Frontofacial Surgery in Children and Adolescents: Techniques, Indications, Outcomes

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Abstract

The techniques of frontofacial surgery are most valuable in the clinical management of complex craniofacial deformity to achieve a range of functional and aesthetic gains in children from infancy to maturity. A variety of complex craniofacial osteotomies that can be used to separate the orbits from the skull base have been described. In addition, the combination of circumorbital release and pterygomaxillary disjunction allows advancement of the orbitomaxillary segment for powerful clinical benefit. For the purpose of this article, the principal frontofacial strategies include the monobloc frontofacial advancement by distraction (MBD), frontofacial bipartition advancement by distraction (BpD), orbital box osteotomy (FFBx), and frontofacial bipartition (FFBp). These techniques are broadly used for two purposes: to allow for the translocation of one or both orbits to correct orbitofacial disproportion (hypertelorism, vertical orbital dystopia, or a combination of both), or to advance the orbitomaxillary segment for orbital volume expansion and protection of the eye in syndromes featuring severe exorbitism (oculo-orbital disproportion).

Here we describe aspects of our experience of frontofacial surgery in the Craniofacial Centre at Great Ormond Street Hospital for Children, London, with reference to the principles underpinning frontofacial surgical techniques, their challenges, and their impact on function and aesthetics.

Keywords

- Apert syndrome
- Crouzon syndrome
- Pfeiffer syndrome
- hypertelorism
- orbital dystopia
- monobloc

Techniques of frontofacial surgery have been in use for syndromes of craniofacial disproportion and its effects upon the function and protection of the airway, breathing, the ocular surface and vision, as well as oropharyngeal function since the mid-1960s. Craniofacial surgery in its contemporary form developed from the innovation of Paul Tessier, whose pioneering surgeries opened a new quality of outcome to patients with syndromic craniosynostosis, hypertelorism, Treacher Collins syndrome, and other syndromes of craniofacial difference.¹ The subsequent establishment of several craniofacial teams led by a new generation of innovators such as Marchac, Van Der Meulen, Ortiz-Monasterio, Kawamoto, and McCarthy established the specialty of craniofacial surgery internationally. Contemporary craniofacial

teams are comprised of a wide number of disciplines and reflect a constantly evolving expertise and technology in every aspect of patient care, from assessment to surgery and the evaluation of outcome. It is this latter aspect of the patient pathway that challenges craniofacial teams in current health care systems that rightly demand value and the practice of evidence-based medicine. Therefore, craniofacial surgery in general has evolved from description of technique to evaluation of outcome across a broad range of disciplines.

What Is Frontofacial Surgery?

Frontofacial surgery includes those operative techniques that separate the intact orbits from the anterior skull base to move

the orbits in relation to each other or as a combined unit with the maxilla in relation to the basicranium. The orbits may be translocated independently (orbital “box” osteotomy), or advanced as a “monobloc” with the maxilla and upper dental arch.

The Circumorbital Osteotomy

The liberation of the orbits from the anterolateral skull base is common to all the principal frontofacial operations: monobloc frontofacial advancement by distraction (MBD), frontofacial bipartition advancement by distraction (BpD), orbital box osteotomy (FFBx), and frontofacial bipartition (FFBp).

The essential osteotomies that achieve this are across the floor of the anterior cranial fossa (requiring transcranial access by removal and replacement of a frontal craniotomy) and down the lateral orbital wall (greater wing of the sphenoid and in addition the zygomatic arch) to meet the orbital floor at the anterior aspect of the inferior orbital fissure (►Fig. 1). The medial orbital wall is sectioned from the anterior cranial fossa (anterior to the cribriform plate) downwards in the coronal plane posterior to the posterior lacrimal crest to reach the orbital floor and then the level of the inferior orbital fissure to complete the internal circumorbital osteotomy part of the surgery. This will allow mobilization of the palpebral complex without medial canthal detachment, although canthal reinforcement may be required.²

