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The Growth Characteristics of Patients with Noonan Syndrome, and First 2 Years Results of GH Treatment: A Nationwide Multicentre Study

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Introduction: Short stature is a common manifestation of Noonan syndrome (NS). GH deficiency, GH insensitivity, and neurosecretory dysfunction have been reported in the literature. The optimal GH treatment for NS is still controversial. In this study, we aimed to evaluate the growth characteristics in addition to clinical features of NS, and the growth response to GH treatment by using a nationwide registration system. **Material and methods:** Children and adolescents with clinical (according to van der Burgt criteria) and/or genetic diagnosis with NS were included to study. Laboratory assessment including standard GH stimulation tests result were evaluated. Height increment of patients with or without GH treatment were analysed after two years therapy. **Results:** A total of 99 patients with NS (68 males, 31 females) have been enrolled. On admission, the mean age of patients was 8.37 ± 4.2 years, height S.D.s was -3.03 ± 1.65, parentally adjusted height deficit was -2.25 ± 1.73, and 30% of them were pubertal. The percentage of frequently seen clinical findings in NS were 77% short stature, 58% cardiac abnormalities, 59% criptorchidism, 34% chest deformity, 30% neuromotor developmental problem, and 23% ophtalmological disorders. GH stimulation tests were performed on 63 patients, and 40 of them showed suboptimal GH response (<10 ng/ml). 36 patients received rhGH (mean dose: 0.25 ± 0.05 mg/kg per week). Height S.D.s increased from -3.69 to -2.85 after 2 years of therapy. Significant differences was observed according to nonGH-treated patients (*n*: 25) (*P*: 0.02) (Table). PTPN11 gene were analysed 45 of patients, and 29 of them (64%) had mutation. Height S.D.s at admission were similar in patients with or without PTPN11 gene mutation. **Conclusion:** In the 1st year GH therapy, increase in ΔHeight SDS is observed as a positive effect. However this effect of therapy waned at the second year. We suggest that growth therapy optimisation is needed for this NS patients.

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Vitamin D in Short Children on GH Therapy: Effects of Vitamin D Status and Vitamin D Supplementation on Glucose Homeostasis

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Background: Glucose metabolism effects of vitamin D deficiency are debated. GH therapy is associated with increased insulin values and decreased insulin sensitivity. **Objective and hypotheses:** To investigate vitamin D status in short children treated with GH- to investigate if the known effects of GH therapy on glucose metabolism are modulated by vitamin D supplementation. **Method:** 41 children treated with GH for short stature where evaluated 6 months before and 6 months after receiving vitamin D 1000 UI/day (colecalciferolum). We analysed: 1. Vitamin D status; 2. Glucose homeostasis evaluated with: glucose, HbA1S, Insulin, HOMA index before starting vitamin D supplementation and 6 month after vitamin D administration. **Results:** Vitamin D level was below 30 ng/ ml in all the patients and bellow 10 ng/ml in 15% of the patients. Vitamin D supplementation with 1000 UI for 6 months increased vitamin D levels over 30 ng/ ml in 56% of the patients and over 10 ng/ml in all the patients. Vitamin D administration had a demonstrable influence on insulin secretion and insulin sensitivity: in vitamin D <10 ng /ml patients insulin correlated positively with vitamin D concentration. In vitamin D >30 ng/ml patients insulin concentration and HOMA index had a decreasing tendency which could be understood as an effect of lowering GH therapy induced hyperinsulinemia and insulin insensitivity and there metabolic consequences. There was no significant influence of vitamin D supplementation for six months on growth parameters. **Conclusion:** Conclusions: vitamin D evaluation and supplementation is needed in short patients on GH therapy for decreasing the glucose metabolism consequences of GH therapy and possibly in the long time for improving response to therapy.