Sociability and Social Vulnerability in Williams Syndrome

ELLEN RIDLEY

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Abstract

Social vulnerability has been emphasised in relation to Williams syndrome (WS), but there is little evidence on its presentation in adolescence, relevance across neurodevelopmental groups or on the underlying factors. This thesis took a multi-methods approach to address these gaps. The first study (Chapter 3) reports findings of an interview study about adults' perceptions and experiences of friendships. The results raised important themes around social decision-making, the role of support networks, and negative peer experiences in school. The childhood/adolescent years emerged as important; therefore, the remainder of the thesis prioritised this developmental stage. Chapter 4 examined aspects of social behaviour relevant to social vulnerability and deemed salient in the WS social profile: prolonged eye contact and propensity to approach others. Using cross-syndrome design, it was established that qualities of eye contact and social interaction styles varied within WS and across neurodevelopmental groups (fragile X syndrome, attention deficit hyperactivity disorder and autism). The findings indicate that social phenotypes are best conceptualised as shared across groups and thus raise questions about the design of cross-group studies on social vulnerability. The final Chapters delineated the profile and correlates of social vulnerability in adolescence in WS. In Chapter 5 social vulnerability was evident at elevated levels and this was supported by parent qualitative data. The study also provided first evidence on family impact and highlighted strategies adopted by families to manage social vulnerability, including increased parental supervision. Extending these findings, Chapter 6 investigated several relevant but untested correlates of social vulnerability. Difficulties in executive functioning, adaptive behaviour and particularly social reciprocity (which showed a predictive role) were associated with greater social vulnerability. Parental protection behaviours increased in line with social vulnerability. Together the findings confirm the need to understand the underpinnings of social vulnerability and provide support at the individual and family level, early on in development in WS.

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Declaration

For Chapter 4 Paper 1, the data included in Study 1 was collected by another researcher as part of a separate study, not connected to the PhD. In addition, some of the Chapter 4 cross-syndrome data (Williams syndrome, attention deficit hyperactivity disorder, autism groups) were collected as part of my MA dissertation. The neurotypical data and fragile X syndrome data were collected as part of the PhD.

Chapter 4 includes two published journal articles. The contributions of each author are outlined below. All authors read and approved the final manuscripts.

Publication features	Author contribution
Ridley, E., Arnott, B., Riby, D. M., Burt, D. M., Hanley, M., & Leekam, S. R. (2022). The quality of everyday eye contact in Williams Syndrome: insights from cross-syndrome comparisons. *American Journal on Intellectual and Developmental Disabilities*, *127*(4), 293-312.	**Study 1 (WS)** **ER:** drafted the revised manuscript and led the peer-review process **BA:** data collection and analysis, drafted the original manuscript **DR:** conceptualisation, helped revise the manuscript **MB:** conceptualisation, provided comments on the manuscript **MH:** provided comments on the manuscript **SL:** conceptualisation, data analysis, drafted the original manuscript, helped revise the manuscript **Study 2 (cross-syndrome)** **ER:** conceptualisation, project administration, data collection, analysis and interpretation, drafted the manuscript and led on the peer review process **BA:** provided comments on the manuscript **DR:** conceptualisation, helped to draft the manuscript **MB:** provided comments on the manuscript **MH:** provided comments on the manuscript **SL:** conceptualisation, data analysis and interpretation, helped draft the manuscript

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ER: conceptualisation, project administration, data collection, analysis and interpretation, drafted the manuscript and led the peer-review process
DR: conceptualisation, participated in the design, coordination and interpretation of the data, helped draft the manuscript
SL: conceptualisation, participated in the design and interpretation of the data, helped draft the manuscript

Statement of Copyright[1]

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1. CHAPTER 1: SOCIAL VULNERABILITY IN WILLIAMS SYNDROME – INTRODUCTION AND LITERATURE REVIEW

1.1. Chapter Overview

The aim of this Chapter is to introduce the core themes of the thesis and synthesise key research. The Chapter is divided into three parts. First, in Part A, Williams syndrome (WS) is introduced as the neurodevelopmental condition of interest for the thesis. The literature on WS is reviewed, focusing on the cognitive, social and mental health features relevant to interpersonal interactions and the topic of social vulnerability. In Part B, the term 'social vulnerability' is introduced before outlining why social vulnerability is particularly relevant and important to understand in the context of individuals with intellectual and developmental disabilities. I argue that the common characteristics of WS map onto an existing theoretical framework of social vulnerability which, when considered together, make social vulnerability in WS a key area for investigation. Part B concludes by reviewing and summarising the evidence to date on social vulnerability in WS. Finally, Part C outlines the research rationale and aims for the current thesis.

PART A – INTRODUCTION TO WILLIAMS SYNDROME

1.2. General Overview

WS is a relatively rare neurodevelopmental condition, that has wide-ranging medical, cognitive and behavioural manifestations. It was first identified in the 1960s, when cardiologists Williams et al. (1961) and Beuren et al. (1962) independently described a group of patients with heart defects supravalular aortic stenosis (SVAS; a narrowing of the aorta), developmental delay and similar, distinctive facial features. The genetic basis of WS was reported three decades later when studies using linkage analysis found the deletion of the Elastin allele (ELN) on one copy of chromosome 7 (Ewart et al., 1993). This discovery kick-started a scientific fascination with WS and since then the WS genotype and phenotype has been studied extensively across many disciplines.

WS is caused by a micro deletion of genetic material, typically 26-28 protein-coding genes, on one copy of chromosome 7 (7q11.23; Ewart et al., 1993; Hillier et al., 2003). The genetic deletion occurs sporadically in most cases yet some familial cases have been documented (see Morris et al., 1993; Sadler et al., 1993; Scherer et al., 2005) and affects boys

and girls equally. A diagnosis of WS is often first queried by a clinician based on presenting clinical characteristics, including cardiovascular disease (most commonly SVAS), hypercalcemia, growth delay, delay in meeting developmental milestones in infancy (termed 'failure to thrive') and dysmorphic facial features. However, given the known variability from person to person, there is no single criteria needed to ascertain a diagnosis of WS. Presently, a diagnosis of WS can be confirmed genetically by fluorescent in situ hybridization (FISH), multiplex ligation-dependent probe amplification (MPLA), or chromosomal microarray analysis (CMA). The FISH test is most used to detect a deletion at the Elastin locus, as the Elastin gene is deleted in 95-97% of cases (Lowery et al., 1995; Palacios-Verdú et al., 2015). The aforementioned genetic discoveries and advances in genomic testing methods have helped to determine diagnosis at a younger age, which in turn has resulted in cases (e.g., in the UK) of prenatal diagnosis due to issues raised during pregnancy regarding heart abnormalities or physical growth issues. However diagnosis remains difficult in low-income countries and with diverse populations (Kruszka et al., 2018; Lumaka et al., 2016; Tekendo-Ngongang et al., 2014).

There has been limited investigation into the prevalence of WS. The most recent, and most cited, population study was carried out in Norway and reported a prevalence of 1 in 7,500 births (Stromme et al., 2002). This prevalence figure is higher than what has been reported in other information sources, for example the National Organisation of Rare Disorders (2008) states 10,000-20,000 in the USA and the Williams Syndrome Foundation UK (2023b) endorses a prevalence of 1 in 18,000.

WS has an impact on multiple organs throughout the body and many of the medical and physical issues are linked to the reduction in Elastin. For a recent and comprehensive account of the aetiology, diagnosis and common medical features associated with WS see Kozel et al. (2021). Heart issues and joint and musco-skeletal problems are common, which in turn impact on coordination, strength and motor skills. Other clinical characteristics include infant hypercalcaemia (increased calcium levels), short stature, and vision problems such as strabismus and amblyopia (Atkinson et al., 2001; Kapp et al., 1995; Winter et al., 1996) and may impact on wider visuo-spatial skills (Atkinson et al., 2001).

While much of the early enquiry on WS focused on describing medical characteristics, focus quickly shifted to the intriguing cognitive and psychological profile. It is the cognitive and behavioural manifestations that are of particular interest to the current

thesis on social vulnerability, therefore the remainder of Part A summarises the literature on the WS behavioural phenotype. A behavioural phenotype refers to the observable features of a condition, and is defined as the increased likelihood that individuals with a certain diagnosis will show behavioural features or characteristics compared to individuals without the diagnosis (Dykens, 1995). In other words, people with WS are likely to share cognitive and behavioural features more so than people who do not have WS, but this is not to say that WS is expressed in the same way for everyone. Rather, studies have documented vast heterogeneity across many aspects of cognition and behaviour in WS (Porter & Coltheart, 2005). The literature summarised below therefore conveys the 'average person' with WS, while acknowledging there are vast individual differences which mean that every person with a diagnosis of WS is unique and will experience different strengths and challenges across these domains.

1.3. Cognitive and Neuropsychological Characteristics

1.3.1. Intellectual Ability

The vast majority of people with WS have a degree of intellectual disability (ID), typically in the range of 'borderline' to 'moderate' (IQ 30-70; Martens et al., 2008). Current evidence indicates that IQ remains relatively stable throughout development in WS, however the evidence on cognitive trajectories is limited (Fisher, Lense, et al., 2016a; Mervis et al., 2012; Mervis & Pitts, 2015; Porter & Dodd, 2011). In fact, overall IQ is not particularly informative in the case of WS, as it masks an intriguing and uneven profile of cognitive strengths and challenges. WS is associated with a discrepancy in cognitive abilities, particularly that of linguistic ability compared to visuospatial skills, memory and numerical concepts. Early investigation into the WS cognitive profile interpreted the discrepancy as a "remarkable juxtaposition of impaired and intact mental capacities" (Rossen et al., 1996, p. 367) with the emergence of language as a strength in contrast to spatial cited as evidence in favour of the modularity account of language (Fodor, 1983) and a specific language module which is "intact" or "preserved" in WS (Bellugi, 1988). This view has since been challenged in light of a transformational shift in our understanding of development. Today, it is widely accepted that developmental outcomes (e.g. performance on a test of cognition) result from complex interactions between genes, environment, brain and behaviour, and that brain areas/functions do not develop in isolation of the rest of the brain (Karmiloff-Smith, 2012; Karmiloff-Smith, Scerif, & Ansari, 2003; Thomas, 2003).

The complexity of the WS cognitive profile is acknowledged and has been illustrated from the findings of studies taking a developmental trajectory approach. They indicate that the WS cognitive profile is not static, looks different at different ages and is underlined by atypical developmental processes (Jarrold et al., 1998, 2001; Sauna-aho et al., 2019). For example, Jarrold et al. (1998) reported evidence of a positive, linear relationship between verbal ability and the size of the verbal/non-verbal discrepancy first in cross-sectional dataset and then longitudinally (2001), indicating that verbal and nonverbal abilities develop at different rates, with the well-documented verbal/non-verbal dissociation emerging as a process of development. There is preliminary evidence that in WS cognitive abilities decline earlier than in neurotypical development, but at different rates with verbal abilities declining in advance of full scale IQ and performance IQ (Sauna-aho et al., 2019).

The complexity of cognition in WS is further evidenced by the fact there is huge variability within and between people with WS, with some individuals scoring low on assessments of language and others scoring in the normal range on visuospatial tasks (Porter & Coltheart, 2005; Stojanovik et al., 2006). Hence, a more accurate depiction of cognition in WS is to say that language ability (and nonverbal reasoning skills) is a strength *relative* to overall level of intellectual ability, whereas visuospatial skill is a considerable challenge *relative* to overall ability. Like all aspects of the WS profile, this is not the case for everyone with WS, but on average there is a higher likelihood that this cognitive profile will be present compared to those who do not have WS.

1.3.2. Social use of Language

Although language is considered a relative strength in WS, there is still a complex profile of strengths and weaknesses within this domain. In the context of the current thesis, it is the social use of language that is most relevant to the topic of social vulnerability. It is true that many people with WS are loquacious and often use mature, engaging language, which has been termed “cocktail party speech” (Udwin & Yule, 1990). However, this command of language coincides with various challenges in the subtle components of linguistics, particularly pragmatic aspects of language (Godbee & Porter, 2013; Laws & Bishop, 2004; Stojanovik et al., 2001). Pragmatics refers to the subtle rules which guide our understanding and use of language within a social context and competence in this area is important for effective communication in everyday interpersonal situations (Berk-Gleason, 2005). There is consistent evidence using multiple methods, of difficulties with pragmatics in WS. Parent and

teacher ratings on communication checklists highlight issues in areas such as inappropriate initiation of conversation and use of stereotyped conversation (e.g. Laws & Bishop, 2004; Philofsky et al., 2007). A study using the Child Communication Checklist (CCC; D. V. Bishop, 1998) to capture the pragmatic profile of nineteen children and young adults with WS (6-25 years) found that 79% of the sample met the cut-off for pragmatic language difficulties. In addition, the group scored lower than neurotypical peers across all five subscales of the CCC and the WS profile was distinguished from that of individuals with Specific Language Impairment (now referred to as Developmental Language Disorder; DLD) and Down syndrome by greater problems with inappropriate initiation of conversation and the use of the stereotyped conversations (Laws & Bishop, 2004)

This pattern of pragmatic language difficulties is supported by research analysing the speech of children with WS (Stojanovik et al., 2001). Udwin and Yule (1990) studied the conversations of school-age children with WS during conversations with an adult researcher. Over one third of the children in the WS sample were described as hyper-verbal, with their speech including overuse of stereotyped phrases, idioms, perseverative responding and a pattern of announcing unrelated personal experiences. Although the quantity or grammatical complexity of the speech did not significantly differ to that produced from a sample of children with ID matched on verbal IQ, the WS sample used more idioms, social phrases and fillers and exhibited a more overfamiliar manner.

Other studies of language profiles in WS have included a comparison group of individuals with DLD as this group tend have specific difficulties with language structure, but rarely have primary difficulties in pragmatics. Stojanovik (2006) reported that compared to children with DLD matched on receptive language ability, the utterances of children with WS during semi-structured conversations tended to include little exchange of information (i.e., significantly fewer continuations compared to both DLD and neurotypical) and inadequate information when responding to requests for further information and clarification. The author labelled the speech as ‘parasitic’ (p.167) to describe the reliance on the conversational partner’s contributions. The findings suggest that children and young people (CYP) with WS have difficulties with the reciprocal nature of a conversation and engaging in behaviours that help to achieve conversational flow. Research has also found that the narratives of children with WS contain a greater proportion of social evaluative language compared to neurotypical children and children with DLD, which has been described as ‘audience hookers’ and linked to the highly sociable nature associated with WS (Reilly et al., 2004).

It has also been reported that people with WS show difficulties with other aspects of pragmatics, including engaging in figurative language in the form of metaphorical expression, idiom and clichés, yet doing so in an appropriate manner or context (Udwin & Yule, 1990), difficulties comprehending the underlying meaning (Karmiloff-Smith et al., 1995; Sullivan et al., 2003) and finding social nuances, particularly non-literal humour such as sarcasm and irony, difficult to follow (Godbee & Porter, 2013). In the context of interactions with others, relatively good expressive language including the use of 'figures of speech', may signal an overestimated level of understanding that does reflect some of the wider challenges. It is important to keep the language profile in mind as other aspects of WS are discussed in the remainder of this section, especially when considering the broader social phenotype.

1.3.3. Executive Functions (EF)

In addition to the presence of mild-moderate ID, many people with WS also show challenges in specific neuropsychological areas which are relevant when thinking about social interaction behaviours and will become more relevant later in the thesis. EF is an umbrella term which encompasses higher-level cognitive skills that allow for goal-directed behaviour, including planning, working memory, self-regulation and inhibition (Diamond, 2013). Various studies have reported that individuals with WS have poorer performance when compared to matched NT individuals or normative scores, across a range of EF domains including inhibition (Carney et al., 2013; J. Greer et al., 2013; Mobbs et al., 2007), planning (Costanzo et al., 2013), attentional shifting (Menghini et al., 2010) and working memory (Rhodes et al., 2010). Studies using standardised parent-report measures of EF, such as the Behavior Rating Inventory of Executive Function (e.g. Gioia et al., 2015), indicate that significant difficulties in EF occur across the developmental spectrum in WS (Camp et al., 2016; Greiner de Magalhães et al., 2022; Hocking et al., 2015). Studies measuring EF using behavioural assessment batteries with individuals with WS lend further support (Rhodes et al., 2010).

Attentional difficulties in WS have been documented from a very young age and research using behavioural tasks has provided evidence of executive dysfunction comparable to what is seen in individuals with ADHD; a neurodevelopmental condition characterised by persistent inattention, hyperactivity and impulsivity (Rhodes et al., 2011). With respect to attention, several studies report the presence of ADHD symptoms in WS samples (e.g.

Carrasco et al., 2005; Leyfer et al., 2006). For example, Leyfer (2006) found that 64.7% of a large sample of children met the Diagnostic and Statistical Manual of Mental Disorders-IV (DSM-IV) criteria for ADHD, which was the most common co-occurring psychiatric diagnosis. Another EF domain that has received particular attention in the case of WS is inhibition (also termed 'response inhibition' or 'inhibitory control') which refers to the ability to control human functions such as emotions, behaviours and attention. During response inhibition tasks, individuals with WS make more errors compared to controls (Menghini et al., 2010). Difficulties in areas of inhibition and attention have been linked to broader aspects of the WS behavioural and social profile, such as the strong propensity to seek out interactions with others including strangers, prolonged gaze to faces and the eye region (Little et al., 2013; Porter et al., 2007) and development of anxiety (Ng-Cordell et al., 2018).

1.4. Social Profile

Many neurodevelopmental conditions are associated with interesting social profiles and there is a particularly striking social profile connected to WS, which has captured the attention of researchers since WS was first described. The term 'social profile' is an umbrella term which encompasses the various social components at play when navigating the social world (Cook & Oliver, 2011). While there is vast variation in the use of terminology and definition of key concepts in this area, Kennedy and Adolphs (2012, p. 559) point out the various "levels of social", which encompasses three broad components (1) *Social behaviour: "*the readily observable inter actions between an individual and other people" (2012, p. 559), (2) *Social cognition:* "the various psychological processes (both conscious and nonconscious) that underlie social behaviour" (2012, p. 560) and (3) *Social functioning:* "the long-term, contextualised ability of an individual to interact with others" (2012, p. 560). The following section outlines the profile associated with WS across the said "levels of social".

1.4.1. Social Behaviour – Heightened Interest in and Approach to Others

Very early reports on WS noted that people with WS "all have the same kind of friendly nature – they love everyone, are loved by everyone and are very charming" (Beuren et al., 1962, p. 1235). People with WS have been described to be "warm", "gregarious", "loving" and "extremely friendly", and it is this unique social disposition that is most often depicted in reports about WS (Gosch & Pankau, 1994; Jarvinen et al., 2013; Klein-Tasman & Mervis, 2003). Over the past three decades of WS research, there has been an accumulation of evidence to support a distinct social profile, which includes overfriendliness, elevated

social drive, high empathy and a tense quality (Jarvinen et al., 2013; Järvinen-Pasley et al., 2008). The findings from research comparing WS with other groups with developmental delay, indicates that this social disposition is somewhat unique to WS (Gosch & Pankau, 1994). For example, Klein-Tasman and Mervis (2003) aimed to capture the personality profile of children with WS and reported that the attributes which best distinguished children with WS (from those with comparable levels of ID of mixed aetiology) included a lack of shyness, high empathy and approach, together with personality traits 'gregarious', 'people-oriented', 'sensitive' and 'tense'.

A hallmark of social interactions is being motivated to seek out contact with others and a heightened social drive is the most salient feature of the WS social profile (often termed 'hypersociability' (Doyle et al., 2004a; W. Jones et al., 2000). Findings from observations of young infants and children with WS across a range of real-life settings suggest that a heightened interest in people manifests early on in development, including a heightened interest in faces (Laing et al., 2002). In a case study of a 10-month old with WS, Mervis et al. (2003) noted twice as much looking behaviour in play sessions with the mother and a stranger compared to mental-age matched children and "intense looking behaviour" towards a stranger was evident by the infant with WS but not a pattern shown by the controls. This description of looking behaviour was also reported when infants with WS (8-43 months old) were observed during their appointments with a geneticist (Mervis et al., 2003). Similarly, children aged 15-58 months who took part in research sessions were reported to look excessively towards the researcher's face and often at the expense of carrying out the task (Jones et al., 2000). These real-world observational findings were taken as evidence of heightened social interest and have since inspired a body of empirical research on social attention and social perception in WS (detailed in next subsection).

The manifestation of hyper sociability in WS has been studied in many ways, across observational and experimental designs, and is borne out in parent reports. The instinctive nature of wanting to approach other people is illustrated in parents' accounts of their child's behaviour: "... 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" (Lough, Rodgers, et al., 2016, p. 1103); "when she was little she liked walking and she would want to stop and talk to everybody coming in the opposite direction" (Riby et al., 2017, p. 186). Findings from questionnaire measures have also helped elucidate profiles of social motivation and social approach in WS. The Salk Institute Sociability Questionnaire (SISQ; see Zitzer-Comfort et al., 2007 for psychometric properties

of the SISQ) was developed by Bellugi and colleagues to capture dimensions of real-life social behaviour specifically in WS, with items of the SISQ tapping into approach behaviours towards familiar and unfamiliar people, eagerness to please others, empathy and ability to remember faces and names. Studies using the SISQ have found that children with WS are rated significantly higher on Global Sociability compared to neurotypical individuals of comparable mental age (Doyle et al., 2004a; Järvinen-Pasley et al., 2010a; Zitzer-Comfort et al., 2007) and individuals with other forms of IDD (Järvinen-Pasley et al., 2010a; W. Jones et al., 2000). Doyle et al. (2004a) reported that even the youngest children with WS in their study showed elevated sociability and every age group within the WS sample showed significantly higher 'global sociability' and 'approach to strangers' compared to neurotypical peers. This pattern of results has been found cross-culturally (Zitzer-Comfort et al., 2007). Further work using the SISQ has identified an interesting motivational difference in the social affiliative behaviours in WS, indicating that increased social approach seen in many people with WS appears to be driven by a desire for social closeness (attributes of e.g. people-oriented, affectionate), whereas in neurotypical people it is related to a desire for social power (traits of persuasiveness and dominance etc Ng et al., 2014).

Experimental, lab-based research has shown that heightened social interest extends into increased social approach behaviours (SAB) towards others. The propensity to engage with and approach unfamiliar people has been documented from a young age in WS (Dodd et al., 2010; W. Jones et al., 2000). Dodd et al. (2010) studied the approach behaviours of pre-schoolers with WS (aged 3-6 years) in the context of social/non-social play scenarios. They reported that the children with WS were more likely to engage with a stranger compared to chronological- and mental-age matched peers, and unlike the neurotypical children who only showed engagement with strangers when toys were present, the children with WS did not show this pattern and approached the stranger simply on sight. The increased approach behaviour towards strangers was evident even when the strangers face was not visible, which the authors interpreted as evidence that heightened social drive is not driven exclusively by the salience of faces (termed the social salience hypothesis; Frigerio et al., 2006).

A body of experimental research has probed hypersociability and SAB by asking individuals with WS to make approach ratings to facial stimuli (Frigerio et al., 2006; Järvinen-Pasley et al., 2010a; W. Jones et al., 2000; Martens et al., 2009). A seminal study by Jones and colleagues (2000) first provided evidence using this paradigm, known as the Adolphs approachability task, where participants view photographs of faces that have been

pre-rated as approachable / unapproachable and are asked to rate how much they would like to approach and talk to the person. Jones et al. (2000) found that adults with WS (mean age 23 years) rated both sets of photographs as more approachable compared to chronological age- and mental age-matched peers, indicating an indiscriminate SAB in WS. While some studies using this task have found a similar pattern of results (e.g. Martens et al., 2009), other studies that have manipulated the facial stimuli and involve emotion processing have revealed a more complex picture (Frigerio et al., 2006; Porter et al., 2007). Further explanation will be given later in Part A when the literature on emotion processing is addressed.

A heightened interest in others and increased desire to approach presents alongside difficulties judging social situations and modulating behaviour once engaged in an interaction. One example is the regulation of interpersonal distance (IPD) – the physical space people naturally keep between themselves and others, and a related concept, personal space (Gessaroli et al., 2013; Perry et al., 2016). For most people, the process of monitoring and regulating IPD is largely instinctive, with judgements of appropriate social space made without much conscious thought. However, issues of invading others' personal space boundaries have been documented in WS, as illustrated by the following parent quote (speaking about their 8-year old) "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" (Lough, Rodgers, et al., 2016, p. 1102). Lough et al. (2015) provided the first evidence that individuals with WS experience difficulties regulating IPD. In this study, the researchers examined one item in the Social Responsiveness Scale (Constantino & Gruber, 2012), 'knows when he or she is too close to someone or is invading someone's space' and found that people with WS were rated, on average, as significantly less aware of others' personal space, compared to a sample of autistic and neurotypical individuals. The study also provided tentative evidence of a lack of developmental change in the pattern in the WS group, whereas in the neurotypical group, children were rated as being significantly less aware of invading the space of others compared to adolescents.

Building upon this work, the findings from a study taking direct assessments of IPD regulation indicate that the interactions of people with WS are not modulated as would be expected by levels of familiarity. Lough et al. (2016) measured the IPD preference of 18 children and young people with WS (8-16 years old) using the stop-distance paradigm. A hip-to-hip measurement was taken under different conditions – when the partner in the interaction was familiar/unfamiliar and when the person with WS was being approached/doing the

approaching. The findings showed that the average preferred IPD of the WS sample when interacting with familiar partner was comparable to chronologically-age matched peers. However, a key difference emerged when the interaction partner was unfamiliar, whereby those with WS on average maintained a smaller IPD and this was true both when approaching and when being approached. In other words, the WS group exhibited a lack of discrimination in their IPD preferences. Irregularities of IPD using the same method have been reported in autistic individuals (Kennedy & Adolphs, 2014) and patients with amygdala damage (Kennedy et al., 2009). To summarise, increased approach to others presents alongside difficulties adapting and modulating behaviour in the moment, in response to changing contextual information.

While hyper sociability has become a defining feature of the WS profile and seems to be present for many individuals with WS, there is less evidence as to whether the intense drive for social interaction and approach to others seen in childhood extends across the lifespan in WS, due to a lack of adult data sets. However, Fisher et al (2014) provided tentative evidence that the degree of social drive may change with age in WS. They observed 30 young adults with WS (average age 26 years; range 16-50) at a community event and found that older individuals exhibited less indiscriminate SAB, were more likely to interact with familiar individuals (compared to strangers) and spent an average of 42% of the time not interacting with anyone. These findings may indicate developmental changes in levels of hypersociability, which may also link to reports of more inhibited temperament / behaviours in adulthood (Gosch & Pankau, 1997).

To date, three main theories have been put forward to account for the intriguing hyper sociability seen in WS. First—The Social Salience Hypothesis—proposes that social information, particularly faces, captures and holds the attention of individuals with WS quicker than non-social elements of a scene (Frigerio et al., 2006). Note that evidence on social perception in WS is outlined in the next section of the Chapter. The second hypothesis—the Frontal lobe Hypothesis—views the executive dysfunction present in WS (discussed earlier in Part A on cognition) to cause problems suppressing the impulse to approach others (Porter et al., 2007). The third hypothesis—amygdala theory—claims that atypicalities in the structure and function of the amygdala result cause a dialled-down reaction to emotional stimuli, particularly negative, threatening stimuli (Haas et al., 2009; Martens et al., 2009). It should be noted that the current thesis is not set up to specifically test these theories in relation to social vulnerability, though they are highly relevant to the topic.

The thesis will touch on elements on these theories later on (for instance Chapter 6 investigation of EF).

Taken together, the findings indicate that many people with WS show an excessively strong prosocial drive for interaction and closeness with others, which emerges from a young age. While these qualities can make people with WS very endearing to interact with, they mask a range of difficulties modulating behaviour (e.g., interpersonal space) once engaged in an interaction and understanding the nuances of social situations. In addition to poor modulation of approach behaviours, many people with WS experience a variety of social cognitive challenges, which when viewed in tandem with the sociability has implications for navigating social interactions and potential vulnerability.

1.4.2. Social Perception and Social Cognition

One aspect of social interaction is the motivation, desire or drive to interact with others, but successful social interaction relies on a variety of socio-cognitive skills. This includes attending to, interpreting and responding to social cues and contextual information. This section outlines the evidence on social attention and emotion perception in WS.

The assessment of social skill or social competence in WS has frequently been made using parent reports on the SRS (Constantino & Gruber, 2012). The SRS was designed to capture the presence and severity of social abilities and difficulties in autism but has been widely used with other neurodevelopmental groups to examine social profiles. A consistent finding across studies using the SRS is that very few parents of CYP with WS (fewer than 20%) report social functioning in the "normal" range when the questionnaire is analysed as a whole (global score) (13% in Klein-Tasman et al., 2011; 18% in Lough, Flynn, et al., 2016; 17% in Van der Fluit et al., 2012), and adults too (e.g. 19% in Ng-Cordell et al., 2018; 17% in Riby, Hanley, et al., 2014). Analysis at the SRS subscale level indicates that the most significant difficulties may fall within the domain of social cognition rather than social motivation (Klein-Tasman et al., 2011). However, it is clear from the evidence presented in the preceding section that such heightened social motivation is far from typical and researchers have proposed that the seemingly 'typical' levels of motivation can be explained by the SRS being designed to capture low levels of motivation (Riby et al., 2017).

Attending to and extracting meaning from faces is a critical cognitive component of social interactions. Observational evidence showing people with WS have an intense interest in faces (e.g. W. Jones et al., 2000; Mervis et al., 2003) inspired a body of research

investigating the profiles of social perception and social attention, using eye tracking. The findings of which support a bias for social information (T. A. Williams et al., 2013) and faces, particularly the eye region, in people with WS compared to both chronological and mental matched peers (Porter et al., 2010; Riby & Hancock, 2008, 2009b, 2009a). In an experiment using scenes which contained faces, Riby and Hancock (2009b) found that individuals with WS spent more time overall fixating on the faces and had longer average fixations. However, those with WS were not quicker in detecting faces compared to NT peers, suggesting that faces really *hold* attention for people with WS. This is supported by the finding that the presentation of faces in a visual search task provides no greater distraction than that shown by NT controls (Riby et al., 2011). These studies indicate that the "face fascination" in WS (Pavlova et al., 2016, p. 2) may be associated with a difficulty in disengaging from faces. Indeed, the issue of disengagement has been indicated by other studies showing a lack of habituation to faces (T. A. Williams et al., 2013).

On the whole, the eye tracking evidence maps onto descriptions of eye gaze from observational research and anecdotal reports and indicates that preference for social information, particularly faces, extends beyond childhood. However, the characterisation of over prolonged looking does not apply to all who have WS (Hanley et al., 2013; Kirk et al., 2013). For example, Kirk et al. (2013) did not find prolonged attention to the eye region in a sample of adolescents and adults with WS compared to chronological and mental-aged matched controls. In addition, they found a relationship between reduced attention to the face and eyes and increased levels of generalised anxiety, suggesting that the allocation of attention may interact with other aspects of the WS profile, particularly levels of anxiety.

Still, if people with WS are over attending to the face and eyes (overall), it raises the question of how this relates to wider social cognitive outcomes. Crucial evidence on this comes from a study conducted by Riby et al. (2013) where participants with WS were shown everyday social scenes that included actors directing their gaze to an object. In the first condition, gaze behaviour was measured when participants viewed the social scene with no instruction from the researcher. In the second condition, the same behaviour was measured but the participant was cued to detect a target (e.g., actor directing gaze towards one cup in an array of three). When viewing the scenes without instruction, participants with WS showed the expected over attending behaviour to the face and eyes of the actor, with less time spent looking at the target objects compared to NT peers. In the cued condition, participants with WS still spent more time than NT participants fixating on faces, however time spent looking

at the correct and plausible targets did increase. While the WS group did change their looking behaviour in response to instruction, they were less accurate in naming what the actor was looking at compared to the NT group. Therefore, the findings from studies exploring social attention in WS in conjunction with wider social cognitive outcomes indicate that over-attending to faces and eyes does not map onto the processing or interpretation of information in ways expected.

Faces provide a wealth of cues relevant for social communication; one of the most important being emotional state. The role of emotion perception has been studied in view of the heightened approach behaviour previously described. Frigerio (2006) modified the classical approachability task used in Jones et al. (2000) using face stimuli with standard facial expressions (happy, angry, disgusted, fearful, sad and neutral). When asked to make approach judgements, people with WS rated faces more approachable than neurotypical CA- and MA- matched comparison participants, but only when the face was showing a happy expression. In fact, participants with WS rated the non-happy faces (neutral, angry, disgusted, fearful and sad) more unapproachable than the NT group. These findings suggest that approach may not be entirely indiscriminate in WS and may be guided by facial emotion expression. Further findings by Porter et al. (2007) add support to that emotion recognition plays an important role, given the emotion cognition ability (of emotion happy, sad, angry, scared) were related to scores on the approach task for participants with WS.

It is important to note that the tasks used in Frigerio et al. (2006) and Porter et al. (2007) used very distinctive emotional expressions (e.g. happy, angry, sad), whereas in reality, faces hold much more subtle emotional expressions. A consistent finding across experimental studies of mental state recognition is that people with WS perform at a level congruent with their mental age (Gagliardi et al., 2003; Plesa-Skwerer et al., 2006; Porter et al., 2010), however negative or more complex emotions may be particularly difficult to discern. Plesa-Skwerer et al. (2009) found that individuals with WS were less accurate in identifying negative emotions when presented as emotional stimuli and voice tones compared to NT mental-aged matched controls. In a study of complex everyday mental states, Hanley et al. (2013) showed that the accuracy of mental state recognition in WS was contingent on the specific emotion portrayed. Specifically, when asked to make judgements of emotion from static and dynamic faces, participants with WS performed at a level comparable to chronological-aged controls when discerning the mental stages ‘deciding’, ‘not sure’ and ‘worried’. In contrast, the emotional states ‘relieved’ and ‘don’t trust’ were particularly

difficult; these states were recognised at a level significantly lower than verbal mental age. Difficulty in discriminating trust is an interesting and important finding because, in the context of interactions with strangers, an assessment of trustworthiness is made solely on facial cues, which in turn guides decisions about whether to approach or avoid others.

In fact, particular difficulties in deciphering trustworthiness have been emphasised in the WS literature. Martens et al. (2012) examined trustworthiness evaluations when viewing computer generated faces which had been pre-rated on trustworthiness. The study used mouse-tracking technology to provide insights into the decision-making process when viewing the faces and making approachability ratings. Relative to neurotypical CA-matched individuals, participants with WS were more likely to approach both the trustworthy and untrustworthy faces and were also significantly more likely to contemplate approaching untrustworthy faces even if they ultimately decided to avoid. This finding indicates that making judgements of trustworthiness may be particularly difficult for people with WS, and this may influence elevated levels of social approach. Yet, other studies have revealed a more nuanced profile of trust evaluations. When making an assessment of the trustworthiness of unfamiliar people (viewing neutral faces) in conjunction with a short scenario describing the character's intentions, adults with WS (mean age 35 years old) show a similar pattern of results to NT participants – they rate characters who lie to avoid hurting another person's feelings more positively than characters who lie to escape trouble (Ng et al., 2015). The authors interpreted this as evidence that people with WS do not show indiscriminate trust (as had previously been reported by Martens et al., 2012) when they have access to contextual information about deception (rather than simply making evaluations from facial appearance). Yet, Ng et al. (2015) also found that the WS group performed at chance level when identifying the intent /meaning of the lies.

1.4.3. Social Functioning – Real World Social Skills in Practice

The evidence reviewed so far across levels of social behaviour and social cognition, indicate that people with WS show a great deal of interest in other people, are highly motivated to seek out social contact with others, but struggle to adapt behaviour in view of social boundaries and contextual factors, and show differences in social attention and making effective use of emotional cues. Much of the findings discussed so far have stemmed from experimental tasks in laboratory settings, but how does this paradoxical social profile

translate in everyday real-life social interactions and what is the impact on developing friendships?

The real-life implications have been emphasised in the few studies that have examined interactions with strangers and awareness of stranger danger. Fisher et al. (2014) observed the behaviours of a group of young adults in community settings (prior to participants undertaking stranger safety training programme). Confederate strangers approached the participants with WS and presented a lure that was either *general* (e.g., would you like to go on a walk around campus with me?), *incentive* (I will buy you an ice cream if you come with me') or *assistance* ('can you help me carry this to my car?'). All but three participants did not show stranger safety skills at the level of saying "no" to the stranger and walking away and 38% of the sample (8/21 participants) agreed to go with the stranger. A lack of awareness of the potential dangers of interacting with strangers is echoed in the accounts given by people with WS when explicitly asked about decision making with strangers. For instance, Riby et al. (2014) examined stranger danger awareness in 16 individuals with WS (mean age 12 years, range 8-17 years) using two video vignette tasks which showed unfamiliar adults interacting with children. After viewing the videos, participants were asked questions about how the child in the video should respond and what they themselves would do if they were in that situation. The responses of participants with WS included significantly less appropriate answers compared to a group of children matched on verbal ability. Specifically, only 27% of the WS sample (compared to 60% of the neurotypical sample) showed appropriate knowledge and awareness of the potential dangers of interacting with unfamiliar adults. The authors reported no relationship between age and stranger danger awareness, indicating that knowledge of stranger danger does not increase with age in childhood, but a "trend towards significance" with IQ.

In terms of building friendships, there is evidence that many people with WS encounter difficulties in developing and sustaining relationships (Davies et al., 1998), which is present in childhood (Gillooly et al., 2021, 2022) and extends into high levels of social isolation by adulthood (Davies et al., 1998). In a study of 108 adults with WS, 30% endorsed difficulty with forming friendships and only half were involved in social activities outside of the home (Elison et al., 2010). Davies et al. (1998) found that most parents of adults with WS reported that their son/daughter engaged in "intense chatter" (83%), asked inappropriate questions or made inappropriate comments when meeting someone (67%) and the overwhelming majority reported problems establishing friendships (96%). However, findings

from more recent research asking adults to self-report on their friendships, portrays a more optimistic view of friendship in WS. Fisher et al. (2020) found that 98% of adults able to name a best friend and while most of the adults did not have a friend who lived close-by, they reported high-quality friendship features (e.g., emotional support) and did not report loneliness.

The implications for interpersonal relationships may become particularly noteworthy when individuals with WS reach adolescence and early adulthood, given this point in development is associated with increased independence and ever more complex social dynamics and expectations (Locke et al., 2010). In a study of outcomes of 19-39 years old, 30% of the sample had some form of daytime occupation (e.g. employed work, voluntary work, or part-time work placements)(Davies et al., 1997). In half of these cases, supervisors indicated that the person with WS was experiencing challenges in their role which "threatened their employment", such as over-friendliness (100%), anxiety (90%) and excessive or inappropriate chatter (86%). Since these findings were published in 1997 there have been huge societal shifts towards the inclusion of people with IDD in the community, hence this evidence is now outdated and there is a timely need for new research on adult outcomes.

The evidence across the 'levels of social' emphasises a striking paradox: people with WS have a strong desire for emotional closeness and connection with others, yet some of the other characteristics associated with the condition, particularly around judging social situations and modulating behaviour, makes navigating interactions with others challenging, and has a real-life impact on friendships and interactions with strangers.

1.5. Mental Health Characteristics

The socially driven personality profile outlined previously coincides with a complex pattern of emotional issues. Anxiety in particular is the most common mental health difficulty in WS and has wide-ranging impact for individuals with WS, as well as the wider family (Dodd et al., 2009; Leyfer et al., 2006, 2009; Royston et al., 2021; Stinton et al., 2010). Within the WS literature, prevalence rates for clinically-significant anxiety (i.e., levels of anxiety that meet diagnostic threshold for a formal diagnosis) vary significantly – from 16% to 84% (Stinton et al., 2010; Woodruff-Borden et al., 2010), however a more recent meta-analysis estimates the prevalence at 48% (Royston et al., 2017). The prevalence estimates are higher than rates seen in samples of neurotypical people (Somers et al., 2006) and while

anxiety is common across multiple genetic syndromes associated with ID, the rates in WS are higher than those reported in Down syndrome, fragile X syndrome and Prader-Willi syndrome (Dimitropoulos et al., 2009; Pegoraro et al., 2014) as well as conditions with mixed aetiology (Reardon et al., 2015).

Findings from a number of studies indicate that individuals with WS experience a range of anxiety disorders, but specific phobia and generalised anxiety disorder (GAD) are most prevalent (Cherniske et al., 2004; Dodd et al., 2009; Dykens, 2003; Leyfer et al., 2006). Indeed, across the 16 studies included in Royston and colleagues' meta-analysis (2017), specific phobia was identified as most prevalent at 36%, followed by GAD at 10% - both significantly higher rates than seen in heterogenous ID. Other genetic syndromes associated with ID vary in the type of anxiety; for instance, fragile X syndrome and Turner syndrome are associated with social anxiety (Cordeiro et al., 2011) and Cornelia de Lange syndrome is associated with separation anxiety (Crawford et al., 2017). Less common anxiety disorders in WS are separation anxiety, social anxiety, panic, post-traumatic stress, agoraphobia, and obsessive-compulsive disorder (Royston et al. 2017). A recent study took a formulation approach to understand the phenomenology of anxiety in adolescents and adults with WS and reported that the triggers or antecedents of anxiety included (in order of most common) uncertainty (e.g. unpredictability and routine changes), specific phobia (e.g. aversive settings), physical (e.g. health worries, lack of sleep, sensory sensitivities), social (e.g. welfare of others, negative emotions in others and separation anxiety), and everyday events (Royston et al. 2021).

Evidence indicates that anxiety often develops early in childhood in WS. For example, 11 of the 13 parents of individuals with WS in Royston et al. (2021) reported anxiety onset before the age of 12 years. While anxiety may develop in childhood, there is consistent evidence to indicate that it endures throughout development and may even increase with age in WS (Dodd et al., 2009; Einfeld et al., 2001; Ng-Cordell et al., 2018). Leyfer et al. (2006) used the Anxiety Disorder Interview Schedule in a sample of 119 children with WS (4-16 years old) and found that rates of GAD were higher among older children (11-16 years old) than would be predicted based on rates in younger children, suggesting that anxiety may increase over time. Further support to the hypothesis that anxiety increases with age comes from the small number of studies employed the gold standard, longitudinal designs. Woodruff-Borden et al. (2010) followed children with WS over 5 years and found that 82% met the clinical criteria for an anxiety disorder at some stage and 62% maintained a chronic

course. This finding is supported by the high prevalence of clinically significant anxiety in research with adult samples (e.g. Cherniske et al., 2004).

Research has identified several mechanisms that may underlie high levels of anxiety seen in WS, but the relationships are not well understood and this is an area where further evidence is needed. One possible mechanism is 'intolerance of uncertainty' which refers to the "dispositional tendency to experience fear of the unknown" (Bomyea et al., 2015, p. 90). Uncertainty is a common trigger of anxiety in WS (Royston et al., 2021) and is relevant in the context of common specific phobias seen in WS, such as noise; blood, injury and injection; storms; and animals (e.g. Stinton et al., 2012; Woodruff-Borden et al., 2010; Leyfer et al., 2006). Research has found that a higher intolerance of uncertainty is associated with increased anxiety in autism (Boulter et al., 2014; Wigham et al., 2015; Rodgers et al., 2016) and syndromes associated with ID (Jones et al., 2023; Crawford et al., 2023). Recent evidence has also provided support for this relationship in WS (South et al., 2021; Uljarevic et al., 2018). Another relevant transdiagnostic issue in the context of anxiety is sensory processing difficulties, which are high in WS (Glod et al. 2020) with the most well-documented being hyperacusis – "an abnormal sensitivity to sounds in which low- and medium-intensity sounds can cause discomfort or even pain" (Silva et al., 2021, p.1). In this sense, anxiety related to noise, or events and environments associated with loud noises, may relate to hyperacusis. In sum, the underlying factors driving high anxiety in WS are likely transdiagnostic features (e.g. intolerance of uncertainty and sensory sensitivities) but may also be related to phenotypic features of WS (e.g. hyperacusis).

A smaller number of studies have examined a broader range of mental health issues in people with WS. For example, Stinton et al. (2010) examined mental health issues in 92 adults with WS using a psychiatric interview developed specifically for adults with ID (Psychiatric Assessment Schedule for Adults with Developmental Disabilities – PAS-ADD). In addition to anxiety disorder (which was the most common mental health issue in the sample), diagnoses included depression, hypomania, schizophrenia and undifferentiated psychosis. Of the people with a psychiatric disorder, 41% had multiple diagnoses. The most common non-anxiety related diagnosis was depression (9%) and other studies have reported similar figures for depressive disorders 10-14% (e.g. Cherniske et al., 2004; Dodd & Porter, 2009), but this is still much lower than the prevalence rates documented for anxiety. Interestingly, Dodd and Porter (2009) found that this figure increased to 25% when the adult

sample was analysed independently, suggesting that depression may be more an issue later in life for people with WS.

1.6. Interim Summary

This Chapter so far has emphasised that WS is a complex neurodevelopmental condition. The review of the literature highlighted the multiple, interesting layers of the WS social profile. We see preference for social information, gaze towards faces, increased social drive including to strangers, engaging language and a very friendly and warm personality profile. This social profile presents in conjunction with the presence of ID, an uneven profile of cognitive abilities and heightened anxiety. The next part of the Chapter outlines why these common characteristics, when considered together, raise concerns about the safety and wellbeing of individuals in their everyday social interactions and relationships. Before outlining what is known about the topic of social vulnerability in WS, it is first important to delineate what is meant by the term social vulnerability.

PART B – INTRODUCTION TO SOCIAL VULNERABILITY IN WILLIAMS SYNDROME

1.7. The Concept of Vulnerability and Social Vulnerability

Vulnerability is a very broad term and can mean that a person is in danger, under threat, highly susceptible to problems, in need of support and/or helpless (Grundy, 2006; Simpson, 2006). There are many reasons why a person or group may be considered vulnerable, and as such, the term has many uses in both academic literature and modern society (K. Brown, 2011). The term vulnerability first appeared in the physical sciences, in reference to geographical location, environmental insecurity and exposure to natural disasters and hazards (Wisner & Luce, 1993). In the medical literature, vulnerability is connected to disease and illness. Within the social sciences—the perspective of the current thesis—vulnerability is linked to many different factors about the lives of individuals and groups of individuals (e.g. Jawaid et al., 2012; Pinsker et al., 2010). Yet, even within the social sciences there is a broad characterisation of the term and concept. The difficulty in defining the term was emphasised in 2017 by the Children's Commissioner for England at the time, who commissioned a report to uncover the number of vulnerable children in England. The foreword to their report states just how difficult a task this is because "the term 'vulnerable'

is used in so many different ways [….] that as soon as you think you've got a grip on it, it floats back out of view" (Children's Commissioner, 2017, p. 1).

One way vulnerability is often conceptualised is in terms of 'within-person' vulnerability (Brown et al., 2017). Within-person vulnerability refers to characteristics about an individual or cohorts that may increase the risk for poor outcomes or danger. It is commonly associated with stages of development, for example childhood and older age. This idea comes from the fact young children lack autonomy and depend on caregivers to meet their basic needs and to provide a level of support and social protection (Bagattini, 2019). At the other end of the developmental spectrum, older age is often associated with a gradual loss of autonomy and ability to take care of oneself due to cognitive decline (Murman, 2015). Vulnerability has also been linked to divergence from the 'typical' developmental processes in individuals with developmental disabilities (Fisher et al., 2012, 2016; Jawaid et al., 2012; Lough, Flynn, et al., 2015; Sofronoff et al., 2011). The concept of vulnerability in this sense assumes that an individual's abilities/disabilities (e.g., physical or psychological impairment, lack of autonomy) may place them at risk of an adverse outcome.

The labelling of groups of individuals as vulnerable has been criticised for neglecting the role of the *situation* (Luna, 2009; Luna & Vanderpoel, 2013). In reality, people can be vulnerable in multiple ways which arise from interactions between individual characteristics and the environment, context or situation experienced. In fact, the entwinement of person and situational factors when thinking about vulnerability is reflected in the updated terminology used in policies and procedures. For example, the term 'vulnerable adult' has been replaced with 'adults at risk' as per the Care Act (2014) and defined as 'any person who is aged 18 years or over and at risk of abuse or neglect because of their needs for care and support'. This shift reflects an attempt to move away from labelling individuals/groups as inherently vulnerable and instead acknowledge the circumstances surrounding the individual (Ann Craft Trust, 2022). This is echoed in the NSPCC Safeguarding Adults Policy and Procedure (NSPCC, 2019, p. 2) which highlights that individual characteristics may make vulnerability more of a concern, but it is the situation or environmental context which increases the potential risk of an adverse outcome. Vulnerability may be relevant for any individual at different times depending on the protective factors around them.

Social vulnerability, in particular, is the focus of the thesis. It has been defined broadly as the "the disadvantages faced by an individual while he or she endeavours to

survive as a productive member of society" (Jawaid et al., 2012, p. 335). Others state that social vulnerability "occurs when individuals are at risk of being unable to avoid adverse events that could affect their emotional, physical or financial well-being" (Fisher et al., 2016, p. 115). Other definitions have emphasised the *social* within social vulnerability, such as "an impaired ability to detect or avoid potentially harmful interpersonal interaction" (Pinsker et al., 2006, p. 110) and "degree of susceptibility to exploitation" (Pinsker et al., 2010, p. 741). As a result, the presence of social vulnerability is thought to contribute to adverse interpersonal experiences such as social exclusion and isolation; psychological, physical and sexual abuse; bullying and neglect.

Issues of social vulnerability in this sense have been emphasised and studied in a range of groups, including children (Seward et al., 2018), people with neurological conditions associated with older age such as dementia (Pinsker, 2011; Pinsker et al., 2006, 2010, 2011; Pinsker & McFarland, 2010) and individuals with intellectual and developmental conditions (Fisher et al., 2012; Fisher, Moskowitz, et al., 2013; Griffiths, Allison, Kenny, Holt, Smith, & Baron-Cohen, 2019; Jawaid et al., 2012, 2012; Riby et al., 2017; Seward et al., 2018; Sofronoff et al., 2011; Wilson & Brewer, 1992). There is extensive evidence that people with neurodevelopmental conditions experience adverse social experiences. Meta-analyses by Jones et al. (2012) and Hughes et al. (2012) examined the prevalence of violent victimisation for adults and children with disabilities. While their reviews included a diverse range of disabilities, the analyses on children/adults with ID are particularly relevant here. They found that adults with an ID were 1.6 times more likely to have experienced past-year violent victimisation compared to their nondisabled peers (Hughes et al., 2012). The figure for children was starker: children with disabilities classified as intellectual or mental health were 4.28 times more likely to have experienced past-year victimisation (physical, sexual or emotional abuse, neglect; L. Jones et al., 2012).

Issues of social vulnerability, social isolation and peer victimisation have been emphasised particularly within the autism literature, beginning in childhood and continuing throughout the lifespan for many autistic people (Cappadocia et al., 2012; van Roekel et al., 2010). Rowley et al. (2012) reported that 40% of their sample of autistic children experienced victimisation, bullying and social isolation due to their social difficulties, and similar findings have been reported in autistic adolescents (Van Roekel, Scholte, & Didden, 2010a). These issues continue throughout the lifespan; for example a report of 141 autistic people over the age of 16 found that over 80% had been victim to mate crime – being bullied or taken

advantage of by someone they thought was a friend (Autism Together, 2015). The age category 16–25-year-old was identified as a particular concern, with 100% of people in that age range reporting difficulty differentiating genuine friends from those who may abuse the relationship. Therefore, the evidence suggests that autistic people across the lifespan tend to experience a variety of adverse social outcomes, which map onto the aforementioned definitions of social vulnerability.

Evidence suggests that victimisation is present across the lifespan for people with ID (Codina et al., 2020). It has been reported that children with developmental conditions are more than three times as likely to experience abuse and neglect compared to children without such conditions (P. M. Sullivan & Knutson, 2000) and during school years, rates of bullying and peer victimisation are significant higher compared to neurotypical peers (Sentenac et al., 2013).

1.8. Importance of Studying Social Vulnerability in WS

The previous section outlined the concept of social vulnerability and illustrated that issues of social vulnerability are particularly relevant in the context of neurodivergent individuals. I now return to the WS profile to consider how the constellation of features (as outlined in Part A of this Chapter) makes social vulnerability highly relevant in this population. In doing so, it is useful to refer to Greenspan and colleagues' (2001) theoretical account of social vulnerability, which was based on their clinical observations of young adults with developmental conditions. In this framework, the authors argue that a core aspect of social vulnerability is a tendency towards *credulity*, ("a tendency to believe something, usually a highly questionable statement or claim, despite scanty evidence" p. 102) and *gullibility* ("a vulnerability to being tricked or manipulated" p. 102). The authors acknowledged that these components are closely connected, in that being credulous almost certainly leads to a gullible consequence (a tangible outcome i.e., handing over money). Whilst being trusting is seen as a positive quality overall, navigating interpersonal interactions requires the ability to differentiate interactions where compliance could result in some form of adverse outcome. In their view, socio-cognitive processing abilities—difficulties in deciphering the intentions of others and identifying potential threats—plays a central role in the presence of credulity and gullibility in individuals with IDD. As outlined in Part A, issues of this nature, particularly in deciphering the nuances of social interactions and evaluating intentions are experienced by many people with WS and this maps closely onto

the definition of social vulnerability as "an impaired ability to detect or avoid potentially harmful interpersonal interaction" (Pinsker et al., 2006, p. 110).

However, there are various other aspects of the WS profile, in addition to social cognition difficulties, that make social vulnerability even more relevant to understand in this group. Indeed, Greenspan et al. (2001) acknowledged the different causes, which may combine to provide various routes by which an adverse outcome may occur, and this is reflected in their framework which pinpoints four broad domain areas (termed personal competencies) that may impact on a gullible/vulnerable outcome; labelled 'intelligence' (including 'social intelligence'), 'communication', 'physical competence' and 'personality/motivation'. According to the framework, social vulnerability can be conceptualised as an interaction between environmental and personal competence factors, whereby in a potentially exploitative / manipulative situation, person factors can either promote opportunities for a vulnerable outcome or safeguard against it. Briefly, the domain of everyday intelligence encompasses both *practical intelligence*, defined by the authors as "understanding of physical objects and processes", and *social intelligence* defined as the "understanding of people and social processes" (mapping onto 'social cognition' as per my initial definitions). When explaining the aspect of social intelligence, Greenspan differentiates it from communication; specifying that the ability to draw upon a repertoire of observable verbal and nonverbal communication skills may be advantageous in terminating a potentially manipulative interpersonal interaction (e.g., averting eye contact, signalling the end of conversation, increasing personal space boundaries).

The WS characteristics outlined in Part A of this Chapter map closely onto the personal competencies outlined in Greenspan's (2001) framework of social vulnerability, though I note the model was not established in reference to WS. First, the strong desire for social connection seen in WS, evidenced by a heightened desire to approach others, is closely relates to Greenspan's category of motivation/personality described as being multifaceted, relating to an individual's goals and needs, efficacy beliefs, affect and attention. The following scenario put forward by Pinsker et al. (2010, p. 743) shows clear links with the WS profile:

"…a cognitively impaired person who is socially isolated may be motivated to interact with strangers, such as door-to-door salespeople, out of personal needs such as loneliness (goals and needs); the same person might have low self-efficacy (lack of

confidence in one's ability to make decisions or carry out tasks, that is, efficacy beliefs); and an inability to track a conversation (attention). With regard to personality traits, excessive agreeableness (affect) on the part of the vulnerable person may be central to establishing rapport with an individual who seeks access to the victim's financial assets under a guise of 'friendship'".

In the case of WS, the desire to approach indiscriminately, alongside a poor understanding of the potential dangers of interacting with strangers, presents an undeniable risk to safety. The longing to form connections with others, coupled with personality traits such as agreeableness and high empathy (Klein-Tasman & Mervis, 2003), could result in the acceptance of inappropriate or manipulative relationships to bring about a sense of connection.

Once engaged in an interaction, the common social cognitive aspects of WS (termed *social intelligence* by Greenspan, 2001) and pragmatic language difficulties (termed *communication* as per the framework), may impact on the flow and success of the interaction. In the context of a potentially manipulative interaction, difficulties with perspective taking, deciphering the motives and intentions of other people and labelling situations as manipulative / adverse, may place individuals with WS in a socially vulnerable position. Prolonged eye contact in addition to misjudging and invading the personal space of others may not only be deemed uncomfortable from the perspective of the other person in the interaction, but crucially might miscommunicate intentions. The use of expressive language and varied vocabulary may give an impression of a more mature level of understanding; however, difficulties with picking up on the social nuance of language (i.e., picking up on sarcasm or when someone is disinterested in the conversation) and deducing subtle non-literal meanings (i.e., jokes), may mean that people with WS miss or misinterpret cues. The following quote from a parent of a 9-year old with WS illustrates how a desire for interaction, alongside expressive language, interacts with a poor understanding of the boundaries of social interactions: "she will ask private questions she will tell things about herself which are just not appropriate" (Lough, Rodgers, et al., 2016, p. 1102).

The category physical competence relates to the fact individuals with IDD may also, but not always, have physical and sensory needs that mean important social cues are missed. The broader medical complications and physical characteristics often associated with WS (outlined at the beginning of Part A) are highly relevant here and may mean that people with

WS miss crucial visual and auditory cues during interactions. The physical characteristics may also mean that people with WS may place more reliance on family members and/or other people for decision making.

It is important to note that Greenspan and colleagues' theorising on social vulnerability was based solely on their observations of individuals with IDD and through conversations with family members, with no empirical literature to evidence their claims. Nevertheless, the theoretical framework provides a useful starting point by which to theorise on potential routes to social vulnerability in clinical populations, like WS, that have relevance to the personal competencies outlined in the model. Indeed, researchers have since applied and extended the framework in empirical research; for example, Pinkser and McFarland (2010) took the model as inspiration and adapted it to include other domains of personal competence relevant in the context of older people, including general cognitive functioning. While the WS profile maps clearly onto the personal competencies identified in the framework, it is not to say that this is comprehensive account of all the factors that may play a role in social vulnerability. For example, heightened levels of anxiety in WS are not accounted for within Greenspan's model per se but do relate to the subcategory 'affect'. Given many people with WS experience heightened anxiety, but less so to social situations, it is important to consider these additional behavioural features when investigating social vulnerability in WS.

