Neurobehavioral dimensions of Prader Willi Syndrome: Relationships between sleep disturbance and psychotic experiences

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Abstract: Background Prader Willi Syndrome (PWS) is a genetic disorder caused by the absence of expression of the paternal copies of maternally imprinted gene(s) located at 15q11–q13. While the physical and medical characteristics of PWS, including short stature, hyperphagia and endocrine dysfunction are well-characterized, systematic investigation of the long-recognized psychiatric manifestations has been recent.

Methods: Here, we report on the first remote (web-based) assessment of neurobehavioral traits, including psychotic-like experiences (Prodromal Questionnaire-Brief Version; PQ-B) and sleep behavior (Pittsburgh Sleep Quality Index), in a cohort of 127 participants with PWS, of whom 48% had a paternal deletion, 36% uniparental disomy, 2.4% an imprinting mutation and 13% unknown mutation (mean age 19.2 years+/-8.4 years; 55.2% female). We aimed to identify the most informative variables that contribute to psychosis risk symptoms. Multiple domains of cognition (accuracy and speed) were also assessed in a subset of PWS participants (N = 39) using the Penn Computerized Neurocognitive Battery (Penn-CNB).

Results: Individuals with PWS reported a range of psychotic-like symptoms, with over half reporting cognitive disorganization (61%) and suspiciousness (51%). Sleep disturbance, particularly the reasons for sleep difficulty, was most strongly associated with distress related to psychotic-like symptoms. Regarding cognition, individuals with PWS showed the most prominent deficits in accuracy on measures of social cognition involving faces, namely Face Memory, Age Differentiation and Emotion Recognition, and greatest slowing on measures of Attention and Emotion Recognition. However, there were no significant differences in psychotic-like experiences or cognitive performance as a function of PWS genetic subtype. Conclusion: PWS is associated with a high prevalence of distressing psychotic-like experiences, which were associated with sleep disturbance. Findings indicate that self/parent-reported neurobehavioral symptoms and cognition can be assessed remotely in individuals with PWS, which has implications for future large-scale investigations of rare neurogenetic disorders.