Long-Term Effects of Premaxillary Excision in Patients with Complete Bilateral Cleft Lips and Palates

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In 1972, Friede and Pruzansky reported on a longitudinal study of complete bilateral cleft lip and palate wherein they divided the populations into subgroups with prognostic distinctions. Utilizing an enlarged sample of 107 patients studied over an even longer period of time, the present report focuses on three patients subjected to premaxillectomy. The rationale for this radical departure from usual and customary treatment and the long-term outcome are analyzed. The results emphasize the importance of considering variations in contiguous structures in treatment planning.

KEY WORDS: Bilateral cleft lip, bilateral cleft palate, premaxillectomy

Introduction

Among the findings that have emerged from the longitudinal studies of children with facial clefts at the Center for Craniofacial Anomalies at the University of Illinois are two concepts that are fundamental to clinical management and research design. The first concept is that not all clefts are alike and that even within a single cleft type there is sufficient variation between patients to affect prognosis (Pruzansky, 1953). The second concept is that not all patients with the same type of cleft possess similar genotypes (Pruzansky, et al., 1973).

In 1972, the first in a series of papers reporting on the natural history of complete bilateral cleft lip and palate was published by Friede and Pruzansky (1972). In a series of 54 patients with unoperated complete bilateral cleft lip and palate (C-BCLP), they found nearly a two-fold variation in the extent of premaxillary protrusion. This variance was predictive of the subsequent state at school age. It was also shown that the increments and direction of mandibular growth could influence the degree and speed of resolution of facial convexity characteristic of this cleft type.

This report demonstrates that variation in the shape and growth pattern of the cranial base and mandible can produce variable outcomes of treatment for seemingly similar complete bilateral clefts of the lip and palate.

Stated another way, two patients with identical clefts operated upon by the same surgeon utilizing similar operative techniques may vary markedly in outcome because of difference between the patients beyond the influence of surgery. It follows that treatment planning has to be individualized to conform with variants in the total craniofacial complex and that the growing child must be monitored to assess the effect of individual growth patterns on the subsequent state.

The basis of this report is a continuing retrospective study of three male patients with C-BCLP subjected to total excision of the premaxilla at ages eight years and eight months, five years and 10 months, and five years and eight months. Experience and a priori reasoning had taught us that premaxillectomy is a radical procedure with predictably deleterious long-term effects that include a concave midfacial skeletal profile, lack of support for the upper lip and nose, and dis-
turbances in arch form with impediments to masticatory function and speech. Despite this foreknowledge, one of us (S.P.) recommended this radical operation. What distinguished our three cases was that none of the deleterious effects occurred. Since the three patients were followed in the longitudinal study with serial data from birth to adulthood, we considered the following questions:

1. What motivated the excision of the premaxilla in the first place?
2. Was the wisdom of this decision sustained as the patients reached adulthood?
3. In what ways did the three patients who underwent premaxillectomy at our Center differ from 9 patients who underwent premaxillectomy elsewhere and were referred to us for rehabilitation because of their poor outcome?
4. What distinguished the three patients who underwent premaxillectomy at our Center from other patients with C-BCLP at our Center who were not subjected to presurgical maxillary orthopedics and experienced neither surgical setback of the premaxilla nor premaxillectomy?
5. Utilizing the information derived from this study, is it possible to define future candidates for premaxillectomy by examining their initial state? If not, at what age is it possible to make such a decision with confidence?

Method

Subjects. At this writing, our serial study includes 107 patients with C-BCLP (75%, 329) for whom the data base begins prior to all surgical intervention. Of this number, four male patients underwent premaxillectomy by staff decision (Friede and Pruzansky, 1972). One of the patients was eliminated from this study (CCFA 38) because the premaxillectomy was not complete in that the basal bone of the anterior nasal floor, including the anterior nasal spine, was retained. For purposes of this investigation, we formed the following groups for comparative purposes:

Group I The experimental group consisted of three male subjects with C-BCLP who had undergone total premaxillectomy.

Group II Ten male subjects with C-BCLP whose premaxillae were not excised or setback and who did not receive presurgical orthopedics. The long-term outcome for these patients was judged to be satisfactory in terms of the facial skeletal profile.

Group III From our cross-sectional studies, 9 male C-BCLP patients were available whose premaxillae were removed elsewhere. These were referred to our Center as adults for rehabilitation because of severe midface hypoplasia.

Group IV Normative control data for the older age periods from publications by the University of Michigan (Riolo et al., 1974) and the Bolton study (Broadbent et al., 1975) at Case Western Reserve University.

