Sensory profiles in autism spectrum disorder and Williams syndrome

Magdalena Glod

Thesis submitted in candidature for the degree of Doctor of Philosophy

Newcastle University

Institute of Neuroscience

May 2017

Abstract

This thesis explored sensory profiles in children with autism spectrum disorder (ASD) and Williams syndrome (WS). The thesis begins with two review papers: the psychological correlates of sensory processing patterns in individuals with ASD were evaluated in a systematic review, followed by a mixed-methods review of sensory processing in Williams syndrome. Next, an investigation of changes in sensory symptoms across different age groups in children with ASD and WS was undertaken and revealed that level of sensory atypicalities in both disorders across age groups were very similar. This was followed with an examination of the sensory profiles of children with ASD without learning disability, ASD with learning disability and WS indicating that the distinction between the diagnostic group based on sensory behaviours and socio-communicative characteristics could hardly be made. A factorial validity of the Spence Children's Anxiety Scale-Parent version was then examined. The conventional SCAS-P structure in the ASD sample was not confirmed, raising concerns regarding the validity of the tool. Further exploration of sensory profiles in ASD and WS was then undertaken, where sensory processing clusters of children with both disorders were examined. The relationships between sensory processing and other clinical features were described and the mediating role of anxiety and intolerance of uncertainty between sensory processing abnormalities and repetitive behaviours was demonstrated. Next, the first comparison of sensory profiles in child-parent dyads in ASD and typical development (TD) was reported indicating some divergent patterns. Finally, a novel direct assessment of auditory and tactile sensory processing was developed and found to show promise as a measure for use with young children with ASD and WS. The synthesis of the evidence across chapters was then discussed and strengths and weaknesses of the current work presented. Suggestions for future research and the implications for clinical and research endeavours were considered.

Dedication

For mum

For my family

And for friends who became family

Acknowledgments

I will never be able to thank my supervisors enough; Jacqui Rodgers, Debbie Riby and Emma Honey for their guidance, support and patience throughout my PhD.

I am extremely grateful for the support of the children and families for their time and enthusiasm to take part in my studies. Recruiting participants would not be possible without the help of the Williams Syndrome Foundation, ASD-UK and schools in the North East of England.

I would also like to acknowledge Cathy Creswell, Polly Waite, Ruth Jameson, Mikle South and Helen McConachie for their help with data acquisition for analysis undertaken in Chapter 6. Cathy Creswell also described the anxious sample presented in that chapter.

The work in this PhD thesis was supported with funding from the Estate of David Murray Garside.

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The following publications relate to research presented in this thesis:

Chapter 2: Glod, M., Riby, D.M., Honey, E. & Rodgers, J. (2015). Psychological Correlates of Sensory Processing Patterns in Individuals with Autism Spectrum Disorder: A Systematic Review. *Review Journal of Autism and Developmental Disorders, 2*, 199–221. DOI: 10.1007/s40489-015-0047-8.

Chapter 6: Glod, M., Creswell, C., Waite, P., Jameson, R., South, M., McConachie, H. & Rodgers, J. (2017). Comparisons of the factor structure and measurement invariance of the Spence Children's Anxiety Scale - Parent version in children with autism spectrum disorder and typically developing anxious children. *Journal of Autism and Developmental Disorders (in press).*

Chapter 8: Glod, M., Riby, D.M., Honey, E. & Rodgers, J. (2016). Sensory Processing Patterns in Dyads of Children with ASD and Their Parents. *Autism Research.* DOI: 10.1002/aur.1680.

Chapter 1. General introduction to autism spectrum disorder and Williams syndrome

This thesis focuses on exploring the phenomenology and impact that sensory processing difficulties have on children with neurodevelopmental disorders. Two candidate disorders, namely autism spectrum disorder (ASD) and Williams syndrome (WS) provide the ideal opportunity to look at the syndrome-specific impact of these widely reported problem behaviours in order for us to understand the needs of individuals with different developmental disorders and provide appropriate support.

First, I define ASD and WS respectively, and introduce the sensory processing framework. A detailed literature review of sensory processing in both ASD and WS can be found in the following chapters (Chapter 2 and Chapter 3).

1.1 Autism spectrum disorder

Autism spectrum disorder (ASD) is a common neurodevelopmental disorder affecting around 1% of the population in the UK (Baron-Cohen et al., 2009). It is characterised by both core features, such as impairments in the social use of nonverbal and verbal communication (such as poor or inappropriate to the situation use of eye contact or gestures; or inability to initiate and maintain typical social interaction), and the presence of restricted, repetitive patterns of behaviour, interests, or activities (Diagnostic and Statistical Manual of Mental Disorders, DSM-5; American Psychiatric Association [APA], 2013). Recently, a subcategory of atypical sensory behaviour was included in the restricted, repetitive patterns of behaviour, interests, or activities diagnostic criterion, which includes a range of behaviours, from simple motor stereotypies to complex circumscribed interests (Richler et al., 2010). According to DSM-5, sensory symptoms can be exhibited as "hyper- or hypo-reactivity to sensory input or unusual interests in sensory aspects of the environment (e.g., apparent indifference to pain/temperature, adverse response to specific sounds or textures, excessive smelling or touching of objects, visual fascination with lights or movement)" (APA, 2013; p.50). Sensory characteristics, hence, alongside impairments in social communication and the presence of restricted and repetitive interests and behaviours, are now part of the diagnostic features of ASD.

Diagnosis of autism can be made on the basis of symptoms present within the early years of life (although these may become more apparent later in one's life) that cause clinically significant impairment in everyday functioning and the difficulties are not caused by intellectual disability nor global developmental delay (DSM-5, APA, 2013). To establish a

diagnosis a number of assessments are undertaken, including behavioural and observational evaluations, interviews with the child and the family (Volkmar et al., 2014).

In the previous diagnostic classification (DSM-IV, APA, 2000) a distinction was made between three subgroups of ASD, Asperger's disorder, autistic disorder and PDD-NOS (pervasive developmental disorder not otherwise specified), however, this classification was replaced more recently with a severity gradient to describe individuals with ASD (DSM-5, APA, 2013). This move towards a unitary category of ASD does not mean, however, that the heterogeneity associated with the disorder is no longer apparent and salient. As Georgiades et al. (2013) pointed out, there are layers of diversity in ASD associated with functional and ability levels (e.g. with some individuals being verbal or non-verbal), different configurations of core features (various degrees of social communication deficits and repetitive behaviours) resulting in different individual profiles, comorbid symptoms (such as anxiety) or casual factors. In this thesis the term autism spectrum disorder or ASD will be used throughout to encompass all autism spectrum conditions.

1.2 Williams syndrome

Williams syndrome (WS) is a neurodevelopmental disorder caused by the microdeletion of approximately 17-28 genes on chromosome 7q11.23 (Donnai & Karmiloff-Smith, 2000; Osborne, 2006). The prevalence rate ranges from 1 in 7500 (Strømme et al., 2002) to 1 in 20000 live births (Morris & Mervis, 1999) and in the UK the current agreed prevalence rate used by the Williams Syndrome Foundation is 1:18,000. Clinical diagnosis is confirmed by fluorescent in situ hybridization (FISH) testing and the detection of a missing copy of the Elastin gene (ELN) which represents a core genetic marker of the disorder. WS is characterised by mild to moderate intellectual disability (Searcy et al., 2004), distinctive facial features, such as a wide mouth with fleshy lips, periorbital fullness or prominent cheeks (Donnai & Karmiloff-Smith, 2000), and cardiovascular difficulties (Morris, 2006). The disorder is also associated with fascinating cognitive profile and personality features (John & Mervis, 2010; Jones et al., 2000).

Unusual cognitive profile, with a clear distinction between relatively stronger verbal abilities and impaired spatial abilities (all against a background of impaired IQ; Donnai & Karmiloff-Smith, 2000), hypersociability and a need to interact with others (Jones et al., 2000), have been the major research focus in the last four decades. More recently, high levels of sensory sensitivity in WS compared to children with other developmental disorders has been reported (Klein-Tasman & Mervis, 2003). Sensory sensitivity has been demonstrated in maladaptive physical and/or emotional reactions to everyday stimuli, in particular

hypersensitivity to certain sounds. As reported by Donnai and Karmiloff-Smith (2000) 85-95% of individuals with WS have been frequently sensitive to the sounds of machines, fireworks and bursting balloons. More recently WS has been also linked with elevated levels of repetitive interests or routine behaviours and a greater range of sensory processing atypicalities (Riby et al., 2013; Rodgers et al., 2012).

1.3 Sensory processing

Effective reception, integration and processing of sensory input, as visual, auditory or proprioceptive information, enables us to respond to environmental signals in an adaptive manner (John & Mervis, 2010), which is essential to everyday functioning and learning. If the process of responding to sensations is disturbed, for example, people may find it difficult to notice certain sensory inputs (Dunn, 2001).

Although there are individual differences in how sensory information is managed, in 1997, Winnie Dunn proposed a general model of sensory processing. In her conceptualization of patterns of sensory processing she suggested taking into consideration two main dimensions: the presence of high or low levels of nervous system reactivity (neurological thresholds) and specific self-regulation strategies, active or passive, used to respond to those thresholds. Our reactions to sensory input in everyday life are the result of the interaction between thresholds and response strategies, and can be presented as a continuum of possible responses to sensory events (Dunn, 2001). An individual's response and behaviours in relation to sensory stimulation could fall at any point on this range. However, some individuals may present with a similar pattern of responses to a number of sensory events. According to the Dunn model, four distinct patterns of sensory processing can be distinguished: Low Registration, Sensation Seeking, Sensory Sensitivity and Sensation Avoiding as depicted in Figure 1.1.

	Responding/Self-Regulation Strategies	
Thresholds/Reactivity	Passive	Active
High	Low Registration	Sensory Seeking
Low	Sensory Sensitivity	Sensory Avoiding

Figure 1.1 Dunn's Model of Sensory Processing. From "The Impact of Sensory Processing Abilities on the Daily Lives of Young Children and Families: A Conceptual Model" by W. Dunn, 1997, Infants and Young Children, 9(4), 23–25.

The first two patterns (upper row) relate to a high threshold for reactivity combined with either passive (Low Registration) or active (Sensory Seeking) responding strategies. As Dunn (2001) explains, individuals who meet criteria for these categories require a high level of sensory input before a response is forthcoming. People described as having low registration may not respond to some of the usual sensory events that other people notice easily and respond to (e.g. turning when a person's name is called). An additional stimulation (e.g. repetitive name calling or touching) might be required to get a response from them. Sensation seekers, on the other hand, actively look for ways of increasing sensory input in their everyday lives. They might be interested in either intensification of the stimulation (e.g. turning up the radio, diving in the pool to experience greater deep pressure) or in an increased range of sensory events by looking for stimulation from different sensory systems (e.g. taste and proprioception by eating different food textures and increasing physical movement by climbing or bouncing or humming).

The other two patterns (lower row) relate to the presence of low neurological thresholds. Those with sensory sensitivity may respond readily to sensory events which are not detected or noticed by other people. They are highly distractible to visual, auditory, vestibular, olfactory, and tactile stimuli (that do not ordinarily cause distraction) such as conversations held by people around them, certain food textures and flavours, and sunlight. Despite the level of irritation and preoccupation caused by noticing too many sensory events in their daily lives, individuals classified by Dunn as 'sensory sensitive' respond to this stimulation passively, not undertaking any actions to reduce the amount of sensory input. 'Sensory avoiders', on the contrary, seek to actively limit or avoid the number of sensory stimuli that could cause any form of distraction or aversion for them. For example, they might avoid crowded places, like shopping malls or buses, to reduce the sensory input created by other people's movement, sound or smell. It has also been hypothesised that may engage in rituals and routines to limit unfamiliar, unpredictable, or frustrating sensory input and create sensory patterns which are predictable and controllable (Brown et al., 2001; Dunn, 1997; Dunn & Brown, 1997; Dunn & Daniels, 2002).

There are several other theoretical approaches to the classification of sensory processing difficulties. The DSM-5 (APA 2013) highlights two sensory processing patterns, hyper- and hypo-responsiveness, understood as either an exaggerated behavioural reaction or lack of, or insufficient behavioural reaction to, sensory stimuli (Boyd et al. 2009). It has been claimed, for example, that features associated with the hyporesponsiveness pattern can discriminate between children with autism, developmental delay, and those of typical

development (Baranek et al. 2006). In addition, sensory atypicalities associated with different patterns of sensory processing may be present within the same individual with ASD as some may be hypo-responsive to certain stimuli and hyper-responsive to other sensory events (Baranek 2002; Baranek et al. 2006; Ben-Sasson et al. 2009).

Another approach taken when investigating sensory atypicalities focuses on sensory modulation disorder (SMD). SMD is characterized by difficulties in regulating and organizing appropriate behavioural responses to sensory input (Miller et al. 2007). The disorder has distinct three subtypes; namely over-responsivity, under-responsivity and sensory seeking associated with the craving of sensory experience (Miller et al. 2007). This classification system has been acknowledged by: the Diagnostic Classification of Mental Health and Developmental Disorders of Infancy and Early Childhood, Revised (known as the DC: 0–3R) (Zero to Three, 2005), the Diagnostic Manual for Infancy and Early Childhood of the Interdisciplinary Council on Developmental and Learning Disorders (ICDL, 2005), and the Psychodynamic Diagnostic Manual (PDM Task Force, 2006).

These multiple theoretical standpoints present in investigating sensory processing atypicalities are reflected in the current literature. However, in this thesis, only the Dunn's model of sensory processing will be further examined as it captures both hypo- and hyper-responsiveness to everyday sensory events and this theory forms the theoretical basis of the subsequent empirical chapters.

Chapter 2. Psychological correlates of sensory processing patterns in individuals with autism spectrum disorder: a systematic review

2.1 Background

2.1.1 Sensory atypicalities in ASD

As highlighted in Chapter 1, effective reception, integration, and processing of sensory input, as visual, auditory or proprioceptive information, enables us to respond to environmental signals in an adaptive manner (John & Mervis, 2010), which is essential to everyday functioning and learning. In autism spectrum disorder (ASD) it has been reported that sensory processing atypicalities are present in over 90% of children (Leekam et al., 2007) and adults (Crane et al., 2009) and sensory processing difficulties are now included in the most recent diagnostic criteria for ASD (DSM-5, APA, 2013) with "hyper- or hyporeactivity to sensory input or unusual interests in sensory aspects of the environment (e.g., apparent indifference to pain/temperature, adverse response to specific sounds or textures, excessive smelling or touching of objects, visual fascination with lights or movement)" (APA, 2013, p.50) as one of the diagnostic features.

2.1.2 Sensory processing patterns in ASD

There are several theoretical approaches to the classification of sensory processing difficulties in ASD. As described in Chapter 1, DSM-5 (APA, 2013) highlights two sensory processing patterns, hyper- and hyporesponsiveness, understood as exaggerated behavioural reaction and lack of, or insufficient behavioural reaction to, sensory stimuli (Boyd et al., 2009). Sensory seeking is also often distinguished and relates to craving of sensory experience (Miller et al., 2007). Following Dunn's model (1997) four sensory patterns can be discussed such as Low Registration, Sensation Seeking, Sensory Sensitivity and Sensation Avoiding. In the current ASD literature all those theoretical stands are represented.

2.1.3 Symptom co-morbidity

Research suggests that there is a relationship between sensory processing difficulties and the clinical features of ASD. Some studies reported significant associations between sensory processing atypicalities, communication and social impairments (Watson et al., 2011) as well as repetitive behaviours (Boyd et al., 2009), the presence of maladaptive behaviours, antisocial behaviours, self-absorption and parent-reported child anxiety (Baker et al., 2008) or perseveration and over focusing attention (Liss et al., 2006). There is also evidence of

significant associations between sensory processing atypicalities and other non-clinical psychological constructs such as temperament (Brock et al., 2012), emotion dysregulation (Samson et al., 2013) or eating difficulties (Nadon et al., 2011). However, there is variability in the methodological approaches used in those studies, including the selection of measures, diagnostic subgroups, and specified inclusion criteria. Due to a vast number of psychological constructs that have been investigated, and a wide range of methods of investigation employed, both interpretation and comparison of findings has been hampered.

2.1.4 Previous reviews

Five literature reviews of sensory atypicalities in individuals with ASD have been published to date (Ben-Sasson et al., 2009; Hazen et al., 2014; Iarocci & McDonald, 2006; O'Neill & Jones, 1997; Rogers & Ozonoff, 2005). However, these evaluations focused on differently defined sensory difficulties: Ben-Sasson et al. (2009) reviewed sensory modulation symptoms in individuals with autism, Hazen et al. (2014) were interested broadly in sensory symptoms, Iarocci and McDonald (2006) investigated multisensory integration, O'Neill and Jones (1997) studied unusual sensory responses, while Rogers and Ozonoff (2005) concentrated on sensory dysfunction. Secondly, the previous reviews employed different methodological approaches, ranging from experimental laboratory findings combined with theoretical and conceptual papers (Hazen et al., 2014; Iarocci & McDonald, 2006; Rogers & Ozonoff, 2005), through reviewing clinical and experimental studies (O'Neill & Jones 1997) to the inclusion of only clinical findings (Ben-Sasson et al., 2009). Thirdly, the previous reviews focused more on the discriminant validity of sensory atypicalities between ASD and typical groups. There is also a growing number of studies investigating physiological reactivity to different types of sensory stimuli (for review see Lydon et al., 2014). However none of the published reviews have described evidence of associations between sensory processing patterns in individuals with ASD and other psychological constructs. Therefore, this current approach to the review is important, because, while there is growing interest and research in sensory processing in individuals with ASD and sensory processing patterns are included in the diagnostic criteria for ASD (APA, 2013), a systematic summary of the recent findings is lacking.

2.1.5 Aim of the review

The current review therefore aims to systematically summarize and evaluate available evidence, recognise and discuss any shortcomings, and identify goals for future research in

order to address the following question: What are the psychological correlates of sensory processing patterns in individuals with ASD?

2.2 Method

2.2.1 Inclusion/ exclusion criteria

Inclusion and exclusion criteria were defined prior to conducting the literature search. Studies were eligible for inclusion if they investigated sensory processing patterns in individuals with ASD and explicitly reported associations with psychological correlates such as cognition, emotions, behaviour or interpersonal relationships. Studies were searched from 1997 onwards. Non-primary studies were excluded from the search (e.g. reviews, book chapters). Also single case studies and case series designs were excluded. This decision was based upon the consideration that results from single case studies would not provide quantitative statistical data which is important from the point of this review and do not allow further generalization of the findings. The search was neither restricted to any particular age group nor particular diagnostic subgroup.

2.2.2 Search Strategy

A systematic literature search aimed to identify studies reporting sensory processing patterns of individuals with ASD conducted up to March, 2016. The search used five electronic databases: Scopus, Web of Knowledge, PsychInfo, Embase and Medline. For both Scopus and Web of Knowledge, which allow authors to search for a number of keywords, the search terms were based on the keywords used in the Ben-Sasson et al. (2009) meta-analysis. After identifying relevant papers, additional keywords that were used in categorising those papers were added into the search terms. The combinations of the following search terms were used: a diagnostic term (autis* or "pervasive developmental disorder*" or Asperger), a sensory term (sensory or reactivity or responsivity or sensation*), and a descriptor term (processing or integration or modulation or regulation or stimul* or input or event* or dysfunction or respons* or profile* or symptom* or unusual or difficulties or interest* or feature* or experience* or hypo* or hyper* or pattern* or sensitiv* or seeking or avoid* or registration or threshold* or defensiveness). In PsychInfo, Embase and Medline databases searches are based on controlled vocabularies. However, because different types of headings are used for each database (e.g. medical subjects headings for Medline, but APA thesaurus for PsychInfo), the vocabulary used in the databases varied. For PsychInfo autism or pervasive developmental disorders or aspergers syndrome were used as diagnostic terms,

combined with sensory integration or intersensory processes or perceptual motor processes or sensorimotor measures or sensory adaptation or adaptation or thresholds or self stimulation. In the Embase database, Asperger syndrome or infantile autism or autism terms were used, combined with sensory dysfunction or abnormal sensation or sensory defensiveness or sensory stimulation or sensation or abnormal sensation or sensation seeking or self stimulation or perceptive threshold or sensorimotor function or sensorimotor integration. When searching in Medline a combination of terms child development disorders, pervasive or autistic disorder or Asperger syndrome, and sensory thresholds or sensation disorders or self stimulation or occupational therapy were used.

A flowchart of the search strategy and numbers of articles identified and excluded at each stage is outlined in Figure 2.1. All databases were searched between 1997 and the 14th of March 2016.

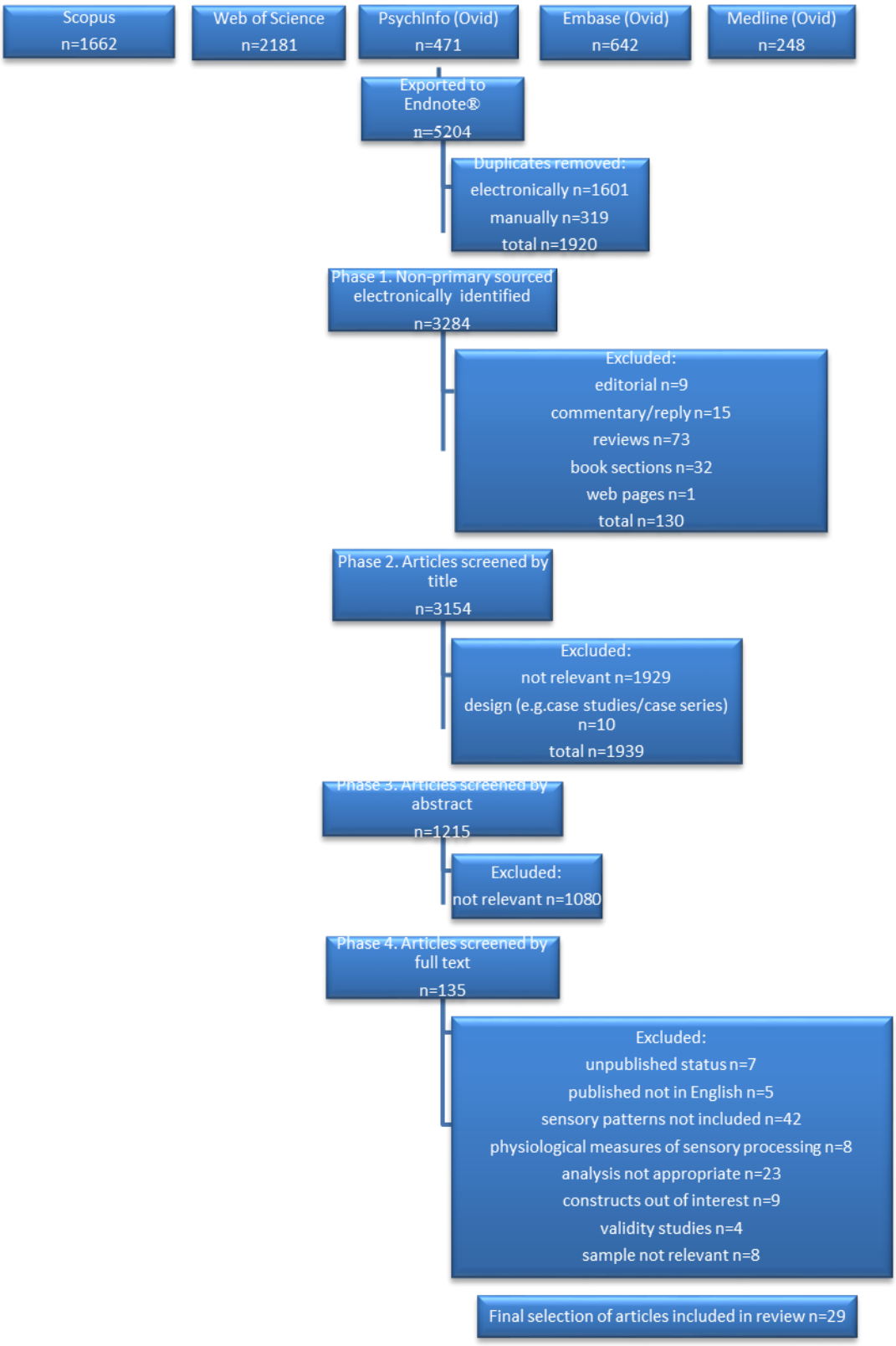


Figure 2.1 Flowchart of search

2.2.3 Electronic search

Results from five electronic databases were exported to Endnote® referencing software resulting in 5204 records in total. Most duplicates of the papers were identified by

Endnote's duplicate identification function and removed from the records' list. Further duplicates not recognised by the software were removed manually, and 3284 records were carried forward to the screening stage.

Screening of electronic search results

Screening of the search results consisted of four main phases. In Phase 1 the non-primary sources were electronically identified and removed (a total of 130 records). In Phase 2 the remaining titles of the records were screened considering their relevance to the search question and 1939 studies were removed. In Phase 3 remaining article abstracts were screened. Only one hundred and thirty five met inclusion criteria and those were carried forward to the final Phase in which articles were screened by full text and the final selection was made.

2.2.4 Final selection

A hundred and six papers were excluded after screening the full text. Seven papers were excluded due to unpublished status (three theses, two conference papers, two editorial). Five were excluded due to being published in languages other than English (Japanese, Italian, Portuguese, Korean and Chinese). In another forty two papers sensory atypicalities in general were investigated mainly reporting the Short Sensory Profile total score or sensory modalities such as auditory or tactile modality. Eight studies used physiological measures of sensory processing. Twenty three papers were not found appropriate due to the lack of correlational analysis. Nine papers did not include any psychological constructs, but examined relationships between sensory processing and for example oral care difficulties, leisure activities, or family life impairment and maternal parenting stress. Four papers were validity studies (investigating psychometric properties of tools). In eight papers a clear ASD sample was not recruited, either studies included participants from the general population, with or without some ASD-traits, or the results were presented for a combined ASD sample with another group (e.g. developmental delay). The remaining twenty nine papers were included in the systematic review. The summary of the descriptive characteristics of these studies can be found in Table 2.1.

Table 2.1 Summary of studies included in the review

Reference	Sample		Sensory processing pattern(s) (Sensory measures)	Analysis type	Psychological correlates measures	Main finding(s)
	N	Age Diagnosis				
Ashburner, J., et al. (2008)	28	Range:6-10 years old ASD	under responsive/seeks sensation (SSP)	correlational	CTRS–R:L ASEBA:TRF GADS GARS	Underresponsive/ seeks sensation was significantly negatively associated with academic performance and attention to cognitive tasks and with autism quotient.
Ausderau, K., et al. (2014)	1307	M=7.7 years (SD=2.7) Range: 2-12 years old ASD	hyporesponsiveness, hyperresponsiveness, sensory interests, repetitions and seeking behaviors, enhanced perception (SEQ-3.0)	correlational	SRS	Autism severity was significantly positively associated with all sensory response patterns.

Baker, A. E. Z., et al. (2008)	22	M=64.86 months (SD=20.70) Range:33-101 months old AD	under responsive/seeks sensation (SSP)	correlational	VABS DBC-P	Poor sensory processing ability was associated with higher levels of behavioural and/or emotional problems.
Baranek, G. T., et al. (2013)	63	Range:20–83months old AD	hyporesponsiveness (SPA)	inferential (series of regression models)	JAA MSEL PLS-4	Sensory hyporesponsiveness was significantly negatively associated with joint attention and language skills.
Bitsika, V., et al. (2016)	140	M=11.2 years (SD=3.3) Range: 6-18 year sold ASD	low registration, sensation seeking, sensory sensitivity, sensation avoiding (SP)	correlational	CASI-D	All sensory processing patterns except the sensory seeking were significantly correlated with the total CASI-D score.
Boyd, B.A. et al. (2010)	67	M=51.69 months (SD=17.07)	hyporesponsiveness, hyperresponsiveness, sensory seeking	inferential (series of	RBS-R	Higher hyperresponsive scores were related to a variety of repetitive behaviours. The significant association was found between

		AD	(SEQ, SP, SPA, TDDT-R)	regression models)		sensory seeking and ritualistic/sameness behaviours.
Brock, M. E., et al. (2012)	54	M=56.17months (SD=3.67) Range:36 - 84 months old ASD	hyporesponsiveness, hyperresponsiveness, sensory seeking (SEQ, SP, SPA, TDDT-R)	inferential (series of regression models)	BSQ	Hyporesponsiveness was most associated with distractibility, slowness to adapt and the threshold subscale. High levels of sensory features were associated with increased withdrawal and more negative mood.
Chen, Y.-H., et al. (2009)	29	Range:8-16 years old Asperger syndrome or ASD	under responsive/seeks sensation (SSP)	correlational and multiple regression	CRI EFT	No significant relationship was found between the presence of sensory abnormalities (underresponsiveness) and restricted and repetitive behaviours and detail-focused cognitive style
Gal, E., et al. (2010)	56	M=9.71 years (SD=1.86) autism	under responsive/seeks sensation (SSP)	correlational	SSIMI	Atypical sensory processing was strongly related to stereotyped movements (underresponsiveness was the best predictor of stereotyped movements).

Green, S. A., et al. (2012)	149	M=28.3 months (SD=5.5) ASD or PDD-NOS	sensory overresponsivity (ITSEA)	correlational	ITSEA	Sensory overresponsivity was positively associated with anxiety (and positively predicted increases in anxiety).
Hilton, C., et al. (2007)	36	Range: 6-10 years old HFASD	low registration, sensation seeking, sensory sensitivity, sensation avoiding (SP)	correlational	SRS	The SRS t scores showed moderate to strong relationships with Sensory Profile quadrant scores.
Jasmin, E., et al. (2009)	35	Range:3-4 years old ASD (AD and PDD-NOS)	low registration, sensation seeking, sensory sensitivity, sensation avoiding (SP)	correlational	PDMS-2 WeeFIM VABS-2	Some sensory responses were associated with motor skills, and there were many correlations between sensori-motor performances and daily living skills.
Lane, A. E., et al. (2010)	54	M=79.02 months (SD=19.22) Range:33-115 months AD	under responsive/seeks sensation (SP)	correlation and multiple regression	VABS	A clear predictive association was evident between sensory processing patterns, communication performance and general maladaptive behaviour.

Lane, S. J., et al. (2012)	23	Range:6-10 years old ASD	Sensory overresponsivity (SP or SensOR Inventory)	correlational	RCMAS	Sensory overresponsivity was strongly linked with anxiety.
Lidstone, J., et al. (2014)	49	M=10.7 years (SD=3.10) Range:3-17;9 years old ASD	low registration, sensation seeking, sensory sensitivity, sensation avoiding (SP)	correlational and mediation	RBQ-2 SCAS-P or PAS	Different sensory features contributed in different ways to the association between anxiety and restricted and repetitive behaviours.
Liss, M., et al. (2006)	144	M=102.4 months old (SD=50.1) ASD	overreactivity, underreactivity, sensory seeking (SQ)	correlational	DSM-IV checklist KOS VABS	The strongest positive correlation was found between overreactivity and overfocusing.
Mazurek, M. O., et al. (2013)	2973	M= 6.0 years (SD=3.5) Range:2-17 years old ASD	sensory overresponsivity (SSP)	correlational	CBCL GI SIQ	Anxiety, sensory overresponsivity, and GI problems were possibly interrelated phenomenon for children with ASD. There was a strong association between anxiety and sensory over-responsivity.

Mazurek, M. O., et al. (2014)	225	M=7.0 years (SD=3.7) Range: 2.4-17.4 years old ASD	sensory overresponsivity (SSP)	correlational	CBCL	Anxiety and sensory overresponsivity were significantly negatively correlated.
Mazurek, M. O., & Petroski, G.F. (2015)	1347	M=7.9 years (SD=3.4) Range: 2-17.6 years old ASD	sensory overresponsivity (SSP)	correlational	CBCL CSHQ	Anxiety and sleep problems were significantly negatively correlated with sensory overresponsivity for both the younger and older groups.
Nadon, G., et al. (2011)	95	M=7.3 years (SD=2.5) Range: 3-10 years old Autism (61%), PDD-NOS (29%) or Asperger syndrome (10%)	under responsive/seeks sensation (SSP)	linear regression	Eating Profile	Under responsive/seeks sensation was not significantly associated with the number of eating problems.

Pfeiffer, B., et al. (2005)	50	M=9.8 years Range:6-16 years old Asperger's disorder	hypersensitivity, hyposensitivity (SP or AASP)	correlational	RCMAS CDI ABAS	There were significantly strong positive correlations between sensory defensiveness and anxiety.
Reynolds, S., et al. (2012)	27	Range:6-12 years old ASD	low registration, sensation seeking, sensory sensitivity, sensation avoiding (SP)	correlational	CBCL	There was a significant correlation between sleep problems and a low sensory threshold/high arousal.
Samson, A. C., et al. (2013)	56	Range:6-16 years old ASD	under responsive/seeks sensation (SSP)	correlational	EDI	Sensory abnormalities were significantly related to emotion dysregulation.
Sullivan, J. C., et al. (2014)	81	M=10.3 years (SD=2.6) Range: 7-17 years old ASD	sensory overresponsivity (SPSI)	correlational	SCAS	Anxiety scores were positively correlated with total sensory overresponsivity score.

Tavassoli, T., et al. (2014)	221	M=38.7 years (SD=12.0) ASC	sensory overresponsivity (SP scale)	correlational	AQ	Sensory overresponsivity was positively correlated with autistic traits.
Tomchek, S., et al. (2015)	400	M=49.58 months (SD=10.54) ASD	hyporesponsivity, sensory seeking/distractibility (SSP)	multivariate regression	Information not obtained	Hyporesponsivity was associated with language limitations. Sensory seeking/distractibility subscale was significantly associated will social behaviour, receptive language, gross and fine motor skills, but not with expressive language.
Tseng, M.-H., et al. (2011)	67	M=64.21 months (SD= 9.01) autism	low registration, sensation seeking, sensory sensitivity, sensation avoiding (SP-C)	correlational	CBCL-C	Correlations between internalizing and externalizing problems and the four quadrants scores of the SP-C were significant, but low.
Watson, L. R., et al. (2011)	72	M=52.3 months (SD=16.5) AD	hyporesponsiveness, hyperresponsiveness, sensory seeking (SEQ, SP, SPA, TDDT-R)	factor analytic model	ADOS MSEL or PLS-4 VABS	Hyporesponsiveness had a significant positive association with social-communicative symptom severity, and was negatively associated with language scores as well as social adaptive scores. Also sensory seeking

						was negatively correlated with language scores.
Wigham, S., et al. (2015)	53	M=12.49 years (SD=2.3) Range: 8-16 years old ASD	overresponsiveness, underresponsiveness (SSP)	correlational	SCAS-P IUS-P RBQ	Sensory overresponsiveness was significantly negatively associated with anxiety, intolerance of uncertainty and repetitive behaviours, while sensory underresponsiveness correlated significantly only with repetitive behaviours' scores.

Note: AASP-Adolescent/Adult Sensory Profile, ABAS-Adaptive Behavior Assessment System, ADOS-Autism Diagnostic Observation Schedule, ASEBA:TRF-Achenbach System of Empirically Based Assessment: Teacher Report Form, AQ-Autism Spectrum Quotient, BAI-Beck Anxiety Inventory, BSQ-Behavioral Style Questionnaire, CASI-D-Child and Adolescent Symptom Inventory-Depressive symptom subscale, CBCL-C-Child Behavior Checklist for ages 4–18 Chinese version, CBCL-Child Behavior Checklist, CDI-Children's Depression Inventory, CES-D-Center for Epidemiological Studies—Depression Scale, CRI-Childhood Routines Inventory, CSHQ-Children's Sleep Habits Questionnaire, CTRS-R:L-Conner's Teacher Rating Scale–Revised Long Version, DBC-P-Developmental Behaviour Checklist—Parent, DSM-IV checklist-Diagnostic and Statistical Manual of Mental Disorders 4th edition checklist, EDI-Emotion Dysregulation Index, EFT-Embedded Figures Test, GADS – Gilliam Asperger's Disorder Scale, GARS – Gilliam Autism Rating Scale, GI SIQ -Gastrointestinal Problems Symptom Inventory Questionnaire, ITSEA-Infant Toddler Social and Emotional Assessment, ITSEA-Infant Toddler Social and Emotional Assessment, IUS-P-Intolerance of Uncertainty Scale-Parent Version, JAA-Joint Attention Assessment, KOS-Kinsbourne Overfocusing Scale, MSEL-Mullen Scales of Early Learning, PAS-Preschool Anxiety Scale, PDMS-2-Peabody Developmental Motor Scales—2nd edition, PLS-4-Preschool Language Scale Fourth Edition, RBQ-Repetitive Behaviour Questionnaire, RBQ-2-Repetitive Behavior Questionnaire 2, RCMAS-Revised Children's Manifest Anxiety Scale Adapted Parent's Version, SCAS-P-Spence Children's Anxiety Scale-Parent Version, SensOR- Sensory Overresponsiveness Inventory, SEQ-Sensory Experiences, Questionnaire, SEQ-3.0-Sensory Experience Questionnaire Version 3.0, SPA-Sensory Processing Assessment, SP-Sensory Profile, SP-C-Sensory Profile-Chinese version, SP Scale- Sensory Processing Scale, SPSI- Sensory Processing Scale Inventory, SQ-Sensory Questionnaire, SRS-Social Responsiveness Scale, SSIMI-Stereotyped and Self-Injurious Movement Interview, SSP-Short Sensory Profile, TDDT-R-Tactile Defensiveness and Discrimination Test, VABS-2-Vineland Adaptive Behavior Scales—Second Edition, VABS-Vineland Adaptive Behaviour Scales, WeeFIM-Functional Independence Measure

2.2.5 Critical evaluation

Each of the retained papers was evaluated against a review quality evaluation grid developed for the purpose of this review. The available checklists for the quality assessment of studies (e.g. PRISMA, Moher et al., 2009; QUADAS, Whiting et al., 2003) or well-known guidelines for conducting systematic reviews in health care (e.g. the Cochrane Collaboration) focus on diagnostic accuracy, evaluation of randomised trials and intervention studies. The newly developed grid aimed to systematically evaluate the overall quality of the studies, their strengths and limitations or potential sources of bias. The grid was divided into four main sections, following the IMRaD structure: introduction, methods, results and discussion (Sollaci & Pereira, 2004). The methods section was of particular importance including items evaluating a studie's quality in participants and method selection. To adequately evaluate the methodology used in the studies, the grid contained items concentrating on appropriate sample characteristics and confirmation of ASD diagnosis. The methods section of the evaluation grid also highlighted the importance of sound psychometric properties of the tools used in the studies as suggested by the COnsensus-based Standards for the selection of health Measurement INstruments (COSMIN) guideline (Mokkink et al., 2010). The total number of criteria that the studies were scored against was kept within the recommended limit to keep clear focus of the review (SIGN, 2008).

Subjective judgement is a part of the evaluation process (Deeks et al., 2003; SIGN, 2008), to minimise the reviewer's subjectivity the following steps were undertaken. First, all scoring criteria were explained in detail. Second, three levels of quality ratings were used, the equivalent of the levels of ratings proposed by SIGN (high, acceptable and low quality). Finally, a proportion of the studies included in the review (14%) were evaluated by an independent rater. The inter-rater reliability between the author's and independent rater's scorings calculated as percentage agreement on individual criteria was 87.5%.

2.3 Results

Of the 3284 unique references identified via the electronic searches, 29 papers met the inclusion criteria and were retained for review.

2.3.1 Evaluation grid – papers' quality

Originally the papers included in the review were scored against 26 criteria. Ten criteria were emphasised during the evaluation. Two criteria were selected from the participants' section ('Was ASD diagnosis confirmed for the study?' and 'Is the sample adequately described?'). They allowed us to assess whether the sample of interest was

included in the study and whether the authors reported participants' characteristics in a high-quality manner. Items from the 'Sensory measures' and 'Psychological correlate measure' sections were also considered as the criteria of the key importance. They allowed us to evaluate the appropriateness, reliability and validity of the tools used in the studies. The chosen criteria are fundamental to evaluate the quality of the studies in the light of the research question asked in this review. For the summary of the information included in the evaluation grid and ten selected criteria, see Table 2.2 and Table 2.3.

Table 2.2 Evaluation grid

Domain	Criterion	Classification	Scoring criteria
Introduction			
Item 1	Are the constructs of interest adequately described?	Yes	The constructs of interest are adequately defined or described
		Partially	The constructs of interest are somewhat unclear or only some constructs are clearly defined
		No/NR	Lack of definitions and descriptions of the constructs of interest
Item 2	Is the research question clearly formulated?	Yes	The research question of the study is clearly formulated
		Partially	The research question is stated but somewhat unclear
		No/NR	The research question of the study is unclear or not stated
Item 3	Are the hypotheses clearly stated and operationalized?	Yes	The hypotheses of the study are clearly stated and operationalized
		Partially	The hypotheses are clearly stated, but not operationalized or operationalization of hypotheses is somewhat unclear or hypotheses are vague, but the operationalization is clear
		No/NR	The hypotheses of the study are unclear and are not operationalized or not stated
Methods			
Participants			

Item 4	Is the sample used in the study representative?	Yes	A population based sample was targeted
		Partially	A convenience sample was used with an attempt to use multiple recruitment sources
		No/NR	A highly selective recruitment method was used (e.g. selectively referred patients already taking part in another study) or recruitment sources are not reported
Item 5	Is the sample used in the study homogenous and recruited at the same time point?	Yes	The sample is recruited for the study at the same time point.
		Partially	The sample is recruited for the study, but the participants are assessed at different time points.
		No/NR	The sample consists of pooled samples from different studies and the data is collected at different time points.
Item 6*	Was ASD diagnosis confirmed for the study?	Yes	Diagnoses have been confirmed for this study by use of a 'gold-standard' diagnostic tool (i.e. ADOS or ADI-R)
		Partially	Diagnoses have been confirmed for this study, but not by use of a gold-standard tool
		No/NR	ASD diagnoses have not been confirmed for this study or diagnoses were confirmed for the study but paper does not provide detail how

Item 7	Are inclusion and exclusion criteria described?	Yes	Inclusion and exclusion criteria are explicitly reported
		Partially	Only inclusion but not exclusion criteria are explicitly reported
		No/NR	Inclusion and exclusion criteria are not explicitly reported
Item 8	Was level of cognitive functioning of participants assessed?	Yes	Level of cognitive functioning is reported and based on assessment using a standardised instrument and was assessed either for the study or within the preceding 3 months
		Partially	Level of cognitive functioning is reported but is based on previous (non-recent) assessment or on method other than standardised instrument (e.g. position in school system) or cognitive function was assessed but very broadly reported (e.g. 'all participants had FSIQs over 75 as assessed by….' or 'MA less than 6 months')
		No/NR	Level of cognitive functioning is not reported
	Are sample characteristics described?		
	Age	Yes	Age range and mean are reported
		Partially	Either age range or mean is reported
		No/NR	Age range and mean are not reported
	Gender	Yes	Gender of participants is reported
		Partially	Gender of participants is somehow reported (proportional data reported)
		No/NR	Gender of participants is not reported

	ASD subtype	Yes	ASD subtypes included are reported
		No	ASD subtypes included are not reported
	Comorbidities	Yes	Presence and detail of relevant comorbidities is reported
		No	Presence and detail of relevant comorbidities is not reported
	Other demographic variables	Yes	Other demographic variables are reported (e.g. location, ethnicity, race)
		No	Other demographic variables are not reported
Item 9*	Based on the above, is the sample adequately described?	Yes	All the above details are given
		Partially	Most of the above details are given
		No/NR	Few or none of the above details are given
Measures: Sensory measures			
Item 10*	Are sensory processing patterns measured using standardised measures of sensory processing?	Yes	Standardised measures are used in this study
		Partially	Non standardised measures are used, but reference to current standardisation work is provided
		No/NR	Non standardised measures are used
Item 11*	Are sensory processing patterns measured using valid measures of sensory processing?	Yes	Evidence of good validity of the measures is provided in this study (e.g. 50% of the variance explained by factors, correlations with 'gold' standard measures ≥ 0.70)

		Partially	Evidence of validity not provided in this study, but reference to cited studies providing evidence of acceptable validity of the measures or evidence provided for the whole measure, but only some items/subscales are used in the study
		No/NR	Non validated measures are used or no reported evidence of validity is provided
Item 12*	Are sensory processing patterns measured using reliable measures of sensory processing?	Yes	Evidence of good reliability of the measures is provided in this study (e.g. Cronbach's alpha(s), ICC $\geq$ 0.7)
		Partially	Evidence of reliability not provided in this study, but reference to cited studies providing evidence of acceptable reliability of the measures or evidence provided for the whole measure, but only some items/subscales are used in the study
		No/NR	Non reliable measures are used or no reported evidence of reliability is provided
Item 13*	Are the measures used appropriate for use with an ASD population?	Yes	Evidence provided that tools used have been standardised and validated for use with ASD population or are ASD-specific
		Partially	Tool has not been standardised for ASD population but it has been validated or is widely used in ASD research or evidence of use with comparable developmental groups is provided in this study
		No/NR	No evidence that tool is appropriate for ASD population
Measures: Psychological correlate measure			

Item 14*	Is the psychological correlate measured using standardised measures of the construct?	Yes	Standardised measures are used in this study
		Partially	Non standardised measures used, but reference to current standardisation work is provided
		No/NR	Non standardised measures are used
Item 15*	Is the psychological correlate measured using valid measures of the construct?	Yes	Evidence of good validity of the measures is provided in this study (e.g. 50% of the variance explained by factors, correlations with 'gold' standard measures ≥ 0.70)
		Partially	Evidence of validity not provided in this study, but reference to cited studies providing evidence of acceptable validity of the measures or evidence provided for the whole measure, but only some items/subscales are used in the study
		No/NR	Non validated measures are used or no reported evidence of validity is provided
Item 16*	Is the psychological correlate measured using reliable measures of the construct?	Yes	Evidence of good reliability of the measures is provided in this study (e.g. Cronbach's alpha(s), ICC ≥ 0.7)
		Partially	Evidence of reliability not provided in this study, but reference to cited studies providing evidence of acceptable reliability of the measures or evidence provided for the whole measure, but only some items/subscales are used in the study
		No/NR	Non reliable measures are used or no reported evidence of reliability is provided

Item 17*	Are the measures used appropriate for use with an ASD population?	Yes	Evidence provided that tool used has been standardised and validated for use with ASD population or is ASD-specific
		Partially	Tool has not been standardised for ASD population but it has been validated or is widely used in ASD research or evidence of use with comparable developmental groups is provided in this study
		No/NR	No evidence that tool is appropriate for ASD population
Results			
Item 18	Are the descriptive statistics appropriately reported?	Yes	The descriptive statistics are appropriately reported (e.g. M, SD, range)
		Partially	Only some of the descriptive statistics are reported or the descriptive statistics are reported for selected constructs
		No/NR	The descriptive statistics are not appropriately reported or not reported at all
Item 19	Are the results presented clearly?	Yes	The results are presented clearly (e.g. tables and figures are easy to read, clearly labelled, the description of the results is easy to follow)
		Partially	The results are presented somehow unclear
		No/NR	The presentation of the results is difficult to follow
Item 20	Are the psychometric properties reported in the current sample?	Yes	Validity and reliability are reported in the current sample

		Partially	Either validity or reliability is reported in the current sample or both reported only for selected constructs
		No/NR	Validity and reliability are not reported in the current sample
Item 21	Are the missing values reported and how they were handled?	Yes	Percentage of missing items and how missing items were handled are described in the study or one-to-one assessments are conducted
		Partially	Percentage of missing items is described, but somehow not clear how missing items were handled
		No/NR	Percentage of missing items not described and not reported how missing items were handled
Analysis			
Item 22	Is the statistical analysis appropriate to the design?	Yes	The analytic strategy is appropriate to the design
		Partially	The analytic strategy is appropriate but has some limitations (e.g. other analytical strategy would have been more powerful or some assumptions have been violated)
		No/NR	Inappropriate statistical tests were used or insufficient information is provided to judge the appropriateness of the analysis
Item 23	Is the sample size sufficient?	Yes	Sample size is based on appropriate power calculations, which are explicitly reported
		Partially	Power calculations are not reported but sample size appears sufficiently large
		No/NR	No justification is given for sample size and sample size appears small

Item 24	Are the effect sizes calculated and reported in the study?	Yes	The effect sizes are calculated for the data and reported in the study
		Partially	The effect sizes are calculated for the data, but not reported in the study
		No/NR	Lack of calculation of the effect sizes for the data
Discussion			
Item 25	Do the conclusions follow adequately from results?	Yes	Main findings are clearly described and follow appropriately from the results and analyses
		Partially	Some limitations in the clarity of description of main findings and their relation to results
		No/NR	Lack of appropriate description of findings and/or findings are over/ understated and do not follow clearly from results
Item 26	Are limitations acknowledged?	Yes	Clear acknowledgement of main limitations of the study and consideration given to the impact of these on interpretation
		Partially	Some limitations are acknowledged but not all, or no consideration given to the impact of limitations on interpretation
		No/NR	No acknowledgement of limitations

Note: NR-not reported, * indicates items included in ten selected criteria of evaluation

Table 2.3 Scoring against selected criteria

Reference	Participants	Sensory measures				Psychological correlate measure						
	Item 6	Item 9	Measure	Item 10	Item 11	Item 12	Item 13	Measure	Item 14	Item 15	Item 16	Item 17
Ashburner et al. 2008	partially	partially	SSP	NR	NR	partially	no	CTRS-R:L	NR	partially	partially	no
								ASEBA:TRF	NR	partially	partially	no
								GARS	NR	partially	partially	yes
								GADS	NR	partially	partially	yes
Ausderau et al. 2014	partially	yes	SEQ-3.0	NR	yes	partially	yes	SRS/SRS-P	NR	NR	NR	yes
Baker et al. 2008	yes	partially	SSP	NR	partially	partially	no	VABS	NR	NR	NR	no
								DBC-P	NR	NR	NR	no
Baranek et al. 2013	yes	partially	SPA	NR	NR	yes	yes	MSEL	yes	partially	partially	no

								PLS-4	yes	partially	partially	no
								JAA	NR	partially	yes	partially
Bitsika et al. 2016	partially	partially	SP	NR	partially	yes	partially	CASI-D	yes	yes	yes	partially
Boyd et al. 2010	yes	partially	SP	NR	NR	NR	no	RBS-R	NR	partially	partially	yes
			SEQ	NR	NR	NR	no					
			TDDT-R	NR	NR	NR	no					
			SPA	NR	NR	NR	yes					
Brock et al. 2012	yes	partially	SP	NR	NR	partially	partially	BSQ	NR	partially	partially	no
			SEQ	NR	partially	partially	yes					
			TDDT-R	NR	partially	partially	partially					
			SPA	NR	NR	partially	yes					

Chen et al. 2009	yes	partially	SSP	NR	NR	partially	partially	CRI	NR	partially	partially	no
								EFT	NR	partially	NR	partially
Gal et al. 2010	partially	partially	SSP	yes	partially	partially	partially	SSIMI	NR	NR	partially	no
Green et al. 2012	yes	partially	ITSEA	NR	NR	yes	no	ITSEA	NR	NR	partially	no
Hilton et al. 2007	partially	partially	SP	NR	partially	partially	partially	SRS	yes	partially	partially	yes
Jasmin et al. 2009	yes	partially	SP	yes	partially	partially	no	PDMS-2	NR	NR	partially	no
								WeeFIM	NR	partially	partially	no
								VABS-2	NR	NR	partially	no
Lane et al. 2010	yes	partially	SSP	yes	partially	partially	no	VABS	yes	partially	partially	partially

Lane et al. 2012	partially	no	SP	yes	partially	partially	no	RCMAS	NR	partially	partially	no
			SensOR	NR	partially	partially	no					
Lidstone et al. 2014	yes	partially	SP	NR	partially	partially	no	RBQ-2	NR	partially	yes	no
								SCAS-P	yes	partially	partially	no
								PAS	yes	partially	NR	no
Liss et al. 2006	partially	no	SQ	partially	partially	NR	yes	DSM-IV checklist	NR	NR	NR	no
								KOS	NR	partially	NR	partially
								VABS	yes	NR	NR	no
Mazurek et al. 2013	yes	partially	SSP	NR	partially	partially	partially	CBCL	yes	partially	partially	partially
								GI SIQ	NR	NR	NR	yes

Mazurek et al. 2014	yes	partially	SSP	NR	partially	partially	partially	CBCL	NR	partially	partially	partially
Mazurek and Petroski 2015	partially	partially	SSP	NR	partially	partially	partially	CBCL	NR	partially	partially	partially
								CSHQ	NR	partially	partially	partially
Nadon et al. 2011	partially	yes	SSP	yes	partially	partially	no	Eating Profile	NR	yes	yes	yes
Pfeiffer et al. 2005	yes	partially	SP	NR	partially	partially	no	ABAS	yes	partially	partially	no
			AASP	yes	yes	yes	no	RCMAS	NR	partially	partially	no
								CDI	NR	partially	partially	no
Reynolds et al. 2012	yes	partially	SP	yes	NR	NR	no	CBCL	NR	partially	NR	no

Samson et al. 2013	yes	partially	SSP	NR	NR	NR	no	EDI	NR	partially	yes	no
Sullivan et al. 2014	partially	partially	SPSI	NR	NR	NR	no	SCAS	NR	NR	NR	no
Tavassoli et al. 2014	partially	no	SensOR	NR	partially	partially	no	AQ	NR	NR	partially	yes
Tomchek et al. 2015	yes	partially	SSP	yes	NR	NR	partially	various	N/O	N/O	N/O	N/O
Tseng et al. 2011	partially	no	SP-C	NR	partially	partially	partially	CBCL-C	NR	NR	partially	no
Watson et al. 2011	yes	partially	SEQ	NR	partially	partially	yes	ADOS	NR	NR	NR	yes
			SP	yes	NR	NR	partially	MSEL	yes	NR	NR	no
			SPA	NR	NR	partially	yes	PLS-4	yes	NR	NR	no
			TDDT-R	NR	partially	NR	yes	VABS	yes	NR	NR	no

Wigham et al. 2015	partially	no	SSP	yes	NR	partially	no	SCAS	NR	partially	partially	partially
								IUS-P	NR	partially	partially	partially
								RBQ	NR	partially	partially	partially

Note: AASP-Adolescent/Adult Sensory Profile, ABAS-Adaptive Behavior Assessment System, ADOS-Autism Diagnostic Observation Schedule, ASEBA:TRF-Achenbach System of Empirically Based Assessment: Teacher Report Form, AQ-Autism Spectrum Quotient, BAI-Beck Anxiety Inventory, BSQ-Behavioral Style Questionnaire, CASI-D-Child and Adolescent Symptom Inventory-Depressive symptom subscale, CBCL-C-Child Behavior Checklist for ages 4–18 Chinese version, CBCL-Child Behavior Checklist, CDI-Children's Depression Inventory, CES-D-Center for Epidemiological Studies—Depression Scale, CRI-Childhood Routines Inventory, CSHQ-Children's Sleep Habits Questionnaire, CTRS-R:L-Conner's Teacher Rating Scale–Revised Long Version, DBC-P-Developmental Behaviour Checklist—Parent, DSM-IV checklist-Diagnostic and Statistical Manual of Mental Disorders 4th edition checklist, EDI-Emotion Dysregulation Index, EFT-Embedded Figures Test, GADS – Gilliam Asperger's Disorder Scale, GARS – Gilliam Autism Rating Scale, GI SIQ -Gastrointestinal Problems Symptom Inventory Questionnaire, ITSEA-Infant Toddler Social and Emotional Assessment, ITSEA-Infant Toddler Social and Emotional Assessment, IUS-P-Intolerance of Uncertainty Scale-Parent Version, JAA-Joint Attention Assessment, KOS-Kinsbourne Overfocusing Scale, MSEL-Mullen Scales of Early Learning, N/O – not obtained, PAS-Preschool Anxiety Scale, PDMS-2-Peabody Developmental Motor Scales—2nd edition, PLS-4-Preschool Language Scale Fourth Edition, RBQ-Repetitive Behaviour Questionnaire, RBQ-2-Repetitive Behavior Questionnaire 2, RCMAS-Revised Children's Manifest Anxiety Scale Adapted Parent's Version, SCAS-P-Spence Children's Anxiety Scale-Parent Version, SensOR- Sensory Overresponsiveness Inventory, SEQ-Sensory Experiences, Questionnaire, SEQ-3.0-Sensory Experience Questionnaire Version 3.0, SPA-Sensory Processing Assessment, SP-Sensory Profile, SP-C-Sensory Profile-Chinese version, SP Scale- Sensory Processing Scale, SPSI- Sensory Processing Scale Inventory, SQ-Sensory Questionnaire, SRS-Social Responsiveness Scale, SSIMI-Stereotyped and Self-Injurious Movement Interview, SSP-Short Sensory Profile, TDDT-R-Tactile Defensiveness and Discrimination Test, VABS-2-Vineland Adaptive Behavior Scales—Second Edition, VABS-Vineland Adaptive Behaviour Scales, WeeFIM-Functional Independence Measure

2.3.2 Participants' section

The two items describing participants' characteristics are essential to establish whether the particular clinical group of interest was selected according to widely accepted research standards. In addition, it was important to confirm whether or not the characteristics were described well enough to allow other researchers to replicate the study and identify some possible important covariates that might influence the study findings. All the studies provided a confirmation of diagnosis of participants. In sixteen papers the assessment of children was carried out prior to inclusion in the study by using 'gold-standard' diagnostic tools such as Autism Diagnostic Observation Schedule (ADOS) or Autism Diagnostic Interview-Revised (ADI-R). In the remaining thirteen papers (Ashburner et al., 2008; Ausderau et al., 2014; Bitsika et al., 2016; Gal et al., 2010; Hilton et al., 2007; Lane et al., 2012; Liss et al., 2006; Mazurek & Petroski, 2015; Nadon et al., 2011; Sullivan et al., 2014; Tavassoli et al., 2014; Tseng et al., 2011; Wigham et al., 2015) documents stating children's and young people diagnosis were gathered or non 'gold-standard' tools were used to confirm diagnosis e.g. medical chart review. However, sample characteristics were not always well described. Five studies (Lane et al. 2012; Liss et al., 2006; Tavassoli et al., 2014; Tseng et al., 2011; Wigham et al., 2015) reported only gender and age of their participants. Only Ausderau et al. (2014) and Nadon et al. (2011) provided all the demographics selected in the evaluation grid characteristics (e.g. age, gender, ASD subtype, comorbidities, and demographic variables). The remaining studies reported three or four of these features.

2.3.3 Sensory measures section

Ten different tools were used to assess sensory processing pattern or patterns in the selected studies. Three authors (Boyd et al., 2010; Brock et al., 2012; Watson et al., 2011) used more than one sensory measure and selected items from each measure to inform a factor analytic model of sensory processing patterns. These models were informed with both observational data and parent reports, and in both studies further confirmatory factor analysis was performed to ensure appropriate model fit to the data (in Table 3 information on each measure separately rather than the final models can be found). Pfeiffer et al. (2005) used two measures depending on the age of their participants and Lane et al. (2012) used two tools, reporting their outcomes as equivalent to each other. Two different versions of the Sensory Experience Questionnaire were used across the studies, with the most updated version (SEQ-3.0) used in Ausderau et al. (2014). Additionally the Sensory Profile was used in two language versions – English and Chinese. Hence, overall there were 10 different sensory

measures used across the 29 selected papers, with the Sensory Profile and Short Sensory Profile being used most frequently.

In ten studies there was information about a sensory measure being standardized (Gal et al., 2010; Jasmin et al., 2009; Lane et al., 2010; Lane et al., 2012; Nadon et al., 2011; Pfeiffer et al., 2005; Reynolds et al., 2012; Tomchek et al., 2015; Watson et al., 2011; Wigham et al., 2015) with Liss and colleagues (2006) providing a reference to a current standardization work. Remaining studies did not report on the measures' standardization. Reliability was more often reported than validity of the measures, with four studies providing calculations of reliability – test-retest reliability (Baranek et al., 2013) and internal consistency (Bitsika et al., 2016; Green et al., 2012; Pfeiffer et al., 2005, but only for the Adolescent/Adult Sensory Profile, AASP). Only Pfeiffer et al. (2005) provided discriminative and convergent validity calculations (for the AASP). Across the papers included in the review, there was no information regarding reliability of nine of the referenced tools used compared to fifteen measures missing information on validity. Across the studies, four measures were referenced as being appropriate for use with ASD population or being ASD-specific (Sensory Processing Assessment, SPA; Tactile Defensiveness and Discrimination Test, TDDT-R; both versions of the Sensory Experiences Questionnaire, SEQ and SEQ-3.0; and Sensory Questionnaire, SQ). Sensory Profile and Short Sensory Profile, in five and six studies respectively, were reported as widely used within the ASD research.

2.3.4 Psychological correlate measure section

Thirty five different measures of psychological correlates were used in the reviewed papers. Some of the tools were used in several publications, Child Behavior Checklist (CBCL) was used in two language versions – English and Chinese, and Vineland Adaptive Behaviour Scales were used in their original version and newest revision (VABS and VABS-2), same as Repetitive Behaviour Questionnaire (RBQ and RBQ-2), resulting in 42 references to psychological correlate measures across selected papers. Only in seven papers (Bitsika et al., 2016; Hilton et al., 2007; Lane et al., 2010; Lidstone et al., 2014; Mazurek et al., 2013; Pfeiffer et al., 2005; Watson et al., 2011) some measures were reported as standardized (CASI-D, SRS, VABS, SCAS-P, PAS, CBCL, ABAS, MSEL and PLS-4). The remaining papers did not indicate standardization status of the tools used. In Liss et al. (2006) a tool measuring exceptional memory was used, however, no information on tool development, measurement properties or scoring criteria were given. Reliability calculations were performed for five tools: inter-rater reliability for the JAA (Baranek et al., 2013); RBQ-2 (Lidstone et al., 2014), EDI (Samson et al., 2013), internal consistency for CASI-D (Bitsika

et al., 2016) and test-retest for Eating Profile (Nadon et al., 2011). Structural validity was only calculated for the RBQ-2 in Lidstone et al. (2014) and CASI-D in Bitsika et al. (2016); and face validity for Eating Profile in Nadon et al. (2011). In the reviewed studies there was no information about reliability of the 13 referenced measures, and about the validity of 15 selected tools. Across the studies, eight measures were referenced as being appropriate for use with ASD population or being ASD-specific (GARS, GADS, GI SIQ, Eating Profile, ADOS, RBS-R, AQ and SRS), further ten were reported as widely used in ASD research or developmental disorders (JAA, EFT, VABS, KOS, CBCL, CASI-D, CSHQ, SCAS, IUS-P, RBQ).

2.3.5 Results – associations

The authors selected different sensory patterns for their investigation. Hyporesponsiveness was examined in Baranek et al. (2013); hyperresponsiveness in Green et al. (2012), Lane et al. (2012), Mazurek et al. (2013), Mazurek et al. (2014), Mazurek and Petroski (2015), Sullivan et al. (2014), Tavassoli et al. (2014); hypo-, hyper-responsiveness and sensation seeking in Boyd et al. (2010), Brock et al. (2012), Watson et al. (2011), a pattern combining under responsiveness and sensation seeking in Ashburner et al. (2008), Baker et al.(2008), Chen et al. (2009), Gal et al. (2010), Lane et al. (2010), Nadon et al. (2013), Samson et al. (2013), and sensory processing patterns from Dunn's model in Bitsika et al. (2026), Hilton et al. (2007), Jasmin et al. (2009), Lidstone et al. (2014), Reynold et al. (2012), Tseng et al. (2011). In Ausderau et al. (2014) hyporesponsiveness, hyperresponsiveness, sensory interests, repetitions and seeking behaviors and enhanced perception were examined. Tomchek et al. (2015) investigated hyporesponsivity and sensory seeking/distractibility, while Wigham and colleagues (2015) focused on over- and underresponsiveness. Liss et al. (2006) used terms sensory seeking and over- and under-reactivity, which were treated as synonyms of hyper- and hypo-responsiveness. Pfeiffer and colleagues (2005) examined hypo- and hyper-sensitivity which were treated same as hypo- and hyper-responsiveness. Some authors preferred using responsiveness, some responsivity – both were also treated as synonyms in this review.

In seventeen papers (Ausderau et al., 2014; Baranek et al., 2013; Bitsika et al., 2016; Boyd et al., 2010; Brock et al., 2012; Gal et al., 2010; Green et al., 2012; Hilton et al., 2007; Lane et al., 2010; Lane et al., 2012, Liss et al., 2006; Mazurek et al., 2014; Nadon et al., 2011; Reynolds et al., 2012; Samson et al., 2013; Sullivan et al., 2014; Tavassoli et al. 2014) investigation of associations between sensory processing patterns and a single psychological construct were carried out. Three of these papers have multiple hypotheses on the sub-

constructs of the phenomenon under investigation that were tested. Baranek et al. (2013) looked at joint attention and reported the results for both initiation of and response to joint attention. Brock et al. (2012) were interested in sensory patterns' association with several dimensions of temperament such as withdrawal, distractibility, persistence, or slowness to adapt; and in Liss et al. (2006) the concept of overarousal was characterised by overfocused behaviour, perseverative preoccupation and exceptional memory for self-selected material. In the remaining studies, the relationship between sensory processing atypicalities and two (Baker et al., 2008; Chen et al., 2009; Jasmin et al., 2009; Lidstone et al., 2014; Mazurek et al., 2013; Mazurek & Petroski, 2015; Tseng et al., 2011; Watson et al., 2011) or more (Ashburner et al., 2008; Pfeiffer et al., 2005; Tomchek et al., 2015; Wigham et al., 2015) constructs were explored. Data extraction was carried out for each construct separately and for this reason those papers investigating multiple constructs were included in the review results' sections more than once.

2.3.6 Participants

Across the 29 studies included in the review, a total of 7923 children and adolescents with ASD were included. One study recruited 2973 participants (Mazurek et al., 2013), two studies included over 1300 participants (Ausderau et al., 2014, and Mazurek & Petroski, 2015) the remaining studies involved between 22 and 400 participants.

The age of participants ranged from 20 months to adulthood. One study focussed particularly on toddlers (Green et al., 2012; with a mean of 28.2 months). Eleven studies (Ausderau et al., 2014; Baker et al., 2008; Baranek et al., 2013; Boyd et al., 2010; Brock et al., 2012; Jasmin et al., 2009; Lane et al., 2010; Nadon et al., 2011; Tomchek et al., 2015; Tseng et al., 2011; Watson et al., 2011) focussed on early and middle childhood (20 to 144 months). A further twelve studies (Ashburner et al., 2008; Bitsika et al., 2016; Chen et al., 2009; Gal et al., 2010; Hilton et al., 2007; Lane et al., 2012; Liss et al., 2006; Pfeiffer et al., 2005; Reynolds et al., 2012; Samson et al., 2013; Sullivan et al., 2014; Wigham et al., 2015) included children and adolescents between middle childhood and mid-teens (6 to 18 years). Four studies included both children and adolescents, Lidstone et al. (2014) recruited 3-17;9 years old participants, and Mazurek et al. (2013, 2014) and Mazurek and Petroski (2015) used a sample between 2 and 17 years old. One study focused specifically on the adult population (Tavassoli et al., 2015).

In all the studies, except for Tomchek et al. (2015), the gender of the participants was reported and 83.3% of participants were male. This percentage mirrors the widely reported

uneven sex ratio for the prevalence of ASD in males; with males being four times more likely to have this condition than females (Anello et al., 2009).

A minority of studies were highly selective when recruiting participants with a particular diagnosis. Pfeiffer et al. (2005) included only children and adolescents who had Asperger's Syndrome, while Hilton et al. (2007) included only children with High Functioning ASD and Tavassoli et al (2014) recruited only adults with autism spectrum condition. Chen et al. (2009) included those with a diagnosis of ASD or Asperger's Syndrome; Green et al. (2012) recruited toddlers with either autism or PDD-NOS; Jasmin et al. (2009) included in their study children with AD or PDD-NOS. In a couple of studies participants were characterised as diagnosed with autism (Tseng et al., 2011; Gal et al., 2010) and further five (Baker et al., 2008; Baranek et al., 2013; Boyd et al., 2010; Lane et al., 2010; Watson et al., 2011) included those with autistic disorder. In the remaining studies, participants fell into the general diagnostic category for ASD. Only Ausderau et al. (2014), Bitsika et al. (2016), Mazurek et al. (2013), Nadon et al. (2011) and Tomchek et al. (2015) reported an exact percentage of ASD children in each diagnostic category (AD, Asperger's disorder, PDD-NOS).

The method of reporting cognitive ability varied markedly across the reviewed studies. Ability in the form of an IQ score was reported by Bitsika et al. (2016) separately for younger and older groups, Lane et al. (2012), Mazurek et al. (2014), Mazurek and Petroski (2015) separately for two age groups, Reynolds et al. (2012), Samson et al. (2013) and Wigham et al. (2015), with the following means (standard deviations): 95.93 (12.98), 93.5 (11.44), 95.5 (18), 82.5 (23.0), 90.56 19.39), 85.56 (22.39), 95.88 (17.8), 82.75 (23.61) and 106.2 (14.79) respectively. Standard score of 61.3 (26.5) were reported in Jasmin et al. (2009). Green et al. (2012) stated nonverbal and verbal developmental functioning (78.1 (18.06) and 58.62 (25.15) of their participants, whereas Baranek et al. (2013), Boyd et al. (2010), Brock et al. (2012) and Watson et al. (2011) reported mental age (23.25 (14.04), 31.97 (20.84), 36.11 (19.88), 32.0 (20.6) respectively). Tavassoli et al. (2014) provided mean Raven score for their sample 50.1 (10.3). Ashburner et al. (2008) included only participants with IQ above 80, while Chen et al. (2009) and Hilton et al. (2007) included individuals with ASD with IQ above 70. In Ausderau et al. (2014) the IQ Proxy was stated (81.4 (28.8)). Mazurek et al. (2013) reported that 3.9% of their sample had an IQ lower than 70, while the remaining sample had IQ above 70. Sullivan and colleagues (2014) relied on parent-reports and according to that description 35% of their sample had their intellectual ability above average, 22% had average scores, mild impairment had 12% and significant impairment had

11% (intellectual ability of the 18.5% of the sample was unknown) Remaining authors did not provide any indicators of cognitive functioning of their participants.

