Social vulnerability in Williams syndrome

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Thesis abstract

This thesis focused on the high levels of social vulnerability experienced by individuals with Williams Syndrome (WS). The investigation began with parent interviews about social approach behaviour, with parents emphasising the lack of awareness of social boundaries that many individuals with WS display. The qualitative analysis also highlighted the within-syndrome variability in the parental accounts, prompting discussion on the heterogeneity of the WS social profile. Based on the atypical social approach behaviour described by parents, the subsequent studies addressed issues of personal space and interpersonal distance. Using a parent report questionnaire, it was found that children with WS were more likely to violate the personal space of others. This was followed up with a stop-distance paradigm which showed that children with WS failed to regulate their distance based on familiarity, and stood the same distance from a stranger as they did their parent, which was not the case for typically developing individuals. Given these findings, the research progressed to explore the issue of trust in WS. It was found that children with WS displayed higher levels of trust behaviour, compared to their mental age matched typically developing peers and struggled to decipher trustworthiness from faces. Taken together, these findings seem to suggest that children with WS could be experiencing high levels of social vulnerability on a daily basis.

It is widely accepted that this social vulnerability continues into adulthood, with increased levels of both independence and isolation posing a new set of challenges. The subsequent chapter probed the level of insight that adults with

WS had about their own vulnerability. Using the Social Vulnerability Questionnaire, it was found that adults with WS consistently reported lower levels of vulnerability, compared to parent reports. This emphasised the need for multi-informant methods, and called for interventions which target self-awareness in order to increase intervention efficacy.

The final chapters looked at how this social vulnerability manifests in the online environment. It was found that adults with WS frequently use the Internet and the majority visit social networking sites every day or almost every day, with little parental supervision or oversight. These individuals were more likely to agree to engage in socially risky behaviours (e.g. meeting an "online friend" in person) compared to risky behaviours that were not social in nature when online (e.g. giving out passwords). A case study interview with an adult with WS and their parent highlighted that this individual held a broad definition of what a friend was and found they used the Internet as a tool to expand their social network, which was of great concern to their parent.

The findings included in this thesis provide in-depth information relating to social vulnerability in WS and offer the first insights into online social behaviour and online vulnerability in adults with WS. The theoretical and real-world implications of these findings are emphasised throughout and a number of suggestions are made to help the research progress towards intervention development.

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Declaration

The following chapters feature published journal articles. The contributions of each author are outlined below. All authors read and approved the final manuscripts.

Chapter	Publication featured	Author contribution
Chapter 4	Lough, E., Rodgers, J., Janes, E., Little, K., Riby, D. M. (2016). Parent insights into atypicalities of social approach behaviour in Williams syndrome. *Journal of Intellectual Disability Research*. doi: 10.1111/jir.12279	EL: conceived of the study, performed the statistical analyses, interpreted the data and drafted the manuscript JR: conceived of the study and participated in the design and coordination of the data EJ: conceived of the study and participated in the design and coordination of the data DR: conceived of the study, participated in the design and coordination of the data and helped draft the manuscript.

Chapter 5	Lough, E., Hanley, M., Rodgers, J., South, M., Kirk, H., Kennedy, D. & Riby, D. M. (2015). Violations of Personal Space in Young People with Autism Spectrum Disorders and Williams Syndrome: Insights from the Social Responsiveness Scale. *Journal of Autism and Developmental Disorders. doi:* 10.1007/s10803-015-2536-0	EL: conceived of the study, participated in the design and coordination of the data, performed the measurement, performed the statistical analyses, interpreted the data and drafted the manuscript MH, JR, MS, HK: performed the measurement and critically revised the manuscript DK: conceived of the study, participated in the design of the study and critically revised the manuscript DR: conceived of the study, participated in the design and coordination of the study, performed the measurement, interpreted the data and drafted the manuscript.
	Lough, E., Flynn, E. G. & Riby, D. M. (2016). Personal space regulation in Williams syndrome: The effect of familiarity. *Journal of Autism and Developmental Disorders. .doi:* 10.1007/s10803-016-2864-8	EL: conceived of the study, participated in its design and coordination, performed the measurement, performed the statistical analysis and drafted the manuscript

		EF: participated in the study design and helped draft the manuscript. DM: conceived of the study, participated in its design and coordination, helped interpret the data and draft the manuscript.
Chapter 7	Lough, E. & Fisher, M. H. (2016). Parent and Self-Report Ratings on the Perceived Levels of Social Vulnerability of Adults with Williams Syndrome. *Journal of Autism and Developmental Disorders. doi:* 10.1007/s10803-016-2885-3	EL: conceived of the study, participated in its design and coordination, performed the measurement, performed the statistical analysis and drafted the manuscript MF: Performed the measurement, aided with the statistical analysis and helped draft the manuscript.
Chapter 8	Lough, E., Flynn, E. & Riby, D. M. (2015). Mapping real-world to online vulnerability in young people with developmental disorders: Illustrations from autism and Williams syndrome. *Review Journal of Autism and Developmental Disorders* 2(1): 1-7.	EL: conceived of the review and drafted the manuscript. EF: helped draft the manuscript DR: conceived of the review and drafted the manuscript.

Chapter 9	Lough, E. & Fisher, M.H. (2016). Internet use and online safety in adults with Williams syndrome. *Journal of Intellectual Disability Research. doi:* 10.1111/jir.12281	EL: conceived of the study, participated in its design and coordination, performed the measurement, performed the statistical analysis and drafted the manuscript MF: conceived of the study, participated in its design and coordination and helped draft the manuscript.

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Chapter One: General Introduction

1.1 Vulnerability

The concept of vulnerability is challenging to define (Larkin, 2009). It can hold a variety of meanings, depending on the context and the population group that it is being applied to (e.g. Pritchard, 2001). Vulnerability can mean that a person is in danger, under threat, at high risk, highly susceptible to problems, in need of support and/or helpless (Simpson, 2006; Grundy, 2006; Mawby, 2004). According to the Department of Health (2000), an adult is considered to be vulnerable if they are unable to care for themselves. But within this generic definition, there exists different types of vulnerability (Rodgers, 1997; Goldson, 2002; Dixon-Woods et al., 2005). For example, innate or person vulnerability refers to traits or characteristics specific to the person concerned which make them vulnerable (Larkin, 2009). Whereas, contextual vulnerability refers to the effect that a set of circumstances has on making a person vulnerable (Goldson, 2002). Other terms encompass both innate and contextual vulnerability definitions. For instance, social vulnerability refers to, "the disadvantages faced by an individual while he or she endeavours to survive as a productive member of the society" (Jawaid et al., 2012, p335). It is this latter form of vulnerability that is of particular interest to the current thesis.

Defining vulnerable groups can also be challenging. The Safeguarding Vulnerable Groups Bill (House of Commons, 2006) defined a vulnerable adult as receiving support from health and social care services, living in sheltered

housing, in need of support to conduct daily affairs, involved with the probation service, or detained under immigration legislation. People who fall into these groups are typically marginalised from society, have limited employment opportunities and thus income and suffer abuse and/ or prejudice (Rodgers, 1997). Some examples of vulnerable groups include older people, children, ethnic minorities, people who have a mental illness, refugees and people with disabilities (Shelter, 2007). Vulnerability is clearly a far-reaching term, however, group members tend to be heterogeneous, and often hold multiple identities (White, 2002). For example, someone with a disability may also experience mental health problems. This can make identification challenging.

People with disabilities are considered to be a vulnerable group (Larkin, 2009). It is estimated that 21% of adults in the UK have a physical or mental disability (Disability Rights Commission, 2006). They face high rates of unemployment, as employers associate disabilities with risk and uncertainty (Heenan, 2002). Even those in employment are reported to experience elevated levels of discrimination, owing to subtle prejudice beliefs (Foster et al., 2006; Morris, 2004; Deal, 2007). Given the high rates of unemployment in this group, it is perhaps unsurprising that this, combined with health problems and low levels of educational attainment, results in low levels of income; it is estimated that 45% of adults with some form of disability live in poverty (Bambra, et al., 2005). However, grouping people together under the umbrella term of people who have ‘disabilities’ ignores the highly heterogeneous nature of this group, as the above information certainly does not apply to everyone in this group. Indeed, there are considerable differences in experiences between people with physical disabilities, and those

with mental disabilities. It is the vulnerability experienced by people who have neurodevelopmental disorders / intellectual and developmental disabilities (IDD) that will be explored further in this thesis (for definitions of the terminology used in this thesis, see Chapter 2, section 2.2).

Individuals with IDD experience high levels of vulnerability throughout the developmental trajectory (Fisher, Baird, Currey & Hodapp, 2016). Sullivan and Knutson (2000) found that children with IDD were more than three times as likely to experience abuse and neglect compared to children without IDD (31% instance rate versus 9% instance rate). They also noted that of these children who had experienced abuse, most had endured multiple forms of maltreatment. During their school years, rates of bullying and peer victimisation are high for individuals with IDD compared to their typically developing peers (e.g. Sentenac, Gavin, Arnaud, Molcho, Godeau, & Gabhainn, 2013). In adulthood, people with IDD are more likely to experience offences against their property, such as robbery or theft, and against their person, such as physical or sexual assault compared to their typically developing peers, particularly if they are living independently (Wilson & Brewer, 1992).

People with Autism Spectrum Disorder (ASD) are susceptible to experiencing high rates of vulnerability. ASD is a collection of pervasive developmental disorders characterised by deficits in social communication and restricted, repetitive patterns of behaviour which are present in the early developmental period (DSM-5; American Psychiatric Association, 2013). Individuals with ASD show communicative and socio-cognitive impairments, and experience high

levels of social isolation (van Roekel et al., 2010). This can place them at risk of peer victimisation (Cappadocia, Weiss, & Pepler, 2012). Rowley et al. (2012) looked at experiences of friendship, victimisation and bullying in 100 children (mean age = 11.4 years) with an ASD diagnosis. They gathered data from parents, teachers and the children themselves (see Chapter 2, section 2.4 for a discussion on the benefits of adopting a multi-informant approach) using the Strengths and Difficulties Questionnaire (SDQ-P, SDQ-T; Goodman, 1997) and the Autism Diagnostic Observation Schedule-Generic Module 3 (ADOS-G; Lord et al., 2000). The authors found that almost all children reported experiencing disagreements with other children and this resulted in feelings of rejection and isolation in 40% of children interviewed. Both parent and teacher reports showed higher levels of victimisation experienced by children with ASD compared to children without special educational needs. Children with ASD therefore offer an example of a group of people with an IDD who are vulnerable to victimisation.

Down syndrome (DS), a genetic disorder resulting from an extra copy of chromosome 21 (typically full trisomy 21), offers another example of an IDD which is linked to high levels of social vulnerability (Fisher et al., 2013). Individuals with DS have moderate levels of intellectual impairment (Gibson, 1978) and are highly social and engaging (Dykens, 2000). Fisher et al. (2013) compared social vulnerability in adults with different types of IDD using the Social Vulnerability Questionnaire (SVQ). They found that individuals with DS had low levels of risk awareness and were perceived by others to be highly vulnerable, which was contributing to their high levels of social vulnerability. Whereas, for individuals with ASD, they had less social protection, in addition to

low levels of risk awareness, which contributed to their vulnerability status. The authors also looked at social vulnerability in a third IDD: Williams syndrome (WS), which will be the focus of this thesis and will therefore be explored in more detail (see section 1.4.4). The findings of Fisher et al. (2013) suggested that the elements which contribute to social vulnerability are likely to be syndrome specific, and further in-depth investigation is needed within syndromes in order to better understand their experience of social vulnerability.

1.2 Williams syndrome

Williams syndrome is a rare neurodevelopmental disorder, which was first identified the 1960s by two independent teams of physicians. Williams, Barratt-Boyes and Lowe (1961) in New Zealand and Beuren, Apitz and Harmjanz (1962) in Germany both described a small group of patients who all showed a distinct cardiovascular problem (supravalvular aortic stenosis – a narrowing of the arteries which results in reduced blood flow), learning difficulties and unique and similar facial features. Williams, Barratt-Boyes and Lowe (1961) proposed that together, these characteristics were indicators of a previously unknown condition. The terms Williams-Beuren syndrome, or more commonly Williams syndrome (WS) were used to describe the condition. Most of the early work on WS focused on the medical profile of the syndrome, such as the supravalvular aortic stenosis and infantile hypercalcemia (elevated blood calcium levels), and during this time, it was medical researchers who were leading the work on WS (e.g. Von Armin & Engel, 1964).

The genetic aetiology of the syndrome during this early period remained unknown, and diagnosis was phenotypical using the triad of supravalvular aortic stenosis, learning difficulties and distinct facial features. However, in the 1990s, several key discoveries were made. The first came from Morris, Thomas and Greenberg (1993) who found that several families showed an autosomal dominant pattern, i.e. there was a 50% chance that parents would transfer the syndrome to their children. Typically, the presence of WS in these parents was undetected until their child received a diagnosis. The second offering came from Morris, Locker, Ensing and Stock (1993) who profiled a family who had supravalvular aortic stenosis caused by a translocation of chromosome 7, which subsequently disrupted the gene for elastin (ELN; a connective tissue protein that provides strength and elasticity to the skin, blood vessels and the walls of organs and arteries). Supravalvular aortic stenosis had long been established as a typical feature of WS, so this disruption in ELN offered a possible etiological hypothesis for the genetic cause of WS (Tassabehji et al., 1999). Support from this hypothesis came from the third key finding of this year; Ewart et al. (1993) found that individuals with WS had a microdeletion on the long arm of one of their chromosome 7s (7q11.23), which included the ELN gene. Subsequent studies have identified that the ELN deletion has been found in almost all individuals with WS (e.g. Lowery et al., 1995 found the deletion in 97% of individuals with WS).

Following the important discoveries in the 1990s, fluorescent in situ hybridization (FISH) testing is now used in the diagnosis of WS. FISH testing detects the deletion of one copy of the elastin gene on chromosome 7, as well as the

hemizygous deletion of approximately 26 – 28 genes on chromosome 7q11.23 (Eisenberg, Jabbi & Berman, 2010; Morris & Mervis, 2000). This is now used as a genetic marker of WS and has resulted in children being more accurately diagnosed, and at a younger age. In addition to the physical heart abnormalities which are typically the first sign of WS, early indicators are known to include infantile hypercalcemia, low birth weight and/or slow weight gain, poor feeding and difficulties sleeping (Sarimski, 1996). Many individuals with WS also experience delays reaching early developmental milestones (Mervis & Velleman, 2011). Prevalence rates for WS range between 1 in 7,500 (Strømme, Bjørnstad & Ramstad, 2002) to 1 in 20,000 births (Morris, Demsey, Leonard, Dilts, & Blackburn, 1988), with the Williams syndrome Foundation placing the current (2016) figure at 1 in 18,000 in the UK. It occurs sporadically in the general population, but as outlined above, there is a 50% chance that an individual with WS could transmit the syndrome to their children. As outlined above, WS is associated with medical and physical atypicalities, however, there is also a unique psychological profile associated with the syndrome, and it is this psychological profile that is most relevant to the current thesis. Now, we are going to focus on aspects of the psychological profile that are relevant to this thesis, starting with the cognitive profile associated with WS. The next section outlines the 'average' cognitive profile seen in WS, although it is acknowledged that there are vast within-syndrome differences (which will be addressed in section 1.3.3).

1.3 Cognitive profile

In the 1980s and 1990s the disorder really captured the attention of cognitive scientists. Traditionally, language acquisition had been seen as directly based on

cognitive development (Torff & Gardner, 1999). However, the cognitive profile associated with WS challenged this notion, proposing that language was modular and developed to some extent independently to cognition (Levy & Herman, 2003). Bellugi, Sabo and Vaid (1988) studied three young people with Williams syndrome (age range = 11 to 16 years). They noted that, despite delayed onset of language development, the linguistic abilities shown by the participants far exceed what would be expected based on their level of cognitive functioning (IQ scores ranged from 43 to 66). The authors described the linguistic abilities of the participants as, "complex, usually grammatically well-formed" and noted that they showed "a great range of grammatical structures with complex embeddings, as well as unusual vocabulary" (p.281), based on their performance on tests such as the Test for Reception of Grammar (Bishop, 1983). This was used as support for their claim of autonomy of systems, dissociation between language abilities and other cognitive functions and also the notion that linguistic abilities are spared in WS. This prompted a great deal of research focused on the cognitive profile of the disorder, not only to understand WS, but also to understand the human mind.

Individuals with WS typically have mild to moderate levels of intellectual impairments, and have a mean IQ of 50 – 60 (e.g. Martens, Wilson & Reutens, 2008; Udwin, Yule & Martin, 1987, Davies, Howlin & Udwin, 1997). Few studies have examined the cognitive trajectories of global IQ scores. Udwin, Davies and Howlin (1996) found an increase in IQ between the first testing point (mean age = 12 years) to the second testing point (mean age = 21 years; though note the change of measures from the Wechsler Intelligence Scale for Children –

Revised (Wechsler, 1974) at time point one to the Wechsler Adult Intelligence Scale – Revised at time point two; (Wechsler, 1981). Conversely, Crisco (1990) reported stability in IQ scores in participants who were first tested aged 4 (mean IQ score of 67 on the Stanford-Binet Form LM; Terman & Merrill, 1960) and then again at age 9 (mean IQ score of 66 on the Stanford-Binet Form LM; Terman & Merrill, 1960). The different measurement tools used in these studies are likely to contribute to the reported difference, particularly given the paradoxical profile associated with WS (Mervis et al., 2000; Karmiloff-Smith et al., 1997). Overall IQ scores mask the intriguing profile of relative cognitive strengths and weaknesses seen in WS (Martens et al., 2008; see sections 1.3.1, and 1.3.2), as well as the extreme within-syndrome heterogeneity (Porter and Coltheart, 2005; see section 1.3.3).

1.3.1 Relative strengths in the WS cognitive profile

Early research into the cognitive profile of WS referred to 'impaired' and 'intact' cognitive functions (e.g. Bellugi, Lichtenberger, Mills, Galaburda & Korenberg, 1999; Pinker, 1999). As alluded to earlier, one ability which was initially described as being 'spared' was language (e.g. Bellugi, Sabo & Vaid, 1988). However, more recent research has dismissed the idea that individuals with WS have an intact language module (Karmiloff-Smith, Brown, Grice & Paterson, 2003). Instead, it has been suggested that language represents a relative strength in WS, though it is still not comparable to the levels seen in mentally age matched peers (e.g. Jarrold, Baddeley & Hewes, 1998). For example, Mervis, Morris, Bertrand and Robinson (1999) examined the lexicons of 123 children and adults with WS. They found that 42% scored in the borderline to typical range on the

Peabody Picture Vocabulary Test – Revised (Dunn & Dunn, 1981). However, they also noted considerable within-syndrome heterogeneity and the group as a whole showed mild levels of delay. This reinforces the idea that whilst language might be a relative strength, it still is not at the level expected for their chronological age.

Bellugi, Lichtenberger, Jones and Lai (2000) investigated the complex pattern of neurocognitive strengths and weaknesses seen in WS. Comparisons were drawn with individuals with DS, who, like WS, show impaired levels of overall cognitive functioning, but unlike WS, show severe impairments in linguistic ability (e.g. Rice, Warren & Betz, 2005). They found that adolescents and adults with WS showed relatively strong grammatical abilities compared to individuals with DS who performed at chance on a task of comprehension of passive sentences. This pattern was repeated on the Test for Reception of Grammar (TROG; Bishop, 1982), the Kempler Test of Syntax (Kempler & VanLancker, 1993), the Curtiss-Yamada Comprehensive Language Evaluation (CYCLE; Curtis & Yamada, 1988) and on the Clinical Evaluation of Language Fundamentals (CELF; Semel, Wiig & Secord, 1987). They also noted that individuals with WS were able to correctly respond to conditional questions ("What would you do if…") 83% of the time, whereas participants with DS only responded correctly 23% of the time. This suggests that individuals with WS have relative strengths in syntax and morphosyntactic rules (Bellugi, Jernigan, Trauner & Doherty, 1990).

However, this claim has come under scrutiny, with others finding that grammatical comprehension is actually impaired in WS. In contrast to the findings of Bellugi et al. (2000) which showed better performance on the TROG (Bishop, 1982) for individuals with WS compared to those with DS, Karmiloff-Smith et al. (1997) and Volterra, Capirci, Pezzini, Sabbadini and Vicari (1996) found that individuals with WS scored below the levels expected for their chronological age and their mental age (see Chapter 2, section 2.6 for a discussion on the role of matched groups design). Karmiloff-Smith et al. (1998) asked participants (mean age = 20.7 years) to complete a sentence-picture matching task, in which a sentence was read aloud and they had to select which picture best represented the sentence. As well as the correct option, there were also options which portrayed a lexical distractor and a syntactic distractor. It was found that the performance of the chronological aged matched control group was largely error free, however, the WS group performed poorly, making an average of 24% errors. Of these errors, 81% were syntactic errors, where the participant chose the reverse role distractor picture. The authors acknowledged that the measures used influence the conclusions that are drawn about language ability, as participants with WS performed relatively well at single word picture matching tasks, such as the British Picture Vocabulary Scale (BPVS, Dunn & Dunn, 1982; see Chapter 2 for a full description of this measure). They also suggested dissociation between representational and integrational impairments in WS syntax.

It has been found that language follows an atypical developmental trajectory in WS (e.g. Klein & Mervis, 1999) and there are severe deficits in some areas of

linguistic ability (e.g. Laing et al., 2002; Karmiloff-Smith, Klima, Bellugi, Grant & Baron-Cohen, 1995). Laws and Bishop (2004) used the Children's Communication Checklist (CCC; Bishop, 1998) to investigate pragmatic language abilities of individuals with WS (mean age = 14.1 years), DS, specific language impairment (SLI) and a group of typically developing children. The CCC has five subscales: appropriateness of the initiation of communication; coherence of conversation; tendency to use stereotyped conversation; use of conversational context; and conversational rapport. They found that all three clinical groups scored significantly lower on the CCC compared to the typically developing control group, however, it was only the individuals with WS who scored below the cut-off indicative of impairment (cut-off score: 132). The WS group scored significantly worse than their TD peers on all of the five subscales, but it was the subscales of inappropriate initiation of conversation and the use of stereotyped conversation which were especially impaired, with the WS group scoring significantly worse than the DS and SLI groups. Therefore, even within the relative strength of verbal abilities, there exists a complex pattern of micro strengths and weaknesses, which impact on the everyday communicative functioning of individuals with WS across the lifespan.

Another area of cognitive functioning which was initially labelled as 'spared' in WS was theory of mind. It was proposed that individuals with WS had a particular aptitude for inferring the mental states of other people (e.g. Karmiloff-Smith, Klima, Bellugi, Grant & Baron-Cohen, 1995). Tager-Flusberg, Boshart and Baron-Cohen (1998) compared the performance of adults with WS on a theory of mind task (the 'Eyes Task'; Baron-Cohen, Jolliffe, Mortimore, &

Robertson, 1997) to an age, IQ and language matched group of adults with Prader-Willi syndrome and a group of age-matched typically developing adults. Prader-Willi syndrome is a genetic disorder, in which individuals often experience delays in language and motor development, along with learning difficulties, sleep disturbances and impulsive eating behaviour (Cassidy, 1997). In the Eyes Task, participants had to match labels to photos of complex mental state expressions in the eye region. There are no executive function components to the task, and it does not involve tracking complex narratives, leading to the suggestions that it has high validity (Baron-Cohen et al., 1997; Happé, 1994). Tager-Flusberg and colleagues found that adults with WS performed significantly better on this task compared to the adults with Prader-Willi, and about half of the adults with WS performed at a similar level to the typically developing adults. This led them to conclude that metalizing is a distinct, and spared, cognitive domain in WS.

More recent research, however, has begun to suggest that theory of mind abilities are in fact far from spared. Porter, Coltheart and Langdon (2008) examined theory of mind abilities in a group of 30 individuals with WS (mean age = 17.02 years), and compared them to a chronologically age matched typically developing control group, and a mental age matched typically developing control group. They used a picture sequencing task (Baron-Cohen et al., 1986), as a non-verbal measure designed to assess understanding of pretence, intention and false belief. Results showed a specific deficit in understanding false belief in WS, although the group performed similarly to the mental age matched control group on stories which required understanding of intention, unrealised goals and pretence.

Interestingly, the authors also noted considerable heterogeneity within the WS group. This is something which will be explored in greater detail in section 1.3.3. Like language, it seems that the original claims of theory of mind abilities being entirely sparred in WS are inaccurate, and even within areas now considered to be relative cognitive strengths, there exists a complex pattern of abilities.

1.3.2 Relative weaknesses in the WS cognitive profile

There are also areas of weakness within the paradoxical cognitive profile associated with WS, the most prominent of which is in the area of visual-spatial functioning (e.g. Bellugi, Sabo, & Vaid, 1988). Bihrle, Bellugi, Delis and Marks (1989) assessed visual-spatial functioning in a group of children and adolescents with WS (mean age = 13.1 years) compared to individuals with DS and typically developing individuals who were matched on chronological and mental age. Participants were asked to draw figures which contained either hierarchically organised global forms composed of local forms, or single items consisting of either one local or one global form. They found that performance on the tasks overall was comparable in the WS and DS groups and significantly poorer than the typically developing chronologically age matched group. However, there was a clear dissociation between the clinical groups in their ability to draw global versus local stimuli. Participants with WS focused exclusively on the local aspects of the hierarchically organised design and drew accurate, small forms of the design, with little attempt at reproducing the global configuration. For the participants with DS, the opposite pattern was found. This led the authors to suggest that individuals with WS show considerable impairment in global relative to local analysis, which is likely to be contributing to the poor visual-spatial

abilities seen in this group. Indeed, using Block Design tasks, it has been suggested that individuals with WS show a propensity to process spatial information at a local level, as seen in younger TD children (Bellugi et al., 1994). This means that they are often able to correctly identify shapes, but are unable to replicate the structure that the shapes were presented in (Georgopoulos, Georgopoulos, Kuz & Landau, 2004).

Other explanations have been proposed for the relative weaknesses in visual-spatial processing. Pani, Mervis, and Robinson (1999) used a visual search task which was sensitive to spontaneous global spatial organisation. Adults with WS (mean age = 30.9 years) and a control group of typically developing adults matched on gender and chronological age took part in the visual search task. The authors found that the adults with WS were more influenced by gestalt grouping than by display size. They were also less affected by the number of distractors compared to the control group, and found it more challenging to switch from global to local processing than the control group. It was suggested that rather than individuals with WS demonstrating deficits in visual-spatial construction abilities due to their reliance on local spatial processing, their impairments in this area instead result from a difficulty in changing between global and local processing strategies.

Interestingly, Farran, Jarrold, and Gathercole (2001) offered a third explanation for the visual-spatial impairments seen in WS. Participants with WS (mean age = 19.9 years) and a group of mental age matched typically developing controls took part in the Children's Figures Test (CEFT; Witkin, Oltman, Raskin, & Karp,

1971) and a mental rotation task. It was found that individuals with WS were significantly more impaired in their ability to use mental imagery, compared to the control group. However, the WS group did not differ from the TD group in their processing style. The authors suggested that differences in performance were more likely the result of poor visual imagery in the WS group. Even within the domain of visuospatial processing there exists some relative strengths, such as a relatively strong ability to recognise and remember faces (e.g. Bellugi et al., 1994; Udwin & Yule, 1991), in spite of their overall levels of impaired spatial cognition (Bellugi et al., 1999). Whilst the explanations for the poor visuospatial performance seen in WS remain contested, the literature is largely equivocal in finding that these deficits exist, and that they are a key component of the WS cognitive profile.

1.3.3 Heterogeneity of the WS cognitive profile

As outlined at the start of this section, the description of the cognitive profile outlined above represents the 'average' cognitive profile, but this ignores the vast within-syndrome differences that exist in WS (e.g. Borg et al., 1995; Pankau et al., 2001). Porter and Coltheart (2005) challenged the idea of a homogenous WS cognitive profile and the over-reliance on group means. They examined whether a universal cognitive profile exists in participants with WS (mean age = 16.9 years) using the Woodcock-Johnson Tests of Cognitive Ability – Revised (WJ-R COG; Woodcock & Johnson, 1990) which measured long-term memory, immediate memory, processing speed, auditory processing, visual processing, comprehension, reasoning and expressive language. They found considerable within-group heterogeneity across tasks. All participants displayed an uneven

cognitive profile, however, the pattern of their strengths and weaknesses differed. In some tasks, verbal ability was impaired for some participants, and non-verbal skills were found to be a strength. Indeed, Porter and Coltheart (2005) used these findings to suggest that there is preliminary evidence for subgroups of WS individuals, based on their heterogeneous cognitive profile. They proposed that there are at least two subgroups within the WS profile, which can be identified based on attention, perceptual and spatial construction abilities and differences in social-emotion skills, although they advise that these suggestions are preliminary. Nevertheless, it seems that individuals with WS do not constitute a homogenous group, and the idea that all individuals display strengths in language and weaknesses in non-verbal spatial skills cannot be substantiated. The importance of the methods used to access this within-syndrome heterogeneity will be explored in Chapter 2.

1.4 Social profile

Whilst the cognitive profile held the attention of researchers in the 1990s, the new millennium saw far more research focused on the social profile of the disorder. Individuals with WS are typically characterised as being outgoing, friendly, gregarious individuals, who display an extreme prosocial drive to interact with others (Frigerio et al., 2006). Jones et al. (2000) investigated the hypersocial phenotype associated with WS. They explored the early development of the social nature of WS in infants (mean age = 1.5 years). The Parental Separation Task was used from the LabTab (Goldsmith & Rothbart, 1991). The frequency and intensity of affective responses on the face, on the body and through the voice were recorded when the infant was separated from their parent and then when

they were reunited. They found that the children with WS displayed fewer negative emotions based on their facial expressions, body movements and vocal expressions compared to chronologically age matched peers. The children with WS were also more likely to quickly resume play with their parent when they returned, whereas the typically developing controls required more consoling. Interestingly, the authors noted that the children with WS displayed a high level of interest in other people, as shown by positive emotional expressions, and engaging behaviour directed towards other people. As an example, several children with WS would try to engage with the experimenter, through eye contact and positive facial expressions, resulting in them failing some of the cognitive tasks because they were focused on interacting with the experimenter. This suggested that the hypersocial profile associated with WS emerges early in life, as they show an attraction to social interaction, even with strangers.

Their excessive friendly behaviour is distinct even from other groups of IDDs. Although individuals with WS and DS are often both considered to be social and friendly, it is the overfriendliness, social disinhibition and difficulty forming and maintaining peer relationships seen in WS that distinguishes the social profile of these two groups (Dykens et al., 2000). Rosner, Hodapp, Fidler, Sagun and Dykens (2004) compared the social competence of individuals with WS, DS and Prader-Willi syndrome (age range 4 – 49 years) using the Child's Behavior Checklist (CBLC; Achenbach, 1991). They found that individuals with DS showed the highest overall social competency scores, followed by the WS and Prader-Willi groups, and that age positively correlated with social competence for individuals with WS and DS. This suggests that although individuals with WS are

more social than individuals with DS (Doyle, Bellugi, Korenberg & Graham, 2004; Jones et al., 2000), they are less socially competent. The hypersociable behavioural phenotype seen in WS therefore has important implications on their social interactions, especially when considered alongside the profile of relative strengths in language abilities and mild-moderate levels of intellectual impairment outlined above.

1.4.1 Social approach behaviour

One key aspect of the WS hypersocial behavioural phenotype is their propensity to approach others (e.g. Bellugi, Adolphs, Cassady & Chiles, 1999). Dodd, Porter, Peters and Rapee (2010) examined the willingness of children with WS (age range = 3 – 6 years) to approach and engage with strangers, compared to children matched on chronological age or mental age. Children took part in four play sessions in an unfamiliar environment, two were non-social and two were social. The first social play sessions assessed the willingness of participants to engage with a stranger whose face was visible. In the second social play session, the strangers face was not visible. Behaviour was observed and coded. The authors found that young children with WS were more willing than their chronological or mental age matched peers to engage with a stranger. Further, this tendency to approach and engage with strangers occurred even when the strangers face was not visible. Children with WS were specifically more likely to initiate interaction with a stranger, which suggested a difference in motivation. For children with WS, they were motivated to engage with a stranger simply when they saw them, whereas the typically developing children were only motivated to engage with the stranger when toys were available.

This social approach behaviour and desire to engage with others seems to extend through the developmental trajectory. Jones et al. (2000) examined the hypersociability of a group of adults with WS (mean age = 23.6 years) towards strangers, compared to a typically developing control group matched on chronological age and gender and one matched on mental age. Participants took part in the Approachability task (Adolphs, Tranel & Damasio, 1998), in which they viewed a series of black and white photographs of faces which had been pre-rated as highly approachable, or highly unapproachable. Participants were asked to indicate how much they would like to go up and talk to the person in the photograph, and they had to respond using a five-point Likert scale. Adults with WS rated both the approachable set of faces and the unapproachable set of faces higher on approachability than the control groups. This reinforces the idea that individuals with WS show an abnormal desire to approach and interact with unfamiliar people.

Reports of elevated social approach behaviour in WS are also present in parent reports. In the Salk Institute Sociability Questionnaire (SISQ; Jones et al., 2000), parents of children with WS aged 1 – 12 years old rated their son/daughter's tendency to approach others and their tendency to be approached, their behaviour in social situations, their eagerness to please other people, their empathetic abilities and their general behaviour in social situations (Doyle, Bellugi, Korenberg & Graham, 2004). Compared to typically developing children, and children with DS, the WS group were rated higher in all areas of sociability, especially on their approach behaviour towards strangers (the groups did not

differ in their approach behaviour towards familiar people). When the WS group was broken down by age (youngest, intermediate and oldest), there was little variability in their social approach behaviour in any of the groups. Indeed, previous work from the group found the same pattern in adolescents and adults with WS, using the SISQ (Jones et al. 2000). This led Doyle and colleagues to suggest that individuals with WS may have an innate predisposition towards hypersociability and in particular, the tendency to approach strangers.

Neuroimaging studies have also supported this notion of high levels of approach behaviour in WS. Martens and colleagues (2009) investigated approachability judgements in relation to amygdala volume in individuals with WS (mean age = 16.9 years) and typically developing, chronologically age matched controls. Participants completed a modified version of the Adolphs Approachability Task (Bellugi et al., 1999). It was found that participants with WS gave higher ratings of approachability to faces showing positive emotions and to faces showing negative emotions, compared to the control group. They also found that the ratings of approachability given by participants with WS correlated with the participant's amygdala volume. Specifically, there was an association between the right amygdala volume and approachability judgements, especially for stimuli depicting negative affect. The same pattern was not found for typically developing individuals. The authors suggested that this offers support for the notion of amygdala dysfunction playing a crucial role in the hypersocial behaviour seen in WS (for a discussion of the amygdala theory of WS social behaviour, see section 1.5.2).

However, others have argued that the emotional expression of the faces presented plays a key role in social approach behaviour in WS (e.g. Porter, Coltheart, & Langdon, 2007). Frigerio and colleagues (2006) examined approach behaviour in WS in response to different facial expressions. Individuals with WS and control groups matched on gender and either chronological age or mental age viewed a series of photographs showing different facial emotions (anger, disgust, fear, happiness, sadness and neutral facial expressions). They were asked to indicate whether they would like to interact with the person in the photograph. If they gave a positive response, then they were further asked to what extent they wished to interact with the person ("a little" or "a lot"). If they gave a negative response, then they were given two further options to clarify their answer ("probably not" or "definitely not"). Unlike the indiscriminate social approach behaviour outlined by the studies above, Frigerio et al. (2006) found that participants with WS only rated the happy faces as being more approachable than the ratings given by the control participants. Indeed, they judged all of the other stimuli (disgust, fear, sadness and neutral facial expressions) as being less approachable compared to control participants. This challenges the notion that individuals with WS always show higher levels of approach behaviour than their typically developing peers. Instead, it seems possible that individuals with WS discriminate their social approach behaviour based on facial expressions; they show higher levels of social approach behaviour towards faces which show positive affect compared to controls and lower levels of approach behaviour towards faces which show negative affect, compared to controls.

Porter et al. (2007) further investigated this possible link between emotion and approachability using the Diagnostic Analysis of Nonverbal Accuracy (DANVA: Nowicki & Duke, 1994) stimuli. Participants with WS (age range = 5 – 43 years) were asked to identify emotions (happy, sad, angry, scared) from a series of faces, postures and voices. Performance was compared to participants with DS, as well as typically developing chronologically age matched and mental age matched controls. They found that emotion recognition abilities affected scores on the social approach task for all groups. Individuals with WS and DS often misidentified a negative emotion as being positive, although this was also found in the typically developing control groups. Similarly, Järvinen-Pasley et al. (2010) found that individuals with WS (mean age = 29.7 years) judged faces of strangers as being more approachable compared to ratings given by their TD peers, but interestingly, they found that these high levels of self-rated willingness to approach strangers correlated with impaired abilities to identify emotions. This indicates that that the hypersociability observed in WS could be in part attributed to atypical perceptual processing of faces, which results in the misidentification of emotions and therefore intent (Kasari et al., 2001).

Indeed, Plesa-Skwerer et al. (2009) found that, compared to typically developing controls matched on verbal mental age, individuals with WS (mean age = 8.1 years) were less able to identify negative facial emotions and negative voice tones and showed automatic hypoarousal to negative faces. They suggested that the lower skin conductance responses seen in WS, as well as the heart rate decelerations, were indicative of interest in faces, rather than arousal. It therefore seems that individuals with WS are less aroused when viewing social images

depicting negative affect (Haas et al. 2009; Meyer-Lindenberg et al. 2005), though general under-arousal in terms of skin conductance is typical in individuals with WS (Doherty-Sneddon, Riby, Calderwood & Ainsworth, 2009). Taken together, these findings suggest that the disinhibited social approach behaviour seen in WS could be driven by a reduced ability to identify social threat signals combined with a positive bias for attending to social stimuli, which is contributing to the atypical development of their emotional processing.

Despite being highly empathetic individuals, it therefore seems that many individuals with WS process emotions atypically (Gagliardi et al., 2003; Plesa Skwerer, Faja, Schofield, Verbalis, & Tager-Flusberg, 2006), especially when they are required to identify complex emotions / mental states of others (e.g. disinterested, worried, sympathetic, Zinck & Newen, 2008; Tager-Flusberg, Boshart & Baron-Cohen, 1998; Plesa Skwerer et al., 2006) rather than basic emotional expression (e.g. happiness, sadness, fear, anger, surprise and disgust; Ekman, 1993; Porter et al., 2007; Karmiloff-Smith, Klima, Bellugi, Grant, & Baron-Cohen, 1995; Tager-Flusberg & Sullivan, 2000; Gagliardi et al., 2003). Plesa Skwerer et al. (2006) tested adults with WS (mean age = 20.8 years) on their ability to recognise complex emotions (e.g. interested, upset, playful, alarmed, preoccupied, grateful) in the revised Reading the Mind from the Eyes task (Baron-Cohen et al., 2001). Participants were presented with pictures of the eye region of the face and had to select the appropriate label for the emotion portrayed by the eyes in the photo (four options were given). Their performance was compared to a general learning difficulty group and a typically developing group, who were matched to the adults with WS on verbal ability. The authors

found that adults with WS performed at a level similar to individuals with learning difficulties on the Reading the Mind from the Eyes task and significantly worse than the typically developing participants. They suggested that the abilities of adolescents and adults with WS in identifying complex mental states were at a level predicted by their mental age, rather than their chronological age.

Tager-Flusberg, Boshart and Baron-Cohen (1998) however, offered alternative findings. They examined the ability of adults with WS (mean age = 27.3 years) to label photographs of the eye region which showed complex mental states ('Reading the Mind from the Eyes' task; Baron-Cohen, Wheelwright & Jolliffe, 1997). Participants were given the option to choose between two semantically opposite emotion labels for each trial (e.g. calm/anxious, decisive/indecisive, flirtatious/not interested). The performance of adults with WS on this task was compared to adults with Prader-Willi syndrome, and chronological age matched typically developing controls. They found that around half of adults with WS scored within the same range as the typically developing control group, and they performed significantly better than adults with Prader-Willi syndrome, who were matched on age, IQ and language ability. The authors suggested that these findings were indicative of a relative sparing of this kind of metalizing capacity, though they acknowledged that generalisations of these findings may be challenging given the comparison group, the number of mental states included in the task, and small participant numbers (n = 13 adults with WS).

Interestingly, this appears to be mirrored in young people with WS. In a study by Riby and Back (2010) young people with WS (mean age = 13.08) were presented

with video clips of dynamic or static faces. They were asked to observe the clip of the person's face then select a word that best describes how that person is thinking or feeling. It was found that the children and adolescents with WS performed at a level comparable to their chronologically age matched peers when identifying complex emotions from moving faces (e.g. relieved, disapproving; taken from Back, Ropar & Mitchell, 2007). By assessing performance on this task when different facial features were frozen, the authors also concluded that the eye region plays a crucial role for individuals with WS when interpreting mental state information. These findings tentatively suggest that deficits in emotion identification are linked to the social interaction difficulties seen in WS.

1.4.2 Face processing

In order to better understand the role of emotion identification in social approach behaviour, other socio-perceptual characteristics of WS should be examined, for example, their propensity for prolonged face fixation. Riby and Hancock (2008) investigated eye gaze patterns in individuals with WS (mean age = 17.06 years), autism (mean age = 13.04 years) and chronologically age matched and non-verbal ability matched control groups. Given the opposing social profiles associated with WS and autism (see Chapter 2, section 2.6.2 for further discussion), the authors were interested in investigating how these individuals view scenes containing people. Participants viewed colour photographs of social scenes involving actors. They were instructed to look at the picture and eye tracking techniques were used to monitor the movement of their gaze. Results showed that both clinical groups spent similar amounts of time viewing the photos, however, the two groups showed differences in their viewing patterns. Individuals with autism spent

significantly less time viewing actors faces, compared to the control groups, whereas, individuals with WS spent significantly longer than the typical control groups viewing the face region. Further, participants with WS spent significantly longer gazing at the eye region of the face (58% of gaze time) compared to participants with autism (17% of gaze time). The eyes are central to expressing communicative signals, as they can provide insight into mental states (Baron-Cohen, 1995). These findings suggest that, in line with the social profile outlined above, individuals with WS show atypical patterns of gaze behaviour and spend significantly longer looking at a person's face, especially their eye region. Riby et al. (2011) suggested that prolonged face fixation may be underpinned by difficulty disengaging from social stimuli, as opposed to faster attentional engagement. They found that participants with WS took significantly longer to disengage their attention from faces compared to objects, a pattern which was not found in the TD comparison group. Thus, atypical patterns of attention, combined with their difficulty disengaging from social stimuli is likely to contribute to the atypical patterns of gaze behaviour reported in WS (Cornish, Scerif, & Karmiloff-Smith, 2007; Mervis et al., 2003; Riby & Hancock, 2009).

Another aspect of gaze behaviour found to be atypical in WS is gaze aversion. Doherty-Sneddon, Whittle and Riby (2013) investigated gaze aversion patterns in WS during face-to-face interactions with familiar and unfamiliar people. Participants with WS (mean age = 21.11 years) were matched to typically developing participants based on their verbal ability. Gaze aversion (defined as, "occurring whenever direction of eye gaze was diverted away from the face of the interlocutor", p620) was scored during interactions with familiar and unfamiliar

adults. It was found that participants with WS showed similar levels of gaze aversion to their typically developing counterparts, although they did engage in less gaze aversion when thinking (rather than listening or speaking) during interactions with unfamiliar people than did controls. It was suggested that individuals with WS display less social inhibition around unfamiliar people (this is explored further in Chapter 5).

However, work by Hanley and colleagues (2013) challenged this widely held assumption that individuals with WS show atypical gaze behaviour and a propensity for prolonged gaze to the face region. Individuals with WS (mean age = 21.6 years) were presented with a series of static and dynamic faces on a computer and were asked to make a judgement on the mental state of that person. They found that participants with WS spent less time looking at the face region when making their decisions about mental state, compared to a chronologically age matched and mental age matched control groups. When analysing the patterns of gaze fixation, they found that participants with WS did not show prolonged gaze towards the eye region, instead they showed reduced attention to the eyes, compared to typically developing participants. They suggested that the complexity of the stimuli used was likely to have affected patterns of attention. Indeed, Riby and Hancock (2009) found that participants with WS can show prolonged face gaze and typical face gaze depending on the type of stimuli used (e.g. cartoon, real human, isolated faces, social scenes). Further, they proposed that there exists wide heterogeneity of social perceptions in WS and that working with such small samples and a heterogeneous condition is a problem for this research area. Therefore, it is likely that the propensity for prolonged gaze

towards the face region exists in WS, but this is dynamic and can change depending on the individual and the context that they are in. When considering the social vulnerability of this group, the behaviour of individuals with WS towards strangers is an important context to explore further.

1.4.3 Stranger danger awareness

Stranger danger awareness develops in typically developing children as young as 6 months old; infants aged between 6 and 12 months begin to show stranger anxiety and a wariness of unfamiliar people (Rheingold & Eckerman, 1973). Moran, Wrden, Macleod, Mayes and Gillies (1997) argue that this instinctual fear, combined with explicit teaching for school aged children, means that an awareness of stranger danger develops between the ages of 8 and 10 years old in typically developing children. Though at this age children can still be vulnerable to the lures of strangers. Dickson and Hutchinson (1988) examined stranger danger awareness in a large sample (n = 585) of 9 – 12 year old typically developing children. They found that the children were aware of the risks that strangers could pose; however, they were unsure how to act when approached by a stranger. Similarly, Krazier, Fryer and Miller (1988) proposed that children may appear to have the conceptual knowledge of the dangers posed by strangers, however, when approached by a stranger to accompany them outside, they struggled to implement this knowledge of what to do. This suggests a possible dissociation between knowledge and actions in young typically developing children. However, less is known about stranger danger awareness in individuals with IDDs.

Riby et al. (2014) investigated stranger danger awareness in young people with WS (mean age = 12.1 years) using a video vignette task. Dickson and Hutchinson (1988) argue that by 12 years old, stranger danger knowledge has developed in typically developing children. Participants viewed an age-relevant clip of a child interacting with a stranger. For example, in one clip, a stranger approached a young boy called Jamie who was playing in a park with other children, while his mum sat nearby. The man told Jamie that he had lost his dog and asked if Jamie would help him find the dog. After the clip, participants were asked, "What do you think Jamie should do?" followed by, "Should he help the man find his dog?" They were asked to give reasons for their answers and were asked what they would do in the same situation. Quantitative and qualitative analysis showed that individuals with WS showed a lack of awareness of the possible risks associated with interacting with a stranger, compared to typically developing children matched on verbal ability. When participants with WS indicated that the child in the video clip should interact with the stranger, thematic analysis showed a lack of understanding that the child should indeed be very cautious during the interaction. The authors offered the preliminary suggestion of a link between impaired everyday social behaviours (including difficulties maintaining peer relationships), and low levels of stranger danger awareness, as reported by parents.

A lack of stranger danger awareness in WS transcends the developmental spectrum, following a vastly different trajectory to that reported in typically developing individuals. Fisher et al. (2013) evaluated a stranger safety training programme for adults with WS. Participants took part in a three day social skills

intervention, specifically designed to teach appropriate responses to stranger lures. Real life scenarios were set up, in which participants were approached by a confederate and an interaction ensued. Fisher et al. (2013) found that, prior to the intervention, only 14% of participants resisted the lure proposed by a stranger. However, after the intervention, 62% gave appropriate responses. Whilst this suggests that stranger danger social skills can be learnt by individuals with WS, 14% still agreed to leave with the stranger post intervention, and there is no longitudinal data showing the longevity of these new skills. Further, without this intervention, this study shows that even adults with WS show low levels of stranger danger awareness (86% of the sample failed to say 'no' and walk away from a stranger when approached with a lure). This is particularly concerning as this is a time when they are also enjoying higher levels of independence (Davies et al., 1997).

1.4.4 Social vulnerability

The social profile associated with WS which has been outlined above, combined with the mild-moderate levels of intellectual impairment, mean that individuals with WS are often considered to be highly socially vulnerable (Jawaid et al., 2012). Their impaired ability to identify social threats and extreme prosocial drive can put them in high risk situations (Riby, Kirk, Hanley & Riby, 2014). Despite their outgoing and friendly personality, many individuals with WS struggle to form and maintain peer relationships, particularly as they get older (Davies et al., 1998). Davies et al. (1997) studied independence in adults with WS aged 18 – 39 years old. Using the Vineland Adaptive Behavior Scale-Interview Edition (Sparrow et al., 1984) they found that only 30% were in employment and almost

all of them had characteristics which their supervisors reported threatened their job stability, such as over-friendliness, anxiety and socially inappropriate behaviour. It is perhaps not surprising therefore that an estimated 73% of adults with WS experience social isolation (Davies et al., 1998).

Individuals with intellectual and developmental disorders are known to experience high levels of victimisation, as outlined in section 1.1. This includes bullying, theft and abuse (Nettelbeck & Wilson, 2002). Fisher et al. (2012) asked parents to complete the Social Vulnerability Questionnaire (SVQ; Fisher, Moskowitz & Hodapp, 2012) about their sons/daughters (mean age = 24.5 years; 80% lived at home with a family member; 40% were in paid employment). They found that social vulnerability in individuals with IDD was linked to characteristics such as: low risk awareness, vulnerable appearance, parental independence and low levels of social protection. 75% of parents also provided examples of victimisation that their son/daughter had experienced. The most frequent examples related to teasing or persuasion (35.6% of the sample), theft (34.2%) and physical or sexual abuse (21.2%).

Interestingly, the patterns of vulnerability experienced by individuals with IDD are thought to be syndrome specific (Fisher, Moskowitz, & Hodapp, 2013). For individuals with WS (mean age = 25.4 years), their social vulnerability was found to be linked to high levels of perceived vulnerability (i.e. physical traits that result in others perceiving them to be an easy target for victimisation) and parental independence (i.e. high levels of time spent away from their parents; Fisher et al., 2013). However, given the differences in the genetic, physical and cognitive

profiles associated with WS (Porter & Coltheart, 2005), the heterogeneity of the social profile is likely to have important consequences when considering the social vulnerability of this group.

