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Autonomic response to approachability characteristics, approach behavior, and social functioning in Williams syndrome

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ABSTRACT

Williams syndrome (WS) is a neurogenetic disorder that is saliently characterized by a unique social phenotype, most notably associated with a dramatically increased affinity and approachability toward unfamiliar people. Despite a recent proliferation of studies into the social profile of WS, the underpinnings of the pro-social predisposition are poorly understood. To this end, the present study was aimed at elucidating approach behavior of individuals with WS contrasted with typical development (TD) by employing a multidimensional design combining measures of autonomic arousal, social functioning, and two levels of approach evaluations. Given previous evidence suggesting that approach behaviors of individuals with WS are driven by a desire for social closeness, approachability tendencies were probed across two levels of social interaction: talking versus befriending. The main results indicated that while overall level of approachability did not differ between groups, an important qualitative between-group difference emerged across the two social interaction contexts: whereas individuals with WS demonstrated a similar willingness to approach strangers across both experimental conditions, TD individuals were significantly more willing to talk to than to befriend strangers. In WS, high approachability to positive faces across both social interaction levels was further associated with more normal social functioning. A novel finding linked autonomic responses with willingness to befriend negative faces in the WS group: elevated autonomic responsivity was associated with increased affiliation to negative face stimuli, which may represent an autonomic correlate of approach behavior in WS. Implications for underlying organization of the social brain are discussed.

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1. Introduction

One powerful method for elucidating the underpinnings of human sociality is to utilize a genetically based disorder associated with altered social functioning as a model. Of particular interest to this line of investigation, Williams syndrome (WS) is a multisystem disorder (Pober, 2010), resulting from a hemizygous deletion of 25–30 genes on chromosome 7q11.23 (Ewart et al., 1993). WS is associated with a unique social phenotype saliently characterized by increased motivation for social interaction and approach (e.g., Doyle et al., 2004; Järvinen-Pasley et al., 2008; Frigerio et al., 2006), which may stem from difficulties with inhibition (Little et al., 2013). The pro-social drive of WS as reflected through a strong affinity toward unfamiliar people has been established through assorted methodologies and paradigms. At the behavioral level, such include observations (Klein-Tasman et al., 2007; Klein-Tasman and Mervis, 2003; Järvinen-Pasley et al., 2008), questionnaires (Doyle et al., 2004), eye tracking approaches (Riby and Hancock, 2008, 2009), and various experimental designs, which have, e.g., compared the willingness of individuals with WS and typically developing (TD) participants to approach strangers (e.g., Bellugi et al., 1999; Frigerio et al., 2006; Martens et al., 2009; Järvinen-Pasley et al., 2010; Martens et al., 2012). However, it is important to emphasize that despite the robustly established "hypersociability", considerable heterogeneity in multiple domains of functioning exists in WS, in e.g., cognition (perception, attention, spatial construction, social-emotional ability) (Porter and Coltheart, 2005) and social behavior (social approach tendency, response inhibition) (Little et al., 2013; Riby et al., 2014a).

Collectively, investigations employing "approachability tasks" have provided mixed findings, suggesting that approach behavior may not be entirely indiscriminate in WS. These tasks typically

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require participants to evaluate on a Likert-type scale how much they would like to approach a person in a facial image, which have been pre-rated for approachability characteristics (e.g., trustworthy/untrustworthy-looking; positive/negative emotional displays). Some studies have reported more positive approachability judgments in WS relative to both chronological age (CA)- and mental age (MA)-matched controls in response to both positively and negatively pre-rated faces (Bellugi et al., 1999; Jones et al., 2000; Järvinen-Pasley et al., 2010; Martens et al., 2009). By contrast, in one investigation, face stimuli displayed positive and negative standard expressions (happiness, fear, anger, disgust, sadness, neutral), and elevated approachability in WS relative to CA- and MA-matched controls was solely evident in relation to the people displaying positive emotion (Frigerio et al., 2006). In a recent study using a mouse-tracking paradigm to examine on-line trustworthiness evaluations of unfamiliar faces (Martens et al., 2012), individuals with WS relative to a CA-matched TD control group showed significantly elevated willingness to approach untrustworthy-looking people. Additionally, a set of studies have examined linkages between approachability ratings and emotion identification skills, and found that atypical approach ratings in individuals with WS were related to deficits in social perception (Järvinen-Pasley et al., 2010; Porter et al., 2007).

Extending the line of work described above, a recent study directly targeted “stranger danger” awareness and perceptions in individuals with WS using video vignettes and pre-determined questions to probe the understanding of interactions with strangers (Riby et al., 2014a). The results suggested that overall participants with WS exhibited difficulties in making trust evaluations and deciding whether to talk with the unfamiliar protagonist. The participants with WS who exhibited decreased awareness of danger also demonstrated difficulties in peer relationships and dysfunctional pro-social behavior. Another recent study reported an interesting qualitative motivational difference between individuals with WS and TD in social approach, namely, whereas high approachability in individuals with WS appeared to be driven by a desire for close interpersonal relations, TD participants demonstrated pro-social behavior with the purpose of exerting social dominance over others (Ng et al., 2014). Taken together, the evidence reviewed above suggests that while the robust appetitive social drive of individuals with WS is motivated by a desire to form relationships, inappropriate social engagement occurs at least partially because of diminished ability to socially evaluate others based on relevant characteristics and contextual cues.

Of importance here is to consider how individuals with WS may understand different types of interpersonal relationships. Using parental reports, a large-scale study by Elison et al. (2010) that included a sample of 92 adults with WS showed that approximately 30% of these individuals had no skills to form friendships and about 50% showed limited grasp of the concept of friendship. Despite this, approximately 40% of the participants were reported to enjoy good quality friendships, encompassing at least one friend of own age. While approximately 30% of the sample was described as showing adequate understanding of intimate relationships, only 12 individuals had experience of such. In line with these observations, Jawaid et al. (2011) state that, “individuals with WS experience overly problematic peer interactions and unstable relationships, despite their friendly demeanour” (p.339), and Plesa Skwerer et al. (2004) also noted that it is very rare of individuals with WS to have actual friendships, let alone a person whom to call a “best friend” (see also Gosch and Pankau (1997)). The picture of WS with respect to relationship understanding is in fact similar to that reported for individuals with other developmental disability conditions. In this vein, Jobling et al. (2000) have postulated that in case of persons with developmental disabilities, relationships are commonly misleadingly and inappropriately classified as “friendships” when they clearly fail to fulfill the concept for such. This pertains to relationships that are clearly superficial or purely instrumental involving support personnel, family friends, and facilitators. Taken together, it is clear that the majority of affected individuals do not show normal understanding of relationships, which may on the other hand be fully expected on the basis of their social-cognitive impairments encompassing the theory of mind (Tager-Flusberg and Sullivan, 2000).