Common to all the frontofacial osteotomies, the midline anterior cranial base cut is placed anterior to the foramen cecum remnant, to avoid risk to the dural reflection and the

cribriform plate (risk to olfaction and a cranial base cerebrospinal leak). If a fontal sinus is present, and the anatomy allows, this cut might be made within the sinus, anterior to its posterior wall, and therefore extracranially.^{3,4}

The detail of the lateral wall cut depends on the detail of what is to be achieved (►Fig. 2). For frontofacial advancement, the cut is made more anteriorly in the lateral wall to allow skeletal advancement of the lateral orbital margin and the Whitnall tubercle, but “leave the globe behind.” In addition, in these syndromic craniofacial dysostosis patients, the temporal lobe bulges forward in the middle cranial fossa and the greater wing of the sphenoid lies at a shallow angle. If the lateral wall cut is made too posteriorly, the middle cranial fossa is breached, risking dural and brain injury. Furthermore, carriage of the lateral posterior wall of the orbit forward in the monobloc will tend to translocate the globe forward also—reducing the clinical benefits of the surgery.

For frontofacial orbital translocation, the aim of the orbital movement is to carry the globe and contents of the orbits; therefore, the lateral wall osteotomy is placed behind the equator of the globe. The aim here is not the orbital expansion of a dysplastic orbit, but orbitopalpebral translocation as a unit (►Fig. 2).

The details of the subsequent osteotomies depend upon the requirements of the surgery: either to move one orbit or both in relation to each other and the skull base as orbital box osteotomies,^{5–7} or to move the orbits together, with the maxilla as a monobloc,^{8,9} which itself can be divided into two as a facial bipartition.^{9–12}

The Box Osteotomy

Movement of the orbits (without the maxilla) as orbital box osteotomies is commonly undertaken for medial and/or vertical translocation of the orbit, behind the equator of the globe, so the consequent globe, canthal, and palpebral

Fig. 1 Three-dimensional skull model. Red line shows ‘internal’ circumorbital osteotomies: orbital roof (accessed via transfrontal osteotomy), to medial wall posterior to posterior lacrimal crest, to orbital floor through anterior aspect of inferior orbital fissure, to lateral wall and back up to orbital roof. The blue line shows the osteotomy for one type of frontofacial box (see text).

Fig. 2 Axial computed tomography scan. Single red line shows medial wall osteotomy through ethmoid complex, posterior to the posterior lacrimal crest. Double red line shows lateral wall osteotomy, the anterior line as for frontofacial monobloc or bipartition distraction to carry the bone but not the globe; the posterior line as for box osteotomy to carry the contents of the ‘effective orbit’.

movements result in greater midfacial symmetry. The orbits are thus moved in relation to each other and separately from the maxilla.

This requires a bone cut across the anterior wall of the maxilla, protecting the infraorbital nerve (which may be included or excluded from the box, but must not be sectioned). This osteotomy extends from the zygoma laterally to meet the nose medially at the base of the pyriform aperture. The cut can be varied across the zygoma specifically to narrow or leave widened the bimalar width of the face.³ The pyriform margin and the nasal margin form the medial part of the anterior aspect of the box up to a point at which an osteotomy is undertaken through the nasal process of the maxilla and nasal bones up to the anterior craniotomy cut, which has already been made for transcranial access. The orbit is thus medialized according to how much bone is resected in the midline (anterior to the cribriform plate), and whether a midline strut of bone is left as a centralizing point of fixation to which to bring the medialized orbital segments (► Fig. 3). Much depends upon the specific requirements of the surgical plan.

Orbital box osteotomies are valuable for the correction of symmetric or asymmetric hypertelorism in a variety of craniofacial syndromes, for the correction of vertical orbital dystopia, or in conjunction with segmental cuts for orbital expansion in micro-orbitism/microphthalmos syndromes.

Monobloc and Bipartition

As a different strategy, movement of the monobloc (intact orbito-maxillary-nasal unit) specifically does not demand osteotomy of the anterior wall of the maxilla. Instead, a pterygomaxillary dysjunction is made at its junction with the pterygoid plates (pterygomaxillary fissure), and continued up into the inferior orbital fissure. The orbital margins (the two orbits are mobilized together as a “pair of spectacles”) retain continuity with the maxilla via its medial and lateral buttresses and the maxillary sidewall. The maxilla is

sectioned from the cranial base at the pterygoids and is therefore freed to mobilize anteriorly with the orbital margin, leaving the lateral wall (greater wing of sphenoid) and medial wall (ethmoid complex) behind (► Figs. 4 and 5).