Due to the unique WS profile, which encompasses features that map onto person factors that are considered to be important in social vulnerability, WS is an important population to examine social vulnerability (Jawaid et al., 2012; Riby et al., 2017). Indeed, concerns about social safety and potential vulnerability have been reported by parents/caregivers of people with WS and although evidence on rates of exploitation in adults with WS is limited, the evidence available is striking (Fisher, Moskowitz, et al., 2013; Lough & Fisher, 2016b). In a sample of 38 adults with WS, 79% were reported to have experienced some form of victimisation in their lifetime (Fisher, Moskowitz, et al., 2013). Parents reported that 37% of adults with WS had experienced teasing or persuasion, 37% had experienced theft or being tricked out of money and 16% had experienced some form of physical or sexual abuse. The authors reported that women with WS are particularly at risk of experiencing sexual abuse. Indeed, serious concerns about the risk of exploitation and abuse relating to levels of overfriendliness have been emphasised in the reports of parents of adults with WS. In a sample of 92 parents/caregivers of adults with WS (aged 19-55 years), two-

thirds were worried about the risk of inappropriate sexual experiences and 15 parents reported that their adult son/daughter had experienced some form of sexual assault (Elison et al., 2010). Therefore, issues of social vulnerability are a cause for concern for the families of people with WS and there is evidence to indicate real-life exploitation during adulthood.

The majority of research in this area has focused on adults with WS and we know far less about issues of social vulnerability in children and young people. However, qualitative research with parents of children with WS indicates that concerns about social safety exist for families of younger individuals with WS as they anticipate the challenges that increased independence will bring, as emphasised by the following quote from a parent of a 14-year old "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" (Lough, Rodgers, et al., 2016, p. 1103).

1.9. Evidence on the Factors Contributing to Social Vulnerability in WS

The issue of social vulnerability has been highlighted in the WS literature (Jawaid et al., 2012; Riby et al., 2017) and there is evidence of high rates of victimization, abuse and adverse social experiences. Despite this, the topic has been relatively understudied and there remains several aspects which relatively little is known. In particular, the factors that predict social vulnerability in WS are less well understood. This question is important as a greater understanding of how aspects of the WS profile may be related to elevated social vulnerability will in turn help to develop support approaches for fostering healthy social interactions and supporting families concerned about social vulnerability. This section considers what is currently known about social vulnerability in WS. Given the lack of research, there is value in reviewing the broader literature on social vulnerability in populations identified as having high levels of social vulnerability and showing divergence in their social, cognitive and/or behavioural profiles in ways similar and different to WS (see Chapter 2 for a discussion of the overlap in the phenotypes of WS and other various forms of IDD). Such findings give an indication of the potential underlying factors connected to heightened social vulnerability, which may be relevant for investigation in WS.

A literature review on social vulnerability and victimisation in adults with IDD found seven studies which explored risk factors for increased vulnerability (Fisher et al., 2016). The factors which emerged pertained to both aspects of the individual (person-centred characteristics) and the environmental/situation. Person factors included the presence of an ID, syndrome-specific facial dysmorphology and poor interpersonal competence. The

environmental/situational factors identified included living in institutional care settings but also individuals afforded increased participation in the community. In a series of studies, Fisher and colleagues (2012; Fisher, Moskowitz, et al., 2013; Fisher, Shivers, et al., 2020) probed patterns of social vulnerability in adults with IDD (including WS) and how these relate to other features of IDD profiles. The findings indicate that high levels of social vulnerability are linked to specific facets of vulnerability, such as low awareness of risk, being perceived by others to be more vulnerable (e.g. through appearance), increased independence from parent/caregivers and having few friends by which to offer social protection (Fisher et al., 2012). This work is valuable in elucidating some potentially relevant factors that may impact on social vulnerability and provides a starting point for more detailed investigation.

In terms of the link between social vulnerability and demographic variables, there is some initial evidence to draw upon. A study of adults with WS (and their parents; Lough & Fisher, 2016b) found that, at the group level, increased social vulnerability may exist across the across the spectrum of intellectual disability in WS and regardless of living arrangement (whether living at home or away from home) and employment status (employed or unemployed). Interestingly, the authors also reported no relationship between age and parent-reported or self-reported social vulnerability, indicating that social vulnerability may be present across the lifespan for people with WS. Of course, this was studied in an adult sample (mean age 28 years) and the relationship might look different in childhood/adolescence. Indeed, research with neurotypical children has shown that parents of younger children report increased social vulnerability compared to parents of older children (Seward et al., 2018). It would be interesting to consider whether this association with age is evident in CYP with WS too, or if social vulnerability is relevant across the full spectrum of age. The research to date on social vulnerability in WS has been almost entirely with adult samples and it will be important to understand the presentation of social vulnerability earlier on in development in WS, particularly in the context of changing social environments in the adolescent years.

Without doubt, the presence of ID is an important risk factor for social vulnerability. However, evidence of increased social vulnerability in neurodivergent groups that vary in their association with ID indicates that the presence of ID does not fully account for increased social vulnerability (Fisher et al., 2012). For example, research has found that autistic children (Asperger syndrome as per older diagnostic manuals) are rated more socially vulnerable compared to NT peers (Sofronoff et al., 2011) and rates of victimisation are higher

than typical levels in autistic adults without cooccurring ID (Hofvander et al., 2009). What is more, Lough et al. (2016b) found initial evidence that levels of social vulnerability on The Social Vulnerability Questionnaire (SVQ; Fisher et al., 2012) did not differ significantly across the spectrum of intellectual ability in WS. This finding, together with the evidence from research with other neurodevelopmental groups, suggests that other features of the WS profile, both cognitive and broader, potentially increase social vulnerability, in addition to ID.

What other aspects of cognition, beyond intellectual ability/disability, might be playing a role here? Looking to the broader literature on social vulnerability, research with other populations has identified executive functions (EF) and social cognition as playing an important role. For example, in a study of social vulnerability in older people, Pinsker et al. (2010) found that EF skills and social intelligence made a particularly important contribution, and led the authors to conclude that the perception of increased social vulnerability in older adults is not simply explained by advanced age, but rather the neurocognitive and socio cognitive changes that occur alongside neurological conditions. Research with young, neurotypical children found that cognitive domains of theory of mind and EF are important in relation to increased social vulnerability (Seward, 2016). As outlined earlier, these are also areas of challenge in WS (e.g. Greiner de Magalhães et al., 2022; Porter et al., 2008)

Jawaid et al. (2012) proposes, using autism and WS as exemplars, that it may be the combination of ID and divergent social profiles (e.g., social cognition capacities or social motivation styles) that result in heightened social vulnerability. There are few studies that have focused on the relationship between social vulnerability and facets of social behavioural profiles. In the context of WS, Lough (2016) provided preliminary evidence in support of a relationship between social vulnerability and social skills (as measured by the SRS). However, on the whole, evidence on the correlates of social vulnerability in WS, including the potential relationship with EF and social cognition, is lacking.

While looking to the broader literature on social vulnerability is useful in identifying potentially relevant underlying factors, researchers have argued the importance of carrying out detailed investigation with specific conditions as "individuals with IDD can be vulnerable in multiple, potentially unrelated ways, and it is important for researchers and clinicians to try to capture these distinct patterns of vulnerability" (Fisher, Shivers, et al., 2020, p. 2355). For example, WS, autism and Down syndrome (DS) are neurodevelopmental conditions associated with divergence from typical in their social phenotypes but vary in their

strengths and challenges. Research examining social vulnerability in these different groups within the same study indicates high levels of social vulnerability across groups, yet the correlates of vulnerability vary by diagnosis (Fisher, Moskowitz, et al., 2013). For example, autistic individuals score higher on issues of social protection, indicating that their social vulnerability might be related to having few friendships by which to offer social protection. In contrast, the pattern of social vulnerability for individuals with WS (and that of DS) was connected to risk awareness and perceived vulnerability through physical characteristics.

To summarise, evidence on the factors underlying social vulnerability in WS is limited. Research on the topic to date has been mostly restricted to samples of adults with WS and have not taken account of the range of factors that could be contributing to heightened social vulnerability. The broader literature on social vulnerability highlights the potential contribution of social cognition and EF.

PART 3 – CHAPTER SUMMARY AND THESIS OUTLINE

In Part A of this Chapter, the neurodevelopmental condition WS was introduced as the population of interest and features of the cognitive, social and mental health profile relevant to social vulnerability were outlined. It was argued that WS is associated with a paradoxical social profile—strong desire for social closeness alongside difficulties understanding social dynamics— together with heightened levels of anxiety and the presence of ID, which makes individuals with WS especially vulnerable in the social environment (Jawaid et al., 2012; Riby et al., 2017). In Part B, the term ‘social vulnerability’ was introduced, and links were made between an existing theoretical account of social vulnerability and the WS profile. The review of the evidence to date on social vulnerability in WS highlighted that issues of vulnerability are evident and although research has begun to outline some predictors of social vulnerability (Fisher, Moskowitz, et al., 2013; Lough & Fisher, 2016b), a greater understanding of the routes to social vulnerability is of critical importance (Thurman & Fisher, 2015). The review also highlighted that WS studies on social vulnerability have focused almost exclusively on adulthood, and one question that remains to be asked is whether increased social vulnerability seen in adult samples is also evident earlier in development in WS. I argue that issues of vulnerability are especially important to consider in the childhood and adolescence years because the heightened drive for social interaction presents early on in development in WS (evidence for hyper sociability outlined in 1.4.1). Additionally, adolescence is a period of rapid developmental change across the

cognitive and social capacities outlined previously (e.g., EFs, social cognition etc.) and is when CYP typically gain increased independence, with peer relationships becoming more salient. Therefore, most of the research in the thesis focuses on CYP with WS.

The overarching goal of the thesis was to contribute towards a better understanding of social vulnerability in people with WS, including the ways in which it might present and its association with the wider WS phenotype, using a mixed method approach. The longer-term aim of the research is to begin to provide researchers with valuable insights about social vulnerability that future research can build upon towards devising tools to support people with WS in their social interactions.

Before directly examining social vulnerability as such, the research first considers broader aspects of social behaviour and social functioning that have may have implications for vulnerability in social interactions. It has been proposed that the strong desire for social connections shown by many people with WS, alongside other challenges navigating social interactions, cause friendships to be an area of difficulty for many people. A lack of friends and, in turn, reduced social protection was identified as an important factor in increased social vulnerability in people with IDD (Fisher et al., 2012). However, current knowledge on friendships in WS is sparse and stems from studies that have tended to ask parents and carers to report on their perception of their son/daughter's friendships using ratings scales. As a result, we currently know little about how people with WS themselves experience friendships and the associated positives and challenges. *Therefore, to address this gap, Chapter 3 describes the conceptualisation and experiences of friendships from the perspective of young adults with WS*. Hearing directly from people with WS about their real-life social interactions and friendships provides first-hand insights about how friendships are experienced by people with WS, social decision-making, and potential challenges and social risk.

Next, Chapter 4 examines aspects of everyday social behaviour previously documented in the WS profile and has been emphasised in relation to the issue of social vulnerability: eye contact with others and social interaction style. As outlined in 1.4.1, intense looking behaviour and a propensity to approach others are two well-documented characteristics of the WS social/behavioural profile, which may impact on the flow and success of an interaction. However, it is unclear whether these behaviours are dominating in WS and whether they are unique to WS, or also evident in the profile of other neurodevelopmental groups known to diverge from 'typical' social behaviour. If the latter, it

would raise the question of social vulnerability in other groups. *Therefore, while WS is the primary focus of the thesis, the two studies in Chapter 4 take a cross-syndrome approach by contrasting a WS group alongside other groups with IDD, to understand the uniqueness of the WS social profile. Furthermore, the Chapter extends what is currently reported about social behaviour in WS by focusing on the 'quality' of eye contact and social interaction style. WS research to date has largely focused on the 'quantity' i.e., excessive eye contact or increased approach.* The rationale for adopting cross-syndrome comparative design is discussed in more detail in Chapter 2.

The remaining Chapters of the thesis focus exclusively on WS to build a richer picture of social vulnerability specifically in WS, as distinct patterns of vulnerability may be related to different forms of IDD (Fisher, Moskowitz, et al., 2013; Fisher, Shivers, et al., 2020). Previous research with adults with WS has reported high rates of victimisation and has outlined the social vulnerability profile, however, there has been little reported about social vulnerability earlier in development. *Therefore, to address this gap, Chapter 5 delineates the profile of vulnerability in a sample of CYP with WS, adding new knowledge about the nature of vulnerability in a younger sample of people with WS, and including two measures of vulnerability.* Following this, the focus of investigation turns to the factors associated with elevated social vulnerability. This is in response to calls from researchers who argue that to understand how best to support concerns about social vulnerability and foster healthy social relationships for people with WS, we need to move beyond documenting rates of victimisation and social vulnerability, to understand the factors that are contributing to social vulnerability (Fisher et al., 2016). *Therefore, Chapter 6 examines the relationship between social vulnerability and individual and family-level factors.* Individual-level factors of interest include some of the cognitive and behavioural characteristics emphasised in Part A, and include EF, anxiety, social cognition. In outlining the concept of social vulnerability in section 1.7 it was noted that social vulnerability is a combination of personal factors and environmental/contextual factors. *Therefore, Chapter 6 also explores, for the first, parental protection behaviours at this age and the association social vulnerability.*

The thesis concludes with Chapter 8, which pulls together the main findings and discusses the key contributions and implications of the research. This Chapter reviewed the literature on WS and social vulnerability and presented evidence that social vulnerability is a key facet of the WS social profile that demands further understanding. Having provided a

background by which to situate the forthcoming research and outlined the main aims of the research, in the next Chapter I give an overview of the methodology used across the thesis.

2. CHAPTER 2: THESIS METHODS AND APPROACHES

2.1. Chapter Overview

The research presented in the thesis uses a range of methodological approaches to further the understanding of social vulnerability in WS. The purpose of this Chapter is to briefly introduce the key approaches taken and outline the rationale for their inclusion. I start by providing context about the conceptualisation and broad design of the research. I then hone in on each of the methods/approaches in turn: self-report (Chapter 3), informant-report (Chapter 4 onwards), cross-syndrome comparative design (Chapter 4) and questionnaire methods (Chapter 4 onwards). A substantial part of the data collection coincided with the emergence of the Covid-19 pandemic in March 2020 and the subsequent cessation of face-to-face data collection. As such, some of the methods described in the thesis (Chapters 3, 5 and 6) were borne out of pandemic-related necessity to transition to remote methods of data collection. This Chapter touches on the impacts and mitigations. Further information on the ethical, moral, and methodological decision-making that informed the modifications, can be found in Appendix A.

2.2. Positionality Statement

My interest in undertaking this programme of research stemmed from a previous experience, working as a Research Assistant on a grant funded WS study. The study itself focused on aspects of the WS profile beyond social vulnerability, but through this role I visited approximately 15 families who had a child with WS. Prior to this, I had never met anyone with WS. In conversations with parents/caregivers during the home visits I learnt about their concerns about their son/daughter's social behaviours and, in particular, the impact these behaviours might have from a safety perspective. The conversations with families also coincided with some of the first published research exploring social vulnerability in WS. One of the researchers laying the groundwork in this area was Dr Emma Lough, who was carrying out her PhD in the same research group where I was based in the UK. Following this RA position, I continued to develop my research skills by working on projects related to child development topics, yet the conversations I had with families stayed with me. The lack of understanding about social vulnerability and the potential impact of new evidence in this area heavily influenced my decision to pursue the PhD.

2.3. Stakeholder Roles and Involvement

It is important to acknowledge the process that was taken in setting the scope and aims of the research. The PhD proposal was conceived between myself and my supervisors, in consideration of the gaps in the literature at the time of writing funding applications (January 2018). The Williams Syndrome Foundation (WSF) who supports people with WS in the UK and their families, was briefed on the proposed research and agreed to support the project as the non-academic collaborator. The WSF did not contribute to setting the research goals nor were members of the community consulted. However, my supervisor had developed a close relationship with the WSF from sitting on the professional advisory panel and attending national meetings of families, therefore was aware of the Foundation's priorities for research. At the beginning of the PhD, I briefed the trustees of the WSF about the aims of the research and collected their feedback. From my perspective, having the WSF as a collaborator was highly valuable as I was able to seek feedback on the study aims and protocols, access recruitment channels, and ensure the findings could be disseminated back to the families through the Foundation's newsletters and social media. Another stakeholder was the research funder, the Baily Thomas Charitable Fund who funds research into learning disabilities. The Trust played no role in setting the research questions or designing the studies, but did give helpful suggestions for consideration. For example, following discussion with the Trust's Medical Trustees (as part of the doctoral fellowship assessment process) I revised the order in which I conducted the studies – I originally intended to collect qualitative data on friendships (Chapter 3) at a later stage of the PhD, but brought this information-gathering forward to the start of the research timeline, to ensure that the qualitative insights could shape the subsequent quantitative research. I updated the funder on an annual basis through discussion with the medical trustees.

2.4. A Focus on Childhood and Adolescence

Across the studies of the thesis, participants range in age from 6 years to 30 years old. However, much of the research focuses on children and adolescents in response to the lack of research that has considered social vulnerability prior to adulthood in WS. As presented in Chapter 1, the reasons why social vulnerability is pertinent to study in WS (i.e., increased sociability, difficulties regulating social behaviour, socio-cognitive differences, and heightened anxiety) are evident from a young age in people with WS. Therefore, there is clear rationale for studying social vulnerability and the ways social vulnerability might present, well before adulthood. The studies described in Chapter 4 examine different features

of social behaviour relevant to social vulnerability and span the years of childhood and adolescence, including CYP aged 6-17 years.

In the context of social vulnerability, adolescence presents a particularly interesting time developmentally and Chapters 5 and 6 focus on this stage of life (participants aged 10-17 years). Adolescence is marked by significant developmental changes in the brain and body, as well as at the socio cognitive and emotion level (for reviews see Andrews et al., 2021; Patton et al., 2016). At the same time as these individual changes, there are substantial shifts in the social environment of CYP. Typically, children move from smaller class and school environments with more pastoral support in primary education to larger cohorts in the busy secondary school environment. This transition can be problematic for pupils with SEND (Evans et al., 2018; Hughes et al., 2013). However, note that this typical transition may look different for young people with learning disabilities like WS (Reilly, Senior, et al., 2015). More generally, time spent outside the home setting and with peers tends to increase. The physical maturation changes that characterise adolescence also impact on others' perceptions and behaviour. For example, a young person who has been through puberty may be treated more like an adult and afforded greater responsibility and independence. Taken together, adolescence presents an important stage to study social behaviours and potential social vulnerability.

In contrast, Chapter 3 focuses on adulthood due to the topic of interest and aims of the study: to learn about how friendships are conceptualised and experienced by people with WS, from the perspective of people with WS. By adulthood, people with WS will have experienced a range of life experiences to draw upon and will likely have experienced a variety of social interactions in varying contexts (i.e., formal education through to post-16 experiences). While the aim of the study was not to examine how friendships develop or change over time, speaking to adults provides a valuable opportunity to probe friendships at different key stages (e.g., by asking adults to reflect on their earlier experiences with friends at school). In addition, the decision to focus on adults was informed by practical and ethical considerations. We know from existing literature that people with WS can experience difficulties establishing and maintaining positive peer relationships, as well as bullying (Davies et al., 1998; Fisher et al., 2017; Gillooly et al., 2021; Udwin, 1990), therefore probing these potentially sensitive and distressing experiences was considered only appropriate with adults.

Overall, the studies included in the thesis represent a static point in time meaning it is not possible to make claims about developmental change. However, where relevant, analyses are conducted to examine relationships with age. I recognise and fully support that, like all behavioural outcomes, social vulnerability will interact with development itself and mechanisms cannot fully be understood without taking developmental change into account (Karmiloff-Smith, 1998; Karmiloff-Smith et al., 2012). However, research of this nature requires longitudinal data collection which is extremely resource intensive, both in time and cost, and not within scope of a doctoral thesis. Given the lack of evidence on social vulnerability, the field would benefit from first having a more informed understanding about the nature of social vulnerability in WS and how it presents earlier on in development. Future studies can then build upon the findings in the thesis, with larger sample sizes and using more complex longitudinal methodologies to answer the question: *what drives social vulnerability?*

2.5. A Note on Sample Size

WS is a relatively rare neurodevelopmental condition compared to conditions like autism and ADHD. As such, the WS samples described in Chapters 4-6 are relatively small (approximately 30 individuals), although this is comparable with previous UK studies involving individuals with WS (Gillooly et al., 2021; Glod et al., 2019; Kleberg et al., 2022; Lough, Flynn, et al., 2016; Rhodes et al., 2010; Rodgers et al., 2012). Indeed, WS studies generally have small sample sizes because recruitment is challenging for several reasons. Primarily, the rarity of the condition means that there is a smaller population from which to sample from. Due to funding constraints researchers may only be recruiting people with WS from one geographical region. Furthermore, attrition can be high because families are leading very busy lives and, lastly, due to the population of interest there are often more conservative ethical constraints. Small sample sizes are evident across the field of neurodevelopmental research more broadly meaning that studies are often statistically underpowered (Farran & Scerif, 2022; Hobson, 2021 estimates that the average power of neurodevelopmental research is 61% when .8 is the recommended standard). With this in mind, appropriate caution is required when interpreting the findings from the thesis.

2.6. A Note on the Impact of the Covid-19 Pandemic

The most substantial part of data collection for the thesis was planned to take place in one large, 12-18 month testing phase, visiting CYP with WS around the UK (due to the geographical dispersion of the WS sample) and inviting families to the University lab to use

specialist research equipment. The original research plans, as funded, involved taking an experimental, multi-methods and cross-syndrome approach to understand pathways to social vulnerability in CYP with WS, with the overarching goal of building a new model of social vulnerability. This included employing novel experimental methods (immersive virtual reality and eye tracking) to measure social behaviours in much more naturalistic contexts than has been possible to date. It also involved administering comprehensive behavioural assessments of cognition (i.e., IQ and executive function batteries) and use of a cross-syndrome approach to understand syndrome-specific pathways to social vulnerability. Most of the envisioned methods and tasks relied on in-person, researcher-participant interaction. Therefore, when Covid-19 hit in March 2020, the accompanying strategies to prevent the spread of the virus (i.e., social distancing measures, national lockdowns and school closures) had a substantial impact on my ability to conduct the experimental research as originally planned (see Table 2.1 for a breakdown of the measures/tasks impacted).

The key barrier I faced was that face-to-face testing with external participants was not permitted as per university regulation and in-person data collection with children and vulnerable groups would not be possible for a considerable amount of time (this type of research activity was the last to recommence at the University). At this time, some people with disabilities and serious underlying medical conditions were identified as being at a higher risk of becoming seriously ill from Covid-19 infection (Centers for Disease Control and Prevention, 2021). Indeed, due to the serious heart conditions and hypertension associated with WS, people with WS were identified as being clinically extremely vulnerable as per 'Group 2' criteria of the guidance issued by the Royal College of Paediatrics and Child Health (2020). Consequently, many individuals with WS and their family members were shielding to manage the risk of contracting the virus.

As a result of these circumstances, the research plans were redesigned to ensure the original goal of the research—to contribute to a better understanding of the ways in which social vulnerability presents for people with WS—could be met without the need for researcher-participant interaction and ensure the research could withstand an extended period of socially distanced data collection. In revising the methods for the parts of the PhD research that were impacted by the pandemic, the focus for the remaining empirical work shifted to a detailed account of social vulnerability using in-depth parent report studies. In these studies, it was possible to gain wide-ranging parent reports across several relevant domains of functioning, to really probe social vulnerability in new and important ways (e.g., looking at

different measures of social vulnerability and examining links to cognition, anxiety and adaptive behaviour). This meant that the original aims around social vulnerability could be addressed and novel insights gained, while minimising any risk to young people with WS and their families. Further information about the process of adapting the research and the ethical and methodological decision-making is given in Appendix A.

It should be noted that this empirical research was carried out in combination with other data collection, as part of a larger programme of research. In this larger programme of work, I led on piloting online methods for collecting data directly from CYP with WS. Although these data do not form part of this thesis, insights gained from the process of revising methods and conducting research at a time of pandemic-related restrictions, will be picked up in the general discussion (Chapter 7). The remainder of this Chapter is dedicated to the methods employed in the studies.

Table 2.1

Summary of Decision-Making: Adapting Methods/Tasks in Light of the Covid-19 Pandemic

Method/task	Rationale / purpose	Can it be achieved remotely?	Action and considerations	Solution
Behavioural measures				
Immersive virtual reality	Capture social approach behaviours in a more naturalistic context	No – use of specialist technology in the lab; participant-researcher interaction	Remove	
Eye-tracking	Social attention to social vignettes	No – use of specialist technology in the lab; participant-researcher interaction	Remove	
Stop-distance paradigm (Lough, Flynn, et al., 2016)	Compare data from novel virtual reality task with this published SAB task	No – hip-to-hip measurement taken during "live" social interaction	Remove	
Standardised assessment of cognition				
NEPSY (Korkman et al., 2007)	Assessment of: (1) executive functioning skills using the subtests Animal Sorting, Inhibition and Auditory Attention and Response Set	No – standardised for in-person protocol[1], requires manipulation of test materials	Remove and identify alternative method. Many online attention tasks online, ideally use one that has been published. Is it needed in addition to questionnaire BRIEF?	Informant-report via the BRIEF (Gioia et al., 2015)
	(2) emotion and mental state understanding using the subtests Affect Recognition and Theory of Mind	No – same as above	Remove and identify alternative online task of emotion perception e.g. (Griffiths, Jarrold, et al., 2019) but high number of trials – is it feasible?	Remove to limit duration of online testing
WASI (Wechsler, 2011)	Comprehensive, reliable but brief measure of intelligence using the four-subtest form (Vocabulary, Similarities, Block Design and Matrix Reasoning)	No – Standardised, in-person protocol Manipulation of test materials (block design) Time-consuming	Remove and replace with alternative measure(s) of cognition for remote delivery - Multiple considerations see Appendix A	Nonverbal skill via new digital version of the RPM Verbal skill via BPVS with permission from publishers (GL Assessment)

[1] Several publishers were in the process of digitising subtests to provide a potential solution to the physical distancing restrictions which clinicians and researchers were working. However, not all subtests could be digitised, and the measures did not yet have standardised norms based on remote assessment

2.7. Methods and Approaches Applied in the Thesis

2.7.1. Overview: A Multi-Method, Multi-Informant Approach

The research in this thesis employs several methods to further understanding about social behaviour and social vulnerability in WS. Most of the research takes a quantitative approach to investigation by, for example, using standardised questionnaire instruments, which affords a high level of control over individual-level variables such as social skills and challenges. To build a richer picture of the concepts of interest, the thesis also embraces qualitative approaches. This is most evident in Chapter 3 where semi-structured interviews are used to explore how friendships are conceptualised and experienced by people with WS (the rationale for combining qualitative with self-report is given below). A qualitative approach is suited to investigations of this nature, where the aim is to capture detailed information about subjective experiences. Qualitative data are also used to add richness and context to some of the quantitative findings. For example, in documenting the profile of social vulnerability in CYP with WS (Chapter 5), open-ended responses are given about real-life examples of vulnerability and strategies to cope with social vulnerability, to compliment a quantitative assessment tool. The addition of qualitative data alongside quantitative is useful to add context to the numbers, especially when studying a nuanced phenomenon like social vulnerability. The purpose and strength of adopting different methods, both quantitative and qualitative, was to gain a broader, more comprehensive understanding of social vulnerability in WS that would not be achieved by using a single design or research method. In addition, the thesis takes a multi-informant design, including both the voice of the person with WS and the views of parents and carers. The thesis prioritised (i) self-report to understand the subjective experience, (ii) informant-report to collect information on behavioural profiles, (iii) cross-syndrome design to examine features of the WS profile that may be shared / distinct with other neurodevelopmental conditions, and (iv) questionnaire measures to provide a relatively quick, valid and reliable assessment of cognition and behaviour.

2.7.2. Chapter 3 – Hearing the Voice of People With WS

Self-report and informant-report are two data collection approaches used in the thesis. Self-report requires participants to reflect and report on their own behaviours, abilities, feelings and experiences. In contrast, informant-report (also known as proxy report or third-party reports) involves an appropriate informant—typically an adult who has a high level of

interaction with the participant, such as a parent or caregiver—to give their perception of the individual's behaviour or capacity. A common trend in WS research, and in neurodevelopmental/ID research more broadly, is to seek the views of caregivers (e.g. WS studies: Gillooly et al., 2021; Lough, Rodgers, et al., 2016; Rodgers et al., 2012; Royston et al., 2021). Yet, in recent years, the field of neurodevelopmental research has evolved with researchers and community advocates acknowledging the importance of including the voice and opinions of people with IDD in research (Fletcher-Watson et al., 2019). There are challenges to taking this approach in research with populations with ID; a critical one being the rarity of appropriate self-report tools among this population. While valid self-report measures have been developed for autistic individuals to report on aspects of wellbeing (e.g. Hull et al., 2019; McConachie et al., 2018; Rodgers et al., 2016), there are considerably less self-report instruments for individuals with ID and those that do exist are mostly for adults with ID (e.g. ASC-ASD; Rodgers et al., 2016). Self-reporting via 'standard' measurement tools is often not appropriate for people with WS as these demand a level of understanding beyond an individual's developmental level to interpret complex wording/phrasing, understand abstract concepts (such as predicting behaviour in hypothetical scenarios), identify and reflect on one's own emotions and thoughts, and respond via rating scales (e.g. Likert scales) (Emerson et al., 2013; Finlay & Lyons, 2001). While studies have obtained information from individuals with WS through surveys and questionnaires (Dodd et al., 2009; Freeman et al., 2010; Lough & Fisher, 2016b), the demands of completing these scales can be exclusionary for some people with WS, meaning that samples are often biased towards individuals with more advanced cognitive skill.

In contrast, a qualitative approach to self-report can be more inclusive as questions can be phrased to align with developmental level. Additionally, in the context of the WS profile, interview methods can capitalise on individuals' strong social drive by engaging them in social conversation to hear their views. Indeed, WS studies using interview methods with people with WS have yielded valuable findings on topics such as peer relationships and friendships (Fisher et al., 2017; Gillooly et al., 2022), self-concept (Plesa-Skwerer et al., 2004) and mental health (Stinton et al., 2012). Overall, though, the WS literature lacks information from people with WS themselves and I argue that we need to place far more value on the perspectives of people with WS, particularly their views about their social behaviours and relationships.

Therefore, the empirical work of the thesis begins by applying a self-report, qualitative approach to the study of social interactions and friendships in WS. The experience of friendship is inherently personal, hence self-report is essential to get at the subjective experience. In addition, asking individuals to reflect on their friendships in their own words can yield valuable insights about individuals' insight into their own vulnerabilities and in turn help identify areas for support (Lough & Fisher, 2016b). For instance, in Chapter 3 adults with WS reflect on issues such as 'what makes someone a good friend?' and 'how do you know if you can trust someone?'. These accounts offer a window into how people with WS understand social relationships and how they go about social decision-making, relevant to social vulnerability. Hearing the perspectives of people with WS about their friendships ensures that the voice of people with WS is heard in the thesis. In addition, starting the thesis with this descriptive piece of work helps to identify issues relevant to social vulnerability that would benefit from more targeted investigation in later stages of the research.

2.7.3. Chapter 4 Onwards – Parent Perspective

From Chapter 4 onwards, the research collects data from parents and carers. There are practical and conceptual reasons for shifting to informant-report. First, a reflection from conducting the interviews in Chapter 3 was that many of the adults found it difficult to reflect and report on their experiences. This likely relates to the cognitive and communication difficulties associated with WS (as outlined in Chapter 1), which make the interview approach not well-suited to capturing the perspectives of all people with WS (discussed more fully in Chapter 3). In view of this, the communication and cognitive challenges were important to consider in the design of subsequent Chapters where questionnaire tools would be administered to measure features of behaviour and cognition in CYP with WS. As outlined above, there is an absence of self-report measures available that are suitable for CYP with WS to report on their behaviour and cognition.

Related to this discussion is the question of who is the right person to report on social behaviour and social vulnerability – young people with WS or their caregivers? There are a few WS studies that have examined the equivalence of self-report and informant-report, and, on the whole, the findings indicate a disparity between the two approaches (Fisher et al., 2014; Freeman et al., 2010; Järvinen-Pasley et al., 2010a; Lough & Fisher, 2016b). Järvinen-Pasley et al. (2010a) compared self-reported and parent-reported approachability to faces in the same sample and found little convergence between the two. In a different study using the

Strengths and Difficulties Questionnaire (SDQ; Freeman et al., 2010), adults with WS and their parents showed good agreement when reporting on difficulties with emotions and hyperactivity, however adults with WS did not report to the same extent as parents challenges with social relationships, via the Peer Problems and Prosocial behaviour subscale. Specifically, parents and adults agreed on items relating to being helpful and being bullied, but parents reported more difficulties on the items "at least one good friend", "being considerate of others" and "getting on better with adults". Fisher et al. (2014) took a step further by contrasting self- and parent-report with naturalistic observations of individuals' social behaviours and found parent-report to be more closely aligned with the observational data.

To draw firmer conclusions about the reliability of reports, we need more observational data alongside self and informant report data. Nonetheless, the evidence suggests that people with WS may underestimate their challenges particularly in areas relevant to social vulnerability. Indeed, the one study to compare self-reported and parent-reported social vulnerability of adults with WS found that adults perceived their overall social vulnerability to be lower than that reported by their parent/caregiver (Lough & Fisher, 2016b). The very nature of social vulnerability means that if an individual is likely to be in position of vulnerability they are highly likely to not realise it and, therefore, there is value in having an outside person to identify the risks that may have been missed. Linked to this, social vulnerability is an abstract concept to reflect on and articulate, especially for CYP with WS. Thus, in drawing assessments of social vulnerability parent-report was adopted (Chapters 4-6).

A further benefit of collecting parent insights and a key reason for taking this approach in the thesis is that parents and carers are arguably the people who spend the most time with CYP with WS. They therefore observe their son or daughter's behaviours in various situations and contexts, and have insights into their experiences, strengths and challenges. Parents and carers also have insight into how *other* people perceive, interact and experience their child's social behaviour, which is important as social vulnerability is a two-way interaction. Taken together, collecting parent reports was considered an appropriate and valuable strategy to probe the phenomenon of social vulnerability.

2.7.4. Chapter 4 (Papers 1 and 2) – A Cross-syndrome Approach

Two studies (Chapter 4) employ a cross-syndrome comparative design—contrasting multiple groups within the same research—to examine features of everyday social behaviour that may impact on social vulnerability. Cross-syndrome design is well established within the field of neurodevelopmental research; however, the impetus for taking this approach has evolved over the years in line with huge shifts in how we understand (neuro)development (Karmiloff-Smith, 1998). Most of the early cross-syndrome neurodevelopmental research prescribed to the adult neuropsychological framework, which viewed developmental conditions from the perspective of "impaired" and "intact" cognitive abilities and, therefore, focused on identifying double dissociations. For example, cognitive scientists in the 1990s proposed language to be 'intact' in WS, based on comparisons with other neurodevelopmental groups (Wang & Bellugi, 1993). The understanding of development then shifted from this simplistic, static notion of parts intact/parts impaired, to an appreciation of the neuroconstructivist approach to neurodevelopmental conditions, which emphasises development as a complex interaction of genes, environment, brain and behaviour (Karmiloff-Smith, 1998, 2009, 2012).

Researchers went on to advocate for the theoretical and clinical value of cross-syndrome comparisons as providing a "very useful tool for unpicking syndrome-specificity" (Hanley, 2015, p. 222) and, in the case of genetic based conditions like WS, studying pathways between genes, brain and behaviour (Haas & Reiss, 2012). From a clinical perspective, being able to delineate phenotypes can provide valuable context for families and professionals, which can guide areas of targeted support. Equally though, cross group design can identify commonalities across groups and there is now plenty of clinical and research evidence against the notion of neurodevelopmental conditions as discrete categories, rather there are multiple shared areas of need across mental health, attention, working memory, sleep, communication and behaviour (for useful reviews see Coghill and Sonuga-Barke (2012), Sonuga-Barke and Thapar (2021) and Thapar et al. (2017). Therefore, cross syndrome approaches to cognition and behaviour have the strength of allowing the unique features of a condition to emerge as well as the commonalities between groups.

In investigations of the WS social profile, a common cross group approach has been to make WS/autism comparisons, based on assumptions about these groups being at opposite ends of the sociability spectrum (Lincoln et al. 2007; Tager-Flusberg et al. 2006). While the

findings from some studies have led to descriptions of divergence within the social domains of these groups (Brock et al. 2008), they have also revealed many overlaps (Asada & Itakura, 2012; Klein-Tasman et al., 2009; Vivanti et al., 2018). Moreover, the notion of WS and autism having opposite social profiles is oversimplistic and developments in the autism literature has challenged the notion of reduced social interest in autistic people (Jaswal & Akhtar, 2019). While the social profiles of WS and autism have attracted significant research attention, other neurodevelopmental groups also are associated with challenges in the social domain. In Chapter 4 I explore how a cross-syndrome approach can provide a richer description of the associations between the social phenotypes of WS and other neurodevelopmental groups, and help to clarify links to enhanced social vulnerability. The research in Chapter 4 includes a group of CYP with WS in addition to CYP with other neurodevelopmental conditions that are associated with well-described social profiles. Previous studies with multiple neurodevelopmental groups has proven valuable in elucidating the phenomenology of cognitive and behavioural phenotypes in more detail than can be achieved by contrasting to a neurotypical sample (Oliver et al., 2011; Waite et al., 2014). Indeed, in Chapter 4, the multiple groups allow social behaviours of interest to be studied across variation in aetiology and presence of ID.

2.7.5. Chapters 4-6 – Questionnaire Measures

Questionnaires are used throughout the thesis to provide a relatively quick assessment of social vulnerability and other behavioural and cognitive variables of interest. While standardised behavioural assessments are the gold-standard approach for measuring cognition and behaviour, these require in person researcher-participant interaction which was not possible at the time of data collection due to the constraints of Covid-19 (see Table 2.1 for the behavioural assessments as per the original research design). Standardised questionnaire measures as an alternative have established validity and reliability to provide an objective assessment on cognitive processes, without the need for in-person interaction. Therefore, in Chapter 6, the Social Responsiveness Scale (SRS; Constantino & Gruber, 2012), the Behaviour Rating Inventory of Executive Functions (BRIEF; Gioia et al., 2015) and the Vineland Adaptive Behaviour Scales (VABS; Sparrow et al., 1984) are standardised tools used to assess social reciprocity, executive functions, and adaptive behaviour. It is important to note that while these measures were not designed specifically for use with individuals with WS, they have all previously been used in research with samples of CYP with WS (Brawn & Porter, 2014; Fisher, Lense, et al., 2016a; Greiner de Magalhães et al., 2022; Riby, Hanley, et

al., 2014; Van der Fluit et al., 2012). An additional advantage of questionnaires is that they are relatively quick to complete, meaning that we can gather information on several variables of interest, without being overly time consuming for participants. This is the case in Chapter 6 where the goal was to examine a range of cross-domain factors that likely impact on social vulnerability. Given the current lack of information about social vulnerability in WS, questionnaires are a useful tool to obtain an initial read on the type of cognitive and behavioural factors associated with social vulnerability, which can be followed up in future research using behavioural assessments.

In Chapters 4 (paper 2), 5 and 6, social vulnerability is the dependent variable of interest therefore its measurement warrants discussion. Across these Chapters the parent report Social Vulnerability Questionnaire (SVQ; Fisher et al., 2012) is relied on heavily, which reflects the lack of tools currently available to assess social vulnerability in individuals with WS, or ID more broadly. In the early stages of designing the research, the literature was searched to identify existing measures of social vulnerability. The Test of Interpersonal Competence and Personal Vulnerability (TICPV; Wilson et al. 1996) is a self-report tool for adults with ID which assesses a person's abilities in areas of social risks (e.g., financial exploitation, sexual abuse). Adults are asked scenario-based questions and have to report the behaviour they would likely adopt in a given scenario, from a multiple-choice option. While the TICPV has several strengths, good psychometric properties (Wilson et al., 1996) and has been used in research with clinical groups (Murphy & O'Callaghan, 2004; Tabin et al., 2021; Wilson et al., 1996), the items were deemed too sensitive for use with CYP with WS. In contrast and as its name suggests, the Children's Social Vulnerability Scale (CSVS; Seward et al., 2018) has been validated on a large sample of parents of children. A limitation of this questionnaire is that it has not been designed for use with CYP with ID. Additionally, as it includes only seven items, it would not allow for a detailed examination of the profile of social vulnerability (aim of Chapters 5 and 6). The chosen tool, the SVQ, has been used in the WS literature on social vulnerability to date (Fisher et al., 2012; Fisher, Moskowitz, et al., 2013; Lough & Fisher, 2016b). It too is not without its limitations; the most notable being that it was developed specifically for adults with ID. However, inspection of the items revealed that they were still appropriate for use with CYP. A key strength of the SVQ is that is allows for a detailed examination across multiple areas of social vulnerability, including emotional awareness, credulity, social protection, risk awareness and parental independence. In weighing up the tools available, I propose that there is currently no ideal assessment tool

for studying social vulnerability in CYP with WS, but the SVQ is most appropriate to meet the aims of the research.

2.8. Summary

This Chapter outlined the key methods and approaches adopted throughout the thesis and highlighted that there are many methodological challenges to conducting research with a rare neurodevelopmental group like WS. Further discussion of the methodology and design is given in the relevant Chapters. The next Chapter begins the empirical research of the thesis—an interview study with adults with WS to learn about social interactions and friendships.

3. CHAPTER 3: FRIENDSHIPS IN WIILLIAMS SYNDROME – HEARING THE VOICE OF PEOPLE WITH WS

3.1. Chapter Overview

Chapter 3 begins the empirical work of the thesis with a focus on friendships, from the perspective of adults with WS. In Chapter 1 it was identified that a lack of friends and, in turn, reduced social protection is a risk factor for heightened social vulnerability in people with ID. Additionally, while people with WS often have a strong desire for social connection, other features of WS raise challenges for interpersonal relationships (Thurman & Fisher, 2015). Yet, surprisingly, little research exists about the nature of friendships for people with WS. In the current study, a self-report qualitative approach was taken to capture what people with WS themselves have to say about their friendships and interactions with others. Hearing from adults with WS (18-30 years) was prioritised as by adulthood people are likely to have experienced various interactions with friends across different contexts (e.g., in formal education through to post-16 routes), thereby providing an opportunity to probe experiences across developmental stages and contexts. The data presented in this Chapter explores the conceptualisation of 'friendship' together with adults' associated experiences of friendships, capturing first-hand insights. To date, the opinions of people with WS have been neglected in research on friendships and social interactions. This self-report study provides important new findings and provides a useful starting point for discussing more creative, self-report methods in this field.

3.1. Introduction

Social connection in the form of friendships and interpersonal relationships plays a pivotal role in physical and psychological wellbeing, and overall quality of life (Chopik, 2017). Friendships can be defined and examined in different ways, such as the number of friends (Ho, 2016) and the quality of the friendship. Evidence suggests that the *quality* counts for more than the *quantity* (Tipton et al., 2013). Higher quality friendships associated with the features of trust, support and intimacy, are related with fewer internalising problems (e.g. lower social anxiety; Rodebaugh et al., 2015; Waldrip et al., 2008), increased self-esteem (Gorrese & Ruggieri, 2013), greater school adjustment (Ladd et al., 1996) and less loneliness (Alsarrani et al., 2022). Furthermore, high-quality friendships in the formative childhood/adolescent years set the stage for future emotional wellbeing (e.g. increased self-worth and lower anxiety; Narr et al., 2019) and healthy relationships in adulthood (Allen et

al., 2020). The positive gains from friendships are evident for people with intellectual and developmental disabilities (IDD). Friendship promotes community participation, increases wellbeing and gives people with IDD a sense of safety and consistency (Friedman & Rizzolo, 2018; Sullivan et al., 2016; Wilson et al., 2017). The importance of community inclusion for people with IDD has been recognised in public policy (e.g. Department for Health, 2001), yet research indicates that people with IDD engage in fewer activities and less frequently than their neurotypical peers (Solish et al., 2010; Taheri et al., 2016) and have smaller social networks (Tipton et al., 2013; van Asselt-Goverts, Embregts, Hendriks, et al., 2015; Verdonschot et al., 2009) that are often restricted to family members and support staff (Bigby & Knox, 2009; Fulford & Cobigo, 2018; Harrison et al., 2021; Kamstra et al., 2015; Mason et al., 2013). High rates of loneliness have also been emphasised with as many as 1 in 3 young people with a learning disability spending less than 1 hour outside of their home on a standard Saturday (Mencap, 2019). In contrast to the benefits of connection and friendship, loneliness is associated with adverse physical and mental health outcomes and poorer quality of life (Gilmore & Cuskelly, 2014). Importantly, a lack of friends has been identified as a risk factor for social vulnerability (Fisher et al., 2012; Fisher, Shivers, et al., 2020).

Social inclusion is more than simply being present in the community; it relies on being able to participate in a meaningful way where *quality* social connections can be formed (Cummins & Lau, 2003) and, in this respect, people with ID can face many barriers. There are major societal barriers to cultivating meaningful social connections and friendships, such as reduced opportunities to engage with others in the community (e.g. in leisure activities), but also social attitudes that have traditionally stigmatised people with disabilities (Abbott & Mcconkey, 2006; Gilmore & Cuskelly, 2014). Interpersonal challenges in areas of communication and social cognition, which are common in WS, are another relevant challenge with respect to friendship formation.

While friendship is an important topic of investigation in the context of people with IDD generally, less is known about the nature of friendships for specific neurodevelopmental groups. It is especially important to understand friendships in WS in view of the paradoxical profile of social desires and social challenges (in social communication and social cognition as outlined in Chapter 1). While other neurodevelopmental conditions are associated with challenges in these areas, it is the addition of the heightened desire for connection that makes research on WS a priority.

3.1.1. Evidence on Friendships in WS

Despite the accumulation of evidence that people with WS have a strong desire for social connections but experience interpersonal barriers that transcends across ages (Thurman & Fisher, 2015), there has been surprisingly little investigation of what this means for the friendships of people with WS. The existing research indicates that friendships are an area of challenge, both in terms of development and maintenance. Claims such as these stem from research in the 1990s which examined the behavioural profile of children with WS (Greer et al., 1997; Udwin et al., 1987) and life outcomes of adults with WS (Davies et al., 1998; Udwin, 1990). For instance, in a study of 70 adults with WS (19-39 years), 96% of parent/carers endorsed issues relating to establishing and maintaining friendships and nearly 75% said that their son/daughter was socially isolated (Davies et al., 1998). Similar reports of social isolation were documented in early investigations of the behavioural profile of children with WS (Udwin et al., 1987). Such reports were published 30 years ago and since then there has been huge societal shifts towards the inclusion of people with IDD, yet we still know relatively little about what this means for the friendships of people with WS.

Overall, what is known about the friendships of people with WS has predominately stemmed from research using parent/caregiver report. For example, as part of a study on health and social outcomes of adults, Elison et al. (2010) found that 40% of adults (19-55 years old) with WS were reported by their parent/caregiver to have one or more friends seen on a regular basis, however 30% reported substantial problems making friends and nearly half described their son/daughter as having a poor understanding of friendship as a concept. Parents also emphasised a lack of social inclusion, with almost half of the sample rarely or never attending clubs. Similar findings have been reported by parents of children with WS, typically through the use of rating scales (Greer et al., 1997; Laws & Bishop, 2004; Udwin et al., 1987). Greer et al. (1997) reported that 60% of a sample of 15 CYP with WS (4-18 years old) showed borderline-significant ‘social problems’ and significantly low levels of social contacts with others on the Child Behaviour Checklist (CBCL; Achenbach, 1991). Similarly, Laws and Bishop (2004) reported that parents of nineteen young people with WS (6-25 years old) endorsed lower ratings of social relationships compared to neurotypical controls on the Children's Communication Checklist (Bishop, 1998). On one item of interest on the CCC ‘tends to be babied, teased, or bullied by other children’, the WS group had significantly more problems in this area compared to neurotypical controls and groups with Down syndrome and DLD. The findings highlight social relationships to be a key area of challenge

for people with WS; however, these studies make inferences about friendships using scores on subscales from much broader, standardised measures of adaptive behaviour and social functioning—the CBCL (Achenbach, 1991), the Vineland Adaptive Behaviour Scales (Sparrow et al., 1984) and the CCC (Bishop, 1998). While items within these measures do address behaviours relevant to friendships, the subscales have not been designed as a measure of friendships per se and they focus on broader elements of social and adaptive behaviour. What is more, the quantitative approach offers limited insight into the nature of friendships.

More recent research has taken a multi-informant approach to the study of peer relationships, acknowledging that relationships may present differently in different contexts. Gillooly et al. (2021) collected parent and teacher assessments of peer relations in a sample of children with WS (parent n=21, teachers n= 18). Both parents and teachers endorsed peer problems (through the Peer Problems subscale of the Strengths and Difficulties Questionnaire, SDQ; Goodman, 1997) that were significantly higher than population norms, indicating that peer relationships are problematic for the children with WS across both home and school environments. Most importantly, however, the researchers moved beyond the reliance on standardised measures (SDQ) by developing a bespoke informant measure ('Peer Interactions Questionnaire') to capture information about the characteristics of peer relations across different contexts. Analysis of this measure revealed interesting differences in the peer relations reported by parents and teachers, specifically in the presence of friendships and ability to sustain friendships. With regard to the school environment, the majority of teachers (78%) reported that children with WS were greeted by peers around the school, however less than half (39%) reported that they were included by peers during recreational breaks or during conversations in the classroom and very few teachers (28%) endorsed that children were chosen by peers as a partner in activities/PE. Therefore, while children with WS may be engaging with peers in a surface-level capacity (i.e., saying hello in a corridor), the results suggest a lack of meaningful peer interaction conducive to friendship formation. A lack of social inclusion was echoed in the parent report, with only 38% of parents endorsing that their child was invited to the homes of peers. Interestingly, none of the teachers reported children being picked on by peers and there was consistency in parent and teacher reports of a lack of peer conflict.

The use of more tailored measures to examine peer relationships is an important step forwards, however, a quantitative approach using ratings scales provides little detail into the nature of friendships. Gathering evidence from multiple sources is also crucial, because

interactions with others vary across situations, but also people can perceive the same behaviours/relationships differently. Related to this is the question of how people with WS perceive their own friendships, as we cannot assume that this equates with the view of informants (e.g., parents and teachers). For example, from an informant perspective the friendships of people with WS might be perceived as lacking in quality, not meeting an assumed criteria of 'friendship', or being problematic in some way, however; this might well contrast with the individual's perspective (Wiener & Schneider, 2002). Indeed, what friendship means to people is inherently personal and can only be gathered using methods that place emphasis on the voice of the individual. To date, very few studies have asked individuals with WS to report information about their friendships.

A review of the published literature revealed that little attention has been given to what people with WS themselves have to say about their friendships. One relevant, self-report study took a survey approach to collect data on the quality of friendship in a large sample of adults with WS (Fisher, Josol, et al., 2020). Participants were first asked to name a best friend, then, with this person in mind, answer questions about the characteristics and qualities of the friendship using the Friendship Qualities Scale (Bukowski et al., 1994) which assesses the qualities 'companionship', 'conflict', 'help', 'security' and 'closeness'. Of the 114 adults who completed the survey, only two could *not* name a best friend. Of those who named a best friend, 60.3% reported speaking to the friend at least a few times per week and that the friendship rated high quality on areas of help, security and closeness, but not companionship. These findings are enlightening in that, in contrast to the parent/caregiver reports from the 1990s (e.g. Davies et al., 1998), adults with WS reported regular contact with at least one friend and that the friendship includes high quality features. However, as acknowledged by the authors, a key limitation is the focus on participants' *best* friend, thus likely painting the most optimistic view of friendship and other friendships may vary quite considerably. In addition, while the use of self-report makes an important contribution, the quantitative approach of using frequencies from survey data is not conducive to obtaining a rich account of the subjective experience of friendships.

There is a lack of research that has taken a self-report, qualitative approach to the study of friendships in people with WS. Most recently, Gillooly et al. (2022) conducted an interview study about friendships with children with WS and their parents and carers. The children, aged between 7 and 16 years, all named at least one friend and did not report feeling excluded, but these interactions were largely restricted to the school context. The parent data

on the other hand documented more nuanced friendship challenges related to behaviour regulation, presence of social anxiety, exclusion from activities and difficulties sustaining friendships with neurotypical peers. While important to obtain the child perspective, the authors reported that the child interview data was not suitable for qualitative analysis due to brevity and endorsed more accessible participatory methods. Moving up the developmental spectrum, Riby et al. (2017; and for a more comprehensive account see Chapter 10 in Lough, 2016) used a case study method to gather qualitative insights from a 23-year-old with WS, and their parent. While the study had a broader focus on social behaviour and internet use in people with WS, 'friendships' emerged as one of the key themes from thematic analysis of the interview. In comparing insights from the adult and the parent, the authors reported a difference in the perception of 'friendship', in that the adult with WS self-reported having hundreds of close friends, whereas the parent endorsed that these would be more accurately described as acquaintances ("to her, a friend is just someone who has smiled at her once"; Riby et al. p. 186). The authors also interpreted that the adult had difficulty in differentiating friendships from relationships, and friends from strangers. The parent endorsed this and highlighted a difficulty in educating on the distinction between friends/strangers as the adult with WS firmly held the view that acquaintances qualified as friends. While the parent insights in tandem with the adult perspective is valuable, more emphasis on the adult perspective would generate useful insights into the nature of the friendship as experienced by people with WS. Furthermore, given the subjective nature of complex social phenomena like friendships, along with the considerable heterogeneity in all aspects of WS, it is important to go beyond the case study approach and obtain insights from more people with WS. Nevertheless, this study provides a valuable starting point by which to build upon with further qualitative research.

Capturing the voice of people with WS through semi-structured interviews has also helped to understand experiences of bullying for people with WS. In a study of 15 adolescents and adults with WS, all reported having experienced bullying: 80% reported having experience verbal bullying, 60% had been left out of a group and 53% had experienced or threatened with physical violence (Fisher et al., 2017). While reducing the interview data to numbers helps to ascertain the prevalence of different forms of bullying, it is the participants' narratives around bullying that add particularly interesting insights. For instance, when asked about their responses to the bullying, Fisher et al. (2017) identified a

core theme of ‘self-advocacy’ whereby many individuals discussed feeling empowered to stand up against the bullying and determination to stay true to oneself.

In summary, the topic of friendships in WS has received relatively little attention since reports of social isolation and general difficulties with friends was documented in the literature over 20 years ago (e.g. Davies et al., 1998). What has been reported has stemmed predominately from parent/caregiver perspectives, evidence on adult outcomes and making inferences from standardised measures of social behaviour (Davies et al., 1998; Elison et al., 2010). While some progress has been made in recent years to advance understanding through multi-informant insights (Gillooly et al., 2021, 2022) and obtaining the perspectives of people with WS (Fisher, Josol, et al., 2020; Gillooly et al., 2022; Riby et al., 2017), more self-report, qualitative research is needed to move beyond documenting the presence/quantity of friendships and instead capture the subjective experience. Qualitative insights about what friendship means to people with WS and the positives and challenges they experience, would help shine light on how best to support people with WS to develop healthy relationships (Athamanah et al., 2019). There is evidence to suggest that people with and without IDD may hold different conceptualisations of friendship and vary in their opinions about what constitutes as good quality friendships (Freeman & Kasari, 2002; Jobling et al., 2000; Matheson et al., 2007; Sigstad, 2017; Webster & Carter, 2007). However, a more recent synthesis of the evidence on friendships in people with IDD indicates that these aspects do align with conventional aspects of friendships, with descriptions of friends as those who offer support, are trustworthy and have common interests (Fulford & Cobigo, 2018). Conceptualisations also likely evolve with age. In young age, it would be expected that friendship is conceptualised around mutual interest in playing together, whereas later in development, this may evolve to centre on more high-level qualities such as support and trust. Hearing directly from people with WS about the way in which they conceptualise friendships may also yield important insights about their own understanding of potential vulnerability.

3.1.2. The Current Study

Given the lack of detailed information about friendships from the perspective of individuals with WS, the current study took an exploratory approach to examine the conceptualisation of ‘friendship’ in adults with WS, together with their associated experiences of friendships. Hearing the voice of adults with WS was prioritised, as by adulthood, people with WS are likely to have experienced various interactions with friends

across different contexts (e.g., the school years moving towards increased independence), therefore, providing opportunity to probe experiences of friendships across age and contexts, and potentially gaining insight into how friendships evolve. Given the existing evidence that peer relationships are an area of challenge in childhood (Gillooly et al., 2021), it is interesting to consider how these experiences with others may impact on friendship development later in life. There is mixed evidence as to whether friendships improve in adulthood in WS. For example, longitudinal research has found slight age-related improvement in aspects of friendships (Elison et al., 2010) and recent insights from people with WS themselves have indicated positive quality friendships in adulthood (Fisher, Josol, et al., 2020).

3.2. Method

The study uses qualitative data collected from interviews with adults with WS. The study received ethical approval through the departmental ethics committee.

3.2.1. Research Participants

Ten adults with WS and/or their caregiver were recruited to the study in response to a study advert circulated through the Williams Syndrome Foundation newsletter and social media channels. Participants were UK-based. One participant's data was removed prior to transcription and analysis as this person did not engage with the interview format and their caregiver reported most of the information in the interview. The final sample included seven women and two men aged between 19 and 30. Employment status varied: student / continued education (n=4), voluntary work (n=2), part-time paid employment (n=2), unable to work (n=1). Parents / caregivers provided basic demographic information through an online questionnaire or over the telephone with the researcher. Given the rarity of WS and the low number of interviewees, personal information beyond gender and age range is not specified to maintain anonymity. An assessment of the participants' ID was not conducted, as it was not within the remit of the research to probe the association between friendships and cognition.

3.2.2. Interview Schedule

A 12-item semi-structured interview schedule was developed with reference to the Social Behaviour and Internet Use Interview – Adult Schedule as had been used in a case study of an adult with WS (Riby et al., 2017; see Lough, 2016 for a full description). The interview schedule was structured around three main themes: the conceptualisation of 'friendships', associated experiences with friends and meeting new people. The interview schedule was designed to meet the purpose of the research question but be accessible to the

participants and allow them to describe their own thoughts and experiences. Supplementary questions and prompts were also asked when necessary (i.e., if a question was met with a very limited answer. See Appendix B for the interview schedule.

