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both approaches. It is a matter of what works best for a particular surgeon. In 90% of simple craniosynostosis cases we obtained good results with one procedure. However, in 10% of our simple cases we had to reoperate to get what we consider a good satisfactory result.

In complex orbital and hypertelorism surgery we do a true three-dimensional correction. If we look at our results critically, we can see these results can also be improved. The same holds true for both the bipartision procedure and the monobloc procedure. Most of our infections have come in these two procedures. The problems of optic atrophy and increased intracranial pressure are also to be taken into consideration. Again, I believe that the monobloc procedure is a high-risk procedure and I would prefer to do it in two stages: the forehead advancement first followed by the jaws. Our general impression is as such: repair the deformity as early as possible (remodeling and reshaping) and then see if this result is satisfactory.

Craniofacial surgery today offers us extensive modalities for reconstruction. However, there is a problem of the skin envelope, and in the future we are going to have to make improvements in the care of the skin envelope. Postsurgically, there is definite growth following infant craniosynostosis correction. The problem of abnormal growth leads us to further evaluation of these patients. On our team an anthropologist takes measurements of the soft tissues of the face. This is where we assess our final results. Our craniofacial surgery standards should be the same as they are for all aesthetic surgery. We should be able to change grotesque deformities into attractive functioning people.

The face is a mask which can be altered by changing the underlying craniofacial structures. But we cannot just take a congenital deformity, cut the bones, and move them into a new position expecting to get a good result. We have to combine cutting, molding, and reshaping of the bones with careful, accurate soft tissue surgery. Many of the surgical procedures in this field are complex and require a dedicated effort. While this is different from aesthetic surgery, the standard should be the same. In the future craniofacial surgery will include soft tissue correction and our results will improve with experience. Finally, the psychological counselling given to our craniofacial surgery patients provides needed support for improved enjoyment and quality of life.

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Treatment Concepts for Craniosynostosis and Hypertelorism

Introduction

The Department of Maxillo-Facial Surgery of the Zürich University Hospital has for the last 20 years mainly focused its attention on craniofacial surgery. Corrections of the five levels of the craniofacial framework (Figs. **1a–c**) have been performed in over 1000 cases using established craniofacial surgical methods (3, 6, 14, 23, 24, 25), individual modifications thereof or totally new methods developed in Zürich (11, 13, 19, 20).

In the following presentation we detail our treatment concepts which are based on long-term results (2, 18) and on the complications we have experienced over many years (1, 12, 16, 21, 22).

