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BRIEF REPORT

Brief Report: Exploring the Relationship Between Sensory Processing and Repetitive Behaviours in Williams Syndrome

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Abstract This study explored the relationship between sensory processing abnormalities and repetitive behaviours in children with Williams Syndrome (WS; n = 21). This is a novel investigation bringing together two clinical phenomena for the first time in this neuro-developmental disorder. Parents completed the Sensory Profile (Short Form; Dunn in The sensory profile manual. San Antonio: The Psychological Corporation, 1999) and the Repetitive Behaviour Questionnaire (Turner 1995). A significant correlation was evident between the total scores on each of these measures; suggesting that children with WS who exhibit increased sensory processing abnormalities also display a higher number of repetitive behaviours. Further exploratory analyses of subscales of the measures indicated potentially important relationships that suggest a role for arousal regulation in the relationship between sensory processing abnormalities and repetitive behaviours in WS.

Keywords Williams syndrome · Sensory processing · Repetitive behaviour

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Abbreviations

WS	Williams syndrome
ASD	Autism spectrum disorder
RBQ	Repetitive behaviours questionnaire
SSP	Sensory profile-short form

Introduction

Williams syndrome (WS) is a neuro-developmental disorder with an estimated prevalence between 1:7,500 (Strømme et al. 2002) and 1:20,000 (Morris and Mervis 1999) and is caused by a sporadic deletion of 1.5 MB including 25–28 genes on chromosome 7 (7q11.23; Donnai and Karmiloff-Smith 2000). Cognitively, the disorder is most often characterised by mild to moderate intellectual difficulty (Searcy et al. 2004) with relative strengths of verbal compared to spatial processing. The disorder is also associated with social, behavioural, and emotional difficulties (for a full review of the literature, see Martens et al. 2008).

In our everyday lives it is essential that we process information from our environment to allow us to respond to that information in an appropriate manner. In both typical and atypical development there is wide variation in the way individuals' process sensory information. 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 et al. 2008: 867). Critical to the current investigation, sensory processing abnormalities have been identified in up to 90 % of children with WS (John and Mervis 2010). Such problems may relate to impairments of visual, auditory, and tactile perception (e.g. Semel and Rosner 2003) and/or sensory modulation difficulties

(including auditory, vestibular, and proprioceptive hyper- and hypo-sensitivity; John and Mervis 2010).

Within Autism Spectrum Disorder (ASD), studies have investigated relationships between sensory processing abnormalities and the presence of repetitive behaviours. Repetitive behaviours are defined as "repetitive, non-functional activities or interests that occur regularly and interfere with daily functioning" (Gabriels et al. 2005: 170). It has been suggested that children with ASD who experience sensory processing abnormalities may also experience more repetitive behaviours (e.g. Chen et al. 2009; Baker et al. 2008). Repetitive behaviours may be functional in regulating arousal levels for children with ASD who experience sensory processing abnormalities (e.g. Gabriels et al. 2008; Liss et al. 2006). Furthermore, sensory seeking may be an intrinsic motivator for repetitive behaviours in children with ASD and those with intellectual disability (Joosten et al. 2009). In a recent review of the literature of repetitive behaviours in ASD (Leekam et al. 2011), it was suggested that repetitive behaviours may be caused by hyper- or hypo-arousal, whereby arousal acts as a key trigger for repetitive behaviours. In relation to hyper-arousal, repetitive behaviours may serve as a coping strategy to enable children with ASD to regulate high levels of arousal or reduce their anxiety, and in the instance of hypo-arousal, they may increase sensory stimulation (Leekam et al. 2011). It is important to explore these relationships in children with other relevant neurodevelopmental disorders; for example WS.

Repetitive behaviours have been reported in up to 86 % of individuals with WS (Davies et al. 1998). Individuals with WS may engage in obsessive–compulsive behaviours, such as the compulsive need to identify the source of sudden noises or compulsive greeting behaviours (Semel and Rosner 2003). Although John and Mervis (2010) found evidence of a relationship between sensory processing abnormalities, problem behaviours, and adaptive functioning in children with WS, there are no studies to date that have looked specifically at the relationship between sensory processing and repetitive behaviours in WS. The aim of this preliminary study was therefore to explore sensory processing abnormalities and repetitive behaviours for the first time in children with WS. In line with research from other neurodevelopmental disorders, we hypothesise that children with WS who demonstrate more sensory processing abnormalities will exhibit more repetitive behaviours.

Method

Participants

Twenty-one children with WS aged 6- to 15-years (mean 9.3 years; 12 male) were recruited via the Williams Syndrome Foundation. All children had previously been clinically diagnosed and their diagnosis had been confirmed by positive fluorescent in situ hybridization testing (FISH). Mean estimated Full Scale IQ (FSIQ) was 52.6 (SD = 11.42), as measured using a Short Form of the WISC-III (Wechsler 1991), this is within the typical range associated with WS (cf. Mervis et al. 2000).

