Atypicalities of Social Functioning in Children with Williams syndrome

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Author’s declaration

This thesis is the result of the author’s original research. It has been composed by the author and has not been previously submitted for examination which has led to the award of a degree.

Signed: Amanda Gillooly

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Abstract

Children with Williams syndrome (WS) are often reported to show a hyper-social phenotype which can pose implications for their interactions with strangers and peers. The present thesis aimed to obtain an in depth examination of two key areas of social difficulty in children with WS: social approach behaviour (SAB) and peer relationships. A multi-informant approach was used in order to examine the nature and stability of these atypicalities across environmental settings and from different perspectives.

Within the present thesis, five studies are reported. Within chapters 3, 4 and 5, SAB was examined in children with WS (aged 7-16 years) by administering an experimental rating task (Adolph's Approachability Task), a parent questionnaire and semi-structured interview. The children with WS indicated significantly higher levels of SAB in Adolph's Approachability Task than typically developing (TD) children matched on verbal ability and TD children matched on non-verbal ability. These high levels of SAB were reported by parents to have substantial implications for the children's vulnerability and family life. However, there was within-syndrome variance in the manifestation of this SAB. Impairment in social functioning, specifically deficits within social cognition and social awareness, were significantly associated with levels of SAB in the children with WS.

Within chapters 6 and 7, questionnaires and interviews were used to solicit the perceptions of children with WS, their parents and teachers regarding the child's peer relationships. Parents and teachers reported that many of the children with WS had difficulties forming and sustaining friendships and demonstrated atypical patterns of behaviour during social interactions. However, these peer relationship difficulties were not reported by the children themselves. Peer relationship difficulties were found to be significantly associated with the children's deficits within social communication, social cognition and social awareness.

The findings from the current thesis provide insight into substantial social difficulties among many children with WS. Importantly, and in line with the heterogeneous nature of the disorder, there was within-syndrome variance in the children's social functioning. The present findings have important implications for both research and practice. These implications will be discussed, with suggestions for future research.

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Chapter 1: General introduction

1.1. Overview of the present thesis

Children with Williams syndrome (WS) are often reported to display a hyper-social phenotype and a strong drive towards social interactions (Jawaid, Riby, Owens, White, Tarar, & Schulz, 2012). Many children with WS show atypicalities within social functioning (Klein-Tasman, Li-Barber, & Magargee, 2011) which pose implications for their vulnerability around strangers (Fisher, 2014; Jawaid et al., 2012). Children with WS also show substantial difficulties forming and sustaining peer relationships (Greer, Brown, Pai, Choudry, & Klein, 1997). The present thesis aims to examine the characteristics of these social difficulties, their stability across environmental settings and the role of underlying social and cognitive functions in these social difficulties. The current thesis will seek to achieve this by using multiple measures, methods and informants.

1.2. What is Williams syndrome?

WS is a complex genetic disorder which stems from the deletion of approximately 25 to 28 genes from chromosome band 7q.11.23 (Haas, Barnea-Goraly, Sheau, Yamagata, Ullas, & Reiss, 2013). WS has been estimated to affect between 1 in 7,500 and 1 in 20,000 individuals worldwide (Sparaci, Stefanini, Marotta, Vicari, & Rizzolatti, 2012). Individuals with WS demonstrate a mild to moderate level of impairment to intellectual functioning (Bellugi, Adolphs, Cassady,

& Chiles, 1999; Greer et al., 1997), a unique profile of cognitive and executive function (EF) impairments (Carney, Brown, & Henry, 2013; Rhodes, Riby, Park, Fraser, & Campbell, 2012) and an atypical social phenotype (Bellugi, Järvinen-Pasley, Doyle, Reilly, Reiss, & Korenberg, 2007; Jawaid et al., 2012). WS is characterised by atypicalities across behaviour and cognition; however, as with many neurodevelopmental disorders, there is substantial within-syndrome variance in the manifestation and severity of these atypicalities (Little, Riby, Janes, Clark, Fleck & Rodgers, 2013; Lough, Rodgers, Janes, Little, & Riby, 2016; Porter & Coltheart, 2005). The nature of these atypicalities will be discussed in further depth throughout this chapter.

1.2.1. Diagnosis of Williams syndrome

The earliest insight into WS was provided by teams of cardiologists (Beuren, Apitz, & Harmjanz, 1962; Williams, Barratt-Boyes, & Lowe, 1961) who recognised an association between cardiac abnormalities, distinct facial features and evident cognitive and intellectual impairments among their patients. Through subsequent investigation, further atypicalities were reported among these patients which included a disparity between verbal and visuo-spatial abilities and an unusually high sociability and drive to approach strangers (Donnai & Karmiloff-Smith, 2000). These discoveries led to the recognition of a genetic disorder which has been termed Williams syndrome.

While previous diagnoses of WS were confirmed by clinical observation, in more recent years, and with advances in technology, there has been an increase in the use of genetic tests as a diagnostic tool. The fluorescence in situ hybridization test (FISH test) is a simple blood test which was developed in the early 1990's and used in the diagnosis of WS. The FISH test detects the absence of the elastin gene (Apps, Newby, & Roberts, 2008) which is absent in an estimated 98% of individuals with WS (Järvinen-Pasley, Bellugi, Reilly, Mills, Galaburda, Reiss, & Korenberg, 2008). The elastin gene is a protein and plays a critical role in the formation of connective tissue within the aorta blood vessel. This genetic deletion can cause narrowing of the blood vessels, increasing the risk of cardiac abnormalities among individuals with WS (Järvinen-Pasley et al., 2008).

1.2.2. Cognitive strengths and weaknesses

WS is characterised by an unusual pattern of cognitive strengths and weaknesses. Accumulated findings support a clear dissociation in performance between verbal and visuospatial domains in WS (Atkinson, Braddick, Anker, Curran, Andrew, & Wattam-Bell, 2003; Bellugi, Korenberg, & Klima, 2001; Järvinen-Pasley et al., 2008; Menghini, Addona, Costanzo, & Vicari, 2010; Mervis & John, 2010). Jones, Hickok, Rossen and Bellugi (1998) used a cross-sectional design to examine verbal and visuospatial skills in WS. Adolescents and adults with WS performed at a similar level to their typically developing (TD) peers in a vocabulary assessment; however, individuals with WS performed significantly poorer than TD peers in a visuospatial task which required them to replicate a drawing. This dissociation between verbal and visuospatial performance has been attributed to patterns of neuropsychological functioning (Meyer-Lindenberg, Kohn, Mervis, Kippenhan, Olsen, Morris, & Berman, 2004).

Neuroimaging research indicates atypical functioning of the dorsal stream in WS. The dorsal stream plays an integral role in the processing of motion stimuli and visually guides actions. This neural impairment is suggested to account for the visuospatial deficits which are widely reported in WS (Akinson, King, Braddick, Nokes, Anker & Braddick, 1997; Atkinson et al., 2003; Meyer-Lindenberg et al., 2004). However, verbal skills are not completely spared in WS, with more recent findings revealing substantial impairments in social communication and pragmatic language in WS (Laws & Bishop, 2004). This will be discussed in greater depth later in this chapter.

1.2.3. Atypical social profile

WS is characterised by an atypical social profile. Individuals with WS can show a predisposition to approach other people, regardless of their level of familiarity (Jawaid et al., 2012) and an absence of fear towards strangers (Fisher, 2014; Jawaid et al., 2012). Despite a strong motivation to approach new people, individuals with WS have been reported to demonstrate atypicalities within social communication and reciprocal social interactions; ultimately leading to their difficulties establishing peer relationships (Bellugi et al., 2007; Davies, Udwin, & Howlin, 1998; Elison, Stinton, & Howlin, 2010).

This thesis will focus on two specific areas of social difficulty among children with WS: SAB towards unfamiliar people and peer relationships. These social difficulties have been identified as key features of the WS behavioural phenotype (Greer et al., 1997; Jawaid et al., 2012). However, as will be discussed below, there is a need to conduct further research to examine the characteristics of these difficulties and the role of psychological variables in these atypicalities of social functioning.

1.3. Social approach behaviour in Williams syndrome

Children with WS are often reported to display little or no fear around strangers (Fisher, 2014) and to approach individuals indiscriminately (Jawaid et al., 2012). From as young as infancy, individuals with WS demonstrate prolonged gaze towards strangers (Mervis, Morris, Klein-Tasman, Bertrand, Kwitny, Appelbaum, & Rice, 2003; Jones, Bellugi, Lai, Chiles, Lincoln, & Adolphs, 2000) and fewer signs of distress when separated from their primary caregiver when compared to TD children matched on mental age and chronological age (Jones et al., 2000).

A study was conducted by Riby, Kirk, Hanley and Riby (2013) which examined stranger danger awareness in a sample of children with WS. The children were shown several video vignettes, each of which depicted a social scenario. In each scenario, a stranger approached a child and asked them for help. After viewing each video clip, a series of questions were posed to the children. The children with WS were asked if the child in the scenario should help the stranger and if they would help the stranger if they were in that situation. The children were asked to provide justifications for their decisions. The children with WS demonstrated a significantly poorer awareness of social danger and indicated a significantly higher intention to approach the stranger than did TD controls. These findings reinforce the social vulnerability of children with WS with regards to stranger danger.

Several methodologies have been used to examine levels of SAB in WS. These include experimental tasks, parental questionnaires and behavioural observations (Dodd, Porter, Peters, & Rapee, 2010; Doyle, Bellugi, Korenberg, & Graham, 2004; Järvinen-Pasley et al., 2010). The accumulated findings support the hypothesis of high levels of indiscriminate friendliness in children and adults with WS (Järvinen-Pasley et al., 2010). These findings will be evaluated, with consideration to the strengths and limitations of each methodology.

1.3.1. Experimental measures of social approach in Williams syndrome

Adolph's Approachability Task (Adolphs, Tranel, & Damasio, 1998) is a standardised experimental task which has been widely used as an assessment of SAB during typical and atypical development (Järvinen-Pasley et al., 2010; Semel & Rosner, 2003). Individuals are presented with an array of black and white images of unfamiliar faces. The participants are instructed to rate the extent to which they would like to approach the depicted person. Researchers have reported that individuals with WS demonstrate a heightened readiness to approach the unfamiliar people within this task than TD individuals (Bellugi et al., 1999; Järvinen-Pasley et al., 2010; Martens, Wilson, Dudgeon, & Reutens, 2009) despite being able to rank the approachability of the faces as accurately as TD individuals (Bellugi et al., 1999). However, contradictory findings have been reported. Frigerio, Burt, Gagliardi, Cioffi, Martelli, Perrett and Borgatti (2006) found that individuals with WS indicated a significantly higher desire to approach the person depicted in the happy faces and a significantly lower desire to approach the person depicted in the negative faces than TD matched controls. This disparity in findings could be suggested to be due to a difference in the nature of the face stimuli used across these studies. Frigerio et al. used face stimuli which depicted six specific emotional expressions in contrast to earlier studies which used a neutral set of face stimuli which had been pre-rated into the categories ‘least approachable’ and ‘most approachable’ (Bellugi et al., 1999; Jarvinen-Pasley et al., 2010; Martens et al., 2009).

Porter, Coltheart and Langdon (2007) controlled for an effect of emotion recognition performance on the participants’ ratings in Adolph’s Approachability Task in an attempt to resolve this disparity in findings. The individuals with WS were required to label the emotion depicted within each face stimulus. The participants’ social approach ratings were restricted to the faces which had been correctly identified. This controlled for any effect of emotion recognition performance on the participants’ levels of SAB. Porter et al. reported that when emotion recognition performance was controlled for, individuals with WS indicated a significantly higher desire to approach the person depicted in the positive faces, but not the negative faces, than TD individuals who were matched on mental age. These findings suggest that individuals with WS are particularly driven to approach and engage with people depicted by positive expressions.

Research using Adolph’s Approachability Task has generated strong support for claims of high levels of SAB in WS (Bellugi et al., 1999; Järvinen-Pasley et al., 2010, Martens et al, 2009). Despite this, researchers have raised concerns regarding the validity of this task. Performance in Adolph's Approachability Task has been found to be poorly correlated with parent ratings of SAB (Fisher, Mello, & Dykens, 2014; Järvinen-Pasley et al., 2010) and behavioural observations of SAB (Fisher, Mello, & Dykens, 2014). Adolph's Approachability Task uses static images of faces. This raises concerns regarding the extent to which this task provides a valid measurement of the children's social interactions in a real-life social setting.

1.3.2. Parent report measures of social approach in WS

Parent ratings have been reported to be strongly correlated with behavioural observations of SAB in WS (Fisher et al., 2014) and may provide a more valid measurement of SAB. The 'Salk Institute Sociability Questionnaire' (SISQ) is a standardised parent report questionnaire designed to measure sociability among children with WS. This scale obtains a measure of the child's level of SAB towards familiar and unfamiliar people (Järvinen-Pasley et al., 2010). Across studies, individuals with WS were rated significantly higher in the SISQ than TD individuals who were matched on mental age (Doyle et al., 2004; Järvinen-Pasley et al., 2010) and other developmental disorder groups (Järvinen-Pasley et al., 2010). A limitation of these studies is that the analysis performed provided little opportunity to examine for within-syndrome variance in SAB. In light of the recent findings which support the heterogeneous nature of WS (Little et al., 2013; Porter & Coltheart, 2005), the present thesis will build on these research findings by examining in more depth for within-syndrome variance in the prevalence and manifestation of SAB among children with WS.

The majority of research thus far has adopted a quantitative approach to measure SAB in WS. Lough et al. (2016) were the first researchers to use a qualitative method to obtain a greater depth of insight into the SAB of children with WS. Lough et al. interviewed the parents of a sample of children with WS regarding their child's SAB. The interview data were subsequently analysed using the qualitative method of thematic analysis. Using thematic analysis, themes are identified from the data which are interpreted as a direct measure of the participants' thoughts, attitudes and motivations (Braun & Clark, 2006). The children with WS were reported by their parents to show disinhibited behaviour towards strangers and lacked an awareness of socially appropriate behaviour. Parents noted that the children with WS often sought physical contact and affection from strangers and were overbearing or inappropriate within conversations. The children with WS were reported to lack an awareness of danger, in particular regarding their interactions with strangers. However, there was heterogeneity in the experiences of SAB within this group. A qualitative analysis of the data extracts indicated that the children's levels of impulsivity, and family variables, such as levels of parental supervision, were reported to be associated with the levels of SAB demonstrated. Lough et al. were the first researchers to use a qualitative methodology to examine SAB in children with WS. The present thesis will build on these findings by soliciting further information on the manifestation of the children's SAB and the impact of this behaviour on the family. In light of previous findings of poor personal space regulation and disinhibited SAB in WS

(Little et al., 2013; Lough et al., 2016), further information will be sought on the nature of this disinhibited behaviour.

Quantitative and qualitative analysis can each provide a valuable insight when measuring social behaviour in neurodevelopmental disorders. The present thesis will build on previous research by using a mixed methods approach. Both quantitative and qualitative measures of SAB will be solicited from the parents of the children with WS. This will provide a greater depth of insight into the characteristics of SAB among children with WS and allow an examination of the associations between social functioning and SAB. Together these findings are believed to be informative in designing appropriate support for the children with WS and their families.

1.3.3. Behavioural observations of social approach in Williams syndrome

Behavioural observations allow a direct assessment of the social behaviour under investigation. Dodd, Porter, Peters and Rapee (2010) were the first researchers to use a behavioural observation method to measure SAB in WS. Pre-school children with WS were invited to the research laboratory which had been designed to resemble a playroom. Actors who played the role of a stranger entered the playroom and the child’s behaviour and interactions with this stranger were recorded. These behaviours included speaking to the stranger, playing with the stranger, waving at the stranger or touching the stranger. The children with WS demonstrated significantly higher levels of SAB towards the stranger than TD controls, and often without any prompts from the stranger. This study provides behavioural evidence of a willingness to approach unfamiliar people in WS. However, a limitation of this study was that it was conducted in a research laboratory and may therefore not reflect SAB in a real-life environment. Caution is required when interpreting the findings.

A study was conducted which investigated SAB in WS in a real-life social situation. Fisher (2014) examined stranger interactions in a sample of adults with WS who were attending a residential summer camp. Using an experimental paradigm, confederate actors were assigned the role of a stranger. The stranger approached the participant and invited this individual to follow them. In some situations the stranger provided an incentive, such as to go and get ice cream. The individuals' interactions with the stranger were observed and coded. Many of the adults with WS demonstrated a willingness to follow the stranger and a lack of awareness of the dangers associated with this behaviour. This was most prominent when the stranger offered an incentive. However, there was heterogeneity within this sample, where not all of the adults with WS followed the stranger. A limitation of this study was that Fisher did not examine stranger interactions in a TD control sample. It is therefore difficult to draw reliable conclusions on the atypical nature of stranger interactions among the adults with WS.

Previous research has predominantly used a single measurement of SAB when assessing the social profile of individuals with WS (Doyle et al., 2004; Porter et al., 2007). The literature presented in section 1.3 has highlighted the strengths and limitations of each methodology and informant. In order to construct an in depth examination of the atypical social profile of children with WS, it is beneficial to adopt a multi-measure and multi-informant approach. The present thesis will build on previous research by soliciting children's ratings of SAB within Adolph's

Approachability Task and parents' ratings of their child's SAB by administering a newly developed questionnaire.

1.3.4. Theoretical perspectives on social approach behaviour in Williams syndrome

Accumulated findings have provided support for a hyper-sociability phenotype and strong drive to approach strangers among individuals with WS (Bellugi et al., 1999; Doyle et al., 2004; Järvinen-Pasley et al., 2010; Martens et al., 2009). Several theoretical models have been proposed to account for high levels of SAB in WS. Three theoretical accounts will be evaluated, with reference to both neurological and experimental research evidence. While the present thesis is not designed to directly test and compare and contrast these theoretical accounts, these theoretical viewpoints are informative in guiding areas for examination in the current thesis.

1.3.4.1. The frontal lobe hypothesis

The frontal lobe hypothesis proposes that the high levels of SAB demonstrated by individuals with WS are caused by EF impairments which occur as a result of impairment to the frontal lobes (Little et al., 2013). It is important to acquire an understanding of the development of EF's in typical and atypically developing populations in order to draw stronger conclusions regarding the degree of cognitive impairment in WS and the role of cognition in the atypical social profile demonstrated by individuals with WS.

1.3.4.1.1. Executive dysfunction in neurodevelopmental disorders

EF’s are higher order cognitive functions which are performed in the attainment of a specific goal (Willcutt, Doyle, Nigg, Faraone, & Pennington, 2005). Many definitions of EF’s have been proposed which differ in the range of aspects of cognitive processes included. The most popular theoretical accounts propose three core components of EF to play a crucial role in an individual’s daily functioning. These are response inhibition, updating (working memory) and attention set-shifting (Miyake, Friedman, Emerson, Witzki, Howerter, & Wagner, 2000). Planning has more recently been implicated as a core component of EF (Diamond, 2013).

Executive dysfunction is prevalent within neurodevelopmental disorders (Gau & Shang, 2010; Happē, Booth, Charlton, & Hughes, 2006; Lee, Fidler, Blakeley-Smith, Daunhauer, Robinson, & Hepburn, 2011). Accumulated research findings support claims of substantial EF impairments in neurodevelopmental disorders which have a known aetiology, for example Down’s syndrome (DS) and fragile X syndrome, and unknown aetiology, for example Autism spectrum disorder (ASD) and Attention Deficit-Hyperactivity Disorder (ADHD) (Castellanos, Sonuga-Barke, Milham, & Tannock, 2006; Costanzo, Varuzza, Menghini, Addona, Gianesini, & Vicari, 2013; Gilotty, Kenworthy, Sirian, Black, & Wagner, 2002; Lanfranchi, Jerman, Dal Pont, Alberti, & Vianello, 2010; Rhodes et al., 2012).

1.3.4.1.2. Executive dysfunction in Williams syndrome

EF’s have been reported to be impaired in individuals with WS (Carney et al., 2013; Lincoln, Lai, & Jones, 2002; Rhodes, Riby, Park, Fraser, & Campbell, 2010). Individuals with WS demonstrate impairments on response inhibition tasks which require them to supress a response, whether that is a verbal or motor response (Carney et al., 2013; Porter et al., 2007). Carney et al. (2013) reported that individuals with WS took significantly longer to generate the appropriate motor response and made significantly greater errors on a verbal inhibition task than TD individuals. Individuals with WS demonstrate impairments on attention set-shifting tasks which require them to transfer attentional resources between different components of a task (Cornish, Scerif, & Karmiloff-Smith, 2007; Lincoln et al., 2002). Rhodes et al. (2010) reported that individuals with WS took significantly longer to shift their attention from one stimulus to another than TD controls who were matched on mental age and chronological age. Individuals with WS have also been found to make significantly more errors than TD controls on working memory tasks which involve simultaneously storing and manipulating information within memory (Carney et al., 2013; Rhodes et al., 2010). Finally, individuals with WS demonstrate difficulties on planning tasks which involve formulating a series of actions in order to achieve a specific end goal, where individuals with WS generated significantly more moves to solve the problem than TD controls matched on mental age and chronological age (Rhodes et al., 2010).

A research study conducted by Rhodes et al. (2010) assessed EF performance in a sample of children and adults with WS. Rhodes et al. administered a standardised testing battery to assess EF performance. The parents of the individuals with WS also completed the standardised Strengths and Difficulties Questionnaire (SDQ) (Goodman, 1997) which provided an assessment of the individual's social behaviour. Individuals with WS demonstrated significantly poorer performance across measures of attention set-shifting, working memory and planning than TD individuals who were matched on mental age and chronological age, who were able to successfully pass these tasks. Furthermore, several relationships were identified between EF performance and social behaviour in this WS sample. Impairments in attention set-shifting were positively correlated with conduct problems and emotional difficulties, and negatively correlated with prosocial behaviour. In addition, impairments on the spatial working memory task were positively correlated with the individuals' total difficulties score within the SDQ. Furthermore, less effective planning strategies were significantly associated with levels of inattention, emotional difficulties and conduct behaviour problems. These findings implicate the role for EF in the wider behavioural difficulties of individuals with WS. Little research thus far has examined the role of EF in the atypical social profile in WS. This will be investigated further in the current thesis.

1.3.4.1.3. Response inhibition deficits in Williams syndrome

During typical development, infants acquire simple inhibition skills within the first year of life (Garon, Bryson, & Smith, 2008) where they are able to inhibit their actions in response to a caregiver's request (Kochanska & Aksan, 1995). These basic response inhibition skills further develop across infancy (Benson & Haith, 2009). There is a significant increase in the complexity of response inhibition performance from 3-4 years of age. At this stage, children are able to pass more advanced tasks such as Luria’s hand game (Hughes, 1998) and the Day Night Stroop task (Gerstadt, Hong, & Diamond, 1993). These complex inhibition tasks have an integral working memory component. The child is required to inhibit their automatic response and produce a conflicting response. The level of working memory demands within a task have been found to impact on the age at which a child is able to successfully complete it, with performance in some tasks continuing to develop into middle childhood and adolescence (Brocki & Bohlin, 2004; Carlson, 2005; Cragg & Nation, 2008; Jonkman, Lansbergen, & Stauder, 2003; Williams, Ponesse, Schachar, Logan, & Tannock, 1999).

Children with WS demonstrate substantial response inhibition deficits when compared to TD children and other developmental disorder groups (Carney et al., 2013; Porter et al., 2007). Carney et al. (2013) measured response inhibition by administering a verbal and motor inhibition task to a sample of children with WS. Within each task, the children were presented with a stimulus and were required to produce an opposing response. In the verbal inhibition task the children were asked to say the word ‘doll’ when presented with the image of a car and say the word ‘car’ when presented with the image of a doll. For the motor inhibition task, two different actions were presented to child, a finger point and a fist clench. The children were required to produce the opposite motor response to the action presented to them. The children with WS demonstrated significantly greater impairments in the verbal and visuo-spatial inhibition tasks than TD children and a sample of children with DS.

Neuro-imaging research has further supported the frontal lobe hypothesis. Individuals with WS often demonstrate reduced activation within the dorsal lateral prefrontal cortex and the dorsal anterior cingulate cortex, with both of these brain structures believed to be critical in response inhibition (Mobbs, Eckert, Mills, Korenberg, Bellugi, Galaburda, & Reiss, 2007). The accumulated experimental and neural findings provide support for response inhibition as an area of substantial difficulty for many individuals with WS.

Research findings indicated that individuals with frontal lobe brain damage demonstrate a similar social profile to individuals with WS; specifically they both engage in uninhibited behaviour towards strangers (Bruce, Tarullo, & Gunnar, 2009; Spurgeon, Davies & Chapman, 1994). These individuals know that they should not approach strangers; however, are unable to inhibit their impulsive response to engage in these interactions. The frontal lobe hypothesis suggests that individuals with WS demonstrate impairments in response inhibition which are caused by underlying deficits to the frontal lobes. This EF deficit is suggested to lead to uninhibited behaviour towards strangers (Little et al., 2013).

The consistency of these research findings across methodological designs (Carney et al., 2013; Mobbs et al., 2007; Porter et al., 2007) strengthens the frontal lobe hypothesis as an account for high levels of SAB in WS. Furthermore, researchers reported that individuals with frontal lobe brain damage displayed a similar social profile to individuals with WS, where both groups were reported to engage in high levels of indiscriminate friendliness (Bruce et al., 2009; Spurgeon et al., 1994). This further supports the role of executive dysfunction in an engagement in high levels of SAB, in line with the assumptions of the frontal lobe hypothesis.

However, response inhibition impairments have been reported in other neurodevelopmental disorders such as ADHD (Willcutt et al., 2005) and ASD (Uzefovsky, Smith, & Baron-Cohen, 2016). While individuals with ADHD and ASD show substantial response inhibition deficits, these individuals do not show high levels of SAB which are characteristic of many individuals with WS (Bellugi et al., 1999). This challenges the strength of the frontal lobe hypothesis as a unitary account of SAB in WS.

1.3.4.2. The amygdala hypothesis

An alternative theory which has been proposed to account for the high levels of SAB demonstrated by individuals with WS is the amygdala hypothesis. The amygdala hypothesis proposes that heightened sociability in WS is a result of abnormalities in the functioning of the amygdala (Porter et al., 2007). The amygdala is a brain structure within the temporal lobes. The amygdala is responsible for the identification of emotional expressions and is essential in the detection of threat (Adolphs, Gosselin, Buchanan, Tranel, Schyns & Damasio, 2005; Winston, Strange, O’Doherty, & Dolan, 2002). Individuals who suffer damage to this brain structure have been reported to experience severe difficulties in the detection of emotions, particularly in the identification of the negative facial expressions of fear, anger and sadness (Adolphs & Tranel, 2004; Herba & Phillips, 2004).

1.3.4.2.1. Atypical functioning of the amygdala in Williams syndrome

Research using functional magnetic imaging has supported the hypothesis of atypical functioning of the amygdala in WS. Individuals with WS demonstrated increased activation of the amygdala in response to positive facial expressions and reduced activation of the amygdala in response to negative facial expressions when compared to TD controls (Haas, Mills, Yam, Hoeft, Bellugi, & Reiss, 2009; Meyer-Lindenberg, Hariri, Munoz, Mervis, Morris, & Berman, 2005). This suggests that individuals with WS process faces differently from TD individuals, where individuals with WS engage in greater processing of positive facial expressions and reduced processing of negative facial expressions. As a consequence, they may be less likely to detect threat in their social environment and may be at greater risk of social vulnerability.

In a similar vein, research has demonstrated that individuals with bilateral amygdala damage indicated a significantly higher level of SAB towards negative faces than TD controls (Adolphs et al., 1998), similar to individuals with WS (Martens et al., 2009). Furthermore, Martens et al. (2009) reported a significant relationship between increased amygdala volume in WS as measured using functional magnetic imaging techniques and higher levels of SAB.

Research findings have provided support for emotion recognition deficits in WS. Plesa-Skwerer, Faja, Schofield, Verbalis and Tager-Flusberg (2006) administered the ‘Diagnostic analysis of non-verbal accuracy scale’ to a sample of adolescents and adults with WS. The participants were presented with images of faces and were required to identify the basic emotion being portrayed. Plesa-Skwerer et al. reported that individuals with WS performed significantly poorer than TD controls when labelling all emotions, particularly negative emotions. These findings of emotion recognition impairments in WS provide support for the amygdala hypothesis as an account for a hyper-social phenotype in WS. However, Capitão, Sampaio, Férnandez, Sousa, Pinheiro, & Gonçalves (2011) reported that while individuals with WS performed poorer than TD controls in their recognition of several negative emotional expressions, they performed comparable to TD individuals in their detection of angry facial expressions. Furthermore, the individuals with WS showed the normal rank order of approachability. They rated the positive faces as more approachable than negative faces which indicates an awareness that some individuals are more approachable than others. These findings challenge the amygdala hypothesis in its assumption that indiscriminate SAB in WS occurs due to the individual's difficulties detecting threat within their social environment.

Findings have been generated across experimental and neuroimaging studies which highlight atypical functioning of the amygdala and difficulties interpreting emotional expressions in individuals with WS. Furthermore, research indicated that individuals with bilateral amygdala damage, similar to individuals with WS, demonstrate higher levels of SAB (Adolphs et al., 1998). However, there are some inconsistent findings within the literature with regards to the emotion recognition impairments in WS. While individuals with WS showed impairments in their identification of negative facial expressions, they performed comparable to TD controls in their recognition of anger (Capitão et al., 2011). In addition, researchers reported that individuals with WS showed the normal rank order of approachability, where they indicated a higher intention to approach positive faces than negative faces (Bellugi et al., 1999; Jones et al., 2000). These inconsistencies within research findings challenge the strength of the amygdala hypothesis as an account for SAB in WS.

1.3.4.3. The social salience hypothesis

The social salience hypothesis proposes that individuals with WS are driven to approach and engage with other people due to an atypically high level of interest in social stimuli, specifically positive stimuli (Frigerio et al., 2006; Porter et al., 2007). This has been termed a ‘social stimulus attraction’ (Frigerio et al., 2006). The atypical social salience of face stimuli among individuals with WS has been demonstrated across eye-tracking studies, where individuals with WS were shown to attend to the person within the social scene for significantly longer than TD controls matched on mental age and chronological age (Riby & Hancock, 2008; Riby & Hancock, 2009).

Through behavioural observations, Jones et al. (2000) reported that infants with WS spent significantly longer looking at the researcher across a range of cognitive tasks and activities when compared with TD infants who were matched on verbal ability. Furthermore, the intensity of the infant’s gaze towards the researcher often interfered with the infant’s performance within the cognitive tasks. Mervis et al. (2003) examined infants’ gaze behaviours during medical assessments with a clinician. The infants with WS demonstrated significantly greater visual attention towards the clinician than infants with a range of other developmental disorders. A case study was also conducted which examined the behaviour of an infant with WS during play interactions with his mother and a stranger. The infant with WS attended to the face of the mother and the stranger significantly more than TD controls who were matched on chronological age. Furthermore, the intensity of gaze towards the stranger was significantly greater among the infants with WS when compared to TD controls matched for both chronological age and developmental age. These studies support the atypical nature of attention allocation among infants with WS when compared to TD controls. These findings also demonstrate an attentional bias in WS towards faces. These atypicalities within attention mechanisms may account for the hyper-social phenotype in WS and drive to approach and engage with strangers (Mervis et al., 2003; Riby & Hancock, 2008, Riby & Hancock, 2009).

Both experimental and neural findings support the hypothesis of a positivity bias within visual perception among individuals with WS (Dodd & Porter, 2010; Hass et al., 2009; Meyer-Linderberg et al., 2005). Hass et al. (2009) measured levels of brain activation in individuals with WS as they viewed faces which depicted different emotional expressions. Using functional MRI scan techniques, individuals with WS demonstrated significantly higher levels of amygdala activation when viewing positive over neutral faces, and significantly lower levels of amygdala activation when viewing negative over neutral faces. These patterns of neural activation were not present in TD individuals. This positivity bias was further demonstrated by Dodd and Porter (2010) who examined for a bias in visual perception among adolescents and adults with WS. The individuals completed a dot probe task. In each trial of this task, two faces were presented parallel to each other on the computer monitor. These faces differed in their emotional valence, with happy, angry and neutral expressions depicted across the images. After a time interval, the faces disappeared and a single dot was presented on the screen in one of these two locations. Dodd and Porter reported that the individuals with WS were significantly faster at detecting the dot when it was presented in the same location as the positive face in comparison to the negative face. This pattern was not demonstrated by the TD comparison groups. The accumulated findings suggest a perceptual bias towards positive social stimuli in WS.

Frigerio et al. (2006) set out to test the theoretical assumptions of the social salience hypothesis. Frigerio et al. administered Adolph's Approachability Task to a sample of children and young adults with WS and TD controls who were matched on mental age and chronological age. The individuals were presented with images of faces which depicted a range of emotional expressions. Frigerio et al. reported that the individuals with WS indicated a higher intention to approach the person depicted by a happy face than did the TD controls. Furthermore, the individuals with WS reported a significantly lower desire to approach the person depicted by a negative face than did the TD controls. These findings provide support for the theoretical assumptions of the social salience hypothesis, where the individuals with WS showed discrimination in their SAB and their responses were driven by the positive valence of the faces presented. The accumulated findings have generated support for a hypothesis of a strong social interest, and in particular a drive towards positive social stimuli among individuals with WS. These findings were generated across neural and experimental studies and have provided support for the theoretical assumptions of the social salience hypothesis.

1.3.4.4. Comparing and contrasting theoretical accounts of social approach in Williams syndrome

Three key theories have been proposed to account for the high levels of SAB demonstrated by individuals with WS: the frontal lobe hypothesis, the amygdala hypothesis and the social salience hypothesis. Experimental and neural evidence have provided support for these theoretical accounts; however, there are evident inconsistencies within the literature. While individuals with WS were impaired in emotion recognition, these individuals demonstrated the normal rank order of approachability where they rated trustworthy faces as significantly more approachable than untrustworthy faces (Bellugi et al., 1999; Capitão et al., 2011; Jones et al., 2000). These findings are argued to challenge the theoretical assumptions of the amygdala hypothesis which proposes that the high levels of SAB in WS are a result of the individual’s impairments perceiving emotional expressions such as threat. The accumulated research findings in support of the frontal lobe hypothesis appear to be more robust with experimental and neural findings which have demonstrated substantial deficits in response inhibition performance in WS (Carney et al., 2013; Porter et al., 2007). Furthermore, individuals with frontal lobe brain damage have been reported to demonstrate a similar social profile to children with WS, in particular, an engagement in high levels of indiscriminate friendliness (Bruce et al., 2009; Spurgeon et al., 1994). However, response inhibition impairments have been reported across neurodevelopmental disorders that do not all demonstrate the high levels of SAB which are shown in WS (Uzefovsky et al., 2016; Willcutt et al., 2005). Finally research has indicated a strong drive towards social stimuli among individuals with WS, specifically positive stimuli. This social drive was illustrated across tasks, where individuals with WS showed the normal rank order of approachability, indicating a stronger interest in approaching the person depicted by positive faces (Frigerio et al., 2006).

Only one study thus far has directly tested the frontal lobe hypothesis and amygdala hypothesis as an explanation for heightened SAB in children with WS. Little et al. (2013) recruited a sample of children with WS aged between 6 and 15 years. The children completed the Sun-Moon Stroop task as a measure of response inhibition performance and an emotion recognition task where they were required to label emotional expressions. The children also completed a modified version of Adolph's Approachability Task which provided a measurement of SAB. Little et al. reported that a sub-group of individuals studied, who demonstrated the highest rating of SAB as indicated within Adolph's Approachability Task, also demonstrated the poorest response inhibition performance. This shed light on the heterogeneous nature of cognition and behaviour in WS which few previous researchers have investigated. The findings of Little et al. suggest that response inhibition impairments may be an important underlying factor in the inappropriate SAB demonstrated in WS. There was no significant association between emotion recognition performance and the children's performance in Adolph's Approachability Task. This challenges the theoretical assumptions of the amygdala hypothesis which propose that high levels of SAB in WS are a result of neural impairments in the amygdala that underlie emotion recognition deficits. The amygdala hypothesis proposes that these emotion recognition deficits account for the indiscriminate nature of the social interactions engaged in by individuals with WS as these individuals are unable to judge the approachability of a person due to their emotion recognition impairments. The findings of Little et al. have provided a substantial contribution to this field of research by testing two theoretical accounts of SAB. This study provides evidence of an association between EF's and the atypical social profile in children with WS, in line with the assumptions of the frontal lobe hypothesis.

Although the findings of Little et al. (2013) expand our understanding of the relationship between EF impairments and SAB in WS, this study has methodological limitations. These limitations will be discussed and the present thesis will aim to address these. Little et al. used Adolph's Approachability Task which experimentally examines a child's SAB towards unfamiliar people. In a subsequent investigation, Fisher et al. (2014) challenged the validity of this task as a measure of SAB. Fisher et al. (2014) noted that the children's scores in Adolph's Approachability Task were poorly correlated with real-life observations of SAB in WS, while parent reports were a strong reflection of real-life SAB. The present thesis takes into consideration this methodological critique and builds on the work of Little et al. by examining for an association between response inhibition performance and two measurements of SAB: Adolph's Approachability Task and parents' ratings of SAB. In light of the validity of parent ratings as an assessment of SAB, the use of multiple measures of SAB will allow more reliable conclusions to be drawn on the strength of the frontal lobe hypothesis as an account for the high levels of SAB in WS.

1.4. Peer relationships in Williams syndrome

A second key area of social difficulty in WS lies within peer relationships. This will form the second focus of the current thesis. Individuals with WS have been reported to demonstrate substantial difficulties forming and sustaining friendships with their peers and to experience high levels of social isolation (Davies et al., 1998; Elison et al., 2010). As will be discussed below, there is a need to examine the peer relationships of children with WS across environmental contexts and across perspectives in order to identify areas of social difficulty.

1.4.1. Typical trajectory of peer relationships

In order to expand our knowledge on the nature of peer relationships in WS, it is important to acquire an understanding of the trajectory of peer relationships during typical development. Peer relationships transform in nature as a child develops and acquires key social communication abilities which facilitate successful peer interactions (Curtis-Tweed, 2011). It is important to understand the characteristic features of peer interactions at each stage of development in order to draw conclusions on the success of a child's peer relationships (Parker, Rubin, Erath, Wojslawowicz, & Buskirk, 2015).

In the first few months of life, infants engage in limited interaction with their peers as they have not acquired the motor and cognitive skills which are necessary in reciprocal social interactions. From approximately six months of age, infants begin to engage with other infants at a basic level. These interactions involve pointing, smiling and basic vocalisations. Infants begin to develop imitation and joint attention abilities which allow them to engage in basic play interactions (Parker et al., 2015). Between 1 and 3 years of age, toddlers begin to differentiate between peers and establish selective friendships with certain children. These early friendships are suggested to provide a context for children to develop interpersonal skills and have been linked with later adaptive behaviour (Howes, 1996; Howes & Phillipsen, 1998).

Within Selman’s framework of friendship development (Selman, 1980), social cognition is proposed to underlie a child’s engagement in successful social interactions. As these social-cognitive skills further develop and advance in their sophistication, the child’s peer relationships are proposed to increase in strength, with these abilities argued to develop in parallel with one another (Curtis-Tweed, 2011; Parker, Rubin, Prince, & DeRosier, 1995).

Children’s peer relationships are suggested to increase in complexity during middle childhood in line with advances in language and social cognition (Curtis-Tweed, 2011). There are also greater opportunities to interact with peers during middle childhood as an increased number of children attend extra-curricular clubs and activities with their peers (Weiss & Smith, 1999). It is argued that children develop a more complex understanding of friendship within middle childhood. In early childhood, friendships are primarily understood in the context of play. As children acquire more complex social cognitive abilities, they are able to infer the intentions and feelings of their peers within social interactions. Children become more aware of the need to cooperate with their peers in order to maintain friendships and are more able to adjust their own behaviour in order to facilitate successful social interactions (Curtis-Tweed, 2011).

During adolescence, there is an increased focus on peer relationships. Adolescents have been reported to spend 29% of the day with their peers which significantly exceeded the time they spend with their parents (Csikszentmihalyi & Larson, 1984).

Social interactions with peers during adolescence are primarily independently led, with minimal involvement or supervision from parents. Friendships also become more selective during adolescence, with a decline in the quantity of friends. Friendships increase in quality as the adolescents invest greater time and resources in these relationships and are characterised by higher levels of intimacy, support and self-disclosure (Gifford-Smith & Brownell, 2003; Zarbatany, McDougall, & Hymel, 2000). It is important to consider the implications of delays at each stage of development on the formation of peer relationships for children with a developmental disability.

1.4.2. Peer relations in neurodevelopmental disorders

Peer relationships are an area of substantial difficulty among many children with neurodevelopmental disorders (Cuckle & Wilson, 2002; Hoza et al., 2005; Rowley, Chandler, Baird, Simonoff, Pickles, Loucas, & Charman, 2012). The neurodevelopmental disorders of ADHD and ASD share characteristic features with WS, which include EF impairments and atypicalities in social functioning. An insight into the peer relations of children with ADHD and ASD will be informative in expanding our understanding of the role of cognition and social functioning in the success of peer relations in children with WS. The present thesis will examine the relationships between response inhibition performance, atypicalities of social functioning and peer relationships in children with WS.

1.4.2.1. Peer relations in Attention Deficit-Hyperactivity Disorder (ADHD)

Peer relationship difficulties have been reported to be prevalent among children with ADHD (Hoza et al., 2005; Mrug, Hoza, Gerdes, Hinshaw, Arnold, Hechtman, & Pelman, 2012). Hoza et al. (2005) measured peer functioning in a sample of children with ADHD by using a peer nomination methodology. TD children rated each of their classmates, including the child with ADHD, on a likability scale. In addition, the TD children nominated which classmates they were the closest to and the least close to. The children with ADHD were significantly less popular and had significantly fewer dyadic friendships than their TD peers. 56% of the children with ADHD did not demonstrate a reciprocal friendship, in comparison to a significantly lower 32% of TD children. These peer relationship difficulties have been widely reported within the ADHD literature (Bagwell, Molina, Pelham, & Hoza, 2001; Diamantopoulou, Henricsson, & Rydell, 2005; Hoza, 2007; Sibley, Evans, & Serpell, 2010). The accumulated findings indicate that peer relationships are an area of substantial difficulty among children with ADHD.

Previous researchers identified high rates of comorbidity between the disorders of ADHD and WS (Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006). These disorders also demonstrate similar profiles of executive dysfunction, with substantial deficits in response inhibition (Rhodes, Riby, Matthews & Coghill, 2011). Children with ADHD, similar to children and adults with WS, experience peer relationship difficulties and have been reported to be overbearing and intrusive within peer interactions (Bagwell et al., 2001).

Previous research supports the hypothesis of a relationship between EF skills and peer relationships in children with ADHD (Chiang & Gau, 2014; Kofler, Rapport, Bolden, Sarver, Raiker, & Alderson, 2011; Rinsky & Hinshaw, 2011) Hay, Payne and Chadwick (2004) developed a psychological framework to account for the development of friendships during infancy. Hay et al. proposed that six cognitive, behavioural and self-regulatory skills underlie an infant's engagement in successful peer relationships, where impairment in one or more of these skills is suggested to contribute to a decline in the quality of the infant's peer relationships. Hay et al. proposed that an important factor in the development of friendships during infancy was behavioural regulation. They argued that disinhibited behaviour during peer relationships can cause conflict, particularly when infants invade others' personal space and boundaries. This psychological framework has clear implications for the peer relationships of children with developmental disorders.

Miller and Hinshaw (2010) recruited a sample of adolescent girls with ADHD. The adolescents completed a battery of EF tasks which measured response inhibition, planning, working memory and attention. Miller and Hinshaw also obtained measures of the adolescents' social functioning by soliciting ratings from parents and peers. There was a positive predictive relationship between the adolescents' EF performance and their peer acceptance and social skills. Furthermore, Chiang and Gau (2014) reported that EF impairment in spatial working memory and planning predicted social interaction deficits at school in children and adolescents with ADHD. Through a consideration of the relationship between EFs and peer relations in children with ADHD and the shared profile of executive dysfunction between WS and ADHD, further research is required to investigate the role of EF impairments in the peer interactions of children with WS.

1.4.2.2.Peer relations in Autism Spectrum Disorder (ASD)

ASD is characterised by substantial impairments within social communication, social interactions and interpersonal relationships (American Psychiatric Association, 2000). While individuals with WS and ASD show apparently opposing social profiles (Lincoln, Searcy, Jones, & Lord, 2007), these individuals share several characteristic behavioural traits and deficits in EF and social-cognition. Individuals with WS and ASD frequently demonstrate high levels of anxiety and restricted and repetitive behaviours (Rodgers, Riby, Janes, Connolly, & McConachie, 2012). These behavioural traits are suggested to be implicated in the peer relationship difficulties and high levels of social isolation reported within these disorders (Bellini, 2004; White & Robertson-Nay, 2009). Furthermore, WS and ASD are characterised by impairments in social and communicative functioning (Klein-Tasman et al., 2011; Klin, Saulnier, Sparrow, Cicchetti, Volkmar, & Lord, 2007). The overlap between these disorders is further highlighted by rates of comorbidity, where a sub-group of individuals with WS were also reported to demonstrate a comorbid diagnosis of ASD (Klein-Tasman, Mervis, Lord, & Phillips, 2007). Through a consideration of this shared social phenotype, it is important to examine the peer relations of children with ASD in order to inform and expand our understanding of the role of social impairments in the peer relationships of children with WS.

Children with ASD experience significantly higher levels of loneliness, peer victimisation and poorer quality peer relationships than their TD peers (Bauminger & Kasari, 2000; Rowley et al., 2012). In an observational study, Rowley et al. (2012) examined experiences of friendship and peer victimisation among children with ASD using the Autism Diagnostic Observation Schedule (ADOS) (Lord, Rutter, Dilavore, & Risi, 1999) The ADOS measures communication, social interactions and social behaviour among children with ASD through play activities. Only half of the children with ASD demonstrated evidence of a reciprocal friendship. Similarly, Orsmond, Krauss and Seltzer (2004) noted that only half of adolescents with ASD were reported to have a reciprocal friendship with a peer. The children with ASD reported high levels of bullying and social exclusion, which often led to feelings of loneliness (Rowley et al., 2012). These findings highlight that peer relationships are an area of difficulty for many children and adolescents with ASD.

Researchers have identified a relationship between social and communicative impairments and peer relations in children with ASD. Orsmond et al. (2004) interviewed the parents of a sample of adolescents with ASD to solicit information on peer relationships and social participation. A measure of social impairment was also derived using the ADOS, where the children’s eye gaze, facial expressions and behaviours during reciprocal interactions were coded. The adolescents’ level of social impairment was a significant predictor of the quality of their peer relationships and participation in social activities. Furthermore, Rowley et al. (2012) reported a significant negative relationship between social interaction and communicative impairments, as measured within the ADOS, and friendship quality in children with ASD. These findings further support the importance of social communicative skills in an engagement in successful peer interactions. With consideration to the shared profile of social and communicative impairments among individuals with WS and ASD, the present thesis will examine for a relationship between social and communicative impairments as assessed within the standardised Social Responsiveness Scale (Constantino & Gruber, 2012) and peer relationships in children with WS.

1.4.3. Peer relations in Williams syndrome

While research investigating peer relations in children with WS is relatively sparse, research has been conducted to examine peer relationships in adults with WS (Davies et al., 1998; Elison et al., 2010; Udwin, 1990). These research studies will be informative in guiding areas for investigation in the present thesis. Davies et al. (1998) administered several standardised interview schedules with the parents of a sample of adults with WS (N=62). These interviews assessed daily functioning, academic performance, emotional and behavioural functioning and social relationships. 96% of the adults with WS were reported by their parents to experience substantial difficulties forming and maintaining friendships. Furthermore, two thirds of the adults with WS were reported to be socially isolated. These high frequency rates highlight the prevalence of peer relationship difficulties among adults with WS.

Many of the adults with WS were reported to show disinhibition within their social interactions which was believed to hinder their peer relationships. Over half of the adults with WS were overly tactile towards familiar people and 10% showed indiscriminate affection towards familiar and unfamiliar people. Furthermore, 80% of the adults with WS were reported to show intense chatter upon meeting someone and often asked inappropriate questions or made inappropriate comments (Davies et al., 1998). These accumulated findings highlight that while the adults with WS were driven to approach peers, their social naivety and poor understanding of social norms and boundaries posed implications for the success of their peer relationships.

