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## Modified Lefort Distraction Osteogenesis for the Treatment of Nager Syndrome-Associated Midface Hypoplasia: Technique and Review

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**Abstract:** The surgical management of midface hypoplasia in the setting of Nager syndrome remains a significant challenge for craniofacial surgeons. This study describes a novel technique using distraction osteogenesis and modified osteotomies for the treatment of midface bony defects in an 11-year-old child with Nager syndrome.

Presurgical 3-dimensional planning was performed to design the osteotomies and placement of distractors. The surgical approach required upper buccal sulcus and extended transconjunctival incisions only. Osteotomies were performed from the pyriform aperture through the orbit to include the lateral orbital wall, with bilateral osteotomy of the zygomas through the anterior arch via the

transconjunctival incision. Distraction of the en bloc midface segment was successfully performed using external distractors. Bone grafting was not required. There were no complications.

External distraction was well tolerated and there were no intraoperative or postoperative complications. The distractors were removed uneventfully after consolidation. The midface was successfully advanced without the need for bone grafting or bicoronal incision. The occlusal plane was leveled and the aesthetic appearance of the child was improved.

Symmetrical midface hypoplasia in the context of Nager syndrome can be successfully corrected with en bloc distraction osteogenesis of the maxilla and bilateral zygomas through modified osteotomies that exclude the upper nasal pyramid. The approach is simplified and the need for bicoronal incision and bone grafting is mitigated in this technique, which the authors have named Lefort 2.5.

**Key Words:** Distraction osteogenesis, facial dysostoses, midface advancement, Nager syndrome

Nager syndrome, or acrofacial dysostosis, is a very rare craniofacial syndrome affecting the mandible, midface, and radial aspect of the hand.<sup>1</sup> Patients typically present with down-slanting palpebral fissures, micrognathia, cleft palate, and maxillary hypoplasia. Fewer than 100 patients with Nager syndrome have been reported in the literature.<sup>2</sup> In addition to hypoplasia, the maxilla is posteriorly malrotated and often associated with protrusion of the central upper midface. Classically, this deformity is treated with Lefort III or Lefort II osteotomies with zygomatic repositioning and bone grafting of the resultant defect.<sup>3</sup> There have been concerns regarding the long-term efficacy and graft resorption experienced with this technique.<sup>4</sup> Additionally, with the already protruding central upper midface, classically described Lefort II osteotomies including this segment may be unnecessary and may lead to an over-projected, beaked nasal appearance.

We describe the case of an 11-year-old boy with Nager syndrome, presenting with concerns regarding aesthetic appearance and occlusal plane abnormalities (Fig. 1). The patient underwent a novel technique for distraction osteogenesis-assisted advancement of the midface and zygomas that corrected the deformity adequately without over-rotating the central upper midface.

### OPERATIVE PROCEDURE

Physical examination of the patient demonstrated midface hypoplasia, with a prominent upper nasal pyramid and posterior open bite (Fig. 2A, C). The patient had previously undergone mandibular distraction. Preoperative computed tomography scans of the facial skeleton with three-dimensional reconstruction confirmed repositioning and hypoplasia of the inferior maxilla with concomitant protrusion of the central upper midface (Fig. 2A, C). Presurgical modeling was performed to design osteotomies to advance the lower midface and zygomas en bloc (Fig. 3). Preoperative modeling based on computed tomography scans was performed using the ProPlan CMF (Materialise, Glen Burnie, MD) platform. Based on the modeling and measured to correct the malocclusion, distraction was planned for 11 mm on the right and 7 mm on the left. The distraction distance was conservative as we prefer to perform a secondary distraction rather than over distract, which is more difficult to correct.

The patient and family provided verbal and written consent for the case details and photographs to be published in peer-reviewed literature.

The patient underwent preoperative assessment and was placed under general anesthesia with reinforced endotracheal tube insertion. Bilateral transconjunctival incisions with lateral canthotomy

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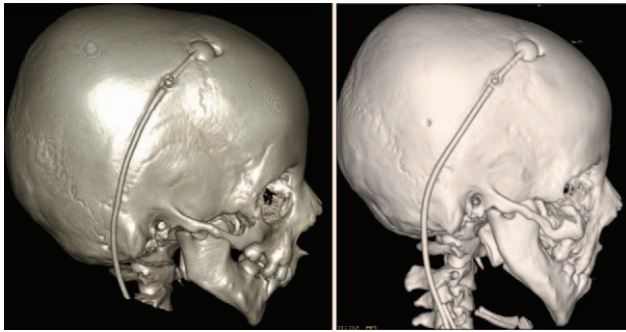
This case was presented at the 17th Biennial Congress of the International Society of Craniofacial Surgery in Cancun, Mexico on October 26, 2017.