3.2.3. Procedure

Following best practice guidance of informed consent, participants and their parents and carers received the study information in written and oral format (video advert with ER providing a verbal overview), using accessible language appropriate for people with ID. The parents/carers who responded to the study advert were invited to complete consent forms (adults with WS also provided consent). Arrangements were made to conduct the interview at a date and time convenient for the adult. All interviews were conducted by ER. At the start of the interview, ER introduced herself and explained the reasons for doing the research. Participants were reminded that the interviews would be audio recorded and assured that there were no right or wrong answers, that their answers would be kept confidential and any personally identifiable information given would be removed when the findings were written up. Three of the interviews took place prior to the Covid-19 pandemic and were conducted in-person at the University. The remainder of the sample was collected remotely during the pandemic, using the videoconferencing software Zoom. The in-person interviews were conducted in a private, quiet room between the researcher and participant. For the interviews conducted remotely, one of the participants requested to have their parent/caregiver present in the room for support, and the remainder were conducted independently between the researcher and the participant. The length of the interviews ranged from 26 to 50 minutes. At the end of the interview, participants were debriefed and given an opportunity to ask questions. Interviews were audiotaped (n = 3) or recorded using in-built recording software (n = 7) ready for transcription.

3.2.4. Data Analysis

Given the variable richness of the participants' accounts, the study used qualitative content analysis to describe participants' thoughts, understanding and experience of friendships. Content analysis is a research tool to ascertain the presence of words, ideas or concepts within qualitative data (e.g., texts, pictures, audio or video material). It is a gradual process of analysis, increasing in the level of abstraction at each stage which can span manifest analysis (literal) to latent analysis (interpretation into categories / themes). Data were analysed using Erlingsson and Brysiewicz (2017) as a guide. Given that there is little

known about friendships in WS adults from the perspective of individuals themselves, an inductive, data-driven approach with no a-priori themes was set. As advised in qualitative analysis, reflection and discussion regarding the researchers pre-understanding was undertaken before analysis, to be mindful not to let it influence the analysis. However, one must acknowledge that our minds are never completely blank from the wider literature. First, the interviews were transcribed verbatim by IM, a research assistant on the project who was a second year undergraduate Psychology student with experience of qualitative analysis. The lead researcher, ER, read 100% of the transcripts several times independently while listening to the recordings, paying close attention to sections of recording marked by IM as inaudible (due to regional accents / background noise), but also as a strategy for quality checking. The transcripts were then read several times for familiarisation by both ER and IM. Notes and comments were made about general impressions. With the research question in mind, the transcriptions were then divided into meaning units (i.e., several words, a sentence or a statement that relate to a single idea) indicating information relating to (a) conceptualisation of friendships, (b) experiences with friends (c) interactions with unfamiliar people. Using NVivo, meaning units were then labelled using descriptive codes to organise and reflect on the data.

3.3. Results

The analysis generated several categories pertaining to (1) the conceptualisation of friendships, (2) experiences with friends and (3) meeting new people. The results below provide a summary of participants' accounts and are accompanied by quotes from the participants using pseudonyms to protect the identity of the participant and their age is given in parentheses. Some quotes are preceded by the interview question (labelled 'ER' where this gives context to the answer). An overview of the key findings is presented in Table 3.1. There was variation in the level of detail reported across the interviews – some participants gave detailed answers whereas others gave short answers or went off topic. A general reflection was that some interviewees reported having lots of friends and being popular (e.g., Jess, aged 19: "yeah I have lots of friends... I can't remember all of them, but they're really, really nice, so I'm like got loads of friends"). However, many interviewees saw these friends infrequently or were drawing upon experiences with one or two friends for the duration of the interview. For instance, when asked about a fun time with friends, Michael (aged 26) replied "Okay so for, um, I'm gonna use, [Name] and [Name] yet again"

Table 3.1

Content Analysis Findings from Interviews with Nine Adults with Williams Syndrome: Conceptualisation of Friendship, Experiences with Friends and Meeting New People

Category	Example codes	Example quote
Conceptualisation of friendship		
Support and encouragement	Look out for each other / help others	You're willing to help other people. It means that you're scarifying a lot for other people. I wanna go the extra mile to go and stop someone from crying and stuff like that and willing to help basically.
Good communication	Listening / communicate	Yeah, you know, I think good communication is certainly one thing. If you have, if you have good communication with them
Laughter, fun and conversation	Bubbly / fun / be funny	…and they seem nice, polite, kind, friendly, bubbly, fun, funny, those are all the qualities that I like in a friend. If I find them boring I wouldn't go with them kind of friends
Meet up and shared activities	Meals out together / activities together / watch films / have a catch up	If we hang out, hang out with them then I would be their friend, but if they don't wanna do that there's no point
Genuine care and trust	Reliable / truthful / honest	What being a good friend to me is being honest with your friends.
Evaluating trust / genuineness	Using body language / having common interests / gut feeling or impressions / trust based on nice gestures / the other person's reaction to them	How I'd be able to work out if I can trust them if for example if I kept visiting them and knowing that each time after I visited them that it's going well and that we're okay taking and being with each other
Change and variation	Different friends / mixing friends / lose friends / loyalty / stable	If friends grow up and they wanna leave I'm fine with that too because in life you grow, you know, you don't just stay where you are all the time. So I know that friends can grow apart from each other and you know that's okay.

Category	Example codes	Example quote
Experiences of friendship		
Friendships from family members and staff / support workers	Family members / personal assistants / youth worker / staff at house	Umm [short pause] my cousins are my friends, they're really nice
Negative peer experiences and conflict	Bullied when younger / verbal bullying / physical bullying / mismatch of needs / not understanding WS / cyberbullying	I had to go through many difficult times of when people had bullied me and you know that was tough
Community participation	Meeting friends at a day centre / meeting friends at work / keeping in touch with friends through activities	Well I got a job at work and I feel I am equal to them, and I feel they genuinely like me and not just 'oh I have to like her'. Do you know what I mean?
Navigating conflict	Apologising when not in the wrong / friends giving support / getting help from family/staff / using communication / standing up for oneself	I was sceptical and I thought, you know what, this isn't right, I don't need that in my life you know. I need people that I can trust that I can go to if I need anything
Meeting new people		
Positive feelings	Positive now older / happy / thrilled	I'm over the moon to do it – that's my motto
Both anxious and excited	Excited / anxious	Like nervous and happy at the same time
Needing to get to know them first	Difficult if unfamiliar / get to know them well enough / takes time	A tiny, a tiny bit hard because if, um, if someone I know I'm alright talking to them, but if it's different I find it a bit hard at first, but then I get used to talking to them…
Finding it hard, feeling judged	Anxiety / feeling judged	'Cos I don't know what they're gonna think of me and what they're gonna say about or how they're gonna approach me, so I do get a bit worried and a bit scared
Greetings	saying hello / asking how they are / shaking hands	I'd say 'hi' at first and then they'd say, 'oh hello' and if they come back they say 'oh hiya' and then I say 'oh hi'
Getting to know each other	Tell them about yourself, compare issues, put effort in, learning their likes and dislikes, finding shared interests	Like what we both like, and what hobbies we like to do, and what we like to do in our spare time.

3.3.1. Conceptualisation of Friendships

When asked about what makes a good friend, adults used a variety of words including 'passionate', 'funny', 'polite', 'nice', 'kindness', 'honesty', 'understanding', 'kind', 'caring'. The participants identified a range of qualities or features about friendship that they value and there was consensus that friendships provide emotional support and help. In addition, participants emphasised friendships providing companionship – doing activities together and sharing fun times. It was important to the adults to have friends that were genuine and could be trusted. The key categories pertaining to the conceptualisation of friendships are presented below.

Friends give support and encouragement. Eight of the nine interviewees mentioned the qualities of support, care and encouragement in good friendships.

> *Anna (29):* That you always look out for each other, like if you see your friend upset- make sure that they're okay, like, like take your time out to do things for them and help them with anything that they need help with and keep reassuring them and encouraging them and like keep their spirits up and don't ever let them down, because, because otherwise you can lose them…
>
> *Sam (30):* Um, helping one another if things are not the way they should be, um just looking after each other.
>
> *Emily (23):* Someone who goes the extra mile, someone who protects you from storms or the vampire, you know, I'm just using it as an example.
>
> *Frances (24):* Never backing out when things get really tough

Importance of good communication. Codes relating to the qualities of communication and listening was identified in three of the adults' accounts.

> *Helen (25***):** Um, understands that what they're talking… um, being a good listener, communicating, uh, trying to be there, um, yeh.
>
> *Michael (26):* Communication is one of the most important things to me of all times, like you know without communication nothing can you know, you can't change a situation, or you can't resolve a situation.

Importance of laughter, fun and conversation. Codes relating to laughter and good conversation were identified in six of the interviews:

> *Sam (30):* making each other laugh
>
> *Bethany (20):* That we just like the same things, and we laugh at each other, like we laugh together at everything.

Helen (25): having a nice laugh and a chat

Friends meet up and share activities. Codes relating to meet up with friends, socialising and doing activities together were present in three of the accounts:

Anna (29): I like friends that like to come out with me and have a laugh and have a catch up.

Georgie (24): Mm like, like birthday meals out together, do activities together and stuff like that.

Friends show genuine care and are honest and trustworthy. This category refers to the emphasis participants placed on having genuine friendships and the qualities of honesty and trustworthiness. Indeed, codes relating to this category were present in eight of the interviews.

Bethany (20): That they're respectful to who you are

Frances (24): And not just..not just having to be a friend for the sake of it – I really appreciate it

Helen (25): because they care, they're bothered about how I'm doing and…like some people don't care, they're just like 'oh hiya' and then they go but some people are like 'Oh, how's this, how's that', you know..

Anna (29): I think what they've got to be is honest and truthful with me, because if they're not truthful with me, how do I know how to trust them or how to cope with them or anything like that. Unless they're honest and truthful with me then I'm not gonna you know

Evaluating trust / genuineness. While participants valued trust in a friendship, when probed further, many found it difficult to articulate how they would evaluate if someone can be trusted or a genuine relationship (n=4), as emphasised by the quotes below.

Frances (24): Just self-consciously thinking: Am I saying the wrong thing here or do they, do they actually pretend to care, or do they actually genuine…? I do know the difference, but it's hard for me to kind of question it all the time. I always question it; I always always question it…Even with a stable relationship, yeh. I don't question it so much, but if, you know, I'm always unsure whether it's true, but I think now, I think I do now when it's true and when it's not true.

Helen (25): I don't, that's the thing, 'cause I don't know if they wanna be my friend for being me or 'cos of my Williams syndrome

Sam (30): Um, I can't really tell

Anna (29): It depends like what they say about their past and if it's true, because I don't know whether it's true or not, you know, so…

Four of the adults reported making judgements of trust automatically / based on gut feeling:

> *Bethany (20):* 'Cos I know then when I've got a good feeling about them, and they've got a good feeling about me
>
> *Frances (24):* Gut feeling. I always try and follow my gut in most things, situations I do, that's how I try and estimate somebody, I just follow my gut, and whether they're actually telling me a lie or not.
>
> *Jess (19):* I can trust in people that I like, but if I cannot trust in people that I don't like, I would not trust them
>
> *Michael (26):* I think I've got a fairly good sense of character to be honest.

Two adults referred to trusting others if they made a gesture or bought something for them:

> *Anna (29):* because if they, if they do things like, let me say, put themselves out there for you, and if they like kind things, it's like [name of friend], she bought me Strictly Come Dancing tickets to go to [place] arena to see the show.
>
> *Michael (26):* It's the actions that you know, like [Name] asking me if we want to go out for dinner is a sign that I could trust him. So, it's you know, if I can trust them, then I believe that they are my friends.

Three adults referred to sussing people out or using body language as information about trustworthiness.

> *Anna (29):* Oh yeah, 'cos I can always tell by the way that they're sitting, by their moves, by the way that they talk to me, you know, like if they aggressively talk to me, then I know that they're lying.
>
> *Georgie (24):* Like the way they speak to me, or if they get snappy with me or anything like that, I just, I just know that that's not a nice person to be friends with and stuff like that
>
> *Emily (23):* Um, when they're not answering me or they're just laughing really fake at me or something.

Five adults referred to using other people's reaction to them, to evaluate trustworthiness:

> *Jess (19):* Um, I would just wait and see if they trust me first, 'cos I'm really trustable and I wouldn't be trustable to them, if they're not trustable to me, 'cos it's really difficult.
>
> *Bethany (20):* They they..that they're trustworthy and that they're kind and that they're happy to be friends with me.
>
> *Frances (24):* Just the warmth that they give you and smiles and genuinely checking that I'm okay, it's just, you know, going out of their way to make sure I'm okay.

Three adults reflected on getting to know the person or taking time to decide if people are to be trusted:

> *Helen (25):* Um, just like take some time with them just to see if they're being who they say they are
>
> *Anna (29):* 'Cos I've learnt in life not to trust people unless I know them and it's worked.

Change and variation in friends. Four adults reported being aware that friendships can change over time in terms of meeting new friends in the future and some friendships coming to an end. Examples of this are highlighted in the quotes below.

> *Helen (25):* you can have different friends, you don't have to have the same friend every time
>
> *Michael (26):* sadly sometimes friendships do kind of progress and sometimes you lose them, which is what happened so.
>
> *Anna (29):* Sometimes I mix my friends up a bit, and I think that's nice because then you get a variety of different, of different people, you know, that come into my life, you know, and I'm happy, you know, I feel that it's better, having a variety of.. it's like my dad lives in [place] so I have life loads of close friends that live down there that I can go and see and like up here, I have lots of close friends that I can, so it's lovely, it's just a nice feeling.

Equally, five adults reflected on the importance of loyalty and stability in friendships:

> *Michael (26):* the good things about my friends are that the friends I've got have always stuck with me. And like, the people like [name] and [name] and my guitar teacher [name] and my friend [name] from [city], they've all stuck with me.
>
> *Bethany (20):* Yeah, I would look for a friend that I could keep, and that wouldn't fight with each other, or be nasty to each other. We'd be best friends forever *laughs*.
>
> ER: Okay, so you've said you have 'best, best, best friends'. I'm just wondering what makes someone a 'best, best, best friend'?
> *Sam (30):* Um, they, they even looked after me at college or school, or they were, um friends from the start

3.3.2. Experience of Friendships

Adults described the joy of having friends in their life and could describe good times spent with friends (n=7). When asked if they considered themselves happy with their friends at the time of the interview, all adults reported being content.

> *Anna (29):* Yeah! And I wouldn't change them for the world, like if anybody wants to come and join my friends then that's great. 'Cos it'll mean that I'll make more friends and I have more fun.

Emily (23): Yeah, um, when we go to the [name of pub] in [place] it's so good, it really is. I have a good dance, yeah it's good times.
ER: Right and how do you feel being with your friends?
*Emily***:** Amazing, just really happy, in my zone.

Frances (24): But you know the rare friends that I have are genuine and I know they're genuine.

Sam (30): If I organise to meet up with someone then I'll take them for lunch in either Costa or Greggs or um McDonalds or KFC. And we'll walk around the shops and have a catch up

Bethany (20): just like a sister to me. We're always having a right laugh together and we go out.

However, there was variation in this, with some adults struggling to recall good times with friends as illustrated by the exchange below:

ER: So, can you tell me about a time you had a lot of fun with your friends or when something good happened?
Helen (25): Umm [short pause], no not really.
ER: No?
Helen: No…
ER: Any events that were fun with a friend?
Helen: Uh, just like going to a friend's like, huh, college events, and getting freebies and stuff like that.

Viewing family members / staff as friends. In their descriptions of, many of the adults (n=5) talked about friendships with family members and paid members of staff (e.g., personal assistants, staff in support living settings).

ER: And who are you thinking about when you're thinking about those friends?
Emily (23): A lot of people…Um, I think about [name of personal assistant], because she takes me out. I think about my mum, obviously. I think about my nana sometimes, 'cos she was a big, big part of my family. She always asks me about my condition and, so that's why last year was a bit hard when she went.

Michael (26): Yes, but before I met, I met them, I met a really nice guy around her called [name], who's like, um, who's like a youth worker, so I met up with him, because I was going through depression about three, two years ago, so, I needed someone to help me get out of it all, and he helped me and then yeh, from there I met his colleagues and we became, I became friends with like two of his main colleagues, so yeh.

Frances (24): I went to, I went away, I had a friend down for a week actually, and we went away for a few days, and it was really lovely...Yes, she's my PA, but equally she's my friend though. I get on really well with her. We went to the seaside, and we just went and did beach stuff, it was really nice, and went to the Sealife Centre, and all sorts.

> And my friend [Name], I always have a lovely time with [Name] every Monday I see him, and it's really lovely.

Negative experiences with peers and bullying. This category of experience was dominant within the adults' accounts (n=8) hence I have included a variety of quotes below to reflect this. The interviews reinforced that despite their desire for connection, participants had experienced negative friendships and interpersonal situations that often include bullying, particularly during school years. Many of the adults were able to identify that these experiences constituted as bullying. Some of the experiences described were quite distressing and participants spoke about them with emotion, even though for many it has been several years since the event. This included physical bullying (n=2) but more common were reports of verbal and emotional bullying (n=5). Participants spoke about adversity relating to peers not understanding them or about WS (n=4). Disagreements, conflict and falling out with peers was generally also common.

> *Michael (26):* So, for example, I had a friend called [Name] at school, and I had to wear back braces for two years, and the one day I was sitting down to have my lunch and he was just removed my chair from me, and I fell to the ground. So, from then, I was like, well you know, I don't really trust anyone at this, you know if that's the kind of thing that I'm gonna have to be subjected to.
>
> *Georgie (24):* I got bullied a lot. I had to leave a couple of times.
>
> *Emily (23):* Mm, I got really badly bullied by these two boys, they were laughing, and they were saying: 'Uh, she's this, she's that.' Yeh, they got put in the exclusion base,'cos they were really harsh to me and…yeah, like they wanted to…My mum wasn't happy. Obviously, you know, they were like, 'she doesn't look nice, she's got nothing, she's got no passion, her condition sucks', the list went on, they would just go, they wanted to pave me in you know.
>
> *Frances (24):* Throughout my whole kind of, until now, I've kind of felt like the odd one out of many social groups going through school and then going into college, it definitely felt I was the odd one out. Nobody really wanted to be friends with.. [break]. I just felt that I wasn't understood by them maybe, or that I didn't understand what they wanted out of me and it was kind of very difficult to keep it and I felt very sad that I couldn't.
>
> *Helen (25):* Um, they're nice, but it's like, uh, it's a bit difficult sometimes to get along with them, 'cos they have different needs and don't have William's Syndrome…So, it's a bit difficult to, and like, if I like don't know something, then they go: 'Oh, [Name] you should know' you know, just like, bad chat at me or make me feel stupid sometimes or…

Anna (29): Yeah ‘cos she was physically abusing me and calling me bad names and kicking me and hitting me and like giving me evil looks and pulling like awful faces at me and I knew, um, that that was wrong.

At the same time, participants reported having found a friend or friends at school/college they could connect with:

Michael (26): Um, yeah, I’d just say that [name of high school] is where everyone accepted, you know, everyone accepted me for who I was without the need to do bad things, so yeah.

Bethany (20): It’s like my own gang of people, like a group of friends that I sit with sometimes when we have dinner and stuff.

Sam (30): well at college I was very, very popular [laughs]. I went to a mainstream college and they had like this group called ‘towards independence’ and, um, there were other people with learning difficulties at that college.

Navigating conflict. Regarding coping strategies, participants reported a range of ways they had managed the challenging experiences with peers, including relying on family members or support staff for emotional support and confidence (n=4), taking the blame or apologising when not in the wrong (n=3), standing up for themselves or cutting off a friendship (n=4), getting support from other select friends (n=2), using communication (n=2), sticking by friends even when not nice (n=1). Example quotes relating to this category are given below.

Bethany (20): I talk to my other friends about it and they help me calm down and stuff, whenever I need it.

Emily (23): Um, my teacher and my sister, just them. Calm me down and just make me feel a lot better about it

Helen (25): I say sorry to her, but like, like even if I didn’t do anything wrong, I still say sorry, and she’s like: ‘Oh, okay.’ But then it’s like, she’ll do it again, like and she’s done it again

Michael (26): We handle it by communication, but he and I have definitely had times where we’ve just been like you know, where I’ve mainly been like- I just don’t feel like talking to you right now, you know.

Anna (29): ‘Cos I have to stand up for myself and I said ‘how would you feel if I did that to you back? You know, you wouldn’t be happy about stealing someone’s…’ and I say, ‘think before you do things or think before you say things to people’, because she nearly lost me, obviously because what she did.

The role of community participation. Codes relating to community activities, employment, volunteering and day centres providing a means for meeting new friends and staying in touch with friends was reflected in seven of the adults' accounts. For example:

> *Frances (24):* They know me for my true colour, you know, they know me inside out, they know when I'm stressed, when I'm happy, you know, and not many people get to see that side of a person unless they're in an environment such as my job, you really understand the true person, I think.
>
> *Anna (29*): some of my friends do drama with me on a Tuesday and I do cooking on a Wednesday and I have fitness on a Friday, so I have a nice… and my friend [name] goes to Arts & Crafts as well, and it's just nice, like it's like one big happy family.
>
> *Michael (26):* …but I've made two lovely friends who are called [name] and [name] who I met at work […] we go out for dinner with each other. Like going out with me was a really big thing for me like mentally, because no one else had gone from work out with me. […] so that was very meaningful because that, you know, I had two people that actually, you know, wanted to be involved with me.

3.3.3. Meeting New People

The data collected regarding meeting new people included participants' feelings and the actions they take in these situations.

Positive feelings (n=4)**.** Participants reported positive feelings about the prospect of meeting new people:

> *Helen (25):* Um, not when I was younger, but now I like meeting new people more
>
> *Michael (26):* I love meeting new people if I, if, you know if I can get to know a new person well enough.

Both anxious and excited feelings (n=3)**.** For others, they described feeling a mix of emotions – both excitement and anxiety – when faced with meeting new people:

> *Anna (29):* Excited and nervous at the same time, but then once I meet them I'm alright
>
> *Sam (30):* I was a bit nervous and a bit excited at the same time

Finding it hard / feeling judged (n=5). The adults also reported meeting new people to be difficult and for some this was related to anxiety or feelings of being judged:

> *Helen (25):* Um, just like being, felt like I'm being judged. Just like when someone looks at you, you feel like you're being judged. You know I wasn't being judged, but I felt like I was being judged.
>
> *Sam (30):* Um, I, I get, well, I, I I, have quite bad anxiety sometimes, so, it's hard for

me to meet new people.

Anna (29): 'Cos I don't know what they're gonna think of me and what they're gonna say about me or how they're gonna approach me, so I do get a bit worried, and a bit scared, and a bit like 'Mm, should I be doing this or not?'

Needing to get to know them first (n=5). Additionally, some of the participants mentioned feeling more positive once they had a chance to get to know the person and become familiar.

Michael (26): I love meeting new people if I, if, you know, I can get to know a new person well enough

Frances (24): I'm, I'm very wary, you know, it takes a lot for them to say: Ah okay, this one's a keeper, or this one's fine. It's just, I have to, I know I'm quite difficult in that sense, but once I warm to them, I warm to them really well.

Bethany (20): Sometimes I get nervous, but one I get to know them more, I don't feel as nervous around them. So, I feel much calmer around them, when, when I know them.

Greeting people. When asked about meeting someone for the first-time adults talked about greeting others.

Helen (25): Um, say it's my sister's friends, I go and say: 'Oh, hiya, you alright?' and then they'd be like: 'Yeh, [Name] you alright?', but like if it's someone that is new, I say: 'Oh hi, I am [Name]' sister'.

Emily (23): Just say 'Hello, I'm [name], nice to meet you. What's your name and how are you?' Just the easy questions: Are you fine? How are you feeling? Yeh…

Sam (30): Oh, um, I sometimes, um, see if they're okay, or how they're, I ask how they're doing.

Asking about them and finding commonalities (n=5). Other participants discussed trying to initiate a more substantial conversation to get to know the new person and identify any shared interests. Some reported sharing about WS and finding commonalities by talking about needs.

Anna (29): I like to sit down with them and I like to get to know them and then like I probably tell them a bit about myself and my issues and they tell me about their issues as well and like we compare, like our issues together and then like we become like really nice friends

Michael (26): Like for me the most important things is like getting to know what their emotions are, like learning what makes them happy, what makes them sad, you know what makes them, you know, what makes them whop they are. That's the first thing that I do and then gradually after that I learn: Okay, this is what makes them happy, this is not what makes them happy, what can I do to make them happy and that's how I go about it.

Jess (19): Um, I would, I would ask them what they got, like what history they got, and what they actually have. And all my, all of my William's Syndrome family have William's Syndrome, so it's really nice to meet them.

3.4. Discussion

Rather than relying on informants as much of the past research on friendships in WS has done, this study put the focus on understanding friendships from the perspective of adults with WS. The findings demonstrate that people with WS have views and experiences to share about their friendships, which can provide valuable insights about where extra support might be needed. The findings are discussed in view of existing literature, before addressing limitations of the study and implications.

3.4.1. Conceptualisation of 'Friendship'

The study first sought to describe how adults with WS conceptualise friends and friendships. The qualities that adults valued in friends were, overall, what has been described in the neurotypical literature—both practical (e.g., having fun together, activities) and psychological qualities (e.g., support and care). This mirrors reports in the ID literature that friendships are described in terms of functional and psychological benefits (Garolera et al., 2021; e.g. Mason et al., 2013). The adults with WS placed emphasis on trustworthiness, described friends as being "honest" and spoke strongly about the importance of having genuine friends. Yet, the findings, although from a small sample, indicate that adults have trouble knowing whether someone can be trusted. In fact, some of the explanations reported by participants about how they evaluate trust in others is concerning and emphasises the key point on social vulnerability. For instance, two participants described having trust if the person had bought them something or invited them somewhere. This indicates that some adults with WS have a superficial understanding of trust, which may feed into the high rates of victimisation and social vulnerability experienced by adults (Fisher et al., 2017; Lough & Fisher, 2016b). These findings are consistent with previous studies that have reported clinically significant social skill challenges in many adults with WS (Fisher & Morin, 2017) and issues with trust evaluations specifically, such as increased approach to untrustworthy faces (Jones et al., 2000; Martens et al., 2012) and problematic evaluations of stranger-danger scenarios (Riby, Kirk, et al., 2014). The current findings add to these reports by providing a glimpse into adults' reasoning when making judgements of trust and emphasise that adults

with WS may be naïve to deception and succumb to lures by people with disingenuous intentions.

3.4.2. Experiences of Friendships

When describing their friendships, adults talked about their family members and professional support staff. This is not uncommon; it has been reported in the wider literature that people with IDD rely on family members and paid staff for friendship (Kamstra et al., 2015; e.g. van Asselt-Goverts, Embregts, & Hendriks, 2015; van Asselt-Goverts et al., 2013). While these family relationships would not be classified as friendships in the conventional sense, it is not to say that these relationships do not provide adults with WS with the same sense of connection and wider-positive gains. This is evidenced by research that asked people with ID about their networks and found that people with ID were satisfied with their networks that included family members and support staff. The research also found an association between the quality of *affection* linked to family and staff , and self-rated quality of life (van Asselt-Goverts, Embregts, & Hendriks, 2015). Indeed, many of the adults in the present study reported being content with their friends. It is important to understand the friendship outcomes that people with WS themselves want to achieve and devise support to work towards that, rather than placing neurotypical expectations (i.e., a certain quantity of friends or friendships with people the same age). Nevertheless, we do need to consider the nature of the friendship from the other person's perspective given that the friendship needs to be reciprocated and meeting the needs of both people to have longevity. It could be that the qualities people with WS see in their family members and support workers justify them as friends. It could also be true that people with WS have limited opportunities to meet new people and develop friendships, therefore place high level of importance on these other relationships. This has been echoed in research on friendships in people with ID more broadly—disabled people tend to be restricted in the physical spaces that they inhabit and, as a result, friendships tend to develop only within particular settings and remain bound to those settings (Milner & Kelly, 2009).

Linked to this, the role of community participation also emerged in the descriptions of experiences with friends. It was apparent that many of the adults had met friends and/or were keeping in touch with friends through employment, volunteering, and groups/activities in the local community (arts and crafts, swimming, going out for dinner). These findings emphasise the importance of independence, community inclusion and participation in providing

meaningful opportunities for adults with WS to develop and sustain their friendships and promote wellbeing and a sense of belonging (Sullivan et al., 2016) . In more recent years there has been a shift towards community inclusion for people with ID and indeed 'inclusion' was one of the key principles identified by the UK government policy *Valuing People* for service provision for people with ID (Department for Health, 2001). Yet, as argued by others, mere exposure to the community is not sufficient to foster a sense of connectedness (Abbott & Mcconkey, 2006; Cummins & Lau, 2003). A lack of community activities has been identified as a barrier to friendships by people with ID (Abbott & Mcconkey, 2006; Rourke et al., 2004). Even when community activities are available, we know that adults experience barriers to participating, such as a lack of travel skills or access to reliable and accessible transport (Ann Bross et al., 2023; Mencap, 2019; Welsby & Horsfall, 2011). Additionally, parental/carer concerns about their son or daughter's social vulnerability and safety might be another barrier to adults with WS participating in the community (Fisher et al., 2017; Lough & Fisher, 2016b). Indeed, people with ID report a desire for more independence (Fulford & Cobigo, 2018) and find overprotective behaviour and a lack of autonomy as having a negative impact on relationships (Sullivan et al., 2016).

A critical theme identified in the adults' experiences relates to conflict, falling out with friends, negative peer interactions and, for at least some, bullying (including physical and verbal). These findings align with reports that people with ID experience greater vulnerability in their social lives, including bullying and victimisation (Cappadocia et al., 2012; Falla et al., 2021; Fisher et al., 2017; Fisher & Taylor, 2016; Griffin et al., 2019; Kloosterman et al., 2014; Sofronoff et al., 2011). A previous interview study on bullying in WS found that all 15 adolescents and adults reported having experienced bullying (Fisher et al., 2017). In their study, Fisher et al. acknowledged that the high endorsement of bullying may have been attributed to a selection bias in that those who responded to the study were those who wanted to report their experiences of bullying. Whereas, in the current study, the interviewer did not prime about bullying, but did probe challenges with friends and difficult experiences. For most adults in the current study, negative peer experiences had occurred when they were younger during school years. This finding is consistent with recent studies by Gillooly et al. where teachers of children with WS reported greater peer problems on the SDQ compared to normative levels (2021) and parents reported a lack of social inclusion (2022). Interestingly, in contrast to the current findings, teachers and parents in Gillooly (2021) did not endorse peer conflict in the school environment. In the current study, for some

adults who reported negative peer experiences at school this was related to peers not understanding WS and/or having a mismatch of needs. Other negative experiences reported were related to cyberbullying and romantic relationships, reflecting the stage of development and styles of interaction in a modern world (Caton & Chapman, 2016; Lough & Fisher, 2016a).

When discussing strategies used to navigate these experiences, there were varied approaches reported. The majority of participants referred to getting support and guidance from family members and professionals, mirroring reports from adults with various forms of ID and highlighting the importance of support networks for people with WS (Fulford & Cobigo, 2018; Garolera et al., 2021; Griffin et al., 2019). Some adults with WS reported taking the blame and apologising even when not at fault, whereas a few participants spoke about empowerment and self-advocacy. The concept of self-advocacy is interesting and also emerged in Fisher et al. (2017) when adults with WS were asked how they respond to bullying. A systematic review of 16 studies found that self-advocacy groups can give adults with ID tools to process trauma, engage in self-development and protect themselves in future scenarios (Tilley et al., 2020). The ways in which adults with WS deal with negative peer experiences is far from understood and warrants further investigation.

Overall, the key findings from the interviews are consistent with reports of friendships in adults with ID generally. For instance, experiences of bullying in the school context are common and the findings reiterated that this is often related to peers not understanding or being accepting of difference. Another example is the importance of relationships with family members and staff which is well documented across the ID literature (van Asselt-Goverts et al., 2013; Giesbers et al., 2018) and likely reflects a lack of community inclusion and social connectedness – an issue not distinct to WS (see Roll & Koehly, 2020 for similar findings in a study of adults with Down syndrome). In summary, the experiences reported by the adults in this study were similar to reports from individuals with other forms of ID. Therefore, support and resources designed for other groups may be usefully applied in the case of WS.

3.4.3. Limitations

There are several limitations to this study that warrant consideration. The current study was limited to adults with WS who were motivated to participate in an interview about friendships, or whose parent or carer deemed they would be interested and able to engage in the study. Within the sample, there was marked variability in individuals' abilities to reflect

on and talk about friendships – while some participants provided detailed answers, drawing upon examples to provide context to their thought processes and decision making, others struggled to reflect and generate detail in their answers. This was evidenced by the variable length of the interviews. Such variation in engagement and level of detail likely reflects the varied intellectual and communication profiles in WS (Searcy et al., 2004), coupled with the abstract subject matter (e.g., what it means to be a good friend). Therefore, it is possible that the categories generated in the analysis are biased towards the accounts of individuals who were engaged, had stronger verbal communication and were more confident articulating their thoughts and experiences. Indeed, adolescents with higher IQ and communication skills have been reported to describe more mature qualities/definitions of friendship and lower satisfaction with their friendships in comparison to those with lower IQ and communication skills (Matheson et al., 2007)

It is also important to recognise that an interview approach may be anxiety-provoking for some individuals. In fact, the study interview data on 'meeting new people' highlighted that despite descriptions that people with WS thrive on meeting others, adults can be shy and nervous in such situations (and evidence of age-related increases in social phobia as per Ng-Cordell et al., 2018). We can apply this to research scenarios where adults are interacting with researchers for the first time and being asked probing questions. Therefore, instances of limited detail in the adults' accounts might not solely reflect the experience of friendships as the method of data collection may also be impacting the results. A small but potentially valuable addition to the study protocol would be to hold a brief 'warm-up' chat with the participants in advance of the interview, to help reduce anxieties and reservations. A priority for future research therefore is to consider methods and tools (e.g. through co-production with adults with WS) to enhance the information-gathering and make self-report more accessible to people with WS who may not have the same level of expressive language or wish to contribute in this format.

It is also important to reflect on the format of the interviews. Given the small sample (three interviews took place face-to-face and the remainder were remote using video conferencing software), it is difficult to draw conclusions about whether engagement levels and the flow of the interviews differed between the two formats. This is also compounded by individual differences in personality and sociability. From my perspective, it was not additionally challenging to build rapport and conduct the interviews online. However, I would note several challenges with remote interviewing: first, unlike the in-person

interviews, it was not possible to control the environment around participants and there were instances of family members being present in the background which may have impacted on the answers given and, at times, this was a source of distraction and it took time to refocus on the interview. Second, being online presented barriers in assessing the wellbeing of participants during the interview (body language cues were more difficult to observe). Finally, instances of poor internet connection disrupted the flow and clarity of the conversation.

Additionally, while we view a strength of the study to be that it placed the emphasise on the participant's own words to understand friendships, it is important to acknowledge that friendship is reciprocal in nature and the friends mentioned by adults in the interview, or other observers, may have varying perceptions of the relationship. Indeed, reports from parents of children with WS indicate that friendships are often not reciprocated to the same level (Gillooly et al., 2021, 2022). As the purpose of the current study was to focus on the perspective of the person with WS, this is not a limitation perse, but it is important to recognise when thinking about the wider profile of vulnerability. For example, if adults with WS are viewing their friendships as high quality and getting a lot out of them but this is not mirrored by the other person, then it may have consequences for the longevity of the friendship and potential vulnerability. While it is true that self-report on friendships has its limitations, we approached this with the acceptance that adults with WS are experts in their experience, and if we want to understand how friendship is experienced then their insights are crucial.

A further limitation is that the study did not collect information about whether the adults had received any support or training on issues such as peer relationships and stranger danger. This additional information would give context as to whether participants who gave more nuanced accounts of friendships and reported self-advocacy approaches in the face of conflict, had received prior coaching or training. Finally, a limitation of the study is the small sample size. While consistent with sample sizes achieved in other qualitative research with people with WS (e.g. Fisher et al., 2017; Gillooly et al., 2022), gathering insights from more adults with WS would paint a greater picture. Given the rarity of WS, interview studies should capitalise on the use of remote technology which has become commonplace since the Covid-19 pandemic.

3.4.4. Implications for Practice and Future Research

This study contributed new information about how young adults with WS conceptualise and experience friendships and, as such, has several implications relevant to those who support individuals with WS. One of the key issues that emerged from the adults' accounts is challenges around trust evaluations. There is, therefore, a definite need to support people with WS in deciphering whether to trust someone and teaching about evaluating intentions behind behaviours and awareness of lures and deception. To date, there has been few interventions developed to support healthy social relationships in people with WS, yet the evidence does show promise for training programmes with adults with WS (Fisher, Burke, et al., 2013; Fisher et al., 2022; Fisher & Morin, 2017). However, it is not enough to teach just about safety with strangers. Abuse and victimisation is often carried out by someone known to person with ID (Fisher et al., 2016; Vadysinghe et al., 2017), therefore adults with WS also need support and guidance on assessing the behaviour and intentions of individuals familiar and close to them, especially as we found that adults with WS may depend and have strong attachments to individuals provide care and support. Support with identifying and evaluating body language may be one avenue for training given we know people with WS may have difficulties picking up on social cues (Hanley et al., 2013; Järvinen-Pasley et al., 2010b).

Another important implication for educators and practitioners who support someone with WS is that despite their overtly sociable nature, by adulthood people with WS are likely to have experienced negative and upsetting interactions with peers, particularly during their school years which they may still need support to talk through and understand (adults spoke emotionally about these experiences even though for many it had occurred many years ago). Research has found that negative emotions in others can be a trigger for high anxiety in some people with WS (Royston et al., 2021), and perhaps distressing peer interactions feed into the increase in anxiety with age (Stinton et al., 2010; and see Ng-Cordell et al., 2018 for evidence of increase in social anxiety). In the school environment specifically, CYP with WS may need significant support to develop a sense of belonging, build friendships, find strategies to negotiate disagreements and cope with bullying and teasing. At the school-level, there is a value in educating other pupils in the classroom about differences and promoting acceptance and understanding, especially when a child with WS is in a mainstream setting – an example of such an approach is Learning About Neurodiversity at School (LEANS) which teaches

neurodiversity concepts to primary school children (for the preregistration of an evaluation of LEANS see Alcorn et al., 2022; Salvesen Mindroom Research Centre, 2023).

Furthermore, an important lesson to take away from this study is that adults with WS rely heavily on the connections and friendships they get from family members and professional staff. Issues may arise here, especially when family members (e.g., siblings) move on and have their own families. This may be a source of sadness and loneliness for people with WS. We know that for some people with WS high anxiety is related to concerns about the health and welfare of significant others, anticipatory worry about the future and separation more generally (Dykens, 2003; Royston et al., 2021), which may become all the more relevant as adults get older. Linked to this, community participation plays a role in developing and maintaining friendships, especially in adulthood when friends from school may not have been retained and many adults will not be in employment and therefore unable to forge new friendships in this setting. Shared activities in the community are therefore essential in providing opportunities for adults with WS to develop new friendships outside of the family and foster a sense of connectedness (see Erasmus & van der Merwe, 2017 for the benefits of engaging in musical activities with others). Ensuring appropriate (inclusive, accessible) community services / activities should be a priority for policymakers.

Compared to earlier research, which described friendships as being difficult for people with WS to establish and maintain (Davies et al., 1998), this study emphasises a richer, more nuanced account. Therefore, there is need for more updated evidence on the topic of social experiences in WS. Future studies should explore how experiences differ relating to levels of independence, living situations, training/employment circumstances as well as individual differences in anxiety, communication, and personality factors. One important area for further investigation is peer relationships in the school context. Recent studies like Gillooly et al. (2021, 2022) have contributed important new insights about friendships at this developmental stage, but further research is needed to understand experiences across different educational settings (e.g., mainstream, or alternate provision) and, crucially, how schools can best support CYP with WS to form healthy peer relationships, feel included and accepted, and cope with negative experiences. The findings highlight that adults with WS (18-30 years) can provide valuable reflections about their relationships during the school years, therefore research with this cohort may be preferred rather than probing sensitive topics with children.

Further self-report research is needed. By listening to people with WS about their understanding of peer relationships, what they value in friends and common challenges, researchers and professionals will be better placed to develop support tools that are meaningful and align to the lives of people with WS. At the time of designing the research, a focus group approach was considered, but individual interviews were deemed more suitable given the sensitivity of the topic. In addition, in a group setting, there is a risk that more confident and sociable individuals dominate the conversation which can create bias towards the insights and experiences of individuals with stronger communication abilities. Importantly though, the same constraints and concerns about the accessibility of semi-structured interviews also apply to the focus group approach. Given these traditional qualitative methods are likely not inclusive for many people with WS, it is crucial that future self-report studies make thoughtful adaptations to better capture the views of people with WS who have a variety of intellectual and communication needs.

Researchers can play an important role in facilitating the information-gathering process by using more inclusive approaches. One example of a more inclusive method is the use of visual techniques such as photographs, videos or creating drawings. In the current study, asking participants to bring a photograph of a friend to the interview could have helped stimulate and ground the discussion, especially for adults who found it more challenging to reflect on their friendships. Taking this further, recent studies have embraced a visual method called 'photovoice' to capture insights from people with intellectual and communication challenges (Jurkowski, 2008; Overmars-Marx et al., 2018; Povee et al., 2014). While photovoice has been used as a tool in ID research on abstract concepts such as identity expression (Krisson et al., 2022), it has not yet been used in WS research. Indeed, WS research can benefit from the methods that have been employed in the ID literature more broadly. To flag just one example, in a study of 109 adults with varying levels of ID, Mayton et al. (2021) used video vignettes to explore reasoning about healthy and unhealthy relationships. In this study, the video vignettes were used pre and post an abuse prevention programme, however the vignettes themselves are thought-provoking and could be applied to WS research to better understand social decision making. The authors cocreated the vignettes and follow-up interview questions with professionals, adults with disabilities and their family members, with the storylines structured around concepts of trust, boundaries, differences between staff and friends.

It would also be interesting to consider a series of in-depth case studies that make use of several data collection techniques to give a more comprehensive and holistic account of friendships in WS. This could take the form of adapted interviews with people with WS (using techniques such as photovoice), but repeating these over an extended period or, in other words, bringing in a longitudinal element. This approach would provide a more nuanced understanding of how challenges with friendships may change over time and interact with contextual factors. While the individual perspective should be prioritised, longitudinal reports from someone familiar to adults with WS, such as a teacher or support worker, would complement the in-depth individual reports.

3.4.5. Conclusion

This study emphasised the value of listening to people with WS about their social relationships. We acknowledge that this was a small-scale study of nine adults with WS and further self-report qualitative work using more inclusive techniques is needed to understand how people with WS perceive and experience friendships in their own words. The study has generated valuable insights about aspects of social decision-making relevant to social vulnerability which will be explored later in the thesis. The childhood/adolescent years also emerged as significant with regard to social challenges and negative peer experiences, and the remainder of the thesis will focus on this stage of development.

4. CHAPTER 4: LESSONS FROM A CROSS-SYNDROME APPROACH TO THE WS SOCIAL PROFILE

4.1 Chapter Overview

The focus of enquiry now turns to consider salient features of the WS social profile that have relevance to the flow of social interactions and potential social vulnerability: the nature of eye contact (Paper 1) and social interaction styles (Paper 2). As outlined in Chapter 1, WS is often associated with having high social interest in others, termed "hyper sociability" in the literature (Doyle et al., 2004b; Jones et al., 2000). Claims of hyper sociability stem from observational and experimental reports of prolonged and intense eye gaze to faces (Jones et al., 2000; Laing et al., 2002; Mervis et al., 2003; Riby & Hancock, 2008, 2009a) and a propensity to approach others regardless of their familiarity (Dodd et al., 2010; Doyle et al., 2004b; Frigerio et al., 2006; Lough, Flynn, et al., 2016; Martens et al., 2009). However, studies to date have largely focused on capturing the 'presence' or 'quantity' of these behaviours (i.e., documenting excessive levels of eye contact and social approach). The current research aims to move beyond quantity to examine the quality or nature of these social behaviours in everyday life. It also aims to probe the proposal that increased social vulnerability in WS may be related to a combination of salient social differences and the presence of ID (Jawaid et al., 2012; Riby et al., 2017).

One question that this Chapter aims to explore is whether descriptions of prolonged eye contact and excessive approach thought to typify WS, describe most people with WS and the extent to which they are seen across other neurodevelopmental conditions. Therefore, to further understanding of social behaviours in WS more broadly, some of the research in this Chapter takes a cross-syndrome approach—including CYP with a range of neurodevelopmental conditions associated with differences in social behaviour, including WS but also autism, ADHD and FXS. It is worth noting that the cross-syndrome study design employed in the current Chapter places emphasis on diagnosis – participants were recruited based on diagnosis and analysis is largely at the diagnostic group level. In the time between designing the studies and writing up the thesis, there have been advances in the understanding of neurodevelopment which, in turn, have inspired more sophisticated approaches to research of this nature (Astle et al., 2021). I discuss this further in the Chapter 4 summary.

The Chapter presents two published papers. Paper 1 focuses on eye contact behaviour and includes two studies: the first is a secondary analysis of a dataset of children and adults with WS, and the second study extends the findings by taking a cross syndrome approach. Paper 2 focuses on social interaction styles across groups and the relationship with social vulnerability. Paper 1 study 2 and paper 2 use the same cross-syndrome sample and were part of a larger study of social interactions in children with and without neurodevelopmental conditions. The papers are presented in their published form, therefore there is some repetition across the two.

Ridley, E., Arnott, B., Riby, D. M., Burt, D. M., Hanley, M., & Leekam, S. R. (2022). The quality of everyday eye contact in Williams Syndrome: insights from cross-syndrome comparisons. *American Journal on Intellectual and Developmental Disabilities*, *127*(4), 293-312.

Ridley, E., Riby, D. M., & Leekam, S. R. (2020). A cross-syndrome approach to the social phenotype of neurodevelopmental disorders: Focusing on social vulnerability and social interaction style. *Research in Developmental Disabilities*, *100*, 103604.

Paper 1: Quality of Everyday Eye Contact in Williams Syndrome: Insights From Cross-Syndrome Comparisons

4.2. Abstract

Past research shows that individuals with Williams syndrome (WS) have heightened and prolonged eye contact. Using parent report measures, we examined not only the presence of eye contact but also its qualitative features. Study 1 included individuals with WS (n = 22, ages 6.0–36.3). Study 2 included children with different neurodevelopmental (ND) conditions (WS, autism spectrum condition, fragile X syndrome, attention-deficit/hyperactivity disorder) and children with neurotypical development (NT; n = 262, ages 4.0–17.11). Unusual eye contact features, including staring, were found in approximately half of the WS samples. However, other features such as brief glances were frequently found in WS and in all ND conditions, but not NT. Future research in ND conditions should focus on qualitative as well as quantitative features of eye contact.

Keywords: Williams syndrome, eye contact, neurodevelopmental condition, cross-syndrome comparison

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4.3. Introduction

Eye contact—the act of looking another person in the eyes—plays a powerful role in our everyday human social interactions. It signals mutual understanding and affiliation between people and promotes social-emotional relationships and communication (Emery, 2000; Falck-Ytter et al., 2015; Kleinke, 1986). Experiences of eye contact also elicit a range of cognitive and

affective reactions in the perceiver (for reviews, see Conty et al., 2016; Hietanen, 2018). In Western European societies, direct eye contact induces a range of positive evaluations (Kreysa et al., 2016; Willis et al., 2011). In contrast, a lack of eye contact may infer disinterest, whereas overly persistent eye contact may be deemed threatening and overly arousing (Akechi et al., 2013; Helminen et al., 2011). Therefore, when an individual's eye contact is reduced or overly prolonged, or unusual in some way, this may adversely affect social impression-formation with consequences for the development of social relationships (Morrison et al., 2020; Sasson et al., 2017).

Several theoretical perspectives have been put forward to explain how eye contact modulates cognition and behavior for those with neurodevelopmental (ND) conditions (for a review, see Senju & Johnson, 2009). The majority of these theoretical accounts apply particularly to the literature on autism spectrum condition (hereafter *autism)* and to the assumption by several different theories (e.g., social motivation theory Chevallier et al., 2012; hyperarousal model, Hadjikhani et al., 2017), that autistic individuals have diminished eye contact. One problem is that the evidence for this view rests mainly on studies that report reduced frequency or presence of eye contact. However, there has been remarkable neglect in considering the nature of the quality of eye contact, which could possibly lead to a different understanding of eye contact in individuals with ND conditions. One reason for the past focus on quantity rather than quality is that much of the research knowledge on eye contact stems from a broader laboratory-based research tradition on eye gaze more generally, which tends to equate looking at the eyes of computerized facial stimuli with "eye contact." Although this paradigm affords a high level of experimental control, the passive viewing of socially relevant stimuli is very different from how eye contact is experienced in everyday dyadic social interactions (see Kingstone, 2009). Research has shown that the realism of the stimuli used in social attention research (e.g., static versus dynamic images; isolated faces versus multiple faces in a social scene), impacts on eye contact (e.g., Hanley et al., 2012; Speer et al., 2007). Consequently, researchers have emphasised the importance of studying everyday situations to understand social attention in real-life interactive situations (e.g., Hanley et al., 2015; Kingstone, 2009; Risko et al., 2012). In the current investigation, we examine both the presence and quality of everyday eye contact of individuals with ND conditions, using the caregiver's perspective of eye contact.

4.3.1. Eye Contact Behavior in Williams Syndrome

Williams syndrome (WS) is a genetic ND condition commonly associated with a heightened desire for social contact (termed hypersociability; for a review of the WS social phenotype, see Thurman & Fisher, 2015). Indeed, WS is a very important ND condition to study various aspects of social behavior because its genetic basis is well-defined (hemizygous deletion of ~25–28 genes on chromosome 7q11.23; Ewart et al., 1993), therefore research with this group has the potential to inform debate about gene-brain-behavior links and further our understanding of the "typical" social brain. Consequently, the WS social profile has garnered a significant amount of research attention at the level of both brain and behavior. For example, evidence that WS is associated with structural and functional atypicalities in key areas of the *social brain network* known to activate in response to eye contact, such as the amygdala (Haas et al., 2009; Martens et al., 2009) and fusiform face area (FFA; Golarai et al., 2010), has informed understanding of how different features of the WS social phenotype may be subserved by neural substrates (for a review, see Haas & Reiss, 2012). At the behavioral level there has been a great deal of interest in capturing various aspects of social behavior in WS, including eye gaze and eye contact behavior. The predominant evidence of gaze behavior in WS comes from face scanning and eye-tracking studies that have examined eye gaze behavior towards images or movies on screen. These studies show that the face, particularly the eye region, attracts and holds the attention of individuals with WS for longer than is typical for young children, adolescents and adults (Porter et al., 2010; Riby & Hancock, 2008, 2009a, 2009b). This tendency for heightened, prolonged looking to faces and eyes has been linked to a lack of habituation to faces (A. M. Järvinen et al., 2012), to physiological reactivity, and to attentional mechanisms related to arousal, suggesting the possibility of hypo-arousal in this group (Doherty-Sneddon et al., 2009; Riby et al., 2012; Skwerer et al., 2009, 2011).

Beyond laboratory studies using eye tracking and measuring gaze to computerized images, a few other observational studies have also reported that young children with WS (< 5 years old) show intense and prolonged looking in real-world settings during interactions in clinics (Mervis et al., 2003) and with experimenters (W. Jones et al., 2000). Although studies using a clinical measure, the Autism Diagnostic Observation Schedule (ADOS; Lord et al., 2000), have reported that up to 52% of children with WS had "definite abnormality" with eye

contact (Klein-Tasman et al., 2007, 2009), we know little about the nature of the unusual eye contact as the ADOS assessment does not capture quality features of eye contact. Given this limited evidence of prolonged, intense eye contact in naturalistic settings, it is still not established whether this quality of eye contact is common in individuals with WS, if it is a feature distinctive to WS, or is frequently found in other ND conditions. Research that examines eye contact behavior in WS alongside other ND conditions will help to identify features of eye contact that may be particularly distinctive to WS (syndrome-specific) or shared across diagnostic groups (syndrome-general). See Asada and Itakura (2012) for further discussion.

4.3.2. Eye Contact Behavior Across ND Conditions

Although WS has been characterized by social interest associated with a heightened and prolonged presence of eye contact, other ND conditions, particularly autism, have traditionally been associated with reduced presence of eye contact (Asada & Itakura, 2012; Senju & Johnson, 2009). Reduced eye contact, in turn, has been connected to a lack of social interest (Chevallier et al., 2012), an assumption that has been challenged by those with subjective, lived experience of autism (Jaswal & Akhtar, 2019) who argue that reduced quantity of eye contact does not necessarily equate with lack of interest. We propose that the clarification of this issue has been hampered by a single dimensional approach to the understanding of eye contact that conflates presence and quality of eye contact. Characterizing eye contact by a single dimension leads to a view that reduced eye contact is poor eye contact and increased eye contact is good eye contact, an assumption that tends to polarize the social phenotypes of ND groups into opposite profiles (see Asada & Itakura, 2012 for review of the autism/WS distinction). By considering multiple qualitative features of eye contact in everyday life contexts across ND conditions, the current study attempts to move away from examining eye contact through a quantitative, single dimensional lens.

Like studies of WS, much previous research on eye contact in autism has also tended to focus on its presence or degree. Eye-tracking studies show that some autistic individuals spend less time than is typical attending to face areas and eye areas on a screen (Sasson et al., 2007; Shic et al., 2011). For reviews of the autism eye tracking literature see Guillon et al. (2014) and Papagiannopoulou et al. (2014). Both eye tracking studies (e.g., Hanley et al., 2014, 2015) and face-to-face observational studies (e.g., S. R. Leekam & Ramsden, 2006) also find differences in

attentional orienting in autistic individuals compared to neurotypical (NT) and intellectually disabled peers, and that reduced eye contact is very dependent on context (R. M. Jones et al., 2017; Kasari et al., 1993). Furthermore, reduced presence of eye contact has been associated with failure to automatically attend to the salience of social cues, rather than to active avoidance of others in several eye tracking studies (Hanley et al., 2013; Klin et al., 2002) and has been associated with overarousal (Hadjikhani et al., 2017). First-hand insights from autistic adults also describe reduced eye contact as a strategy for arousal reduction (McGlensey, 2016; Trevisan et al., 2017) and report the use of qualitative strategies used such as non-eye fixation, blurring focus, and strategic fixation (Trevisan et al., 2017). However, the perceived experience of unfocused eye gaze in these first-hand accounts has not been measured from another person's perspective and the research reported here targets this by exploring parents' perspective of eye contact taken from their everyday experience.

Although autism and WS are two frequently studied ND conditions in the eye gaze and eye contact literature, these are not the only ND conditions that are associated with social differences related to eye contact. Like WS, fragile X syndrome (FXS) is a genetic condition associated with intellectual disability (ID) and impacts upon social functioning. The FXS social phenotype can be summarized as a mix of both social approach (Cornish et al., 2008) and social withdrawal behaviors (Roberts et al., 2007, 2019), alongside heightened social anxiety (Crawford et al., 2017). Studies to date consistently show that FXS is associated with gaze avoidance (Hall et al., 2006, 2009, 2010; Hessl et al., 2006), which increases when the interlocutor is unfamiliar (Hall & Venema, 2017), but which may improve over the course of an interaction ('warm up effect'; Hall et al., 2009; Roberts et al., 2007). People with FXS show a tendency for shorter gaze episodes towards another person and for brief glances when the person is looking elsewhere rather than making direct eye contact (Cohen et al., 1991; Hall et al., 2006, 2015; Klusek et al., 2020).

Although social difficulties are not part of the diagnostic criteria for attention-deficit/hyperactivity disorder (ADHD), there is a growing literature reporting socio-cognitive difficulties, problematic peer relationships (for reviews, see Gardner & Gerdes, 2015; Soucisse et al., 2015), and high rates of social vulnerability (Ridley et al., 2020b). Studies reporting on aspects of gaze orienting and attention indicate diminished attention to socially relevant

information (Airdrie et al., 2018; Marotta et al., 2014, 2017; Muszkat et al., 2015), however everyday eye contact behaviors in this population have scarcely been documented. One relevant study using the ADOS found that unusual eye contact was reported statistically more frequently in a sample of autistic children compared to children with ADHD (Grzadzinski et al., 2016). Nevertheless, 31% of the ADHD sample were reported to have "abnormal" eye contact, yet the nature of the unusual eye contact was not described.

4.3.3. The Current Study

In this study, we explored the quality of everyday eye contact in individuals with WS in comparison with each of these ND groups using parent report. First, we studied the single dimension of "presence" (or degree of presence). Second, we included a specific measure of different qualitative features that have been associated with different ND conditions. A two-stage approach was adopted. First, given the gap in the literature on the quality of eye contact in WS, particularly from a parent perspective, Study 1 used a set of standard interview questions to explore the qualitative features that parents might observe in their child's everyday eye contact. Although we expected a high presence of eye contact in WS, we also expected, given the findings of Mervis et al. (2003) and Jones et al. (2000), that parents might observe a quality of intense, prolonged eye contact (equated with staring in this study). However, we did not know whether other qualitative features would be frequently seen or the extent to which staring would be found across all WS individuals and across all ages.

In Study 2 we used a parent questionnaire method to examine further the eye contact quality features used in Study 1 as well as other qualitative features, making cross-syndrome comparisons across children with WS, autism, FXS, and ADHD. In addition, we included a NT comparison group to examine whether particular qualitative aspects of eye contact were specific to the presence of an ND condition. The research will contribute new evidence to an ongoing debate about the similarities and differences in eye contact in ND conditions, particularly between WS and autism. The study will also add new findings to the literature on eye contact behavior in FXS and in ADHD, a topic that has received limited attention.

Paper 1, study 1: Examining the Nature of Eye Contact in WS

The first study explored the presence and quality of eye contact used by individuals with WS in their everyday life. A semi structured set of interview items was used that enabled parents to describe both the presence of eye contact and qualitative features such as brief glances, staring behavior and blank, unfocused gaze. The individual's developmental level of language and visuospatial ability was also recorded during the interview.

