Cranioplasty For Craniosynostosis In The Negev: Our Experience And Surgical Concepts And Early Post Operative Results

E Silberstein, O Perez, T Silberstein, L Rosenberg, A Cohen

Citation

Abstract
Introduction: Craniosynostosis is characterized by early fusion of cranial sutures resulting in a variety of structural phenotypes and neurological sequelae due to alteration in cranial volume and restriction of brain growth. Tessier introduced the modern concept of cranioplasty that included wide subperiosteal exposure of cranial vault and orbits complete separation and reposition of large parts of the skull including the orbits. The goal of this study is to describe our experience with surgical correction of craniosynostosis with its operative and early post-operative course and complications.

Materials and methods: This is a retrospective chart review of the first 62 consecutive children that underwent surgical cranioplasty for craniosynostosis at Soroka university medical center between Jan 1991 and Dec 2005.

Results: Our surgical technique for each type of craniosynostosis is described as well as intra-operative and post-operative early complications. 57 out of 62 patients had isolated craniosynostosis. There was no gender difference with regard to presence of synostosis or suture involvement. We had one case of intra-operative death. Six cases of significant complications including one CSF leak, one infection that required drainage, one case of transient leg ischemia and two cases of hematoma, 10 cases of minor complications and 46 cases with no complications. All patients required blood transfusion.

Conclusion: Cranioplasty for craniosynostosis is a safe procedure but it should be practiced in dedicated centers with advanced intra and post-operative monitoring and intensive care facilities with personnel experienced in these type of procedures.

INTRODUCTION
Craniosynostosis is characterized by early fusion of cranial sutures resulting in a variety of structural phenotypes and neurological sequelae due to alteration in cranial volume and restriction of brain growth. Virchow suggested in 1851 that premature fusion along the suture lines results decreased growth of the skull in the perpendicular axis and a compensatory growth along a plane parallel to the fused suture [1]. This theory was dominant for over 100 years. More recent studies led to better understanding the influence of cranial base pathology on the development of craniosynostosis [2-4] as well as its molecular and genetic mechanism involving mutations in the receptor for fibroblast growth factor (FGFR) [5]. It was Tessier [6] who introduced the modern concept of cranioplasty that included wide subperiosteal exposure of cranial vault and orbits complete separation and reposition of large parts of the skull including the orbits. He found that the orbits can be moved and repositioned in any direction without impairment of vision or ocular movement. The progress in intra-operative monitoring and pediatric anesthesia enables surgeons today to implement these anatomical and surgical principles. However, these procedures did not become less risky and easier and proper surgical and anesthetically setting with caution and adherence to non traumatic tissue handling principles are prerequisites for safety and success.

The goal of this study is to describe our experience with surgical correction of craniosynostosis with its operative and early post-operative course and complications.

MATERIALS AND METHODS
This is a retrospective chart review of the first 62 consecutive children that underwent surgical cranioplasty for craniosynostosis at Soroka university medical center between Jan 1991 and Dec 2005. Demographical, anatomical, surgical and post operative data were collected from medical records operative and anesthesia reports. Statistical analysis was performed using non-parametric
Kruskal–Wallis Test for quantitative or ordinal parameters. To test the difference of qualitative variables between groups we used Chi-square or Fisher exact test when appropriate.

**SURGICAL TECHNIQUE**

All procedures were executed by a combined team of plastic surgeons and neurosurgeons. All were done under general anesthesia in a supine position. All were approached thru a bi-coronal incision followed by wide exposure of skull vault in two different layers: sub-galeal sharp dissection followed by raising two large, as long as possible anteriorly based periosteal flaps (fig 1). The entire surgical process was done under copious saline drip in order to avoid desiccation of the tissue’s surface. One reason for raising these flaps was to enable closure of the distinct layers in two different plains so that suture lines are not over opposing. The other reason to raise these flaps was that they may be used to fill soft tissue defects, cover the bone graft’s areas or even to repair dural tear. Osteotomies were planed according to the type and severity of skull deformation. In cases of brachycephaly we performed fronto-orbital advancement and frontal bones remodeling (fig 2). In cases of plagiocephaly fronto-orbital advancement or unilateral temporal advancement with frontal bone remodeling was done. In cases of trigonocephaly bi-temporal advancement and frontal remodeling was the procedure of choice. In cases of scaphocephaly strip craniotomies and vault remodeling was performed (fig 3). Bone grafts were secured in most cases using Vicryl™ 3/0 threads and stainless steel wires until 2002 and wires and titanium clamp fixation system Craniofix® later on. In few cases absorbable or titanium mini plates were used. All patients were transferred for initial recovery to pediatric intensive care unit (PICU) followed by few days of hospitalization in the pediatric surgery department.

