Exploring the prevalence and phenomenology of repetitive behaviours and abnormal sensory processing in children with Williams syndrome

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Abstract

Background: A small amount of research with individuals who have Williams Syndrome (WS) suggests that children with the condition may be vulnerable to sensory processing abnormalities and present with repetitive and restricted behaviours.

Methods: Parents of twenty-one children with WS aged 6-15 years completed a semi-structured interview designed to elicit the form, frequency, impact and developmental course of a range of sensory processing abnormalities and repetitive behaviours.

Results: Findings indicate that sensory processing difficulties are predominantly characterised by hypersensitivities, particularly in relation to vestibular, auditory, gustatory and proprioceptive functioning. Parents also reported the presence of a range of restricted and repetitive behaviours, which were often associated with their child’s sensory symptoms.

Conclusions: This study makes a significant contribution to our understanding of sensory functioning and repetitive behaviours in WS. It also highlights the need for a multidisciplinary assessment of the difficulties experienced by children with the disorder.

Keywords: Williams syndrome, sensory processing abnormalities, repetitive and restricted behaviours
Exploring the prevalence and phenomenology of repetitive behaviours and abnormal sensory processing in children with Williams syndrome

1. Introduction

Williams Syndrome (WS) is a neurodevelopmental disorder caused by the microdeletion of approximately 25-28 genes on chromosome 7q11.23 (Blomberg, Rosander, & Andersson, 2006). It occurs in approximately 1 in every 20,000 births (Korenberg, Bellugi, Salandanan, Mills, & Reiss, 2003), and is most often characterised by craniofacial and cardiac abnormalities, hypercalcemia, a distinctive “hypersociable” personality, and cognitive, social, and behavioural difficulties (cf. Martens, Wilson, & Reutens, 2008). Although there is considerable variation within WS, individuals generally function within the mild to moderate range of intellectual disability (e.g. Searcy et al., 2004), and often possess relative strengths in verbal ability and auditory rote memory, with comparative deficits in visuospatial construction (Mervis, et al., 2000). Further difficulties include impulsivity and attention difficulties (e.g. Rhodes, Riby, Matthews, & Coghill, 2011), sensory processing impairment, and repetitive or obsessive behaviours (Semel & Rosner, 2003). In relation to the emotional and psychological characteristics of WS, research has consistently identified high levels of anxiety and fear in many individuals with WS, in comparison to other developmental groups (e.g. Dykens, 2003).

Sensory processing can be defined as “the way that sensory information e.g. visual, auditory, vestibular, or proprioceptive stimuli is managed in the cerebral cortex and brainstem for the purpose of enabling adaptive responses to the environment” (Baker, Lane, Angley, & Young,
In typical development and developmental disability there is wide variation in the way individuals process sensory information and interact with their physical and social environment, with particular profiles of sensory processing being associated with difficulties with daily functioning and behavioural and emotional problems (Ben-Sasson et al., 2008; Liss, Mailloux, & Erchull, 2008). Within WS relatively little is known about sensory processing difficulties (John & Mervis, 2010). Semel and Rosner (2003) report significant impairments in visual, auditory, and tactile areas of perception in WS, and John and Mervis (2010) found children with WS to have abnormalities in sensory modulation; including auditory, vestibular, and proprioceptive hyper- and hypo-sensitivity. Anxiety in individuals with WS has often been related to the problems they experience in auditory hypersensitivity and tactile defensiveness (Blomberg, et al., 2006; Dilts, Morris, & Leonard, 1990; Gallo, Klein-Tasman, Gaffrey, & Curran, 2008; Gosch & Pankau, 1994; Levitin, et al., 2005; Semel & Rosner, 2003; Udwin et al., 1987) indicating that a clearer understanding of the nature of sensory processing difficulties in the disorder may have significant downstream implications for our understanding of the aetiology of mental health difficulties in this group.

