Management of Exorbitism Using Midface Distraction Osteogenesis

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Abstract

Introduction Management of patients with syndromal craniosynostosis is probably one of the greatest challenges. They usually present with variable deformities yet share common characteristics. Patients with Crouzon and Aperts syndrome present with exorbitism and midface hypoplasia. Exorbitism is a protrusion of the eyeball due to a decrease in the capacity of the orbital container, and it may lead to drastic consequences if left untreated.

Material and methods Four cases with exorbitism and midface hypoplasia. Three presented with Aperts syndrome and one with Crouzon syndrome. They all underwent an intraoral modified Le Fort III osteotomy followed by midface distraction osteogenesis.

Conclusion Distraction osteogenesis of the midface is a safe and highly effective method in managing exorbitism associated with midface deficiency.

Keywords Distraction osteogenesis · Exorbitism · Le fort III · Crouzon · Aperts

Introduction

Exorbitism is a manifestation of an abnormal protrusion of the eye globe. The volume of the orbital contents is usually normal, but the marked globe protrusion in exorbitism is a result of a reduced bony orbital volume in other words “shallow orbit” [1]. This condition is seen in most facio-craniosynostosis syndromes including Aperts, Crouzon and Pfeiffer syndrome.

The degree of globe projection in relation to the orbital cavity is of great importance for both function and aesthetic reasons. When the eye globes are abnormally protruded, the consequences are countless. These complications include exposure keratitis and corneal ulcers due to the inability of the upper eyelid to properly protect and cover the globe.

In extreme situations, patients will present with lagophthalmos and globe subluxation, due to the contraction of the upper eyelid behind the globe [2]. Avoiding such drastic complications are usually the aim when dealing with such cases. Therefore, the type and timing of surgical intervention is of paramount importance.

The literature mentions many surgical options for correction of craniofacial exorbitism in syndromal faciocraniosynostosis. These surgical options include, fronto-orbital advancement followed by Le Fort III osteotomy advancement or distraction osteogenesis [3, 4]. The correction of exorbitism by monobloc fronto facial advancement with distraction osteogenesis was also mentioned in the literature [5].

However, there is only a handful of publications regarding the management of exorbitism and the effect of midface advancement on the orbital cavity.

We discuss the management of cases with syndromic exorbitism. They all underwent Le Fort III using distraction

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osteogenesis. We also discuss and demonstrate the effect of Le Fort III using distraction osteogenesis on exorbitism.

Patients and Methods

Four patients with exorbitism and midface hypoplasia as a result of syndromal craniofacial synostosis presented to the oral and maxillofacial surgery clinics at King Abdulaziz University. Three of them with Aperts syndrome and one with Crouzon syndrome. Ages ranged between 7 and 16 years. They were all medically free and gave a history of frontofacial advancement surgery except for one patient. They all underwent presurgical clinical and radiographic examinations including CT scans. They were managed with midface advancement using distraction osteogenesis. Three patients underwent Le Fort III advancement using distraction osteogenesis. A modified Le Fort III osteotomy excluding nasal complex (Wassmund III) was made via an intraoral incision and midface degloving. The osteotomy included the orbital floor and narrow portion of the zygomatic arch extending inferiorly to the pterygomaxillary junction. Rigid external distractors (RED) were used in three patients and internal distractors were used in one patient. After a 5 day latency period the distractors were activated at a rate of 1 mm/day. The length of distraction ranged between 20 and 36 mm. The midface and

exorbitism improvement was our guide. After a consolidation period of 3 months, all patients underwent CT examination before the removal of the distractor to confirm bony consolidation.

All four patients showed correction in the midface deficiency and exorbitism. They have all been under follow up for 3 years with no reported complications.

Case 1

A 7 year old boy with Crouzon syndrome presented with a midface hypoplasia and exorbitism which resulted in severe visual impairment. He had undergone a fronto-orbital advancement at the age of 3 years to correct the brachycephaly, which resulted in a cranial bone defect as demonstrated on the 3D CT image. The degree of exorbitism is evident on the axial CT image. He underwent 20 mm Le Fort III advancement using internal distractors, due to the cranial bone defects. A clear improvement in the midface deficiency and exorbitism was noted on the CT scan image taken 6 months later (Fig. 1).