1.4.5 Heterogeneity of the WS social profile

Similar to the suggestions of Porter and Coltheart (2005) about heterogeneity in the cognitive profile of WS (see section 1.3.3), recent work has also begun to question the homogenous nature of the social profile (e.g. Porter et al., 2007; Jarvinen-Pasley et al., 2010) outlined in the sections above. Little et al. (2013) investigated the heterogeneity of social approach behaviour in children with WS, by exploring performance on tasks assessing social salience, emotion recognition and response inhibition. They were interested in establishing whether there was evidence for social approach behaviour subgroups based on these constructs. Twenty-five children with WS (mean age = 9.5 years) completed the Adolphs Approachability Task (Adolphs et al., 1998) which is designed to assess participants' willingness to approach unfamiliar people, an emotion recognition task (which used the basic emotions of happy, sad, angry, surprised, scared and neutral) and The Sun–Moon Stroop Task (Archibald & Kerns, 1999) as a measure of response inhibition. In the approach task, participants showed comparably high levels of approachability for positive affect faces to previous studies (e.g. Jones et al., 2000; Martens et al., 2009). They also found approach ratings for faces showing negative affect to be higher than the levels found in these other studies. Interestingly however, they noted substantial variability in the approachability ratings which were indicative of distinct subgroups. Subgroups could be identified based on their levels of response inhibition. Those who displayed high

levels of social approach behaviour were also found to have the lowest levels of inhibitory control, whereas those who scored highly on the response inhibition task formed a cluster of individuals who also showed low levels of approach behaviour. Each subgroup contained individuals of varying ages and IQ levels, suggesting that developmental variables were not predictive of subgroup classification. This lends support to the frontal lobe hypothesis (which will be discussed in section 1.6.1), and reinforces the notion that individuals with WS are not a homogenous group. This is important because, combined with the heterogeneous cognitive profile outlined in section 1.3.3, this suggests that social vulnerability in WS is likely to be a heterogeneous entity. This means that appropriate methods must be selected to access the experience of the individual, and this thesis should not be reliant solely on group means in its exploration of social vulnerability in WS (for discussion of the methods selected in this thesis, see Chapter 2)

1.5 Psychopathology in WS

Intertwined in the social profile that has been outlined in the above sections is a complex profile of psychopathology that is prevalent in WS (Riby et al., 2014). Given that mental health issues were outlined in section 1.1 as a pre-cursor to increased vulnerability, it is important to consider the psychopathological profile that is seen in WS when exploring the notion of social vulnerability in this cohort. However, identifying psychopathology in individuals with IDD can be challenging. Measurement tools have often been developed in a typically developing population (see Lecavalier et al., 2014 for a discussion on the use of anxiety measurements normed in typically developing populations for individuals

with ASD), and diagnostic over-shadowing can result in under-reporting given the over-lapping symptoms between the IDD and the psychopathology (Mason & Scior, 2004; van Steensel, Bögels & Dirksen, 2012; MacNeil, Lopes & Minnes, 2009; see Chapter 2 for further discussion on the applicability of research measures used in this thesis).This confusion is shown in initial estimates of mental health problems in adults with learning difficulties, which ranged from 7% to 97% (Wright, 1982; Linaker & Nitter, 1990; King et al., 1994).

More recently, Cooper, Smiley, Morrison, Williamson and Allan (2006) conducted a large scale (n = 1,023) population based survey to investigate the point prevalence of mental health problems in adults with learning difficulties (mean age = 43.9 years, range = 16 – 83 years). They found a point prevalence of 40.9% of the sample meeting clinical diagnosis criteria for mental health problems. Aside from problem behaviours and co-morbid ASD diagnoses, the most common mental health problems were affective disorders (6.6%; most commonly unipolar depressive episodes: 4.1%), psychotic disorders (4.4%; most commonly schizophrenia: 2.9%) and anxiety disorders (3.8%; most commonly generalised anxiety disorder: 1.7%). The authors found that severe learning difficulties, experiencing a high number of life events in the past 12 months, living with paid carer support, not having a severe physical disability and being female as some of the factors that were independently associated with mental health problems in adults with learning difficulties. However, other factors which have been implicated in mental health problems in the typically developing population, such as living in more deprived areas, no daytime occupation and marital status, were not found to be associated with mental health problems in

adults with learning difficulties. They argued that healthcare services may not be adequately meeting the mental health needs of adults with learning difficulties, which could further compound the impact these problems have on their daily lives. Indeed, Smiley et al. (2007) noted that the two year incidence of mental health problems in adults with learning difficulties was 16.3%. In comparison to the general population, children and adults with learning difficulties are at an increased risk of developing mental health problems (Dykens, 2000).

1.5.1 Depression

Rates of depression in individuals with WS are considered to be relatively low in comparison to other types of psychopathology (Leyfer et al., 2006), and the levels experienced by individuals with other forms of IDD. For example, review papers on the rates of depression in individuals with ASD have suggested a prevalence rate as high as 57% (Lainhart, 1999; Ghaziuddin, Ghaziuddin & John Greden., 2002). Depression in ASD has been found to be associated with an increase in non-cooperative behaviour and self-harming (Kim et al., 2000). It has been highlighted as the most common psychiatric diagnosis in ASD (Wing, 1981; Ghaziuddin et al., 1998; Ghaziuddinet al., 2002)

In comparison, Cherniske et al. (2004) found a prevalence rate of 10% for depression in adults with WS (mean age = 38.8 years). Similarly, Dodd and Porter (2009) found a prevalence rate of 14% for depressive disorders in children and adults with WS (mean age = 18.5 years), and a rate of 25% when only the adult group was examined (mean age = 27.3 years). This suggests that individuals with WS may become more susceptible to depression later in life. This coincides

with a time when their levels of isolation increase (Davies et al., 1998), which could have important implications for their vulnerability status. Exploration of social vulnerability in adults with WS is offered in Chapters 7, 9 and 10.

1.5.2 Anxiety

Dykens (2003) noted that several studies had indicated, based on informal questioning or the use of global rating scales such as the Rutter Questionnaire (Rutter, 1967) or the Child Behaviour Checklist (Achenbach, 1991), that children and adults with WS seem to be more fearful compared to other individuals with IDDs and their typically developing peers. However, the psychiatric features of WS had not been systematically studied. Dykens (2003) therefore set out to investigate anxiety and fears in WS through a series of studies using a DSM based interview, which encompassed both parent and self-report (see Chapter 2 for a discussion on the benefits of multi-informant approaches). Children and adults with WS (mean age = 16.6 years) were found to score higher on the Fear Survey Schedule for Children – Revised (FSSC-R; Ollendick, 1983) compared to a chronologically age matched group of individuals with learning difficulties. They found that specific phobias had the highest prevalence in this population (35%), followed by generalised anxiety disorder (GAD; 16 – 18%) and separation anxiety disorder (4%). Interestingly, 57% were said to be excessively worried about the future, 35% had become sick from worry and 25% showed an inability to relax. This suggested that anxiety disorders were likely to be highly prevalent in individuals with WS, with rates even surpassing the high levels seen in the learning difficulties population.

Leyfer, Woodruff-Borden, Klein-Tasman, Fricke and Mervis (2006) examined the prevalence rate of anxiety disorders in school-aged children with WS. They noted that Dykens (2003) had employed a broad age range, and used a child interview measure for both child and adult participants so argued that more research was needed to define the psychiatric phenotype associated with WS. The authors used the Anxiety Disorders Interview Schedule for DSM-IV Parent Interview Schedule (ADIS-IV Parent; Silverman & Albano 1996) to assess the prevalence of anxiety disorders in the sample (n = 119). They found that Attention Deficit Hyperactivity Disorder (ADHD; a condition characterised by inattention, hyperactivity and impulsivity; APA, 2000) was the most frequent co-morbid diagnosis in children with WS. The prevalence rate of 64.7% was higher than the prevalence rate for the general population (3-7%; APA, 2000) but similar to the prevalence rate reported for children and adolescents with Down syndrome (Myers & Pueschel, 1991). Specific phobias were also found to be highly prevalent in children with WS. 53.8% of the sample was reported to have a specific phobia, which is higher than the rates reported in the work of Dykens (2003), and rates reported in both of these studies are higher than levels of specific phobias reported in children and adolescents with Down syndrome (Myers & Pueschel, 1991). The most common phobias reported for children with WS were of loud noises and visits to doctors/dentists. Finally, 12% of the sample met the diagnostic criteria for GAD, which is considerably higher than the rates reported in typically developing children (2-4%; e.g. Bowen et al., 1990).

Similar results have also been obtained in adults with WS. Stinton, Elison and Howlin (2010) examined mental health problems in adults with WS (mean age =

32 years) using the Psychiatric Assessment Schedule for Adults with Developmental Disabilities (PAS-ADD; Moss et al., 1996). They found that anxiety disorders were common in their sample, with 16.5% receiving a diagnosis. Specific phobias were the most common type of anxiety disorder diagnosed (12%). They concluded that anxiety was the most significant mental health problem facing adults with WS. Despite the inconsistency in the methods and measurement tools used across studies and across age groups, high levels of anxiety are consistently reported.

It is the impact that these high levels of anxiety could have on social behaviour that is of greatest interest to the current thesis. Riby et al. (2014) looked at the interplay between anxiety and social functioning in WS. Parent reports of anxiety indicated that 46% of sample (n=59; age range 6 – 36 years) experienced high levels of anxiety (with scores indicative of clinical levels of anxiety), which was not related to age or IQ. When asked about their son/daughter's social behaviour, only 17% of parents reported that their child showed social behaviour within the 'normal' range, suggesting that most individuals with WS in this study struggled with social reciprocity. In line with Klein-Tasman et al. (2011), participants showed higher levels of impairment in the social-cognition domains, compared to the domains of prosocial functioning. Interestingly, Riby et al. (2014) also showed that there was a difference in the social profile (social awareness, social cognition and social communication) of individuals who experienced high levels of anxiety, compared to those who experienced low levels of anxiety. Those who have higher levels of anxiety showed greater levels of social dysfunction than those who scored lower on the measure of anxiety. There was no difference

between the groups on their level of social motivation or autistic mannerisms. This shows that anxiety is important in shaping the pattern of social abilities and impairments experienced by both children and adults with WS, which has consequences for their resilience and levels of social vulnerability.

1.6 Theories of social behaviour

In an attempt to better understand the patterns of social behaviour displayed by individuals with WS, two dominant theories have been applied in the WS literature: the frontal lobe hypothesis, and the amygdala theory. More recently, the social motivation hypothesis (which has become well established in the ASD field) has also been applied to help explain social behaviour in WS. Lewis (2003) outlined criteria that theories of developmental disorders should fulfil. When applied to WS, they are as follows: the theory must reference a deficit that is experienced by all individuals with WS, it must be specific to WS rather than learning difficulties in general, the deficit described must causally precede the behaviour of interest and it must remain present throughout development. This section will explore these theories in turn and conclude with a brief discussion about the exclusivity of the theories and the extent to which these explanations are syndrome specific.

1.6.1 Frontal lobe hypothesis *(e.g. Porter et al., 2007; Little et al., 2013; Rhodes, Riby, Matthews & Coghill, 2011)*

The frontal lobe theory suggests that the atypical social behaviour seen in WS is the result of frontal lobe dysfunction, which impairs response inhibition (Porter et al., 2007). Porter et al. (2007) used the Diagnostic Analysis of Nonverbal

Accuracy (DANVA: Nowicki & Duke, 1994) stimuli to test emotion recognition and social approach behaviour in adolescents with WS (mean age = 16.13 years) and DS (mean age = 16.38) compared to typically developing control groups. The Shape School Test (Espy, 1997) was also used as a measure of response inhibition. The authors proposed that if atypical social approach behaviour was the result of frontal lobe impairment, then people with WS and DS would show impaired response inhibition on the Shape School Test (Espy, 1997) and they should know not to approach strangers or people with threatening expressions, meaning they should perform on the approach task at a similar level to the control groups. Generalised difficulties with emotion identification would also be indicative of frontal lobe impairment (Rosen et al., 2004). The results supported the frontal lobe hypothesis. Participants with WS and DS showed difficulties with emotion recognition, did not perform atypically on the social approach task, and displayed impairments in response inhibition relative to their mental age. Although this does not offer a direct link, these findings suggest that the tendency to approach strangers seen in WS is the result of low levels of response inhibition resulting from frontal lobe dysfunction. The authors argue that, like patients with acquired frontal lobe impairments, individuals with WS and DS show a dissociation between knowing and doing; they know not to approach strangers, but they still indiscriminately approach strangers, due to their low levels of response inhibition. This was supported by the findings of Little et al. (2013), which have been outlined in section 1.4.5 above.

Comparisons have been drawn between the executive functioning difficulties seen in WS and those seen in individuals with ADHD (e.g. Rubia et al., 2005;

Rhodes, Riby, Matthews & Coghill, 2011). Individuals with ADHD typically display deficits in executive functioning and impulse control which have been noted to be similar to the lack of inhibition seen in WS (Carrasco et al., 2005; Leyfer et al., 2006). ADHD has also been linked to frontal lobe impairments (e.g. Barkley, Grodzinsky & DuPaul, 1992). Rhodes et al. (2011) compared the neuropsychological functioning and behavioural symptoms of ADHD in individuals with WS (mean age = 18.4 years) to individuals with ADHD and typically developing controls matched on verbal ability. Participants completed the Conners Parent Rating Scale (CPRS; Conners, Parker, Sitarenios, & Epstein, 1998). They found no difference between the scores of participants with WS and those with ADHD on all subscales of the CPRS: both groups scored within the abnormal range. WS and ADHD also showed similar patterns of neuropyschological functioning, for example in working memory strategies. This suggests that individuals with WS experience high levels of ADHD symptoms (Carrasco et al., 2005; Leyfer et al., 2006). As outlined in section 1.4.2, Leyfer et al. (2006) found that ADHD was the most common co-morbid psychopathology in WS. This adds weight to the frontal lobe hypothesis and suggests that strategies used in ADHD to improve impulse control could be relevant for individuals with WS.

Neurological evidence also points to frontal lobe abnormalities in WS (e.g. Myer-Lindenberg et al., 2005). Mobbs et al. (2007) examined performance on a Go/NoGo response inhibition task in a group of adolescents and adults with WS (mean age = 31.4 years). In this task, participants were instructed to press a key when every letter except the letter X was presented on the screen. For the letter X

they had to withhold their response. The study used functional magnetic resource imaging (fMRI) to measure blood oxygenation level dependence signal maps during the task. The authors found that participants with WS showed reduced activity in the striatum, dorsolateral prefrontal and dorsal anterior cingulate cortices during the response inhibition task compared to the age matched control group. These findings suggest reduced engagement of the frontal lobes during inhibition tasks and offer tentative biological markers for the difficulties in response inhibition and the atypical social phenotype seen in WS.

1.6.2 Amygdala theory *(e.g. Haas et al., 2009; Martens et al., 2009; Kennedy, Glascher, Tyszka & Adolphs, 2009)*

The amygdala theory offers an alternative explanation for the social behaviour seen in WS. The amygdala (part of the limbic system) is involved in the regulation of socio-emotional behaviour, such as personal space regulation (Kennedy, Glascher, Tyszka & Adolphs, 2009; see Chapter 5 for studies on personal space regulation in WS), and the identification of facial expressions (Adolphs, 2002, Adolphs, 2003, Phelps, 2006, Phelps & LeDoux, 2005). It also forms part of the 'social brain' (a combination of the amygdala, orbitofrontal cortex and superior temporal sulcus provide the neural substrates of social intelligence; Brothers, 1990). Adolphs et al. (1999) examined the recognition of facial emotion in adult patients with bilateral amygdala damage, compared to individuals with other acquired brain inquiries (for whom the amygdala was not affected) and typically developing controls. Participants were asked to identify the basic emotion (e.g. happiness, surprise, fear, anger, disgust and sadness) expressed by the person in the photograph. They found that the patients with

bilateral amygdala damage were significantly impaired in their ability to recognise the negative emotions presented, especially fear (though there was also vast variability in performance). None of these patients were impaired in their ability to recognise positive expressions. The authors suggested that the amygdala plays a crucial role in triggering responses related to threat and danger which are shown in facial expressions. Similarities have therefore been drawn between the socio-perceptual patterns of patients with bilateral amygdala damage and those seen in WS.

Haas et al. (2009) implicated the amygdala in the WS phenotype. Using fMRI and event-related potentials (ERP), they found that adults with WS (mean age = 31.01) showed reduced amygdala activation when shown faces depicting negative emotions, such as threatening expressions. They also showed heightened amygdala activity to positive social stimuli compared to the control group. Atypical amygdala reactivity in WS could therefore be increasing attention to social stimuli depicting positive affect and decreasing attention to social stimuli depicting negative affect, such as fear. This muted amygdala reaction to social threat helps explain the lack of stranger danger awareness and disinhibited behaviour seen in WS (Martens et al., 2008).

Several studies have shown amygdala volume in individuals with WS is atypically large (e.g. Reiss et al., 2004). Martens et al. (2009) investigated the link between approachability ratings and amygdala volume in children and adults with WS (mean age = 16.9), compared to typically developing chronologically age matched controls. They found that participants with WS showed increased

amygdala volumes, as well as giving significantly higher ratings of approachability for faces displayed both positive and negative facial expressions. Interestingly, they also noted a positive association between the volume of the right amygdala and approachability ratings given by participants with WS, especially when rating faces showing negative affect. This supports the theory that structural and functional atypicalities of the amygdala could be linked to the hypersocial behaviour profile seen in WS.

The amygdala theory has also been widely applied to individuals with ASD (e.g. Baron-Cohen, Ring, Bullmore, Wheelwright, Ashwin & Williams, 2000; Grelotti et al., 2002, Howard et al., 2000). Baron-Cohen et al. (2000) proposed the amygdala theory of autism: the characteristics of social impairment seen in ASD are the result of an inability to process the emotional relevance of social stimuli. Howard et al. (2000) added some early support to this theory. They found that individuals with high functioning autism show impairment in the recognition of facial expressions of fear, identification of eye gaze direction and facial recognition memory. MRI techniques also showed bilaterally enlarged amygdala volumes in these participants. This suggests that atypical amygdala structure and activation could underpin the socio-cognitive impairments seen in ASD. This is particularly interesting given the similarities to WS. Both of these clinical groups appear to show atypical social perceptual skills, have an enlarged amygdala volume and an impaired ability to identify threat. Yet, behaviorally, they are considered to show 'opposing' social profiles (Courchesne, Bellugi, & Singer, 1995; Jones et al, 2000). The broad spectrum of difficulties associated with ASD, alongside considerable heterogeneity in both groups, contributes to much of the

overlap between the two conditions (e.g. Klein-Tasmin et al., 2009; see Chapter 2, section 2.6.2 for further discussion of the similarities and differences between individuals with WS and ASD). Theory development in WS may therefore be able to make use of these between group similarities and differences in order to further isolate neuroanatomical areas of interest to help explain social behaviour in WS.

However, the amygdala theory of WS has not gone unchallenged. Porter et al. (2007; described in section 1.5.1 above) found that the emotion recognition abilities of participants with WS were typical for their level of cognitive functioning, challenging the principles of the amygdala theory. Instead, they found that it was scores on a response inhibition task that were significantly lower than expected based on their level of functioning, offering further support for the frontal lobe theory and the role of inhibition in social functioning.

1.6.3 Social motivation theory *(e.g. Chevallier, Kohls, Troiani, Brodkin and Schultz, 2012)*

The social motivation theory has been gaining strength in the autism literature, by breaking away from the conceptualisation that social cognitive deficits account for the pervasive social impairments seen in ASD. Chevallier, Kohls, Troiani, Brodkin and Schultz (2012) proposed that social motivation deficits effect the development of social cognition, resulting in atypical social interest. Motivational mechanisms are therefore seen within this model as primary deficits in ASD. In their review of the literature, Chevllier et al. (2012) highlighted the behavioural elements of social motivation in autism. For example, a lack of social orienting is

one of the first manifestations of ASD in early life (Elsabbagh et al., 2012). Infants with ASD show infrequent orienting to their own name, diminished levels of eye contact and mannerisms akin to social aloofness (e.g. Osterling et al., 2002). Further, typically developing children tend to seek out social stimulation and demonstrate an interest in social stimuli (Berridge et al., 2009), characteristics which are largely absent in individuals with ASD. They have few meaningful friendships (e.g. Howlin et al., 2004), experience high levels of social exclusion (e.g. Rowley et al., 2012) and many experience social anhedonia which is correlated with the severity of their impairments (e.g. Chevallier et al., 2012). When these atypicalities are combined with impairments at the biological and evolutionary levels, the authors argue that the social motivation framework accounts for the diminished levels of social interest seen in ASD. They present the deficits in social cognition as a consequence, rather than a cause, of low levels of social motivation. The low levels of social interest restrict the learning opportunities that a child has by depriving them of social input. This prevents the development of expertise in social cognition that we see in typically developing children.

In comparison to the diminished levels of social interest seen in ASD, we know that individuals with WS display extreme elevated levels of social interest (Courchesne, Bellugi, & Singer, 1995; see section 1.4 above). Importantly, both show impairments in social perceptual abilities (Lincoln et al., 2007; Klein-Tasmin et al., 2009; Asada & Itakura, 2012). The social motivation theory of autism could therefore be extended to WS. In the same way that the hyposociability seen in autism can be attributed to deficits in social motivation

mechanisms, the hypersociability displayed by individuals with WS may too be rooted in atypical prosocial motivation forces. The merits of this relatively new framework will be discussed further in Chapter 11, and it will be evaluated in light of the findings of this thesis.

It is important to note that the theories presented in this section are far from mutually exclusive. In reality, most researchers acknowledge that these theories are unlikely to be absolute and rather each makes a partial contribution to our understanding of social approach behaviours in WS (Gaser et al., 2006; Meyer-Lindenberg et al., 2005) and indeed in typical development. There exists substantial overlap between the neuroanatomical regions implicated in each theory (e.g. there are a dense set of neural networks that connect the amygdala and the frontal lobes; Ghashghaei et al., 2007). Within-syndrome heterogeneity in WS also makes establishing a theory that can explain social behaviour in all individuals with WS challenging. In their current form, it is clear that these theories are not syndrome-specific. Despite their differing social profiles, theory development in WS has been closely linked to theories applied to individuals with ASD. Although the studies presented in the current thesis will not directly test these hypotheses, they offer an important context within which to situate our knowledge of social behaviour.

1.7 Ethical issues

Research on a sensitive topic, such as social vulnerability, in children and adults with an IDD merits careful consideration of the associated ethical issues. Iacono (2006) investigated the ethical complexities of including people with intellectual

disabilities as participants in research. She proposed a trade-off between protection and discrimination; researchers working with vulnerable groups must protect their participants from harm and exploitation, yet this protectionist stance can run the risk of excluding individuals with IDD from important research studies. Careful consideration of the key ethical issues relevant in this arena of work is therefore necessary.

When planning the research for the current thesis, there were many discussions around how to approach research in this sensitive area, particularly as the ecological validity of the research was important to maximise applicability to real life situations. In-depth interviews with individuals with WS about their experiences of vulnerability and victimisation were likely to cause significant distress and harm to the participant. We therefore decided to focus the research on the social behaviours which feed in to the vulnerability (e.g. personal space, trust), rather than experiences of vulnerability itself. Any qualitative work which probed issues of risk directly was carried out with the parents of individuals with WS rather than with the individual themselves (e.g. Chapter ten). The design of the studies in this thesis were set up to avoid deception and to minimise distress to the participant, whilst carrying out important research that was relevant to the issues faced in their daily lives.

In order to do this, however, we had to consider the capacity of individuals with IDD to consent to participation in research. It is widely acknowledged that capacity to consent is transient and is likely to be context dependent (Mental Capacity Act, 2005). A study by Fisher, Cea, Davidson and Fried (2006)

examined the capacity of 50 adults with mild IDD and 50 adults with moderate ID to consent to a hypothetical clinical trial, compared to individuals without IDD. They found that as a group, individuals with IDD showed consent deficits; particularly demonstrating a lack of awareness of the purpose of the research and their reasoning about their decision to participate. However, the authors noted that most individuals with IDD could make a choice about participation, they understood the methods involved in the research and the consequences of taking part. Further, many individuals showed consent capacity which was comparable to persons without an IDD. This led the authors to suggest that the need to consider individual differences when working with individuals with IDD is crucial. This approach was applied to the current thesis. For example, in Chapter ten, the adult with WS who took part was highly functioning and showed an acute awareness of the research process resulting from vast amounts of previous experience as a research participant. She could engage in a meaningful conversation about the information sheet and consent form provided. As such, it was deemed that for this study, she had capacity to consent. However, this is certainly not the case for all individuals with IDD (e.g. Dye, Hare & Hendy, 2007; Arscott, Dagnan & Krosse, 1998; Brown & Thompson, 1997).

Indeed, the ethical challenges can become more convoluted in adults with IDD who are deemed not to have capacity. As the parent in Chapter 10 said, "*The situation is becoming more complex as she gets older. It's just a question of who's got authority really, cause I don't really legally have any authority over her*". During adulthood, most individuals with WS experience increased levels of

independence which in itself is an important ethical consideration when considering informed consent by proxy and the sharing of information.

However, whilst the autonomy of adults with IDD should be fostered in research where possible, working with a vulnerable population on sensitive issues means the ethical considerations around potential disclosures should be considered. The researcher has a responsibility when working with vulnerable populations (e.g. children, individuals with intellectual difficulties) to protect their participants against harm (BPS, 2010), and a protocol should be in place should any disclosures be made. Adults with WS who participated in research for this thesis were informed that if they said anything that worried us or gave us a cause for concern, then we would have to break confidentiality and discuss what they had said with their parent/caregiver. Participants were asked to give explicit consent to this as a condition of their participant in the research.

The issues explored above represent important considerations for any researcher working on a sensitive topic with a vulnerable population. However, McDonald and Kindey (2012) suggest that there has been a movement towards scrutinising these issues at the expense of acknowledging the benefits that research on these sensitive topics can bring for individuals with IDD and their families. Indeed, Rosenthal and Rosnow (1984) have argued that the cost-benefit analysis carried out by ethics committees are often incomplete when they fail to consider the cost-benefit analysis of not carrying out the research. For individuals with WS, anecdotal reports tell us that their social behaviour is placing them in extremely vulnerable situations, where the risk of victimisation is high. The aim of the

current thesis was to develop our knowledge of some of the social behaviours which are likely to increase their levels of vulnerability, which can be used to inform future interventions to better support individuals with WS. There is therefore an argument that not conducting research in this area also holds important ethical considerations. Thorough consideration of the ethical issues associated with the current thesis has been applied in accordance with the British Psychological Society Code of Human Ethics (2010; see Chapter two for further discussion).

1.8 Aims of this thesis

The research presented in this thesis aims to further explore the issue of social vulnerability. The mixed-methods research presented here uses data from multiple informants where possible and draws comparisons between individuals with WS and their typically developing peers (e.g. in Chapters 3, 5 and 6). Given their opposing social profiles, and the more established literature base associated with ASD, comparisons with autism are also included in the literature throughout (Asada & Itakura, 2012; Brock et al., 2008; Jones et al., 2000; Riby & Hancock, 2008).

The overall aim of the thesis is to explore social behaviour which is likely contributing to the high social vulnerability seen in this population (Jawaid et al., 2012). The first aim is to explore types of social behaviour which could place children with WS at risk during social interactions. The experimental chapters start with Chapter 3 which presents a profile based on parent reports of the social behaviour, anxiety, communication and social vulnerability in WS, which are

constructs that have been introduced in this Chapter. The aim of this Chapter is to provide primary evidence of the atypical social behaviour reported in WS. To progress this further, Chapter 4 explores social approach behaviour and stranger danger awareness through parent interviews. The within-syndrome variability of social approach behaviour highlighted in section 1.4.5 will be explored here within the thematic analysis. Personal space regulation in WS, which has yet to be examined in the literature, is examined in Chapter 5. Here, parental reports and experimental paradigms are used to assess awareness of personal space and preferred interpersonal distance. It has been shown that typically developing children regulate their interpersonal distance based on the familiarity of the person they are approaching (Gessaroli et al., 2013). This effect of familiarity is therefore explored in children with WS using a stop-distance paradigm and the theories proposed in section 1.5 to explain social behaviour in WS are also discussed. Chapter 6 investigates trust in WS, which is central to making any claims about vulnerability in WS (e.g. Martens, Hasinski, Andridge & Cunningham, 2012), and builds on the findings of Chapters 3 – 5. Together, this body of research presents information on the characteristics of the social interactions of young people with WS, which may be acting as a precursor to the high levels of vulnerability reported in WS (Jawaid et al., 2012).

The second aim is to investigate the level of insight that individuals with WS have into their own vulnerability, using the Social Vulnerability Questionnaire (SVQ; Fisher et al., 2012). Previous research by Fisher, Mello & Dykens, (2014) has shown a discrepancy between self-reports, parent reports and behavioural observations. In order to work towards intervention development, it is important

to consider the self-insight of individuals with WS (Emmerson, Granholm, Link, McQuaid & Jeste, 2009). How individuals with WS believe that they are perceived by others could be simultaneously indicative of their resilience and their vulnerability and therefore offers a valuable insight when considering experiences of social vulnerability in this population (Fisher et al., 2014).

The final aim is to offer the first investigations into online vulnerability in adults with WS (see Chapter 2 for a discussion on the merits of including children and adults with WS in this thesis). The distinction between the offline and online world is becoming increasingly blurred (Subrahmanyam & Greenfield, 2008). A review in Chapter 8 outlines what is known about atypical social behaviour in WS, their high levels of social vulnerability, and what this could mean for online vulnerability (e.g. Mazurek, 2013). This paves the way for Chapter 9 which uses a mixed-methods approach to provide descriptive data about how often and why adults with WS use the internet. It also uses a scenario-based task (Wilson, Seaman, & Nettelbeck, 1996) to assess the types of risks that they could be facing when online. An exploratory case study of social vulnerability and online behaviour is presented in Chapter 10 to provide some in-depth information about the experiences of one individual and her family.

Taken together, this thesis provides information relating to social vulnerability in WS derived from mixed-methods approaches. It also offers the first insights into online social behaviour and online vulnerability in adults with WS. The real-world implications of these findings are emphasised throughout. In Chapter 11, the theoretical contributions of this thesis are outlined and a number of

suggestions are made to help subsequent research progress towards intervention development.

1.8 Conclusion

This chapter has introduced relevant experimental studies within the framework of the wider developmental disorders field and provided a theoretical context to support the forthcoming chapters. These issues will be explored in greater detail at relevant points throughout the thesis. The focus of this introductory chapter has been on the social profile associated with WS, as this is highly salient to the investigation of social vulnerability in this population. The exploratory nature of this work requires careful consideration when selecting appropriate methodology, therefore, this thesis will begin (Chapter 2) with an overview of the methodological choices made in the subsequent experimental chapters.

Chapter Two: General Methods

2.1 Introduction

This chapter provides a brief overview of some of the methodological considerations relevant to the current thesis. A variety of methods, informants and age ranges are used throughout the thesis in order to extract the most insightful data to further what is known about social vulnerability in WS. This Chapter outlines some of the methodological issues relevant to the study of social vulnerability in WS and presents the methodological choices that have been made in subsequent chapters of this thesis. It begins with an overview of the terminology used throughout the thesis.

2.2 Terminology

The language used to refer to individuals who experience some form of developmental delay has evolved in recent years. 'Mental retardation' was a term coined by the American Association on Mental Retardation in 1961 to describe individuals with intellectual and/or developmental disabilities. It was subsequently adopted by the American Psychiatric Association (APA) for use in its series of Diagnostic and Statistical Manuals for Mental Disorders (DSM), to replace previously commonly used terms such as 'feeblemindedness' and 'idiocy'. However, over the last decade, the term 'mental retardation' has fallen out of favour, owing to its prejudicial connotations. Instead, the DSM-5, produced by the APA (2013) and the International Classification of Diseases (ICD-11) manual, which will be produced by the World Health Organisation in 2018, opt to use phrases such as 'intellectual disability', 'intellectual and

developmental disorders' (IDD) and 'neurodevelopmental disorders'. Person first language is also now encouraged to show respect for the individual. Throughout the thesis, WS and ASD will therefore be referred to as 'neurodevelopmental disorders' or IDD.

Neurodevelopmental disorders are severe impairments which present early in life, typically through one or more specific cognitive deficit. These impairments are life-long and are not acquired: rather they are present from birth. Some are also accompanied by co-morbid learning difficulties (IDD). The DSM-5 criteria for neurodevelopmental disorders broadly encompasses disorders which, through developmental deficits, produce impairments in a wide range of domains linked to everyday functioning (APA, 2013). The developmental deficits range from domain specific impairments of functioning, (e.g. learning) to global impairments of functioning (e.g. social skills), providing a broad spectrum of everyday functioning. Not all symptoms shown by individuals with neurodevelopmental disorders are centred on delays or deficits, but some may be symptoms of excess. For example, excessive restricted and repetitive behaviours are typically found in ASD and extreme prosocial behaviour is phenotypical of WS.

In the DSM-5 (APA, 2013), the previously used categories of 'autistic disorder', 'Asperger disorder', 'childhood disintegrative disorder' and 'Pervasive Developmental Disorder – Not Otherwise Specified (PDD-NOS)' have been replaced by the term 'autism spectrum disorder (ASD)'. This term encompasses a broad continuum of performance related to the condition. The category of ASD

will therefore be used when referring to participants on the autism spectrum in this thesis.

Learning difficulties (previously referred to as learning disabilities and intellectual disabilities) are defined by the Department of Health (2001) as a reduced ability to compute complex information, learn new skills and cope independently with everyday tasks, as expected for their chronological age. Overall intelligence used to be seen as a central component to identifying learning difficulties (e.g. with the IQ-achievement discrepancy formulae; Frankenberger & Fronzaglio, 1991). However, the identification of learning difficulties has now moved to focus on a low level of proficiency within a learning field irrespective of IQ and IQ-achievement disparities. Learning difficulties are often seen in individuals with ASD, and with WS and can be an important consideration when determining whether participants understand the complexities of a task. As an example of this, receptive vocabulary abilities were assessed using the British Picture Vocabulary Scale (BPVS second edition; Dunn, Dunn, Whetton & Burley, 1997) in Chapter 6 to facilitate the matched groups design (see section 2.6).

2.3 Quantitative and qualitative methods

Quantitative and qualitative methodologies were used in tandem throughout this thesis to provide a multi-methods insight. Quantitative methods have dominated mainstream psychology, arising from an emphasis on the importance of measurement. In this 1930's, the need for scientific rigour in research became paramount and a focus on isolating 'variables' and the use of statistics became

integral to psychological research (Howitt, 2010). It was not until the 1980's that qualitative methodology made significant inroads into being accepted as an important element of psychological research and theory development (Howitt, 2010). According to Denzin and Lincoln (2000), qualitative methodology is concerned with capturing the richness of description, capturing the individual's perspective, rejecting positivism in favour of post-modern perspectives (i.e. data does not show reality, as there are multiple versions of reality), adherence to post-modern sensibility (i.e. researchers should aim to move away from the artificiality of laboratory settings, and instead aim to get close to the real life experiences of individual people) and examination of the constraints of everyday life (i.e. striving to understand how the everyday social world could shape the experiences of participants). Qualitative methodology was used in the current thesis either as an additional methodology to what has been used previously in the literature, offering a novel insight into a heavily research topic, or as an exploratory method of enquiry when there was no previous literature on a topic. In both instances, the aim was to provide in-depth personal experience, providing insight that is not always achievable through experimental design.

However, qualitative methodology has been criticised for lacking reliability and validity (Smith, Flowers & Larkin, 2009). The subjective nature of qualitative analysis and the idea that there exists multiple versions of reality goes against the scientific rigour that psychology promotes (Howitt, 2010). Guba and Lincoln (1982) argued that constructs such as reliability and validity are not crucial features when controlling for the quality of qualitative research. Instead, they proposed that 'trustworthiness' was a key quality marker, containing four aspects:

credibility (confidence that the findings represent the 'truth'), transferability (demonstrating that the findings are relevant to other contexts), dependability (showing that there is some consistency in the findings) and confirmability (the extent to which the findings represent the views of the respondent, rather than researcher bias). These markers can be attained through triangulation, detailed description, and reflexivity.

For the purpose of the current thesis, the following reflexive statement is offered: I begun this work with no prior research experience of issues linked to vulnerability, and I had never met a person with Williams syndrome. My motivation to undertake this work was based on a fascination with the Williams syndrome social profile and an awareness of the real-life applicability that work of this nature could have. The work evolved over the three year period, and did not seek to confirm any pre-held beliefs. Indeed, in the case of Chapter 6, some unexpected results were reported. The thematic analyses carried out in Chapters 4 and 9 were reviewed by multiple coders to minimise researcher bias, although a critical realist stance in the analysis process is acknowledged.

Recently, literature has focused less on the merits of quantitative versus qualitative methods, and has dismissed the idea that quantitative and qualitative methods are distinct and should not be mixed (see 'the incompatibility thesis'; Howe, 1988). Instead, there has been more emphasis on the importance of employing a mixed methods approach (Johnson & Onwuegbuzie, 2004). Johnson and Onwuegbuzie (2004) defined mixed methods research as a form of research that combines quantitative and qualitative research approaches, methods and

concepts in a single study. Chapters 7 embodies this idea by quantitatively analysing closed-response questionnaire data about social vulnerability as well as qualitatively analysing open-ended questionnaire data about personal experiences of victimisation. Mixed research methods are also used across studies within the thesis, using a variety of informants.

2.4 Multi-informant approach

In typically developing children, we know that behaviour varies considerably across interpersonal situations (e.g. Dirks, Treat & Weersing, 2007), and so too do people's perspectives on this behaviour (De Los Reyes & Kazdin, 2005). Multi-informant approaches can therefore produce widely differing reports (Dumenci, Achenbach, & Windle, 2011). This can be problematic for clinical diagnoses of psychopathology, and can create uncertainty around measurement error and reporting biases (De Los Reyes, Kundey, & Wang, 2011). However, rather than seeking to confirm one accurate report of behaviour (Wright et al., 2011), these different reports should in themselves be seen as insightful, as behaviours occurring in different situations, perceived by different informants, could contribute to our knowledge of distinct phenotypes and within-syndrome heterogeneity (Dirks, De Los Reyes, Briggs-Gowan, Cella & Wakschlag, 2012). Parent reports and/or self-reports are therefore used throughout the thesis in order to access multi-informant information about social vulnerability in WS.

Finlay and Lyons (2001) highlighted the widely held assumption by researchers that parents of individuals with IDD can accurately report their son/daughter's thoughts, behaviours and emotions. Parents were seen as vital informants as some

individuals with intellectual difficulties have difficulties responding to complex questions, and show high rates of inconsistent reporting (Finlay & Lyons, 2001). Many individuals with IDD also experience high levels of alexithymia, and struggle to identify and verbalise their thoughts, feelings and emotions (Davies et al., 2015; Mellor & Dagnan, 2005). Indeed, in the WS literature, Fisher, Mello and Dykens (2014) found no agreement between parent report and self-report of adults with WS (mean age = 26.4 years) about social approach behaviour. Self and parent-report approachability scales asked the participant about the likelihood that they/ their son or daughter would approach a person in a given scenario. This was followed by a faces task, in which participants had to indicate whether or not they or their son/daughter would approach faces presented on a computer screen. The authors found that parents reported significantly higher levels of approach behaviour, compared to the reports given by their son/daughter. When these reports were compared to behavioural observations, it was found that parent reports most accurately reflected the behaviour observed, reinforcing the value of parent reports when working with individuals with IDD. Parent reports, either in isolation or alongside self-reports, are used throughout this thesis.

However, more recently, the importance of also gathering information from the individual themselves has been acknowledged (Kassam-Adams, Garcia-Espana, Miller, & Winston, 2006). Chappell (2000) claimed that, "people with learning difficulties are the best people to ask if researchers want to know their views and experiences" (p.40). She highlighted the emerging trend for emancipatory and participatory research, which place individuals with disabilities as central to the research process. A great deal of emphasis was placed on the value of self-report

in this thesis, particularly in Chapter 7. Although self-reports may differ from reports provided by other informants (De Los Reyes et al., 2015), it is nevertheless important to understand how individuals with IDD see themselves, other people, and the events around them. This links to the discussion of the principles of qualitative methodology above, which acknowledges that each individual constructs their own unique version of reality. Increased self-insight has been found to be a key predictor of intervention success in the mental health literature (Emmerson, Granholm, Link, McQuaid & Jeste, 2009), suggesting that the value of self-report should not be dismissed.

2.5 Static-point approach

The research included in this thesis represents a static time point in development and is therefore unable to capture developmental changes over time. When appropriate, comparisons between age groups are made (e.g. in Chapter 5), which offers some insight into the development of the phenomenon of interest. The aim of the studies in the current thesis was to provide initial insights into novel phenomenon which could be relevant to the study of social vulnerability. At this early stage, a longitudinal design could not be advocated in the absence of any supporting evidence. Further, a cross-sectional design would require wide age ranges to reduce the risk of floor and ceiling effects (Thomas, Purser & Van Herwegen, 2012), and, given what we know about heterogeneity in WS (e.g. Little et al., 2013; Porter & Colheart, 2005; Porter et al., 2007), the assumption of cross-sectional designs that all individuals with WS will follow the same trajectory is troublesome. Instead, a static point approach was selected, which

used a relatively narrow age range in each study, and a matched groups design (as outlined in section 2.6) in several of the chapters (e.g. Chapters 3, 5, 6, 10).

2.6. Matched groups design

The use of appropriate control groups when working with people with WS is highly important (Landau & Ferrara, 2013). A factorial matching paradigm was used in this thesis to examine differences between participant groups (Hermelin & O'Connor, 1970; Baddeley & Gathercole, 1999). This design enables us to highlight areas of functioning which differ to 'typical' performance for their chronological age or level of intellectual ability. Research on issues related to vulnerability in WS have typically contrasted performance to groups of typically developing (TD) individuals, either matched on mental age (e.g. Tager-Flusberg et al., 2003), chronological age (e.g. Tager-Flusberg, Plesa-Skewer, Faja & Joseph, 2000) or to other individuals with neurodevelopmental disorders (e.g. Wang, Doherty, Rourke & Bellugi, 1999; Doyle, Bellugi,Korenberg & Graham, 2004). This section will outline the different types of matched groups designs used in this thesis and the justification for their selection.

2.6.1 Comparisons between WS and typical development

Typically developing individuals were used as the primary comparison group in this thesis (e.g. in Chapters 3, 5 and 6) to address questions of typicality. In order to better understand the extent to which the behaviours displayed by individuals with WS are divergent from the norm, typical performance must be established. One method of matching participants is based on their chronological age (e.g. Bishop, 1997; Bellugi, Wang & Jernigan, 1994; Tager-Flusberg et al., 2003). This

provides some insight into how the performance of individuals with WS compares to the level expected based on the chronological developmental trajectory shown by their typically developing peers. For example, Tager-Flusberg et al. (2003) examined the performance of individuals with WS (mean age = 20.1 years) on a face processing task (the part-whole paradigm; Tanaka & Farah, 1993) compared to chronologically age matched typically developing controls (mean age = 19.5 years). They found that adolescents and adults with WS showed a whole-face advantage for upright faces but not inverted faces. The inclusion of a chronologically age matched control group here allowed them to argue that the results suggested that people with WS process faces 'normally'. Matching on chronological age also acknowledges that more years of experience can impact upon performance (Jarold & Brock, 2004). The decision was made to match individuals with WS to typically developing individuals based on their chronological age, rather than their mental age in the chapter on personal space (Chapter 5). This is because we were interested in the initial level of vulnerability derived from their approach behaviour and interpersonal distance. The unfamiliar person that they are approaching only had an estimate of their chronological age available at this stage guide their response to the approach.

Matching groups based on intellectual ability is a common matched groups design in research with individuals with IDD (see Mottron, 2004 for a meta-analysis of matching procedures in autism). One of the most common measures of functioning identified by Mottron (2004) was the BPVS (Dunn et al., 1997; used in 23.3% of autism studies reviewed). Given the paradoxical cognitive profile associated with WS (e.g. Martens et al., 2008; Mervis et al., 2000)

matching participants on overall level of function would likely be misleading, due to the dissociations seen between verbal and non-verbal performance (e.g. Klein & Mervis, 1999). It was therefore decided to match participants on the aspect of functioning deemed most salient to the task. Chapter 6 matched individuals with WS to a group of chronologically age matched typically developing controls and a group of typically developing controls matched on their receptive vocabulary skills (as measured by the BPVS II, Dunn et al., 1997) to investigate trust behaviour.

The BPVS II (Dunn et al., 1997) assesses receptive vocabulary abilities, and provides raw and standardised scores which link to an equivalent mental age (given in years and months). This task is suitable for varying levels of ability, as it has been designed for use in 3 – 16 year olds. In the task, the experimenter says a word and presents four pictures to the child. The child must select which of those four pictures best corresponds to the word spoken. A basal set is first established by finding the set of words in which the participant gets no more than one answer incorrect. The child then works through each subsequent set which increase in difficulty, until he/she makes eight errors in a set and this is established as the ceiling set. The BPVS II has frequently been used in individuals with WS to assess intellectual functioning (e.g. Howlin, Davies & Udwin, 1998; Karmiloff-Smith et al., 2004; Annaz, Karmiloff-Smith, Johnson & Thomas, 2009; Laing, Hulme, Grant & Karmiloff-Smith, 2001).

2.6.2 Comparisons between WS and other IDD

The inclusion of comparison groups with IDD allow for identification of syndrome specific patterns of performance, rather than behaviours which are a characteristic of more general developmental delay (Burack et al., 2002). Early work by Bellugi and her team (e.g. Wang & Bellugi, 1999) favoured comparing the performance of individuals with WS to individuals with Down syndrome, as both groups were supposed to have comparable mental ages due to the mild – moderate levels of intellectual difficulties seen in both syndromes. This allowed them to investigate the extent to which spatial abilities were syndrome specific, rather than generic to people with learning difficulties (Burack et al., 2002). Given the prosocial behaviour seen in WS, Down syndrome has been a frequently selected comparison group. For example, Doyle et al. (2004) compared the social behaviour (as measured by the Salk Institute Sociability Questionnaire; Jones et al., 2000) of 3 – 9 year olds with WS and Down syndrome. They reported elevated and distinct patterns of social behaviour in the WS group, compared to the DS group. They argued that these differences in hypersociability are unlikely to be the result of cognitive impairments which lead to a lack of understanding of social norms, as both groups had cognitive impairments. Instead, the authors suggested that individuals with WS may have an innate predisposition towards hypersociability, and in particular, increased approach to strangers. Here, the inclusion of another developmental disorder participant group allowed for these syndrome-specific conclusions to be drawn.

More recently, attention has shifted to comparing and contrasting the social profiles associated with WS and ASD (e.g. Asada & Itakura, 2012; Brock et al., 2009), and it is this comparison that is of particular interest to the current thesis. It

has been widely proposed that WS and ASD display polar opposite profiles, especially in the domain of social functioning (Courchesne, Bellugi, & Singer, 1995). Jones et al. (2000) highlight that many individuals with WS show high levels of prosocial behaviour which they seemingly find difficult to inhibit, whereas individuals with ASD tend to avoid interactions with others. These opposing social profiles can be illustrated through work on face processing. Riby and Hancock (2008) showed photographs of people to individuals with ASD (mean age = 13 years), individuals with WS (mean age = 17 years) and individuals matched for chronological age and non-verbal ability. Using eye-tracking techniques, they found that individuals with ASD spent less time looking at the faces in the photographs compared to the control groups, whereas individuals with WS spent more time viewing the faces. Specifically, individuals with ASD spent less time looking at the eye region, whereas individuals with WS spent more time attending to this region when compared to the control groups. These insights into the differing socio-communicative patterns seen in these groups have important implications when considering the "uniqueness" of the social vulnerability experienced by individuals with WS.

However, more research has started to also emphasise the similarities between individuals with WS and ASD (e.g. Lincoln et al., 2007; Klein-Tasmin et al., 2009), though direct cross-syndrome comparisons are still relatively rare (Asada & Itakura, 2012). Klein-Tasmin et al. (2009) used the Autism Diagnostic Observation Schedule (ADOS; Lord et al., 1999) Module 1 in children with WS. They found that 10% of the children were classified as having 'autism' and 40% as being on the 'autism spectrum'. They found that whilst the socio-

communicative impairments in the WS group were not as substantial as those seen in individuals with autism, they were comparable to the PDD-NOS group. This suggests that, despite their differing social behaviour, some individuals with WS also have traits which are indicative of an autism spectrum disorder. This is likely to be explained by the considerable amount of within-syndrome heterogeneity in WS (e.g. Porter & Coltheart, 2005; see Chapter 1 for further discussion) as well as the broad spectrum of abilities seen in ASD (Joseph, Tager-Flusberg & Lord, 2002). This complex profile of similarities and differences therefore makes ASD a particularly interesting comparison group, especially as individuals with WS and ASD have both been shown to experience social vulnerability (Fisher et al., 2013). Chapter 5 includes TD and ASD control groups, both matched on chronological age, to investigate personal space violations in WS.

2.7 Syndrome specific studies

Some chapters in the thesis focus solely on WS (Chapters 4, 7, 9 and 10), and investigate in-depth behavioural phenotypes associated with the condition. In these chapters, it was decided that the inclusion of a TD comparison group would be inappropriate. For example, the comprehensive interview presented in Chapter 4 on social approach behaviour and stranger danger awareness (mean age of participants = 9.8 years) covered issues that are not relevant to the majority of the TD population of this age (awareness of strangers has been found in TD children as young as 6 – 12 months; Rheingold & Eckerman, 1973). Instead, the aim was to better understand these phenomenon in WS and the individual experiences of people with WS and their families. This approach is not novel in

the WS field. For example, Little et al. (2013) investigated social approach behaviour and the role of inhibition in twenty-five children with WS and did not include any comparison groups. The aim of the study was to look within-syndrome at heterogeneity of behaviour within one syndrome, which is similar to the aims of the research presented in Chapter 4.

Syndrome specific studies are more common in qualitative work than quantitative work (e.g. Jones, Quigney & Huws, 2003; King, Zwaigenbaum, King, Baxter, Rosenbaum & Bates, 2006; Woodgate, Ateah & Secco, 2008; Kuhaneck, Burroughs, Wright, Lemanczyk & Darragh, 2010) and do not share the aims of identifying issues of typicality or uniqueness that are central to matched groups studies. For example, Woodgate et al., (2008) used hermeneutic phenomenology to explore the experiences of parents who had a child with autism. They uncovered themes such as 'living in a world of our own,' 'society's lack of understanding' and 'feeling disconnected from the family'. Here, the authors were concerned with the experience of individuals in their idiographic approach, rather than between group comparisons. Chapters 9 and 10 used a syndrome specific design, in order to investigate a topic which had previously not examined in WS. By only including participants with WS, we were able to build up a more comprehensive picture of computer use and internet safety in this group, which could form the basis for future matched group comparisons. For the purposes of this thesis however, it acted as an appropriate method of exploratory enquiry.

2.7.1 Case study approach

Case study approaches often focus on one participant group, rather than making between group comparisons. A case study approach is typically an exploratory method of research enquiry, in which real-life phenomenon can be explored in great detail within the context in which it exists (Yin, 1984). Simons (1996) highlights that a case study approach can be beneficial when seeking to better understand the particular, unique experiences of individuals; but that the importance of generalisation, comparability and certainty must be relegated. Indeed, according to Flyvbjerg (2006), frequent criticisms of the case study approach include i) difficulties generalising from a single case which impacts upon contributions to scientific development, ii) the case study method can only serve to generate hypotheses, rather than to test hypotheses and contribute to our theoretical understanding and, iii) there exists a researcher bias towards verification. However, Gerring (2004) argues that case studies look to make comparable links to similar others, rather than seeking generalisation, though he concedes that the exploratory nature of case studies often presents intrinsic challenges to demonstrating the falsification of hypotheses. In Chapter 10, a qualitative case study was used to investigate internet use in WS by speaking to an adult with WS and her mother. The aim of this was to build on anecdotal reports from parents about internet safety concerns by gaining an in-depth understanding of the experiences of an individual to stimulate further investigation on this topic.

2.8 Applicability of research measurements

There has been an emerging body of literature on the suitability of measures for individuals with IDD which have been developed for use in the typically

developing population (e.g. Rodgers et al., 2016). At several points throughout the current thesis, there is a discussion about why the measurements used were developed, and for whom. For example, in Chapter 3, we discuss the relevance of the Social Communication Questionnaire (SCQ; Rutter, Bailey & Lord, 2003), which was initially developed as a screener for autism, to WS and the typically developing population. Similarly in Chapter 5, it is acknowledged that the Social Responsiveness Scale (SRS; Constantino & Gruber, 2005) was not developed as a measure of personal space, though the items relating to personal space are still of interest.

