

LINKING LEARNING WITH NEURODEVELOPMENTAL FUNCTIONING - FINAL REPORT

This [project](#) was funded by the Foundation for Prader Willi Research in 2006 and conducted by Drs. Gregory Olley and Anne Wheeler and their team at the University of North Carolina, Chapel Hill (UNC), Center for Development and Learning. Dr. Wheeler is currently finishing the analysis and preparing publications from the study. Following publication, a more detailed analysis will be available, but Dr. Wheeler is pleased to share the following general summary and preliminary results with the PWS community.

The first goal of this study was to design teacher and parent rating forms specific to children with PWS, based on the links between academic skills and the eight neurodevelopmental constructs outlined by Dr. Mel Levine (author of “All Kinds of Minds”). Questionnaires were developed and given to parents and teachers for completion. Dr. Wheeler and her team felt that the information provided by parents and teachers in these questionnaires has been very informative. Analysis of specific sections of the rating scale (e.g., writing skills, behaviors, attention and behavior/learning strategies) has been conducted and will be published soon.

The second goal of the study was to describe the neurodevelopmental and educational strengths and needs of children with PWS. Forty-six children with PWS (mean age 8 years old, range 3-16 years) underwent extensive cognitive and neurodevelopmental testing for this study. An initial look at the data indicates the following patterns.

- Higher Order Cognition. The overall average estimated IQ for the sample was in the low 80s. Since an average IQ (by definition) is 100, the mean of low 80s is below average. In keeping with the fact that those with PWS exhibit a wide range of ability, the range of IQs was from the mid 40s to above 120. Most of the children in the study had estimated IQs in the average or low average range, with 10 in borderline, 6 in mildly impaired, and 5 in the moderately impaired range of intellectual functioning. Two children scored in the above average range. Overall, younger children scored higher on the IQ tests than the older children. In terms of specific areas:

- Memory. On average, the children displayed better visual memory skills than verbal memory skills. Working memory (which is our ability to temporarily store and manage information required to carry out more complex cognitive tasks) scores are in the lower range indicating that working memory skills is likely to be a relative weakness for this sample of children with PWS.

- Attention. On average, the children in this sample had below average visual attention skills. Parent reported of attention problems in approximately a quarter of children.
- Language. On average, the children in this sample displayed similar expressive and receptive vocabulary skills, which are in the below average range. However, more than half of the children scored in the average range on receptive language, and almost as many scored in the average range on expressive language.
- Neuromotor. Neuromotor skills appear to be a significant weakness in the sample; in fact, only 2 children scored in the average range on these tests. A large percentage scored in the well below expected level reflecting significant challenges with fine motor skills and motor planning.
- Educational Skills. Many of the children in the sample were too young to complete the education tests, and instead completed school readiness testing. The majority of the younger children in this sample scored in the average range, indicating good pre-academic skills. The older children's' average scores on the Woodcock Johnson (for this test the Mean score is 100.) overall were below average for math, reading, and writing, but here again, there was a wide range in test scores, ranging from significantly below average to average and even above average for some children in some subject areas. There was the greatest variability in math scores, and writing appears to be one of the more challenging academic areas for the children tested.
- Behavior. Parents rated their child's internalizing (withdrawal, somatic complaints, anxiety, and depressive symptoms) and externalizing (delinquent, aggressive behavior, tantrums) behaviors and almost half reported problems in these areas.

In summary, the children with PWS in the sample demonstrate strength in visual memory and significant relative weaknesses in fine motor abilities, motor planning and working memory. Academic skills appear to be relatively intact in regard to overall cognitive abilities, although more children struggle with writing skills than other academic areas. The youngest children tested appear to have the highest overall cognitive and academic skills. While this is likely an artifact of measurement, as many standardized measures have elevated scores for the lowest end of the age range, it is also possible that this may be related to earlier intervention and/or growth hormone replacement therapy. More research needs to be done to determine this. Finally, behavioral concerns were significant relative to what would be expected in the general population, with nearly half of the children exhibiting elevated global problem behaviors. These behaviors almost certainly have an impact on the child's ability to learn and therefore need to be explored further.

Each family whose child has participated in the assessment portion of this study received a detailed report that provides specific learning strategies derived from clinical observations during the testing and the existing intervention data base.

The third goal of this study was to evaluate any differences that may exist secondary to genetic subtypes. Preliminary results indicate no significant differences between genetic subgroups on any measured variable, except expressive vocabulary (EVT), where children with UPD scored higher than those with deletion. Dr. Wheeler and her colleagues have received funding from other sources to collect additional genetic information on the children in this sample who were reported to either have the deletion, or whose parents were unsure of the subtype.

Summary

This study is one of the largest studies, to date, that looks specifically at the neurodevelopmental, academic skills, and behavioral concerns of children with PWS as young as three years of age. Using both standardized testing instruments and information collected from parents and teachers through the development of a PWS specific Neurodevelopmental and Educational Questionnaire, areas of strengths and needs in children with PWS have been identified. The above results are a preliminary summary of the study and the investigators are in the process of publishing the final results. The investigators are also developing a data base of learning strategies specifically for those with PWS based on the profiles of the children in this study. This information will be used both in future research studies as well as clinically to support learning in children with PWS.