OUTSTANDING GROWTH RESPONSE TO GROWTH HORMONE REPLACEMENT THERAPY IN 3 DIFFERENT CASES OF GROWTH HORMONE DEFICIENCY (GHD)

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Background

Case Reports

Growth response in growth hormone deficient children during growth hormone (GH) replacement therapy rarely fulfill our projections and patient's expectations. It is well known that prediction models suffer from over-optimism. We here report 3 cases with outstanding growth response.

Case 1

First patient was diagnosed as gluten enteropathy in early childhood, but the diet didn't improve his growth. At age of 7 years the diagnosis of isolated growth hormone deficiency (GHD) was established and GH substitution therapy introduced. Next 2 years his growth velocity increased to 10cm per year. Routine head MRI revealed hypothalamic tumor.

Case 2

Severe growth failure in eight-years-old boy with history of purulent meningitis in toddler period and head trauma at age of six. MRI discovered congenital pituitary abnormality (pituitary hypoplasia and ectopic posterior pituitary). Combined substitution therapy improved his height from -3SD to + 0.67 SD

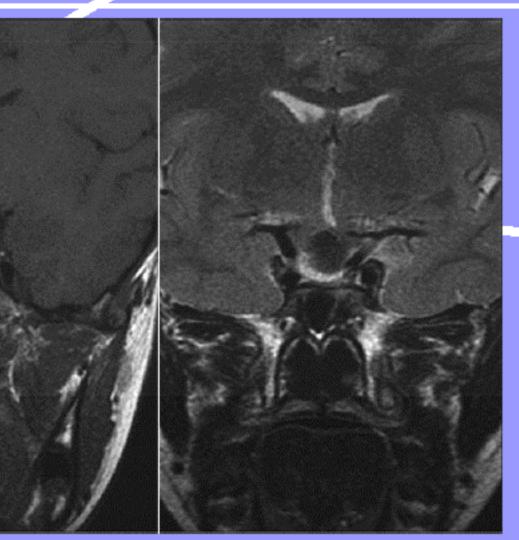
Case 3

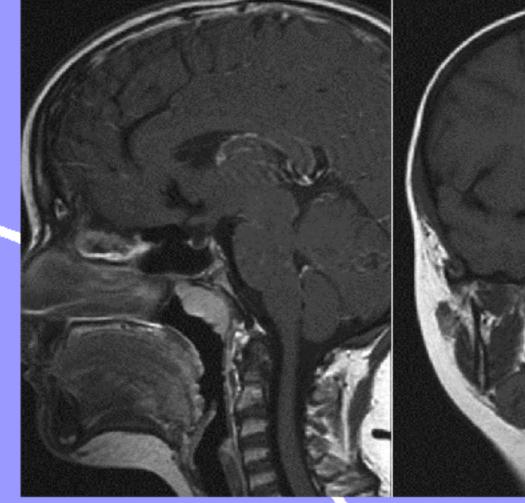
An Empty Sella Syndrome was diagnosed in short obese boy with delayed puberty. At diagnosis his height of 148.8 cm was -1.88 SD (P3). During 4 years of combined therapy he acheved height of 184 cm (P90), +1.28 SD.

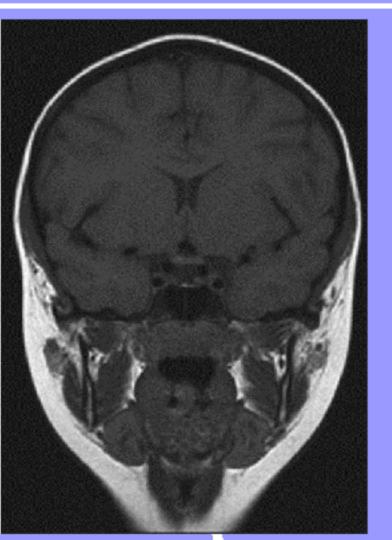
	At Admission	Last Visit
Age (years)	6.83	11.42
Height (cm)	109.0 (-2.25 SD)	144.0 (0 SD)
Height age (yrs)	5	7.25
Body Mass (kg)	18.4kg (+0.4kg)	42.6 (+6.6)
BMI	15.31 (>P50)	20.54 (P85)
BA (yrs)	5.5	9.5
MPH		182.5 (P75)
GH peak (mU/l)		
Clonidine test	2.8	
Insuline test	4.0	
Cortisol	1009.0	
fT4 (nmol/l)	13.2	15
TSH (mU/l)	4.0	1.03
PRL (mU/l)	375	572
IGF1 (ng/ml)	187	242
Hormonal therapy:		
GH (mg/kg)	0.025	0.023
Na-l-thyroxine (µg/day)	/	75
Mean HV (cm/year)	7.6	

