Heterogeneity of Social Approach Behaviour in Williams syndrome: The Role of Response Inhibition

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Abstract

The developmental disorder of Williams syndrome (WS) is associated with an overfriendly personality type, including an increased tendency to approach strangers. This atypical social approach behaviour (SAB) has been linked to two potential theories; the amygdala hypothesis and the frontal lobe hypothesis. The current study aimed to investigate heterogeneity of SAB in WS by exploring whether subgroups of SAB profiles could be identified using cluster analytic techniques. Twenty-five children with WS aged 6-15 years completed three behavioural tasks tapping i) social approach behaviour, ii) emotion recognition ability and iii) response inhibition. Cluster analyses revealed preliminary evidence of WS subgroups based on SAB profiles and indicated that response inhibition ability was the key differentiating variable between SAB cluster profiles. The findings provide tentative support for the frontal lobe hypothesis of SAB in WS and highlight the importance of investigating SAB at a heterogeneous level.

Keywords: Williams syndrome, social approach, inhibition, frontal lobe, amygdala

Abbreviations: SAB, Social approach behaviour

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1.1 Introduction

Williams Syndrome (WS) is a neurodevelopmental disorder, with estimated prevalence rates ranging from 1:20,000 (Morris & Mervis, 2000) to 1:7,500 (Stromme, Bjornstad & Ramstad, 2002) and is caused by the deletion of 25-28 genes on the long arm of chromosome 7 (7q11.23; Donnai & Karmiloff-Smith, 2000). The disorder is characterised by distinct, yet variable cognitive, physical and behavioural profiles (Hepburn, Fidler, Hahn & Philofsky, 2011). Most individuals with WS have mild to moderate intellectual difficulties (Searcy, Lincoln, Rose, Klima, Bavar & Korenberg., 2004); with verbal processing (Morris & Mervis, 1999) and certain aspects of language (Mervis & Klein-Tasman, 2000) identified as relative strengths within their cognitive profile. Specific areas of deficit include nonverbal processing and visuospatial skills (Farran & Jarrold, 2004). Individuals with WS have also been reported to display distractible behaviours (Dykens, 2003) and higher levels of anxiety than typically developing (TD) children and other groups with intellectual disabilities (Einfield, Tonge & Florio, 1997).

Hypersociability is frequently cited to be a defining feature of the social phenotype associated with WS (Järvinen-Pasley, Adolphs, Yam, Hill, Grichanik, Reilly & Bellugi, 2010) and has been described as a 'general presentation of extreme happiness' (Levine & Wharton, 2000; p.364); being 'unusually sociable, friendly and empathic' (Jones, Bellugi, Lai, Chiles, Reilly, Lincoln & Adolphs, 2000 p. 30), an excessive interest in others and a distinct lack of inhibition with regard to approaching others in social contexts (Bellugi, Järvinen-Pasley, Doyle Reilly, Reiss & Korenberg, 2007; Jones et al., 2000). Individuals with WS appear

hypersociable from an early age (Doyle, Bellugi, Korenberg & Graham, 2004), demonstrating an eagerness to make eye contact with and to approach strangers (Mervis & Klein-Tasman, 2000). An interest in looking at faces remains in childhood and into young adulthood (Riby & Hancock, 2008). Parents of children with WS often report concerns regarding the subsequent increased vulnerability and risk of exploitation that their children are exposed to as a result of their overfriendly behaviour and drive to approach strangers (Jones et al., 2000). This is especially relevant when considered alongside the developmental delay experienced by many individuals with the disorder (for a discussion of issues of social vulnerability see Jawaid, Riby, Owens, White, Tarar & Schulz, 2012). Developing an understanding of social approach behaviour (SAB) has been increasingly prioritised over recent years and two hypotheses have been proposed: the amygdala hypothesis and the frontal lobe hypothesis (Porter, Coltheart & Langdon., 2007). However, the literature on SAB in WS is fractionated by conflicting findings.