4.4.Methods

4.4.1. Participants

Twenty-two individuals with WS and their families were recruited throughout the North of England and Scotland following institutional ethical approval and study approval from the Professional Advisory Panel of the Williams Syndrome Foundation. Informed consent was given by all participants. The researcher conducting the interviews with parents, BA, was trained in its use by SL. In all cases, it was the primary caregiver who completed the interview with the researcher, either at home or in the University.

Individuals were sampled across a wide age range. At the time of the parent interview, individuals with WS ranged between 6 years 0 months and 36 years 3 months of age (male, 10, female 12), M_{age} 196 months (*SD* 98 months). All individuals were attending school, college, or work placements, including five in mainstream school with support, 10 in special educational provision, and five in supported work or college (two had information missing). All individuals had previously been diagnosed phenotypically by clinicians and their diagnosis had been confirmed with positive fluorescent in situ hybridization (FISH) testing.

Information on language delay, and current language and visuospatial ability was collected from parents during the interview. As Table 4.1 shows, the group was developmentally delayed. In terms of language delay, 78% of individuals (14/18, four missing) were late to use 2-3 phrases and 84% (16/19, three missing) were late to understand word meanings. In terms of current language ability, 21 participants (one missing) had sentence-level expressive language and all but one participant had sentence-level receptive language (simple or complex sentences). However, only two-thirds (14 individuals) used expressive language at the highest level (complex age-appropriate grammatical constructions) and only one third (seven individuals)

understood language at this level. Visuospatial data (two missing) showed that only three individuals (15%) had age-appropriate level of current skill.

Table 4.1

Data for Each Individual with WS for DISCO items Assessing Eye Contact and Language and Visuospatial Skills Level

Age in months	M/F	Language Delay		Age-appropriate level of current skill			Eye contact present	Unusual quality of eye contact (marked or frequent)		
		Late to use 2-3 phrases	Late to understand word meanings	Expressive language Level 1-9 [a]	Receptive language Level 1-7 [b]	Visuospatial skill Level 1-12 [c]		Brief glances	Blank unfocused gaze	Staring
72	F	Yes	Yes	8	5	9	+	+	+	+
89	M	Yes	Yes	8	5	5				
100	F	No	No	9	6	9	+			
101	M	–	–	9	3	12	+			
106	M	Yes	Yes	9	7	10	+	+		
115	F	Yes	Yes	7	6	10	+			
124	F	Yes	Yes	8	7	10	+			
153	M	Yes	Yes	9	7	9	+		+	
159	M	Yes	Yes	9	7	9	+			
161	F	Yes	Yes	9	5	12	+			
172	F	Yes	Yes	9	4	10	+	+	+	+
193	M	No	–	9	4	10	+	+	+	+
193	M	–	Yes	8	5	8	+	+	+	+
205	F	Yes	Yes	8	4	–				
206	M	Yes	Yes	9	4	12	+		+	+
210	F	No	Yes	9	7	6	+			
258	F	Yes	Yes	8	5	8	+			
277	M	Yes	Yes	9	5	6	+	+		
286	F	–	No	9	7	8	+	+	+	+
301	M	Yes	Yes	9	7	3	+			+
396	F	No	No	9	6	8	+			+
435	F	–	–	–	6	–	+	+	+	+

Note. DISCO = Diagnostic Interview for Social and Communication Disorders; M = male; F = female. Dash sign (–) = parent data was not available. Cells with plus sign (+) indicate endorsement of either (a) presence of eye contact and/or (b) unusual quality of eye contact at a marked level.

[a] Language expression: 0–2 = No speech or babbles; 3–4 = Says names for things only; 5 = says phrases of 2 words only; 6 = Says longer phrases; 7 = Uses spontaneous sentences, present tense only; 8 = Uses sentences/phrases including "but" and "because"; 9 = Uses past, present, and future tenses and complex grammatical constructions.

[b] Language comprehension: 0–1 = No response or responds to name only; 2 = Understands simple words from phrases in context (learned from gestural cues, e.g. time for bed); 3 = Knows the meaning of some words and can responds (e.g., "Give me your cup"; 4 = Follows instructions involving 2 new objects "Put the doll on the chair"; 5 = Can reliably follow instruction to fetch two or more objects from outside of the room; 6 = Understands a sequence of commands; 7 = Understands instructions involving decisions (conditionals) "See if my phone is in my bedroom and if not look for it in the bathroom."

[c] Visuospatial skill: 0 = Does not hold objects in hands; 1 = Holds objects in hands; 2 = Examines objects; 3 = Handles objects; 4 = Rolls toys on floor; 5 = Builds tower of 2–5 bricks; 6 = Builds tower of 6 bricks; 7 = Arranges objects in size order; 8 = Completes puzzle 6 pieces; 9 = Completes puzzle 10 pieces; 10 = completes puzzle 20–30 pieces; 11 = Completes puzzle 50 pieces; 12 = completes puzzle 150 pieces.

4.4.2. Materials and Procedure

A research form of the Diagnostic Interview for Social and Communication Disorders (DISCO; S. Leekam, 2021; Wing et al., 2002) was used. The DISCO is a semistructured clinical interview used with parents and carers. It is most commonly used for parents of autistic individuals of any age, but is also suitable for use with individuals with other ND conditions and includes items applicable for ADHD, WS, and FXS. The current interview followed the format of previous research that has used and published subsets of DISCO items (e.g., Prior et al., 1998). The eye-contact and language items used in Study 1 are included in the published DSM-5 algorithm (Kent et al., 2013) and DISCO ICD-10 Childhood Autism algorithm (S. R. Leekam et al., 2002), and the visuospatial skill item is a non-algorithm item in the DISCO (Wing et al., 2002). Each of the four eye contact items and each of the language and visuospatial items has a high level of interrater reliability ranging from $k = .89$ to $k = 1.00$ (Wing et al., 2002).

Information on language delay and current language and visuospatial ability was collected using age-appropriate scales within the DISCO (see Table 4.1). Items from the current language scales have been published (Honey et al., 2007). Age-equivalent visuospatial skill was indicated by the ability to construct complex puzzles according to age group. Language delay (use of phrases, comprehension of word meanings without visible cue) was indicated by delay after 48 months old. Age-appropriate current sentence skills were recorded when complex grammatical constructions and past, present, and future tense were present.

Information on the presence and quality of eye contact was collected using four eye contact items and scored using the DISCO syntax rules that have previously been applied in both interview (Kent et al., 2013) and questionnaire (C. R. G. Jones et al., 2020) research formats. The first item related to the presence of eye contact. The interviewer asked the caregiver whether it was easy to get eye contact with the individual. The item was scored as "eye contact present" if the answer was *yes*, even if the eye contact given was described as unusual in some way, and *no* if the parent reported little or no eye contact. The next three questions related to the quality of eye contact seen as usual behavior on an everyday basis. These were whether the individual (a) makes eye contact only in brief glances (e.g., out of the corner of eyes, but not for the purpose of gaining another's attention); (b) whether the individual has a blank, unfocussed gaze; and (c) whether the individual stares too long and hard, perhaps holding another person's face to make

eye contact and/or looking closely into another's eyes. Each item was sequentially assessed by the interviewer who established whether this was a typical behavior for the individual (used routinely with adults and age peers) and whether it was marked (or frequent), occasional, or rarely/never seen. Following DISCO syntax rules, each item was scored as having a markedly unusual quality if judged to be "marked" (brief glances), "marked and frequent" (blank, unfocused gaze), and "marked staring or otherwise inappropriate" (staring) in that individual, but not if the feature was sometimes, rarely or never seen.

4.5.Results and Discussion

Case-by-case profiles of eye contact patterns are shown in Table 4.1 together with age and language/visuospatial level. The cells that include the plus symbol (+) indicate endorsement of a score for each individual (e.g., presence of eye contact or a marked quality of eye contact) whereas the blank cells indicate nonendorsement. Results showed that 20 (91%) individuals (male n = 9, female n = 11) gave eye-contact easily (even if inappropriately), whereas two (9%), gave little or no eye contact. Subsequent analyses focused on these 20 individuals, 13 of whom (65%; male n =7, female n = 6), had a "marked" unusual quality of eye contact, as indicated by at least one out of three unusual features—brief glances, unfocused gaze, or staring. Brief glances at marked level were endorsed by eight (40%), unfocused gaze by eight (40%), and staring was endorsed by 10 (50%; see Table 4.1). Six individuals had marked scores for all three features.

Further exploration was made of the characteristics of the 13 individuals with marked unusual quality of eye contact. More than half, nine of the 13 (69%), had early developmental delay in understanding of word meanings (two had no delay, two had missing data), and of these nine individuals, all but one (data missing) were also delayed in using two-to-three word phrases. The gender distribution was also approximately equal for endorsement of each of the three eye contact quality features.

To explore how each of the unusual eye contact quality features was affected by other variables (current age, current language level, and visuospatial level), Mann-Whitney tests were carried out with the 20 participants who were reported by parents as giving eye contact easily. For each analysis, the group of individuals with "marked" responses was compared with the group without marked features (scoring *sometimes* or *rarely/never*). Analyses were repeated to

examine brief glances, unfocused gaze, and staring features separately and Bonferonni adjustment was applied to accommodate multiple comparisons (.05/3, $p = .02$). An age difference was found (see Table 4.1), as the group with marked staring features was older, having a mean age of 20 years 11 months ($M_{age} = 251.20$ months, $SD = 108.37$, $n = 10$) whereas those without marked staring features had a mean age of only 12 years 6 months, ($M_{age} = 150.60$ months, $SD = 56.44$, $n = 10$), $U = 99.0$, $p < .010$. However, there were no age differences for the other unusual quality features (blank, unfocused gaze, $p = .92$; brief glances $p = 1.00$). No differences were found in visuospatial ability, current expressive and receptive language for those with marked unusual eye contact quality.

In summary, Study 1 used a set of parent interview questions for the first time, to explore the qualitative features of everyday eye contact in individuals with WS. The results showed positive presence of eye contact by 91%, together with an unusual quality of staring in 50%. This pattern supports previous evidence from laboratory and clinic studies (W. Jones et al., 2000; Mervis et al., 2003). However, in addition, new evidence was found. Results showed that staring was more frequent among older ages. However, staring was not an exclusive or predominant quality feature and parents endorsed features of unusual quality of eye contact beyond staring, including brief glances and unfocused gaze. These were reported by parents in 40% of individuals with at least one of these features often co-occurring alongside staring.

Paper 1, study 2: Comparing Eye Contact in WS, Other ND Conditions, and NT Development

To gather a larger sample of reports, Study 2 asked the same questions as in Study 1 but used a questionnaire measure with parents of children with WS. In addition, we adopted a cross-syndrome approach to examine potential syndrome-specific aspects of eye contact behavior in WS, autism, FXS and ADHD, as well as NT development.

Given the research findings reviewed above and the results of Study 1, we predicted a high presence of eye contact in WS compared with other ND groups. We also predicted that unusual qualitative features would be found in WS and also in the other ND groups, with staring reported for children with WS (Klein-Tasman et al., 2007; Mervis et al., 2003), brief glances and avoidance reported for FXS children (Klusek et al., 2020) and a blurred or unfocused gaze

(Trevisan et al., 2017) and/or avoidant gaze (Senju & Johnson, 2009) reported for autistic children. Given the results for the WS group in Study 1, we expected not only staring but also other qualitative features to be reported. However, it was not known whether other ND groups might have particular distinctive and predominating qualitative features.

4.6.Methods

4.6.1. Participants

Parents/caregivers of children were recruited for this study as part of a larger investigation of social interaction behaviors in children with and without ND conditions. Survey responses were received for 276 caregivers/parents in total. Responses were included for data analysis based on the child's primary diagnosis if the parent reported that their child (a) had a primary diagnosis of WS, autism, FXS, or ADHD, or had NT development and did not have an ID or a statement of Special Educational Need (SEN); and (b) was aged 4–17 years. Of the 276 respondents, 262 met the aforementioned inclusion criteria and fell in the following groups: WS (n = 29), autism (n = 29), FXS (n = 18), ADHD (n = 36), and NT (n = 150). None of the participants in Study 1 were included in the WS sample in Study 2.

Table 4.2 shows the child characteristics per group. Fifty-nine percent of the full sample were males. The ND groups (apart from the WS group) included significantly more males than the NT group. Of the ND groups, FXS included significantly more males than the WS group. However there was no significant difference in the distribution of genders between the other ND groups. The ND groups differed in parent-reported ID status as seen in Table 4.2, $\chi^2(df = 3) = 50.98$, $p < .001$. As expected, the WS and FXS groups included a significantly higher frequency of children with ID compared to the autism and ADHD groups (but no difference in the frequency of ID status between WS and FXS, or between autism and ADHD). For receptive language ability, the WS and FXS groups had a higher frequency of children without full sentences compared to the autism and ADHD groups. Likewise, for expressive language ability, the WS and FXS groups had a higher frequency of children without full sentences compared to the ADHD group, but no difference with the autism group.

Table 4.2

Demographic Characteristics of the Sample (Percentage Reported) Split by Diagnostic Group

Demographic variables	Autism (*n* = 29)	WS (*n* = 29)	ADHD (*n* = 36)	FXS (*n* = 18)	NT (*n* = 150)
Males/females/prefer not to say	72/28/0	59/41/0	78/19/3	94/6/0	48/51/1
Age (months)					
M (*SD*)	127 (28.4)	100 (36.3) [a]	127 (38.8) [b]	118 (36.9)	107 (45.8) [c]
Range	59-187	48-204	54-179	54-197	48-215
Presence of an intellectual disability	21	90	28	89	0
Expressive language					
None	3	7	0	11	1
Single words	3	7	0	17	0
Simple phrases	7	24	6	33	0
Full sentences	86	62	94	39	99
Receptive language					
None	0	0	0	6	0
Single words	0	7	0	0	0
Simple phrases	17	28	6	28	0
Full sentences	83	66	94	67	100

Note. WS = Williams syndrome; ADHD = attention-deficit/hyperactivity disorder; FXS = fragile X syndrome; NT = neurotypical.

[a] Missing data (*n* = 1). [b] Missing data (*n* = 1). [c] Missing data (*n* = 1).

4.6.2. Procedure

Separate advertisements invited parents of (a) children with a diagnosis of WS, autism, ADHD, or FXS; and (b) parents of children with NT development, to complete an online survey about their child's social interactions and were distributed via a university research participation database for local families, social media, and United Kingdom charity networks. Informed consent was obtained from all participating caregivers/parents following positive ethical opinion from the University ethics committee. Parents did not receive financial remuneration.

4.6.3. Materials

Parents/caregivers reported on their child's eye contact behaviors as part of a larger bespoke survey on social interactions throughout development via online survey software (www.onlinesurvey.ac.uk). In addition to the questions addressing the research aims, parents provided demographic information concerning the child's date of birth, gender, diagnostic status, and ID status. To gather information about language abilities we included the following questions "Does your child use language to communicate?" (*none*; *single words*; *simple phrases*; *full sentences*), and "Does your child understand language?" (*none*; *single words*; *simple phrases*; *full sentences*).

The eye contact items corresponded exactly with interview items of Study 1 but the method was distinct as the items were presented in a fixed response format more suitable for a questionnaire. Items were presented as statements with options to select as follows: Item 1 "He/she makes eye contact (even if inappropriate, learned, or occasional)" with a response option *yes/no*. The next set of items relating to quality of eye contact, unlike Study 1, were not presented sequentially. Instead, they were presented as a forced choice format and caregivers could select only one item in response to the following question: "Please tell us more about the quality of eye-contact. Which of the following applies most usually?" Six response options were offered (shown in full in Table 4.3). In addition to the three items in Study 1 (staring; blank, unfocused gaze; brief glances), two other items were offered to capture a wider range of qualitative features that might be seen in any of the children. These were (a) "always appropriate and natural" and (b) "avoids eye contact." One of the six (indicating the one that applies most usually) could be ticked. The next item, "If none of the above applies you can give more

information here if you wish (this is optional)" allowed parents to elaborate on their child's eye contact behavior if it did not easily fit one of the prespecified categories

4.7.Results and Discussion

The first hypothesis, that there would be a high presence of eye contact in WS compared with other ND groups, was not supported. Instead, results showed that the vast majority of all children with a ND condition engaged in eye contact. Although as many as 93% (*n* = 27/29) of parents of children with WS endorsed this item, similar to Study 1, endorsement was also high for autism: 86% (*n* = 25/29), FXS: 72% (*n* = 13/18) and ADHD: 86% (*n* = 31/36). A Chi-Square test of Independence showed no significant difference between the four ND groups, χ^2 (3) = 3.98, *p* = .264. Nevertheless, the strong presence of eye contact in all ND groups (96/112, 86%), was still lower than for the NT sample, virtually all of whom were endorsed as showing eye contact (146/149, 98%, one missing response), *p* < .001 (Fisher's Exact Test).

The second hypothesis was that unusual qualitative features would be found in WS and in other ND groups. This hypothesis was examined in several ways. Table 4.3 presents the distribution of responses (i.e., children with endorsement of *yes* to Item 1 reporting presence of eye contact). First, taking the responses for "eye contact always natural and appropriate" (Column 3 of Table 4.3), this was the most highly endorsed option for 87% of the parents of NT children and significantly higher than endorsement for the ND sample as a whole (31%; *p* < .001 Fisher's Exact Test) or for the WS group alone (44%; *p* < .001 Fisher's Exact Test). This evidence supports the prediction that even when children with a ND condition do give eye contact, the quality of their eye contact is not predominantly natural or appropriate. Nevertheless, the WS group did show a significantly higher frequency of "appropriate" eye contact compared to the autism group (12%; *p* =.01), but no difference compared to FXS (15.4%; *p* =.09) or ADHD groups (42%; *p* = 1; 2 x 2 Fishers Exact Chi Square analyses, with WS compared with each ND group for responses to the "appropriate" option versus the remaining response options).

Second, initial examination of the pattern of unusual qualitative features revealed that the option "avoids eye contact" was rarely selected for any of the ND groups. This was surprising, given descriptions of avoidance in the autism and FXS literature (Hall et al., 2006; Senju & Johnson, 2009), but it demonstrates parents' interpretation of their child's eye contact quality when selecting from different behavioral options.

Table 4.3

Quality of Eye Contact Behavior Endorsed by Parents in Each Group

Group	*n* [a]	Quality of eye contact applied most usually					
		Eye contact always appropriate and natural	Brief glances	Blank, unfocused gaze	Staring	Avoids eye contact	None of these apply
WS	27	12 (44.4)	5 (18.5)	1 (3.7)	7 (25.9)	1 (3.7)	1 (3.7)
Autism	25	3 (12)	11 (44)	2 (8)	1 (4)	3 (12)	5 (17.2)
FXS	13	2 (15.4)	9 (69.2)	0 (0)	1 (7.7)	0 (0)	1 (7.7)
ADHD	31	13 (41.9)	13 (41.9)	1 (3.2)	1 (3.2)	2 (6.5)	1 (3.2)
NT	143 [b]	124 (86.7)	11 (7.7)	1 (0.7)	0 (0)	2 (1.4)	5 (3.5)
Total ND sample	96	30 (31.3)	38 (39.6)	4 (4.2)	10 (10.4)	6 (6.3)	8 (8.3)

Note. WS = Williams syndrome; FXS = fragile X syndrome; ADHD = attention-deficit/hyperactivity disorder; NT = neurotypical; ND = neurodevelopmental. Percentages are presented in parentheses.

[a] Parents who reported *yes* to Q1 about the presence of eye contact. [b] Of the 146 parents in the NT group who reported *yes* to Q1, 3 data points were missing.

Subsequent analysis therefore focused on the three unusual quality descriptors from Study 1 (staring; brief glances; and blank, unfocused gaze). Results showed that the majority of parents in the ND sample selected one of these features as the most usual qualities of their child's eye contact (ranging from 48% to 77% of each group and 54% of the total ND sample) in comparison to only 8% of the NT group. A Fisher's Exact Test confirmed higher endorsement any of these three (see Table 4.3) in the ND groups taken together (54%) compared to the NT group ($p < .001$).

Given the result of Study 1, we did not predict specificity or dominance in one qualitative feature (e.g., staring) for the WS group. However, it was not known whether other ND groups might have specific qualitative features that are distinctive or dominating. To analyze this, a series of 2 x 2 Fishers Exact Chi square analyses were carried out, using only the samples endorsed with brief glances, unfocused gaze or staring (totals from columns 4–6 of Table 4.3; i.e., WS $n = 13$; autism $n = 14$; FXS $n = 10$; ADHD $n = 15$). The categories "unfocused, blank gaze" and "staring" were collapsed together (due to small expected frequencies) and compared with "brief glances." This confirmed a different distribution of response: brief glances were more frequently selected for autism (78.6%, $p = .05$), FXS (90%, $p = .03$) and ADHD (86.7%, $p = .02$) groups compared to the WS group (5/13, 38.5%), whereas the presence of staring behavior (with unfocused gaze) was more frequently endorsed in the WS group (7/13, 61.5%) This finding supports previous descriptions of persistent and prolonged eye contact in young children (Klein-Tasman et al., 2007; Mervis et al., 2003), showing these behaviors are also found in older children and adolescents. In summary, although dominance of one specific qualitative feature was neither predicted nor found, the results indicate that when given a forced choice format, a small but significant proportion of parents of children with WS tend to preferentially select "staring/unfocused gaze" in favor of "brief glances" whereas the majority of parents of all other ND groups select "brief glances."

Only a very small minority of parents selected the option "none of the above apply" (5.4% of the full sample: NT $n = 5$, ND $n = 8$), indicating that the options provided were mostly consistent with the range of parent experiences. All of these parents also answered "if none of the above apply please leave further information here (this is optional)." The majority of the free-

text responses (NT $n = 4$, ND $n = 5$) reported that the child might show more than one type of eye contact behavior according to situational or person context.

Follow-up analyses examined the relationship between eye contact behavior, first for presence and then for quality ("blank, unfocused gaze" collapsed with "staring") and the demographic variables: Age, Gender, ID-status (*yes/no*), and language-status (*with/without full sentences*) analysed using Chi-square tests. Small samples limited the opportunities for finding significant associations with other demographic variables throughout. No significant associations were found between type of unusual eye contact and language ability (expressive or receptive), ID, gender, or age, and it was not meaningful to test the comparison between staring and age found in Study 1 because of the sample sizes.

4.8.General Discussion

Eye contact strengthens the communication process during human social interaction and shapes our judgements about others (Conty et al., 2016; MacDonald, 2009). For this reason, it is important to understand how eye contact manifests in everyday life for those with WS and with other ND conditions. The results of Study 1 and 2 show that parents of individuals with WS, nearly all of whom described their child as making eye contact, also described their child's eye contact as unusual rather than natural and appropriate. Our findings support previous evidence showing prolonged and intense looking in individuals with WS, and Study 1 also found first evidence of an association between staring and increased age. However, importantly, staring was not the only type of unusual feature as many parents also reported the use of brief glances and blank, unfocused gaze.

The cross-syndrome comparison with other ND groups in Study 2 revealed surprising insights. First, the research literature for autism and FXS often describes individuals as having reduced or avoidant eye contact. But parents of these children, who must be looking at their children's eyes on an everyday basis, tend not to describe a lack of eye contact. Like the parents of children with WS, most parents in the autism, FXS, and ADHD groups reported that their child does make eye contact; however, when given different options to indicate the quality of that eye contact, they indicated an unusual quality to it. The most frequently endorsed feature for parents of all three groups was brief glances, whereas this was not the case for the parents of the WS group who more frequently than the other groups, selected staring or unfocused gaze in this

forced choice question format. However, staring/unfocused gaze was not unique to WS and many parents also endorsed brief glances in their children with WS.

This study contributed to the literature by moving beyond the conventional measurement of eye contact as being either present or absent, in varying degree. By separating the measurement of "presence" from an additional measurement of "quality," we found different results from studies that have used a single measure of presence of eye contact as an indicator that eye contact is good versus poor. In contrast, our results suggest that nearly all individuals with WS (Study 1), and nearly all children whether WS, autism, FXS, or ADHD (Study 2), do make eye contact even if in an unusual manner. The type of this unusual quality also seems to be consistently identified by parents as taking the form of brief glances, unfocused gaze or staring, as evidenced by the fact the "none apply" was rarely endorsed in Study 2. In Study 2 we also found that the option of "avoids eye contact" was rarely endorsed by parents in preference to these other three items. However, it is not clear why they made this preference. Possibly, the choice of one of six forced choice options constrained them and resulted in few cases of "avoids eye contact." Further research is needed to test out why parents did not choose "avoids" in preference to other items and to evaluate whether this is because it is not a feature of eye contact according to caregiver perspective, or whether it is because other types of contact behavior are merely more common.

We learn from the cross-syndrome comparison design of Study 2 that unusual eye contact is found across multiple ND conditions, rather than specific conditions being associated with specific patterns of eye contact. It is unclear the extent to which this is due to direct yet variable effects of the ND condition on eye contact, or whether these behaviors are differently acquired through factors which may vary but show commonalities across ND conditions, along with external and internal environments. To disentangle this further, the next stage of research enquiry may benefit from moving towards a more transdiagnostic design. In a recent review on the transdiagnostic model for understanding neurodevelopment, Astle et al. (2021) outline a spectrum of study designs that can offer transdiagnostic insights, which vary in the emphasis placed on diagnostic status. Based upon this classification, studies like ours that test for syndrome-specific associations offer value in elucidating where aspects of cognition and behavior crossover different ND conditions, or are distinctive. However, this traditional,

categorical approach is problematic as it rests on the assumption that ND conditions are homogenous and have clear-cut boundaries; an assumption that does not match up with the clinical reality. Consequently, researchers have argued for the need to reconceptualize neurodevelopment and embrace more transdiagnostic features of design throughout the research process (Astle et al., 2021; Casey et al., 2014; Sonuga-Barke & Thapar, 2021). In the case of research on eye contact, there would be value in following a model similar to that used in research areas of cognition and learning (e.g., Bryant et al., 2020; Mareva et al., 2019), by recruiting a large heterogeneous sample of individuals with ND conditions known to impact on social attention and social interaction, and stratifying on the basis of particular eye contact styles (see the 'diagnostic-blind' approach in Astle et al., 2021).

An important consideration for studies such as ours that do compare groups according to diagnostic label is that children and adults who receive a diagnosis of any ND condition may also receive other associated diagnoses (Cleaton & Kirby, 2018). Autism frequently co-occurs with other conditions and as difficulty with eye contact is a diagnostic feature of autism, this might explain eye contact differences in other conditions as well. As information on co-occurring autism diagnoses had been collected at the time of recruitment, we were able to carry out further analysis of those with associated diagnoses (WS n = 2, FXS n = 9, ADHD n = 9). The pattern of results for presence of eye contact and for unusual quality of eye contact remained unchanged; therefore, significant effects of an associated autism diagnosis were not evident in this study, but given the small sample sizes, future research designs should test more fully for the effect of co-occurring diagnoses on eye contact presence and quality (see model of study designs outlined in Astle et al., 2021).

4.8.1. Limitations

There are several important limitations to this study. Although the results from parent reports in these studies appear striking, it should also be remembered that there are problems using subjective methods of this kind. Parents were aware that this was an interview or questionnaire studying social interactions in those with ND conditions and responses could be attributed to a response bias. Therefore, a recommendation for future research would be for the inclusion of different measures that combine insights from direct observations and experiments, along with multi-informant reports of everyday eye contact. Teacher insights would make a

valuable addition given teachers are interacting with children on a regular basis but within a different setting compared to parents.

Another limitation was that the measure adapted from Study 1 for use in Study 2 did not use exactly the same format. Parents in Study 2 were given a forced choice which did not include options for reporting overlapping types of eye contact quality, as measured in Study 1. This means we cannot make exact comparisons between the measures. Nevertheless, despite differences in the presentation format, the measurement of common behavior indicators of quality of eye contact (staring, unfocused gaze, brief glances) in each of the two studies contributes new evidence to this sparse literature on the quality of eye contact within WS and across other ND groups. Further testing and replication is still a priority however. Although we might be encouraged by the endorsement rates for Study 2 across the options linked to Study 1, with few choosing the option "none of these apply," still further validation of the Study 2 method is needed. For example, we recommend further testing of internal, convergent, and discriminant validity as has been carried out for other questionnaires using DISCO items (e.g., C. R. G. Jones et al., 2020).

The most serious limitation of the study was that the lack of associations with ID, age, and gender, were likely due to a lack of power due to small samples distributed across the ND groups. Although the sample size for the WS group in both studies was the same as the sample size for other studies (Klein-Tasman et al., 2007, 2009), there were limitations in making group-wise comparison for each ND condition and in drawing conclusions on the effects of ID, age, gender and language level. As this was compounded by the constraint on caregivers to select only one of six options to describe their child's eye contact, further replication is needed by comparing larger participant groups and testing different research designs.

4.8.2. Future Directions and Implications

The relationship between older age and staring behavior in Study 1 is an intriguing finding. One explanation is that staring behavior emerges throughout development in WS. Another interpretation is that the reporting of marked staring in adults relates more to a change in the perception of this behavior. From the perspective of the interlocutor, an adult showing staring behavior may be more striking and deemed less socially acceptable compared to a child staring. However, it is important to note this association with age was not found in the child-only sample

of Study 2; therefore, future research should help to corroborate differences and similarities across age and ND groups.

Future cross-syndrome comparisons will also benefit from a fine-grained analysis of the differential qualitative aspects of unusual eye contact in relation to social interaction and communication. Klein-Tasman et al. (2007, 2009) noted findings of "abnormal eye contact" in young children with WS as measured within the ADOS domain of reciprocal social interaction. Common difficulties were also found in the ADOS domains of declarative pointing, showing and giving objects, reciprocal social interactions and social communication, and cognition. However, as the qualitative nature of eye contact (e.g., specific type of qualitative features) is not recorded by the ADOS, follow-up research using the ADOS, DISCO, or other assessment measures could help to clarify the relation between particular qualitative types of eye contact and other social interaction, communication, and social cognition difficulties. The prediction would be that unusual qualitative features have particular implications for other aspects of social interaction and for social cognition as the flow of interaction is affected.

Our findings may also prove useful in future transdiagnostic research, with respect to (a) separating out the cognitive processes involved in attention and arousal, (b) elucidating the neural circuitry associated with eye contact, and (c) the psychosocial factors associated with qualities of eye contact. In terms of the cognitive processes, it may be possible to test whether unfocused gaze is related to slow allocation of automatic attention (Kuhn et al., 2010), whether staring is related to attentional shifting and hypo arousal (Riby et al., 2011), and whether brief glances are linked to gaze aversion strategies during information processing (Doherty-Sneddon et al., 2012). In the case of neural processes, a more transdiagnostic analysis would be particularly informative for revealing the neural processes associated with qualities of eye contact in people with genetic and nongenetic ND conditions. Not only is there a dearth of research documenting how the brain circuitry responds to eye contact in people with ND conditions, to our knowledge, no research has examined how qualitative features of eye contact are subserved by neural substrates. Indeed, the characteristic use of qualitative features of eye contact early in life may itself have a role in neural development, indicating bidirectional biology-behaviour relations, rather than a simple underpinning of neural processes driving eye contact quality. The results also address psychosocial influences on eye contact and how

different qualitative features may serve as adaptive functions to increase or avoid social contact when eye contact is experienced as overly stimulating, distracting in some way, or not as socially rewarding. With respect to brief glances for example, for some people who find it aversive to look in the eyes of others (hyperarousal), brief glances may serve to reduce the uncomfortable sensation, as indicated by evidence of increased activation of the subcortical system when focusing on the eye region (Hadjikhani et al., 2017) and first-hand insights from autistic people (McGlensey, 2016; Trevisan et al., 2017). However, brief glances may also indicate an opportunity for information processing during gaze aversion (Doherty-Sneddon et al., 2012). Collecting further parental data on the quality of eye contact used by their child in varying contexts (e.g., interaction partners, social situations) would add valuable insights into the psychosocial factors that may influence eye contact behaviour.

The findings also point to the direction for future research priorities in the areas of FXS and ADHD. Our findings regarding brief glances support previous research with children with FXS. However, the previous research has largely referred to brief glances made while the individual looks elsewhere rather than as part of making eye contact. Therefore, further fine-grained observational research is needed to examine the extent to which the well documented finding of brief glances in FXS (e.g., Hall et al., 2015) provides a communication strategy for eye contact, at least as far as parents are concerned. At the same time, the results open a new direction of research in ADHD, a ND condition in which eye contact profiles have previously been neglected. The fact that only 42% of this group showed eye contact that is always appropriate and natural, and similarities in the pattern of unusual eye contact quality to that seen in other ND conditions, should be investigated in relation to their known challenges establishing and maintaining friendships (Normand et al., 2011, 2013) and broader sociocognitive skills (Bora & Pantelis, 2016a; Sibley et al., 2010; Uekermann et al., 2010). Further research is also needed with this group to understand eye contact patterns in those with co-occurring ADHD and autism.

From a clinical and societal perspective, the findings emphasise that eye contact given by people with ND conditions may look different from the NT preference of direct, steady gaze, but that the observable qualities may vary across individuals with the same diagnosis. Difference from a NT pattern of eye contact should not be interpreted as a call for intervention, given these

behaviours likely serve an adaptive role. One important consideration, however, is the potential impact that different eye contact behaviours may have on the wider social interaction, in terms of impression formation and potential stigma (Morrison et al., 2020; Sasson et al., 2017). Unusual qualities of eye contact may miscommunicate information about the intentions and attitudes of people with ND conditions. For example, brief glances may infer that the person is disinterested in the interaction. Equally, being on the receiving side of prolonged eye contact may be an uncomfortable experience. Prolonged staring at a time of greater social independence during adolescence and young adulthood is particularly important given the vulnerability issues that have been emphasised in people with ND conditions (Fisher, Moskowitz, et al., 2013; Jawaid et al., 2012; Ridley et al., 2020b).

To conclude, it is known that measurement differences lead to particular interpretations of eye contact (Jongerius et al., 2020). We argue that the previous single dimension interpretation, based on measurement of the degree or strength of eye contact, has led to the oversimplified assumption that reduced eye contact equates to poor eye contact, whereas eye contact that is not reduced equates to good eye contact. This in turn has led to an interpretation that polarises different ND groups such as WS and autism and makes the incorrect assumption about underlying social motivational and cognitive factors. Given our findings on similarities across ND conditions, we think it is time to focus on describing eye contact profiles more in terms of different qualitative styles, and less in terms of a single dimension (i.e., degree of presence/absence). This new perspective would have implications for research on psychological and neural mechanisms related to eye contact as it indicates that quality of eye contact subtypes may be studied independently of traditional diagnostic groupings and divisions.

Paper 2: A cross-syndrome approach to the social phenotype of neurodevelopmental disorders: Focusing on social vulnerability and social interaction style

4.9.Highlights

- Heightened social vulnerability is evident across multiple neurodevelopmental disorders.
- The limitation of IQ to explain social vulnerability is shown by a cross-syndrome approach.
- Atypical social interaction styles vary within and across neurodevelopmental groups.
- Social interaction styles make a unique contribution to heightened social vulnerability.
- Social phenotypes are best understood as distributed across diagnostic boundaries.

4.10. Abstract

Background: Following Annette Karmiloff-Smith's approach to cognitive research, this study applied a cross-syndrome approach to the social phenotype, focusing on social vulnerability (SV) and the factors that contribute to it. **Aims:** To (i) identify syndrome-specific differences in SV across four neurodevelopmental disorder (NDD) groups, (ii) determine the contribution of intellectual disability (ID), age or gender to SV, and (iii) explore its relationship with social interaction style (SIS). **Methods and Procedures**: 262 parents of children: Autism (n = 29), Williams syndrome (n = 29), Attention Deficit Hyperactivity Disorder (n = 36), Fragile X syndrome (n = 18), and Neurotypical (n = 150) reported on their child's SV, quality of SIS and other factors (ID, age, gender). **Outcomes and Results:** Heightened SV was not syndrome-specific. Instead it was found equally across NDD groups (and not in the neurotypical group), and independently of ID, age and gender. Different atypical SISs were also distributed across NDD groups and each were significantly related to SV, independent of the factors above and beyond neurodevelopmental diagnosis. **Conclusions and Implications:** The findings emphasise that social phenotypes are best understood as distributed across diagnostic boundaries and offer opportunities to further test the role of varied atypical SISs in the development of heightened SV.

Keywords: Social interaction style; social vulnerability; cross-syndrome comparisons; neurodevelopmental disorder; autism; Williams syndrome; Attention deficit hyperactivity disorder, Fragile X syndrome.

4.11. What this paper adds

In the first cross-syndrome comparison of social vulnerability (SV) profiles and social interaction styles (SISs), the current study emphasised that neurodevelopmental disorders (NDDs) of Autism, Williams syndrome, Attention Deficit Hyperactivity Disorder, and Fragile X syndrome are equally associated with atypical and heightened SV and that this cross-syndrome effect is not associated with intellectual disability (ID), age or gender. Furthermore, the study showed that SV is associated with the presence of distinctive, atypical patterns of SIS and that these are also found within and across these diagnostic groups. The results substantially extend previous evidence on cross-syndrome variability in both SV and SIS, highlighting the case for non-specificity in the social phenotype of different NDDs. The study also indicates the potential contribution of SIS as a factor in heightened SV beyond the effect of diagnostic group and other factors such as ID, age and gender. Crucially these initial findings strongly support a cross-syndrome approach to the study of SV in NDDs, and make a case for further consideration of the role of atypical SISs in our understanding of SV and the development of social phenotypes more generally.

4.12. Introduction

Annette Karmiloff-Smith pioneered a cross-syndrome approach to the study of cognition in neurodevelopmental disorders (NDDs; J. H. Brown et al., 2003; Paterson et al., 2006; Scerif et al., 2004). Her cross-syndrome approach has advanced the understanding of a wide range of phenomena, including language development (Kelly et al., 2013; Lindgren et al., 2009), face and emotion recognition (Annaz et al., 2009; Dimitriou et al., 2015; Martínez-Castilla et al., 2015), attention (Cornish et al., 2012; Scerif et al., 2004), sleep (A. Ashworth et al., 2013, 2017; D'Souza et al., 2020), psychopathology (Rodgers et al., 2012; Royston et al., 2019; Woodcock et al., 2009), sensory processing (Hannant et al., 2018; Heald et al., 2020) and social/adaptive behaviour (Hamner et al., 2019; Sumner et al., 2016; T. A. Williams et al., 2013).

The goal of a cross-syndrome design is often to identify differences in abilities between disorders while capturing variability. For example, Karmiloff-Smith encouraged researchers to study cross-syndrome associations in order to understand cognitive mechanisms that drive development in specific disorders. However, in addition to helping identify these mechanisms, the focus on cross-syndrome associations in itself illuminates invariance in some areas of functioning in comparison to specific differences (see also Asada & Itakura, 2012; Farran & Karmiloff-Smith, 2012). The aim of the current study was to apply a cross-syndrome approach to the social domain in order to explore specificity and/or invariance in particular aspects of the social phenotype. The main focus of the study is social vulnerability (SV), defined as "*the disadvantages faced by an individual while he or she endeavours to survive as a productive member of society*" (Jawaid et al., 2012, p. 335) or an "*impaired ability to detect or avoid potentially harmful interpersonal interactions*" (Pinsker et al., 2006, p. 109).

The cognitive and social mechanisms that drive the development of SV are not yet understood. Jawaid et al.(2012) has proposed that a combination of intellectual disability (ID) and atypical social behaviours result in heightened SV. However, this proposal has not been tested and research to date has been carried out only with one or two specific groups. First, with respect to the role of ID, evidence is sparse but preliminary studies suggest that this may not be a primary influence. For example, initial evidence with adults with Williams Syndrome who have heightened SV indicates that they do not differ across levels of ID (Lough & Fisher, 2016b).

Evidence also shows that those with Autism Spectrum Disorder (hereafter 'autism') who have heightened SV may not have IDs (Hofvander et al., 2009). Second, with respect to the role of atypical social interaction behaviour as a predictor of SV, to our knowledge there is no evidence available on this. Therefore, the current study explored and described for the first time the cross-syndrome variability of SV across five neurodevelopmental groups and the contribution made by ID and atypical social interaction style (SIS) as well as by other factors such as age and gender. Given the lack of previous evidence in this area, this exploratory method offers the potential to elicit factors relevant to understanding the development of heightened SV.

The current study focused on the relationship between SV and the variables above across a broad range of NDD groups (Autism, Williams Syndrome [WS], Attention Deficit Hyperactivity Disorder [ADHD], Fragile X Syndrome [FXS]) and neurotypical development

[TD]. The motivation for including the four NDDs as well as TD was three-fold. First, all four NDDs are characterised by unusual social interactions in the literature. Social difficulties are definitive for autism (American Psychiatric Association, 2013) and an unusual over-approaching SIS is associated with WS (Doyle et al., 2004b; Jarvinen et al., 2013; Järvinen-Pasley et al., 2010a; Riby, Hanley, et al., 2014). FXS is associated with high social motivation alongside significant social anxiety and social communication difficulties (Cordeiro et al., 2011; Kau et al., 2004; Kaufmann et al., 2004; Roberts et al., 2007). Many children and youth with an ADHD diagnosis also show socio-cognitive impairments in areas of social problem-solving and perspective taking (Bora & Pantelis, 2016a; Sibley et al., 2010) and experience interpersonal challenges, including an absence of mutual friends (Bagwell et al., 2001; Hoza et al., 2005), less stable and lower quality friendships (Normand et al., 2013) and high rates of peer rejection and victimisation (Holmberg & Hjern, 2008; Taylor et al., 2010). For an overview of peer difficulties in ADHD see Gardner & Gerdes (2015).

Secondly, we know there is significant within-disorder heterogeneity in all areas of cognition and behaviour in developmental disorders (Charman, 2015; Masi et al., 2017; Porter & Coltheart, 2005) as well as in TD. Research emphasises the overlapping characteristics between syndromes and a potential lack of discrete diagnostic boundaries at the behavioural level (Asada & Itakura, 2012; D. Bishop & Rutter, 2009; Dyck et al., 2011; Kaplan et al., 2006; Moreno-De-Luca et al., 2013; Zorlu et al., 2015). For example, studies often adopting a cross-syndrome approach have revealed the many shared social features between individuals with Autism and WS (Asada & Itakura, 2012; Hamner et al., 2019; Klein-Tasman et al., 2009; Vivanti et al., 2018). Consequently, the field has moved away from the notion of these two neurodevelopmental conditions as polar opposite of social functioning. Utilising a cross-syndrome approach to the study of SV and SIS in a much broader range of neurodevelopmental groups should help pinpoint where there are both group differences and shared features.

Finally, variability in social interaction abilities is also found in the TD population and therefore the inclusion of a TD group allows us to consider the behaviours that fall within the range of ‘typical’ variation including the extremes of individual differences. In a study design that examines both syndrome differences and cross-syndrome similarities, the issue of ‘typicality’, can only be considered by the inclusion of a TD group.

Both SV and SIS were operationalised using established methods. However, as these methods were adapted for the study, the measurement format of each construct was tested for the first time. SV was measured using a subset of items from the Social Vulnerability Questionnaire (Fisher et al., 2012), while SIS was measured using Wing and Gould's (1979) clinical classification system of SIS subtypes (see Scheeren et al., 2020 for recent description).

In summary, the present study aimed to address the following research questions: First, are there syndrome-specific differences between NDD groups in SV? Second, can SV be explained by other factors such as ID, age, or gender? Third, is there a cross-syndrome *association* between SV and SIS and if so, does SIS itself make a unique contribution to SV, independent of other factors above?

4.13. Method

4.13.1. Participants

276 parents or guardians were recruited for the study. Data from 14 participants were removed prior to analysis due to parents reporting that their child did not meet the inclusion criteria because they (i) presented with a variety of difficulties changeable over time or had a diagnosis beyond the focus of the study (n = 5), (ii) fell outside of the age range (n = 1), or (iii) were recruited to the TD group but parents reported an intellectual disability or presence on the special educational needs register (n = 8). The final sample included parents or guardians of 262 4- to 17-year-old children (M_{Age} = 112 months, SD = 42.43) living in the UK (93% Mothers), of which 118 were parents of children with a diagnosis of a NDD and 150 were parents of TD children. The children were categorised into 4 NDDs: Autism (n = 29), WS (n = 29), ADHD (n = 36), FXS (n = 18; see Table 4.4). Parents were recruited through a university research participation database for local families, social media, and via UK charity networks (e.g. Williams Syndrome Foundation, ADHD Foundation, and Fragile X Society). The study complied with ethics (as per BPS requirements) and GDPR legislation (as per University requirements) and received favourable ethical opinion from the local ethics committee. Parents opted-in to the study and were not reimbursed for their time.

Age was normally distributed for each of the neurodevelopmental groups but not for the TD group. Preliminary analysis using Kruskall-Wallis analysis across all 5 groups (M_{Age}: TD =

107 months, Autism = 127 months, WS = 100 months, ADHD = 126 months, FXS = 120 months) found a main effect of chronological age ($H(4) = 16.25$, $p = .003$) however this difference was confined to a difference between the TD and NDD subgroups (specifically Autism and ADHD groups). Follow up tests using the Bonferroni correction revealed no significant differences in age between the four NDD subgroups.

The NDD subgroups differed in parent-reported ID status as seen in Table 4.4, $\chi^2(df = 3) = 50.98$, $p < .001$. With respect to language, parents reported that the majority of participants in all groups had expressive language and receptive language at the level of full sentences, Table 4.4. However, the number of children with and without full sentences differed between the four NDD subgroups for receptive language $\chi^2(df = 3) = 10.41$, $p = .015$) and expressive language $\chi^2(df = 3) = 24.31$, $p < .001$).

4.13.2. Materials

Parents completed a bespoke online questionnaire about their child's social functioning and social interactions, via Online Survey software (www.onlinesurvey.ac.uk). Of the items included in the online questionnaire, only the quality of SIS and SV items are reported here[2]. A set of demographic questions were asked at the end (e.g. parents provided their child's date of birth, gender, Special Educational Needs status (SEN) and gave information about diagnosis, schooling and presence of ID, some of which are provided below in terms of describing the sample [also see Table 4.4]).

[2] This research formed part of a larger study exploring social interactions in children with and without developmental disabilities

Table 4.4

Demographic Characteristics of the Sample (% Reported) and SV-Total, Split by Diagnostic Group

	Autism (N = 29)	WS (N = 29)	ADHD (N = 36)	FXS (N = 18)	TD (N = 150)
Males/females/prefer not to say	72/28/0	59/41/0	78/19/3	94/6/0	48/51/1
Age					
Mean (SD) (months)	127 (28.4)	100 (36.3)	126 (35.5)	120 (43.7)	107 (45.8)
Range (months)	59-187	48-204	54-197	52-178	48-215
Presence of a physical disability	21	21	3	17	1
Presence of a hearing impairment	0	10	6	0	0
Presence of a visual impairment	10	37	14	11	6
Presence of an intellectual disability	21	90	28	89	0
Stage of education					
Preschool	3	10	3	11	7
Primary	62	77	53	45	63
Secondary	35	10	44	45	27
Post-16 education	0	0	0	0	3
Educational provision					
Mainstream school	66	50	86	22	97
Special Educational school	17	43	11	67	0
Home-schooling	10	3	3	0	3
Other[1]	7	3	0	11	0
Special educational needs register					
Yes	69	69	50	67	0
No	17	3	33	6	98
I don't know	14	24	17	28	2
Statement of SEN/EHCP					
Yes	52	90	33	89	-
No	45	7	61	6	-
I don't know	3	3	6	6	-

	Autism (N = 29)	WS (N = 29)	ADHD (N = 36)	FXS (N = 18)	TD (N = 150)
Use of language to communicate					
None	3	7	0	11	1
Single words	3	7	0	17	0
Simple phrases	7	24	6	33	0
Full sentences	86	62	94	39	99
Understanding of language					
None	0	0	0	6	0
Single words	0	7	0	0	0
Simple phrases	17	28	6	28	0
Full sentences	83	66	94	67	100
SV-Total					
Mean (SD)	17.83 (5.05)	17.07 (4.29)	14.83 (4.61)	16.78 (5.61)	6.08 (4.01)

[1]Four parents reported 'other'. This included children not currently in educational provision (due to pupil/school
Choice) and pupils with a mix of provision.

4.13.3. Measure of level of SV

SV was measured using nine items from the Social Vulnerability Questionnaire (SVQ). The SVQ is a 30-item parent-report measure of vulnerability, which taps *Emotional Bullying, Risk Awareness, Social Protection, Perceived Vulnerability, Parental Independence* and *Credulity* (Fisher et al., 2012). The SVQ was validated on 144 parents of individuals with intellectual and developmental disabilities and has previously been used to examine SV in autism, WS and Down syndrome (Fisher et al., 2012; Fisher, Moskowitz, et al., 2013; Lough & Fisher, 2016b). As the SVQ focuses on many broader issues of vulnerability than the current study aimed to explore (e.g. physical threat) we selected nine items specifically social in nature (SVQ items 1, 3, 4, 12, 13, 14, 16, 19, 25). Parents rated statements on a Likert scale of 0 ("not true/never") to 3 ("very true/always"). Potential SV total scores (SV-Total) ranged from 0-27, with a higher SV-Total score indicative of greater SV. Cronbach's alpha for the nine SV items on the current total sample was 0.87 (per NDD subgroup: TD: α = 0.69, Autism: α = 0.70, WS: α = 0.64, ADHD: α = 0.7, FXS: α = 0.81).

4.13.4. Measure of SIS

SIS was measured using Wing & Gould's (1979) original clinical classification system of SIS subtypes ('typical', 'aloof', 'passive' and 'active-but-odd (hereafter 'active-but-unusual'). This classification system shows good internal consistency when used in a parent questionnaire format (Castelloe & Dawson, 1993; Roeyers, 1997; Scheeren et al., 2012). The classification also shows good external validity (Borden & Ollendick, 1994; Waterhouse et al., 1996). Extended versions of the classification system have been developed and these have been validated in behavioural observation studies using a single checklist judgement by naïve observers (Roeyers, 1997) and in parent interview studies with judgements by interviewers blind to diagnosis (S. R. Leekam et al., 2002; Wing et al., 2002). The current study followed the extended interview classification developed by Wing (2006) described in Kent (2014) and used within the validation study of Leekam et al., (2002). For the current study it was adapted into a parent questionnaire context (see Table 4.5). Parents/guardians were asked to select one description that best described their child's social interactions. From the original classification, five subtypes were selected, four of which were, 'typical', 'aloof', 'passive' and 'active-but-unusual. The fifth, "shy but social contact is appropriate for mental age with well-known people, including age peers"

was also selected because together with the ‘typical’ description it formed part of the original measure of ‘appropriate’ social styles previously validated (S. R. Leekam et al., 2002). Several subtype descriptions were combined to form the ‘aloof’ subtype. Subtypes that specifically referred to (a) WS and (b) FXS, and also the subtypes; “selective mutism” and “over- formal, stilted, rigid, over-polite and calmly outspoken” were excluded.

Table 4.5

Percentage of Children Within Each Quality of Social Interaction Style Subtype Across Diagnostic Groups (%)

Quality of Social Interaction (%)	Interaction label	Group					
		Autism (n=29)	WS (n=28)	ADHD (n=36)	FXS (n=17)	TD (n=150)	NDD[3] (n=112)
1. Social contacts with children and adults are appropriate for level of ability. Looks up with interest and smiles when approached. Responds to the ideas and interests of people of similar mental/developmental age and contributes to the interaction	Appropriate	6.9	31	38.9	0	74.0	22.3
2. Shy but social contact is appropriate for mental age with well-known people, including age peers. Might refuse to talk to adults but interacts with other children	Shy	17.2	0	8.3	23.5	17.3	10.7
3. Makes social approaches actively but these are usually inappropriate / the behaviour is not modified according to the needs, interests and responses of the person approached	Active-but-unusual	48.8	51.7	36.1	17.6	4.0	40.2
4. Generally does not initiate but responds to social contact if others make approaches. May join in passively and shows pleasure in passive role and may try to copy but with little understanding	Passive	24.1	13.8	13.9	41.2	4.0	20.5
5. Does not interact; aloof and indifferent (though may interact to obtain physical needs, including physical contact needs, rough and tumble play, cuddle)	Aloof	3.4	0	2.8	17.6	0.7	4.5
Unusual / inappropriate for mental age[1]		75.9	65.5	52.8	72.2	8.7	65.2
Typical / appropriate for mental age[2]		24.1	31.0	47.2	22.2	91.3	33.0

[1] Descriptions 3, 4 and 5 collapsed to form one category
[2] Descriptions 1 and 2 collapsed to form one category
[3] NDD = Autism, Williams syndrome, ADHD and FXS collapsed to form one category

4.13.5. Analytic Approach

Shapiro-Wilk test of normality indicated that the data for SV-Total was not normally distributed for the sample as whole, or for the TD and Autism groups (when normality tests were run per diagnostic group). Therefore, nonparametric tests were used but results from parametric tests were reported if they did not differ. To examine RQ1, group differences in SV were tested using Kruskal-Wallis H test / one-way ANOVA. To examine effects of ID status, age and gender (RQ2) Spearman's correlations and Mann Whitney tests/ t tests were used. Finally, to examine associations between SIS and SV and whether SIS was uniquely related to level of SV (SV-Total score), a multiple regression analysis was conducted with SIS subtypes as the predictor variables, and SV-Total as the dependent variable (RQ3), while statistically controlling for age, gender, ND status and ID status. For all tests, an alpha value of .05 was set, unless multiple comparisons required Bonferroni adjustment. In addition, analyses were re-run to equalise the size of the TD comparison group.

4.14. Results

4.14.1. SV and NDD

Mean SV-Total scores for each diagnostic group are shown in Table 4.4. To examine differences across groups (TD, Autism, WS, ADHD and FXS), SV-Total scores were analysed using a one-way ANOVA. A significant group difference was found, $F(4,257) = 90.81$, $p < 0.001$, $\eta_p^2 = 0.59$. Scheffe post hoc comparisons revealed that SV-Total score for the TD group was significantly lower than all four NDD subgroups (all p's < .001; see Table 4.4). Pairwise comparisons showed no significant difference in SV-Total score between each of the NDD subgroups (all p's > .05). Atypically heightened SV was a feature of NDD diagnosis and distinctive from TD, but it was not syndrome-specific; instead scores were equivalently elevated across all the four NDD subgroups. Given the unequal size of the TD group, the ANOVA was re-run using the first 35 respondents who were recruited into the TD group. Results showed a significant group difference in SV-Total score $F(4,142) = 38.38$, $p < .001$, $\eta_p^2 = 0.52$. As above, mean SV-Total score for the TD group was significantly lower than all four ND groups, with no significant difference between the ND groups. The result from the full sample analysis was therefore maintained.

4.14.2. SV and age, ID and gender

SV-Total score was not related to Age for the sample as a whole (TD, Autism, ADHD, WS, FXS combined; $r_s(259) = .08$, $p = .21$), or for the NDD group taken together ($r_s(259) = .10$, $p = .29$). Follow-up comparisons for each sub-group also showed no significant SV correlation with Age for the Autism, WS, or ADHD groups (all p's > .05) and although there was a significant, positive relationship between SV-Total score and Age for the FXS group ($r_s(17) = .54$, $p = .03$) and a significant negative relationship for the TD group ($r_s(149) = -.19$, $p = .02$, the significance level did not survive when Bonferroni adjustment for multiple comparison was applied (.05/6 = p.01).

For the analysis of ID, only the NDD subgroups were included as none of the TD group had ID. For the NDD group as a whole, there was no significant difference in SV-Total score associated with the presence of ID ($M = 16.67$, $SD = 4.66$) / absence of ID ($M = 15.96$, $SD = 5.24$), $t(104) = 0.74$, $p = .46$, $d = 0.14$. For Gender, taking the sample as a whole, there was a significant difference in SV-Total score due to Gender, with higher SV reported for males ($M = 11.70$, $SD = 7.12$) than females ($M = 8.62$, $SD = 5.70$); $t(248.93) = 3.87$, $p < .001$, $d = 0.48$. This difference was not significant when all NDD subgroups were analysed together ($t(64.44) = 1.03$, $p = .31$, $d = 0.21$), but was significant for the Autism group independently, as males ($M = 18.52$, $SD = 5.13$) scored higher than females ($M = 14.38$, $SD = 3.54$; $t(18.53) = 2.47$, $p = .02$, $d = 0.94$), although this effect did not survive Bonferroni adjustment (.05/4 = p.01). Note that there was a substantial imbalance in gender in the autism group and other NDD groups (see Table 4.4 for a breakdown of gender per group). There was no significant gender difference for each of the remaining four developmental groups (including TD) (all p's > .05). The aforementioned results remained unchanged when the analysis was applied with the reduced TD sample.

4.14.3. SV and SIS

To examine the relation between SV and different types of SIS, several analyses were conducted. First, the relation between SV-Total score and SIS was explored for the whole sample independently of NDD subgroup status. An initial ANOVA, with SIS subtype as the independent variable (5 categories as shown in Table 4.5) and SV-Total score as the dependent variable showed a significant effect of SIS subtype on SV-Total score, $F(5, 256) = 36.02$, $p < .001$, $\eta_p^2 = 0.41$. Post hoc comparisons also revealed that children with SIS subtypes 'appropriate (1)' ($M = 7.03$, $SD = 5.06$) and 'shy (2)' ($M = 9.03$, $SD = 6.00$) had

significantly lower SV-Total scores than children with the atypical SIS subtypes 'active-but-unusual (3)' (*M* = 16.69, *SD* = 4.60), 'passive (4)' (*M* = 16.34, *SD* = 5.94) and 'aloof (5)' (*M* = 16.67, *SD* = 4.80). However, SV-Total scores did not differ between 'appropriate' and 'shy' SIS subtypes ($p > .05$). Neither were differences found between each of the three atypical SIS subtypes (all *p*'s > .05). This result remained even when subtype 5 'aloof' (n = 6) was collapsed with subtype 4 'passive' (n = 29) into a social withdrawal subtype (*M* = 16.40, *SD* = 5.70), and compared with subtype 3, 'active-but-unusual' (n = 51; *M* = 16.69, *SD* = 4.60) due to unequal samples (*t* test; $p = .8$, $d = 0.06$). The results reported above were maintained when analysed using the reduced TD sample.

