Splenectomy in Infants and Children*

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The following report is concerned with the end results of splenectomy in infants and children, performed at the Childrens Hospital, Los Angeles, California, for a variety of indications. Recently the advisability of performing splenectomy in infancy,[2] and the necessity of "emergency splenectomy"[4] have been questioned and merit some discussion.

During the period 1932 to 1952, there were 72 splenectomies performed. Indications for surgery, together with results are listed in Table I.

CONGENITAL HEMOLYTIC ICTERUS

There were 37 cases with this diagnosis established. All but nine of these children had a history of similar disease in the family, and the spleen was usually large. The ages varied from four and one-half weeks to 14 years, and there were ten cases in patients under the age of seven months. The age distribution in this latter group of infants is as follows:

4½ weeks	1
7 weeks	1
8 weeks	1
10 weeks	1
14 weeks	1
4 months	1
6 months	2
7 months	2

This age distribution is of significance since it is felt that the establishment of the diagnosis of congenital hemolytic icterus is a clear-cut indication for splenectomy. Delay in operation may only mean the necessity of repeated transfusions, chronic ill health, anemia, and retarded development. In a recent report,[2] King and Shumacker suggest the possibility of an increased susceptibility to infection in infants in whom splenectomy is performed during the first few months of life. They report five cases in which splenectomy was performed for congenital hemolytic icterus in infants varying in age from two weeks to six months. Meningitis developed in four of these infants with one death, and an acute illness of unknown etiology with death in another. These complications occurred within a period of two and one-half months to three years postoperatively, and suggested the possibility of a cause and effect relationship between splenectomy in infancy and increased susceptibility to infection. The question of the necessity of splenic function to adequately combat infection, especially in infancy, is raised by these authors, together with a review of the literature, which indicates that the spleen may have the most functional activity in early life. Experimental work is reviewed, which indicates that in certain animal species splenectomy depresses resistance to infections and reduces the immunological response to antigen injection.

In this series of ten infants, splenectomy was accomplished in each case without difficulty, and a satisfactory clinical and hematological response occurred in all. There was one case of otitis media occurring on the fourth postoperative day, with a prompt response to sulfadiazine and a return to a normal temperature in 24 hours. Follow-up has been obtained in each of these patients

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Diagnosis	No.	Mortality	Complications	Results	
Congenital Hemolytic Icterus	37	0	Subarachnoid hemorrhage	Good	36
			Atelectasis	Poor	1
Thrombocytopenic Purpura	6	0	0	Good	6
Acquired Hemolytic Icterus	6	0	0	Good	2
				Fair	1
				Poor	3
"Banti's Syndrome"	4	1	Evisceration	Good	1
				Poor	3
Traumatic Rupture	10	1	0	Good	9
				Poor	1
Splenic Hematopenia	1	0	0	Poor	1
Splenic Neutropenia	1	1	Tracheobronchitis	Poor	1
Cooley's Anemia	2	0	0	Fair	1
				Poor	1
Congenital Hypoplastic Anemia	2	0	0	Good	1
				Fair	1
Gaucher's Disease	2	0	0	Fair	1
				Poor	1
Hepatosplenomegaly Unknown Cause	1	0	0	Good	1
Total	72	3	4		

for what is considered an adequate period. The youngest patient has been followed ten months, and the next three youngest have been followed nine, 13, and 12 years, respectively. The remainder have been seen for seven years in two instances, and for six years, four years, and three years, in the others. Careful evaluation has failed to disclose any significant illness, except for the otitis media noted previously, either in the immediate postoperative period or in the long term follow-up. All patients are in excellent health.

