ASSOCIATION OF OROFACIAL FEATURES OF β-THALASSEMIA MAJOR PATIENTS

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ABSTRACT

BACKGROUND

Patients with β-thalassemia major or Cooley's anaemia will have severe anaemia, which triggers several defence mechanisms in the body, the most significant of which is the expansion of bone marrow. This expansion of bone marrow in turn forces the bones to expand, produces deformities of the skull and face bones.

The purpose of this study was to find out the relationship between different orofacial parameters of labio-oral region in patients with β-thalassemia major and to identify differences by comparing them with age and sex matched controls.

MATERIALS AND METHODS

A case-control study was carried out in 400 study participants (200 cases and 200 controls). Mouth breadth (ch-ch), Height of integumental lips (ls-li) and Height of the total upper lip (sn-sto) of all the study participants were measured.

RESULTS

All the measurements used in the present study except height of integumental lips were found significantly associated in sex-wise subgroups as well as in overall study group.

CONCLUSION

The result of the present study increased our knowledge about orofacial anatomy of β-thalassemia major patients and enabled us to achieve quantitative results.

KEY WORDS

Cooley's Anaemia, Mouth Breadth, Labio-Oral, Craniofacial, Height of Upper Lip.

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BACKGROUND

Patients with β-thalassemia major or Cooley's anaemia will have severe anaemia and they live only for a short period without blood transfusion. The severe anaemia triggers several defence mechanisms in the body, the most significant of which is the expansion of bone marrow to about 30 times to produce more cells. This bone marrow hyperplasia produces frontal bossing and prominent malar eminences, which are seen on the skull x-ray as 'hair on end' appearance. Haemoglobin with absent β-chains and only with soluble alpha chains is toxic to the erythroblasts resulting in their intramedullary destruction, which causes ineffective bone marrow expansion by the release of erythropoietin in response to anaemia. This hyperplasia/ expansion of bone marrow in turn forces the bones to expand, produces deformities of the skull and face, and protrusion of upper tooth. These deformations of craniofacial bones changes hard as well as soft tissue profiles of the patients suffering from β-thalassemia major and gives classical 'chipmunk facies' or a 'rodent' like face appearance.[1] The features of chipmunk facies are characterised by frontal bossing, depressed nasal bridge, maxillary hypertrophy (most common) with protrusion of the upper incisors.[2-4]

There are only a few studies available on association of thalassemia on anthropometric measurements of face and head (Craniofacial anthropometry). Moreover, increasing prevalence of β-thalassemia major and concerns of researchers for abnormal anthropometric form of maxillary region in few previous studies led to this study to find out relationship between different orofacial parameters of labio-oral region in patients with β-thalassemia major and to identify differences by comparing them with age and sex matched controls.

MATERIALS AND METHODS

This case-control study was carried out in 400 study participants (200 cases and 200 controls) in Department of Anatomy, RNT Medical College and attached group of hospitals, Udaipur (Rajasthan) after getting approval from the Institutional Ethical Committee and Institutional Research Board. 200 haemoglobin electrophoresis confirmed cases of β-thalassemia major were randomly selected from thalassemia ward of Paediatric Department of MB Government Hospital, Udaipur receiving regular blood transfusion were selected as cases and their Age and Sex matched 200 controls were selected from school going children and undergraduate medical students of Rabindranath Tagore Medical College, Udaipur who were having normal haemoglobin level and willing to participate and cooperate in the study. Controls with any major illness and history of craniofacial abnormalities and facial surgery were excluded from the study.

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Sample size was calculated at 80% study power and α-error of 0.05 assuming standard deviation of 2.8 centimetres in head circumference of β-thalassemia major patients as found in study of Karakas et al (Eurasian J Med 2016; 48: 204-8). For minimum detectable difference of 0.8 centimetres in mean head circumference of cases and controls, 194 participants in each group were required as sample size. It was rounded off to 200 participants in each group as final sample size for present study.

