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Review

Distraction osteogenesis in the surgical management of syndromic craniosynostosis: a comprehensive review of published papers

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Abstract

Our aim was to summarise current published evidence about the prognosis of various techniques of craniofacial distraction osteogenesis, particularly its indications, protocols, and complications. Published papers were acquired from online sources using the keywords "distraction osteogenesis", "Le Fort III", "monobloc", and "syndromic craniosynostosis" in combination with other keywords, such as "craniofacial deformity" and "midface". The search was confined to publications in English, and we followed the guidelines of the PRISMA statement. We found that deformity of the skull resulted mainly from Crouzon syndrome. Recently craniofacial distraction has been achieved by monobloc distraction osteogenesis using an external distraction device during childhood, while Le Fort III distraction osteogenesis was used in maturity. Craniofacial distraction was indicated primarily to correct increased intracranial pressure, exorbitism, and obstructive sleep apnoea in childhood, while midface hypoplasia was the main indication in maturity. Overall the most commonly reported complications were minor inflammatory reactions around the pins, and anticlockwise rotation when using external distraction systems. The mean amount of bony advancement was 12.3 mm for an external device, 18.6 mm for an internal device and 18.7 mm when both external and internal devices were used. Treatment by craniofacial distraction must be validated by long-term studies as there adequate data are lacking, particularly about structural relapse and the assessment of function.

Keywords: Syndromic craniosynostosis; distraction osteogenesis; midface; treatment strategies; distraction devices

Introduction

Craniosynostosis is premature fusion of the craniofacial sutures that causes disturbance of craniofacial growth. The function of cranial sutures is to allow deformation of the skull during passage through the birth canal and growth of the calvarium. Craniosynostosis can be an isolated event, which results in non-syndromic craniosynostosis, or it can happen in conjunction with other anomalies in distinct patterns that make up clinically-recognised syndromes. Despite there being a broad range, the most commonly identified syndromes of which craniosynostosis is a part include Crouzon, Apert, Pfeiffer, Muenke, and Saethre-Chotzen syndromes. The condition may develop when a gene mutates, or it may be genetically inherited.

Patients with syndromic craniosynostosis have impaired growth of the skull and facial bones that makes it hard to achieve lasting correction of the appearance and to maintain adequate space in the skull for the growing brain and other vital structures. Affected patients often have serious problems with breathing, hearing, speech, and mastication, and may also be born with anomalies of the limbs that

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give rise to additional functional and operative challenges. Functional assessment involves the evaluation of patients' problems that are associated with structural anomalies in syndromic craniosynostosis, and the main functional issues are caused by raised intracranial pressure as a result of restricted intracranial volume secondary to early fusion of the cranial sutures. Visual functions that can be affected include the optic nerve, which may be injured by the raised intracranial pressure; and inability to close the eyelids as a result of severe exophthalmos, which requires orbital protection. The patient may also have breathing difficulties secondary to the narrow nasopharyngeal airway caused by a severely hypoplastic midface. The most serious problem is the rise in intracranial pressure, which is detrimental to the brain. By using an epidural monitor in children with craniosynostosis, workers have shown that 47% of the children with "multiple-suture fusion" have increased intracranial pressure.[1] This renders those with syndromic craniosynostosis more difficult to care for, and requires a multidisciplinary team to address their needs effectively.

Those with additional midface retrusion, which is one problem of syndromic craniosynostosis, may present with raised intracranial pressure, exophthalmos, malocclusion, respiratory difficulty, and developmental delay.[2] Several techniques that have been developed (including surgical intervention in infancy, the advent of computed tomography, the introduction of rigid (and later resorbable) plating systems, and advances in distraction osteogenesis) now help to solve this problem. The Le Fort III midface advancement technique was introduced by Sir Harold Gillies in 1949 and later refined and popularised by Tessier.[3,4] More recently the development of the application of distraction osteogenesis to the craniofacial skeleton has raised the possibility of a new treatment (Le Fort III distraction osteogenesis), which does not involve bone grafting and has few complications. There are also lower rates of relapse and infection that had previously been caused by the massive frontonasal dead space behind the advancement segment.[5] Le Fort III has been replaced with monobloc or Le Fort III with facial bipartition, depending on the need for cranial expansion.[6]

Distraction osteogenesis was introduced into craniomaxillofacial surgery in 1992 when McCarthy et al successfully used it to lengthen a human mandible.[7] It involves an osteotomy followed by rigid fixation to lengthen the bone, muscles, and soft tissues, and it results in a serious degree of correction over time.[8] After a brief period of latency, the segments of bone are gradually detached at a specific rate and time. After the bone has regenerated, a period of consolidation allows for the mineralisation of the lengthened bone. This is a valuable advance in the treatment of children with mandibular deficiencies, and provides effective correction of micrognathia with associated expansion of soft tissue.[9] Technically, craniosynostosis can be treated more successfully in this way with different distraction devices.[10,11]

Early craniofacial correction (within the first years of life) depends on the neurosurgical and functional indications. During the first six years, reconstruction is focussed on cranial decompression and reshaping procedures associated with it. Reconstruction of craniofacial deformities is recommended after the age of 6 years, and will achieve stable adult dimensions in the cranio-orbitozygomatic regions. Finally, correction of occlusal relations can be achieved after skeletal maturity (Table 1).[12]

Table 1
Timing of distraction of osteogenesis.

Age	Distraction osteogenesis
Less than 6 months/6–12 months	Posterior distraction of the cranial vault (particularly in case of posterior stenosis of the skull)
4–8 or 9–12 years	Le Fort III advancements can combine with distraction osteogenesis when a lack of bone and soft tissue forces preclude a single stage
4–12 years	Monobloc frontofacial advancement
14–18 years	May be needed, may be not

The treatment of syndromic craniosynostosis is still evolving, and there is an increasing number of reports about distraction osteogenesis and the types of device being used.[12] Our aim was to compare the success of this treatment based on published reports (with particular emphasis on clinical indications, surgical techniques, advantages and disadvantages, types of distraction device, and complications). Such a review may alter the management of patients.

Methods

Objective

We have tried to synthesise evidence from current research to assess the effectiveness and prognosis of the different techniques of distraction osteogenesis that have been used in patients with syndromic craniosynostosis, with emphasis on their indications, protocols, and complications. Our question was: what was the effect of different distraction protocols and distraction devices in such patients who required craniofacial distraction, and what were their adverse effects, stability, and complications?

Selection criteria

We included studies of all patients of any age with syndromic craniosynostosis who had Le Fort III, monobloc or bipartition distraction and required one operation or more.

Exclusion criteria

We excluded patients who had been treated conventionally, those with non-syndromic craniosynostosis, and those who had been treated conservatively. Excluded studies were reviews, abstracts, debates between authors, summary

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