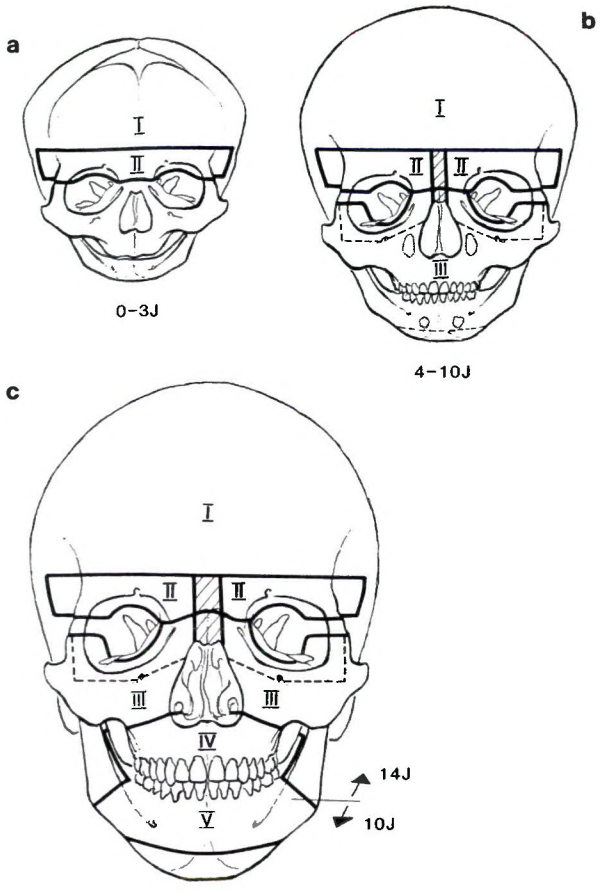


Fig. 1 Surgical correction of the 5 levels of the craniofacial framework at age groups 0 to 3 years, 4 to 10 years and older than 10 years. **a** Up to 3 years of age only corrections of the levels I and II. **b** Between 4 and 10 years corrections of levels I, II, and III. **c** Over 10 years of age surgical corrections of all levels can be performed excepting mandibular retropositioning which should not be done before age of 14 years.

Timing

The timing for the correction of craniofacial stenosis and other related anomalies has been developed as follows:

1) Surgical procedures in the first year of life (Fig. **1a**):
   a) Correction of the first and second level in patients with increased intracranial pressure and extreme exophthalmus.
   b) Correction of the clover-leaf deformity.
   c) Correction of occipital plagiocephaly by the floating occiput method (5). The anterior position of the pyramid complex on the affected side shows backward movement and normal development of the medial cranial fossa, if operated early.
   d) A tracheostomy is performed in cases of severe airway obstruction. This facilitates an excellent examination of the young patient, enables better general development, and is simultaneously a prophylactic measure against repeated asphyctic attacks and chronic bronchial infections. The Le Fort III-osteotomy in this stage is not indicated and should be postponed; tooth-germ damage is almost always certain.

2) Surgical procedures performed between the first and third year of life (Fig. **1a**):
   a) Correction of brachycephaly, oxycephaly, and plagiocephaly affecting the craniofacial levels I and II when *not* accompanied by hypertelorism (Fig. **2a–b**). The correction of cases with hypertelorism should be postponed (see paragraph 3).
   b) Correction of severe airway obstruction by Le Fort III-osteotomy, especially in patients which had undergone tracheotomy during their first months of life. However, Le Fort III-procedure performed during this age period involves the risk of damaging the tooth germs.

3) Surgical procedures between the fourth and tenth year of life (Fig. **1b**):
   a) Frontofacial advancement (group of Crouzon's disease and Apert's syndrome). This may be done in one block if the amount of movements at all levels is similar. However, if the skeletal deficiency of the levels differ, then individual osteotomies for their individual corrections are necessary by division between the levels I, II and III. Osteotomy between the levels III and IV (Le Fort I-osteotomy) are contraindicated because of injury to the dental anlage.
   b) Correction of the levels I, II, and III combined with moving the orbits in the vertical (Fig. **3**) and/or horizontal planes in cases of hypertelorism (Fig. **1b**). In this age group the germs of the canines are usually, but not always, positioned caudally enough to allow a high infraorbital osteotomy. Careful radiological investigation will indicate the exact position of the germs of the permanent teeth.
   Concomittant brachyorbitism and hypertelorism is corrected simultaneously (19) by elongating and medially rotating the orbits. The olfactory nerve filaments are preserved using our method (20).
   c) A genioplasty using Obwegeser's sliding osteotomy (10) is only possible if the position of the germ of the canine has developed far enough in the cranial direction to allow the osteotomy of the chin prominence. In cases with caudally positioned tooth germs, an onlay chin augmentation is performed using homologous lyophilized cartilage grafts (15).
   d) In orthodontic patients (6 years or older) treatment using fixed bonding is sometimes indicated to achieve acceptable postoperative occlusions. Early relapses of the midface position are avoided by always paying special attention to the occlusion.

