Advancement of the midface, from conventional Le Fort III osteotomy to Le Fort III distraction: review of the literature


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Abstract. Since its introduction in about 1950, the Le Fort III (LF III) procedure has become a widely accepted treatment for correction of midface hypoplasia and related functional and esthetic problems. As long-term surgical experience grows and improvements are made in technique, equipment and peri-operative care, the number of LF III procedures performed worldwide is increasing. A number of fundamental questions concerning the technique remain unclear, and large, conclusive studies are lacking owing to the relative rarity of severe midface hypoplasia. This literature review aims to address problems, such as the indication field, timing of surgery, rate of relapse and the use of distraction osteogenesis. An overview of the history and technique of LF III osteotomy and distraction is provided, together with a comprehensive review of the available clinical data.

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aim is to provide more insight into problems related to indications, surgical technique and relapse.

History

Conventional LF III osteotomy

Owing to the increasing success and experience achieved with LF I osteotomy, attention in the 1950s was turned to developing surgical techniques to cope with hypoplastic midface and/or aberrant skull shapes, such as those seen in patients with CFD syndromes. In this respect Gillies’ reports were breaking new ground. In 1941, as a military surgeon, Gillies performed a refraction of a badly healed traumatic LF III fracture. Nine years after his initial attempt, he pioneered LF III osteotomy in a patient with oxycephaly. The indication for this procedure was marked prognathism and exophthalmus. He mobilized the entire midface, achieved rigid fixation with intermaxillary wiring and maintained this for 5 weeks. Although the operation was successful and esthetically beneficial, considerable relapse, resulting scars overlying the nasomaxillary and frontomalar junctions and damage to the lacrimal apparatus was noted.

Paul Tessier, a French plastic surgeon, operated on 35 patients with various CFD syndromes and standardized the procedures for surgical treatment of many types of deformities. His aims were: to restore a normal projection of the facial skeleton. Obwegeser suggested opening the maxillary arch simultaneously with the combination osteotomy, in case widening of the LF I segment might be necessary to correct the dysmorphia. In 1971, Converse et al. reported another modification, the ‘tripartite osteotomy’, a surgical technique that divides the entire midface into three segments: one central nasomaxillary segment and two orbitozygomatic segments, each separately mobile in a sagittal as well as a transverse or vertical direction. All these modifications aimed to give more remodeling options and thus better esthetic results. Important research into combination osteotomies, together with bimaxillary corrections, was continued by Freihofer among others.

The basic LF III operation is now established, although minor modifications on the surgical technique are still being reported.

History of distraction osteogenesis (DO)

In 1993, Cohen et al. were the first to apply the DO technique to the midface in a 4-month-old boy with unilateral craniofacial microsomia and anophthalmia. In their report they used a buried (intraoral) system of miniature distraction devices that permitted maxillary, orbital and mandibular distraction on the LF III level. Since then, several reports have been published dealing with DO on the LF III level. As experience grew with the technique, research has focused on developing new internal and external devices and optimizing DO protocols. An overview is provided in the Surgical technique section below.

Indications

Advancement of the midface on the LF III level is indicated in those syndromes that include midface hypoplasia involving the nasal and zygomatic complex and bony orbits, for example the Crouzon, Apert and Pfeiffer syndromes. Midface hypoplasia presents with several clinical problems, most notably at the level of the airway, orbits, occlusion and facial esthetics with their associated psychosocial problems.

CFD patients are at high risk for upper airway obstruction and undetected obstructive sleep apnea syndrome (OSAS). Almost 50% of CFD patients develop OSAS and need airway intervention at some time. OSAS can be treated pharmacologically, non-surgically (nocturnal oxygen, continuous positive airway pressure, nasopharyngeal tube) or surgically depending on its severity and cause. The standard surgical procedure to alleviate airway obstruction is tracheotomy, which is used in 17–50% of CFD patients. Major complications occur in nearly 7% of all pediatric tracheotomy procedures in the early postoperative phase and in nearly 5% of procedures in the late postoperative phase. CFD patients are also at higher risk for other airway abnormalities, notably tracheal cartilaginous sleeve, laryngomalacia, tracheomalacia, and bronchomalacia. The complication rate in CFD patients is estimated to be even higher. Timely advancement of the midface with minimal intra-operative strain, enlarging the nasopharynx and the palatopharyngeal space, can allow faster decanulation. Decreasing the duration