Recent advances from brain-imaging studies have elucidated the neural correlates of increased approach behavior in WS, and as a result, two major hypotheses of the increased approach behavior have been proposed. First, the amygdala hypothesis postulates that alterations in the amygdala structure and/or function and its connectivity with the orbitofrontal cortex (OFC) underpin the major social features of WS (Haas et al., 2009; Meyer-Lindenberg et al., 2005; Reiss et al., 2004). The role of the amygdala in the perception of emotional facial expressions is well established (Adolphs, 2003; Herba and Phillips, 2004), and bilateral amygdala damage has been linked to atypically positive approachability judgments in response to untrustworthy-looking or negative faces (Adolphs et al., 1998). Studies of individuals with WS have reported drastically diminished amygdala activation in response to threatening faces (Meyer-Lindenberg et al., 2005) and increased activation to threatening non-social scenes (Meyer-Lindenberg et al., 2005; Thornton-Wells et al., 2011). In a similar vein, Haas et al. (2009) reported decreased amygdala activation in response to fearful faces, and increased activation to happy facial expressions, in participants with WS as contrasted with TD controls. Subsequently, it has been suggested that the amygdala dysfunction in response to threatening stimuli indexes diminished recognition of social danger, and thus is linked to the disinhibited behavior in social settings (Bellugi et al., 1999; Martens et al., 2009).

Two magnetic resonance imaging (MRI) investigations have specifically examined amygdala features in tandem with approachability tendencies in individuals with WS. First, Martens et al. (2009) related amygdala volume to approachability ratings using the Adolphs Approachability task (e.g., Bellugi et al., 1999; Jones et al., 2000; Järvinen-Pasley et al., 2010). Consistent with previous studies, individuals with WS relative to TD demonstrated increased amygdala volume and elevated approachability ratings in response to both positively and negatively pre-rated faces. Moreover, higher approachability ratings in response to negative faces were positively related right amygdala volume in the WS group, providing support to the amygdala hypothesis. In the other study, Haas and colleagues utilized the Salk Institute Sociability Questionnaire (SISQ), a parental report tapping into approach tendencies, and an implicit task testing facial expression processing in combination with functional MRI (fMRI) (Haas et al., 2010). The results showed that in individuals with WS, decreased amygdala activation to fearful facial expressions was linked to an amplified tendency to approach strangers. The authors concluded that the evidence supported the idea that abnormal amygdala response to fear is indeed associated with dysregulated social behavior in WS.

The second hypothesis posits that the increased approach behavior is underpinned by frontal lobe dysfunction resulting in impaired response inhibition. This postulation is founded upon the finding that the striatum is implicated in decision-making outcomes of social interactions related to social approval/rejection judgments. Mobbs et al. (2007) found decreased frontostriatal activation during a non-social response inhibition task and hypothesized that this may also be linked to the uninhibited social affiliation in WS, reflecting a generalized deficit. In a similar vein, Porter et al. (2007) tested participants with WS and controls on a battery comprising behavioral emotion recognition, social approach, and response inhibition tasks. While results indicated that their emotion-processing performance was consistent with mental-age level and their pattern of approachability ratings was similar to the controls, those with WS performed at a lower level than expected on the basis of their MA on the response inhibition task. The authors interpreted their results as suggesting that increased approachability in WS was linked to poor response inhibition stemming from frontal lobe dysfunction.

The "hypersocial" nature of individuals with WS described above camouflages panoply of deficits in social-perceptual skills, social-cognition, and communication, as well as maladaptive behaviors impacting daily living skills. Indeed, the increased affiliative behavior of individuals with WS has been suggested to predispose such individuals to vulnerability to exploitation and abuse (Fisher et al., 2014a). As social evaluations are intimately linked to decisions to approach others, it is important to consider how these processes may be altered in WS. Social interactions are modulated by social norms and expectations, which facilitate the learning of reputation-related information from others (Rilling and Sanfey, 2011). Trust is founded on the idea that generous or kind behavior toward others will be reciprocated, and it is a crucial ingredient for the development of meaningful social relationships (van der Bos et al., 2009). Crude judgments of trust are made rapidly (e.g., Adolphs et al., 1998; Todorov et al., 2008), although decisions about pursuing deeper social relationships involve repeated exposures and "trial-and-error" learning (Chang et al., 2010).

A recent line of relatively systematic investigation has focused on profiling social reciprocity skills in WS by employing the Social Responsiveness Scale (SRS; Constantino and Gruber, 2005). Klein-Tasman et al. (2011) reported more pronounced deficits in social cognitive (communication and cognition) than to pro-social (social awareness and motivation) functions in participants with WS. Riby et al. (2014b) reported that less than 20% of their WS sample covering a broad age range scored within the normative range on the SRS. Finally, van der Fluit et al. (2012) explored associations between social cognition, social perception, and social communication in young persons with WS. On the SRS, the most profound impairments were found in social cognition, while social motivation appeared unimpaired in those with WS. The results further indicated that participants with WS who performed similarly to TD individuals in interpreting ambiguous social dynamics also exhibited decreased problems in real-life social reciprocity. These associations remained after controlling for intelligence, suggesting that problems with interpreting social situations may be pivotal in interpersonal difficulties of individuals with WS. On the basis of the above evidence, it seems plausible that heterogeneity in social affiliative tendencies in WS (Little et al., 2013; Riby et al., 2014a) stems from not only individual differences in social reciprocity, but also from the ability to interpret social information including cues to others' approachability characteristics (Bellugi et al., 1999; Martens et al., 2009, 2012; Jones et al., 2000; Porter et al., 2007; Frigerio et al., 2006; Järvinen-Pasley et al., 2010). For example, within the domain of social perception, Martens et al. (2009) found that when making approachability judgments, individuals with WS appeared to utilize peripheral (hair, earrings, distinctive physical marks) as compared to eye/mouth region of the face stimuli more frequently than age-matched controls, although the use of mouth and eye features increased with age in WS.

In light of the widespread alterations of the social brain in WS (Järvinen et al., 2013; Haas and Reiss, 2012), further insight into the social drive in WS can be derived from approaches that employ social interaction paradigms and physiological indices modulated by the hypothalamic–pituitary–adrenal (HPA) axis. The HPA axis is responsive to an array of social-emotional behaviors (Pfaff, 1999; Goodson, 2005), e.g., psychosocial stressors (Dickerson and Kemeny, 2004). Its activity is regulated by a feedback loop implicating the amygdala, PFC, and hippocampus (Dedovic et al., 2009), which are altered in WS (Haas and Reiss, 2012; Meyer-Lindenberg et al., 2005; Haas et al., 2014). In one study focusing on cortisol reactivity, which is the end product of the HPA axis, Lense and Dykens (2013) tested participants with WS in two social settings: cognitive challenge and solo musical performance. The results showed that whereas adults of WS relative to TD exhibited diminished cortisol response during neurocognitive testing, their cortisol response remained stable during the musical performance (no data from TD individuals were collected for the latter condition). The authors interpreted the results as indicating that the psychophysiological responses of individuals with WS are sensitive to different types of social situations.