**RESULTS**

As mentioned before, these are the first 62 children operated for craniosynostosis during the study period. Gender distribution was statistically equal, 32 males patients and 30 females. 57 (92%) cases where non symptomatic with 5 (8%) children having craniofacial syndromes (4 Crouzon syndrome and 1 Apert syndrome). There where 17 (27%) cases of family history of craniosynostosis. There was no correlation between ethnicity and family history of isolated craniosynostosis or syndromic craniosynostosis. The frequency of sutures involvement was as follow: coronal suture was involved in 27 cases (44%), of those 19 (31%) were unilateral and eight (13%) bilateral. Sagittal suture was involved in 20 cases (32%). Metopic suture was involved in ten (16%) and lambdoid suture in five (8%) cases only. There was no difference between genders regarding the suture type involvement (Fig 4). Time of diagnosis: 25 (38.5%) were diagnosed at birth, 57 (90%) were diagnosed before the age of one. Two children were diagnosed at 12 months, one child at 18 months and two at 24 months. Mean and standard deviation of age at diagnosis was 5.2 ± 3.4 months. Time of surgery: 38 children (61%) were operated before the age of 9 months. 44 (70%) before 12 months. 17 children (30%) were operated at the age of 12 months or later. Patients that had positive family history were operated at significantly younger age then patients without family history 7.2 ± 2.6 months vs. 11.7 ± 6.9 Months respectively (MW=177, p<0.05). We found positive correlation between the age of diagnosis and age of surgery with Spearman's correlation coefficient of 0.645 (p<0.05). The younger the patients age at surgery the shorter the duration of surgical procedure (r=0.235 , p<0.05). All patients required blood transfusion. The length of the procedure was directly correlated to the amount of blood administrated to the patient. In procedures taking up to 4.5 h (n=34) the amount of packed RBC transfused was 166 ± 99 ml whereas in cases longer than 4.5 h administration of 287 ± 232 ml. was needed (r=0.445, p<0.01). Immediate surgical complications are summarized in table 1. There was one unfortunate intra-operative death due to anesthesia accident (airway obstruction). Other significant complications include CSF leak, two cases of hematoma, one infection that required drainage and one lower extremity transient ischemia due to arterial line. Minor complications included: Minor infections and fever greater then 38.5°C for up to 48h. All patients were admitted to PICU after surgery. 58 patients (94%) stayed in the PICU for less then 24 h. Three patients were moved to pediatric department within 72h and one patient stayed in the PICU for more then 3 days.
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**Figure 1**
Table 1: Immediate post operative Complications

<table>
<thead>
<tr>
<th></th>
<th>No of cases</th>
<th>(%)</th>
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</thead>
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<tr>
<td>No complication</td>
<td>46</td>
<td>74</td>
</tr>
<tr>
<td>Significant complications</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Death</td>
<td>1</td>
<td>1.6</td>
</tr>
<tr>
<td>CSF leak</td>
<td>1</td>
<td>1.6</td>
</tr>
<tr>
<td>Wound Infection</td>
<td>1</td>
<td>1.6</td>
</tr>
<tr>
<td>Leg ischemia</td>
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<td>1.6</td>
</tr>
<tr>
<td>Hematoma</td>
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<td>3.2</td>
</tr>
<tr>
<td>Minor Complications</td>
<td>10</td>
<td>16</td>
</tr>
</tbody>
</table>

**Figure 2**
Fig 1: Anteriorly based perioseal flaps

Periosral flaps elevated at the beginning of procedure B. Periosteal flaps sutured back in place after bone fixation.
Figure 3
Fig 2: brachicephaly
A. Fronto-orbital bar on the back table. B. Bone grafts in place.

Figure 4
Fig 3: Scaphocephaly
A. Pre-operative picture. B. Intra-operative strip cranioplasty
Figure 5
Fig 4: Gender distribution versus suture type involvement

DISCUSSION
This paper summarizes our first experience over a period of 14 years. As such, there are obvious development and changes in management concepts and surgical techniques. We did not find any difference between genders with regarding to the prevalence of craniosynostosis in general nor with distribution of suture involvement. Most of our patients had isolated non symptomatic craniosynostosis, similar to some previous report [6] and differing from other centers [6, 7]. We found that in symptomatic cases or in cases with family history of craniosynostosis diagnosis was made earlier then in other cases, probably due to the family and physician awareness, leading to higher index of suspicion or due to the more severe condition and therefore more noticeable skull deformation. The younger the age at diagnosis the earlier the time of surgery. This may be explained by the assumption that more severe cases were diagnosed earlier, presenting also signs of elevated ICP as well as parents actively encouraging surgery. We had one case of mortality due to anesthetic complication at the beginning of our craniofacial service. This was a severe, 2.5 years old neglected case of Cruzon’s syndrome with parents continuously refusing surgery. This child has been diagnosed at birth and suffered among other, from increasing ICP and brain damage leading to his nearly blindness and severe and chronic upper airways obstruction and pulmonary infections. Thick mucus that filled the alveoli, obstructing the bronchial tree and the tube as well was the direct cause of death. In view of our safety results after the initial learning curve, we do think that cranioplasty for craniosynostosis is a safe procedure but it should be practiced in dedicated centers with advanced intra and post-operative monitoring and intensive care facilities with personnel experienced in these type of procedures. Further analysis of long term aesthetic and social outcome of these patients is needed.

References
Author Information

Eldad Silberstein
Center of R&D in plastic surgery, Soroka University Medical Center, Ben-Gurion University of the Negev

Odelia Perez
Center of R&D in plastic surgery, Soroka University Medical Center, Ben-Gurion University of the Negev

Tali Silberstein
Obstetric and Gynecology, Soroka University Medical Center, Ben-Gurion University of the Negev

Lior Rosenberg
Center of R&D in plastic surgery, Soroka University Medical Center, Ben-Gurion University of the Negev

Abraham Cohen
Neurosurgery department, Soroka University Medical Center, Ben-Gurion University of the Negev