental factors although if environmental function is not identified, it may be more likely that pain influences the challenging behaviour. These findings indicate a complex relationship between the influential factors of challenging behaviour whereby biological and social factors simultaneously influence the behaviour.

This relationship could take several forms. Firstly, pain could act as a setting event, as suggested by Carr and Smith (1995). According to this theory, the presence of pain may increase the motivation for a consequence previously associated with a behaviour, thus increasing the likelihood of the behaviour (see section 1.3.8). Previous research has supported this theory by demonstrating that children are more likely to engage in more episodes of challenging behaviour and for greater lengths of time on days when they are unwell. The same findings have been shown with women with disability during times of menses (Carr et al., 2003), when they are more likely to be experiencing stomach pains.

Another explanation could be that pain may initially introduce challenging behaviour into the behavioural repertoire, through processes such as the gate control, but then once the behaviour has been introduced, environmental factors can shape it. The mean age of the children in this study was 10 (see section 5.3.1). Therefore, it is likely that the challenging behaviour exhibited by these children would have been exposed to environmental influences. It would be useful for future research to recruit younger children who have only recently began to engage in challenging behaviour and investigate whether it is easier to categorise the challenging behaviour as being influenced only by pain in that case.

In this study, the positive association between behavioural indicators of pain and self injury and aggression was demonstrated, which is in line with previous research discussed throughout this thesis and the findings in chapters 3 and 4. The results also show that there is a significant proportion of challenging behaviour, for which an environmental function cannot be attributed. In these cases, there is potentially an increased likelihood that pain influences challenging behaviour.

These findings provide strong evidence to support the idea that challenging behaviour may be treated successfully using health and behavioural interventions and therefore, demonstrate the importance of effective early identification and treatment of challenging behaviour. It is essential that parents understand the importance of seeking advice regarding challenging behaviour as soon as it occurs. A standard protocol is required to identify the causes of challenging behaviour in order to match appropriate treatments to behaviour. It is especially important for health conditions, which may cause pain and discomfort, to be identified and treated as early as possible before environmental influences take effect.

6.5.1 Limitations and implications for future research

There are a number of potential limitations associated with the present study. For example, participants were separated into the challenging behaviour groups depending on teacher reports. Previous research has shown that teacher and parental reports of challenging behaviour often differ between respondents and different environmental settings (Greenbaum et al., 1994; Grietens et al., 2004; Satake et al., 2003; Verhulst & Akkerhuis, 1989). In the present study, teacher reports of challenging behaviour were used when assessing the difference in pain scores between individuals with and without challenging behaviour. Therefore, the results may have been different if the children had been categorised as having challenging behaviour based on parent report or researcher observations.

Caution is advised when interpreting the findings from the experimental functional analysis assessments for a number of reasons. Firstly, the results vary depending on how challenging behaviour is grouped. For instance, one participant engaged in self injury, aggression and destruction of property during the functional analysis assessments. Self injury and aggression were associated with attention maintenance and destruction of property was associated with social escape and access to tangibles. However, when all challenging behaviours were combined to form an 'overall challenging behaviour' variable, this was then associated with only attention maintenance. This example demonstrates the importance of examining each topography of challenging behaviour independently as different behaviours may have different environmental functions. This has implications for designing interventions for challenging behaviour as it would be ineffective to respond to a collection of challenging behaviours with one intervention (Day et al., 1994; Derby et al., 1994). In this study, different topographies of challenging behaviour were grouped to form the three composite variables; self injury, aggression and destruction of property. Therefore, the proportion of challenging behaviour reported to be associated with each environmental function may be different to if the topographies of self injury, aggression and destruction of property were investigated separately.

Secondly, using this method of categorising challenging behaviour is problematic because only a very short occurrence of challenging behaviour is sufficient to have a significant impact on the results. For example, if two independent episodes of self injury occurred in two separate low attention conditions but at no other time, this would be sufficient to categorise that behaviour as ‘attention maintained’, even if the behaviour only lasted for a second in each instance. Using such a sensitive method of categorising challenging behaviour increases the likelihood of over inclusion in the ‘functional’ group. Also, using this method means that it is not possible to determine the strength of the association other than knowing the proportion of experimental conditions the behaviour occurred in compared to the number of control conditions. So, behaviour would be described as having the same strength of association if it occurred for 100% of the test condition time as it occurred for just 2% of the test condition time.

A third issue is that the functional analysis conditions used in the present study are only able to identify behaviour that was associated with demand escape or attention maintenance. During data analysis, it appeared that, on some occasions, more challenging behaviour was observed in the control conditions than the test conditions. This lead to a categorisation of function associated with social escape or access to tangibles. Given the functional analysis assessments were not designed to systematically examine these functions, caution is advised in these instances. In the same sense, the design used in this study did not allow the automatic reinforcement of behaviour hypothesis to be investigated. Many of the instances of behaviours being defined as ‘attention maintained’ or ‘access to tangibles’ in this study could be confounded by behaviour that is, in fact, automatically reinforced. It is possible that the lack of sensory stimulation experienced throughout the ‘low attention’ experimental condition leads to higher rates of challenging behaviour rather than the motivation for social attention or access to tangibles.

The most common function identified for destructive behaviour was demand-escape. However, this result could be confounded by the fact that there was more opportunity for destructive acts to occur during the demand condition as the child was given access to materials whereas in the other conditions, the researcher attempted to limit the participant’s access to materials.

Despite these limitations in methodology, this study was valuable as it was the first to incorporate the assessment of both pain-related and environmental influences on challenging behaviour. Future research should build on these findings to provide a more detailed description of the characteristics of individuals engaging in functional and non-functional challenging behaviour. Given the methodological limitations discussed, it would be beneficial for future studies to employ the use of more extensive experimental functional assessments, which investigate the possible influence of a broader range of environmental factors. It would also be useful to conduct functional assessments on different days, to increase the likelihood of observing behaviour influenced by setting events such as pain. Finally, when analysing the results from experimental functional assessments, it would be helpful to separate the forms and topographies of challenging behaviour to improve the validity of these assessments for attributing environmental function to challenging behaviour.

CHAPTER 7

THE TEMPORAL RELATIONSHIP BETWEEN BEHAVIOURAL INDICATORS OF PAIN AND SELF INJURY

7.1 Preface

The previous chapters have demonstrated a potential association between pain and challenging behaviour in people with intellectual disability. However, the findings reported thus far do not allow a causal model between pain and challenging behaviour to be developed. In this chapter, the temporal association between behavioural indicators of pain and self injury will be assessed. The aim is to identify temporal patterns between pain behaviour and self-injury which might indicate whether pain causes, or is the result of, self injury.

7.2 Introduction

Estimated rates of challenging behaviour, such as self injury, aggression and destruction of property, in people with intellectual disability, range from 10% to 45% (Emerson et al., 2001b; Grey et al., 2010; Lowe et al., 2007). There are numerous negative personal, carer and financial implications associated with challenging behaviour (Emerson et al., 2001b) and these testify to the importance of developing effective interventions to prevent or reduce its occurrence. There is emerging evidence that once established, challenging behaviours persist over time (Chadwick et al., 2005; Taylor et al., 2011; Totsika et al., 2008) and can be moulded through the process of social reinforcement to develop into more severe challenging behaviour (Carr & Durand, 1985; Iwata et al., 1982/1994, Oliver et al., 2005). The increase in both severity and prevalence with age, highlights the importance of early intervention, which, to be successful, requires an understanding of the underlying causes of the behaviour.

There are several descriptions of how challenging behaviours first develop. Operant learning theory postulates that stereotyped, accidental or pain related behaviours are inadvertently shaped through social reinforcement into challenging behaviours (Carr & Durand, 1985; Guess & Carr, 1991; Iwata et al., 1982/1994) (see section 1.3.1). Neuro-transmitter theories suggest that disrupted functioning of the opioid (see section 1.3.6.2), dopaminergic or serotinergic (see section 1.3.5) systems influence the development of self injury (Breese et al., 1995; Cataldo & Harris, 1982; Sandman, 1990/1991; Rojahn et al., 2008; Symons, 2011; Winchel & Stanley, 1991). Psychiatric models propose that challenging behaviour may be a depressive equivalent in people with intellectual disability and should be considered a symptom of depression (Marston et al., 1997) (see section 1.2.6). Finally, recent models suggest that underlying health problems and pain cause challenging behaviour, predominantly self injury (Carr & Owen-DeSchryver, 2007; Kennedy et al., 2007; Symons et al., 2009a) (see chapter 2).

These theories of challenging behaviour are not mutually exclusive. For example, challenging behaviour introduced into the behavioural repertoire because of internal influences could then be subject to operant conditioning and develop into more severe challenging behaviour (Tunnicliffe & Oliver, 2011). Alternatively, pain could act as a ‘setting event’ altering the pre-existing relationship between challenging behaviour and environmental influences (Carr & Smith, 1995; Carr et al., 2003) (see section 1.3.8). Therefore, the identification of environmental function associated with challenging behaviour does not eliminate the possibility of pain being present or it influencing challenging behaviour. However, if environmental function is not identified, this suggests that internal factors are likely to be driving the challenging behaviour. The lack of environmental function associated with challenging behaviour could therefore be used in clinical and research practice to identify cases of challenging behaviour likely to be influenced by pain or discomfort.

Understanding the influence of pain is important as targeted interventions addressing pain in addition to environmental factors can yield successful outcomes when treating challenging behaviour (Carr et al., 2003; Carr & Blakely-Smith, 2006; Luzzani, et al., 2003). Furthermore, greater awareness of the role of pain in challenging behaviour may prompt investigation, diagnosis and treatment of underlying health conditions when challenging behaviour is present.

Researchers have proposed different theoretical accounts of how pain may be related to challenging behaviour. Firstly, self injury could act as a 'coping' mechanism for pain by modulating the transmission of nerve impulses travelling from pain receptors (gate control theory) or by influencing the pain perception threshold (Melzack & Wall 1965/1982; Woolf & Slater, 2000) (see section 1.3.6.4). Secondly, the endogenous opioid response to pain caused by self injury may be rewarding and therefore be reinforced through operant learning (Sandman, 1990/1991; Rojahn et al., 2008) (see sections 1.3.6.2 & 1.3.1) and finally, insensitivity to pain from self-injury could allow behaviours to be shaped through social reinforcement without pain acting as a deterrent (see section 1.3.6.3).

There are a number of methodological approaches used to evaluate the relationship between pain and challenging behaviour. Studies employing informant based observational pain measures report a positive association between pain and challenging behaviour (Kennedy et al., 2007: Symons & Danov, 2005; Symons et al., 2009a). Other studies infer pain from the presence of health problems, identifying significant increase in challenging behaviour on days when pain ratings were higher and there were observable (e.g. diarrhoea) or measurable (e.g. high temperature) indications of ill health (Carr & Owen-DeSchryver, 2007). Increases in challenging behaviour were also observed in women during menses when abdominal pain was likely (Carr et al., 2003). Caution needs to be applied when using this methodology because the validity of using health problems to infer pain is questionable. However, one advantage of using this method, as opposed to investigating the association between behavioural indicators of pain and challenging behaviour, is that it is more theoretically viable to infer causation between the two variables. As it is theoretically implausible that challenging behaviour could cause menstrual pain or diarrhoea, it can be tentatively assumed that the discomfort associated with the health problem causes the challenging behaviour.

An alternative method for investigating the causal relationship between pain and challenging behaviour is to record the occurrence of behavioural indicators of pain and episodes of challenging behaviour on a second by second basis. This would provide a description of how pain and challenging behaviour are temporally related. This methodology has been used by Courtemanche et al., (2012) who investigated the temporal sequence of self injurious behaviour and pain indicators in four individuals with intellectual disability. Participants were selected who typically engaged in self injury at least once a day, on most days and who had engaged in self injury for at least the last twelve months. Results revealed no predictable or standard pattern of association between the two variables. Some behavioural indicators of pain were observed before the self injury, some after and in some cases, no expressions of pain were observed. This identified a novel research methodology that provides methods of data collection and analysis for future studies. It may be beneficial to expand this methodology with more careful selection of participants. If participant inclusion was based on prior information indicating the influence of pain on the challenging behaviour, it may be possible to attain a clearer description of the relationship between these two factors. This is purpose of the study reported here. The aim of this study is to describe the temporal association between behavioural indicators of pain and episodes of self injury in carefully selected individuals who engage in challenging behaviour that is likely to be associated with pain.

7.3 <u>Method</u>

7.3.1 <u>Participants</u>

Participants were recruited as part of a larger study (see section 5.3.1). Three participants were selected based on the following criteria, which indicate the influence of pain on their self injury;

1. Pain identified as a function for at least one topography of self injury on the Questionnaire About Behavior Function (QABF)
2. Completion of experimental functional analysis where no environmental function was identified for any topography of challenging behaviour
3. Behavioural indicators of pain were displayed for a minimum of three minutes (3.75% of the time) during experimental functional analysis.
4. Engaged in five or more episodes of self injury across two or more experimental functional analysis conditions (see section 6.3.3.3.2)

The table below (table 7.1) describes the participants selected for this study and states the forms of challenging behaviour each of the participants engaged in during the experimental functional assessments.

Table 7.1. Participants' expressive and receptive language, gross motor skills and personal daily living skills age equivalent scores (months) are derived from the Vineland Adaptive Behavior Scale. Challenging behaviour exhibited during the experimental functional analysis is also noted.

Participant	**Syndrome**	**Gender**	**Age (years)**	**Expressive language** [a]	**Receptive language** [a]	**Gross motor skills** [a]	**Personal daily living skills** [a]	**Challenging behaviour during experimental functional analysis**
1	Fragile X with Autism[b]	Male	13	8	11	31	27	Aggression and self injury
2	Tuberous Sclerosis Complex with ASD[b]	Female	5	19	16	17	24	Destruction of property, aggression and self injury
3	Tuberous Sclerosis Complex with Autism[b]	Female	6	3	9	28	10	Destruction of property, aggression and self injury

[a] Age equivalent scores (months) derived from the Vineland Adaptive Behaviour Scale
[b] Scored past the cut-off for Autism Spectrum Disorder (ASD) or Autism on the Social Communication Questionnaire

7.3.2 Procedure

Questionnaire and interview data were collected as part of a larger study (see section 6.3.2). Participants completed experimental functional assessments with the primary researcher at the children's school (see section 6.3.3.3.2). These assessments were recorded by the secondary researcher who did not interact with the child during the assessment.

7.3.3 Measures

7.3.3.1 Experimental functional analysis

The environmental setting is systematically altered to test the effect of social/environmental factors on challenging behaviour (see section 6.3.3.3.2).

7.3.3.2 Questionnaire About Behavioral Function (QABF: Paclawskyi, Matson, Rush, Smalls & Vollmer, 2000) (appendix V)

Questions about behavioural function (QABF) is an informant based questionnaire comprising of 25 questions related to the possible function of the behaviour (e.g. social attention, escape from demands, physical discomfort etc). Each item is rated on a four point Likert scale, from 'never' (0) to 'often' (3). For example, questions 25 asks *'Does he/she seem to be saying "give me that (toy item, food item)" when engaging in the behaviour?'* Internal consistency for the total scale ranges from 0.79 to 0.99 and test-retest reliability is reported to be good with correlations between raters ranging from 0.64 to 1.0 (Paclawskyj et al., 2000).

7.3.3.3 Direct behavioural codes of challenging behaviour.

Challenging behaviour was recorded during the experimental functional assessments. Each form and topography of challenging behaviour was coded separately. For a list of the codes (and therefore definition of self injury to be applied to this study), see appendix M.

7.3.4 Data analysis

To ensure there were sufficient data for analysis, participants were only included if they engaged in at least five episodes of self injury across a minimum of two separate experimental conditions.

Video footage collected during experimental functional assessments was analysed using Obswin software (Martin, Oliver & Hall, 2001), which records the onset and offset of operationally defined behaviours. This allows for the calculation of both frequency and duration of behaviours. Inter-rater reliability data were collected for 25% of the video footage for each participant. Cohen's Kappa (Cohen, 1960) was used to evaluate agreement between observers. The Kappa value represented the proportion of agreements between variables recorded by each researcher within five second windows. A Kappa value of .6 or higher was achieved for all target variables for all participants, which reflects good to excellent inter-rater agreement (Fleiss, 1981).

A D-Stat value was calculated for the experimental functional analysis assessments. A D-stat value of .50 or above indicated 'functional' challenging behaviour (see section 6.3.4).

7.3.4.1 The temporal association between behavioural indicators of pain and challenging behaviour

Yule's Q indicates the magnitude of the association between pairings of variables, in this case behavioural indicators of pain and challenging behaviours. The behaviours of interest were operationally defined prior to data coding. The FLACC pain measure was used as a guide for selecting the behaviours that would be coded. A time-based sequential lag analysis was conducted, which meant Yule's Q values were calculated based on twelve, ten second windows prior to and after the occurrence of challenging behaviour. Therefore, a series of Yule's Q values were created, indicating the strength of the association between the behavioural indicators of pain and challenging behaviour every ten seconds starting from 120 seconds before the onset of self injury until 120 seconds after the onset of self injury. The lag analysis was restricted to the next occurrence of self injury, meaning observations of behavioural indicators of pain would only be included in the Yule's Q analysis up until the next occurrence of challenging behaviour, even if they occurred within the 120 second time frame. Lag zero indicates the onset of self injury. Each time lag indicates a ten second window. Therefore lag -1 is ten seconds prior to the occurrence of challenging behaviour and lag +1 indicates ten seconds after the onset of challenging behaviour and so on.

Episodes of self injury, aggression and destruction of property were observed during the experimental functional assessments. However, after inspection of the data, it was evident that self injury was the main form of challenging behaviour exhibited by the participants selected for this study. Also, the pain theories of challenging behaviour only have limited value when applied to other forms of challenging behaviour. For these reasons, only self injury will be assessed in this study.

Following on from the findings discussed in chapter 5, all behavioural indicators of pain were combined to form one 'total pain indicator' variable as this was found to be the most valid method for measuring pain using direct observations (see section 5.4.3).

7.3.5 Participant selection

7.3.5.1 QABF data

Inclusion criteria stated that participants had to engage in self injury that was associated with the function of pain, by scoring at least '1' on four out of the five items on the pain subscale on the QABF. Table 7.2 shows the challenging behaviour reported by parents during the QABF interviews. Behaviours identified as being associated with pain are noted.

Table 7.2. Behaviours reported by the parents during the QABF interviews with a note detailing whether they were endorsed as being associated with pain.

Participant	Behaviour 1	Behaviour 2	Behaviour 3	Behaviour 4
1	Self injury- Biting hands, knuckles and wrist [a]	Aggression- pulling hair[a]	Aggression- hitting [a]	na
2	Self injury- chewing hand [a]	Self injury- hitting head [a]	Aggression- pinching [a]	Aggression- pulling hair [a]
3	Self injury- biting	Self injury- hitting face [a]	na	na

[a] Behaviour defined as being associated with pain according to the QABF.

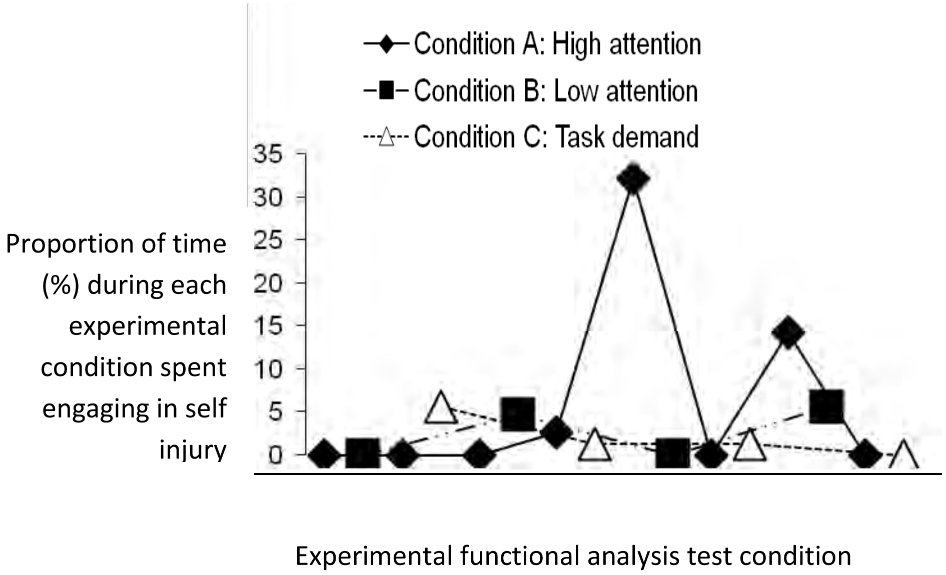
7.3.5.2 Function of challenging behaviour; experimental functional analysis results

The second criterion for selection was that challenging behaviour could not be associated with environmental function identifiable with the use of experimental functional assessments. Moderate function is defined as a D-stat value of .5 or higher (see section 6.3.4). The graphs below (figure 7.1) show the percentage of time during each experimental condition that the participant engaged in self injury and the associated D-stat values for self injury.

Figure 7.1. Graphs depicting the proportion of time, during each five minute experimental functional analysis condition, that was spent engaging in self injury.

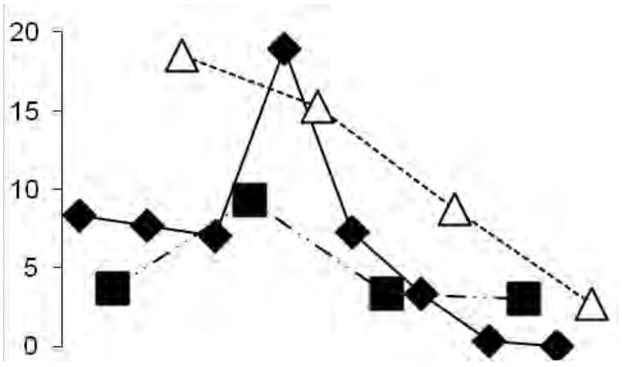
Participant One

Attention maintained self injury D-stat: 0.06
Demand escape self injury D-stat: 0.16



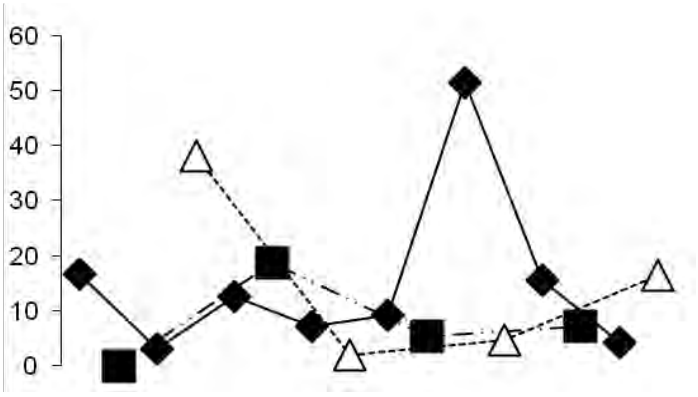
Participant Two

Attention maintained self injury D-stat: -0.09
Demand escape self injury D-stat: 0.44



Participant Three

Attention maintained self injury D-stat: -0.25
Demand escape self injury D-stat: -0.06



The D-stats calculated from these experimental functional analysis results were all below .50, meaning no environmental function was associated with self injury. D-stat calculations for aggression, destruction of property and overall challenging behaviour for these participants were also below .50 (see appendix X).

