Craniofacial Disproportions in Apert's Syndrome: An Anthropometric Study

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Twenty craniofacial indices composed of 26 surface measurements taken directly from the head and face were determined in 14 Apert's syndrome patients 18 days to 5 years old (younger subgroup) and 14 patients ages 6 years to 15 years old (older subgroup). All of the patients were North American Caucasians who had undergone early suture release but no facial repair. The indices were compared with those in healthy controls of the same age and sex. The wide intercanthal distance in relation to the narrow soft nose was the most frequent (81.5%) and extensive (17.7% above the maximum normal index value) disproportion. Abnormal indices occurred most often with the combination of one abnormal and one normal measurement (61.0%).

All seven of the 16 disproportions seen in both age subgroups increased in frequency nonsignificantly with age: the supernormal cephalic, intercanthal, nasal, and vertical mandibulofacial indices and the subnormal nasofacial, upper face, and jaws' arcs indices. Of the six disproportions that decreased in frequency with age, four changed significantly (the supernormal frontoparietal and frontozygomatic indices and the subnormal mandibulofacial and nasozygomatic indices) and two changed nonsignificantly (the supernormal intercanthoalar and subnormal cheilozygomatic indices). With the exception of two nasal proportions, the extent of the disproportionality decreased in all of the indices that increased with age.

Apert's syndrome is the most studied of the acrobrachycephalic anomalies (Gray and Dickey, 1947; Lewin, 1953; Kahn and Fulmer, 1955; Woolf et al, 1959; Gorlin and Pindborg, 1976; Becker et al, 1974; Cohen et al, 1971; Tessier, 1971; Dyken and Miller, 1980; Whitaker et al, 1981). Morphological studies are based more on visual observation (Lewin, 1953; Kahn and Fulmer, 1955; Woolf et al, 1959; Dyken and Miller, 1980) than on anthropometry or cephalometry (Gray and Dickey, 1947; Cohen et al, 1971; Tessier, 1971; Cohen, 1975). The study of disproportions in Apert's patients has been restricted to the determination of the head and orbit type (brachycephaly and hypertelorism).

To help distinguish between Apert's syndrome and other types of acrobrachycephaly and to gain information for planning correction, we investigated (1) the main disproportions in the cranio-orbito-facial area of Apert's syndrome patients, using facial indices; (2) the measurement revealing the source of such disproportions; and

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(3) the age-related changes in frequency and intensity of these disproportions.

MATERIAL

The study group consisted of 28 North American Caucasians with Apert's syndrome (8 males and 20 females) who had undergone early suture release but no facial repair. The patients were divided into two subgroups of 14: children 18 days to 5 years old (younger subgroup) and those between 6 years and 15 years of age (older subgroup); they were compared to normal populations of corresponding age groups. The data from the younger subgroup were compared to norms calculated for the West German population (Hajniš, 1974). The older subgroup was compared to North American population norms (Farkas, 1981).

METHODS

Twenty craniofacial proportion indices were investigated in the patients (Tables 1 to 6): six cranial, five facial, three orbital, three nasal, two labio-oral, and one in the ears. Each index was composed of two surface measurements and the 20 indices involved a total of 25 measurements: six cranial, seven facial, four orbital, two nasal, two labio-oral, and four in the ears. The methods of measurement and their reliability have been described elsewhere (Farkas, 1981). All measurements were taken by one of the authors (LGF).

EVALUATION OF FINDINGS

In general, the smaller measurement is the numerator of the index and the larger the denominator. Thus, the index indicates the smaller measurement as a percentage of the larger. The relationship of the two measurements was classified as proportionate or normal if the index value was within the range of the mean ± 2 standard deviations (SD) of the control population. Values more than 2 SD below the mean were qualified as subnormal, while values more than 2 SD above the mean were marked as super-

TABLE 1. Cranial Proportions

Index	*Refer to*	*Formula (A/B)*	*Subnormal Index*	*Supernormal Index*
Cephalic	Figures 1A and 1B	$\frac{\text{eu-eu} \times 100}{\text{g-op}}$	Narrow, elongated (dolichocephalic) head	Wide, short (brachycephalic) head
Vertical craniofacial	Figure 2	$\frac{\text{v-n} \times 100}{\text{n-gn}}$	Short face, high head	Long face, low head
Vertical frontocranial	Figure 3	$\frac{\text{tr-n} \times 100}{\text{v-n}}$	Low forehead in high head	High forehead in low head
Parietocranial	Figure 4	$\frac{\text{eu-eu} \times 100}{\text{v-po}}$	Narrow, high head	Wide, low head
Frontoparietal	Figure 5	$\frac{\text{ft-ft} \times 100}{\text{eu-eu}}$	Narrow forehead, wide head	Wide forehead, narrow head
Frontozygomatic	Figure 6	$\frac{\text{ft-ft} \times 100}{\text{zy-zy}}$	Narrow forehead, wide face	Wide forehead, narrow face