Elison et al. (2010) built on previous findings by examining social inclusion in adults with WS. Almost half of the adults with WS were reported to rarely or never attend extra-curricular clubs and activities. Among the adults who did engage in extra-curricular activities, the majority of these adults experienced difficulties participating in the activities and engaging with peers. Furthermore, the majority of the adults with WS who attended extra-curricular activities relied on their parents to arrange these social events. These findings highlight the adults' lack of social independence and low levels of inclusion with peers.

The accumulated findings provide support for a hypothesis of social functioning and peer relationship difficulties in adults with WS (Davies et al., 1998; Elison et al., 2010; Udwin, 1990). Only a few studies thus far have investigated peer relations in children with WS. Greer et al. (1997) measured social skills and adaptive functioning in a sample of children with WS by soliciting parent ratings across standardised scales. 60% of the children with WS were reported to demonstrate social problems. Furthermore, many of the children with WS had few enduring social contacts and a low level of engagement in extra-curricular activities. This supports previous findings of high levels of social isolation among children with WS (Gosch & Pankau, 1994; Gosch & Pankau, 1997; Udwin & Yule, 1991; Udwin, Yule, & Martin, 1987). However, Greer et al. did not compare the performance of the children with WS to normative population scores and were therefore limited in their ability to draw reliable conclusions on levels of social impairment.

1.4.4. Atypicalities of social functioning in Williams syndrome

Accumulated research findings have provided support for atypicalities in the social functioning of children with WS. Two areas of social functioning have been identified as particularly problematic in WS: social cognition and social communication. These social impairments will be discussed, with reference to research evidence.

1.4.4.1.Social cognitive impairments in Williams syndrome

Social cognition is the cognitive mechanism which underlies an individual's engagement in reciprocal social interactions (Senju, 2013). Social cognition incorporates several mechanisms including attention to social cues, attributions and interpretation of mental states (Tager-Flusberg et al., 2006). One aspect of social cognition which is implicated in the success of a child's social interactions is 'theory of mind'. 'Theory of mind' is the ability to interpret the mental states which underlie a person's behaviour. These mental states can include an individual's knowledge, beliefs and intentions. An understanding of the mental states which underlie overt behaviours is informative in allowing an individual to respond appropriately within a social interaction (Morris et al, 2006; Riby & Back, 2010). Porter et al. (2008)

administered a story sequence task to a sample of children with WS and TD children who were matched on mental age and chronological age. The story sequence task provided a measurement of false belief performance. The children and adults with WS performed significantly poorer in this false belief task than the matched controls. The accumulated findings support substantial theory of mind impairments among children and adults with WS when compared to children with an intellectual disability (Tager-Flusberg & Sullivan, 2000) and TD children who were matched on mental age and chronological age (Porter et al., 2008).

1.4.4.2.Social communication impairments in Williams syndrome

Individuals with WS can often display a very chatty and outgoing personality profile (Doyle et al., 2004; Jones et al., 2000; Järvinen-Pasley et al., 2010). Despite this, individuals with WS show evident pragmatic language impairments (Laws & Bishop, 2004; Stojanovik et al., 2001). These pragmatic language deficits could be suggested to contribute to the peer relationship difficulties which are prevalent in WS. Pragmatic language refers to a set of implicit communicative rules which guide successful social interactions. Laws and Bishop (2004) administered the Children’s Communication Checklist to the parents and teachers of a sample of children with WS as an assessment of pragmatic language skills. The children and adults with WS performed significantly poorer than TD children who were matched on mental age across all domains of this measure. Furthermore, the children and adults with WS showed a significantly higher level of stereotyped speech and inappropriate initiation of conversations than TD controls and individuals with developmental disabilities.

These research findings align with research studies which reported that individuals with WS frequently made irrelevant statements which diverged from the conversational topic, relied on others to lead and maintain the conversation, demonstrated difficulties interpreting meanings within conversations and picking up on relevant social cues (Davies et al., 1998; Laws and Bishop, 2004; Philofsky, Fidler, & Hepburn, 2007; Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004; Stojanovik, 2006; Udwin & Yule, 1991). Pragmatic language impairments have been suggested to disrupt the flow of communication and an individual's ability to engage in reciprocal social interactions (Stojanovik et al., 2001).

Through a consideration of the accumulated findings of atypical social functioning in children with WS (Klein-Tasman et al., 2011; Porter et al., 2008) and the importance of these social functions in an engagement in successful peer relationships (Stojanovik et al., 2001), there is a need to examine the role of these social constructs in the peer relationships of children with WS. The present thesis will build on previous work by directly investigating the relationship between atypicalities in social functioning and the quality of peer relations in children with WS. Social functioning will be assessed by administering the standardised SRS.

1.4.4.3. A multi-informant assessment of social functioning and peer relations in Williams syndrome

Previous research which has been conducted to examine social functioning in WS has predominantly used a single informant approach (Klein-Tasman et al., 2007; Tager-Flusberg et al., 2006; Stojanovik, 2001). Klein-Tasman et al. (2011) were the first researchers to utilise a multi-informant approach to examine social functioning in children with WS. Klein-Tasman et al. (2011) administered the standardised Social Responsiveness Scale (SRS) to the parents and teachers of a sample of children with WS (4-16 years). The SRS measures social and communicative functioning across five subscales: social awareness, social cognition, social communication, social motivation and autistic mannerisms. This scale has been widely validated with both typical and atypically developing populations as a strong measurement of these social constructs (Constantino & Grubber, 2012). Parents and teachers reported that the children with WS were most impaired in the social cognition and autistic mannerisms sub-scales, while the children with WS were significantly less impaired in the social motivation sub-scale. These findings highlight that this pattern of strengths and difficulties persists across parent and teaching ratings. This suggests that these social difficulties are a core feature of the WS social phenotype. However, Klein-Tasman et al. revealed that the parents rated the children with WS as significantly more impaired within social and communicative functioning than the teachers' ratings of the children. This suggests that social functioning impairments may be more prominent in the home environment, potentially due to the unstructured nature of this social setting. The present thesis aims to further develop these findings by obtaining measures of peer relations and social functioning from the parents and teachers of children with WS in order to examine the peer relations of children with WS across informants.

While previous researchers used a multi-informant approach to examine social functioning in children with WS (Klein-Tasman et al., 2011), only one study to the researcher's awareness has investigated peer relations in children with WS using a multi-informant approach (Laws & Bishop, 2004). Laws and Bishop (2004) administered the child communication checklist with the parents and teachers of a sample of children and young adults with WS. This standardised scale measures several aspects of social communication and social relationships. The children with WS were reported to have significantly poorer social relationships than TD children who were matched on mental age and children with DS. This study has expanded our knowledge of peer relationship difficulties in children and young adults with WS; however, social relationships were one sub-scale of a larger measure which assessed several aspects of social communication and language skills. In consequence, this study provides a limited examination of where these difficulties lie.

The present thesis will build on previous findings by administering standardised and specifically designed measurements of both social functioning and peer relationships with the parents and teachers of children with WS. The parents' and teachers' ratings within the standardised scales will be compared to normative population scores in order to draw conclusions regarding levels of impairment. The use of a multi-informant approach will allow an examination of the children's social functioning and peer relations across environmental settings and from different perspectives.

Qualitative studies have been conducted to examine peer relations in children with a range of neurodevelopmental disorders, including ADHD, ASD and DS (Bagwell et al., 2001; Cuckle & Wilson, 2002; Orsmond et al., 2004; Rowley et al., 2012). Interviews are a valuable method which can provide a qualitative assessment of personal experiences and an examination of individual differences within a population. A research study was conducted by Cuckle and Wilson (2002) to investigate the nature of peer relations in adolescents with DS. Cuckle and Wilson conducted semi-structured interviews with a sample of adolescents with DS, their parents and learning support teachers regarding the adolescents' peer relations and social inclusion. By soliciting the perceptions of different informants, this study gained insight into the adolescents' difficulties developing and sustaining friendships and the adolescents' experiences of social exclusion across settings. No comparable qualitative studies have been conducted thus far with a sample of children with WS. Interview schedules were developed for use in the present thesis to measure specific characteristics of peer relations in children with WS from the perspectives of the child with WS, their parent and their teacher.

1.5. Aims of the present thesis

The present thesis has two overarching aims. The first aim of this thesis is to examine the characteristics of SAB and peer relationships in children with WS. In order to achieve this, the current thesis will solicit quantitative and qualitative information from several informants, the children with WS, their parents and teachers, in order to expand our knowledge on the prevalence and the manifestation of these social difficulties in children with WS. This thesis will build on previous research by examining the characteristics of social functioning in children with WS across environmental contexts and across informants and will examine for within-syndrome variance in these behaviours. The second aim of the present thesis is to examine the role of psychological processes, specifically, the role of response inhibition, information processing and social functioning skills in the SAB and peer relationships of children with WS. By identifying areas of social impairment in WS, the findings can inform the development of interventions in the future to support children with WS and their families within the domains of friendship, social skills and stranger danger awareness.

1.6. Chapter structure of the present thesis

Chapter 2 will detail the methodologies used in this thesis. There are several important considerations to take when recruiting children with a developmental disorder, for example the need to recruit appropriate comparison groups. These methodological considerations are discussed in depth within this chapter. In addition to standardised tasks, several questionnaires and interview schedules were developed or modified for application in the present thesis. This chapter describes the development and psychometric properties of each measure. This chapter provides a sufficient level of detail on the participant sample, measures and procedure to allow future replication.

Chapters 3, 4 and 5 report on investigations into SAB in children with WS. Chapter 3 examines the prevalence and manifestation of SAB in children with WS. The children with WS completed an experimental task (Adolph’s Approachability Task), within which they were required to indicate the extent to which they would like to approach an unfamiliar person depicted across the images. The children’s performance in this task was compared to TD children who had been matched on verbal ability and TD children who had been matched on non-verbal ability. The children with WS indicated a significantly higher level of SAB in Adolph’s

Approachability Task than both TD comparison groups. The parents of the children with WS also completed a questionnaire designed to examine the characteristics of the children's SAB. All of the children with WS were reported to frequently approach strangers and to show a strong interest in conversing with strangers. However, there was within-syndrome variance in the manifestation of SAB, where only a sub-sample of the children with WS were reported to engage in disinhibited tactile behaviour towards strangers.

Chapter 4 investigates which psychological processes underlie an engagement in high levels of SAB. The relationships between response inhibition, social information processing and SAB were examined in a sample of children with WS and a sample of TD children. Response inhibition was measured using the standardised Sun Moon Stroop task. In order to examine information processing, the children were presented with social vignettes which were designed to elicit the children's causal attributions and predictions. Within each social vignette, the children were instructed to identify the cause of the event or to predict the outcome of the event from three response options, a positive, negative and neutral option. There was no significant difference in the valence of the causal attributions and predictions formed by the children with WS and TD children matched on verbal ability and TD children matched on non-verbal ability. While there was no significant relationship between response inhibition performance, information processing and levels of SAB among the children with WS, there was a significant relationship between the positivity of the attributions formed by the TD children and the children's levels of SAB as measured in Adolph's Approachability Task.

Within chapter 5, an interview methodology was used to further solicit parents' perceptions on their child's SAB. These interviews tapped into the manifestation of the children's SAB and the implications of this SAB for the child and family life. A common theme across the interview extracts was the indiscriminate nature of the children's social interactions. The children's high levels of SAB were accounted for by some parents as due to the child's poor impulse control, while some parents attributed this behaviour to their child's lack of an ingrained sense of stranger danger. As a result of their perceived social vulnerability, many parents reported that their child required constant supervision, with implications for family functioning.

Chapters 6 and 7 expand our knowledge of peer relationships in children with WS. Within chapter 6, the parents and teachers of the children with WS completed standardised questionnaires which measured social functioning. Parents and teachers also completed questionnaires designed to measure the characteristics of peer relations in WS. The use of a multi-informant approach builds on previous research and allowed an investigation into the stability of peer relationships across informants and contexts. The children with WS were rated significantly poorer across measures of social functioning than TD children matched on verbal ability and TD children matched on non-verbal ability. This was consistent across parents' and teachers' reports. Furthermore, this chapter provided a detailed account on the characteristics of peer relationships in children with WS. Many of the children with WS were reported to have difficulties establishing and sustaining peer relationships and to experience social exclusion from peer social activities. This chapter also examined the role of response inhibition and social functioning skills in the peer relationships of children with WS. The children's social functioning skills were measured using the SRS which was completed by parents. There were no significant relationships between inhibition performance and peer relations among the children with WS. However, the children's impairment in social cognition, social communication and social awareness, as measured in the SRS, were significantly associated with their peer relationship difficulties. This suggested that peer problems in WS are a consequence of features of the social phenotype in WS, rather than simply a consequence of intellectual delay.

Chapter 7 extended our knowledge of peer relations in WS by soliciting qualitative information from the children with WS, their parents and teachers. The children with WS and their parents individually took part in a semi-structured interview about the child's peer relationships and social inclusion. Teachers completed a semi-structured questionnaire, where open ended responses were obtained regarding their perceptions of the children's peer relations. Across the data extracts, many of the children with WS were reported to have difficulties sustaining friendships with peers. This was attributed to several factors, including their communication difficulties and deficits in their play skills. These peer difficulties were reported by parents and teachers, suggesting that these are core aspects of the WS social phenotype which are stable across environmental contexts. However, these difficulties in peer relationships were not perceived by the children themselves.

Finally, in chapter 8, conclusions are drawn on the atypical patterns of social functioning in children with WS, with a focus on the children's SAB and peer relationships. This chapter evaluates the research findings in terms of their contribution to research and practice. The findings from the current thesis are discussed, with a focus on how this thesis has developed previous research, in particular its methodological advances. The practical implications of the findings are also discussed. This includes the potential to inform the design of behavioural interventions in the future which support children with WS in their daily social lives and the role of the current findings in raising awareness of the social difficulties experienced by children with WS.

Chapter 2: General methodology

This chapter will detail the methodological decisions which were taken in the present thesis and provide clear justifications to support these decisions by drawing on previous research where applicable.

2.1. Design considerations

This section will describe the research designs that were employed within the empirical chapters of this thesis.

2.1.1. Factorial design

Careful consideration is required to control for an effect of intellectual delay on task performance when conducting research with children with neurodevelopmental disorders. The use of a factorial design allows direct comparisons to be made between the social functioning of children with WS with that of TD children who have been matched to the children with WS on intellectual ability. This factorial design allows stronger conclusions to be formed, where any observed differences in task performance or questionnaire ratings can be interpreted as a consequence of the disorder rather than due to a general intellectual delay (Jarrold & Brock, 2004).

2.2. Group matching criteria

WS is characterised by a dissociation in performance between verbal and non-verbal domains. Children with WS demonstrate relative strength in verbal tasks and weakness in visuo-spatial tasks (Jarrold, Baddeley, & Hewes, 1998; Jarrold, Baddeley, & Phillips, 2007; Vicari, Bellucci, & Carlesimo, 2003). Through a consideration of this dissociation in performance, two TD comparison groups were recruited in the present thesis. Each child with WS was matched with a TD child on verbal ability in the British Picture Vocabulary Scale II – BPVS II (Dunn, Dunn, Whetton, & Burlay, 1997). Each child with WS was also matched with another TD child on non-verbal ability in Raven's Coloured Progressive Matrix – RCPM (Ravens, Ravens, & Court, 1998). These comparison groups allowed an examination of the extent to which the children with WS deviated from normative scores when intellectual ability was controlled for. The chronological age, verbal and non-verbal performance scores for the children with WS and TD comparison groups are reported in the individual participant sections throughout this thesis.

Many previous researchers used the Wechsler's intelligence scale for children (Wechsler, 1991) to match participant groups on intellectual ability (Capitão et al., 2011; Santos, Silva, Rosset, & Deruelle, 2010). Through a consideration of the dissociation in performance between verbal and non-verbal domains in WS, the Wechsler's intelligence scale for children (Wechsler, 1991) was not believed to be appropriate for use in the present thesis as this measure does not allow verbal and non-verbal performance to be examined separately.

The BPVS II (Dunn et al., 1997) is a standardised measure of receptive vocabulary. This was used in the present thesis to match the children with WS to a sample of TD children on verbal ability. The BPVS II has been widely used to assess verbal functioning in TD children and children and adults with intellectual disabilities (Ferguson & Murphy, 2014; Kirk, Hocking, Riby, & Cornish, 2013; Rhodes, Riby, Fraser, & Campbell, 2011; Rhodes et al., 2010). The BPVS II consists of 4 practice sets and 14 experimental sets. There are 12 items in each of the experimental sets within this task. This generates a total of 168 items. Each set increases gradually in the level of difficulty from the previous set. For each item the participants are presented with four simple black and white images and are asked to identify which image they consider to best illustrate the stimulus word. A baseline score is established when the participant completes a set with one or less errors. A ceiling score is established when the participant demonstrates 8 or more errors within a set at which point the testing session is terminated. The BPVS II has been reported to be effective for application with individuals of a wide age range and spectrum of abilities.

RCPM (Ravens et al., 1998) is a standardised measure of abstract reasoning which has been administered in previous research to match participant groups on non-verbal performance (Riby & Hancock, 2009; Riby et al., 2011). This was used in the present thesis to match the children with WS with TD children on non-verbal ability. This task consists of 36 abstract reasoning problems which are presented across 3 sub-sets. In each item the participants are presented with a coloured matrix which has one piece missing and they are required to identify the missing part from 6 illustrated options. The RCPM is a modification of the Standard Raven’s Progressive Matrix

(Ravens, 1938). Within this modified task, coloured images are used in place of the original black and white images. This modified task also uses fewer trials and therefore has a shorter completion time. In consequence to these modifications, the RCPM is argued to be suitable for application with younger children (aged 5-11 years) and individuals with intellectual impairments.

2.3. Participant characteristics

The rare nature of WS and characteristics of this disorder poses implications for the research design and subsequent data analysis. This section will detail the nature of the participants recruited in this thesis and the implications of this sample on research design and implementation.

2.3.1. Sample size

WS is a rare neurodevelopmental disorder with an estimated prevalence of between 1 in 7,500 and 1 in 20,000 individuals within the UK (Eisenberg, Jabbi, & Berman, 2010). 21 children with WS (12F, 9M) were recruited in the present thesis. 14 of the children with WS attended a mainstream school and seven children attended a SEN school. This sample size is in line with previous research in this area (Capitão et al., 2011 (N = 15); Fisher, 2014 (N=21); Frigerio et al., 2006 (N=21); Järvinen-Pasley et al., 2010 (N=20); Rhodes et al., 2010 (N=19); Riby, Janes, & Rodgers, 2013 (N=21)). Martens et al. (2008) conducted a review of 178 published studies from the WS literature and reported that the median sample size across these studies was between 6 and 17 participants with the exception of those studies which obtained parental reports of their children’s behaviours through postal or telephone correspondence.

The relatively small sample size recruited (N=21) suggests that caution and careful consideration are required when conducting statistical analysis with these data. A regression analysis requires a larger sample size (Field, 2009) than was possible to recruit in the present thesis due to the rare nature of WS and geographical spread of the participants across the UK. Statistical tests such as correlational analysis and analysis of variance (ANOVA) have been shown to be suitable for application with smaller sample sizes and therefore are considered to be appropriate for use in the present thesis (Doyle et al., 2004; Järvinen-Pasley et al., 2010).

2.3.2. Confirmation of diagnosis

All children in the clinical group had a formal diagnosis of WS which had been previously confirmed through positive genetic fluorescent in situ hybridisation testing (FISH test). The FISH test was developed in the 1990’s, prior to which all diagnoses of individuals with WS were based on clinical observations (Martens et al., 2008). WS is caused by the deletion of approximately 28 genes from chromosome seven. The FISH test is a blood test which is used to detect the absence of the elastin gene. The elastin gene has been found to be consistently deleted among individuals with WS and therefore has been identified as a method of diagnosis of WS (Apps et al., 2008). The elastin gene functions as a protein which forms connective tissues within the aorta blood vessels. The deletion of this gene has been linked to cardiac problems which are widely reported in WS (Järvinen-Pasley et al., 2008).

2.3.3. Screening procedure

In order to make clear comparisons between the children with WS and TD children, it was important to screen for the presence of mental or behavioural disorders among the TD children. The standardised Strengths and Difficulties Questionnaire (Goodman, 1997) was used as a screening measure in the present thesis. Further details on this questionnaire are provided in section 2.6.1.1. Parents and teachers were both invited to complete this questionnaire in order to control for incidences of non-response. A composite score termed ‘the total difficulties score’ (TDS) was generated for each child that indicated their level of social impairment. Where both the parent and teacher of a child had completed the SDQ, the TDS from the teacher was used as a measure of social behaviour in line with previous research (Docherty-Sneddon et al., 2012; Riby & Hancock, 2008; Riby & Hancock, 2009; Rhodes et al., 2010). The children’s TDS were subsequently categorised into ‘normal’, ‘borderline’ and ‘abnormal’ in accordance with the SDQ guidelines. Exclusion criteria were set out prior to testing. Any children who scored in the abnormal category in the SDQ were removed from the dataset and any participants who scored in the borderline range across both parent and teacher reports were removed from the dataset. Four children were removed from the dataset prior to analysis as they met these exclusion criteria. No SDQ data were provided from either informant for an additional three children. These children were subsequently removed from the dataset prior to analysis as it was not possible to assess their eligibility to participate in this research.

2.3.4. Age considerations

Children with WS aged between 6 and 16 years old were contacted regarding participation in this research. The experimental tasks were chosen with careful consideration to the characteristics of the participant sample. It was important that the tasks were not too simple or too difficult for the children in order to avoid the occurrence of ceiling or floor level performance. All tasks were considered suitable for the intellectual ability of the children recruited. The Sun-Moon Stroop task has been successfully administered as a measure of response inhibition performance among TD children (Archibald & Kerns, 1999; Pasalich, Livesey, & Livesey, 2010) and children with WS (Little et al., 2013) which supported the suitability of this task for the present thesis. The attribution and prediction scales were read aloud to the children and visual aids were provided to optimise the suitability of this task for young children and children with developmental disorders. This was intended to avoid demands on the children’s literacy skills. Adolph’s Approachability Task has been successfully administered to children with WS of a similar age range to the participants in the present thesis and was therefore considered appropriate for use in the present thesis (Little et al., 2013; Martens et al., 2009). This task was further modified by incorporating emotive faces alongside the words in the Likert scale. These visual aids were again intended to avoid demands on the children’s literacy skills.

2.4. Recruitment procedure

Full ethical approval was granted from the University of Strathclyde ethics committee. Sections 2.4.1 and 2.4.2 will provide information on the recruitment of the WS and TD participant samples.

2.4.1. Williams syndrome sample

All participants were recruited through the UK William Syndrome Foundation (WSF). All families who had a child with WS aged between 6 and 16 years and resided within Scotland, Northern England, Central England and North Wales were contacted by letter (N = 79). Information sheets were sent out to the parent and the young person. These provided details on the nature of the research project and used language which was deemed appropriate for the recipient. The parents were invited to read this information sheet and to indicate if they wished to participate in this research by returning the enclosed consent form. 21 families provided consent to participate in this research. This provided a 27% participation rate. In all cases, permission was granted for the child with WS and their parent to participate in this research. The mother (N=11), father (N=2) or both the mother and father (N=8) participated in this research. The participants were recruited from Northern and Central England (71%), Scotland (24%) and North Wales (5%). All participants were visited at their home address where they took part in this research.

20 (95%) parents provided permission for the researcher to contact the child’s class teacher to seek their participation in this research project. Permission was initially sought from the relevant local education authorities (LEAs). All LEAs provided permission to contact the school in question. A letter was sent to the Head teacher of each school to seek permission to invite the child’s class teacher to participate in the research. On occasions where there was no response from mail correspondence, the head teacher was contacted by telephone. 20 Head Teachers provided permission to proceed with the research. Subsequently 18 of the class teachers agreed to participate in this research.

2.4.2. Typically developing sample

Seven Scottish LEAs were contacted by letter. An application form was completed for each LEA which detailed the research project and the recruitment procedure. These applications were reviewed by a panel at each individual LEA and four authorities provided permission to proceed with the recruitment process. A total of 110 primary schools were contacted across these four LEAs. The Head Teacher of three primary schools on the outskirts of Glasgow consented to participate in this research study.

Information sheets and consent forms were sent to the parent/carer of all children between five and seven years of age at each school. Parental permission was sought regarding their child’s participation and their own participation in this project. Permission was sought from the child’s class teacher for their own participation in this research. Finally verbal consent was obtained from the children.

A total of 71 TD children aged between 5.02 and 7.96 years were recruited (M: 6.54, SD: .90; 31F, 40M). Seven of these children were removed from the dataset prior to analysis because they had scored in the ‘abnormal’ range in the SDQ screening measure (N=3), SDQ data were not provided for the child (N=3), or the child chose to withdraw from the study (N=1). The final dataset for the TD sample was 64 children. Of these, 21 TD children were individually matched with the children with WS on verbal ability in the BPVS II and 21 TD children were individually matched with the children with WS on non-verbal ability in RCPM.

2.5. Experimental measures

This section will detail the experimental measures which were administered within the present thesis. Several of these measures were developed or modified for use in this thesis. This section will detail the construction of these measures with reference to previous research literature and internal reliability statistics.

2.5.1. Adolph’s Approachability Task

The standardised Adolph’s Approachability Task (Adolphs et al., 1998) was modified for use in the present thesis (please see appendix 2). This provided a measurement of the children’s SAB to unfamiliar people. Adolph’s Approachability Task has been widely employed by researchers as a measurement of SAB in both TD children and children with developmental disorders (Bellugi et al., 1999; Frigerio et al., 2006; Little et al., 2013; Martens et al., 2009; Porter et al., 2007). Adolph’s

Approachability Task utilises black and white images of human faces. However, individuals perceive the world in colour and therefore these images could be argued to lack real life generalizability. For the present research, coloured digital photographs of human faces were used. The task contained 20 trials. In 10 trials, an unfamiliar male adult was presented and in 10 trials an unfamiliar female adult was presented. All face stimuli depicted a neutral expression. The face stimuli were taken from the ‘Centre for Vital Longevity Face Database’ (Minear & Park, 2004) which was freely available for use.

The children were asked to view each image and to rate the extent to which he or she would like to talk to the person in the image on a five point Likert scale. This Likert rating scale was colour coded to maximise its usability for young children and children with intellectual difficulties (Lamberski & Dwyer, 1983). Within this task, a rating of 1 indicates that the child definitely would not want to talk to the person within the image and a rating of 5 indicates that the child would really like to talk to the person within the image. Two features, colour and facial expression, were manipulated to emphasise the range. Thus, the Likert scale ranged from bright red and very sad (=1) to bright green and very happy (=5). Little et al. (2013) employed this rating scale and found it to be effective in its application with young children and children with WS. The possible score range in this task was a minimum of 20 and a maximum of 100.

2.5.1.1. Pre-rating of face stimuli

The face stimuli used in Adolph's Approachability Task were pre-rated by a sample of TD children (N=64). Although all of the faces were neutral in valence, pre-ratings allowed an examination of the rank order of approachability in a sample of TD children. The TD children's ratings for each face were subsequently grouped into two categories, the 10 faces which had been rated the highest by the TD children, 'the most approachable' (M: 2.73, SD: .22) and the 10 faces which had been rated the lowest by the TD children, 'the least approachable' (M: 2.34, SD: .17). There was no significant difference between the children with WS's ratings for the faces which had been pre-rated as 'most approachable' (M: 3.33, SD: .90) and the faces which had been pre-rated as 'least approachable' (M: 3.63, SD: .86), t (19) = -2.0, p = .060 (two tailed). Although this statistical test was close to significance, an observation of the mean scores indicated that the children rated both sets of face stimuli within the mid-range of the scale. Furthermore given the relatively small number of face stimuli administered in the present thesis (N=20), the author did not deem it reliable to split the face stimuli into two independent categories in light of the close proximity of these ratings. In consequence, all analysis within this thesis will be performed on the combined set of faces.

2.5.2. Sun Moon Stroop Task

The Sun-Moon Stroop task (Archibald & Kerns, 1999) is a standardised measure of inhibitory control performance. This task was a modification of the Day-Night Stroop task (Gerstadt et al., 1993). The Day-Night Stroop task was suggested to be too simplistic as indicated by findings of ceiling level performance by the age of 7 (Archibald & Kerns, 1999).

The Sun-Moon Stroop task has been widely administered with both TD children and children with developmental disorders as a measurement of inhibitory control performance (Archibald & Kerns, 1999; Little et al., 2013; Pasalich et al., 2010). The Sun-Moon Stroop task demonstrated strong test-retest reliability (r = .90) and was strongly correlated with other measures of response inhibition (r = -.2, p = .03) when tested with a sample of TD children (Archibald & Kerns, 1999).

There are two conditions within the task, a control condition and an experimental condition. In the control condition, the children are presented with a piece of paper with 30 pictures of the sun and the moon which are arranged in a randomised order. These images were taken from the global website Google. The images are freely available to use and free from copyright laws. The children were instructed to name each picture e.g. respond “sun” when they saw a picture of a sun and “moon” when they saw a picture of a moon. In the experimental condition, the children were instructed to inhibit their initial response by responding “sun” when they saw a picture of a moon and “moon” when they saw a picture of a sun. In both conditions, the children were instructed to respond to as many pictures as they could in a 45 second interval and to correct themselves if they answered incorrectly before proceeding to the next picture. The researcher pointed at each picture sequentially and if the child answered incorrectly the researcher continued to point at this picture until the child had corrected himself or herself. The children were given a brief practice block where they were asked to name the first four images while following each rule. In order to avoid ceiling level performance in this task as has been found in past research (Gerstadt et al., 1993), an interference score was generated for each participant. The interference score was calculated as the difference in accuracy between the control condition and the experimental condition, where lower scores indicated stronger inhibitory control performance. This has been shown to be a strong measure in both clinical and TD samples (Archibald & Kerns, 1999; Little et al., 2013).

A computerised task was developed as an additional measure of response inhibition. This was piloted with a sample of TD children. However, the children's scores were negatively skewed (-1.84), which indicated ceiling level performance, and were poorly correlated with performance in the standardised Sun Moon Stroop Task (r = 16, p = .208). This task was not administered further in this thesis.
Further information on the development of this Stroop task can be found in appendix 1.

2.5.3. Attribution/prediction scales

To the author's awareness, only one study thus far has examined social information processing in WS. Godbee and Porter (2013) developed an intention attribution task. The participants were presented with black and white cartoon images. The intentions of the actor within these drawings were ambiguous. The participants were asked to describe what was happening within each scene and to predict what was going to happen next in the scene. The participants' references to the actor's intentions as negative or neutral were coded. Cartoon line drawings can be argued to lack real-life generalisability and may not measure the true nature of children’s attributions. In addition, this task did not code for positive responses and therefore did not allow an examination for the presence of a positive attribution bias in WS.

Within the present thesis, three measures, a social attribution measure, a social prediction measure and a non-social prediction measure were developed (see appendices 4 - 6). These measures aimed to overcome the methodological limitations of previous research by directly examining for a positive bias in information processing in WS. Within each of these measures, seven vignettes were presented which depicted various social scenarios. Scenarios included going to the park, attending a birthday party and going to the cinema. The scenarios were believed to be familiar to the age group under investigation. These social vignettes were believed to yield greater real-life validity than the cartoon line drawings used by Godbee and Porter (2013). The vignettes were designed to elicit the children’s causal attributions and their future predictions across different scenarios. Each vignette was read aloud to the child and the child was provided with three possible responses to choose from: a positive attribution/prediction, a negative attribution/prediction and a neutral attribution/prediction. The children’s responses to each vignette were scored ‘+1’ for a positive attribution/prediction, ‘-1’ for a negative attribution/prediction and ‘0’ for a neutral attribution/prediction. This scoring system was constructed in order to generate a score for each measure which reflected a balance of the children’s responses. There were 7 items in each scale; hence, scores could range from -7 to 7, where the more positive the score, the more positive the child’s causal attributions and predictions.

2.5.3.1. Causal attribution measure

The children completed an attribution measure which was designed to elicit causal attributions within seven different social vignettes. In each case, the children were asked to identify what had caused the event, where the possible causes were uncertain or ambiguous. An example scenario is “Simon has been sent to the head teacher’s office. Why has he been sent here?” There are three options for the children to choose from. A positive attribution e.g. “to receive an award for doing well”, a negative attribution e.g. “because he is in trouble” and a neutral attribution e.g. “because the teacher needed some textbooks”. The present author used three response options, in contrast to earlier tasks which used two options, in order to reduce the risk of chance performance results. All vignettes and responses were read aloud to the children. This was intended to reduce demands on the children’s literacy skills. All scenarios were presented on PowerPoint presentation slides. The scenarios were accompanied by a coloured photograph which visually depicted the event.

2.5.3.2. Prediction measures

Prediction tasks were designed to elicit the children’s expected outcomes across a series of vignettes. The vignettes were ambiguous in nature and the children were asked to predict what the outcome would be in each scenario. The vignettes and responses were read aloud to the children and all response options were illustrated by an image. This aimed to increase the simplicity of this task for young children and children with developmental disorders.

2.5.3.2.1. Social prediction measure

In the social prediction task, seven social vignettes are presented to the child, and the child is required to predict the outcome of the event, by being asked the question “what will happen next?” The children are presented with three options to select from, a positive outcome, a negative outcome and a neutral outcome. All outcomes involve an interaction with another person, hence making them social in nature. An example vignette is “Lucy arrives at the restaurant for dinner. What will happen next?” There are three response options: a positive response (“The waiter will be really friendly”), a negative response (“the waitress will be really grumpy”) and a neutral response (“the waiter will arrive at the table”).

2.5.3.2.2. Non-social prediction task

In the non-social prediction task, the same seven social vignettes are presented to the child and the child is again required to predict the outcome of the event by answering the question “what will happen next?” The children are presented with three options to select from, a positive outcome, a negative outcome and a neutral outcome. However, in contrast to the social prediction task, all outcomes involve an interaction with an inanimate object. An example vignette is “Lucy arrives at the restaurant for dinner. What will happen next?” For the non-social prediction scale the children are asked to select from three alternative options: a positive response (“the food will taste really nice”), a negative response (“the food will taste horrible”) and a neutral response (“there will be some knives and forks on the table”).

2.6. Questionnaire instruments

2.6.1. Social Responsiveness Scale (Constantino & Gruber, 2012)

This is a 65 item questionnaire which measures a child's reciprocal social interactions. This scale is used within research and clinical practice to identify children with social impairments associated with ASD and in these cases to measure the frequency and degree of these impairments. This questionnaire assesses social impairments in children and adolescents aged between 4 and 18 years. This scale measures items in five domains: social awareness (8 items), social cognition (12 items), social communication (22 items), social motivation (11 items) and restricted interests and repetitive behaviour (12 items). The parents are asked to read each statement and rate the extent to which that applies to their child using a 4 point Likert scale, where 1 indicates 'not true' and 4 indicates 'almost always true'. Scores are generated for each domain together with an overall score which indicates the severity of the child's social ASD deficits. The SRS demonstrated strong internal reliability ($\alpha = .95$) and strong correspondence between informants ($r = .61$) when tested with a large normative sample (Constantino & Gruber, 2012). The SRS has been widely applied to measure social functioning in children with WS (Hanley, Riby, Caswell, Rooney, & Back, 2013; Klein-Tasman et al., 2011; Lough et al., 2015; Lough et al., 2016). Klein-Tasman et al. (2011) reported strong internal consistency in a sample of children with WS ($\alpha = .92$), supporting the suitability of this scale for application in the present research. The SRS demonstrated strong internal reliability when administered with the parents in the current thesis ($\alpha = .84$).

2.6.2. Strengths and Difficulties Questionnaire (Goodman, 1997)

The SDQ is a screening tool which assesses for the presence of mental and behavioural disorders in children aged 3-16 years old. The SDQ consists of 25 items which measure five constructs: emotional symptoms, conduct problems, hyperactivity/inattention, peer relationship problems and prosocial behaviour. This scale can be completed by either parents or teachers. The participants are instructed to read each statement carefully and to rate the extent to which the item applies to the child. All items are rated on a three point Likert scale, where 0 indicates ‘not true’, 1 indicates ‘somewhat true’ and 2 indicates ‘certainly true’. A score is generated for each sub-scale. The possible score range for each sub-scale is a minimum of 0 and a maximum of 10. The participants’ scores from the first four domains are combined to form a composite total score which is termed ‘total difficulties score’ (TDS). The participants’ scores are subsequently grouped into three categories: ‘normal’, ‘borderline’ or ‘abnormal’. A high score indicates more problematic social behaviour. For the teacher informants, a TDS between 16 and 40 is classified as abnormal and for parent informants a TDS between 17 and 40 is classified as ‘abnormal’. The SDQ has been widely administered with TD children and children with neurodevelopmental disorders (Muris, Meester, & Van Den Berg, 2003; Rhodes et al., 2010; Riby et al., 2013). Goodman (2001) reported that the SDQ showed strong internal reliability ($\alpha = .73$) and strong test re-test stability after 4 to 6 months ($r = .62$). The SDQ demonstrated acceptable internal consistency when administered with the parents ($\alpha = .69$) and teacher ($\alpha = .64$) within the present thesis.

2.6.3. Novel questionnaires

Several novel questionnaires were developed for use in the present thesis. The relatively small sample size of the WS group (N=21) limited the ability to perform robust statistical assessments of the inter-relationships between the items of the scales. Several researchers have proposed sample size estimates for conducting factor analysis. Nunnally (1978) recommended a participant and variable ratio of 10:1, while other researchers argued that a sample size of at least 300 was required for accurate results to be gained (Tabachnick & Fidell, 2007).

Costello and Osbourne (2005) examined the success of factor analysis across a range of sample sizes. They reported that larger sample sizes produced significantly more accurate solutions than smaller sample sizes. Costello and Osbourne (2005) reported that for studies with a participant-variable ratio of 2:1, only 10% of the factors identified were accurate. However, larger studies which used a participant-variable ratio of 20:1 demonstrated a 70% accuracy rate from the factor solutions derived. These findings have highlighted the importance of recruiting a large sample size when applying factor analysis to a dataset. In consequence of the small sample size (N=21) in the present study, Cronbach's alpha measure of internal reliability will be conducted on the individual constructs within the questionnaires. Cronbach (1951) developed this measure to determine the extent to which all items within a questionnaire consistently measure the same dimension. A Cronbach's alpha value of .7 or above has been identified as an indicator of adequate internal reliability (Fields, 2009). Individual constructs within the questionnaires will be identified based on observations of correlation values, where those items which are closely correlated will be grouped together as one construct.

2.6.3.1. Social approach questionnaire (parent report)

A 12 item questionnaire was developed for application in the present thesis (see appendix 3). These items were developed through reference to previous research as detailed below. The items in this questionnaire were developed to gauge three aspects of SAB: the manifestation of SAB (5 items), the perceived vulnerability of the child (4 items) and the impact of SAB on daily and family life (3 items).

While previous researchers have reported higher levels of SAB in children with WS when compared to TD children (Doyle et al., 2004; Martens et al., 2009), few researchers have examined the manifestation of these social approach behaviours. In line with previous findings of a poor awareness of personal space (Lough et al., 2015) and difficulties inhibiting behaviour among children with WS (Little et al., 2013; Porter et al., 2007), items were developed which specifically tapped into the disinhibited nature of communication and physical interactions in WS.

Individuals with WS have been reported to experience high levels of victimisation and social vulnerability (Fisher, Moskowitz, & Hodapp, 2012). Through a consideration of this social vulnerability and poor stranger danger awareness (Riby et al., 2014), items were incorporated into this questionnaire which measured the children’s social vulnerability around strangers and the parents’ concerns regarding this vulnerability.

Previous researchers reported that high levels of SAB in adults with WS had a negative impact on the individuals’ relationships, independence and vulnerability to victimisation and abuse (Davies et al., 1998; Elison et al., 2010). Few researchers have investigated the negative impact of this SAB on daily functioning in children with WS. Items were devised for inclusion in this questionnaire to measure the impact of this SAB on the children's independence, their vulnerability and on family life.

This questionnaire had an acceptable Cronbach's alpha value of .75. Further observation of the reliability statistics indicated that removing item 9 "If asked, my child would say that they know they should not approach strangers" further increased the internal reliability of this questionnaire. This generated an 11 item construct which had a Cronbach's alpha value of .80. Parents were asked to rate the extent to which each item in this scale applied to their child (1: Strongly disagree, 5: Strongly agree) or the frequency with which their child engaged in this behaviour (1: Never, 5: Often). The possible scores in this questionnaire ranged from 11 to 55. A high score indicates a higher degree of problematic SAB.

2.6.3.2. Peer interaction questionnaire (Parent report)

A peer relationship questionnaire was developed for application in the present thesis (see appendix 8). The peer problems sub-scale within the SDQ provides a strong measure of the quality of a child's peer relationships (Goodman, 1997). The SDQ has been widely administered with both typical and atypically developing populations (Muris et al., 2003; Rhodes et al., 2010; Riby et al., 2013). To the author's awareness, no questionnaires have been developed to investigate peer relationships in children with WS. The present questionnaire aims to build on the SDQ peer problems sub-scale by achieving further insight into the children's inclusion by peers and the appropriateness of their behaviour within these peer interactions. The items in this questionnaire were developed with reference to previous research as detailed below.

Research has provided evidence of peer relationship difficulties among children with WS (Greer et al., 1997). Previous research indicated low levels of social inclusion in adults with WS (Davies et al., 1998; Elison et al., 2010) and children with developmental disorders (Chamberlain, Kasari, & Rotheram-Fuller, 2007). Few researchers have examined social inclusion in children with WS. Taking into account the importance of peer relationships on psychological wellbeing (Liem & Martin, 2011), items were included in this questionnaire to examine the children's extra-curricular engagement with peers.

Previous researchers reported high levels of tactile behaviour in adults with WS which could be suggested to hinder their peer relations (Davies et al., 1998; Elison et al., 2010; Udwin, 1990). In view of more recent findings that children with WS demonstrated difficulties understanding the concept of personal space (Lough et al., 2015), further investigation is sought to identify how these conceptual difficulties impact on their social interactions with peers. Items were incorporated into this questionnaire which measured the children's engagement in tactile behaviour and their awareness of personal space boundaries within peer interactions.

Six items in this scale measure the quality of the children's peer relations. These items generated an acceptable Cronbach's alpha value of .78 when administered in the present thesis. This six item sub-scale was termed 'peer relationship quality'. This sub-scale was negatively correlated with the peer problems sub-scale within the parent rated SDQ (r = -.66, p = .001) which indicated strong construct validity. Three items in this scale measure the children's engagement in tactile behaviour. This three item sub-scale also showed strong internal consistency when administered in the present thesis, with a Cronbach's alpha value of .77. This sub-scale was termed 'tactile behaviour in peer interactions'.

Parents are required to rate the extent to which each statement within this questionnaire applies to their child on a five point Likert scale, where 1 indicates 'Strongly disagree' and 5 indicates 'Strongly agree'. The possible scores in the 'peer relationship quality' sub-scale range from 6 to 30. A higher score indicates a higher quality of peer relationships. The possible scores in the 'tactile behaviour in peer interactions' sub-scale range from 3 to 15. A higher score indicates a higher engagement in tactile behaviour.

2.6.3.3. Peer interaction questionnaire (Teacher report)

Previous researchers have predominantly used parent reports to measure social behaviour in WS (Davies et al., 1998; Doyle et al., 2004). The recruitment of multiple informants can provide valuable insight into the nature of these behaviours across contexts (Cuckle & Wilson, 2002; Klein-Tasman et al., 2011). A 21 item questionnaire was developed for use in the present study to solicit teachers' perceptions regarding the peer relations of the child with WS (see appendix 9). All items in this questionnaire are presented on a five point Likert scale where 1 indicates 'Strongly disagree' and 5 indicates 'Strongly agree'. Additional open ended questions are included throughout the questionnaire to allow the teacher to provide any further information or examples to support their ratings. This questionnaire solicits both quantitative data through the Likert scale items and qualitative data through the open ended questions.

In line with the gaps in research identified in section 2.6.4, items were developed for inclusion in this questionnaire to measure the nature of the child's peer relationships and the appropriateness of their social interactions. The first construct in this scale is 'peer relationship quality'. This is a 12 item construct which assesses the children's friendship patterns and their social inclusion within school. This construct demonstrated strong internal reliability when administered with the participants in the present thesis (α = .78). This construct was negatively correlated with teachers' ratings in the peer problems sub-scale of the SDQ ($r = -.71$, $p = .001$). This correspondence between measures supports the validity of this construct as an assessment of peer relationship quality. Four items were developed to measure the child's engagement in tactile behaviour and the negative impact of this behaviour on peer relations. One of these items was removed ("Other children have reported to dislike this behaviour") as this item was not endorsed by any of the participants and therefore was poorly correlated with the other three items in this construct. The final three item construct 'display of tactile behaviour' showed adequate internal reliability (α = .67). Two items were developed which measured the level of support which the child received within school. One item measured levels of support within the classroom and one item measured levels of support within leisure time. There was a significant positive correlation between these two items ($r = .56$, $p = .020$). The remaining three items in this questionnaire measured emotional difficulties, social anxiety and interaction with strangers. These items did not correlate significantly with the other items in this scale and together produced a very low

Cronbach's alpha value of .18. In consequence, these items were removed from this questionnaire as they were deemed to not provide a reliable measure of peer relationship functioning. The final questionnaire contained 17 Likert scale questions which were intended to gauge three areas of peer interaction: quality of peer relationships, display of tactile behaviour and level of support received.

2.7. Interview instruments

2.7.1. Vineland Adaptive Behavior Scales, Second Edition – Survey Interview Form (Sparrow, Cicchetti, & Balla, 2005)

The Vineland Adaptive Behavior Scales (VABS) is a standardised measure which is frequently employed by educational and clinical psychologists to identify individuals with intellectual and developmental disabilities. This scale provides a measurement of adaptive behaviour and daily living skills across the lifespan (0-90 years). The items in this scale measure five core constructs: communication, daily living skills, socialisation, motor skills and maladaptive behaviour. The VABS is a paper based task which is administered in the format of an interview. This measure has an estimated completion time of between 20 and 60 minutes for the full interview. Due to the lengthy nature of this interview and the broad scope of behaviours measured, only the socialisation domain of this interview will be administered in the present thesis as this is the focus of investigation. The socialisation construct measures the children's behaviour across three sub-domains: interpersonal relationships, play and leisure time and coping skills. The participants' scores across these three sub-domains are combined to form a domain composite score. Previous research reported that the VABS socialisation domain demonstrated high internal reliability with split half reliability scores which ranged between .84 to .93, strong test-retest reliability with correlation values which ranged between .76 to .92, and strong inter-rater reliability with correlation values which ranged between .71 to .81 (Sparrow et al., 2005). This measure showed strong internal consistency when administered with the participants in the current thesis (α = .76).

2.7.2. Newly developed interview schedules

Several interview schedules were developed for use in the present thesis. A multi-informant approach was used. The children with WS and their parents were interviewed. The construction of these interview schedules is detailed in section 2.7.2.1, 2.7.2.2 and 2.7.2.3.

2.7.2.1. Social approach interview schedule (Parent informant)

A semi-structured interview was developed for use in the present research to solicit qualitative information from parents regarding their child's SAB (see appendix 7). In line with the gaps in research identified in section 2.6.3, items aimed to measure three aspects of SAB: the manifestation of SAB in WS, the wider impact of SAB on the child and family, and stranger danger awareness.

2.7.2.2. Peer relationships interview schedule (Parent informant)

A semi-structured interview was developed for use in the present thesis (see appendix 10). This interview schedule was an adapted version of the friendship interview schedule administered by Cuckle and Wilson (2002) in their assessment of friendships in adolescents with Down’s syndrome. This interview schedule was modified in order to meet gaps in the research literature and tap into specific aspects of the WS social phenotype. Cuckle and Wilson (2002) used a single item to measure social inclusion. Taking into consideration the high levels of social isolation reported among children and adults with WS (Davies et al., 1998; Elison et al., 2010; Greer et al., 1997), additional items were developed for inclusion in this interview schedule to gain further insight into the nature of these peer relationship difficulties and the children’s extra-curricular engagement with peers. Cuckle and Wilson (2002) measured the number of extra-curricular clubs and activities attended by the child. Taking into account the social impairments and peer difficulties reported among children with WS (Greer et al., 1997; Klein-Tasman et al., 2011) additional items were incorporated into this interview schedule to gain further insight into the children’s experiences of these clubs, with a focus on the children’s peer relationships and inclusion at these clubs. Finally research has identified substantial communication difficulties (Laws & Bishop, 2004; Stojanovik, 2006) and disinhibition within interactions among individuals with WS (Davies et al., 1998; Elison et al., 2010). Additional items were incorporated in this interview schedule to further tap into these difficulties within the context of peer relations.

