

Final height and endocrine complications in patients with β -thalassemia intermedia: (TI) Our experience in non-transfused versus infrequently transfused patients and correlations with liver iron content

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INTRODUCTION

We evaluated the final height and the endocrine complications encountered in young adult patients with Thalassemia Intermedia (TI) followed at Hematology Section, Doha (Qatar) in relation to the liver iron content (LIC) in non-transfused versus infrequently transfused TI patients

Patients and Methods

28 young adults with TI who were randomly selected from the Hematology clinic, National Centre for Cancer Care and Research, Hamad Medical Corporation of Doha (Qatar).

Group 1 included 9 patients who did not receive any blood transfusion and

Group 2 included 19 patients who infrequently received a blood transfusion.

Data recorded included :

1. transfusion frequency, chelation therapy, and splenectomy, auxological and pubertal data .
2. Endocrine and hepatic complications were assessed.
3. Iron overload was assessed by direct (liver iron content; (LIC) and indirect methods; serum ferritin (SF), and
4. bone mass index (BMA) by dual-energy X-ray absorptiometry (DXA)

Results

1. Short stature [Final Height (Ht) SDS < -2] occurred in 25% of patients with no difference between the two groups of patients.
2. (IGF-1) SDS was low in 35.7 % of patients with no statistical difference between the two groups.
3. Impaired fasting blood glucose occurred in 17.8% of patients, diabetes mellitus in 25%.
4. Hypogonadotropic hypogonadism occurred in 10.7%
5. Morning cortisol was low in one patient.
6. No thyroid or hypo-parathyroid abnormalities detected.
7. Liver iron content (LIC) > 15 mg/g dry weight and SF > 2,000 ng/mL were detected in 75% of the patients.
8. Both LIC and SF were significantly higher in the transfused group (Group 2).

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Management

9. High liver enzyme level (ALT) was detected in 42.8 % of patients.

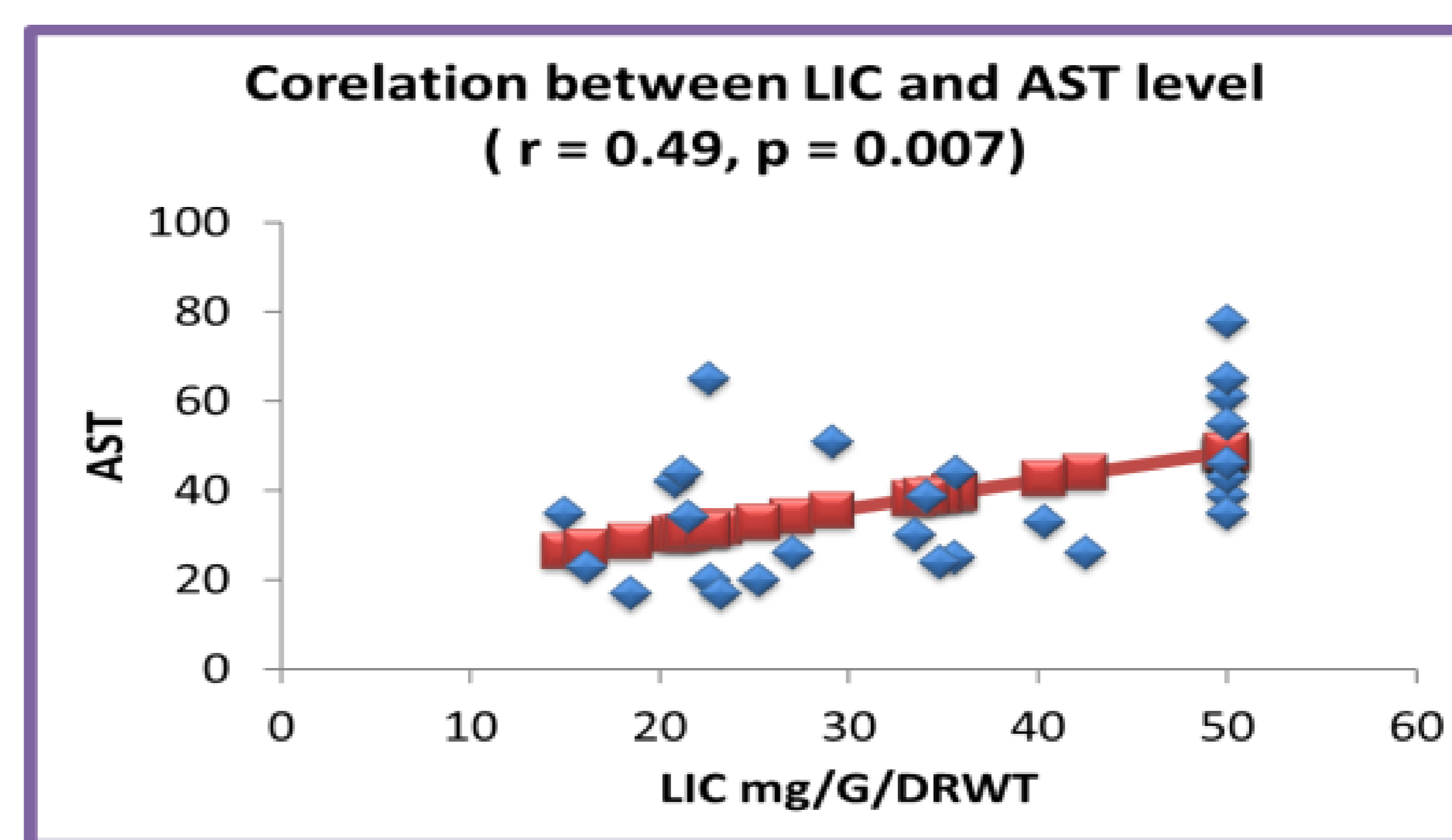
10. Total and fetal Hb was significantly higher in group 1 versus group 2.

