European Journal of Orthodontics 24 (2002) 9–19

Cephalometric measurements and facial deformities in subjects with β-thalassaemia major

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SUMMARY This study was performed to identify cephalometric and facial features of patients with β-thalassaemia major. A total of 54 thalassaemic subjects were examined for craniofacial deformities, including 37 patients (24 males and 13 females, aged 5–16 years) who had lateral cephalometric radiographs. The thalassaemic groups were compared with a normal control group matched for sex and dental age, using a *t*-test.

All thalassaemic patients had a Class II skeletal base relationship. The average ANB angle was significantly larger than the controls in dental stages 2 and 3 ($P < 0.05$). Mandibular base length (Ar–Gn) was significantly less in thalassaemic patients than in controls, with the greatest differences ($P < 0.001$) found in the younger age group. The maxilla was of normal length (PNS–ANS, Ptm′–ANS′) and appeared prominent (3.3 mm in males and 5.1 mm in females) due to a reduced cranial base length (Ar′–S′) and a short mandible (Ar′–P′). Vertically, thalassaemic patients showed a significantly increased maxillary/mandibular planes angle in all groups, with differences ranging between 6.19 and 12.55 degrees ($P < 0.001$). Thalassaemic patients also showed a reduced posterior facial height (S–Go, Ar–Go) and increased anterior facial proportions. Of the 54 thalassaemic patients examined, 17 per cent had severe facial disfigurements (grade 3).

Introduction

Thalassaemia is a group of inherited defects in the synthesis of either the α or β polypeptide chains of haemoglobin, referred to as α and β thalassaemia, respectively. Based on genetic and clinical entities, thalassaemias are classified as homozygous, heterozygous, or compound heterozygous (Weatherall and Clegg, 1981). The heterozygous form of the disease (thalassaemia minor) is mild, with minimal clinical expression. The homozygous form of β-thalassaemia (thalassaemia major) exhibits the most severe clinical symptoms with marked orofacial defects. A less severe form, thalassaemia intermedia, also occurs.

Homozygous β-thalassaemia (also known as Cooley's anaemia and Mediterranean anaemia) is transfusion-dependent and is commonly manifested during the first year of life. Frequently, affected infants are severely anaemic, fail to thrive and gain weight normally, and become progressively paler. Feeding problems, diarrhoea, recurrent fever, spontaneous fractures, bleeding, susceptibility to infection, hepatosplenomegaly, and retardation of growth are common presenting symptoms.

The suggested causes of growth retardation and maturation of the skeleton include chronic anaemia (Caffey, 1957), hyperparathyroidism (Flynn *et al*., 1976), and somatomedin deficiency (Saenger *et al*., 1980), a factor that stimulates cartilage growth. If children survive early childhood, they usually die during the second decade from cardiac failure due to iron deposition in the myocardium, chronic anaemia, and hypoxia (Modell, 1976; Weatherall and Clegg, 1981).

The best-known orofacial manifestations of β-thalassaemia are prominent cheek bones and a

protrusive premaxilla due to erythroid hyperplasia with depression of the bridge of the nose, often referred to as 'rodent' or 'chipmunk' faces (Kaplan *et al*., 1964; Cannel, 1988). Cephalometric radiographs show dilatation of the diploic space, especially in the frontal region and the subperiosteal bone grows in radial striations producing a bristle-like or 'hair on end' appearance of the skull (Poyton and Davey, 1968; Roy *et al*., 1971; Weel *et al*., 1987). The maxillary sinus is partially obliterated due to erythroid hyperplasia.

The dentition shows protrusion, flaring and spacing of the maxillary anterior teeth, open bite, and other degrees of malocclusion (Kaplan *et al*., 1964; Weel *et al*., 1987; Cannel, 1988; Hes *et al*., 1990). The tooth-crown size (Tas *et al*., 1976) and tooth length (Poyton and Davey, 1968) in thalassaemic subjects are significantly smaller than in unaffected groups. Panoramic and intra-oral radiography may show a generalised loss of bone density similar to that seen in osteomalacia or osteoporosis, and a thin cortex of the mandible. The trabeculae of the jaws appears coarse in pattern with enlarged marrow spaces described as 'chicken-wire' (Van Dis and Langlais, 1986; Hes *et al*., 1990; Barnard and Smallridge, 1998).

The prevalence of thalassaemia in Greece, Cyprus, Sardinia, and Turkey is as high as 15–20 per cent. The disease is also prevalent in some African and Far Eastern countries, as well as in India, Pakistan, Iran, and Israel (Kurdish Jews). In Jordan, approximately 1000 transfusion-dependent thalassaemic patients are registered (1:4600 of the total population) with an annual increase of 80 cases and a carrier rate of 7–10 per cent of the population.