Next, to examine whether each of the atypical SIS subtypes uniquely predicted SV independent of other factors, including diagnostic status, a regression analysis was conducted. The SV data were entered for the whole sample, including TD data (original sample) in order to increase variability. An initial model was run with only SIS subtypes as predictor and SV-Total as the dependent variable. The model generated (adjusted $R^2 = .40$) was a significant predictor of overall SV-Total score, $F(4,257) = 44.58$, $p < .001$, with each of the four interaction subtypes entered (excluding "appropriate") making a significant contribution to the model (all *p*'s < .05). In order to probe the unique contribution of SIS, a sequential, multiple-regression strategy was conducted where Age, Gender, ID status (presence/absence of ID) and NDD status (the presence of a NDD compared to TD) were entered in Model 1; Model 2 added SIS subtypes. Of the variables entered in Model 1, only NDD status was a significant predictor of SV-Total ($p < .001$). Age ($p = .39$), Gender ($p = .68$) and ID status ($p = .43$) did not significantly contribute therefore, the regression was rerun with only NDD status statistically controlled for. The regression revealed that at Model 1, NDD status entered alone contributed significantly to the model ($p < .001$) and accounted for 57.3% of the variation in SV-Total. Introducing the SIS subtypes explained an additional 5.3% of variation in SV-Total and this change in R^2 was significant ($p < .001$). With ND status statistically controlled for, the SIS subtypes of *active-but-unusual*, *passive* and *aloof* significantly contributed to the model (all *p*'s <.001). The SIS subtype *shy* was no longer a significant predictor of SV-Total ($p = .2$).

4.15. Discussion

Inspired by Annette Karmiloff-Smith's approach to cognitive research, this study applied a cross-syndrome approach to the social phenotype; focusing on SV and its relation to SIS, ID and other factors.

4.15.1. Are there syndrome specific differences in SV?

One of the main findings was that heightened SV was found across multiple NDDs. Parents/guardians in the Autism, WS, ADHD and FXS groups all endorsed higher levels of SV compared to parents of neurotypical children. While autism and WS are already known to be two particularly socially vulnerable populations (Fisher et al., 2012; Fisher, Moskowitz, et al., 2013; Griffiths, Allison, Kenny, Holt, Smith, & Baron-Cohen, 2019; Jawaid et al., 2012; Lough, Flynn, et al., 2015; Lough & Fisher, 2016b; Riby et al., 2017; Sofronoff et al., 2011) this is the first evidence of heightened SV in ADHD and FXS groups. This finding suggests that heightened SV may be a clinical phenomenon that is a shared feature of NDDs, even those distinguished by specific genetic and biological aetiologies.

To date we know little about the developmental mechanisms of SV and further research beyond this study will be needed in order to isolate and test out these mechanisms. Our starting point was to address Jawaid et al's. (2012) proposal that a combination of ID and atypical social behaviours contribute to heightened SV and we used parent questionnaires to explore the concurrent contribution of these and other factors (age, gender) to SV.

4.15.2. Can SV be explained by ID, age, or gender?

First, we found, like previous studies, that ID did not fully explain SV (Hofvander et al., 2009; Lough & Fisher, 2016b; Wilson et al., 1996). Comparable, heightened SV was also found in the NDD groups not characterised by ID (ADHD and Autism, of whom 72% and 79% did not have an ID as reported by parents, respectively).

Age also did not explain the presence of heightened SV scores either for the whole sample or for the NDD groups, except for small correlations with FXS and TD which did not survive adjustment for multiple testing. Although these findings for IQ and age might be surprising, the participants were young. Therefore, further research should investigate whether the lack of age and ID would replicate in older groups of individuals. A major limitation is also that our measure of ID was limited by parent-report ID (yes/no response) and formal standardised measurement of ID would be needed in order to probe this more

accurately. Finally, gender also did not significantly contribute to SV although there were indicative findings of higher scores in males in the autistic group only. However, inequality in gender grouping size constrained the analysis. Further research is needed with matched gender samples to clarify these effects.

The results for ID, age and gender support other studies using the SVQ (e.g., Lough & Fisher, 2016b) and extend findings of heightened SV for the first time to ADHD and FXS, as well as consolidating the finding of reduced SV within a large sample of neurotypical children. As the study used only a small subset of items from the longer SVQ this may indicate the effectiveness of this format for this purpose. However, while internal consistency was good, the lack of psychometric testing on this abbreviated version was a limitation, potentially restricting its capacity to capture differences and further research comparing the question sets is needed.

Nevertheless, these findings emphasise that SV may potentially be an issue that transcends diagnostic boundaries, irrespective of ID, and this reinforces the view that "*individuals with intellectual and developmental disabilities can be vulnerable in multiple, potentially unrelated ways, and it is important for researchers and clinicians to try to capture these distinct patterns of vulnerability*" (Fisher et al., 2018, p. 8).

4.15.3. Understanding SV through the lens of SIS

To explore the role of SIS, we used the classification of quality of social interaction based on Wing and Gould's original typology (1979), drawing on Wing's (2002, 2006) clinical classification and applying this within a parent questionnaire. Five SIS subtypes were analysed; three atypical (aloof, passive and active-but-unusual) together with two appropriate styles (typical and shy). Although Scheeren et al., (2012) found an active-but-unusual style in children with autism who were also reported as having ADHD features and/or disruptive or social-emotional behaviours, this is the first study to apply Wing & Gould's classification system to those with diagnoses beyond autism and pervasive developmental disorder. The results showed that atypical SIS was strongly associated with SV and this association was not specific to any particular type of atypical SIS subtype (aloof, passive, active). Furthermore, like the results for SV we also found no syndrome-specific effects; atypical SIS was found across all the NDD groups.

To further explore the possibility of a unique contribution made by SIS, the regression analysis revealed that each of the atypical SISs (aloof, passive and active) made a significant

contribution to SV even when neurodevelopmental status was accounted for. In contrast the shy style did not significantly contribute. However, the magnitude of contribution made by SIS to SV was smaller than that of NDD status. Furthermore, of the child characteristics examined (ID, age, gender), only the presence /absence of a neurodevelopmental diagnosis was a significant predictor in the model, indicating a qualitative difference between typical and atypical development (Autism, WS, ADHD or FXS) regardless of a range of other factors.

In summary, these findings show that atypical SIS uniquely contributes to SV. Importantly not one, but each of these SISs separately (aloof, passive, active-but-unusual) were significant predictors of SV. Each of these SISs are very different from each other, yet each may still be relevant to the characterisation of any NDDs. Given that between half to three quarters of the children in all NDD groups studied here were classified with one of these particular social interaction profiles (see Table 4.5), it is possible that these profiles are not part of the normal distribution across the population and that difficulties with social interaction may serve as a consistent flag or indicator when clinical neurodevelopmental diagnosis is considered.

4.15.4. Considerations and future research

The current study has a number of limitations that require consideration. In terms of measurement, the two main measures had been adapted from pre-existing measures and used for the first time in this research. While the adapted measure of SV showed good internal validity, an assessment of external validity is needed. Similarly, although the SIS checklist judgement method used for the first time with parents appeared to be effective, it requires validation against clinical judgement and in comparison with the well-established Wing Subtypes Questionnaire (e.g. Castelloe & Dawson, 1993). A considerable measurement concern is also that both measures collected parent information, raising the possibility of informant bias and follow up studies using cross-informant analysis and other forms of testing are needed. In terms of design, correlational studies of this kind are insufficient to provide insight into the directionality of the relationships between variables. An experimental design would help to disentangle the concepts of SV, for example, by separating particular types of individual style (e.g. aloof, passive, active-but-unusual) from particular types of behaviours by others (e.g. taunting, exploiting a child (e.g. for favours) or rejecting a child's social approach). An experimental design would also be necessary in order to test the effect

of interventions, both to support children and reduce stigma and victimisation by other people.

It has been argued that the SIS of children with autism may be a predictor of intervention success, with several studies giving insights on how to tailor interventions to support different children with different SIS (Begeer et al., 2015; Beglinger & Smith, 2005). Also, studies with autistic children show that an active-but-unusual style is seen more commonly in older children with higher IQ and that these children have less severe autism symptoms especially across time (Scheeren et al., 2020), while those classified as 'aloof' make fewer improvements after intensive intervention (Beglinger & Smith, 2005). Our findings across NDD groups suggest the need for further exploration of this evidence, given that half of the WS group also show the active-but-unusual SIS accompanied by ID. For this group, an approaching social style may lead to greater social learning opportunities while their ID may limit their ability to take up the opportunity to learn. In the case of higher IQ in children with autism and ADHD, their higher IQ may be a protective factor to enable them to learn and adapt to complex social challenges and in turn possibly change their social interactions (Scheeren et al., 2020).Yet even for this group, at some point the demands of complex social environments may exceed adaptive capacity.

In the current study, presence of ID was controlled in the analysis as we examined the contribution of SIS on SV; however future studies should further examine the role played by cognitive ability, both general IQ and specific cognitive skills (e.g. executive functioning, theory of mind) in a cross-syndrome approach extending existing work on SIS and autism intervention (Begeer et al., 2015; Beglinger & Smith, 2005). Like these studies, future work should be directed towards developing interventions that are sensitive to SIS. However new work should also focus on SV to provide a cross-syndrome understanding of how SIS and adaptive cognitive skills can help the individual to buffer particular kinds of challenges that they face in the social environment. Such focus on SV ensures that interventions can also work to assess and intervene on disadvantages experienced by the individual that can be identified in their social environment, including the contribution of other individuals, the social group and organisations.

In summary, the current study is the first to compare SV profiles and the role of SIS, in TD children and children with a range of NDDs. While exploratory in nature, this study provides preliminary evidence that SISs may play an important role in SV and opens up

potential avenues for future research to delineate the nature of the association more comprehensively. We know that "equivalent behavioural outcomes stem from different underlying processes" (Karmiloff-Smith, 1997, p. 513), therefore studies adopting a cross-syndrome approach are key in understanding whether pathways to SV are the same or different across neurodevelopmental groups.

4.16. End of Chapter Discussion

This Chapter has advanced the understanding of the WS social phenotype by focusing on the *quality* of salient social behaviours and studying across multiple neurodevelopmental groups. One core finding emphasised from taking a cross-syndrome design is that social phenotypes can be conceptualised as shared across groups. The qualities of eye contact behaviour and social interaction styles were distributed within CYP with WS and across the neurodevelopmental groups studied. In the example of social interaction style, CYP in the sample did not neatly fit into a category of social interaction style based upon their diagnostic label. Similarly, while staring/prolonged eye contact was more likely to be evident in WS compared to other qualitative styles, parents in the WS group also endorsed other styles of eye contact and staring was not exclusive to WS. This challenges the traditional characterisation of the WS social profile and highlights the variability within WS. In addition, we found tentative evidence that elevated social vulnerability is evident in WS but also autism, ADHD and FXS. These findings support an increasing body of evidence that there is huge variability in people who share the same neurodevelopmental diagnosis, but also overlap between people with different diagnoses. In view of this, the idea that we can or should seek to identify behavioural outcomes based on a diagnostic label is oversimplistic. Furthermore, while we found evidence that an over approaching style was related to higher social vulnerability, this was equally the case for other styles of interaction. Therefore, the factors that increase social vulnerability are therefore likely to be shared across neurodevelopmental groups and this raises the question of social vulnerability in groups beyond WS. It also raises questions about the design of studies that include multiple neurodevelopmental groups. Indeed, since designing the study, researchers have advocated for more transdiagnostic approaches to answering questions about neurodevelopment (Astle et al., 2021) and this is relevant to future studies seeking to understand the factors that underlie heightened social vulnerability in CYP with various neurodevelopmental profiles. I will return to this in the General Discussion (Chapter 7) when proposing implications.

Nevertheless, the overarching aim of the PhD research was to advance understanding of social vulnerability in WS, therefore the remaining Chapters of the thesis return to focus on CYP with WS. The next Chapter details the profile and nature of social vulnerability during childhood and adolescence in WS. Chapter 6 then builds upon the evidence that social interaction styles contribute to social vulnerability, by examining the relationship with other aspects of behaviour and cognition.

Addendum to Chapter 4

Addition to Paper 1 Study 2 (section 4.8.1) to provide further evaluation of the eye contact measure used:

A strength of the eye contact measurement is that it used items from a standardised instrument, the DISCO. Study 1 used the semi-interview format (as intended for the DISCO) and allowed parents to endorse multiple qualities of eye contact. In contrast, Study 2 adapted the measure to be suitable for survey design and adopted a fixed-choice format whereby parents had to make a forced choice between the different styles (select only one of six options). In reality, it is likely that qualities of eye contact co-occur or overlap depending on context (i.e. familiarity of the interlocutor and situational factors). Indeed, parents stated this as the reason for selecting "none apply" to the qualitative features of eye contact given. Context matters when it comes to eye contact. For example, evidence from the FXS literature suggests that individuals show a 'warm-up' effect whereby gaze aversion is high in novel settings but diminishes as the person in the interaction becomes more familiar (Roberts et al., 2019; Hall et al., 2009). The measurement approach taken in this study—to assess quality of eye contact based on one or two questions—is inadequate for understanding the subtleties of eye contact behaviour across many types of real-life social interaction contexts. Asking parents to endorse features of eye contact across various contexts (i.e. interactions with close peers versus strangers or adults) would provide a more nuanced account. The Eye Contact Avoidance Scale (ECAS; Hall & Venema, 2017) is a potentially useful measure to extend the current findings as it probes eye contact across five domains of social functioning (e.g. when listening/speaking to others) and three levels of familiarity (the informant, friends and family, unfamiliar people). However, the ECAS is designed to measure eye contact avoidance specifically, whereas the findings from the current study emphasised that qualities beyond avoidance are commonly seen within and across neurodevelopmental groups. Another approach to better understand the interaction between eye contact and contextual factors would be to collect self-reported information from adults with WS about the experience of using and receiving eye contact (see Trevisan et al., 2017 for an example of a qualitative approach with autistic people).

Addition to Paper 1 Study 2 (section 4.8.2) to provide discussion about sex-based phenotypic variation in FXS and the potential impact on the results:

Another consideration is whether the sex-based phenotypic variation seen in FXS impacts on the pattern of results. FXS is an X-linked syndrome. Males have only one X chromosome, therefore if they inherit the X chromosome with the FXS mutation then all their X chromosomes are impacted. Females on the other hand have two X chromosomes and, in most cases, only one is affected by the FX mutation with the second X chromosome providing some level of compensation (see Bartholomay et al., 2019 for an overview). The clinical presentation in females is variable, but generally females present with milder symptoms than males, particularly in areas of cognition. Males with FXS tend to have ID in the moderate to severe range along with attention deficit characteristics. The female profile is more often typified by socio-emotional and mental health challenges. While evidence on eye contact/gaze behaviour between males and females with FXS is lacking, a recent study by Miller et al. (2023) reported patterns of eye contact in young females with FXS (mean age 11 years) similar to what has been reported in research with males with FXS. The females with FXS in the study had lower levels of eye contact (measured using eye tracking) compared to a comparison group of females without FXS matched on verbal IQ. They also showed greater physiological arousal (increased pupil dilation) when looking into the eyes of the researcher (as part of prompting), indicating that gaze aversion in individuals with FXS may signal hyperarousal. Given the known phenotypic variation between sexes, it is noteworthy that only one participant in the FXS group was female (the underrepresentation of females is a wider issue of FXS research; Bartholomay et al., 2019). Therefore, the pattern of eye contact described in this study may vary should the study be replicated with a sample of young females with FXS.

Addition to Paper 2 (section 4.14 Results) to test the direct comparison of SIS profiles between groups:

SIS and diagnostic groups: A 4 x 5 chi-square analysis was carried out to test the association between primary diagnosis (autism, WS, FXS, ADHD) and social interaction style. A statistically significant association was found, $\chi\,(12) = 35.98$, $p < .001$. However, the chi-square analysis indicated that 50% of the cells had expected frequencies below 5 with the lowest cell count being .77. This violates an assumption of the test that no more than 20% of cells should have expected counts below 5 (Moore & McCabe, 1999). The analysis was

repeated with the social interaction style ‘aloof’ removed due to low endorsement and the assumption was still violated (37.5% of cells with expected counts below 5). It is therefore not appropriate to draw conclusions about the association between diagnostic group and social interaction style from this analysis.

Addition to Paper 2 (section 4.14 Results) to provide further detail on the outcome of the regression analyses:

Table A

Regression coefficients of social interaction style on social vulnerability

Variable	*B*	β	SE	Adjusted R^2	Δ R^2
Model summary				.40	.41***
Constant	7.03***		.45		
Shy	1.20*	.11	.96		
Active	9.66***	.57	.86		
Passive	9.32***	.43	1.07		
Aloof	9.64***	.21	2.18		

Note. An initial model of the impact of social interaction style on social vulnerability score. The social interaction style ‘appropriate’ is not entered as a predictor as this is the reference group.
*$p < .05$, *** $p < .001$

Table B

Regression coefficients of social interaction style on social vulnerability after controlling for demographic variables (initial model)

Variable	*B*	β	SE	Adjusted R^2	ΔR^2
Step 1				.58	.58***
Constant	6.66***		.82		
Age	-.01	-.05	.01		
Gender[a]	.22	.02	.60		
ID status[b]	.82	.05	.87		
ND status[c]	10.03***	.73	.77		
Step 2				.63	.06***
Constant	6.05***		.81		
Age	-.01	-.04	.01		
Gender[a]	.01	.00	.57		
ID status[b]	.69	.04	.84		
ND status[c]	7.72***	.56	.83		
Shy	.89	.05	.80		
Active	4.18***	.25	.85		
Passive	4.85***	.22	.99		
Aloof	4.04*	.09	1.84		

Note. In step 1, the control variables of age, gender, ID status and ND status were entered to predict social vulnerability score. In step 2, the social interaction styles were entered as predictors. The social interaction style 'appropriate' is not entered as a predictor as this is the reference group.
[a] Female = 0, Male = 1. [b] no ID = 0, presence of ID = 1. [c] neurotypical = 0, neurodevelopmental (ND) diagnosis = 1.
* $p < .05$, *** $p < .001$

Table C

Regression coefficients of social interaction style on social vulnerability after controlling for ND status (final model)

Variable	*B*	β	SE	Adjusted R^2	Δ R^2
Step 1				.58	.58***
Constant	6.08***		.36		
ND status[a]	10.39***	.76	.55		
Step 2				.62	.05***
Constant	5.55***		.38		
ND status[a]	8.05***	.59	.66		
Shy	.94	.05	.77		
Active	4.04***	.24	.83		
Passive	4.41***	.21	.94		
Aloof	4.41*	.10	1.79		

Note. In step 1, the control variable of ND status was entered to predict social vulnerability score. In step 2, the social interaction styles were entered as predictors. The social interaction style 'appropriate' is not entered as a predictor as this is the reference group.
[a] neurotypical = 0, neurodevelopmental (ND) diagnosis = 1.
* $p < .05$, *** $p < .001$

5 CHAPTER 5: A PROFILE OF SOCIAL VULNERABILITY IN CHILDREN AND YOUNG PEOPLE WITH WS

5.1 Chapter Overview

Chapter 4 examined two aspects of everyday social behaviour relevant to social vulnerability: eye contact behaviour and social interaction style. The next two Chapters focus on social vulnerability itself. To date, research on social vulnerability in WS has focused on adulthood and far less is known about its presentation earlier in development. Therefore, the present Chapter aims to establish the profile of social vulnerability in a sample of children and young people (CYP) with WS. While the findings reported in Chapter 4 (published paper 2) provided initial evidence of elevated social vulnerability in CYP with WS, further research is needed to understand this issue in adolescence, which is a critical period for social development and independence. In addition to a sample of CYP with WS, this Chapter includes data from a sample of neurotypical CYP to examine whether levels are elevated and whether there is something different about social vulnerability in WS. The study also extends what is currently known about everyday experiences of social vulnerability by collecting qualitative data on real-life examples of social vulnerability in addition to a measure of 'vulnerable experiences' not previously used in WS research, thus providing new insights into vulnerability in this group. A novel aspect of the study is that it provides the first evidence on the strategies used by families to manage issues of vulnerability and ensure social safety. Having established the profile of vulnerability in this Chapter, the data are taken forward in Chapter 6 to examine how social vulnerability relates to core features of the WS cognitive and behavioural profile.

5.2 Introduction

There is a small but emerging literature on social vulnerability in WS. To date, the handful of studies on this topic are limited to adulthood and have used the Social Vulnerability Questionnaire (SVQ; Fisher et al., 2012) as a tool to capture levels of social vulnerability (Fisher et al., 2012; Fisher, Moskowitz, et al., 2013; Lough & Fisher, 2016b). The SVQ was developed to quantify levels of social vulnerability and identify patterns of social risk in adults with individuals with intellectual and developmental disabilities (IDD), considering the lack of existing measures that met this remit. The measure uses an informant

approach whereby parents/caregivers rate statements about an individual using a Likert scale, with higher scores for the measure as a whole and across each subscale being indicative of greater social vulnerability. An initial exploratory factor analysis of the SVQ with 146 individuals with IDD revealed 30 items that loaded onto six factors or separate ways in which people could be vulnerable, namely Risk Awareness, Parental Independence, Social Protection, Credulity, Perceived Vulnerability and Emotional Bullying, and totalled 49.1% of the variance (Fisher et al., 2012). To date, researchers have administered the SVQ with parents/caregivers of adults with WS and adapted the measure for self-report (Lough & Fisher, 2016b). The SVQ has also been used in cross-syndrome research to determine whether different neurodevelopmental conditions (WS, autism and Down syndrome [DS]) are associated with differences in overall levels of social vulnerability and facets of vulnerability (Fisher, Moskowitz, et al., 2013). The findings from this approach indicate that adults with WS experience overall levels of social vulnerability that are comparable to adults with other forms of IDD, but that there might be syndrome-specific patterns of vulnerability. For instance, there is evidence that adults with WS (average age 25 years) are more vulnerable in the area of parental independence (e.g., being left alone overnight; being with individuals of the opposite sex without supervision) relative to autistic adults and adults with DS. In addition, compared to autistic adults (but not dissimilar to adults with DS), adults with WS are perceived as being more vulnerable through physical appearance (Fisher, Moskowitz, et al., 2013).

Having reviewed the WS literature to date, it is striking that very little research has investigated social vulnerability in childhood and adolescence. Adolescence is often defined as the years between the onset of puberty and beginning of adulthood (typically 10-19 years, but see Sawyer et al., 2018 who suggests 10-24 years is a better fit with the current understanding of development) and is an important stage to examine social vulnerability for several reasons. First, adolescence is a period of significant social change: social networks diversify and expand as young people typically spend more time outside of the family home, bringing with it new social experiences and pressures, and helping to shape a sense of self-identity (Meeus et al., 2005). It is also characterised by significant brain growth and development, particularly in emotion regulation and goal-directed behaviour (Blakemore & Choudhury, 2006). In addition, adolescence is marked by rapid physical maturation which signals a transition to adulthood and may give others the perception that an individual is able to take on greater responsibility. We also know that adolescence coincides with the peak

prevalence of peer victimisation (11-14 years; Hymel & Swearer, 2015). Adolescence is therefore an especially important developmental stage for social interactions.

While little is known about social vulnerability in adolescence in WS, other studies have documented social behaviours during this developmental stage that are relevant in the context of social vulnerability. For example, poor awareness of the risks posed by strangers (Lough, Rodgers, et al., 2016; Riby, Kirk, et al., 2014) and difficulties regulating social approach behaviour and personal space boundaries (Lough, Flynn, et al., 2016), have been reported in research with CYP with WS and are relevant in the context of behaviours that might indicate social vulnerability (Riby et al., 2017). Linked to this, we know from speaking to adults with WS in Chapter 3 that negative social experiences with peers, including bullying, are common in the secondary school/adolescent years. Therefore, the wider social phenotype associated with WS makes adolescence an important developmental stage to understand the profile of social vulnerability.

To my knowledge, Lough (2016)[3] is the only study to have investigated social vulnerability in CYP with WS. This study sampled 23 CYP aged between 8.1 and 16.8 years (mean age 12.4 years) using the SVQ (Fisher, Moskowitz, et al., 2013) and found significantly higher social vulnerability compared to that in a neurotypical group, both in overall levels and across subscales of the SVQ, except for the subscale Parental Independence. The findings reported in Chapter 4 of the thesis (paper 1, sample of CYP aged 6-17 years) lend further support to social vulnerability being elevated in childhood in WS. However, the measure of vulnerability used in Chapter 4 was limited in that it included only a small number of items from the SVQ and therefore prevented meaningful comparison to Lough (2016). It also did not allow for an in-depth look at different facets of vulnerability. The present study adds evidence of social vulnerability in CYP with WS to examine whether levels are elevated and/or different to neurotypical CYP.

How social vulnerability changes across age and different stages of life is still largely unknown. The few studies of adults with WS have reported no association between chronological age and levels of social vulnerability when examined at the composite level (total score on the SVQ). However, there is a small body of evidence to suggest an interesting link between independence-related vulnerability and chronological age/developmental stage. Specifically, both Lough and Fisher (2016b) and Fisher et al. (2013) reported an association

[3] Lough et al. (2016) is a study carried out as part of a doctoral dissertation.

between chronological age and the score on the SVQ subscale 'parental independence', whereby older adults were reported by their parents to be, on average, afforded more independence and hence considered more vulnerable in this domain. As per the design of the SVQ, increased parental independence contributes towards a greater social vulnerability score, on the assumption that greater autonomy (or less parental supervision) increases the risk of an adverse outcome. Therefore, it may be the case that there are age/developmental stage factors associated with vulnerabilities and here the evidence points towards independence-related vulnerability increasing with age. Evidence of other age/developmental stage related aspects of vulnerability include younger adults being less aware of social risk (Fisher, Moskowitz, et al., 2013) and more credulous (Lough & Fisher, 2016b). If greater independence-related vulnerability is associated age, then this is likely to emerge earlier in adolescence when children are typically encountering more opportunities for independence. We currently do not have sufficient data on this yet, hence the current study aimed to address this gap by capturing the profile of social vulnerability, including independence-related vulnerability and its association with age, in CYP with WS.

In addition to quantifying social vulnerability and considering the profile with age, an important question is how social vulnerability presents in the context of everyday situations. In other words, if CYP experience heightened social vulnerability, is this mirrored in their everyday social experiences and outcomes? There is a dearth of literature on this topic, however the available evidence from the adult literature suggests that everyday experiences of victimisation are commonly experienced by adults with WS. In Fisher et al. (2013), 79% of parents identified an instance when their son/daughter had been taken advantage of. Similarly, in the adult sample studied in Lough and Fisher (2016b), this figure was 75% (when asking about experiences in the past year). Of the examples given in Fisher et al. (2013), most experiences related to teasing/persuasion (37%) or money/theft (37%), but also instances of physical or sexual abuse (16%). Less frequently reported in Lough and Fisher (2016b), but noteworthy all the same, were examples of sexual abuse/grooming (9% of those who reported an example), physical abuse (3%) and social risk with unfamiliar people such as letting strangers into the house and giving out phone numbers (frequency not reported). There is currently no evidence on the real-life presentation of social vulnerability in CYP with WS: neither the data reported in Chapter 4, nor in Lough (2016)—the only two existing WS child studies on social vulnerability—go beyond quantifying social vulnerability to explore specific features of vulnerability and document real-life examples of vulnerable

outcomes. The current study addresses this gap in knowledge by providing first evidence of real-life vulnerable experiences of CYP with WS, using the Vulnerable Experiences Quotient (VEQ; Griffiths et al. 2017). The VEQ originates from the autism literature but asks about negative life events that tap into different areas of vulnerability relevant to CYP with WS, such as education, mental health and interpersonal issues. Understanding how vulnerability presents in the everyday lives of CYP with WS is an important step forward in understanding the key areas for support.

Linked to this, there is currently a dearth of research that has explored support or intervention for social vulnerability in WS. The only published study with this aim is an evaluation of an intervention to teach adults with WS about interactions with strangers (Fisher, 2014). If social vulnerability is elevated in CYP with WS, it is likely to be very concerning for the wider family and support network, therefore an important question is how the wider family manages these concerns and ensures their child's safety. It is currently unknown how families navigate the issue of social vulnerability during the childhood/adolescent years; therefore, the current study addresses this question for the first time, taking an exploratory approach to collect evidence on parent/caregiver strategies.

5.2.1 The Current Study

Social vulnerability has been emphasised in review articles on WS and a handful of empirical studies have started to examine this concept (Fisher, Moskowitz, et al., 2013; Lough & Fisher, 2016b; Riby et al., 2017). Yet, to date the profile and nature of social vulnerability has been examined only in adults with WS. The current study examined the social vulnerability profile in a sample of CYP with WS to understand its presence in adolescence, a time of increasing independence and significant social development. In doing so, the study adds to the limited evidence on this topic.

In WS research the SVQ is the only measure that has been used to assess social vulnerability. Therefore, to understand how the profile in the current sample compares to existing reports, it was essential to administer the SVQ. Alongside capturing levels of social vulnerability and examining whether these are elevated compared to neurotypical CYP, the study extends what is currently known about the nature of social vulnerability in WS by capturing data on real-life vulnerable experiences using a measure not used in WS research to date. Therefore, the study provides new insights about the presentation of vulnerability in this group alongside qualitative insights to better understand the real-life manifestations. Finally,

the study sought to document the protective strategies parents and families adopt to manage concerns of social vulnerability. The specific research questions were:

1) Do parents of CYP with WS report heightened social vulnerability (total levels and across domains) in comparison to a sample of neurotypical CYP? Based on evidence of elevated levels in a child sample (Lough, 2016) and the findings reported in Chapter 4, it was predicted that CYP with WS would be reported to have higher levels of overall social vulnerability (SVQ-Total) and across social vulnerability domains (SVQ subscales).
2) What does the profile of social vulnerability look like in terms of different components of vulnerability and is there an association with child chronological age? While evidence from adult studies has indicated no relation between age and overall levels of social vulnerability (Lough & Fisher, 2016b), the relationship in the current age group is currently an open question. Given we do not have any evidence with this age group to make a clear prediction, the study took an exploratory approach.
3) How does social vulnerability present in the everyday lives of CYP with WS? To answer this question, data were collected on examples of vulnerability experienced by CYP. In addition, the pattern of endorsement across a range of negative life events (measured using the VEQ) was examined.
4) What are the common support strategies used by families concerned about social vulnerability? Currently there are no data on this topic, therefore; this question adopted an exploratory approach.

5.3 Method

5.3.1 Participants

WS sample: Thirty-three parents or caregivers of children with WS were recruited to the study. Inclusion criteria included being a caregiver of a child aged 10-17 years with a diagnosis of WS. Exclusion criteria was the child having an additional diagnosis of autism. One parent reported on the demographics questionnaire that their child had co-occurring autism and was excluded from further analysis. The final sample included 32 parents of CYP with WS aged 10-17 years (all birth parents, 29 females/3 males; all of whom reported living with their child at the time of completing the questionnaire). All parents endorsed that their child's diagnosis of WS had been confirmed via genetic testing (n=31) or a cardiologist (n=1). The average age of diagnosis was 23 months, but this ranged considerably from 4

months to 7 years 3 months. The majority of the CYP in the sample (65.6%) were currently attending a school/college for children with special education needs (see Table 5.1 for a breakdown across school provision). Most of the sample (94%, 30/32) reported currently having (or previously having had) a statement of Special Educational Needs (SEN) or an Education, Health and Care Plan (EHCP). Fifty-nine percent of the CYP (n = 19/32) had been diagnosed by a clinician as having a co-occurring condition. Of those with a co-occurring condition, approximately half had multiple co-occurring conditions. The most common was ID (n = 12) and language delay (n = 14), but other conditions featured too[4]. Parents were asked "do you suspect that your child has (or has had) any condition that they have not been diagnosed with?" to which 50% (n = 16/32) reported "yes". The most common suspected condition was an anxiety disorder (n = 9)[5]. Demographic information about the groups is summarised in Table 5.1.

Neurotypical group: Thirty-seven parents/caregivers of neurotypical CYP were recruited. Inclusion criteria included being a caregiver of child aged 6-17 years (the neurotypical group was from a broader age range to act as a reference group due to the vast heterogeneity of functioning levels associated with WS). Exclusion criteria included children with a diagnosis of a developmental condition (e.g., autism, ADHD) or additional support needs (e.g., intellectual disability). The study was also not open to parents of children in the process of diagnostic assessment. Dat from two participants was removed prior to analysis – one parent had not completed the child demographic information and the other reported that their child was on the diagnostic pathway. The final sample for analysis included 35 parents (all female birth parents who lived with the child at the time of answering the questionnaire). One parent reported that their child had received a diagnosis of language delay. In addition, two parents responded "yes" to the question "do you suspect that your child has (or has had) any condition that they have not been diagnosed with?"[6]. One child in the sample was home educated and the remainder were currently attending mainstream school or college (one missing response).

[4] Other cooccurring diagnoses included sensory processing disorder (n = 4), anxiety disorders (n = 4), ADHD/attention deficit disorder (n = 3), depression (n = 1), eating disorder (n = 1), dyspraxia (n = 1)

[5] Other suspected conditions included autism (n = 2), ADHD/attention deficit disorder (n = 4), eating disorder (n = 1), sensory processing disorder (n = 2), language delay (n = 1), dyslexia (n = 2), dyspraxia / developmental coordination disorder (n = 1), Tourette syndrome or tic disorder (n = 1), ID (n = 1).

[6] Suspected conditions included dyslexia (n = 1) and sensory processing disorder (n = 1).

Table 5.1

Participant Demographics

Variable	WS (n = 32)	NT (n = 35)
Mean chronological age in years (SD)	13.93 (2.57)	9.74 (2.8)
Range of chronological age in years	10.00-17.83	6.25-16.67
Gender	12m; 20f	15m; 20f
Current school provision [a]		
Mainstream school/college	6	33
SEN school/college	21	0
SEN unit within a mainstream school/college	4	0
Home educated	1	1
Not currently attending school/college	0	0
SEN/EHCP statement (current or in the past)		
Yes	30	1
No	2	34
Diagnoses		
Presence of a clinical diagnosis [b]	19	1
Suspected conditions (current or historic) [c]	16	2
Parent highest level of education		
No formal quals	0	0
High school/secondary school	2	0
Further vocational quals	13	2
University undergraduate	10	14
University postgraduate	7	19

Note. SEN = Special Educational Needs; EHCP = Education and Health Care Plan

[a] One missing response for the neurotypical (NT) group. [b] For the WS group, this refers to the presence of a cooccurring condition (alongside a WS diagnosis). [c] Parent's own opinion

5.3.2 Materials

Demographic diagnostic history. Information was collected on the child's date of birth, gender, school provision, primary diagnosis (for the WS group e.g., age of diagnosis and method of diagnosis), and diagnosed and suspected co-occurring conditions. The form

also collected information on the parent informant, including their relationship status to the child, living arrangements, date of birth, gender, and highest level of qualification. The full list of demographics questions asked is provided in Appendix C.

Social Vulnerability Questionnaire (SVQ; Fisher et al., 2012)**.** The SVQ is a 30-item parent measure to assess social vulnerability in individuals with IDD, by quantifying levels and identifying domains of risk. The measure been used in research with adults with WS and other forms of IDD (Fisher et al., 2012; Fisher, Moskowitz, et al., 2013; Lough & Fisher, 2016b) and in one study with CYP with WS, as part of doctoral research (Lough, 2016). The SVQ produces a total score of social vulnerability (minimum score 30; maximum 120) and the 30 items load onto six subscales which represent distinct areas in which people with IDD can be socially vulnerable (see Table 5.2 for a description of the subscales). Parents/caregivers are asked to rate statements on a 4-point Likert Scale from 1 (not true or never) to 4 (very true or always). After reverse scoring relevant items on the subscales Risk Awareness and Social Protection, higher scores on each factor indicate greater vulnerability in that domain. Two separate studies running exploratory factor analysis on the SVQ with a large sample of individuals with IDD support the 6-factor structure accounting for 49.1% of the variance (Fisher et al., 2012). The SVQ also includes three open-ended questions which were included in this study: “currently, how worried are you about your child being taken advantage of? What makes you concerned?”, “can you give an example of a time when your child has been taken advantage of in the past six months?” and “what do you do to help prevent your child from being taken advantage of?”.

The SVQ was administered in full for the WS group and showed good internal consistency ($\alpha = .82$) for the measure as a whole. Two of the SVQ subscales showed poor/questionable levels of internal consistency (see Table 5.2 for reliability analysis per subscale). In data collection with the neurotypical group, three disability-specific items were removed from the questionnaire: item 5 (“knows he or she has a disability”; Risk Awareness subscale), item 9 (“others perceive him or her to have a disability; Perceived Vulnerability subscale) and item 26 (“can accurately describe his or her disability to others”; Risk Awareness subscale). The total possible score for this abbreviated measure was 108 (refer to Data Analysis section for further information). Cronbach’s alpha for the abbreviated measure was acceptable for the WS group ($\alpha = .78$) but questionable for the neurotypical group ($\alpha = .63$). To note, for the neurotypical group, three of the SVQ subscales had unacceptable Cronbach’s alpha values (< .5)

Table 5.2

Overview of the Six Subscales of the Social Vulnerability Questionnaire and Reliability Statistics

SVQ subscale	Description	N of items in full measure [N of items in abbreviated measure]	Example item	Cronbach's alpha		
				WS	WS abv [a]	NT abv [a]
Emotional Bullying	Experiences of teasing or otherwise unkind actions performed by others	5	Is often called names by others	.83	-	.75
Risk Awareness	ability to detect and avoid potentially dangerous situations	9 [7]	Knows not to talk to strangers and follows that rule	.81	.77	.72
Social Protection	presence of a protection from a peer group	4	Is considered part of a social group	.65	-	.49
Perceived Vulnerability	Presence of physical characteristics that could be perceived by others as vulnerable	4 [3]	Others perceive him/her to look different from same age peers	.56	.42	.25
Parental Independence	Amount of social autonomy afforded by parents/caregivers	3	Parents allow to be with older individuals with no supervision	.72	-	.73
Credulity	Propensity to believe false information	5	Is likely to believe a claim when there is evidence it should not be believed	.80	-	.42

Note. Cronbach's alpha < .5 = unacceptable, .5-.6 = poor, .6-.7 = questionable, .7-.8 acceptable, .8-.9 – good, >.9 = excellent

[a] items removed from the Risk Awareness (n = 2) and Perceived Vulnerability (n = 1) subscales

Vulnerability Experiences Quotient (VEQ). The VEQ parent report is a 44-item measure of negative life experiences. The parent version used in the current study is unpublished but is based on the self-report version (60 items) developed using a participatory approach for use with autistic adults (Griffiths et al., 2019). Each item of the VEQ is a statement about a life experience e.g., "my child moved school because the school was unable to cater for his/her needs" (see Table 5.6 for the full set of items). Parents were asked to report whether their son/daughter had experienced each event by selecting 'yes', 'no', or 'no opportunity'. At the end of the list of items, there was a text box for parents to add details of any other life experiences they felt were relevant (not analysed for this study). Each item was scored 1 if 'yes', and 0 if 'no' or 'no opportunity' except for three items which were reverse scored (0 if 'yes', 1 if 'no' or 'no opportunity'). The total score was between 0 and 44. Cronbach's alpha for the WS sample was .78, indicating acceptable reliability.

Impact of Covid-19. The study took place during the Covid-19 global pandemic in 2020-2021 and the vast majority of WS individuals were classified as highly clinically vulnerable. Issues of social vulnerability (and other variables studied as part of the research reported in Chapter 6) are likely to have been impacted by the circumstances of repeated lockdowns and school closures (i.e., changes to routines) and new societal rules (i.e., introduction of social distancing), therefore it was important to gather information about potential changes in behaviour during this time. Parents completed a bespoke questionnaire which asked them to report on their child's awareness of Covid-19, change to anxiety levels, awareness of and ability to socially distance from others, as well as parents' concern about their child's friendships and wellbeing during this time. The list of items is provided in Appendix D.

5.3.3 Procedure

Ethical approval was obtained from the Department of Psychology Ethics Committee at Durham University. Recruitment into the study took part during November 2020-January 2022. Families of CYP in the age group were recruited via online flyers and short videos (for each group separately) explaining the aims of the research, and the inclusion and exclusion criteria. For the WS sample, the recruitment materials were distributed via the Williams Syndrome Foundation (a national support charity for individuals with WS and their families), through their newsletters and social media channels. The neurotypical sample was recruited via a University FaceBook page for families interested in taking part in developmental

research. All participants (parents/caregivers) provided written/digital opt-in consent to take part prior to completing the questionnaires.

The data collection formed part of a larger, multi-phase project examining social vulnerability and its relationship with the cognitive and behavioural profile in CYP with WS. Overall, parents completed a total of nine questionnaires which were divided between two packs and posted to participating parents a few weeks apart to make their participation more manageable. Pack 1 included the demographics questionnaire, experience of Covid-19 questionnaire, the SVQ (Fisher et al., 2012), the Social Responsiveness Scale-Second Edition (SRS-2; Constantino & Gruber, 2012) and the Behaviour Rating Inventory of Executive Function, Second Edition (BRIEF-2; Gioia et al., 2015). The second pack of questionnaires included the VEQ (Griffiths, Allison, Kenny, Holt, Smith, & Baron-Cohen, 2019), the Vineland Adaptive Behaviour Scales (VABS-3; Sparrow et al., 2016), the Anxiety Scale for Children with Autism Spectrum Disorder (ASC-ASD; Rodgers et al., 2016) and the Overprotection scale (Clarke et al., 2013). The questionnaires that relate to the aims of this Chapter (to delineate the profile and nature of vulnerability) are described above, and the remaining measures are outlined in Chapter 6. On return of both questionnaire packs, parents received a £10 gift voucher as a thank you for their participation.

5.3.4 Analytic Approach

The dependent variables were individual item-level responses, subscale scores and total scores of the SVQ and VEQ. Six participants (1/32 WS, 5/35 neurotypical) did not return Pack 2 of the questionnaires resulting in missing VEQ data for these CYP. Participants who responded to less than 95% of the SVQ or VEQ (> 2 missing items) were excluded from associated analyses. Missing responses were replaced with 0 when calculating a total SVQ or VEQ score. Participants were excluded from analysis of individual items in the SVQ or VEQ if responses were missing. All statistical analyses were performed using IBS SPSS version 26 or 27. The level of significance was set at $\alpha < .05$, except when adjusted to account for multiple comparisons (stated below). Effect sizes were calculated to aid interpretation of the correlational analyses and group comparisons. For correlations an effect size of .1 is small, .3 is medium, and .5 is large, according to Cohen (1988). For Cohen's *d* an effect size of < .2 indicates a small effect, between .2 and .5 a medium effect and > .8 a large effect.

Given the small sample size, it was important to determine the distribution of the dependent variables for choosing an appropriate statistical test. Exploration of the variable

SVQ-Total using the Shapiro-Wilk statistic indicated that the data was normally distributed for the WS group (W = .06, p = .21), as were the SVQ subscales Emotional Abuse (W = .96, p = .21), Risk Awareness (W = .96, p = .35), and Social Protection (W = .95, p = .13). The subscales Perceived Vulnerability (W = .93, p=.05), Parental Independence (W = .76, p < .001) and Credulity (W = .92, p = .02) were not normally distributed. Examination of the variable VEQ-Total indicated that the data was not normally distributed for the WS group (W = .9, p = .005). Non-parametric statistical tests were used (Spearman's correlations and Mann Whitney U tests) where data varied from the normal distribution. Data were inspected for outliers using boxplots[7]. It was decided that outliers would be retained as extreme scores were expected and these are meaningful in capturing the full range of social vulnerability.

To understand if average levels of social vulnerability were elevated compared to neurotypical children (RQ1), a new total SVQ score was computed for the WS group, with the three disability-specific items removed, therefore mirroring the neurotypical measure. This produced a new total score and two new subscale scores (see section 5.3.2 for the removed items), which were used in independent samples t-tests to make comparisons between the WS and neurotypical groups on SV-Total and SVQ subscale scores. Alpha values were adjusted using Bonferroni correction for the six subscale comparisons.

To describe the profile of social vulnerability and examine the potential association with chronological age (RQ2), the SVQ was scored and analysed using all items, as per the original measure to allow for comparison with previous literature. The SVQ subscales vary in the number of items, therefore, to allow for comparison across subscales, the mean score per item was calculated. Pearson's/Spearman's correlations were used to examine the relationship between SVQ scores and chronological age.

To examine how social vulnerability presents in the everyday lives of CYP with WS (RQ3), two approaches were taken. First, by examining the frequency of life experiences endorsed on the VEQ, using the neurotypical pattern as a comparison. An exploratory Pearson correlation was performed to analyse the relationship between the SVQ and VEQ. Second, by analysing parents' qualitative reports in response to the open-ended question "can you give an example of a time when your child has been taken advantage of in the past six months?". Taking an inductive approach, responses were coded first by ER to identify the

[7] Outliers were identified for the SVQ subscale emotional abuse (n=1, score 18); social protection (n = 2, both scores of 16); and parental independence, (n=3, all scores of 9). One outlier was identified on the VEQ (score of 21).

nature of the vulnerable example given. Codes were then reviewed by two further coders to ensure inter-rater reliability and any discrepancies were resolved through discussion.

Finally, to assess parents' concern about social vulnerability, responses to the question "currently, how worried are you about your child being taken advantage of? What makes you concerned?" were analysed quantitatively to identify endorsement of concern. Where parents did not report concern, these responses were further examined at the qualitative level by assigning codes. To examine the type of strategies implemented by families to reduce the risk of social vulnerability, responses to the question "what do you do to help prevent your child from being taken advantage of?" were analysed qualitatively. Participant quotes are used to add richness to the data for research questions 3 and 4. Names in quotes have been pseudonymised.

5.4 Results

First, contextual data on social behaviour and anxiety during the Covid-19 pandemic are outlined in Table 5.3, before addressing the core research questions. All parents reported that their child was aware of Covid-19. Over half of the parents in the neurotypical group reported that their child was not experiencing anxiety prior to the pandemic and that this had not changed. In contrast, over 60% of parents in the WS group endorsed that anxiety levels had increased. The overwhelming majority reported that their son/daughter with WS had been anxious about changes to seeing friends and about keeping their distance from others ("social distancing"), whereas this was evidenced to a lesser extent in the neurotypical group. Note that while WS and neurotypical recruitment did crossover, most of the recruitment to the WS group was conducted first. Therefore, the impact and memory of Covid-19 may have been more salient for parents in the WS group.

Table 5.3

Descriptive Results about Child Behaviour During the Covid-19 Pandemic (%)

Variable	WS (n = 32)	NT (n = 35)
Child is aware of Covid-19	100%	100%
Chane in child's anxiety levels		
Anxiety not present before Covid-19 and no change	6%	57%
Anxiety present before Covid-19 and no change	31%	9%
Anxiety levels increased somewhat/significantly	63%	34%
Is/has been anxious about changes to routines with friends	91%	51%
Is/has been anxious about social distancing	94% [a]	20%
Social distancing behaviour		
Child is aware of the need to social distance	88%	97%
Child appropriately social distances/ed	63%	89%
Parent concerned about impact on child's friendships [b]	87%	49%
Parent concerned about impact on child's emotional wellbeing[c]	94%	74%

[a] One missing response (19/31). [b] somewhat concerned: 59% (WS), 40% (NT); very/extremely concerned: 28% (WS), 9% (NT). [c] somewhat concerned: 59% (WS), 69% (NT); very/extremely concerned: 35% (WS), 6% (NT)

5.4.1 RQ1: Is Social Vulnerability Heightened?

The first research question sought to examine whether social vulnerability in the WS group was elevated. As there is no clinical cut-off for the SVQ, comparison with the neurotypical pattern provides helpful context as to whether levels are elevated on average. Analyses explored potential differences in parent-reported social vulnerability scores (total levels and across domains) between the WS and neurotypical groups using the abbreviated SVQ measure. As shown in Table 5.4, the WS group scored significantly higher on the SVQ-Total compared to the neurotypical group with a large effect size. The WS group also scored significantly higher on the SVQ subscales Emotional Abuse, Risk Awareness, Social Protection, Perceived Vulnerability and Credulity, all with large effect sizes thereby indicating greater vulnerability across these domains in the WS group (see Table 5.4). As illustrated in Figure 5.1, the SVQ subscale Parental Independence showed the opposite pattern, with the WS group scoring lower than the neurotypical group, on average. A lower

score on the Parental Independence subscale (and all subscales of the SVQ) is indicative of less social vulnerability in this domain.

Table 5.4

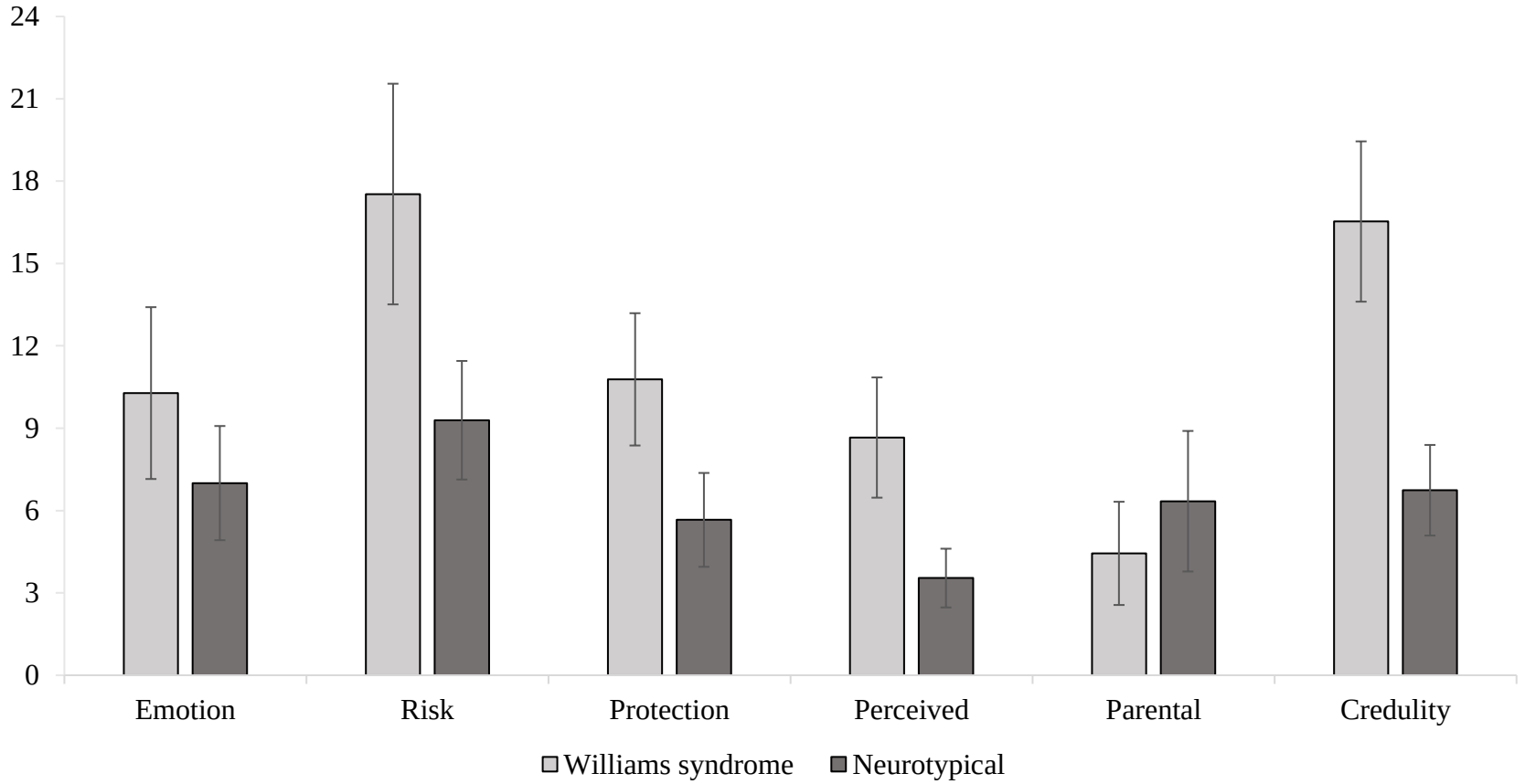
Descriptive Statistics and Group-Level Comparisons on the Abbreviated Social Vulnerability Questionnaire (SVQ)

SVQ (total possible score)	WS (n = 32)		Neurotypical (n= 35)		Group difference				
	Mean (SD)	Range	Mean (SD)	Range		t	df	*p*	*d*
SVQ-Total (108)	68.22 (9.08)	50-87	38.57 (5.29)	31-56	WS > NT	16.13	48.94	< .001	3.99
Emotional abuse (20)	10.28 (3.13)	3-18	7.00 (2.08)	5-13	WS > NT	5.09	65	< .001	1.23
Risk Awareness (28)	17.53 (4.02)	11-26	9.29 (2.16)	7-15	WS > NT	10.33	46.64	< .001	2.55
Social Protection (16)	10.78 (2.41)	2-16	5.66 (1.71)	4-9	WS > NT	10.11	65	< .001	2.45
Perceived Vuln (12)	8.66 (2.19)	3-12	3.54 (1.07)	3-7	WS > NT	11.96	43.98	< .001	2.97
Parental Ind (12)	4.44 (1.88)	2-9	6.34 (2.56)	3-12	WS < NT	-3.45	65	.001	0.85
Credulity (20)	16.53 (2.92)	3-20	6.74 (1.65)	5-12	WS > NT	16.69	48.05	< .001	4.13

Note. Independent sample t-tests are reported for the group comparison. Levene's Test for Equality of Variance indicated unequal variances between groups for SVQ-Total, Risk Awareness, Perceived Vulnerability and Credulity, therefore adjusted Welshes *t*-statistic and adjusted *P* values are reported for these variables. Adjusted α level = .008 to account for the six multiple comparisons.

Figure 5.1

Bar Chart Showing the Average Score Across the Subscales of the Social Vulnerability Questionnaire



Note. subscales vary in maximum possible score: Emotion = 20, Risk = 28, Protection = 16, Perceived = 12, Parental = 12, Credulity = 20. Error bars represents the standard deviation.

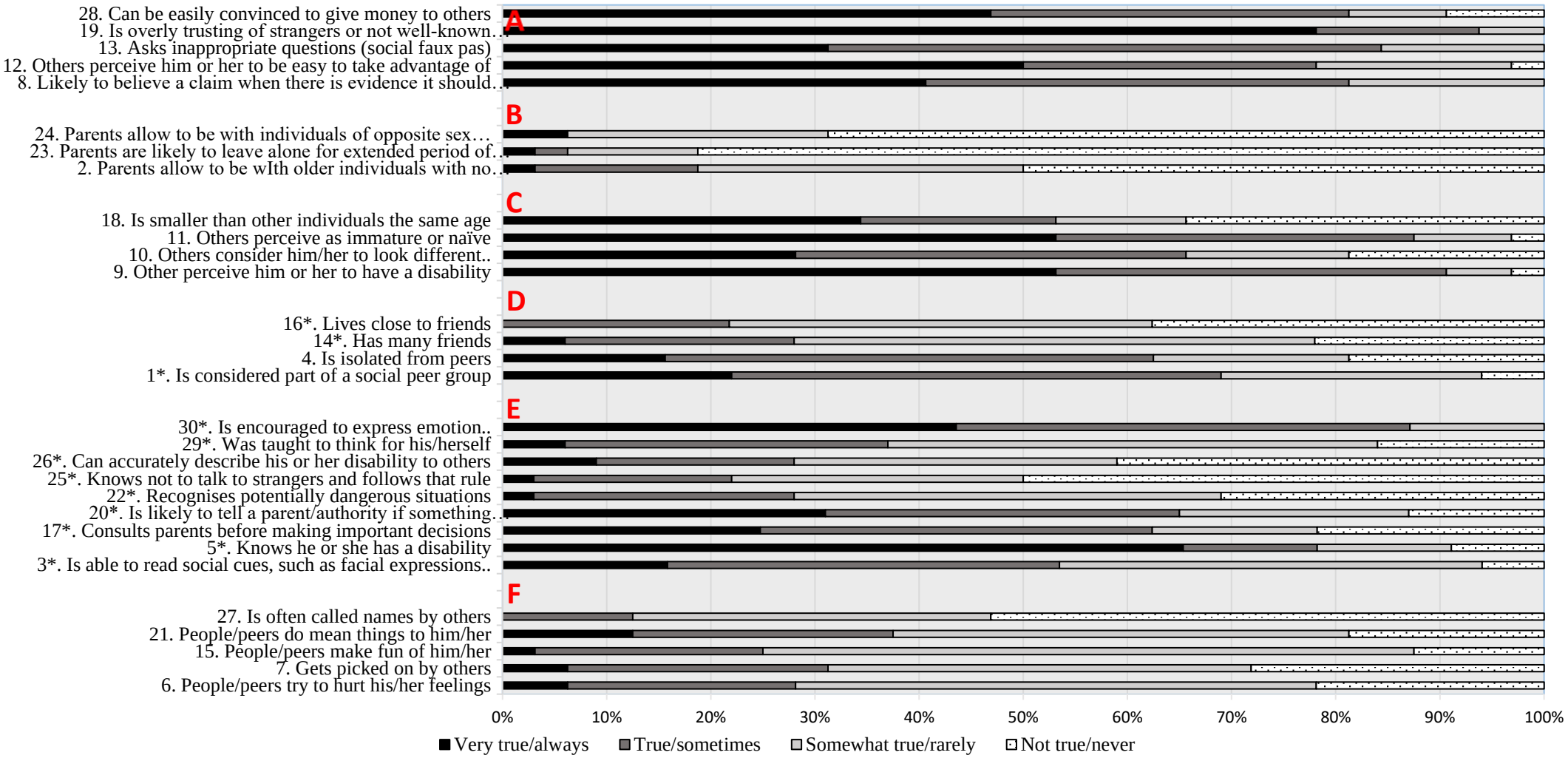
5.4.2 RQ2: What is the Profile of Social Vulnerability and its Association With Age?