A related question concerns the role of autonomic arousal in social settings and related evaluations in WS. For example, electrodermal activity (EDA) is an amygdala-associated, HPA regulated, non-invasive measure of autonomic function indexing sensitivity to social-affective information at physiological levels (Adolphs, 2001; Laine et al., 2009; LeDoux, 2000). While a growing body of literature indicates that individuals with WS demonstrate altered autonomic response while processing socially relevant stimuli, results are disparate (see Järvinen and Bellugi (2013), for a review). Briefly, Plesa Skwerer et al. (2009) documented hypoarousal in response to dynamic face expressions in adolescents and adults with WS in relation to controls with TD and developmental delay matched on CA. In another study, adults with WS contrasted with a CA-matched TD group demonstrated a lack of typical electrodermal habituation when viewing affective face stimuli, suggesting increased arousal (Järvinen et al., 2012). It was further speculated that the lack of habituation may be linked to the increased affiliation and attraction to faces. Finally, Järvinen et al. (2015) found while overall arousal patterns to emotional face stimuli did not differ between adults with WS and CA-matched TD controls, those with WS demonstrated the highest arousal to happy, and lowest arousal to fearful stimuli, while the TD participants demonstrated the contrasting pattern, as indexed by EDA. Furthermore, in WS, more normal social functioning as measured by the Social Responsiveness Scale was related to higher autonomic arousal to facial expressions, suggesting that low autonomic arousal to faces may be linked to greater social impairments in this population. However, as the above-cited studies utilized emotional facial expressions as test stimuli, it is difficult to determine the extent to which the observed autonomic response occurred in response to the face per se, or to the emotion it conveyed.

Of particular relevance here is to consider scientific endeavors that have examined autonomic arousal directly in naturalistic social settings in individuals with WS, without focusing on specific facial emotions. In one such study, Doherty-Sneddon et al. (2009) found that individuals with WS displayed general hypoarousal and reduced gaze aversion in a naturalistic context involving a math problem. However, similar to the TD controls, their arousal levels accelerated in response to eye contact to the examiner as compared to gaze aversion, and both groups regulated their gaze on the basis of the task difficulty (i.e., increasing gaze aversion was related to more difficult math problems). In another study, Riby et al., (2012) compared baseline EDA reactivity in response to live versus video-mediated displays of happy, sad, and neutral affect in individuals with WS and TD controls matched for CA. The results showed that only live faces increased the level of arousal for both groups. Similar to Doherty-Sneddon et al.'s (2009) findings, participants with WS displayed lower baseline arousal as compared to the TD group, which the authors interpreted as suggesting hypoarousal in this group. Despite the potential of psychophysiological approaches, very little is still known about the psychophysiology associated with the social drive of WS, and what the relations between these salient features of the phenotype are. In this

Table 1
Mean characteristics of the participant groups.

	CA (SD; *range*)	VIQ (SD; *range*)	PIQ (SD; *range*)	FSIQ (SD; *range*)
WS (n=22)	33.94 (9.93; *19.0–55.9*)	71 (8.88; *57–94*)	65 (4.93; *58–75*)	66 (6.49; *54–80*)
TD (n=22/18)	26.06 (6.20; *17.9–43.2*)	101 (16.87; *73–127*)	100 (15.55; *62–127*)	101 (16.18; *69–127*)

vein, Jawaid et al. (2011) recognize that in light of the alterations in autonomic responsivity in social context in individuals with WS, it is important to relate "autonomic responses between individuals with WS who rate negative faces as approachable and those who do not".

To this end, the aim of the current study was examine the underpinnings of approach judgments of individuals with WS by employing a multidimensional approach combining measures of autonomic arousal, social reciprocity (SRS), and two levels of approach evaluations. Specifically, given that the approach motivation in WS appears to be linked to a specific desire to form friendships (Ng et al., 2014), that individuals with WS show diminished awareness of "stranger danger" and demonstrate increased willingness to engage in conversation with strangers in inappropriate situations (Riby et al., 2014a), and that in TD, "deeper level" interpersonal relations associated with friendships require repeated experiences with the individual (Chang et al., 2010), approach judgments will be compared across "shallow" (talking) and "deep" (befriending) interpersonal levels. Employing the Adolphs Approachability task (Adolphs et al., 1998; Bellugi et al., 1999; Martens et al., 2009; Järvinen-Pasley et al., 2010), participants will be asked to indicate their willingness to approach unfamiliar positive and negative faces shown in photographs in light of two questions: (1) "How much would you like to go up and talk to the person", and (2) "How much would you like to be a friend to the person". Autonomic arousal in response to the stimuli will be measured. We hypothesized that TD individuals would be more willing to talk than befriend strangers, and that their overall ratings may be lower as compared to those with WS, reflecting more discriminative and discerning behavior especially with regard to "deeper" social affiliations. If individuals with WS fail to understand the implications between "one-off" versus longer-term social relations, as evidence indicating difficulties in such individuals with understanding the concept of, e.g., friendships may suggest (e.g., Elison et al., 2010; Plesa Skwerer et al., 2004), they were predicted to show indiscriminative approach judgments across conditions. We further hypothesized that for individuals with WS, higher social functioning may be associated with more selective approach behavior. As heterogeneity in approachability has been reported in WS (Järvinen-Pasley et al., 2010; Little et al., 2013; Riby et al., 2014a), this study will address the possibility that underlying physiological arousal and/or the level of the individual's social functioning may contribute to this variability.

2. Material and methods

2.1. Participants

A total of 44 individuals participated in the current study: 22 individuals with WS (9 females), and 22TD comparison individuals (12 females). The genetic diagnosis of WS was established using fluorescence in situ hybridization (FISH) probes for elastin (ELN), a gene invariably associated with the WS microdeletion (Ewart et al., 1993; Hillier et al., 2003). In addition, all participants with WS exhibited the medical and clinical features of the WS phenotype, including cognitive, behavioral, and physical features (Bellugi et al., 2000). The TD participants were screened for history of brain trauma, psychiatric concerns, and central nervous system disorders, and were required to be native English speakers. All participants were recruited through the Salk Institute as a part of a multi-site multidisciplinary program of research addressing neurogenetic underpinnings of human sociality, and were given written informed consent before participation. Written informed assent was also obtained from WS participants' parents, guardians, or conservators. This study was approved by the Institutional Review Board at the Salk Institute for Biological Studies, and all procedures were conducted according to the principles of Declaration of Helsinki.

2.2. Materials

2.2.1 Sample characterization

Measure of cognitive functioning: the participants' cognitive functioning was assessed using the Wechsler Intelligence Scale. Participants with WS were administered the Wechsler Adult Intelligence Scale Third Edition (WAIS-III; Wechsler, 1997), while the TD participants were administered the Wechsler Abbreviated Scale of Intelligence (WASI; Wechsler, 1999). Due to an experimenter error, four TD participants had missing IQ scores; however, based on the screening and their performance on the experimental task, there was no reason to suspect below-normal level cognitive functioning. Table 1 shows the demographic characteristics of the sample of participants with WS and TD. The WS group was higher in terms of CA as compared to the TD individuals (t (42)=3.16, p=0.003). TD participants obtained higher scores as compared to their counterparts with WS on verbal intelligence quotient (VIQ) (t (38)=−7.26, $p<0.001$), performance IQ (PIQ) (t (38)=−9.93, $p<0.001$), and full-scale IQ (FSIQ) (t (38)=−9.36, $p<0.001$).