4) Surgical procedures after the tenth year of life (Fig. **1c**):
   a) Osteotomies are usually possible between all levels I, II, III, IV including the combined Le Fort III plus Le Fort I-osteotomy (Figs. **4–5**) (11). The Le Fort III-osteotomy minus Le Fort I-osteotomy (Fig. **6**) is indicated in patients with normal occlusion (Fig. **6**) or in cases in which the upper (infraorbital) part of the midface must be advanced further than the lower tooth-bearing part of the maxilla. A typical example is in the case of the "top Crouzon"-patient or at times with the Saetre-Chotzen-Syndrome. Careful radiological examination is necessary before Le Fort I-osteotomies because the positions of the canines must be caudally enough to allow the horizontal osteotomy.
   b) Mandibular movements (Fig. **1c**; level 5) to correct retromandibulism (class II) are possible from the age of 10 years onwards. Mandibular movements (level 5) to correct prognathism (class III) are not usually per-

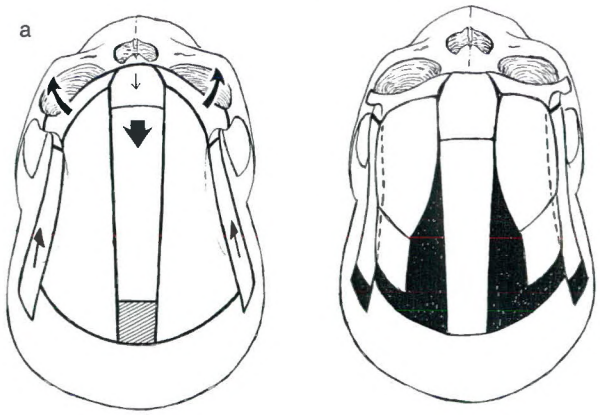
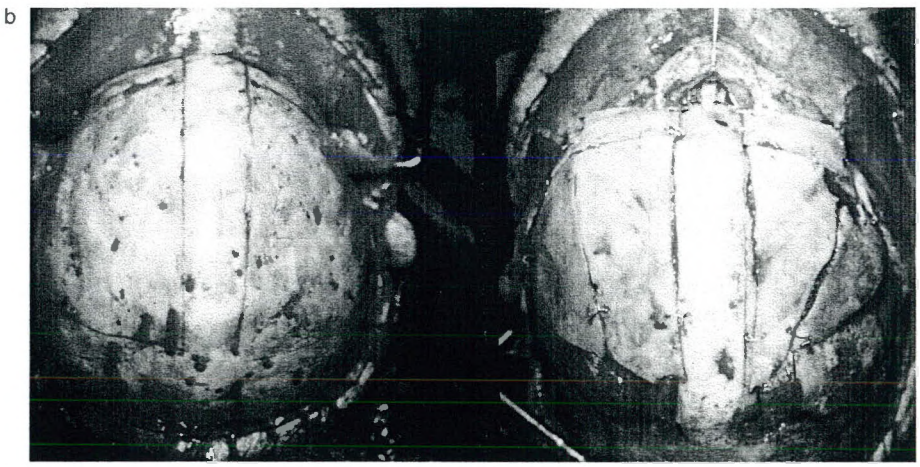


Fig. 2 Method for correction of scaphocephalus. The central part of the forehead is retropositioned, both lateral parts are advanced, left the osteotomies; right the condition after correction **(a)**. The intraoperative views **(b)** correspond in principle to the drawing **(a)** detailing the skull enlargement.



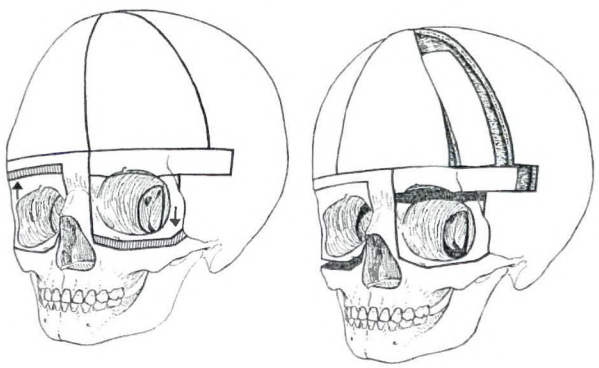


Fig. 3 Plagiocephaly with orbital dystopia; the drawing details the surgical procedure: elevation of the right and lowering of the left orbit; the infraorbital osteotomy traversed the left and right infraorbital foramina because of high positioned canines. The infraorbital nerves were preserved. Simultaneously the reseated left supraorbital rim and left forehead region were advanced (right).

