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Uvulo-glosso-pharyngeal dimensions in subjects with β-thalassaemia major

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SUMMARY The purpose of this study was to investigate cephalometrically the uvulo-glosso-pharyngeal dimensions in subjects with β-thalassaemia major. The subjects were 15 thalassaemic patients (eight males, seven females) with a mean age of 14.13 ± 1.06 years. The variables measured on the lateral cephalograms were tongue height and length, soft palate length and thickness, superior, middle and inferior pharyngeal airway space, and hyoid bone position. The thalassaemic group was compared with a normal control group matched for sex and age using a *t*-test.

The results showed that thalassaemic patients had a smaller tongue size (length $P < 0.05$, height $P < 0.001$), shorter soft palate ($P < 0.001$), smaller upper ($P < 0.001$) and middle ($P < 0.05$) pharyngeal airway spaces, and a shorter vertical pharyngeal length ($P < 0.05$). The hyoid bone in thalassaemic patients was closer to the mandibular plane ($P < 0.001$).

Introduction

The thalassaemias are a group of inherited haemoglobinopathies caused by the defective synthesis of either the α or β polypeptide chains referred to as α- and β-thalassaemia, respectively (Weatherall and Clegg, 1981). A wide variety of genetic defects produce diverse clinical findings from mild morphological abnormalities (thalassaemia minor) to a life-threatening disease with severe clinical symptoms and marked orofacial defects (thalassaemia major).

β-thalassaemia major (also known as Cooly's anaemia) is transfusion-dependent and is commonly manifested during the first year of life. The affected infants are severely anaemic, highly susceptible to infection, and have growth retardation. The suggested causes of growth retardation include chronic anaemia (Caffey, 1957), hyperparathyroidism (Flynn *et al*., 1976), and somatomedian deficiency (Saenger *et al*., 1980).

Cephalometric radiography has been extensively used in orthodontics to study craniofacial morphology and to provide information about the soft and hard tissue of the upper airway (Taylor *et al*., 1996). The reliability of lateral cephalograms in measuring airway landmarks was evaluated by Miles *et al.* (1996), who found that the commonly used landmarks of the airway structures could be identified reliably, irrespective of the quality of the radiograph.

The typical craniofacial features of β-thalassaemia major are a Class II skeletal pattern, a shorter cranial base length, shorter mandible, and increased anterior face height (Bassimitci *et al*., 1996; Abu Alhaija *et al*., 2002). The dentition shows protrusion, flaring, and spacing of the maxillary anterior teeth (Kaplan *et al*., 1964; Weel *et al*., 1987; Cannell, 1988; Hes *et al*., 1990).

The prevalence of thalassaemia in Greek, Turkish, and Cypriot populations is 15 per cent, and up to 25 per cent in the people of Rhodos and Malta (Weatherall and Clegg, 1981). In Jordan, approximately 1000 transfusion-dependent thalassaemic patients are registered, with an annual increase of 80 cases and a carrier rate of 7–10 per cent of the population.

Although thalassaemia major is considered to be a common genetic disorder, there are no reports in the literature concerning the upper airway and soft tissue dimensions.

The aim of this study was to investigate the uvulo-glosso-pharyngeal dimensions of patients with thalassaemia major and to compare the measurements with a control group without any known systemic diseases or syndromes.

Materials and methods

The material comprised the lateral cephalograms of 15 thalassaemia major subjects (eight males and seven females, aged 12–16 years). The control group consisted of 15 subjects (eight males and seven females, aged 13–16 years) who had a lateral cephalogram taken for orthodontic assessment. All subjects had a Class II skeletal pattern (ANB° > 3) and were in the full dentition stage (all permanent teeth were erupted except the third molars). The mean age and sex distribution for the thalassaemic and control groups are shown in Table 1. Because of the limited sample size and insignificant differences between sexes, male and female measurements were pooled.

Cephalometric radiographs were taken with a Siemens Orthophos-5 machine using a standardized technique with the teeth in maximum intercuspation and a fixed anode–mid-sagittal plane distance. The magnification of the radiographic machine, which was not corrected, was 11.3. Lateral skull radiographs were traced on acetate paper, and 10 hard and soft tissue cephalometric points were registered yielding 12 linear measurements (Figure 1). The measurements were performed manually using a ruler to the nearest 0.1 mm.