The between-group differences in CA were controlled for in the statistical analyses by covarying this variable. However, it is noteworthy here that previous evidence has established that individuals with WS across the lifespan exhibit significantly increased approach behavior as compared to both CA and mental age (MA) matched, and both typically and atypically developing, control groups (e.g., Doyle et al., 2004; Jones et al., 2000; Järvinen-Pasley et al., 2008, 2010); thus, no effect of CA on approachability was expected. Between-group differences in cognitive functioning were not controlled for in the data analyses on the basis of postulations that controlling for IQ does not contribute to the clarity of the meaningful differences seen in the autonomic responses to valence and social content in neurodevelopmental conditions (see Cohen et al. (2015), for a discussion). It has been postulated that IQ impairments may stem from shared ANS alterations that lead to the social–emotional processing deficits, and thus, it could be argued that IQ does not lie at the root of social–emotional processing difficulties but they rather have a shared origins founded on the underlying neurodevelopmental disorder (Dennis et al., 2009). As previous studies employing either the identical or similar paradigm to the current study have clearly indicated that elevated approachability is specific to WS, and both CA- and MA-matched groups tend to perform similarly (e.g., Bellugi et al., 1999; Jones et al., 2000; Frigerio et al., 2006), this suggests that the approachability characteristic is an independent function of IQ.

Due to the between-group differences in CA and IQ in the experimental groups, an enriched characterization of the sample is

Table 2
Mean scores for the BAI, MPQ, and ATQ (standard deviations in parentheses) for participants with WS and TD.

Standardized measure	WS	TD	t	P
BAI (WS $n=18$; TD $n=14$)				
Neurophysiological symptoms	0.26 (0.40)	0.11 (0.23)	1.23	n.s.
Subjective symptoms	0.47 (0.55)	0.29 (0.33)	1.11	n.s.
Panic symptoms	0.32 (0.58)	0.09 (0.58)	1.44	n.s.
Autonomic symptoms	0.44 (0.58)	0.32 (0.47)	0.64	n.s.
MPQ (WS $n=20$; TD $n=11$)				
Agentic positive emotionality	8.82 (1.91)	12.55 (2.40)	−4.74	0.001
Communal positive emotionality	12.28 (2.03)	12.12 (2.03)	0.19	n.s.
Negative emotionality	6.80 (1.76)	5.51 (0.82)	2.28	0.030
Constraint	11.93 (1.55)	12.15 (2.03)	−0.34	n.s.
ATQ (WS $n=17$; TD $n=12$)				
Negative affect	4.49 (0.89)	3.65 (0.70)	2.74	0.010
Extraversion/surgency	5.17 (0.69)	4.64 (0.55)	2.26	0.032
Effortful control	3.54 (1.12)	4.67 (0.72)	−3.08	0.005
Orienting sensitivity	4.34 (0.78)	4.61 (0.67)	−0.98	0.337

provided below, including standardized measures of anxiety, temperament, and personality. To maintain consistency with the existing literature (Järvinen-Pasley et al., 2010; Ng et al., 2014), questionnaires were filled in by a parent/caregiver of an individual with WS. The TD participants who were living apart from family members were instructed to complete the inventories with the assistance of a family member or a spouse. Please note that we had missing data across the different measures, and the respective sample sizes are reported in Table 2.

Beck Anxiety Inventory (BAI) (Beck and Steer, 1990). All participants with the exception of five individuals with WS and two TD participants completed the BAI. Consistent with Ng et al. (2014), caregivers of those with WS completed the inventory based on their interactions and observations of their child, and TD individuals were instructed to complete the inventories with a close kin or spouse for reference. This inventory consists of 21 items indexing four domains of anxiety (subjective, autonomic, neurophysiological, panic). Subjective subscale assessed the psychological feelings associated with anxiety (e.g., feeling terrified, nervous). Autonomic subindex consists of physiological symptoms (e.g., indigestion or discomfort in abdomen, feeling hot). Panic symptoms refer to those related to panic attacks (e.g., heart pounding or racing, difficulty breathing, fear of losing control). Finally, neurophysiological subscale refers to anxious symptoms such as shakiness and trembling hands. This four-scale model has been supported in psychometric investigations (Osman et al., 1997). Respondents were instructed to rate the degree each symptom was endorsed. Ratings were measured on a 4-point scale: 0 (*Not at all*), 1 (*Mildly*), 2 (*Moderately*), and 3 (*Severely*). The mean averages per subindex were computed for statistical analysis.

The Multidimensional Personality Questionnaire – Parent version (MPQ-P) (Tellegen, 1985; Klein-Tasman and Mervis, 2003). The inventory consists of 34 items that comprise the following 11 lower-factor subscales: Absorption, Achievement, Aggression, Alienation, Control, Harm Avoidance, Social Closeness, Social Potency, Stress Reaction, Traditionalism, and Well-being. Parents were asked to rate their child on a 4-point scale, resulting in a high or a low score for each of the items: 1-*definitely low on the trait*; 2-*probably low on the trait*; 3-*probably high on the trait*; 4-*definitely high on the trait*. For brevity, we report below the four higher-order factors that can be computed from aforementioned lower-factor items according to instructions reported in Klein-Tasman and Mervis (2003). (1) Agentic Positive Emotionality assesses the desire for social achievement, and it is computed as 2x (Achievement)+Social Potency+Well-Being. (2) Communal Positive Emotionality is intended to capture social attributes pertaining to personality factors and emotionality, and is computed as Well-Being+Social Potency+2x(Social Closeness). Thus, while Social Potency subscale taps into social leadership characteristics, Social Closeness subscale indexes the desire to socially engage with others as well as social-affective traits. (3) Constraint indexes the degree of self-regulation, and is computed as Harm Avoidance+Control+Traditionalism+Absorption. Finally, (4) Negative Emotionality indexing the feelings or expression of negative affect is computed as Stress Reaction+Aggression+Alienation.

Adult Temperament Questionnaire – Short Form (ATQ-S) (Rothbart et al., 2000). The short form of the ATQ was used to assess temperament. Rothbart and Derryberry (2002) define temperament as constitutionally based individual differences in emotional, motor, and attentional reactivity and regulation. Temperament is influenced by experience, and in turn influences experience, and is gradually transformed and integrated into our adult personality (Rothbart et al., 2000). The ATQ is a self-report questionnaire that consists of 77 items rated on a 7-point Likert scale and includes four factor scales: effortful control, negative affect, extraversion, and orienting sensitivity. Negative affect factor scale is the composite of Fear, Sadness, Discomfort, and Frustration. Extraversion/Surgency factor scale is the composite of Sociability, Positive Affect, and High Intensity Pleasure. Effortful Control factor scale is the composite of Attentional Control, Inhibitory Control, and Activation Control. Finally, the Orienting Sensitivity factor scale is a composite of Neutral Perceptual Sensitivity, Affective Perceptual Sensitivity, and Associative Sensitivity. For brevity, we report participant data for these factor scales in Table 2.

Independent t-tests were carried out to explore between-group differences (WS versus TD) in personality dimensions in childhood/adolescence versus adulthood. Table 2 displays the mean ratings and t-test values for the BAI, MPQ, and ATQ scales for participants with WS and TD.

Table 2 shows that the WS and TD samples did not differ in terms of anxiety. On the MPQ, participants with WS scored higher than the TD group on Negative Emotionality, while the TD group was rated higher than participants with WS on Positive Emotionality. On the ATQ, individuals with WS were rated higher than their TD counterparts on Extraversion/Surgency and Negative Affect, while the TD group was rated higher than the WS group on Effortful Control. Given the relevance of the BAI subscales and ATQ Extraversion construct to the current study, Pearson correlations (two-tailed) were applied between the participants' questionnaire scores and their experimental approach ratings. The anxiety measures were not significantly associated with approach ratings for either group of participants (all p values $>.115$). The results of the correlations between ATQ extraversion and approach ratings are reported in Section 3.