Fig. 1. (A) An 8-year-old patient with Pfeiffer syndrome, which involves synostosis of the lambdoid and coronal sutures, hypoplastic shallow orbits and midface hypoplasia. (B) On the lateral radiograph no airway is detected in the nasopharynx (arrow).
of endotracheal intubation improves the patient’s quality of life and reduces long-term endotracheal intubation-related morbidity. In contrast to adults with OSAS, children often manifest a pattern of persistent partial airway obstruction during sleep, leading to obstructive hypoventilation, rather than cyclical, discrete obstructive apneas, making the disease difficult to diagnose. In the infant, leaving OSAS untreated may result in failure to thrive, feeding difficulties, recurrent infections, disturbed cognitive functions, developmental delay, cor pulmonale or infant sudden death.

Clinical findings suggest that frequent desaturations, changes in blood pressure and cerebral perfusion may cause deterioration of vision. A close association between OSAS and raised intracranial pressure has been suggested. The authors’ CFD protocol includes that all patients with clinical signs of OSAS are screened for raised intracranial pressure by the consulting ophthalmologist. In case of papiledema, a sign for raised intracranial pressure, the surgical plan is adjusted according to the neurosurgical indication.

One of the most prominent clinical features of CFD is ocular proptosis with corneal distortion, leading to ocular (sub-)luxation in the most severe cases. Functional loss of vision at the causal orbital level can be due to papiledema as a result of cranial overpressure, corneal exposure and/or amblyopia. Papiledema occurs in 10–15% of untreated CFD patients. Corneal exposure, in conjunction with an affected lacrimal apparatus and inefficient tear film can lead to anatomical loss of vision due to exposure keratitis, keratoconjunctivitis sicca and infection leading to corneal ulceration and cataract. Major visual impairment is due to amblyopia. Strong risk factors for amblyopia include strabismus, hypermetropia, astigmatism and anisometropia, which are more prevalent in CFD-patients than in the non-affected population.

Achieving a balanced, esthetically pleasing appearance is the major factor in determining the surgical outcome satisfaction of the patient, family and surgeon. Several studies have mentioned the negative impact of facial distortion on the mother–child attachment, which occurs during the first year of life. This bond is a major influence on the infant’s early psychosocial development, some authors have advocated surgery in infancy for esthetic and psychosocial reasons.

Recent comparative studies in patients with cleft palate have since shown no long-term difference in mother–child attachment in children with affected facial appearance and controls at 24 months of age, making esthetics an elective rather than a pressing indication for surgery.

Timing of surgery

Posnick wrote in 1997: ‘The current approach to the correction of the deformities associated with CFD is to stage the reconstruction to coincide with facial growth patterns, visceral function, and psychosocial development’. Facial growth occurs in 2 distinct periods; during the first 6–7 years of life, craniofacial growth is mostly determined by growth of brain, eyes and nasal cartilage, leading to satalurial growth. After the age of 7 years, growth occurs because of bony surface deposition or apposition, development of the maxillary alveolar process and enlargement of the nasal cavity. As stated and reviewed below, the CFD patient shows little, if any, maxillary growth during the period of craniofacial growth and development, whether operated or unoperated. No detrimental or beneficial effect of surgery on subsequent growth was seen with CFD patients. The LF III procedure should not be postponed in order not to compromise the inherent growth potential through scarring, as there is minimal inherent growth potential in the CFD midface. One should be aware that repeated surgery is necessary to overcome OSAS, which carries a higher risk of complications.

In summary, midface advancement can be scheduled in the first years of life for absolute indications, such as OSAS or severe exorbitism. If the patient is only mildly afflicted, elective surgery can be postponed until skeletal maturity has been reached after puberty and it can then be performed for relative functional and esthetic reasons. The surgeon should always allow for an individual, patient-based approach towards the best possible treatment.