7.3.5.3 Behavioural indicators of pain

The third criterion for inclusion in this study was that the individual had to engage in behavioural indicators of pain for a minimum of 3.75% of the time during the experimental functional analysis, which equates to three minutes. Table 7.3 shows the number of discrete incidents of pain indicators and the proportion of time each participant engaged in the behaviours during the experimental functional analysis.

Table 7.3. Number of distinct episodes of, and proportion of experimental functional assessment spent engaging in; negative affect, self sooth, non-relaxed body movement and non relaxed leg movement. 'Pain' is a composite measure of these other behaviours. .

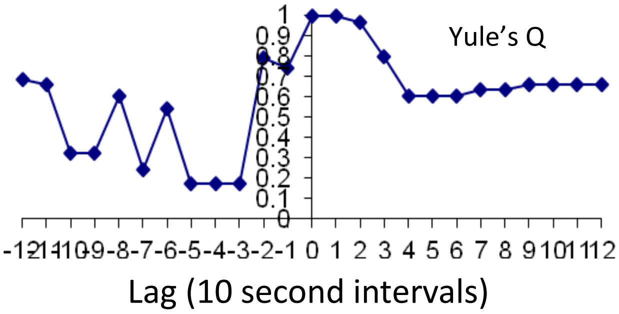
Participant	Negative affect (% time)	Self Sooth (% time)	Non relaxed body movement (% time)	Non relaxed leg movements (% time)	Pain (% time)
1	18 (3.45)	0	0	3 (.31)	21 (3.76)
2	18 (3.45)	0	0	2 (.62)	20 (4.07)
3	160 (21.82)	0	46 (3.50)	0	179 (24.21)

7.4 Results

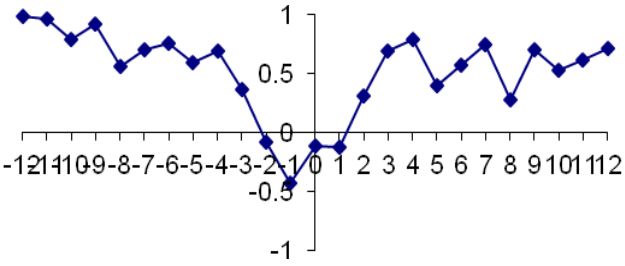
A time-based sequential lag analysis was conducted to examine how the magnitude of the association between behavioural indicators of pain and the occurrence of self injury changed over time. Figure 7.2 shows Yule's Q values over twelve ten-second time lags before and after the onset of self injury.

Figure 7.2. Graphs depicting Yule's Q value in the 120 seconds prior to and after the onset of self injury.

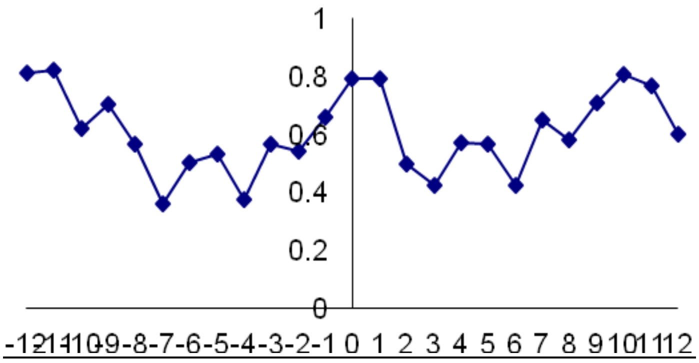
Participant one



Participant two



Participant three[1]



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[1] Following the research visit with participant 3, the researcher received a letter from the Mother stating that the child's challenging behaviour had been associated with constipation. Medical intervention to remedy the constipation resulted in a dramatic improvement to the child's challenging behaviour. This was significant enough to allow the family to go on holiday, which is something they had never been able to do previously because of the severity of the child's challenging behaviour.

For participant one, the association between behavioural indicators of pain and self injury increased steeply in the thirty seconds prior to the onset of self injury and peaked at lag zero, which is the time self injury began. The association between behavioural indicators of pain and self injury then decreased steadily for the forty seconds after the onset of self injury before reaching a plateau. The pattern was similar in participant three. The association between behavioural indicators of pain and self injury increased for the forty seconds before the onset of self injury and then peaked in the ten seconds immediately following the onset of self injury. The association then reduced for the following twenty seconds.

The pattern of association was distinctly different in participant two. There was a steady decrease in association between behavioural indicators of pain and self injury from forty seconds prior to self injury until ten seconds prior to self injury. The association between the two factors then increased steadily until around forty seconds after the onset of self injury.

7.5 Discussion

In this study, participants were selected based on criteria which signified an influence of pain on their challenging behaviour. This was indicated by absence of an environmental function associated with challenging behaviour identified through experimental functional analysis, pain function identified for the challenging behaviour on the QABF and a high proportion of time spent engaging in behavioural indicators of pain.

The results from participant one and three followed the same general pattern. There was a clear increase in the association between behavioural indicators of pain and challenging behaviour before the onset of self injury. This suggests that pain plays a causal role in the expression on self injury in these individuals. This finding supports the pain theory of challenging behaviour that suggests self injury may, in some cases, act as a ‘coping strategy’ for dealing with pain (Peebles & Price, 2012) (see section 1.3.6.4). According to this theory, in a typically functioning nervous system, the presentation of noxious stimuli below detection levels increases the threshold at which pain is perceived. It is proposed that some individuals with intellectual disability have altered nervous systems, which means more intense stimulation, such as self injury, is required to evoke the same response.

The pattern of results for participant two was strikingly different to participants one and three. The association between behavioural indicators of pain and self injury reduced in the lead up to the self injury. Therefore, the pain theory is inappropriate for explaining self injury in this individual. After closer inspection of the experimental functional assessment graphs, it appears likely that self injury in participant two was associated with a 'demand escape' function. However, this was not reflected in the D-Stat result (.42). This reveals a limitation to using pre-selected values to define 'function' as in some cases, this method results in the possible misclassification of challenging behaviour. It is possible that some challenging behaviour associated with an environmental function gets classed as non-functional, as with participant two and the opposite could be true for other individuals. This shows the value of using alternative strategies to complement the experimental functional assessments such as more fine grained investigation of the temporal association between variables or the use of informant reports using measures such as the QABF.

The results of this study also show the value of using comprehensive assessment protocols which investigates all possible factors associated with challenging behaviour. Just within these three individuals, omitting the use of either experimental functional analyses or observations of behavioural indicators of pain would have meant that key influences on self injury would not have been identified. Therefore a multi-faceted assessment protocol is required to identify all the influential variables on challenging behaviour, which can then be used to design an individualised intervention strategy.

It is worth noting that the results of this study could be explained by an alternative theory to the pain theory of challenging behaviour. It is possible that the behaviours coded as ‘behavioural indicators of pain’ could serve as proto-imperative behaviours, which are defined as “gestures intended to make another person do something for one’s benefit” (Sarria, Gomes & Tamarit, 1996). According to this theory, behaviours such as negative vocalisations, negative facial expressions or fidgety movement could be viewed as communicative behaviours that are functionally equivalent to the challenging behaviour expressed during the experimental functional assessments. Previous research findings support this notion. Petty, Allen and Oliver (2009) demonstrated that crying (including negative vocalisations) and going limp were functionally equivalent to self injurious behaviour in a group of five children with intellectual disability. Therefore, in this study, the temporal association between proto-imperative and challenging behaviours may not be explained by pain, but by the motivation to evoke a rewarding outcome previously associated with the behaviour.

CHAPTER 8

GENERAL DISCUSSION

8.1 Introduction

In the introduction to this thesis, an overview of different theoretical accounts of challenging behaviour in people with intellectual disability was presented. The most plausible and comprehensive explanation for challenging behaviour was derived from combining a number of separate theories, as each theory alone was potentially insufficient for explaining all challenging behaviour (see section 1.3.8). It is quite possible that each theory discussed in the introductory chapter is correct but only applicable to certain cases of challenging behaviour. Also, in some cases, the operant learning processes and biological/internal factors, such as pain and discomfort, could interact to initially cause, and then influence, challenging behaviour in people with intellectual disability. For example, Carr and McDowell (1980) demonstrated how a child's self injurious scratching, initially introduced in response to contact dermatitis, was subsequently influenced by social reinforcement. In this example, scratching may have been introduced into the behavioural repertoire to moderate the pain or discomfort associated with contact dermatitis (Melzack & Wall 1965/1982; Woolf & Slater 2000) then, the scratching became associated with environmental factors through operant learning. In a separate study, O'Reilly (1997) showed that the self injury of a 26 month old girl with intellectual disability was associated with sensory escape (escape from noise) and demand escape, but only during times when she had otitis media. In this example, pain caused by the otitis media appeared to influence the established behaviour-reinforcement relationship between self injury and the removal of demands (Carr & Smith, 1995; Carr & Blakeley-Smith, 2006; Carr et al., 2003). The importance of understanding the role of pain in challenging behaviour was highlighted by consideration of the possible implications for assessment and treatment.

In addition to this model building based on theoretical accounts there is empirical evidence that a significant proportion of challenging behaviour cannot be ascribed an environmental function (Iwata et al., 1994; Toogood & Timlin, 1996). Consequently, the operant learning theory is potentially unable to explain these behaviours and interventions based on operant learning principles are likely be ineffective for treating these cases of challenging behaviour and pain as a cause of challenging behaviour in these cases should be considered, alongside other possibilities such as the presence of depression. It follows therefore that the investigation of the influence of pain on challenging behaviour may lead to the development of successful intervention strategies that treat cases of challenging behaviour via pain relief. Investigating the association between pain and challenging behaviour may also have implications for identifying pain in non-verbal individuals. Pain is inherently difficult to recognise in people with intellectual disability with limited communication skills because it is a personal and subjective experience. Without the ability to self report, pain cannot be directly assessed. The aim of this thesis was to address some of these methodological difficulties by employing a range of techniques that indirectly assess pain. The association between pain and challenging behaviour in people with intellectual disability could then be examined.

8.2 Main findings from each chapter

In chapter 2, the results of the first systematic literature review on the association between pain and challenging behaviour in people with intellectual disability were presented. Examination of the methodology used in these studies identified a number of possible strategies for appraising the association between pain and challenging behaviour. In several studies, poor health was used to infer the presence of pain or discomfort. Observations of behavioural indicators of pain were also used to identify pain. The methodological approaches identified by reviewing previous literature were used to guide the design of the empirical studies described within this thesis.

In chapter 3, the use of a rating scale to identify behavioural indicators of gastro-oesophageal distress was employed. This study was the first to use the Gastro-oesophageal Distress Questionnaire (GDQ) as a proxy measure of pain for two groups at high risk of gastro-oesophageal reflux. This study further established the understanding of the association between pain and challenging behaviour and demonstrated the value of using health problems to identify probable cases of pain. Rates of self injury, aggression, destruction of property and stereotyped behaviours were significantly higher in individuals with probable gastro-oesophageal pain compared to those without. Rates of challenging behaviour were also significantly associated with low affect and hyperactivity, signifying that these behaviours may be useful, although less well defined, indicators of pain in people with intellectual disability. In chapter 3, the possibility of using difference in activity level to distinguish between challenging behaviour associated with pain and challenging behaviour associated with depression was also discussed.

In chapter 4, individuals with Tuberous Sclerosis Complex (TSC) were recruited as a high-risk group for probable pain. TSC is characterised by benign growths in all organs of the body, which are likely to cause pain and discomfort. Compared to people with Down syndrome, a low-risk group for pain, rates of self injury, aggression and destruction of property were high in those with TSC, although this between group difference was not statistically significant. Expanding on the findings from the study described in chapter 3, a number of person characteristics were shown to be associated with challenging behaviour. As in chapter 3, low mood and hyperactivity were associated with challenging behaviour. Other person characteristics were also shown to be associated with challenging behaviour including socialisation and communication difficulties, and repetitive and compulsive behaviours. Importantly, more behavioural indicators of pain were also expressed by people with TSC who engaged in challenging behaviour compared to those with no challenging behaviour.

In chapter 5, the value of using observations of behavioural indicators of pain was appraised. The psychometric properties of the FLACC, NCCPC-R and direct observation codes were assessed in a group of people with intellectual disability during times not previously defined as being associated with pain. Good inter-rater reliability of the teacher and researcher FLACC ratings was reported. Also, good concurrent validity was reported between the teacher FLACC and teacher NCCPC-R total scores as well as the researcher FLACC and the ‘overall pain indicator’ code (used during live observations), which combined all behavioural indicators of pain into one composite variable. Based on these findings, the teacher FLACC and NCCPC-R, researcher FLACC and ‘overall pain indicator’ direct observational code, were selected for use in studies discussed in chapters 6 and 7 and may prove to be useful in other research studies.

Chapter 6 added to previous findings reported in chapter 4 by identifying higher levels of behavioural indicators of pain in people with challenging behaviour than those who do not show challenging behaviour. In chapter 6, a protocol was trialled that assessed operant functions associated with challenging behaviour and also the potential influence of pain. Experimental functional analysis was used to assess the function of challenging behaviour and pain was assessed using teacher ratings on the FLACC and NCCPC-R. 26% of self injury, 47% of aggression and 44% destruction of property was associated with an environmental function. This was the first study to assess behavioural indicators of pain in relation to environmental causes of challenging behaviour. It was shown that, there was a weak to moderate negative correlation between many behavioural indicators of pain and the environmental influence on challenging behaviour. This indicates that, if an environmental function for challenging behaviour cannot be ascribed, an association between the challenging behaviour and pain is more likely. Therefore, this highlights the potential use of 'undifferentiated function' to signify cases of challenging behaviour that are more likely to be linked to underlying pain.

In chapter 7, detailed coding of behavioural indicators of pain and episodes of self injury were used to describe the temporal association between pain and self injury. The participants in this study were carefully selected based on a number of criteria, which were indicative of an influence of pain on challenging behaviour. The participants in this study engaged in challenging behaviour which was not associated with an environmental function according to experimental functional assessment results. The behaviour was associated with pain according the Questionnaire About Behavior Function and each individual engaged in behavioural indicators of pain for at least three minutes of the experimental functional assessment. In two of the three case studies described in this chapter, there was an increase in behavioural indicators of pain immediately prior to the onset of self injury. This suggests that, in these individuals, pain may cause self injury.

8.3 Plan for discussion

In this general discussion, the implications of the main findings presented in previous chapters will be described and theoretical, clinical and research implications discussed. Methodological limitations and considerations for future research will be presented.

8.4 Theoretical implications

Throughout this thesis, results have suggested that the association between behavioural correlates of pain and self injury may be higher than the association between pain and other forms of challenging behaviour such as aggression and destruction of property. For example, in chapter 3, pain associated with gastro-oesophageal distress was shown to be associated with self injury in people with Cri du Chat syndrome but not with aggression or destruction of properly. In chapter 6, a seemingly greater proportion of self injury compared to aggression and destruction of property was associated with 'undifferentiated' function, although the statistical significance of this difference was not open to testing due to the low numbers. Therefore, it appeared that a smaller proportion of self injury could be explained by environmental factors, and by assumption, this may indicate the role of internal/biological factors such as pain instead. The gate control theory suggests that acts of self injury could moderate the perception of pain (Melzack & Wall 1965/1982; Woolf & Slater 2000) (see section 1.3.6.4). The opioid theory suggests that opioids, released in response to self injury, may be associated with a pleasant and rewarding sensory response (Rojahn et al. 2008; Sandman, 1990/1991) (see section 1.3.6.4). These theories are only useful for explaining the association between pain and self injury but may not apply equally to other forms of challenging behaviour. A subset of self injury, associated with pain in the ways described here, could explain the higher proportion of self injury, compared to other forms of challenging behaviour, for which an environmental function cannot be ascribed.

A second theory of how pain could influence self injury was also discussed in the introductory chapter. The ‘reduced pain perception’ theory asserts that reduced or altered pain perception would allow self injury to be moulded through operant learning processes without the typical ‘costs’ of pain (Peebles & Price, 2012; Rojahn et al. 2008; Sandman, 1990/1991) (see section 1.3.6.3). The findings reported throughout this thesis suggest that this theory cannot be applied to all people who engage in self injury. In chapters 3, 4, 6 and 7, a positive association between behavioural correlates of pain and self injury was demonstrated, suggesting that increased, not decreased, levels of pain are associated with self injury in these individuals. Also, in chapter 7, for two people who engaged in self injury, behavioural indicators of pain were shown to increase before an episode of self injury, indicating that experiencing pain could cause an episode of self injury in these two children.

Another theory presented in the introductory chapter to this thesis, and then explored in chapter 3, is that challenging behaviour could be a ‘symptom’ of depression in people with intellectual disability (Marston et al., 1997; Meins, 1995) (see section 1.2.6). This theory is based on the observation that low mood is often associated with challenging behaviour. However, the findings of this thesis indicate that it is entirely possible that both low mood and challenging behaviour are explained by the common third variable of pain. In chapters 3 and 4, hyperactivity and low mood were associated with challenging behaviour (see sections 3.4.3 & 4.4.3). Hyperactivity has been identified as a possible behavioural indicator of pain (Luzzani et al., 2003; Breau et al., 2004, Merkel et al., 1997). Therefore, the occurrence of hyperactivity, in conjunction with low mood and challenging behaviour, indicates that all three factors may be associated with pain. If challenging behaviour was indicative of depression alone, it would not be expected to be associated with other correlates of pain in this way.

8.5 Clinical implications

The possible association between pain and challenging behaviour has been demonstrated in each empirical study of this thesis using different strategies identified in the systematic literature review in chapter 2. This potential association shows that it is imperative for the assessment of pain to be incorporated into any standard protocol for investigating causes of challenging behaviour. It is also apparent that the influence of pain should be considered when designing intervention strategies to treat challenging behaviour in people with intellectual disability. Failure to identify and treat underlying pain is unethical and the less acceptable error as this would lead to unnecessary suffering. It would also jeopardise the success of other methods for treating challenging behaviour, such as behavioural interventions based on operant learning processes, as the underlying cause of the challenging behaviour would not be addressed. Although errors of commission are arguably more justifiable, it is also important to consider the risk of over-diagnosing pain. This could result in unnecessary intrusive and distressing medical examinations or procedures and the possibility of adverse side effects to wrongly prescribed medication.

The literature review in chapter 2 and findings from the experimental functional assessments in chapter 6 highlight the importance of assessing and treating pain as a cause of challenging behaviour in the context of environmental factors (Carr et al., 2003; Carr & Blakeley-Smith, 2006). Even if challenging behaviour is initially caused by pain, it is likely to then be subject to operant learning processes (Carr & McDowell, 1980). Pain could also impact on some challenging behaviour by acting as an ‘establishing operator’, which increases the likelihood of challenging behaviour being expressed (Carr & Smith, 1995; Carr & Blakeley-Smith, 2006; Carr et al., 2003) (see section 1.3.8). Therefore, to ensure the success of intervention strategies for all cases of challenging behaviour, it is essential to address the influence of both environmental factors and pain.

Aside from the obvious benefits of identifying unrecognised pain, early recognition of pain in people with intellectual disability has important implications. As already mentioned, even if challenging behaviour is initially caused by pain, it can then be moulded through operant learning processes (Carr & McDowell, 1980). This can result in challenging behaviour persisting over time (Chadwick et al., 2004; Taylor et al., 2011; Totsika et al., 2008) and also means, even if pain is relieved, the challenging behaviour may continue. This testifies to the importance of early identification and treatment of pain in people with intellectual disabilities as this may prevent challenging behaviours acquiring an environmental function. The findings reported in this thesis may bring researchers and clinicians one step closer to being able to achieve this. Several person characteristics (hyperactivity and negative affect) were shown to be potentially associated with pain in people with intellectual disability (see sections 3.4.3 & 4.4.3). These have potential for use as risk markers for pain in this population, prompting further investigation. Also, the work in this thesis demonstrated the value of using observation measures (chapters 4, 6 & 7), which enable contemporary rating of the presence of behavioural indicators of pain. The items listed on these measures could be used as a guide to inform clinicians of behavioural correlates of pain in people with intellectual disability.

8.6 Methodological/research implications

8.6.1 Methods for investigating the association between pain and challenging behaviour

In the introductory chapter, the difficulty of identifying pain in people with intellectual disability in the absence of self-report was discussed (see section 1.1). This problem makes it extremely difficult to investigate the association between pain and challenging behaviour in people with intellectual disability. In an effort to overcome this problem, a number of different methodological approaches for identifying pain in non-verbal individuals have been employed. It has been demonstrated that behavioural indicators of health problems, such as gastro-oesophageal reflux disorder, can be used to successfully identify a group of people likely to be experiencing pain (see chapter 3). Similarly, questionnaire measures, which require observers to rate the presence or frequency of behavioural indicators of pain can also be used effectively (see chapters 4, 5 & 6). For example, the NCCPC-R (Breau et al., 2004) and FLACC (Merkel et al., 1997) were used in chapter 6 to evaluate the pain levels of individuals with and without environmentally functional challenging behaviour. Also, in chapter 4, the NCCPC-R was used to show that individuals who engaged in self injury, aggression and destruction of property, had significantly higher rates of behavioural indicators of pain compared to people without challenging behaviour. Direct observation and real-time coding of behavioural indicators of pain also provide a second-by-second account of the expression of pain behaviours (Courtemanche et al., 2012). This method was used to illustrate the temporal relationship between indicators of pain and episodes of self injury (see section 7.4). Finally, in chapter 4, individuals with TSC represented a group at high risk of pain. Each of these methods allowed the association between pain and challenging behaviour to be successfully investigated in a population of people with intellectual disability. Therefore, the empirical studies in this thesis have highlighted the effective use of a number of different approaches for recognising pain and investigating its relationship with challenging behaviour. These strategies can now be adopted for future research meaning the association between pain and challenging behaviour can be more thoroughly investigated. One area for future research could be to refine the method for identifying which cases of challenging behaviour are likely to be influenced by pain, and therefore, which cases of challenging behaviour would benefit from medical interventions aimed at pain relief. The predictive validity of the method of case identification could then be evaluated in an intervention trial.

8.6.2 Discrepancy between observers

In chapter 5, results indicated that there were differences between parent and teacher ratings of behavioural indicators of pain (see section 5.4.1). This finding is in line with previous research, which has demonstrated consistent and systematic differences between parent and teacher ratings of challenging behaviour (Greenbaum et al., 1994; Grietens, et al., 2004; Satake et al., 2003; Verhulst & Akkerhuis, 1989). This has a number of clinical and research implications. Firstly, it is important to ensure that ratings of pain and challenging behaviour are taken from a number of different sources (e.g. teachers and parents) to provide a comprehensive description of an individual's behaviour. Secondly, it important to be cautious when generalising findings from one setting to another.

