Nasopharyngeal Dysmorphology In The Syndromes of Apert and Crouzon

SALLY J. PETERSON-FALZONE, Ph.D.
SAMUEL PRUZANSKY, D.D.S., M.S.
PAMELA J. PARRIS, M.S.
JOANNE L. LAFFER, M.S.
Chicago, Illinois 60680

Serial cephalometric studies on 29 patients with Apert syndrome and 26 patients with Crouzon disease confirmed and expanded previously reported observations of dysmorphology of the nasopharynx and contiguous structures. Particularly remarkable were alterations in pharyngeal depth, pharyngeal height, length of the posterior cranial base, and length of the hard and soft palate. The distortions and displacements contributing to diminition of the nasopharyngeal space were typically present early in life and became more severe as the patient matured. Implications for care and treatment planning are discussed.

KEY WORDS: Cephalometrics, Apert syndrome, Crouzon disease, nasopharynx

In 1974, the current authors reported on palatal anomalies in a series of 19 patients with Apert syndrome and 13 patients with Crouzon disease. The focus of that report was twofold: (a) a detailed description of the anomalous configuration of the maxillary arch and palatal vault, both of which had been at least partially described by previous investigators but only in Apert patients (Blank, 1960; Buchanan, 1968; Buckley and Yakovlev, 1948; Ferriman, 1941; Gorlin and Pindborg, 1964; Solomon et al., 1973), and (b) the first documentation of increased length and thickness of the soft palate in comparison to the norms of Subtelny (1957). Solomon et al. in 1973 had described the age-dependent changes in the anomalous configuration of the palatal vault in Apert syndrome, the major thrust of that report being a histochemical description of the contents of the soft tissue accumulations along the palatine processes. Peterson and Pruzansky (1974) found these accumulations to occur in both Apert and Crouzon patients but to be more consistent in occurrence and more severe in the former. No relationship was found between presence of either a bifid uvula or an overt cleft palate and either the abnormal size of the soft palate or the aberrant configuration of the palatal vault, which creates the image of a pseudo-cleft. Although the data utilized in that report were cross-sectional, the cephalometric measurements and dental study casts taken on patients ranging in age from less than 3 months to over 30 years strongly suggested growing severity of these palatal anomalies with increasing age. The significance of these findings in relationship to velopharyngeal function, nasal respiratory physiology, and speech did not escape our attention.

The authors are affiliated with the Center for Craniofacial Anomalies, Abraham Lincoln School of Medicine, University of Illinois Medical Center, Chicago, Illinois. Dr. Peterson-Falzone is Associate Professor, Department of Otolaryngology, and Head, Speech and Hearing Section of CCFA. Dr. Pruzansky is Professor of Dentistry, Department of Pediatrics and Director of Research of CCFA. Ms. Parris and Ms. Laffer are Research Associates.

This investigation was supported in part by grants from the National Institute for Dental Research, DE-02872, and from Maternal and Child Health, Department of Health and Human Services, MCPD-0128842.

Review of the Literature

As a result of the surgical reconstruction procedures introduced by Tessier (1971), there has been a proliferation of literature concerned with the clinical study and management of patients with syndromes including premature craniofacial synostosis. Previous radiographic studies (Aleksandroya et al., 1979; Ebel, 1974; Giuffre et al., 1978; Matras et al., 1977) focused mainly on the osseous deformities of the neurocranium and facial

skeleton with scant attention to the nasopharyngeal space. An exception was a 1979 paper by McCarthy et al. These authors used airflow studies, articulation tests, and lateral roentgencephalometry to study velopharyngeal function before and after maxillary advancement in 40 patients, including four with Apert syndrome and 15 with Crouzon disease. With specific regard to preoperative radiographic findings, McCarthy et al. (1979) reported that:

(a) nasopharyngeal height and depth were less than normal in all four Apert patients;

(b) nasopharyngeal depth was less than normal in 14 of the 15 patients with Crouzon disease.

Unfortunately, the remainder of the findings with regard to nasopharyngeal height and velar length are not interpretable for the Apert and Crouzon patients since data on them were not segregated from those for other patients.

Although not primarily a radiographic study, the 1977 report by Witzel and Munro on the effects of facial osteotomies on speech and velopharyngeal function included some pertinent observations on nasopharyngeal air-

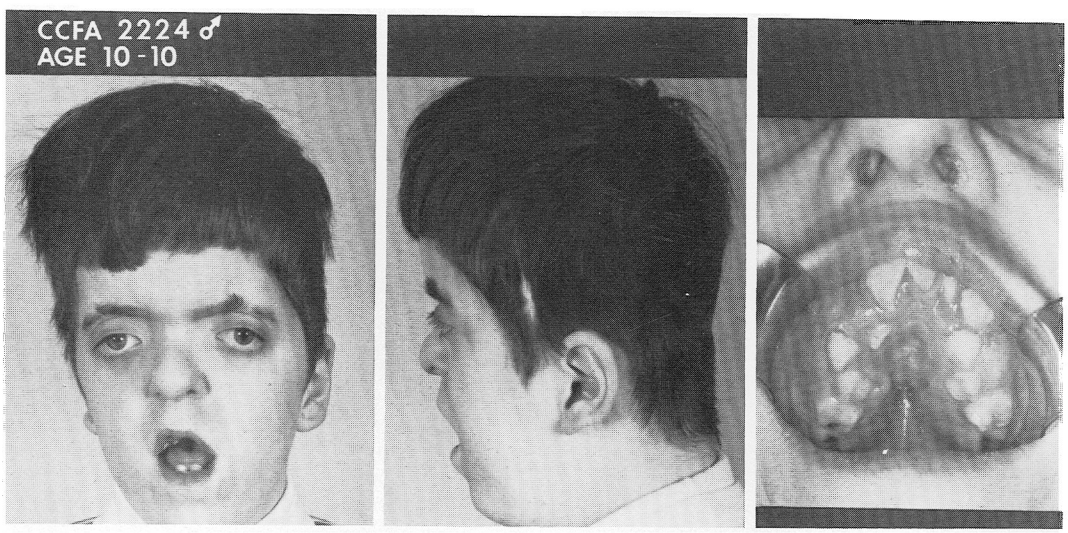

FIGURE 1. Facial and intraoral features in Apert syndrome (CCFA 2224). Note accumulations of soft tissue in palatal vault.

