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.

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)

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.