Repetitive behaviours (RRB) can be defined as “repetitive, non-functional activities or interests that occur regularly and interfere with daily functioning” (Gabriels et al., 2005, p. 170). Within WS, RRB have been reported in up to 86% of individuals (Davies, et al., 1998). Semel and Rosner (2003) state that individuals with WS are likely to engage in many obsessive-compulsive behaviours, such as the compulsive need to identify the source of sudden noises, compulsive greeting behaviours, and so on. Similar to that observed within other developmental groups (e.g. autism), a relationship between RRB and anxiety has been suggested. Semel & Rosner (2003) propose that the compulsive forms of behaviour observed
in WS may serve to reduce anxiety. However, at present there is very little evidence to support this suggestion in WS.

Some studies have begun to investigate a relationship between sensory processing and RRB. Chen, Rodgers, and McConachie (2009) found children with autism who demonstrated more RRB, also exhibited significantly more sensory processing difficulties; in particular, related to tactile, visual, and auditory hypersensitivity. Similar findings have been reported by Gabriels, Agnew, Goldson, Ledbetter, and Cuccaro (2005). Although to date this relationship has not been as extensively explored within WS, similarities between WS and other neurodevelopmental groups, as discussed, support the need for further investigation.

In a recent study, Baker et al. (2008) found strong correlations between sensory processing abnormalities and maladaptive behaviour (believed to constitute mainly RRB) and between sensory processing difficulties and anxiety in children with autism spectrum disorders (ASD). These results suggest greater sensory processing difficulties may be associated with higher levels of RRB and anxiety in this group. Joosten, Bundy, and Einfeld (2009) examined the extrinsic and intrinsic motivators for stereotyped and repetitive behaviours in children with ASD and intellectual disability (ID) and those with ID without ASD. They reported both anxiety and sensory seeking to be intrinsic motivators for RRB in children with ASD and ID, with anxiety as the strongest motivator. In those without ASD, sensory seeking was the strongest motivator. This suggests within ASD there may be a specific and atypical motivator for the presence of RRB.

These potential relationships have begun to be explored in WS. Based on parental report questionnaires Riby, Janes and Rodgers (2012) report a significant correlation between
sensory processing abnormalities and RRB in a sample of children with WS, mirroring the relationship reported in ASD. No comparable relationship was found between RRB and anxiety in WS (Rodgers, Riby, Janes, Connolly & McConachie, 2012) indicating that at least through the use of questionnaire measures the patterns of relationships and interactions across these developmental disorders may differ. However, the questionnaire measures utilised have not been developed specifically for use with families with a child who has WS and as such may not capture the syndrome-specific nature of the relationship between RRB and sensory processing in this population. Given this, it was felt that these relationships need further and more detailed exploration within WS where we have less evidence than is currently available from our explorations of ASD.

This emerging body of research indicates that there are potential relationships between sensory processing difficulties and RRB in WS which warrant further exploration and therefore provide a clear rationale for the study. Furthermore much of the work exploring these to date is based on data from questionnaire completion by parents and whilst valuable to enable relationships to be identified, such data have limitations. These measures were not developed for, nor have they been validated with families with a child who has WS and as such important aspects of the phenomenology of these experiences in WS may not be captured by the use of these standardised techniques. Therefore the rationale and aim for the current work was to use a different method of capturing information about sensory processing experiences and RRBs from parents of children with WS, namely a semi-structured interview. It was hoped that utilising this method would enable parents to identify aspects of their day to day experiences not captured by the standardised measures, to provide information about developmental changes associated with these phenomena, and to describe more fully the impact that these characteristics have on their everyday lives.
2. Methodology

2.1 Recruitment

Participants with WS were identified and recruited through the Williams Syndrome Foundation (WSF), and were located throughout the North of England and Scotland. A total of 29 participants were contacted by letter and asked to participate in the study; 21 participants consented to take part, one person did not consent, and seven did not respond. This gave a total consent rate of 72%. Participants were included if they had a diagnosis of WS (previously confirmed through positive genetic fluorescent in situ hybridisation testing), if they were aged between 5- and 15-years-old, and if they were members of the WSF. A favourable ethical opinion was granted by Newcastle University Faculty of Medical Sciences Ethics Committee.

