Surgical Treatment of Craniosynostosis: Outcome Analysis of 250 Consecutive Patients

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ABSTRACT. Objective. Surgery for craniosynostosis has evolved rapidly over the past two decades, with increased emphasis on early, extensive operations. Older published series may not accurately reflect more recent experience. Our study was designed to analyze outcome in a large series of consecutive patients treated recently at a single center.

Methods. We reviewed 250 consecutive patients who underwent surgical treatment of craniosynostosis between January 1, 1987 and December 31, 1992. They were divided into nine groups by suture involvement: sagittal, unilateral coronal, bilateral coronal, unilateral lambdoid, bilateral lambdoid, metopic, multiple suture, the Kleeblattschädel deformity (cleft/leaf skull), and acquired craniosynostosis. Outcome was analyzed in terms of residual deformities and irregularities, complications, mortality, as well as the need for additional surgery.

Results. There were 157 males (62.8%) and 93 females (37.2%), with most of the male preponderance accounted for by the large sagittal synostosis group, which consisted of 82 males and 25 females. Median age at first operation was 147 days. A named syndrome was present in 23 patients (9.2%) and was more common than expected with bilateral and unilateral coronal synostosis, the Kleeblattschädel deformity, and multiple suture synostosis. There were two deaths (0.8%), both with Kleeblattschädel patients, and 17 other complications (6.8%). Morbidity and mortality were significantly associated with secondary vs primary operations and syndromic vs nonsyndromic patients. Outcome analysis revealed the best surgical results with metopic synostosis and significantly less good results with the Kleeblattschädel deformity, multiple suture synostosis, and bilateral coronal synostosis.

Conclusions. Using modern surgical techniques, craniosynostosis can be corrected with good outcomes and relatively low morbidity and mortality, particularly for otherwise healthy, nonsyndromic infants. Pediatrics 1997;100(1). URL: http://www.pediatrics.org/cgi/content/full/100/1/e2; craniosynostosis, craniofacial anomalies, craniofacial surgery, facial deformities.

MATERIALS AND METHODS

A total of 260 patients underwent a surgical procedure for craniosynostosis at Childrens Hospital Los Angeles between January 1, 1987 and December 31, 1992. Seven patients were excluded because their first operation for craniosynostosis had been performed before January 1, 1987. Three other patients were excluded because their first operation, although falling within the 6-year study period, was performed elsewhere. The remaining 250 patients had undergone initial surgical treatment of craniosynostosis.
at Childrens Hospital Los Angeles between January 1, 1987 and December 31, 1992.

The medical records of the 250 patients were reviewed. Data were collected, including name, medical record number, date of birth, gender, involved sutures, other medical diagnoses, dates of all surgical procedures performed for craniosynostosis, complications, dates of follow-up visits, findings at follow-up, and the most recent assessment of outcome. Any other relevant data were also noted.

Data were analyzed for the entire patient group, as well as for nine subgroups based on suture involvement: sagittal, unilateral coronal, bilateral coronal, unilateral lambdoid, bilateral lambdoid, metopic, multiple suture, Kleeblattschädel, and acquired. The Kleeblattschädel group consisted of those patients with the classic cloverleaf skull and total sutural synostosis. The multiple suture synostosis group was defined as those patients with synostosis of more than one suture, excluding bilateral coronal, bilateral lambdoid, and Kleeblattschädel patients.

To be able to analyze surgical results, we developed a seven-category classification system (Table 1). In this system, classes 1 through 4 represent good to excellent overall correction of the deformity, but with varying degrees of minor visible and/or palpable irregularities: none in class 1, palpable but not visible irregularities in class 2, visible irregularities in class 3, and requiring reoperation in class 4. Examples would include a palpable but not visible surgical wire in the temporal region (class 2), a visible and palpable bone spicule in the forehead (class 3), or a visible and palpable surgical plate requiring surgical removal because of impending exposure (class 4). Classes 5 through 7 represent patients with significantly compromised correction: not requiring further surgery in class 5, requiring further surgery in class 6, and believed by the surgeon to require reoperation by the surgeon, but family declines further surgery in class 7.