It is now clear that thalassaemia probably represents the most common genetic disorder causing a major public health problem in the world's population. Surprisingly, very few detailed studies on craniofacial measurements and facial deformities in thalassaemic patients have been published.

The aim of the present study was to conduct a cephalometric analysis of patients with thalassaemia major and to compare measurements with normal subjects of similar ages. In addition, craniofacial deformities in thalassaemia major were clinically recorded and graded according to severity of involvement.

Materials and methods

The study was conducted at the Dental Centre of the Jordan University of Science and Technology, Irbid (the second largest city in Jordan with a population of 777,000). Approximately 310 thalassaemic patients are registered in Irbid, representing one-third of the diagnosed cases in Jordan. All patients were referred from two main thalassaemic units in the city.

A total of 54 thalassaemia major subjects were examined cephalometrically. Of those 37 patients (24 males and 13 females, aged 5.5–16 years) had lateral cephalometric radiographs. The average heights and weights of the patients were in the 3rd and 10th percentile, respectively, of the standard chart for the country population. The control group consisted of 37 normal subjects (24 males and 13 females, aged 6–15 years) who had a lateral cephalogram taken for orthodontic assessment. The participants (thalassaemic and control subjects) were divided into three groups according to dental stage (Table 1). In dental

Table 1 Distribution of thalassaemic patients and controls according to age (sexes pooled).

Dental stage	Number of subjects	Thalassaemia		Control	
		Mean age	SD	Mean age	SD
Dental stage 1	16	7.50	2.15	7.47	1.09
Dental stage 2	10	11.2	1.91	12.14	0.90
Dental stage 3	11	13.8	1.44	13.55	0.80

stage 1, the permanent first molars and incisors were erupted (mean age 7.5 ± 2.15 years for thalassaemia and 7.47 ± 1.09 years for control). In dental stage 2, the permanent lower canines and at least two premolars were erupted (mean age 11.2 ± 1.91 years for thalassaemia and 12.14 ± 0.90 years for control). In dental stage 3, all permanent teeth (except the third molars) were erupted (mean age 13.8 ± 1.44 years for thalassaemia and 13.55 ± 0.80 years for control). Because of the limited sample size and insignificant differences between sexes, male and female measurements in each dental age group were pooled.

Cephalometric radiographs were taken with a Siemens Orthophos-5 machine using a standardized technique; that is, the posterior teeth were in maximum intercuspation and the anode-mid-sagittal plane distance was fixed. The magnification of the radiographic machine was × 1.3. The lateral skull radiographs were traced on acetate paper and 16 cephalometric points were registered (Figure 1). In addition to the conventional cephalometric assessment, Wylie's analysis was performed (Wylie, 1947). This analysis is designed to detect horizontal malrelationship or dysplasia between maxillary and mandibular prognathism. The Frankfort plane was chosen as the horizontal with articulare as the origin. The projected lengths of the cranial base elements, maxilla, and mandible were measured, and compared with standard values. A maxillary or mandibular length, which exceeded the standard, indicated maxillary or mandibular prognathism, respectively. If the maxillary length was less than standard the deficit was added to mandibular prognathism and *vice versa*. If the effective length of the cranial base element exceeded the standard, the excess contributed to maxillary prognathism; if it was less than the standard, the deficit contributed to mandibular prognathism. Wylie's antero-posterior analysis is shown in Figure 2. The antero-posterior distances between points articulare (Ar), sella (S), pterygomaxillary fissure (Ptm), and anterior nasal spine (ANS) projected on to the Frankfort plane were