2.2 Sample

Participants were 21 children with WS aged between 6 and 15 years (mean age 9.3 years; 12 male, 9 female). The mean estimated Full Scale IQ (FSIQ) was 52.6 (sd 11.3, range 40-83), which is within the range reported as being typical for WS (cf. Mervis, et al., 2000). Mean Verbal IQ (VIQ) was 59.5 (sd 14.2, range 46-98) and mean performance IQ (PIQ) was 54.0 (sd 7.7, range 46-73). This discrepancy between verbal and performance IQ mirrors the widely reported pattern of dissociation between the two domains of functioning (cf. Martens et al., 2008). Nine of the children were reported to be taking medication, although in no cases was this for behavioural or mental health difficulties. In the majority of cases medication was
related to stomach or bowel disorders or sleep difficulties. Further data for this sample are reported by Riby et al. (2012).

2.3 Measures

Sensory Processing

Sensory Profile – Short Form (SSP; Dunn, 1999)

The SSP is a 38-item parent-report questionnaire asking parents to rate the frequency with which their child displays a number of sensory behaviours on a five-point scale (always, frequently, occasionally, seldom, or never) (Dunn, 1999). There are seven subscales; Tactile Sensitivity, Taste/Smell Sensitivity, Movement Sensitivity, Under-responsive/Seeks Sensation, Auditory Filtering, Low Energy/Weak, and Visual/Auditory Sensitivity. A total overall behaviour score was used for the main analyses. A lower-score on this measure is indicative of greater impairment. The SSP has shown to have good internal consistency for the various subscales (Cronbach’s alpha = .47 – .91), and has established content validity and strong inter-rater reliability (Dunn, 1999). Studies have reported that the SSP has discriminate validity of >95% in identifying children with and without sensory modulation difficulties (McIntosh, Miller, & Shyu & Dunn 1999). It has been recommended as a good measure for research protocols (Dunn, 1999). The SSP was used here to provide an indication of the general level of sensory processing of the sample in comparison to published norms.


The SRAF-SSI is a 29-item parent-report interview. It was developed by the authors to explore a range of experiences in families with a child with WS, including anxiety, fear, RRB
and sensory processing difficulties. Those data relating specifically to sensory processing abnormalities and RRB will be reported here.

The SRAF-SSI was constructed by using a number of resources such as the Autism Diagnostic Interview (Lord, Rutter & Le Couteur, 1994) and literature on sensory processing (e.g. Bogdashina, 2003; Smith-Myles, Tapscott-Cook, Miller, Rinner, & Robbins, 2000). Other questions were developed by the author in relation to gaps in current measures which corresponded to points of interest in WS.

The SRAF-SSI begins with questions related to sensory processing and covers seven different aspects of the phenomena (tactile hypersensitivity & hyposensitivity; vestibular hypersensitivity & hyposensitivity; proprioception, visual hypersensitivity & hyposensitivity; auditory hypersensitivity & hyposensitivity, gustatory features and olfactory features). The interview continues with a section relating to RRB. The focus of this element of the interview was higher order repetitive interests (Turner 1997) and included questions related to unusual interests (e.g. parts of objects), circumscribed interests/hobbies (e.g. timetables etc.). Whilst a large number of standardised measures of RRB are currently in use (See Honey Rodgers & McConachie, 2012, for a review) none of these measures have been developed for and validated with individuals who have WS. As such the development of an SSI would allow us to capture some of the rich detail which may be specific to WS that would potentially not be captured by a tool developed for a different population.

For each question the interview began with a description of the target behaviour. Parents were then asked to identify whether this applied to their child and if so to describe in what
They were then asked to consider the previous three months and indicate on a five point likert scale (not at all to always) how often this behaviour was shown (frequency) and how much the behaviour impacted on family life (intensity). They were then asked to rate on a five point likert scale (much better now to much worse now) how much this behaviour might have changed throughout development. At the end of each section parents were provided with the opportunity to report any other sensory or repetitive behaviours that they believed it would be important to capture. For the RRB questions, parents were also provided with the opportunity to report what seemed to influence whether their child was more or less likely to engage in RRB and when these behaviours were most or least frequent. The interview schedule therefore yielded a mixture of quantitative and descriptive information.

