

Craniofacial distraction for syndromic craniosynostoses: evidence of an operative learning curve

Distração craniofacial para tratamento das craniossinostoses: evidência de uma curva de aprendizagem operatória

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RESUMO

Introdução: Avaliar criteriosamente a utilização de distração osteogênica no tratamento das craniossinostoses sindrômicas, com base na experiência adquirida em um serviço terciário de cirurgia craniofacial, enfatizando os aspectos cirúrgicos e complicações. **Método:** Análise retrospectiva de 11 pacientes operados na mesma instituição. Para tanto, prontuários médicos, exames radiológicos e fichas operatórias foram utilizados para obtenção dos dados clínicos. **Resultados:** Onze pacientes foram submetidos a distração osteogênica craniofacial com distratores rígidos. Avanço frontofacial em monobloco foi realizado em 10 casos. Um paciente foi submetido a osteotomia do tipo LeFort III para avanço do terço médio da face. Todos os pacientes apresentavam exorbitismo e sinais de hipertensão intracraniana, e 8 (73%) casos possuíam algum grau de obstrução respiratória. A distração osteogênica craniofacial promoveu melhora clínica e radiológica em todos os pacientes. Fístula líquórica foi a complicação mais frequente (5 casos - 45%). Não houve mortalidade cirúrgica. **Conclusões:** A evolução das técnicas cirúrgicas possibilitou que a distração osteogênica com distrator rígido se tornasse uma importante ferramenta no tratamento das anomalias craniofaciais complexas presentes nas craniossinostoses sindrômicas, uma vez que seus resultados são muito satisfatórios.

Descritores: Craniossinostose/cirurgia. Anormalidades craniofaciais. Cranio/anormalidades.

ABSTRACT

Purpose: A critical evaluation of the use of distraction osteogenesis in the surgical treatment of syndromic craniosynostoses, based on the experience of a single institution, with an emphasis on surgical pitfalls and complications. **Methods:** Retrospective review of the clinical data of eleven patients operated on in the same institution. Medical charts, surgical notes and other relevant data were analyzed and the patients' symptoms and signs, diagnosis, surgical treatment details and complications were assembled. **Results:** All patients underwent craniofacial distraction with rigid distractors. Ten of them underwent monobloc frontofacial advancement. In one patient a LeFort III advancement was performed. All patients had exorbitism and signs of intracranial hypertension pre-operatively, whilst eight (73%) had upper airway obstruction. Craniofacial distraction surgery promoted an improvement in all patients. Cerebrospinal fluid leakage was the most frequent complication in this series, occurring in 5 cases (45%). **Conclusions:** The evolution of surgical techniques allowed distraction osteogenesis with rigid distractors to be an important tool for treating the craniofacial issues related to syndromic craniosynostoses.

Key words: Craniosynostosis/surgery. Craniofacial abnormalities. Skull/abnormalities.

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INTRODUCTION

The use of distraction osteogenesis in the treatment of craniofacial deformities started in 1992, by McCarthy¹. Since then, there have been outstanding advances in this field, both technological and surgical, leading to a more comprehensive management of complex defects and with good results overall.

The occurrence of craniosynostosis, defined as the premature closure of calvarial sutures, inevitably produces a restriction in the growth vectors of the skull and ultimately leads to typical deformities of facial and cranial bones^{2,3}. Syndromic craniosynostosis often have a retraction of both the frontal region, including skull base and the midface, and consequently show ophthalmological, intellectual and breathing disturbances⁴⁻¹⁰.

The frontofacial retraction that occurs in complex syndromic craniosynostoses is traditionally managed with advancements of both the upper (frontal) and midface (maxilla), in two steps^{3-5,9}. In the past, standard craniofacial advancements had some technical limitations and high morbidity and mortality³. Recently, the use of rigid devices for craniofacial distraction has become the gold standard for the management of these complex cases, producing a gradual expansion of the restrained structures (specially the frontal lobes), better compliance of soft tissues and progressive development of newly formed bone tissue⁵.

