Statural growth and prevalence of endocrinopathies in relation to liver iron content (LIC) in adult patients with beta thalassemia major (BTM) and sickle cell disease (SCD)

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Introduction

Despite regular blood transfusion and iron chelation therapy, growth impairment and pubertal delay are commonly seen in children and adolescents with transfusion-dependent Beta thalassaemia major (BTM) and sickle cell disease (SCD).

Objectives -Patients and Methods

We evaluated growth parameters and endocrine disorders in relation to the liver iron concentration (LIC) assessed by the Ferriscan® method in a cohort of adults with SCD (n=40) and BTM (n=52) receiving blood transfusions and iron chelation therapy since early childhood.

Before transfusion, hemoglobin concentration had not been less than 9 g/dl in the past 12 years; subcutaneous daily desferrioxamine was administered for all of them since early childhood (2-5 years of age). All patients were shifted to oral therapy with deferasirox iron chelation, 20 mg/daily for the past 5 years.

Results

BTM patients with higher LIC (>15 mg Fe/g dry weight) had significantly shorter stature, lower insulin-like growth factor-I SDS (IGF-I SDS), higher alanine transferase (ALT) and serum ferritin concentrations compared to thalassemic patients with lower LIC.

Patients with SCD with LIC >8 mg Fe/g dry weight had significantly shorter stature, lower IGF-I SDS and higher ALT compared to SCD patients with lower LIC.

Patients with BTM had significantly shorted final height (Ht-SDS), IGF-I SDS and FT4 level compared to patients with SCD.

LIC and mean fasting blood glucose (FBG) were significantly higher in patients with BTM compared to those with SCD.

The linear regression analysis showed a significant correlation between LIC and serum ferritin level in SCD and BTM.

LIC and serum ferritin level were also correlated significantly with IGF-I level in patients with BTM. LIC was correlated significantly with ALT in patients with BTM

Prevalence endocrinopathies in adult patients with BTM in relation to LIC.

Complication	LIC > 15 (n = 24) (I)	LIC 8 :15 (n = 7) (II)	LIC < 8 (n= 29) (III)	P value (I vs II)	P value (I vs III)
Ht- SDS < -2)	33%	14.0%	21%	0.34	0.34
IGF-I SDS < -2	66%	14.0%	14%	<0.001	<0.01
Hypothyroid	13.0%	14.0%	3.0 %	0.46	0.17
DM	29%	0 %	3%	0.16	0.0085
IFG	13%	29.0%	17.0 %	0.33	0.69
Hypogonadism	45%	14%	7%	0.15	0.00

Prevalence endocrinopathies in adult patients with SCD in relation to LIC.

Complications	LIC > 8 mg Fe/g d.w. (n = 8)	LIC < 8 mg Fe/g d.w. (n= 32)	p value
(Ht- SDS < -2)	25%	21.8%	0.8
IGF-I SDS < -2	50%	28%	0.24
Hypothyroidism	13%	0%	0.04
DM	25%	6.2 %	0.11
IFG	25%	40.6%	0.42
Hypogonadism	13%	6.2%	0.25

HT: standing height, IGF-I SDS: insulin-like growth factor 1 SDS, DM: diabetes mellitus, IFG: impaired fasting glucose

Conclusions

The prevalence of endocrinopathies especially hypothyroidism, DM, and hypogonadism was significantly higher in BTM patients versus SCD patients and higher in patients with higher LIC versus those with lower LIC. These complications occurred less frequently, but still considerable, in chronically transfused patients with SCD

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