2.4 Procedure

The researcher met with the parent(s) to carry out the SRAF-SSI (approximately 45 minutes to complete), and with the child to carry out the WISC-III Short Form (Wechsler, 1991; approximately 20 minutes to complete). The SSP was posted to parents to complete in advance of the meeting. Families were interviewed in their own homes. For 14 families the mother was interviewed alone. For the remaining 7 families both parents were interviewed together.

3. Results

Sensory processing was measured using the SSP. The SSP allows categorisation of participants into three categories based on their total score. The categories are: typical performance, probable difference and definite difference. The mean score fell within the ‘definite difference’ range, indicating that the majority of the sample experienced sensory processing difficulties (M= 124.3 SD 21.4; definite difference range: scores of 38-141). This
indicates that for participants in the present study significant degree of sensory processing difficulties were present.

Using methods derived from thematic analysis items on the SRAF-SSI were summarised according to their frequency, the impact on family life and in relation to any changes in that behaviour over time (See Table One). The data are described below in relation to these features and where the behaviour was reported to be present for the majority of the sample (the most frequent endorsement being for often/always) the responses to the interview items are illustrated with quotes.

[Insert Table One]

3.1 Sensory Processing

3.1.1 Vestibular hypersensitivity

Parents were introduced to this theme with the following description ‘I will ask you about whether _________ is over-sensitive to their body movements. If this is the case people often have low tolerance for activities that involve movement, they may have difficulty changing direction and speed, or may have difficulty maintaining a body position other than upright with feet on the floor.’

Eleven parents reported that their child was over-sensitive to their own body movements. Five parents reported this to be an occasional difficulty. Six of these parents reported that the presence of vestibular sensitivity impacted on family life ‘often’ or ‘always’. Changes over time were reported. Thirteen families reported improvements over time.

‘He has difficulty changing direction and maintaining body position…’ (age 15)
'She struggles with balance... if we take her somewhere busy and there are children running around she will have to stop and get her bearings and her balance is worse' (age 10)

'She has a lot of problems [going] up and down stairs' (age 12)

'When he is running along he has difficulty in changing speed and direction, he finds it difficult on his bike and rocks and wobbles...he can’t get his balance' (age 8)

3.1.2 Proprioceptive hypersensitivity

Parents were introduced to this theme with the following description ‘The next area that I’m interested in is the ability to sense the position, orientation, and movement of the body and the different parts of the body that helps us to move, sit, hold items, and balance. A person may have poor posture and may slump. They may have difficulty climbing stairs, following directions such as left and right, difficulty with PE at school, and may miss their mouth cleaning teeth, etc.’

Fourteen parents reported that their child experienced difficulties with proprioceptive hypersensitivity and 8 parents reported that this impacted often or always on family life. Eleven parents reported improvement over time, and 9 reported no change.

The type of behaviours reported here included:

‘We still have to clean his teeth for him’ (aged 8)

‘He finds writing difficult’ (aged 9)

‘She falls over a lot and breaks things...she drops everything, and she finds it difficult to use a knife and fork’ (age 7)

‘He has difficulty brushing his teeth, feeding himself, especially soup...he gets covered in it...he goes up and down stairs on all fours and holding the banister...he likes to join in with activities like ball games but finds catching a ball really difficult’ (age 6)
3.1.3 Auditory hypersensitivity

Parents were introduced to this theme with the following description ‘Some people show over-sensitivity to sounds – a person may respond negatively to loud noises. Sometimes noises can be painful and cause distress. Even small noises can be uncomfortable and some children can’t concentrate in the presence of noise, such as background noise.’

Fourteen parents reported that their child was over-sensitive to auditory stimuli ‘often’ or ‘always’. Ten parents reported that the behaviour impacted ‘often’ or ‘always’ on family life. Nine parents indicated improvement over time, 9 parents reported that the behaviour had not changed with development, whereas 3 parents reported worsening over time.