### RESULTS

**Suture Involvement**

Distribution of patients by suture involvement is shown in Table 2. By far the largest group was sagittal synostosis, 107 patients (42.8%). Next most frequent were multiple suture synostosis (12.0%), unilateral lambdoid synostosis (12.0%), and unilateral coronal synostosis (11.2%).

**Gender**

Gender distribution, presence of an identified syndrome, and median age at first operation are shown in Table 3. There was a clear male preponderance among patients with sagittal synostosis (76.6%) (P < .0001). However, the bilateral coronal synostosis patients were 76.2% female (P < .05). Taking the entire group of 250 craniosynostosis patients, there were 64 more males. Almost the entire difference can be explained by the sagittal synostosis group, which had 57 more males than females.

**Syndrome Diagnosis**

More than half of the bilateral coronal synostosis patients, 12 of 21, or 57.1%, carried the diagnosis of a specific named syndrome (P < .001). Six of the twelve had Crouzon, four Apert, one Antley-Bixler, and one Pfeiffer syndrome. A named syndrome was present in 4 of 30 multiple suture synostosis patients or 13.3% (P < .001), with three having Crouzon and one Apert syndrome. Two of the six Kleeblattschädel patients, or 33.3% (P < .01), carried a syndrome diagnosis, one Antley-Bixler and one Pfeiffer. Two unilateral coronal synostosis patients had syndrome diagnoses, one Baller-Gerold and one Saethre-Chotzen. Additionally, there was one metopic synostosis patient with Klippel-Feil syndrome, one unilateral lambdoid synostosis patient was a 47 xxx female, and one sagittal synostosis patient had the Goldenhar variant of the facio-auriculo-vertebral spectrum (Fig 1).

It is striking that the Kleeblattschädel patients were operated on at a much earlier age (median 25 days) than any other group. Because of the total sutural involvement and high risk of resultant damage to the central nervous system from increased intracranial pressure, these patients were treated quite urgently. The sagittal synostosis group was the next youngest group at the time of initial surgery (median 101 days). Many of those patients were treated by sagittal craniectomy using the technique that has been described by McComb. Some of the younger (less than 6 months) patients in the sagittal synostosis group had a strip craniectomy. Some of
the older patients underwent the $\pi$ or reverse $\pi$ procedure.\footnotemark[10]

**Complications**

Complications, deaths, and number of patients undergoing planned as well as unplanned reoperation are shown in Table 4. The only two deaths, as well as two nonfatal complications, occurred in the Kleeblattschädel group. One of the deaths was a male infant with Antley-Bixler syndrome, who underwent radical posterior craniectomy at 9 days, followed by bilateral frontal-orbital advancement and remodeling at 18 days. He subsequently developed turricephaly, for which barrel stave osteotomies were performed at 16 months. He died, shortly after that surgery, of acute brain herniation. The second death was a male infant with Pfeiffer syndrome who underwent posterior craniectomy at 25 days, which was complicated by an intraoperative dural sinus hemorrhage requiring transfusion of 2 units of packed red blood cells and 40 units of platelets. At 3 months, bilateral frontal orbital advancement and remodeling was performed, which was followed by postoperative hydrocephalus, requiring ventriculo-peritoneal shunting 2 months later. On follow-up examination at 14 months, there was found to be no evidence of head growth since the shunt placement. The infant was developmentally delayed. Posterior vault remodeling was performed and was complicated by venous sinus hemorrhage. This was controlled intraoperatively, but the patient died of cerebral edema 3 days later.
Five of 21 bilateral coronal synostosis patients (23.8%, \( P < .001 \)) had complications, which included tongue edema requiring reintubation, a dural and cortical laceration resulting in a postoperative cerebrospinal fluid leak that resolved, femoral artery thrombosis at a catheter site, a subdural fluid collection that required shunting, and a pyogenic granuloma at the surgical incision. Four of 30 multiple suture synostosis patients (13.3%, \( P < .001 \)) had complications: postoperative apnea requiring reintubation, postoperative seizures, severe conjunctival edema and herniation, and superficial stitch abscesses along the incision. Two of 30 unilateral lambdoid synostosis patients (6.7%, \( P < .05 \)) had complications: a unilateral supranuclear facial palsy that resolved 8 months after surgery, and Salmonella sepsis in a patient whose father had a diarrheal illness shortly before surgery. Four of 107 sagittal synostosis patients (3.7%, \( P < .001 \)) had complications: pneumonia, postoperative hydrocephalus requiring shunting, intraoperative metabolic alkalosis, and a superficial wound infection.