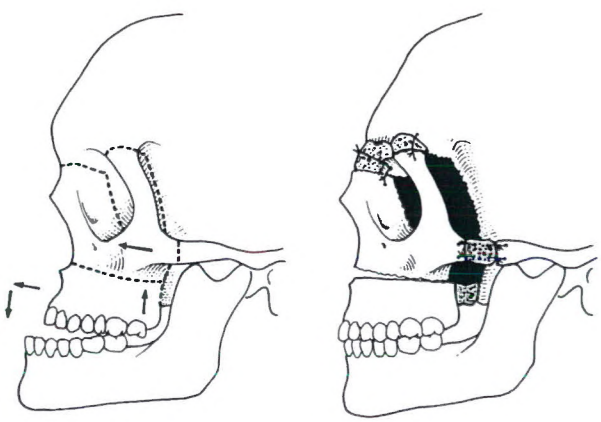


Fig. 4 Combined Le Fort III- and Le Fort I-osteotomy: This procedure allows the individual correction of the tooth-bearing and the infraorbital part of the midface. Correction of the open-bite deformity is possible without causing an extreme prominence of the glabella region. Simultaneously elongation or shortening of the midface and narrowing and widening of the maxillary arch are possible (18).

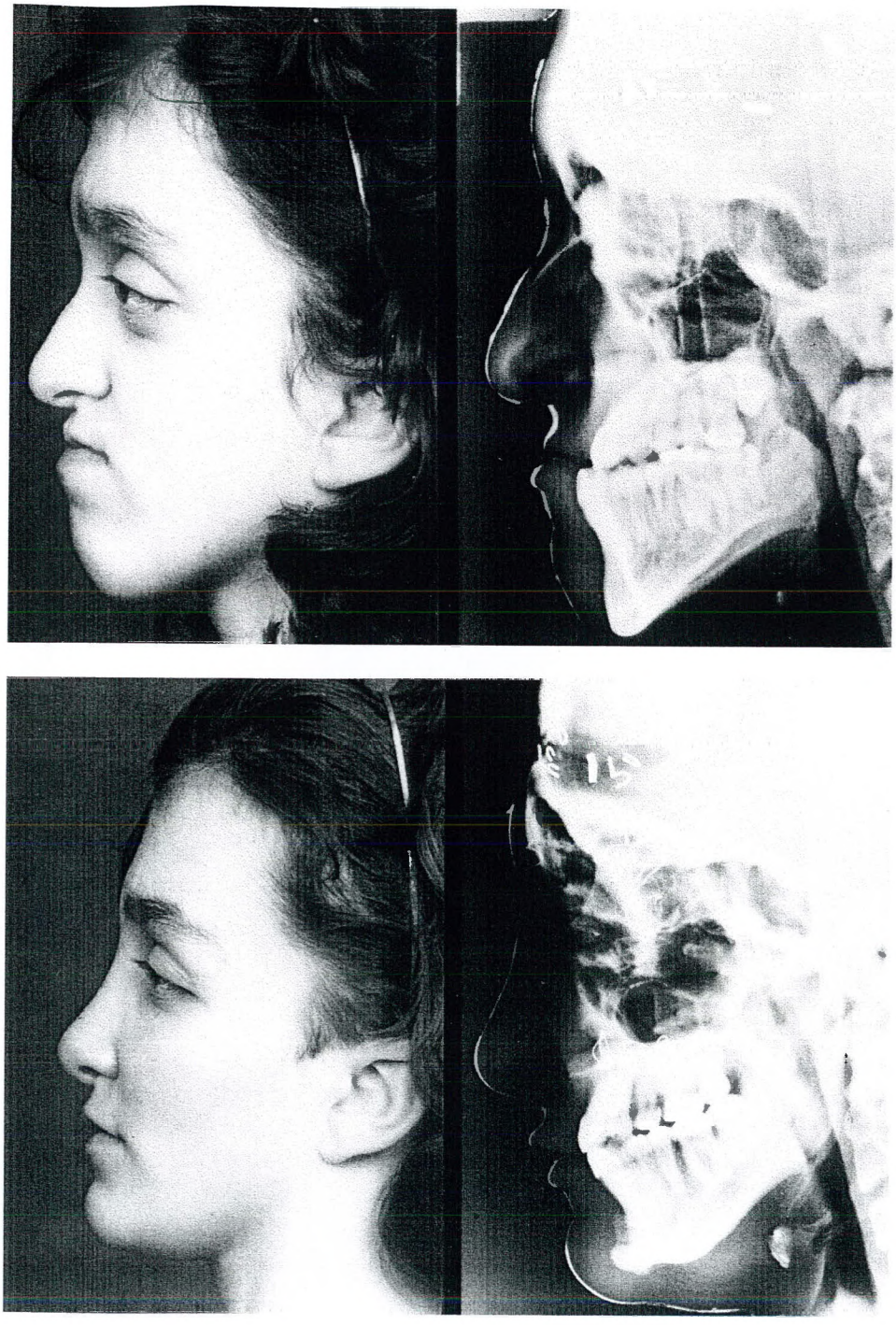


Fig. 5 The preoperative **(a)** and postoperative **(b)** photographs and cephalometric radiographs of the 13 year old girl with Crouzon's disease detail the method: the infraorbital rim was advanced further than that of the tooth-bearing part of the maxilla, and a sliding genioplasty (10) was done at the same time.