‘He doesn’t like loud noises, especially unexpected loud bangs’ (age 15)

‘He doesn’t like unexpected sounds...he covers his ears, cries, gets sweaty palms, has palpitations, gets shaky...any fire alarms, small children and babies’ (age 6)

‘She is petrified of noises – she never used to be and now over the last two years it has developed. She is scared of the hoover, drill.... She doesn’t like the hairdryer, or the doorbell. She has to check everything is OK.’ (age 7)

‘She gets frightened by sounds and puts her hands over her ears. But she likes music. She [has] developed a tolerance of some sounds. She doesn’t like the sound of fireworks, motorbikes, drills, mechanical noises, popping balloons, bottles, lawnmowers. Sometimes she doesn’t want to leave the house.’ (age 6)

3.1.4 Gustatory hypersensitivity

Parents were introduced to this theme with the following description ‘Some people may avoid certain foods and only eat certain foods with certain tastes. They may often be described as a
picky eater. Would you describe ___________in this way? Can you give some examples? Have they always been like this? Do they eat/like to taste any unusual substances (i.e. PICA)?’

Sixteen parents reported that their child had oversensitivity to certain foods/tastes. Twelve of these parents reported that this impacted on family life ‘often’ or always’. Five families reported improvement in this area, 9 reported no change and 7 reported worsening of this feature over time.

‘She is a very picky eater. She won’t eat bread, only homemade bread or French stick. She won’t eat toast or sandwiches or try new things’ (age 10)

‘She finds liquids to solids difficult’ (age 7)

‘He doesn’t like sticky or chewy things…. he is anxious about new foods. When he was younger he ate better than he does now’ (age 15)

‘He is obsessive about foods. It is worse now than when he was younger’ (age 13)

‘He likes "pink chicken" which is what he refers to meaning ham or gammon…he only likes one type of steak pie - he nearly made himself sick when we bought a different one once’ (age 7)

3.1.5 Unaffected aspects of sensory processing

There are also a number of areas of sensory processing abnormalities which were not described as a feature of their child’s every day life by the majority of parents in the sample, including Tactile hyper- & hypo- sensitivity, Vestibular hyposensitivity, Proprioceptive hyposensitivity, Visual hyper- & hypo- sensitivity, Auditory hyposensitivity, Gustatory hyposensitivity and Olfactory hyper- & hypo sensitivity.

3.2 Repetitive Behaviours
3.2.1 Overall Repetitive Behaviours

Parents were introduced to this theme with the following description ‘We are now going to move on to some of the ways in which they might behave. I am especially interested in behaviours that may appear to be repetitive. Repetitive behaviours can be defined as “repetitive, activities or interests that don’t appear to have a purpose that occur regularly and interfere with daily life”. There are lots of different types of repetitive behaviours, such as repetitive use of objects or interest in parts of an object, compulsions and rituals, difficulties with changes in routine or environment, unusual attachment to objects, hand, finger or body mannerisms, and so on.’

Eighteen parents reported RRB to be present ‘often’ or ‘always’ for their child. Thirteen parents reported that the presence of RRB had an impact on family life. Eight parents thought that the presence of these behaviours had remained stable over time and 11 thought that they had worsened.

‘She bounces her ball repeatedly, all day long’ (age 6)

‘She loves to look at faces a lot and gets frustrated if she can’t’ (age 7)

‘He repeatedly bangs his knees together’ (age 14)

‘She doesn’t like changes in routine. Her speech is often very repetitive’ (age 7)

‘He takes toys apart and looks at how they work repetitively. He is also repetitive in his language - he seeks reassurance saying "have I been good?" or "what are we doing next?"’ (age 8 years)
‘He likes to switch lights on and off in the hall and upstairs. He gets up in the night and turns lights on and off. We have to sleep with him now as he switches all the lights on in the house’ (age 7)

‘He likes anything that goes up and down like barriers in a car park - he will sit and watch them for hours’ (age 7)

3.2.2 Unusual interests

Parents were introduced to this theme with the following description ‘I would like to ask you about some repetitive behaviours which were not covered in sufficient detail on the questionnaires you completed. Some children may have unusual or peculiar interests – interests that preoccupy them even when the focus of the interest is not physically present and that might seem odd to other people. For example, metal objects, traffic lights, street signs or toilets. Does ________ have any unusual or peculiar interests? How much does he/she talk about them? Does this interest influence how he/she behaves?’

