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Expecting the worst: Observations of reactivity to sound in young children with Williams syndrome

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Abstract

The study examined behavioral reactions to sound, including startle eye blinks, in young children with Williams syndrome (WS) using video-based observational techniques. Participants were 21 children with WS and 20 children with other developmental disabilities of mixed etiology between the ages of 2.5 and 6. Groups were matched for chronological age and developmental level. All children participated in a semi-structured play interaction including exposure to mild intensity sounds as emitted from conventional toys. Overall, 90% of the children in the WS group were observed to exhibit overt behavioral reactivity to mild intensity sounds, compared to only 20% in the mixed etiology group. Examination of the temporal sequence indicated that children with WS generally exhibited these behaviors before exposure to sound stimuli, suggesting a relation to anticipatory anxiety. Children with WS also exhibited significantly greater acoustic startle eye blinks, often viewed as an indication of heightened emotional state. Taken together, the current findings confirm the presence of heightened reactivity to sound in WS, behaviors previously investigated using parent report alone. The observed behaviors and their potential relation to anxiety are also discussed.
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In recent years there have been a number of examinations of the behavioral characteristics of people with Williams syndrome, as a thorough and precise understanding is necessary for the study of gene–brain–behavior relationships. Williams syndrome (WS) is caused by a sporadic microdeletion of at least 19 genes on chromosome 7q11.23 (Hillier et al., 2003). In addition to a

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distinct cognitive profile, individuals with WS exhibit unique behavioral and emotional difficulties (Dilts, Morris, & Leonard, 1990; Dykens, 2003a; Klein-Tasman & Mervis, 2003; Levitin, Cole, Lincoln, & Bellugi, 2005; Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006). One recurrent observation in the literature is that people with WS report an intense sensitivity to and/or dislike for noises not generally perceived as loud or aversive by others (Don, Schellenberg, & Rourke, 1999; Gosch & Pankau, 1994; Klein, Armstrong, Greer, & Brown, 1990; Levitin et al., 2005; Udwin, 1990). For example, Nigam and Samuel (1994) described a young child with WS who became deeply distressed at the sound of doorbells and telephones. This aversion to commonly encountered sounds is accompanied by marked behavioral responses and is reported to occur in as many as 95% of individuals with WS (Gothelf, Farber, Raveh, Apter, & Attias, 2006; Klein et al., 1990).

To date, the majority of research on behavioral reactivity to commonly encountered sounds in WS has focused on documenting its existence using information gathered almost exclusively from parent-report measures and researchers' informal observations (Gothelf et al., 2006; Klein et al., 1990; Levitin et al., 2005; Van Borsel, Curfs, & Fryns, 1997). Reports often highlight the occurrence of complex defensive behaviors such as covering ears with hands, crying, cringing, hiding, and/or making verbalizations to stop the sound in question. Observed less frequently are severe reactions to sound such as violent inconsolable screaming, repetitive rocking, and self-injurious behavior. In addition to overt defensive behaviors, Gothelf et al. (2006) also suggested that more basic reflexive reactions to sound such as exaggerated startle eye blinks fall within this constellation. Based on reports of parents with children with WS, Down syndrome, autism spectrum disorders, and typically developing controls, Levitin et al. (2005) found that these reactions are by far most prevalent in individuals with WS, have an early time of onset (approximately 1 year of age), and generally decrease in severity as individuals reach adulthood.

Observation of these behaviors has led to numerous investigations attempting to highlight potential neurological and auditory abnormalities that may contribute to sound reactivity in this population (Gothelf et al., 2006; Hickok, Neville, Mills, Jones, Rosen, & Bellugi, 1995; Holinger et al., 2005; Johnson, Comeau, & Clarke, 2001; Marler, Elfenbein, Ryals, Urban, & Netzloff, 2005). In addition, recent literature has suggested an association to emotional difficulties also commonly reported in WS (Blomberg, Rosander, & Andersson, 2006; Dykens, 2003b). Individuals with WS are repeatedly characterized in the literature as having heightened levels of anxiety, fear, and phobias when compared to others with developmental delay (Davies, Udwin, & Howlin, 1998; Dykens, 2003a; Gosch & Pankau, 1994, 1997; Leyfer et al., 2006; Udwin, 1990). As a heightened reactivity to sound is documented for numerous clinical populations experiencing emotional difficulties (Andersson, Lindwall, Hursti, & Carlbring, 2002; Stanfield, 1992), a similar finding for individuals with WS might be expected. Additionally, Blomberg and co-workers found strong relationships between reports of sound reactivity and scores on the Fear Survey Schedule for Children-Revised (FSSC-R; Ollendick, King, & Frary, 1989), a commonly used parent rating scale of childhood fears. Of interest is the finding that sound reactivity correlates robustly with ratings on items on the FSSC-R that have little to do with noise-related fears, for example a fear of animals. This suggests that sound reactivity in WS may indeed be more generally related to a proclivity to act with fear or anxiety in a variety of situations.

As mentioned, observations of exaggerated startle eye blinks have been reported as early as the first year of life in WS, based on maternal interview (Gothelf et al., 2006). The inclusion of exaggerated startle as a behavior characteristic of sound reactivity in this population is potentially of great interest given its demonstrated relation to anxiety in the literature (Grillon & Baas, 2003; Lang, Bradley, & Cuthbert, 1990; Vrana, Spence, & Lang, 1988). The acoustic startle eye blink

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