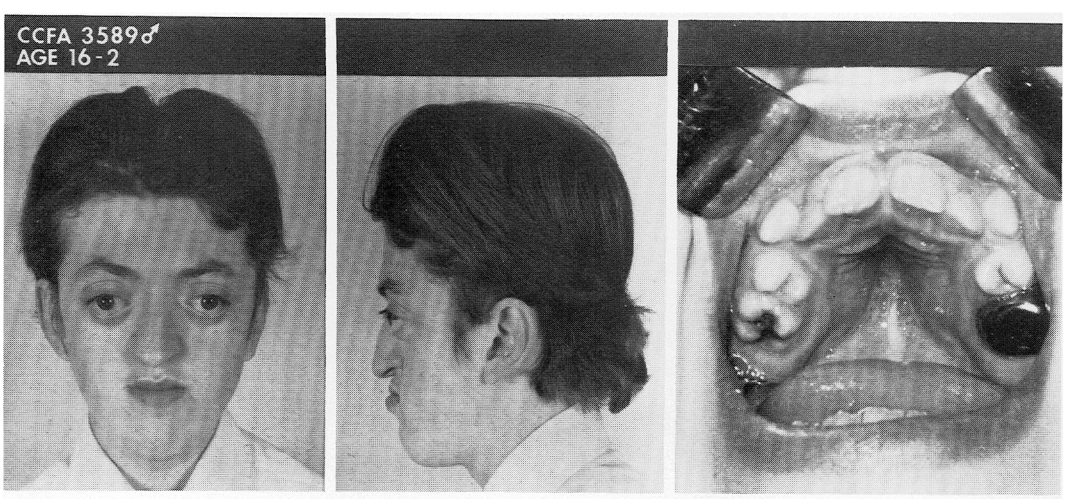

FIGURE 2. Facial and oral features in Crouzon disease (CCFA 3589).

space in a series of 11 Apert and Crouzon patients. The authors reported that, prior to LeFort III osteotomies, five Apert and two Crouzon patients had hyponasality, "absent" nasal air emission, abnormally long soft palates, and pharyngeal depth described as "almost negligible." Following surgery, "velopharyngeal function became possible" in one Crouzon and four Apert patients. The authors did not report on the postoperative state in the remaining Apert and Crouzon patients in their series.

As is evident from the above, a concurrence of interest in nasopharyngeal morphology among those centers treating patients with Apert syndrome and Crouzon disease is predictable since this morphology directly affects several areas of function—speech, hearing, breathing, head-to-neck posture, and tongue posture—particularly in the preoperative state. The current report represents an extension of earlier roentgenographic studies (Peterson and Pruzansky, 1974, and Peterson-Falzone et al., 1978). Relationships to other forms of assessment are discussed.

Procedure

Data utilized for this report were a mixture of longitudinal and cross-sectional cephalometric measurements on a series of 29 patients with Apert syndrome and 26 with Crouzon disease. Whereas our previous paper (1978) utilized only data on patients for whom longitudinal records were available, for purposes of this study we used all films available within each age category. Thus some patients were represented only once, (i.e., infants with only one set of films) while, for others, longitudinal records extending over a number of years were available. The longest series of records extended from three to 24 years of age.

A total of 232 preoperative lateral cephalometric films were digitized on an IMLAC system. The digitization protocol operates on a series of 28 points designed for analysis of craniofacial morphology in a variety of syndromes. Among the points recorded are

1. Na = nasion
2. H = an erected point where a perpendicular from the palatal plane at PNS intersects with line Na-Ba
3. S = midpoint of sella
4. Ba = basion
5. D = a point on the posterior pharyngeal wall where the extended palatal plane intersects with the wall
6. W = a point on the posterior superior surface of the soft palate which is one end point of the line marking the greatest width of the resting soft palate
7. L = the inferior tip of the resting soft palate
8. W′ = a point on the anterior inferior surface of the resting soft palate which is the end of the line W-W′ marking the greatest width of the resting soft palate
9. PNS^1 = anatomical posterior nasal spine when visible
10. PNS^2 = the point at which the pterygomaxillary fissure crosses the palatal plane
11. ANS = anterior nasal spine

The following measurements were selected from among those produced by the protocol:

1. Soft palate length at rest (PNS-L)
2. Soft palate thickness at rest (W-W′)
3. Hard palate length measured from anatomic PNS (ANS-PNS^1)
4. Hard palate length measured from constructed PNS (ANS-PNS^2)
5. Pharyngeal height (H-PNS)
6. Pharyngeal depth measured from anatomic PNS (D-PNS^1)
7. Pharyngeal depth measured from constructed PNS (D-PNS^2)
8. Anterior cranial base (S-Na)
9. Posterior cranial base (Ba-S)
10. Cranial base angle (Ba-S-Na)

Results

Separate means and standard deviations were computed for the Apert and Crouzon patients at each of the ages indicated in Table 1. Because the number of patients for whom a readable lateral cephalometric film was available varied from age to age and because many patients were represented more than once in the data by virtue of their longitudinal records, no attempt was made to analyze the data statistically other than plotting the means against graphs representing the 95% confidence intervals as computed from the data of Subtelny (1957) for measurements of soft palate length (Figure 3), palatal thickness

TABLE 1. Number of Apert and Crouzon Patients from Whom Cephalometric Records Were Available at the Ages Indicated. Years Taken as Whole Years (i.e., 9 Years = 9-0-0 Through 9-11-30).

Age	*Apert*	*Crouzon*
3 mos	7	4
6 mos	6	1
9 mos	5	3
1 yr	5	2
2 yrs	9	6
3 yrs	6	4
4 yrs	8	5
5 yrs	7	7
6 yrs	8	2
7 yrs	4	5
8 yrs	8	6
9 yrs	6	4
10 yrs	6	5
11 yrs	3	5
12 yrs	4	6
13 yrs	7	4
14 yrs	6	1
15 yrs	4	5
16 yrs	4	5
17 yrs	5	3
18+ yrs	2	4

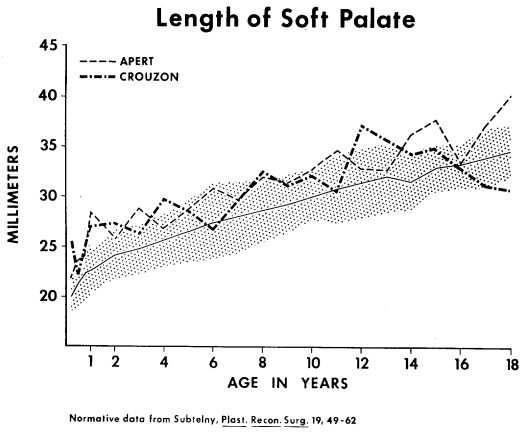

FIGURE 3. Mean for soft palate length plotted against the normative data of Subtelny (1957). The solid line connects the Subtelny means, with the shaded area representing the 95% confidence interval (± 2 standard deviations).