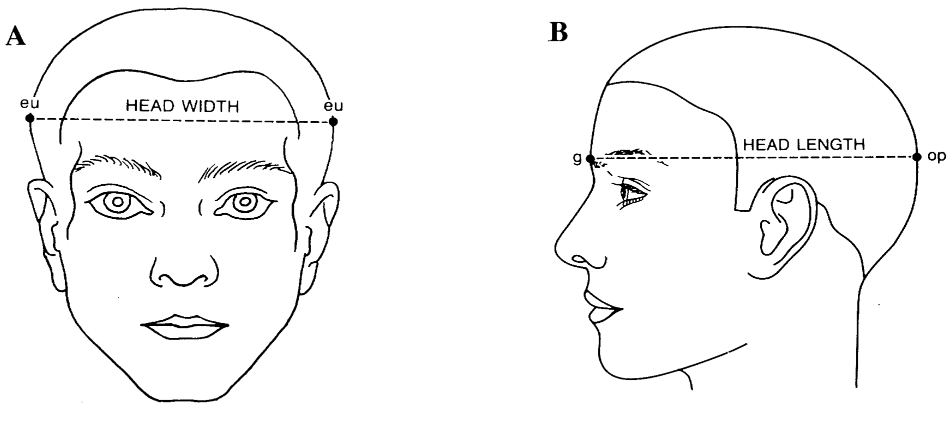


FIGURE 1. Cephalic index (eu = eurion; g = glabella; op = opisthocranion)

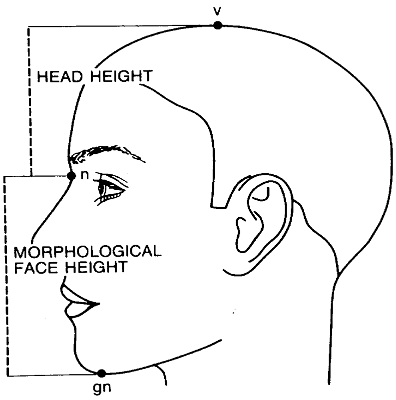


FIGURE 2. Vertical craniofacial index (v = vertex; n = nasion; gn = gnathion)

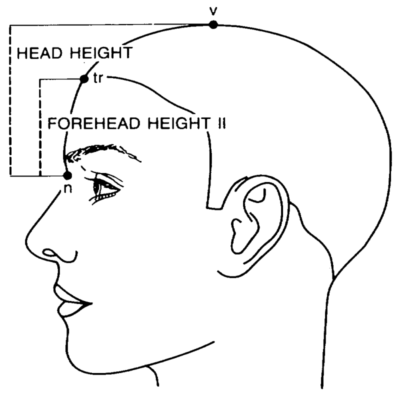


FIGURE 3. Vertical frontocranial index (tr = trichion)

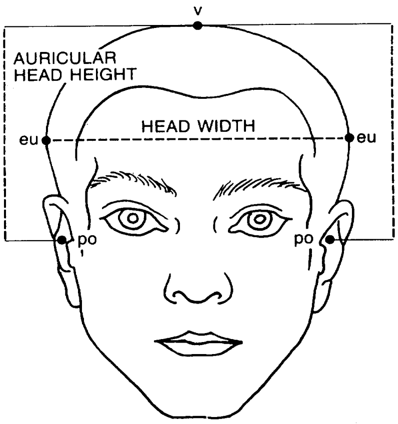


FIGURE 4. Parietocranial index (po = porion)

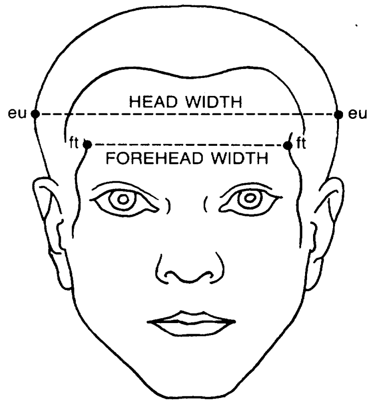


FIGURE 5. Frontoparietal index (ft = frontotemporale)

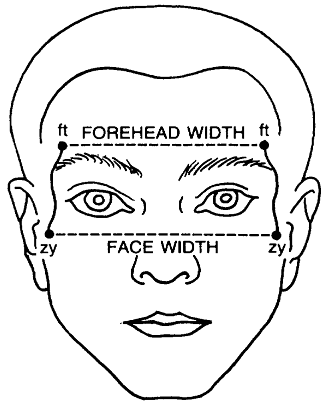


FIGURE 6. Frontozygomatic index (zy = zygion)

normal. Thus, either a subnormal or supernormal index signified a statistical disproportion. The visual appearance of the significant disproportion depended on its extent, which was expressed as a percentage of the smallest normal value for subnormal findings (relative reduction) or of the maximum normal for supernormal findings (relative increase).