Only three studies reported co-occurring medical conditions for their participants. Nadon et al. (2011) reported attention deficit disorder, hyperactivity and mental retardation as the most common co-occurring conditions, while in Hilton et al. (2007) attention deficit/hyperactivity disorder, learning disability, anxiety disorder, depression, and Tourette syndrome were reported as additional diagnoses. In Sullivan et al. (2014) comorbid diagnoses included sensory processing disorder, anxiety disorder, obsessive-compulsive disorder, dyspraxia or movement disorder, language disorder, cognitive delay, conduct of defiance disorder, seizure disorder, depression, dyslexia, Tourette's syndrome and bipolar disorder.

2.3.7 Psychological constructs

In the selected studies, the authors examined relationships between sensory processing patterns and a variety of psychological constructs. In order to present our findings in a systematic way, the papers have been grouped. In the most recent Diagnostic and Statistical Manuals of Mental Disorders, core features of ASD, such as impairments in the social use of both nonverbal and verbal communication and presence of restricted, repetitive patterns of behaviour, interests, or activities are diagnostic components for the disorder (APA, 2013). In addition to these core features that are present in individuals with ASD, a number of associated difficulties has been listed in the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV-TR, APA, 2000), these include emotional, attentional, cognitive and behavioural problems. The psychological constructs examined in the selected papers have been grouped accordingly, either belonging to the core features of ASD, such as social functioning and repetitive behaviours or characterised as associated conditions of ASD, e.g. affective and cognitive difficulties. As a result six main groups of psychological constructs were created: symptom severity, social functioning, restricted and repetitive behaviours, emotional and behavioural functioning, affective and cognitive symptoms, and physical skills.

In the identified groups the following constructs were included (as indicated by the authors):

- symptom severity: social communicative symptoms (Watson et al., 2011), social competence (Hilton et al., 2007), social symptoms/communication impairment (Liss et al., 2006), autism quotient and Asperger's disorder quotient (Ashburner et al., 2008), autism severity (Ausderau et al., 2014), autistic traits (Tavassoli et al., 2014);

- social functioning: language skills (Watson et al., 2011), language abilities (Baranek et al., 2013), social and communication adaptive skills (Watson et al., 2011) and joint attention (Baranek et al., 2013), social skills and communication (Tomchek et al., 2015);
- restricted and repetitive behaviours: restricted and repetitive behaviours (Chen et al., 2009; Boyd et al., 2010; Lidstone et al., 2014, Wigham et al., 2015) and stereotyped movement (Gal et al., 2010);
- emotional and behavioural functioning: emotional, behavioural, and educational outcomes (Ashburner et al., 2008), emotional and behavioural problems (Tseng et al., 2011), emotion dysregulation (Samson et al., 2013), adaptive/maladaptive functioning (Baker et al., 2008; Lane et al., 2010; Liss et al., 2006; Pfeiffer et al., 2005), behavioural responsiveness (Baker et al., 2008), gastrointestinal problems (Mazurek et al., 2013), eating (Nadon et al., 2011) and sleep (Mazurek & Petroski, 2015, Reynolds et al., 2012) problems;
- affective and cognitive symptoms:
  affective: temperament (Brock et al., 2012), anxiety (Green et al., 2012; Lane et al., 2012; Lidstone et al., 2014; Mazurek et al., 2013, Mazurek et al., 2014; Mazurek & Petroski, 2015; Pfeiffer et al., 2005; Sullivan et al., 2014, Wigham et al., 2015), depression (Bitsika et al., 2016; Pfeiffer et al., 2005), intolerance of uncertainty (Wigham et al., 2015);
  cognitive: memory (Liss et al., 2006), cognitive style (Chen et al., 2009), attention (Liss et al., 2006);
- physical skills: motor skills (Jasmin et al., 2009; Tomchek et al., 2015) and daily living skills (Jasmin et al., 2009).

Symptom severity

Six papers investigated associations between sensory atypicalities and symptom severity. Ashburner et al. (2008) found a significant negative correlation between the underresponsive / seeks sensation subscale of the Short Sensory Profile and GARS autism quotient (r=-.53 p=.003), but not with GADS Asperger's disorder quotient, suggesting more sensory problems being associated with more autism symptoms (low score on the SSP indicates more sensory issues). Ausderau et al. (2014) showed that autism severity measured with the SRS was significantly positively associated with all sensory response patterns calculated from the Sensory Experience Questionnaire (hyporesponsiveness: r=.57, p<.001, hyperresponsiveness: r=.50, p<.001, sensory interests, repetitions and seeking behaviors:

r=.50, p<.001, enhanced perception r=.33, p<.001). Hilton et al. (2007) reported significant associations between all sensory processing patterns as measured by the Sensory Profile and SRS scores, both total score (correlations with Sensory Sensitivity: r=-.745, p<.01, Sensory Avoiding: r=-.796, p<.01, Low Registration: r=-.578, p<.01 and Sensation Seeking: r=-.527, p<.01) and subscales, with the exception of SRS social awareness for which only sensory sensitivity and sensation avoiding were found to be negatively correlated. Liss and colleagues (2006) found significant correlations between overreactivity, underreactivity and sensation seeking and all the subscales of DSM-IV checklist. Only the DSM-IV communication impairment subscale was not significantly associated with overreactivity. Significant associations between sensory overresponsivity and autistic traits measured with the Autism Spectrum Quotient (r=.34, p<.001). Watson et al. (2011) used ADOS as one of the outcome measures in their study and found associations between social-communicative algorithm scores and both hyporesponsiveness (β=0.48, SE=.023, p=.040) and sensation seeking (β=0.78, SE=.025, p=.002).

Social functioning/social skills

The relationship between sensory processing patterns and verbal and nonverbal communication skills in individuals with ASD was investigated in three studies. Baranek and colleagues (2013) were interested in associations between sensory difficulties and language abilities and joint attention. Watson et al. (2011) explored the relationships between sensory atypicalities and language skills, social and communication adaptive skills. All verbal and nonverbal variables were associated with sensory hyporesponsiveness (Receptive language ratio scores: β=-2.0, SE=.68, p=.004, Expressive language ratio scores: β=-2.1, SE=.73, p=.005, Receptive Joint Attention: β=-0.83, SE=.37, p=.025, Initiating Joint Attention: β=-1.63, SE=.59, p=.006, Aggregate language quotient scores: β=-0.010, SE=.004, p=.018, Social adaptive scores: β=-0.017, SE=.007, p=.011). Also in Watson et al. (2011) language skills (aggregate language quotient scores) were correlated with sensory seeking (β=-0.011, SE=.004, p=.005). Tomchek and colleagues (2015) examined a contribution of sensory processing patterns to adaptive behaviour and receptive and expressive language, and found the same pattern of associations with hyporesponsivity and sensory seeking/distractibility associated with language limitations and restricted social behaviours. Only expressive language scores and sensory seeking/distractibility were not significantly associated.

Restricted and repetitive behaviours (RRBs)

Restricted and repetitive behaviours (RRBs) is a broad term which includes behaviours ranging from self-injurious behaviour and stereotyped motor mannerisms through insistence on sameness and circumscribed interests (Bodfish et al., 2000). Turner (1999) suggested distinguishing two levels of behaviours - 'lower level' including motor repetitions and stereotyped behaviours, and 'higher level' relating to insistence on sameness and circumscribed interests. This division of RRBs into two separate levels is present in the studies included in our review, hence we present the results distinguishing between 'lower' and 'higher' levels of RRBs.

Five papers looked at the relationship between sensory processing patterns and the presence of restricted and repetitive behaviours. Boyd et al. (2010) reported a significant association between hyperresponsiveness and stereotypy (β=3.40, SE=1.35, p=.012). Gal et al. (2010) found a significant negative correlation between the number of Different Stereotyped Movements and the underresponsiveness/seeks sensation subscale of Short Sensory Profile (r=-.43, p<.001). Lidstone at al. (2014) reported significant negative correlations between repetitive motor behaviours and sensation avoiding and sensation seeking (r=-.42, p<.01 for both). In the same study significant negative correlations were found between all sensory processing patterns and insistence of sameness (correlations with Sensory Sensitivity: r=-.43, p<.01, Sensory Avoiding: r=-.49, p<.01, Low Registration: r=-.38, p<.01 and Sensation Seeking: r=-.49, p<.01). Similarly, significant negative associations between repetitive motor behaviours and insistence on sameness and both sensory over- and underresponsiveness were reported in Wigham et al. (2015) study (for repetitive motor behaviours and hyper-responsiveness r=-.386, p<.01, and hypo-responsiveness r=-.695, p<.001; and insistence on sameness and hyper-responsiveness r=-.558, p<.001 and hypo-responsiveness r=-.358, p<.01). Chen et al. (2009), however, did not find any associations between under responsiveness/seeks sensation patterns and 'compulsive-like behaviours'. Compulsions were associated with hyperresponsiveness in Boyd et al. (2010) study (β=3.50, SE=1.41, p=.013). The authors found also significant associations between rituals and both hyperresponsiveness (β=4.47, SE=1.35, p=.001) and sensory seeking (β=5.92, SE=2.97, p=.046).

Emotional and behavioural functioning

Seven papers examined associations between sensory difficulties and the emotional and behavioural functioning of individuals with ASD. Ashburner et al. (2008) reported significant correlations between the underresponsive/seeks sensation subscale of the Short

Sensory Profile and three subscales of Conner's Teacher Rating Scale–Revised Long Version, cognitive problems/inattention (r=-.48, p<.01), social problems (r=-.32, p<.05) and inattentive (r=-.42, p<.05). They also found significant associations of the Short Sensory Profile under responsiveness/seeks sensation subscale and two of the subscales of the Achenbach System of Empirically Based Assessment: Teacher Report Form, namely thought problems (r=-.39, p<.05) and academic performance (r=.62, p<.01). Baker and colleagues (2008) using the Short Sensory Profile reported correlations with the following subscales of the Developmental Behaviour Checklist subscales: self-absorbed (r=-.523, p=.012), Autism Screening Algorithm (r=-.533, p=.011) and total score (r=-.491, p=.020); and maladaptive behaviour scale of Vineland Adaptive Behaviour Scales (r=-.482, p=.023). Lane et al. (2010) also found similar associations with the maladaptive behaviour scale of the VABS. Using the VABS, Liss et al. (2006) reported correlations between the socialization subscale and hyperresponsiveness (r=-.195, p<.05), VABS daily living and adaptive behaviour composites and hyporesponsiveness (r=-.326, p<.01 and r=-.221, p<.01respectively) and sensory seeking with the VABS communication (r=-.263, p<.01), daily living (r=-.165, p<.05) and adaptive behaviour composite (r=-.235, p<.01). Pfeiffer et al. (2005) investigated relationships between sensory processing patterns and adaptive behaviours. They found significant negative associations between both hypo- and hypersensitivity and community use (r=-.271, p=.05 and r=-.291, p=.041) and social skills subscales of the Adaptive Behavior Assessment System (r=-.298, p=.036 and r=-.278, p=.05 respectively). Samson and colleagues (2013) looked at emotion dysregulation and its relationship with sensory atypicalities, reporting higher emotion regulation difficulties in those individuals with ASD who also had high scores on the under responsive/seeks sensation subscale of the SSP (r=-.57, p<.001). Tseng et al. (2011) were interested in sensory processing dysfunction and children's emotional and behavioural problems. They used the Child Behavior Checklist to measure both internalizing and externalizing difficulties and found a number of significant associations of those dimensions with all the sensory processing patterns. Internalizing was negatively associated with Sensory Sensitivity: r=-.24, p=.047, Sensory Avoiding: r=-.43, p<.001, Low Registration: r=-.28, p=.020 and Sensation Seeking: r=-.43, p<.001, while externalizing correlated significantly with Sensory Sensitivity: r=-.30, p=.013, Sensory Avoiding: r=-.29, p=.016, and Sensation Seeking: r=-.29, p=.016.

The relationship between associated behavioural problems with ASD and sensory processing difficulties was investigated in four studies. Mazurek et al. (2013) reported that those children with ASD who had chronic GI problems such as chronic constipation, chronic abdominal pain, chronic bloating, chronic nausea, chronic diarrhoea had significantly lower

sensory overresponsivity scores (greater levels of overresponsivity) than those children with ASD who had no additional GI problems (d=-.36 to -.71, $p<.0001$). Nadon and colleagues (2011) did not find any significant associations between underresponsive/ seeks sensation subscale of the Short Sensory Profile and the mean number of eating problems in children with ASD. Reynolds et al. (2012) reported significant positive correlation between sensation avoiding and sleep problems ($r=.502$, $p=.11$), associations with other sensory processing patterns were not significant. Different sleep difficulties, such as bedtime resistance, sleep duration or sleep anxiety, were however significantly associated with sensory sensitivity in Mazurek and Petroski (2015) in both younger (2-5 years old) and older (6-18 years old) age group.

Affective symptoms

Eleven papers investigated the relationships between sensory processing patterns and affective symptoms such as dimensions of temperament, anxiety, depression and intolerance of uncertainty.

Brock et al. (2012) looked at how sensory atypicalities relate to temperament dimensions in children with ASD. Three out of the nine investigated dimensions were associated with only one particular pattern, namely hyporesponsiveness (adaptability: $\beta=0.38$, $p=.001$, distractibility: $\beta=-0.46$, $p<.0001$, reactivity $\beta=-0.28$, $p=.04$), reporting that children with ASD who show hyporesponsive behaviours, may be more susceptible to various distractions and their optimal level of engagement with the environment may be narrower, elongating adjustment to change.

In all nine papers in which the relationship between anxiety and sensory patterns was examined (Green et al., 2012; Lane et al., 2012; Lidstone et al., 2014; Mazurek et al., 2013; Mazurek et al., 2014; Mazurek & Petroski, 2015; Sullivan et al., 2014; Pfeiffer et al., 2005; Wigham et al., 2015), correlations between hyperresponsiveness and anxiety were found and reported by four authors although a number of different measures were used across the studies (Green et al., 2012: $r=.52$, $p<.001$(time 1) and $r=.60$, $p<.001$(time 2); Lane et al., 2012: $r=.18$, $p<.001$; Mazurek et al., 2013: $r=-.45$, $p<.0001$; Mazurek et al., 2014: $r=-.42$, $p<.001$; Mazurek & Petroski, 2015: $r=-.46$, $p<.001$ (2-5 years old), $r=-.39$, $p<.001$ (6-18 years old); Pfeiffer et al., 2005: $r=.476$, $p<.001$; Wigham et al., 2015: $r=-.350$, $p<.05$). Sullivan et al. (2014) found a similar pattern of association, although the study investigated the relationship between the hyperresponsiveness and generalized anxiety disorder specifically ($r=-.31$, $p<.01$). Lidstone at el. (2014) looked at dimensions of hyperresponsiveness (both sensory sensitivity and sensation avoiding), and further moderate

to strong correlations were reported (r=-.61, p<.01 and r=-.71, p<.01 respectively). Only Lidstone et al. (2014) stated a relationship between anxiety and low registration (r=-.40, p<.01). In Wigham et al. (2015) the relationship between hyporesponsiveness and anxiety was also examined, however, a non-significant association was found.

Depression was not only associated with hyperresponsiveness (r=.394, p=.005 for the total sample, and for the younger children r=.449, p=.013, but not for the teenage group) as reported by Pfeiffer and colleagues (2005), but also with hyporesponsiveness (r=.214, p=.05 for the total sample, non-significant associations for the younger children and significant for the teenagers: r=.492, p=.027), and hyporesponsiveness dimensions (low registration was significantly associated with depression only in the teenage group r=.483, p=.031, and sensation seeking correlated with depression when the total sample was used r=.299, p=.035). In Bitsika et al. (2016) participants of a similar age (6-18 years old) to Pfeiffer et al. (2005) were included. Sensory sensitivity, sensation avoiding and low registration were associated with depression scores, for both parent and self-reports (for parent reports: r=-.355, p<.01; r=-.315, p<.01; r=-.345, p<.01 and self-reports: r=.357, p<.01; r=.351, p<.01 and r=.406, p<.01 respectively). Sensation seeking was not, however, significantly correlated with depressive symptoms in that sample.

Only one study examined the relationship between the intolerance of uncertainty and sensory processing patterns. Wigham et al. (2015) examined both hyperresponsiveness and hyporesponsiveness patterns, but the significant association was found only between intolerance of uncertainty and hyperresponsiveness (r=-.356, p<.01).

Cognitive symptoms

The relationship between cognitive functioning and sensory abnormalities in children with ASD was examined in two studies. Chen et al. (2009) were interested in exploring the relationship between sensory difficulties and an individual's detail-focused cognitive style. Only non-significant associations between the under responsive/seeks sensation subscale of the Short Sensory Profile and the Embedded Figure Test were reported. Liss and colleagues (2006) looked at both ability to shift attentional focus and exceptional memory for self-selected material in individuals with ASD. They showed that although underreactivity and sensory seeking were significantly correlated with Kinsbourne Overfocusing Scale (r=.293, p<.01 and r=.235, p<.01 respectively), the strongest positive correlation was found with overreactivity (r=.608, p<.01). Overreactivity was also negatively correlated with the reverse log of the exceptional memory score (r=-.196, p<.05), showing an association between greater exceptional memory and individual's overreactivity to sensory stimuli.

Physical skills

Only one study investigated the relationship between sensory processing patterns and motor skills with daily living skills (as self-care skills) in children with ASD. Jasmin et al. (2009) reported significant correlations between only one sensory pattern, namely sensation avoiding and self-care domain of the WeeFIM (r=.388, p<.025), personal (r=.457, p<.011) and daily living skills (r=.372, p<.033) domains on the VABS-2. Also sensation seeking was positively correlated with gross motor skills as measured by PDMS-2 (r=.39, p<.03). The authors also looked at the association separately for AD and PDD-NOS groups. For the AD group significant correlations were reported between the sensation avoiding and self-care (r=.44, p<.04), personal (r=.56, p<.01) and daily living skills (r=.48, p<.02) domains; and between low registration and personal skills (r=.44, p<.05). For the PDD-NOS group, however, the only significant correlation was found between sensation seeking and the self-care domain (r=.71, p<.03). In addition, Tomchek et al. (2015) investigated a contribution of sensory processing patterns to gross and fine motor skills. That study reported significant associations between sensory seeking/distractibility and both gross and fine motor skills (β=.261, SE=.119, p<.05 and β=.257, SE=.118, p<.05 respectively), but no significant relationship was found for the hyporesponsivity and motor variables.

2.4 Discussion

This systematic review focused on 29 studies that examined relationships between sensory processing patterns and psychological constructs in individuals with ASD.

2.4.1 Evaluation grid - papers' quality

The evaluation grid was designed for the purpose of this review, although, it could be used in other reviews evaluating studies using correlational analysis methods in ASD research. The grid could be also easily adapted to be used in a wider context of developmental disorders research or even typical development. However, as it was used first time in the review, its validity is not established.

A confirmation of the diagnosis of ASD is provided in all the papers included in the review. Some authors selected participants with a particular ASD subtype, with most of the authors reporting their participants as children and/or young people with ASD. In the new DSM-5 (APA, 2013), all the ASD subtypes that were present in the previous version of the Manual (APA, 1994), namely, autistic disorder, Asperger's disorder, and PDD-NOS (pervasive developmental disorder not otherwise specified) were merged together under the umbrella of one term – autism spectrum disorder. This is important to bear in mind, because

findings from those studies, in which participants with only one ASD subtype were included, might be less generalizable to future studies, in which participants from across the spectrum will be included. Interestingly, in the study in which the results were presented for a total sample, and for two subtypes separately – AD and PDD-NOS groups (Jasmin et al., 2009), the findings differed for each subtype and for the total sample. ASD is a very heterogeneous disorder, with a diverse presentation across individuals. It seems therefore important to report both basic participants' characteristics such as gender and age, as well as features such as cognitive ability in order to make some comparisons and generalizations between and within such a varied population.

A wide variety of measures were used to assess sensory processing difficulties in individuals with ASD in the selected review papers. Some authors, however, did not report whether the selected tools were appropriate to use with this clinical population. Only a few were reported as widely used within the ASD population or were ASD-specific. Also, reliability and particularly validity of the tools were poorly reported. There is a lack of reliable and valid measures of sensory processing designed for use with ASD individuals. By using measures developed with and for typically developing individuals in ASD research without at least reporting their psychometric properties in this population, we have little evidence that the tools selected are appropriate. Therefore researchers should consistently report psychometric properties of the tools used in the sample selected. Moreover, there are a great number of questionnaires and observational measures of sensory atypicalities available for use for researchers. The decision regarding which tools should be used to examine a research question might be based on a number of reasons, e.g. the measures selected in the previous studies, common use of tools by particular research group. There is no consensus between researchers about which measures of sensory processing should be used in future studies. This lack of consensus on 'best-measures' makes the comparison and interpretation of the results, obtained by employing different measures, problematic. Time spent identifying and developing 'gold-standard' sensory processing measures would help in understanding and interpreting the findings. Some authors (Boyd et al., 2010; Brock et al., 2012; Watson et al., 2011) rather than using a single measure, developed a sensory processing model based on information obtained from a range of measures and informants. Through this approach the authors tried to overcome some limitations associated with using single, mainly parent-report based measures and they yielded stronger sensory constructs scores. Building the factor analytic models is an interesting suggestion in sensory atypicalities measurement field. The models not only conglomerate information from different measures, but also have excellent

structural validity scores. Researchers might consider implementing this form of measuring sensory processing patterns in their studies.

Across the ten instruments of sensory atypicalities used in the paper selected for this review, the Sensory Profile and Short Sensory Profile were most frequently used. It should be remembered, however, that the Short Sensory Profile provides very limited information in regards to sensory processing patterns of individuals with ASD. Researchers might consider using tools which provide information on at least hypo- and hyper responsivity to follow the dimensions of sensory atypicalities as suggested by DSM-5 (APA, 2013).

Thirty five different measures were used in the reviewed papers to assess psychological constructs in the ASD samples studied. Similar to the sensory processing measures, the psychometric properties of the selected tools were poorly reported (particularly the validity of the measures). Also their appropriateness for use with this particular population was not justified. If excluding those tools which were used for both diagnosis and outcome measurements, only eight were reported as appropriate for use in autism research. There is not only a lack of tools designed specifically to assess a number of problems associated with the ASD, but also a lack of consensus regarding which measures are best suited to each specific phenomenon. As a result, researchers use different measures to investigate the same constructs (e.g. anxiety). Interpretation of the results and their generalizability is therefore hampered. As already highlighted in the systematic review conducted by the MeASURe team (McConachie et al., 2015), for children with ASD under 6, psychometric work still needs to be done in order to select those tools which are reliable and valid within autism research.

2.4.2 Sensory processing patterns and correlates

Concentrating on two main dimensions of sensory responsiveness – hyper- and hypo-responsiveness, as distinguished and suggested in DSM-5 (APA, 2013), most of the measures of autism symptom severity were associated with hyporesponsiveness (GARS autism quotient in Ashburner et al., 2008; SRS in Ausderau et al., 2014; DSM-IV communication impairment in Liss et al., 2006; ADOS social-communication algorithm score in Watson et al., 2011; and DBS ASA in Baker et al., 2008). What is notable, however, is that in those papers investigating the relationships between symptom severity and sensory atypicalities, associations were found despite a wide range of symptom severity outcome measures being used, different groups included (HFASD in Hilton et al., 2007; ASD in Ashburner et al., 2008 and Liss et al., 2006; AD in Watson et al., 2011) and different age groups of participating children (although they all were up to 12 years old except the Tavassoli et al. (2014) study

that focused on adults). It might indicate that those sensory atypicalities were so prevalent that they could be detected across ASD subtypes and with different measures. However, when the SRS was used (Hilton et al., 2007), correlations were found with both hypo- and hyper-responsiveness, with the social awareness subscale correlating only with hyperresponsiveness. Also DMS-IV social symptoms subscale (Liss et al., 2006) was associated solely with hyperresponsiveness. Hyperresponsiveness was also associated with autistic traits measured with the Autism Spectrum Quotient (Tavassoli et al., 2014). Language and socio-communication variables (Baranek et al., 2013; Pfeiffer et al., 2005; Tomchek et al., 2015; Watson et al., 2011), joint attention (Baranek et al., 2013), stereotyped movement (Gal et al., 2010), a number of cognitive and social problems (Ashburner et al., 2008), maladaptive behaviours (Baker et al., 2008; Lane et al., 2010), some mood dimensions (Brock et al., 2012), emotion dysregulation (Samson et al., 2013) and gross motor skills (Jasmin et al., 2009; Tomchek et al., 2015) were all associated with hyporesponsiveness. On the other hand, self-care variables (Jasmin et al., 2009), anxiety (Green et al., 2012; Lane et al., 2012, Lidstone et al., 2014; Mazurek et al., 2013; Mazurek et al., 2014; Mazurek & Petroski, 2015; Sullivan et al., 2014; Pfeiffer et al., 2005; Wigham et al., 2015), intolerance of uncertainty (Wigham et al., 2015), socialization subscale on the VABS (Liss et al., 2006), GI problems (Mazurek et al., 2013) and sleep difficulties (Mazurek & Petroski, 2015; Reynolds et al., 2012) were correlated with hyperresponsiveness. Some variables were also associated with both sensory patterns, repetitive motor behaviours (Lidstone et al., 2014; Wigham et al., 2015), insistence on sameness (Lidstone et al., 2014; Wigham et al., 2015), depression (Bitsika et al., 2016; Pfeiffer et al., 2005), anxiety in Lidstone et al. (2014); attention (Liss et al. 2006), community use and social skills in Pfeiffer et al. (2005) study, and internalizing and externalizing scores (Tseng et al., 2011).

This evidence suggests that sensory hyporesponsiveness is more often associated with core features of ASD such as communication impairment, emotional, cognitive, behavioural problems while social awareness difficulties and affective disorders are associated with hyperresponsiveness. Similarly, Gay et al. (2008) suggested that hyporesponsiveness and sensory seeking may be more associated with difficulties in social-communication domains in children with ASD. That supports Baranek et al. (2006) findings proposing that sensory hyporesponsiveness discriminated individuals with autism from those diagnosed with other developmental disorders or typically developing individuals. However, investigating other sensory processing patterns in the light of the findings of this review seems as important. Not only are high frequencies of hyperresponsiveness also present in individuals with ASD, but also hypo- and hyperresponsiveness were reported to be present in the same individuals

(Baranek et al., 2006), and some associations between both hypo- and hyperresponsiveness and other ASD features remain unclear (e.g. repetitive behaviours). Hyperresponsiveness, nevertheless, seems to be an under-researched sensory pattern. For example, in the studies investigating associations between sensory processing patterns and anxiety, primarily the relationship between anxiety and over responsivity was examined. While the link between children's sensory over responsivity, negative reactivity to complex sensory events and anxiety, has been made in the reviewed papers, other associations were not explored. Only Lidstone et al. (2014) investigated other sensory processing patterns' associations with more sensory atypicalities than hyperresponsivity and did find significant associations between anxiety and other sensory processing difficulties. Furthermore, because researchers widely use the Short Sensory Profile which includes an under responsive/seeks sensation subscale only, finding and reporting associations with hyperresponsiveness is impossible.

It should also be noted, that in some papers relatively small sample sizes were used (Ashburner et al., 2008; Baker et al., 2008; Chen et al., 2009; Lane et al., 2012; Reynolds et al., 2012) and therefore type II error might have occurred and some of the associations might have not been detected although a relationship between sensory atypicalities and measured constructs could exist in the population (Field, 2009).

2.4.3 Study limitations

The current review highlighted that sample selection processes varied across studies. There was also a lack of consistency in the methods employed. First, in some reviewed areas, a small number of studies was included, which limits the conclusions that can be drawn. Secondly, studies with a wide age range of participants were often pooled together, ignoring possible age related differences in the presentation of both sensory atypicalities and ASD related difficulties. Thirdly, the wide variety of methods assessing sensory processing patterns and psychological constructs used in the reviewed studies made the interpretation of the results very difficult.

2.4.4 Conclusions

In summary, the current research reports a number of associations between sensory processing patterns and the clinical and non-clinical features of ASD, highlighting that sensory atypicalities play an important role in the disorder. However, there are several theoretical and measurement approaches to the classification of the sensory processing patterns. Consensus on using a singular theoretical framework and set measures would help with clarifying results, but should be preceded with more psychometric work. In the absence

of the agreement on measurement tools, multiple informant measures and sensory processing models based on information obtained from a range of measures and informants might be a bridging alternative.

There are also several questions that require further investigation. Hyperresponsiveness remains under-researched sensory processing pattern; hence, establishing its associations with psychological constructs is an apparent research need. The current evidence provided for some constructs (e.g. repetitive behaviours) has mixed findings. Further research examining these correlations and establishing whether there are clear associations with a particular processing pattern or whether some psychological constructs correlate with a number of sensory atypicalities, would benefit our understanding of the complexity of sensory processing difficulties in ASD.

Finally, at present, the research focuses on children and adolescents with ASD, without including adult participants in the recruited samples. Investigating associations between sensory processing patterns and psychological constructs in adults might shed some light into developmental changes of ASD characteristics.

Chapter 3. Sensory processing in Williams syndrome: a narrative review

3.1 Background

To recap, 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, p. 867). Under that broad term, therefore, a number of sensory features can be characterised, from discrimination of a single stimulus, as for example visual or auditory information to sensory modulation as an ability to regulate the degree by which an individual is affected by sensory information (Dunn, 1997; Gal et al., 2007; Mulligan, 2002). Sensory atypicalities are common amongst individuals with a number of neurodevelopmental disorders, including attention deficit hyperactivity disorder, Fragile X (Ermer & Dunn, 1998; Rogers et al., 2003), and are very common in autism spectrum disorder (Ben-Sasson et al., 2009) and WS (John & Mervis, 2010).

Interestingly, in WS hypersensitivity to certain sounds has been the main research focus as up to 85-95% of individuals with the disorder are sensitive to the sounds, such as those of machines, fireworks and bursting balloons (Donnai & Karmiloff-Smith, 2000). Only very recently have a broader range of sensory processing atypicalities been reported in the disorder (Riby et al., 2013; Rodgers et al., 2012).

3.1.1 Previous reviews

There are only a handful of reviews available on WS in general (e.g. Kaplan et al., 2001; Martens et al., 2008) with even more limited systematic presentation of findings on sensory processing or sensory aspects of the disorder. There has been a surge of research activity over the last two decades on sensory atypicalities in WS, which has made undertaking reviews more feasible. Kaplan et al. (2001) summarised clinical features of WS with only brief information on hyperacusis as affecting 95% of individuals with the disorder, which was described as painful by older children and with abnormal responses usually found to high-frequency auditory tones. Martens et al. (2008) presented the cognitive, behavioural, and neuroanatomical phenotype of individuals with WS. In that review musical skills, including absolute and relative pitch rather than sensory processing *per se*, were discussed. Auditory and visual processing were presented in a review conducted by Zarchi et al. (2010), however, the link between visuospatial ability and oversensitivity to sound, and the underlying structural and functional brain abnormalities were its main focus.

3.1.2 Aims

To date, very little is known about sensory processing in WS and the similarities of the sensory profile of individuals with WS compared to people with other neurodevelopmental disorders. Understanding sensory symptoms in WS would allow us to gain a better insight into strengths and weaknesses associated with this condition and further explore syndrome-specific characteristics that might guide clinical assessments, interventions and future research.

The current review therefore aims to summarise available evidence on sensory processing in WS, recognise and discuss any shortcomings, and identify goals for future research. Specifically, the review aims to: (I) explore the prevalence and phenomenology of sensory processing in WS; (II) assess the presentation of sensory processing in WS related to age, gender, intellectual ability; (III) assess the presentation of sensory processing in WS related to other clinical and behavioural features; (IV) compare sensory profiles of individuals with WS, those typically developing and with other neurodevelopmental disorders.

3.2 Method

The review followed a mixed methods model. A systematic literature search was undertaken to identify papers relevant to the review topic. Subsequently, a narrative approach was used to identify prominent themes in the literature and interpret the findings of the reviewed studies.

3.2.1 Inclusion/ exclusion criteria

Inclusion and exclusion criteria were defined prior to conducting the systematic literature search. Studies were eligible for inclusion if they reported data or information on sensory processing (including sensory modulation, processing patterns and modalities) in individuals with WS. Studies were not limited to any time frame, except the time limits specific to each of the databases. Non-primary studies were excluded from the search (e.g. reviews, book chapters). The search was not restricted to any particular age group. Case studies, if relevant, were included in the review.

3.2.2 Search Strategy

A systematic literature search aimed to identify studies investigating sensory processing of individuals with WS conducted up to 29th of March, 2016. The search used five electronic databases: Scopus, Web of Science, PsychInfo, Embase and Medline. Two of the

databases, Scopus and Web of Science, allow searching for a number of keywords. Combinations of the following search terms were used: a diagnostic term (Williams syndrome or Williams-Beuren syndrome or infantile hypercalcaemia as all three terms were used over years in relation to WS), a sensory term (sensitiv* or reactivity or processing or integration or modulation or sensory or stimul* or pattern* or input or event* or dysfunction or respons* or profile* or symptom* or unusual or difficulties or interest* or feature* or experience* or hypo* or hyper* or seeking or avoid* or registration or threshold* or defensiveness), and a modulation term (visual or tactile or auditory or propriocepti* or gustatory or vestibular or olfactory or vision or hearing or touch or smell or taste or balance).

In the other three databases (PsychInfo, Embase and Medline) the searches were based on controlled vocabularies. However, different types of headings were used for each database (e.g. medical subjects headings for Medline, but APA thesaurus for PsychInfo), hence the vocabulary used in the databases varied. For PsychInfo Williams syndrome was used as diagnostic terms, combined with sensory integration or intersensory processes, or perceptual motor processes, or sensorimotor measures, or sensory adaptation, or adaptation, or thresholds, or self stimulation, or perception, or perceptual stimulation, or tactual perception, or proprioception. In the Embase database, Williams-Beuren syndrome term was used, combined with sensory dysfunction or abnormal sensation, or sensory defensiveness, or sensory stimulation, or sensation, or abnormal sensation, or sensation seeking, or self stimulation, or perceptive threshold, or sensorimotor function, or sensorimotor integration, or sensory system, or hearing, or touch, or vision, or odor, or taste, or proprioception, or vestibular function, or loudness recruitment. When searching in Medline a combination of terms Williams syndrome and sensory thresholds or sensation disorders, or self stimulation, or occupational therapy, or sensation were used.

Additionally, a hand search of literature was performed to ensure that all the relevant papers were included in the review.

A flowchart of the search strategy and number of articles identified and excluded at each stage, and included in the final search, is outlined in Figure 3.1. All databases were searched up to the 29th of March 2016.

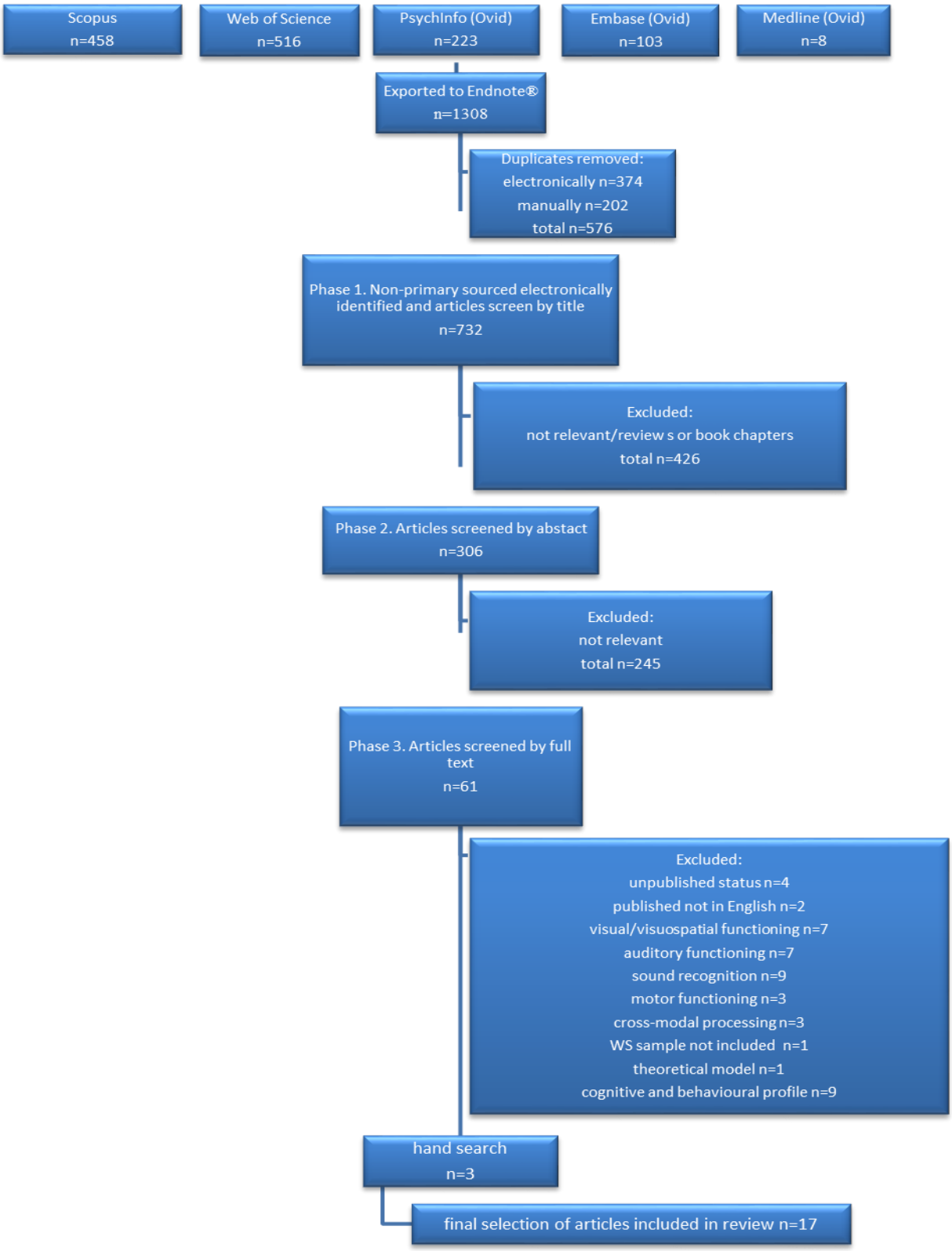


Figure 3.1 Flowchart of search

3.2.3 Electronic search

Results from five electronic databases were exported to Endnote® referencing software resulting in 1308 records in total. Most duplicates of the papers were identified by Endnote's duplicate identification function and removed from the record list. The duplicates not recognised by the software were searched for and removed manually, and 732 records were carried forward to the screening stage.

3.2.4 Screening of electronic search results

Screening of the search results consisted of three main phases. In Phase 1 the non-primary sources were removed and the remaining titles of the records were screened considering their relevance to the search question, and 426 studies were removed. In Phase 2 the remaining article abstracts were screened. Only sixty one articles met inclusion criteria and those were carried forward to the final Phase, in which articles were screened by full text and the final selection was made.

3.3 Results

3.3.1 Final selection

Forty six papers were excluded after screening the full text. Four papers were excluded due to lack of published status (conference papers and dissertation). Two were excluded due to being published in languages other than English (French and Italian). In one paper individuals with intellectual disabilities rather than WS were included. Another paper was found inappropriate due to lack of empirical data as only theoretical associations between genes and sensitivity to sounds were presented. Seven papers focused on visual or visuospatial functioning (such as pattern recognition) rather than sensitivity to everyday visual stimulation, which was a main interest of this review. In seven papers auditory functioning and in another nine papers sound recognition (including perfect pitch and timbre investigations) were reported. Three papers focused on motor functioning in WS and another three described cross-modal processing (such as audio-visual functioning). Cognitive and behavioural profiles of individuals with WS were investigated in a further nine papers. The remaining fourteen papers were included in the narrative review.

From the hand search an additional three papers met the inclusion criteria. In total, seventeen papers were included in the review. A summary of the descriptive characteristics of these studies can be found in Table 3.1.

Table 3.1. Summary of the papers included in the review

Authors	Research Question/Hypotheses	Participants	Methods	Findings	Conclusions
Hyperacusis					
Bedeschi et al. 2011	To investigate medical problems in WS	45 WS (23 m, 22 f) Age range: 17-44 years old (M=23.6 years)	audiograms	Audiologic problem were recorded in 13/45 subject. Five of them (38.4%) had been diagnosed with hyperacusia in infancy, and 8 (61.6%) in adulthood	The onset of hyperacusis in majority of WS patients was in adulthood
Blomberg et al. 2006	To investigate the prevalence of fear and hyperacusis and to explore the possible connections between fear, hyperacusis and musicality in a	38 WS (25 m, 13 f) M=21.00, SD=8.13 years	Fear Survey Schedule for Children – Revised (FSSC-R), Hyperacusis Questionnaire (HQ), Musicality Interest Scale (MIS)	Mean score on the HQ was 19.55 (SD=7.58) with 13% of the participants scoring above suggested cut-off for hyperacusis. There were many significant correlations between the HQ (total score and the attention, social aspects, emotional aspects subscales) and	A high reported prevalence of fear and hyperacusis was reported among the WS participants and correlations between reported fears and hyperacusis were found. Female individuals with WS (particularly adult women) had higher reported fears and

	Swedish sample of individuals with WS			the FSSC-R (total score and the danger and death, failure and criticism, the unknown, animals, agoraphobic situations subbscales), but very few between the HQ and the MIS	hyperacusis compared to male individuals with WS
Don et al. 1999	To examine the music and language skills in children with WS	19 WS (10 m, 9 f) Age range:8-13 years old (M=10y6m, SD=1y10m) 19 TD (11 m, 8 f) Age range:5-12 years old (M=7y11m, SD=2y5m)	Parent Music Questionnaire	History of hyperacusis evident for all of the WS group, but only for 10% TD group. All the WS children had unusual fearfulness toward sounds (comparing to 47% of TD children) and 75% exhibited unusual liking for specific sounds (comparing to 1 TD child)	The overwhelming prevalence of hyperacusis and unusual emotional responses to specific sounds were characteristic of the children with WS and distinguished them from the TD children

Einfeld et al. 1997	To assess psychopathology in WS	70 WS M=9.2 years old 454 TD M=12 years old	Developmental Behavior Checklist	80% of the WS participants covered ears to avoid particular sounds comparing to 35% of the TD participants (t=7.15, p<.001)	Children with WS reported hyperacusis more often than TD children
Elsabbagh et al. 2011	To assess the relationship between speech perception in noise in WS and their subjective rating of the severity of hyperacusis in everyday situations	32 WS Age range: 7.5-56.7 years old 32 TD Age range:7.4-11.8 year sold 24 TD Age range: 19.1-58.1 years old	Hyperacusis questionnaire, word-pairs discrimination/speech perception task	Higher severity of hyperacusis was associated with worse discrimination performance on the speech perception task	Hyperacusis influences speech perception, which may thus contribute to a qualitatively different process of language acquisition in WS

Gallo et al. 2008	To broaden the understanding of atypical behavioural reactivity to everyday sounds in the WS population using observational methods	21 WS (14 m, 7 f) Age range: 30-65 months old (M=44.48, SD=10.84 months) 20 mixed ethology (ME; 12 m, 8 f) Age range: 30-78 months old (M=44.8, SD=12.55 months)	Autism Diagnostic Observation Schedule-Module 1	Approximately 90% of the young children in the WS showed discomfort, fear, and/or anxiety in response to (or in anticipation of) everyday sounds, compared to 20% of mixed ethology controls. Over half the children with WS exhibited two or more different behaviours reflective of sound reactivity during the brief play interaction, compared to 15% in the ME group	Adverse reactions to sound were very common. A large proportion of the behaviours may be interpreted as ‘anticipations’, rather than ‘direct responses’ to aversive sound stimuli, highlighting the presence of anxiety that is pervasive among children with WS

Gothelf et al. 2006	To describe the clinical characteristics of hyperacusis and phonophobia in WS and to investigate the audiologic and neurologic abnormalities in subjects with WS and hyperacusis	49 WS (20 m, 29 f) Age range: 1-35 years old (M=11.1, SD=7.4 years)	Hyperacusis Screening Questionnaire	83.7% of WS participants were reported to be frightened or bothered by normal environmental sounds, the hyperacusis was most severe at age 5.7 +/- 3.8 years and tended to decline thereafter; children sensitive most frequently to electric machines, thunder, bursting balloons, and fireworks	Hyperacusis occurred in 84% of the participants; aversive responses to noise were present as early as infancy
Honjo et al. 2015	To investigate medical problems in WS	55 WS (34 m, 21 f) Age range: 2-30 years old	Clinical and laboratory assessments	Hyperacusis prevalent in 94.5% of the individuals	High prevalence of hyperacusis in WS
Klein et al. 1990	To obtain more data on the characteristic of hyperacusis	65 WS (36 m, 29 f)	questionnaire	Hyperacusis found in 95% of WS participants (and 12% of TD	High prevalence of hyperacusis in WS; many of the adverse reactions noted prior to 1 year of age,

		Age range: 1-28 years old 65 TD (32 m, 33 f) Age range: 2-17 years old		participants), with a slight decrease in severity with age	suggesting that hyperacusis may be an innate condition
Lense and Dykens 2013	To identify correlates of musical instrument learning	46 WS (47.8% m) Age range: 7-49 years old (M=23.13, SD=9.55)	Sensitivity to Sounds Questionnaire	The mean of the sound sensitivity score was 18.27, with SD = 6.61 (range 5-30)	The achievement of learning a new instrument was not associated with sound sensitivity
Lense et al. 2013	To examine how the auditory sensitivities and love of music that characterise WS relate to their variable musical	73 WS (49.3 % m) Age range: 10-51 years old (M=26.2,	Sensitivity to sounds questionnaire, musical questionnaires and behavioural assessments	The mean of the sensitivity to specific (non-musical) sounds was 50.1 (SD=19.1), and for the sensitivity to sound characteristics was 18.2 (SD=6.4). Musical interest was	Musical perception in WS is not related to general auditory sensitivities

	perception and production abilities	SD=9.4 years)		not associated with sensitivity to sound characteristics or sensitivity to specific (non-musical) sounds.	
Levitin et al. 2005	To clarify and document the incidence of auditory abnormalities in and among people with WS	118 WS (61 m, 57 f) M=20.4 (SD=10.4) years old 30 autism (24 m, 6 f) M=18.2 (SD=7.7) years old 40 Down syndrome (20 m, 20 f)	Open-ended questionnaire	4.7% of the WS sample reported true hyperacusis. 79.8% of the WS sample reported odynacusis (compared to 33% of the people with autism and Down syndrome, and 4% of the TD sample). Auditory aversions were reported by 90.6% of the WS sample (and 27% of the autism, 7% of the Down syndrome and 2% of the TD groups). There was a significant difference in the age of onset of auditory aversions with onset in WS occurring significantly	Unusual auditory behaviours were more common in the WS sample than in any other comparison group. The concepts of aversion, awareness and attraction seem to characterise the auditory abnormalities observed in WS

			M=17.2 (SD=9.2) years old 118 TD (28 m, 90 f) M=20.9 (SD=7.4) years old		earlier than in other groups. Auditory fascinations were found in 9% of the WS sample (and only 1 TD participant (0.8%) reported auditory fascinations.	
	O'Reilly et al. 2000	To examine how hyperacusis influenced operant responding under functional analysis assessment conditions for an individual with WS who exhibited problem behaviour	Case study (f) 5 years 2 months	Williams Syndrome Questionnaire and functional analysis	Little problem behaviour was observed during the functional assessment under the no-noise condition; during the noise condition, high level of problems were observed under the demand assessment condition, but little problem behaviour occurred during the attention and play conditions	Background noise seemed to influence responding under functional analysis conditions by increasing the aversiveness of task demands

Udwin 1990	To collect more information on adults with WS	119 WS (51 m, 68 f) Age range: 16y2m – 38y10m M=22y10m	Survey/questionnaires	110 adults (92%) were hypersensitive to sounds as children, and 93 of these (78% of the total group) remained hypersensitive as adults	The high rate of hyperacusis was found in the sample
Sensory processing					
Janes et al. 2014	To capture information about sensory processing experiences and repetitive behaviours	21 WS (12 m, 9 f) Age range: 6-15 years old (M=9.3 years)	Short Sensory Profile (SSP); Assessment of Sensory Processing, Repetitive Behaviour, Anxiety, Fears in WS - Semi-structured Interview (SRAF-SSI)	The majority of the sample experienced sensory processing difficulties. The areas of sensory processing most frequently endorsed by parents were vestibular, auditory, gustatory and proprioceptive hypersensitivities.	The sensory profile of children with WS is characterised by hypersensitivities. Visual and tactile processing was not frequently reported as problematic.
John and Mervis 2010	It was hypothesised that children with WS would demonstrate	78 WS (34 m, 44 f) Age range: 4-10.95 years	Short Sensory Profile (SSP), Behavior Rating Inventory of Executive Functioning (BRIEF),	Only 7 children (9.9%) were classified as ‘typical performance’ based on their SSP total score. 56.3 % was	Most children with WS demonstrated abnormalities in sensory modulation. The ability to use muscles to move, noticing

	symptoms of poor sensory modulation and that these sensory modulation abnormalities contribute to the phenotype	old (M=6.63, SD=2.14)	Children's Behavior Questionnaire (CBQ), Scales of independent Behavior – Revised (SIB-R), Conner's Parent Rating Scale – Revised (CPRS-R(L))	classified as definitely having overall sensory modulation issues and 33.8% as probably having overall sensory modulation issues. Definite abnormalities on the Auditory Filtering were found in 59% of the children, on the Low Energy/Weak in 64.1% and on the Under-responsive/Seeks Sensations in 62.8%. Executive functioning, temperament, adaptive functioning and problem behaviours were associated with sensory modulation difficulties	everyday sensory events and hypo- and hyper-responsiveness to sounds were the most problematic areas of sensory modulation. Children with high impairments in sensory modulation had significantly poorer executive functioning and adaptive functioning, had also more problem behaviours and more difficult temperament
Riby et al. 2013	To explore sensory processing abnormalities and	21 WS (12 m, 9 f)	Short Sensory Profile (SSP), Repetitive	A significant negative correlation between the total score of the RBQ and the total	Children with WS who experienced more sensory processing abnormalities

repetitive behaviours in children with WS. It was hypothesised that children with WS who demonstrated more sensory processing abnormalities would exhibit more repetitive behaviours	Age range: 6-15 years old (M=9.3 years)	Behaviour Questionnaire (RBQ)	score of the SSP (r=-.60, p=.01) Significant correlations existed between RBQ Repetitive Movement and 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 Under-responsive/Seeks Sensation subscale (r=-.54, p=.01). RBQ Sameness of Behaviours was significantly correlated with the Taste/Smell Sensitivity subscale (r=-.58, p=.01)	demonstrated more repetitive behaviours. Engagement in some of the behaviours reported in the RBQ Repetitive Movement subscale occur as a consequence of tactile sensitivity.

3.4 Discussion

The primary aim of this review was to explore sensory processing in WS. The focus was placed on (I) the prevalence and phenomenology of sensory difficulties, (II) differences related to age, gender and intellectual ability, (III) presentation of sensory processing in relation to other clinical and behavioural features, and (IV) comparison of sensory profiles of individuals with WS, those developing typically and those with other neurodevelopmental disorders. It is important to note, that interestingly, the papers included in the review clustered into two groups. The majority of the papers explored and discussed the phenomenon of hyperacusis in WS (n=14), and only three papers investigated sensory processing more broadly. The findings will be discussed for each group of papers.

3.4.1 Hyperacusis

Prevalence and phenomenology

In the reviewed papers the prevalence of hyperacusis ranged from between 4.7% (Levitin et al., 2005) to 100% (Don et al., 1999), with the majority of authors reporting prevalence rates above 80% (Einfeld et al., 1997; Gallo et al., 2008; Gothelf et al., 2006; Honjo et al., 2015; Klein et al., 1990; Udwin, 1990). However, the authors did not agree on the definition of the term hyperacusis. Don et al. (1999) and Klein et al. (1990) defined hyperacusis as "aversive reactions to sounds that do not cause such reactions in normal individuals" (Don et al., 1999, p.155). In Blomberg et al. (2006), Gothelf et al. (2006), O'Reilly et al. (2000) and Udwin (1990) hyperacusis is seen as "an oversensitivity or excessive perception of normal environmental sounds" (Gothelf et al., 2006, p.390). Hypersensitivity to certain sounds was also mentioned by Elsabbagh et al. (2011) in addition to fascination by sounds. Gallo and colleagues (2008) instead of using the term hyperacusis, which was for them associated with heightened sensitivity to sound and auditory abnormality, decided to refer to *sound reactivity* to describe a range of behaviours associated with exposure to sound. Lense and Dykens (2013) and Lense et al. (2013) followed the distinction made by Levitin and colleagues (2005). These authors distinguished four categories of abnormal reactions to sounds, including true hyperacusis - understood as lowered hearing thresholds, odynacusis – lowered uncomfortable loudness level, auditory allodynia – auditory aversions to or fear of certain sounds and auditory fascinations. Einfeld et al. (1997) used covering ears or avoiding particular sounds as a description of hyperacusis. No definition was provided by Bedeschi and colleagues (2011), who used term hyperacusia, which usually is associated with abnormal acuteness of hearing, increased sensation to sound (Ghanizadeh, 2008).

Across various studies, the term hyperacusis has been used inconsistently. Hyperacusis has a medical origin and is defined as abnormal sensitivity to sound (Dirckx, 2001; Venes et al., 2001), where the hearing threshold is lowered enabling individuals with hyperacusis to hear sounds that are too soft for other people to hear. Yet, as seen in the WS literature, the meaning of the term has been widened, used inconsistently and has become less clear. Aversive reactions to certain sounds (Don, 1999; Klein et al., 1990), or the opposite, fascination by particular sounds (Elsabbagh et al., 2011), move away from the original meaning and may hamper our understanding and the interpretation of the findings on hyperacusis in WS.

Subsequently, the measures used to explore hyperacusis in WS, varied greatly, from audiograms (Bedeschi et al., 2011), various questionnaires including, the Hyperacusis Questionnaire (Blomberg et al., 2006; Elsabbagh et al., 2011) and Sensitivity to Sounds Questionnaire (Lense & Dykens, 2013; Lense et al., 2013), through to the Autism Diagnostic Observation Schedule-Module 1 (Gallo et al., 2008). The range of measures used to determine the prevalence and phenomenology of hyperacusis in WS hinders comparisons between these studies and hampers us in estimating an accurate rate of hyperacusis present in the disorder. Furthermore, none of the articles provided reliability or validity data for their measures for the WS sample, or any psychometric information on the tools used. In most of the studies, bespoke, author-developed questionnaires and interviews were used, such as the Sensitivity to sounds (Lense & Dykens, 2013) questionnaire or the Hyperacusis Screening Questionnaire (Gothelf et al., 2006). The lack of psychometric properties of the tools used in WS further hampers the interpretation of the prevalence data and making comparisons across disorders.

Individual differences

Age

The majority of the studies recruited WS participants across different ages, with very wide age ranges such as 2-30 years old (Honjo et al., 2015) or even 7.5-56.7 years old (Elsabbagh et al., 2011). A broad recruitment strategy is not surprising considering the rarity of the disorder and the desire to include appropriately sized samples for analysis (Morris & Mervis, 1999; Strømme et al., 2002).

Five papers explored developmental changes in the presentation of hyperacusis in WS. Gothelf et al. (2006) recruited participants between 1 and 35 years old and reported that aversive responses to noise were present in infancy. They also found that hyperacusis was most severe in young childhood, at age 5.7 +/- 3.8 years, and tended to decline with age.

Similarly, Klein and colleagues (1990), whose participants were between 1 and 28 years of age, noticed that many of the adverse reactions to sounds were present prior to 1 year of age with a slight decrease in severity overtime. Some decrease in hyperacusis was also described by Udwin (1990). In that study 110 adults with WS reported being hypersensitive to sounds as children, and 93 individuals remained hypersensitive as adults. Children and young adults with WS, autism, Down syndrome and those typically developing took part in Levitin et al. (2005) study. A significant difference was found in the age of onset of auditory aversions among the participants. Onset in WS occurred significantly earlier than in other groups. In contrast, Bedeschi et al. (2011) using clinical interviews and audiograms, thus focusing on assessing responses to loudness and pitch, reported that only 5 out of 13 individuals with WS were diagnosed with hyperacusia in infancy and the majority of the sample (61.6%) was diagnosed in adulthood, suggesting that the onset of oversensitivity to sound was more common later in life. It is likely that the different pattern emerging across these studies reflects differences in methodologies and understanding of the term hyperacusis across the studies.

Although in the reviewed articles different questionnaires were used to obtain the information about developmental changes in hyperacusis and auditory atypicalities in WS, in most of them similar findings were stated. The authors agreed on a very early onset of hypersensitivity to sounds and a slight decrease in severity with age, with the exception of Bedeschi et al. (2011) study. Early presentation of oversensitivity to sounds and general sensory oversensitivity, with characteristic decreasing over age in the severity of the presentation, has been found not only in other developmental disorders, such as autism (for the review see Ben-Sasson et al., 2009), but also in typical development (Kern et al., 2007), hence this pattern in WS shows a general developmental trait. Bedeschi et al. (2011), however, relying on the audiologic examination, found more individuals suffering with hyperacusis later in age than in the childhood. These findings need to be replicated.

Gender and intellectual ability

In relation to gender playing a role in the hyperacusis, only Blomberg et al. (2006) reported that female individuals with WS displayed higher levels of fears and hyperacusis compared to male individuals with WS, even though cognitive and behavioural differences across gender are minimal (John & Mervis, 2010). None of the studies included in the review investigated the possible role of intellectual ability/disability in the presentation of hyperacusis. Chapter 1 emphasised that many individuals with WS have mild to moderate intellectual difficulties, but there is significant cognitive and intellectual heterogeneity within

the disorder (Donnai & Karmiloff-Smith, 2000). Intellectual functioning might be an important factor in hyperacusis presentation as it has been reported in other neurodevelopmental disorders such as ASD that more sensory atypicalities are present in those individuals with lower ability levels (Maskey et al., 2016). Both areas of research require further investigation so that the evidence can contribute to models / theories of hyperacusis in WS.

Presentation of hyperacusis in relation to other clinical and behavioural features

In five papers the associations between hyperacusis and other clinical and behavioural features were examined. Blomberg et al. (2006) explored the relationship between fear, hyperacusis and musicality in individuals with WS. The authors reported many significant correlations between hyperacusis and fears, but very few between hyperacusis and musicality. Associations between musical perception, musical instrument learning and auditory sensitivities were investigated in two further studies (Lense et al., 2013; Lense & Dykens, 2013). Neither musical perception nor the achievement of learning a new instrument were related to sound sensitivity in WS.

The investigation of the association between speech perception and subjective rating of the severity of hyperacusis in everyday situations in individuals with WS was the main focus in the Elsabbagh et al. (2011) study. It was found that hyperacusis influenced speech perception, with higher severity of hyperacusis negatively correlated with discrimination performance on a speech perception task. O'Reilly et al. (2000) in their case study examined the relationship between hyperacusis and problem behaviour. The authors reported that for their 5 years and 2 months old female participant who took part in three conditions of the study (play, attention and demand) increased level of problem behaviours were found in the increased noise condition only. However, caution is required when extrapolating results from a case study to other individuals with the disorder, especially due to within-syndrome heterogeneity.

Comparison of hyperacusis of individuals with WS, those typically developing and those with other neurodevelopmental disorders

Very few studies reviewed here included a comparison group. Don and colleagues (1999) recruited a control group of typically developing children matched on mental age to WS individuals. Hyperacusis was present in all of the WS group in contrast to only 10% of the typically developing group. It was also reported that all the children with WS were unusually fearful towards certain sounds, in comparison to 47% of typically developing

children. Interestingly, 75% of individuals with WS exhibited unusual liking for specific sounds, while only one typically developing child showed similar behaviour. Similarly, Einfeld et al. (1997) compared children with WS to their typically developing peers. The authors found that 80% of the WS participants covered their ears to avoid and limit particular sounds. The same behaviour was observed in just 35% of the TD participants when age, gender and intellectual ability were controlled for. The prevalence of hyperacusis in WS individuals and typically developing children and adolescents was also compared in Klein et al. (1990). Hyperacusis was found in as many as 95% of WS participants, age range between 1 and 28 years old (median 8 years), while only 12% of TD participants between 2 and 17 years old (median age of 7) reported being oversensitive to sounds. Levitin et al. (2005) compared WS individuals not only to typically developing participants, but also to those with autism and Down syndrome. They found that true hyperacusis was only present in the WS sample and odynacusis was very common in WS individuals compared to 33% of the people with autism and Down syndrome, and 4% of the TD sample. Auditory aversions were reported by 90.6% of the WS sample and were present in other groups, however less frequently (in 27% of the autism sample, 7% of the Down syndrome and 2% of the TD group). Auditory fascinations, although found in only 9% of the WS sample were much more WS specific as only one TD participant (0.8%) reported auditory fascinations and they were not found in any of the comparison neurodevelopmental disorder groups. Gallo et al. (2008) similarly to Levitin and colleagues (2005), compared individuals with WS to those with other neurodevelopmental disorders, however their comparison group was highly heterogeneous, including children with autism, Down syndrome, Kabuki syndrome, Isodicentric 15 and other developmental delays. According to the findings gathered through the Autism Diagnostic Observation Schedule-Module 1, approximately 90% of the young children in the WS and 20% of children in the mixed aetiology group, indicated discomfort, fear, and/or anxiety when presented with everyday sounds. Sound reactivity was found in over half the children with WS and 15% of the mixed aetiology group during the play sessions. The findings suggested that sensitivity to sound was more prevalent in WS than in other neurodevelopmental disorders.

In all five studies, the overwhelming prevalence of hyperacusis and unusual auditory responses to everyday sounds were found frequently in children and young people with WS and these features distinguished those with WS from typically developing individuals and those with other neurodevelopmental disorders, including autism and Down syndrome. These findings clearly contrast with the autism literature (see Chapter 2), where oversensitivity to sensory stimulation, including oversensitivity to sounds, is one of the diagnostic features of

autism spectrum disorder (DSM-5, APA, 2013) and individuals with WS are considered to be exhibiting autistic behaviours (i.e. hypersensititivty to sounds; www.autism.com). It is however, possible, as shown by the papers reviewed here, that individuals with autism are exhibiting WS features. Further work is needed to establish whether oversensitivity to sounds is specific to one of these conditions or whether it is a common feature present across different neurodevelopmental disorders.

3.4.2 Sensory processing

Prevalence and phenomenology

Two papers provided some indication of prevalence of general sensory processing difficulties in WS. In Janes et al. (2014) study it was reported that the majority of the sample scored within the 'definite difference' range on the Short Sensory Profile questionnaire, showing sensory processing atypicalities (it is worth noting here that in Janes et al. (2014) and Riby et al. (2013) the same sample was used, hence the same prevalence rate applies to Riby et al. (2013) indirectly). John and Mervis (2010) provided more detailed information, stating that 90.1% of children with WS in their sample showed atypical performance based on the Short Sensory Profile total score. They also described that over half of the children (56.3%) were reported as definitely having overall sensory modulation issues and further 33.8% showed probable overall sensory modulation issues. Furthermore, on the subscales Auditory Filtering, Low Energy/Weak and Under-responsive/Seeks Sensations over 50% of the children were classified as having definite abnormalities.

Although all the authors used the same measure of sensory processing - the Short Sensory Profile (SSP; Dunn, 1999), they used slightly different terminology for their constructs. Janes et al. (2014) and Riby et al. (2013) defined sensory processing, following Baker et al. (2008), as the way that sensory information is managed. John and Mervis (2010) on the other hand focused more on sensory modulation that they described as an efficient processing of sensory input, where "sensations from one or more sensory systems (e.g., auditory, tactile, vestibular) are detected and integrated allowing the body to regulate and manage sensory input from multiple modalities in a graded and adapted manner" (p.266). Sensory processing and sensory modulation are, however, associated. Sensory modulation, alongside sensory motor behaviours and sensory discrimination, form sensory processing (Miller et al., 2007), hence sensory processing is a broader term than sensory modulation. In the literature, however, the terms are often used as synonyms, which can make it more difficult to compare and interpret the presented findings.

It is worth mentioning, that the Short Sensory Profile, although as reported in Janes et al. (2014) and Riby et al. (2013), has good psychometric properties, including internal consistency for the subscales, inter-rater reliability, content and discriminant validity in the general population, the measure has not been validated for individuals with WS. Due to the rarity of the disorder and small sample sizes recruited for the individual studies, the psychometric work on the measures used with the WS population has been hampered; nevertheless, the reliability and validity of the tools should be established for future research.

Individual differences

Age, gender and intellectual ability

None of the studies investigating sensory processing in WS examined the possible role of developmental changes, gender or intellectual ability/disability in the presentation of sensory profiles. These areas of research require and deserve further investigation with sufficiently large samples.

Presentation of sensory processing in relation to other clinical and behavioural features

Janes et al. (2014) and Riby at al. (2013) were interested in exploring the relationship between sensory processing and repetitive behaviours. Janes and colleagues (2014) interviewed parents of children with WS using the bespoke Assessment of Sensory Processing, Repetitive Behaviour, Anxiety, Fears in WS - Semi-structured Interview (SRAF-SSI; Janes, 2010, unpublished document). Parents reported an association between sensory processing difficulties and repetitive behaviours. The support for that qualitative work can be found in Riby et al. (2013), who correlated scores obtained from the Short Sensory Profile and the Repetitive Behaviour Questionnaire (RBQ; Turner, 1995, 1999) and found a significant negative relationship between the total score of the RBQ and the total score of the SSP (r=-.60, p=.01), suggesting that the more repetitive behaviours a child was presenting, the more sensory processing difficulties they had (as a low score on the SSP indicates more sensory processing atypicalities); and a number of correlations between the subscales of both measures. John and Mervis (2010) investigated the relationship between sensory modulation difficulties and adaptive functioning. It was reported that the group of children with WS who had a higher severity of sensory modulation impairment, also showed more difficulties in executive functioning, temperament, adaptive functioning and problem behaviours compared to children with WS who did not have so many sensory modulation atypicalities.

These findings of a relationship between sensory processing and a number of clinical and behavioural features, where increased degree of sensory processing difficulty is

associated with more behavioural problems and higher severity of other clinical symptoms, are not unique to WS. Analogous patterns of relationships can be found in other neurodevelopmental disorders such as autism (for the review see Chapter 2) or Down syndrome (Bruni et al., 2010). However, the evidence of sensory processing features being related to behavioural and clinical symptoms in WS is very limited. A number of possible associations between sensory processing and other psychological correlates could be investigated, including anxiety, attention or emotional functioning, enhancing our understanding of the disorder as well as our theories of sensory processing.

Comparison of sensory processing of individuals with WS, those typically developing and those with other neurodevelopmental disorders

None of the studies included in the review, included a comparison group; neither a typically developing sample nor a sample consisting of participants with other neurodevelopmental disorders nor mixed aetiology. Investigating sensory profiles across different groups would allow researchers to specify the strengths and weaknesses of sensory processing in a disorder and to consider the theoretical links between sensory atypicalities and other WS features. Further work should be done in this particular area.

3.4.3 Limitations of the review

This review has several limitations. First, different terminology and a variety of methodologies were used across the studies included in the review. Although it might have impacted on the clear understanding of the field and interpretation of the findings, it supports the narrative approach undertaken in this review. Implementing a fully systematic method could increase reviewer bias and reduce the review's replicability. The narrative approach enabled the reviewer to present the outcomes of the studies and to draw together the major concurrent themes presented in the literature, and to summarise the research findings in a comparable manner across two main clusters of papers that were identified – hyperacusis and sensory processing. The systematic approach would not be appropriate to use, especially in the relation to sensory processing studies, as the number of the papers that met the inclusion criteria was very small.

Secondly, parent-reports were the main, or often only, source of information regarding sensory processing in individuals with WS. Parents might be more aware of their child's problems when the child is older and can express their difficulties better. Especially in young children direct assessments should be undertaken and information combined with

parental data in order to fully understand sensory processing in WS at the early stage of development (and indeed across the lifespan).

Thirdly, only papers written in English were included in the review due to limited access to translation. It is possible that some relevant papers presented in other languages were excluded from the review. Furthermore, the review was performed by only one reviewer. The potential bias of the author was however minimized by the systematic approach used to identify relevant studies for review.

3.4.4 Conclusions

In summary, the current research on sensory processing in WS is dominated by studies investigating hyperacusis and only a handful of papers have examined broader sensory processing issues / characteristics. Nevertheless, a high prevalence rate of both hyperacusis and sensory processing difficulties was reported in the reviewed studies and these were associated with younger age, were discriminant between other developmental disorders and typically developing samples, and were associated with more behavioural problems and greater severity of other clinical symptoms. Several theoretical and measurement approaches to the classification of hyperacusis and sensory processing were used. Consensus on a single theoretical framework and gold standard measures would help with understanding and interpretation of the results. This research should be underpinned by psychometric work on sensory processing tools, an endeavour that has never been undertaken in WS research. Furthermore, further research should focus on developing a better understanding of sensory processing difficulties and their impact on everyday life and functioning of individuals with WS, across different ages, genders, and levels of intellectual abilities/disabilities. Sensory profiles, are yet to be determined in WS.