Separation of the orbital spectacles segment from the anterior skull base is completed by cuts across the floor of the anterior cranial fossa, anterior to the cribriform (or via the frontal sinus), and caudally through the nasal septum to a level above the palate. This cut generally joins the medial orbital wall cuts, thus bringing the nose forward with the spectacles segment.

Monobloc osteotomies and advancement of the orbito-maxillary-nasal unit by external frame distraction osteogenesis are fundamental to the contemporary management of the syndromic craniosynostoses such as Crouzon and related syndromes. The primary clinical benefits of monobloc advancement by distraction relate to the orbital expansion achieved, which corrects oculo-orbital disproportion, globe subluxation, and ocular surface exposure from severe exorbitism. Anterior movement of the orbital margins (spectacles segment) with the medial canthus, lateral canthal resuspension, and the Whitnall apparatus renders competent, protective, palpebral sphincter function of the eyelids. Additional benefits include airway expansion at the level of the nasopharynx, and a degree of volume expansion at the anterior skull in the treatment of syndromic intracranial hypertension. The advancement of the maxilla tends to correction of the skeletal class III relationship; however, definitive correction of this is achieved by subsequent orthognathic and orthodontic techniques. Postoperative orthodontics may be required to realign the dentition, and also affords the opportunity to correct the interocclusal relationships in isolation or in conjunction with orthognathic surgery.

The monobloc may be additionally divided in a midline plane to separate the facial dysjunction into two halves as a monobloc bipartition by a simple vertical osteotomy from the frontal craniotomy cut to the nasal keystone at the

Fig. 3 (A–C) Three types of box osteotomy design. (A) Midline excision of bone, osteotomies to carry the zygoma in entirety and narrow the bimalar distance. The anterior maxillary osteotomy is a closing wedge below the infraorbital nerve. (B) The inferior border of the zygoma is not carried, leaving a wider bimalar width (suited a male face). The osteotomy is superior to the infraorbital foramen, as might be the case with a high canine tooth root. (C) Asymmetric design with microphthalmos. A midline strut is preserved as a reference point to build toward in the reconstruction.

Fig. 4 (A–E) Shows complete monobloc osteotomies through the (A) orbital roof, (B) laterally through the lateral orbital walls, (C) the zygomatic arch, (D) the pterygoids, and (E) showing the distraction wires in the medial buttress of the maxilla.

osseocartilaginous junction of the nose, and from the anterior nasal spine to the alveolar margin between the central incisors. By designing the cut in the upper middle third of the face to remove bespoke amounts of bone in a “V,” the technique can be used to treat symmetrical forms of axial

Fig. 5 The three-dimensional computed tomography scan in lateral view shows the frontal craniotomy and plate fixation to the monobloc segment in an advanced position. The zygomatic arch is shown sectioned in lateral view, and the advanced maxillary position optimizes the jaw relationship for subsequent orthognathic surgery.

hypertelorism or to correct symmetrical exorotational orbital deformities such as in Apert syndrome (►**Fig. 6**). The V-shaped bone excision creates a fulcrum at the rhinion such that the medialized orbits travel through an arc of rotation to close in the midline. Closing this wedge osteotomy in the upper middle third of the face opens a wedge in the midsagittal osteotomy of the maxillary alveolus that is the lower part of the bipartition. The characteristic biconcave facial form of Apert syndrome responds particularly well to the bipartition osteotomy in combination with advancement by distraction.

Preparation for Surgery

Frontofacial techniques can be used in the management of crisis events such as recurrent ocular subluxation, for urgent interventions for functional gain such as for nasopharyngeal airway expansion, or perhaps more commonly for elective functional improvement. In the later childhood or teenage years, frontofacial techniques are used to improve aesthetic balance and the attendant psychological gain.