Procedures. The infant casts utilized in this study were described by Pruzansky (1953), and the techniques of roentgencenchalometry of infants were reported by Pruzansky and Lis (1958). Since Friede and Pruzansky (1972) have already defined the anthropometric parameters for quantifying the C-BCLP deformity, only those roentgencenchalometric measurements relevant to this investigation will be described herein (Figure 1).

1. Shape of the Cranial Base
   (a) The angle nasion-sella-basion (N-S-Ba) is a measure of the flexure of the cranial base.

This measurement can affect the spatial relationship of mandible to maxilla because the latter is attached to the anterior cranial base while the mandible articulates with the middle cranial fossa. All other factors being equal, an excessive cranial base angle (platybasia) will position the mandible posteriorly, while an acute cranial base angle (basilar kyphosis) will project the mandible anteriorly.

2. Size and Shape of the Mandible
   (a) The linear measurement gonion-pogonion (Go-Pg) is a measure of mandibular body length.
   (b) The linear measurement articulare-gonion (Ar-Go) is a measure of mandibular ramus height.
   (c) Gonial angle—the angle achieved through intersection of the plane Ar-Gol with the plane Gol-Me (Riolo et al., 1974). Alternatively, the gonial angle can be defined by the intersection of plane Ar-Go with plane Go-Gn (Broadbent et al., 1975).

3. Spatial relationship of the mandible to the cranial base.
   (a) The angle sella-nasion-pogonion (S-N-Pg) is a measure of the protrusion of the
Points, angles, and planes employed in the evaluation of the craniofacial complex

Angular measurements

1. $\angle N-S-Ba$
2. $\angle S-N-Pg$
3. $\angle N-S-Gn$
4. $\angle Ar-Go-Gn$
5. $\angle Ar-Gol-Me$

Linear measurements

a. Ar-Go
b. Go-Pg

FIGURE 1. Index of roentgencephalic landmarks and angular and linear measurements employed in this study.

(a) The angle nasion-sella-gnathion (N-S-Gn) is a measure of the rotation of the chin relative to the cranial base.

(b) The angle nasion-sella-gnathion (N-S-Gn) is a measure of the rotation of the chin relative to the cranial base.

Group I—Summary Case Reports

(CCFA 449) White, male, second born at full term to a gravida 2, para 1, ab 4 mother. Birth weight 2295 gms. No other malformations were noted. Longitudinal data available from age two months to 10 years (Figures 2 through 4).

At three months of age, the left side of the lip was repaired and was followed at age 9 months by surgical setback of the premaxilla with simultaneous repair of the right side of the lip. At age two years, the clefts of the hard and soft palates were repaired. Tonsillectomy and adenoidectomy were performed at age six. At age eight years and eight months, premaxillectomy and pharyngoplasty were performed. Between ages 13 and 16, he underwent orthodontic treatment elsewhere. An Abbe flap was constructed at age 17-1/2 with secondary reconstruction of the nose at 18 years of age and pharyngeal flap revision at age 19.

He was subsequently fitted with a multibutment fixed prosthesis described by Gold and Pruzansky (1979). His speech was characterized as being inconsistently hypernasal with multiple articulation errors. The patient declined speech therapy. Mild bilateral conductive hearing loss was reported.

(CCFA 742) White, male, full-term, birth weight 3626 gms. born to gravida 6, para 5, ab 0 mother. The patient was sixth in birth order. Mother was 29 years old and father 36 at birth. No other malformations were present. Serial records were available from age two months to 18 years (Figures 5 through 7).

At eight months of age, he underwent a single-stage repair of the bilateral cleft lip. Closure of the soft palate was performed at 26 months of age. Premaxillectomy and closure of the cleft of the hard palate were carried out at age five years and 10 months. Lip revision was performed at age seven years, seven months. Closure of an oronasal fistula and columella lengthening were done at age 12 years, 10 months. He wore a removable prosthesis to replace the missing maxillary incisors. Because of poor hygiene and general indifference toward further therapy, orthodontic treatment and fixed dental prosthesis were not recommended.

(CCFA 751) White, male, born to a gravida 4, para 3, ab 0 mother. Normal delivery at full term. Birth weight was 3683 gms. Both parents were 31 years of age. No other malformations. Serial records were available from age seven weeks to 19 years (Figures 8 and 9).

At four months of age, he had bilateral lip closure in a single stage followed by palatal surgery at 15 months of age. The premaxillectomy was performed at age five years and 8 months. Millard revisions of the lip were carried out in two operations the following year. During the next two years, he underwent elongation of the columella by composite graft from the right ear and further lip revision. At age 15, he had additional nasal reconstruction with submucous resection. The next year he
FIGURE 2. (CCFA 449) Unoperated complete bilateral cleft lip and palate at age two months.