Table 1 Mean age ± SD, and sex distribution in the thalassaemic and control groups.

Sex	Thalassaemic group		Control group	
	Mean	SD	Mean	SD
Male (n = 7)	14.88	0.64	14.75	0.89
Female (n = 8)	13.29	0.76	13.29	0.49
Males + females (n = 15)	14.13	1.06	14.06	1.03

Method error

Ten randomly selected films were retraced and measured and the method errors calculated as recommended by Dahlberg (1940) and Houston (1983). Dahlberg error varied from 0.27 mm for

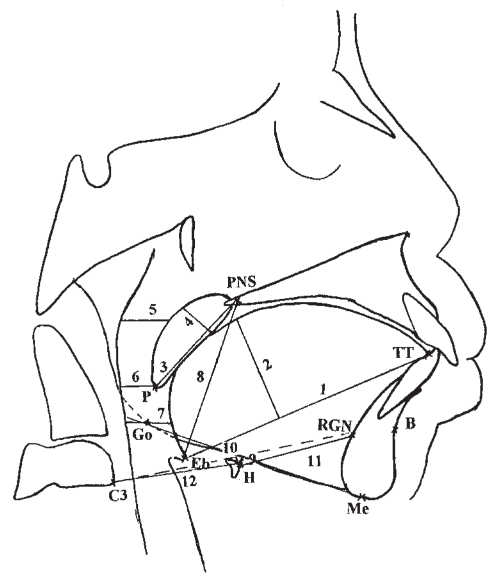

Figure 1 Cephalometric points, lines, and linear variables used in the cephalometric analysis. TT, tongue tip; Eb, base of epiglottis; P, tip of soft palate; PNS, posterior nasal spine; Me, menton; Go, gonion; B, point B; RGN, retrognathion; H, hyoidale; C3, antero-inferior limit of third cervical vertebra; Go–Me, mandibular plane; Go–B line 1, TGL, tongue length (Eb–TT); 2, TGH, tongue height (maximum height of tongue along perpendicular line of Eb–TT line to tongue dorsum); 3, soft palate length (PNS–P); 4, MPT, soft palate thickness (maximum thickness of soft palate measured on line perpendicular to PNS–P line); 5, SPAS, superior posterior airway space (width of airway behind soft palate along a line parallel to Go–B line); 6, MAS, middle airway space (width of airway along a line parallel to Go–B line through P); 7, IAS, inferior airway space (width of airway space along Go–B line); 8, VAL, vertical airway length (distance between PNS and Eb); 9, perpendicular distance from hyoid bone to mandibular plane; 10, HH1, perpendicular distance from hyoid bone to the line connecting C3 and RGN; 11, distance between hyoid bone and RGN; 12, C3H, distance between hyoid bone and C3.

IAS to 0.58 mm for PNS–Eb. Houston's coefficient of reliability ranged from 0.90 to 0.96 mm.

Statistical analysis

Descriptive statistics including the mean, standard deviation (SD), and difference between the means for each group were computed using SPSS PC+. The differences between the thalassaemic patients and controls were evaluated using the independent *t*-test.

Results

The results of the statistical analysis are shown in Table 2. Tongue dimension was smaller in the thalassaemic patients. Tongue length was 69.77 ± 5.95 mm in the thalassaemic subjects compared with 75.07 ± 5.75 mm in the control group. The difference was statistically significant ($P < 0.05$). Tongue height was 26.20 ± 2.60 mm in the thalassaemic patients and 29.97 ± 2.57 mm in the control group. The difference was significant ($P < 0.001$).

The thalassaemic patients showed a shorter soft palate (PNS–P) compared with the control group. The difference was statistically significant ($P < 0.05$).

Superior and middle airway spaces were narrower in the thalassaemic patients. Superior and middle airway spaces averaged 8.37 ± 1.93 and 8.50 ± 3.11 mm in the thalassaemic patients compared with 14.50 ± 3.35 mm and 10.57 ± 2.29 mm in the control group ($P < 0.001$ and $P < 0.05$, respectively).

