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Cross-modal influences of affect across social and non-social domains in individuals with Williams syndrome

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ABSTRACT

The Williams syndrome (WS) cognitive profile is characterized by relative strengths in face processing, an attentional bias towards social stimuli, and an increased affinity and emotional reactivity to music. An audio-visual integration study examined the effects of auditory emotion on visual (social/non-social) affect identification in individuals with WS and typically developing (TD) and developmentally delayed (DD) controls. The social bias in WS was hypothesized to manifest as an increased ability to process social than non-social affect, and a reduced auditory influence in social contexts. The control groups were hypothesized to perform similarly across conditions. The results showed that while participants with WS exhibited indistinguishable performance to TD controls in identifying facial affect, DD controls performed significantly more poorly. The TD group outperformed the WS and DD groups in identifying non-social affect. The results suggest that emotionally evocative music facilitated the ability of participants with WS to process emotional facial expressions. These surprisingly strong facial-processing skills in individuals with WS may have been due to the effects of combining social and music stimuli and to a reduction in anxiety due to the music in particular. Several directions for future research are suggested.

1. Introduction

Williams syndrome (WS) is a multifactorial genetic disorder resulting from a hemizygous deletion of 25–30 genes on chromosome 7q11.23 (Ewart et al., 1993; Korenberg et al., 2000). It is associated with a unique combination of distinct facial characteristics, widespread clinical symptoms, and an asymmetrical, complex profile of cognitive and behavioral features (see Järvinen-Pasley et al., 2008; Meyer-Lindenberg, Mervis, & Berman, 2006; Morris & Mervis, 2000, for reviews). The neuropsychological profile is characterized by a mean IQ estimate between 40 and 90 (Searcy et al., 2004), with a typically higher verbal IQ (VIQ) than performance IQ (PIQ) (Howlin, Davies, & Udwin, 1998; Udwin & Yule, 1990). In addition, the neurocognitive phenotype is characterized by a unique pattern of dissociations: while relative strengths are evident in socially relevant information processing (e.g., in face and language), significant impairments are apparent in non-verbal intellectual functioning (e.g., planning, problem solving, spatial and numerical cognition) (Bellugi, Lichtenberger, Jones, Lai, & St George, 2000; Bellugi, Wang, & Jernigan, 1994). However, rather than being "intact", evidence indicates that near typical performance in some socially relevant tasks, such as face processing, is associated with atypical neural processing (e.g., Haas et al., 2009; Mills et al., 2000; Mobbs et al., 2004), which may be related to significantly increased attention to faces (Riby & Hancock, 2008, 2009), as well as to a relative enlargement in some major brain structures involved in social information processing (Reiss et al., 2004). Emerging data suggests that at least some of the characteristic "excessive" social functions, specifically an increased tendency to approach unfamiliar people, can be linked to the genetic features of the WS full deletion (Dai et al., 2009). It remains to be investigated, however, whether areas of deficit may be common to general intellectual impairment. Dai et al. (2009) report evidence from a rare individual with a deletion of a subset of the WS genes, who displays a subset of the WS features. These data suggest that GTF2I, the gene telomeric to GTF2IRD1, may contribute disproportionately to specific aspects of social behavior, such as indiscriminant approach to strangers, in WS. However, the pathways of the "dissociation" characterizing the WS social phenotype, that is, the increased sociability and emotionality on one

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hand, and the clear limitations in complex social cognition on the other, are currently poorly understood.

While great progress has been made in characterizing aspects of the social phenotype of WS, and in mapping out some of its major behavioral components, a somewhat unsymmetrical profile has emerged, with major enigmas remaining with respect to the "hypersocial" phenotype. Perhaps the most robust behavioral characteristic is an increased drive for social interaction, including the initiation of social contacts with unknown people, and increased social engagement (e.g., eye contact, use of language, staring at the faces of others)—a feature readily observable even in infancy (Doyle, Bellugi, Korenberg, & Graham, 2004; Jones et al., 2000). Other characteristics that appear unique to this syndrome include a relative strength in identifying (e.g., Rossen, Jones, Wang, & Klima, 1996, Special issue) and remembering (Udwin & Yule, 1991) faces, empathetic, friendly, and emotional personality (Klein-Tasman & Mervis, 2003; Tager-Flusberg & Sullivan, 2000), as well as socially engaging language in narratives (Gothelf et al., 2008; Järvinen-Pasley et al., 2008). Remarkably, overly social behavior and language of individuals with WS in relation to typical individuals extend across different cultures (Järvinen-Pasley et al., 2008; Zitzer-Comfort, Doyle, Masataka, Korenberg, & Bellugi, 2007). At the same time, the social profile of WS is poorly understood and appears paradoxical, in that, for example, the emotional and empathic personality is accompanied by significant deficits in social–perceptual abilities (Gagliardi et al., 2003; Plesa-Skwerer, Faja, Schofield, Verbalis, & Tager-Flusberg, 2006; Plesa-Skwerer, Verbalis, Schofield, Faja, & Tager-Flusberg, 2005; Porter, Coltheart, & Langdon, 2007). This pattern of strengths and deficits suggests that social functioning may have several dissociable dimensions, including affiliative drive and certain aspects of face and social–perceptual processing.