Measures

The *sensory profile—short form* (*SSP;* Dunn 1999) is a 38-item parent-report questionnaire asking parents to rate the frequency that their child displays 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 lower total overall behaviour score indicates greater impairment. The SSP has good internal consistency for the subscales (Cronbach's alpha = .47–.91), and established content validity and strong inter-rater reliability (Dunn 2005). Studies have reported that the SSP has discriminate validity of >95 % in identifying children with and without sensory modulation difficulties (McIntosh et al. 1999). It has been recommended as a good measure for research protocols (Dunn 1999).

The *repetitive behaviour questionnaire* (*RBQ;* Turner 1995, 1999) is a 33-item parent-report questionnaire measuring the prevalence, frequency, and duration of repetitive behaviours (Turner 1995). The RBQ was chosen as it is a widely used measure of repetitive behaviours in studies of children with ASD and it has been found to have good reliability (Honey et al. 2012). Furthermore, it has been found to have excellent inter-rater agreement (mean k value = .99) and test–retest reliability (mean agreement = .83; Turner 1999). There are three sub-scales; Repetitive Language, Sameness Behaviour, and Repetitive Movements. Scores are calculated for each subscale and a Total score.

Procedure

Questionnaire packs including the SSP and RBQ were sent to parents of individuals with WS who had agreed to participate in the study. An information sheet was also provided to each parent and child alongside the consent form. The researcher visited each child with WS to complete the WISC-III Short Form in their home. Favourable ethical opinion was granted by Newcastle University Faculty of Medical Sciences Ethics Committee.

Results

SSP Total Scores and RBQ Total Scores were normally distributed and achieved Cronbach's alpha coefficients above .8, indicating good to excellent reliability. Non-significant correlations were found between FSIQ and the SSP ($r = -.21$, $p = .37$) and the FSIQ and the RBQ ($r = .05$, $p = .83$); therefore the FSIQ was not controlled for in the subsequent analyses.

A two-tailed Pearson correlation revealed a significant negative correlation between the total score of the RBQ ($M = 13.3$, $SD = 8.6$) and the total score of the SSP ($M = 124.3$, $SD = 21$) ($r = -.60$, $p = .01$). As repetitive behaviours increased so did sensory processing abnormality.

Further exploration of the subscales of each measure was conducted (see Table 1). The three subscales of the RBQ were correlated with the seven subscales of the SSP. Significant correlations existed between RBQ Repetitive Movement and three subscales of the SSP; Tactile Sensitivity ($r = -.48$, $p = .03$), Taste/Smell Sensitivity ($r = -.52$, $p = .02$), and Under-responsive/Seeks Sensation ($r = -.58$, $p = .01$). RBQ Repetitive Language was significantly correlated with only the Under-responsive/Seeks Sensation subscale ($r = -.54$, $p = .01$). RBQ Sameness of Behaviour was significantly correlated with only the Taste/Smell Sensitivity subscale ($r = -.58$, $p = .01$).

Discussion

This study revealed a significant relationship between sensory processing abnormalities and repetitive behaviours in children with WS; those who experienced more sensory processing abnormalities demonstrated more repetitive behaviours. The findings mirror reports from other neurodevelopmental disorders such as ASD (e.g. Baker et al. 2008; Chen et al. 2009; Joosten et al. 2009). Critically, it is not possible to infer causality or make assumptions about the function of this relationship, but we provide new preliminary insights into the existence of this relationship that can inform future research and have clinical implications.

We use examples from the subscale correlations to propose specific aspects of the relationship between sensory processing and repetitive behaviours in WS. First, RBQ Repetitive Movements were significantly correlated with SSP Tactile Sensitivity. The RBQ Repetitive Movement subscale includes items addressing motoric, physical repetition, such as touching body parts or clothes, repetitive body movements, spinning, etc. The SSP Tactile Sensitivity scale includes rubbing or scratching where being touched, reacting emotionally to touch, not being able to stand too close to others, etc. We propose that engagement in some of the behaviours reported in the RBQ Repetitive Movement subscale occur *as a consequence* of tactile sensitivity. This relationship may be enforced as the child with WS attempts to regulate their hyper-arousal, however further research is required to investigate this proposal.

This possible role of arousal may gain some support from the highly significant relationship between RBQ Repetitive Movements and SSP Sensory Under-Responsiveness/Seeks Sensation scale and the significant relationship between RBQ Repetitive Language and SSP Sensory Under-Responsiveness/Seeks Sensation scale (see Table 1). This SSP scale includes behaviours such as seeks movement and fidgets, over excitable during movement activity, touches people and objects, etc. Again, these relationships may link to the requirement to seek sensory

Table 1 Pearson correlations between subscale scores on the SSP and RBQ for children with WS (n = 21)

Score	RBQ sameness of behaviour	RBQ repetitive movement	RBQ repetitive language
SSP tactile sensitivity	−.40	−.48*	−.20
	.08	.03	.39
SSP taste/smell sensitivity	−.58**	−.52*	−.29
	.01	.02	.22
SSP movement sensitivity	−.10	.04	.18
	.67	.86	.45
SSP under-responsive/seeks sensation	−.34	−.58**	−.54*
	.14	.01	.01
SSP auditory filtering	−.38	−.41	−.31
	.10	.07	.18
SSP low energy/weak	−.23	−.23	−.01
	.32	.33	.96
SSP visual/auditory sensitivity	−.02	−.14	.27
	.94	.55	.25

