A Cephalometric Evaluation of Midface and Mandibular Effective Lengths and the Maxillomandibular Differentials in Children with Down Syndrome in Benin City, Nigeria.

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ABSTRACT

Objective: To determine the effective lengths of the midface, mandible and the maxillomandibular differentials of individuals with Down syndrome and compare with the standard average McNamara norms

Methods: Twenty-two children with Down syndrome (10-15 years; 13 males and 9 females) were recruited for the study. The effective lengths of the midface (MxTL) and mandible (MnTL), maxillomandibular differentials (MxMnD) and the anterior cranial base (S-N) length were evaluated on the lateral cephalograph. Independent t-test was used to comparatively evaluate the continuous variables in relation to gender. Pearson correlation coefficient was used to determine the relationship of the variables. A significant level of $p<0.05$ was set for this study.

Results: The average effective length of the midface (MxTL) and mandible (MnTL) were 68.75±6.73mm and 89.18±10.63mm respectively. The maxillomandibular differential (MxMnD) was 20.32±5.93mm while the average anterior cranial base (S-N) length was 53.14±5.16mm. Male patients with Down syndrome had a larger MxTL and MnTL, MxMnD and S-N length than females ($p<0.05$). Pearson correlation coefficient (r) showed a statistically significant level ($p=0.0001$) of strong correlation between the S-N length and the MxTL ($r=0.832$) and MnTL ($r=0.929$). Linear regression showed the relationship and impact of S-N on the variables as follows: on MxTL, ($r=0.831$, $r^2=0.690$), MnTL ($r=0.923$, $r^2=0.862$), and MxMnD. ($r=0.740$, $r^2=0.548$).

Conclusion: This study, therefore, shows that the average effective lengths of the midface and mandible were shorter than the McNamara values for young non-Down syndrome children while the maxillomandibular differentials were within normal values as described by McNamara. The reduction in the effective lengths of the midface and mandible could be a contributing factor to development of malocclusion among individuals with Down syndrome.

Keywords: Down syndrome, midface, mandible, maxillomandibular differentials

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INTRODUCTION

Down syndrome phenotypic characteristics were first described by John Langdon Down but in 1959, Jerome Lejeune, a French physician discovered the chromosomal abnormality associated with this group of individuals.[1] Trisomy 21 accounts for 94% of the aetiology of Down syndrome while translocation and mosaic account for the other causes of Down syndrome.[2]

Certain facial features are common among individuals with Down syndrome which include: flattened face, mid-face depression and reduced skull size. These features have been attributed to the significant hypoplasia in the endochondral, mesodermal, and ectomesenchymal derived structures of the cranium and face of individuals with Down syndrome.[3]

Malocclusion is an appreciable deviation from ideal occlusion which is functionally and aesthetically unpleasant.[4] Individuals with Down syndrome have a high prevalence of malocclusion with large deviation in occlusal relationship.[5,6] Mouth breathing, characteristic tongue thrusting, delayed eruption and/or exfoliation of both the primary and permanent dentition, brachycephaly have been attributed to the development of malocclusion among individuals with Down syndrome.[7,8,9]

Though there seems to be an agreement regarding a reduction in the dimension of the cranial base and the mid-face length among individuals with Down syndrome,[10] Spitzer et al[11] and some other authors[12-13] have reported a reduction in the effective length of the mandible while Fischer-Brandies[14] and Quintanilla et al[15] have observed no difference in the length of the mandible of individuals with Down syndrome when compared with individuals without Down syndrome.[14,15] Maxillary deficiency has been reported not to be so expressive in the face of individuals with Down syndrome when compared to individuals without Down syndrome with mid-face deficiency because of the overall reduction in craniofacial dimensions.[13] Premature fusion of the cranial sutures due to mutations in fibroblast growth factor receptors and the transcription factor MSX2 associated with individuals with Down syndrome is responsible for the reduced maxillary length with midface retrusion, a small cranial base and an increased cranial base angle.[14-16]

Moyers reported that the cranial base plays a significant role in the positioning of the midface and the lower jaw.[17] Individuals with Down syndrome generally have a defective cranial base development resulting in reduced cranial base length and broad cranial base angle.[3,18] A reduced cranial base length not only affects its sagittal positions with the jaws but also affects the growth potential of the jaws themselves.[19]

Evaluation of the sagittal jaw relationship using the ANB angle has shown that individuals with Down syndrome have the tendency of developing class III jaw relationship[3,13,20] when compared to normal individuals. However, the study conducted by Clarkson et al.[21] did not show a significant difference in the ANB angle among individuals with Down syndrome and individuals without Down syndrome. The relationship of the lower jaw to the cranial base was also observed not to be significantly different from normal control even though it was larger among the individuals with Down syndrome.[10]