Within the WS phenotype, increased sociability is accompanied by an intriguing profile of auditory processing. Reports suggest that individuals with WS demonstrate a high affinity to music, including a high engagement in musical activities (Don, Schellenberg, & Rourke, 1999; Levitin et al., 2005), which may be linked to increased activation of the amygdala, reduced planum temporale asymmetries, and augmented size of the superior temporal gyrus (STG) (Galaburda & Bellugi, 2000; Levitin et al., 2003; Reiss et al., 2004). However, this is not to say that individuals with WS demonstrate enhanced music processing abilities (e.g., Deruelle, Schön, Rondan, & Mancini, 2005). In addition, in as much as 95% of cases, WS is accompanied by hyperacusis, including certain sound aversions and attractions (Levitin, Cole, Lincoln, & Bellugi, 2005; Gothelf, Farber, Raveh, Apter, and Attias, 2006).

Of specific interest to the current study is the notion that in individuals with WS, heightened emotionality has been reported to extend from their social interactions with others (e.g., Reilly, Losh, Bellugi, & Wulfeck, 2004; Tager-Flusberg & Sullivan, 2000) to the experience of music (Don et al., 1999; Levitin et al., 2005). In one study, Levitin et al. (2005) utilized a comprehensive parental questionnaire designed to characterize the musical phenotype in WS. Participants included 130 children and adults with WS ($M = 18.6$ years), as well as controls with autism, Down syndrome, and typical development (TD) (30 in each group), matched for chronological age (CA). Findings suggested that people with WS exhibited a higher degree of emotionality than Down syndrome and TD groups when listening to music. Individuals with WS were also reported to show greater and earlier interest in music than the comparison groups. Similarly, a study by Don et al. (1999) reported that, in addition to inducing feelings of happiness, individuals with WS differed from the comparison groups (TD, autism, Down syndrome) in that music had a significantly greater propensity to induce sadness in these participants. These findings are interesting in light of the fact that a genetic link between musicality and sociability has been postulated (Huron, 2001). More specifically, according to this view, during the history of human evolution, music is assumed to have played a role in social communication and social bonding, and thus shared genes may be implicated in both social and musical behaviors. However, reports of increased emotionality in response to music are largely anecdotal in the WS literature, a question of significant interest concerns the ways in which musical information may influence the processing of emotion in other modalities and domains in individuals with WS.

Social behavior is arguably tightly coupled to emotion, and the understanding of the emotions of others is critical for successful social interactions. Previous evidence from affect identification studies utilizing standardized face and voice stimuli have robustly established that individuals with WS are significantly impaired when compared to TD CA-matched controls, but perform at the level expected for their mental age (MA). 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 TD participants were significantly better at labeling disgusted, neutral, and fearful faces than their counterparts with WS. Similarly, a study by Gagliardi et al. (2003) included animated face stimuli exhibiting neutral, angry, disgusted, afraid, happy, and sad expressions. The results showed that participants with WS showed noticeably lower levels of performance than CA-matched controls particularly with disgusted, fearful, and sad face stimuli. Another study by Plesa-Skwerer et al. (2006) utilized The Diagnostic Analysis of Nonverbal Accuracy—DANVA2 test (Nowicki & Duke, 1994), which includes happy, sad, angry, and fearful expressions, across both voice and still face stimuli. The results showed that, across both visual and auditory domains, individuals with WS exhibited significantly poorer performance than CA-matched controls with all but the happy expressions. In all of the above-mentioned studies, the performance of participants with WS was indistinguishable from that of MA-matched controls. However, these studies fail to elucidate the potential interactions between emotion processing across different domains (e.g., visual and auditory, social and non-social), and reports of increased emotionality in WS.

Affective expressions are often multimodal, that is, simultaneous and often complementary information is provided by, for example, a face and a voice. Thus, the integration of information from visual and auditory sources is an important prerequisite for successful social interaction, particularly during face-to-face conversation. Recent studies with typical individuals utilizing multi-modal affective face/voice stimuli have shown that a congruence in emotion between the two facilitates the processing of emotion (Dolan, Morris, & de Gelder, 2001); that multimodal presentation results in faster and more accurate emotion processing than unimodal presentation (Collignon et al., 2008); that information obtained via one sense affects the information-processing of another sensory modality, even when individuals are instructed to attend to only one modality (De Gelder & Vroomen, 2000; Ethofer et al., 2006); and that visually presented affect tends to be more salient than aurally presented emotion (Collignon et al., 2008). In the context of music, research has shown that musicians' facial expressions have a significant impact on the experience of emotion in the musical sound (Thompson, Graham, & Russo, 2005; Thompson, Russo, & Quinto, 2008; Vines, Krumhansl, Wanderley, & Levitin, 2006). These results suggest that the processes underlying the integration of facial and vocal information are automatic. Only one known study has examined audiovisual integration abilities in WS (Böhning, Campbell, & Karmiloff-Smith, 2002). In this study, which focused upon natural speech perception, individuals with WS were found to be impaired in visual but not auditory speech identification, with decreased effects of visual information upon auditory processing in the audiovisual speech condition. Neverthe-