Chapter 4. Developmental changes in sensory profiles in autism spectrum disorder and Williams syndrome

4.1 Background

As shown in Chapters 2 and 3, sensory atypicalities play an important role in the manifestations of both ASD and WS; yet our understanding of sensory processing difficulties and their impact on everyday life and functioning of individuals with ASD and WS across different ages is still limited. There is some indication in the literature that sensory symptoms in individuals with ASD might change with chronological age; yet the findings are inconclusive, and the direction of the association between sensory symptoms and age remains unclear. Kern et al. (2007) recruited 103 participants diagnosed with ASD, aged 3 to 43 years of age, and age- and gender-matched typically developing individuals, and used the parent-report Sensory Profile. Interestingly, Sensation Avoidance behaviours were the only sensory processing pattern which occurred as frequently in younger as in older participants with autism, in contrast to the behaviours associated with the Low Registration, Sensation Seeking, and Sensory Sensitivity frequency lessened as participants got older. In contrast, Talay-Ongan and Wood (2000) investigated hypo- and hyper-sensitivities in 30 ASD children between 4 and 14 years of age using the self-developed tool the Sensory Sensitivity Questionnaire - Revised. The authors reported an increase of sensory sensitivities over age in their sample. Interestingly, Adamson et al. (2006), who used the Short Sensory Profile in 44 children with ASD, found no relationship between chronological age and the severity of sensory difficulties. Ben-Sasson et al. (2009) in their meta-analysis of sensory modulation symptoms in individuals with autism were particularly interested in examining whether chronological age could contribute to nature of the presentation of the sensory symptoms. The authors divided the reviewed studies into four age categories: up to 3.4 years; under 6.5 years old; between 6.5–9.5 years old and above 9.5 years old. They reported that an increase in hyperresponsivity and sensation seeking could be observed up to age of 6-9 years, with a decrease in those sensory behaviours at later ages. The authors were not able to describe a consistent pattern for hyporesponsivity based on research findings in autism.

To our knowledge, it is not known whether sensory symptoms in WS change their intensity or frequency over age. None of the studies investigating sensory processing in WS examined the possible role of developmental changes in the presentation of sensory difficulties (Janes et al., 2013; John & Mervis, 2010; Riby et al., 2013). Some indication of a possible relationship between sensory symptoms and chronological age comes from studies

focusing on hyperacusis in WS as reviewed in Chapter 3 and suggesting an early onset of hypersensitivity to sounds in that condition.

Considering that difficulties with sensory processing have been associated with problems with managing daily life in autism (Dunn, 2001; Kern et al., 2006), it is crucial to understand which sensory processing patterns could impact everyday functioning at different ages. Also, around 40% of individuals with autism experience comorbid learning disability so it is important to determine whether there is any within syndrome variability in sensory processing atypicalities based on ability. There is no evidence to date regarding how learning disabilities may impact on sensory processing patterns in individuals with ASD, hence investigating a possible influence of learning disability on sensory atypicalities in ASD would benefit our current knowledge and understanding of this condition. Additionally, investigation of sensory processing patterns or modalities, using the Sensory Profile, has never been undertaken in WS and our knowledge of sensory atypicalities in this disorder is very limited. Providing further evidence on sensory issues in WS and investigating any age-related changes would allow us to gain a better insight into difficulties associated with this condition. Furthermore, comparing sensory patterns and modalities of individuals with ASD (with and without additional learning disabilities), to those presented in WS and in typically developing children, will allow us to explore syndrome-specific characteristics which are crucial to the formulation of theories of sensory processing.

To better understand sensory processing in ASD and WS, the aims of this study were two-fold, to investigate:

- The sensory processing patterns and modalities from Dunn's model of sensory processing in autism and WS,
- Determine any age related differences within the sensory processing patterns and modalities in order to examine possible developmental changes within sensory processing trajectories. To facilitate making comparisons with previous findings, the distinguished age groups related to those reported in Ben-Sasson et al. (2009).

It was predicted that sensory processing patterns and modalities would be atypical in the ASD and WS groups compared to typically developing individuals, with ASD and WS children having significantly lower scores on the SP than TD children. It was also predicted that atypical sensory patterns and modalities in individuals with autism and WS would be lower in the older age range age, however compared to typically developing individuals their sensory symptoms would remain within an atypical range at all age ranges.

4.2 Methods

4.2.1 Recruitment

Parents of typically developing children, those with ASD or WS, between 3 and 16 years of age were invited to take part in the 'Sensory Hotspots in Children' study (all recruitment documents related to this study are included in Appendices A-F). Parents of typically developing children were recruited via the Newcastle University research volunteers' database and word of mouth. Parents of children with ASD were recruited via ASD-UK (www.ASD-UK.com), a major UK family research database of children with ASD (Warnell et al., 2015). WS families were approached via the Williams Syndrome Foundation, which supports research into social aspects of the condition and enables researchers to contact individuals with WS and their families. Additionally, data obtained from the 'Touch, hear, react' study, that aimed to recruit children between 4 and 9 years of age, were included in the dataset (see Appendices G-L). For that study parents of typically developing children were recruited via local schools (based in the North East of England). Parents of children with ASD were recruited via local mainstream and special schools; and via 'Contact a family' which is a national charity for families with disabled children. WS families were again approached via Williams Syndrome Foundation. In both studies children with any other comorbid diagnosis of neurodevelopmental disorders or with visual, hearing or motor impairments were excluded. For ASD participants recruited to the 'Sensory Hotspots in Children' study additional LD were noted if previous diagnosis was confirmed by the parents. Those recruited to the 'Touch, hear, react' study, had undergone cognitive ability assessments (Raven's Coloured Progressive Matrices, RCPM; Raven et al., 1998 and British Picture Vocabulary Scale - Third Edition, BPVS3; Dunn et al., 2009) and if a score below 70 was obtained on both measures, they were considered as having additional LD.

Families whose children met the study criteria were initially sent information about the study by email or letter, and reminders were sent to non-responders. Parents participated on a voluntary basis. Newcastle University Faculty of Medical Sciences Ethics Committee granted favourable ethical opinion.

4.2.2 Participants

After merging both datasets, the data consisted of 55 parents of typically developing children, 38 parents of children with WS and 80 parents of children with ASD (23 typically developing children, 17 children with WS and 23 children with ASD were recruited to the 'Touch, hear, react' study, the remaining children were recruited through the 'Sensory

hotspots' study). Eleven typically developing children were removed from the dataset, 5 due to their Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997) total scores which were within abnormal range (17 or above) and a further 6 due to a high percentage of missing data. Three children with WS were recruited to both studies, hence only data obtained for the 'Sensory Hotspots in Children' study were included. Another 3 WS children did not have a complete dataset. The LD status of two ASD children was unclear and a further 5 children did not have complete data. The final samples consisted of 44 typically developing children, 32 children with WS and 73 children with ASD of whom 37 had additional LD.

All children with ASD had previously been diagnosed with ASD based on a multidisciplinary team assessment following the guidelines of the UK National Autism Plan for Children (Le Couteur, 2003) as stated by the parents. Additionally, for 58 children with ASD data from the Social Responsiveness Scale - Second Edition (SRS-2; Constantino & Gruber, 2012) were available (for 15 children total raw score was not calculated due to a large amount of missing data, due to the presence of LD making the items inappropriate), with a range between 58 and 176, mean=114.09, SD=29.95. Only 8 children fell into mild to moderate range, with 50 into severe range. Children with ASD for whom the SRS-2 total score could not be calculated, did not differ on gender ($t_{(34.459)}$=-1.639, p=.110), but did differ on age ($t_{(71)}$=-2.754, p=.007) compared to children for whom the SRS-2 data were available. Those for who the data were not available were significantly younger (mean=69.80 months old, SD=37.87) than children for whom the SRS-2 data were available (mean=97.84 months old, SD=34.46).

All WS children had previously been clinically diagnosed with the diagnosis confirmed by positive fluorescent in situ hybridization testing (FISH). Moreover, the SRS-2 data for 27 children with WS were also available and the raw total scores ranged from 34 to 146, mean=88.22, SD=28.23. Three children fell within normal, 12 within mild to moderate and 12 within severe range. Five children with WS for whom the SRS-2 total score could not be calculated due to large amount of missing data did not differ on gender ($t_{(30)}$=.325, p=.747) or age ($t_{(30)}$=.600, p=.553) compared to children for whom the SRS-2 data were available.

Data on the Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997) were available for all 44 TD children, who obtained scores within the normal range (0-13; mean=6.82, SD=3.71).

4.2.3 Measures

The parents were asked to complete the following questionnaires:

1. Sensory Profile (SP; Dunn, 1999) - a caregiver questionnaire that measures a child's sensory processing abilities. The questionnaire consists of 125 items, rated on a five-point Likert scale, ranging from almost never to almost always. The measure is divided into three main sections: Sensory Processing, Modulation, and Behavioural and Emotional Responses and 14 sensory processing categories (e.g. auditory, visual, tactile). Children can also be classified as fitting into one of the four general sensory processing patterns: sensation seeking, sensation avoiding, sensory sensitivity, and low registration. The SP is commonly used with 3 to 10 year olds; however it has been used with older ASD participants (in Kern et al., 2007 the oldest participant for who the SP was filled in was 43 years old). Cronbach's alpha, as reported in the manual, ranged from .47 to .91 across different subscales and the tool is reported to have a good convergent and discriminant validity (SP; Dunn, 1999).
The completion of the SP usually does not exceed 25 minutes (see Appendix M).
Parents of children with ASD and WS also completed:

2. Social Responsiveness Scale – Second Edition (SRS-2; Constantino & Gruber, 2012) - a 65-item rating scale which takes 15–20 minutes to complete. It is a parent-report of autistic traits that covers unusual interpersonal behaviours, communication or repetitive/stereotyped behaviours. The SRS-2 describes a degree of autistic social impairment and the severity of autistic symptoms (Appendix N).

To ensure that the typically developing children included in the study did not experience any emotional, social or behavioural problems, parents of those children were also required to complete:

3. The Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997) – a 25-item caregiver-report of children of 4-16 years old that screens whether the child has any emotional, conduct, hyperactivity/inattention, and peer relationship problems or displays prosocial behaviour. It takes between 5 and 10 minutes to complete the form (Appendix O).

4.2.4 Data analysis

The data were analysed using SPSS version 22. To reduce the likelihood of type I error, two multivariate analysis of variance (MANOVA) were conducted with the four sensory processing patterns and five sensory modalities as the repeated measures, with diagnostic group (WS, ASD with LD, ASD no LD, TD) and age (3 categories: under 6,5 years old; between 6,5–9,5 years old; above 9,5 years old) as between-group factors. To test for specific differences within each of the sensory processing patterns and modalities, individual analysis of variance (ANOVA) was conducted with each of the sensory measures.

4.3 Results

The analyses were performed on the selected participants. The participants' characteristics are presented in Table 4.1. Also in Table 4.1 'typical performance' as indicated in the manual was included (Dunn, 2006). Participant characteristics for each age category are reported in Table 4.2.

Table 4.1 Descriptive statistics (mean (SD)) of participants' characteristics

	TD (n=44)	WS (n=32)	ASD no LD (n=36)	ASD with LD (n=37)	Typical performance
Gender: male	26	15	26	34	
Age in months	92.98 (32.00)	97.69 (37.41)	85.08 (28.53)	98.89 (42.55)	
Age range	51-159	49-181	36-161	36-184	
Sensory Profile					
Registration	68.93 (6.38)	43.25 (12.97)	53.72 (11.78)	54.62 (12.67)	72-64
Seeking	111.40 (13.20)	88.61 (17.81)	82.75 (19.79)	83.19 (15.89)	123-103
Sensitivity	88.86 (8.70)	63.48 (14.18)	66.64 (14.98)	65.29 (10.88)	94-81
Avoiding	122.56 (12.83)	90.53 (17.42)	90.09 (17.66)	94.70 (16.98)	133-113
Auditory	33.18 (4.91)	21.31 (6.16)	20.94 (6.66)	22.29 (5.32)	38-30
Visual	37.89 (4.54)	28.53 (6.52)	30.47 (6.66)	30.34 (6.05)	41-32
Vestibular	49.64 (4.97)	41.72 (6.99)	42.06 (7.32)	41.89 (7.07)	55-48
Touch	81.48 (7.54)	64.53 (12.23)	59.53 (14.84)	60.62 (12.95)	88-73
Oral	53.30 (6.11)	40.58 (11.55)	41.28 (11.34)	39.00 (11.52)	59-46

Table 4.2 Descriptive statistics (mean (SD)) of participants' characteristics for each age category

	TD (n=44)			WS (n=32)			ASD no LD (n=36)			ASD with LD (n=37)		
	under 6,5	6.5-9.5	above 9.5	under 6.5	6.5-9.5	above 9.5	under 6.5	6.5-9.5	above 9.5	under 6.5	6.5-9.5	above 9.5
n	19	14	11	11	11	10	15	14	7	14	9	14
Gender: male	12	8	6	3	6	6	10	10	6	13	7	14
Sensory Profile												
Registration	67.74 (4.29)	68.57 (9.80)	71.45 (2.70)	39.27 (10.29)	47.64 (14.66)	42.80 (13.41)	58.20 (11.03)	52.43 (9.51)	46.71 (14.77)	56.86 (10.72)	54.11 (15.30)	52.71 (13.26)
Seeking	105.28 (12.48)	112.14 (13.30)	120.45 (8.90)	84.30 (21.34)	88.73 (13.97)	92.80 (18.58)	87.20 (22.98)	78.93 (17.39)	80.86 (17.58)	89.23 (13.71)	81.56 (14.23)	78.64 (17.92)
Sensitivity	86.37 (7.88)	88.21 (10.76)	94.00 (4.67)	63.20 (15.40)	63.09 (13.01)	64.20 (15.63)	72.60 (15.39)	62.71 (12.81)	61.71 (15.75)	63.45 (9.15)	65.67 (11.29)	66.50 (12.37)
Avoiding	119.72 (11.23)	125.07 (15.45)	124.00 (11.89)	84.91 (17.11)	93.00 (15.16)	94.00 (20.13)	96.53 (20.11)	85.15 (14.09)	85.43 (15.63)	94.36 (15.46)	91.11 (18.33)	97.46 (18.10)

Auditory	30.53	34.79	35.73	18.73	21.64	23.80	24.80	17.86	18.86	21.92	20.33	23.86
	(4.53)	(4.59)	(3.90)	(6.00)	(5.87)	(6.11)	(7.04)	(4.50)	(5.87)	(5.07)	(4.58)	(5.82)
Visual	35.63	39.36	39.91	27.27	30.82	27.40	32.67	30.64	25.43	30.67	29.78	30.43
	(4.59)	(4.03)	(3.53)	(7.07)	(4.36)	(7.75)	(6.94)	(4.65)	(7.64)	(5.38)	(5.02)	(7.44)
Vestibular	48.53	49.07	52.27	40.82	43.27	41.00	42.93	42.14	40.00	43.71	40.44	41.00
	(4.80)	(6.11)	(2.37)	(7.72)	(5.50)	(8.01)	(7.59)	(5.96)	(9.71)	(6.46)	(9.11)	(6.30)
Touch	79.32	81.07	85.73	63.91	65.82	63.80	65.67	56.57	52.29	62.00	60.11	59.57
	(6.05)	(10.16)	(3.95)	(10.13)	(10.19)	(16.81)	(15.81)	(11.59)	(15.31)	(15.37)	(10.96)	(12.29)
Oral	51.95	53.21	55.73	41.00	36.73	44.40	43.73	38.07	42.43	41.62	43.33	33.79
	(5.78)	(6.75)	(5.57)	(15.32)	(8.28)	(9.97)	(10.15)	(12.34)	(11.77)	(12.54)	(8.41)	(10.94)

First, one way ANOVA analyses were performed to establish whether the groups differed in age and gender. The main effect of age was not significant, $F_{(3, 137)}$=1.12, p=.343. A main effect of gender was found, $F_{(3, 137)}$=6.68, p<.001 with significantly more male participants in the ASD with LD group than in both WS (p<.001) and TD (p=.006) groups as indicated by the post-hoc Tukey HSD test.

The normality of the data and homogeneity of variance were tested thereafter. As indicated by the Kolmogorov-Smirnov test of normality, the distribution of the data was non-normal in the TD sample for all variables except Visual, Seeking and Avoiding scores ($D_{(43)}$=.11, p=.200; $D_{(43)}$=.09, p=.200 and $D_{(43)}$=.09, p=.200 respectively). For the ASD with LD group data were non-normally distributed for Registration and Seeking scores ($D_{(32)}$=.22, p=.001; $D_{(32)}$=.20, p=.003). For some of the sensory variables, the variances were equal, as indicated by the Levene's test, Seeking $F_{(3, 137)}$= 1.91, p=.131; Avoiding $F_{(3, 137)}$=1.31, p=.274; Auditory $F_{(3, 137)}$=1.36, p=.257; and Visual $F_{(3, 137)}$=1.20, p=.313. However, for the remaining variance were significantly different in the four groups, Registration $F_{(3, 137)}$=7.78, p<.001; Sensitivity $F_{(3, 137)}$=5.52, p=.001; Vestibular $F_{(3, 137)}$=3.93, p=.01; Touch $F_{(3, 137)}$=7.73, p<.001; Oral $F_{(3, 137)}$=6.51, p<.001. Due to a number of variables being non-normally distributed and heterogeneous, and the violation of assumptions of normality and homogeneity, the bootstrapping procedure was implemented.

To control for the use of multiple outcome measures, a MANOVA was conducted twice using the sensory processing patterns and sensory modalities as repeated measures. There was a significant sensory processing pattern by group effect ($F_{(9, 51)}$ = 2.76, p = 0.01), indicating that there were significant differences between the participants across different diagnostic groups on sensory processing patterns scores. A significant main effect of age or interaction between group and age were not found.

In addition a sensory modality by group significant effect ($F_{(12, 72)}$=7.51, p<.001) was found. Also a significant interaction was found between sensory modality and age ($F_{(8, 72)}$=2.40, p=.024). To test for specific differences, a two-way ANOVA was conducted separately for each of the sensory symptoms and Bonferroni post-hoc test was applied.

4.3.1 Sensory processing patterns

Registration: lack of response to some of sensory events that other people easily notice and respond to

A significant main effect of group was found for Registration ($F_{(3, 136)}$=35.26, p<.001). The two ASD groups did not differ significantly from each other (p=1), while TD and WS groups differed significantly from all the other groups. The TD group had higher scores on

Registration than the remaining groups (please, see Table 4.1 for the summary of the descriptive characteristics of the participants, all $p<.01$), indicating more typical behaviours. In contrast, the WS group had the lowest scores (more atypicalities) on Registration compared to the other groups (all $p<.01$). The main effect for age was non-significant ($F_{(2, 136)}=.65$, $p=.525$; partial $\eta^2=.009$, observed power$=.157$). The interaction between group and age was also found to be non-significant ($F_{(6, 136)}=1.77$, $p=.111$; partial $\eta^2=.072$, observed power$=.650$).

Seeking: involves actively looking for ways of increasing sensory input in everyday situations

A significant main effect of group was found ($F_{(3, 133)}=29.11$, $p<.001$). Post-hoc analysis indicated that the TD group obtained significantly higher scores than all other groups ($p<.001$), showing more typical sensory behaviours than the other groups. The differences between the other groups were non-significant (all at the $p=1$ level). The main effect of age ($F_{(2, 133)}=.39$, $p=.679$; partial $\eta^2=.006$, observed power$=.112$) and the interaction between group and age ($F_{(6, 136)}=1.89$, $p=.088$; partial $\eta^2=.078$, observed power$=.684$) were non-significant.

Sensitivity: readily responding to sensory events which are not detected or noticeable by other people

A significant main effect of group was found ($F_{(3, 132)}=40.03$, $p<.001$). The TD group obtained significantly higher scores than all other groups ($p<.001$), engaging in more typical sensory behaviours than the other groups. The differences between the other groups were non-significant (all at the $p=1$ level). The main effect of age ($F_{(2, 132)}=.28$, $p=.76$; partial $\eta^2=.004$, observed power$=.093$) and the interaction between group and age ($F_{(6, 132)}=1.40$, $p=.221$; partial $\eta^2=.060$, observed power$=.530$) were non-significant.

Avoiding: actively limiting or avoiding the number of sensory stimuli

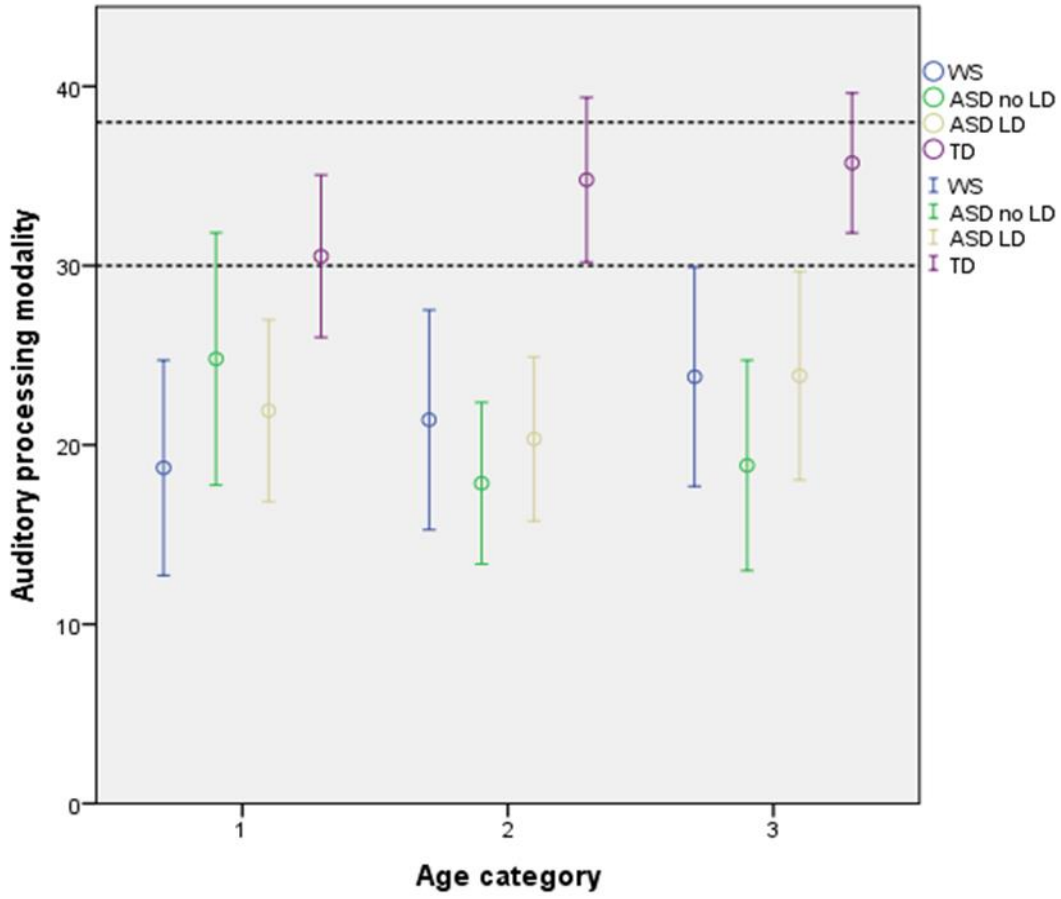
The main effect of group was found for Avoiding ($F_{(3,130)}=36.92$, $p<.001$). The TD group obtained significantly higher scores than all other groups ($p<.001$). The differences between the other groups were non-significant (all at the $p=1$ level). A significant main effect for age ($F_{(2, 130)}=.10$, $p=.90$; partial $\eta^2=.002$, observed power$=.065$) or interaction between group and age ($F_{(6, 130)}=1.31$, $p=.26$; partial $\eta^2=.057$, observed power$=.498$) were not found.

4.3.2 Sensory modalities

Auditory

A significant main effect of group was found for Auditory ($F_{(3, 134)}=51.97$, $p<.001$). The TD group obtained significantly higher scores ($p<.001$) than the neurodevelopmental groups indicating more typical sensory behaviours. The differences between the other groups were

non-significant (all at the p=1 level). The main effect of age was non-significant ($F_{(2, 134)}$=1.56, p=.214; partial η^2=.023, observed power=.326). A significant group by age interaction was found ($F_{(6, 134)}$=4.04, p=.001). The TD and WS group' scores increased for each age category (for TD group within typical range: 30.53 (1.24), 34.79 (1.44), 35.73 (1.62), for WS group within atypical range: 18.73 (1.62), 21.40 (1.70), 23.80 (1.70) respectively), indicating a reduction in atypicality across age categories. Both ASD groups, on the other hand, showed an initial decrease and then an increase in scores with age (for ASD no LD: 24.80 (1.39), 17.86 (1.44), 18.86 (2.04) and for ASD with LD: 21.92 (1.55), 20.33 (1.80), 23.86 (1.44) respectively) (Figure 4.1).

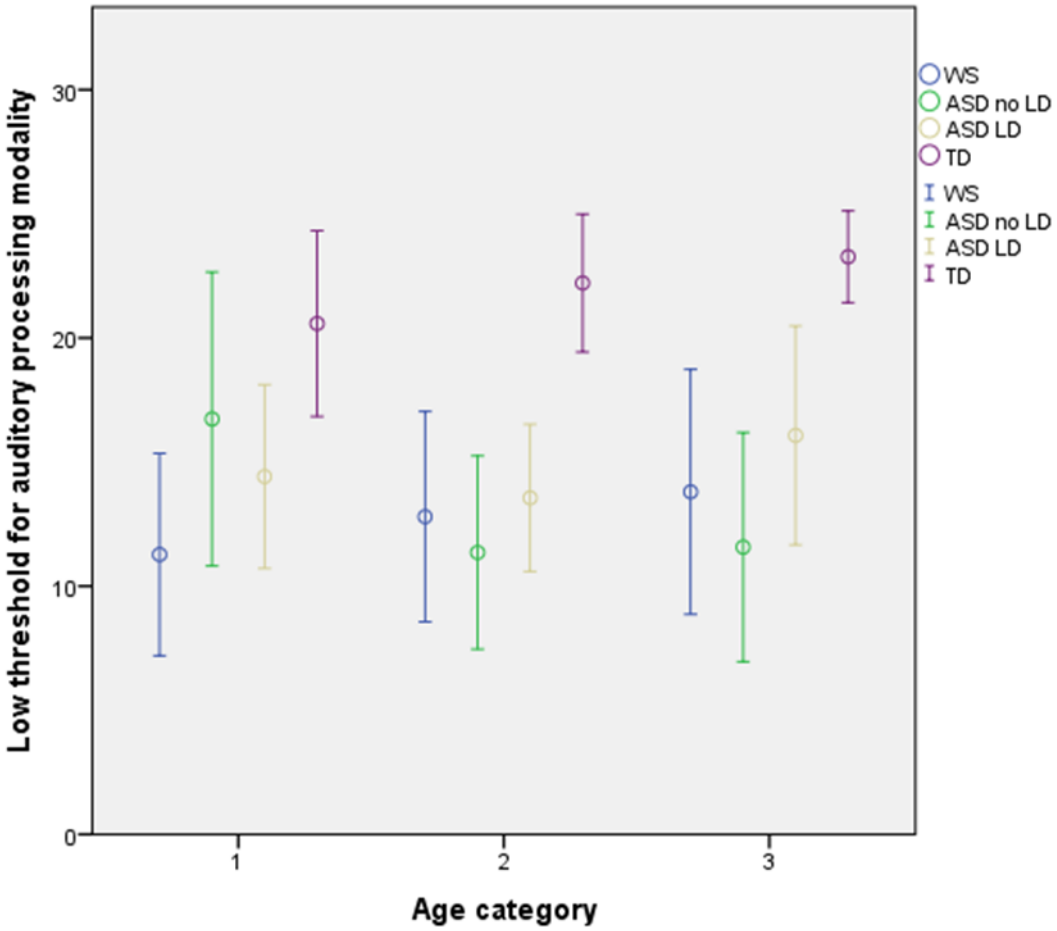


Note: Error bars indicate +/- 1 SD from the mean. Dotted lines indicate typical range of the auditory processing scores.

Figure 4.1 Auditory modality for three time points and four groups.

A significant main effect of group was found for low threshold auditory ($F_{(3, 134)}$=43.69, p<.001). The TD group obtained significantly higher scores (p<.001) than the neurodevelopmental groups indicating more typical sensory behaviours. There were no significant differences between the other groups (ASD with LD and ASD without LD; and

WS and ASD no LD, p=1; WS and ASD with LD, p=.151). The main effect of age was non-significant ($F_{(2, 134)}$=.95, p=.388). A significant group by age interaction was found ($F_{(6, 134)}$=3.24, p=.005). The TD and WS group' scores increased for each age category (for TD group within typical range: 20.58 (.94), 22.21 (1.09), 23.27 (1.23), for WS group within atypical range: 11.27 (1.23), 12.80 (1.29), 13.80 (1.29) respectively), indicating a reduction in atypicality. Both ASD groups, on the other hand, showed an initial decrease and then an increase in scores (for ASD no LD: 16.73 (1.05), 11.36 (1.09), 11.57 (1.54) and for ASD with LD: 14.42 (1.18), 13.56 (1.36), 16.07 (1.09) respectively) (Figure 4.2).

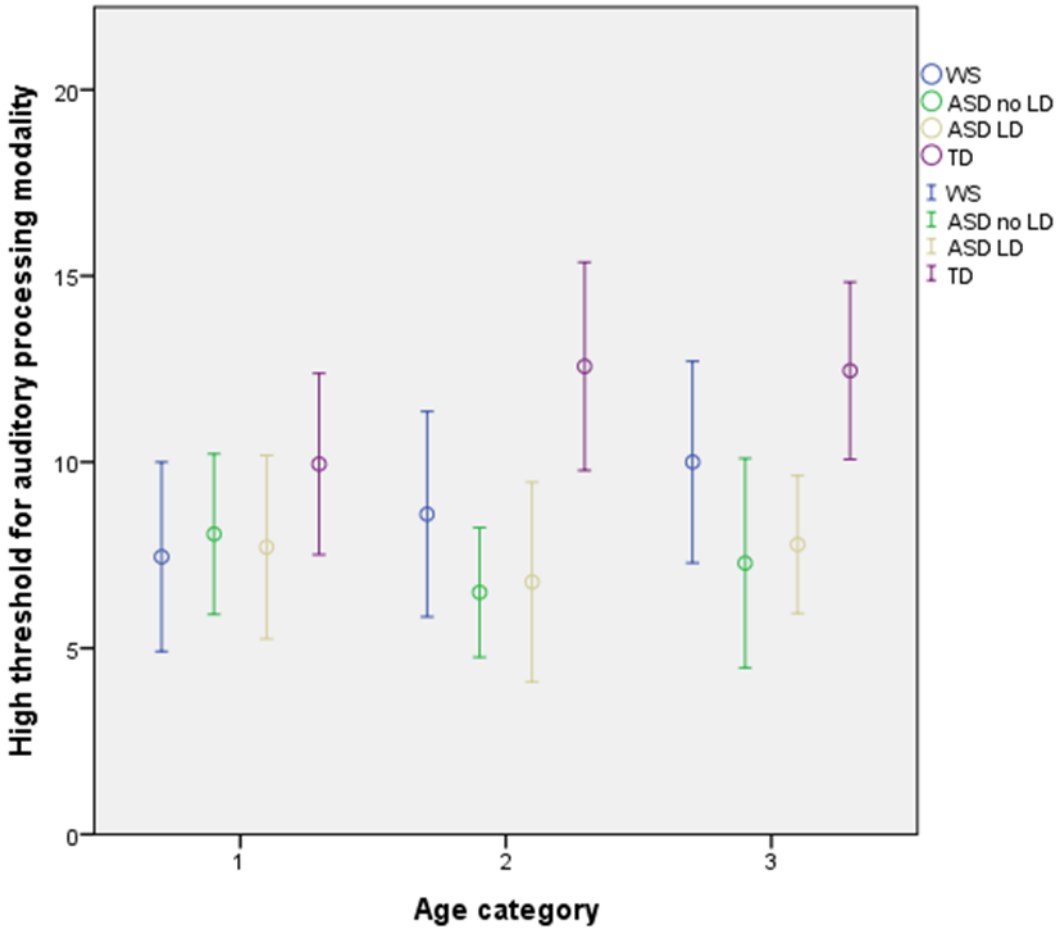


Note: Error bars indicate +/- 1 SD from the mean.

Figure 4.2 Low threshold auditory modality for three time points and four groups

A significant main effect of group was found for high threshold Auditory ($F_{(3, 136)}$=27.33, p<.001). The TD group obtained significantly higher scores (p<.001) than the neurodevelopmental groups indicating more typical sensory behaviours. There were no significant differences between the other groups (ASD no LD and ASD with LD, p=1; ASD

no LD and WS, p=.153; ASD with LD and WS, p=.342). The main effect of age was non-significant ($F_{(2, 136)}$=2.39, p=.096). A significant interaction between age and group was found ($F_{(6, 136)}$=2.77, p=.014). The TD group scores increased between the first and second age category (9.95 (.56) and (12.57 (.65)) and remained constant thereafter (12.46 (.73)), while WS group scores increased for each age category (7.46 (.73), 8.60 (.77), 10.00 (.77) respectively). Both ASD groups, on the other hand, showed an initial decrease and then an increase in scores (for ASD no LD: 8.07 (.63), 6.50 (.65), 7.29 (.92) and for ASD with LD: 7.71 (.65), 6.78 (.81), 7.79 (.65) respectively) (Figure 4.3).



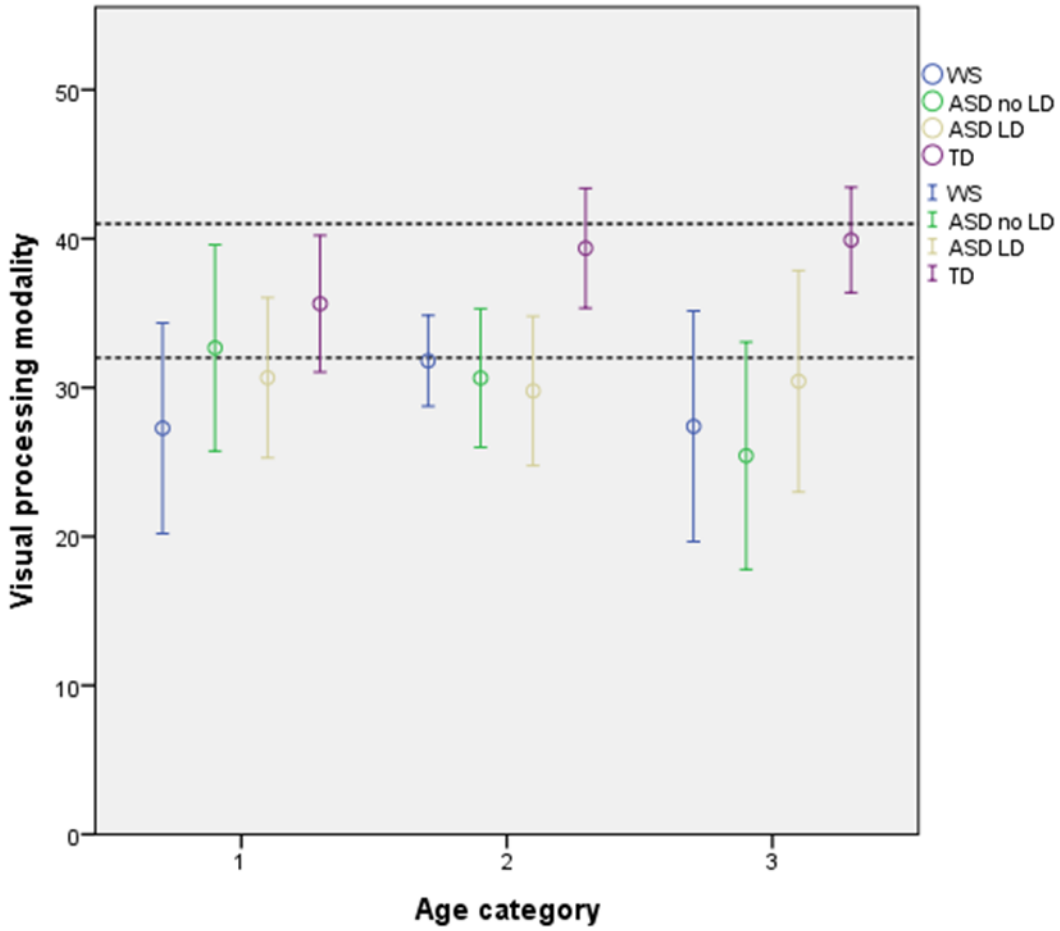
Note: Error bars indicate +/- 1 SD from the mean.

Figure 4.3 High threshold auditory modality for three time points and four groups

Visual

A significant main effect of group was found for Visual ($F_{(3, 134)}$=22.97, p<.001). The TD group obtained significantly higher scores (p<.001) than the other groups indicating that the TD group engaged in more typical sensory behaviours. The differences between the other

groups were non-significant (all at the p=1 level). The main effect of age was non-significant ($F_{(2, 134)}$=1.56, p=.214; partial $\eta 2$=.022, observed power=.313). A significant group by age interaction was found ($F_{(6, 134)}$=2.48, p=.026). The TD group showed an increase in scores over age categories (35.63 (4.59), 39.36 (4.03), 39.91 (3.53) respectively), ASD without LD group showed decrease (32.67 (6.94), 30.64 (4.65), 25.43 (7.64) respectively), ASD with LD remained constant over three age categories (30.67 (5.38), 29.78 (5.02), 30.43 (7.44) respectively), while WS group showed increase in early years, and then a later decrease (27.27 (7.07), 31.80 (3.05), 27.40 (7.75) respectively) (Figure 4.4).

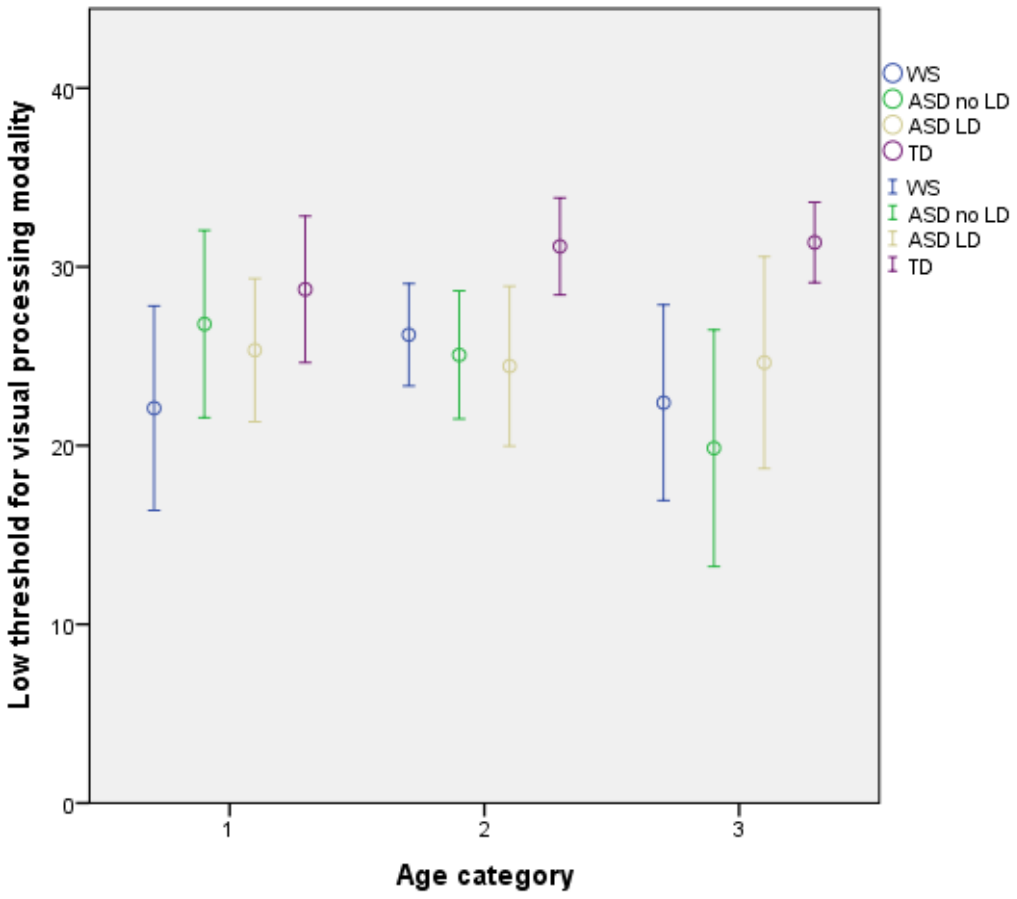


Note: Error bars indicate +/- 1 SD from the mean. Dotted lines indicate typical range of the visual processing scores.

Figure 4.4 Visual modality for three time points and four groups

A significant main effect of group was found for Low threshold Visual ($F_{(3, 134)}$=19.42, p<.001). The TD group obtained significantly higher scores (p<.001) than the neurodevelopmental groups indicating more typical sensory behaviours. The differences

between the other groups were non-significant (all at the p=1 level). The main effect of age was non-significant ($F_{(2, 134)}$=2.39, p=.095). A significant group by age interaction was found ($F_{(6, 134)}$=2.82, p=.013). The TD group showed an increase in scores for each age category (28.74 (4.09), 31.14 (2.71), 31.36 (2.25) respectively). WS group showed an initial increase and then a decrease in scores (22.09 (5.72), 26.20 (2.86), 22.40 (5.48) respectively). ASD with LD group had similar scores across different age categories (25.33 (4.01), 24.44 (4.48), 24.64 (5.93) respectively), while ASD without LD group showed a decrease in scores across the age categories (26.80 (5.24), 25.07 (3.58), 19.86 (6.62) respectively) (Figure 4.5).



Note: Error bars indicate +/- 1 SD from the mean.

Figure 4.5 Low threshold visual modality for three time points and four groups

A significant main effect of group was found for High threshold Visual ($F_{(3, 134)}$=15.76, p<.001). TD group obtained significantly higher scores (p<.001) than the neurodevelopmental groups indicating more typical sensory behaviours. The differences between the other groups were non-significant (all at the p=1 level). The main effect of age

was non-significant ($F_{(2, 134)}$=.67, p=.514). Also a non-significant group by age interaction was found ($F_{(6, 134)}$=.93, p=.473).

Vestibular

A significant main effect of group was found for Vestibular ($F_{(3, 136)}$=15.13, p<.001). TD group obtained significantly higher scores (p<.001) than the other groups. The differences between the other groups were non-significant (all at the p=1 level). The main effect of age ($F_{(2, 136)}$=.05, p=.95; partial $\eta 2$=.001, observed power=.058) and the interaction between group and age ($F_{(6, 136)}$=.98, p=.441; partial $\eta 2$=.041, observed power=.378) were non-significant.

Low threshold Vestibular showed a significant group effect ($F_{(3, 136)}$=7.19, p<.001). TD group obtained significantly higher scores (p<.001) than the neurodevelopmental groups indicating more typical sensory behaviours. The differences between the other groups were non-significant (for both ASD groups, p=1; for WS and ASD with LD, p=.231; for WS and ASD no LD, p= .342). The main effect of age was non-significant ($F_{(2,136)}$=.14, p=.868). Also a non- significant group by age interaction was found ($F_{(6, 136)}$=.24, p=.963).

High threshold Vestibular showed a significant group effect ($F_{(3, 136)}$=15.80, p<.001). TD group obtained significantly higher scores (p<.001) than the neurodevelopmental groups indicating more typical sensory behaviours. The differences between the other groups were non-significant (for both ASD groups, p=1; for WS and ASD with LD, p=.651; for WS and ASD no LD, p= .948). The main effect of age was non-significant ($F_{(2, 136)}$=.03, p=.974). Also a non-significant group by age interaction was found ($F_{(6, 136)}$=1.99, p=.071).

Touch

The main group effect was found for Touch ($F_{(3, 136)}$=32.30, p<.001). TD group obtained significantly higher scores (p<.001) than the other groups. The differences between the other groups were non-significant (for ASD with LD and ASD without LD; and for WS and ASD with LD p=1, for WS and ASD without LD p=.73). The main effect of age ($F_{(2, 136)}$=.62, p=.54; partial $\eta 2$=.009, observed power=.152) was not significant and neither was the interaction between group and age ($F_{(6, 136)}$=1.48, p=.19; partial $\eta 2$=.061, observed power=.559).

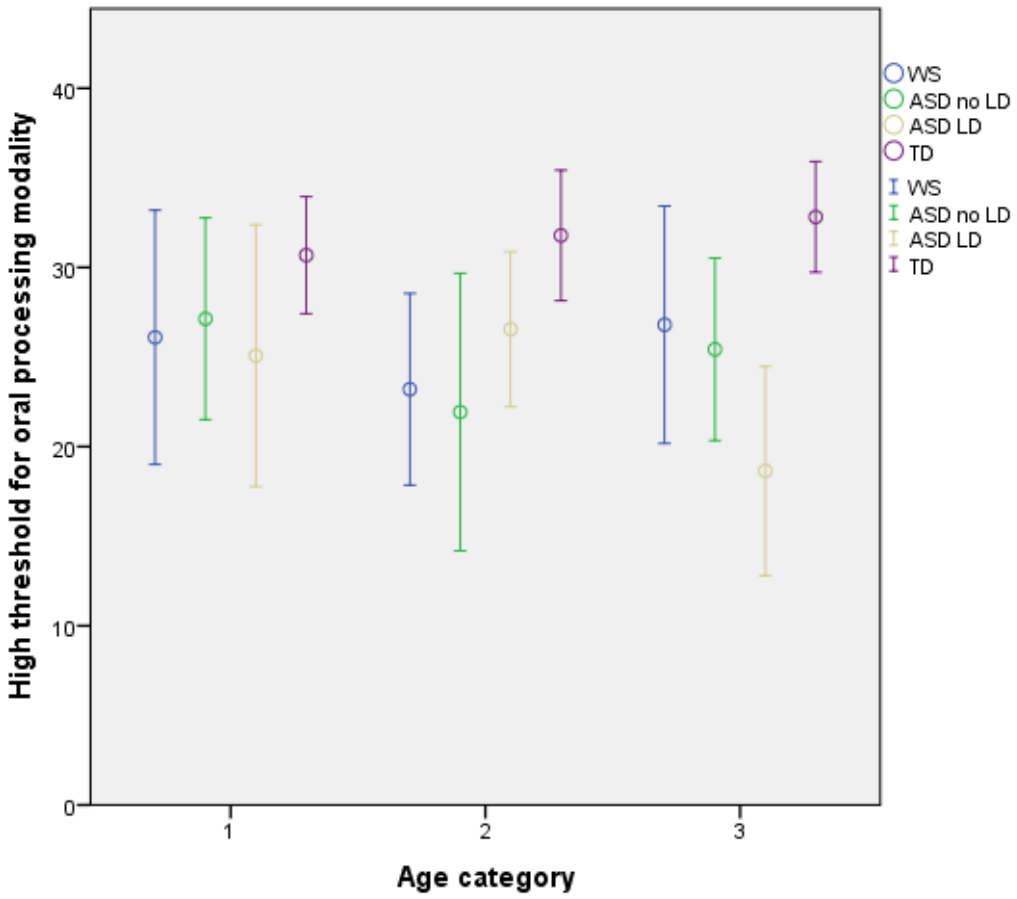
Low threshold Touch showed a significant group effect ($F_{(3, 136)}$=25.78, p<.001). TD group obtained significantly higher scores (p<.001) than the neurodevelopmental groups indicating more typical sensory behaviours. The differences between the other groups were non-

significant (for both ASD groups, p=1; for WS and ASD with LD, p=1; for WS and ASD no LD, p= .259). The main effect of age was non-significant ($F_{(2,136)}$=1.12, p=.329). Also a non-significant group by age interaction was found ($F_{(6, 136)}$=1.36, p=.234).
High threshold Touch showed a significant group effect ($F_{(3, 136)}$=21.60, p<.001). TD group obtained significantly higher scores (p<.001) than the neurodevelopmental groups indicating more typical sensory behaviours. The differences between the other groups were non-significant (all at the p=1 level). The main effect of age was non-significant ($F_{(2,136)}$=.10, p=.901). Also a non-significant group by age interaction was found ($F_{(6, 136)}$=1.05, p=.395).

Oral

The main group effect was found for Oral ($F_{(3, 134)}$=17.08, p<.001). TD group obtained significantly higher scores (p<.001) than the other groups. The differences between the other groups were non-significant (all at the p=1 level). The main effect of age ($F_{(2, 134)}$=.47, p=.623; partial $\eta 2$=.007, observed power=.126) and the interaction between group and age ($F_{(6, 134)}$=2.06, p=.062; partial $\eta 2$=.085, observed power=.730) were non-significant.
Low threshold Oral showed a significant group effect ($F_{(3, 134)}$=10.32, p<.001). TD group obtained significantly higher scores (p<.001) than the neurodevelopmental groups indicating more typical sensory behaviours. The differences between the other groups were non-significant (all at the p=1 level). The main effect of age was non-significant ($F_{(2, 134)}$=.59, p=.555). Also a non- significant group by age interaction was found ($F_{(6, 134)}$=.61, p=.720).

High threshold Oral showed a significant group effect ($F_{(3, 134)}$=17.21, p<.001). TD group obtained significantly higher scores (p<.001) than the neurodevelopmental groups indicating more typical sensory behaviours. The differences between the other groups were non-significant (for both ASD groups, p=1; for WS and ASD with LD, p=.994; for WS and ASD no LD, p= 1).The main effect of age was non-significant ($F_{(2, 134)}$=.97, p=.381). A significant group by age interaction was found ($F_{(6, 134)}$=3.51, p=.003). TD group showed an increase in scores for each age category (30.68 (3.27), 31.79 (3.64), 32.82 (3.09) respectively). WS and ASD without LD groups showed an initial decrease and then an increase in scores (26.10 (7.09), 23.20 (5.35), 26.80 (6.63) and 27.13 (5.63), 21.93 (7.75), 25.43 (5.09) respectively). ASD with LD group showed an increase in scores between the first two age categories and then a decrease in scores (25.08 (7.31), 26.56 (4.33), 18.64 (5.84) respectively) (Figure 4.6).



Note: Error bars indicate +/- 1 SD from the mean.

Figure 4.6 High threshold oral modality for three time points and four groups

4.4 Discussion

In this study, the examination of sensory processing profiles in WS and ASD was undertaken. It was predicted that sensory processing patterns will be atypical in the ASD and WS groups compared to typically developing individuals, with ASD and WS children having significantly lower scores on the SP than TD children. The study also aimed to determine any age related differences within the sensory processing profiles in order to identify possible developmental changes within sensory processing trajectories. It was predicted that sensory patterns in individuals with autism and WS will decrease over age, however compared to typically developing individuals their sensory symptoms will remain within the atypical range over time. Significant group differences were found for all sensory processing patterns and modalities. For all sensory features the TD group had higher (more typical) scores than the WS, ASD with LD and ASD without LD groups. Additionally, for Registration, the WS group differed significantly from all the other groups. A significant main effect for age was not found for either sensory processing patterns or sensory modalities, indicating that sensory

symptoms have similar severity and frequency across the age categories for the sample overall. Only two significant group by age category interactions were found – for auditory and visual modalities, with mixed patterns of changes in these modalities.

The finding that both ASD groups had lower sensory scores, hence showed more atypical sensory responses as reported by parents, compared to the TD group, is not surprising. The first studies to use the Sensory Profile reported the same pattern, with significant differences between sensory symptoms in autism and typical development (Kientz & Dunn, 1997; Watling et al., 2001). Interestingly, when children with autism were compared to other clinical groups, they showed more sensory symptoms than children with general developmental delays, but when compared to a few specific clinical groups, such as fragile X syndrome and deaf-blind children, children with autism showed similar or lower levels of sensory dysfunction (for the review see: Rogers & Ozonoff, 2005). Previous research in WS suggested that 90% of children with WS demonstrated sensory atypicalities (John & Mervis, 2010). In this study, directly comparing ASD and WS groups, we found that children with ASD and WS have very similar sensory profiles, although it is important to highlight that individuals showed high variability within the groups. This suggests that children with WS and ASD, experience significant sensory atypicalities. Also, in this study the two ASD groups, one with LD and the other without LD, showed a very similar level of sensory difficulties across both sensory processing patterns and sensory modalities. This is in contrast to Klintwall et al. (2011), who reported that sensory abnormalities in preschool children with an ASD diagnosis, were more common in those with ASD without LD than in other ASD subgroups. It is possible that their findings are accounted for ability, with the young children in their sample being able to better express their sensory difficulties than young children with ASD and LD.

A number of individual research studies have reported a linear relationship between sensory features and age in ASD (Kern et al., 2007; Saulnier, 2003; Talay-Ongan & Wood, 2000). Yet, evidence from a meta-analysis of sensory modulation symptoms in individuals with ASD (Ben-Sasson et al., 2009) suggested that a trajectory of sensory symptoms across age had a nonlinear course. The authors established that over-responsivity and seeking behaviours increased in frequency up to 6-9 years, and decreased in later years. The inconsistent pattern of changes was found for under-responsivity. In this study, however, the relationship between chronological age and sensory under-responsivity was not found as child's age group was not associated with Registration or Seeking scores for any of the groups. Although using a different measure and age categories, Adamson et al. (2006) found no relationship between age group (50-100 months, 101-150 months and 151+ months) and

the definite difference category on the Short Sensory Profile. Kern et al. (2007) like the current study used the Sensory Profile and investigated sensory processing patterns in autism, reported an age effect in the Low Registration, Sensation Seeking, and Sensory Sensitivity with older individuals showing less sensory difficulties than younger persons with autism. In that study, however, participants across much wider age range (3-43 years old) were recruited and included into one of seven age categories (3–7; 8–12; 13–17; 18–22; 23–27; 28–32; and 33+). It has been suggested, that a neurological normalization process might be taking place (Kern et al., 2007). The same age categories were distinguished in Kern et al. (2006), in which the examination of auditory, visual, oral and touch processing was undertaken. The authors reported, similarly to this study, significant group differences between ASD and TD participants on auditory, visual, touch, and oral sensory processing modalities, as well as on low and high thresholds of these modalities. In this study significant interactions between age and group were found for the auditory high threshold, visual processing and oral high threshold patterns, suggesting that the four groups have different patterns of change in these variables; the TD group showed either an increase or remained constant in all investigated sensory modalities, while the clinical groups showed mixed patterns of change with age. Whereas Kern et al. (2006) examined the sensory processing differences between only ASD and TD groups across seven age categories, we compared ASD with and without LD, WS and TD groups and the age range of participants was narrower.

There is no previous study examining changes in sensory symptoms across different age groups in WS. Based on studies investigating hyperacusis in WS, it was expected that sensory symptoms would slightly decrease in severity with age (Gothelf et al., 2006; Udwin, 1990). In contrast to previous research in general, sensory processing patterns did not significantly differ between the three age groups in WS, showing similar level of sensory atypicalities in children and teenagers with WS. Our methods were different to those used in previous studies and in both the Gothelf et al. (2006) and Udwin (1990) studies the age range of samples was much broader (up to 35 years old), making comparisons of the findings difficult. It is possible, of course, that the decrease in severity of sensory symptoms takes place in adulthood.

4.4.1 Study limitations

This study has some limitations. Although the groups did not differ on age and represented a typical gender radio for each sample, the participants were not matched on age and gender. While the three age groups were included in the study and some age-related conclusion have been made, the study design was not longitudinal. Only following up the same participants at

several time points would allow us to fully examine the developmental trajectory of sensory symptoms. Within-syndrome heterogeneity may play an important role in the presentation of the symptoms as the variability even within the age groups for each condition was high. Although total sample sizes were relatively large for the groups of interest, age sub-groups were much smaller and hence multiple comparisons were undertaken on relatively small samples; yet, the alpha values were adjusted accordingly as Bonferroni post-hoc tests were applied to control Type I error and analyses were powered to detect associations between chronological age and sensory symptoms presentation. Every effort has been made to include participants with WS and those with ASD with LD, and ASD without LD, however, inclusion of other clinical groups, particularly those with general developmental delay and fragile X syndrome could further benefit our understanding of syndrome-specificity of sensory atypicalities.

4.4.2 Conclusions

In this study, directly comparing ASD and WS groups, we found that children with ASD and WS have very similar sensory profiles that are distinct from those present in typically developing children. Similar levels of sensory atypicalities in WS and ASD across both sensory processing patterns and sensory modalities across age groups suggests that sensory difficulties remain a persistent characteristic of both disorders during childhood and adolescence.

Chapter 5. Sensory processing profile and autistic symptoms as predictive factors in autism spectrum disorder and Williams syndrome

5.1 Background

As shown in Chapter 4, children with ASD and WS have very similar sensory profiles that are distinct from those present in typically developing children. In addition to the growing evidence of the pervasiveness of sensory processing atypicalities across neurodevelopmental disorders, there is also mixed evidence in relation to the specificity of socio-communicative abnormalities to ASD, as assessed by the Social Responsiveness Scale (SRS; Constantino & Gruber, 2005). In previous research that involved the SRS with over 1,900 children, scores distinguished between children with pervasive developmental disorders and those with other child psychiatric disorders (Constantino et al., 2000; Constantino & Todd, 2000, 2003). However, there is also evidence that socio-communicative abnormalities are common in children with WS (Klein-Tasman et al., 2007) and these atypicalities are very similar to the difficulties observed in children with ASD (Klein-Tasman et al., 2011). Indeed a comorbid autism diagnosis has been reported in several WS cases (Gillberg & Rasmussen, 1994; Herguner et al., 2006). Klein-Tasman et al. (2011) who investigated social functioning of children with WS between 4 and 16 years old using the SRS, both parent and teacher versions, reported that a number of children with WS displayed marked social difficulties across the subscales, most commonly in the areas of Social Cognition, Social Communication, and Autistic Mannerisms, with a relative strength in Social Motivation. Also Riby et al. (2014) showed that 58% of individuals with WS (reported by parent-completed SRS) had severe deficits of reciprocal social interaction. In summary, there is emerging evidence that some ASD symptoms are also common in WS. Yet, a cross-syndrome comparisons of the social-communication features and sensory profiles of the two disorders, has been never undertaken.

The aims of this study were: (a) to examine and compare the sensory and social responsiveness profiles in three groups of children and adolescents with a neurodevelopmental disorder, those with a diagnosis of ASD without learning disability, (LD), those with ASD with additional learning disability, and those with WS; (b) to investigate whether autistic symptoms and sensory processing scores can predict group membership.

5.2 Methods

5.2.1 Participants

Parents of children with ASD or WS, between 4 and 16 years of age were recruited to take part in the study ('Sensory Hotpots in Children' study; see Chapter 4). The initial dataset consisted of 35 parents of children with WS and 74 parents of children with ASD. Nine WS children did not have complete datasets. The LD status of two ASD children was unclear and further 16 children did not have a complete data. The final samples consisted of 26 children with WS and 56 children with ASD, of whom 26 had additional LD.

The WS children were aged between 53 to 181 months (mean 96.77, sd=35.71; 13 male). All of the children had previously been clinical diagnosed and had their diagnosis confirmed genetically with fluorescent in situ hybridization testing (FISH) to detect the deletion of one copy of the ELN gene. All of the children with ASD had previously been diagnosed based on a multidisciplinary team assessment following the guidelines of the UK National Autism Plan for Children (Le Couteur, 2003), as stated by the parents. Within the ASD sample, two subgroups were distinguished – those with ASD without additional diagnosis of LD, and children with ASD with comorbid learning disability (LD). LD status was assigned either in agreement with a parent report (for 43 children data were available and 22 ASD children were classified as having additional LD) or examined in a direct assessment. Thirteen children were assessed directly and four who obtained standard scores below 70 on both Raven's Coloured Progressive Matrices (RCPM; Raven et al., 1998) and British Picture Vocabulary Scale - Third Edition (BPVS3; Dunn et al., 2009) were identified as having additional LD. Thirty children with ASD without LD were aged between 50 and 161 months old (mean 90.10, sd=27.11; 21 male) and 26 children with ASD with LD were aged were between 54 and 184 months (mean 108.85, sd=39.36; 24 male). The groups significantly differed on age $t_{(43.453)}$=-2.044, p=.047.

5.2.2 Measures

Sensory Profile (SP; Dunn, 1999). The SP is a caregiver questionnaire that measures a child's sensory processing abilities. The questionnaire consists of 125 items, rated on a five-point Likert scale, ranging from almost never to almost always (see Chapter 4 for more details).

Social Responsiveness Scale – Second Edition (SRS-2; Constantino & Gruber, 2012). The SRS-2 is a 65-item parent-report rating scale of autistic traits in 4- to 18-year-olds. The questionnaire is commonly used as either a screener or as an aid to clinical diagnosis of ASD.

The items cover a range of unusual interpersonal behaviours, communication or repetitive/stereotyped behaviours and are rated on a four-point Likert scale, ranging from not true to almost always true. Parents during the questionnaire completion are asked to focus on the child's behaviour in the past 6 months. The SRS-2 provides scores in five subscales including Social Awareness, Social Cognition, Social Communication, Social Motivation, and Restricted Interests and Repetitive Behaviour (RRB); and the Total score. Higher scores indicate greater impairment, with a total T-score of 76 or higher indicating severe, between 75 and 66 moderate, between 65 and 60 mild deficits in social interactions, and a T-scores below 59 representing typical range.

5.2.3 Procedure

Questionnaire packs including an information sheet, the consent form, the SP and SRS-2 were sent to parents of children with ASD and with WS. In the case of missing data in the questionnaires, parents were contacted again and asked to provide missing information. Those ASD children who underwent a direct ability assessment were evaluated either at their homes or schools. Favourable ethical opinion was granted by Newcastle University Faculty of Medical Sciences Ethics Committee.

5.2.4 Data analysis

In this study, 1.4% of the SP and 1.2% of the SRS-2 item scores were missing. There was no pattern within missing data. Missing values were treated as missing completely at random and replaced by the mean of the non-missing subscale items when less than 20% of the data within the subscale was missing. To determine whether there were significant differences between the means of the three groups and to investigate whether autistic symptoms and sensory quadrant scores could predict group membership one-way ANOVA and regression analyses were subsequently undertaken on the complete dataset. Adaptions for multiple comparisons were dealt with by applying Bonferroni corrections.

5.3 Results

One-way ANOVA and Chi-square test were performed to determine whether there were any significant differences in mean scores between the three groups on age and gender. The groups did not differ on the mean age ($F_{(2,79)}=2.14$, $p=.125$), however they did differ on gender ($\chi^2_{(2)}=10.14$, $p=.005$) with ASD with LD group having significantly more males than WS group (34 to 13, $p=.002$). Descriptive statistics for the participants on the outcome variables are presented in Table 5.1.

The analysis showed that the groups were significantly different on most of the SRS-2 subscales: SRS-2 Total score ($F_{(2,79)}$=7.86, p=.001), Social Awareness ($F_{(2, 79)}$=8.42, p<.001), Social Communication ($F_{(2,79)}$=13.20, p<.001, Social Motivation ($F_{(2,79)}$=11.71, p<.001). The differences in mean scores for the SRS-2 on Social Cognition ($F_{(2,79)}$=1.35, p=.265) and RRB ($F_{(2,79)}$=2.06, p=.134) were not significant. For the SP the groups were significantly different on the Low Registration ($F_{(2,79)}$=3.73, p=.028) only, while the differences in mean scores on Sensation Seeking ($F_{(2,79)}$=.69, p=.505), Sensory Sensitivity ($F_{(2,79)}$=.65, p=.524) and Sensation Avoiding ($F_{(2,79)}$=.27, p=.763) were not significant.

For the SRS-2 total score the WS group differed from both ASD groups, with significantly lower scores in both cases (as indicated by Bonferroni test for ASD LD: p=.001, for ASD no LD: p=.01). The same pattern was found for the SRS-2 subscales, with the WS sample having significantly lower scores than both ASD groups (Social Awareness: WS and ASD no LD, p=.018, WS and ASD LD, p<.001; Social Communication: WS and ASD no LD, p<.001, WS and ASD LD, p<.001; Social Motivation: WS and ASD no LD, p=.001, WS and ASD LD, p<.001). As post-hoc analysis indicated, for Low Registration only WS and ASD without LD groups different significantly (p=.042) with WS group having lower scores than ASD without LD group.

Table 5.1 Mean (SD) scores on outcome variables

Variable	WS (n=26)	ASD with LD (n=26)	ASD without LD (n=30)
SRS-2:			
Total score	87.85 (28.72)	118.81 (29.76)	111.80 (30.38)
Social Awareness	11.54 (3.74)	15.85 (3.81)	14.43 (4.01)
Social Cognition	20.00 (5.79)	22.54 (5.99)	20.60 (5.76)
Social Communication	26.42 (9.69)	40.00 (10.92)	37.63 (10.09)
Social Motivation	10.04 (5.90)	18.58 (6.83)	17.17 (7.63)
RRB	19.85 (7.50)	23.92 (7.39)	21.97 (6.89)
SP Quadrant:			
Low Registration	43.85 (13.51)	52.23 (13.89)	52.53 (12.29)
Sensation Seeking	87.62 (17.07)	82.08 (16.07)	82.97 (21.14)
Sensory Sensitivity	61.65 (14.56)	65.58 (11.14)	65.23 (15.27)
Sensation Avoiding	90.65 (17.89)	92.12 (16.21)	88.77 (17.05)

Multinomial logistic regression was undertaken with sensory quadrants (Low Registration, Sensory Sensitivity, Sensation Seeking, and Sensory Avoiding) and SRS-2 subscales (Awareness, Cognition, Communication, Motivation and RRB) as covariates. The model was significant $\chi^2_{(18)}=91.62$, $p<.001$. Low Registration ($\chi^2_{(2)}=23.57$, $p<.001$), Sensation Avoiding ($\chi^2_{(2)}=8.82$, $p=.012$), Social Communication ($\chi^2_{(2)}=8.75$, $p=.013$), Social Awareness ($\chi^2_{(2)}=8.00$, $p=.021$) and Social Cognition ($\chi^2_{(2)}=8.26$, $p=.016$) had a significant main effect on diagnostic group. Non-significant main effects were found for Sensory Seeking ($\chi^2_{(2)}=.40$, $p=.82$), Sensory Sensitivity ($\chi^2_{(2)}=4.18$, $p=.123$), RRB ($\chi^2_{(2)}=1.12$, $p=.572$) and Social Motivation ($\chi^2_{(2)}=1.87$, $p=.392$). Additionally, gender was added to the model, however the main effect of gender was non-significant ($\chi^2_{(2)}=3.86$, $p=.144$).

As indicated by parameter estimates with the WS group as a comparison, Low Registration ($\beta=.43$, Wald $\chi^2_{(1)}=5.33$, $p=.021$), Social Cognition ($\beta=-.54$, Wald $\chi^2_{(1)}=4.71$, $p=.03$) and Social Communication ($\beta=-.56$, Wald $\chi^2_{(1)}=4.71$, $p=.041$) significantly predicted whether a participant had WS or ASD without LD. The odds ratio indicated that as Low Registration and Social Communication increased in unit and Social Cognition decreased in unit, a participant was more likely to be diagnosed with ASD without LD rather than with WS. Only Low Registration significantly predicted whether an individual had WS or ASD with LD, ($\beta=.42$, Wald $\chi^2_{(1)}=5.10$, $p=.024$), with the odds ratio showing that as Low Registration increased, it was more likely for a child to be diagnosed with ASD with LD rather than WS. The summary of the results is presented in Table 5.2.

Additionally, when the ASD with LD group was placed in the model as the comparison group, the only variable that significantly predicted whether a participant had a diagnosis of ASD with LD or ASD without LD was Sensation Avoiding ($\beta=.04$, Wald $\chi^2_{(1)}=4.25$, $p=.039$) with the odds ratio showing that as Sensation Avoiding increased, it was more likely for a child to be diagnosed with ASD with LD rather than ASD without LD.

Table 5.2 Multinomial logistic regression

		95% CI for Odds Ratio		
	β (SE)	Lower	Odds Ratio	Upper
WS vs. ASD without LD				
Intercept	-29.17 (20.48)			
Low Registration	.43 (19)*	1.07	1.53	2.21
Sensation Seeking	-.03 (.06)	.86	.97	1.09
Sensory Sensitivity	.18 (.12)	.96	1.20	1.51
Sensation Avoiding	-.18 (.10)	.69	.84	1.02
Social Awareness	.90 (.58)	.80	2.46	7.62
Social Cognition	-.54 (.25)*	.36	.58	.95
Social Communication	.56 (.27)*	1.02	1.74	2.97
Social Motivation	.17 (.17)	.86	1.19	1.65
RRB	-.21 (.22)	.53	.81	1.25
WS vs. ASD with LD				
Intercept	-41.47 (20.82)*			
Low Registration	.42 (19)*	1.06	1.52	2.20
Sensation Seeking	-.02 (.06)	.87	.98	1.11
Sensory Sensitivity	.17 (.12)	.94	1.19	1.49
Sensation Avoiding	-.09 (.10)	.75	.91	1.11
Social Awareness	1.06 (.58)	.92	2.88	8.97
Social Cognition	-.42 (.26)	.40	.66	1.09
Social Communication	.52 (.27)	.99	1.68	2.86
Social Motivation	.21 (.17)	.88	1.24	1.73
RRB	-.17 (.22)	.55	.85	1.30

Note: R^2=.67 (Cox & Snell), .76 (Nagelkerke), *p<.05

5.4 Discussion

The aim of this study was to examine and compare the sensory and social responsiveness profiles in three groups of children and adolescents with a neurodevelopmental disorder, those with a diagnosis of ASD without learning disability, (LD), those with ASD with additional learning disability and those with WS to examine both within-syndrome variability and cross-syndrome comparisons. The study also aimed to investigate whether autistic symptoms and sensory processing scores can predict group membership.