The use of maxillomandibular differential (MMD) to determine the sagittal relationship of the lower and upper jaws was first proposed by McNamara.[22] An earlier study showed that individuals with Down syndrome have a comparable maxillomandibular differential with normal individuals.[13] Silva Jesuino and Valladares-Neto,[13] further observed an overall reduction of the cranio-facial dimension among individuals with Down syndrome, including the mandible.[13] Therefore, it is important to further assess the linear measurements of the jaws among individuals with Down syndrome and to determine if they contribute significantly to the development of malocclusion. If the jaws are not in correct sagittal position in relation to the cranial base, is it also possible that the dimensions of the jaws are reduced? This study was conducted with the aim of determining the effective lengths of the midface and mandible, and ultimately the maxillomandibular differentials in individuals with Down syndrome. Furthermore, their level of correlation to the anterior cranial base will be determined. The sagittal jaw relationship will also be determined using the maxilla-mandibular differential.[22]

Methods

Ethical approval (ADM/E 22/A/VOL VII/1236) for this research protocol was obtained from the hospital Ethic and Research Committee before data were collected. Written informed consents were obtained from the guardians of the individuals with Down syndrome before they were recruited for the study. Verbal assents were also obtained from study participants. This cross-sectional descriptive study was conducted among 22 individuals with Down syndrome aged 10 -15 years of age, comprising of 13

males and 9 females. The mean age of the study participants was 13.14 ± 1.78 years. It was conducted in the Orthodontic unit of the University of Benin Teaching Hospital. The study participants were recruited from schools for special need individuals within the Benin City metropolis and their lateral cephalographs taken at a private dental clinic.

Convenience sampling method was used to recruit the study participants due to peculiarity of the individuals and the need to recruit sizeable study participants. All the participants were objectively karyotyped and confirmed to be Trisomy 21 using cytogenetic analysis. The inclusion criteria included: those within the study age group, individuals for whom informed consent had been given and those confirmed via karyotyping. Individuals with Down syndrome with previous orthodontic treatment, difficult neck stability and those with distorted radiographs were excluded. The lateral cephalographs of the study individuals were manually traced on a matte acetate paper using a pointed HB pencil under a light box. For intra-investigator reliability, five lateral cephalographs were initially traced at two different sessions and in a two weeks interval with a kappa statistical value of 0.79.

The data were analyzed using the IBM SPSS version 20. The cephalometric measurements obtained from individuals with Down syndrome were analyzed using independent t-test. Pearson correlation coefficient was used to determine the association among the variables. The effect of the anterior cranial base (S-N) on the effective maxillary length (MxTL), effective mandibular length (MnTL) and the maxillomandibular differentials (MxMnD was conducted using the regression model analysis. A significant level of $p<0.05$ was set for this study

Description of landmarks (Figure 1)

ANS = Anterior Nasal Spine, the most anterior point of hard palate; intersection of anterior and upper maxilla part with nasal fosse floor

Go = The most posterior–inferior point at the angle of the mandible, located by bisecting the angle formed by the lines that are tangent to the posterior border of the ramus and the inferior border of the mandible

Gn = Ggnathion, the most anterior and inferior point on the body outline of the chin, situated at equidistance from Pog and Me

A-point = the deepest part of the curvature between the Anterior Nasal Spine (ANS) and the alveolar crest supporting the maxillary central incisor

Co = Condylion, the most upper and posterior point of mandibular condylar outline

PNS = Posterior Nasal Spine, the most posterior point of hard palate

S = Sella, the centre of sella turcica

N = Nasion, the most anterior point on the frontonasal suture

- Total mandibular length (MnTL), also called effective length of the mandible: the distance from Co – Gn
- Total maxillary length (MxTL), also called effective length of the midface: the distance from Co – A point
- Mandibular body length (MnBL): the distance from Go-Gn
- Maxillary body length (MxBL): the distance from PNS-A point
- Anterior cranial base (S-N): the linear distance from S-N points
- Maxillo-mandibular differentials (MxMnD): the difference between effective maxilla and mandibular length
- The values of the effective midface length, effective mandibular length and maxillomandibular differential will be compared with the original McNamara values for young children, as shown in Table 1
- Total posterior facial height: the vertical distance from the S point to the gonion