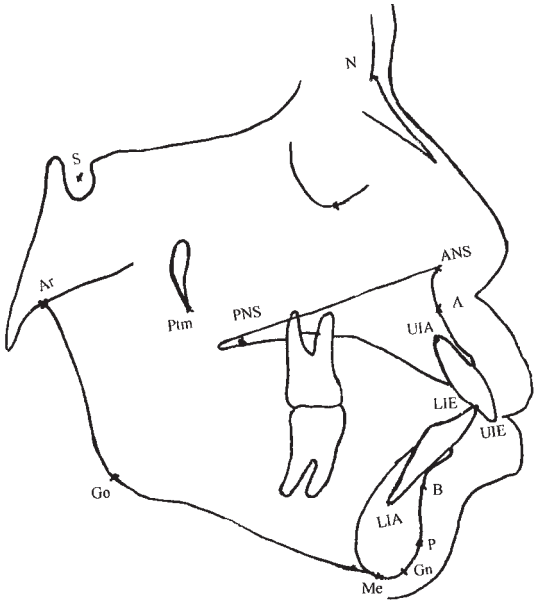

Figure 1 Points used in the cephalometric analysis. S: sella, N: nasion, Ar: articulare, ANS: anterior nasal spine, PNS: posterior nasal spine, Ptm: pterygomaxillary fissure, point A, point B, Me: menton, Gn: gnathion, P: pogonion, Go: gonion, UIE: mid-point of the upper central incisor edge, UIA: apex of the upper incisor, LIE: mid-point of lower central incisor edge, LIA: apex of lower central incisor.

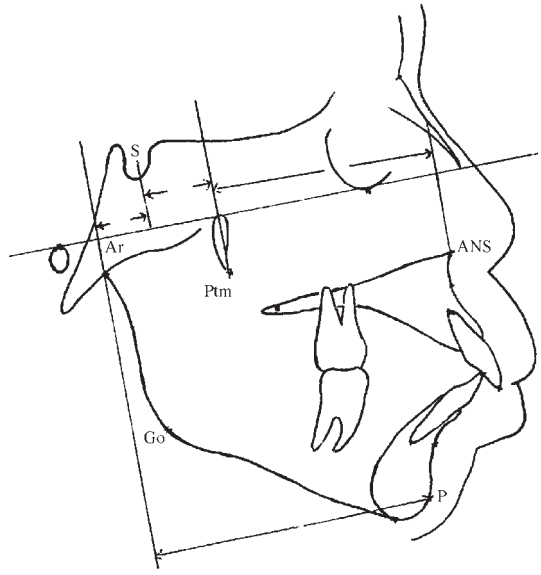

Figure 2 Horizontal skeletal analysis after Wylie. Measurements: Ar′–S′, the distance from articulare to sella projected on the Frankfort plane; S′–Ptm′, the distance between sella and pterygomaxillary fissure projected on Frankfort plane. Ptm′–ANS′, the distance between pterygomaxillary fissure and anterior nasal spine projected on Frankfort plane. Ar′–P′, the distance between articulare and pogonion projected on the Frankfort plane.

measured, and also the effective length of the mandible from Ar to Pogonion (P). Measurements were performed manually using a ruler and protractor, and recorded to the nearest 0.1 mm or 1 degree. The incisor relationship was recorded as follows:

Overjet
Normal: between 2 and 4 mm.
Increased: > 4 mm.
Reduced: ≥ 0 mm to < 2 mm.
Reversed: < 0 mm.

Overbite
Normal: between 2 and 3 mm.
Increased: > 3 mm.
Reduced: ≥ 0 mm to < 2 mm.
Open bite: < 0 mm.

Craniofacial deformities were clinically assessed for the 54 thalassaemic patients assigned into four grades according to severity of involvement:

Grade 0: No cephalofacial deformity present.
Grade 1: Slight depression of the nose, puffiness of the eyelids with no apparent maxillary overgrowth (Figure 3a).
Grade 2: Mild maxillary overgrowth and slight bulging of the cheeks and frontal bones (Figure 3b).
Grade 3: Prominent overgrowth of the maxilla, frontal and cheek bones, distinct depression of the bridge of the nose and protrusion of the anterior teeth 'chipmunk' faces (Figure 3c).

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 between 0.42 mm for overjet and 1.17 mm for S–Go, and from 0.60 degrees for SNB to 1.19 degrees for Li/Mand. Houston's coefficient of reliability ranged from 0.94 to 0.99.

Statistical analysis

Descriptive statistics, including the mean, standard deviation (SD) and difference between means for each group were computed using SPSS PC+. The variables were tested for normality. The differences between thalassaemic patients and controls were evaluated using the independent *t*-test if the variable was normally distributed, otherwise, an approximation by *z*-test was performed. A Chi-squared test was applied to test for differences in overjet and overbite distribution between the two groups.

Results

The results of the statistical analysis are shown in Tables 2–6.

Dental stage 1 (Table 2)

Angular measurements. The thalassaemic patients showed a reduced anterior cranial base angle (NSAr). This angle was 120.63 ± 4.39 in thalassaemic patients compared with 126.53 ± 6.48 in the controls. A difference of 5.90 degrees was statistically significant at $P < 0.01$. In the vertical plane, the maxillary/mandibular planes angle in thalassaemic patients was greater than in the control ($P < 0.001$). Upper incisor inclination was 100.19 ± 7.56 degrees in thalassaemic patients and 111.34 ± 6.51 degrees in controls. The difference of 11.15 degrees was highly significant ($P < 0.001$).

