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EFFECTIVENESS OF RECOMBINANT IGF-I TREATMENT IN A PATIENT WITH ISOLATED GH IA DEFICIT PRODUCER OF ANTI-GH ANTIBODIES

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BACKGROUND

GH deficiency type IA represents the most serious form of isolated deficit growth hormone (IDGH). It's transmitted as an autosomal recessive pattern and in most cases there is a homozygous deletion of the GH1 gene. Good initial response to treatment is characteristic, although often could appear antibodies against recombinant GH.

CASE PRESENTATION

First visit 5 years (no previosuly data)
Irrelevant neonatal history
Normal psychomotor development
Parents blood relatives (first cousins) with normal height.

Phenoype: truncal obesity, prominent forehead, and craniofacial disproportion.
Karyotype 46 XY

MRI pituitary hypoplasia

GH deficit (clonidin test) No other abnormal hormonal results.

It was suspected **IDGH type IA**, and genetic study showed **absence of GH1 gene in homozygous.**

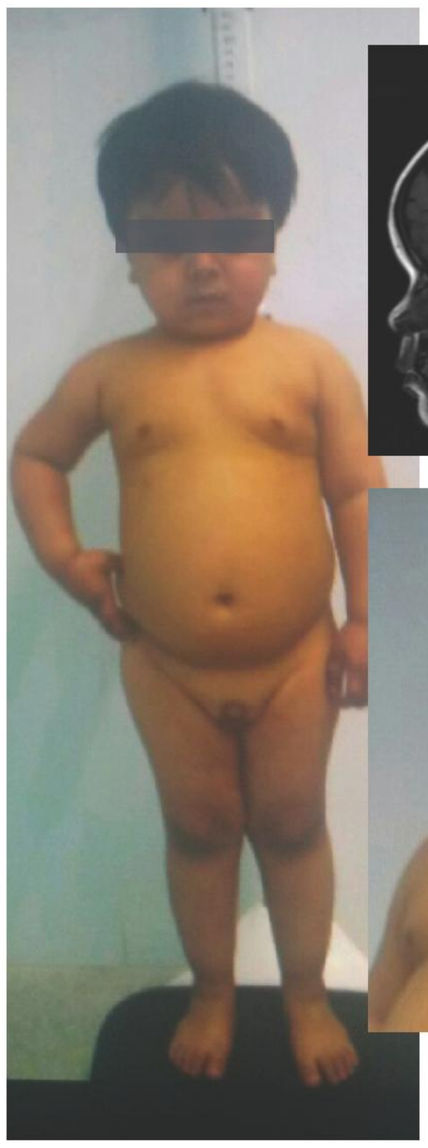
Started on r-hGH therapy.
Heigh velocity 12 cm/year with normal IGF-1 levels initially, but it dropped to 3,3cm/year (-3,5 SD) six months later, with undetectable IGF1 levels.

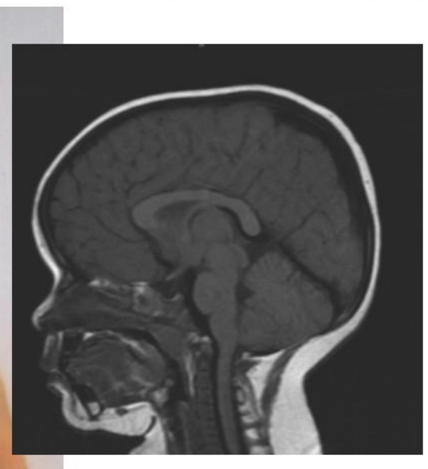
Presence of **anti-hGH antibodies** was suspected and confirmed on laboratory analysis.

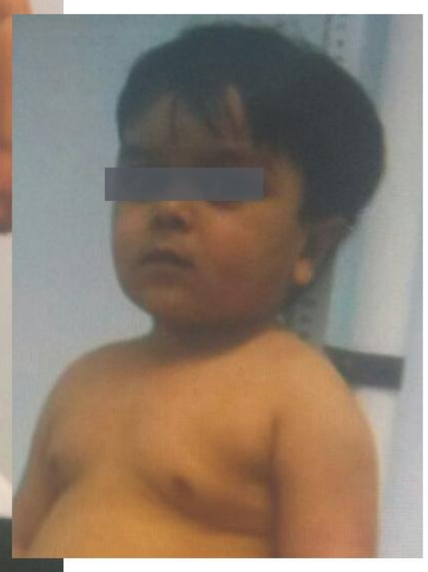
Recombinant IGF-1 treatment was started, increasing growth velocity to **10cm/year** without complications in his evolution.