One area which has stimulated a great deal of research in recent years is the measurement of anxiety in individuals with developmental disorders, with the characteristics associated with ASD being used to highlight measurement issues (Wood et al., 2015). The process of disentangling the characteristics associated with having an ASD diagnosis from those seen in anxiety can be challenging (Gjevik, Eldevik, Fjaeran-Granum, & Sponheim, 2011). For example, restricted and repetitive behaviours may be difficult to identify as being distinct from compulsive behaviours seen in some anxiety sub-types (e.g. OCD; Zandt, Prior, & Kyrios, 2009), and the social withdrawal seen in ASD could be challenging to differentiate from elements of social anxiety (e.g. Pellecchia et al., 2015). A recent systematic review by Wigham and McConachie (2014) of the measurement tools used in cognitive behavioural therapy trials for anxiety in children with ASD found only three questionnaire measures to be robust in this population: The Spence Children's Anxiety Scale (SCAS; Spence, 1998), the Revised Children's Anxiety and Depression Scale (RCADS; Chorpita, Yim,

Moffitt, Umemoto, & Francis, 2000), and the Screen for Child Anxiety Related Emotional Disorders (SCARED; Birmaher et al., 1999). The SCAS (Spence, 1998) has been widely used with individuals with WS (e.g. Rodgers et al., 2012, Riby et al., 2014). This, combined with the suggestions of Wigham & McConachie (2014), meant that the SCAS (Spence, 1998) was used as the primary measurement of anxiety in the current thesis. However, the appropriateness of this measure and others is reflected on again in the thesis limitations in Chapter 11.

2.9 Participant characteristics

The characteristics of the samples included in the current thesis differ between studies. It is therefore important to provide some relevant information on the samples included, to allow for informed interpretation of the findings presented.

2.9.1 Recruitment

The investigations in the current thesis involving children with WS were conducted in the UK (age range 6 – 16 years), and those involving adult participants were primarily conducted in the USA. The exceptions to this are the internet use case study (Chapter 10) which involved an adult participant but was conducted in the UK, and the first experiment in Chapter 5, which was a multi-site project involving participants from the UK, Ireland, the USA and Australia. For the studies carried out in the UK, all participants were recruited through the Williams syndrome Foundation (WSF). The adult WS sample was collected in the USA through the Williams syndrome Association (WSA) and at a residential summer camp in conjunction with the Vanderbilt Kennedy Center.

Some participants took part in more than one study in this thesis (as outlined in Table 2.1). Given the recruitment challenges associated with a rare condition like WS, some of the same individuals were recruited for several different studies due to their availability and willingness to participate in research. This overlap of participants between Chapters should be considered when interpreting the results and considering the wider social behaviour profile of individuals with WS.

Table 2.1. A summary of the number of participants who took part in multiple studies

	Participant overlap
Chapter four	Five out of 23 participants in this Chapter also took part in Chapter three (representing 22% overlap in participants)
Chapter five *(Paper one)*	Seven out of 77 participants in this Chapter also took part in Chapter three (representing 9% overlap in participants). Sixteen out of 77 participants in this Chapter also took part in Chapter four (representing 21% overlap in participants).
Chapter five *(Paper two)*	Five out of 18 participants in this Chapter also took part in Chapter three (representing 28% overlap in participants) Seven out of 18 participants in this Chapter also took part in Chapter four (representing 39% overlap in participants).

	Nine out of 18 participants in this Chapter also took part in Chapter five (Paper one; representing 50% overlap in participants)
Chapter six	All of the participants (n = 18) took part in the study in Chapter five (Paper two)
Chapter nine	All of the participants (n = 28) took part in the study in Chapter seven.

2.9.2 Sample size

The importance of sample size in psychological research has attracted a great deal of attention (e.g. Holmes, 1979, 1983; Holmes, Holmes, & Fanning, 1981; Cochrane & Duffy, 1974; Marszalek, Barber, Kohlhart & Holmes, 2011). As a result of concerns about methodological issues which threatened the credibility of psychology as a science, the Task Force on Statistical Inference was formed by the American Psychological Association (APA) in 1996 (Wilkinson, 1999). One issue they addressed was sample size; as small sample sizes in leading journals were said to be affecting generalisability, estimation of effect sizes and the development of recommendations for future studies. After the Task Force was established, Marszalek et al. (2011) reviewed the samples sizes in four leading journals from 1995 to 2006. They found that, overall, sample sizes did not differ over time, and remained relatively small. However, there was variability between fields, with a slight increase in sample size being found in *Developmental Psychology.* They highlighted that accessibility to research populations could be

one barrier to increasing sample size, something which is relevant when conducting research on WS. The sample sizes used in research involving individuals with WS are typically relatively small (18 – 25 individuals; e.g. Riby & Hancock, 2008; Rhodes, Riby, Park, Fraser & Campbell, 2010; Tager-Flusberg & Sillivan, 2000; van der Fluit, Gaffrey & Klein-Tasman, 2012).

Linked to sample size is the issue of power, which Rossi (1990) found had remained low since the development of Cohen's study surveying power in 1962. Maddock and Rossi (2001) reviewed the statistical power reported in three volumes of health psychology journals. They found that the statistical power in studies which reported a small effect size to be on average 0.36, which is considerably lower than the minimum value of 0.8 originally suggested by Cohen (1988). Marszalek et al. (2011) suggested that small sample sizes are relevant when considering why power has failed to improve; "Increased sample size is likely to prove the most effective general prescription for improving power" (Cohen, 1962, p. 153). When an increased sample size is not possible, such as in the second half of Chapter 5, then an a priori power analysis was used to ensure that the study had sufficient power to detect a large effect size.

2.9.3 Confirmation of diagnosis

All participants who took part in the research had a diagnosis of WS. For most, this was obtained through genetic testing using fluorescent in situ hybridisation (FISH) testing which was outlined in Chapter 1. However, as this genetic testing is a relatively recent advancement, some of the adult sample (in Chapters 7, 9 and 10) had their diagnosis confirmed through clinical assessment of the physical,

behavioural and cognitive characteristics linked to WS, rather than through FISH testing.

2.9.4 Participant age

The age of participants in the samples included throughout the thesis range from 4 to 54 years old. This is because some of the lines of enquiry are more relevant to a particular age range. As an example, pilot testing showed that internet safety was not a concern for parents of children aged 8 – 16, as their son/daughter spent little time on the internet unsupervised. Whereas, in the adult population, anecdotal parent reports highlighted that this was a highly pertinent issue for the older age range, as they become more independent but also more socially isolated in the real world at the same time. The investigations included within this thesis do not attempt to track the developmental trajectory of the issues of interest. Instead a static time point approach was used (as outlined in section 2.5). However, where relevant, the impact of age is explored (e.g. in Chapter 5). Despite selecting a matched groups static point approach, we are aware that the phenomenon investigated occur within the wider context of development (as discussed in Chapter 1).

2.10 Ethics

Ethical approval was sought for all studies included in this thesis. The table below (Table 2.2) outlines the institutional boards which awarded ethical approval for each empirical study.

Table 2.2. Record of ethical approval for each study in the current thesis

	Title	Approved by
Chapter three	A profile of social behaviour, communication, anxiety and vulnerability in Williams syndrome	Durham University Psychology Ethics Committee
Chapter four	Parent insights into atypicalities of social approach behaviour in Williams syndrome	Durham University Psychology Ethics Committee
Chapter five *(Paper one)*	Violations of personal space in young people with Autism Spectrum Disorders and Williams syndrome: Insights from the Social Responsiveness Scale	Durham University Psychology Ethics Committee
Chapter five *(Paper two)*	Personal space regulation in Williams syndrome: The effect of familiarity	Durham University Psychology Ethics Committee
Chapter six	Examining trust behaviour in young people with Williams syndrome	Durham University Psychology Ethics Committee
Chapter seven	Parent and Self-Report Ratings on the Perceived Levels of Social Vulnerability of Adults with Williams Syndrome	Vanderbilt University's Institutional Review Board

Chapter nine	Internet Use and Online Safety in Adults with Williams Syndrome	Vanderbilt University's Institutional Review Board
Chapter ten	A case study analysis of social behaviour and internet use in Williams syndrome	Durham University Psychology Ethics Committee

All work was carried out in accordance with the British Psychological Society's Code of Human Ethics (2010) and the European Federation of Psychologists' Associations (EFPA) meta-code of ethical guidelines (2005). Information sheets were provided for each study and informed consent was obtained for all participants. It was made clear that participation was on a voluntary basis, and there would be no adverse consequences should they decide not to take part. Where applicable, informed assent was also obtained from children who were under the age of 18 years old. If informed consent was obtained from the parent, but informed assent was not obtained from the child, then the child did not participate in the research. Participants were informed that all data collected would be anonymised to remove any identifiable information. For the qualitative studies, participants were asked to give their consent for extracts of the interview to be presented at conferences and written up for publication. Participants were made aware that they could withdraw from the study at any time, without reason, and a full debrief was given at the end of each study.

2.11 Conclusions

Research with a rare population such as WS involves many methodological considerations which have been outlined in this chapter. The experimental chapters of this thesis will now follow, beginning with Chapter 3 which employed a matched groups design to look at parent reports of some of the key characteristics associated with WS, at a static time point.

Chapter Three: A profile of social behavior, communication, anxiety and vulnerability in Williams syndrome

Chapter 3 begins the experimental work of this thesis by providing an overview of social behaviour, communication, anxiety and vulnerability in a sample of individuals with WS. These constructs have been introduced in Chapter 1, and have been selected for further investigation in this chapter because of their relevance to understanding the wider social profile of typicalities and atypicalities that we associate with WS. By studying the range of concepts within one group, the chapter takes a multi-methods explorative approach (see Chapter 2) and offers tentative insight into the potential relationships and interactions between these issues for individuals with WS. This Chapter contributes to the literature by profiling these key constructs in an adolescent sample, as adolescence is known to be a challenging transitional period for young people with WS which could have implications for their vulnerability status.

3.1 Introduction

As emphasised in Chapter 1, the social profile associated with WS is of particular interest to the research focus of this thesis. Individuals with WS are known to be hypersociable; they experience an exaggerated prosocial desire to interact with other people (Frigerio et al., 2006), including strangers (Jones et al., 2000). Studies of social approach behaviour in WS have shown that individuals with WS indicate that they would be more willing to approach both trustworthy and untrustworthy faces compared to typically developing (TD) individuals (Jones et al., 2000). Martens, Hasinski, Andridge and Cunningham (2012) used computer mouse tracking technology to show the nature of approach decisions to computer generated faces which had been pre-rated on trustworthiness. Participants with WS (mean age = 20 years) were significantly more likely than the chronologically age matched control group to approach untrustworthy faces. They were also significantly more likely to consider approaching untrustworthy faces, even if they eventually decided to avoid the face. Further, Frigerio et al., (2006) found that it was faces which displayed positive emotions which individuals with WS (mean age = 16.5 years) rated as being most approachable, suggesting a link to emotion in social approach decisions (supported by Porter et al., 2007).

Individuals with WS also lack stranger danger awareness (Fisher, 2013). Using a series of video vignettes, Riby et al. (2014) found that when children with WS (mean age = 12.1 years) indicated that they would interact with a stranger, they showed a lack of understanding of the dangers posed by the interaction. Showing the benefit of multi-informant information and mixed methodologies, it was those individuals who were more likely to engage with strangers who were rated by

their parents as showing more atypical social behavior. Interestingly, Jones et al. (2000) found that atypical social behaviour is evident in infants, toddlers, school aged children and adults with WS. General social functioning therefore has important consequences for the daily lives of individuals with WS across the developmental spectrum.

Interestingly, this prosocial behavior exists alongside high levels of anxiety (Rodgers et al., 2012). As noted in Chapter 1, anxiety is often considered the most significant mental health challenge faced by individuals with WS, certainly by adulthood (Stinton et al. 2010). Recent work by Riby and colleagues (2014) found that 46% of the individuals with WS in their study (n = 59; age range 6 – 36 years) experienced high levels of anxiety. The mean anxiety scores for the high anxiety WS group were greater than those reported for clinically anxious children (Nauta et al., 2004). They also found that individuals with high versus low levels of anxiety displayed distinctly different social profiles; differing on aspects of social awareness, social cognition, and social communication as measured by parent reports on the Social Responsiveness Scale (SRS; Constantino & Gruber, 2005). Those who experienced the highest levels of anxiety were found to also be most impaired in their reciprocal social behaviour. This shows the within-syndrome heterogeneity that exists in WS, and the importance of taking measures of anxiety when assessing social behaviour.

Emphaising the applied implications of the social profile we link to WS, despite their gregarious and sociable personality (Ng, Jarvinen & Bellugi, 2014), many individuals with WS struggle to form and maintain peer relationships throughout

development and experience high levels of isolation in adulthood (Davies et al., 1998). Adolescence has been identified by parents as being a particularly challenging transitional period for young people with WS, as independence increases and social relationships become more complex (Lough et al., 2016). When considering their intellectual impairment, extreme prosocial drive, high levels of anxiety and social isolation, it is clear why individuals with WS are thought to be socially vulnerable in their everyday lives (for a review, see Jawaid et al., 2014).

The aim of this chapter is to profile the social behavior, social communication, anxiety and social vulnerability status of a small sample of young people with WS compared to their typically developing peers (therefore also exploring typicality of abilities / dysfunction). Several studies have examined these characteristics in isolation (e.g. Jones et al., 2000; Riby et al., 2014; Rodgers et al., 2012), but to our knowledge, no published study to date has taken these four measures from one sample allowing the opportunity to look at the relationship across these areas of functioning. An age range of 8 – 16 years was selected to examine these issues and the relationship between them before adulthood, when the claims of social isolation are most prevalent (Davies et al., 1998). There were three research questions. First, will individuals with WS score atypically on measures of social behavior, anxiety, communication and vulnerability (compared to their typically developing peers)? Second, will those individuals with WS identified as being especially vulnerable experience significantly higher levels of anxiety, as well as more atypical social behavior and communication compared to those deemed relatively low in vulnerability? Third, do scores on these four

measures correlate with one and other for individuals with WS? It was predicted that the WS group will score significantly higher than the TD group on all four measures (research question 1), and that those identified as experiencing high levels of vulnerability will show a different social and anxiety profile to those who display low levels of vulnerability (research question 2). Finally, it was hypothesised that scores on all four measures would be highly correlated (research question 3).

3.2 Method

3.2.1 Participants

Parent reports were obtained for 23 individuals with WS (mean age = 12.4, SD = 2.98, range = 8.1 – 16.8, 39.1% male) and for 29 typically developing children (mean age = 13.6, SD = 1.45, range = 11.3 – 16.5, 58.6% male). There was no significant difference of chronological age between the groups of young people for whom parental reports were obtained ($t(50) = -1.7$, $p = 0.09$). Parents of individuals with WS were recruited through the Williams Syndrome Foundation UK. All children with WS were required to have previously had their diagnosis confirmed through positive genetic florescent in situ hybridisation testing (FISH), detecting the deletion of one copy of the elastin gene on chromosome 7 as the genetic marker of the disorder. Any participants who had co-morbid diagnoses of an Autism Spectrum Disorder (ASD) or Attention Deficit Hyperactivity Disorder (ADHD) were excluded from the study (n = 1). One TD participant was excluded (female, 11 years old) due to a Spence Children's Anxiety Scale (SCAS; Spence,

1998) score of 36, which is above the recommended threshold for clinical diagnosis (24; Nuata et al, 2004).

3.2.2 Materials

Social Responsiveness Scale (SRS; *Constantino & Gruber, 2005*)

The SRS is 65-item parent report standardised questionnaire, measuring the typicality / atypicality of social functioning. This measure has previously been reported to show good psychometric properties, including an internal consistency of .93 (Constantino & Gruber, 2005). Items are coded on a 0-3 scale which generates a total score as well as scores on five subscales. The subscales are: social awareness (e.g. is aware of what others are thinking or feeling), social cognition (e.g. takes things too literally and doesn't get the real meaning of a conversation), social communication (e.g. has trouble keeping up with the flow of a normal conversation), social motivation (seems self-confident when interacting with others) and autistic mannerisms (e.g. behaves in ways that seem strange or bizarre). Raw scores are converted into T scores, with higher T scores being indicative of greater levels of impairment. Total T scores (range = 34 to >90) can be used to classify individuals as showing "normal" social functioning (i.e. scores of 59 or less), "mild to moderate" deficits in social behaviour (i.e. scores of 60 – 75) or "severe interference in everyday social functioning" (i.e. scores of 76 and above).

Social Communication Questionnaire (SCQ; *Rutter, Bailey & Lord, 2003)*

The SCQ (current form) is a 40 item measure completed by parents/caregivers, designed to assess normality/abnormality of social communication (range of

scores = 0 = 39). Parents must read each item, and select a "yes" or "no" response. It was originally designed to screen for autism spectrum disorder (ASD) symptomology (Chandler et al., 2007), but has since been used in a variety of typical and atypical populations (e.g. Howlin & Karpf, 2004). Based on responses, a total score is produced, as well as scores for 3 subdomains: "reciprocal social interaction", "communication" and "restricted, repetitive and stereotyped patterns of behaviour". Higher scores (i.e. scores of 15 and above) are linked to more atypical styles of social communication, and are indicative of a possible diagnosis of ASD.

Spence Children's Anxiety Scale – Parent version (SCAS-P; *Spence, 1998*)

The SCAS-P was used to assess symptoms of anxiety. It has been used in typically developing and clinically anxious populations (e.g. Nauta et al., 2004; Spence, 1998), as well as with individuals with WS (e.g. Rodgers et al., 2012, Riby et al., 2014) . In this 38-item questionnaire, parents are asked to rate statements on a 4-point Likert scale ("never", "sometimes", "often" and "always"). Their answers are scored from 0 to 3, yielding a maximum possible score of 114. These scores produce overall scores on six subscales relating to anxiety: panic/agoraphobia (9 items; e.g. suddenly starts to tremble or shake when there is no reason for this), separation anxiety (6 items; e.g. feels afraid of being on their own at home), physical injury fears (5 items; e.g. scared of the dark), social phobia (6 items; e.g. feels afraid when they have to talk in front of the class), obsessive compulsive (6 items; e.g. can't seem to get bad or silly thoughts out of their head), and generalised anxiety disorder (6 items; e.g. worries about things). While there is no formal clinical cut-off for the SCAS-P, total

SCAS-P scores of 24 or above have been suggested to indicate clinical levels of anxiety (Rodgers et al., 2012).

Social Vulnerability Questionnaire- Revised (SVQ; *Fisher, Moskowitz & Hodapp, 2012*)

The SVQ is a 31-item parent report measure of vulnerability. Parents are asked to rate statements on a scale of 1 – 4 of "not true or never", "somewhat true or rarely", "true or sometimes" and "very true or always". This generates a total score (maximum score = 120), and scores on six sub-domains: emotional abuse (e.g. people try to hurt his/her feelings), risk awareness (e.g. he/she can recognise potentially dangerous situations), social protection (e.g. he/she is isolated from their peers), perceived vulnerability (e.g. others consider him/her to look different from same age peers), parental independence (e.g. you are happy to leave him/her alone for an extended period of time) and credulity (e.g. others perceive him/her to be easy to take advantage of). There are currently no markers of severity available for this measure.

3.2.3 Procedure

Parents were asked to complete the questionnaires, and returned them to the researcher using the stamp-addressed envelope provided. Favourable ethical approval was granted from the local ethics committee prior to the research commencing.

3.3 Results

3.3.1 Social Responsiveness Scale (SRS)

The findings showed that individuals with WS had significantly higher mean SRS Total T scores than the TD group (see Table 3.1), showing that as a group, the individuals with WS exhibit, on average, severe impairments of social functioning. The TD group had mean total scores within the normal range. The scores for the WS group were also significantly higher than the TD comparison group in all five of the subscales generated by the SRS. Indeed, the mean scores for the group with WS were indicative of 'severe' atypicalities of social functioning in three out of the five subscales (cognition, communication and mannerisms).

Table 3.1. SRS T scores and standard deviations for individuals with WS and TD individuals.

	WS	TD	p
SRS Total	79.26 (±13.65)	47.79 (±9.12)	0.001
Awareness	65.91 (±16.9)	47.48 (±8.79)	0.001
Cognition	80.96 (±10.78)	44.72 (±7.24)	0.001
Communication	76.35 (±14.55)	48.14 (±11.14)	0.001
Motivation	63.39 (±13.64)	51.62 (±8.97)	0.001
Mannerisms	83.57 (±11.88)	47.14 (±9.34)	0.001

Scores of 76 and higher indicate severe atypicalities in a domain of social functioning. T scores of 60 – 75 suggest a mild-moderate level of impairment, and scores of 59 and under are in the normal range of functioning.

It is clear from the standard deviations presented in Table 3.1 that there is a great deal of within-group variability in the Total T score, and indeed all five of the

sub-domains. Figures 3.1 and 3.2 below shows the percentage of each group who scored within each degree of functioning (severe, mild/moderate and normal). 69.6% of the WS group received overall scores indicative of severe atypicalities in social functioning, compared to just 4.4% of the TD group. Only 17.4% of the WS group were in the 'normal' range of social functioning based on their Total T scores, with only 13% within the 'normal' range for social awareness, 8.7% for social cognition, 17.4% for social communication, 39.13% for social motivation and 8.7% for social mannerisms. This shows that it was only on the social motivation subscale that more than 20% of participants showed 'normal' functioning.

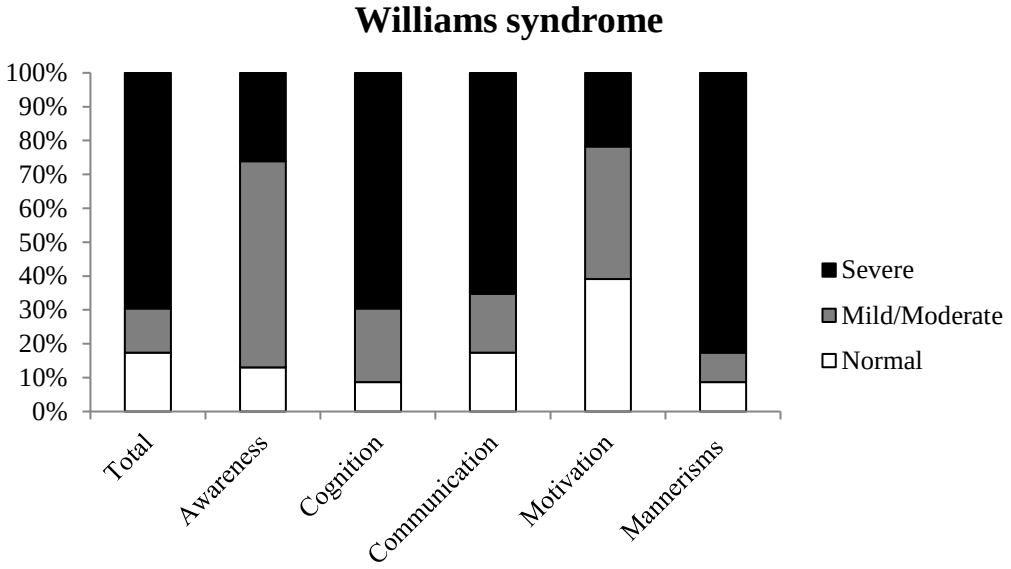


Figure 3.1. Distribution of classifications on the SRS for individuals with WS.

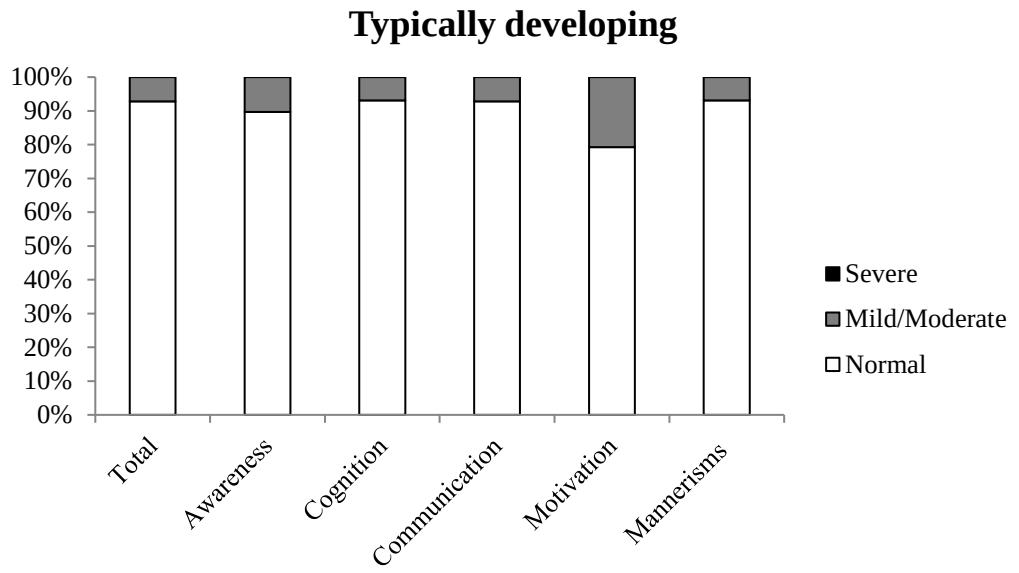


Figure 3.2. Distribution of classifications on the SRS for individuals who are typically developing.

3.3.2 Social Communication Questionnaire (SCQ*)*

On the SCQ, the WS group had a significantly higher mean total score (M = 14.13, SD = 4.38) than the TD group (M = 5.07, SD = 3.52; t(50) = 8.27, p<0.001). As the cut-off score for the SCQ is suggested to be 15 and above, the mean of the WS group was nearing the marker for severe atypicalities of social communication. There were also significant between group differences found when looking at the subdomains of reciprocal social interaction (WS: M = 4.09, SD = 2.21; TD: M= 1.21, SD = 1.86; t(50) = 5.1, p <0.001) , communication (WS: M = 5.91, SD = 1.62; TD: M = 3.55, SD = 1.84, t(50) = 4.84, p <0.001) and on the restricted, repetitive behaviour subdomain, (WS: M = 3.39, SD = 2.35; TD: M = 0.2, SD = 0.62, t(50) = 7.01, p<0.001). Individual level data (see Figure 3.3) shows the variability within the WS and TD samples.

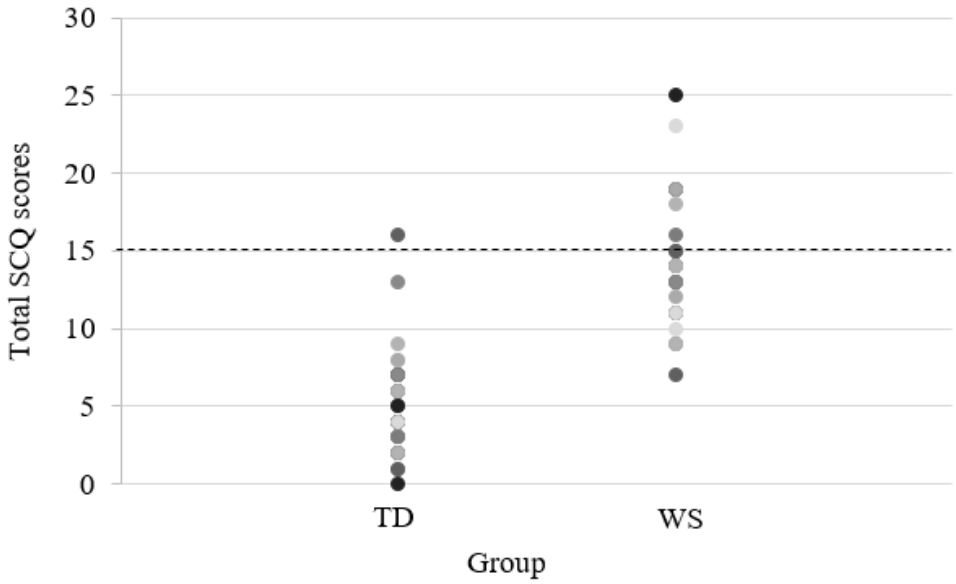


Figure 3.3. Individual total SCQ scores for the TD and WS groups.

3.3.3 Spence Children's Anxiety Scale – Parent version (SCAS-P)

Scores on the SCAS-P show that individuals with WS had significantly higher mean total scores compared to the TD group (see Table 3.2). Indeed, the mean total score for the WS group was indicative of clinical levels of anxiety (>24) but the mean score for the TD group was significantly below this cut off value ($t(28) = 6.51$, $p<0.001$). Figure 3.4 shows the distribution of the individual level data, to highlight the variance within the groups.

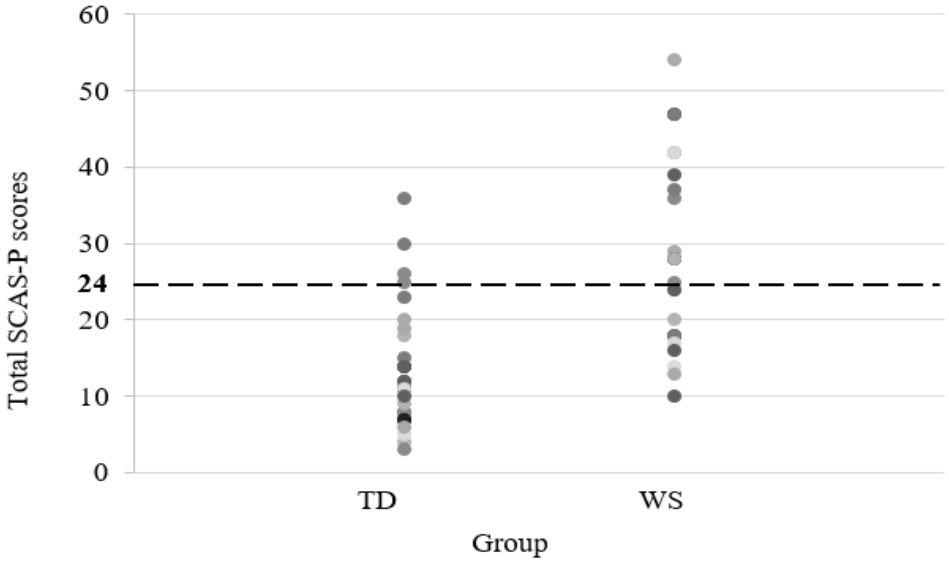


Figure 3.4. Individual total SCAS-P scores for the TD and WS groups.

The individuals with WS also scored significantly higher on five of the six subdomains. Interestingly, the only subdomain where there was no significant difference between groups was the area of social phobia. Indeed the TD group scored higher on this subscale than the WS group, though this was non-significant ($p=.42$).

Table 3.2. SCAS-P raw scores

	WS	TD	p
SCAS Total	28.52 (±12.37)	14.1 (±8.18)	<0.001
Panic	4.09 (±2.97)	1.07 (±1.67)	<0.001
Separation anxiety	5.35 (±1.62)	3.69 (±1.66)	<0.001
Physical injury fears	4.9 (±2.46)	2.55 (±2.2)	<0.001
Social phobia	4.52 (±3.31)	5.21 (±2.74)	0.42
OCD	3.17 (±2.69)	0.89 (±1.47)	<0.001
GAD	6.43 (±2.69)	2.76 (±1.38)	<0.001

3.3.4 Social Vulnerability Questionnaire (SVQ)

Individuals with WS scored significantly higher on the SVQ compared to the TD group when examining total scores (see Table 3.3). They also scored significantly higher on all sub-scales of the SVQ, with higher scores being indicative of higher levels of vulnerability. This relatively new scale does not yet provide standardised scores or cut off values for the severity of vulnerability but the between group differences clearly show heightened variability in the WS, compared to the TD, group.

Table 3.3. SVQ scores for individuals with WS and typically developing individuals

	WS	TD	p
SVQ Total	76.35 (±9.3)	50.72 (±7.82)	<0.001
Emotional bullying	10.43 (±2.74)	7.72 (±2.7)	<0.001
Risk awareness	23.3 (±4.52)	16 (±2.82)	<0.001
Social protection	9.96 (±3.11)	6.52 (±2.13)	<0.001
Perceived vulnerability	11.83 (±2.37)	5 (±1.51)	<0.001
Parental independence	5.52 (±2.15)	9.03 (±2.63)	<0.001
Credulity	15.3 (±3.42)	6.45 (±1.9)	<0.001

3.3.5 Relationship between measures

To explore the relationship between these aspects of functioning for the first time, a number of exploratory approaches were applied to the data. The data for individuals with WS who scored in the upper quartile (M = 86, SD = 2.68, n = 6) and in the lower quartile (M = 63.67, SD = 6, n = 6) of the SVQ were extracted for further analysis (with extreme caution due to the reduced power and sample sizes). It was found that there was a significant difference in anxiety levels between the high social vulnerability group and the low social vulnerability group on the SCAS (M = 32.67, SD = 11.72 and M = 19, SD = 6.39 respectively, $t(10) = 2.5$, $p<0.05$); those who experienced high levels of vulnerability also displayed high levels of anxiety. Similarly, these two sub-groups showed a significant

difference in the atypicality of their social behaviour (high vulnerability: M = 84.67, SD = 8.19, low vulnerability: M = 64, SD = 15.47; $t(10) = 2.89$, $p<0.05$) on the SRS. Those in the high vulnerability group showed severe deficits in their social functioning, whereas individuals in the low vulnerability group showed moderate impairments. However, the two groups did not differ on their social communication, measured by the SCQ (high vulnerability: M = 16.5, SD = 6.16, low vulnerability: M = 13.83, SD = 4.49; $t(10) = 0.86$, $p = 0.41$). Despite the lack of significant difference between groups, the high vulnerability group met the cut off score on the SCQ (15 and above) which is indicative of severe atypicalities of social communication, whereas the low vulnerability group had a mean score below this marker.

Finally, looking at the full sample of individuals with WS, there were strong correlations between scores on the SRS, SVQ and the SCAS[2]. Figure 3.5 shows high correlations between the SRS and the SVQ, and also between the SRS and the SCAS-P. The SVQ and the SCAS-P were found to have a moderately strong correlation. Scores on the SCQ did not significantly correlate with any of the other measures.

[2] We note caution here due to the small sample size; however, it is relevant that a strong correlation exists even with a small sample size.

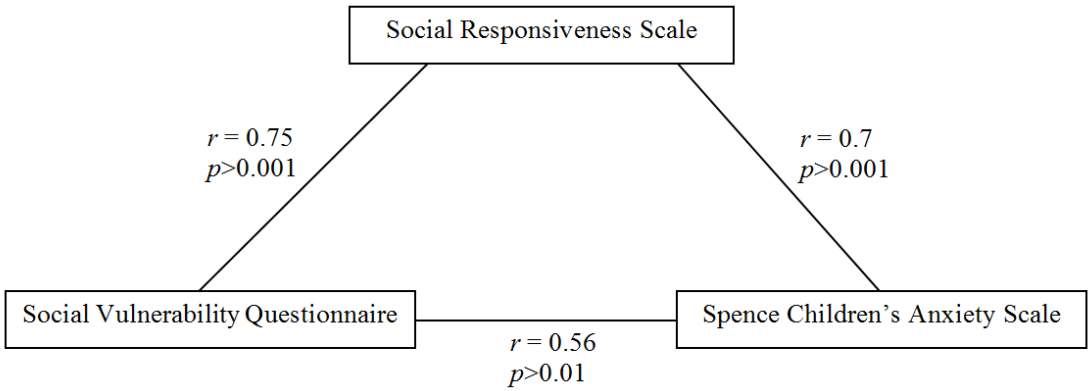


Figure 3.5. Correlations between scores on the SRS, SVQ and SCAS for individuals with WS.

As a pilot exploration for future follow up with a larger sample, a partial mediation analysis showed that social behaviour (SRS) was driving the relationship between scores on the SVQ and the SCAS. After partialling out anxiety, the correlation between social vulnerability and social behaviour remained significant (r = 0.6, p>0.01), however, when social behaviour was partialled out, the correlation between social vulnerability and anxiety was non-significant (r = 0.06, p = 0.79).

3.4 Discussion

Results from the current study showed that young people with WS experience a profile of elevated levels of anxiety and social vulnerability and displayed more atypical social behaviour and atypicalities of communication than their typically developing peers. Atypicalities of social behaviour are well documented as being present throughout the developmental trajectory (e.g. Riby et al., 2014), however, these findings reinforce that high levels of anxiety and social vulnerability are not

just relevant to adults with WS (Dykens, 2003; Fisher et al., 2012), but also adolescents too. When considering timings of potential interventions, these findings suggest that any interventions should begin pre-adolescence in order to better support young people with WS with these issues before they reach adulthood.

The first research question asked whether individuals with WS would score atypically on measures of anxiety, social behaviour, communication and vulnerability (compared to TD peers). This question focused on the 'average' for the WS group as later questions would explore individual differences in more detail. The findings of the current study show that individuals with WS did score atypically on these measures. Participants with WS scored significantly higher on the SCAS-P than their TD peers, suggesting that they experience much higher levels of anxiety (Porter, Dodd, & Cairns, 2009; Stinton, Elison & Howlin, 2010; Riby et al., 2014; Dodd & Porter, 2009; Dykens, 2003). This is in line with previous findings from Rodgers et al. (2012) who found individuals with WS (mean age = 9.4 years) experienced an elevated risk of anxiety compared to typically developing children. Interestingly, in the current findings, there was no difference between the WS group and the TD group on the social phobia subscale. Individuals with WS indeed displayed a non-significant lower mean score for this subscale than their TD peers. Dodd and Porter (2009) found that there was no difference between scores of social anxiety individuals with WS compared to TD individuals. They suggested that their gregarious, social behaviour marks underlying feelings of social anxiety. Rodgers et al. (2012) also proposed that the areas of social functioning in which individuals with WS show

relative proficiency, combined with their prosocial drive, could be protecting against the development of social anxiety.

Social behaviour was found to be significantly more impaired compared to typically developing controls, based on total SRS T scores. Indeed, the mean score for the WS group indicated that the group as a whole experienced severe deficits in social reprocracy. Interestingly, it was only on the social motivation subscale that more than 20% of participants showed 'normal' functioning. This is supported by findings from Klein-Tasman et al. (2011) who used parent and teacher report versions of the SRS to assess cross-situational consistency in social functioning in children with WS (mean age = 9.5 years). They found that parents rated their children as having significant impairments in reciprocal social interactions, with 39% of participants scoring within the severe range of deficits in everyday social functioning. There were good levels of correspondence (moderate significant correlations) between parent and teacher reports on the SRS, suggesting that atypical social behaviour in WS is evident in multiple contexts. Similar to the findings in the current study, Klein-Tasman and colleagues (2011) noted that difficulties with prosocial areas of social functioning (e.g. social motivation) were less common than social-cognitive difficulties in reciprocal social functioning (e.g. social communication, social cognition) based on parent and teacher reports. This suggests that despite their friendly and outgoing personalities, individuals with WS display poor social skills, which impact on the success of their everyday social interactions. Given the high levels of social isolation reported in WS (73%; Davies et al., 1998), it would seem that reciprocal social function is highly salient when considering the issue of social vulnerability.

Findings from the Social Communication Questionnaire (SCQ) showed that individuals with WS had a significantly higher mean total score, compared to the TD group, suggesting that they experience elevated levels of social communication impairments. Good social communication skills used to be considered as a relative strength in WS (e.g. Jones et al., 2000; Bellugi, Lichtenberger, Jones, Lai & St. George, 2000; Mervis, Klein-Tasman & Mastin, 2001). However, more recent work has suggested that individuals with WS are in fact less sensitive to the conversational input needed when interacting with others (Stojanovik, 2006), provide too little information to sustain meaningful conversation (Stojanovik, Perkins & Howard, 2001), and display pragmatic language impairments and social deficits (Laws & Bishop, 2004). The significantly higher levels of impairment found in the current study in the subdomains of reciprocal social interactions and communication suggest that, although individuals with WS display a strong interest in interacting with others, they experience significant difficulties in social communication which affect the success of their social interactions. The WS group also scored significantly higher on the restricted and repetitive behaviour subdomain of the SCQ. Restricted, repetitive patterns of behaviour are a core feature of ASD (The Diagnostic and Statistical Manual of Mental Disorders, 5th ed.; *DSM–5*; American Psychiatric Association, 2013). As explored in Chapter 2, there exists overlap between the profiles associated with WS and ASD, despite their seemingly opposing social behaviours. For example, Philofsky, Filder and Hepburn (2007) found that children with WS (mean age = 9.1 years) and children with ASD (mean age = 9.6 years) both displayed communicative and pragmatic language impairments. The

findings from the current study therefore question the extent to which the reported deficits in social communication are syndrome specific.

The current findings also highlighted the high rates of social vulnerability experienced by individuals with WS, compared to those who are TD, which is in line with a recent review on the high levels of social vulnerability in WS (Jawaid et al., 2014). Fisher et al. (2013) found that whilst many individuals with intellectual and developmental disabilities (IDD) experience high levels of vulnerability, the nature of this vulnerability differs depending on the type of IDD. For example, they found that the high levels of vulnerability experienced by individuals with WS (mean age = 25.4 years) was linked to their high levels of parental independence and perceived vulnerability, i.e. other people perceive these individuals to be vulnerable. The current findings show significant differences between the WS group and the TD group on all sub-domains of the SVQ, suggesting that during adolescence, there are several different facets contributing to the social vulnerability seen in WS, emphasising the need to provide support prior to adulthood. Preliminary work by Fisher (2014) showed that stranger danger awareness in adults, which is linked to vulnerability levels, could be improved by a stranger safety training programme. She found that, prior to the intervention, only 14% of adults with WS walked away when approached by a stranger. After a three day behavioural skills training programme focused on how to respond to lures from strangers, 62% of participants walked away. Whilst this showed vast improvement, 14% of participants still agreed to leave with a stranger post-intervention, and the longitudinal effects of an intervention of this sort remain unclear. The findings in the current study suggest that there is reason

to trial interventions with a younger age group, and track progress longitudinally to improve intervention success.

The second research question asked whether individuals with WS identified as being especially vulnerable experience significantly higher levels of anxiety, as well as more atypical social behavior and communication compared to those with relatively low levels of vulnerability. Indeed, we found a significant difference in anxiety levels and of social behavior between a sub-group of individuals with WS deemed to experience high levels of social vulnerability, compared to a low-vulnerability sub-group (using an upper and lower quartile approach as the SVQ does not provide cut off values) therefore supporting the proposed hypothesis.

The second research question also allowed us to begin to think about the variability seen within WS – for example, the fact that we could split the group on the basis of vulnerability scores and also that there was vast heterogeneity across all measures in both the WS and TD groups. Whilst the social behaviour displayed by individuals with WS has traditionally been thought of as a homogenous concept, recent work has begun to acknowledge the variability that exists within the syndrome at both the behavioural and cognitive levels (e.g. Porter & Coltheart, 2005; Jarvinen-Pasley et al., 2010). Little et al. (2013) found evidence supporting the notion of WS sub-groups of social approach behaviour which could be identified based on inhibition ability. They emphasised the need to look at approach behaviour at an individual level in order to understand the variability within the WS social profile. The variability within the WS profile is evident in the current study from the scatterplots presented of the SCQ and

SCAS-P total scores. They outline the range and spread of scores for both the WS and TD groups. According to Porter et al. (2007), cognitive heterogeneity in WS reflects differing patterns of strengths and weaknesses, rather than degree of impairment. In the current study, individuals with high versus low vulnerability levels displayed differing behavioural profiles, with higher levels of anxiety and a more atypical social profile being found in those with high vulnerability levels. A clearer understanding of the within-syndrome heterogeneity in WS therefore seems important in furthering our understanding about the social vulnerability profile associated with this syndrome.

The third research question addressed whether abilities on the four constructs correlated with one and other for individuals with WS. Findings from the current study showed significant positive correlations between scores on the SRS, SVQ and the SCAS. As the same parent completed all of the measures, it is relevant to consider how the parents see the overlap between these issues. Previous work by Riby et al. (2014) looked at the interplay between anxiety and social functioning. They found the severity of anxiety, as measured by the SCAS, was positively correlated with scores on the SRS; individuals with higher levels of anxiety showed more atypical social behaviour. Indeed, when the sample was split according to their anxiety scores (high versus low), there was a significant difference between the social skills profile of these two groups. The authors emphasised the importance of taking measures of anxiety when investigating social behaviour. Anxiety has also been linked to the way social information is attended to / perceived (Kirk et al., 2013; Freeth, Bullock & Milne, 2013).

However, Ng et al. (2014) found no significant relationship between social-emotional functioning (using the Salk Institute Sociability Questionnaire; Jones et al., 2000) and anxiety characteristics (measured by the Beck Anxiety Inventory; Beck, Epstein, Brown, & Steer, 1988; Beck & Steer, 1993), but found a positive association between intellectual abilities and anxiety. The mean age of the WS sample in this study was 32 years, compared to a mean age of 17 years in the work by Riby et al. (2014) which is likely to contribute to the differences observed, however, future research should clarify the link between social behaviour, anxiety and vulnerability.

The pilot exploration of the relationship between social behaviour, anxiety and social vulnerability offers some interesting, but tentative, theoretical considerations. Partial mediation analysis revealed that social behaviour was driving the relationship between anxiety and social vulnerability. This suggests that high anxiety alone is unlikely to result in increased social vulnerability. Rather, high anxiety which occurs alongside severely impaired social functioning is a prelude to high levels of vulnerability. The strong correlations between the SRS, SCAS-P and the SVQ suggest that social behaviour and anxiety are very much linked to the construct of social vulnerability, however, preliminary suggestions from the current data would suggest that interventions designed to improve social skills are more likely to reduce levels of vulnerability, compared to interventions designed to reduce anxiety. Future work with a larger sample size is needed to improve our understanding of the relationship between these issues, and in turn, our theoretical understanding of social vulnerability.

The patterns of behaviour noted in this chapter have important applied implications for the daily lives of individuals with WS. However, there are limitations which should be addressed. First, although the findings have indicated some within-syndrome variability in the WS phenotype, the small sample size makes it hard to capture the true extent of this variability. The sample size also did not allow for a full mediation analysis (Fritz & MacKinnon, 2007). Further large-scale research is needed into the link between anxiety, social behaviour and social vulnerability, which will enable us to develop a more holistic view of the social profile associated with WS to aid intervention and theory.

Secondly, we did not find a significant correlation between the SCQ and the other measures. As the SCQ was designed as a screening tool for autism and is widely used in the autism field (e.g. Chandler et al., 2007; Allen, Silove, Williams & Hutchins, 2007; Eaves, Wingert, Ho & Mickelson, 2006; Wiggins, Bakeman, Adamson & Robins, 2007), it may be that this measure is not sensitive enough to specific characteristics of the WS profile. Indeed this links to a wider issue of the relevance of the measures and the groups that they have been designed to be used with (see Chapter 2 for further discussion). The measures used have not been specifically designed to capture typicality / atypicality in WS. For example, although the SRS has been used repeatedly in WS populations (e.g. Klein-Tasman, Li-Barber & Magargee, 2011; Riby et al., 2014; Karmiloff-Smith et al., 2012), it was not designed to capture the nuances of the disorder. The applicability of measures designed for use in TD populations, or indeed for other developmental disorders requires careful consideration.

Importantly, the characteristics of the WS social phenotype captured by the current exploratory study will inform the subsequent chapters of this thesis where some of these issues are probed further.

Chapter Four: Parent insights into atypicalities of social approach behaviour in Williams syndrome

The findings from Chapter 3 reinforced the notion that individuals with WS experience high levels of vulnerability, albeit with likely within-syndrome variability. Given the impact that this vulnerability has on the daily lives of individuals and their parents/carers, closer examination of the behaviours which may contribute to this vulnerability is needed. Anecdotal reports from parents have highlighted concerns about social interactions with strangers. An important precursor to this is the social approach behaviour which allows for these interactions. The published literature features a number of studies of social approach utilizing rating scale methodologies (e.g. Jones et al., 2000) but rich, qualitative data on the nature of the social approach behaviour seen in WS is currently lacking and would be a particularly useful next step in understanding more about social vulnerability in WS.

This paper has appeared as:

Lough, E., Rodgers J, Janes E, Little K, Riby D. M. (2016). Parent insights into atypicalities of social approach behaviour in Williams syndrome. *Journal of Intellectual Disability Research*. doi: 10.1111/jir.12279

4.1 Introduction

Williams syndrome (WS) is a genetic neurodevelopmental disorder with a prevalence of approximately 1:20,000 and is caused by the micro-deletion of 25-28 genes on chromosome 7 (7q11.23; Hiller et al., 2003). Alongside mild-moderate levels of intellectual impairment (Searcy et al., 2004), individuals with WS have been reported to display a paradoxical cognitive profile of relative strengths in verbal processing and relative weaknesses of spatial ability (Mervis et al., 2000). Although, it is acknowledged that even the relative strengths in verbal processing are select (Paterson et al., 1999; Laing et al., 2002; Mervis et al., 1999), as language acquisition often follows an atypical developmental pathway, showing deficits in areas such as past tense formation and atypical phonological representations (Thomas et al., 2001). Most relevant to the current study, the social phenotype of WS has attracted significant research attention, largely due to claims of hyper-sociability (Jarvinen et al., 2013). This translates as an extreme prosocial drive to approach and interact with other people, irrespective of whether the person is known to them or not (Jones et al., 2000).

Several studies have explored and characterised social approach behaviours in WS by asking individuals to rate faces for approachability using a Likert scale, when given a hypothetical situation of whether they would like to talk to the presented face or not. However, such studies have produced conflicting findings, which can typically be accounted for by the type of task used and the emotional expression of the faces that have been presented (Porter et al., 2007). For example, in some studies individuals with WS report higher approachability ratings for trustworthy and untrustworthy faces compared to their typically

developing (TD) peers (Jones et al., 2000; Martens et al., 2009). Yet, other studies have shown that individuals with WS only give high approachability ratings to happy faces, rather than those expressing negative emotions such as anger or fear (Frigerio et al., 2006). The role of emotion in social approach decisions is thought to be further complicated by impairments in emotion recognition (Porter et al., 2007). Thus the exact nature of social approach behaviour in WS remains unclear, but the issue remains of great importance because of the social vulnerability status associated with increased approach to unfamiliar people (for a discussion of vulnerability issues see both Jawaid et al., 2012 and Lough et al., 2015a). This vulnerability is heightened by considering the increased social approach in addition to the previously mentioned intellectual impairments (Searcy et al., 2004) and an abundance of social functioning atypicalities such as staring at faces (e.g. Riby & Hancock, 2008) and an inability to make accurate socio-cognitive judgements (Tager-Flusberg & Sullivan, 2000). Social vulnerability warrants further exploration using multiple methods to gain complementary insights into the social approach behaviours and underlying issues that could be tackled as part of a social skills training programme

Riby and colleagues (2014) approached the issue of stranger danger awareness in WS in a novel way by conducting a qualitative analysis of discussions with young people with WS that stemmed from stranger danger video vignettes. Based on the qualitative data produced, it was clear that young people with WS showed heightened vulnerability compared to typically developing individuals. Crucially, 73 per cent of the answers given by the young people with WS (mean 12 years) failed to show an appropriate knowledge or awareness of any risks of interacting

with unfamiliar adults. This compared to an average 40 per cent of the responses given by a younger group of typically developing children (mean age 7 years). Riby and colleagues recommended that further qualitative data were needed from a variety of sources on the issue of social approach behaviour and stranger danger awareness, especially based on the within-syndrome variability observed in the responses for the WS group (also captured by Little et al., 2013). This work would allow us to tailor training programmes to complement individual differences of social approach and stranger danger awareness in WS.