Parent reports of Social Awareness, Social Communication, Social Motivation and Low Registration were found to differ significantly for children with WS, ASD with LD and ASD without LD. Only Low Registration from the SP, and Social Cognition and Social Communication subscales from the SRS-2 were significant predictors of whether a child had WS or ASD without LD. With a change in Low Registration and Social Cognition scores being more typical, and Social Communication scores being more atypical, diagnosis of ASD without LD was more likely than diagnosis of WS. Yet, only Low Registration scores significantly predicted whether an individual had WS or ASD with LD where with Registration scores increasing, a child was more likely to be diagnosed with ASD with LD rather than with WS. These findings indicate that some autistic symptoms and sensory processing scores can predict a group membership between WS and ASD without LD; however, only Low Registration from the SP (and none of the SRS-2 scores) was able to predict whether a child had a diagnosis of WS or ASD with LD. This is particularly important as individuals with WS do have LD and when compared to those with ASD and LD, the similarity between the disorders is striking examining sensory and social responsiveness profiles of children.

That the WS group obtained significantly lower (however, still within atypical range) scores on the SRS-2 than both ASD groups is not surprising. Some authors report that SRS scores can distinguish children with pervasive developmental disorders from those with other disorders (Constantino et al., 2000; Constantino & Todd, 2000, 2003), whereas others report commonality of socio-communicative abnormalities in children with WS (Klein-Tasman et al., 2007) and the similarity of difficulties present in children with ASD (Klein-Tasman et al., 2011). As stated by Klein-Tasman et al. (2011) the most commonly seen difficulties in children with WS are in the areas of Social Cognition, Social Communication, and Autistic Mannerisms (the RRB subscale in the SRS-2). Interestingly, in this study, results indicate that there was a relationship between difficulties in social communication and social cognition and a diagnosis of either ASD without LD or WS; however the relationships differ in their

direction. We found an increase in social communication difficulties and decrease in social cognition scores was associated with ASD without LD or WS. The Social Communication scale of the SRS-2 assesses reciprocity of social communication (e.g. ability to keep the flow of a conversation) (Bruni et al., 2014) and this in turn relates to social communication and interaction, one of two core diagnostic symptoms of autism (Frazier et al., 2008, 2012; Snow et al., 2009). In addition, earlier studies suggested a single global severity factor in autism was strongly associated with social communication behaviour (Constantino et al., 2004). It seems that these social communication difficulties can predict group membership in relation to ASD without LD and WS. Social Cognition, on the other hand, assesses processing of social information (e.g. understanding meaning of social behaviours) (Bruni et al., 2014). As found by Klein-Tasman and colleagues (2011), Social Cognition was rated by the parents of children with WS significantly higher than Social Communication. Similarly, we found the relationship between Social Cognition and likelihood of diagnosis of WS rather than ASD without LD. Surprisingly, neither Social Communication nor Social Cognition, nor any other SRS-2 subscale, significantly predicted either WS or ASD with LD diagnosis. This is particularly theoretically and clinically important as the LD status did not play a role when distinguishing between the diagnoses. Further research is needed to replicate these results and establish whether sensory processing and social responsiveness profiles can discriminate between the groups.

The findings of this study also suggested that sensory symptoms in children and adolescents with WS or ASD are very similar. Only Low Registration predicted whether a child had WS or ASD and no other sensory variable was related to the diagnostic status. Low Registration is associated with high threshold to sensory experiences, passive responses to sensory events and limited detection of changes in sensory situations (Dunn, 1997). There is evidence that features associated with the hyporesponsiveness pattern (Low Registration and Sensory Seeking belong to that pattern) in both social and non-social context can discriminate between children diagnosed with autism, developmental delays and those typically developing (Baranek et al., 2006). Baranek et al. (2006) used the Sensory Experiences Questionnaire, which is a parent report and assessed children between 5 months and 6 years old. In this study the Sensory Profile was used and Low Registration scores rather than hyporesponsiveness pattern scores were calculated. Similarly, we found that Low Registration scores significantly predicted a diagnostic status distinguishing between WS and ASD both with and without LD. Surprisingly, however, lower Low Registration scores (indicating more atypical sensory behaviours) was associated with higher likelihood of a diagnosis of WS, than ASD with or without LD. Further research investigating sensory

profiles in WS and examining cross-syndrome comparisons is needed to establish unique discriminative sensory difficulties for each of these conditions in order to contribute to both theory and intervention.

5.4.1 Study limitations

There are several notable limitations of the current study. First, although in the current study three groups of children with neurodevelopmental disorders, namely WS, ASD without LD and ASD with LD, were included, the sample sizes were still relatively small, especially in light of the analysis strategy. Secondly, only parent-reports were used in this investigation. It is important to note that the SRS-2 was designed solely for use with ASD individuals and was not aimed to be discriminable. The questions focus on the presence rather than a nature of an atypicality and therefore more fine social behaviours present in the WS might not be reported (such as the atypical increased social motivation in WS; Lough et al., 2016). Moreover, data from multiple raters and measures, including direct assessments of social and sensory behaviours would provide better understanding of children's strengths and difficulties. Finally, only a limited number of predictors were entered into the model. However, there are possibly other features that can change the likelihood of WS or ASD diagnosis, for example comorbid behaviour problems that were found to influence autism symptoms (Hus et al., 2013; Mayes & Calhoun, 2010). Incorporating more symptoms could enhance the power of the model.

5.4.2 Conclusions

In conclusion, the current study is one of the first investigations of the relationship between autistic and sensory symptoms, and diagnostic status in three groups of children with neurodevelopmental disorders. The study found that parent reported social responsiveness and sensory profiles in children with WS and ASD were very similar. The distinction between ASD without LD and WS appeared dependent on Social Communication, Social Cognition and Low Registration scores, while the distinction between ASD with LD and WS was related only to Low Registration scores, with those with WS exhibiting more atypicalities. Further work is needed to establish which aspects of socio-communicative and sensory behaviours are unique to each of the disorders.

Chapter 6. Comparisons of the factor structure and measurement invariance of the Spence Children's Anxiety Scale - Parent version in children with autism spectrum disorder and typically developing anxious children

As reported in Chapter 2, research suggests that there is a relationship between sensory processing difficulties and clinical features of ASD. The undertaken systematic review highlighted that affective disorders are associated with hyperresponsiveness in that disorder. Before further investigating the relationship between sensory atypicalities and anxiety and other co-morbid symptoms in both ASD and WS, we would like to consider some of the potential measurement issues.

6.1 Background

Anxiety is a common health concern in children with autism spectrum disorder (ASD), affecting between 11-84% (White et al., 2009) compared to 3-24% of typically developing children (Green & Ben-Sasson, 2010). A meta-analysis (Van Steensel et al., 2011) reported that nearly 40% of individuals with ASD display clinical levels of anxiety and anxiety is one of the most common comorbid psychiatric disorders in children with ASD (Simonoff et al., 2008). Furthermore, anxiety problems can lead to increased maladaptive behaviour (Kim et al., 2000), unemployment, and chronic mental health difficulties among young people with ASD (Farrugia & Hudson, 2006). Although the recognition of anxiety problems in ASD has a long history, starting as early as with the first description of autism by Kanner (1943), the assessment and treatment of anxiety in individuals with ASD has only recently begun to receive the empirical attention it needs and deserves (Rodgers et al., 2012; White et al., 2009). There remains a critical need for the development of valid and reliable assessment measures to accurately identify anxiety in children and young people with ASD.

MacNeil, Lopes and Minnes (2009) reported that young people with ASD have higher levels of anxiety than typically developing (TD) children and comparable levels of anxiety to typically developing clinically anxious children. As is the case among typically developing populations, some forms of anxiety appear to be more common than others in children with ASD (Van Steensel et al., 2011); for example specific phobias are more common than separation anxiety and panic disorder. Sukhodolsky et al. (2008) report the prevalence rates for specific phobias, separation anxiety and panic disorder in children with ASD aged between 5 and 17, as 31%, 10.5% and 0.0%, respectively. Rates reported for Obsessive

Compulsive Disorder (OCD), Social Anxiety Disorder (SAD) and Generalised Anxiety Disorder (GAD) vary widely across studies in ASD (2.6-36.7% for OCD; 0.5- 27.3% for SAD; and 1.2-45.2% for GAD; Van Steensel et al., 2011). Understanding this variability is important and it may be that it is influenced by a number of factors, including the specific challenges of accurately measuring anxiety in ASD.

The presentation of anxiety in children with and without ASD shares some common features, such as social fears that are characteristic of social phobia (Settipani et al., 2012). However, there may also be some unique aspects of anxiety in ASD, for example there is evidence for an association between anxiety and both sensory over-responsivity (Ben-Sasson et al., 2008; Green & Ben-Sasson, 2010) and impairment in social functioning in ASD (Bellini, 2004; Bellini, 2006). Thus, young people with ASD may be predisposed to anxiety as a result of a range of ASD-specific factors. Furthermore, there is also evidence that anxiety can exacerbate some of the features of ASD, such as repetitive behaviours (Sofronoff et al., 2005). Kanner (1943) observed that "an insistence on sameness, and the repertoire of fixed behaviours and routines" appeared to have a strong association with anxiety (Kanner, 1943 as cited in Gillot et al., 2001, p.277). Features of ASD and symptoms of anxiety may however overlap and prove difficult to delineate (Gjevik et al., 2010). For example, repetitive behaviours seen in ASD can be difficult to differentiate from the compulsive behaviours found in OCD (Zandt et al., 2009). Also atypical anxiety symptoms have been reported to be associated with ASD symptomatology, strengthening the overlap and relationship of anxiety and repetitive and restricted behaviours in ASD (Kerns et al., 2014). Furthermore, Mikita et al. (2016) suggested putative links between predisposing ASD traits and subsequent anxiety responses, possibly underpinned by a distinct pathophysiological mechanism. The authors indicated a possibility of distinguishing a distinct nosological category of individuals with ASD and comorbid anxiety that should be researched in its own right. That highlights the need for measures that include anxiety-related items that are specific to the phenomenology of anxiety in ASD (Rodgers et al., 2016). Rodgers and colleagues (2016) have recently developed the first autism-specific anxiety scale (ASC-ASD) with evidence of good reliability and validity.

Generally, the assessment of anxiety in ASD has relied on measures originally validated for use in typically developing populations (White et al., 2009). Given the distinct challenges of measuring anxiety in ASD, the precision of these instruments has been called into question. Van Steensel, Deutschman and Bögels (2013) evaluated the parent-report Screen for Child Anxiety-Related Emotional Disorders (SCARED-71; Bodden et al., 2009) for use in ASD. They reported that although psychometric properties of the measure were

comparable for ASD and anxiety-disordered groups, alternative cut-off scores were recommended for young people with ASD. White, Schry and Maddox (2012) provided mixed evidence for the reliability and validity of both the Multidimensional Anxiety Scale for Children (MASC) and the Child and Adolescent Symptom Inventory-4 ASD Anxiety Scale (CASI-Anx) when used with adolescents diagnosed with high functioning autism. The authors found that the measures had acceptable internal consistency, and there was evidence of discriminant validity, however, the youth self-report was found to have a questionable validity. Kaat and Lecavalier (2015) evaluated the self- and parent-reported Revised Child Anxiety and Depression Scale (RCADS) and a more recent version of the MASC among youth with ASD and raised some concerns regarding the construct validity of anxiety in ASD as measured by these scales. More concerns were particularly raised about the interpretation and validity of child/youth self-report anxiety screening measures in the ASD group (Mazefsky et al., 2011; White et al., 2012). Moreover, acceptable internal consistency, modest convergent validity, and questionable divergent validity in separating anxiety from attention problems in ASD on the RCADS suggested that more convincing evidence is needed to use the tool in ASD (Sterling et al., 2015).

The Spence Children's Anxiety Scale-Parent (SCAS-P; Spence, 1998) is frequently used in ASD research (Chalfant et al., 2006; McConachie et al., 2014; Rodgers et al., 2012; Russell & Sofronoff, 2005; Sung et al., 2011). The SCAS-P is a parent-completed questionnaire for assessing the severity of a range of anxiety symptoms. It has been reported to be a reliable and valid tool for screening anxiety symptoms in typically developing children (Nauta et al., 2004). The parent-report measure also has high correspondence with the well-validated self-report Spence Children's Anxiety Scale (SCAS; Nauta et al., 2004). Russell and Sofronoff (2005) found both parent and child versions of the questionnaire had high internal reliability in ASD samples. Findings from the recent psychometric work done on the questionnaire showed that there was overall moderately good agreement between caregivers' and ASD children's reporting of anxiety symptoms using the SCAS-P and the SCAS (Magiati et al., 2014); and suggested that the SCAS-P could be a useful screening tool for anxiety disorders in ASD (Zainal et al., 2014). A recent systematic review of outcome measures used in anxiety intervention studies for high-functioning children with ASD suggested that the SCAS-P, its revised version, the RCADS, and the SCARED had the most robust measurement properties (Wigham & McConachie, 2014). However, there was little or no evidence for some aspects (e.g. responsiveness to change and content validity). Little is yet known about the reliability or validity of the SCAS-P as a measure of anxiety in children with ASD

It remains unclear whether the SCAS-P measures the same constructs in ASD as it does in typically developing clinically anxious children (without ASD). Moreover, the subsequent question of whether this instrument measures the construct in the same way, should also be addressed to enable valid comparisons of observed scores across groups to be made. Further investigation is required to enable confidence that the scale functions in the same way across clinical groups.

In order to establish whether a given measure of a particular latent construct (such as anxiety) performs similarly across the groups, it has been suggested that measurement invariance should be first performed (Vandenberg & Lance, 2000). Only then can meaningful comparisons between groups be made as measurement invariance analysis indicates whether the instrument measures the same construct in the same way across different populations or groups (Millsap & Kwok, 2004). For example, Garnaat and Norton (2010) assessed measurement invariance of the Yale-Brown Obsessive Compulsive Scale across four racial/ethnic groups (namely, White, Black, Asian, and Hispanic). They found generally stable properties although highlighted some concern that some scales may underestimate diagnosis of OCD in Black groups.

To our knowledge, there has been no attempt to use measurement invariance to compare separate clinical groups. The aims of this study were two-fold. Firstly, to determine the factor structure for the SCAS-P in a sample of young people with ASD and to compare it with the factor structure derived from a sample of clinically-anxious young people without ASD, and in the combined sample to ensure adequate fit to consider invariance. Secondly, to use measurement invariance techniques to determine whether SCAS-P items function in the same way in children with ASD and anxious children without ASD, in order to establish whether cross-groups comparisons using the SCAS-P are appropriate and meaningful. Due to concerns raised about both validity and interpretation child/youth self-report anxiety measures in the ASD group, the parent version of the SCAS was the main focus of this study.

6.2 Methods

6.2.1 Measure

The Spence Children's Anxiety Scale-Parent Version (SCAS-P; Spence 1998) is a 38-item, checklist where parents rate the frequency of occurrence of anxiety symptoms on a four-point Likert-type scale, ranging from 0 (never) to 3 (always). Thus, higher scores indicate increased levels of anxiety. SCAS-P mean norms for the total score in healthy children and young people range between 11.8 and 16, increasing to 30.1 to 33 in anxiety

disordered children and adolescents (Nauta et al., 2004). The scale provides a total anxiety score as well as six subscale scores developed to reflect symptoms characterized by the Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV-R, APA, 2000): panic and agoraphobia; separation anxiety; social phobia; physical injury fears; obsessive–compulsive disorder (OCD), and generalized anxiety disorder (GAD) (see Appendix P). The proposed 6-factor structure has been supported by confirmatory factor analyses (Nauta et al., 2004). The SCAS-P is reported to have satisfactory to excellent reliability and shows acceptable validity for anxious children (Nauta et al., 2004).

6.2.2 Participants

The study involved analysis of archival data pooled from several different settings.

ASD sample. This group consisted of parents of 285 children and adolescents with ASD, recruited from four sources. Most children and adolescents (211participants, 181 male, mean age in months=147.95, SD=24.1; range: 8-16 years old) were seen by health and education teams in the North East of England, recruited through Daslne (Database of children with autism spectrum disorder living in North East; McConachie et al., 2009). The second group consisted of those who took part in the Beating Anxiety Together (BAT) project (McConachie et al., 2014), an intervention programme created for children and adolescents with ASD who also had comorbid high anxiety (21 participants, 20 male, mean age in months=137.05, SD=16.22; range: 8.92-13.58 years old). The third group (19 participants, 16 male, mean age in months=139.74, SD=29.66; range: 8.83-15.58 years old) took part in the UK part of the 'Fun and Games' study investigating decision making styles used by individuals with ASD (Boulter et al., 2014; South et al., 2014). Finally, 34 participants (29 male, mean age in months=139.50, SD=35.90; range: 7.05-17.09 years old) were recruited for a study based at Newcastle University, UK; investigating the relation between executive functioning, sensory processing and anxiety (Darus, unpublished PhD). All children were diagnosed through a multidisciplinary team assessment following the guidelines of the UK National Autism Plan for Children (Le Couteur, 2003). All met criteria for ASD on the Autism Diagnostic Observation Schedule (ADOS; Lord et al. 2000), administered and rated from video by trained raters who maintained over 80% agreement with consensus ADOS ratings. In all cases, one parent completed the SCAS-P, reporting on their child's symptoms of anxiety. The mean of the SCAS-P total score was 33.85 (SD=19.65) in the ASD sample. The means of subscales were as follow: Panic attack and agoraphobia: 4.75 (SD=4.48), Separation anxiety: 5.97 (SD=4.12), Physical injury fears: 4.87 (SD=3.24), Social phobia:

7.26 (SD=4.90), Obsessive compulsive: 5.00 (SD=3.93), Generalized anxiety disorder: 6.00 (SD=3.76).

Anxious sample. The anxiety-disorder group included data from parents of non-ASD, clinically anxious children and adolescents referred to the Berkshire Child Anxiety Clinic at the University of Reading, UK. SCAS-P data from this sample was collected from parents of 224 (150 male) children and adolescents with a mean age in months of 144.92 (SD=32.82, range: 8-17 years old). The mean total score of the SCAS-P was 38.47 (SD=17.02). The means of subscales were as follow: Panic attack and agoraphobia: 5.44 (SD=4.93), Separation anxiety: 7.56 (SD=4.26), Physical injury fears: 4.60 (SD=2.77), Social phobia: 9.11 (SD=4.35), Obsessive compulsive: 3.98 (SD=3.57), Generalized anxiety disorder: 7.78 (SD=3.63).

For that sample, on receipt of referral, parents completed a number of screening questionnaires to ensure that anxiety was the primary concern. This screening included the Social Communication Questionnaire to screen for characteristics of ASD (Rutter et al., 2003). Where children scored above clinical cut-offs (≥15) further investigations were conducted to ensure that children did not meet criteria for ASD. All children met diagnostic criteria for a primary anxiety disorder as established by the Anxiety Disorders Interview Schedule for DSM-IV structured interview (ADIS-C/P; Silverman & Albano, 1996), a structured diagnostic interview with well-established psychometric properties (Silverman et al., 2001). Where children met symptom criteria for a diagnosis they were assigned a clinical severity rating (CSR) ranging from 0 (complete absence of psychopathology) to 8 (severe psychopathology). As is conventional, overall diagnoses and CSRs were assigned if the child met diagnostic criteria on the basis of either child or parent report, and the higher CSR of the two was taken. Only those who met symptom criteria with a CSR of 4 or more (moderate psychopathology) were considered to meet diagnostic criteria. Assessors (psychology graduates) were trained on the administration and scoring of the ADIS and ADIS-C/P through verbal instruction, listening to assessment audio-recordings and participating in diagnostic consensus discussions. The first 20 interviews conducted were then discussed with a consensus team, led by an experienced diagnostician (Consultant Clinical Psychologist). The assessor and the consensus team independently allocated diagnoses and CSRs. Following the administration of 20 child or 20 parent interviews, inter-rater reliability for each assessor was checked, and if assessors achieved reliability of at least .85 they were then required to discuss one in six interviews with the consensus team (to prevent inter-rater

drift). Overall reliability for the team was excellent. As different assessors interviewed the parent and child simultaneously reliability figures for parent and child report were calculated separately. Reliability for presence or absence of diagnosis on the ADIS-C/P was kappa = .98 (child report), .98 (mother report); and for the CSR intra-class correlation = .99 (child report), .99 (mother report). Reliability for presence or absence of maternal diagnosis on the ADIS was kappa = .97; and for the CSR intra-class correlation = .99. Primary anxiety diagnoses for the sample were Generalised Anxiety Disorder (n=55), Social phobia (n=61), Separation Anxiety Disorder (n=40), Specific Phobia (n=41), Obsessive-Compulsive Disorder (n=3), Agoraphobia without Panic Disorder (n=9), Anxiety Disorder Not Otherwise Specified (ADNOS; n=5), and Panic Disorder (n=10).

6.2.3 Analysis plan

All analyses were conducted using SPSS 21 (IBM SPSS Statistics for Windows, Released 2012) and AMOS 21.0.0 (Arbuckle, 2012) software programs. There were missing values only in our anxious sample. There were no particular patterns in the missing data, allowing the data to be treated as Missing Completely at Random (MCAR). Participants with over 20% of missing item level data were removed (n=3) to minimalize randomness in our dataset. For the remaining participants the Maximum Likelihood Estimation (MLE) method of data imputation was used to complete the dataset.

Confirmatory factor analysis. In order to determine the factor structure of the SCAS-P, a confirmatory factor analysis (CFA), using structural equation modelling (SEM), in AMOS, was conducted with data from the anxious and ASD samples separately, and then in the combined sample, in order to determine the best-fitting factor structure and assess invariance. Six hypothesised models were tested subsequently. Five were the DSM-IV-based symptom models suggested by Nauta et al. (2004) including: (1) one factor, (2) six uncorrelated factors, (3) six correlated factors, (4) six correlated factors and one higher order factor, and (5) five correlated factors and generalized anxiety as one higher-order factor. For anxiety disordered children, as suggested by Nauta et al. (2004), support was found for six intercorrelated factors (separation anxiety, generalized anxiety, social phobia, panic/agoraphobia, obsessive–compulsive disorder, and fear of physical injuries) and a model with generalized anxiety as the higher order factor for the other five factors. There is no support in the literature that either of the models would fit ASD sample. The sixth model tested in this study was based on work done by Jamieson et al. (unpublished thesis, 2012) who suggested that five correlated factors (with GAD subscale excluded) might be the best-fitting factor structure for children and adolescents with ASD. All models were tested in

order to establish whether any of the hypothesised models would provide the fitting factor structure for either of the samples.

Model fit was evaluated using established recommendations identified as "best behaved" on the basis of previous research (Brown, 2006, p.85; Hu & Bentler, 1999). For example, we followed recommendations that χ^2/df ratio (Bryant & Yarnold, 1995) should be close to zero and that Root mean square error of approximation (RMSEA) values close to .06 represent good fit (Hu & Bentler, 1999), whilst values less than .08 are indicative of acceptable fit, and values between .08 and .10 represent poor model fit (Browne & Cudeck, 1993). It is recommended that the Comparative Fit Index (CFI) is greater than .95, but a level greater than 0.9 being acceptable (Hu & Bentler, 1999). It is also recommended that the Tucker-Lewis Index (TLI) is greater than .90 to demonstrate good fit (Brown 2006). The non-significant Chi-square (χ^2) statistic (Brown, 2006) may be used an indicator of fit, however, because it is greatly influenced by sample size (Stevens, 2002), we did not use in isolation from other recommended goodness of fit indices. The chi-square difference test was also used to compare competing models.

Measurement invariance. The measurement invariance technique can be implemented by running a multi-group analysis of the factor structure that underlies the data of two groups (Byrne & Campbell, 1999). The following sequence of four nested models is usually tested (see Cheung & Rensvold, 2002; Schmitt & Kuljanin, 2008): configural invariance; metric invariance; scalar invariance; and residual (uniqueness) invariance. In the configural invariance model, the same factor structure is implied for two or more groups of participants entered into the analysis. The values of the parameters (i.e. factor loadings, intercepts, residual variances) may vary across the groups, as no equality constraints are imposed. In the metric invariance model whether the values of the factor loadings are the same across groups is tested; hence item loadings are constrained to be equal across groups. Scalar invariance tests latent factor mean differences across groups and is evaluated by constraining the intercepts of measures to be the same across groups. In the residual model items unique variances are constrained to be equal across the two (or more) comparison groups. As suggested by Chen (2007), suggested differences in both CFI (delta CFI < 0.01) and RMSEA (RMSEA < 0.015) values were considered when comparing two nested models e.g. metric and scalar invariance.

6.3 Results

6.3.1 Preliminary and descriptive statistics

Examining the SCAS-P samples, anxious and ASD participants did not significantly differ on age. A significant difference was found for gender, with more female participants in the anxious sample. However, this difference represents the general sex ratio typical for the ASD population, with more males than females diagnosed with the condition (Werling & Geschwind, 2013).

6.3.2 Confirmatory factor analysis

In the anxious, ASD and combined (both anxious and ASD) samples, six models, including: (1) one factor, (2) six uncorrelated factors, (3) six correlated factors, (4) six correlated factors and one higher order factor, (5) five correlated factors and generalized anxiety as one higher-order factor, and (6) five correlated factors (with GAD subscale excluded), were tested. The goodness of fit indices are summarised in Table 6.1.

Overall, fit indices fell below the generally recommended ranges for good fit in each model. Due to poor models' fit subsequent invariance testing was not conducted as there was not enough evidence to assess invariance.

Table 6.1 Fit indices for six hypothesised models for the anxious, ASD and combined sample

Hypothesised Model:	χ^2	df	χ^2/df	*p*	CFI	TLI	RMSEA
Model 1: one factor							
ANX	2250.07	665	3.38	<0.001	.53	.50	.103
ASD	2428.89	665	3.65	<0.001	.66	.64	.097
Combined	3984.07	665	5.99	<0.001	.60	.58	.099
Model 2: six uncorrelated factors							
ANX	2171.05	665	3.27	<0.001	.55	.53	.101
ASD	2833.2	665	4.26	<0.001	.58	.55	.107
Combined	4173.27	665	6.28	<0.001	.58	.56	.102
Model 3: six correlated factors							
ANX	1685.40	650	2.59	<0.001	.69	.67	.085
ASD	1908.08	650	2.94	<0.001	.76	.74	.083
Combined	2855.66	650	4.39	<0.001	.73	.71	.082
Model 4: six correlated factors and one higher order factor							
ANX	1703.40	659	2.59	<0.001	.69	.67	.084
ASD	1937.68	659	2.94	<0.001	.75	.74	.083
Combined	2878.88	659	4.37	<0.001	.73	.72	.081
Model 5: five correlated factors and generalized anxiety as one higher-order factor							
ANX	1711.32	661	2.59	<0.001	.69	.67	.084
ASD	1941.39	661	2.94	<0.001	.75	.74	.083
Combined	2880.18	661	4.36	<0.001	.73	.72	.081
Model 6: five correlated factors (with GAD subscale excluded)							
ANX	1134.13	454	2.49	<0.001	.73	.70	.082
ASD	1257.59	454	2.76	<0.001	.79	.77	.079
Combined	1839.60	454	4.05	<0.001	.77	.75	.077

Note: Recommended goodness of fit indices values demonstrating good model fit: $\chi 2/df$ ratio close to zero, RMSEA <0.6, CFI >0.95 and TLI >0.9 (Brown, 2006; Hu & Bentler, 1999).

6.3.3 Post-hoc analysis

Due to the poor model fit with any of the six hypothesised models, we investigated the factor structure of the SCAS-P in the anxious and ASD samples with exploratory factor analysis (EFA). Parallel analysis and Velicer's minimum average partial (MAP) test were

performed to determine the number of components in the factor analyses. These validated procedures are superior to the eigenvalues greater-than-one rule (O'Connor, 2000). In the ASD and anxious sample parallel analysis indicated an eight factor solution. The MAP test indicated six factors in the ASD sample and seven factors in the anxious sample. When differences in test results emerge, optimal decisions should be made after considering the results of both analytic procedures bearing in mind that the MAP test tends to underextract the number of factors, whereas parallel analysis tends to overextract the number of factors (O'Connor, 2000). In both the eight and seven factor solutions in the ASD sample and the eight factor solution in the anxious sample, one factor consisted only of two items. The six factor solution in the ASD sample and seven factor solution in the anxious sample were considered as the most optimal. The summary of factors loading can be find in Table 6.2 and Table 6.3. Maximum Likelihood extraction with oblique rotation was used because high correlations between the components were found (above .4 and below -.4 in both groups).

Table 6.2 Rotated factor loadings in Exploratory Factor Analysis of SCAS-P in ASD sample

Item	Content	Communalities	Factor 1	Factor 2	Factor 3	Factor 4	Factor 5	Factor 6
			R^2 = 33.46	R^2 = 6.27	R^2 = 5.05	R^2 = 4.41	R^2 = 4.19	R^2 = 3.29
			E = 12.72	E = 2.38	E = 1.92	E = 1.67	E = 1.59	E = 1.25
4	Feeling afraid	.67	**.51**	-.12	-.04	.41	.05	.04
36	Bothered by bad or silly thoughts or pictures	.70	**.47**	-.32	-.11	.00	-.03	.30
17	Bad or silly thoughts	.54	**.44**	-.34	.05	.03	.04	.21
20	Something bad will happen to him/her	.59	**.41**	-.33	-.13	.15	.04	.05
26	What other people think of him/her	.74	.03	**-.84**	-.08	-.18	-.02	.08
9	Make a fool	.68	.02	**-.77**	-.06	.01	-.00	.07
10	Do badly at school	.61	.03	**-.71**	-.12	-.00	.03	-.02
6	Take a test	.47	.02	**-.59**	-.05	.20	.02	-.08
31	Talk in front of the class	.43	-.04	**-.54**	-.03	-.02	.21	.05
18	Heart beating really fast	.79	-.07	-.04	**-.91**	.08	-.04	-.09
32	Heart suddenly starting to beat too quickly	.78	-.03	-.01	**-.89**	.04	-.12	.09
12	Can’t breathe	.41	-.07	-.08	**-.53**	-.05	.13	.10
30	Becoming dizzy or faint	.42	-.00	-.08	**-.44**	.05	.20	.06
5	Own at home	.55	-.02	-.09	-.11	**.69**	-.01	-.06
8	Being away from parent	.57	-.10	-.14	-.10	**.59**	.05	.11
2	Dark	.43	.20	-.02	-.04	**.58**	.01	-.02
14	Sleep on his/her own	.39	.07	.06	-.09	**.58**	.08	-.05
38	Stay away from home overnight	.45	.01	-.09	.02	**.47**	.25	.06

19	Tremble or shake	.55	.28	.18	-.27	-.05	**.55**	-.02
25	Travel in the car, or on a bus or train	.46	-.07	-.09	-.16	.05	**.52**	.05
22	Feels shaky	.57	.26	-.03	-.18	-.11	**.51**	.13
21	Doctor or dentist	.29	-.04	-.11	.11	.12	**.45**	.06
28	Scared for no reason	.64	.30	.00	-.04	.12	**.45**	.24
27	Crowded places	.46	-.14	-.20	-.05	.14	**.40**	.18
35	Do some things over and over again	.60	-.10	-.05	.05	-.06	.04	**.80**
37	Certain things in just the right way	.55	.16	.02	-.04	.03	.03	**.65**
13	Keep checking	.49	.00	-.04	-.11	.01	-.02	**.64**
24	Think special thoughts to stop	.39	.16	.05	-.22	-.05	.07	**.43**
1	Worries about things	.56	.39	-.36	.04	.25	-.02	.12
3	Funny feeling in stomach	.37	.18	-.18	-.25	.22	.07	.00
7	Public toilets and bathrooms	.46	-.21	-.23	-.09	.24	.39	.01
11	Something awful will happen to someone in the family	.48	.21	-.33	-.11	.28	-.22	.22
15	School in the mornings	.34	.09	-.34	-.07	.05	.27	-.02
16	Dogs	.08	-.12	.10	.03	.21	.01	.14
23	Heights	.20	.15	.10	-.09	.27	.10	.09
29	Insects or spiders	.16	.02	-.12	.01	.22	.11	.11
33	Suddenly get a scared feeling	.56	.35	.02	-.22	.07	.26	.22
34	Small closed places	.25	-.07	-.06	-.05	.16	.33	.08

Note: loading derived from Maximum Likelihood estimation with Oblimin rotation. Content – summarized items content. E – Eigenvalue. Communalities reported are post-extraction. Reported R^2 and E derived from unrotated factor solution. **Bold loadings** > |.40|.

Table 6.3 Rotated factor loadings in Exploratory Factor Analysis of SCAS-P in anxious sample

Item	Content	Communalities	Factor 1 R^2 = 25.66 E = 9.57	Factor 2 R^2 = 8.32 E = 3.16	Factor 3 R^2 = 6.35 E = 2.41	Factor 4 R^2 = 5.50 E = 2.09	Factor 5 R^2 = 4.16 E = 1.58	Factor 6 R^2 = 3.79 E = 1.44	Factor 7 R^2 = 3.69 E = 1.40
32	Heart suddenly starting to beat too quickly	.71	**.86**	.10	-.06	.04	.02	.05	-.12
18	Heart beating really fast	.67	**.81**	-.02	-.04	.04	-.05	.06	.02
12	Can't breathe	.63	**.75**	.07	.01	.09	.02	.00	.06
19	Tremble or shake	.51	**.51**	-.09	-.17	-.17	-.04	-.10	.16
22	Feels shaky	.51	**.42**	-.07	-.17	-.30	-.06	-.19	.17
9	Make a fool	.71	-.06	**.83**	-.09	-.01	-.07	-.02	-.08
26	What other people think of him/her	.69	-.03	**.83**	-.02	-.01	-.14	.06	-.05
10	Do badly at school	.59	.01	**.76**	.03	.04	-.01	.04	.06
31	Talk in front of the class	.41	.05	**.58**	-.04	-.15	.12	-.05	.02
6	Take a test	.37	.21	**.52**	.02	-.01	.09	-.03	.07
37	Certain things in just the right way	.84	-.01	-.12	**-.93**	.00	-.08	.01	-.06
35	Do some things over and over again	.56	.03	.07	**-.71**	-.02	.01	-.08	.02
13	Keep checking	.47	.24	.21	**-.48**	.15	-.00	.07	-.05
5	Own at home	.49	.07	-.14	-.06	**.58**	-.02	-.25	.02
14	Sleep on his/her own	.43	-.04	-.06	-.10	**.55**	-.10	-.19	.03
2	Dark	.36	-.04	.08	-.01	**.54**	-.15	-.02	.08
17	Bad or silly thoughts	.65	.04	.06	-.07	.03	**-.74**	-.02	-.00
36	Bothered by bad or silly thoughts or pictures	.66	-.04	-.05	-.19	.06	**-.74**	.02	.04

20	Something bad will happen to him/her	.52	.23	.13	.08	.16	**-.45**	-.05	.14
38	Stay away from home overnight	.53	-.02	.01	-.16	.19	.11	**-.65**	.09
8	Being away from parent	.54	.03	.08	-.05	.24	-.07	**-.61**	-.02
15	School in the mornings	.38	.01	.23	-.03	-.14	-.07	**-.48**	-.06
4	Feeling afraid	.48	.10	-.02	.03	.23	-.26	**-.47**	-.04
3	Funny feeling in stomach	.42	.17	.02	.07	-.11	-.15	**-.46**	.13
33	Suddenly get a scared feeling	.50	.15	-.03	-.05	-.15	-.30	**-.45**	.06
34	Small closed places	.47	.05	-.02	-.05	.08	.09	-.01	**.67**
25	Travel in the car, or on a bus or train	.39	.01	.10	-.04	-.14	-.07	-.19	**.48**
27	Crowded places	.45	.01	.19	-.07	-.19	-.14	-.18	**.42**
23	Heights	.21	.06	-.12	.02	.03	-.08	.12	**.41**
1	Worries about things	.44	.09	.32	-.05	.18	-.22	-.24	.04
7	Public toilets and bathrooms	.35	-.01	.20	-.05	.19	.18	-.25	.38
11	Something awful will happen to someone in the family	.47	.27	.12	.07	.31	-.28	-.20	.00
16	Dogs	.12	.04	-.07	-.04	.30	.04	.15	.03
21	Doctor or dentist	.24	.10	.09	.04	.10	-.08	-.04	.36
24	Think special thoughts to stop	.46	.21	-.08	-.20	.19	-.11	.03	.35
28	Scared for no reason	.43	.16	-.05	-.05	-.06	-.26	-.37	.14
29	Insects or spiders	.26	-.08	.19	-.18	-.02	-.11	.26	.31
30	Becoming dizzy or faint	.43	.39	.04	-.04	-.25	-.15	-.03	.21

Note: loading derived from Maximum Likelihood estimation with Oblimin rotation. Content – summarized items content. E – Eigenvalue. Communalities reported are post-extraction. Reported R^2 and E derived from unrotated factor solution. **Bold loadings** > |.40|.

For both groups the social phobia factor was derived and was very similar to the original social phobia factor (Nauta et al., 2004), with only item 7 ('My child is afraid when (s)he has to use public toilets or bathrooms') not loading onto that factor. Also an OCD factor was derived that was similar to the original suggested by Nauta and colleagues (2004), however it consisted of only four items in the ASD group and three items in the anxious group. For the ASD group the other four factors comprised mostly of items belonging to the OCD, GAD, panic attack and agoraphobia, and separation anxiety subscales. Interestingly, panic attack and agoraphobia items loaded on two different factors. One factor included four items (item 19 'My child suddenly starts to tremble or shake when there is no reason for this', item 25 'My child feels scared if (s)he has to travel in the car, or on a bus or train', item 27 'My child is afraid of being in crowded places (like shopping centres, the movies, buses, busy playgrounds)' and item 28 'All of a sudden my child feels really scared for no reason at all') grouped together with one GAD item (item 22 'when my child has a problem, (s)he feels shaky) and one physical injury item (item 21 'My child is scared of going to the doctor or dentist'). A second factor related to the majority of the physiological symptoms of anxiety (item 32 'My child's complains of his/her heart suddenly starting to beat to quickly for no reason', item 12 'My child complains of suddenly feeling as if (s)he can't breathe when there is no reason for this', item 30 'My child complains of suddenly becoming dizzy or faint when there is no reason for this' and GAD item 18 'when my child has a problem, (s)he complains of his/her heart beating really fast'). These three items relate to physiological symptoms of panic experience, including the ability to recognise those symptoms (e.g. increased heart beat) and communicate those changes in the body functions to others.

A split in the original panic and agoraphobia factor was also found in the anxious sample. Some of the items loaded on to a physiological symptoms of anxiety factor (with additional items from the original GAD factor) while the other factor was more agoraphobia specific (e.g. item 34 'My child is afraid of being in small closed places, like tunnels or small rooms'). Also OCD items separated into two distinct factors in the anxious typically developing group, with one relating to compulsions (e.g. item 37 'My child has to do certain things in just the right way to stop bad things from happening'), the other to obsessive thoughts (e.g. item 17 'My child can't seem to get bad or silly thoughts out of his/her head'). Another factor that was indicated for the anxious group comprised of various separation anxiety, GAD and panic attack and agoraphobia items (e.g. item 33 'My child worries that (s)he will suddenly get a scared feeling when there is nothing to be afraid of'). The last factor consisted of two separation anxiety items (item 5 'My child would feel afraid of being on his/her own at home' and item 14 'My child is scared if (s)he has to sleep on his/her own')

and one physical injury fears item (item 2 'My child is scared of the dark'). Items from across a range of the original subscales loaded on to the other factors in the anxious sample, with factor four including items ranging from separation anxiety to being scared of darkness, and factor five including items related to anxious thoughts and factor seven encompassing specific phobias.

6.4 Discussion

The first aim of this study was to determine the factor structure for the SCAS-P in a sample of young people with ASD and to compare it with the factor structure derived from a sample of clinically-anxious young people without ASD, and in the combined sample to ensure adequate fit to consider invariance. However, due to poor model fit and inability to find an adequate baseline model for further between-group model testing, measurement invariance analyses could not be performed. Inability to find a model with a fixed number of factors in each group for the measure that has an established factor structure for use with typically developing samples was an unexpected outcome. Similarly, White et al. (2015) could not pursue the multigroup invariance factor analysis on the MASC parent version (but could on the MASC self-report), because the CFA undertaken on the typically developing anxious youth did not confirm the conventional MASC-P structure. It is important to bear in mind that parents might not always be aware of all anxiety-related behaviours that children exhibit, unless they verbalize their subjective and individual experiences. It is likely, particularly for our ASD sample, that parents were not aware of some of the symptoms or their severity and frequency. The reason why we could not find the baseline model of the SCAS-P in the anxious sample is unknown.

Using EFA, a six-factor model was established for the ASD sample, and a seven-factor model was found to describe the anxious sample best. The findings here for both groups differ from the SCAS-P factor structure suggested by Nauta et al. (2004), who found that six correlated factors fit the data obtained from the parents/caregivers of anxiety-disordered children best. Indeed, for the clinically anxious group we only found partial support for the panic attack and agoraphobia, OCD and social phobia factors. However, even within these factors some anomalies were found. Even less support for the original factor structure of the SCAS-P was found in the ASD sample.

The study showed limited support for the original factor structure of the SCAS-P. It is a novel, inconsistent with previous emotional functioning and personality literature (e.g., Hoelzle & Meyer, 2009; Hopwood & Donnellan, 2010; O'Connor, 2002) finding. Some concerns, however, have been raised previously with regards to the validity of the SCAS-P,

particularly of the GAD subscale for use with typically developing children. Spence, Rapee, McDonald and Ingram (2001) argued that this sub-scale could indicate more negative affect and autonomic responding than generalized anxiety, and found little support for a separate GAD-subscale. The content validity of the GAD subscale has been also questioned because it lacks overt reference to excessive worry (Chorpita et al., 1997), which is considered to be a central feature of GAD in childhood and adolescence. Our findings support these concerns, as a distinct GAD factor was not found in either our anxious or ASD samples. The physical injury fear factor was also not established for either of the samples. The reliability of the subscale, however, has been questioned previously, with unacceptable to questionable Cronbach's alpha reported across community and clinical samples in various countries (Arendt et al., 2014; Whiteside & Brown, 2008; Zainal et al., 2014). Although in the RCADS, a revised version of the SCAS-P, the measurement properties of GAD appeared to have improved (Wigham & McConachie, 2014), evidence on psychometric properties of this tool remains patchy and requires further investigations.

According to our findings, further work is needed on the SCAS-P to establish its reliability and validity, particularly when used with the ASD population. Zainal and colleagues (2014) reported in their preliminary investigation, the SCAS-P might be a useful screening tool of anxiety in children with ASD when assessing elevated anxiety symptoms and relying on the total score. We suggest that a further caution is needed when using the tool to assess particular anxiety subtypes and make cross-groups comparisons between children with ASD and children diagnosed with anxiety disorder based on the SCAS-P scores. Although Wigham and McConachie (2014) reported that the SCAS-P was one of the tools to have the most robust measurement properties in comparison to other measures, there was lack of evidence for a number of reliability and validity characteristics of the questionnaire.

6.4.1 Study limitations

An important limitation to this study is that our anxious sample consisted of clinically referred individuals; and our ASD sample consisted of participants recruited to various studies, hence our sampling procedure might have impacted our findings. Further qualitative work is recommended to explore the validity of SCAS-P items in ASD samples. In line with other studies we recommend that the GAD and physical injury fears subscales require additional reliability and validity checks across clinical and community samples. Adaptation of the questionnaire is needed for reliable and valid use with ASD individuals. Qualitative interviews with parents should be conducted to better understand the context and particular situations in which caregivers base their answers.

6.4.2 Conclusions

The SCAS-P has been developed and validated for use with typically developing youth. To use the scale as a reliable measure of anxiety in young people with ASD further work is needed. Researchers and clinicians should not rely solely on the scores obtained from the SCAS-P when assessing anxiety symptoms in individuals with ASD. Further and more systematic quantitative and qualitative research would be required to turn the SCAS-P into a robust measure of anxiety for use in ASD practice or research.

Chapter 7. Relationships between sensory atypicalities, repetitive behaviours, anxiety and intolerance of uncertainty

7.1 Background

There is evidence suggesting that sensory atypicalities are associated with other symptoms in both ASD and WS e.g. sensory hyporesponsiveness is more often associated with core features of ASD such as communication impairment, emotional, cognitive, and behavioural problems while social awareness difficulties and affective disorders are associated with hyperresponsiveness (Chapter 2); also a higher degree of sensory processing difficulties is associated with more difficulties in executive functioning, temperament, adaptive functioning, problem behaviours and repetitive behaviours in WS (Chapter 3). The associations, however, within and between the symptoms are still not well understood and the degree of the co-occurrence of these features in the disorders is still not well explored. Examining the complexity of the mechanisms underlying the relationship between sensory atypicalities, repetitive behaviours and anxiety across neurodevelopmental disorders would help us establish their symptom-specificity and potentially develop treatment protocols tailored to a specific disorder or cluster of symptoms.

7.1.1 Sensory atypicalities and repetitive behaviours

Investigating the relationship between sensory features and repetitive behaviours has recently gained more research attention as a high degree of co-occurrence between sensory atypicalities and repetitive behaviours has been reported in a number of studies. For example, Gabriels et al. (2008) divided their ASD sample into two subgroups – participants with either high or low levels of repetitive behaviours. Further analysis showed that the high repetitive behaviours subgroup showed more sensory atypicalities than the subgroup with less repetitive behaviours. That relationship was also found and reported in other studies (e.g. Baker et al., 2008; Chen et al., 2009; Joosten et al., 2009). The relationship, however, was less clear when the associations between types of repetitive behaviours and various sensory features were examined. In the recent review investigating psychological correlates of the sensory processing patterns (Chapter 2), two types of repetitive behaviours ('lower level' such as motor repetitions and stereotyped behaviours, and 'higher level' relating to insistence on sameness and circumscribed interests; Turner, 1999) were found to be associated with different sensory symptoms, such as hyperresponsiveness, hyporesponsiveness and sensory

seeking (over-reactivity, under-reactivity and craving or fascination with certain stimuli; Boyd et al., 2010). The reported pattern of associations, hence, is very mixed. There is also very limited evidence supporting a relationship between atypicalities within sensory modalities and presence of restricted and repetitive behaviours, with only tactile and auditory modalities associated with RRBs in individuals with ASD (Chen et al., 2009; Foss-Feig et al., 2012). Moreover, Riby et al. (2013) concluded that some repetitive behaviours (e.g. repetitive movement) may be a consequence of specific types of sensory problems (e.g. tactile sensitivity) in individuals with WS. Considering a cross-syndrome approach, sensory abnormalities may be associated with different repetitive behaviours in ASD and WS and further exploration of these relationships is needed.

With regards to the relationship between sensory processing and repetitive behaviours in WS, evidence is very limited as there is only one study which has investigated this association. Riby et al. (2013) reported that increased levels of sensory processing abnormality were associated with higher levels of repetitive behaviours. In particular, significant relationships were reported between repetitive movement and underresponsive/seeks sensation and tactile sensitivity; also taste/smell sensitivity was associated with both repetitive movement and sameness of behaviour. Riby and colleagues (2013) suggested that some repetitive movement behaviours were a consequence of tactile sensitivity in individuals with WS.

7.1.2 Sensory processing and anxiety

Anxiety is a common health concern in children with ASD affecting between 11%-84% (White et al., 2009) compared to 3-24% of typically developing children (Green & Ben-Sasson, 2010). Sensory processing abnormalities have been linked to anxiety in ASD (Ben-Sasson et al., 2008; Green et al., 2011; Liss et al., 2006; Pfeiffer et al., 2005). A strong relationship between hyper-responsiveness and anxiety has been reported several times (for review see: Chapter 2). Furthermore, research with toddlers indicated that sensory over-responsivity was not only stable across time, but also emerged earlier than anxiety, and predicted later development of anxiety (Green et al., 2012). The link between sensory processing abnormalities and anxiety has been strengthened by functional magnetic resonance imaging (fMRI) findings in ASD (Green et al., 2013), indicating that sensory sensitivity was related to increased activity in the amygdala and prefrontal areas (including orbitofrontal cortex, OFC). These brain areas are involved in emotion regulation and response to threat that directly link to anxiety (Green et al., 2013).

Elevated risk of anxiety has also been found in WS; more specifically this psychopathology is one of the most frequently co-occurring with the disorder (Rodgers et al., 2012). Anxiety disorders in children with WS, similar to those with ASD, include social phobia, generalized anxiety disorder or separation anxiety disorder (Rodgers et al., 2012). It has been reported that individuals with WS can develop an intense fascination for certain sounds that they found frightening as children (Levitin et al., 2005). Anxiety associated with certain sounds in children with WS has been also found by Leyfer et al. (2006). Leyfer and colleagues (2006) reported that the most common type of Specific Phobia present in WS individuals between 4 and 16 years of age was a phobia of loud noises. Yet, to date, there is a dearth of studies that have investigated the relationships between sensory processing difficulties and anxiety in WS.

7.1.3 Repetitive behaviours and anxiety

Vulnerability to anxiety in both ASD (White et al., 2009) and WS (Dykens, 2003) has been reported. Also, elevated levels of RRBs occur in both disorders (Rodgers et al., 2012), with up to 86% of individuals with WS (Davies et al., 1998) and all individuals with ASD (Lewis & Bodfish, 1998) engaging in some form of repetitive behaviours. However, to date, there is limited evidence suggesting that higher levels of anxiety are associated with restricted and repetitive behaviours in general (Rodgers et al., 2012), and with both repetitive motor behaviours (Rodgers et al., 2012) and insistence on sameness (Lidstone et al. 2014; Rodgers et al. 2012). Although it has been suggested in the literature (Semel & Rosner, 2003) that repetitive behaviours in WS might function to reduce anxiety, interestingly, the relationship between RRBs and anxiety was not found in WS (Rodgers et al., 2012). As Rodgers and colleagues (2012) proposed the lack of that association might suggest that high levels of RRBs may not play a role in presentation of anxiety in WS.

7.1.4 Sensory atypicalities, repetitive behaviours, anxiety and intolerance of uncertainty

There are only two studies to date, investigating the relationships between sensory atypicalities, repetitive behaviours and anxiety in individuals with ASD (Lidstone et al., 2014, Wigham et al., 2015). To our knowledge, there is no single study examining these associations in WS. As Lidstone and colleagues (2014) reported both Low Registration and Sensation Seeking were related to insistence on sameness behaviours and anxiety, however, sensation avoiding was a mediator between anxiety and insistence on sameness behaviours. Only in Wigham et al. (2015) intolerance of uncertainty was considered in understanding of the relationship between sensory atypicalities, repetitive behaviours and anxiety. Intolerance

of uncertainty may be presented as the way of perceiving information in uncertain situations and responding to it in a cognitive, emotional and behavioural way (Freeston et al., 1994). Those individuals who are intolerant of uncertainty are more likely to perceive everyday events as unacceptable and disturbing (Dugas et al., 2001). It has been shown that children with ASD not only have significantly higher levels of intolerance of uncertainly, but also that intolerance of uncertainty accounted for the increased levels of anxiety in those individuals with ASD (Boulter et al., 2014). According to Wigham and colleagues (2015) anxiety combined with intolerance of uncertainty mediates the relationship between sensory atypicalities and restricted and repetitive behaviours in ASD. These results highlight the presence of the inter-relationships between the phenomena and the complexity of the associations between sensory features, repetitive behaviours and anxiety in individuals with ASD.

7.1.5 Heterogeneity in ASD and WS

Although three subgroups of ASD, Asperger's disorder, autistic disorder and PDD-NOS (pervasive developmental disorder not otherwise specified) are replaced with a severity gradient to describe individuals with ASD (see Chapter 1), it has been recently suggested (Lane et al., 2014) that children with ASD can be classified meaningfully by their sensory differences. Research has suggested that sensory based phenotypes are not explained by differences in age, non-verbal intellectual ability or autism severity. The heterogeneity of sensory difficulties in WS has never been investigated.

7.1.6 Rationale

The empirical evidence to date indicates that there are relationships between sensory processing abnormalities and repetitive behaviours, between sensory features (sensory overresponsivity), anxiety and intolerance of uncertainty, and between repetitive behaviours and anxiety, and that these phenomena co-occur in individuals with ASD. Less is known with regards to the associations between sensory features, repetitive behaviours, anxiety and intolerance of uncertainty in WS, however, all these symptoms are very common in both disorders. The studies examining the relationships between repetitive behaviours and sensory atypicalities present a number of limitations, including not only the differences in defining and measuring repetitive behaviours, difficulties in distinguishing between repetitive behaviours and sensory features, but also in recruiting participants across the spectrum, age, and ability levels (Boyd et al., 2010; Glod et al., 2015). The heterogeneity of ASD and WS seems to be understudied, along with the findings suggesting that patterns of sensory

processing (Kern et al., 2006), repetitive behaviours (Richler et al., 2010) and emotional symptoms (Howlin, 2005) change in adolescence in individuals with ASD. Furthermore, investigating contributions of different sensory modalities to associations between sensory atypicalities and RRBs, anxiety and intolerance of uncertainty have been very rarely examined with only selected modalities included in analyses (e.g. touch only in Foss-Feig et al., 2012). Investigating relationships between sensory symptoms, repetitive behaviours, anxiety and intolerance of uncertainty is critical for establishing diagnostic criteria reflecting the complexity of the disorders and understanding the pathogenesis of ASD and WS.

7.1.7 Study aims and objectives

In the present study, the primary aim is to investigate the following research questions: (1) What are the patterns of sensory clusters in children with ASD or WS? (2) Are any of the patterns of sensory clusters syndrome-specific? (3) Are there sensory-pattern based subgroups that have higher levels of anxiety, intolerance of uncertainty or repetitive behaviours in children with ASD or WS? (4) Are there sensory-modality based subgroups that have higher levels of anxiety, intolerance of uncertainty or repetitive behaviours in children with ASD or WS? The secondary aim of the study is to examine the relationship between sensory atypicalities, repetitive behaviours, anxiety and intolerance of uncertainty in children with ASD and WS.

In regards to the primary aim, it is hypothesized that there would be distinct patterns of sensory clusters in children with ASD and children with WS, and that some of these may be syndrome-specific (at present the exact nature of those that will be syndrome-specific is unclear). Also the subgroup with the highest frequency of sensory processing difficulties would show the highest level of anxiety, intolerance of uncertainty and repetitive behaviours irrespective of ASD or WS diagnosis.

In regards to the secondary aim, it is hypothesized that sensory processing abnormalities would predict repetitive behaviours directly in both ASD and WS groups. Intolerance of uncertainty and anxiety would mediate this relationship in the ASD and WS groups. A greater role of intolerance of uncertainty is expected in the ASD sample.

7.2 Methods

7.2.1 Recruitment

Children between 4 and 9 years of age, with ASD or WS, and their parents, were invited to take part in the research project (‘Touch, hear, react’ study; see Chapter 4 for more

details). Those children, who apart of their main diagnosis had any other comorbid diagnosis of neurodevelopmental disorder or had visual, hearing or motor impairments, were excluded from the study. Families whose children met the study criteria were initially sent information about the study by email or letter, and reminders were sent to non-responders. Children and their parents participated on a voluntary basis. Parents were asked to give consent for themselves and their child to take part in the study. Additionally, a verbal assent was sought from each child. Favourable ethical opinion was granted by the Newcastle University Faculty of Medical Sciences Ethics Committee.

7.2.2 Participants

Twenty-three children with ASD and seventeen children with WS and their parents were recruited to the study. Children with ASD were recruited through two different routes: local mainstream and special needs primary schools from the North East of England and a newsletter distributed by a local branch of 'Contact a Family'. 'Contact a Family' is a national charity for families with disabled children which on daily basis provides information, advice and support to the families. The charity also releases a weekly newsletter in which research studies can be advertised. The advertisement used for this study can be found in Appendix Q. All children with ASD had previously been diagnosed with ASD based on a multidisciplinary team assessment following the guidelines of the UK National Autism Plan for Children (Le Couteur, 2003) as stated by the parents. The questionnaire data were returned for nineteen children with ASD. Out of those, data from the Social Responsiveness Scale - Second Edition (SRS-2; Constantino & Gruber, 2012) were available for sixteen children (for three children total raw score was not calculated due to large amount of missing data), with a range between 62 and 175, mean=111.13, SD=35.94. Eight children fell into mild to moderate, and 8 into severe range. Children for whom the SRS-2 total score could not be calculated, did not differ on gender, age and any sensory variable compared to children for whom the SRS-2 data were available. Children with WS were recruited via the Williams Syndrome Foundation which actively supports research into the condition. All WS children had previously been clinically diagnosed with the syndrome with the diagnosis confirmed by positive fluorescent in situ hybridization testing (FISH). The questionnaire data were obtained for sixteen children with WS. The SRS-2 raw total scores ranged from 34 to 141, mean=80.0, SD=25.54. Three children fell within normal, ten within mild to moderate and three within severe range.

7.2.3 Measures

The parents were asked to complete a set of questionnaires, including:

1. Sensory Profile (SP; Dunn, 1999) - a caregiver questionnaire that measures a child's sensory processing abilities. The questionnaire consists of 125 items, rated on a five-point Likert scale, ranging from almost never to almost always (see Chapter 4).

2a. Spence Children's Anxiety Scale-Parent Version (SCAS-P; Spence, 1998) is a 38-item questionnaire. Parents rate the frequency of occurrence of anxiety symptoms on a four-point Likert-type scale, ranging from 0 (never) to 3 (always) (see Chapter 6 for more details).

2b. Preschool Anxiety Scale (PAS, Spence et al., 2001) is a version of SCAS-P adapted for use with very young children. It consists of 28 anxiety items, rated on a 5-point scale from 0 'not at all' to 4 'very often true'. The PAS provides the total score and subscale scores for generalized anxiety, social anxiety, obsessive compulsive disorder, physical injury fears and separation anxiety Appendix R).

In both SCAS-P and PAS higher scores indicate higher level of anxiety in an individual.

3. Anxiety Scale for Children-ASD, parent-version (ASC-ASD©, Rodgers et al., 2016) – is a 24-item anxiety questionnaire assessing anxiety symptoms specific to ASD population. Based on the findings presented in Chapter 6, we decided to include an additional anxiety measure in order to ensure that symptoms specific to children with ASD are reported and considered. It takes around 5 minutes to complete the ASC-ASD and the measure has four sub-scales: Separation Anxiety, Uncertainty, Performance Anxiety and Anxious Arousal (Appendix S). The ASC-ASD was derived from the parent version of Revised Child Anxiety and Depression Scale (RCADS; Chorpita et al., 2000), which is a well-validated measure of anxiety symptoms and depression developed for use with typically developing children. Higher scores indicate higher level of anxiety.

4. Repetitive Behaviour Questionnaire (RBQ; Turner, 1995) – is a 33-item questionnaire designed for parents of children with or without ASD. Parents are asked to rate on a 3 or 4 point Likert scale the severity or frequency of repetitive behaviours that their child engaged in over the last month (Appendix T). Higher scores reflect grater engagement in repetitive behaviours. Behaviours reported include repetitive movements, sameness behaviour, circumscribed interests and repetitive use of language. The RBQ has been previously used with children with ASD between 4 and 16 years old (Barrett et al., 2004; Honey et al., 2012; Zandt et al., 2009).

5. Social Responsiveness Scale – Second Edition (SRS-2; Constantino & Gruber, 2012) - a 65-item rating scale which takes 15–20 minutes to complete. It is a parent-report of autistic trait that covers unusual interpersonal behaviours, communication or

repetitive/stereotyped behaviours. The SRS-2 describes a degree of autistic social impairment and the severity of autistic symptoms, with higher scores suggesting grater severity of symptoms.

The children underwent a cognitive ability assessment, comprising of:
1. Raven's Coloured Progressive Matrices (RCPM; Raven et al., 1998) - a jigsaw-like test assessing reasoning ability, designed for children between 4 and 11 years old, including those with intellectual disability and limited language skills. The child is presented with a pattern with a missing piece and is asked to choose one of the six pattern blocks that best fits into the missing gap.
2. British Picture Vocabulary Scale: Third Edition (BPVS3; Dunn et al., 2009) - a tool assessing a child's receptive vocabulary that can be used with children as young as 3 years old. During the assessment, an examiner says a word and the child is asked to select one from four pictures that best illustrate the word. The measure has been used with children with autism and other communication difficulties.

7.2.4 Data analysis

SPSS 22 was used to analyse the data. Descriptive statistics were calculated for the ASD and WS samples. A hierarchical agglomerative cluster analysis was performed as an exploratory method of grouping data based on high associations between grouped objects. First, a hierarchical cluster analysis using Ward's method with squared Euclidean Distance was carried out to determine the optimum number of cluster for the sample used. Second, the k means technique was applied to include all the participants in the final analysis. This two-stage approach is commonly used in ASD research (Ben-Sasson et al., 2008; Liss et al., 2006). Pearson's two tailed correlations were calculated to examine the relationship between sensory processing abnormalities, anxiety, intolerance of uncertainty and repetitive behaviours; and Bonferroni corrections were applied. T-scores from the SCAS-P/PAS were entered as an anxiety measure and the Uncertainty subscale of ASC-ASD was used as a measure of intolerance of uncertainty. Pearson's two tailed correlations were also calculated to examine the relationship between the outcome variables, and between the outcome variables and demographic characteristics. Finally, PROCESS (Hayes, 2012) was used to test serial mediation models.

The normality of the data was tested before performing correlational analyses. As indicated by the Kolmogorov-Smirnov test of normality, the distribution of the data was non-

normal in the WS sample for some of the anxiety-related variables: combined SCAS-P and PAS subscales of GAD (D(14)=.24, p=.024), Social Anxiety (D(14)=.40, p<.001), and OCD (D(14)=.43, p<.001), and the following ASC-ASD variables: Total score (D(14)=.28, p=.004), Performance anxiety (D(14)=.31, p=.001), Anxious arousal (D(14)=.36, p<.001) and Uncertainty (D(14)=.30, p=.001). Also non-normal distribution of the data was found in the ASD sample for the repetitive behaviours variables: RBQ total (D(14)=.24, p=.029), RBQ Sensory Motor (D(14)=.24, p=.028), RBQ Sameness (D(14)=.30, p=.001), as well as for some anxiety variables: combined SCAS-P and PAS subscales of GAD (D(14)=.25, p=.016), Social anxiety (D(14)=.25, p=.022), Separation anxiety (D(14)=.24, p=.026), ASC-ASD Performance anxiety (D(14)=.35, p<.001) and ASC-ASD Anxious arousal (D(14)=.28, p=.005). Bootstrapping procedure was implemented in all analyses to minimize the measurement error.

7.3 Results

Descriptive characteristics and descriptive scores on the outcome measures are shown in Table 7.1 and Table 7.2.

Table 7.1 Descriptive statistics (mean (SD)) of participant characteristics

	WS (n=16)	ASD (n=19)
Gender: male	8	16
Age in months	85.13 (22.56)	84.74 (21.81)
Verbal IQ	85.38 (8.67)	89.73 (13.62)
Non-verbal IQ	78.75 (6.29)	96.82 (11.89)
SRS-2 total score	80.00 (25.54)	111.13 (35.94)

Note: due to low ability level in the WS sample verbal IQ data were collected only for 4 participants and non-verbal IQ data were available for 8 children; in the ASD sample verbal and non-verbal IQ were assessed in 11 participants

Table 7.2 Mean scores on outcome variables

Outcome variable	WS (n=16)	ASD (n=19)
Sensory profile		
Registration	48.75 (14.32)	56.84 (12.88)
Seeking	87.06 (17.22)	87.63 (16.58)
Sensitivity	62.25 (14.17)	66.18 (14.83)[d]
Avoiding	93.94 (17.22)	89.73 (19.90)[b]
Auditory	22.56 (5.02)	20.35 (6.12)[d]
Visual	30.75 (5.86)	31.00 (6.36)[d]
Vestibular	41.88 (6.71)	42.16 (8.16)
Touch	65.06 (10.01)	63.47 (16.32)
Oral	36.19 (11.73)	41.74 (13.72)
RBQ		
Total score	12.00 (10.17)	23.00 (16.94)
Sensory/Motor	4.69 (4.25)	8.63 (5.52)
Insistence on Sameness	4.94 (4.85)	10.32 (7.30)
SCAS-P/PAS T-scores		
Total score	48.63 (12.69)	52.28 (17.67)[e]
Separation	53.44 (11.67)	56.94 (14.65)[e]
Physical injury fears	55.19 (11.33)	55.83 (13.39)[e]
Social Anxiety	44.63 (6.51)	51.06 (15.71)[e]
OCD	47.87 (13.87)	54.72 (20.16)[e]
GAD	58.00 (17.78)	53.94 (17.48)[e]
ASC-ASD		
Total score	13.00 (10.08)[a]	19.94 (18.37)[d]
Performance anxiety	1.13 (1.87)[b]	3.47 (5.71)[d]
Anxious arousal	2.33 (2.97)[b]	3.22 (4.53)[e]
Separation anxiety	3.57 (2.93)[a]	4.56 (3.88)[e]
Uncertainty	7.19 (5.98)[c]	10.17 (7.88)[e]

Note: [a] n=14, [b] n=15, [c] n=16, [d] n=17, [e] n=18; SCAS-P data were available for 11 WS and 11ASD participants, PAS data were available for 5 WS and 7 ASD participants

7.3.1 Cluster analysis

A hierarchical agglomerative cluster analysis was performed to identify subgroups based on sensory characteristics for WS and ASD groups. Ward's (1963) method was performed to determine clusters. In this procedure, each child in the beginning is considered as a separate cluster, and with each step is merged with the closest cluster (another child). Squared Euclidean distance was used to determine the similarity between the clusters (Hair & Black, 2000). Sensory variables that were obtained from the Sensory Profile were standardized to minimize the effect of unequal scaling on the clusters' determination. The hierarchical tree and the agglomeration schedule were examined to determine the optimal number the clusters for each set of sensory variables. The selected number of clusters was then examined with a k-means iterative partitioning cluster analysis method (Punj & Stewart, 1983). This procedure compliments the hierarchical method by indicating the stability of clusters (Hair & Black, 2000).

7.3.2 Sensory processing patterns by diagnostic group

In the next step of the analysis, two neurodevelopmental groups were examined separately. Two and four possible sensory processing pattern clusters were identified using hierarchical clustering. In the WS group, all participants kept their cluster membership in the two-cluster solution, while only 44% remained in the same cluster after the partitioning procedure. While in the ASD group, one participant changed cluster membership in the two-cluster-solution, whereas all remaining participants maintained their original cluster membership in the four-cluster solution after the partitioning procedure. However, as one of the clusters contained only one participant, the two-cluster solution was selected for the further analyses.

In the two-cluster solution for the WS group, the following clusters emerged: (1) a cluster with a low frequency of all sensory behaviours, with Registration having the lowest frequency (n=10; 'low-atypical' cluster); (2) a cluster with a higher frequency of all sensory behaviours (n=6; 'high-atypical' cluster). Figure 7.1 presents the SP sensory processing patterns z-score centered for each cluster for the two-cluster solution.

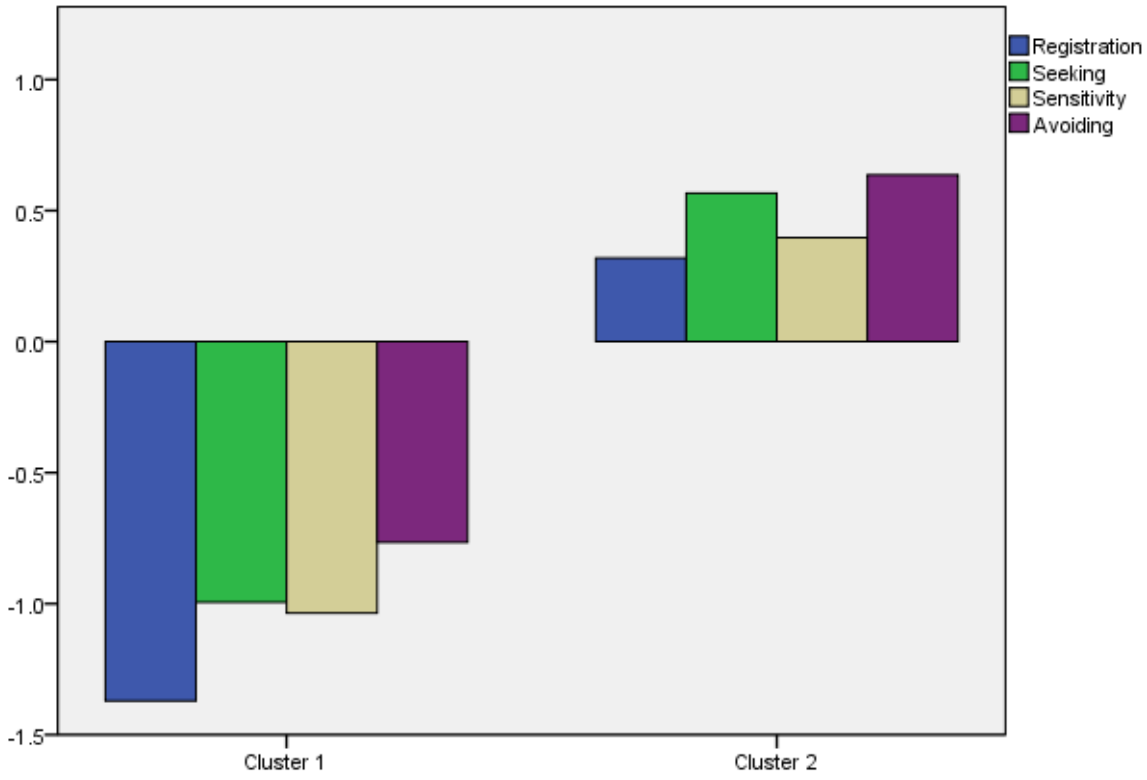


Figure 7.1 SP sensory processing patterns z-scores by cluster in a two-cluster solution for the WS group

In the two-cluster solution for the ASD group, similar clusters emerged: (1) a cluster with a higher frequency of all sensory behaviours (n=11; 'high-atypical' cluster); (2) a cluster with a low frequency of all sensory behaviours, with Avoiding having the lowest frequency (n=4; 'low-atypical' cluster). Figure 7.2 presents the SP sensory processing patterns z-score centered for each cluster for the two-cluster solution.

