27 Surgical Treatment of Hypertelorism

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DEFINITION

Orbital hypertelorism, or hypertelorbitism, is an increased distance between both orbits. The distance between the medial and lateral orbital walls is also increased. The normal interorbital distance (IOD) of an adult individual is approximately 25 mm. Tessier classified three types of hypertelorism in adults: In type I, the IOD is between 30 and 34 mm; in type II, between 35 and 39 mm; and in type III, wider than 39 mm.[1] In infants and children, the IOD is much smaller and age dependent. An IOD of 25 mm in a 1-year-old child is considered hyperteloric and must be closely observed, and the etiology must be investigated. The IOD (Fig. 27–1) may be measured with a caliper, the arms of which are pressed upon the medial orbital rims. Accurate measurements can be made by posteroanterior cephalometric radiograph and by computed tomography (Fig. 27–2). The intercanthal soft tissue distance (ICD) is measured with a ruler. In an adult, the normal ICD is approximately 30 mm; in a 3-year-old child, approximately 26 mm.[2,3] There is no direct correlation between the IOD and the ICD. Often both distances necessitate independent corrections. Hypertelorism is practically always combined with a malformation of the nose, ranging from wide nasal bones over a bifid nose to complete or partial nasal duplication and also hypoplasia or aplasia of parts of the nasal hard and soft tissue structures.

FORMS OF HYPERTELORISM

Hypertelorism is either symmetric or asymmetric (Fig. 27–3). In addition, the orbits can have an "antimongoloid" position, or they can have a normal shape and volume but can also be shallow, as in Crouzon and Apert syndromes (Fig. 27–4). The four orbital rims can be situated symmetrically in the three planes, or they may be individually malpositioned. These rims must be correctly positioned during the operation. There are also unilateral forms of hypertelorism (Fig. 27–5; see also Fig. 27–11*A*) as well as cases of hypertelorism with eyeless orbits (Fig. 27–6).

ETIOLOGY

Hypertelorism is not a pathologic entity. It is usually a symptom and can be caused by hyperpneumatization of the ethmoids, craniostenosis of the anterior skull base, persistence of the metopic suture, syndromes such as Crouzon or Apert, medial facial clefts, or meningoencephaloceles (Fig. 27–7; see also Fig. 27–24). Many patients with cleft lip and palate present with mild hypertelorism (Fig. 27–8). Tumors situated between the orbits (osseous keloids, fibrous dysplasia, neurofibromastosis), mucoceles of the frontal and ethmoidal sinuses, and massive facial trauma may induce hypertelorism (Fig. 27–9): only the medial orbital walls show an increased distance from each other, but not the lateral walls. A condition like this is defined as pseudohypertelorism, and correction is often possible by subcranial procedures (to be described).

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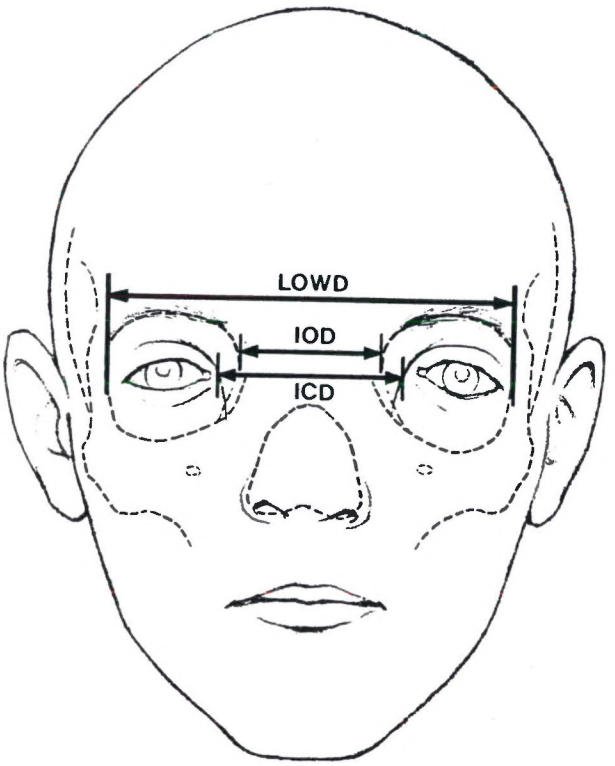


Figure 27–1. The interorbital distance (IOD) is difficult to determine because of the overlying soft tissues; the intercanthal distance (ICD), which is very important for esthetic reasons, is easily measured by a ruler. In true hypertelorism, the distance of the lateral orbital walls (LOWD) is also increased, in contrast to cases of pseudohypertelorism.

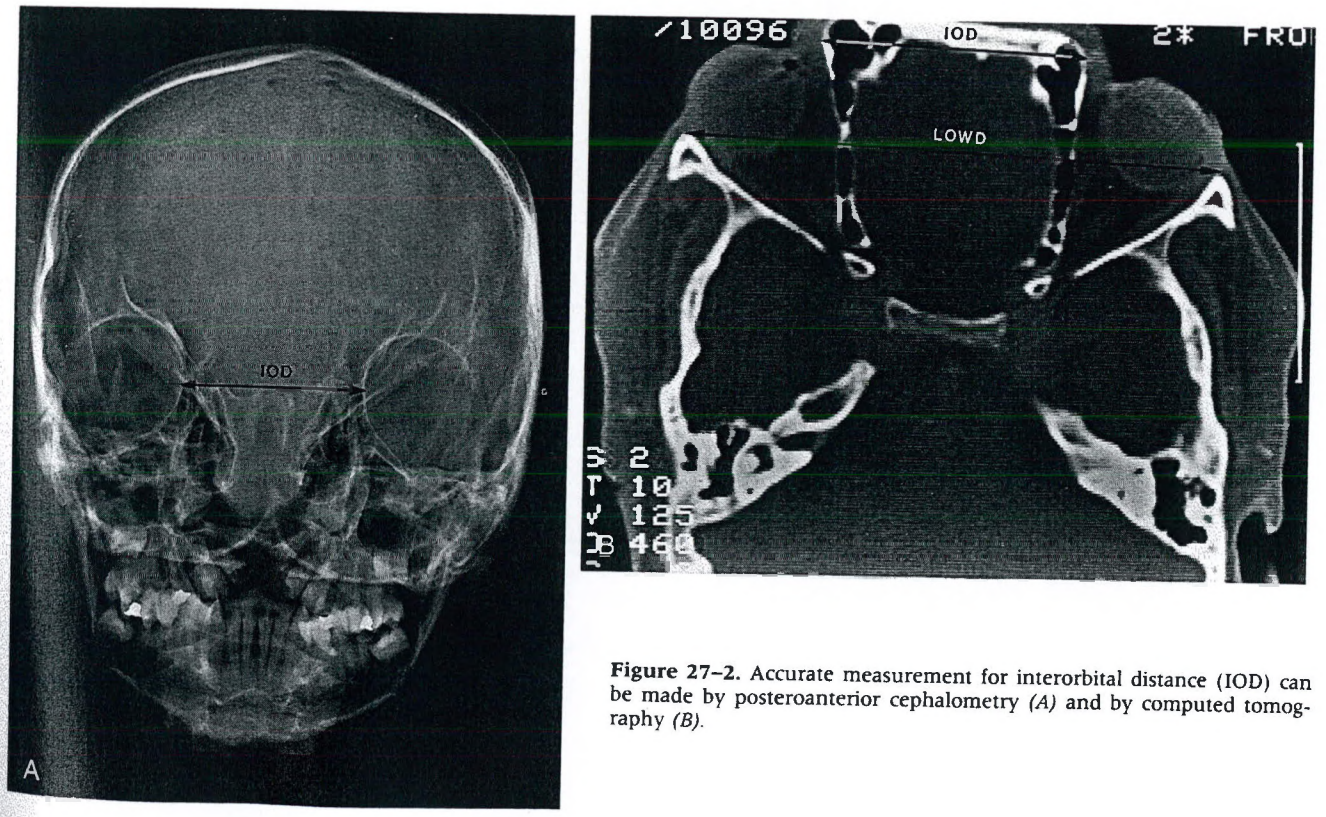


Figure 27–2. Accurate measurement for interorbital distance (IOD) can be made by posteroanterior cephalometry *(A)* and by computed tomography *(B)*.