8.6.3 The confounding effect of intellectual disability

In chapters 3 and 4, the confounding effect of level of intellectual disability on the association between pain behaviours and challenging behaviour was considered (see sections 3.4.4 & 4.4.4). In chapter 4, when level of intellectual disability was removed as a variable in the analysis, the association between behavioural indicators of pain and challenging behaviour was no longer significant. However, it was also pointed out that in some cases, level of intellectual disability is correlated with other clinical features of genetic syndromes (de Vries et al., 2007), including health problems that cause pain. Therefore, by removing intellectual disability as a possibly confounding variable, the impact of pain may also be removed. Given the importance of identifying pain in people with intellectual disability, not only so that treatment can be provided to alleviate discomfort, but also because of the implications for challenging behaviour interventions, it is arguable that over-diagnosis of pain is justifiable compared to the alternative, which is discounting a possible influence of pain. How the issue of intellectual disability is dealt with in this context is an important consideration for future research.

8.6.4 Alternative interpretations of the presence of behavioural indicators of pain

In chapter 7, an increase in behavioural indicators of pain was observed before the onset of self injury, which was interpreted as pain causing self injury (see section 7.4). However, ‘negative affect’, ‘negative vocalisations’, ‘non-relaxed body’ and ‘non-relaxed legs’, which are the behaviours that make up the composite ‘pain indicators’ variable, could also be interpreted as communicative behaviours associated with an environmental function instead of being indicators of pain. It could be that these precursor behaviours serve an environmental ‘function’ such as causing the initiation of social attention or the removal of a demanding task (Petty et al., 2009). Given that the pattern of results predicted by both of these interpretations is the same, it is not possible to determine which account is accurate for which cases of challenging behaviour. It is arguable however, that the negative impact of interpreting these behaviours as communicative as opposed to indicators of pain, is far greater than incorrectly inferring pain. If these behaviours are associated with environmental function, results from experimental functional analysis could be used to design an intervention procedure, which is likely to be successful. The resources deployed through investigating the possibility of pain would be the only cost associated with inaccurately interpreting the behaviours as being indicative of pain. Alternatively, if the behaviours were pain related, and this was overlooked, pain and discomfort could go untreated. Also, interventions for challenging behaviour that only address the associated environmental influences would be likely to be unsuccessful because they fail to address the underlying cause of the behaviour. Therefore, in clinical and research practice, the risk of over inclusion is justifiable compared to the risk of erroneous omission of the impact of pain on challenging behaviour.

Also, within this thesis, low mood has been used as an indicator of pain. When viewed in this way, the association between low mood and challenging behaviour can be used to support the assertion that pain is associated with challenging behaviour. However, some researchers may suggest that challenging behaviour may be a symptom of depression, which would explain the association between low mood and challenging behaviour. There are important implications and considerations associated with each of these interpretations. If depression is present, then treatment with anti-depressants may improve challenging behaviour and it would be unethical and ineffective to not consider this diagnosis during treatment. However, it is equally unethical to not treat pain, and failure to consider the influence of pain during treatment, if it is present, would also result in ineffective treatment strategies for challenging behaviour. It may also be the case that depression is the result of chronic pain. In this instance, it would be inappropriate to treat the depression without addressing the underlying cause of the depression. It is important that future research focuses on how to determine which of these interpretations is accurate for each case of challenging behaviour when low mood is present. Until this can be achieved, it is important that clinicians investigate the possibility of both depression and pain when challenging behaviour presents.

8.6.5 Methodological limitations

8.6.5.1 Recruitment limitations

A number of methodological limitations were identified throughout the thesis, which are important to note for future research. Firstly, in chapter 4, participants with TSC were recruited from a parent/carer support group (see section 4.3.1). Membership of such organisations may be biased towards carers of people with more challenging behaviours (Karloff & Friesen 1991). This may have produced an inflated estimate of rates of challenging behaviour within this population. This bias can result from a number of other recruitment strategies, including recruitment through treatment centres, residential care homes, questionnaire surveys and more. The possibility of recruitment bias, and the impact this has on research findings, should be considered in future research.

8.6.5.2 Experimental functional analysis limitations

In chapter 6, experimental functional assessments were used to assess environmental influences on challenging behaviour. Caution is advised when interpreting findings from experimental functional assessments for a number of reasons. Firstly, the outcome of the assessment (whether behaviour is deemed functional and, if so, which function it is associated with), depends largely on the operational definition of the challenging behaviour, i.e. whether different forms of challenging behaviour are grouped together (e.g. self injury, aggression and destruction of property) or if different topographies are grouped together (e.g. self scratching, self pinching and pulling one’s own hair grouped into ‘self injury’) (Day et al., 1994; Derby et al. 1994). This means that the function identified for behaviours could be misleading. Secondly, using experimental functional assessments may lead to certain functions being missed if the assessment protocol does not test the specific antecedent associated with challenging behaviour for that individual (Richards, 2012) or if the control condition does not provide a suitable ‘demand free’ comparison condition. Furthermore, using a D-stat value as was employed in chapter 6, does not distinguish between challenging behaviour that occurs for a very brief amount of time compared to challenging behaviour that lasts for a much greater proportion of time during the experimental condition. Therefore, a D-stat value is not sufficient for fully describing the behaviour. It is also necessary to look at the proportion of time spent engaging in each behaviour when making a judgement about function.

In summary, taking all of these limitations of experimental functional analysis into consideration, it is advised that the results from these assessments should be complemented by additional information. For example, the number of episodes or the proportion of time spent engaging in the challenging behaviour might be used as filters before using the data to indicate the strength of the association between an environmental influence and challenging behaviour. This method could also be used to identify cases where function may have been erroneously attributed and further investigation is warranted. More detailed analysis could also improve the validity of the findings from experimental functional assessments. For example, real-time coding of challenging behaviours, antecedent events and consequences could be effective for identifying function associated with challenging behaviour that typical experimental functional assessment protocols do not identify.

8.7 Closing statement

The aim of this thesis was to employ a range of methodological approaches and strategies to investigate the association between pain and challenging behaviour in people with intellectual disability. Because pain is a personal and subjective experience, the first methodological hurdle was to find a way to identify pain in this population, when self report is not possible. In this thesis, a number of successful approaches were employed that measured pain indirectly through the use of behavioural correlates of pain. Through use of these strategies, a robust and reliable association between pain and challenging behaviour was established. The findings discussed throughout this thesis testify to the importance of identifying and treating pain in people with intellectual disability. It is imperative that the possibility of pain is investigated when challenging behaviour is present, not only to alleviate unnecessary discomfort but also because of the potential impact on challenging behaviour.

As yet, there is no standard protocol for assessing challenging behaviour which incorporates the assessment of pain. The findings reported in this thesis can be used to inform future assessment protocols. The combined use of experimental functional assessment, assessment of other factors such as depression, and observation of behavioural indicators of pain could be the initial standard protocol for assessing challenging behaviour. A number of outcomes could be used to indicate the potential involvement of pain: failure to identify an environmental function, observation of a high number of behavioural indicators of pain, observation of person characteristics associated with pain such as hyperactivity and low mood, the increase of pain indicators prior to the onset of challenging behaviour, a history of health problems and the diagnosis of a genetic syndrome associated with painful health problems, could all be used to signify the possible influence of pain. Identification of pain alongside other factors such as mental health problems and environmental factors influencing challenging behaviour could lead to the successful implementation of effective intervention strategies to treat challenging behaviour, which in turn, could significantly improve the well being of a large proportion of people with intellectual disabilities and their carers. The next step is to test the use of the proposed protocol for identifying cases of challenging behaviour that are influenced by pain and treat the challenging behaviour with physical interventions targeted at pain relief. The results of such investigations could help to refine the criteria used to identify probable pain-related challenging behaviour. This could lead to the implementation of more evidence based, targeted, and therefore successful, interventions for reducing challenging behaviour in the future.

REFERENCES

Adams, D., & Oliver, C. (2011). The expression and assessment of emotions and internal states in individuals with severe or profound intellectual disabilities. *Clinical Psychology Review, 31,* 293-306.

Affleck, G., Tennen, H., Urrows, S., & Higgins, P. (1992). Neuroticism and the pain-mood relation in rheumatoid arthritis: insights from a prospective daily study. *Journal of Consulting and Clinical Psychology*, 60, 119-126.

Ahlsen, G., Gillberg, I.C., Lindblom, R., & Gillberg, C. (1994). Tuberous Sclerosis in Western Sweden. A population study of cases with early childhood onset. *Archives of Neurology, 51*, 76-81.

Allen, D.G., Lowe, K., Moore, K., & Brophy, S. (2007). Predictors, costs and characteristics of out of area placement for people with intellectual disability and challenging behaviour. *Journal of Intellectual Disability Research*, *51,* 409–416.

Allerton, L., A., Welch, V., & Emerson, E. (2011). Health inequalities experienced by children and young people with intellectual disabilities: a review of literature from the United Kingdom. *Journal of Intellectual Disability Research, 15(4),* 269-78.

Aman, M.G., & Singh, N.N. (1986). *Aberrant Behavior Checklist manual.* East Aurora, NY: Slossen.

Anderson, L.T., & Ernst, M. (1994). Self-injury in Lesch-Nyhan disease. *Journal of Autism and Developmental Disorders, 24,* 67–81.

Applegate, H., Matson, J.L., & Cherry, K. (1999). An evaluation of functional variables affecting severe problem behaviours in adults with mental retardation by using the Questionnaire about Behavioural Function Scale (QABF). *Research in Developmental Disabilities, 20(3),* 229-237.

Arron, K., Oliver, C., Berg, K., Moss, J., & Burbidge, C. (2011). Prevalence and phenomenology of self-injurious and aggressive behaviour in genetic syndromes. *Journal of Intellectual Disability Research*, 55, 109-120.

Baghdadli, A., Pascal, C., Grisi, S., & Aussilloux, C. (2003). Risk factors for self-injurious behaviours among 222 young children with autistic disorders. *Journal of Intellectual Disability Research, 47,* 622-627.

Baker, P., Piven, J., & Sato, Y. (1998). Autism and Tuberous Sclerosis Complex: Prevalence and Clinical Features. *Journal of Autism and Developmental Disorders, 28,* 279- 285.

Baldini, A. (2004). DiGeorge syndrome: an update. *Current Opinion in Cardiology, 19*, 201-204.

Barr, O., Gilgunn, J., Kane, T., & Moore, G. (1999). Health screening for people with learning disabilities by a community learning disability service in Northern Ireland. *Journal of Advanced Nursing, 29*, 1482-1491.

Beck, B. (1987). Psycho-social assessment of 36 de Lange patients. *Journal of Mental Deficiency Research, 31,* 251–257.

Berg, K., Arron, K., Burbidge, C., Moss, J., & Oliver, C. (2007). Carer reported contemporary health problems in people with severe learning disability and genetic syndromes. *Journal of Policy and Practice in Intellectual Disabilities*, *4,* 120-128.

Berument, S.K., Rutter, M., Lord, C., Pickles, A., & Bailey, A. (1999) Autism screening questionnaire: diagnostic validity. *British Journal of Psychiatry,175,* 444-451.

Berney, T.P., Ireland, M., & Burn, J. (1999). The behavioural phenotype of Cornelia de Lange syndrome. *Archives of Diseases in Childhood*, *81*, 333 *-336.*

Bhuiyan, Z., Klein, M., Hammond, P., van Haeringen, A., Mannens, M., Berckelaer-Onnes, I., & Hennekam, R. (2006). Genotype-phenotype correlations of 39 patients with Cornelia De Lange syndrome: the Dutch experience. *Medical Genetics, 43,* 568-575.

Bodfish, J.W., Crawford, T.W., Powell, S.B., Parker, D.E., Golden, R.N., & Lewis, M.H. (1995). Compulsions in adults with mental retardation: prevalence, phenomenology, and comorbidity with stereotypy and self-injury. *American Journal of Mental Retardation, 100(2),* 183–92.

Böhmer, C.J.M., Taminiau, J.A.J.M., Klinkenberg-Knol, E.C., & Meuwissen, S.G.M. (2001). The prevalence of constipation in institutionalized people with intellectual disability. *Journal of Intellectual Disability Research, 45*, 212–218.

Bohmer, C.J., Niezen-de Boer, M.C., Klinkenberg-Knol, E.C., Deville, W.L., Nadorp, J.H., & Meuwissen, S.G. (1999). The prevalence of gastroesophageal reflux in institutionalized intellectually disabled individuals. *American Journal Gastroenterology, 94,* 804–810.

Bosch, J., Van Dyke, D.C., Smith, S.M., & Poulton, S. (1997). Role of medical conditions in the exacerbation of self-injurious behavior: an Exploratory Study. *Mental Retardation*, *35(2),* 124-130.

Bouras, N., Dykens, E.M., & Smith, A.,C.,M. (2003). Distinctiveness and correlates of maladaptive behaviour in children and adolescents with Smith–Magenis syndrome. *Journal of Intellectual Disability Research, 42*, 481–489.

Branford, D., Bhaumik, S., & Duncan, F. (1998). Epilepsy in adults with learning disabilities. *Seizure, 7,* 473-477.

Breau, L.M., Camfield, C., McGrath, P.J., Rosmus, C., & Finley, G.A. (2001). Measuring pain accurately in children with cognitive impairments: Refinement of a caregiver scale. *Journal of Pediatrics, 138,* 721-727.

Breau, L.M., Camfield, C., Symons, F.J., Bodfish, J.W., Mackay, A., Finley, G.A., & McGrath, P.J. (2003). Relation between pain and self-injurious behavior in nonverbal children with severe cognitive impairments. *The Journal of pediatrics, 142,* 498-503.

Breau, L.M., McGrath, P.J., Camfield, C.S., & Finley, G.A. (2002) Psychometric properties of the non-communicating children's pain checklist-revised. *Pain, 99,* 349-357.

Breau, L.M., McGrath, P.J., Camfield, C., Rosmus, C., & Finley, G.A. (2000). Preliminary validation of an observational checklist for persons with cognitive impairments and inability to communicate verbally. *Developmental Medicine and Child Neurology, 42,* 609-616.

Breau, L., McGrath, P, Finley, A., & Camfield, C. (2004). *The Non-Communicating Children's Pain Checklist- Revised.* Nova Scotia, Canada.

Breese, G.R., Criswell, H.E., Duncan, G.E., Moy, S.S., Johnson, K.B., Wong, D.R., & Mueller, R.A. (1995). Model for reduced brain dopamine in Lesch–Nyhan Syndrome and the mentally retarded: neurobiology of neonatal-6-hydroxydopamine lesioned rats. *Mental Retardation and Developmental Disabilities Research Reviews, 1,* 111–119.

Breese, G.R., Knapp, D.J., Criswell, H.E., Moy, S.S., Papadeas, S.T., & Blake B.L. (2005). The neonate-6-hydroxydopamine-lesioned rat: a model for clinical neuroscience and neurobiological principles. *Brain Research Reviews, 48,* 57-73.

Bruno, E., Rossi, N., Thüer, O., Córdoba, R., & Alday, L. (2003). Cardiovascular findings, and clinical course, in patients with Williams syndrome. *Cardiology in the Young, 13,* 532-536.

Bull, M.J., Fitzgerald, J.F., Heifetz, S.A., & Brei, T.J. (1993). Gastrointestinal abnormalities: A significant cause of difficulties and failure to thrive in Brachman–de Lange syndrome. *American Journal of Medical Genetics, 47,* 1029–1034.

Burbidge, C., & Oliver, C. (2008). *The Activity Questionnaire. Manual for administration and score interpretation.* University of Birmingham, Birmingham.

Burbidge, C., Oliver, C., Moss, J., Arron, K., Berg, K., Hill, L., Trusler, K., Furniss, F., & Woodcock, K. A. (2010). The association between repetitive behaviours, impulsivity and hyperactivity in people with intellectual disability. *Journal of Intellectual Disability Research*, *54*, 1078-1092.

Carr, E. (1977). The Motivation of Self-Injurious Behavior: A Review of Some Hypotheses. *Psychological Bulletin, 84,* 800-816.

Carr, E.G., & Durand, V.M. (1985). Reducing behavior problems through functional communication training. *Journal of Applied Behaviour Analysis, 18(2),*111–126.

Carr, E., & Blakeley-Smith, A. (2006). Classroom intervention for illness-related problem behavior in children with developmental disabilities. *Behaviour Modifications, 30(6),* 901-924.

Carr, E., & McDowell, J.J. (1980) Social control of self-injurious behavior of Organic etiology. *Behavior Therapy, 11,* 402-409.

Carr, E.G., & Owen-DeSchryver, J.S. (2007). Physical illness, pain, and problem behavior in minimally verbal people with developmental disabilities. *Journal of Autism and Developmental Disorders, 37(3),* 413-424

Carr, E.G., & Smith, C.E. (1995). Biological setting events for self-injury. *Mental Retardation and Developmental Disabilities Research Reviews, 1*, 94-98.

Carr, E.G., Smith, C.E., Giacin, T.A., Whelan, B.M., & Pancari, J. (2003). Menstrual discomfort as a biological setting event for severe problem behavior: Assessment and intervention. *American Journal on Mental Retardation, 108*, 117-133.

Carvill, S. (2001). Review: Sensory impairments, intellectual disability and psychiatry. *Journal of Intellectual Disability Research, 45*, 467-83.

Cataldo, M.F., & Harris, J. (1982). The biological basis for self-injury in the mentally retarded. *Analysis and Intervention in Developmental Disabilities, 2,* 21-39.

Chadwick, O., Kusel, Y., Cuddy, M., & Taylor, E. (2005). Psychiatric diagnoses and behaviour problems from childhood to early adolescence in young people with severe intellectual disabilities. *Psychological Medicine, 35,* 751-760.

Chandler, K., & Dahlquist, C. (2006). *Functional assessment: Strategies to prevent and remediate challenging behavior in school settings.* New Jersey: Pearson Education, Inc.

Chapman, R., & Hesketh, L. (2000). Behavioural phenotype of individuals with Down Syndrome. *Mental Retardation and Developmental Disabilities Research Reviews, 6,* 84-95.

Choi, J.U., Bae, E.J., Noh, C., Yoon, Y., & Hwang, Y.S. (1995). Cardiac rhabdomyoma in childhood tuberous sclerosis. *Cardiology in the Young, 5,* 166-171.

Christie, R., Bay, C., Kaufman, I.A., Bakay, B., Borden, M., & Nyhan, W.L. (1982). Lesch-Nyhan disease: clinical experience with nineteen patients. *Developmental Medical Child Neurology, 24,* 293–306.

Clarke, D.J., & Dykens, E.M. (1997). Correlates of maladaptive behaviour in individuals with 5p-(cri du chat) syndrome. *Developmental Medicine and Child Neurology, 39 (11),* 752-756.

Clarke, A., Hancock, E., Kingswood, C., & Osborne, J. (1994). Nephrology Dialysis. *Transplantation, 14*, 988-991.

Clayton, J., & Smith, M.M. (1993). Clinical research on Angelman syndrome in the United Kingdom: observations on 82 affected individuals. *American Journal of Medical Genetics, 46,* 12-15.

Cliff, N. (1993). Dominance Statistic- Ordinal Analyses to Answer Ordinal Questions. *Psychological Bulletin, 114,* 494-509.

Cohen, J. (1960). A coefficient of agreement for nominal scales. *Educational and Psychological Measurement, 20*, 37–46.

Collacott, R., Cooper, A., Branford, D., & McGrother, C. (1998). Behaviour Phenotype for Down's syndrome. *The British Journal of Psychiatry, 172,* 85-89.

Cooper, S., Smiley, E., Allan, L., Jackson, A., Finlayson, J., Mantry, D., & Morrison, J. (2009). Adults with intellectual disabilities: prevalence, incidence and remission of self injurious behaviour, and related factors. *Journal of Intellectual Disability Research, 53,* 200-216.

Courtemanche, A., Schroeder, S., Sheldon, J., Sherman, J., & Fowler, A. (2012). Observing signs of pain in relation to self-injurious behaviour among individuals with intellectual and developmental disabilities. *Journal of Intellectual Disability Research, 65(5),* 501-515.

Craig, K.D., Hadjistavropoulos, H.D., Grunau, R.V.E., & Whitfield, M.F. (1994). A Comparison of 2 Measures of Facial Activity During Pain in the Newborn Child. *Journal of Pediatric Psychology, 19,* 305-318.

Crino, P.B., Nathanson, K.L., & Henske, E.P. (2006). The Tuberous Sclerosis Complex. *New England Journal of Medicine, 355,* 1345–56.

Critchley, M., & Earl, C. (1932). Tuberous Sclerosis and allied conditions. *Brain, 55,* 311-46.

Crocker, A., Mercier, C., Lachapelle, Y., Brunet, A., Morin, D., & Roy, M. (2006). Prevalence and types of aggressive behaviour among adults with intellectual disabilities. *Journal of Intellectual Disability Research, 50*, 652-661.

Curatolo, P., Bombardieri, R., & Jozwiak, S. (2008). Tuberous Sclerosis. *The Lancet, 372,* 657-68.

Davies, L. (In review). Pain, mood and challenging behaviour in intellectual disability. Unpublished Clinical Psychology Doctoral Thesis, University of Birmingham, Birmingham.

Davis, J.P., Judd, J.K., & Herman, H. (1997). Depression in adults with intellectual disability. *Australian and New Zealand Journal of Psychiatry*, 31, 232-242.

Day, H., Horner, R., & O'Neil, R. (1994). Multiple functions of problem behaviours; assessment and Intervention. *Journal of Applied Behavior Analysis, 27*, 279- 289.

Deb, S., Thomas, M., & Bright, C. (2001). Mental disorder in adults with intellectual disability. 2: The rate of behavior disorders among a community-based population aged between 16 and 64 years. *Journal of Intellectual Disability Research, 45(6),* 506-514.

Derby, K., Wacker, D., Peck, S., Sasso, G., DeRaad, A., Berg, W., Asmus, J., & Ulrich, S. (1994). Functional analysis of separate topographies of aberrant behaviour, *Journal of Applied Behavior Analysis, 27*, 267-278.

de Vries, P., Hunt, A., & Bolton, P. (2007). The psychopathologies of children and adolescents with Tuberous Sclerosis Complex, A postal survey of UK families. *European Child Adolescent Psychiatry, 16,* 16-24.

de Vries, P.J., Gardiner, J., & Bolton, P.F. (2009). Neuropsychological attention deficits in tuberous sclerosis complex (TSC). *American Journal of Medical Genetics, 149,* 387–395.

de Vries, P. J. (2010). Neurodevelopmental, Psychiatric and Cognitive Aspects of Tuberous Sclerosis Complex, in *Tuberous Sclerosis Complex: Genes, Clinical Features and Therapeutics* (eds D. J. Kwiatkowski, V. H. Whittemore and E. A. Thiele), Wiley-VCH Verlag GmbH & Co. KGaA, Weinheim, Germany.

de Vries, P.J., & Prather, P.A. (2007). The Tuberous Sclerosis Complex. *The New England Journal of Medicine, 365,* 92-94.

Dinca, O., Paul, M., & Spencer, N.J. (2005). Systematic review of randomized controlled trials of atypical antipsychotics and selective serotonin reuptake inhibitors for behavioural problems associated with pervasive developmental disorders. *Journal of Psychopharmacology, 19,* 521–532.

Donovan, J. (2002). Learning disability nurses' experiences of being with clients who may be in pain. *Journal of Advanced Nursing, 38,* 458–466.

