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Study of Growth Parameters in Patients of Thalassemia In Rajendra Institute of Medical Sciences,Ranchi

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I. Introduction

The β thalassemias are a heterogeneneous group of inherited disorders of haemoglobin (Hb) synthesis characterized by a reduction(β+) or absence of (β0) synthesis of the β globin chains of Hb, resulting in an imbalanced chain synthesis.[1]

As per WHO estimates 4-5% of the world's population are carriers of hemoglobinopathies. World wide 15 million people have clinically apparent thalassemic disorders. There are about 240 million carriers of β thalassemia world wide and in India alone ,the number is approximately 30 million with a mean prevalence of 3.3.Every year approximately 100,000 children with β thalassemia major are born world over of which 10,000 are born in India. In our country with around 9,000 to 10,000 cases being added every year. The carrier rate for β thalassemia major gene varies from 1 to 3 percentage in Southern India to 3to 15 percentage in Northern India.[2]

Anaemias results when rate of destruction exceeds the capacity of marrow to produce RBCs . During haemolysis RBC survival is shortened, RBC count falls, erythropoietin is increased and marrow activity results in increased RBC production reflected by increased percentage of reticulocytes in blood (Nelson 20th edition).

Management of thalassemia is by repeated blood transfusions and iron chelators as and when indicated. In India, the diagnosis of thalassemia is often delayed due to unawareness and lack of knowledge regarding the disease and lack of facilities for diagnosis everywhere. The children with Thalassemia become symptomatic after 3-6 months of age. Common clinical features are pallor, fatigue, poor appetite, lethargy due to anaemia. Children with severe disease who are not frequently transfused have ''thalassaemic facies'' which include frontal bossing, depressed nasal bridge, maxillary hyperplasia. The other features are growth failure, hepatosplenomegaly, and in some cases mild to moderate jaundice due to liver dysfunction. There is requirement of regular blood transfusion and iron chelating agents to remove excess iron which results from repeated blood tansfusions . However definitive treatment is bone morrow transplantation . It has been seen that children who are well transfused have normal growth and development. The most dreaded complication of thalassemia is iron overload and results in high mortality and morbidity . Iron overload results from ineffective erythropoiesis, increased gastrointestinal absorption, lack of physiological mechanism for excreting excess iron. Due to repeated blood transfusions, the patient have excess iron. Iron is very toxic to tissues, normally iron is bound to carrier protein transferrin . Due to repeated blood transfusions, the patients have excess iron that saturates transferrin and ''free iron'' level increases in blood and accumulates in different tissues mainly heart, exocrine gland, liver and leads to congestive heart failure, cardiac arrhythmias . Exocrine dysfunction like hypothyroidism, hypoparathyroidism, hypogonadotropic gonadism, growth deficiency , diabetes mellitus can occur.

Blood transfusions, though a well established form of management is not available to majority of thalassemia patients because of its limited availability.Patients often have to travel too far off distances for blood transfusion to centres where blood is regularly available. There are only few centres where these children are regularly given blood transfusion and monitored for various parameters. There are very few studies regarding benefits of regular blood transfusion, endocrine and cardiac complications and growth in children, however there is no statistics available regarding total number of thalassemia patients.

This prospective study is aimed to anthropology parameters in thalassemia patients.

II. Material And Methods

A prospective observational study will be carried out at pediatrics Out Patient Department (O. P. D.) and in patent department, RIMS, Ranchi.

The study will include 110 cases of children between the age of 1-18 years with beta thalassemia . All the cases and control registered in the study will be interrogated for detail history, clinically examined thoroughly and investigated. It covers following parameters.

Ethical consideration:

The synopsis of the study along with the informed consent form (in English & Hindi) was submitted to the Institutional Ethics Committee of Rajendra Institute of Medical Sciences (RIMS) ,Ranchi for approval. Study was done after approval was obtained in writing.

Consent :

Written informed consent was taken from each participant . illiterate patients gave their fingerprint (left thumb impression) instead of signature on consent form .

a) Anthropometric measures

i) Height
ii) Weight
iv) Upper segment:lower segment ratio

b) Detailed medical history

i)Family genetic studies
ii)Pedigree analysis
iii)Total amount of transfused blood and hemoglobin level at the time of transfusion

c) Investigations

i) Complete blood count (CBC)
ii) High performannce liquid chromatography (HPLC)

Duration of Study:

May, 2017 to June, 2018

Statistical analysis

Stastical analysis of the data was done using statistical package for the social sciences (SPSS) software. Data master sheet was generated for variables study . the quantitative data (age) will be expressed as mean ±standard deviation (SD) . P values less than 0.05 were considered statistically significant.