The importance of individual differences was again highlighted in recent work by Ng, Jarvinen and Bellugi (2014). They emphasized the impact that the WS personality profile could have in explaining maladaptive social behaviours. They outlined the case of atypical social motivation in WS. Individuals with WS (both children and adults) were driven by a desire for social closeness in their social interactions, which was underpinned by their "gregarious, people-orientated and affectionate personality features" (p1844) whereas their typically developing peers sought social power driven by "persuasive, dominant and visible personality attributes" (p1844). They argued that identifying the role that personality traits play in the elevated levels of social drive seen in WS could allow us to target interventions towards these areas. Further research on individual social motivation and the underlying mechanisms of social motivation in WS is clearly warranted.

So why do individuals with WS struggle to make appropriate social judgements? There have been several theories proposed to explain the WS social phenotype.

Specifically, the neural systems underpinning this social behaviour have attracted significant interest. The amygdala hypothesis proposes atypicalities in amygdala structure and function of individuals with WS. It suggests that those with WS have an atypically enlarged amygdala volume which is linked to the atypical social approach behaviours (Martens, 2009). According to Haas et al. (2009), individuals with WS show decreased amygdala activation in response to threatening faces, which the authors suggest could explain the disinhibited approach behaviour. Therefore both structure and function of the amygdala appear critical. An alternative has been proposed by the frontal lobe hypothesis (e.g. Porter et al. 2007). According to this theory, individuals with WS show similarities of social approach behaviour to individuals who have experienced frontal lobe damage. Both groups share deficits in response inhibition, which leads to atypical approaches, such as approaching strangers. This occurs in spite of 'knowing' that this type of approach behaviour is not appropriate. It could therefore be that inhibitory control is key (Little et al., 2013). However, the proposed theories are far from mutually exclusive. In reality, most researchers acknowledge that these theories are unlikely to be absolute, and rather each makes a partial contribution to our understanding of social approach behaviours in WS (Gaser et al., 2006; Meyer-Lindenberg et al., 2005).

Indeed, recent work has noted considerable variability in areas such as frontal lobe functioning, social functioning, anxiety and social approach behaviours in WS (e.g. Porter et al., 2007; Little et al., 2013; Jarvinen-Pasley et al., 2010; Riby et al., 2014). Little et al. (2013) proposed the notion of sub-groups within WS based on social approach. Through cluster analysis of children's responses on

Adolphs Approachability Task (Adolphs et al., 1998), an emotion recognition task and a response inhibition task (the Sun-Moon Stroop Task; Archibald & Kerns, 1999), they noted substantial variability of approach desires. They argued that WS subgroups could be identified based on the social approach profile of an individual, with inhibition being the strongest indicator of subgroup membership. This highlights the need to look at social approach behaviour in a manner that captures individual differences and without reliance on group 'means'. This is especially important for accurately evaluating intervention needs.

The methods employed to investigate social approach behaviour have been discussed. Recent work by Fisher, Mello and Dykens (2014) highlighted a discrepancy between self-report and parental reports of social approach behaviour in adults with WS. They found that the responses given by individuals with WS in a number of different tasks (e.g. self-report approachability scale, self-report faces task) suggested that they displayed much lower levels of abnormal social approach behaviour compared to the levels reported by their parents. Indeed, behavioural observation in a community setting showed it was parent report responses which were more consistent with observations of social behaviour in a natural setting, suggesting that parents could more accurately report their child's social approach behaviour towards strangers. This may be something that individuals with WS find very hard to reflect upon, especially during childhood.

Parent report has been used in the existing literature on social approach behaviour, however, it has predominantly been in the form of questionnaire responses (e.g. Doyle et al., 2004). Considering the value attached to parental

reports, and the current discrepancy of findings in WS, the current study aims to extract more in-depth, rich, qualitative data through semi-structured parent interviews. Using the Social Responsiveness Scale (SRS; Constantino & Gruber, 2005) and the Spence Children's Anxiety Questionnaire – Parent Version (SCAS-P; Spence, 1998) we will gain insight into the general social and anxiety profile of this group (previous research has shown anxiety levels to be high in WS; Riby et al., 2014; Rodgers et al., 2012) as well as establishing whether there is heterogeneity and thus within-syndrome variability in the parental accounts given. This will provide a novel and valuable insight into the social competence of the group, their patterns of social approach behaviour and within-syndrome variability.

4.2. Method

4.2.1 Participants

The parents of twenty-one children with WS (range 6 – 15 years; mean age 9.8 years; SD3.2; 10 males, 11 females) were recruited through the Williams Syndrome Foundation. Their child must have had a formal WS diagnosis which had been confirmed through positive genetic florescent *in situ* hybridisation testing. We used the Wechsler Intelligence Scale for Children (WISC-IV; Wechsler, 2003) which generated an overall intellectual ability mean of 54.14 (SD 7.57; Full Scale Intelligence Quotient; FISQ). The sample had a mean verbal IQ (VIQ) score of 63.62 (SD 9.93) and a performance IQ (PIQ) of 51.29 (SD 6.86). For two families, both parents took part in the interview, and for the remaining 19 families, the mother was interviewed. The ethnicity of the cohort

was entirely white British. Participants who had a co-morbid diagnosis of an Autism Spectrum Disorder were excluded from the study. The study received favourable ethical approval from the local ethics committee. Informed consent was obtained from parents who took part in the interview.

4.2.2 Materials and Procedure

Social Responsiveness Scale

The Social Responsiveness Scale (SRS; Constantino & Gruber, 2005) is a 65-item parent report questionnaire that measures the normality / abnormality of a child's social functioning. It was originally designed as a screener for Autism Spectrum Disorders, and has since been used to detail the social profile of a variety of typical and atypical populations including with individuals who have Williams syndrome (see Lough et al., 2015b; Riby et al., 2014; Klein-Tasman et al., 2011; Van der Fluit et al., 2012; Channell et al., 2015). Each item on the SRS is coded on a scale of 0 – 3, which generates scores across five sub-domains - social awareness, social cognition, social communication, social motivation and autistic mannerisms, as well as an overall T score as a degree of severity of social abnormality. Higher scores represent greater deficits of everyday social functioning. Previous research using the SRS has suggested that only a small percentage of individuals with WS are likely to be classified as showing 'normal' social behaviours; far more are likely to show either mild-moderate or severe impairments that impact on daily functioning. For example, van der Fluit, Gaffrey, and Klein-Tasman (2012) reported only 17% of individuals with WS (total n= 24) were classified within the 'normal' range and this was corroborated by Riby et al. (2013) who also reported 17% of their sample to fall within this

range (sample size n=59). In that same study, 58 % were classified by parents as showing severe deficits of reciprocal social interaction behaviour that would significantly impair everyday social functioning and 25% showed mild deficits of social behaviour (Riby et al., 2014).

Spence Children's Anxiety Questionnaire – Parent Version

The Spence Children's Anxiety Questionnaire – Parent Version (SCAS-P; Spence, 1998) was completed by 18 of the parents (86%) in the sample. The SCAS-P has previously been used in the literature to measure anxiety in children with WS (e.g. Rodgers et al., Riby et al., 2014) and in relation to the link between social behaviour and anxiety in this population. It is a 38-item measure, on which parents must rate statements on a four point Likert scale, which correspond to the options *never*, *sometimes*, *often* and *always*. This measure provides an overall indication of anxiety levels, as well as scores in six subdomains: separation anxiety, physical injury fears, social phobia, obsessive compulsive disorder, and generalised anxiety disorder.

Social Approach Behaviour Interview

A bespoke semi-structured interview was developed by the authors and completed with parents of children with WS. The interview had four modules; auditory sensitivity, social approach behaviour, understanding of emotion and anxiety; of which the social approach behaviour module is explored here (see Appendix A for interview schedule). Relating to the child's social behaviour, the questions covered themes such as interest in social situations, confidence around strangers, and knowledge not to approach strangers.

The researchers met with the parent individually to complete the SRS, the SCAS-P and the semi-structured interview. The interviews were conducted in the homes of families, and the whole interview (including the social approach / social behaviour module) took approximately 60 minutes.

4.2.3 Data analysis strategy

Thematic analysis was used to systematically analyse the data in line with the suggestions of Braun and Clarke (2006). The interviews were transcribed and initial codes and conceptualisations were generated from line-by-line coding of the accounts given by parents. These codes were analysed and developed into themes which were deemed to fit the data as closely as possible. These themes were processed and reprocessed until final themes were generated and could be reviewed.

4.3 Results

4.3.1 Social Responsiveness Scale

The mean SRS T score for the sample showed that the group as a whole experienced severe levels of impairment in their social functioning (mean T score = 79.6), although there were high levels of variability within the sample (SD = 13.5). There was no significant difference in the overall T scores of males (M = 77.9, SD = 15.02) versus females (M = 81.18, SD = 12.45; t(19) = 1.77, p = 0.59), and there was no significant correlation between total SRS T score and FSIQ (r = 0.19, p = 0.41), VIQ (r = 0.04, p = 0.88) or PIQ (r = 0.27, p = 0.24). Figure 4.1 shows that 72% of the group had overall T scores in the severe range,

whilst 14% scored within the mild-moderately impaired range, and 14% within the normal range of social functioning. It is worth noting that the proportion of WS participants being classified as having mild-moderate and severe social deficits is similar to previous reports with larger WS samples using the SRS (van der Fluit, et al., 2012; Riby et al., 2014).

There was a significant correlation between the age of the participants and their total T score ($r = -0.56$, $p<0.01$). There was also a significant correlation between age and scores on 4 out of the 5 sub-domains of the SRS (awareness: $r = -0.52$, $p<0.05$; cognition: $r = -0.63$, $p<0.01$; communication: $r = 0.64$, $p<0.01$; mannerisms: $r = -0.58$, $p<0.01$), suggesting that the most socially impaired were, on average, younger. However, this was not the case for the sub-domain of social motivation ($r = 0.21$, $p = 0.37$), indicating that atypicalities in social motivation do not differ with age. Of further interest, all of the sub-domains of the SRS were significantly correlated with each other (all at $p<0.05$), with the exception of social motivation and awareness ($r = 0.3$, $p = 0.19$), and social motivation and cognition ($r = 0.23$, $p = 0.31$). Social motivation is something that is clearly atypical in WS across ages and that has been captured in the WS literature to date as an identifying aspect of the WS social phenotype (e.g. Doyle et al., 2004; Frigerio et al., 2006; Jawaid et al., 2012).

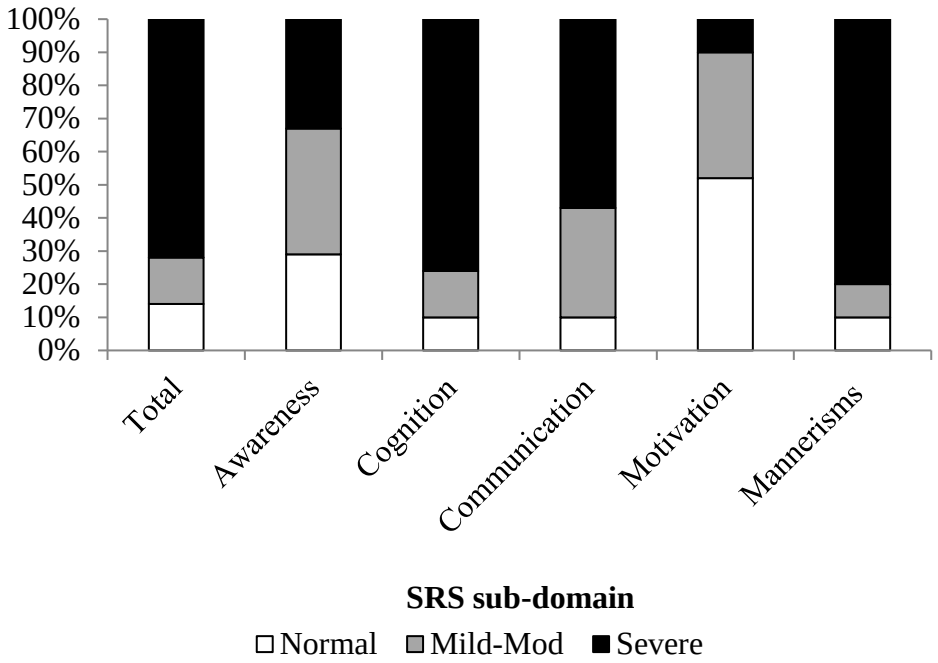


Figure 4.1: Levels of impairment shown for total SRS scores and scores on the five sub-domains

4.3.2 Spence Children's Anxiety Scale – Parent Version

The mean raw score for overall anxiety was 20.23 (SD 12.18), suggesting the sample experience low levels of anxiety (Rodgers et al., 2012). From this it can be proposed that this will have limited influence on social approach behaviours (Riby et al., 2014). The mean sub-scale scores are shown in Table 4.1. Interestingly, participants scored highest on the GAD subscale of anxiety, with low scores on the OCD, social phobia and panic subscales. There was no significant correlation between total SCAS scores and total SRS T scores ($r = 0.38$, $p = 0.12$), age ($r = 0.19$, $p = 0.45$) or IQ (FISQ: $r = 0.05$, $p = 0.84$; VIQ: $r = 0.05$, $p = 0.83$; PIQ: $r = 0.2$, $p = 0.42$).

Table 4.1: Mean SCAS-P total score and sub-scale scores

SCAS-P	T scores
Total score	20.23 (12.18)
Panic/Agoraphobia	2.22 (2.51)
Separation anxiety	4.83 (4.08)
Physical injury fears	4.17 (2.79)
Social phobia	2.11 (2.14)
OCD	1.89 (1.99)
GAD	5.06 (3.19)

4.3.3 Social approach behaviour interview

The thematic map shown in Figure 4.2 depicts the themes that arose from the semi-structured interviews with parents of children with WS.

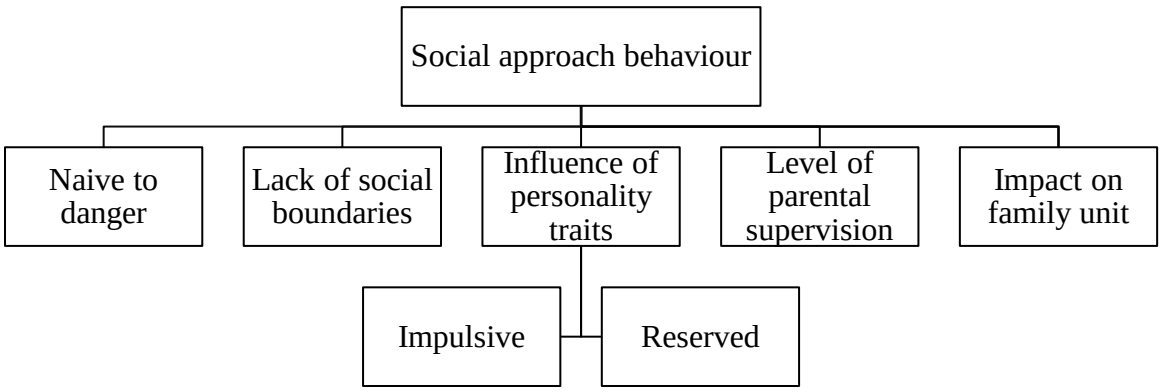


Figure 4.2: Thematic map for parent interviews on social approach behaviours

Naivety to danger and a lack of social boundaries were prominent themes in the accounts of parents with children with WS. However, there were qualitative differences in the nature of their social behaviour, personality traits and the level of parental supervision employed, reinforcing the heterogeneous nature of social approach behaviour in WS.

The parents talked about the naivety of their children, in particular to dangerous or potentially risky situations and as seen by the ages of the illustrations below, this was an issue across ages:

> "She can't understand why she can't talk to people she doesn't know she will say they're nice and she liked them so she doesn't understand why that's bad" (female, 8 years)
>
> "I just know for a fact anyone could come up in a car and say come on Natalie and she would climb in and go with them" (female, 6 years)
>
> "I think he's too trusting particularly of adults … he would be very easily led" (male, 15 years)
>
> "I picked her up because she was sick and we crossed the road and a man walked past and she just starts waving and says hello as he got closer, asking him what his name was" (female, 6 years)

They also frequently highlighted the difficulties experienced by their children with regards to understanding and respecting social boundaries:

> "She will ask private questions she will tell things about herself which are just not appropriate" (female, 9 years)

> "She's not got boundaries … if she was going to talk to someone she would put her hand on their knee or arm she would break that personal space and not understand that it wasn't right" (female, 8 years)
>
> "… he will hold hands and try and hug people whether he knows someone or not is irrelevant" (male, 9 years)
>
> "She has no concept of personal space … if someone has a nice necklace she will touch it and tell them she likes it, she can get that close to them" (female, 6 years)

These observations seemed to be tied in with the extreme outgoing and gregarious behaviour reportedly displayed by some of the children. Several parents described their children as highly impulsive in their social approach behaviour:

> "I don't know, you can tell her until you're blue in the face but it's like it is inbuilt it's something that she can't stop" (female, 12 years)
>
> "It's just an instinct for her it's part of her genetic make-up its spontaneous it's not something she thinks" (female, 9 years)
>
> "She doesn't ever really think about what she's doing" (female, 8 years)

Parents were also concerned about the longevity of this behaviour, and many shared their concerns for the future:

> "I don't know that she will ever be aware that you don't approach strangers" (female, 14 years)
>
> "I keep saying she will never be in a situation on her own, but she's going to get older and you don't know what's going to happen" (female, 14 years)

However, there was a notable amount of variability in the accounts, as not all parents reported these impulsive behaviours. Some parents discussed the reserved personality of their child, which they saw as serving to minimise inappropriate social approach behaviour:

> "He wouldn't like to be the centre of attention or to stand up and talk in front of lots of people so I think he would be more comfortable in familiar surroundings with people he knows" (male, 15 years)
>
> "I see him hold back sometimes if he doesn't like someone" (male, 13 years)
>
> "She doesn't actively seek others out, she's quite quiet" (female, 15 years)

The above quotes begin to illustrate the heterogeneity in the accounts given. With differing degrees of social approach behaviour, as well as distinctly different personality traits, it is perhaps unsurprising that the level of parental supervision employed was also varied. Some of the parents referenced the high level of parental supervision they employed to ensure that their children were safe around strangers.

> "I think because he doesn't go out by himself I don't really worry about strangers" (male, 15 years)
>
> "I think what holds him back most of all is … he's very restricted by having to need us to be there or take him somewhere so I think that has stunted his social life"(male, 9 years)

The first of the above quotes is interesting given the age of the individual with WS and the likelihood that if they were typically developing this is an age (15

years) when we would expect social independence to be evident. It seems that such a high level of parental supervision has curtailed opportunities for social approach, but at a cost to their level of independence. The primary driving force when parents are considering this equation was their need to protect their child.

> "If we weren't there, she would be easy picking" (*female, 6 years*)

For other parents, they have been able to build up confidence and trust in their child, allowing them less parental supervision, and greater autonomy:

> "At first I was worried because I'm a mum and he was going up talking to people he doesn't know, but now I've got confidence in him and knowing his own mind." (male, 13 years)

Perhaps unsurprisingly, many families spoke about the impact that their child's social approach behaviour had on the family unit and daily living:

> "His sister gets embarrassed. She's younger … and she will start to talk to people as well because she sees him doing it" (male, 10 years)
>
> "We have to do holidays different my husband would love to do an all-inclusive somewhere but I can't possibly go somewhere where she can pester the same people through breakfast at the pool through the afternoon and at dinner as well so for holidays we always go self-catering and we always go to the same places so we know our containment areas." (female, 6 years)

4.4 Discussion

By analysing interviews conducted with parents of children with WS, the current study identified impaired social competence and high levels of social approach

behaviour across the sample. We also noted considerable heterogeneity of social approach behaviours in this clinical group, consistent with previous research (e.g. Little et al., 2013; Porter et al., 2007). Based on our findings, it would seem that this variability cannot be predicted solely by age or IQ. Indeed there were some themes in the parent interviews that were evident for all parents irrespective of the age of their child. As expected, all of the children were reported to display inappropriate social behaviour, and to be naïve to danger, but crucially their personality traits (e.g. their level of impulsiveness) as well as familial factors (e.g. level of parental supervision) influenced the nature of this behaviour. This is in line with previous research (e.g. Porter et al., 2007; Ng et al., 2014) and complements findings by Porter and Colheart (2005) on the heterogeneity of cognitive strengths and weaknesses in WS. The differences found in the qualitative interview data are likely to help shape the individual atypical social profiles of these children, and impact upon the effectiveness of interventions which assume a homogenous WS social behaviour profile. Furthermore the individual nature of the social approach profiles in these children will impact upon the way that such behaviours influence family life in each of these family units.

Based on the interviews, and the data obtained from the questionnaire rating items, it is clear that the children with WS in this study showed an interest in social situations and as evident across both the SRS and the interview data, were strongly socially motivated (in line with Frigerio et al., 2006); however only some were reported to be especially confident and disinhibited around strangers. When considering the theoretical explanations offered by the amygdala hypothesis and

the frontal lobe hypothesis, this heterogeneity proves problematic. The frontal lobe hypothesis centres on difficulties with response inhibition, yet not all of the participants were reported to experience this, or indeed not to the same extent. Unfortunately we do not have cognitive or behavioural inhibition data for this sample of children but it would be interesting to explore the role of the cognitive heterogeneity in WS with the social heterogeneity reported here. Furthermore, it may be that individual differences in personality factors could play an important mediating role in prosocial WS drive as recently suggested (Ng et al., 2014). Certainly some parents suggested that their children were outgoing and extraverted, whereas others emphasised the reserved nature of their child. This issue suggests an interesting area for further exploration. Finding an appropriate theoretical framework for social approach behaviours in WS is dependent on acknowledgement of the heterogeneity and subgroups that exist within the disorder and the role of both cognitive and social profiles. Therefore taking an in-depth individual / holistic approach to understanding such issues is crucial for both theory and practice.

These findings offer a novel insight into the vulnerability status of some individuals with WS. Given that individuals with WS struggle to form and maintain peer relationships (Davies et al., 1998), experience high levels of anxiety (Riby et al., 2014; Rodgers et al., 2012) and lack stranger danger awareness (Riby et al., 2014), the increased social approaches of some individuals with WS is of particular concern. These individuals may be targeted for intervention. Indeed the qualitative data provided by parents in this study allows us to delve deeper into the social approach profile of individuals with the disorder

than face rating tasks used previously (e.g. Jones et al., 2000). The work can have a significant impact by highlighting the heterogeneity of social approach in WS, but also by emphasising the impact of the atypicalities of social behaviour and social approach on the wider family unit. Parents noted this in their responses as highlighted in a number of quotes in the Results section. Therefore supporting these family needs is important.

The limitations of the current study merit consideration. The qualitative interview data have provided us with a rich insight into how parents view their child's social approach behaviour. However, these data do not allow for analysis of the link between SRS scores, SCAS scores and social approach behaviour. Therefore, whilst these measures are useful in outlining the profile of the sample, the relationship between social functioning, anxiety and social approach remains unclear. Furthermore, although we have outlined the impact that age and IQ has on social functioning and anxiety in our sample, it is not clear how these factors relate to the social approach behaviour described in the interviews. It seems likely that age will have an effect on social approach, although it is worth noting that quotes about abnormal social approach behaviour were provided by parents of children of varying ages, implying that it could transcend age boundaries. Finally, as parental report offers an indirect measure of social approach, it is important that it is considered alongside other methodologies, in order to adopt a multi-informant approach to understanding social approach behaviour.

The findings from this study open up numerous avenues for future research. First, the developmental trajectory of social approach behaviours in WS remains

unclear, and in particular whether the heterogeneity reported here persists into adulthood. Furthering our knowledge on this area is particularly important when considering the increased levels of independence associated with adulthood, and the potential impact of social approach on social vulnerability (e.g. Lough et al., 2014). Secondly, as the literature base on heterogeneity in WS begins to build, future research should look to bridge the gap between the reported heterogeneous social profile, and the heterogeneous cognitive profile, in order to generate more comprehensive ideas on how to define these subgroups. This could be invaluable in helping to tailor support and avoid a one size fits all approach to intervention. Finally, the current study emphasises the importance of considering social approach behaviours and subsequent issues of vulnerability at the individual level, moving away from reliance on group means in order to formulate effective interventions.

Chapter Five: Personal space regulation in Williams syndrome

Chapter 4 provided a rich, qualitative insight into the social approach behaviour of young people with WS, as reported by parents. Although there was considerable within-syndrome variability, many parents indicated that their son/daughter would approach a stranger, and would have little awareness of personal space boundaries. Chapter 5 will extend these parental reports, and offer the first insight into personal space regulation in WS, taking a multi-methods approach (see Chapter 2) by using a combination of parent report questionnaires, and experimental paradigms. Considering the lack of stranger danger awareness reported in Chapter 4, personal space regulation when interacting with unfamiliar people is of particular interest in Chapter 5.

This chapter includes two published papers that have appeared as:

Lough, E., Hanley, M., Rodgers, J., South, M., Kirk, H., Kennedy, D. & Riby, D. M. (2015). Violations of Personal Space in Young People with Autism Spectrum Disorders and Williams Syndrome: Insights from the Social Responsiveness Scale. *Journal of Autism and Developmental Disorders. doi:* 10.1007/s10803-015-2536-0

Lough, E., Flynn, E. G. & Riby, D. M. (2016). Personal space regulation in Williams syndrome: The effect of familiarity. *Journal of Autism and Developmental Disorders. .doi:* 10.1007/s10803-016-2864-8

Violations of personal space in young people with Autism Spectrum Disorders and Williams syndrome: Insights from the Social Responsiveness Scale

5.1 Introduction

Personal space refers to the distance that individuals strive to maintain between themselves and other people (Hall, 1966). Intrusion of another person's personal space can have significant implications on social interactions, prompting feelings of discomfort and anxiety (Perry et al., 2013) or transferring fallacious social intentions (Kaitz et al., 2004). In order to proactively avoid such intrusions, we automatically regulate the boundaries for our personal space, and these boundaries are continuously re-assessed dependent on social dynamics and context (Lloyd, 2009). For example, the physical distance maintained between two people, i.e. interpersonal distance, can vary as a function of many factors, including familiarity, age and gender (Horne, 2006; Beaulieu, 2004). As such, successful interpretation of these social cues and subsequent appropriate decisions on context-dependent personal space regulation, play a vital role in positive social interactions (Gessaroli et al., 2013).

Despite the regulation of personal space being an automatic and adaptive process in typically developing individuals, several studies have shown that patterns of personal space regulation are altered in individuals who follow an atypical developmental trajectory. For instance individuals with developmental disorders such as Autism Spectrum Disorder (ASD), which is characterised by notable difficulties in social interactions (APA, 2013), often find interpreting and responding to social situations challenging (Smith et al., 2010). A recent study by

Gessaroli and colleagues (2013) used an experimental stop-distance paradigm (Hayduk, 1978) to examine the issue of interpersonal distance in children with ASD. For that task, the participants were asked to approach the experimenter and stop at the distance that felt most comfortable to them, which was in turn measured using a digital laser measurer. Gessaroli et al. (2013) found that children with ASD maintained a greater interpersonal distance than their typically developing (TD) peers, and this was not modulated in response to social cues such as familiarity, as it was in TD children. Therefore, they suggested that individuals with ASD not only maintained a greater personal space distance, but their personal space boundaries were also more rigid and less socially responsive than other children of the same age who did not have ASD. In contrast, Kennedy and Adolphs (2014) employed parent rated behavioural measures to examine interpersonal distance in ASD using the Social Responsiveness Scale (SRS) questionnaire (Constantino & Gruber, 2005). Despite the greater self-boundaries identified by Gessaroli et al. (2013), Kennedy and Adolphs found that 79% of parents report that their children with ASD were more likely to violate the personal space of others (e.g. have smaller self-boundaries) when compared to their TD siblings. Although both studies therefore suggest a difficulty of regulating personal space, these two previous studies provide equivocal findings, which could be in part attributed to the different forms of assessment.

A theoretically important developmental disorder, which features atypical social interactions and a lack of appropriate responsiveness to complex social stimuli, is Williams syndrome (WS; Pinheiro et al., 2011). WS is a rare genetic neuro-developmental disorder, affecting approximately 1 in 20,000 individuals

(Korenberg et al., 2003). Unlike the variable and largely underdetermined etiology of ASD, WS is caused by the microdeletion of 25 – 28 genes on chromosome 7q11.23 (Hillier et al., 2003). Predominant characteristics of WS include mild-to-moderate intellectual impairment (Searcy et al., 2004) and a hyper-sociable behavioural phenotype (Jarvinen et al., 2013). People with WS often experience difficulties interpreting social nuances and forming and maintaining relationships, especially with peers (Davies et al., 1998). The social profiles of WS and ASD are both viewed as atypical, but the atypicalities are likely to be syndrome-specific (Tager-Flusberg et al, 2006). For example, individuals with WS show an extreme prosocial drive, with excessive face attention, in particular to the eye region (Riby & Hancock, 2009). In contrast, individuals with ASD show a lack of gaze fixation on the eye region (Riby & Hancock, 2008), and may be considered socially aloof (Wing, 1981).

Little is known about how individuals with WS regulate their personal space, and whether this has any bearing on their social interaction style. The inability to interpret and regulate appropriate interpersonal distance may intensify everyday social vulnerability for both ASD and WS individuals. However, we do not know whether the nature of these interactions may be qualitatively different across syndromes.

In the current study, we adopted the questionnaire-based approach to collect data on social distancing, previously employed by Kennedy and Adolphs (2014). Our aim was to measure parent reports of social functioning in relatively large, multi-site samples of individuals with ASD and WS using the Social Responsiveness

Scale (Constantino & Gruber, 2005) to compare social profiles between ASD and WS groups. We then sought to verify the robustness of the findings offered by Kennedy and Adolphs (2014) on personal space violations in ASD, as well as offering the first insight into personal space regulation in WS, and directly compare social distancing abnormalities between ASD and WS groups. Based on the work of Kennedy and Adolphs (2014) and what is known about the WS social phenotype,it was hypothesised that the parents of both the ASD and WS individuals would be more likely to report interpersonal distance atypicalities than the parents of typically developing individuals.

5.2 Method

5.2.1 Participants

Parent reports were provided for individuals with ASD (n = 101; mean age = 13.5; age range = 8 - 37), WS (n = 77; mean age = 15.3; age range = 4 - 36) and typically developing individuals (n = 118; mean age = 13.5; age range = 3 - 36). Diagnosis of an ASD had previously been confirmed using the Social Communication Questionnaire (SCQ; Rutter et al., 2003), the ADOS or the ADI-R, and all individuals with WS who participated had previously had their diagnosis confirmed with positive *fluorescent in situ hybridization* (FISH) testing. The typically developing individuals were not reported to have any difficulties with everyday functioning or to have any developmental or neurological deficits. A one way ANOVA revealed that there was no significant difference in chronological age across the three groups (p = .09; see Table 5.1).

5.2.2 Social Responsiveness Scale

The parent report SRS (Constantino & Gruber, 2005) is a 65-item questionnaire that measures the normality/abnormality of social functioning. It was originally designed not just as an autism screener, but also to detect milder traits of autism in the typically developing population. As such it has been used in a range of typical and atypical populations (Barttfeld et al., 2013; Channell et al., 2015; Klein-Tasman, Li-Barber, & Magargee, 2011; Riby et al., 2014). Each item is coded on a scale of 0 – 3, and scores are generated across five subscales: social awareness (e.g. – aware of what others are thinking or feeling), social cognition (e.g. – recognizes when something is unfair), social communication (e.g. – is able to communicate feelings to others), social motivation (e.g. – self-confident when interacting with others) and autistic mannerisms (e.g. - has an unusually narrow range of interests). Higher scores on these subscales are indicative of greater impairments. Of interest, item 55 directly addresses interpersonal space ("Knows when he or she is too close to someone or is invading someone's space"). Kennedy and Adolphs (2014) also noted three other items which were highly correlated with this statement: item 52 ("Knows when he or she is talking too loud or making too much noise"), item 56 ("Walks in between two people who are talking") and item 63 ("Touches others in an unusual way e.g., he or she may touch someone just to make contact with them then walk away without saying anything"). These items were therefore examined independently as part of a separate interpersonal space subdomain.

5.2.3 Procedure

The study was a multi-site project between UK, USA1, Australia and Ireland. Parents completed the questionnaires and returned them to the researcher. Ethical approval was obtained from all the host institutions.

5.3 Results

5.3.1 Profiles of social functioning in ASD and WS

An initial one-way ANOVA was conducted to assess differences in overall social functioning of individuals with ASD, WS and TD controls using total score on the SRS as completed by parents. The results revealed a statistically significant effect of group/diagnosis on total SRS T-score ($F(2, 293) = 406.2$, $p<.001$). A Tukey post-hoc comparison revealed that the total SRS score for the ASD group (mean = 110.0, ±25.0) was significantly higher than both the WS group (mean = 84.4, ±32.5; $p<0.001$) and the TD group (mean = 21.1, ±17.1; $p<0.001$). Likewise, the WS group scored significantly higher than the TD group ($p<0.001$; see Table 5.1).

The mean total SRS T-score for both the ASD and the WS group was in the severely abnormal range, whereas the TD group was within the normal range of social functioning (see Table 5.1). Crucially, only 1 per cent of the ASD group, and 18 per cent of the WS group were reported by parents to function within the 'normal' range (compared to 92 per cent of the typically developing group).

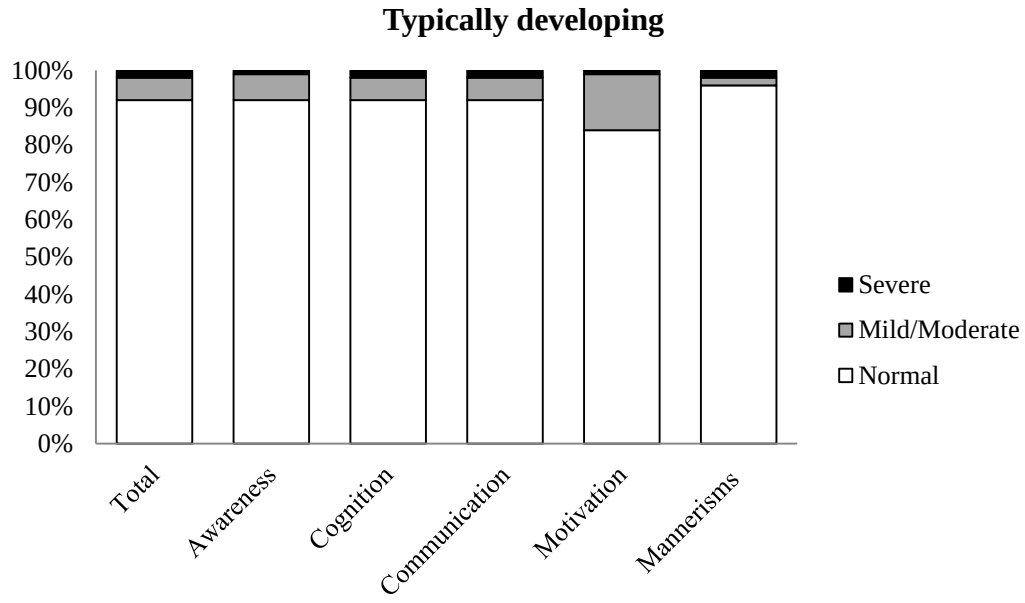
Table 5.1. Participant characteristics and SRS scores for individuals with ASD, WS and those who are typically developing

	ASD (n = 101)	WS (n = 77)	TD (n = 118)
Mean age (± SD)	13.5 (±5.1) years	15.3 (±8.3) years	13.5 (±5.8) years
Males/Females (%)	84/16	51/49	60/40
SRS T scores			
Total score	110.0 (±25.0)	84.4 (±32.5)	21.1 (±17.1)
Social awareness	68.37 (±12.6)	64.7 (±15.45)	45.47 (±9.04)
Social cognition	81.61 (±9.32)	78.14 (±13.06)	44.74 (8.97)
Social communication	80.94 (±9.26)	71.52 (±14.47)	44.69 (±9.4)
Social motivation	77.75 (±11.96)	62.31 (±13.56)	48.26 (±9.73)
Autistic mannerisms	84.43 (9.55)	79.3 (±13.5)	45.99 (±8.09)

Exploring patterns at the subscale level, there were significant effects of group on all five sub-domains of the SRS, assessing social awareness, social cognition, social communication, social motivation, and autistic mannerisms (all $p<0.001$; Table 5.1). Post hoc comparisons showed that the WS and ASD groups both scored significantly higher than the TD group in all five domains ($p<0.001$; Tukey HSD). The ASD and WS groups also scored significantly different to each other in the sub-domains of communication, motivation and mannerisms as the ASD group were more atypical (all at $p<0.001$), but the groups did not differ on the social awareness subscale ($p = 0.12$) and the social cognition subscale ($p = 0.07$). The difference in the profile of individuals categorised in the severe, moderate and normal ranges across these domains is displayed in Figure 5.1 and

shows the similarities / differences in the profiles between the two clinical groups. The mean T scores in all sub-domains indicate that the ASD and WS groups did not show social functioning in the 'normal' range in any of the five areas.

A



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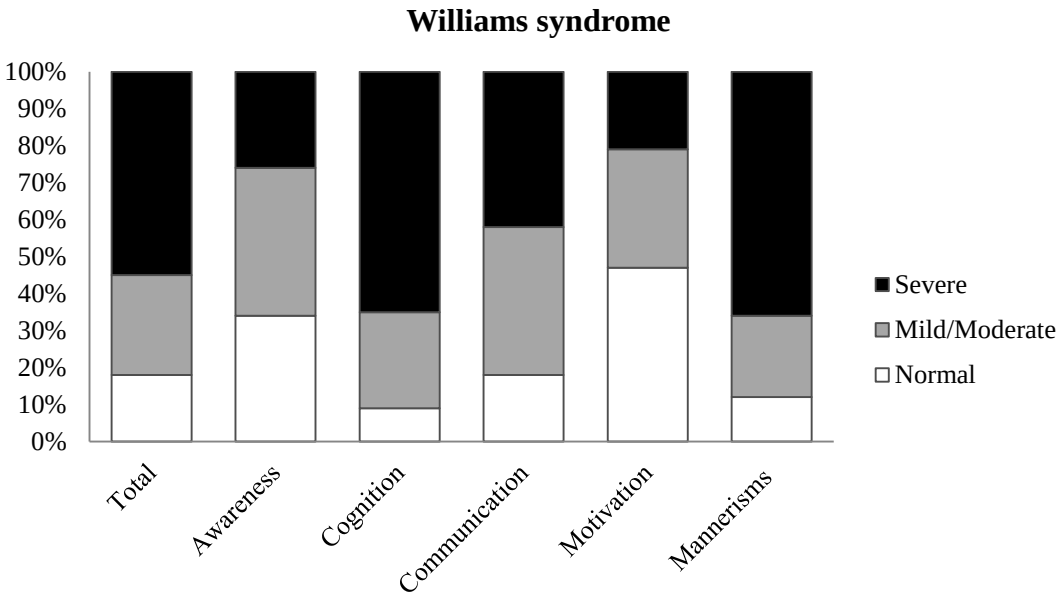

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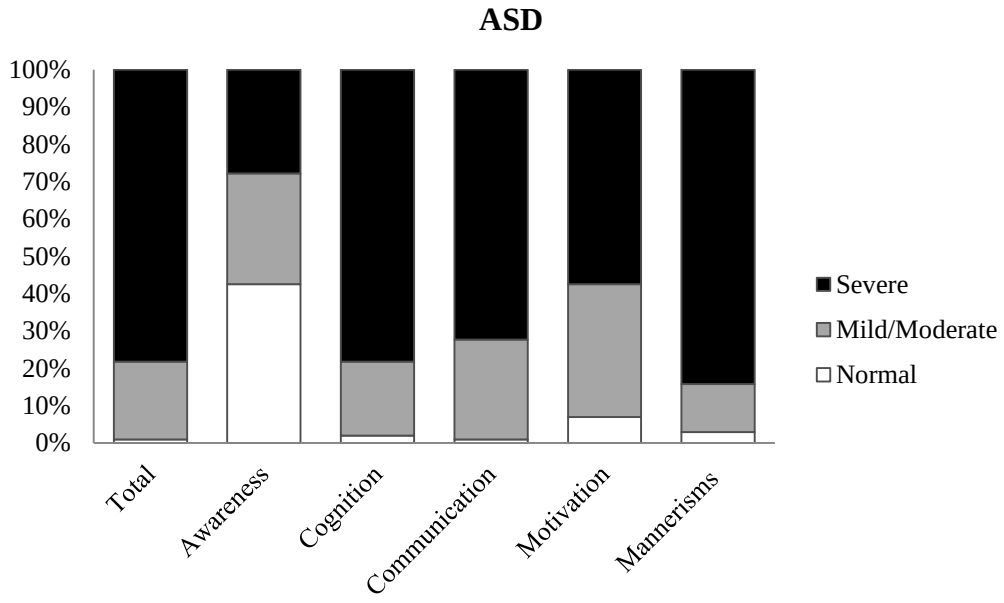


Figure 5.1. The percentage of individuals classified in the severe, mild-moderate and normal range using the SRS for individuals with ASD, WS and TD.

5.3.2 Personal space in ASD and WS

In order to specifically assess the characteristics of personal space across individuals with ASD, WS and those who are TD, a Kruskal-Wallis H test was conducted to explore the parent ratings. It was found that scores significantly differed on this item between the three groups ($H(2) = 114.2$, $p<0.001$); with a mean rank item score of 173.3 for the ASD group, 208.6 for the WS group and 88.1 for the TD group. These rank scores suggest that on average the WS group was reported to be significantly less aware of someone else's personal space

(i.e., more atypical in this behaviour) compared to the ASD group (U = 2899, Z = -3.12, p<0.001) and to the typically developing group (U = 903, Z = -9.93, p<0.001, Mann-Whitney U test; Figure 5.2). The ASD group were also reported to be less aware of invading another person's personal space when compared to the typically developing group (U = 2466, Z= - 7.89, p<0.001, Mann-Whitney U test). Therefore, individuals with ASD and WS were reported by parents to display personal space difficulties in comparison to TD children, however the parents of those with WS reported the greatest deficits.

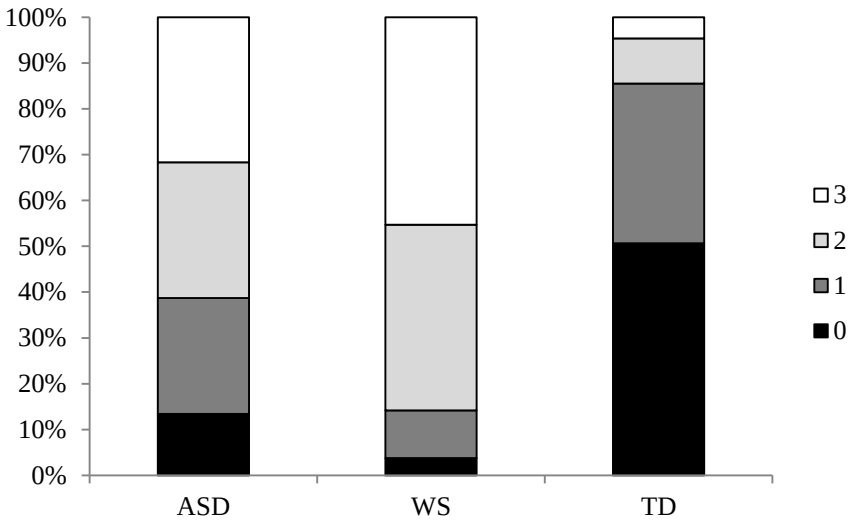


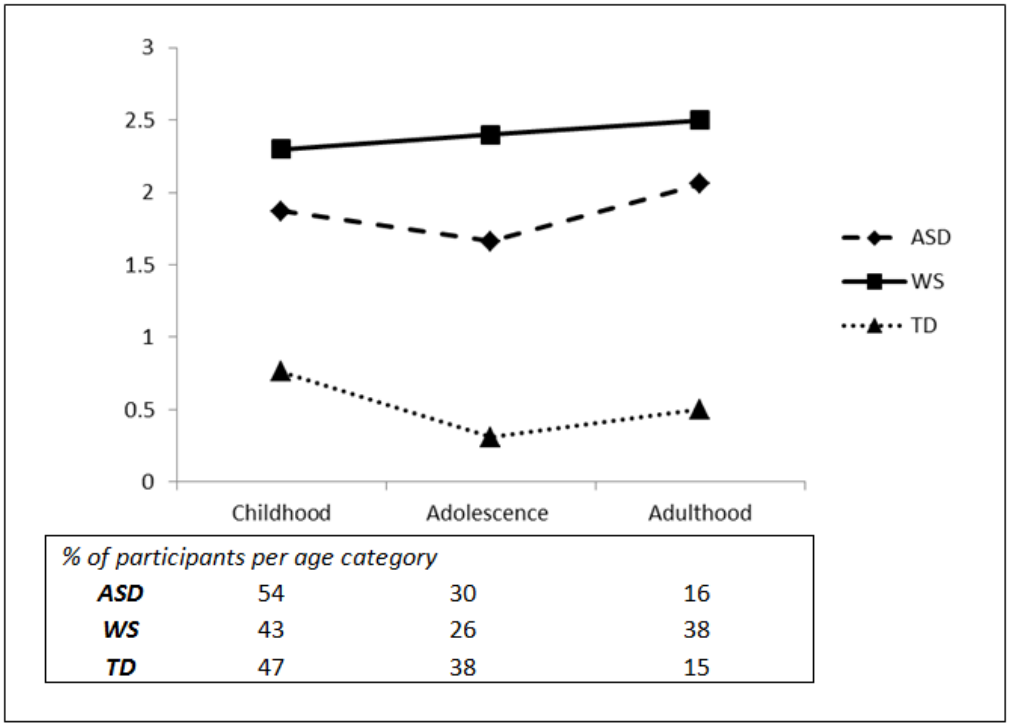
Figure 5.2. Percentage of scores relating to each group on item 55. Scores range from 0 – 3, with higher scores highlighting greater social distancing abnormalities.

As suggested by Kennedy and Adolphs (2014), our sense of space is a multimodal construct. Thus, we looked for items on the SRS which were highly correlated with item 55 regarding personal space. We found that item 52 (the volume at which words are spoken) had a high correlation with item 55 in both

the ASD and WS groups (r = 0.35, r = 0.47 respectively, both at $p<0.001$), but not the TD group (r = 0.29, $p<0.001$). Likewise item 56 (walks in between two people who are talking) was highly correlated with item 55 in the WS group and the TD group (r = 0.4, r = 0.4 respectively, both at $p<0.001$), but not in the ASD group (r = 0.08, $p = 0.46$). Both of these items relate to the broad construct of social distancing.

5.3.3 The impact of age on personal space

To explore the impact of age on personal space judgements, the participant groups were split into broad age categories of 'Child' (age 3-12 years), 'Adolescent' (age 13 – 17 years) and 'Adult' (age 18+years) and the data were explored for item 55. Figure 5.3 illustrates the lack of developmental change in response to this item in the clinical groups, thus suggesting there is little evidence of an age-specific atypicality. These data were analysed using a Kruskal-Wallis H test which revealed that there was no significant difference between the three age categories for the ASD group ($H(2) = 1.53$, $p = 0.46$) or for the WS group ($H(2) = 0.74$, $p = 0.69$). In the TD group, a significant difference was found ($H(2) = 7.09$, $p<0.05$), with children being significantly less aware of invading another person's personal space than adolescents ($U = 912.5$, $Z = 2.64$, $p<0.05$; Mann-Whitney U test) but no difference between adolescents and adults. Some caution is required due to the uneven proportion of participants per age category in each group.



% of participants per age category			
ASD	54	30	16
WS	43	26	38
TD	47	38	15

Figure 5.3. Group average scores on item 55 per age, with the percentage of participants per group in each of the age categories. Scores range from 0 – 3, with higher scores highlighting greater social distancing abnormalities.

5.4 Discussion

By analysing parent-reported Social Responsiveness Scale scores acquired from relatively large samples of individuals with ASD and WS, the current study identified significant difficulties of social functioning, variation across subdomains of communication, social motivation, and autistic mannerisms, and specific impairment in the regulation of appropriate interpersonal distance in the two clinical groups. Crucially, individuals with ASD and with WS were reported by their parents to be less aware of invading another person's personal space, compared to reports from parents of typically developing children. In line with

the findings of Kennedy and Adolphs (2014), we also found that a lack of awareness of other people's personal space was correlated with abnormalities in other forms of social distancing. For example, a lack of awareness of personal space may also manifest itself as atypicality of invading another person's space with intrusive loud noise. These data therefore suggest that personal space is a 'multimodal construct' that may be regulated in an atypical manner by individuals with ASD and those with WS. Such atypicalities are highly likely to feed into the profiles of atypical social interaction we associate with both of these developmental disorders and impact upon the range of social difficulties experienced in daily living for both groups.

Interestingly, parents of individuals with WS rated their sons / daughters as being the least aware of another person's space boundaries compared to TD and ASD individuals, thus showing severe abnormality in this domain of social functioning. The findings offer the first insight into interpersonal distance regulation abilities in individuals with WS and suggest that, like individuals on the autism spectrum, this group can also struggle with personal space behaviours, perhaps to an even greater extent. The data here strongly support anecdotal evidence from parents of individuals with WS in terms of the nature of their interactions with unfamiliar people. Given the wide ranging reports of hypersociability associated with the disorder (e.g. Frigerio et al., 2006), increased approach to unfamiliar people (e.g. Jones et al., 2000) and a lack of stranger awareness (e.g. Riby et al., 2014), prolonged fixation on faces during an interaction (e.g. Riby & Hancock, 2008) and generally reduced intellectual capacity to accurately interpret cues during an interaction (e.g. Searcy et al.,

2004), a dysregulation of personal distancing may play a crucial role in social vulnerability of individuals who have the disorder (e.g. Lough et al,. 2014). This issue clearly warrants further investigation using a variety of methods to probe its relation to other components of the social profile and wider aspects of the disorder. In addition, it motivates the need to develop interventions that teach individuals with WS and ASD how to maintain appropriate space between themselves and others. This is relevant across ages in these developmental disorder groups given the lack of evidence of developmental change with regards to personal space distancing in the current data.

Similarities between individuals with ASD and WS in social distancing abnormalities are of particular interest as the two developmental disorders have been considered to be associated with such different social profiles (e.g. Brock, Einav & Riby, 2008). Individuals with ASD are often considered to be hypo-social with a lack of social priority for people, which can be very different from the hyper-sociability and extreme social motivation towards people that has been associated with WS (e.g. claims of a prosocial drive, Frigerio et al, 2006). Despite these differences, both groups are considered socially vulnerable (Lough et al., 2014) as a consequence of the atypicalities of their social profiles and the impact upon daily functioning. Indeed, the current study shows that both group show atypical social distancing regulation that will feed into those atypical social profiles. It follows that appropriate social distancing plays a vital role in positive social interactions (Gessaroli et al., 2013), and positive social interactions can be protective against social vulnerability.

The role of the neural systems underpinning social behaviour regulation is of interest in light of our findings, especially social distance. Kennedy et al. (2009) demonstrated that a patient with bilateral amygdala damage (known as patient SM) also showed substantially reduced personal space boundaries. They suggested that the amygdala is therefore a key component of the neural substrate regulating interpersonal distance. This proposal drew strength from the findings of Gessaroli et al. (2013) and Kennedy and Adolphs (2014) who found diminished personal space regulation in individuals with ASD, a condition with known anatomical abnormalities of the amygdala (Baron-Cohen et al., 2000). Individuals with WS are also known to have structural and functional abnormalities of the amygdala (e.g. Bellugi et al.1999; Haas et al., 2010; Haas et al., 2009; Meyer-Lindenberg et al., 2005), and the current study has shown that these individuals demonstrate severely impaired interpersonal distance awareness. However, although amygdala function may play a role in interpersonal space this region does not necessarily function in the same manner for individuals with WS as it does with ASD. Indeed when engaged in viewing faces individuals with WS show reduced amygdala activation whereas individuals with ASD shown amygdala hyper- responsiveness (Kliemann et al., 2012). Therefore, it remains speculative as to whether interpersonal distance regulation could be an endophenotype for amygdala dysfunction in WS and ASD (Kennedy & Adolphs, 2014).