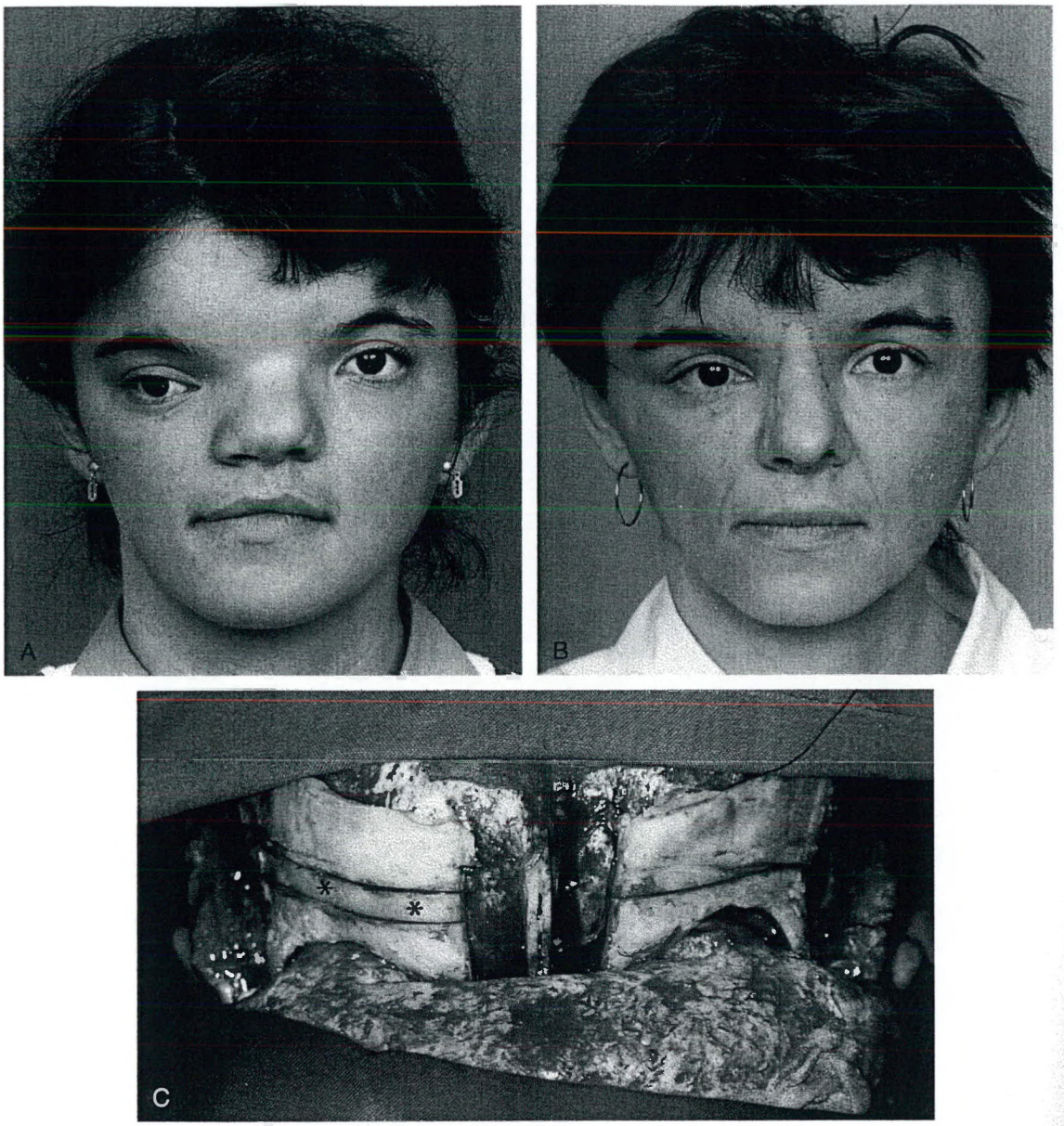


Figure 27–3. Case of asymmetric hypertelorism before *(A)* and after *(B)* correction. The intraoperative picture *(C)* shows the condition after interorbital osteotomy and before removal of the supraorbital horizontal bone band *(asterisk)* to elevate the right orbit.

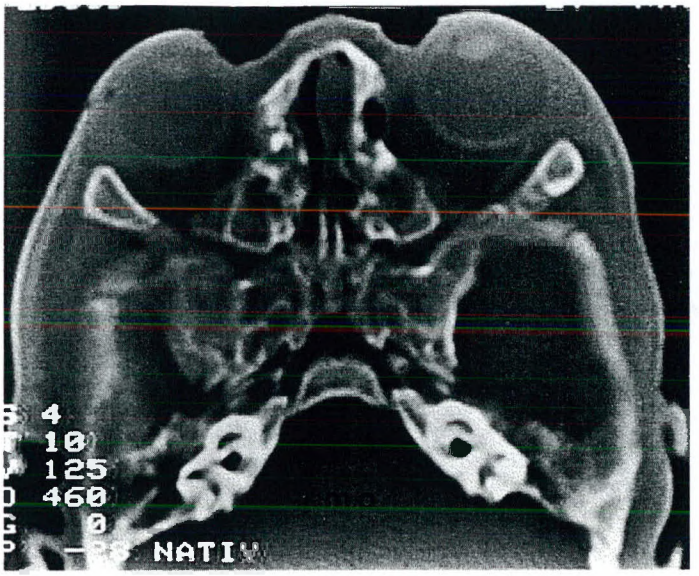


Figure 27–4. Extremely shallow orbits in a case of Crouzon syndrome with medium degree of hypertelorism.

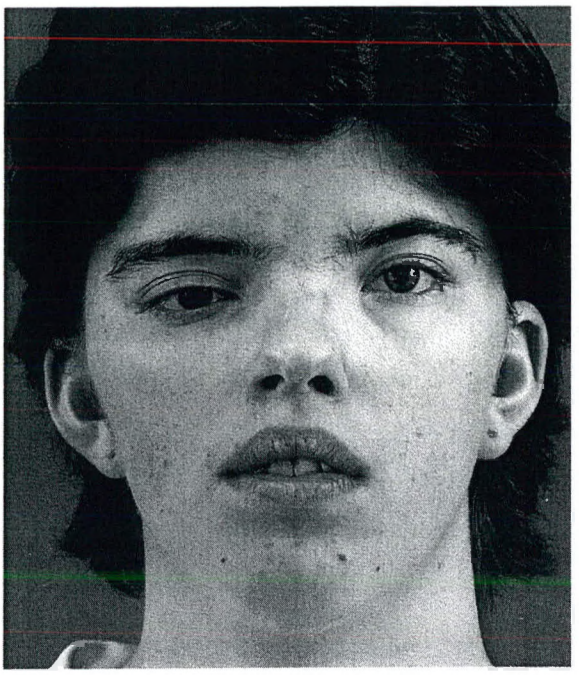


Figure 27–5. Unilateral form of hypertelorism (see also Fig. 27–11*A*) in a case of plagiocephaly.



Figure 27–6. Monstrous malformation, including extremely hyperteloric, eyeless orbits, several kinds of facial clefts, and interorbital encephalocele.



