IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 18, Issue 2 Ser. 7 (February. 2019), PP 25-30
www.iosrjournals.org

A Study on Growth Parameters and Clinical Profile in Children with Thalassemia Major

ParthaSarathiDas[1] ,Saikat Majumdar[2]
[1]*MBBS, MD Paediatrics, Senior Resident, Paediatrics, Kharagpur Sub divisional Hospital*
[2]*MBBS, MD Anesthesiology, DM Post Doctoral Trainee in CTVA (Associate Professor Cum Post Doctoral Trainee,*
Dept. of CardioThoracicVascularAnaesthesia, Nil RatanSircar Medical College
Corresponding Author: ParthaSarathiDas

Abstract:
Objectives: *To study the growth parameters & clinical & demographic profile in pediatric patients living with thalassemia major.*
Methods: *This cross-sectional, descriptive and analytical study was carried out on 30 thalassemic patients who received blood transfusion therapy at Kharagpur SD Hospital with respect to growth parameters such as Height/Length, Weight, Head Circumference; demographic profile such as age, gender & clinical profile such as presenting complaint & blood groups were studied. Collected data was compared to existing standardized IAP & WHO growth chart.*
Results*: Age distribution in our study group was bimodal (4 to 6 yrs& 9-12 yrs) with a male female ratio of 1:1, B positive blood group being the most common blood group. Most common presenting complaint was generalized weakness with mean hemoglobin of 4.33 ± 0.4 at presentation. Weight for age, height for age both were significantly low when compared to the standard growth curves with relative sparing of head growth (head circumference) with trend lines showing decline in Height & weight for age with age. A very high prevalence of both underweight (70%) and stunting (67%) noted in our study group, with almost 90% had a height for age & weight for age below mean.*
Conclusions: *Thalassemia major patients, are highly prone to growth failure with high prevalence of stunting and wasting which aggravates with age with relative sparing of head implying the need for effective early childhood interventions, growth monitoring, properly timed regular blood transfusion, sensitizing patients' family regarding early recognition of anemia, elevating the average pre transfusion Hemoglobin level (Hb) is the key to combat this severe growth faltering.*
Keywords: *Thalassemia major, growth failure, Hemoglobin.*

Date of Submission: 29-01-2019 Date of acceptance: 14-02-2019

I. Introduction

Thalassemia syndromes are a heterogeneous group of single gene disorders, inherited in an autosomal recessive manner, prevalent in certain parts of the world, including India. [1] The β thalassemias and sickle cell disorders pose a significant health burden in India. The average prevalence of β thalassemia carriers is 3–4%[2]. Growth failure in thalassaemia major (TM) has been recognized for many years, and has persisted despite major therapeutic advances. The child with TM has a particular growth pattern, which is relatively normal until age 9-10 years; after this age a slowing down of growth velocity and reduced or absent pubertal growth spurt are observed. The pathogenesis of growth failure is multifactorial. The fundamental problem is the free iron and hemosiderosis-induced damage of the endocrine glands. Additional factors may contribute to the etiology of growth delay including chronic anaemia and hypoxia, chronic liver disease, zinc and folic acid and nutritional deficiencies, intensive use of chelating agents, emotional factors, endocrinopathies (hypogonadism, delayed puberty, hypothyroidism, disturbed calcium homeostasis and bone disease) and last but not least dysregulation of the GH-IGF-1 axis. Three phases of growth disturbances according to age of presentation are well recognized, and have different etiologies: in the first phase growth disturbance is mainly due to hypoxia, anemia, ineffective erythropoiesis and nutritional factors. During late childhood (second phase), growth retardation is mainly due to iron overload affecting GH-IGF-1 axis and other potential endocrine complications.[3] Here we present the clinical & demographic pattern of paediatric patients living with thalassemia major that we experienced in our clinical practice in a period of 3 months at Kharagpur Sub Divisional Hospital.