2.2.2 Experimental measure

The modified version of the Adolphs Approachability Task (see Adolphs et al. (1998), Bellugi et al. (1999); more recently utilized by Martens et al. (2009)) included 42 black-and-white photographs of faces of unfamiliar people. Out of the original 100 stimuli (Adolphs et al., 1998), 21 photographs that had been most consistently pre-rated positively, and 21 photographs that had been most consistently pre-rated negatively, and that together spanned as wide a range of ratings as possible, were selected as test stimuli.

2.2.3 Psychophysiological recording

EDA was recorded during the initial, passive viewing portion of the experimental paradigm (see procedure below) using BioPac MP150 Psychophysiological Monitoring System (BioPac systems Inc., Santa Barbara, CA) at a 1000 Hz sampling rate. We sampled seven seconds subsequent to stimuli presentation and a three-second pre-stimulus baseline on a trial-by-trial basis, in order to compute event-related change scores. This approach allowed us to obtain weighted trial-specific percentage variations of autonomic activity, thus minimizing the influence of large-scale tonic fluctuations and assessing small-scale ANS reactivity and sensitivity.

2.2.4 Index of social functioning

The Social Responsiveness Scale (SRS) (Constantino and Gruber, 2005) is a 65-item questionnaire that parents or caregivers complete for their child. It is aimed for individuals aged 4–18 years to screen for symptomatology associated with autism, encompassing atypical communication, interpersonal relationships, and the presence of repetitive/stereotypic behaviors. While the SRS was developed as a screening tool for autism, it has also proven helpful in identifying individuals with problem behaviors whose level of functioning in the targeted domains fall at sub-threshold levels. The caregivers' responses to questionnaire items result in T-scores across the scales: Social Awareness, Social Cognition, Social Communication, Social Motivation, and Autistic Mannerisms, in addition to a Total Score. T-scores below 60 indicate no clinically significant concerns in social functioning; T-scores of 60–75 indicate mild-to-moderate social dysfunction; and T-scores higher than 75 indicate severe social dysfunction.

As was mentioned in the introduction, consistent with the existing literature (Klein-Tasman et al., 2011; Riby et al., 2014b; van der Fluit et al., 2012), this measure was included in the study with a purpose of examining *within* WS population differences in social functioning, and thus was only administered to the WS sample. The rationale for this is that all individuals in the TD group were above the targeted age range for this instrument, and were living independently of their families, which would have complicated data collection. Additionally, we had no reason to suspect atypical social functioning in any individual on the basis of our screening, interactions with the participants, and their performance in the experimental tasks. One parent did not return the inventory; therefore, one individual with WS had missing scores on this measure.

2.3. Procedure

The stimuli were randomized with respect to both pre-rated level of approachability or face valence (positive/negative) and gender (male/female), and were preceded by a blinking fixation cross. The order of the stimulus presentation was separately randomized for each participant in the passive and active phases of the experiment.

To prevent autonomic habituation effects, the passive psychophysiological portion of the study was always administered first, followed by the active behavioral portion, during which participants made approachability judgments. The experiment was conducted in a quiet room. Participants sat in a comfortable chair in a well-lit room, 130 cm away from a TFT monitor (screen resolution of 1680×1050 pixels). The stimuli were presented on a desktop computer running Matlab (The MathWorks Inc., Natick, MA), which delivered a digital pulse embedded in the recording at the onset of each stimulus. To measure physiological responses, after a fixation cross for 2000 ms, each stimulus was presented for 3000 ms, separated by an interstimulus interval (ISI) of 8000 ms (blank screen) to allow enough time for autonomic activity to return to near baseline levels.

For the passive psychophysiological portion of the experiment, participants were told that they would see pictures of faces. Participants were only instructed to look at the images while attending to a monitor displaying a fixation cross, and staying as quiet and still as possible. Ag/AgCl electrodes where applied to the skin with an isotonic NaCl electrolyte gel placed on the index and middle medial phalanges of the participant's left hand to record EDA, according to a standard bipolar placement (Venables and Christie, 1980). The recording sessions were divided into four bins separated by brief pauses, during which participants were allowed to stretch and relax and recordings were checked for misplacement and movement artifacts. The sessions were also preceded by a five-minute baseline period, during which ANS activity at rest was qualitatively inspected and participants were given the time to habituate to the sensors. During the experiment, stimulus onsets were marked with trigger codes, embedded into the recordings.

For the active behavioral portion, participants were presented each of the stimuli again individually, and asked to rate them in terms of approachability using a five-point Likert type scale with verbal descriptor labels. The rating scale was displayed on the computer screen. Participants were asked to consider how much they would like to walk up and talk with the person in the photograph. The task was untimed, and the participant was instructed to verbally indicate their willingness to approach using the verbal descriptor labels of the response scale, to ensure that responses were unaffected by participants' potential misinterpretation of the numerical Likert scale. The experimenter operated the computer keyboard on the participant's behalf. After the participant had indicated their response, they were asked to look at the person again and this time, decide how much they would like to be a friend to the person. Each response was coded numerically on a scale from 1 (“No” response) to 5 (“Yes” response), and where 2 and 4 represented “Probably no/yes” response, and 3 represented “Don't know” response. Prior to administering the active portion of the experiment, participants were familiarized with the rating scale using a training set of faces.

2.4. Statistical analyses of ANS data

In order to investigate potential associations between “first-sight” autonomic reactions in response to the positive and negative facial images and subsequent overt ratings reflecting the participants' willingness to engage in social interaction with the persons portrayed in the stimuli, we designed mixed-effects models that were applied on the data. Statistical analyses were run and checked using R (R Development Core Team, 2008), and the R package nlme (Pinheiro et al., 2012). Group (WS/TD), face valence (positive/negative, based on Adolphs' original ratings), and standardized event-related change scores for EDA were included as fixed effects in our models. To account for time-related confounds, we included trial number as a discrete variable in the model, and we modeled autocorrelations between subsequent trial measurements. The autocorrelation structure has been designed as a first-order autoregressive covariance matrix. The models have been designed taking into account random effects due to individual differences between participants and potential confounding covariates, i.e., gender and age. Moreover, all trials containing outliers, i.e., psychophysiological indices that exceeded 2.5 SDs above or below the individual mean, were removed from analyses.

As suggested by Pinheiro and Bates (2000), we assessed the significance of the fixed effects by applying conditional *F*-tests (*F* and *p* values of the Type III Sum of Squares computations). Here, we chose to report the degrees of freedom of our comparisons, but note that the calculations of the relevant denominator degrees in mixed-effects models are approximations. The normality and homogeneity assumptions for linear mixed-effects models were assessed by examining the distribution of residuals.

3. Results

3.1. Behavioral analysis of experimental data

Fig. 1 displays the mean approachability ratings for positive and negative faces for individuals with WS and TD in response to the "Talk" and "Friend" conditions of the experiment. Individuals with WS provided greater approach ratings than the TD group in response to the Friend question, while TD participants provided higher approach ratings than the WS group in the Talk condition.