FIGURE 3. (CCFA 449) Patient in Figure 2 at 20 years and four months of age.

had an Abbe flap. Further nose revision occurred during his seventeenth year.

Orthodontic treatment was carried out between 10 and 13 years of age. He was fitted with a removable prosthesis to replace the incisor teeth.

Although the cosmetic and functional results in terms of soft tissue reconstruction and freedom from hypernasality were less than ideal, we noted that there was no midface skeletal hypoplasia or concavity.

Results

For the sake of clarity, the results of our analysis will be presented as answers to the questions posed in the introduction.

1. Why did we elect premaxillectomy?

All three patients were severely handicapped by gross facial malformations resulting from severely projecting premaxillae. They were at a critical stage of their social development. Two were about to enter the school system, and the eldest of the three had already suffered the taunts of schoolmates for more than two years.

All three patients exhibited Class II malocclusions (distocclusion) of the buccal dentaion suggesting that the mandible was relatively posterior to the maxilla. This was confirmed by roentgencephalometric studies.

In all three cases, the prior growth record
FIGURE 4. (CCFA 449) Selected serial cephalometric radiographs beginning at age two months prior to surgical intervention; at age 9 years and three months following excision of premaxilla, and at age 20 years and four months with fixed prosthesis in place.

demonstrated unequivocally that the skeletal pattern was retrognathic and that this accentuated the premaxillary protrusion. Further, the growth pattern suggested that this state would persist and even grow worse because of downward and backward mandibular growth and the associated platybasia.

In one instance (CCFA 449), surgical premaxillary setback did not resolve the problem. This procedure was not considered in the other two cases because the degree of the facial convexity could not be significantly reduced by setback. Orthopedic manipulation was not considered.

In view of the magnitude of the deformity and the consequent functional and psychosocial handicap combined with the poor skeletal pattern and growth prognosis, there seemed to be no alternative to total excision of the premaxilla.

2. Was the wisdom of this decision sustained as the patient reached young adulthood?

The answer is yes. In evidence, we point to the final lateral cephalometric radiographs which did not exhibit the expected severe midface concavity. In addition, the adverse skeletal and growth pattern, noted earlier, persisted into adulthood (Table 1).

3. Comparison of Group I and Group III.

The three patients in our series who underwent premaxillectomies (Group I) were compared with the 9 adult male patients (Group III) whose premaxillectomies were performed elsewhere and who were referred to us for
rehabilitation because of severe midface hypoplasia. Clearly, the subjects in Group III represent a biased group since their ascertainment was predicated on the existence of a deformity presumed to evolve from the premaxillectomy. However, this bias does not negate the point to be made. Did they differ from our patients in the configuration of adjacent structures and if so, did these differences account for the deformity in Group III and relative lack of deformity in Group I?

Again, the answer is yes on both counts. The patient in Group III showed more severe midface hypoplasia than those in Group I. This can be attributed to the fact that the development of contiguous anatomy in Group III was closer to the norm and closer to Group II patients who did not undergo premalignectomy and ended with a relatively satisfactory profile (Table 2).

4. How did the patients in Group I compare with those in Group II?

It was of interest to compare the three patients who underwent premaxillectomy (Group I) with ten male patients with similar clefts subjected to surgery of the lip and palate without presurgical orthopedics, premaxillary setback, or excision- (Group II). Comparing the data longitudinally, it is clear that those who underwent premaxillectomy had a more obtuse cranial base angle throughout their growth period. The growth axis of the mandible relative to the cranial base (N-S-Gn) demonstrated a retrognathic pattern for Group I. The chin relative to the facial profile was more retrognathic in Group I (S-N-Pg); while in two of the three cases in Group I, linear measurements showed the mandible in Group I to be smaller (Table 2).

Discriminant analysis of the cross-sectional data at age 18 years revealed that, on the basis of three parameters, it was possible to discriminate among the three groups. The critical measures involved the cranial base and those defining the position of the mandible relative to the cranial base. All of these measurements showed the patients in Group I to have greater mandibular retrognathia than those in Group II.

3. Although candidates for premaxillectomy are rare, is it possible to determine who they are by examining their initial state? If not, at what age does it become evident that such a radical procedure is necessary?

