A Study Comparing Blood Transfusion Requirement And Frequency Of Blood Transfusion In Splenectomized And Non-Splenectomized Children With Beta Thalassemia Major - Our Institutional Experience

* Dr. Prajakta Ghatage, ** Dr. Ramesh Kothari,*** Dr. Sunil Mhakse

* Resident, ** Professor and Head,Department of Paediatrics, *** Dean, Dr Vithalrao Vikhe Patil Foundation's Medical College, Ahmednagar

Corresponding Author : Dr. Prajakta Ghatage

Mail id : prajaktaghatage38@gmail.com

Mobile No.: 7276613919

Address : Department of Paediatrics, Dr. Vithalrao Vikhe Patil Foundation's Medical College, Ahmednagar

Abstract :

Introduction : Beta thalassemia major is an autosomal recessive blood disorder. Thalassemia refers to a group of genetic disorders of globin chain production in which there is an imbalance between the α-globin and β-globin chain production. β-Thalassemia syndromes result from a decrease in β-globin chains, which results in a relative excess of α-globin chains. **Material and Methods :** Sample size : 45. Duration of Study: June 2016- June 2018. Children from 5 to 15 years of age who are diagnosed case of Beta thalassemia Major with blood requirement more than 200 ml/kg/year. The patients were divided into 2 groups: 5 to 10 years and 11 to 15 years. History and complete physical examination with haematological workup were done in all cases. Pre-transfusion haemoglobin of all 45 patients was noted. The risks and benefits of splenectomy were explained, 20 patients were selected for splenectomy. **Result :** From the Age group of 5 to 10 years there were Total 22 patients out of which 11 underwent splenectomy. The normal haemoglobin level in the children of this age group is 12.5 gm/dl on an average. The average haemoglobin level in non-splenectomised patients were 7 gm/dl. On the other hand the average haemoglobin level in a splenectomised patient was 9 gm/dl, 6 months after splenectomy. The annual blood requirement of all splenectomised and non-splenectomised patients was compared. In non splenectomised patients the annual requirement was 260 ml/kg/year while in splenectomised patients it was 210 ml/kg/year. **Conclusion :** The mean haemoglobin levels were on a higher side in splectomized patients as compared to their pre-operative levels as well as compared to non-splenectomized patients. The blood requirement significantly reduced in splenectomized patients to 210 ml/kg/year.

Keyword : Thalassemia Major, Splenectomy, Blood transfusion.

Introduction : Beta thalassemia major is an autosomal recessive blood disorder. Thalassemia refers to a group of genetic disorders of globin chain production in which there is an imbalance between the α-globin and β-globin chain production. β-Thalassemia syndromes result from a decrease in β-globin chains, which results in a relative excess of α-globin chains. The free α-globin chains and inclusions are very unstable, precipitate in red cell precursors, damage the red cell membrane, and shorten red cell survival leading to anemia and increased erythroid production.[1]

The clinical features of Thalassemia Major are pallor, jaundice, Hepatosplenomegaly. The other feature include thalassemic facies (maxilla hyperplasia, flat nasal bridge, frontal bossing), pathologic bone fractures, marked hepatosplenomegaly, and cachexia. If not treated, children with homozygous β0-thalassemia usually become symptomatic from progressive hemolytic anemia, with profound weakness and cardiac decompensation during the 2nd 6 months of life. Depending on the mutation and degree of fetal hemoglobin production, transfusions in β-thalassemia major are necessary beginning in the 2nd month to a 2nd year of life.[2]

Bone marrow transplantation, the only curative treatment is available to very few patients and most of the patients are treated by regular blood transfusion and iron chelation. The frequency of blood transfusion and the amount of blood transfusion requirement keep on increasing with increasing age leading to iron overload.

Splenectomy has a role in reducing the frequency of blood transfusion and transfusion related complications.[3]

Mass effect symptoms may be considered as one of the indications for splenectomy in children with Thalassemia major. Another important indication is repeated episodes of abdominal pain caused by infarctions in the spleen.

Splenectomy can be done by open, Laparoscopic or robotic methods. However most commonly used approach for elective surgical management for massive splenomegaly is open approach.[4]

Material and Methods :

- **Sample size :** 45
- **Duration of Study :** June 2016- June 2018.
- **Inclusion criteria :** Children from 5 to 15 years of age who are diagnosed case of Beta thalassemia Major with blood requirement more than 200 ml/kg/year.
- **Exclusion Criteria :**

1. Thalassemia minor and intermedia.
2. Patients with non-compliance to regular treatment.
3. Patients whose parents were not giving consent for the study.

The patients were divided into 2 groups : 5 to 10 years and 11 to 15 years. History and complete physical examination with haematological workup was done in all cases. Pre transfusion haemoglobin of all 45 patients was noted.

The frequency of blood transfusion was noted in all patients and the requirement was above 200 ml/kg/ year in all the patients.

Hence the risks and benefits of splenectomy were explained, 20 patients were selected for splenectomy. After giving Pneumococcal, Hemophilus Influenzae and Varicella vaccines the patients underwent splenectomy.

Haemoglobin Levels of all 45 patients were followed up and the frequency of blood transfusion noted. The haemoglobin levels in splenectomized and non-splenectomized patients as well as pre and post-operative levels in splenectomized patients were noted.

