SPLENECTOMY IN THALASSEMIA

Report on 12 Cases *

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The value of splenectomy as a therapeutic measure in thalassemia has been recently established for certain cases of this disease.[1,2] Previously its value had been questioned and a lot was said about severe infections occurring after splenectomy in children.

In recent years, however, several authors have reported on splenectomized cases of thalassemia [3,4] and it seems evident from these reports that splenectomy has a beneficial effect on longevity and also results in a decreased need of transfusions.

Thalassemia is fairly common in Lebanon as it is the most commonly encountered congenital form of anemia. Although there is no official report of case series from the Lebanon, the fact is that we observe more than 10 new patients per year in our American University hospital. One case with aplastic crisis in thalassemia was reported by the author.[5]

Diagnosis

The diagnosis of thalassemia is made when the following criteria are present in a case of hemolytic anemia and splenomegaly:

RBC. Anisocytosis, poikilocytosis - severe.

Hypochromia - marked.

High fetal hemoglobin.

Absence of abnormal hemoglobins as determined by electrophoresis.

Normal or high serum iron.

All the 12 cases met these criteria and in addition there was in most instances a positive family history and evidence of thalassemia minor in the parents. Transfusions were given at intervals when-

* Paper presented at the Second Middle Eastern - Mediterranean Pediatric Congress, Ankara, 1961.

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ever the level of hemoglobin fell to a value below 6 grams per 100 ml. The indication for splenectomy was not the same in all cases. In 9 cases the frequency of transfusions was increasing but in the other 3 instances marked splenomegaly and pancytopenia were the only indications for splenectomy which was performed in the hope of improving the general well - being of the patients. Table 1 shows the age of these patients and the period of observation after splenectomy.

TABLE 1

CASE	AGE, START OF ILLNESS	AGE AT SPLENEC-TOMY	FOLLOW-UP	AGE AT DEATH	SPLENIC WT.
1	24 months	6 years	13 years	19 years	
2	6½ years	11 years	4 years	15 years	
3	12 months	3 years	7 years		
4	24 months	5½ years	20 days	5 years, 7m.	
5	9 months	2 years, 1 m.	3 years		650 Gm.
6	2 months	6 years	2½ years		
7	5 months	2 years, 5 m.	1 year, 3 m.		
8	6 months	2 years, 4 m.	8 months		
9	2 months	2 years	8 months		710 Gm.
10	8 months	3½ years	3 months		
11	2 months	2 years, 10 m.	3 months		
12	1 year	6 years	3 months		

Infections after Splenectomy

During the follow - up of this series we have not observed any fulminating infections. The first two patients died as a result of cardiac complications (large heart, heart failure, severe arrhythmia). The fourth patient died 20 days after operation and the autopsy showed multiple thrombosis in the splenic vein, portal vein and mesenteric vessels. One patient (case 3) had pneumococcus meningi-

tis at the age of 9 years (6 years after splenectomy) which was treated with success. In general, splenectomy is not performed below 2 years of age because it is well known that fulminating infections are more common in the younger age group. [6]

Results after Splenectomy

In all our patients, except one who died 20 days after the operation, there was marked general improvement, an increase in weight, and in particular a decrease in the need for transfusions as shown in the following table:

TABLE 2

TRANSFUSIONS NEEDED BEFORE AND AFTER SPLENECTOMY

CASE	BEFORE cc./MONTH	AFTER cc./MONTH	REMARKS
1	500	125	
2	500	120	
3	50	None	Splenectomy mainly for large spleen.
4	200	—	Died 20 days after operation.
5	250	—	Splenectomy mainly for very large spleen and mild thrombocytopenia.
6	500	0	
7	500	160	
8	1000	0	
9	500	250	Bone marrow passed through a long period of aplasia.
10	500	0/3mos.	
11	750	100	
12	250	0	Splenectomy mainly for low retuculocyte count, leukopenia, thrombocytopenia.

The other important changes observed after splenectomy are those of the value obtained for reticulocyte counts, white blood

cells, (excluding the nucleated RBCs) and the platelet counts. These changes are shown in table 3.

Discussion

Unlike those with other hemolytic conditions, a good percentage of these patients showed rather low values for white blood cells, platelets and reticulocytes. All of these patients had very active and highly celluar marrows before splenectomy. It was shown that the bone marrow in thalassemia is already relatively ineffective [7,8] and it seems from the present series that the large spleen of these patients, in addition to hemolysis and destruction, exerts another hypersplenic effect on formed elements, depressing their appearance in the peripheral blood. It is noteworthy to add here that the spleens removed from our patients were very large, weighing between 650 and 800 grams.

Splenectomy in thalassemia is certainly absolutely indicated when a patient with active bone marrow needs more and more transfusions. The role of the spleen as a hemolyzing agent could well be proven by survival studies using Cr 51, a method that we did not employ. I find, however, that lowering of the values for formed elements, especially the platelets, would be another relative indication for splenectomy, provided the bone marrow is rich, cellular and *without evidence of megaloblastic erythropoiesis.* Clinical observation in this series has proved the beneficial effect of splenectomy under these conditions, even though the need for repeated transfusions was not great (case 5 - platelets, and case 12 - platelets, WBC, retic. See Tables 2 and 3).

Thus the indication for splenectomy in thalassemia would be as follows :

1. Greater need for transfusions.
2. Tendency to moderate thrombocytopenia, leukopenia and low reticulocytosis and active bone marrow.

Summary

Twelve cases of thalassemia are presented with hematological observations before and after splenectomy. The author thinks that it is possible to detect hypersplenism in thalassemia even before the increased need of repeated transfusions makes it more evident.

TABLE 3

RETICULOCYTES, WBC PLATELETS BEFORE AND AFTER SPLENECTOMY (*)

CASE	Reticulocytes before (%)	Reticulocytes after (%)	WBC(**) b.	WBC(**) a.	Platelets b.	Platelets a.
1	4	5	17,850	24,300		310,000
2	5.1	6.7	6,086	14,890	89,140	179,000
3	13.25	7.5	23,000	20,000	144,300	156,600
4	1.8	n.a	11,750	17,500	85,000	125,000
5	12.5	12.5	12,500	22,000	94,000	188,500
6	5.5	15	6,000	12,000	161,000	213,000
7	10.5	10.9	10,500	27,000	87,000	250,000
8	3.3	7.25	13,500	47,500	140,000	215,000
9	2.9	8.8	16,000	37,800	85,000	263,000
10	3.3	10	10,500	20,500	150,000	250,000
11	4.1	5.1	9,500	20,000	85,000	228,000
12	1.5	6	10,000	20,000	68,000	247,000

* All values are based on average of several examinations before and after splenectomy.

** Nucleated RBCs excluded.

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