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PREVALENCE OF SHORT STATURE IN PEDIATRIC β -THALASSEMIA MAJOR PATIENTS ADMITTED TO A TERTIARY CARE CENTER

Pediatrics

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ABSTRACT

Background: Growth failure is an important complication of β-Thalassemia major in pediatric patients.
Aims and objectives: To estimate the prevalence of short stature in the pediatric patients of β-thalassemia major.
Materials and methods: 122 β-thalassemia major patients between 1 to 12 years of age, receiving regular blood transfusions with or without iron chelation therapy were enrolled in this record based study. The data was analyzed using WHO growth standards.
Results: 34.43% of the patients were of short stature. 59.52% of the short stature patients were underweight. The maximum number of short stature patients was seen in the age group of 11 to 12 years.
Conclusion: The prevalence of short stature in β-thalassemia major is 34.43%. Patients in the age group of 11 to 12 years seem to be most affected. Malnutrition is more prevalent in short stature than in normal stature thalassemia patients.

KEYWORDS

Growth failure, Prevalence, Short stature, Thalassemia

INTRODUCTION

Thalassemia is a group of autosomal recessive hemoglobinopathies characterized by chronic hemolytic, microcytic, hypochromic anaemia with ineffective erythropoiesis and extra-medullary hematopoiesis due to defective production of haemoglobin in the bone marrow.(1) Thalassemia is classified as α or β depending on the defective synthesis of α or β globin chains respectively. β-Thalassemia can present in three forms: major, intermedia and minor.(2) β-Thalassemia major is the most severe form that usually presents within the first two years of life with severe anaemia, requiring regular red blood cell transfusions. The untreated or poorly transfused individuals with β-thalassemia major develop complications like growth retardation, jaundice, poor musculature, hepatosplenomegaly, skeletal changes etc.(3)

Growth failure is a well-recognized complication of thalassemia and has persisted over the years despite major therapeutic advances. The cause of growth failure is attributed to multiple factors such as chronic hypoxia, anaemia, ineffective erythropoiesis, malnutrition, iron overload-induced endocrinopathy, delayed or arrested puberty, lack of growth spurt etc.(4)

Growth failure leads to a short stature which is defined as height below the 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.(5)

Short statures possess a number of physical as well as psychological problems in childhood. Short children have lower social competence and show more social problems than children with normal stature. This simply adds to the morbidity of the disease.(6)

Many studies have previously evaluated the nutritional and the constitutional aspects of growth failure in Indian population. However, few studies have been done on growth failure due to chronic diseases in children. Hence, 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 regular blood transfusions with or without iron chelation therapy, in Sassoon General Hospital, a tertiary care teaching hospital, in Maharashtra, India and also to study the association of malnutrition with growth failure in these patients.

MATERIAL & METHOD

A record-based retrospective study was conducted in Sassoon General Hospital involving patients admitted for a blood transfusion from 1st of January 2018 to 1st of April 2018. The required sample size was calculated to be 119. A total of 122 patients, 55 girls and 67 boys, with β-thalassemia major, in the age group of 1 to 12 years, receiving regular 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 the body mass index were calculated. The data were analyzed using the WHO standard growth charts. Patients were divided into 6 age groups and the prevalence and the severity of short stature in each age group were assessed. Chi-square test was used for the statistical analysis.

RESULTS

The study involved 122 β-thalassemia major patients, 55 females & 67 males in the age-group of 1 to 12 years. The mean age of all the patients was 6.56±0.27 years; that of females was 6.52 ± 0.40 years and that of males was 6.51 ± 0.36 years. The mean weight of all the patients was 17.90±0.50 kg; that of females was 17.55 ± 0.64 kg and of males was 18.19 ± 0.75 kg. The mean height of all the patients was 108.21±1.48 cm; that of females was 107.6 ± 2.02 cm and of males was 108.7 ± 2.13 cm. The mean BMI of all the patients was 14.95±0.14; that of females was 14.91±0.17 and of males was 14.98±0.22. 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 34.43% (N=122) of the total patients were short stature. Also, 32.73% (N=55) of females and 35.82% (N=67) of males were of short stature. There was no significant difference between females and males (table 1).

We observed that 47.54% (N=122) of the total patients were moderately stunted whereas 21.31% (N=122) were severely stunted. In females, 47.27% (N=55) were moderately stunted and 18.18% (N=55) were severely stunted. In males, 47.76% (N=67) showed moderate stunting whereas 23.88% (N=67) showed severe stunting. There was no significant difference between females and males (table 1).

The highest number of short stature cases was seen in the age-group of 11 to 12 years, both in females and males. The number of severe short stature cases was also highest in the age-group of 11 to 12 years, however; in males where it was seen in the age-group of 1 to 3 years (table 1).