Midface distraction

Conventional procedure versus DO

DO can achieve advancements exceeding the advancement of the conventional procedure by 2- to 3-fold. This is because DO can overcome the natural soft-tissue resistance by means of gradual stretching and accommodation, generating new soft-tissue (histogenesis) simultaneously with skeletal augmentation. Some authors consider that relapse rates are lower because of this (see Relapse section below). Application of external distraction devices allows for better vector control, making traction more effective and precise. DO is associated with decreased operative and post-operative morbidity. Operation time is reduced, blood loss is lowered, postoperative pain is less and the hospital stay is shorter, while also reducing costs. Morbidity might also be lower because of the lesser degree of undercorrection and the lower relapse rate, often eliminating the need for a second surgical procedure.

Advantages associated with the DO technique mainly involve material-related complications, the need for high patient compliance and the high psychological impact of the treatment, which can lead to difficulties when treating children. Also the need for a second surgical procedure to remove the distractor (in particular with intraoral devices) is a disadvantage. DO can also provoke pseudorelapse when patients undergo surgery in early childhood. The main advantages of the traditional technique are the absence of a distraction device (and thus the associated complications, prolonged distraction period and high patient compliance) and the requirement for a second surgical procedure to remove the device.

Surgical technique

LF III osteotomy is performed following exposure of the frontotemporal skull, lateral orbital region, nasion, zygomatic arch, and the zygomatic body via a coronal incision. The anterior surface of the maxillary antrum can be approached through the gingivobuccal sulcus. Osteotomies, following the LF III – Tessier III design, are then made through the frontozygomatic suture, floor of the orbit, and the nasion using a reciprocating saw (Fig. 2). A cephalo-osteotome is used to separate the vomer and ethmoid from the cranial base in the midline. The pterygomaxillary junction is separated either from the bicomeral approach or the gingivobuccal access. Rowe forceps are then used to mobilize the Le Fort III segment including an maxillary acrylic plate to prevent unwanted fracture of the maxilla (Fig. 3). Mobilization of the midface is an extensive procedure, carrying with it a high degree of morbidity in blood loss. Surgeons have sought less invasive techniques to limit morbidity. The greatest advance has been the advent of DO, eliminating the need for immediate advancement, graft harvesting and immediate

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internal stabilization. SCHULTEN et al. combined the use of an internal and external distractor, called the ‘push-pull technique’, to better control the distraction process and force vectors. In their experience, the use of both types of distractor allows for the advantages of both devices, while the disadvantages are not additive. Combining sagittal and transverse distraction devices is also possible and is called ‘multidirectional DO’. UEKI et al. performed this technique in a patient with Crouzon syndrome by using both a Rigid External Distractor (RED) system and hyrax expansion screw in the maxilla.

DENNY et al. developed ‘rotational advancement’. After standard LF III osteotomy and full mobilization of the midface, an internal distractor is fixated to the zygomatic arch, with only one screw in the anterior plate, which acts as a pivot. A hinge plate is fixed across the fronto-zygomatic osteotomy, and a single axial plate is fixed across the nasofrontal osteotomy, which bends with distraction. The objective is to achieve a differential advancement with enough advancement at the occlusal level to establish class I occlusion and an acceptable esthetic facial contour and profile in cases where there is an unequal severity of retrusion at the orbital, nasal root, malar and maxillary alveolar ridge level. Trials have been undertaken to limit incisions by using an endoscopic approach and to lower morbidity by using ultrasound osteotomes in craniofacial surgery. Following experimental animal studies by STAFFENBERG et al. and MCCARTHY, PELLERIN et al. and LIU et al. performed midface advancement in children aged 6–12 years by applying distraction force to the midface with a midfacial pin but without osteotomy. Computer-aided surgical simulation is now being used in the fully virtual pre-operative planning of complex mid-facial deformities.

Distraction devices

Distraction devices are extraoral or intraoral, and many advantages and disadvantages of both types have been recorded. Of the extraoral distraction devices, two haloframes are commercially available (External Midface Distractor, manufactured by Synthes, Oberdorf, Switzerland and Rigid External Distractor, manufactured by Martin, Tuttingen, Germany). Both have similar advantages: the ability to control and modify the vectors of force during the distraction period, the central distribution of forces, easy application and removal of the device and employability in case of thin cortical bone segments. The disadvantages of the two haloframes include patient discomfort (psychosocial as well as physical), halo-related complications (traumatic injuries, scarring, pin loosening) and the need for an upper dental arch to fix the oral splint. With only bony anchorage paranasally, at the aperture piriformis and in the zygomatic region the mobilized segment can be brought forward successfully. In order to minimize halo-related complications with external distractors, the authors advise taking a CT scan of the cranium preoperatively to detect any possible bony defects.

