Experience of Splenectomy in Thalassaemia

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Fourteen Thalassaemic, male and female patients, age ranging from 5 years to 20 years, under went splenectomy in our surgical unit after careful preparation. Non of them required more than one transfusion during the procedure. Respiratory and wound infection was the problem in 9 out of 14 patients. One bled through the abdominal drain site and managed with pressure dressing. No serious sepsis or mortality was observed in this series.

Key words: Thalassaemia, splenectomy

These hereditary haemolytic anemias result from a defect in haemoglobin synthesis in which one of the haemoglobin polypeptide chains is synthesized at a markedly reduced rate. Specific pairs of genes are responsible for synthesis of the alpha, beta, gamma, and delta chains of the haemoglobin molecule and a deficiency in synthesis of one of these subunits may lead to one of the thalassaemia syndromes. Thalassaemia is classified by the deficient peptide chain[5,6] Beta-Thalassaemia, in which there is a quantitative reduction in the rate of beta chain synthesis, is the most common type of thalassaemia. When the abnormal gene is inherited from both parents (homozygous), severe anemia termed thalassaemia major results. Heterozygous patients have a mild anemia termed thalassaemia minor. The term thalassaemia intermedia is used to describe some homozygous patients who have a milder than usual course and some heterozygous patients who have a more severe course than usual[6]. In thalassaemia major, the reduction in the rate of beta chain synthesis produce a marked decrease in the amount of normal adult haemoglobin (HbA) with a compensatory increase in fetal haemoglobin (HbF). Elective splenectomy is most commonly performed for haematological disorders. The operation is usually performed through an upper midline incision but subcostal and thoraco-abdominal incisions as also used. If the procedure is indicated for blood dysrasias it is important to make a careful search for splenunculi which should be removed[2].

Aims and Objectives:

The study was directed towards following objectives.

- To highlight the benefits of splenectomy in thalassaemia syndromes.
- To observe the complications associated with splenectomy in thalassaemic patients.

Material and methods

This prospective study of 14 patients was conducted in Sir Ganga Ram Hospital from May 2002 to November 2002 in collaboration with the Thalassaemia Center, Department of Paediatrics, Fatima Jinnah Medical College, Lahore. These patients were first seen in their center, investigated, selected and prepared for surgery. Splencetomy was carried out by our department, Surgical Unit-III, of Sir Ganga Ram Hospital, Lahore.

Once the children were in the surgical ward, a detail clinical examination was performed which revealed retarded growth, pallor, occasional jaundice and hepatosplenomegaly. Laboratory investigation showed haemoglobin to be around 6gm [increased to 8gm just prior to surgery], Increased total bilirubin, increased indirect bilirubin, abnormal LFT's reticulocytosis, leucopenia and thrombocytopenia, PT and APTT were checked as well. Many children were hepatitis B positive and a few were hepatitis C positive. Ultrasound was done to see the paranchymal details of liver, spleen, cholelithiasis and choledocolithiasis. We did not see any stones in the gall bladder or CBD in our patients. Immunization with polyvalent pneumococcal vaccine and haemophilus influenzae was carried out two weeks in advance. Antibiotic prophylaxis was used at the time of surgery and one paediatric unit of blood was available in the theatre and another one was on standby in the recovery room.

Splenectomy: Splenectomy was performed through standard abdominal incisions. Chemoprophylaxis was carried out with the induction of anaesthesia and was continued for 24 hours. We used left transverse incisions. Accessory splenic tissue was identified in two of our patients and removed as well. Gall bladder was palpated for stones and it was confirmed that none of them have had stones. The left upper quadrant was not drained routinely. Following the surgery patients were managed in recovery room and next day straight forward cases were shifted to the Thalassaemia Center. `Follow-up was carried out in the OPD, both by Paeds and Surgical departments for 1 year.

Results

Table 1 Sex distribution

	n=	%age
Male	6	43
Female	8	57

Table 2: Age Distribution

Ages (years)	n=	%age
1-5	Nil	-
6-10	7	50
11-20	7	50
21-30	Nil	-

Table 3: Diagnosis

Diseases	n=	%age
Thalassaemia major	11	79
Thalassaemia Intermedia	3	21

Table 4: Morbidity

Diseases	n=	%age
Respiratory infection	6	43
Wound infection	3	21
Wound dehiscence	1	7
Stomach dilatation	2	14
Congestive failure	2	14
Bleeding from drain site	1	7
Thrombocytosis	02	14.28
Gastric fistuala	Nil	-
Pancreatitis	Nil	-
Venous thrombosis	Nil	-
Left hypochondrial abscess	Nil	-
Generalized abdominal abscess	Nil	-
Splenic abscess	Nil	-

Discussion

Splenectomy is rewardable procedure in thalassaemia patients as it prevents the destruction of red blood cells and improves anaemia in these patients. By doing so splenectomy also decrease blood transfusion requirements and the complications associated with blood transfusion[4]. Hypersplenism develops in thalassaemia patients very gradually or not at if all transfusion requirement is adequately met with as in high transfusion regimen. The spleen also does not get enlarge massively and danger of rupture is eliminated[7]. In low transfusion regimen in which transfusion are not given frequently and haemoglobin level remains 6 or 7 gm% (below 8.5gm%) extramedullary erythropoeisis is stimulated[3].

The constant stimulation of intra- medullary haemopoeisis leads to massive hepato spleno megaly. The splenic enlargment leave to increased trapping of blood cells. (because of increase volume) and further aggravates hypersplenism. Splenectomy cures the hypersplenism completely but it does not alter the basic pathology of thalassaemia. Fifty percent in our study were less than 10 years old. And the immune function of spleen is very active during that age group. Therefore prophylactic vaccination is mandatory and was performed in all patients.

Splenectomy improved the symptoms of patient by decreasing transfusion requirement, improving haemoglobin level and increased level of platelets and levcocytes. The mean age of the patients in our study is comparable to other studies[7]. Due to loss of immune function of the spleen, the patients are more susceptible to infection and this was seen in 6 our patients (43%). Culture and sensitivity revealed S.pneumoniae as a causative organism. Fatal post splenectomy haemophilus influenzae group B bacteraemia may occur in spite of pre-operative pneumococcal vaccination and penicillin prophylaxis after surgery. We in this small series with limited follow-up did not see any such fatal infection in our patients[1].

Patients with thalassaemia and other sickle-cell diseases are at a greater risk of developing splenic abscess but this phenomenon was negative in our study. The association of splenic abscess with thalassaemia[8].

Patients with thalassaemia are at a greater risk of developing pigmented stones due to chronic haemolysis. It was observed that 17% patients with HB-S beta thalassaemia had gall-stones though we did not see any stones in our study.

Thrombocytosis occurs immediately after splenectomy in most parties and is often the desired therapeutic result. Up to 75% of patients who have had splenectomy develop thrombocytosis [platelet count of greater than 400,000 per cu mm] and platelets counts in excess if 1,000,000 per cu. mm. develop in some patients. Generally this has not been associated with an increased risk of thromboembolism[3]. We have similar effect in all the patients.

Conclusion

Splenectomy has a definitive role in decreasing blood transfusions requirements in thalssaemia major patients. It also reduces iron overload in these patients. In thalassaemia intermedia the transfusion requirement was totally eliminated in our study. Slpenectomy also eliminates the danger of spontaneous rupture (a phenomenon still observed in thalassaemic patients treated by low transfusion regimens). If performed carefully in well-prepared patients in collaborations with pediatric department the rate of per and postoperative complications decreased and patients tolerates splenectomy very well in the term.

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