Original Research:

Study of Growth Profile of Children with βoThalassemia in ChildrenHaving Undergone Splenectomy

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Abstract:

A longitudinal study was conducted for 5 years to observe the growth profile of Thalassemia children who had to undergo splenectomy. Their growth profile was compared with a age and sex matched control group. It is seen that the growth profile was satisfactorily maintained in Thalassemic children with splenectomy. Hemoglobin level is also was well maintained and less frequent blood transfusion was required.

Key words- Growth profile, Thalessemic children with splenectomy.

Introduction:

Thalassemia is a single gene disorder1. First clinical entity was recognised by Dr. Thomas B. Cooley in the year 19252 amongst Italian children. Thalassemia was recognised first in West Bengal by Dr. M. Mukherjee3 in the year 1936. Whipple and Bradfort first used the term Thalassemia in 1932. The word was taken from Greek word meaning 'The sea'4. The magnitude of the problem of thalassemia is very high. Approximately 4% of the world population carry ? thalassemia gene. Various studies have revealed that 0-17 % of Indian population with an average of 3 % in the general population5 are affected.

Extensive work has been done in thalassemia in regard to early detection by haemoglobin electrophoresis and molecular studies. The growth profile of thalassemic children without splenectomy has been studied6,7. However, there has been no study on growth of thalassemic children with splenectomy. As it was observed that children with splenectomy required less blood transfusion than the children without splenectomy, the present study was undertaken. The usual indications for splenectomy were hypersplenism and huge splenomegaly causing mechanical discomfort.

Material and Methods:

The study was conducted in the Paediatric and Surgery Departments of Burdwan Medical College and Hospital Burdwan, West Bengal, India. Both indoor and outdoor cases were included.

The age of the patients were between 5 to 18 years, as splenectomy is not recommended below 5 years due to higher risk of infections. The study was confined to children belonging to district of Burdwan only.

Inclusion criteria (Selection of patients for splenectomy):

1. Patients who are unable to maintain the desired hemoglobin level as per age and sex.

2. Those that have irregular chelation or no chelation therapy due to financial problems.

3. Patients with features of hypersplenism.

4. Patients with mechanical discomfort due to huge splenomegaly.

Exclusion criteria:

Evidence of hemosiderosis in vital organs like liver, pancreas, heart.

Any congenital malformation.

HIV (Human Immunodeficiency Virus) positive cases.

100 children with ?o Thalassemia were included in the study, divided into 2 groups:

Group A: ?o Thalassemia children with splenectomy.

Group B: Normal control.

Age at which splenectomy was done are as follows:

20% children had splenectomy between 5 to 6 years of age, 40% had splenectomy between 6 to 7 years, 30% between 7 to 8 years and 10% between 8 to 10 years.

The growth profile was assessed in three scales 1) ICMR (Indian Council Of Medical Research) 2) NCHS (National Centre For Health Statistics), and 3) Affluent India (9).

It was very interesting to note that normal children were not comparable in their growth profile with the NCHS and Affluent India standards. Hence, the comparison was done with the ICMR standard.

The socio-economic status of the children is as per modified Kupuswami scale.

Investigations:

1. CBC: Haemoglobin level, total leucocyte count, differential count, ESR (Erythrocyte Sedimentation Rate), PCV (Packed cell volume), MCV (Mean corpuscular volume), MCH (Mean corpuscular haemoglobin), MCHC (Mean corpuscular haemoglobin concentration) and peripheral smear examination.

2. High Performance Liquid Chromatography (HPLC).

3. Serum sodium, potassium and calcium ions.

4. Hepatic enzymes (serum glutamate oxaloacetate transaminase, serum glutamate pyruvate transaminase)

5. Hepatitis B surface antigen

6. Blood urea and serum creatinine.

7. Septic screening and screening for Tuberculosis.

8. ECG (Electrocardiography)

9. Thyroid function tests.

10. Growth profile in standard method as per Indian Council of Medical Research (ICMR) and National Centre for Health Statistics (NCHS) recommendations.