II. Methods

We analyzed 30 paediatric patients with thalassemia over a period of 3 months. Data were collected in a predesigned proforma. Study design was cross sectional, observational, descriptive - analytical in nature. We organized & analyzed data in Microsoft Excel 2010 using standard statistical techniques. Statistical significance was analyzed with paired t test.

III. Results

Data from a total of 30 children aged birth to 12 years living with thalassemia attending thalassemia clinic for blood transfusion was collected in a pre designedproforma.

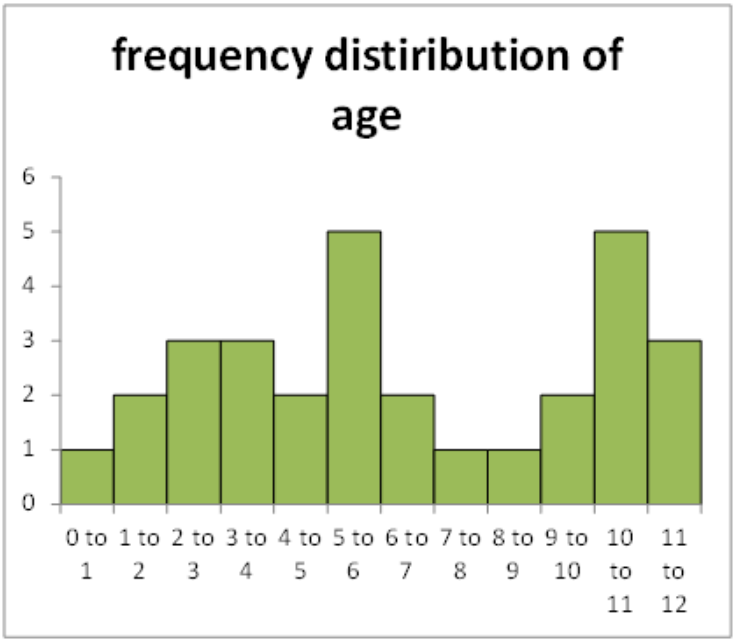

Figure 1: The bar diagram above shows the polymodal nature of age distribution in the study population with peaks at 5 to 6 & 10-11 years. The ratio of male to female was 1:1. There was no significant intergroup differences in demographic & growth parameters between 2 sex.

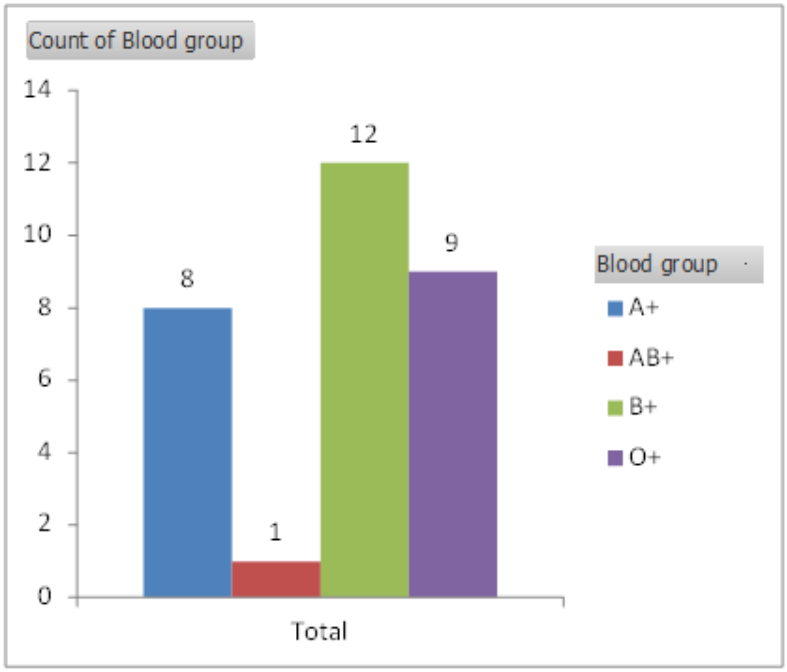

Figure 2: Distribution of various blood groups among study population, blood group B+ being the most common followed by O+.