Fourteen parents reported that their child had unusual interests. Nine parents reported that these unusual interests impacted ‘often’ or ‘always’ on family life. Twelve parents reported that the presence of these behaviours had been stable over time, whereas 8 parents felt that the impact was worse now than previously.

‘Lawnmowers, car washes, hair, food blenders, circuses, helicopters, army cadets’ (age 15)

‘Wheels on cars, he sits outside and watches them for hours’ (age 14)

‘….dangling things – she likes wool, glittering things, wire or thread’ (age 13)

‘Electric fans and things that spin’ (age 15)
‘He also loves hoovers - especially Henry Hoover...he will ask people randomly what sort of hoover they have got. He likes to dismantle them. When he was in ikea once he saw a hoover, took off and dismantled it in the shop’ (age 6)

3.2.3 Special hobbies/circumscribed interests

Parents were introduced to this theme with the following description ‘Some children also have special hobbies or interests which differ from ordinary hobbies in terms of their intensity. For example, some children have strong interests in computer games, trains, mechanics, etc. Often they may involve a high level of expertise or knowledge, they may be unusually focused, they may tend to be non-social activities (not shared with another individual), etc. Does __________ have any special hobbies/interests that are unusual in their intensity? In what way is it unusual? Does he/she share the interest with other people? In what way? What happens if you interrupt him/her? Does it interfere with his/her doing things?’

Special hobbies or interests were reported by 17 parents, with 9 of these parents indicating that the presence of these interests impacted on family life. 12 parents reported stability in this characteristic over time, whereas for 8 families the impact had worsened over time.

‘Bagpipes.... He knows all about them. The army, he knows a lot about the army, he doesn’t really notice if you’re interested just keeps on talking’ (age 15)

‘Computer games, talking about cars. He used to be obsessed by Thomas the Tank Engine’ (age 8)

‘Diggers and excavators – he has very high expertise’ (age 15)

‘Pieces of music and emotional parts of films – she gets obsessed with them. She feels music very intensely. It’s hard to get her off the subject’. (age 13)
'He is obsessive about Michael Jackson and has a lot of specialist knowledge about him. When he died on TV, he was fascinated - he asked a lot of questions about death... ' (age 7)

3.2.4 Triggers to Repetitive Behaviours

Parents were asked to reflect upon what might trigger the onset of a RRB. The majority view of those parents who responded to this item was that anxiety was a trigger for the onset of RRB. Some parents also indicated that illness may trigger RRB.

‘When he is anxious he will constantly go on about things – his obsessions’ (age 15)

‘When she is poorly she pulls down her hair repeatedly and ends up making herself worse but I think she is trying to soothe herself’ (age 10)

‘When she is ill’ (age 10)

‘When she is anxious it makes them worse’ (age 13)

‘If going somewhere unknown or she is worried about something she will do them more’ (age 12)

‘Anxiety increased the frequency as he uses them as a comfort’ (age 15)

‘She has shaky legs and shakes them up and down repeatedly when she is anxious’ (age 7)

‘If he doesn’t know what’s happening next or if he knows something is happening like swimming on a Friday, he will ask from Monday about it’ (age 8)

3.3 The relationship between sensory processing abnormalities and repetitive behaviours

During the interview a number of parents spontaneously described a relationship between their child’s sensory processing difficulties and their repetitive behaviours. In the instances when this occurred parents described aspects of the auditory environment which had initially been aversive for the child which over time became the source of fascination.
‘Vacuum cleaners [are a special interest], but when he was little he didn’t like the noise’ (age 15)

‘Sirens, when they are not present he talks about them a lot. Motorbikes are another thing. He used to be afraid of the noises….now he loves them’ (age 13)