Durand, V.M., & Carr, E.G. (1991). Functional communication training to reduce challenging behavior: Maintenance and application in new settings. *Journal of Applied Behavior Analysis, 24,* 251–264.

Dykens, E.M. (1995). Measuring behavioral phenotypes: provocations from the 'new genetics'. *American Journal on Mental Retardation, 99,* 522-32.

Dykens, E., Shah, B., Sagun, J., Beck, T., & King, B. (2002). Maladaptive behaviour in children and adolescents with Down's syndrome. *Journal of Intellectual Disability Research, 46,* 484-492.

Easterbrook, P.J., Berlin, J.A., Gopalan, R., & Matthews, D.R. (1991). Publication bias in clinical research. *Lancet, 337(8746),* 867-72.

Eaves, L.C., Wingert, H.D., Ho, H.H., & Mickelson, E.C.R. (2006). Screening for autism spectrum disorders with the social communication questionnaire. *Developmental and Behavioral Pediatrics, 27,* 95–103.

Ekman, P., & Friesen, W. V. (1978). Facial action coding system: A technique for the measurement of facial movement. Palo Alto, CA: Consulting Psychologists Press.

Embregts, P.J.C.M., Didden, R., Schreuder, N., Huitink, C., & Van Nieuwenhuijzen, M. (2009). Aggressive behavior in individuals with moderate to borderline intellectual disabilities who live in a residential facility: An evaluation of functional variables. *Research in Developmental Disabilities, 30*, 682–688.

Emerson, E. (2003). Mothers of children and adolescents with intellectual disability: social and economic situation, mental health status, and the self-assessed social and psychological impact of the child's difficulties. *Journal of Intellectual Disability Research, 47,* 385–399.

Emerson, E. (1995). *Challenging behaviour: analysis and intervention in people with learning disabilities.* Cambridge Press, Cambridge.

Emerson E., Cullen C., & Hatton C. (1996). *Residential Provision for People with Learning Disabilities: Summary Report.* Hester Adrian Research Centre, Manchester University, Manchester.

Emerson, E., & Hatton, C. (2007a). The contribution of socio-economic position to the health inequalities faced by children and adolescents with intellectual disabilities in Britain. *American Journal on Mental Retardation, 112(2),* 140-50.

Emerson, E., & Hatton, C. (2007b). Poverty, socio-economic position, social capital and the health of children and adolescents with intellectual disabilities in Britain: a replication. *Journal of Intellectual Disability Research, 51(11),* 866-74.

Emerson, E., Kiernan, C., Alborz, A., Reeves, D., Mason, H., Swarbrick, R., Mason, L., & Hatton, C. (2001a). Predicting the persistence of severe self-injurious behavior. *Research in Developmental Disabilities, 22,* 67-75.

Emerson, E., Kiernan, K., Alborz, A., Reeves, D., Mason, H., Swarbrick, R., Mason, L., & Hatton, C. (2001b). The prevalence of challenging behaviors: a total population study, *Research in Developmental Disabilities, 22*, 77-93.

Epstein, M.H., Cullinan, D., & Gadow, K.D. (1986). Teacher ratings of hyperactivity in learning disabled, emotionally disturbed, and mentally retarded children. *Journal of Special Education, 20,* 219-229.

Eysenck, S.B.G., Easting, G., & Pearson, P.R. (1984). Age norms for impulsiveness, venturesomeness and empathy in children. *Personality and Individual Differences, 5,* 315-321.

Fanurik, D., Koh, J.L., & Harrison, R.D., Conrad, T.M., & Tomerlin, C. (1998). Pain assessment in children with cognitive impairment: an exploration of self report skills. Clinical Nursing Research, *7*, 103–19.

Fass, R., & Dickman, R. (2006). Non-cardiac chest pain: an update. *Neurogastroenterology & Motility, 18,* 408–417.

Feldman, S.I., Downey, G., & Schaffer-Neitz, R. (1999). Pain, negative mood, and perceived support in chronic pain patients: a daily diary study of people with reflex sympathetic dystrophy syndrome. *Journal of Consultant Clinical Psychology, 67,* 776-85.

Feldt, K.S. (2000). The checklist of nonverbal pain indicators (CNPI). *Pain Management Nursing, 1,* 13-21.

Fidler, D. (2005). The emerging Down syndrome Behavioural Phenotype in Early Childhood; Implication for Practice. *Infants and young children, 18*, 86-103.

Fisher, W.W., Bowman, L.G., Thompson, R.H., & Contrucci, S.A. (1998). Reductions in self injury produced by transcutaneous electrical nerve stimulation. *Journal of Applied Behavior Analysis, 31,* 493-496.

Fleiss J. (1981) *Statistical Methods for Rates and Proportions*, 2nd edn. John Wiley, New York.

Fox, R.A., & Wade, E.J. (1998). Attention deficit hyperactivity disorder among adults with severe and profound mental retardation. *Research in Developmental Disabilities, 19*, 275-280.

Gath, A., & Gumley, D. (1986). Behaviour problems in retarded children with special reference to Down's syndrome. *The British Journal of Psychiatry, 149,* 156-161.

Grey, I., Pollard, J., McClean, B., MacAuley, N., & Hastings, R. (2010). Prevalence of psychiatric diagnoses and challenging behaviors in a community-based population of adults with intellectual disability. *Journal of Mental Health Research in Intellectual Disabilities, 3,* 210-222.

Greenbaum, P.E., Dedrick, R.F., Prange, M.E., & Friedman, R.M. (1994). Parent, teacher and child ratings of problem behaviours of youngsters with serious emotional disturbances. *Psychological Assessment, 6(2),* 141-148.

Grietens, H., Onghena, P., Prinzie, P., Gadeyne, E., van Assche, V.A., Ghesquiere, P., & Hellinckx, W. (2004). Comparison of mothers', fathers and teachers reports on problem behaviour in 5- to 6-year old children. *Journal of Psychopathology and Behavioural Assessment, 26(2),* 137-146.

Guess, D., & Carr, E. (1991). Emergence and maintenance of stereotypy and self-injury. *American Journal on Mental Retardation, 96(3),* 299-319.

Gutierrez, G., Smalley, S., & Tanguay, P. (1998). Autism in Tuberous Sclerosis Complex. *Journal of Autism and Developmental Disorders, 28 ,* 97-103.

Hagopian, L.P., Fisher, W.W., Sullivan, M.T., Acquisto, J., & LeBlanc, L.A. (1998). Effectiveness of functional communication training with and without extinction and punishment: a summary of 21 inpatient cases. *Journal of Applied Behavior Analysis, 31(2),* 211–235.

Hall, S., Arron, K., Sloneem, J., & Oliver, C. (2008). Health and sleep problems in Cornelia de Lange Syndrome: A case control study. *Journal of Intellectual Disability Research, 52,* 458-468.

Hammock, R., Levine, W.R., & Schroeder, S.R. (2001). Brief report: effects of clozapine on self-injurious behavior of two risperidone nonresponders with mental retardation. *Journal of Autism and Developmental Disorders, 31,* 109–113.

Handen, B.L., McAuliffe, S., Janosky, J., Feldman, H., & Breaux, A.M. (1994). Classroom behaviour and children with mental retardation- comparisons of children with and without ADHD. *Journal of Abnormal Child Psychology, 22 (3),* 267-280.

Hanley, G.P., Iwate, B.A., & McCord, B.E. (2003). Functional analysis of problem behaviour: a review. *Journal of Applied Behaviour Analysis, 36,* 147-185.

Harding, J.W., Wacker, D.P., Berg, W.K., Lee, J.F., & Dolezal, D. (2009). Conducting functional communication training in home settings: A case study and recommendations for practitioners. *Behavior Analysis in Practice. 2(1),* 21–33.

Hartman, E.C., Gilles, E., McComas, J.J., Danov, S.E., & Symons, F. (2008). Clinical observation of self-injurious behaviour correlated with changes in scalp morphology in a child with congenital hydrocephalus. *Journal of Child Neurology, 23(9),* 1062-1065.

Hayes, S., McGuire, B., O'Neill, M., Oliver, C., & Morrison, T. (2011). Low mood and challenging behaviour in people with severe and profound intellectual disabilities. *Journal of Intellectual Disability Research, 55(2),* 182-189.

Hellings, J.A., & Warnock, J.K. (1994). Self-injurious behavior and serotonin in Prader-Willi Syndrome. *Psychopharmacology Bulletin, 30(2),* 245-250.

Holden, B., & Gitlesen, J.P. (2006). A total population study of challenging behaviour in the county of Hedmark, Norway: Prevalence and risk markers. *Research in Developmental Disabilities, 27,* 456-465.

Hollins, S., Attard, M., van Fraunhofer, N., McGuigan, S.,M., & Sedgwick, P. (1998). Mortality in people with learning disability: risks causes, and death certification findings in London. *Developmental Medicine and Child Neurology, 40,* 50-56.

Hopkins, W.G. (1997). New view of statistics. Retrieved September 2nd 2012 from http://www.sportsci.org/resource/stats/effectmag.html. Cited in; Kotrlik, J.W., & Williams, H.A. (2003). The Incorporation of Effect Size in Information Technology, Learning , and Performance Research. *Infomation Technology, Learning and Performance Journal, 2(1),* 1-7.

Horvath, K., Papadimitriou, J.C., Rabsztyn, A., Drachenberg, C., & Tildon, J.T. (1999). Gastrointestinal abnormalities in children with autistic disorder. *Journal of Pediatrics, 135,* 559-563.

Horvath, K., & Perman, J.A. (2002). Autistic disorder and gastrointestinal disease. *Current Opinion in Pediatrics, 14,* 583-587.

Howells, G. (1986). Are the medical needs of mentally handicapped adults being met? *Journal of the Royal College of General Practitioners,* 36, 449-53.

Hunt, A. (1983). Tuberous sclerosis: a survey of 97 cases. III: family aspects. *Developmental Medicine and Child Neurology, 25,* 353–357.

Hunt, A. (1997). A comparison of the abilities, health and behaviour of 23 people with Tuberous Sclerosis at age 5 and as adults. *Journal of Applied Research in Intellectual Disabilities, 11,* 227-238.

Hyman, P., Oliver, C., & Hall, S. (2002). Self-injurious behavior, self-restraint, and compulsive behaviors in Cornelia de Lange Syndrome. *American Journal of Mental Deficiency*, *107,* 146–54.

Irazábal, M., Marsà, F., García, M., Gutiérrez-Recacha, P., Martorell, A., Salvador-Carulla, L., & Ochoa, S. (2012). Family burden related to clinical and functional variables of people with intellectual disability with and without a mental disorder. *Research in Developmental Disabilities*, *33(3)*, 796-803.

Iwata, B.A., Dorsey, M.F., Slifer, K.J., Bauman, K.E., & Richman, G.S. (1982). Toward a functional analysis of self-injury. *Analysis and Intervention in Developmental Disabilities, 2,* 3-20.

Iwata, B.A., Dorsey, M.F., Slifer, K.J., Bauman, K.E., & Richman, G.S. (1994a). Toward a functional analysis of self-injury. *Journal of Applied Behavior Analysis, 27,* 197–209.

Iwata, B.A., Pace, G.M., Dorsey, M.F., Zarcone, J.R., Vollmer, T.R., Smith, R.G., ... Willis, K.D. (1994b). The functions of self-injurious behavior: An experimental-epidemiological analysis. *Journal of Applied Behaviour Analysis, 27(2),* 215–240.

Iwata, B., Pace, G., Kalsher, M., Cowdery, G., & Cataldo, M. (1990). Experimental analysis and extinction of self-injurious escape behavior. *Journal of Applied Behavior Analysis, 23,* 11-27.

Jansen, D.E.M.C., Krol, B., Groothoff, J.W., & Post, D. (2004). People with intellectual disability and their health problems: a review of comparative studies. *Journal of Intellectual Disability Research, 48,* 93-102.

Joinson, C., O'Callaghan, F., Osborne, J., Martyn, C., Harris, T., & Bolton, P. (2003). Learning disability and epilepsy in an epidemiological sample of individuals with Tuberous Sclerosis Complex. *Psychological Medicine, 33(2),* 335-344.

Kennedy, C.H., Juárez, A.P., Becker, A., Greenslade, K., Harvey, M.T., Sullivan, C., & Tally, B. (2007). Children with severe developmental disabilities and behavioral disorders have increased special healthcare needs. *Developmental Medicine & Child Neurology, 49,* 926–930.

Kennedy, C., & O'Reilly, M.F. (2006). Pain, Health Conditions, and Problem Behavior in People with Developmental Disabilities. In T.Oberlander & F. J. Symons (Eds.), *Pain in Children and Adults with Developmental Disabilities* (pp. 121-135). Paul H Brookes, Baltimore, Md.

Kline, A.D., Krantz, I.D., Sommer, A., Kliewer, M., Jackson, L.G., FitzPatrick, D.R., Levin, A.V., & Selicorni, A. (2007). Cornelia de Lange syndrome: Clinical review, diagnostic and scoring systems, and anticipatory guidance. *American Journal of Medical Genetics Part A, 143,* 1287-1296.

Kopp, C.M.C., Muzykewicz, D.A., Staley, B.A., Thiele, E.A., & Pulsifer, M.B. (2008). Behavior problems in children with Tuberous Sclerosis Complex and parental stress. *Epilepsy & Behavior, 13,* 505-510.

Karloff, N.M., & Friesen, B.J. (1991). Support groups for parents of children with emotional disorders: A comparison of members and non-members. *Commuunity Mental Health Journal, 27,* 265-279.

Kushlick, R., Blunden, G., & Cox, G. (1973). A method of rating behaviour characteristics for use in large scale surveys of mental handicap. *Psychological Medicine, 3(4),* 466-478.

Lalli, J.S., Casey, S., & Kates, K. (1995). Reducing escape behavior and increasing task completion with functional communication training, extinction, and response chaining. *Journal of Applied Behavior Analysis, 28(3),* 261–268.

Laud, R.B., & Matson, J.L. (2006). The relationship between manic symptoms on the DASH-II and YMRS and feeding/mealtime behaviour problems. *Research in Developmental Disabilities, 27,* 559-566.

Leung, A., & Robson, W. (2007). Tuberous Sclerosis Complex: A Review. *Journal of Pediatric Health Care, 21,* 108-114.

Lewis, J.C., Thomas, H.V., Murphy, K.C., & Sampson, J.R. (2004). Genotype and psychological phenotype in Tuberous Sclerosis. *Journal of Medical Genetics, 41*, 203–207.

Lotan, M., Ljunggren, E.A., Johnsen, T.B., Defrin, R., Pick, C.G., & Strand, L.I. (2009). A modified version of the Non-Communicating Children Pain Checklist-Revised, Adapted to adults with intellectual and developmental disabilities: Sensitivity to pain and internal consistency. *Journal of Pain, 10(4),* 398-407.

Lovaas, O.I., Freitag, G., Gold, V.J., & Kassorla, I.C. (1965) Experimental studies in childhood schizophrenia: Analysis of self-destructive behavior. *Journal of Experimental Child Psychology, 2,* 67-84.

Lowe, K., Allen, D., Jones, E., Brophy, S., Moore, K., & James, W. (2007). Challenging behaviours: prevalence and topographies. *Journal of Intellectual Disability Research, 51*, 625–636.

Luckasson, R., Borthwick-Duffy, S., Buntinx, W.H.E., Coulter, D.L., Craig, E.M., Reeve, A., ... Tasse, M.J. (2002). *Mental retardation: Definition, classification, and systems of supports* (10th ed.). Washington, DC: American Association on Mental Retardation.

Luzzani, S., Macchini, L., Valade, A., Milani, D., & Selicorni, A. (2003). Gastroesophageal Reflux and Cornelia de Lange Syndrome: Typical and Atypical Symptoms. *American Journal of Medical Genetics, 119*, 283–287.

Mace, F.C., & Lalli, J.S. (1991). Linking descriptive and experimental analyses in the treatment of bizarre speech. *Journal of Applied Behavior Analysis, 24*, 553-562.

Mace, F.C., Lalli, J.S., & Shea, M.C. (1992). Functional analysis and treatment of self-injury. In J. Luiselli, J. Matson, & N. Singh (Eds.), *Assessment, analysis, andtreatment of self-injury.* (pp. 122-152). New York: Springer-Verlag.

Maes, B., Broekman, T.G., Došen, A., & Nauts, J. (2003). Caregiving burden of families looking after persons with intellectual disability and behavioural or psychiatric problems. *Journal of Intellectual Disability Research, 47,* 447–455.

Malten, K.E. (1981). Thoughts on irritant contact dermatitis. *Contact Dermatitis, 7*, 238–247.

Malviya, S., Voepel-Lewis, T., Merkel, S., & Tait, A.R. (2005). Difficult pain assessment and lack of clinician knowledge are ongoing barriers to effective pain management in children with cognitive impairment. *Acute Pain, 7,* 27-32.

Malviya, S., Voepel-Lewis, T., Burke, C., Merkel, S., & Tait, A.R. (2006). The revised FLACC observational pain tool: improved reliability and validity for pain assessment in children with cognitive impairment. *Pediatric Anesthesia, 16,* 258-265.

Marston, G. M., Perry, D. W., & Roy, A. (1997). Manifestations of depression in people with intellectual disability. *Journal of Intellectual Disability Research, 41*, 476– 480.

Martin, N., Oliver, C., & Hall, S. (2001). *Obswin: Software for the Collection and Analysis of Observational Data*. University of Birmingham, Birmingham UK.

Matson, J., Minshawi, N., Gonzalez, M., & Mayville, S. (2006). The relationship of comorbid problem behaviurs to social skills in persons with profound mental retardation. *Behavior Modification, 30*, 496-506.

Matthews, T., Weston, N., Baxter, H., Felce, D., & Kerr, M. (2008). A general practice-based prevalence study of epilepsy among adults with intellectual disabilities and of its association with psychiatric disorder, behaviour disturbance and carer stress. *Journal of Intellectual Disability Research, 52*, 163-73.

McClintock, K., Hall, S., & Oliver, C. (2003). Risk marker associated with challenging behaviour: a meta-analytic study. *Journal of Intellectual Disability Research, 47,* 405-416.

McGill, P. (1999). Establishing Operations: Implications for the assessment. Treatment and prevention of problem behavior. *Journal of Applied Behavior Analysis, 3,* 393-418.

McGuigan, S.M., Hollins, S., & Attard, M. (1995). Age specific standardised mortality rates in people with learning disability. *Journal of Intellectual Disability Research, 39,* 527-31.

Meins, W. (1995). Symptoms of major depression in mentally retarded adults. *Journal of Intellectual Disability Research, 39,* 41-45.

Melzack, R., & Wall, P.D. (1965). Pain mechanisms: A new theory. *Science, 150,* 171-179.

Merskey, H., & Bogduk, N. (1994). Classification of Chronic Pain. 2nd ed. *Seattle: IASP Press.*

Merkel, S.I., Voepel-Lewis, T., Shayevitz, J.R., & Malviya, S. (1997). The FLACC: a behavioural scale for scoring postoperative pain in young children. *Pediatric Nursing Journal, 23,* 293–7.

Michael, J., & Richardson, A. (2008). Healthcare for all: the independent inquiry into access to healthcare for people with learning disabilities. *Tizard Learning Disability Review, 13(4),* 28-35.

Moss, S., Emerson, E., Kiernan, C., Turner, S., Hatton, C., & Alborz, A. (2000). Psychiatric symptoms in adults with learning disability and challenging behaviour. *British Journal of Psychiatry, 177,* 452-456.

Moss, J., Oliver, C., Arron, K., Burbidge, C., & Berg, K. (2009). The prevalence and phenomenology of repetitive behavior in genetic syndromes. *Journal of Autism and Developmental Disorders, 39,* 572-588.

Nathan, P.W., & Rudge, P. (1974). Testing the gate theory of pain in man. *Journal of Neurology, Neurosurgery and Psychiatry, 37,* 1366-1372.

Nellist, M., Janessen, B., Brook-Carter, P.T., Hesseling-Janssen, A.L., Maheshwar, M.M., Verhoef, S., ... Breuning, M.J. (1993). Identification and characteriszation of the Tuberous Sclerosis gene on chromosome 16. *Cell, 75*, 1306-1315.

Oberlander, T., & Symons, F.J. (2006). *Pain in Children and Adults with Developmental Disabilities*. Paul. H Brookes, Baltimore, Md.

O'Callaghan, F. (1999). Tuberous Sclerosis. Epidemiological research is needed to complement new findings in genetics. *British Medical Journal, 318(7190),* 318- 1019.

O'Callaghan, F., Sheill, A., Osbornem, J., & Martyn, C. (1998). Prevalence of tuberous sclerosis estimated by capture-recapture analysis. *The Lancet, 351(9114)*, 1490-1499.

Oliver, C. (1995). Annotation: self-injurious behaviour in children with learning disabilities: recent advances in assessment and intervention. *Journal of Child Psychology and Psychiatry, 30,* 909-927.

Oliver, C., Arron, K., Powis, L., & Tunnicliffe, P. (2011a). Cornelia de Lange, Cri du Chat and Rubinstein Taybi syndrome. In P.Howlin, T. Charman and M. Ghaziuddin (Eds.) *The SAGE Handbook of Developmental Disorders* (pp 239 – 260). Sage, London.

Oliver, C., Berg, K., Burbidge, C., Arron, K., &. Moss, J. (2011b). Delineation of behavioral phenotypes in genetic syndromes. Comparison of autism spectrum disorder, affect and hyperactivity. *Journal of Autism and Developmental Disorder*, *41,* 1019-1032.

Oliver, C., Hall, S., & Murphy, G. (2005). The early development of self-injurious behaviour: Evaluating the role of social reinforcement. *Journal of Intellectual Disability Research,* 49, 591-599.

Oliver, C., McClintock, K., Hall, S., Smith, M., Dagnan, D., & Stenfert-Kroese, B. (2003a). Assessing the severity of challenging behaviour: Psychometric properties of the challenging behaviour interview. *Journal of Applied Research in Intellectual Disabilities, 16,* 53-61.

Oliver, C., Murphy, G., Crayton, L., & Corbett, J. (1993). Self-injurious behavior in Rett syndrome: Interactions between features of Rett Syndrome and operant conditioning. *Journal of Autism and Developmental Disorders, 23(1),* 91-109.

Oliver, C., Murphy, G., Hall, S., Arron, K., & Leggett, J. (2003b). Phenomenology of self-restraint. *American Journal of Mental Retardation, 108,* 71–81.

Oliver, C., Sloneem, J., Hall, S., & Arron, K. (2009). Self-injurious behaviour in Cornelia de Lange syndrome: 1. Prevalence and phenomenology. *Journal of Intellectual Disability Research, 53,* 575–589.

Oliver, C. & Wilkie, L. (2005). *Gastro-Oesophageal Distress Questionnaire*. University of Birmingham, Birmingham.

O'Reilly, M.F. (1997). Functional analysis of episodic self-injury correlated with recurrent otitis media. J*ournal of Applied Behavior Analysis, 30,* 165-167.

