

Preliminary Results of Strengths and Weaknesses in Neuropsychological Testing in Children with Prader-Willi Syndrome

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Introduction: Prader-Willi syndrome is well known for negative physical, cognitive, social, and behavioural outcomes. Less is known however, about the neuropsychological effects of the disorder, particularly in children. This ongoing study has examined executive function and attention abilities in 11 children with PWS. Executive functions include a number of specific abilities including inhibition, working memory, set-shifting, strategy employment, and flexible thinking. Executive functions have been defined as being a variety of higher-order mental processes necessary for complex goal-directed behaviour and adaptation to environmental changes and demands. The present study utilized a set of standardized neuropsychological instruments to shed light on a neuropsychological profile of children with PWS. Such a profile will assist intervention efforts so that we can appropriately capitalize on the strengths and address the needs of these children.

Method: The present sample comprised 11 children with a confirmed PWS diagnosis who were recruited from Alberta and British Columbia, Canada. We will be reporting on measures of participants' IQ (Stanford Binet Intelligence Test – Fifth Edition), attention (Test of Variables of Attention- TOVA), and executive function (selected subtests from the NEPSY-II and parental ratings on the Behaviour Rating Inventory of Executive Function- BRIEF). Scale and standard scores were transformed to z scores in order to meaningfully compare performance across measures.

Results and Conclusions: The study sample demonstrated an uneven profile suggesting certain areas of strength and weakness and that children's IQ does not wholly account for performance on these measures. On average, children with PWS obtained abbreviated intelligence scores in the extremely low range. Participants' verbal and nonverbal working memory scores are comparable to their IQ. Results from the TOVA indicate that these children demonstrate significant difficulty maintaining attention for sustained periods of time and show signs of fatigue and impulsivity. On the NEPSY-II, children with PWS demonstrated relative strengths on Animal Sort and Response Set but showed relative and normative weaknesses on Auditory Attention and Inhibition. On the BRIEF, parents reported children with PWS to have relative strengths with organizing materials, initiating tasks, and holding information in memory. Parents reported normative and relative weaknesses controlling their emotions, changing tasks, resisting impulses, regulating their behaviour, and self-awareness. Although a small sample size limits our ability to generalize results and make detailed conclusions, this preliminary information can begin to inform practice. As such, we will also discuss implications for intervention and support based on this early information.