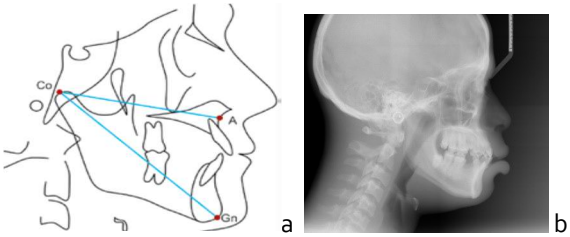

Figure 1. Schematics of the cephalometric landmarks used for the study and a cephalometric radiograph of a Down syndrome participant used in the study

Table 1: Original average mean values given by McNamara for young children

Variables	Effective midface length (mm)	Effective mandibular length(mm)	Maxilloman-dibular differentials (mm)
Values	85	105-108	20-23

Results

The study was conducted among 22 individuals with Down syndrome aged 10-15 years. It comprised of 13 males and 9 females. The mean age of the study participants was 13.14 ± 1.78 years.

The average mean value for the anterior cranial base length (S-N) was 53.14 ± 5.16 mm for the entire study participants. Effective midface (MxTL) and mandibular (MnTL) lengths were 68.75 ± 6.73 mm and 89.18 ± 10.63 mm respectively. The sagittal jaw relationship measure as the average maxillomandibular differential (MxMnD) was 20.32±5.93 mm, see Table 2. Table 3 shows that the anterior cranial base length (S-N), effective midface length (MxTL), mandibular lengths (MnTL) length, maxillary body length (MxBL) and maxilla-mandibular differentials (MxMnD) were lesser among the females compared to the males, p >0.05. However, female Down syndrome had larger mandibular body length (MnBL) and a higher posterior facial height (S-Go), p >0.05.

Pearson correlation coefficient shows a statistically significant level of strong correlation association between the anterior cranial base length and the effective midface and mandibular length. This shows that a reduction in the length of the anterior cranial base is accompanied by a significant reduction in the effective jaw lengths (MxTL, MnTL). There is also a strong positive and significant correlation between the anterior cranial length and the maxillomandibular differentials as shown in Table 4.

Although the maxillomandibular differentials had a positive correlation with the effective lengths of the maxilla and mandibular, the results, however, show weak correlation with the effective midface length but a strong correlation with the effective mandibular length, refer to Table 4. Figure 2 and 3 show the direction of Pearson correlation line of best fit.

Tables 5 shows the relationship and the impact the anterior cranial base (S-N) has on the effective maxillary length (MxTL), effective mandibular length (MnTL) and the maxillomandibular differentials (MxMnD using the regression model analysis. The results show a good correlation between S-N and MxTL (r=0.831), S-N and MnTL (r=0.923) and S-N and MxMnD (r=0.740). The anterior cranial base appears to explain more variation in the effective mandibular length (r^2=0.862) compare to the effective maxillary length (r^2=0.690). The significance of the coefficient shows that impact of S-N on MxTL, MnTL and MxMnD was statistically significant, p<0.001 for each. The result, however, rejects the null hypothesis that anterior cranial base does not have significant impact on the effective lengths of the midface, mandible and the maxillomandibular differentials, see Table 5.

Table 2: Distribution of the average mean values of the variables

VARIABLES	MEAN±SD	MIN	MAX
S-N (mm)	53.14±5.16	48.00	66.00
MxTL (mm)	68.75±6.73	59.00	83.00
MnTL (mm)	89.18±10.63	76.00	116.00
MxMnD (mm)	20.32±5.93	9.00	33.00
MxBL (mm)	36.61±4.08	30.00	48.00
MnBL (mm)	62.96±7.10	53.00	79.00
S-Go (mm)	44.11±7.36	33.00	64.00

Table 3: Distribution of the variables in relation to gender

Variables	Group	No	Mean	Mean difference	p value	95% CI	
						Lower	Upper
SN(mm)	Male	13	53.37±1.49	0.325	0.887	-4.362	5.011
	Female	9	52.94±1.64				
	Total	22					
MxTL(mm)	Males	13	69.00±1.89	0.611	0.840	–5.621	6.843
	Female	9	68.39±2.34				
	Total	22					
MnTL(mm)	Males	13	90.00±3.35	2.000	0.675	-7.811	11.811
	Female	9	88.00±2.91				
	Total	22					
MxBL(mm)	Males	13	37.04±1.16	1.039	0.570	-2.711	4.789

	Female	9	36.00±1.37				
	Total	22					
MnBL(mm)	Males	13	62.92±2.08	-0.769	0.981	-6.654	6.500
	Female	9	63.00±2.30				
	Total	22					
MxMnD(mm)	Males	13	21.04±1.59	1.761	0.507	-3.671	7.193
	Female	9	19.28±2.13				
	Total	22					
S-Go(mm)	Males	13	43.12±1.60	-2.440	0.458	-9.168	4.288
	Female	9	45.56±3.13				
	Total	22					