Paclawskyj, T.R., Matson, J.L., Rush, K.S., Smalls, Y., & Vollmer, T.R. (2000). Questions About Behavioral Function (QABF): A behavioral checklist for functional assessment of aberrant behaviour. *Research in Developmental Disabilities, 21*, 223–230.

Palmer, J., & Jenkins, J. (1982). The'Wessex' behaviour rating system for mentally handicapped people: reliability study. *British Journal of Mental Subnormality*, *28*, 88 –96.

Parliamentary and Health Service Ombudsman (2009) Six lives: the provision of public services to people with learning disabilities. Part one: overview and summary investigation reports. Crown Copyright, 2009.

Peebles, K.A., & Price, T.J. (2012). Self-injurious behaviour in intellectual disability syndromes: evidence for aberrant pain signalling as a contributing factor. *Journal of Intellectual Disability Research, 56(5),* 441-452.

Peters-Scheffer, N., Didden, R., & Korzilius, H. (2012). Maternal stress predicted by characteristics of children with autism spectrum disorder and intellectual disability**.** *Research in Autism Spectrum Disorders, 6 (2),* 696-706.

Petty, J., & Oliver, C. (2005). Self injurious behaviour in individuals with intellectual disability. *Current Opinion in Psychiatry, 18*, 484-489.

Petty, J., Allen, D., & Oliver, C. (2009). The relationship between self-injurious, potentially-injurious, repetitive, challenging and pragmatic communicative behaviors in children with severe intellectual disabilities. *American Journal on Intellectual and Developmental Disabilities, 114,* 256-358.

Pellicer, F., Buendia-Roldan, I., & Pallares-Trujillo, V.C. (1998). Self-mutilation in the Lesch-Nyhan syndrome: a corporal consciousness problem? —a new hypothesis. *Medical Hypotheses*, *50 (1)*, 43-47.

Poppes, P., van der Putten, A.J.J., & Vlaskamp, C. (2010). Frequency and severity of challenging behaviour in people with profound intellectual and multiple disabilities. *Research in Developmental Disabilities*, *31(6)*, 1269-1275.

Puri, B.K., Lekh, S.K., Langa, A., Zaman, R., & Singh, I. (1995) Mortality in a hospitalized mentally handicapped population: a 10-year survey. *Journal of Intellectual Disability Research, 39*, 442-46.

Quine, L., & Pahl, J. (1989). *Stress and Coping in Families Caring for a Child with Severe Mental Handicap,* Unpublished doctoral thesis. University of Kent, Canterbury.

Qureshi, H. (1995). Impact on families: young adults with learning disabilities who show challenging behaviour. In: *Research to Practice? Implications of Research on the Challenging Behaviours of People with Learning Disability* (ed. C. Kiernan), pp. 89–115. BILD Publications; Kidderminster.

Rakowski, S.K., Winterkorn, E.B., Paul, E., Steele, D.J.R., Halpern, E.F., & Thiele, E.A. (2006). Renal manifestations of Tuberous Sclerosis Complex: Incidence, prognosis, and predictive factors. *Kidney International, 70,* 1777–1782.

Ramesh, V. (2003). Aspects of tuberous sclerosis complex (TSC) protein function in the brain. *Biochemical Society Transactions, 31(3),* 579-583.

Reiss, S., & Rojahn, J. (1993). Joint occurrence of depression and aggression in children and adults with mental retardation. *Journal of Intellectual Disability Research, 37,* 287–294.

Richards, C. (2012). *Experimental functional analysis of self-injurious behaviour in people with Autism Spectrum Disorder.* Unpublished doctoral thesis, University of Birmingham.

Richards, C., & Oliver, C. (in preparation). Validity of using the Gastro-Oesophageal Distress Questionnaire in people with Cornelia de Lange Syndrome.

Roeleveld, N., Zielhuis, G.A., & Gabreels, F. (1997). The prevalence of mental retardation: a critical review of recent literature. *Developmental Medicine and Child Neurology*, *39,* 125–132.

Royal College of Psychiatrists, British Psychological Society and the Royal College Of Speech and Language Therapists (2007) *Challenging Behaviour: A Unified Approach. Clinical and Service Guidelines for Supporting People with Learning Disabilities who are at risk of receiving abusive or restrictive practices*.

Rojahn, J., Schroeder, S.R., & Hoch, T.A. (2008). *Self-Injurious Behavior in Intellectual Disabilities*. Elsevier: Amsterdam.

Rojahn J. (1986) Self-injurious and stereotypic behavior of noninstitutionalized mentally retarded people: prevalence and classification. *American Journal of Mental Deficiency, 91,* 268-76.

Ross, E., Arron, K., & Oliver, C. (2008). *The Mood Interest and Pleasure Questionnaire. Manual for administration and scoring*. University of Birmingham, Birmingham.

Ross, E., & Oliver, C. (2002). Self-injurious behaviour in people with intellectual disability, *Mental retardation, 15*, 477-481.

Ross, E. & Oliver, C. (2002). The relationship between levels of mood, interest and pleasure and 'challenging behaviour' in adults with severe and profound intellectual disability. *Journal of Intellectual Disability Research, 46,* 191-197.

Ross, E., & Oliver, C. (2003). Preliminary analysis of the psychometric properties of the Mood, Interest & Pleasure Questionnaire (MIPQ) for adults with severe and profound learning disabilities. *British Journal of Clinical Psychology, 42* , 81- 93.

Rutter, M., Bailey, A., & Lord, C. (2003). *Social Communication Questionnaire-WPS SCQ-WPS*. Los Angeles, CA: Western Psychological Services.

Sandman C. A. (1990/1991). The opiate hypothesis in autism and self-injury. *Journal of Child and Adolescent Psychopharmacology, 1(3),* 237–48.

Sanlaville, D., & Verloes, A. (2007). CHARGE syndrome: an update. *European Journal of Human Genetics, 15*, 389-399.

Sarria, E., Gomes, J.C., & Tamarit, J. (1996). Joint attention and alternative language intervention in autism: Implications of theory for parctice. In: S. von Tetzchner, & M.H. Jensen (Eds.), *Augmentative and alternative communication: European perspectives* (s. 49-64). London: Whurr.

Satake, H., Yoshida, K., Yamashita, H., Kinukawa, N., & Takagishi, T. (2003). Agreement between parents and teachers on behavioural/emotional problems in Japanese school children using the child behaviour checklist. *Child Psychiatry and Human Development, 34(2),* 111- 126.

Schopler, E., Reichler, R.J., De Vellis, R.F., & Daly, K. (1980). Toward objective classification of childhood autism rating scale. *Journal of Autism and Developmental Disorders, 10(1),* 91-103.

Shepherd, C., & Stephenson, J. (1992). Seizures and intellectual disability associated with Tuberous Sclerosis Complex in the west of Scotland. *Developmental Medicine and Child Neurology, 34,* 766-774.

Sieg, A., Hachmoeller-Elsenbach, U., & Elsenbach, T. (2001). Prospective evaluation of complications in outpatient GI endoscopy: a survey among German gastroenterologists. *Gastrointestinal Endoscopy, 53(6),* 620- 627.

Smalley, S., Burger, F., & Smith, M. (1994). Phenotypic variation of tuberous sclerosis in a single extended kindred. *Journal of Medical Genetics, 31,* 761-765.

Smalley, S. (1998). Autism and Tuberous Sclerosis Complex. *Journal of Autism and Developmental Disorders, 28(5),* 408-414.

Smiley, E., & Cooper, S. (2003). Intellectual disabilities, depressive episode, diagnostic criteria for psychiatric disorders for use with adults with learning disabilities/mental retardation (DC-LD). *Journal of Intellectual Disability Research, 47(1),* 62-71.

Smith, R. G., & Iwata, B. A. (1997). Antecedent influences on behavior disorders. *Journal of Applied Behavior Analysis, 30,* 343–375.

Sommer, A. (1993). Occurrence of the Sandifer complex in the Brachman–de Lange Syndrome. *American Journal of Medical Genetics, 47,* 1026–1028.

Staley, B., Montenegro, M., Major, P., Muzykewicz, D., Halpern, E., Kopp, C., Newberry, P., & Thiele, E. (2008). Self-injurious behavior and Tuberous Sclerosis Complex: Frequency and possible associations in a population of 257 patients. *Epilepsy and Behavior, 13*, 650-653.

Strachan, R., Shaw, R., Burrow, C., Horsler, K., Allen, D., & Oliver C. (2009). Experimental functional analysis of aggression in children with Angelman syndrome. *Research in Developmental Disabilities, 30,* 1095–1106.

Symons, F.J. (2011). Self-injurious behavior and neurodevelopmental disorders: relevance on nociceptive and sensory mechanisms. *Neuroscience and Biobehavioral Reviews, 35,* 1266-1274.

Symons, F.J., Clark, R.D., Hatton, D.D., Skinner, M., & Bailey, D.B. (2003). Self injurious behavior in young boys with fragile X syndrome. *American Journal of Medical Genetics Part A, 118,* 115–121.

Symons, F.J., & Danov, S.E. (2005). A prospective clinical analysis of pain behavior and self-injurious behavior. *Pain, 117,* 473-477.

Symons, F.J., Harper, V.N., McGrath, P.J., Breau, L.M., & Bodfish, J.W. (2009a). Evidence of increased non-verbal behavioral signs of pain in adults with neurodevelopmental disorders and chronic self-injury. *Research in Developmental Disabilities, 30,* 521-528.

Symons F.J., Harper V., Shinde S.K., Clary J., & Bodfish J.W. (2010). Evaluating a sham-controlled sensorytesting protocol for nonverbal adults with neurodevelopmental disorders: self-injury and gender effects. *Journal of Pain, 11*, 773–781.

Symons, F.J., Sutton, K.A., & Bodfish, J.W. (2001). Preliminary study of altered skin temperature at body sites associated with self-injurious behavior in Adults Who Have Developmental Disabilities. *American Journal on Mental Retardation, 106,* 336-343.

Symons, F.J., Thompson, A., & Rodriguez, M.C. (2004*).* Self-injurious behavior and the efficacy of naltrexone treatment: a quantitative synthesis. *Mental Retardation Developmental Disabilities Research Reviews, 10*, 193-200.

Symons, F.J., Wendelschafer-Crabb, G., Kennedy, W., & Heeth, W. (2009b). Degranulated mast cells in the skin of adults with self injurious behavior and neurodevelopmental disorders. *Brain, Behavior and Immunity, 23,* 365-370.

Taylor, L., Oliver, C., & Murphy, G. (2011). The chronicity of self-injurious behaviour: a long-term follow-up of a total population study. *Journal of Applied Research in Intellectual Disabilities, 24,* 105–117.

Taylor, D.V., Rush, D., Hetrick, W.P., & Sandman, C.A. (1993). Self-injurious behavior within the menstrual cycle of women with mental retardation. *American Journal on Mental Retardation, 97(6),* 659-664.

Tiger, J.H., Hanley, G.P., & Bruzek, J. (2008). Functional communication training: A review and practical guide. *Behavior Analysis in Practice, 1,* 16–23.

Tiller, S., Wilson, K.I., & Gallagher, J.E. (2001). Oral health status and dental service use of adults with learning disabilities living in residential institutions and in the community. *Community Dental Health, 18,* 167-71.

Toogood, S., & Timlin, K. (1996). The functional assessment of challenging behaviour: A comparison of informant-based, experimental and descriptive methods. *Journal of Applied Research in Intellectual Disabilities, 9,* 206–222.

Totsika, V., Toogood, S., Hastings, R.P., & Lewis, S. (2008). Persistence of challenging behaviours in adults with intellectual disability over a period of 11 years. *Journal of Intellectual Disability Research, 52,* 446–457.

Tsiouris, J.A. (2001). Diagnosis of depression in people with severe/profound intellectual disability. *Journal of Intellectual Disability Research, 45(2),* 151-120.

Tunnicliffe, P., & Oliver, C. (2011). Phenotype-environment interactions in genetic syndromes associated with severe or profound intellectual disability. *Research in Developmental Disabilities*, 32, 404-418.

Turner, M. (1997). Towards and executive dysfunction account of repetitive behaviour in autism. In James Russell (Ed.), *Autism as an executive disorder* (pp. 57–100). Oxford: Oxford University Press.

van Schrojenstein Lantman-de Valk, H.M.J., Metsemakers, J.F.M., Haveman, M.J., & Crebolder, H.F.J.M. (2000). Health problems in people with intellectual disability in general practice: a comparative study. *Family Practice, 17*, 405- 407.

van Schrojenstein Lantman-de Valk, H., Linehan, C., Kerr M., & Noonan-Walsh P. (2007). Developing health indicators for people with intellectual disabilities. The method of the Pomona project. *Journal of Intellectual Disability Research*, *6,* 427-434

van Slegtenhorst, M., de Hoogt, R., Hermans, H., Nellist, M., Janssen, B., Verhoef, S., Lindhout, D., van den Ouweland, A., Halley, D., & Young, S., ... Kwiatkowski, D.J. (1997). Identification of the tuberous sclerosis gene *TSC1* on chromosome 9q34. *Science, 277,* 805–808.

Verhulst, F.C., & Akkerhuis, G.W. (1989). Agreement between parents' and teachers' ratings of behavioural/emotional problems of children aged 4-12. *Journal of Child Psychiatry, 30(1),* 123-136.

Voepel-Lewis, T., Merkel, S., Tait, A.R., Trzcinka, A., & Malviya, S. (2002). The reliability and validity of the Face, Legs, Activity, Cry, Consolability observational tool as a measure of pain in children with cognitive impairment. *Anesthesia and Analgesia, 95,* 1224-9.

Walters, R.M. (1990). Suicidal behaviour in severely mentally handicapped patients. *British Journal of Psychiatry, 151,* 444–446.

Webb, D., Fryer, A., & Osborne, J. (1991). On the incidence of fits and mental retardation in tuberous sclerosis. *Journal of Medical Genetics, 28,* 395-397.

Webb, D.W., & Osbourne, J.P. (1992). New research in tuberous sclerosis. *British Medical Journal, 30,* 1647-8.

Weiner, D.M., Ewalt, D.E., Roach, E.S., & Hensle, T.W. (1998). The TSC: a comprehensive review. *Journal of the American College of Surgeons, 187*, 548-561.

Wilkins, L.E., Brown, J.A., Nance, W.E., & Wolf, B. (1983). Clinical heterogeneity in 80 home-reared children with Cri du Chat syndrome. *Journal of Pediatrics, 102 (4),* 528–533.

Winchel, R.M., & Stanley, M. (1991). Self-Injurious Behavior: A review of the behavioural and biology of self-mutilation. *American Journal of Psychiatry, 148(3),* 306-318.

Woolf, C.J., & Slater, M.W. (2000). Neuronal plasticity: increasing the gain in pain. *Science, 288*, 1765–1768.

APPENDICES

APPENDIX A

Parent cover letter

The Cerebra Centre for
Neurodevelopmental Disorders

UNIVERSITY OF
BIRMINGHAM

«AddressBlock»

20, November 2008

Re: «subject_name»

Dear «Title» «First_name_initial» «Surname»,

You may remember that you have taken part in our research before by completing questionnaires about the person you care for. We hope you found the feedback that we sent to you helpful. We are now extending this project by carrying out a five year follow-up to find out about changes since we first contacted you. This is the first study to follow people up over such a long period of time and the results of this study will be important for understanding how people change as they grow older. The more people that take part in the research, then the more meaningful the results are. A good response at this five year follow up will provide new and valuable information.

We would like to invite you and the person you care for to continue to take part in this research by completing the enclosed questionnaires. When we have analysed the information you send to us, we will provide personalised feedback about the person you care for and we will report any changes from previous assessments.

There is an information sheet enclosed that gives you more details about why the research is being carried out and what it will involve.

Please read the information sheet before completing the questionnaires and if you are unclear about any aspect of the study or have any questions then contact Professor [redacted] at the address below or on [redacted]

Thank you for your time and we look forward to hearing from you.

Yours sincerely

[redacted]
Professor of Neurodevelopmental Disorders

APPENDIX B

Information Sheet

INFORMATION SHEET

Background:
A team at the University of Birmingham is carrying out a questionnaire study to look at behaviour of people with [syndrome name].

What does it involve?
The booklet you have received contains several short questionnaires about some aspects of behaviour. The questionnaires should take no longer than one hour to complete.

Consent:
It is up to you whether or not you want your child or the person you care for to take part in the study. If you feel that it is appropriate, you may wish to discuss the project with your child or the person you care for.

Withdrawal:
Should you or the person you care for decide that you no longer wish to be involved in the study, the information that you have provided can be withdrawn at any time without you giving a reason. Even after the questionnaires have been completed, consent can be withdrawn and any data collected will be destroyed. This will not restrict access to other services and will not affect the right to treatment.

Confidentiality:
All details collected will be kept on a confidential database that is only accessible to those working on the project. Anonymity is ensured by storing the questionnaire data separately from any material that identifies participants. If published, information will be presented without reference to any identifying information.

At the end of the study:
Each parent/carer will receive a personalised feedback report on their child or the person they care for. A summary of the project's findings will be circulated to anyone involved who wishes to see a copy and a report will be written for syndrome support groups' newsletters. Any requests for advice concerning your child/the person you care for will be referred to Professor [redacted], Clinical Psychologist. It is possible that you may be invited to participate in further research after the study however, consenting to participate in this study does not mean that you are obliged to do so.

Review:
This study has been reviewed by the University of Birmingham, School of Psychology Research Ethics Committee. If you have any concerns about the conduct of this study please contact Professor [redacted] at the School of Psychology, University of Birmingham, Edgbaston, Birmingham, B15 2TT in the first instance.

Thank you very much for taking the time to read this information

APPENDIX C

Consent forms

CONSENT FORM A

For people aged below 16 or people over 16 who are unable to give consent.

Please initial the boxes

I confirm that I have read and understood the information sheet for the above study and have had the opportunity to ask questions. ☐

I understand that participation in the study is voluntary and that I am free to end my child's / the person I care for's involvement at any time, or request that the data collected in the study be destroyed, without giving a reason. ☐

I agree to the participation of my child's / the person I care for in the above study ☐

Please complete the information below

Participant's name……………………………………….date of birth…………………

Parent or guardian's name………………………………Mr/Mrs/Miss/Ms (please circle)

Parent or guardian's signature…………………………………Date………………..

Please state relationship with participant……………………………………………….

FOR OFFICE USE ONLY

Signature of researcher…………………………………………..Date………………...

CONSENT FORM B

For people aged 16 or over who are able to give consent.

Please initial the boxes

I confirm that I have read and understood the information sheet for the above study or that it has been explained to me and have had the opportunity to ask questions. ☐

I understand that participation in the study is voluntary and that I am free to end my involvement at any time, or request that the data collected in the study be destroyed, without giving a reason. ☐

I agree to take part in the above study ☐

Please complete the information below

Your name………………………………………. Your date of birth…………………

Your signature……………………………………Date……………………………….

FOR OFFICE USE ONLY

Signature of researcher…………………………………………..Date………………...

APPENDIX D1

Gastro-Oesophageal Distress Questionnaire (GDQ)

The GDQ

Instructions:

- This questionnaire asks about behaviours sometimes shown by people with learning disabilities.
- Please read the questions and examples carefully and indicate how often each behaviour has occurred in the **last two weeks** by circling the appropriate answer.

Does the person you care for:	**More than once an hour**	**Once an hour**	**Once a day**	**Once a week**	**Not occurred**
1. Arch his/her back, lie over arms of chairs or people on his/her back? ….........	*4*	**3**	**2**	**1**	**0**
2. Lie over an object on his/her stomach? e.g. a side of an arm chair.	*4*	**3**	**2**	**1**	**0**
3. Salivate excessively? …………………………………………………...........	*4*	**3**	**2**	**1**	**0**
4. Fidget, wriggle or move their body a great deal? ………………………….....	*4*	**3**	**2**	**1**	**0**
5. Place their hands or fingers in back of their mouth? ………………………	*4*	**3**	**2**	**1**	**0**
6. Chew on his/her clothes, fingers, hands or other parts of the body, objects or material? ……………………………………………………………………….	*4*	**3**	**2**	**1**	**0**
7. Grind their teeth? ……………………………………………………….......	*4*	**3**	**2**	**1**	**0**
8. Scratch, hit, press or rub around the upper chest or throat? ………..............	*4*	**3**	**2**	**1**	**0**
9. Drink, request or seek out an excessive amount of fluids? …………............	*4*	**3**	**2**	**1**	**0**
10. Cough, gag or regurgitate? ………………………………..........................	*4*	**3**	**2**	**1**	**0**
11. Appear in pain or discomfort (cry, groan or moan)? ………………..............	*4*	**3**	**2**	**1**	**0**
12. Refuse food even though they are probably hungry? ……………..................	*4*	**3**	**2**	**1**	**0**

13. Does the person you care for appear indecisive about food (edging towards table or food then moving away

repeatedly, taking food and putting it back)? *(please tick)* **Yes** ☐ **No** ☐

14. Does the person you care for wake during the night?	**Never**	**Once a week**	**Most nights**	**Every night**
15. Does the person you care for sleep sitting or propped up?	**Never**	**Once a week**	**Most nights**	**Every night**
16. Does the person you care for seem to have bad breath?	**Never**	**Once a week**	**At the same time everyday**	**All day every day**

17. Has the person you care for prone to respiratory tract infections? *(please tick)* **Yes** ☐ **No** ☐

If 'yes' please indicate how often they occur:

Monthly **Quarterly** **Every six months** **Annually**

Other (please specify)____

APPENDIX D2

Challenging Behaviour Questionnaire (CBQ)

THE CBQ

1) Has the person shown self-injurious behaviour <u>in the last month</u>? (e.g. head banging, head-punching or slapping, removing hair, self-scratching, body hitting, eye poking or pressing).

Yes ☐ No ☐

If the behaviour has not occurred, please go to question 6.
If the behaviour occurred in the past month please answer questions 2 to 5:

2) Place a tick next to the item for any of the following list of behaviours which the person displays in a repetitive manner (repeats the same movement/ behaviour twice or more in succession):

Hits self with body part (e.g. slaps head or face)…………………………………… ☐
Hits self against surface or object (e.g. bangs head on floor or table)…………… ☐
Hits self with object……………………………………………………………… ☐
Bites self (e.g. bites hand on wrist or arm)………………………………………... ☐
Pulls (e.g. pulls hair or skin)……………………………………………........... ☐
Rubs or scratches self (e.g. rub marks on arm or leg)……………………………. ☐
Inserts finger or objects (e.g. eye poking)………………………………........... ☐
Other form of self-injury, please specify:………………………………………… ☐

3) In the last month, for how long did the **longest** episode or burst of his behaviour last? *(Please circle one number)*

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) In the last month as a result of this behaviour, has physical contact or prevention or restraint by others been necessary e.g. blocking, taking objects from an individual, temporary restraint of an arm? *(Please circle one number)*

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

5) Think about how often this behaviour occurred in the last month. If there was no change and you watched the person now, then would you definitely see the behaviour:

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

6) Has the person shown physical aggression <u>in the last month</u>? (e.g. punching, pushing, kicking, pulling hair, grabbing other's clothing).

