Crouzon Syndrome: A Case Series of Craniomaxillofacial Distraction Osteogenesis for Functional Rehabilitation

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Crouzon syndrome (CS) is the most common craniosynostosis syndrome and requires a comprehensive management strategy for the optimization of care and functional rehabilitation. This report presents a case series of 6 pediatric patients diagnosed with CS who were treated with distraction osteogenesis (DO) to treat serious functional issues involving severe orbital proptosis, an obstructed nasopharyngeal airway, and increased intracranial pressure (ICP). Three boy and 3 girls were 8 months to 6 years old at the time of the operation. The mean skeletal advancement was 16.1 mm (range, 10 to 27 mm) with a mean follow-up of 31.7 months (range, 13 to 48 months). Reasonable and successful outcomes were achieved in most patients as evidenced by adequate eye protection, absence of signs and symptoms of increased ICP, and tracheostomy tube decannulation except in 1 patient. Complications were difficult fixation of external stabilizing pins in the distraction device (n = 1) and related to surgery (n = 4). Although DO can be considered very technical and can have potentially serious complications, the technique produces favorable functional and clinical outcomes in treating severe CS.

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More than 100 craniosynostosis syndromes have been described, with an estimated birth prevalence of 1 in 2,000 to 2,500.[1] Syndromic craniosynostoses are estimated to constitute 15% of all craniosynostoses and more than 180 craniosynostosis syndromes have been identified to date, of which approximately 8% of cases are inherited or familial.[2] Crouzon syndrome (CS) is one of the most common syndromic craniosynostoses related to multiple fibroblast growth factor receptor 2 (*FGFR2*) mutations and was first reported by Louis Edouard Octave Crouzon in 1912 who described craniofacial dysostosis with the triad of calvarial deformities, facial anomalies, and exophthalmos in a woman and her son.[2,3]

Over the years, conventional craniofacial surgical techniques, such as strip craniectomy, fronto-orbital

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advancement, and Le Fort III procedures, have proved reliable to treat symptomatic syndromic craniosynostosis. However, in severe conditions, large segmental advancement requires the gap to be grafted, stabilized, and closed primarily because inadequate stability secondary to soft tissue restriction and unstable bone segment fixation can cause graft resorption, thus causing relapse and creating less than an ideal long-term outcome.

The introduction of distraction osteogenesis (DO) to craniofacial surgery has provided a reliable surgical alternative in achieving superior segmental advancement compared with conventional techniques in treating functional issues in syndromic craniosynostosis. Apart from obviating an additional bone grafting procedure, the natural process of bone regeneration through gradual traction simultaneously produces new histogenesis, which overcomes the soft tissue limitation.

In cases of severe CS, patients can present with major functional disturbances, namely increased intracranial pressure (ICP), severe exorbitism with the inability to achieve eyelid closure for orbital protection, and serious upper airway obstruction with progressive obstructive sleep apnea (OSA) secondary to a severely hypoplastic maxilla, which eventually might require a tracheostomy to bypass the obstructed airway. As such, the indication for each major surgery in pediatric patients with this condition should be agreed to by the craniofacial team members because the procedure carries substantial mortality and morbidity risks.[4]

This report presents a case series of pediatric patients with CS who underwent craniomaxillofacial DO to manage functional deficiencies, with the focus on surgical indications, choice of device, and the distraction protocol and its associated complications.

Report of Cases

This study was approved by the medical ethics committee of the Faculty of Dentistry of the University of Malaya (Kuala Lumpur, Malaysia; institutional review board reference number DF OS1516/0053[P]) and all participants signed an informed consent agreement.

This report describes 6 pediatric patients with CS (3 boys and 3 girls; age range, 8 months to 6 years). Five patients (patients 1 to 5) presented with increased ICP, severe exorbitism with an inability to achieve eyelid closure, and OSA secondary to a narrow nasopharyngeal space, and 1 patient (patient 6) presented with increased ICP only. All patients underwent a standard craniofacial protocol as routinely practiced in the authors' multidisciplinary craniofacial clinic, which involved computed tomographic (CT) analysis, fabrication of a 3-dimensional (3D) skull bio-model, and comprehensive assessment from neurosurgeons, maxillofacial surgeons, pediatric ophthalmologists, pediatric otolaryngologists, pediatric respiratory therapists, anesthetists, and clinical genetics. As part of the presurgical workup, the baseplates for the midface internal devices were pre-bent and fixed on a 3D skull bio-model for each patient to allow surgical simulation and vector determination and minimize operating time.

Based on specific functional indications, 5 patients underwent monobloc DO to achieve intracranial decompression, orbital protection, and nasopharyngeal airway relief and 1 patient underwent posterior cranial vault DO to address the isolated increase in ICP.

To treat the 3 functional issues optimally, 4 patients (patients 1 to 4) with severe structural deficiency received a combination of bilateral internal midface devices and a rigid external device (Synthes, Oberdorf, Switzerland) and 1 patient (patient 5) with moderate functional discrepancies received only bilateral internal midface distractors (Synthes). Because patient 6 presented only with signs of a potential progressive increase in ICP, posterior cranial vault expansion was indicated using internal distractors (Synthes).

All procedures were performed through the coronal approach. Before the osteotomy, the internal devices were placed to mark the planned placement site. In patients who underwent monobloc DO, the osteotomy was performed at the fronto-orbital region before completion of the Le Fort III osteotomy and completed through the maxillary tuberosity cut intraorally. Once the midface was separated, the internal devices were fixed and trial activation was performed to ensure correct vector trajectory. For patients receiving an external device, bilateral protective titanium temporal plates were fabricated and placed subperiosteally to prevent temporal bone perforation before the external frame was placed at the end of surgery. Patient 6, who underwent posterior cranial vault expansion, had a similar presurgical workup of the distractor application on his skull bio-model for surgical simulation and vector determination.

All patients were admitted to the pediatric intensive care unit for 3 to 5 days for close monitoring before being transferred to the pediatric ward. The mean latency period was 2.5 days (range, 1 to 3 days). The activation rate was 1 mm per day and the mean skeletal advancement was 16.1 mm (distraction range, 10 to 27 mm), with a mean consolidation period of 24 weeks (12 to 48 weeks). Mean follow-up was 31.7 months (range, 13 to 48 months). Reasonable and successful functional rehabilitation outcomes were documented in most patients as evidenced by the absence of signs and symptoms of increased ICP, ability for eyelid closure to achieve adequate eye protection, tracheostomy tube decannulation, resolved OSA, and an