The aim of this paper is to present a single-center experience in managing syndromic craniosynostosis with distraction osteogenesis, with an emphasis on surgical techniques and pitfalls and perioperative complications.

METHODS

Patient population

Eleven patients were submitted to craniofacial distraction osteogenesis between 2003 and 2010, in the Hospital of Clinics, Ribeirão Preto School of Medicine, University of São Paulo (HCFMRP-USP) by the same multidisciplinary craniofacial team. They underwent a total of 14 surgical procedures regarding their craniofacial deformities. Their ultimate treatment was based on craniofacial osteogenesis using rigid devices for distraction. Medical charts, surgical notes and other relevant data were assessed and analyzed individually for each patient.

Monobloc frontofacial advancement was performed in ten patients as their definite treatment; a Le-Fort III osteotomy for midface advancement was done in one patient. Three patients needed other surgical procedures before craniofacial distraction: two patients underwent calvarial expansion due to intracranial hypertension and one patient needed a posterior fossa decompression due to symptomatic Chiari I malformation.

Nine internal distraction devices (KLS Martin®, Germany) and two external devices (Synthes®, West Chester, PA, USA) were used.

All patients underwent pre-operative Computerized Axial Tomography (CT) scans, both for planning the surgery and to look for intracranial associated pathologies.

Preoperative evaluation

With the purpose of analyzing the midface advancement an open Source radiological free software (Osiri® for Macintosh®, version 3.6, California, USA) was used. Using three

dimensional reconstruction, we were able to measure the distance between a standard fixed anatomical landmark (the most anterior point of the anterior margin of the foramen magnum - AFM), the glabella and the maxillary point (the posteriormost point in the concavity of the alveolar process of the maxilla bilaterally - MP), establishing the upper and midface advancement, respectively. Afterwards, the pre and post-operative images were fused and the advancement measurements were confirmed.

For evaluation of exorbitism, a Hertel exophthalmometer was used. Measurements were obtained from the lateral orbital rim to the corneal apex.

All patients underwent a preoperative polysomnography.

Statistical analysis

Statistical analysis was performed using SPSS for Macintosh, version 16.0 (SPSS, Inc.). The chi-square and Fisher exact tests were used to compare categorical data, and the nonparametric Mann-Whitney U test was used to compare independent groups. A probability value was deemed significant at less than 0.05.

RESULTS

Table 1 summarizes the demographic data for the 11 patients.

Of these patients, 7 (64%) had Crouzon Syndrome, 2 (18%) had Apert Syndrome and the last 2 (18%) Pfeifer type I Syndrome. Their mean age was 5.3 years, ranging from 4 months to 14 years. There were 7 (64.3%) boys and 4 (36.7%) girls.

All our patients presented with symptoms and signs of intracranial hypertension, including developmental delay. The mean pre-operative distance between the AFM and the glabella was 7.17 cm; post-operatively, it was 8.61 cm. Likewise, the mean distance between the AFM and the MP was 6.19 cm before surgery and 7.75 cm after. As a result, a mean advancement of 1.44 cm was obtained for the upper face and 1.56 cm for the midface (Figure 1).

All patients had an eye ball protrusion greater than 21 mm, 3 (27%) of them being greater than 30 mm (severe exorbitism). The mean pre-operative protrusion measurement was 27.4 mm. After the distraction process, all patients had a protrusion of 21 mm or less (mean 18.5 mm), resulting in a significant improvement in their proptosis ($p=0.0065$) (Figure 2).

Eight (73%) patients showed clinical features of upper airway obstruction, and three of them (27%) were even submitted to a tracheostomy before their arrival in our service (in one of them, we had to reopen the tracheostomy in the perioperative period). After surgery, all patients had a clinical improvement of their breathing pattern confirmed by polysomnography.

Complications were observed in 7/11 (63%) patients. Five (45%) cases presented a CSF leak; four of them required a post-operative lumbar drain, resolving within no longer than five days; the other one had to be reoperated for dural repair.

All cases required intra and post-operative blood transfusion. Two patients showed a perioperative hypovolemic shock that recovered with adequate blood transfusion.