Yes ☐ No ☐

7) Has the person shown disruption and destruction of property or the environment <u>in the last month</u>? (e.g. tearing or chewing own clothing, tearing newspapers, breaking windows or furniture, slamming doors, spoiling a meal).

Yes ☐ No ☐

8) Has the person shown stereotyped behaviours i<u>n the last month?</u> (e.g. rocking twiddling objects, patting or tapping part of the body, constant hand movements, eye pressing).

Yes ☐ No ☐

APPENDIX D3

Wessex Questionnaire

WESSEX QUESTIONNAIRE

These items refer to the person you care for. For each question (A, B, C, D etc …), please enter the appropriate code in each box.

A) Wetting (nights) **1** = frequently **2** = occasionally **3** = never

B) Soiling (nights) **1** = frequently **2** = occasionally **3** = never ☐

C) Wetting (days) **1** = frequently **2** = occasionally **3** = never ☐

D) Soiling (days) **1** = frequently **2** = occasionally **3** = never ☐

E) Walk with help **1** = not at all **2** = not up stairs **3** = up stairs and elsewhere ☐

(note: if this person walks *by himself* upstairs and elsewhere, please also code '3' for 'walk with help')

F) Walk by himself **1** = not at all **2** = not up stairs **3** = up stairs and elsewhere ☐

G) Feed himself **1** = not at all **2** = with help **3** = without help ☐

H) Wash himself **1** = not at all **2** = with help **3** = without help ☐

I) Dress himself **1** = not at all **2** = with help **3** = without help ☐

J) Vision **1** = blind or almost **2** = poor **3** = normal ☐

K) Hearing **1** = deaf or almost **2** = poor **3** = normal ☐

L) Speech **1** = never a word **2** = odd words only **3** = sentences and normal **4** = can talk but doesn't ☐

If this person talks in sentences, is his/her speech:

1 = Difficult to understand even by acquaintances, impossible for strangers?

2 = Easily understood for acquaintances, difficult for strangers?

3 = Clear enough to be understood by anyone? ☐

M) Reads **1** = nothing **2** = a little **3** = newspapers and/or books ☐

N) Writes **1** = nothing **2** = a little **3** = own correspondence ☐

O) Counts **1** = nothing **2** = a little **3** = understands money values ☐

APPENDIX D4

The Activity Questionnaire (TAQ)

ACTIVITY QUESTIONNAIRE © C Burbidge and C Oliver, 2003

Instructions:

- **Please read each item carefully and circle the appropriate number on the scale, for the person you care for.**
- **Please ensure that you indicate a response for every item. If the particular behaviour does not apply, for example, if the person is not verbal or not mobile, please circle 0 on the scale.**

		Never/ almost never	**Some of the time**	**Half of the time**	**A lot of the time**	**Always/ almost all the time**
1.	Does the person wriggle or squirm about when seated or lying down?	0	1	2	3	4
2.	Does the person fidget or play with their hands and/or feet when seated or lying down?	0	1	2	3	4
3.	Does the person find it difficult holding still?	0	1	2	3	4
4.	Does the person find it difficult to remain in their seat even when in situations where it would be expected?	0	1	2	3	4
5.	Does the person prefer to be moving around or becomes frustrated if left in one position for too long?	0	1	2	3	4
6.	When the person is involved in a leisure activity (e.g. watching TV, playing a game etc.) do they make a lot of noise?	0	1	2	3	4
7.	When the person is involved in an activity, are they boisterous and/or rough?	0	1	2	3	4
8.	Does the person act as if they are "driven by a motor" (i.e. often very active)?	0	1	2	3	4
9.	Does the person seem like they need very little rest to recharge their battery?	0	1	2	3	4
10.	Does the person often talk excessively?	0	1	2	3	4
11.	Does the person's behaviour seem difficult to manage/contain whilst out and about (e.g. in town, in supermarkets etc.)?	0	1	2	3	4
12.	Do you feel that you need to "keep an eye" on the person at all times?	0	1	2	3	4
13.	Does the person you care for seem to act/do things without stopping to think first?	0	1	2	3	4
14.	Does the person blurt out answers before questions have been completed?	0	1	2	3	4
15.	Does the person start to respond to instructions before they have been fully given or without seeming to understand them?	0	1	2	3	4
16.	Does the person want things immediately?	0	1	2	3	4
17.	Does the person find it difficult to wait?	0	1	2	3	4
18.	Does the person disturb others because they have difficulty waiting for things or waiting their turn?	0	1	2	3	4

APPENDIX D5

Mood, Interest and Pleasure Questionnaire (MIPQ)

MOOD, INTEREST AND PLEASURE QUESTIONNAIRE

This questionnaire contains 12 questions – <u>you should complete all 12 questions</u>. Each question will ask for your opinion about particular behaviours, which you have observed in the last 2 weeks. For every question you should circle <u>the most appropriate</u> response e.g.

6) In the last two weeks, how interested did the person appear to be in his/her surroundings?

interested all	***interested most***	***interested about***	***interested some***	[✓ ***never***]

1) In the last two weeks, did the person seem…

sad all of the time	sad most of the time	sad about half of the time	sad some of the time	never sad

Please comment if anything has happened in the last two weeks which you feel might explain sadness if it has been observed (e.g. a bereavement):

2) In the last two weeks, how often did you hear positive vocalizations* when the person was engaged in activities*?

all of the time	most of the the time	about half of the time	some of the time	never

*<u>positive vocalizations</u>: e.g. laughing, giggling, “excited sounds” etc.
*<u>engaged in activities</u>: i.e. when someone is actively involved in any activity such as a mealtime, a social interaction, a self-care task or social outing etc.

3) In the last two weeks, do you think the facial expression of the person looked “flat”*…

all of the time	most of the the time	about half of the time	some of the time	never

*<u>flat expression</u>: expression seems lifeless; lacks emotional expression; seems unresponsive.

4) In the last two weeks, would you say the person…

cried every day	cried nearly every day	cried 3-4 times each week	cried once or twice each week	cried less than once each week

5) In the last two weeks, how interested did the person appear to be in his/her surroundings?

interested all of the time	interested most of the time	interested about half of the time	interested some of the time	never interested

6) In the last two weeks, did the person seem to have been enjoying life…

all of the time	most of the the time	about half of the time	some of the time	never

Please comment if there are any reasons why this person might not have been enjoying him/herself e.g. illness, being in pain, experiencing a loss etc.:

7) In the last two weeks, would you say the person smiled…

at least once every day	at least once nearly every day	3-4 times each week	once or twice each week	less than once each week

8) In the last two weeks, how disinterested did the person seem to be in his/her surroundings?

disinterested all of the time	disinterested most of the time	disinterested about half of the time	disinterested some of the time	never disinterested

9) In the last two weeks, when the person was engaged in activities*, to what extent did his/her facial expressions* suggest that s/he was interested in the activity?

interested all of the time	interested most of the time	interested about half of the time	interested some of the time	never interested

*engaged in activities: i.e. when someone is actively involved in any activity such as a mealtime, social interaction, self-care task or social outing etc.
*facial expressions: interest might be indicated by the degree to which the person's gaze is being directed at the person/things involved in an activity.

10) In the last two weeks, would you say that the person…

laughed every day	laughed nearly every day	laughed 3-4 times each week	laughed once or twice each week	laughed less than once each week

11) In the last two weeks, how often did you see gestures which appeared to demonstrate enjoyment* when the person was engaged in activities*?

all of the time	most of the the time	about half of the time	some of the time	never

*gestures which appear to demonstrate enjoyment: e.g. clapping, waving hands in excitement etc.
*engaged in activities: i.e. when someone is actively involved in any activity such as a meal time, social interaction, self-care task or social outing etc.

12) In the last two weeks, did the person's vocalizations* sound distressed…

all of the time	most of the the time	about half of the time	some of the time	never

*vocalizations: any words, noises or utterances.

APPENDIX D6

Non-Communicating Children’s Pain Checklist

Non-Communicating Children's Pain Checklist

This questionnaire asks how often the person you care for shows signs that they are in pain. Please answer according to how your child has behaved within the last two weeks.

Please read each question carefully and circle the number that best describes their behaviour over the last two weeks

0 = NOT AT ALL 1 = JUST A LITTLE 2 = FARILY OFTEN 3 = VERY OFTEN NA = NOT APPLICABLE

Over the last two weeks, how often has the person you care for made the following vocal sounds?

1. Moaning, whining, whimpering (fairly soft)…………………………………………………………	0	1	2	3	NA
2. Crying (moderately loud)……………………………………………………………………………	0	1	2	3	NA
3. Screaming/yelling (very loud)……………………………………………………………………….	0	1	2	3	NA
4. A specific sound or word for pain (e.g., a word, cry or type of laugh)……………………………………	0	1	2	3	NA

Over the last two weeks, how often has the person you care for shown these social behaviours?

5. Not cooperating, cranky, irritable, unhappy……………………………………………………………	0	1	2	3	NA
6. Less interactive with others, withdrawn………………………………………………………………	0	1	2	3	NA
7. Seeking comfort or physical closeness…………………………………………………………………	0	1	2	3	NA
8. Being difficult to distract, not able to satisfy or pacify…………………………………………………	0	1	2	3	NA

Over the last two weeks, how often has the person you care for shown the following facial signs?

9. A furrowed brow…………………………………………………………………………………………	0	1	2	3	NA
10. A change in eyes, including: squinching of eyes, eyes opened wide eyes frowning……………………	0	1	2	3	NA
11. Turning down of mouth, not smiling……………………………………………………………………	0	1	2	3	NA
12. Lips puckering up, tight, pouting, or quivering…………………………………………………………	0	1	2	3	NA
13. Clenching or grinding teeth, chewing or thrusting tongue out…………………………………………	0	1	2	3	NA

Over the last two weeks, how often has the person who you care for had a change in activity level in the following way?

14. Not moving, less active, quiet……………………………………………………………………………	0	1	2	3	NA
15. Jumping around, agitated, fidgety…………………………………………………………………………	0	1	2	3	NA

Over the last two weeks, how often has the person you care for shown the following limb and body activity?

16. Floppy………………………………………………………………………………………………………	0	1	2	3	NA
17. Stiff, spastic, tense, rigid………………………………………………………………………………	0	1	2	3	NA
18, Gesturing to or touching part of the body that hurts……………………………………………………..	0	1	2	3	NA
19. Protecting, favouring or guarding part of the body that hurts……………………………………………	0	1	2	3	NA
20. Flinching of moving the body part away, being sensitive to touch………………………………………	0	1	2	3	NA
21. Moving the body in a specific way to show pain (e.g., head back, arms down, curls up etc.)…………..	0	1	2	3	NA

Over the last two weeks, how often has the person you care for displayed the following physiological behaviours?

22. Shivering…………………………………………………………………………………………………	0	1	2	3	NA
23. Change in colour, pallor…………………………………………………………………………………	0	1	2	3	NA
24. Sweating, perspiring…………………………………………………………………………………….	0	1	2	3	NA
25. Tears………………………………………………………………………………………………………..	0	1	2	3	NA
26. Sharp intake of breath, gasping…………………………………………………………………………..	0	1	2	3	NA
27. Breath holding………………………………………………………………………………………………	0	1	2	3	NA

Over the last two weeks, how often has the person you care for displayed the following behavioural changes?

28. Eating less, not interested in food…………………………………………………………………………	0	1	2	3	NA
29. Increase in sleep…………………………………………………………………………………………	0	1	2	3	NA
30. Decrease in sleep…………………………………………………………………………………………	0	1	2	3	NA

APPENDIX D7

Social Communication Questionnaire (SCQ)

SOCIAL COMMUNICATION QUESTIONNAIRE

1. Is she/he now able to talk using short phrases or sentences? If no, skip to question 8.	**Yes**	**No**
2. Do you have a to and fro "conversation" with her/him that involves taking turns or building on what you have said?	**Yes**	**No**
3. Does she/he ever use odd phrases or say the same thing over and over in almost exactly the same way (either phrases she/he has heard other people use or ones that she/he makes up)?	**Yes**	**No**
4. Does she/he ever use socially inappropriate questions or statements? For example, does she/he ever regularly ask personal questions or make personal comments at awkward times?	**Yes**	**No**
5. Does she/he ever get her/his pronouns mixed up (e.g., saying *you* or *she/he* instead of *I*)	**Yes**	**No**
6. Does she/he ever use words that she/he seems to have invented or made up her/himself; put things in odd, indirect ways; or use metaphorical ways of saying things (e.g., saying *hot rain* for *steam*)?	**Yes**	**No**
7. Does she/he ever say the same thing over and over in exactly the same way, or insist that you say the same thing over and over again?	**Yes**	**No**
8. Does she/he ever have things that she/he seems to have to do in a very particular way or order, or rituals that she/he insists that you go through?	**Yes**	**No**
9. Does her/his facial expression usually seem appropriate to the particular situation, so far as you can tell?	**Yes**	**No**
10. Does she/he ever use your hand like a tool, or as if it were part of her/his own body (e.g., pointing with your finger, putting your hand on a doorknob to get you to open the door)?	**Yes**	**No**
11. Does she/he ever have any interests that preoccupy her/him and might seem odd to other people (e.g., traffic lights, drainpipes, or timetables)?	**Yes**	**No**
12. Does she/he ever seem to be more interested in parts of a toy or an object (e.g., spinning the wheels of a car), rather than using the object as it was intended?	**Yes**	**No**
13. Does she/he ever have any special interests that are *unusual* in their intensity but otherwise appropriate for her/his age and peer group (e.g., trains, or dinosaurs)?	**Yes**	**No**
14. Does she/he ever seem to be *unusually* interested in the sight, feel, sound, taste, or smell of things or people?	**Yes**	**No**
15. Does she/he ever have any mannerisms or odd ways of moving her/his hands or fingers, such as flapping or moving her/his fingers in front of her/his eyes?	**Yes**	**No**
16. Does she/he ever have any complicated movements of her/his whole body, such as spinning or repeatedly bouncing up and down?	**Yes**	**No**
17. Does she/he ever injure her/himself deliberately, such as by biting her/his arm or banging her/his head?	**Yes**	**No**
18. Does she/he ever have any objects (*other* than a soft toy or comfort blanket) that she/he *has* to carry around?	**Yes**	**No**
19. Does she/he have any particular friends or a best friend?	**Yes**	**No**
20. Does she/he ever talk with you just to be friendly (rather than to get something)?	**Yes**	**No**
21. Does she/he ever spontaneously copy you (or other people) or what you are doing (such as vacuuming, gardening, or mending things)?	**Yes**	**No**
22. Does she/he ever spontaneously point at things around her/him just to show you things (not because she/he wants them)?	**Yes**	**No**
23. Does she/he ever use gestures, other than pointing or pulling your hand, to let you know what she/he wants?	**Yes**	**No**
24. Does she/he nod her/his head to indicate *yes*?	**Yes**	**No**

25. Does she/he shake her/his head to indicate *no*?	**Yes**	**No**
26. Does she/he usually look at you directly in the face when doing things with you or talking with you?	**Yes**	**No**
27. Does she/he smile back if someone smiles at her/him?	**Yes**	**No**
28. Does she/he ever show you things that interest her/him to engage your attention?	**Yes**	**No**
29. Does she/he ever offer to share things other than food with you?	**Yes**	**No**
30. Does she/he ever seem to want you to join in her/his enjoyment of something?	**Yes**	**No**
31. Does she/he ever try to comfort you if you are sad or hurt?	**Yes**	**No**
32. If she/he wants something or wants help, does she/he look at you and use gestures with sounds or words to get your attention?	**Yes**	**No**
33. Does she/he show a normal range of facial expressions?	**Yes**	**No**
34. Does she/he ever spontaneously join in and try to copy the actions in social games, such as The Mulberry Bush or London Bridge is Falling Down?	**Yes**	**No**
35. Does she/he play any pretend or make-believe games?	**Yes**	**No**
36. Does she/he seem interested in other people of approximately the same age whom she/he does not know?	**Yes**	**No**
37. Does she/he respond positively when another person of approximately the same age approaches her/him?	**Yes**	**No**
38. If you come into a room and start talking to her/him without calling her/his name, does she/he usually look up and pay attention to you?	**Yes**	**No**
39. Does she/he ever play imaginative games with another person in such a way that you can tell that each person understands what the other is pretending?	**Yes**	**No**
40. Does she/he play cooperatively in games that need some form of joining in with a group of other people, such as hide-and-seek or ball games?	**Yes**	**No**

APPENDIX E

Descriptive data of participants in chapter 3

		CdC				**ASD**			
		GD+	**GD-**	**t/χ^2**	**P value**	**GD+**	**GD-**	**t/χ^2**	**P value**
N		18	28			105	176		
Age*	Mean (SD)	16.11 (11.0)	22.52 (12.60)	1.77	.08	10.24 (4.60)	12.94 (7.51)	3.33	.001
Gender	Male (%)	27.8	35.7	.31	.58	88.6	83	1.63	.20
Self [1] Help	Partly able/able[2] (%)	47.1	60.7	4.60	.71	82.9	94.4	41.51	<.001
Mobility[1]	Fully mobile[3] (%)	47.1	75.0	8.51	.74	93.3	96.0	2.57	.46
Vision[1]	Normal (%)	88.2	92.9	.29	.60	94.3	96.6	.87	.35
Hearing[1]	Normal (%)	100	82.1	3.14	.07	94.3	98.3	3.44	.06
Speech[1]	Partly verbal/Verbal (%)	76.5	78.6	.03	.88	88.6	96.6	6.98	<.01

		CdC				**ASD**			
		GD+	**GD-**	**t/χ^2**	**P value**	**GD+**	**GD-**	**t/χ^2**	**P value**
N		15	31			83	203		
Age*	Mean (SD)	16.6 (11.45)	21.66 (12.52)	1.32	.19	10.06 (4.80)	12.64 (7.18)	3.01	<.01
Gender	Male (%)	26.7	35.5	.36	.55	86.7	84.7	.21	.65
Self [1] Help	Partly able/able[2] (%)	42.9	61.3	5.46	.60	79.5	94.1	38.70	<.001
Mobility[1]	Fully mobile[3] (%)	42.9	74.2	13.31	=.01	91.4	96.6	5.05	.17
Vision[1]	Normal (%)	85.7	93.5	.73	.40	92.8	97.0	2.28	.10
Hearing[1]	Normal (%)	100	83.9	2.54	.11	92.8	98.5	6.34	.01
Speech[1]	Partly verbal/Verbal (%)	71.4	80.6	.47	.49	86.7	96.0	8.02	.01

APPENDIX F

Parent cover letter for participants with Tuberous Sclerosis Complex

The Cerebra Centre for
Neurodevelopmental Disorders

UNIVERSITY OF
BIRMINGHAM

April 2010

Dear Parent,

We are writing to inform you of a new research project that is being carried out at the Cerebra Centre for Neurodevelopmental Disorders at the University of Birmingham. Briefly, the research is a questionnaire study looking at different behaviours in children and adults with Tuberous Sclerosis Complex (TSC) that have received minimal attention within the literature. We would like to invite you and the person you care for with TSC to take part in this new research project.

The assessments used in this questionnaire study are designed for parental report and not self report. Unfortunately, this means that this study is currently only applicable to children with TSC (of all abilities) and adults with TSC who have an intellectual disability. We have contacted you through the Tuberous Sclerosis support group, meaning that your personal details will not be known to us unless you decide to take part in the study. If you are an adult with TSC who does not have an intellectual disability and you have received this information pack, we apologise as we have contacted you in error. If you are unsure whether or not you are eligible to take part in this study or if you do not think that you are eligible to take part but would like to hear about future research projects at the Cerebra Centre for Neurodevelopmental Disorders, please contact Professor [redacted] at the address below or [redacted]. Please also use these details if you would like a further explanation of why you are not eligible to take part in this current questionnaire study.

There is an information sheet enclosed that gives you more details about why the research is being carried out and what participation will involve. If you feel it is appropriate you may wish to discuss the research with the person you care for with TSC before a decision is made about taking part. When we have analysed the information you send to us, we will provide personalised feedback about the person you care for and we will report any changes from previous assessments.

If you and your child/person you care for with TSC would like to take part in the study then please complete the enclosed consent form and questionnaire pack that applies to you and return them in the pre-paid envelope provided. Only one of the enclosed questionnaire packs will apply to you depending on whether the child/person you care for is under or over the age of 16. There are two questionnaire packs enclosed;

- One **blue** questionnaire pack to be completed if the person with TSC is **16 years of age or older.**
- One **yellow** questionnaire pack to be completed if the person with TSC is **under the age of 16**.

Please take care to complete the correct consent form and questionnaire pack.

Please read the information sheets before completing the questionnaires and if you are unclear about any aspect of the study or have any questions then contact Professor [redacted] at the address below or on [redacted].

Thank you for your time and we look forward to hearing from you.

Yours sincerely

[redacted]

Professor of Neurodevelopmental Disorders

APPENDIX G

Parent information sheet for participants with Tuberous Sclerosis Complex

INFORMATION SHEET

Background:
A team at the University of Birmingham is carrying out a questionnaire study to look at behaviour of people with Tuberous Sclerosis Complex.

What does it involve?
The booklet you have received contains several short questionnaires about some aspects of behaviour. The questionnaires should take no longer than one hour to complete.

Consent:
It is up to you whether or not you want your child or the person you care for to take part in the study. If you feel that it is appropriate, you may wish to discuss the project with your child or the person you care for.

Withdrawal:
Should you or the person you care for decide that you no longer wish to be involved in the study, the information that you have provided can be withdrawn at any time without you giving a reason. Even after the questionnaires have been completed, consent can be withdrawn and any data collected will be destroyed. This will not restrict access to other services and will not affect the right to treatment.

Confidentiality:
All details collected will be kept on a confidential database that is only accessible to those working on the project. Anonymity is ensured by storing the questionnaire data separately from any material that identifies participants. If published, information will be presented without reference to any identifying information.

At the end of the study:
Each parent/carer will receive a personalised feedback report on their child or the person they care for. A summary of the project's findings will be circulated to anyone involved who wishes to see a copy and a report will be written for syndrome support groups newsletters. Any requests for advice concerning your child/the person you care for will be referred to Professor Chris Oliver, Clinical Psychologist. It is possible that you may be invited to participate in further research after the study however, consenting to participate in this study does not mean that you are obliged to do so.

Review:
This study has been reviewed by the University of Birmingham, School of Psychology Research Ethics Committee. If you have any concerns about the conduct of this study please contact Professor [redacted] at the School of Psychology, University of Birmingham, Edgbaston, Birmingham, B15 2TT in the first instance.

Thank you very much for taking the time to read this information

APPENDIX H

Personal/nominated consultee information sheets

Personal Consultee Information Sheet

Please read this information sheet if you care for a person who you have judged *is not* able to make an 'informed' decision about whether or not they would like to take part in the study or *is not* able to communicate that decision to you.

We would like to invite you to act as a personal consultee for the person that you care for.

What is a Personal Consultee?

In order to understand illness and disability, and to improve treatment and care, research is essential. That research may focus on the people with the illness or disability or on children under the age of 16, and may invite those people to participate. Some people will have capacity to make their own decision whether to take part in the research.

Others, possibly the youngest children or those most affected by the illness or disability, may not have that capacity. They may not be able to understand enough of the research to be able to give 'informed consent'. They may not be able to communicate a decision. The research provisions of the Mental Capacity Act are designed to allow such people to take part in research even though they cannot give valid consent of their own.

