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Research Article

Hematological Parameters in Beta Thalassemia Major Children

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Abstract
Background: Thalassemia is a hematologic disease caused by mutations in the genes coding for hemoglobin chains. The most common causes of genetic disorders in humans are mutations within the β-globin gene, of which 350 β-thalassemia mutations have been identified to date. The study aimed to assess hematological parameters in beta thalassemia major children in pediatric department of hematology, El-Minia university children and maternity hospital. **Methods:** This cross-sectional study was carried out from January 2021 to November 2021.Thirty beta thalassemic major children were taken during their regular follow up in the pediatric hematology outpatient clinic and hematology internal department, El-Minia University Children and Maternity Hospital the patients were divided into 2 groups; children underwent splenectomy and children without splenectomy. Blood samples were collected and analyzed for hematological parameters. The two groups: were subjected to careful detailed history taking, complete clinical examination, laboratory investigations including: complete blood count (CBC), serum ferritin. **Results**: There was a statistically significant difference as regard platelet count between the two groups (P value 0.047). splenectomized children showed a higher platelet count than the other group. There was statistically significantly increase in age (P value 0.006) and duration of disease (P value 0.014) in splenectomized children than non splenectomized children. **Conclusion:** The higher level of platlets in splenectomized children may be due to absence destruction of platelets by spleen.

Keywords; Beta Thalassemia; iron overload; hematological biomarker; ferritin.

Introduction
Thalassemia is the most common of all inherited disorders which are resulted by reduced or absent synthesis of the hemoglobin chains[(1)]. It is estimated that there are 270 million carriers of different hemoglobinopathies of which 30% are carriers of β-thalassemia in the world's population [(2)]. About 300,000–400,000 new cases are born with a serious hemoglobin disorder each year[(3)]. Ineffective erythron-poiesis is one of the major pathogenic mechanisms of disease manifestations, since years ago to now, blood transfusion has been used as regular therapy[(4)]. This makes them particularly vulnerable to iron overload in several organs that led to organ dysfunction and serious complications such as cardiac complications[(5)].

Patients and methods
This cross-sectional study included thirty beta thalassemic major children. patients were taken during their regular follow up in the pediatric hematology outpatient clinic and hematology internal department, El-Minia University Children and Maternity Hospital.

Our children were ranging from 6 to 16 years. They were classified into; Group (a): splenectomized children and group (b): non splenectomized children.

Then included children were subjected to the following:
a- ***Careful history taking including:***
Name, age, sex, residence, socioeconomic standard, and family history of blood diseases.

b. ***Full clinical examination: including***
1- Vital data: respiratory rate, heart rate, blood pressure, temperature.
2- Systemic ex: full chest, cardiac and abdominal examinations.
c- ***Laboratory investigation:*** CBC, serum ferritin.

Results

There was statistically significantly increase in age (P value 0.006) and duration of disease (P value 0.014) in splenecto-mized children than non splenictomized children. (Table 1).

There was a statistically significant difference as regard platelet count between the two groups. splenectomized children showed a higher platelet count than the other group (P value 0.047). (Table2).

Regarding serum ferritin level, our results showed in significantly increase in serum ferritin level in splenectomized group than the other group (P value 0.4). (Table2).

There was statistically insignificantly differ-rence in hemoglobin level between splenectomized children and non splenic-tomized children. (P value 0.812).(Table 2)

There was statistically insignificantly increase in total leucocytic count in splenectomized children than non splenic-tomized children. (P value 0.841).(Table 2)

Table1: Comparison between Splenectomized vs non Splenectomized patients as regarding demographic date

Splenectomized vs non Splenectomized patients	Splenectomized patients (N = 11)		Non Splenectomized patients (N = 19)		P value
	Mean ± SD	Range	Mean ± SD	Range	
Age (yr.)	12.4±3.62	7 -16	8.81±3.09	6 -16	0.006*
Sex: **Male N (%)** **Female N (%)**	5 (45.5%) 6 (54.5%)		8 (42.1%) 11 (57.9%)		0.858
Duration of disease	11.4±3.63	6 -15.6	8.06±2.9	5 -15	0.014*

Table 2: Comparison between Splenectomized vs non Splenectomized patients as regarding hematological date

Splenectomized vs non Splenectomized patients	Splenectomized patients (N = 11)		Non Splenectomized patients (N = 19)		P value
	Mean ± SD	Range	Mean± SD	Range	
Hb (g/ dl)	7.21±0.54	6.5 -8.1	7.23±0.84	6 -8.7	0.812
TLCs ($\times 10^3$ /cmm)	12.3±6.43	6.7 -25.3	11.4±4.36	4.6 -19.2	0.846
platelets ($\times 10^3$/cmm)	453±149	208 - 625	335±90.5	184 -515	0.047*
Ferritin (ng /ml)	2065±772	1267-3415	2109±694	1200-3350	0.74

Discussion

β-thalassemia major (TM) is one of the most prevalent inherited hemoglobino-pathies. It has one of the highest prevalences of transfusion-dependent TM patients globally, with an estimated greater than 100,000 active cases. Blood transfusions (BT) are essential in the management of severe TM [(6)]. In our study is that; there is a higher platelet level in

splenectomized patients than the other group and this was statistically significant (P value 0.047). Regarding the thrombocytosis in our splenectomized patients, the thrombocytosis persists indefinitely after splenectomy. This usually appears to be a consequence of continuing anemia with a hyperplastic marrow[7,8]. Although reactive thrombocytosis is not usually associated with thromboembolic problems, the high Platelet count may have contributed to the serious and sometimes fatal episodes of pulmonary embolism and deep vein thrombosis that have occurred following splenectomy in some of our deceased βTM patients. Therefore, antiplatelet therapy is mandatory given to our splenectomized patients to avoid thromboembolic problems. Splenectomy and transfusion naivety have been considered an important risk factor for hypercoagulability and thromboembolic events in thalassemic patients [9].

The study of Ammara et al., 2014, about the long-term follow-up after splenectomy in thalassemia patients revealed thrombocytosis and the risk of thromboembolism. Moreover, splenectomy has been considered as an important risk factor for infections;[10] therefore, proper preoperative vaccination can reduce the risk of overwhelming post-splenectomy infections [11]. The current study showed that there was statistically non-significant increase in serum ferritin level in splenectomized group than the other group as splenectomy did not reduce the iron burden or the requirement for blood transfusions in various transfusion-dependent disorders [12]. After splenectomy there will be a potential shift of the splenic iron to extra splenic tissue, and further increase in iron overload should be borne in mind in considering removal of this organ [13].

Conclusion

Splenectomy increase incidence of thrombocytosis may be due to a consequence of hyperplastic marrow. the high Platelet count may have contributed to the serious and sometimes fatal episodes of pulmonary embolism and deep vein thrombosis that has occurred following splenectomy in some of our deceased βTM patients.

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