III. Observations And Results

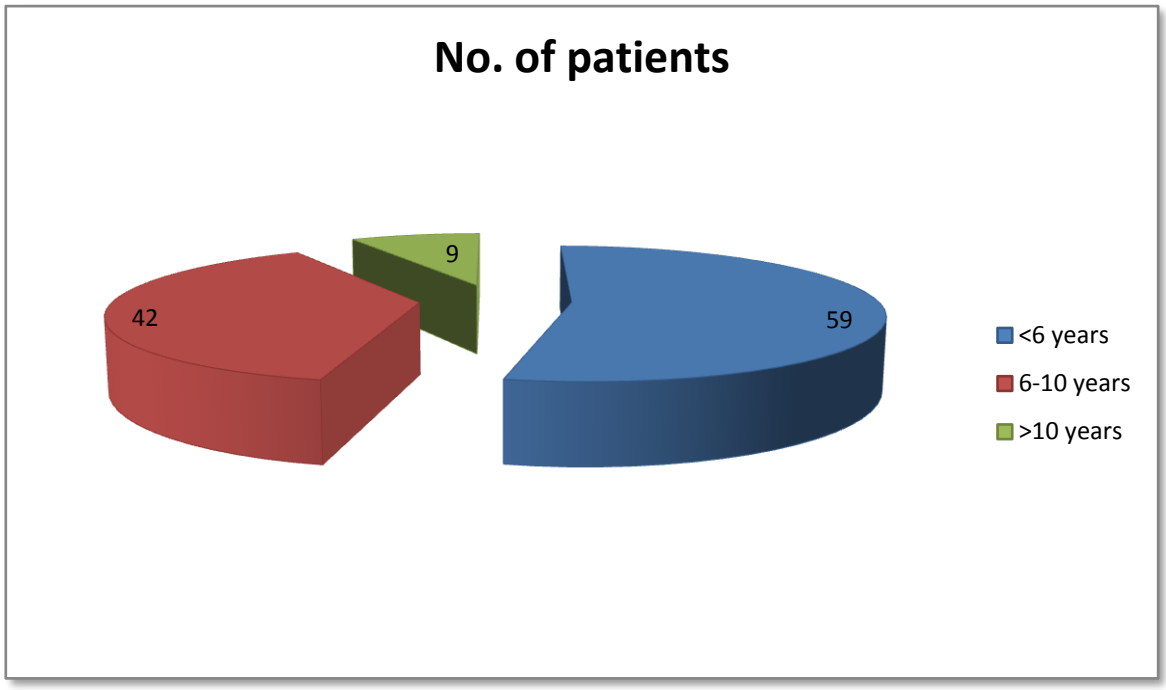
In the present study 110 cases of thalassemic children admitted in Department of Paediatrics and Neonatology of Rajendra Institute of Medical Sciences, Ranchi from the period May 2017 to June 2018 were studied. The children were on only blood transfusion and some were on single iron chelator (Deferasirox)
The results and observation of the present study has been presented in following tables.

Table -1

CASE DISTRIBUTION ACCORDING TO AGE

Among 110 transfusion dependent it was seen that majority of children i.e 59 children (53.63%) were less than 6 years ,42 children (38.18%) were in the age group 6-10 years and 9 children (8.1%) were more than 10 years.