In our practice, we have evolved a 3-day craniofacial assessment (CFA) in which patients see all the members of the multidisciplinary team (MDT) in their own clinics over a concentrated period, culminating in a summation meeting by the MDT to review the opinions and data collected. The

Fig. 6 The facial bipartition. The frontal “D” craniotomy has been performed. The facial segment has been cut as a monobloc, with a central V of bone removed from craniotomy cut to the rhinion (osseocartilaginous junction of the nose). The shape of the V determines the arc of medialization of the orbits.

decisions made at that meeting are recorded in the chart, and our clinical nurse specialist calls the family at home to discuss the outcome. In this way, the family sees all MDT members individually but contemporaneously without the potential inefficiencies of a daunting single clinic roomful of medical team members. The CFA is a valuable tool in data collection for frontofacial surgery and helps to prepare the family and answer questions arising during a concentrated experience with the team. Sessions with the ward and specialist nurses are invaluable in preparing children for the perioperative experience. Clinical psychologists with expertise in managing the impact of life with a facial difference discuss the family's long-term expectations for change and offer help with managing anxiety, boosting confidence, self-esteem, and any difficulties with peer relationships. For those children undergoing advancement of the monobloc or bipartition osteotomies, there is an opportunity to familiarize with the external distraction system, and speak to other children who have undergone similar procedures.

Data are collected in a wide range of disciplines including psychological assessment; speech, language, and feeding skills; ophthalmic parameters including ocular movement, the state of the ocular surface, and a comprehensive vision assessment; airway and hearing analysis; and genetic advice. Any missing investigations such as radiology and sleep-study analysis can be undertaken during the CFA, and are reviewed with the clinical reports at the summation meeting. The CFA report is a means of communication between specialists in the hospital, local caregivers at the district level, and the family. The standardization of the data collected across the wide range of disciplines acts as a longitudinal record of progress.

Complex orthodontic preparation is often a useful preparation for elective facial bipartition surgery to protect the central incisor teeth against damage from the midsagittal osteotomy in the maxillary alveolus. The aim is to create a diastema between the central incisors through which a safe vertical osteotomy can be made, and this is usually done with

fixed appliances to the upper dental arch over a period of several months. In these bipartitions, the closing wedge osteotomy of the upper midface segment creates an open wedge gap in the alveolus. Our experience is that this closes spontaneously under the closed mucosa with an upper arch collapse. Postoperative orthodontics is only occasionally required to realign the occlusion.

The preparation and postoperative care pathway create a potential burden of care of clinic appointments, investigations, and assessments, which can be tiring and costly for families often traveling some distance to the hospital. Good communication with local pediatricians, orthodontists, speech and language therapists, and the school systems is essential. The number of appointments can serve to increase both frustration and expectation of the postoperative results for both function and aesthetics, and the role of psychological assessment and preparation for frontofacial surgeries cannot be underestimated. A current focus of research and audit activity is to match our functional outcome data with patient-reported outcome measures in all forms of frontofacial surgery.

Craniosynostosis Syndromes

Advancement of the orbitomaxillary monobloc in craniofacial dysostosis syndromes is a valuable tool for clinical gain across several parameters. Expansion of the orbital volume effectively treats the oculo-orbital disproportion and midface retrusion with extreme negative vector that causes exorbitism, and at its extreme, the subluxation of the globe onto the cheek. Such ocular dislocation is caused by a lack of skeletal support to the eye from an orbital margin that lies behind the equator of the globe, with a shallow orbit and oblique greater wing of the sphenoid creating an angled lateroposterior wall of the orbit.¹³ This is a common feature of severe infant Crouzon and Pfeiffer syndromes (→Fig. 7). Although temporizing measures such as tarsorrhaphy can bring the eyelids into a protective position over the ocular surface, these are self-limiting by the effects of stretch and dehiscence, and potentially morbid by increasing intraocular pressure. The dislocation of the globe across a reduced palpebral fissure after tarsorrhaphy may render globe relocation at the bedside impossible and endanger the sight in that eye.