Unplanned reoperation was required in 18 of the 250 patients (7.2%). Specifically, unplanned reoperation was needed in two Kleeblattschädel (33.3%), four bilateral coronal (19.0%), five multiple suture (16.7%), two unilateral coronal (7.1%), two unilateral lambdoid (6.7%), one metopic (5%), and two sagittal synostosis patients (1.9%).

There were 17 nonfatal complications and 2 deaths, a combined morbidity and mortality rate of 19 of 250 patients (7.6%) or 19 of 297 operations (6.4%). For 250 primary operations, there were 10 complications and no deaths (a complication rate of 4.0%). For 47 secondary or tertiary operations, there were two deaths and seven other complications, for a complication rate of 14.9% (\( P < .01 \) compared to primary operations by \( \chi^2 \) analysis), and a combined morbidity and mortality rate of 19.1% (\( P < .001 \)).

Analyzing morbidity and mortality by the presence of an associated syndrome, it is striking that...
both deaths and 9 of 17 nonfatal complications occurred in syndromic patients. Because our entire patient group included 23 patients with syndrome diagnoses, the mortality rate was 8.7% for syndromic patients and 0% for nonsyndromic patients. The nonfatal complication rate was 39% for syndromic patients as opposed to 3.5% for patients who did not carry a syndrome diagnosis ($P < .0001$).

### Multiple Suture Synostosis

The multiple suture synostosis group is a particularly interesting and challenging group of patients that has not been well described elsewhere (Fig 2). In this group, we include all patients with synostosis of two or more cranial sutures other than isolated bilateral lambdoid synostosis, bilateral coronal synostosis, or the classic Kleeblattschädel (clover leaf skull) deformity, which three groups are classified separately. In our series, this group was surprisingly large, consisting of 30 patients, or 12% of the entire population. Such a large number of unusual suture combinations may reflect either increased awareness and better ability to diagnose multiple suture involvement with modern imaging techniques, or a selection bias based on referral to a tertiary care center. The specific sutures involved for those 30 patients are listed in Table 5. The most common combination was 10 patients with unilateral coronal and unilateral lambdoid synostosis. It is striking that the involvement of the two sutures was ipsilateral in all 10 patients; the right side for 7 patients and the left for 3 patients. The resulting deformity was usually quite severe (Fig 3). We have never seen a patient with unilateral coronal and unilateral lambdoid synostosis on opposite sides.

### Acquired Craniosynostosis

One female infant in our series had acquired craniosynostosis with congenital hydrocephalus who had undergone ventriculo-peritoneal shunting and developed fusion of multiple cranial sutures. This is a known, but uncommon, sequela of shunting that has been previously described.$^{12}$ Our patient was treated with extensive calvarial remodeling with multiple bone flaps at 23 months. She did well after surgery, but died 6 months later of unrelated sepsis secondary to necrotizing enterocolitis and bowel perforation.

### Outcome

Outcome after completion of planned surgery, whether one or two operations, was analyzed according to the classification system listed in Table 1 (Fig 4). Sagittal synostosis patients were not included in this analysis, because they will be analyzed separately and reported elsewhere. All patients followed for a minimum of 6 months after completion of planned surgery were included in this analysis. The actual numbers are shown in Table 6, which includes 115 of 143 possible patients. To allow statistical analysis we assigned a numerical value to each class, as follows: Class 1, 0 points; class 2, 0.5 point; class 3, 1 point; class 4, 2 points; class 5, 3 points; class 6, 4 points; and class 7, 4 points.

