Final adult height, Insulin-like Growth Factor 1 (IGF-I) concentration in adolescents and young adults with β-thalassemia major (BTM) with and without Growth hormone Deficiency

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Introduction

Patients with beta thalassemia major (BTM) had high incidence of short stature and variable degree of disturbance in growth hormone (GH) – insulin-like growth factor -1 axis. However, many other factors may affect their final adult height.

***Aims of the study*: To measure the final adult standing height (FA-Ht), liver iron content (LIC) and insulin-like growth factor 1 (IGF-I) concentration in BTM patients with and without GH deficiency.**

Methods and Materials

We studied the FA-Ht, body mass index (BMI), and measured IGF-I concentration in two selected groups of patients according to their growth hormone (GH) response to provocation.

9 with normal GH secretion (GHN) and 8 with GH deficiency GHD; peak GH response to provocative test with clonidine < 7 ng/ml), who were on iron chelation therapy with DFO given subcutaneously that was changed to oral deferasirox during the last 5-6 years.

In both groups LIC was measured using FerriScan ® R2-MRI method. These 17 patients were not treated with rhGH.

Results

Thalassaemic adolescents with defective GH secretion had decreased FA-Ht and HtSDS (159.1± 6.42 cm, and -2.5 ± 0.9 respectively) compared to those with normal GH secretion (163.5 ± 5.2 cm and -1.74 ± 0.83 respectively).

The IGF-1-SDS did not differ between the two groups. Neither ferritin level nor IGF-1 concentrations were correlated with the Ht-SDS. (table)

Correlations

The final FA-Ht-SDS was correlated significantly with :

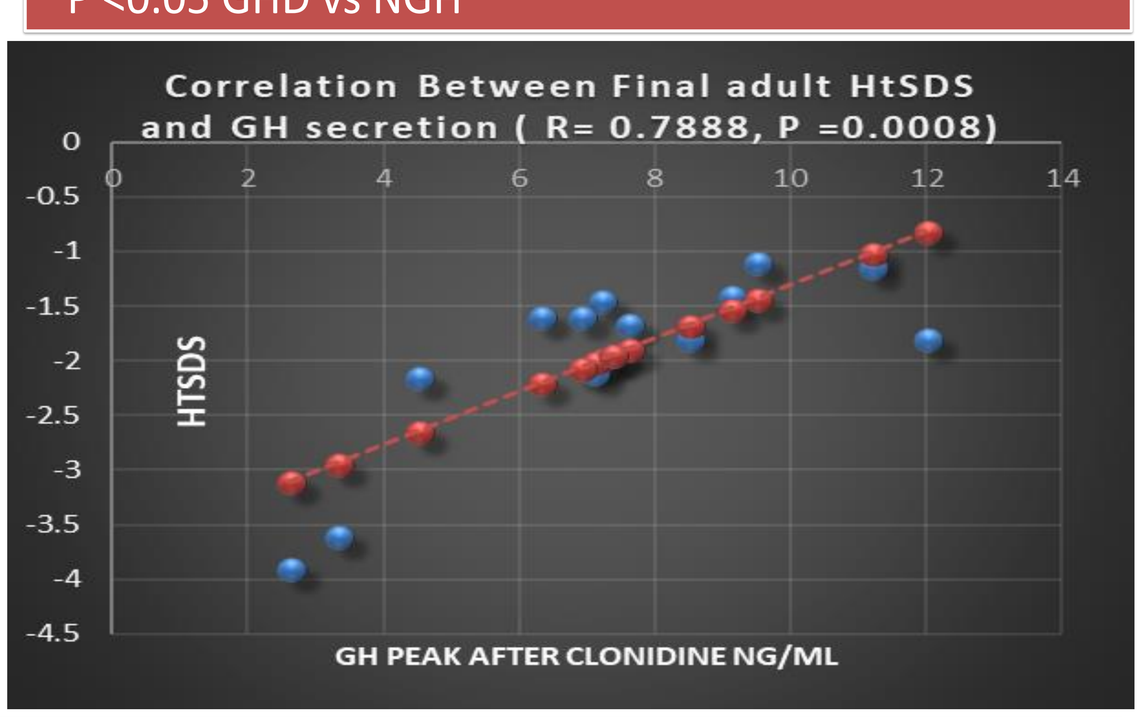
1. the peak GH secretion (r = 0.788, p = 0.0008), (fig)
2. mid-parental height SDS (r=0.58, P <0.01) and LIC (r = 0.26, p = 0.08).

Neither ferritin level nor IGF-1 concentrations were correlated with the Ht-SDS

Final adult height and IGF-1 in adults with (GHD) and without (NGH) GH deficiency diagnosed before the onset of puberty.

	Age years	HtSDS	BMI-SDS	Ferritin ng/ml	Basal GH ng/ml	Peak GH ng/ml	IGF-1 SDS
GHD							
Mean	21.2	-2.5	-0.3	2529	0.4	4.7	-2.7
SD	3.4	0.9	0.3	1225	0.4	1.5	0.7
NGH							
Mean	23.4	-1.7*	-0.6	2666	1	9.9*	-2.8
SD	4.3	0.5	0.4	1455	1.3	1.8	1.3

* P <0.05 GHD vs NGH



Conclusions

Patients with BTM and GHD were significantly shorter compared to their pears with NGH. Therefore, rhGH therapy can be recommended for the treatment of thalassemic children and adolescents with GHD in addition to proper blood transfusion and intensive chelation to improve their final height.

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