The GHRH + arginine stimulated pituitary GH secretion in children and adults with Prader-Willi syndrome shows age- and BMI-dependent and genotype-related differences.

E. Marostica, G. Grugni, G. De Nicolao, N.Marazzi, A. Crinò, M. Cappa, A. Sartorio

Growth Hormone & IGF Research, 23: 261-266, 2013.

Objective: The quantitative and qualitative aspects of the pituitary response in children and adults with Prader-Willi syndrome (PWS) are compared in order to verify the possible age-dependent and genotype-related differences in terms of GH secretion.

Design: 29 young subjects (21 males and 8 females) and 65 adults (24 males and 41 females) with PWS were studied. All subjects underwent a standard GH Releasing Hormone (GHRH 1-29, 1 μg/kg as i.v. bolus at 0 minutes) + arginine (0.5 g/kg) test. Peak GH values, standard GH area under the curve (AUC), AUC of the instantaneous secretion rate (ISR), and secretion response analysis (i.e. half-secretion time) were evaluated. A regression analysis was performed to investigate which are the patient characteristics that affect the amplitude and shape of the GH secretion response.

Results: Peak GH values and AUC_{GH} were significantly higher in PWS children than in PWS adults, these differences being also significant both in PWS DEL15 (only peak GH value) and PWS UPD15.Moreover, PWS children showed significantly lower half secretion time than PWS adults, this delayed response being present both in PWS DEL15 and PWS UPD15.

Significant negative correlations between AUC_{GH} and BMISDS were observed in the two groups (adults and children), as well as in adults and children DEL15, but not in adults and children PWS UPD15. A regression analysis performed on the whole dataset showed that for PWS DEL15 the statistically significant variable explaining GH responsiveness was BMISDS ($p < 0.0001$), while for UPD15 no statistically significant covariate was found. Conversely, when the delay of the secretion response was considered, the regression model yielding the best performances was the one with only age as a regressor ($p < 0.001$).

Conclusions: The quantitative and qualitative analyses of GH responsiveness to GHRH + arginine highlight relevant differences between PWS children and PWS adults and genotype-related traits. The negative influence of BMISDS on GH secretion reinforces the need for an early start of life-long weight management in PWS subjects.

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