Result:

Table No.1 Comparison of anthropometry in two groups

	ICMR STANDARD													NCHS STANDARD								
	weigh	Group A		Group A				Group B		Group B				Group A		Group A			Group B		Group B	
		Beginning		End				Beginning		End				Beginning		End			Beginning		End	
Age (years)	No.	Above mean	Below mean	Above mean	Below mean	Age (years)	No.	Above mean	Below mean	Above mean	Below mean	No.	Above mean	Below mean	Above mean	Below mean	No.	Above mean	Below mean	Above mean	Below mean
5-9	15	10 (66.6%)	5 (33.3%)	14 (93.3%)	1 (6.6%)	5-9	18	18 (100)	0	18 (100%)	0										
>9-12	17	16 (94%)	1 (6%)	13 (76.4%)	4 (23.6%)	>9-12	15	15 (100%)	0	15 (100%)	0										
>12-18	18	11 (61.1%)	7 (38.8%)	9 (50%)	9 (50%)	>12-18	17	13 (76.4)	4 (23.5%)	16 (87.5%)	1 (12.5%)										
Total	50	37 (74%)	13 (26%)	36 (72%)	14 (28%)	Total	50	46 (92%)	4 (08%)	49 (98%)	1 (2%)										

	ICMR STANDARD													NCHS STANDARD							
	weigh	Group A		Group A				Group B		Group B			Group A		Group A			Group B		Group B	
		Beginning		End				Beginning		End			Beginning		End			Beginning		End	
Age (years)	No.	Above mean	Below mean	Above mean	Below mean	Age (years)	No.	Above mean	Below mean	Above mean	Below mean	No.	Above mean	Below mean	Above mean	Below mean	No.	Above mean	Below mean	Above mean	Below mean
5-9	15	10 (66.6%)	5 (33.3%)	5 (33.3%)	10 (66.6%)	5-9	18	18 (100%)	0	18 (100%)	0										
>9-12	17	12 (70.5%)	5 (29.4%)	7 (41%)	10 (59%)	>9-12	15	15 (100%)	0	15 (100%)	0										
>12-18	18	12 (66.6%)	6 (33.3%)	8 (44.4%)	10 (55.5%)	>12-18	17	17 (100%)	0	17 (100%)	0										
Total	50	34 (68%)	16 (32%)	20 (40%)	30 (60%)	Total	50	50 (100%)	0	50 (100%)	0										
5-9>	18			18 (100%)	0		15			7 (46.6%)	8 (53.3%)										
9-12	15			12 (80%)	3 (20%)		17			6 (35.2%)	11 (64.7%)										

	ICMR STANDARD													NCHS STANDARD							
	weigh	Group A		Group A				Group B		Group B			Group A		Group A			Group B		Group B	
		Beginning		End				Beginning		End			Beginning		End			Beginning		End	
Age (years)	No.	Above mean	Below mean	Above mean	Below mean	Age (years)	No.	Above mean	Below mean	Above mean	Below mean	No.	Above mean	Below mean	Above mean	Below mean	No.	Above mean	Below mean	Above mean	Below mean
>12-18	17			15 (88.2%)	2 (11.7%)		18			7 (38.8%)	11 (61.1%)										
Total	50			45 (90%)	5 (10%)		50			20 (40%)	30 (60%)										
5-9	18			18 (100%)	0		15			14 (93.3%)	1 (6.6%)										
>9-12	15			14 (93.3%)	1 (6.6%)		17			4 (23.52%)	13 (76.4%)										
>12-18	17			7 (41.1%)	10 (58.8%)		18			2 (11.1%)	16 (88.8%)										
Total	50			39 (78%)	11 (22%)		50			20 (40%)	30 (60%)										
5-9	18			1 (5.5%)	17 (94.4%)		15			0	15 (100%)										
>9-12	15			0	15 (100%)		17			0	17 (100%)										
>12-18	17			1 (5.8%)	16 (94.1%)		18			0	18 (100%)										
Total	50			2 (4%)	48 (96%)		50			0	50 (100%)										

Sexual growth as per Tanner criteria in boys and girls [8]:

Sexual Maturation Rating (SMR) in Thalassemia with splenectomy is studied only in the age group of 12-18 years who are in the pubertal age. Out of 18 thalassemia children with spenectomy there were 7 males and 11 females. Out of the 7 males, 6 had SMR score 4 and 1 had SMR score 3. Whereas among female children, 7 had SMR score 4 and 4 females had SMR score of 3. In the control group there were 17 children out of whom there were 9 males and 8 females. Out of the 9 males, 8 had SMR score of 4 and 1 had score 3. In the female control group, 6 had SMR 4 and 2 had SMR 2. In thalassemic female children SMR is low as the female child gets less care than the male. Statistical analysis of different growth parameters in ?o thalassemic children with splenectomy and normal child:

	Weight		Height		Chest circumference	
Age(years)	X2	P value	X2	P value	X2	P value
5-9	1.25	<0.01	7.13	<0.01	24.06	<0.001
>9-12	4.18	<0.02	5.42	<0.02	19.69	<0.001
>12-18	24.48	<0.001	6.89	<0.02	4.26	<0.005

The statistical analysis shows that all the parameters of growth profile are statistically significant. It clearly indicates that growth profile of thalassemic children cannot be maintained only by giving blood. They need more nutrition than normal children as their haemoglobin is not more than 10 gm/dl. Though blood transfusion and chelation therapy are main treatment but to maintain the growth profile, dietetic supply is essential.

