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ORIGINAL ARTICLE

Dentofacial and Cranial Changes in Down Syndrome

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Abstract
Objectives: This study aimed to determine the prevalence of certain oral characteristics usually associated with Down syndrome and to determine the oral health status of these patients.
Methods: The cross-sectional study was conducted among patients attending a special education program at Faculty of Dentistry, Jamia Millia Islamia, Delhi, India. The study design consisted of closed-ended questions on demographic characteristics (age, sex, and education and income of parents), dietary habits, and oral hygiene habits. Clinical examination included assessment of oral hygiene according to Simplified Oral Hygiene Index (OHI-S), dental caries according to decayed, missing, and filled teeth (DMFT) index, periodontal status according to the Community Periodontal Index of Treatment Needs (CPITN), and malocclusion according to Angles classification of malocclusion. Examinations were carried out using a using a CPI probe and a mouth mirror in accordance with World Health Organization criteria and methods. Craniometric measurements, including maximum head length and head breadth were measured for each participant using Martin spreading calipers centered on standard anthropological methods.
Results: The majority of the patients were males ($n = 63$; 82%) with age ranging from 6–40 years. The Intelligence Quotient (IQ) score of the patients indicated that 31% had moderate mental disability and 52% had mild mental disability. 22% exhibited hearing and speech problems.12% had missing teeth and 15% had retained deciduous teeth in adult population. The overall prevalence of dental caries in the study population was 78%. DMFT, CPITN and OHI scores of the study group were 3.8 ± 2.52, 2.10 ± 1.14 and 1.92 ± 0.63 respectively. The vast majority of patients required treatment (90%), primarily of scaling, root planing, and oral hygiene education. 16% of patients reported CPITN scores of 4 (deep pockets) requiring complex periodontal care. The prevalence of malocclusion

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was 97% predominantly of Class III malocclusions. Further 14% presented with fractured anterior teeth primarily central incisor. The percentage means of cephalic index was 84.6% in the study population. The brachycephalic and hyperbrachycephalic type of head shape was dominant in the Down syndrome individuals (90%).
Conclusion: The most common dentofacial anomaly seen in these individuals was fissured tongue followed by macroglossia.

1. Introduction

Down syndrome (DS) is a genetic disorder produced by the (complete or partial) presence of three copies of chromosome 21 [1−4]. The syndrome is characterized by a distinctive and immediately recognizable craniofacial phenotype [5,6]. The peculiar aspect of these subjects is partly a result of developmental anomalies of the craniofacial skeleton [4,6]. Many published studies have reported relatively poor dental health practices, relatively poor oral hygiene, and high levels of periodontal disease in children with Down syndrome than in normal children [7−9]. It has been reported that individuals with Down syndrome consistently show higher prevalence of periodontitis compared with that of other patients with mental retardation [10,11].

A search of the literature reveals that a large number of studies indicate that certain oral findings are concomitant with Down syndrome. Some of these findings are centered on clinical observations and some are on studies with a small number of patients. Some of the studies contradict and some support previous findings. Therefore, the purpose of this study was to determine the prevalence of certain oral characteristics usually associated with Down syndrome and to determine the oral health status of these patients.

2. Material and methods

The cross-sectional study was conducted among patients attending a special education program at Faculty of Dentistry, Jamia Millia Islamia, Delhi, India. The study protocol was approved by the Institutional Review Board prior to the start of the study**.** Patients were included in the study if they had parental consent/proxy consent, were present on the day of examination, and were willing to participate. Children were excluded from the study if they were uncooperative or had medical conditions, which contraindicated an oral examination without appropriate modifications, such as infective endocarditis, coagulopathy, abscess, etc. Informed consent was obtained from their guardian by whom they were accompanied. The intelligence quotient (IQ) of these children in these schools ranged between 20−80. This IQ had been determined prior to placing the children in schools by educational diagnosticians involved in the assessment of mentally handicapped children.

The study design consisted of closed-ended questions on demographic characteristics (age, sex, and education and income of parents), dietary habits, and oral hygiene habits. Clinical examination included assessment of oral hygiene according to Simplified Oral Hygiene Index (OHI-S) [12], dental caries according to decayed, missing, and filled teeth (DMFT) index [13], periodontal status according to the Community Periodontal Index of Treatment Needs (CPITN) [14], and malocclusion according to Angles classification of malocclusion [15]. Examinations were carried out using a using a Community Periodontal Index (CPI) probe and a mouth mirror in accordance with World Health Organization criteria and methods [16]. Craniometric measurements, including maximum head length and head breadth were measured for each participant using Martin spreading calipers centered on standard anthropological methods. The craniometric measurements were taken according to the technique defined by Kalia et al [11]. The head length was measured as the straight distance from the opisthocranion to the glabella and the head width was measured as the distance between the two most lateral points of the skull above the level of the supramastoid crest at right angles to the median sagittal plane. Subsequently, the cephalic index was calculated using the formula: head breadth/head length $\times$ 100. All the examinations were carried out by two dentists; however, throughout the examinations, every 10^{th} child was reexamined independently by each examiner to test for possible intra- and interexaminer variation, which was $< 5\%$ for each of the studied variables. Recording procedures were carried out according to the criteria described by WHO [13].