1.2 Amygdala Hypothesis

The amygdala hypothesis suggests that atypically large amygdala volumes and subsequent amygdala dysfunction play a role in the aetiology of atypical SAB in WS (Bellugi, Adolphs, Cassady & Chiles, 1999; Martens, Wilson, Dudgeon & Reutens, 2009). The amydgala is a limbic structure that guides socio-emotional behaviour, plays a role in the identification of facial emotional expression (Adolphs & Spezio, 2006), and is required for accurate social judgment of individuals on the basis of their facial expressions (Adolphs, Tranel & Damasio, 1998). Haas, Mills, Yam, Hoeft, Bellugi & Reiss (2009) reported findings of disparity in the amygdala functioning of individuals with WS compared to typical controls. The WS group demonstrated reduced amygdala reactivity in response to threatening faces and a heightened reactivity to happy expressions. It is suggested that the decreased amygdala activation to

threatening faces evident in individuals with WS indicates a reduced reaction to social danger and helps explain the social disinhibition and reduced fear towards strangers observed in this population (Bellugi et al., 1999; Martens, 2005). Amygdala volume as well as functionality is likely to be related to social behaviours in WS and likely to be atypical (Martens, Wilson, Dudgeon & Reutens, 2009). Martens et al. (2009) investigated the relationship between amygdala volume and approachability ratings in individuals with WS compared to TD controls. The findings revealed a significant relationship between increased volumes and higher approachability ratings in WS to both 'negative' faces and 'positive' faces which supports this hypothesis. However, Frigerio et al. (2006) and Porter, Coltheart & Langdon (2007) found that individuals with WS rated only the 'positive' faces as more approachable than controls whilst 'negative' faces were rated as less approachable. They concluded that individuals with WS are able to discriminate the approachability of individuals and their SAB was not a function of underlying emotion recognition difficulties.

1.3 Frontal Lobe Hypothesis

The frontal lobe hypothesis postulates that the atypical SAB in WS may result from impairment in response inhibition subsequent to frontal lobe dysfunction (Porter et al., 2007). Porter et al. (2007) describes the similarities in the atypicalities of SAB in WS and the SAB of patients with frontal lobe damage, and state that both groups seem to demonstrate a dissociation between 'knowing' and 'doing' which is reflected by their tendency to approach strangers in day-to-day life. Porter et al suggest that individuals with frontal lobe damage 'know' that they shouldn't talk to or approach strangers but still go ahead and do so due to poor impulse control. Furthermore, several studies report neurological evidence to suggest that frontal lobe abnormalities do exist in WS (Meyer-Lindenberg et al., 2005, Mobbs et al., 2007) and behavioural tasks show evidence of executive functioning difficulties similar to

those seen in individuals with ADHD (e.g. Rhodes, Riby, Matthews, & Coghill, 2011). Mobbs et al. (2007) used functional magnetic resonance imaging (fMRI) to investigate frontal lobe activation and found that the WS group demonstrated reduced frontostriatal activation compared to TD controls during a response inhibition task. They suggest that individuals with WS display a generalised deficit in response inhibition which subsequently impacts upon their behaviour in social situations.

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Porter et al., (2007) investigated SAB in WS in relation to the theories discussed. They found that WS participants displayed emotion recognition abilities that were appropriate to their general level of cognitive functioning and did not display atypical responses on the social approach task. However, performance on a response inhibition task was well below the level expected on the basis of their mental age or level of intellectual functioning. They therefore concluded that the tendency for WS individuals to approach strangers in everyday life may be due to poor response inhibition.

1.5 Heterogeneity of social behaviours in Williams syndrome

Research to date has focussed on describing SAB in WS as a homogenous construct. However, Porter et al. (2007) and Järvinen-Pasley et al. (2010) observed substantial variability in the approachability ratings given by individuals with WS which differed from the consistency of ratings demonstrated in the TD control groups. Many developmental disorders are heterogeneous (Abrahams & Geschwind, 2008) and this is a particularly pertinent feature of WS, with cognitive, social, genetic and physical characteristics varying considerably from one individual to the next (Porter & Coltheart, 2005). Porter and Coltheart, (2005) challenged the notion of a "syndrome specific" (Howlin, Davies & Udwin, 1998) WS cognitive profile and suggested that subgroups might exist within WS based on their similarities in cognitive profile. They discovered evidence for two groups differing in terms of perception, attention and spatial construction abilities and differences in social-emotion skills. Subgroup one displayed a perceptual integration deficit, but good spatial construction abilities, response inhibition and emotion perception abilities, whereas subgroup two showed good perceptual integration skills, but poor spatial construction abilities, poor response inhibition and poor emotion perception abilities. This research supports the notion of cognitive and social heterogeneity in WS, furthermore, the heterogeneity did not appear to reflect differences in degrees of impairment but rather distinct patterns of strength and weaknesses (Porter et al., 2007).