Statistical Evaluation

In statistical analysis, the standard error of the difference (SED), a simple method for

TABLE 2. Facial Proportions

Index	*Refer to*	*Formula (A/B)*	*Subnormal Index*	*Supernormal Index*
Facial	Figure 7	$\frac{\text{n-gn} \times 100}{\text{zy-zy}}$	Short, wide face	Long, narrow face
Horizontal mandibulofacial	Figure 8	$\frac{\text{go-go} \times 100}{\text{zy-zy}}$	Narrow lower face with wide face	Wide lower face with narrow face
Vertical mandibulofacial	Figure 9	$\frac{\text{sto-gn} \times 100}{\text{n-gn}}$	Short chin in elongated face	Long chin in short face
Upper facial	Figure 10	$\frac{\text{n-sto} \times 100}{\text{zy-zy}}$	Short upper face, wide face	Long upper face, narrow face
Jaws' arcs	Figure 11	$\frac{\text{t-sn-t} \times 100}{\text{t-gn-t}}$	Recessed maxilla or pseudoprognatic mandible	Protruding maxilla or receding mandible

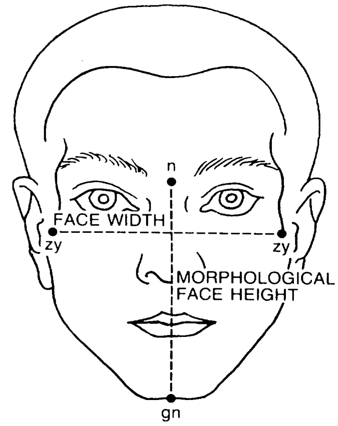


FIGURE 7. Facial index

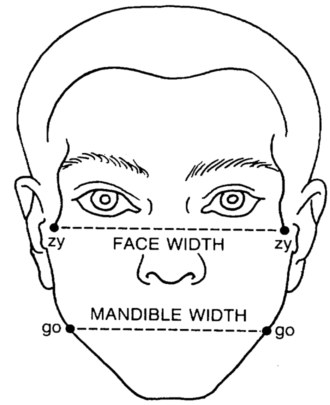


FIGURE 8. Horizontal mandibulofacial index (go = gonion)

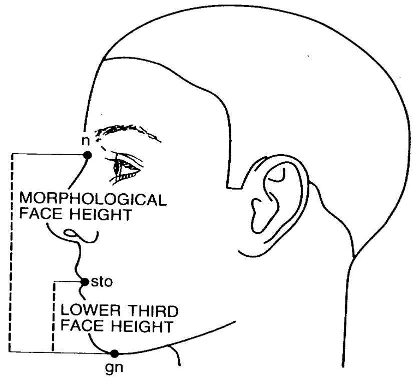


FIGURE 9. Vertical mandibulofacial index

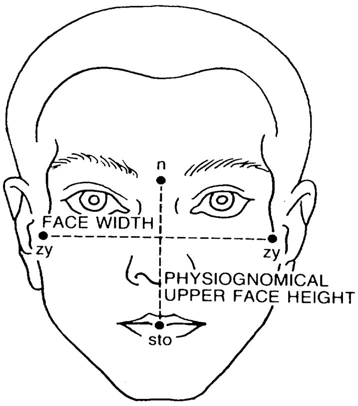


FIGURE 10. Upper facial index

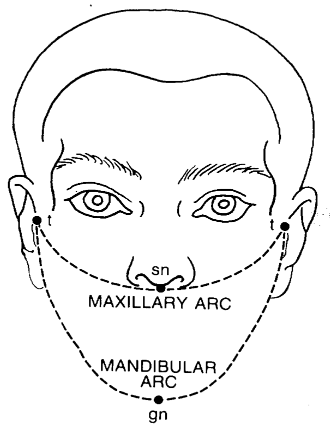


FIGURE 11. Jaws' arcs index (t = tragion; sn = subnasale)

analysis of a small sample with large expected SDs, was applied (Hughes, 1967). "As the standard error of the difference between two proportions is given by the formula:

$$\sqrt{p_1 q_1 / n_1 + p_2 q_2 / n_2}$$

where p_1 and q_1 are the proportions in the one sample and p_2 and q_2 those in the other, and n_1 and n_2 are the respective samples, a difference between two proportions may be said to be significant if it exceeds twice its own standard error" (Hughes, 1967) (p=percentage, $q=100-p$, n=size of sample, difference between two proportions = difference between p_1 and p_2).