formed before the patient is 14 years old. The sagittal splitting procedure (9) is performed, and the germ of the third molar can be preserved or removed at the same time.

Principles in Treating Craniofacial Stenosis

1. Maximal correction in every age group:
   The maximal correction possible in each age group must be achieved. Two-stage procedures should be avoided when simultaneous corrections of several levels are possible.
2. Eye-glass-osteotomy (Fig. 7):
   This osteotomy was designed to treat cases by frontofacial advancement in several segments. This osteotomy

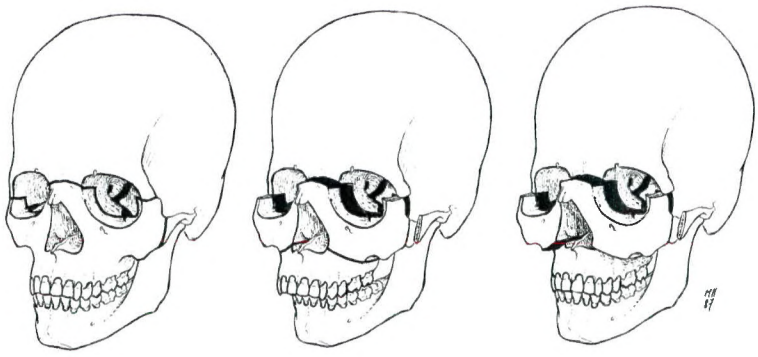


Fig. 6 Le Fort III- minus Le Fort I-osteotomy: The drawing shows that first the midface is mobilized as in a routine Le Fort III-procedure; not before mobilisation is perfect the Le Fort I-osteotomy is performed and the tooth-bearing part of the maxilla retropositioned and put into occlusion.