The final interview schedule contained 14 broad questions with additional probe questions. These questions aimed to measure six areas of peer interactions: the nature of the child’s peer relationships, frequency of contact with peers, extra-curricular engagement, social inclusion, communication skills and the appropriateness of social behaviour.

2.7.2.3. Peer relationship interview schedule (Child informant)

A semi-structured interview was developed for use in the present thesis to solicit the children’s perceptions of their own peer relationships (see appendix 11). This interview was a modification of the friendship interview schedule developed by Cuckle and Wilson (2002) in their assessment of friendships in adolescents with Down’s syndrome. Additional items were incorporated into this interview schedule in order to address key gaps in research. The present measure aimed to gain insight into the children’s perceptions of their peer relationships. This modified interview contained 13 questions, with additional probe questions. The questions in this interview aimed to measure the children’s perceptions of the quality of their peer relationships, their extra-curricular engagement and their understanding of friendships. Through a consideration of the social isolation reported in WS (Elison et al., 2010; Greer et al., 1997), additional items were included which tapped into the children’s social inclusion both in and out with school.

Chapter 3: Social approach behaviour in children with Williams syndrome: A quantitative study

3.1. Introduction

WS can be characterised by high levels of interest in unfamiliar people and an engagement in indiscriminate SAB (Jones et al., 2000; Martens et al., 2009). This strong social drive has been found to develop early in infancy and to be present across the lifespan (Bellugi et al. 1999; Doyle et al., 2004; Jones et al., 2000; Martens et al., 2009; Mervis et al., 2003). The accumulated effect of mild to moderate intellectual delay (Searcy, Lincoln, Rose, Klima, Bavar, & Korenberg, 2004), atypicalities within social functioning (Klein-Tasman et al., 2011) and poor stranger danger awareness (Riby et al., 2014) could be suggested to account for the high levels of social vulnerability which have been reported in children with WS (Fisher et al., 2013). This heightened social vulnerability reinforces the importance of conducting further research to expand our understanding of the prevalence and manifestation of atypical SAB in children with WS.

Within this chapter, Adolph's Approachability Task will be administered to experimentally measure levels of SAB in children with WS. Comparisons will be drawn between the performance of the children with WS, TD children who have been matched on verbal ability and TD children who have been matched on non-verbal ability. Furthermore, a parent questionnaire will be used to solicit parents' perceptions of the prevalence and characteristics of their child's SAB.

Several methodologies have been used to examine SAB in WS (Bellugi et al., 1999; Dodd et al., 2010; Doyle et al., 2004). The strengths and limitations of these methods should be considered when interpreting the research findings. Adolph's Approachability Task (Adolphs et al., 1998) is a standardised experimental instrument which has been used to measure SAB in children and adults with WS (Bellugi et al., 1999; Järvinen-Pasley et al., 2010; Martens et al., 2009). Järvinen-Pasley et al. (2010) recruited a sample of individuals with WS (13 – 52 years) and a TD comparison group who were matched on chronological age. The participants completed Adolph's Approachability Task. Within this task, the individuals were presented with black and white images of unfamiliar people and were asked to rate the extent to which they would like to approach the person depicted within each image on a five point Likert scale. The individuals with WS indicated a significantly higher willingness to approach the person depicted within the image than the TD controls. These findings of high levels of SAB were in line with previous research evidence (Bellugi et al., 1999; Martens et al., 2009). However, across these research studies, the individuals with WS were compared to a TD comparison group who were matched on chronological age. It is therefore difficult to draw conclusions regarding whether this atypical SAB is a consequence of the individuals' intellectual delay or is a specific result of the WS phenotype.

While Adolph's Approachability Task has generated support for atypically high levels of SAB in WS, researchers have raised concerns regarding the validity of this task. Fisher et al. (2014) examined levels of correspondence between performance in Adolph's Approachability Task, parent questionnaire ratings and real-life behavioural observations of SAB in a sample of young adults with WS. Fisher et al.

reported that the participants' performance in Adolph's Approachability Task was poorly correlated with both parents' questionnaire ratings (*rho* = .23, *ns*) and behavioural observations of SAB (*rho* = .01, *ns*). These findings challenge the validity of Adolph's Approachability Task as a measurement of SAB in WS. Adolph's Approachability Task requires the participants to form judgements of approach based on black and white static images. It could be argued that due to the artificial nature of this task, this may not provide an accurate measurement of SAB in WS.

Parental ratings have been reported to correspond strongly with real-life observations of SAB in WS (r*ho* = .45, *p* < .005) (Fisher et al., 2014). Parent reports could therefore be suggested to provide a more reliable measurement of SAB. Previous researchers used the standardised 'Salk's Institute Sociability Questionnaire' (SISQ) to measure SAB in WS. Doyle et al. (2004) administered the SISQ to the parents of a sample of children with WS, DS and TD children. All children were between the ages of 1 and 12 years old. There was a significant effect of participant group on SAB (p < .001). The children with WS were reported to demonstrate significantly higher levels of SAB towards strangers than children with DS and TD children who were matched on chronological age. Järvinen-Pasley et al., (2010) further reported that individuals with WS (13 – 52 years) were rated by their parents to engage in significantly higher levels of SAB towards unfamiliar people than a TD comparison group that was matched for chronological age (p < .001).

Previous research has generated strong support for high levels of SAB in individuals with WS when compared to TD comparisons and groups with other developmental disorders (Bellugi et al., 1999; Järvinen-Pasley et al., 2010; Martens et al., 2009).

Although previous researchers (Little et al., 2013; Porter & Coltheart, 2005) identified within-syndrome variance in levels of SAB in WS, few researchers have examined this variance in detail. Through a consideration of previous findings of a heterogeneous profile of cognition and behaviour in WS (Little et al., 2013; Porter & Coltheart, 2005), there is a need to further develop these findings by examining the within-syndrome variance in the manifestation and impact of SAB among children with WS.

Children with WS have been reported to demonstrate substantial impairments within social functioning (Klein-Tasman et al., 2011). Very little research thus far has examined the implications of these impairments for the children's interactions with strangers. Riby et al. (2014) examined the relationship between social functioning and stranger danger awareness in children with WS (8 – 17 years). The children completed a video vignette task. The vignettes depicted several social scenarios where a child was approached by a stranger. After viewing each video, a series of questions was posed to the child. The participants were asked if the child in the video clip should help the stranger and if they themselves would help the stranger. The children were asked to provide justifications for their responses. The children's responses in this task were coded in order to generate a rating of stranger danger awareness. Parents also completed the standardised SDQ which provided a measurement of social functioning. There was a significant negative relationship between the parents' ratings of peer problems and levels of stranger danger awareness among the children with WS. Riby et al. suggested that peers may have an important influence in a child's understanding of socially appropriate behaviour within interactions. While Riby et al. provided insight into the relationship between social functioning and stranger danger awareness, further research is required to directly examine the relationship between social functioning and levels of SAB in children with WS.

This study aims to examine the characteristics of SAB in children with WS, including the manifestation of SAB and the impact of SAB on the child and family. The present study will develop the methodologies of previous research which has predominantly used a single informant approach when examining SAB in WS (Doyle et al., 2004; Porter et al., 2007) by soliciting information from the children with WS and their parents. Furthermore, the present study will compare the children's performance in Adolph's Approachability Task to TD comparison groups who have been matched on verbal and non-verbal ability. This will allow more reliable conclusions to be drawn on the role of intellectual delay in the high levels of SAB demonstrated in children with WS which previous researchers were unable to examine. Finally, the present study aims to further develop the findings of Riby et al. (2014) by directly examining the relationship between peer relations, social functioning and SAB in children with WS.

3.2. Research question and hypotheses

The present study aims to address the research question 'What are the characteristics of SAB in children with WS?'

The current study will test three hypotheses:

1. The children with WS will demonstrate significantly higher levels of SAB in Adolph’s Approachability Task than TD children matched on verbal ability and TD children matched on non-verbal ability.
2. There will be a significant negative relationship between peer relationship quality and levels of SAB in the children with WS.
3. There will be a significant positive relationship between social functioning impairments and levels of SAB in the children with WS.

3.3. Method

3.3.1. Design

Hypothesis 1: An independent groups design was employed. The independent variable (IV) was participant group. This IV had 3 levels: children with WS, TD children matched on verbal ability and TD children matched on non-verbal ability. The dependent variable (DV) was level of SAB as measured in Adolph’s Approachability Task. The DV was measured on an interval scale.

Hypotheses 2 and 3: A correlational design was recruited in order to examine the relationships between peer relations (parent rated peer problems – SDQ, teacher rated peer problems – SDQ, parent rated peer relationship quality, teacher rated peer relationship quality), social functioning (Social Responsiveness Scale) and SAB (Adolph’s Approachability Task & parental ratings of SAB). All variables were measured on an interval scale.

3.3.2. Participants

21 children with WS aged between 7.5 and 16.49 years (M: 11.83, SD: 2.69; 12M, 9F) participated. The Adolph's Approachability Task responses of one child were removed prior to analysis due to the child's difficulties in following the verbal task instructions. The mother (N=19) or father (N=2) of the children with WS also participated. The class teacher of 18 of the children with WS participated. All of the children with WS had previously received a formal diagnosis of WS which had been confirmed through positive genetic fluorescent *in situ hybridisation* testing (FISH test).

40 TD children were recruited from primary schools in central Scotland. 20 of these children were individually matched with the children with WS on verbal ability on the BPVS II (Dunn et al., 1997) (5.08 – 7.85 years, M: 6.47; SD: .94, 11M, 9F). There was no significant difference between the children with WS and TD controls in their scores on the BPVS II, *t* (38) = .52, *p* = .608 (two tailed). 20 TD children were individually matched with the children with WS on non-verbal ability on RCPM (Ravens et al., 1998) (5.08 – 7.96 years; M: 6.15; SD: .91; 9M, 11F). There was no significant difference between the children with WS and TD controls in their scores on RPCM, *t* (38) = -.08, *p* = .934 (two tailed). All of the TD children scored within the acceptable range on the SDQ which assessed for the presence of any mental or behavioural disorders.

3.3.3. Measures

3.3.3.1. Adolph's Approachability Task (Adolph et al, 1998)

A modified version of the standardised Adolph's Approachability Task was used. This task contains 20 experimental trials. In 10 trials, an unfamiliar male adult is presented and in 10 trials an unfamiliar female adult is presented. All faces depicted a neutral expression. Coloured photographs were used, in contrast to the black and white photographs used in the original version of this task. In each experimental trial the child is asked to rate the extent to which he or she would like to approach and converse with the person depicted. The children are required to rate their response on a five point Likert scale, where higher ratings indicate a greater desire to approach the person. The possible score range in this task was a minimum of 20 and maximum of 100.

3.3.3.2. Social approach questionnaire (Parent report)

This 11 item parent report questionnaire was designed to measure the manifestation of SAB, perceived vulnerability of the child and the impact of SAB on daily/family life. This scale showed strong internal consistency when applied in the current thesis ($\alpha = .80$). Parents are asked to rate the extent to which each item in this scale applies to their child (1: Strongly disagree, 5: Strongly agree) or the frequency with which their child engages in this behaviour (1: Never, 5: Often). The possible scores in this questionnaire ranged from 11 to 55. A high score indicates a higher degree of problematic SAB.

3.3.3.3. Social Responsiveness Scale (SRS) (Constantino & Gruber, 2012)

The SRS is a 65 item questionnaire which measures social functioning across five domains: social awareness, social cognition, social communication, social motivation and autistic mannerisms. Scores are generated for each domain and together with an overall score which indicates the severity of the child's social impairments. The parent informant is required to rate the extent to which each statement is applicable to their child on a four point Likert scale, where 1 indicates 'not true' and 4 indicates 'almost always true'. The participants' ratings in this questionnaire demonstrated strong internal reliability (α = .84).

3.3.3.4. Strengths and Difficulties Questionnaire (Goodman, 1997)

The SDQ is a 25 item scale which assesses five constructs of social functioning: emotional symptoms, conduct problems, hyperactivity/inattentiveness, peer problems and prosocial behaviour. The parent or teacher informant is asked to rate the extent to which each of the items applies to the child. All responses are rated on a 3-point Likert scale, where 0 indicates 'not true' and 2 indicates 'certainly true'. Scores are generated for each sub-scale and together with a composite score termed the 'total difficulties score'. This questionnaire demonstrated adequate internal reliability when administered with the parents (α = .69) and teachers (α =.64) in the current study.

3.3.3.5. Peer relationship quality questionnaire (Parent report)

This six item questionnaire measures the quality of the child's peer relationships and his or her social inclusion with peers. Parents are asked to rate the extent to which the items apply to their child. All responses are rated on a five-point Likert scale, where 1 indicates 'strongly disagree' and 5 indicates 'strongly agree'. The possible scores in this questionnaire range from 6 – 30. A high score indicates a stronger quality of peer relations. This scale demonstrated strong internal consistency when administered with the parents in the present study ($\alpha = .78$).

3.2.3.6. Peer relationship quality questionnaire (Teacher report)

This 12 item questionnaire measures the quality of the child's peer relationships and his or her social inclusion by peers. Teachers are asked to rate the extent to which the items apply to the pupil. All responses are rated on a five-point Likert scale, where 1 indicates 'strongly disagree' and 5 indicates 'strongly agree'. The possible scores in this scale range from 12 to 60, with higher scores indicating stronger peer relationships. This scale showed strong internal reliability for the current participant sample ($\alpha = .78$).

3.3.4. Procedure

The children with WS and their parent were visited at home where all measures were administered. The TD children were visited at school. Both the children with WS and the TD children completed Adolph's Approachability Task. In addition, the parents of the children with WS completed questionnaires which assessed their child's SAB and aspects of their social functioning. These questionnaires were not completed by the parents of the TD children. The teachers of both the children with WS and the TD children completed questionnaires on the children's social functioning which were posted out to the school.

3.4. Results

3.4.1. 'What are the characteristics of SAB in children with WS?'

Table 1 provides the mean, standard deviation (SD) and range for each individual item in the parent social approach questionnaire for the sample of children with WS, together with the mean total score. All items were rated on a scale from 1 to 5. The possible range of scores in this scale was a minimum score of 11 and a maximum score of 55. Higher scores in this questionnaire indicated a higher engagement in SAB.

Table 1:

Mean, SD and range of scores among the children with WS in the parent social approach questionnaire (N=21)

	Mean rating (SD)	***Range***
Approached a stranger in a public place	4.10 (1.04)	2 – 5
Tactile behaviour towards unfamiliar people	3.19 (1.40)	1 – 5
Tactile behaviour towards adults in authority	3.76 (1.04)	1 – 5
Interest in speaking with unfamiliar adults	4.76 (0.44)	4 – 5
Interest in speaking with unfamiliar children	4 (0.89)	2 – 5
Requires constant supervision in public	4.67 (0.73)	2 – 5
SAB is a parental concern	4.57 (0.68)	3 – 5
SAB impacts on the family	4.05 (0.80)	2 – 5
Stranger danger awareness	3.71 (1.01)	2 – 5
Placed in an awkward/vulnerable situation in the past as a result of SAB	3.76 (1.09)	2 – 5
Would follow a stranger if invited	4.52 (0.68)	3 – 5
Very trusting	4.67 (0.48)	4 – 5
Total SAB	39.10 (4.31)	33 – 55

As shown in table 1, the children with WS were reported by their parents to show high levels of SAB. There was, however, within-syndrome variance in the levels of SAB and impact of SAB on daily functioning. A closer inspection of the scores for the individual questionnaire items provides further insight into this within-syndrome variance. The highest rated items were a strong interest in speaking with unfamiliar adults and high levels of trust, while engagement in tactile behaviour and levels of stranger danger awareness showed the greatest variance in the children with WS.

3.4.2. Hypothesis 1: The children with WS will demonstrate significantly higher levels of SAB in Adolph's Approachability Task than TD children matched on verbal ability and TD children matched on non-verbal ability.

The descriptive statistics for the children's ratings in Adolph's Approachability Task are presented in table 2. The children rated the faces on a five point Likert scale from 1 ('I definitely would not approach') to 5 ('I definitely would approach').

Table 2:

Adolph's Approachability Task performance (Mean, SD and range) for the children with WS and TD comparison groups

	Williams syndrome group (N=20)		TD verbal comparison group (N=20)		TD non-verbal comparison group (N=20)	
	Mean (SD)	Range	Mean (SD)	Range	Mean (SD)	Range
Adolph's Approachability Task (All faces)	3.48 (.81)	2.05 – 4.55	2.56 (1.23)	1.00 – 4.95	2.54 (1.21)	1.00 – 3.95
Adolph's Approachability Task (Female faces)	3.37 (.93)	1.70 – 5.00	2.57 (1.17)	1.00 – 4.97	2.41 (1.26)	1.00 – 5.00
Adolph's Approachability Task (Male faces)	3.59 (.87)	2.00 – 5.00	2.54 (1.19)	1.00 – 5.00	2.67 (1.43)	1.00 – 5.00

As indicated in table 2, the children with WS showed a higher intention to approach unfamiliar adults than both TD comparison groups. All children rated the male and female faces similarly.

A one way between groups analysis of variance (ANOVA) was conducted to examine the effect of participant group on performance in Adolph's Approachability Task. The data met the parametric assumption of an interval scale of measurement. An analysis of skewness indicated that the data showed a normal distribution. An observation of Q-Q plots also indicated the data points to lie close along a straight line and there to be no significant outliers. Levene's test was conducted to test the assumption of homogeneity of variance. The variance in SAB scores was equal between groups, F (2, 57) = 1.77, p = .180, therefore the assumption of homogeneity of variance was met.

There was a significant main effect of participant group on performance in Adolph's Approachability Task, F (2, 57) = 5.10, p = .009, $\eta 2$ = .15 A post hoc Tukey HSD test showed that the WS group rated the faces as significantly more approachable than did the TD verbal matched controls (p = .022) and the TD non-verbal matched controls (p = .019). There was no significant difference between the verbal and non-verbal matched controls in their performance in Adolph's Approachability Task (p = .999). The findings provide support for the first hypothesis of this study that children with WS will show higher levels of SAB within Adolph's Approachability Task when compared to TD children matched on verbal ability and TD children matched on non-verbal ability[1].

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[1] Additional analysis was conducted to examine the effect of gender of the face on the participants' performance in Adolph's Approachability Task, however, this proved non-significant, F (1, 57) = 1.94, p = .169.

3.4.3. Hypothesis 2: There will be a significant negative relationship between peer relationship quality and levels of SAB in the children with WS.

Parents' ratings of their child's peer problems (SDQ) were non-normally distributed. In consequence, Spearman's rank order correlations were conducted to examine the relationship between this variable and the children's performance in Adolph's Approachability Task and parents' ratings of SAB among the children with WS. There were no significant relationships between these variables ($p > .05$) and therefore this will not be reported further.

For all other variables, the data met the parametric assumptions of an interval scale of measurement, no significant outliers, linearity of variables and normally distributed data points. Bivariate Pearson's correlations were conducted to examine for a relationship between three measures of peer relations (teacher rated peer problems - SDQ, parent rated peer relationship quality and teacher rated peer relationship quality) and the children's SAB (performance in Adolph's Approachability Task and parent rated SAB). As a small number of planned tests were conducted, the standard alpha value of $p < .05$ was used as the test criterion. The correlation output is presented in table 3

Table 3:

Bivariate Pearson's correlations between parents' and teachers' ratings of peer relations and levels of SAB in children with WS (N = 21)

	1	2	3	4	5
1. Teacher rated peer problems (SDQ)		-.59**	-.71**	-.05	.08
2. Parent rated peer relationship quality			.66**	-.29	-.12
3. Teacher rated peer relationship quality				-.22	-.28
4. Adolph's Approachability Task					-.18
5. Parent rated SAB					

* $p < .05$, ** $p < .01$, *** $p < .001$

There were no significant relationships between parents' and teachers' ratings of peer relations and levels of SAB in the children with WS as measured in Adolph's Approachability Task and parents' questionnaire ratings ($p > .05$).

3.4.4. Hypothesis 3: There will be a significant positive relationship between social functioning impairments and levels of SAB in the children with WS.

The data met the parametric assumptions of an interval scale of measurement, no significant outliers, linearity of variables and normally distributed data points. In consequence, Bivariate Pearson's correlations were conducted to examine the relationships between indices of social functioning as measured in the SRS (social awareness, social cognition, social communication, social motivation and autistic mannerisms), the children's total score in the SRS, and the children's levels of SAB (performance in Adolph's Approachability Task and parent rated SAB). As a small number of planned tests were conducted, the standard alpha value of $p < .05$ was used as the test criterion. The correlation output is presented in table 4

Table 4:

Bivariate Pearson's correlations between parents' ratings of social impairment (SRS) and levels of SAB in the children with WS (N=21)

	1	2	3	4	5	6	7	8
1. Social awareness (SRS)		.64**	.48*	.29	.49*	.51**	.45*	-.10
2. Social cognition (SRS)			.59**	.46*	.52**	.63**	.45*	.38*
3. Social communication (SRS)				.78***	.55**	.83**	.13	-.15
4. Social motivation (SRS)					.65**	.88***	-.23	.16
5. Autistic mannerisms (SRS)						.72***	.22	.27
6. Total score (SRS)							.09	.15
7. Adolph's Approachability Task								-.18
8. Parent rated SAB								

p* < .05, ** *p* < .01 * *p* < .001

There was a significant positive relationship between impairments in social awareness and parent rated SAB (*p* = .021). There were significant positive relationships between impairments in social cognition and the children's ratings in Adolph's Approachability Task (*p* = .047) and parent rated SAB (*p* = .020).

3.5. Discussion

3.5.1. Summary of findings

The accumulated findings indicate high levels of SAB in children with WS. This was demonstrated in Adolph's Approachability Task and in the parental questionnaire ratings. This suggests that high levels of SAB are a core feature of the WS social phenotype. This builds on previous research which predominantly used a single informant approach (Bellugi et al., 1999; Martens et al., 2009). However, there was within-syndrome variance in the manifestation of this SAB and severity levels among the children with WS. Impairments in social functioning, specifically deficits in social cognition and social awareness were significantly associated with an engagement in high levels of SAB in WS, providing support for this hypothesis. The findings within this chapter will be evaluated in further depth in this discussion, with note to their contribution to this field of research.

3.5.2. What are the characteristics of SAB in children with WS?

Previous researchers primarily analysed mean scores to compare levels of SAB between individuals with WS and TD children and groups with other developmental disorders. Although previous research (Doyle et al., 2004) has reported within-syndrome variance in SAB, few investigators have examined this in detail. The present findings provide further insight into this within-syndrome variance. All of the parents of the children with WS reported that their child showed a strong interest in speaking with unfamiliar adults and was very trusting of other people. These items were rated highly for all children and adolescents with WS in this sample. This suggests that these are prominent features of the WS social phenotype and that they persist across development. This finding provides supports for previous research which indicated that children with WS demonstrated a strong drive to approach and converse with unfamiliar adults (Bellugi et al., 1999; Doyle et al., 2004).

An interesting finding from the present study was the discrepancy between the children's interest in speaking with unfamiliar adults and the children's interest in speaking with unfamiliar children. The children with WS were reported to show a higher level of interest in speaking with unfamiliar adults compared to unfamiliar children. This suggests that some children with WS do show discrimination in their SAB.

This discrimination in approach behaviour challenges the theoretical assumptions of the frontal lobe hypothesis as these children were able to inhibit their approach behaviour towards children. Caution should be taken as this data is based on experimental ratings and parents' perceptions of SAB. This account should be further tested by obtaining behavioural observations of the children's social interactions with unfamiliar adults and children. The present finding suggests that adults are particularly salient to children with WS. This is in line with previous research which reported that adults with WS showed a preference for older individuals over their peers (Udwin, 1990). This suggests that this social drive to engage with older individuals is present across children with WS, as well as adults with WS. While some children demonstrated discrimination and only approached adults, other children were reported to show an interest in speaking with both children and adults. This builds on previous findings of within-syndrome variance in the social behaviour of individuals with WS (Little et al., 2013; Lough et al., 2016) by providing insight into the aspects of SAB which demonstrate variability. This variability will be further examined in chapter 5 by soliciting parents' observations on their child's SAB through semi-structured interviews.

It is important to acknowledge the presence of within-syndrome variance in the display of SAB in WS, with this having potentially important implications for the design of interventions and support. However, caution should be taken when drawing conclusions on within-syndrome variance in WS as this distribution was not subjected to statistical analysis. Furthermore, as the parental social approach questionnaire was developed for use in the present thesis, the absence of normative data and scaling reinforces the need for further caution to be taken when drawing interpretations regarding levels of within-syndrome variance.

While previous researchers reported that adults with WS engaged in a high level of tactile behaviour towards strangers (Davies et al., 1998), there was variance in the level of tactile behaviour and the target of the tactile behaviour demonstrated by the children with WS in the current study. While many parents reported that their child would not randomly hug a stranger in the street, several of the children were reported to engage in tactile behaviour towards adults in positions of authority which included teachers and health practitioners, indicating that there is a motivation to engage in tactile displays. It could be suggested that this high drive to engage in physical contact with other people is present in WS; however, may be restricted by high levels of parental supervision which minimise the contexts in which this is manifested. All of the children with WS were reported to be highly trusting of others and the majority of parents believed that their child would be easily led by a stranger.

Many of the children with WS were reported to show an awareness of stranger danger. This contrasted with the previous findings of Riby et al. (2014) who reported significant impairments in stranger danger awareness among children with WS. This discrepancy in the children's levels of stranger danger awareness could be accounted for by differences in the measurement of stranger awareness. Riby et al. measured stranger awareness by administering a video vignette task where the children were required to apply their understanding of stranger danger across different contexts. In contrast, the parents in the present study rated their child's stranger awareness on a Likert scale based on their own perceptions. Further research is sought to tap into stranger danger awareness in WS and account for this discrepancy in findings.

3.5.3. Adolph's Approachability Task (Hypothesis 1)

As predicted, the children with WS demonstrated significantly higher levels of SAB towards unfamiliar people than did TD children who were matched on verbal ability and TD children who were matched on non-verbal ability. The findings provide support for previous evidence that children with WS are driven to approach individuals indiscriminately (Bellugi et al., 1999; Jones et al., 2000; Martens et al., 2009). Importantly, the present study addresses the methodological limitations of previous research (Järvinen-Pasley et al., 2010; Martens et al., 2009) by directly comparing the children with WS to TD children who were individually matched on verbal and non-verbal ability. The children with WS showed significantly higher levels of SAB than the comparison groups when intellectual ability was controlled for. These findings suggest that the high levels of SAB demonstrated by children with WS cannot be attributed to a general intellectual delay alone, with other components of the WS disorder such as response inhibition impairments and attention bias playing a potential role in this behaviour.

Several previous researchers recruited a wide age range, incorporating both children and adults with WS (Bellugi et al., 1999; Järvinen-Pasley et al., 2010; Martens et al., 2009). This is a common practice in research involving rare populations given the restrictions on recruitment; however, a limitation of recruiting a wide age range is that there may be variance in the nature of SAB within this sample which can distort the findings. The present study focused on children with WS and therefore allowed conclusions to be drawn regarding the atypical nature of SAB in this period of development.

3.5.4. Relationships between social functioning, peer relations and SAB in children with WS (Hypothesis 2 & 3)

Previous researchers (Riby et al., 2014) identified a significant negative relationship between peer problems and stranger danger awareness in children with WS. This was in line with previous findings where peers were reported to play a protective role in minimising a child's social vulnerability (Fisher et al., 2013).

The present study is the first to the author's knowledge which has directly examined the relationships between social functioning, peer relations and SAB in children with WS. There was a significant positive relationship between impairments in social awareness and an engagement in inappropriate SAB in children with WS. Social awareness concerns an individual’s understanding of social norms and rules and the ability to detect appropriate social cues within interactions (Constantino & Gruber, 2012). It could be suggested that some children with WS may engage in disinhibited social interactions towards unfamiliar people due to a difficulty detecting social cues and applying this knowledge within social interactions.

There was also a significant positive relationship between impairments in social cognition and SAB in the children with WS. During typical development, children often make reference to a person’s facial expression or tone of voice in order to form a judgement on whether to approach that person (Meyer-Lindenberg et al., 2005). This interpretation of social cues is vitally important as it provides a child with the necessary resources to identify if he or she is being taking advantage of and promotes safe social practices. Previous research has demonstrated that children with WS experience significant social cognitive impairments (Porter et al., 2008; Tager-Flusberg et al., 2006; Tager-Flusberg & Sullivan, 2000), with these difficulties in interpreting social cues and information often impeding successful social interactions and potentially accounting for their high engagement in SAB. This inappropriate SAB could be attributed to the child’s impairments extracting and interpreting relevant social information within an interaction. The children with WS may be unable to interpret another person’s discomfort at their disinhibited behaviour. The children with WS may also violate social norms regarding personal space boundaries due to difficulties understanding rules and regulations of socially appropriate behaviour. These findings have provided support for the role of social functioning in the high levels of SAB demonstrated by children with WS and provided support for the hypothesis of this study that there would be a significant positive relationship between social functioning impairments and SAB.

There was no significant relationship between the quality of the children's peer relationships and the children's levels of SAB as measured in Adolph's Approachability Task and parental questionnaire ratings. Riby et al. (2014) reported a relationship between peer relations and the children's understanding of appropriate practices within social interactions. The current study identified poor correspondence between the children's understanding of stranger danger principles and their practice of these behaviours. Although many parents reported that their child had an understanding of stranger danger principles, the majority of the children did not apply these principles within social interactions. With consideration to this, it could be suggested that while friendships may facilitate a child's understanding of socially appropriate behaviour, they may be unable to translate this understanding into practice.

Due to the nature of correlational analysis, while conclusions can be drawn on the association between atypicalities in social functioning and high levels of SAB towards strangers, correlational analyses do not allow conclusions to be drawn on causality (Fields, 2009). Within future work, researchers should seek to recruit a larger sample size. Increasing the sample size will allow more powerful statistics to be conducted, which can investigate and draw conclusions on the predictive nature of the relationship between these variables.

3.5.5. Conclusions

This study has expanded our knowledge of SAB in children with WS by soliciting information from children with WS and their parents. This advances the methods of previous researchers who predominantly used a single informant methodology. The children with WS indicated significantly higher levels of SAB in Adolph's Approachability Task than TD children matched on verbal ability and TD children matched on non-verbal ability. Furthermore, many of the children with WS were reported by their parents to demonstrate high levels of SAB. However, there was within-syndrome variance in the manifestation of this SAB. While the children with WS were rated highly in their interest in approaching and conversing with strangers, only a sub-sample of children with WS demonstrated disinhibited tactile behaviour towards these strangers. The children's engagement in SAB was found to be significantly associated with the children's impairments in social awareness and social cognition, providing support for the hypothesis that impairments in social functioning are associated with high levels of SAB in WS.

Chapter 4: Response inhibition and social information processing in the social approach behaviour of children with Williams syndrome

4.1. Introduction

Many children with WS are reported to demonstrate excessive friendliness within their daily social interactions (Jawaid et al., 2012) and to engage in high levels of SAB. These high levels of SAB in WS have been demonstrated using a range of methodologies including experimental rating tasks, parent report questionnaires and behavioural observations (Bellugi et al., 1999; Dodds et al., 2010; Doyle et al., 2004; Fisher et al., 2013; Järvinen-Pasley et al., 2010; Martens et al., 2009; Riby et al., 2013). As reported in chapter 3, the children with WS in the current thesis indicated significantly higher levels of SAB in Adolph's Approachability Task when compared to TD children who were matched on verbal ability and TD children who were matched on non-verbal ability. These high levels of SAB were further illustrated within the parental questionnaire ratings, where many of the children with WS were reported to engage in high levels of indiscriminate friendliness. This was reported to be manifested verbally through the children's interest in conversing with strangers and physically through the children's disinhibited physical interactions with strangers.

Three key theoretical accounts: the frontal lobe hypothesis, the amygdala hypothesis and the social salience hypothesis, have been proposed to account for these high levels of SAB in WS. The frontal lobe hypothesis proposes that high levels of SAB in WS are a result of deficits in response inhibition which result from underlying impairment to the frontal lobes. The frontal lobe hypothesis proposes that individuals with WS struggle to physically inhibit their drive to approach strangers due to this EF deficit (Little et al., 2013; Porter et al., 2007). These EF impairments have been widely reported in WS (Carney et al., 2013; Porter et al., 2007; Rhodes et al., 2010). Patients with frontal lobe brain damage have also been reported to demonstrate similar patterns of social behaviour to children with WS, with both groups displaying indiscriminate friendliness towards strangers (Bruce et al., 2009).

As an alternative theoretical account, the amygdala hypothesis argues that impairments in emotion recognition are implicated in the high levels of SAB shown in WS. The amygdala hypothesis proposes that high levels of SAB in WS are a result of the individual's impairments interpreting negative emotions, in particular threat. In consequence, individuals with WS are proposed to approach other people indiscriminately due to this poor threat detection (Little et al., 2013; Porter et al., 2007). This hypothesis is supported by neural findings of atypical activation of the amygdala in WS (Hass et al., 2009; Meyer-Lindenberg et al., 2005).

Finally, the social salience hypothesis proposes that individuals with WS are driven to approach and engage with other people due to their atypically high level of interest in social stimuli, specifically positive stimuli (Frigerio et al., 2006; Porter et al., 2007). This has been termed a 'social stimulus attraction' (Frigerio et al., 2006). Frigerio et al. (2006) administered Adolph's Approachability Task to a sample of children and young adults with WS and TD controls who were matched on mental age and chronological age. The individuals were presented with images of faces which depicted a range of emotional expressions. Frigerio et al. reported that the individuals with WS indicated a higher intention to approach the people depicted by the happy faces when compared to the ratings provided by TD controls. Furthermore, the individuals with WS reported a significantly lower desire to approach the people depicted by the negative faces when compared to the ratings provided by TD controls.

Only one group of researchers thus far have tested the theoretical assumptions of the frontal lobe hypothesis and the amygdala hypothesis as an account for atypical SAB in WS. Little et al. (2013) recruited a sample of children with WS aged between 6 and 16 years. The children completed the standardised Sun Moon Stroop task which measured response inhibition performance and an emotion labelling task which assessed emotion recognition skills. The children also completed Adolph's Approachability Task (Adolph et al., 1998) which provided a measurement of their SAB. Little et al. reported that a sub-sample of children with WS who performed the poorest in the Sun-Moon Stroop task also demonstrated the highest ratings of SAB. However, Little et al. reported that there was no significant relationship between the children's performance in the emotion labelling task and their SAB. The findings of Little et al. have provided empirical support for the theoretical assumptions of the frontal lobe hypothesis as an account for the high levels of SAB in WS.

However, Adolph's Approachability Task has received criticism regarding the validity of this task as a measurement of SAB. Fisher et al. (2014) examined SAB in young adults with WS using multiple methods and informants. Fisher et al. reported that there was poor correspondence between the individuals' ratings in Adolph's Approachability Task and behavioural observations of the individuals' interactions with strangers in a community setting (*rho* = .01, *ns*). In contrast, Fisher reported that parent ratings of SAB were strongly correlated with behavioural observations of the individuals’ interactions with strangers ($rho = .45$, $p < .005$). These findings indicate the value of utilising an additional measurement of SAB, for example parental ratings, when investigating the relationship between response inhibition performance and SAB in children with WS. This can provide a stronger examination of the strength of this association.

Previous experimental and neural findings have provided support for the role of response inhibition in the high levels of SAB demonstrated in WS (Bruce et al., 2009; Little et al., 2013; Porter et al., 2007). However, response inhibition impairments have been reported across neurodevelopmental disorders such as ADHD (Wåhlstedt, Thorell, & Bohlin, 2008; Willcutt et al., 2005) and ASD (Uzefovsky et al.., 2016). Researchers directly compared the EF performance of children with WS and ADHD and identified substantial overlap between these groups (Rhodes et al., 2011). In particular, children with WS and ADHD both demonstrate impairments in response inhibition (Porter et al., 2007). Despite this shared profile of executive dysfunction, children with ADHD do not demonstrate the atypically high levels of SAB which are a characteristic of many children with WS. This challenges the frontal lobe hypothesis as a unitary account for SAB in WS.

The parents’ questionnaire rating in chapter 3 indicated that SAB among the children with WS was more pertinent to interactions with adults than interactions with children. This suggests that children with WS do show a level of discrimination in their SAB. These findings challenge the assumptions of the frontal lobe hypothesis and suggest that this theoretical account may not fully account for the high levels of SAB demonstrated in WS. The present author suggests that several factors may be involved in the high levels of SAB in WS. This chapter will examine the relationships between response inhibition, social-cognition and SAB in children with WS.

Previous research has indicated that individuals with WS show a positivity bias in their visual perception. Dodd and Porter (2010) recruited a sample of children and adults with WS and TD controls who were matched on mental age and chronological age. The participants completed a dot probe task which measured attention to face stimuli. In each trial of this task, a dot probe appeared on one side of the computer screen. This screen had previously presented participants with two images on opposing sides, a positive and negative face. The individuals were instructed to respond as fast as possible to indicate the location of the probe. Dodd and Porter reported that the individuals with WS responded to the location of the dot significantly faster when it was presented in the location of the happy in contrast to the angry face. This positivity bias was not present in the TD controls. This study has provided empirical support for an attentional bias towards positive stimuli in individuals with WS.

Neural findings have also provided support for a positivity bias in visual perception in WS (Meyer-Lindenberg et al., 2005). Hass et al. (2009) used functional MRI scans with individuals with WS to examine the activation of the amygdala when the participants' viewed positive, negative and neutral faces. Hass et al. reported that individuals with WS showed greater activation of the amygdala when viewing positive faces than when viewing neutral faces. This positivity bias was not present in the TD controls. In contrast, the TD controls showed significantly greater activation of the amygdala when presented with angry faces than individuals with

WS. These findings support a positivity bias within visual perception among individuals with WS.

If individuals with WS have a positivity bias in perception, it could be suggested that these individuals may also have a positivity bias in their social cognition. The social information processing theory (Crick & Dodge, 1994) has provided an account of the processes through which children understand their social environment. This theory proposes that when a child enters a social interaction, he or she proceeds through a series of mental processes which determine his or her behavioural response. Upon encoding sensory information from the social environment, the child is argued to interpret this information with regard to the cause behind the event. This process is termed attribution formation. Attributions are believed to be vital in successful social functioning as they allow an individual to choose the appropriate behavioural response within a situation through a consideration of the other person’s behaviour intentions (Leffert, Siperstein, & Widaman, 2010).

While previous researchers have identified biases in visual perception in WS, only one study to the author’s awareness has examined for an attribution bias in WS. Godbee and Porter (2013) administered a negative intention attribution task to a sample of children and adults with WS and TD controls who were matched for mental age and chronological age. The individuals were presented with cartoon line drawings which depicted a social interaction between two or more characters. The intentions of the characters in the scenarios were ambiguous. The children were instructed to describe what was happening in the scenario and based on these descriptions, the children’s reference to the negative or neutral intentions of the actor were coded. Godbee and Porter reported that the individuals with WS generated significantly fewer attributions of negative intent than TD individuals matched for chronological age. Godbee and Porter reported that there was no significant difference in the valence of the attributions formed by the individuals with WS and TD individuals matched on mental age. A limitation of this study was that the researchers did not measure positive attributions and therefore were unable to draw conclusions regarding a positive attribution bias in WS. Furthermore, Godbee and Porter compared the children with WS to a single intellectual ability control group. Given the dissociation in performance between verbal and visuo-spatial domains in WS, it is important to match the children with WS with TD control groups separately on verbal and non-verbal ability.

Outcome expectancy biases occur when an individual has a biased expectation of positive or negative outcomes across events (O'Sullivan, Szczepanowski, El-Deredy, Mason, & Bentall, 2011; Pornari & Wood, 2010). Depressed individuals, for example, have been reported to expect more negative outcomes and less positive outcomes across scenarios. This has been reported to contribute to the stability of a depressive mood state in these individuals (MacLeod & Salaminiou, 2001). While research has identified an attribution bias in WS, no research thus far to the author's knowledge has investigated for an outcome expectancy bias in WS. If children with WS demonstrate positivity biases when forming causal attributions, these individuals could be suggested to show a bias where they expect more positive outcomes across events.

Furthermore, no previous research to the author's awareness has examined for differences in patterns of information processing across scenarios where the outcomes are social in nature, versus non-social in nature. If the children with WS do show a bias within information processing, the use of social and non-social response options will provide insight into the specificity of this information processing bias to the child's social interactions. This can provide insight into whether children with WS are predisposed to perceive other people in a positive or hostile manner, or whether the children exhibit a generalised predisposition to view the world in a positive or hostile manner, such that they expect positive or negative outcomes across many different situations.

Within their social information processing theory, Crick and Dodge (1994) propose that there is a strong correlation between an individual's attributional style and their subsequent behaviour. This relationship has been demonstrated in typical and atypically developing populations (Dodge & Price, 1994; Nelson & Crick, 1999; Frey, Nolen, Edstrom, & Hirschstein, 2005). Accumulated research has provided support for an association between a hostile attribution bias and overt aggressive displays (King, Waschbusch, Pelham, Frankland, Andrade, Jacques, & Corkum, 2009; Mikami, Hinshaw, Lee & Mullin, 2008, Milich & Dodge, 1984). Previous research also identified a significant relationship between low levels of negative intention attributions and prosocial displays in a sample of TD primary school aged children (Nelson & Crick, 1999). These studies provide support for an association between social-cognition and behaviour. Previous research measured the formation of hostile or benign intention attributions; however, did not examine positive attributions. There is a need for further research to directly examine the relationship between a positivity bias in information processing and an engagement in SAB in TD children and children with WS.

4.2. Research aims of the current study

The overall aim of the present study is to examine the factors which underlie the high levels of SAB in WS. With careful consideration to theoretical accounts and inconsistencies within the research findings, the present study will investigate the role of response inhibition and information processing biases in the SAB of children with WS. Previous experimental and neural findings have provided support for a positivity bias within visual perception in WS (Dodd & Porter, 2010; Hass et al., 2009). In light of this, it could be suggested that this positivity bias may extend to social cognition. The present study aims to build on previous research by directly assessing for a positivity bias in WS in the formation of causal attributions and predictions. Social and non-social response options will be utilised in order to examine the extent to which any information processing biases are specific to the processing of social stimuli.

In line with the assumptions of the social information processing theory, both attributions and EF’s are suggested to have an important influence on the appropriateness of a child’s social interactions (Sanders & Phye, 2004). This challenges previous theoretical approaches which propose that a single impairment can explain the high levels of SAB in WS (Little et al. 2013) and reinforces the importance of investigating these two constructs and their associations with SAB in WS. In order to achieve this research aim, the present study sets out to test four hypotheses.

4.3. Research hypotheses

1. In making causal attributions and in outcome predictions, children with WS are hypothesised to show greater positivity biases than TD children matched on verbal and non-verbal ability.

2. The children with WS will show greater positivity biases when making social predictions than when making non-social predictions.

3. There will be an inverse relationship between response inhibition and SAB (children with poorer inhibition should score higher on SAB).

4. Positivity biases in the attribution and prediction tasks are hypothesised to be associated with higher SAB scores.

4.4. Method

4.4.1. Participants

21 children with WS participated in this study (7.50-16.49 years, M: 11.83 years, SD: 2.69, 12F, 9M). The data from one child were removed prior to analysis due to the child's difficulties following the verbal task instructions. The mother (N=19) or father (N=2) of each child with WS was also recruited. Further information on the participant sample has been reported in chapter 2 of this thesis.

71 typically developing (TD) children were recruited (5.02-7.96 years; M: 6.54, SD: .90, 31F, 40M). A large sample of TD children was recruited to allow the children with WS to be individually matched with a TD child on verbal and non-verbal performance. All TD children were recruited via three Scottish primary schools. The presence of mental and behavioural disorders within this sample were screened for using the SDQ (Goodman, 1997). Seven children were removed from the dataset prior to analysis as they showed abnormal ratings in the SDQ or the researcher was unable to obtain SDQ data from either the child's parent or teacher.

The children with WS were individually matched with 20 TD children on verbal ability (5.08-7.92 years, M: 6.47, SD: .94, 9F, 11M) on the BPVS II (Dunn et al., 1997). An independent samples t-test indicated that there was no significant difference between the children with WS (M: 67.24, SD: 19.15) and TD controls (M: 65.50, SD: 15.04) on the BPVS after matching, *t* (38) = .52, *p* = .608 (two tailed). The children with WS were also individually matched with 20 TD children on non-verbal ability (5.08-7.96 years, M: 6.15, SD: 91, 11F, 9M) on RCPM (Ravens et al., 1998). An independent samples t-test indicated that there was no significant difference between the children with WS (M: 16.71, SD: 3.76) and TD controls (M: 16.90, SD: 3.81) on RCMP after matching, *t* (38) = - .08, *p* = .934 (two tailed).

4.4.2. Measures

4.4.2.1. Sun-Moon Stroop Task (Archibald & Kerns, 1999)

The Sun-Moon Stroop task is a standardised measure of response inhibition. This task has been previously administered with TD children and children with WS (Little et al., 2013; Pasalich et al., 2010). In the control condition, the children are presented with a sheet of paper which contains 30 images of the sun and the moon arranged in a randomised order. The children are instructed to name each image, e.g. respond “sun” when they see a picture of a sun and “moon” when they see a picture of a moon. In the experimental condition, the children are required to respond “sun” when they see a picture of a moon and “moon” when they see a picture of a sun. An interference score is generated for each participant as the difference in accuracy rate between the control condition and the experimental condition. A lower score in this task indicates stronger response inhibition performance. Further details are provided in chapter 2.

4.4.2.2. Attribution/prediction measures

Three attribution/prediction measures were developed for use in the present study. Each measure consists of seven social vignettes. These vignettes describe different social scenarios, which include going to school, going to the park and attending a birthday party. The vignettes were developed to elicit the children’s causal attributions and/or outcome predictions across different scenarios. Each vignette is read aloud to the child and the child is required to choose from three response options: a positive attribution/prediction, a negative attribution/prediction and a neutral attribution/prediction. The children’s responses to each vignette were scored ‘+1’ for a positive attribution/prediction, ‘-1’ for a negative attribution/prediction and ‘0’ for a neutral attribution/prediction. There were 7 items in each scale; hence, scores could range from -7 to 7, where the more positive the score, the more positive the child’s causal attributions/predictions. Further details on the development of these measures have been reported in the general methodology chapter (chapter 2).

4.4.2.3. Adolph’s Approachability Task (Adolphs et al., 1998)

Adolph’s Approachability Task is a standardised task which measures an individual’s SAB to unfamiliar people (Bellugi et al., 1999; Frigerio et al., 2006; Little et al., 2013; Martens et al., 2009; Porter et al., 2007). Please see chapter 2 for full details on administering this task.

4.4.2.4. Social approach questionnaire (parent report)

This 11 item parent report questionnaire measures three aspects of SAB: the manifestation of SAB, perceived vulnerability of the child and the impact of SAB on daily/family life. Full details on the development and scoring of this questionnaire are detailed in chapter 2.

4.4.3. Procedure

The children with WS and their parent were visited at their home. The TD children were visited at school. The children with WS and TD children completed the Sun-Moon Stroop task, attribution and prediction measures and Adolph’s Approachability Task as described above. The parents of the children with WS completed the SAB questionnaire. Full ethical approval was granted from the University of Strathclyde ethics committee.

4.5. Results

4.5.1. In making causal attributions and in outcome predictions, children with WS are hypothesised to show greater positivity biases than TD children matched on verbal and non-verbal ability (Hypothesis 1)

The mean, SD and range of scores on the attribution and prediction measures are reported in table 5. Descriptive statistics are reported for each group (children with WS, TD verbal matched controls and TD non-verbal matched controls).