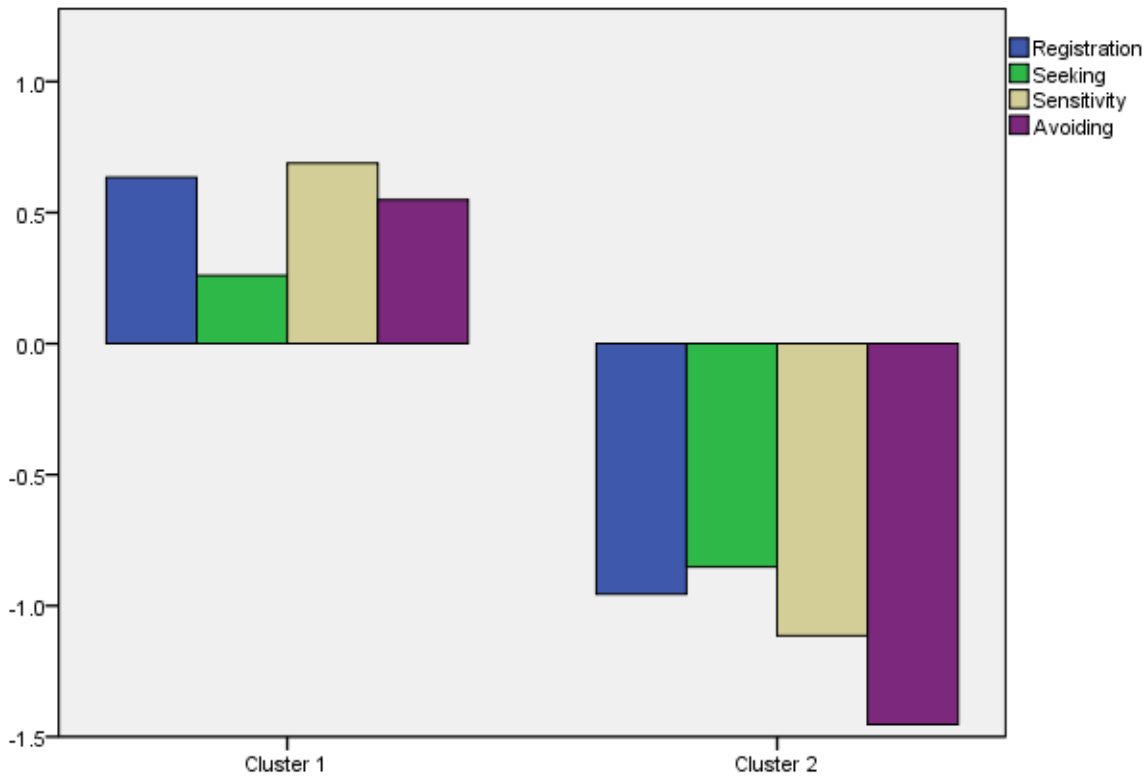


Figure 7.2 SP sensory processing patterns z-scores by cluster in a two-cluster solution for the ASD group

The two WS clusters did not differ on age ($F_{(1,2)}$=.17, p=.724), verbal IQ ($F_{(1,2)}$=3.44, p=.205), non-verbal IQ $F_{(1, 2)}$=1.56, p=.338 and SRS-2 total score ($F_{(1,2)}$=.78, p=.47). The difference in gender distribution was not computed by the SPSS.

The ASD clusters did not differ on age ($F_{(1,7)}$=.02, p=.905), gender distribution ($F_{(1,7)}$=1.17, p=.316), verbal IQ ($F_{(1,7)}$=1.80, p=.222), non-verbal IQ $F_{(1,7)}$=1.38, p=.279, but significantly differed in their SRS-2 total score ($F_{(1,7)}$=45.33, p<.001). The SRS-2 total score was significantly lower in 'low-atypical' cluster (m=87.67, sd=14.45) comparing to 'high-atypical' cluster (m=155.67, sd=13.87).

MANOVA on the non-standardized sensory processing pattern scores showed significant differences between the clusters for both the WS and ASD groups ($F_{(4,11)}$=8.24, p<.001 and $F_{(4,10)}$=16.99, p<.001 respectively). The summary of the findings can be found in Table 7.3.

Table 7.3 Summary of the MANOVA analysis of the non-standardized sensory processing pattern scores for WS and ASD groups

	WS				ASD			
Sensory processing patterns	'low-atypical' n=10	'high-atypical' n=6	F	F (SRS-2 as covariate)	'low-atypical' n=4	'high-atypical' n=10	F	F (SRS-2 as covariate)
Registration	57.60 (9.44)	34.00 (6.07)	29.64***	5.99*	61.55 (7.05)	35.50 (5.20)	44.78***	15.02**
Typical performance	3	0			4	0		
Probable difference	4	0			4	0		
Definite difference	3	6			3	4		
Seeking	96.80 (5.25)	70.83 (18.29)	18.43**	2.75	89.64 (16.60)	74.25 (10.08)	2.95	.008
Typical performance	1	0			2	0		
Probable difference	7	0			2	0		
Definite difference	2	6			7	4		

Sensitivity	70.00	49.33	15.91**	1.86	73.00	45.00	28.80***	8.93*
	(10.56)	(9.00)			(9.13)	(8.25)		
Typical performance	1	0			3	0		
Probable difference	5	0			3	0		
Definite difference	4	6			5	4		
Avoiding	103.60	77.83	17.81**	1.84	99.45	63.00	30.77***	8.42*
	(8.93)	(15.74)			(12.61)	(4.32)		
Typical performance	2	0			1	0		
Probable difference	3	0			4	0		
Definite difference	5	6			6	4		

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

For the WS participants MANOVA analysis exposed a significant cluster effect for the three RRB scores ($F_{(3,12)}$=8.25, p=.003), but non-significant cluster effects on the six SCAS/PAS scores ($F_{(6,9)}$=1.35, p=.331) and five ASC-ASD scores ($F_{(4,9)}$=1.66, p=.243). In the ASD group, significant cluster effects were found for the three RRB scores ($F_{(3,11)}$=4.13, p=.031) and six SCAS/PAS scores ($F_{(6,8)}$=3.87, p=.041), but not for the ASC-ASD mean scores ($F_{(4,9)}$=2.66, p=.103).

Univariate analysis indicated that the clusters differed in repetitive behaviours and anxiety for both the WS and ASD group. Parents of children in 'high-atypical' cluster reported significantly more repetitive behaviours, both in total as well as across both sensory motor and insistence on sameness behaviours than parents of children grouped in 'low-atypical' cluster. Also, parents of children in 'high-atypical' cluster indicated a significantly higher level of anxiety in children than parents of children in 'low-atypical' cluster, as shown by total scores of both measures and several subscale scores. The summary of the univariate analysis can be found in Table 7.4.

Table 7.4 Summary of the univariate analysis for the WS and ASD groups

	WS					ASD				
	'low-atypical' cluster	'high-atypical' cluster	F	partial η^2	observed power	'low-atypical' cluster	'high-typical' cluster	F	partial η^2	observed power
RBQ Total score	5.80 (4.24)	22.33 (8.55)	27.24***	.579	.961	17.45 (7.27)	42.00 (23.76)	10.34**	.390	.720
RBQ Sensory/Motor	2.20 (2.90)	8.83 (2.48)	21.70***	.493	.880	6.91 (3.56)	13.25 (7.37)	5.29*	.272	.496
RBQ Insistence on Sameness	2.40 (1.84)	9.17 (5.49)	13.26**	.497	.884	8.27 (3.17)	19.25 (9.22)	12.95**	.409	.753
SCAS-P/PAS Total score	42.80 (9.60)	58.33 (11.67)	8.38*	.284	.518	48.36 (11.53)	76.75 (11.35)	17.91**	.499	.887
SCAS-P/PAS Separation	49.50 (11.07)	60.00 (10.26)	3.55	.174	.309	54.55 (9.02)	74.75 (16.58)	9.51**	.328	.604
SCAS-P/PAS Physical injury fears	52.50 (9.17)	59.67 (13.98)	1.55	.297	.543	56.18 (12.17)	66.75 (10.56)	2.35	.163	.291
SCAS-P/PAS Social Anxiety	43.70 (6.26)	46.17 (7.22)	.52	.011	.063	46.64 (8.62)	71.50 (19.98)	12.15**	.262	.475

SCAS-P/PAS OCD	42.80 (6.20)	56.33 (19.24)	4.38	.081	.157	48.36 (11.92)	83.25 (19.45)	18.16**	.481	.864
SCAS-P/PAS GAD	51.50 (12.78)	68.83 (20.72)	4.36	.064	.133	48.18 (10.97)	79.50 (12.97)	21.89***	.548	.938
ASC_ASD Total score	9.70 (4.24)	21.25 (16.09)	4.87*	.289	.528	17.27 (12.95)	46.00 (20.08)	9.40*	.439	.803
ASC-ASD Performance anxiety	1.50 (2.17)	0.50 (.577)	.79	.062	.130	2.73 (4.74)	9.67 (8.39)	3.73	.237	.428
ASC-ASD Anxious arousal	1.10 (.74)	4.25 (4.72)	4.75	.283	.517	2.09 (3.42)	7.00 (5.20)	3.99	.249	.451
ASC-ASD Separation anxiety	2.50 (2.37)	6.25 (2.63)	6.77*	.261	.666	4.00 (2.83)	8.67 (5.69)	4.26	.262	.475
ASC-ASD Uncertainty	4.60 (2.41)	10.25 (9.22)	3.56	.229	.412	8.45 (5.92)	20.67 (4.16)	10.95**	.477	.859

Note: **p<.01, *p<.05

In the ASD group, when the SRS-2 total score was controlled for in the MANOVA comparing clusters on RRB scores, the effect of cluster on repetitive behaviours was not significant $F_{(3,10)}$=.96, p=.450. Similarly, when the SRS-2 total score was controlled for in the MANOVA comparing clusters on ASC-ASD and SCAS/PAS scores, the effect of cluster on anxiety was not significant ($F_{(4,8)}$=.63, p=.65, $F_{(6,7)}$=2.22, p=.160 respectively).

This suggests that differences in the severity of autistic traits may contribute to both the higher presentation of repetitive behaviours and to the higher presentation of anxiety in those children with ASD who have greater sensory difficulties.

7.3.3 Sensory processing modalities by diagnostic group

In the next step of the analysis, the two neurodevelopmental groups were examined separately, examining sensory modalities. Two, three and four possible sensory processing modalities clusters were identified using hierarchical clustering. In the WS group, all participants kept their cluster membership in the two-cluster solution, 87.5% remained in the same cluster in the three-cluster solution, and 75% remained in the same cluster in the four-cluster solution, after the partitioning procedure. In the ASD group, 88% of participants kept their original cluster membership in the two-cluster-solution, while only 56% and 82% of all participants remained in their original cluster membership in the three-cluster and four-cluster solution, after the partitioning procedure.

In the two-cluster solution for the WS group, the following clusters emerged: (1) a cluster with a low frequency of all sensory behaviours, (n=12; 'low-atypical' cluster); (2) a cluster with a higher frequency of all sensory behaviours (n=4; 'high-atypical' cluster). Figure 7.3 presents the SP sensory processing modalities z-score centers for each cluster for the two-cluster solution.

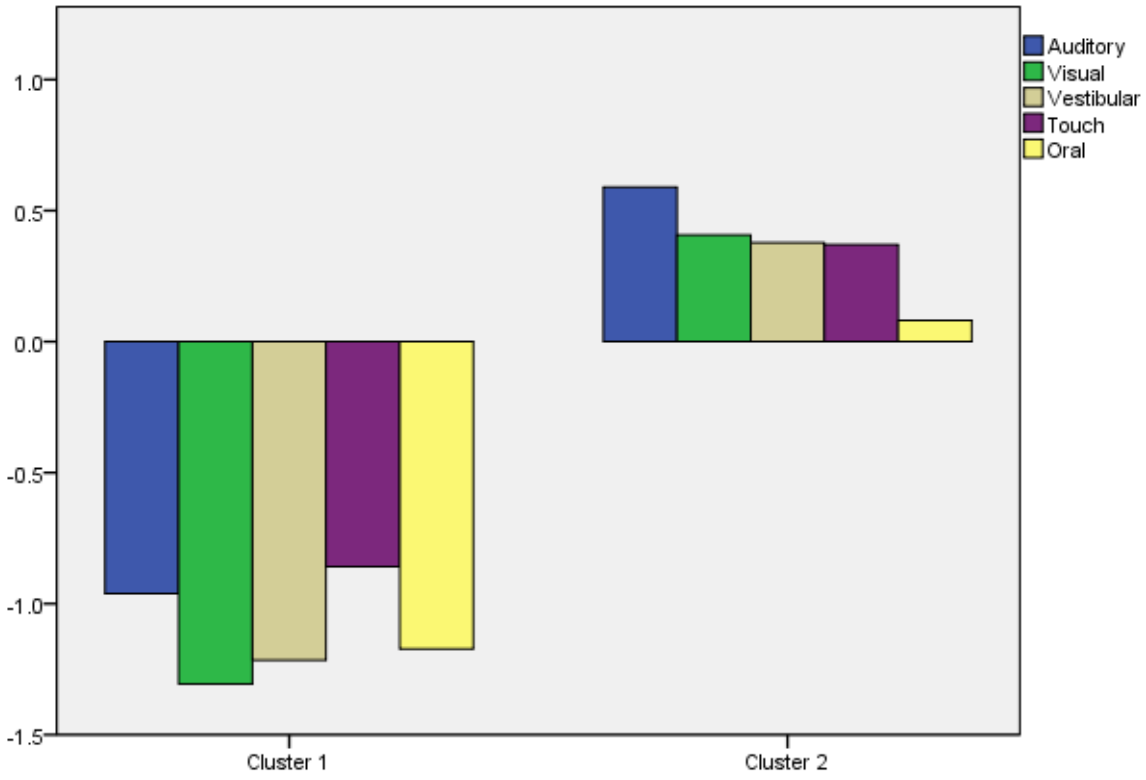


Figure 7.3 SP sensory processing modalities z-scores by cluster in a two-cluster solution for the WS group

In the two-cluster solution for the ASD group, similar clusters emerged: (1) a cluster with a higher frequency of all sensory behaviours (n=8; 'high-atypical' cluster); (2) a cluster with a low frequency of all sensory behaviours (n=9; 'low-atypical' cluster). Figure 7.4 presents the SP sensory processing patterns z-score centered for each cluster for the two-cluster solution.

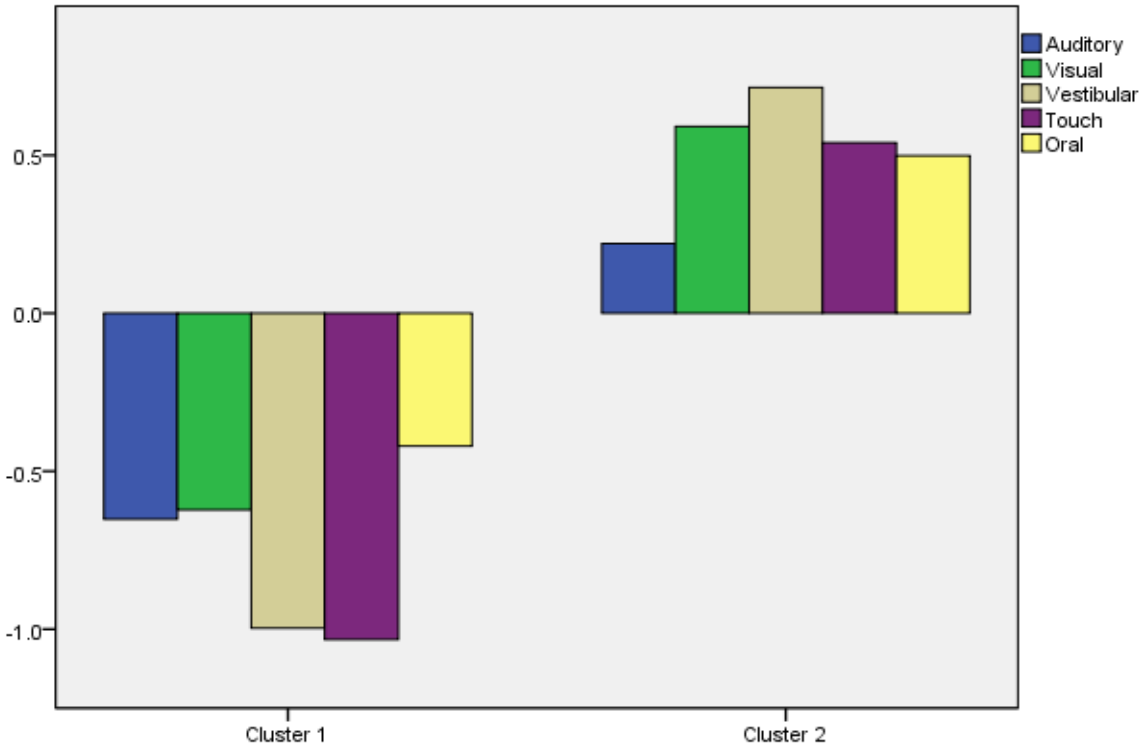


Figure 7.4 SP sensory processing modalities z-scores by cluster in a two-cluster solution for the ASD group

Due to uneven cluster sizes, the bootstrapping procedure was not performed and the standard outcomes have been reported for the WS group. There was a trend suggesting age differences in the WS clusters with younger children grouping in the 'high-atypical' cluster and older children in the 'low-atypical' cluster ($F_{(1,14)}$=4.32, p=.056), The clusters did not differ on gender distribution ($F_{(1,14)}$=1.27, p=.278) or verbal IQ ($F_{(1,6)}$=.04, p=.859). However, the SRS-2 total score differed between the clusters ($F_{(1,14)}$=15.41, p=.002) with participants in 'low-atypical' cluster having significantly lower SRS-2 total score (m=69.67, sd=16.76) than participants in 'high-atypical' cluster (m=111.00, sd=22.85). The difference in nonverbal IQ could not be computed by SPSS due to small sample size.

The ASD clusters did not differ on age ($F_{(1,8)}$=.17, p=.690), gender distribution ($F_{(1,8)}$=1.60, p=.242), verbal IQ ($F_{(1,8)}$=2.93, p=.125), non-verbal IQ ($F_{(1,8)}$=.25, p=.629), but differed in their SRS-2 total score ($F_{(1,8)}$=14.96, p=.005). The SRS-2 total score was

significantly lower in ‘low-atypical’ cluster (m=87.78, sd=16.58) comparing to ‘high-atypical’ cluster (m=141.14, sd=31.54).

MANOVA analysis of the non-standardized sensory processing modalities scores showed significant differences between the clusters for both the WS and ASD groups ($F_{(5,10)}$=8.91, p=.002 and $F_{(5,11)}$=14.09, p<.001 respectively). Results remained significant when the SRS-2 total score was entered into the analysis as a covariate only for the ASD group $F_{(5,9)}$=6.26, p=.009, but not for the WS group $F_{(5,9)}$=2.88, p=.080. The summary of the findings can be found in Table 7.5.

Table 7.5 Summary of the MANOVA analysis of the non-standardized sensory processing modalities scores for WS and ASD groups

	WS				ASD			
Sensory processing modality	'low-atypical' cluster n=12	'high-typical' cluster n=4	F	F (SRS-2 as covariate)	'low-atypical' cluster n=9	'high-typical' cluster n=8	F	F (SRS-2 as covariate)
Auditory	24.75	16.00	21.69***	10.80**	18.50	22.00	1.42	.18
	(2.80)	(4.55)			(4.38)	(7.19)		
Typical performance	1	0			0	2		
Probable difference	3	0			0	1		
Definite difference	8	4			8	6		
Visual	33.33	23.00	23.04***	8.48*	28.25	33.44	3.21	.16
	(3.42)	(4.69)			(4.74)	(6.86)		
Typical performance	9	0			2	5		
Probable difference	3	1			4	2		
Definite difference	0	3			2	2		

Vestibular	44.83	33.00	23.00***	4.60	33.63	48.22	71.46***	28.82***
	(4.43)	(3.65)			(4.81)	(1.86)		
Typical performance	4	0			0	6		
Probable difference	2	0			0	3		
Definite difference	6	4			8	0		
Touch	69.25	52.50	17.82**	1.78	51.88	70.00	7.59*	.24
	(6.50)	(8.10)			(14.45)	(12.70)		
Typical performance	5	0			0	5		
Probable difference	3	0			1	1		
Definite difference	4	4			7	3		
Oral	40.25	24.00	8.73*	.63	33.62	45.78	4.07	5.43*
	(10.57)	(3.74)			(12.46)	(12.35)		
Typical performance	2	0			1	6		
Probable difference	4	0			3	1		
Definite difference	6	4			4	2		

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

The WS group MANOVA showed a significant cluster effect for the three RRB scores ($F_{(3,12)}$=6.30, p=.008), but a non-significant cluster effect for the six SCAS/PAS scores ($F_{(6,9)}$=2.12, p=.150) and five ASC-ASD scores ($F_{(4,9)}$=2.13, p=.159). Univariate analysis indicated that the clusters differed in repetitive behaviours and anxiety. Parents of children in 'high-atypical' cluster reported significantly more repetitive behaviours, both in total as well as across both sensory motor and insistence on sameness behaviours than parents of children grouped in the 'low-atypical' cluster. Also, parents of children in the 'high-atypical' cluster indicated significantly higher levels of anxiety in children than parents of children in the 'low-atypical' cluster, as shown by total scores of both measures and several subscale scores. The summary of the univariate analysis can be found in Table 7.6. However, when the SRS-2 total score was controlled for in the MANOVA comparing clusters on RRB scores, the effect of cluster on repetitive behaviours was not significant $F_{(3,11)}$=1.07, p=.400. Similarly, when the SRS-2 total score was controlled for in the MANOVA comparing clusters on ASC-ASD and SCAS/PAS scores, the effect of cluster on anxiety was not significant $F_{(4,8)}$=.28, p=.886 and $F_{(6,8)}$=1.55, p=.276 respectively.

For the ASD group the MANOVA showed a non-significant cluster effect for the three RRB scores ($F_{(3,13)}$=2.51, p=.105), six SCAS/PAS scores ($F_{(6,9)}$=1.36, p=.326) and five ASC-ASD scores ($F_{(4,10)}$=2.64, p=.097). Univariate analysis indicated that the clusters differed in repetitive behaviours and anxiety. Parents of children in 'high-atypical' cluster reported significantly more repetitive behaviours, both in total as well as across both the sensory motor and insistence on sameness behaviours than parents of children grouped in the 'low-atypical' cluster. Also, parents of children in the 'high-atypical' cluster indicated significantly higher level of anxiety in children than parents of children in the 'low-atypical' cluster, as shown by the total scores of both measures and several subscale scores. The summary of the univariate analysis can be found in Table 7.6. However, when the SRS-2 total score was controlled for in the MANOVA comparing clusters on RRB scores, the effect of cluster on repetitive behaviours was not significant $F_{(3,11)}$=.14, p=.931. Similarly, when the SRS-2 total score was controlled for in the MANOVA comparing clusters on ASC-ASD and SCAS/PAS scores, the effect of cluster on anxiety was not significant $F_{(4,9)}$=.12, p=.973 and $F_{(6,8)}$=.51, p=.789 respectively.

Table 7.6 Summary of the univariate analysis for the WS and ASD groups

	WS					ASD				
	'low-atypical' cluster	'high-atypical' cluster	F	partial η^2	observed power	'low-atypical' cluster	'high-atypical' cluster	F	partial η^2	observed power
RBQ Total score	7.75 (5.96)	24.75 (9.91)	17.72**	.387	.714	15.00 (6.95)	35.25 (19.51)	8.54*	.272	.533
RBQ Sensory/Motor	2.92 (3.12)	10.00 (2.16)	17.43**	.356	.657	6.00 (3.54)	12.50 (5.86)	7.89*	.272	.532
RBQ Insistence on Sameness	3.33 (2.87)	9.75 (6.80)	7.54*	.232	.418	7.44 (3.43)	15.38 (8.25)	7.01*	.220	.425
SCAS-P/PAS Total score	43.83 (9.10)	58.33 (11.67)	11.75**	.354	.653	46.67 (14.12)	64.14 (17.85)	4.80*	.194	.375
SCAS-P/PAS Separation	50.17 (10.50)	63.25 (10.24)	4.70*	.252	.456	74.75 (16.58)	67.14 (15.55)	5.25*	.213	.411
SCAS-P/PAS Physical injury fears	54.00 (9.42)	58.75 (17.15)	.51	.266	.483	52.56 (9.90)	65.29 (12.31)	5.44*	.280	.547
SCAS-P/PAS Social Anxiety	43.08 (5.84)	49.25 (6.99)	3.06	.179	.320	47.56 (9.28)	58.71 (21.37)	2.00	.103	.206

SCAS-P/PAS OCD	43.17 (6.06)	62.00 (21.73)	8.19*	.093	.176	50.00 (12.70)	65.00 (26.60)	2.24	.069	.149
.149SCAS-P/PAS GAD	51.50 (11.58)	77.50 (20.34)	10.45**	.154	.274	49.67 (11.67)	63.00 (22.55)	2.37	.084	.174
ASC_ASD Total score	10.33 (4.38)	29.00 (22.63)	9.92**	.453	.824	15.78 (15.15)	31.50 (20.46)	2.94*	.185	.356
ASC-ASD Performance anxiety	1.33 (2.02)	0.50 (.71)	.32	.026	.081	3.22 (5.14)	5.00 (7.38)	.31	.023	.081
ASC-ASD Anxious arousal	1.33 (1.07)	6.00 (7.07)	7.15*	.373	.690	2.00 (3.87)	4.33 (4.41)	1.17	.083	.171
ASC-ASD Separation anxiety	2.83 (2.29)	8.00 (2.83)	8.36*	.411	.756	3.44 (3.00)	6.67 (4.50)	2.80	.177	.341
ASC-ASD Uncertainty	4.83 (2.29)	14.50 (13.44)	8.07*	.402	.742	7.11 (6.27)	15.50 (7.01)	5.88*	.311	.611

Note: $^{**}p<.01$, $^{*}p<.05$

This suggests that differences in severity of autistic traits contribute to the higher presentation of repetitive behaviours and to the higher presentation of anxiety in those children with ASD and WS who have greater sensory difficulties across different sensory modalities.

7.3.4 Correlational analysis

Correlations between sensory processing, anxiety, intolerance of uncertainty and repetitive behaviours variables were calculated. Neither gender nor age was significantly correlated with any of the sensory processing variable in any of the groups, however gender was significantly different between the ASD and WS groups.

For the WS group, significant negative relationships were found between the RBQ total score and all sensory processing patterns, and vestibular and touch sensory modalities. The RBQ Sensory/Motor subscale was associated with Registration and vestibular and touch sensory modalities, while the Insistence on sameness subscale was negatively correlated with Registration, Sensitivity and Avoiding.

The SCAS/PAS total score in the WS sample was associated with Registration and Sensitivity and with vestibular modality. Also a significant negative association was found between the SCAS/PAS Separation subscale and Sensitivity. The ASC-ASD total score was negatively correlated with Seeking, Avoiding, Auditory and Touch processing, Anxious Arousal subscale correlated with Seeking, Avoiding and Visual processing, Separation subscale was associated with Sensitivity and Touch processing, while Uncertainty was correlated with Seeking and Touch processing.

The correlations are presented in Tables 7.7-7.12.

Table 7.7 Correlations between sensory processing patterns and repetitive behaviours variables for the WS sample after bootstrapping procedure

	Registration	Seeking	Sensitivity	Avoiding
RBQ Total	**-.821**	**-.729**	**-.718**	**-.792**
p	**<.001**	**.001**	**.002**	**<.001**
RBQ sensory/Motor	**-.806**	-.619	-.663	-.634
p	**<.001**	.011	.005	.008
RBQ Insistence on Sameness	**-.708**	-.567	**-.697**	**-.736**
p	**.002**	.022	**.003**	**.001**

Note: significant correlations after Bonferroni correction are in bold

Table 7.8 Correlations between sensory modalities and repetitive behaviours variables for the WS sample after bootstrapping procedure

	Auditory	Visual	Vestibular	Touch	Oral
RBQ Total	-.556	-.564	**-.804**	**-.758**	-.584
p	.025	.023	**<.001**	**.001**	.018
RBQ sensory/Motor	-.478	-.442	**.772**	**-.671**	-.475
p	.061	.087	**<.001**	**.004**	.063
RBQ Insistence on Sameness	-.406	-.474	-.653	-.629	-.575
p	.118	.063	.006	.009	.020

Note: significant correlations after Bonferroni correction are in bold

Table 7.9 Correlations between sensory processing patterns and SCAS/PAS variables for the WS sample after bootstrapping procedure

	Registration	Seeking	Sensitivity	Avoiding
SCAS/PAS Total	**-.735**	-.522	**-.805**	-.684
p	**.001**	.038	**<.001**	.003
SCAS/PAS Separation	-.506	-.261	**-.717**	-.302
p	.046	.328	**.002**	.256
SCAS/PAS Physical Injury	-.538	-.501	-.571	-.506
p	.032	.048	.021	.046
SCAS/PAS Social Anxiety	-.044	-.217	-.371	-.176
p	.872	.419	.157	.514
SCAS/PAS OCD	-.510	-.275	-.516	-.425
p	.044	.302	.041	.101
SCAS/PAS GAD	-.669	-.390	-.641	-.623
p	.005	.136	.008	.010

Note: significant correlations after Bonferroni correction are in bold

Table 7.10 Correlations between sensory modalities and SCAS/PAS variables for the WS sample after bootstrapping procedure

	Auditory	Visual	Vestibular	Touch	Oral
SCAS/PAS Total	-.589	-.476	**-.754**	-.624	-.513
p	.016	.063	**.001**	.010	.042
SCAS/PAS Separation	-.428	-.181	-.558	-.557	-.449
p	.098	.503	.025	.025	.081
SCAS/PAS Physical Injury	-.425	-.191	-.476	-.635	-.476
p	.101	.479	.063	.008	.062
SCAS/PAS Social Anxiety	-.487	-.368	-.335	-.103	-.176
p	.056	.161	.204	.704	.514
SCAS/PAS OCD	-.252	-.366	-.492	-.307	-.268
p	.347	.163	.053	.247	.316
SCAS/PAS GAD	-.422	-.520	-.567	-.440	-.385
p	.104	.039	.022	.088	.141

Note: significant correlations after Bonferroni correction are in bold

Table 7.11 Correlations between sensory processing patterns and ASC-ASD variables for the WS sample after bootstrapping procedure

	Registration	Seeking	Sensitivity	Avoiding
ASC-ASD Total	-.673	**-.787**	-.694	**-.818**
p	.008	**.001**	.006	**<.001**
ASC-ASD Performance	.362	.221	.040	.108
p	.204	.448	.893	.714
ASC-ASD Arousal	-.625	**-.780**	-.559	**-.890**
p	.017	**.001**	.038	**<.001**
ASC-ASD Separation	-.722	-.621	**-.791**	-.690
p	.004	.018	**.001**	.006
ASC-ASD Uncertainty	-.654	**-.789**	-.579	-.715
p	.011	**.001**	.030	.004

Note: significant correlations after Bonferroni correction are in bold

Table 7.12 Correlations between sensory modalities and ASC-ASD variables for the WS sample after bootstrapping procedure

	Auditory	Visual	Vestibular	Touch	Oral
ASC-ASD Total	**-.743**	-.672	-.699	**-.761**	-.577
p	**.002**	.008	.009	**.002**	.031
ASC-ASD Performance	-.174	-.095	.156	.366	.028
p	.552	.748	.595	.198	.925
ASC-ASD Arousal	-.718	**-.732**	-.599	-.705	-.541
p	.004	**.003**	.024	.005	.046
ASC-ASD Separation	-.650	-.401	-.721	**-.807**	-.517
p	.012	.156	.004	**<.001**	.058
ASC-ASD Uncertainty	-.591	-.613	-.590	**-.731**	-.515
p	.026	.020	.026	**.003**	.060

Note: significant correlations after Bonferroni correction are in bold

For the ASD group, significant negative correlations were found between the RBQ total score and Registration, Sensitivity and Avoiding; and vestibular sensory processing. The RBQ Sensory/Motor subscale was associated with Sensitivity, while the Insistence on Sameness subscale was associated with Registration, Sensitivity, Avoiding, and vestibular and touch modalities. The SCAS/PAS total score and the GAD subscale were negatively correlated with touch processing modality. There were no significant associations between any of the sensory variables and ASC-ASD scores. The summary of all association found can be seen in Tables 7.13-7.18.

The findings suggest that greater sensory processing difficulties were associated with more repetitive behaviours and higher anxiety levels in both the WS and ASD samples, with stronger associations between sensory processing, anxiety and repetitive behaviours found in the WS group.

Table 7.13 Correlations between sensory processing patterns and repetitive behaviours variables for the ASD sample after bootstrapping procedure

	Registration	Seeking	Sensitivity	Avoiding
RBQ Total	**-.706**	-.575	**-.738**	**-.713**
p	**.003**	.025	**.002**	**.003**
RBQ sensory/Motor	-.605	-.638	**-.691**	-.628
p	.017	.010	**.004**	.012
RBQ Insistence on Sameness	**-.739**	-.523	**-.722**	**-.745**
p	**.002**	.045	**.002**	**.001**

Note: significant correlations after Bonferroni correction are in bold

Table 7.14 Correlations between sensory modalities and repetitive behaviours variables for the ASD sample after bootstrapping procedure

	Auditory	Visual	Vestibular	Touch	Oral
RBQ Total	-.398	-.549	**-.724**	-.648	-.223
p	.113	.023	**.001**	.005	.389
RBQ sensory/Motor	-.352	-.543	-.653	-.572	-.303
p	.166	.024	.005	.016	.237
RBQ Insistence on Sameness	-.418	-.570	**-.735**	**-.744**	-.129
p	.095	.017	**.001**	**.001**	.621

Note: significant correlations after Bonferroni correction are in bold

Table 7.15 Correlations between sensory processing patterns and SCAS/PAS variables for the ASD sample after bootstrapping procedure

	Registration	Seeking	Sensitivity	Avoiding
SCAS/PAS Total	-.604	-.504	-.665	-.718
p	.017	.055	.007	.003
SCAS/PAS Separation	-.663	-.338	-.385	-.665
p	.007	.217	.157	.007
SCAS/PAS Physical Injury	-.288	-.334	-.490	-.647
p	.298	.224	.064	.009
SCAS/PAS Social Anxiety	-.493	-.166	-450	-.586
p	.062	.554	.092	.022
SCAS/PAS OCD	-.652	-.460	-.640	-.551
p	.008	.084	.010	.033
SCAS/PAS GAD	-.644	-.372	-.599	-.644
p	.010	.172	.018	.010

Table 7.16 Correlations between sensory modalities and SCAS/PAS variables for the ASD sample after bootstrapping procedure

	Auditory	Visual	Vestibular	Touch	Oral
SCAS/PAS Total	-.586	-.467	-.690	**-.758**	-.049
p	.022	.068	.003	**.001**	.857
SCAS/PAS Separation	-.424	-.201	-.639	-.643	.139
p	.101	.455	.008	.007	.607
SCAS/PAS Physical Injury	-.496	-.363	-.515	-.511	-.029
p	.051	.167	.041	.043	.916
SCAS/PAS Social Anxiety	-.401	-.312	-.466	-.513	.035
p	.124	.240	.069	.042	.898
SCAS/PAS OCD	-.435	-.363	-.650	-.694	-.077
p	.092	.167	.006	.003	.778
SCAS/PAS GAD	-.486	-.402	-.617	**-.712**	-.012
p	.056	.123	.011	**.002**	.964

Note: significant correlations after Bonferroni correction are in bold

Table 7.17 Correlations between sensory processing patterns and ASC-ASD variables for the ASD sample after bootstrapping procedure

	Registration	Seeking	Sensitivity	Avoiding
ASC-ASD Total	-.482	-.234	-.560	-.677
p	.081	.421	.037	.008
ASC-ASD Performance	-.314	.025	-.329	-.465
p	.274	.933	.250	.094
ASC-ASD Arousal	-.347	-.252	-.394	-.478
p	.224	.385	.163	.084
ASC-ASD Separation	-.539	-.106	-.331	-.620
p	.047	.717	.247	.018
ASC-ASD Uncertainty	-.458	-.398	-.718	-.699
p	.099	.159	.004	.005

Table 7.18 Correlations between sensory modalities and ASC-ASD variables for the ASD sample after bootstrapping procedure

	Auditory	Visual	Vestibular	Touch	Oral
ASC-ASD Total	-.449	-.505	-.538	-.636	-.015
p	.093	.055	.038	.011	.958
ASC-ASD Performance	-.362	-.393	-.203	-.351	.021
p	.185	.148	.468	.199	.940
ASC-ASD Arousal	-.300	-.413	-.442	-.673	.049
p	.278	.126	.099	.006	.863
ASC-ASD Separation	-.323	-.180	-.524	-.442	.080
p	.240	.522	.045	.099	.777
ASC-ASD Uncertainty	-.484	-.609	-.647	-.687	-.120
p	.068	.016	.009	.005	.670

Similar analysis was then performed with controlling for gender. For the WS group, significant negative relationships were found between the RBQ total score and all sensory processing patterns, and vestibular and touch sensory modalities. The RBQ Sensory/Motor subscale was associated with Registration and vestibular and touch sensory modalities, while the Insistence on sameness subscale was negatively correlated with Registration, Sensitivity and Avoiding.

The SCAS/PAS total score was associated with Registration and Sensitivity and with vestibular modality. Also a significant negative association was found between the SCAS/PAS Separation subscale and Sensitivity. The ASC-ASD total score was negatively correlated with Seeking, Avoiding, Auditory and Touch processing, Anxious Arousal subscale correlated with Seeking, Avoiding and Visual processing, Separation subscale was associated with Sensitivity and Touch processing, while Uncertainty was correlated with Seeking and Touch processing (Tables 7.19-7.24).

Table 7.19 Correlations between sensory processing patterns and repetitive behaviours variables for the WS sample after bootstrapping procedure and controlling for gender

	Registration	Seeking	Sensitivity	Avoiding
RBQ Total	**-.831**	**-.729**	**-.724**	**-.792**
p	**<.001**	**.002**	**.002**	**<.001**
RBQ sensory/Motor	**-.809**	-.622	-.663	-.633
p	**<.001**	.013	.007	.011
RBQ Insistence on Sameness	**-.740**	-.567	**-.722**	**-.745**
p	**.002**	.028	**.002**	**.001**

Note: significant correlations after Bonferroni correction are in bold

Table 7.20 Correlations between sensory modalities and repetitive behaviours variables for the WS sample after bootstrapping procedure and controlling for gender

	Auditory	Visual	Vestibular	Touch	Oral
RBQ Total	-.574	-.564	**-.822**	**-.758**	-.589
p	.025	.029	**.001**	**.001**	.021
RBQ sensory/Motor	-.482	-.44	**-.781**	**-.671**	-.473
p	.069	.098	**.001**	**.006**	.075
RBQ Insistence on Sameness	-.453	-.475	-.700	-.636	-.600
p	.090	.073	.004	.011	.018

Note: significant correlations after Bonferroni correction are in bold

Table 7.21 Correlations between sensory processing patterns and SCAS/PAS variables for the WS sample after bootstrapping procedure and controlling for gender

	Registration	Seeking	Sensitivity	Avoiding
SCAS/PAS Total	**-.739**	-.525	**-.807**	-.684
p	**.002**	.045	**<.001**	.005
SCAS/PAS Separation	-.491	-.301	**-.737**	-.318
p	.014	.276	**.002**	.247
SCAS/PAS Physical Injury	-.620	-.515	-.641	-.539
p	.014	.049	.010	.038
SCAS/PAS Social Anxiety	-.014	-.229	-.357	-.175
p	.960	.412	.191	.534
SCAS/PAS OCD	-.498	-.284	-.507	-.426
p	.059	.304	.054	.113
SCAS/PAS GAD	-.685	-.389	-.651	-.625
p	.005	.152	.009	.013

Note: significant correlations after Bonferroni correction are in bold

Table 7.22 Correlations between sensory modalities and SCAS/PAS variables for the WS sample after bootstrapping procedure and controlling for gender

	Auditory	Visual	Vestibular	Touch	Oral
SCAS/PAS Total	-.597	-.477	**-.764**	-.624	-.513
p	.019	.072	**.001**	.013	.051
SCAS/PAS Separation	-.371	-.207	-.528	-.600	-.439
p	.173	.459	.043	.018	.102
SCAS/PAS Physical Injury	-.540	-.193	-.579	-.673	-.544
p	.038	.490	.024	.006	.036
SCAS/PAS Social Anxiety	-.461	-.380	-.306	-.101	-.156
p	.084	.163	.267	.720	.580
SCAS/PAS OCD	-.223	-.375	-.475	-.308	-.253
p	.425	.169	.074	.264	.363
SCAS/PAS GAD	-.447	-.520	-.591	-.441	-.394
p	.095	.047	.020	.100	.146

Note: significant correlations after Bonferroni correction are in bold

Table 7.23 Correlations between sensory processing patterns and ASC-ASD variables for the WS sample after bootstrapping procedure and controlling for gender

	Registration	Seeking	Sensitivity	Avoiding
ASC-ASD Total	-.707	**-.799**	-.708	**-.821**
p	.007	**.001**	.007	**.001**
ASC-ASD Performance	.362	.216	.035	.100
p	.224	.478	.909	.744
ASC-ASD Arousal	-.647	**-.786**	-.561	**-.892**
p	.017	**.001**	.046	**<.001**
ASC-ASD Separation	-.724	-.618	**-.790**	-.689
p	.005	.024	**.001**	.009
ASC-ASD Uncertainty	-.716	**-.824**	-.603	-.725
p	.006	**.001**	.029	.005

Note: significant correlations after Bonferroni correction are in bold

Table 7.24 Correlations between sensory modalities and ASC-ASD variables for the WS sample after bootstrapping procedure and controlling for gender

	Auditory	Visual	Vestibular	Touch	Oral
ASC-ASD Total	**-.839**	-.637	-.739	**-.776**	-.607
p	**<.001**	.019	.004	**.002**	.028
ASC-ASD Performance	-.168	-.118	.162	.363	.028
p	.584	.701	.597	.223	.928
ASC-ASD Arousal	**-.795**	**-.709**	-.651	-.710	-.561
p	**.001**	**.007**	.016	.006	.046
ASC-ASD Separation	-.669	-.399	-.734	**-.806**	-.519
p	.012	.177	.004	**.001**	.069
ASC-ASD Uncertainty	-.722	-.560	-.694	**-.766**	-.565
p	.005	.046	.008	**.002**	.044

Note: significant correlations after Bonferroni correction are in bold

For the ASD group, significant negative correlations were found between the RBQ total score and Sensitivity and Avoiding; and vestibular sensory processing. The RBQ Sensory/Motor subscale was associated with Sensitivity, while the Insistence on Sameness subscale was associated with Registration, Sensitivity, Avoiding, and vestibular and touch modalities. The SCAS/PAS total score was negatively correlated with touch processing

modality. There were no significant associations between any of the sensory variables and ASC-ASD scores. The summary of all association found can be seen in Tables 7.25-7.30.

Table 7.25 Correlations between sensory processing patterns and repetitive behaviours variables for the ASD sample after bootstrapping procedure and controlling for gender

	Registration	Seeking	Sensitivity	Avoiding
RBQ Total	-.702	-.568	**-.758**	**-.715**
p	.005	.034	**.002**	**.004**
RBQ sensory/Motor	-.610	-.647	**-.751**	-.657
p	.020	.012	**.002**	.011
RBQ Insistence on Sameness	**-.737**	-.518	**-.749**	**-.754**
p	**.003**	.058	**.002**	**.002**

Note: significant correlations after Bonferroni correction are in bold

Table 7.26 Correlations between sensory modalities and repetitive behaviours variables for the ASD sample after bootstrapping procedure and controlling for gender

	Auditory	Visual	Vestibular	Touch	Oral
RBQ Total	-.393	-.545	**-.719**	-.646	-.193
p	.133	.029	**.002**	.007	.473
RBQ sensory/Motor	-.381	-.543	-.655	-.571	-.321
p	.146	.030	.006	.021	.226
RBQ Insistence on Sameness	-.433	-.568	**-.733**	**-.742**	-.106
p	.094	.022	**.001**	**.001**	.695

Note: significant correlations after Bonferroni correction are in bold

Table 7.27 Correlations between sensory processing patterns and SCAS/PAS variables for the ASD sample after bootstrapping procedure and controlling for gender

	Registration	Seeking	Sensitivity	Avoiding
SCAS/PAS Total	-.599	-.487	-.629	-.694
p	.024	.078	.016	.006
SCAS/PAS Separation	-.657	-.318	-.339	-.647
p	.011	.268	.236	.012
SCAS/PAS Physical Injury	-.265	-.302	-.404	-.608
p	.360	.294	.152	.021
SCAS/PAS Social Anxiety	-.486	-.150	-.439	-.579
p	.078	.608	.116	.030
SCAS/PAS OCD	-.650	-.440	-.599	-.511
p	.012	.115	.024	.062
SCAS/PAS GAD	-.638	-.354	-.581	-.627
p	.014	.214	.029	.016

Table 7.28 Correlations between sensory modalities and SCAS/PAS variables for the ASD sample after bootstrapping procedure and controlling for gender

	Auditory	Visual	Vestibular	Touch	Oral
SCAS/PAS Total	-.543	-.464	-.683	**-.764**	.047
p	.037	.081	.005	**.001**	.869
SCAS/PAS Separation	-.401	-.193	-.632	-.642	.231
p	.138	.490	.012	.010	.408
SCAS/PAS Physical Injury	-.421	-.360	-.504	-.517	.114
p	.118	.187	.056	.048	.687
SCAS/PAS Social Anxiety	-.406	-.307	-.460	-.510	.083
p	.133	.265	.085	.052	.769
SCAS/PAS OCD	-.369	-.359	-.643	-.702	.034
p	.176	.189	.010	.004	.903
SCAS/PAS GAD	-.476	-.398	-.609	-.712	.058
p	.073	.142	.016	.003	.838

Note: significant correlations after Bonferroni correction are in bold

Table 7.29 Correlations between sensory processing patterns and ASC-ASD variables for the ASD sample after bootstrapping procedure and controlling for gender

	Registration	Seeking	Sensitivity	Avoiding
ASC-ASD Total	-.481	-.232	-.589	-.692
p	.096	.446	.034	.009
ASC-ASD Performance	-.317	.020	-.370	-.493
p	.291	.948	.214	.087
ASC-ASD Arousal	-.367	-.281	-.510	-.558
p	.217	.352	.075	.047
ASC-ASD Separation	-.536	-.092	-.304	-.610
p	.059	.764	.312	.027
ASC-ASD Uncertainty	-.455	-.388	-.721	-.692
p	.118	.190	.005	.009

Table 7.30 Correlations between sensory modalities and ASC-ASD variables for the ASD sample after bootstrapping procedure and controlling for gender

	Auditory	Visual	Vestibular	Touch	Oral
ASC-ASD Total	-.511	-.506	-.541	-.636	-.020
p	.062	.065	.046	.014	.945
ASC-ASD Performance	-.444	-.398	-.209	-.351	-.007
p	.112	.159	.473	.219	.980
ASC-ASD Arousal	-.465	-.437	-.472	-.687	-.053
p	.094	.119	.088	.007	.858
ASC-ASD Separation	-.315	-.175	-.521	-.446	.137
p	.273	.549	.056	.110	.641
ASC-ASD Uncertainty	-.497	-.607	-.644	-.692	-.086
p	.071	.021	.013	.006	.771

7.3.5 Mediation analysis

Serial mediation models were tested using PROCESS (Hayes, 2012). This computational tool for mediation, moderation, and mediated moderation models of observed effects runs under SPSS (Statistical Package for Social Science software, version 22.0). The model was based on previous computational work (Wigham et al., 2015) and similar direct

paths from both sensory hyporesponsiveness and sensory hyperresponsiveness to both repetitive sensory/motor behaviours and insistence on sameness and an indirect path through intolerance of uncertainty (IoU) and anxiety were tested (see Figure 7.5 in which direct paths are marked as black lines and indirect paths as blue lines). Sensory hyporesponsiveness and sensory hyperresponsiveness were calculated as sums of items marked as either low or high neurological threshold as indicated by the Sensory Profile (Dunn, 1999). As a measure of intolerance of uncertainty, a subscale of ASC-ASD was used. T-scores from the SCAS-P/PAS were entered as an anxiety measure.

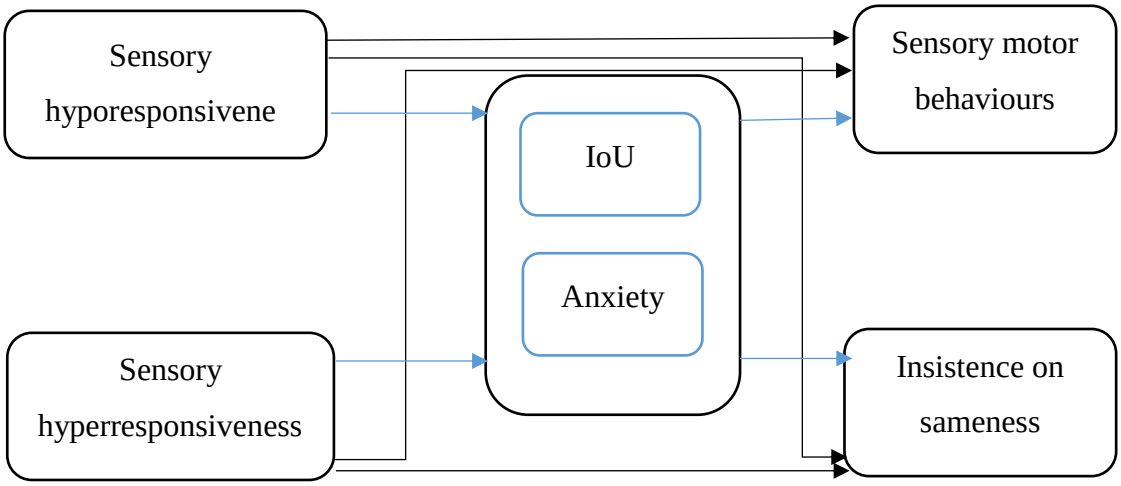


Figure 7.5 Hypothesised model of the relationship between sensory processing atypicalities, intolerance of uncertainty, anxiety and repetitive behaviours

Total, direct and indirect effects were calculated. As Kenny (2016) postulates, total effect refers to a path (c) between a causal variable (X) and an outcome variable (Y). When the effect of X on Y is mediated by a mediated variable (M), direct effect of X on Y can be calculated after controlling for M (path c'). Complete mediation takes place when variable X no longer affects Y after M has been controlled, making path c' nonsignificant and closer to zero. Partial mediation takes place when the size and significance level of the path from X to Y are reduced when the mediator is introduced.

In the serial mediation models, PROCESS estimates the total indirect effect as well as single (via intolerance of uncertainty and via anxiety) and double mediator path (via intolerance of uncertainty and anxiety). Due to small sample size, bootstrapping technique with 1000 resamples and generated accelerated 95 % confidence was implemented to adjust for measurement error when interpreting indirect effects (Shrout & Bolger, 2002). Non-significant paths in the models are indicated by confidence intervals of zero; effect sizes are

indicated by R^2 values. Pearson's correlations between age, gender, verbal IQ and non-verbal IQ and covariates were calculated. Age was entered as covariate for WS sample as it correlated with intolerance of uncertainty (r=-.642, p=.007), sensory/motor repetitive behaviours (r=-.591, p=.016) and insistence on sameness (r=-.520, p=.039). None of the remaining correlations were significant. Inter correlations between the covariates for both the WS and ASD samples are presented in Table 7.31 and Table 7.32.

Table 7.31 Inter correlations between the covariates for the WS sample

	Sensory under responsiveness	Sensory over responsiveness	Intolerance of uncertainty	Anxiety	RBQ SM
Sensory over responsiveness	.866**				
Intolerance of uncertainty	-.775**	-.778**			
Anxiety	-.845**	-.683**	.792**		
RBQ SM	-.706**	-.751**	.797**	.786**	
RBQ Sameness	-.757**	-.723**	.888**	.785**	.800**

Note: ** correlation is significant at the .01 level

Table 7.32 Inter correlations between the covariates for the ASD sample

	Sensory under responsiveness	Sensory over responsiveness	Intolerance of uncertainty	Anxiety	RBQ SM
Sensory over responsiveness	.791**				
Intolerance of uncertainty	-.780**	-.561*			
Anxiety	-.685**	-.629*	.826**		
RBQ SM	-.713**	-.739**	.465	.577*	
RBQ Sameness	-.776**	-.748**	.536*	.663**	.866**

Note: ** correlation is significant at the .01 level, * correlation is significant at the .05 level

Based on the findings of Wigham et al. (2015) it was predicted that for the ASD sample, direct paths will emerge from sensory hyporesponsiveness to both sensory/motor behaviours and insistence on sameness, and from sensory hyperresponsiveness to insistence on sameness behaviours. Also, it was expected that indirect paths involving both intolerance

of uncertainty and anxiety in all cases will occur. The significant total and direct effects were found in all cases: from sensory hyporesponsiveness to sensory/motor behaviours (β=-.12, *p*=.003 and β=-.14, *p*=.018 respectively) and insistence on sameness behaviours (β=-.17, *p*<001 and β=-.19, *p*=.007); and from sensory hyperresponsiveness to sensory/motor behaviours (β=-.13, *p*=.002 and β=-.11, *p*=.031) as well as to insistence on sameness (β=-.17, *p*=.001 and β=-.13, *p*=.040). Reduced significance level from total to direct effects suggests that all the paths are partially mediated by at least one of the mediation variables (intolerance of uncertainty or anxiety). Significant indirect effects through anxiety (β=-.09, LL=-.21, UL=-.00) and intolerance of uncertainty and anxiety (β=.19, LL=.02, UL=.46) from sensory hyporesponsiveness to insistence on sameness behaviours were found.

None of the paths, either direct or indirect, were significant for the 'low-atypical' cluster. In the 'high-atypical' cluster significant total and nonsignificant direct effects were found from sensory hyporesponsiveness and insistence on sameness (β=-.22, *p*=.03 and β=-.19, *p*=.12 respectively) and sensory hyperresponsiveness and insistence on sameness behaviours (β=-.31, *p*=.04 and β=-.24, *p*=.13 respectively). In addition, significant indirect effects from sensory hyperresponsiveness and insistence on sameness through intolerance on uncertainty was found (β=-.06, LL=.00, UL=.46) suggesting that intolerance of uncertainty fully mediates that path.

In the WS sample, the significant total and non-significant direct effects were found in all cases: from sensory hyporesponsiveness to sensory/motor behaviours (β=-.09, *p*<.001 and β=-.01, *p*=.80 respectively) and insistence on sameness behaviours (β=-.11, *p*<.001 and β=-.01, *p*=.81); and from sensory hyperresponsiveness to sensory/motor behaviours (β=-.07, *p*=.01 and β=-.04, *p*=.24) as well as to insistence on sameness (β=-.09, *p*=.01 and β=-.01, *p*=.84). Diminished to non-significance level paths from total to direct effects suggest that all the paths are completely mediated by at least one of the mediation variables (intolerance of uncertainty or anxiety) in the WS sample. Significant indirect total effects and indirect effect through intolerance of uncertainty were found from sensory hyporesponsiveness to insistence on sameness behaviours (β=-.10, LL=-.28, UL=-.04 and β=-.08, LL=-.22, UL=-.01 respectively) and from sensory hyperresponsiveness to insistence on sameness behaviours (β=-.08, LL=-.18, UL=-.03 and β=-.06, LL=-.18, UL=-.02).

In the 'low-atypical' cluster in the WS sample significant direct effect was found only for sensory hyperresponsiveness to insistence on sameness path (β=-.12, *p*=.04). In 'high-atypical' cluster significant total effect and nonsignificant direct effect was found from

sensory hyporesponsiveness to sensory/motor behaviours (β=-.11, p=.03 and (β=-.12, p=.30). None of the other paths were found to be significant.

Table 7.33 Mediation analysis summary for the WS group with age entered as covariates

Variables		Total, direct and indirect effects																									
Dependent	Predictor	Total effect						Direct effect					Total indirect effects				Indirect effect IU				Indirect effect Anx				Indirect effect IU → Anx			
		R^2	B	se	LL	UL	p	B	se	LL	UL	p	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL
Sensory under responsiveness	SM	.65	-.09	.03	-.14	-.03	<.001	-.01	.05	-.12	.09	.80	-.08	.05	-.18	.02	-.01	.04	-.11	.05	-.06	.05	-.15	.03	.05	.07	-.09	.17
Sensory under responsiveness	Sameness	.67	-.11	.03	-.18	-.05	<.001	-.01	.05	-.11	.09	.81	-.10*	.05	-.28	-.04	-.08*	.05	-.22	-.01	-.02	.05	-.15	.06	-.05	.08	-.25	.08
Sensory over responsiveness	SM	.65	-.07	.02	-.12	-.03	.01	-.04	.03	-.10	.03	.24	-.04	.03	-.10	.01	.00	.04	-.06	.05	-.04	.03	-.11	.01	.04	.06	-.06	.15
Sensory over responsiveness	Sameness	.57	-.09	.03	-.15	-.02	.01	-.01	.03	-.07	.06	.84	-.08*	.03	-.18	-.03	-.06*	.04	-.18	-.02	-.02	.03	-.08	.03	-.04	.05	-.18	.04

Table 7.34 Mediation analysis summary for the ASD group

Variables		Total, direct and indirect effects																									
Dependent	Predictor	Total effect						Direct effect					Total indirect effects				Indirect effect IU				Indirect effect Anx				Indirect effect IU → Anx			
		R^2	B	se	LL	UL	p	B	se	LL	UL	p	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL
Sensory under responsiveness	SM	.51	-.12	.03	-.19	-.05	.003	-.14	.05	-.25	-.03	.018	.02	.06	-.08	.13	.08	.07	-.05	.19	-.06	.04	-.13	.01	.14	.09	-.03	.28
Sensory under responsiveness	Sameness	.60	-.17	.04	-.25	-.09	<.001	-.19	.06	-.31	-.06	.007	.02	.05	-.06	.18	.11	.07	-.00	.29	-.09*	.05	-.21	-.00	.19*	.12	.02	.46
Sensory over responsiveness	SM	.55	-.13	.03	-.20	-.06	.002	-.11	.04	-.21	-.01	.031	-.02	.04	-.11	.06	.01	.05	-.05	.17	-.03	.04	-.12	.03	.04	.08	-.08	.27
Sensory over responsiveness	Sameness	.56	-.17	.04	-.26	-.08	.001	-.13	.06	-.25	-.01	.040	-.04	.05	-.13	.03	.01	.08	-.06	.24	-.06	.06	-.20	.05	.07	.14	-.14	.40

Table 7.35 Mediation analysis summary for the WS 'low-atypical' cluster with age entered as covariate

Variables		Total, direct and indirect effects																									
Dependent	Predictor	Total effect						Direct effect					Total indirect effects				Indirect effect IU				Indirect effect Anx				Indirect effect IU → Anx			
		R^2	B	se	LL	UL	p	B	se	LL	UL	p	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL
Sensory under responsiveness	SM	.28	.02	.12	-.28	.32	.86	.06	.07	-.16	.27	.45	-.04	.43	-.23	.21	-.01	.41	-.17	.10	-.02	.06	-.17	.07	.01	.39	-.13	.23
Sensory under responsiveness	Sameness	.56	-.10	.05	-.21	.02	.09	-.05	.04	-.19	.09	.33	-.04	.16	-.21	.06	-.04	.15	-.15	.19	.00	.04	-.13	.04	-.04	.15	-.16	.33
Sensory over responsiveness	SM	.34	.09	.14	-.26	.44	.53	.02	.11	-.33	.36	.89	.08	.13	-.13	.44	.01	.08	-.14	.20	.06	.10	-.03	.38	-.05	.12	-.34	.12
Sensory over responsiveness	Sameness	.45	-.10	.06	-.26	.06	.17	-.12	.04	-.23	.00	.04	.02	.10	-.38	.15	-.01	.06	-.31	.03	.02	.07	-.14	.19	-.03	.08	-.11	.28

Table 7.36 Mediation analysis summary for the WS 'high-atypical' cluster with age entered as covariate

Variables		Total, direct and indirect effects																									
Dependent	Predictor	Total effect						Direct effect					Total indirect effects				Indirect effect IU				Indirect effect Anx				Indirect effect IU → Anx			
		R^2	B	se	LL	UL	p	B	se	LL	UL	p	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL
Sensory under responsiveness	SM	.81	-.11	.04	-.20	-.01	.03	-.12	.10	-.42	.18	.30	.01	.11	-.26	.26	.00	.11	-.13	.21	.02	.10	-.14	.27	-.02	.18	-.27	.31
Sensory under responsiveness	Sameness	.48	-.14	.10	-.41	.12	.23	-.14	.10	-.35	.72	.35	-.33*	.21	-.88	-.04	-.26	.20	-.52	.11	-.06	.16	-.25	.32	-.20	.30	-.62	.35
Sensory over responsiveness	SM	.67	-.05	.03	-.12	.03	.16	-.03	.04	-.17	.12	.60	-.02	.10	-.42	.01	-.02	.15	-.49	.07	.00	.07	-.32	.10	-.01	.22	-.70	.09
Sensory over responsiveness	Sameness	.34	-.05	.07	-.22	.13	.50	.04	.07	-.20	.28	.61	-.09	.20	-1.21	.01	-.09	.28	-1.71	.10	.00	.10	-.20	.12	-.08	.38	-2.20	.22

Table 7.37 Mediation analysis summary for the ASD 'low-atypical' cluster

Variables		Total, direct and indirect effects																										
Dependent	Predictor	Total effect						Direct effect					Total indirect effects				Indirect effect IU					Indirect effect Anx				Indirect effect IU → Anx		
		R^2	B	se	LL	UL	p	B	se	LL	UL	p	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL
Sensory under responsiveness	SM	.02	.03	.08	-.17	.23	.74	.09	.10	-.21	.40	.40	-.07	.08	-.21	.14	-.04	.09	-.21	.17	-.02	.08	-.37	.01	-.02	.16	-.20	.47
Sensory under responsiveness	Sameness	.06	.03	.05	-.09	.15	.58	.04	.09	-.26	.35	.68	-.02	.10	-.32	.09	-.02	.14	-.39	.24	.00	.17	-.31	.14	-.02	.31	-.95	.44
Sensory over responsiveness	SM	.10	-.05	.07	-.23	.12	.48	-.02	.08	-.28	.25	.86	-.04	.24	-.19	1.12	.00	.09	-.10	.51	-.03	.20	-.23	.01	.03	.19	-.10	.21
Sensory over responsiveness	Sameness	.17	-.04	.04	-.14	.06	.35	-.06	.07	-.27	.15	.41	.02	.29	-.09	1.45	.01	.12	-.11	.70	.01	.25	-.01	1.37	.00	.26	-.18	.13

Table 7.38 Mediation analysis summary for the ASD 'high-atypical' cluster

Variables		Total, direct and indirect effects																										
Dependent	Predictor	Total effect						Direct effect					Total indirect effects				Indirect effect IU					Indirect effect Anx				Indirect effect IU → Anx		
		R^2	B	se	LL	UL	p	B	se	LL	UL	p	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL	B	se	LL	UL
Sensory under responsiveness	SM	.37	-.12	.06	-.27	.03	.11	-.15	.08	-.36	.06	.12	.04	.05	-.05	.15	.08	.09	-.01	.32	-.05	.09	-.17	.21	.13	.17	-.10	.46
Sensory under responsiveness	Sameness	.57	-.22	.08	-.42	-.03	.03	-.19	.10	-.45	.08	.12	-.04	.09	-.19	.18	.10	.14	-.07	.50	-.14	.13	-.36	.16	.24	.26	-.18	.83
Sensory over responsiveness	SM	.31	-.16	.09	-.39	.08	.15	-.15	.12	-.48	.17	.25	.00	.19	-.10	.70	.06*	.08	.00	.46	-.06	.23	-.42	.05	.13	.28	-.05	.86
Sensory over responsiveness	Sameness	.52	-.31	.12	-.61	-.01	.04	-.24	.13	-.58	.11	.13	-.08	.22	-.19	.71	.07	.10	-.05	.43	-.15	.26	-.43	.53	.22	.33	-.64	.73

7.4 Discussion

In this study, we primarily aimed to investigate the patterns of sensory clusters in children with ASD or WS, examine whether any of the patterns of sensory clusters were syndrome-specific or associated with higher levels of anxiety, intolerance of uncertainty or repetitive behaviours. Two distinct patterns of sensory processing clusters emerged in both the ASD and WS groups, when either sensory processing patterns or sensory processing modalities were entered into the analysis: one cluster with a more typical presentation of sensory processing features and the other one with a more abnormal presentation of sensory characteristics. However, more children with ASD (71.4% of the sample for the sensory processing patterns and 47% for the sensory processing modalities) were classified to the 'high atypical' cluster in comparison to the children with WS (37.5% of the sample for the sensory processing patterns and 25% for the sensory processing modalities). Also in both groups, parents of those children who had greater sensory processing difficulties reported that their children had more repetitive behaviours, higher levels of anxiety and greater intolerance of uncertainty. Interestingly, in both groups, differences in severity of autistic traits contributed to the higher presentation of repetitive behaviours and to the higher presentation of anxiety in those children who have greater sensory processing difficulties.

In some previous research involving ASD samples, three sensory clusters have been identified, low frequency of sensory symptoms, high frequency of symptoms and a mixed cluster with either high frequency of under-and over-responsivity and low frequency of seeking (Ben-Sasson et al., 2008) or sensory processing difficulties across all sensory domains except for Low Energy/Weak and Movement Sensitivity (as measured by the Short Sensory Profile; Lane et al., 2010) rather than the two cluster-solution found here. However, parents of toddlers with ASD with high frequency of hypo- and hyper-responsivity also reported their children to have higher negative emotionality, depression and anxiety symptoms than parents of children with ASD who had low frequency of sensory symptoms (Ben-Sasson et al., 2008). Additionally, it has been reported (Liss et al., 2006) that features that clustered together in ASD were overreactivity, overfocused behaviours and perseverative and stereotyped behaviours, suggesting that sensory hyperresponsivity in individuals with ASD leads to repetitive behaviours. The findings from this and previous studies (Ben-Sasson et al., 2008; Lane et al., 2010) that identified sensory clusters do not therefore support the sensory modulation disorder classification of sensory over-responsivity, under-responsivity and sensory seeking (Miller et al., 2007) as distinct, mutually exclusive subtypes of sensory abnormalities. Instead, they indicate that a more individual differences approach in describing

performance across sensory processing patterns or modalities is needed, as well as indicating that atypicalities across different subtypes may be present in one individual. The latter further supports reports of different patterns of sensory processing present within the same individual with ASD (Baranek, 2002; Baranek et al., 2006; Ben-Sasson et al., 2009) and suggests that similar groupings of sensory processing difficulties can occur in children with WS. Although the high rates of sensory atypicalities in WS have been reported previously (John & Mervis, 2010), this is the first study showing the similarities in presentation of sensory processing abnormalities in WS and ASD, not only in relation to presentation across individuals but also in relation to further associations between greater sensory difficulties and higher repetitive behaviours (as previously reported by Riby et al., 2013) and anxiety levels across these two neurodevelopmental disorders.

The secondary aim of the study was to examine the relationship between sensory atypicalities, repetitive behaviours, anxiety and intolerance of uncertainty in children with ASD and WS. Several significant negative relationships were found between sensory processing scores and RBQ total score, SCAS/PAS total score and ASC-ASD total score in both WS and ASD groups. These findings suggested that greater sensory processing difficulties were associated with more repetitive behaviours in both groups and higher anxiety levels in the WS and ASD group. Moreover, some of the associations appear to be syndrome-specific. In the WS sample only we found a significant negative relationship between the RBQ total score and Seeking, and the touch modality. The RBQ Sensory/Motor subscale was associated with Registration and vestibular and touch sensory modalities. In the ASD sample only, the RBQ Sensory/Motor subscale was negatively associated with Sensitivity, while Insistence on the Sameness subscale correlated negatively with vestibular and touch processing modalities. Interestingly, there were a number of significant associations found between sensory atypicalities and anxiety (e.g. the SCAS/PAS total score was associated with Registration and Sensitivity and with vestibular modality, while the ASC-ASD total score was negatively correlated with Seeking, Avoiding, Auditory and Touch processing), yet, these relationships were found only in the WS group. Similar patterns of relationships were found in both samples after controlling for gender.

A high degree of co-occurrence between sensory atypicalities and repetitive behaviours in ASD (Baker et al., 2008; Chen et al., 2009; Gabriels et al., 2008; Joosten et al., 2009) and in WS (Riby et al., 2013) have been previously reported. In contrast to previous findings, the relationship between a number of sensory atypicalities and anxiety in

individuals with ASD has not been supported (Ben-Sasson et al., 2008; Green et al., 2011; Liss et al., 2006; Pfeiffer et al., 2005) as only some of the anxiety scores were associated with touch modality. Interestingly, in this study some associations were found to be syndrome-specific (even after controlling for gender). In the ASD group a higher degree of insistence on sameness behaviours was associated with more vestibular and touch sensory processing atypicalities, while in the WS sample the same relationship was found between sensory/motor behaviours and vestibular and touch modalities. It is likely that some sensory difficulties may lead to specific repetitive behaviours to each neurodevelopmental disorder. For example, hyper-sensitivity to certain sounds and to certain food textures, tastes or smells in WS may result in displaying more insistence on sameness behaviours as a way to avoid and limit unpleasant sensations.