An alternative explanation is offered by the frontal lobe hypothesis. Frontal lobe dysfunction is thought to be related to impaired response inhibition (Porter et al., 2007). Parallels in social functioning, and specifically approach behaviour, have

been drawn between patients with frontal lobe damage and those with WS. Porter and colleagues (2007) suggest that whilst these individuals report knowing not to approach a stranger, they have difficulty inhibiting the impulse they experience to carry out this behaviour. This lack of inhibitory control has also been shown in individuals with ASD (Christ et al., 2006). Therefore, the atypical interpersonal distance findings in the current study for these two groups could be in part explained by their lack of inhibitory control.

Anxiety has also been shown to mediate social behaviour in WS (Kirk et al, 2013) and it is suggested that high levels of anxiety (which are present in both WS and ASD; Riby et al., 2014; Rodgers et al., 2012) may influence the ability to process socially meaningful stimuli. It is therefore possible that mental health issues associated with the disorder are impacting the ability to gauge appropriate social behaviour, and thus affecting interpersonal distance regulation. Considering the contribution of each of these previously mentioned theoretical standpoints to our understanding of interpersonal distance regulation in these clinical groups is a challenge for future research and emphasises the need to consider the whole individual and the cognitive / behavioural profiles associated with these disorders in a more comprehensive manner. Certainly the parent report data provided here suggest that future research is warranted in much greater detail. Indeed it is only once we consider the full profile at an individual level and capture both within- and between-disorder variability that we can begin to disentangle the above interpretations.

A significant strength of the current study is the cross-syndrome approach in a large sample size. This is especially insightful considering the rarity of conditions such as WS. However, there remain limitations which should also be addressed. As the SRS was not originally designed to measure social distancing, it lacks the detail and insight that could be acquired through experimental work. The method reported here is not offered as a replacement for observational or experimental work, rather it serves to assess a large sample of individuals with relatively rare developmental disorders, certainly in the case of WS. As social distancing in WS was previously unexplored, this study offers the first insight into whether or not there is an abnormality that requires further attention in this population – and indeed, the results suggest this to be the case. A further limitation is that item 55 only addresses one direction of social distancing abnormalities by only asking about social violations arising from close proximity rather than violations from being abnormally distant from others. It is therefore entirely possible that the few individuals with ASD and WS who score in the typical range for this question still have social distancing abnormalities related to maintaining too great a distance from other people during social interactions. Furthermore, the fact that item 55 did not correlate with other assessments of personal distancing in the ASD group warrants future exploration to explore syndrome-specific patterns of social behaviour atypicality that may inform intervention. Finally, a measure of general cognitive functioning was not taken in this study, which could have mediating effects on the social distancing phenotype. However, previous work on a large sample of individuals with ASD found that social distancing abnormalities cannot be entirely explained by intelligence (Kennedy & Adolphs, 2014).

Moreover, when considering the social vulnerability status of these groups, a stranger can get a strong cue from social distancing abnormalities; however, intelligence levels are more hidden and thus may not be immediately apparent. The stranger therefore only has access to estimates of age based on physical appearance with which to make their decision of how to respond as the individual with WS or ASD approaches. The social impact of inappropriate personal distancing may be compounded by reduced intellectual abilities once an interaction begins, further emphasising the importance of social distancing in individuals with lower IQ.

In conclusion, the current findings provide new evidence that individuals with WS have difficulties with social distance regulation, and are rated to be more likely than individuals with ASD and their typically developing individuals of the same chronological age to infringe upon the personal space of others. However, these preliminary findings need to be followed up with experimental paradigms, and the real-world implications of these behaviours need to be considered for these vulnerable individuals with developmental disorders. By doing so, we will begin to develop a greater understanding of the relationship between interpersonal distance, successful interpersonal interactions and social vulnerability status.

Mid Chapter Summary

The first study in this Chapter highlighted that individuals with WS show social distancing abnormalities, as reported by their parents. However, it was acknowledged that the SRS was not designed to measure personal space, and therefore experimental paradigms are needed to further investigate this issue. Consequently, the second study in this chapter used a mixed methods, multi-informant approach, using both the parent report SRS and a stop-distance paradigm in an attempt to better understand personal space regulation in young people with WS. In Chapter 4, parents reported that personal space regulation was difficult for individuals with WS, but it was especially around strangers that it was of concern. The second study in this Chapter therefore also seeks to investigate the impact of familiarity on personal space regulation, as this could have considerable consequences for the social vulnerability of people with WS.

Personal space regulation in Williams syndrome: The effect of familiarity

5.5 Introduction

When engaging in social interactions, individuals must regulate the distance that they maintain between themselves and other people (Hall, 1996). Personal space is defined as the area around a person's body, which if invaded, can cause feelings of discomfort and anxiety (Perry et al., 2013). Vignemont and Iannetti (2015) outline different types of personal space. Peripersonal space refers to the space around one's body where an object can be grasped, whereas extrapersonal space is the area around the body that is just beyond reach. Indeed, they highlight that there also exists functional differences within the definition of peripersonal space. The Function-Specific Model of personal space identifies two functions of peripersonal space: the protective (or defensive) space (Sambo & Iannetti, 2012) and the working personal space (Rizzolatti et al., 1997). The current study is concerned with the protective peripersonal space (herein referred to as simply 'personal space'), and its implications for social vulnerability levels in Williams syndrome.

For typically developing individuals, regulating this personal space is a largely automatic process, guided by situational cues, social cues and cultural norms (Beaulieu, 2004). The ability to successfully collate these cues and maintain an appropriate interpersonal distance contributes to successful and positive social interactions (Gessaroli et al., 2013). However, it is known that some individuals with developmental disorders find social interactions challenging, and they may also struggle to regulate their personal space. Gessaroli et al. (2013) studied

personal space regulation in children with autism spectrum disorders (n=15, mean 9 years; ASD) and compared them to typically developing (n=23, mean 9 years; TD) children. Using a stop-distance paradigm, they found that children with ASD maintained a greater distance from a confederate compared to their TD peers. Further, whilst TD children were able to regulate their personal space based on the familiarity of the person they were interacting with, children with ASD failed to do so, suggesting that they lack flexibility in personal space regulation.

Kennedy and Adolphs (2014) also examined the issue of personal space in young people with ASD. They used the Social Responsiveness Scale (SRS; Constantinno & Gruber, 2005), which is a 65 item parent report questionnaire designed to measure the typicality/atypicality of social functioning. They were specifically interested in item 55 ("Knows when he or she is too close to someone or is invading someone's space"). In stark contrast to the findings of Gessaroli and colleagues (2013), Kennedy and Adolphs (2014) found that individuals with ASD were more likely to be reported by parents to violate the personal space of others. Indeed, 79% of parents report that their children with ASD have smaller personal space boundaries compared to their TD siblings. However, the different methods of assessment used in these studies do not allow for direct comparisons of results, and if personal space regulation is disorder-specific, then they offer little insight into how individuals with other developmental disorders regulate their personal space.

Williams syndrome (WS) is a rare neuro-developmental disorder, which affects approximately 1 in 20,000 individuals (Korenberg et al., 2003). It is caused by the

microdeletion of 25-28 genes on chromosome 7 (7q11.23; Hiller et al., 2003). Individuals with WS typically have mild-moderate levels of intellectual impairment (Searcy et al., 2004), and experience a powerful prosocial drive to interact with others, i.e., they display a hypersocial behavioural phenotype (Jarvinen et al., 2013). Despite their social nature, individuals with WS can struggle to pick up on social cues, and many find it hard to form and maintain peer relationships, resulting in high levels of isolation (Udwin, 1990). This occurs against a backdrop of high anxiety levels (Stinton et al., 2010). Recent work by Riby and colleagues (2014) found that 46% of children and adults with WS experienced high levels of anxiety, with a mean for this high anxiety group above that found in clinically anxious children (Nauta et al. 2004). Interestingly, they also noted differing patterns of social behaviour, as measured by the Social Responsiveness Scale (SRS). Those individuals who experience higher anxiety showed more severe social dysfunction, suggesting that anxiety levels are linked to social behaviour in WS. As individuals with WS show indiscriminate approach behaviour (Little et al., 2013), and a lack of stranger danger awareness (Riby et al., 2013), their personal space regulation when interacting with others is an important facet when looking at their social vulnerability profile (Jawaid et al., 2012; Lough et al., 2015b).

Lough et al. (2015a) offered the first insights into personal space regulation in WS and ASD, using the same methods employed by Kennedy and Adolphs (2014). They found that individuals with WS were reported by their parents to show the least awareness of personal space boundaries, when compared to reports from parents of individuals with ASD and TD individuals. This is of particular

concern given the wider social vulnerability profile associated with WS (see Jawaid et al., 2012 for a review). Despite the large sample size used by Lough and colleagues, it was acknowledged that the SRS was not designed to measure personal space, and using a method such as the stop-distance paradigm could provide more clarity and insight into this issue. In the current study, we used both the SRS and the stop-distance paradigm to provide multiple measures of personal space regulation in young people with Williams syndrome. Our aim was to obtain a more comprehensive insight into this issue by utilising information from a parent report questionnaire and experimental work involving the individuals with WS themselves. Based on the work of Lough et al. (2015a), as well as what we know about the WS social profile, large effect sizes were predicted, and the hypotheses were as follows: 1) Children and adolescents with WS would receive higher scores on SRS items relating to personal space than their TD peers; 2) In the stop-distance task, young people with WS would let unfamiliar people approach and stand at a closer interpersonal distance than TD children and adolescents, and 3) In the same task, young people with WS would approach and stand closer to an unfamiliar person than their TD peers.

5.6 Method

5.6.1 Participants

Eighteen young people with WS (mean age = 11.4; age range = 8 – 16) and eighteen typically developing children and adolescents (mean age = 11.3; age range = 8 – 16) participated in the study (see Table 5.1). An a priori power analysis indicated that 12 participants were needed in each group to have 80%

power for detecting a large effect size (d = 0.8, α = 0.05). The participants were matched on chronological age and gender. All participants with WS had previously had their diagnosis confirmed using *fluorescent in situ hybridization* (FISH) testing.

5.6.2 Social Responsiveness Scale (Constantinno & Gruber, 2005)

This parent report questionnaire consists of 65 items which measure the typicality/atypicality of social functioning. It has frequently been used in the typically-developing population, but also with young people with WS (e.g. Klein-Tasman et al., 2011; Riby et al., 2014; Lough et al., 2015a). From the responses, five sub-scale scores can be generated in the areas of: social awareness, social cognition, social communication, social motivation and autistic mannerisms (see Table 5.2). Higher scores suggest greater levels of impairment.

There are several items relating to personal space that have been examined in the work of Kennedy and Adolphs (2014) and Lough and colleagues (2015a) and are therefore of interest to the current study. They examined item 55 which asks parents to rate the following statement: "knows when he or she is too close to someone or is invading someone's space". Other items also examined by both Kennedy and Adolphs (2014) and Lough et al., (2015a) refer to multi-modal construct of personal space, including item 52 ("Knows when he or she is talking too loud or making too much noise), item 56 ("Walks in between two people who are talking") and item 63 ("Touches others in an unusual way e.g. he or she may touch someone just to make contact with them then walk away without saying anything").

5.6.3 Spence Children's Anxiety Scale – Parent version (Spence, 1998)

The Spence Children's Anxiety Scale–Parent Version (SCAS-P; Spence, 1998) was used to assess symptoms of anxiety. This issue is particularly relevant due to increased anxiety in WS and reports of an association between increased anxiety and atypical social behaviours (Riby et al., 2014). The SCAS-P has been reported to have good psychometric qualities including a high internal consistency of .92 (Spence, Barrett, & Turner, 2003), and has been used in both TD and clinically anxious populations (Nauta et al., 2004; Spence, 1998). This 38-item parent report questionnaire is divided into six subscales of anxiety relating to panic/agoraphobia, separation anxiety, physical injury fears, social phobia, obsessive compulsive, and generalised anxiety disorder. Parents rate each item on a 4-point Likert scale according to how often their child exhibits the symptoms, from 1 (never) to 4 (always). Their answers are scored from 0 to 3, yielding a maximum possible score of 114. While there is no formal clinical cut-off for the SCAS-P, total SCAS scores of 24 or above have been suggested to indicate clinical levels of anxiety (Spence, 2008).

Table 5.2. Participant characteristics and SRS scores for individuals with WS and those who are typically developing

	WS (n = 18)	TD (n = 18)	*p* value
Mean age (± SD)	11.4 (±2.5) years	11.3 (±2.5) years	
Males/Females (%)	44/56	44/56	
SRS T scores			
Total score	77.17 (±13.37)	44 (±7.98)	*
Social awareness	66.06 (±11.53)	45.67 (±8.04)	*
Social cognition	79.72 (±10.34)	44.22 (±6.62)	*
Social communication	74.5 (±12.95)	46.83 (±8.72)	*
Social motivation	63.39 (±12.57)	47.5 (±7.08)	*
Autistic mannerisms	82.22 (±12.25)	46.28 (±6.09)	*

* = $p < 0.001$

5.6.4 Procedure

A stop-distance paradigm was used to assess preferred interpersonal distance (Kennedy et al., 2009). This procedure has been used extensively for assessing preferred interpersonal distance under different conditions, yielding reports of high reliability and validity (Greenberg, Strube, & Myers, 1980; Hayduk, 1978, 1983, 1985). The task began with the participant standing 3 metres away from the experimenter. There were four conditions: two of which involved completing the task with an unfamiliar person (the experimenter) and the other two were undertaken with a familiar person (mother/father; see Figure 5.4). In condition A the participant was asked to approach the experimenter and stop at a location that felt comfortable to them. The experimenter maintained a neutral expression and

no eye contact was made. Once the participant had decided on a location which felt comfortable to them, a hip-to-hip measurement was taken using a digital laser distance measurer (RZE-40). Three measurements were taken in succession and averaged together. Each condition consisted of three trials. The average distance across these three trials was taken as the preferred distance in each condition. This procedure was repeated for condition B, in which the experimenter approached the participant and the participant instructed them to stop at a distance that felt comfortable. Conditions C and D mirrored the first two conditions, but a familiar person took the place of the experimenter. The conditions were presented in a random order for each participant. Ethical approval was obtained from the host institution.

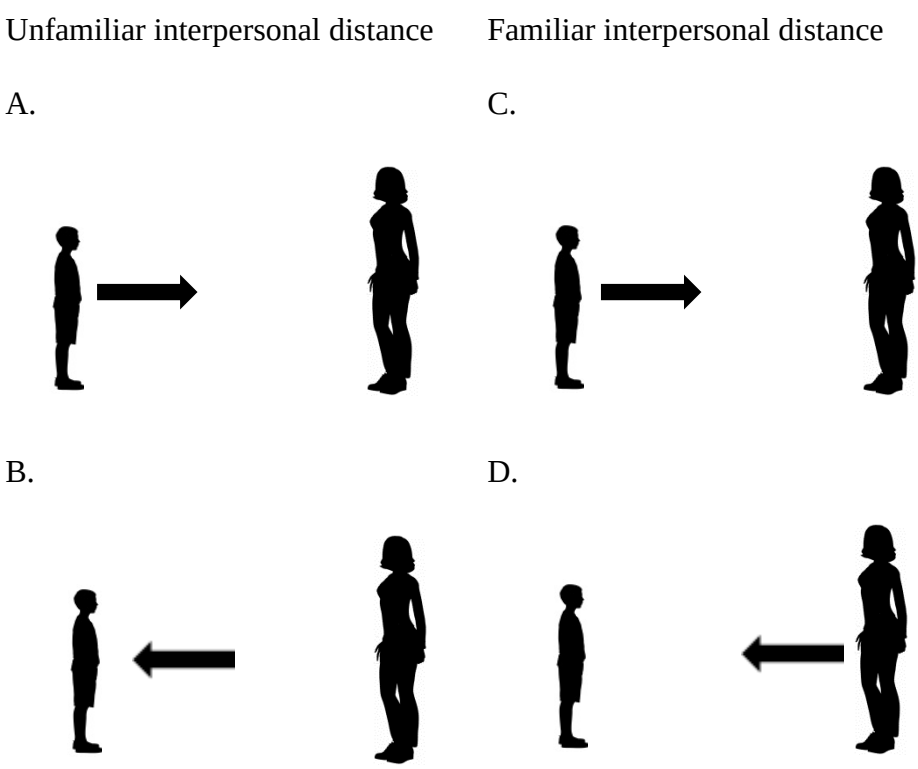


Figure 5.4. Stop-distance paradigm. In condition A, the participant approached the experimenter (unfamiliar). In condition B, the experimenter (unfamiliar) approached the participant. In condition C, the participant approached their parent (familiar). In condition D, the parent (familiar) approached the participant.

5.7 Results

5.7.1 Social Responsiveness Scale

To examine the overall social profile of the two groups, Mann-Whitney U tests were used. We found statistically significant differences between groups on the SRS total T scores, as well as on all of the five subscales (all $p<0.01$). In all cases the WS group showed more atypical social behaviour. 92% of TD young people displayed overall social behaviour that was deemed to be in the normal range of

functioning, whereas only 18% of young people with WS scored within the normal range (this maps directly onto levels reported in van der Fluit et al., 2012 and Riby et al., 2014). Importantly, the results also revealed statistically significant differences between the two groups on the four items in the SRS that relate to personal space, with parents of individuals with WS reporting greater atypicalities in their son/daughter on these items (all $p<0.01$; Mann-Whitney U tests; see Figure 5.5).

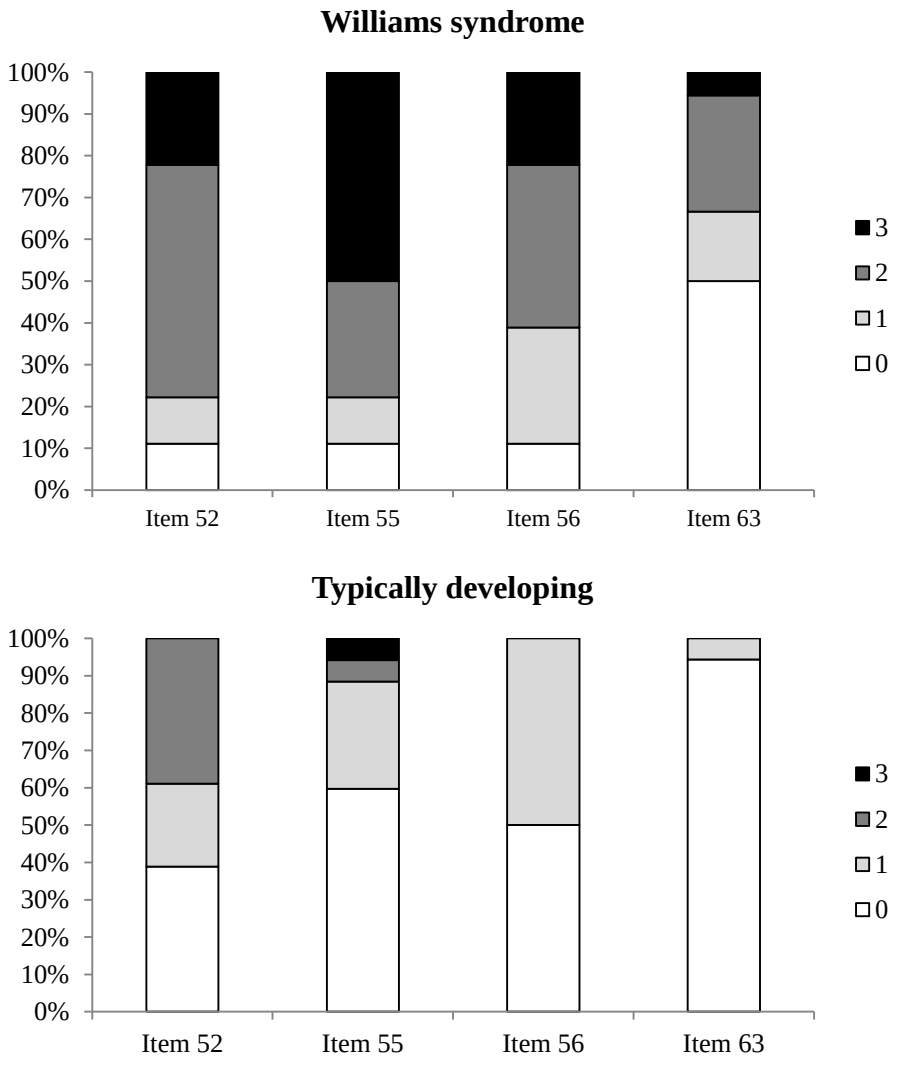


Figure 5.5. The percentage of scores on item 52, item 55, item56 and item 63 on the SRS for children with WS and TD children. Higher scores indicate greater atypicalities of behaviour. * Item 52: "Knows when he or she is talking too loud

or making too much noise; item 55: “Knows when he or she is too close to someone or is invading someone’s space”; item 56: “Walks in between two people who are talking”; item 63: “Touches others in an unusual way e.g. he or she may touch someone just to make contact with them then walk away without saying anything”.

5.7.2 Stop-distance paradigm

Being Approached by an Adult

A two-way mixed methods ANOVA was used on the measurement of interpersonal distance (m), with Familiarity (unfamiliar, familiar) as a within-subjects variable and Group (TD, WS) as a between subjects variable. There was a significant main effect of Familiarity ($F(1,34) = 4.74$, $p < 0.05$; $\eta_p^2 = 0.12$), showing that participants maintained a larger distance when approached by an unfamiliar person compared to a familiar person. There was also a significant main effect of Group ($F(1,34) = 4.75$, $p < 0.05$; $\eta_p^2 = 0.12$), as young people with WS showed reduced interpersonal space compared to their TD peers when they are being approached. Crucially, there was a significant interaction between Familiarity and Group ($F(1,34) = 15.18$, $p < 0.001$; $\eta_p^2 = 0.31$; see Figure 5.6), showing that individuals with WS maintained a much closer distance when approached by unfamiliar people than TD individuals do, with a large effect size .

Post hoc analyses showed that the WS group (M = 0.74, SD = 0.29) and the TD group (M = 0.75, SD = 0.2) did not significantly differ in their interpersonal distance when approached by a familiar person ($t(34) = -0.13$, $p = 0.9$, $d = 0.04$; independent t-test). However, when approached by an unfamiliar person, the WS

group (M = 0.68, SD = 0.26) let the unfamiliar person stand at a significantly closer distance to them than the TD group (M = 0.98, SD = 0.22; t (34) = -3.7, p< 0.001, d = 1.3). Indeed, there was a significant difference for the preferred interpersonal distance of the TD group when approached by familiar versus unfamiliar people (t (17) = 4.8, p<0.001), but there was no significant difference for the WS group (t (17) = 1.28, p = 0.22).

Approaching an Adult

The above tests were repeated in order to examine interpersonal distance when the child was doing the approaching. A two-way mixed methods ANOVA on Familiarity (unfamiliar, familiar) and Group (TD, WS) showed a significant main effect of familiarity ($F(1,34) = 15.6$, $p < 0.001$; $\eta_p^2 = 0.31$). When the child is the one approaching, there was no significant main effect of Group ($F(1,34) = 2.71$, $p = 0.11$; $\eta_p^2 = 0.07$). However, there was a large effect size and a trend towards a significant interaction between Familiarity and Group ($F(1,34) = 3.72$, $p = 0.06$; $\eta_p^2 = 0.1$; see Figure 5.6), with the WS group coming much closer when they approach unfamiliar people compared to the TD group. Between subjects *t*-tests revealed that there was no difference between groups when approaching a familiar person (WS: M = 0.7, SD = 0.23; TD: M = 0.76, SD = 0.22; t (34) = -0.77, p = 0.44). There was, however, a significant difference between groups when approaching an unfamiliar person, with the TD group standing further away (M = 0.97, SD = 0.31) than the WS group (M = 0.78, SD = 0.25, t (34) = -2.08, p<0.05, d = 0.7). The TD individuals showed a significant difference between the distance they maintained when approaching a familiar adult compared to an

unfamiliar adult (t(17) = 6.1, p<0.001), whereas no significant difference was found for the WS group (t(17) = -0.99, p = 0.34).

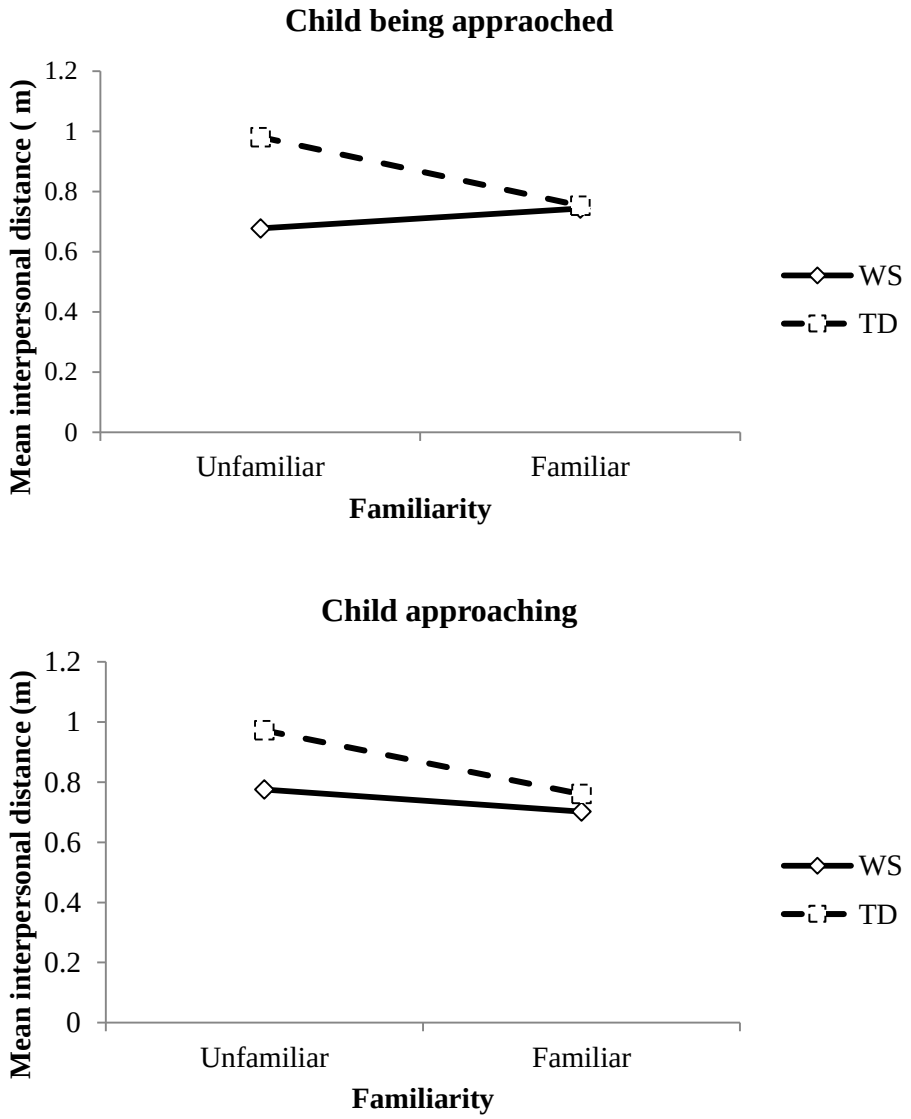


Figure 5.6. Interaction graphs showing interpersonal distance when the child is being approached by / is approaching familiar and unfamiliar people.

5.7.3 Factors impacting on personal space regulation

Age

In the WS group, there was no significant correlation between age and the distance these individuals maintained between themselves and the experimenter (r = 0.28, p = 0.27). However, there was a correlation between age and how close they let a stranger come to them, with older individuals requesting that the unfamiliar person kept a greater distance (r = 0.53, p<0.05). There was no correlation between age and interpersonal distance found in any of the conditions for TD individuals

When looking at age effects on item 55 of the SRS, there was a significant negative correlation between age and the likelihood of violating another person's personal space in the WS group (r = -0.62, p = 0.01). No significant correlation was found between age and item 55 for the TD group (r = -0.08, p = 0.75).

Anxiety

There was a significant difference between the WS group (M = 26.78, SD = 14.38) and the TD group (M = 14.56, SD = 6.32; t(34) = 3.3, p<0.01, d = 1.1) on their total SCAS scores, with higher anxiety in the WS group. There was no significant correlation between anxiety (SCAS total scores) and the distance maintained when approaching a familiar (WS: r = -0.29, p = 0.24; TD: r = -0.25, p = 0.32) or unfamiliar person (WS: r = -0.22, p = 0.07; TD: r = -0.43, p = 0.07). Anxiety did not correlate with approach by a familiar person (WS: r = -0.62, p = 0.81; TD: r = -0.21, p = 0.39), but there was a significant negative correlation

between anxiety and how close both groups let an unfamiliar person stand from them (WS: r = -0.55, p<0.05; TD: r = -0.52, p <0.05), suggesting that young people high in anxiety let unfamiliar people stand closer to them. Although this might appear counterintuitive this result requires further investigation as it may be that there is a bi-directional effect where anxiety also feeds off an inability to make appropriate judgements about others in such social situations.

5.7.4 Relationship between scores on the Social Responsiveness Scale and stop-distance paradigm

There was no significant correlation between the four conditions and the individuals' total SRS t scores in the WS group (Condition A: r = -0.04, p = 0.87; Condition B: r = -0.38, p = 0.12; Condition C: r = -0.24, p = 0.33; Condition D: r = -0.02, p = 0.94), or in the TD group (Condition A: r = -0.14, p = 0.59; Condition B: r = -0.02, p = 0.95; Condition C: r = -0.03, p = 0.91; Condition D: r = -0.09, p = 0.71). Similarly, overall approach behaviour was not significantly correlated with the SRS items related to personal space in the WS group (Item 52: r = 0.12, p = 0.64; Item 55: r = -0.16, p = 0.53; Item 56: r = -0.19, p = 0.44; Item 63: r = -0.34, p = 0.17) or the TD group (Item 52: r = -0.21, p = 0.39; Item 55: r = -0.26, p = 0.3; Item 56: r = -0.1, p = 0.69; Item, 63: r = -0.12, p = 0.63). Of interest, there was a significant negative correlation found between being approached by an unfamiliar person (Condition B) and levels of social awareness on the SRS (r = -0.54, p<0.05); i.e., those with the most impaired levels of social awareness let unfamiliar people come closer to them.

5.8 Discussion

Individuals with WS are reported by their parents to be more likely to violate the personal space of others than their TD peers, supporting our first hypothesis. This replicates the findings by Lough et al (2015a); reinforcing the notion that interpersonal distance regulation is highly atypical in individuals with WS, although the participant demographics between the two groups differ. In the second part of the study, the stop-distance paradigm was utilised, and found that individuals with WS maintained an overall shorter interpersonal distance than the TD individuals. Indeed, differences between the young people with WS and their TD peers were found only in their interpersonal distance around unfamiliar people, not familiar people. Specifically, young people with WS maintained a smaller interpersonal distance when approaching, and when being approached by, unfamiliar people, supporting the second and third hypotheses. Taken together with the findings from the SRS, it would seem that young people with WS show atypical interpersonal distance behaviour, and struggle to regulate their personal space in accordance with the familiarity of the person with whom they are interacting. This is the first study to empirically study social distance violations in this population and shows that this is a critical issue in Williams syndrome.

Our current findings also highlighted a significant negative correlation between anxiety and how close both individuals with WS and TD individuals let an unfamiliar person stand from them. This finding requires further investigation. It may be particularly relevant here to examine the role of social cognition in personal space regulation or vice versa. Individuals with WS have been shown to have deficits in understanding and predicting the actions of others (Sparaci et al., 2012). They show more pronounced difficulties in understanding ‘what’ action is

being performed than individuals with ASD, and when compared to TD individuals matched on chronological and mental age (Sparaci et al., 2014). Both individuals with WS and those with ASD also showed impaired 'why' understanding. This may be pertinent in the context of the stop-distance paradigm, in which an approaching act is being performed, and participants are required to read the observed actions in order to respond appropriately. Indeed, Tager-Flusberg and Sullivan (2000) proposed that there exists dissociation in WS between perceptual and cognitive components of social intelligence. They argue that whilst many individuals with WS are able to make immediate judgements about the mental states of other people (perceptual judgements), they struggle to make inferences about the content of these mental states (cognitive judgements). Elements of social cognition are likely to have a direct bearing on social approach behaviour in WS, and therefore require further investigation.

These findings are of particular concern given what is known about the indiscriminate approach behaviour using face-rating tasks with WS adults (Jones et al., 2000) and the lack of stranger danger awareness in WS (Riby et al., 2014). Invading the personal space of others, particularly strangers, can also transfer fallacious social intentions (Kaitz et al., 2004). If individuals with WS are more likely to approach strangers, invade their personal space and not have an awareness of the dangers this could pose, then they could be facing significant levels of risk during social interactions (Lough et al., 2015a). These issues become even more problematic when combined with the reduced intellectual functioning of individuals with WS (Searcy et al., 2004), staring at faces (Riby & Hancock, 2008), and problems interpreting socio-communicative signals (Porter

et al., 2007) meaning that they may miss important subtle cues from those with whom they are approaching and interacting.

The role of the neural structures underlying social behaviour in WS is of particular interest, given the current findings on atypical personal space regulation. Frontal lobe dysfunction has been related to the hypersociability behavioural phenotype observed in WS. It is known that regions in the frontal lobe are involved in regulating and suppressing socially inappropriate actions (Meyer-Lindenberg et al., 2005). Porter et al. (2007) drew parallels between social approach behaviour in individuals with WS, and individuals with frontal lobe damage. They observed how both groups displayed impulsive social behaviour, which they attributed to impairments in response inhibition. Indeed, Little et al. (2013) proposed that frontal-lobe controlled response inhibition was indicative of social approach behaviour. Frontal lobe theory and lack of inhibitory control have also been implicating in ASD (Christ et al. 2007), leading Lough et al. (2015a) to suggest that difficulty with inhibitory control could be used to explain atypical social distancing regulation in individuals with WS and with ASD.

Alternatively, the amygdala theory has been proposed to help explain the atypical social behaviour seen in WS, and indeed ASD (Jawaid et al., 2012). The amygdala is involved in processing and recognising emotions from faces, generating and controlling anxiety, and mediating eye gaze (Fried, MacDonald & Wilson, 1997). Kennedy et al. (2009) showed that a patient with bilateral amygdala damage showed considerably reduced personal space boundaries.

Increased amygdala volume has been repeatedly found in individuals with WS (e.g. Haas et al., 2014). When viewing faces, individuals with WS show reduced activation of the amygdala compared to controls (Kliemann et al. 2012). It may therefore be that abnormalities in amygdala development could be central to the deficits in social judgement and face perception processing seen in WS, which results in atypical emotional reactions and social behaviour linked to social distance regulation. However, as emphasised by Lough et al. (2015a), the current methodology does not allow for further differentiation between these explanations due to the methodology used.

There was no significant relation between age and how close young people with WS stood from other people, but there was a relation between age and how close they let other people stand from them (although caution is advised when interpreting these findings due to the small sample size). Older individuals asked the unfamiliar person to maintain a greater distance than the younger participants did, implying that as they get older, they become more protective of their own personal space boundaries, but still lack awareness of invading other people's personal space boundaries. It is thought that children display adult-like personal space regulation by age 12 years (Aiello, 1987), which could explain why there were no relation between age and personal space in the TD group who had a mean age of 11.3 years.

The relationship between interpersonal distance and age is less clear when looking at the findings from the SRS. Results from the current study show that there was a significant negative correlation between age and Item 55 ("Knows

when he or she is too close to someone, or is invading their personal space"), suggesting that as children get older, their parents rate them as being less likely to invade the personal space of other people. This is in contrast to both the behavioural data collected from the stop-distance task, and also the findings from Lough et al. (2015a), who did not observe any age related changes in interpersonal distance for individuals with WS. However, Lough et al. (2015a) employed a wide age range of 4 – 36 years and, for the purposes of their age analyses, split the sample into "childhood", "adolescence" and "adulthood" age categories, which could not be done in the present study. It seems likely that the different age range of the sample and analysis method contributed to the differences observed. Future work tracking the developmental trajectory of personal space regulation in individuals with WS, and other developmental disorders, is warranted.

Interestingly, our results show that there was not a significant correlation between SRS scores addressing personal space and distance on the stop-distance paradigm, which could explain why the relationship between age and interpersonal distance differed depending on the methodology used. As the items on the SRS relating to personal space are scored on a 0-3 scale, it is likely that there is not enough variability in scores to allow for meaningful correlations (in addition to a relatively small sample size). Nevertheless, the reliability of measures used to assess personal space still requires further investigation, particularly in individuals with an intellectual disability (ID).

Despite the contrasting findings on the impact of age, there are congruent findings of atypical personal space regulation in WS across studies (e.g. Lough et al., 2015a, Gessaroli et al., 2013, Kennedy & Adolphs, 2104) and methodologies (i.e., SRS and stop-distance paradigm). Yet, findings on personal space in ASD show discrepant findings depending on the methodology used. Like individuals with WS, individuals with ASD experience high levels of anxiety (Rodgers et al., 2012). Recent work by Perry et al. (2015) suggested that the discrepancy in findings on interpersonal distance in the ASD literature could be partially explained by levels of social anxiety (SA). Perry et al., (2013) found a positive correlation between SA traits and interpersonal distance preference, with individuals with high SA traits preferring to stay further away from a stranger compared to those with low SA traits. Future work on interpersonal distance in ASD would therefore benefit from careful consideration of the anxiety profiles of the participants included in the sample.

This study offered a novel insight into the issue of personal space regulation in young people with WS, and has clear real world implications for these individuals. However, the limitations of this work merit attention. First, a relatively small sample size was used. This is due to the rarity of the condition, meaning relatively small sample sizes are often seen across the WS literature. The number of participants is also comparable to the sample size of the previously mentioned stop-distance study by Gessaroli et al. (2013) involving children with autism. Second, the stop-distance paradigm could be seen as an artificial task, and may not capture behaviour that is reflective of real life. This task has been used in several previous studies on personal space (e.g. Gessaroli et al., 2013; Kennedy et

al., 2009), but further observational work would be welcomed to offer more insight into this behaviour. Third, while the findings appear congruent with the WS behavioural phenotype, it is unclear whether the patterns observed are specific to WS, or rather a feature of having an ID. Future work would therefore benefit from the inclusion of an ID comparison group. Fourth, IQ data were not collected for participants in the current study. Previous work with individuals with ASD has shown that social distancing abnormalities cannot entirely be explained by intelligence levels (e.g. Kennedy & Adolphs, 2014); however, the inclusion of IQ assessment is an important next step in furthering the work on interpersonal distancing in WS. This would be particularly useful in further exploring the age-related findings outlined in the current study. Finally, although the familiarity of the person conducting the stop-distance task was manipulated, all testing took place in familiar environments. It may be, therefore, that the young people with WS viewed the stranger as a 'trusted stranger'. Whilst this would be true for both the WS and the TD groups, it is likely that individuals with WS have more experience of unfamiliar professionals interacting with them which could impact upon their behaviour. This further emphasises the need for future work to include an ID comparison group, in order to draw conclusions on syndrome-specific patterns of personal space regulation.

In conclusion, the current study offers a new insight into social distance regulation in young people with WS. These issues become especially concerning in light of the constellation of issues/abilities we associate with the disorder – it is when you look at them all together that the vulnerabilities as so enhanced. When considering the wider social profile associated with WS, these findings feed into

what is already known about social vulnerability in these individuals (Jawaid et al., 2012), and raise significant concerns about their safety when interacting with strangers. Future work needs to expand on this with large sample, cross-syndrome studies, in order to gain more insight into the degree of disorder-specific patterns of personal space regulation. It is also important to consider the developmental trajectory associated with personal space regulation, and the real world implications associated with this. Adults with WS are more likely to be independent and have more encounters with strangers, and as a result could face a greater degree of risk by displaying atypical social distancing.

End of Chapter Discussion

The studies in this Chapter have shown that interpersonal distance regulation is highly atypical in people with WS. Parents report that their sons/daughters are more likely to violate the personal space of others compared to individuals with ASD, and typically developing participants. The stop distance paradigm found that young people with WS maintain a significantly smaller interpersonal distance compared to their typically developing peers when approaching, or being approached by, an unfamiliar person. The congruent findings across these studies, which employed differing methodologies and participant demographics, suggest that personal space regulation is likely to be a pervasive impairment for individuals with WS. This is particularly concerning given the wider social profile associated with WS of hypersociability (Frigerio et al. 2006), lack of stranger danger awareness (Riby et al. 2014), increased approach behaviour (Jones et al. 2000) and reduced intellectual capacity (Searcy et al. 2004). Invading the personal space of strangers is likely to transfer fallacious intentions (Kaitz et al. 2004), and heighten their levels of social vulnerability.

One core issue which emerged across the two studies in this Chapter was the relationship between age and interpersonal distance regulation. The first study in this chapter (age range 4 – 36 years) found no relationship between age and likelihood of violating the personal space of others using the SRS. However, the stop-distance paradigm in the second study (age range 8 – 16 years) showed that older participants were more protective of their own personal space; they asked unfamiliar people to stand further away compared to younger participants. Despite this, both younger and older children were still equally likely to violate

the personal space of others, suggesting i) a differentiation between own personal space and the personal space of others, and ii) that the lack of awareness of other people's personal space transcends age boundaries. It is acknowledged in Chapter 2 that work in this thesis takes a static approach to investigating phenomena of social vulnerability, however, future work in this area would benefit from tracking the developmental trajectory of personal space regulation in WS.

A second core issue which arose in both studies was the impact of anxiety on personal space regulation. The first study in this chapter proposed that anxiety was a theoretically relevant explanation for the findings. Anxiety mediates social behaviour in WS (Kirk et al., 2013), meaning it could be impacting on the ability of individuals with WS to gauge appropriate social behaviour. Interestingly, the second study in this Chapter found that individuals with WS, as well as TD individuals, who were high in anxiety allowed an unfamiliar adult to approach and stand closer to them compared to those who were low in anxiety. Pilot work in Chapter 3 suggested that social behaviour was driving the relationship between social vulnerability and anxiety. Intervention design should therefore consider the link between interpersonal distance regulation, anxiety and levels of social vulnerability.

The third core issue in this Chapter concerns the theoretical explanations proposed to explain the atypical social distancing seen in WS. Parallels were drawn between the patterns of personal space regulation reported in this Chapter and the atypical social distancing observed in patients with bilateral amygdala damage (Kennedy et al., 2009), implicating the amygdala in the personal space

regulation seen in individuals with WS. The similarities in social approach behaviour between individuals with WS and people who have frontal lobe damage (Porter et al., 2007) were also acknowledged, offering that deficits in response inhabitation attributed to frontal lobe dysfunction could better explain the observed behaviour. However, the methodologies used in the current studies do not allow for differentiation between these theoretical explanations. It is also unlikely that these explanations are mutually exclusive, as outlined in Chapter 1. It is hoped that the exploratory work presented within this Chapter will pave the way for exploration of personal space regulation at an individual level, and across several different developmental disorders to capture within and between syndrome variability which could help disentangle these theoretical explanations.

Chapter Six: Examining trust behaviour in young people with Williams syndrome

Chapters 4 and 5 outlined a WS social profile that includes high levels of social approach behaviour, combined with a lack of awareness of stranger danger and inappropriate interpersonal distancing regulation. This means that young people with WS are likely to approach strangers, stand close to them and show a lack of awareness of the dangers the interaction poses (Haas et al., 2010; Doyle, Bellugi, Korenberg & Graham, 2004; Jones et al., 2000; Riby, Kirk, Hanley & Riby, 2014b; Fisher, 2013). Less is known, however, about whether these individuals also show elevated levels of trust behaviour. Martens, Hasinski, Andridge and Cunningham (2012) found that individuals with WS were significantly more likely than TD controls to approach untrustworthy faces. However, Ng, Fillet, DeWitt, Heyman and Bellugi (2015) showed relatively low levels of trust when presented with deceptive behaviour, and failed to show indiscriminate trust as the authors predicted. Further research on trust behaviour in WS would contribute significantly to the profile of social vulnerability that has been built up so far in this thesis; if young people with WS are interacting with unfamiliar people and are also unable to evaluate trust then this could further heighten their level of vulnerability. The current chapter extends the previous work by investigating trust behaviour in WS, compared to typically developing chronologically age matched peers, and typically developing peers matched on receptive vocabulary.

6.1 Introduction

It is widely accepted that trust is a fundamental factor in guiding everyday social interactions (Rotter, 1971). Rousseau et al. (1998) defined trust as, "a psychological state comprising the intention to accept vulnerability based upon positive expectations of the intentions or behavior of another" (p.395). Existing literature has demonstrated that we use attributes such as self-similarity (e.g. Holm & Nystedt, 2005), gender (e.g. Buchan, Croson & Solnick, 2008), experience (e.g. Jones & George, 1998), the ability to infer the mental state of others (Vanderbilt et al., 2011) and facial cues (Ewing et al., 2014a) to help guide our trust behaviour. It has been suggested that children as young as 5 years old have the ability to accurately decipher trustworthiness from facial expressions (Willis & Todorov, 2006), and this trust behaviour matures through development in conjunction with advances in social perspective taking (van den Bos et al., 2011; Evans, Athenstaedt & Krueger, 2013) and executive functioning (Anderson, 20002).

Self-similarity is thought to be a key factor when forming decisions on trustworthiness in adults. Holm and Nystedt (2005) found that 20 year olds and 50 year olds who were developing typically were more likely to trust players who were in the same age cohort as they were during a mail-based trust game. They suggested that this was because players found it easier to infer the mental state of others who were of similar age to them. Indeed, McCabe and colleagues (2001) argued that the ability to infer the mental state of co-players is extremely important in games of reciprocal exchange. However, it is less clear whether this is also true for children who engage in games which involve reciprocal trust.

Many studies have shown that children are more likely to place their trust in adults than in their peers based on judgements of knowledge and authority (e.g. Lampinen & Smith, 1995), however, there is also contradicting evidence showing that children are in fact more likely to trust their peers than adults (e.g. Jaswal & Neely, 2006). This typically occurs in situations where children believe their peers to be more knowledgeable than adults, for example when seeking advice on how to play a child's game (VanderBorght & Jaswal, 2009).

It is unlikely, however, that similar appearances are always used when forming trust judgements. Recent work by Ewing et al. (2014a) used the economic trust game, 'Token Quest', in which participants must make investment decisions when playing against unknown faces (pre-rated as trustworthy or un-trustworthy). They found that both 5 year old and 10 year old participants invested more highly when they had access to written reputation information about their partner, than when they could see their partner's face. In other words, they could override the principles of the self-similarity hypothesis when written character information was available. However, they also noted that the younger children were still motivated to seek out faces to help them with their trust decisions. They were more likely than the older children to pay some of their tokens to access photographs of their opponents, suggesting that physical appearance remains an important source of information for younger children.

Yet, not all individuals find deciding who to trust an easy process. Ewing et al. (2014b) found that children with Autism Spectrum Disorder (ASD; mean age = 9 years old) were able to modulate their trust behaviour in line with reputation

information, but they were equally likely to place their trust in someone deemed to have an untrustworthy face, as they were a trustworthy face. Whereas, the typically developing control group of similar age (mean age = 8.2 years) consistently invested less tokens in untrustworthy faces compared to trustworthy faces. The authors concluded from these findings that children with ASD fail to use facial cues to extract information about trustworthiness.

Indeed, Riby et al. (2014) found that individuals with Williams syndrome (WS; age range 8 – 17 years) also struggle to form trustworthiness judgements. They used a video vignette task, and found that young people with WS had difficulties deciding whether or not they should trust and engage in conversation with a stranger. They suggested that assessments of trust are key to making good social judgements, and difficulties making judgements of trustworthiness could have serious implications for social vulnerability levels. A lack of stranger danger awareness therefore could offer some insight into trust behaviour in WS.

Martens, Hasinski, Andridge and Cunningham (2012) used computer mouse tracking technology to measure approach behaviour in WS to faces that had been pre-rated as trustworthy or untrustworthy. The trajectory of movement in the mouse was used to provide some insight into the process of deciphering trustworthiness and subsequent approach behaviour. It was found that individuals with WS (mean age = 20 years) were significantly more likely than their chronologically age matched peers to approach untrustworthy faces. Further, individuals with WS were more likely to consider approaching untrustworthy faces, even if their final decision was to avoid. The authors suggested that

deciphering trustworthiness from faces is likely to be challenging for individuals with WS because of their impairments in labelling facial expressions (Plesa-Skwerer et al., 2006; Jarvinen-Pasley et al., 2010; Capitão et al., 2011; see Chapter 1 for discussion of the role of the amygdala in identifying facial expressions).

Ng, Fillet, DeWitt, Heyman and Bellugi (2015), using different methodology, found starkly different results. Participants with WS (mean age = 34.8 years) were presented with a series of neutral faces, accompanied by a short scenario. In the transgression lie scenarios, the scenario depicted a liar who had done something wrong and was trying to avoid the negative consequences. In the polite lie scenarios, someone was lying to avoid hurting another person's feelings. Participants were asked some questions about their prosocial judgement of the liar, and they were asked to indicate how willing they would be to socially engage with that person. Contrary to their predictions, the authors found that individuals with WS showed relatively low levels of trust behaviour across the conditions, and rather than showing indiscriminate trust behaviour, they showed some differentiation between transgression lie scenarios and polite lie scenarios, in favour of the polite lie scenarios. However, their distrust extended across the conditions, suggesting that individuals with WS hold a more rigid definition of deception, and consider people who lie to be bad irrespective of their motivations.

From the findings in the thesis so far and the literature to date, we know that individuals with WS show increased approach behaviour, are naïve to danger and show inappropriate social distancing regulation. We also know that in adults with

WS, trust behaviour seems to be atypical (e.g Martens et al., 2012). However, less is known about trust behaviour in young people with WS. Therefore, the current study investigated whether young people with WS were more likely to show increased levels of trust behaviour, compared to their chronological age (CA) matched TD peers and their mental age (MA) matched TD peers (matched on receptive vocabulary skills). Token Quest, a behavioural economic trust game used by Ewing et al. (2014b) was used to assess trust. Participants had to invest tokens in their partners when they could see their silhouette, their face and written hints about their past behaviour in the game. It was hypothesised that, i) in line with their social vulnerability profile, individuals with WS would display higher levels of trust behaviour throughout the game compared to the typically developing control groups, ii) individuals with WS would invest more tokens than their CA and MA typically developing matched peers in the faces round, compared to the silhouettes round, given their interest in socially salient stimuli, iii) like individuals with ASD, participants with WS will be equally likely to trust someone with a trustworthy face, as they would someone with an untrustworthy face, and iv) there would be a self-similarity effect in the silhouettes and faces condition with participants investing more in partners who are children rather than adults, but this would be overridden when reputation information was available in the hints condition, where there will be no significant difference.