TABLE 3. Orbital Proportions

Index	*Refer to*	*Formula (A/B)*	*Subnormal Index*	*Supernormal Index*
Intercanthal	Figure 12	$\frac{\text{en-en} \times 100}{\text{ex-ex}}$	Hypoteloric-type orbit	Hyperteloric-type orbit
Orbital	Figure 13	$\frac{\text{en-ex} \times 100}{\text{en-en}}$	Short palpebral fissures with wide interocular space	Long palpebral fissures with narrow interocular space
Intercanthoalar	Figure 14	$\frac{\text{en-en} \times 100}{\text{al-al}}$	Narrow intercanthal space with wide soft nose	Wide intercanthal space with narrow soft nose

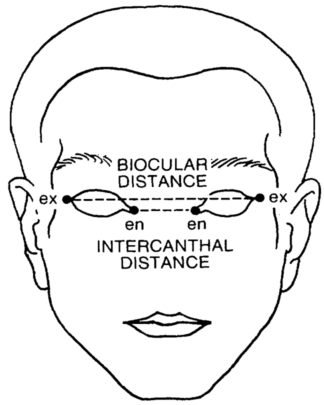


FIGURE 12. Intercanthal index (en = endocanthion; ex = exocanthion)

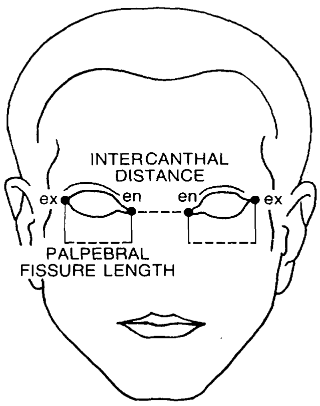


FIGURE 13. Orbital index

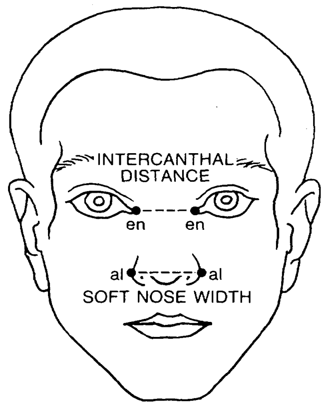


FIGURE 14. Intercanthoalar index (al = alare)

TABLE 4. Nasal Proportions

Index	*Refer to*	*Formula (A/B)*	*Subnormal Index*	*Supernormal Index*
Nasal	Figure 15	$\frac{\text{al-al} \times 100}{\text{n-sn}}$	Narrow, elongated nose	Wide, short nose
Nasofacial	Figure 16	$\frac{\text{n-sn} \times 100}{\text{n-gn}}$	Short nose in long face	Long nose in short face
Nasozygomatic	Figure 17	$\frac{\text{al-al} \times 100}{\text{zy-zy}}$	Narrow, soft nose in wide face	Wide nose in narrow face

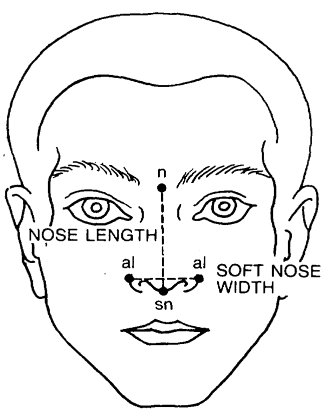


FIGURE 15. Nasal index

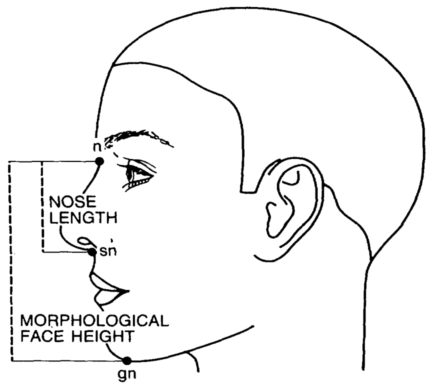


FIGURE 16. Nasofacial index

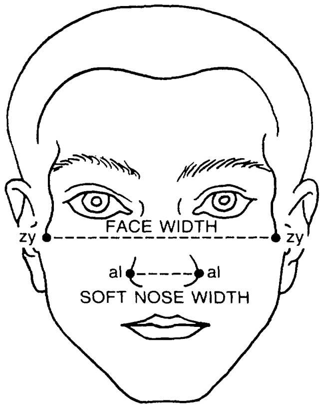


FIGURE 17. Nasozygomatic index

TABLE 5. Labio-Oral Proportions

Index	*Refer to*	*Formula (A/B)*	*Subnormal Index*	*Supernormal Index*
Labial	Figure 18	$\frac{\text{sn-sto} \times 100}{\text{ch-ch}}$	Short lip and wide mouth	Long lip and narrow mouth
Cheilozygomatic	Figure 19	$\frac{\text{ch-ch} \times 100}{\text{zy-zy}}$	Narrow mouth in wide face	Wide mouth in narrow face

