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Original Research Article

To study the growth retardation in relation to age, pre-transfusion haemoglobin level, serum ferritin level in pediatrics patients.

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Abstract:

Background &Method: The aim of this study is to study the growth retardation in relation to age, pre-transfusion haemoglobin level, serum ferritin level in pediatrics patients.The serum ferritin level was measured using ADVIA Centaur system at Endocrine laboratories. The ADVIA Centaur ferritin assay is a two-site sandwich immunoassay using direct chemiluminometric technology, which uses constant amounts of two anti-ferritin antibodies.

Result: Out of 160 cases, 66 cases were from age group 1-5 years, 60 cases were from age group 6-10 years and only 34 cases were from age group 11-14 years.

Conclusion: There is significant growth retardation in boys and girls in all the age-groups when compared with 50th percentile scores by N.C.H.S. Out of 160 cases, 66 cases were from age group 1-5 years, 60 cases were from age group 6-10 years and only 34 cases were from age group 11-14 years. Adequate pre-transfusion haemoglobin level should be maintained so that growth retardation because of anaemia should be minimized.

Keywords: pre-transfusion, haemoglobin, serum ferritin and iron chelation.

Study Designed: Observational Study.

1. INTRODUCTION

Thalassemia is the genetic disorders in globin chain production. In individual with β-thalassemia, there is either a complete absence of β-globin gene production or a partial reduction. Although, β-thalasemia has more than 200 mutations, most are rare. Approximately 20 common alleles constitute 80 percent of the known thalassemia worldwide. 3 percent of the world's population carries gene for β-thalassemia[1]. It has high incidence in a broad tropical belt extending from Mediterranean region through the middleeast and far east. The largest concentration of thalassemia patients is seen in South-east Asia, Bangladesh, North-west India, Pakistan, Middle-east countries, North-Africa, Greece and Italy[11]. Frequency of thalassemia gene in Indian population varies between 0-17 percent in different ethnic group with average of over 3 percent. Its prevalence is high among Gujratis, Punjabis, Sindhis, Lohanas etc. Over thirty million people are carriers of thalassemia gene in our country. Ten thousands thalassemic children are born every year in India[2].

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The major features contributing to the pathogenesis of sequelae of β-thalassemia are inadequate β gobin chain production leading to decreased levels of normal haemoglobin(Hb A) and an imbalanced α and β globin chain production. There is disruption of maturation of red blood cells, resulting in ineffective erythropoeisis. Though the marrow is hyperactive, the patient has relatively few reticulocytes and severe anaemia. Since there is an excess of α chains relative to β and γ chains, α tetramers are formed. These inclusions shorten the survival of red blood cells by interacting with red cell membrane, thus leading to anaemia. There is elevated Hb Fand Hb A2 levels due to increased production of γ globin and δ globin chains respectively[3].

Anaemia stimulates production of erythropoietin. This leads to increased erythropoiesis which is ineffective. Increased erythropoiesis results in expansion of medullary cavities of various bones. The liver and spleen are enlarged as a result of extra-medullary haematopoiesis[4].

2. MATERIAL & METHOD

This study was conducted on 160 children with β-thalassemia major aged between 1-14 years being regularly transfused at Dept. of Pediatrics, ESIC Hospital, IndoreDec 2019 to Jan 2021, after taking the consent from the parents and explaining them the purpose and method of study.

The serum ferritin level was measured using ADVIA Centaur system at Endocrine laboratories. The ADVIA Centaur ferritin assay is a two-site sandwich immunoassay using direct chemiluminometric technology, which uses constant amounts of two anti-ferritin antibodies. The antibody, in the Lite Reagent, is a polyconal goat anti-ferritin antibody labelled with acridine ester.

