Effectiveness of rhIGF-I treatment in a girl with Leprechaunism

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Background

Infants with severe insulin resistance syndrome show failure to thrive.

Objective and hypotheses

Effect of rhIGF-I treatment on growth in a patient with severe insulin resistance syndrome.

Methods

Case report.

Results

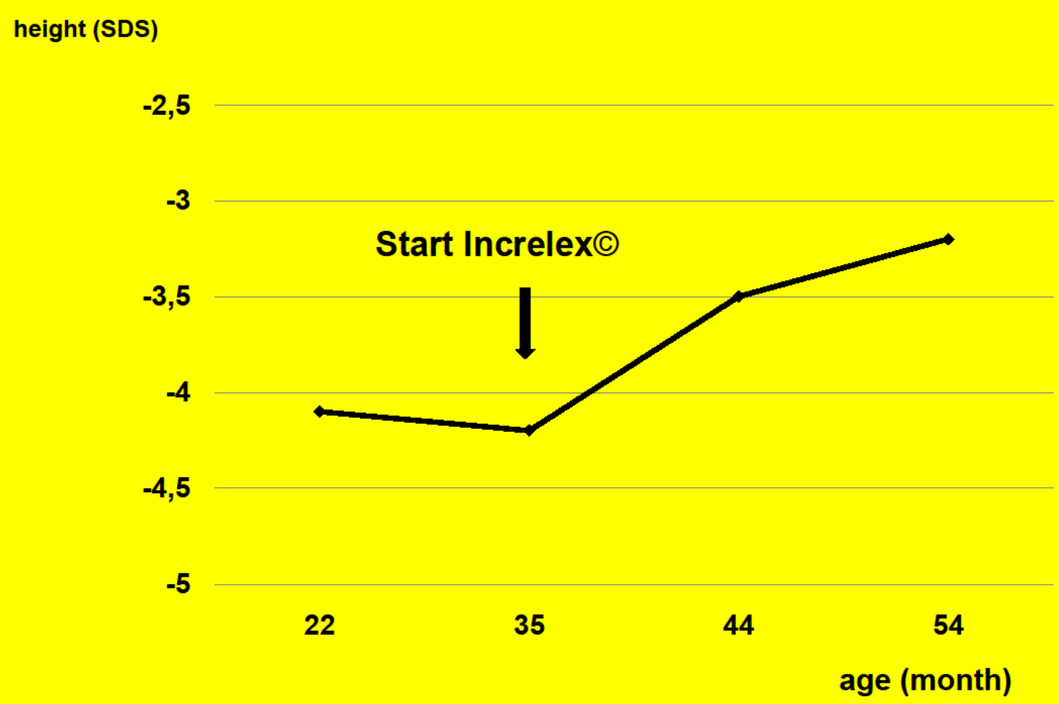
The patient is a now 4 ½ -yr old Caucasian girl of unrelated healthy parents. She was born after a 40 week gestation as a small for gestational age infant with a birth weight of 1970 g (-3.56 SDS). After birth she developed high blood glucose levels of >400 mg/dL with high levels of insulin (>2.000 mU/mL) and C-peptide (>60 ng/mL), fasting hypoglycaemias and high blood pressure. At the age of 22 months she was admitted to our hospital. She had decreased subcutaneous fat, hypertrichosis, ovarial cysts, and acanthosis nigricans. She was extremely short and had very low levels of IGF-I and IGFBP-3 (Table 1). We started treatment with rhIGF-I (Increlex©, Ipsen, Ettlingen, Germany) with a dose of twice daily 0.6 mg s.c. at the age of 35 months. She showed a catch-up growth of +1.0 SDS over the first 19 months (Fig. 1). Levels of IGF-I were in the middle-normal range bevor and in the upper normal range two hours after s.c. injection (Table 1). Levels of IGFBP3 remained low (Table 1). Treatment was well tolerated, blood glucose levels remained stable and haemoglobin A_{1c} level was within the normal range (5.7%).

Sponsored by a travel grant from „Forum Wachsen“.

Table I

Age (months)	Height (cm) (SDS)	Weight (kg) (SDS)	Increlex© dosage (µg/kg/d)	IGF-I (µg/L) (SDS)	IGFBP-3 (mg/L) (SDS)
22	73.4 (-4.1)	8.0 (-3.9)	-	1 (-8.0)	0.18 (-9.4)
35	80.4 (-4.2)	8.8 (-4.9)	-	2 (-7.9)	0.55 (-5.6)
Start Increlex©: 2 x 0.6 mg/ d (136 µg/kg/d) at the age of 35 months					
44	87.3 (-3.5)	10.6 (-3.9)	113 (2 x 0.6 mg)	91 (-0.1) (pre-dose)	0.82 (-4.3)
54	94.1 (-3.2)	11.9 (-3.7)	117 (2 x 0.7 mg)	225 (+1.7) 2 hrs post-dose	N.D.

Figure I



Conclusions

This leprechaun patient has an IGF-I deficient state and rhIGF-I treatment induced catch-up growth.

DOI: 10.3252/pso.eu.54espe.2015