* Correlation significant at the .05 level (2-tailed)

** Correlation significant at the .01 level (2-tailed)

stimulation in order to regulate hypo-arousal. There has previously been a suggestion of low baseline arousal levels (hypo-arousal) associated with WS using very different paradigms and measured through galvanic skin responses (e.g. Doherty-Sneddon et al. 2009; Plesa-Skwerer et al. 2009). Equally relevant is evidence that abnormal regulation and structure of the amygdala in WS may play a role in atypicalities of arousal modulation in this group (e.g. Haas et al. 2009). Repetitive behaviours have been proposed to regulate arousal in children with ASD (Gabriels et al. 2008; Leekam et al. 2011; Liss et al. 2006), a disorder also associated with amygdala modulation abnormalities (e.g. Adolphs et al. 2001). Research of the nature reported here questions the specificity of these relationships to ASD and considers the possible link between these phenomena across neuro-developmental disorders.

An alternative explanation for this relationship (and indeed for others that we do not have sufficient space to contemplate here), may relate to overlap at the item level between the two scales; reflecting a lack of theoretical clarity between low level repetitive behaviours and sensory abnormalities. For example, a child rated on the SSP as having high levels of tactile sensitivity is also potentially likely be rated as frequently touching parts of the body or clothes by their parent. It is unclear whether the relationships reported here results from 'true relationships' between distinct clinical phenomena or are an artefact of poor construct independence and overlapping measurements. However, the relationship between the RBQ Repetitive Movements and RBQ Sameness of Behaviour with SSP Taste/Smell Sensitivity is less likely to be due to consequences of overlapping constructs. It may be that children who are sensitive to tastes and smells experience anxiety around food and use repetitive movements (e.g. self-soothing strategies) to reduce their anxiety (and associated arousal). Similarly, the desire for sameness of behaviour (e.g. wanting to eat the same foods, difficulty reacting to changes in routine etc.) may reduce anxiety for children with WS who are highly sensitive to taste and smell and help regulate hyper-arousal when it becomes uncomfortable. This suggestion once again contemplates a role for arousal when considering repetitive behaviours and sensory processing.

There are several clinical implications of the findings of the current study. At present little is known about the experience of these clinical phenomena in WS, or indeed the proposed relationship between them, thus emphasising the novelty and timeliness of the reported study. If more is known about the function of repetitive behaviours in relation to sensory processing abnormalities this knowledge could inform assessment and interventions for children with WS. For example, the development of comprehensive functional assessments of repetitive behaviours, with particular focus upon whether a child is demonstrating hyper- or hypo-arousal, may lead to intervention programmes that aim to reduce the level of repetitive behaviours whilst continuing to regulate the levels of arousal through the strategic use of more appropriate sensory activities.

This novel exploration makes a significant contribution to the understanding of sensory processing abnormalities and repetitive behaviours in children with WS. A total of 29 families were contacted to take part in the study which included all those children with WS within the North of England and Scotland known to the WSF in the age range of 5–15 years. Of the 29, 21 consented to take part, giving a moderately high consent rate of 72 %, reducing the likelihood of consent bias. However, it is also important to acknowledge some of the limitations of this study. Firstly, the relatively small sample size achieved was due to the low incidence of WS in the general population. As a result of the small sample size this study was underpowered, however, effect sizes were calculated for all analyses and despite a small sample size, moderate to large effects were found. Secondly, as highlighted, very little is known about the phenomenology of sensory processing abnormalities and repetitive behaviours in WS, and therefore the measures used may not be sensitive to assessing these clinical features in this group. Although these measures have been used with children with neuro-developmental disorders, and the SSP has been validated upon samples of children with and without disabilities, both have yet to be standardised on a WS population. Furthermore, as stated, parents' who report excessive repetitive movements are likely to endorse similar items on other scales of the SSP, such as tactile sensitivity, under-responsiveness, etc. Gabriels et al. (2008) recognised that many measures label a behaviour as repetitive on one scale and as sensory on another. In future studies it would be interesting to control for overlapping items to be able to infer more about the pure relationship between sensory processing and repetitive behaviours. Those future studies will also need to explore the mechanisms/functions of repetitive behaviours in relation to sensory processing and whether they serve to regulate arousal as this may suggest links with other clinical features of WS such as anxiety. In addition, as much of what we know about the relationship between these two constructs comes from research of ASD, comparisons between WS and ASD groups would help us to understand the specificity of the relationship between repetitive behaviours and sensory processing across neuro-developmental disorders. Using a combination of standardised measures and direct observations in future studies may also strengthen the findings. Further research should also explore the effect of chronological age on the developmental of this relationship between mechanisms and any

change with age, which is not possible at present due to the small sample size. This study was exploratory and therefore the findings offer an initial understanding of these constructs in WS. Critically, there is a timely need for further research to support and extend the preliminary findings reported here.

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