Since the spleen has been said to exert an inhibitory action on the hemopoietic function of the bone marrow, as evidenced by the regular occurrence of thrombocytosis and leucocytosis following splenectomy, and since a leucocytic response is a partial index of not only the severity of infection, but the organism's capacity to respond, it was felt a review of the ten infants' leucocytic response would be of interest to determine if it differed in any respect from the standard response in older age groups. In two cases an inadequate number of serial white cell counts was present to permit interpretation. In the remaining cases leucocytosis occurred in five patients, with an increase varying between 2,000 and 9,000, as compared to the preoperative level and with a peak at about the tenth day. In the two remaining cases, counts done on the fifth postoperative day revealed a leucocyte count 2,000 cells less than the preoperative level. A serial platelet count was made in only one case, and here the platelets increased from 256,000 to 1,050,000. In this small series it would appear that the hematological response of the infant to splenectomy, so far as leucocytosis and thrombocytosis are concerned, parallels the adult response.

It is believed that in this study there is no clinical evidence that splenectomy in infants is associated with any unusual risk, or with increased susceptibility to infection. Leucocytosis and thrombocytosis occur in most cases in a standard fashion in infants as in adults following splenectomy. Since results following splenectomy for congenital hemolytic icterus are universally good, it is felt that the earlier splenectomy is performed following the establishment of the diagnosis, the sooner patient rehabilitation will occur.

In the older ages there were 27 cases. These varied from 14 months of age to 14 years. Excellent clinical and hematological responses were obtained in 26 of these

cases. A poor result was obtained in one seven-year-old boy. This patient had an uneventful postoperative convalescence, but he had a poor subsequent clinical and hematological response. The anemia recurred with bouts of jaundice and episodes of fainting and convulsions requiring hospitalization four times. Six years after splenectomy, recurrent episodes of cramping upper abdominal pain occurred, associated with nausea and vomiting. The presence of gall stones had been established previously by cholecystogram, and the parents were advised that cholecystectomy should be performed. Operation was repeatedly refused, and three years later, after admission to another hospital for an acute episode of pain, rapid collapse and death occurred. Autopsy revealed a perforated colon ulcer, a part of an ulcerative colitis.

One serious postoperative complication occurred in a ten-year-old boy who developed a subarachnoid hemorrhage on the sixth postoperative day. Complete recovery followed, and the patient was released from the hospital on the 26th postoperative day.

There were three instances of gall stones found incidentally in the 37 cases, an incidence of 8 per cent. These patients were seven, 14, and 11 years of age at the time of splenectomy and simultaneous discovery of the calculi. The first patient has already been discussed relative to an eventual poor result. The second patient was operated upon ten months, and the third patient two years after splenectomy; cholecystectomy was performed.

ACQUIRED HEMOLYTIC ICTERUS

There were five cases of acquired hemolytic icterus. The age distribution was one and one-half years, five years, two of six years, and one of nine years. The five-year-old patient and the nine-year-old patient each had a good clinical and hematological response in a two and three year follow-up. One of the six year old patients had a fair result. Prior to splenectomy this patient had had 66 transfusions, and hemolysis had continued progressively. Since operation, the hemoglobin has remained at a low level of 48 per cent, but transfusion has not been done up to the present time, four years postoperatively. The second six-year-old patient had a poor result. Hemolysis continued after surgery, and there was a progressive downhill course, with death three months postoperatively.

In this small series, in only one case was the hemolytic process present as a complicating feature of a known primary disease. In the youngest child a diagnosis of Hodgkin's disease was established by lymph node biopsy three years after splenectomy. Hemorrhagic phenomena recurred, and the patient expired six years after operation. This latter case is illustrative of the difficulty in establishing the etiology in the acquired hemolytic group of cases, and also indicates the necessity of considering a primary disease such as Hodgkin's disease, leukemia, carcinomatosis, or even severe liver damage as being the underlying process in a hemolytic disease. This is especially true when one appreciates that spherocytosis and increased red cell fragility may be present in the acquired form of hemolytic anemia as well as in the congenital form.

The final case is discussed in some detail because Cortisone was used early in the course of therapy, yet hemolysis proceeded at a rapid rate. The patient, in critical condition, was subjected to emergency splenectomy which resulted in recovery.