Interestingly, with regards to the relationship between sensory processing abnormalities and anxiety, only syndrome-specific associations were found in the WS group. Associations between Low Registration and Sensation Seeking and anxiety scores across measures were established. As hypo-responsiveness consists of Low Registration and Sensation Seeking in Dunn's model (1999) these findings are rather surprising. In the ASD research a strong relationship between hyper-responsiveness, overreactivity and anxiety has been reported a number of times (for review see: Chapter 2). There is some evidence, however, that individuals with WS can develop an intense fascination for certain stimulation that they found frightening at first (Levitin et al., 2005). Hence, it is possible that some children with WS avoid and then seek for anxiety-provoking stimulation.

The examination of the direct relationship between sensory processing abnormalities (both hypo- and hyperresponsiveness) and repetitive behaviours and the indirect path through intolerance of uncertainty and anxiety showed syndrome-specific paths. In the ASD group significant direct effects were found from sensory hyporesponsiveness to both sensory/motor behaviours and insistence on sameness, and from sensory hyperresponsiveness to both sensory/motor behaviours and insistence on sameness. None of these direct effects were significant in the WS sample. However, significant total effects across the groups and paths suggested that in the ASD group all the paths between sensory processing abnormalities and repetitive behaviours were partially mediated by at least one of the mediation variables (intolerance of uncertainty or anxiety), in the WS group the relationships between sensory processing abnormalities and repetitive behaviours were completely mediated via intolerance of uncertainty and/or anxiety, showing for the first time the potentially greater role of

intolerance of uncertainty and/or anxiety in the presentation of repetitive behaviours in WS sample.

Significant indirect effects from sensory hyporesponsiveness to insistence on sameness behaviours through anxiety, and intolerance of uncertainty and anxiety were found in the ASD group. The same path in the WS sample was significantly mediated via intolerance of uncertainty only. Similarly, in the WS sample, the relationship between sensory hyperresponsiveness and insistence on sameness behaviours was also mediated via intolerance of uncertainty. Such an importance of mediating role of intolerance of uncertainty between sensory atypicalities and repetitive behaviours in WS was not predicted. This novel finding suggests that those children with WS who find sensory environment unpredictable, can display more repetitive behaviours perhaps in order to regain predictability in their world. Although the role of intolerance of uncertainty has been already taken into account in anxiety treatments in typically developing population (McEvoy and Mahoney, 2012) and in one treatment programme for young people with ASD (Rodgers et al., 2016), it has not been targeted in any interventions designed for children with WS. The examination of the mediational role of intolerance of uncertainty between sensory processing abnormalities and repetitive behaviours in the WS group needs to be further undertaken.

Additionally, when the analysis was performed for each group separately for the clusters that emerged in the cluster analysis, within the ASD group the relationships between sensory processing abnormalities and repetitive behaviours were found only for participants who had greater sensory difficulties where the relationship between sensory hyperresponsiveness and insistence on sameness was fully mediated via intolerance of uncertainty. In the WS sample, in the group of participants who had more typical sensory processing, a relationship between sensory hyperresponsiveness and insistence on sameness was found. For those with WS who had greater sensory difficulties, the relationship between sensory hyporesponsiveness and sensory/motor behaviours was fully mediated via anxiety and/or intolerance of uncertainty.

Similarly to Wigham et al. (2015), direct relationships between sensory processing abnormalities and repetitive behaviours were found in our ASD group, including a direct path between sensory hyperresponsiveness and sensory/motor behaviours that was non-significant in Wigham et al. (2015) study. Lidstone et al. (2014), however, reported a significant relationship between repetitive motor behaviours and Sensation Avoiding, suggesting that sensory hyperresponsiveness may play a role in presentation of motor repetitive behaviours.

7.4.1 Strengths and limitations

A small sample size was used in the study. Adjusting for small sample size statistical methods (bootstrapping) were implemented, and the RRB variance accounted by the main models was large, ranging from 51% to 67%. Nevertheless, in the models in which participants with particular sensory subtype were entered, the RRB variance accounted by ranged from small to large, 28% to 81% for the WS sample and 2% to 57% for the ASD sample. Replication of the findings with a larger sample is required. Secondly, our sample comprised of children across ability and communication levels for both neurodevelopmental disorders, and as such the study findings may be more generalizable to other children with WS and ASD. While this is a strength of the current study it also carries some limitations. Comparing this study's findings with previous work may be hampered as commonly only participants with good language skills and with average or higher IQ are included in the research. Undertaking further research with less able individuals with ASD and WS is needed. Third, evidence of the direction of the association between sensory symptoms and mental age is mixed in the ASD literature, with some authors reporting that less developmentally mature children experience the most sensory processing problems compared to their more developed peers (Baranek et al., 2006). Others, on the other hand, do not find any association between the severity of sensory symptoms and IQ in ASD (or in a developmentally delayed group, however, IQ was moderately associated with sensory difficulties in Fragile X syndrome; Rogers et al., 2003). Although chronological rather than mental age contributes to the presentation of sensory processing difficulties in individuals with ASD (Ben-Sasson et al., 2009), the role of IQ as a contributing factor to the nature of sensory symptoms in WS is unknown and requires further investigation. Fourth, the intolerance of uncertainty subscale from the ASC-ASD as a measure of intolerance of uncertainty was used in this study. The ASC-ASD is primarily an ASD-specific anxiety measure that has not been validated in the WS population or in younger children with ASD. Although, the intolerance of uncertainty subscale consists of only 8 items comparing to 27 item Intolerance of Uncertainty Scale, (Freeston et al., 1994) or 12 item Intolerance of Uncertainty Scale - Short Form (IUS-12; Carleton et al., 2007), it has good psychometric properties (Rodgers et al., 2016). Further psychometric work on an intolerance of uncertainty scale for use with younger children with ASD and children with WS is needed. Finally, the motor behaviour subscale of the RBQ contains some sensory-related items which could impact part of the examination of the relationship between sensory processing and repetitive behaviours. Clear distinction between sensory only and motor only repetitive behaviours should be undertaken in the future research.

7.4.2 Conclusion

This is the first study which has explored the role of intolerance of uncertainty and anxiety in relation to sensory processing abnormalities and repetitive behaviours in young children with ASD across the spectrum and in young children with WS. A high degree of co-occurrence between sensory atypicalities and repetitive behaviours and anxiety in both neurodevelopmental disorders and some syndrome-specific associations have been highlighted. The greater role of intolerance of uncertainty and anxiety in relation to sensory difficulties and repetitive behaviours than expected in WS is surprising, yet the results support the value of considering the complexity of the mechanisms underlying the relationship between sensory processing abnormalities and repetitive behaviours across neurodevelopmental disorders. Understanding these relationships would support developing uncertainty- and anxiety-related treatment protocols better tailored to a specific disorder.

Chapter 8. Sensory atypicalities in dyads of children with autism spectrum disorder and their parents

8.1 Background

It has been shown in Chapter 7 that sensory processing atypicalities can be associated with some other characteristics of ASD, such as anxiety and repetitive behaviours. It has seen also been previously reported that sensory processing difficulties can present significant challenges across a wide range of daily life for a child with ASD, including attention, ability to learn, emotion regulation and effective management of interpersonal relationships with both peers and family members.

It is known that there is a heritable component to ASD (Silverman et al., 2002) as shown by twin studies (Bailey et al., 1995; for the review see Ronald & Hoekstra, 2011). Interestingly, some unaffected relatives of individuals with ASD, including parents have been reported to have a number of autism-related traits, and subclinical atypicalities in social and communication skills (Gerdts & Bernier, 2011), including language skills (Ruser et al., 2007) and memory (Baron-Cohen & Hammer, 1997). This phenomenon of increased likelihood of autism-related traits in some family members of individuals with ASD (Bernier et al., 2012), known as the broader autism phenotype (BAP), has rarely been investigated in relation to sensory atypicalities.

Only one study to date (Uljarevic et al. 2014) has examined sensory processing in parents of individuals with ASD. The authors reported elevated levels of sensory atypicalities in mothers of children and adolescents with ASD, with 98% of mothers of children with ASD having sensory processing scores within an atypical range on the Adolescent/Adult Sensory Profile (AASP; Brown & Dunn, 2002) compared to a normative sample. In a similar study De la Marche, Steyaert and Noens (2012) assessed sensory processing in adolescent siblings of individuals with ASD and reported that non-affected siblings shared some aspects of an atypical sensory processing profiles with their affected sibling. In addition, data from baby siblings of children with ASD show that sensory processing differences, in particular difficulties with auditory processing and lowered registration of sensory stimulation, were more common in high-risk siblings subsequently diagnosed with ASD than in typically developing infants (Germani et al., 2014; Loh et al., 2007; Mulligan & White, 2012). These findings suggest that behavioural responses to sensory input may serve as an early risk marker of ASD, particularly in high-risk infants.

The relationships between sensory processing profiles in ASD families may not be unique to the disorder. A level of sensory heritability (perceptual sensitivity) and sensory

over-responsivity in relation to both tactile and auditory processing has also been reported in the general population in monozygotic and dizygotic typically developing twins (Goldsmith et al., 1997; Van Hulle et al., 2012). Furthermore, a strong association between sensory sensitivities and autistic traits in the general population has been shown (Robertson & Simmons, 2013). Taking these findings together the limited evidence to date suggests that parents of children with ASD may also present with atypicalities in their sensory processing profiles. Surprisingly, the relationship between sensory atypicalities in matched dyads of children with ASD, and developing typically children and their parents has not been investigated.

Investigation of similarities and differences in sensory processing in parent-child dyads in neurodevelopmental disorders will inform our understanding of how phenotypic profiles may be inherited within families. The concordance in sensory profiles between individual parent and child dyads in ASD families has never been examined. Therefore, the aim of this study was to explore the profiles of sensory processing in child-parent dyads within ASD families in comparison to TD dyads. We hypothesised that (1) parents of children with ASD would present with more sensory atypicalities than parents of typically developing children and (2) sensory processing patterns in child-parent dyads would be more similar in ASD families than in typically developing families.

8.2 Methods

8.2.1 Participants

Forty-four parents (38 mothers and 6 fathers) of children with ASD and thirty parents (25 mothers and 5 fathers) of typically developing (TD) children were recruited. All children with ASD had previously been diagnosed with ASD based on a multidisciplinary team assessment following the guidelines of the UK National Autism Plan for Children (Le Couteur, 2003). Additionally, for the children with ASD data from the Social Responsiveness Scale, Second Edition (SRS-2; Constantino & Gruber, 2012) were available for all children of an appropriate developmental age, with the exception of four (due to a large amount of missing data), with the scores falling between the mild to moderate (n=4; total raw score ranging from 58 to 80, mean=70, SD=9.38) and severe range (n=31; total raw score ranging from 88 to 171, mean=116.9, SD=23.73). Children for whom the SRS-2 total score could not be calculated, did not differ on gender, age and any sensory variable compared to children for whom the SRS-2 data were available. All TD children obtained scores within the normal

range (0-13; mean=6.70, SD=3.73) on the Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997).

Children with ASD were recruited via ASD-UK (www.ASD-UK.com), a major UK family research database of children with ASD (Warnell et al., 2015). Families whose children met the study criteria were initially sent information about the study by email or letter, and reminders were sent to non-responders. In order to ascertain whether ability plays a role in sensory atypicalities presentation, children across the ability range were recruited, so the sample included those with and without comorbid intellectual disability (ID) as reported by parents. Twenty-three children in the ASD sample also had an intellectual disability (ID). TD children were recruited through local schools, a University research volunteers' database and word of mouth.

8.2.2 Measures

1. The Sensory Profile (SP; Dunn, 1999) is a caregiver questionnaire that measures a child's sensory processing abilities. The questionnaire consists of 125 items, rated on a five-point Likert scale, ranging from always (1) to never (5) (see Chapter 4).

2. The Adolescent/Adult Sensory Profile (AASP; Brown & Dunn, 2002) is a self-report questionnaire designed for individuals between 11 and 65 years old evaluating their responses to everyday sensory events. In this 60-item questionnaire, 15 questions are related to each of the four sensory quadrants—low registration, sensation seeking, sensory sensitivity, and sensation avoiding. Scores for taste/smell, movement, visual, touch and auditory processing can also be calculated (to be consistent with the SP domains, we refer to taste/smell sensory processing using oral sensory processing term, and to movement sensory processing, using vestibular sensory processing term). As in the SP, each statement is rated on a five-point Likert scale; however, the rating system is reversed, ranging from almost never (1) to almost always (5) (see Appendix U). Some individuals may have atypical scores in more than one sensory quadrant. The internal consistency of the measure is s good with alpha values ranged from .63 to .77, as reported in the measure manual, for the various quadrant scores. Evidence of good convergent and discriminant validity was also provided (AASP; Brown & Dunn, 2002).

3.The Social Responsiveness Scale – Second Edition (Constantino & Gruber, 2012) is a 65-item parent-report four-point Likert-like rating scale of autistic trait that covers unusual interpersonal behaviours, communication or repetitive/stereotyped behaviours. The SRS-2 describes a degree of autistic social impairment and the severity of autistic symptoms. It is reported to have good psychometric properties (Bruni, 2014).

4.The Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997) is a 25-items caregiver-report of children of 4-16 years old that screens whether the child has any emotional, conduct, hyperactivity/inattention, and peer relationship problems or displays prosocial behaviour. The SDQ has been widely used in large epidemiological studies and is well adapted for studies of the general population (Goodman & Goodman, 2009).

8.2.3 Procedure

Questionnaire packs including an information sheet, consent form, the Sensory Profile (Dunn, 1999), Adolescent/Adult Sensory Profile (Brown & Dunn, 2002), Social Responsiveness Scale – Second Edition (Constantino & Gruber, 2012; parents of ASD children only), and the Strengths and Difficulties Questionnaire (Goodman, 1997; parents of TD children only) were sent to parents who had agreed to participate in the study. Favourable ethical opinion was granted by Newcastle University Faculty of Medical Sciences Ethics Committee.

8.2.4 Data analysis

After initial data entry, parents were contacted again and asked to provide missing information, if relevant. Some parents did not respond resulting in 1.27% of the SP and 0.09% of the AASP item scores missing. There were no patterns within missing data. Missing values were treated as missing at random and replaced by the mean of the non-missing subscale items when less than 20% of the data within the subscale were missing. Descriptive statistics, inferential and Intraclass Correlation Coefficient (ICC) analyses were subsequently undertaken on the complete dataset for both quadrant scores and sensory processing modalities scores. Intraclass Correlation Coefficients were used to quantify the agreement between parent-child pairs and establish consistency between the sensory processing measurements for the pairs.

8.3 Results

Descriptive statistics of participant characteristics are presented in Table 8.1.

Table 8.1 Mean (SD) scores and effect sizes on participant demographics and outcome variables

	ASD total (*n*=44)	TD (*n*=30)	
Variable	Mean (SD)	Mean (SD)	Cohen's d
Child data			
Male	36	18	
Age in years	8.07 (3.33)	8.41 (2.98)	
Registration	52.68 (12.02)	70.0 (4.10)	-1.93
Seeking	83.34 (17.20)	114.6 (10.68)	-2.18
Sensitivity	66.41 (11.68)	91.13 (6.55)	-2.61
Avoiding	93.16 (16.62)	123.37 (11.47)	-2.12
Auditory	22.41 (5.79)	34.10 (4.71)	-2.21
Visual	30.45 (6.26)	38.63 (4.09)	-1.55
Vestibular	42.05 (6.59)	50.63 (3.96)	-1.58
Touch	59.52 (11.72)	83.03 (5.50)	-2.57
Oral	40.66 (9.47)	54.43 (5.30)	-1.79
Parent data			
Male	6	5	
Age in years	41.43 (7.03)	41.72 (4.67)	
Registration	28.89 (7.21)	25.53 (6.7)	0.48
Seeking	42.93 (7.92)	44.6 (5.39)	-0.25

Sensitivity	35.57 (9.37)	30.0 (5.22)	0.73
Avoiding	34.14 (10.65)	28.77 (5.73)	0.63
Auditory	25.18 (7.29)	22.13 (4.72)	0.50
Visual	23.61 (5.19)	20.90 (3.43)	0.62
Vestibular	19.73 (4.87)	18.43 (3.18)	0.32
Touch	28.75 (6.20)	27.03 (5.77)	0.29
Oral	19.82 (3.16)	18.50 (2.27)	0.48

Note: lower scores in child data and higher scores in parent data indicate more sensory atypicality

8.3.1 Sensory quadrants

There were no significant differences in the sensory scores between mothers and fathers in each group and between ASD children with ID and without ID. Further analyses were performed on all parents together (irrespective of gender) and all ASD participants together (irrespective of ability level).

First, one way ANOVA analyses were performed to compare group means on the sensory scores. Parents of children with ASD had significantly higher scores than parents of TD children in the Registration, Sensitivity and Avoiding quadrants ($F_{(1,72)}$=4.08, p=.047 $F_{(1,72)}$=8.72, p=.004 and $F_{(1,72)}$=6.36, p=0.014 respectively), with a higher score indicative of more atypicality. There were no other differences between the parent groups (see Table 8.2).

Table 8.2 One way ANOVA statistics on the mean sensory quadrants and modality scores between ASD and TD children; and parents of children with ASD and those typically developing (only mothers included in the analysis of modalities)

	Child data (ASD vs TD)		Parent data (ASD vs TD)	
Variable	*F* value	*p*	*F* value	*p*
Registration	57.48	<.001	4.08	.047
Seeking	78.31	<.001	1.01	.318
Sensitivity	110.42	<.001	8.72	.004
Avoiding	74.67	<.001	6.36	.014
Auditory	84.26	<.001	3.06	.085
Visual	39.55	<.001	3.73	.058
Vestibular	40.81	<.001	1.05	.309
Touch	104.58	<.001	0.43	.512
Oral	52.16	<.001	5.69	.020

Subsequently, Intraclass Correlation Coefficient analyses (ICC; two-way mixed, consistency) were undertaken. Two approaches were implemented for the sensory quadrants scores.

The first approach aimed to evaluate the categorical nature of the data. Child and parent sensory quadrants scores were coded into one of the five categories: 'much less than most people', 'less than most people', 'similar to most people', 'more than most people', 'much more than most people' following the manuals' guideline (Brown and Dunn, 2002; Dunn, 1999). The level of agreement for sensory quadrants between parent-child dyads was then calculated for the categorical data. Significant agreement was obtained between parents and their children *sensory sensitivity* scores in ASD dyads and on *low registration* and *sensation avoiding* in TD dyads.

The second approach aimed to mirror the dimensional nature of the data. Due to directional differences in the Likert scale scoring of the SP and AASP (e.g. score 1 is interpreted as 'always' in the SP and refers to 'almost never' in the AASP), the Z scores of sensory quadrants were calculated (and reversed for the parental data) to estimate the level of

agreement for sensory quadrants between parent-child dyads. The ICC results are shown in Table 8.3. Significant agreement was obtained between parents and their children in both groups on *low registration* and *sensory sensitivity* scores. There were no significant correlations between parent-child *sensation seeking* scores in either group. A significant association was found between parental and child scores on *sensation avoiding* within ASD dyads, however, that correlation was non-significant within TD dyads.

Table 8.3 Intraclass Correlation Coefficients (ICC) for parent-child dyads for ASD and TD samples with corresponding 95% confidence intervals (CI)

	ASD			TD		
Variable	ICC	*p*	95% CI	ICC	*p*	95% CI
Registration	.42*	.040	-.07 to .68	.78*	<.001	.53 to .89
Seeking	.19	.245	-.48 to .56	.29	.183	-.50 to .66
Sensitivity	.48*	.018	.04 to .71	.55*	.019	.05 to .78
Avoiding	.45*	.026	.01 to .70	.41	.077	-.23 to .72
Auditory	.47*	.028	-.02 to .72	.36	.140	-.45 to .72
Visual	.77*	<.001	.55 to .88	.33	.164	-.51 to .71
Vestibular	.45*	.038	-.07 to .71	.47	.064	-.20 to .77
Touch	.60*	.003	.23 to .79	.81*	<.001	.58 to .92
Oral	.05	.43	-.82 to .51	.39	.116	-.38 to .73

Note: * indicates significant results

8.3.2 Sensory processing modalities

There were significant differences on the sensory processing modality scores between mothers and fathers in each group (ASD group: taste/smell: $t(42)=-1.997$, $p=.05$, movement: $t(42)=-1.401$, $p=.17$, visual: $t(42)=-.645$, $p=.52$, touch: $t(42)=-.035$, $p=.97$, auditory: $t(42)=2.338$, $p=.02$; TD group: taste/smell: $t(28)=-.106$, $p=.92$, movement: $t(28)=-2.345$, $p=.03$, visual: $t(28)=-2.206$, $p=.04$, touch: $t(28)=-1.582$, $p=.12$, auditory: $t(28)=1.873$, $p=.07$). Further analyses were therefore performed only on mothers.

First, one way ANOVA analyses were performed to compare group means on the sensory processing modality scores. Children with ASD had significantly lower scores ($p<.001$) than TD children in all modalities, with a lower score indicative of more atypicality. Mothers of children with ASD had significantly higher scores than mothers of TD children in

the taste/smell modality ($F_{(1,62)}$=5.69, p=.020), indicating more atypicality. There were no other differences between the mothers' groups (see Table 8.2).

Intraclass Correlation Coefficient analyses (ICC; two-way mixed, consistency) showed that significant agreement was obtained between mothers and their children in both groups on touch processing scores. A significant association was found between parental and child scores on auditory, visual and vestibular sensory processing within ASD dyads, however, those correlations were non-significant within TD dyads (see Table 8.3).

ICC analysis was not performed for the sensory processing modalities categorical data as the AASP manual did not provide information on classification of sensory processing modalities scores.

8.4 Discussion

This is the first study exploring sensory processing atypicalities in dyads of children with ASD and their parents, compared to typically developing children. Parents of children with ASD showed significantly more *over responsivity* sensory atypicalities, with higher scores on *sensory sensitivity* and *sensation avoiding* and more *low registration* difficulties compared to parents of TD children. Also mothers of children with ASD showed more taste/smell sensory processing related difficulties than mothers of TD children. The effect sizes between the groups ranged from small to medium. A similar level of agreement was obtained within ASD and TD parent-child dyads on sensory atypicalities, showing that to a degree sensory processing might be universally heritable within families, irrespective of ASD status. Categorical data analysis suggested a significant relationship between parent and child *sensory sensitivity* scores in ASD families. Further ICC analysis of Z-scores showed significant associations between parent and child quadrant scores on *sensation avoiding,* and sensory processing scores on auditory, visual and vestibular processing in ASD families only.

In this study parents of children with ASD showed atypical sensory processing on three sensory quadrants (*low registration*, *sensory sensitivity* and *sensation avoiding*) in comparison to parents of typically developing children. These data are in contrast to the Uljarevic et al. (2014) study, where parent group differences were found for all sensory quadrants. However, in the current study, TD parent data were obtained directly from a control group and inferential analyses were performed. In Uljarevic et al. (2014) sensory scores of parents of children with ASD were compared to the original American normative

sample (Brown & Dunn, 2002). Further work on psychometric properties of the tool and replication of this study are required.

With regards to the results on sensory quadrants, our findings might suggest a genetic contribution for *sensory sensitivity*, in parent-child dyads. Interestingly, a similar level of agreement was found between parent and child data for both the ASD and TD groups, on the *sensitivity* quadrant suggesting that that aspect of sensory processing might be heritable, irrespective of ASD status in the dimensional data analysis. Findings from the categorical data analysis, however, suggested that ASD status might play a role in sensory sensitivity heritability. We did not find agreement between parent and child scores on the *sensation seeking* quadrant in either group. De la Marche et al. (2012) reported that both adolescents with ASD and their siblings had reduced *sensation seeking* and argued that sensory seeking atypicalities might be a candidate endophenotype. In this study, ASD participants showed more difficulties related to *sensation seeking* than their TD peers. Also in contrast to the familial relationship reported by De la Marche et al. (2012) we found no significant difference between parents of children with ASD and parents of typically developing children on that quadrant. This might suggest that *sensation seeking* atypicalities are not heritable, but may be more related to the presence of sensory atypicalities common for individuals with ASD or inherent in the other aspects of the disorder. The sensory processing differences in the ASD participants between the studies could also be explained by age discrepancies in the samples as younger individuals with ASD are reported to show more sensory atypicalities than adolescents (for review, see Ben-Sasson et al., 2009).

Although support for the familiality of *sensation seeking* was not found, agreement between parent and child scores on the s*ensation avoiding* quadrant was found for the ASD dyads only, which suggests that this aspect of the atypical sensory processing profile may be heritable solely within ASD families. This phenomenon needs further investigation.

As in previous studies (Kern et al., 2006; Kientz and Dunn, 1997), we found that children with ASD had more sensory processing difficulties across different modalities than typically developing children. Goldsmith et al. (2006) investigated heritability of auditory and tactile defensiveness in twin study of the general population. They found that tactile defensiveness demonstrated greater heritability than auditory defensiveness. Our study supports that, as a similar level of agreement was found between parent and child data for both the ASD and TD groups on the *tactile sensory processing* quadrant. However, the findings also showed that for auditory, visual and vestibular sensory processing an agreement

was found between child and parent scores, suggesting that for these aspects of sensory processing familial factors might play a role only within ASD families.

While our data might support the notion that sensory atypicalities may form part of the broader autism phenotype we cannot rule out the role of the environment on the development of atypical sensory profiles. There is a strong evidence that fearful behaviours can be modelled by parents and in turn increase fear in children (de Rosnay et al., 2006; Gerull & Rapee, 2002). It has been shown that parents who experience anxiety think about their children's environments as threatening and are more likely to interpret ambiguous situations, including those child-related, as possibly distressing (Gallagher & Cartwright-Hatton, 2009). According to Rachman's three pathways to fear (Rachman, 1977), anxiogenic learning experiences can be provided by the parents by verbal threat information, negative vicarious learning and direct aversive conditioning experiences. It is possible that the same process takes place in the intergenerational transmission of sensory-related anxieties. Parents may react to or describe certain sensory situations as threatening, modelling to their children how distressing sensory experiences can be, resulting in the attribution of fear or distress to those stimuli by the child. However, this intergenerational transmission might also occur in the opposite direction, from the child to the parent. It is possible that some parents of children with ASD become more avoidant of certain sensory events because of their child's often aversive, anxious and avoidant response to those sensory stimuli and this this pattern is subsequently reinforced. It has been suggested that parents of children with ASD may use an escape-avoidance coping style to deal with stressful situations more often than parents of typically developing children (Dabrowska & Pisula, 2010). It has been also shown that those mothers who were more anxious compared to nonanxious mothers, expected their children to perform more poorly on a number of experimental tasks (Creswell, Apetroaia, Murray, & Cooper, 2012), hence their perception of their children performance was biased. It is then possible that parental anxiety or stress could have influenced parental reporting of children's sensory problems.

In order to assess whether increased levels of *sensation avoidant* behaviours are a consequence of genotype or learnt coping strategies, longitudinal studies are needed. To establish whether auditory, visual and vestibular sensory processing atypicalities constitute a part of the broader autism phenotype, a replication study is required.

8.4.1 Study limitations

The present study has a number of limitations. Two different baseline tools were used in the children's evaluation of autistic symptoms and emotional and behavioural difficulties

(SRS-2 and SDQ). Although the measures were appropriate for the samples, using only the SRS-2 would allow for more direct comparison of some of the behavioural features between the groups. A small sample size restricted further investigation of the level of agreement between parent and child sensory profile scores for young children and adolescents with ASD separately. There is evidence suggesting that patterns of sensory processing change with development in individuals with ASD (Ben-Sasson et al., 2009) and it is unknown which aspects of sensory profiles would be shared between parents and their young or adolescent children with ASD. Also the data were obtained only from parental reports and no direct measures of sensory processing were applied. Moreover, children with ASD without co-morbid ID were not asked to complete the SP questionnaire themselves, which could enrich our understanding of sensory processing in individuals with ASD. Information on sensory quadrant scores from mothers and fathers were combined, and presented for mothers only on sensory processing modalities. It has been suggested that females present more sensory atypicalities than males (Goldsmith et al., 2006) and further investigation of whether a similar pattern can be found in parents of children with ASD is needed, requesting recruitment of fathers of children with ASD. Although a control group of parents of TD children was recruited to the study, including the children and parents of children with other neurodevelopmental disorders would benefit our understanding of the specificity of these findings to ASD. Last, but not least, in this preliminary study investigating sensory processing patterns in parent-child dyads, a measure of parental broader autism phenotype traits was not used. Elevated BAP features in parents could not only possibly indicate parents with atypical sensory processing, but also impact parental ability to report on their children's sensory experiences. It is likely that highly sensitive parents might have been biased toward perceiving similar traits in their children, and equally, parents who are less sensitive might have been reporting their children as less bothered by everyday sensory input. Further studies investigating sensory processing in parents of children with ASD would benefit from including a BAP measure.

8.4.2 Conclusions

In conclusion, sensory profiles were similar for parent-child dyads across both groups, however children with ASD and their parents shared more sensory avoidant behaviours, and auditory, visual and vestibular sensory processing atypicalities compared to TD dyads. Some sensory characteristics might therefore need to be included into the broader autism phenotype features, alongside well-established social communication skills and personality traits (Gerdts & Bernier, 2011). It is also possible that attitudes towards sensory experiences are transmitted

inter-generationally. Further investigation of whether sensation avoiding, auditory, visual and vestibular atypicalities in parents of children with ASD have genetic or environmental origin, or are a result of interaction between the two, is needed.

Chapter 9. Development of the tactile and auditory sensory observation schedules

9.1 Background

To assess sensory atypicalities in children with autism and WS, researchers have mostly relied on caregiver-report questionnaires (Glod et al., 2015; see Chapter 2 and Chapter 3 for review) with the most commonly used the Sensory Profile (SP; Dunn, 1999) and Short Sensory Profile (SSP; McIntosh, Miller, Shyu & Dunn, 1999a). Although retrospective parent- or caregiver-sensory questionnaires are considered valuable screening tools for sensory symptoms, they may also be a source of recollection bias or provide inaccurate responses (Hoyle et al., 2001). There are only a handful of observational tools administered directly to participants that assess sensory difficulties, which have been used with children with ASD; yet, the literature suggests that semi-structured direct observation is a thorough and accurate assessment that, combined with the parent-report, provides a more reliable assessment (Achenbach & Rescorla, 2004). The Sensory Processing Assessment for Young Children (SPA; Baranek, 1999b) is a semi-structured play-based assessment evaluating hyporesponsiveness, hyperresponsiveness and sensory seeking patterns of sensory processing across different modalities. The Tactile Defensiveness and Discrimination Test (TDDT-R; Baranek, 1998; Baranek & Berkson, 1994) is an observational assessment of tactile processing, examining hyperresponsiveness and discrimination skills in children with autism and other developmental disabilities. The SensOR Assessment (Schoen et al., 2008) evaluates the severity of sensory hyperresponsiveness across different modalities in participants between 3 and 55 years old, either typically developing or with symptoms of sensory overresponsivity. Similarly, the Sensory Processing Scale Assessment (SPS; Schoen et al., 2014) evaluates sensory reactivity difficulties in typically developing individuals with and without SMD. The tool measures sensory hyper-reactivity, hypo-reactivity and craving/seeking behaviours across three domains (vision, hearing and touch) and has been used with children with ASD (Tavassoli et al., 2016). The SPS is the only measure assessing sensory processing modalities, taking into account both hypo- and hyper-reactivity patterns in children with ASD that are compatible with new diagnostic criteria (DSM-V; APA, 2013). None of the direct observational tools evaluating sensory modulation have been administered to children with WS. At the beginning of this study, neither the Tactile Defensiveness and Discrimination Test, the SensOR Assessment, nor the Sensory Processing Scale Assessment were available for research or clinical use.

Through parent-reports and direct assessments various modalities can be evaluated. In both autism and WS, and in typical development, atypicalities in the tactile and auditory modalities have attracted a lot of research attention. Goldsmith and colleagues (2006) showed evidence of a distinction between tactile and auditory over-responsivities in a population based sample of toddlers, finding low correlations between the subscales of the Sensory Defensiveness subscales of the revised Toddler Behavior Assessment Questionnaire (TBAQ; Goldsmith, 1996), differences in the gender distribution of auditory and tactile defensiveness and distinct patterns of heritability. Thus, specific genetic factors were attributed to individual variation in auditory and tactile defensiveness. Further findings from a factor analytic study of individuals with developmental disabilities (including intellectual disability, autism and mixed aetiology) suggests that tactile sensitivity behaviours cluster separately from sensitivities in other modalities (Baranek et al., 1997). Both tactile symptoms and auditory filtering were found to be significantly elevated in children with autism compared to those children who were typically developing or had a developmental delay (Rogers et al., 2003), and were independent of social-communication symptoms.

Some studies have investigated the relationships between sensory modalities and other symptoms, present across neurodevelopmental disorders. For example, tactile and auditory processing significantly correlated with restricted and repetitive behaviours in individuals with ASD (Chen et al., 2009; Foss-Feig et al., 2012). This evidence supports making a distinction between the tactile and auditory modalities and other sensory modalities. In WS, nearly 60% of children with WS are reported as having definite abnormalities on the auditory filtering subscale of the Short Sensory Profile (John & Mervis, 2010). Although little work has been undertaken examining tactile symptoms, Riby et al. (2013) concluded that some repetitive behaviours (e.g. repetitive movement) may be a consequence of specific types of sensory problems (e.g. tactile sensitivity) in individuals with WS.

Despite the fact that sensory symptoms are very common in both autism and WS and are believed to impair social functioning and communication (Boyd et al., 2009; John & Mervis, 2010; Watson et al., 2011), there is no agreement on a gold standard set of measures that best evaluates sensory processing, not only in neurodevelopmental disorders, but also in typical development. Parent-reports have been widely used to assess sensory symptoms in children, yet, more objective, direct assessments of specific sensory modalities in young children, are needed. The goal of this study was to develop, administer and evaluate the Tactile and Auditory Sensory Observation Schedule (TASOS) for typically developing, ASD and WS children. Therefore, the aims were fourfold, to examine:

(1) the feasibility and acceptability of the newly developed measure with all three samples,
(2) group differences between typical, ASD and WS samples on the auditory and tactile processing subscales of the SP and the TASOS,
(3) convergent validity of the TASOS relative to the SP,
(4) TASOS items that were most informative and discriminative between the typical and neurodevelopmental samples, and between ASD and WS samples.

9.2 Methods

9.2.1 Recruitment

Children between 4 and 9 years of age, those typically developing, with ASD or WS were invited to take part in the research project ('Touch, hear, react' study, see Chapter 4 for further details). Those children who, as well as their main diagnosis, had any other comorbid diagnoses of neurodevelopmental disorders or had visual, hearing or motor impairments were excluded from the study. Families whose children met the study criteria were initially sent information about the study by email or letter, and reminders were sent to non-responders. Children and their parents participated on a voluntary basis. Parents were asked to give consent for themselves and their child to take part in the study. Additionally, a verbal assent was sought from each child. Favourable ethical opinion was granted by Newcastle University Faculty of Medical Sciences Ethics Committee.

9.2.2 Participants

Twenty-three typically developing (TD) children, twenty-three children with ASD and seventeen children with WS between 4 and 9 years old and their parents were recruited to the study ('Touch, hear, react' study). Typically developing children were recruited through local primary mainstream schools in the North of England. Out of twenty-three typically developing children, data on the Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997) was available for seventeen. Fourteen children obtained scores within the normal range (1-12; mean=7.07, SD=3.79). Three TD children who obtained their SDQ total score within atypical range (17-21) were removed from further analyses.

Children with ASD were recruited through two different routes: local mainstream and special needs primary schools and a newsletter distributed by a local branch of 'Contact a Family'. 'Contact a Family' is a national charity for families with disabled children which provides information, advice and support to the families. The charity releases a newsletter in

which research studies can be advertised. All children with ASD had previously been diagnosed with ASD based on a multidisciplinary team assessment following the guidelines of the UK National Autism Plan for Children (Le Couteur, 2003) as stated by the parents. Additionally, for nineteen children with ASD data from the Social Responsiveness Scale - Second Edition (SRS-2; Constantino & Gruber, 2012) were collected, however data were only available for sixteen (for three children total raw score was not calculated due to large amount of missing data). Scores ranged between 62 and 175, mean=111.13, SD=35.94. Scores for eight children were in the mild to moderate range, and 8 were in the severe range. Children for whom the SRS-2 total score could not be calculated, did not differ on gender, age and any sensory variable compared to children for whom the SRS-2 data were available.

Children with WS were recruited via the Williams Syndrome Foundation. All WS children had previously been clinically diagnosed with the syndrome with the diagnosis confirmed by positive fluorescent in situ hybridization testing (FISH). Moreover, the SRS-2 data on sixteen children with WS were also collected and the total scores ranged from 34 to 141, mean=80.0, SD=25.54. Three children were within the normal range, ten within the mild to moderate range and three within the severe range. In order to ascertain whether ability plays a role in sensory atypicalities presentation, children across the ability range were recruited.

9.2.3 Measures

All children recruited to the study were asked to undertake the following tasks:

1. Raven's Coloured Progressive Matrices (RCPM; Raven et al., 1998) is a measure of non-verbal reasoning ability, designed for children between 4 and 11 years old, including those with intellectual disability and limited language skills. The children were presented with a pattern with a missing piece and asked to choose one of the six pattern blocks that best fits into the missing gap. This engaging and jigsaw-like test took between 15 to 30 minutes to administer.

2. British Picture Vocabulary Scale - Third Edition (BPVS3; Dunn et al., 2009) is a tool assessing a child's receptive vocabulary, and can be used with children as young as 3 years old. During the assessment an examiner said a word and a child was asked to select one from four pictures that best illustrate the word. The measure has been used with children autism and other communication difficulties. It took between 5 and 30 minutes to complete the test, depending on a child ability.

3. Tactile and auditory sensory observation schedule is a new play-based measure assessing hypo- and hyper-responsiveness to auditory and tactile stimuli, developed for this

study. The content was guided by statements included in the Sensory Profile (SP; Dunn, 1999), which measures sensory processing abilities in children. Other sensory questionnaires were also consulted (e.g. Sensory Questionnaire, Liss et al., 2006; Sensory Checklist, Biel & Peske, 2005; Sensory Processing Disorder checklist, http://www.sensory-processing-disorder.com/sensory-processing-disorder-checklist.html) to make sure that all appropriate items were considered and included in the tool.

Those questionnaire-based auditory and tactile items that could be tested experimentally and easily observed in everyday play situations, were used in the assessment. Some of the questionnaire items were not suitable to be included in a direct assessment, for example those relating to the perception of pain or responsiveness to temperature changes. Short auditory and tactile games were created in order to facilitate detecting targeted sensory behaviours.

Moreover, the relevant literature was searched in order to identify the most common sounds and textures to which typically and atypically developing children were particularly sensitive. Furthermore, a Facebook ad posted via the National Autistic Society (NAS) was created asking parents of children with ASD to describe textures that their children enjoyed touching and those that they did not enjoy. The summary of the feedback can be found below:

- ‘My daughter hates fluffy or hairy things and hates jelly but loves anything wooden’
- ‘Dislikes flour/sand/sugar (anything fine powder/grainy that gets under the nails)’
- ‘My son likes soft fluffy things. Hates sand, gooey things, paint etc.’
- ‘Particularly loves silky smooth i.e. clothes labels to touch’
- ‘My son hates things like glue paint sand etc. but loves water!’
- ‘Doesn't like fluffy things, sand or anything gluey or sticky’
- ‘Loves to scratch along an elasticated waistband on his trousers or pjs’
- ‘Glue is not a good one for my son and he doesn’t like labels in clothes’
- ‘He can only wear cotton tracksuits (occasionally jeans but he's not comfy in them). He hates the feel of embroidered labels or normal labels in clothes. He can't bear light touch, particularly can't stand his hair or back being touched (explains why he used to bite me as a toddler when I picked him up lol). We once had to send a brand new sofa back because he couldn't stand the soft dralon feel to it, short piled fluffy fabric, yet loves long piled rugs. He doesn't like the super soft feel of our current sofa, which is essentially a fake suede so we have to have a cotton woven throw on it. He doesn't

like bed linen, particularly flannelette but that's more to do with him fearing restraint as he rips them (deliberately) if his feet get tangled. He has the same problem restraint wise with coats but he also can't stand the way the sleeves "swoosh". Towels have to be proper cotton terry ones (posh deep or cheap thin ones, doesn't matter as long as it isn't the over soft velvety feeling ones). So basically he doesn't do really super scratchy and doesn't do uber-soft velvety. In between is good'

- 'My son loves blankets, feels them, sniffs them, covers his head in them. The cellular type. Doesn't like any tight fitting of clothes or anything with mesh in'.

This information was used to ensure that the assessment included tactile and auditory stimuli that could provoke hypo- or hyper-responsive reactions in children.

In the auditory domain eight items were included:
Item 1. Sound seeking (codes the child's unusual interest in sound/making sound),
Item.5. Response to hearing specific sounds (codes the child's response to a number of unexpected sound presses e.g. flushing toilet, dog barking, car horns, police siren, aeroplane),
Item 6. Response to radio while playing on the tablet/colouring in (codes the child's response to radio sounds),
Item 7. Response to background noise (refrigerator, people talking, traffic noise) (codes the child's response to background noise - social and non-social),
Item 8. Response to hearing specific sounds and pointing to a picture that best matches the sound (codes the child's response to a number of loud sound presses),
Item 9. Response to name (codes the child's response to hearing his/her name),
Item. 10. Response to non-social sound – whistle (codes the child's response to non-social sound),
Item 11. Response to 'special' interest word (codes the child's response to 'special' interest word).

In the tactile domain 13 experimental tasks were included:
Item 2. Need for touching certain fabrics (codes the child's display of unusual need for touching certain fabrics),
Item 3. Touching objects and others (codes the child's touching behaviours),
Item 4. Child's need to be touched (codes child's unusual need to be touched by others or objects),

Item 12. Sensitivity to certain fabrics (codes the child's sensitivity to certain fabrics (e.g. cotton, wool, silk, stone, sand, wood, hay, sand paper, plastic, carpet, dried noodles, sticky tape)),
Item 13. Response to finding animals in 'messy' things (codes the child's response to finding a plastic animal in 'messy' things),
Item 14. Response to being made 'messy' with lotion by the examiner (codes the child's response to his/her arm being made messy by the examiner),
Item 15. Response to being 'messy' (codes the child's response to his/her hands being messy),
Item 16. Response to standing/sitting close to others,
Item 17. Response to splashing water,
Item 18. Response to finding animal in sand (codes the child's response to being barefoot),
Item 19. Response to putting the socks/shoes back,
Item 20. Over-response to unexpected touch,
Item 21. Under-response to unexpected touch.

The parents were asked to complete the following questionnaires:

1. Sensory Profile (SP; Dunn, 1999) - a caregiver questionnaire that measures a child's sensory processing abilities. The questionnaire consists of 125 items, rated on a five-point Likert scale, ranging from almost never to almost always, including tactile and auditory items (see Chapter 4).

Parents of children with ASD and WS were also completing:

2. Social Responsiveness Scale – Second Edition (SRS-2; Constantino & Gruber, 2012) - a 65-item rating scale which takes 15–20 minutes to complete. It is a parent-report of autistic trait that covers unusual interpersonal behaviours, communication or repetitive/stereotyped behaviours. The SRS-2 describes a degree of autistic social impairment and the severity of autistic symptoms.

In order to ensure that the typically developing children included in the study were not experiencing any emotional, social or behavioural problems, parents of those children were also required to complete:

3. The Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997) – a 25-items caregiver-report of children of 4-16 years old that screens whether the child has any emotional, conduct, hyperactivity/inattention, and peer relationship problems or displays prosocial behaviour. It takes between 5 and 10 minutes to complete the form.

9.2.4 Procedure and materials

Pilots:

First, the tasks were pre-piloted on two typically developing boys, aged 4 and 6 years in order to establish task order and ensure that taking part in the tasks was enjoyable for the children (i.e. task acceptability). Then the tactile and auditory observation schedules were piloted on another two typically developing children (a 4 year old boy and a 9 year old girl) to test the adjusted task order and ensure that children across the age range could engage and show interest in the tasks (feasibility and acceptability). Video recordings were made of these pilot assessments for discussion in the research team. The videos were watched and scored independently by the three experts in order to verify and adjust a scoring schedule. The initial reliability of the schedule was established and consensus regarding the scoring items and system was reached (89.75% percentage agreement).

Parental involvement

After consenting to take part in the study, a brief telephone interview was undertaken with each parent. The parents were asked to provide basic demographic information (such as confirming a date of birth of their child, describing their child's ethnic origin). The parents of children with ASD were also asked to provide information on the age of their child when they received their initial diagnosis. Additional questions related to a possible vision, hearing, movement difficulties, and the use of medication. Moreover, parents were asked whether any tactile items or sounds could provoke anxiety in their child, and if they were named, they were asked to describe the child's possible reactions to those and parental strategies used to calm the child down. They also provided information on their child's interest and favourite activities. Finally, the interviewer enquired regarding any allergies to ensure that none of the materials were going to cause harm to the child. Those parents of children with ASD whose children were to be seen at school were also asked whether they wished to be contacted again with the exact date and time of the assessment to give them an opportunity to inform their child when the assessment would take place. The questionnaire can be found in Appendix V.

Questionnaire packs including an information sheet, the Sensory Profile (Dunn, 1999), Social Responsiveness Scale – Second Edition (Constantino & Gruber, 2012; parents of ASD and WS children only), and the Strengths and Difficulties Questionnaire (Goodman, 1997; parents of TD children only), were sent to parents who had agreed to participate in the study. The parents could either return the questionnaires by post (in the stamped addressed

return envelope that was included in the pack) or hand them in to the examiner during the visit (WS and some of the ASD parents only).

Child involvement

Direct assessments with the children took place either at child's home or at the child's school. All the WS children were visited at home, all the TD children were seen at school, and the ASD children were assessed either at home (n=9) or at school (n=14).
The assessments always began with the examiner introducing herself, explaining what the research was about and asking the child if they were willing to help the examiner by playing pattern, word, touch and hearing related games. Each child was also asked if they agreed for part of the games (TASOS) to be video-recorded. After obtaining a verbal agreement from the child, the Raven's Coloured Progressive Matrices and the British Picture Vocabulary Scale - Third Edition were administered. After the ability assessment, the tactile and auditory sensory observation schedules began. All children agreed to take part in the study.

Apparatus

HP Pavilion dm1 notebook, Samsung Galaxy Tab Pro, HD Camcorder (Panasonic HC-VX870EB-K) and tripod, Sound Meter app installed on a Samsung Galaxy S6 mobile phone were used in the study.

Materials

Auditory domain

A number of short (30 seconds each) sound clips were used across the tasks and played from the laptop. The clips included: a popular radio song - 'Happy' by Pharrell Williams (item 6); refrigerator, people talking, traffic noise (item 7); flushing toilet, vacuum cleaner, hair dryer, dog barking, clock ticking, pencils/pens scratching, candy wrappers, car horns, train, alarm clock, police siren, a balloon popping, fireworks, an aeroplane (item 8 and some of the sound clips (flushing toilet, dog barking, car horns, police siren, aeroplane) also used in item 5); whistle sound (item 10).
All the sound clips were downloaded from YouTube® and converted (shortened) with Audacity® software (version 2.1.1).

Tactile domain

To assess tactile processing, two sets of materials were needed. To administer a task relating to sensitivity to certain fabrics (item 12), 14 wooden cube blocks (2x2x2 inches) and

a cardboard box (without the back side and with the flap cut in the middle of the front side, like a letter box) were needed. Thirteen wooden cubes had a square (2x2 inches) piece of fabric/texture glued on to one side. The fabrics/textures used in the study were as follow: leather, sponge, sandpaper, fluffy towel, lace, t-shirt label, velvet, suede, plastic, carpet, sticky tape, feathers, and artificial grass. The fourteenth cube was kept plain in order to provide a wooden texture for the children. In that task also fourteen laminated photographs of the textured side of the wooden blocks were included. To administer tasks related to 'being messy' (item 13 and 14) five transparent cylinder-shaped food containers were used. Each of the containers was filled up with one of the materials: dry pasta (400g), uncooked rice (800g), sand (1500g), unscented body lotion (800g) and a gooey dough (made of plain flour, salt and water; 1200g). For the tasks that required water and sand exposure (item 17 and 18), two transparent cuboid-shaped food containers were used, filled up with water (1200ml) and sand (2500g) respectively.

Procedure

The tactile and auditory sensory observation schedules had three main parts: a free-play time, auditory and tactile tasks. Games and activities were designed to reflect everyday sensory experiences that might prompt atypical responses in children with both autism and Williams syndrome. Each child's assessment was video-recorded to enable their responses to be coded in a systematic manner. In the free-play time the child was presented with commonly used toys: magnetic blocks, a tea set, a jack in a box, two fluffy and soft mascots (Kermit and Animal), a tambourine, an electronic toy smartphone, dolls, cars, a range of miniature animals, two neon puffer balls and asked to play for 10 minutes. This allowed the examiner to observe and code the extent to which the child explored materials in a sensory way, was seeking auditory and/or tactile stimulation.

The assessment materials selected for this part of observation are presented in Figure 9.1.

Figure 9.1 Materials selected for a free-play time

Additionally, four auditory tasks were administered during the free-play time in a set order: 'Item 5. Response to hearing specific sounds (codes the child's response to a number of unexpected sound presses e.g. flushing toilet, dog barking, car horns, police siren, aeroplane)', 'Item 9. Response to name', 'Item 10. Response to non-social sound – whistle' and 'Item 11. Response to 'special' interest word' in order to obtain reaction to unexpected stimuli. The first auditory task was always administered after the child engaged in play for two minutes. The unexpected sounds (item 5) were played at up to 70 decibels (dB) volume level and the experimenter was always within 2 metres distance from a child. The whistle sound was played at three different volume levels: around 45, 60 and 75 (dB).

After the free-play time, each child participated in two groups of semi-structured tasks assessing hypo- and hyper-auditory and tactile processing. First, the children were asked to play one or more self-selected games on a Tablet. They could choose from: Animal Farm for Kids, Coloring Princess, Crossy Road, Cut the Rope, Hill Climb Racing, Minecraft, Peppa Paintbox, Super Puzzle, Temple Run. The Tablet itself and all the games were muted.
To assess hyper-responsivity to auditory stimulation, when a child started playing a selected game, without cueing the child the experimenter played first a radio song and then the refrigerator, people talking and traffic noise sound clips. Each of the four sound clips were played three times, at different volume levels, around 45, 60 and 75 dB (as perceived by a

child), for 30s each time, with no break between the stimuli, with the examiner sitting next to the child.

A further task assessing hyper-responsivity to auditory stimulation involved exposing a child to background noise when engaged in an activity task. The child was asked to listen to fourteen brief sound clips and identify the sounds (e.g. dog barking, police siren, balloon popping, clock ticking) by either naming them or pointing to pictures presented to him/her. Only pictures of the sounds used were included on the answer sheet (see Figure 9.2). The presentation of a sound clip was ended when the child provided an answer (either verbal or non-verbal) and the following sound clip was introduced. Each sound was played twice at around 50 dB and up to 70 dB. The child was not given any feedback.

Figure 9.2 The answer sheet used in the sound recognition task

In the assessment of hypo- and hyper-responsivity to tactile stimulation, the child was first introduced to 'the magic box' game to measure child's sensitivity to certain fabrics. It was explained that there was a box with a mobile flap. The rules of the game were that there two pictures of the stimuli (textured wooden cube blocks) would be presented to the child, yet only one of them was hidden in the box. The child had to first look at the pictures, then place a hand in the box, and after touching the texture, match the stimulus in the box with one of the pictures. The same procedure was repeated fourteen times, to administer all stimuli during the task. Feedback was given to the child after each trial. The order was randomised for each

child. After completing the game, children were asked to help the examiner to tidy up and put all the wooden blocks away onto the cover of the magic box.

The next game was then introduced. Five containers were always placed on a table and a child was told they will be asked to find a surprise (small plastic animal) hidden in each of the containers. Before starting the game, the examiner sat very close to the child to examine his/her response to sitting next to others and to assist him/her to roll their sleeves up by touching both forearms to assess child's response to unexpected touch. If a child had a short-sleeve top on, the examiner would still touch both child's forearms (justifying it as making sure that child was ready to take part in the game). The task would start by presenting a child with containers filled with pasta, rice, and sand. During this part of the task, unexpectedly, the child's face would also gently be splashed with some water. Children were offered a towel to dry their face. After finding surprises in the first three containers, the examiner opened the container with body lotion, getting some on their index finger and told the child that she was going to show him/her how this one feels before putting his/her hand in the container in order to find another surprise. Body lotion was then spread around child's wrist to assess child's hypo-responsivity to tactile stimulation. Children could use a towel to wipe lotion off if they wanted to remove it.

Next children were asked to look for a hidden plastic animal toy in lotion and gooey dough. If they wanted, they could wash their hands in a container filled with water and/or use a towel. Finally, each child was asked to take their shoes and socks off and find one more surprise in a cuboid-shaped container filled with sand using their feet. After the task, the examiner would wipe the sand off their feet and help the children to put their socks and shoes back on. If a child was wearing tights, this game was not administered. All children received a certificate of achievement after completion of taking part in the study. The tasks were usually completed within 45-60 minutes.

9.2.5 Scoring procedure

The tactile and auditory sensory observation schedule is composed of two main domains, which reflect tactile and auditory sensory systems. Each of the domains is composed of games and activities assessing either hypo- or hyper-responsivity to tactile or auditory stimulation which were individually scored for response. Scoring took place after game administration as the assessments were video-recorded. Two coders were involved in scoring the videos – the main coder (the experimenter) rated all the videos, while the second independent coder (an undergraduate final year psychology student) scored 10% of the videos

(randomly selected across the three groups, gender and time of assessment as data collection took place over 6 months).

Each domain had multiple games and activities that were administered in a standardised manner to the child participants. Within the domains, the coders used standard criteria to score the responses in a consistent manner. Items within each domain were scored on a three point scale ranging from 0 (typical, expected response), 1 (some atypicality) to 2 (atypical, unexpected response). However, item 10 in the auditory domain was scored in a binary manner, with 0 relating to the observed response and 2 relating to the lack of response. Detailed definitions for each item are included in the scoring sheet (see Appendix W).

9.2.6 Data analysis

SPSS 22 was used to analyse the data. The inter-rater agreement was calculated to establish the degree of agreement among raters. To investigate group differences ANOVAwas conducted. Regression analysis was performed to quantify overlap between auditory and tactile scores obtained from both measures. Item distribution was used to identify the most discriminative tactile and auditory sensory processing tasks for all participants, and for the ASD and WS samples. Cramer's V were calculated as a measure of effect sizes, with >.01 indicating small, >.03 medium and >.5 large effect size (Field, 2009).

Inter-rater reliability

A proportion of the video-recordings of the auditory and tactile assessment (10%) were evaluated by an independent rater. The inter-rater reliability between the examiner's and independent rater's scorings calculated as percentage agreement on all items (including sub-items such as responding to individual sounds in item 5) was 88% and on the main 21 items was 85%.

9.3 Results

9.3.1 Feasibility and acceptability

All children completed all the auditory tasks. Four TD children and one ASD child were unable to complete 'Response to finding an animal in sand' and 'Response to putting the socks/shoes back on' tasks (item 18 and item 19) due to wearing tights. One child with WS and further five children with ASD were not able to complete some of the tasks from the tactile domain due to difficulties with understanding the instructions. None of the children requested to stop the assessment. These data show that the observation schedule is feasible

for use with verbal children, both typically and atypically developing. Also, minimally verbal and non-verbal children with ASD and WS (11 ASD and 9 WS respectively) were able to complete the tasks. Six children, however, were not able to complete the tasks due to their very low IQ status.

9.3.2 Group differences between typical, ASD and WS samples on the auditory and tactile processing subscales of the SP and the TASOS

The analyses were performed on all ASD and WS participants, and on eligible TD children. The descriptive statistics for three groups can be found in Table 9.1.

Table 9.1 Descriptive statistics (mean (SD)) of participants' characteristics and TASOS scores

	TD (n=20)	WS (n=17)	ASD (n=23)
Gender: male	11	9	19
Age in months	77.20 (7.77)	84.65 (21.93)	84.09 (20.72)
Verbal IQ	91.95 (13.99)	85.38 (8.67)	89.58 (12.99)
Non-verbal IQ	100.25 (14.00)	78.75 (6.29)	96.25 (11.51)
TASOS scores			
Tactile Total Score	5.74 (1.94)	6.06 (1.73)	7.17 (2.90)
Hypo-responsiveness	4.42 (2.12)	3.00 (1.79)	4.61 (1.88)
Hyper-responsiveness	1.32 (1.97)	3.06 (1.69)	2.56 (2.06)
Auditory Total Score	2.73 (1.35)	5.18 (1.81)	4.92 (2.28)
Hypo-responsiveness	1.95 (1.21)	2.00 (.87)	2.96 (1.88)
Hyper-responsiveness	.74 (1.10)	3.18 (1.78)	1.96 (1.55)

Note: in the TD sample verbal IQ data were available for 19 participants and non-verbal IQ data for all participants; due to low ability level in the WS sample verbal IQ data were collected only for 4 participants and non-verbal IQ data were available for 8 children; in the ASD sample verbal and non-verbal IQ were assessed in 12 participants. TASOS Tactile Total Score range: 0-26, TASOS Auditory Total Score range: 0-16. Higher TASOS scores indicate more atypicality.

The groups did not significantly differ on age $F_{(2,63)}=.62$, $p=.541$, but did significantly differ on ability $F_{(2,63)}=8.55$, $p=.001$ for non-verbal and $F_{(2,63)}=7.17$, $p=.002$ for verbal ability, with TD group being significantly different to both ASD and WS groups as indicated by the Bonferroni post-hoc test. ASD and WS groups did not significantly differ on any of the variables (for age and verbal ability $p=1.0$, for non-verbal ability $p=.072$).

Sensory Questionnaire: Sensory Profile (SP)

Using ANOVA with group as a fixed factor and SP auditory and tactile processing scores as the dependent variable, it was found that there was a significant effect of group on both auditory ($F_{(2,45)}$=16.356, p<.001) and tactile ($F_{(2,47)}$=6.229, p=.004) processing scores. Levene's test showed that the variances were equal for the groups for both auditory ($F_{(2,45)}$=.250, p=.780) and tactile ($F_{(2,47)}$=2.674, p=.079) scores. Post-hoc analysis showed that children with ASD and WS had significantly lower auditory processing scores (p<.001) and tactile processing scores than TD children (TD and ASD, p=.005; TD and WS, p=.016) as reported by parents. Tactile and auditory scores were not significantly different for ASD and WS groups (p=1.0) The SP manual classifies children as having 'definite difference' in auditory processing if the scores fall between 8 and 25; and in tactile processing if the scores fall between 18 and 64. For both auditory and tactile processing only 1 child in the TD group had the scores within the 'definite difference' range (7.1%). Atypical auditory processing was reported in 72.2% of children with WS and 82.4% children with ASD, while scores on tactile processing within 'definite difference' range were characteristic for 52.9% of children with WS and 52.5% of ASD children. Table 9.2 presents the summary of the SP scores in three groups.

Table 9.2 Mean (SD) values for the Sensory Profile auditory and tactile processing scores for the three groups

SP variable	TD	WS	ASD
n	14	17	17
Auditory processing	31.21 (4.92)	21.88 (5.61)	20.35 (6.12)
Auditory 'definite difference'	7.1%	72.2%	82.4%
n	14	17	19
Tactile processing	78.14 (10.15)	64.94 (9.70)	63.37 (16.20)
Tactile 'definite difference'	7.1%	52.9%	52.6%

Note: lower mean scores on auditory and tactile processing indicate more atypicalities

9.3.3 Correlation and regression analysis, convergent validity of the TASOS

Correlation analysis showed that for the whole sample the auditory processing score from the SP was significantly associated with gender (r=.358, p=.014), also non-verbal IQ score was negatively correlated with the auditory domain as measured by the TASOS (r=-.389, p=.023), indicating that higher scores on the auditory processing domain (greater atypicality) were associated with lower non-verbal intellectual ability.

At the group level, in the TD sample, the auditory processing domain score as measured by the TASOS was significantly associated with gender (r=.582, p=.009) with girls showing more auditory processing atypicalities than boys, as indicated by a t-test (t(20)=-2.74, p=.013). In the WS sample, the SP auditory processing domain score was significantly negatively associated with non-verbal IQ score (r=-.969, p=.031), demonstrating that the lower non-verbal intellectual ability, the greater auditory processing atypicality was present in the WS participants.

Two linear regression analyses with the SP auditory and tactile scores as the dependent variable and the auditory and tactile processing domain scores as the independent variable were conducted to examine the relationship between two measures. For the total sample, scores on the auditory processing domain significantly predicted auditory scores on the SP (R=.43, R^2=.19, $F_{(1,45)}$=10.194, p=.003), however, the tactile processing domain scores did not significantly predicted tactile scores on the SP (R=.08, R^2=.01, $F_{(1,41)}$=.249, p=.620). When investigating group level data, tactile processing domain scores significantly predicted tactile scores on the SP in the WS group (R=.58, R^2=.34, $F_{(1,14)}$=6.552, p=.024) only.

9.3.4 Score distribution in the observation schedule (discriminative items between the typical and neurodevelopmental samples, and between ASD and WS samples)

A Chi-squared test was performed to examine whether there was a difference in the distribution of the TASOS scores for both auditory and tactile processing domain, across three groups. Some of the expected frequencies were lower than 1, hence a 2-tailed Fisher's exact test was calculated. A significant difference in the distribution of the scores was found in the auditory domain (p=.001, Fisher's exact test), but not in the tactile domain (p=.203, Fisher's exact test).

The Fisher's exact test was then calculated for TD and ASD, TD and WS, and ASD and WS groups separately. For the TD and ASD groups and TD and WS groups, a significant difference in the distribution of the scores was found in the auditory domain (p=.006; p=.007, Fisher's exact test respectively), but not in the tactile domain (p=.589; p=.196, Fisher's exact test respectively). The Fisher's exact test was also performed for the ASD and WS groups to examine whether the auditory or tactile processing domain scores as measured by the TASOS were similarly distributed in the neurodevelopmental groups. Fisher's exact tests were non-significant (for auditory: p=.082; for tactile: p=.123), showing that distribution of the auditory and tactile processing scores were similar in these groups.

The distribution analysis was also run for the auditory and tactile hypo- and hyper-responsivity subscales. Auditory hyper-responsivity scores were significantly differently

distributed between all three groups (p=.002, Fisher's exact test) and between TD group and neurodevelopmental groups (ASD: p=.014, Fisher's exact test; WS: p>.001, Fisher's exact test). Also tactile hyper-responsivity scores were significantly differently distributed between TD and WS groups (p=.031, Fisher's exact test). None of the remaining distribution analyses were significant.

9.3.5 Item distribution analysis and item reduction

An item distribution analysis was conducted for the tactile and auditory sensory observation schedule. The score distribution data are presented in Table 9.3 and Table 9.4. The analysis was performed four times, once for all three groups, and then again for TD and WS, TD and ASD, and ASD and WS groups only to identify items that best distinguish between the groups.

Table 9.3 Contingency table for auditory processing domain items

		TD (n=20)			WS (n=17)			ASD (n=23)		
	Item #	0	1	2	0	1	2	0	1	2
hypo	Item 1. Sound seeking	9	3	8	1	4	12	1	2	20
	Item 9. Response to name	20	0	0	16	0	1	15	4	4
	Item 10. Response to non-social sound – whistle	12	0	8	15	0	2	21	0	2
	Item 11. Response to 'special' interest word	19	0	1	17	0	0	18	1	4
hyper	Item 5. Response to hearing specific sounds	19	1	0	10	4	3	16	2	5
	Item 6. Response to radio while playing on the Tablet	17	3	0	4	13	0	14	7	2
	Item 7. Response to background noise (refrigerator, people talking, traffic noise) while playing on the Tablet	19	1	0	5	11	1	11	10	2
	Item 8. Response to hearing specific sounds and matching the sound	16	1	3	6	4	7	17	2	4

Table 9.4. Contingency table for tactile processing domain items

		TD (n=20)			WS (n=17)			ASD (n=23)		
	Item #	0	1	2	0	1	2	0	1	2
hypo	Item 2. Need for touching certain fabrics	6	8	6	3	4	10	8	4	11
	Item 3. Touching objects and others	18	2	0	15	2	0	12	9	2
	Item 4. Child's need to be touched	13	6	1	13	3	1	13	6	4
	Item 14. Response to being made 'messy' with lotion by the examiner	7	2	11	12	5	0	14	2	4
	Item 15. Response to being 'messy'	4	7	9	15	2	0	13	6	0
	Item 19. Response to putting the socks/shoes back on	16	0	0	10	2	4	10	2	6
	Item 21. Under-response to unexpected touch	19	0	1	17	0	0	20	0	1
hyper	Item 12. Sensitivity to certain fabrics	19	1	0	16	1	0	13	5	0
	Item 13. Response to finding an animal in 'messy' things	15	2	3	2	3	11	5	6	8
	Item 16. Response to standing/sitting close to others	18	1	1	17	0	0	21	0	1
	Item 17. Response to splashing water	15	3	1	7	7	3	12	6	1
	Item 18. Response to finding an animal in sand	10	4	2	11	1	4	13	0	4
	Item 20. Over-response to unexpected touch	20	0	0	17	0	0	19	1	1

Note: Due to the inability of all child participants to undertake all the tasks, for some of the items the totals of all three coding categories (0, 1, 2) do not add up to the indicated n

As indicated by the analysis, from the auditory processing domain all items discriminated between the three groups except item 11 (response to special interest word); whereas in the tactile processing domain only items 3, 13, 14, 15 and 19 discriminated between the three groups (see Table 9.5).

Table 9.5 Fisher's exact test and effect sizes for all three groups

Item #	Fisher's exact test p value	Cramer's V
Auditory processing domain		
Item 1. Sound seeking	**.002**	**.373**
Item 9. Response to name	**.007**	**.319**
Item 10. Response to non-social sound – whistle	**.037**	**.355**
Item 11. Response to 'special' interest word	.183	.225
Item 5. Response to hearing specific sounds	**.046**	**.272**
Item 6. Response to radio while playing on the Tablet	**<.001**	**.397**
Item 7. Response to background noise (refrigerator, people talking, traffic noise) while playing on the Tablet	**<.001**	**.393**
Item 8. Response to hearing specific sounds and matching the sound	**.051**	**.282**
Tactile processing domain		
Item 2. Need for touching certain fabrics	.495	.171
Item 3. Touching objects and others	**.023**	**.304**
Item 4. Child's need to be touched	.606	.164
Item 14. Response to being made 'messy' with lotion by the examiner	**.001**	**.382**
Item 15. Response to being 'messy'	**<.001**	**.486**
Item 19. Response to putting the socks/shoes back on	**.020**	**.309**
Item 21. Under-response to unexpected touch	1	.122
Item 12. Sensitivity to certain fabrics	.118	.315
Item 13. Response to finding an animal in 'messy' things	**<.001**	**.421**
Item 16. Response to standing/sitting close to others	.905	.156
Item 17. Response to splashing water	.181	.236
Item 18. Response to finding an animal in sand	.231	.255
Item 20. Over-response to unexpected touch	1	.177

Note: Significant and approaching significance items are in bold

Similarly, the distribution analysis was repeated for the ASD and WS groups only. For the auditory domain the analysis indicated that only item 6 discriminated between the groups; however, item 8 approached significance (Table 9.6). In the tactile domain no items significantly discriminated between the groups, Item 3 ‘touching objects and other’s was approaching significance.

Table 9.6 Fisher's exact test and effect sizes for ASD and WS groups

Item #	Fisher's exact test p value	Cramer's V
Auditory processing domain		
Item 1. Sound seeking	.406	.213
Item 9. Response to name	.102	.355
Item 10. Response to non-social sound – whistle	1	.051
Item 11. Response to 'special' interest word	.123	.325
Item 5. Response to hearing specific sounds	.575	.206
Item 6. Response to radio while playing on the Tablet	**.011**	**.465**
Item 7. Response to background noise (refrigerator, people talking, traffic noise) while playing on the Tablet	.544	.210
Item 8. Response to hearing specific sounds and matching the sound	**.054**	**.387**
Tactile processing domain		
Item 2. Need for touching certain fabrics	.834	.126
Item 3. Touching objects and others	**.050**	**.304**
Item 4. Child's need to be touched	.451	.220
Item 14. Response to being made 'messy' with lotion by the examiner	.094	.376
Item 15. Response to being 'messy'	.236	.238
Item 19. Response to putting the socks/shoes back on	.885	.091
Item 21. Under-response to unexpected touch	1	.148
Item 12. Sensitivity to certain fabrics	.177	.290
Item 13. Response to finding an animal in 'messy' things	.262	.288
Item 16. Response to standing/sitting close to others	1	.143
Item 17. Response to splashing water	.317	.252
Item 18. Response to finding an animal in sand	.838	.186
Item 20. Over-response to unexpected touch	1	.212

Note: Significant and approaching significance items are in bold

Fisher's exact test was also calculated for auditory and tactile observation schedule items for the TD and WS groups. Item 1, item 5, item 6, item 7, item 8 from the auditory domain had a significantly different distribution between the groups, while in the tactile domain critical items were item 13, item 14, item 15, and item 19 (Table 9.7)

Table 9.7 Fisher's exact test and effect sizes for TD and WS groups

Item #	Fisher's exact test p value	Cramer's V
Auditory processing domain		
Item 1. Sound seeking	**.029**	**.439**
Item 9. Response to name	.459	.181
Item 10. Response to non-social sound – whistle	.073	.317
Item 11. Response to 'special' interest word	1	.160
Item 5. Response to hearing specific sounds	**.022**	**.447**
Item 6. Response to radio while playing on the Tablet	**<.001**	**.618**
Item 7. Response to background noise (refrigerator, people talking, traffic noise) while playing on the Tablet	**<.001**	**.685**
Item 8. Response to hearing specific sounds and matching the sound	**.022**	**.458**
Tactile processing domain		
Item 2. Need for touching certain fabrics	.242	.290
Item 3. Touching objects and others	1	.028
Item 4. Child's need to be touched	.725	.143
Item 14. Response to being made 'messy' with lotion by the examiner	**<.001**	**.603**
Item 15. Response to being 'messy'	**<.001**	**.698**
Item 19. Response to putting the socks/shoes back on	**.018**	**.480**
Item 21. Under-response to unexpected touch	1	.154
Item 12. Sensitivity to certain fabrics	1	.019
Item 13. Response to finding an animal in 'messy' things	**<.001**	**.645**
Item 16. Response to standing/sitting close to others	1	.220
Item 17. Response to splashing water	.083	.388
Item 18. Response to finding an animal in sand	.345	.280
Item 20. Over-response to unexpected touch	NC	NC

Note: NC – not computed as a constant; significant and approaching significance items are in bold

The distribution analysis performed for the TD and ASD groups indicated that item 1, item 5, item 7, and item 9 from the auditory domain had a significantly different distribution between

the groups, while in the tactile domain item 3, item 13, item 15 and item 19 had a significantly different distribution for these groups (see Table 9.8).

