Final adult height, Insulin-like Growth Factor 1 (IGF-I) concentration and endocrine complications in adolescents and young adults with β-thalassemia major (BTM) who received oral iron chelation (OIC) in comparison with those who did not use OIC

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Introduction

Relatively little is known about endocrine function, bone mineral health, and growth during oral iron chelation therapy (OIC) in β-thalassemia major patients (BMT) on treatment with deferasirox

Aim of the study

To measure the final adult standing height (FA-Ht) and the frequency of endocrine complications in relation to their liver iron content (LIC) and insulin-like growth factor 1 (IGF-I) concentration . Patients were grouped into two groups according to their iron chelation therapy.

Methods and Materials

The first group (Group A; 15 patients, 6 females and 9 males) received oral iron chelation therapy (OIC) with deferasirox for 6 years before puberty. The second group (Group B; 40 patients) attained the FA-Ht before the use of OIC (iron chelation therapy with deferoxamine (DFO) given subcutaneously, since the age of 2 years). In both groups LIC was measured using FerriScan ® R2-MRI method.

Results

Patients with BTM who received OIC for 6 years or more before their end of growth were significantly taller and had lower LIC assessed by FerriScan® R2-MRI, and lower fasting glucose level (FBG) and liver enzymes (ALT and AST) concentration, and higher IGF-1 SDS versus those who did not receive OIC before attaining FA-Ht.

The prevalence of endocrinopathies, including hypothyroidism, impaired fasting glucose and hypogonadism were significantly lower in Group A versus Group B.

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

Results

Final adult height and biochemical parameters of adolescents with BTM who were on oral chelation therapy (OCT) versus those who attained their final adult height before using OCT

		Age years	Ht-SDS	BMI Kg/m2	FT4 pmol/L	TSH mIU/l	Serum ferritin ng/ml	IGF-1 - SDS ng/ml	LIC mg/Fe /g dw	AST U/l	ALT U/l	ALP U/l	FBG mmol/l
Group A (OCT) n= 15	Mean ±SD	20.0 ±1.4	-1.2* ±0.7	21.7 ±3.0	12.4 ±0.8	2.6 ±0.7	1520.0 ±985.0	-1.3 ±0.9	10.1 ±6.7	39.1 ±13.2	46.5 ±22.2	129.4 ±98.5	6.6* ±4.5
Group B (no OCT) n=40	Mean ±SD	26.83* ±8.1	-1.9 9±1.1	24.2* ±4.9	12.4 ±1.6	2.7 ±2.2	1726.0 ±899.0	-2.9 ±1.2	14.9* ±15.4	58.2* ±189.7	70.5* ±308.4	113.9 ±88.3	5.5 ±1.3

Legend: Ht- SDS=height SDS; FT4=free thyroxine; IGF-1-SDS=insulin like growth factor-1 SDS; LIC=liver iron concentration by FerriScan; ALT=alanine aminotransferase; AST=aspartate aminotransferase; ALP=alkaline phosphatase; (*p<0.05).

Growth and endocrine function in adults who received oral iron chelation (OIC) vs those who did not receive OIC before attaining final adult height

	Group A: OIC	Group B: No OIC
Number of patients	15	40
DM	6.6%	2.5%
IFG	6.6%	17.5%*
Hypothyroid	0.0%	10.0%*
IGF-1 <-2	20.0%	87.5%*
HtSDS < -2	6.6%	52.5%*
Hypogonadism	13.3%	40.0%*

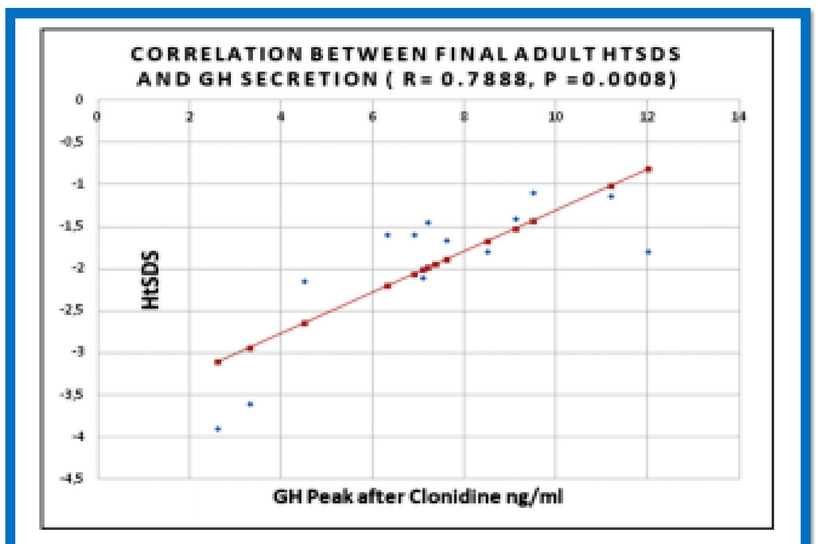

Figure 1. Correlation between peak GH secretion and final adult height expressed in SDS in patients with BTM

Conclusions

The use of OIC years before the end of puberty was associated with a significantly lower prevalence of endocrinopathies, lower LIC and higher FA-Ht. Proper blood transfusion and early use of intensive oral chelation can improve the final height of patients with thalassemia major.

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