Splenectomy in thalassaemia major: experience at Madina Maternity and Children’s Hospital, Saudi Arabia

ZAKARIA MOH’D AL HAWSAWI, TARIG ISMAIL HUMMAIDA* & GHOUSIA AHMED ISMAIL

Departments of Paediatrics and *Paediatric Surgery, Madina Maternity & Children’s Hospital, Madina Al Munawara, Saudi Arabia

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Summary  Eighteen (32%) of 56 children with thalassaemia major, whose ages ranged from 5 to 12 years (mean 8.8), underwent splenectomy at Madina Maternity and Children’s Hospital, Saudi Arabia during the period January 1992 to December 1999. This retrospective study was undertaken with the aim of discovering the outcome. The indications for splenectomy were increased transfusion requirements and massive splenomegaly in 17 children and splenic abscess in one. Polyvalent pneumococcal and Haemophilus influenza vaccines were not available and all the children therefore received intramuscular benzathine penicillin prophylaxis prior to surgery and oral penicillin prophylactically afterwards. Post-splenectomy sepsicaemia did not occur. The mean transfusion requirement reduced from 2 to 4 weeks and the mean pre-transfusion haemoglobin rose from 6 to 9 g/dl. There were no deaths. We conclude that splenectomy can be performed safely in children over 5 years of age with thalassaemia, and that pre- and post-operative penicillin can be given prophylactically in the absence of the recommended vaccines.

Introduction

Thalassaemias are a group of genetic disorders caused by decreased biosynthesis of one or more of the globin chains of haemoglobin, presenting clinically as hypochromic microcytic anaemia. In \( \beta \)-thalassaemia, \( \beta \) globin synthesis is either reduced or totally absent. \( \beta \)-thalassaemia major, historically known as Cooley anaemia, is the homozygous form of this disease. It occurs at a gene frequency that varies between the different regions of Saudi Arabia.\(^1\) In Madina, the estimated gene frequency of \( \beta \)-thalassaemia is 0.1.\(^2\) Management of \( \beta \)-thalassaemia major includes hypertransfusion and splenectomy when patients have increasing transfusion requirements.\(^3\)

Madina Maternity & Children’s Hospital (MMCH) has 400 beds, 200 of which are paediatric, and it is the main referral hospital for the Madina region. The upper age limit for paediatric admissions is 13 years. The approximate number of children served by the hospital is 350,000 in an estimated population of 800,000. In 1992, the thalassaemia centre was established in the paediatric section to provide comprehensive management of children with thalassaemia, which includes regular blood transfusion at 3–4-week intervals, as indicated.

The objective of the study was to demon-
strate the outcome of splenectomy among the children with thalassaemia attending the centre.

Patients and methods
The case records of 56 children with \( \beta \)-thalassaemia major who were managed and followed up in the thalassaemia centre in MMCH between January 1992 and December 1999 were reviewed retrospectively. The diagnosis of \( \beta \)-thalassaemia major was based on a clinical history of pallor, jaundice and hepatosplenomegaly with haemoglobin electrophoresis showing high HbF values (95–98%) and raised HbA\(_2\) (2–5%) on a cellulose acetate medium at an alkaline pH of 8.4 (Helena Laboratories, 1530 Lindbergh Drive, Beaumont, Texas, USA). In addition, detection of \( \beta \)-thalassaemia minor in both parents was of diagnostic value. All patients were on a hypertransfusion regimen, receiving 10–15 ml/kg of packed RBCs at 3–4-week intervals with the aim of maintaining the mean pre-transfusion Hb at 9–10 g/dl. The main criterion for inclusion in the study was splenectomy in children with \( \beta \)-thalassaemia major who were then followed up in our centre for a minimum period of 2 years. Eighteen children fulfilled this criterion. The following variables were studied: age at splenectomy, sex, nationality, duration of follow-up, evidence of septicaemia and mean pre-transfusion Hb levels and leukocyte and platelet counts pre- and post-splenectomy. All these laboratory values were derived from many pre-transfusion complete blood counts.

Indications for splenectomy were increased transfusion requirements (>250 ml/kg/yr) and, in one instance, splenic abscess. The following procedure was followed: (i) all patients received 1.2 million units of IM benzathine penicillin 1 week before splenectomy; (ii) total splenectomy was performed by paediatric surgeons; (iii) on discharge, patients were given oral penicillin prophylaxis, 500 mg twice daily throughout the follow-up period; (iv) no patient received pneumococcal or Haemophilus influenzae vaccine pre- or post-splenectomy as they were not available at the time.

