Relations between social-perceptual ability in multi- and unisensory contexts, autonomic reactivity, and social functioning in individuals with Williams syndrome

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Compromised social-perceptual ability has been proposed to contribute to social dysfunction in neurodevelopmental disorders. While such impairments have been identified in Williams syndrome (WS), little is known about emotion processing in auditory and multisensory contexts. Employing a multidimensional approach, individuals with WS and typical development (TD) were tested for emotion identification across fearful, happy, and angry multisensory and unisensory face and voice stimuli. Autonomic responses were monitored in response to unimodal emotion. The WS group was administered an inventory of social functioning. Behaviorally, individuals with WS relative to TD demonstrated impaired processing of unimodal vocalizations and emotionally incongruent audiovisual compounds, reflecting a generalized deficit in social-auditory processing in WS. The TD group outperformed their counterparts with WS in identifying negative (fearful and angry) emotion, with similar between-group performance with happy stimuli. Mirroring this pattern, electrodermal activity (EDA) responses to the emotional content of the stimuli indicated that whereas those with WS showed the highest arousal to happy, and lowest arousal to fearful stimuli, the TD participants demonstrated the contrasting pattern. In WS, more normal social functioning was related to higher autonomic arousal to facial expressions. Implications for underlying neural architecture and emotional functions are discussed.

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1. Introduction

Affective communication lies at the heart of successful social interactions and thus interpersonal relationships. Impairments in processing emotional expressions have been suggested to significantly contribute to dysfunctional social behavior and communication in neurodevelopmental disorders, autism spectrum disorder (ASD) being a case in point (Bachevalier and Loveland, 2006). Williams syndrome (WS), resulting from a clearly defined hemideletion of 25–30 genes in the chromosome region 7q11.23 (Ewart et al., 1993; Hillier et al., 2003), is associated with a "hypersocial" albeit relatively poorly understood social and emotional phenotypes. Individuals with WS display a strong drive to socially engage with others (e.g., an increased propensity to approach strangers), and idiosyncratic language features that facilitate social engagement (e.g., atypically high affective content in speech) (see Järvinen-Pasley et al., 2008; Järvinen et al., 2013; Haas and Reiss, 2012, for reviews). Another prominent feature is that social information appears atypically salient individuals with WS, reflected as an attentional bias toward social over non-social stimuli both in social interaction contexts (e.g., Järvinen-Pasley et al., 2008; Mervis et al., 2003) and experiments (Riby and Hancock, 2008, 2009). These social attributes combine with a full-scale intelligence quotient (IQ) profile characterized by the mild-to-moderate intellectual disability range (mean of 50–60) (Mervis et al., 2000; Searcy et al., 2004). Notably, there is substantial heterogeneity in skills tapping into both cognitive (perception, attention, spatial construction, and social-emotional ability) (Porter and Coltheart, 2005) and social domains (social approach tendency in conjunction with response inhibition) (Little et al., 2013).
1.1. Social functioning in WS

Perhaps paradoxically, despite hypersociability, profound impairments in reciprocal social communicative and interactive behavior are evident in individuals with WS (Klein-Tasman et al., 2011; van der Fluit et al., 2012; Riby et al., 2014), including a lack of interpersonal relationships and subsequent social isolation (Davies et al., 1998; Jawaid et al., 2011), impacting such individuals’ well-being. A growing body of literature has focused on characterizing the nature and extent of social dysfunction evident in WS by utilizing diagnostic instruments commonly employed to screen for ASD. Empirical studies have delineated the socio-communicative impairments in individuals with WS employing the Social Responsiveness Scale (SRS; Constantino and Gruber, 2005). In one such study involving 4–16 year-olds with WS, Klein-Tasman et al. (2011) reported more profound impairments in social-cognitive domains (communication and cognition) as opposed to pro-social functions (social awareness and motivation). Consistent with this, Riby et al. (2014) reported normative social functioning as measured by the SRS in merely ~17% of their sample of individuals with WS aged 6–36 years; this implicates that approximately 80% of the WS population exhibit severe social communicative deficits. Finally, van der Fluit et al. (2012) utilized the SRS in tandem with an experimental social attribution paradigm in 8–15 year-olds with WS. On the SRS, the most severe deficits were observed in social cognition, while social motivation appeared unimpaired in those with WS. The results further showed that individuals with WS who performed similarly to typically developing (TD) individuals in interpreting ambiguous social dynamics also demonstrated more normal social functioning in real life. Notably, these associations remained after controlling for intelligence, suggesting that problems with interpreting social situations may play a unique role in interpersonal difficulties experienced by individuals with WS, beyond intellectual functioning. This profile suggesting more pronounced impairments in social-cognitive over pro-social/motivational functions appears stable across development in WS (cf. Klein-Tasman et al., 2011; Riby et al., 2014; Ng et al., 2014).