Table 5

Mean, SD and range of scores on the social attribution, social prediction and non-social prediction tasks (children with WS & TD comparison groups)

	Children with WS (N=20)		TD verbal matched controls (N=20)		TD non-verbal matched controls (N=20)	
	Mean (SD)	Range	Mean (SD)	Range	Mean (SD)	Range
Social attribution ratings	.45 (3.14)	-4 – 7	-.30 (2.66)	-5 – 4	-.55 (1.82)	-5 – 2
Social prediction ratings	3.25 (2.77)	-2 – 7	2.0 (2.68)	-5 – 7	1.75 (2.91)	-5 – 7
Non-social prediction ratings	3.35 (2.58)	-1 – 7	2.35 (2.56)	-5 – 7	2.65 (2.39)	-1 – 6

Table 5 indicates that the children with WS showed comparable performance to the TD comparison groups on the attribution scale. However, the children with WS formed more positive predictions than the TD control groups. There was within-syndrome variance in the valence of the attributions and predictions formed within all participant group, where some children demonstrated a strong positivity bias when forming attributions and predictions as illustrated by their extreme scores in this task.

A between groups analysis of variance (ANOVA) was conducted to examine for a statistically significant effect of participant group on the positivity of the attributions formed. The data met the parametric assumption of an interval scale of measurement. An analysis of skewness indicated that the data were normally distributed. An observation of Q-Q plots also indicated that the data points were lying along a straight line and there were no significant outliers. There was no significant effect of participant group on the valence of the causal attributions, *F* (2, 57) = .80, *p* = .453 (two tailed).

A mixed ANOVA was subsequently conducted to examine the effect of participant group and task type on the valence of the predictions formed. The between groups variable was participant group (children with WS, TD verbal matched controls and TD non-verbal matched controls). The within group variable was task type (social versus non-social). There was no significant effect of participant group on the valence of the predictions formed, *F* (2, 57) = 1.48, *p* = .235 (two tailed). There was no significant effect of task type on the valence of the predictions formed, *F* (1, 57) = 2.05, *p* = .158 (two tailed). Furthermore, there was no significant interaction between participant group and task type on the valence of the predictions formed, *F* (2, 57) = .57, *p* = .572 (two tailed).

4.5.2. The children with WS will show greater positivity biases when making social predictions than when making non-social predictions (Hypothesis 2)

As indicated in table 4, the children with WS showed similar mean scores in the social prediction scale and the non-social prediction scale. Paired samples t-tests were conducted to examine for a statistically significant difference in the valence of the predictions formed by the children with WS in the social and non-social prediction scales. The data met the parametric assumptions of an interval scale of measurement, normally distributed data points and no significant outliers. There was no significant difference in the valence of the predictions formed by the children with WS in the social prediction and non-social prediction tasks, *t* (19) = -.18, *p* = .858 (two tailed).

4.5.3. It is hypothesised that there will be an inverse relationship between response inhibition and SAB (children with poorer inhibition should score higher on SAB) (Hypothesis 3)

The mean, SD and range of scores in the Sun-Moon Stroop task, Adolph's Approachability Task and the parent SAB questionnaire are reported in table 6. For the parent SAB questionnaire, data were only obtained for the children with WS.

Table 6:

Mean, SD and range of scores on the Sun Moon Stroop task (response inhibition), Adolph's Approachability Task and parent social approach questionnaire (children with WS and TD comparison groups)

	WS (N=20)		TD verbal matches (N=20)		TD non-verbal matches (N=20)	
	Mean (SD)	Range	Mean (SD)	Range	Mean (SD)	Range
Sun Moon Stroop task	-.08 (.09)	-.22 - .09	-.13 (.12)	-.46 – 0	-.19 (.12)	-.38 - .01
Adolph's Approachability Task	69.55 (16.26)	41 – 100	51.10 (22.53)	20 – 99	50.80 (24.21)	20 – 100
Parent rated SAB	46.20 (5.75)	33 – 55				

Table 6 indicates that the children with WS performed marginally better than the TD comparison groups on the Sun Moon Stroop task, showing less interference. There was within-group variance across all groups in the children's performance in this task; however, this variance was larger in the TD comparison groups. The children with WS scored higher than both TD comparison groups on Adolph's Approachability Task.[2] There was substantial within syndrome variance in the children's scores in Adolph's Approachability Task across all groups; however this variance was largest for the TD comparison groups.

Bivariate Pearson's correlations were conducted to examine for a significant relationship between response inhibition performance and SAB among the children with WS (N=20) and TD children (N=64). The data met the parametric assumptions of an interval scale of measurement, no significant outliers, linearity of variables and normally distributed data points.

There was no significant relationship between response inhibition performance and performance in Adolph's Approachability Task in the children with WS (r = -.12, p = .610). There was no significant relationship between response inhibition performance and parents' ratings of SAB in the children with WS (r = .10, p = .671). There was no significant relationship between response inhibition performance and performance in Adolph's Approachability Task in the TD children (r = .04, p = .798).

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[2] The children with WS scored significantly higher in Adolph's Approachability Task than the TD children matched on verbal ability (p = .022) and the TD children matched on non-verbal ability (p = .019). Full analysis was reported in chapter 3.

4.5.4. Positivity biases in the attribution and prediction tasks are hypothesised to be associated with higher SAB scores (Hypothesis 4)

Pearson's Bivariate correlations were conducted to investigate the associations between the valence of causal attributions, social predictions, non-social predictions and levels of SAB. Correlations were examined for the WS sample (N=20) and a larger sample of TD children (N=64). The data met the parametric assumptions of an interval scale of measurement, no significant outliers, linearity of variables and normally distributed data points.

There were no significant relationships between the positivity of the children's social attributions (r = .04, p = .862), social predictions (r = -.10, p = .662), non-social predictions (r = -.08, p = .731) and performance in Adolph's Approachability Task in the children with WS. There were no significant relationships between the positivity of the children's social attributions (r = .10, p = .672), social predictions (r = .08, p = .740), non-social predictions (r = -.02, p = .936) and parent rated SAB in the children with WS.

There was a significant positive relationship between the positivity of the attributions formed by the TD children and performance in Adolph's Approachability Task (r = .23, p = .032, one tailed). There were no significant relationships between the positivity of the predictions formed in the social (r = .14, p = .139) and non-social measures (r = .13, p = .184) and performance in Adolph's Approachability Task in the TD children.

4.6. Discussion

4.6.1. Summary of findings

With consideration to the inconsistencies across research findings, the current study proposed that high levels of SAB in WS could not be accounted for by response inhibition performance alone. The present study examined the role of response inhibition and information processing biases in the high level of SAB demonstrated among children with WS. There was no significant difference in the valence of the attributions and predictions formed by the children with WS and TD children who were matched for verbal ability and non-verbal ability. Furthermore, there was no significant relationship between response inhibition performance, information processing and SAB in WS. These findings will be further examined within the present discussion.

4.6.2. In making causal attributions and in outcome predictions, children with WS are hypothesised to show greater positivity biases than TD children matched on verbal and non-verbal ability (Hypothesis 1)

Children with WS have been reported to demonstrate preferential processing of positive stimuli over negative and neutral stimuli (Dodd & Porter, 2010; Hass et al., 2009). While previous researchers have provided insight into a positivity bias in visual perception, no research thus far has directly examined for a positive attribution bias in WS. The current study reported there to be no significant difference in the valence of the attributions and predictions generated by the children with WS and TD children matched on intellectual ability. These findings differ from previous research which identified lower levels of negative attributions in individuals with WS when compared to TD controls (Godbee & Porter, 2013). This disparity in findings could be accounted for by differences in the methodologies used by these researchers. Godbee and Porter (2013) reported that individuals with WS formed significantly fewer attributions of a negative intent than TD individuals who were matched on chronological age but not TD individuals who were matched on mental age. In consequence, it could be suggested that this attribution bias may be a result of the child's developmental delay, as this group difference was not found when intellectual ability was controlled for. In light of these findings, caution should be considered when interpreting these between group differences. In order to control for the effect of developmental delay on the children's performance across the attribution and prediction tasks, the children with WS in the current study were matched with TD children on verbal and non-verbal ability with consideration to the dissociation in performance between verbal and visual-spatial domains in WS (Jarrold et al., 2007; Vicari et al., 2003). This builds on previous research which used a single intellectual ability control group (Godbee & Porter, 2013).

The difference in findings between the present study and previous research could also be accounted for through an evaluation of the strengths and limitations of the tasks used by these researchers. Godbee and Porter (2013) presented the individuals with WS with pictures of cartoon line drawings which depicted a social interaction between two or more characters. The individuals with WS were required to explain the cause of the event and their formation of negative and neutral intention attributions were coded. This task could be suggested to have poor ecological validity due to the artificial nature of the stimuli. In light of this critique, caution should be exercised when interpreting these research findings. Within the present study, the children were presented with coloured photographs which depicted a real-life social scenario. Furthermore, the vignettes described various social scenarios which were considered to be familiar to the age group of the children who were participating in this study (5-16 years). This included going to school and going to the park. These modifications to the task design could be considered to have a greater level of ecological validity than previous tasks.

The descriptive statistics indicate that the children with WS generated more positive outcomes within the social prediction task than both TD comparison groups; however, this group difference did not reach statistical significance. This non-significant difference could be accounted for by a lack of statistical power as a result of the relatively small sample in the current study. Furthermore, there were high levels of within-syndrome variance among the children regarding the valence of the attributions and predictions formed. A few children demonstrated a strong positivity bias when forming attributions and predictions as highlighted by extreme scores on the positive end of the valence spectrum. This suggests that while the children with WS did not make attributions or predictions which were significantly different from the TD children, this positivity bias was present for a sub-sample of the group. These findings provide further support for the heterogeneous nature of WS. However, these distributions were not subjected to statistical analysis and therefore caution is required when interpreting these findings.

4.6.3. The children with WS will show greater positivity biases when making social predictions than when making non-social predictions (Hypothesis 2)

The present study is the first to the author's knowledge to examine for differences in the manner with which children with WS process information when the outcomes are social in nature versus non-social in nature. If an information processing bias exists, it is important to identify whether this bias is specific to social stimuli, for example, are children with WS predisposed to expect positive outcomes from their social interactions, or if children with WS show a general positivity bias when interpreting social and non-social stimuli. The children with WS in the current study generated predictions of a similar valence when the response options were social and non-social in nature. The current results differ from previous research findings which found that individuals with WS exercised a positivity bias when interpreting social stimuli (Dodd & Porter, 2010). In this present study, the findings suggest that within the context of social information processing, children with WS may process social and non-social stimuli in a similar way. However, this is the first study to test this hypothesis. The small sample size in the current study limits the ability to accurately test this hypothesis due to a lack of statistical power. Researchers should seek to further investigate the nature of information processing in children with WS by recruiting a larger sample size, thus increasing the power of their study.

4.6.4. It is hypothesised that there will be an inverse relationship between response inhibition and SAB (children with poorer inhibition should score higher on SAB) (Hypothesis 3)

The present study found that there was no significant relationship between response inhibition performance and SAB. The current study used a similar methodology to previous researchers, with regards to the characteristics of the participant sample and the measures used (Little et al., 2013). This allowed more reliable comparisons to be drawn between the present results and these previous research findings. The current findings differed from Little et al. (2013) who reported that there was a significant negative relationship between response inhibition performance and Adolph’s Approachability Task performance in the children with WS. However, Little et al. reported that this relationship was only significant for a sub-sample of the children with WS who demonstrated the poorest inhibition performance and highest levels of SAB. These findings were analysed using cluster analysis. While an advantage of cluster analysis is that it can be applied with small sample sizes, cluster analysis has also received some criticism regarding the exploratory nature of this test and the potential effects of researcher bias on the clusters identified (Kent, 2015). Caution should therefore be taken when interpreting these research findings. High levels of within-syndrome variance were reported in the present sample with regards to the children’s response inhibition performance and their SAB ratings. This could have masked any potential relationship between inhibition performance and SAB and may account for this disparity in findings.

Previous researchers used Adolph’s Approachability Task to measure SAB in children with WS (Little et al., 2013). Adolph’s Approachability Task has received criticism regarding its poor ecological validity (Fisher, 2014). In light of this critique, the present study examined the relationship between response inhibition, performance in Adolph's Approachability Task and parents' reports of SAB in order to provide a stronger examination of this relationship. There was no significant relationship between response inhibition performance and SAB as measured using the standardised Adolph's Approachability Task and parents' ratings of the children's SAB. The current results challenge the theoretical assumptions of the frontal lobe hypothesis which propose that high levels of SAB in WS are the result of the individual's impairments in executive functioning. This raises further questions regarding the principle role of EF impairments in an engagement in high levels of SAB in WS and reinforces the need to investigate which other factors underlie an engagement in high levels of SAB.

In the present study, the children with WS scored higher than the TD controls on the Sun Moon Stroop task. These findings challenge previous research findings of substantial response inhibition deficits in children with WS (Carney et al., 2013; Porter et al., 2007). This disparity in findings could be accounted for by the procedure used to measure inhibition performance. Menghini et al. (2010) administered a verbal Stroop task to measure response inhibition performance in a sample of individuals with WS. Menghini et al. reported that although the individuals with WS showed greater errors in the verbal inhibition task than TD controls, there was no significant difference in the participants' interference score. An interference score is generated as the difference in the time taken to inhibit a response between the control condition, where participants are simply instructed to say a word, and the experimental condition where participants are instructed to inhibit their response of naming a word and to generate an alternative response. The Sun Moon Stroop task which was administered in the current study assessed the children’s response times only. This could therefore account for the non-significant difference between the children with WS and TD controls in the present study.

The Sun Moon Stroop Task was used in line with the methodology of Little et al. (2013); however, this task only taps into verbal inhibition. Carney et al. (2013) administered a verbal and a visuo-spatial inhibition task with a sample of children with WS. In the verbal inhibition task, the children with WS were required to inhibit a verbal response, while in the visuo-spatial inhibition task, the children with WS were required to inhibit a motor response. Carney et al. reported that the children with WS performed significantly poorer than TD children in both the verbal and visuo-spatial inhibition task. However, the children’s reaction times were significantly poorer in the motor inhibition task. Previous research has indicated that individuals with WS demonstrate difficulties with personal space regulation (Lough et al., 2015) and often engage in disinhibited tactile behaviours towards strangers (Bellugi et al., 1999) and peers (Davies et al., 1998; Elison et al., 2010). With a consideration to these reports of difficulties in the self-regulation of behaviour, it would be informative to examine the relationship between motor inhibition performance and levels of SAB in children with WS.

4.6.5. Positivity biases in the attribution and prediction tasks are hypothesised to be associated with higher SAB scores (Hypothesis 4)

There was a significant positive relationship between the positivity of the causal attributions formed by the TD children and performance in Adolph's Approachability Task. These findings are in line with previous research which indicated a positive attributional style to predict prosocial behaviour in TD children (Nelson & Crick, 1999). There was no significant association between the positivity of the causal attributions or predictions formed by the children with WS and the children's SAB. However, as has been raised throughout this chapter, there were high levels of within-syndrome variance among both the children with WS and the TD children in their scores on the attribution and prediction tasks.

The present findings suggest that a positive attributional style may be associated with SAB at a moderate level as was demonstrated in the TD children, where children who attribute positive intentions to other people are more likely to be motivated to engage in social interactions. However, there was no significant relationship between the valence of the attributions and predictions formed and SAB in the children with WS. These findings provide partial support for the theoretical assumptions of the social information processing theory, suggesting that attributions have an important influence on children's social interactions during typical development; however, this relationship was not present for the neurodevelopmental disorder of WS.

The non-significant relationship between information processing and SAB in WS is in contrast to previous findings of a significant association between information processing biases and social behaviour in children with ADHD (King et al., 2009; Mikami et al., 2008). This suggests that the relationship between information processing and social behaviour differs between neurodevelopmental disorder groups. While previous researchers identified high levels of hostile attributions in children with ADHD (King et al., 2009; Sibley et al., 2010, Zentall et al., 2001), the children with WS in the current study did not show a positive or negative bias within information processing.

4.6.6. Conclusion

The present study was conducted to investigate which psychological variables can account for the high levels of SAB demonstrated by children with WS. The present author suggested that response inhibition performance alone could not account for these high levels of SAB, as these inhibition impairments were reported across neurodevelopmental disorders. In consequence, this present study examined the relationships between response inhibition, information processing biases and levels of SAB in children with WS and TD children.

The current study found that there was no significant relationship between response inhibition performance and SAB in TD children or children with WS. This challenged the findings of Little et al. (2013) and the theoretical assumptions of the frontal lobe hypothesis. This study found there to be no significant difference in the positivity of the attributions and predictions formed by children with WS and TD children matched on intellectual ability. There was a positive association between the positivity of the causal attributions formed by TD children and SAB; however, there was no significant relationship between information processing and SAB in children with WS. These non-significant findings were interpreted with reference to the tasks utilised and high levels of variance in the children's performance across these tasks.

A challenge when working with rare disorder groups such as WS is the potential barriers to recruitment, with low prevalence rates posing constraints on the sample sizes obtained and level of statistical power. This low level of power can increase the risk of type two errors, where a researcher may falsely accept a null hypothesis due to a lack of statistical power to detect an effect. With consideration to the small sample size in the present study, it is suggested that caution is required when interpreting the non-significant findings across this chapter. Furthermore, where true effects are reported, low levels of statistical power can result in over-estimates of effect size, and consequently, lower rates of replication across research (Button, Loannidis, Mokrysz, Nosek, Flint, Robinson, & Munafo, 2013). It is therefore vitally important that researchers working with low prevalence populations consider the challenges of low sample sizes, lower levels of statistical power and the associated implications when interpreting research findings. Future research should be conducted to further investigate these relationships using larger sample sizes where possible.

Chapter 5: Social approach behaviour in children with Williams syndrome: A qualitative approach

5.1. Introduction

Individuals with WS often demonstrate a hyper-social phenotype and a strong motivation to engage in social interactions with both familiar and unfamiliar people (Järvinen-Pasley et al., 2010; Jones et al., 2000). Previous research has generated strong support for claims of higher levels of SAB among individuals with WS when compared to TD individuals and groups with other developmental disorders (Bellugi et al., 1999; Dodd et al., 2010; Doyle et al., 2004; Järvinen-Pasley et al., 2010; Martens et al., 2009). This was further illustrated across the experimental ratings and parental questionnaire ratings in chapter 3 of the present thesis. In light of previous findings of high levels of social vulnerability (Fisher et al., 2013) and poor stranger danger awareness in WS (Riby et al., 2014), it is vital to expand our understanding of stranger interactions in children with WS.

High levels of SAB in WS have been suggested to be a result of EF impairments (Little et al., 2013; Porter et al., 2007). Although this has not been shown in the present thesis (chapter 4), this could be accounted for by the nature of the task used which only assessed verbal inhibition. It is suggested that behavioural inhibition may be more affected in WS. Preliminary research indicates that individuals with WS show difficulties regulating their behaviour during social interactions. Lough et al. (2015) investigated personal space regulation in a sample of individuals with WS (4 – 36 years) by soliciting parents' ratings within the SRS. The individuals with WS were reported to be significantly poorer at regulating personal space boundaries during social interactions when compared to TD individuals matched on chronological age and a comparison group of individuals with ASD. Furthermore, adults with WS have been reported to engage in high levels of disinhibited affection and tactile behaviour within their daily social lives (Davies et al., 1998; Elison et al., 2010). This disinhibited SAB was reported to negatively impact on several aspects of life for these adults with WS, including the individuals' independence, peer relations and vulnerability (Davies et al., 1998; Elison et al., 2010).

Research thus far has predominantly used a quantitative approach to examine levels of SAB in WS, where researchers have administered experimental rating tasks and parent questionnaires (Bellugi et al., 1999; Järvinen-Pasley et al., 2010; Martens et al., 2009). These research studies identified high levels of SAB in many children with WS. Further research is sought to expand our knowledge of the implications of atypically high levels of SAB for the child with WS and his or her family.

Qualitative analysis can provide valuable insights into the personal experiences of individuals and an examination of the diversity within these experiences (Braun & Clark, 2006; Fischer, 2011). Qualitative analysis has been used to measure stranger danger understanding among children with WS (Riby et al., 2014). However, only one study to our awareness has used a qualitative methodology to examine SAB in WS. Lough et al. (2016) interviewed the parents of a sample of children with WS (6 – 15 years). The interviews solicited information on several aspects of the children's SAB, including the children's social motivation and their stranger danger awareness.

The interview data were analysed using the qualitative method of thematic analysis. When using thematic analysis, themes are identified from the data extracts which provide a direct measure of the individual's thoughts and motivations (Braun & Clarke, 2006). The interview data illustrated high levels of SAB among the children with WS. Many of the parents reported that their child with WS engaged in atypical approach behaviour towards strangers, showed a poor understanding of social boundaries and lacked an awareness of social danger; however, there was substantial within group variance in the children's social approach patterns. This variance was accounted for by differences in the personality traits of the children, where the children who were more introverted were reported to engage in less SAB. The levels of parental supervision also varied within this sample in accordance with the parents' levels of trust in their child. These findings support heterogeneity in the characteristics of SAB in children with WS, in line with previous research evidence (Little et al., 2013; Porter & Coltheart, 2005).

The research findings of Lough et al. (2016) have contributed to a very limited area of research by providing the first qualitative examination into the characteristics of SAB in children with WS. With a consideration to the lack of qualitative research studies in this field and the high levels of heterogeneity in WS (Porter & Coltheart, 2005), there is a need to further develop the findings of Lough et al. (2016) in order to obtain further insight into the characteristics of SAB in WS and the implications of this SAB for the child with WS and family life.

Like all children, those with WS live and develop in a social context (Barbarin & Wasik, 2009). It is important to understand how the child's exceptional characteristics are perceived by key parties, such as their parents. This study aims to solicit parents' perceptions on their child's atypical SAB. This study further aims to investigate how the characteristics of the children's SAB impacts on family life and parenting strategies. This will be achieved through a qualitative analysis of interview data solicited from parents. This study aims to address the research question "What are parents' insights into the nature of their child's interactions with strangers and the wider impact of this behaviour?"

5.2. Method

5.2.1. Participants

The parents of 21 children with WS were recruited from across the UK. The mother (N=11), father (N=2) or both the mother and father (N=8) participated in this research. The children with WS were aged between 7.5 and 16.49 years (M: 11.83, SD: 2.69; 12M, 9F). Further information regarding participant recruitment is provided in chapter 2.

5.2.2. Measures

5.2.2.1. Social approach interview (parents)

A semi-structured interview was designed to solicit qualitative information from parents regarding their child's SAB. This interview contained seven broad questions and additional probe questions. The items measured three factors: the manifestation of SAB, stranger danger awareness and the wider impact of SAB on the child and family life. This interview schedule was developed through reference to previous literature on SAB in WS (as detailed in chapter 2).

5.2.3. Procedure

The parents completed a semi-structured interview as an assessment of their child's SAB. The interviews were audio recorded with the permission of the participants.

5.2.4. Data analysis

The interview data were analysed using the qualitative methodology of thematic analysis (Braun & Clarke, 2006). Thematic analysis involves the identification of patterns or themes in a dataset and allows a rich interpretation of these themes. An inductive approach was used in the present study, where the final themes were generated directly from the interview data, rather than being driven by the analytical or theoretical interests of the researcher. In line with the principles of a realist account, the data were interpreted as a direct communication of the participants' beliefs and motivations. This is in contrast to a constructionist approach, where the individuals' experiences are interpreted as a product of society (Guest, MacQueen, & Namey, 2012).

In line with the guidance of Braun and Clark (2006), several steps were taken when analysing the data. The audio data was initially transcribed verbatim. Codes were formed across the dataset which were subsequently condensed into five main themes through meticulous review.

5.3. Results

Five themes were identified from the data, as illustrated in figure 1. All themes and sub-themes will be discussed and extracts from the data which illustrate these themes will be provided. All names within the extracts will be replaced with pseudonyms in order to protect the identity of the participant. Across the results section the gender and age of the child being discussed will be presented in parentheses.

1. Indiscriminate nature of social interactions

One theme identified was the indiscriminate nature of the children's social interactions. Several parents reported that their child showed an interest in approaching people, regardless of the level of familiarity of the person:

"Yes she will go up to anybody. I mean I usually have her grasped tightly in one hand but if she sees somebody she will want to talk to them whether she knows them or not" (Female, age 8)

"She will talk to people all the time when you're out and sometimes I'll say to her 'Who was that? Did you know them?' No, but yet she is chatting away as if she does" (Female, age 11)

"He will talk to anybody…it's not really exclusive" (Male, age 9)

Parents' reports, as expected, highlight an unusually high level of interest in social interactions among children with WS and the indiscriminate nature of these interactions. Unsurprisingly, but importantly, it becomes clear that this has repercussions on parent-child interaction. Parents are concerned about these aspects of the child's social behaviour and attempt to regulate them, physically or through verbal interrogation and warnings.

1.1. Inquisitive personality

Several parents accounted for this indiscriminate approach behaviour as due to their child's inquisitive personality:

"She has great curiosity about new people and that drives her" (Female, age 11).

"She has always been very confident and keen to meet new people" (Female, age 15).

"We are sat at a table and she will go off to any table, talking to anybody...she has got an enquiring mind to other people all of the time" (Female, age 8).

These extracts suggest that this sociability is driven by the children's interest in other people and their need for knowledge about these individuals. These quotes also highlight the parents' acceptance of this atypical communication style as being part of their child's character with reference to what could be perceived to be positive character traits such as "great curiosity" and "very confident".

2. Social Disinhibition

Another prominent theme within the data was the children's uninhibited behaviour when meeting new people. Several parents reported that their child showed disinhibition in their communication and physical interactions with strangers.

2.1. Atypical communication

Many of the children were reported to engage in what could be considered an inappropriate or unusual style of communication.

2.1.1. Intense questioning

Several parents reported that their child often engaged in intense questioning when they encountered someone for the first time:

"You could be stood in the queue at the supermarket and the next thing you know he has got someone's life story as he has asked them so many questions" (Male, age 11).

"She always likes to ask people their name, ask people where they stay, do you have any children... It's as if she has been programmed to ask that" (Female, aged 14).

One parent recalled a situation when her daughter approached a new parent at a swimming club and engaged in repeated questioning: *"She just walked up to him and started questioning him, "how did you get here?", "did you come on the motorway?", "what's your name?", "how old are you?", "where do you live?", "what's your telephone number?" and that's typical" (Female, age 8).*

These extracts suggest that these children with WS have difficulties restraining their curiosity within social interactions as highlighted by their engagement in uninhibited questioning. The style of questioning was reported to be one way and repetitive. This could be suggested to highlight the children's difficulties within communication.

2.1.2. Inappropriate nature of conversations

Several parents also reported that their child often asked inappropriate or personal questions within social interactions with strangers:

"She doesn't necessarily understand that what she is asking, some people might find offensive… It's anything that comes to her mind really; she will just ask what she thinks" (Female, age 8).

"Yeah she has a lot of inappropriate conversations with people…yeah she would ask quite personal things of people as well. She wouldn't sort of think I shouldn't ask that as I don't know them" (Female, age 11).

"He can say inappropriate things, I suppose a lot of the time it is innocent questions, he just doesn't understand that actually that can be quite offensive" (Male, age 13).

Parents' noted their child's difficulties differentiating and moderating their conversations based on the familiarity of the other person. This could be suggested to highlight the child's deficits in social cognition. Across the extracts, parents acknowledge that their child's questions could unintentionally cause upset or embarrassment for the recipient. This in turn could cause a knock on effect of embarrassment or discomfort among the family members who find themselves amid these awkward situations.

A few parents noted that some individuals were unsure how to respond to the child's uninhibited behaviour, with strangers' reactions reported to be more favourable when the children were younger:

"When she was younger, people thought it was lovely because she was this small toddler and it was very much older people seemed to think that was lovely, but it worried me, but now that she's bigger you can see people standing back looking as if she say why is that girl asking me that" (Female, aged 14).

"Obviously when they are little tots, it's sort of quite endearing and people will tend to talk back but some people can sort of look at her now as if to say 'what are you saying? Why are you talking to me? I don't even know you'. You know I think because of her age now" (Female, age 11).

These extracts highlight that the children's overly friendly interactions with strangers become more problematic with age as some adults were reported to be less receptive to these behaviours by older children and adolescents. These quotes indicate the parents' awareness and concerns that the inappropriateness of their child's social interactions will become more evident and problematic with the child's increasing age as other people become less tolerant.

2.2. Tactile behaviour

Several parents reported that their child engaged in high levels of tactile behaviour:

“He wants to kiss people and hug them, all the time” (Male, age 9).

“If you were to smile at her she would just run up and hug you” (Female, aged 14).

“She gives the doctor a cuddle; she gives the teachers a cuddle. Everybody gets a cuddle” (Female, age 15).

Several parents also noted the indiscriminate nature of their child’s tactile behaviour, where these children were reported to seek out physical contact with other people regardless of their level of familiarity:

“She will give anybody a hug…she is very affectionate, which isn’t so much of a problem when it’s somebody you know of or are familiar with but when it’s a complete stranger” (Female, age 8).

One parent also noted her son’s poor judgement of other people’s personal space and her need to frequently remind him about this behaviour:

“He is quite tactile and he also doesn’t really know where other people’s personal space is so he gets quite close to people. This is anyone” (Male, age 8).

This indiscriminate tactile behaviour was raised as a major concern among the parents. This engagement in uninhibited tactile behaviour and affection was reported across children and adolescents. This would not normally be considered a socially acceptable pattern of behaviour, particularly during adolescence. This further supports the problematic nature of this behaviour given the older age range of these children.

However, there were individual differences among the parent reports, where a few parents noted that their child would not initiate tactile behaviour towards a stranger:

"I don't think she would go up and randomly hug a stranger. She's not one for going up and hugging them but if she started chatting to a stranger and a stranger went to hug her, she would hug them back" (Female, age 8).

"No, I don't think he's ever really ran up and hugged complete strangers...He will go up and hug somebody but generally only if he knows them or he has spoken to them quite a few times and he knows more about them" (Male, age 9).

These extracts indicate within-syndrome variance in the levels of tactile behaviour engaged in by children with WS. This provides support for a heterogeneous profile of social behaviour in WS.

3. Difficulty translating knowledge into practice

Another theme identified in the data was the children's difficulties translating their knowledge of stranger danger into practice. Several parents reported that while their child demonstrated a theoretical awareness of stranger danger, their child struggled to apply this knowledge in a real-life social interaction:

"We have very much got issues with him being able to in a classroom scenario give all the right answers, but then when he is put into a real life situation he doesn't make the right call" (Male, age 13).

"If you asked my child not to approach a stranger she will 100% say that she won't but two seconds later she has. She knows it's wrong but can't seem to help herself. She can talk the talk as you would say i.e. answers you the way you would expect her to, but always does the opposite" (Female, age 14).

"We try to say the dangers of going with strangers so obviously we are trying to embed that into her so she knows but if the situation comes around and if they said the right thing, she would go. She hasn't got stranger danger, she would give you probably the right answers...but I'm quite certain she would go" (Female, age 15).

One parent also noted her daughter's denial when she has spoken to a stranger and her awareness that this behaviour was inappropriate:

"Whenever she is going out, mum and dad say to her "don't stare, don't speak to anyone you don't know", and then her brother will come in and say "mum, Jessica was talking to that man up there"... when she comes back in she will say "no, no" when you ask if she spoke to anyone... She will be all apologetic and gets upset when she's done it because she's upset you, but at the same time this thing takes over*" (Female, aged 14).*

"Like if there was someone sitting beside her, she would be cuddling into them but she looks over at you while she's doing it as she knows you won't be happy with her" (Female, aged 14).

However, there was diversity within the interview responses as a few children were reported to be able to apply their understanding of stranger danger with practice:

"He does have some awareness, and I don't think he would ever go off with anybody" (Male, age 9).

"She knows that you always have to ask before you stroke a dog, so she wouldn't just go off and speak to someone straight away" (Female, age 16)

Parents’ concerns regarding their child’s vulnerability around strangers was evident across these extracts. There was a sense of despair among the parents that despite their efforts to educate their child about the dangers of approaching strangers, the parents acknowledged that the probability would be that their child would not be able to execute these principles in a real-life situation. This is suggested to have ramifications for the parent-child relationship, where several parents expressed their frustrations that despite their extensive efforts to instil an understanding of stranger danger in their child, the children were often unable to implement these behavioural requests.

Several of the parents reported that their children with WS showed an awareness that their behaviours were inappropriate and were a violation of their parents’ wishes. In some cases, this led to further behavioural and socio-cognitive adjustments in the child. For example, a few of the children were reported to attempt to deceive their parents regarding their SAB lapses. Several of the children often demonstrated guilt about their SAB and frequently became upset when confronted by their parents.

3.1. Inhibition deficit

Some parents suggested that their child’s difficulties applying their knowledge in practice could be a result of inhibition impairments:

“You can see her sometimes trying to sit on her hands like as if she knows but it’s just overwhelming, it just comes right over her and she can’t stop” (Female, aged 14).

"She will be all apologetic and gets upset when she's done it because she's upset you, but at the same time this thing takes over" (Female, aged 14).

"As she is chatting to them she will actually say '"I shouldn't be speaking to you because you're a stranger"…By the end of this conversation, we were getting off the train and she said do you want to come to our house?" (Female, age 13).

These extracts highlight the inner struggle many children with WS experience as they attempt to regulate their strong impulses to engage in social interactions and their wishes to abide by their parents teachings. The children showed efforts to self-regulate their behaviour by their own attempts of physical restraint, where one child was reported to sit on her hands. The children also demonstrated emotional self-regulation where they were reported to become upset and rueful of their actions. This also indicates a level of social-cognitive understanding, where the children showed an awareness when they had upset or disappointed their parents.

3.2. Interpretation of stranger danger

However, other parents believed that their child's understanding of stranger danger was very limited:

"It's like he does understand when we are saying it to him in part but he doesn't really understand what a stranger is as he thinks if he knows your name, you're not a stranger, you're a friend" (Male, age 11).

"She understands but I think if you were in that situation her understanding of it would go, as if that person were being nice to her, instantly she would think oh they are a nice person then" (Female, age 11).

"I think he knows that we don't want him to do it but I don't think he knows why it's dangerous" (Male, age 12).

These extracts highlight the parents' perceptions of vulnerability regarding their children's naivety and the potential consequence of this for the child's safety. While concerns regarding stranger danger could be considered to be a typical worry among all parents, these quotes illustrate that these anxieties are heightened for the parents of children with WS due to their child's very trusting nature, with this a core feature of the WS social phenotype.

4. Changing nature of SAB with age

Another theme identified was a change in the nature of the child's approach behaviour with age. Several parents reported that the nature and frequency of their child's SAB had declined and become more manageable with age:

"When he was younger, he was a lot worse than he is now…he would have gone off with any Tom, Dick or Harry" (Male, age 12).

"(When she was younger) she would be inappropriately friendly towards strangers. She is now more restrained. If you take her to a cafe or a restaurant she wouldn't be looking to talk to people around her… she wouldn't just start talking to another customer in the shop but somebody on the till she would be keen to talk to or the waitress she would be keen to talk to" (Female, age 15).

"She would have to have built up a bit of a relationship beforehand before she would just randomly hug. Whereas I think a few years ago, she would have just hugged everybody" (Female, age 15).

"Yeah it's something that she's doing quite well with; they discourage it at secondary school. They swap the hugs for high 5's and they've done quite a lot of work on personal space, so she is getting better at it… she would run up to the teacher when they came out in the morning and then just stop a couple of inches before them like "oh I've remembered I haven't to do that". So yeah she's learnt quite well since she's been at secondary school" (Female, age 11).

"When he was younger, he was a lot worse than he is now. We have taught him stranger danger so it's a learnt behaviour now" (Male, age 12).

These extracts highlight a decline in the inappropriate nature of interactions with strangers among some children with WS as a result of education and regular reminders on socially appropriate behaviour. These quotes indicate that the children continue to show a strong pull towards social interactions; however, with increasing age and maturity, the children showed greater efforts to self-regulate their behaviour in accordance with their teachings. These extracts also reinforce that stranger danger is an area of concern for many parents of children with WS and as such they recognise the value in devoting their time in repetitively training their child in stranger danger principles and closely monitoring their child's actions.

5. Social vulnerability

Another theme identified within the data was the children's social vulnerability. Many parents reported that their child did not have an ingrained sense of danger and many of the children were reported to require close supervision as a consequence of their social vulnerability.

5.1. Lacks an ingrained sense of danger

Many parents reported that their child did not have an ingrained sense of danger and had a very trusting nature:

"You have to be careful as she would walk away with anybody. There is no fear; she thinks everybody is an angel. Anybody that gives her a smile" (Female, age 11).

Within this extract, the parent indicates the triggering effect of another person's smile on the child. It is suggested, that the child with WS is highly sensitive to the emotional expressions of other people and is particularly drawn to positive facial expressions. This is in line with the theoretical assumptions of the amygdala hypothesis.

"If anybody approached him and chatted to him, he would quite happily talk to them. If they said oh come I'll go get you a bag of sweets, he would come easily" (Male, age 13).

"She is very trusting. If someone stopped her in the street and said "your mum said I've got to come and pick you up' and it was especially an adult, she would just go" (Female, age 15).

"She would just wander off to talk to them (strangers). She doesn't see any danger in doing that whatsoever and she thinks everybody is her friend. If I say to her, you didn't know that person, "I did!" because she had already asked them their name and so she thinks they are instantly a friend" (Female, age 11).

These extracts emphasise the highly trusting nature of children with WS and the vulnerability of these children as a result. There was evidence of high levels of parental anxiety regarding the children's vulnerability to strangers. Within these quotes the parents incorporate the 'stranger danger' narrative of fear to reinforce their concerns about their child.

The parents of one child described a situation when their son found himself in a vulnerable situation when he initiated an interaction with a group of teenagers:

"They had started trying to beat him up, they popped his tyre and everything… he said they were his friends…So he didn't understand that he had been in danger at all' (Male, age 11).

This highlights the vulnerability of children with WS and the ramifications of this social naivety for the child's wider social life, including the child's risk of peer victimisation. This also reinforces the parents' fears of the child's vulnerability when unsupervised.

Another parent reported that her daughter did not have any fears or anxieties around strangers which she believed contributed to her child's social vulnerability:

"She doesn't feel threatened by strangers. So that's kind of a difficult issue…she doesn't have the same feeling or fear that we would as an ordinary society about strangers" (Female, age 8).

Several of the parents expressed concerns regarding their child's vulnerability as a result of their very trusting nature:

"You just worry about how other people will react to her. We have never had any bad reactions but it's the vulnerability that the wrong person at the wrong time might take advantage, that's the big thing" (Female, aged 14).

These extracts emphasise the child's lack of fear around strangers and parents' concerns as a result of this. One parent distinguished between her child's relative absence of fear around strangers and the relative fear and caution demonstrated by society. This contrast reinforces the parent's perceptions of the atypical nature of their child's interactions with strangers.

5.2. Need for parental supervision

In consequence to this poor awareness of danger, many of the children were reported to require constant supervision:

"He can't be by himself at all; he's got no sense of danger or stranger danger" (Male, age 9).

"You've always got eyes on the back of your head" (Female, aged 14).

One parent reported that she could not allow her son out of her sight for even a few minutes due to the potential for him to be led by strangers:

"I wouldn't even let him walk to the car as I know that if someone came and said "oh I've got some cats in my car...or your mum told me to come and get you", he would just take that as face value, yeah you've told me that and I'll go" (Male, age 9).

Several parents also emphasised their child's vulnerability and need for supervision by making comparisons to their TD siblings:

"When we are out, I watch her more than I watch the boys despite the fact they are 3 years younger. They are just more aware, they will stay close to me and they won't wander, whereas she will" (Female, age 8).

"Yeah it is a constant, constant worry. Whereas our other two would have been playing out at this age, you couldn't let Max play out...he would go with anybody as he would trust anyone" (Male, age 11).

These extracts suggest that the children's indiscriminate friendliness may constrain their opportunities for independence, with many parents reporting a need to supervise their child due to their vulnerable nature. Across these quotes, the children's trusting nature and vulnerability are shown to have evident implications for the parents' anxieties, stress levels and day to day functioning. This need for constant supervision and the parents' anxieties regarding their child are suggested to impact on the family unit, with the need for planning and additional supervision potentially constraining family activities. The parents' also drew comparisons between the child with WS and their younger siblings regarding levels of support. This further reinforces the heightened vulnerability of children with WS when compared to TD children.

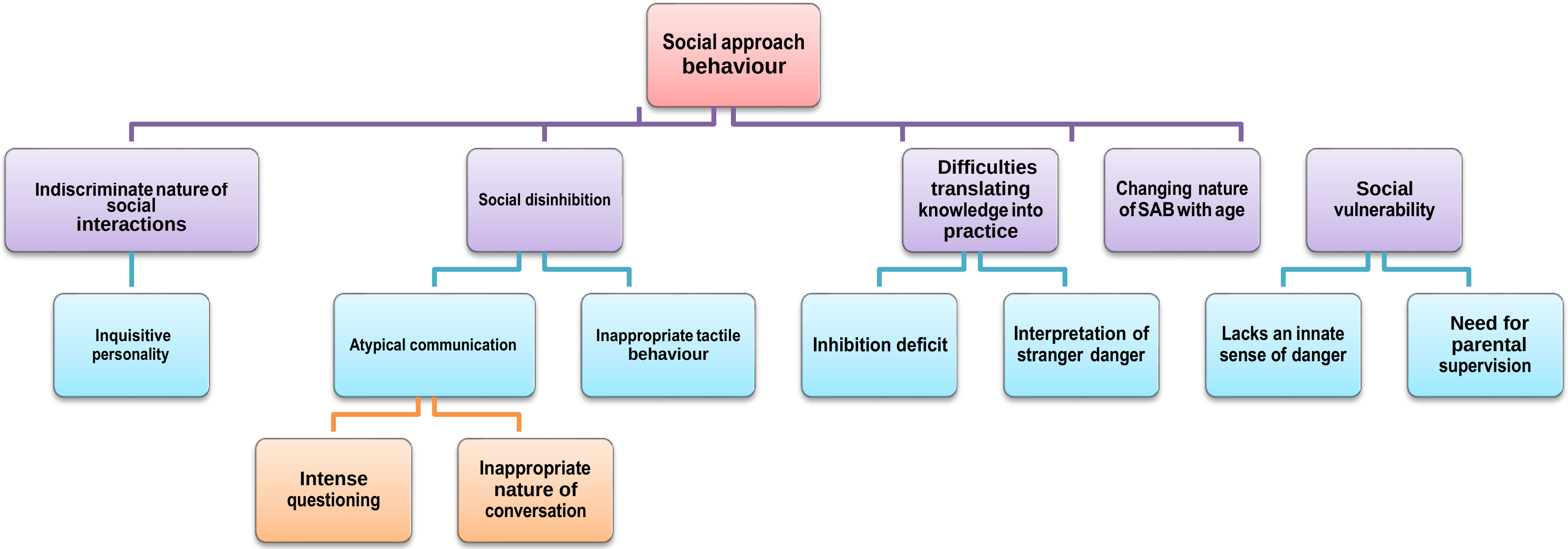

Figure 1: *Thematic analysis framework – parents' perceptions of their child with William syndrome's atypical SAB*

5.3. Discussion

Semi-structured interviews were conducted with the parents of a sample of children with WS. Through this qualitative methodology, descriptive information was obtained regarding the children's interactions with strangers. Across the interviews, several key themes were identified, including the indiscriminate nature of the child's SAB, social disinhibition within interactions and social vulnerability due to a naivety to danger. Previous researchers primarily used rating scale tasks and parental questionnaires to measure SAB. The accumulated findings provided support for high levels of SAB in children with WS (Bellugi et al., 1999; Doyle et al., 2004; Järvinen-Pasley et al., 2010; Jones et al., 2000; Martens et al., 2009). The present study builds on previous research by obtaining a qualitative insight into parents' accounts of this atypical SAB, including the manifestation of SAB and the wider implications of this behaviour for the child with WS and his or her family.

Only one study to the author's knowledge has achieved a qualitative investigation into SAB in children with WS. Lough et al. (2016) solicited parents' accounts on their child's SAB. Several themes were identified from this study, including the children's social disinhibition around strangers and heightened social vulnerability. With consideration to the high levels of heterogeneity in WS (Little et al., 2013; Porter & Coltheart, 2005) and the lack of qualitative research which has been conducted thus far, it was deemed important to further investigate the SAB of children with WS.

A dominant theme across the extracts was the indiscriminate nature of the children's SAB. Several parents reported that their child did not discriminate between familiar and unfamiliar people when seeking out social engagement, with this illustrated across their communication patterns and tactile displays. This builds substantially on previous research which measured parents' ratings on their child's SAB towards familiar and unfamiliar people (Doyle et al., 2004; Jones et al., 2000). The current findings provide further insight into the manifestation of SAB in a real-life situation and parents' understandings of this behaviour, with several parents attributing this in part to their child's inquisitive nature. These findings align with Lough et al. (2016) who reported that levels of SAB among the children with WS varied in accordance with the personality traits of the child, where the children who were more extroverted, showed higher levels of SAB.

Several parents reported that their child often engaged in inappropriate tactile behaviour towards strangers and lacked an awareness of personal space boundaries. Previous research by Lough et al. (2015) used parent questionnaires to assess personal space regulation in WS. The current study builds on this by identifying the contexts within which these behaviours are demonstrated and the indiscriminate nature of these physical interactions. This disinhibited behaviour was reported as a concern by several parents. However, there was within-syndrome variance in the parents' reports, where some children were reported to discriminate between familiar and unfamiliar people when engaging in these tactile behaviours. This highlights the heterogeneity in the social behaviour of children with WS in line with previous findings (Little et al., 2013; Porter & Coltheart, 2005).

Previous research identified substantial communication deficits in children with WS (Laws & Bishop, 2004; Stojanovik, 2006). The current findings build on this by providing insight into the implications of the child's atypical patterns of communication for his or her potential social vulnerability around strangers. Several parents reported that their child often engaged in intensive questioning when they met someone for the first time. Several parents noted that their child had a stock set of questions that they repeatedly used within social interactions. This intense questioning could be accounted for by impairments to the EF of cognitive flexibility (Rhodes et al., 2010). Cognitive flexibility is the ability to shift attention or mental thoughts from one construct to another. It could be suggested that children with WS fixate on one mental task and have difficulty disengaging from this task during a social interaction. This could account for the children's engagement in repetitive questioning during social interactions.

Several parents reported that their child often asked what could be considered to be inappropriate or personal questions within social encounters with strangers. This further illustrates the children's non-discriminate interactions, where many of the children with WS showed over friendliness with other people, regardless of the familiarity of the person. This suggests that children with WS may lack an awareness of social boundaries and social norms. During typical development, children begin to develop a basic understanding of socially appropriate behaviour in early childhood which regulates their actions within social interactions (Csoti, 2001; Perner & Lang, 1999). Given the older age range of the children in the present study (7-16 years), it could therefore be suggested that this social awareness may be impaired in WS, potentially accounting for this disinhibited behaviour. Several of the parents reported that their child's intentions were well meaning and without malice and attributed this to their child's naivety. This implies an impairment in social cognition, where the children with WS are suggested to have difficulties interpreting social cues and moderating their behaviour with consideration of the recipient’s potential discomfort. Alternatively, the children’s atypical patterns of communication may reflect his or her executive inhibition deficits, where these children are suggested to struggle to inhibit their verbal interactions. These findings align with the results reported in chapter 3 of the present thesis, where the children’s impairments in social awareness and social cognition were positively associated with their levels of SAB.

Previous research predominantly assessed SAB at a single time point (Doyle et al., 2004; Jones et al., 2000). The current research builds on this by soliciting parents’ observations of changes in their child’s SAB across age and development. Although approach behaviours were reported by parents across the sample, several parents noted that the inappropriateness of their child’s SAB had declined with age. Several parents reported that their child still had an innate drive towards social interactions; however, was better able to manage these behaviours than when they were younger. This decline in inappropriate behaviour was attributed in most cases to teaching and repetitive practice of socially appropriate behaviour, with some parents describing this as a learnt behaviour. The school was also credited by some parents with playing a role in their child’s understanding of social boundaries and self- management of their behaviour.

In many cases parents reported that their child still engaged in high levels of SAB; however, the context with which their child engaged in SAB had become more appropriate. One parent reported that her daughter was able to regulate her behaviours according to the situation and environment; often identifying an appropriate person to converse with based on their professional status, for example shop attendants. While this could still be argued to constitute approaching a stranger, this behaviour was considered to be more acceptable by the parents. This highlights a level of understanding of stranger danger principles through this child's ability to differentiate between different types of strangers.