Järvinen-Pasley et al. (2010) highlighted that future research should elicit the sources and extent of variability in social behaviour in the WS population and look for explanations which go beyond the assumption of a relatively homogenous syndrome profile of social cognition in individuals with WS.

Given the suggested heterogeneity of SAB in WS, the current study aims to examine this variability by exploring performance on a range of tasks assessing social salience, emotion recognition and response inhibition to determine whether there is evidence for clusters of SAB based on these constructs. It is hypothesised that if the amygdala hypothesis provides an accurate account for the SAB seen in WS, clusters will be characterised on the basis of emotional recognition ability, whereas if the frontal lobe hypothesis provides a more accurate account of the phenomena clusters will be characterised by response inhibition abilities.

2.0 Method

2.1 Participants

Twenty five children with WS aged between 6- and 15-years-old (mean age 9.5years; SD 8.95; 12 male, 13 female) participated in the research. All children had previously been diagnosed with WS using genetic testing (fluorescent *in situ* hybridization testing; FISH) and clinical diagnosis. The mean estimated Full Scale IQ (FSIQ) using the Wechsler Intelligence Scale for Children – Third Edition (WISC-III) Short Form (Wechsler, 1991) was 54.7 (sd 8.95), mean verbal IQ was 65.6 (sd 8.95) and mean performance IQ was 50.58 (sd 6.77). Parental consent was received prior to participation for all children and the study had received favourable ethical approval from the local ethics committee.

2.2 Materials

2.2.1 Social Approach Behaviour

Adolph's Approachability Task (Adolph, 1998) provided a measure of an individual's willingness to approach an unfamiliar person. Photographs of unfamiliar faces were presented to participants and they were asked to rate how much they would like to approach the person on a 5 point likert scale (ranging from 0= not at all to 4= yes definitely). The task has been used in numerous studies with individuals who have WS (Bellugi et al., 1999; Martens et al., 2009; Järvinen-Pasley et al., 2010).

2.2.2 Emotion Recognition Task

The emotion recognition task used face stimuli from Ekman and Friesen (1976) with 30 pictures of faces used as stimuli and participants required to make a forced choice to determine whether the face looks happy, sad, angry, surprised, scared or neutral (thus tapping

basic expressions of emotion). This task has been used previously with individuals who have WS (Järvinen-Pasley et al., 2010). The task was self-paced and stimuli remained on screen until a response was provided.

2.2.3 *Response Inhibition Task*

The Sun-Moon Stroop task (Archibald & Kerns, 1999) is a pictorial modification of the Stroop Test, developed for pre-literate children (Pasalich, Livesey & Livesey 2010). In condition A, participants are shown a single page consisting of 30 sun and moon pictures which have been randomly arranged into equal rows and columns. They are instructed to respond "sun" to the pictures of the suns, and "moon" to the pictures of the moons, as fast as they can (within a 45-second time limit) and to correct themselves if they made a mistake before moving on. The experimenter points at each picture as it is named and if a participant makes an error on a picture, the experimenter leaves their marker on this picture until the participant corrects themselves. If they name all the pictures on the page within the given time limit, participants are asked to start from the top again. As a practice trial, children are asked to name the first four pictures. In condition B, participants are asked to respond “sun” to pictures of moons and “moon” to the pictures of the sun, thus having to inhibit their initial response. Archibald and Kerns (1999) found that the task correlated significantly with other measures of inhibitory control and had high test-retest reliability (.86) among 7- to 12-year olds. To the authors knowledge the task has not been used with WS, although alternative Stroop tasks have been used (Menghini et al., 2010).