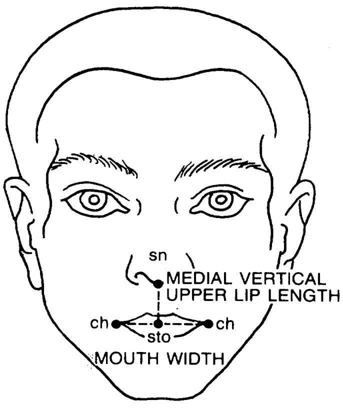


FIGURE 18. Labial index (ch = cheilion)

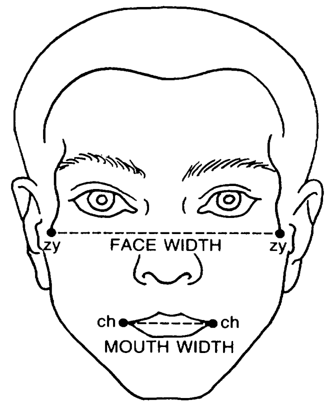
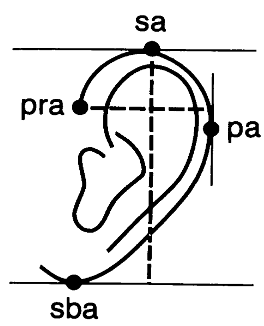


FIGURE 19. Cheilozygomatic index

TABLE 6. Ear Proportion

Index	*Refer to*	*Formula (A/B)*	*Subnormal Index*	*Supernormal Index*
Auricular	Figure 20	$\frac{\text{pra-pa} \times 100}{\text{sa-sba}}$	Narrow, elongated ear	Wide, short ear

FIGURE 20. Auricular index (pra = preaurale; pa = postaurale; sa = superaurale; sba = subaurale)



FINDINGS

Cranial Disproportions

The most frequent cranial disproportion was the brachycephalic type head (cephalic index), seen in almost two-thirds of patients, followed by a high head in relation to the face (vertical craniofacial index) (Table 7). The forehead was markedly low in relation to the head height in one-half of the older patients (vertical frontocranial index) and the head was long in relation to its width in almost as many (parietocranial index). About one-third to one-half had a wide forehead in relation to the facial width (frontozygomatic index) and head width (frontoparietal index).

Extent of Disproportions

The largest disproportion (13.3% below the minimum normal index value) was reported in the vertical frontocranial index (low head). The extent of disproportion between the forehead and face widths (frontozygomatic index) was the smallest (3.9% above the maximum normal index value).

Sources of Disproportionate Indices

Of the 55 cranial disproportions, 35 (64%) had one abnormal measurement. Marked difference between the measurements resulted in 14 disproportionate indices (25%) in which both

TABLE 7. Cranial Proportions in Apert's Syndrome

	Younger Subgroup			*Older Subgroup*			*Total Group*		
Index	*Normal*	*Subnormal*	*Supernormal*	*Normal*	*Subnormal*	*Supernormal*	*Normal*	*Subnormal*	*Supernormal*
Cephalic									
Number/total	7/14		7/14	4/14		10/14	11/28		17/28
% of patients	50		50	29		71	39		61
% from nearest normal index			12.9			5.6			9.3
Vertical Craniofacial									
Number/total				5/12		7/12	5/12		7/12
% of patients				42		58	42		58
% from nearest normal index						5.2			5.2
Vertical Frontocranial									
Number/total				6/12	6/12		6/12	6/12	
% of patients					50			50	
% from nearest normal index					13.3			13.3	
Parietocranial									
Number/total				7/13	6/13		7/13	6/13	
% of patients				54	46		54	46	
% from nearest normal index					6.5			6.5	
Frontoparietal									
Number/total	3/9		6/9	10/13		3/13	13/22		9/22
% of patients	33		67	77		23	59		41
% from nearest normal index			11.2			4.4			7.8
Frontozygomatic									
Number/total	1/9		8/9	11/13		2/13	12/22		10/22
% of patients	11		89	85		15	55		45
% from nearest normal index			5.9			1.8			3.9

measurements were abnormal and six (11%) in which both measurements were in the normal range.

Age-Related Differences

Three of the six cranial indices were calculated in both subgroups and the other three only in the older subgroup because normal data for the younger children were not available.

The brachycephalic type head (cephalic index) increased nonsignificantly (SED=18.0; diff, 21.4) in frequency but decreased markedly in extent. Wide foreheads decreased significantly both in relation to the head width (frontoparietal index, SED=19.6; diff, 43.6) and face width (frontozygomatic index, SED=14.5; diff, 73.5). The extent of the disproportions also decreased markedly.