Several internal devices have been reported. Most consist of two bilaterally placed, bone-attached, standardized or customized plates that can be extended during DO. Advantages of these are their smaller size, better patient acceptance (esthetics as well as physical), independence of the presence of an upper dental arch and lesser major complication rates. Disadvantages include the need for a second intervention to remove the device, the impossibility of adjusting the vectors of force during DO, possible fracture at the zygo-maxillary junction in case of thin cortical bone, technical difficulties in placing the 2 devices bilaterally parallel and applying lateral forces onto the midfacial complex (which undesirably extend the concavity of the advanced midfacial segment) instead of forces with a central action.

COHEN et al. have introduced biodegradable plates for internal distractors, but a
second (minor) surgical procedure is still necessary to remove the distractor screw and cable-drive. Burstein et al. designed a one-stage internal biodegradable device. No long-term follow-up studies with internal biodegradable devices on the LF III level have been published.

Only two published reports have compared external and internal distractors. Gosain et al. consider the RED system to be a viable alternative to internal distraction systems, preferably to be used in older patients. Fearon considers the external system to be superior to internal distraction devices when performing LF III DO. Both authors report that both systems yield stable long-term results.

Complications related to LF III osteotomy

Minor and major complications have been reported with the traditional LF III osteotomy. Minor complications include cutting the infra-orbital nerve, ptosis, strabismus, partial anemia, fracturing the zygoma during mobilization, partial exposure of the nasal bone graft and localized infections/abscesses of the surgical area. Major complications include respiratory distress requiring tracheotomy, gastric stress ulcer development, infection of ventriculo-atrial shunt, generalized infection, subgaleal hematoma, cerebrospinal fluid leakage and fistula and visual loss after retro-orbital hemorrhage. In one case-report, lethal intracranial arterial bleeding was described following a skull base fracture due to perioperative maneuvers (most likely pterygoidmaxillary dysjunction and downfracture manipulation).

Complications related to DO

Concerning the DO procedure several authors report no or only a minimal risk of complications in midfacial distraction. A systematic review by Swennen et al. showed that DO on the midfacial and cranial level in 96 patients was associated with a considerable level of complications. This is supported by a recent report from the authors’ group in which a substantial number of complications in DO on the midfacial level using an extraoral distraction device were recorded. Complications mainly constitute mechanical problems with the distraction device (pin loosening, frame migrations, traumatic injuries, intracranial migration of halo-fixation pins), technical difficulties (including fracture of the zygomaticomaxillary junction, intraoperative fragment disjunction, problems with maxillary splint attachment to the teeth), localized or pin-site skin infections, problems with advancement (less or asymmetrical advancement) and severe infections requiring hospitalization.

Fearon was the first to compare the 2 techniques in two retrospective studies. He concluded that the incidence of complications and length of hospital stay were lower in the distraction groups, while advancements were significantly higher in these groups. Sleep apnea was more successfully corrected by means of DO. According to Fearon, DO should be able to prevent a second distraction procedure. Fearon recommended the use of DO on the midfacial level in younger patients with more severe retrusions of the midface, which need greater advancements than can be achieved using the conventional method. Nevertheless, the conventional procedure is recommended in patients who need moderate advancement (8–10 mm) and who have completed growth.

Relapse

Conventional LF III and DO

Long-term follow-up studies on LF III osteotomy that include a substantial number of patients are rare. Considering the various studies available, the authors conclude that the standard LF III procedure provides a relatively stable postoperative position of the midface. Relapse, when it occurred, could be attributed to inadequate postoperative fixation leading to backward rotation of the midface at the level of the orbits or to ’pseudo-relapse’, defined as relapse at the occlusal plane because of normal mandibular growth combined with decreased maxillary growth. Pseudorelapse is observed in patients who were operated on in childhood and can be corrected successfully by a LF I procedure after skeletal maturity. Studies agree that the conventional LF III advancement procedure renders stable results with regard to the position of the skeletal midfacial segment, irrespective of the various cephalometric landmarks and analyses used by the different authors.