2.2.4 Intellectual Ability

A short form of the Wechsler Intelligence Scale for Children – Third Edition (WISC-III; Wechsler, 1991) was used to assess intellectual functioning. The Similarities, Vocabulary,

Picture Arrangement and Block Design subtests were administered. This collection of subtests has been recommended by Minshew, Turner and Goldstein (2005) as the most extensive short form for individuals with autism and has been previously used in a study with children with WS (Rodgers, Riby, Janes, Connolly & McConachie, 2012).

2.3 Procedure

Due to the similarity of the Emotion Recognition Task and Adolph's Approachability Task the order of administration of these tasks alternated to control for the potential confounding effects of one task on the other. Therefore each child first completed either the Emotion Recognition Task or Adolph's Approachability Task then a short form of the WISC-III was administered. Following this either the Emotion Recognition Task or Adolph's Approachability Task was completed and finally all children completed the Sun & Moon Task was completed. Data collection took approximately 30-45 minutes to complete.

2.4 Data Analysis Strategy

Cluster analyses are mathematical methods that can be used to find out which objects in a group are similar (Romesburg, 2004). Cluster analysis methods group 'objects' based on their similarities along one or more constructs of interest, and their dissimilarities from the objects in other groups (Steele et al., 2007). Cluster analytic techniques are often used to identify patterns of differences across multiple measures at a single point in time (cross-sectional) and the clusters that are formed can then be used for descriptive purposes. This approach was considered suitable to examine variability in SAB within WS. Our aim was to determine whether clusters of different SAB profiles could be identified from the data. Cluster analysis can be used with small sample sizes and has been used with samples of participants with developmental disorders e.g. Barton et al. (2004). It has been used

previously to investigate sensory abnormalities in children with WS (John et al., 2010). The variables used for cluster analysis here were selected on the basis of predictions from theories of SAB; emotion recognition and response inhibition. Chronological age and IQ were also included to investigate whether developmental stage masked the effect of other key variables. It is acknowledged that any conclusions that are drawn from cluster analysis and applied to a population must be based on analogy and not inference (Romesburg, 2004).

3.0 Results

3.1 Approachability

Approachability behaviour was measured using Adolph's Approachability Task (Adolphs, Tranel, & Damasio, 1998). During this task participants were required to make judgments about 20 photos of unfamiliar people in terms of how much they would like to approach the person on a 5 point scale ranging from, 0-4 (0; no, 4; yes, definitely). The mean approachability rating given by participants was 3.05 (SD 0.73, range 1.9 to 4). Table 1 compares participant's ratings of the stimuli to the norms derived from the pre-ratings of the stimuli (Adolphs et al., 1998), as well as to previous studies (Jones et al., 2000; Martens et al., 2009) which used the task with participants with WS. Mean scores are reported for the pre-rated 'positive' and 'negative' faces. Previous studies had used a five point likert scale ranging from - 2 to + 2 rather than 0 to 4 therefore data from the current study were transformed to enable comparison.

Table 1

3.2 Emotion Recognition (ER) Task

This task sought to address the ability to recognise facial affect. Participants were shown photographs of faces depicting various emotions and were required to make a forced choice about how they thought the person in the photograph was feeling. A summary of the results are shown in table 2.

Table 2

Recognition errors were specifically evident on the disgusted, surprised, scared, and neutral expressions.

3.3 Response Inhibition Task

The Sun-Moon Stroop Task (Archibald & Kerns, 1999) was used as a measure of inhibitory control. In condition (A), participants are instructed to respond "sun" to pictures of the suns, and "moon" to pictures of the moons, in condition B they are required to respond “sun” to pictures of moons and “moon” to pictures of suns within a 45-second time limit. An interference score (no. of items completed (B) - no. of items completed (A)}/ {no. of items completed) was calculated for each child, where higher scores indicate less interference. The mean interference score was –0.28, (SD; 0.24).

3.4 Cluster Analysis

Participants were the ‘objects’ subjected to cluster analysis using ClustanGraphics (Wishart, 2006). This software was chosen for the benefits of being able to handle heterogeneous data. By standardising the data, the package strips the identity from each attribute, changes its

numerical value, and recasts it in dimensionless form in order to ensure that each variable in the data is given appropriate weight in the analysis and removes any difficulties created by comparing attributes that are measured using different scales. Data were subjected to cluster analysis across the three key variables, as predicted by the models of SAB in WS: emotion recognition and response inhibition.