Several parents also noted a transition in strangers' reactions to their child's overfriendly demeanour in line with the child's increasing age. This suggests that as the children get older and the gap between them and their TD peers widens, the atypical nature of their behaviours become more apparent, with strangers often experiencing discomfort at the child's friendly overtures which could be suggested to go against societal norms of acceptable behaviour. It was evident from the interview data that there were increased levels of concern among parents regarding the inappropriateness of their child's behaviour as they reached the adolescent period, with these behaviours suggested to become more problematic with age. The current study sheds light on the transition in the nature of the child's SAB with age and the implications of this for the child's vulnerability and parental concerns. It would be valuable within future research to examine these age related changes further by using a longitudinal design to assess the developmental changes in SAB across childhood and adulthood. These findings are suggested to have potentially important implications in the design of interventions and support for individuals with WS by expanding the understanding of the difficulties experienced at different stages of development.

To the author’s knowledge, while previous research has examined stranger awareness and SAB independently, the current study was the first to apply a qualitative analysis to examine the relationship between these constructs. Many of the children with WS were reported to show an understanding of stranger danger concepts; however, struggled to translate this awareness into practice during social interactions. Across the extracts, there was evidence of an inner conflict among the children as they attempted to regulate their social impulses, while also trying to appease their parents. This constant battle was evident through the children’s efforts to self-regulate their behaviour by physical restraint. Furthermore, several of the children became upset following their interactions with strangers. This indicates a level of social-cognitive understanding, where the children were aware of the implications of their behaviour, which in turn resulted in frustration felt by the parents. Further research is sought to identify which factors are disrupting this pathway between knowledge and practice. In line with the assumptions of the frontal lobe hypothesis (Little et al., 2013; Porter et al.,, 2007), it could be suggested that children with WS, similarly to patients with frontal lobe damage, understand the rules regarding socially appropriate behaviour; however, are unable to perform these actions due to their executive inhibition deficits which impair their ability to regulate their actions.

The present data develops previous questionnaire findings which indicated high levels of social vulnerability in WS (Fisher et al., 2013) by expanding our understanding of the nature of this vulnerability. This study identified the role of self-regulation deficits and poor social judgements in the children’s social drive to approach and engage with strangers.

While several parents perceived their child to have an understanding of stranger danger, these parents often attributed their child’s social vulnerability around strangers to deficits in self-regulation, where the child was reported to struggle to physically inhibit their behaviours upon meeting a stranger. These findings can be suggested to provide support for the theoretical assumptions of the frontal lobe hypothesis which proposes that high levels of SAB in WS are caused by the child’s deficits within response inhibition, which result from underlying neural impairment to the frontal lobes.

High levels of vulnerability in WS were also attributed to the children’s poor social judgements. Many of the children were reported to be highly trusting of other people. This was identified as a cause of concern among parents with regards to their child’s safety. Several parents believed that their child was particularly trusting of adults and would be easily led by a stranger due to their naivety to danger. One mother reported that her daughter identified a person as a friend as soon as she knew their name. This highlights the child’s misconceptions on what constitutes a trusted person and a friend, further highlighting the child’s potential vulnerability.

Several parents noted that their child did not show a level of wariness around strangers, as is shown during typical development (Benson & Haith, 2009). Parents, not surprisingly, tended frequently to draw on the experiences of the child’s typical siblings to illuminate differences in the individual with WS that the parent found problematic. One parent reported that her daughter with WS required significantly greater supervision than her younger siblings despite a difference in age.

Across the interview data, the inappropriateness of the child's SAB could be suggested to have implications for the parent-child relationship. Parents indicated their extensive efforts to protect their children's safety through educating them about stranger danger and regulating their behaviours through close supervision. While previous research identified high levels of SAB in WS, the current findings provide insight into the implications of this behaviour for the child and wider family unit. Across the interviews, parents' concerns regarding their child's lack of anxiety around strangers, disinhibited interactions with strangers and naivety to danger are suggested to pose implications for parents' wellbeing, stress levels and anxiety. The current findings also develop previous research by providing insight into the implications of the child's SAB for everyday family functioning, where an increased need for support and supervision posed implications for family life.

The themes derived from the present study have provided strong support for Lough et al. (2016) with social disinhibition, a naivety to danger and the need for parental supervision identified as prominent themes across both of these studies. Through a consideration of the heterogeneity reported in the social behaviour of children with WS (Little et al., 2013; Porter & Coltheart, 2005), the consistency between the present findings and the work of Lough et al. suggests that these experiences are key features of the WS social phenotype. In addition to providing strong support for these characteristic features of SAB in children with WS, the current research has expanded our understanding of SAB in children with WS by examining the transitions in the nature of this SAB across age and development, the disparity between the children's understanding of stranger danger and their practice of these principles, and the wider implications of SAB for the child and their family. These accumulated findings contribute substantially to expanding our understanding of SAB in children with WS.

Chapter 6: The characteristics of peer relations in children with Williams syndrome: A quantitative study with parents and teachers

6.1. Introduction

Individuals with WS can be characterised by a strong motivation to seek out social contact with other people (Bellugi et al., 1999; Dodd et al., 2010; Doyle et al., 2004) and a relative lack of fear when in the presence of strangers (Fisher, 2014; Jawaid et al., 2012). These high levels of SAB were illustrated in the experimental ratings of the children with WS in chapter 3 and further indicated across the parents' questionnaire ratings (chapter 3) and interview data (chapter 5).

Many individuals with WS have been reported to demonstrate atypicalities of social functioning. These atypicalities in social functioning have been suggested to hinder the success of the individual's social interactions (Laws & Bishop, 2004; Philofsky et al., 2007). Children and adults with WS demonstrate extensive social and communicative impairments (Laws & Bishop, 2004; Stojanovik et al, 2001). On the surface, individuals with WS appear extremely chatty and outgoing; however, researchers have reported that children with WS demonstrate substantial pragmatic language impairments (Laws & Bishop, 2004; Stojanovik et al., 2001). Many individuals with WS demonstrate what has been termed "cocktail party speech" (Laws & Bishop, 2004; Udwin, Yule, & Martin, 1987; Udwin & Yule, 1990). Individuals with WS frequently produce complex stereotyped social phrases; however, they often struggle to extend conversations beyond these polite social greetings (Laws & Bishop, 2004). These communication difficulties have evident implications for the success of peer relations among individuals with WS (Davies et al., 1998).

These atypicalities in social functioning have also been reported to be present within the domain of social-cognition. Children and adults with WS have been reported to demonstrate substantial difficulties identifying and interpreting relevant social cues during interactions (Davies et al., 1998; Laws & Bishop, 2004; Philofsky et al., 2007). Children with WS have been reported to show greater impairments in social cognition than TD children who have been matched on mental age and chronological age (Porter et al., 2008) and children with a non-specific intellectual disability (Tager-Flusberg & Sullivan, 2000), where the children with WS demonstrated significantly greater impairments across assessments of theory of mind. This accumulated research evidence highlights impairments in social language and social cognition in children with WS. As these are two important components of social functioning, these findings provide evidence for atypical social functioning in many children with WS.

Previous researchers have utilised a range of methodologies to measure social functioning impairments in children and adults with WS. These methods have included parental questionnaires, experimental tasks and behavioural observations (Klein-Tasman et al., 2007; Laws & Bishop, 2004; Stojanovik et al., 2001). Klein-Tasman et al. (2011) measured social and communicative impairments among children with WS aged between 6 and 15 years old. Parents and teachers completed the standardised Social Responsiveness Scale (SRS) (Costantino & Gruber, 2012). The SRS measures impairments across social cognition, social communication, social awareness, social motivation and the presence of autistic mannerisms. The children with WS were reported by their parents and teachers to show substantial impairments in social cognition, social communication and social awareness; however, the children were reported to demonstrate significantly fewer impairments within social motivation. The SRS only captures one aspect of social motivation, the children's social withdrawal. While children with WS do not show difficulties in social withdrawal, these children have been reported to engage in atypical patterns of social approach (Bellugi et al., 1999; Dodd et al., 2010). In light of this, the children's social motivation scores in this scale should be interpreted with caution. The findings from this study implicate the role of social and communicative impairments in children with WS's difficulties engaging in reciprocal social interactions with peers. This pattern of impairment was consistent across parent and teacher ratings. This suggests that impairments in social cognition, social communication and social awareness are core features of the WS social phenotype.

Klein-Tasman et al. (2011) noted that the parents rated the children with WS as significantly more impaired in social functioning than the teachers' ratings. This suggests that social impairments may be more prominent in the home environment, potentially due to the unstructured nature of this social setting. These findings highlight the importance of using a multi-informant approach when examining social behaviour in WS. This study builds on previous research which used a single informant approach (Greer et al., 1997) by providing insight into the children's social functioning across contexts. Very few studies thus far have achieved an in depth examination of peer relationships in children with WS across contexts. The current research aims to further develop these research findings by soliciting information from parents and teachers on the characteristics of peer relations among children with WS.

Peer relationships are an area of substantial difficulty for many children with neurodevelopmental disorders (Cuckle & Wilson, 2002; Hoza et al., 2005; Rowley et al., 2012). Children with ASD have been reported to experience high levels of social isolation and a poorer quality of peer relationships than their TD peers (Bauminger & Kasari, 2000; Rowley et al., 2012). Peer relationship difficulties and social isolation were also reported to be prevalent among children with ADHD (Bagwell et al., 2001; Diamantopoulo et al., 2005). Hoza et al. (2005) used a peer nomination method to measure peer relationships in children with ADHD. The children with ADHD were reported to be significantly less popular than their TD peers. Furthermore, only 56% of the children with ADHD had a reciprocal friendship with a classmate. The accumulated findings highlight that peer relationships are an area of difficulty for many children with neurodevelopmental disorders.

Impairments in social cognition and social communication have been identified as significant predictors of peer relations in children with neurodevelopmental disorders. Rowley et al. (2012) administered the autism diagnostic observation schedule to a sample of children with ASD. This paradigm measures social and communication skills by observing children across several structured play activities. Rowley et al. reported that there was a significant negative relationship between impairments in social communication and peer relationship quality among the children with ASD. Furthermore, Sibley et al. (2010) reported that impairments in social cognition were a significant predictor of peer relationship difficulties among adolescents with ADHD.

Researchers also identified a role for EF's in an engagement in successful peer relationships (Fabes et al., 1999). Miller and Hinshaw (2010) investigated the relationship between EF and peer relations in a sample of adolescents with ADHD. The participants completed a battery of EF tests which measured response inhibition, planning and working memory skills. Measures of peer relationships and social functioning were also solicited from parents and peers. The adolescents' EF performance was found to positively predict peer acceptance and social skills.

EF impairments, in particular, deficits in response inhibition, are prevalent in WS (Carney et al., 2013). Although this was not found in the present thesis (chapter 4), this can be accounted for by the task utilised. These impairments in response inhibition have been shown to be expressed cognitively (Carney et al., 2013; Porter et al., 2007), and behaviourally within social interactions (Davies et al., 1998; Elison et al., 2010). Researchers indicated that individuals with WS showed difficulties with personal space regulation (Lough et al., 2015) and disinhibited tactile behaviour during interactions with both strangers (Lough et al., 2016) and peers (Davies et al., 1998). While researchers have identified a significant relationship between EF's and peer relations in neurodevelopmental disorders, such as ADHD, no researchers thus far have directly examined for a relationship between self-regulatory deficits and peer relations in children with WS.

The previous findings of an association between impairments in social functioning, impairments in EF and peer relations in the neurodevelopmental disorders of ADHD and ASD align with psychological frameworks which propose social cognition, social communication skills and regulatory skills to underlie a child's engagement in successful social interactions (Hay et al., 2004). There is evident overlap between

WS and the disorders of ASD and ADHD with their shared cognitive, social and behavioural impairments (Asada & Itakura, 2012; Costanzo et al., 2013; Klein-Tasman, Phillips, Lord, Mervis, & Gallo, 2009; Rhodes et al., 2011). In light of these shared impairments, there is a need to further examine the role of social functioning and EF in the peer relationships of children with WS.

A relatively small body of research has investigated peer relationships in WS. Fisher, Lough, Griffin and Lane (2017) conducted interviews with a sample of adolescents and adults with WS about their experiences of bullying. 53% of the individuals with WS had experienced physical bullying and 80% had experienced verbal bullying. Furthermore, Fisher et al. noted that 60% of the individuals with WS reported that they had experienced social exclusion from their peer group.

Researchers have provided support for claims of substantial peer relationship difficulties and high levels of social exclusion in adults with WS. Davies et al. (1998) administered several standardised questionnaires to the parents of a sample of adults with WS. These questionnaires measured various aspects of social and adaptive functioning. Adults with WS were reported to demonstrate substantial difficulties establishing friendships, high levels of social isolation and inappropriate behaviour within social interactions which included an engagement in over-tactile behaviour towards peers. Adults with WS have also been reported to show a low level of engagement in extra-curricular clubs and activities with their peers (Udwin, 1990).

Only a few researchers thus far have investigated peer relationships in younger children with WS. Greer et al. (1997) measured social skills and adaptive functioning in a sample of children with WS (4-16 years old) by soliciting parent ratings across standardised scales. 60% of the children with WS (N=9) were reported to demonstrate evident social problems. Furthermore, many of the children with WS had few enduring social contacts and a low level of engagement in extra-curricular activities. This supports previous findings of high levels of social isolation among children with WS (Gosch & Pankau, 1994; Gosch & Pankau, 1997; Udwin & Yule, 1991; Udwin et al., 1987).

Previous research has provided an initial insight into peer relationship difficulties among children and adults with WS. These studies have several methodological limitations which should be addressed within future research. The majority of the researchers who have investigated peer relationships in WS did not make comparisons between the individuals with WS and the normative population (Davies et al., 1998; Greer et al., 1997). Consequently, it is difficult to draw conclusions on the extent of social impairment among these individuals. Furthermore, the researchers used questionnaire schedules which measured a broad range of social behaviours (Davies et al., 1998; Greer et al., 1997; Udwin, 1990). Further research is required to selectively focus on peer relationships in WS in order to obtain a greater depth of insight into the characteristics of these social difficulties. In particular, there is a need to utilise measures which tap into the characteristic features of peer relationships in children with WS by soliciting information from both parents and teachers. This will build on previous research which has predominantly used a single informant methodology.

6.1.1. Research aims

The present study aims to examine the characteristics of social functioning and peer relationships in children with WS. Parents' and teachers' perceptions will be solicited in order to investigate social behaviour in children with WS across environmental contexts. In light of previous findings of predictive relationships between social functioning impairments, EF and peer relationships in neurodevelopmental disorders, the second aim of this study is to examine the role of psychological processes, specifically response inhibition and social functioning skills, in the quality of peer relationships in children with WS. In order to achieve these research aims, the present study will address five research questions.

6.1.2. Research questions

1. Do children with WS demonstrate impairments within social and communicative functioning?
2. Do children with WS demonstrate greater peer relationship difficulties than TD children?
3. What are the characteristic features of peer relationships among children with WS?
4. Are chronological age, verbal ability, non-verbal ability, self-regulatory skills and the severity of social and communicative impairments significantly associated with the quality of peer relationships among children with WS?
5. Are the peer relationships of children with WS stable across parents' and teachers' ratings?

6.2. Method

6.2.1. Participants

The parents of 21 children with WS (7.50-16.49 years, M: 11.83, SD: 2.69, 12F, 9M) were recruited. 14 of the children with WS attended a mainstream school and seven children attended a SEN school. The class teachers of the children with WS were also contacted, and 18 consented to take part.

The children with WS were individually matched with TD children on verbal ability in the BPVS II (Dunn et al., 1997) and with another group of TD children on non-verbal ability in RCPM (Ravens et al., 1998) in order to make between group comparisons on a measure of social functioning which was administered in the current study (the SDQ). As not all of the TD children had received SDQ ratings from both their parent and their teacher, the children with WS were matched with two groups of TD controls (verbal and non-verbal matched) to compare ratings on the parent SDQ and matched with two other groups of TD controls (verbal and non-verbal matched) to compare ratings on the teacher SDQ. This resulted in four TD comparison groups, two matched on verbal ability and two matched on non-verbal ability.

6.2.1.1. Participant matching

6.2.1.1.1. Matching for parent SDQ ratings

The children with WS were individually matched with TD children on verbal ability in the BPVS II. The TD matched control group were aged 5.08-7.92 years (M: 6.38, SD: .93; 10F, 11M). An independent samples t-test indicated that there was no significant difference between the performance of the children with WS (M: 67.05, SD: 19.52) and TD controls (M: 63.76, SD: 14.53) on the BPVS, *t* (40) = .62, *p* = 540. The children with WS were individually matched with TD children on non-verbal ability in RCPM. The TD non-verbal matched control group were aged 5.02 – 7.96 years (M: 5.97, SD: .92; 12F, 9M). An independent samples t-test indicated that there was no significant difference between the performance of the children with WS (M: 16.67, SD: 3.79) and TD controls (M: 16.62, SD: 3.49) on the BPVS, *t* (40) = .04, *p* = .966.

6.2.1.1.2. Matching for teacher SDQ ratings

The children with WS were individually matched with TD children on verbal ability in the BPVS II. The TD verbal matched control group were aged 5.08 – 7.92 years (M: 6.7, SD: .90; 10F, 8M). An independent samples t-test indicated that there was no significant difference between the performance of the children with WS (M: 70.56, SD: 18.74) and TD controls (M: 66.83, SD: 14.01) on the BPVS II, *t* (34) = .68, *p* = .504. The children with WS were individually matched with TD children on non-verbal ability in RCPM. The TD non-verbal matched control group were aged 5.02 – 7.96 years (M: 6.07, SD: .97; 10F, 8M). An independent samples t-test indicated that there was no significant difference between the performance of the children with WS (M: 17.11, SD: 3.72) and TD children (M: 17.39, SD: 3.65) in RCPM, *t* (34) = -.23, *p* = .823.

To compare scores on the Vineland Adaptive Behavior Scales (Sparrow et al., 2005), the performance of the children with WS was compared to the published scores of a normative population sample (Sparrow et al., 2005).

6.2.2. Measures

6.2.2.1. The Social Responsiveness Scale - SRS (Constantino & Gruber, 2012)

The SRS contains 65 items which measure five sub-domains of social functioning. A score is generated for each of: the child's social awareness, social cognition, social communication, social motivation and restricted interests and repetitive behaviour. A total score is also generated across these domains termed the T- score which represents the degree of the child's social impairments relative to TD children matched on gender. This scale showed strong internal reliability when administered with the present participant sample (α = .84). Further details on administering this scale are provided in chapter 2.

6.2.2.2. The Vineland Adaptive Behavior Scales, Second Edition – Survey Interview Form (Sparrow et al., 2005).

Due to the lengthy nature of this scale and the broad scope of behaviours which are measured, only the socialisation domain was administered in the present study. The socialisation construct contains items which measure the individuals' behaviour across three sub-domains: interpersonal relationships, play and leisure time and coping skills. The participants' scores across these three sub-domains are combined to form a domain composite score. The socialisation scale showed strong internal consistency for this participant sample (α = .76).

6.2.2.3. The Strengths and Difficulties Questionnaire (Goodman, 1997)

The SDQ measures five constructs of social functioning: emotional symptoms, hyperactivity/inattention, conduct problems, peer relationship problems and prosocial behaviour. This questionnaire demonstrated adequate internal reliability for the current sample across both parent (α = .69) and teacher ratings (α = .64). Further details on administering this scale are provided in chapter 2.

6.2.2.4. Peer interaction scale (parent report)

This 9 item questionnaire measures the children’s peer interactions across two sub-scales: the quality of the children’s peer relationships (α = .78) and tactile behaviour towards peers (α = .77). Full details on the development of this measure are provided in chapter 2.

6.2.2.5. Peer interaction scale (teacher report)

This questionnaire measure peer functioning across two constructs: the quality of peer relationships (α = .75) and tactile behaviour towards peers (α = .67). Full details on the development of this measure are provided in chapter 2.

6.2.3. Procedure

The children with WS and their parents were visited at their home. The children with WS and TD control children completed the BPVS II and RCPM as described in chapter 2. The parents and teachers of the children with WS completed the questionnaire measures as described above. The parents and teachers of the TD children completed the SDQ.

6.3. Results

6.3.1. Research question 1: Do children with WS demonstrate impairments within social and communicative functions?

There was adequate internal consistency within the parents' ratings in the SRS (α = .84). In addition to raw scores, T-scores were also calculated, where the children's social functioning performance was compared to a normative sample of children of the same gender. According to Constantino & Gruber (2012), a T score of below 60 indicates that the child is within the normal range of social functioning, a score of 60-65 indicates that the child shows mild impairments within daily social interactions, a score between 66 and 75 indicates that the child shows moderate impairments within daily social interactions and scores above 75 indicate severe impairments within social interactions. The descriptive statistics for the raw and T-scores in the SRS are reported in table 7.

Table 7:

Mean, SD and range of scores for the children with WS on the Social Responsiveness Scale (N=21)

	Raw score		**T-Score**	
	Mean (SD)	Range	Mean (SD)	Range
Social awareness	12.81 (3.40)	4 – 18	73.33 (10.48)	47 – 89
Social cognition	21.10 (5.46)	8 – 30	78.52 (9.72)	55 – 90
Social communication	29.62 (8.49)	12 - 47	72.29 (9.62)	53 – 90
Social motivation*	11.48 (5.69)	0 – 23	62.48 (12.48)	38 – 89
Autistic mannerisms	21.33 (7.55)	4 – 30	79.48 (12.01)	50 – 90
SRS total score	94.10 (24.99)	28 - 131	75.95 (9.93)	50 - 90

*Impairments in social withdrawal

Table 7 demonstrates that the children with WS show high levels of impairment across all sub-scales of the SRS. The large range of scores also indicates substantial within-syndrome variance in social functioning among the children with WS.

Table 8 reports the frequency of children with WS who were classified (following Constantino & Gruber, 2012) in the normal range of social functioning, mild impairments, moderate impairments or severe impairments in the sub-scales and total scores in the SRS.

Table 8:

Frequency (N and %) of children with WS who scored in each SRS impairment category

	Normal range	Mild impairment	Moderate impairment	Severe impairment
Social awareness	2 (9.52%)	3 (14.29%)	5 (23.8%)	11 (52.38%)
Social cognition	1 (4.76%)	0	6 (28.57%)	14 (66.67%)
Social communication	3 (14.29%)	2 (9.52%)	12 (57.14%)	4 (19.05%)
Social motivation	6 (28.57%)	7 (33.33%)	6 (28.57%)	2 (9.52%)
Autistic mannerisms	3 (14.29%)	1 (4.76%)	1 (4.76%)	16 (76.19%)
Total T-score	2 (9.52%)	1 (4.76%)	8 (38.10%)	10 (47.62%)

Table 8 indicates that the majority of the children with WS demonstrated moderate or severe impairments of social functioning, with only 2 children scoring within the normal range in the total T-score. The most severe impairments were reported in the social cognition, social awareness and autistic mannerism sub-scales, with only 2 children reported to demonstrate severe impairments in social motivation.

6.3.2. Research question 2: Do children with WS demonstrate greater peer relationship difficulties than TD children?

6.3.2.1. Ratings on the VABS socialisation scale

Parents rated the children with WS on the VABS socialisation scale. Mean, SD and range of scores on the VABS socialisation scale are reported in table 9 for the children with WS. Descriptive statistics are reported for the socialisation domain score and raw scores on the three sub-scales: interpersonal relationships, play and leisure time and coping skills. Published means and SD's are also reported for a normative child population sample (Sparrow et al., 2005).

Table 9:

Mean, SD and range of scores on the VABS socialisation domain (children with WS and a normative population sample)

	WS		**Normative population sample**
	Mean (SD)	Range	Mean (SD)
Interpersonal relationships	57.43 (12.82)	34 – 80	66.09 (6.67)
Play and leisure time	34.05 (15.68)	8 – 67	53.27 (5.59)
Coping skills	31.48 (12.16)	9 – 54	45.59 (10.47)
Socialisation Standardised score	73.38 (17.30)	50 – 119	100.45 (14.4)

Table 9 indicates that the children with WS demonstrate socialisation skills which are substantially below the normative population. The children with WS scored lower than the TD children in all sub-scales of the VABS socialisation scale and in the socialisation domain score. Play and leisure time was reported to be the most problematic sub-scale among the children with WS. The large range of scores across the sub-scales and the standardised socialisation score also indicates that there is substantial within-syndrome variance in the socialisation skills of children with WS.

Independent samples t-tests were conducted to compare the children with WS with normative population scores in the VABS socialisation scale (Sparrow et al., 2005). The data met the parametric assumptions of an interval scale of measurement, normally distributed data points and no significant outliers. The children with WS were rated significantly lower than the normative population on the interpersonal relationships scale, t (20) = -3.10, p = .006 (two tailed); d = 0.889. The children with WS were rated significantly lower than the normative population on the play and leisure time scale, t (20) = -5.62, p < .001 (two tailed); d = 1.807. The children with

WS were rated significantly lower than the normative population on the coping skills scale, *t* (20) = -5.32, *p* < .001 (two tailed); *d* = 1.247. The children with WS were rated significantly lower than the normative population in the standardised socialisation score on the VABS, *t (*20) = -7.17, *p* = < .001 (two tailed); *d* = 1.708.

6.3.2.2. Ratings on the SDQ peer problems sub-scale

The parents' and teachers' ratings in the peer problems sub-scale of the SDQ are reported in table 10. The mean, SD and range of scores are reported for the children with WS and TD comparison children who were matched on verbal and non-verbal ability.

Table 10:

Mean, SD and range of scores on the parent and teacher SDQ peer problems sub-scale (children with WS and TD comparison groups)

	WS (N=21)		TD verbal matched controls (N=21)		TD non-verbal matched controls (N=21)	
	Mean (SD)	Range	Mean (SD)	Range	Mean (SD)	Range
Parents' ratings	4.95 (2.04)	0 – 7	1.62 (1.63)	0 – 6	1.10 (1.22)	0 – 4
Teachers' ratings	3.67 (2.17)	0 – 7	1.17 (1.82)	0 - 6	0.61 (1.09)	0 – 3

*Possible values range from 0 – 10

As highlighted in table 10, the children with WS showed greater peer problems than TD children who had been matched on verbal and non-verbal ability. This pattern was present across parent and teacher ratings.

An analysis of skewness statistics indicated that the data were non-normally distributed. Furthermore, an observation of Q-Q plots indicated that there were significant outliers in the dataset. In consequence, the Kruskal-Wallis H test was used to analyse the data. The data met the assumptions for this non-parametric test.

The parents' ratings of peer problems were significantly different between the groups, $\chi2$ (2) = 27.902, $p < .001$, $r = .45$. Subsequently, pairwise comparisons were conducted using Dunn's (1964) procedure. This post hoc analysis revealed statistically significant differences in peer problem ratings between the children with WS (Mean rank = 29.45) and TD children matched on verbal ability (Mean rank = 13.55), $\chi2$ (1) = 17.988, $p < .001$, $r = .44$. There was also a statistically significant difference in peer problems ratings between the children with WS (Mean rank = 30.33) and TD children matched on non-verbal ability (Mean rank = 12.67), $\chi2$ (1) = 22.327, $p < .001$, $r = .54$. There was no significant difference between the peer problem ratings of the TD children who were matched on verbal ability (Mean rank = 23.38) and the TD children matched on non-verbal ability (Mean rank = 19.62), $\chi2$ (1) = 1.063, $p = .303$.

The teachers' ratings of peer problems were significantly different between the groups, $\chi2$ (2) = 20.236, $p < .001$, $r = .32$. Post hoc analysis revealed that there was a statistically significant difference in peer problem ratings between the children with WS (Mean rank = 24.14) and TD children matched on verbal ability (Mean rank =

12.86), $\chi 2$ (1) = 10.918, *p* = .001, *r* = .55. There was also a statistically significant difference in peer problems ratings between the children with WS (Mean rank = 25.47) and TD children matched on non-verbal ability (Mean rank = 11.53), $\chi 2$ (1) = 17.217, *p* < .001, *r* = .86. There was no significant difference between the peer problem ratings of the TD children matched on verbal ability (Mean rank = 19.72) and the TD children matched on non-verbal ability (Mean rank = 17.28), $\chi 2$ (1) = .691, *p* = .406.

6.3.3. Research question 3: What are the characteristic features of the peer relationships of children with WS?

The parents and teachers of the children with WS completed a bespoke questionnaire which measured the quality of the children’s peer relationships and levels of tactile behaviour within peer interactions. The mean, SD and range of scores in these scales are presented in table 11. A higher score in each scale indicates a higher quality of peer relationships or a higher level of tactile behaviour.

Table 11:

Mean, SD and range of scores for the children with WS in the parent and teacher peer interaction scales

	Mean (SD)	Possible min value	Possible max value	Range
Parent rated peer relation quality	18.62 (5.63)	6	30	8 – 29
Parent rated tactile behaviour towards peers	11.38 (2.65)	3	15	4 – 15
Teacher rated peer relation quality	39.65 (7.47)	13	65	29-56
Teacher rated tactile behaviour towards peers	7.24 (3.07)	3	15	3 – 11

Table 11 indicates that there were high levels of within-group heterogeneity in the quality of the children's peer relationships. This within-syndrome variance was reported by parents and teachers.

Frequency statistics were computed using the parents' and teachers' ratings across the individual items on these scales. The items assessed parents' and teachers' perceptions of the quality of the child's peer relationships, inclusion by peers and the appropriateness of their social behaviour. All items in the scales were measured on a 5 point Likert scale, where 1 indicates strongly disagree and 5 indicates strongly agree. A rating of 4 or 5 (agree or strongly agree) was computed as the endorsement of an item.

Table 12 and 13 provide frequency data from the parents' and teachers' ratings in these questionnaires. Frequencies are reported for the sample as a whole. Individual frequencies are also reported by school type: the children who attended mainstream school and the children who attended a special educational needs (SEN) school.

Table 12:

Frequency statistics (N and %) on parents' reports of the characteristics of peer relations in children with WS (N=21)

	Total sample (N=21)		Mainstream school (N=14)	SEN school (N=7)
	N	%	N	N
Has one or more close friends	9	42.9	5	4
Ability to maintain friendships for several months or longer	10	47.62	7	3
Frequent fall outs with peers	5	23.81	4	1
High levels of tactile behaviour towards peers	16	76.19	12	4
Awareness of personal space boundaries	3	14.29	2	1
Invited to peers' homes	8	38.10	5	3
Invited to peers' birthday parties	10	47.62	5	5
Attends extra-curricular clubs/activities with peers	13	61.90	8	5

Table 13:

Frequency statistics (N and %) on teachers' reports of the characteristics of peer relations in children with WS (N=18)

	Total sample (N=18)		Mainstream school (N=12)	SEN school (N=6)
	N	%	N	N
Has a best friend (reciprocal)	3	16.67	1	2
One or more close friends in the class	8	44.44	5	3
Difficulties forming friendships	6	33.33	4	2
Difficulties maintaining friendships	7	38.89	7	0
Frequent fall outs with peers	4	22.22	4	0
Included in conversations by peers	7	38.89	5	2
Chosen as a partner for work activities/P.E.	5	27.78	3	2
Included by peers during break/lunch time	7	38.89	4	3
Picked on by peers	0	0	0	0
Greeted by other children	14	77.78	11	3
Over-tactile towards peers	4	22.22	3	1
Awareness of personal space boundaries	6	38.89	3	3

Table 12 and 13 indicate that peer relationships are an area of substantial difficulty for many of the children with WS. This pattern was present across both the parent and teacher ratings. Less than half of the children with WS were reported by their parents and teachers to have a close friendship with a peer. However, the presence of friendships was higher among the children who attended a SEN school than those who attended a mainstream school. Table 12 indicates that less than half of the children with WS were reported by their parents to be invited to the homes of the child's peers or to the birthday parties of their peers, and just over 60% of the children were reported to attend extra-curricular clubs and activities with their peers. Less than 40% of the children with WS were reported by their teacher to be involved in conversations with their peers and to be included by their peers during recreational breaks. Furthermore, just over one quarter of the children with WS were chosen by their peers as a partner for work activities and P.E. These findings suggest that social exclusion is an area of difficulty for many children with WS, with this reported within parents' and teachers' accounts. The findings indicate that social exclusion was more problematic among the children with WS who attended a mainstream school, when compared to the children who attended a SEN school. However, caution is required when making comparisons across school type with consideration to the small sample sizes recruited, in particular for the SEN group (N=7). Furthermore, while initial observations are drawn between these groups, these differences were not qualified through statistical significance testing.

Over three quarters of parents reported that their child demonstrated high levels of tactile behaviour towards peers; however, less than one quarter of teachers reported an engagement in high levels of tactile behaviour towards peers. Less than 15% of the children were reported by their parents to show an awareness of personal space boundaries within peer interactions, with just under 40% of teachers reporting that the child with WS showed a poor awareness of personal space boundaries.

Despite difficulties establishing friendship and high levels of social exclusion, peers were reported to be friendly towards the children with WS. Over three quarters of the children with WS were reported to be greeted by other school children, and none of the children were reported to have been picked on by their peers.

The accumulated findings highlight that peer relationships are an area of substantial difficulty for many children with WS. Many of the children with WS were reported by their parents and teachers to experience difficulties forming and maintaining friendships with their peers and to experience high levels of social exclusion from their peer group. However, these difficulties were more prevalent among the children who attended a mainstream school than those who attended a SEN school. Despite these difficulties within friendships and experiences of social exclusion, none of the children with WS were reported to be bullied by their peers.

6.3.4. Research question 4: Are chronological age, verbal ability, non-verbal ability, self-regulatory skills and the severity of social and communicative impairment associated with the quality of peer relationships in children with WS?

Parent ratings of the children’s peer problems (SDQ) were non-normally distributed and therefore the data violated the assumptions required when conducting parametric tests. In consequence, Spearman’s rank order correlations were conducted to examine for a relationship between chronological age, verbal ability, non-verbal ability, social functioning impairments, response inhibition and parent rated peer problems (SDQ). As there were no significant relationships between these variables ($p > .05$), these are not reported further.

For all other variables, the data met the parametric assumptions of an interval scale of measurement, no significant outliers, linearity of variables and normally distributed data points. Bivariate Pearson’s correlations were conducted to examine the relationships between chronological age, verbal ability, non-verbal ability, response inhibition and peer relations in children with WS. Across the correlational analysis, a more conservative alpha value of $p < .01$ was used to control for type 1 errors when conducting multiple tests.

There was no significant relationship between the variables of chronological age, verbal performance, non-verbal performance, and any indices of social functioning and peer relations in the children with WS (*p* > .05). There was also no significant relationship between response inhibition performance, disinhibited tactile behaviour and any indices of social functioning and peer relations in the children with WS (*p* > .05). As these analyses proved non-significant, these variables will not be reported further. The correlation output for the remaining variables is presented in table 14.

Table 14:

Pearson's bivariate correlations between social functioning impairments (SRS) and parents' and teachers' ratings of peer relations in children with WS

	1	2	3	4	5	6	7	8	9	10	11	12	13
1. Social awareness (SRS)		.64**	.48*	.29	.49*	.51*	.36	-.63**	-.47*	-.48*	-.10	-.33	.01
2. Social cognition (SRS)			.59**	.46*	.52*	.63**	-.58**	-.62**	-.59**	-.54*	.10	-.67**	-.44
3. Social communication (SRS)				.78***	.55**	.83***	-.51*	-.54*	-.51*	-.66**	.41	-.64**	-.43
4. Social motivation (SRS)					.65**	.88***	-.23	-.24	-.24	-.44*	.57*	-.51*	-.61**
5. Autistic mannerism (SRS)						.72***	-.40	-.46*	-.13	-.52*	.52*	-.46*	-.62**
6. T-Score (SRS)							-.33	-.43	-.28	-.51*	.41	-.49*	-.63**
7. Interpersonal relations (VABS)								.54*	.47*	.73***	-.24	.51*	.27
8. Play/leisure skills (VABS)									.70***	.72***	-.32	.56**	.35
9. Coping skills (VABS)										.64**	.00	.48*	.11
10. Standardised score (VABS)											-.30	.63**	.38
11. Teacher rated peer problems (SDQ)												-.59*	-.71**
12. Parent rated peer relation quality													.66**
13. Teacher rated peer relation quality													

*$p < .05$,** $p < .01$,*** $p < .001$

As reported in table 14, there were several significant relationships between the children's social functioning impairments, as measured in the SRS, and the children's peer relationships. Impairments in social awareness were negatively correlated with play and leisure time skills (*p* = .002) as measured in the VABS. Impairments in social cognition were negatively correlated with interpersonal relationships (*p* = .006), play and leisure time (p = .003) and coping skills (*p* = .005) as measured in the VABS. Impairments in social cognition were also negatively correlated with parent rated peer relationship quality (*p* = .001). Impairments in social communications were negatively correlated with the socialisation domain score (*p* = .001) as measured in the VABS, and parent rated peer relationship quality (*p* = .002). Autistic mannerisms were negatively correlated with teacher rated peer relationship quality (*p* = .007). These findings suggest that children with WS may struggle to engage in reciprocal social interactions with their peers due to their atypicalities in social functioning. This includes their difficulties interpreting another person's intentions during a social interaction, inflexible patterns of behaviour and poor awareness of socially appropriate practices of behaviour. All of these social skills are considered to play an important role in establishing and sustaining successful peer relationships.

6.3.5. Research question 5: Are the peer relationships of children with WS stable across parents' and teachers' ratings?

Measures of peer relationship quality and peer problems were obtained from the parents and teachers of the children with WS. Peer relationship quality scores were obtained using the total scores in the parent and teacher peer relation scales which were designed for application in the present thesis. Peer problem scores were obtained using the standardised SDQ. Table 15 presents the mean, SD and range of scores for each measure.

Table 15:

Mean, SD and range of scores for the children with WS on parent and teacher peer relationship measures

	Parents			**Teachers**		
	Mean (SD)	Possible values	Range	Mean (SD)	Possible values	Range
SDQ peer problems	4.95 (2.04)	0-10	1-7	3.67 (2.17)	0-10	1-7
Peer relationship quality	18.62 (5.63)	6-30	8-29	39.65 (7.47)	13-65	29-56

Table 15 highlights that the parents rated the children with WS higher in peer problems than the teachers. Ratings of peer relationship quality were similar between parent and report reports.

Paired samples t-tests were conducted to examine for a statistically significant difference in mean scores between the parent and teacher SDQ ratings. The parents rated the children with WS significantly higher in peer problems than the teachers, *t* (17) = -2.17, *p* = .045 (two tailed), *d* = .42. Parent and teacher ratings on the quality of peer relationships were converted to Z-scores in order to allow comparison between the means of different scales. There was no significant difference between the parents' and teachers' ratings of peer relationship quality for the children with WS, *t* (17) = .86, *p* = .404 (two tailed).

6.4. Discussion

6.4.1. Summary of findings

The findings across this chapter support substantial impairments within social functioning and peer interactions in children with WS. The children with WS were rated poorer than the normative population across several standardised measures of socialisation, with these impairments reported across social settings. Furthermore, the use of specifically designed questionnaires provided insight into the atypical nature of the child's peer relationships and the children's struggles to integrate within their peer group. Impairments in several aspects of social functioning were identified as strong underlying factors in the children's peer relationship difficulties. These findings will be evaluated further in the present discussion section.

6.4.2. Do children with WS demonstrate impairments within social and communicative functioning? (Research question 1)

The vast majority of the children with WS were reported to demonstrate moderate or severe impairments within social functioning. These findings have provided strong support for previous research (Klein-Tasman et al., 2011; Lough et al., 2015; Lough et al., 2016; Riby et al., 2014; Van der Fluit, Gaffney, & Klein-

Tasman., 2012). The use of the standardised SRS, in line with the methodologies of previous researchers, allowed reliable comparisons to be drawn between the research findings. When examining the percentage of individuals within each category of impairment, the current findings closely mirror previous research. Klein-Tasman et al. (2011) reported that 39% of children with WS scored in the severe range of impairments, 47.6% scored in the mild to moderate level of impairment and 13.4% scored in the normal range of social functioning. These previous findings closely align with the percentages reported for the present sample, where 47.7% of children with WS scored in the severe range of impairment, 42.9% scored in the mild to moderate level of impairment and 9.5% scored in the normal range of social functioning. The similarity in the percentages reported between these studies allow stronger conclusions to be drawn on levels of social impairment in individuals with WS. However, there were high levels of within-syndrome variance in the children's overall scores in the SRS. This finding provides further support for the heterogeneous nature of WS, in line with previous research (Little et al., 2013; Porter & Coltheart, 2005).

Following the SRS guidelines, over half of the sample of children with WS were classified as severely impaired within the social cognition, social awareness and autistic mannerism sub-scales, and over half of the sample were moderately impaired in the social communication sub-scale. Social cognition was identified as the most problematic area of social functioning among the children with WS. These findings provide further support for previous experimental research which has identified social cognitive deficits in children with WS, including substantial impairments within theory of mind performance (Porter et al., 2008; Tager-Flusberg & Sullivan, 2000). The present findings further support these claims of social cognitive impairments in children with WS by using a standardised questionnaire which has been widely validated with typical and atypically developing populations (Costantino & Gruber, 2012). These atypicalities in social cognition expressed themselves cognitively, where the children with WS were reported to have difficulties interpreting other people’s facial expressions and tone of voice, with these skills considered to be important in guiding appropriate social interactions. The children’s social cognitive impairments were also expressed behaviourally, where many of the children with WS were reported to have difficulties engaging in pretend play. These social-cognitive impairments can be suggested to pose implications for the children’s social interactions and peer relations.

6.4.3. Do children with WS demonstrate greater peer relationship difficulties than the normative population? (Research question 2)

The children with WS were rated significantly poorer in the socialisation domain of the VABS when compared to the published ratings of TD children. These findings are in line with previous research (Greer et al., 1997). However, the present study has developed previous research which administered the VABS by analysing the children’s socialisation scores at both the domain level and sub-domain level. This provides a greater depth of insight into the severity of peer relationship difficulties in children with WS. The children with WS performed significantly poorer than the normative population on the interpersonal relationships, play and leisure time and coping skills sub-scales of the VABS socialisation scale. This highlights that children with WS experience substantial difficulties during social interactions with others, difficulties engaging in effective play with their peers and a low level of participation in social activities. The children with WS performed poorer than the normative population across all sub-scales; however, they were rated the lowest in the play and leisure time sub-scale. This finding suggests that children with WS experience substantial difficulties engaging in effective play and activities with their peers. Many of the children with WS were also reported to show a lack of independence in planning and partaking in social activities with peers. This could be suggested to reflect EF impairments in WS within the domain of planning (Rhodes et al., 2010). These difficulties engaging in effective social exchanges and establishing reciprocal friendships can be suggested to provide an account for the high levels of social isolation in WS (Gosch & Pankau, 1997; Udwin & Yule, 1991) and social anxieties in WS (Riby et al., 2014). These findings could be used to inform the design of future interventions in order to support children with WS from a young age to develop stronger play and interaction skills.

Previous researchers have primarily used a single informant approach by soliciting parent ratings to examine social behaviour among children with WS (Greer et al., 1997; Udwin & Yule, 1991). Klein-Tasman et al. (2011) were one of the first research groups to utilise a multi-informant approach by soliciting social functioning ratings from the parents and teachers of a sample of children with WS. The present study builds on the findings of Klein-Tasman et al. (2011) by obtaining a measurement of both social functioning and peer relations from parents and teachers in order to expand our understanding of social strengths and difficulties in children with WS.

The children with WS were reported by their parents and teachers to demonstrate significantly greater peer problems than TD children who were matched on verbal ability and TD children matched on non-verbal ability. These findings suggest that peer problems in children with WS cannot be explained by the children's intellectual delay alone. It is suggested that these peer problems may be due to other aspects of the syndrome's phenotypic profile, for instance, cognitive or behavioural deficits. There was a significant association between impairments in social cognition, social communication, social awareness and peer problems among the children with WS in the current study. This highlights an integral role for social functioning skills in the peer relationships of children with WS.

There was within-syndrome variance in the children's ratings in the SRS and VABS, both at the sub-domain and domain levels. This indicates that although impairments in social functioning and peer relations were prevalent, there was variation in the levels of impairment within this sample. There was variance in the children's scores across the sub-scales of the SRS, as well as their overall level of social impairment. Furthermore, there were high levels of variance in the children's socialisation scores, ratings of peer problems and peer relationship quality. The within-syndrome variance within parents' and teachers' ratings of peer problems were larger than comparison TD groups. These findings provide support for the heterogeneous nature of the social behaviour of children with WS across social functioning and peer relations. This develops previous research findings which identified within-syndrome variance in cognition and behaviour in individuals with WS (Little et al., 2013; Porter & Coltheart. 2005) by illustrating within-syndrome variance in the social functioning of children with WS. While it is important to acknowledge the presence of within-syndrome variance in the social functioning and peer problem ratings of the children with WS, caution should be exercised when interpreting this variance as these distributions were not subjected to statistical analysis.

6.4.4. What are the characteristic features of the peer relationships of children with WS? (Research question 3)

Previous research has indicated that children with WS experience higher levels of social isolation and have fewer enduring social contacts than their TD peers (Gosch & Pankau, 1994; Greer et al., 1997). The present study builds on these research findings by examining the characteristics of the children's peer relationships in greater depth. The parents and teachers of the children with WS completed a questionnaire which measured several aspects of the child's peer relations, including the quality of their peer relationships, social inclusion by peers, engagement in social activities and the appropriateness of their social interactions. These questionnaires were designed to measure the characteristic features of peer relationships which are associated with the home and school environment.

Peer relationship difficulties were experienced by several of the children with WS. Less than half of the children with WS were reported to have a close friendship with another child. This difficulty in establishing friendships was reported by parents and teachers. This suggests that this is a prevalent feature of the WS social phenotype as these difficulties were reported to persist across social environments. However, friendships were more prevalent among the children with WS who attended a SEN school than a mainstream school. This suggests that SEN schools may provide greater opportunities for children to develop friendships with peers whose interests and abilities may be more on par with them. However, caution is required when making comparisons across school type with a consideration to the relatively small sample sizes recruited and the absence of statistical significance testing.

Social exclusion was identified as an area of particular difficulty inside and outside of school. Less than 40% of the children with WS were reported by their teachers to be included by their peers during recreational breaks. When the children were split by school type, there were higher levels of inclusion in the SEN schools, where 3/6 of the children who attended a SEN schools, compared to 4/12 of the children who attended a mainstream school were reported by their teacher to be included by peers during recreational breaks. Social exclusion was particularly evident in mainstream schools when pupils were required to choose partners for work activities, with only 3/12 of the children with WS reported to be chosen as a partner by their peers. It could be suggested that TD children may seek out partners of a similar intellectual ability to themselves in order to maximise their success in the work activity.

Less than half of the children with WS were reported to be invited to their peers' homes on a regular basis or to be invited to birthday parties. Social invites were higher among the children who attended SEN schools. This suggests that socialisation with other children, who experience similar difficulties and needs, often led to more opportunities to engage with peers outside of the school setting. Across the participant sample, attendance at birthday parties was significantly greater than visits to a peers' home. This suggests that the children may have acquaintances, such that they would be invited to a group activity or party; however, may be less likely to be selected as a best friend. The difference in friendships and social inclusion between the children who attended a mainstream and a SEN school highlight the integral role of the peer group on a child's social adjustment. While caution should be taken when drawing conclusions from a small sample, these findings can be suggested to support the advantages of SEN schools in providing an environment where children are able to form peer relations with others who are similar to them, in terms of intellectual and social needs and who may share similar interests to them.

This is in line with previous research on the importance of shared interests in the development of friendships (Benton, Hollis, Mahler, & Womer, 2011).

Previous researchers reported that children with WS had few enduring social contacts (Greer et al., 1997). The present study further develops these findings by investigating the characteristics of these peer relationship difficulties and the prevalence of these experiences among children with WS. Over 60% of the children with WS were reported to attend extra-curricular clubs and activities with their peers. This exceeds previous findings from the adult literature, where less than half of adults with WS were reported to participate in extra-curricular activities (Davies et al., 1998; Udwin, 1990). This difference in extra-curricular engagement suggests that there are greater opportunities for inclusion in clubs among children and adolescents with WS in comparison to adults. This difference could be accounted for through the increased opportunities available to this age group through the education system and similar establishments. Schools often provide opportunities for social engagement and provide a social network for children which may not be available to adults with WS. These findings also indicate a decline in individuals' participation in social activities with development. It would be valuable to investigate changes across development by conducting a longitudinal assessment of peer relations and social inclusion across childhood and adulthood in order to draw stronger conclusions on degrees of social inclusion in children and adults with WS.