Although admittedly arbitrary, we attempted to assign numbers proportionate to the amount by which a result varied from class 1. Classes 6 and 7 have the same numerical value because they were

### TABLE 4. Complications, Planned and Unplanned Reoperations, and Deaths Among 250 Craniosynostosis Patients, by Sutural Involvement

<table>
<thead>
<tr>
<th>Sutural Involvement</th>
<th>Complications (%)</th>
<th>Patients Undergoing Planned Reoperation (%)</th>
<th>Patients Requiring Unplanned Reoperation (%)</th>
<th>Deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sagittal</td>
<td>+4 (3.7)</td>
<td>0</td>
<td>2 (1.9)</td>
<td>0</td>
</tr>
<tr>
<td>Unilateral Coronal</td>
<td></td>
<td>0</td>
<td>$\geq 2 (7.1)$</td>
<td>0</td>
</tr>
<tr>
<td>Bilateral Coronal</td>
<td>+5 (23.8)</td>
<td>5 (23.8)</td>
<td>+4 (19.0)</td>
<td>0</td>
</tr>
<tr>
<td>Unilateral Lambdoid</td>
<td>+2 (6.7)</td>
<td>0</td>
<td>$\geq 2 (6.7)$</td>
<td>0</td>
</tr>
<tr>
<td>Bilateral Lambdoid</td>
<td></td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Metopic</td>
<td></td>
<td>0</td>
<td>1 (5)</td>
<td>0</td>
</tr>
<tr>
<td>Multiple suture</td>
<td>+4 (13.3)</td>
<td>13 (43.3)</td>
<td>+5 (16.7)</td>
<td>0</td>
</tr>
<tr>
<td>Kleeblattschädel</td>
<td>+2 (33.3)</td>
<td>5 (83.3)</td>
<td>+2 (33.3)</td>
<td>2 (33.3)</td>
</tr>
<tr>
<td>Acquired</td>
<td></td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>17 (6.8)</td>
<td>23 (9.2)</td>
<td>18 (7.2)</td>
<td>2 (0.8)</td>
</tr>
</tbody>
</table>

* $P$ values compared to zero, for complications and for unplanned reoperations, based on binomial ($n \leq 20$) or Poisson ($n > 20$) distributions.

$+$ $P < .001$.

$\ddagger$ $P < .05$.

$\S$ $P < .01$.

### TABLE 5. Distribution of Involved Sutures for 30 Patients With Multiple Suture Synostosis

<table>
<thead>
<tr>
<th>Sutural Involvement</th>
<th>No. % of Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unilateral coronal, unilateral lambdoid</td>
<td>10 33.3</td>
</tr>
<tr>
<td>Metopic, sagittal</td>
<td>5 16.7</td>
</tr>
<tr>
<td>Sagittal, unilateral coronal</td>
<td>2 6.7</td>
</tr>
<tr>
<td>Sagittal, bilateral coronal</td>
<td>2 6.7</td>
</tr>
<tr>
<td>Sagittal, unilateral lambdoid</td>
<td>2 6.7</td>
</tr>
<tr>
<td>Unilateral coronal, bilateral lambdoid</td>
<td>2 6.7</td>
</tr>
<tr>
<td>Metopic, unilateral coronal</td>
<td>1 3.3</td>
</tr>
<tr>
<td>Metopic, unilateral lambdoid</td>
<td>1 3.3</td>
</tr>
<tr>
<td>Metopic, sagittal, bilateral coronal</td>
<td>1 3.3</td>
</tr>
<tr>
<td>Sagittal, unilateral coronal, bilateral lambdoid</td>
<td>1 3.3</td>
</tr>
<tr>
<td>Sagittal, bilateral lambdoid</td>
<td>1 3.3</td>
</tr>
<tr>
<td>Bilateral coronal, unilateral lambdoid</td>
<td>1 3.3</td>
</tr>
<tr>
<td>Bilateral coronal, bilateral lambdoid</td>
<td>1 3.3</td>
</tr>
</tbody>
</table>
believed to represent the same outcome, differing only as to whether or not the family elected to have further surgery. The mean and SE of the outcome scores are listed in Table 7. The best outcome (lowest score) was found in the metopic synostosis group (Fig 5). Comparing the seven groups by the Tukey test, looking for pair differences, significantly worse outcomes were found for Kleeblattschädel, bilateral coronal, and multiple suture synostosis patients compared with metopic synostosis patients. No

Fig 4. A and B, 3-month-old female infant with left unilateral coronal synostosis. C and D, The same patient at 16 months, after bilateral frontal-orbital remodeling at 5 months. She had three palpable stainless steel wires, above the hairline, which have not needed to be removed. Therefore, this was a class 2 result.