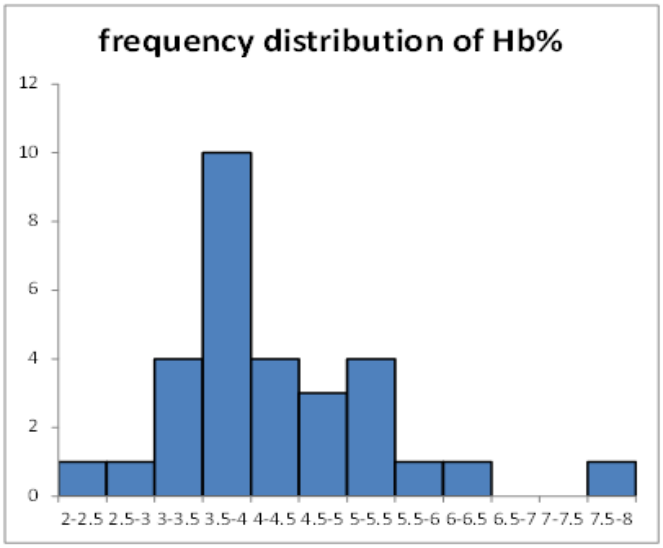

Figure 3: Histogram showing presenting Hb% with average being (4.33 $\pm$ 0.4) gm/dl.

Presenting Complaint	Number	Percentage
chest pain	3	10
cough & cold	6	20
generalized weakness	1	3.33
pain abdomen	1	3.33
pain in leg	5	16.67
shortness of breath	6	20
Muscular cramps	1	3.33
Generalized weakness	7	23.33
Grand Total	30	100

Table 1: Tabular depiction of various presenting complaints among the study population, most common being generalized weakness (23.33%) followed by Pain in legs (16.67%).

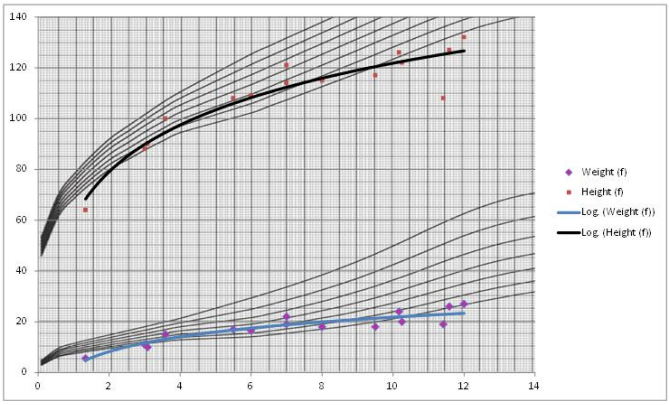

Figure 4: The distribution of weight & height for age among girls with thalassemia. Analysis shows almost all data points are below the mean of reference curve with trend line suggestive of worsening pattern of growth failure with advancing age. The best growth observed between 3 to 8 years suggesting early intervention at this point with regular follow up may prevent this to some extent. In the late childhood the height appears to be more affected than the weight strengthening the role of endocrine cause to the growth failure. The growth chart used as reference are WHO-IAP combined growth chart for Girls from 0-18 years. 11 out of 15 (73%) these patients were underweight & 10 out of 15 (67%) patients were stunted.