Table 9.8 Fisher's exact test and effect sizes for TD and ASD groups

Item #	Fisher's exact test p value	Cramer's V
Auditory processing domain		
Item 1. Sound seeking	**.001**	**.526**
Item 9. Response to name	**.013**	**.430**
Item 10. Response to non-social sound – whistle	.078	.316
Item 11. Response to 'special' interest word	.363	.230
Item 5. Response to hearing specific sounds	**.045**	**.341**
Item 6. Response to radio while playing on the Tablet	.251	.278
Item 7. Response to background noise (refrigerator, people talking, traffic noise) while playing on the Tablet	**.002**	**.492**
Item 8. Response to hearing specific sounds and matching the sound	1	.073
Tactile processing domain		
Item 2. Need for touching certain fabrics	.369	.216
Item 3. Touching objects and others	**.013**	**.436**
Item 4. Child's need to be touched	.557	.186
Item 14. Response to being made 'messy' with lotion by the examiner	.067	.375
Item 15. Response to being 'messy'	**<.001**	**.587**
Item 19. Response to putting the socks/shoes back on	**.008**	**.500**
Item 21. Under-response to unexpected touch	1	.011
Item 12. Sensitivity to certain fabrics	.091	.295
Item 13. Response to finding an animal in 'messy' things	**.009**	**.500**
Item 16. Response to standing/sitting close to others	.727	.167
Item 17. Response to splashing water	.366	.221
Item 18. Response to finding an animal in sand	.089	.392
Item 20. Over-response to unexpected touch	1	.213

Note: Significant and approaching significance items are in bold

9.4 Discussion

This study aimed to develop a new tactile and auditory sensory observation schedule (TASOS) that would facilitate the assessment of auditory and tactile hypo- and hyper-reactivity in typical and atypical development. All children completed all the auditory tasks. One child with WS and further five children with ASD were not able to complete some of the tasks from the tactile domain due to low ability level. The study showed that the observation schedule is feasible for use with verbal children, both typically and atypically developing. Also, it is the first study that included minimally verbal and non-verbal children with ASD and WS (11 ASD and 9 WS respectively). The feasibility and acceptability of the measure looks promising in this group of less able children, as the majority of children completed all the tasks, with 100% of children completing auditory processing tasks, 94% of WS and 78% of ASD children completing tactile processing tasks.

As indicated by the results, on the subscale level, auditory domain scores were distributed significantly differently across three groups, distinguishing between the TD sample and both neurodevelopmental samples, yet the ASD and WS group scores were distributed similarly. Also, the scores were distributed equally for all the groups in the tactile domain. However, the SP auditory and tactile processing scores indicated significant group differences between TD and both neurodevelopmental samples, showing greater difficulties in sensory processing in ASD and WS groups. The study, hence, confirmed differences for children with ASD on the SP (e.g. Kientz & Dunn, 1997; Kern et al., 2006) as compared to TD children. On the SP 82.4% of ASD participants fell into the category of 'definite difference' on the auditory processing subscale and 52.6% on the tactile processing subscale. This was the first study to examine sensory processing differences in WS children using the SP. On the parent-questionnaire 72.2% of children with WS showed a 'definite difference' in auditory processing and 52.9% in the tactile processing. The prevalence of sensory processing difficulties in those two domains was hence very similar to the ASD children and consistent with the previous study showing definite abnormalities on the Auditory Filtering domain as measured by the Short Sensory Profile in 59% of the WS children (John & Mervis, 2010).

Regarding hypo- and hyper-responsivity, the distribution analysis indicated that for auditory and tactile modalities some of the hypo- and hyper-responsivity scores were significantly differently distributed between all three groups and between the TD group and neurodevelopmental groups, with medium to large effect sizes. These findings are surprising, especially in relation to our ASD sample, as it has been claimed that features associated with the hyporesponsiveness pattern in both social and non-social contexts can discriminate

between children diagnosed with autism, developmental delays and those typically developing (Baranek et al., 2006), but not the features associated with the hyper-responsiveness pattern. However, Baranek et al. (2006) relied in their study on the Sensory Experiences Questionnaire, which is a parent report and assessed children between 5 months and 6 years old. It has been reported that in autism sensory hyper-responsivity increases up to age of 9 while a non-consistent pattern of chronological age related changes was found for hypo-responsivity (Ben-Sasson et al., 2009). It is therefore possible that hyper-responsive behaviours are more prevalent in young children, however we found that only the auditory hyper-responsivity subscale distinguished between ASD and TD children. Although no analysis of developmental changes in sensory processing is available for WS, hyperacusis seems to be most severe in young childhood, at age 5.7 +/- 3.8 years (Gothelf et al., 2006). More research is needed investigating hypo- and hyper-responsivity patterns across modalities in both ASD and WS, especially in terms of distinguishing these two disorders from each other.

Using item distribution analysis, items related to sound seeking and a range of background noise in the auditory domain (item 1, item 6, item 7) and those related to being messy or being made messy (item 13, item 14, item 15) in the tactile domain were best at distinguishing between all the three groups and between TD and WS groups. Most of these items were also implicated when differentiating between TD and ASD participants. The administration of these six tasks takes up to 25 minutes. Rather than using all the tasks, only these selected tasks could be used when assessing auditory and tactile hypo- and hyper-responsivity in children with typical and atypical development. Future replication studies are required to further validate these findings.

Only one item (Item 6. Response to radio while playing on the Tablet) distinguished between ASD and WS children clearly, with more ASD children not noticing the sound compared to the WS children, who were more likely to react to it. A further two items approached significance (Item 8. Response to hearing specific sounds and matching the sound and Item 3. Touching objects and others), with all three items having a medium effect size. The groups, in general, performed similarly, in line with sensory hypo- and hyper-reactivity included as one of the diagnostic features of autism spectrum disorder (DSM-V, APA, 2013), and WS literature in which sensory processing abnormalities were proposed as a fundamental characteristic of the disorder (John & Mervis, 2010). Interestingly, however, on the two hyper-responsivity auditory items that were distributed differently in both groups, WS children showed more atypicalities compared to the ASD children. The opposite was found for the hypo-responsivity tactile item that distinguished between the groups, as

children with ASD showed more tactile seeking behaviours than children with WS. These findings support previous research on ASD and WS. Gallo et al. (2008) and Levitin and colleagues (2005), compared individuals with WS to those with other neurodevelopmental disorders, including children with autism. The authors reported the overwhelming prevalence of hyperacusis and unusual auditory responses to everyday sounds in children and young people with WS and claimed that these features distinguished individuals with WS from not only typically developing individuals but also from those with other neurodevelopmental disorders, including autism. Prior research demonstrated that the hypo-responsivity pattern best differentiates ASD from other developmental disabilities (Baranek et al., 2006; Watts et al 2016), yet we found only one hypo-responsive tactile item that distinguished between ASD and WS groups. In our sample we included children with ASD who were low functioning. Foss-Feig et al. (2012) reported tactile seeking and hypo-responsivity behaviours as associated with greater levels of social impairment, repetitive behaviours and non-verbal communication impairment. Hence, it is likely, that non-verbal ASD children are particularly similar to those with WS in regards to sensory behaviours. Further work is needed to establish whether oversensitivity to sounds and tactile hypo-responsivity are specific to one of these conditions or whether they are a common feature present across different neurodevelopmental disorders.

9.4.1 Limitations

The current study has some limitations. The sample size of each group was relatively small. Despite this, we identified auditory and tactile differences between TD, WS and ASD samples on the SP and on the TASOS indicating some differences between both neurodevelopmental groups. The unequal gender ratio may also have influenced our findings. We found using the TASOS that in the TD sample girls showed more auditory processing atypicalities than boys. This finding is consistent with previous research suggesting that females present more sensory atypicalities than males (Goldsmith et al., 2006). It is unclear whether a similar pattern can be found in individuals with ASD. Lai et al. (2011) reported that females with ASD have more sensory issues than males while Lane et al. (2014) did not find any associations between gender and sensory subtypes in their ASD sample. Further research is needed to investigate the role of gender in the presentation of unusual sensory responses in both ASD and WS. Furthermore, a relationship between the tactile processing domain on the TASOS and the tactile processing subscale on the SP was not found. It is possible that tactile behaviours are more difficult to be noticed by the parents than auditory behaviours. Moreover, although the tasks aimed to assess only single modality (either

auditory or tactile) at the time, a cross-modal transfer possibly occurred, particularly in item 6 and item 12 a visual component might have played an additional role . As previously reported, integration of multisensory information is impaired in individuals with ASD (Stevenson et al., 2014), and that in turn could impact individual performance on the tasks. Further work is required on the validation of the TASOS and its psychometric properties to provide researchers and clinicians with an accurate measure of auditory and tactile processing in children with and without neurodevelopmental disorder.

9.4.2 Conclusion

This is the first study to develop a test of auditory and tactile sensory observation schedule and explore its use in samples of TD children and those with ASD and WS. It is the first study using direct assessments in evaluating auditory and tactile processing in individuals with WS. Feasibility and acceptability of the measure were promising. In summary, we found that similar high proportions of children with ASD and WS presented with both auditory and tactile unusual sensory responses to everyday stimuli as measured by the TASOS and by parent-report using the SP. Six items best discriminated between TD children and those with neurodevelopmental conditions. Future research should focus on further investigating sensitivity of these items to assess the presence of sensory processing difficulties in children. Three items could differentiate between ASD and WS children. More work is needed to evaluate whether some of the sensory responses are specific to ASD or WS; or whether they are a common feature present across different neurodevelopmental disorders.

Additional research is also required to establish a consensus on a set multiple informant measures of sensory processing that could be used across disciplines and settings.

Chapter 10. General Discussion

10.1 Overview

The studies presented in this thesis investigated sensory profiles of children and adolescents with a diagnosis of ASD and their parents, as well as children with a diagnosis of WS. This chapter will begin with a very brief summary of what was included in each chapter in the thesis and will then move on to synthesise the evidence across chapters and discuss what we can learn from these findings. It will then continue with an exploration of the strengths and weaknesses of the current work and conclude with an exploration of what remains to be done and the implications for future clinical and research endeavours.

The thesis focuses on exploring the phenomenology and impact that sensory processing difficulties have on children with autism spectrum disorder and Williams syndrome. A general introduction to both disorders and sensory processing can be found in Chapter 1. Dunn's model of sensory processing (1997) served as a theoretical stand point in all of the studies undertaken. In that approach, hyper- and hyporesponsiveness to sensory stimulation are divided according to the presence of passive and active self-responding strategies used to respond to sensory stimulation by an individual. As a result, four patterns of sensory processing are described: Low Registration, Sensation Seeking, Sensory Sensitivity and Sensation Avoiding. All four sensory processing patterns have been reported to be present in individuals with ASD (Kern et al., 2007), though there has been scant examination of these patterns in those with WS. A systematic review presented in Chapter 2 evaluated evidence of the psychological correlates of sensory processing patterns in individuals with ASD. The findings of that evaluation suggested that sensory hyporesponsiveness was correlated with core features of ASD, while social awareness difficulties and affective disorders were associated with hyperresponsiveness. Equivocal evidence was found for the associations between sensory processing patterns and repetitive behaviours. A systematic evaluation of these correlates could not be undertaken on the Williams syndrome literature due to the small number of studies available. As revealed in the mixed-methods review on sensory processing in WS (Chapter 3), the majority of the papers explored and discussed the phenomenon of hyperacusis in the disorder, while only three papers investigated sensory processing in general, highlighting the need to conduct more empirical work in this under-researched area.

As far as we are aware, Chapter 4 presents the first investigation of changes in sensory symptoms across different age groups in children with WS. Direct comparison of ASD and WS groups revealed that children with ASD and WS have very similar sensory

profiles that are distinct from those present in typically developing children that remain a persistent, life-long characteristic of both disorders.

The results were further developed in the empirical work reported in Chapter 5 which investigated the sensory and social responsiveness profiles of three groups of children and adolescents with a neurodevelopmental disorder, those with a diagnosis of ASD without learning disability (LD), ASD with additional learning disability or WS.

The comparison of the factorial validity of the Spence Children's Anxiety Scale-Parent version (SCAS-P; Spence, 1998) in a sample of young people with ASD and a sample of typically developing young people with anxiety disorders was undertaken in Chapter 6 to examine whether cross-group comparisons between ASD and anxious samples based on the SCAS-P were appropriate.

Further exploration of the sensory processing profiles in both ASD and WS was reported in Chapter 7. First, sensory processing clusters of children with ASD or WS were examined and similarities between the groups were highlighted. Also the relationships between sensory processing and repetitive behaviours were reported and the role of intolerance of uncertainty and anxiety in mediating between sensory atypicalities and repetitive behaviours was described.

Also, in this thesis, sensory processing related familiar factors were investigated and the first examination of profiles of sensory processing in child-parent dyads within ASD families in comparison to TD dyads was reported in Chapter 8. Although sensory profiles were found similar in parent-child dyads across both groups, children with ASD and their parents shared more sensory avoidant behaviours, and auditory, visual and vestibular sensory processing atypicalities compared to TD dyads. The role of genetic and environmental factors in the inter-generational transmission of sensory atypicalities was discussed.

The work reported thus far has relied solely on data provided by parent questionnaire. In order to obtain observational data a novel direct assessment of hypo- and hyper-responsiveness of auditory and tactile sensory processing modalities was developed (TASOS) and implemented and evaluated with young children with ASD, WS and those typically developing and described in Chapter 9. The development and preliminary evaluation of this new observational measure of auditory and tactile processing was reported and some indications in regards to most discriminant tasks and further simplifying of the tool were made.

10.2 Synthesising the evidence: A Comparison of sensory profiles in individuals with ASD and Williams syndrome

The presence of sensory difficulties in ASD was included in the very first descriptions of the disorder (Asperger, 1944/1991; Kanner, 1943). More recent reports of over 90% of children (Leekam et al., 2007) and adults (Crane et al., 2009) with autism to have extreme levels of sensory processing, including hypersensitivity, hyposensitivity and sensory seeking (Ben-Sasson et al., 2009; Leekam et al., 2007; Rogers & Ozonoff, 2005; Tomchek & Dunn, 2007) led to sensory processing difficulties being recognised as a diagnostic criterion of the disorder (DSM-V, 2013). Sensory problems, however, are common among individuals with neurodevelopmental disorders, including attention deficit hyperactivity disorder (ADHD), Fragile X syndrome and WS (Ermer & Dunn, 1998; Rogers et al., 2003). Interestingly, a claim that sensory processing abnormalities in general are a fundamental characteristic of WS was made as sensory atypicalities were found in 90% of children with WS (John & Mervis, 2010). Although it has been reported that features associated with the hyporesponsivness pattern can discriminate between children diagnosed with autism, developmental delays and those typically developing (Baranek et al., 2006), support for that claim has not been found in our studies. Two comparisons of sensory profiles on WS and ASD were undertaken. In the first, children and adolescents with WS were compared to children with ASD with or without additional learning disabilities (Chapter 4). In the second, young children between 4 and 9 years old with either diagnosis of ASD or WS were recruited (Chapter 7). The samples in these studies consisted of different participants and thus the findings are not a consequence of the same sample being included in each study. Interestingly, it was found, that children with WS had significantly lower Low Registration scores that constitute the hyporesponsivness dimension compared to children diagnosed with ASD, regardless of their learning disability status, contradicting Baranek et al. (2006) findings. In general, the sensory profiles of children in both neurodevelopmental groups were very similar.

Similarities in sensory profiles in ASD and WS were also evident when clusters of sensory processing patterns were compared (Chapter 7). The findings showed the same pattern emerging in both ASD and WS with two distinct clusters present: one cluster with more typical presentation of sensory processing features and the other one with more abnormal presentation of sensory characteristics. These data suggest that children with WS show as much or even more sensory atypicalities than children with autism. Sensory dysfunction to a similar or greater level in individuals with Fragile X syndrome and those who are deaf-blind compared to children with ASD has been previously reported (Rogers & Ozonoff, 2005), suggesting that sensory processing difficulties might be a more global

characteristic of individuals with developmental disorders rather than syndrome-specific feature.

10.3 Synthesising the evidence: A Comparison of sensory profiles in individuals with ASD and WS across age groups

We followed our sensory profile comparisons in both ASD and WS with further investigation of sensory difficulties across age groups in both disorders, aiming to determine any possible developmental changes within sensory processing trajectories (Chapter 4). From the available ASD literature an unclear picture of the relationship between sensory symptoms and age emerges, with some studies reporting the frequency of the majority of symptoms decreasing over time (Kern et al., 2007), whilst others suggest an increase in sensory sensitivities with age (Talay-Ongan & Wood, 2000), and some reporting no relationship between chronological age and the severity of sensory difficulties (Adamson et al., 2006). To our knowledge, the investigation of intensity or frequency in changes in sensory symptoms in WS across age groups has not previously been undertaken. Examination of the sensory profiles in ASD and WS in three group categories (under 6.5 years old, between 6.5 and 9.5 years old, above 9.5 years old) was carried out. The findings indicated that sensory symptoms had similar severity and frequency across the all age categories for each of the groups, with the exception of the auditory and visual modalities for which mixed patterns of change emerged. This first investigation of sensory profiles in WS across different age groups shows that the level of sensory difficulties remains fairly stable in individuals with WS, comparable to those diagnosed with ASD. That suggests that not only the degree of difficulties is very similar in both disorders, but also the developmental trajectory time might be comparable. Further longitudinal studies investigating changes in sensory profiles in different neurodevelopmental disorders are of course needed to enhance our understanding of the course of change in sensory symptoms over lifespan across clinical groups.

10.4 Direct assessment of auditory and tactile processing

The sensory profile comparisons in children with autism and WS presented in Chapter 4 and 7 and the reported sensory processing similarities across groups were based on caregiver-report questionnaire assessment of sensory atypicalities. Yet, the literature suggests that semi-structured direct observation is a more thorough and accurate assessment that when combined with parent-report provides a more reliable evaluation (Achenbach & Rescorla, 2004). Therefore, an observational measure of tactile and auditory hypo- and hyper-responsiveness was developed and administered directly to young children with ASD and

WS, and those with typical development (Chapter 9). Similarly to previous findings, when the Sensory Profile auditory and tactile scores were compared, significant group differences between TD and both neurodevelopmental samples were found, showing greater difficulties in sensory processing in ASD and WS groups. This finding, yet again, confirmed differences for children with ASD on the Sensory Profile (e.g. Kientz & Dunn, 1997; Kern et al., 2006) and showed similar prevalence of sensory processing difficulties in auditory and tactile domains in children with ASD and WS, with 82.4% of ASD participants falling into the category of definite difference on the auditory processing subscale and 52.6% on the tactile processing subscale, and 72.2% of children with WS showing a definite difference in auditory processing and 52.9% in the tactile processing. Similarly, when direct assessment data were compared, at the subscale level, auditory domain scores were distributed significantly differently between the TD sample and both neurodevelopmental samples, yet the ASD and WS group scores were distributed similarly; however, the scores were distributed equally for all the groups in the tactile domain. Furthermore an item distribution analysis showed a significant difference between ASD and WS children on one item only (Item 6. Response to radio while playing on the Tablet), whilst a further two items approached significance in distinguishing between ASD and WS children (Item 8. Response to hearing specific sounds and matching the sound and Item 3. Touching objects and others). The neurodevelopmental disorders groups, therefore, performed similarly when assessed using direct assessment as well as care-giver questionnaire.

10.5 A review of the evidence: Correlates of sensory profiles in individuals with ASD and WS

To better understand the degree of syndrome-specificity and cross-syndrome overlap of sensory processing in both ASD and WS, systematic and mixed-methods literature reviews were carried out to evaluate evidence of the psychological correlates of sensory processing patterns in both conditions (Chapter 2 and 3). The discrepancy between the number of publications on the topic of interest in ASD and WS, with Williams syndrome literature being very scarce, did not allow for a full comparison of relationships between sensory atypicalities and associated features in both disorders. However, it became evident that sensory difficulties play an important role in both conditions. In the ASD literature, evidence suggested that sensory hyporesponsiveness was more often associated with core features of ASD such as communication impairment, emotional, cognitive and behavioural problems, while social awareness difficulties and affective disorders were associated with hyperresponsiveness. These findings are in line with claims made by Gay and colleagues

(2008) who suggested that hyporesponsiveness and sensory seeking may be more associated with difficulties in social-communication domains in children with ASD. Interestingly, mixed association between sensory atypicalities and repetitive behaviours were found across the reviewed papers. Only more general conclusions can be made in relation to the presentation of sensory processing and its relationship with other clinical and behavioural features in WS. In the available literature associations between sensory difficulties in general and other psychological and behavioural characteristics have been examined. It was reported that in children with WS a higher degree of sensory atypicalities was associated with more difficulties in executive functioning, temperament, adaptive functioning and exhibiting more repetitive and problem behaviours (John & Mervis, 2010; Riby et al., 2013). On that general level, analogous patterns of relationships were also found in autism (Chapter 2) and Down syndrome (Bruni et al., 2010). More detailed conclusion however were hampered due to limited evidence of sensory processing features being related to behavioural and clinical symptoms in WS.

Relationship between sensory atypicalities, repetitive behaviours and anxiety

To further investigate the relationship between sensory profiles and behavioural and clinical features in both ASD and WS, an examination of the relationship between sensory processing and repetitive behaviours, anxiety and intolerance of uncertainty was undertaken in children with ASD and WS and presented in Chapter 7. A number of significant relationships were found between the sensory processing scores and RBQ total score, SCAS/PAS total score and ASC-ASD total score in both WS and ASD groups. These findings suggested that greater sensory processing difficulties were associated with more repetitive behaviours and higher anxiety levels in the WS and ASD groups and support previous report of a high degree of co-occurrence between sensory atypicalities and repetitive behaviours in ASD (Baker et al., 2008; Chen et al., 2009; Gabriels et al., 2008; Joosten et al., 2009) and in WS (Riby et al., 2013); and between sensory atypicalities and anxiety in individuals with ASD (Ben-Sasson et al., 2008; Green et al., 2011; Liss et al., 2006; Pfeiffer et al., 2005).

A more in depth examination of the relationships between sensory processing and repetitive behaviours and anxiety suggested some syndrome-specific associations. In the WS sample, significant relationships between the RBQ total score and Sensation Seeking, auditory and oral sensory processing scores were found, as well as associations between Insistence on Sameness and Sensation Seeking and oral sensory processing; while in the ASD sample only the RBQ total score, RBQ Sensory/Motor subscale and Insistence on Sameness were associated with visual sensory processing. There is some evidence, hence, that some

sensory difficulties may be associated with specific repetitive behaviours and the relationship might be different for each neurodevelopmental disorder. For example, hyper-sensitivity to certain sounds and to certain food textures, tastes or smells in WS may result in more insistence on sameness behaviours as a way to avoid and limit unpleasant sensations. The significant relationship between the presence of visual abnormalities and restricted and repetitive behaviours in ASD has been reported previously (Chen et al., 2009), however, evidence for syndrome-specificity of that association has been first suggested in Chapter 7. Further work is needed to explore the specificity of the links between sensory abnormalities and repetitive behaviours.

Interestingly, in relation to the association between sensory processing abnormalities and anxiety, the only syndrome-specific association was found in the WS group with anxiety being significantly associated with Low Registration and Sensation Seeking (when SCAS/PAS total score was used), and with Low Registration, Sensation Seeking, visual, vestibular, auditory and oral sensory processing (when ASC-ASD total score was used). Independently of measures, associations between Low Registration and Sensation Seeking and anxiety scores were established. As hypo-responsiveness consists of Low Registration and Sensation Seeking in Dunn's model (1999) these findings are rather surprising. In the ASD research a strong relationship between hyper-responsiveness, overreactivity and anxiety has been reported a number of times (for review see: Glod et al., 2015). There is some evidence, however, that individuals with WS can develop an intense fascination for certain stimulation that they found frightening at first (Levitin et al., 2005). Hence, it is possible, that in children with WS at first anxiety-provoking stimulation is avoided while with time the same stimulation, although still anxiety-provoking, is sought out. It is very interesting that a relationship between hyper-responsiveness and anxiety that has been reported a number of times in the ASD literature (Green et al., 2012; Lane et al., 2012; Lidstone et al., 2014; Mazurek et al., 2013; Mazurek et al., 2014; Mazurek & Petroski, 2015; Sullivan et al., 2014; Pfeiffer et al., 2005; Wigham et al., 2015) has not been found to be syndrome-specific. Our novel findings show how similar presentations of sensory processing abnormalities in WS and ASD, are not only present in relation to individuals but also in relation to further associations between greater sensory difficulties and higher repetitive behaviours (as previously reported by Riby et al., 2013) and anxiety levels across these two neurodevelopmental disorders.

The examination of the direct relationship between sensory processing abnormalities (both hypo- and hyperresponsiveness) and repetitive behaviours and the indirect path through intolerance of uncertainty and anxiety showed some syndrome-specific paths (Chapter 7).

While in the ASD group significant direct relationships between both sensory hypo- and hyperresponsiveness and both sensory/motor behaviours and insistence on sameness behaviours were found, supporting Wigham et al. (2015), such direct associations were not present in the WS group. On the other hand, in the WS group the relationships between sensory processing abnormalities and repetitive behaviours were entirely mediated via intolerance of uncertainty and/or anxiety. The key role of intolerance of uncertainty and/or anxiety in the presentation of repetitive behaviours in WS sample was shown, as intolerance of uncertainty or anxiety played only a partial role in mediating between sensory processing abnormalities and repetitive behaviours in the ASD group.

Moreover, the relationships between sensory hypo- and hyperresponsiveness and insistence on sameness behaviours in the WS group was mediated via intolerance of uncertainty only, while in the ASD group the relationship between sensory hyporesponsiveness and insistence on sameness behaviours was mediated via anxiety, suggesting syndrome-specific mechanisms between some of the sensory abnormalities and repetitive behaviours. Furthermore, when the analysis was performed for those children with ASD and WS who had either more typical or atypical sensory profiles, within the ASD group the relationships between sensory processing abnormalities and repetitive behaviours were found only for participants who had greater sensory difficulties and it was suggested that the relationship between sensory hyperresponsiveness and insistence on sameness was fully mediated via intolerance of uncertainty. In the WS sample, in the group of children who had more typical sensory processing profiles, the relationship between sensory hyperresponsiveness and insistence on sameness was found. For those with WS who had greater sensory difficulties, the relationship between sensory hyporesponsiveness and sensory/motor behaviours was fully mediated via anxiety and/or intolerance of uncertainty.

Although the mediating role of intolerance of uncertainty between ASD and anxiety has been previously reported (Boulter et al., 2014), its mediating role between sensory processing abnormalities and repetitive behaviours in the WS groups is a novel finding. That suggests that those children with WS who find the sensory environment unpredictable, can display more repetitive behaviours to order to regain predictability in their world. Although the role of intolerance of uncertainty has been already taken into account in anxiety treatments in typically developing population (McEvoy & Mahoney, 2012) and in treatment programme for young people with ASD (Rodgers et al., 2016), it has never been targeted in any intervention designed for children with WS. It is likely that reducing intolerance of uncertainty would impact on both sensory processing abnormalities and repetitive behaviours in WS individuals and therefore, needs to be further addressed.

10.6 Strengths and limitations

10.6.1 Novel findings

The systematic review of psychological correlates of sensory processing patterns in individuals with autism spectrum disorder and the narrative review of sensory processing in WS (Chapter 2 and 3) are first in the field to systematically explore these topics. Both literature reviews highlighted associations between sensory symptoms and other difficulties such as higher level of anxiety and repetitive behaviours, emphasizing that sensory atypicalities play an important role in both disorders. Searches for both reviews were carried out in March 2016, hence the most recent eligible papers might have not been included in the reviews.

Moreover, for the first time, sensory symptoms were detailed examination of sensory profiles were undertaken with children with WS, including an overview of changes in sensory atypicalities presentations across different age groups and an exploration of the relationship between sensory processing and anxiety, intolerance of uncertainty and repetitive behaviours was described. In addition the sensory profiles of individuals with WS were described and also compared to both typically developing group and to those with a diagnosis of ASD, adding strength to the comparisons made as both control and other neurodevelopmental condition were included. Novel findings have been added to the ASD research field. A detailed investigation of the sensory processing patterns in three rigorously selected age groups of individuals with a diagnosis of ASD without learning disability and those with ASD with additional learning disability has never previously been undertaken. Finally a novel approach to the examination of the patterns of sensory clusters based on Dunn's model (1997) in children with ASD and its associations with other clinical symptoms was introduced.

As information about sensory symptoms in children is commonly obtained through caregiver questionnaires, and only a handful of observational assessments of some of sensory behaviours are being used in a research context, the development of a tactile and auditory observation schedule facilitating the assessment of auditory and tactile hypo- and hyper-responsiveness in typical and atypical development was a novel venture. The schedule was found to be feasible for use with verbal children, both typically and atypically developing. Also, it was the first study that included minimally verbal and non-verbal children with ASD and WS (11 ASD and 9 WS respectively) in a sensory direct assessment. The majority of the

less able children were able to complete all the tasks, suggesting that with a further development of the schedule, the tool could be used in children across ability levels.

The main focus of this piece of work was the comparison of sensory profiles in children with ASD and WS. Although, the broader investigation of similarities and differences in sensory profiles in parent-child dyads in ASD was also undertaken in order to inform our understanding of how phenotypic profiles may be inherited within families. That was the first study exploring sensory processing atypicalities in dyads of children with ASD and their parents, compared to typically developing children. The findings suggested that familial factors might play a role for some aspects of sensory processing (such as sensation avoiding, and auditory, visual and vestibular sensory processing) only within ASD families; however, the role of the environment on the development of atypical sensory profiles and familiarity is yet to be established. Also, including children with other neurodevelopmental disorders and their parents would further benefit our understanding of similarities and differences in phenotypic sensory profiles within families.

10.6.2 Measurement

Most of the findings reported in this piece of work, are based on caregiver-report questionnaire assessment of sensory atypicalities, anxiety, repetitive behaviours and intolerance of uncertainty in children with ASD and WS. Although the Sensory Profile is the most commonly used tool to assess sensory symptoms in children with ASD (see review Chapter 2), questionnaire data can be a source of recollection bias or inaccurate responses (Hoyle et al. 2001). To overcome that limitation an observational measure of tactile and auditory hypo- and hyper-responsiveness was developed and administered directly to young children with ASD and WS, and those typically developing (Chapter 9). Moreover, the investigation of the factor structure and measurement invariance of the Spence Children's Anxiety Scale - Parent Version in children with ASD and typically developing anxious children was undertaken (Chapter 6) in order to determine whether the SCAS-P measures the same constructs in ASD as it does in typically developing clinically anxious children (without ASD). The confirmatory factor analysis did not confirm the conventional SCAS-P structure in the ASD sample and the findings enhanced concerns that have been raised previously with regards to the validity of the SCAS-P (Chorpita et al., 1997; Spence et al., 2001), particularly in the ASD sample, as the SCAS-P has been developed and validated for use with typically developing youth. Another anxiety measure, therefore, was also completed by the parents of children with ASD and WS when the investigation of relationship between sensory atypicalities, repetitive behaviours and anxiety was undertaken (Chapter 7). Anxiety Scale for

Children-ASD, parent-version (ASC-ASD©, Rodgers et al., 2015) assesses anxiety symptoms specific to ASD population. However, the tool has not been validated in younger children with ASD (under 8 years of age) or in the WS population. Also, it is important to highlight that parents might not always be aware of all anxiety-related behaviours that children exhibit, unless they verbalise their fears and worries. It is likely, particularly for our ASD sample and younger children, that parents were not aware of some of the symptoms or their severity and frequency, even if a questionnaire addressed ASD-specific anxiety symptoms. Furthermore, the Intolerance subscale of the ASC-ASD was used in Chapter 7 as a proxy of intolerance of uncertainty. Although the subscale consists of only 8 items compared to 27 item Intolerance of Uncertainty Scale, (Freeston et al., 1994) or 12 item Intolerance of Uncertainty Scale - Short Form (IUS-12; Carleton et al., 2007), it has good psychometric properties (Rodgers et al., 2016). Yet, the psychometric properties of the subscale were not examined in either younger children with ASD or WS children. Last, but not least, the Repetitive Behaviour Questionnaire (RBQ; Turner, 1995) includes repetitive movements, sameness behaviour, circumscribed interests and repetitive use of language related-items, however, some of the questions in the motor behaviour subscale of the RBQ could be interpreted as sensory-related (e.g. Question 1. Does he/she operate light switches, taps, the toilet flush etc. repeatedly when it is not necessary to do so?) and as such could impact part of the examination of the relationship between sensory processing and repetitive behaviours presented in Chapter 7. However, the RBQ has been previously used in investigations of relationship between sensory processing and repetitive behaviours in both children with ASD (Wigham et al., 2015) and WS (Riby et al., 2013).

10.7 Theoretical perspectives

As presented across the chapters (Chapter 4, 5 and 7), both hypo- and hyperresponsiveness have been reported to occur in the same individuals in both ASD and WS, supporting previous findings from ASD research (Baranek, 2002; Baranek et al., 2006; Ben-Sasson et al., 2009) and adding to the literature in WS. High levels of hyperresponsiveness reported in individuals with ASD are consistent with the Enhanced Perceptual Functioning hypothesis (EPF; Mottron & Burack, 2001), which proposes that superior local processing abilities (such as low-level perception required in discrimination and pattern perception) are exhibited by individuals with ASD (Shah & Frith, 1983; 1993). The EPF theory, however does not explain the presence of hyporesponsiveness in ASD. Intra-individual variability seen in autism is supported, however, by the neural noise hypothesis (Simmons et al., 2009). Neural 'noise' refers to variation in neural responses that

usually limits an ability to detect or discriminate between stimuli by reducing the signal-to-noise ratio (Baker & Meese, 2012; McDonnell & Ward, 2011), which would result in hyporesponsiveness. However, it is also likely that signal-to-noise ratio can be increased under certain circumstances (Wiesenfeld & Moss, 1995) and enhance stimulus detection or discrimination, hence improve performance in autism (Simmons et al., 2009) or result in hyperresponsiveness. Although there are compelling arguments supporting either high levels of endogenous neural noise or the contrary, reduced neural noise, as enhancing or disrupting stimulus detection and discrimination in ASD (Davis & Plaisted-Grant, 2015); both hypo- and hypersensitivity, even within the same modality, could be explained from the neural noise perspective. Given the similarity in sensory profiles reported here 'neural noise' processes are likely to occur also within WS individuals.

10.8 Clinical implications

As reported in this piece of work, children with WS show as many or even more sensory atypicalities than children with autism. Sensory processing difficulties are likely to be more global rather than syndrome-specific characteristic of individuals with developmental disorders. Surprisingly, sensory atypicalities are a significantly understudied aspect of WS and to date only three papers have focused on sensory difficulties (other than hyperacusis) in that condition (Chapter 3). Clinically, WS is still not fully understood, which might impact on professional treatments and services provided to individuals with WS and their families. This is particularly important, as first of all, in both the ASD and WS groups, parents of those children who had greater sensory processing difficulties reported more repetitive behaviours, higher level of anxiety and greater intolerance of uncertainty in their children (Chapter 7). That not only shows how similar the presentations of sensory processing abnormalities are in WS and ASD, in both relation to presentation of sensory difficulties across individuals but also in relation to further associations between greater sensory difficulties and higher repetitive behaviours and anxiety levels in these two neurodevelopmental disorders. That suggests that those individuals with ASD and WS, who have a high degree of sensory difficulties may need additional support, not only to manage their sensory difficulties, but also other co-occurring clinical symptoms. The full list of features associated with sensory atypicalities is still yet to be made. Secondly, some associations between repetitive behaviours and anxiety were found to be syndrome-specific (Chapter 7). Further examination of these relationships requires more research and clinical attention.

Moreover, the mediating role of intolerance of uncertainty and anxiety between sensory atypicalities and repetitive behaviours has been reported in ASD (Chapter 7) and interestingly, intolerance of uncertainty fully mediated relationships between sensory processing abnormalities and repetitive behaviours in the WS group. This finding suggests that those children with WS who find the sensory environment unpredictable, may display more repetitive behaviours in order to regain predictability in their world. Although the role of intolerance of uncertainty has already been taken into account in anxiety treatments in typically developing population (McEvoy & Mahoney, 2012) and is beginning to feature in treatment programmes for young people with ASD (Rodgers et al., 2016), it has never been targeted in any intervention designed for children with WS. It is likely that reducing intolerance of uncertainty would impact on both sensory processing abnormalities and repetitive behaviours in WS individuals.

There is evidence that a substantial proportion of individuals with intellectual disability, of genetic origin, also display autistic features, or meet criteria for ASD (Kaufmann et al., 2008). Behavioural profiles consistent with the diagnostic classification for ASD and overlapping features with idiopathic ASD have been reported across a number of genetic disorders, including Fragile X syndrome, Down syndrome, Velocardiofacial syndrome, Rett Syndrome, Prader-Willi syndrome, Smith-Magenis syndrome, Angelman syndrome, Turner Syndrome, San Filippo syndrome, Cohen syndrome, Smith-Lemli-Opitz syndrome, Tuberous sclerosis, phenyloketonura, adenylosuccinate lyase deficiency and Williams syndrome (Cohen et al., 2005; Feinstein et al., 2007). As demonstrated in this thesis, participants with ASD and WS had very similar sensory profiles, and as highlighted in Chapter 5, socio-communicative profiles. The overlap of core behavioural features present in both conditions reported here could lead to better understanding of many aspects of idiopathic ASD. Williams syndrome, alongside Fragile X syndrome (Kaufmann et al., 2008), could be further studied as a putative genetic model, to identify both genetic and neurological mechanisms present in idiopathic ASD.

10.9 Future directions

First of all, more research into sensory profiles in ASD and WS is required, particularly in the Williams syndrome field, as only a handful of studies have so far examined sensory processing difficulties in WS. It is important to notice that sensory difficulties in WS are not only related to sensitivity to sound and the research focus should be shifted from hyperacusis to sensory processing in general. It has been highlighted a number of times in

this thesis that individuals with WS present with as much or even more sensory difficulties then individuals with ASD, yet this phenomenon is highly understudied.

However, to fully examine the developmental trajectory of sensory symptoms in both ASD and WS, studies with longitudinal designs with several follow up points are needed. It would be interesting to include not only those children who have good language skills and average or higher IQ, but also explore the full spectrum of ability of both ASD and WS as sensory profiles, their everyday presentation and associated difficulties may differ in less and more able individuals. In addition, listening to first-hand sensory experiences of those with ASD and WS, including children could further enhance our understanding of the complexity of the subjective sensory world in neurodevelopmental disorders.

As shown in Chapter 9 tactile and auditory observation schedule developed specifically for this piece of work, was feasible for use with verbal children, both typically and atypically developing and could, with further modifications, be used as a direct assessment with minimally verbal and non-verbal children with ASD and WS. The six items that best discriminated between TD children and those with neurodevelopmental conditions could be further investigated in terms of their sensitivity to assess the presence of sensory processing difficulties in children. That work could help with a development of a brief, 25 minutes long assessment of tactile and auditory processing in both typically and atypically developing young children.

It would be fascinating to further expand research into the broader phenotype of autism. As suggested in Chapter 8 better understanding of a role of familial factors into presentation of sensory atypicalities may provide greater insight into the mechanisms underlying atypical sensory processing. Ideally, the research should focus on both biological parents and siblings of individuals with ASD, with a rigorous assessment of degree of autistic traits in family members. Investigating a role of environment in development of atypical sensory processing through qualitative interviews, longitudinal designs and comparisons made with other families with individuals with neurodevelopmental disorders could also enhance our understanding of other factors possibly playing a role in development of sensory difficulties.

10.10 Conclusion

Sensory profiles in ASD and WS were very similar and the degree of atypicalities across the sensory processing patterns and modalities was highly comparable. Similar patterns in age-related changes in sensory processing in both disorders were also observed. Yet, examination of relationship between sensory atypicalites and other clinical features such

as repetitive behaviours, anxiety and intolerance of uncertainty allowed us to trace some syndrome-specific associations. Further investigation of presentation of sensory features in association with other symptoms might help us better understand generality and specificity of the sensory profiles in ASD and WS.

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Appendices

27 March 2014

Magdalena Glod
PhD student
Institute of Neuroscience

Faculty of Medical Sciences
Newcastle University
The Medical School
Framlington Place
Newcastle upon Tyne
NE2 4HH United Kingdom

FACULTY OF MEDICAL SCIENCES: ETHICS COMMITTEE

Dear Magdalena

Title: Sensory Hotspots in Children. Sensory processing patterns in children with Autism Spectrum Disorder, Williams Syndrome and typically developing children and their parents
Application No: 00747/2014
Start date to end date: 01 April 2014 to 30 September 2016

On behalf of the Faculty of Medical Sciences Ethics Committee, I am writing to confirm that the ethical aspects of your proposal have been considered and your study has been given ethical approval.

The approval is limited to this project: **00747/2014**. If you wish for a further approval to extend this project, please submit a re-application to the FMS Ethics Committee and this will be considered.

During the course of your research project you may find it necessary to revise your protocol. Substantial changes in methodology, or changes that impact on the interface between the researcher and the participants must be considered by the FMS Ethics Committee, prior to implementation.*

At the close of your research project, please report any adverse events that have occurred and the actions that were taken to the FMS Ethics Committee.*

Best wishes,

Yours sincerely

M. Holbrough

Marjorie Holbrough
On behalf of Faculty Ethics Committee

cc.
Professor Andy Hall, Chair of FMS Ethics Committee
Ms Lois Neal, Assistant Registrar (Research Strategy)

*Please refer to the latest guidance available on the internal Newcastle web-site.

tel: +44 (0) 191 222 6000
fax: +44 (0) 191 222 6621

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I, the undersigned, confirm that (please tick box as appropriate):

1.	I have read and understood the information sheet about the project.	☐
2.	I have been given the opportunity to ask questions about the project and my participation.	☐
3.	I voluntarily agree to participate in the project.	☐
4.	I understand I can withdraw at any time without giving reasons and that I will not be penalised for withdrawing nor will I be questioned on why I have withdrawn.	☐
5.	The procedures regarding confidentiality have been clearly explained (e.g. use of names, pseudonyms, anonymisation of data, etc.) to me.	☐
6.	The use of the data in research, publications, sharing and archiving has been explained to me.	☐
7.	I understand that other researchers will have access to this data only if they agree to preserve the confidentiality of the data and if they agree to the terms I have specified in this form.	☐
8.	I understand that data may be retained for use in future projects, subject to approval by a Research Ethics Committee.	☐
10.	I agree to sign and date this informed consent form.	☐

Participant:

_____ _____ _____

Name of Participant Signature Date

Home/Postal address _____

_____

Telephone Number (house) _____ (mobile) _____

Email Address _____

Researcher:

_____ _____ _____

Name of Researcher Signature Date

Sensory Hotspots in Children

We would like to invite you to take part in a research study. Please read this information sheet before deciding to take part. It will explain why the research is being done, and what it will involve. If there is anything you are unclear about after you have read this information, please feel free to ask further questions. We can be contacted by email or by phone using the contact details at the bottom of the sheet.

Purpose of the study

Children with ASD often see the world around them differently than their siblings or peers and the way they see the world may change as they get older. We are interested in how your child reacts to different everyday sensory events, for example how she or he responds to noisy environment or bright light, at his or her age. We will first compare his or her sensory experiences of the world to the experiences of other children, of the same and different ages and those with Williams Syndrome (a rare developmental disorder). We will also ask you about your reactions to sensory information to better understand your child's responses. We hope this research will benefit the families of children with ASD by providing a wider understanding of how children with ASD experience the world, and that this will further impact future interventions.

Who is the researcher?

The lead researcher is *Magdalena Glod* who is a PhD student at Newcastle University. The researcher is being supervised in this study by *Dr. Jacqui Rodgers* (Senior Lecturer in Clinical Psychology at Newcastle University), *Dr. Deborah Riby* (Senior Lecturer and Researcher in the Psychology Department at Durham University) and *Dr. Emma Honey* (Associate Clinical Lecturer at Newcastle University and Clinical Psychologist at the Regional Complex Neurodevelopmental Disorders Service at Wakergate, NHS, Newcastle).

Why have I been asked to take part?

Parents of children from local schools, and parents who enrolled in research databases who have at least one child with either Autism Spectrum Disorder or Williams Syndrome, have been asked to participate. We would like parents of children of a wide variety of ages to be involved in the study.

What will the study involve?

The study will involve the completion of three questionnaires. These will ask about:
Sensory Profile (Caregiver Questionnaire): your child's responses to everyday sensory events,
SRS-2: his or her social and communication behaviours,
Adult/Adolescent Sensory Profile (Self Questionnaire): <u>your</u> personal reactions to sensory experiences.
You will be asked to return them to the researcher in a stamped addressed envelope enclosed with the questionnaires. It will take you up to 1 hour to fill in the questionnaires. Your child will not be directly involved in the project.

What are the benefits of this research?

There are no direct benefits from participating in this study, however we hope that this research will benefit both families and children through greater knowledge amongst professionals of how children process sensory information, and how parents and their children respond to sensory events. This may help in developing interventions for children or families who may be struggling with some of these issues.

What are the disadvantages of taking part in the study?

We hope that there will be very few disadvantages of taking part in this study. One disadvantage may be the time you will need to complete the questionnaires. However we have tried to keep this to a minimum and hope it will take as little time as possible.

Do I have to take part in the study?

You do not have to take part in this study. Participation is on a voluntary basis. If you decide to take part and you change your mind, you can withdraw from the study at any time without giving an explanation. If you wish to take part, please, keep the information sheet, sign a consent form (see attached) and send it to the researcher.

What will happen to the data?

All information collected from you will be kept confidential. Your name and personal details have been used for the purpose of contacting you. Your name and any personal details are not recorded on your responses to ensure they remain anonymous. Following completion of the data collection, your name and any personal details will be destroyed.

What will happen to the results?

The results of these studies will be available in a report. A copy of this report will be available on request. It will not be possible to identify participants from this report. We are aware that some parents may be very interested in their child's individual results from the measures completed, however due to the research nature of the study, this information cannot be given on an individual basis.

The study is being undertaken as part of the PhD programme and will form part of a PhD thesis. The results will also be published in a peer-reviewed journal and presented at national/international conferences.

Any further questions...

If you have any further questions, we would be happy to discuss them with you. You can contact us using the details below:

Jacqui Rodgers:
Jacqui.Rodgers@ncl.ac.uk
Institute of Neuroscience
4th Floor, Ridley Building
Newcastle University
Newcastle Upon Tyne
NE1 7RU

Phone: 0191 222 7562

Magdalena Glod:
m.glod@ncl.ac.uk
Sir James Spence Institute
Royal Victoria Infirmary
Newcastle University
Newcastle Upon Tyne
NE1 4LP

Sensory Hotspots in Children

Thank you for consenting to take part in this research study. Please read this information sheet. It will explain why the research is being done, and what it will involve. If there is anything you are unclear about after you have read this information, please feel free to ask further questions. We can be contacted by email or by phone using the contact details at the bottom of the sheet.

Purpose of the study

Children with Williams Syndrome often see the world around them differently than their siblings or peers, and the way they see the world may change as they get older. We are interested in how your child reacts to different everyday sensory events, for example how he or she responds to a noisy environment or bright light at his or her age. We will first compare your child's sensory experiences of the world to experiences of other children, of the same and different ages, and those with Autism Spectrum Disorder (a common developmental disorder). We will also ask you about your reactions to sensory information to better understand your child's responses. We hope this research will benefit the families of children with Williams Syndrome by providing a wider understanding of how children with this condition experience the world and this will further impact future interventions.

Who is the researcher?

The lead researcher is *Magdalena Glod* who is a PhD student at Newcastle University. The researcher is being supervised in this study by *Dr. Jacqui Rodgers* (Senior Lecturer in Clinical Psychology at Newcastle University), *Dr. Deborah Riby* (Senior Lecturer and Researcher in the Psychology Department at Durham University) and *Dr. Emma Honey* (Associate Clinical Lecturer at Newcastle University and Clinical Psychologist at the Regional Complex Neurodevelopmental Disorders Service at Wakergate, NHS, Newcastle).

Why have I been asked to take part?

Parents of children from local schools, and parents who enrolled in research databases who have at least one child with either Autism Spectrum Disorder or Williams Syndrome, have been asked to participate. We would like parents of children of a wide variety of ages to be involved in the study.

What will the study involve?

The study will involve the completion of four questionnaires. These will ask about:
Parent Questionnaire WS: some basic information about your child's diagnosis and difficulties he or she might be facing,
Sensory Profile (Caregiver Questionnaire): your child's responses to everyday sensory events,
SRS-2: his or her social and communication behaviours,
Adult/Adolescent Sensory Profile (Self Questionnaire): <u>your</u> personal reactions to sensory experiences.
You will be asked to return the completed questionnaires to the researcher in an enclosed stamped addressed envelope. It will take you around 1 hour to fill in the questionnaires. Your child will not be directly involved in the project.

What are the benefits of this research?

There are no direct benefits from participating in this study, however we hope that this research will benefit both families and children through greater knowledge amongst professionals of how children process sensory information, and how parents and their children respond to sensory events. This may help in developing interventions for children or families who may be struggling with some of these issues.

What are the disadvantages of taking part in the study?

We hope that there will be very few disadvantages of taking part in this study. One disadvantage may be the time you will need to complete the questionnaires. However we have tried to keep this to a minimum and hope it will take as little time as possible.

Do I have to take part in the study?

You do not have to take part in this study. Participation is on a voluntary basis. If you decide to take part and you change your mind, you can withdraw from the study at any time without giving an explanation.

What will happen to the data?

All information collected from you will be kept confidential. Your name and personal details have been used for the purpose of contacting you. Your name and any personal details are not recorded on your responses to ensure they remain anonymous. Following completion of the data collection, your name and any personal details will be destroyed.

What will happen to the results?

The results of these studies will be available in a report. A copy of this report will be available on request. It will not be possible to identify participants from this report. We are aware that some parents may be very interested in their child's individual results from the measures completed, however due to the research nature of the study, this information cannot be given on an individual basis.

The study is being undertaken as part of the PhD programme and will form part of a PhD thesis. The results will also be published in a peer-reviewed journal and presented at national/international conferences.

Any further questions...

If you have any further questions, we would be happy to discuss them with you. You can contact us using the details below:

Jacqui Rodgers:
Jacqui.Rodgers@ncl.ac.uk
Institute of Neuroscience
4th Floor, Ridley Building
Newcastle University
Newcastle Upon Tyne
NE1 7RU

Phone: 0191 222 7562

Magdalena Glod:
m.glod@ncl.ac.uk
Sir James Spence Institute, 3rd floor
Royal Victoria Infirmary
Newcastle University
Newcastle Upon Tyne
NE1 4LP

Appendix E

Sensory Hotspots in Children

We would like to invite you to take part in a research study. Please read this information sheet before deciding to take part. It will explain why the research is being done, and what it will involve. If there is anything you are unclear about after you have read this information, please feel free to ask further questions. We can be contacted by email or by phone using the contact details at the bottom of the sheet.

Purpose of the study

The way children see the world around them may change as they get older. We are interested in how your child reacts to different everyday sensory events, for example how he or she responds to a noisy environment or bright light, at his or her age. We will first compare your child's sensory experiences of the world to experiences of other children, including children of the same age, different ages, those with Autism Spectrum Disorder (a common developmental disorder) and those with Williams Syndrome (a rare developmental disorder). We will also ask you about your reactions to sensory information to better understand your child's responses. We hope this research will help us better understand age-related changes in perceiving sensory events in children, and will further impact future interventions for children with Autism Spectrum Disorder and Williams Syndrome.

Who is the researcher?

The lead researcher is *Magdalena Glod* who is a PhD student at Newcastle University. The researcher is being supervised in this study by *Dr. Jacqui Rodgers* (Senior Lecturer in Clinical Psychology at Newcastle University), *Dr. Deborah Riby* (Senior Lecturer and Researcher in the Psychology Department at Durham University) and *Dr. Emma Honey* (Associate Clinical Lecturer at Newcastle University and Clinical Psychologist at the Regional Complex Neurodevelopmental Disorders Service at Wakergate, NHS, Newcastle).

Why have I been asked to take part?

Parents of children from local schools, and parents who enrolled in research databases who have at least one child with either Autism Spectrum Disorder or Williams Syndrome, have been asked to participate. We would like parents of children of a wide variety of ages to be involved in the study.

What will the study involve?

The study will involve the completion of three questionnaires. These will ask about:
Sensory Profile (Caregiver Questionnaire): your child's responses to everyday sensory events,
Strengths and Difficulties Questionnaire: his or her social and emotional behaviours,
Adult/Adolescent Sensory Profile (Self Questionnaire): <u>your</u> personal reactions to sensory experiences.
You will be asked to return them with a signed consent form to the researcher. It should not take you more than 45 minutes to fill in the questionnaires. Your child will not be directly involved in the project.

What are the benefits of this research?

There are no direct benefits from participating in this study, however we hope that this research will benefit both families and children through greater knowledge amongst professionals of how children process sensory information, and how parents and their children respond to sensory events. This may help in developing interventions for children or families who may be struggling with some of these issues.

What are the disadvantages of taking part in the study?

We hope that there will be very few disadvantages of taking part in this study. One disadvantage may be the time you will need to complete the questionnaires. However we have tried to keep this to a minimum and hope it will take as little time as possible.

Do I have to take part in the study?

You do not have to take part in this study. Participation is on a voluntary basis. If you decide to take part and you change your mind, you can withdraw from the study at any time without giving an explanation. If you do take part, please, keep the information sheet and you will be asked to sign a consent form (see attached).

What will happen to the data?

All information collected from you will be kept confidential. Your name and personal details have been used for the purpose of contacting you. Your name and any personal details are not recorded on your responses to ensure they remain anonymous. Following completion of the data collection, your name and any personal details will be destroyed.

What will happen to the results?

The results of these studies will be available in a report. A copy of this report will be available on request. It will not be possible to identify participants from this report. We are aware that some parents may be very interested in their child's individual results from the measures completed, however due to the research nature of the study, this information cannot be given on an individual basis.

The study is being undertaken as part of the PhD programme and will form part of a PhD thesis. The results will also be published in a peer-reviewed journal and presented at national/international conferences.

Any further questions...

If you have any further questions, we would be happy to discuss them with you. You can contact us using the details below:

Jacqui Rodgers:
Jacqui.Rodgers@ncl.ac.uk
Institute of Neuroscience
4th Floor, Ridley Building
Newcastle University
Newcastle Upon Tyne
NE1 7RU

Phone: 0191 222 7562

Magdalena Glod:
m.glod@ncl.ac.uk
Sir James Spence Institute
Royal Victoria Infirmary
Newcastle University
Newcastle Upon Tyne
NE1 4LP

Appendix F

Debriefing Letter for Parents

Sensory Hotspots in Children Project

First of all we would like to thank you for taking part in the 'Sensory Hotspots in Children' study. We really appreciate your participation. If there is anything you are unclear about after you have read this sheet, please feel free to ask questions. We can be contacted by email or by phone using the contact details at the bottom of the sheet.

We invite you to read this debriefing letter which will further explain the purpose of our study, describe how any of your questions can be answered, and what happens next.

Purpose of the study

Children, as they get older, change the way they see the world around them and can react differently to the same sensory events. Children with ASD and Williams Syndrome sometimes respond to sensory information in a more unusual way than other children.

The aim of the 'Sensory Hotspots in Children' project was to understand how children's perception of the world changes with age and to test if it is affected by a developmental disorder. In order to do this we needed to compare results from sensory profile questionnaires of children of the same and different ages, and those with ASD, WS, and without any of those conditions. We also aimed to understand similarities and differences in perceiving the world between children and their parents. This is why we asked you to share with us some information about your way of responding to everyday sensory events. Thank you for helping us achieve our aims. We hope this research will contribute to a better understanding of age-related changes in perceiving sensory events in children, and will benefit the families of children with ASD and WS by providing a wider understanding of how children with ASD and WS experience the world and this will further impact future interventions.

100 families with children with ASD, WS and those without any disorders aged between 4-16 years were invited to take part in the 'Sensory Hotspots in Children' project, and we hope that there were very few disadvantages to taking part. However one disadvantage may have been the time you needed to complete the questionnaires. We tried to keep this to a very minimum and hope it was not too burdensome.

What happens next?

All information collected from you will be kept confidential. Your name and any personal details will not be recorded in the same place as your questionnaire responses in the database. The questionnaires will be stored securely and the data from them stored on an encrypted network drive. We will use the data to see how children and their parents respond to everyday sensory experiences.

Appendix F

The results of this study will be available in a report, which will be available on request from the researchers. The results will also be published in peer-reviewed journals and presented at conferences. It will not be possible to identify participants from the reports.

Any further questions...

If you have any further questions, we would be happy to discuss them with you. You can contact us using the details below:

Jacqui Rodgers:
Jacqui.Rodgers@ncl.ac.uk
Institute of Neuroscience
4th Floor, Ridley Building
Newcastle University
Newcastle Upon Tyne
NE1 7RU

Phone: 0191 222 7562

Magdalena Glod:
m.glod@ncl.ac.uk
Sir James Spence Institute
Royal Victoria Infirmary
Newcastle University
Newcastle Upon Tyne
NE1 4LP

Thank you very much for taking part.

10 March 2015

Magdalena Glod
School of Psychology

Faculty of Medical Sciences
Newcastle University
The Medical School
Framlington Place
Newcastle upon Tyne
NE2 4HH United Kingdom

FACULTY OF MEDICAL SCIENCES: ETHICS COMMITTEE

Dear Magdalena

Title: 'Touch, hear, react' study. Stimulation, repetition and anxiety in autism spectrum disorder (ASD) and Williams syndrome (WS).
Application No: 00848/2015
Start date to end date: 01 February 2015 to 31 September 2016

On behalf of the Faculty of Medical Sciences Ethics Committee, I am writing to confirm that the ethical aspects of your proposal have been considered and your study has been given ethical approval.

The approval is limited to this project: **00848/2015**. If you wish for a further approval to extend this project, please submit a re-application to the FMS Ethics Committee and this will be considered.

During the course of your research project you may find it necessary to revise your protocol. Substantial changes in methodology, or changes that impact on the interface between the researcher and the participants must be considered by the FMS Ethics Committee, prior to implementation.*

At the close of your research project, please report any adverse events that have occurred and the actions that were taken to the FMS Ethics Committee.*

Best wishes,

Yours sincerely

M. Holbrough

Marjorie Holbrough
On behalf of Faculty Ethics Committee

cc.
Professor Andy Hall, Chair of FMS Ethics Committee
Ms Lois Neal, Assistant Registrar (Research Strategy)

*Please refer to the latest guidance available on the internal Newcastle web-site.

tel: +44 (0) 191 222 6000
fax: +44 (0) 191 222 6621

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Consent form Appendix H

I, the undersigned, confirm that:

- I have read and understood the information sheet about the project.
- I have been given the opportunity to ask questions about the project and my participation.
- I voluntarily agree to participate in the project.
- I voluntarily agree to have my child take part in this study.
- I understand that I can withdraw myself and my child at any time without giving reason(s) and that I will not be penalised for withdrawing nor will I be questioned on why I have withdrawn.
- The procedures regarding confidentiality have been clearly explained (e.g. use of names, pseudonyms, anonymisation of data, etc.) to me.
- Separate terms of consent for video-recordings have been explained and provided to me.
- The use of the data in research, publications, sharing and archiving has been explained to me.
- I understand that other researchers will have access to this data only if they agree to preserve the confidentiality of the data and if they agree to the terms I have specified in this form.
- I understand that data may be retained for use in future projects, subject to approval by a Research Ethics Committee.
- I, along with the Researcher, agree to sign and date this informed consent form.

Parent Participant:

_____ _____ _____

Name of Participant Signature Date

Child Participant:

_____

Name of Child

Home/Postal address _____

_____

Telephone Number (house) _____ (mobile) _____

Email Address _____

Researcher:

_____ _____ _____

Name of Researcher Signature Date

Appendix I

'Touch, hear, react' study

We would like to invite you and your child to take part in a research study. Please read this information sheet before deciding to take part. It will explain why the research is being done, and what it will involve. If there is anything you are unclear about after you have read this information, please feel free to ask further questions. We can be contacted by email or by phone using the contact details at the bottom of the sheet.

Purpose of the study

We all experience the world around us through our senses. We rely on what we see, smell or taste to understand what happens around us and feel safe. Children with autism spectrum disorder (ASD) often see the world around them differently than their siblings or peers. Sometimes they can be more and sometimes less sensitive to everyday sensory experiences like sunlight or standing close to other people. We are interested in how your child reacts to different everyday sensory events, particularly those that relate to hearing and touch, for example how he or she responds to background noise or sticky textures. We have designed a new play-based measure of some of sensory experiences and would like your child to take part in the games to see whether he or she enjoys undertaking our tasks.

Everyone feels some degree of anxiety from time to time, and children are no different. We would like to know more about your child's anxiety and repetitive behaviours as many parents of children with ASD report them as very common in their children. Then we will compare your child's experiences of the world and possible difficulties to experiences of children with Williams Syndrome (a rare developmental disorder) to see if some of these experiences are more common for children with one of these conditions or if they are universal. We hope this research will benefit the families of children with ASD by providing a wider understanding of how children with ASD experience the world and if the way they respond to sounds and textures relates to some of their other difficulties.

Who is the researcher?

The lead researcher is *Magdalena Glod* who is a PhD student at Newcastle University. The researcher is being supervised in this study by *Jacqui Rodgers* (Senior Lecturer in Clinical Psychology at Newcastle University), *Deborah Riby* (Senior Lecturer and Researcher in the Psychology Department at Durham University) and *Emma Honey* (Associate Clinical Lecturer at Newcastle University and Clinical Psychologist at the Regional Complex Neurodevelopmental Disorders Service at Walkergate, NHS, Newcastle).

Why have I been asked to take part?

Parents of children from local primary schools, including special schools, and parents who have at least one child with Williams Syndrome, have been asked to participate. We would like parents and their young children to be involved in the study.

What will the study involve?

For parents, the study will involve:

- Completion of five questionnaires. These will ask about your child's responses to everyday sensory events, his or her social and communication behaviours, anxiety and repetitive behaviours. Questionnaires will be sent out to you if you consent to participate in the project. You will be asked to return them to the researcher in a stamped addressed envelope enclosed with the questionnaires. It will take you up to 1 hour to fill in the questionnaires.

- Answering some further questions (via phone call) about your child allergies and anxiety, to make the tasks he or she will be undertaking more enjoyable. You will also decide whether you prefer us to see your child at school or home.

For you child, the study will involve:

- Undertaking some jigsaws and vocabulary tests (Raven's Coloured Progressive Matrices and *British Picture Vocabulary Scale - Third Edition) to give the researcher an idea about your child's cognitive strengths and difficulties.*
- Playing some games involving listening to everyday sounds (e.g. radio, clock ticking, police siren) and touching various textures (e.g. cotton, sand paper, feathers). The tasks will be video-recorded. It will take up to 1 hour for your child to complete all the tasks.

What are the benefits of this research?

There are no direct benefits from participating in this study, however we hope that this research will benefit both families and children through greater knowledge amongst professionals of children with ASD or children with WS by providing a wider understanding of how children with these conditions experience the world and if their responses to sounds and textures relate to some of their other difficulties. This may help in developing interventions for children or families who may be struggling with some of these issues. We also hope that this work will help with establishing a new direct measure of sensory processing. Currently, there is lack of a direct assessment of sensory experiences in children.

What are the disadvantages of taking part in the study?

We hope that there will be very few disadvantages of taking part in this study. One disadvantage may be the exposure of children to unpleasant sounds and textures for them. However, before working with children directly, a brief interview with you will be undertaken (via phone). We will ask about the child's allergies to make sure that any of the materials is not going to cause any harm to the child. You will also be asked about any tactile items or sounds that are highly anxiety provoking for your child. These, if included in the tasks, will not be administered to the child. The child will be also able to switch off/stop any stimuli particularly unpleasant to them. If possible, you will be asked to be present during the examination. Another disadvantage may be the time you will need to complete the questionnaires. However we have tried to keep this to a minimum and hope it will take as little time as possible.

Do I have to take part in the study?

You and your child do not have to take part in this study. Participation is on a voluntary basis. If you decide to take part and you change your mind, you can withdraw from the study at any time without giving an explanation. We will also ask your child if he or she wants to take part. If your child refuses to take part, even if you consent the child, or change his/her mind, we will not carry on with the study.

If you do take part, please, keep the information sheet and you will be asked to sign and send back to us a consent form (see attached).

What will happen to the data?

All information collected from you and your child will be kept confidential. Your and your child's name and any personal details are not recorded on your or his/her responses to ensure they remain anonymous. Following completion of the data collection, your and your child's name, any personal details will be destroyed. The video-recordings will be watched only by the researcher to make sure that your child's reactions, e.g. smile or surprise were noticed.

What will happen to the results?

Appendix I

The results of these studies will be available in a report. A copy of this report will be available on request. It will not be possible to identify participants from this report. We are aware that some parents may be very interested in their child's individual results from the measures completed. However, due to the research nature of the study, this information cannot be given on an individual basis.

The study is being undertaken as part of the PhD programme and will form part of a PhD thesis. The results will also be published in a peer-reviewed journal and presented at national/international conferences.

Any further questions...

If you have any further questions, we would be happy to discuss them with you. You can contact us using the details below:

Jacqui Rodgers:
Jacqui.Rodgers@ncl.ac.uk
Institute of Neuroscience
4th Floor, Ridley Building
Newcastle University
Newcastle Upon Tyne
NE1 7RU

Phone: 0191 222 7562

Magdalena Glod:
m.glod@ncl.ac.uk
Sir James Spence Institute
Royal Victoria Infirmary
Newcastle University
Newcastle Upon Tyne
NE1 4LP

‘Touch, hear, react’ study

We would like to invite you and your child to take part in a research study. Please read this information sheet before deciding to take part. It will explain why the research is being done, and what it will involve. If there is anything you are unclear about after you have read this information, please feel free to ask further questions. We can be contacted by email or by phone using the contact details at the bottom of the sheet.

Purpose of the study

As part of my research I am investigating how young children process sensory information and how they may react to these experiences. We are particularly interested in children’s sensory experiences of textures and sounds in everyday settings. We know from previous research that children with developmental disabilities sometimes have difficulties dealing with sensory experiences. At the moment we do not have the right tools to measure these responses accurately. The study will help to develop a direct, observational assessment of tactile and auditory sensory processing for use with all children, including children with autism spectrum disorder (a common developmental disorder) and Williams syndrome (a rare developmental disorder).

We are also interested in looking at anxiety and repetitive behaviours in children. Everyone feels some degree of anxiety from time to time, and children are no different. We would like to know more about your child’s anxiety and repetitive behaviours as many parents report them as very common emotion and behaviours for their children. We hope this research will benefit all the families by providing a wider understanding of how children experience the world and emotions such as anxiety.

Who is the researcher?

The lead researcher is *Magdalena Glod* who is a PhD student at Newcastle University. The researcher is being supervised in this study by *Jacqui Rodgers* (Senior Lecturer in Clinical Psychology at Newcastle University), *Deborah Riby* (Senior Lecturer and Researcher in the Psychology Department at Durham University) and *Emma Honey* (Associate Clinical Lecturer at Newcastle University and Clinical Psychologist at the Regional Complex Neurodevelopmental Disorders Service at Walkergate, NHS, Newcastle). The researcher has had a recent DBS check. The study has ethical approval from Newcastle University.

Why have I been asked to take part?

Parents of children from local primary schools, including special schools, and parents who have at least one child with Williams syndrome, have been asked to participate. We would like parents and their young children to be involved in the study.

Appendix J

What will the study involve?

For parents, the study will involve:

- Completion of five questionnaires. These will ask about your child's responses to everyday sensory events, his or her social and communication behaviours, anxiety and repetitive behaviours. Questionnaires will be sent out to you after you have consented to participate in the project. You will be asked to return them to the researcher in a stamped addressed envelope enclosed with the questionnaires. It will take you up to 1 hour to fill in the questionnaires.
- Answering some further questions (via phone call) about your child allergies and anxiety, to make the tasks he or she will be undertaking more enjoyable. *You will also decide when you would prefer us to visit you and your child at your home.*

For you child, the study will involve:

- Undertaking some jigsaws and vocabulary tests (Raven's Coloured Progressive Matrices and *British Picture Vocabulary Scale - Third Edition) to give the researcher an idea about your child's cognitive strengths and difficulties.*
- Playing some games involving listening to everyday sounds (e.g. radio, clock ticking, police siren) and touching various textures (e.g. cotton, sand paper, feathers). The tasks will be video-recorded. It will take up to 1 hour for your child to complete all the tasks.

What are the benefits of this research?

There are no direct benefits from participating in this study; however study will help to develop a direct, observational assessment of tactile and auditory sensory processing for use with all children.

What are the disadvantages of taking part in the study?