‘He is attracted to some sounds though even though he doesn’t like them - like the hoover he has a hoover obsession. It’s like he takes an interest in an item and like he wants to understand it. He was completely against hand driers – scared of the noise – couldn’t get him out of the toilet the other time as he wanted to stay with the hand drier’ (age 6)

‘Any fire alarms...he can’t be in school when they are testing the alarms. He is fascinated by alarms though and always asks his dad if his alarm went off at work’ (age 6)

‘He hates screaming with emotion too. But he goes to have a look when he is crying now and looks at himself in the mirror or looks at the child’s face who is screaming even though he doesn’t like the noise. He has a fascination with motorbikes...but he doesn’t like the noise’ (age 7)

‘The electric toothbrush...she was anxious of the noise but drawn towards it – now she is vaguely obsessed with the toothbrush. She wants to put the electric toothbrush on but will then be anxious about it. She asks other people if they have one’ (9 years)
4. Discussion

The overall aim of the study was to use a parental semi-structured interview to explore the prevalence and phenomenology of RRB and sensory processing abnormalities in children with WS. The study builds on previous work by providing first hand parental accounts of these phenomena.

As expected, based on parent interview data sensory processing abnormalities were found to be highly prevalent in our sample, supporting Riby et al. (2012) and John and Mervis (2010) who both explored sensory processing abnormalities in children with WS using the Short Sensory Profile. This suggests sensory processing abnormalities are a fundamental characteristic of WS and require further understanding.

Those areas of sensory processing which were most frequently endorsed by parents in our sample were vestibular, auditory, gustatory and proprioceptive hypersensitivities. No instances of hyposensitivity were endorsed by the majority of parents indicating that the sensory profile of children with WS is characterised by hypersensitivities. Furthermore visual and tactile processing was not frequently reported as problematic. Parents were asked to provide examples of the nature of the difficulties their children experienced across the sensory domains and despite the unprompted nature of the questioning, certain themes emerged. With regard to vestibular difficulties, the impact was described in relation to its impact on play. Proprioceptive difficulties impacted on everyday activities like teeth cleaning and resulted in parents continuing to be involved in this activity longer than is typical. Auditory hypersensitivities were frequently associated with an aversion to everyday noises and frequently resulted in avoidance behaviours. Difficulties with texture combinations were
reported in relation to gustatory sensitivities and parents frequently reported concerns relating to restricted diets.

Frequent reports of the presence of RRB in children with WS supports anecdotal clinical evidence reported in the literature (e.g. Semel & Rosner, 2003). RRB reported by parents included repetitive language and questioning, dislike for changes in routine, and obsessional interests. Accounts of RRB in the SRAF-SSI, largely reflected higher order RRB rather than the more sensori-motor behaviours. This suggests the types of RRB seen in WS reflect more high level behaviours (Turner, 1999). The assessment of RRB in WS in both clinical and research contexts therefore should include more “compulsive-like” behaviours to be sensitive to this phenomenon in WS. We asked parents to reflect on potential triggers to the onset of a repetitive behaviour. The majority of parents who responded to this item indicated that anxiety was a trigger for the onset of repetitive behaviours. This is in contrast to Rodgers et al (2012) who reported, using standardised measures of RRB and anxiety no significant association relationship between RRB and anxiety in WS. This discrepancy may reflect the lack of sensitivity of the questionnaire measures to the specific associations between anxiety and RRB in WS. Some parents also indicated that illness may trigger RRB. This finding may in fact reflect a specific type of anxiety and is in keeping with the reports of heightened fear of physical injury reported previously in WS (Rodgers et al., 2012).