The hyoid bone was positioned close to the lower border of the mandible in the thalassaemic patients. The distance from hyoid to the mandibular plane (MPH) and from hyoid to retrognathion (HRGN) was 6.83 ± 4.50 and 33.33 ± 5.36 mm in the thalassaemic patients compared with 15.60 ± 3.64 and 39.73 ± 4.56 mm in the control group ($P < 0.001$).

Discussion

β-thalassaemia major is a life threatening condition, characterized by severe anaemia, hepatosplenomegaly, growth retardation, endocrine dysfunction and skeletal changes due to hypertrophy, and expansion of haematopoietic marrow. The best known oral manifestations of the condition are the enlargement of the maxilla,

Table 2 Means, standard deviations, differences between the means and significance (*P*) for thalassaemia and control groups.

Variable	Thalassaemia (*n* = 15)		Control (*n* = 15)		Difference between the means	Level of significance
	Mean	SD	Mean	SD		
Tongue						
TGL (mm)	69.77	5.95	75.07	5.75	–5.30	*
TGH (mm)	26.20	2.60	29.97	2.57	–3.77	***
Soft palate						
PNSP (mm)	29.23	4.10	34.37	3.12	–5.14	***
MPT (mm)	9.57	1.94	9.00	1.62	0.57	NS
Upper airway						
SPAS (mm)	8.37	1.93	14.50	3.35	–6.13	***
MAS (mm)	8.50	3.11	10.57	2.29	–2.07	*
IAS (mm)	12.57	2.96	12.37	3.30	0.20	NS
VAL (mm)	49.20	5.56	55.07	7.25	–5.87	*
Hyoid						
MPH (mm)	6.83	4.50	15.60	3.64	–8.77	***
HH1 (mm)	6.10	4.16	7.40	3.83	–1.30	NS
HRGN (mm)	33.33	5.36	39.73	4.56	–6.40	***
C3H (mm)	33.07	3.34	32.97	3.72	0.10	NS

*$P < 0.05$, ***$P < 0.001$; NS = not significant.

frontal bones, and zygoma due to bony expansion with depression of the bridge of the nose and flaring and spacing of the maxillary anterior teeth (Kaplan *et al.*, 1964; Van Dis and Langlais, 1986; Hes *et al.*, 1990).

Previous studies have been undertaken to investigate the airway space and morphology, tongue, and hyoid bone position in normal subjects and those with breathing difficulties. Patients with breathing difficulties have been found to have significant craniofacial abnormalities when compared with matched controls (Jamieson *et al.*, 1986). These abnormalities include a shorter mandibular body (Miles *et al.*, 1996), increased anterior face height (Lowe *et al.*, 1986; Bacon *et al.*, 1988; Tangugsorn *et al.*, 1995), inferiorly displaced hyoid bone (Jamieson *et al.*, 1986), enlarged tongue and soft palate (Tangugsorn *et al.*, 1995), and decreased posterior airway space (Battagel and L'Estrange 1996; Solow *et al.*, 1996).

Limited information is available concerning the craniofacial morphology of thalassaemia major patients and almost no evidence exists regarding the soft tissue structures and airway space in these subjects. Bassimitci *et al.* (1996) reported that thalassaemic patients usually present with a Class II skeletal pattern and a pronounced vertical mandibular growth direction. Abu Alhaija *et al.* (2002) investigated a group of thalassaemic patients and reported that a Class II skeletal pattern, a shorter cranial base length, a shorter mandible, and increased anterior vertical dimensions are typical features.

In the present study, it was found that all the variables studied were smaller. The thalassaemic patients showed a shorter soft palate and smaller tongue (length and height). The upper and middle airway spaces were found to be smaller than in the normal control group. Vertical airway length was shorter and the hyoid bone was found to lie closer to the mandibular plane. These findings could be explained by the fact that thalassaemic patients have a significant growth retardation caused by a variety of environmental factors including severe chronic anaemia, endocrine dysfunction, and somatomedin deficiency, which is marked as puberty is approached (Weatherall and Clegg, 1981).

Conclusions

β-thalassaemic major patients have a smaller tongue size, a shorter soft palate, narrower superior and middle pharyngeal airway spaces, a shorter vertical pharyngeal length, and the hyoid bone lies closer to the mandibular plane.

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