Table 4: Pearson correlation coefficient

Variables		MxTL	MnTL	MxBL	MnBL	MxMnD
S-N	R	0.832	0.929	0.633	0.805	0.740
	p-value	0.0001	0.0001	0.002	0.0001	0.0001
MxMnD	R	0.431	0.824			
	p-value	0.045	0.0001			

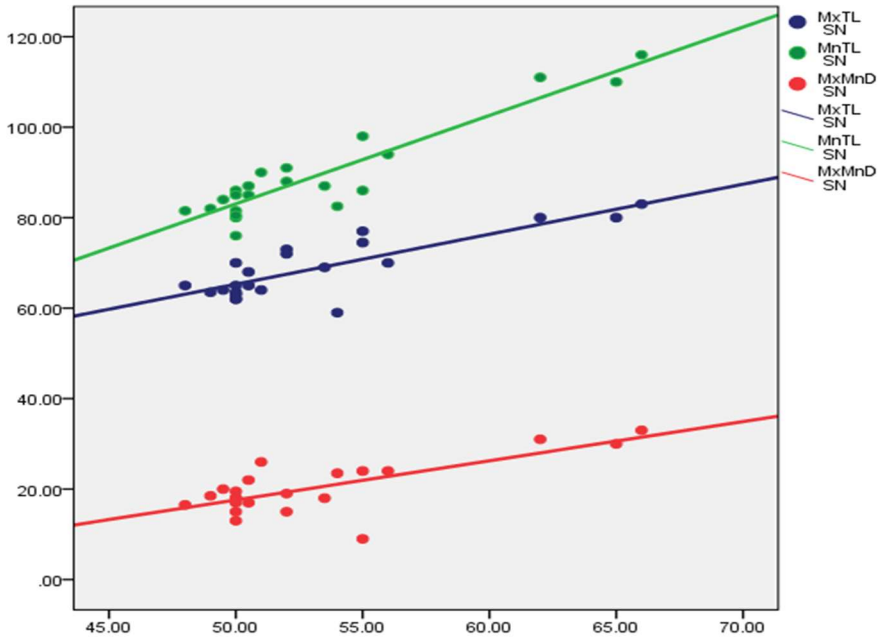

Figure 2. Directions of Pearson correlation (*r*) of the anterior cranial base length (S-N) with the effective mandibular length MnTL (green), effective midface length MxTL (blue) and the maxillomandibular differentials MxMnD (red).

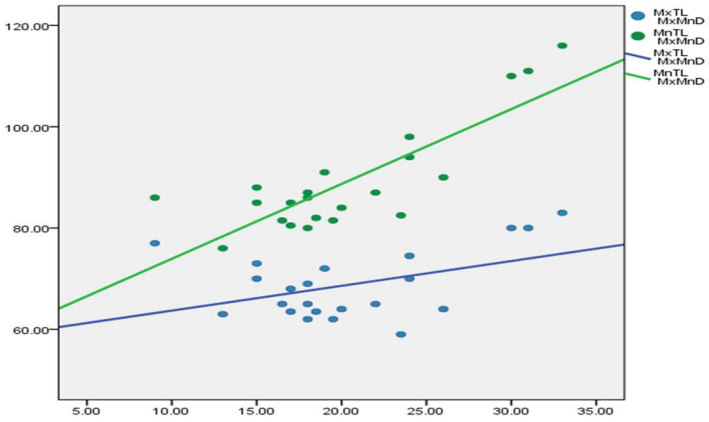

Figure 3. Directions of Pearson correlation (*r*) of the maxillomandibular differentials MxMnD, with the effective mandibular length MnTL (green) and effective midface length MxTL (blue).

Table 5: Linear regression model to determine the relation and impact of S-N on MxTL, MnTL and MxMnD.

Variables	R	R^2	Adjusted R^2	P value
MnTL	0.923	0.862	0.857	*0.000
MxTL	0.831	0.690	0.675	*0.000
MxMnD	0.740	0.548	525	*0.000

*: p value < 0.001 which indicates the level of significance of the impact of the regression coefficient of the main predictor (independent variable S-N) on the variable

Discussion

McNamara has suggested that there is a corresponding relationship of the effective length of the midface to the effective length of the mandible.[22] According to him, the geometric relationship of the midface and the mandible is more important than the age of the patient.[22] This geometric relationship is the difference in the effective length of the midface and the effective length of the mandible, otherwise referred to as the maxillomandibular differentials with normal range of values given. McNamara has proposed that for young individuals with a well-balanced face, the effective midface length should be around 85mm while the effective mandibular length would be between 105mm to 108mm with a maxillomandibular differential of 20-23mm.