For further analysis, the patients were divided into two groups: short stature and normal stature. The proportion of normal stature patients (65.57%, N=122) was significantly higher than that of short stature patients (34.43%, N=122) ($p < 0.0001$). The mean age of short stature patients (7.29 years) was significantly higher than that of normal stature patients (6.11 years) (p=0.0335). The mean weight of short stature patients (17.57 kg) was less than that of normal stature patients (18.07 kg). Also, the mean BMI of short stature patients (15.16 kg/m^2) was less than that of normal stature patients (14.75 kg/m^2).

The number of underweight children amongst short stature patients (59.52%, N=42) was significantly higher than that amongst normal stature patients (15%, N=80) ($p < 0.0001$). It was seen that acute malnutrition was more common in short stature patients (14.29%, N=42) than that in normal stature patients (7.5%, N=80).

DISCUSSION

This study has shown that the prevalence of short stature in pediatric β-thalassemia major patients is 34.43%. A study done by De Sanctis V et al. compared the prevalence data from four different countries, all of them showing comparable results that support our finding.(7) We found that there is no significant difference in the prevalence (p=0.7219) and severity (p=0.4461 for severe short stature, p=0.9572 for moderate short stature) of short stature between females and males.

We found that the highest prevalence of short stature was in the age-group of 11 to 12 years. Many studies have emphasized on the iron-overload induced endocrinopathy as the main cause of growth failure in β-thalassemia major that usually sets in around 10 years of age.(8) A study by N. Skordis et al. states that the growth failure in thalassemia occurs in three phases. The growth is usually normal till 9 to 10 years of age, followed by a steady decline in growth velocity and a delay or absence of pubertal growth spurt.(4) However, in this study, we found that the mean age of patients with short stature was 7.29 years. Also, 57.14% (N=42) of short stature patients were under 9 years of age. This could be explained by factors other than iron-overload induced endocrinopathy such as chronic anaemia, hypoxia, osteopathy, malnutrition etc. that affect the growth at an early age.(9)

We also observed that malnutrition was more common in patients with short stature than in patients with normal stature. The prevalence of underweight in short stature patients (59.52%, N=42) was significantly higher than that in normal stature patients (15%, N=80) ($p < 0.0001$). Also, acute malnutrition was more prevalent in short stature patients than in normal stature patients. The prevalence of SAM was equal to that of MAM in both normal and short stature patients. Although the mean weight of short stature patients was smaller than that of normal stature patients, the mean BMI was higher due to the short stature. Our findings are supported by the study done by Kumari Vasundhara which showed that almost 70% of pediatric patients with β-thalassemia major had grade 2 and 3 malnutrition and most of the children had weight-for-age and height-for-age <50th percentile of which majority were below the 3rd percentile.(10)

In conclusion, the prevalence of short stature in β-thalassemia major is 34.43%. The patients in the age group of 11 to 12 years seem to be most affected. There is no significant difference in the prevalence and severity of short stature between females and males. Malnutrition is more common in short stature patients than in normal stature patients.

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.

TABLE 1: Comparison of anthropometric data between female and male patients with β-thalassemia major

	Total patients	Females	Males	P value
	(N=122)	(N=55)	(N=67)	
Mean age (years)	6.56±0.27	6.52±0.40	6.51±0.36	0.9952
Mean weight (kg)	17.90±0.50	17.55±0.64	18.19±0.75	0.5279
Mean height (cm)	108.21±1.48	107.6±2.02	108.7±2.13	0.7197
Mean BMI (kg/m²)	14.95±0.14	14.91±0.17	14.98±0.22	0.4237
Prevalence of short stature	34.43%	32.73%	35.82%	0.7219
Prevalence of severe short stature	21.31%	18.18%	23.88	0.4461
Prevalence of moderate short stature	47.54%	47.27%	47.76%	0.9572
Prevalence of underweight	30.23%	34.54%	26.86%	0.3605
Prevalence of stunting without wasting	29.51%	29.09%	29.85%	0.9273
Prevalence of wasting without stunting	4.92%	9.09%	1.49%	0.0544
Prevalence of stunting with wasting	4.92%	3.63%	5.97%	0.5536
Age-group with highest prevalence of short stature	11 to 12 years	11 to 12 years	11 to 12 years	
Age-group with the lowest prevalence of short stature	5 to 7 years	5 to 7 years	7 to 9 years	
Age-group with highest prevalence of severe short stature	11 to 12 years	11 to 12 years	1 to 3 years	
Age-group with lowest prevalence of severe short stature	7 to 9 years	1 to 9 years	7 to 9 years	

TABLE 2: Comparison of anthropometric data between short stature and normal stature patients with β-thalassemia major

	Short Stature	Normal stature	P value
Number of patients	42 (34.43%, N=122)	80 (65.57%, N=122)	$p < 0.0001$
Mean age (years)	7.29	6.11	0.0335
Mean height (cm)	106.58	109.06	0.427
Mean weight (kg)	17.57	18.07	0.6402
Mean BMI (kg/m²)	15.16	14.75	0.1527
Underweight%	59.52	15	$p < 0.0001$
Acute malnutrition%	14.29	7.5	0.2334
SAM%	7.14	3.75	0.4126
MAM%	7.14	3.75	0.4126

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