(Figure 4), pharyngeal height (Figure 5), and pharyngeal depth (Figure 6). Means for the length of the hard palate and the length of the anterior and posterior cranial base are plotted in Figures 7, 8, and 9 against similar graphs computed on the Bolton standards (1975).

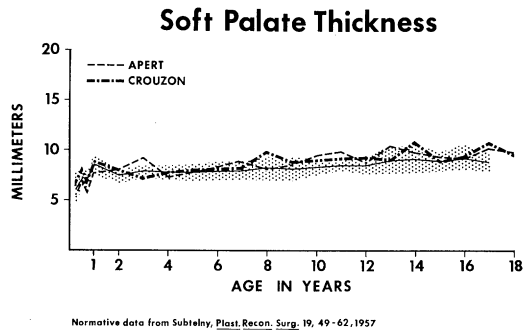

FIGURE 4. Means for palatal thickness plotted against the normative data of Subtelny (1957).

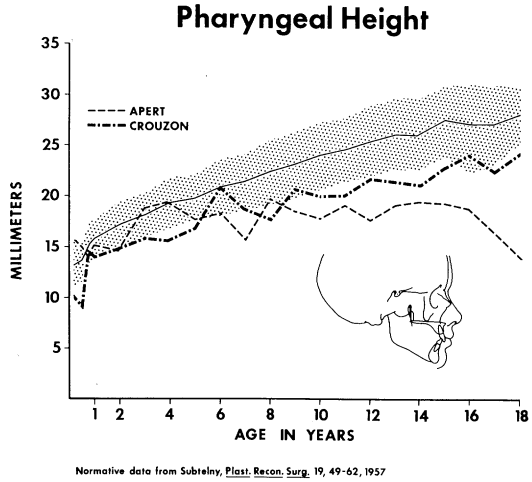

FIGURE 5. Means for pharyngeal height plotted against the normative data of Subtelny (1957).

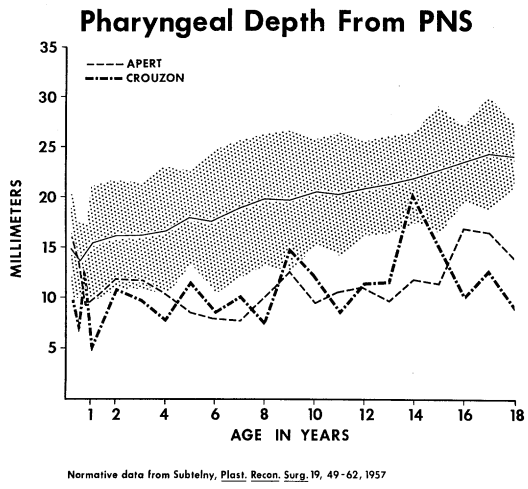

FIGURE 6. Means for pharyngeal depth plotted against the normative data of Subtelny (1957).

Although the means and standard deviations for cranial base angle in the Apert and Crouzon patients are presented in Table 11, no graph is included for this measurement. As we reported in 1978 on a more limited sample, marked distortions of this angle in individual patients are obscured in the averaging process because individual measurements can vary from platybasia to acute angulation. Although virtually all the means in Table 11 fall within the "normal range" reported by Brailsford (1944) and by McGregor (1948), individual measurements in the current series ranged from 111.0° to 164.6° in the Apert patients and 119.8° to 157.1° in the Crouzon patients. Such extremes are of far more importance in clinical evaluation and treatment planning for the individual patient (Pruzansky, 1977) than is the attempt to identify a central tendency in either patient group.

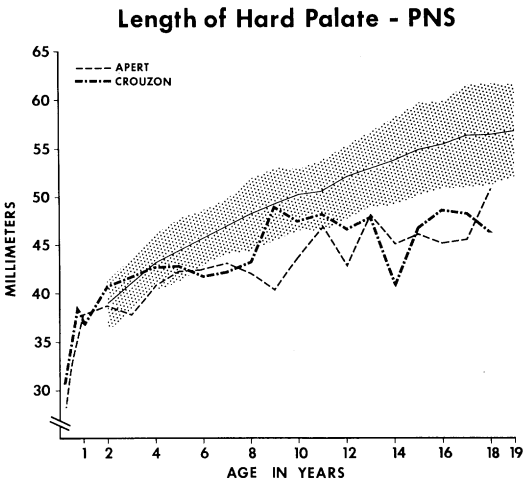

FIGURE 7. Means for hard palate length plotted against the Bolton standards (1975).

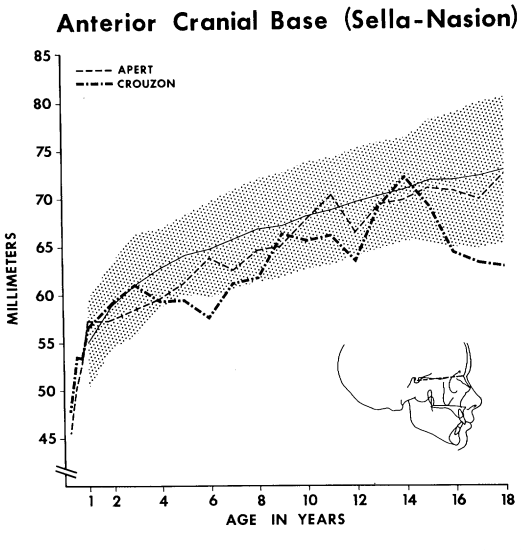

FIGURE 8. Means for anterior cranial base length plotted against the Bolton standards (1975).

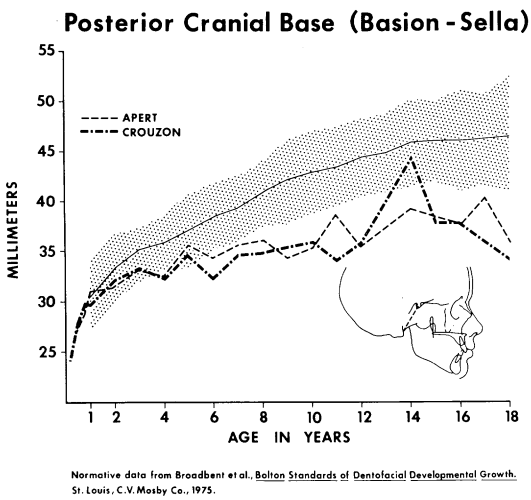

FIGURE 9. Means for posterior cranial base length plotted against the Bolton standards (1975).