	At Admission	Last Visit
Age (years)	8.55	13.25
Height (cm)	114.7 (-3SD)	163.5 (+0.67)
Height age (yrs)	5.75	14.0
Body Mass (kg)	22 (+2kg)	43.6 (-6.4kg)
BMI	16.72 (>P50)	16.31 (P25)
BA (yrs)	5.0	13.0
MPH		182 (P75)
GH peak (mU/l)		
Clonidine test	0.63	
Insuline test	0.93	
Cortisol	273.0	289.0
fT4 (nmol/l)	9.0	16.24
TSH (mU/l)	1.1	0.01
PRL (mU/l)	188.0	183.0
IGF1 (ng/ml)	25 (-2.5 SD)	356 (+0.6)
Hormonal therapy:		
GH (mg/kg)	0.035	0.018
Na-l-thyroxine (µg/day)	25	75
Mean HV (cm/year)	10.38	
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	At Admission	Last Visit
Age (years)	14	18.25
Height (cm)	148.8 (-1.88SD)	184.0 (+1.28)
Height age (yrs)	12.0	>18.0
Body Mass (kg)	50 (+11kg)	77
BMI	23 (P85)	22.74 (>P50)
BA (yrs)	11.0	16.0
MPH		180 (P75)
GH peak (mU/l)		
Clonidine test	0.66	
Insuline test	0.29	
Cortisol	643.0	513.0
fT4 (nmol/l)	12.0	16.0
TSH (mU/l)	4.3	3.2
PRL (mU/l)	445.0	141.0
IGF1 (ng/ml)	63 (-2.8)	294 (+0.67)
Hormonal therapy:		
GH (mg/kg)	0.035	0.018
Na-I-thyroxine (µg/day)	25	75
Mean HV (cm/year)	8.28	