6.2 Method

6.2.1 Participants

Participants were 18 young people with a diagnosis of WS (8 – 16 years, M = 11.4, SD = 2.5, 8 males; referred to as the WS CA group) who were recruited from the Williams syndrome Foundation. BPVS-II (Dunn, Dunn, Whetton & Burley, 1997) data was collected for 14 of these individuals (8 – 16 years, M = 12.1, SD = 2.6, 6 males; referred to as the WS MA group). Eighteen TD individuals, matched on age and gender to the WS group were recruited (8 – 16 years, M = 11.3, SD = 2.4, 8 males; referred to as the TD CA group) as well as an additional 14 TD individuals matched on BPVS-II raw scores to the WS group (5 – 10 years, M = 6.7, SD = 1.77, 7 males; referred to as the TD MA group). This was to allow for comparisons between groups based on chronological age and receptive vocabulary age. Informed consent was received from all parents prior to participation, and child assent was also obtained. The study received favourable ethical approval from the local ethics committee.

6.2.2 Stimuli

Stimuli were eight colour photographs of Caucasian faces, four that were deemed to have a trustworthy appearance (two male) and four untrustworthy faces (two male). Half of the images were of children's faces, and the other half were of adult faces. Twenty-five participants were recruited for the pilot phase for selection of stimuli (11 male; mean age 22.6 years, SD 3.41). The images were sourced from the Radboud faces database (Langner et al., 2010), and pre-rated for trustworthiness using a 7-point Likert scale, where 1 = very untrustworthy and 7

= very trustworthy. Faces were chosen for each gender and each age group that were assigned very high or very low trustworthiness scores to ensure a difference in perceived trustworthiness was present (all at $p<0.05$). These scores are shown in Table 6.1. These faces were used in the Token Quest game.

Table 6.1. Pilot rating scores (means and standard deviations) for the faces used in the Token Quest Game

Trustworthiness rating	Adult female	Adult male	Child female	Child male
High	5.72 (1.5)	5.04 (1.1)	5.2 (1.53)	4.8 (1.63)
Low	3.44 (0.98)	2.48 (1.36)	4.2 (2.04)	2.88 (1.56)

Token Quest. Participants took part in an economic trust game, designed to test their willingness to trust others (see Ewing et al., 2014a for a detailed description of the task). They played three rounds, in each they had to decide how many of their tokens to invest (minimum 0, maximum 6). They were told that investing in some partners would be more fruitful than others; however, adopting a risk-free approach and investing no tokens in each round would make it difficult to collect more tokens (Ewing et al., 2014b). The number of tokens invested in each partner was taken as a measure of their trust. The aim of the game was to collect as many tokens as possible.

6.2.3 Procedure

The game began with some simple instructions and the opportunity to ask any questions. The notion of trust was not mentioned during this time. The children took part in two practice trials, which were set for the participant to experience one high token return and one low token return. In each trial, the participants were asked, "How many tokens do you want to give this partner?" Feedback on the number of tokens returned during this practice was given after each trial. The game then began with the first round, in which participants were presented with silhouettes of faces. In the second round they viewed faces which had been pre-judged in the pilot phase to be very trustworthy or very untrustworthy. In the final round, information about their partner's reputation was on screen and read aloud to the child (see Figure 6.1 for examples). Feedback on the number of tokens returned was given at the end of each round. The number of tokens won was dependent on the number of tokens invested, but all investments were rewarded, in line with the rules set by Ewing et al. (2014a).

Figure 6.1. Examples of the stimuli presented in Token Quest.

At the end of the game, participants were asked, "What do you think trust is?" to ensure they understood the concept of trust, which all of the participants were

able to do, with varying levels of complexity. In addition, children were asked to rate the trustworthiness of the eight faces used in Token Quest on a scale of 1 – 7, with 7 being extremely trustworthy. This confirmed that each group could detect a significant difference between trustworthiness of the pre-rated high trustworthy faces.

6.3 Results

6.3.1 Tokens invested overall and in each condition (silhouettes, faces, written hints)

Independent t-tests showed that there was no significant difference in the overall number of tokens invested throughout the game for the WS CA participants (M = 80.5, SD = 22.37), compared to the TD CA control group (M = 67.44, SD = 22.61, t(34) = 1.56, $p = 0.3$). There was no difference between groups in the amount of tokens they invested in the silhouettes condition (t (34) = 1.84, $p = 0.08$), the faces condition (t (34) = 0.99, $p = 0.33$) or the written hints condition (t(34) = 1.4, $p = 0.17$).

There was, however, a significant difference in total tokens invested for the WS MA group (M = 75.78, SD = 24.43) compared to the TD MA group (M = 45.7, SD = 29.99, t(26) = 2.91, $p<0.01$). When receptive vocabulary is the same in both groups, individuals with WS showed higher levels of overall trust behaviour then their TD peers. When looking at the three conditions, there were significant differences between these groups in the silhouettes condition (t (26) = 3.15, $p <0.01$), the faces condition (t (26) = 2.18, $p <0.05$) and the written hints condition

(t(26) = 2.46, p <0.05). Table 6.2 shows that individuals with WS invested more tokens in each of the 3 conditions compared to their BPVS-II matched peers, however, it was the silhouettes condition in which the WS group invested the most tokens.

Table 6.2. Means and standard deviations of the number of tokens invested in each condition

Group	Mean number of tokens invested in silhouettes	Mean number of tokens invested in faces	Mean number of tokens invested in written hints
Williams syndrome MA	28.07 (8.72)	23.79 (12.09)	23.93 (7.72)
Typically developing MA	14.79 (13.18)	14.21 (11.1)	16.71 (7.81)

6.3.2 Tokens invested in trustworthy versus untrustworthy faces

The TD CA group showed significant differences in the face condition between the number of tokens they invested in the trustworthy faces (M = 3.32, SD = 1.37), compared to the untrustworthy faces (M = 2.26, SD = 1.19; t(17) = 3.4, p<0.01). The same was found for the TD MA group (trustworthy: M = 2.11, SD = 1.55; untrustworthy: M = 1.45, SD = 1.42; t(13) = 2.32, p<0.05). Interestingly, this pattern was not evident for the WS group. The WS CA group showed no difference between the number of tokens they invested in trustworthy faces (M = 3.47, SD = 1.72) compared to untrustworthy faces (M = 3, SD = 1.66; t(17) = 1.52, p = 0.15). The WS MA group also showed no significant difference in their

investments based on facial cues of trustworthiness (trustworthy: M = 3.19, SD = 1.77; untrustworthy: M = 2.75, SD = 1.49; t(13) = 1.3, p = 0.21).

6.3.3 Self-similarity

The WS CA group showed no significant difference in the number of tokens they invested in silhouettes of children (M = 14.22, SD = 5.54) compared to silhouettes of adults (M = 14.5, SD = 3.79; t(17) = 0.37, p = 0.71), or the number of tokens they invested in child faces (M = 12.39, SD = 6.57) versus adult faces (M = 13.5, SD = 7.03; t(17) = 0.86, p = 0.4). They did however invest significantly more tokens in adult partners in the written hints condition (M = 15.78, SD = 4.83) compared to child partners in this condition (M = 10.11, SD = 5.64; t(17) = 5.02, p<0.001).

The TD CA group showed a similar pattern. There was no significant difference in the number of tokens they invested in child versus adult silhouettes (child: M = 12.17, SD = 6.54; adult: M = 10.5, SD = 5.31; t (17) = 1.36, p = 0.19) or in child versus adult faces (child: M = 10.72, SD = 4.16; adult: M = 11.61, SD = 5.44; t(17) = 0.95, p = 0.35). But they too invested more tokens in adult partners (M = 15.22, SD = 2.88) compared to child partners (M = 7.22, SD = 3.44) when written hints were available (t(17) = 7.82, p<0.001).

In the TD MA group, the participants invested significantly more tokens in the silhouettes of child partners (M = 9.21, SD = 7.89) compared to the silhouettes of adult partners (M = 5.57, SD = 6.19, t(13) = 2.6, p<0.05). There was no significant difference between the number of tokens they invested in child faces

(M = 6.71, SD = 5.72) and in adult faces (M = 7.5, SD = 5.96, t(13) = 0.81, p = 0.43), but like the other groups, this group invested more tokens when written hints were available about an adult partner (M = 12.07, SD = 3.89) compared to when written hints were available about a child partner (M = 4.64, SD = 4.81; t(13) = 7.03, $p<0.001$).

6.3.4 Ability to differentiate between trustworthy and untrustworthy faces

At the end of the task, all participants were asked to rate the faces on their trustworthiness. Despite their performance in the game, individuals with WS were able to differentiate between trustworthy and untrustworthy faces. Table 6.3 shows the mean trustworthiness ratings given to each group of faces. From this, it is clear that individuals with WS were more favourable in their ratings for trustworthy faces, compared to the TD groups. Interestingly, the WS MA group showed more of a differentiation between rating for trustworthy and untrustworthy faces, compared to the TD MA matched controls.

Table 6.3. Means and standard deviations of trustworthiness ratings given to trustworthy and untrustworthy faces

Group	N	Mean untrustworthy faces	Mean trustworthy faces	P
Williams syndrome CA	18	3.13 (1.21)	5.32 (1.37)	<0.001
Typically developing CA	18	3 (0.94)	4.5 (0.93)	<0.001
Williams syndrome MA	14	2.98 (0.99)	5.18 (1.41)	<0.001
Typically developing MA	14	2.77 (1.48)	3.82 (1.38)	<0.01

6.4 Discussion

Findings from the current study show that there was no difference in the number of tokens invested throughout the game between the WS group and their CA matched peers. There was, however, a difference between the WS group and the TD group matched on receptive vocabulary ability. When receptive vocabulary was the same in both groups, individuals with WS showed higher levels of overall trust behaviour then their TD peers, supporting the first hypothesis. This contradicts the findings of Ng et al. (2015) who found that adults with WS showed relatively high levels of distrust across scenarios. However, in addition to the differing sample demographics and task requirements, the work by Ng et al. (2015) focused on deceit, rather than trust, framing the task in a negative valence

which could affect participants' responses, and could account for the difference in results.

The current findings also show that trust behaviour increases with age in typically developing individuals, suggesting that younger children are more cautious with their trust behaviour. Erikson (1950) suggested that trust behaviour increases as children get older and develop a greater sense of self-identity. Indeed, Bernath and Feshbach (1995) suggested that trust in young children in centered on a concern for one's own interests and concrete rewards during interpersonal exchanges, whereas older children emphasise mutuality. Selman and Selman (1979) found that between the ages of 6 and 12 years old, children begin to see trust as a reciprocal need, as they become more concerned with the thoughts of their social partners. The increase in trust behaviour seen in older children may therefore reflect this understanding of mutuality.

When investigating the second hypothesis, it was found that there was no difference between how many tokens individuals with WS invested in the silhouettes, faces and written hints condition, compared to the CA matched control group. However, when compared to the TD group matched on receptive vocabulary scores, individuals with WS invested significantly more tokens in all three conditions. Contrary to our prediction, they invested the most tokens in the silhouettes condition, rather than the faces condition. It seems that when trust judgements are down to chance (i.e. in the silhouette condition), our participants were willing to make generous trust behaviours. Van den Bos et al. (2010) indicated that young children are more likely to show high levels of trust behavior

when the risks and benefits are low. It may be that in the silhouette condition, where they had no information about their partner and their return was based on chance, they saw this as a low risk, low benefit condition and therefore displayed elevated levels of trust behavior.

The third hypothesis postulated that like individuals with ASD, participants with WS would be equally likely to trust someone with a trustworthy face, as they would someone with an untrustworthy face. It was found that the TD control groups both showed that they could decipher trustworthiness from faces, by investing more tokens in faces that were pre-rated as being trustworthy compared to untrustworthy. However, individuals with WS invested the same amount of tokens in trustworthy and untrustworthy faces, suggesting that they were unable to decipher trustworthiness from faces during the game. This is in line with the findings of Ewing et al. (2014b) who found that children with Autism Spectrum Disorder (ASD; mean age = 9 years old) were equally likely to place their trust in someone deemed to have an untrustworthy face, as they were a trustworthy face. Despite having opposing social profiles, it therefore seems that both developmental disorders are characterised by an inability to decipher trustworthiness from faces, despite being a skill which emerges in typically developing children as young as 5 years old (Willis & Todorov, 2006). It is interesting, however, that participants with WS could successfully rate the faces on trustworthiness in the task at the end of the game, implying that individuals with WS are able to decipher trustworthiness from the faces. This contradicts findings by Martens et al. (2012) who found that adults with WS struggle to decipher trustworthiness from faces, and are therefore more likely to approach

untrustworthy faces. The difference in performance during the game, compared to the trustworthiness ratings provided at the end of the game suggests that other aspects of the task are likely to be affecting children's ability to decipher trustworthiness from faces, for example the cognitive demands of the tasks, or the social encounters in the game context.

Finally, it was predicted that there would be a self-similarity effect in the silhouettes and faces condition with participants investing more in partners who were children rather than adults, but this would be overridden when reputation information was available in the hints condition, where there would be no significant difference. This hypothesis was not supported. It was found that there was no difference between the WS group and the typically developing CA group on the number of tokens they invested in child versus adult partners in the silhouette and faces condition, showing no self-similarity effect. Both groups did however invest more tokens in adult partners than child partners when written hints were available, showing the opposite of the self-similarity effect. The typically developing MA group invested more tokens in child partners than adult partners in the silhouettes condition, there was no difference in investment in child and adult partners in the faces condition, and they invested more in adult partners when hints were available than child partners. This is surprising given the findings of Holm and Nystedt (2005) who emphasised the importance of a shared identity when regulating trust behaviour. It seems that the participants in the current study instead valued the 'knowledgeability' of their partner (related to source credibility cues, including their expertise and age; Lampinen & Smith, 1995). It seems that when both an adult and a child have a positive reputation, all

participants were more trusting of the adult. In the context of the wider social vulnerability profile associated with WS, and the concerns reported by parents in Chapter 4, it would seem that individuals are more likely to trust adult strangers than child strangers. This provides a valuable insight for targeting interventions.

The findings from the current study have offered some interesting insights into trust behaviour in WS, however, the limitations of the study merit consideration. First, the trust behaviour observed in the current findings is situated within the wider context of development. The Token Quest game, as well as offering a measurement of trust, could also be functioning as a delay gratification game. It would therefore be important for future work to investigate how other developmental achievements, such as executive functioning, affect children's performance in the game. Second, it is acknowledged that using an economic investment game, such as Token Quest, to measure trust behaviour could be somewhat arbitrary and may not reflect real-life trust behaviour (Schwieren & Sutter, 2008). The current sample was older than the age of the sample in the work by Ewing et al. (2014a) which focused on 5 and 10 year olds. Indeed the mean age of the TD sample in Ewing et al. (2014b) was 8.2 years. The lack of significant results when performance by the WS group was compared to their typically developing CA matched peers could be because the measure was too arbitrary for the age of the participants. This measure may therefore be more developmentally appropriate for younger children, or individuals with developmental disorders who have a younger MA. Third, several studies have identified a link between trust and anxiety (e.g. Muris, Meesters, van Melick & Zwambag, 2001; Kosfeld et al., 2005; Guastella et al., 2009) which could be

impacting on the results. Anxiety levels are also likely to vary depending on the condition. For example, anxiety (in particular, social anxiety) could play a greater role when evaluating the trustworthiness of another person's face, rather than when looking at a silhouette. As individuals with WS experience high levels of anxiety, future work investigating trust behaviour in this population would benefit from including a measure of anxiety.

Together, the findings from the current study offer further information about trust behaviour in young people with WS. These findings extend the work of Ewing et al. (2014b) by showing that, like individuals with ASD, individuals with WS who played the Token Quest game also had difficulties extracting trustworthiness information from faces. However, the ability of the participants with WS to decipher trustworthiness from faces outside of the game context raises serious questions about the validity of the results. Further, there were also many unexpected results, such as the lack of differences in trust behaviour between individuals with WS and their CA matched peers, which require further investigation. Future studies are now needed to further explore trust behaviour in young people with WS, within the wider context of social vulnerability. Further work in this area holds the potential for early interventions to be explored to promote social competence and reduce vulnerability levels later in life.

Chapter Seven: Parent and Self-Report Ratings on the Perceived Levels of Social Vulnerability of Adults with Williams Syndrome

Chapters 4, 5 and 6 outline a WS social profile that includes high levels of social approach behaviour, combined with a lack of awareness of stranger danger, inappropriate interpersonal distancing regulation and atypical patterns of trust behaviour. This fits with the literature on the WS social phenotype (Jones et al., 2000; Järvinen, Korenberg, & Bellugi, 2013; Martens, Wilson & Reutens, 2008; Doyle, Bellugi, Korenberg & Grahm, 2004) and contributes to the notion that individuals with WS experience high levels of social vulnerability (Jawaid et al., 2012; Fisher et al., 2013). However, it is unclear how individuals with WS see their own levels of vulnerability. This is of interest given their intellectual impairments (e.g. Searcy et al., 2004) that will not only hinder their understanding of social situations (e.g. Riby, Kirk, Hanley & Riby, 2014) but also their understanding of their own abilities and functioning levels (e.g. Mellor & Dagnan, 2005). In order to progress towards intervention development, more needs to be known about the level of self-insight individuals with WS have into their own behaviour. The remainder of the chapters in this thesis use an adult sample (see Chapter 2 for discussion about the methodological choices made).

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7.1 Introduction

Compared to those without disabilities, individuals with intellectual and developmental disabilities (IDD) are at increased risk of experiencing abuse (including verbal, physical and sexual abuse; Wilson & Brewer, 1992) and victimisation (i.e. physical or verbal forms of assault or damage to property; Nettelbeck & Wilson, 2002). They also experience high levels of social vulnerability (Fisher, Moskowitz, & Hodapp, 2012). Social vulnerability is defined as, "the disadvantages faced by an individual while he or she endeavours to survive as a productive member of the society" (Jawaid et al., 2012, p335). Increased social vulnerability has been related to individuals with IDD who: are less aware of risk, appear more vulnerable (e.g., through action or appearance), are afforded more independence from their parents, and have few friends to offer social protection (Fisher et al., 2012).

Recent research has shown that individuals with specific types of developmental disorders (compared to individuals without IDD) are uniquely vulnerable (Fisher, Moskowitz, & Hodapp, 2013). For example, while individuals with autism spectrum disorder (ASD), Williams syndrome (WS), and Down syndrome are all socially vulnerable, their risk factors for social vulnerability are qualitatively different in nature (as measured by the Social Vulnerability Questionnaire, SVQ; Fisher et al., 2012). Specifically, individuals with ASD are socially vulnerable because they have low levels of risk awareness (i.e. protective skills to detect and avoid victimization) and little social protection (i.e. a lack of supportive peer networks); whereas individuals with WS are socially vulnerable because of their high levels of perceived vulnerability (i.e. seeming more vulnerable to other

people) and parental independence (i.e. time spent away from parents/caregivers). Based on these findings, there have been repeated calls for disorder-specific interventions to target the specific correlates of social vulnerability in each disorder (e.g., see Thurman & Fisher, 2015).

In response to the need to better understand the unique social vulnerabilities of individuals with specific developmental disorder conditions, the current study was designed to describe the social vulnerability of adults with WS. WS is a neurodevelopmental disorder caused by a micro-deletion of genes on chromosome 7. Prevalence estimates range between 1 in 7,500 (Stromme, Bjornstad & Ramstad, 2002) to 1 in 20,000 individuals (Korenberg et al., 2003). Individuals with WS typically have mild to moderate levels of intellectual disability (estimated mean IQ of 50 – 60; Searcy et al., 2004) and display a paradoxical cognitive profile of relative strengths and weaknesses in language and spatial perception, respectively (Mervis, Kistler, John & Morris, 2000). Their high level of verbosity when engaged in a social interaction, however, can mask their true level of intellectual functioning, as well as their ability to follow and evaluate complex emotional prosody during a conversation (Mervis et al., 2000).

A key facet of the WS profile is their hypersociability. Individuals with WS are highly social and gregarious, and disinhibited in their social approach behaviour towards both familiar and unfamiliar people (Järvinen, Korenberg, & Bellugi, 2013). Yet, despite their extreme prosocial drive, individuals with WS struggle to maintain peer relationships, and as adults can experience high levels of social isolation (Davies, Udwin & Howlin, 1998). They are also known to experience

elevated levels of anxiety, given their co-existing social difficulties and social motivation (Riby et al., 2014). Overall, then, their lack of inhibition and powerful impulse to engage with other people, coupled with their relative lack of social protection, can result in high levels of social vulnerability in their everyday lives (Jawaid et al., 2012).

Due to the nature of the social-cognitive profile associated with WS, this developmental disorder represents an important population for the study of social vulnerability. Indeed it has previously been claimed that individuals with WS are highly vulnerable and at risk of victimisation (Fisher et al., 2013; Rosner, Hodapp, Fidler, Sagun & Dykens, 2004; Lough, Flynn & Riby, 2015), but less is known about the specific risk factors unique to individuals with WS and how these can be addressed when designing interventions (Thurman & Fisher, 2015).

The Social Vulnerability Questionnaire (SVQ) was designed to identify risk factors for social vulnerability and the types of victimisation experienced by individuals with IDD (e.g. teasing, theft, and abuse; Fisher et al., 2012). Previous research found that individuals with intellectual disability, with few friends, and who displayed more behaviour problems were more socially vulnerable (Fisher et al., 2012). The social vulnerability of adults with WS was then reported to be uniquely related to their vulnerable appearance and parental independence (Fisher et al., 2013).

Three important caveats to the previous findings, however, are that (1) the majority of existing research has relied on parent reports, without accounting for

the individuals' own ratings of their perceived social vulnerability; (2) the previous research did not examine how risk for social vulnerability in Williams syndrome related to specific demographic variables, and (3) there is no qualitative data available about the types of victimisation experienced by individuals with WS. As a result, it remains unclear whether individuals with WS recognise their own levels of vulnerability and thus whether they might feel a need to engage with interventions to learn self-protection and advocacy skills. Additionally, because WS is a heterogeneous condition, it is unclear how individuals' demographic characteristics (e.g., independent functioning, gender) relate to levels of social vulnerability. Finally, until accounts of the types of victimisation experienced by individuals with WS are examined, designing a targeted intervention to address social vulnerability remains challenging.

Recent research has highlighted a clear disparity between parent and self-report measures in adults with WS (Fisher, Mello & Dykens, 2014; Järvinen-Pasley et al., 2010). In two separate studies, no agreement was reported between parents and individuals with WS's rating of social approach behaviour, with parents reporting higher levels of approach behaviour for their son/daughter than was reported by the individual with WS. Fisher, Mello & Dykens (2014) then compared these reports to direct observations of the individuals' behaviours; thus, confirming the accuracy of parent report (and inaccuracy of self-report for adults with WS). Additionally, there was no difference in the inaccuracy of self-reporting based on specific demographic characteristics, including age and gender. While these findings indicate that adults with WS, overall, are not

accurate reporters of their own behaviours, it is not known if they are able to accurately report their own levels of social vulnerability.

Identifying and accurately reporting one's own behaviour and levels of vulnerability has important implications for intervention. For example, inaccurate reporting is problematic when considering intervention efficacy, as an individuals' insight into their own behaviours is an important factor related to the individual's ability to recognize their need for intervention. Like WS, individuals with schizophrenia show lower levels of functioning and self-awareness (Medalia & Lim, 2004). Emmerson, Granholm, Link, McQuaid and Jeste (2009) identified self-insight, specifically insight into the need for treatment, as a key predictor of intervention success for individuals with schizophrenia in a Cognitive-Behavioural Social Skills Training program. Extending these findings with individuals with schizophrenia, understanding how adults with WS view their own social vulnerability is therefore a crucial first step when working toward intervention design.

The current study therefore aimed to address this void in the literature by comparing parent and self-reported levels of social vulnerability using the SVQ and examining social vulnerability in adults with WS related to specific demographic characteristics. Further, to better understand the types of victimisation commonly experienced by adults with WS, qualitative data on parent reported examples of victimisation was also collected. Three research questions were examined. First, compared to parent report scores, do individuals with WS report significantly lower levels of social vulnerability? Second, how do

age, gender, functional ability, intellectual ability, and living arrangement and employment status relate to SVQ scores, as reported by adults with WS and by their parents? Third, in the past year, what types of victimisation have adults with WS experienced?

7.2 Method

7.2.1 Participants

Overall, 102 adults with WS (55 males, 47 females) and 102 parents (95 mothers, 7 fathers) participated. The majority of participants (n = 148; 74 parent/adult with WS pairs) were recruited from the Williams Syndrome Association (WSA) listserv. Specifically, all WSA members who fit the inclusion criteria (had a child with WS aged 18 or older) received an email inviting them to contact the research team for a link to the survey if they were interested in participating. The email described the study as a research project examining the social functioning of adults with WS. A subsample of participants (n = 28) was recruited from a residential summer camp program for adults with WS. On average, adults with WS were 27.83 years (*SD* = 7.96). The mean age of participants did not significantly differ based on recruitment method. Full scale IQ data were available for the subsample of participants recruited through the summer camp, with an average full-scale IQ of 69.11 (SD = 15.28; KBIT-2, Kaufman & Kaufman, 2004). Parents recruited through the WSA completed the Activities of Daily Living (Seltzer & Li, 1996) as a proxy for level of functioning, with an average total score of 45.82 (7.66), range 25-58.

7.2.2 Measures

Demographics. Demographic information was collected from the parents about the adult with WS. This questionnaire asked for information such as age, gender, living situation, employment status, and daily activities (see Table 7.1).

Table 7.1. Demographic information of individuals with WS based on parent report

	M (SD)	%
Age	27.83 (7.96)	
Gender: Male (Female)		53.9 (46.1)
Living arrangement		
Lives with parent/guardian		85.7
Supported living		7.2
Own apartment		7.2
Daily Activities		
Employed*		57.1
Attends school		7.1
Attends day programme		25
In job training		3.6
Volunteers		46.4

*For those who were in employment, the number of hours spent at work ranged from 1 – 40 hours per week, with an average of 17 hours per week.

Social Vulnerability Questionnaire (SVQ; Fisher et al., 2012). The SVQ includes 30 items that parents rate on a four point scale (1 = *not true or never* to 4 = *very*

true or always) about their child. The 30 items load on to six subscales: emotional bullying (e.g. "Gets picked on by others"), risk awareness (e.g. "Recognizes potentially dangerous situations"), social protection (e.g. "Is considered a part of a social peer group"), perceived vulnerability (e.g. "Others consider him/her to look different from same aged peers"), parental independence (e.g. "Parents are likely to leave alone for an extended period of time"), and credulity (e.g. "He/she is likely to believe a claim when there is evidence it should not be believed"; see Fisher et al. 2012 for further description of the SVQ). High scores are indicative of higher levels of social vulnerability. The maximum total score that can be obtained is 120. Reliability statistics for each subscale of the SVQ ranged from .59 to .89 for the parent version and .39 to .84 for the self-report version. The vulnerable appearance subscale for the self-report version had alphas well below acceptable levels (.39) and was therefore considered unreliable and removed from the analyses (Tavakol & Dennick, 2011). The majority of the remaining alphas were between the recommended levels of .70 and .90, indicating the SVQ was a reliable measure for both parent and self-report. The questionnaire also included an open-ended question – "Can you give us an example of a time within the past year that your child/the individual has been taken advantage of?" Individuals with WS were also asked to complete the SVQ, which was adapted to use simplified language and did not include the final open-ended question. The number of items and the rating scale remained the same.

Activities of Daily Living (ADL; Seltzer & Li, 1996). The ADL measures the functional abilities of individuals with IDD through 15 items rated on a 5-point scale (1 = *not at all* to 5 = *very well*). Sample items include the degree to which

the individual is able to walk, read, participate in leisure activities, and work (Cronbach's α = .83). Variables are summed into a single, cumulative score ranging from 15 to 75, with higher scores indicating greater functional independence. This scale has been reliably used in previous research to provide an estimate of functional abilities of individuals with IDD (Burke, Taylor, Urbano & Hodapp, 2012; Seltzer & Li, 1996).

Kaufman Brief Intelligence Test- 2nd Edition (K-BIT-2; Kaufman & Kaufman, 2004). The KBIT-2 is a brief assessment of cognitive functioning, usually administered in 20 minutes or less. The assessment provides a Composite IQ, Verbal IQ, and Nonverbal IQ. Standard scores are normally distributed with a general population mean of 100 and standard deviation of 15 (range from 40-160). Verbal IQ is derived from two subtests measuring receptive vocabulary, general knowledge, and verbal reasoning (Verbal Knowledge; Riddles), and Nonverbal IQ is derived from a subtest assessing nonverbal reasoning (Matrices). The KBIT-2 has been used as a reliable estimate of IQ for individuals with Williams syndrome in previous studies (e.g., Fisher, Lense, & Dykens, *in press*; Mervis, Kistler, John, & Morris, 2012).

7.2.3 Procedure

To ensure that adults with WS would be able to understand and complete the adapted version of the SVQ, a subsample of participants was recruited from a residential summer camp program to pilot the questionnaire. For these participants, parents of individuals with WS were contacted about their interest in participating in research as a part of the camp. Interested parents were then sent a

consent form and an assent form for the participant with WS to sign. Once the signed forms were received by the research team, parents were sent the link to questionnaires to complete online. The rate of completion for parents was 93.6%. The individuals with WS subsequently completed adapted forms of the measures with the support of one of the authors while at the residential camp. The completion rate with this group was 96.8%; one individual was unable to complete the questionnaire. After observing that the remaining adults with WS were able to answer the questions without assistance (e.g., did not ask for clarification), we determined the questionnaire could be placed online and a second, larger sample could be recruited.

For the second group, interested parents from the WSA listserv were emailed a link to the parent questionnaires. The rate of completion was 92.1%. Once parents completed their questionnaires, they were directed to a page that contained the link to the self-report questionnaires. Parents were asked to save the link and to open it when the individual with WS was present and ready to complete their portion of the study. Parents were asked to support the individual with WS while completing the questionnaire (e.g., reading questions aloud, using the mouse, clicking the correct option), but to not provide any influence or guidance in answering the questions. The rate of completion was 97.4%. For the purposes of this study, only those individuals for whom we received both parent and self-report responses were included in the analyses. This research received ethical approval from the local Institutional Review Boards. All participants received a gift card for their participation.

7.2.4 Data analysis

To confirm the reliability of the parent and self-report versions of the SVQ, we conducted Cronbach's alphas for each subscale. We then conducted preliminary t-test analyses to compare differences in parent and self-report scores on the SVQ based on recruitment method. Next, to examine whether there was a difference between parent reported levels of social vulnerability and self-reported levels of social vulnerability, we conducted a MANCOVA using recruitment method as a covariate, and then conducted follow-up ANCOVA analyses to examine differences on each factor within the SVQ. To examine the relation between scores on the SVQ and specific demographic variables, we used partial correlational analyses (controlling for recruitment method), MANCOVAs and ANCOVAs, using recruitment method as a covariate.

Finally, the open-ended questions were coded by two coders to identify the types of victimisation experienced by adults with WS. There was a high level of inter-rater reliability (percentage agreement 100%). Fisher et al. (2013) coded responses to these open ended questions into three types of victimisation: teasing and persuasion; money or theft; and physical or sexual abuse These categories were used to inform the first round of the coding process in the current study, using a deductive approach. Importantly, however, in a review of the victimisation literature, Nettlebeck and Wilson (2002) noted that the types of victimisation experienced by individuals with an IDD are as diverse as those experienced by individuals without an IDD. Therefore, in the second round of coding, the previous framework was removed, and an inductive approach was employed, where themes were generated from the data using unrestricted coding.

The themes generated from both methods were synthesised and refined, and the final themes were generated.

7.3 Results

7.3.1 Preliminary Analyses

We first examined whether parent and self-report total and subscale scores on the SVQ differed by recruitment method. While few parent scores differed, several participant scores differed significantly. Specifically, although total scores on the SVQ did not differ for parents who were recruited through the camp versus those who were recruited through the WSA listserv (total score = 70.23 (8.80) vs. 67.32 (8.05), respectively, t (100) = -1.52, *ns*), camp parents rated their children higher on parental independence than did WSA-recruited parents (7.67 (2.14) vs. 6.22 (2.52), respectively, t (100) = -2.72, $p < .01$).

Next, adults with WS who completed the SVQ online rated themselves as significantly more socially vulnerable than did those who completed the SVQ at the summer camp (total score = 63.59 (7.90) vs. 59.50 (8.57), respectively, t (100) = 2.28, $p < .05$). These differences were specifically related to risk awareness (15.75 (2.97) vs. 13.46 (2.55), t (100) = 3.61, $p < .01$)), social protection (9.38 (2.59) vs. 7.46 (3.16), t (100) = 3.13, $p< .01$), and credulity (11.48 (2.61) vs. 9.36 (2.90), t (100) = 3.56, $p< .01$). Adults from the summer camp, on the other hand, scored higher on parental independence than did adults from the WSA listserv (7.82 (2.26) vs. 6.41 (2.55), respectively, t (100) = -2.58,

$p < .05$). Because of these differences, the remaining analyses were conducted using recruitment site as a covariate.

7.3.2 Differences in Parent and Self-Report on the SVQ

To address our first research question, we examined whether parents and adults with WS provided significantly different responses to the SVQ. Indeed, parents reported their child to be more socially vulnerable (mean = 69.43, SD = 8.66, range = 49 - 90) than the individuals with WS reported themselves (mean = 62.47, SD = 8.26, range = 46 - 80; $F(1, 201) = 35.57$, $p < .01$, $\eta^2 = .15$).

We then compared differences between the respondents on each of the subscales of the SVQ. Table 7.2 shows that there was a significant difference in four out of the five areas of social vulnerability (all p's < 0.01), with parents and individuals with WS only rating the level of parental independence as similar. Compared to self-report, the parents reported higher levels of social vulnerability on the risk awareness, social protection, and credulity subscales. Adults with WS, however, rated their levels of emotional abuse (e.g. being picked on by others, peers making fun of him/her) higher than parents rated emotional abuse.

Table 7.2. SVQ subscale scores

	Parent		Individual		F	p	η^2
	Mean	SD	Mean	SD			
Emotional abuse (maximum score 20)	8.46	2.97	10.25	3.68	14.58	0.00	.07
Risk awareness (maximum score 36)	17.81	3.92	15.12	3.02	31.81	0.00	.14
Social protection (maximum score 16)	10.42	2.98	8.85	2.87	15.41	0.00	.07
Parental independence (maximum score 12)	6.62	2.50	6.79	2.54	.27	0.61	
Credulity (maximum score 20)	13.76	2.96	10.90	2.83	51.65	0.00	.20

7.3.3 Relation of SVQ Scores to Demographic Variables

To address the second research question, we examined whether social vulnerability of individuals with WS was related to specific demographic characteristics, including age, gender, functional abilities, intellectual functioning, and living arrangement and employment status.

Age. Parent reports indicate that there was no significant correlation between age and total SVQ scores (r = -0.06, *ns*). However, increasing age was significantly related to higher scores on the parental independence subscale (r = 0.42, $p<.01$) and increasing age was significantly related to decreasing scores on the credulity subscale (r = -0.27, $p<.01$). Similar to parent report, self-report total scores on the SVQ were not significantly related to age (r = 0.02), but older age was related to higher scores on the parental independence subscale (r = 0.30, $p<.01$). Additionally for self-report, increasing age was related to a decrease in scores on the emotional abuse subscale (r = -0.21, $p<.05$).

Gender. Parents reported higher levels of social vulnerability for males (M = 70.89, SD = 7.72) compared to females (M = 67.72, SD = 9.44) on the total SVQ score (F (1,102) = 6.23, p = 0.01, η^2 = .05); however, there was no difference for gender on self-report total SVQ score (males: M = 62.44, SD = 7.58; females: M = 62.50, SD = 9.06; F (1,102) = 0.48, *ns*). Examining the SVQ subscales, parents rated males (compared to females) to be more socially vulnerable on the risk awareness subscale (18.60 (4.00) vs. 16.89 (3.65), respectively, F (1, 101) = 8.89, $p < .01$, η^2 = .08) and on the parental independence subscale (7.49 (2.59) vs. 5.60 (1.99), respectively, F (1, 101) = 11.78, $p < .01$, η^2 = .11). Females with WS rated

themselves higher on self-reports of the emotional abuse subscale (10.93 (3.91)) compared to males with WS (9.67 (3.39), F (1, 101) = 4.52, $p < .05$, $\eta^2 = 0.04$).

Functional Abilities. Total ADL score (n = 74) was significantly negatively related to the total score on the parent version of the SVQ (r = -0.42, p<.01), and to all of the parent-report SVQ subscales (r's range from -.29 [credulous] to .51 [parental independence], all p's < .01), with the exception of the emotional abuse subscale (r = -0.20, *ns*). These correlations indicate that higher levels of functional abilities are related to lower levels of social vulnerability. Similarly, total ADL score was also significantly negatively related to the total score on the self-report version of the SVQ (r = -0.49, p< .01), and to all of the self-report SVQ subscales (r's range from -.30 [emotional abuse] to -.51 [social protection], all p's < .01), with the exception of the credulity subscale (r = -0.22, *ns*).

Intellectual Abilities. Interestingly, when examining the subsample of those who had KBIT IQ scores (n = 28), there was no significant correlation between IQ and total SVQ score, as reported by the parent (r = 0.11, *ns*) or by the adult with WS (r = -0.04, *ns*), nor were any subscales significantly correlated with IQ.

Living Arrangement and Employment Status. Finally, there was no significant difference in parent report total SVQ scores between individuals who lived at home versus those who lived away from home F (1,101) = 0.01, *ns*, nor was there a difference for self-report scores (F (1,101) = 3.73, *ns*). Neither parent report total SVQ scores nor self-report total SVQ scores were related to differences

between those who worked and those who did not work F (1,101) = 1.71, *ns* and F (1, 101) = 3.00, *ns*, respectively.

7.3.4 Experiences of victimisation

Finally, to determine whether adults with WS did indeed experience victimisation and to better understand the types of victimisation most often experienced by adults with WS, we asked parents to give us an example of a time when their child had been taken advantage of in the past year. The majority (75%) of parents provided an example of victimisation. The most common response to this open-ended question referenced instances relating to money/theft (n = 43). Examples given included:

> "He has become friends with customers of his store. One of these individuals convinced him to give her money because she was going to lose her house. He used his debit card and gave her $300."
>
> "She buys food for friends because she is always generous. She has no concept of money and is just so glad to have a friend."
>
> "I also found out that he allowed a stranger to teach him how to use an ATM to withdraw money with his debit card."

Other parents noted examples of teasing or persuasion (n = 10):

> "One of his co-workers would bother him while being transported to his job site."
>
> "One of his friends was over to our house and I was not at home. His friend talked him into letting him drink an alcoholic beverage."

Instances of sexual abuse and grooming also featured in parents' accounts as examples of victimisation that their son/daughter had experienced (n = 9):

> "Bus driver asked her to teach him how to slow dance when they were alone on the bus, which she did."
>
> "Chatted with someone on the internet and she believed they wanted to be her boyfriend and she wanted a boyfriend so badly she was willing to meet them without supervision."
>
> "She met a man … he ended up leading her into a forest trail system … he tried to touch her inappropriately."
>
> "Shared inappropriate photos because was told she was loved by a stranger on the internet."

Some adults with WS had experienced physical violence, or had been threatened with physical violence (n = 3):

> "She was assaulted in the local shopping centre toilets. A drugged woman was hiding in the cubicle and came out to grab her bag…my daughter was unable to defend herself properly."

Finally, 11 parents recounted other examples of victimisation, which included being let down by other family members, letting strangers into their house, and giving out their phone number to strangers.

7.4 Discussion

The findings from the current study offer a novel insight into how adults with WS view their own social vulnerability and how interventions can be developed to

address the unique social vulnerabilities of this population. This study extends the literature by comparing parent report to self-report to determine whether adults with WS are aware of their own levels of social vulnerability. Additionally, we describe important factors that should be considered when designing interventions for this vulnerable group.

First, because interventions are more effective when the recipients acknowledge their need for training, we sought to determine whether adults with WS are aware of their own levels of social vulnerability. Indeed, our findings showed that adults with WS perceive their overall social vulnerability to be lower than that reported by their parents. Specifically, adults with WS rated themselves lower than their parents did in four out of the five subscales; adults with WS and their parents were only in agreement about their level of parental independence. On the majority of the remaining subscales (risk awareness, social protection, and credulity) the individuals with WS reported lower social vulnerability levels than their parents. However, this was not the case for the subscale of emotional abuse, where the WS group reported higher social vulnerability levels than their parents.

Because the actual behaviours of individuals with WS were not observed, it is difficult to truly determine if parents were a more accurate reporter of social vulnerability than the individual with WS. Previous research, however, has highlighted that adults with WS are in fact unreliable reporters of their own social approach behaviours (Fisher et al., 2014; Järvinen-Pasley et al., 2010). Given the similarities of the behaviours being assessed in this study to those queried in the previous research, we lean toward the conclusion that the individuals with WS in

this sample were less aware of their own social vulnerability. This lack of insight points to the need to first address self-awareness with adults with WS before teaching them self-protection. It also emphasises the importance of parent-report, and how it should continue to be considered an important piece of information when assessing individuals with WS.

Addressing the second research question, to ensure interventions are designed to address the characteristics that are most likely related to increased social vulnerability for individuals with WS, we examined the relation between specific demographic characteristics and levels of social vulnerability. Overall, the individual's age, IQ, and living situations were not related to parent or self-report total social vulnerability scores. These findings indicate that individuals with WS may be socially vulnerable throughout their lifespan and regardless of the severity of their intellectual disability, highlighting the importance of providing intervention to all individuals with WS. We also found that employment status was not related to parent or self-report total social vulnerability scores, suggesting that those who were employed experienced similar levels of vulnerability to those who were unemployed. However, for those participants who were in employment, the number of hours worked ranged from 1 to 40 hours per week. It is therefore likely that individuals within this group had highly heterogeneous experiences of employment, meaning caution is advised when interpreting these results.

Regarding gender, parents reported males to be more socially vulnerable than females, but males did not report themselves to be more socially vulnerable. Such

increased social vulnerability for males was more specifically related to risk awareness and parental independence; parents indicated that males with WS (compared to females) are less able to detect risky situations but they are given more independence from their parents. This is coupled with the finding that males with WS did not rate themselves as more socially vulnerable, highlighting that males may lack insight even beyond those with WS in general. This pattern is reversed for those without disabilities. Females are typically considered to experience higher levels of vulnerability than males (e.g Mitchell, Jones, Finkelhor & Wolak, 2007). However, it is widely reported that victimization against males is underrepresented due to social stigmas attached to being the victim of a crime (O'Leary & Barber 2008). Therefore, rather than females without disabilities being more vulnerable than males, it may be that they are simply more likely to report instances of victimization. The lack of gender differences in the self-report of vulnerability in the current study could suggest that males with WS do not experience the same level of stigma associated with vulnerability and victimization. Alternatively, like individuals without disabilities, they too may be more likely to underreport, hence the discrepancy between parent and self-reported levels of vulnerability in males. Nevertheless, the parent reported differences in vulnerability between males and females require further exploration in any intervention design, and perhaps individualized, gender-specific trainings should be considered.

While IQ was not related to scores on the SVQ, the individual's level of functioning was related to the SVQ. Both parents and individuals with WS rated individuals with greater functional independence to be less socially vulnerable.

This finding suggests that the individual's ability to perform daily living tasks independently serves as a greater protective factor than an individual's intellectual ability. Similar to previous calls for action, this finding highlights the importance of teaching functional independence from an early age, so that adults with WS might be better prepared to be on their own and to protect themselves (Thurman & Fisher, 2015). Additionally, similar to the gender differences, this finding again highlights the importance of individualising interventions for individuals with WS who display specific demographic characteristics. It will be important to teach additional safety skills to individuals with WS who are not as functionally independent.

Finally, while there was no relation between age and social vulnerability overall, older age was indeed related to the parental independence subscale on both the parent and self-report ratings. These findings are similar to previous research (Fisher et al., 2013) and indicate that adults with WS are afforded more independence as they age. This finding raises concern, however, because even older adults with WS are not often aware of the risks they may encounter when on their own, which could then lead to instances of victimisation. Again, this highlights the importance of including individuals of any age in interventions.

Our third research question found that the majority of the individuals with WS in this sample experienced victimisation and parents reported several different forms of victimisation. The most common form of victimisation was related to problems with money. Financial victimisation has been identified as a commonly experienced form of victimisation for individuals with IDD (Fisher et al., 2012;

Greenspan, Loughlin & Black, 2001; Nettelbeck & Wilson, 2001), with financial abuse reported to encompass 14.6% of referrals to Adult Protective Services (Mansell, Beadle-Brown, Cambridge, Milne & Whelton., 2009). Unfortunately, few interventions have directly addressed ways to recognise and avoid financial victimisation. Future interventions for adults with WS should consider the specific forms of victimisation experienced by adults with WS so that they can learn to recognise and address the situations they are most likely to experience.

7.4.1 Implications for Intervention

Evidence from the clinical literature frequently advocates adopting a multi-informant approach in order to increase intervention efficacy (for an example, see De Los Reyes, Thomas, Goodman & Kundey, 2013; De Los Reyes et al., 2015). Indeed, based on our current findings it would seem that individuals with WS lack insight into their own behaviours that could lead to victimisation.

Such findings highlight the need for interventions that address self-awareness. For example, if individuals with WS do not see themselves as easy to take advantage of (e.g., many report that they do not talk to strangers and cannot easily be convinced to give away money to others, yet parents report these behaviours as a problem), then interventions addressing these concerns may be ineffective. Rather, interventions should first address ways to teach individuals with WS to evaluate potentially vulnerable situations and to then teach them how to handle such situations. It is this ability to recognise risky behaviour which could be key to successful social safety interventions.

7.4.2 Implications for Future Research

Parental scores on the SVQ are in line with the work by Fisher et al. (2013), which offered comparisons between the SVQ scores of adults with WS, ASD and Down syndrome. For example, in the work carried out by Fisher and colleagues (2013), scores on the domains of risk awareness, perceived vulnerability, parental independence and credulity were 16.38, 12.81, 7.16 and 13.72 respectively. In the current study, scores on these domains were 16.75, 12.18, 7.68 and 13.46. Whilst these cross-study comparisons offer insight into the validity of the current findings, future work would benefit from comparison groups (e.g. other developmental disabilities and typically developing individuals) to look at the syndrome-specific pattern of social vulnerability. This will allow us to really understand the areas of most extreme social vulnerability within WS specifically, and will aid the development of tailored intervention approaches.

A clear strength of this work is it has highlighted the importance of self-report, and the need for multi-informant studies. However, there remain limitations which should be addressed. First, the current study cannot offer insights into which informant can most accurately report social vulnerability levels. This preliminary study focused on examining differences between respondents, but it is hoped that these findings can be used as a basis for behavioural observation studies to provide more insight into this issue. Second, further work is needed on the SVQ with a normative sample to generate severity markers. Without this, we are unable to draw inferences about the severity of the social vulnerability reported. Third, although no relationship was found between IQ and social vulnerability levels, IQ data was only collected for a sub-sample (n = 28). This

means that the link between IQ and social vulnerability remains unknown for the majority of the sample. Fourth, the varied methods of data collection (e.g. in person versus on the Internet) could have impacted on the results. For example, although parents of individuals who completed the measure online were asked not to influence or provide any guidance in answering the questions, we cannot fully ascertain the effect of parents on the online self-report data. This would be important to manipulate in future work comparing parent and self-report. Fifth, the measure was deemed appropriate and accessible based on pilot testing, in which participants were able to complete the questionnaire without asking for clarification. However, Llewellyn and Northway (2008) highlighted that self-advocacy can be challenging for individuals with IDD, Future research should therefore seek to ask participants to summarize their understanding of the measure, to further ensure readability. Finally, the demographic data shows that this is a group that is highly likely to be dependent upon others into adulthood. This high level of dependence could indeed be viewed as a protective factor, as they are likely to have less freedom to get into vulnerable positions. Equally, this could be a risk factor due to their reliance upon others and inability to cope independently. Further exploration of the relationship between parental independence and vulnerability is warranted.

In conclusion, the findings from the current study highlight the need to teach self-awareness as part of a multi-informant approach to intervention design. Previous research, (e.g. Fisher et al., 2014) has identified parental reports as being more representative of behaviour, compared to self-report. The use of parent reports when investigating social vulnerability is therefore crucial in understanding the

risks faced by individuals with WS. However, the inclusion of self-report captures the importance of self-awareness, which has the potential to be overlooked in training programs solely based on parent report. Indeed, the authors are unaware of any studies to date which have looked at the impact of self-awareness on intervention design and success in individuals with IDD. Future studies should consider adopting a multi-informant approach to interventions designed to target social vulnerability, and consider the importance of syndrome-specific patterns of social vulnerability.

Chapter Eight: Mapping real-world to online vulnerability in young people with developmental disorders: Illustrations from Autism and Williams syndrome

Moving on from the real world interactions which have been the focus of the thesis up to this point, the following three chapters address social vulnerability in the online world. The internet has become increasingly accessible over the past few decades, and offers a wealth of new opportunities for individuals with intellectual and developmental disabilities (IDD), including an alternative platform to engage in social interactions. However, the internet also poses significant risks, for those with and without an intellectual disability. This chapter reviews how the characteristics which contribute to making individuals with WS and ASD socially vulnerable in the real world could also be making them vulnerable to victimisation online. Characteristics which have played a key role in the thesis so far, such as anxiety (as explored in Chapter 3), disinhibition (explored in Chapter 4) and trust (explored in Chapter 6), are reviewed to examine whether they could be contributing to online vulnerability.

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8.1 Introduction

The vulnerability and safety of children and young people is a prominent issue for the parents, media and government of today. High profile cases of child victimisation and abuse have nurtured fear within society about the safety and resilience of young people (Mitchell et al., 2011). Concern regarding vulnerability and risk has recently been extended to internet conduct, with cyber-bullying, trolls and online grooming posing a new type of risk to young people (Livingstone et al., 2011). The 'EU Kids Online' survey, which interviewed twenty-five thousand children and their parents throughout Europe (Livingstone et al., 2011), found that young people with an intellectual or physical disability face an elevated level of risk compared to others. Such findings reinforce the recommendations of the UK 'Children Go Online' survey (Livingstone & Bober, 2005), which called for in-depth, targeted research on the internet conduct of vulnerable minority groups, specifically highlighting ethnic minorities and individuals with disabilities as priorities for future research. Yet, despite these findings and recommendations, there has been little research to advance our knowledge about the vulnerability of these groups in an online environment (Whittle et al., 2013a). This current paper aims to use what is known about the social profiles of individuals with developmental disorders (namely Autism Spectrum Disorder and Williams syndrome), as well as our knowledge of offline social vulnerability, to inform predictions about the vulnerability of these cohorts online, in order to stimulate the critical research and debate for action in this area.

8.1.1 Autism Spectrum Disorder & Williams syndrome

While vulnerability and resilience are relevant to all young people, Autism Spectrum Disorders (ASD) and Williams syndrome (WS) are developmental disorders that offer case illustrations showing when the interaction between social characteristics and online/offline vulnerability are especially pertinent. As detailed in subsequent sections, these two disorders have been chosen as they are characterised by atypicalities of social behaviour, but the nature of those atypicalities varies across the two disorders. Importantly, the impact of the atypicality has implications for everyday social functioning in both groups. The reasons for postulating that offline – online social vulnerability is critical in these groups are as follows:

8.1.2 Autism Spectrum Disorders

ASDs are a set of lifelong neuro-developmental disorders, typically characterised by a triad of impairments in the domains of communication, social interaction and restricted, repetitive behaviours (APA, 2000). Thus, many individuals who are functioning on the autism spectrum have difficulty interpreting social situations and responding appropriately to them (Smith et al., 2010). These difficulties have implications for face-to-face interaction; for example, individuals may miss communicative cues (Rump et al., 2009). As a consequence of the array of social difficulties and possible intellectual impairments, individuals who have autism represent a highly socially-vulnerable cohort in their real (offline) world (Howlin et al., 2004).