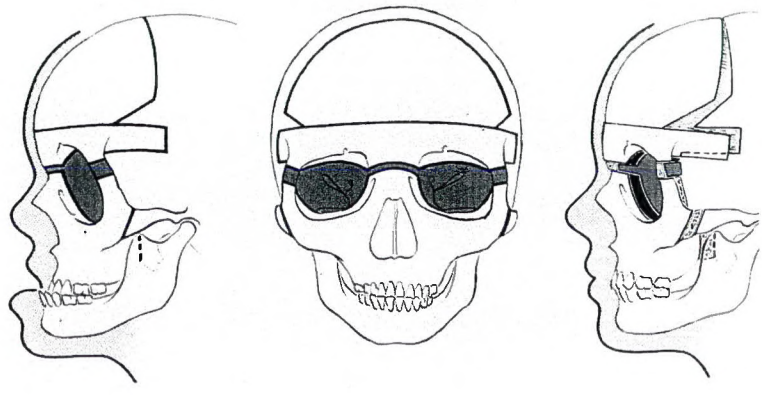


Fig. 7 Eye-glass osteotomy: This kind of osteotomy was designed for difficult cases of frontofacial advancement with several segments. The osteotomy leaves a horizontal stable skeletal structure between the cranial and midface segments. The drawing (centre) simulates eyeglasses; the lateral orbital parts function as fixation points for the segments and transplants serving as reference points for orientation and measurements. At the end of the operation, the gap between the supraorbital and lateral orbital rims is bridged using a graft of lyophilized cartilage. The midface is already advanced, and the gap within the zygomatic arch was bridged by bank bone. Craniotomy and frontal advancement will now follow.

has been used for 6 years and is helpful to orientate, measure, and fix segments and transplants.

3. The intraoperative timing in frontofacial advancement:
   In severe cases of frontofacial advancement in several parts it is imperative first to mobilize the midface. This is especially important in infants and children. If the craniotomy is performed first it is almost impossible to fix the small skulls of children when the midface must be mobilized.
4. Avoid autologous grafts and use allogeneic ones instead!
   We use allogeneic transplants to reconstruct all defects in the cranial vault area, the orbit, the skull base, the zygomatic complex, and the maxilla.
   The most reliable are:
   a) Gas-sterilized, lyophilized allogeneic cartilage which has been used in over 4000 cases involving all areas of the craniomaxillofacial region. Lyophilized cartilage is transformed into the patients own bone (17).
   b) Gas-sterilized, lyophilized allogeneic ribs, iliac or calvarial bone. Deep-frozen allogeneic bone should *not* be used, particularly without other additional sterilization procedures. The risk of transmission of infectious diseases must be totally avoided. Autologous transplants of rib and iliac crest bone are now rarely used in our department.
   Autologous bone obtained by splitting of the calvarium is used less and less. This is because it is time-consuming and the results are not better than for reconstructions of defects obtained after craniofacial movements than lyophilized cartilage or bone.
5. The empty-space problem in craniofacial surgery:
   The epidural cavity created by advancing the frontal bone in adults can remain because of lack of cerebral expansion. The empty space can be obliterated by using lyophilized cartilage to avoid infections. The same holds for the empty-space management in the temporal region after hypertelorism procedures.
6. Use of metal plates in craniofacial surgery:
   Metal plates in the non-hair-bearing forehead area should be abandoned since they are visible under the skin after several months and often cause reactive bone production. Metal plates in the forehead area have not been used in Zürich since 1983. However, in the hair-bearing region of the skull the plates can be helpful but rarely remain the procedure of choice.
7. Sagittal overcorrection:
   A sagittal overcorrection of the tooth-bearing midface region is done in children younger than twelve years of age operated by a Le Fort III-osteotomy. This results in a class II-occlusion, and the cases are not stable because of insufficient intercuspidation. Special dental splints and rubber bands must often be used over many months to maintain the position of the midface.
8. Skin incisions:
   Only coronal skin and enoral incisions are employed. Transconjunctival and other approaches to the orbit are not necessary.
9. Sequence of osteotomies in cases of frontofacial advancement:
   In patients with combined mobilization of the calvarium, frontal bone, and midface, first mobilization

and fixation of the midface (Le Fort III-osteotomy) is performed. If the craniotomy is done first the head cannot be fixed well enough manually during mobilization of the midface.