To answer this question, we reviewed the measurements published by Friede and Pru-
zansky (1972) on 54 unoperated patients with complete bilateral clefts of the lip and palate. The three patients in Group I were included in this series. From the tabulated comparison (Table 3), it is clear that two of the three patients were distinguished by their more severe retrognathia (S-N-Pg) and by their greater convexity of the facial skeletal profile (N-\(\chi\)-Pg). Although Friede and Pruzansky did not report on the cranial base angle (N-S-Ba), in the present study it was noted that by age one year, Group I already exhibited platybasia (Table 1).

The hindsights provided by these studies indicate that, in the initial state, we could detect faint signals suggesting that we were dealing with an infant whose mandible would not grow as much or in a direction favorable for minimizing the premaxillary protrusion. As we monitored the growth of our patients, their differences from the group became more apparent with increasing age. This is understandable, for initial minor differences in size and growth rate, if sustained, become more obvious with increasing age. As the children reached school age, their differences were fully revealed, and the decision to take the heroic step of premaxillectomy became unavoidable.

Discussion

The danger of lumping together patients with similar diagnosis for clinical investigative purposes has been stressed in previous publications from this Center. The hazard resides in the marked variation in severity within a
single cleft type. In the present report, we stress yet another variable, that found in the growth pattern of contiguous structures. Since the morphology of contiguous structures can minimize or maximize the dysmorphology of the primary defect, the need to consider such variables becomes clear.

This variability must be understood in relationship to clinical management since the long-term therapeutic outcome can be dependent upon factors not under the control of the surgeon. Individualized treatment planning demands long-term monitoring and documentation by the quantitative means illustrated herein.

Since this report has focused on the radical decision to perform premaxillectomy, we want to demonstrate also how similar documentation was used to disuade the surgeon from undertaking a premaxillectomy or a premaxillary setback because of a clearly favorable growth pattern. For this purpose, we selected one case out of a larger series of similar background.

A white male with complete bilateral cleft lip and palate was followed from the age of 10 months. When he was referred for orthodontic treatment at age eight, he showed marked facial convexity, incisal overjet of 12.5 mm., deep overbite, and class II malocclusion (distoocclusion). Although premaxillary setback had been urged on several occasions, we resisted this recommendation because his growth pattern was judged to be favorable. Accordingly, we instituted conventional orthodontic treatment. The lateral film obtained at the conclusion of orthodontic treatment reflects our success and demonstrates the contribution of mandibular growth to the resolution of the extreme convexity of the skeletal profile (Figure 10).

We have justified premaxillectomy in three
Motohashi and Pruzansky, eraxiLLARY
[46x628]CFA 75184
AGE 19 -O
p"
[52x400]FIGURE 9. (CCFA 751) Photographs of patient in
Figure 8 reveal that with prosthesis replacing missing
teeth in the at-rest position, concavity of midface profile
is minimized.

Summary
Utilizing longitudinal information on 107
patients with complete bilateral cleft lip and
apalate, the data on three male subjects who
underwent total premaxillectomy at early
school age were reviewed to determine out-
come in adulthood. The decision to take this
heroic step was dictated by the handicapping
effects of the severe facial convexity and the
special cases. However, this is not a generally
recommended procedure and should not be
so interpreted. We elected to take this step in
only four out of 107 patients with C-BCLP. It
should not be assumed, however, that a simi-
lar percentage would be found by others. It
may be that there is an ascertainment bias in
our sample similar to the more complex cases
referred to our Center.

Finally, we must reiterate the caveat that
no one should resort to premaxillectomy un-
less evidence similar to that presented here is
available to ascertain when mother nature works
to the advantage or disadvantage of the pa-
tient and to plan treatment accordingly.

TABLE 1. Group I, consisting of three patients who underwent premaxillectomy, is compared with matched Group II, consisting of 10 similar patients who did not undergo premaxillectomy or surgical setback. Note that three variables, N-S-Ba, S-N-Pg, and N-S-Gn, remain distinctive throughout the growth period. These findings attest to the constancy of the platybasia and mandibular retrognatia.