The authors report no conflicts of interest.

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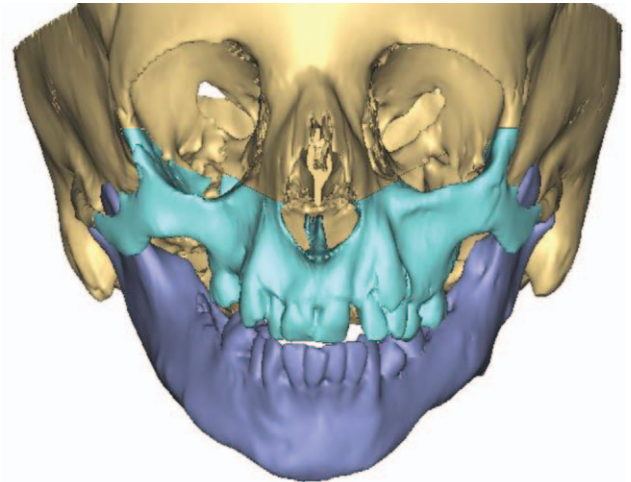


**FIGURE 1.** (A) Preoperative computed tomography three-dimensional reconstruction demonstrating inferior maxillary hypoplasia and retrusion. (B) Six months postoperative computed tomography three-dimensional reconstruction demonstrating improved maxillary position, consolidated bone across zygomatic arch, and improved bite position with no anterior or posterior open bite.

extensions were performed to expose the orbital floor, medial and lateral orbital walls, and zygomatic arch. Bilateral upper buccal sulcus incisions were performed to expose the pyriform triangle and the anterior maxillary walls. Bicoronal incision was performed, but was ultimately not required nor utilized for the procedure. Osteotomies (Fig. 3) were performed beginning at the mid-pyriform aperture bilaterally, extending through the lower medial orbital wall, the orbital floor, and the lateral orbital walls. The zygomas were osteotomized at the anterior arches just posterior to the body via the lateral canthotomy extension. The maxilla was completely mobilized.



**FIGURE 2.** (A) Preoperative sagittal clinical photograph demonstrating midface hypoplasia and prominent upper nasal pyramid. (B) Six months postoperative midface and mandibular distraction sagittal clinical photograph showing improved midface projection and upper nasal pyramid projection relative to lower nose. (C) Preoperative frontal clinical photograph demonstrating midface hypoplasia, mandibular retrusion, and downslanting palpebral fissures. (D) Six months postoperative midface and mandibular distraction frontal clinical photograph demonstrating improved projection of the midface and mandible.



**FIGURE 3.** Computed tomography three-dimensional frontal reconstruction of proposed osteotomies using the Proplan CMF (Materialise, Glen Burnie, MD) platform. The osteotomies extend from the mid-pyriform aperture medially through the orbital floor, lateral orbital rim and across the zygomatic arch, mobilizing the midface en bloc.

The distraction plates were placed on the zygomas according to the preoperative modeling design. Due to intraoperative problems with a previously placed ventriculo-peritoneal shunt, an external halo head-frame was applied by neurosurgery. The pin site of the halo interfered with the planned temporal distractor plate location and therefore external distraction was used as the fixed point.

The patient began distraction on postoperative day number 3, which is standard at our center for a patient this age. Distraction continued at 0.5 mm per day for a total advancement of 11 mm on the right and 7 mm on the left, as planned. The device was then removed after 40 total days in situ. There were no complications. Blood transfusion was not required and there was no pin-site infection. The patient tolerated both procedures and the distraction well. He underwent a second mandibular distraction subsequent to midface distraction to improve occlusion.

### DISCUSSION

Distraction osteogenesis is an increasingly accepted technique for treatment of midface abnormalities associated with craniofacial syndromes.<sup>5</sup> There has been much debate regarding the choice of osteotomies for facial dysostoses, with no clear superiority of a single technique. In the case of maxillary hypoplasia and retrusion, Lefort III, Lefort II with zygomatic repositioning or monobloc osteotomies are typically chosen depending on the presence of associated deformities and surgeon preference. Lefort II and III techniques are traditionally described with osteotomies through the upper nasal bones in the glabellar region, requiring either bicoronal or direct transglabellar incisions for access.