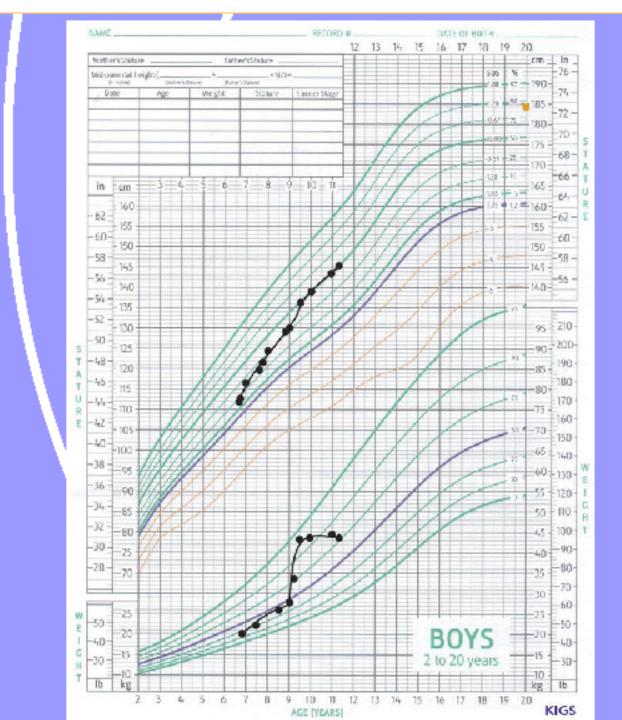




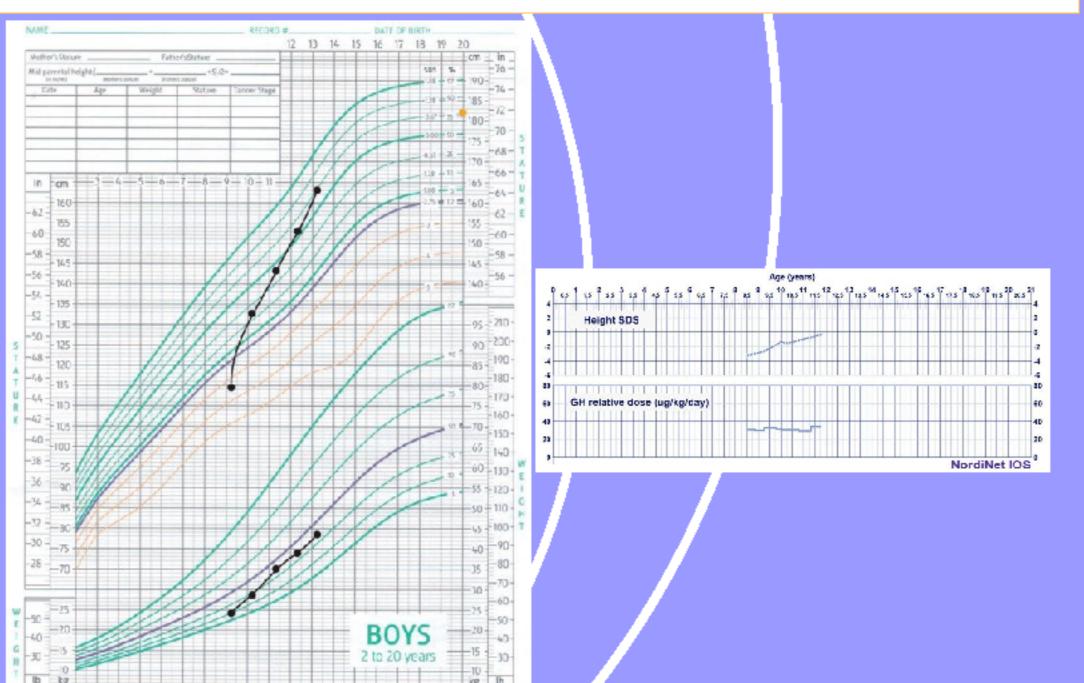


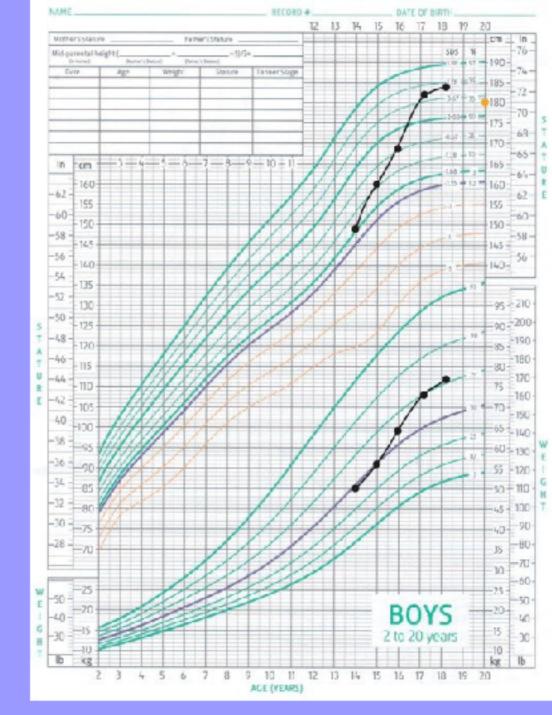
MRI – Pituitary hypoplasia and ectopic posterior pituitary

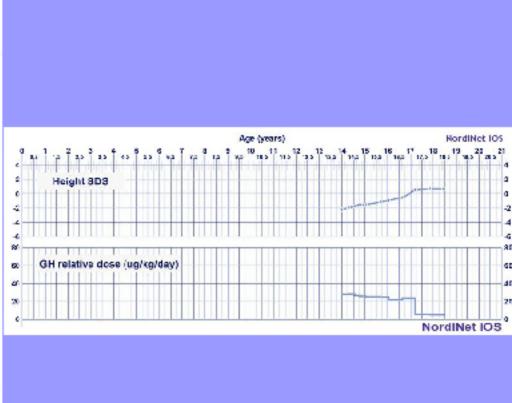
MRI – Empty sella



MRI –TU hypothalamici







Conclusions

These 3 boys showed excellent growth on average doses of GH at start of therapy and during followup. GH doses were adjusted according to ideal body weight for height, body surface and IGF values. What could influence achievement of the best growth response in these patients?

- They all had combined pituitary hormones deficiency (concomitant hypogonadism)
- Influence of central satiety regulation and obesity (growth because of insulin)
- Genetic influence (tall parents)
- Unsupressed GH activity (impaired somatostatin release).