All the procedures were done by equally experienced surgeons by Department of Surgery. Splenectomy was done by left subcostal incision, followed by mobilization of the ligaments of spleen including Splenophrenic ligament, Splenorenal ligament, Gastrosplenic ligament and Splenocolic ligament. Then approaching to the hilum of spleen and ligating splenic artery and vein which is finally divided.

Results :

The two groups were compared on the basis of

1. Haemoglobin levels
2. The frequency of blood transfusion
3. Annual blood requirement

1. Heamoglobin Level :

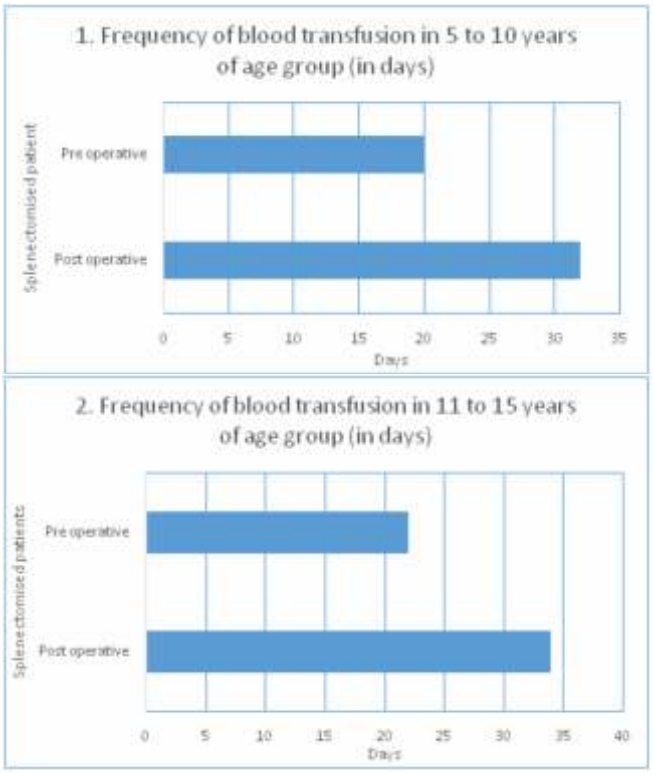
Age Group	Number (n)	Normal Levels	Non-Splenectomised patients	Splenectomised patients
5 to 10 years	22	12.5 gm/dl	7 gm/dl	9 gm/dl
11 to 15 years	23	13.7 gm/d/	8 gm/dl	8.5 gm/dl

From the age group of 5 to 10 years there were total 22 patients out of which 11 underwent splenectomy. The normal haemoglobin level in the children of this age group is 12.5 gm/dl on an average. The average haemoglobin levels in non-splenectomised patients was 7 gm/dl. On the other hand the average haemoglobin level in splenectomised patient was 9 gm/dl, 6 months after splenectomy. The Normal Haeamoglobin level from 11 to 15yeas is 13.7 gm/dl. on average. In non-splenectomised group it was 8 gm/dl and splenectomised was 8.5 gm / dl.

2. The frequency of blood transfusion : The frequency of blood transfusion was compared in splenectomised and non splenectomised patients as well as pre-operative and post-operative blood transfusion frequency in splenectomised patients.

The frequency of blood transfusion pre-operatively in 5 to 10 years age group was an average of 20 days which after splenectomy was an average of 32 days. The average frequency of blood requirement in the age group of 11 to 15 years was 22 pre-operatively and 34 post splenectomy.

The frequency of blood transfusion in non-splenctomised patients was an average of 20 days in 5 to 10 age group and 21 in 11 to 15 age group which further increased in frequency in follow up of 2 year time span.



3. Annual blood requirement : The annual blood requirement of all splenectomised and non-splenectomised patients was compared. In non splenectomised patients the annual requirement was 260 ml/kg/year while in splenectomised patients it was 210 ml/kg/year.

Non-Splenectomised patients	Splenectomised patients
260 ml/kg/year	210 ml/kg/year

Discussion : Beta thalassemia Major is a haemoglobinopathy with defect in beta-globin chain synthesis. Due to excess alpha chains, the RBC are prematurely lysed and the iron in deposited in body tissues.

The life span of RBCs is further reduced due to secondary hypersplenism which is seen in thalassemia. Abolition of secondary hypersplenism is the rationale for splenectomy.[5]

Propper et al[6] have suggested that persistent maintenance of haemoglobin above 10 gm/dl is required for preventing overt hypersplenism early in life and since patients of beta thalassemia demonstrate a gradual increase in transfusion requirement, enlargement of the spleen may result in iron overload.

Cohen et al[7] suggest splenectomy when blood transfusion is more than 200 ml/kg/year.

The basic inborn haemoglobinopathy is not corrected after splenectomy and also it further increases the chances of life threatening infections in splenectomized patients, some authors do not consider splenectomy of any benefit.[8]

But in our study, a significant reduction of blood requirement and increased level of haemoglobin was observed. And no infectious complications were noted post-operatively.

Conclusion : The mean haemoglobin levels were on a higher side in splectomized patients as compared to their pre-operative levels as well as compared to non-splenectomized patients. The blood requirement significantly reduced in splenectomized patients to 210 ml/kg/year. The frequency of transfusion was reduced as so were the long and short term transfusion related complications overall establishing the beneficiary role of splenectomy in patients with Beta thalassemia major.

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