ORIGINAL ARTICLE

Improved quality of life in post splenectomized thalassemic patients

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Abstract

Objective: To see the change in quality of life and the effect on transfusion requirement after splenectomy in thalassemia major patients.

Study Design: Cohort study.

Setting and Duration: Liaquat National Hospital and Medical Colege, Karachi from January 2009 to December 2010.

Methodology: We reviewed 28 patients with Thalassemia Major referred by Thalassemia Control Centre (TCC) at Hussaini Blood Bank and underwent splenectomy at Liaquat National Hospital. Data regarding age, sex, indication for splenectomy, morbidity and mortality associated with the procedure was collected from the hospital records, and change in transfusion requirements was collected from the TCC. Change in quality of life was assessed by using a questionnaire that was filled by the patients or their families.

Results: Of the 28 patients, 15 (53.5%) were males and 13(46.5%) females. Mean age was 23 years (range 5-26).The mean unit of blood transfusions over a six month period prior to splenectomy was 18.1, which reduced to 9.09 over six months, post-surgery. One patient expired on the first post-operative day due to intractable cardiac arrhythmias. No incidence of post-splenectomy severe infection, or long term morbidity was recorded. Majority of the patients (90.4 %) felt a definite improvement in the quality of life after splenectomy. Interestingly 95 % of the patients were happy with the outcome because of reduced number of transfusion and a decrease in hospital visits.

Conclusion: Splenectomy in thalessemia is a safe procedure, and in children with massive splenomegaly leading to high transfusion requirements, selective splenectomy may help reduce this requirement and improve their quality of life.

Keywords: Thalassaemia, Splenectomy, Blood Transfusions, Quality of life

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Introduction:

The spleen has important functions related to cell mediated immunity and filtration of abnormal blood cells. Removal of the spleen affects an individual's immunity, especially against encapsulated pathogens. However, splenectomy is justified in patients with Thalassaemia Major as it has been shown to decrease transfusion requirements[1]. and may help with symptoms of right upper quadrant pain and risk of splenic rupture in patients with massive splenomegaly[2]. Patients who require more than 200-250 ml/kg of packed red blood cells (PRBC) per year to maintain a Hb level around 10 mg/dl benefit from splenectomy. The risk of severe infections and malaria in the immunocompromisation associated with spleen removal are the chief concerns when deciding about splenectomy in these patient[3]. Pre-operative immunization against encapsulated bacteria and prophylactic antibiotics should be routinely prescribed to minimize this risk[4].

The surgical options available for removal of spleen in these patients include traditional open splenectomy, partial splenectomy, laparoscopic

splenectomy and splenic embolization[5,6]. Indication of splenectomy for hematological diseases and complications after splenectomy in children have been described however there is paucity of literature regarding the outcome analysis in terms of improvement in quality of life. The aim of this study was to see if the splenctomized thalassaemic patients actually benefited after the procedure with a decrease in transfusion requirement and improvement in general quality of life.

Methodology:

Thalassaemia patients are registered at the Thalassaemia Control Centre (TCC) at Hussaini Blood Bank. Detailed records from regular visits are kept at the centre. Patients whose transfusion requirements exceed the protocols, who develop massive splenomegaly or have hypersplenism, are referred for splenectomy. We reviewed 28 patients with Thalassaemia Major who underwent splenectomy at Liaquat National Hospital (LNH) from January 2009 to December 2010.

All patients were immunized against encapsulated bacteria at least two weeks prior to surgery. Pre operative investigations included recent hematological status, ECHO and evaluation of liver function. Open Splenectomy was performed by a single surgeon through a left sub costal incision, splenenculi, if any, were removed and patient was kept post operatively in the ward on IV antibiotics and analgesics. The patient was discharged after recovery from the procedure on monthly Long acting Penicillin and appropriate instructions regarding Opportunistic Post Splenectomy Infections. Follow up was conducted at LNH and TCC. Data regarding age, sex, indication for splenectomy, hospital stay, morbidity/mortality associated with the procedure was collected from the hospital records. Incidence of post-splenectomy sepsis was recorded from follow-up notes.

Data for transfusion requirements 6 months prior to surgery and 6 months after surgery (2 patients had only a 4 month record of transfusion prior to and after surgery) was collected from the TCC. Change in quality of life was assessed by using a questionnaire that was filled by the patients or their families. The Parameters assessed included: Change in school/work attendance, change in participation in outdoor activity/sports, long term discomfort resulted by the procedure, change in social activity level, change in number of hospital visits for transfusion/checkup, and self assessment of any change in quality of life.

