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Original Research Article

A Study to Assess the Ocular Biometric Parameters and Prevalence of Refractive Errors among Thalassemic Children in a Rural Based Tertiary Hospital

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ABSTRACT

Introduction: Thalassaemia major patients have characteristic skeletal changes, including typical craniofacial changes and deformities of the long bones that result from expansion of the bone marrow. Craniofacial changes in thalassaemia major patients might lead to an abnormal bony orbit and subsequently might lead to distinctive ocular biometry and/or refraction.
Materials and Methods: 523 Thalassemic children and 502 age and sex matched control population were selected by systematic random sampling. Comprehensive ophthalmological examination was done on both case and control groups. The obtained data has been analysed by standard statistical software.
Results and Analysis: The Thalassemic children had lower mean weight than the control group (p<0.00, t=99.0, SED: 0.17, 95%CI:-16.72 to -16.07). The mean axial length of the eyeball was less among the Thalassemic children (p=0.04, t=2.00, SED: 0.10, 95% CI:-0.40 to 0.00). The average lenticular thickness was comparatively more among Thalassemic Children (p=<0.00, 10.64, SED:0.00, 95%CI: 0.05 to 0.08). The average keratometry value was lower among the Thalassemia children (p=0.04, t=2.08, SED: 0.10, 95%CI: -0.39 to -0.01). Astigmatism was more among the Thalassemic children (p=0.04, z=2.06).
Conclusion: Due to differential phenotypic expressions Thalassemic children may have abnormal ocular biometric characteristics.

Key Words: Thalassemic Children, Ocular Biometry, errors of Refraction.

INTRODUCTION

Thalassemia is an autosomal recessive disorder characterized by the reduced or absent expression of the β-globin gene, leading to an imbalance of α and β globin chains. [1] It is one of the most common autosomal recessive disorders worldwide. [2] In India alone 30 million people carry the defective gene and nearly 10,000 lethally affected homozygotes are born annually comprising 10.0% of the children born in the world annually. [3] The

Thalassemias are a major health problem in India. Yet, it has received little attention due to other health priorities, such as malnutrition and communicable diseases. Moreover, the burden of the disease is not uniform in the country with certain communities having appreciably greater prevalence of the disease than the others. [4]

Individuals with thalassemia major usually present within the first two years of life with severe anemia, requiring regular red blood cell [RBC] transfusions. Findings in untreated individuals with Thalassemia major are growth retardation, pallor, jaundice, poor musculature, hepatosplenomegaly, leg ulcers, development of masses from extramedullary hematopoiesis, and skeletal changes that result from expansion of the bone marrow. Peripheral blood smear shows, in addition to microcytosis and hypochromia, anisocytosis, poikilocytosis and nucleated red blood cells. Hemoglobin pattern [by cellulose acetate electrophoresis or high performance liquid chromatography (HPLC)] varies according to the type of β-Thalassemia. In β-Thalassemia, characterized by the lack of beta globin chain synthesis, HbA is absent, HbF is 95–98%, and HbA2 is 2–5%. [2]

Thalassaemia major patients have characteristic skeletal changes, including typical craniofacial changes and deformities of the long bones that result from expansion of the bone marrow. Craniofacial changes comprise bossing of the skull, prominent malar eminence, depression of the bridge of the nose, tendency to a mongoloid slant of the eye and hypertrophy of the maxillae. [5-12] Ocular growth is intimately related to the growth of the adjacent bony orbit. In principle, craniofacial changes in thalassaemia major patients might lead to an abnormal bony orbit and subsequently might lead to distinctive ocular biometry and/or refraction. [6-12]

There is not enough data regarding any significant change in the shape of the eye ball due to changes in the bony orbit in Thalassemic children. Whether these changes in ocular biometry, if any, are getting translated into errors of refraction among these children also need to be thoroughly investigated. The present study intends to investigate the ocular biometric parameters and prevalence of refractive errors among Thalassemic Children in a rural based tertiary hospital in Eastern India.

MATERIALS AND METHODS

523 Thalassemic children who have met the inclusion criteria were selected for the study after a systematic random sampling. Preverbal children and children who were too sick to undergo comprehensive ocular examination were excluded from this cross sectional institution based one year prevalence study. Parents or care providers of the selected Children had undergone a detailed personalised interview and pretested and predesigned questionnaires were filled up to document the elicited information. Additional information was collected from the registers of the Thalassemia unit and In-patient registers of Department of Paediatric Medicine and Department of Internal Medicine. Then the children had undergone anthropometric examination and detailed comprehensive Ophthalmological examination including USG A scan, Keratometry and objective refraction with Auto refractometer with cycloplegics as per age wise recommendations. The following study definitions were considered:

a. Myopia: More than -0.25 D reading in the auto refractometer in either eye was documented as Myopia.
b. Hypermetropia: More than +0.25D reading in the auto refractometer in either eye was documented as Hypermetropia.
c. Astigmatism: Cylindrical power present in any axis in either eye was documented as Astigmatism.
d. Anisometropia: Difference of Spherical power of 5D or more between the two eyes was considered as Anisometropia.