First, the research has to be approved by a Research Ethics Committee. Then, instead of asking the research participant for consent, the researcher must ask a consultee for an opinion whether the research participant would have wished to take part in the research.

Who can be a personal consultee?

Any person interested in the welfare of the proposed participant, for example:

- A family member, unpaid carer or friend
- A person acting under a Lasting Power of Attorney
- A court appointed deputy

Who cannot be a personal consultee?

- Paid carers and professionals
- People connected with the research (e.g. members of the research team)

Why have I been asked?

You have been asked to act as a personal consultee by a researcher because the researcher thinks you might be willing and able to do this because of your close relation with the proposed research participant.

If I agree to be a personal consultee, what will I have to do?

You will need to think about what the proposed participant's wishes and feelings about the research would be if they had capacity to make an informed decision and decide whether in your view the person should be involved in the research or not. This means you need to

- Look at the study information sheet.
- Think about whether or not the person would want to be involved in the research project if he or she had the capacity to make that decision.

You should not put forward your personal views on participation in the specific project or research in general, you must consider only what the person's views and interests are or would likely be. You should think about:

- What the broad aims of the research and the practicalities of taking part will mean for the proposed participant.
- How the specific activities in the research might impact the participant. For example, if the study involves activities in the afternoon when the person is most tired they might find it a strain or the research might involve an activity that the person particularly enjoys and thus would give them more pleasure.
- Any view previously expressed by the person on the overall nature of the research.

If you advise that the proposed participant would not have wanted to be involved in the research, they cannot be included in the research.

If you advise that the proposed participant would want to be involved, they may be included in the research. If the research commences but the person shows any sign at any stage that they are not happy to be involved in the research you can change your advice at any time without giving a reason, whereby the researcher must withdraw the person from the research. If the person seems unhappy at any point or shows any signs of objection, then they will be withdrawn from the research.

The research project has been approved by the (name) NHS Research Ethics Committee. If you wish to see proof of approval from this body, or you wish to discuss any concerns about acting as a personal consultee for the person that you care for, please contact (name of researcher) on (telephone) or by email at (email address).

I don’t want to be a personal consultee/ I am a paid carer and so cannot be a personal consultee- what do I do?

Please try to suggest an alternative person who might like to act as a personal consultee for the potential participant, please pass the project information pack on to that person.

Where can I get more information and guidance?

More information is available from:

Department for Constitutional Affairs (2007) *Mental Capacity Act 2005 Code of Practice*
http://www.dca.gov.uk/legal-policy/mental-capacity/mca-cp.pdf

Department of Health (2007) *Guidance on nominating a consultee for research involving adults who lack capacity to consent* (consultation)
http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_076207

Mental Capacity Implementation Programme (2007) *Making Decisions: a guide for family, friends and unpaid carers. Second edition*
http://www.dca.gov.uk/legal-policy/mental-capacity/mibooklets/booklet02.pdf
A printed copy of this booklet is available by telephoning 023 80878038.

I have decided that I want to be a personal consultee- what do I do?

Please go back to the (Study Title) Information Sheet and continue reading.

Nominated Consultee Information Sheet

You may need this information if you are a paid carer for a person who you have judged *is not* able to make an 'informed' decision about whether or not they would like to take part in the study or *is not* able to communicate that decision to you. If this person does not have any family members, unpaid carers or friends who are in regular contact and are able and willing to act as a personal consultee, does not have anyone acting under Lasting Power of Attorney or a court appointed deputy, then we would like to invite you to act as a nominated consultee for the person that you care for.

What is a Nominated Consultee?

In order to understand illness and disability, and to improve treatment and care, research is essential. That research may focus on the people with the illness or disability or on children under the age of 16, and may invite those people to participate. Some people will have capacity to make their own decision whether to take part in the research.

Others, possibly the youngest children or those most affected by the illness or disability, may not have that capacity. They may not be able to understand enough of the research to be able to give 'informed consent'. They may not be able to communicate a decision. The research provisions of the Mental Capacity Act are designed to allow such people to take part in research even though they cannot give valid consent of their own.

First, the research has to be approved by a Research Ethics Committee. Then, instead of asking the research participant for consent, the researcher must ask a consultee for an opinion whether the research participant would have wished to take part in the research.

Who can be a nominated consultee?

- Any person interested in the welfare of the proposed participant who works with the participant in a professional capacity.

Who cannot be a nominated consultee?

- People connected with the research (e.g. members of the research team)

Why have I been asked?

You have been asked to act as a nominated consultee by a researcher because the researcher thinks you might be willing and able to do this because of your professional relationship with the proposed research participant.

If I agree to be a nominated consultee, what will I have to do?

You will need to think about what the proposed participant's wishes and feelings about the research would be if they had capacity to make an informed decision and decide whether in your view the person should be involved in the research or not. This means you need to

- Look at the study information sheet.
- Think about whether or not the person would want to be involved in the research project if he or she had the capacity to make that decision.
- You may need to seek the advice of friends/ family/ other paid carers of the person you care for in order for you to best advise us on what the person's wishes and feelings would be.

You should not put forward your personal views on participation in the specific project or research in general, you must consider only what the person's views and interests are or would likely be. You should think about:

- What the broad aims of the research and the practicalities of taking part will mean for the proposed participant.
- How the specific activities in the research might impact the participant. For example, if the study involves activities in the afternoon when the person is most tired they might find it a strain or the research might involve an activity that the person particularly enjoys and thus would give them more pleasure.
- Any view previously expressed by the person on the overall nature of the research.

If you advise that the proposed participant would not have wanted to be involved in the research, they cannot be included in the research.

If you advise that the proposed participant would want to be involved, they may be included in the research. If the research commences but the person shows any sign at any stage that they are not happy to be involved in the research you can change your advice at any time without giving a reason, whereby the researcher must withdraw the person from the research. If the person seems unhappy at any point or shows any signs of objection, then they will be withdrawn from the research.

The research project has been approved by the (name) NHS Research Ethics Committee. If you wish to see proof of approval from this body, or you wish to discuss any concerns about acting as a personal consultee for the person that you care for, please contact (name of researcher) on (telephone) or by email at (email address).

I don't want to be a nominated consultee - what do I do?

Please try to suggest an alternative person who might like to act as a nominated consultee for the potential participant, please pass the project information pack on to that person.

If no-one can be found who is willing and able to act as a consultee for the person you care for then the person will not be able to participate in the research study.

Where can I get more information and guidance?

More information is available from:

Department for Constitutional Affairs (2007) *Mental Capacity Act 2005 Code of Practice*
http://www.dca.gov.uk/legal-policy/mental-capacity/mca-cp.pdf

Department of Health (2007) *Guidance on nominating a consultee for research involving adults who lack capacity to consent* (consultation)
http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_076207

Mental Capacity Implementation Programme (2007) *Making Decisions: a guide for family, friends and unpaid carers. Second edition*
http://www.dca.gov.uk/legal-policy/mental-capacity/mibooklets/booklet02.pdf
A printed copy of this booklet is available by telephoning 023 80878038.

I have decided that I want to be a nominated consultee- what do I do?

Please go back to the (Study Title) Information Sheet and continue reading.

APPENDIX I

Consent forms for participants with Tuberous Sclerosis Complex

Consent Form

Section 1: Please initial ONE of the following statements:

I am over 16 years old and I have read and understood the (study title) information sheet. I understand what the study is trying to find out and why, and what I would have to do if I take part. I have enough information to decide whether or not I want to take part.	

I am under 16 years old and I have read and understood the (study title) information sheet. I understand what the study is trying to find out and why, and what I would have to do if I take part. I have enough information to decide whether or not I want to take part.	

I have read and understood the (study title) information sheet on behalf of someone that I care for. I have discussed the information with the person that I care for. I feel that the person I care for understands what the study is trying to find out and why, and what they would have to do if they take part. The person that I care for has made an informed decision about whether or not they wish to participate and they have communicated this decision to me.	

I have read and understood the (study title) information sheet on behalf of someone that I care for. I feel that the person that I care for *does not* have the capacity to make an informed decision about whether or not they wish to participate in the study or that they cannot communicate this decision to me.	

Section 2: Please ONLY fill in this section if you are completing this form on behalf of someone that you care for who you feel *does not* have the capacity to make an informed decision about whether or not they wish to participate.

Please read the following statements and initial each one to show that you agree.

I have read and understood the 'Personal Consultee Information sheet **or** if I am acting as a 'Nominated Consultee' then I have read and understood the 'Personal Consultee Information Sheet' and the 'Nominated Consultee Information Sheet'.	

I agree to act as a personal consultee for the person that I care for and will advise the researchers about what I feel would be that person's wishes throughout the study **or** there is no appropriate person able/ willing to act as a personal consultee for the person that I care for (in a professional capacity) so I agree to act as a nominated consultee for the person that I care for and will advise the researchers about what I feel would be that person's wishes throughout the study.	

I understand that if I feel that the wishes of the person that I care for have changed at any point during the study then I must advise the researchers so that they can act accordingly.	

I will complete the rest of this form to advise the researchers of what I feel would be the wishes/ interests of the person that I care for in my capacity as a personal consultee **or** nominated consultee.	

Name: ____

Relationship to participant: ____

Signature: ____

Date: ____

Section 3: Please read the following statements and initial each one to show that you agree.

I have read and understood the (study title) information sheet.	
I understand that it is up to me whether or not I take part in the study **or** if I am acting as a consultee, I understand that it is up to me to advise on whether or not the person that I care for would want to take part.	
I understand that even after consent has been given, participants can withdraw from the study at any time simply by telling the researchers of their decision or by asking someone else to tell the researchers for them **or** if I am acting as a consultee, I can advise the researchers if I feel that the person that I care for would want to withdraw from the study at any time.	
I understand that all information collected during the study will be confidential. Only members of the research team at the Cerebra Centre for Neurodevelopmental disorders will know who has participated in the study. All information collected during the study will be stored in locked cabinets that only members of the research team will have access to. No names will be published in any reports. Information will be treated as strictly confidential and handled in accordance with the provisions of the Data Protection Act 1998.	
I understand that as part of the above study, video/voice recordings of participants and members of participants' families who are involved in the research may be made and stored for further review.	
I understand that the University of Birmingham will hold the copyright of any video/voice recordings collected during the study but that this does not entitle the University of Birmingham to edit, copy or use the videos for teaching purposes without my written permission.	
I am happy to be contacted in the future by the University of Birmingham regarding the use of video/ audio recordings for teaching purposes.	
I would like to give my 'informed' consent to take part in the study (study title) **or** if I am acting as a consultee I would like to advise the researchers that in my opinion the person that I care for would wish to provide their consent to take part in the study.	

Participant's name: ____

Your name if you are not the participant: __________________________

Your relationship to the participant: ______________________________

Signature: ____

Date: ____

Section 4: This section is to be completed by the parent/ legal guardian of participants under 16 years old who have made the informed decision that they would like to participate in the research and have completed sections 1 and 3 above.

Please read the following statements and initial each one to show that you agree.

I have read and understood the (study title) information sheet.	
I understand that I can withdraw my consent for my child to participate in the study at any time and that this will not affect the future access that I/ my child has to services.	
I understand that all information collected during the study will be confidential. Only members of the research team at the Cerebra Centre for Neurodevelopmental disorders will know who has participated in the study. All information collected during the study will be stored in locked cabinets that only members of the research team will have access to. No names will be published in any reports.	
I would like to give my 'informed' consent for my child to take part in the study (study title).	

Name: ____

Signature: ____

Date: ____

APPENDIX J

Parent cover letter: The influence of pain and environmental factors on challenging behaviour

The Cerebra Centre for
Neurodevelopmental Disorders

UNIVERSITY OF
BIRMINGHAM

[redacted]

[redacted]

«AddressBlock»
<<Date>>

Re: Invitation to participate in research

Dear «Title» «First_name_initial» «Surname»,

We have contacted you via the <insert syndrome group> foundation in order to invite you and your child to take part in a research project being carried out at the Cerebra Centre for Neurodevelopmental Disorders at the University of Birmingham. This research will investigate health related pain and challenging behaviour in individuals with X syndrome and other genetic syndromes. Although we will be looking at pain and challenging behaviour, it is important that we recruit children with a wide range of both states of health and levels of challenging behaviour. Therefore, please do not be put off from being involved in this study if you feel that the person you care for does not experience pain or display any challenging behaviour- the information we will get from these individuals' participation will still be extremely valuable to us.

Through this study we hope to further our knowledge about painful health conditions in X syndrome and the link between pain and challenging behaviour within this group. By enhancing our knowledge in this area we hope to improve upon intervention strategies for tackling challenging behaviour and also to increase our ability to recognise the signs of pain and discomfort in children with X syndrome and other genetic syndromes.

There is an information sheet enclosed that gives you more details about why the research is being carried out and what it will involve.

Please read the information sheet before completing the expression of interest form and if you are unclear about any aspect of the study or have any questions then contact Professor [redacted] using the details above or [redacted] [redacted] at [redacted]@bham.ac.uk or on [redacted].

If you would like to be involved in this study or would like to receive further information, please complete the 'expression of interest' form and return it in the pre-paid envelope. Expressing interest in the study will not commit you to participation and you will be able to decide at a later date whether or not you would like to take part.

Many thanks for your support.

Yours Sincerely,

Professor [redacted]　　　　[redacted]
(Clinical Psychologist)　　　　(Research Psychologist)

APPENDIX K

Parent information sheets: The influence of pain and environmental factors on challenging behaviour

Investigating the health related pain and challenging behaviour in children with genetic syndromes

Parent Information Sheet

Please read this information carefully before deciding whether you wish to take part in the study. If you have any further questions please contact Professor [redacted] on ([redacted] or at [redacted]@bham.ac.uk. If you have any medical/ other problems which make it difficult for you to read this information, please contact Professor Chris Oliver 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 expression of interest form and return it to us in the prepaid envelope provided

Background

We would like to invite you to take part in a research study being conducted at the Centre for Neurodevelopmental Disorders, University of Birmingham. This research work is led by Professor Oliver and will investigate health related pain and challenging behaviour such as aggression and self injury in children with **X syndrome** and other neurodevelopmental disabilities. Through this study we hope to further our understanding of the relationship between pain and discomfort associated with health problems and challenging behaviour. We also aim to produce an easy to administer pain assessment tool that can be used to identify pain in children with an intellectual disability. The results from this study will be used directly to design the pain assessment measure. The more people that take part in this research, the more meaningful the results will be and the more effective the pain measure will be in identifying and rating pain in children with **X syndrome**. There will also be an optional follow up aspect to the study that some participants will be asked to take part in. This follow up study will allow researchers to test the newly developed pain assessment tool.

Aims of the study

1. To investigate health related pain and challenging behaviour in children with X syndrome and other genetic syndromes associated with an intellectual disability.

2. To develop an easy-to-use pain assessment tool.

Review

The study has been approved by Birmingham East, North and Solihull NHS Research Ethics Committee.

Who can be involved in the study?

We are looking for children with a confirmed diagnosis of **X syndrome,** who are aged between 4 and 15.

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

What does it involve?

Participation in the research project will involve the following:

- A day of activities/ assessments with your child either at their school or at home, depending on where suits you best. These activities will mirror activities that your child would usually encounter in a typical school day.
- Completion of a questionnaire pack asking questions regarding your child's behaviour.
- Completion of two short interviews, conducted over the phone at a time that suits you. Your child's teacher will also be asked to complete one of these interviews.
- Several short observations of your child followed by the completion of a short assessment form. This will take a maximum of 5 minutes each time. These observations and assessments will be conducted by yourselves, your child's teacher and the researchers.
- We will ask for access to your child's medical records for the past 12 months. This will only be done with your explicit consent

An example of the timetable for collecting the above information from you, your child/person you care for is shown below.

Stage 1: Return consent form
Stage 2: Complete questionnaire pack.
Stage 3: Two research workers will visit you and your child/the person you care for at home or you and your child/the person you care for will travel to the University of Birmingham for a day of activities and assessments.

You may be asked to repeat the questionnaire pack from stage two.

Your child's teacher will be asked to complete an interview assessment regarding your child's behaviour.
Stage 4: Interviews conducted over the phone and GP records collected.
Stage 5: You will receive a detailed individual feedback form about your child's/person you care for's assessments.
Follow up stage: Previous assessments will be repeated. This stage is optional and will only involve some participants.

Where will the research take place?

The research will involve several stages. Completion of the questionnaire packs can be completed by you in your own time. Interviews with you will take place over the phone at a time that suits you and they will take place with the teacher at a time that suits them during the school day. The assessment visit will take place either at your home or your child's/ person you care for's school, depending on whatever suits you best.

Who will be involved in collecting the data?

Members of the research team at the Cerebra Centre for Neurodevelopmental Disorders including Professor [redacted] and research psychologist, [redacted].

How long will participation in the study take?
Participation in this study will take up to 3 months from the time of the returned consent forms and questionnaire pack until the research visit.

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

There is a chance that your child might engage in their usual challenging behaviour throughout the assessment day. The assessments that we use are designed to mirror usual day-to-day activities so we would therefore anticipate that your child's behaviour would be at the same level expected from 'every day life'. Therefore, your child would not be at any heightened risk through being involved in this study. Before the day of our visit, we would ask you for information about your child's typical challenging behaviour levels and if your child's challenging behaviour does exceed what would normally be expected, we would end the assessment.

Participation in this study will not restrict your ability to access any services or support for yourself or the person you care for.

What are the potential benefits for participants from taking part?
All participants will receive an individual feedback report detailing how your child/person you care for performed on all of the assessments. If requested, this feedback report will be circulated to other interested individuals. This information could be used within an education and/or medical setting to ensure your child gets the help that is most appropriate for their needs.

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.

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. Yours and your child's personal details will be held separately to data collected throughout the study and it will only be possible to connect results to individuals via a unique identification number.

How video recordings will be made?
Observations and video recordings will only take place during previously specified times that have been agreed by teachers and parents/ legal guardians. Your child's privacy and dignity will be respected and video recordings will not take place if children are in a state of undress or when there is evidence that the observations are causing distress. Parents/ legal guardians and teachers can ask to see a copy of the videotape. When videotapes are not in use they will be stored in a locked cabinet in the School of Psychology, University of Birmingham and will only be viewed by research workers from the University of Birmingham. Information identifying your child will not be stored on or with the tape.
The video recordings may only be viewed by legal guardians, individuals providing a service to the person, Professor Chris Oliver and research staff at the University of Birmingham. Any data that are derived from the tape will remain anonymous. Video recordings will not be shown for the purpose of teaching.

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

You and your child or 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.

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.

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 unique identification number. 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.

Consent

After having read all of the information and having received appropriate responses to any questions that you may have about the study you will be asked to give your consent to participate in the study if you decide that you do wish to participate. We need to receive consent for your own involvement in the study and that of the person you care for before we can arrange for them to participate. If you feel that the person you care for has the ability to understand what participation in the study involves for them, and they wish to provide their consent to participate, please help them to complete ‘consent form A’ before completing ‘consent form B’ yourself. If you feel that a symbol sheet would aid your child in understanding what is involved in the study, then please contact us and we will send you a simplified symbol information sheet through the post. If you feel that your child does not have the ability to fully understand what is being asked of them, please only complete ‘consent form B’

Any concerns of queries?

If you are unclear about any aspect of the study or have any questions, please do not hesitate to contact Professor [redacted] by telephone: [redacted], by email: [redacted] or at the address below:

[redacted]

Birmingham
B15 2TT

If your questions are not answered to your satisfaction then you can contact Glyn Humphreys, Head of Psychology, who is external to this project at the University of Birmingham. He can be contacted on [redacted]

Thank you very much for taking the time to read this information – please keep this information sheet for future reference

APPENDIX L

Consent forms: The influence of pain and environmental factors on challenging behaviour

Investigating the health related pain and challenging behaviour in children with genetic syndromes

Study Director: Professor [redacted]

Consent form A: Please complete this form if you are a person with X syndrome.

1. Has somebody else explained the project to you or have you read the information? YES/NO
2. Do you understand what the project is about? YES/NO
3. Have you asked all of the questions you want? YES/NO
4. Have you had your questions answered in a way you understand? YES/NO
5. Do you understand it is OK to stop taking part at any time? YES/NO
6. Are you happy to take part? YES/NO

If any answers are ‘no’ or you don’t want to take part, don’t 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:

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

Your name:____

Date:______________________

The person who explained this project to you needs to sign too. This should be your parent/guardian.

Print name:____________________________ Sign:__________________________ Date:___________________

PTO…continued overleaf

Consent form B: Please complete this form if you are a parent/carer/guardian of a person with X syndrome.

Please initial box…

1. I confirm that I have read and understood the 'pain and challenging behaviour' information sheet. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily. ☐

2. 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. ☐

3. I understand that relevant sections of my child's/person I care for's GP medical notes regarding 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. ☐

4. 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. ☐

5. I agree to take part in the above study. ☐

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

6. I agree for researchers to contact me regarding mine and my child's involvement in futures aspects of this study. I understand that neither I nor my child is obligated to take part in future aspects of the study. ☐

Print Name: ______________________________ Telephone number: ______________

Address: ____

____

____________ Relationship to participant:____________ Signature:

__________________ Date: ______________

PTO…continued overleaf

Access to Medical Records

Please fill in the details below so that we can locate your child's medical records:

Name of the medical professional (GP or clinical geneticist) who has these records regarding genetic diagnosis:____
____

Name of the hospital/ organisation where the records are held or were initially recorded: _______
____
____
____

Address of the hospital/ organisation where the records are held or were initially recorded:______
____
____
____
____

Any other contact details for the professional/ institution holding the records (e.g. telephone number, email address): ____
____
____

APPENDIX M

Direct observation behaviour codes

*Duration variable

(vprom)Verbal prompt- verbal demand to complete a task. Includes pointing whilst prompting.
(gprom)*Gestural prompt- model of correct response by researcher, guiding ppt towards a location with hands
(pprom)*Physical prompt- hand over hand demand to complete a task
(tout)*Timeout- removal of demand and task materials contingent on CB

(den)* Denial - taking items away from ppt if they are resisting the item being removed, physically moving ppt away from something, blocking attempts to escape room or damage items, blocking access to items that could cause injury including blocking SIB, verbal request to stop doing or not do something (not referring to consequences in low attention condition- these to be coded as attention).
(ratt) *Attention- physical contact initiated by researcher, verbal contact from researcher

(PhyIn)* Physical initiation- physical contact initiated and maintained by the ppt with someone else in the room
(PhyDi)* Physical dissent- pulling/pushing away from physical contact or physical prompts

(CoSter)*Contact stereotypy- repetitive behaviour involving contact between body parts or body and object i.e. hand mouthing or face/body tapping ("potentially injurious" often coded as individual behaviours i.e. hand mouth and chin tap, especially if there is more than one topography shown)
(NCoSter)*Noncontact stereotypy- such as hand flapping, body rocking etc (again, different topographies often coded separately)
(ObjSter)*Object stereotypy- stereotypical use of an item i.e., twirling or spinning materials etc

(VocPos)*Voc pos/neutral- positive vocalisations/laughing. If the vocalisations are ongoing, count two clear seconds before turning off the vocalisations.
(PosFac)*Pos. face - positive facial expression/smiling

(Legs) *Legs- no weight on legs. Legs free to move (i.e. when sitting/lying being held etc.)
(ReLeg)*Relaxed legs- relaxed leg position/movement when free to move. (NOT uneasy, restless, tense, increased tone, rigidity, intermittent flexion/extension of limbs, kicking, legs drawn up, tremors).

