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An 8-year-old boy was admitted to our hospital with the complaint of short stature. The patient was born at term with normal vaginal delivery after an uneventful pregnancy. His birth weight was 3550 g, his birth length was unknown but his mother noted that his height was always shorter than his peers. The patient had met developmental milestones in time. He received routine immunization and he had uneventful medical history to this age. His parents were first-cousins. He lived with his parents and had two siblings. His father's and mother's heights were 165 and 167 cm, respectively. His 19-year-old brother was 174 cm and his 16-year-old sister was 141 cm in height. On physical examination, a significant growth retardation was seen. His vital signs were normal. His weight and height were 13.3 kg and 98 cm (below 3rd percentile) respectively and the height SDS value was very low (-5.75 SDS) (Figure 1). His body-mass index was 13.8 kg/m^2 (between 3-10th percentile). The patient was at Tanner stage 1 for puberty and his stretched penis length was 3 cm (below 10th percentile for his age). He had frontal bossing, hypoplastic nasal bridge (Figure 2)

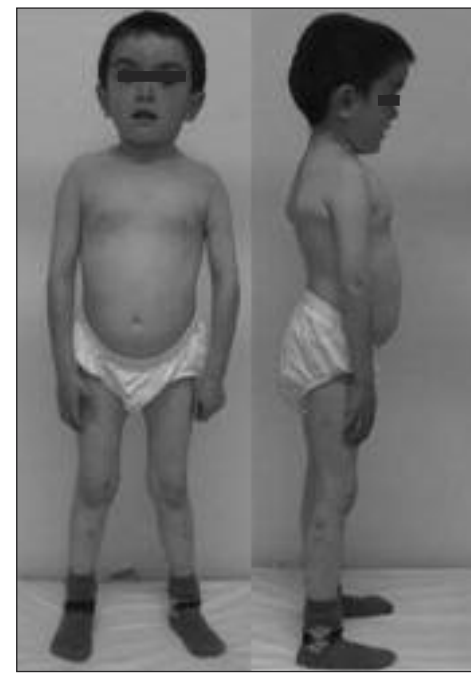

FIGURE 1: Appearance of the patient. The photo is published with the written permission of the parents

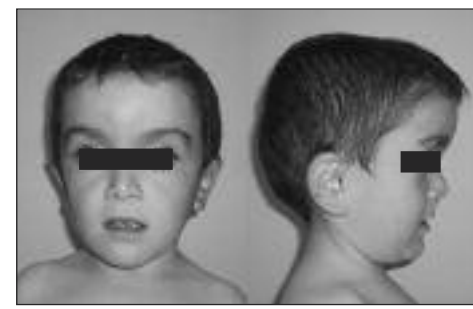

FIGURE 2: Characteristic features of the patient face. The photo is published with the written permission of the parents

and high-pitched voice. Results of complete blood count as well as liver, renal and thyroid functions were normal for his age. Screening for Celiac disease was negative. His bone age was markedly delayed, estimated as four years. Laboratory tests showed markedly low serum insulin-like growth factor-1 (IGF-1) and insulin-like growth factor binding protein-3 (IGFBP-3) (25 and 775 ng/ml, respectively). According to these data, growth hormone (GH) deficiency was considered and peak growth hormone response to L-dopa and clonidine were evaluated. Results of GH stimulation tests are shown in Table 1.

Provocative testing of pituitary GH secretion revealed elevated basal GH and increased peak GH levels. Subsequently response to exogenous GH was evaluated with insulin like growth factor generation test. Serum concentrations of IGF-1 and IGFBP-3 (Table 2) did not increase after injections of GH (0.1 U/kg body weight for 4 days).

TABLE 1: Growth hormone values (ng/mL) before and after stimulation.

	Basal value	30th minute	60th minute	90th minute
L-dopa	21.4	27	40	40
Clonidine	17.7	20	11.3	13.7

TABLE 2: Insulin like growth factor generation test.

	Basal value	5th day of test	Normal range
IGF-1 ng/mL	<25	<25	70-480
IGFBP-3 ng/mL	750	<500	3000-6500

On the basis of given medical history, physical examination and laboratory data what is your diagnosis?