Under these circumstances, frontofacial monobloc for crisis intervention can be extremely valuable, even in the very young (our experience includes 23 monobloc distractions in children younger than 5 years old, with 10 children less than 2 years of age).¹⁴ The monobloc osteotomies and some on-table advancement will provide immediate operative orbital expansion, and take the pressure off on-table tarsorrhaphies. The use of external frame distraction osteogenesis with 5-point fixation in the face¹⁵ gives a further controlled advancement and brings the orbital margin anterior to the equator of the globe, resolving the negative malar vector and rendering the eyelid position favorable for a protective palpebral sphincter. Distraction is usually delayed for a few postoperative days, and then commenced at 1.4 mm per day.

Fig. 7 Clinical image of child with Crouzon-Pfeiffer syndrome, with constricted orbital hypoplasia and oculo-orbital disproportion. The constricted orbit predisposes to palpebral insufficiency, ocular surface exposure, and extremely ocular subluxation. The panel of axial computed tomography scans demonstrates the inadequacy of orbital volume and shape and degree of exorbitism, with skeletal support behind the equator of the globe.

Although not as effective as posterior cranioplasty for vault expansion,¹⁶ the expansion of the anterior cranial fossa that accompanies monobloc advancement is a useful adjunct for the management of intracranial hypertension in syndromic craniofacial dysostosis. In addition, monobloc advancement may have beneficial effects upon the airway, such that tracheotomy can be reversed or avoided. These gains are very valuable in elective or urgent surgical planning in complex cases of Crouzon and Pfeiffer syndromes, where multifactorial problems of interlinking airway compromise, intracranial hypertension, and ocular surface protection often occur. Monobloc advancement for these purposes can be valuable for functional compromise in infancy and childhood.

Often, in the less-severe phenotypes of the spectrum of Crouzon and Pfeiffer syndromes and their related syndromes, the approach to monobloc advancement surgery will be based upon the patient's drive to achieve improved facial balance. Frontofacial surgery delivers favorable aesthetic change in many congenital syndromes, and can facilitate

integration of the more confident individual into an uncompromising society. A keystone of preparation for aesthetic frontofacial surgery includes comprehensive psychological preparation and the management of expectation in both adolescent and family, which includes building psychosocial resilience and offering specific strategies for managing changes to appearance.

It is important to remember that syndromic craniofacial dysostosis takes many phenotypic forms, all closely related and interlinking both in appearance and genetic cause. The shape and form of the face is variable, and not all such faces respond equally in aesthetic terms to the monobloc advancement. Aesthetic adjuncts such as mid-facelifts, eyelid and canthal surgery, browlifts, and aesthetic orthognathic surgery are often offered to these patients to improve facial balance and ameliorate the stigmata of the syndrome. In Apert syndrome, in particular, the narrow genotype generates a restricted range of facial phenotype that responds very well to monobloc bipartition with distraction. The Apert face is biconcave in axial and sagittal planes, and the advancement of the facial halves of the monobloc bipartition successfully breaks up the concavity, also allowing medial rotation of the orbits and correction of the lateral facial cant and negative canthal axis (►Fig. 8). The bipartition advancement of the Apert midface changes orbital shape, treating the oculo-orbital disproportion (as well as providing medial orbital translocation), and providing relief of the ocular surface symptoms of exposure.¹⁷

Orbital Translocation for Hypertelorism and Vertical Orbital Dystopia

Orbitofacial disproportion may be a vertical asymmetry (vertical orbital dystopia) or a horizontal (axial) feature. In complex craniofrontonasal dysplasia, there may be aspects of orbital disproportion in vertical, axial, and rotational planes, and also the plane of anteroposterior tilt of the face. Hypertelorism (hypertelorbitism) has been variously defined,^{18–20} but is essentially characterized by the wide horizontal separation of a relatively normally configured orbit. The relationship between the medial and lateral walls of each orbit approaches normal, whereas the bony interorbital distance (anterior lacrimal crest to anterior lacrimal crest), and therefore the intercanthal distances are excessive. Furthermore, hypertelorism can perhaps be usefully described as symmetrical axial, symmetrical rotated (in coronal plane), asymmetrical symmetrical rotated, and multiplanar. This approach classifies the clinical nature of the orbital relationship rather than the genetic/phenotypic classification by disease; thus, it is useful in surgical planning.³