Case 2

This 9 year old boy with Aperts syndrome, presented with brachycephaly, midface hypoplasia and exorbitism evident

Fig. 1 **a** *Left*: preoperative photograph showing the exophthalmos and the hypoplastic midface. *Center*: preoperative 3D CT scan. *Right*: preoperative axial CT scan. **b** *Left*: post operative photograph

showing an improvement in the exorbitism. *Center*: 3D CT scan obtained 6 months postoperatively. *Right*: axial CT scan 6 months postoperatively

Fig. 2 **a** *Left*: preoperative picture. *Center*: 3D CT scan showing the midface hypoplasia. *Right*: axial CT scan showing the degree of exorbitism. **b** *Left*: postoperative picture. *Center*: 3D CT obtained

6 months postoperatively. *Right*: axial CT scan obtained 6 months postoperatively showing the improvement in the exorbitism

in the CT scan images. He underwent a midface advancement using distraction osteogenesis. Le Fort III osteotomies were performed intraorally. An external rigid distractor was used and was active at a rate of 1 mm per day for 23 days, until there was a clinical improvement in the midface hypoplasia. Six months later the CT scan image demonstrated a successful midface distraction and correction of the degree of exorbitism (Fig. 2).

Case 3

A 16 year old male with Aperts syndrome had undergone craniotomy with fronto-orbital remodelling at age seven. He presented with midface hypoplasia, severe exorbitism, recurrent keratoconjunctivitis and lagophthalmos. The axial CT scan demonstrates the exorbitism. We performed Le Fort III intraoral osteotomies. An external rigid distraction device was fixed and activated after a latency period of 7 days at a rate of 1 mm per day for 26 days.

When a clinically evident improvement in the midface hypoplasia and exorbitism was noted the distraction was discontinued. In this case the midface hypoplasia was overcorrected with a slight correction in the degree of exorbitism. A CT scan taken 6 months later demonstrates osteogenesis of the midface with correction of the midface hypoplasia and exorbitism. When followed up 2 months

later, the patient was able to close his eyes slightly better than before distraction, but he still presented with lagophthalmos, which will require further lower eyelid surgery. This may include medial canthopexy or lower eyelid suspension using a graft (Fig. 3).

Discussion

The surgical correction of exorbitism in syndromal facio-craniosynostosis includes several options, but the traditional two-staged approach is still mainly used. Fronto-orbital advancement (FOA) is done first followed by Le Fort III advancement by using distraction osteogenesis [3, 4, 6] or osteotomy [7].

Children with craniosynostosis often have reduced orbital volumes due to the restricted ability to attain normal orbital growth. Fronto-orbital advancement helps restore normal growth of the orbital cavity and orbital volume. However, the timing of fronto-orbital advancement in craniosynostosis is still quite controversial. It is generally agreed that surgeons need to select the optimal time to operate in order to maximise the effects of normal orbital growth and therefore minimising the need for further surgical intervention. But it has been noted that the underlying mechanism that leads to restriction of orbital growth and

Fig. 3 **a** *Left*: preoperative picture showing the severe exorbitism and lagophthalmos. *Right*: preoperative axial CT scan. **b** *Left*: postoperative picture. *Center*: 3D CT scan obtained 6 months postoperatively

showing the midface distraction. *Right*: axial CT scan showing the improvement in the exophthalmos

volume seems to lose its effect during the end of the child's first year of life. Therefore, it is advised to delay the fronto-orbital advancement until the second half of the first year to maximise the effects of accelerated normal orbital growth and reducing relapse [8].

All previously mentioned patients gave a history of undergoing Fronto-orbital advancement, except for case 3. However, his exorbitism did not appear to be more severe in comparison to the other three cases.

Le Fort III midface advancement with or without distraction osteogenesis allows separation of the facial bones from the skull base, therefore the face can be moved independent of the cranial bone and mandible. An improved facial appearance can be achieved by balancing key projecting components of the midface, cranium, zygoma and mandible. A reduction in the amount of exorbitism is also achieved along with airway improvement and a possible correction of class III malocclusion. It has been noted that immediate advancement and gradual Le Fort III distraction osteogenesis to enlarge orbital cavities are the best approaches for the treatment of exorbitism in craniofacial synostosis [9].

However, distraction osteogenesis has the advantage of avoiding bone grafts and alleviates the restriction of soft

tissue to the bony advancement of the midface. The soft tissue envelope does not limit the amount of advancement when using distraction osteogenesis as in traditional Le Fort I, II, III osteotomy procedures. This is due to the formation of the functional matrix that compensates for as much of the growth deficit as possible. The resulting regenerate possesses the same morphology as the native hard and soft tissue and is considered to be more stable against relapse [8, 9].

In cases with severely hypoplastic facial bones, extensive advancement is usually the answer, but the amount of craniofacial advancement is usually limited by the surrounding soft tissue envelope [10]. It had been noted in the literature that the distance of craniofacial advancement is limited to approximately 10 mm. On the other hand, distraction osteogenesis allows an advancement of 20 mm or more by distraction of the soft tissue envelope [8]. The severity of the exorbitism and the midface hypoplasia in our cases required large advancements for correction. For this reason, our choice was gradual Le Fort III distraction osteogenesis. Most of our patients required an advancement of 20 mm or more. We performed a modified Le Fort III and used an external D.O in cases 2, 3 and 4. We used an internal D.O in case 1 due to the defects in cranial bone.

