

ABSTRACT

Wigren, M. (2004). *Psychiatric problems in Prader-Willi syndrome: Symptom development and clinical management*. Department of Psychology, Göteborg University, Sweden.

Prader-Willi syndrome (PWS) is a rare mental retardation syndrome caused by abnormalities in chromosome 15. The syndrome is primarily distinguished by its severe eating disorder and was until recently best known for being the most common genetic cause of human obesity. Individuals with PWS have an increased risk for developing specific psychiatric problems supposed to be part of the PWS behavioral phenotype. The main aim of this thesis was to delineate those psychiatric features by exploring symptom development, symptom constellations, and parents' service needs. **Study I** mapped past and current indices of skin picking, comorbidity of self-mutilating behaviors, compulsivity, and impulsive-aggressive symptoms among 37 adolescents and adults aged 12 to 30 years. Findings indicated that skin picking has an early onset, is stable over time, and co-occurred with other grooming symptoms and impulsive aggression.

The compulsive symptoms in PWS resemble the adaptive rituals seen in normal children. The aims of **Study II** were to develop a Swedish version of the Childhood Routines Inventory (CRI) and to investigate the extent to which manifestations of compulsivity among 50 children and adolescents with PWS aged 5 to 18 years compare with childhood rituals displayed by healthy 4-year-olds. Results showed that child-like rituals do not subside with age in PWS and that affected individuals display an extremely high adherence to insistence on sameness. On the other hand, common clinical compulsive features related to washing compulsions, like concerns about details, imperfections, dirt, and cleanliness are less common in PWS. Study II suggests that compulsivity in PWS is manifested in ways different from those seen in non-retarded clinical groups.

The purpose of **Study III** was to delineate syndrome specific behavior problems by exploring indices of attention-deficit/hyperactivity disorder (ADHD) among 58 children and adolescents with PWS. Another aim was to investigate how ADHD-related problems associated with self-mutility and excessive rates of child-like insistence on sameness. It was found that indices of ADHD are common and are highly associated with behavior problems. Rigid adherence to sameness, ADHD-related problems, and skin picking constitute a complex symptom constellation that seems to exacerbate over time. The PWS clinical picture is multifaceted and parents need etiology-related support. **Study IV** investigated parental needs of family- and child directed service support on the basis of age, clinical characteristics, and current management of their child. Results revealed that parents wanted information as to availability of external resources and future child needs and that child-focused support was deemed more important than family-directed interventions.

Keywords: Attention-deficit/hyperactivity disorder (ADHD), behavioral phenotypes, childhood rituals, obsessive-compulsive disorder, Prader-Willi syndrome, skin picking

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