Since the introduction of the DO technique on the midfacial level in 1993, only a few reports have been published dealing with its long-term stability. All these studies report minimal or no relapse in conjunction with DO of the midface. In contrast with conventional osteotomy, no statements are made in these reports about postoperative retention. The authors’ CFD protocol includes a 1-year retention phase using night-time face-mask traction.

Responding to a questionnaire, 31% of craniofacial surgeons reported relapse of the midface with DO in their practice. It is unknown whether this observed relapse was assessed subjectively or objectively. Most respondents encountered relapse within the first 6 months after finishing DO.

Postsurgical growth

There are contrasting views about postsurgical growth of the midface portion. When considering postsurgical growth it is essential to consider the presurgical/normal growth potential of CFD patients. Bachmayer et al. established the growth potential of 52 unoperated CFD patients.
Horizontal growth was measured as the horizontal distance from basion to A-point. Their findings indicate that the horizontal growth of CFD patients is about 0.7 mm/year. Kreiborg et al. and Meazzi et al. attribute the measured growth to the posterior cranial base, and state that measurements of the midfacial horizontal growth in these patients towards the anterior cranial base (sella-nasion line) showed no sagittal displacements of A-point; they conclude that sagittal growth in unoperated CFD patients is negligible. Significant vertical growth was measured in these patient groups, irrespective of the use of different cephalometric tracing methods. Bachmayer et al. report a vertical lengthening of Anterior Nasal Spine (ANS) towards the true horizontal, and both Meazzi et al. and Kreiborg et al. report a discrepancy between the anterior and posterior vertical lengthening. A greater increase in the distance from ANS to the anterior cranial base was found, when compared with the distance of the posterior occlusal point to the anterior cranial base. As horizontal growth turns out to be nil, vertical growth seems to be preserved in unoperated CFD patients, stressing the importance of considering sagittal growth in its distinct components.

Considering presurgical growth data, a further deterioration of craniofacial growth in CFD patients is not expected. Several authors report some postsurgical sagittal growth of the midface, but do not differentiate between horizontal and vertical growth. Some vertical growth is to be expected, whether the patients undergo surgery or not, owing to remodeling and appositional growth rather than to sutureal growth. Fearon compared postsurgical growth between conventional and distracted LF III osteotomy, is performed in a second surgical procedure, but in the authors’ opinion always after the age of skeletal maturity. Close cooperation with the orthodontist of the craniofacial team is mandatory to plan the surgery with pre- and postoperative orthodontic treatment; patients should be seen together in the peri-operative phase.

There is no consensus on the growth potential of the midface after surgery therefore decisions and timing of surgery before skeletal maturity should be strictly bound by the indications. Absolute indications for surgery are OSAS and ocular proptosis with corneal distortion as a result of orbital deficiency. Younger patients are generally treated with DO to achieve the greater advancement and overcorrection they need in order to correct the OSAS effectively and compensate for future restricted growth. Little information is available regarding the impact of the clinical signs of OSAS and abnormal outcomes of the polysomnography in CFD patients. It is unclear how aggressive one should be with the diagnosis of even mild OSAS in order to prevent irreversible damage. It is also unclear how much advancement is necessary to correct the OSAS. With bronchoscopy and CT scanning the upper airway can be monitored more precisely and volumes can be measured. These outcomes could be linked to the results of the polysomnography. With improved imaging techniques, the size and shape of the distraction segment can be investigated, giving insight into the long-term stability of the segment in relation to the surrounding tissues.

The authors recently observed growth retardation of the mandible and functional pharynx problems possibly contributing to persistent OSAS, despite considerable advancement of the midface with DO, in patients with Apert and Crouzon syndromes. Bronchoscopy before midface advancement is advised to monitor all possible levels of obstruction. In a large prospective study of CFD patients the relation between OSAS and raised intracranial pressure is being investigated in the authors’ Craniofacial Centre in an attempt to elucidate the pathophysiologic pathway of OSAS leading to raised intracranial pressure and vice versa.

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