Although the current findings demonstrate that many children with WS experience difficulties forming and maintaining friendships with their peers and experience relatively high levels of social exclusion, none of the children with WS were reported to be picked on by their peers. This suggests that the children's exclusion from their peer group may not be underpinned by malicious intentions. Instead, these peer relationship difficulties could be suggested to be a result of the children's social functioning deficits which may make sustaining friendships with their peers significantly more challenging. This absence of peer victimisation challenges the previous research findings of Fisher et al. (2017) who reported high levels of physical and verbal bullying among adolescents and adults with WS. This disparity in findings could be accounted for by the different age range of participants recruited in these studies. The present study recruited a sample of children aged between 7 and 16 years, in comparison to an older sample of adolescents and adults (12 to 37 years) who were recruited by Fisher et al. These findings suggest that bullying in WS may become heightened during later adolescence and adulthood. Furthermore, the present study used teacher reports as an assessment of bullying, in comparison to a self-report methodology which was used by Fisher et al. (2017). Teachers within a school environment often operate a zero tolerance policy regarding bullying. The low level of victimisation reported by teachers could be suggested to be a reflection of this.

Many of the children with WS were reported to engage in high levels of tactile behaviour during peer interactions. Tactile behaviour was reported to be more prevalent by parents, where three quarters of the parents, in comparison to only one quarter of teachers reported that the child with WS engaged in high levels of tactile behaviour towards their peers. These reports of tactile behaviour supports previous research findings where children with WS were reported to engage in disinhibited SAB, and is in line with the assumptions of the frontal lobe hypothesis (Bellugi et al., 1999; Little et al., 2013). However, the present study builds on these previous findings by measuring tactile behaviour specifically within peer interactions. This disparity between the parents' and teachers' ratings of tactile behaviour could be explained by a low tolerance of tactile behaviour within the school environment. School rules and regulations may restrict the children's display of tactile behaviour, with a core component of the school curriculum involving education and promotion of socially appropriate behaviour. In contrast, the home environment is often an unstructured setting, where there are potentially less rules regarding socially appropriate behaviour. In consequence, there are suggested to be greater opportunities for children to engage in inappropriate social interactions outside of the school setting.

Many of the children with WS were reported to have a poor awareness of personal space boundaries. This finding supports previous research by Lough et al. (2016) who reported that children with WS showed a poorer awareness of personal space boundaries than TD children and children with ASD. The present study builds on the findings of Lough et al. by measuring an awareness of personal space boundaries specifically within the context of peer relationships. These high levels of tactile behaviour and poor awareness of social boundaries could provide an explanation for the peer relationship difficulties which are reported in this sample of children with WS.

The present study builds on previous research findings by examining the characteristics of the peer relationships of children with WS and the prevalence of these experiences. Although there were evident patterns of peer relationship difficulties within this sample of children with WS, there was within-syndrome variance in the quality of the children's peer relationships, their levels of social inclusion and the appropriateness of their social interactions. This sheds light on the heterogeneous nature of social behaviour in WS, building on previous research findings (Little et al., 2013; Lough et al., 2016). However, the absence of normative data and scaling for the parent and teacher peer relationship scales limit the conclusions which can be drawn with regards to this within group variability.

Another potential challenge when conducting research with small sample sizes is the need to exercise greater caution when drawing conclusions on the generalizability of the findings to the wider population. This could also be argued to pose ethical implications given the time and involvement of participants. However, when working with rare populations, small sample sizes are often a common occurrence and these investigations allow important initial insights into a topic which has received very little attention previously. The present findings provided an important insight into the peer relationship experiences of children with WS and identified several problematic areas of social functioning. Future researchers should seek to further examine these atypicalities of social functioning in children with WS. This will allow stronger conclusions to be drawn regarding the extent to which these characteristics of peer relationships are generalizable to the wider population of children with WS.

6.4.5. Are chronological age, verbal ability and non-verbal ability, self-regulatory skills, and the severity of social and communicative impairment associated with the quality of peer relationships in children with WS? (Research question 4)

There were no significant relationships between chronological age, verbal ability and non-verbal ability and any measurements of peer relationships among the children with WS. These results suggest that peer relationship difficulties are not a consequence of intellectual delay alone, but are the result of one or several features of the WS phenotype. Furthermore, these findings indicate that peer relationship difficulties may be a stable aspect of WS, irrespective of age.

Previous researchers reported that inhibition was a significant predictor of peer relationship quality in children with ADHD (Miller & Hinshaw, 2010). The present study was the first, to the author’s knowledge, to examine the relationship between inhibitory control and peer relations in children with WS. There was no significant relationship between the children’s self-regulatory skills, as measured using a standardised cognitive task and behavioural observations of self-regulation skills during peer interactions, and the children’s peer relationships. It is suggested that the lack of statistical power in the current study, as a result of a relatively small sample size, may restrict the ability to reliability test this relationship. Several measures of social functioning were significantly associated with peer relations among the children with WS. Impairments in social awareness were negatively correlated with the children’s play skills as measured within the VABS. Social awareness is an understanding of socially appropriate behaviour and the ability to detect important social cues within interactions with peers (Constantino & Gruber, 2012). This ability to identify social cues is argued to be informative in guiding a child’s behaviour within a social interaction (Csoti, 2001). It is suggested that the children with WS may struggle to engage in effective play interactions with their peers due to their impairments in social awareness.

Impairments in social communication and social cognition were significantly negatively correlated with peer relationship functioning in the children with WS. Social communication allows a child to engage in reciprocal conversations with their peers and to share important thoughts and feelings in order to establish a close relationship (Laing et al., 2002). Previous researchers identified a significant negative relationship between social communicative impairments and peer relationship quality in children with ASD (Cappadocia, Weiss, & Pepler, 2012; Rowley et al., 2012). The present study is the first, to the author’s awareness, which has investigated the relationship between social communication and peer relationships in children with WS. The current findings suggest that social communication skills play an important role in establishing successful peer relationships in children with WS. Further research is sought to identify which shared features of these developmental disorders can account for this relationship.

Social cognition is a child's ability to interpret another person's intentions within a social interaction. This ability is vital for reciprocal social interactions as it guides a child's behaviour by allowing him or her to predict the intentions of their peer within an interaction (Meyer-Lindenberg et al., 2005). Furthermore, social cognitive skills are suggested to have a protective function as they increase a child's ability to identify if he or she is being taken advantage of by a peer and to withdraw themselves from such interactions (Constantino & Gruber, 2012). In line with this, the peer relationship difficulties in WS are suggested to be due to the children's impairments interpreting their peers' intentions within an interaction which are suggested to hinder their ability to respond appropriately.

The accumulated findings on the associations between social functioning impairments and peer relationship quality in children with WS emphasise the importance of these social skills. These findings are suggested to have the potential to inform the design of future social intervention to support the children with WS in developing stronger peer relationships.

Finally, there was a significant negative relationship between the children's autistic mannerisms, specifically their restricted interests and engagement in repetitive behaviours, and the quality of the children's peer relationships. During childhood, play has an integral role in a child's social development and formation of friendships (Parker et al., 2015). In order to develop successful peer relationships, it is important for children to show flexibility within their social interactions. Furthermore, shared interests provide a foundation for children to establish friendships (Benton et al., 2011). Consequently, it is suggested that the children with WS may struggle to develop effective friendships with their peers as a result of their restricted interests and often inflexible behaviour and need for routine (Rodgers et al., 2012).

6.4.6. Are the peer relationships of children with WS stable across parents’ and teachers’ ratings? (Research question 5)

The school environment is suggested to provide an optimal setting to measure peer relationships. The school setting provides many opportunities for children to interact with their peers and can provide valuable insight into peer relationship and social interaction deficits (Wentzel & Miele, 2016). Previous researchers have predominantly used a single informant methodology by measuring peer relationships from the perspective of the parent (Elison et al., 2010; Greer et al., 1997; Davies et al., 1998). Klein-Tasman et al. (2011) utilised a multi- informant methodology to examine social functioning in children with WS. Klein-Tasman et al. (2011) recruited the parents and teachers of the children with WS who completed standardised measures of social functioning. Both the parents and teachers reported that the children with WS showed substantial impairments within social functioning. However, the parents rated these impairments as significantly more problematic than the teachers. The present study builds on the findings of Klein-Tasman et al. (2011) by examining peer relationship across contexts.

The present study found that parents and teachers rated the children with WS similarly in peer relationship quality, where impairments were reported by both informants. This suggests that the characteristic features of peer relationships, which include difficulties establishing friendships and high levels of social exclusion, are present across contexts. Although parents and teachers rated the children with WS significantly higher than TD children on the SDQ peer problems index, the parents reported significantly more peer problems than the teachers. This suggests that while peer relationship difficulties are prevalent in the home and school environment, and are therefore a core feature of the WS social phenotype, these difficulties are more prominent in the home environment as reflected by higher parental ratings. This could be suggested to be due to the less structured nature of the home environment which may provide greater opportunities for peer difficulties to arise. Alternatively, this difference in ratings could be explained by a difference in the informants' reference of comparison. Parents may perceive their child with WS to experience substantial peer relationship difficulties when compared to the peer relationships of their TD siblings. In contrast, teachers may have a lower standard of peer relationship quality, particularly teachers within SEN schools. This difference in ratings between informants highlights the importance of recruiting both parents and teachers in order to develop a strong understanding of peer relationships in children with WS.

The findings across this chapter have provide support for claims of substantial deficits of social functioning and peer relations in children with WS. Many of the children with WS were reported by their parents and teachers to have difficulties forming and sustaining friendships, to show atypical patterns of behaviour and exclusion from peers' social activities. The children with WS were rated significantly higher than TD children across standardised measures of social functioning and peer relations. The children's difficulties in peer relations were reported to be significantly associated with atypicalities in social functioning within the domains of social cognition, social communication and social awareness.

Chapter 7: Peer relations in children with Williams syndrome: A qualitative study with parents, teachers and children with Williams syndrome

7.1. Introduction

Individuals with WS have been reported to demonstrate indiscriminate friendliness within their social interactions (Dodd et al., 2010; Doyle et al., 2004) as highlighted by an engagement in high levels of SAB (Bellugi et al., 1999; Järvinen-Pasley et al., 2010; Jawaid et al., 2012). These high levels of SAB were further demonstrated in chapters 3 and 5 of the present thesis. Despite their hyper-social phenotype, many individuals with WS have been reported to show impairments within reciprocal social interaction (Bellugi et al., 2007; Klein-Tasman et al., 2011). These atypicalities within social functioning (Klein-Tasman et al., 2011) in combination with the individuals' mild to moderate levels of intellectual impairment (Searcy et al., 2004), high levels of anxiety (Dykens, 2003; Rodgers et al., 2012) and repetitive behaviour and restricted interests (Janes, Riby & Rodgers, 2013) could be suggested to influence the success of peer relationships (Davies et al., 1998; Elison et al., 2010; Greer et al., 1997).

Previous research has provided support for atypical social functioning in children and adults with WS (Laws & Bishop, 2004; Klein-Tasman et al., 2011; Porter et al., 2008; Stojanovik et al., 2001). This was further illustrated in chapter 6 of the present thesis, where 38% and 47%, respectively, of the children with WS scored in the moderate and severe level of impairment in the SRS.

Many individuals with WS have been reported to show pragmatic language impairments (Laws & Bishop, 2004; Stojanovik, 2006). As reported in chapter 6, over three quarters of the children with WS were classified as demonstrating moderate or severe levels of impairment in social communication. These atypicalities in communication were further illustrated in chapter 5, where many parents reported that their child with WS demonstrated unusual patterns of communication, often asking inappropriate questions or making inappropriate statements during interactions with familiar and unfamiliar people. The present findings provide further support for atypical patterns of communication in WS, in line with previous research (Davies et al., 1998; Laws & Bishop, 2004; Philofsky et al., 2007; Rosner et al., 2004; Stojanovik, 2006; Udwin & Yule, 1991). These communicative impairments have been suggested to hinder the individuals' ability to engage in reciprocal social interactions (Stojanovik, 2001). This was illustrated in chapter 6, where impairments in social communication were negatively associated with parents' ratings of peer relationship quality among the children with WS.

Individuals with WS have also been reported to demonstrate a poor awareness of social norms and boundaries (Lough et al., 2015). This social naivety has been illustrated through the individuals' engagement in high levels of disinhibited behaviour during social interactions (Davies et al., 1998; Udwin, 1990). Lough et al. (2015) solicited questionnaire ratings from the parents of individuals with WS (aged 4-36 years). The individuals with WS were reported to have a significantly poorer awareness of personal space boundaries than individuals with ASD and TD controls. In chapter 6 of the present thesis, over three quarters of parents reported that their child with WS engaged in high levels of tactile behaviour during peer interactions, with only 14% of the children with WS reported to have an awareness of personal space boundaries. The accumulated findings suggest that personal space regulation is an area of substantial difficulty for many individuals with WS. Further research is sought to investigate the implications of this disinhibited behaviour for the peer relations of children with WS.

Despite a hyper-social personality profile (Jones et al., 2000), many children with WS have been reported to experience elevated levels of anxiety within novel environments and situations (Dykens, 2003; Leyfer et al., 2006; Leyfer, Woodruff-Borden, & Mervis, 2009). Rodgers et al. (2012) recruited the parents of a sample of children with WS (6 – 15 years). The parents completed the Spence Children's Anxiety Scale (Spence, 1998). The children with WS were reported to show clinically significant levels of anxiety. Peer relationships are often unpredictable across environments (Rubin & Ross, 2012). In consequence, it could be suggested that the children's experiences of anxiety may hinder the success of their peer relationships.

Previous research has provided an initial examination into the peer relationship difficulties of children with WS (Greer et al., 1997; Laws & Bishop, 2004; Udwin et al., 1997). Greer et al. (1997) administered questionnaires to the parents of a sample of children with WS (4 – 18 years old) to measure aspects of the children's social behaviour. Many of the parents indicated that their child had little or no enduring social contacts and low levels of inclusion in social clubs and activities with peers. In another study, Udwin et al. (1997) solicited ratings from the parents and teachers of a sample of children with WS (6 – 16 years) on various aspects of adaptive functioning. Peer relationships were identified as problematic for many of the children with WS. 84% of the children were reported by their parent to be socially isolated, while 50% of the children were rated as socially isolated by their teacher. This variance across ratings suggests that peer relations differ across settings and reinforces the importance of using a multi-informant approach to examine peer relationships across environmental contexts and across perspectives. These research studies have provided support for claims of peer relationship difficulties in children with WS.

The findings from the present thesis have developed previous research by using a multi-measure and multi-informant approach to examine the characteristics of peer relations in children with WS. As indicated in chapter 6, the children with WS were reported to show greater peer problems than TD comparison groups who had been individually matched on verbal ability and non-verbal ability. These impairments were consistent across parents' and teachers' reports. The findings from chapter 6 further developed previous research by examining parents' and teachers' perceptions of the characteristics of the children's peer relations. Many of the children with WS were reported to have few enduring social contacts and manifested difficulties forming and sustaining friendships with their peers. Several of the children with WS were also reported to be excluded from peer social activities and to have little interaction with their peers outside of the school context.

To the author's awareness, all of the studies conducted thus far to investigate peer relations in children with WS have administered questionnaires. Questionnaires are highly informative in providing an examination of levels of impairments and associations between variables (Fields, 2009); however, questionnaires can only provide a limited depth of insight into the experiences of the individuals (Brannen, 2017; Taylor, 2005). In order to identify and support children with WS within their interpersonal relationships, an in depth understanding of these atypicalities and social difficulties is sought using an interview method.

Semi-structured interviews have been conducted with the parents of adults with WS to solicit information on the individuals’ peer relations (Udwin, 1990; Davies et al., 1998; Elison et al., 2010). Davies et al. (1998) reported that 96% of the adults with WS were reported by their parent to have substantial difficulties forming and maintaining friendships. Furthermore, three quarters of the adults were identified as socially isolated. These interviews also provided insight into the atypicalities in the individuals’ peer interactions. Many of the adults with WS were reported to have a poor understanding of social norms and boundaries, showed disinhibited affection and tactile behaviour towards peers and often made inappropriate comments or asked inappropriate questions during interactions. This social disinhibition within interactions could be suggested to be a contributing factor in the peer relationship difficulties experienced in WS. Elison et al. (2010) further developed previous research by obtaining insight into the individuals’ social exclusion. Almost half of the adults with WS were reported by their parents to rarely or never attend extra-curricular clubs or activities. Furthermore, many of the adults who did engage in extra-curricular activities were reported to experience difficulties at these clubs and were dependent on their parents to arrange and facilitate these social activities. These findings highlight that peer relationships difficulties are prevalent in adults with WS. However, these studies used a single informant approach. Previous researchers found that peer relations and social functioning ratings varied across environmental contexts (Klein-Tasman et al., 2011; Udwin et al., 1987). This reinforces the importance of using a multi-informant approach to solicit the perceptions of the children with WS, their parents and teachers regarding the nature of the child's peer relationships.

Previous researchers have used an interview method to examine peer relationships in children with a range of neurodevelopmental disorders, including ADHD, ASD and DS (Bagwell et al., 2001; Cuckle & Wilson, 2002; Osborne et al., 2004; Rowley et al., 2012). In one research study, Cuckle and Wilson (2002) used a multi-informant approach by conducting semi-structured interviews with parents, learning support teachers and adolescents with DS to examine peer relationships in DS. Many of the adolescents with DS had a poor understanding of the term friendship. The adolescents with DS were reported to have few friends outside of school and struggled to sustain friendships, in particular with their TD peers. This was attributed in many cases to a gap in intellectual ability and a difference in interests. These interviews provided support for claims of peer relationship difficulties across contexts in adolescents with DS. No comparable qualitative studies have been conducted to examine peer relations in children with WS. This will be achieved in the present study.

7.2. Research aims

The aim of this study is to further expand our understanding of the characteristics of peer relationships in children with WS. The present study aims to build on the findings from chapter 6 by using a qualitative methodology to solicit the perceptions of children with WS, their parents and teachers regarding the nature of the child's peer relations. This will allow an examination into the characteristics of peer relations in WS across environmental contexts and across informants. Previous research findings suggest that peer relations are an area of difficulty for many children with WS (Greer et al., 1997; Udwin et al., 1997). There is a need to develop these findings by obtaining insight into the characteristics of these peer relationship difficulties, the children's social inclusion, extra-curricular engagement and the implications of atypicalities in social functioning for the children's peer relationships. Furthermore, although we know from previous research that children with WS tend to have peer relationship difficulties (Greer et al., 1997; Udwin et al., 1987; Laws & Bishop, 2004), this has rarely been examined in a developmental context. The present study aims to use interviews to solicit parents' observations on changes in the children's relationships over time and with development. The current study aims to advance qualitative research in this field by addressing the research question: 'What are the experiences of peer relationships in children with Williams syndrome?'

7.3. Method

7.3.1. Participants

21 children with WS participated in this study (7.50 - 16.49 years; M: 11.83, SD: 2.69; 12F, 9M). The parent(s) of all children were also recruited. The interviews were conducted with the mother (N = 11), father (N = 2) or the mother and father together (N = 8). 18 teachers also participated. All children had a formal diagnosis of WS which had been previously confirmed through positive genetic fluorescent *in situ*

hybridisation testing and were contacted through the Williams Syndrome Foundation UK. Full ethical approval was granted by the University ethics committee and informed consent was obtained from the parents and teachers who participated in this study.

7.3.2. Measures

Full details on the development of these interview schedules is provided in chapter 2.

7.3.2.1. Peer relations interview (child informant)

A 13 item interview schedule was developed for use with the children with WS. This interview schedule measured three constructs: the children's perceptions of the quality of their peer relationships, extra-curricular engagement and the children's understanding of the term 'friendship'.

7.3.2.2. Peer relations interview (parent informant)

A 14 item interview schedule was developed for use with the parents participating in this study. This interview schedule measured six factors: the nature of the child's peer relationships, the appropriateness of social behaviour, communication skills, inclusion by peers, frequency of contact with peers and engagement in extra-curricular clubs and activities.

These interview schedules were adaptions of the friendship interview schedules administered by Cuckle and Wilson (2002) in their assessment of friendships in adolescents with DS. Through reference to previous literature on social difficulties in WS, these interview schedule were modified to tap into specific aspects of the WS social phenotype.

7.3.2.3. Peer relations questionnaire (teacher report)

A 21 item questionnaire was developed for use with the teachers recruited. The items in this questionnaire measured five factors: the nature of the child's peer relationships, the appropriateness of their social behaviour, inclusion by peers, anxiety symptoms and level of support received. This questionnaire contained both Likert scale items (which were analysed in chapter 6) and open ended questions. For the purpose of the present study, only the open ended questions will be analysed.

7.3.3. Procedure

The children with WS and their parents individually took part in a semi-structured interview as described above. All interviews were conducted in the participants' homes and were audio recorded with the permission of the participant using a Dictaphone recording devise. The interviews ranged in duration from 6.46 to 48.36 minutes (M: 28.90, SD: 12.92). The teachers completed a questionnaire as described above. The questionnaire was posted to the school and the teachers completed this by providing written responses.

7.3.4. Data analysis

Parent and teacher interview and questionnaire data were analysed using the qualitative method of thematic analysis (Braun & Clark, 2006). An inductive approach was taken where the themes were driven by the data. In addition, thematic analysis was conducted within a realist paradigm, where the data were interpreted as a direct communication of the participants' thoughts, attitudes and motivations (Braun & Clark, 2006; Clark & Braun, 2013; Joffe, 2012).

In line with the guidance of Braun and Clark (2006), each stage of thematic analysis was followed in sequence. The audio data were initially transcribed verbatim. Initial codes were generated across the dataset and points of interest were noted. Through careful review and consideration, these initial codes were condensed into a final set of three themes.

7.4. Results

7.4.1. Interview findings (parents' and teachers' reports)

Through the process of thematic analysis, three broad themes were identified from the data: quality of peer relationships, atypical social interactions and extra-curricular engagement with peers (as shown in table 14). Each theme and the associated sub-themes will be discussed in turn, with reference where applicable to the prevalence of the theme. All themes and sub-themes identified from the data will be discussed and extracts from the interviews and questionnaires which highlight these themes will be provided. All names within the extracts will be replaced with pseudonyms in order to ensure the anonymity of the participants. Across the result section the gender and age of the child being discussed will be presented in parenthesis.

Table 14: Thematic analysis framework on parents' and teachers' perceptions of peer relations in children with WS

1. Quality of peer relationships

1.1. Absence of a best friend
1.2. Mainstream peers as a source of support
1.3. Contrasting peer relationships between the child with WS and his/her siblings
1.4. Perceptions of friendships
1.5. Difficulties sustaining friendships
1.6. Gap between the children with WS and their mainstream peers has widened
1.7. Changing relationships in the context of school transitions

2. Atypical social interactions

2.1. Lack of interest in peers
2.2. Intense behaviour towards peers
2.3. Atypical communication
2.4. Atypical patterns of play
2.4.1. Engagement in solitary play
2.4.2. Remains on the periphery from peers
2.4.3. A loss of interest during peer interactions
2.4.4. The need for adult involvement

3. Extra-curricular engagement with peers

3.1. Level of contact with peers
3.2. Engagement in extra-curricular clubs
3.2.1. Impairments limit participation at clubs
3.2.2. Need for additional support
3.2.3. Parental involvement

1. Quality of peer relationships

One prominent theme which was identified within the data was the quality of the child's peer relationships.

1.1. Absence of a best friend

Twelve of the parents interviewed reported that their child did not have a best friend that they regularly spent time with:

"I mean he has acquaintances, I wouldn't say they would be friends" (Male, age 11).

"He flits from people to people. He doesn't have anybody that he repeatedly talks about or anything like that" (Male, age 13).

"He gets invited to more birthday parties than other people's houses, which kind of again I suppose is that thing about him having lots and lots of superficial friends who you would say invite to a birthday party but not one who you would spend loads and loads of time with as a best friend" (Male, age 8).

The absence of a best friend was also observed within the school context. One teacher reported:

"He will be friendly but it is to everyone and not to anyone in particular. He doesn't engage in any social interaction with peers" (Male, age 11).

Although many of the children with WS did not have a best friend, several parents and teachers reported that the child was well-liked by their peers:

"Everybody seems to love her" (Female, age 7).

"He is a well-liked, accepted member of the school community" (Male, age 12).

"He is a well-liked child around the school. Not just with his class but with children from all years. All support him" (Male, age 11).

These extracts highlight a differentiation between acquaintances and dyadic friendships. While the children with WS are reported to be well-liked by their peers, they do not have the same quality of friendships as would be expected during typical development.

There was, however, diversity in the individuals' experiences of friendships and five parents reported their child to have a strong reciprocal friendship with a peer:

"She's got 2 friends in the street…they all play together and she's invited into both of their houses and they both know she's got Williams syndrome and they both understand about it. So she's welcome in both of their houses, they come here and she goes there" (Female, age 8).

"They've known each other since they were tiny so although he knows Sam is different and he knows Sam has Williams syndrome, he is just Sam to him so he doesn't see him as any different. He has got an amazingly lovely nature, really does look after him" (Male, age 9).

The individual differences within the parents' reports on the quality of their child's peer relationships have highlighted the heterogeneous nature of WS. This reinforces the value of utilising interviews to gain insight into the diverse experiences of individuals with WS.

1.2. Mainstream peers as a source of support

Four parents reported that their child had a close friendship with a peer; however, they emphasised that this friendship was of a different nature from the friendships engaged in by their mainstream peers. Parents explained that peers often took an older sibling or a carer role within the friendship and that they did not engage in activities which friends typically would:

“They grew up together and are both the same age…and she’s great with her…but she has now took on an older sister role” (Female, age 15).

“She has got a best friend…but it’s my friend’s daughter and they have been brought up together…they get on brilliant and are very close but it’s not a friendship where, they wouldn’t go out and do a girlie shopping day or anything like that. She looks after her…more of a carer than a friend” (Female, age 13).

Teachers also reported that peers were very supportive and protective towards the young person:

“Most children in the class would try to help her if she looks like she needs it or is distressed etc. but they see her as someone to look after, like someone much younger than themselves, rather than someone to make a friendship with” (Female, age 8).

“The other pupils in the class are aware of her needs and always include and support her” (Female, age 15)

“The children are very inclusive and mindful of her needs...the children in her own class and older children in the school feel protective over her. She has a strong group of peers in her own class for group work” (Female, age 8).

These extracts suggest that peers are attuned to the child's need for support. Both parents and teachers consistently reported that many of the mainstream peers showed a natural desire to protect the child with WS. These quotes highlight that the mainstream peers often modified their own behaviour to compensate for the child with WS's limitations. As a result, this posed evident implications for the child's friendships and ability to engage in age appropriate activities with their peers.

1.3. Contrasting peer relationships between the child with WS and his/her sibling

Several parents made comparisons between their child's friendships and the friendships of their TD siblings in order to further highlight the atypical nature of these social relationships:

"When you've got another child you know the difference, my other daughter comes home and she asks almost every night can such a person come on Thursday and can I go here and do that and that. It's different isn't it?" (Female, age 13).

"I would say no, not in the same way as perhaps her sister has relationships. She has had her favourites in the class, but again I wouldn't say they were necessarily friends in the way I would describe friendship and in the way her sister (aged 10) or perhaps her younger brothers (aged 5) would" (Female, age 8).

These extracts further reinforce differences in the quality of the children's friendships through a direct comparison to the friendship patterns of their TD siblings. These direct comparisons highlight the parents' awareness of their child's peer relationship difficulties and their need for different levels of support and monitoring.

1.4. Perception of friendships

Although many parents believed that their child had difficulties in establishing close friendships, they reported that their child perceived their friendships with peers to be strong:

"She's got lots of friends who she thinks are all her best friends. That can be from people she's met at primary school to someone she's met 5 minutes ago" (Female, aged 14).

"It's almost like everybody is his best friend" (Male, age 8).

"You are only in an hour and she has got 10 best friends" (Female, aged 14).

Both parents and teachers reported that these friendships were rarely reciprocated by the other child:

"Mainstream classmates would tolerate him more than befriend him" (Male, age 11).

"I find if he bumps into people quite often it is quite one way. He is very enthusiastic and they are much less" (Male, age 8).

"She would class them as more her best friends but I don't think it would be the other way around" (Female, age 8).

"Children are compassionate and friendly and will take time to play with her but it's not equal friendship" (Female, age 8).

These extracts suggest that the children with WS have a distorted perception of what constitutes a friendship. These quotes further highlight a differentiation between the child with WS and his or her peers on their perception of the shared relationship. While the children with WS were consistently reported to be enthusiastic over the relationship, this was rarely reciprocated to the same degree by their mainstream peers.

1.5. Difficulties sustaining friendships

Fourteen parents reported that their child experienced difficulties sustaining friendships with their peers, with these children reported to frequently alternate between peer groups:

"Yeah she has difficulty maintaining friendships over a period of time. Yeah she changes friends frequently" (Female, aged 14).

"He flits from people to people. He doesn't have anybody that he repeatedly talks about or anything like that" (Male, age 13).

"She just floats, she flits, I wouldn't say she feels she has this one friend and she has to stick with them at all times" (Female, age 15).

"He has friends, like specific people that he plays with all the time but there's a lot of them. So he goes from one person to the other…he just rotates around them all" (Male, age 12).

This difficulty in sustaining friendships was also present within the teachers' reports:

"This young person develops friendships but due to his immaturity relative to his peers, he finds these difficult to maintain" (Male, age 12).

One parent suggested that her son's difficulties in sustaining friendships were in part due to his peers developing in maturity:

"He hasn't changed so I think he could maintain the friendship but other children are getting older and they are growing out of the friendship. So I think if everybody stayed the same, they would be fine but they're not, so if he was each time going a year below, he probably would keep friends for longer but as I say the children are getting more grown up" (Male, age 9).

These quotes highlight the temporary nature of friendships where the children with WS were reported to alternate between their peers with no close bond to a specific peer. These extracts suggest that the child's friendships lack depth and longevity. These concerns were reported across parent and teacher reports. This highlights that the child's difficulties sustaining friendships with peers extend across environmental contexts.

1.6. Gap between the children with WS and their mainstream peers has widened

Many parents emphasised that the gap between their child with WS and the child's mainstream peers had widened significantly with age as the peers became more advanced in their interests and abilities:

"They are away far ahead now, I mean they are probably out socialising in places that they shouldn't be but that's what kids do but there's no way that she could be put in that situation" (Female, aged 14).

"As she got to the later years of primary school, the gap widened between her and others so she didn't get invited out as much and not really to people's houses. We would invite people here but they wouldn't often invite her back" (Female, age 11).

"They all seem a bit more advanced in the games that they play, she's not quite on their game level if you know what I mean, so she's a bit on the outside" (Female, age 8).

"When she was younger you didn't see it as much, the gap between her and her peers would be a lot smaller when she was younger. Now she is older, the gap between her and her peers is massive and it stands out, you can see it" (Female, age 14).

"I think she is just out of sync and they have all become quite independent and so getting public transport. I think when it got to the stage where the parents were no longer organising their social activities and even in the village, they can get quite a degree of independence at quite a young age as they are able to go to the swimming pool on their own or go to the park on their own but she just couldn't do that without adult support and so I think as that began to happen it was a struggle to maintain" (Female, age 15)

This gap between the children with WS and their mainstream peers was also recognised by teachers:

“The students in my class are very supportive of Jack but as they get older, the gap *socially and emotionally is growing” (Male, age 12).*

One parent expressed her concern that this gap would become more prominent when her son transitioned to secondary school:

“I think that gap is just going to get wider and wider and the worst thing is, he is *going to be more aware of it whereas now he is quite oblivious, whereas in a few years’ time he is really going to notice that he behaves and is treated differently from other children” (Male, age 9).*

Some parents also noted a difference in interests between their child and his or her peers which they believed to further contribute to the expansion of this gap:

“She’s not interested in make-up or listening to music, you know she wouldn’t do anything that a 13 year old would appropriately do” (Female, age 13).

“Other children her own age, she’s 15 nearly, they are all off going to town or the cinema or wherever they want to go with their friends, whereas she is not bothered” (Female, age 14).

These extracts highlight that peer relationships became more difficult to sustain as the child with WS had got older. This was attributed in many cases to their peers’ increased social independence, progressive maturity and the development of new interests with age. The peers’ increased independence was identified as a concern among several parents who worried about the potential implications of this increased freedom for their child’s vulnerability.

1.7. Changing relationships in the context of school transitions

Several parents and teachers reported an improvement in the quality of the child's peer relationships as a result of their transition from mainstream school to a SEN school:

"Jenny had difficulties when younger and in mainstream school but she is much more confident now as she is part of a small supportive community" (Female, age 15).

"The unit helps Samantha to develop friendships which would have been more difficult in mainstream school" (Female, age 13).

"He went to a mainstream middle school initially and I dropped him off at the school disco and you could just tell that the kids didn't know how to interact with him…he was very much an onlooker, very much watching what was going on rather than being a part of what was going on, it broke my heart. So now we have moved him to a special school, with kids with moderate learning difficulties and I dropped him off at youth club the first couple of times and he is just fully part of it… Yeah, he's found his little niche in the world... So yeah when it comes to mainstream children he really struggles and when he is around children who are kind of around his sort of understanding at the clubs he goes to, he absolutely loves it". (Male, age 13).

"She's settled in really well because primary school she found really difficult… it's been good for her to sort of go to school with other people that find learning difficult as well, not just her" (Female, age 11).

These extracts highlight that peer problems became more evident as the children reached the latter years of primary school and the gap between the child with WS and his or her mainstream peers widened. In consequence, many of the parents transitioned their child to a special needs secondary school thus alleviating the expanding gap between the children with WS and their peers. Across these quotes there is a common theme on the benefits of socialising with other children who share similarities in their social and intellectual functioning. This was reported to lead to perceptions of greater inclusion and stronger friendships.

However, there were individual differences in the responses and some parents reported that these peer relationship difficulties persisted, or in some cases were heightened when socialising with other children with disabilities:

"They don't mix together very well, although they want to be friends but they don't know how to be friends as both of them have got social and emotional immaturity...they don't know how to play without being led a bit, and if there's no-one else in the group with the maturity to lead them, they just clash" (Male, age 8).

"She just wants to be socially active whereas a lot of the other kids don't. It's a big thing for them to come to your house but they don't want you to come to their house, so you are kind of caught in that situation" (Female, aged 14).

"There's certain children that she will want to stay clear of, so children that are rowdy and throw things, she's frightened, any children that make loud noises or anything, she shies away from them" (Female, age 11).

These extracts suggest an inner conflict among parents when weighing up the advantages of mainstream versus special needs schools. While many parents acknowledge the benefits of special needs schools, they must also give consideration to the diverse range of disabilities regarding the children's support needs and characteristics. These parents highlighted the value of a level of compatibility within a friendship, with several of the children with WS facing challenges due to their own unique requirements.

2. Atypical social interactions

Another key theme which was identified from the data was the children's engagement in atypical social interactions with peers.

2.1. Lack of interest in peers

Many of the children with WS were reported to lack an interest in socialising with peers. Fifteen parents reported that their child frequently sought the company of older teenagers or adults over children their own age:

"He wouldn't go for the children his age, he would go for the teenage lads, we have seen him do it, he prefers people that are older…even the little guys his age were asking him to play football and he didn't want to, he wanted to go to these teenage lads hanging around on the swings" (Male, age 11).

"He is more sociable with adults, he is more comfortable with adult company… like if we were away on holiday, he wouldn't go and strike up a conversation with another child. He talks to the parent rather than the child" (Male, age 11).

“He does still prefer the company of adults…I have to tell him to go and play with his cousins rather than sit with my sisters and I” (Male, age 13).

“She will seek out an adult more readily than a child” (Female, age 13).

“If there’s adults here, she wants to speak to them, if there were children here she’s not bothered…If we had friends around and there were children in the other room playing, she would be sat in here with us” (Female, age 15).

Several teachers also observed this behaviour within school, suggesting this to persist across contexts:

“She is very keen to talk to adults and can be over-friendly” (Female, age 11).

“She definitely prefers communicating and spending time with adults” (Female, age 14)

“This individual prefers to talk to adults, rather than peers” (Female, age 14).

Some parents suggested that their child’s preference for adult company was due to adults providing their child with greater attention:

“She gets more of a reaction off adults, I think that’s what it is. A lot of adults will communicate with her, as she will just go randomly and say something to them and they will talk to her...so she gets more out of talking to adults because they are more receptive to her” (Female, age 8).

“I think they are just able to entertain her more. They have the skills to give her the attention that she wants really” (Female, age 15).

"Quite often at school he will spend time with the ladies cleaning up or in the kitchen. I think it is attention because adults would be more interested" (Male, age 11).

While one parent accounted for his child's preference for adult contact as due to his daughters need for security:

"The children are very kind to her but she doesn't want to be with them as they are too rough for her. She likes the gentle life and she would rather be with adults as she knows she is going to be looked after" (Female, age 11).

Across these extracts, a common theme was the children's motivation to seek out social interactions with adults. Several parents reported that while there were ample opportunities to engage with peers, the children with WS did not show an interest in pursuing these interactions. This unusually high level of interest in adults could be suggested to pose implications on the success of the children's peer relationships.

A few of the children with WS were also reported to seek out the company of younger children over their peers:

"I would say she associates with younger children, so for example if her brother brings a friend around, so they will be 10 or 11, she would mentally wise associate with them" (Female, age 15).

"In school she has occasionally been moved down and in fact she gets on better with year 1 and 2 children as their interests are on a par than perhaps the year 4 children" (Female, age 8).

"I've got friends who have got children his age and then younger children, he has more in common and he will play more with the younger children than he would do with the child that is his age" (Male, age 9).

He seems to prefer the company of younger pupils and activities aimed at their age and stage" (Male, age 12).

These extracts suggest that some children with WS may be better able to establish a connection with younger children due to the children's similarities in ability level and interests. These quotes indicate that some of the children with WS felt more comfortable and better able to relate to younger children than with their peers.

The extracts reported across this section suggest that the peer relationship difficulties experienced by many children with WS may not be due to a lack of opportunities to engage with peers, but rather, the children's lack of interest in manifesting friendships with their peers.

2.2. Intense behaviour towards peers

Nine parents reported that their child was overly affectionate and tactile towards their peers and often lacked an awareness of other people's personal space:

"Yeah he is quite tactile and he also doesn't really know where other people's personal space is so he gets quite close to people" (Male, age 8).

"Yeah because he would just wrap his arms around their neck and not let them go. You would have to go and pull him off and say 'leave them alone' " (Male, age 12).

"He wants to kiss people and hug them, all the time" (Male, age 9).

"He would still say to friends I love you, we are friends we love each other don't we, and other kids now would be like 'oh you don't say that type of thing'" (Male, age 9).

These over tactile and affectionate behaviours were also reported by several teachers:

"He is sometimes unaware of personal space" (Male, age 12).

"She is very affectionate and loves being hugged" (Female, age 15).

"This young person does often display over tactile actions e.g. hugging, arms around shoulders. Staff remind him of appropriate behaviour" (Male, age 12).

A few parents also reported that their child could become infatuated with certain peers:

"She gets a bit obsessional as in she'll maybe be on her messenger thing and I'm always checking it and have noticed that she has sent them a message like 15 times in a row, like every minute, 'Hi, Hi, Hi, Hi' waiting for them to reply" (Female, aged 14).

"*So this girl he was talking about, he gets fixations on certain people sometimes and he got a fixation with her and so he just liked watching her and following her around and of course I think a couple of years ago when he did it, she didn't mind but it's now she's a bit older and other people notice it…now we have a conversation every day that you can say hello to her but you don't follow her around…but unless you say that to him all the time he will forget and do it again" (Male, age 9).*

Several parents and teachers reported that this affectionate or tactile behaviour caused other children to feel uncomfortable, potentially impacting their peer relationships:

"The kids that are his age, sometimes you can see if they don't know him, they are not quite sure what to make of somebody being quite that forward" (Male, aged 12).

"It is sometimes a bit overwhelming for people who don't know her and so they tend to kind of stand back…you can see them looking as if, she doesn't know me but she's shouting hello" (Female, age 14).

"You are only in an hour and she has got 10 best friends but again it's quite draining, especially for kids as they are quite over powered by her I think…I think she is too full on sometimes that it kind of takes people back. There's no inhibitions" (Female, aged 14).

"When I used to take her to primary school and she would run over to people and fling her arms over them and you could kind of see them stood uncomfortably you know, with their arms by their side thinking let me go, I don't like this" (Female, age 11).

"She is over bearing and intense which causes discomfort to peers" (Female, age 13).

While many parents reported this intense behaviour to be a negative factor in their child's peer relationships, one parent emphasized the difference between the reaction of familiar and unfamiliar peers to her daughter's tactile behaviour:

"You can see other children who don't know her that well totally don't know how to take it and are totally standoffish and a bit like get off me…It's only the kids who really know her and have known her a long time. They see it as part of her personality" (Female, age 8).

These extracts raise several important points. Within chapter 6, parental questionnaire ratings indicated that several of the children with WS engaged in disinhibited tactile behaviour towards their peers and lacked an awareness of personal space boundaries. The interview data in the present chapter developed this further by shedding light on parents' perceptions of the implications of this tactile behaviour for the child's peer relationships. Numerous parents indicated their concerns regarding their child's overly tactile behaviour, believing this to have negative implications on the success of the child's peer relationships. Both parents and teachers indicated their need to monitor the child's behaviour and to regularly remind him or her about appropriate conducts of behaviour. Parents indicated their struggles to moderate their child's behaviour through repeated reminder and education regarding appropriate behaviour; however, noted that despite these efforts the children were often unable to control their actions. This could be suggested to put a strain on the parent-child relationship. Several parents and teachers also raised concerns regarding the peers' visible discomfort regarding the child's overfriendly manner. In particular, older peers were reported to show a greater awareness that the child with WS's behaviours were different, with this believed to have implications on the nature of friendships.

2.3. Atypical communication

Several of the children were also reported to experience difficulties in their communication with peers. This aligns with the findings reported in chapter 6, where over 70% of the children with WS were rated within the SRS as showing a moderate or severe level of impairments in social communication. Across the interview extracts, many of the children with WS were reported to be repetitive, fixate on certain subjects and show a lack of depth in their conversations. These difficulties were reported by both parents and teachers which supported these communicative impairments to be present across contexts:

"*She tends to repeat herself quite a lot or say something inappropriate or daft or nothing to do with the conversation and then that kind of stops the conversation" (Female, aged 14).*

"It's the same things like what's your name, where do you stay, it's as if she has been programmed to ask that. She wouldn't maybe ask someone where they had been on holiday, she's not really that great a conversationalist" (Female, aged 14).

"I'd say it's superficial" (Female, age 8).

"She likes to talk about herself and what she likes to do and if you want her to talk about something completely different, you can't, she will carry on talking about what she was talking about" (Female, age 13).

"She may say something in conversation that is relevant but doesn't respond to what others say directly. Most conversations between her peers

are beyond her level of understanding so largely inaccessible" (Female, age 8).

These extracts highlight that many of the children with WS showed a limited capacity to engage in reciprocal conversation due to their communicative impairments. This poses potentially substantial implications on the children's ability to establish friendships with peers.

However, there were individual differences within the interviews. A few parents reported that their child had strong communication skills and was able to engage in reciprocal interactions:

"She sits down with us and we talk about the day and so she can have very sensible conversations definitely. I mean she is getting older and she is quite interested in things and things that she has seen on the news even. She does take in quite a bit and asks a lot of questions and is quite curious about things" (Female, age 15).

"We did a lot of work with him when he was about 4 or 5 on turn taking and things like that through speech and language therapy so when he has a conversation he has a conversation… He has got his stock questions and answers but he listens to what people say" (Male, age 9).

These quotes further support claims of social communicative impairments in children with WS as was reported earlier in the thesis. These communicative impairments were perceived by some parents to pose implications for the children's social interactions with peers, hence could be argued to hinder the children's ability to develop and sustain relationships with their peers.

2.4. Atypical patterns of play

Another theme which was identified from the data was the children's engagement in atypical patterns of play.

2.4.1. Engagement in solitary play

Many parents reported their child to primarily engage in solitary activities and to prefer to play alone. An engagement in musical activities (N=16) and the use of technology, particularly searching YouTube (N=15) were the most common pastimes among this sample of young people.

"It doesn't keep her interest does it, like having a friendship, she would rather be on her own. She would rather be on her own at home too" (Female, age 13).

"Her favourite past time is to sit on her own in her bedroom, if we let her, on her iPad watching One Direction or on the Wii" (Female, age 13).

"She's quite happy on her iPad, playing on it, she would be on it all day long if you would let her. You know she is quite happy with her own company really" (Female, age 15).

"We all get together, my group of friends and they have all got children. She will quite often go off, doing her own thing, not playing with them. It's not that they are leaving her out but she will just choose to do that. Or if my other daughter has got her friends round, Chloe won't interact and play with them, she would rather be on her own" (Female, age 13).

The children were also reported to show a preference for solitary activities within school:

"He likes time on his own at break times and he will often go and find a quiet space. He will watch others but not speak to them or join in" (Male, age 11).

"He generally plays on his own" (Male, age 13).

"He sometimes just floats around the playground...he chooses to simply hang around alone" (Male, age 11).

These extracts emphasise the child's choice and preference to play alone, despite there being opportunities available to interact with peers. Although the concept 'solitary' may usually have negative connotations of exclusion, these quotes indicate that the children found comfort in these solitary activities and frequently selected solitary play over interaction with their peers. These extracts also highlight the parents' strive to reach a balance between solitary play and social interaction. The use of technology was highlighted as a core interest among the children with WS, with many of the children's pastime revolving around this.

2.4.2. Remains on the periphery from peers

Parents also reported that their child tended to play alongside their peers, rather than interacting directly with peers in a game:

"She would more be playing alongside them. That's what you get, generally. If you try to get her to play with anybody, they are playing and she is playing alongside them but she isn't actually playing with them" (Female, age 14).

"He would be more on the periphery. Things like if they are playing a computer game, he is happier just to watch than to take part" (Male, age 11).

"He didn't play with any of them, he was just playing alone and is happy to do so… didn't play with a single person there or talk to them or anything really but he had a great party" (Male, age 11).

These extracts suggest that many of the children with WS have difficulties within their play skills, with this posing implications on their ability to engage with peers and establish friendships. These quotes align with the parents' ratings in the VABS as reported in chapter 6, where the children with WS scored significantly poorer in the 'play and leisure time' sub-scale of the VABS than TD children of comparable chronological age. The interview data develops these questionnaire ratings by soliciting parents' observations of the implications of these play deficits for the children's interactions with peers. Despite these challenges, several parents noted that their child was more content to be an onlooker in social situations. These extracts highlight the complexity of WS, where the children were happy to be in the vicinity of other people; however, due to the children's impairments they struggled to engage appropriately with these peers.

2.4.3. A loss of interest during peer interactions

Many parents reported that their child often lost interest during social interactions with peers. Parents described occasions where peers from school came to visit and their child had lost interest in playing with the visitors and retreated back to his or her own solitary activities:

"They will turn up, have some juice and something to eat and then she will just take herself upstairs… her brother generally ends up entertaining whoever's came round" (Female, age 15).

"She can lose interest really when someone is here and so her music is her big thing and she spends most of her time plugged in to her music and she can lose interest in someone and want to go and do that. When her friend came for a sleepover, we organised a movie, but I had all that lined up for them to do…she could still abandon a friend really, have someone round, be really delightful that they are there and then abandon them and gravitate back to her solitary interests really" (Female, age 15).

"Even with her friends here, they will come here and play shops and such like and she will play for a couple of minutes and then she will probably go off herself and play on the computer" (Female, age 8).

"She will start off playing and will join in the game, but she gets either bored very quickly or she loses track of the game so she will just go off" (Female, age 7).

One teacher also reported:

"Peers are respectful and do try to include her but she will often turn her back, rather than get involved" (Female, age 14).

These extracts highlight the children's difficulties sustaining an appropriate level of interest within peer interactions. Despite their initial enthusiasm at having a visitor, many of the children with WS lost focus and retreated back to their solitary interests. This could be a result of the child's EF impairments within attention which may hinder their ability to sustain interest during peer interactions.

2.4.4. The need for adult involvement

Several parents reported their involvement in many aspects of their child's daily social life. Parents described their child's lack of independence in arranging social activities, with most social contact being initiated by the parents. A few parents also reported that they had to encourage their child to engage with peers and provide direction in their social interactions:

"It's more so if I'm friendly with the mum and then it's maybe going out for lunch or it's going out to the cinema and then we go for something to eat after it, not very much maintaining on their own" (Female, age 14).