2.1. Statistical analysis

The Chi-square test was used to compare between categorical variables. Independent sample t tests and Z-tests were performed for comparison of means between two groups for quantitative variables, with $p < 0.05$ indicating statistical significance. Statistical analysis of the data was done using SPSS version 11 (SPSS Inc., Chicago, IL, USA).

3. Results

Of 94 individuals selected for the study, 77 patients could be examined. The rest of the patients did not cooperate for an oral examination, which gave a response rate of 82%. The demographic profile of the study

Table 1. Demographic characteristics of study population.

		Number of patients (%)
Age (y)	1−10	7
	11−20	55
	21−30	15
Sex	Male	70
	Female	7
Diet	Vegetarian	14
	Mixed	63
IQ score	Mild (50−70)	40
	Moderate (35−49)	24
	Severe (20−34)	13
Family history	Present	8
	Absent	69
Dentition	Permanent	61
	Mixed	16
	Tongue thrusting	18
	Mouth breathing	8
Brushing frequency	Once daily	54
	Two or more times/d	23
Mode of cleaning teeth	Toothpaste	70
	Toothpowder	7
Material used for cleaning teeth	Toothbrush	71
	Finger	6

IQ = intelligence quotient.

population revealed that the majority of the patients were males ($n = 63$; 82%) with age ranging from 6 years to 40 years (Table 1). The IQ score of the patients indicated that: 31% had moderate mental disability and 52% had mild mental disability; 22% exhibited hearing and speech problems; 12% had missing teeth; and 15% had retained deciduous teeth as adults. Prevalence of dentofacial abnormalities are presented in Table 2. The overall prevalence of dental caries in the study population was 78%. DMFT, CPITN, and OHI scores of the study group were 3.8 ± 2.52, 2.10 ± 1.14, and 1.92 ± 0.63, respectively (Table 3). Most patients (90%) required treatment, primarily scaling, root planing, and oral hygiene education. Sixteen percent of patients reported CPITN scores of 4 (deep pockets), requiring complex periodontal care (Table 4). The prevalence of malocclusion was 97%, predominantly of Class III malocclusions (Table 1). In addition, 14% presented with fractured anterior teeth, primarily the central incisor. The percentage means of cephalic index was 84.6% in the study population (Table 5). The brachycephalic and hyperbrachycephalic type of head shape was dominant in individuals with Down syndrome (90%; Table 6)

Table 2. Distribution of dentofacial abnormalities among patients with Down syndrome patients.

Abnormalities		Number of patients (%)
Fissured tongue		52 (67.5)
Macroglossia		45 (58.4)
Ankyloglossia		10 (13)
Angular cheilitis		17 (22.1)
High arched palate		65 (84.4)
Delayed eruption		10 (13)
Microdontia		35 (45.5)
Lack of lip seal		40 (51.9)
Fractured teeth		11 (14.3)
Malocclusion	Class 1	42 (54.5)
	Class 2	2 (2.6)
	Class 3	33 (42.9)
Crossbite		26 (33.8)
Open bite		15 (19.5)
Crowding of anterior teeth		18 (23.4)
Retained deciduous teeth		8 (10.4)
Congenitally missing teeth		26 (33.8)

Table 3. Distribution of mean decayed, missing, and filled teeth (DMFT) index/DMFT and Community Periodontal Index of Treatment Needs scores in patients with Down syndrome.

Variables	Downs syndrome (Mean ± SD)
DMFT scores	3.8 ± 2.52
CPITN scores	2.10 ± 1.14
OHI score	1.92 ± 0.63

CPITN = Community Periodontal Index of Treatment Needs; DMFT = decayed, missing, and filled teeth; OHI = Oral Hygiene Index; SD = standard deviation.

4. Discussion

In the current study, the most common dentofacial anomaly seen in these individuals was fissured tongue followed by macroglossia which is consistent with previous studies [10,17]. Most of the patients with fissured tongue presented with multiple fissures and various fissural patterns on the dorsal surface of the anterior two thirds of the tongue. The cause of fissure tongue is possibly

Table 4. Distribution of Community Periodontal Index of Treatment Needs scores by type of disability.

CPITN scores	Down syndrome (%)
0 (Healthy)	8 (10.4)
1 (Bleeding)	6 (7.8)
2 (Calculus)	36 (46.8)
3 (Shallow pockets)	15 (19.5)
4 (Deep pockets)	12 (15.6)
Total	30
TN0 (No need for treatment)	8 (10.4)
TN1 (Oral hygiene instruction)	6 (7.8)
TN2 (Prophylaxis)	51 (66.2)
TN3 (Complex treatment)	12 (15.6)

CPITN = Community Periodontal Index of Treatment Needs.

Table 5. Cranial values in patients with Down syndrome.

