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GROWTH FAILURE IN BETA THALASSEMIA MAJOR PATIENT ADMITTED IN TERTIARY CARE CENTRE

Medicine

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ABSTRACT

Introduction: Thalassemia is one of the most common genetically transmitted diseases in the world and is associated with reduced synthesis of structurally normal haemoglobin. Growth failure in thalassemia is one of the common complications . Many factors like iron overload, irregular blood transfusion, iron chelating agent toxicity, socioeconomic factors, nutritional deficiency, hormonal deficiency will lead to growth failure. **Aims and Objectives:** To estimate the prevalence of short stature in the paediatric patients of β-thalassemia major. **Materials and Methods:** 98 β-thalassemia major patients between 1 to 12 years of age, visiting the centre for blood transfusions with or without iron chelation therapy were enrolled in this observational study. The data was analysed using WHO growth standards. **Results:** 43.88% of the patients were of short stature. **Conclusion:** Physical growth parameters such as height, weight, and BMI are reduced in children with beta thalassemia

KEYWORDS

INTRODUCTION

The inherited haemoglobin disorders are the most common single gene defect in man. The frequency of the carrier state has been estimated to be 270/million with about 400,000 annual births a year of infants with serious haemoglobinopathies worldwide.This is of special importance in developing countries, where it increases the burden of 1 health care delivery systems .[1]

There are two main types, alpha and beta Thalassemia. The severity of alpha and beta thalassemia depends on how many of four genes for alpha or two genes for beta globin are missing. [2]

B-Thalassemia major is the most severe form of the thalassemia group, that usually presents within the first two years of life with severe anaemia, requiring regular red blood cell transfusions[3]. The untreated or poorly transfused individuals with β-thalassemia major develop complications like growth retardation, jaundice, poor musculature, hepatosplenomegaly, skeletal changes etc.[4]

Growth failure in thalassemia is one of the common complications. Many factors like iron overload, iron chelating agent toxicity, socioeconomic factors, nutritional deficiencies, hormonal deficiencies will lead to growth failure[5]. Regular blood transfusions, regular iron chelators use in proper dose and other therapeutic facilities are important factors involved in the growth and development of these patients. Providing appropriate therapeutic methods and education for these patients and their families, and also the importance of regular measurement of their physical growth parameters as an index to evaluate the therapeutic effectiveness.

Growth failure leads to short stature which is defined as height below 3rd centile or 2 standard deviations less than the mean for the age, sex and the population. Height for age below -2 SD means moderate to severe stunting whereas that below -3 SD means severe stunting.[6]

We conducted this study to estimate the prevalence of short stature in β-thalassemia major patients, in the age group of 1 to 12 years, receiving blood transfusions with or without iron chelation therapy in the tertiary care centre.

MATERIALAND METHODS

An observational study was conducted in a tertiary care centre in Patna, involving patients admitted for a blood transfusion. A total of 98 patients, 34 girls and 64 boys, with β- thalassemia major, in the age group of 1 to 12 years, receiving blood transfusions with or without iron chelation therapy were enrolled in the study. Their anthropometric records were studied and the indices: height-for-age, weight-for-age, weight-for-height, and thebody mass index were calculated. The data were analysed using the WHO standard growth charts. Chi-square test was used for the statistical analysis.

Inclusion Criteria

Children between the age of 1-12 years diagnosed as beta thalassemia major based on HPLC without splenectomy.

Exclusion Criteria

- Patients > 1 Year and < 12 years
- Patients of other types of thalassemia sydromes.
- Patients diagnosed with other causes of anaemia.
- Patients with inconclusive HPLC results.

RESULTS

The study involved 98 β-thalassemia major patients, 34 females & 64 males in the age-group of 1 to 12 years. The mean age of all the patients was 4.95 years; that of females was 5.11 years and that of males was 4.79 years. The mean weight of all the patients was 16.3 kg; that of females was 16.93 kg and of males was 15.68 kg. The mean height of all the patients was 98.2 cm; that of females was 99.41 cm and of males was 97.39 cm. The mean BMI of all the patients was 15.94; that of females was 16.16 and of males was 15.72. There was no significant difference in the mean age, weight, height and BMI between females and males (Table 1).