The Squared Euclidean distance was used as a proximity coefficient to measure the overall resemblance (the degree of similarity) between each pair of objects and create a resemblance matrix. In order to provide a visual representation of the degree of similarity between all pairs of objects, the resemblance matrix was used to create a tree using Ward's [30] minimum variance clustering method. This method has been used in previous studies with similar samples e.g. autism spectrum disorder (Hrdlicka et al., 2005) and has shown to be effective when applied to behavioural data. To explore the best cluster solution we used the Bootstrap Validation procedure available in ClustanGraphics. In this procedure the proximity matrix is randomized and compared to the obtained proximity matrix, highlighting the cluster solutions which significantly differ from random. The Bootstrap Validation procedure indicated that the greatest departure from a random pattern occurred at four clusters. Table 3 shows the profiles of each cluster.

Table 3

Cluster four comprised four participants who demonstrated the highest interest in approaching others as measured by Adolph's approachability task (1999). Participants in this group had the lowest IQ scores and also demonstrated the poorest response inhibition abilities which suggest that both of these variables may be associated with increased approachability behaviour. However, examination of the remaining cluster profiles indicated that response

inhibition appeared to make a stronger contribution in distinguishing between the clusters than IQ. For example, cluster one and three had very similar IQ scores, yet substantially different approachability scores and these clusters could best be differentiated on the basis of the response inhibition score. Cluster two were the lowest approachability group and contained participants who were the most able to inhibit responses.

Further cluster analyses explored cluster solutions when age and IQ were removed as variables in order to establish if developmental variables were masking the effect of the other variables. These solutions supported the original cluster solution. With age and IQ removed, bootstrap validation revealed that the greatest departure from random occurred at a five cluster solution with the high approachability clusters (cluster three, four and five) demonstrated poor scores on the response inhibition task. Scores on the emotion recognition task were much less indicative of approachability, e.g. cluster three and four demonstrated very similar response inhibition scores and approachability ratings, yet substantially different emotion recognition scores which would seem to indicate that response inhibition is a key variable in determining approachability behaviour.

4.0 Discussion

The primary aims of this study were to examine SAB in WS in the context of the amygdala and frontal lobe hypotheses and to examine variability by exploring whether clusters of different SAB experiences can be detected.

In relation to the Approachability Task the data are to some degree consistent with Jones et al. (2000) and Martens et al. (2009) who report WS participants rating both the positive and negative stimuli higher than the normative ratings established with adult participants in Adolphs et al. (1998) study. In the current study, participants rated the positive stimuli similarly to participants with WS in the studies by Jones et al. (2000) and Martens et al. (2009). However the ratings for the negative stimuli were rated more favourably which indicates that participants in this study were more willing to approach the negative faces than the participants in studies by Jones et al. (2000) and Martens et al. (2009).

The results support previous findings with regard to social approach behaviour in WS (Bellugi et al., 1999; Martens et al., 2009; Capitao et al., 2011) that individuals with WS report a high willingness to approach unfamiliar faces. Consistent with Jones et al. (2000), Porter et al. (2007) and Martens et al. (2009) the findings here revealed that participants rated positive faces as more approachable than negative faces. The mean approachability ratings for positive faces are similar to Jones et al. (2000) and Martens et al. (2009).

4.1 The Amygdala Hypothesis

In this sample emotion recognition errors were specifically evident on the disgusted, surprised, scared, and neutral expressions. This pattern of findings is consistent with Plesa-

Skwerer et al., (2005), Järvinen-Pasley et al. (2010) and Gagliardi et al. (2003) who also used the Ekman and Friesen emotion recognition task with WS participants. These data suggest that while the ability to perceive more basic expressions of emotion is relatively good in individuals with WS, difficulties can occur identifying more complex or subtle emotions. These studies also report that the performance of participants with WS is comparable to that of mental age matched controls and suggest that these difficulties are best understood in a developmental context rather than being syndrome specific. Previous studies (Meyer-Lindenberg et al., 2005; Haas et al., 2009) have reported that individuals with higher facial affect identification abilities tend to be more discriminative of unfamiliar people and therefore will be less inclined to approach them. However, findings here indicate that emotion recognition scores were not associated with SAB. Although participants in this sample of individuals with WS can recognise facial affect at an appropriate developmental level, this ability does not determine their social approach behaviour. These findings highlight the need for further investigation into social-perceptual abilities of individuals with WS that go beyond emotion recognition ability and their link to SAB.