Symmetrical axial hypertelorism requires medialization of the orbits along a symmetrical axial plane (►Fig. 3 middle). Either a horizontal sliding box osteotomy (medialization in horizontal plane) or facial bipartition (medialization in a gentle arc of rotation) might be considered, depending upon the circumstances. Key determinants are the age of the patient and the state of the dentition, as well as features of the underlying syndrome or cause. Facial bipartition in

Fig. 8 (A,B) Vertex view of an adolescent with Apert syndrome, (A) pre- and (B) postfrontofacial bipartition. The midface has been advanced differentially and the upper eyelid given a mechanical and functional advantage. The lateral canthus has been elevated relative to the medial canthus, and the nose is advanced. Nasal and forehead balance is improved.

midline clefting syndromes is facilitated by the cleft. In younger patients, the unerupted permanent dentition is high in the anterior maxilla and is vulnerable to the use of a box osteotomy—which must be deferred until the canine root descends. This allows enough space below the infraorbital margin to complete the anterior maxillary horizontal cut while minimizing tooth or infraorbital nerve damage. Generally, the descent of the secondary dentition allows box osteotomy by the age of 8 to 9 years where indicated.

Whereas box osteotomies do not require disruption of the upper dental arch, facial bipartition requires a midline palatal alveolar split; in our unit, this is facilitated by orthodontic preparation. Orthodontic techniques create a midline interdental space, or diastema, between the central incisors. Such a separation of the teeth in the midline protects the teeth and periodontal tissues from damage during the osteotomy, preventing the risk of tooth loss or ankylosis. The orthodontic preparation requires fixed upper arch appliances, is generally coordinated between the craniofacial orthodontist and local practitioners, and adds to the burden of care in preparation for the hypertelorism correction.

Medial orbital translocation by facial bipartition will necessarily rotate the orbit and generate an upslanting or “positive” canthal axis (upwards angle from the horizontal plane), which can have desirable effects in symmetrical axial hypertelorism as well as symmetrical rotated hypertelorism. The amount of rotation can be fine-tuned by the width of the V-shaped midline bone resection at the nasion, and its fulcrum at the rhinion (osseocartilaginous junction) of the nose (► Fig. 4). The wider the V resection, the greater is the orbital rotation and the tendency to a positive canthal axis. There is a coincident increase in upper facial height and an increase in upper dental arch width. The upper dental arch generally tends to collapse passively after facial bipartition without orthodontic encouragement. By contrast, box osteotomy for symmetrical axial hypertelorism narrows the upper face width and does not lengthen the face (► Fig. 3). There is no interruption of the dentition requiring no orthodontics. The choice of box versus bipartition for symmetrical forms of axial or rotated hypertelorism thus reflects the age of the patient,

the desired effects upon facial height and width, decision making about the dental arch, and the degree of orbital rotation required.

Box osteotomy, by facilitating independent movement of the orbits, is a particularly valuable option in asymmetric rotated or multiplanar hypertelorism (► Fig. 3). The position of the liberated orbit and palpebral apparatus can be independently referenced to the contralateral side and the nose. Box osteotomy allows correction of orbital cant or tilt in three planes, which would not be possible by facial bipartition without a major disruption to occlusion.

As the orbits move, the soft tissue envelope must be redraped appropriately for the function of the eyelids and the aesthetics of the cheek, brow, and canthi. The lateral canthus is degloved and refixed for the circumorbital osteotomy and to restore the tension and position of the lower eyelid. Medial canthal fixation is not degloved, but may require reinforcement. The soft tissues of the brow and midface are usually resuspended to restore volume, vector, and symmetry. Where large medial translocations are undertaken, there is midline soft tissue excess and this may be resected by elliptical excision leaving a straight midline scar. Alternative strategies to achieve soft tissue contraction include the use of splints and internalized mattress sutures.²¹ Often, the midline soft tissue excess also may be accommodated by nasal augmentation or reconstruction with a cranial bone graft to the nose at the time of hypertelorism surgery. Secondary surgical modifications include fat transfers for volume augmentation and the use of flap transfers. Attention to detail in these aesthetic adjuncts can make a great difference to the patient and perception of result.²

Although a few children who present with hypertelorism remain unclassifiable by diagnostic group, the majority of children who come to hypertelorism correction by box or bipartition surgery have a syndromic diagnosis.