Figure 27–7. *A,* Meningoencephalocele causing hypertelorism; *B,* the intraoperative picture shows the perforation of the encephalocele (already resected) into the left orbit *(arrows).*

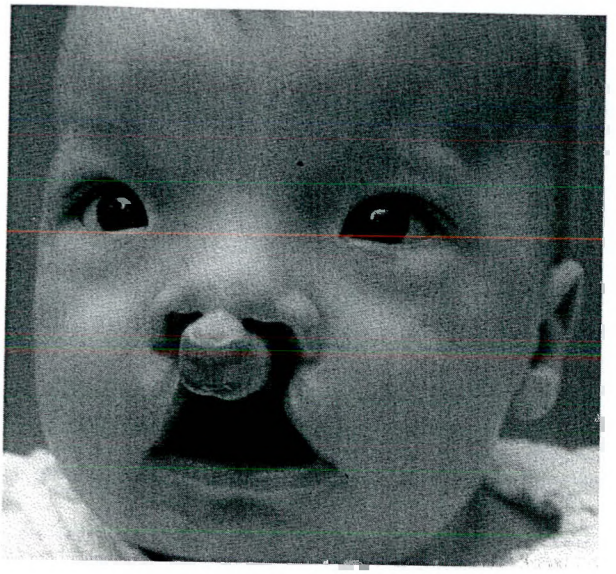


Figure 27–8. Bilateral cleft lip, cleft palate, and orbital hypertelorism.

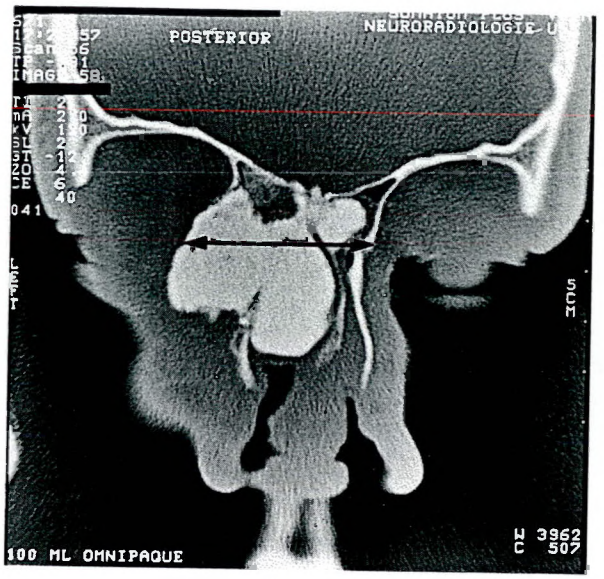


Figure 27–9. Interorbital tumor-like lesion (osseous keloid) causing widening of the interorbital distance (IOD) but not of the lateral walls.

TIME OF SURGERY

The development of the deciduous and permanent dentition is the most important determinant of the timing of surgery.[4] As long as the maxillary sinus has not yet developed and the tooth germs have not yet grown caudally enough, the infraorbital horizontal osteotomy cannot be performed without damage to the tooth buds (Fig. 27–10). Therefore, the earliest age for hypertelorism correction is between 4 and 10 years, when the maxillary sinus is developing.

The development of the roots of the canines always merits special consideration. Often their apices are positioned far cranially in adolescents and even in adults because of missing space within the alveolar arch. It is imperative that the canines be preserved. Therefore, orthodontic treatment is often required after extraction of the first premolars in order to guide the eruption of the canines.

An exception to this is the facial bipartition procedure, in which no infraorbital horizontal osteotomy is performed. In this procedure there is less risk to the canines and premolars, but the molars may nonetheless be damaged, depending on the age of the patient.

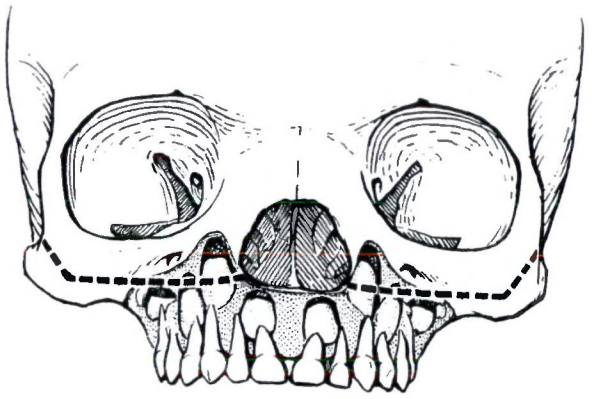


Figure 27–10. The drawing shows the state of tooth development in 4- to 5-year-old children and the infraorbital horizontal osteotomy that causes damage to the permanent dentition.

Craniomaxillofacial surgeons should always be aware that damage to the tooth buds results in growth impairment of the midface.[4] According to Enlow, the growth of the primary and the secondary dentition is also responsible for the development of the maxilla in all three dimensions.[5]

In some of the syndromes associated with hypertelorism (e.g., Crouzon and Apert syndromes), growth impairment of the midface is typical of the malformation; this is aggravated if tooth buds are damaged by early surgery as it is recommended by others.[6,7]

PLANNING

The surgical correction of hypertelorism is difficult and risky because of the highly important contiguous anatomic structures. The diagnostic and planning aspect of treatment are multidisciplinary tasks, but the surgeon should be the leader of the team because he or she is responsible for the result and for possible complications.

For planning, all modern diagnostic means should be available, including three-dimensional imaging and stereolithography. The most important rule gained by experience is that intercanthal distance reduction corresponds to only half of the amount of the removed bone; for example, interorbital bone resection of 30 mm results in a reduction of the ICD of about 15 mm. As mentioned, exact measurements in the posteroanterior cephalometric radiograph or in the computed tomographic scan (see Fig. 27–2) are necessary for preoperative assessment. Life-size photographs of the patient's face are beneficial (Fig. 27–11). Stereolithographic models allow the exact simulation of the surgical procedure (Fig. 27–12).

Every case of hypertelorism correction demonstrates profile alterations of various degrees (see Fig. 27–11*E*,*F*), necessitating simultaneous corrections of nasal form and of the projection of the zygomatic area as well as of the chin.

POSSIBILITIES IN SURGICAL CORRECTION

In the authors' department, correction of as much as possible in a single operation is the goal. The simultaneous transcranial and transoral approach is used in all hypertelorism corrections, and a higher incidence of infection has not been observed.

The correction of hypertelorism always involves hard and soft tissue procedures. Not only are the bony structures of the orbit, the maxilla, the nose, and the forehead involved; the soft tissues of the forehead, the nose, the cheek, the upper lip, the eyelids, and the medial and lateral canthi must also be restructured. In transcranial procedures, the cribriform plate is exposed and dissected, and consequently the olfactory nerve system may be damaged. The whole midface is degloved, including the zygomatic bone and arch, and the temporal muscles are elevated. The surgeon must therefore have experience in handling soft and hard tissue. He or she should also be accustomed to contemporary means of fixation such as micro- and miniplates in the treatment of maxillofacial trauma.

