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Social Responsiveness and Competence in Prader-Willi Syndrome: Direct Comparison to Autism Spectrum Disorder.

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Source

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Abstract

Prader-Willi syndrome (PWS), a neurodevelopmental disorder primarily characterized by hyperphagia and food preoccupations, is caused by the absence of expression of the paternally active genes in the proximal arm of chromosome 15. Although maladaptive behavior and the cognitive profile in PWS have been well characterized, social functioning has only more recently been systematically examined. Findings to date indicate the social impairment exhibited may reflect specific difficulty interpreting and using social information effectively. In addition, evidence suggests that there is an increased risk of social deficits in people with the maternally-derived uniparental disomy (mUPD) subtype of PWS in comparison to those with 15q11-13 paternal deletion (DEL). Using the Social Responsiveness Scale (SRS) and the Social Competence Inventory, our goal was to compare social functioning in PWS to individuals with autism spectrum disorder (ASD). Participants with mUPD scored similarly to the ASD group across most SRS domains. All groups had difficulty with social competence, although the DEL group scored highest on prosocial behavior. Findings suggest further characterization of social behavior in PWS is necessary to aid in advancing the understanding of the contributions of genes in the 15q11-13 critical region to ASD susceptibility, particularly with respect to the overexpression of maternally expressed genes in this region, as well as aiding in awareness and development/implementation of interventions.

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