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Original article:

Cephalic index in children with E Beta thalassemia: study at a tertiary care centre in eastern India

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Abstract:

Introduction: Hemoglobin E Beta thalassemia (EB thal) a clinically diverse disorder, is common in eastern India. Facial deformities are a prominent feature in transfusion dependent EB thal. This study attempts to show the clinical variation in an objective manner. The purpose of this study is to evaluate the craniofacial dimensions of EB thal patients and to identify differences by comparing them to those of control group.

Method: The study was conducted at a tertiary care centre in Kolkata city of eastern India, over one year period. Craniometric measurements were taken by spreading callipers , from children, upto 12 years age. Similar measurements were also done in controls. The head length was measured from the glabella to the inion, while the head breadth was measured as the distance between the euryons. The Cephalic Index (CI) is calculated as the ratio of the head width to the head length multiplied by 100.

Results and conclusion: The commonest and rarest head shape overall were mesocephaly and dolicocephaly respectively. Further mesocephaly was the commonest head shape in children who required blood transfusion before the age of four years. In addition , when CI was compared between children of EB thal and control groups, across all age groups, no significant difference was found.

Keywords: Hemoglobin E Beta thalassemia , Cephalic Index

Introduction:

The population of India is diverse with numerous ethnic groups. Hemoglobinopathies are the commonest hereditary disorder in India and pose a major health problem . Hemoglobin E Beta thalassemia(EB thal) is a common and clinically diverse disorder. The marked clinical variability ranges from mild asymptomatic anemia to a life threatening disorder requiring regular blood transfusion[1,2,3]. High prevalence of the disease is observed in eastern India where it is common for individuals to inherit alleles for both HbE and Beta thalassemia.[1,4,5]. Attempts to categorize the severity of EB thal have included the assignment of patients to severe and mild groups between which putative genetic and environmental factors are then compared. Genetic factors include the type of beta thalassemia mutation coinherited with HbE, the coinheritance of alpha thalassemia, and that of polymorphisms shown to be associated with increased synthesis of fetal haemoglobin [3].The characteristic physical features of severe Hb EB thal include frontal bossing of the skull , maxillary

hyperplasia , depressed bridge of nose and general growth retardation. The study of cephalometric indices is important in areas of Pediatrics , Plastic surgery, Oral surgery and Forensic medicine. Cranial or Cephalic Index (CI) is one of the clinically recognized anthropometric parameter in the investigation of craniofacial skeleton because of its validity and practicability[6,7,8] .CI is based on the length and breadth of the head .

Aim and objective : There is paucity of data on the CI of children and adults with EB thal , across the clinical spectrum of severity, in India . This study attempts to show the clinical variation in an objective manner. The purpose of this study was to evaluate the craniofacial dimensions of Hb EB thal patients and to identify differences by comparing them to those of a control group.

Material and method:

Study location design and duration: The study was conducted at RG Kar Medical College and Hospital, a tertiary care centre, in Kolkata, West Bengal. This is an observational descriptive study done over a period of one year,(October 2018 to Sep 2019).

Measurement and procedure: The head measurements were carried out in the dept of Pediatrics, at RGKMCH. Craniometric measurements were taken from 45 children, (upto 12 years old) with transfusion dependent thalassemia. Fifty normal growing children, without Thalassemia/chronic haemolytic disorder, visiting the department due to other ailments , were also included in the study. Further, children with craniofacial malformation, or history or sign of craniofacial syndromes were excluded from this study.

The details of the children as regard their name, age, sex, age at first blood transfusion, and present frequency of transfusion. Measurements were taken using standard recognised anatomical landmarks. All measurements were taken to the nearest 0.1 cm. The CI was calculated using measurements from each subject. The head length (greatest anteroposterior diameter) and head width (maximum transverse diameter between two definite points) were measured with the help of spreading callipers. The head length was measured from the glabella to the inion, while the head breadth was measured as the distance between the euryons. The CI is calculated as the ratio of the head width to the head length multiplied by 100. The CI has been used to classify head shapes into four categories, namely Dolicocephalic (CI≤ 74.9), Mesocephalic (CI=75.0-79.9), Brachycephalic (CI=80.0-84.9) and hyperbrachycephalic (CI≥85.0)(7,8). Ethical clearance was obtained from relevant department and the institution. The CI from patients and age and sex matched controls were entered and analysed using SPSS version 17. Student t-test was used for comparison between means and statistical significance set at p=<0.05.