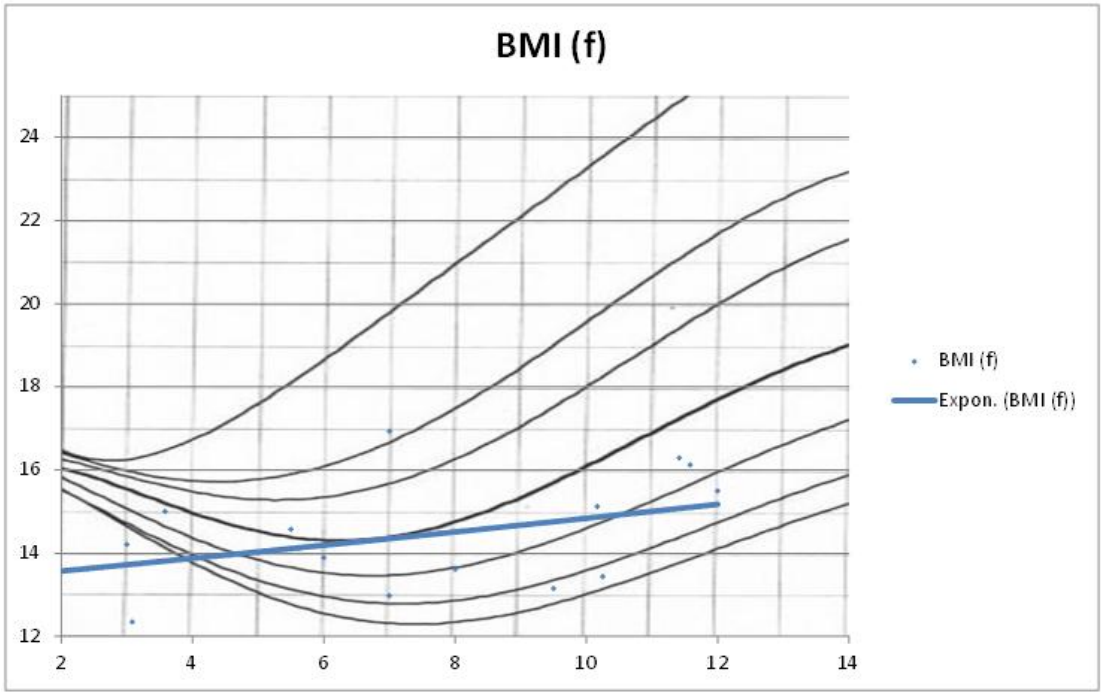

Figure 5: BMI percentiles were initially low compared to the standard growth curve (IAP-2007) but later it appeared to be within 2SD of mean suggesting proportionate affection weight & height & emphasizing the fact that BMI is not a good marker of childhood malnutrition mainly due to its wide range of variability.

