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A Comparative Study of Blood Transfusion versus Splenectomy in Thalassemia Major Patients with Splenomegaly

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Abstract: ***Basics and Objectives****: Thalassemia represents an inherited disorder with either lack of or reduced hemoglobin beta or alpha chains production with destruction of red blood cells in spleen resulting in splenomegaly. Blood transfusions are usually performed in patients of Thalassemia major frequently but few Patients with Splenomegaly having increased blood requirement that prevents control with iron chelating agents, hypersplenism with Pancytopenias and symptomatic Splenomegaly with left upper quadrant pain require elective splenectomy respectively. This study includes observational study of 50 Patients with Thalassemia major admitted in Paediatric department who have been treated either conservatively with only blood transfusions or undergone elective splenectomy and comparing the outcomes in both cases. Our study results confirm that elective Splenectomy patients had better outcomes compared to those who were treated only conservatively with blood transfusions every 2 - 3 weeks.*

Keywords: Splenectomy, Hypersplenism, Siderosis

1. Introduction

Thalassemia is an inherited autosomal recessive disorder where there is reduced or absent alpha or beta hemoglobin chain synthesis. Beta Thalassemia major (Cooleys anemia) is defined as complete absence of beta chain synthesis leading to abnormal shape of red blood cells that are destroyed by reticuloendothelial system particularly spleen leading to Splenomegaly, Anaemia and iron overload presenting with symptoms within 1st year of life and these patients are severely transfusion dependent. Beta Thalassemia minor is moderate suppression of beta chain synthesis and these patients are usually asymptomatic. Beta Thalassemia intermedia patients require occasional blood transfusions and presents with symptoms later in life. Patients with Thalassemia major are managed with frequent blood transfusions and iron chelation therapy to prevent iron overload. Candidates for Splenectomy were selected if the patients had - increased blood transfusion requirement that prevents control with iron chelation therapy, Hypersplenism with Pancytopenia and symptomatic Splenomegaly with left upper quadrant pain. There are various complications after Splenectomy that are prevented with pre - operative vaccination and post - operative long - acting antibiotics as well.

2. Methods

It is an observational study done in the department of general surgery in association with department of Paediatrics and department of Haematology during the period of May, 2019 to May, 2021. A total of 50 patients suffering from Thalassemia major who had undergone either Splenectomy or went for conservative treatment with blood transfusion only. Patients were assessed based on blood investigations, chest x - ray, Echocardiography, Abdominal ultrasound, clinical examinations with detailed family history. Prophylaxis was given with vaccines against Pneumococcus, Heamophilus influenza type - B and Meningococcus - C 2weeks before surgery. Among those Patients who undergone Splenectomy were given post - operative care in surgical ward and Ryles tube was inserted before surgery and removed 48 hours after surgery and patients were discharged with proper advice for follow up for suture removal and with meningococcal vaccine to be repeated every 5years and Haemophilus influenza - B vaccine to be repeated every 10years. Pneumococcal vaccine is given to those patients only aged over 2 years. Antibiotic prophylaxis was given with long acting Penicillins on discharge. Patients with asymptomatic Splenomegaly with requiring frequent blood transfusions were also studied.

3. Results

Total number of patients was 50 out of which 34 were male and 16 were female with a male to female ratio of 2: 1. Age group of patients was 8 - 15 years respectively. Splenectomy was done in 5 (10%) patients with Hypersplenism and Pancytopenia and 3 (6%) patients with left upper quadrant pain and rest of 42 (84%) patients were given blood transfusions only who had asymptomatic Splenomegaly with risk for surgery.

Table - 1

Interval of blood transfusion before Splenectomy was 2 - 3weeks and overall volume of blood transfusion was 260 - 300ml/kg/year . After Splenectomy interval of blood transfusion was 1.5 - 2 months and overall volume of blood transfusion was 150 - 175ml/kg/year. They were kept on iron Chelation therapy post - operatively to prevent iron overload due to blood transfusions. Blood transfusion requirement and number of follow - ups to clinics reduced and interval between blood transfusions is increased. In majority of patients pre - operative serum ferritin levels were found to be more than 2000 ng/ml even with iron chelation therapy but post - operative serum ferritin levels were maintained less than 900ng/ml with iron chelation therapy.