It remains unclear how the social profile associated with functioning on the autism spectrum and resulting vulnerability might manifest online. While on-line and off-line worlds are becoming more integrated, it may be that environments that rely more on virtual interactions (on-line environments) make different interactional demands to environments with less, or no virtual interactions; but what effect do these differing environments have for individuals who are functioning on the autism spectrum? Online platform offers individuals with autism an alternative method of engaging in social interaction, which may be appealing due to the diminished levels of social presence, reciprocity and social anxiety, whilst also drawing on frequent strengths and interests in screen-based technology (Mazurek, 2013). Conversely, impairments associated with ASD could lead to inappropriate interactions styles online, as they do offline (Happe, 1999), as social acuity is still required in conversations online as it is offline. A dialogue of the similarities and differences that suggested for the offline - online interactional environments of individuals who are developing typically will be provided to inform discussion of the proposed online vulnerability faced by individuals with ASD.

8.1.3 Williams syndrome

Williams syndrome (WS) is a rare genetic neuro-developmental disorder, caused by the microdeletion of 25 – 28 genes on chromosome 7q11.23 (Hiller et al., 2003). Characteristics of WS include mild – moderate intellectual impairment (estimated mean IQ of 50-60; Searcy et al., 2004) and a distinct social profile, notably, a hyper-sociable behavioural phenotype (Jarvinen et al., 2013). This latter characteristic refers to an exaggerated desire to interact with others – both

familiar and unfamiliar people (Jones et al., 2000; Jawaid et al., 2012). During such interaction, it has been suggested that some individuals with WS display atypicalities of social communication and behaviour, for example prolonged gaze behaviour to faces, especially the eye region (Riby & Hancock 2008). Their verbosity masks their level of intellectual impairment, allowing an easy flow of conversation but at a superficial level (Mervis et al., 2000). This relative proficiency hides an array of subtle deficits and atypicalities of evaluating complex emotional prosody during interactions (Pinheiro et al., 2011). These characteristics can occur in parallel with aspects of psychopathology, such as highly prevalent anxiety as the main mental health concern (Stinton et al., 2010). Indeed individuals with WS who show the highest anxiety also show more severe problems with everyday social behaviours (Riby et al., 2013).

As with ASD, it remains unclear how this social profile may be exhibited in an online environment, and the consequent level of risk that these traits pose when interacting online. What is known is that individuals with WS are considered highly vulnerable in the offline environment (Jawaid et al., 2012). Based on what we know about the level of transference in vulnerable typically-developing young people (Whittle et al., 2013a), there is reason to postulate that this vulnerability could be matched if not intensified during online interactions for individuals with WS. In order to pursue this line of argument, we must first acknowledge what it means to be vulnerable in the offline environment.

8.1.4 Social vulnerability

Due to the substantial social atypicalities in the groups discussed above, as well as our interest in the social online conduct of these groups, it is *social* vulnerability that is of particular interest in this context. Social vulnerability specifically refers to, "the disadvantages faced by an individual while he or she endeavours to survive as a productive member of the society" (Jawaid et al., 2012 p335), encompassing bullying (Fox & Boulton, 2005), abuse (Sidebotham, 2013), victimisation (Fisher et al., 2013), and social exclusion (Hofvander et al. 2009). When considering social vulnerability as an outcome, the relation between the contribution of 'risk' and 'protective' factors must be considered (Whittle et al., 2013b). The existing body of literature has established a common understanding that risk factors constitute a characteristic, a situation or a combination of both that result in a heightened probability that an individual will experience harm (Masten & Powell, 2003). Conversely, the consensus of opinion on protective factors revolves around definitions that acknowledge their protective influence in minimising the impact incurred by the risk (Blum et al., 2002). Rather than a single risk factor, young people tend to hold a socially vulnerable position due to the interaction between several risk factors, which occur in lieu of sufficient protective factors (Dixon et al., 2009). It is important to reflect on how this information can help make predictions about online vulnerability, before considering how using the profiles of ASD and WS as case examples, we can highlight risk factors that elevate levels of online vulnerability.

8.2 Interaction between online and offline vulnerability

With the rise of interactive technology, most notably the internet, the social environment in which young people interact has expanded to encompass a virtual world of communication. The portability of devices that allow access to the internet, such as iPads, mobile phones and Kindles, permit constant communication and accessibility. It is estimated that over 80% of 5–15 year olds have access to the internet at home (European Commission, 2008), with as many as 31% having access in their bedroom and 66% having a mobile phone by their tenth birthday (Ofcom, 2010). This has led to a debate on the extent to which there is a distinction between one's online and offline existence, and in turn the level of transference between offline and online existence (Child Exploitation and Online Protection Centre, 2010).

It has been argued that the characteristics of those individuals considered to be most vulnerable offline can be used to make predictions regarding their vulnerability online (Whittle et al., 2013b). Numerous studies have identified qualities such as immaturity (Olson, et al., 2007), agreeableness (Talbot et al., 2003) and mental health problems (Mitchell et al., 2007) as precursors to offline social vulnerability. It has been suggested that these offline characteristics shape how an individual presents him- or herself to others online (CEOP, 2010). For example, those with fewer and weaker social relationships in the offline world may seek to form relationships online to compensate for this void (Mesch, 2001). As suggested above, the online and offline platforms could be so complexly intertwined that they cannot be viewed in isolation.

Similarities between the demographics of offline and online vulnerable populations have also been identified. For instance, as with traditional 'offline abuse', girls have been found to be targets of online sexual solicitations almost twice as often as boys (Mitchell et al., 2007). This supports the argument that similar characteristics and profiles precede vulnerability both online and offline. However, the integrity of these findings has been called into question as research on 'sexual solicitations' does not solely encompass dangerous predatory behaviour from older adults but includes advances from peers (Whittle et al., 2013b). This means that the motivations behind these solicitations and the danger they pose to vulnerable individuals remain unclear. Likewise, the stigma attached to boys being sexually abused may repress male victims from reporting these experiences, meaning they could be underrepresented in the majority of prevalence estimates (O'Leary & Barber, 2008). Without clear estimates of the rates of under-reporting, it is unfounded to use such prevalence rates to suggest that the vulnerable population is the same both online and offline. Rather, the data highlight that females are more likely to report inappropriate solicitations, both online and offline, than males – although this in itself remains speculative.

However, Whittle et al., (2013a), among others including Suler (2004), propose that the fundamental differences between the offline and online environment are so substantial that each environment needs to be examined separately in order to accurately understand vulnerability. For example, the disinhibition effects experienced by some people when interacting online could be fundamentally important when considering online grooming behaviour (European Online Grooming Project et al., 2012). According to Suler (2004), anonymity, invisibility

and minimal authority presence all feed into how people interact online. Suler (2004) acknowledged that disinhibition can be benign or indeed even be positive by helping to facilitate the discussion of thoughts on difficult issues. However, it can also be toxic and used to justify the use of criticism or threatening behaviour.

Whilst this holds some validity, the definition of grooming appears to remain constant irrespective of the environment, with only the techniques used to pursue this offence differing (Whittle et al., 2013a). Craven, Brown and Gilchrist (2006, p287) defined grooming as, "a process by which a person prepares a child, significant adults and the environment for the abuse of this child. Specific goals include gaining access to the child, gaining the child's compliance and maintaining the child's secrecy to avoid disclosure". Therefore, it could be that the online and offline environments share similar risk factors to vulnerability, and the impact this vulnerability has on the individual can be seen as comparable in both; with the methods used for pursuit in exploiting this vulnerability differing (Whittle et al., 2013a). Critically, we can use what we know about offline risk factors, along with the social phenotypes of ASD and WS, to offer suggestions on key factors that could place some individuals with developmental disorders in a vulnerable position online. The essential component here is that not all individuals who are developing typically, or indeed all individuals who have ASD or WS, will be placed in a vulnerable position online (e.g. it is not inevitable) but an awareness and a call for an understanding of the key issues is required at this time when the use of online social engagement is increasing.

8.3 Risk factors for vulnerability

For children following an atypical developmental trajectory, social vulnerability and the possible consequences of social vulnerability present a sizeable risk (Jawaid et al., 2012). When translated to the online world, this population are vulnerable as they may be unable to identify the inappropriate advancements of others, and may lack the intellectual capacity to evaluate why their own exploits may place them in a vulnerable situation (Whittle et al., 2013b). There are several risk factors to online social vulnerability, and the subsequent section highlights factors considered to be most congruent with having a developmental disorder, as illustrated by ASD and WS. We present important avenues for future research, which will allow us to enhance our understanding of online vulnerability in individuals with developmental disorders.

8.3.1 Trust

High levels of trust towards strangers have been implicated as a risk factor to vulnerability for young people with WS (Riby et al., 2013). It has been suggested that children as young as 5 years who are developing typically have the ability to use their judgement of facial expressions to decipher levels of trustworthiness to an accurate level (Willis & Todorov, 2006). However, the stranger-danger awareness literature has emphasized that many individuals with developmental and intellectual disabilities display elevated levels of stranger trust (Pinkham et al., 2008), which remain after their age-matched typically developing peers have learnt to make informed judgements of trustworthiness.

Adults with WS engage in a degree of indiscriminate trust and increased social approach behaviour, suggesting that even as adults this cohort has yet to refine their ability to make trustworthiness judgements (Fisher, 2013). On approachability tasks, Hanley and colleagues (2013) found that 'do not trust' judgements were especially difficult for WS participants (aged between 7 years 2 months and 38 years 10 months) to correctly identify when compared to typically developing controls matched on verbal ability or chronological age. Individuals with WS have difficulty making sense of the socio-communicative cues that help guide these judgements of trust. This difficulty to discriminate levels of trustworthiness, combined with their strong motivation to engage in social encounters (Frigerio et al., 2006) and reduced inhibitory control (Little et al., 2013), often leaves individuals with WS in a socially vulnerable position (Jawaid et al., 2012). However, the majority of this research has used methods concerning the emotional processing of faces, which may not map onto more naturalistic environments. Furthermore, it may not be possible to draw transferable conclusions between these data and an online environment, where faces have diminished presence. Thus, it remains unclear whether individuals with WS extend this trust behaviour to text presented online, and also what level of intellectual capacity and emotional awareness they have concerning potential deception. Indeed it is known that individuals with WS struggle with the subtle nuances of social interaction such as telling the difference between a joke and a lie (Sullivan et al., 2003). If it is hard for individuals with WS to make this distinction in face-to face-communication it may be equally hard, or even harder, online. Such an area is ripe for future exploration.

8.3.2 Involvement of parents

Fisher et al. (2012) devised the Social Vulnerability Questionnaire for use with individuals who had WS, which outlined four key contextual facilitators: risk awareness, social protection, perceived vulnerability and parental independence. One of the most significant correlates of social vulnerability for individuals with WS was parental independence. Parents of young people with WS were more likely to leave them unsupervised for prolonged periods of time and allow their child to spend unsupervised time with members of the opposite sex in the offline world compared to parents of children with ASD and parents of children with Down syndrome. This reduced level of parental involvement is of concern when considering the findings of the EU Kids Go Online survey (Livingstone et al., 2011), which noted that young people with disabilities experienced an elevated contact risk, meaning they were more likely than other groups to arrange meetings with people they have met online. This suggests that, not only are individuals with WS more likely to arrange meetings online, but these meetings may go undetected by parents due to the individuals' higher levels of parental autonomy. Further research is needed on the relationship between parental autonomy and contact risk in vulnerable young people.

8.3.3 Social isolation

Social isolation is also thought to be a significant precursor to social vulnerability (Jawaid et al., 2012). The lack of support networks mean that socially isolated individuals not only miss the protective influence of significant others, but it also minimises their opportunity to confide in someone about any untoward behaviour they experience, which has significant repercussions on their ability to cope

(Whittle et al., 2013b). Such a dynamic reinforces the cycle of vulnerability. Individuals with ASD have been widely reported to experience social isolation (Billstedt et al., 2005), and likewise up to 73% of adults with WS have been found to demonstrate social isolation as adults, with difficulties in relationship formation and maintenance (Davies et al., 1998). Ultimately, these difficulties result in feelings of loneliness (Bauminger et al., 2003). In the typical population, use of the internet, and specifically social media, has been found to enhance friendship quality, increase social interactions and minimise feelings of loneliness (Valkenburg & Peter, 2007). Thus, it may be that individuals with WS or ASD who have the intellectual ability to access the internet use this platform as a means of seeking out the social relationships that they lack in their offline world. Mazurek (2013) offered the first findings on social media use amongst adults with ASD, finding that around 80% used social networking sites, with 'forming social connections' being the most common reason for engaging in this activity. Content analysis of the data revealed that many individuals with ASD acknowledged that they used social media as an alternative way to try and engage with others. Heightened loneliness may result in an exaggerated desire to establish friendships online, which could result in the risk-taking behaviour described earlier, meaning these individuals could be facing an elevated level of online social vulnerability, in comparison to their typically developing age-matched peers. This is particularly pertinent as the European Online Grooming Project (2012) reported that offenders seek out loneliness and the subsequent desire for attention when selecting their victims, and successively exploit this to groom the individual online.

8.3.4 Mental health problems

Mental health problems have also been linked to enhanced vulnerability (European Online Grooming Project, 2012). Those experiencing depression or loneliness may be more motivated to seek solace in contacting strangers online (Wolak et al., 2004), and likewise, those with anxiety may feel protected by the diminished social presence that they usually struggle with, providing social stimulation without much of the social anxiety (Tian, 2013). This is of particular significance when considering the online vulnerability of individuals with ASD, as a recent meta-analysis estimated that anxiety disorders were present in around 40% of children with ASD (van Steensel et al., 2012). Thus, social networking could provide these individuals with a platform to communicate with minimal anxiety; particularly reduced social evaluation and social anxiety. Similarly, anxiety has been noted in individuals with WS as one of the most common mental health challenges (Stinton et al., 2010). Recent findings by Riby and colleagues (2013) found that higher anxiety levels were linked to more severe impairments in social functioning, which could be used as a possible explanation of why over 70% of adults with WS experience social isolation (Davies et al., 1998). As a result those with mental health problems may be driven to gain social connectedness online to overcome their social isolation. Research into this triangular relationship between mental health, social isolation and vulnerable presentation online in individuals with developmental disorders would offer much needed information on the impact and intensity of this relationship.

8.3.5 Disinhibition

When deciding how to present ourselves in both online and offline worlds, it may be necessary for us to use attentional control and execute planning behaviours – for example restraining and inhibiting impulsive behaviours that might have negative longer-term consequences. Indeed it has previously been proposed that some of the deficits of executive and inhibitory control associated with WS (e.g. Rhodes et al., 2010; Greer et al., 2013) could play a role in disinhibited social approach behaviours towards unfamiliar people (e.g. Little et al., 2013). Again it is unclear whether this may also relate to online social behaviours and the nature of information presented by an individual with WS or indeed with ASD due to the similar association between the disorder and attention and executive control deficits (e.g. see Kenworthy et al., 2010 for a review). Again this is an avenue for debate and research attention.

8.4 Discussion

It is argued that if real world vulnerability can be used as a predictor for online conduct and subsequent online vulnerability, then individuals with developmental disorders, as illustrated by the cases of ASD and WS, can face a high level of risk when interacting with others via the internet (Livingstone et al., 2011). This is supported by similar trends for transference in the typically developing literature (Whittle et al., 2013b), as well as the social phenotypes associated with ASD and WS. If individuals with ASD express a desire to form social connectedness (Mazurek, 2013), are unable to achieve this in the offline world (Chamberlain et al., 2007) and they are frequent users of screen based technology (Mazurek & Wenstrup, 2013), it would be reasonable to assume that this pursuit online

combined with their lack of social awareness could be placing them in a vulnerable position online. Similarly, we know that individuals with WS harbour an extreme prosocial drive to interact with others, both familiar and unfamiliar (Jarvinen et al., 2013), yet experience difficulty in forming and maintaining these relationships in the offline environment (Jawaid et al., 2012). Therefore, the online platform provides a novel way to interact with others, with minimised anxiety due to lack of physical presence, and this could combine with their lack of social awareness to place them in an extremely vulnerable position online. While we are not suggesting that all individuals with WS and ASD who engage in online interactions will be exploited or will be vulnerable, we do need to acknowledge that there may be risk factors that need to be taken into consideration and targeted for increased awareness and/or intervention. It is suggested that these suppositions do not just ring true for ASD and WS, but other individuals with developmental disorders who present as vulnerable in the offline world could be facing unprecedented online risks during their computer time. Whilst these prepositions currently remain provisional, we would argue that they follow a logical theoretical rationale to suggest that there is a serious issue of online vulnerability in individuals with developmental disorders that is currently being ignored by the literature.

Although it has been preliminarily reported that some individuals with developmental disorders use the internet as frequently as their typically developing peers (Mazurek, 2013), the first step in addressing the aforementioned void should be to extend such research to gain more information on the level of internet usage amongst young people with developmental disorders. This call for

research will stimulate questions on issues such as: the type of usage, level of supervision whilst online and whether there is an interaction between internet use, IQ and vulnerability. Such research would constitute the first stage in enhancing our knowledge with a view to informing an intervention needs that could help keep vulnerable individuals safer online.

Chapter Nine: Internet Use and Online Safety in Adults with Williams Syndrome

Based on the review presented in Chapter 8, it seems likely that individuals with WS could be at a heightened risk of experiencing online social vulnerability as a result of their social phenotype (Jones et al., 2000), and in particular their high levels of social motivation. Chapter 4 outlined parents' concerns about their son/daughters motivation to approach unfamiliar people, with little or no awareness of the dangers the interaction could pose. The internet allows individuals with developmental disorders to engage in this social approach behaviour through a different medium, offering a new world of friendships and relationships that may not be readily available to them in real life (Mazurek, 2013; Chamberlain, Kasari, & Rotheram-Fuller, 2007). However, there is little research on how often and why individuals with IDD use the internet. This chapter builds on the review presented in the previous chapter by exploring online social behaviour in adults with WS. This research offers the first insights into how often and why individuals with WS use the internet, and the types of internet scenarios which pose the biggest risk to their safety.

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9.1 Introduction

Internet use has been rising at a meteoric rate amongst the current generation. It is estimated that 83.3% of households in the US have a computer, and 74.4% have access to the Internet (compared to just 18% who had Internet access in 1997; File & Ryan, 2014). The Internet itself is now estimated to have almost three billion users worldwide (International Telecommunications Union, 2014). The Internet is now a necessity of everyday life, particularly as devices continue to become more portable, allowing for constant communication. This has substantial implications for the lives of today's 'Internet generation,' as the distinction between the real world and the virtual platform becomes increasingly blurred (Subrahmanyam & Greenfield, 2008).

The Internet has also become an important tool for individuals with intellectual and developmental disabilities (IDD). Adults with mild intellectual disability often have the skills needed to access the Internet independently (Katz, 2001; Davies, Stock, King, Brown, Wehmeyer, & Shogren, 2015). This is especially true as programmes become more user-friendly and the Internet becomes more intertwined with daily life (Boyd & Ellison, 2007; Davies et al., 2015). It is estimated that 97% of individuals with IDD have access to the Internet at home, and 41% have a computer in their own bedroom (Didden et al., 2009). Although the majority of individuals report using the Internet primarily for downloading music or playing games, several also report using the Internet for making social connections, such as sending or receiving e-mail, posting information about themselves on the Web, and chatting with friends (Didden et al., 2009).

While access to the Internet offers undeniable advantages to individuals with and without IDD-- providing access to resources and the opportunity to feel a part of an online community (Ridings & Gefen, 2004)-- there are risks to these connections. Recent high profile cases in the media have highlighted the dangers of the Internet for individuals without IDD, which too often include instances of cyberbullying, identity theft, and online grooming (e.g., behaviours to gain access to the individual by exploiting their trust, whilst maintaining the secrecy of the relationship; Craven, Brown, & Gilchrist, 2006; Mitchell, Jones, Finkelhor, & Wolak, 2011). Currently, little research has sought to understand the Internet use patterns and potential online victimisation of individuals with IDD.

9.1.1 Online Vulnerability of Individuals with IDD

Whilst everyone faces the risks posed by the Internet, individuals with IDD may be especially vulnerable to online victimisation, particularly if they have reduced IQ, difficulties interpreting social communication and understanding social nuances, elevated levels of trust, and feelings of social isolation and loneliness in their daily lives (Lough, Flynn, & Riby, 2015). Overall, compared to individuals without IDD, individuals with IDD are less likely to use social networking sites (82% versus 68%) or video chat rooms (32% versus 24%), but they are equally likely to talk with people whom they meet online (40% versus 41%) and more likely to display sexualized behaviour online (13% versus 20%; Wells & Mitchell, 2014). These latter two behaviours have been related to increased instances of sexual solicitation and harassment (Mitchell, Finkelhor, & Wolak, 2001). Thus, while the Internet allows individuals with IDD to engage in a world of friendships and relationships that are not necessarily available to them in real

life (Mazurek, 2013; Chamberlain, Kasari, & Rotheram-Fuller, 2007), being a part of the online community may also lead to increased vulnerability.

Just as risk for victimisation differs for individuals with different disabilities in the real world (Fisher, Moskowitz, & Hodapp, 2013), Internet use and online vulnerability might also differ by type of disability. Specific characteristics of certain disabilities may be more or less related to risk of online victimisation (Lough et al., 2015; Normand & Sallafranque-St-Louis, 2015; Wells & Mitchell, 2014). For example, individuals with Williams syndrome (WS) may be especially vulnerable to online victimisation. WS is a rare genetic neurodevelopmental disorder caused by the microdeletion of 25-28 genes on chromosome 7 (7q11.23; Hillier et al., 2003), affecting approximately 1 in 20,000 people. Individuals with WS display mild to moderate levels of intellectual disability (Searcy, Lincoln, Rose, Klima, Bavar, & Korenberg, 2004) as well as an extreme prosocial drive to engage with other people (Järvinen, Korenberg, & Bellugi, 2013), irrespective of whether or not they are familiar (Jones et al., 2000). Individuals with WS are described as being overly-friendly and trusting, with a lack of social inhibitions (Little, Riby, Janes, Clark, Fleck, & Rodgers, 2013). Yet, despite this, individuals with WS typically struggle to form and maintain peer relationships, resulting in high levels of social isolation (Davies, Udwin, & Howlin, 1998). Taken together, this profile indicates that individuals with WS are often considered to be socially vulnerable (Fisher et al., 2013; Jawaid, Riby, Owens, White, Tarar, & Schulz, 2012).

A key facet in the social vulnerability profile of individuals with WS is their social approach behaviour. Several studies (e.g. Bellugi, Adolphs, Cassady, &

Chile, 1999; Fisher et al., 2014; Jones et al., 2000; Martens, Wilson, Dudgeon, & Reutens, 2009) have examined social approach behaviour by presenting photographs of faces displaying various emotions and asking participants to indicate how much they would like to approach and interact with each person (e.g. Adolphs Approachability Task; Adolphs, Tranel, & Damasio, 1998). Research consistently reports that compared to chronologically age-matched peers, individuals with WS are more willing to approach the faces displaying positive (e.g., happy) facial expressions (Bellugi et al., 1999; Fisher et al., 2014; Frigerio et al., 2006; Jones et al., 2000; Martens et al., 2009; Porter, Coltheart, & Langdon, 2007). Such heightened willingness to approach someone simply based on their picture (without knowing anything about the person) has significant implications when considering the online vulnerability of individuals with WS, where often the only information available about a person is their online profile (e.g., a photograph).

No research has examined whether the real world social vulnerability and social approach behaviour of individuals with WS is also manifested online. To begin to understand the risk for online victimisation for individuals with WS the current study was designed to describe the online behaviour of adults with WS, and to examine the likelihood that individuals with WS might put themselves in high risk situations online. Three research questions were examined. First, what are the primary reasons young adults with WS use the Internet and how often do they go online? Second, do young adults with WS use social networking and how accessible are their social networking profiles? Third, how do adults with WS

respond to scenarios which have been designed for the purposes of this study to assess their online vulnerability?

9.2 Method

9.2.1 Participants

Participants were recruited from a residential summer camp programme for adults with WS. The sample included 28 adults with WS (22 male, 6 female) and their parents (3 fathers, 25 mothers). The average age of the participants with WS was 27.7, ±8.4 years and average full-scale IQ was 69.11, ±15.28. The majority of participants (24) lived in their family home, while three others lived in the community with supports, and one lived in supported living. The parents who took part in the study had an average age of 56.9, ±7.4 years.

9.2.2 Procedure

After receiving approval from the University's Institutional Review Board, parents of individuals with WS were contacted about their interest in participating in research as a part of the camp. Interested parents were then mailed a consent form and an assent form for the participant with WS to sign. Once the signed forms were received by the research team (100% return rate) parents were sent the link to questionnaires to complete online. The rate of completion was 93.6%, as 31 consent forms were received from the research team and 29 parents completed the questionnaires. The 31 individuals with WS subsequently completed adapted forms of the parent measures whilst at the residential camp. The completion rate with this group was 96.8%; one individual was unable to

complete the questionnaire due to limited comprehension of the survey questions. For the purposes of this study, only those individuals for whom we received both parent and participant responses were included in the analyses (n= 28).

9.2.3 Measures

Demographics. Parents completed a demographic questionnaire about their child and their family circumstances. In this questionnaire, parents were asked about age, living situation and employment status for both them and their child.

Kaufman Brief Intelligence Test, 2nd Edition (KBIT-2; Kaufman & Kaufman, 2004). The KBIT is a psychometric measure used to assess verbal, nonverbal and full-scale IQ. It can be used with individuals aged 4 - 90 years, and has been used in several previous studies with individuals with WS (for examples see Dykens, Rosner & Ly, 2001; Fisher, Mello & Dykens, 2014; Mervis, Kistler, John, & Morris, 2012).

Internet Use Questionnaire. The Internet Use Questionnaire was adapted from the EU Kids Online Survey (Livingstone, Haddon, Görzig, & Olafsson, 2011). The parents were asked to complete a questionnaire about the online activity of their child, and the individual with WS was asked to complete the same questionnaire about their own behaviour. The individuals with WS were provided with visual aids created for this study (e.g., pictures of thumbs up/thumbs down, pictures of a calendar with various number of days shaded, various social media icons) to support their comprehension of the questions and of the Likert scales. The questions covered topics such as how long they spend on the Internet, where they

use the Internet, which websites they visit, who they talk to online, and their social media activity.

E-Safety Scenarios. An e-safety scenarios task was designed to be completed by the individual with WS. This task was based on currently available online safety programmes for children, and was influenced by the Test of Interpersonal Competence and Personal Vulnerability (Wilson, Seaman, & Nettelbeck, 1996). It included 12 scenarios, which assessed the likelihood of the participants talking to or arranging to meet strangers they met online, sharing personal information and photographs, hiding information from parents, and giving away money. Three options were presented for each scenario, and the selected answers were scored on a scale of 1 - 3, with higher scores representing higher risk options. Specifically, the option considered the lowest risk (e.g., said 'no' to the request) was scored (1). The option considered the highest risk (e.g., agreed to the request) was scored (3). The final option (scored 2) had the potential to lead toward a riskier situation but provided a delay in agreeing to the request. The options were presented in a random order for each scenario. An example of a scenario presented was as follows; 'You met a new friend online named Alex. You like all of the same things and have a lot in common, but you have never met before in real life. Alex wants to meet up soon so you can do something fun together. What would you do?'. The participants could select their answer from the following options; (scored 3) 'Make plans to meet Alex as soon as possible', (2) 'Tell Alex you're busy but would love to meet sometime soon' or (1) 'Say no, you don't think it's a good idea'. (See Appendix B for the e-safety scenarios task). A total score was calculated with higher scores indicating higher risk taking online.

Scenarios were also rated as either social (e.g., meeting a person; n = 8) or non-social (e.g., giving out bank information; n = 4), and a mean score was calculated for each grouping. Cronbach's alpha reveal this scale had high reliability (alpha = .886).

9.3 Results

The data were analysed to determine Internet use, Social Networking use, and E-Safety behaviours of adults with WS. We then compared responses to social and non-social scenarios using t-test analyses.

9.3.1 Overall Internet and Social Networking Use

Internet Use. Based on parent report, 85.7% (24) of the adults with WS used the Internet every day or almost every day; 7.1% (2) used it once or twice a week; 3.6% (1) used it once or twice a month; and 3.6% (1) did not use the Internet. Of those individuals who used the Internet, 48.1% (13) used it for more than two hours each day, and 25.9% (7) used it for more than four hours a day (on weekend days). When asked how they access the Internet, 96.3% (26) used a mobile phone, 74.1% (20) went online using a portable device such as a tablet, 55.6% (16) used a laptop in their bedroom, 40.7% (11) used a gaming console, and 33.3% (9) used a PC in their bedroom.

Parental Supervision. Participants reported that few parents provide supervision while the individual with WS uses the Internet. Only 11.1% (3) reported that their parents sit with them while they use the Internet. Participants also indicated that

parents are not always aware of what they are doing when they are online; 29.6% (8) said that their parents knew nothing or just a little, 66.6% (18) said that their parents knew most or a lot of what they did online, and 3.7% (1) said they were not sure how much their parents knew. When asked what they do most often when they are online, the most common reasons for using the Internet were to watch videos (e.g., YouTube) and to access social networking sites (see Table 9.1 for additional activities).

Table 9.1. Participant reported online activities

	% use Internet (n = 27)	How often (%)		
		Once or twice a month	Once or twice a week	Everyday or almost everyday
Video (e.g. YouTube)	100	3.7	11.1	85.2
Social networking	85.2	13.0	21.7	65.2
Email	59.3	6.3	50.0	43.8
Instant message	51.9	7.1	50.0	35.7
Chatroom	44.4	33.3	25.0	41.7

Social Networking. Of the 85.2% of participants (n = 23) who reported using social media, 95.7% (22) reported that they could use it anytime, and 4.3% (1) could only use social media with permission or supervision from a parent. All of the participants reported that they most frequently visited Facebook for social networking. In further inquiring about their Facebook user profile, we found they had an average of 655 friends (range 15 – 1722) and 56.5% (13) had their profile set to 'public' (e.g., anyone can access), compared to 39.1% (9) who had a 'private' profile (4.3% didn't know). When asked about who they talk to on Facebook, 95.7% (22) said they talked to people they knew in real life, and 78.3% said they also talked to people they did not know in real life. Table 9.2 details the specific information that participants share in their Facebook profile.

Table 9.2. Percent of participants (n = 23) who provide specific identifying information on their Facebook profile

9.3.2 Internet Safety

Facebook profile information	% (*n*)
Picture that clearly shows face	95.7 (22)
Last name	91.3 (21)
Address	30.4 (7)
Phone number	56.5 (13)
School or job	73.9 (17)
Birthday	91.3 (21)

Responses are reported for the 27 participants who reported using the internet. First, Table 9.3 shows what percentage of respondents provided each answer for each of the e-safety scenarios. On the surface, these findings seem promising, in that participants seem most likely to select the option presenting the lowest risk. A closer examination, however, comparing the responses given to the different categories of scenarios (e.g. social versus non-social) presents a more nuanced picture.

Table 9.3. Responses to e-safety scenarios with percentage of respondents who selected each answer option for each scenario.

E-safety Scenario	3 (high risk) % (*n*)	2 (medium) % (*n*)	1 (low risk) % (*n*)	Social (S)/ non-social (NS)
Putting video camera on for unknown person	11.1 (3)	25.9 (7)	63 (17)	S
Sending photos of self to unknown person	33.3 (9)	14.8 (4)	51.9 (14)	S
Arranging to meet unknown person	37 (10)	18.5 (5)	44.4 (12)	S
Arranging to go to unknown person's house	29.6 (8)	3.7 (1)	66.7 (18)	S

Opening unknown photo file	3.7 (1)	48.1 (13)	48.1 (13)	S
Sharing password	7.4 (2)	7.4 (2)	85.2 (23)	NS
Paying to enter competitions	22.2 (6)	7.4 (2)	70.4 (19)	NS
Giving out bank account information	7.4 (2)	22.2 (6)	70.4 (19)	NS
Accepting friend request from unknown person	14.8 (4)	29.6 (8)	55.6 (15)	S
Clicking link to e-mail unknown person	3.7 (1)	33.3 (9)	63 (17)	S
Keeping online relationship a secret from parents	29.6 (8)	18.5 (5)	51.9 (14)	S
Hiding online behaviour from parents	14.8 (4)	7.4 (2)	77.8 (21)	NS

To further explore the e-safety responses, we examined the mean differences for social versus non-social scenarios (see Table 9.3 for rating for each item on a scale from 1 to 3). A paired samples t-test revealed that the participants were significantly more likely to engage in risky situations that were social (mean = 1.65, ± .54) rather than non-social (1.37, ± .50; $t(26) = 4.62$, $p<.001$) in nature. According to Cohen's (1988) suggestions, this was considered to be a moderate

sized difference, with a *Cohen's d* value of 0.54. To examine this one step further, we divided the social behaviours into meeting people in real life versus talking with people on the Internet. Adults with WS were significantly more likely to agree to arrange to meet with an unknown person in real life (1.85, ± .72) compared to talk to an unknown person online (1.60, ± .50; t(26) = 2.54, p<=017, *Cohen's d* = .40) or engage in a non-social risky online activity (1.37, ± .50; t(26) = 4.88, p<.001, d = .77). Participants were also more likely to talk to an unknown person online compared to engage in a engage in a non-social risky online activity (t(26) = 4.42, p< .011, d = .46). There were no differences between gender and living situation and scores on the total E-Safety Scenarios task, nor on the social and non-social groupings. Scores also did not correlate with age or total IQ.

9.4 Discussion

Previous research has already identified individuals with WS as being a socially vulnerable group in the real world (for a review, see Jawaid et al., 2012). Such real life social vulnerabilities could be exaggerated in the online world (Lough et al., 2015). By asking individuals with WS about their online behaviour and responses to specific scenarios, the current study provides the first insight into their Internet use patterns and level of online vulnerability. These findings not only help us to understand how often and why individuals with WS are using the Internet, but they also help to inform the development of Internet safety interventions.

First, we found that adults with WS frequently use the Internet and the majority of our participants used social networking sites such as Facebook everyday or almost everyday. Such findings are in line with reported Internet use (97%; Didden et al., 2009) and frequency (25.9%; Wells & Mitchell, 2014) in other samples of individuals with other IDD. Of interest, we found that parental supervision or oversight when individuals with WS used the Internet was low and several participants indicated that their parents did not know everything that they did online. The individuals with WS in this sample therefore seem to be relatively autonomous when using the Internet.

The second finding is that adults with WS share a large amount of identifiable information on their social networking profiles. Additionally, individuals with WS are Facebook "friends" with several hundred people, both known and unknown to them in real life and participants indicated they often spoke to people online who they did not know in real life. These findings are significant as previous research indicates that talking with strangers (i.e. individuals met on the Internet) is a prominent risk factor for sexual solicitation online (Mitchell et al. 2007, 2008). Previous research has also highlighted the concern that lack of sexual education for people with IDD, combined with their desire to meet people online, could increase the risk of sexual cyber-victimisation (Murphy & O'Callaghan 2004). Thus, considering the amount of personal information that participants are sharing with virtual strangers, concerns for their safety are raised.

In our third finding, we note that adults with WS are more likely to agree to engage in socially risky behaviours compared to risky behaviours that are not

social in nature. In fact, the more risky the behaviour, the more willing the individual was to engage in the behaviour. Thus when their online visibility is considered alongside their propensity to agree to meet up with strangers who they have only spoken to online, it would seem that this group is at very high risk when interacting with other people online (Lough et al., 2015). It may be, however, that individuals with WS do not have the same opportunities as their peers to meet with someone they have spoken to online. Whilst this is a viable suggestion that could alleviate some concern for their vulnerability, recent work by Fisher and colleagues (2013) found parental independence in WS to in fact be a disability-specific correlate of their social vulnerability. This suggests that they are likely to have at least some opportunities to meet with "online friends" in person. Further, the EU Kids Online survey (Livingstone et al., 2011) noted that people with intellectual disabilities faced an elevated contact risk compared to their peers, suggesting they would be more likely to arrange to meet with strangers they have met online.

These findings are situated within an on-going debate about the overlap between real world and online existence (Whittle, Hamilton-Giachritsis, Beech, & Collings, 2013). It has been argued that the online and offline worlds have become so embedded in one another, that it has become impossible to disentangle one's offline and online existence. For individuals with IDD, this means that the factors that feed into their vulnerability in the real world are likely to also serve as risk factors in the online world. The findings from the current study certainly lend support to this argument. Individuals with WS are known to be highly trusting and disinhibited during social interactions (Pinkham, Hopfinger, Pelphrey, Piven,

& Penn, 2008; Riby, Kirk, Hanley, Riby, 2014), and show a diminished stranger danger awareness (Riby et al., 2014; Fisher, 2014). When presented with the e-safety scenarios in the present study, almost half of the participants indicated that they would go and meet with someone who they had been speaking to online. It would therefore seem that the traits that feed into the social vulnerability of these individuals in real life, also help shape their vulnerability online (Lough et al., 2015).

There are also fundamental differences, however, between the offline and online environments that could shape the qualitative nature of the vulnerability experienced. As an example, the Internet offers increased freedom as well as anonymity during online social interactions which would not be available in face-to-face interactions (Suler, 2004). Whilst this may be liberating and facilitate openness in conversations, it can be problematic for individuals who struggle to understand social boundaries. It also affords anonymity to the people that they are interacting with, which is likely to prove dangerous considering the high levels of trust that individuals with WS employ (Riby et al., 2014).

Given the social nature of adults with WS, both on and offline, it seems imperative that a parent or guardian should monitor Internet use and social interactions initiated through social networking. Taking such a stance may be difficult, however, as adults with IDD often wish to (and should be allowed to) maintain their own independence and autonomy (Northway, 2015; Wehmeyer & Garner, 2003). An alternative approach, often taken by parents of youth without disabilities, would be to use filtering software (e.g., CyberPatrol, NetNanny) that

can block access to dangerous sites or to insist their children do not visit social networking sites (Tynes, 2007). Yet results from the current study and from previous research indicate that the individuals with WS might not tell their parents what they do online or they might hide certain activities.

Perhaps a different approach, then, would be to teach Internet safety skills to adults with WS. Equipping individuals with WS with specific strategies to use when responding to risky situations may help to decrease vulnerability online. For example, individuals without IDD often develop their own strategies for staying safe online, such as increasing their privacy settings or minimizing interactions from unknown individuals (Tynes, 2007). Our results indicate that adults with WS do not similarly employ these techniques, as more than half had a public Facebook profile and almost half indicated they would agree to meet an "online friend" in person. Thus, while individuals without IDD might be able to employ Internet safety skill on their own, individuals with WS may need more explicit Internet safety skills instruction. Unfortunately, we are unaware of any such research on interventions for teaching Internet safety skills to individuals with WS, or any disability.

It light of our current findings, it seems pertinent to start with teaching adults with WS about what personal information is okay to share and which should be kept private (e.g., address, school), who is and is not appropriate to accept as a friend, and how to decide whether an "online friend" is okay to become an "offline friend". Perhaps even more simply, teaching adults with WS about safety in general could enhance their safety online. For example, similar to the approach

taken in a stranger safety training conducted with adults with WS (Fisher, 2014), adults with WS should be taught to always let a trusted individual know where they are. It is estimated that from 9% to over 40% of young Internet users have face-to-face meetings with a person first encountered online and in 30–61% of cases, their parents were unaware of these meetings (Baumgartner, Valkenburg, & Peter, 2010; Livingstone et al. 2011; Helweg-Larsen, Schutt, & Larsen, 2012). If this simple rule is followed, then at least someone will know of their whereabouts if they go to meet a stranger who they have met online.

While the results of this study are an important first step in understanding the online social vulnerability of adults with WS, certain limitations should be addressed. First, the participants with WS were a part of an overnight camp that required them to display few behaviour problems and to be able to stay away from home. As such, these participants might not be a representative sample of adults with WS. Additionally, because these participants were potentially more independent than the broader population of adults with WS, it is possible that they were subsequently afforded more independence online as well. While we found no relation between age, IQ, gender, or living situation and responses to the E-Safety Scenarios, a larger sample of adults with WS with a broader range of functioning is needed to more fully understand the online social vulnerability of adults with WS. This study also did not have a control group to which our findings could be compared. The percent of Internet and social networking users, however, were similar to those reported in previous studies (Didden et al., 2009; Wells & Mitchell, 2014).

Despite these limitations, the research presents important findings for future research and intervention development. These results also re-affirm the anecdotal evidence we often hear from parents expressing their concern for their children's online safety, especially as their child gets older and becomes more independent. While the Internet provides a wealth of opportunities and resources to enhance the everyday lives of adults with WS, it also poses threats which are arguably more dangerous than those they face in the real world. As the Internet continues to become more accessible, future research should further examine the online vulnerability of individuals with WS, and intellectual disability more broadly. Such research should continue to explore specific disability status information to determine whether features of certain disabilities have different implications to risk of online victimisation (Wells & Mitchell, 2014). Once a more complete knowledge base is built, then the effectiveness of both existing and novel e-safety educational strategies can be examined.

Chapter Ten: A case study analysis of social behaviour and internet use in Williams syndrome

Based on the review presented in Chapter 8, and the results from Chapter 9, it is clear that some individuals with WS could face a high level of risk when online, particularly when they are interacting with other people over the internet. For individuals with WS who seek social interactions (Frigerio et al., 2006), whilst simultaneously experiencing high levels of loneliness (Davies et al. 1998), it is clear why connecting with others via the internet is an attractive prospect. However, their atypical patterns of trust behaviour, outlined in Chapter 6, could place them at a disadvantage in identifying danger online. We know from Chapter 4 that parents are concerned about their son/daughter's social approach behaviour in the real world, but little is known about how parents view their son/daughter's social behaviour online. This chapter contains data from a pilot case study, which provides in depth detail about the online behaviour of one adult with WS. This is drawn from a semi-structured interview carried out with the adult, and a separate interview with their parent. The methodological strengths and limitations of this multi-informant approach are discussed in Chapter 2, along with the merits of employing a case study design for this type of research.

10.1 Introduction

A common behavioural characteristic of Williams syndrome (WS) is hypersociability (e.g. as seen in chapter 4); they experience an exaggerated desire to interact with other people, showing little, if any, stranger danger awareness (Riby et al., 2014b). Individuals with WS typically display a gregarious and outgoing personality (Klein-Tasman & Mervis, 2003), and they are highly driven to seek out social interactions (Little et al., 2013). Recent work by Ng, Järvinen and Bellugi (2014) identified that the likely motivation underpinning this extreme prosocial behaviour is a desire to form affectionate relationships. This is in contrast to their typically developing peers who sought out social interactions to enhance their positive emotional well-being, and to exert influence over others. Yet despite this hypersociability, these individuals struggle to form and maintain peer relationships, and experience high levels of loneliness, especially during adulthood (Davies et al., 1998).

Atypicalities of social cognition have been linked to the hypersociability witnessed in WS (Jones et al., 2000). Individuals with WS show atypicalities in the way that they process facial features and interpret affect. They show prolonged gaze towards the eye region within faces (Riby & Hancock 2009), and experience difficulties disengaging (especially but not solely from faces). Whilst they can recognise simple facial expressions, such as happy or sad, they struggle to decipher more complex emotional expressions (Plesa-Skwerer et al., 2006). Indeed, although individuals with WS are known to have relatively more proficient language abilities in comparison to their visuospatial profile (Losh, Bellugi, Reilly & Anderson, 2000), they in fact struggle to follow the subtle

social nuances of a conversation, such as sarcasm or irony (Pinheiro et al. 2011). Indeed, Laws and Bishop (2004) found that despite the strong social interest shown by individuals with WS, they show significant pragmatic language deficits, and poor social relationships. This means that individuals with WS typically show an extreme desire to approach and interact with other people irrespective of their familiarity, they maintain prolonged eye gaze during interactions yet struggle to fully understand the complexities of the conversation they are engaging in. When combined with their mild-moderate levels of intellectual impairment (Searcy et al., 2004), they are thought to be highly vulnerable to victimisation (Jawaid et al., 2012). Perhaps unsurprisingly, therefore, parents' report that they are extremely worried about their son/daughter's social behaviour and in particular their propensity to approach strangers (Lough et al., 2016), as emphasised throughout this thesis.

The hypersociability observed in WS is likely to be displayed not just in their everyday social interactions, but also in their online social interactions (Lough et al., 2015). The rise of the internet has brought with it new methods of communication, and as such, has opened up new opportunities for social interaction. Little is known about how or why individuals with developmental disorders use the internet, and whether the reasons for using the internet are the same as those seen in TD individuals. Due to the level of cognitive functioning needed for internet use, an adult with WS was selected for the study. Isolation and high levels of anxiety are thought to be especially evident in adulthood (Bregman, 1996). Plesa Skwerer et al. (2006) found that adults with WS (age range 18 – 36.9 years) showed impaired social-perceptual abilities, which were comparable to

those seen in adolescents aged (12 – 17.11 years). It therefore seems that impaired social functioning in WS persists into adulthood.

Previous research in individuals with developmental disorders has shown discrepancies between parent and self-reports about social profile. Recent work by Schriber, Robins, and Solomon (2014) found that young people with ASD (8 – 18 years) gave overly favourable reports of their personality characteristics, whereas typically developing children showed a tendency to "self-diminish". Individuals with ADHD have also been found to show a lack of awareness of their own deficiencies (Hoza et al., 2002). In WS, Fisher, Mello and Dykens (2014) examined parent versus self-report of social approach behaviour and found parents reported higher levels of approach behaviour for their son/daughter than the individual reported for themselves. When these reports were compared to direct behavioural observations, it was found that parent reports were more similar to the observed behaviour than self-reports. It is therefore important to adopt a multi-informant approach, in order to highlight similarities and differences in accounts of social behaviour and internet use.

A case study approach offers a method of exploratory research enquiry, in which a real-life phenomenon can be explored in the context in which it exists (Yin, 1984). Qualitative case study data will enable us to capture the complexities of the social behaviour seen in WS, which may be over-simplified if an experimental approach was employed. As there is currently no literature on internet use in WS, this approach will provide some in-depth information which will be used to guide any future studies on this issue. The aim of the current study

was therefore to employ a case study approach to gain further insight into social behaviour in WS, and to present one of the first insights into internet use in WS. The overlap between social behaviour and online social behaviour was also explored. A multi-informant method was selected in order to gain insight from the perspective of an adult with WS, and from their parent.

10.2 Method

10.2.1 Participants

An adult with WS (age 23 years) and her mother (age 48 years) were recruited through the Williams Syndrome Foundation UK. To ensure anonymity, the adult with WS was given the pseudo name of Alaina. Alaina had previously received a formal diagnosis of WS using fluorescent *in situ* hybridisation testing. The study received favourable approval from the local ethics committee.

10.2.2 Materials

Social Behaviour and Internet Use Interview – Parent and Adult Schedules

A semi-structured parent interview was designed to address social behaviour and internet use in Williams syndrome. There were four sections covering a general introduction, social profile, computer use and risk and resilience (see Appendix C). The first section was developed to find out about their immediate family structure, as well as their daily routine and interests, and any particular strengths they have. Here, information about any co-morbid diagnoses was also obtained. This information was designed to provide sufficient grounding to move on to the second section, which focused in on their social profile. Questions were asked

about peer social groups and how these have changed over time and about their interaction style with familiar and unfamiliar people. This section progressed to questions about trust and deceit, and ended with a question about experiences of anxiety. By understanding more about their social profile, the third section was able to address their computer use and online social persona. Questions were developed to probe reasons for internet use, benefits and dangers of internet use, rules, supervision and parental concerns. The final section was titled risk and resilience, which posed questions about vulnerability issues more directly. An adapted version of this interview schedule, focusing on the areas of social profile and compute use was used for the adult with WS (see Appendix C for full interview schedule).

10.2.3 Procedure

The parent interview took place over the telephone, and lasted for 68 minutes. The researcher met with the adult with WS in their home to conduct the interview, which lasted 47 minutes. It was made clear to the participants that they did not have to take part in the study, and could withdraw at any time. The interviews were recorded, and the content was transcribed verbatim.

10.2.4 Data analysis strategy

A case study approach was employed, and data was analysed using thematic analysis, in line with the principles of Braun and Clarke (2006). The interviews were analysed separately, and initial conceptualisations were developed from line-by-line coding. These codes were developed into themes, and the themes were applied back to the transcripts until saturation was reached. The same

procedure was used to analyse the interview with the adult with WS. At this point, similarities and differences between the themes generated by the two accounts were analysed, in an attempt to generate higher order themes which encompassed the accounts given by both participants. Within each theme, the narratives were compared and contrasted to produce results which explore different perspectives on the same issues. Each transcript was coded for reliability. Cohen's kappa showed "substantial" agreement between coders for the account given by the adults with WS ($\kappa = .804$, $p<.001$) and "almost perfect" agreement between raters for themes generated from the parent account ($\kappa = .834$, $p<.001$; Viera & Garrett, 2005).

10.3 Results

The results from a thematic analysis are shown in Figure 10.1 below. These themes were generated from the responses given by the adult with WS, and from her parent. Whilst the themes generated from the accounts given by Alaina and her mother are the same, the content of these themes is different. The differences between the participants' accounts are therefore highlighted throughout.

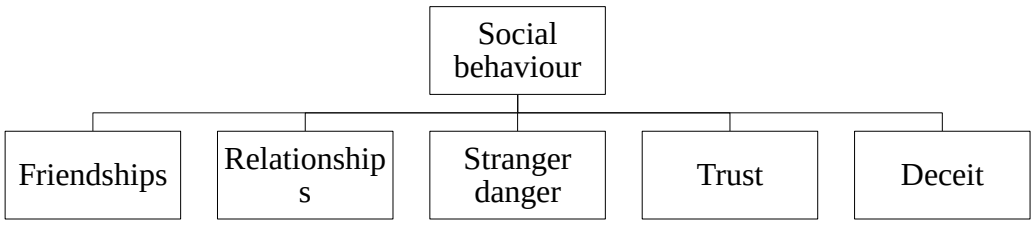

Figure 10.1 Thematic map of themes linked to social behaviour

10.3.1 Social behaviour

The overarching themes which emerged from the interview were accounts of the hypersociable behaviour displayed by Alaina. The parent gave examples of her daughter's strong desire to interact with others, in contexts where it may not be considered socially acceptable:

> "She will want to tell people who are serving her in shops one or two things about herself while she's getting the money out of her purse or tells them a funny thing that's happened so she will try to engage people in more than the functional thing that she's doing wherever she goes" (parent)
>
> "She will contact people daily, people she's actually not seen for a year. She often gets obsessed with certain people, and there was this family she became quite obsessed with, she kept ringing the house and unbeknown to us she was ringing daily and at first they humoured her and answered her calls then finally the parents rang us up and said do you know and we didn't because she goes upstairs and rings them from her bed so we can't hear she's on to them again" (parent)

She also commented on how this behaviour worried her, and how even though her daughter was 23 years old, this type of social behaviour meant that her independence was still limited:

> "There's the worry about trying to tell her she has to come straight home from places and not go round chatting to people so it has affected her independence. We can't let her go out in the evenings really because she's just too small and vulnerable to be travelling alone after dark" (parent)

Interestingly, the focus on social behaviour primarily came from the parent interview, with Alaina talking less directly about her social behaviour, and more about the elements that make up social behaviour, such as friendships.