INCLUSION CRITERIA

1. Child suffering from β-thalassemia major only as confirmed by Hb electrophoresis.
2. Age of thalassemic child should be between 1-14 years.
3. Attending dept. of pediatrics

EXCLUSION CRITERIA

1. Child suffering from β-thalassemia minor or any other associated haemoglobinopathy.
2. Other cases of anaemia requiring regular blood transfusion.All the thalassemic patients made 2 visits at an interval of 6mth.

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3. RESULTS

TABLE-1: AGE-WISE DISTRIBUTION OF CASES

AGE – GROUP (YEARS)	NO. OF CASES	% OF CASES
1-5	66	41%
6-10	60	38%
11-14	34	21%
TOTAL	160	100%

Out of 160 cases, 66 cases were from age group 1-5 years, 60 cases were from age group 6-10 years and only 34 cases were from age group 11-14 years.

TABLE-2: Mean pre-transfusion Hb levels in age between 1-5 years

AGE (years)	MEAN – PRETRANSFUSION Hb IN GIRLS (g/dl) ± S.D.	MEAN- PRETRANSFUSION Hb IN BOYS (g/dl) ± S.D.
1	7.9±0.307	-
2	5.34±1.654	7.4±1.51
3	6.9±0.107	6.2± 1.92
4	6.41 ±1.93	3.5 ± 0.6
5	6.6±0.37	6.81 +1.33

TABLE-3: Mean serum ferritin levels in age between 1-5 years

AGE (years)	MEAN SERUM FERRITIN in GIRLS (ng/ml) ± S.D.	MEAN SERUM FERRITIN LEVELS IN BOYS (ng/ml) ± S.D.
1	587± 73.69	-
2	1413.2±1381.67	1017.15± 1259.23
3	1793.9±94.51	2791.7±814.57
4	2247.51± 1031.55	2960.89
5	3523.9± 207.1	2701± 1169.44

4. DISCUSSION

Growth retardation has been accounted for to happen in many patients with thalassemia major. This is accepted to be expected, to some degree, to an immediate impact of iron over-burden on the endocrine instruments of pubescence. Development Chemical (GH) impedance at various levels (hypothalamic or pituitary) or potentially decreased insulin like development factor-1 (IGF-1) amalgamation have been proposed to be the primary drivers of hindered development in these patients[5]. Different causes incorporates - persistent sickliness, hypersplenism, ongoing liver illness (HBV,HCV), zinc and folic lack, skeletal dysplasia, desferrioxamine harmfulness and profound aggravations.

In the concentrate by Moayeri and Oloomi (2006) [5], short height was recognized in 98 patients (62%) with thalassemia major (59 % females and 65% guys). Of these 98 patients, 78 were no less than 3SD underneath the typical level.

In a cross-sectional investigation of 68 kids with β-thalassemia major in 75 % of the young ladies and 62% of the young ladies were underneath the third percentiles in level. Out of 233 thalassemic patients, 68.7% were put underneath the I.C.M.R. mean for weight and 71.2% were put underneath the I.C.M.R. mean for height[6].

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In the current review, 59% of thalassemic patients (61% young ladies and 57% young men) were underneath the I.C.M.R. standards for weight and half of thalassemic patients (53% young ladies and 48% young men) were underneath the I.C.M.R. standards for level. When contrasted with N.C.H.S. standards, 100 % of thalassemic patients were beneath the 50th percentile scores by N.C.H.S. While, 36% of thalassemic patients had short statured in concentrate by Aydinok et al. (2002)[7] in Turkey. Shamshiraz et al. of Iran (2003)[8] enlisted 220 thalassemic patients in their review. Short height (level underneath third percentile) was seen in 39% of their patients.

5. CONCLUSION

There is significant growth retardation in boys and girls in all the age-groups when compared with 50^{th} percentile scores by N.C.H.S. Out of 160 cases, 66 cases were from age group 1-5 years, 60 cases were from age group 6-10 years and only 34 cases were from age group 11-14 years. Adequate pre-transfusion haemoglobin level should be maintained so that growth retardation because of anaemia should be minimized.

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