Observation and results:

The commonest head shape overall was mesocephaly (40%), while dolicocephaly was the least common(table1). Mesocephaly was also the commonest head shape in children above eight years of age. In children upto four years age, brachycephaly was the commonest head shape. Further, hyperbrachycephaly was found to be in similar frequency in 5-8 years and greater than eight years age groups(table 2).

In all of the children, blood transfusion was initiated before eight years age. Only one child had undergone splenectomy. In majority of children (40/45), it has been started by the age of four years. In this age group again mesocephaly was the commonest head shape, while the distribution of hyperbrachycephaly and dolicocephaly was found to be equal (six each, table 3).

Further, CI was compared between children with EB thal and their control groups. In all age groups, viz upto four years, 5-8 years, and 9-12 years, the difference was not significant($p>0.05$, table 4)

Table1

Head shape	Male	Female	Overall (%)
Dolicocephalic	2	4	6(13.3)
Mesocephalic	5	13	18(40)
Brachycephalic	4	9	13(28.9)
Hyperbrachycephalic	3	5	8(17.8)

Table 2

Head shape	Age group		
	Upto 4 yrs	5-8 yrs	>8 yrs
Dolicocephalic	1	1	4
Mesocephalic	nil	5	13
Brachycephalic	2	6	5
Hyperbrachycephalic	nil	4	4

Table 3

Head shape	Age of initiation of Blood Transfusion		
	Upto 4 yrs	5-8 yrs	>8 yrs
Dolicocephalic	6	nil	nil
Mesocephalic	16	2	nil
Brachycephalic	12	1	nil
Hyperbrachycephalic	6	2	nil

Table 4

Age group(in yrs)	Mean CI		
	EB thal	Control	p value
Upto 4	78.7	72.9	p=0.48
5-8	81.2	84.1	p=0.11
9-12	78.9	81.7	P=0.09

Discussion:

The present study shows that commonest head shape in children with EB thal from our centre was mesocephaly. Since there is a dearth of Indian studies which analyse this perspective , numerous other studies [8,9,10,11,12,13,14] have shown mesocephaly to be the most common head shape in healthy Indian adults.Craniofacial measurements and assessments especially within the early years of life not only help in physical examination of the child, but also are helpful in analysing the development of the child. They are also useful in evaluation of craniofacial defects[15]. Thus this information may significantly contribute towards management of craniofacial anomalies as well as a quick indicator of brain and cranial growth in children. In a study by Karakas et al [16], no significant difference in head width was found between beta thalassemia major and those of control group. This

is in keeping with our study where no significant difference was found between cases and controls across the age groups studied. In a study on beta thalassemia major from the Indian population by Samba et al [17] ,though cephalometric measurements were made of maxilla and mandible, cephalic index was not assessed. The study by Girinath P et al[18], has evaluated orofacial manifestations in 50 thalassemic patients. Though bossing of the skull was noted to be prominent feature, cephalic index was not discussed.

Pathogenesis of bone changes in beta thalassemia major have been explained through multiple mechanisms like impaired osteoblast activity, abnormal osteoclast activity, hormonal abnormalities, iron overload and miscellaneous factors like nutritional deficits , in particular , vitamin C deficiency(19).Whether these same factors apply to patients of EB thal is not clear. In this aspect , the present study may contribute to understanding of craniofacial anomalies in children with EB thal.

Conclusion:

In this study from a tertiary care centre in eastern India, among children with E B thal, mesocephaly was found to be the commonest head type. No significant difference was found between cases and controls across the age groups studied. Several studies have dealt with craniofacial measurements, but though EB thal has common prevalence in eastern India, this study is first of its kind from this region in the country. However, a larger study is needed to corroborate the present findings.

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