In this study, we found that 43.88% of the total patients were short stature. Also, 44.12% of females and 43.75% of males were of short stature. There was no significant difference between females and males (Table 1). We observed that 23.47% of the total patients were moderately stunted whereas 20.41% were severely stunted. In females, 23.53% were moderately stunted and 20.59% were severely stunted. In males23.43% showed moderate stunting whereas 20.31% showed severe stunting. There was no significant difference between females and males

For further analysis, the patients were divided into two groups: short stature and normal stature (Table 2). The proportion of normal stature patients (55.1%, N=98) was significantly higher than that of short stature patients (43.88%, N=98) ($p < 0.0001$). The mean age of short stature patients (5.44 years) was significantly higher than that of normal stature patients (4.36 years) . The mean weight of short stature patients (13.95 kg) was less than that of normal stature patients (18.15 kg). The mean BMI of short stature patients (16.55 kg/m^2) was slightly more than that of normal stature patients (15.47 kg/m^2).The number of underweight children amongst short stature patients (48.83%,N=43) was significantly higher than that amongst normal stature patients (21.81%, N=55) ($p < 0.0001$).Out of 98 patients, only 48 patients (48.98%) were on iron chelation therapy (Table 4). Only 15 female patients (44.12%) and 33(51.56%) male patients were taking iron chelators. When investigated, serum ferritin was >1000 in 23 (67.64%) female patients and 47(69.1%) male patients(Table 3)

The patients were divided into 3 groups according to the frequency of blood transfusion (Table 5). Out of the total 98 patients, 35 patients

were transfused every 3 weeks regularly. Most patients (49) visited the centre for transfusion every 3-6 weeks. The rest 14 patients visited for transfusion after more than 6 weeks.

Table1: : Comparison Of Anthropometric Data Between Female And Male Patients With B-thalassemia Major

	Total Patients	Total Female	Total Male	P Value
	N=98	N=34	N=64	
Mean Age(Years)	4.95	5.11	4.79	0.49
Mean Weight(Kg)	16.302	16.928	15.676	0.63
Mean Height(Cm)	98.402	99.412	97.391	0.41
Mean Bmi(Kg/M2)	15.941	16.163	15.718	0.76
Prevalence Of Short Stature	43.88% (43)	44.12% (15)	43.75% (28)	0.06
Prevalence Of Moderate Short Stature	23.47% (23)	23.53% (8)	23.43% (15)	0.95
Prevalence Of Severe Short Stature	20.41% (20)	20.59% (7)	20.31% (13)	0.86

Table 2: Comparison Of Anthropometric Data Between Short Stature And Normal Stature Patients With B-thalassemia Major

	Short stature	Normal stature	P-value
No.of patients	43(43.88%)	55(56.12%)	P<0.0001
Mean age(years)	4.36	5.44	0.68
Mean weight(kg)	13.95	18.15	1.43
Mean height(cm)	89.77	105.18	0.11
Mean BMI (kg/m2)	16.55	15.47	0.19
Underweight(%)	48.83%	21.81%	<0.0001

Table 3:status Of Serum Ferritin

SERUM FERRITIN	FEMALE	MALE
<1000	11(32.35%)	17(25%)
>1000	23(67.64%)	47(69.1%)

Table 4:status Of Chelation Therapy

	Total	Females	Males
Taking Chelators	48(48.98%)	15(44.12%)	33(51.56%)
Not Taking Chelators	50(51.02%)	19(55.88%)	31(48.43%)

Table 5:frequency Of Blood Transfusion

	Total	Females	Males
<3 Weeks	35(35.7%)	10(29.4%)	25(39.06%)
3 Weeks-6 Weeks	49(50%)	18(52.94%)	31(48.4%)
>6 Weeks	14(14.29%)	6(17.6%)	8(12.5%)

CONCLUSION:

Physical growth parameters such as height, weight, and BMI are reduced in children with beta thalassemia Physical growth in thalassemia is dependent on multiple factors including endocrinopathies, iron overload, improper iron chelator therapy and its toxicity, nutritional deficiencies, irregular blood transfusion leading to chronic anaemic states. Endocrinal function progressively declines with age due to iron deposition in thepituitary and endocrine organs. In conclusion, the prevalence of short stature in β- thalassemia major is 43.88%.

From our study, we strongly suggest a stringent monitoring of the growth velocity of β- thalassemia major patients for an early detection and early intervention.

Due to technical limitations, a thorough evaluation of the causes of growth failure in these patients was not possible, however; this will be covered in our future studies.

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