All the eligible study participants were approached by the investigator herself. Nature and purpose of the study was explained. After obtaining their informed and written consent, their socio-demographic data and detailed history was taken. Blood for haemoglobin level was collected and sent to a designated laboratory for examination.

Orofacial measurements (labio-oral region) of both the study groups were measured by using standard landmarks, instruments (sliding caliper) and techniques by investigator personally (Figure 1). These measurements were as follows: Mouth breadth or lip length (ch-ch), Height of integumental lips (ls-li) and Height of the total upper lip (Sn-Sto) (Table 1). Mouth breadth or lip length (ch-ch) was measured by the straight distance between two chelion (ch), ensuring that the facial muscles of the subject were relaxed, and the jaws closed. Height of integumental lips (ls-li) was measured by the straight distance between labrale superior (ls) and labrale inferior (li). The subject was asked to keep his mouth closed and the arms of the sliding caliper were placed in such a manner that they cut the margins of the integumental lips tangentially and that the caliper was vertical. Height of the total upper lip (sn-sto) was measured by the straight distance between subnasale (sn) and stomion (sto) (Farkas method).[5]

Accordingly, desired data of each and every study subject was taken and recorded on separate sheet of prescribed proforma and was subjected to statistical analysis for achieving the purpose of the study.

Statistical Analysis

Shapiro-Wilk test was used to ensure normal distribution of data. Unpaired 't' test was used to compare variables. 'P' value < 0.05 was taken as significant. SPSS 22 version software was used for all statistical calculations.

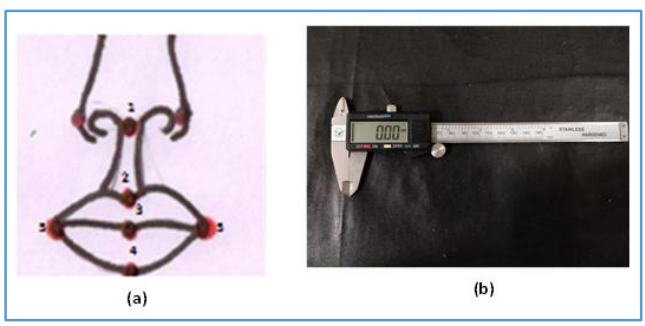

Figure 1. (a) Showing Landmarks used for taking Measurements (b) Sliding Caliper

RESULTS

Sl. No.	Landmark	Definition	Landmark in Fig.
1	Subnasale (sn)	It is the point where the lower margin of the nasal septum meets the integument of upper lip. This point should be sought where the tangent drawn to the nasal septum meets the upper lip.	1
2	Labrale superior (ls)	It is the point in the mid-sagittal plane cut by a tangent drawn at the highest elevation of the upper margin of the integumental lip.	2
3	Labrale inferior (li)	It is the point on the lower margin of the lower lip in the mid-sagittal plane.	4
4	Stomion (sto)	It is the point where the slit of the mouth with close lips cut the mid-sagittal plane.	3
5	Chelion (ch)	It is the point on the mouth-opening where the lateral margins of the upper and lower lips meet, i.e. corners of the mouth.	5

Table 1. Showing various Landmarks used in the Present Study

Measurement (mm)	Group	N	Mean	SD	'P' Value*
Mouth breadth (ch-ch)	Case	200	39.86	5.53	<0.001
	Control	200	42.27	5.97	
Height of integumental lips (ls-li)	Case	200	17.49	2.91	0.106
	Control	200	17.01	2.98	
Height of total upper lip (sn-sto)	Case	200	20.25	3.04	<0.001
	Control	200	18.65	2.48	

Table 2. Comparison of Orofacial Measurements (Labio-Oral Region) between Cases and Controls

*Unpaired 't' test.

Measurement (mm)	Group	N	Mean	SD	'P' Value*
Mouth breadth (ch-ch)	Case	107	40.19	5.51	0.009
	Control	90	42.41	6.37	
Height of integumental lips (ls-li)	Case	107	17.41	2.73	0.192
	Control	90	16.88	2.88	
Height of total upper lip (sn-sto)	Case	107	20.79	3.17	<0.001
	Control	90	19.01	2.68	

Table 3. Comparison of Orofacial Measurements (Labio-Oral Region) between Male Cases and Male Controls

*Unpaired 't' test.