Fig 5. A, 14-month-old male child with untreated metopic synostosis. B, The same patient on the operating table, before correction, at 17 months. C, At the completion of surgery. D and E, At 2 years, 10 months.
TABLE 6. Surgical Result After Reconstruction for Craniosynostosis in Patients Followed for a Minimum of 6 Months, Analyzed by Sutural Involvement

<table>
<thead>
<tr>
<th>Class</th>
<th>Unilateral Coronal</th>
<th>Bilateral Coronal</th>
<th>Unilateral Lambdoid</th>
<th>Bilateral Lambdoid</th>
<th>Metopic</th>
<th>Multiple Suture</th>
<th>Kleeblattschädel</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8 (30.8)</td>
<td>6 (31.6)</td>
<td>7 (36.8)</td>
<td>3 (60)</td>
<td>8 (50)</td>
<td>2 (7.7)</td>
<td>1 (25)</td>
</tr>
<tr>
<td>2</td>
<td>3 (11.5)</td>
<td>1 (5.3)</td>
<td>3 (15.8)</td>
<td>0</td>
<td>6 (37.5)</td>
<td>7 (26.9)</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>2 (7.7)</td>
<td>3 (15.8)</td>
<td>6 (31.6)</td>
<td>1 (20)</td>
<td>1 (6.2)</td>
<td>5 (19.2)</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>1 (3.8)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1 (6.2)</td>
<td>2 (7.7)</td>
<td>0</td>
</tr>
<tr>
<td>5</td>
<td>11 (42.3)</td>
<td>5 (26.3)</td>
<td>1 (5.3)</td>
<td>1 (20)</td>
<td>0</td>
<td>6 (23.1)</td>
<td>0</td>
</tr>
<tr>
<td>6</td>
<td>1 (3.8)</td>
<td>4 (21.1)</td>
<td>2 (10.5)</td>
<td>0</td>
<td>0</td>
<td>3 (11.5)</td>
<td>2 (50)</td>
</tr>
<tr>
<td>7</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1 (3.8)</td>
<td>1 (25)</td>
</tr>
</tbody>
</table>

* Percentage of each group shown in parentheses; see Table 1 for definitions of classes 1 to 7.

TABLE 7. Numerical Outcome Scores for Patients After Surgical Treatment of Craniosynostosis, Ranked by Sutural Involvement, Minimum Follow-up 6 Months

<table>
<thead>
<tr>
<th>Sutural Involvement</th>
<th>No.</th>
<th>Mean</th>
<th>SE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Metopic</td>
<td>15</td>
<td>0.38</td>
<td>0.13</td>
</tr>
<tr>
<td>Bilateral lambdoid</td>
<td>5</td>
<td>0.80</td>
<td>0.58</td>
</tr>
<tr>
<td>Unilateral lambdoid</td>
<td>19</td>
<td>0.97</td>
<td>0.30</td>
</tr>
<tr>
<td>Unilateral coronal</td>
<td>26</td>
<td>1.63</td>
<td>0.28</td>
</tr>
<tr>
<td>Multiple suture</td>
<td>26</td>
<td>*1.79</td>
<td>0.28</td>
</tr>
<tr>
<td>Bilateral coronal</td>
<td>19</td>
<td>*1.82</td>
<td>0.38</td>
</tr>
<tr>
<td>Kleeblattschädel</td>
<td>4</td>
<td>*3.00</td>
<td>1.00</td>
</tr>
</tbody>
</table>

* Significant pair difference, compared to metopic synostosis patients, by Tukey test.

