

Summary and Conclusion

Summary:

The present study was designed to assess the prevalence of delayed growth, delayed puberty and HCV infection in patients with hemolytic anemias. Our work was carried on 200 patients of hemolytic anemia, their ages ranged from 2 months up to 18 years, attending hematology clinic, Benha Specialized Children Hospital.

They were divided into 5 groups according to the type of hemolytic disease: Beta thalassemia major group (n=80), sickle cell anemia group (n=8), hereditary spherocytosis group (n=6), autoimmune hemolytic anemia group (n=6) and Glucose-6-phosphate dehydrogenase deficiency group (n=100). Beta thalassemia major constitutes 80% of chronic hemolytic pediatric patients.

All patients were subjected to detailed history taking, complete physical examination including: general examination Anthropometric measurement: weight and height and correlating them with normal measurement using growth chart, Pubertal status and complete systemic examination

All cases were investigated including Complete blood picture, Blood film, Reticulocytic count, Serum ferritin, Detection of hepatitis C virus antibodies

Appropriate investigations for hemolysis including:

Hemoglobin electrophoresis, Glucose-6-phosphate dehydrogenase activity, Osmotic Fragility tests, Coombs' test (direct and indirect) and Sickling cell test

Our study reveled the following results

There is no statistically significant difference between all groups of hemolytic anemia as regard general characteristic data (age, age of onset, gender and consanguinity).

All groups were matched as regard anthropometric measures (height,weight) and puberty. There was a statistically significant short stature, under weight and delayed puberty in chronic hemolytic studied group

There was no a statistically significant difference in the serum ferritin level among the studied groups. That it was found to be high among all patients with chronic hemolytic anemia . however, the highest values were found among some patients with beta thalassemia major.

As regard hepatitis C virus infection, There was a statistically significant increase in HCV infection in chronic hemolytic studied group with the highest value in 58 (72.5%) patients of beta thalassemia, 2(25%) patients of sickle cell disease and 5(83.3%) patients of auto immune hemolytic anemia. There was a significant increase in HCV infection in cases who had a higher average of transfusion numbers compared to negative cases, on the other hand, there was no significant difference of HCV infection as regard age of the first transfusion.

In our study, there was a significant increase splenectomy in all types of chronic hemolytic studied group. While, Age of splenectomy had no significance in all studied groups. splenectomy was done usually at the age of 6 years in 13 thalassemia major patients (16.25%), one patient sickle thalassemia (12.5%) and 4 cases of hereditary spherocytosis (66.6%).

In our study, Patients with serum ferritin ≥ 1000 ng/ml had significantly lower anthropometric measures and delayed puberty than those with serum ferritin < 1000 ng/ml. short stature was reported in 69 (86.25%) patients. delayed puberty reported in 16 (53.4%) of 30 beta thalassemia patients reach pubertal age, Mean serum ferritin level during the study period was 3984.09 ± 1759.10 ng/mL. Delayed puberty was noted in 53.4% of thalassemic patients who reached pubertal age, Mean ferritin level of 2560 ng/mL during puberty was the cut-off for hypogonadim and ferritin level of 2800 ng/mL during prepuberty was the cut-off for final short stature .

In our study, Age at starting chelation was significantly higher among short stature , delayed puberty , subjects had serum ferritin ≥ 1000 ng/ml. this reflect that delaying in starting chelation with subsequent iron overload play essential role in growth failure and delayed puberty

Our study revealed that, Repeated blood transfusion therapy produces chronic iron overload which causes many complications as frequent endocrine complications, delayed growth, delayed puberty and HCV infection in children.