It was observed in this study that individuals with Down syndrome have a reduction in the effective lengths of the midface and mandible when compared to the norms provided by McNamara for younger normal individuals. This study takes into consideration that several studies have shown that McNamara cephalometric norms can differ between racial groups.[23-25] Although the maxillomandibular differential in this study appeared to be within the normal range proposed by McNamara, this is possibly attributed to the overall reduction in the effective lengths.

Again, when comparison was made in relation to the age range of individuals with Down syndrome in this study, the effective lengths of the midface and mandible of individuals with Down syndrome in this study still appeared shorter compared to the Bolton standard and the values derived from the Burlington research centre for normal individuals.[22,23] A reduction in the length of the maxilla and mandible among individuals with Down syndrome has been previously documented.[3,24]

Despite the lower age range (6-12 years) in the McNamara norms developed for normal Egyptian children,[25] individuals with Down syndrome in this study were aged 10-15 years yet they expressed a lower effective length of the midface and mandible,

even though the maxillomandibular differential appeared minimal. The overall reduction in the effective lengths of the midface and mandible as observed in this study supports earlier observation made by Silva Jesuino and Valladares-Neto.[13] The implication of these findings is that there is a less obvious prognathic lower jaw due to the symmetric reduction in the effective length.

The reduction in the dimensions of the jaw and midface in individuals with Down syndrome is likely due to the significant hypoplasia in the structures of the cranium and face that are of endochondral, mesodermal, and ectomesenchymal origin.[3] The use of ANB angle for evaluating skeletal jaw relationship have also shown that individuals with Down syndrome have the tendency of developing class III jaw relationship. [3,13,20,24] The study conducted by Clarkson et al, however, did not show a significant difference in the ANB angle among individuals with Down syndrome and normal individuals.[21]

Controversy exists between authors whether the size of the mandible is normal or reduced among individuals with Down syndrome.[3,12-15,24] This study corroborates earlier findings that individuals with Down syndrome have a reduction in both the maxillary and mandibular effective lengths[3,12,13,24] but varies with observation made by other researchers[14,15] who observed no difference in the length of the mandible.

The pertinent question that needs clarification is that if there is an overall reduction in the dimension of the anterior cranial base and the midface/jaws, why do individuals with Down syndrome have the appearance of a "prognathic" mandible? The apparent prognathic appearance has been attributed to several factors which include anterior tongue position among individuals with Down syndrome which slides the lower jaw forward.[26] A more pronounced midface reduction also makes the mandible appear prognathic as suggested by Brown RH, Cunningham VM.[27]

This study did not show that the midface was more significantly reduced when compared to the mandible. When compared with the standard average McNamara values for young children, findings from this study shows that there is symmetric geometric reduction in both effective lengths of the midface and mandible. Fink et al suggested that the midface is more deficient than the mandible which gives the prognathic appearance,[12] but it is at variance with this study.

However, Roche AF[28] observed a normal growth rate of the midface and jaws among normal populations and individuals with Down syndrome.[28] The contour of the labial and lingual mandibular chin also gives a more prognathic appearance to the face of individuals with Down syndrome.[29]

The dimension of the anterior cranial base can influence the sagittal positioning of the maxilla and mandible.[19] This study shows that the reduction in the sagittal position of the jaws and midface of individuals with Down syndrome is accompanied by a similar reduction in the anterior cranial base length as evident by the positive Pearson correlation. Observations from this study also shows that the effect of the deficient anterior cranial base was more on the effective length of the midface than the effective length of the mandible. A shorter anterior cranial base among individuals with Down syndrome is related to the small brain tissue.[30] This study shows an overall reduction in the effective lengths of the midface and mandible with the maxillomandibular differentials appearing somewhat within normal range.

Study limitation

The dearth of information regarding the McNamara norms for Nigerian children limited the specific comparison with the individuals with Down syndrome in this study.

Conclusion

The findings from this study implies a geometric reduction in the effective length of the midface and mandible in the population studied.

The mean maxillomandibular differential of the population studied was within the normal value as described by McNamara, which can be attributed to the overall reduction in the dimension of the cranial structure, making the class III skeletal pattern less obvious.

This study also shows a strong positive correlation between the effective length of the midface, effective length of the mandible, the maxillomandibular differentials and the anterior cranial base.

Recommendation

A larger scale longitudinal study among individuals with Down syndrome is advocated to evaluate the serial growth pattern of the midface and mandible.

Also, there is need to develop McNamara norms for Nigerian children for appropriate comparison.

Sponsorship: Nil

Conflict of interest: None declared

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