Table - 2

Out of 42 patients who were on frequent and repeated blood transfusions with iron chelation therapy and not undergone Splenectomy, about 9 (21.4%) patients developed secondary heart failure due to cardiac Siderosis, 6 (14.3%) patients developed short stature and growth retardation due pituitary dysfunction, 5 (11.9%) patients developed Hypothyroidism due to Thyroid dysfunction, 4 (9.5%) patients developed diabetes mellitus due to pancreatic insufficiency secondary to iron - deposition, 3 (7.14%) Patients developed liver cirrhosis, 1 (2.4%) patient developed Hypogonadism and 2 (4.8%) patients developed leg Ulcers. Iron overload in Thalassemia major is due to increased blood transfusions and increased intestinal absorption of iron. This excess of non - transferrin bound iron is transported through calcium channels into respective tissues like Liver, Heart, Spleen, Pancreas, Pituitary gland, Thyroid glands etc. where reactive oxygen species formed by metabolism of this non - transferrin bound iron leads to tissue necrosis and apoptosis. Out of 42 Patients, 30 Patients developed complications due to iron overload with serum ferritin levels between 1500 - 1900ng/ml due to poor compliance to iron Chelation therapy and rest of 12 Patients didn't have any complications at all with serum ferritin levels between 600 - 900ng/ml respectively with good compliance to iron Chelation therapy.

Table 1: Changes in overall volume of blood transfusion and frequency of clinic follow ups for blood transfusions before and after Splenectomy

	Overall volume of blood transfusion (ml/kg/year)	Interval of blood transfusions
1 - Before Splenectomy	260 - 300ml/kg/year	Every 2 - 3 weeks
2 - After Splenectomy	150 - 175 ml/kg/year	Every 1.5 - 2 months

Table 2: Complications in patients of Thalassemia major who were given only blood transfusions without Splenectomy (total patients with only blood transfusions - 42)

Percentage of patients (%)	Complications
1- 21.4 %	Heart failure with Cardiomyopathy due to Cardiac siderosis
2- 14.3 %	Short stature and growth retardation due to Pituitary dysfunction
3- 11.9 %	Hypothyroidism due to Thyroid dysfunction
4- 9.5%	Diabetes mellitus due to Pancreatic insufficiency
5- 7.14%	Hepatic dysfunction due to hepatic siderosis
6- 2.4%	Hypogonadism
7- 4.8%	Leg ulcers

4. Discussion

50 patients with Thalassemia major were included in this observational study with 34 male and 16 female with male to female ratio of 2: 1 indicates the disorder is more common in males and the age group was 8 - 15 years. Thalassemia presents with symptoms very early in age and Splenectomy is usually done by 1st - 2nd decade of life. In this study of 50 patients about 8 patients undergone Splenectomy and the major indication was Hypersplenism with Pancytopenia (10 %) and left - upper quadrant pain (6%). It was found that patients who undergone elective Splenectomy had reduced overall volume of blood transfusion and increased interval of blood transfusion and clinical follow ups respectively. After Splenectomy serum Ferritin levels were reduced with continuation of blood transfusions and iron Chelation therapy. Variety of iron chelating agents used were Deferoxamine, Deferiprone and Deferasirox. Post - operative patients were monitored for adverse effects of iron chelating agents. Patients on Deferasirox are prone to develop raised serum Creatinine, increased liver enzymes and gastrointestinal disturbances. Patients on deferiprone may develop gastrointestinal disturbances with Agranulocytosis, Arthralgia and increased liver Enzymes. Those on deferoxamine may have infusion site reactions, retinal and auditory complications. Iron chelation therapy is costly and poor patients may not afford such therapy and hence Splenectomy especially in government set ups have reduced the financial stress on poor families with improved compliance of patients for follow ups with reduced blood transfusions and reduced complications due to iron overload in tissues with improved quality of patient's life compared to conservative approach of frequent blood transfusions only with complications of iron overload even with iron chelation therapy and cost of iron chelation therapy posing financial and psychological burden on patient's family with frequent follow ups to clinic with overall increased blood transfusions. Splenectomy with prophylaxis via vaccinations and long acting Penicillins reduced the chances of sepsis after Splenectomy. One of the most common complications found in patients of thalassemia major due to blood transfusion is Hepatitis - C infection.

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