The second research question sought to examine the profile of social vulnerability in the WS group. To answer this question and compare to existing literature, the full SVQ measure was used (30 items, as published in Fisher et al., 2012). Descriptive results for the WS sample on the SVQ Total score and subscale scores are summarised in Table 5.5 and item-level data are shown in Figure 5.2. It is worth noting that the average total score in the current sample (all items of the SVQ) was comparable to that reported in the only other study to use this measure with a sample of CYP with WS (76.35 in Lough, 2016) and is higher than that reported by parents of adults with WS (69.43 in Lough & Fisher, 2016b). To examine endorsement across different aspects of vulnerability, frequency was examined across the SVQ subscales (using mean score per item as subscales vary in their items). Of the six SVQ subscales, items within the subscale Credulity were most frequently endorsed, followed by Perceived Vulnerability, Social Protection, Risk Awareness, Emotional Abuse and Parental Independence. This pattern of endorsement is illustrated in Figure 5.2. On closer inspection of the most frequently endorsed subscale, Credulity, 81% of parents endorsed that their child can be easily convinced to give money to others, 94% reported their child as overly trusting of strangers/not well-known individuals and 78% said others perceive him/her to be easy to take advantage of. Examination of the least endorsed SVQ subscale, Parental Independence, showed that at least half of the parents did not endorse the three items on this subscale – 69% reported 'not true/never' to "parents allow to be with individuals of opposite sex with no supervision", 81% reported 'not true/never' to "parents are likely to leave alone for an extended period of time (overnight)" and 50% of parents reported 'not true/never' to "parents allow to be with older individuals with no supervision".

Correlations (Bonferroni corrected) were performed to investigate potential associations between child chronological age, SVQ-Total, and each of the six SVQ subscales. The correlation between age and SVQ total score was weak and not significant (r = -0.34, p = .06), but showed a negative relationship, suggesting that younger children were reported to be more socially vulnerable, albeit not at a meaningful level. As shown in Table 5.5, of the six SVQ subscales, only Social Protection was significantly associated with child chronological age (r = -.5, p = .004). Thereby indicating that younger children were more vulnerable due to having less social protection (less support from a social network).

Figure 5.2

Item-Level Endorsement on the Social Vulnerability Questionnaire for the Williams Syndrome Sample



Note. A = credulity; B = parent independence; C = perceived vulnerability; D = social protection; E = risk awareness; F = emotional bullying. Items marked with an Asterix (*) are reverse scored – less endorsement on these items is associated with greater vulnerability.

Table 5.5

Descriptive Statistics for the Social Vulnerability Questionnaire and Relationship with Chronological Age for the WS Group (SVQ)

SVQ	Mean (SD)	Range [total possible score]	Mean score per item (SD)	Correlation between SVQ and chronological age	
				Co-efficient	p
SVQ-Total	76.31 (10.43)	58-99 [120]		-.34	.06
Emotional abuse	10.28 (3.13)	5-18 [20]	2.06 (0.24)	-.14	.44
Risk Awareness	22.22 (5.19)	14-34 [36]	2.47 (0.54)	-.29	.10
Social Protection	10.78 (2.41)	6-16 [16]	2.70 (0.37)	-.5*	.004
Perceived Vuln	12.06 (2.64)	6-16 [16]	3.02 (0.38)	.03	.86
Parental Ind	4.4 (1.88)	3-9 [12]	1.48 (0.18)	.12	.51
Credulity	16.53 (2.92)	9-20 [20]	3.31 (0.21)	-.22	.22

Note. Higher mean scores represent greater social vulnerability. Perceived Vuln = perceived vulnerability; Parental Ind = Parental Independence.

*Significant at the 0.008 level (2-tailed) (adjusted α-value to align with the 6 subscale comparisons

5.4.3 RQ3: How Does Social Vulnerability Present in the Everyday Lives of CYP With WS?

Research question 3 was addressed in two ways. First, by exploring the VEQ data about the frequency of real-life vulnerable experiences. Second, by examining the real-life examples of vulnerability reported by parents.

VEQ results. The neurotypical VEQ data were used alongside the WS VEQ data to contextualise the frequency of negative life events experienced by the WS sample. Of those who returned the VEQ (31/32 WS, 30/35 neurotypical), two further cases (1 WS, 1

neurotypical) were excluded from analysis at the total level due to <95% of items completed. Out of the 44 life experiences listed in the VEQ, CYP in the WS group had experienced 6.97 on average whereas this figure was 2.03 for the neurotypical group. The number of experiences endorsed ranged from 0 to 20 for the WS group, showing greater variability than for the neurotypical group (range 0-8). Indeed, 30 items on the VEQ were not endorsed by any parent in the neurotypical group, whereas this was only 12 items in the WS group. As the measure is scored using binary 0 or 1, it is more informative to analyse at the item-level. Table 5.6 shows the number of parents/caregivers in each group who endorsed that their child had experienced each type of vulnerability experience on the VEQ (listed in order of most-to-least frequently endorsed for the WS group). The most frequently experienced life events tended to be those that were interpersonal in nature. For both groups, the most frequently endorsed negative experience was "my child has been left out of activities by his/her peers". An exploratory analysis was run to explore whether the frequency of negative life experiences was associated with levels of social vulnerability for the WS group. The correlation between SVQ-Total score and VEQ-Total score was significant, meaning that a greater number of negative life events was associated with greater social vulnerability, r = .41, p = .02 (see Figure 5.3).

Figure 5.3

Scatterplot Showing the Significant, Positive Relationship Between Total Scores on the Social Vulnerability Questionnaire (SVQ_Total) and the Vulnerability Experiences Quotient (VEQ_Total)

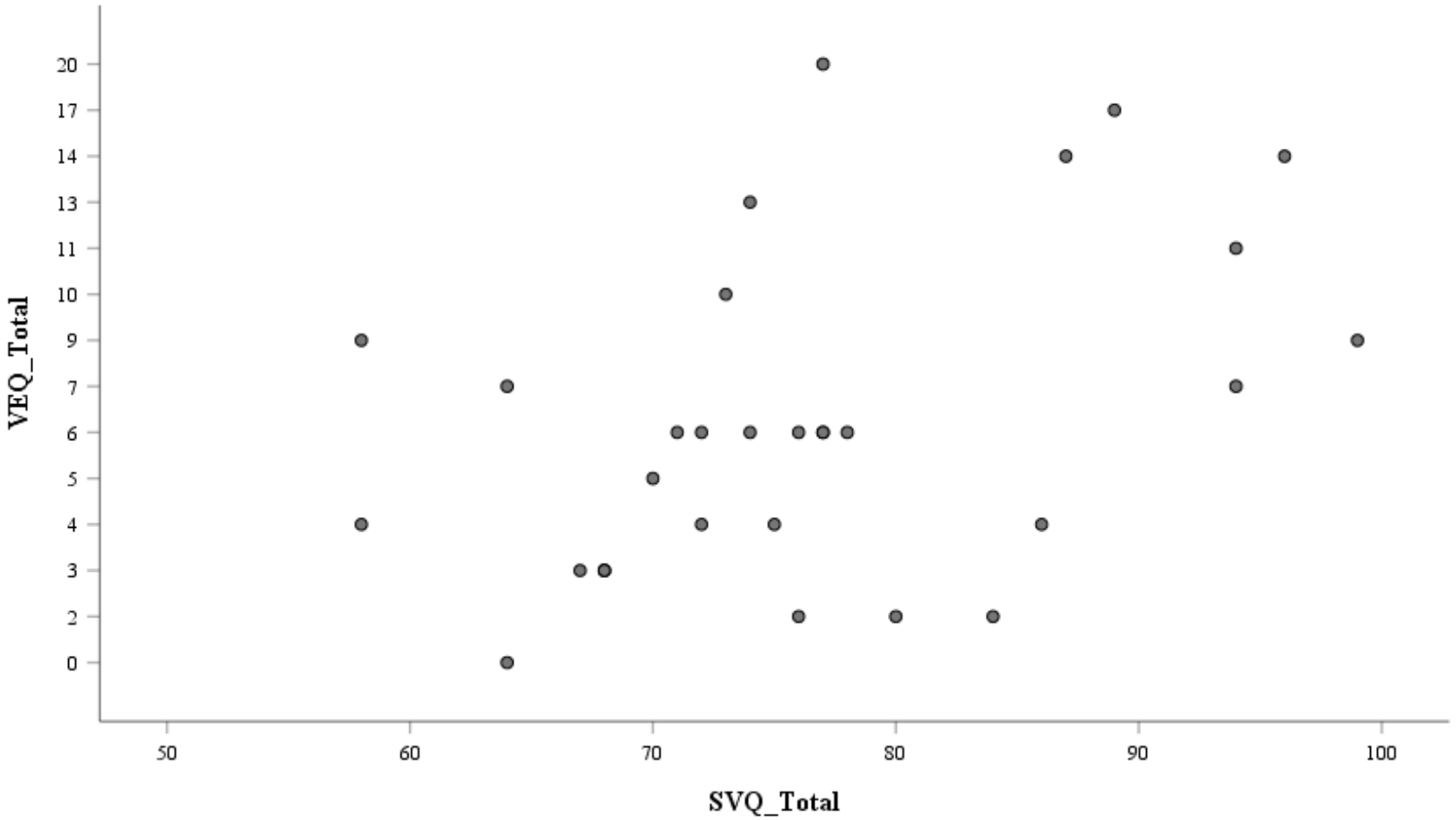


Table 5.6

Percentage of Parents Who Responded 'yes' for Each Item on the Vulnerability Experiences Quotient, in Order of Endorsement as per the WS Group.

Item	WS (n)	NT (n)
4. My child has been left out of activities by his/her peers	77 (24/31)	40 (12/30)
12. My child has been called names or insulted by his/her peers	63 (19/30)	27 (8/30)
1. My child has been bullied by someone that s/he considered to be a friend	58 (18/31)	23 (7/30)
14. My child has been bullied by his/her peers	48 (15/31)	23 (7/30)
20. My child has a mental health condition that has affected his/her daily life	42 (13/31)	0 (0/30)
42. My child does not have friends who he/she sees outside of school*	39 (12/31)	3 (1/30)
21. My child moved school because the school was unable to cater for his/her needs	35 (11/31)	0 (0/30)
13. My child does not have friends who s/he speaks to over the telephone/internet*	32 (10/31)	37 (11/30)
15. My child has been tricked or pressured into breaking rules	32 (10/31)	7 (2/29)
32. My child has had rumours spread about him/her or been talked about unkindly by his/her peers	32 (10/31)	17 (5/30)
3. My child has been hurt by his/her peers badly enough that it left marks on his/her body (e.g., bruises or scratches)	26 (8/31)	10 (3/30)
2. My child has been taken advantage of by someone who was his/her girl/boy friend	23 (7/30)	7 (2/30)
7. My child has been sworn at or called names like stupid, ugly or lazy by an adult	19 (6/31)	0 (0/29)
33. My child often gets into fights with their peers	19 (6/31)	0 (0/30)
37. My child has avoided attending school/college because s/he found it too stressful	16 (5/31)	0 (0/30)
19. My child has not made friends with other children at school*	13 (4/31)	7 (2/30)
27. My child has been humiliated, embarrassed, or scared by an adult	13 (4/31)	4 (1/28)
28. My child has talked about suicide	13 (4/31)	0 (0/30)
35. My child has been tricked or pressured in to giving someone money or possessions	13 (4/31)	3 (1/30)
41. My child has been refused additional support at school when I have asked for it	13 (4/31)	0 (0/30)
6. My child has been bullied by someone in his/her family	10 (3/31)	3 (1/30)
17. My child has deliberately harmed him/herself	6 (2/31)	0 (0/30)
18. My child missed more than 2 weeks of school/college due to anxiety, depression or another reason related to his/her mental health	6 (2/31)	0 (0/30)
29. My child has been pressured into sexual activity	6 (2/31)	0 (0/30)
38. My child is often in trouble for misbehaving at school	6 (2/31)	0 (0/30)
8. My child was subject to a child protection investigation due to concerns about his/her care	3 (1/31)	0 (0/30)
16.My child was charged with a criminal offence	3 (1/31)	0 (0/30)
22. An adult has made sexual comments or advances towards my child	3 (1/31)	0 (0/29)
24. My child has been in trouble for inappropriate sexual behaviour	3 (1/31)	0 (0/30)
25. An educational, medical or social work professional questioned my ability to care for my child	3 (1/31)	0 (0/30)
34. My child has been physically forced into sexual activity	3 (1/31)	0 (0/30)

Item	WS (n)	NT (n)
44. My child was referred to social services due to concerns about his/her care	3 (1/31)	0 (0/30)
5. My child has a criminal record	0 (0/30)	0 (0/30)
9. My child was incorrectly diagnosed with a mental health condition (e.g. ADHD instead of autism)	0 (0/31)	0 (0/30)
10. My child left a school/college without a qualification because s/he failed exams	0 (0/31)	0 (0/30)
11. My child regularly uses alcohol or another (non-prescribed) drug	0 (0/31)	0 (0/30)
23. My child was temporarily or permanently excluded from school	0 (0/31)	0 (0/30)
26. My child has attempted suicide	0 (0/30)	0 (0/30)
30. My child has been hurt by an adult badly enough that it left marks on his/her body	0 (0/31)	0 (0/30)
31. My child has been sectioned because of a mental health condition	0 (0/31)	0 (0/30)
36. My child was arrested by the police	0 (0/31)	0 (0/30)
39. My child spent time in a juvenile detention centre	0 (0/31)	0 (0/30)
40. My child was taken into the care of the local authority	0 (0/31)	0 (0/30)
43. My child was cautioned by the police	0 (0/31)	0 (0/30)

Note. Items with asterisks (*) indicate the item is positively worded on the VEQ but has been reworded here to align with the majority of items.

Real-life examples: To better understand the real-life nature of social vulnerability and outcomes experienced for CYP with WS, parents were asked (via the SVQ) to report an example of a time when their child had been taken advantage of in the past six months. Of the 32 respondents who completed the SVQ, 15 (47%) provided an example in response to this open-ended question. Fifty-three percent of the sample (17/32) therefore did not report an example of their child being taken advantage of in the past six months, of which seven parents provided further explanation. Parents cited the Covid-19 lockdown and subsequent lack of interaction (n = 3) and constant supervision (n = 4) as reasons for providing an example. For instance, one parent commented: "She is never left without supervision. We are very conscious of her trusting nature. At her previous school [name] was found in a compromising position with a boy." Another parent commented "No, because we have been in lockdown, he has had very few interactions with people in a position to take advantage of him." Of the 15 parents who did provide an example, some of the responses described a specific event, whereas others were more general descriptions of indicators of vulnerability. All responses were coded into categories of experience (see Table 5.7). Examples mostly related to instances of the child experiencing social pressure, such as being manipulated or coerced (n = 13).

Table 5.7

Real-Life Examples of Social Vulnerability Reported by the WS Group (n=15)

Category	N	Example
Manipulation / peer pressure	7	She was made to feel like she had done something wrong because her peers didn't like how she was friends with someone else. They pressured her into telling the person she said not to be their friend anymore and bullied her on social media until she did that.
Coercion	3	Another child (the same age) in his special school produced a stock cube and told my child to eat it (which he did). This was a very dangerous situation because: my child often chokes on food and requires first aid […]; my child did not ask what it was or where it had come from or worry about the consequences.
Bullying	3	At school her friends can sometimes be mean and on zoom calls in the evening things can get nasty when they start picking on each other. But she can hold her own and only gets upset rather than being taken" advantage of. Sometimes her friends will ask her to set up a play date or they won't be her friend anymore if she doesn't try.
Taking the blame	1	Polly can often be blamed for things in certain situations, when it's not always clear if she was at fault. Because she is unable to articulate herself as well as her peers, she can often be claimed to be in the wrong, when she might not be, i.e., for taking food from the fridge at home for example.
Handing over possessions	1	She gave her friend a toy at school she'd had for Christmas that she took to school to show and tell.

Note. N = number of parents who gave an example in a category out of a total of 15 parents reporting an example.

5.4.4 RQ4: Parent Concern and Strategies Used to Mitigate Social Vulnerability

The final research question sought to extend the focus beyond the person with WS, to consider parental concern and strategies used by families to mitigate social vulnerability. This RQ took an exploratory approach with no hypotheses set.

Concern: When asked the question "currently, how worried are you about your child being taken advantage of" most parents (24/32) endorsed concern. For instance, parents commented:

> I am very worried about him being taken advantage of. However, he is only 10 and has a full-time 1:1 TA at school so he is rarely alone with an adult present. Children seem to instinctively know that he is different and hence vulnerable.

> I am concerned about Ruth being taken advantage of as Ruth is incredibly trusting of others and has no awareness of stranger danger. Ruth will often do things people tell her to do to please people, regardless as to whether it is wrong or right.
>
> She wants to please people and loves making people happy. She wants to buy presents for people for Christmas. I am worried that she will not be able to see when people will take advantage of her by pretending that being a friend means that you have to pay or give everything you have.

Of the 24 parents who reported being concerned, seven linked their concern to the child getting older and how this would impact on their safety in the social world:

> Highly concerned, especially as he gets older. He knows the rules regarding stranger danger but cannot always follow them, or he will behave inappropriately and shout at point to a stranger who has not even approached him. If someone tried to take him away and knew the right bribes to offer him, he would go.
>
> Very - as he gets older and wants more independence, I am increasingly concerned he could be convinced to do almost anything.

For the eight parents who expressed less or no concern about social vulnerability, reasons mirrored those given by parents who could not provide an example of their vulnerability (as per RQ3) – issues related to limited interaction due to Covid-19 and enforced supervision. One parent endorsed lack of concern due to good school support. Examples include:

> She is rarely away from home except at school or clubs with friends. So we know she is safe wherever she goes. We would never allow her to be in a situation where we thought she would or might be taken advantage of. She is very naïve and vulnerable so there is always someone looking out for her.
>
> Currently lucky, in a mainstream school with EHCP and has amazing supportive teachers and 6 friends also with special needs who she spends a lot of time chatting too. Think she is still in a safe school bubble but not sure when she leaves school.
>
> Not overly worried at the moment as the opportunities for this to happen are rare and limited. However, if someone she went to school with (primary) who she liked were to fabricate something she would still trust them and believe them- it would then by an internal battle between what they asked and upsetting me.

Strategies: In response to the question "what do you do to help prevent your child from being taken advantage of?" Answers related to four main categories, and these are outlined in Table 5.8. In outlining the strategies used, some parents reported more than one

strategy, indicating that they are using multiple strategies to reduce the risk of their child experiencing an adverse outcome. An example of this is highlighted in the quote below.

> Almost never leave him with peers unsupervised. If we do his brother is almost always present or we check on him very regularly. We try to educate other people about his vulnerability. We are slowly trying to educate and demonstrate situation-specific appropriate behaviour and trying to teach him to say 'no' if he doesn't like what somebody is doing. This is very tricky.

Table 5.8

Strategies Reported by Parents and Carers to Manage Social Vulnerability Concerns/Risk

Strategies	endorsement[8]	Example quotes
Child education on the dynamics and dangers of social interactions and relationships	17	Social stories, have set phrases 'we do not x, y, z because x, y, z'... We try to talk about 'what ifs'. Get my child to 'goal plan, do, check' and think things through e.g., teach him to always be suspicious 'question everything' e.g., a stranger or problem may not be obvious – women/children can be just as unkind as a man. I explain to her that we only give gifts on special occasions. Because she want people to love understand her. I explain that it is not because they tell her that they love her that she had to give what she has. We talk and act out scenarios, he also learns about it at school. We teach him to talk to people he trusts like his family, us. Ultimately it is a big worry
High levels of parental supervision / restricting opportunities for social interaction	17	Although he is 15 ½ he does not ever do anything unsupervised – e.g. go to the shops, go to the park etc. there is always an adult present to make sure he is not taken advantage of or hurt in any way. He lives a very sheltered life. Becky is constantly supervised by an adult, and if someone is not on her shoulder, we always make sure we can see where she is and who is with her. I will intervene if I feel Becky is being inappropriate or if I feel uncomfortable about who she is talking to, in a park for example. We are always with him or another trusted adult is. We never leave him unsupervised. All his social interactions are carefully considered by us. Limit the people she is around. We don't socialise much. She doesn't go to any mainstream clubs. She needs supervision at all times. When out, she doesn't go to the toilet alone or go to kids clubs
Educating others about WS and vulnerability	3	.. I have visited her school and done presentations on WS to teachers and older children to flag up her vulnerability. I will speak with her friends and her and make her understand boundaries and limitations of events or actions. We try to educate other people about his vulnerability.

[8] Where parents reported multiple strategies, these were coded separately, therefore this colum does not total 100%

5.5 Discussion

The findings add to the small body of literature on social vulnerability in WS, which to date has focused almost entirely on adulthood (Fisher et al., 2012; Fisher, Moskowitz, et al., 2013; Lough & Fisher, 2016b). Indeed, the current study is the first to detail the profile of social vulnerability during childhood/adolescence in WS, a time of changing social dynamics and increased independence. It also provides new evidence on the profile of social vulnerability during this developmental stage, combining evidence across the SVQ (Fisher et al., 2012; Fisher, Moskowitz, et al., 2013) and VEQ (Griffiths et al., 2019), together with exploratory qualitative insights. The study also provides the first insights into the strategies used by parents and families to manage concerns of social vulnerability.

5.5.1 Elevated Social Vulnerability in CYP with WS

As predicted, and in line with the one other study on social vulnerability in a similar age group (Lough, 2016), parents in the current study reported elevated levels of social vulnerability overall compared to parents of neurotypical CYP. Indeed, the average SVQ total score was near-equal to that reported in the only other child sample (Lough, 2016). The results at the SVQ subscale level lend further support to heightened levels of SV in WS across broad areas of vulnerability, including emotional abuse, risk awareness, social protection, perceived vulnerability, and credulity. We did not find evidence of greater independence-related vulnerability in the WS group, and this finding is addressed further in relation to the overall profile. These findings build upon and support the finding of elevated social vulnerability reported in Chapter 4 by using a more comprehensive measure of social vulnerability.

In comparing the current findings with the existing literature on social vulnerability in adults with WS, we see evidence that heightened levels are evident well before adulthood. The average social vulnerability score in the current sample was higher than levels reported in a sample of adults with WS (Lough & Fisher, 2016b). Compared to the adult levels reported in Fisher et al. (2013), the CYP in the current sample were reported on average to have higher scores (and therefore more socially vulnerable) in the domains of emotional abuse (current child sample M = 10.28, adult sample in Fisher et al. 2013 = 8.5), risk awareness (current child sample M = 22.22, adult sample M = 16.38) and credulity (current child sample M = 16, adult sample M = 13.72). In other words, not only is social vulnerability elevated in adolescence in WS, but parents are reporting children are less aware (compared to

adults with WS) of potential social risks associated with victimisation, experience greater emotional abuse and are more credulous. The same pattern of results was found in the only other study to measure social vulnerability in a child sample (Lough, 2016). In contrast, parents of adults with WS report greater parental independence compared to the parents of CYP in this study. Overall, the results highlight that social vulnerability is a significant issue earlier in development than has been previously studied in WS and thus requires understanding and support well before adulthood.

5.5.2 Profile of Social Vulnerability and Association With Age

The research builds upon the one existing study to have documented social vulnerability in CYP with WS (Lough, 2016) by moving beyond total levels to describe the profile of social vulnerability, examining item-level and subscale-level across of the SVQ. We found that parents most endorsed the SVQ subscale Credulity. According to these data, we can infer that the high levels of social vulnerability in CYP with WS may be particularly linked to high levels of credulity (the effect size for this subscale was largest when comparing the WS and neurotypical groups). As introduced in Chapter 1, credulity refers to a tendency to believe things that are unproven or unlikely to be true (Greenspan et al., 2001) and has been identified as a key factor in models of social vulnerability (Pinsker et al., 2011). High endorsement of credulous behaviours links to existing evidence of high levels of trusting behaviours towards strangers (Lough, Rodgers, et al., 2016; Riby, Kirk, et al., 2014) and difficulty making trust evaluations on emotion / mental state recognition tasks compared to chronological-age and verbal-matched controls (Hanley et al., 2013). It also accords with the earlier finding reported in Chapter 3 that adults with WS described some questionable reasoning around trust judgements and difficulties discriminating intentions of others. Researchers have argued that the hallmarks of the WS social profile, including indiscriminate trust and difficulty evaluating socio-communicative cues, make social vulnerability particularly relevant in this group (Jawaid et al., 2012; Riby et al., 2017). The current study provides evidence that these are highly endorsed features of the vulnerability profile for CYP with WS. However, researchers have differentiated credulity from trust in that trusting people will generally believe information to be true, until given evidence to suggest otherwise. However, a credulous person is likely to believe information even in the face of false or deceptive information (Pinsker et al., 2011). Therefore, further work is needed to disentangle the construct of credulity to understand how it presents in CYP with WS and the factors underlying credulous behaviours.

It is interesting that of the different facets of vulnerability examined in the SVQ, parents least endorsed vulnerability related to parental independence. Indeed, the majority of parents did not endorse social independence in their child aged between 10-17 years (approx. 70% of parents sampled disagreed with the independence statements across this subscale). The neurotypical findings on the same subscale provide useful context here, as parental independence was the only facet of social vulnerability where neurotypical CYP scored higher (greater vulnerability) compared to the WS group, thereby indicating that neurotypical CYP are afforded more independence during this developmental stage. In a cross-syndrome study of adults with IDD, parents of adults with WS reported being more likely to leave their son/daughter unsupervised (more parental independence) compared to parents of autistic adults and adults with DS (Fisher, Moskowitz, et al., 2013). Although the two study designs were different—Fisher et al. (2013) was cross-syndrome and here the comparison is between WS and NT—the lack of endorsement about parental independence in this study suggests that CYP with WS may not be given independence from parents, and perhaps are less vulnerable in this domain. However, this interpretation rests on the assumption that greater independence makes individuals more vulnerable, and—the reverse scenario—greater supervision equates to less vulnerability. An important issue that has not yet been addressed is the relation between supervision and vulnerability. It is possible high levels of supervision/protection may reinforce vulnerability if CYP are less exposed to social situations and have fewer opportunities to develop skills. This point is addressed further when discussing measurement limitations and implications for support.

In detailing the profile, one aim was to examine potential age-related associations with social vulnerability. While previous research with adult samples has reported no relationship with age (Lough & Fisher, 2016b), it was important to reconsider this relationship in adolescence when we see increasingly complex and changing social expectations. However, consistent with the adult literature, we found no meaningful link between overall levels of social vulnerability and age in the sample of CYP, beyond a trend towards younger CYP with WS being more socially vulnerable. These data must be interpreted with caution because of the small sample size and age range studied (10-17), yet it is worth considering this finding. One interpretation is that social vulnerability does not increase as CYP with WS get older, during a developmental period associated with greater opportunities for social engagement. However, we know from the broader profile of vulnerability that parents did not endorse independence and, linked to this, we found no

evidence of age-related change in levels of independence-related vulnerability (parental independence subscale). This differs from adults WS studies which have reported that adults are afforded greater parental independence with age (Fisher, Moskowitz, et al., 2013; Lough & Fisher, 2016b). The evidence here suggests, at the group level, CYP with WS are not afforded more independence as they get older (but note this may explained by within-group variability in functioning and support levels). Therefore, the lack of independence afforded to CYP with WS is perhaps protecting against vulnerability at an age when independence usually increases, and we might expect more vulnerability-related challenges. Again, some caution is required about drawing conclusions about age-related change at the subscale level, given that a number of subscales consist of few items (e.g., 3 items on the parental independence subscale). Future research that spans a wider developmental spectrum, from childhood to young adulthood, will make an important contribution in capturing potential age/developmental-related change in social vulnerability in WS. Research that takes a longitudinal approach to understand developmental changes in social vulnerability (tracking the same individuals through adolescence and into adulthood) would be the best way to explore this issue in future research to avoid the confound of individual differences associated with WS. However, it is noted that this would be a significant research investment.

5.5.3 Real-Life Manifestation of Social Vulnerability

A unique and important aspect of the study was to document the real-life presentation of social vulnerability during this stage of development, using insights from the VEQ and parent-reported examples. First, findings from the VEQ indicate that the most frequent life events experienced by CYP with WS relate to those that are interpersonal in nature (e.g., being insulted by peers, being bullied by someone considered to be a friend) and that these events are experienced at a higher rate compared to neurotypical CYP. These parental data reinforce the accounts given by adults with WS when reflecting on their peer interactions during the school years (Chapter 3). Some life events on the VEQ were not endorsed at all in the neurotypical group but were commonly experienced by CYP with WS, thereby highlighting experiences that may be particularly relevant to WS. One example of this is moving schools because the school was unable to cater for the young person's needs (35% of CYP in the WS group had experienced this) which is noteworthy when thinking about the disruption to social networks and having to build new friendships in the new school environment. Another relatively common experience was not seeing friends outside of school (39% endorsed in WS, 3% neurotypical) which chimes with recent reports of CYP with WS

experiencing social exclusion outside of school (i.e. not being invited to peers' homes and birthday parties; Gillooly et al., 2021, 2022).

The real-life examples given by parents add further support that vulnerability is inherently social in nature, with many examples relating to interpersonal pressures and negative experiences with peers (e.g., bullying). This is significant given reports that CYP with WS have a strong desire for social connection (Lough, Flynn, et al., 2016; Lough, Rodgers, et al., 2016). On the topic of peer relationships, recent studies have reported that CYP with WS had significantly greater friendship difficulties than a neurotypical sample, including difficulty sustaining friendships and social exclusion, which may not be identified by children with WS themselves (Gillooly et al., 2021, 2022). Furthermore, in Chapter 3, when asked about friendships, adults reported negative experiences particularly during the school years, and the examples given by parents in this Chapter also suggest that social adversity is common during adolescence.

Taking together the VEQ and qualitative evidence, one conclusion is that the real-life experience of vulnerability is very much social in nature. Of course, this could change with developmental stage as indicated from adult reports, which has highlighted vulnerability extending to areas such as finance (Lough & Fisher, 2016b). It is noteworthy that 50% of the sample reported a real-life example of social vulnerability, which is lower than reported in a sample of adults with WS (75% of parents reported an example in Lough & Fisher, 2016b). While 50% is not a low endorsement, it is likely the societal context in which the data were collected—reduced socialisation and lockdowns due to Covid-19—impacted the responses and, indeed, parents cited these factors when explaining their lack of example. It is likely that we would find a greater proportion of the sample providing an example of social vulnerability and future research should ascertain a more realistic account of everyday scenarios encountered by CYP with WS when not socially restricted. For others, not providing an example was linked to their child being always highly supervised, which links to the evidence that parents least endorsed independence-related vulnerability on the SVQ. Finally, the finding that the level of social vulnerability (on the SVQ) was correlated with the frequency of vulnerable experiences (on the VEQ), provides preliminary evidence that heightened social vulnerability is associated with adverse experiences in everyday life for CYP with WS. Further work is needed to understand how the SVQ and VEQ relate (i.e., overlapping items), but the addition of this measure in the current study provides new evidence to suggest that vulnerability is social in nature in WS, and it is impacting everyday social outcomes.

5.5.4 Family Impact and Protective Strategies

The final aim of the study was to examine for the first time the impact of social vulnerability and strategies to manage social vulnerability risk. Despite its exploratory nature, the findings offer some interesting insights which future research can build upon in hypothesis-driven enquiry. First on impact, most parents in the sample expressed high concern and many reported being particularly concerned in relation to their child getting older (the question did not prime about age). This finding adds context to the earlier finding of no age-related increases in parental independence. If parents are worried about their child being taken advantage of, and this is more likely to be the case if they encounter increasingly complex social situations with age, then it makes sense that parents may not grant increased independence as their child gets older.

Indeed, one of the key strategies to protect against social vulnerability reported by parents was to ensure high levels of supervision and/or reduce their son/daughter's opportunities for social interaction. This finding is consistent with interview data from parents of CYP with WS (Gillooly, 2018) highlighting parental support and supervision as a means to manage inappropriate social approach behaviour. Another strategy reported by parents in the current research was to educate their young person about social dynamics and risks, and the examples given relate to common core aspects of difficulty associated with the WS profile such as interpreting social cues (e.g., signs someone is being untruthful; social stories) and stranger danger awareness (Riby, Kirk, et al., 2014). An intriguing finding is that a smaller number of parents reported that they educate others about WS and vulnerability. This is interesting as so often we focus on the how to equip the individual with useful skills, however social vulnerability it is a two-way interaction – individuals are only vulnerable in the environment if there's an interactor who wants to take advantage, be manipulative or bully. The importance of educating others also emerged from Chapter 3 in relation to peers in the school environment not understanding different needs.

5.5.5 Considerations and Future Research

There are several methodological limitations and barriers to measuring social vulnerability that should be noted and considered when designing future studies on this topic. First, although the SVQ has shown evidence of good psychometric properties in adults with ID (Fisher et al., 2012; Fisher, Shivers, et al., 2020), it has not been validated for use with CYP with ID. While the questions were deemed appropriate in the current age group, some

subscales fell below acceptable levels and further work is required to establish whether the SVQ is a valid and reliable instrument for assessing social vulnerability in CYP with WS. Second, and linked to this, the SVQ was adapted in the current study to enable comparison between WS and neurotypical profiles. The adapted measure has not undergone psychometric testing and we found that many of the SVQ subscales showed unacceptable scale reliability. This could be a function of low covariances among items and/or the lack of items in some subscales. Thirdly, further consideration should be given to the theoretical assumptions that underpin the design of the SVQ. For instance, and as previously noted, the design of the Parental Independence subscale is such that greater scores (greater independence from parents) equate to greater overall social vulnerability, on the assumption that being afforded autonomy means there is a greater likelihood of a vulnerable outcome. Yet, is it the case that individuals with higher levels of parental supervision are less vulnerable? While parental protection likely makes an adverse outcome less likely to occur, it could also be true that being closely supervised and having less opportunity to encounter and learn from social experiences, could reinforce social vulnerability. If we are to make accurate assessments of social vulnerability, a better understanding of the different domains of vulnerability is required.

Overall, the reliance on the SVQ stems from the lack of instruments available to assess social vulnerability in people with IDD, particularly CYP. Indeed, a review of the literature identified five published tools, many of which have been designed to for use with clinical groups but may not be appropriate for use with CYP with WS. For example, the Test of Interpersonal Competence and Personal Vulnerability (TICPV; Wilson et al., 1996) was one of the first measures developed to assess social vulnerability in people with IDD. It is a self-report measure which requires adults with IDD to choose the behaviour they would likely adopt in a range of hypothetical, social situations. This self-reflection about likely decision-making can be difficult for people with WS and the items were deemed too sensitive to include in research with CYP with WS. Conversely, informant-report scales designed for childhood, such as the Children's Social Vulnerability Scale (Seward et al., 2018) was developed to measure social vulnerability in neurotypical CYP and therefore likely misses aspects of vulnerability that are specific to having an IDD. It also includes only seven items and does not allow for a detailed profile across different features of vulnerability. Therefore, it is true that WS research on this topic is restricted to a limited pool of instruments. Nevertheless, the approach used in the current study of cross-referencing different sources of

assessment, including quantitative scales and qualitative insights, is a strength and future research should devise tools which collect the voice of the person themselves, in a sensitive and meaningful way.

A further limitation worth considering is the possibility of sample selection bias and priming effects. The study was advertised as being about social skills and social vulnerability, therefore it may have appealed to parents and carers who were particularly concerned about their child's social interactions and social vulnerability. To challenge this, it would be useful to probe social vulnerability as part of a study that has a wider, multi-disciplinary focus and if framed more broadly (i.e. "experiences during adolescence in WS"). Another strategy, albeit logistically challenging given the rarity of WS, would be to obtain a more random sample of participants by conducting research through community settings such as GP appointments. In addition, the sequencing of the questions on social vulnerability may have implications for the pattern of results. For instance, the open-ended questions which asked parents to report on their concerns and provide examples of social vulnerability were placed immediately after the 30 SVQ Likert items which described specific features of social vulnerability. Respondents were likely prompted by these descriptions and ideas when responding about their concerns and experiences. Future survey research on this topic should consider placing broader questions that measure the overall experience (e.g. levels of concern) first, followed by more specific questions on the topic.

Despite the limitations, the study certainly adds to our understanding of the profile of social vulnerability in childhood and has identified important avenues for future enquiry. Having established in this study (and consistent with Lough, 2016) that levels of social vulnerability are elevated in CYP with WS at a level considerably higher than neurotypical CYP, future work should move beyond making WS/neurotypical comparisons and focus efforts into understanding the issue specifically in WS. If the goal is to support families of CYP with WS, then much more research is needed with this group. The qualitative insights from caregivers in this study have taken us a step further in recognising the impact of social vulnerability and the strategies already adopted by families. However, to provide tailored support we first need to understand the factors that contribute to social vulnerability in WS (both risk and protective factors) and the ways in which these high levels impact on CYP and the wider family.

The topic of independence and its association with vulnerability has emerged throughout the study in that (i) independence-related vulnerability was the least endorsed subscale in the WS group, (ii) CYP with WS were reported to have less parental independence compared to neurotypical CYP, (iii) no evidence of age-related increases in independence in WS unlike in the neurotypical sample and (iv) caregivers reported high levels of supervision as a strategy to manage concerns. This combination of findings raises preliminary evidence of an interplay between social vulnerability and independence, whereby independence is reduced as a protective strategy and one which could be usefully explored in further research. Considerably more work is needed on trajectories of independence during the childhood and adolescence years. While researchers have studied adaptive behaviour (which relates to independence) in childhood (see Brawn & Porter, 2018), less is known about independence, supervision and protection, or how having less independence in adolescence impacts on later outcomes in adulthood. Consideration should be given to how best to measure independence in CYP with WS as only three items of the SVQ contribute to this subscale and are arguably ambiguous. For instance, "parents allow to be with older individuals with no supervision" could be interpreted as unsupervised interactions with grandparents or other members of the family, which is very different to unsupervised interactions with acquaintances or unfamiliar adults. Therefore, to develop a richer picture of the interplay here, studies should consider alternative, more comprehensive assessments of independence/supervision. As a starting point, qualitative research with parents and carers of people with WS may be helpful in identifying what independence looks like and the barriers to gaining independence.

Many of the findings emerging from this study, including those on parental independence, raise important clinical implications. With vulnerability high and real-life negative outcomes common in this group, there is a need to raise awareness of social vulnerability and devise support strategies appropriate for this developmental stage. A review of the literature on victimisation and social vulnerability identified a lack of intervention studies on this topic in the context of people with ID (Fisher et al., 2016). In the context of WS, this is limited to one study teaching adults about interacting with strangers (Fisher, 2014; Fisher, Burke, et al., 2013). In addition, the current study clearly demonstrates that support is needed at the family-level too. Parents report being very concerned about their child's social vulnerability as they get older, meaning that support is needed to help families navigate the transition from adolescence to adulthood. The strategies outlined by parents give a good

insight into where support is needed. For instance, high levels of parent supervision are a well-intentioned support strategy to protect CYP from harm, however this likely reduces opportunities for independence, which in turn could have wider implications for skill development (potentially feeding back into social vulnerability). We need to provide support to parents/caregivers so that they can take manageable steps towards granting opportunities for independence.

In conclusion, this study set out to address the lack of evidence on the profile and nature of social vulnerability in adolescence in WS. The findings emphasise that social vulnerability is present at elevated levels during this key developmental period and requires support at the individual and family level. There appears to be no clearcut relationship between social vulnerability and age, but further work is needed to disentangle how social vulnerability presents at different developmental stages and in relation to cognitive capacity and emotional awareness. Despite its exploratory nature, the study offers insight into the impact of social vulnerability and strategies adopted by families to manage social risk. The relevance of independence is clearly supported by the results and requires further exploration, both in terms of measurement and in thinking about the nature of vulnerability and support. Having outlined the profile of vulnerability, the next Chapter examines the association between social vulnerability and other core aspects of the WS social, cognitive and behavioural profile, in the same sample of CYP with WS.

6. CHAPTER 6: SOCIAL VULNERABILITY IN WILLIAMS SYNDROME: THE ROLE OF INDIVIDUAL AND FAMILY FACTORS

6.1 Chapter Overview

Chapter 5 examined the profile and nature of social vulnerability in a group of CYP with WS. A clear finding from the perspective of parents was that CYP with WS experience high levels of social vulnerability not evident in a neurotypical comparison group. Such heightened vulnerability was a significant concern for most parents/caregivers in the WS sample. To understand how best to support families concerned about their child's social vulnerability and help support CYP with WS with healthy social interactions, we first need to clarify the factors that contribute to heightened social vulnerability. The next stage of inquiry therefore extends the findings of Chapter 5 to investigate several relevant but untested personal and family factors that could impact on social vulnerability in WS. In this study, the individual factors of interest were those that have been identified as important in the social vulnerability literature more broadly and/or are common features of WS known to be associated with the social profile. This study is also the first to investigate the relationship between social vulnerability and external characteristics about CYP's family environment. In Chapter 5, parents endorsed a lack of parental independence and the qualitative findings highlighted high levels of supervision as a strategy to protect against high social vulnerability. This study builds on these findings by examining parent protection behaviours during childhood and adolescence in WS, and considers the relationship with social vulnerability. The data reported in this Chapter come from the same sample of CYP as described in Chapter 5.

6.2 Introduction

The significance of social vulnerability has been emphasised in the WS literature and studies have started to delineate profiles of social vulnerability in adulthood (Fisher, Moskowitz, et al., 2013; Jawaid et al., 2012; Riby et al., 2017). Findings from Chapter 5 provided important insights about the presence and nature of social vulnerability earlier on in development than has previously been studied in WS, indicating that social vulnerability is elevated and requires support during adolescence. However, there currently exists little systematic research to establish why social vulnerability is elevated in WS. This evidence-gathering is a crucial first step towards knowing how best to support CYP with WS develop healthy social interactions and support the wider family network for whom social

vulnerability is a real concern (as emphasised in the findings of Chapter 5). To better understand the factors that contribute to elevated social vulnerability in CYP with WS, the current study aimed to examine how social vulnerability relates to individual and family characteristics.

The literature reviewed in Chapter 1 demonstrated that WS is associated with cognitive, behavioural and emotional features which have significance in the context of social vulnerability, but their association has not been fully explored. So, what does the evidence to date suggest about the individual factors of importance? Studies using the Social Vulnerability Questionnaire (SVQ; Fisher et al., 2012; Fisher, Moskowitz, et al., 2013) have started to identify domains in which people with WS may be particularly vulnerable. In adults, the evidence suggests that parental independence and physical characteristics play a role (Fisher, Moskowitz, et al., 2013). The findings from the SVQ subscales reported in Chapter 5 also provide insight into potentially relevant factors that contribute to social vulnerability in CYP. The profile indicated that CYP with WS are particularly vulnerable in areas relating to credulity, which refers to the tendency to believe information even in the face of competing information. High credulity may be linked to difficulties making socio cognitive evaluations which has been well documented in WS. For instance, challenges in making appropriate judgements of trustworthiness (Martens et al., 2012 and also relevant in Chapter 3) and a lack of stranger danger awareness (Riby, Kirk, et al., 2014). In Chapter 4 it was also reported that different styles of social interaction relate to higher social vulnerability and may contribute beyond the presence of ND, although to a small extent.

To date, only a few studies have examined factors underlying social vulnerability in WS. Therefore, in theorising on the factors that might be relevant, a valuable source of information comes from theoretical models of social vulnerability and studies with other neurodevelopmental and clinical groups reported to experience high social vulnerability (Greenspan et al., 2001; Pinsker et al., 2010; Pinsker & McFarland, 2010; Sofronoff et al., 2011). Some of the factors identified as important from this work are transdiagnostic issues and commonly reported in people with WS, such as executive difficulties and social cognition. Thus, drawing upon the broader social vulnerability literature can guide hypotheses in how features of the WS profile might relate to social vulnerability (Thurman & Fisher, 2015). The introduction of this Chapter draws upon both the WS literature and wider neurodevelopmental and social vulnerability literature, to set out the rationale for studying several individual factors which may place CYP with WS at increased risk of social

vulnerability. The second half of the introduction argues the importance of studying social vulnerability through a wider lens about the lives of CYP with WS.

6.2.1 The Role of Individual Characteristics

Cognition and executive functions (EF). Taking cognition first, many people with WS have mild-moderate ID (Searcy et al., 2004) and the presence of an intellectual disability (ID) has been identified as a clear risk factor for experiencing issues related to social vulnerability such as interpersonal violence (Hughes et al., 2012; Jones et al., 2012; Sullivan & Knutson, 2000). However, the findings from Chapter 4 (Paper 2) indicate that ID may not entirely explain elevated social vulnerability, given we found evidence of heightened levels across neurodevelopmental groups not typically associated with ID (ADHD and autism). Additionally, in a study of adults with WS, Lough et al. (2016b) found no meaningful relationship between intellectual ability, as measured by the Kaufman Brief Intelligence Test-2nd Edition (K-BIT-2; Kaufman and Kaufman, 2004) and total score on the SVQ or any of the SVQ subscales, lending support that social vulnerability may be elevated across the spectrum of intellectual ability in WS. Therefore, it appears that social vulnerability may be associated with characteristics beyond ID. To date, no studies have investigated how different facets of cognition may link to social vulnerability in WS.

A specific component of cognition known to be challenging in WS and has been identified in models of social vulnerability is executive control or executive functions (EFs). EFs refer to a suite of cognitive processes that contribute to goal-directed behaviour such as planning, organisation and problem solving (Diamond, 2013). While there has been no research investigating the EF correlates of social vulnerability in WS, evidence from related research suggests that EFs play a role. For example, EFs are associated with peer relationships and victimisation in neurotypical development (Holmes et al., 2016; Verlinden et al., 2014). Additionally, research with groups considered to experience high social vulnerability, such as older adults (Pinsker & McFarland, 2010) and autistic adolescents (Kloosterman et al., 2014) has found executive dysfunction to be associated with greater social vulnerability.

As outlined in Chapter 1, many people with WS have specific neuropsychological challenges in areas of EF (Camp et al., 2016; Capitão et al., 2011; Greiner de Magalhães et al., 2022; Hocking et al., 2015; Ng-Cordell et al., 2018). A recent study found that EF difficulties were highly prevalent in 308 6–17-year-olds with WS (Greiner de Magalhães et

al., 2022). Research has established that EF, particularly the component of inhibition, is linked to the disinhibited social behaviours that characterise the WS social profile. In a study of children with WS (6-15 years) Little et al. (2013) explored the relation between social approach behaviours using Adolph's Approachability Task (Adolph et al., 1998) and response inhibition, using the Sun-Moon Stroop task (Archibald & Kerns, 1999). Taking a cluster analysis approach, they identified behavioural subgroups based on levels of response inhibition and found that those with high approachability behaviours were also those with the greatest inhibition difficulties. The opposite pattern was found for those with the lowest approachability behaviours. Ng-Cordell et al. (2018) provided further evidence of the relationship between EF and the social profile in a study of individuals with WS. Using a validated parent questionnaire of EF, The Behaviour Rating Inventory For Executive Function (BRIEF) (Gioia et al., 2000a), the authors reported a strong, positive relationship between EF and social reciprocity difficulties. Evidence such as this has been used in support of the frontal lobe hypothesis of hyper sociability (Porter et al., 2007), which claims that the propensity to approach others seen in WS stems from impairments in inhibitory control, whereby individuals with WS may 'know' that approaching others is not appropriate but struggle to stop the behaviour. Indeed, researchers have drawn parallels between the disinhibited social behaviour seen in WS and those documented in individuals with acquired frontal lobe damage (Porter et al., 2007).

Taken together, the evidence of an association between EF/social challenges in WS and the findings from the broader social vulnerability literature, provide a compelling rationale for studying EFs in the context of elevated social vulnerability in CYP with WS. To date, there has been a focus on inhibition and its relation to the disinhibited social behaviours that typify the WS social phenotype (e.g. Little et al., 2013), however EF is not a unitary construct and it will be important to establish how different components of EF relate to the profile of social vulnerability. Indeed, previous research with CYP with WS using the BRIEF has found an uneven profile across the EF indexes, with relative strengths in behaviour and emotional regulation, alongside relative challenges in cognitive regulation (Greiner de Magalhães et al., 2022; Hocking et al., 2015). Studies also suggest that aspects of EFs are differently related to other aspects of functioning. For instance, behavioural and/or emotion regulation, but not cognitive regulation, has been associated with attention difficulties (Hocking et al., 2015) and anxiety (Woodruff-Borden et al., 2010). The BRIEF is a common rating instrument of EF which examines the EF domains of behavioural, cognitive and

emotional regulation. Theoretically, all three of the BRIEF domains could be related to social vulnerability. CYP with WS who find it difficult to inhibit behavioural impulses (e.g., approaching others) may lead to participation in situations that increase the likelihood of a vulnerable outcome or may impact on the flow and experience of the interaction from the perspective of the partner. Difficulties with cognitive regulation may make it difficult to switch attention from one situation to the next, problem-solve and initiate a course of action if presented with a threatening interaction.

Adaptive behaviour. Adaptive behaviour refers to a set of everyday skills and behaviours that help individuals to function in their everyday lives (Adaptive Behavior (aaidd.org)). An assessment of adaptive behaviour is valuable for identifying the level of support an individual might benefit from in order to engage and be safe in areas such as community participation, school and learning, and employment (Brawn & Porter, 2014). WS is associated with challenges in adaptive functioning which may increase (or plateau) with age in adolescence and into adulthood (Fisch et al., 2007; Fisher, Lense, et al., 2016b; Mervis & John, 2010; Mervis & Klein-Tasman, 2000). Studies that have examined the profile across different dimensions of adaptive behaviour have reported that adolescents and adults with WS usually show greater abilities in the domains of socialisation and communication (Elison et al., 2010; Fisch et al., 2007) but have greater challenges in daily living skills (Fisch et al., 2007; Greer et al., 1997; Hahn et al., 2014). However, like many behavioural features of WS, there is heterogeneity in adaptive behaviour, with some individuals scoring very low in adaptive functioning, and others scoring at a level commensurate with their chronological age (Brawn & Porter, 2014).

There is some evidence that adaptive behaviour relates to social vulnerability. A study of 29 adults with ID found that individuals with greater support needs in daily activities were more socially vulnerable (Tabin et al. 2020). A similar finding was reported in a study of 102 adults with WS whereby Lough et al. (2016) found evidence of an association between social vulnerability and functional ability as measured by the Activities of Daily Living (Seltzer & Li, 1996), which is used as an estimate of functional abilities of individuals with IDD (e.g., ability to read and participate in leisure activities). Individuals reported to have greater functional independence were also rated as less socially vulnerable, as per total score on the SVQ and all SVQ subscales, apart from the subscale emotional abuse. This finding suggests that greater functional abilities are associated with lower levels of social vulnerability overall in adults with WS but may not protect against vulnerability to emotional abuse. To date, we

have no evidence as to whether this relationship exists in childhood in WS and whether we see associations between different aspects of functional skills (e.g., socialisation, communication, daily living). If, for example, we find evidence that greater daily living skills are a protective factor in social vulnerability outcomes, then this provides a potential avenue for supporting concerns (i.e., by teaching functional skills).

Social competencies. According to theoretical frameworks of social vulnerability, broader aspects of social behaviour, such as social cognition and social communication, play an important role (Greenspan et al., 2001; Pinsker et al., 2010). This is intuitive – social interactions are dynamic and navigating these involves paying attention and interpreting social cues, identifying truthful versus deceitful behaviour, and understanding social exchanges. Empirical studies with clinical populations known to have socio-cognitive difficulties provide support for this hypothesis. For example, older adults rated as more self-aware using the Patient Competency Rating Scale (Prigatano, 1986) and with greater social skills, as measured by the Social Performance Survey (Lowe & Cautela, 1978), were rated as being less socially vulnerable (Pinsker & McFarland, 2010). Similarly, studies with autistic CYP have reported those with high social vulnerability to have poorer social skills (Sofronoff et al., 2011) and those who experienced victimisation were 5 times more likely to have greater communication difficulties than those who had not (Cappadocia et al., 2012).

There is a substantial WS literature documenting social differences in childhood and adolescence. We know from Chapter 1 that this includes differences across all levels of the social phenotype, including social attention, social motivation, and social communication (Thurman & Fisher, 2015). We also learnt from Chapter 4 that CYP with WS show differences in social interaction styles and the quality of everyday eye contact, and found first evidence that different styles of interaction contribute to social vulnerability. Despite the salience of the social features of WS, we know very little about how social vulnerability relates to other aspects of the WS social profile. Lough (2016) is the first and only study to have examined social vulnerability and its relation to social behaviour and social communication in CYP with WS. The findings indicated a strong association ($r = .75$) between greater social vulnerability and poorer social reciprocity, as measured by the Social Responsiveness Scale (SRS) (Constantino & Gruber, 2012). In contrast, there was no meaningful relationship between social vulnerability and social communication, as measured by the Social Communication Questionnaire. The study provides preliminary evidence of association between social vulnerability and some features of social behaviour in WS.

Furthermore, we know from the profile of social vulnerability reported in Chapter 5 that parents highly endorsed vulnerability related to credulity.

Anxiety. The role of emotional behaviour in social vulnerability is far from understood, but related research has identified it to be of interest. For example, a separate but related body of research on peer victimisation has highlighted an association with internalising psychopathology – meta analyses of longitudinal studies found a bi-directional relationship whereby internalising problems predicted increased likelihood of experiencing peer victimisation and, equally, peer victimisation predicted increased in internalising problems (Christina et al., 2021; Reijntjes et al., 2010). The role of emotional behaviour is further supported by a study of social vulnerability in autistic CYP which found higher levels of social vulnerability to be associated with higher levels of anxiety, anger, and behavioural problems (Sofronoff et al., 2011).

For many people with WS, anxiety is a common and significant mental health challenge, often developing at a young age and persisting into adulthood (Leyfer et al., 2006; Ng-Cordell et al., 2018; Royston et al., 2017; Woodruff-Borden et al., 2010). While little is known about the relationship between anxiety and social vulnerability, studies have reported a link between heightened anxiety and other aspects of the social profile in WS. For instance, in a sample of 59 individuals with WS, Riby et al. (2014; mean age 17 years old, 6-36) reported that those with higher levels of anxiety (as measured by the Spence Children's Anxiety Scale-Parent version) also showed greater impairments in social reciprocity (as measured by the Social Responsiveness Scale). The interplay between anxiety and social functioning was further borne out when the sample was split in two groups high/low anxiety, showing that the group with high anxiety had greater difficulty in all social areas measured by the SRS, except for social motivation where the groups did not differ. These findings indicate that an association between greater anxiety and more social difficulties.

To the author's knowledge, only one study to date has investigated the link between anxiety and social vulnerability in CYP with WS. Lough (2016) reported a moderately strong association ($r = 0.56$) between level of social vulnerability, measured by the SVQ (Fisher et al., 2012), and severity of anxiety, measured by the Spence Children's Anxiety Scale (Spence, 1998). The relationship was further supported when anxiety levels were analysed between those with high/low social vulnerability (based on upper and lower quartiles on the SVQ), showing that CYP with high social vulnerability experienced significantly greater

levels of anxiety. However, results from a partial mediation analysis indicated that the association between anxiety and social vulnerability was driven by social reciprocity (as measured by the SRS-2). In other words, high anxiety in of itself did not account for social vulnerability. These findings are valuable in starting to think about how social vulnerability and anxiety may relate, however the analyses require caution given the high/low social vulnerability analysis was based on 6 participants per group. The current study builds upon these initial findings, to clarify the association between social vulnerability and anxiety, and to understand if anxiety is related to social vulnerability, then to what extent does it contribute.

To summarise so far, difficulties in EFs, adaptive functioning skills and social reciprocity, as well as heightened anxiety, are common features of the WS profile and show associations with aspects of the WS social phenotype. Additionally, these individual characteristics have been identified to be important in the wider social vulnerability literature with other ID and/or clinical groups. Such evidence provides a useful starting point in theorising on how individual factors might relate to social vulnerability in WS, however it is crucial that research examines these relationships specifically in WS as pathways to social vulnerability may be different in different neurodevelopmental groups (Fisher, Moskowitz, et al., 2013). In Chapter 4 all neurodevelopmental groups studied showed heightened social vulnerability, but we see differences in the nature of aspects of social behaviour. To date, few studies have examined how individual factors relate to social vulnerability in people with WS and it is currently unknown whether and, to what extent, the cognitive and behavioural features outlined above relate to the elevated levels of social vulnerability in CYP with WS.

6.2.2 The Role of Family Characteristics

Thus far, the Chapter has focused on features of cognition, behaviour and emotion where people with WS commonly experience difficulties, and which may make social vulnerability more likely compared to peers without WS. However, as discussed in Chapter 1, social vulnerability is not intrinsic nor is it fixed; personal factors interact with the external environmental and contextual factors (Greenspan et al. 2001; Luna, 2009; Luna & Vanderpoel, 2013). Therefore, the personal factors outlined above are important to examine in relation to social vulnerability but should not be considered in isolation. In general, the role of environmental factors and the wider social context has been overlooked in studies of social behaviour in WS. In a study of adaptive behaviour in children and adults with WS, Brawn

and Porter (2014) considered characteristics about the family environment (e.g. quality of relationships, emphasis on personal growth and degree of control and organisation in family life). Greater adaptive behaviour skills overall and in the domains of socialisation and daily living skills were associated with family engagement in more recreational and social activities outside of home/work, and intellectual and cultural activities. As the authors highlight, it could be that participation in these activities provides learning opportunities for individuals with WS to develop further skill in areas of socialisation and daily living. It could also be that families of individuals with WS who have stronger abilities in these domains are more able to participate in activities in the community. Interestingly, families of individuals with higher levels of externalising behaviours (e.g., problems with inhibition and impulsivity) reported lower scores on the Independence scale which measured the family's focus on self-sufficiency and making independent decisions. Again, we have no information of causality in these associations, but the authors speculate that difficulties in areas such as inhibition may make it more challenging for families to encourage autonomy in individuals with WS.

Characteristics about the family environment, particularly around issues of independence and control, is also relevant to consider in the context of social vulnerability. Lough et al. (2016) explored levels of social vulnerability between adults with WS who lived at home compared to those living away from home, in addition to adults in employment and those who did not work. In both cases, there was no meaningful difference in levels of social vulnerability, indicating social vulnerability may extend across levels of independence. However, the authors noted that individuals currently in employment had variable experiences of this—from 1 to 40 hours per week—therefore the conclusion that social vulnerability is not related to levels of independence is tentative. In Chapter 5 of the thesis, parents reported high levels of supervision over their young person's social interactions and endorsed a lack of parental independence, which did not increase with age. Typically, parents of young children exert a high level of control over the child's exposure to and engagement in social situations which tends to gradually decrease as children get older. In families of CYP with WS, the trajectory of autonomy may look different. Close supervision and restricted opportunities for independence may be a well-intentioned strategy to manage social safety concerns. It is also interesting to consider whether family protection behaviours feed back into social vulnerability. Tabin et al. (2021, p. 2) states that "any individual, including a person with ID, learns to independently navigate potential challenges and thus decrease their social vulnerability through exposure to social situations". Therefore, a greater dependence

on others may reduce opportunities to encounter and learn from social interactions, which in turn may lead to less proficiency navigating interactions and more reliance on others for decision making (Lough & Fisher, 2016b).