10. Sequence of osteotomies in cases of Le Fort III- plus/minus Le Fort I-osteotomies:
The midface as a whole is first mobilized in all cases with combined Le Fort III- plus Le Fort I- or Le Fort III-minus Le Fort I-osteotomies. The Le Fort I-osteotomy is subsequently done either to further advancement (Le Fort III plus Le Fort I) or to backward movement (Fig. 23.**6**) of the tooth-bearing part of the maxilla (Le Fort III minus Le Fort I).
11. Cerebral spinal fluid-drainage:
Cerebral spinal fluid-drainage systems are ligated temporarily in patients with craniosynostonis to avoid a ventricular collapse during craniofacial surgery.
12. Intermaxillary fixation (IMF):
Individual splints (8) can be fixed to either the deciduous or the permanent teeth. Intermaxillary fixation longer than 6 weeks is not practised. Using rigid fixation by plating IMF can sometimes be avoided.
13. Interskeletal suspensions (4) in children are done with wires fixed around the mandible or through the bone at the most inferior border of the mandible to avoid injuring the tooth germs.

Discussion

There is no doubt that the growing craniofacial skeleton reacts differently to osteotomies performed in the different craniofacial levels described above (Fig. **1**).

In most cases corrections of the cranial vault using the floating forehead (6) or the floating occiput procedure (5) predispose to normal skull growth. In some cases the floating occiput procedure can also improve the shape of the deformed cranial base.

Osteotomies in the growing midface (Le Fort III-osteotomies) do not lead to a release of the inhibited growth in every case. An acceptable intermaxillary relationship was achieved in most cases published, but several showed a skeletal class III-condition which was just corrected by orthodontic appliances. A normal occlusion, therefore, does not necessarily prove a continuing normal growth of the midface. In many cases of early midface advancement a Le Fort I-osteotomy is necessary after proper physiognomy planning and orthodontic treatment.

Growth of both the mandible and the maxilla is influenced by the development and eruption of the tooth germs. Therefore, surgical interventions of the midface (Le Fort III-osteotomies performed during the first year of life (7) and Le Fort I-osteotomies in early childhood do traumatize the tooth-germs. This produces growth disturbance of the skeletal structures whose growth potential is already impaired by the underlying malformation. Craniofacial surgery can no longer ignore this important fact. The timing of craniofacial surgery must consider other biological aspects as well including knowledge of tooth development as it is established in the field of orthopedic jaw surgery.

In early childhood the anlage of the permanent teeth and tooth germs is not visible on radiographs because of lack of calcification. The anlage of the permanent maxillary dentition is closely related to the orbital floor and the outer parts of the retrolateral walls of the maxilla. Therefore, advancement of the midface in the first months of life is indicated only in patients with malignant exophthalmus.

Children with respiratory obstruction, e.g. severe forms of Crouzon's and Apert's disease, are usually high-risk patients. Respiratory insufficiency is not only caused by the severe hypoplasia and retropositioning of the midface but also by anomalies of the tracheal system. These young patients are also in a poor general medical condition and early tracheotomy is recommended. The children gain weight and their general condition improves. Simultaneously an excellent presurgical examination is also facilitated. The surgical risk is reduced if these patients are about one year old. Further, the size of the midface skeleton is conducive to a lowered surgical risk, and the mobilization and fixation of the midface is easier to achieve.

Correction of the hypertelorism depends on the position of the tooth germs in children. These procedures are usually delayed until the germs of the permanent teeth are calcified and visible on the radiographs. The position of the canine (Fig. **1b**) can remain high till the age of 10 years and infraorbital horizontal osteotomy is, therefore, risky in such patients.

Cases of brachyorbitism combined with hypertelorism can be corrected simultaneously using the method discussed by Sailer and Landolt (19). In this technique olfactory nerve filaments can always be preserved (20) eliminating this draw-back associated with of the original hypertelorism operation (25).