Age	No. of patients
<6 years	59
6-10 years	42
>1 0years	9

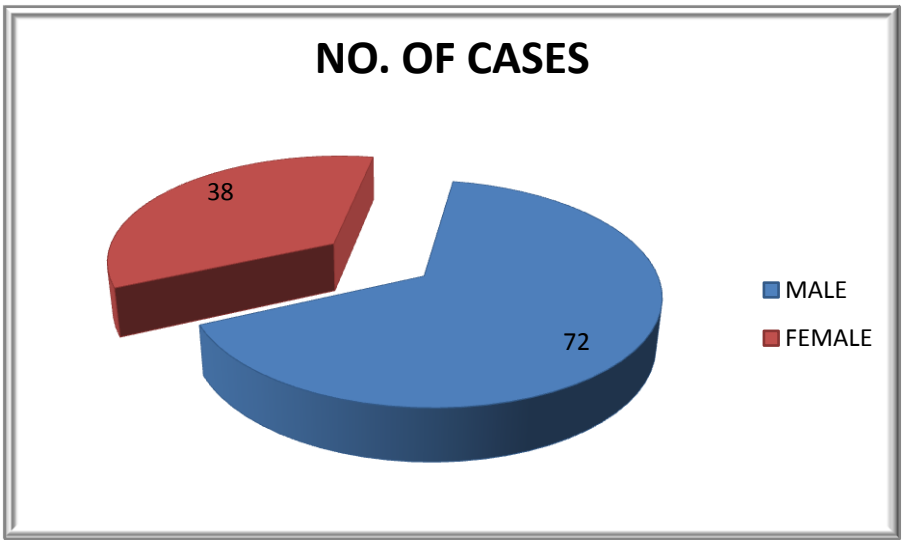


1. GRAPH SHOWING CASE DISTRIBUTION ACCORDING TO AGE

SEX DISTRIBUTION OF THE STUDY

Table no-2

SEX	NO. OF CASES	% AGE
MALE	72	65.45
FEMALE	38	34.55



2. GRAPH SHOWING SEX DISTRIBUTION AMONG PATIENTS

CASE DISTRIBUTION ACCORDING TO ANTHROPOMETRIC OBSERVATION IN COMPARISION TO ICMR IN DIFFERENT AGE GROUP

Table no-5

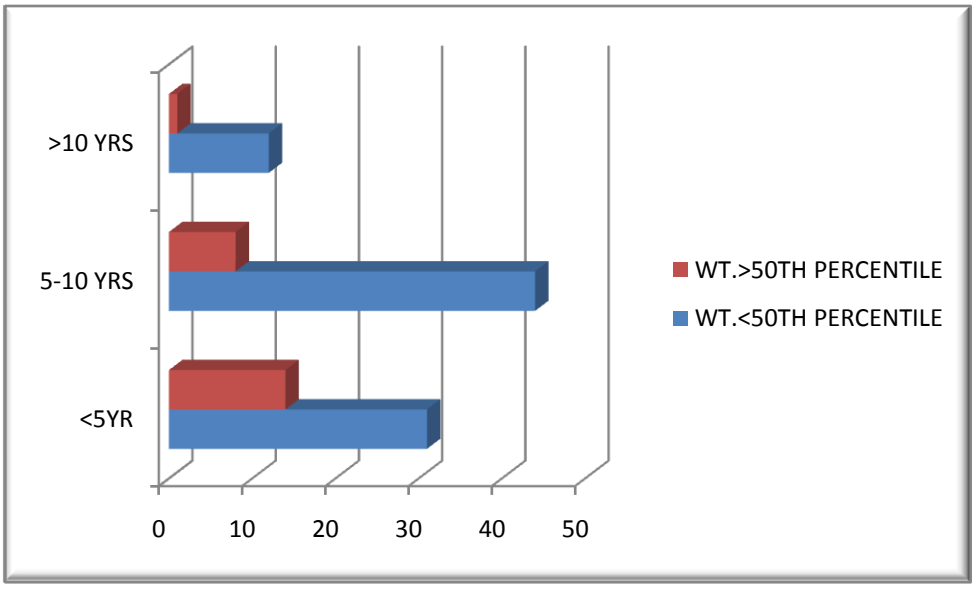
AGE GROUP	TOTAL NO. OF PATIENTS	WT. OF PATIENT		HT. FOR AGE	
		< ICMR (50^{TH} Percentile)	>ICMR (50^{TH} Percentile)	< ICMR (50^{TH} Percentile)	>ICMR (50^{TH} Percentile)
< 5 Yrs	45	31	14	17	18
5-10 Yrs	52	44	8	23	29
>10 Yrs	13	12	1	6	7

A total of 110 children were enrolled for this study and children were grouped according to their age. It was seen that in children less than 5 years, out of 45 children, 31 children (68.8%) and 17 children(37.7%) had weight and height less than ICMR standards.

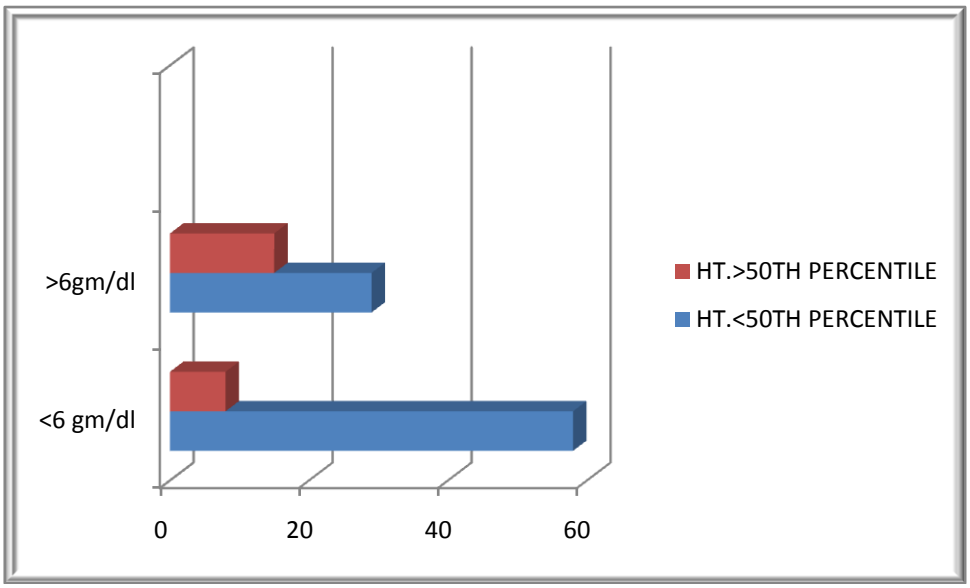
In children of age group 5-10 years, it was seen that out of 52 children, 44 children (84.6%) and 23 children(44.23%) had weight and height less than ICMR standards.