DISCUSSION

We found a striking male preponderance among the sagittal synostosis patients; 62 males and 25 females. That finding is consistent with the hypothesis of Graham et al.,13 that fetal head constraint may contribute to sagittal synostosis. The argument is that the increased occurrence of sagittal synostosis in males is related to larger fetal head size during the third trimester of pregnancy, resulting in a higher degree of physical constraint of the head in the maternal pelvis. Overall, our surgical results with sagittal synostosis were excellent, with only four complications (3.7%) and only two unplanned reoperations (1.9%). The median age at surgery for this group (a little more than 3 months) was lower than for all other groups except the Kleeblattschädel patients.

The unilateral coronal synostosis patients had no perioperative complications, but a high proportion, 46%, had a compromised aesthetic result because of residual postoperative asymmetry. In only one patient was the asymmetry severe enough for reoperation to be recommended. However, that is a very subjective decision. In this series, the median age at first operation for unilateral coronal synostosis was almost 7 months. We are presently operating on these patients at a younger age (4 to 6 months) hoping to achieve better symmetry because the bone is more pliable and the deformity is less severe at a younger age.

The bilateral coronal synostosis group is striking for the preponderance of females (76.2%) and the frequency of named syndromes (57.1%), the highest of any group in this study. Median age at first operation was 4.5 months. Five of 21 patients underwent planned second operation, and 4 of 21 required an unplanned reoperation ($P < .001$). There was a high complication rate, 23.8% ($P < .001$), and all five complications occurred in syndromic patients. Outcome was significantly worse than for our best group, the metopic synostosis patients. Bilateral coronal synostosis patients are a challenging group, at least partly due to the frequent presence of named syndromes with other associated problems and abnormalities in these patients. Many, such as the Aport and Crouzon patients, also have severe mid-face deformities, making it difficult to achieve or even judge the proper amount of frontal and superior orbital advancement, whether done as the initial operation or as a combined monobloc advancement.14 There is still not a consensus as to the optimal timing and approach for these patients.8,15–19

The unilateral and bilateral lambdoid synostosis patients did well overall, although the unilateral lambdoid synostosis group did have a 6.7% complication rate, and two patients required unplanned reoperation. As with the unilateral coronal patients, the major problem encountered with unilateral lambdoid synostosis has been asymmetry. In fact, one of our patients had such severe secondary forehead asymmetry that she eventually underwent frontal-orbital remodeling after two occipital procedures.

Although unilateral lambdoid and bilateral lambdoid synostosis patients comprised 12.0% and 2.8% of the patients in this series, respectively, that has changed as of 1992. We are now operating on a far smaller number of these patients. At present, when we have an opportunity to see patients before 1 year, we treat them with a molding headband20 (Dynamic Orthotic Cranioplasty, Southwest Orthotic-Prosthetic Laboratory, Phoenix, AZ). Initial results are very encouraging and have been reported separately.21 A recent report22 described a dramatic increase as of 1992 in the number of infants seen at a single center for plagiocephaly without synostosis. These infants characteristically present with unilateral occipital flattening that could be mistaken for unilateral lambdoid synostosis. We do not believe that any of our unilateral lambdoid synostosis patients actually had this entity, because radiographic and actual surgical findings were, in all our cases, consistent with true lambdoid synostosis. It has been hypothesized that plagiocephaly without synostosis has been seen with increased frequency as of 1992 because that is the year that the American Academy of Pediatrics...
formally launched a campaign to educate the public about the association of the prone sleeping position with sudden infant death syndrome. An infant positioned supine for sleep, it is suggested, may favor one side and thus expose that side to gentle but constant pressure of enough magnitude to affect head shape. Because that series ended in 1992, it seems unlikely that such factors played a role in many, if any, of these cases.

Our best surgical results were obtained in the metopic synostosis patients (Table 7). We had no complications, performed no planned reoperations, and only one patient (5%) required unplanned reoperation in the metopic synostosis group. A recent study, with 1-year follow-up CT measurements in 10 metopic synostosis patients, demonstrated improved but persistent anterior orbital hypotelorism. Although our assessment was that these patients had excellent outcome overall, a number of these patients do have residual mild hypotelorism even after surgical correction.