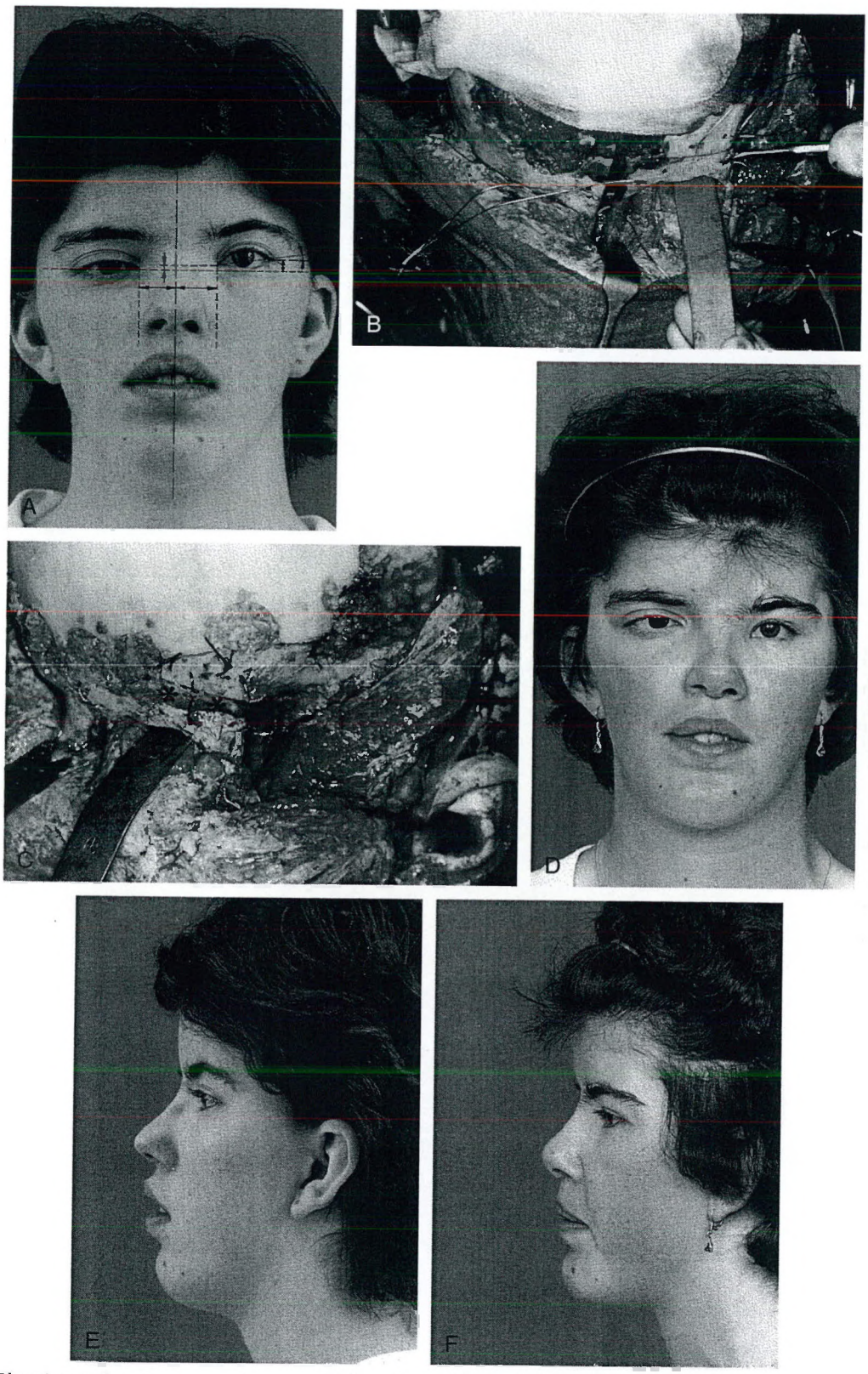


Figure 27–11. Planning and performance of a case of left unilateral hypertelorism with vertical orbital discrepancy. Exact measurements on the 1:1 life-size photograph *(A)* suggest removal of interorbital nasofrontal bone on the left, as seen in the intraoperative picture *(B)*, by lowering the left orbit *(C)* by interpositional bone graft *(asterisk)*. The result *(D)* shows both orbits on a horizontal level, but eyelid soft tissue differences remain. Simultaneous correction of the facial profile *(E*, before surgery; *F*, after surgery) is necessary.

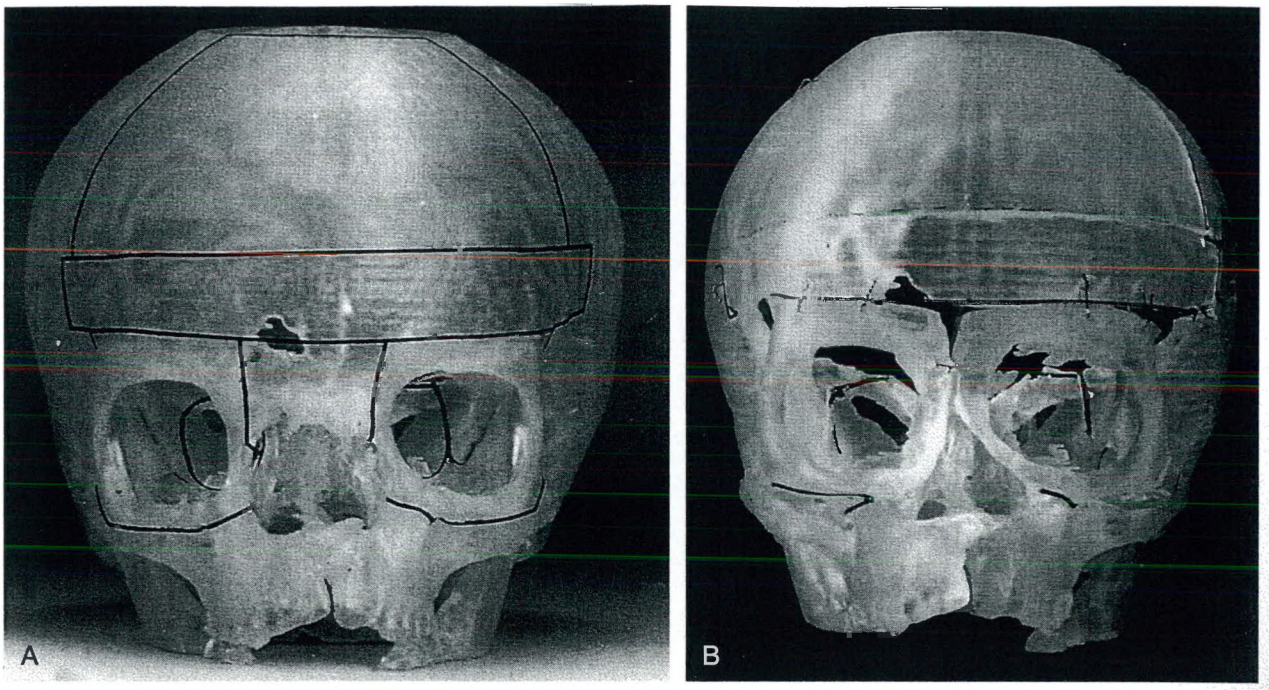


Figure 27–12. Stereolithographic models allow an exact analysis of the morphology of the malformation *(A)* and perfect simulation of the operation *(B)*.

Correction of hypertelorism can be accomplished subcranially or transcranially. There are many variations and modifications. The most common procedures used in the Department of Cranio-Maxillofacial Surgery, University Hospital Zurich, are presented and described step by step.

Subcranial Procedures

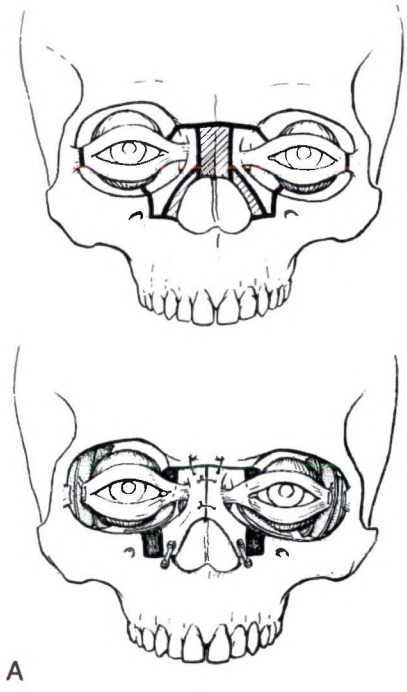
Subcranial correction of hypertelorism or pseudohypertelorism is indicated in mild forms only when the IOD is about 35 to 40 mm. This operation is indicated especially in posttraumatic cases or for tumors in which the lateral orbital walls are in a normal position. The procedure involves medial orbital wall approximation, including the nasal and lacrimal bones along with the medial canthi.