FACIAL DISPROPORTIONS

The facial proportions are shown in Table 8. The most frequent facial disproportion was the narrow lower face within a relatively wide face (horizontal mandibulofacial index). The least frequent disproportion was observed in the relatively elongated, narrow face (facial index).

Extent of Disproportions

The greatest disharmony (11.0% below the minimum normal index value) was reported in the horizontal mandibulofacial index (narrow lower face in wide face). The mildest disproportion (2.7% above the maximum normal index

TABLE 8. Facial Proportions in Apert's Syndrome

	Younger Subgroup			*Older Subgroup*			*Total Group*		
Index	*Normal*	*Subnormal*	*Supernormal*	*Normal*	*Subnormal*	*Supernormal*	*Normal*	*Subnormal*	*Supernormal*
Facial									
Number/total	13/14	1/14		13/14		1/14	26/28	1/28	1/28
% of patients	93	7		93		7	93	4	4
% from nearest normal index		9.6				2.7		9.6	2.7
Horizontal Mandibulofacial									
Number/total		10/10		7/14	7/14		7/24	17/24	
% of patients		100		50	50		29	71	
% from nearest normal index		15.1			6.8			11.0	
Vertical Mandibulofacial									
Number/total	5/10		5/10	3/14		11/14	8/24		16/24
% of patients	50		50	21		79	33		67
% from nearest normal index			10.6			8.0			9.3
Upper facial									
Number/total	7/12	5/12		5/14	9/14		12/26	14/26	
% of patients	58	42		36	64		46	54	
% from nearest normal index		11.5			8.0			9.8	
Jaws' arcs									
Number/total	6/9	1/9	2/9	10/14	4/14		16/23	5/23	2/23
% of patients	67	11	22	71	29		70	22	9
% from nearest normal index		2.1	4.5		1.4			1.8	4.5

value) was seen between face height and width (facial index).

Sources of Disproportionate Indices

Of the 56 facial disproportions, 29 (52%) had one abnormal measurement. Marked difference between the measurements was the cause of 19 disproportionate indices (34%) in which both measurements were normal and seven (13%) in which both measurements were abnormal.

Age-Related Differences

All 5 facial indices were calculated in both subgroups. The only significant age-related difference was the decreasing frequency of disproportion in the horizontal mandibulofacial index (SED=13.7; diff, 50.0). The extent also decreased. The long chin relative to the face (vertical mandibulofacial index) and short upper face in the wide face (upper facial index) increased nonsignificantly in frequency but decreased in extent. Relatively large maxillary arcs were present only in the younger children, but relatively small maxillary arcs increased nonsignificantly with age (jaws' arcs index). The extent of the disproportions decreased markedly.

Orbital Disproportions

The orbital proportions are shown in Table 9. The most frequent orbital disproportion was the markedly wide space between the eyes compared with nose width (intercanthoalar index). Hyperteloric orbits (intercanthal index) were the least frequent.

Extent of Disproportions

The largest disproportion (17.0% above the maximum normal index value) was reported in the intercanthoalar index (wide intercanthal space with narrow nose). The extent of disproportion

TABLE 9. Orbital Proportions in Apert's Syndrome

Index	*Younger Subgroup*			*Older Subgroup*			*Total Group*		
	Normal	*Subnormal*	*Supernormal*	*Normal*	*Subnormal*	*Supernormal*	*Normal*	*Subnormal*	*Supernormal*
Intercanthal									
Number/total	9/14		5/14	6/14		8/14	15/28		13/28
% of patients	64		36	43		57	54		46
% from nearest normal index			7.6			6.6			7.1
Orbital									
Number/total				4/14	10/14		4/14	10/14	
% of patients				29	71		29	71	
% from nearest normal index					10.8			10.8	
Intercanthoalar									
Number/total	2/13		11/13	3/14		11/14	5/27		22/27
% of patients	15		85	21		79	19		82
% from nearest normal index			17.7			16.3			17.0

in the hyperteloric orbits was the smallest (7.1% above the maximum normal index) (intercanthal index).

Sources of Disproportionate Indices

Of the 45 orbital disproportions, 27 (60%) were caused by a supernormal intercanthal distance (one hyperteloric orbit, 10 relatively short eye fissures, and 16 wide intercanthal distances in relation to soft nose width). Of the 22 supernormal intercanthoalar disproportions, 16 were due to a supernormal intercanthal space (en-en), one was the result of subnormal width of the soft nose (al-al) and 5 resulted from a supernormal intercanthal space and a relatively narrow soft nose. All 10 subnormal orbital indices were caused by supernormal intercanthal width with normal eye-fissure widths. In 12 of 13 hyperteloric orbits, both distances were supernormal, the intercanthal distance greater than the biocular distance; in the other case, only the intercanthal space was supernormal.