We have described a novel osteotomy technique, which we term Lefort 2.5, to advance the lower midface selectively. In Nager syndrome the maxilla is often hypoplastic and rotated posteriorly with respect to the upper third of the facial skeleton. By placing our central osteotomies through the lateral pyriform aperture rather than through the nasal root (Fig. 3), we prevent overcorrection of the upper nasal pyramid and maintain the ability to advance en bloc, leading to a well-corrected mid-face and improved nasal root position relative to the lower nose and midface that persisted 6 months postoperatively (Figs. 1B and 2B, D). Following the midface distraction, a secondary mandibular distraction was performed to further improve occlusion and projection. The patient achieved good occlusion anteriorly and posteriorly, no relapse was

experienced. Although this technique may increase anterior nostril show, this was mild and preferable to the overcorrection that would have resulted from distraction at the radix in standard Lefort II or III osteotomies. The parents were pleased with the aesthetic result in this case; however, this difference should be discussed preoperatively.

There is little information in the literature on midface management in Nager syndrome.<sup>6</sup> To our knowledge, this is the first reported patient to have undergone successful midface distraction in this small patient population. The described modified Lefort technique is well suited to advance the hypoplastic segment and correct the rotational deformity without overcorrecting the upper nasal pyramid. The technique also mitigates the need for bicoronal incision and bone grafting, both of which may increase operative morbidity and surgical time. Although ideal in the treatment of Nager-associated maxillary hypoplasia, Lefort 2.5 could also correct similar deformities in other facial dysostoses. Given the straightforward nature of the technique and potential broad applicability, we believe it is a useful adjunct for craniofacial surgeons.

### SUMMARY

Nager syndrome is a very rare craniofacial syndrome characterized by mandibular and midface hypoplasia. We describe a novel modified Lefort osteotomy technique that corrects maxillary hypoplasia and malrotation while sparing the upper nasal vault using external distraction osteogenesis. The procedure is straightforward, does not require bicoronal incision or bone grafting, and could be used in multiple examples of facial dysostoses.

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## Late Complication Associated With the Treatment of Orbital Floor Fracture With Titanium Mesh

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**Abstract:** Treatment of orbital fractures involves restoration of lost bone anatomy and orbital volume. Among the materials used for

this purpose, the titanium mesh is widely used due to its effectiveness and low complication rate related to distortion especially in non-traumatic events. This study shows an atypical and late complication involving the deformation of the titanium mesh used during reconstruction of the orbital floor in a patient with orbital zygomatic complex fracture 5 months after the procedure and without traumatic etiology.

**Key Words:** Fracture fixation, orbit/surgery, orbital fractures, surgical mesh

Orbitozygomatic complex (OZC) fractures account for approximately 13% of all craniofacial fractures<sup>1</sup> and may occur either in isolation or in combination with other facial bones. When the internal orbital walls are impaired, complications such as diplopia, ophthalmoplegia, enophthalmos, and ocular dystopia may be present due to extraocular muscle entrapment.

The treatment of OZC fractures is based on 3-dimensional recovery of the bone anatomy through the reconstruction of the facial and orbital volume.<sup>2</sup> Of the materials most commonly used to reconstruct the orbital walls, titanium mesh has proved to be effective and has gradually replaced autogenous bone grafts.<sup>3</sup> Its stiffness associated with some elasticity provides good support to the orbital content and implant fixation to the bone ridge, which prevents a secondary detachment.<sup>4</sup> However, its stiffness can be considered a disadvantage in recurrent facial traumas, in which the dissipated energy may cause implant distortion, thus threatening adjacent anatomic structures, especially the eye globe.<sup>5</sup> We report an unusual case of titanium mesh deformation 5 months after the treatment of OZC fracture and with no previous traumatic events.

A 26-year-old man who was a victim of car accident was diagnosed with OZC fracture (Fig. 1A). He underwent fracture reduction and fixation using one plate and four 5-mm monocortical screws from the 2.0 system (Bioplate, Los Angeles, CA) in the left zygomaticomaxillary buttress, and one semilunar plate and five 3-mm monocortical screws from the 1.5 system (Bioplate) in the left infraorbital ridge. Orbital floor reconstruction was then performed using one titanium mesh, which was fixed with two 1.5 system screws (Bioplate).

No complications occurred during the surgery, and the patient had no pain or ocular complaints in the immediate postoperative period. Ophthalmoplegia and diplopia were absent. Postoperative radiographs showed a good pattern of fracture reduction and good adaptation of the orbital floor mesh implant (Fig. 1B). Therefore, the patient was discharged and referred to the outpatient clinic for follow-up.

In the fifth month of postoperative control, the patient sought the emergency department complaining of intense pain in the left eye

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