Vertical orbital translocation by box osteotomy is a valuable strategy in hemifacial microsomia, late-operated unicoronal synostosis, and some skeletal dysplasias such as fibrous dysplasia. The circumorbital osteotomies are made as for horizontal box osteotomy, but the translocation is a

Fig. 9 (A–C) Intraoperative panel. (A) Coronal flap reflected to show ink marks of planned orbital access and expansion with elevation. The periorbital soft tissues fill the orbit in the reflected soft tissue flap. (B) Frontal craniotomy removed for orbital roof access. (C) Postreconstruction with frontal craniotomy replaced and fixed with the supraorbital margin elevated (black arrow). The periorbital are flanked by a medial space (white arrow) showing the effective orbital expansion.

vertical one, with the resected bone superiorly being used for the bone gap created inferiorly. In hemifacial microsomia, the vertical translocation can be accompanied by orbital expansion with osteotomy to open up a contracted socket (→ Fig. 9), to insert a prosthesis, or allow conjunctival sac expansion and a conformer.

Globe movement and vision may be negatively affected by translocation of the orbit and globe. A third of hypertelorism patients presenting for orbital translocation without advancement have stereoscopic vision, with visual and motor fusion for both aesthetic balance and binocularity. Visual tolerance for change in visual axis is greater for horizontal movements than for vertical movements, where small change may precipitate intolerable double vision. Where presurgical vision is bilaterally independent, and there is no visual or motor fusion experience, orbital translocation and a shift in globe position will not restore binocularity, despite bringing the globes into better alignment. In these patients, secondary squint surgery is for aesthetic gain.²²

Careful preassessment by orthoptist and ophthalmologist is therefore required to prevent the risk that orbital translocation for aesthetic reasons will not disrupt visual function. What is not determined is whether early orbital translocation for ocular balance, followed by strabismus surgery, can rescue the potential for any level of binocularity, prior to the maturation of postretinal visual pathways.

Outcomes and Adversities

The risks and responsibilities of frontofacial surgery are many.²³ However, in common with many areas of surgical

practice, a concentration of experience in dedicated high-volume craniofacial centers allows the audit of properly collected outcome data. The government of the United Kingdom has recognized this and funds craniofacial care for designated conditions in regional pediatric hospital hubs. A review of the practice at the Craniofacial Centre, Great Ormond Street hospital has been instructive. We find that frontofacial surgery carries a major adversity rate of 11%: 114 frontofacial cases over a 12-year period, including monobloc and bipartition advancements ($n = 83$) and box or bipartition procedures for orbital translocation without advancement ($n = 31$), where major adverse events are for life-saving intervention such as unplanned tracheostomy, revision of transcranial procedure, or death ($n = 1$) and permanent disability. The overall infection rate runs at approximately 25% of cases. Transfusion requirements are often high, especially in revision cases; however, the use of autologous intraoperative transfusion and the cell saver has been valuable and can avoid the need for donated blood for frontofacial surgery in the optimized patient. The risk of cerebrospinal fluid leak is insignificant in nondistractive cases, rising to a third of distractive cases; however, the real morbidity that results is generally minor, and the majority of these leaks resolve spontaneously. It does not seem to be an independent variable in the length of hospital stay of distractive (24 days) over nondistractive cases (14 days). In broad terms, orbital translocation by either technique is more predictably free of complication than monobloc or bipartition surgery advanced by distractive. What is clear is that the gains of frontofacial surgery are functional—measured in terms of airway, vision, and overall performance—and aesthetic—as reported by

independent review, patients, and stakeholders. A robust set of patient-reported outcome measures is clearly required to be able to quantitatively rate the reported highly positive outcome of these surgeries against the attendant risks.

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