Linear measurements. Thalassaemic patients showed a reduced anterior cranial base length (S–N) compared with the controls. The difference was statistically significant ($P < 0.05$). The mandibular length (Ar–Gn) in thalassaemic patients was lower than in controls (88.63 ± 7.99 mm versus 98.88 ± 6.99 mm, $P < 0.001$). In the vertical plane, the total facial height (Na–Me) in thalassaemic patients was significantly less than in controls (105.44 ± 5.93 mm versus 112.84 ± 4.87 mm, $P < 0.01$). In thalassaemic patients, the lower (Ar–Go) and the total (S–Go) posterior facial heights averaged 35.34 ± 4.97 mm and 61.13 ± 5.36 mm, respectively. The corresponding heights in controls were 40.16 ± 3.76 mm and 66.59 ± 3.96 mm. These differences were statistically significant ($P < 0.01$).

Proportional measurements. Anterior facial proportion [lower anterior face height (ANS–Me)/

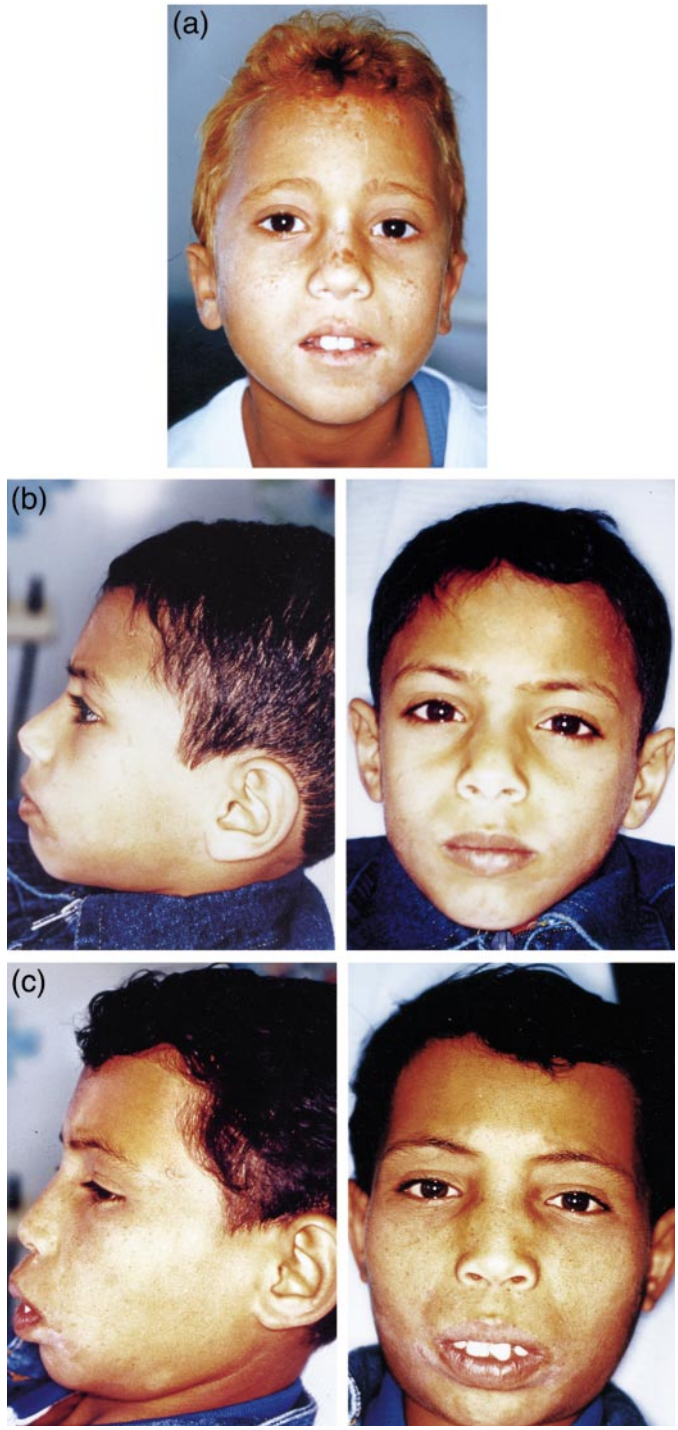

Figure 3 Grades of cephalofacial deformities in thalassaemia patients. (a) Puffiness of the eyelids with no maxillary overgrowth (grade 1). (b) Mild maxillary overgrowth (grade 2). (c) Gross maxillary overgrowth and prominent malar bones and maxillary anterior teeth, known as 'chipmunk' faces (grade 3).