4.2 Frontal Lobe Hypothesis

To the author's knowledge, the sun-moon task has not been used previously on WS population. However, Pasalich, Livesey & Livesey (2010) used the task with typically developing children (aged 4-5 years old) and similar interference scores were reported. The cluster analyses provide tentative support for the response inhibition hypothesis of SAB. Response inhibition ability appeared to be the key differentiating variable between clusters. The 'high approachability' clusters contained participants who demonstrated the poorest response inhibition abilities, whereas participants who demonstrated greater ability on the response inhibition task were clustered in the 'low approachability' groups. These findings

are consistent with Porter et al. (2007) and Jarvinen-Pasley et al. (2010) who interpreted their findings as best supporting the frontal lobe hypothesis.

4.3 The impact of developmental stage

In order to investigate the effect of developmental variables (age and IQ) on the cluster solutions, further analyses explored cluster solutions when age and IQ were removed as variables. These solutions supported the original cluster and indicated that response inhibition remained the most significant variable in determining SAB. With age and IQ removed, bootstrap validation revealed that the greatest departure from random occurred at the five clusters solution. Similarly the high approachability clusters (cluster three, four and five) demonstrated poor scores on the response inhibition task. Scores on the emotion recognition task were much less indicative of approachability, e.g. cluster three and four demonstrated very similar response inhibition scores and approachability ratings, yet substantially different emotion recognition scores. These findings support previous studies (Mobbs et al., 2006; Porter et al., 2007; Menghini et al., 2010) which have proposed that deficits in inhibition are a key executive characteristic for individuals with WS and that SAB in WS may be linked to an inhibitory deficit for social responses (Jones et al., 2000; Frigerio et al., 2006; Porter et al., 2007).

4.4 Heterogeneity in WS

It is important to note that substantial variability was found for the approachability ratings provided by participants. These findings support studies which have demonstrated heterogeneity in WS (Järvinen-Pasley et al., 2010; Porter et al., 2007; Stojanovik, Perkins & Howard, 2006; Porter & Coltheart, 2005). Interestingly there was variability in age and IQ within clusters, indicating that developmental variables did not consistently or reliably predict

SAB profiles. The results highlight the importance of investigating SAB at an individual level rather than looking at group means. Heterogeneity in WS independent of age and IQ is consistent with reports of varying genetic patterns, varying physical features, and clinical variability within the syndrome (Borg et al., 1995; Fryssira et al., 1997; Pankau et al., 2001). Although it is important that these findings are interpreted with caution due to the exploratory nature of the analysis and the small sample size, they do offer a preliminary suggestion of WS subgroups based on SAB profiles.

4.5 Social Salience Hypothesis

A further hypothesis postulated to account for the SAB in WS but not examined here is the social salience hypothesis. This hypothesis proposes that social stimuli, e.g. faces, are more salient for individuals with WS (Porter et al., 2007) and that this motivates their SAB. Following observations that individuals with WS tended to look intensely at researchers during experimental procedures (Jones et al., 2000; Mervis et al., 2003), Frigerio et al. (2006) proposed that individuals with WS have high 'social stimulus attraction' (p.258) and that this drives their SAB. Research using eye-tracking methodology has provided experimental evidence that individuals with WS tend to look at faces for extended periods (Riby & Hancock., 2008) in particular the eye region (Mervis et al., 2003; Riby & Hancock 2008) which further supports this hypothesis. However, Dodd and Porter (2010) employed an observational paradigm to investigate the role of the face in motivating SAB in WS and found that that the face did not need to be visible for WS children to display atypical SAB. They concluded that attraction to the face may not be the principal motivating factor of SAB. Future research examining individual variability in SAB in WS should incorporate social salience variables.