In craniofacial synostosis there are clear growth deficits, therefore it is likely that patients who undergo Le Fort osteotomies early in life, will require further surgical corrections for developing malocclusions at the end of adolescence [10]. Malocclusion after Le Fort III advancement can be attributed to mandibular growth. While the maxilla continues to show inferior growth, however shows minimal anteroposterior growth. In fact, it has been mentioned in the literature that maxillary growth after Le Fort III is virtually absent in syndromal faciocraniosynostosis [4]. This was observed in most of our patients. They all developed an anterior open bite and will require orthognathic surgery for correction.

Fronto-facial monobloc advancement using internal or external distraction devices has received a lot of interest. As noted with distraction osteogenesis we have the ability to produce large monobloc advancements thus minimising the risk of complications that could arise when performing advancements in the midface or frontal area. The effect of monobloc advancement on exorbitism correction has been reported in the literature with promising outcomes [5].

Fronto-facial advancement involves transaction of some or all of the orbital walls and moving them forward, therefore increasing the bony orbital volume; however, the literature has emphasised that in adults the improvement in globe protrusion was dependent on several factors. Luckily, these factors were under the surgeon's control. The first factor is the amount of fronto-facial advancement achieved. The second factor is the amount of orbital expansion resulting from fracturing the four orbital walls, especially the medial and inferior walls. The expansion on the anterior aspect of lateral orbital wall has minimum significance due to the presence of the temporalis muscle that acts as a barrier which permits orbital contents to prolapse into the temporal fossa [5]. The final factor to consider which is of less significance is the cicatricial contraction of the orbital fat. This happens if the orbital fat prolapses through the orbital wall when the periorbita is disrupted [11].

In children who undergo frontal or midface distraction, the final axial position of the eye globe is affected by factors that are beyond the surgeon's control. However, these factors play a significant role in the axial globe position. These include the growth of the eye ball and the change in axial globe position with age. Studies show that postnatal ocular growth from birth to 18 months is of great importance, as the axial length increases 3.8 mm. After that the rate of growth is much slower, so ocular growth from a surgical point of view would be of significance in cases where advancements are performed at early infancy [12, 13].

The difference in orbital morphology between Aperts and Crouzon syndrome is an important feature that must be

considered when correcting exorbitism in such cases. A recent study showed that in Aperts syndrome the posterior orbital wall is much more protruded than in Crouzon syndrome. On the other hand in Crouzon syndrome, the lateral and inferior orbital margins are much more retruded in relation to the cranial base than in Aperts syndrome. The most striking finding in Aperts syndrome is a marked protrusion of the lateral orbital wall in the sagittal plane (greater wing of the sphenoid). This was observed in sagittal CT records of our patients with Apert, especially case 3. This could mean that the globe would be pushed outwards due to the pressure exerted by the lateral wall even after midface distraction. Another point to consider is that midface distraction in patients with Aperts syndrome, rarely leads to normalization of the relation between the eye and the orbit. It also doesn't usually show normalization between the eye and the facial plane as often observed in Crouzon syndrome. In other words, Aperts syndrome characteristics most often remain, even after craniofacial surgery [14].

It is noted in the literature that changes in axial position of the globe that happen with age are of great importance. Using exophthalmometry the globe protrusion increases 2.57 mm from age 3 years to 10 years [15] and then continues to increase throughout puberty until adulthood and reaches values of 15–16 mm [16]. However, as the orbital volume also increases from birth until puberty [8], the change in globe protrusion is a reflection of the increased retrobulbar tissues and some amount of remodelling of the lateral orbital rim [16]. So the final position of the globe will not only depend on the amount of advancement, but other procedures may need to be implemented in order to achieve satisfactory results. We noticed an improvement in all four cases. However, our eldest patient case 3 showed a slight improvement in the amount of exorbitism. His retrobulbar tissues have made the exorbitism worse but further remodelling of the orbital wall may improve his condition. In addition, he still shows severe lagophthalmos which can only be corrected surgically by several techniques including lateral canthopexy.

Conclusion

We have demonstrated the role of Le Fort III distraction osteogenesis in treating exorbitism. During the course of the treatment numerous observations and questions were raised. The main question revolves around orbital volume changes during Le Fort III distraction. Orbital volume measurements would be effective in predicting the exact amount of distraction needed to correct the exorbitism. The other serious question is does increasing orbital volume actually improve the exorbitism or are we only creating an

optical illusion by advancing the midface? Finally, does the orbital morphology, age and orbital contents play a role in the amount of distraction needed? Further multicenter studies with standard CT protocols are needed to measure the orbital volume changes in relation to amount of midface distraction osteogenesis. We look forward to finding the answers in the near future.

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