Table 2 Means, standard deviations, differences between the means and significance for thalassaemia and control groups, dental stage 1.

Variable	Thalassaemia		Control		Difference between the means	Level of significance
	(n = 16) Mean	SD	(n = 16) Mean	SD		
Angular measurements (°)						
N–S–Ar	120.63	4.39	126.53	6.48	–5.90	**
SNA	80.84	3.33	79.03	2.70	1.81	NS
SNB	74.56	3.37	74.78	3.69	–0.22	NS
ANB	6.28	2.18	4.19	4.13	2.09	NS
Max/Mand	37.22	5.31	31.03	4.49	6.19	***
Ui/Max	100.19	7.56	111.34	6.51	–11.15	***
Li/Mand	91.75	6.50	92.09	8.10	–0.34	NS
Linear measurements (mm)						
S–N	64.41	3.80	67.28	2.71	–2.87	*
S–Ar	28.22	3.10	30.50	3.26	–2.28	NS
PNS–ANS	45.41	3.35	47.41	2.38	–2.00	NS
Ar–Gn	88.63	7.99	98.88	6.99	–10.25	***
Na–Me	105.44	5.93	112.84	4.87	–7.40	***
ANS–Me (LAFH)	63.94	3.69	63.47	3.82	0.47	NS
Ar–Go (LPFH)	35.34	4.97	40.16	3.76	–4.82	**
S–Go	61.13	5.36	66.59	3.96	–5.46	**
Proportional measurements						
AFP% (ANS–Me/N–Me)	61	0.02	56	0.03	0.05	***
PFP% (Ar–Go/S–Go)	58	0.04	60	0.06	–0.02	NS

NS = not significant, *P < 0.05, **P < 0.01, ***P < 0.001.

total anterior face height (Na–Me)] averaged 61 ± 2 per cent in thalassaemic patients compared with 56 ± 3 per cent in the control groups (P < 0.001).

Dental stage 2 (Table 3)

Angular measurements. ANB angle was the only variable that showed a statistically significant difference (P < 0.05). In absolute terms, the average angle was 8.85 ± 2.36 degrees in the thalassaemics and 5.86 ± 2.34 degrees in the controls.

Linear measurements. Posterior cranial base length (S–Ar) was smaller in thalassaemia patients compared with the controls. On average, it was 31.20 ± 3.11 mm and 35.29 ± 3.53 mm, respectively. The difference was statistically significant at P < 0.05.

Dental stage 3 (Table 4)

Angular measurements. In the antero-posterior plane the thalassaemic patients showed an increased ANB angle. The difference between the upper and lower dental bases (ANB) was 7.18 ± 2.23 degrees in thalassaemic patients compared with 3.73 ± 3.66 in controls. The difference of 3.45 degrees was significant (P < 0.05). In the vertical plane, the maxillary/mandibular planes angle in thalassaemic patients was significantly greater than in the controls (P < 0.001).

Linear measurements. Posterior cranial base (S–Ar) was reduced in those with thalassaemia. The average length was 30.18 ± 2.49 mm and, in controls, 35.00 ± 3.69 mm (P < 0.01). In the antero-posterior plane, the length of the mandible (Ar–Gn) in thalassaemic patients was less than in controls (P < 0.01). In the vertical plane, the lower anterior face height (ANS–Me) was increased (P < 0.05) and both the lower

Table 3 Means, standard deviations, differences between the means and significance for thalassaemia and control groups, dental stage 2.

Variable	Thalassaemia		Control		Difference between the means	Level of significance
	(n = 10) Mean	SD	(n = 10) Mean	SD		
Angular measurements (°)						
N–S–Ar	122.70	5.34	121.86	3.08	0.84	NS
SNA	81.45	3.48	81.29	3.25	0.16	NS
SNB	72.60	3.98	75.43	3.78	–2.83	NS
ANB	8.85	2.36	5.86	2.34	2.99	*
Max/Mand	39.85	11.22	33.00	7.55	6.85	NS
Ui/Max	105.75	5.37	106.07	8.93	–0.32	NS
Li/Mand	92.75	18.89	99.86	5.34	–7.11	NS
Linear measurements (mm)						
S–N	67.90	2.83	68.14	2.61	–0.24	NS
S–Ar	31.20	3.11	35.29	3.53	–4.09	*
PNS–ANS	47.90	4.75	49.43	5.56	–1.53	NS
Ar–Gn	95.50	7.76	101.86	9.46	–6.36	NS
Na–Me	115.10	5.99	118.29	5.53	–3.19	NS
ANS–Me (LAFH)	70.90	5.59	69.00	3.42	1.90	NS
Ar–Go (LPFH)	37.55	5.27	42.71	6.68	–5.16	NS
S–Go	65.75	6.28	73.07	10.28	–7.32	NS
Proportional measurements						
FP% (ANS–Me/N–Me)	62	0.03	58	0.04	0.04	NS
FP% (Ar–Go/S–Go)	57	0.05	59	0.04	–0.02	NS