4.5 *Strengths and Limitations*

This is the first known study to use cluster analysis to investigate SAB in WS and this analytic approach overcomes many of the limitations faced by previous studies e.g. enabling investigation of variability within the syndrome. However, it is important to recognise that the exploratory nature of the analysis is subject to experimenter bias vulnerable to results that are biased in the direction of the 'framing decisions' (Romesburg, 2004) made by the experimenter throughout the analytic process. Framing decisions refer to the choices made that shape or frame the data from it input to output (Wishart, 2006). If these decisions are made differently, the output (clusters) will be different. In order to reduce the risk of bias, the researcher selected the variables for the cluster analysis from existing theories of SAB. Triangulation of data also enhanced the robustness and validity of the findings.

Due to the inconsistencies in the SAB literature, researchers have questioned the ecological validity of approachability tasks. Future research should further examine the reliability and validity of this task and seek to develop a robust and standardised measure of SAB.

The small sample size of the current study has implications for how well the findings can be generalised to the WS population. Cluster analysis techniques are increasingly used with neurodevelopmental research in order to capture heterogeneity and can be used with comparable small sample sizes to the one reported here e.g. Barton et al. (2004) and John et al. (2010). In order to avoid a type one error, variables used for cluster analysis were selected on the basis of predictions from underlying theories of SAB. Given the planned data analytic strategy of employing cluster analysis, a power calculation was not possible for this study. In comparison to other research employing cluster analysis techniques with populations with developmental disorders e.g. Barton et al. (2004) who used a **s**ample of 24, the current sample was deemed adequate for the exploratory nature of the study. In order to further

maximise the generalisability of the findings, the age range of participants was restricted to 6-15. Whilst still broad this age range is narrower than other SAB studies with WS participants, which included wider age ranges, e.g. Porter et al. (2007; age range 5 years to 43 years), Martens et al. (2009; age range 8 to 41 years), Jarvinen-Pasley et al. (2010; age range 13 to 53 years) it is nevertheless acknowledged that the range included in the current study remains broad. Future studies should investigate approachability profiles with larger WS samples and with different age ranges in order to begin to establish a robust developmental trajectory of SAB profiles.

4.6 Conclusion

This is the first known study to investigate the heterogeneity of SAB in WS. The findings demonstrate that substantial variability exists in the social profile of individuals with WS. Cluster analysis revealed preliminary evidence of subgroups of WS based on their SAB profiles and indicated that response inhibition ability is the strongest indicator of SAB. Interestingly the results were not masked by developmental variables. The results provide tentative support for the frontal lobe hypothesis. The findings highlight several important directions for research which will be essential in furthering our understanding of this phenomenon and for the development of effective assessments and interventions.

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Table 1: Approachability Ratings for Unfamiliar Faces on the Adolph's Task

Study	N	Overall Mean	SD	Mean Score Positive faces	SD	Mean Score Negative faces	SD
Current Study (transformed scores)	25	0.97	0.47	1.28	0.36	0.66	0.35
Adolph's (1999) Normative data (TD adults)	26	-0.03	1.29	1.06	0.65	-1.12	0.65
Jones et al (2000) WS	26			1.32	1.1	-0.54	1.39
Martens et al (2009) WS	27			1.20	0.53	-0.50	-0.66

Table 2: Performance on the Emotion Recognition task

Correctly Identified	**Overall**	**Happy**	**Sad**	**Angry**	**Scared**	**Disgusted**	**Surprised**	**Neutral**
					(max score = 5)			
Mean	17.92	5	3.48	2.96	2.12	0.88	2.08	1.68
SD	3.29	0	1.26	1.14	1.45	1.2	1.75	1.63
Range	12-25	0	0-5	0-5	0-5	0-4	0-5	0-5

Table 3. Cluster Solutions.

Cluster	N	Mean approachability Rating*	Age	Mean FSIQ	Mean emotion recognition score	Mean response inhibition score
1	8	2.74	7.38	62.63	15.25	-0.18
2	5	2.13	12	54.20	17.80	-0.06
3	8	3.21	10.88	52.63	21.25	-0.32
4	4	3.95	9.25	41.25	16.75	-0.66

- Higher score is indicative of more willingness to approach