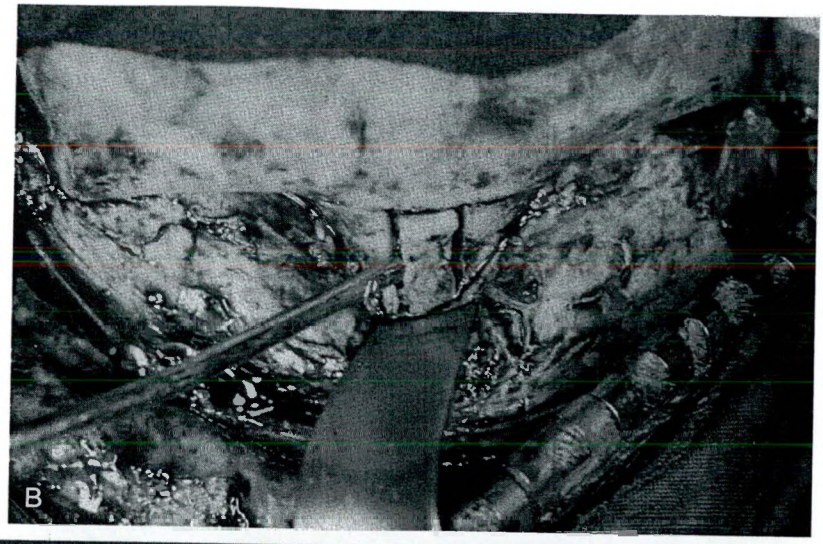
Another subcranial procedure involves orbital medialization without orbital roof osteotomy. In this procedure, both the medial and the lateral orbital walls and the orbital floor are shifted medially. Both subcranial operations can be simultaneously combined with a LeFort III osteotomy, if necessary.

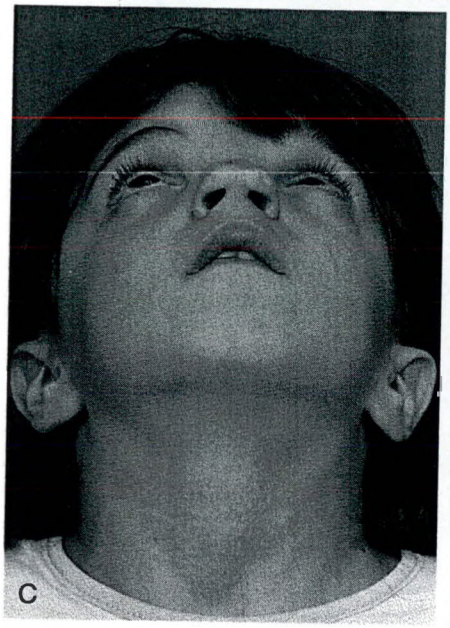
The Medial Orbital Wall Approximation (Fig. 27–13*A,B,C,D*)

Step-by-Step Procedure

1. For the coronal incision, a 1.5-cm–wide strip of hair running from one ear to the other is removed; the remaining hair is disinfected, and the posterior part of the head is covered with a sterile cloth fixed to the galea by clamps. The anterior part of the hair-bearing scalp is covered with a cloth that is fixed by stitches, leaving the forehead free (Fig. 27–14). A local anesthetic containing epinephrine is injected, and an incision made along the hair-free strip of galea running from one ear to the other and, from there, forward and downward close to the helix. The wound edges are slightly mobilized on both sides. Hemostatic clips are applied along the posterior wound edge, and a hemostatic continuous running suture along the anterior wound edge is placed. Then a supraperiosteal dissection is performed, together with the dissection between the superficial and deep temporal fascia. The fat pad between both fasciae is dissected by knife to avoid damage to the facial nerve. About 2 cm above the supraorbital rims,







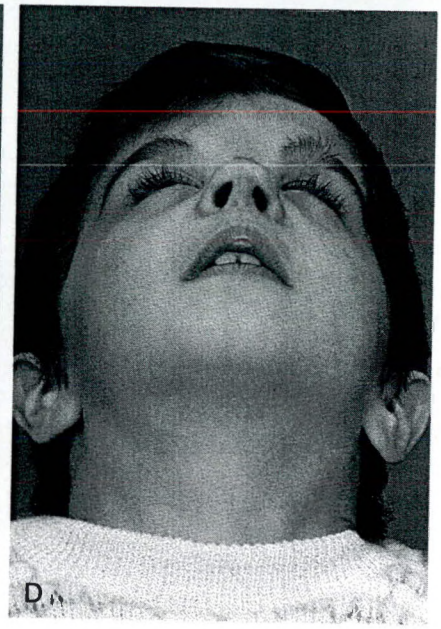


Figure 27–13. *A,* Schematic drawing of the medial orbital wall approximation as it is often used in cases of pseudohypertelorism. The osteotomy along the nasofrontal suture must be performed in a caudoposterior direction to avoid intracranial penetration. Along the pyriform aperture, some bone is removed to avoid nasal obstruction. The slight enlargement of the orbital volume is compensated by transplantation of bone or cartilage grafts to the lateral walls. The lateral canthal ligaments should also be repositioned. Orbital wall defects are bridged by grafts. *B,* The intraoperative photograph shows the amount of dissection and the osteotomies for interorbital nasal bone resection; patient with slight hypertelorism before *(C)* and after *(D)* medial orbital wall approximation and nasal correction.

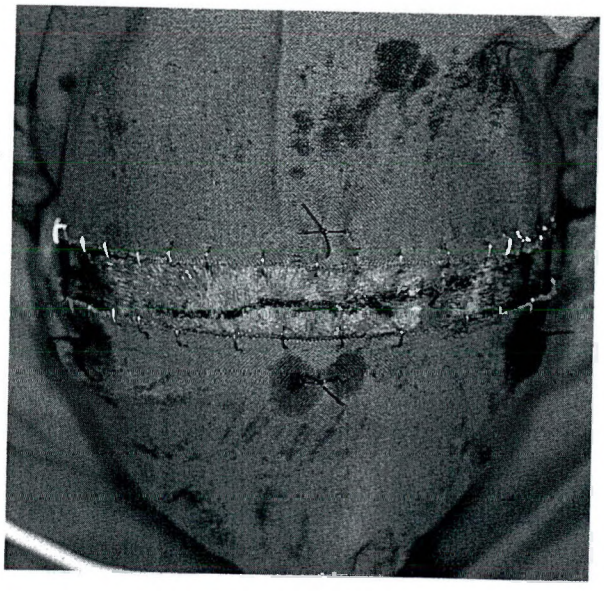


Figure 27–14. Preparation of the scalp. Only a 1.5-cm–wide strip of hair is removed, and the remaining hair anteriorly and posteriorly is disinfected and covered with a sterile cloth.

the pericranium is incised horizontally, and the bone around the supraorbital rims, the nasal bones, and the glabella are degloved. In most cases, the supraorbital neurovascular bundle must be removed from its canal on both sides.

2. The nasal bones, parts of the cartilaginous structures of the nose, the orbital roofs, and the medial, lateral, and inferior walls of the orbits are subperiosteally exposed, and the medial canthal ligaments (including the lacrimal duct) are dissected but not detached.