Tables 2 through 10 list the means for the remaining measurements and the standards against which they were plotted in the graphs. No graphs are included for those measurements which were based on "constructed PNS" since the comparisons between these measurements and those based on anatomic PNS (for pharyngeal depth and hard palate length) showed too little difference to warrant comment. It should be noted that means for all measurements represent n's ranging from one to eight (see Table 1) and thus cannot be expected to show an orderly progression in value from year to year. The smaller the n, the greater the influence of any extreme values. For example, the films available on the 14-year-old patient with Crouzon disease yielded measurements of soft palate thickness, hard palate length, pharyngeal depth, and posterior cranial base length which were markedly "out of line" with the mean values at proximal ages.

TABLE 2. Mean Length in Millimeters of the Soft Palate in Apert and Crouzon Patients in Comparison to the Norms of Subtelny (1957).

Age	*Subtelny norm*	*s.d.*	*+ 2 s.d.*	*Apert* $\bar{X}$	*Crouzon* $\bar{X}$
3 mos	20.1	1.33	17.5–22.7	22.4	25.5
6 mos	21.3	1.43	18.5–24.1	23.6	22.2
9 mos	22.4	1.81	18.8–26.0	24.2	24.8
1 yr	22.6	1.36	19.8–25.4	28.5	27.1
2 yrs	24.3	1.44	21.5–27.1	25.9	27.4
3 yrs	24.8	1.52	21.8–27.5	28.8	26.4
4 yrs	25.7	1.51	22.7–28.7	26.9	29.7
5 yrs	26.5	1.86	22.7–30.3	28.9	28.6
6 yrs	27.4	2.11	23.2–31.6	30.8	26.9
7 yrs	28.0	2.18	23.6–32.4	29.8	29.5
8 yrs	28.6	1.80	25.0–32.2	32.0	32.6
9 yrs	29.3	1.70	25.9–32.7	31.5	31.0
10 yrs	30.1	1.72	26.7–33.5	32.8	32.2
11 yrs	30.8	2.09	26.6–35.0	34.7	30.5
12 yrs	31.4	2.14	27.2–35.6	32.9	37.2
13 yrs	32.0	2.15	27.6–36.4	32.7	35.9
14 yrs	31.5	1.76	27.3–35.1	36.4	34.4
15 yrs	32.9	1.31	27.5–34.7	37.7	34.9
16 yrs	33.1	1.31	29.5–34.7	33.5	33.0
17 yrs	33.8	1.81	30.2–36.4	37.3	31.2
18 yrs	34.5	1.57	31.3–37.7	40.2	30.8

TABLE 3. Mean Thickness in Millimeter of the Soft Palate in Apert and Crouzon Patients in Comparison to the Norms of Subtelny (1957).

Age	*Subtelny norm*	*s.d.*	*+ 2 s.d.*	*Apert* $\bar{X}$	*Crouzon* $\bar{X}$
3 mos	5.9	.84	4.3–7.5	5.5	6.8
6 mos	7.2	.40	6.4–8.0	6.7	8.1
9 mos	7.3	.58	6.5–8.5	5.8	6.8
1 yr	7.5	.66	6.1–8.9	7.7	8.8
2 yrs	7.4	.46	6.4–8.4	8.0	8.0
3 yrs	7.8	.39	7.0–8.6	9.1	7.2
4 yrs	7.7	.42	6.9–8.5	7.3	7.7
5 yrs	7.7	.54	6.7–8.7	7.8	8.0
6 yrs	7.9	.56	6.7–9.1	8.4	8.2
7 yrs	7.9	.56	6.7–9.1	8.7	8.3
8 yrs	8.2	.75	6.6–9.8	8.2	9.8
9 yrs	8.1	.71	6.7–9.5	8.6	8.9
10 yrs	8.3	.49	7.3–9.3	9.5	9.0
11 yrs	8.6	.40	7.8–9.4	9.9	9.2
12 yrs	8.5	.58	7.3–9.7	8.7	9.2
13 yrs	9.0	.83	7.4–10.6	10.5	9.0
14 yrs	9.2	.83	7.6–10.8	9.7	10.8
15 yrs	8.9	.62	7.7–10.1	9.4	8.9
16 yrs	9.3	.78	7.7–10.9	9.3	9.5
17 yrs	8.8	.56	7.6–10.0	10.2	10.8
18 yrs				9.9	9.6

TABLE 4. Mean Values in Millimeters for Length of Hard Palate Measured From ANS to Anatomical PNS in Apert and Crouzon Patients in Comparison to the Bolton Standards (1975).

Age	*Bolton standard*	*s.d.*	*+ 2 s.d.*	*Apert* $\bar{X}$	*Crouzon* $\bar{X}$
3 mos				28.1	30.6
6 mos				32.6	34.0
9 mos				35.1	38.8
1 yr	38.8	1.5	35.8–41.8	37.9	35.8
2 yrs	41.0	1.7	37.6–44.4	38.6	40.8
3 yrs	43.2	1.8	39.6–46.8	37.9	41.7
4 yrs	44.5	1.9	40.7–48.3	40.7	42.7
5 yrs	45.8	1.8	42.2–49.4	42.3	42.8
6 yrs	47.0	1.7	43.6–50.4	42.5	41.9
7 yrs	48.2	2.3	43.6–52.8	43.2	42.3
8 yrs	49.3	2.2	44.9–53.7	42.1	43.4
9 yrs	50.2	1.6	47.0–53.4	40.4	48.9
10 yrs	50.5	1.8	46.9–54.1	43.9	47.5
11 yrs	52.0	1.9	48.2–55.8	46.9	48.2
12 yrs	53.0	2.4	48.2–57.8	42.9	46.6
13 yrs	53.7	2.7	48.3–57.1	48.2	47.9
14 yrs	54.9	2.8	49.3–60.5	45.1	40.8
15 yrs	55.4	2.7	50.0–60.8	46.2	46.7
16 yrs	56.3	3.1	50.1–62.5	45.2	48.6
17 yrs	56.4	3.2	50.0–62.8	45.6	48.3
18 yrs	56.8	2.9	51.0–62.6	50.7	46.2

TABLE 5. Mean Values in Millimeter for Length of the Hard Palate Measured From ANS to constructed PNS in Apert and Crouzon Patients in Comparison to the Bolton Standards (1975).