<table>
<thead>
<tr>
<th>Measurements</th>
<th>Group I (3) C-BCLP</th>
<th>Group II (10) C-BCLP</th>
<th>t-test</th>
<th>Group I (3) C-BCLP</th>
<th>Group II (10) C-BCLP</th>
<th>t-test</th>
<th>Group I (3) C-BCLP</th>
<th>Group II (10) C-BCLP</th>
<th>t-test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age in Years</td>
<td>1 year</td>
<td></td>
<td>6 year</td>
<td></td>
<td></td>
<td>16 year</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>C-BCLP</td>
<td></td>
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<td>C-BCLP</td>
<td></td>
<td></td>
<td>C-BCLP</td>
<td></td>
<td></td>
</tr>
<tr>
<td>N-S-Ba</td>
<td>145.0 ± 4.4</td>
<td>136.4 ± 3.3</td>
<td>*</td>
<td>141.4 ± 4.9</td>
<td>134.4 ± 3.1</td>
<td>*</td>
<td>141.2 ± 7.5</td>
<td>131.5 ± 4.4</td>
<td>*</td>
</tr>
<tr>
<td>S-N-Pg</td>
<td>59.5 ± 1.8</td>
<td>63.2 ± 2.0</td>
<td>*</td>
<td>64.8 ± 2.0</td>
<td>71.3 ± 3.2</td>
<td>*</td>
<td>68.7 ± 2.1</td>
<td>74.6 ± 3.3</td>
<td>*</td>
</tr>
<tr>
<td>N-S-Gn</td>
<td>79.7 ± 3.2</td>
<td>73.2 ± 3.2</td>
<td>*</td>
<td>79.3 ± 3.4</td>
<td>69.8 ± 4.4</td>
<td>*</td>
<td>79.3 ± 3.1</td>
<td>71.1 ± 3.0</td>
<td>*</td>
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<tr>
<td>Ar-Go-Me</td>
<td>142.7 ± 1.5</td>
<td>132.7 ± 7.5</td>
<td>*</td>
<td>140.5 ± 6.5</td>
<td>134.0 ± 5.9</td>
<td>*</td>
<td>130.7 ± 4.6</td>
<td>129.4 ± 3.7</td>
<td>*</td>
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<tr>
<td>Ar-Go</td>
<td>27.7 ± 1.2</td>
<td>28.7 ± 1.6</td>
<td>*</td>
<td>34.9 ± 3.2</td>
<td>38.7 ± 2.3</td>
<td>*</td>
<td>48.1 ± 4.0</td>
<td>50.0 ± 4.3</td>
<td>*</td>
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<tr>
<td>Go-Pg</td>
<td>41.1 ± 1.6</td>
<td>42.9 ± 3.0</td>
<td>*</td>
<td>59.5 ± 3.4</td>
<td>59.6 ± 3.8</td>
<td>*</td>
<td>76.9 ± 5.1</td>
<td>75.4 ± 6.4</td>
<td>*</td>
</tr>
</tbody>
</table>

* Student t-test—statistically significant differences at 0.05%
Group I: Premaxillectomy (n = 35)
Group II: C-BCLPs without premaxillectomy or setback (n = 105)
Values in Table 1 represent means and standard deviations.
TABLE 2. Group I is compared with Groups II, III, & IV. Group I is distinguished from the other groups by virtue of the platybasia (N-S-Ba) and retrognathia (S-N-Pg and N-S-Gn). The patients who underwent premaxillectomy elsewhere with consequent deleterious effects (Group III) resembled Groups II and IV more than they did Group I.

<table>
<thead>
<tr>
<th>Group I-Initial State</th>
<th>Age</th>
<th>P1-P2</th>
<th>ml-m2</th>
<th>α-M</th>
<th>SNa</th>
<th>SNPg</th>
<th>NaPg</th>
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<tr>
<td>CCFA*</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>449</td>
<td>1m, 26d</td>
<td>18.2</td>
<td>13.5</td>
<td>16.8</td>
<td>95°</td>
<td>57°</td>
<td>112°</td>
</tr>
<tr>
<td>742</td>
<td>2, 10d</td>
<td>20.4</td>
<td>11.2</td>
<td>17.7</td>
<td>99°</td>
<td>58°</td>
<td>112°</td>
</tr>
<tr>
<td>751</td>
<td>1m, 0d</td>
<td>15.7</td>
<td>16.1</td>
<td>15.4</td>
<td>95°</td>
<td>61.5°</td>
<td>117°</td>
</tr>
<tr>
<td>Mean ±1 s.d.</td>
<td>1.7 ± 0.7</td>
<td>18.1 ± 2.35</td>
<td>13.6 ± 2.45</td>
<td>16.6 ± 1.20</td>
<td>96.3 ± 2.31°</td>
<td>58.8 ± 2.36°</td>
<td>113.7 ± 2.89°</td>
</tr>
</tbody>
</table>

*Measurements of casts and radiographs at the initial stage (Friede and Pruzansky, 1972).
recognition of factors contributing to mandibular retrognathia. This study demonstrates the importance of considering variation in contiguous structures in diagnosis and treatment planning. In treatment, no less than in research design, it must be recognized that not all clefts of a single type are alike, nor are patients with similar clefts alike. Attention to variability in form through the growth period of the patient is essential for optimal care.

References