11. Osteopenia was diagnosed in 14.3% of patients.

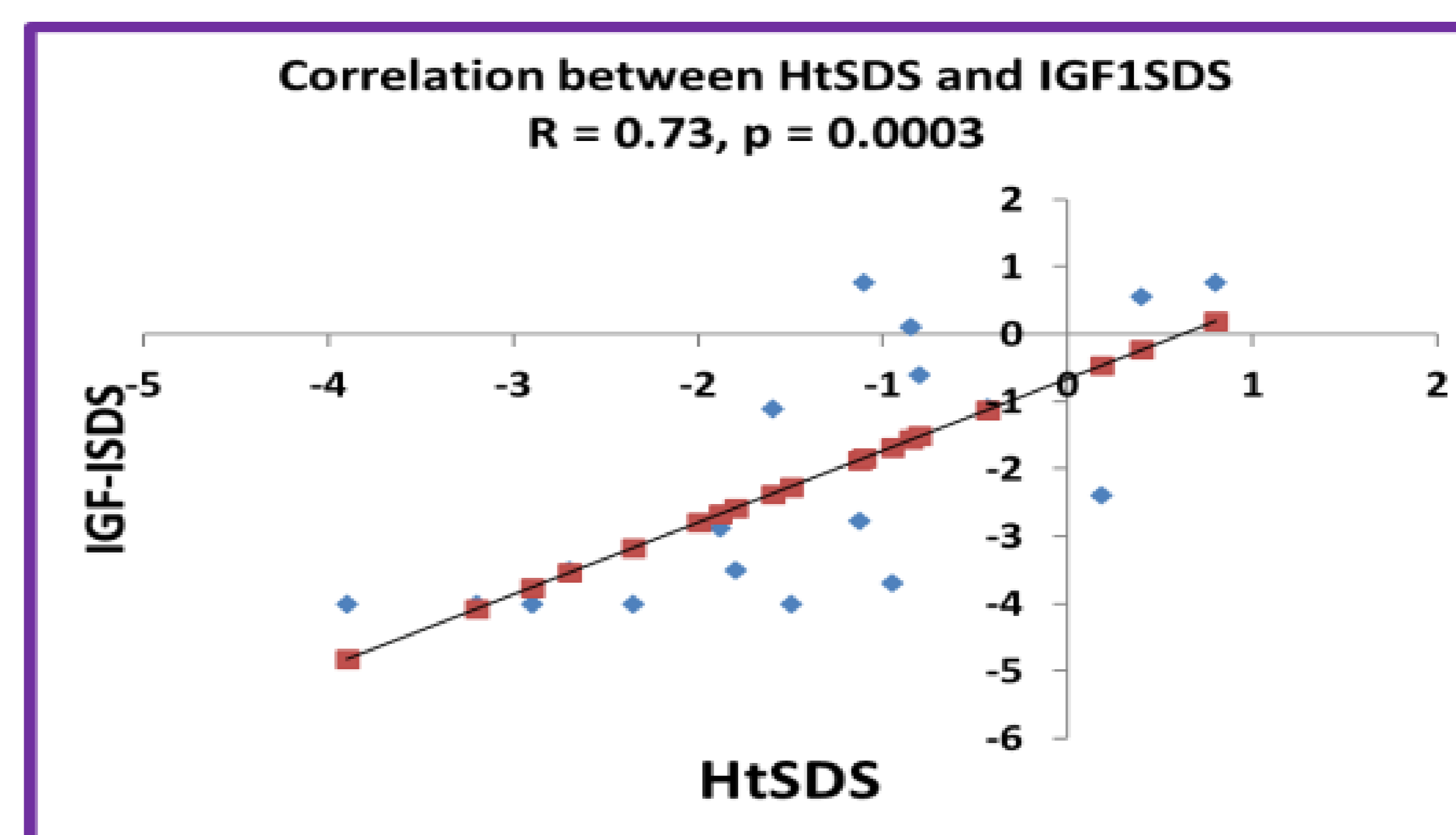
12. Females had significantly better final height SDS, higher IGF-1 SDS, lower LIC and fasting blood glucose level compared to males.

Ht-SDS was correlated significantly with IGF-1 SDS.

AST concentrations were correlated significantly with LIC and SF levels.



The HtSDS was correlated significantly with IGF1SDS



LIC was correlated significantly with ferritin concentrations.

Total and fetal Hb did not correlate significantly with Ht-SDS or IGF-1 level Ht-SDS was correlated significantly with IGF-1 SDS.

Conclusions

A significant number of TI patients have high LIC, short stature and endocrine disorders.

We also recommend close monitoring of endocrine and other complications, according to the international guidelines



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