A three-year-old patient was admitted to the hospital two and one-half weeks after the onset of an acute upper respiratory infection for which a sulfa drug had been prescribed, and four days after the onset of grossly dark urine and progressive weakness. Upon admission the patient was acutely ill. The liver was enlarged 3 cm. below the costal margin. The day preceding admission, blood study revealed R. B. C., 2,400,000; hemoglobin, 7.5 Gm.; W. B. C., 11,000; Coombs test, 2 plus. The urine was positive for occult blood, albumin 3 plus, 0-2 W. B. C., and 0-1 R. B. C. /h.p.f. Upon admission the hemoglobin was 6.9 Gm., and the R.B.C.'s num-

bered 2,221,000. Platelets were normal. The urine was dark red in color. A transfusion was started, and after 100 ml. had been given a severe chill and fever to 104° occurred. The pulse was 180 at this time. The transfusion was stopped. An initial dose of 25 mg. Cortisone was administered, and repeated in 8 hours. Hemolysis continued and 8 hours after admission the hemoglobin had dropped to 4.3 Gm., and the red cells numbered 1,290,000. Two doses of Cortisone had been given by this time. The patient's condition was considered critical, and with hemolysis continuing, the hematological and surgical consultants recommended splenectomy. The operation was performed rapidly and was without complications. Hemolysis continued for 12 hours, and then the urine rapidly cleared. Convalescence was uneventful. Cortisone was continued for 3 days. Three hundred ml. of blood was given during the operation after the pedicle was clamped, and 500 cc. of sedimented cells were given on the first postoperative day. At this time the hemoglobin was 12.0 Grams.

ACTH or Cortisone has been used in the therapy of acquired hemolytic icterus with success in producing a remission in the disease which, even if not sustained, can be reproduced later in preparation of a patient for splenectomy. Results have been favorable enough to warrant a recommendation that emergency splenectomy be a thing of the past[4] in a hemolytic crisis. However, it would seem that further study is indicated concerning this problem. It was felt by both the surgical and hematological staff that the last patient would have expired had splenectomy not been done as an emergency measure. Close hematological and surgical observation must prevail, and if a patient fails to respond to ACTH or Cortisone, splenectomy may be a life-saving procedure.

Thrombocytopenic Purpura:

There were 41 cases of thrombocytopenic purpura and five of these had splenectomy in the period studied. The results may be classified as follows:

Conservative Therapy	36
Spontaneous improvement	31
Deaths	5
Splenectomy	
Cured or improved	5

The mortality of 11 per cent in the patients not operated upon when compared to no deaths in the splenectomized patients does not represent an accurate comparison. Two of the patients who were not operated upon died on the day of admission, presumably from cerebral hemorrhage, although no autopsies were obtained. Another died after a prolonged course, and autopsy revealed an absent left kidney, double ureter, nephritis and pneumonia.

Two cases of death due to hemorrhage were in children under observation, in one instance for one month and in the other for two months, and serve to remind us that, although fatal hemorrhage is less common in childhood thrombocytopenia, it occurs nevertheless. The patient who continues to bleed can be supported for a limited time only by transfusion. Splenectomy may have to be considered in these patients even if conditions are not ideal, since bleeding may stop when the pedicle is ligated. Neither of the two deaths occurred during a period when ACTH or Cortisone was available, and it has now been demonstrated that these drugs can produce both clinical and hematological improvement in these cases with a rise in platelet count, reduction of bleeding,[3] and improvement in capillary resistance.[1] These beneficial results have been obtained either prior to splenectomy or in the splenectomized patient with recurrence of bleeding phenomena. The time of onset of clinical improvement following adrenal hormone therapy is an important factor, especially in an acutely bleeding patient, and is variously reported from "almost immediately" to up to four days. Although ACTH and Cortisone have proved to be of great value in the management of the thrombocytopenic patient, it would seem that in the actively bleeding patient who may die if the hemorrhage is not stopped, careful consideration must still be given to the possibility that a splenectomy may be a life-saving procedure.

The five children operated upon have all done well clinically. Two patients have had persistently low platelet counts since splenectomy, but have had greatly diminished hemorrhagic tendencies amounting now to only easy bruising. One of these patients is 15 years postoperative, and the other two and one-half years. The remaining patients have had normal platelet counts since operation and are well one year, four years, and 21 years since splenectomy.