Results
Splenectomy was performed in 18 (32%) of 56 children with \( \beta \)-thalassaemia. Nine were Saudis and nine were non-Saudis, of whom seven were Pakistani, one Yemeni and one Syrian. There were ten boys and eight girls and their ages at splenectomy ranged from 5 to 12 years (mean 8.8). The majority of them (15, 83%) had splenectomy at between 5 and 10 years of age. Spleen sizes ranged from six to 12 cm below the costal margin. None of the children developed progressive hepatomegaly following splenectomy. The follow-up period ranged from 2 to 7 years (mean 2.7). The transfusion requirement post-splenectomy was reduced from 250–300 ml/kg/yr to 160–180 ml/kg/yr and the interval between transfusions increased from 2 to 4 weeks.

The mean pre-transfusion Hb level of 6 g/dl before splenectomy rose to 9 g/dl after splenectomy. The mean pre-transfusion platelet count after splenectomy ranged from 500,000/mm\(^3\) to 1,000,000/mm\(^3\). None of the children had thrombo-embolic complications, post-splenectomy septicaemia did not occur, and there were no deaths. Table I summarises the laboratory data of all the children before and after splenectomy.

Discussion
This is the first study in the Madina region to record the experience and outcome of splenectomy in children with \( \beta \)-thalassaemia major. It has long been known that splenectomy increases the risk of infection, particularly in younger patients,\(^{4,5}\) but it does reduce transfusion requirements\(^{6,7}\) and eliminates discomfort from mechanical pressure. Furthermore, a growth spurt might in some cases follow splenectomy.\(^{8}\)

Our study probably encompasses the largest number of splenectomies in children in Saudi Arabia with \( \beta \)-thalassaemia major. All were
TABLE I. Haematological data pre- and post-splenectomy (pre-transfusion mean values)

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Pre-splenectomy</th>
<th>Post-splenectomy</th>
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<tbody>
<tr>
<td></td>
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<td>Hb g/dl</td>
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homozygous $\beta$-thalassaemia, unlike the compound heterozygous haemoglobinopathies reported by Sumer et al. from the Eastern Province of Saudi Arabia. This probably reflects the fact that the Eastern Province has the highest prevalence and heterogeneity of haemoglobinopathies in the world. Over 80% of this population has one or more type of structural haemoglobinopathies or thalassaemia syndromes.

The prevalence of splenectomy among our children with thalassaemia is 32%, similar to other reports from Saudi Arabia. Some patients included in the study attended before the establishment of the thalassaemia centre and were not on the hypertransfusion regimen, and some showed poor compliance, resulting in significant splenomegaly and hypersplenism. There were no significant sex differences.

In 17 (94%) children, the indication for splenectomy was an increased requirement for transfusion, which is a more sensitive indicator of hypersplenism than thrombocytopenia or leucopenia. One of our patients had a splenic abscess, which is a rare but recognised complication of sickle cell disease, although to the best of our knowledge it has not previously been reported in thalassaemia. This child presented with fever, abdominal pain and tender splenomegaly, and ultrasonography and computerised tomography provided diagnostic imaging, as observed by Al-Salem et al.

Pneumococcal and *Haemophilus influenza* vaccines were not available but, despite this, none of our patients developed overwhelming post-splenectomy sepsis (OPSS) during 7 years of follow-up. Children under 5 years of age are at risk of OPSS after splenectomy but older children and even adults can also be at risk. Although overall transfusion requirements were reduced and mean pre-transfusion haemoglobin improved post-splenectomy, some of the patients (nos 3, 11 and 17) did not show significant differences (Table I). The reduction in blood transfusion requirements from every 2 weeks before splenectomy to every 4 weeks afterwards and the reduction of annual transfusion requirements by 60% are consistent with other reports. All the patients developed thrombocytosis ranging from 500,000 to 1,000,000/mm$^3$ but required no
intervention, as observed by Sumer et al.9 There were no deaths among our children.

Our findings show that splenectomy can safely be performed in thalassaemic children over 5 years of age and reduces transfusion requirements, thereby protecting from iron overload. Long-acting IM penicillin before splenectomy and oral penicillin prophylaxis post splenectomy can prevent septicemia. When the recommended polyvalent pneumococcal and Haemophilus influenza vaccines are not available, however, post-splenectomy septicemia is still considered a major risk in children under 5 years of age, and in children under 4 the risk of septicemia is twice as great. In the 1st year of life, the risk can be 21% or higher.17-20 Because of this risk, some authors have advocated partial splenectomy and splenic embolisation to preserve splenic function and avoid post-splenectomy complications.21,22

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References

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