Age-Related Differences

Two of the three orbital indices were calculated in both subgroups. The frequency of hyperteloric orbits (intercanthal index) increased nonsignificantly with age but the percentage of wide intercanthal spaces with relatively narrow noses (intercanthoalar indices) decreased. For both measurements, the extent of the disproportions decreased slightly.

NASAL DISPROPORTIONS

Nasal proportions are shown in Table 10. The most frequent nasal disproportion was a narrow soft nose in relation to face width (nasozygomatic index). The nose was normally or abnormally short in more than two-thirds of patients and became disproportionate to the face height (nasofacial index) in 41 percent of the patients. Approximately one-fifth of the noses were disproportionately wide for their length (nasal index).

Extent of Disproportions

The largest disproportion (10.8% below the minimum normal index value) was in nose length in relation to face height.

Sources of Disproportionate Indices

Of the 29 nasal disproportions, 18 (62%) had one abnormal measurement (short nose, narrow nose, or wide face). In seven (24%) of the disproportions both measurements were in the normal range and in four others (14%), there was a combination of one supernormal and one subnormal measurement.

TABLE 10. Nasal Proportions in Apert's Syndrome

Index	*Younger Subgroup*			*Older Subgroup*			*Total Group*		
	Normal	*Subnormal*	*Supernormal*	*Normal*	*Subnormal*	*Supernormal*	*Normal*	*Subnormal*	*Supernormal*
Nasal									
Number/total	12/13		1/13	9/14		5/14	21/27		6/27
% of patients	92		8	64		36	78		22
% from nearest normal index			3.7			11.0			7.4
Nasofacial									
Number/total	9/13	4/13		7/14	7/14		16/27	11/27	
% of patients	69	31		50	50		59	41	
% from nearest normal index		10.4			11.2			10.8	
Nasozygomatic									
Number/total	4/13	9/13		11/14	3/14		15/27	12/27	
% of patients	31	69		79	21		56	44	
% from nearest normal index		7.8			5.3			6.6	

Age-Related Differences

All three nasal indices were calculated in both subgroups. A nose disproportionately wide for its length increased nonsignificantly (nasal index, SED = 14.4; diff, 25.6) in frequency and extent. A relatively narrow nose in relation to face width was significantly more frequent and greater in extent in the younger subgroup (nasozygomatic index, SED = 16.9; diff, 47.8). The frequency and extent of short noses in relation to face height (nasofacial index) increased nonsignificantly (SED = 18.5; diff, 19.2).

LABIO-ORAL DISPROPORTIONS

As is seen in Table 11, in more than one-third of the patients the labial fissure was short in relation to the face width (cheilozygomatic index).

Extent of Disproportions

The largest disproportion (7.8% above the maximum normal index value) was in the relatively high upper lip in relation to labial fissure length (labial index).

Sources of Disproportionate Indices

Of the 10 disproportions between labial fissure length and face width (cheilozygomatic index), five (50%) were caused by subnormal mouth width, two by supernormal face width, two by relative disproportions within the normal ranges, and one by a combination of a subnormal labial fissure and a supernormal face. Disproportions between upper lip height and mouth width (labial index) were due to subnormal lip height in one case and supernormal height in the other.

TABLE 11. Labio-Oral Proportions in Apert's Syndrome

Index	*Younger Subgroup*			*Older Subgroup*			*Total Group*		
	Normal	*Subnormal*	*Supernormal*	*Normal*	*Subnormal*	*Supernormal*	*Normal*	*Subnormal*	*Supernormal*
Labial									
Number/total	13/13			12/14	1/14	1/14	25/27	1/27	1/27
% of patients	100			86	7	7	93	4	4
% from nearest normal index					2.2	7.8		2.2	7.8
Cheilozygomatic									
Number/total	7/13	6/13		10/14	4/14		17/27	10/27	
% of patients	54	46		71	29		63	37	
% from nearest normal index		7.3			6.3			6.8	

TABLE 12. Ear Proportions in Apert's Syndrome

Index	Younger Subgroup			Older Subgroup			Total Group		
	Normal	Subnormal	Supernormal	Normal	Subnormal	Supernormal	Normal	Subnormal	Supernormal
Auricular									
Number/total	12/14		2/14	13/14	1/14		25/28	1/28	2/18
% of patients	86		14	93	7		89	4	7
% from nearest normal index			9.9		3.4			3.4	9.9

Age-Related Differences

Both labio-oral indices were calculated in both subgroups, but labial index disproportions were noted only in the older children. Disproportional cheilozygomatic indices decreased nonsignificantly with age in frequency and extent (SED=18.4; diff, 17.6).

AURICULAR DISPROPORTION

Disproportionally wide ears were almost twice as common as disproportionally narrow (elongated) ears (Table 12).

