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Prospect of Splenectomy in Thalassemia

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Thalassemia is one of the most severe genetic defects which presents as a major

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public health problem in the world population(1). Over 180 million people in the world and around 20 million in India carry the gene for (3-thalassemia(2). Every year about 1 lakh children are born in the world with thalassemia, 5000-7000 of whom are born in India(3) and it is quite likely that a large number of them die even before a diagnosis is made. Treatment of thalassemia consists of mainly blood transfusion, chelation therapy and bone marrow transplantation. With the recent introduction of bone marrow transplantation in the management of thalassemia, it is possible to have a cure for this disease and one can hope for a complete cure of the disease in the future by genetic engineering. Regular transfusion is still the only treatment used in the rural hospitals of our country. However, several life threatening complications like iron overloading, hepatitis, hypertension, convulsions, cardiac dysfunctions and renal failure may develop due to multiple transfusions(4-6). Splenectomy is often done to avoid these complications associated with repeated transfusions and to minimize the need and frequency of blood

transfusions(7). The present study has been undertaken to see the effects of early splenectomy on some serum glycolytic enzymes as well as on the requirement of transfusion.

Material and Methods

This study was conducted at the Departments of Pediatrics, Biochemistry and Anatomy of Burdwan Medical College in collaboration with R.G. Kar Medical College and B.C. Roy Hospital, Calcutta.

Twenty cases of p-thalassemia were selected for splenectomy while another twenty of a similar age and sex distribution were taken as the control group to see the effects of splenectomy in thalassemia patients. The indication for splenectomy in most cases was repeated blood transfusions at least once every month. Subjects between 2 to 6 years old were of either sex with an average age of 5 years. Thalassemia was diagnosed on the basis of their clinical profile and hemoglobin electrophoresis. Prior to splenectomy, complete hemogram estimations of RBC enzyme like phosphohexoisomerase, lactate dehydrogenase(9) and

aldolase(10), chest X-ray and an electrocardiogram were done. These investigations were repeated 12 weeks after splenectomy and the results were compared to see the effect of early splenectomy.

Results

Serum levels of phosphohexose isomerase, lactic dehydrogenase and aldolase of thalassemic patients and splenectomized thalassemic children are shown in *Table I*. It is evident from the results that the serum levels of these glycolytic enzymes are significantly lower in the splenectomized children compared to those of control thalassemic patients. The enzymatic activities in control thalassemic children are about 4 times higher than that of usual normal value while they show a general tendency to become normal due to splenectomy.

The histopathology of liver revealed deposition of iron and dilatation of the sinusoids but there was no evidence of fibrosis. The histopathology of spleen showed deposition of, iron and the architectural pattern was well maintained with no evidence of fibrosis.

TABLE I - Serum Levels of Phosphohexose Isomerase (PHI), Lactic Dehydrogenase (WH) and Aldolase of Thalassemic Patients with and without Splenectomy

Enzyme assayed	Enzyme activity	
	Splenectomized thalassemic patients	Thalassemic patients without splenectomy
pm (units/ml)	34.6±3.8	20.2±0.82*
LDH (units/ml)	752.4±16.2	266.6±5.62*
Aldolase (units/ml)	38.6±3.2	19.8±1.41*

Values are expressed as mean ± SE from 20 patients.

Values compared to corresponding controls by Students' t-test p values * <0.001.

Discussion

A transfusion requirement in excess of 180-200 ml/kg of packed cells per year is taken as an evidence of hypersplenism and is an indication for splenectomy(11). In the present series, the indication for splenectomy was also repeated transfusions, *i.e.*, one transfusion/month. Of the 20 cases under study, 16 had received prior immunization with pneumococcal and hepatitis-B vaccines. All the patients were on prophylactic penicillin therapy irrespective of their vaccination status.

Although the efficacy of prophylactic penicillin is not conclusive(12), we have routinely given it to all patients. During our 2-12 years follow up, none of the patients had severe septicemia or meningitis.

The RBC enzymes may be used as an index for blood transfusion and splenectomy in thalassemia(12). In the present study, more importance was given to the level of the RBC enzymes than the hemoglobin level to assess the degree of nemolysis. It is clear from the results that the enzyme levels are markedly reduced in splenectomized patients compared to those of controls. This reduced activity may be due to less hemolysis as these glycolytic enzymes are usually located in RBCs. Moreover, it was noted in clinical practice that the transfusion requirements fall from 180 ml/kg/year to 80 ml/kg/year. This observation along with the reduced glycolytic enzyme activity indicated that remarkable improvement was achieved with early splenectomy.

Histopathology of the liver did not show any cirrhotic changes as noted by various authors(13,14) who had done splenectomy quite late. In the present series, splenectomy was done quite early and there were no unwanted changes in the liver and the heart.

Pancreatic biopsy, was purposely avoided because of its complications, but serial blood sugar estimations did not reveal any evidence of diabetes mellitus.

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Parentral Ciprofloxacin in Persistent Diarrhea

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The role of empiric antibiotic therapy in hospitalized persistent diarrhea without identified pathogen is under investigation. At present use of a single systemically absorbed antibiotic may be justified(1).

Ciprofloxacin has a, broad spectrum bactericidal coverage and has been successfully used in multidrug resistant Gram negative infections in children(2-4). It has been also recommended in treatment of nosocomial Gram negative infections, which fail to respond to third generation cephalosporins(5). It was planned to study

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the effect of parenteral ciprofloxacin as a mono antibiotherapy in contrast to that of a combined antibiotherapy in hospitalized persistent diarrhea of infancy.

Material and Methods

Thirty children (0-1 yr) suffering from diarrhea for more than 14 days were enrolled. Fifteen children (Group A) were treated with parenteral ciprofloxacin (10 mg/kg/day) from the onset. The remaining 15 infants (Group B) were treated with parenteral ampicillin and chloramphenicol/ampicillin and amikacin (in neonates) in therapeutic dosage. Malnutrition and dehydration were treated with parenteral solutions (including plasma and/or blood)' as per standard practices of the Institute. Antibiotherapy was resorted to all the cases after drawing blood, stool, urine, CSF and other samples as appropriate for pathological and bacteriological examinations.

Results

Twenty one (70%) infants had malnutrition (below ICMR Grade II). Six (20%) cases had demonstrable radiological changes in the chest. Stool culture was negative in 18 (60%) cases probably due to previous antibiotherapy. *E. coli*, *Salmonella*, Gram positive cocci, proteus and other coliform organisms were present in the rest 12 infants (40%). They were all sensitive to ciprofloxacin and ampicillin *in vitro*. Blood culture was negative in all of them.