Age	*Bolton standard*	*s.d.*	*+ 2 s.d.*	*Apert* $\bar{X}$	*Crouzon* $\bar{X}$
3 mos				28.2	30.6
6 mos				32.5	34.0
9 mos				35.3	38.8
1 yr	38.8	1.5	35.8–41.8	37.9	35.8
2 yrs	41.0	1.7	37.6–44.4	38.9	40.8
3 yrs	43.2	1.8	39.6–46.8	38.3	41.7
4 yrs	44.5	1.9	40.7–48.3	40.6	42.7
5 yrs	45.8	1.8	42.2–49.4	42.4	42.9
6 yrs	47.0	1.7	43.6–50.4	41.8	42.4
7 yrs	48.2	2.3	43.6–52.8	43.2	42.3
8 yrs	49.3	2.2	44.9–53.7	42.4	43.7
9 yrs	50.2	1.6	47.0–53.4	40.5	49.4
10 yrs	50.5	1.8	46.9–54.1	43.8	47.8
11 yrs	52.0	1.9	48.2–55.8	47.3	48.6
12 yrs	53.0	2.4	48.2–57.8	43.2	46.6
13 yrs	53.7	2.7	48.3–57.1	48.6	47.9
14 yrs	54.9	2.8	49.3–60.5	45.3	41.8
15 yrs	55.4	2.7	50.0–60.8	46.8	47.1
16 yrs	56.3	3.1	50.1–62.5	45.5	48.3
17 yrs	56.4	3.2	50.0–62.8	46.0	49.4
18 yrs	56.8	2.9	51.0–62.6	50.7	46.4

TABLE 6. Mean Values in Millimeters for Pharyngeal Height in Apert and Crouzon Patients in Comparison to the Norms of Subtelny (1957).

Age	*Subtelny norm*	*s.d.*	+ *2 s.d.*	*Apert* $\bar{X}$	*Crouzon* $\bar{X}$
3 mos	13.3	1.5	10.3–16.3	15.6	9.8
6 mos	13.7	1.2	11.3–16.1	14.9	9.0
9 mos	15.2	1.3	13.6–17.8	14.0	14.7
1 yr	16.0	1.1	13.8–18.2	15.2	14.0
2 yrs	17.4	1.3	14.8–20.0	14.7	14.9
3 yrs	18.3	1.3	15.7–20.9	18.9	15.8
4 yrs	19.5	1.5	16.5–22.5	19.4	15.7
5 yrs	19.9	1.4	17.1–22.7	17.7	17.0
6 yrs	21.0	1.7	17.6–24.4	18.4	20.9
7 yrs	21.5	1.6	18.3–24.7	15.8	18.7
8 yrs	22.5	1.9	18.7–26.3	19.8	17.7
9 yrs	23.2	2.0	19.2–27.2	18.6	20.8
10 yrs	24.2	2.0	20.2–28.2	17.9	20.1
11 yrs	24.6	1.8	21.0–28.2	19.3	20.2
12 yrs	25.5	2.2	21.1–29.9	17.8	21.8
13 yrs	26.3	2.1	22.1–30.5	19.3	21.6
14 yrs	26.2	2.1	22.0–30.4	19.6	21.2
15 yrs	27.5	2.1	23.3–31.7	19.4	23.0
16 yrs	27.2	2.6	22.0–32.4	18.8	24.2
17 yrs	27.2	2.4	22.4–32.0	16.6	22.6
18 yrs	28.1	1.7	24.7–31.5	14.0	24.4

TABLE 7. Mean Values in Millimeters for Pharyngeal Depth Measured from Anatomic PNS in Apert and Crouzon Patients in Comparison to the Norms of Subtelny (1957).

Age	*Subtelny norm*	*s.d.*	+ *2 s.d.*	*Apert* $\bar{X}$	*Crouzon* $\bar{X}$
3 mos	14.7	2.9	8.9–20.5	16.0	9.8
6 mos	13.9	2.2	9.5–18.3	12.4	7.0
9 mos	14.6	1.8	11.0–18.2	9.4	12.8
1 yr	15.4	3.5	8.4–22.4	9.7	5.0
2 yrs	16.3	3.2	9.9–22.7	11.9	10.9
3 yrs	16.3	3.2	9.9–22.7	11.9	9.9
4 yrs	16.8	3.9	9.0–24.6	10.4	7.9
5 yrs	18.1	2.8	12.5–23.7	8.6	11.5
6 yrs	17.7	4.4	8.9–26.5	8.0	8.6
7 yrs	19.0	4.1	10.8–27.2	7.8	10.2
8 yrs	19.9	3.9	12.1–27.7	10.1	7.6
9 yrs	19.7	4.2	11.3–28.1	12.7	15.0
10 yrs	20.6	3.2	14.2–27.0	9.6	12.4
11 yrs	20.4	3.7	13.0–27.8	10.7	8.7
12 yrs	21.0	2.8	15.4–26.6	11.0	11.5
13 yrs	21.4	2.9	15.6–27.2	9.9	11.6
14 yrs	22.0	2.7	16.6–27.4	13.9	20.2
15 yrs	22.9	3.8	15.3–30.5	13.6	15.5
16 yrs	23.5	2.3	18.6–28.1	17.0	10.2
17 yrs	24.4	3.4	17.6–31.2	16.5	12.7
18 yrs	24.2	1.9	20.4–28.0	14.0	9.3

TABLE 8. Mean Values in Millimeters for Pharyngeal Depth Measured From Constructed PNS in Apert and Crouzon Patients in Comparison to the Norms of Subtelny (1957).

Age	*Subtelny norm*	*s.d.*	*+ 2 s.d.*	*Apert* $\bar{X}$	*Crouzon* $\bar{X}$
3 mos	14.7	2.9	8.9–20.5	16.2	9.7
6 mos	13.9	2.2	9.5–18.3	12.4	7.0
9 mos	14.6	1.8	11.0–18.2	9.7	12.8
1 yr	15.4	3.5	8.4–22.4	9.7	5.0
2 yrs	16.3	3.2	9.9–22.7	12.2	10.9
3 yrs	16.3	3.2	9.9–22.7	12.3	9.9
4 yrs	16.8	3.9	9.0–24.6	10.2	7.9
5 yrs	18.1	2.8	12.5–23.7	8.8	11.6
6 yrs	17.7	4.4	8.9–26.5	8.3	9.1
7 yrs	19.0	4.1	10.8–27.2	7.8	10.2
8 yrs	19.9	3.9	12.1–27.7	10.3	7.9
9 yrs	19.7	4.2	11.3–28.1	12.9	15.6
10 yrs	20.6	3.2	14.2–27.0	9.4	12.5
11 yrs	20.4	3.7	13.0–27.8	11.1	9.1
12 yrs	21.0	2.8	15.4–26.6	11.3	11.5
13 yrs	21.4	2.9	15.6–27.2	10.4	11.6
14 yrs	22.0	2.7	16.6–27.4	14.2	21.2
15 yrs	22.9	3.8	15.3–30.5	14.4	15.9
16 yrs	23.5	2.3	18.6–28.1	17.3	10.9
17 yrs	24.4	3.4	17.6–31.2	16.9	13.8
18 yrs	24.2	1.9	20.4–28.0	14.0	9.6

TABLE 9. Mean Values in Millimeters for Length of Anterior Cranial Base in Apert and Crouzon Patients in Comparison to Bolton Standards (1975).