Discussion:

This study was conducted on two groups of children. Group A consists of children with ?o Thalassemia who had splenectomy and is on blood transfusion along with or without chelation therapy. Group B consists of the normal control group.

As per the incidence of the occurrence of thalassemia of West Bengal scenario, total 1000 patients are expected to be born every year. In Burdwan district it should be 86. This disease needs a continuous treatment including blood transfusion, chelation therapy and a continuous assessment of their growth.

Blood transfusion is the mainstay in the treatment of thalassemia, but the number of blood banks in West Bengal is only 103 of which 29 are in the private sector. This part is to be improved in both the government and non-government levels. Total blood collection at the government and non-government levels has increased proportionately from the year 2004 to the year 2007 at both the government and non-government levels. However there is decline in replacement of blood by the family members of the patients. Therefore the family members need more motivation for blood donation as otherwise there will be lack of blood and the thalassemic children will be affected as the government has decided to give blood to thalassemic children without donation by the family members. It is very difficult for the family members of the thalassemic children to arrange 8 to 10 transfusions per year.

The study shows that as per ICMR standards, all the values with regards to weight were 100% at the beginning and the end of study in control group. In case of height it was 96% at the beginning and end of study.

In group A that is thalassemic children with

splenectomy, those in the age group of 5 to 9 years 14 children out of 15 maintained normal growths at the begining and the end of study.

In the age group 9 to 12 years, 13 children out of 17 maintained normal growth.

In the age group 12 to 18 years, 9 out of 18 children maintained normal growth.

Thus 72% of the children in group A are seen to maintain normal growth in respect of weight for age at the end of the study.

Similarly, in respect of height for age, 40% of the children with splenectomy have heights above mean in comparison to normal children who have heights above mean in 100% cases.

In respect of head circumference 40% of thalassemic subjects with splenectomy have levels above mean and 90% of normal children have head circumference above mean.

In respect of chest circumference only 40% thalassemic children have chest circumference above mean and 78% of the thalassemic children have chest circumference above mean.

In respect of Mid arm circumference (MAC) only 4% normal children have MAC above mean and none of the thalassemic children have MAC above normal.

The incidence of Thalassemia in the district of Burdwan is 86 per year. The study sample was taken randomly. It is a longitudinal study. The normal study population was selected belonging to the same geographic region and similar linear follow-up was conducted with the study group. The ICMR standard has been used for the assessment of growth profile, excepting Mid Arm Circumference for which the Agarwal standard has been used.

The study was conducted on patients admitted and operated at Guru Gobind Singh Hospital, Jamnagar(10) in various surgical units between June 2007 and March 2009. This is a study of 50 cases operated on elective basis. The main indication for splenectomy was higher blood transfusion requirement. After splenectomy, a dramatic increase in the Hb level occurred. All the patients had Hb >8g% with an average of 10g%. Most of the patients had a decrease in blood transfusion requirement with 96% of patients having blood transfusion requirements <150ml/kg/year. The frequency of blood transfusion requirement also decreases. Quality of life improved after splenectomy because of improved hemoglobin. The patients felt less fatigue and were able to carry out their daily activity in a better way. Decreasing blood transfusion requirement helped them reduce the hospital visit. The cost of iron chelation therapy was also reduced. Most patients had improved school attendance and better academic performance. Their family members also benefited due to decreased hospitalization. Overall, quality of life of the entire family improved and thus it proved to be a boon for the patient and his family.

Conclusion:

It was seen that the growth profile is satisfactorily maintained in Thalassemic children with splenectomy. Hemoglobin level is also well maintained and less frequent blood transfusion is required. Chelation therapy should be given to all such children. Early splenectomy may be considered when the child attained the age to combat the infections, besides regular blood transfusion and chelation therapy in resource scarce settings.

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