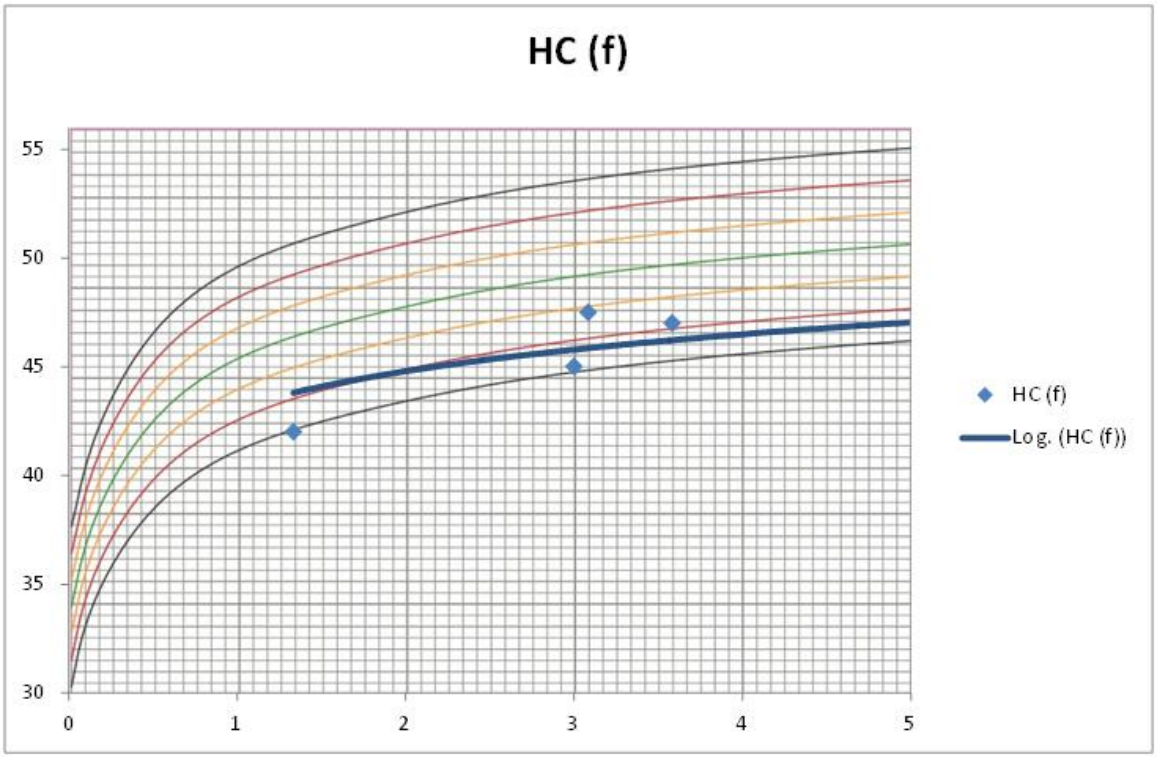

Figure 6: Head circumference were within 2-3 SD below the mean.

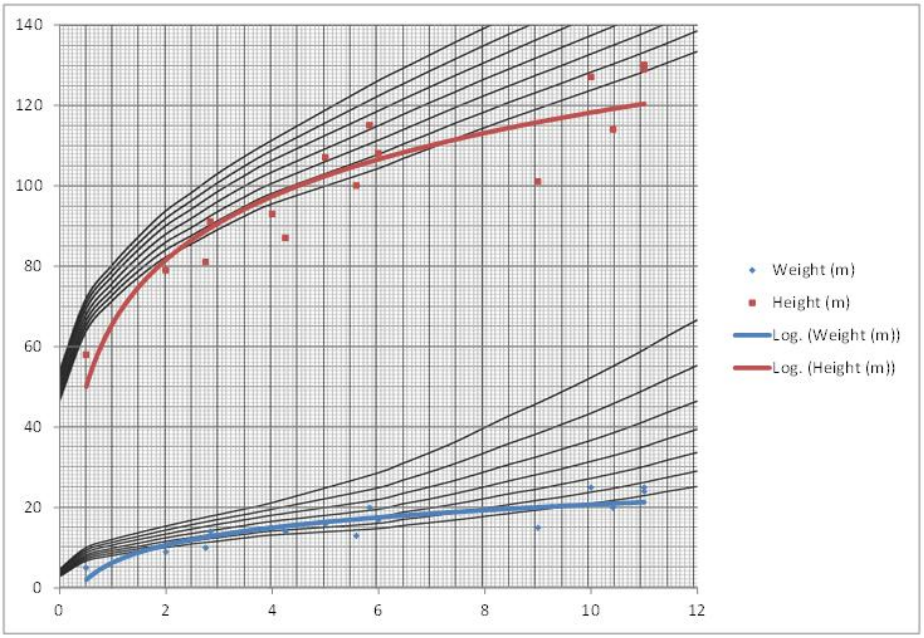

Figure 7: Weight & Height for age in boys with thalassemia shows the same trend with more pronounced growth failure with majority of data points falling below the 3rd percentile line with best growth parameters observed between 3-8 years & a worsening trend line with age. 10/15 (67%) patients were underweight & 10/15 (67%) patients were stunted.