502 age and sex matched children were selected as control population from

Out Patient Department of Paediatric Medicine.

RESULTS AND ANALYSIS

The demographic data between the Thalassemia Group and the control Group were comparable. The mean height among the Thalassemia Group was 124.2±11.8 cms as compared to 126.0±12.4 cms among the Control Group (p=0.02, t=2.38, SED: 0.76, 95% CI:-3.28 to -3.10). The mean weight among the two groups were 37.8±2.5 kg and 54.2±2.8 kg respectively and the difference is statistically significant (p<0.00, t=99.0, SED: 0.17, 95% CI:-16.72 to -16.07). However, when the head circumference between the two groups were compared, the difference was statistically non-significant (52.9±2.4 cms vs 53.1±2.3 cms, p=0.17, t=1.36, SED: 0.15, 95% CI: -0.49 to 0.09).

In the present study it was seen that the mean axial length of the eyeball among the children in the Thalassemia Group was 21.6±1.6 mm and that found among the children in the control group was 21.8±1.6 mm. The difference was investigated to be statistically significant (p=0.04, t=2.00, SED: 0.10, 95% CI:-0.40 to 0.00). Similarly, the thickness of the lens was also found to be statistically significant. The mean thickness among the Thalassemic Group was 4.04±0.11 mm as compared to 3.97±0.10 mm among control Group (p=<0.00, 10.64, SED: 0.00, 95% CI: 0.05 to 0.08). However the mean anterior chamber depth among the Thalassemia Group was 3.52±0.09 mm and that among the Control Group was 3.53±0.08 mm and the difference was statistically non-significant (p=0.06, t=1.88, SED: 0.01, 95% CI:-0.02 to 0.00). In the same manner, the mean depth of the vitreous Chamber between the Thalassemia Group and the Control Group was found to be 15.44±0.45 mm and 15.50±0.53 mm respectively. The difference was not statistically significant (p=0.05, t=1.96, SED: 0.03, 95% CI: -0.12 to 0.00).

Average Keratometry values of both eyes were 43.6±1.8 D among the Thalassemia Group where as among the Control Group it was 43.8±1.2 D. This difference was found to be significant when analysed statistically (p=0.04, t=2.08, SED: 0.10, 95%CI: -0.39 to -0.01).

While analysing the prevalence of errors of refraction, no significant statistical difference was documented between the Thalassemia Group and the Control Group (16.82% vs 20.52%, p=0.13, z=1.52). 10.7% of the children in the Thalassemia Group were found to have Hypermetropia as compared to 13.35% of the children in the Control Group (p=0.19, z=1.29). However, 9.18% of the children in the Thalassemia Group were found to have astigmatism where as 5.78% of the children in the Control Group were found to have astigmatism. The difference was detected to be statistically significant (p=0.04, z=2.06). The number of anisometropia among the Thalassemia Group and the Control Group were 2.87% and 2.59% respectively. The difference was not statistically significant (p=0.79, z=0.27).

Table 1: Showing the demographic and anthropometric Data among the Study population and Control population (N=523,n=502)

Parameters	Thalassemia Group (N=523)	Control Group (n=502)	P value
Age (in years)	9.0±3.88	9.0±3.76	1.0
Sex(male%)	62.14%	61.55%	0.85
Height (in cms)	124.2±11.8	126.0±12.4	0.02
Weight (in kg)	37.8±2.5	54.2±2.8	<0.00
Head Circumference (in Cms)	52.9±2.4	53.1±2.3	0.17

Table 2: Showing the biometric and keratometric data between the study population and control population (N=523,n=502):

Parameters	Thalassemia Group (N=523)	Control Group (n=502)	P value
Axial Length (in mm)	21.6±1.6	21.8±1.6	0.04
Anterior Chamber Depth (in mm)	3.52±0.09	3.53±0.08	0.06
Lens Thickness (in mm)	4.04±0.11	3.97±0.10	<0.00
Vitreous Chamber Depth (in mm)	15.44±0.45	15.50±0.53	0.05
Keratometry (D)	43.6±1.8	43.8±1.2	0.04

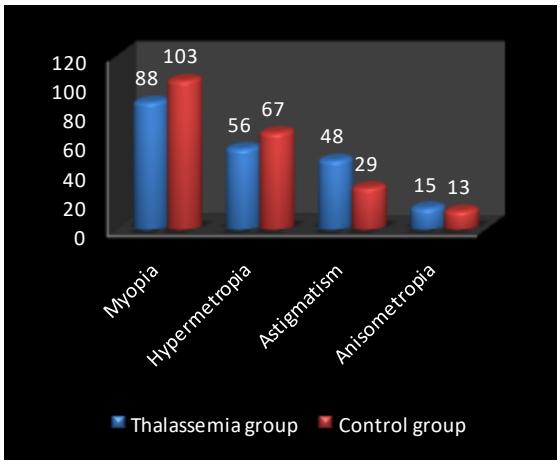

Figure 1: Showing the prevalence of refractive errors among study population and control population (N=523, n=502):