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

(Ar–Go) and the total (S–Go) posterior face heights were reduced in thalassaemic patients ($P < 0.05$ and < 0.01, respectively).

Proportional measurements. The anterior facial proportion in thalassaemic and control groups averaged 62 ± 4 and 56 ± 2 per cent, respectively. The difference was significant at $P < 0.001$.

Wylie's analysis (Table 5)

A relative maxillary prognathism amounting to 3.3 mm in males and 5.1 mm in females was demonstrated in thalassaemic patients compared with Wylie's standard.

The overall distribution of overjet and overbite in thalassaemic patients and controls (age groups pooled) are presented in Table 6. A Chi-squared test revealed no significant difference in overjet and overbite distribution between the two groups.

A summary of the maxillary (PNS–ANS) and the mandibular (Ar–Gn) lengths in the three investigated groups of thalassaemia and controls are shown in Figures 4 and 5, respectively. The curves show that the maxilla in the thalassaemic patients grew similar to the controls. On the other hand, the mandible showed a rapid increase after dental stage 2 (mean age 11.2 ± 1.91 years) that was not present in the thalassaemic group.

These observations on craniofacial deformities indicate that facial disfigurement increased with age and duration of symptoms in uncontrolled cases. Of the 54 patients examined, 18 (33 per cent) had a normal appearance (grade 0), 14 (26 per cent) were classified as grade 1, 13 (24 per cent) as grade 2, and 9 (17 per cent) as grade 3.

Discussion

Little is known about the cephalometric characteristics and the posibility for orthodontic treatment in thalassaemic patients. Successful surgical correction and osseointegrated implant surgery of the maxilla have been reported in

Table 4 Means, standard deviations, differences between the means and significance for thalassaemia and control groups, dental stage 3.

Variable	Thalassaemia		Control		Difference between the means	Level of significance
	(n = 11) Mean	SD	(n = 11) Mean	SD		
Angular measurements (°)						
N–S–Ar	122.95	4.47	126.91	6.11	–3.96	NS
SNA	80.36	4.20	79.64	3.53	0.72	NS
SNB	73.18	3.66	75.91	4.18	–2.73	NS
ANB	7.18	2.23	3.73	3.66	3.45	*
Max/Mand	38.55	4.91	26.00	4.38	12.55	***
Ui/Max	107.45	8.26	114.77	9.51	7.32	NS
Li/Mand	98.64	9.60	97.46	8.78	1.18	NS
Linear measurements (mm)						
S–N	69.59	3.67	69.09	2.13	0.50	NS
S–Ar	30.18	2.49	35.00	3.69	–4.82	**
PNS–ANS	48.73	4.84	50.46	2.21	1.73	NS
Ar–Gn	98.45	7.44	108.05	8.16	–9.60	**
Na–Me	116.41	8.57	119.00	5.98	–2.59	NS
ANS–Me (LAFH)	72.09	5.50	66.73	5.34	5.36	*
Ar–Go (LPFH)	40.59	4.12	45.68	4.45	–5.09	*
S–Go	68.05	5.06	76.27	5.50	–8.22	**
Proportional measurements						
FP% (ANS–Me/N–Me)	62	0.04	56	0.02	0.06	***
FP% (Ar–Go/S–Go)	60	0.04	60	0.04	0.00	NS

NS = not significant, *P < 0.05, **P < 0.01, ***P < 0.001.

Table 5 Results of Wylie's analysis for the male (M) and female (F) subjects.

Measurement	Wylie standard (mm)		Sample mean ± 2SE (mm)		Maxillary prognathism (mm)		Mandibular prognathism (mm)	
	M	F	M	F	M	F	M	F
Ar′–S′	18.0	17.0	12.7 ± 0.85	13.3 ± 1.48			5.3*	3.7*
S′–Ptm′	18.0	17.0	17.5 ± 1.04	18.8 ± 2.24		1.8	0.5	
Ptm′–ANS′	52.0	52.0	51.6 ± 1.83	50.0 ± 2.70			0.4	2.0
Ar′–P′	103.0	101.0	93.5 ± 3.48	92.0 ± 2.63	9.5*	9.0*		
Prognathism totals					9.5	10.8	6.2	5.7

*P < 0.05.

some cases (Weel *et al*., 1987; Hes *et al*., 1990; Misch *et al*., 1998).