Variables	Mean ± standard deviation
Head breadth	14.31 ± 0.91
Head length	16.91 ± 0.86
Cephalic index (%)	84.6

developmental [18] and is not associated with sex. Macroglossia in Down syndrome patients could be attributed to inadequate lymphatic drainage [19]. The dorsal surface of the tongue in most cases was dry because of mouth breathing. A few patients also presented with scalloped margins and imprints of teeth due to abnormal pressure of the enlarged tongue on the teeth. The protruding tongue can possibly lead to speech problems, which are common in these patients. Angular cheilitis and lack of lip seal was also observed, which could conceivably result from hypotonia of the orbicularis, zygomatic, masseter, and temporalis muscles reported in Down syndrome [20]. The hypotonic upper lip and lower lip and large tongue in a reduced oral cavity may lead to mouth breathing, drooling, and angular cheilitis [21]. The high prevalence of high arched palate in this study could be due to midface hypoplasia resulting in a reduction of the length, height, and depth of the palate [21].

Congenitally missing teeth are seen in 34% of the study group, which is in agreement with reports in the literature [22]. Genetic modes of transmission reportedly is the cause of increased prevalence of oligodontia and in Down syndrome. Research in this area has revealed that this "trisomic insult" could greatly increase the host susceptibility to this anomaly while not disturbing specific tooth buds [23]. The most frequently missing teeth were third molars followed by second premolars and incisors. A few patients also presented with retained primary teeth as adults. Of the patients with Down syndrome, 46% presented with microdontia, a finding concurrent with that of Spitzer et al and Kissling et al who reported that all teeth, except the upper first molars and lower incisors, were reduced in size, but that the root formation was always complete [22].

In India, only 20−36% of children in the general population have been found to have a definitive malocclusion [24], the individuals with Down syndrome showed 93% incidence of definitive malocclusion primarily of Angle Class III malocclusion. Our results are in strong agreement with previous studies reporting an increase in Class III malocclusion coexistent with a reduction of Class II cases in these patients compared to controls [24−27]. This could be attributed to altered cranial−base relationships, decreased arch length, reduced dental arch size, and diminished maxillary size in patients with Down syndrome [9,28−30].

Table 6. Distribution of head shapes in patients with Down syndrome.

	Number of patients (%)	*p*
Dolicocephalic (< 74.9)	6 (7.8)	**< 0.05***
Mesocephalic (75−79.9)	2 (2.6)	
Brachycephalic (80−84.9)	48 (62.3)	
Hyperbrachycephalic (85−89.9)	21 (27.3)	

*$p < 0.05$ is considered significant.

Another interesting finding in the current study was that patients with Down syndrome had a higher incidence of tooth fractures predominantly affecting maxillary incisors in comparison with the general population than in the general population in India [31]. This is consistent with the findings of the other studies [32−34],which also suggest that higher frequency of injuries to the maxillary incisors could be due to the higher frequency of extreme maxillary overjet, Angle Class II Division I malocclusion, short or incompetent upper lip, and accident-proneness of children with disabilities [35].

Furthermore, the current study states that the mean cephalic index of the study group is 84.6%, thus classifying the patients with Down syndrome as brachycephalic, which confirms the stigmata of Down syndrome reported in the literature. The principal stigmata of DS comprises overall reduction in head size and brachycephaly with a flattened occipital bone resulting from developmental anomalies of the craniofacial skeleton [4,6,17,36,37].

A review of the literature shows that there has been a disparity regarding the caries susceptibility in individuals with Down syndrome [9,38−41]. Our results suggest that the prevalence of dental caries in Down syndrome was higher than that found in the general population [42,43].These findings were in agreement with previous studies reporting 78−90% prevalence of dental caries in Down syndrome [42]. Increased dental caries in these individuals could be due to muscle weakness and inadequate muscular coordination interfering with daily hygiene procedures.

In addition, patients with Down syndrome reported poorer oral hygiene and periodontal status than that reported in the general population, thus agreeing with findings of previous studies [10,19,20]. Furthermore, a significant proportion of these patients (23%) had deep pockets, requiring complex periodontal surgical care. These results are consistent with previous studies [12,43,44] that reported high a prevalence of periodontal disease in Down syndrome. Marginal gingivitis was seen in patients as young as 12 years. Patients also presented with advanced periodontitis, gingival recession, horizontal and vertical bone loss with suppuration, bifurcation involvement in the molar area, and marked mobility of posterior and anterior teeth. These results could be due to the low physical capabilities of these individuals, limited understanding on the

importance of oral health management [38,45], difficulties in communicating oral health needs [23], and dependence on other people such as parents or employees with assisted living services [24]. An oral health promotion program should be started as early as age 6 months but no later than age 18 months. These intervention programs should be targeted to special-needs schools and parents and guardians of patients with Down syndrome.

The current study explores the prevalence of various dentofacial anomalies and oral health status in Down syndrome that may require medical consultation. However, given the rising number of patients with Down syndrome living in the community, the assessment of the features of these patients may be of help to clinicians and basic researchers.

Conflicts of interest

The authors declare they have no conflicts of interest related to the studies performed.

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