

## **Stimulated GH levels during the transition phase in Prader-Willi Syndrome.**

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Purpose: early institution of GH therapy in children with Prader-Willi syndrome (PWS) yields beneficial effects on their phenotype and is associated with a persistent improvement of body composition, both in the transition age and in adulthood. Reports from GH stimulation testing in PWS adults, however, suggest that GH deficiency (GHD) is not a universal feature of the syndrome, and the current Consensus Guidelines suggest to perform a reassessment of persistent GHD so as to continue GH therapy after reaching adult height. Few data about GH responsiveness to stimulation testing throughout the transitional period in PWS are available to date. Thus, we investigated the prevalence of GHD in a large cohort of patients with PWS during the transition phase.

Patients and methods: one hundred forty-one PWS patients, 72 females and 69 males, aged 15.4-24.9 years, were evaluated by dynamic testing with growth hormone-releasing hormone (GHRH) plus arginine (GHRH + ARG). To define GHD, both BMI-dependent and BMI-independent diagnostic cut-off limits were considered.

Results: according to BMI-dependent criteria, 10.7% of normal weight (NW), 18.5% of overweight and 22.1% of obese PWS maintained a status of GHD. Similar results were obtained by adopting a cut-off limit specific for the adult age (26.2%), as well as criteria for the transition phase in NW subjects (25%).

Conclusion: our study shows that about 20% of patients with PWS fulfilled the criteria for GHD during the transitional age, suggesting the need of an integrated analysis of GH/IGF-I axis, in the context of the general clinical picture and other endocrine abnormalities, in all subjects after attainment of final stature.

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