Adelman (1965) treated a severe case of thalassaemia orthodontically using an extra-oral facebow with neck traction. He concluded that the disease factor of thalassaemia does not interfere with osteoclastic and osteoblastic activity that occurs with orthodontic tooth movement. The present findings indicate that the cephalofacial deformities increased with age and duration of symptoms with 17 per cent of the patients having severe facial disfigurement. Similar findings were reported by Logothetis *et al*. (1971), who examined 138 Greek patients with thalassaemia major. In the present sample, however, the facial appearance was found to be more 'chipmunk' or

Table 6 Summary of overjet and overbite distribution in thalassaemic and control groups.

	Thalassaemia		Control	
	Overjet *n* (%)	Overbite *n* (%)	Overjet *n* (%)	Overbite *n* (%)
Normal	18 (48.7)	11 (29.7)	15 (40.5)	15 (40.5)
Increased	14 (37.8)	6 (16.2)	13 (35.1)	12 (32.4)
Reduced	4 (10.8)	12 (32.4)	5 (13.5)	6 (16.2)
Reversed	1 (2.7)	8 (21.6)	4 (10.8)	4 (10.8)

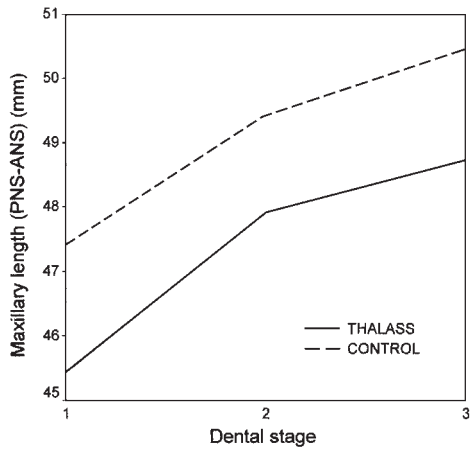

Figure 4 Length of the maxilla (PNS–ANS) in thalassaemic patients and controls at different dental stages.

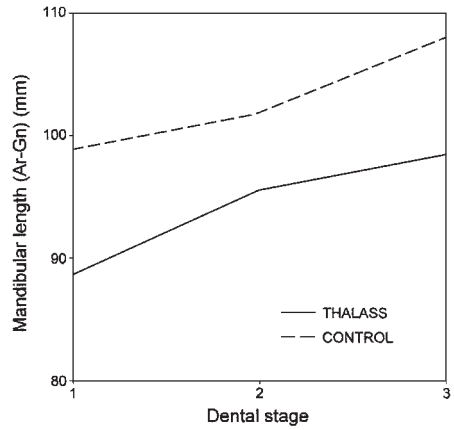

Figure 5 Length of the mandible (Ar–Gn) in thalassaemic patients and controls at different dental stages.

'rodent'-like than 'mongoloid' as reported by others (Cooly and Lee, 1925; Logothetis *et al.*, 1971).

Cephalometric analysis posed the problem that point nasion, which represents the anterior limit of the cranial base, may give misleading values for the angles SNA and SNB because the bridge of the nose is depressed. The less conventional Wylie analysis does not identify this point, but was used in addition to the conventional analysis.

Thalassaemic patients exhibited a large intermaxillary discrepancy (ANB) that produced a Class II skeletal pattern. The intermaxillary discrepancy was pronounced in dental stages 2 and 3. The maxilla was of normal length (PNS–ANS) in all dental age groups and the mandible was unusually short (Ar–Gn). Wylie's analysis showed maxillary prognathism in thalassaemic patients was attributed to a short cranial base (Ar′–S′, S′–Ptm′) and a short mandible (Ar′–P′).

In the vertical dimension, the thalassaemic patients showed an increased maxillary/mandibular planes angle, increased anterior facial proportions and a reduced total (S–Go) and lower (Ar–Go) posterior face heights. The posterior face height is largely determined by growth at the condyles, which is deficient probably due to anaemia. This results in a shorter posterior face height and subsequently an increased maxillary/mandibular planes angle. The increased maxillary/mandibular planes angle and the anterior face height indicate a vertical growth direction in thalassaemic patients. These finding confirm those of Bassimitci *et al.* (1996)

who found that typical thalassaemic patient had a moderate Class II skeletal pattern with a pronounced vertical mandibular growth direction.