The multiple suture synostosis patients were a surprisingly large and heterogeneous group. Four of the 30 patients had identified syndromes. There were high rates of complications (13.3%), planned two-stage correction (43.3%), as well as unplanned reoperations (16.7%). It is difficult to generalize about such a diverse group, except to emphasize the need to individualize the treatment plan based on the involved sutures and the resulting deformity. If there is significant lambdoid and/or coronal involvement, posterior release (craniectomy or cranioplasty) at a young age, perhaps 1 to 3 months, can relieve intracranial hypertension and allow waiting until 4 to 6 months for the definitive frontal-orbital reconstruction. By 4 to 6 months, we find the bone easier to work with to accomplish and stabilize frontal-orbital advancement and remodeling. The high incidence of multiple suture synostosis in our series, 12%, is striking, particularly because bilateral coronal, bilateral lambdoid, and Kleeblattschädel patients are grouped separately and make up an additional 13.6% in our series. Little has been written specifically about multiple suture synostosis. Hoffman and Reddy reported the experience from the Hospital for Sick Children in Toronto, Canada, but what they described was a somewhat different entity. They found 11 patients (1.7%) in a retrospective review of 665 craniosynostosis patients over a 58-year period. However, their reports emphasized delayed and progressive multiple suture synostosis, their patients were older at presentation (24 to 117 months), most of their cases were holocranial, and four of their cases followed previous surgery and might better be considered acquired craniosynostosis. Interestingly, Shillito and Matson, in their 1968 work based on 619 surgical patients in a 40-year period at Children’s Hospital Medical Center in Boston, reported 76 of the 619 patients (14.6%) as having multiple suture synostosis. Of those, 10 had involvement of two unpaired sutures, 36 had involvement of three sutures, and 30 had involvement of four or more sutures. Because that last group probably included patients that we have separately listed as Kleeblattschädel deformity, it is striking how close their incidence, 14.6%, is to the combined incidence of our multiple suture patients and Kleeblattschädel patients, 14.4%. Also striking is that 38% of their multiple suture patients were operated on more than once, in comparison with 43.3% of our multiple suture patients who underwent planned reoperation and 16.7% who required unplanned reoperation. Our rates of reoperation for the six Kleeblattschädel patients were even higher.

The Kleeblattschädel deformity is characterized by a trilocular or cloverleaf cranial configuration, associated facial malformations, hydrocephalus, and (in some cases) micromelia and skeletal anomalies. This has been a particularly difficult problem in our experience, as well as that of others. Despite planning a two stage approach in five of our six patients, two of those patients still required additional unplanned operations. The only two deaths in our series were those two patients.

Our one patient with acquired craniosynostosis developed the problem after ventriculo-peritoneal shunting for hydrocephalus. The presumed mechanism is rapid decompression of the cranial vault, after shunting, resulting in overlapping of the bones across what had been widely spaced sutures. If brain growth does not soon spread the bones again, the sutures can become synostosed.

We found a striking association of mortality and morbidity with the presence of a named syndrome. There are two possible explanations, both of which may play a role. First, 16 of the 23 syndrome patients had bilateral coronal synostosis, multiple suture synostosis, or Kleeblattschädel deformity, three groups that are particularly challenging surgically and would have required longer, more extensive, and more complicated surgical approaches. Second, many of the associated findings in the syndromic patients, such as a retracted midface with airway narrowing, would put those patients at increased risk for complications.

In conclusion, we have reviewed a 6-year experience with surgical management of craniosynostosis at a single center. This is the largest such series to be analyzed and reported in almost 30 years. This report provides outcome data for craniosynostosis right up to the beginning of the recent controversies regarding the possible association of supine sleep positioning and plagiocephaly without synostosis, an entity that should be managed without surgery. Our results demonstrate that true craniosynostosis can be successfully corrected, using modern surgical techniques, with relatively low morbidity (6.8%) and mortality (0.8%). Many unanswered questions remain, but we hope that this report of a large, recently treated series of craniosynostosis patients will contribute to our evolving understanding and treatment of these problems.

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Surgical Treatment of Craniosynostosis: Outcome Analysis of 250 Consecutive Patients
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