3. A horizontal bone cut is made along the nasofrontal suture running into the medial orbital wall on both sides and then downward behind the canthal ligaments and the lacrimal duct system, in the direction of the infraorbital rim. Two vertical cuts are then made through the nasal bones in front of the medial canthal ligaments. The distance between the cuts is coincident with the desired medial wall movement. A Toller bur is used for this osteotomy and helps to avoid damage to the underlying nasal mucosa. The interorbital nasal bone piece is now carefully removed without damaging the mucosa.

4. For the transoral osteotomies, a horizontal incision is made in the maxillary vestibule, and the canine fossa, including the infraorbital rims and the infraorbital foramen, is degloved. The nasal mucosa is elevated inside the pyriform aperture on both sides. An osteotomy beginning laterally is now performed through the infraorbital rim medially to the infraorbital foramen (but far enough laterally not to damage the lacrimal duct), running downward into the pyriform aperture beneath the inferior turbinate. With the use of chisels, the bony structures containing the medial canthal ligaments and the lacrimal ducts are mobilized medially and fixed into position with direct wiring (see Fig. 27–13). There is no absolute indication for the use of plates and screws in such a procedure.

5. The medial orbital wall approximation causes enlargement of the orbital volume, which necessitates compensation. Many surgeons use autologous rib grafts or split calvarial grafts placed onto the lateral orbital walls. The authors' material of choice is lyophilized cartilage.

6. If necessary, a strut of lyophilized cartilage can be placed to project the nasal dorsum. Other minor corrections are performed with the same biomaterial. In an alternative procedure, it is possible to detach the medial canthal ligaments to perform the osteotomies and replace the ligaments in the old or a new position. But usually the ligaments are left in place.

The Orbital Medialization Without Roof Osteotomy (Fig. 27–15)

This procedure is performed if a more extensive medial shifting of the orbits is necessary. This procedure allows 5- to 8-mm medial shifting of each side. It is possible only if the anterior cranial fossa with the cribriform plate does not extend downward between the orbits and is situated at the level of the orbital roofs. If the cribriform plate's position is farther downward, there is a high risk of entering the intracranial space and damaging the dura mater with the consequence of cerebrospinal fluid leakage and other complications.

Radiologic documentation (by computed tomography) is necessary to define the exact position of the cribriform plate and the anterior and medial cranial fossae. The authors recommend three-dimensional imaging for better evaluation. If there is any doubt about the extent of the cranial cavity, it is safer to make a bur hole in the temporal fossa and protect the cranial contents with a brain spatula than to perform a blind orbital osteotomy.

The transcranial procedure is the safer one.

Step-by-Step Procedure

1. Coronal approach and dissection of the periorbital and nasal bones are as described earlier; in addition, the zygomatic arch and complex are extensively exposed, and the anterior parts of the temporal muscles are elevated.

2. A horizontal osteotomy through the frontonasal suture is made. It is continued posteriorly beneath the cranial base and downward into the orbital floor through the infraorbital fissure, into the lateral orbital wall, and forward beneath the cranial base and through the frontozygomatic suture. The zygomatic complex, including the lower lateral orbital rim, is included in the osteotomy. This procedure is also performed on

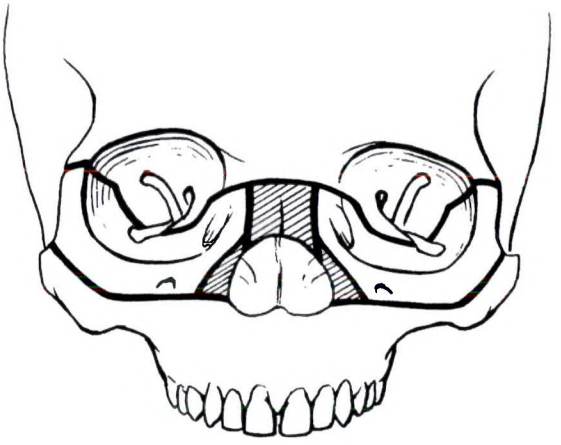


Figure 27–15. Osteotomy of the orbital walls, excluding the orbital roof, is shown. The lower part of the orbits is rotated to the midline after the nasal ostectomy.

the other side, and a strip of interorbital bone of the desired width is removed from the nasal structure.

3. The transoral approach is as described earlier. The zygomatic osteotomy is exposed, and a horizontal osteotomy running beneath the infraorbital foramen is made through the maxillary sinus into the pyriform aperture beneath the inferior turbinates. The orbital structures are then mobilized from intraoral and extraoral exposure. Triangular bone areas are removed along the pyriform margin to avoid nasal obstruction. The medially rotated midfacial structures are fixed with wires.

4. The defects within the lateral orbital walls and the orbital floor are bridged by lyophilized cartilage sheets (to be described). The nasal form is corrected by using a strut of lyophilized cartilage, which is fixed by a wire to the lacrimal bones or to the frontal bone. Wire fixation is also necessary at the lateral orbital rims and sometimes within the canine fossae.

5. The defects within the zygoma are bridged with lyophilized bone and microplates.

6. The lateral canthal ligaments are fixed inside the lateral orbital wall through bur holes.

Combination Procedure of Subcranial Hypertelorism Operation and LeFort III Osteotomy (see Fig. 27–23)

This procedure is indicated if the midface, including the zygomatic complex, has to be advanced.

Step-by-Step Procedure

1. The coronal approach and dissection of the periorbital soft tissues are as described earlier.
2. The osteotomy is performed as described earlier.
3. The connecting osteotomy is made between the infraorbital fissure and the pterygomaxillary suture on both sides; the pterygoid plates are separated from the maxillary tuberosity. An osteotomy of the zygomatic arch is also made.
4. The orbital walls are mobilized, and the orbital structure is approximated.
5. The midface is mobilized.
6. All bony structures are fixed according to the preoperative planning with wiring and mini- or microplates.

Combination Procedure of Subcranial Hypertelorism Operation and LeFort I Osteotomy (see Fig. 27–24)

This procedure is indicated only when the tooth-bearing part of the upper jaw has to be mobilized for occlusal reasons but the zygomatic complex does not.

The infraorbital horizontal osteotomy is connected to the LeFort I bone cut running under the zygomatic buttress to the tuberosity area, where the pterygomaxillary junction is separated by a chisel.

The Transcranial Hypertelorism Procedure

Tessier's Transcranial Hypertelorism Operation (THO) (Fig. 27–16)

Tessier's hypertelorism procedure[8] revolutionized the correction of monstrous craniofacial malformations. Through this and other methods, Tessier inaugurated new dimensions of cranial base surgery; this transcranial approach to the orbits and cranial base has also widely influenced the field of craniofacial trauma and the treatment of craniofacial tumors. The THO allows movements of the orbits in all directions and also the correction of the hypotelorism by outward rotation of the orbits (Fig. 27–17). The procedure is not easy to understand; three-dimensional imagination is necessary in order to perform the cranial, orbital, and midfacial osteotomies and movements (Fig. 27–18).

Anesthetic Considerations

The transcranial hypertelorism procedure is one of the longest operations in the whole field of surgery. Operations lasting 8 to 15 hours and even longer are not rare. The anesthesiologist requires experience in delivering long-term anesthesia and must be equipped with all modern monitoring modalities. Blood loss during surgery can be severe, and in contrast to the subcranial procedures described earlier, it is usually not possible to avoid homologous blood transfusions. It is the surgeon's responsibility to reduce or avoid such transfusions; therefore, the hemodilution technique and intraoperative autotransfusion with the cell saver apparatus are routinely used.[9] The cell saver is also used in the oral cavity.