"I just maybe say to my friend who has the little boy with Downs syndrome, does he want to come to tea and one night she will go there but it's not that they have asked each other. They are quite happy for it but we have instigated it" (Female, age 13).

"She wouldn't come home and go can I go to Jessica's house for tea or that kind of thing, she doesn't understand how to ask for things like that really" (16, Female, age 8).

"If we usher him off to play with kids, he is prepared to do it, he just needs encouragement to do it" (Male, age 13).

Several teachers also reported that the children required adult encouragement and support in order for them to engage in social interactions with their peers:

"Peers try to include her and staff encourage integrations – even so she requires support and prompting to join in" (Female, age 14).

"She needs to be encouraged to participate, although if she is comfortable in situations she will join in, particularly anything musical" (Female, age 11).

These extracts all illustrate the crucial role of the parents in the children's social interactions. Although the children with WS were reported to be happy to engage in these social activities, there was a dependency on the parents to establish social contacts and arrange and facilitate all social activities. The children's reliance on adult support was evident within the home and the school environment, suggesting that these social difficulties persist across social settings.

3. Extra-curricular engagement with peers

Another important theme was the children's extra-curricular engagement with their peers.

3.1. Level of contact with peers

Three individuals were reported to socialise frequently with peers outside of school. The remaining children had infrequent contact or no contact with their peers outside of school:

"He doesn't socialise out of school, so he never has anybody back for tea, he is never invited to other people's for tea, it just doesn't happen" (Male, age 13).

"No-one would ever come and knock on the door and ask if he is playing out" (Male, age 11).

"We have been looking at trying to find some sort of club for her as there's nothing for her to do. She's lucky she has got siblings but apart from that, it's quite a lonely life for her sometimes" (Female, age 11).

These extracts highlight the challenges faced by parents in their extensive efforts to improve their child's social relationships. Some parents expressed their disappointment for their child and concerns regarding their child's exclusion from peer groups. Parents indicated their efforts to increase their child's social inclusion by seeking out appropriate clubs and the challenges associated with this. These quotes highlight the exasperation experienced by some parents in their strive to gain equal opportunities for their child and identify an area where further support is required.

Some parents also noted their child's exclusion from birthday parties:

"Yes and it's most noticeable when invites have been handed out in the class and it can be very difficult to explain to her then that no you haven't got an invite. I think that she can find upsetting and the fact she's not been included in things that she perhaps wants to be. Other times I think it's less obvious as she doesn't necessarily see the other children going home and having playdates after school. For parties it's very obvious, so that's why she has come out and she has been upset when I've had to break it to her that no you haven't had an invite" (Female, age 8).

"She gets left out of parties and stuff like that, she doesn't very often get invited to stuff like that" (Female, age 15).

These extracts highlight the emotional turmoil experienced by parents on witnessing their child's exclusion by peers and managing the fallout from this. This exclusion was reported to be particularly apparent when party invites were handed out within the class, with evident implications for the child's emotional distress.

One mother also reported that she could not allow her son to visit a peer's home alone due to his social impairments and need for supervision:

"I wouldn't let him go to someone else's house as he doesn't understand, he wouldn't know there's certain rooms you don't go in or he wouldn't know if somebody was being inappropriate with him" (Male, age 9).

There were individual differences in the participant sample however with regards to the child's socialisation with peers, with some children reported to have regular contact with their peers out with school:

"Every fortnight at least, they are at one or another" (Male, age 9).

"She's got 2 friends in the street, Jack and Ben, they all play together and she's invited into both of their houses and they both know she's got Williams syndrome and they both understand about it. So she's welcome in both of their houses, they come here and she goes there" (Female, age 8).

These quotes further highlight the heterogeneous experiences within the social relations of children with WS.

3.2. Extra-curricular clubs

While many of the children with WS attended extra-curricular clubs with their peers, which included swimming, dancing and youth groups, parents often reported that their child experienced difficulties at these clubs. This was particularly evident when attending mainstream clubs.

3.2.1. Impairments limit participation at clubs

Many of the parents noted their child’s difficulties when participating in sports and other activities and their struggles to keep up with mainstream peers at these clubs:

“He just doesn’t keep up. He can’t keep up with dance routines and taekwondo, he just couldn’t” (Male, age 13).

“She went to kayaks for a bit, and brownies for a bit when she was young but didn’t keep up with that and then there was dancing as well, didn’t keep up with that” (Female, age 15).

“She doesn’t necessarily do everything at the same time she’s meant to. I think she can manage the steps but not necessarily when they are meant to be. She can’t follow the instructions as quickly” (Female, age 8).

These extracts indicate that several of the children faced obstacles within mainstream clubs due to their motor impairments. A general theme from the interviews was the need for those around the child, for example adults and peers, to understand the child’s needs and if possible moderate the activities to facilitate the child’s participation.

3.2.2. Need for additional support

Some parents noted careful consideration to select clubs which are suitable for their children’s needs, with some children attending clubs with a younger age group or smaller group size due to their requirement for additional support:

"They are mainstream clubs but it's a younger group, the actual dance class she is in is aged 2 to 6, so they are all a lot younger because if she was in a group her own age, they would all be much more advanced in abilities" (Female, age 8).

"It is mainstream but it is a much smaller pool. So it is a 10 metre pool and the staff are in the water and it's only groups of 8 with 2 teachers…there's a greater level of support there. It's warm as well, as she gets very cold, so that's another thing. Her brothers (aged 5) swim at the local leisure centre in the 25 metre pool and her sister did, and that's 1 to 12 in terms of the ratios, whereas she wouldn't cope with that" (Female, age 8).

"We are a bit more selective about which groups we send him into and for how long. So Beavers is only an hour long and he is usually fine at that, it's literally just up the road" (Male, age 8).

These extracts further highlight the careful thought process of the parents regarding the suitability and safety of clubs for their child. This highlights the value of special needs clubs which are suggested to be better equipped to cater for the individual needs of children with disabilities.

3.2.3. Parental involvement

Attendance at the clubs was predominantly initiated by parents and the children often required encouragement to attend:

"It's us that have kind of initiated that…she would be quite happy to go home and sit on her own every night in her bedroom, quite happy and it's us that say 'no you're

doing this'. So 3 nights a week she goes out and spends this time with children her own age because it's important" (Female, age 13).

"We are trying to encourage her with this club now so she can interact with people her own level, so she gets more interactive" (Female, age 14).

One parent also emphasised a difference in the level of encouragement and initiation required for her child to attend a club in comparison to her TD sibling's participation in these activities:

"It's all been driven and instigated by us. Sometimes she will say I don't want to go to dancing, I want to stay in, but it's making her go and be out there. Whereas my other daughter wants to do certain clubs because she wants to do it" (Female, age 13).

However there were individual differences within the reports, with some children reported to initiate their own involvement in extra-curricular activities:

"Everything that he has ever done, it's him, all sorts of activities at school. He has done every instrument and he has tried every club at school, the football club, running club. So we let him do whatever he wants at school and when he wants to stop, he stops. So what he is doing at the moment are the things that he wants to do… Yeah he is definitely kept busy and he is doing what he wants to do really" (Male, age 11).

These extracts highlight the parents' views on the value of their child integrating socially with other children. Several parents noted that they were the driving force behind the child's attendance at these clubs. This was found to have implications for the parent-child relationship, where some children showed resistance over their parent’s decisions regarding extra-curricular engagement.

7.4.2. Interview findings (children’s reports)

When conducting the interviews with the children with WS, it was evident that many of the children had very limited verbal skills. Many of the children with WS provided short responses across the interviews which limited the level of insight which could be obtained into the children’s perceptions of their peer relationships. This in line with previous research where children with WS were reported to struggle to engage in reciprocal conversations (Klein-Tasman et al., 2011; Laws & Bishop, 2004) and the findings of substantial communication deficits as reported in chapter 6 of the current thesis. Furthermore, across the interviews, many of the children with WS showed difficulties maintaining focus on the present subject in line with substantial evidence of attention deficits in WS (Leyfer et al., 2006) and often diverted the topic of conversation to their own niche interests or obsessions. The children’s verbal communication skills are therefore suggested to limit opportunities for qualitative analysis in the present chapter. The responses across the children’s interview data will be reported as frequency statistics.

All 20 children reported that they had at least one best friend and were able to name this friend. All of the children were able to describe activities which they took part in with their friends. This included sports, technology based games and general play. All children reported that they spent break and lunch time at school with their friends. When asked how often they argued with their friends, 13 children reported that they never argued with their friends and the remaining 7 children reported that they only occasionally argued with their friends or only with one specific peer. A few of these children reported that this made them feel angry, sad or frustrated. 16 of the children with WS reported that they had never been left out by their peers. Among the children who reported to being left out by peers, 3 children reported that this only happen occasionally or referred to one specific child. However, one boy reported that he was left out by peers “because they aren’t kind”. Many of the children showed difficulties defining the concept friendship. 7 children defined friendship around engaging in activities together, 10 children listed positive character traits e.g. kind, loving and helpful, however, many of the children were unable to expand on this. 3 children were unable to provide a definition of friendship.

7.5. Discussion

Qualitative insights were obtained from the children with WS, their parents and teachers through the use of semi-structured interviews and open ended questionnaires. This expanded the understanding of peer relations in children with WS, building on previous research which primarily used rating scale measures (Greer et al., 1997; Laws & Bishop, 2004). Across the responses, several themes were identified regarding the children’s peer relationships, social interactions and inclusion.

There were evident difficulties within peer relations, with substantial implications for the children with WS and their families. Most of the children did not have a best friend and struggled to sustain friendships. Some parents accounted for these peer relationship difficulties through their child’s atypical interactions with peers. Several parents and teachers reported that the child with WS lacked a sustained interest in their peers and showed social interaction deficits. As a result, the children frequently partook in solitary activities. Many parents also reported that their child had low levels of social contact with peers out with the school context and was often excluded from birthday parties and other social events. Although the majority of the parents reported that their child attended extra-curricular clubs, most parents indicated that their child experienced difficulties at these clubs due to their social and motor impairments. Previous research examined rates of attendance at extra-curricular clubs in adults with WS (Elison et al., 2010). The current findings develop this research by providing further insight into the difficulties encountered within these clubs and the implications for the child's inclusion.

However, there were individual differences in the parents' responses. A small number of parents reported that their child had a strong reciprocal friendship with one or more peers. These parents noted that their child was able to sustain friendships over a prolonged period of time and was well integrated in their peer group. These reports build on previous findings on the heterogeneous nature of cognition and behaviour in WS (Little et al., 2013; Porter & Coltheart, 2005) and indicate that individual differences are also evident in the social skills of children with WS.

Several parents and teachers emphasised the difference in the nature of the child's peer relationships in comparison to the friendships of their TD peers. Both parents and teachers reported that peers often mothered the child with WS or adopted a carer or sibling role within the friendship. This highlights the peers' awareness of the young person's vulnerability and their need for additional support. It was evident from the interviews that the individuals surrounding the child with WS, for example parents, teachers and peers, recognise his or her difficulties, and in many cases, attempt to accommodate the child by intervening to organise or moderate social activities or instructing the child in appropriate patterns of behaviour. Several parents also directly compared their child's friendships with the friendship patterns of their TD siblings in order to further emphasise the atypical nature of these peer relationships. These parents have a clear reference point and prior experience of typical patterns of peer relations during childhood. In light of this enhanced understanding, these parents could be suggested to be better able to draw stronger conclusion regarding their child’s peer relationship difficulties.

Several parents reported that despite the challenges associated with friendships, their child believed that they had numerous close friends. As indicated in the interviews with the children with WS, all of the children reported that they had a close friend and were able to name this peer. This was in contrast to the perceptions of many of the parents and teachers who indicated that the child with WS did not have a close reciprocal friendship and struggled to sustain relationships with peers. The evident disparity between the perspectives of the children with WS and their parents and teachers suggests that children with WS have a poor perception of their own peer relationships. This poor social awareness was further highlighted through the children’s difficulties defining the concept of a friend, with some parents also noting that their child considered someone to be a friend instantly upon meeting them. These findings further reinforce the complexity of peer relationships for children with WS. The current findings are in line with previous research which assessed SAB in WS and reported there to be a poor correlation between the young persons’ reports on their own approach behaviour and real-life observations of their behaviour (Fisher et al., 2014). In light of the current findings of poor self-awareness and the children’s difficulties understanding the concept of friendship, these findings suggest that children with WS may interpret other people’s intentions and natures favourably. They may therefore be less aware of other people potentially taking advantage of them.

Several parents and teachers reported that the child with WS was over-friendly and inappropriately tactile towards their peers, with some peers believed to be overwhelmed by this behaviour. Previous research identified an engagement in high levels of tactile behaviour in adults with WS (Davies et al., 1998; Elison et al., 2010; Udwin, 1990). This was further illustrated in chapter 6 of the present thesis, where over three quarters of parents reported that their child with WS frequently engaged in over-tactile behaviour during peer interactions. The present study builds on these findings by soliciting parents' and teachers' observations on the implications of this tactile behaviour for the peer relationships of children with WS across social settings. Several parents and teachers believed that the child's disinhibited behaviour towards peers was problematic, causing the peer to feel uncomfortable. This was identified as a barrier in the children's ability to sustain a close friendship with their peers. The consistency between parent and teacher reports suggests that this tactile behaviour is prevalent in WS and is manifested across contexts. Some teachers also reported that the child had to be regularly reminded by staff about socially appropriate behaviour. This highlights that this behaviour is a recognised problem within school.

This engagement in inappropriate levels of tactile behaviour could be explained through the theoretical assumptions of the frontal lobe hypothesis. The frontal lobe hypothesis proposes that the high levels of SAB demonstrated by individuals with WS are caused by impairments in response inhibition, resulting from abnormalities in the frontal lobes (Little et al., 2013). These principles could therefore be extended to account for the inappropriate tactile displays which have been reported by several of the parents and teachers in this current study.

Children with WS are socially driven to approach other people and seek out social interaction (Dodd et al., 2010). Despite this hyper-social profile, many of the parents reported that their child experienced significant difficulties in sustaining friendships with their peers, in line with the findings in chapter 6. Children with WS have been reported to show deficits in the identification of facial expressions (Karmiloff-Smith, Klima, Bellugi, Grant, & Baron-Cohen, 1995) and inferring other people's mental states (Hanley et al., 2013) when compared to TD children matched on chronological age. Selman (1980) proposed that social-cognitive functions play a critical role in the development of friendships. This was reported in chapter 6 of the present thesis, where there was a significant negative association between impairments in social cognition, as measured in the SRS, and the quality of peer relationships in the children with WS. These findings suggest that the children's impairments in social-cognition contribute to their difficulties sustaining friendships.

Several parents and teachers noted that the gap between the children with WS and their mainstream peers had expanded significantly with age. Teachers accounted for this increased gap as due to the peers advancing in their social and emotional maturity relative to the children with WS. Numerous parents reported that their child did not have the ability to arrange social contact with their peers independently, further contributing to the expansion of this gap. The need for parental involvement was present across age groups, with several adolescents also reported to rely on this support. It would be interesting in future research to examine whether this need for parental support declines in adulthood or is a requirement for many individuals with WS across their lifespan. Teachers also reported that any interactions with peers required encouragement from staff members. One teacher also observed that when her pupil felt comfortable in the situation, such as in musical activities, she was more likely to engage with peers. This is in line with previous research which reported children with WS to experience anxiety in novel situations and environments (Dykens, 2003) and suggests that familiarity may be an important factor in successful peer interactions.

A few of the parents stated that initiating social activities for their child was simpler when the child was younger. Some parents noted the challenges they encountered in their quest to initiate social interaction between their child and his or her peers. Parents reported that they were no longer in contact with other families as a natural consequence of the peers' increased independence from their parents. Many of the parents interviewed also highlighted the difference in interests between their child and their peers with age, which they believed to further hinder their peer relationships. Many of these parents reported that their child had unusual interests or obsessions which were not age appropriate. This reflects past research findings of repetitive and restricted interests in WS (Rodgers et al., 2012). Friendships develop through shared interests (Gifford-Smith & Brownell, 2003). This difference in interests could therefore be suggested to account for the peer relationship difficulties experienced by this group. The current findings build substantially on previous research by investigating peer relations in children with WS in a developmental context. The accumulated findings suggested that peer relationships became more problematic with development, with this primarily attributed to the expansion of the gap between the child with WS and their mainstream peers.

Several parents and teachers reported an improvement in the quality of the child's peer relationships following their transition from mainstream to a SEN school. These findings are in line with the results reported in chapter 6, where parents and teachers reported lower levels of social exclusion and a higher number of friendships among the children who attended SEN school compared to those who attended mainstream school. By using an interview method, the author was able to further explore these findings. SEN schools are specifically catered for children with additional support needs. Several parents reported that these schools provided their child with greater opportunities for social inclusion and facilitated the development of stronger peer relationships than was possible in mainstream school. This was suggested to alleviate the difficulties previously discussed regarding the expanding gap between the children with WS and their mainstream peers and the associated peer relationship problems. Both parents and teachers noted the greater levels of support provided at SEN schools. These schools were believed to provide a supportive environment which allowed the children to form friendships with other children of a similar intellectual ability. One parent noted the direct benefit of this transition in connection with her son's social inclusion. She recalled a particular social event at a previous mainstream school where she observed her son struggle to integrate with his classmates and recounted her anguish when observing her son's isolation. This parent emphasised the stark improvement in her son's inclusion by peers and his eagerness to interact when attending clubs with other children with disabilities through the SEN school. This suggests that children with WS may feel more comfortable when attending clubs which are specifically catered for children with additional support needs and feel more able to participate in these surroundings.

However, there were individual differences with regards to the benefits of SEN schooling for the child's peer relationships. Some parents reported that these peer relationship difficulties persisted following their child's transition to a SEN school, and in some cases these difficulties became heightened. A few parents accounted for these peer relationship difficulties as due to a difference in the nature of WS and other complex disabilities, with many children with WS seeking out social situations in contrast to some of their peers with ASD who were reported to be less socially active. This difference in characteristics and interests was reported by some parents to hinder their child's development of friendships at school.

The majority of mainstream peers were reported to be very accommodating and supportive of the children with WS. It was suggested that these peers were able to recognise the young person's vulnerability and had the skills to compensate for this within their social interactions. In contrast, some parents suggested that other children with additional support needs may not have the social skills or level of understanding required to engage in an effective friendship. Some parents also reported their child's anxiety to be another barrier in their development of friendships with other children with additional support needs due to the occasional unpredictable nature which may occur within these disabilities. Children with WS have been reported to experience anxiety in novel situations and to have over-sensitive hearing and heightened anxiety for loud noises (Dykens, 2003). This anxiety could be suggested to further contribute to the peer relationship difficulties experienced by children with WS.

Another prominent theme was the children's engagement in atypical social interactions with their peers. Almost three quarters of the children were reported to lack an interest in peers, instead seeking out contact with older children and adults. Previous research reported a strong motivation in children with WS to seek out contact with older children and adolescents (Dodd et al., 2010; Doyle et al., 2004). These findings were further highlighted in chapter 3 of the present thesis, where many of the children with WS were reported by their parents to show a greater level of interest in approaching and conversing with unfamiliar adults than unfamiliar children. The present research further developed these findings by soliciting parents' perspectives on the factors underlying this behaviour. Parents accounted for this preference for adult contact as due to older individuals providing their child with a greater level of attention, interest or security which they seek. Some parents also reported that their child was often invited to play by peers but declined these invitations in order to seek out adult company. It could be suggested that the children's difficulties in peer relationships may be due to their attention being fixated on adults in any social situations to the detriment of their interest and interactions with peers.

Several parents and teachers reported that the children with WS sought out adult contact. This consistency between the parents and teachers highlights that this unusually high level of interest in interacting with adults is a core feature of the WS social phenotype and is present across contexts. This also suggests that this behaviour is not due to a lack of opportunities to engage with peers, as this behaviour persisted when in a school setting where there are numerous opportunities for peer interaction.

Some parents reported that their child sought out the company of younger children over their peers, with these children reported to relate more with the younger year groups at school than with their classmates. This could be explained by a similarity in intellectual ability, hence the gap in maturity and interests between these groups is minimised, providing greater grounds for establishing friendships.

Many of the children with WS were reported to primarily engage in solitary activities, with a particular interest in musical activities (Rosner et al., 2004). This interest in solitary activities is in line with previous research (Davies et al., 1998; Rosner et al., 2004; Udwin, 1990). However, the present findings develop this research further by soliciting parents' and teachers' observations of the children's solitary play across social contexts. There was stability in the children's engagement in solitary play across the home and school environment. During the pre-school period, TD children are argued to progress through several phases of play. Pre-school children initially show a period of solitary play, where their attention is primarily focused on their own activity, with little interest in their peers. As children transition though the pre-school period, they enter a phase of parallel play. During this stage, the children more frequently play alongside their peers; however, there is minimal interaction between the children. At the end of the pre-school period, children are proposed to begin to engage in collaborative play with their peers, where the play is reciprocal and involves frequent social exchanges and the children working together to achieve a common goal (Parten, 1932). Both the children and adolescents with WS in the present study were reported by parents and teachers to engage in solitary play. This suggests that unlike TD children who transition past the solitary phase with development, this pattern of solitary play may persist across childhood in WS.

Several parents reported that their child often lost interest during peer interactions and reverted back to their solitary activities. This loss of interest in peer interactions could be explained by attention deficits in WS (Leyfer et al., 2006). Several parents noted the need to have pre-arranged structured activities in place for their child to engage in with peers in order to sustain their attention for longer. This highlights the children's need for routine and structure (Riby et al., 2013). This can be argued to provide support for the theoretical assumptions of the frontal lobe hypothesis, where the children's difficulties in peer relationships can be accounted for by neural impairments to the frontal lobes, resulting in EF deficits.

Teachers also reported that the children with WS frequently engaged in solitary activities within school, which indicates that this behaviour persists across contexts. Schools provide an optimal environment for group play with peers under the encouragement of staff members, therefore the children with WS were reported to engage in these solitary activities despite numerous opportunities for peer interaction. Parents and teachers reported that the children frequently chose to play alone, despite their peers' best efforts to incorporate them in any games or activities. This may suggest that these children enjoy their own company while playing independently. Alternatively the decision taken by the child to play alone could have a direct link to their own anxieties surrounding social interactions.

Many parents also reported that their child engaged in unusual patterns of play. These findings further develop the results reported in chapter 6, where the children with WS showed significantly greater impairments in their play skills when compared to the normative population. The use of an interview method in the current chapter allowed further exploration of this. Several parents reported that their child tended to remain on the periphery, often as an onlooker, rather than engaging directly in play with peers.

Children with WS have been reported to demonstrate pragmatic language impairments and difficulties in reciprocal social interactions (Klein-Tasman et al., 2011; Laws & Bishop, 2004), with this further illustrated in chapter 6 of the present thesis. However, no research thus far has examined the implications of communicative impairments for the children's peer relationships. The present study builds on previous findings by soliciting parents' and teachers' observations on the impact of communication deficits for the children's peer interactions.

Several parents and teachers noted that the children with WS showed atypicalities in their communication with peers. Conversations were often reported to be repetitive in nature. The children were reported to struggle to achieve any level of depth within a conversation. Some parents noted that their child had stock questions which they regularly drew on during social interactions; however, the children were reported to struggle if the conversation deviated from within their control. This supports previous findings where children with WS on the surface appeared to be highly sociable; however, struggled to further develop these conversation (Laws & Bishop, 2004; Stojanovik et al., 2001). The present thesis builds on this research by obtaining insights into parents' accounts for their child's struggles within communication with peers. Some parents accounted for these communication difficulties as primarily due to the gap in intellectual ability between their child and their mainstream peers which resulted in a loss of focus. Parents reported that their child often attempted to engage in conversation with their peers; however, due to their inability to maintain control of the topic, their input was sometimes irrelevant and at times resulted in the termination of the conversation. This highlights the children's willingness to engage in conversation. This need to maintain control within conversations could reflect the difference in interests between the child with WS and their mainstream peers, with some children reported to fixate their conversations around their obsessions and specific interests and to lose focus when the conversation drifted from them.

Language is reported within Hay et al. (2004) framework as a critical factor in the development of successful peer relationships during infancy and later childhood. These communication skills are argued to enable children to share their thoughts and feelings and avoid situations of conflict, therefore strengthening their peer relationships. These communication difficulties, along with their impairments in social cognition, could be suggested to account for the atypical social interactions reported in this sample, making play with peers significantly more challenging.

Many parents reported their child to have little or no contact with their peers out with school. This provides support for previous findings of high levels of social isolation in children and adults with WS (Davies et al., 1998; Greer et al., 1997). The current study builds on previous research by obtaining parents' accounts of real-life scenarios which illustrate the children's level of social inclusion. Several parents also indicated their child to be excluded from parties, with both young children and adolescents reported to experience social exclusion, highlighting that this is a problem across the developmental spectrum. Variability was again evident, with a few children reported to have frequent contact with their peers, reflecting the heterogeneous nature of this disorder.

Within chapter 6, over 60% of the children with WS were reported by their parents to attend extra-curricular clubs with their peers. This current chapter develops these findings by exploring the children's experiences at these clubs. Despite their attendance at clubs, many of the children with WS were reported to experience difficulties at these clubs which limited their participation in the activities. Many parents reported their children to experience significant difficulties in keeping up with sports or dance routines, which may reflect an impairment in visual-spatial skills (Carney et al., 2013). Some parents also reported that their child was slower at following instructions than their mainstream peers which again limited their ability in these activities. As a consequence, many parents reported the need for careful consideration when selecting clubs which were appropriate for their child to attend.

Amendments included smaller group sizes, greater levels of support and attending classes with a younger age group.

The accumulated findings from this chapter provide insight into the perceptions of parents', teachers' and children with WS regarding the nature of the child's peer relationships. These findings have provided support for claims of substantial peer relationship difficulties in children with WS, in line with the findings from chapter 6. However, the current chapter builds on this by using qualitative analysis to gain a deeper level of insight into the nature of these social difficulties and the implications of this for the child and the family. Many of the children with WS were reported by their parents and teachers to demonstrate difficulties establishing friendships and to show atypical patterns of behaviour within social interactions with peers, including unusual patterns of communication and poor awareness of social boundaries. These social difficulties were reported to have implications for the child's vulnerability and parents' anxieties and family functioning. While these social difficulties were evident across both parents' and teachers' accounts, these social difficulties were not reported by the children themselves, suggesting that children with WS have a poor perception of their own peer relationships.

Chapter 8: General discussion

8.1. Aims of the present thesis

The present thesis had two overarching aims. The first aim was to examine the characteristics of SAB and peer relationships in children with WS, including an examination of within-syndrome variance in these social experiences. These social difficulties have been identified as core features of the WS social phenotype (Jawaid et al., 2012; Jones et al., 2000). However, research in this field is relatively sparse. With consideration to the relative lack of research and the methodological limitations across previous studies, there was a need for further research to be conducted to obtain insight into the characteristics of these social difficulties across environmental settings and across perspectives.

Previous research has predominantly used a single informant approach when examining social functioning in WS (Bellugi et al., 1999; Davies et al., 1998; Martens et al., 2009). The present thesis builds on this by soliciting information from the children with WS, their parents and teachers in order to examine the stability of social functioning across social contexts and informants.

Furthermore, previous researchers have predominantly used either a quantitative or a qualitative methodology to solicit information on the phenomenon under investigation (Cuckle & Wilson, 2002; Elison et al., 2010; Martens et al., 2009). Both quantitative and qualitative methods merit consideration within research, with each providing valuable insights (Braun & Clark, 2006; Fields, 2009). The present research builds on this by using a mixed method approach. Quantitative data were obtained in chapter 3 and chapter 6 on the children's SAB and peer relations through an analysis of questionnaire ratings. This provided insight into the prevalence of these social difficulties and associations between factors. This was supported by qualitative data in chapter 5 and chapter 7 on the manifestation of these social difficulties and the implications of these behaviours for the child with WS and his or her family which were obtained using semi-structured interviews.

The second aim of the present thesis was to examine the role of psychological processes, specifically, the role of response inhibition, information processing and social functioning skills in the SAB and peer relationships of children with WS. Achieving an insight into the psychological variables which underlie these social difficulties was suggested to contribute to the understanding of these behaviours. Within chapters 3 and 4, the relationships between EF impairments, information processing biases, atypicalities of social functioning and levels of SAB in WS were examined. In chapter 6 and 7, the relationship between atypicalities of social functioning and peer relations in children with WS was investigated. These findings will be discussed in greater depth within this chapter.

8.2. Key findings

8.2.1. Evidence for atypical social approach in children with Williams syndrome

Previous research indicated that individuals with WS engage in high levels of indiscriminate SAB (Järvinen-Pasley et al., 2010; Jawaid et al., 2012). This hyper-social profile has been illustrated using a range of methods including experimental rating tasks, parental questionnaires and behavioural observations (Bellugi et al., 1999; Dodd et al., 2010; Doyle et al., 2004). The accumulated findings provided support for higher levels of SAB in individuals with WS when compared to TD individuals matched on chronological age (Bellugi et al., 1999; Doyle et al., 2004; Järvinen-Pasley et al., 2010) and groups with other developmental disorders (Doyle et al., 2004).

The present thesis aimed to build on the methodologies of previous researchers who utilised a single informant approach (Doyle et al., 2004; Porter et al., 2007). Within chapter 3, SAB ratings were solicited from the children with WS and their parents, thus providing an assessment of SAB across environmental settings and from different perspectives. The children with WS indicated a significantly higher willingness to approach an unfamiliar person in Adolph's Approachability Task than TD children who were matched for verbal ability and TD children who were matched for non-verbal ability. By individually matching the children with WS to TD children on verbal and non-verbal performance, it was possible to control for the effect of intellectual delay on task performance. As a result, stronger and more reliable conclusions could be drawn regarding SAB in WS.

The experimental and parental ratings obtained in chapter 3 illustrated high levels of SAB in children with WS. All of the children with WS were reported by their parents to show a high level of interest in speaking with unfamiliar adults and to be very trusting of strangers. There was very little variance in the parents' ratings in these items, indicating that these experiences were prevalent across this sample. These findings were further supported by qualitative interview extracts from the parents within chapter 5. Many parents reported that their child with WS was indiscriminate in their SAB. Several parents attributed this to their child’s inquisitive personality, while other parents attributed this to their child’s difficulties inhibiting their behaviour within social situations. Furthermore, several parents indicated that their child with WS did not have an ingrained sense of danger. This was identified as an area of concern for several of the parents who worried about the implications of this for their child’s vulnerability to strangers. In consequence, several parents highlighted their need to closely supervise their child when in public places in order to monitor their child’s interactions.

While all of the children with WS showed an interest in speaking with strangers, there was variance in the children’s levels of tactile behaviour towards these strangers. This finding suggested that while SAB was prevalent in the children with WS, where all of the children demonstrated a strong social interest, there was variability in the severity of this SAB. However, as has been noted across this thesis, caution is required when interpreting these findings as this variance was not subjected to statistical analysis. These quantitative findings were further supported by qualitative data in chapter 5 where individual differences in the manifestation of SAB were explored.

The use of both quantitative and qualitative data builds substantially on previous research which used a single methodology to examine SAB in WS (Doyle et al., 2004; Lough et al., 2016; Martens et al., 2009). While some children were reported to engage in high levels of indiscriminate affection, where parents attributed this to the child’s difficulties inhibiting their behaviour in social interactions, this was not reported across the entire sample. Some parents indicated that while their child showed a strong interest in speaking to other people, he or she only engaged in tactile behaviour towards familiar people. In some cases, this was attributed to the child’s enhanced understanding of socially appropriate behaviour which they had developed with age as a result of maturity and education.

Previous research indicated that children with WS had a poor understanding of stranger danger (Riby et al., 2014). Within the current research, several parents reported that their child showed an understanding of stranger danger principles. The present findings are in contrast to the previous findings of Riby et al. (2014). This was suggested to be due to differences in the measurement of stranger danger between studies. Although several of the parents' reported that their child with WS understood stranger danger principles, many of the children were reported to engage in indiscriminate SAB. Questionnaire data alone cannot provide an explanation for this disparity in findings. This thesis was able to explore this in further depth through an analysis of the parents' interview extracts. Many parents reported that while their child with WS demonstrated a basic understanding of stranger danger principles and was able to provide the correct answers on paper when questioned, he or she often struggled to put this understanding into practice when in real-life social interactions.

While it was evident across the parents' reports that the children with WS showed difficulties regulating their behaviour around strangers, the findings from chapter 4 indicated that there was no significant relationship between the children's response inhibition performance and their levels of SAB. These findings contrast with Little et al. (2013) and could be suggested to challenge the assumptions of the frontal lobe hypothesis. However, this non-significant relationship is suggested to be accounted for by the task used in the present thesis. Within the present thesis, the Sun Moon Stroop task was administered which provided a measurement of the children's reaction time when required to execute a verbal response. However, previous researchers reported that children with WS demonstrated poorer performance in a motor inhibition task than they did in the verbal inhibition task (Carney et al., 2013). This aligns with the findings in the current thesis where many of the parents attributed their child's difficulties within social interactions as due to their poor behavioural regulation. It is therefore suggested that the Sun Moon Stroop task may not have provided a reliable test of the theoretical principles of the frontal lobe hypothesis. Within future research, it would be informative to further examine the relationship between inhibition and SAB using tasks which tap into the children's motor inhibition in social settings in order to draw strong conclusions regarding the theoretical assumptions of the frontal lobe hypothesis.

The accumulated findings across the experimental task, parental questionnaire ratings and parental interview extracts have highlighted high levels of SAB in children with WS, with implications for the children's vulnerability and family functioning. There was within-syndrome variance in the questionnaire ratings and individual differences in the interview data. These findings further highlight a heterogeneous profile of SAB in children with WS.

8.2.2. Evidence for impaired peer relations in children with Williams syndrome

Previous researchers identified substantial social impairments and peer relationship difficulties in adults with WS (Davies et al., 1998; Elison et al., 2010; Udwin, 1990). Very little research has been conducted to examine peer relations in children with WS. The present thesis addressed this gap in the literature by using a multi-informant approach to investigate peer relationships in children with WS across settings and across informants. This methodology builds substantially on previous research which predominantly solicited parent reports on peer relations in WS (Davies et al., 1998; Greer et al., 1997; Udwin, 1990).

The present thesis involved administering a range of questionnaires, standardised and bespoke, and semi-structured interviews to investigate peer relations in children with WS. The children with WS were reported by their parents and teachers to demonstrate significantly greater peer relationship difficulties than TD children who were matched on verbal and non-verbal ability. This is the first study, to the author's awareness, to achieve an in depth examination of peer relations in children with WS using multiple measures and multiple informants. The current findings indicate that peer problems persist across environmental settings, reinforcing the importance of expanding our understanding of these social difficulties. The children with WS also showed difficulties in several areas of socialisation when compared to a normative population sample. The children with WS showed significantly greater deficits than the normative population within their interpersonal relationships, play skills and social participation.

The use of standardised questionnaires allowed conclusions to be drawn regarding the atypical nature of peer relationships in children with WS. Novel questionnaires were also developed for use in the current thesis to expand our understanding of the characteristics of peer relations in WS which existing measures did not examine. This provided insight into the quality of the children's peer relationships, their social inclusion, extra-curricular engagement with peers and the appropriateness of their social interactions.

Parent and teacher questionnaires were developed for use in the present thesis. These questionnaires examined the characteristics of peer relations in children with WS across the home and school environment. Many of the children with WS were reported by their parents and teachers to show substantial difficulties forming and sustaining friendships with peers. Both parents and teachers also reported low levels of social inclusion, where the children with WS were often reported to be social isolated during recreational breaks and rarely engaged with peers outside of school. These questionnaire ratings were further supported by qualitative data in chapter 7 which provided further insight into these social difficulties. This provided insight into parents' accounts for their child's social difficulties, where several parents attributed their child's peer relationship difficulties as due to their communitive impairments, difficulties with personal space regulation and the difference in interests from their peers. These insights would not have been possible to obtain using questionnaire data in isolation.

Across the questionnaire ratings, several parents reported that their child with WS attended group birthday parties; however, the children's engagement in one to one activities with their peers was reported to be significantly lower. The interview data in chapter 7 provided further insight into this disparity in findings. Several parents reported that their child with WS enjoyed interacting with children, however, did not have a steady reciprocal friendship. In consequence, these children were reported by parents and teachers to rotate between their classmates. Parents and teachers also reported that the children with WS struggled to sustain a reciprocal conversation with peers and had substantial deficits within their play skills which impacted their ability to engage with peers. Together these findings are suggested to provide an explanation for the low levels of social inclusion and extra-curricular engagement with peers which were indicated across the questionnaires. 60% of the children with WS were reported to attend extra-curricular clubs and activities. The interview data in chapter 7 developed this further by providing insight into the nature of these clubs and the success of the children's interactions. While 60% of children attended clubs, the majority of children with WS were reported to experience difficulties participating in the activities and engaging in social interactions at these clubs. Furthermore, several parents reported a need for high levels of parental support and supervision. This study has greatly expanded our understanding of peer relations in children with WS by using various measures and informants. By obtaining both quantitative and qualitative data, questionnaire ratings allowed an examination of the prevalence of social difficulties in children with WS, while the interview extracts provided a greater depth of insight into these social difficulties by drawing on personal experiences.

The recruitment of parents and teachers in the current thesis was highly informative in providing accounts into the children's peer relationships across environmental settings. Both parents and teachers reported that the children with WS experienced substantial difficulties sustaining friendships, engaging in play with peers, low levels of social inclusion and evident difficulties engaging in reciprocal social interactions. This consistency across informants indicates that these social difficulties are core features of the WS social profile which persist across contexts.

However, the parents rated the child's peer problems within the SDQ significantly higher than teachers' ratings. Certain aspects of the children's social interactions were rated as more problematic by parents than teachers. For example, the questionnaire data indicated that three quarters of parents reported that their child engaged in problematic tactile behaviour within peer interactions in comparison to one quarter of teachers. This was further supported by qualitative extracts, where parents described their child’s overly affectionate behaviour towards peers. Several parents reported that this posed implications for their child’s peer interactions, leading to a level of discomfort among peers. It is suggested that there are less opportunities for children to exercise tactile behaviour within school settings due to greater rules and regulations. These findings highlight the importance of using a multi-informant approach in order to examine for differences in behaviour across environmental settings.

In chapter 7 of the current thesis, the children with WS were interviewed regarding their peer relationships. While parents and teachers reported substantial peer relationship difficulties among the children with WS, the children themselves believed that they had a close friend. These findings suggest that children with WS may have a poor perception of their own peer relationships. These findings were supported by interview extracts from several parents who indicated that despite apparent difficulties within friendships, their child perceived that they had a best friend. However, many parents reported that this friendship was not reciprocated by the peer. Several parents also noted that their child had a poor understanding of the concept of friendship and considered someone to be a friend immediately upon meeting. This was further illustrated within the interviews with the children with WS where many of the children struggled to define the term friendship. These findings further highlight the complexity of peer relationships for many children with WS.

While peer relationship difficulties were evident in this sample of children with WS, there was a small degree of variance in the children's social experiences. A few of the children with WS were reported to have a strong friendship with a peer and to be well integrated in their peer group. Furthermore, a few children with WS were also reported to have an awareness of social norms and appropriate practices within social interactions. This heterogeneity in WS further highlights the importance of adopting an individual approach when assessing the support needs of children with WS.

8.2.3. Negative association between atypicalities of social functioning, SAB and quality of peer relations in children with Williams syndrome

Atypicalities in social functioning, specifically impairments in social cognition and social communication have been identified as significant predictors of peer relations in children with a range neurodevelopmental disorders including autism and ADHD (Rowley et al., 2012; Sibley et al., 2010). To the author's awareness, no research thus far has investigated the relationship between atypicalities in social functioning and interactions with strangers and peers in children with WS. The current thesis addressed this gap in research. In chapter 3, the relationships between social functioning impairments, as measured using parents' ratings in the SRS, and levels of SAB were examined in a sample of children with WS. There was a significant positive relationship between impairments in social awareness and the children's levels of SAB within Adolph's Approachability Task. There was also a significant positive relationship between impairments in social cognition and the children's levels of SAB as indicated within Adolph's Approachability Task and parental questionnaire ratings. In chapter 6, the relationships between social functioning impairments, as measured using parents' ratings in the SRS, and the quality of the child's peer relationships were examined. Peer relationships were examined by administering standardised measures of social functioning and peer relationships and questionnaires specifically designed to measure the characteristics of peer relations. These questionnaires were administered to the parents and teachers of the children with WS. There were several relationships between impairments in social awareness, social cognition and social communication and measures of socialisation and peer relations in the children with WS.

The findings across chapters 3 and 5 of the present thesis suggest that children with WS may experience difficulties engaging in socially appropriate interactions with peers and strangers due to impairments across social functioning, specifically impairments detecting and interpreting social cues within interactions. The findings from the present thesis are in line with previous research findings which provided evidence of a poor awareness of societal norms and boundaries (Lough et al., 2015), and difficulties interpreting other people's mental states in individuals with WS (Porter et al., 2008; Tager-Flusberg & Sullivan, 2000). These findings build on this previous research by demonstrating the implications of these atypicalities of social functioning for the children's social adjustment. These findings are suggested to align with Selman's psychological framework on friendship development, where social-cognitive skills are proposed to play a critical role in the development of effective peer relationships during childhood (Selman, 1980).

8.3. Limitations and future directions of the current research

The present thesis has contributed to an increased understanding of SAB and peer relations in children with WS. However, it is important to acknowledge the limitation of the current research. These limitations will be discussed, with suggestions for future research.

The primary focus of the present thesis was to achieve an in depth examination of social behaviour in WS across contexts and across informants. In achieving this level of insight, the current research was constrained to a small sample size (N=21). The rarity of WS and as a result, the geographical spread of families across the UK, posed evident implications for participant recruitment. The present author obtained a sample size that was in line with previous research (Järvinen-Pasley et al., 2010; Riby et al., 2013). However, the relatively small sample size recruited posed implications for statistical analysis. Within the present thesis, significant relationships were identified between several indices of social functioning and peer relations and SAB in children with WS. However, it was not possible to examine the predictive relationship between these variables due to a lack of statistical power. Where possible, postal methods could be explored as an avenue to maximise sample size within future studies. Utilising postal correspondence methods has the potential to maximise opportunities for participant recruitment and in consequence, opportunities for statistical analysis.

Several novel measures of SAB and peer relations were designed for use in the present thesis to meet gaps within existing published measures. This achieved a valuable insight into a profile of social difficulties in children with WS by soliciting further information on the manifestation of these social difficulties across environmental contexts and insight into the implications of these social difficulties for the children with WS and their families. The specific focus of the present thesis was to achieve an examination of the patterns of behaviour and relationships between these behaviours in children with WS. Future research could obtain more extensive comparisons with TD children by administering these measures with both the children with WS and a sample of TD controls. This would allow the author to draw further conclusions regarding the atypical nature of the peer relations and SAB of the children with WS in the current research.

Within future research, it would also be valuable to administer the current questionnaires with groups of children with other developmental disorders. Developmental disabilities are characterised by unique profiles of social, behavioural and cognitive impairments. A between-group comparison of social impairment could be informative in allowing the author to draw conclusions regarding the syndrome specificity of these social traits and impairments to WS, or alternatively, if these social difficulties are generalizable across developmental and intellectual disabilities. Furthermore, with consideration to the similarities and differences in the phenotypic profiles of developmental disabilities, between-group comparisons could be informative in providing insight into the role of cognition, behaviour and intellectual impairments in the manifestation of these social difficulties in WS.

This study can be considered to have advanced the methodological approaches of previous researchers through the use of a multi-informant approach. Although parent reports have been identified as a strong measure of social behaviour in WS (Fisher et al., 2014), this is an indirect measurement of social behaviour. Further researchers should seek to further validate these findings by soliciting behavioural observations of the children's peer relations and SAB.

8.4. Implications of the present research findings

As detailed in the key findings earlier in this chapter, the current thesis has contributed substantially to a very limited area of research by expanding our understanding of social difficulties in children with WS. In addition to addressing important gaps within the literature, the current thesis has developed existing methodologies in this field through the design of several new instruments. The present thesis also has potentially important practical implications. It is suggested that the current findings have the potential to inform the design of future interventions and can contribute to raising the awareness of WS. These implications will be discussed in greater depth below.

8.4.1. Methodological advances

Several questionnaires and tasks were developed for use in the present thesis due to the lack of suitable measures within the literature. The design of novel measures provided a valuable insight into these social difficulties in WS by tapping into constructs of social behaviour which had not been examined by previous researchers. With a consideration to the gaps within the literature, a parent report social approach questionnaire was developed to examine the manifestation of SAB, specifically, the children's disinhibited communication and physical interactions with strangers. Items were also included which examined the children's social vulnerability around strangers and parents' concerns regarding this. Furthermore, while previous researchers examined peer relations in adults with WS by conducting interviews with parents (Davies et al., 1998; Udwin, 1990), there was a need for both questionnaire and interview schedules to be devised which could tap into the characteristics of peer relations in children with WS. The present thesis builds substantially on previous measures in this field by developing questionnaire and interview schedules to solicit parents' and teachers' perceptions of the child's peer relations. Through further application, these measures can be used by future researchers to investigate social difficulties in WS, TD populations and groups with other developmental disorders. These methodological advances therefore have potential implications for researchers not only in this specific field of research, but can be applicable to researchers across related fields.

8.4.2. Practical implications

The findings from the current thesis have greatly expanded the understanding of social vulnerability in children with WS. The use of a multi-informant approach provided insight into the presence of social difficulties in children with WS across social environments. While there is no simple solution to eradicate these difficulties, the findings within the present thesis can have practical implications in raising awareness of problematic areas of social functioning for children with WS.

Within the Scottish Educational Curriculum, there is an integral focus on promoting positive social relationships. An important component of the Scottish Educational Curriculum for Excellence is 'personal and social development'. The Scottish

Educational Curriculum for Excellence sets several aims which educators are required to attain through their teachings. This includes the acquisition of friendship skills, developing an understanding and execution of appropriate behaviour within peer interactions e.g. personal space regulation, and the development of interactive play skills (The Scottish Government, 2004). By further expanding teachers' awareness of social difficulties in children with WS, this knowledge could be better incorporated by teachers into lesson plans in order to target specific areas of social difficulty.

On national and international levels, legislations promote the social wellbeing and inclusion of children. At the national level, The Children and Young People (Scotland) Act 2014 adopts an approach termed 'getting it right for every child' where there is a core focus on promoting equality and social inclusion in children across social, educational and economic barriers (The Scottish Government, 2014). Furthermore, on an international level, in line with the Rights Respective School Award, teachers have a duty of care to support all children to exercise their rights of access to friendship and their rights to play and engage in recreational activities (UNICEF, 1990). In consequence to these legislations, peer relationships are now a fundamental component within the education system. It is suggested that the integration of the findings across the present thesis will be informative to teachers. While the teachers were able to report on their observations of a single child within the classroom environment, the current findings can help to expand teachers' understanding of this rare disorder. This can include raising their awareness of the factors believed to underlie these social difficulties, where social functioning deficits, including impairments within social cognition, social awareness and social communication were positively associated with peer relationship difficulties among the children with WS. It is suggested that by expanding teachers' understanding of WS, teachers will be better equipped to support children with WS in their everyday peer interactions.

As a result of national and global policies which promote equality among children, many schools have implemented various systems to facilitate the development of positive peer relationships. Nurture units are aimed at providing social support for children with emotional, social and behavioural difficulties. Through these nurture schemes, children are supported in their development of social skills and peer relationships through guidance from trained professionals (Bennathan & Boxall, 2013). Nurture groups have been widely reported to be successful in promoting positive social relationships during primary and secondary education (Cefai & Cooper, 2011; Colley, 2012; McKinney & Hall, 2016). With consideration to the social interaction, social communication and play deficits which were widely reported in the present thesis, this acquired knowledge could be put to practical use in supporting children with WS to develop stronger peer relationships through the use of these nurture schemes. Another system which could be utilised to support children with WS in their peer relations at school is a 'buddy system'. Through the use of this practice, children with social difficulties can be paired with another pupil who would be encouraged to look out for this child and to provide a source of companionship (Bucher & Manning, 2003).