DISCUSSION

The mean age among the children suffering from Thalassemia was 9±3.88 years and that among the Control population was 9±3.76 years. In a similar study conducted by Aksoy A et al [13] among children suffering from Thalassemia in Mediterranean region, the mean age among Thalassemic children was 9.31±3.89 years and that among the control population was 8.05 ±2.19 years. About 62.14% of the children in the Thalassemia Group were males where as about 61.55% were males in the Control Group. However in the study conducted by Nowroozzadeh MH et al, [12] there were about 36.2% and 38.6% males in the Thalassemia Group and the Control Group respectively. The male predominance in the present study may be explained with the fact that in a rural Indian society like the present study venue, the male children enjoy a preferential attitude than their female counterparts from the family members.

The mean height among children suffering from Thalassemia in the present study was 124.2±11.8 cms as compared to 126.0±12.4 cms among children in the Control Group. In a similar fashion, in the study conducted in Iran by Nowroozzadeh MH et al [12] showed that the mean height among Thalassemia Group to be 143.37±13.22 cms and among the Control Group to be 155.22 ±14.77 cms ($p<0.00$). This supports the observation from the present study that children suffering from Thalassemia have stunted growth. The above study also shows in sync with the present study that children suffering from Thalassemia are also under weight as compared to their normal counterparts. However, the present study has found no significant correlation with Head circumference between the Thalassemia Group and the Control group in harmony with the findings of Nowroozzadeh MH et al. [12]

The mean Axial Length of both eyes of the children in the Thalassemia Group was less than that of Control Group in the present study ($p=0.02$). Similarly, the study conducted in Iran[12] showed that the mean Axial Length among the Thalassemia Group was 23.01 ±0.12 mm as compared to 23.46 ±0.13 mm among the Control Group ($p=0.03$). However , the study conducted by Aksoy A et al [13] in the Eastern Mediterranean showed no statistically significant difference between the Mean Axial Length between the Thalassemia Group and that of the Control Group ($p>0.05$). The genetic mutations in Thalassemia are diverse with direct association with the phenotypical expression of the disease. It may be a possibility that the orbital changes bringing changes in the length of the eyeball secondary to extramedullary erythropoiesis may not be uniformly expressed in all population. The effects of Thalassemia on the Axial Length of the eyeball need further investigation from population based longitudinal studies in future.

The present study has shown average thickness of crystalline lens was greater in the Thalassemic Group than the Control Group ($p<0.00$). However, there was no statistical difference between the two groups while assessing the mean Anterior Chamber Depth and Depth of Vitreous Chamber ($p=0.06$, 0.05). Similar findings were echoed in the study conducted by Nowroozzadeh MH et al. [12] Both the present study and the study conducted in Iran has shown steeper average Keratometry

values among the Thalassemia Group and the Control Group.

The mean visual acuity among the children suffering from Thalassemia in the present study is 0.32±0.14 and that among the Control group is 0.30±0.12 (p=0.01). Though the mean acuity among the Thalassemia Group is not unsatisfactory yet the difference is statistically significant when compared with the Control Group. The study conducted by Aksoy A et al [13] showed that the mean visual acuity among Thalassemia group was 1.34±0.75 where as that among the Control Group was 1.08±0.28 and the difference was statistically significant (p<0.05). However, the study conducted by Nowroozzadeh MH et al [12] has shown no significant difference in logMAR visual acuity at presentation between the two groups (p=0.71). Unlike our study the study conducted by Nowroozzadeh MH et al [12] has shown no significant difference in the prevalence of Astigmatism between the Thalassemia Group and the Control Group (p=0.83).

CONCLUSIONS

The difference in the ocular Biometric and refractive characteristics as evidence in the present study and elsewhere can be explained with the following postulation: there is no significant change in the head circumference among the children suffering from Thalassemia despite reduced height and weight suggests that there is should not be any appreciable difference in ocular dimensions between the two groups. However, there is no denying the fact that there is some amount of shortening of the Axial Length of the eyeball in children suffering from Thalassemia than their normal counterparts. To outweigh the adverse refractive outcome of the possibly there is steeper corneal curvature and thicker Crytalline lens leading to a state of emmetropia more or less comparable with the general population. This explains why there is no shift in the pattern of refractive errors among the Thalassemia Group and Control Group in this study and previous studies. However, the present study has additionally showed more prevalence of Astigmatism among the Thalassemia Group than the Control Group. This raises the possibility that the ocular biometric adjustments to achieve emmetropia among the children suffering from Thalassemia are not uniform in all populations and in some cases residual Astigmatism may remain causing visual impairment. This may be at a larger aspect attributed to the non-uniformity in the phenotypic expression of Thalassemia especially among the Indian population. A longitudinal community based study of ocular biometric parameters among the same ethnic population in near future may reveal a brighter perspective on the matter.

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