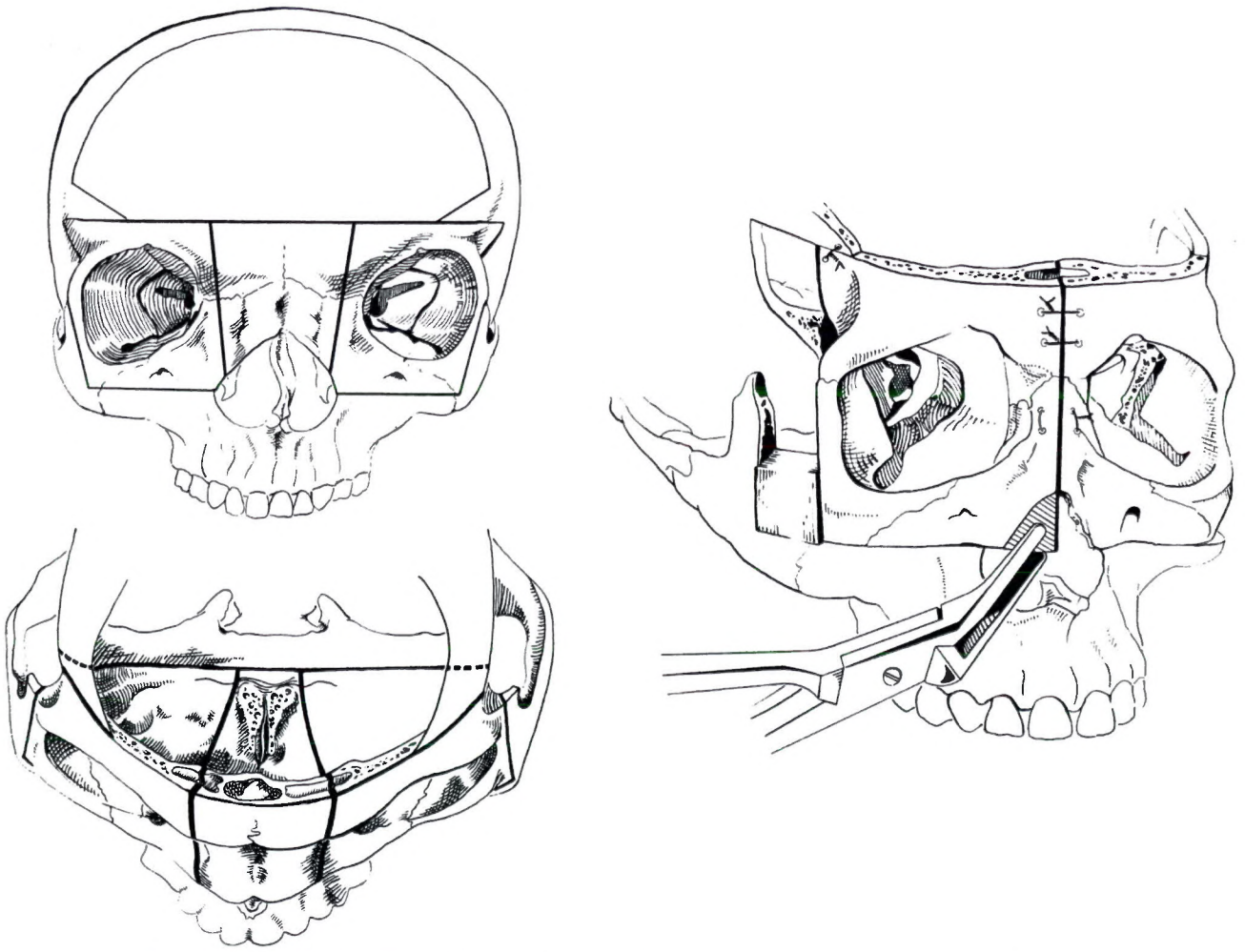


Figure 27–16. Tessier's transcranial hypertelorism procedure with resection of the cribriform plate.

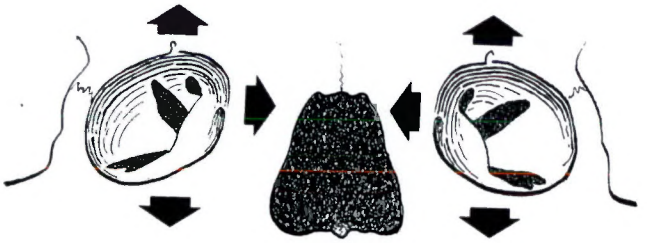
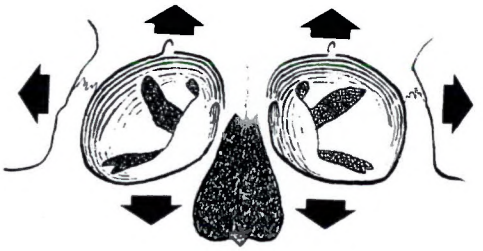


Figure 27–17. Tessier's THO procedure allows movements of the orbits in all necessary directions and also makes possible the correction of hypotelorism by lateralization of the orbits.



Neurosurgical Considerations

A transcranial procedure should be performed in collaboration with a neurosurgeon, especially when several malformations such as encephalomeningoceles or tumors such as neurofibromas are present. During elevation of the frontal lobe from the anterior cranial base, care should be taken not to damage the olfactory nerve filaments. Tessier's original hypertelorism procedure[8] includes the resection of the cribriform plate with the consequence of loss of olfaction (see Fig. 27–16). Soon they and others, such as Converse and colleagues[10] and Obwegeser and Farmand[11] attempted to preserve the cribriform plate by removing only a strip of bone laterally to the cribriform plate on each side (Fig. 27–19) to facilitate the rotation of the orbits. This is possible

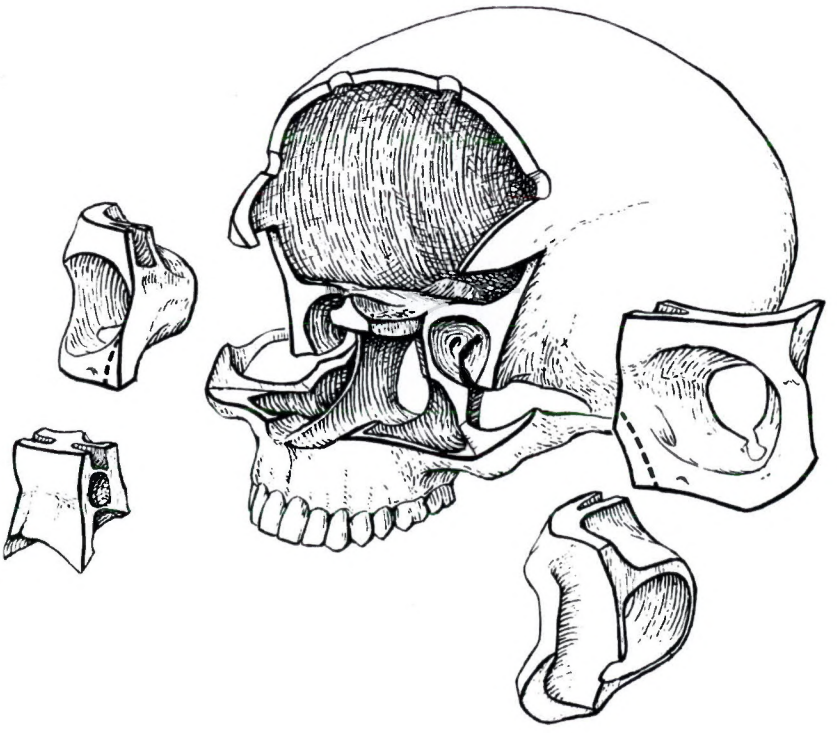


Figure 27–18. For three-dimensional understanding of the movements of the orbits, an explosion picture is presented. The cribriform plate in this picture is left in place but must be reduced or resected, as described in the next two figures.