Friendships

The theme of friendships emerged from both interviews, offering very different perspectives. Alaina considered herself to have hundreds of close friends, whereas her parent identifies these friendships as being more like acquaintances.

> "To her, a friend is just someone who has smiled once at her" (parent)
>
> "I have like over 200 close friends… I'm still in contact with them all and I have a friend, this is amazing right, I have a friend and my mum pays her because my mum goes to work and she can't take me out so this friend gets paid by my mum to take me out"

According to her parent, Alaina struggles to form and maintain friendships, particularly with her peers:

> "It was very difficult with friends she always wanted to see people more than they wanted to see her and unless you've got kind hearted people who deliberately give her a little bit of time she couldn't really hold friends her own age very well" (parent)

Her understanding of what a friendship is could be particularly important when considering issues of vulnerability. Her parent gave an example of Alaina's broad definition of a friend, and the socially inappropriate interactions that follow:

> "She was at school locally and she left her high school when she was 17 so it's now 7 years ago and she's really really good with faces so people I wouldn't have a clue about because they've all changed from 16 year old boys to 23 year old men but she knows them and she says he's my friend he was at school with me and she will run up to them and expect a hug and that's a bit worrying because it was a big school" (parent)

Relationships

Alaina's difficulty interpreting social cues and responding appropriately expended to romantic relationships. Her mother explained how her daughter can misconstrue kindness from males as a sign that they would like a relationship with her:

> "There are boys who she likes, but they're all people who have face-to-face been quite kind to her, that tends to be the people who she wants to be friends with. So they're all really nice people and they all try to do their best and give her a bit of attention but then to her it gives the wrong message. Some of the boys she's then thought they fancy her and want to go out with her but then she'll start ringing them or contacting them on Facebook a lot until they finally tell us that they can't cope, it's the same pattern each time" (parent)

From Alaina's perspective, she thinks that she has been in a relationship, but she seems to struggle to differentiate friendship behaviour with relationship behaviour:

> "I met a boy on my course and he accepted that I liked him actually he didn't run away from me so we were on and off for like 3 years we didn't kiss or hold hands or anything but he did really like me. I'm going to stay single until I find the right right right guy. But the people I've been dating have been Mr Right, they've ticked all my boxes but they just didn't want to go out with me!"

Strangers

There were discussions around strangers with both interviewees. Alaina was asked what a stranger was, and how she could tell the difference between who was a "good" stranger, and who was a "bad" stranger:

> "I think strangers could be crazy they could do anything. One of them said would you like to come with me in my car, just like you have to be careful, like I knew they were going to kidnap me so I said no I don't want that"
>
> "A bad stranger…. I can tell like if they've got a smirk on their face or if they're wearing glasses … and like the way they smell, and the things they're saying to me… but a good stranger is if they're friendly and chatty, then you know it's OK. They will be smiley and happy, I could talk to them"

Her differentiation appears to be on a surface level, focusing on how they look and their demeanour. Based on this, she shows little awareness of the possibility of deceit. This blurred definition seems to be something that she has held since

childhood, as her parent explains the logic Alaina applies when making judgements on friendships and strangers:

> "When she was little she liked walking and she would want to stop and talk to everybody coming the opposite direction and we tried to teach her what the difference between a stranger and a friend was, so strangers you just say hello and friends so people she knew she could talk to a little more but once people had said hello she would say well they said hello so now I know they are my friend and even though we've explained it many many times to her she has never really let go of that so once people have said hello they are her friends." (parent)
>
> "She just seems unable to understand she strongly strongly disagrees to the point where she really does believe with great conviction that she's right and these people are her friends! When I say but they're strangers but she just says well you're wrong they're very nice they asked me about my day." (parent)

Her strong conviction that many strangers are her friends means that she frequently interacts with strangers, and opens up to them about personal information. This inability to tailor her social behaviour to the familiarity of the other person is something which her mother reports as concerning:

> "There are just some people on the bus who I talk in person to I only see them on the bus but they're just people from my town and who have been on the bus with me for ages and who I thought yeh I want to talk to them"
>
> "Because of her voluntary works he travels regularly on the bus into town at commuting time so there's a regular set of people on the same bus as

her in the morning and she calls them my friends and we try to say they're not your friends and she must never try and get off the bus and go anywhere with them which she's never actually tried to do but there's no distinction between these people and people she knows more deeply" (parent)

"There was a day recently when we fell out and she had basically a counselling session from a stranger on the bus and she repeated back to me all the things that this lady had said to her which thankfully were all very appropriate but I thought afterwards that's not good she had obviously left home slightly annoyed by our conversation and had opened up to a complete stranger about it." (parent)

Indeed, when Alaina was asked what she does when she meets someone for the first time, she said:

"Well for me I just say like hi, how are you? And then when I see them again I say like I miss you to ensure that they're ready for me because you know the friends that I see a lot give me cuddles and stuff and that shows me they've missed me. Then next time I give them a hug and literally I'm very emotional. One of my friends I had a big cry of joy with that I got to meet up with them again … it has been a long, long while, like a month"

It is clear from this passage that strangers are quickly seen as friends by Alaina, and there is a great deal of emotion attached to these "friendships".

Trust

The issue of trust seems highly relevant given the high level of hypersociability described, as well as the broad definition of friendship. When Alaina was asked how she knew if she could trust someone, she replied:

> "Well it's like if they show it to me if they act as though they love me and comfort me … like my friends said to me we'll look after you and things like that and that showed me I could trust them so saying nice things to me"

Again, her definition of trust is closely linked to kindness. For Alaina, people who say nice things to her are people who she can trust. According to her mother, Alaina's default position when interacting with other people is to trust them:

> "Very easy that's her basic position, to trust everybody … It's not that she struggles, I don't think she has the ability there at all to make these trust judgements." (parent)

Deceit

Linked to trust, Alaina's mother explains how her daughter struggles to identify deceit:

> "If someone sophisticated lied to her I don't think she would have a clue. I think she would be very vulnerable to grooming and that's still a concern really" (parent)

However, Alaina identifies herself at being very good at knowing when someone is lying. It seems that the difference could be attributed to Alaina's understanding of what a lie is:

> "I know what lies are because my teacher said to me that saying sorry is not enough. And my friend said to me that he didn't want my help and he didn't want my comfort and now he does 3 or 4 years later. People do that all the time to me, they lie to me."
>
> "If she said she will be there at a certain time and the bus lets her down, she gets anxious that she's lied she can't seem to understand that she's actually been held up by public transport and she didn't lie because she didn't know." (parent)

Again, she seems to rely on facial expressions to guide her judgements about deceit. She identified people who lie as looking "*very angry and guilty*", whereas being who are not lying are "*happy and smiley*". In terms of Alaina's ability to deceive other people, her mother said it is rare, but when it does happen, it is usually socially motivated:

> "So she doesn't like deceiving people. She just tends to go silent. In terms of deceiving me, there have been one or two people I've felt were not so suitable and she's been adamant that she is going to meet up with them and she just goes off radar by text, she turns her phone off then she'll appear back home 3 hours later with me on tenterhooks but that's quite rare."(parent)

10.3.2 Internet use

A second thematic diagram was produced to depict themes which came from the discussion of internet use. Responses from the adult with WS and her parent were used to generate these themes.

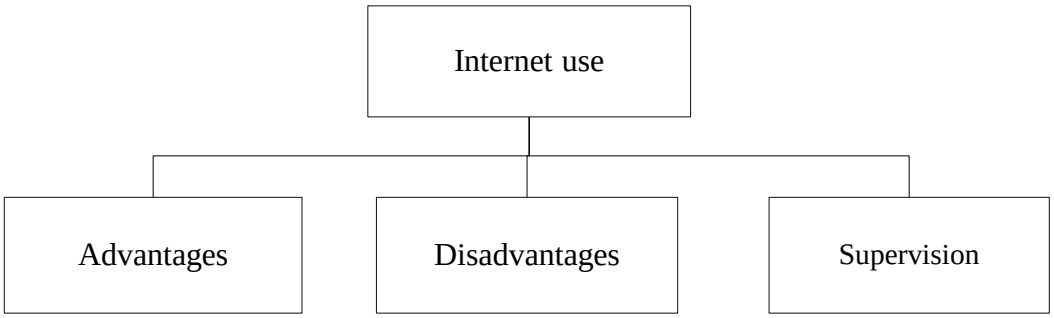

Figure 10.2 A thematic map summarising the key themes on internet use

Alaina uses the internet for a variety of reasons, and her mother reports that now she is an adult, she spends most of her days on the computer:

> "I go on Facebook. I go on YouTube and watch music videos and watch videos of things like washing machines and proposals you know nice little things"
>
> "We restricted her computer use quite a lot until she was 18 to half an hour twice a day then up to an hour then now that she chooses what she does especially when we're out of the house she will be on the computer most of the time … she always has lots of tabs up and it's usually a soap, Facebook, a washing machine and a heavy metal thing those are mostly what she does." (parent)

Interestingly, her broad definition of friendship extends to online social networking, as Alaina said she has over "1,000 friends on Facebook". Her

hypersociability, and difficulties understanding social nuances, also seem to be evident in her virtual friendships:

> "When people want space or when people want me to end the friendship they tell me. I've ended a few friendships over Facebook because they weren't wanting me as a friend they weren't wanting my love they just didn't see how I felt. They messaged me saying that loads and loads and loads of times so I ended the friendship. I've only started to speak to a few of them again properly now because this time they're ready they weren't ready last time I just wish they could have seen it. I just know they're ready. They need to understand that it's just me trying to show love to them. My mum says if they don't want me I have to let them go but you know I'm a special needs person so I can't do that it's very hard"

Advantages

Alaina was keen to stress the advantages of the internet for her, highlighting that it opens up new methods of social interaction and communication.

> "To find out interesting things and communicate with friends and stuff. And I can talk to friends I don't get to see at all. So like I can see their face, I can see what's happening in their life, I can see how I can help them out … I like the word comfort in a way that most people don't really understand what comfort is. I like it in the way that you can look after people because they're nice and friendly"
>
> "I just feel that like Facebook has opened up so many new things like before Facebook I was ringing up people's houses and sometimes people don't answer the house phone and now I can message them without

having to ring them. So one time I was in trouble for ringing my friend's house but it was their fault it wasn't mine because they didn't know by then what I was meaning they didn't know that I was just being a friend then they rang my mum behind my back. I've had to end friendships because they didn't show me love they didn't want me to call them or contact them at all they didn't want to hear my news or anything"

This was something that was also emphasised by her mother. These online friendships give Alaina some of the social stimulation that she seeks:

"It gives her proxy friendships really when friendship is difficult elsewhere" (parent)

"Facebook it keeps her in touch with people who perhaps wouldn't keep in touch with her so she follows some people who she admires, she follows them avidly, so she'll know the names of their families and friends she'll know what parties people are going to even if she's not invited herself so that gives her a sort of virtual friendship with people which doesn't depend on their will to keep it going. There's also some people who will give her 5 or 10 minutes and they chat in the chat box and she just loves that I think that's one of the reasons she does it and people do comment on her status and will answer one or two questions for her and then log off" (parent)

Disadvantages

Both Alaina and her mother acknowledged that there were disadvantages to using the internet, although they saw the types and severity of these disadvantages very differently. The focus for Alaina was on being tricked by scam e-mails:

> "Little things like they might trick you and some things like e-mails that come up that you shouldn't open but you want to open, like my church had a virus going round and I so wanted to read the email and I couldn't so little things like that really but apart from that its good"

It seems that Alaina's awareness of the wider risks associated with the internet is low:

> "Because she has Facebook on her phone, she was going to a course, and she put as her status I'm going to this place, who wants to meet me, and there was no-one accompanying her so I think she made herself very vulnerable saying she would be in a certain café at a certain time and 1,000 people know that she's there and waiting to talk, it's very very open to abuse." (parent)
>
> "She knows that we think there are some dangerous things on the internet but she doesn't really agree with us … In her mind if you asked her she might be able to give you the right answer, but in her heart she doesn't really believe any of it." (parent)

Many of the disadvantages that her mother associates with the internet again link back to Alaina's definition of friendship:

> "It's this definition of a friend because anyone who was at the same high school as her at the same time she sees as a friend so that's mostly how she's got these massive numbers and she had quite a phase of if those people started going out with somebody she would want to be friends with the new person as well so friend of a friend and that's the one that worries me really because she can't see the difference between that and someone who she has met face to face. (parent)

Indeed, whilst her mother said that Alaina is likely to talk to her about things that have upset her online, she also talks to people online about things that have upset her. This lack of social boundaries is something that seems to be present for Alaina both offline and online:

> "If we have caused upset through trying to limit activities or insist on certain activities at home, then she will pour her heart out to a relative stranger online so it's the flipside of it if they upset her she'll come to us, but if we upset her she'll go to them." (parent)

These similarities led her parent to conclude that the vulnerability experienced by Alaina in her daily life is very similar to that she experiences online:

> "It's the same really. Except it's a bit more difficult online because it's there in the home isn't it, you can't lock the door and it's gone." (parent)

Supervision

Alaina's mother tries to set rules to keep her safe online, but acknowledges that they are difficult to enforce:

> "We've said that her computer has to stay downstairs in the living room so there are people looking over her shoulder quite a bit but not when we're at work and she's at home so it is difficult to monitor it because she doesn't want us to monitor it and there's plenty of time for her to do it alone." (parent)

Alaina views her mother's input in a positive way, seeing her as being helpful rather than over-protective:

> "She doesn't act over protective which is nice she just helps me and I tell her if anything goes wrong and she tells me what to do then I don't speak to that person for a while until they come back. Like my friends I've known for years I wanted to help them out with something and they were like sorry we can do it on our own thanks we don't need you so I talked to mummy and the friends rang up and literally talked to mummy loads of times saying that they don't want me and I wasn't letting them go"

Interestingly, her mother thinks that limiting access to the internet is particularly dangerous, as she believes that Alaina will seek social interaction elsewhere:

> "We used to do it by restricting access so giving her limited hours and not having the internet on and things like that but I find that more dangerous now to be honest because she just wants to look outside the house for company if she can't be on the internet at home watching videos and things." (parent)

But as Alaina gets older, her mother has conceded that it is becoming more challenging to supervise her activity online.

> "The situation is becoming more complex it's just a question of who's got authority really cause I don't really legally have any authority over her although she doesn't really know that." (parent)

10.4 Discussion

Parents who have children with WS consistently report concerns about their social behaviour and in particular their hypersociability, as reported elsewhere in this thesis (e.g. see Chapter 4). The aim of the current case study was to gain an in-depth insight into one adult with WS, from her perspective and from the perspective of her mother. Findings from the current study identified key themes relating to social behaviour and internet use. Hypersociability and a lack of stranger danger awareness were apparent across these themes. This is echoed in recent work by Riby et al. (2013), who used video vignettes to explore issues of stranger danger awareness in individuals with WS aged 8 – 17 years old. They found that, compared to their typically developing peers, many individuals with WS show a lack of awareness of the dangers of interacting with strangers. When examined alongside parental reports, it was suggested that stranger danger awareness was closely linked to everyday social behaviours. Whilst the participant in the current study was older than those involved in the work of Riby et al. (2013), the same issues are clearly still evident in early adulthood. Indeed it seems likely that Alaina's social behaviour is an important pre-cursor to her vulnerability.

Linked to Alaina's prosocial approach towards strangers was her definition of a friendship. Her broad, encompassing definition, included justifications which may be more typically associated with strangers. Previous research on friendship in individuals with developmental disorders has noted that what constitutes a friendship may be very different for these individuals (see Jobling, Moni & Nolan 2000; Freeman & Kasari, 2002). Indeed, adolescents with ASD have been shown to find describing the difference between a friend and an acquaintance challenging (Carrington, Templeton & Papinczak, 2003). However, the qualities, definitions and criteria for friendships in individuals with intellectual and developmental disorders remain poorly understood. When considering stranger danger safety training programmes, it seems important to consider definitions of friendship, in order to develop effective interventions. It is interesting to note, however, that Alaina's mother said that her daughter's definition of friendship was particularly resistant to change, demonstrating the emotion invested in these friendships, and the challenge facing intervention design.

Yet, based on the parental account, it is clear that Alaina experiences difficulties in identifying deceit. She bases her judgements on immediately identifiable perceptual information, such as facial expressions. Tager-Flusberg and Sullivan (2000) proposed that there are two distinct components of theory of mind: a social cognitive component and a social-perceptive component. Social cognitive theory of mind refers to the, "conceptual understanding of the mind as a representational system" (p. 61), that is, the ability to reason the content of other people's minds (Tager-Flusberg and Sullivan, 2000). This is strongly linked to other cognitive abilities as well as language abilities (Tager-Flusberg & Sullivan,

1994). In contrast, social-perceptive theory of mind focuses on person perception, for example, using immediately available perceptual information to make judgements on the intentions and emotional states of others. It has been suggested that social-perceptive theory of mind acts as a precursor to the development of more sophisticated social cognitive theory of mind (Baron-Cohen, 1994). As such, it could be seen that whilst Alaina show relatively preserved social-perceptive theory of mind, she has atypical or delayed development of social-cognitive theory of mind, which is why she struggles to identify lies and deceit. This is supported by Plesa-Skwerer and colleagues (2006; 2008) who have shown that indidivudals with WS demonstrate relative abilities in basic expression perception, but deficits in identifying more complex expressions.

As proposed by Lough et al. (2015), there was also evidence of an overlap between offline and online vulnerability. For example, Alaina's mother said, "*It's the same really. Except it's a bit more difficult online because it's there in the home isn't it, you can't lock the door and it's gone.*" Alaina reported using social networking sites daily, stating that she had over 1,000 friends. Her mother acknowledged that it was much harder to protect her from victimisation when she was online, as opposed to when she was out in the real world. She was especially worried about how open her daughter was online, and her susceptibility to grooming. The recent EU Kids Online survey (Livingstone et al., 2011) identified individuals with an intellectual difficulty as facing an elevated risk of victimisation when online, compared to their typically developing peers. Specifically, they experienced high contact risks, meaning they would be more likely to arrange a meeting with a stranger they had met online. Alaina's atypical

definitions of what a friend is and what a stranger is are likely to contribute to this elevated risk, heightening her level of vulnerability online.

It was interesting however, to hear that her mother believed that preventing internet access would only increase social vulnerability in the real world, as her daughter would seek her social interaction from elsewhere. Both Alaina and her mother said that the internet offered the opportunity to form proxy friendships. In typically developing individuals, use of social media has been found to increase social interactions, and decrease feelings of loneliness (Valkenburg & Peter 2007). For individuals with WS who seek social interactions (Frigerio et al., 2006), whilst simultaneously experiencing high levels of loneliness (Davies et al. 1998), it is clear why connecting with others via the internet is an attractive prospect. This may be especially true during adulthood when, according to parental anecdotes, we see the isolation and social withdrawal increase in WS. During this time, we also see an increase in social anxiety. If individuals with WS are withdrawing from social situations because of their heightened anxiety and their difficulties interpreting complex interactional nuances (Laws & Bishop, 2004), then the internet offers an alternative route for interactions. However, as Alaina gets older, her mother said that supervision, both offline and online, has become more challenging, as it is less clear who has the authority of Alaina's social behaviour.

The findings from the current study highlight the impact that social behaviour has on vulnerability, and also offer one of the first insights into computer use in an adult with WS. However, within-syndrome heterogeneity is widely reported in

the WS literature (e.g. Little et al., 2013), and as such, it is acknowledged that the themes generated from the interviews in the present study are unlikely to be representative of all adults with WS. They do, however, suggest that it is likely that at least some adults with WS are using the internet frequently, and are using social networking as a new medium of social interaction. The case study approach employed here however has been criticised for its epistemological and ontological basis. Researcher subjectivity, it has been argued, steers the analysis towards verification of previously held beliefs. However, Flyvbjerg (2006) argues that this is true of many methodologies, and that the case study approach actually contains a bias towards falsification of ideas, rather than verification. Despite the inherent limitations of the case study approach, it does offer the in-depth, exploratory methodology needed to begin investigating this novel field. Based on the findings in the current study, it is predicted that the characteristics that prelude social vulnerability in the real world, are also feeding vulnerability in the online environment. Further investigation of internet use and online vulnerability in WS is therefore needed.

Chapter Eleven: General Discussion

11.1 Introduction

This thesis has introduced the reader to important elements which contribute to the experience of social vulnerability in the developmental disorder Williams syndrome (WS). The first chapter reviewed the relevant literature on the cognitive and social profiles associated with WS (e.g. Jones et al., 2000), as well as the impact that within-syndrome heterogeneity and high levels of psychopathology could have on their behavioural profile (e.g. Porter & Coltheart, 2005; Porter et al., 2007; Little et al., 2013). These characteristics are of interest given the hypersociable behavioural phenotype already associated with WS in the literature (e.g. Frigerio et al., 2006; Jones et al., 2000). Methodological considerations were outlined in Chapter 2, where important issues such as the use of matched groups design and a static time point approach were discussed. Chapter 3 presented primary evidence of the social, anxiety, communication and vulnerability profiles associated with WS, where participants with WS performed significantly more atypically on the measures than an age-matched typically developing control group. It also highlighted the strong correlation between social behaviour, anxiety and vulnerability, which provided some context for the subsequent experimental work.

The first aim was to explore the types of social behaviour which could place children with WS at risk during social interactions. Therefore, the first part of the thesis (Chapters 4 to 6) focused on the events leading up to a social interaction, such as approach behaviour, interpersonal distance and trust. Previous studies

have relied heavily on ratings of faces as indicators of approachability, but have yielded contradictory findings (e.g Jones et al., 2000; Martens et al., 2009; Frigerio et al., 2006). Porter et al. (2007) and Little et al. (2013) proposed that there was in fact considerable variability in the social approach behaviour of individuals with WS. Chapter 4 used parental interview data to gain a rich, in-depth insight into how parents see their son/daughter's social approach behaviour. The results showed that 72% of parents reported that their child had severe impairments in reciprocal social functioning. Whilst these impairments improved with age, atypicalities of social motivation remained constant. Thematic analysis showed that there was variation in the personality traits and level of parental supervision, reinforcing the heterogeneous nature of social approach behaviour. However, the themes of naivety to danger, and lack of social boundaries were prominent throughout the accounts, which are likely to be a part of the high levels of social vulnerability experienced in WS (Jawaid et al., 2012).

The theme in Chapter 4 of lack of social boundaries was particularly interesting, with parents saying that their son/daughter has, "no concept of personal space". No previous research had addressed this issue in WS. Given the significance of personal space and interpersonal distance when considering successful social interactions (Gessaroli et al., 2013) and indeed levels of social vulnerability, Chapter 5 extended the social approach findings in the previous chapter to investigate violations of personal space. Previous work by Kennedy and Adolphs (2014) in children with ASD found that 79% of parents rated that their sons/daughters were more likely to violate the personal space of others compared to their TD siblings. Conversely, Gessaroli et al. (2013) found that children with

ASD maintained a greater interpersonal distance than their TD peers, when taking part in a stop-distance paradigm. The first study in Chapter 5 used the same method as Kennedy and Adolphs (2014), and assessed violations of personal space through parent reports on the Social Responsiveness Scale (SRS; Constantino & Gruber, 2005). The findings showed that individuals with WS were reported to be more likely than individuals with ASD and TD individuals to impede on personal space of other people. The second study in that chapter used the stop-distance paradigm, as used in Gessaroli et al. (2013), and showed that individuals with WS maintained a smaller interpersonal space than their age-matched TD peers. They also failed to regulate their personal space boundaries depending on the familiarity of the person they were interacting with; they stood as close to a familiar person as they did an unfamiliar person. Strangers can get strong cues from interpersonal distance abnormalities and we know that appropriate social distancing plays a key role in social interactions, and positive social interactions are protective against social vulnerability (Gessaroli et al., 2013). The atypical social distancing seen in WS, particularly when interacting with strangers, could therefore be heightening their levels of social vulnerability.

From Chapters 4 and 5, it was clear that many individuals with WS were likely to approach unfamiliar people with a lack of awareness of the danger this could pose, and they were also likely to invade the personal space of others, i.e. stand very close to them as they interact. It seemed imperative here to the story of social vulnerability in WS to find out whether individuals with WS were also more likely to trust people (e.g. Ng et al., 2015; Martens et al., 2012), compared to their chronologically age matched and mental age matched peers. Chapter 6

presented the findings from a behavioural economics trust game. It was found that, compared to their mental age matched peers, participants with WS were significantly more trusting of other people, although, contrary to prediction, they invested more tokens when their partner's face was not visible, compared to when it was. Interestingly, the participants with WS were not able to decipher the trustworthiness of faces during the game, although they were able to after the game had finished. This difference in performance during the game, compared to the trustworthiness ratings provided at the end of the game suggests that other aspects of the task are likely to be affecting their ability to decipher trustworthiness from faces, for example the cognitive demands of the tasks, or the social encounters in the game context. These findings suggest it is unclear how consistently individuals with WS can identify trustworthiness from faces which could be important when considering their social vulnerability status.

Chapters 4, 5 and 6 reinforce the notion that people with WS could be experiencing high levels of social vulnerability. However, they offer no insight into how these individuals perceive their own vulnerability, especially when considering that individuals with WS experience intellectual impairments (e.g. Searcy et al., 2004) and socio-cognitive impairments (e.g. Tager-Flusberg & Sullivan, 2000) that will impact upon their ability to self-reflect. The second aim of this thesis therefore was to investigate the level of insight that individuals with WS have into their own vulnerability using the Social Vulnerability Questionnaire (SVQ; Fisher et al., 2012). Given the substantial real world implications of this vulnerability (e.g. elevated lefts of theft, victimisation, abuse and bullying; Nettelbeck & Wilson, 2002; Fisher et al., 2013), researchers have

begun to consider the effectiveness of possible interventions. For example, Fisher (2013) trialled a stranger safety intervention with adults with WS (n=21), and found that adults with WS could learn to respond appropriately to lures from strangers, with 62% walking away from strangers. Self-insight has also been found to be a crucial element of intervention success in other clinical populations (Emmerson et al., 2009). Chapter 7 therefore investigated how individuals with WS saw their own vulnerability, and it how this compared to reports from their parent, taking a multi-informant insight into this issue (see the benefits of utilising a multi-informant approach in WS research in Klein-Tasman, Li-Barber & Magargee, 2011 and in Chapter 2). In Chapter 7 it was found that individuals with WS perceived their own levels of vulnerability to be lower than that reported by their parents, except on the subscale of emotional abuse, where the WS group reported experiencing higher levels of vulnerability than their parents reported. Although parent reports are undoubtedly valuable when working with a WS population, over-reliance on parent reports for future interventions may not be the most effective way of addressing social behaviour. Instead, this thesis advocates the use of multi-informant approaches when investigating social vulnerability.

The final aim was to offer the first investigations into online vulnerability in adults with WS. The remainder of the thesis therefore investigated social vulnerability in the online context. Chapter 8 reviewed the parallels between the risk factors for online vulnerability, and the WS behavioural phenotype, presenting the notion that the social vulnerability experienced by individuals with WS in their daily lives is likely to also be present in the online world. Chapter 9 therefore investigated internet use and online safety in a group of adults with WS.

It was found that 86% of the adults in the sample were using the internet daily, and 85% of those were frequent users of social networking sites. On an internet scenarios task, participants indicated that they would be more likely to engage in a socially risky behaviour (e.g. agreeing to meet with a stranger they have met online), compared to a risky behaviour that was non-social in nature (e.g. giving out passwords). To investigate this in more depth, a case study was presented in Chapter 10 with an adult with WS and her mother. This outlined how the portability and accessibility of the internet has provided a new platform for individuals with WS to engage with others, and fulfill their need for social interaction. A superficial understanding on what a friend was, and how to differentiate between a 'good stranger' and a 'bad stranger' offered some clarification on how adults with WS view social interactions. It therefore seems that social vulnerability is a wide, far-reaching concept that is closely tied into the socio-behavioural profile seen in WS, particularly their high levels of social motivation. It also has a considerable impact on the everyday lives of individuals with WS, and their families.

11.2 Theoretical implications

The theoretical context of social behaviour in WS was outlined in Chapter 1. It is important to emphasise that the thesis did not set out to explicitly test any theory of social behaviour in WS. However, the body of research presented here does contribute to our theoretical understanding of typical and atypical social behaviour by providing descriptive data which can be placed in the context of previous research which has investigated the neuroanatomical functioning of individuals with WS (e.g. Myer-Lindenberg et al., 2005; Mobbs et al., 2007; Haas

et al., 2009). It also highlights similarities and differences in social behaviour to more theoretically established IDD, such as ASD. The within and between syndrome differences discussed in this thesis offer important theoretical contributions to the understanding of social behaviour in WS. For example, the social motivation theory of autism (Chevallier et al., 2012) offers an exciting new theoretical approach when applied to WS.

One of the most prominent theoretical explanations in the WS field is the amygdala theory (e.g. Haas et al., 2009; Martens et al., 2009; Kennedy, Glascher, Tyszka & Adolphs, 2009). The amygdala theory suggests that individuals with WS have a large amygdala volume, and subsequent atypical amygdala functioning (Haas et al., 2009, Martens et al., 2009). They are likely to approach other people because of reduced activation of the amygdala in response to threatening stimuli (Martens et al., 2005). It is this muted reaction to negative affect that is used to explain the indiscriminate social approach behaviour seen in WS. The face stimuli used in the current thesis (e.g. Chapter 6) depicted neutral facial expressions, meaning it is difficult to comment on the validity of the amygdala hypothesis in explaining the extreme prosocial behaviour seen in WS. However, the amygdala hypothesis has also been used to explain personal space regulation. Kennedy, Gläscher, Tyszka & Adolphs (2009) found that a participant with complete bilateral amygdala lesions displayed no sense of personal space. In one of the trials, the participant approached the experimenter and standing nose-to-nose whilst maintaining eye contact, and rated the interpersonal distance as being "perfectly comfortable" on a 10-point Likert scale (1 = extremely comfortable, 10 = extremely uncomfortable). Similarly, when an unfamiliar male

confederate approached the participant, she was happy for him to stand abnormally close when engaging in conversation. Again, the participant reported that this distance felt "perfectly comfortable", whereas the confederate rated the distance as being very uncomfortable, by giving a score of 7 on the Likert scale. They also found that there was amygdala activation in TD individuals when they were in close personal proximity to another person. We know that individuals with WS have atypical amygdala functioning and a reduced response to threat (Haas et al., 2009). It could therefore be that this atypical amygdala functioning is not triggering the appropriate emotional reactions when personal space violations occur, leading to reduced interpersonal distancing in WS (Kennedy et al., 2009), particularly around unfamiliar people, as found in Chapter 5.

More relevant to the wider findings of the current thesis is the frontal lobe hypothesis. This suggests that the social behaviour seen in WS is the result of poor impulse control (Porter et al., 2007). Whilst these individuals may know not to approach strangers, they struggle to inhibit that behaviour. This was something that was repeated anecdotally by parents during testing for this thesis. Porter et al. (2007) highlighted the similarities between the social approach behaviour seen in WS and the social approach behaviour seen in patients with frontal lobe damage. For example, participants with WS showed difficulties with emotion recognition, did not perform atypically on the social approach task, and displayed impairments in response inhibition relative to their mental age. Although these links were not causal, these findings suggest that the tendency to approach strangers seen in WS is the result of low levels of response inhibition resulting from frontal lobe dysfunction. This notion of 'knowing' versus 'doing' was emphasised by a

parent in Chapter 4 who said about her daughter's social approach behaviour, "*I don't know, you can tell her until you're blue in the face but it's like it is inbuilt it's something that she can't stop*". This strengthens the idea that the behaviour she described is the result of difficulties with inhibitory control.

In the same chapter, findings from the Social Responsiveness Scale (SRS) showed that the most socially impaired individuals tended to be younger, except when it came to the domain of social motivation. Here, it was found that atypical social motivation does not reduce with age. It could be that a combination of high levels of social motivation combined with poor inhibitory control is contributing to the hypersociable behaviour documented throughout this thesis. This thesis emphasises a lack of 'typical' social behaviour throughout, and discounts the early view of intact social behaviour in WS and impaired social behaviour in ASD. Rather, as was shown in Chapter 5, social behaviour is impaired in both WS and ASD but in some similar and some different ways. Extreme atypical social motivation could be the defining characteristic of the social profile in WS and ASD, which would make it key to understanding social vulnerability in the wider developmental disabilities field.

Indeed, the application of the social motivation theory to WS offers an interesting framework to better understand social behaviour and social vulnerability in this population. Chevallier, Kohls, Troiani and Schultz (2012) present an integrated model of social motivation. At the behavioural level, social motivation manifests as prioritising items of social relevance, experiencing social interactions as rewarding and striving to maintain and enhance social relationships (Fletcher-

Watson, et al., 2008). From the work in the current thesis, we know that individuals with WS are drawn towards socially salient stimuli, and that they enjoy engaging in social interactions. Biologically, social motivation has been linked to several regions of the brain, including the amygdala and the prefrontal cortex, and the dense connections between them (Ghashghaei et al., 2007). The Social Motivation Theory therefore transcends both the amygdala hypotheses and the frontal lobe hypotheses which are prominent in the WS literature. Yet, the findings in Chapter 3 show that 39% of individuals with WS scored within the 'normal' range for social motivation on the Social Responsiveness Scale (SRS; Constantino & Gruber, 2005), which was much higher than any of the other sub-domains. Future research should investigate the relevance of this theory to social behaviour in WS, and consider to what extent it can help explain other deficits in WS (e.g. non-verbal abilities, high levels of anxiety) beyond social behaviour.

11.3 Strengths and limitations

This thesis has reinforced the notions of naivety to danger and lack of awareness of appropriate social boundaries in WS using mixed methods and multi-informant approaches. It has offered the first investigations into personal space regulation, self-insight, online behaviour and internet safety in WS, and has made a substantial contribution to our knowledge of social vulnerability in WS. However, there are some limitations which should be addressed. First, several of the chapters in the thesis do not include a comparison group, meaning conclusions on typicality of behaviour cannot be reached in these chapters. For example, in Chapters 9 and 10, it is not known how the levels of internet engagement, and the types of online activities, are similar / different to TD peers,

or indeed different to individuals with other developmental disorders, such as ASD or DS. Though, as outlined in Chapter 2, the inclusion of a TD control group here would likely produce floor effects when considering the online safety scenarios task (Chapter 9), given the mean age of the sample was 27.7 years old. Likewise, inclusion of a TD control group in Chapter 4, as the questions on social approach behaviour are unlikely to be relevant to parents of 10 year old children. This does however raise the question of the value of including a mental age matched TD control group. Although some of the strands of enquiry in this thesis are tailored to the WS profile specifically, other chapters, such as the work on personal space, have paved the way for subsequent studies to investigate these phenomenon in comparison to mental age matched controls. The domains on which these groups are matched (e.g. overall ability, verbal ability or non-verbal ability) will require careful consideration given the complex paradoxical cognitive profile associated with WS. Further, this static point approach employed throughout the thesis does not consider the impact of developmental change. Understanding of the developmental trajectory of social vulnerability, and indeed its related facets, will be essential to the development of any targeted interventions.

Secondly, although the findings throughout the thesis are explored in the context of social vulnerability, the majority of the chapters do not directly assess this phenomenon. This is because the only current measure available to assess social vulnerability in individuals with IDD is the parent report Social Vulnerability Questionnaire (SVQ; Fisher et al., 2012). Although this questionnaire provides some insight into social vulnerability, it has not been standardised or normed

using a TD population (or any other developmental disorders in large enough samples to look at specificity), and cut-off markers of severity are not available. Therefore the links between personal space, trust and online behaviour with social vulnerability remain indirect. Despite this, when these behaviours are considered alongside the wider WS profile, it is clear to see how elevated levels of social vulnerability are produced (Jawaid et al., 2012).

11.4 Future directions

There are several ways to extend the research presented in the current thesis, in order to enhance our understanding of social vulnerability in WS. The first consideration is the inclusion of comparison groups with different IDD. Fisher et al. (2013) found differences between the subscale predictors of social vulnerability for adults with WS, DS and ASD, suggesting that there are syndrome specific patterns of vulnerability. Based on the current thesis, it is unclear if the behaviour reported is a general feature of having an IDD, or if it is specific to WS. We know that individuals with IDD as a whole experience high levels of vulnerability and victimisation (Nettelbeck & Wilson, 2002), but the uniqueness of the patterns of behaviour seen in this thesis to WS requires further investigation. Inclusion of an ASD comparison group would be of particular benefit. Both groups are known to show atypical social behaviour, yet have opposing social profiles. It could be of interest to investigate the similarities and differences in online social motivation and social behaviours in these groups, as introduced in Chapter 8. Mazurek (2013) found that the most common reason for individuals with ASD using social networking sites is for ‘forming social

connections', suggesting that their social motivation may be more similar to individuals with WS than different.

Further exploration of the within-syndrome heterogeneity seen in WS and its implications for the social vulnerability of individuals with the disorder would advance our knowledge on the topic. Little et al. (2013) suggested that rather than WS being a homogenous syndrome, behavioural subgroups can be identified based on levels of response inhibition. Participants who had high levels of social approach behaviour, were also shown to have low levels of inhibitory control and vise-versa. The subgroups identified contained individuals of varying ages and IQ levels, which suggests that developmental variables may not be predicative of within-group heterogeneity. Future research should move beyond assessing group means, to look at data at the individual level, in order to increase identification of within-syndrome variability in social vulnerability. It would be particularly interesting to explore the characteristics of a sub-group of individuals who score low on the SVQ, to find out which characteristics might be linked to low levels of vulnerability.

Given the considerable real world implications of the social vulnerability reported in WS, future work in this area should look towards intervention development. The development of a programme of syndrome-specific social skills training sessions, centred around stranger danger awareness could be hugely beneficial for individuals with WS. The high levels of vulnerability reported in Chapter 3 in adolescents aged 8 – 16 years suggest that the pre-adolescent age range would benefit from an intervention, which could in turn prevent some of the more

serious cases of victimisation reported in adults who have more independence and opportunities to interact with strangers (Fisher, 2014). Indeed, incorporation of online social skills training would be beneficial in light of the findings in Chapters 9 and 10.

11.5 Conclusions

The current thesis makes a significant contribution to our knowledge of social vulnerability in WS. The findings from this research suggest that young people with WS show increased social approach behaviour; they are highly likely to approach other people, regardless of whether or not they are familiar, and are naïve to potential dangers. Following their approach, they are likely to violate the personal space of others, as they maintain a reduced interpersonal distance, especially around unfamiliar people. There is also evidence of inconsistencies in the abilities of individuals with WS to decipher trustworthiness from faces. At the same time, they lack insight into their own behaviour, and how it could be perceived by others. Online, adults with WS are frequent users of social networking, and they deem thousands of online people to be their 'friends'. Online vulnerability in adults with IDD is a relatively new direction for research, and more needs to be done to understand the transference of social vulnerability in the real world to the online world. Therefore, this thesis has provided a valuable insight into social vulnerability in WS, and has opened up several new avenues for future research.

Appendices

Appendix A: Chapter 4 interview schedule

Social approach behaviour interview schedule

To begin with, I would like to find out about _____level of interest in social interaction with other people in general. Some children really enjoy social interaction with others and actively seek out opportunities for this to happen, whereas other children do not show this level of interest.

How would you describe _____'s behaviour in this area? (Are they interested in social interaction? How do you know? What do they do?)

Does _____ show more interest in social interaction with certain people?

Does _____show more interest in social interaction with children or with adults? What makes you say this

Does _____show more interest in social interaction with familiar people or with unfamiliar people? What makes you say this?

Now, I would like you to think about how _____behaves around people they don't know (a stranger). Children vary in how confident they feel around people they don't know. Some children are very confident and will approach them without hesitation, whereas other children feel less confident and are quite cautious

Can you tell me a little bit about how _____ behaves around people he/she doesn't know? (How do they respond to strangers? What do they do?)

Do you think that the setting _____ affects how they behave around strangers? In what way? (e.g. is it the same at home/school?)

Does _____ seem more **confident** around strangers in familiar or unfamiliar settings? What makes you say this?

Do you think that _____**knows** that they shouldn't approach a stranger? What makes you say this?

To what extent do you think that _____**knows** that they shouldn't approach a stranger?

How **likely** it is that _____would approach a stranger? What makes you say this?

Could you describe an example in the last month when _____has approached a stranger? What happened? (Get specific detail) Including: What exactly happened before, during and after. Why do they think child approached stranger? How did parent respond? What did child do following parent's response?

How does the way _____behaves around strangers make you feel?

Sometimes parents report feeling **worried** about the way their child behaves around strangers. Do you ever feel worried about the way _____behaves around strangers? What makes you say this?

Sometimes parents report feeling **stressed** about the way their child behaves around strangers. Do you ever feel stressed about the way _____behaves around strangers? What makes you say this?

Sometimes parents report feeling **embarrassed** about the way their child behaves around strangers. Do you ever feel embarrassed about the way _____behaves around strangers? What makes you say this?

Does the way _____behaves around strangers have any impact upon family life? (Do you have to make any changes as a family to accommodate this behaviour?)

Does the way _____behaves around strangers ever make you feel that you need to be more protective of him/her?

Does the way _____ behaves around strangers ever cause you to avoid going to certain places?

Does the way _____ behaves around strangers make you feel that you need to prepare before going somewhere?

Now, I am interested in finding out about how much _____thinks about what they are doing in a social situation. I am also interested in how well you think they can stop themselves from doing something they know they shouldn't do in a social situation. Sometimes children can find this difficult and tend to behave without thinking about the potential consequences or risks. For example; they may say inappropriate things to other people, or look through someone's bag/possessions without asking.

Can you describe _____'s behaviour in this area? (e.g. does your child tend to behave without thinking? Do they often take risks?)

To what extent do you think that _____ **thinks** about what he/she is doing in a social situation? What makes you say this?

To what extent do you think that your child acts on impulse in a social situation? What makes you say this?

Can you describe a specific incident in the last month when your child has done something they know they shouldn't do in a social situation? (e.g. saying something inappropriate/looking through someone's bag) What happened?

Appendix B: Chapter 9 e-safety scenarios task

Appendix A: E-safety scenarios task

I want to talk to you about certain things that might happen online. I want you to choose the option that is closest to what you think you would do. There is no right or wrong answer and you will not get in trouble for any of the answers. We will not talk to your parents about your answers. We just want to know more about what you are doing online.

1 You have been talking to a friend online but you have never met them in real life, they ask you to put your video camera on, what do you do?

a) Tell them you don't want to

b) Say not now, but you will do it next time you talk

c) Say yes sounds like a great idea

2 You have met a really cool person online and you have been getting along really well. They say they want to date you and ask you to send a picture of yourself. What do you do?

a) Say no, you don't want to send a picture of yourself

b) Promise you will send them one tomorrow

c) Send them a nice picture of you

3 You met a new friend online named Alex. You like all of the same things and have a lot in common, but you have never met before in real life. Alex wants to meet up soon so you can do something fun together. What should you do?

a) Make plans to meet Alex as soon as possible

b) Tell Alex you're busy but would love to meet sometime soon

c) Say no, you don't think it's a good idea

4 You are playing an online game, and you are chatting to other people that are also playing the game. One person asks if you want to come over and play the game together at their house. You have only ever talked to them online. What do you do?

a) Ask them if they want to come over to yours instead

b) Tell them no thanks

c) Say yes you would love to

5 Someone tries to send you a picture online. You have never met this person before or talked to them online before. They say it is a picture of a friend that you know. What do you do?

a) Ignore them

b) Say sure, send the photo over

c) Start talking to them to find out more

6 Someone you have met online asks you for your e-mail password because they need a new password and want some good ideas. What do you do?

a) Give them your e-mail password, it's good to share things

b) Suggest some of the other passwords you use instead

c) Say no, it's private information

7 You've found a really cool competition online. They say if you enter now you could win a lot of money. It only costs $5, and they ask for your name, address and phone number. What do you do?

a) Pay the $5 but don't give them all of your details

b) Pay the $5 and give them all your details

c) Ignore it

8 Someone you don't know has e-mailed you to say you have won a competition. You don't remember entering any competition, but it says you have won $10,000! They ask you for your bank account number so they can put the money in your account. What do you do?

a) Ignore it, you didn't enter a competition

b) Write them back and ask what the competition was before giving them your bank account information

c) Give them your bank account information so you can get the $10,000

9 You have received a few friend requests on Facebook from people you don't know. What do you do?

a) Click accept, you could become friends

b) Click decline, you don't know who they are

c) Leave the friend requests and decide later

10 When you're online, a message pops up saying "Hi I'm Danny from England. I'm looking to make friends in America. Click here to send me an email". What do you do?

a) Send Danny an e-mail and forward the message to your friends so he can make lots of new friends

b) Ignore the message

c) Send Danny an e-mail, you could be his friend

11 You have been talking to someone online, but you have never met them in real life. They are flirting with you, but ask you to keep your relationship a secret because it will make it more exciting. Do you keep it a secret?

a) No, you don't think you should keep this kind of thing a secret

b) Yes you will keep it a secret

c) Say you will keep it a secret for a while but then you want to tell people

12 You have been on a social networking site that you know your parents don't like you using. They ask you if you have been on it recently. What do you say?

a) Say no you haven't been on it, they would be mad if they knew you had been on that site

b) Say yes you have been on it and talk to them about it

c) Avoid answering the question

Appendix C: Chapter 10 interview schedule

Interview schedule: Adult WS

SECTION ONE: SOCIAL PROFILE

Remember, you don't have to answer any questions that you don't want to and you can stop the interview at any point, without giving a reason. We are going to start by talking about your friends, and what you like to do when you see them.

1. To begin with, can you tell me about the friends you had at school? (Prompts: how many close friends they had, how often they saw them, how they formed the friendships, what they did together etc.)

2. Can you tell me about who your friends are now? (Prompts: how often they see them, what they do together)

3. When you meet someone for the first time (i.e. someone you don't know), what do you do?

We are now going to talk about secrets.

4. Do you think you are good at keeping secrets? (Prompts: can you give me an example?)

5. Can you think of a time when it would not be OK to keep a secret?

6. Can you tell when someone else is lying to you or trying to trick you? (Prompts: can you give me an example?)

7. How do you know if you can trust someone?

SECTION TWO: COMPUTER USE

Now, we are going to talk about using the internet.

8. How often do you go on the internet?

9. Why do you go on the internet? (Prompts: work, watch videos, go on Facebook.)

10. What is your favourite website and why?

11. What do you think the best thing about having the internet is?

12. What do you think the worst thing about the internet is?

13. Do you use websites where you can talk to other people, such as Facebook?

If yes….

14. Who do you talk to on <insert name of website>? (Prompts: how do you know them, have you met in real life, do you talk to people you have only met on the internet?)

15. Has anything bad ever happened to you on the internet? Can you tell me about it?

16. Do you think that your mum/dad worries about you when you're on the internet?

17. Finally, do your parents set any rules for you when you're on the internet? Do you follow these rules?

Thank you for answering these questions. If you want to ask any questions about the study, then I can answer them for you.

End of Interview

Interview schedule: Parent

SECTION ONE: GENERAL PROFILE

You are reminded that you do not have to answer any questions which you do not wish to. You can stop the interview at any point, without giving a reason. We are going to start with some general information on _____ and day-to-day living.

1. Can you please start by telling me a bit about _____, for example their age, their daily routine, their interests…?

2. What would you say are the main challenges _____ faces on a day-to-day basis?

3. How, if at all, have these challenges changed as your son/daughter has grown up?

4. Can you tell me about any particular strength that _____ has and how s/he uses them? (for example, social, behavioural, hobby)

5. How does _____ fit into the structure of your immediate family – for example do they have siblings? (if necessary probe the general family structure in terms of immediate family, involvement of grandparents, ages where relevant)

6. Does _____ have any other diagnoses? (for example of another learning difficulty or developmental disorder and / or of any mental health difficulties?)

SECTION TWO: SOCIAL PROFILE

I would now like to move on to talk about _____'s general social functioning. For this study we are especially interested in looking at social abilities. In this interview we are going to be thinking about _____'s abilities to interact with others, and to form and maintain relationships.

7. To begin, can you tell me about your son/daughter's peer social group during their school years? (for example, how many close friends they had, how often they saw them, how they formed the friendships, what they did together etc.)

8. Can you now tell me about your son/daughter's current peer social group?

- What would you say the differences were between their peer social group now, compared to when _____ was a teenager?

9. Tell me a little bit more about their interaction style with people who are familiar to him/her? Has there been any change as they have grown up?

10. Can you also tell me about _____'s interaction style with people who are unfamiliar to him/her? Has there been any change as they have grown up?

11. Can you talk me through how _____ would interact with an unfamiliar person they had just met for the first time?

We are going to go on to talk about some aspects of emotion and social behaviours, for example _____'s trust behaviour and awareness of deception.

12. Thinking about deception, can you tell me about _____'s ability to deceive others? Can you give me an example?

13. Can you also tell me about _____'s ability to recognise when someone else is lying? Can you give me an example?

14. How easy does _____ find it to trust other people? Can you give an example?

- What makes them more/less likely to trust someone?
- Do they ever change their mind about how trustworthy someone is?

15. Is there anything else that you feel is particularly important about the way in which _____ interacts with other people, for example whether they suffer from anxiety?

SECTION THREE: COMPUTER USE

Technology is becoming an integral aspect of the lives of young people, offering new ways to play, socialise and gain knowledge. For this section, I would like you to think about your son/daughter's computer usage, and in particular their use of the internet.

16. Can you tell me about your son/daughter's computer usage, for example how often they use it?

- Why do they use it?
- Do they use the internet?
- What are their favourite websites?

17. What do you worry about, if anything, when _____ is on the internet? Why does this make you worry?

18. Does _____ know that you're worried about this?

19. How aware is _____ of the potential dangers of the internet? (ask about cyber-bullying, online grooming, inappropriate content, identity fraud.)

20. How aware would you say you are of these risks?

21. Do you take any precautions or set any rules for _____'s internet use? If so, can you tell me about them and why you have set them?

22. Can you tell me about how easy or difficult you find it to monitor your son/daughter's internet use? Can you tell me why?

23. What do you think the benefits are of _____ using the internet? Can you give me some examples of how they have benefitted from having access to the internet?

24. Is there anything else that you feel is particularly important about _____'s computer and internet use that we have not already discussed?

SECTION FOUR: RISK AND RESILIENCE

Now that we have discussed _____'s offline and online world, we want you to think about their risk and resilience in these two environments. We will start by talking about the offline environment.

25. Can you tell me about how resilient you think _____ is? Can you tell me about a time when they coped well in a risky situation?

26. How vulnerable would you say your son/daughter is now? By vulnerable, we mean how 'at risk' would you say your son/daughter is currently? How vulnerable were they when they were younger?

27. Is there anything that you can pin-point that contributes to this vulnerability?

28. Can you give me an example of a time when they showed this vulnerability?

29. Can you give me an example of a time when they showed good resilience?

30. What strategies, if any, have you tried to promote resilience and reduce vulnerability? Were they effective?

Now we are going to talk about risk and resilience in the online environment, i.e. – while _____ is on the internet.

31. Can you tell me how resilient _____ is online? For example, how does s/he cope with upsetting information, how aware is s/he of potential dangers, does s/he know what to do if they are unhappy with anything that is said online?

32. Would you say the vulnerability experienced offline is the same or different to the vulnerability _____ faced online? What are the similarities? How are these environments different for _____?

33. What advice would you give to parents who are concerned about how vulnerable or at risk their son/daughter might be offline? Would this advice be the same if the vulnerability was online?

34. Finally, is there anything else you would like to add that you feel we haven't already covered?

Thank you for taking the time to participate in this research. If you have any questions for me I can try to answer them for you.

End of Interview

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