

PRADER-WILLI SYNDROME PSYCHOBEHAVIORAL PROFILE IN A CLINIC BASED SAMPLE

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Background: Maladaptive behaviors and cognitive impairment can be main problems in patients with Prader-Willi Syndrome. Understanding difficulties underlying the maladaptive behavior through early diagnosis and management allow tremendous improvement in the whole clinical picture.

Aim: To study the psychobehavioral deviations in a sample of Egyptian children with Prader-Willi syndrome and possible relation to genotype.

Methods: We included 15 out of 72 patients of both sexes, aged from 5-15 years old from the National Research Centre. All patient were fulfilling the diagnostic threshold of modified list of Prader-Willi criteria. Detailed cytogenetic analysis was done using: conventional cytogenetic analysis of peripheral blood lymphocytes and Fluorescent in Situ Hybridization. Cognitive and psychological profile of Patients were assessed using

- (a) standard and semi structured psychiatric sheet and interview
- (b) Wechsler Intelligence Scale for Children,
- (c) Child Behavior Checklist for ages 4-18years,
- (d) Conner's' Parent Rating Scale-Revised; long version,
- (e) Childhood Autism Rating Scale.

Results: 66.7 % of the patients were deletion positive and 33.3% were negative. 100% of the sample were mentally subnormal. 26.3% had autistic symptoms ranging from features to severe form , all were deletion negative . High prevalence of inattention, hyperactivity, anxiety, social, cognitive and behavioral problems were evident in the patients, manifested in more frequency and severity in non-deletion patients.

Conclusion: Cognitive and psychological challenges were evident in PWS patients'. There were some clinical differences according to different genotypes.

Keywords: Prader- willi Syndrome, Cognitive, Psychobehavioral profile, Genotype.