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ASOCIACIÓN MADRILEÑA  
PARA EL SÍNDROME DE  
PRADER-WILLI



## WORKING MEMORY IN ADULTS WITH PRADER-WILLI SYNDROME. BASELINE DATA FROM A PROSPECTIVE, RANDOMIZED STUDY

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**INTRODUCTION:** The Prader-Willi syndrome is a rare, complex, and genetically determined neuro-developmental disorder. The subjects have their own individual characteristics; however share in common particular physical features and specific cognitive strengths and weaknesses. These characteristics change across the lifespan and make up the physical and behavioural "phenotype" of the syndrome. With age, certain behaviours become more evident, for instance repetitive and ritualistic behaviour in older children and adults. According to the literature, subjects score moderate to high on subscales measuring compulsory behaviour. Learning difficulties are common and range the whole spectrum, from mild to severe. Most subjects find reading, spelling and especially mathematics very difficult. By means of a computer based cognitive test (CogState), the aim of the present study was, on a cross-sectional basis, to analyse aspects of the adults working memory.

**METHODS:** Twenty individuals (mean age 28.3 years, range 21-37 years, 7 men) with genetically verified PWS included in the Nordic Prader-Willi study were investigated with selected parts of the CogState battery and compared with normative data and the findings were related to data on anthropometry. According to the definition by WHO of overweight and obesity in normal subjects, 4 individuals had a normal weight, 3 were overweight and the rest obese. The mean Body Mass Index (BMI) was 33.5 k/m<sup>2</sup> (SD = 10.6). The genetic background was a deletion in most of the patients as only one individual had verified UPD.

**RESULTS:** Nineteen subjects were able to perform the computer based test. We found that adults with PWS have significantly decreased elements of working memory compared to normative data. In general, the subjects had median scores within two standard deviations below the normal mean, however with large differences between the cognitive tests performed. Moreover, extensive inter-individual ranges were observed. Females did significantly better than males in some of the items. Related to BMI, obese subjects performed slightly better in working memory than the less obese.

**DISCUSSION:** Adults with PWS were both positive and cooperative in this time limited, motivating, and visual based cognitive test situation. In general, our population had markedly decreased working memory, as detected with the CogState battery. The impairment for all items was of major magnitude, however the impairment of visual memory was less pronounced. This should be taken into account, when teaching strategies and learning are considered. The visual pathway is obviously the best functioning and should be used in educational settings and when giving general information.