The results also reveal that parents perceive a relationship between RRB and sensory processing abnormalities in their children with WS supporting the conclusions of Riby et al. (2012). Indeed some parents reported that features of the auditory environment which had initially been associated with sensory discomfort developed in to special interests over time.
The complex relationship between these variables requires further understanding. It is unclear what the mechanism is whereby the presence of RRB and sensory processing abnormalities interact in WS. This phenomenon is similar to that described by Levitin, et al. (2005), who stated that all WS individuals in their study who were reported to have an auditory fascination, had previously been afraid of the object of fascination. Furthermore, children were described by their parents as “going beyond merely wanting to hear the sound” and seeking out pictures of the objects, wanting to learn as much as possible about the object, perhaps in a similar way to which a child with repetitive behaviours displays unusual or peculiar interests. This suggests a possible link between anxiety or fear of a sound, sensory modulation difficulties, and subsequent RRB. One speculative hypothesis might be RRB serve as a coping mechanism for the child by reducing acute anxiety and enabling some control to be exerted over their immediate environment. However given the unexpected nature of this finding we were not able to address this hypothesis directly here. This is an important area for future research, not least to determine whether this effect is restricted only to auditory processing abnormalities.

WS is a developmental disorder and as such it is appropriate to assume that the characteristics of the disorder will change with development. Whilst the cross-sectional nature of the present study precludes a detailed analysis of the developmental trajectory of sensory processing difficulties and RRB our data do provide some preliminary indications of the changing nature of these features over time. The majority of parents report that the frequency and impact of RRB has increased over time for their child. The developmental trajectory of sensory difficulties appears more complex, the impact of some sensory sensitivities are reported as improving over the course of development (e.g. proprioceptive and vestibular hypersensitivity) whereas other aspects of hypersensitivities appear to be relatively stable or
worsening (auditory hypersensitivity and gustatory hypersensitivity). Given the small sample size here, these suggested developmental patterns certainly require further exploration ideally with longitudinal research.

This study makes a significant contribution to our knowledge of sensory processing abnormalities, and RRB in children with WS. When interpreting the findings, it is important to acknowledge some of the weaknesses of the study. First, a relatively small sample size was achieved due to the low incidence of WS in the general population (1 in 20,000; Korenberg, et al., 2003) and therefore limited recruitment opportunities. The interview schedule was developed for the purposes of this study and this is the first reported use of the tool. Whilst steps were taken during the development of the measure to try to ensure that the measure has face validity through the use of piloting future work is required to ensure that it is a valid and reliable method of capturing the phenomena of interest. Furthermore although the development of a measure related to WS specific difficulties could be regarded as a key strength of the current study there will inevitably be a compromise in terms of generalizability. Similarly the use of a single group design limits the conclusions that can be drawn regarding specificity of the findings to WS. Whilst our intention was to explore the phenomenology of RRB and sensory difficulties in WS rather than to compare and contrast with other groups it is clear that a comparison of the findings from our WS sample with those from other conditions would be a useful endeavour. In addition the data were derived from only one informant per child. Future work would benefit from the inclusion of multiple sources of data.

The findings reported here provide a number of directions for future research. Firstly, as discussed, the specific impact that RRB and sensory processing difficulties have in WS
remains unclear and this is a crucial area for further investigation. Understanding these characteristics and the relationships between them would have significant clinical implications, in particular for the assessment, formulation, and treatment.

Limited research to date has mainly focused upon the prevalence of sensory processing abnormalities in individuals with WS, with little attention given to the presence of RRB and only one study to our knowledge has attempted to explore the relationships between the two (Riby et al. 2012). Until more is known about the phenomenology of these two features of WS and the interactions between them, it is difficult to develop effective treatment interventions to alleviate any negative impact for the individual and their family. Many families discussed how much their child’s RRB, and sensory processing abnormalities impacted upon family life, and how challenging it was to manage these difficulties. The majority of families reported that they wanted practical advice as to how to manage their child’s behaviour but were unable to find any specific to children with WS. Therefore, if more is understood about the role of sensory processing abnormalities and RRB in WS, this could inform directions for future clinical research to increase our knowledge of these constructs and also develop future practice, specifically the assessment process and treatment approaches. It also highlights the need for a multi-disciplinary assessment of the difficulties experienced by children with WS, such as the specialised assessment of sensory processing abnormalities by Occupational Therapists, a comprehensive emotional and behavioural assessment by Clinical Psychologists, and shared input from families and teachers.
5. References