A sliding genioplasty (10) affects the physiognomy favorably in many patients. This method can be used as soon as the tooth germs of the mandibular canines leave sufficient space for the osteotomy of the chin prominence and is usually possible after the age of 10 years. However, chin augmentation can also successfully carried out before this age using lyophilized cartilage onlay grafts (17).

Mandibular osteotomies should be correlated with the growth period as well. In case of craniosynostosis, mandibular growth is usually not primarily but often secondarily impaired. The age of the patient must be considered when retropositioning the mandible. The risk of relapse seems lower in females because of their earlier growth termination. The risk of osteotomies relapsing increases when the procedure is timed too soon after the onset of puberty (2).

A different situation exists in patients with impaired mandibular growth as in the Treacher-Collins syndrome and other forms of retromandibulism. In these patients mandibular advancement is usually possible at the age of 10 years because important mandibular forward growth cannot be expected anymore. The sagittal splitting procedure (9) is then performed.

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Facial Splitting for Correction of Orbital Hypertelorism

Introduction

The correction of orbital hypertelorism in adults can often be well accomplished according to the original Tessier osteotomies with infraorbital cuts horizontally through the maxilla (5). In children, however, these osteotomies are likely to interfere with the development of the permanent tooth buds, especially the canines, and should not be used until the age of 13 or 14 years.

The "keel deformity" of the hard palate and midline alveolar process together with marked orbital hypertelorism as described by Tessier (4) appears to be a developmental arrest of the face. It has consequently tempted several craniofacial teams to complete the inward rotation of the facial halves after splitting the face sagittally (1, 3, 6).

The technique of facial splitting involves a sagittal osteotomy through the hard palate between the central incisors with inward rotation of the facial halves and circumvents the problems of horizontal osteotomies through the maxilla endangering the tooth buds.

For several years we have felt that in children with orbital hypertelorism a pronounced "keel deformity" of the dental arch would have to be a prerequisite for a facial splitting operation. Urged by the need to correct these children early we have started to do facial splits regardless of the dental arches. We assume that the distortion of the dental arch that may result after surgery in fact can be treated orthodontically later on, if self-correction proves to be insufficient.

The purpose of this paper is to testify to the usefulness of the facial splitting procedure for hypertelorism correction, but also to question its use in certain situations.

Material and Surgical Techniques

During a 12-month period 9 hypertelorism patients were operated on, 8 of these with the facial splitting technique. Their data are shown in Table **1**.

Preoperative analysis: All subjects underwent CT scanning with a section thickness of 1.5 mm (GE 800 CT scanner). The magnetic tapes were then analyzed with a 3-D imaging program (U.S.). In addition, standard cephalometric records were taken.

Facial split. Surgical procedure: General anesthesia is maintained with hypotension monitored through an arterial line. The tracheal tube is placed in the mouth and fixed in a manner that permits visual inspection of the hard palate.

No hair is cut or shaved at all for the bicoronal approach which is carried out freeing up the temporal muscles completely for subsequent transpositioning at the end of the procedure.

The orbits are dissected in a standard manner with the important difference that the medial orbital wall is only dissected behind the lacrimal sack and the medial palpebral tendon. In fact, these two structures should not be visualized at all and the soft tissues in this area are as much as possible left attached to the underlying bone.

After planning and drawing with ink, a semilunar "window" is made in the frontal bone and taken out. Caudal to this a horizontal bar, ca 15 mm wide, is removed to facilitate access to the anterior cerebral fossa (Fig. 1). The bar will be wired back in place later. The anterior fossa is dissected and the frontal bone between the orbits is removed down to the glabella with a saw. The nose is entered with a burr making it possible to save the nasal mucosa and dissect it free before making the osteotomies.

The orbital roof, floor and lateral wall are cut with a saw in the ordinary way. The medial wall, however, which is less dissected, leaves relatively little room and the blade of the reciprocating saw is therefore inserted from the inside of the anterior fossa, joining the osteotomies of the floor and the roof moving the saw in one cut medially behind the lacrimal sack and all the way into the nose and