We hope that there will be very few disadvantages of taking part in this study. One disadvantage may be the exposure of children to unpleasant sounds and textures for them. However, before working with children directly, a brief interview with you will be undertaken (via phone). We will ask about the child's allergies to make sure that any of the materials is not going to cause any harm to the child. You will also be asked about any tactile items or sounds that are highly anxiety provoking for your child. These, if included in the tasks, will not be administered to the child. The child will be also able to switch off/stop any stimuli particularly unpleasant to them. You could be present during the examination. Another disadvantage may be the time you will need to complete the questionnaires. However we have tried to keep this to a minimum and hope it will take as little time as possible. We have undertaken piloting of the tasks with children prior and children report finding the tasks enjoyable and fun to complete.

Do I have to take part in the study?

You and your child do not have to take part in this study. Participation is on a voluntary basis. If you decide to take part and you change your mind, you can withdraw from the study at any time without giving an explanation. We will also ask your child if he or she wants to take part. If your child refuses to take part, even if you consent the child, or change his/her mind, we will not carry on with the study.

If you do take part, please, keep the information sheet and return a signed consent form (see attached) **in the stamped self-addressed envelope provided**.

What will happen to the data?

All information collected from you and your child will be kept confidential. Your and your child name and any personal details are not recorded on your or his/her responses to ensure they remain anonymous. Following completion of the data collection, your and your child name, any personal details will be

destroyed. The video-recordings will be watched only by the researcher to make sure that your child's reactions, e.g. smile or surprise were noticed.

What will happen to the results?

The results of these studies will be available in a report. A copy of this report will be available on request. It will not be possible to identify participants from this report. We are aware that some parents may be very interested in their child's individual results from the measures completed. However, due to the research nature of the study, this information cannot be given on an individual basis.

The study is being undertaken as part of the PhD programme and will form part of a PhD thesis. The results will also be published in a peer-reviewed journal and presented at national/international conferences.

Any further questions...

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Royal Victoria Infirmary
Newcastle University
Newcastle Upon Tyne
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‘Touch, hear, react’ study

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Purpose of the study

As part of my research I am investigating how young children process sensory information and how they may react to these experiences. We are particularly interested in children’s sensory experiences of textures and sounds in everyday settings. We know from previous research that children with developmental disabilities sometimes have difficulties dealing with sensory experiences. At the moment we do not have the right tools to measure these responses accurately. In order to do that we need to first do some work with children who are typically developing. The study will help to develop a direct, observational assessment of tactile and auditory sensory processing for use with all children, including children with autism spectrum disorder (ASD, a common developmental disorder) and Williams syndrome (WS, a rare developmental disorder).

We are also interested in looking at anxiety in children. Everyone feels some degree of anxiety from time to time, and children are no different. We would like to know more about your child’s anxiety as many parents report anxiety as a very common emotion for their children. We hope this research will benefit all the families by providing a wider understanding of how children experience the world and emotions such as anxiety.

Who is the researcher?

The lead researcher is *Magdalena Glod* who is a PhD student at Newcastle University. The researcher is being supervised in this study by *Jacqui Rodgers* (Senior Lecturer in Clinical Psychology at Newcastle University), *Deborah Riby* (Senior Lecturer and Researcher in the Psychology Department at Durham University) and *Emma Honey* (Associate Clinical Lecturer at Newcastle University and Clinical Psychologist at the Regional Complex Neurodevelopmental Disorders Service at Walkergate, NHS, Newcastle). The researcher has had a recent DBS check. The study has ethical approval from Newcastle University.

Why have I been asked to take part?

Parents of children from local primary schools, including special schools, and parents who have at least one child with Williams Syndrome, have been asked to participate. We would like parents and their young children to be involved in the study.

Appendix K

What will the study involve?

For parents, the study will involve:

- Completion of four questionnaires. These will ask you about your child's responses to everyday sensory events, his or her social, communication, emotional behaviours and anxiety. Questionnaires will be sent out to you after you have consented to participate in the project. You will be asked to return them to the researcher in a stamped addressed envelope enclosed with the questionnaires. It will take you up to 40 minutes to fill in the questionnaires.
- Answering some further questions (via phone call) about your child allergies and anxiety, to make the tasks he or she will be undertaking more enjoyable. You will also decide whether you prefer us to see your child at school or home.

For you child, the study will involve:

- Undertaking some jigsaws and vocabulary tests (Raven's Coloured Progressive Matrices and *British Picture Vocabulary Scale - Third Edition) to give the researcher an idea about your child's cognitive strengths and difficulties.*
- Playing some games involving listening to everyday sounds (e.g. radio, clock ticking, police siren) and touching various textures (e.g. cotton, sand paper, feathers). The tasks will be video-recorded. It will take up to 1 hour for your child to complete all the tasks.

What are the benefits of this research?

There are no direct benefits from participating in this study; however we hope that this research will benefit both families and children through greater knowledge amongst professionals of children with autism or children with Williams syndrome by providing a wider understanding of how children with these conditions experience the world. The study will help to develop a direct, observational assessment of tactile and auditory sensory processing for use with all children.

What are the disadvantages of taking part in the study?

We hope that there will be very few disadvantages of taking part in this study. One disadvantage may be the exposure of children to unpleasant sounds and textures for them. However, before working with children directly, a brief interview with you will be undertaken (via phone). We will ask about the child's allergies to make sure that any of the materials is not going to cause any harm to the child. You will also be asked about any tactile items or sounds that are highly anxiety provoking for your child. These, if included in the tasks, will not be administered to the child. The child will be also able to switch off/stop any stimuli particularly unpleasant to them. If possible, you will be asked to be present during the examination. Another disadvantage may be the time you will need to complete the questionnaires. However we have tried to keep this to a minimum and hope it will take as little time as possible. We have undertaken piloting of the tasks with children prior and children report finding the tasks enjoyable and fun to complete.

Do I have to take part in the study?

You and your child do not have to take part in this study. Participation is on a voluntary basis. If you decide to take part and you change your mind, you can withdraw from the study at any time without giving an explanation. We will also ask your child if he or she wants to take part. If your child refuses to take part, even if you consent the child, or change his/her mind, we will not carry on with the study.

If you would like to take part, please, send us an email (m.glod@ncl.ac.uk) with your name and postal address, so we could send you a consent form to sign (see attached).

Appendix K

What will happen to the data?

All information collected from you and your child will be kept confidential. Your and your child name and any personal details are not recorded on your or his/her responses to ensure they remain anonymous. Following completion of the data collection, your and your child name, any personal details will be destroyed. The video-recordings will be watched only by the researcher to make sure that your child's reactions, e.g. smile or surprise were noticed.

What will happen to the results?

The results of these studies will be available in a report. A copy of this report will be available on request. It will not be possible to identify participants from this report. We are aware that some parents may be very interested in their child's individual results from the measures completed. However, due to the research nature of the study, this information cannot be given on an individual basis.

The study is being undertaken as part of the PhD programme and will form part of a PhD thesis. The results will also be published in a peer-reviewed journal and presented at national/international conferences.

Any further questions...

If you have any further questions, we would be happy to discuss them with you. You can contact us using the details below:

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Phone: 0191 222 7562

Magdalena Glod:
m.glod@ncl.ac.uk
Sir James Spence Institute
Royal Victoria Infirmary
Newcastle University
Newcastle Upon Tyne
NE1 4LP

Debriefing Letter for Parents

'Touch, hear, react' study

First of all we would like to thank you for taking part in the 'Touch, hear, react' study. We really appreciate your participation. If there is anything you are unclear about after you have read this sheet, please feel free to ask questions. We can be contacted by email or by phone using the contact details at the bottom of the sheet.

We invite you to read this debriefing letter which will further explain the purpose of our study, describe how any of your questions can be answered, and what happens next.

Purpose of the study

The aim of the 'Touch, hear, react' study was to understand how children's reactions to different everyday sensory events, particularly to those involving sound and touch, relate to their anxiety and repetitive behaviours. In order to do this we needed to design a play-based measure of auditory and tactile sensory experiences, and first invite typically developing children to take part in our games to help us to make sure that the tasks are enjoyable and develop scoring instructions. Then we invited children with ASD and WS to undertake the tasks. All parents were also asked to complete some questionnaires. You were asked to fill in questions about sensory experiences of your child and his or her anxiety, and if your child has one of the developmental disorders, you also completed questionnaires about his/her level of repetitive behaviours and socio-communication difficulties. Thank you for helping us with that. We hope this research will help us understand how children with ASD and WS experience the world and if their responses to sounds and textures relate to some of their other difficulties. This may help in developing interventions for children or families who may be struggling with some of these issues. We also hope that this work will help with establishing a new direct measure of sensory processing.

50 families with children with ASD, WS and those without any disorders aged between 4-9 years were invited to take part in the 'Touch, hear, react' study, and we hope that there were very few disadvantages to taking part. However one disadvantage may have been the time you needed to complete the questionnaires. We tried to keep this to a very minimum and hope it was not too burdensome. Another disadvantage may have been the exposure of your child to unpleasant sounds

and textures for him/her. However, after talking to you about your child's allergies and responses to different sounds and materials, we hope that your child had a great time playing with us.

What happens next?

All information collected from you will be kept confidential. Your and your child's name and any personal details will not be recorded in the same place as your questionnaire responses in the database. The questionnaires and video-recordings will be stored securely and the data from them will be stored on an encrypted network drive.

The results of this study will be available in a report, which will be available on request from the researchers. The results will also be published in peer-reviewed journals and presented at conferences. It will not be possible to identify participants from the reports.

Any further questions...

If you have any further questions, we would be happy to discuss them with you. You can contact us using the details below:

Dr Jacqui Rodgers:
Jacqui.Rodgers@ncl.ac.uk
Institute of Neuroscience
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Phone: 0191 222 7562

Magdalena Glod:
m.glod@ncl.ac.uk
Sir James Spence Institute
Royal Victoria Infirmary
Newcastle University
Newcastle Upon Tyne
NE1 4LP

Thank you very much for taking part.

SENSORY PROFILE

Winnie Dunn, Ph.D., OTR, FAOTA

Caregiver Questionnaire

Child's Name: _____ Birth Date: _____ Date: _____

Completed by: _____ Relationship to Child: _____

Service Provider's Name: _____ Discipline: _____

INSTRUCTIONS

Please check the box that **best** describes the frequency with which your child does the following behaviors. Please answer all of the statements. If you are unable to comment because you have not observed the behavior or believe that it does not apply to your child, please draw an X through the number for that item. Write any comments at the end of each section. Please do not write in the Section Raw Score Total row.

Use the following key to mark your responses:

ALWAYS	When presented with the opportunity, your child always responds in this manner, 100% of the time.
FREQUENTLY	When presented with the opportunity, your child frequently responds in this manner, about 75% of the time.
OCCASIONALLY	When presented with the opportunity, your child occasionally responds in this manner, about 50% of the time.
SELDOM	When presented with the opportunity, your child seldom responds in this manner, about 25% of the time.
NEVER	When presented with the opportunity, your child never responds in this manner, 0% of the time.

Printed in the United States of America

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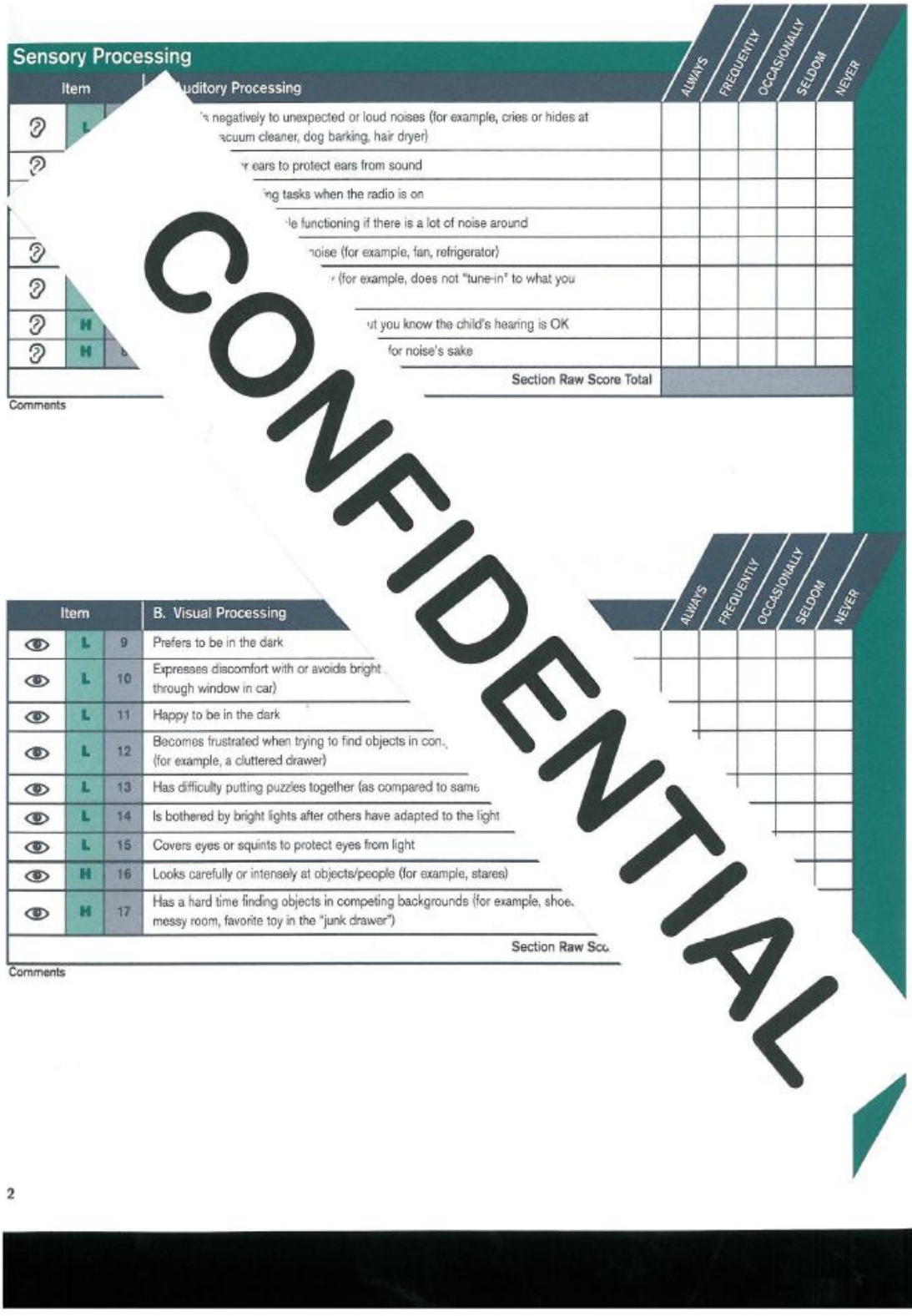

Sensory Processing

Item			Auditory Processing	ALWAYS	FREQUENTLY	OCCASIONALLY	SELDOM	NEVER
	L		's negatively to unexpected or loud noises (for example, cries or hides at acuum cleaner, dog barking, hair dryer)					
			r ears to protect ears from sound					
			ng tasks when the radio is on					
			le functioning if there is a lot of noise around					
			noise (for example, fan, refrigerator)					
			(for example, does not "tune-in" to what you					
	H		ut you know the child's hearing is OK					
	H		for noise's sake					
			Section Raw Score Total					

Comments

Item			B. Visual Processing	ALWAYS	FREQUENTLY	OCCASIONALLY	SELDOM	NEVER
	L	9	Prefers to be in the dark					
	L	10	Expresses discomfort with or avoids bright through window in car)					
	L	11	Happy to be in the dark					
	L	12	Becomes frustrated when trying to find objects in con (for example, a cluttered drawer)					
	L	13	Has difficulty putting puzzles together (as compared to same					
	L	14	Is bothered by bright lights after others have adapted to the light					
	L	15	Covers eyes or squints to protect eyes from light					
	H	16	Looks carefully or intensely at objects/people (for example, stares)					
	H	17	Has a hard time finding objects in competing backgrounds (for example, shoe messy room, favorite toy in the "junk drawer")					
			Section Raw Sco					

Comments

SRS Profile Sheet — John N. Constantino, M.D. — PARENT REPORT — FEMALE

Child's Name: _____ Chronological Age: _____

Ethnicity: _____ Respondent's Name: _____

Administration Date: _____ Relationship to Child: ☐ Mother ☐ Father ☐ Other _____

This profile for the SRS is designed to aid in diagnosis and treatment planning. The user should be familiar with the materials presented in the SRS Manual (WPS Product No. W-9998). No diagnostic or treatment decisions should be made solely on the basis of this report without confirming information from independent sources.

SRS TOTAL SCORE RESULTS

T-Score	Total Raw Score
≥100	≥100
	98–99
	96–97
	94–95
	92–93
65	91
	89–90
	87–88
	85–86
	83–84
	81–82
60	80
	78–79
	76–77
	74–75
75	72–73
	71
	69–70
	67–68
	65–66
70	63–64
	62
	60–61
	58–59
	56–57
65	54–55
	53
	51–52
	49–50
	47–48
60	45–46
	43–44
	42
	40–41
	38–39
	36–37
55	34–35
	33
	31–32
	29–30
	27–28
50	25–26
	24
	22–23
	20–21
	18–19
45	16–17
	15
	13–14
	11–12
	9–10
40	7–8
	5–6
	4
	2–3
35	0–1
30	

Total Raw Score _____

T-Score _____

TOTAL SCORE DISCUSSION

Total T-score of 76T or higher. The result is in the severe range. Scores in this range are strongly associated with a clinical diagnosis of Autistic Disorder, Asperger's Disorder, or more severe cases of PDD-NOS. They suggest a severe interference in everyday social interactions. In most clinical and educational settings, SRS scores of or above 76T from two separate informants provides very strong evidence of the presence of a clinically diagnosable autism spectrum condition.

T-score of 66T through 75T. The result is in the mild to moderate range. Scores in this range indicate deficiencies in reciprocal social behavior that are clinically significant and are resulting in mild to moderate interference in everyday social interactions. These scores are typical for children with mild or "high functioning" autism spectrum conditions, such as PDD-NOS, and higher functioning children with Asperger's Disorder.

T-score of 59T or less. The result is in the normal range. Children in the general population and not affected by autism spectrum conditions typically obtain scores in this range. When obtained on ratings in clinical settings, scores in this range usually suggest the absence of an autism spectrum condition, and any psychosocial dysfunction noted at referral is more likely to be due to a non-autism-related condition. Rarely, children with very mild, "high functioning" autism spectrum conditions may be rated in the upper end of this range (55T to 59T).

TREATMENT SUBSCALE RESULTS

T-Score	Social Awareness	Social Cognition	Social Communication	Social Motivation	Autistic Mannerisms
≥100	≥10	≥22	≥28	≥20	≥17
			26–27		
	9	20–21		19	16
			24–25		
90		18–19	22–23		14–15
	8		20–21	18	
		16–17	18–19		12–13
80	7		16–17	17	
		14–15	14–15		10–11
70	6		12–13	16	
		12–13	10–11	15	8–9
60	5	10–11	8–9	14	
		8–9	6–7	13	6–7
50	4	6–7	4–5	12	4–5
	3	4–5	2–3	11	2–3
40	2	2–3	0–1	10	0–1
	1	0–1		9	
30	0			8	

T-Score	Social Awareness	Social Cognition	Social Communication	Social Motivation	Autistic Mannerisms
Raw Score	_____	_____	_____	_____	_____
T-Score	_____	_____	_____	_____	_____

Social Responsiveness Scale AutoScore™ Form — John N. Constantino, M.D. — PARENT REPORT

DIRECTIONS

For each questi… circle the n… best d… ch…

…'s Name: _____ Chronological Age: _____

…(required) ☐ Female ☐ Male Ethnicity: _____

…'s Name: _____ Administration Date: _____

…ild: ☐ Mother ☐ Father ☐ Other _____

…OUR RESPONSES:

…TIMES TRUE 3 = OFTEN TRUE 4 = ALMOST ALWAYS TRUE

1. Si… …es than when alone.	1	2	3	4
2. Expr… …hat he or she is saying.	1	2	3	4
3. Seems … …ys.	1	2	3	4
4. When und… …ble patterns of behavior that seem odd.	1	2	3	4
5. Doesn't reco… …ntage of him or her.	1	2	3	4
6. Would rather be…	1	2	3	4
7. Is aware of what o…	1	2	3	4
8. Behaves in ways tha…	1	2	3	4
9. Clings to adults, seems…	1	2	3	4
10. Takes things too literally a… …versation.	1	2	3	4
11. Has good self-confidence.	1	2	3	4
12. Is able to communicate his or h…	1	2	3	4
13. Is awkward in turn-taking interacti… …derstand the give-and-take of conversations).	1	2	3	4
14. Is not well coordinated.	1	2	3	4
15. Is able to understand the meaning of othe… …stions.	1	2	3	4
16. Avoids eye contact or has unusual eye conta…	1	2	3	4
17. Recognizes when something is unfair.	1	2	3	4
18. Has difficulty making friends, even when trying hi…	1	2	3	4
19. Gets frustrated trying to get ideas across in conversa…	1	2	3	4
20. Shows unusual sensory interests (e.g., mouthing or spinning… …ys.	1	2	3	4
21. Is able to imitate others' actions.	1	2	3	4
22. Plays appropriately with children his or her age.	1	2	3	4
23. Does not join group activities unless told to do so.	1	2	3	4
24. Has more difficulty than other children with changes in his or her …	1	2	3	4
25. Doesn't seem to mind being out of step with or "not on the same wav…	1	2	3	4
26. Offers comfort to others when they are sad.	1	2	3	4
27. Avoids starting social interactions with peers or adults.	1	2	3	4
28. Thinks or talks about the same thing over and over.	1	2	3	4
29. Is regarded by other children as odd or weird.	1	2	3	4
30. Becomes upset in a situation with lots of things going on.	1	2	3	4
31. Can't get his or her mind off something once he or she starts thinking about it.	1	2	3	4
32. Has good personal hygiene.	1	2	3	4

wps
Test with Confidence

W-9998

Strengths and Difficulties Questionnaire

P 4-16

For each item, please mark the box for Not True, Somewhat True or Certainly True. It would help us if you answered all items as best you can even if you are not absolutely certain or the item seems daft! Please give your answers on the basis of the child's behaviour over the last six months.

Child's Name ..... Male/Female

Date of Birth.....

	Not True	Somewhat True	Certainly True
Considerate of other people's feelings	☐	☐	☐
Restless, overactive, cannot stay still for long	☐	☐	☐
Often complains of headaches, stomach-aches or sickness	☐	☐	☐
Shares readily with other children (treats, toys, pencils etc.)	☐	☐	☐
Often has temper tantrums or hot tempers	☐	☐	☐
Rather solitary, tends to play alone	☐	☐	☐
Generally obedient, usually does what adults request	☐	☐	☐
Many worries, often seems worried	☐	☐	☐
Helpful if someone is hurt, upset or feeling ill	☐	☐	☐
Constantly fidgeting or squirming	☐	☐	☐
Has at least one good friend	☐	☐	☐
Often fights with other children or bullies them	☐	☐	☐
Often unhappy, down-hearted or tearful	☐	☐	☐
Generally liked by other children	☐	☐	☐
Easily distracted, concentration wanders	☐	☐	☐
Nervous or clingy in new situations, easily loses confidence	☐	☐	☐
Kind to younger children	☐	☐	☐
Often lies or cheats	☐	☐	☐
Picked on or bullied by other children	☐	☐	☐
Often volunteers to help others (parents, teachers, other children)	☐	☐	☐
Thinks things out before acting	☐	☐	☐
Steals from home, school or elsewhere	☐	☐	☐
Gets on better with adults than with other children	☐	☐	☐
Many fears, easily scared	☐	☐	☐
Sees tasks through to the end, good attention span	☐	☐	☐

Do you have any other comments or concerns?

Please turn over - there are a few more questions on the other side

SPENCE CHILDREN'S ANXIETY SCALE
(Parent Report)

Your Name: _____ **Date:** _____

Your Child's Name: _____

BELOW IS A LIST OF ITEMS THAT DESCRIBE CHILDREN. FOR EACH ITEM PLEASE CIRCLE THE RESPONSE THAT BEST DESCRIBES YOUR CHILD. PLEASE ANSWER ALL THE ITEMS.

1.	My child worries about things.....	Never	Sometimes	Often	Always
2.	My child is scared of the dark.....	Never	Sometimes	Often	Always
3.	When my child has a problem, s(he) complains of having a funny feeling in his / her stomach	Never	Sometimes	Often	Always
4.	My child complains of feeling afraid.....	Never	Sometimes	Often	Always
5.	My child would feel afraid of being on his/her own at home.....	Never	Sometimes	Often	Always
6.	My child is scared when s(he) has to take a test.....	Never	Sometimes	Often	Always
7.	My child is afraid when (s)he has to use public toilets or bathrooms.....	Never	Sometimes	Often	Always
8.	My child worries about being away from us / me.....	Never	Sometimes	Often	Always
9.	My child feels afraid that (s)he will make a fool of him/herself in front of people.....	Never	Sometimes	Often	Always
10.	My child worries that (s)he will do badly at school.....	Never	Sometimes	Often	Always
11.	My child worries that something awful will happen to someone in our family.....	Never	Sometimes	Often	Always
12.	My child complains of suddenly feeling as if (s)he can't breathe when there is no reason for this.....	Never	Sometimes	Often	Always
13.	My child has to keep checking that (s)he has done things right (like the switch is off, or the door is locked)..	Never	Sometimes	Often	Always
14.	My child is scared if (s)he has to sleep on his/her own.....	Never	Sometimes	Often	Always
15.	My child has trouble going to school in the mornings because (s)he feels nervous or afraid.....	Never	Sometimes	Often	Always
16.	My child is scared of dogs	Never	Sometimes	Often	Always
17.	My child can't seem to get bad or silly thoughts out of his / her head.....	Never	Sometimes	Often	Always
18.	When my child has a problem, s(he) complains of his/her heart beating really fast.....	Never	Sometimes	Often	Always

19.	My child suddenly starts to tremble or shake when there is no reason for this.....	Never	Sometimes	Often	Always
20.	My child worries that something bad will happen to him/her.....	Never	Sometimes	Often	Always
21.	My child is scared of going to the doctor or dentist	Never	Sometimes	Often	Always
22.	When my child has a problem, (s)he feels shaky.....	Never	Sometimes	Often	Always
23.	My child is scared of heights (eg. being at the top of a cliff).....	Never	Sometimes	Often	Always
24.	My child has to think special thoughts (like numbers or words) to stop bad things from happening.....	Never	Sometimes	Often	Always
25.	My child feels scared if (s)he has to travel in the car, or on a bus or train	Never	Sometimes	Often	Always
26.	My child worries what other people think of him/her.....	Never	Sometimes	Often	Always
27.	My child is afraid of being in crowded places (like shopping centres, the movies, buses, busy playgrounds).....	Never	Sometimes	Often	Always
28	All of a sudden my child feels really scared for no reason at all.....	Never	Sometimes	Often	Always
29.	My child is scared of insects or spiders.....	Never	Sometimes	Often	Always
30.	My child complains of suddenly becoming dizzy or faint when there is no reason for this.....	Never	Sometimes	Often	Always
31.	My child feels afraid when (s)he has to talk in front of the class.....	Never	Sometimes	Often	Always
32.	My child's complains of his / her heart suddenly starting to beat too quickly for no reason	Never	Sometimes	Often	Always
33.	My child worries that (s)he will suddenly get a scared feeling when there is nothing to be afraid of.....	Never	Sometimes	Often	Always
34.	My child is afraid of being in small closed places, like tunnels or small rooms.....	Never	Sometimes	Often	Always
35.	My child has to do some things over and over again (like washing his / her hands, cleaning or putting things in a certain order).....	Never	Sometimes	Often	Always
36.	My child gets bothered by bad or silly thoughts or pictures in his/her head	Never	Sometimes	Often	Always
37.	My child has to do certain things in just the right way to stop bad things from happening	Never	Sometimes	Often	Always
38.	My child would feel scared if (s)he had to stay away from home overnight.....	Never	Sometimes	Often	Always
39.	Is there anything else that your child is really afraid of?	YES	NO		
	Please write down what it is, and fill out how often (s)he is afraid of this thing: _____	Never	Sometimes	Often	Always
	_____	Never	Sometimes	Often	Always
	_____	Never	Sometimes	Often	Always

'Touch, hear, react' advert

Do you have a young child with a diagnosis of ASD, aged between 4-9 years? Would you like to help out with some research? We are a group of researchers at Newcastle University. We are interested in understanding how young children experience their sensory world and how they deal with and react to everyday sounds and textures. We know from previous research that children with autism sometimes have difficulties dealing with sensory experiences. At the moment we do not have the right tools to measure these responses accurately. This study will help to develop a way gaining a better understanding of these experiences. We are also interested in hearing from parents about young children's worries and repetitive behaviours. We hope this research will benefit all the families by providing a better understanding of how children experience the world and emotions such as anxiety. If you agree to take part one of the researchers (Magda Glod) will visit you at home. Mums or Dads will be asked to complete some questionnaires about their child's experiences and reactions. Children will be asked to play some games involving listening to everyday sounds and touching various textures. We will video-record the session. It will take up to 1 hour for the child to complete all the games. If you are interested or want to hear more please email m.glod@ncl.ac.uk or Jacqui.rodgers@ncl.ac.uk and we can provide you with an information sheet. Thank you.

PRESCHOOL ANXIETY SCALE
(Parent Report)

Your Name: _____ **Date:** _____

Your Child's Name: _____

Below is a list of items that describe children. For each item please circle the response that best describes your child. Please circle the **4** if the item is **very often true, 3** if the item is **quite often true, 2** if the item is **sometimes true, 1** if the item is **seldom true** or if it is **not true at all** circle the **0**. Please answer all the items as well as you can, even if some do not seem to apply to your child.

		Not True at All	Seldom True	Sometimes True	Quite Often True	Very Often True
1	Has difficulty stopping him/herself from worrying.....	0	1	2	3	4
2	Worries that he/she will do something to look stupid in front of other people.....	0	1	2	3	4
3	Keeps checking that he/she has done things right (e.g., that he/she closed a door, turned off a tap).....	0	1	2	3	4
4	Is tense, restless or irritable due to worrying.....	0	1	2	3	4
5	Is scared to ask an adult for help (e.g., a preschool or school teacher).....	0	1	2	3	4
6	Is reluctant to go to sleep without you or to sleep away from home.....	0	1	2	3	4
7	Is scared of heights (high places).....	0	1	2	3	4
8	Has trouble sleeping due to worrying.....	0	1	2	3	4
9	Washes his/her hands over and over many times each day.....	0	1	2	3	4
10	Is afraid of crowded or closed-in places.....	0	1	2	3	4
11	Is afraid of meeting or talking to unfamiliar people.....	0	1	2	3	4
12	Worries that something bad will happen to his/her parents.....	0	1	2	3	4
13	Is scared of thunder storms.....	0	1	2	3	4
14	Spends a large part of each day worrying about various things.....	0	1	2	3	4
15	Is afraid of talking in front of the class (preschool group) e.g., show and tell.....	0	1	2	3	4
16	Worries that something bad might happen to him/her (e.g., getting lost or kidnapped), so he/she won't be able to see you again.....	0	1	2	3	4
17	Is nervous of going swimming.....	0	1	2	3	4

		Not True at All	Seldom True	Sometimes True	Quite Often True	Very Often True
18	Has to have things in exactly the right order or position to stop bad things from happening.....	0	1	2	3	4
19	Worries that he/she will do something embarrassing in front of other people.....	0	1	2	3	4
20	Is afraid of insects and/or spiders.....	0	1	2	3	4
21	Has bad or silly thoughts or images that keep coming back over and over.....	0	1	2	3	4
22	Becomes distressed about your leaving him/her at preschool/school or with a babysitter.....	0	1	2	3	4
23	Is afraid to go up to group of children and join their activities.....	0	1	2	3	4
24	Is frightened of dogs.....	0	1	2	3	4
25	Has nightmares about being apart from you.....	0	1	2	3	4
26	Is afraid of the dark.....	0	1	2	3	4
27	Has to keep thinking special thoughts (e.g., numbers or words) to stop bad things from happening.....	0	1	2	3	4
28	Asks for reassurance when it doesn't seem necessary.....	0	1	2	3	4
29	Has **your child ever experienced anything really bad or traumatic** (e.g., severe accident, death of a family member/friend, assault, robbery, disaster)	**YES**	**NO**			

Please briefly describe the event that your child experienced.....

_____

_____

_____

_____

_____

If you answered **NO** to **question 29**, please **do not** answer questions 30-34. **If you answered YES, please DO** answer the following questions.

Do the following statements describe your child's behaviour since the event?

30	Has bad dreams or nightmares about the event.....	0	1	2	3	4
31	Remembers the event and becomes distressed.....	0	1	2	3	4
32	Becomes distressed when reminded of the event.....	0	1	2	3	4
33	Suddenly behaves as if he/she is reliving the bad experience.....	0	1	2	3	4
34	Shows bodily signs of fear (e.g., sweating, shaking or racing heart) when reminded of the event	0	1	2	3	4

ASC-ASD - Parent Version

Anxiety Scale for Children – Autism Spectrum Disorder – Parent version (ASC-ASD -P) ©

Jacqui Rodgers, Sarah Wigham, Helen McConachie, Mark Freeston, Emma Honey, Jeremy Parr
Newcastle University, Newcastle UK

Name of child:_____ **Age of Child (years/months):**_____

Date:_____ **Relationship to Child:**_____

Please put a circle around the word that shows how often each of these things happens for your child.

1. My child suddenly gets a scared feeling when there is nothing to be afraid of	Never	Sometimes	Often	Always
2. My child worries what other people think of him/her e.g. that he/ she is different	Never	Sometimes	Often	Always
3. My child's heart suddenly starts to beat too quickly for no reason	Never	Sometimes	Often	Always
4. My child feels scared when taking a test in case they make a mistake or don't understand the questions	Never	Sometimes	Often	Always
5. My child worries that people will bump into him/ her or touch him/ her in busy or crowded environments	Never	Sometimes	Often	Always
6. My child is afraid of being in crowded places (like shopping centres, the movies, buses, busy playgrounds) in case he/ she is separated from his/ her family	Never	Sometimes	Often	Always
7. My child worries about doing badly at school work	Never	Sometimes	Often	Always

Developed with the generous support of The Baily Thomas Fund
Jacqui.Rodgers@ncl.ac.uk

ASC-ASD - Parent Version

8. My child suddenly feels so anxious he/ she feels as if he/she can't breathe when there is no reason for this	Never	Sometimes	Often	Always
9. My child is afraid of new things, or new people or new places	Never	Sometimes	Often	Always
10. My child is afraid of entering a room full of people	Never	Sometimes	Often	Always
11. My child worries when in bed at night because he/ she does not like to be away from his her parents/ family	Never	Sometimes	Often	Always
12. When my child has a problem, he/she feels shaky	Never	Sometimes	Often	Always
13. My child suddenly starts to tremble or shake when there is no reason for this	Never	Sometimes	Often	Always
14. Feeling unsure stops my child from doing most things	Never	Sometimes	Often	Always
15. My child worries when he/she thinks he/she has done poorly at something in case people judge him/ her negatively	Never	Sometimes	Often	Always
16. My child always needs to be prepared before things happen	Never	Sometimes	Often	Always
17. My child feels afraid that he/she will make a fool of him/herself in front of people	Never	Sometimes	Often	Always
18. My child worries about being away from me	Never	Sometimes	Often	Always
19. My child worries that something awful will happen to someone in the family	Never	Sometimes	Often	Always
20. My child feels scared to be away from home because his/ her parents are familiar with his/ her bedtime routine	Never	Sometimes	Often	Always

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Jacqui.Rodgers@ncl.ac.uk

ASC-ASD - Parent Version

21. My child worries about being in certain places because it might be too loud, or too bright or too busy	Never	Sometimes	Often	Always
22. My child suddenly becomes dizzy or faint when there is no reason for this	Never	Sometimes	Often	Always
23. My child worries if they don't know what will happen next e.g. if plans change	Never	Sometimes	Often	Always
24. My child worries that something bad will happen to him/her	Never	Sometimes	Often	Always

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Jacqui.Rodgers@ncl.ac.uk

REPETITIVE BEHAVIOUR QUESTIONNAIRE

Thank you for taking time to complete this questionnaire.

Although there are several pages of questions, you will find that many can be answered with a quick ‘no’ response. In this way you should be able to complete the questionnaire quite quickly.

Please record the behaviour that your son or daughter a shows at the moment (over the last three months). Please describe and rate the most usual way he/she displays this behaviour. Each question is followed by a list of alternatives. Please tick the box next to the alternative that best describes the behaviour shown by your son or daughter. Where he/she shows two or more behaviours of the type probed by one question then please describe the code each separately. The examples given in each question are only a guide to the type of behaviour that can be shown; please describe any other behaviours of the type probed by each question. If your son daughter shows any behaviour that is not covered by the questionnaire please describe this and provide as much information as you can on additional sheet of paper.

For those items that ask about the frequency with which behaviour is shown, please rate how frequently your son or daughter might display the behaviour over the course of the day if you were watching them all day. Think about this either in terms of the number of bouts of this behaviour he/she would show over the course of the entire day, or if it is more appropriate, the number of bouts of this behaviour that might occur in a typical hour.

Please try to complete each question as accurately as you can and try not to leave any question, or any part of a question, unanswered.

REPETITIVE BEHAVIOUR QUESTIONNAIRE

Your name: _____

Today's date: __ __ / __ __ / __ __ __ __

Young person's name: _____

Young person's date of birth: __ __ / __ __ / __ __ __ __

---

1. Does he/she operate light switches, taps, the toilet flush etc. repeatedly when it is not necessary to do so?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Please describe this behaviour

Is there any specific time or situation when this behaviour is especially likely to occur?

---

2. Does he/she arrange toys or other items in rows or patterns

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Please describe this behaviour

Is there any specific time or situation when this behaviour is especially likely to occur?

---

3. Does he/she repeatedly fiddle with toys or other items?
For example, does he/she spin, twiddle, bang, tap, twist, flick or wave anything repetitively?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Please describe this behaviour

Is there any specific time or situation when this behaviour is especially likely to occur?

4. Does he/she touch parts of his/her body or clothing repeatedly?
For example, does he/she repeatedly rub his legs, pull at the buttons on his/her clothing, or touch his/ her ear or elbow etc.?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Please describe this behaviour

Is there any specific time or situation when this behaviour is especially likely to occur?

5. Is he/she attached to anything in particular?
For example, does he/she carry a teddy, a blanket or stick etc. around with him/her?

- ○ **No** particular attachment to any object
- ○ Attachment to an object commonly used as a comforter (e.g. teddy, blanket etc.)
- ○ Attachment to an unusual object (e.g. stick, glove etc.)

Please describe this behaviour

6. Does he/she obsessively collect or hoard items of any sort?

- ○ **No** obsessive or unusually keen collecting or hoarding
- ○ Very keen collector of **usual** items (e.g. stamps, football cards etc.)
- ○ Very keen collector of **unusual** or odd items (e.g. leaflets, jar lids, sticks etc.)

Please describe this behaviour

---

7. Does he/she spin him/herself around and around?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Is there any specific time or situation when this behaviour is especially likely to occur?

---

8. Does he/she rock backwards and forwards, or side to side, either when sitting or when standing?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Is there any specific time or situation when this behaviour is especially likely to occur?

---

9. Does he/she bang his/her head? Does he/she do this repetitively?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Is there any specific time or situation when this behaviour is especially likely to occur?

---

10. Does he/she pace or move around repetitively?
For example, does he/she walk to and fro across a room, or around the house or garden repetitively?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Please describe this behaviour

Is there any specific time or situation when this behaviour is especially likely to occur?

---

11. Does he/she make repetitive hand and/or finger movements?
For example, does he/she repetitively wave, flick, flap or twiddle his/her hands or fingers repetitively?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Please describe this behaviour

Is there any specific time or situation when this behaviour is especially likely to occur?

---

12. Does he/she make other repetitive body movements?
For example, does he/she repeatedly clasp his/her hands, tap his/her feet, swing his/her legs or jump etc.?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Please describe this behaviour

Is there any specific time or situation when this behaviour is especially likely to occur?

---

13. Does he/she ever injure him/herself?
For example, does he/she bite, scratch, knock or pick him/herself? Does he she do this repeatedly?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Please describe this behaviour

Is there any specific time or situation when this behaviour is especially likely to occur?

14. Does he/she insist on things about the house staying the same?
For example, does he/she insist on furniture staying in the same place, or curtains being open or closed etc.?

- ○ **No**
- ○ **Mild** problem which does not effect others
- ○ **Serious** problem which effects others on a regular basis

Please describe this behaviour

15. Does he/she insist on other items being put out, kept or stored in the same way?
For example, does he/she like ornaments, toys or cassette tapes kept in the same places or positions?

- ○ **No**
- ○ **Mild** problem which does not effect others
- ○ **Serious** problem which effects others on a regular basis

Please describe this behaviour

16. Does he/she play the same music, game or video, or read the same book repeatedly?

- ○ **Never** or rarely
- ○ **Regular** feature of behaviour, but will tolerate alternatives when necessary
- ○ **Highly regular** and **highly rigid** feature of behaviour. Will not tolerate any alternatives

Please describe this behaviour

---

17. Does he/she insist on using the same objects or items in any other situation?
For example, does he/she insist on using the same chair, plate, bed linen or door? (DO NOT count any insistence on using the same mug or cup)

- ○ **Never** or rarely
- ○ **Regular** feature of behaviour, but will tolerate alternatives when necessary
- ○ **Highly regular** and **highly rigid** feature of behaviour. Will not tolerate any alternatives

Please describe this behaviour

---

18. Does he/she insist on wearing the same clothes or refuses to wear new clothes?

- ○ **Never** or rarely
- ○ **Regular** feature of behaviour, but will tolerate alternatives when necessary
- ○ **Highly regular** and **highly rigid** feature of behaviour. Will not tolerate any alternatives

Please describe this behaviour

---

19. Does he/she insist that certain items of clothing must always be worn or worn in the same situation or in the same way?
For example, does he/she insist on always wearing a vest, or wearing a hat to the shops, or always buttoning a shirt to the collar?

- ○ **Never** or rarely
- ○ **Regular** feature of behaviour, but will tolerate alternatives when necessary
- ○ **Highly regular** and **highly rigid** feature of behaviour. Will not tolerate any alternatives

Please describe this behaviour

---

20. Does he/she insist on eating the same foods, or a very small range of foods, at every meal?

- ○ **Never** or rarely
- ○ **Regular** feature of behaviour, but will tolerate alternatives when necessary
- ○ **Highly regular** and **highly rigid** feature of behaviour. Will not tolerate any alternatives

Please describe this behaviour

---

21. Does he/she insist on moving or travelling by the same route?
For example, does he/she insist on taking the same route when moving about the house, going for a walk, or travelling in the car?

- ○ **Never** or rarely
- ○ **Regular** feature of behaviour, but will tolerate alternatives when necessary
- ○ **Highly regular** and **highly rigid** feature of behaviour. Will not tolerate any alternatives

Please describe this behaviour

---

22. How does he/she react if any changes are made to his/her surroundings at home?
For example, if you move the furniture, or rearrange the way that certain items are stored or organised?

- ○ May comment on, or notice the change but shows no negative reaction
- ○ Accepts the change, but shows some degree of anxiety or mildly negative reaction
- ○ Will accept the change, but shows extreme anxiety or strong negative reaction (e.g. tantrum)
- ○ Will **not** accept the change. Persistently attempts to rearrange the items

---

23. Are there any aspects of routine that he/she insists must remain the same?
For example, does he/she insist on always bathing before breakfast, on going to the shops every afternoon, or on watching a video after every meal?

- ○ **No**
- ○ **Mild** problem which does not effect others
- ○ **Serious** problem which effects others on a regular basis

Please describe this routine

---

24. Does he/she make rituals out of everyday activities such as eating, dressing, getting in the car, walking up stairs etc.?

- ○ **No**
- ○ **Mild** problem which does not effect others
- ○ **Serious** problem which effects others on a regular basis

Please describe this activity and ritual(s)

---

25. Does he or have any rituals that are linked to particular occasions or places?
For example, does he/she have specific rituals for the supermarket, the Doctor's surgery or a relative's house?

- ○ **No**
- ○ **Mild** problem which does not affect others
- ○ **Serious** problem which affects others on a regular basis

Please describe this ritual(s)

---

26. How does he/she react his/her daily routine is changed

- ○ May comment on, or notice the change but shows no negative reaction
- ○ Accepts the change, but shows some degree of anxiety or mildly negative reaction
- ○ Will accept the change, but shows extreme anxiety or strong negative reaction (e.g. tantrum)
- ○ Will **not** accept any change to routine

---

27. Does he/she 'echo' or repeat what other people say?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Is there any specific time or situation when this behaviour is especially likely to occur?

---

28. Does he/she say the same things, or make the same noises, repeatedly?
For example, does he/she say the same word repeatedly or other sounds such as hums or growls or clicking noises? Or does he/she use the same 'stock phrases' frequently?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Please describe this behaviour

Is there any specific time or situation when this behaviour is especially likely to occur?

---

29. Does he/she talk about the same topic over and over again?

- ○ **Never** or rarely
- ○ **One** or more bouts of this behaviour daily
- ○ **15** or more bouts of this behaviour daily (or at least one bout an hour)
- ○ **30** or more bouts of this behaviour daily (or at least two bouts an hour)

Please describe this behaviour

Is there any specific time or situation when this behaviour is especially likely to occur?

---

30. Does he/she have any interests or hobbies? Please describe these briefly.

In particular, does he/she have any interests or preoccupations which you would describe as overly keen, obsessional, or unusual in any way?
Please describe any such interests in as much detail as you can.

30 (continued) In summary would you say that he/she has:

- ○ A varied pattern of interests which he/she will pursue spontaneously and without prompting
- ○ One or more obsessional interests, but also other usual interests which he/she will pursue spontaneously and without prompting
- ○ Only obsessional interests which he/she will pursue spontaneously
- ○ Has no particular interests or hobbies that he/she will pursue spontaneously

(DO NOT include watching TV as an interest or hobby)

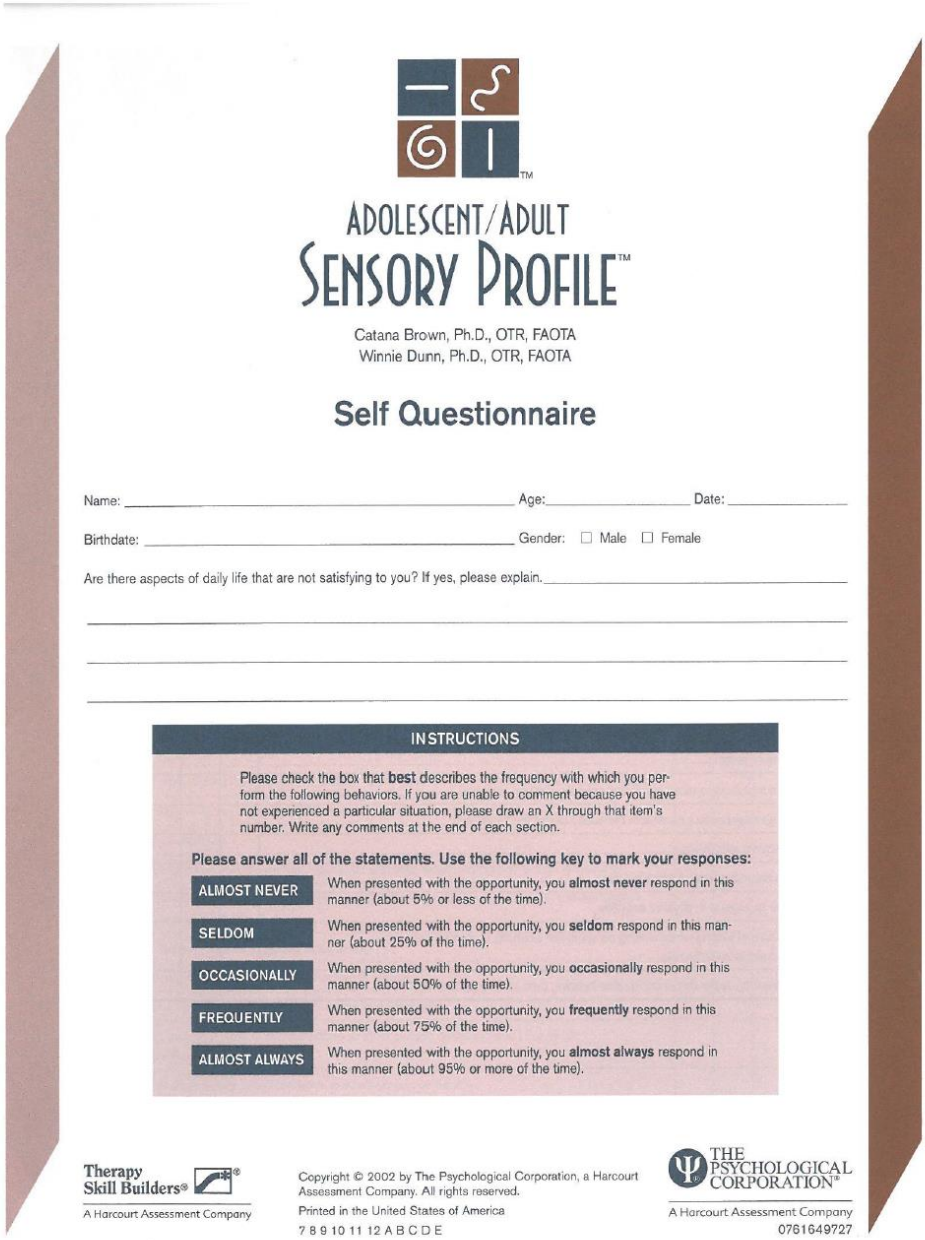
31. What was the earliest repetitive activity that you remember your son or daughter showing?

How old was he/she when this began?

32. Of all the behaviours in this questionnaire that your son or daughter engage in, which one would you say is the most marked or the most noticeable?

33. Of all the behaviours in this questionnaire that your son or daughter engage in, which one would you say causes the greatest problem in day-to-day life?

Thank you for completing this questionnaire



ADOLESCENT/ADULT SENSORY PROFILE™

Catana Brown, Ph.D., OTR, FAOTA
Winnie Dunn, Ph.D., OTR, FAOTA

Self Questionnaire

Name: _____ Age: _____ Date: _____

Birthdate: _____ Gender: ☐ Male ☐ Female

Are there aspects of daily life that are not satisfying to you? If yes, please explain. _____

_____

_____

_____

INSTRUCTIONS

Please check the box that **best** describes the frequency with which you perform the following behaviors. If you are unable to comment because you have not experienced a particular situation, please draw an X through that item's number. Write any comments at the end of each section.

Please answer all of the statements. Use the following key to mark your responses:

ALMOST NEVER	When presented with the opportunity, you **almost never** respond in this manner (about 5% or less of the time).
SELDOM	When presented with the opportunity, you **seldom** respond in this manner (about 25% of the time).
OCCASIONALLY	When presented with the opportunity, you **occasionally** respond in this manner (about 50% of the time).
FREQUENTLY	When presented with the opportunity, you **frequently** respond in this manner (about 75% of the time).
ALMOST ALWAYS	When presented with the opportunity, you **almost always** respond in this manner (about 95% or more of the time).

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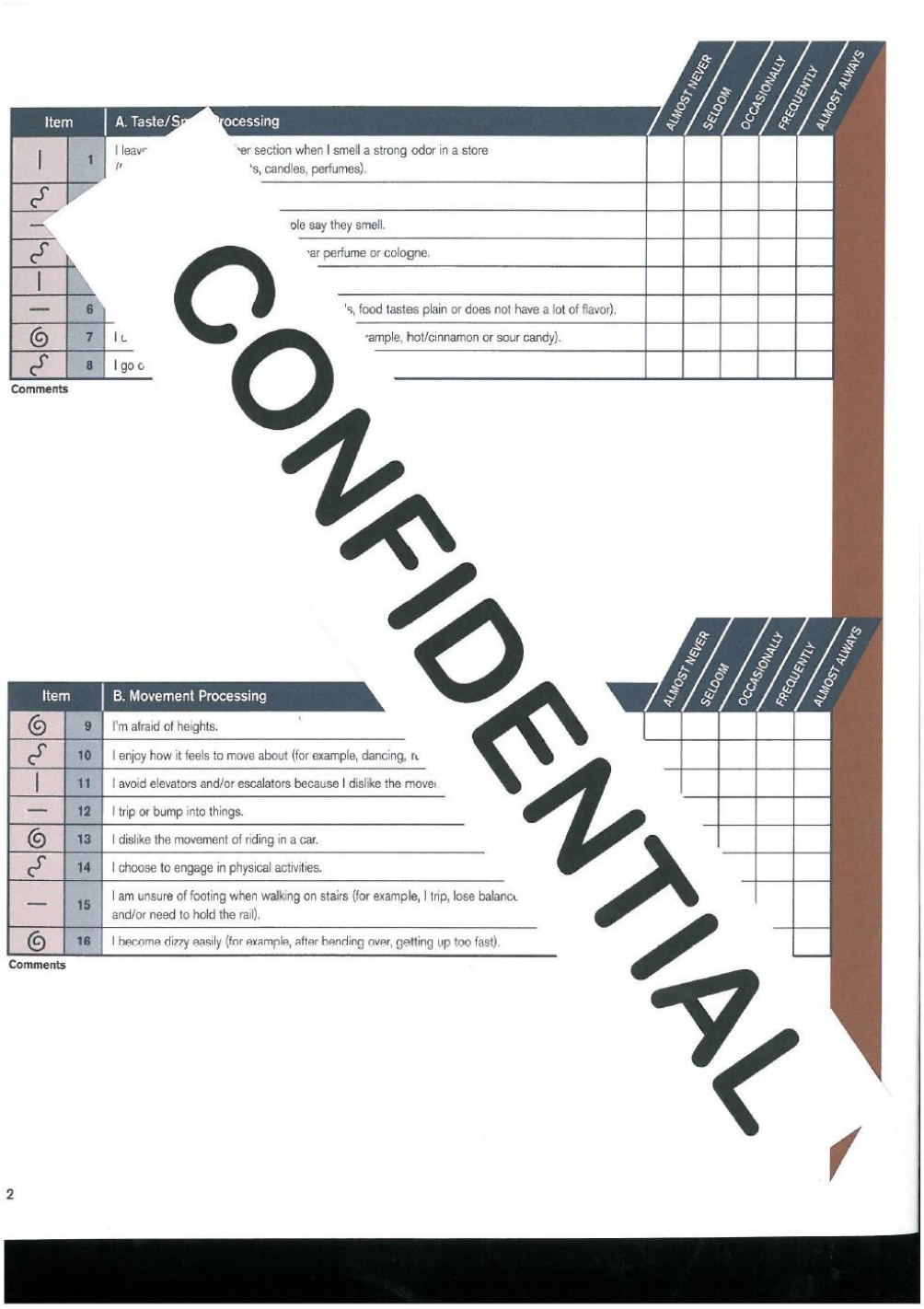

Item		A. Taste/Sm…rocessing	ALMOST NEVER	SELDOM	OCCASIONALLY	FREQUENTLY	ALMOST ALWAYS
	1	I leav…er section when I smell a strong odor in a store (f… …s, candles, perfumes).					
		…ole say they smell.					
		…ar perfume or cologne.					
	6	…s, food tastes plain or does not have a lot of flavor).					
	7	I … …ample, hot/cinnamon or sour candy).					
	8	I go …					

Comments

Item		B. Movement Processing	ALMOST NEVER	SELDOM	OCCASIONALLY	FREQUENTLY	ALMOST ALWAYS
	9	I'm afraid of heights.					
	10	I enjoy how it feels to move about (for example, dancing, r…					
	11	I avoid elevators and/or escalators because I dislike the move…					
	12	I trip or bump into things.					
	13	I dislike the movement of riding in a car.					
	14	I choose to engage in physical activities.					
	15	I am unsure of footing when walking on stairs (for example, I trip, lose balanc… and/or need to hold the rail).					
	16	I become dizzy easily (for example, after bending over, getting up too fast).					

Comments

Telephone questionnaire:

Appendix V

Demographic information:

1. Name and date of birth ……
2. Is English first language of your child? What languages can he/she speak?

……..

3. What's your child ethnic group (e.g. White, mixed, Asian/Asian British, Black/African/Cariban/Black British, other)

3a. <u>ASD ONLY</u> What age your child was given the diagnosis?.....

Questions about well-being and development:

4. Is your child's vision/hearing/motor skills normal/corrected to normal?

Is your child seeing ok or does your child need to wear glasses? …….

Is your child hearing ok or does your child need any hearing aid? ……...

Are your child writing, using a spoon, walking ok or does your child need any motor aid?

……

5. Does your child has any medical diagnosis (developmental, neurological etc)?

……

6. Does your child take any medication?.....

.....

Questions to ensure that the child enjoys the tasks/games:

7. Is your child in particular interested in something, so you know when you mention it, you will get his/her attention (e.g. favourite computer game, favourite activity)?

……

8. Are there any sounds that your child really does not like or even make him/her anxious (he/she is upset, frustrated, cries or hides when hearing the sound)?

……

9. Are there any materials/textures that your child really does not like or even make him/her anxious (he/she is upset, frustrated, cries or hides when hearing the sound)?

……

10. Does your child has any allergies?.....
11. Would you like me to see your child at home or school?

Free-play time/general observation

Item 1. Sound seeking (Hypo)

Item 8. Enjoys strange noises/seeks to make noise for noise's sake

Codes the child's unusual interest in sound/making sound

☐ 0=does not seem to be unusually interested in any of the sounds presented in the free-time session or across the tasks

1=wants to listen to a stimulus two or three times OR shows some additional interest in the stimulus OR plays with noisy toys for a prolonged time OR makes sounds himself/herself occasionally (**one to three times**)

2=wants to listen to a stimulus a number of times, to a stage when it interrupts with carrying on the task OR tries to re-play the stimulus OR keeps talking about the stimulus OR seeks to make sound for sound's sake OR plays only with noisy toys OR makes sounds himself/herself a number of times

Item 2. Need for touching certain fabrics (Hypo)

Item 41. Displays unusual need for touching certain toys, surfaces, or textures (for example, constantly touching objects)

☐ 0=does not seem to be particularly attracted to any particular fabric, carries out the tasks smoothly and does not show any unusual interest in any toy, object, material during the study

1=verbally expresses appreciation for touching the item OR wants to touch at least one of the fabrics/toys/objects **two or three times,** but does not interrupt with carrying on the next task

2=wants to touch at least one of the fabrics/toys/objects **a number of times**, to a stage when it interrupts with carrying on the next task OR keeps talking about the task OR unusually plays with toys/objects (focusing on their texture/material rather than function)

Item 3. Touching objects and others (Hypo)

Item 40. Touches people and objects to the point of irritating others

Item 45. Touches people and objects

Codes the child's touching behaviours

☐ 0=does not seem to be interested in touching objects or other people beyond the tasks' requirements

1= touches objects and other people (e.g. parent, examiner) occasionally in a non-functional manner OR one clear example of unusual touch

2=touches objects and other people (e.g. parent, examiner) frequently, even to the point of irritating others, shows unusual interest in different textures

Item 4. Child's need to be touched (Hypo)

Codes child's unusual need to be touched by others or objects

☐ 0=does not look for any additional tactile stimulation from other people or objects

1=occasionally (1-2 times) seeks for additional tactile stimulation from other people or objects (e.g. drives a car on his/her arm/leg, tickle his/her face with one of the toys or stimulus material; note: mouthing objects e.g. pencils, t-shirts, hijabs should not be coded here)

2=wants to be touched either by toys, materials or the examiner/parent a number of times (e.g. uses toys as blankets, uses examiner's hands to press his/her cheeks or wrap his/her body)

Auditory processing

Item 5. Response to hearing specific sounds (Hyper)

Item 1. Responds negatively to unexpected or loud noises (for example, cries or hides at noise from vacuum cleaner, dog barking, hair dryer)

Codes the child's response to a number of unexpected sound presses e.g. flushing toilet, dog barking, car horns, police siren, aeroplane

☐ 0=does not seem to be affected by the sound presses, carries out the task smoothly OR is surprised by sounds, but goes back to the task straight away

1=does not like at least one of the sound presses, shows some form of discomfort, such as facial grimacing OR verbally expresses the dislike of the sound stimulus, however, continues with the task

2=tries to eliminate or avoid at least one of the sound presses (e.g. covering the ears), OR after one of the sound presses verbally requests to stop OR switches the tablet off OR does not want to take part in the study OR shows anxiety

If coded 1 or 2, please, indicate to which stimuli the child reacted to:

☐ flushing toilet ☐ dog barking ☐ car horns

☐ police siren ☐ aeroplane

Item 6. Response to radio while playing on the tablet/colouring in (Hyper)

Item 3. Has trouble completing tasks when the radio is on

Codes the child's response to radio sounds

☐ 0=does not seem to be affected by the radio, carries out the task smoothly
1=does not like the radio, shows some form of discomfort, such as facial grimacing OR verbally expresses the dislike of the radio, however, continues with the task OR **gets distracted** (to the level of disengaging with the task) at least once
2=tries to eliminate or avoid the radio sound (e.g. covering the ears) OR verbally requests to stop OR switches the tablet off OR does not want to take part in the study OR shows anxiety OR cannot proceed with the task due to difficulty with concentrating

Item 7. Response to background noise (refrigerator, people talking, traffic noise) while further colouring in (Hyper)

Item 4. Is distracted or has trouble functioning if there is a lot of noise around

Item 5. Can't work with background noise (for example, fan, refrigerator)

Codes the child's response to background noise (social and non-social)

☐ 0=does not seem to be affected by the background noise, carries out the task smoothly
1=does not like the background noise, shows some form of discomfort, such as facial grimacing OR verbally expresses the dislike of the sound stimulus, however, continues with the task OR gets distracted at least once
2=tries to eliminate or avoid the background noise (e.g. covering the ears) OR verbally requests to stop OR switches the tablet off OR does not want to take part in the study OR shows anxiety OR cannot proceed with the task due to difficulty with concentrating

If coded 1 or 2, please, indicate to which stimuli the child reacted to:

☐ Refrigerator ☐ Traffic ☐ People talking

Notes:

……

Item 8. Response to hearing specific sounds and pointing to a picture that best matches the sound (Hyper)

Item 1. Responds negatively to unexpected or loud noises (for example, cries or hides at noise from vacuum cleaner, dog barking, hair dryer)

Codes the child's response to a number of loud sound presses e.g. flushing toilet, vacuum cleaner, hair dryer, dog barking, clock ticking, pencils/pens scratching, candy wrappers, car horns, train, alarm clock, police siren, balloon popping, fireworks, aeroplane

☐ 0=does not seem to be affected by the sound presses, carries out the task smoothly
1=does not like at least one of the sound presses, shows some form of discomfort, such as facial grimacing OR verbally expresses the dislike of the sound stimulus, however, continues with the task
2=tries to eliminate or avoid at least one of the sound presses (e.g. covering the ears), OR after one of the sound presses verbally requests to stop OR switches the tablet off OR does not want to take part in the study OR shows anxiety

If coded 1 or 2, please, indicate to which stimuli the child reacted to:

☐ flushing toilet	☐ pencils scratching	☐ police siren
☐ vacuum cleaner	☐ candy wrappers	☐ balloon popping
☐ hair dryer	☐ car horns	☐ fireworks
☐ dog barking	☐ train	☐ aeroplane
☐ clock ticking	☐ alarm clock	

Item 9. Response to name

Item 7. Doesn't respond when name is called but you know the child's hearing is OK

Codes the child's response to hearing his/her name

☐ 0=looks toward the examiner or verbally acknowledges his/her name being called (e.i. 'yeah') on at least one of the first two presses made by the examiner
1=looks toward the examiner's or verbally acknowledges his/her name being called (e.i. 'yeah') on third or fourth press of name only

2=does not respond in any way to the name being called or responds only when an interesting or familiar vocalization or verbalization is made (e.g., tongue clucking; 'I'm going to get you')

Item 10. Response to non-social sound – whistle (Hypo)

Codes the child's response to non-social sound

☐ 0=child's is visibly distracted, looks toward the examiner/parent or looks for the source of the sound OR asks about the sound (for example, 'What was that?')

Reaction:

☐ non-social ☐ social

2=does not seem to hear the stimuli

Item 11. Response to 'special' interest word (Hypo)

Item 6. Appears to not hear what you say (for example, does not "tune-in" to what you say, appears to ignore you

Codes the child's response to 'special' interest word

☐ 0=child's is visibly distracted from the task

Reaction:

☐ non-social ☐ social

2=does not seem to hear the stimuli

Not included: *Item 2. Holds hands over ears to protect ears from sound*

<u>Tactile processing</u>

Item 12. Sensitivity to certain fabrics (Hyper)

Item 33. Is sensitive to certain fabrics (for example, is particular about certain clothes or bedsheets)

Codes the child's sensitivity to certain fabrics (e.g cotton, wool, silk, stone, sand, wood, hay, sand paper, plastic, carpet, dried noodles, sticky tape)

☐ 0=does not seem to be sensitive to any particular fabric, carries out the task smoothly

1=shows some form of discomfort when touching at least one of the fabrics, such as facial grimacing OR verbally expresses the dislike of touching the fabric, however, continues with the task

2=tries to eliminate or avoid at least one of the fabrics (e.g. moving the hand away, moving quickly to another one) OR verbally requests to stop OR does not want to take part in the study OR shows anxiety

☐ leather	☐ label	☐ sticky tape
☐ sponge	☐ velvet	☐ feathers
☐ sandpaper	☐ suede	☐ wood
☐ fluffy towel	☐ plastic	☐ grass
☐ lace	☐ carpet	

Item 13. Response to finding an animal in 'messy' things (Hyper)

Item 29. Avoids getting "messy" (for example, in paste, sand, finger paint, glue, tape)

Codes the child's response to finding a plastic animal in 'messy' things (e.g. in sand, rice, dried noodles, salt dough, lotion)

☐ 0=does not seem to be avoiding getting 'messy', carries out the task smoothly

1=does not like to get 'messy', shows some form of discomfort, such as facial grimacing OR verbally expresses the dislike of getting 'messy', however, continues with the task

2=tries to eliminate or avoid at least one of the 'messy' stimulus (e.g. moving the hand away, skipping a container) OR verbally requests to stop OR does not want to take part in the study OR shows anxiety

Item 14. Response to being made 'messy' with lotion by the examiner (Hypo)

Item 46. Doesn't seem to notice when face or hands are messy

Codes the child's response to his/her arm being made messy by the examiner

☐ 0=wants to clean his/her arm straight away, looks for a towel/water or asks for it

1=seems to notice that his/her arms are messy, does not look for opportunity to clean them straight away, however, wants to clean his/he hands in a while

2= does not seem to notice when arms are messy, happy to take part in the next task while his/her hands are sticky/dirty

Item 15. Response to being 'messy' (Hypo)

Item 46. Doesn't seem to notice when face or hands are messy

Codes the child's response to his/her hands being messy

☐ 0=wants to clean his/her hands straight away, looks for a towel/water or asks for it

1=seems to notice that his/her hands are messy AND/OR does not look for opportunity to clean them straight away, however, wants to clean his/he hands in a while

2= does not seem to notice when hands are messy, happy to take part in the next task while his/her hands are sticky/dirty

Item 16. Response to standing/sitting close to others (Hyper)

Item 38. Has difficulty standing in line or close to other people

☐ 0=does not seem to be bothered by the close presence of the examiner

1=appears to be bothered by siting close to the examiner, but does not undertake any actions

2=has difficulty standing close to the examiner OR constantly moves away OR verbally asks the examiner to stay away or pushes the examiner away

☐ moves away ☐ moves towards

Item 17. Response to splashing water (Hyper)

Item 37. Withdraws from splashing water

Codes the child's response to splashing water

0=does not seem to be avoiding splashing water, is excited, surprised playful

1=does not like being splashed, shows some form of discomfort, such as facial grimacing OR verbally expresses the dislike of splashing

2=tries to eliminate or avoid splashing (e.g. moving away, covering eyes with hands) OR verbally requests to stop OR does not want to take part in the study OR shows anxiety

Item 18. Response to finding an animal in sand (Hyper)

Item 35. Avoids going barefoot, especially in sand or grass

Codes the child's response to being barefoot

0=does not seem to be bothered by the suggestion of being barefoot and happy to carry the task out being barefoot

1=needs some encouraging to be barefoot OR shows some form of discomfort, such as facial grimacing or verbally expresses the dislike of being barefoot, however, carries the task out and completes it

2=tries to eliminate or avoid the 'barefoot' stimulus (e.g. moving the foot away, skipping a container) OR verbally requests to stop OR does not want to take part in the task OR shows anxiety

Item 19. Response to putting the socks/shoes back on (Hypo)

Item 44. Avoids wearing shoes; loves to be barefoot

0=puts the socks/shoes on straight away

1=is not happy to put the socks/shoes back on (shows some facial grimacing or verbally expresses willingness to stay barefoot), however listens to the instructions

2=does not want to put the socks/shoes on, wants to stay barefoot or barefoot during the whole assessment time

Item 20. Over-response to unexpected touch (Hyper)

Item 36. Reacts emotionally or aggressively to touch

Item 39. Rubs or scratches out a spot that has been touched

Codes the child's over-response to unexpected touch

☐ 0=looks at the examiner/parent after being touched OR quickly rubs or scratches out a spot that has been touched

1=moves away slightly or shows some form of discomfort OR rubs or scratches out a spot that has been touched longer than expected

2= reacts emotionally or aggressively to unexpected touch or moves rapidly away or gets anxious or asks to stop being touched OR keeps rubbing or scratching out a spot that has been touched

Item 21. Under-response to unexpected touch (Hypo)

Item 43. Doesn't seem to notice when someone touches arm or back (for example, unaware)

Codes the child's lack of response to light touch

☐ 0=looks at the examiner/parent or on the spot that has been touched on at least one of the first two presses made by the examiner

1=looks at the examiner/parent or on the spot that has been touched on third or fourth press made by the examiner

2=does not seem to notice that has been touched