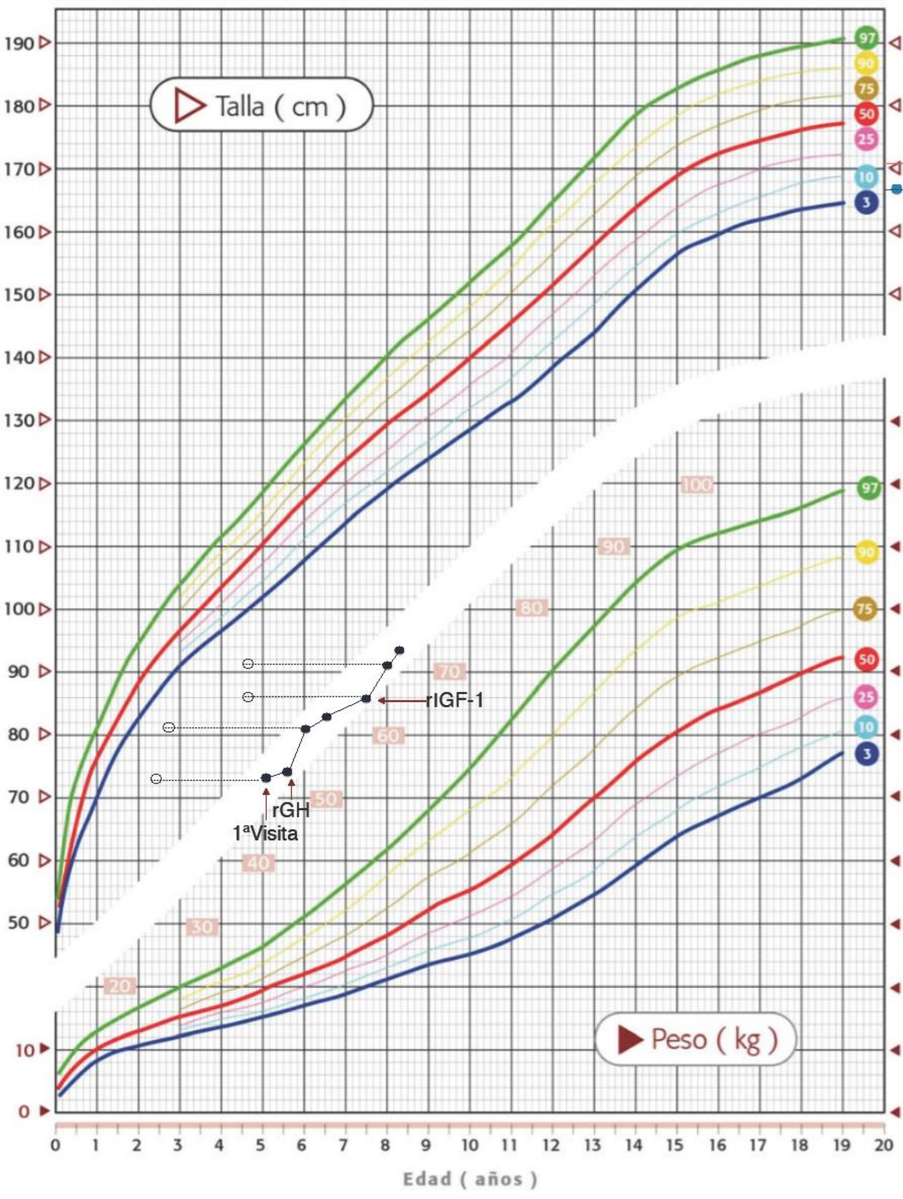
5 years HEIGH 74,2cm (- 8, 07 DE)

8 years 2m HEIGH 93 cm (-6,87 DE)









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An Pediatr (Barc) 2008;68:552-69

CONCLUSION

Development of anti-GH antibodies is an inconstant finding despite identical molecular defects. Response to r-hGH treatment could be different. In our reported case, rIGF-1 treatment has been shown as the only possible alternative therapy, resulting highly effective with no side effects. We consider the importance of reporting clinical experience and response to new treatments available for an uncommon pathology

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