Age	*Bolton standard*	*s.d.*	*+ 2 s.d.*	*Apert* $\bar{X}$	*Crouzon* $\bar{X}$
3 mos				46.0	47.8
6 mos				50.3	53.5
9 mos				52.9	53.4
1 yr	55.0	2.3	50.4–59.6	57.2	56.6
2 yrs	58.9	2.2	54.5–63.3	57.4	59.1
3 yrs	61.1	2.7	55.7–66.5	58.5	61.1
4 yrs	62.7	2.1	58.5–66.9	59.5	59.2
5 yrs	64.2	2.1	60.0–68.4	61.3	59.4
6 yrs	64.7	2.5	59.7–69.7	63.8	57.6
7 yrs	65.8	2.6	60.6–71.0	62.6	61.2
8 yrs	66.8	2.7	61.4–72.2	64.6	61.6
9 yrs	67.2	2.7	61.8–72.6	65.1	66.3
10 yrs	68.2	2.8	62.6–73.8	67.8	65.6
11 yrs	68.7	2.8	63.1–74.3	70.4	66.1
12 yrs	69.6	2.8	64.0–75.2	66.4	63.5
13 yrs	70.3	2.8	64.7–75.9	69.4	69.3
14 yrs	70.9	2.7	65.5–76.3	69.8	72.2
15 yrs	71.8	3.2	65.4–78.2	70.9	69.1
16 yrs	71.8	3.5	64.8–78.8	70.6	64.1
17 yrs	72.3	3.8	64.7–79.9	69.8	63.2
18 yrs	72.8	3.8	65.2–80.4	72.3	62.8

TABLE 10. Mean Values in Millimeters for Length of Posterior Cranial Base in Apert and Crouzon Patients in Comparison to Bolton Standards (1975).

Age	*Bolton standard*	*s.d.*	*+ 2 s.d.*	*Apert* $\bar{X}$	*Crouzon* $\bar{X}$
3 mos				24.1	24.8
6 mos				27.2	27.7
9 mos				28.8	29.8
1 yr	30.4	2.2	26.0–34.8	31.0	28.6
2 yrs	33.4	2.0	29.4–37.4	31.6	32.1
3 yrs	35.1	1.4	32.3–37.9	33.2	33.4
4 yrs	35.8	1.7	32.4–39.2	32.6	32.3
5 yrs	37.0	2.2	32.6–41.4	35.4	32.8
6 yrs	38.4	2.2	34.0–42.8	34.4	32.3
7 yrs	39.2	2.0	35.2–43.2	35.6	34.6
8 yrs	40.8	2.1	36.6–45.0	36.2	34.7
9 yrs	42.1	2.5	37.1–47.1	34.4	35.4
10 yrs	42.8	2.6	37.6–48.0	35.3	35.9
11 yrs	43.4	2.4	38.6–48.2	38.6	34.1
12 yrs	44.4	2.4	39.6–49.2	35.5	35.9
13 yrs	44.8	2.4	40.0–49.6	37.3	39.7
14 yrs	45.1	2.7	39.7–50.5	39.2	44.4
15 yrs	46.0	2.4	41.2–50.4	38.4	37.9
16 yrs	46.0	3.0	40.0–52.0	37.7	37.9
17 yrs	46.2	2.8	40.6–51.8	40.4	35.9
18 yrs	46.5	3.4	39.7–53.3	35.8	34.2

TABLE 11. Mean Values for Cranial Base Angle in Apert and Crouzon Patients.

Age	*Apert* $\bar{X}$	*s.d.*	*Crouzon* $\bar{X}$	*s.d.*
3 mos	145.8°	11.3°	144.6°	11.2°
6 mos	138.5°	9.9°	123.2°	0
9 mos	136.4°	9.8°	137.7°	8.5°
1 yr	131.3°	11.0°	137.8°	18.8°
2 yrs	135.4°	13.2°	134.1°	6.7°
3 yrs	139.5°	12.5°	131.2°	6.8°
4 yrs	134.6°	13.7°	131.4°	8.6°
5 yrs	129.9°	12.6°	131.9°	10.9°
6 yrs	132.5°	15.0°	137.2°	18.5°
7 yrs	129.6°	11.8°	134.7°	10.2°
8 yrs	138.2°	13.9°	127.9°	2.9°
9 yrs	133.1°	14.5°	134.6°	7.4°
10 yrs	128.4°	10.3°	132.8°	4.6°
11 yrs	141.1°	11.0°	134.6°	7.4°
12 yrs	127.9°	10.5°	136.7°	10.1°
13 yrs	132.7°	10.5°	134.4°	6.8°
14 yrs	129.7°	10.6°	131.6°	0
15 yrs	129.6°	9.0°	137.6°	10.2°
16 yrs	129.8°	12.2°	142.6°	11.5°
17 yrs	127.9°	8.3°	137.9°	15.3°
18 yrs	126.4°	14.1°	126.1°	7.7°

The plotted data suggest the following:

1. Soft palate length
   a. From the first months of life in both Apert and Crouzon infants, soft palate length is greater than the norm.
   b. By the teenage years, this deviation places the measurements for the Apert patients more than two standard devia-

tions above the mean and this remains generally true into the adult years.[1]

c. The trend for Crouzon patients is less clear.

2. Soft palate thickness

   Thickness of the soft palate remains above the normal mean from the age of five with no clear difference between Apert and Crouzon patients. Most of the means fall within two standard deviations of the normal mean.

3. Length of the hard palate

   a. Whether anatomic or constructed PNS is used as the posterior landmark, hard palate length is reduced by two or more standard deviations from the mean by the age of three years in Apert and six years in Crouzon patients.

   b. In both Apert and Crouzon patients, deviation from the norm becomes more marked with increasing age.