(GORA) *GORA (relaxed activity) – Any normal movement such as walking, reaching, moving objects. Only to be turned off if ppt is completely inactive (no movement at all) for 2 seconds.

Physical aggression

(AggHit) *Agg. hit- contact made with intended force between the ppts hand or object that ppt is holding and the researchers body.
(AggPinch) *Agg. pinch- ppt pinch/ or attempt to pinch (if clothes prevent it) researcher
(AggHair)*Agg. hair pull- ppt holds onto and deliberately pulls the researcher's hair
*(AggBite)- ppt bite/ attempt to bite (if clothing prevents it) the researcher
(AggHBang)Agg. head bang- contact made with deliberate force between the ppt's head and the researcher.
(AggSpit) Spit- Ppt. spit in the direction of the researcher.
(AggKick) *Agg. kick- contact made with intended force between the ppt.'s feet and the researcher
(AggGrab)* -Participant holding onto and pulling researcher towards the participant.

(AggPush)* - Participant pushing the examiner away when an interaction has NOT been initiated by the researcher. E.g. during the low attention condition. 5
(Shove)* - Participant using force to move the researcher out of the way to get access to something/somewhere.
(scratch)*- Dragging nails accross researchers or attempt to drag nails accross researcher's skin if clothes prevent contact.
(AggPress)* Participant pushing on the examiner, without the intention of moving the examiner.

AGGRESSION- all occurrences of aggression. Includes all variables listed under aggression.

Self injurious behaviour

When naming body part, use separate codes for each separate body part specifically targeted.

(hit)*Self hit (name body part) - contact made with intended force between the ppt's hand or object in a hand and other body part/attempted contact (if prevented by clothing/protective devices)
(pinch)*Self pinch (name body part) - ppt pinch self
(hair)*Self hair pull- ppt holds onto and deliberately pulls their own hair
(bite)*Self Bite (name body part) - ppt bite/ attempt to bite (if clothing prevents it) self
(HeadBang)*Self head bang- contact made with deliberate force between the ppt's head and an object (can include door/floor etc.). If the participant uses the researcher as an object to facilitate the head bang, then code as SIB, if the researcher is the receiver of the behaviour, then code as Agg.
(HeadBangNC) Throwing head back and forward violently (head bang without connecting with object- non contact head bang)
(FinMouth)*Finger mouthing- pushing finger or toe into mouth (not biting)
(ObjMouth)*Object mouthing- pushing object into mouth
(kick) *Self kick- contact made with intended force between the ppt.'s feet and other body part.
(EyePoke)- using hand/finger or object to poke eye
(FinEar)*- pushing finger into ear with force
HeadPush- using hand to push on head
HeadPushO- using researchers hand to push on head
ChinPush-using hand or object to push on chin.
NosPic- scratching of picking the inside of the nose

SELFINJURY- all occurrences of self injury. Includes all variables listed under the self injury section

Destruction

(destr)*Destruct- destructive behaviour- i.e. swiping items off table, throwing, tearing, biting object, kicking or hitting objects with force etc
(Spit)*- deliberately putting saliva on an object or person and playing with the saliva.

DESTRUCT- destruct and spit combined

CHALLENGE- Any occurrence of DESTRUCT, SELFINJURY or AGGRESSION.

Pain indicators

(Ges)Gesturing- gesturing or touching body part that hurts, guarding body part that hurts. Not as part of playful interaction.
(SeSo)*Self sooth- pressure on body part/ rubbing body part. Not as part of playful interaction.

NEGFACUSE- Negative face when the face is on camera.
NONReLegs- Non relaxed legs when the legs are free to move.
(VocNeg)*Voc neg- negative vocalisations i.e. moaning, whining etc
(cry)*Crying- crying (with tears/ obvious crying if tears not possible)
(NegFac)*Neg. face- Frown/ worried face
(NGA) *NGA (not GORA)- Squirming, shifting back and forth, jerking etc. Not rocking.
(RigAc) *Rigid activity- rigid/tense position movement (NOT GORA).
NegAFFECT*- negative face (when on camera) or negative vocalisation or cry.

Self Stimulation
(mast) masturbating, self-stimulating genitals/genital area
GroPush- pushing on groin/lower stomach area.
(noise) making noises with/from mouth to self-stimulate

Extras

(loff)* – legs off camera. If you can’t see enough to infer what action the whole legs are making (both legs). If you are unsure, always code loff.
(foff)*- face off camera- If you can’t see enough to infer the facial expressions. If you are unsure, always code foff.
(mat)*- holding/manipulating objects
(ComSign)*- using sign language or gestures to communicate
(Verb)- using words or sounds to communicate

APPENDIX N

Face, Legs, Activity , Cry, Consolability (FLACC)

Child's name___________________ Date____________________

FLACC Pain assessment

Please observe your child/person you care for over a period of **3-5 minutes**. Following your observation, please place a score in the score column relating to each category of the scale (Face, legs, activity, cry and consolability). Use the scoring criteria to determine what score to give your child.

Categories	Scoring Criteria			**Score**
	0	**1**	**2**	
Face	No particular expression or smile, eye contact and interest in surroundings	Occasional grimace or frown, withdrawn, disinterested, worried look to face, eyebrows lowered, eyes partially closed, cheeks raised, mouth pursed	Frequent to constant frown, clenched jaw, quivering chin, deep furrows on forehead, eyes closed, mouth opened, deep lines around nose/lips	
Legs	Normal position or relaxed	Uneasy, restless, tense, increased tone, rigidity, intermittent flexion/extension of limbs	Kicking or legs drawn up, hypertonicity, exaggerated flexion/extension of limbs, tremors	
Activity	Lying quietly, normal position, moves easily and freely	Squirming, shifting back and forth, tense, hesitant to move, guarding, pressure on body part	Arched, rigid, or jerking, fixed position, rocking, side to side head movement, rubbing of body part	
Cry	No cry/moan (awake or asleep)	Moans or whimpers, occasional cries, sighs, occasional complaint	Crying steadily, screams, sobs, moans, grunts, frequent complaints	
Consolability	Calm, content, relaxed, does not require consoling	Reassured by occasional touching, hugging, or 'talking to'. Distractible	Difficult to console or comfort	

APPENDIX O

Non-Communicating Children’s Pain Checklist- Revised

(parent modified version)

NCCPC-R

This questionnaire asks how often the person you care for shows signs that they are in pain. Please answer according to how your child has behaved within THE LAST WEEK. Please read each question carefully and circle the number that best describes their behaviour over the last two weeks

0 = not at all 1 = just a little 2 = fairly often 3 = very often NA = not applicable

Over the last week, how often has the person you care for made the following vocal sounds?

1. Moaning, whining, whimpering (fairly soft)…………………………… 0 1 2 3 NA
2. Crying (moderately loud)………………………………………………… 0 1 2 3 NA
3. Screaming/yelling (very loud)…………………………………………… 0 1 2 3 NA
4. A specific sound or word for pain (e.g., a word, cry or type of laugh)…… 0 1 2 3 NA

Over the last week, how often has the person you care for shown these social behaviours?

5. Not cooperating, cranky, irritable, unhappy……………………………… 0 1 2 3 NA
6. Less interactive with others, withdrawn…………………………………… 0 1 2 3 NA
7. Seeking comfort or physical closeness……………………………………… 0 1 2 3 NA
8. Being difficult to distract, not able to satisfy or pacify…………………… 0 1 2 3 NA

Over the last week, how often has the person you care for shown the following facial signs?

9. A furrowed brow……………………………………………………………………… 0 1 2 3 NA
10. A change in eyes, including: squinching of eyes, eyes opened wide eyes frowning…… 0 1 2 3 NA
11. Turning down of mouth, not smiling…………………………………………………… 0 1 2 3 NA
12. Lips puckering up, tight, pouting, or quivering……………………………………… 0 1 2 3 NA
13. Clenching or grinding teeth, chewing or thrusting tongue out………………… 0 1 2 3 NA

Over the last week, how often has the person who you care for had a change in activity level in the following way?

14. Not moving, less active, quiet……………………………………………………… 0 1 2 3 NA
15. Jumping around, agitated, fidgety…………………………………………………… 0 1 2 3 NA

Over the last week, how often has the person you care for shown the following limb and body activity?

16. Floppy…………………………………………………………………………………… 0 1 2 3 NA
17. Stiff, spastic, tense, rigid…………………………………………………………… 0 1 2 3 NA
18, Gesturing to or touching part of the body that hurts………………………………… 0 1 2 3 NA
19. Protecting, favouring or guarding part of the body that hurts…………………………… 0 1 2 3 NA
20. Flinching of moving the body part away, being sensitive to touch………………………… 0 1 2 3 NA
21. Moving the body in a specific way to show pain (e.g., head back, arms down, curls up etc.) 0 1 2 3 NA

Over the last week, how often has the person you care for displayed the following physiological behaviours?

22. Shivering…………………………………………………………………………… 0 1 2 3 NA
23. Change in colour, pallor…………………………………………………………… 0 1 2 3 NA
24. Sweating, perspiring……………………………………………………………… 0 1 2 3 NA
25. Tears…………………………………………………………………………….. 0 1 2 3 NA
26. Sharp intake of breath, gasping…………………………………………….. 0 1 2 3 NA
27. Breath holding……………………………………………………………………… 0 1 2 3 NA

Over the last weeks, how often has the person you care for displayed the following behavioural changes?

28. Eating less, not interested in food………………………………………………… 0 1 2 3 NA
29. Increase in sleep…………………………………………………………………… 0 1 2 3 NA
30. Decrease in sleep…………………………………………………………………… 0 1 2 3 NA

APPENDIX P

Non-Communicating Children’s Pain Checklist-Revised

(teacher version)

NCCPC-R

This questionnaire asks how often the person you care for shows signs that they are in pain. Please answer according to how your child has behaved within THE LAST HOURS. Please read each question carefully and circle the number that best describes their behaviour over the last two weeks

0 = not at all 1 = just a little 2 = fairly often 3 = very often NA = not applicable

Over the last two hours, how often has the person you care for made the following vocal sounds?

1. Moaning, whining, whimpering (fairly soft)…………………………… 0 1 2 3 NA
2. Crying (moderately loud)………………………………………………… 0 1 2 3 NA
3. Screaming/yelling (very loud)…………………………………………… 0 1 2 3 NA
4. A specific sound or word for pain (e.g., a word, cry or type of laugh)…… 0 1 2 3 NA

Over the last two hours, how often has the person you care for shown these social behaviours?

5. Not cooperating, cranky, irritable, unhappy……………………………… 0 1 2 3 NA
6. Less interactive with others, withdrawn…………………………………… 0 1 2 3 NA
7. Seeking comfort or physical closeness……………………………………… 0 1 2 3 NA
8. Being difficult to distract, not able to satisfy or pacify…………………… 0 1 2 3 NA

Over the last two hours, how often has the person you care for shown the following facial signs?

9. A furrowed brow…………………………………………………………………… 0 1 2 3 NA
10. A change in eyes, including: squinching of eyes, eyes opened wide eyes frowning…… 0 1 2 3 NA
11. Turning down of mouth, not smiling……………………………………………… 0 1 2 3 NA
12. Lips puckering up, tight, pouting, or quivering………………………………… 0 1 2 3 NA
13. Clenching or grinding teeth, chewing or thrusting tongue out………………… 0 1 2 3 NA

Over the last two hours, how often has the person who you care for had a change in activity level in the following way?

14. Not moving, less active, quiet……………………………………………………… 0 1 2 3 NA
15. Jumping around, agitated, fidgety…………………………………………………… 0 1 2 3 NA

Over the last two hours, how often has the person you care for shown the following limb and body activity?

16. Floppy…………………………………………………………………………………… 0 1 2 3 NA
17. Stiff, spastic, tense, rigid…………………………………………………………… 0 1 2 3 NA
18, Gesturing to or touching part of the body that hurts………………………………… 0 1 2 3 NA
19. Protecting, favouring or guarding part of the body that hurts…………………… 0 1 2 3 NA
20. Flinching of moving the body part away, being sensitive to touch………………… 0 1 2 3 NA
21. Moving the body in a specific way to show pain (e.g., head back, arms down, curls up etc.) 0 1 2 3 NA

Over the last two hours, how often has the person you care for displayed the following physiological behaviours?

22. Shivering………………………………………………………………………………… 0 1 2 3 NA
23. Change in colour, pallor……………………………………………………………… 0 1 2 3 NA
24. Sweating, perspiring…………………………………………………………………… 0 1 2 3 NA
25. Tears…………………………………………………………………………………. 0 1 2 3 NA
26. Sharp intake of breath, gasping…………………………………………. 0 1 2 3 NA
27. Breath holding………………………………………………………………………… 0 1 2 3 NA

Over the last two hours, how often has the person you care for displayed the following behavioural changes?

28. Eating less, not interested in food…………………………………………………… 0 1 2 3 NA
29. Increase in sleep………………………………………………………………………… 0 1 2 3 NA
30. Decrease in sleep………………………………………………………………………… 0 1 2 3 NA

APPENDIX Q

Inter-rater reliability of the NCCPC-R between parent and teacher ratings

		N	Kendall's tau	P
Parent (one week) X teacher (two hours)	Total	34	.14	.28
	Vocalisations	31	.17	.23
	Socialisation	31	.14	.34
	Facial	32	.02	.88
	Activity levels	30	.02	.91
	Body posture	31	.16	.28
	Physiological	31	.02	.92
	Eating and sleeping	24	.02	.91

APPENDIX R

Stability of the NCCPC-R and FLACC observations

		Parent NCCPC-R time one							
	Kendall's taun (N)	Total	Vocal	Social	Facial	Activity	Body	Physiological	Eating and sleeping
Parent NCCPC-R time two	Total	**.61** (38) .	.37 (37)	.39 (38)	.39 (38)	.20 (38)	.20 (38)	.39 (38)	.29 (38)
	Vocal	.35 (37)	**.45** (36)	.34 (37)	.27 (37)	.25 (37)	.21 (37)	.24 (37)	.13 (37)
	Social	.37 (38)	.26 (37)	.33 (38)	.27 (38)	.20 (38)	.11 (38)	.25 (38)	.33 (38)
	Facial	.33 (37)	.18 (36)	.27 (37)	**.43** (37)	.15 (37)	.11 (37)	.25 (37)	.30 (37)
	Activity	.38 (37)	.39 (36)	.35 (37)	.26 (37)	**.46** (37)	.09 (37)	.26 (37)	.09 (37)
	Body	.25 (38)	.15 (37)	.24 (38)	.17 (38)	.01 (38)	.27 (38)	.28 (38)	.15 (38)
	Physiological	37 (38) .	.35 (37)	.36 (38)	.21 (38)	.07 (38)	.27 (38)	**.41** (38)	.16 (38)
	Eating and sleeping.	.28 (38)	.27 (37)	.35 (38)	.19 (38)	.04 (38)	.24 (38)	.21 (38)	.05 (38)

APPENDIX S

Stability of researcher FLACC across the three observations

		Researcher 1 FLACC	
	Kendall’s tau (N)	Observation 2	Observation 3
Researcher 1 FLACC	Observation 1	.30 (29)	.32 (29)
	Observation 2	-------	.39 (29)

APPENDIX T

Challenging Behaviour Interview

Challenging Behaviour Interview

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

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 the category and then giving some examples from the appendix. Check the response by ensuring the month criterion and that the behaviour fulfils the operational 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. For challenging behaviours which are included, read each question whilst the respondent looks at the question and then ask for a rating for each of the behaviour 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 examples)

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 this person now, then would 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[2]?

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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[2] 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 service users?**

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 service users e.g. disruption to planned activities, service users are frightened or upset, belongings or clothing are damaged or lost, meals are spoiled etc.?**

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 individual lives?**

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) or specially designed 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 designed clothing ☐, Other ☐, (please specify) ____)

11. Has the environment in which the individual currently lives been modified because of this behaviour (examples of modification are given 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 reach, furniture is secured, a cupboard door is secured, a door is secured, curtains are absent (because they will be torn down), pictures are out of reach, hard or sharp surfaces are padded, service users are always visible, a room is out of bounds, cutlery is plastic, furniture is deliberately heavy, door closers are fitted to prevent slamming, wallpaper is washable in rooms apart from kitchen and bathroom, fridge is secured, ornaments are out of reach, furniture or fittings have 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. discouraging the behaviour, 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 carers been necessary e.g. 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 behaviour occurred?

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**

APPENDIX U

Challenging Behaviour Interview appendix

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

Behaviour	Description	Examples	Notes
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 include anal-poking but do include poking of other body orifices. Any stereotyped behaviours which fulfil the criteria should be coded here and not under stereotyped behaviours.
Pica (PIC)	Eating inedible substances.	Eating, paper, leaves, discarded food etc.	Must include swallowing the substances. Include eating cigarette ends.
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 include verbal aggression such as threats, insults etc.
Verbal aggression (VAG)	Verbal statements which are likely to induce fear or distress.	Threats, insults etc.	Exclude inappropriate 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 includes temporary disruption of the environment, for example caused by tipping furniture.
Anal poking (AP)	Insertion of objects, fingers or hand into the anus.		

Behaviour	Description	Examples	Notes
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 stereotyped behaviours may result in tissue damage e.g. hair loss from head weaving when in a chair. This should be coded under self-injury.
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 taking food from 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 the vomiting does not have a medical cause such as hiatus hernia.

APPENDIX V

Questionnaire About Behavioural Function (QABF)

THE QABF

Please indicate which type self-injurious (skin picking, head banging etc) or aggressive behaviour (hitting or biting others) that you have been most concerned about in the last month______________________________

Rate how often the person you care for demonstrates the stated behaviour (above) in situations where they might occur. Be sure to rate how often each behaviour occurs, not what you think a good answer would be.

		Does not apply	Never	Rarely	Some	Often
1	Engages in the behaviour to get attention.	X	0	1	2	3
2	Engages in the behaviour to escape work or learning situations.	X	0	1	2	3
3	Engages in the behaviour as a form of ‘self-stimulation’.	X	0	1	2	3
4	Engages in the behaviour because he/she is in pain.	X	0	1	2	3
5	Engages in the behaviour to get access to items such as preferred toys, food, or beverages.	X	0	1	2	3
6	Engages in the behaviour because he/she likes to be reprimanded	X	0	1	2	3
7	Engages in the behaviour when asked to do something (get dressed, brush teeth, work, etc).	X	0	1	2	3
8	Engages in the behaviour even if he/she thinks that no one is in the room.	X	0	1	2	3
9	Engages in the behaviour more frequently when he/she is ill.	X	0	1	2	3
10	Engages in the behaviour when you take something away from him/her	X	0	1	2	3
11	Engages in the behaviour to draw attention to him/herself.	X	0	1	2	3
12	Engages in the behaviour when he/she does not want to do something.	X	0	1	2	3
13	Engages in the behaviour because there is nothing else to do.	X	0	1	2	3
14	Engages in the behaviour when there is something bothering him/her physically.	X	0	1	2	3
15	Engages in the behaviour when you have something he/she wants.	X	0	1	2	3
16	Engages in the behaviour to try and get a reaction from you.	X	0	1	2	3
17	Engages in the behaviour to try to get people to leave him/her alone.	X	0	1	2	3
18	Engages in the behaviour in a highly repetitive manner, ignoring his/her surroundings.	X	0	1	2	3
19	Engages in the behaviour because he/she is physically uncomfortable.	X	0	1	2	3
20	Engages in the behaviour when a peer has something he/she wants.	X	0	1	2	3
21	Does he/she seem to be saying ‘come see me’ or ‘look at me’ when engaging in the behaviour?	X	0	1	2	3
22	Does he/she seem to be saying ‘leave me alone’ or ‘stop asking me to do this’ when engaging in the behaviour?	X	0	1	2	3
23	Does he/she seem to enjoy the behaviour, even if no one is around?	X	0	1	2	3
24	Does the behaviour seem to indicate to you that he/she is not feeling well?	X	0	1	2	3
25	Does he/she seem to be saying ‘give me that (toy item, food item)’ when engaging in the behaviour?	X	0	1	2	3

APPENDIX W

Difference in pain scores between people with functional and non-functional challenging behaviour

		Median (Inter-quartile Range)		U score	P value	Effect size (r)
		Non-functional challenging behaviour	Functional challenging behaviour			
Researcher FLACC		0.00 (0.00-1.00)	0.00 (0.00-3.00)	55.00	.57	.12
Teacher FLACC totals over five days	Total	9.50 (5.00-13.50)	6.00 (1.75-7.25)	27.50	**.05**	.42
	face subscale	1.50 (1.00-2.75)	1.50 (0.00-2.13)	49.00	.62	.10
	legs subscale	1.50 (1.00-4.25)	0.00 (0.00-1.13)	25.00	**<.05**	.47
	activity subscale	1.00 (.25-2.00)	0.00 (0.00-1.63)	40.50	.26	.24
	cry subscale	2.00 (1.25-3.00)	1.00 (0.00-2.25)	34.00	.12	.33
	Consolability subscale	1.50 (.25-2.75)	0.00 (.00-1.00)	28.00	**<.05**	.44
Teacher NCCPC	total	6.00 (2.50-14.50)	5.00 (3.50-9.50)	31.50	.92	.02
	vocal subscale	0.00 (0.00-3.00)	1.00 (.50-2.00)	22.50	.30	.25
	social subscale	3.00 (.50-6.25)	2.00 (1.00-2.00)	18.50	.38	.21
	facial subscale	1.00 (0.00-2.50)	2.00 (0.00-3.50)	26.50	.54	.14
	activity subscale	1.00 (0.00-2.50)	0.00 (0.00-1.50)	28.50	.67	.10
	body subscale	0.00 (0.00-2.00)	0.00 (0.00-1.00)	31.50	.91	.03
	physiological subscale	1.00 (0.00-1.50)	0.00 (0.00-.50)	20.50	.16	.33
	eat/sleep subscale	0.00 (0.00-0.00)	0.00 (0.00-0.00)	15.00	.60	.14
GDQ	number of items scoring past the cut-off	5.00 (3.50-7.00)	5.00 (4.50-6.25)	60.00	.85	.04

APPENDIX X

D-Stat results for all forms of challenging behaviour

	Attention maintained				Demand escape			
	Aggression	Self injury	Destruction of property	Overall challenging behaviour	Aggression	Self injury	Destruction of property	Overall challenging behaviour
Participant 1	0.25	0.06	n/a	0.22	0.00	0.16	n/a	0.16
Participant 2	-0.22	-0.09	0.00	-0.13	-0.38	0.44	-0.25	0.19
Participant 3	-0.13	-0.25	-0.13	-0.25	0.09	-0.06	-0.13	0.00

APPENDIX Y

Expressive communication subscale of the Vineland Adaptive Behavior Scale