Within the current thesis, many of the children with WS were reported to struggle to sustain friendships with their TD peers. These difficulties were reported to become more evident as the children got older. This was attributed in many cases to the peers' increased independence and a difference in interests. In chapters 6 and 7 of the current thesis, information was solicited from the parents regarding their child's attendance at extra-curricular clubs. While more than 60% of the children were reported to attend extra-curricular clubs, many parents identified their children's difficulties participating at these clubs, with this most prominent when attending mainstream clubs. This further reinforces the importance and the necessity of special needs clubs for children with disabilities to establish friendships with other children of a similar intellectual ability. Furthermore, the current findings can inform teachers' behaviour within the classroom setting, where teachers' awareness of the value of interactions with other children with disabilities could influence their decisions when assigning pupils to groups for activities, further promoting opportunities to establish friendships.

Across chapters 3, 4 and 5 of this thesis, there was evidence to support high levels of indiscriminate friendliness towards strangers in children with WS. This was illustrated using an experimental rating task and by soliciting parents' perceptions using questionnaires and interviews. Stranger awareness is an integral part of the Scottish Educational Curriculum for Excellence under the element 'personal and social development'. This addresses the need for all children to acquire an awareness of stranger danger, to be able to differentiate between familiar and unfamiliar people and to be equipped to engage in socially appropriate practices in these situations (The Scottish Government, 2004). In light of the current findings of inappropriate and often indiscriminate interactions with strangers, this knowledge on the vulnerability of children with WS can have important implications for teachers who will be better informed to monitor the child’s behaviour across different situations in order to protect the child’s safety and wellbeing.

The present thesis findings are suggested to have important implications in raising awareness among peers regarding social difficulties in children with WS. It is proposed that if peers understand that children with WS have these difficulties in personal space regulation and a poorer awareness of socially appropriate behaviour, the peers may be better prepared if they find themselves within these situations. Expanding peers’ awareness of the characteristics of WS could result in peers being more understanding of the children’s behaviours and idiosyncrasies. Increasing peers’ understanding of different disabilities and promoting respect and anti-bullying through school policies, can in turn reduce the potential for hazardous conflicts among peers.

This thesis identified associations between several indices of social functioning and social difficulties in WS. In light of the current findings of a significant relationship between atypicalities of social functioning and both SAB and peer relations in children with WS, these findings raise practical implications which can be tested in the course of behavioural interventions. Future interventions should be designed which facilitate the development of important social skills among children with WS, with a particular focus on developing children’s abilities within social awareness, social cognition and social communication due to their association with the atypical social profile in WS. Future interventions could target these key social skills through education and practical application of techniques which promote reciprocal communication, interpretation of social cues and personal space regulation.

8.5. Conclusion

The present thesis has greatly expanded our understanding of two key areas of difficulty among children with WS: SAB and peer relationships. Through methodological advances, the current thesis builds substantially on previous research by providing insight into the characteristics of these social difficulties across different environmental contexts. The use of a quantitative method of analysis allowed an examination of the prevalence of these characteristics, while qualitative methods provided a detailed insight into the manifestation of these social difficulties and the implications for the children with WS and their families. There was also within-syndrome variance in this sample with regards to the children's engagement in SAB and the quality of their peer relationships, in line with the heterogeneous nature of this disorder.

The present thesis has evident implications for this field of research. Several novel measures were designed for use in the present thesis. Through further testing, these measures can be used by researchers to further explore these areas of social impairment. The current thesis findings also have important practical implications. This thesis is suggested to be informative in further expanding the awareness of the child's teachers and peers regarding areas of social difficulty among children with WS. While parents' and teachers' reports in chapter 7 indicated that many of the child's peers were sensitive to the needs of the child with WS, several parents and teachers noted that peers were often overwhelmed by the child's disinhibited nature.

By educating peers regarding the characteristics of WS, these findings can be used to promote more prosocial interactions between the children with WS and their peers.

Overall the current thesis has highlighted several areas of social difficulty for children with WS, with the children's atypical interactions with both strangers and peers being identified as areas of concern among parents. These findings highlight the importance of research in this area and the need for future research to be targeted towards designing interventions and support for children with WS and their families. By supporting children with WS within their daily social lives, this can have implications in reducing the child's social vulnerability and the negative impacts on family life.

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Appendices

Appendix 1: Development of computerised Sun Moon Stroop Task

Rationale:

Pasalich et al. (2010) argued that the standardised Sun-Moon Stroop task did not provide a sensitive measure of the accuracy of the children's inhibition performance. Pasalich et al. (2010) suggested that the children's response patterns may have influenced their results in this task, where children who focused more on the speed of their responses may have scored higher than those who focused on accuracy.

Task design:

A modified computerised version of this task was developed for application in the present thesis which aimed to overcome this limitation. This bespoke task was trialled on a sample of TD children (N=64). This task was developed using the programming software 'E-Prime'. There was a 10 trial practice block, followed by 20 experimental trials. In the practice trials the children were provided with feedback on the accuracy of their responses. This task followed a similar procedure to the standardised Sun-Moon Stroop task. In the experimental condition, the participants were instructed to press the Sun button whenever they saw an image of a moon and to press the Moon button whenever they saw an image of a Sun. In contrast to the standardised Sun Moon Stroop task, each stimulus was presented in an independent trial and the trials did not move on until a response was made. This was believed to achieve a more accurate measurement of performance accuracy.

Results:

An analysis of performance in the TD sample showed that there was a weak correlation in scores between the paper and computerised versions of the Sun-Moon Stroop task (r = 16, p

= .208). The children’s scores in the computerised task showed a negative skew (-1.84) which indicated ceiling level performance. Due to the poor correspondence between measures and the skewed nature of the data in the computerised task, the standardised Sun-Moon Stoop task was used as a measure of response inhibition across subsequent experimental chapters.

Appendix 3: Social approach behaviour scale (Parent report)

1. My child has approached a stranger in the street/public place.

1	2	3	4	5
Never	Rarely	Sometimes	Quite often	Often

2. My child has hugged, kissed or held hands with a stranger when out in public.

1	2	3	4	5
Never	Rarely	Sometimes	Quite often	Often

3. My child has been tactile towards adults in authority e.g. teacher, dentist, doctor.

1	2	3	4	5
Never	Rarely	Sometimes	Quite Often	Often

4. My child shows a desire to speak to adults they meet, even those that they have not met before.

1	2	3	4	5
Strongly disagree	Disagree	Neither agree nor disagree	Agree	Strongly Agree

5. My child shows a desire to speak to other children they meet, even those that they have not met before.

1	2	3	4	5
Strongly disagree	Disagree	Neither agree nor disagree	Agree	Strongly Agree

6. My child requires constant supervision when out in public due to their tendency to approach other people.

1	2	3	4	5
Strongly disagree	Disagree	Neither agree nor disagree	Agree	Strongly Agree

7. My child’s approach behaviour towards strangers worries me.

1	2	3	4	5
Strongly disagree	Disagree	Neither agree nor disagree	Agree	Strongly Agree

8. My child’s approach behaviour impacts upon the family.

1	2	3	4	5
Strongly disagree	Disagree	Neither agree nor disagree	Agree	Strongly Agree

9. If asked my child would say that they know they should not approach strangers.

1	2	3	4	5
Strongly disagree	Disagree	Neither agree nor disagree	Agree	Strongly Agree

10. My child’s over-friendly behaviour has placed them in a vulnerable or awkward situation in the past.

1	2	3	4	5
Strongly disagree	Disagree	Neither agree nor disagree	Agree	Strongly Agree

11. In my opinion, my child would follow a stranger if invited.

1	2	3	4	5
Strongly disagree	Disagree	Neither agree nor disagree	Agree	Strongly Agree

12. My child is very trusting of other individuals.

1	2	3	4	5
Strongly disagree	Disagree	Neither agree nor disagree	Agree	Strongly Agree

Appendix 7: Social approach behaviour interview schedule (Parent report)

1. **Can you tell me about your child’s interactions with strangers?**

How often does your child approach strangers?

Does your child show an interest in conversing with stranger?

Can you provide any examples which illustrate this?

2. **Does your child engage in tactile behaviour towards strangers?**

Does your child engage in tactile behaviour towards adults in positions of authority e.g. teachers, doctors?

How often does your child engage in these behaviour? Can you provide any examples which illustrate this?

3. **In your opinion, is your child very trusting of other people?**

Can you provide examples which illustrate this?

4. **Does your child require constant supervision when in public?**

Can you provide examples which illustrate this?

5. **Is your child’s SAB a concern for you**?

Can you provide examples which illustrate this?

6. **Does your child’s behaviour with strangers have an impact on family life?**

Can you provide examples which illustrate this?

7. **In your opinion, does your child understand that they should not approach strangers?**

Can you provide examples which illustrate this?

Appendix 8: Quality of peer relationship questionnaire (Parent report)

Scoring system:
1- Strongly disagree
2 - Disagree
3- Neither agree nor disagree/neutral
4 – Agree
5 – Strongly agree

1. My child has one or more close friends.

1 2 3 4 5

2. My child can maintain long lasting friendships for several months, or longer.

1 2 3 4 5

3. My child often experiences falling outs with his/her peers.

1 2 3 4 5

4. My child will go right up to other children and give them a hug.

1 2 3 4 5

5. My child displays high levels of physical contact towards other children (e.g. holding hands).

1 2 3 4 5

6. My child understands that you should not get so physically close to another person that it makes that person uncomfortable.

1 2 3 4 5

7. My child is invited to the homes of other children.

1 2 3 4 5

8. My child is invited to the birthday parties of other children.

1 2 3 4 5

9. My child attends social clubs or activities with peers.

1 2 3 4 5

Appendix 9: Quality of peer relationships questionnaire (Teacher report)

1 – Strongly disagree
2 – Disagree
3 – Neither agree nor disagree
4 – Disagree
5 – Strongly disagree

1. This child has a best friend in the class.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

2. If applicable: This friendship is reciprocated by the other person.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

3. This young person has one or more friends in the class that he/she associates with.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

4. This young person experiences difficulties in forming friendships.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

5. This young person experiences difficulties in maintaining friendships.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Regarding questions 4 & 5, can you provide any relevant examples or further information to support this?

6. This young person is frequently left out by peers in school.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

7. This young person often experiences falling outs with his/her peers.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

8. This young person is included by peers in class e.g. conversations.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

9. This young person is chosen as a partner for work activities/P.E.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

10. This young person is included by peers during breaks/lunch time.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

____

____

____

____

11. This young person is picked on by his/her peers.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this? (Please do not include any names).

____

____

____

____

12. This young person is greeted/talked to by other children around the school.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

____

____

____

____

13. This young person requires one to one support in class.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

____

____

____

____

14. This young person requires one to one support at break/lunch time.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

____

____

____

____

15. This young person shows an interest to speak to other adults in the school than would normally be expected.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

____

____

____

____

16. This young person becomes upset if his/her peers choose to play with other children?

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

____
____
____
____

17. This young person becomes anxious when in larger groups of children/social situations.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

____
____
____
____

18. This young person is overly tactile towards other children (hugging, holding hands).

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

____
____
____
____

If applicable:

Have any children reported to dislike this behaviour?

Yes No

19. This young person understands that you should not get so physically close to another person that it makes that person uncomfortable.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

20. If applicable, I believe this tactile behaviour to hinder their social interactions.

1 **2** **3** **4** **5**

Strongly disagree Strongly agree

Can you provide any relevant examples or further information to support this?

21. Any further comments on this young person’s experiences of friendships and the difficulties faced.

Appendix 10: Quality of peer relationships (Parent semi-structured interview)

1. **In your opinion, do you consider your child to have a best friend?**

Further probes:

If yes: How does your child know this person? Does this person have a disability? In your opinion is this friendship reciprocated by this person?

If no: Does your child have any friends or acquaintances that he/she plays with and/or talks to?

2. **Can you tell me anything about your child's ability to make friends?**

Probes: Are there any examples which you feel highlight this?

3. **When it comes to activities with peers, do you ever find the situation where your child is left out to a certain extent?**

Probes: If you don't mind elaborating, are there any examples which spring to mind? Do you feel that your child notices this/feels left out?

4. **In your opinion, do you think your child is drawn to certain age groups when initiating contact with others**?

   Probes: For instance, does he/she initiate contact with both children and adults equally? Are there any examples which illustrate this?

5. **Are there any situations that spring to mind where your child has been picked on by peers?**

6. **Does your child experience anxiety in social situations?**

Probes: Are there any examples of this which spring to mind? In your opinion, has this anxiety affected his/her social relationships?

7. **Does your child demonstrate any of the following when in social situations: excessive chatter, intense behaviour towards others, asking inappropriate questions?**

Probes: Are there any examples which spring to mind? Do you feel that any of these behaviours affect his/her social relations?

8. **Can you tell me anything about your child's ability to hold conversations with other people?**

9. **Does your child understand that his/her friends may also have others friends who are not in the same group whom they spend time with?**

Probe: Does this upset your child? Is he/she able to play by himself/herself if these friends are unavailable?

10. **Does your child engage in any of the following activities with peers? (Computer/video games, listening to or making music, engaging in sports, general play or anything else?).**

Probe: If yes, how often does he/she engage in these activities?

If no, how does your child spend his/her free time?

11. **Does your child attend any organised clubs/activities?**

Probes: Does he/she enjoy these? Is he/she included by the other children e.g. in teams, playing, talking. Does he/she experience any difficulties at these clubs?

12a. **How often, if ever, does your child get invited to the homes of other children from school or in the neighbourhood?**

12b. **How often, if ever, does he/she get invited out to activities by other children's parents e.g. dinner, the cinema, bowling?**

12c. **How often, if ever, does he/she get invited to the birthday parties of other children?**

13. **From your experience, does your child often go up to other children and hug them or hold their hand?**

Probe: Has this had any positive or negative effects to his/her social relationships?

14. **Do you have any further comments on the quality of your child's social life and your hopes/concerns for his/her future?**

Appendix 11: Peer relations interview schedule (children with WS)

1. **Do you have a best friend?**

Probes: If yes: How do you know this friend?

2. **Are the other children in school nice?**

3. **What do you like to do with your friends?**

Probes:

Do your friends come over to your house?

Do you go over to their house?

Do you play inside or out in the garden/street?

Do you sometimes go out with your friends and their families? What do you do?

What's your favourite game to play with friends?

4. **Do you go to any clubs?**

Probes:

Do you like these clubs?

What is your favourite thing about these clubs?

Do you have friends that you talk to at the clubs?

Do you have friends that you play with at the clubs?

5. **One girl told me that she argues with her friends sometimes. Do you ever fall out or argue with your friends?**

Probes: When does this happen? How does this make you feel?

6. **One boy told me that his friends said that he couldn't play football with them. Do your friends ever tell you that you can't play with them?**

Probes: Does this happen a lot or just sometimes? How does this make you feel?

7. **Do you like talking to the teachers at school?**

8. **Do you like talking to the children in your class?**

Probes: What kind of things do you talk about?

9. **What do you do at break time at school?**
   What do you do at lunch time at school?

Probes: Do you spend them with the other children? Who do you eat lunch with? Who do you talk to?

10. **What makes a good friend? What do you like about your friends? Do you think friends are important?**

Appendix 12: Parent information sheet 1 (WS recruitment)

University of Strathclyde
Humanities & Social Sciences

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

Participant information sheet

Name of Department: School of Psychological Sciences and Health
Title of the study: Constructing a profile of social behaviour in Williams Syndrome

---

Introduction

My name is Amanda Gillooly and I am a postgraduate researcher at the University of Strathclyde, undertaking my PhD in psychology. I am working with Dr Sinead Rhodes and Professor Kevin Durkin within the School of Psychological Sciences and Health at the University of Strathclyde and Dr Deborah Riby from Durham University and this project is part of the North East Williams Syndrome research group. Your child is invited to take part in a research study. Before you decide, it is important for you to understand why the research is being carried out and what it will involve. Please take the time to read the following information sheet carefully. If you have any questions regarding this study please do not hesitate to contact either myself or my supervisors on the contact details below. I can be contacted via email at amanda.gillooly@strath.ac.uk.

What is the purpose of this investigation?

This study is being conducted to investigate some of the factors which may underlie the heightened social approach behaviour and peer relationship difficulties experienced by some individuals with Williams Syndrome. The results of this study will contribute significantly to researchers' understandings of this disorder, facilitate the development of further support for individuals with Williams Syndrome and help to further raise awareness.

Do you have to take part?

No, you don't have to take part and participation in this study is entirely voluntary. Your child may withdraw from this study at any time without giving a reason. You can withdraw your child and his or her responses from the study prior to the completion of the testing if you wish to do so. After the testing is complete and questionnaires have been returned, the researcher will destroy the record of your child's identity and their responses will be anonymised (represented by a code number) where it will not be possible to identify him or her from their responses. At this stage, you will be unable to remove your child's responses as it will not be possible to identify his or her responses.

What will you and your child do in the project?

Your child will be invited to complete a series of short and simple paper and computer based tasks. Your child will be presented with a series of words and asked to point to the picture which best describes that word. In another task your child will be shown a series of patterns with one shape missing in each and will be asked to identify which shape would complete the pattern. Your child will take part in a simple picture task where pictures of the sun and moon will be presented on a sheet of paper and your child will be asked to name each image e.g. sun when they see the sun image and moon when they see the moon image. At some points in this task your child will be asked to respond by saying the opposite word, e.g. moon when they see a sun image and sun when they see a moon image. Your child will also be invited to complete a social behaviour task where they will be shown several images of males and females and will be asked to rate on a scale of 1-5 how much they would like to talk to the person. This task will provide a measure of your child's desire to approach unfamiliar people. Your child will listen to a series of short stories which will be read aloud to them and they will be asked to either identify the cause behind the event or to predict the outcome of the event from three response options. The stories will describe a boy or girl taking part in several different activities, such as going to the park, going to a restaurant for dinner and having a birthday party. Finally the researcher will invite your child to take part in a short informal interview where they will be asked about their friendships and their social activities. The total estimated completion time for this study will be 60-90 minutes, taking into consideration regular breaks.

Why have you been invited to take part?

Families who are registered with the Williams Syndrome Foundation and who have a child with Williams Syndrome aged between 6 and 16 years are being invited to take part.

What happens to the information in the project?

Your child's responses will initially be pseudo-anonymous (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from your responses). After your child's testing has been completed and questionnaires have been returned the researcher will make your child's responses anonymous by destroying information on his or her identity and he or she will not be identifiable in any way from their responses. All data will be stored in a locked cabinet within the University of Strathclyde and will only be accessible to the researchers on this study.

The University of Strathclyde is registered with the Information Commissioner's Office who implements the Data Protection Act 1998. All personal data on participants will be processed in accordance with the provisions of the Data Protection Act 1998.
Thank you for reading this information – please ask any questions if you are unsure about what is written here.

What happens next?

If you would like your child to take part in this project you will first be asked to complete a consent form. You will then be contacted regarding a suitable time for you to complete this study. This can be midweek or at the weekend and daytime or night time, depending on when is most suitable for you.

If you do not wish to take part thank you very much for your interest and for taking the time to read this information sheet.

This investigation was granted ethical approval by the University of Strathclyde Ethics Committee.

If you have any questions/concerns, during or after the investigation, or wish to contact an independent person to whom any questions may be directed or further information may be sought from, please contact:

Secretary to the University Ethics Committee
Research and Knowledge Exchange Services
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Appendix 13: Parent information sheet 2 (WS recruitment)

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

University of Strathclyde
Humanities & Social Sciences

Name of Department: School of Psychological Sciences and Health
Title of the study: Constructing a profile of social behaviour in Williams Syndrome

---

Introduction

My name is Amanda Gillooly and I am a postgraduate researcher at the University of Strathclyde, undertaking my PhD in psychology. I am working with Dr Sinead Rhodes and Professor Kevin Durkin within the School of Psychological Sciences and Health and Dr Deborah Riby from Durham University and this project is part of the North East Williams Syndrome research group. You are invited to take part in a research study. Before you decide, it is important for you to understand why the research is being carried out and what it will involve. Please take the time to read the following information sheet carefully. If you have any questions regarding this study please do not hesitate to contact either myself or my supervisors on the contact details below. I can be contacted via email at amanda.gillooly@strath.ac.uk.

What is the purpose of this investigation?

This study is being conducted to investigate peer relationships in children and adolescents with Williams Syndrome. Previous research has reported some individuals with Williams Syndrome to experience difficulties in forming and maintaining friendships. The present research aims to expand our understanding of this by using both questionnaire and interview methods to assess peer relationships across contexts. We hope that the results of this study will contribute to future interventions to help individuals develop peer relationships skills and raise awareness of this disorder.

Do you have to take part?

No, you don’t have to take part and participation in this study is entirely voluntary. You may withdraw from this study at any time without giving a reason. After your child’s testing has been completed and questionnaires have been returned, the researcher will destroy the record of you and your child’s identities and your responses will be anonymised (represented by a code number) where it will not be possible to identify you from your responses. At this stage, you will be unable to remove your responses as it will not be possible to identify individual participants’ responses.

What will you do in the project?

You will be asked to complete a few short questionnaires on your child's behaviour. These questionnaires will ask about your child's display of various social behaviours, their peer relationships and their social approach behaviour. You will also be invited to take part in an informal interview with the researcher, where you will be asked questions about your child's peer relationships and their social activities. The questionnaires should take approximately 30 minutes to complete in total and the interview should take approximately 20 minute.

Why have you been invited to take part?

Families who are registered with the Williams Syndrome Foundation and who have a child with Williams Syndrome aged between 6 and 16 years are being invited to take part.

What happens to the information in the project?

Your responses in this questionnaire will initially be pseudo-anonymous (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from your responses). After your child's testing has been completed and questionnaires have been returned, the researcher will make your responses anonymous by destroying information on your identity, where you will not be identifiable in any way from your responses. All data will be stored in a locked cabinet within the University of Strathclyde and will only be accessible to the researchers.

The University of Strathclyde is registered with the Information Commissioner's Office who implements the Data Protection Act 1998. All personal data on participants will be processed in accordance with the provisions of the Data Protection Act 1998.

Thank you for reading this information – please ask any questions if you are unsure about what is written here.

What happens next?

If you would like to take part in this project you will first be asked to complete a consent form. You will then be contacted regarding a suitable time for you and your child to complete this study.

If you do not wish to take part thank you very much for your interest and for taking the time to read this information sheet.

This investigation was granted ethical approval by the University of Strathclyde Ethics Committee.

If you have any questions/concerns, during or after the investigation, or wish to contact an independent person to whom any questions may be directed or further information may be sought from, please contact:

Secretary to the University Ethics Committee
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Dr Deborah Riby (Supervisor)
School of Psychology
Durham University
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Phone: 0191 33 43247

Appendix 14: Parent consent form 1 (WS recruitment)

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

University of Strathclyde Humanities & Social Sciences

Department: School of Psychological Sciences and Health

Title of study: Constructing a profile of social behaviour in Williams Syndrome

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- I confirm that I have read and understood the information sheet about the project and the researcher has answered any queries to my satisfaction.

- I understand that my child's participation in this project is voluntary and that my child is free to withdraw from the project at any time, without having to give a reason and without any consequences.

- I understand that my child's responses will initially be pseudo-anonymous (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from my child's responses). I understand that after my child's has completed his/her testing and all questionnaires have been returned, the document containing my child's identity will be destroyed and all of my child's responses will become fully anonymous (i.e. represented by a code name, where there is no possible way of identifying my child from their responses).

- I understand that my child's responses across these tasks will not be linked to information about my child provided in this consent form.

- I understand that after my child's testing has been completed and questionnaires have been returned I will be unable to remove my child's responses from this project as all responses will be made anonymous by the researcher and it will not be possible to identify the responses of individual participants.

- I understand that the researcher will not be able to provide me with any information on how my child has performed in this project.

- I consent to my child participating in this project.

I (PRINT NAME)	agree to take part in the above project
Signature of Participant:	Date

NAME (of child): ____________________________________

CHILD'S DATE OF BIRTH ____

- I give permission for audio recording equipment to be used to record my child's interview.

Yes ☐ No ☐

Appendix 15: Parent consent form 2 (WS recruitment)

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

Department: School of Psychological Sciences and Health
Title of study: Constructing a profile of social behaviour in Williams Syndrome

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- I confirm that I have read and understood the information sheet about the project and the researcher has answered any queries to my satisfaction.

- I understand that my participation is voluntary and that I am free to withdraw from the project at any time, without having to give a reason and without any consequences.

- I understand that my responses in this questionnaire will be pseudo-anonymous initially (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from my responses). I understand that after my child has completed his/her testing and all questionnaires have been returned, the document containing my identity will be destroyed and my responses in this questionnaire will become fully anonymous (i.e. represented by a code name, where there is no possible way that I can be identified from my responses).

- I understand that my responses in this questionnaire will not be linked to information about my child and I provided in this consent form.

- I understand that after my child's testing has been completed and questionnaires have been returned I will be unable to remove my questionnaire responses from this project as my responses will be made anonymous by the researcher and it will not be possible to identify individual participants' responses.

- I consent to being a participant in this project

I (PRINT NAME)	agree to take part in the above project
Signature of Participant:	Date

NAME (of child): ____________________

CHILD'S DATE OF BIRTH ____________________

- I give permission for audio recording equipment to be used to record my interview.

Yes ☐ No ☐

- I give permission for you to invite the teacher of my child to complete questionnaires on my child's social behaviour and peer relationships.

Yes ☐ No ☐

If yes:

Name of school ____________________

Class ____________________

Signature ____________________

Appendix 16: Child consent form (WS recruitment)

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

Department: School of Psychological Sciences and Health
Title of study: Constructing a profile of social behaviour in Williams syndrome

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My name is Amanda. I am trying to learn about what children do when they meet new people. I am also trying to learn about how well children can follow the rules of the games we will play and the way children understand stories. The final thing I am trying to learn about is what children like to do with their friends. If you would like, you can take part in some activities. You will be asked to play a picture game, read some stories and to solve different puzzles. In the picture game I will show you pictures of the sun and pictures of the moon. Sometimes when you see these pictures you will be asked to say the name aloud e.g. sun when you see a picture of a sun and moon when you see a picture of a moon. At other times you will be asked to say the opposite word, so you would say the word sun when you see a picture of a moon and moon when you see a picture of the sun. I will ask you to solve two types of puzzles in my picture books. In one game I will ask you to point to the picture which best describes the word I have said. In the other game I will show you pictures that have a missing shape and I will ask you to choose which shape would complete this picture. I will also read you some short stories which will describe a boy or girl taking part in different activities such as going to the park or having a birthday party and I will ask you to guess what will happen next in the story or to explain why something has happened in the story. In another activity you will see lots of pictures of different people and I will ask you to look at each picture and to tell me how much you would like to talk to that person. The last thing we will do is talk about what you like to do with your friends.

Nobody else will know what answers you have given and how you did in the activities as your results will be mixed in with the other children’s results.

Your parents/guardian have said that it’s OK for you to take part in these activities but if you don’t want to no one will be annoyed at you. If you want to take part in the activities now and change your mind later, that’s OK. You can stop at any time.

Would you like to take part in these activities?

YES/NO

(The appropriate response will be circled by the researcher)
THIS WILL BE READ ALOUD TO EACH CHILD BY THE RESEARCHER

Appendix 17: Teacher information sheet (WS recruitment)

University of Strathclyde
Humanities & Social Sciences

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

Teacher information sheet

Name of Department: School of Psychological Sciences and Health
Title of the study: Constructing a profile of social behaviour in Williams Syndrome

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Introduction

My name is Amanda Gillooly and I am a postgraduate researcher at the University of Strathclyde, undertaking my PhD in psychology. I am working with Dr Sinead Rhodes and Professor Kevin Durkin within the School of Psychological Sciences and Health and Dr Deborah Riby from Durham University and this project is part of the North East Williams Syndrome research group. You are invited to take part in a research study. Before you decide, it is important for you to understand why the research is being carried out and what it will involve. Please take the time to read the following information sheet carefully. If you have any questions regarding this study please do not hesitate to contact either myself or my supervisors on the contact details below.

What is the purpose of this investigation?

This study is being conducted to investigate peer relationships in children and adolescents with Williams Syndrome. Previous research has reported some individuals with Williams Syndrome to experience difficulties in forming and maintaining friendships. The present research aims to expand our understanding of this by assessing peer relationships across contexts. We hope that the results of this study will contribute to future interventions to help individuals develop peer relationships skills and further raise awareness of this disorder.

Do you have to take part?

No, you don't have to take part and participation in this study is entirely voluntary. You may withdraw from this study at any time without giving a reason. After the young person's testing has been completed and questionnaires have been returned, the researcher will destroy the record of your identity and your responses will be anonymised (represented by a code number) where it will not be possible to identify you from your responses. At this stage, you will be unable to remove your responses as it will not be possible to identify individual participants' responses.

What will you do in the project?

You will be asked to complete a few short questionnaires on the young person's behaviour. These questionnaires will ask about this young person's display of various social behaviours, their peer relationships and social inclusion.

Why have you been invited to take part?

You have been invited to take part in this study as you are the class teacher of a young person with Williams Syndrome aged between 6 and 16 years old and the young person's parent/guardian has provided permission for me to contact you regarding this.

What happens to the information in the project?

Your responses in this questionnaire will initially be pseudo-anonymous (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from your responses). After the young person's testing has been completed and all questionnaires have been returned the researcher will make your responses anonymous by destroying information on your identity, where you will not be identifiable in any way from your responses. All questionnaire responses will be stored in a locked cabinet within the University of Strathclyde and will only be accessible to the researchers.

The University of Strathclyde is registered with the Information Commissioner's Office who implements the Data Protection Act 1998. All personal data on participants will be processed in accordance with the provisions of the Data Protection Act 1998.
Thank you for reading this information – please ask any questions if you are unsure about what is written here.

What happens next?

If you would like to take part in this project you should complete the attached consent form and the questionnaires described above in a time of convenience and return these to me using the stamped addressed envelope provided.
If you do not wish to take part thank you very much for your interest and for taking the time to read this information sheet.

This investigation was granted ethical approval by the University of Strathclyde Ethics Committee.

If you have any questions/concerns, during or after the investigation, or wish to contact an independent person to whom any questions may be directed or further information may be sought from, please contact:

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Durham University
Email: deborah.riby@durham.ac.uk
Phone: 0191 33 43247

Appendix 18: Teacher consent form (WS recruitment)

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

University of Strathclyde Humanities & Social Sciences

Department: School of Psychological Sciences and Health
Title of study: Constructing a profile of social behaviour in Williams Syndrome

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- I confirm that I have read and understood the information sheet about the project and the researcher has answered any queries to my satisfaction.
- I understand that my participation is voluntary and that I am free to withdraw from the project at any time, without having to give a reason and without any consequences.
- I understand that my responses in this questionnaire will be pseudo-anonymous initially (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from my responses). I understand that after the child has completed the testing and all questionnaires have been returned, the document containing my identity will be destroyed and my responses in this questionnaire will become fully anonymous (i.e. represented by a code name, where there is no possible way that I can be identified from my responses).
- I understand that my responses in this questionnaire will not be linked to information about me provided in this consent form.
- I understand that after I return the questionnaires I will be unable to remove my questionnaire responses from this project as my responses will be made anonymous by the researcher and it will not be possible to identify individual participants' responses.
- I consent to being a participant in this project

I (PRINT NAME)	agree to take part in the above project
Signature of Participant:	Date

NAME (of child): ____________________

Appendix 19: Parent information sheet 1 (TD sample recruitment)

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

University of Strathclyde
Humanities & Social Sciences

Participant information sheet

Name of Department: School of Psychological Sciences and Health

Title of the study: The ability of young children to self-manage their behaviour, young children's thought processes and social behaviour towards new people.

Introduction

My name is Amanda Gillooly and I am a postgraduate researcher at the University of Strathclyde, undertaking my PhD in psychology. I am working with Dr Sinead Rhodes and Professor Kevin Durkin within the School of Psychological Sciences and Health. Your child is invited to take part in a research study. Before you decide, it is important for you to understand why the research is being carried out and what it will involve. Please take the time to read the following information sheet carefully. If you have any questions regarding this study please do not hesitate to contact either myself or my supervisors on the contact details below. I can be contacted via email at amanda.gillooly@strath.ac.uk or by telephone at 0141 548 4239.

What is the purpose of this investigation?

This study is being conducted to investigate the relationship between the way children manage their behaviour, their style of thinking and the way they approach and interact with other people. A child's ability to approach new people and engage in conversation with them will help them to make friends and become integrated into their peer group. When children show over-friendliness to other individuals it may cause concern among some parents that their child may approach strangers when they are out in public. This study aims to examine how children approach and interact with new people and to identify any factors which influence this behaviour.

Do you have to take part?

No, you don't have to take part and participation in this study is entirely voluntary. Your child may withdraw from this study at any time without giving a reason. You can withdraw your child and his or her responses from the study prior to the second testing session if you wish to do so. After both testing sessions have been completed the researcher will destroy the record of your child's identity and their responses will be anonymised (represented by a code number) where it will not be possible to identify him or her from their responses. At this stage, you will be unable to remove your child's responses as it will not be possible to identify his or her responses.

What will you and your child do in the project?

First of all you and the class teacher will be invited to complete a 5 minute multiple choice questionnaire which will ask questions about your child's behaviour in social situations and their interactions with other children. This questionnaire will be sent home to you via your child and we ask that you complete this as soon as is convenient and give this to your child to hand in at school. Your child will then be invited to complete a series of short and simple computer based and paper based tasks. Your child will be presented with a series of words and asked to point to the picture which best describes that word. In another task your child will be shown a series of patterns with one shape missing in each and will be asked to identify which shape would complete the pattern. Your child will take part in a simple computer task where pictures of the sun and pictures of the moon will appear on the screen and your child will be asked to respond by pressing the button with the same picture when they see each image. At some points in this task your child will be asked to respond by pressing the button with the opposite picture e.g. the sun in response to a picture of a moon and the moon in response to a picture of a sun. Your child will also be invited to complete a social behaviour task where they will be shown several images of males and females and will be asked to rate on a scale of 1-5 how much they would like to talk to the person. This task will provide a measure of your child's desire to approach new people. Finally your child will listen to a series of short stories which will be read aloud to them and they will be asked to either identify the cause behind the event or to predict the outcome of the event from three response options. The stories will describe a boy or girl taking part in several different activities, such as going to the park, going to a restaurant for dinner and having a birthday party. The total estimated completion time for this study will be 60 minutes which will be completed over two 30 minute sessions on different days to avoid your child becoming tired.

Why have you been invited to take part?

You have been invited to take part in this study due to your child being within the age range of interest (5-7 years old).

What happens to the information in the project?

Your child's responses will initially be pseudo-anonymous (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from your responses). After your child's testing has been completed the researcher will make your child's responses anonymous by destroying information on his or her identity and he or she will not be identifiable in any way from their responses. The questionnaires will be stored in a locked cabinet within the University of Strathclyde. The computerised data will be stored on a password protected computer within the University of Strathclyde and will only be accessible to the researchers on this study.

The University of Strathclyde is registered with the Information Commissioner's Office who implements the Data Protection Act 1998. All personal data on participants will be processed in accordance with the provisions of the Data Protection Act 1998.

Thank you for reading this information – please ask any questions if you are unsure about what is written here.

What happens next?
If you would like your child to take part in this project you will first be asked to complete a consent form. Your child will then be invited to complete a series of computer and paper based tasks as described above. This will take place within the school during school hours.

If you do not wish to take part thank you very much for your interest and for taking the time to read this information sheet.

This investigation was granted ethical approval by the School of Psychological Sciences and Health ethics committee.

If you have any questions/concerns, during or after the investigation, or wish to contact an independent person to whom any questions may be directed or further information may be sought from, please contact:

Dr James Baxter (Chair of Ethics Committee)
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Dr Sinead Rhodes (Chief Investigator/Supervisor)
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Phone : 0141 548 2574

Appendix 20: Participant information sheet 2 (TD sample recruitment)

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

Participant information sheet

Name of Department: School of Psychological Sciences and Health

Title of the study: The ability of young children to self-manage their behaviour, young children's thought processes and social behaviour towards new people.

Introduction

My name is Amanda Gillooly and I am a postgraduate researcher at the University of Strathclyde, undertaking my PhD in psychology. I am working with Dr Sinead Rhodes and Professor Kevin Durkin within the School of Psychological Sciences and Health. You are invited to take part in a research study. Before you decide, it is important for you to understand why the research is being carried out and what it will involve. Please take the time to read the following information sheet carefully. If you have any questions regarding this study please do not hesitate to contact either myself or my supervisors on the contact details below. I can be contacted via email at amanda.gillooly@strath.ac.uk or by telephone at 0141 548 4239.

What is the purpose of this investigation?

This study is being conducted to investigate the relationship between the way children manage their behaviour, their style of thinking and the way they approach and interact with other people. A child's ability to approach new people and engage in conversation with them will help them to make friends and become integrated into their peer group. When children show over-friendliness to other individuals it may cause concern among some parents that their child may approach strangers when they are out in public. This study aims to examine how children approach and interact with new people and to identify any factors which influence this behaviour.

Do you have to take part?

No, you don't have to take part and participation in this study is entirely voluntary. You may withdraw from this study at any time without giving a reason. After your child's testing has been completed, the researcher will destroy the record of you and your child's identities and your responses will be anonymised (represented by a code number) where it will not be possible to identify you from your responses. At this stage, you will be unable to remove your responses as it will not be possible to identify individual participants' responses.

What will you do in the project?

You will be asked to complete one short questionnaire on your child's behaviour. This questionnaire will ask questions about your child's display of various social behaviours and their interactions with other children. You will be asked to identify which of the options shown you consider to apply to your child. This questionnaire will take approximately 5 minutes to complete. If you consent to participate, we will ask you to complete this questionnaire in the nearest time of convenience and give this to your child to hand in at school. All children, regardless of their score in this measure, will be invited to complete the study and their results will be included in the final dataset. The researcher will use the responses you provide to organise the participants datasets into sub-groups for analysis purposes e.g. those who scored high in hyperactivity symptoms and those who scored low in hyperactivity symptoms. The symptoms measured in this questionnaire are frequently observed in children and will not represent a diagnosis for your child.

Why have you been invited to take part?

You have been invited to take part in this study as you the parent/guardian to a child between the ages of 5-7 years (the age group of interest).

What happens to the information in the project?

Your responses in this questionnaire will initially be pseudo-anonymous (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from your responses). After your child's testing has been completed the researcher will make your responses anonymous by destroying information on your identity, where you will not be identifiable in any way from your responses. The questionnaires will be stored in a locked cabinet within the University of Strathclyde. The computerised data will be stored on a password protected computer within the University of Strathclyde and will only be accessible to the researchers.

The University of Strathclyde is registered with the Information Commissioner's Office who implements the Data Protection Act 1998. All personal data on participants will be processed in accordance with the provisions of the Data Protection Act 1998.

Thank you for reading this information – please ask any questions if you are unsure about what is written here.

What happens next?

If you would like to take part in this project you will be given a consent form to complete. You will then be sent a questionnaire to complete as discussed above. We ask if you can complete this at a time of your nearest convenience and give this to your child to hand in to the school.

If you do not wish to take part thank you very much for your interest and for taking the time to read this information sheet.

This investigation was granted ethical approval by the School of Psychological Sciences and Health ethics committee.

If you have any questions/concerns, during or after the investigation, or wish to contact an independent person to whom any questions may be directed or further information may be sought from, please contact:

Dr James Baxter (Chair of Ethics Committee)
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Phone : 0141 548 2574

Appendix 21: Parent consent form 1 (TD sample recruitment)

University of Strathclyde
Humanities & Social Sciences

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

Department: School of Psychological Sciences and Health

Title of study: The ability of young children to self-manage their behaviour, young children's thought processes and social behaviour towards new people.

☐ I confirm that I have read and understood the information sheet about the project and the researcher has answered any queries to my satisfaction.

- I understand that my child's participation in this project is voluntary and that my child is free to withdraw from the project at any time, without having to give a reason and without any consequences.

- I understand that my child's responses in the first testing session will be pseudo-anonymous (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from my child's responses). I understand that after both testing sessions have been completed, the document containing my child's identity will be destroyed and all of my child's responses will become fully anonymous (i.e. represented by a code name, where there is no possible way of identifying my child from their responses).

- I understand that my child's responses across these tasks will not be linked to information about my child provided in this consent form.

- I understand that after my child's testing has been completed I will be unable to remove my child's responses from this project as all responses will be made anonymous by the researcher and it will not be possible to identify the responses of individual participants.

☐ I understand that the researcher will not be able to provide me with any information on how my child has performed in this project.

- I give permission for you to ask the class teacher of my child to complete a questionnaire about my child's behaviour.

☐ I consent to my child participating in this project.

I (PRINT NAME)	agree to take part in the above project
Signature of Participant:	Date

NAME (of child): ____

CHILD'S DATE OF BIRTH____

University of Strathclyde
Humanities & Social Sciences

Appendix 22: Parent consent form 2 (TD sample recruitment)

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

Department: School of Psychological Sciences and Health

Title of study: The ability of young children to self-manage their behaviour, young children’s thought processes and social behaviour towards new people.

- ☐ I confirm that I have read and understood the information sheet about the project and the researcher has answered any queries to my satisfaction.

- ☐ I understand that my participation is voluntary and that I am free to withdraw from the project at any time, without having to give a reason and without any consequences.

- ☐ I understand that my responses in this questionnaire will be pseudo-anonymous initially (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from my responses). I understand that after my child has completed both testing sessions, the document containing my identity will be destroyed and my responses in this questionnaire will become fully anonymous (i.e. represented by a code name, where there is no possible way that I can be identified from my responses).

- ☐ I understand that my responses in this questionnaire will not be linked to information about my child and I provided in this consent form.

- I understand that after my child’s testing has been completed I will be unable to remove my questionnaire responses from this project as my responses will be made anonymous by the researcher and it will not be possible to identify individual participants’ responses.

- ☐ I consent to being a participant in this project

I (PRINT NAME)	agree to take part in the above project
Signature of Participant:	Date

NAME (of child): ______________________________

CHILD’S DATE OF BIRTH ______________________________

Appendix 23: Child consent form (TD sample recruitment)

University of Strathclyde
Humanities & Social Sciences

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

Department: School of Psychological Sciences and Health

Title of study: The ability of young children to self-manage their behaviour, young children's thought processes and social behaviour towards new people.

My name is Amanda. I am trying to learn about what children do when they meet new people. I am also trying to learn about how well children can follow the rules of the games we will play. The final thing I am trying to learn about is the way children understand stories. If you would like, you can take part in some activities. You will be asked to play a computer game and you will be asked to solve different puzzles in my picture books. The computer game you will be asked to play will show you pictures of the sun and pictures of the moon. Sometimes when you see these pictures you will be asked to press the button with the same picture on it, so you would press the sun button when you see a picture of a sun and you would press the moon button when you see a picture of a moon. At other times you will be asked to press the button with the opposite picture on it, so you would press the sun button when you see a picture of a moon and the moon button when you see a picture of a sun. I will ask you to solve two types of puzzles in my picture books. In one game I will ask you to point to the picture which best describes the word I have said. In the other game I will show you pictures that have a missing shape and I will ask you to choose which shape would complete this picture. I will also read you some short stories which will describe a boy or girl taking part in different activities such as going to the park or having a birthday party and I will ask you to guess what will happen next in the story or to explain why something has happened in the story. Last of all you will see lots of pictures of different people and I will ask you to look at each picture and to tell me how much you would like to talk to that person.

Nobody else will know what answers you have given and how you did in the activities as your results will be mixed in with the other children's results.

Your parents/guardian have said that it's OK for you to take part in these activities but if you don't want to no one will be annoyed at you. If you want to take part in the activities now and change your mind later, that's OK. You can stop at any time.

Would you like to take part in these activities?

YES/NO

(The appropriate response will be circled by the researcher)

THIS WILL BE READ ALOUD TO EACH CHILD BY THE RESEARCHER

Appendix 24: Teacher information sheet (TD sample recruitment)

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

Participant information sheet

Name of Department: School of Psychological Sciences and Health

Title of the study: The ability of young children to self-manage their behaviour, young children's thought processes and social behaviour towards new people.

Introduction

My name is Amanda Gillooly and I am a postgraduate researcher at the University of Strathclyde, undertaking my PhD in psychology. I am working with Dr Sinead Rhodes and Professor Kevin Durkin within the School of Psychological Sciences and Health. You are invited to take part in a research study. Before you decide, it is important for you to understand why the research is being carried out and what it will involve. Please take the time to read the following information sheet carefully. If you have any questions regarding this study please do not hesitate to contact either myself or my supervisors on the contact details below. I can be contacted via email at amanda.gillooly@strath.ac.uk or by telephone at 0141 548 4239.

What is the purpose of this investigation?

This study is being conducted to investigate the relationship between the way children manage their behaviour, their style of thinking and the way they approach and interact with other people. A child's ability to approach new people and engage in conversation with them will help them to make friends and become integrated into their peer group. When children show over-friendliness to other individuals it may cause concern among some parents that their child may approach strangers when they are out in public. This study aims to examine how children approach and interact with new people and to identify any factors which influence this behaviour.

Do you have to take part?

No, you don't have to take part and participation in this study is entirely voluntary. You may withdraw from this study at any time without giving a reason. After the child's testing has been completed, the researcher will destroy the record of your identity and your responses will be anonymised (represented by a code number) where it will not be possible to identify you from your responses. At this stage, you will be unable to remove your responses as it will not be possible to identify individual participants' responses.

What will you do in the project?

You will be asked to complete one short questionnaire on each child in your class who is participating in this study. Each questionnaire will take approximately 5 minutes to complete. If you consent to participate, you should complete this questionnaire in the nearest time of convenience and return this to the school office. This questionnaire will contain questions about the behaviour of the child within the school environment and their interactions and behaviours with other children. All children, regardless of their score in this measure, will be invited to complete the study and their results will be included in the final dataset. The researcher will use the responses you provide to organise the participants datasets into sub-groups for analysis purposes e.g. those who scored high in hyperactivity symptoms and those who scored low in hyperactivity symptoms. The symptoms measured in this questionnaire are frequently observed in children and will not represent a diagnosis for your child.

Why have you been invited to take part?

You have been invited to take part in this study as you are the class teacher of the child participating in this study.

What happens to the information in the project?

Your responses in this questionnaire will initially be pseudo-anonymous (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from your responses). After the child's testing has been completed the researcher will make your responses anonymous by destroying information on your identity, where you will not be identifiable in any way from your responses. The questionnaires will be stored in a locked cabinet within the University of Strathclyde. The computerised data will be stored on a password protected computer within the University of Strathclyde and will only be accessible to the researchers.

The University of Strathclyde is registered with the Information Commissioner's Office who implements the Data Protection Act 1998. All personal data on participants will be processed in accordance with the provisions of the Data Protection Act 1998.

Thank you for reading this information – please ask any questions if you are unsure about what is written here.

What happens next?

If you would like to take part in this project you will be given a questionnaire to complete for each child in your class who is participating in this study. We ask if you are able to complete these at a time which is convenient to you and return these to either myself or the school office.

If you do not wish to take part thank you very much for your interest and for taking the time to read this information sheet.

This investigation was granted ethical approval by the School of Psychological Sciences and Health ethics committee.

If you have any questions/concerns, during or after the investigation, or wish to contact an independent person to whom any questions may be directed or further information may be sought from, please contact:

Dr James Baxter (Chair of Ethics Committee)
School of Psychological Sciences and Health
University of Strathclyde
Graham Hills Building
40 George Street
Glasgow G1 1QE
Email : j.baxter@strath.ac.uk
Phone : 0141 548 2242

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Appendix 25: Teacher consent form (TD sample recruitment)

SCHOOL OF PSYCHOLOGICAL SCIENCES & HEALTH

University of Strathclyde
Humanities & Social Sciences

Department: School of Psychological Sciences and Health

Title of study: The ability of young children to self-manage their behaviour, young children's thought processes and social behaviour towards new people.

- I confirm that I have read and understood the information sheet about the project and the researcher has answered any queries to my satisfaction.

- I understand that my participation is voluntary and that I am free to withdraw from the project at any time, without having to give a reason and without any consequences.

- I understand that my responses in this questionnaire will be pseudo-anonymous initially (i.e. responses will be anonymised and given a code name, with the key for that code name stored in a secure document which will be kept separate from my responses). I understand that after the child has completed both testing sessions, the document containing my identity will be destroyed and my responses in this questionnaire will become fully anonymous (i.e. represented by a code name, where there is no possible way that I can be identified from my responses).

- I understand that my responses in this questionnaire will not be linked to information about me provided in this consent form.

- I understand that after the child's testing has been completed I will be unable to remove my questionnaire responses from this project as my responses will be made anonymous by the researcher and it will not be possible to identify individual participants' responses.

- I consent to being a participant in this project

I (PRINT NAME)	agree to take part in the above project
Signature of Participant:	Date

NAME (of child): ______________________________