A $2 \times 2 \times 2$ repeated measures mixed analysis of covariance (ANCOVA) was carried out, with condition (Talk/Friend) and face valence (positive/negative) entered as within-subjects variables, group (WS/TD) entered as between-subjects variable, and CA as a covariate. This analysis revealed a significant main effect of condition ($F(1,41)=6.92$, $p=0.012$, $\eta^2=0.14$), with higher overall ratings occurring in the Talk as compared to the Friend condition; face valence ($F(1, 41)=6.87$, $p=0.012$, $\eta^2=0.14$), with greater ratings occurring in response to the approachable as compared to unapproachable faces overall, and a condition by group interaction ($F(1, 41)=4.71$ $p=0.036$, $\eta^2=0.10$). Thus, the effect of CA was not significant ($F(1,41)=0.59$, $p=0.45$). Post-hoc Bonferroni-corrected t-tests examining the interaction effect indicated that while no significant between-group differences emerged for the total ratings in response to either the Talk ($t(42)=-1.54$, $p=0.13$) or the Friend ($t(42)=0.91$, $p=0.37$) condition, within groups, individuals with WS provided similar overall ratings across the Talk and Friend conditions ($t(21)=1.43$, $p=0.17$), whereas the TD individuals provided significantly greater approachability ratings in response to the Talk as compared to the Friend condition ($t(21)=4.20$, $p<0.001$).

Pearson correlations (two-tailed) were conducted between the participants' approach ratings across the Talk and Friend conditions, to examine the possibility that individuals with WS failed to discriminate between the two levels of interpersonal exchange. Ratings for approachable faces across talk and friend conditions were correlated with each other for both the WS ($r(22)=0.86$, $p<0.001$) and TD ($r(22)=0.58$, $p<0.005$) groups. Similarly, ratings for unapproachable faces across talk and friend conditions were correlated with each other for both the WS ($r(22)=0.85$, $p<0.001$) and TD ($r(22)=0.47$, $p<0.03$) groups.

Finally, Pearson correlations (two-tailed) were applied between the ATQ Extraversion score and the participants' approach ratings across the Talk and Friend conditions, to explore potential relationships between temperament and self-rated willingness to approach. ATQ extraversion scores were associated with approach ratings in response to the approachable-looking faces in the Talk condition for both the participants with WS ($r(19)=0.47$, $p=0.041$) and TD ($r(12)=0.62$, $p=0.033$). All other correlations failed to reach significance.

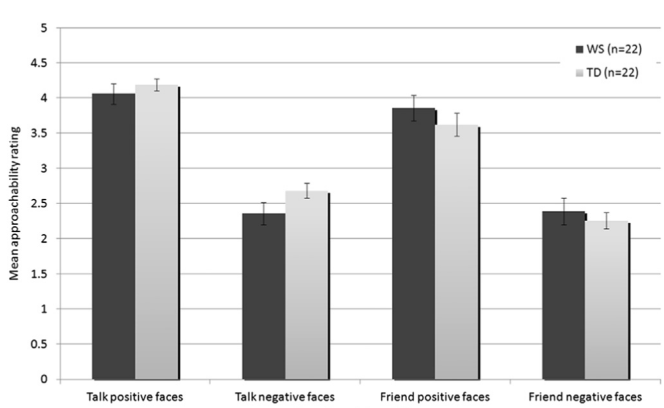

Fig. 1. Mean approachability ratings for positive and negative faces across the Talk and Friend experimental conditions for individuals with WS and TD. (Error bars represent ± 1 SEM).

Table 3
Mean SRS T-scores for participants with WS.

SRS domain	WS ($n=21$) mean T-score (SD)
Social awareness	58.81 (10.68)
Social cognition	71.67 (14.55)
Social communication	64.95 (12.31)
Social motivation	57.00 (14.17)
Autistic mannerisms	81.10 (17.67)
Total score	69.62 (13.11)

Note: Higher T-scores reflect greater deficits in the domain.

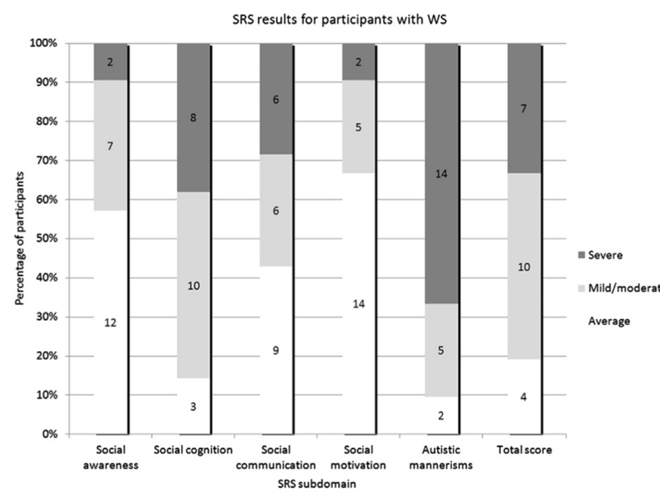

Fig. 2. Percentage of individuals with WS falling in the average, mild-to-moderate, and severe ranges on the SRS parent report inventory (number of participants indicated).

3.2. Social functioning (SRS)

Mean T-scores and SDs for each subscale for participants with WS are displayed in Table 3. Fig. 2 illustrates the number of participants with WS with T-scores falling within the different classifications across the six domains of social functioning (including the total score).

3.3. Relations between approachability ratings and SRS

Pearson correlations (two-tailed) with a Bonferroni correction were applied between the approachability ratings data and SRS scores to examine how social skills may be related to the willingness of participants with WS to engage in social interaction with strangers at conversing and befriending levels. Higher approachability ratings in response to approachable-looking faces in the Talk condition were associated with more normal (i.e., lower) scores on SRS Social Motivation ($r(21)=-0.51$, $p=0.02$) and Autistic Mannerisms ($r(21)=-0.50$, $p=0.02$) subscales. Higher approachability ratings in response to approachable-looking faces in the Friend condition were associated with more normal (i.e., lower) scores on SRS Social Motivation subscale ($r(21)=-0.52$, $p=0.015$). All other correlations failed to reach significance.

3.4. Analysis of EDA data

Reported below are significant findings as revealed by statistical models incorporating standardized event-related EDA change scores. All main and interaction effects failed to reach significance for the Talk condition. This suggests that standardized event-related EDA change scores were not predictive of participants' individual approach ratings in response to the Talk question.

3.4.1 Friend condition

Analyses focusing on the approachability data in the Friend condition indicated that EDA reactivity significantly modulated the participants' overt ratings ($F(1,1612)=9.99$, $p=0.002$). There were also significant two-way interactions between EDA reactivity and face valence ($F(1,1612)=8.52$, $p=0.004$), and EDA reactivity and group ($F(1,1612)=6.58$, $p=0.010$), and a three-way interaction between EDA reactivity, face valence, and group ($F(1,1612)=6.47$, $p=0.011$).

In order to explore these interactions further, we conducted focused statistical models on subsets of data on the approach ratings in response to approachable and unapproachable faces in the Friend condition. Analyses applied on the data in response to approachable-looking faces failed to reveal significant effects in either group.

