

Age and indications for osteotomy for frontofacial advancement in patients with syndromic craniosynostosis

Idade e indicações de osteotomias para avanço frontofacial em pacientes com craniossinostoses síndrômicas

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ABSTRACT

Background: Craniofacial surgery has overcome many challenges since its initiation into clinical practice. Several technical issues have been addressed and the basic infrastructure of the specialty has now been developed. At present, 25 years after the first publications on frontofacial advancement, questions still remain as to the appropriate age for surgery and the appropriate type of surgery that should be performed. The aim of this study was to evaluate patients surgically treated for syndromic craniosynostosis over the last 10 years at our institution. **Methods:** All syndromic patients who underwent monobloc frontofacial advancement or only isolated facial advancement from 2001 to 2011 were selected. Out of 70 patients in total, 56 underwent monobloc frontofacial advancement and 14 underwent facial advancement after fronto-orbital remodeling. All data concerning these patients were correlated with patient age and final result. Moreover, age at surgery, complications, and final results were correlated with the main preexisting problems. **Results:** Final results for syndromic patients varied, depending on the syndrome and the age at which the procedure was performed. Monobloc frontofacial advancements had a low index of immediate postoperative complications, but there was a clear need for further procedures at the time of final facial growth. The index of positive outcome was higher in patients who underwent surgery at an older age. **Conclusions:** In cases of severe craniosynostosis with functional problems, monobloc frontofacial advancement is still the best therapeutic option.

Keywords: Craniosynostoses/surgery. Craniofacial abnormalities. Outcome assessment.

RESUMO

Introdução: Desde o início da Cirurgia Craniofacial, muitos desafios foram ultrapassados. Problemas operatórios técnicos e de infraestrutura básica de atendimento especializado foram solucionados. Agora, 25 anos após as publicações iniciais dos avanços frontofaciais, há ainda algumas dúvidas quanto às indicações precisas da idade e do tipo de cirurgia a ser realizada. O objetivo deste estudo foi avaliar a evolução de pacientes submetidos a tratamento de craniossinostoses síndrômicas operados nos últimos 10 anos em nossa instituição. **Método:** Todos os pacientes síndrômicos submetidos a avanço frontofacial em monobloco ou somente facial isolado foram selecionados no período de 2001 a 2011. Foram selecionados 70 pacientes, 56 submetidos a avanço frontofacial em monobloco e 14, a avanço facial após remodelagem frontorbitária prévia. Todos os dados referentes a esses pacientes foram corre-

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lacionados, avaliando a idade e o resultado final. Os pacientes foram selecionados de acordo com idade à época da cirurgia, complicações existentes e resultados finais correlacionados com os principais problemas existentes previamente. **Resultados:** Os pacientes síndrômicos apresentaram graus variados de resultados finais, dependendo da síndrome e da idade de realização do procedimento. Os avanços frontofaciais em monobloco apresentaram baixo índice de complicações pós-operatórias imediatas, porém ficou demonstrada a necessidade de procedimentos futuros ao final do crescimento facial. Nos pacientes submetidos a cirurgias mais tardiamente, o índice de resultados positivos foi maior. **Conclusões:** Nos casos de craniossinostoses graves, com problemas funcionais, a indicação de avanço frontofacial em monobloco continua sendo a melhor opção terapêutica.

Descritores: Craniossinostoses/cirurgia. Anormalidades craniofaciais. Avaliação de resultados.

INTRODUCTION

Premature closing of the cranial sutures in children leads to profound craniofacial alterations, both aesthetic and functional. Complex craniosynostosis, involving the closure of several cranial and facial sutures, interferes with vital functions, and the craniofacial skeleton becomes compromised in 3 dimensions: cerebral expansion is restricted; protection of the ocular globe is deficient; and, most importantly, breathing is compromised.

The treatments that have been proposed to date aim at releasing the cranial sutures, with bone remodeling to allow for cerebral expansion; this is occasionally combined with orbital remodeling to protect the ocular globe. Additionally, in craniosynostosis with respiratory involvement, which is generally more serious, complex frontofacial advancement is prematurely advised. The greatest difficulty is in precisely identifying the perfect age to perform these surgical procedures. Although there are centers that perform these procedures in younger patients, others perform the procedures in older individuals because of the higher risk for complications associated with performing these operations on younger patients¹⁻⁴.

The aim of this study was to evaluate the clinical evolution of patients treated for syndromic craniosynostosis who underwent surgery at our institution over the past 10 years.

METHOD

Seventy patients with syndromic craniosynostosis who were operated on from 2001 to 2011 were analyzed in this study. The patients were operated on at different centers by the senior author (NA).

Among these 70 patients, 56 were subjected to monobloc frontofacial advancement and 14 to facial advancement with osteotomy type Le Fort III.

The patients were distributed into 3 groups according to age: younger than 5 years (G1), 5 to 12 years (G2), and older than 12 years (G3).

The correlations among the diagnosis, age, and complications associated with each type of procedure were examined.