Table 1 - Summary of the clinical characteristics of the children in the series.

Case	Age at surgery	Syndrome	Exorbi- tism (mm)	Airway Obstruc- tion	Management	Follow-up	Complications
1	7 years and 2 months	Pfeiffer	31	No	MFFA	5 years	CSF leak
2	4 years and 6 months	Crouzon	27	Severe	Midface advancement	6 years	CSF leak
3	4 months	Pfeiffer	26	Severe	MFFA	5 years and 6 months	CSF leak
4	8 years and 7 months	Crouzon	28	Severe	MFFA	1 year and 4 months	Orbital-zygomatic frac- ture CSF leak
5	3 years and 4 months	Crouzon	34	Severe	MFFA	1 year	Hypovolemic shock CSF leak Right orbital fracture
6	3 years and 8 months	Crouzon	29	Severe	MFFA	1 year and 6 months	Hypovolemic shock Orbital cellulitis
7	14 years and 3 months	Crouzon	25	Moderate	MFFA	1 year and 2 months	None
8	7 months	Apert	24	Moderate	MFFA	9 months	None
9	2 years and 3 months	Apert	23	No	MFFA	6 months	Displacement of distrac- tor
10	11 years and 8 months	Crouzon	31	Severe	MFFA	6 months	None
11	6 months	Crouzon	24	No	MFFA	5 months	None

MFFA = monobloc fronto-facial advancement; CSF = cerebrospinal fluid.

One patient exhibited an orbital cellulitis, probably because of the communication between the ethmoid sinuses and the orbital compartment during facial osteotomies, that recovered after a course of antibiotics. Two cases presented with a lower orbital rim fracture (both requiring surgical osteosynthesis) (Figure 3) and another case showed a displacement of the zygomatic part of the internal distractor device, which needed to be repositioned.

The follow-up ranged from 3 months to 7 years (mean follow-up 2.7 years).

DISCUSSION

In 1978, Ortiz-Monasterio developed a technique that enabled the advancement of both the upper and middle facial heights in a single procedure¹¹, thus sparing at least one surgery. Albeit a huge development, it also brought a high morbidity that outweighed its benefits³.

The correction of craniofacial deformities associated with syndromic craniosynostoses was initially based on a two-staged approach, whose first step was the fronto-orbital

advancement and the second one was the LeFort III osteotomy for the advancement of the midface^{3,12}. Indeed, although this technique provided some good results, it also carried out some substantial problems, specially regarding the need for more than one surgical procedure and the disadvantages of reoperations³. It must be remembered that these patients already had to undergo a number of surgeries for the other issues related to their syndrome.

Craniofacial advancements with distraction osteogenesis allow a progressive bone tissue formation between the separated bones without leaving a retrofrontal dead space^{5,6}. Therefore, good results can be achieved and the complications minimized. One might argue that the second operation required for the removal of the distractor is a drawback, but we agree with Arnaud et al.³ that, in this particular case, the benefits outweigh the disadvantages and that, in the future, the development of resorbable devices may solve this problem.

Many authors have published their experience with craniofacial distraction osteogenesis^{2,3,5,6,13-21}. It is now well established that gradual distraction reduces the risk of complications, preventing the occurrence of a retrofrontal dead space; also, it

Figure 1 - Monobloc frontofacial advancement in a 9-year-old patient with Crouzon's syndrome. **A** and **B**: preoperative photographs; **C** and **D**: postoperative photographs.