4. Pharyngeal height

   a. Although pharyngeal height in Crouzon patients remains generally below the norm at all ages, most measurements fall within two standard deviations from the mean.

   b. Apert patients also generally fall below the norm. By the age of 9 years, their means consistently fall below the 95% confidence interval.

   c. In the Apert patients, the reduction in this measurement from the norm increases with age.

5. Pharyngeal depth

   a. Whether anatomic or constructed PNS is used as the anterior landmark, pharyngeal depth is markedly reduced in comparison to the norm in both Apert and Crouzon infants by the age of one year.

   b. In the age range of five to 13 years, the reduction may be as much as 50% of the normal depth.

6. Anterior cranial base

   a. The averaged values tend to fall below the norm, with a particularly marked reduction for the Crouzon patients at ages five and six and again above the age of 16.

   b. Virtually all Apert means fall within two standard deviations of the normal mean.

7. Posterior cranial base

   The posterior cranial base is markedly foreshortened by the age of four years in both Apert and Crouzon patients and remains so at all ages. The deviation from the mean increases with age.

In addition to the generalizations based on the combined cross-sectional and longitudinal data, we offer selected serial cephalometric tracings on a single Apert patient (Figure 10) and a single Crouzon patient (Figure 11) to demonstrate the progression in these distortions as the patient matures.

Discussion

The utility or sufficiency of cephalometric radiography as a means of evaluating the morphology of the nasopharynx is currently both a popular and a controversial topic (Fujioka et al., 1979; Hibbert and Stell, 1979; Holmberg and Linder-Aronson, 1979; Poole et al., 1980; Sorensen et al., 1980; Vig and Hall, 1980). Certainly airflow studies, endoscopy, and multiview fluoroscopy contribute important information regarding the *physiology* of the system in respiration, deglutition, and speech. Cephalometric radiography represents a basic, first step in assessment of the *morphology*, affording accurate, repeatable measures of the sizes and relationships of structures and the anatomic changes which accompany growth. Serial tomography offers the advantage of additional information but at a cost of greatly increased radiation. Irradiation of these patients for any purpose must, of course, be well justified and carefully monitored since the very nature of their malformations necessitates pre- and postoperative radiographic studies, often on a repeated basis.

The overwhelming impression of the data reported here is one of significant crowding of the nasopharyngeal airway which is often present early in life and which tends to become more severe with age. In addition to the distortions revealed in the lateral films, there is evidence of significant diminution in nasopharyngeal width as well: Since the pterygoid plates which form the lateral bony boundaries

[1] Kaye et al. (1978) found the same accumulations of acid mucopolysaccharides which had been reported in the palatal vault by Solomon et al. (1973) to be present in the soft palate of Apert patients. It is speculated that such accumulations could account, in part, for the abnormal size of the velum.

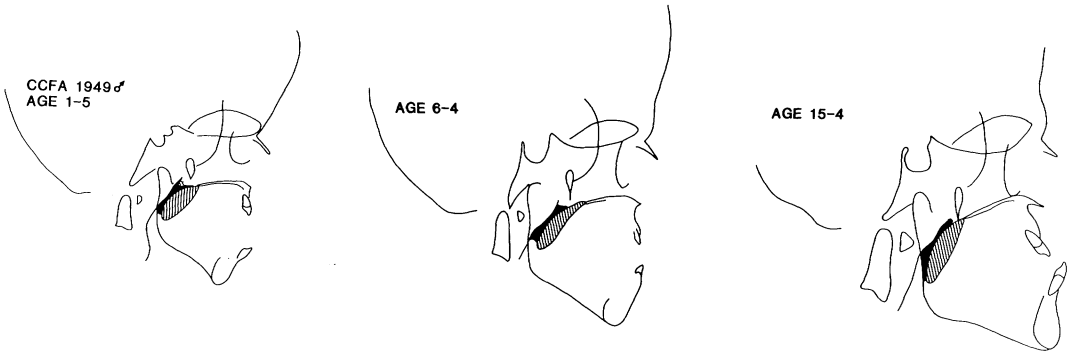

FIGURE 10. Selected serial tracings on a single Apert patient (CCFA 1949) at ages one, six and 15.

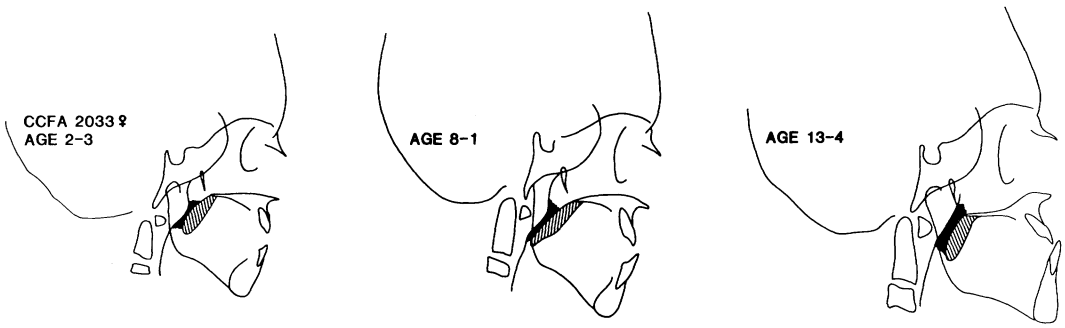

FIGURE 11. Selected serial tracings on a single Crouzon patient (CCFA 2033) at ages two, eight, and 13.

of the posterior choanae and walls of the pharynx abut against the maxillary tuberosities, an indirect measure of nasopharyngeal width was obtained by measuring the width across the maxillary tuberosities of the palatal casts. This measure was consistently reduced from the norm in Apert patients, indicating diminution of the nasopharyngeal space in the lateral as well as the A-P dimension.[2] Concomitantly, downward and forward displacement of the cranial base reduces the vertical height of the posterior choanae. This reduction in patency of the choanae can preclude utilization of nasendoscopy for examination of the nasopharynx.

The reduction in pharyngeal depth by as much as 50% in the age range of five to 13 may reflect a "peaking" of adenoid size during these years, at least in some patients. However, any consideration of surgical removal of adenoid tissue should be carefully weighed in balance with plans for advancement of the midface.

The current data appear to suggest greater reduction in length of the anterior cranial base in Crouzon than in Apert patients. The significance of this finding in concert with other measurements on the varying degrees of exorbitism in the two syndromes as a function of age is currently under investigation.