Extent of Disproportions

The disproportion in relatively wide-short ears was greater (9.9% above the maximum normal index value) than in relatively long-narrow auricles (3.4% below the minimum normal index value).

Sources of Disproportionate Indices

All auricular disproportions occurred because the ears were supernormal in either width or length.

Age-Related Differences

Relatively wide-short ears were reported in only two children of the younger subgroup, and relatively long-narrow ears in only one older child.

DISCUSSION

In this study we analyzed certain proportions of the patients' faces rather than just the individual measurements. Each absolute measurement gives information about only one line, inclination, or angle in the face. Even a large number of facial measurements does not reflect the relation among measurements unless the proportion indices are calculated. In the past, many investigators and clinicians have emphasized the significance of proportions in studies of facial morphology, both in normal studies (Retzius, 1842; Dzierżykraj-Rogalski and Modrzewska, 1955; Figalová and Šmahel, 1972) and in patients (Burian, 1971; Firmin et al, 1974; Šmahel and Figalová, 1976; Maue-Dickson, 1979). When judging the face, we subconsciously receive an impression of a number of measurements and their relation as they appear to us in the given conditions (e.g., position of the head, distance, light). For example, when a face is observed in the frontal view, the length of the nose is obviously related to the height of the face, the width of the eye fissure to the space between the eyes, and the width of the mouth to the width of the face. Since morphologic defects in a malformed face are the second most important reason for surgical correction (after functional defects) (Tessier, 1971), objective identification of facial disproportion is indispensible.

The extent of abnormality of an index was expressed as a percentage of the difference from the terminal (the highest or lowest values of the normal range); we believe percentage changes of the index demonstrate the degrees of disproportion better than the absolute difference alone.

The division of the study group into younger and older subgroups was necessary because we have no valid North American Caucasian norms for children less than 6 years of age; it was not intended to provide information about changes during growth. As a surrogate, we used the West German population norms (Hajniš, 1974), which have fewer measurements (and, therefore, proportion indices) than are available for our controls over 6 years of age.

Our study confirmed the wide scale of proportion variations in the cranio-orbito-facial complex of Apert's syndrome patients, which resulted from the varying extent in premature closure of the coronal suture (Lewin, 1953; Cohen et al, 1971; Rogers, 1977; Dyken and Miller, 1980; Whitaker et al, 1981) and/or various cranial fossa and cranial base defects (Kreiborg et al, 1976; Stewart, 1976; Converse et al, 1977). In contrast with the frequently cited brachycephaly or hypertelorism (Gray and Dickey, 1947; Cohen et al, 1971; Tessier, 1971; Whitaker et al, 1981), a disproportionally wide interorbital space compared with the soft nose width was our most frequent finding. The brachycephalic head and hypertelorism were the fourth and ninth most common findings.

The abnormal midface or maxillary hypoplasia mentioned frequently by others (Lewin, 1953; Woolf et al, 1959; Becker et al, 1974; Cohen et al, 1971; Tessier, 1971; Rogers, 1977; Dyken and Miller, 1980; Whitaker et al, 1981) could not be demonstrated fully in our study by using the jaws' arcs index. The maxillary arc, running in a mild curve between the tragion of the ears and the base of the columella (subnasale), could not reveal all maxillary deficiencies. Although only 22 percent of maxillary arcs were disproportionally short in relation to the mandibular arcs, there was a nonsignificant but definite trend to short (normal and subnormal) maxillary arcs (83%, 19 of 23) compared to mandibular arcs (65%, 15 of 23). This area showed the smallest extent of disproportion. In agreement with Tessier's (1971) observation, the degree of hypertelorism was moderate (7.1%) in our sample.

The greatest single disproportion found in one patient was a wide intercanthal space with narrow soft nose (62.9% above the maximum normal index value). The greatest average disproportion (17.7% above the maximum normal index value) was also seen between the wide intercanthal space and the narrow soft nose.

Facial indices at various ages offer valuable information about the rate of growth (Hunter, 1968; Šmahel and Figalová, 1976). Of the 20 craniofacial proportion indices in this study, 16 were recorded in both subgroups; seven (44%) increased nonsignificantly, while six (38%) decreased in frequency with age. In four indices the decrease was significant: the head and face width increased more than the forehead and the mandible, and soft nose width more than the face.

Of the 200 disproportions we detected in the faces of our patients, almost two-thirds (122) were signalled by the combination of normal and abnormal measurements. Two normal measurements resulted in 34 disproportionate indices, a combination of a subnormal and a supernormal measurement was responsible for 27, and two supernormal measurements caused the remaining 17.

The extent of disproportion, the identification of the measurements leading to disproportionate indices, and the effect of age-related changes on these disproportions are all important when planning surgical repair.

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