3.4.2 Focused analyses: unapproachable faces and the Friend condition

The analyses carried out on the subset of trials comprising unapproachable faces revealed a significant effect of EDA reactivity ($F(1,768)=7.53$, $p=0.006$), which was also mediated by group ($F(1,768)=4.97$, $p=0.026$). These results suggest that EDA change scores are at least partially predictive of participants' ratings in response to the friend question: greater EDA reactivity was associated with more positive ratings ($b=0.45$). Moreover, the significant interaction between group and EDA change scores suggests potential group-specific differences in the extent to which standardized event-related EDA change scores mediate individual ratings in response to the negative faces in the Friend condition. In order to probe this interaction further, we further split the dataset and tested the association between EDA reactivity and approach ratings to negative faces within groups. For the TD group, standardized event-related EDA change scores were not significantly predictive of the magnitude of participants' ratings in response to the negative faces in the Friend condition ($F(1,320)=0.14$, n.s.). However, within the WS group, standardized event-related EDA change scores modulated the magnitude of participants' individual ratings in response to negative faces in the Friend condition ($F(1,405)=6.67$, $p=0.010$). Specifically, higher EDA reactivity in response to negative face stimuli was associated with more positive ratings ($b=0.47$), suggesting that hyperresponsivity to negative-looking faces may drive the social disinhibition characterizing WS.

4. Discussion

The main results from this study indicated that while overall level of self-rated approachability toward strangers did not differ between WS and TD groups, an important qualitative between-group difference emerged when the desire to engage with strangers was probed at two different "levels" of social interaction: talking versus befriending. Whereas individuals with WS demonstrated a similar willingness to approach strangers across both experimental conditions, TD individuals were significantly more willing to talk to than to befriend strangers. As such, the current findings provide an important contribution to the existing literature by implying that when approach tendencies are examined in specific social contexts, clear behavioral differences between WS and TD emerge that are modulated by the intended outcome or purpose of the social interaction. It should be noted, however, that the positive association between the ATQ extraversion score and approach ratings in response to the approachable-looking faces in the Talk condition for both the participants with WS and TD and suggests some underlying similarities in the tendency to be socially outgoing in both groups. Importantly, correlational analysis indicated that the high positive associations for participants with WS relative to TD between Talk and Friend conditions across approachable and unapproachable faces suggested that individuals with WS struggled to understand the difference between these two levels of social interaction; they tended to provide similar approachability ratings to the individual stimuli independent of the condition. The pattern of autonomic data failed to support this behavioral finding at least insofar as such appearance-based judgments appeared not to be related to autonomic reactivity, as there was no relationship between arousal and approach judgments in the Talk condition.

In WS, high approachability in response to positive-looking faces across both social interaction levels was further associated with more normal parent-reported social motivation as measured by the SRS. Those who demonstrated high willingness to talk with positive-looking people further exhibited fewer autistic mannerisms, reflecting diminished repetitive interests and stereotyped behaviors. A novel finding linked autonomic responses with willingness to befriend negative-looking people in the WS group: higher electrodermal responses appeared to regulate behavioral ratings in such a way that they resulted in increased approachability. As such, this result may represent an ANS correlate of approach behavior in WS. Typically, elevations in EDA, which indexes the sympathetic branch of the ANS, are elicited by stimuli evoking threat (Bradley et al., 2001) or threat appraisal, which may be an adaptive feature to avoid social dangers (Wessing et al., 2011). Taken together with the current finding, while the ANS appears to alert individuals with WS to social threat, it nevertheless fails to modulate their behavior accordingly, suggestive of a disorganized autonomic system.

The overall behavioral result illustrating that both groups of participants provided higher approachability ratings in response to approachable than unapproachable-looking faces supported the idea that approach behavior in WS is largely driven by their compulsion to approach positive-looking people (cf. e.g., Frigerio et al., 2006). Of interest here is to consider the lack of relationship between autonomic arousal and approachability ratings in the TD participants. One potential explanation is that since the experimental stimuli were static black-and-white images of faces, they were not "real" enough to warrant a distinctive autonomic response in the TD individuals (cf. Riby et al., 2014b). Support to this suggestion is provided by (non-significant) trends of the EDA data, characterized by low overall EDA and responsivity, together with quick habituation to stimuli, particularly in the TD group.

The lack of between-group differences in overall motivation to approach strangers warrants further consideration, as this finding appears to be in odds with previous studies utilizing the same paradigm (e.g., Bellugi et al., 1999; Jones et al., 2000; Martens et al., 2009). However, focusing on the Friend condition, the finding that individuals with WS exhibited elevated approachability relative to the TD participants for both positive and negative faces is in line with the extant literature, suggesting that the social context in which the approach behavior occurs plays a crucial role in the decision processes and motivation particularly for TD individuals. The significantly increased willingness of the TD group to converse as compared to befriend strangers suggests that approaching is considered acceptable when its purpose is to engage in a brief conversation, while intending to pursue a durable interpersonal relationship as in the Friend condition appears to be linked to significantly more discerning and selective judgments. By contrast, approach motivation of individuals with WS did not discriminate across the two contexts, suggesting that the specific social context plays a diminished role in their approach behavior. This difference may also reflect the compromised cognitive capacities of individuals with WS, as social decision-making and trust judgments involve higher-level cognition. Indeed, deficits in the skills linked to social evaluations are likely to relate to the difficulties individuals with WS experience in regulating their social behavior in response to others (Dykens, 2003), reflected as disregard for the social context by individuals with WS. At the same time, the current results provide support to reports suggesting that an important motivation of individuals with WS to approach unfamiliar people is the desire to form close interpersonal relationships (Ng et al., 2014). Alternatively, it is also possible that individuals with WS do not share an understanding of the necessary steps to achieve close interpersonal relationships, and therefore their motivation to talk to others may not be perceived as as differently than the act of befriending others. One potential explanation for the increased approach behavior observed in the Friend condition is that individuals with WS find social interaction and other people highly rewarding. Perhaps the lack of relationship between autonomic arousal and approach ratings in the Talk condition reflects the fact that individuals with WS specifically yearn for and value friendships, and higher approach ratings are given to faces, which produce a higher autonomic effect. Likewise, it is possible that those with WS have atypical autonomic responsivity that act as physiological signals to approach others in casual social situations. At the same time, it is noteworthy here, however, that in studies comparing correspondence of self-ratings of approachability of individuals with WS to their actual approachability in real life, self-perceptions have been found to lack ecological validity (Fisher et al., 2014b; Järvinen-Pasley et al., 2010). Specifically, parents of adults with WS have been found to provide most accurate predictions of real-life approach behavior of individuals with WS (Fisher et al., 2014b).

Correlations between approachability ratings and social functioning provided further interesting insight into approach behavior in WS. Namely, individuals with WS who were reported as showing more normal social motivation as reported by their caregivers demonstrated a greater willingness to engage in both the talking and befriending levels of social interaction with people represented by positively prejudged faces. This finding appears to demonstrate a good agreement between self- and caregiver reports, as the social motivation subscale assesses the extent to which a respondent is generally motivated to engage in social-interpersonal behavior, and includes elements of social anxiety, inhibition, and empathic orientation. Although no significant association emerged between social functioning and approach ratings to negative faces, the current results nevertheless suggest that individuals with more sophisticated social-reciprocal skills were able to discriminate between positive and negative faces, as they were to eager to socially interact specifically with approachable-looking people. In a study by Riby et al. (2014a), it was found that individuals with WS who demonstrated increased awareness of stranger danger also experienced fewer problems with peer relationships and exhibited better pro-social behavior as reported by parents. The current pattern of correlations supports this view, and underscores the importance of employing interdisciplinary approaches (e.g., parent versus self-ratings, psychophysiological and neurobiological techniques, assessments of real-life social behavior, and formal assessments of social/cognitive functioning and psychopathology) toward enhancing the current understanding of approach behavior in WS.