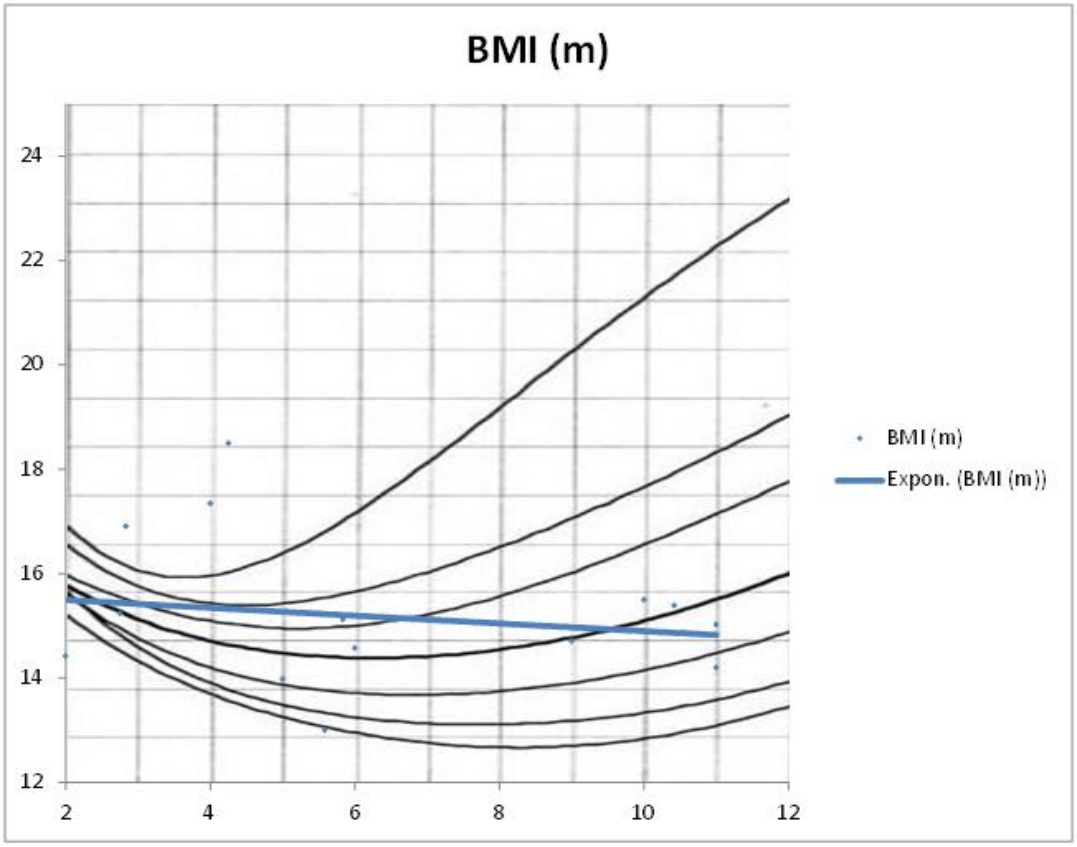

Figure 8: BMI, here also, appears to be a relatively less important marker as childhood growth parameter, though a decreasing trend has been observed in this population.