"BANTI'S SYNDROME"

There were four cases with this diagnosis. Their ages were three weeks, one and one-half years, four years, five and one-half years, and six years. Results were poor in three of the cases in which splenectomy alone was performed, but a fourth case in which splenectomy was combined with a splenorenal shunt has had the only good result to date. The oldest child died after an evisceration on the eleventh postoperative day. The four-year-old patient died three months postoperatively with recurrent bleeding. The youngest child was lost to follow-up after three years, but had recurrent gastro-intestinal bleeding up to that period. These results are consistent with results of splenectomy in Banti's syndrome published by most authors, and emphasize that very little improvement can be expected in this disease by splenectomy with the exception of thrombosis of the splenic vein. In this instance splenectomy may be curative. Therapy directed to the relief of the increased portal tension and bleeding from collateral veins may prove of benefit in children to a degree comparable to that reported in adults.

There has been one splenorenal anastomosis performed at Children's Hospital. The patient was a five-year-old boy with gastro-intestinal bleeding and hepatosplenomegaly. Portal vein pressure at operation was 22 cm. of saline, and after the shunt splenic vein pressure was 5 cm. of saline. Convalescence was uneventful and there has been no bleeding for two years since operation.

MISCELLANEOUS TYPES

Gaucher's Disease. There were two patients with this disease. In each instance the spleen was very large and was removed for "mechanical" reasons. One 17-month-old female had a stormy postoperative course and remained well for only a period of seven months. She was readmitted with a pneumococcus meningitis, and expired one month later. The second patient, an eight-year-old boy, has remained well for a two year period.

Splenic Neutropenia. There was one case of this type, an acutely ill two-year-old boy with a severe pustular dermatitis, whose bone marrow showed almost complete absence of segmented neutrophilic granulocytes. Both the bone marrow and peripheral blood had an excellent response to ACTH, but there was a poor clinical response. A severe tracheobronchitis requiring tracheotomy followed splenectomy, and death occurred seven weeks postoperatively. Autopsy revealed multiple lung abscesses and bronchopneumonia.

Splenic Hematopenia. There was one case in a 3½ year old boy with dwarfism. There was an excellent clinical and hematological response to splenectomy, but 13 months postoperatively the patient was readmitted with an elevated temperature, and an acute upper respiratory infection, and expired.

Cooley's Anemia. One three-year-old child was benefited for a very short period, but has required transfusions about every two to three months since splenectomy, three years ago. A second eight-year-old patient has not been followed for a sufficently long period to evaluate.

Congenital Hypoplastic Anemia. A nine-year-old female with this diagnosis had had repeated transfusions. Following splenectomy she had three transfusions in a ten

month period postoperatively, and has maintained a hemoglobin of 75 per cent for the last five years, without therapy. A second eight-year-old girl has not been followed long enough to determine the long range result. The immediate postoperative hematological status has been good.

Splenic Trauma. There was one death in the ten children who had splenectomy for trauma to the spleen. This four-year-old child had associated cerebral injury and a laceration of the jejunum.

Hepatosplenomegaly of Unknown Etiology. A three-and-one-half-year-old female was operated upon in 1940 after prolonged observation of persistent anemia, splenomegaly, and adenopathy. Following splenectomy this patient has remained well to the present date. The value of long term follow-up is well illustrated by this case since the original pathological diagnosis, based upon examination of the spleen, a liver biopsy, and a mesenteric lymph node, was a reticuloendothelial lymphoblastoma. The passage of time has indicated the error of this diagnosis, and a review of the original sections, together with the obvious clinical status of the patient, has corrected the error without, however, making a specific diagnosis.

SUMMARY

1. The results of splenectomy performed on infants and children are reported.

2. Splenectomy for congenital hemolytic icterus is indicated upon the establishment of the diagnosis.

3. No evidence of increased susceptibility to infection nor increased mortality is present when splenectomy is performed in infancy.

4. Further study is indicated before "emergency splenectomy" is discarded as a therapeutic weapon in the treatment of the hemolytic crisis or the acutely bleeding thrombocytopenic patient.

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