The prominence of the premaxilla in thalassaemic patients (Figures 3c and 6) has led some authors to conclude that the maxillary incisors are proclined (Kaplan *et al*., 1964; Weel *et al*., 1987). However, cephalometric analysis showed that the upper incisors tended to be upright or even retroclined in some cases. Although lower incisor inclination in thalassaemic patients was within normal range, it is considered proclined in respect to the increased maxillary/mandibular angle (Houston *et al*., 1994). This incisor compensation could explain the normal overjet seen in 48.7 per cent of the thalassaemic patients and the increased overjet (>4 mm) in 37.8 per cent. This is comparable with those of the controls i.e. increased in 35.1 per cent (Table 6).

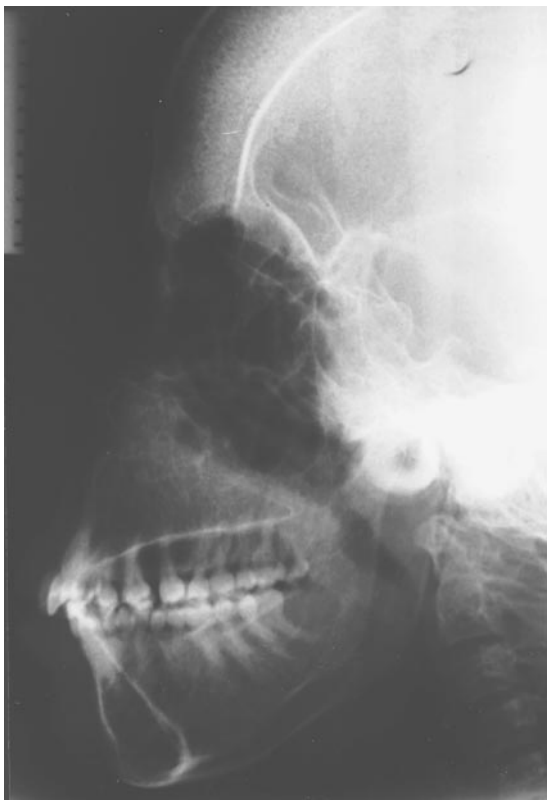

Figure 6 Cephalometric radiograph of patient with thalassaemia (grade 3) showing a prominent maxilla and a Class II skeletal pattern, increased vertical dimensions, a depressed nasal bridge, short roots, and a spaced dentition.

The overbite in the thalassaemic patients was normal in 29.7 per cent, increased in 16.2 per cent, reduced in 32.4 per cent, and an anterior open bite was present in 21.6 per cent. In the control group, only 27 per cent showed a reduced overbite and an anterior open bite. Kaplan *et al*. (1964) reported an increased overbite in 36 per cent and an anterior open bite in 30 per cent of the thalassaemic patients they examined. No measurements on the degree of malocclusion was given to allow direct comparison with the present findings.

A review of the literature reveals some inconsistency regarding the age at which growth retardation occurs in patients with thalassaemia major. Caffey (1957) and Weatherall and Clegg (1981) reported that slowing of growth is more marked as puberty is approached, while Erlandson *et al*. (1964) stated that there was a particular tendency for retarded growth to occur at 8–10 years of age. Lapatsanis *et al*. (1978) stated that in the 5–7 year age group, half of the children showed bone retardation (>6 months), whereas after this age bone retardation was found in almost two-thirds of the subjects. The results of the present investigation show that thalassaemic patients have significantly reduced craniofacial dimensions (Tables 2–4), apparently as part of a total growth retardation. Retarded growth of the maxilla and mandible was prominent at 7.5 years of age (dental stage 1) in thalassaemic patients (Figures 4 and 5). Although growth retardation almost invariably occurs in thalassaemia major, it may occur early in association with severe anaemia and in the under-transfused child. Further studies are needed to determine the pattern and magnitude of growth during early childhood.

The craniofacial manifestations of orthodontic concern include the Class II malocclusion associated with prominent maxillary anterior teeth and spacing of other teeth, as well as increased overjet and reduced overbite. It is therefore important to understand the changes associated with thalassaemia and their implication for orthodontic treatment.

Conclusions

The typical findings in subjects with thalassaemia are a Class II skeletal pattern, short cranial base length, short mandible, increased anterior and reduced posterior vertical dimensions, and severe facial disfigurement in 17 per cent of patients.

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Acknowledgements

The authors would like to thank Professor Andrew Richardson, Queen's University of Belfast for revising our manuscript and Dr Hani Samawi, Yarmouk University for advice on the statistical analysis.

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