A closely related concept, parental overprotection, refers to a parent's protective behaviours that may be deemed excessive in view of the child's developmental level. Parental overprotection has been linked to child-related factors which may relate to parents' perceptions of child vulnerability, including chronic pain (Anno et al., 2015), paediatric conditions (Hullman et al., 2010) and anxiety disorders (Hudson & Rapee, 2001). Indeed, parents of adolescents with disabilities report higher overparenting behaviours than parents of children without disabilities (Gagnon et al., 2020). With these findings in mind, parental protection is likely to be relevant in the context of CYP reported to be vulnerable in the social domain coupled with the presence of ID. Building on the findings of high levels of supervision reported in Chapter 5, the interest here is to further examine parent protection behaviours in this age group and the potential association with social vulnerability, in recognition of wider family and environmental factors related to social vulnerability.

6.2.3 Research Goals and Predictions

In light of the above, the primary objective of this study was to examine whether, in a sample of CYP with WS, social vulnerability was related to individual- and family-level characteristics and if so, outline the nature of this relationship. The individual-level factors chosen for focus in this study were common co-occurring features of WS, namely EFs, adaptive behaviour, anxiety, and social reciprocity. The study also sought to address the absence of evidence about the relation between family characteristics and social vulnerability. Therefore, a second aim was to provide the first evidence on parental protection behaviours and how this relates to social vulnerability. It is hoped that this study—the first to examine all these aspects within the same study in WS—will contribute to a deeper understanding of the factors related to high social vulnerability. The research aims and associated hypotheses were:

1. To investigate the association between social vulnerability and individual-level factors of EF skills, anxiety, social reciprocity differences and adaptive behaviour.
    a. **EF predictions:** Based upon the evidence for the role of EF in the WS social profile (Little et al., 2013; Ng-Cordell et al., 2018; Porter et al., 2007), it was hypothesised that greater difficulties in this domain would be positively

correlated with more social vulnerability. We also explored whether specific domains of EF (cognitive, behavioural and emotional regulation) are differentially related with social vulnerability. Given inhibitory control is known to relate to WS social behaviour, a specific prediction was made about behavioural EF (includes components inhibition) relating to social vulnerability, but no other predictions were made about the two other components.

   b. **Adaptive behaviour:** based on evidence from adults with WS (Lough & Fisher, 2016b), greater adaptive behaviour was predicted to be correlated with lower social vulnerability
   c. **Social functioning:** Given the findings of Lough (2016) and the wider literature on social skills feeding into social vulnerability, social vulnerability was predicted to be positively correlated with social reciprocity difficulties (so more social vulnerability related to great social reciprocity difficulty).
   d. **Anxiety:** Leading from the work of Lough (2016) and knowing that an interplay has been found between anxiety and broader features of the WS social profile (Riby et al. 2014) it was expected that greater levels of anxiety would be related to greater social vulnerability.

2. To identify predictors of social vulnerability using a regression model. As no studies to date have considered all of the individual characteristics above in relation to social vulnerability, this was an exploratory aim and no specific hypotheses were set.
3. To document the profile of parent protection behaviours and examine the relationship with social vulnerability. Even though this was the first study to explore this question, it was hypothesised, based on the initial evidence in Chapter 5, that parents of CYP with WS would endorse greater protection behaviours compared to parents of neurotypical CYP. It was also expected that higher levels of social vulnerability would be associated with greater parent protection behaviours.

6.3 Method

6.3.1 Participants

The study included the same sample of parents of CYP diagnosed with WS (aged 10-17 years) reported in Chapter 5. In relation to Aim 3, data is also included from the neurotypical sample described in Chapter 5.

6.3.2 Materials

Parents/caregivers completed a battery of questionnaire measures that asked them to report on their young person's social reciprocity, executive functions, anxiety symptoms, and adaptive behaviour. The study also used data on the young person's social vulnerability and demographic information, which were outlined in Chapter 5 and are not repeated here. As outlined in Chapter 2, the decision to take a questionnaire design to this study was in response to the restrictions on face-to-face data collection because of the pandemic.

Everyday executive functions. The parent version of the Behaviour Rating Inventory of Executive Functioning – Second Version (BRIEF-2; Gioia et al., 2015) was administered to measure the child's EFs in the home setting. The BRIEF-2 is an updated version of the original BRIEF questionnaire measure (Gioia et al., 2000a, 2000b); an informant-report ratings scale which was designed to assess "real-world" expressions of EF behaviours (including many items related to EF within social settings). The BRIEF-2 is a widely used as tool for the assessment of EFs in children with neurodevelopmental conditions, for example ADHD and autism and WS (Gioia et al., 2002; Hovik et al., 2017). The parent-report version used in the current study is validated for CYP aged 5-18 years. The BRIEF-2 includes 63-items which map onto three index scales: Behaviour Regulation Index (BRI), Emotion Regulation Index (ERI) and Cognitive Regulation Index (CRI). The BRI represents a child's ability to regulate and monitor behaviour effectively and is comprised of the subscales Inhibit (8 items) and Self-Monitor (4 items). The ERI represents a child's ability to monitor and regulate emotional responses and comprises of the subscales Shift (8 items) and Emotional Control (8 items). The final index, CRI, represents a child's ability to control and manage cognitive processes and problem solve effectively, and is comprised of the subscales Initiate (5 items), Working Memory (8 items), Plan/Organise (8 items), Task-Monitor (5 items), and Organisation of Materials (6 items). An overall summary score, the Global Executive Composite (GEC), is generated by summing the three index scales.

Using the BRIEF-2, parents rate their child's behaviour using a three-point Likert scale (never, sometimes, often). Higher scores indicate greater executive dysfunction. All scales and indices use T-scores to generate clinical cut-offs. T-scores exceeding 64 are considered clinically significant, 60-64 are classed as *mildly elevated*, 65-69 classed as *potentially clinically elevated*, and 70 or above is classed as *clinically elevated*. The BRIEF-2 parent form was validated on a sample of 1,400 parents of CYP aged 5-18 years, and shows

good correlations with other measures of IQ, supporting its validity. The BRIEF-2 shows good psychometric properties: internal consistency for the subscales is good (ranging 0.76-0.97) and good test-retest reliability of above 0.8. The BRIEF-2 has shown good discriminant validity by distinguishing between clinical and non-clinical groups (Jacobson et al., 2020) (e.g. Gioia et al., 2000; McCandles & O'Laughlin, 2007). In the current study, the measure as a whole (GEC) showed excellent internal consistency (a = .93) and the individual scales ranged from questionable at .63 (task-monitor) to good at .87 (emotional control).

Adaptive behaviour skills. The Vineland Adaptive Behaviour Scales–Third Edition (VABS-3; Sparrow et al., 2016) is an assessment of an individual's ability to cope with environmental changes, learn everyday skills and be independent. It is widely used in research with individuals with IDD and is the most commonly used tool of adaptive behaviour in WS studies (Brawn & Porter, 2018). The VABS-3 assesses adaptive behaviour in the domains of communication, daily living and socialisation, which correspond to the three broad domains of adaptive functioning specified by the Diagnostic and Statistical Manual of Mental Disorders-Fifth Edition (DSM-5) and the American Association on Intellectual and Developmental Disabilities (AAIDD), and together they generate the Adaptive Behaviour Composite (ABC). The measure uses a 3-point scale to indicate whether an individual engages in a behaviour never (0), sometimes (1) or usually/often (2) without help or reminders. Higher raw scores indicate greater adaptive behaviour skills. The VABS-3 can be administered using a choice of three forms: parent interview, parent-report form and teacher-report. All three delivery formats are also available in Comprehensive version and Domain-Level version. In this study, the parent/caregiver, domain-level form (for individuals aged 3-90 years) was used to provide a relatively quick overview of adaptive behaviour, including the subscales Communication (40 items), Daily Living Skills (40 items) and Socialisation (40 items). Parents also completed the Maladaptive Behaviour Domain (35 items), but the results are not reported in this study. For the current sample, the measure showed excellent internal consistency for the full measure (a = .97) and for the three subscales (α ranged from .93 to .95). Raw scores can be converted to standard scores (M =100, SD = 15).

Social reciprocity. The Social Responsiveness Scale – Second Edition (SRS-2; Constantino & Gruber, 2012) School-Age form was used to gather information about social reciprocity. The SRS-2 is a 65-item, standardised parent-report measure for CYP aged 4-18 years, designed to assess impairments of social reciprocity in areas of communication,

interpersonal relationships and motivation. While the SRS-2 has been specifically developed to capture social impairments characteristic of autism, it also a widely used measure for capturing the social profile of other neurodevelopmental groups known to show differences in social behaviour, albeit that might fall at sub-threshold levels, including WS (Fisher & Morin, 2017; Glod et al., 2020; A. Järvinen et al., 2015; Klein-Tasman et al., 2011; Lai et al., 2021; Lough, Hanley, et al., 2015; Riby, Hanley, et al., 2014; Van der Fluit et al., 2012). Parents/caregivers respond to the items using a 1 (not true) to 4 (almost always true) Likert scale based on the frequency of the behaviour. The SRS-2 generates a total score in addition to scores on five subscales: Social Awareness, Social Cognition, Social Communication, Social Motivation, and Restricted and Repetitive Behaviour (RRB). Higher scores are indicative of greater difficulty in that domain of social reciprocity. Total and subscale scores can be converted into T-scores with the accepted clinical cuts as follows: scores <59 indicate no clinically significant concerns; scores between 60 and 65 indicate 'mild' social impairment, scores of between 66 and 75 indicate moderate levels of social impairment; and scores of 76 or higher are considered 'severe' clinically significant social impairment. In the current analysis T-scores were used to classify the sample into levels of functioning. All other analyses with the SRS-2 data used the raw scores to maximise variability, in line with other WS studies (e.g. Riby et al., 2014; Ng-Cordell, 2017). For the current sample, the total measure showed excellent internal consistency (a = .92), but subscales varied: social awareness (.49), social cognition (.77), social communication (.82), social motivation (.71), RRB (.77).

Anxiety. The Anxiety Scale for Children with Autism Spectrum Disorder – parent version (ASC-ASD-P; Rodgers et al., 2016; freely available at https://research.ncl.ac.uk/neurodisability/leafletsandmeasures/anxietyscaleforchildren-asd/asc-asdparent/) was designed to capture anxiety symptoms in autistic children. It was developed with knowledge of the Revised Child Anxiety and Depression Scale (RCADS; Chorpita et al., 2000) to document both the typical and autism-specific anxiety symptomatology. The measure consists of 24 items (12 from RCADS, 5 modified from RCADS and 7 ASD-related anxiety items) rated on a 4-point Likert Scale ranging from 0 (never) to 3 (always). Items are grouped into four subscales: Separation Anxiety (5 items), Uncertainty (8 items), Performance Anxiety (5 items) and Anxiety Arousal (6 items). The measure is not a diagnostic instrument, but a tool to describe the profile of anxiety symptoms and the authors of the ASC-ASD have suggested two scores indicative of elevated anxiety:

scores of ≥20 indicate “significant anxious symptoms” and scores of >24 is “a more specific indication of significant anxiety”. Psychometric evaluation of the ASC-ASD with 170 autistic children (8-16 years old) and their caregivers showed good convergent validity, strong test-retest reliability, excellent internal consistency of total and subscale scores for the parent version (a = 0.94) (Rodgers et al. 2016).

While the ASC-ASD has not been psychometrically evaluated to capture anxiety in children with WS, it is used in this study on the basis that anxiety likely presents differently in WS compared to NT as we know from the autism literature, and one other study has used the ASC-ASD on sample of children with WS (Glod et al., 2019). In the current study, a total score of ≥ 20 was used in support of elevated anxiety. For the current sample, Cronbach’s alpha indicated that internal consistency of the ASC-ASD-P subscales ranged from good (Separation Anxiety *a* = .84; Anxious Arousal *a* = .86; Performance Anxiety *a* = .82) to excellent (Uncertainty a = .92), and the measure as a whole showed excellent internal consistency (*a* = .94).

Parental protection. Assessment of parental protection was made using the Parental Overprotection (OP) measure (Edwards et al., 2008, 2010). This is a 19-item parent report questionnaire developed for use with parents of preschool children to study parenting behaviours that may limit a child’s exposure to situations perceived to be potentially dangerous or harmful for the child. Parents rate statements (e.g., “*I protect my child from criticism*”) based on how much each behaviour applies to their usual parenting practices using a 5-point Likert Scale from 0 (not at all) to 4 (very much). Item scores are summed to produce a total score, with higher total scores indicative of greater protection behaviours. The scale has shown high internal consistency (0.87), strong test-re-test reliability, good construct validity and to correlate with observations of maternal behaviour with a sample of parents of 3–5-year-olds (Edwards, 2010). While the scale was developed for use with parents of preschool children, it has since shown high levels of internal consistency in research with older children aged 7-12 years with and without a clinical diagnosis of anxiety (Clarke et al., 2013). The OP measure was identified as a relevant tool by which to provide an insight into parenting behaviours and level of supervision in parents of young people with WS. For the current sample, the OP measure had a Cronbach’s alpha of .88, indicating good internal consistency and mirroring other studies using this measure (e.g. Clarke et al., 2013; Edwards et al., 2010). Reliability analysis indicated that the internal consistency would be improved by

deleting 4 items, however the improvement was so minor, it was decided that all items would be retained in the analysis to allow for comparison with previous literature.

6.3.3 Procedure

The current data collection was part of a larger multi-phase study on social vulnerability in CYP with WS. The procedure is reported in the Method section of Chapter 5.

6.3.4 Analytic Approach

Raw scores obtained from the BRIEF-2, VABS-3 and SRS-2 were adjusted for chronological age to produce standard scores, according to the instrument manuals. As per BRIEF-2 scoring protocol, scale raw scores were not computed if more than one than one item that contributes to a subscale was missing response. One participant had four missing responses on the plan/organisation subscale and was not computed. On inspection of the ASC-ASD-P data, two outliers were identified but were not removed as we wanted to capture the full spectrum of anxiety. All variables were first explored to assess distribution. Before running analyses to answer the research questions, analyses were conducted to describe the profile across the individual factors of interest, and examine potential associations with the demographic variables, age, and gender. For two participants, their BRIEF-2 subscale score (inhibit =1, shift =1) was so high (EF so poor) that it was beyond the reference table in the BRIEF manual, therefore the highest raw score from the table was used. The OP measure is not standardised; therefore, the NT profile on this measure is outlined to provide contextual information about parent protection behaviours in WS.

To examine the relation between social vulnerability and EF, adaptive behaviour, social reciprocity, and anxiety (aim 1), correlational analyses were conducted to examine the nature of the relationship. The total SVQ scale (SVQ-Total) was used in these analyses (as explained in Chapter 5). When interpreting correlations, it is important to note that for all measures a higher score is indicative of greater impairment, apart for the VABS-3 whereby lower scores are indicative of greater challenge. To investigate if individual-level factors predict heightened social vulnerability (aim 2), a multiple linear regression was performed with SVQ-Total as the dependent variable. To document the profile of parent protection behaviours and test the relationship with social vulnerability (aim 3), the pattern of endorsement across the OP measure was examined and a correlation was run between total score on the OP measure and SVQ-Total.

Assessment of internal consistency was made using Cronbach's alpha, with a score of between 0.7 to 0.8 considered satisfactory and a values of 0.9 and above deemed desirable. The level of significance was set at $p < .05$. Effect sizes were calculated and used throughout the Results section to aid interpretation of the analyses. For correlations an effect size of .1 is small, .3 is medium, .5 is large, according to Cohen (1988). For Cohen's *d* an effect size of < .2 indicates a small effect, between .2 and .5 a medium effect, and > .8 a large effect. The results to each of the research questions are presented in turn.

6.4 Results

The first aim of the study was to investigate the association between social vulnerability and individual characteristics of EF skills, levels of anxiety, social reciprocity and adaptive behaviour skills. The profile of social vulnerability is reported in Chapter 5.

6.4.1 EF Skills and Social Vulnerability

Table 6.1 displays descriptive results for the BRIEF-2 subscale, indices, and GEC scores. The percentage of CYP in the clinical ranges for BRIEF-2 indexes is shown in Figure 6.1.- Eighty-eight percent of the total sample had a GEC T-score in the clinically significant range (63% significantly clinically elevated, 25% potentially clinically elevated). Similarly, the mean score across the three BRIEF indices was in the clinically elevated range. At the subdomain level, the average score on Shift, Initiate, Working Memory and Task Monitor was clinically elevated. The remaining scales had an average score in the potentially clinically elevated range (see Figure 6.2 for the classification breakdown across the nine BRIEF subscales). Therefore, at the group level, the sample demonstrated significant difficulties in EF. A repeated measures ANOVA found no significant difference between T-scores on the three BRIEF indexes, $F(2, 60) = 1.77$, $p = .18$, $\eta_p^2 = .06$). T-scores on GEC, BRI, ERI and CRI showed a negative association with chronological age meaning that greater proficiency in these EF related to increasing age. However, this association was only statistically significant in the case of BRI (see Table 6.1). There was no significant difference in the GEC or three index scores between males and females in the sample (all p's > .05).

GEC, BRI, ERI and CRI were normally distributed as per Shapiro-Wilk (p's > .05). To test the hypothesis that greater challenges in EF overall and in EF domains would be associated with greater social vulnerability, correlations were performed between SVQ Total and GEC, and the three index scores. SVQ Total was significantly, positively correlated with

GEC (r = .6, p < .001, 1-tailed test), BRI (r = .54, p < .001, 1-tailed test) and CRI (r = .59, p < .001). The association between SVQ Total and ERI was not significant (r = .29, p =.05).

Figure 6.1

BRIEF-2 T-Scores at the Total Level and for the Three Indexes: Percentage of CYP in the Typical Range (60 < T), Mildly Elevated Range (60 ≤ T ≤ 64), Potentially Clinically Elevated Range (65 ≤ T ≤ 69) and Clinically Elevated Range (T ≥ 70)

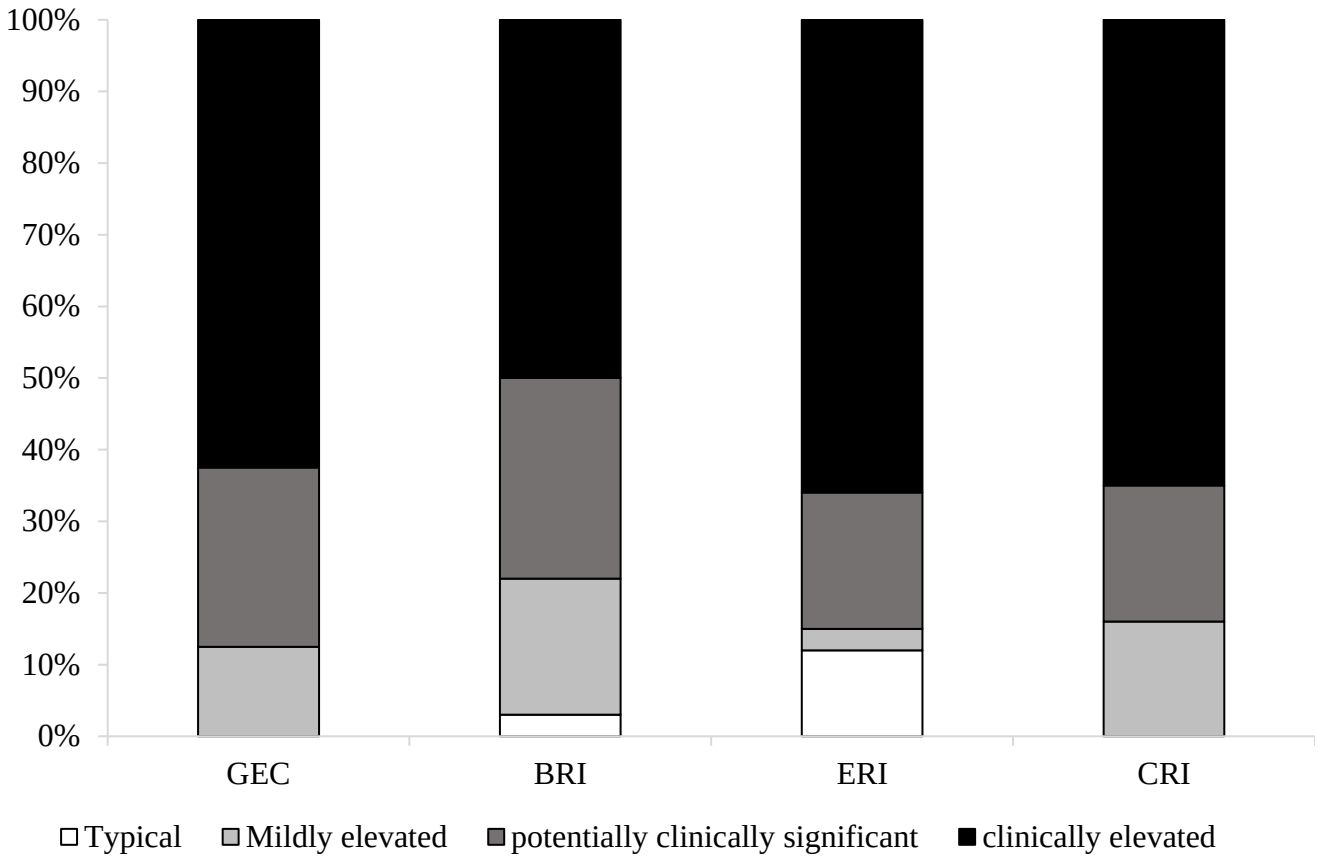


Figure 6.2
Classification of T-Scores for the Nine BRIEF-2 Scales

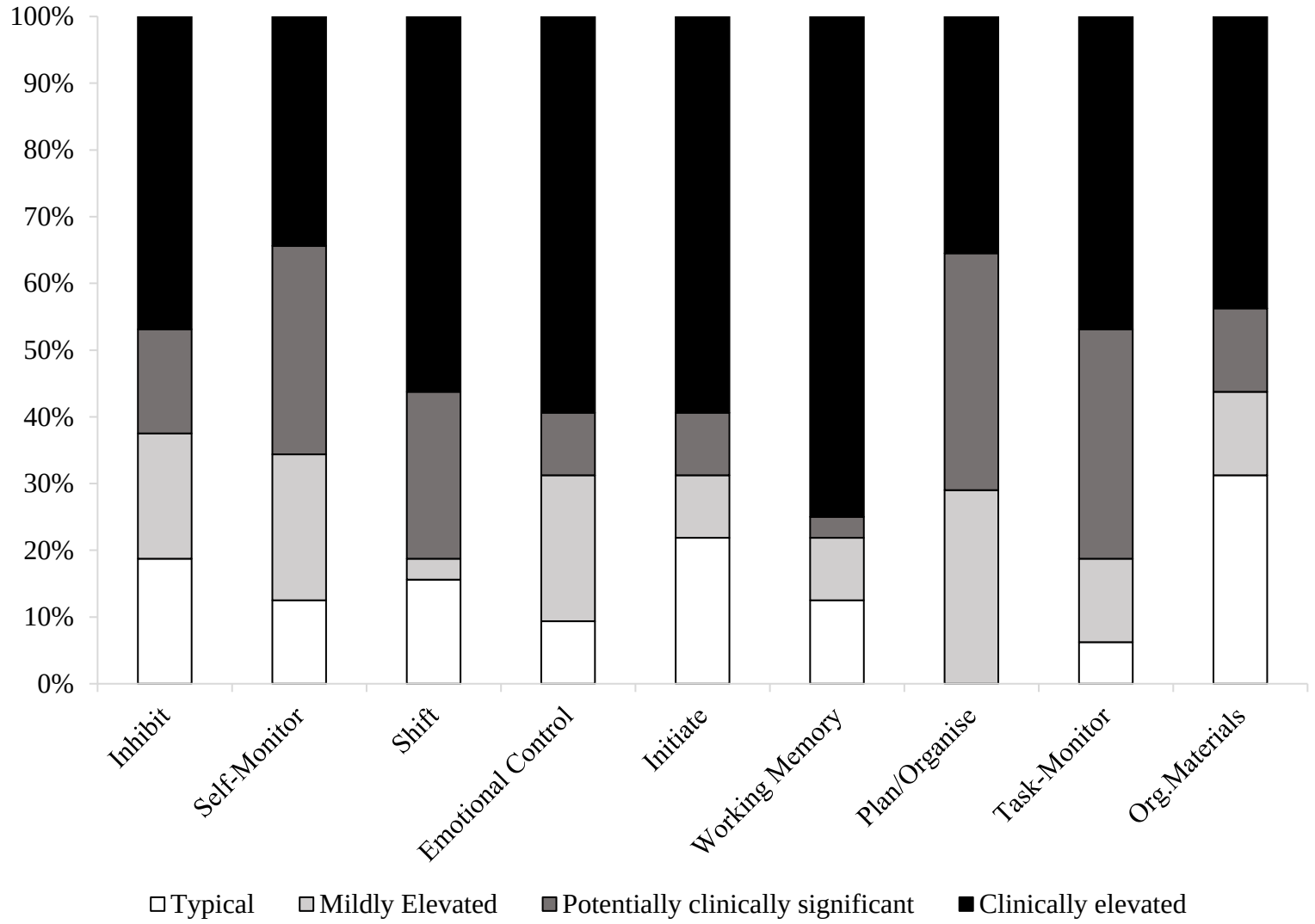


Table 6.1

Descriptive Statistics for the Subscale, Index and Scale Scores on the BRIEF-2 and Correlations With Chronological Age

BRIEF-2 component	Raw score		T-score			
	M (SD)	Range	M (SD)	Range	r	*p*
GEC	142.75 (16.62)	111-169	74.03 (7.57)	60-86	-.23	.21
BRI	27.16 (5.11)	17-36	70.06 (10.64)	50-90	-.36	.04
Inhibit	17.63 (3.73)	11-24	69.56 (11.47)	51-90		
Self-Monitor	9.53 (1.87)	5-12	67.09 (9.33)	44-80		
ERI	35.81 (5.77)	22-47	72.09 (9.82)	49-90	-.22	.24
Shift	17.16 (3.01)	11-23	71.03 (10.26)	50-90		
Emotional Control	18.66 (3.71)	10-24	69.56 (9.65)	46-83		
CRI	80.55 (8.63)	65-95	73.26 (7.01)	62-86	-.1	.6
Initiate	12.22 (2.24)	7-15	70.06 (10.12)	48-84		
Working Memory	20.13 (2.87)	14-24	73.44 (8.10)	56-84		
Plan/Organise	20.10 (2.32)	17-24	69.06 (6.07)	61-80		
Task-Monitor	13.41 (1.54)	9-15	70.69 (7.14)	57-82		
Org. Materials	14.25 (2.6)	10-18	65.78 (8.17)	51-77		

GEC = Global Executive Composite; BRI = Behaviour Regulation Index; ERI = Emotional Regulation Index; CRI = Cognitive Regulation Index; Org. Materials = Organisation of Materials.
Higher raw and T scores are indicative of greater executive dysfunction. T score classifications: mildly elevated ($60 \leq T \leq 64$), potentially clinically elevated ($65 \leq T \leq 69$), clinically elevated ($T \geq 70$)
One response missing for Plan/Organise and CRI.

6.4.2 Adaptive Behaviour and Social Vulnerability

Descriptive statistics for the Adaptive Behaviour Composite (ABC) and domain standard scores on the VABS-3 are outlined in Table 6.2. As per the VABS-3 manual, a score of 70 or below is indicative of low (poor) adaptive behaviour. Eighty-three percent of the sample had an ABC score that signified low adaptive behaviour (see Figure 6.3). At the domain level, the average score across the three domains was in the impaired range, with the lowest score for Daily Living Skills. However, there was much variability with some individuals scoring well below 70 (see Table 6.2 for variability in daily living skills) and others scoring in the adequate range (standard scores 86-114). A repeated measures ANOVA determined that the mean scores on the three domains of the VABS-3 differed significantly ($F(2, 58) = 32.41$, $p < .001$, $\eta_p^2 = .53$). A post hoc pairwise comparison using the Bonferroni correction showed a statistically significant difference between the Daily Living score and Communication ($p < .001$) and Socialisation scores ($p < .001$). The difference between Socialisation and Communication was not significant ($p = .84$). This indicated that daily living skills were a relative challenge compared to communication and socialisation skills; a pattern which has been documented in other WS studies (Fisch et al., 2007; Greer et al., 1997; and see Brawn & Porter, 2018 for a systematic review).

Correlations between SV-Total and VABS-3 ABC and VABS-3 domains were conducted to examine whether facets of adaptive behaviour were related to greater social vulnerability. Lower adaptive abilities overall and in the areas of communication, daily living skills and socialisation were all associated with greater social vulnerability (see Table 6.2 for the results of the correlational analyses).

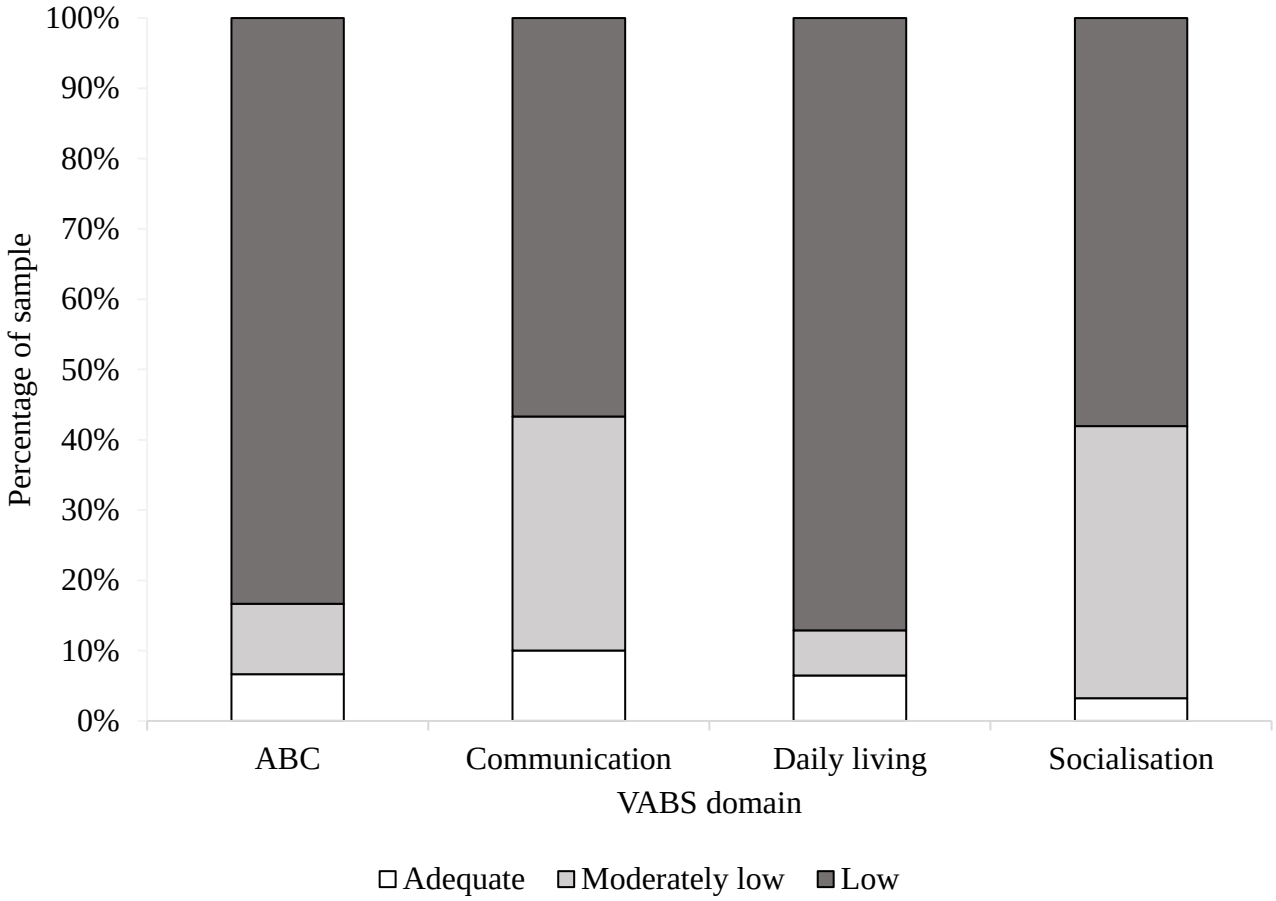
Table 6.2
Mean Scores for the Adaptive Behaviour Composite (ABC) and Vineland-3 Domains

VABS-3 domains	Mean (SD)	Range	Correlation with SVQ-Total	
			r	P
ABC	64.90 (9.13)	50-87	-.59	<.001
Communication	70.17 (12.94)	49-105	-.6	<.001
Daily living skills	55.39 (14.38)	34-94	-.49	.003
Socialisation	68.35 (9.67)	48-89	-.51	.002

Scores are standard scores with a mean of 100 (SD=15), range 20-140. As per the VABS manual, scores ≤ 70 are considered 'impaired'. Correlations are one-tailed.

Figure 6.3

Percentage of the Sample with Adaptive Behaviour in the VABS classification



6.4.3 Social Reciprocity and Social Vulnerability

Descriptive results for SRS-2 total and subscales scores are reported in Table 6.3. The percentage of children and young people in the 'normal', 'mild', 'moderate' and 'severe' classifications for each subscale are shown in Figure 6.4. For the sample as a whole, the average SRS-2 score was indicative of 'moderate' impairment in social reciprocity at the total level. Only 3 children and young people with WS (9%) received scores indicative of 'normal' social reciprocity overall (Total T-score <59). A similar pattern was found across most of the social reciprocity domains, with average scores for Social Awareness and Social Communicative indicative of 'moderate' impairments, and average scores for Social Cognition and Autistic Mannerisms indicative of 'severe' impairments (see Table 6.3). In contrast, the average score for Social Motivation fell within the 'normal' range, and 53% of the sample received scores within this range. Less than 20% of the sample scoring in the

'normal' range for Social Awareness (19%), Social Cognition (9%), Social Communication (16%) and autistic mannerisms (3%). This profile is comparable to what has been reported in other studies of CYP with WS.

Exploration of the variable SRS-2 Total (raw) indicated the data were normally distributed (W = .97, p = .49). A Pearson correlation found chronological age was not significantly correlated with Total score on the SRS-2 (r = -.16, p = .39) indicating that social reciprocity difficulty was not related to age in the WS sample. Similarly, there was no significant difference in SRS-Total score between males (M = 89.42, SD = 25.91) and females (M = 89.20, SD = 22.56), $t(30)$ = -.03, p = .98, d = .009 (Levene's test not significant – p = .45).

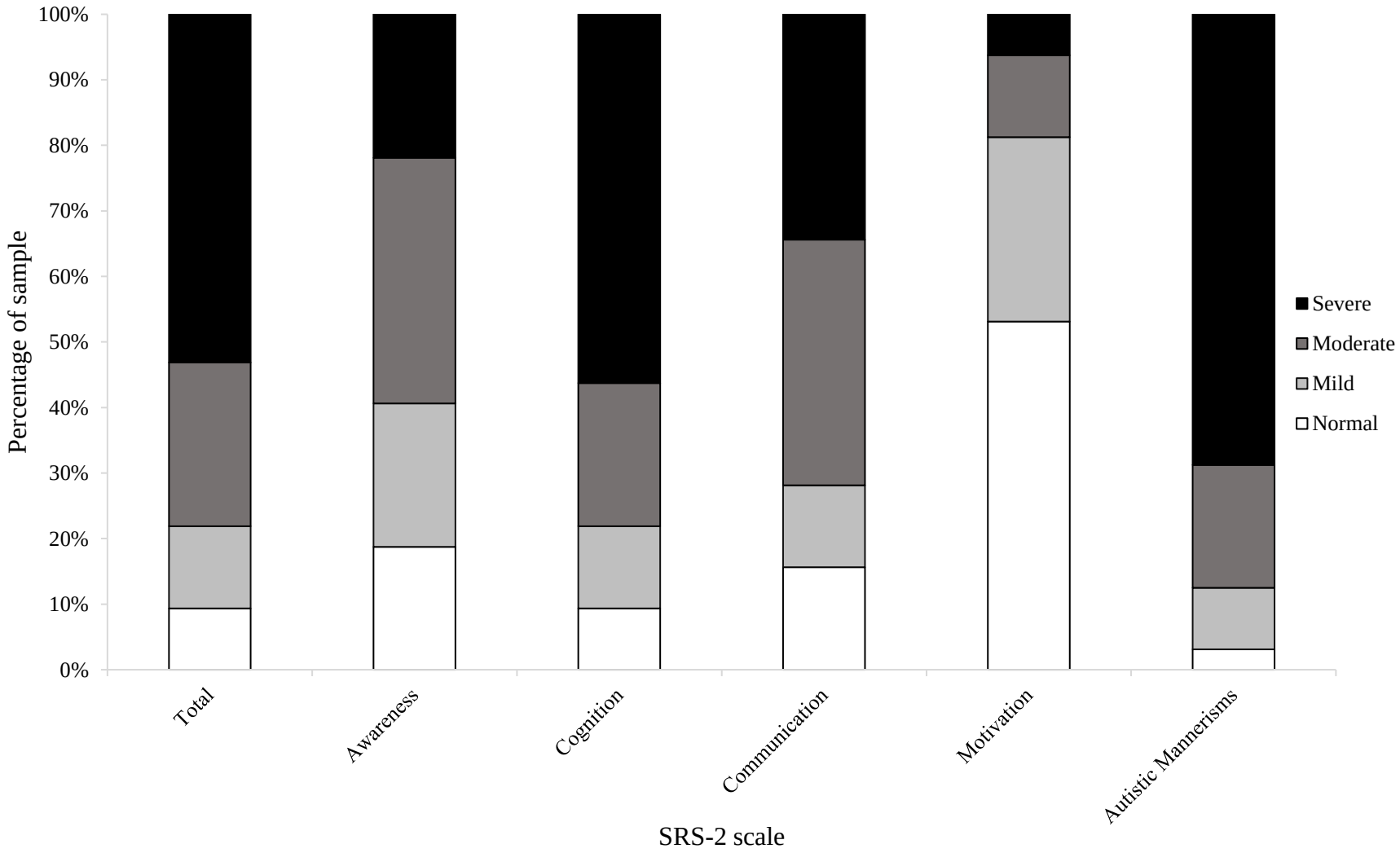
To test the hypothesis that greater social vulnerability would be related to greater difficulties in social reciprocity, correlation analysis was conducted. As predicted, there was a significant positive correlation found between SRS-Total and SVQ-Total score with a large effect size (r = .66, p < .001, one-tailed test), suggesting that greater social reciprocity impairments are associated with increased social vulnerability in CYP with WS.

Table 6.3

Descriptive Results on the Social Responsiveness Scale-2

SRS-2 scales	Raw scores		T scores	
	Mean (SD)	Range	Mean (SD)	Range
Total	89.28 (23.46)	47-134	74.06 (9.43)	55-90
Social Awareness	11.41 (3.22)	4-18	68.97 (9.91)	45-90
Social Cognition	19.59 (5.62)	10-31	76.25 (10.13)	59-90
Social Communication	28.13 (8.64)	12-45	71.03 (9.80)	51-90
Social Motivation	9.63 (4.83)	2-21	58.63 (10.49)	43-86
Autistic Mannerisms	20.53 (6.08)	7-35	79.59 (9.78)	56-90

Figure 6.4
Bar Chart Showing the Classification on the SRS-2 as per T-scores



6.4.4 Anxiety and Social Vulnerability

To examine the profile of anxiety, ASC-ASD scores were examined at the total scale, subscale and item level. Total scores ranged from 1 (*n* = 1) to 56, out of a maximum possible score of 72, highlighting the variability. The mean total score was indicative of elevated anxiety at the group level (see Table 6.4 for ASC-ASD-P descriptive results). For reference, a study of children with WS aged 4-9 years using the ASC-ASD-P reported an average score of 13 (Glod et al., 2019). Of the 31 parents/caregivers who completed the measure, 48.4% (15/31) reported the presence of significant levels of anxiety (5/15 scored above the primary cut-off of 20 ≤ total ≤ 24, 10/15 scored above the more conservative cut off point of >24).

As ASC-ASD subscales vary from five to eight items, analysis of subscales was explored as mean score per item to allow for comparison between subscales. Mean scores per item were highest for Uncertainty ($\overline{x}$ = 1.11, SD = 0.41), followed by Separation Anxiety ($\overline{x}$ 0.90, SD = 0.17), Performance Anxiety ($\overline{x}$ = 0.86, SD = 0.14) and Anxious Arousal ($\overline{x}$ = 0.58, SD = 0.31). Hence, uncertainty seemed to characterise the anxiety profile and the most frequently endorsed item on the ASC-ASD was within the Uncertainty subscale: 96% of parents reported that their child "*always needs to be prepared before things happen*", at least sometimes. While Uncertainty was most frequently endorsed, item "*my child is afraid of entering a room full of people*" showed a different pattern with 55% of parents endorsing 'never', 42% 'sometimes', 3% often and 0% 'always', which provides tentative evidence that anxiety around uncertain situations may not extend into social situations. Lowest endorsement rates were for the items within the Anxious Arousal subscale.

A Spearman's correlation was conducted to assess the relationship between ASC-ASD Total and chronological age as the variable ASC-ASD Total was not normally distributed (Shapiro-wilk W = .93, *p* = .034). The correlation was not significant, *rho* = -.32, *p* = .08 (two-tailed), nor were the correlations between ASC-ASD-P subscales scores and chronological age (all *p*'s >.5, but negative relationship). There was a significant difference in ASC-ASD-P total score and gender with a large effect size, with females scoring significantly higher (*n* =19, *M* = 24.05, *SD* = 14.22) compared to males (*n* = 12, *M* = 13.83, *SD* = 9.12), *t*(29) = 2.21, *p* = .04, *d* = 0.82. Equal variances were assumed as Levene's Test for Equality of variances was not significant.

Table 6.4

Total and Subscale Scores on the ASC-ASD-P

ASC-ASD-P	Mean (SD)	Range (max possible score)	Correlation with SVQ-Total	
			R / rho	p
Total	20.10 (13.32)	1-56 (72)	.23[a]	.11
Separation Anxiety	4.52 (3.72)	0-14 (15)	.19[a]	.31
Uncertainty	8.87 (5.55)	0-22 (24)	.18	.34
Performance Anxiety	4.32 (3.07)	0-13 (15)	.01	.97
Anxiety Arousal	3.45 (3.47)	0-13 (18)	.42[a]	.02

Note. Spearman's rho correlations as subscale was not normally distributed

Only total score was a one-tailed correlational analysis, others two-tailed

To test the hypothesis that greater social vulnerability would be associated with greater anxiety, correlation analysis was conducted. Contrary to the prediction, the correlation between SVQ-Total and ASC-ASD Total was not significant (see Table 6.4*).* Exploratory correlations were conducted between SVQ-Total and subdomains. Note that Anxious Arousal shows as significant but did not hold up after corrected for multiple comparisons (.05/4 = .01). The lack of a meaningful relationship between anxiety and SV was further supported when the sample was split into high/low anxiety groups (high = total ASC-ASD-P ≥20, low = ASC-ASD-P <20). An exploratory independent sample t-test found no difference in SVQ-Total between elevated anxiety (*M* = 79.2, *SD* = 11.72, *n* = 16) and non-elevated anxiety (<20; *M* = 73.63, *SD* = 8.97, *n* = 15) groups, *t*(29) = 1.49, *p* = .15). See Figure 6.5.

Figure 6.5

Social Vulnerability Score for CYP in the Sample With Elevated Anxiety (n=15) and Non-Elevated Anxiety (n = 16), as per the ASC-ASD-P

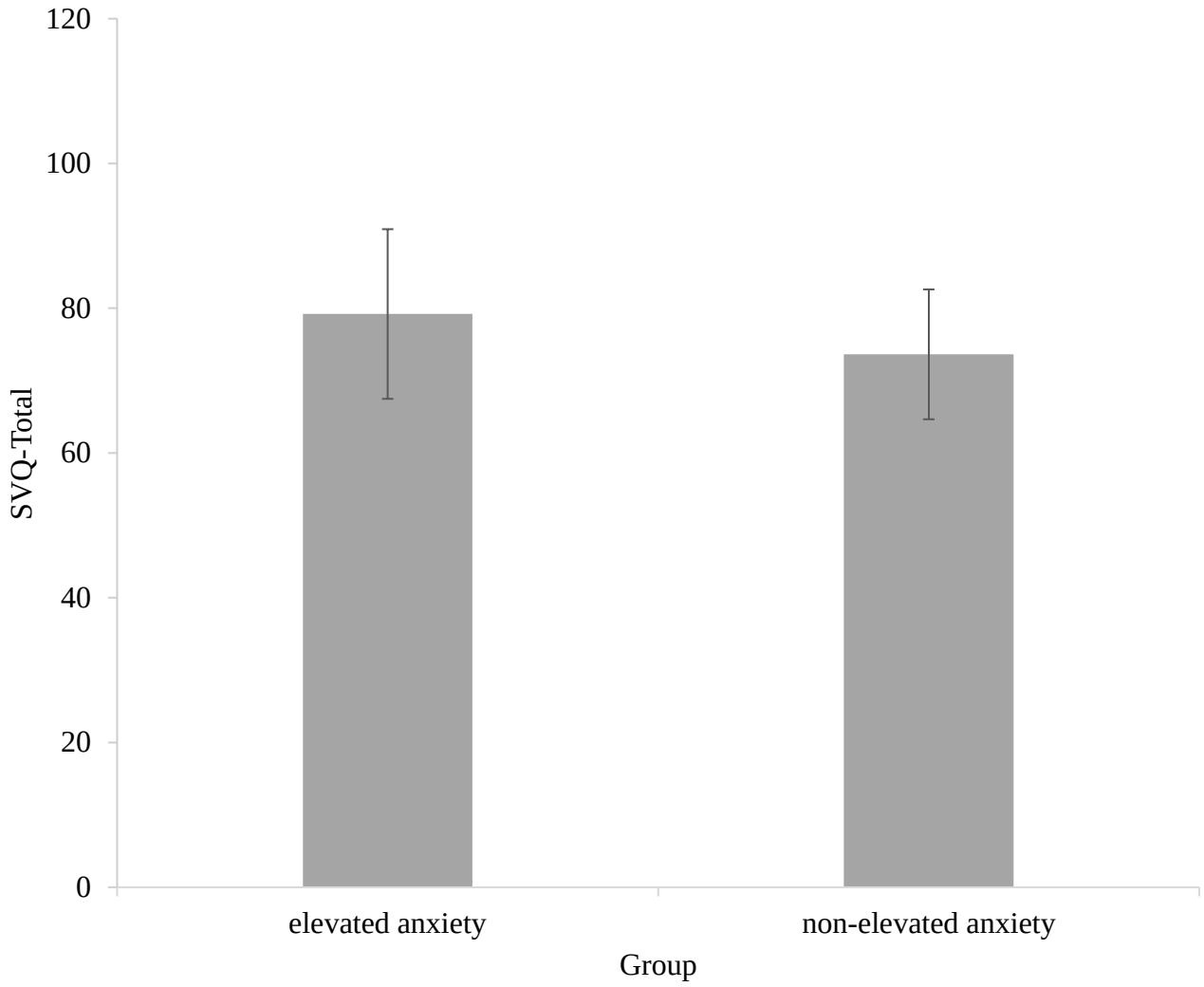
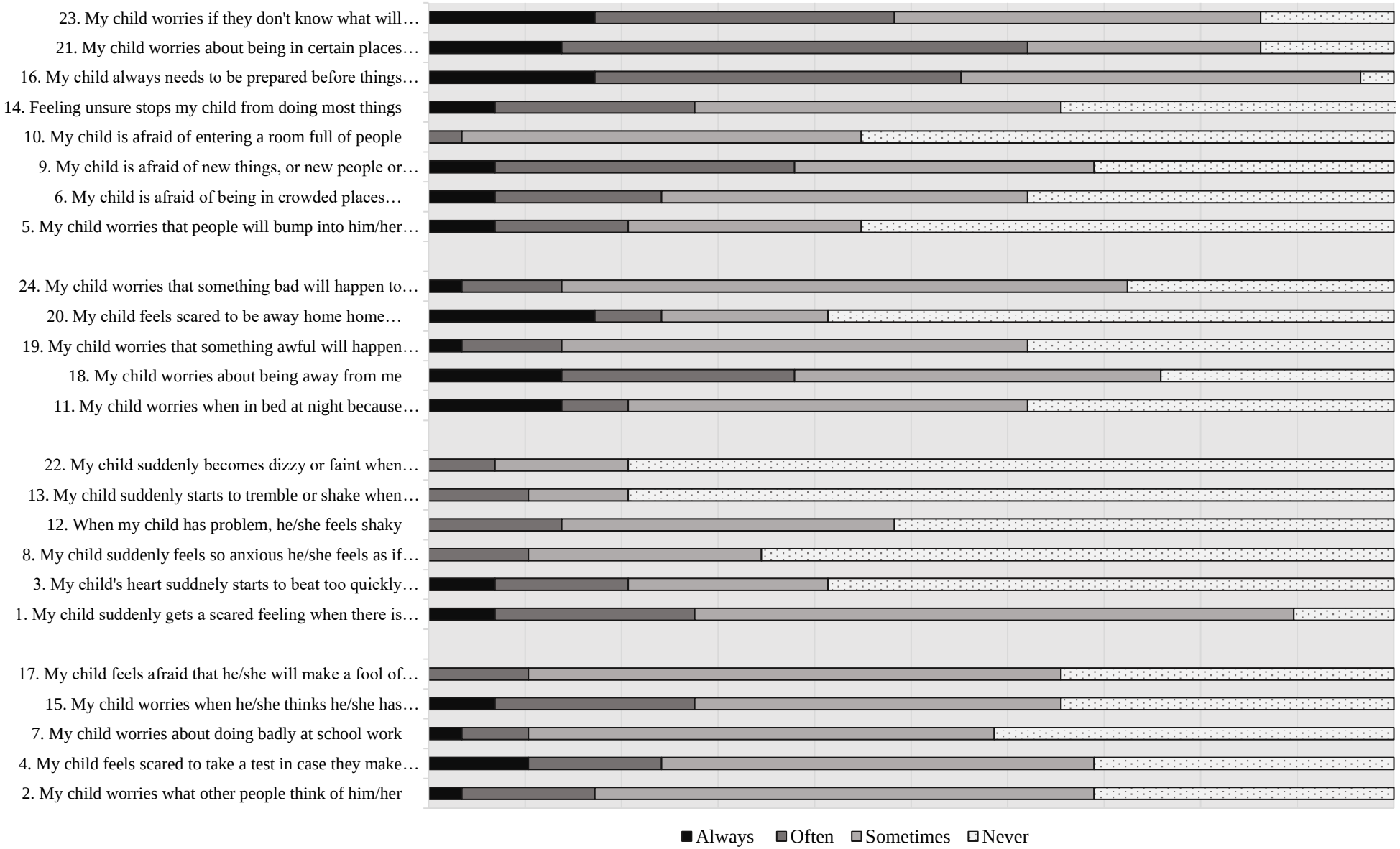


Figure 6.6

Endorsement Patterns for Each of the Items on the ASC-ASD-P



6.4.5 Predictors of Social Vulnerability

In the regression analysis, the following variables were entered as predictors as previous analysis had identified a significant association with SVQ-Total: SRS-2 total, BRIEF BRI, BRIEF CRI and VABS-3 ABC. Using the Enter method (as the sample size was not sufficient for stepwise), the initial model generated was significant and explained 49.7% of the variance in SVQ-Total, $F(4,28) = 7.91$, $p < .001$. However, BRIEF BRI ($p = .24$), BRIEF CRI ($p = .22$) and VABS-3 ABC ($p = .81$) were not significant predictors of SVQ-Total. The only significant predictor was SRS-total ($p = .04$). Therefore, the model was rerun with only SRS-Total as predictor. The model explained 45% of the variance in SVQ-Total and was significant, ($F(1,28) = 24.03$, $p < .001$).

6.4.6 Parent Protection Behaviours and Social Vulnerability

The final aim was to document the profile of parent protection behaviours and investigate the relation with social vulnerability. One participant in the WS group and five participants in the neurotypical group did not return the questionnaire. A further two participants in the neurotypical group responded to <95% of the items (responded 'not appropriate') and were excluded from analysis. Therefore, descriptives and analysis relating to this research question were run on 31 CYP with WS and 28 neurotypical CYP. The average OP score was significantly higher in the WS group ($M = 40.35$; $SD = 13.13$, range 14-66, maximum score = 76) compared to the neurotypical group ($M = 28$, $SD = 11$, range 12-59). There was no significant difference between OP score and child gender for the WS group ($t(29) = .82$, $p = .42$, 2-tailed) or the neurotypical group ($t(26) = .89$, $p = .38$, 2-tailed). The OP score was however significantly correlated with child chronological age in the WS group, $r = -.41$, $p = .02$ and the same relationship was found for neurotypical group, but a stronger association ($r = -.69$, $p < .001$). The negative significant relationship here indicates that parents/caregivers of younger children tended to report higher levels of protection behaviours. An exploratory Pearson correlation was run to examine the association between protection behaviours and social vulnerability for CYP with WS. SVQ-Total was positively correlated with OP total, $r = .48$, $p = .003$ (one-tailed test). Parents of CYP reported to be more socially vulnerable also endorsed higher parent protection behaviours.

6.5 Discussion

This study addressed a critical gap in the literature by exploring social vulnerability and its association with individual and family characteristics in a group of CYP with WS.

Correlational analyses provided support that greater challenges in areas of EF, adaptive behaviour and social reciprocity (but not anxiety), are related to greater social vulnerability. The regression analysis provided evidence that challenges in social reciprocity are a meaningful predictor of heightened social vulnerability. A particularly novel aspect of the study was to consider social vulnerability through a wider lens about family characteristics, providing the first evidence on the association with parent protection behaviours. Findings are discussed per research question below, before turning to the limitations and future directions.

6.5.1 Relationship Between Social Vulnerability and Individual Factors

Few studies have gone beyond documenting rates of social vulnerability and victimisation in WS, to understand the factors that contribute to heightened social vulnerability for some people with WS (but see Fisher, Moskowitz, et al., 2013; Lough, 2016; Lough & Fisher, 2016b). Therefore, the first aim of the study was to examine how social vulnerability relates to common features of the WS cognitive, behavioural and emotional phenotype that are reported to be important in the broader literature on social vulnerability (Greenspan et al., 2001; Pinsker et al., 2010; Pinsker & McFarland, 2010; Sofronoff et al., 2011). Consistent with previous studies on WS, the group of CYP in the current study were reported to have extensive challenges across EF, adaptive behaviour, social reciprocity and anxiety (Brawn & Porter, 2014; Fisher, Lense, et al., 2016b; Greiner de Magalhães et al., 2022; Thurman & Fisher, 2015; Woodruff-Borden et al., 2010).

Greater EF challenges associated with higher social vulnerability. As predicted, greater difficulties in EF overall were associated with higher social vulnerability, providing support for the role that cognitive factors play in social vulnerability. This finding aligns with previous reports of a link between challenges in EF and social vulnerability in older adults (Pinsker & McFarland, 2010) and EF and peer victimisation in autistic adolescents (Kloosterman et al., 2014). In the current sample, challenges in the EF domains of behavioural and cognitive regulation, but not emotional regulation, were associated with greater social vulnerability. Taking behavioural regulation first, the behavioural regulation index (BRI) of the BRIEF-2 includes items on inhibition and self-regulation. Therefore, the association between greater difficulties in these domains and higher social vulnerability supports the hypothesis and is consistent with previous evidence on the role of inhibitory control in the WS social profile (Little et al., 2013; Ng-Cordell et al., 2018). For instance,

Little et al. (2013) reported a link between poor response inhibition in WS, measured using the sun-moon stroop task (Archibald & Kerns, 1999) and high social approach behaviours to unfamiliar faces. Poor response inhibition is also relevant in the context that young people with WS struggle with interpersonal distance regulation (Lough et al., 2015) and maintain a smaller personal space boundary when interacting with unfamiliar people (Lough et al., 2016). It could be that CYP with particular challenges in behavioural regulation are also the same individuals making more inappropriate social approaches, which contributes to greater social vulnerability. Future research should examine the associations between behavioural regulation, social approach behaviours and social vulnerability in the same study, but using different methods to overcome the issue of shared variance across parent report.

The study also generated the first evidence of an association between poorer cognitive regulation and greater social vulnerability. As per the design of the BRIEF-2, the cognitive regulation index (CRI) assesses the specific EF areas of working memory, planning, initiating, monitoring and organisation. While the CYP in the study had significant difficulties across all these subdomains, challenges in working memory were most evident with 75% of the sample scoring in the clinically elevated range. Working memory difficulties are well documented in WS (Vicari et al., 2003; Rhodes et al., 2011), but there is little evidence on the role that working memory plays in relation to the WS social profile. To navigate social interactions involves constantly holding, updating, and manipulating social information about others (e.g. their mental states, past behaviours, personality traits and relationships). Therefore, memory likely interacts with social cognition in various important ways (Spreng, 2013; Meyer & Collier, 2020). For CYP with poorer working memory, this social information integration may be more challenging, which in turn may impact on interpersonal outcomes, like social vulnerability.

The wider literature has reported that other aspects of cognition and EF may also play a role in relation to social vulnerability. For instance, of the EF components analysed in a study of social vulnerability in older adults, verbal fluency and abstract reasoning explained the greatest variance. The authors proposed that having proficient language and reasoning skills may help identify deception, problem-solve and find a strategy when faced with an adverse situation (Pinsker & McFarland, 2010). Similarly, adults with ID with lower social vulnerability scores have higher scores on the Raven's Coloured Progressive Matrices (RCPM), a measure of fluid intelligence which assess non-verbal, abstract reasoning skills (Tabin et al., 2020). There has been far less research on how profiles of cognitive EF skills

relate to WS social behaviours, and the findings here indicate that future research on social vulnerability should consider the relative contribution of specific cognitive and behavioural EF domains.