The cumulative effect of (1) a nasopharyngeal space reduced in three dimensions, (2) increased size of the velum, and (3) reduced height and width of the posterior choanae can be life-threatening for the infant and young child. Radiographic measurements alone cannot, of course, be taken as indication of a need for physical intervention, i.e., tracheostomy. Clinical signs such as excessively noisy respiration and evidence of sleep apnea will alert the physican to possible functional problems

[2] The validity of this approach is currently being investigated by comparing measurements from cases with measurements from coronal tomograms at the same level in selected cases.

in the individual patient. In some cases, the presence of an overt cleft palate may be an unexpected benefit, providing the patient with "additional" patency of the nasopharyngeal airway. Certainly surgical closure of a cleft in a child whose airway patency is suspect should not be undertaken without thorough preoperative studies.

Both Apert syndrome and Crouzon disease vary in severity, and management decisions must be based upon the clinical picture presented by the individual patient. The current data should alert the clinician (1) to some of the anatomic factors which may affect patency of the nasopharyngeal airway in these syndromes and (2) equally as important, to the time-dependent nature of both the appearance and the severity of these factors.

Conclusions

Results of this cephalometric study of the dimensions of the nasopharynx in the syndromes of Apert and Crouzon concur with information previously reported both from this and other centers, enlarging the data base to direct attention to changes in these dimensions with growth of the patient. Alterations of the nasopharyngeal architecture in these syndromes include reduction in pharyngeal height, width, and depth; increased length and thickness of the velum; decreased length of the hard palate; and marked reduction in the posterior cranial base with somewhat less remarkable changes in the anterior cranial base. Increased basilar kyphosis evident in some patients contributes to the reduction in nasopharyngeal space. Most of these deviations are present early in life and tend to become worse as the patient matures. The combination of reduced nasopharyngeal dimensions and reduced patency of the posterior nasal choanae poses the threat of respiratory embarrassment and cor pulmonale, particulary in the young child.

Acknowledgment: The authors gratefully acknowledge the assistance of M. Esler, D. Leu, R. Balmes, and A. Figueroa, D.D.S., in the preparation of illustrations.

Reprints: Sally J. Peterson-Falzone Ph.D.
Center for Craniofacial Anomalies
Box 6998 Chicago, Illinois 60680

References

ALEKSANDROYA, I., KHOREA, N., and PETRUNIA, G., Changes in facial skeleton in Apert-Crouzon's syndrome. *Stomatologiia* (Mosk), *58*, 1, 51–54, 1979.

BLANK, C., Apert's syndrome (a type of acrocephalosyndactyly)—observations on a British series of thirty nine cases. *Ann. Hum. Gen.*, *24*, 151–163, 1960.

BRAILSFORD, J., *The Radiology of Bones and Joints* (3rd edition). Churchill, 1944.

BUCHANAN, R., Acrocephalosyndactyly, or Apert's syndrome. *Br. J. Plast. Surg.*, *21*, 4, 406–418, 1968.

BUCKLEY, R., and YAKOVLEV, P., Dysostosis of skull, face and extremities (acrocephalosyndactyly). *Am. J. Dis. Child.*, *75*, 688–694, 1948.

EBEL, KL.-D., Craniostenosis. *Ped. Radiol.*, *2*, 1–14, 1974.

FERRIMAN, D., *Acrocephaly and Acrocephalosyndactyly.* Oxford University Press, 1941.

FUJIOKA, M., YOUNG, L., and GIRDANY, B., Radiographic evaluation of adenoid size in children: Adenoidal-nasopharyngeal ratio. *Am. J. Roentgen.*, *133*, 3, 401–404, 1979.

GIUFFRE, R., VAGNOZZI, R., and SAVINO, S., Infantile craniosynostosis: Clinical, radiological, and surgical considerations based on 100 surgically treated cases. *Acta Neurochir.*, *44*, 49–67, 1978.

GORLIN, R., and PINDBORG, J., *Syndromes of the Head and Neck.* New York: McGraw-Hill Book Co., 1964.

HIBBERT, J., and STELL, P., A radiological study of the adenoid in normal children. *Clinical Otolaryn.*, *4*, 321–327, 1979.

HOLMBERG, H., and LINDER-ARONSON, S., Cephalometric radiographs as a means of evaluating the capacity of the nasal and nasopharyngeal airway. *Am. J. Orthod.*, *76*, 5, 479–490, 1979.

KAYE, C., MATALON, R., and PRUZANSKY, S., The natural history of Apert syndrome with speculations on pathogenesis (abstract). *Teratology*, *17*, 2, 28A, 1978.

MATRAS, H., WATZEK, G., and PERNECSKY, A., Cephalometric observations in premature craniosynostosis. *J. Maxillofac. Surg.*, *5*, 298–303, 1977.

MCCARTHY, J., COCCARO, P., and SCHWARTZ, M., Velopharyngeal function following maxillary advancement. *Plast. Reconst. Surg.*, *64*, 2, 180–189, 1979.

MCGREGOR, M., The significance of certain measurements of the skull in the diagnosis of basilar impression. *Br. J. Radiol.*, *2*, 171–181, 1948.

PETERSON-FALZONE, S., PRUZANSKY, S., LAFFER, J., and PARRIS, P., Longitudinal changes in palatal configuration in the syndromes of Apert and Crouzon. Presented before the American Cleft Palate Association, Atlanta, 1978.

PETERSON, S., and PRUZANSKY, S., Palatal anomalies in the syndromes of Apert and Crouzon. *Cleft Pal. J.*, *11*, 394–402, 1974.

POOLE, M., ENGEL, G., and CHACONAS, S., Nasopharyngeal cephalometrics. *Oral Surg.*, *49*, 3, 266–271, 1980.

PRUZANSKY, S., Radiocephalometric studies of the basicranium in craniofacial malformations. In Bosma, J. (Ed.), *Symposium on Development of the Basicranium.* HEW Publications, 1977.

SOLOMON, L., MEDENICA, M., PRUZANSKY, S., and KREIBORG, S., Apert syndrome and palatal mucopolysaccharides. *Teratology*, *8*, 287–292, 1973.

SORENSEN, H., SOLOW, B., and GREVE, E., Assessment of the nasopharyngeal airway. *Acta Otolaryng.*, *89*, 227–232, 1980.

TESSIER, P., The definitive plastic surgical treatment of the severe facial deformities in craniofacial dysostosis. *Plast. Reconst. Surg.*, *48*, 419–442, 1971.

VIG, P., and HALL, D., The inadequacy of cephalometric radiographs for airway assessment (letter to the editor). *Am. J. Orthod.*, *77*, 230–232, 1980.

WITZEL, M., and MUNRO, I., The effects of facial osteotomies on speech and velopharyngeal function. Presented before the Third International Congress on Cleft Palate and Related Craniofacial Anomalies, 1977.