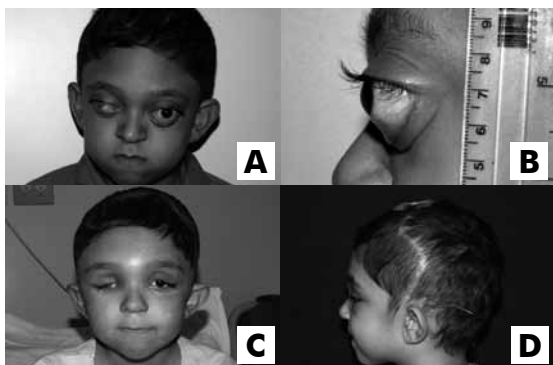
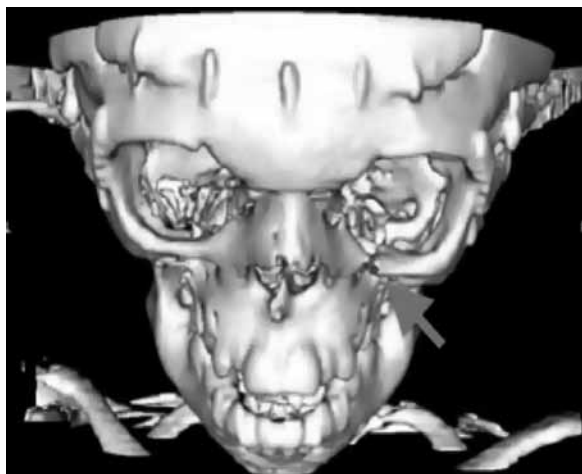


Figure 2 - CT scan (sagittal reconstruction) images of a 5-year-old patient with Crouzon's syndrome. All images were produced using a radiological software (OsiriX®). **A**: Measuring the distance between the AFM and the glabella. **B**: Measuring the distance between the AFM and the MP. **C**: Fusion of the pre (red shadow) and post-operative (gray) images, allowing a better visualization of the craniofacial advancement after distraction.



Figure 3 - 3D reconstructed CT scan showing a post-operative zygomatic-maxillary fracture (red arrow) due to distraction in a patient with Crouzon's syndrome.



promotes a slowly progressive bone formation, with a smaller number of bony defects^{3,5,6}. In addition, there appears to be a consensus that monobloc frontofacial advancement is an adequate method to improve the breathing disturbances and the exorbitism related to faciocraniosynostosis, as well as intracranial hypertension³.

Our experience has shown that the distraction interval between the separated bones provided by the distractors (15-30 mm) is enough to enlarge the intracranial, orbital and retropharyngeal and upper airway spaces, thus reducing intracranial pressure and improving the ocular proptosis and breathing obstruction that these patients present with. As described by other authors^{3,5,6,14,22}, in order to prevent complications related to residual hematomas, we also perform an immediate small advancement (usually 3-5 mm). This maneuver stabilizes to bony flap as well.

It should also be stressed out that, for the use of the distraction techniques, a multidisciplinary team is mandatory, as well as the awareness that it involves a learning curve²³, which is clearly demonstrated in our casuistry. In our first cases, we have had a relatively high rate of complications, but as we became more familiar with this surgical technique, we were able to lower our complications to a minimum (in the last five cases, four experienced no complications and the other case had a minor one). We have had no deaths.

Significant blood loss occurs in most of the patients operated on, since these surgeries involve large osteotomies and soft tissue dissection; additionally, children do not tolerate blood losses as well as adults^{5,6,22}. However, we believe that blood transfusion is safe and have not seen any major complications related to it.

CSF leaks are still a common complication, especially in patients that have already undergone previous craniofacial surgeries^{3,14,22}. Five of our patients (45.4%) needed additional procedures to correct CSF leaks; fortunately, all the leaks resolved and the distraction process did not need to be interrupted. The use of periosteal pedicled flaps to fill in the gaps in the anterior cranial fossa, ethmoid cells and orbital and nasal cavity may reduce the incidence of this complication^{3,14}.

Lastly, complications related to the distraction process itself may appear²². We have had two cases of periorbital (inferior rim) fractures, due to an uneven distraction. Close clinical and radiological follow-up is an important tool to prevent that, and confirmation that the distractors are well positioned and that the distraction vectors are synchronic is imperative.

CONCLUSIONS

Craniofacial distraction osteogenesis has become a safe method for the correction of deformities of the craniofacial skeleton in children with syndromic craniosynostoses. The use of these techniques encompasses a learning curve and a multidisciplinary team, and, with that in mind, the rate of complications can be minimized and the results turn out to be very satisfactory.

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