Results:

Out of the 28 patients, 15 (53.5%) were males and 13 (46.4%) females. Mean age was 23 years (range 5-26). The average hospital stay was 5.5 days. Range of Follow up period was form 4 months to 2 years. One patient expired on the first post-operative day due to intractable cardiac arrhythmias (Mortality 3.5 %). No incidence of post-splenectomy severe infection, or long term morbidity was recorded.

Out of 28 patients, 7 could not be assessed for quality of life assessment. Transfusion requirements before and after surgery were not available for 5 patients who were stationed out of the city. The mean unit of blood transfusions over a six month period prior to splenectomy was 18.1, which reduced to 9.09 in six months, post-surgery.

Quality of life was assessed for only 21 patients who were available for interview regarding the impact of splenectomy on their general wellbeing. Out of 21 patients only 10 attended school or worked somewhere prior to surgery. 5 (50 %) out of these 10 patients had a definite improvement in attendance at school/work. 42.8 % felt a definite improvement in sporting/outdoor activities whereas 47.6 % felt no change in activity level. Only 4 patients felt that the procedure had resulted in long term discomfort in terms of pain and distress.

Majority of the patients (90.4 %) felt a definite improvement in the quality of life after splenectomy. Interestingly 95 % of the patients were happy with the outcome because of reduced number of transfusion and a decrease in hospital visits.

Table 1: *Quality of life assesment after splenectomy (n = 21)*

Change in average number of days missed from school/work after splenectomy:		
Improvement	5	23.8%
No Change	3	14.2%
Worsened	2	9.5%
Do not attend/ never attnded prior to surgery	11	52.3%
Change in patients ability to participate in games/sports after splenectomy:		
Improvement	9	42.8%
No Change	10	47.6%
Worsened	2	9.5%
Long term pain/discomfort/distress resulted by the procedure:		
Yes	4	19.0%
No	16	76.1%
Don't Know	1	04.7%
Change in patients ability to be involved socially after splenectomy		
Improvement	8	38.0%
No Change	10	47.6%
Worsened	3	14.2%
Change in the number of visits to hospital/clinic for transfusion/ check up after splenectomy		
Improvement	20	95.2%
No Change	1	04.7%
Worsened	0	0%
Improvement in quality of life after splenectomy:		
Yes	19	90.4%
No	2	9.52%
Don't Know	0	0%

Discussion:

Open splenectomy is a widely performed procedure in the pediatric age group for various hematological disorders. In patients with thalassaemia who have massively enlarged spleens, the high transfusion requirements are decreased after splenectomy[1]. Iron overload is directly proportional to the number of blood transfusions[7,8] and may cause mortality due to cardiac or liver failure. Significant morbidity due to organ damage is also associated with high transfusion rates, along with the risk of infection transmission[9, 10]. The decrease in transfusion justifies splenectomy despite the risk of post-splenectomy severe or even fatal infection

Chelation therapies and transfusion regimens have improved the survival rates in patients with thalessemia[11, 12]. Our review of literature showed dearth of data regarding change in the quality of life in these patients. Surgical procedures that retain a part of spleen, such as partial splenectomy, partial splenic embolization[13-15] or slice grafting may further help improve the quality of life in these patients and at the same time preserve some immune function in this vulnerable age group.

A study among local population, conducted on 20 patients who underwent Splenectomy for thalassaemia showed that it is a relatively safe procedure after proper preoperative anesthesia and cardiac assessment. None of their patient suffered from serious infection related to splenectomy. Preoperative vaccination was done for all patients. Complication like wound infection (10 %) and bleeding from splenic bed (5%) was observed. They had a mortality of 5 %. A significant reduction was noted in the number of transfusions after the procedure [16].

We performed splenectomy for 28 thalassaemic patients over a two year period. Only one patient expired in the immediate post operative period due to cardiac arrhythmia. None of the patients had any post surgical complications like, hemorrhage, wound infection, pancreatic trauma. All patients were immunized prior to surgery and recent hematological, liver functions and cardiac assessment was performed. Post operatively they were given intramuscular long acting penicillin prophylaxis. There were no sepsis related complications. 21 of the patients, who were interviewed regarding an improvement in quality of life, were happy with the procedure. Our patients had a significant improvement in the quality of life post-splenectomy including improvement in pain, ability to interact socially, improved attendance at school and participation in sports. Many of them suggested that this procedure should be offered to other thalassaemics, more so, at an earlier stage, as it helps in improving the quality of life.

Conclusion:

Most of the thalassaemic patients suffer from years of worsening medical condition and health due to repeated transfusions and rising serum ferritin levels. It is arguable that splenectomy to

these children should be offered at an earlier age so that they can benefit from the procedure and become an active member of society.

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