In children of age group > 10 years, it was seen that out of 13 children,12 children(92.3%) and 6 children (46.15%) had weight and height less than ICMR standards respectively.

According to the results it was seen that growth retardation was observed more, in early childhood more than 5 years.



3. Graph showing weight variation according to age



4. Graph showing height variation according to age

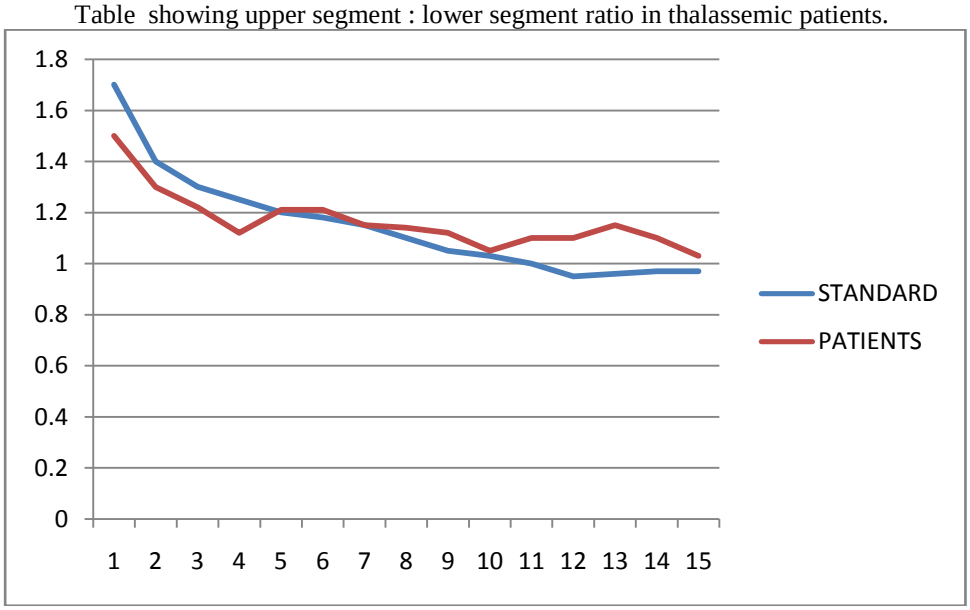
UPPER SEGMENT : LOWER SEGMENT RATIO IN THALASSEMIA PATIENTS

Table

AGE	STANDARD	PATIENT
1	1.7	1.5
2	1.4	1.3
3	1.3	1.22
4	1.25	1.12
5	1.2	1.21

6	1.18	1.21
7	1.15	1.15
8	1.1	1.14
9	1.05	1.12
10	1.03	1.05
11	1	1.1
12	0.95	1.1
13	0.96	1.15
14	0.97	1.1
16	0.97	1.03

Table showing upper segment : lower segment ratio in thalassemic patients.



GRAPH SHOWING UPPER SEGMENT LOWER SEGMENT RATIO IN THALASSEMIC AND NORMAL CHILDREN

Upper segment/lower segment ratio was low compared to standards till he age of 5 years depicting rapid growth in the leg length compared to trunk length. In the rest of the growth period, the ratio was high suggesting disproportion in body segments of patients.

IV. Conclusion

In our study, 46 (41.81%) patients had short stature (height for age Z score less than-2SD). Among 45 thalassemic patients *not* on iron chelator, 27 patients (60%) had height less than <- 2 SD . In β thalassemic patients growth disturbance is the main clinical feature that affects the life and wellbeing of such individuals. Our study has revealed that patients with beta thalassemia suffer from reduced height ,which is enhanced in patients having high levels of serum ferritin (ng/ml) and low haemoglobin (gm/dl).

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