Evaluation of results was conducted according to the position of the medial third of the face and orbits, as well as the presence of breathing problems after surgical correction. Patients operated upon in the last few years were not included in the final evaluation, which was limited to the 26 patients with a postoperative follow-up period of at least 5 years.

RESULTS

The distribution of patients according to diagnosis and procedure is shown in Table 1; patients suffering from Crouzon and Apert syndromes were predominant.

The groups of patients according to age are shown in Table 2, indicating that the majority of monobloc facial advancements were performed on patients between the ages of 5 and 12 years.

The average age of patients who underwent monobloc frontofacial advancement was 6.97 years and ranged from 9 months to 15 years. For frontal advancements, the mean age of patients was 17.21 years and ranged from 10 to 25 years.

Complications related to the surgery are illustrated in Table 3.

The results of the postoperative evaluation are summarized in Table 4 and showed that retrusion of the medial third of the face was found in 18 patients, all of whom were subjected to monobloc frontofacial advancements.

Table 1 – Case distribution according to diagnosis and procedure.

Diagnosis	Monobloc	Le Fort III	Total
Crouzon syndrome	32	8	40
Apert syndrome	18	6	24
Pfeiffer syndrome	5	–	5
Saethre-Chotzen syndrome	1	–	1

Eight patients required tracheostomy (G1 and G2); removal was possible in 6 of the cases.

Figures 1 to 3 illustrate some of the cases reported in this study.

DISCUSSION

Since its beginnings in the pioneering work of Paul Tessier, the field of craniofacial surgery has shown that there is hope for the treatment of severe facial deformities. In the past 15 years, with the development of frontofacial advancements using facial distraction, there has been a pronounced decrease in complications related to these procedures, which has encouraged many surgeons to use them. However, large osteotomies in immature skeletons

Table 2 – Case distribution according to age at the time of the procedure.

Age range	Monobloc	Le Fort III	Total
< 5 years (Group 1)	14	–	14
5-12 years (Group 2)	39	–	39
> 12 years (Group 3)	3	14	17
Total	56	14	70

Table 3 – Observed surgical complications.

Complications	n = 26
Cerebrospinal fluid fistula	3 (Group 2)
Infection	1 (Group 2)
Temporary vision problems	1 (Group 2)
Displaced bone fracture	3 (Group 2)
Breaking of distractor	3 (Group 2)

n = number of patients.

Table 4 – Late postoperative results.

Unsatisfactory results	n = 26	Syndrome
Ocular protrusion (Groups 1 and 2)	1	Apert syndrome
	1	Crouzon syndrome
Face retrusion (Groups 1 and 2)	4	Apert syndrome
	10	Crouzon syndrome
	3	Pfeiffer syndrome
	1	Saethre-Chatzen syndrome
Breathing problems (Group 2)	2	Crouzon syndrome

n = number of patients.



Figure 1 – Patient with Crouzon syndrome who underwent previous fronto-orbital advance. In A and B, lateral and front views, respectively, in pre-operative facial advancement Le Fort III type. In C and D, lateral and front views, respectively, in the postoperative period of 10 years of facial advancement Le Fort III type.



Figure 2 – Patient with Crouzon syndrome. In A and B, lateral and front views, respectively, preoperatively fontofacial monobloc advancement, demonstrating severe exophthalmos with retrusion of the midface. In C and D, lateral and front views, respectively, in the postoperative period of 7 years, demonstrating maintenance of the advancement of the orbital region.



Figure 3 – Patient with Apert syndrome. In **A** and **B**, lateral and front views, respectively, preoperatively frontofacial monobloc advancement, showing hypoplasia of the periorbital region and retrusion of the midface. In **C** and **D**, lateral and front views, respectively, in the postoperative period of 10 years, demonstrating maintenance of the advance obtained.

must start from the premise of evaluating the growth of the skeleton.

In the most dramatic clinical situations, with the presence of tracheostomy and pronounced ocular protrusion (generally in syndromic patients), it is our opinion that, to immediately improve the clinical situation, earlier and more aggressive facial advancements are indicated (monobloc frontofacial advancements), even in the absence of

adequate posterior growth of the medial third of the face. However, because of the inadequate facial growth in patients subjected to large advancements before the age of 5 years (G1), as well as in the 5- to 12-year-old group (G2), it is important to consider, whenever possible, postponing facial advancements until after the age of 12. Although the patients operated upon for facial advancement also had alterations in facial occlusion, none showed retrusion of the medial third when operated upon after the age of 12.

CONCLUSIONS

The low rate of perioperative and immediate postoperative complications in frontofacial osteotomies (monobloc frontofacial advancements) is encouraging, and results in the increased use of these procedures; however, the need for further operations after skeletal maturity strongly suggests that these procedures may eventually be performed in individuals at a older age, depending on clinical priorities.

Emphasis should primarily be given to functional problems but, even in less serious cases, patients with syndromic craniosynostosis undoubtedly have problems with the growth of the bones of the face.

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