Greater adaptive behaviour challenges associated with higher social vulnerability. In line with the hypothesis, greater difficulties in adaptive behaviour overall and across the domains of communication, daily living and socialisation were associated with higher social vulnerability. These findings are consistent with those of Lough and Fisher (2016b) which found that adults with WS who had greater functional abilities, as measured by the Activities of Daily Living (Seltzer & Li, 1996), were reported to have lower social vulnerability (when social vulnerability was measured by self and informant-report). Similarly, in a study of adults with ID, individuals with less support needs in areas of community living and health and safety had lower social vulnerability (Tabin et al., 2020). The results of the current study indicate that everyday functional skills in communication, daily living and socialisation may be important in protecting against social vulnerability in adolescence in WS. All associations were strong, but adaptive difficulties in communication showed the strongest association with higher social vulnerability ($r = .6$). This is an interesting finding as a previous study of CYP with WS found no association between social vulnerability and social communication when measured using the Social Communication Questionnaire (Lough, 2016). Future research is therefore needed to clarify the relationship. Communication challenges was raised in Chapter 1 when outlining common aspects of the WS social profile. The evidence in this area suggests that CYP with WS can often be verbose (Reilly et al., 2004), but experience difficulties in pragmatic language (Laws & Bishop, 2004), sustaining conversational flow (Stojanovik, 2006), and may engage in seemingly sophisticated language, such as idioms and cliches, but do so in an inappropriate manner without fully understanding the meaning of social nuance (Udwin & Yule, 1990). Therefore, difficulties in these areas may impact on social vulnerability in various ways, such as failing to detect subtle deceptive cues in language or disrupting the flow of an interaction if not picking up on jokes (Sullivan et al., 2003).

One question is to what extent greater skills in adaptive behaviour is a proxy of greater intellectual ability overall. This study did not include an assessment of IQ and the literature to date on the relation between adaptive functioning and IQ is inconclusive – some studies have reported no meaningful difference (M. K. Greer et al., 1997), while others have found a discrepancy in both directions with higher/lower levels of adaptive skills in relation

to IQ (Brawn & Porter, 2014; Fisch et al., 2007; Mervis & John, 2010). In a subsample of 28 adults with WS, Lough and Fisher (2016b) found that IQ (as measured by the KBIT) showed no meaningful relationship with social vulnerability at the total level or across domains on the SVQ, yet an individual's ability to perform daily living tasks independently did correlate with social vulnerability. While further studies are needed to confirm the role of IQ in these relationships, the findings from the current study alongside Lough and Fisher (2016b) indicate that skills in adaptive behaviour may help to protect against social vulnerability.

Greater social reciprocity challenges associated with higher social vulnerability. The results also showed that higher social vulnerability was associated with greater challenges in social reciprocity. This finding is consistent with the work of Lough (2016) using the same measures with a similar aged group of CYP with WS. Navigating social interactions relies on an array of social skills, therefore it is logical that challenges in these areas may impact on social vulnerability. However, the evidence of a relationship here requires interpreting with caution as it is not clear to what extent the SVQ and SRS-2 overlap in the constructs measured. Nevertheless, if we assume that social aptitude generally is important, future research should consider making assessments of specific components of social skills (e.g., social drive versus understanding of others' intentions) and teasing apart the relation with social vulnerability. The importance of social reciprocity skills will be addressed further when discussing the findings from research question 2 (regression analysis).

No evidence of an association between levels of anxiety and social vulnerability. In this study, levels of anxiety were not meaningfully related to social vulnerability when examined through correlational analysis or when the sample was divided into high/low anxiety groups. The lack of association with anxiety is further supported by the finding that emotional regulation skills (BRIEF-2 analysis) were not related to social vulnerability. A lack of relationship with anxiety was contrary to expectations and contrasts with Lough (2016) who reported a relationship between heightened social vulnerability and heightened anxiety in CYP aged 8-16 years. On the one hand, this discrepancy could be due to methodological differences in the assessment of anxiety – the current study used the ASC-ASD (Rodgers et al., 2016) whereas Lough (2016) used the SCAS-P (Spence, 1998). While the ASC-ASD has not been validated for use in CYP with WS, the group profile in the current study showed that approx. 50% of the sample had elevated anxiety which is consistent with previous reports of anxiety in this age group (Lough, 2016; Royston et al., 2017), providing cautious preliminary

support for the measure. It is also worth noting that the association between social vulnerability and anxiety reported in Lough (2016) was found to be mediated by SRS-2 score, which suggests that attention should be directed towards the association between social vulnerability and social reciprocity. Nonetheless, as this is only the second study to have considered the relationship between anxiety and social vulnerability and due to the relatively small samples, future studies should bring clarity to this issue.

6.5.2 Predictors of Social Vulnerability

The second aim of the research was to investigate whether the personal correlates of social vulnerability would be predictive of greater social vulnerability. In the regression model, lower social reciprocity skills were the only significant predictor of higher social vulnerability. While difficulties in behavioural EF, cognitive EF and adaptive behaviour all showed strong associations with social vulnerability, these variables did not meaningful contribute to social vulnerability in the model. This finding indicates that strengths in areas of social reciprocity may protect against social vulnerability in CYP with WS and this is important because less than less than 10% of the sample were reported to have social reciprocity skills in the 'normal' range as per the SRS-2 classification. The role of social skills was also emphasised in Chapter 5 when parents reported teaching their young person social skills, awareness of vulnerable situations and how to respond, as a strategy to protect against social vulnerability. The findings are consistent with theoretical conceptualisations of social vulnerability that have identified social intelligence as an important factor (Greenspan et al., 2001). However, social reciprocity is multi-faceted, and it is well established that many CYP with WS show differences across social domains, including communication, social perception and social interest/drive (for a review see Thurman & Fisher, 2015). In the context of a potentially manipulative situation, challenges across all these areas are relevant to consider. One potential domain to examine in more detail in relation to social vulnerability is social cognition. Difficulties evaluating social cues and the intentions of others may make individuals more vulnerable to being manipulated or deceived. Making appropriate trust judgements was identified as an area of challenge from the interviews in Chapter 3 and other studies have emphasised that individuals with WS can find deciphering more complex emotional states like trustworthiness difficult (Hanley et al., 2013) and this is evident in social decision-making (Martens et al., 2012; Ng et al., 2015; Riby, Kirk, et al., 2014). Indeed, in the broader literature, theory of mind skills have been identified to play an important role in explaining social vulnerability (Pinsker & McFarland, 2010). For example,

theory of mind mediated the relationships between EF and language ability and social vulnerability in neurotypical children (Seward, 2016).

6.5.3 Social Vulnerability and Family Characteristics – The Role of Parent Protection

A novel aspect of the study was to examine social vulnerability in relation to broader characteristics about the lives of CYP with WS, focusing on parent protection behaviours. As hypothesised, the average score on the parent protection measure for the WS group was higher than seen in the neurotypical group. Like in previous research using the same parent protection measure, parent protection score reduced with CYP age (Clarke et al., 2013; Edwards et al., 2008). While the group of CYP in the current study were older than the measure was initially designed for, the average score was higher (indicating greater parent protection) than in CYP aged 7-12 years (total score of 26.31 in Clarke et al., 2013) and even higher than seen in very young children (32.78 in Edwards et al., 2008). Therefore, while we see the same pattern of protection behaviours decreasing as CYP with WS get older, parents are reporting greater levels of protection behaviours that exceed rates seen in young children. In Chapter 5, when parental independence was examined using the SVQ subscale, it did not increase as CYP got older, however as the SVQ subscale consists of only three items, this research has provided a useful extension. Of course, the constructs measured might be different, but it could also mean that while parents may reduce protection behaviours as CYP get older, they are not necessarily granting greater opportunities for independent social interaction. In line with the prediction, greater parent protection behaviours were associated with higher social vulnerability. While the study was not designed to clarify causation between these two factors (i.e., does greater parental protection feed into higher social vulnerability or is greater protection a consequence of heightened social vulnerability), the study provides preliminary evidence of an association that should be followed up in future studies.

6.5.4 Considerations and Future Directions

Findings from the current study should be considered within the context of several limitations. First, the study was not set up to tell us anything about causality, therefore the relationships identified should be followed up in future inquiry to understand the nature of the relationships more fully. A major limitation is that the study used parent report across all measures which presents the issue of subjective biases and shared variance – for instance, a parent who considers their child to be high on social vulnerability also likely reports high on

protection behaviours. Future research should re-examine the relationships identified in this study using multiple informants and, ideally, multiple methods, including direct behavioural assessments of cognition and behaviour. Additionally, studies should include a standardised measure of IQ to understand if cognitive and behavioural components contribute to social vulnerability above and beyond the contribution of ID.

Another consideration pertains to the analytical approach. This study focused on group-level analysis, however there was heterogeneity in the level of social vulnerability within the group of CYP with WS (Figure 5.3). Previous studies have also recognised wide within-group variability at the cognitive and behavioural level in WS (Järvinen-Pasley et al., 2010a; Little et al., 2013; Porter & Coltheart, 2005). Therefore, it would be valuable to explore the data at the level of individual profiles and consider clustering techniques which have proven fruitful in other WS studies. For example, using cluster analysis Little et al. (2013) identified subgroups of social approach behaviour in CYP with WS and found that membership to a subgroup was best distinguished by inhibitory control skills. Building on this and as advocated by Lough (2016), it would be interesting to examine a subgroup of CYP with WS who score low on social vulnerability to probe heterogeneity of social vulnerability. Such an approach could yield valuable evidence on the characteristics that may be protective and should be prioritised in developing support tools.

One important consideration is the measurement of anxiety and whether the ASC-ASD appropriately captures anxiety as it presents in WS. There are currently no anxiety measures designed specifically for use in WS and, as such, it is common for studies to use tools designed for the general population (e.g. the SCAS). The issue with this approach is that anxiety likely manifests differently in people with neurodevelopmental differences (Kerns et al., 2014). Indeed, the ASC-ASD (Rodgers et al., 2016) was developed in response to this issue and it aims to be a more sensitive measure of anxiety for use with autistic young people. A strength of using the ASC-ASD in the current study is that some of the characteristics of anxiety in autism are also commonly seen in WS, such as sensory sensitivity and fears around uncertainty (Royston et al., 2021). Nevertheless, the ASC-ASD is not tailored for use with groups with ID, nor has it been validated empirically for use with people with WS. It is important to note that a lack of appropriate anxiety measures (and measures of mental health more broadly) is a challenge for research with many genetic syndromes associated with ID (see Crawford et al., 2023 for a discussion in the case of FXS). Given the high prevalence of

anxiety in young people with WS, it is crucial that researchers have tools that can accurately detect and measure anxiety in this group.

While the study has extended knowledge about social vulnerability by examining the wider family context of parent protection behaviours, the parental protection measure used was developed for use with parents of preschool children (Edwards et al., 2008) and has not been validated on the current age group. Nevertheless, the pattern of endorsement indicated that the questionnaire items were relevant, and this is further supported by the reported high internal consistency. It is possible that the items are appropriate in the context of CYP with WS who typically have a mental age that is lower than their chronological age. In other words, parents of adolescent CYP with WS may be exerting protection behaviours that are comparable to parents of much younger neurotypical children. This is supported by the accounts of parents, emphasising that they monitor the social behaviour of their child with WS has WS to a greater extent than their other younger children[9] (Gillooly, 2018). Future research should examine parent protection behaviours in more detail in relation to social vulnerability, but it will be important to first establish whether the OP measure is appropriately capturing these behaviours in the context of families with CYP with WS.

Overall, much more research is needed to disentangle the likely interplay between social vulnerability, adaptive behaviour skills and parent protection. High parental protection may be a consequence of parents' concerns about their son or daughter's social vulnerability, and therefore warrants further investigation in relation to family impact. Equally though, researchers should consider the potential impact of high parent protection behaviours in relation to independence and social inclusion. Tabin et al. (2020) proposes that individuals with greater adaptive skills may be those who experience richer social environments and, in turn, experience opportunities that foster social and problem-solving skills (likely beneficial in navigating vulnerable situations). Future studies should consider the role of other socio-environmental factors in the context of social vulnerability.

Finally, considering the evidence that social reciprocity skills may be an important area for targeted support, a useful next step would be to understand which components of social reciprocity (e.g., awareness, communication, theory of mind, social drive) make the most important contribution to heightened social vulnerability. In Chapter 4 (Paper 2)

[9] Parent quote from Gillooly (2018) in reference to the subtheme 'need for parental supervision' "When we are out, I watch her more than I watch the boys despite the fact they are 3 years younger. They are just more aware, they will stay close to me and they won't wander, whereas she will"

different styles of social interaction contributed to increased social vulnerability, but this study has not answered whether CYP with particularly high levels of social motivation/drive experience social vulnerability to a greater extent. To answer this question, studies should identify a robust measure of social motivation that can capture subtle variations. The SRS-2 is commonly used in WS studies and includes a social motivation subscale. However, Riby et al. (2017) argues against its use in capturing social motivation in WS because the measure is designed to identify low social motivation, as presumed to be the presentation in autistic people. Therefore, by the SRS classification, anything other than reduced social motivation, is deemed 'typical' and indeed over 50% of the CYP in the current study were reported to have social motivation in the 'typical' levels. Further investigation of the association between social vulnerability and aspects of social functioning, including profiles of social motivation, would be a useful extension of this work.

6.5.5 Conclusion

This was the first study to examine the correlates of social vulnerability in CYP with WS, using standardised assessments to measure aspects of cognitive, behaviour, and emotion. WS. It was also the first investigation of social vulnerability in the context of characteristics about the wider family environment. The study highlights noteworthy associations between greater social vulnerability and challenges in areas of EF and adaptive behaviour, and provides preliminary evidence that skills in social reciprocity may play an important protective role. Understanding the factors that relate to increased social vulnerability is fundamental to the development of appropriate support tools and this study has raised important questions for future studies to take forward. Moving beyond personal characteristics and placing more emphasis on the social environment around CYP with WS is important to provide a full account of social vulnerability.

7. CHAPTER 7: GENERAL DISCUSSION

7.1. Chapter Overview

Having described the population of interest and put forward the rationale for studying social vulnerability in WS, this thesis set out to enhance understanding on this topic. Five studies yielded qualitative and quantitative data plus multi-informant insights to address (i) the ways in which social behaviours present in WS and across neurodevelopmental groups (Chapter 4), (ii) friendships from the perspective of people with WS (Chapter 3), (iii) the profile of social vulnerability in CYP (Chapter 4 Paper 2; Chapter 5) and (iv) new evidence on individual and family factors related to social vulnerability in CYP with WS (Chapter 4 Paper 2; Chapter 6). I found abundant evidence to support that social vulnerability is an important feature of the WS social phenotype that warrants further understanding and support early on in development. This final Chapter synthesises the key findings across all Chapters and reflects on the strengths and limitations. It also provides discussion of the implications of the findings and offers suggestions for future research.

7.2. Summary of Key Results

7.2.1. Social Vulnerability is High in Adolescence in WS

The research contributed several noteworthy findings to the literature on social vulnerability in WS, particularly considering the small evidence base to date. The research that came before and provided impetus for the thesis documented poor awareness of stranger danger and increased approach to others, which in turn raised the question of social vulnerability in WS (Jawaid et al., 2012; Lough, Flynn, et al., 2016; Riby, Kirk, et al., 2014; Riby et al., 2017). However, there had been little systematic investigation of social vulnerability itself, particularly before adulthood in WS. An important contribution of the thesis has been to confirm the significance of adolescence in the context of social vulnerability. Using an established tool of social vulnerability, I found evidence to indicate that social vulnerability is high in childhood-adolescence in WS compared to neurotypical levels at this developmental stage (Chapter 4 Paper 2, Chapter 5), but not unlike peers with other forms of IDD (Chapter 4 Paper 2). Taking the WS-neurotypical difference first, there has been one other study to have documented elevated social vulnerability in adolescence in WS (Lough, 2016) and the findings here add further support. In a sense, elevated levels

compared to neurotypical peers is an unsurprising pattern given the constellation of cognitive and behavioural factors associated with WS is not seen in neurotypical development. However, in the first comprehensive investigation of social vulnerability in adolescence (Chapter 5), CYP with WS had elevated levels across all domains of vulnerability measured by the SVQ (except parental independence), indicating a multi-faceted nature to the vulnerability which may be related to different underlying mechanisms and require different types of support.

The salience of the secondary school years and school environment was further emphasised when detailing the profile of social vulnerability (Chapter 5). The real-life examples of vulnerability reported by parents and carers were often linked to the school context (e.g., experiences of peer pressure in school) and this also emerged in the interviews with adults (Chapter 3); many of whom reported bullying and distressing interactions with peers during the formal school years. Taken together, the findings underline adolescence as an important developmental stage for understanding social vulnerability and providing support. More broadly, these results strengthen the idea that the socially gregarious profile often connected with WS (Doyle et al., 2004a; Järvinen-Pasley et al., 2010a; Zitzer-Comfort et al., 2007) is overly simplistic. Rather elevated social vulnerability is an important feature of the social profile for many adolescents with WS that demands more research attention and practical support.

The thesis has also shed new light on the implications of social vulnerability beyond the individual-level, for the lives of families with CYP with WS – a topic that has been given little attention to date. I found that high social vulnerability is a great concern for many parents and families. It is particularly striking that where social vulnerability was less of a concern this was often because families were already implementing close supervision to ensure their young person's social safety. The impact for families was further emphasised by evidence of high levels of parent protection behaviours, which were seen to a greater extent in families of CYP with higher social vulnerability. The findings here compliment those of Gillooly (2018) who also found that parents of CYP with WS endorsed the need for constant supervision when discussing their child's social approach behaviours, indiscriminate friendliness and trusting nature. In addition to parental supervision and restricting social independence, families also reported protecting against social vulnerability by teaching on issues of social awareness and social cognition. It can therefore be assumed that this an area where families would welcome formal support. In sum, the findings underline the importance

of identifying the mechanisms that underpin high social vulnerability and developing approaches to support CYP with WS in their social interactions and support the wider family navigate the concerns.

7.2.2. Concerns Around Social Vulnerability may not be Specific to WS

By studying social vulnerability in WS alongside multiple other neurodevelopmental groups, I found elevated levels of social vulnerability were comparable across autism, FXS and ADHD groups (Chapter 4 Paper 2). This suggests the phenomenon of social vulnerability may be relevant across neurodevelopmental groups with different aetiologies. A note of caution is due here since this research was based on small sample sizes using an abbreviated measure of vulnerability. Nevertheless, the findings raise important questions about the significance of social vulnerability in clinical groups not previously studied in the literature to date and provide impetus for extending research on social vulnerability to other neurodevelopmental groups, including ADHD and FXS. The findings raise the possibility that some factors underlying social vulnerability are transdiagnostic. For instance, many neurodevelopmental groups show differences in EFs (Camp et al., 2016; Carney et al., 2013; Costanzo et al., 2013; Hovik et al., 2017; Rhodes et al., 2011), pragmatic language (Hawkins et al., 2016; Mandy et al., 2017; Mareva et al., 2019; Smith et al., 2017) and social cognition (Bora & Pantelis, 2016b; Pastorino et al., 2021). However, different developmental pathways can result in the same cognitive/behavioural outcome (Karmiloff-Smith, 1998), hence it may also be the case that different pathways related to social profiles, ID, or a combination of these factors, give rise to the same outcome of heightened social vulnerability. Considerably more detailed work is needed to determine the developmental pathways to social vulnerability in CYP with a range of neurodevelopmental profiles.

More broadly, the cross-syndrome findings extend the neurodevelopmental literature by adding support that social phenotypes are best understood as distributed across diagnostic boundaries (Astle et al., 2021). I focused on two aspects of social behaviour implicated in social interactions and which have previously been emphasised in the WS social phenotype – an over approaching interaction style and prolonged eye contact (W. Jones et al., 2000). To date, the WS literature has documented the excessive nature (in quantity) of these features of social behaviour and, in turn, raised the question of social vulnerability (Jawaid et al., 2012; Riby et al., 2017). I found that an active-but-unusual social interaction style was highly endorsed in WS but was also evident (to varying extents) in other clinical groups studied

(e.g., 49% in autistic CYP). Similarly on qualities of eye contact, prolonged eye contact was most evident in the WS group, but also endorsed across groups. Taken together, the evidence indicates that some of the social behaviours deemed 'characteristic' of WS are not unique to WS, rather they are evident across other groups. Again, this supports a rationale for considering social vulnerability across multiple neurodevelopmental groups, but also emphasises non-specificity in the social phenotype within and across neurodevelopmental groups. Of course, the cross-syndrome studies explored just two aspects of social behaviour, and when we consider the entirety of a social interaction the picture across groups is likely more nuanced.

7.2.3. Why Heightened Social Vulnerability?

Having established that social vulnerability is elevated in CYP with WS and a concern for family members, the next logical line of enquiry was to probe the factors that contribute to such high levels. This was a particular focus of Chapter 6, but in fact cross-chapter findings go some way to enhancing our understanding of the factors related to heightened social vulnerability. Overall, the evidence points towards the role of social awareness and social cognition. For instance, difficulties making assessments of trust was a theme that came out of the interviews with adults with WS (Chapter 3) and was echoed in the parent questionnaire data where CYP with WS were reported to have particularly high levels of vulnerability in the domain of credulity. Further, some of the parent strategies to protect against social vulnerability related to increasing knowledge about social dynamics (e.g., teaching on social interactions, identifying others' intentions and socially risky situations). These findings align with experimental WS studies that have documented difficulties with complex social cognitive judgements, including trust evaluations (Hanley et al., 2013; Martens et al., 2012; Ng et al., 2015), and judgements in real life social interactions (Lough, Flynn, et al., 2016).

These initial findings were taken forward for further exploration in Chapter 6 and provided a rationale for including a measure of social reciprocity (SRS-2) when exploring correlates of social vulnerability. Using standardised measures of cognition and behaviour, I found that difficulties in areas of EF, adaptive behaviour and social reciprocity were related to greater social vulnerability, but only social reciprocity was a meaningful predictor. These specific personal characteristics were identified as potential correlates based on evidence on social vulnerability in other groups, and indeed the thesis findings support theoretical

conceptualisations that identify 'social intelligence' as playing an important role in social vulnerability (Greenspan et al., 2001; Sofronoff et al., 2011). This finding makes sense – avoiding adverse social interactions relies on being able to identify cues related to deceptive or manipulative behaviour and then navigating a course of action. Other studies of social vulnerability with autistic and young neurotypical CYP have identified the role of theory of mind (Seward et al., 2018; Sofronoff et al., 2011) and there is wider evidence that social cognition is associated with detecting deceit (Mills & Elashi, 2014; Vanderbilt et al., 2011). Differences in social cognition is transdiagnostic in the neurodevelopmental groups studied in the cross-syndrome work (Chapter 4), so it is plausible that social cognition challenges provide a risk marker for social vulnerability.

Social skills/cognition is an important factor, yet social vulnerability is likely to be a combination of various critical components. While not significant predictors in the model of social vulnerability, the associations with personal factors of EF and adaptive behaviour are novel and warrant further attention. The relation with EF supports existing models of social vulnerability (Greenspan et al., 2001) and has also been shown in studies with older adults with neurological issues (Pinsker & McFarland, 2010). Difficulties in EF play a role in socially disinhibited behaviours common in WS (Little et al., 2013) and increased approach behaviours are likely a component of how social vulnerability is perceived by parents and carers. Therefore, an interesting question for future studies is the potential mediating role of social disinhibition in the relationship between EF and social vulnerability. In other words, does poor EF skills lead to more social disinhibition in real-life social interactions, which in turn leads to the perception of greater social vulnerability? I reported in Chapter 4 that social interaction styles contributed to greater social vulnerability when studied in a large sample (neurotypical and neurodevelopmental samples combined), which suggests it is important to consider the quality of the interaction style.

7.3. Strengths and Limitations

I now turn to the strengths and limitations of the research. A key strength is the range of methods used, including quantitative and qualitative approaches, from questionnaire design with multiple neurodevelopmental groups, to individual-level analysis of adults' own words. Beginning the thesis with qualitative insights from adults generated information on social decision-making and experiences with peers, relevant to social vulnerability. Turning to a cross-syndrome design in Chapter 4 enabled me to consider whether aspects of social

behaviour thought to characterise WS are distinct to the WS profile or shared across other neurodevelopmental profiles. Parent/caregiver quantitative and qualitative data in Chapter 5 provided the first detailed account of the social vulnerability profile in CYP with WS. Finally, the use of standardised tools in Chapter 6 yielded valid and reliable data on cognition and behaviour, to examine how social vulnerability relates to the wider WS profile. A single methodology would not have yielded the same richness and by including multiple approaches the disadvantage of each individual method is more balanced. As limitations have been noted per Chapter, the aim here is to reflect on the limitations of the methodology and approaches across the thesis overall.

7.3.1. Methodological Quality and Design

First, all but one study relied on parent-reported questionnaire measures alone and with often the same parent or carer completing multiple measures (e.g., in Chapter 6 where informants completed a battery of questionnaires on cognition and behaviour). Relying on a single informant across multiple measures is problematic as it may have resulted in shared variance. Additionally, the data are subject to parents and carers' biases and interpretations, and parents might not always be aware of their adolescent son/daughter's behaviour across different environmental contexts. In adolescence, CYP spend most of their time at school and encounter various peer interactions. It is reasonable to expect that social behaviours and challenges related to social vulnerabilities present differently across home and school settings (Klein-Tasman et al., 2011). Indeed, the school environment was dominant in the examples of social vulnerability given by parents (Chapter 5), therefore teacher perspectives on the presentation and impact of social vulnerability in the school/classroom context would help to triangulate parent reports and strengthen the conclusions about where support is needed. The value of a multi-informant approach including teacher perspectives has been evidenced in other WS studies on social functioning and peer relationships (see Gillooly et al., 2021; Klein-Tasman et al., 2011).

Linked to this, assessments of social vulnerability, behaviour and cognition were made through questionnaire report only. This was an essential approach given the constraints at the time of the conducting the research (covid-19 preventing face to face research). However, as these are indirect measurements, I can't draw conclusions about how this relates to real-life behaviour. Hence, taking a multi-method approach (e.g., the addition of behavioural, observational, or experimental data) to the measurement of social vulnerability

would have strengthened the evidence and provided a more objective account (to note that experimental data on social approach preferences was collected as part of an online follow-up study, however the results were not presented in the thesis). Likewise, the informant measures used to examine behaviour and cognition in Chapter 6 were well-established, validated tools for CYP with ID, but direct behavioural assessments would have provided a more robust measure of CYP's abilities and challenges across these domains (see Gerst et al., 2017 for discussion that different types of EF measures likely yield different information).

The SVQ was relied on heavily across Chapters where social vulnerability was the dependent variable of interest. The SVQ was identified as an appropriate tool to meet the aims of the research as it is designed for use with individuals with IDD and can provide assessment across a range of social vulnerability domains. However, it is not without its limitations, and these should be stressed again. Chapter 4 used an abbreviated version of the SVQ to draw conclusions about levels of vulnerability across different groups. Had the full measure been used it would have provided a more reliable estimate of whether social vulnerability is indeed elevated at a comparable level across multiple groups. In Chapter 5, the full SVQ was used to investigate whether levels of social vulnerability in CYP with WS were elevated compared to neurotypical peers. However, this is also problematic because the measure was designed for use with adults with ID not CYP, therefore using an adult tool means that we might have missed information about the profile of vulnerability as it presents in childhood and adolescence. Additionally, the SVQ was not designed for neurotypical comparisons meaning that the measure was adapted to meet this aim. A final note on the SVQ relates to the conclusion that social reciprocity plays an important role in social vulnerability (the only predictor of increased social vulnerability in Chapter 6). It should be noted that the research did not examine potential overlap between the SVQ and the SRS. Therefore, overlap in the constructs being measures (multicollinearity) may be reflected in the significance of the relationship here. Reliance on the SVQ signifies a broader limitation of conducting research on this topic, in that there are very few tools available to measure social vulnerability in CYP with ID.

Another methodological limitation is that the studies did not include carefully matched comparison groups. The neurotypical samples of CYP in Chapters 4-6 provided contextual information about the levels and nature of social vulnerability seen in neurotypical development. However, ideally, the neurotypical and WS groups would have been individually matched on mental age, to draw conclusions about whether levels of social

vulnerability seen in WS are congruent with developmental level. While valuable from a theoretical perspective, I caution placing too much emphasis on neurotypical/WS comparisons as the parent data tells us that social vulnerability is a concern and we would expect that this is something different going on in WS. If future studies do consider matching, this needs careful thought. A mental-age matched comparison group would indeed provide insights about social vulnerability in relation to developmental level but would not provide a comparison on life experience. A group of CYP with Down syndrome would be more suitable for this type of comparison.

Regarding the conclusions about factors related to social vulnerability, it should be noted that the thesis has not meaningfully considered the role of intellectual ability/disability. In Chapter 4 I reported that ID did not fully account for elevated social vulnerability as comparable levels were found in groups not characterised by ID (autism and ADHD). However, a major limitation of this work is the reliance on parent-reported ID status. An objective measure of cognitive ability / ID would have yielded a more robust estimate of the role of ID in explaining social vulnerability. In Chapter 5, social reciprocity was identified as important in explaining social vulnerability, however without a measure of ID, I can't draw conclusions about whether factors like social reciprocity contribute to social vulnerability above and beyond having ID. Previous research has not found an association between general intelligence ability and social vulnerability in neurotypical CYP (Seward, 2016) or adults with WS (Lough et al., 2016), however there is not good evidence on this in WS and ID likely contributes to the ability to detect and respond to manipulative social situations (Greenspan et al., 2001).

A final limitation is the cross-sectional methodology across Chapters which means I cannot make inferences about casual associations between individual factors and social vulnerability (Chapter 6). In other words, it remains to be understood whether high social vulnerability leads to greater challenges in areas of EF, adaptive behaviour and social reciprocity, and increased parent protection behaviours; or whether these characteristics lead to high social vulnerability. Longitudinal data would provide the most robust account of how social vulnerability emerges throughout development and interacts with other developmental processes, but I acknowledge this is a huge undertaking for research with rare groups like WS especially in the context of a PhD.

7.3.2. Sample Size and Generalisability

The nature of the samples recruited also requires consideration. Based on the recruitment approach, it is possible that the studies have a self-selection bias, in that parents who were particularly concerned about their child's social interactions and social safety were more likely to respond to the study adverts. The participant-facing materials in Chapters 5 and 6 stated that we were interested in hearing from families where social vulnerability was not currently a concern and, indeed, we saw variation in levels of social vulnerability. Nevertheless, it is possible that the high levels of social vulnerability reported in the thesis may not be representative of the broader population of CYP with WS or families' experiences. A strategy to make research of this nature more representative would be to foster greater links between researchers and community settings or medical practices where data collection could be carried out in clinic. Additionally, I note that no data was collected on ethnicity or socio demographics of families (beyond parent highest qualification).

Another key limitation is the relatively small sample size across the studies which has likely resulted in reduced power to detect significant results. While not such an issue with the qualitative work in Chapter 3 (see Guest et al., 2006 for suggestion that 6-12 interviews can reach saturation; Namey et al., 2016), the small samples is a significant issue in the quantitative Chapters and was a barrier in being able to conduct more sophisticated analytical models of social vulnerability. Small samples were also evident in the clinical groups recruited in Chapter 4, therefore the finding of elevated social vulnerability across neurodevelopmental conditions should be interpreted with caution. Hence, further detailed enquiry into social vulnerability in different neurodevelopmental groups is required. To draw larger sample sizes and probe social vulnerability across development, I endorse the recommendations of others who have advocated for better collaboration in WS research, drawing upon multi-lab data pooling so that studies are designed to answer the most pressing research questions rather than just what is possible with sample size (e.g. Farran & Scerif, 2022).

7.4. Implications

The research presented here would greatly benefit from replications and extensions. Nonetheless, it is worthwhile considering how these findings might be applied in practice, should they be replicated and considered robust.

7.4.1. For the WS Community

A great deal more work is needed to disentangle what is driving social vulnerability before we understand how best to support families in this area. However, the findings so far should be used by families and organisations like The Williams Syndrome Foundation (WSF), to raise awareness of social vulnerability in WS and advocate for support and resource in this area. The WSF will be particularly interested in the finding that most families are concerned about social vulnerability, particularly how it might exacerbate as their son or daughter gets older. The WSF will also be interested in the strategies implemented by families to protect against social vulnerability as this provides a useful indication of where the WSF can focus efforts when developing support and resources for families with CYP with WS. Many families spoke about teaching their young people about safe social interactions and broader social skills, and this was supported by the fact greater challenges in this area was predictive of greater social vulnerability (Chapter 6). Therefore, indicating that giving CYP with WS support in social cognitive skills is a valuable approach. The accounts of adults with WS (Chapter 3) also emphasise that adults have a hard time making trust evaluations and deciphering the intentions of others, therefore adults may benefit from guidance and support in this area. Currently, routes to support or intervention are lacking and, to my knowledge, the only support programmes are directed to adults with WS (Fisher, 2014; Fisher et al., 2022; Fisher & Morin, 2017). Most recently, Fisher and colleagues conducted an evaluation of a social skills training programme delivered over 8 weeks with teaching on topics including conversations, acquaintances versus friends, and social boundaries. They reported encouraging evidence, albeit preliminary, of its feasibility, acceptability, and effectiveness. Future studies should focus on designing support tools and programmes that are targeted and feasible with CYP with WS.

7.4.2. For Educators and Practitioners

The finding that social vulnerability is elevated in CYP is relevant to various people and organisations that support CYP with WS and their families – educators, health practitioners, and community groups. The formal school years emerged as salient across the thesis findings; therefore, it is important to raise awareness of social vulnerability amongst educators and other professionals who support a young person with WS. Educators and practitioners need to be aware that a young person with WS will likely need significant support beyond the academics, such as support with interpersonal skills, guidance on recognising manipulative behaviour and navigating social situations, and responding to peer

pressure. This is especially important because so few educators will work with more one child with WS and will likely not have received training specifically on WS[10]. The data from adults with WS, together with parent insights, tells us that negative peer experiences and bullying is common in the school environment. Therefore, educators need to be aware that aspects of the WS profile may place young people at risk of being bullied and there is a need to monitor the child's safety and wellbeing in school. Building on this, the adults' accounts in Chapter 3 highlighted that negative interactions often occurred due to a lack of understanding about differences in needs and, indeed, when asked about strategies to manage social vulnerability (Chapter 5), a few parents said they had given talks to the school about WS. Schools therefore should promote respect and anti-bullying campaigns, but also try to go further in fostering an understanding of diversity at an early age. The Learning About Neurodiversity at Schools (LEANS; Salvesen Mindroom Research Centre, 2023) programme is an excellent example of a whole-school approach to fostering a more inclusive and understanding school environment starting in the primary school years. In trying to support individuals with WS in their social relationships, practitioners can learn from the adults' accounts of strategies that have helped e.g., empowerment and self-advocacy skills.

The findings highlight the need for wider family support too. Parent and carers reported being very concerned about their CYP's social safety and consequently implement their own strategies to manage the social vulnerability concerns. For instance, parents endorsed high levels of close supervision, which no doubt impacts on the wellbeing of parents, siblings and wider family life in terms of family activities and participation. We know that child emotional and behavioural difficulties are strong predictors of parental distress in parents of children with rare genetic syndromes (Fitzgerald & Gallagher, 2022). In WS specifically, evidence suggests that parental stress can be high (Lanfranchi & Vianello, 2012; Papaeliou et al., 2012) and excessive sociability in CYP with WS is a common parent-reported challenge that distinguishes parents of CYP with WS (aged 4-19 years) from those with other neurogenetic syndromes (C. Reilly, Murtagh, et al., 2015). Support with issues of social vulnerability therefore needs to be targeted to the whole family unit. Additionally, if CYP with WS are being closely supervised during adolescence, it is important to consider the implications for independence, social participation and friendships. In the long-term, there is thus a need for practitioners to support parents and carers give their young person

[10] The Williams Syndrome Foundation (2023a) estimates that currently about 1 in 30 GPs ever meets someone with WS

opportunities for independence and to practice their social skills in a safe and supported environment at this stage of development.

7.4.3. For Policymakers

The findings from the interviews with adults with WS have implications for policy. I urge policymakers to recognise that people with WS get joy and meaning from having friends, but their social networks are often restricted to family relationships and staff members, which likely reflects the family as the main social support mechanism once an adult leaves formal education. In addition, we know that social vulnerability concerns present a barrier for families providing opportunities for their son/daughter to foster and maintain connections. Adults with WS need more opportunities to be part of a social group and develop a sense of connectedness. Adults with WS may face barriers to community participation in terms of their functional skills and therefore require additional support from professionals in this area (so that it doesn't all fall on family members).

7.4.4. For Researchers

Many of the thesis findings will be of interest to researchers in the field of WS and neurodevelopment more broadly, but I wish to highlight two methodological implications for future research. First, the cross-syndrome comparison findings raise intriguing questions about the design of studies with multiple groups. In Chapter 4 the approach taken to contrast groups side by side in the same study demonstrated that social phenotypes are best understood as distributed within and across groups. In other words, features of behaviour likely relevant to social vulnerability (i.e., qualities of eye contact and social interaction styles, but also EF difficulties and adaptive behaviour) cut across diagnostic groups. The results therefore challenge the idea of discrete diagnostic categories and syndrome-specificity, and add support to the growing evidence base that the clinical reality of neurodevelopmental profiles is one of overlapping phenotypes (Coghill & Sonuga-Barke, 2012). The results should encourage further debate about whether seeking syndrome-specificity is meaningful from a theoretical and applied perspective. A transdiagnostic approach to neurodevelopmental research has gained support from researchers in the field (Astle et al., 2021; Bryant et al., 2020; Fletcher-Watson, 2022; Mareva et al., 2019, 2023; Sonuga-Barke et al., 2016). It has also led to initiatives like Embracing Complexity, a coalition of over 60 organisations with a focus on neurodevelopmental conditions or mental health (including the WSF) to work towards a more united approach in recognising the

overlap of conditions (see reports Autistica, 2019; Sapiets, 2021). Inspired by Astle et al. (2021), I advocate that future work seeking to disentangle the factors underpinning social vulnerability would benefit from incorporating more transdiagnostic elements into the study design. This could take the form of recruiting participants based on diagnostic groupings, but running analyses trans diagnostically (i.e., diagnostic label is removed from the equation, termed 'diagnostic blind' in Astle et al. 2021). Studies taking this approach would capitalise on the larger sample size afforded by combining the data across participants (a common challenge associated with rare genetic syndrome research like WS and FXS) and draw more robust conclusions about features of behaviour and cognition that are shared across neurodevelopmental profiles and contribute to social vulnerability.

The second implication I propose is that researchers should build upon the self-report approach taken in Chapter 3 and strive to collect data on the thoughts and experiences of people with WS themselves, as a primary source of information. The voice of people with WS has been neglected in research to date and while there are obvious challenges in asking people with WS about abstract topics such as their own social vulnerability, the findings demonstrate the value of taking this approach to generate insights into social reasoning. I strongly recommend that WS researchers give thoughtful consideration to the methodology and embrace more creative, inclusive tools that have been used in the wider ID literature (Watt et al., 2010). An example is photovoice which is a qualitative research method that has potential to capture the voice of individuals who might face barriers in contributing their perspective via traditional interviews or surveys (Chinn & Balota, 2023). The use of photovoice in research with people with ID has been growing, but to my knowledge no WS study has used this as an inclusive tool.

7.5. Future Research

Social vulnerability is an important issue and the thesis findings inspire several avenues for future research. Ultimately, the goal should be to develop targeted support tools that can help support individuals with WS build and sustain healthy social interactions. To inform interventions, additional research is needed to better understand the factors that contribute to social vulnerability over development. However, before embarking on these larger scale studies, I encourage researchers to reconsider how we conceptualise and measure social vulnerability. Is the SVQ appropriate for use with CYP? To my knowledge, the SVQ was designed based on researcher input only and in the context of adulthood. It would be

valuable to use the SVQ as a tool to facilitate focus group discussion with parents and carers of CYP with WS and assess whether the SVQ fully captures social vulnerability as it presents in childhood and adolescence. Once tools have been validated, future studies should attempt to study social vulnerability using methods that do not depend solely on informant report. For instance, it would be valuable to collect parental reports alongside lab-based assessments of social behaviour or self-report using scenario-based assessments of vulnerability (e.g. similar to the approach used in Riby, Kirk, et al., 2014), to determine how informant reports relate to real-life social behaviour of CYP with WS.

The thesis correlates of social vulnerability highlighted in the thesis provide indications about underpinnings of high social vulnerability, which may inform intervention work. Future research should re-examine the links between social vulnerability and social reciprocity, EF, adaptive behaviour using direct behavioural assessments, but crucially also take account of intellectual ability. The regression model in Chapter 6 has provided a starting point for identifying relevant factors, but only accounted for approximately half of the variance in social vulnerability, therefore the question becomes, what other factors play a role in social vulnerability? Future research should consider other aspects of the behavioural and cognitive profile of WS. The investigation of factors related to social vulnerability was broad in scope and this was important as a first step in identifying relevant domains. However, a caveat is the lack of detail within each domain. For instance, social reciprocity appears to be important, but this is comprised of social awareness, social communication, social motivation and social cognition. Therefore, future studies should provide more detailed enquiry about the contribution of social reciprocity and tease apart the relative contribution of these components.

The issue of individual variability is an interesting one and not an area that was not meaningfully addressed in the thesis. Not everyone with WS will experience social vulnerability, therefore it would be interesting to consider protective factors in a subgroup of individuals with WS who score low on social vulnerability. Linked to this, it will be important to place greater emphasis on social vulnerability as a two-way interaction between personal and socio-environmental factors. Greenspan (2001) noted that when presented with a potentially adverse interaction such as a manipulative situation, it is the interaction between the personal competence factors and the situation itself that leads to either a successful or negative outcome. A greater focus on socio-environmental factors could produce interesting findings that account for more variation in social vulnerability. Further research could

usefully explore social vulnerability in relation to social participation / integration, social support and stability of relationships, to name just a few avenues of interest.

7.6. Concluding Remarks

The research findings presented in this thesis have advanced understanding of social vulnerability during childhood and adolescence in WS. The key takeaway message is that social vulnerability is a concerning feature of the WS social profile, that is elevated for many during this developmental stage, and requires support at the individual and family level. The findings on cognitive and behavioural predictors of social vulnerability have extended theoretical conceptualisations of social vulnerability and yielded interesting associations about underlying factors. Consequently, the thesis has provided new evidence about why some CYP with WS experience high social vulnerability which future studies should clarify with larger samples using multi-informant and multi-method data. Moreover, the findings indicate that heightened social vulnerability and behaviours relevant to social vulnerability may apply to CYP with neurodevelopmental conditions beyond WS. It is vital that these findings are developed in future research to clarify the pathways to social vulnerability and understand how best to support individuals with WS navigate social interactions safely.

Appendix A: Context About Covid-19 Mitigations During the PhD

Original Plans, Pre Pandemic

The experimental research, as funded, involved a multi-method, cross-syndrome design to understand pathways to social vulnerability in individuals with neurodevelopmental conditions. The proposed research aimed to further understanding of social vulnerability in people with neurodevelopmental conditions by:

- Employing innovative, ecologically valid, experimental methods to capture social decision-making in more naturalistic contexts. The most novel aspect of the experimental work was to develop an immersive virtual reality (VR) task to measure social approach behaviours (SAB). As described in Chapter 1, WS has been associated with an interesting profile of SAB, however research to date has relied on methods that fail to capture the complexities of real-life social interaction. I intended to use traditional SAB tasks (e.g., ratings tasks and the Stop-Distance Paradigm), but also develop a new VR approach/avoidance task to measure precisely how people with WS chose to engage with an avatar under different experimental conditions (e.g., direction of eye gaze, facial expression and age and gender of the avatar).
- Conducting an in-depth examination of the relationship between intellectual abilities/disabilities and levels of social vulnerability, using gold-standard standardised measures of cognition including the Wechsler Abbreviated Scale of Intelligence–Second Edition (WASI-II; Wechsler, 2011) to measure IQ, and the NEPSY–Second Edition (NEPSY-II; Korkman et al., 2007) to make a comprehensive assessment of executive functioning skills.
- Adopting a cross-syndrome approach throughout the research, focussing on the neurodevelopmental conditions WS and autism as the ideal candidate conditions by which to address routes to social vulnerability. While both groups may be at risk of heightened social vulnerability, the factors that impact on socially vulnerable outcomes may be different between groups.

Pandemic-Related Considerations

Most of the envisioned methods and tasks involved in-person, researcher-participant interaction. Therefore, when Covid-19 hit in March 2020, the accompanying strategies to

prevent the spread of the virus (i.e., social distancing measures, national lockdowns and school closures) had a substantial impact on my ability to conduct the experimental research as originally planned. During this time, some people with disabilities and serious underlying medical conditions were identified as being at a higher risk of becoming seriously ill from covid-19 infection (Centers for Disease Control and Prevention, 2021). Due to the serious heart conditions and hypertension associated with WS, people with WS were identified as being clinically extremely vulnerable as per 'Group 2' criteria of the guidance issued by the Royal College of Paediatrics and Child Health (2020). This meant that many individuals with WS and their wider family members were shielding to manage the risk of contracting the virus. Therefore, before adapting the research questions, methods and design, it was first important to consider the ethical and moral implications of asking CYP with WS and their families to take part in research at this time. Many families were going through an incredibly challenging time; with their son/daughter shielding, school closures for other siblings and juggling this alongside work / day-to-day life. Therefore, as a researcher my ethical responsibility was to avoid adding additional stress and burden to the lives of these families. Consequently, one of the first decisions made was to pause the research.

Instead, focus shifted to trying to identify new ways by which to answer the original research questions. This involved extensive information-gathering on alternative ways of collecting data, keeping up to date with wider discussions within the field and culminated in an extensive ethical amendment. In October 2020, with support from my supervisory team, I considered it an appropriate time to restart the research, following in the footsteps of many other child development labs in the UK. At this time, many CYP with WS were still shielding or being home-schooled, and face-to-face research with external participants was still not permitted. Consequently, a priority in redesigning the experimental research, was to develop a protocol that did not require any researcher–participant interaction and could withstand an extended period of socially distanced data collection.

Many research groups tried to eliminate or minimise the need for face-to-face interaction, by opting for online data collection (Omary et al., 2020). While a plethora of psychological research has capitalised on advancements in remote technology and sophisticated software for hosting research online (Lefever et al., 2007), shifting to online is no simple feat in the case of child development / neurodevelopmental research. The field has a long-standing tradition of face-to-face testing in university laboratories, schools, the home environment and at science festivals. A key reason for this is that the majority of commonly

used neuropsychological measures in neurodevelopmental studies require face-to-face delivery between a researcher and participant. However, remote assessment is a fundamental deviation from the standardised protocols, which in turn threatens measurement reliability and validity (The British Psychological Society, 2020b). The two standardised cognitive batteries in the proposed research—the WASI-II (Wechsler, 2011) and the NEPSY-II (Korkman et al., 2007) —both involve highly structured interpersonal procedures and are comprised of subtests which require participants to manipulate the test materials, for example using on a block design task. Consequently, it was not methodologically appropriate or practically possible to replicate via teleconferencing software.

A further challenge for studies like mine that involved data collection with CYP with neurodevelopmental conditions, is that the intellectual disabilities and other cognitive difficulties often associated with neurodevelopmental diagnoses, can make virtual assessment/tasks particularly challenging (The British Psychological Society, 2020b). For example, attentional difficulties are a common feature across a range of neurodevelopmental conditions. Therefore, the traditional face-to-face approach is vital in enabling the researcher to monitor participant engagement levels, judge when a break is needed and support the participant to refocus (M. Ashworth et al., 2021). Remote sessions can also make it more difficult to form researcher-participant rapport, which is important in the context of high rates of anxiety associated with WS. Despite the well-founded reliance on face-to-face protocols, online remote methods with individuals with clinical and neurodevelopmental needs, were showing promise even before the pandemic, with diagnostic assessment (“telehealth”) and treatment/intervention moving in this direction (Adamou et al., 2021).

Actions Taken to Adapt the Research

In revising the experimental protocol for remote methods, the first stage was identifying which tasks could reasonably and appropriately be conducted using a teleconferencing session with CYP. Tasks which required the use of sophisticated laboratory equipment or intended to capture “live” social behaviour were removed (VR and eye tracking). Cognitive assessments which could not be administered without deviating significantly from standardised procedures were removed and alternative measure(s) of intellectual ability / cognition were researched. The alternative measure(s) needed to be (1) digitised or amenable to digital, remote administration with small modifications (2) appropriate for use with children/young people with neurodevelopmental / intellectual

conditions (3) relatively quick to administer and (4) ideally standardised norms. One key concern in administering the assessment remotely with CYP with WS was whether it would be feasible and conducive to collecting meaningful data. At this time there was very little evidence on how remote psychological assessment compared to the traditional face-to-face procedure. However, one timely and highly relevant paper by Ashworth et al. (2021) provided promising initial evidence of the feasibility and appropriately of conducting cognitive assessments online with children with WS. The researchers compared scores on the British Picture Vocabulary Scales (Dunn et al., 2009) and the Ravens Coloured Progressive Matrices with one group of children with WS assessed online (n=14) and another face-to-face (RCPM n=12; BPVS n=24). They found that online format produced comparable RCPM scores, but BPVS scores were higher when administered online. The possibilities of remote methods was also acknowledged by several professional boards, such as the American Psychological Association (2020) and The British Psychological Society (BPS; 2020a, 2020b), who issued guidance about delivering psychological assessments under physical distancing constraints. The guidance was key in shaping my decision-making.

The actions that were taken to adapt the research included (1) streamlining the research i.e., removing tasks like VR and eye tracking that could not be delivered online (2) identifying paper-based measures that could yield standardised data on cognition and behaviour (3) developing a new social approach task that could be delivered online via remote research sessions with CYP, alongside measures of cognition suitable for online delivery (BPVS adapted with permission from the publisher and the Ravens-2 designed for one-screen administration).

Appendix B: Chapter 3 Semi-structured Interview Schedule

Hello! This chat is to find out about your experiences of having friends. I have some questions that I will ask you. You can give as much information as you wish.
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. I will be recording the interview using this device. This is so that I can type up what we talk about.

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?**
   how many close friends they had,
   how often they saw them,
   how they formed the friendships,
   what they did together,
   did you find it easy to make friends

2. **Can you tell me a bit about your friends now?**
   are they the same friends that you had at school or different?
   how often they see them,
   what they do together,
   do you use the internet to keep in touch with friends?
   are you happy with the friends you have now?

3. **What does being a good friend mean to you?**
   how do you know when someone is your friend?
   do you have some friends who are more important than others?

4. **Is it important to you to keep the same friends?**
   how do you make sure you keep friends?

5. **Can you tell me some good things about your friends?**
   do your friends help you out?

6. **Can you tell me some difficult things about your friends?**
   Do you and your friends ever argue / fall out and what about?

7. **Can you tell me about a time when you had a lot of fun with your friends, or when something good happened with them?**
   how did you feel

8. **Can you tell me about a time when something bad happened with a friend, or when your friends did something you didn't like?**

9. **How do you feel about meeting new people?**
   When you meet someone for the first time (i.e. someone you don't know), what do you do?

10. **Have you ever had not-so-good friends, or people who pretended to be your friend?**
    how did you know?
    How did you feel?)

11. **Can you tell when someone else is lying to you or trying to trick you?**
    Can you give me an example?

12. **Is everyone a friend?**
    How do you know if you can trust someone?

Appendix C: Chapter 5: Demographics Questionnaire

This questionnaire asks for information about you and about your child. If you have any questions as you go through please do not hesitate to contact the leader researcher, Ellen, via email (ellen.ridley@durham.ac.uk) or telephone on XXX

Date of completion: _ _ / _ _ / _ _ _ _

Part 1 – About you (parent / caregiver)

What is your date of birth? _ _ / _ _ / _ _ _ _

Which gender do you identify with?

☐ Male
☐ Female
☐ Other _____

What is your relationship with the child you are answering about?

☐ Birth parent
☐ Foster parent
☐ Adoptive parent
☐ Step-parent
☐ Other, please state your relationship _____

Do you currently live with the child you are answering about?

☐ Yes
☐ No

Do you have a diagnosis of a developmental condition or mental health condition given by a clinician?

☐ Yes, please specify

_____

☐ No

What is your highest level of qualification?

☐ No formal qualifications
☐ High school / secondary school level qualifications
☐ Further vocational qualifications
☐ University undergraduate level qualification (BA, BSc etc.)
☐ University post-graduate level qualification (MA, MSc, PhD, Certificate, etc.)

Part 2 – About your child

What is your child's date of birth? _ _ / _ _ / _ _ _ _

Which gender does your child currently identify with?

- ☐ Male
- ☐ Female
- ☐ Non-binary
- ☐ Don't know

What type of school or college does your child currently attend?

- ☐ Mainstream school / college
- ☐ School/college for children with special educational needs
- ☐ Special educational unit within a mainstream school/college
- ☐ My child is home schooled
- ☐ My child is not currently attending school/college
- ☐ Other, please specify _____

Has your child ever had a statement of special educational needs or an education, health and care plan?

- ☐ Yes
- ☐ No
- ☐ Do not know

Has your child ever been diagnosed with any of the following conditions <u>by a clinician</u>? Please tick all that apply.

- ☐ Anxiety Disorder
- ☐ Attentional deficit hyperactivity disorder (ADHD) / Attentional deficit disorder (ADD)
- ☐ Autism spectrum condition (Asperger Syndrome/Autism/PDD-NOS)
- ☐ Bipolar disorder
- ☐ Conduct disorder
- ☐ Depression
- ☐ Dyslexia
- ☐ Dyspraxia/Developmental coordination disorder
- ☐ Eating disorder
- ☐ Intellectual disability
- ☐ Generalised anxiety disorder
- ☐ Language delay
- ☐ Obsessive compulsive disorder (OCD)
- ☐ Oppositional defiant disorder (ODD)
- ☐ Panic disorder
- ☐ Personality disorder
- ☐ Post-traumatic stress disorder
- ☐ Sensory processing disorder
- ☐ Social phobia/social anxiety disorder
- ☐ Specific phobia
- ☐ Tourette syndrome / Tic disorder
- ☐ Schizophrenia/psychosis

Do you <u>suspect</u> that your child has (or has had) any condition that they <u>have not</u> been diagnosed with?

☐ Yes
☐ No

Which condition(s) do you suspect that your child has (or has had)? Please tick all that apply

☐ Anxiety Disorder
☐ Attentional deficit hyperactivity disorder (ADHD) / Attentional deficit disorder (ADD)
☐ Autism spectrum condition (Asperger Syndrome/Autism/PDD-NOS)
☐ Bipolar disorder
☐ Conduct disorder
☐ Depression
☐ Dyslexia
☐ Dyspraxia/Developmental coordination disorder
☐ Eating disorder
☐ Intellectual disability
☐ Generalised anxiety disorder
☐ Language delay
☐ Obsessive compulsive disorder (OCD)
☐ Oppositional defiant disorder (ODD)
☐ Panic disorder
☐ Personality disorder
☐ Post-traumatic stress disorder
☐ Sensory processing disorder
☐ Social phobia/social anxiety disorder
☐ Specific phobia
☐ Tourette syndrome / Tic disorder
☐ Schizophrenia/psychosis

Was your child's diagnosis of Williams syndrome confirmed with genetic testing?

☐ Yes
☐ No

Any comments please leave here

_____

_____

_____

Has your child ever been diagnosed with any other mental health or developmental condition(s) (not including Williams syndrome)?

☐ Yes, please state your child's diagnosis

_____

☐ No

How old was your child when they were diagnosed with Williams syndrome?

_____years, ___months

Do any of your child's other relatives have a diagnosis of a developmental condition? Please tick all that apply.

- ☐ Their other parent (not yourself)
- ☐ Their grandparent
- ☐ Their brother and/or sister
- ☐ Their aunt and/or uncle
- ☐ Their cousin
- ☐ Other, please specify

End of questionnaire. Thank you.

Appendix D: Chapter 5: Experience of Covid-19 Questionnaire

The following questions/statements ask about child's behaviour in the context of COVID-19 (please reflect over the past year or so, including periods of lockdown/school closures etc.)

1. **Is your child aware of covid-19?**
   ☐ Yes
   ☐ No

2. **What impact has covid-19 had on your child's anxiety?**
   ☐ Anxiety was not an issue before covid-19 and that's still the case
   ☐ Anxiety was an issue before covid-19 and this has stayed the same
   ☐ Anxiety levels have increased somewhat
   ☐ Anxiety levels have increased significantly

3. **My child has been anxious / is currently anxious about not being able to see and meet up with friends in the ways they are used to**
   ☐ Definitely disagree
   ☐ Slightly disagree
   ☐ Slightly agree
   ☐ Definitely agree

4. **My child is <u>aware</u> that they need to keep their distance from other people (i.e. social distancing)?**
   ☐ Definitely disagree
   ☐ Slightly disagree
   ☐ Slightly agree
   ☐ Definitely agree

5. **My child has been anxious or is currently anxious about keeping their distance from other people (i.e. social distancing)**
   ☐ Definitely disagree
   ☐ Slightly disagree
   ☐ Slightly agree
   ☐ Definitely agree

6. **My child appropriately keeps their distance from other people when we are out and about (e.g. not approaching people and standing at the required distance)**
   ☐ Definitely disagree
   ☐ Slightly disagree
   ☐ Slightly agree
   ☐ Definitely agree

7. **How concerned are you about the impact of covid-19 on your child's friendships? (e.g. maintaining their friendships)**
   ☐ Not at all concerned
   ☐ Somewhat concerned
   ☐ Very concerned
   ☐ Extremely concerned

8. **How concerned are you about the impact of covid-19 on your child's emotional wellbeing?**
   ☐ Not at all concerned
   ☐ Somewhat concerned
   ☐ Very concerned
   ☐ Extremely concerned

9. **Please feel free to leave further information about the impact of covid-19 on your child's anxiety, friendships and social approach behaviours, or any other information you think may be relevant. Thank you.**

_____
_____
_____
_____
_____
_____
_____
_____

End of questionnaire. Thank you.

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