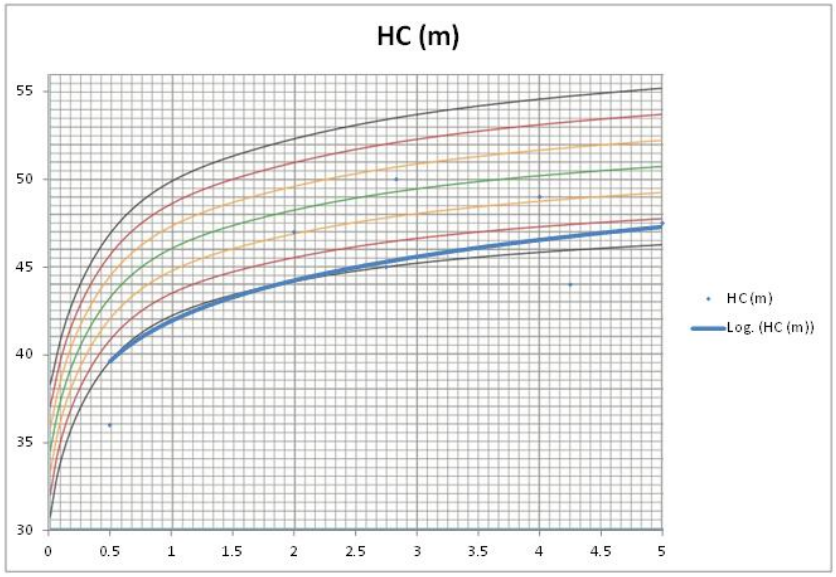

Figure 9: The pattern of Head growth in the population is relatively unaffected though individual data points are between 2 - 3SD below mean.

IV. Discussion

In our study growth failure appeared to be a common occurrence among patients with thalassemia with high prevalence of both underweight (70%) and stunting (67%) in our study group, with almost 90% had a height for age & weight for age below mean, which correlates with previous study by Tienboon et al[4]. Average pre transfusion Hb% was 4.33±0.4 gm/dl, which is lower than previously reported by Mona M Hamdy[5] in this regard (5.7 ± 1.16 gm/dl). We found the most prevalent ABO & Rh group in our study population being B+>O+>A+>AB+ unlike previous study by SoumyajitMaiti et al[6] . Best growth period in thalassemia turns out to be around 4 to 7 years in our study group, indicating frequent follow up, regular blood transfusion, chelation & other relevant intervention started on or before this point may have a lasting effect in halting growth faltering[7,8].

Contributors:PSD : study conceptualization, data collection, analysis and manuscript writing. SM: data analysis and manuscript writing; AS: manuscript writing. All authors approved final version of manuscript.

Acknowledgement

We thank Dr.Krishnendu Mukherjee, Dr. ArabindoMahato, Dr. Indrani Das, Dr. AshmitaSaha for their immense support, enthusiasm regarding study design & sample collection and data analysis.
Additional Clinical Investigators: Dr. SekhAftab, Dr. Rathindranath Paik, Dr. Raju Das.
Funding: None.

References

[1]. Naveen Thacker, Prevention of Thalassemia in India, Indian Pediatrics 2007; 44: 647-648 .
[2]. N. Madan, S. Sharma, S.K. Sood, R. Colah, H.M. Bhatia Frequency of ß thalassemia trait and other hemoglobinopathies in northern and western India; Indian J Hum Genet, 16 (2010), pp. 16-25 .
[3]. Skordis N, Kyriakou A, The multifactorial origin of growth failure in thalassaemia, PediatrEndocrinol Rev. 2011; Suppl 2:271-7 .
[4]. Tienboon P, Sanguansermsri T, Fuchs GJ; Southeast Asian J Trop Med Public Health. 1996;27:356-61.
[5]. Mona M Hamdy, Lamis A Ragab, Iman A Shaheen, Rania N Yassin; Blood transfusion among thalassemia patients: A single Egyptian center experience; Asian Journal of Transfusion Science, Vol. 7, No. 1, 2013, pp. 33-36 .
[6]. BikashMonda, SoumyajitMaiti, Biplab Kumar Biswas, Debidas Ghosh and Shyamapada Paul; Prevalence of hemoglobinopathy, ABO and rhesus blood groups in rural areas of West Bengal, India; J Res Med Sci. 2012 ; 17: 772–776.
[7]. Rodda CP, Reid ED, Johnson S, Doery J, Matthews R, Bowden DK. Short stature in homozygous beta-thalassaemia is due to disproportionate truncal shortening. ClinEndocrinol1995 ;42:587–92 .
[8]. Andreas Kyriakou, and NicosSkordis, Thalassaemia and Aberrations of Growth and Puberty; Mediterr J Hematol Infect Dis. 2009; 1:e2009003.Published online 2009 .

ParthaSarathiDas. "A Study on Growth Parameters and Clinical Profile in Children with Thalassemia Major." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 18, no. 2, 2019, pp 25-30.