1.2. Social-perceptual processing in WS

Within the domain of emotion processing, empirical investigations have largely reported deficits in the perception of basic emotions in individuals with WS within both visual and auditory modalities. For example, a study by Plesa Skwerer et al. (2005) included dynamic face stimuli with happy, sad, angry, fearful, disgusted, surprised, and neutral expressions. The findings showed that chronological age (CA)-matched TD participants were superior at labeling disgusted, neutral, and fearful faces as compared to their counterparts with WS. The performance level of the participants with WS was similar to that of a mental age (MA)-matched group of individuals with mixed developmental disability (DD) conditions. Similarly, a study by Gagliardi et al. (2003) included animated faces displaying neutral, angry, disgusted, afraid, happy, and sad expressions. The results showed that participants with WS relative to CA-matched TD controls demonstrated difficulties particularly with disgusted, fearful, and sad face stimuli, while performance of these individuals was indistinguishable from that of a MA-matched, albeit a significantly younger TD control group. Another study by Plesa Skwerer et al. (2006) utilized The Diagnostic Analysis of Nonverbal Accuracy – DANVA2 test (Nowicki and Duke, 1994), which includes happy, sad, angry, and fearful expressions, across both vocal and still face stimuli. The results showed that, across modalities, individuals with WS exhibited significantly poorer performance than CA-matched controls with all but the happy expressions. Taken together, in all of the above-mentioned studies, the performance of participants with WS was indistinguishable from that of MA-matched controls, with the exception of processing happy expressions, which appears relatively preserved.

Studies examining the processing of emotional prosody in individuals with WS are sparse; however, compromised ability has been reported with lexically/semantically intact utterances (Caterral et al., 2006; Plesa Skwerer et al., 2006), while significantly higher performance with emotional filtered speech sentences has been found in such individuals as compared to participants with developmental disabilities matched for IQ and CA (Plesa Skwerer et al., 2006). A dichotic listening study focusing on the hemispheric organization for positive and negative human non-linguistic vocalizations in participants with WS and CA-matched TD individuals found that abnormalities in auditory processing in WS were restricted to the realm of negative affect (Järvinen-Pasley et al., 2010a; Plesa Skwerer et al., 2006). Taken together, this evidence may suggest relatively more competent affect processing in WS in contexts that are free of semantic/lexical interference. However, a recent ERP study using neutral, positive, and negative utterances with both intact and impoverished syntactic and semantic information reported abnormalities in all ERP components of interest linked to prosodic processing (N100, P200, and N300) in individuals with WS relative to TD controls (Pinheiro et al. 2011). This included diminished N100 for semantically intact emotional sentences, more positive N200 particularly for happy and angry semantically intact stimuli, and diminished N300 for both semantically intact and impoverished information. This suggests atypical localization of early auditory functions in WS, showing a bottom-up contribution to the compromised processing and understanding of affective prosody, as well as top-down influences of semantic processing at the level of sensory processing of speech. Overall, impairments in social-perceptual skills have been postulated to contribute to the increased approachability and inappropriate social engagement in WS (Meyer-Lindenberg et al., 2005; Järvinen-Pasley et al., 2010b; Jawaid et al., 2011), urging studies to be directed at investigating social-emotional processing parallel to social functioning in this population. Importantly, however, the evidence discussed above fails to shed light into affect processing capabilities of individuals with WS required in naturalistic social interaction settings, such as the integration of emotion originating from different sensory modalities.

1.3. Audiovisual integration in social context

As discussed above, emotional messages can be transmitted via both visual (e.g., facial expressions, gestures) and auditory (e.g., affective prosody) channels and, in fact, in naturalistic social settings emotional information is rarely purely unimodal. As humans are constantly exposed to competing, complex audiovisual emotional information in social interaction contexts, a reliable interpretation of others’ affective states requires the integration of multimodal stimuli into a single, coherent percept (see De Gelder and Bertelson, 2003, for a review); an automatic function that is evident already at seven months of age in TD (Grossmann et al., 2006). Moreover, multisensory affective perception precedes unisensory affective perception in development (e.g., Flom and Bahrick, 2007). Existing behavioral literature into multisensory emotional face and voice integration in TD indicates that a congruence in affect between the two stimuli aids in the decoding of emotion (Dolan et al., 2001); that multisensory presentation leads to more rapid and accurate emotion processing than unimodal presentation (Collignon et al., 2008); that signals obtained via one sense influence the information-processing of another sensory modality, even in situations where participants are instructed to orient to only one modality (de Gelder and Vroomen, 2000; Ethofer et al., 2006); and that visually presented emotion appears more salient
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