Measurement (mm)	Group	N	Mean	SD	'P' Value*
Mouth breadth (ch-ch)	Case	93	39.48	5.55	<0.001
	Control	110	42.15	5.64	
Height of integumental lips (ls-li)	Case	93	17.58	3.11	0.287
	Control	110	17.11	3.06	
Height of total upper lip (sn-sto)	Case	93	19.63	2.78	<0.001
	Control	110	18.35	2.27	

Table 4. Comparison of Orofacial Measurements (Labio-Oral Region) between Female Cases and Female Controls

*Unpaired 't' test.

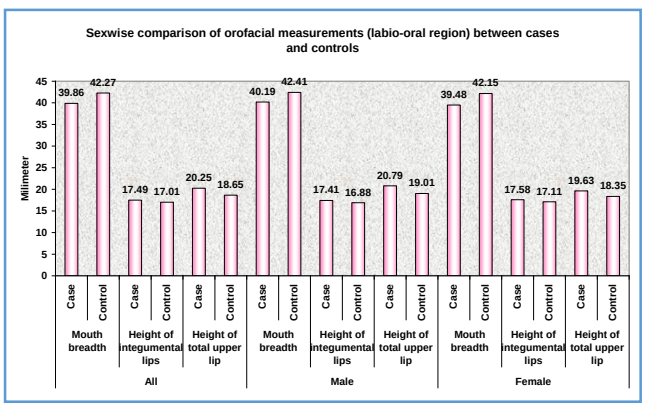

Figure 2. Sex-Wise comparisons of Orofacial Measurements (Labio-Oral Region) between Cases and Controls

DISCUSSION

Cases and controls were found comparable. Mean age of cases and controls was 9.8 years and 10.28 years respectively and was not statistically significant ('p' value= 0.296). There were 53.5% and 45.0% males among cases and controls respectively ('p' value= 0.110).

Mean mouth breadth (ch-ch) of cases was 39.86 mm and it was found significantly lower than controls in this study. (Table 2). When comparisons were done separately for male and female participants, similar observations were made there also (Table 3 and 4) (Figure 2). Similarly, Karakas S et al also found significantly lower mouth breadth in cases of their study.[1] However, Naimi AJ et al did not observe significantly lower mouth breadth, but their study was conducted at Iran and racial/ geographical variations in characteristics of participants may be the possible reason for this difference.[6]

Mean height of integumental lips (ls-li) of cases was 17.49 mm and it was found larger than controls, but the difference was negligible and not statistically significant (Table 2). When comparisons were done separately for male and female participants, similar observations were made there also (Table 3 and 4) (Figure 2). This parameter could not be discussed here, because in previous studies height of integumental lips was not analysed.

Mean height of total upper lip (sn-sto) of cases was 20.25 mm and it was found significantly larger than controls in the study (Table 2). When comparisons were done separately for male and female participants, similar observations were made there also (Table 3 and 4) (Figure 2). Similarly, Naimi AJ et al also found significantly larger mouth breadth in cases.[6] This finding of present study also commensurate with the study of Karakas S et al who concluded significantly larger total upper lip height (sn-sto) in their study.[1]

CONCLUSION

The present study concluded that all measurements used in present study except height of integumental lips were found significantly associated in sex-wise subgroups as well as in overall study group. The result of the present study increased our knowledge about orofacial anatomy of β-thalassemia major patients and enabled us to achieve quantitative results. These quantitative or numerical changes in the orofacial surface anatomy of β-thalassemia patients can also be useful for clinicians to identify deformity as well as for oral-maxillofacial surgeons during surgical interventions.

Limitations

Possibility of selection bias cannot be ruled out as cases were selected from Govt. hospital. Scarcity of literature was limitation for discussion of findings.

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