As was mentioned above, while participants' level of social functioning was not associated with approach judgments in response to negative faces in WS, higher EDA in response to negative faces in the Friend condition was associated with increased willingness to approach unapproachable-looking faces. It is possible that unlike in typical function, the higher autonomic response acted to disinhibit approach behavior. Indeed, autonomic arousal has been suggested to modulate amygdala activity, and therefore fear conditioning (Critchley et al., 2002). Critchley et al.'s investigation included individuals with autonomic denervation, and showed that the absence of peripheral autonomic responsivity was related to diminished fear conditioning in the insula, the amygdala, and the hippocampus toward angry faces. Thus, it is possible that the irregularities of autonomic functioning in those with WS attenuate their conditioning of fear in response to threatening stimuli, thereby manifesting in disinhibited social behaviors. Further, Critchley et al. (2002) contended that their findings to support claims that long-term memories are enhanced by emotional arousal (Cahill and McGaugh, 1998). Considering that individuals with WS demonstrate overly social tendencies across the lifespan (Jones et al., 2000; Järvinen-Pasley et al., 2008), it is possible that the poor regulation of social-emotional behaviors is linked to deficits in fear conditioning and consolidation and retrieval of relevant memories (e.g., instances in which their social behaviors did not yield positive instrumental outcomes).

Two major hypotheses for the neural underpinnings of increased approachability were outlined in the introduction. The current result showing a specific autonomic-mediated behavioral response to negative faces may provide support to the amygdala hypothesis insofar as autonomic responses are mediated by the amygdala, and diminished amygdala activation to fearful faces has been shown to be linked to increased tendency to approach strangers (Haas et al., 2010). In a similar vein, in the context Martens et al.'s (2009) study in which higher approach tendency toward negative people was associated with increased right amygdala volumes in the WS group. This initial ANS-based result warrants further neurophysiological research to be carried out to further elucidate its potential correlates in the brain.

The current pattern of results may also be considered as at least partially supporting the hypothesis of frontal lobe dysfunction, as the increased autonomic response linked to elevated approachability suggests that these individuals with WS were unable to inhibit approachability toward negative-looking individuals. However, it is unclear whether the results of this study actually fully support this hypothesis given that increased autonomic arousal to negative faces was linked to greater approach/befriending. The abnormality may not stem from poor inhibition control, but rather dysorganized autonomic system that fails to offer appropriate signaling of threat. E.g., not receiving physiological cues to a danger and thus responding inappropriately to it seems to be linked to bottom up (i.e., perception and interpretation of it) rather than top down (ability to effortfully withhold from responding) process. Thus, the results do not clearly ascertain whether it is the top down versus bottom up processes that are driving the hyper-approach behavior. However, the finding indicating that individuals with WS with more normal social functioning solely showed an approach tendency toward positively pre-rated people, suggests that some individuals were capable of inhibiting approach toward unapproachable-looking strangers. This finding also consolidates previous evidence indicating increased approachability selectively in response to positive-looking faces in WS (Frigerio et al., 2006), by suggesting that this reflects the performance pattern of individuals with WS with higher social reciprocity skills. Indeed, as the effect reported by Frigerio et al. (2006) emerged in response to a paradigm requiring the appreciation of standard expressions of emotion (as compared to judging approachable versus unapproachable facial features without any emotional expression, as in the Adolphs Approachability Task), the suggestion that it is linked to higher social functioning is plausible. In a similar vein, the current results are compatible with previous findings suggesting that amplified approach tendencies of individuals with WS are linked to compromised social-perceptual ability (Järvinen-Pasley et al., 2010; Porter et al., 2007), as higher level of social functioning encompasses such social-cognitive skills as emotion perception. Taken together, this suggests that both the level of social functioning and autonomic nervous system responsivity play a role in approach behavior in WS, with implications to underlying architecture of the social brain. As such, the main finding from the current study suggests that hyperresponsive autonomic function to negative faces may be associated with indiscriminate social approach behavior in individuals with WS. However, in light of the small sample sizes of the current study and a lack of a MA-matched group to the participants with WS, the current preliminary results should be considered with caution. A further limitation is posed by the fact that no measure of inhibitory control was included in the current study; therefore, as indicated above, it is unclear whether the approach behavior is a function of poor ability to monitor and control one's impulses to act on social drive. Future large-scale studies with more carefully matched control groups (e.g., MA-matched group, individuals with developmental disabilities) should be directed toward further elucidating the psychophysiological and neural correlates of the uniqueness of approach behavior in WS.

It is also of importance here to consider the questions posed to distinguish between "low level" (talking) interpersonal relationships and "deeper level" (befriending) interpersonal relationships in the current study. The two questions used were different, not only because the question in the Talk condition referred to a more "superficial" approach behavior, but also in the level of specification. In other words, while going up to a person to talk to him/her may have appeared to participants as a very specific type of approach, the second question may have a multitude of interpretations. To clarify, asking "How much would you like to go up and talk to the person" versus "How much would you like to be a friend to the person, and play with him/her at his/her house once a week" would represent a contraposition between two very concrete actions. Consequently, future research studies utilizing a similar approach to the current study may benefit from incorporating graduating questions (between the two extremes of talking versus befriending) in the study design.

Toward a similar vein, the current pattern of results may partially stem from the unusually high approach ratings of the TD participants in the Talk condition. Unlike in the studies of Frigerio et al. (2006) and Bellugi et al. (1999), in which TD participants were administered an isolated social scenario, our study offered two different possibilities to interact with a stranger. However, the participants were consistently presented with the Talk condition prior to the Friend condition, which may have potentially primed the TD individuals to rate Talk items based on different considerations than when asked to evaluate the social situation alone. In the future, researchers should counter-balance the different social conditions to prevent possible order or priming effects. Alternatively, investigators may consider increasing sample size and data-collection of both individuals with WS and TD, but split within each group and provide solely one interactive condition; however, this method would reduce the ability to assess individual differences across social appraisals.

Results from the parental report of social functioning indicated that approximately 60–70% of the WS sample demonstrated unimpaired social awareness and social motivation, and that social communication was substantially less affected than social cognition and autistic mannerisms; this profile is broadly consistent with the existing evidence highlighting markedly more severe impairments in domains of social cognition as compared to prosocial functioning (e.g., Klein-Tasman et al., 2011; van der Fluit et al., 2012; Riby et al., 2014b). The relative strengths in pro-social and communicative aspects fit in well with the social profile of individuals with WS, which is associated with an elevated tendency to engage in social interaction behaviors with unfamiliar people.

In conclusion, the results from the current study, although preliminary and exploratory, suggest interesting associations between autonomic response to social danger and subsequent social behavior in WS. As such, they may have important implications for new behavioral intervention programs aimed at training stranger safety (see e.g., Fisher, 2014), and open up several avenues for future research. The current results further suggest that approach behavior is not indiscriminate in WS, but is instead mediated by pro-social attributes of others' faces, autonomic responsivity, and the level of social functioning. At the same time, unlike in normative social development, individuals with WS appear insensitive to the social interaction context when making approach judgments toward strangers.

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