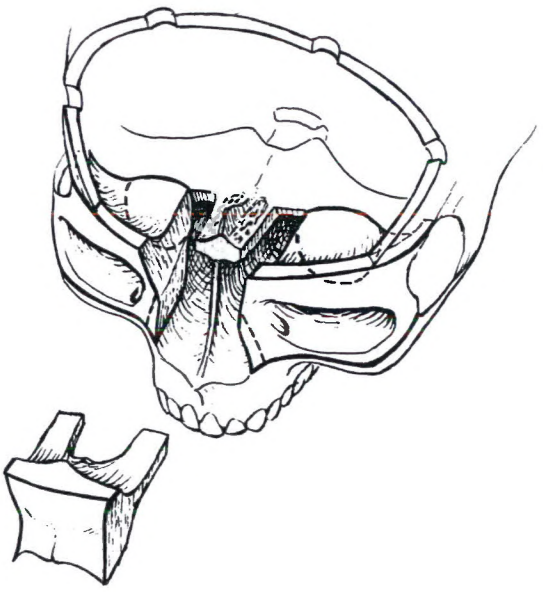
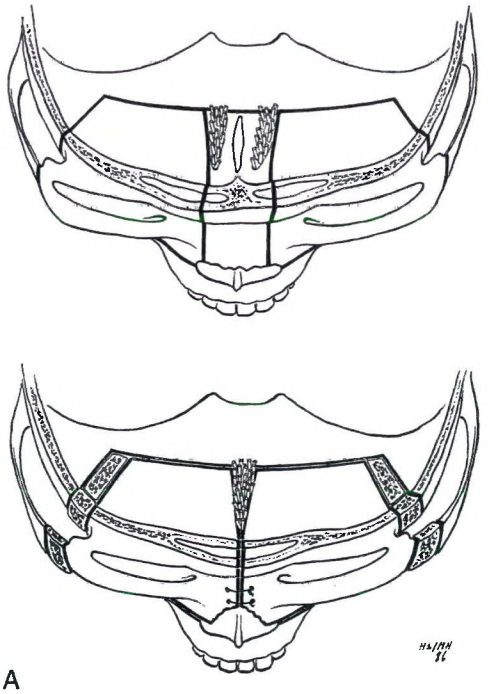


Figure 27–19. Transcranial hypertelorism procedure without resection of the cribriform plate. The osteotomy is performed laterally to the cribriform plate and allows medial orbital rotation (Converse method). This technique should be employed only in less severe cases.

only in cases of mild hypertelorism in which the cribriform plate is not too wide and especially not separated in the midline with dislocation of one or the other half. For severe cases, the authors have introduced their own method of preservation of the olfactory nerves[12,13] by dissecting every nerve filament out of the cribriform plate (Fig. 27–20) to permit medial orbital rotation.

Ophthalmologic Aspects

As a member of the craniofacial team, the ophthalmologist is responsible for an extensive examination. It is important to recognize that all orbital movements can cause diplopia, especially in cases of hypertelorism in which binocular vision is present preoperatively. Experience indicates that when focusing was not possible



A

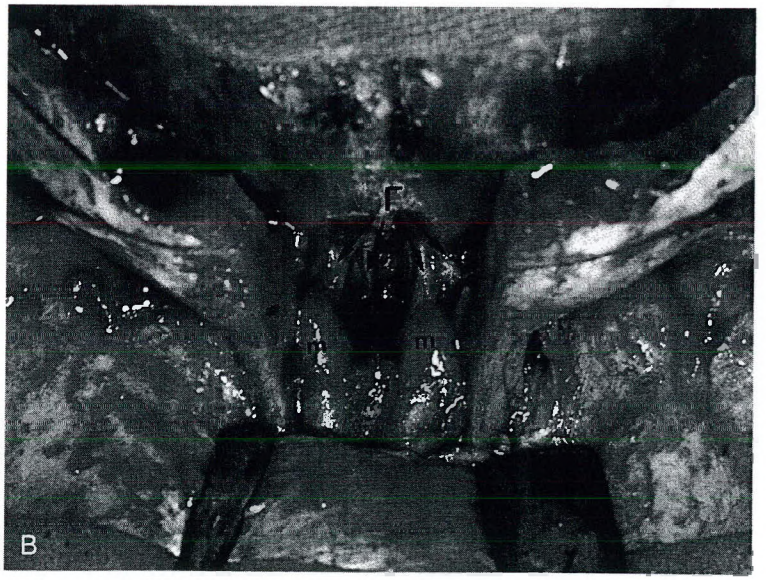


Figure 27–20. Sailer's transcranial hypertelorism procedure with resection of the cribriform plate but preservation of the olfactory nerve filaments (Sailer's method). The drawing *(A)* demonstrates nerve filaments assembled in the midline after orbital rotation (bottom); the intraoperative photograph *(B)* depicts the free filaments (F) running into the intact nasal mucosa (m).

preoperatively, the diplopia sometimes disappears after hypertelorism correction without further ophthalmologic measures.

Step-by-Step Procedure

1. The coronal approach is used; the skull, the orbital region, the nose, and the zygomatic area are dissected subperiosteally.
2. The anterior halves of the temporal muscles are elevated by freeing the temporal fossa.
3. The osteotomies and cranial bur holes are marked with methylene blue and bur lines (Fig. 27–21*A,B*).
4. The craniotomy is performed and the frontal bone removed (see Fig. 27–21*C*).
5. The frontal lobes of the brain are elevated, and the anterior cranial fossa and the cribriform plate are exposed as well as possible.
6. An osteotomy of the interorbital frontal and nasal bone (see Fig. 27–21*D*) is made, with preservation of the nasal mucosa. Now the access to the cribriform plate is free: With the use of magnifying glasses, the bone around the olfactory nerve filaments is removed (see Fig. 27–21*E*) with a mosquito clamp; this is usually necessary in the anterior and the lateral parts of the cribriform plate.
7. The ethmoids and the mucosal lining of the frontal sinus are removed.
8. A circular osteotomy of all orbital walls is made within the posterior part of the orbital pyramid just in front of the supraorbital fissure.
9. An osteotomy is made through the zygomatic complex.
10. The intraoral incision is made as described earlier, and the midface and the pyriform aperature are exposed subperiosteally.
11. A horizontal infraorbital connecting osteotomy is made between the zygomatic bone cut and the pyriform aperature; the osteotomy lies beneath the inferior turbinates.
12. Careful slow mobilization of both orbits is accomplished with the use of chisels.
13. A triangular bone piece along the pyriform aperature is removed to allow medial movements of the orbits without compromising the nasal airways.
14. Bur holes and wires are placed in the glabella region, and the orbits are slowly approximated by twisting the wires with direct control of the orbital contents and the olfactory nerve filaments. If bradycardia occurs because of compressing or pulling the orbital contents, mobilization is discontinued until normal heart rhythm returns. The process then continues.
15. The positions of the supraorbital and infraorbital rims are determined according to the preoperative planning, and lyophilized bone blocks are interpositioned into the defect. Adjustments of the orbital position can be made once the final position is achieved. The segments are fixed with wires, miniplates, or microplates.
16. Now the defects within the orbital walls are bridged with lyophilized cartilage slices, and those within the area of the cribriform plate are bridged with bone powder gained from the craniotomy plus lyophilized cartilage chips and bone morphogenetic proteins.
17. The frontal calvaria is then placed into the desired position, adapted by trimming edges, and fixed, preferably by wires (see Fig. 27–21*F,G*).
18. The temporal muscles are fully mobilized, advanced, and fixed to the lateral orbital walls and rims by sutures running through bur holes. The lateral canthal ligaments are now fixed into their new position through bur holes within the lateral orbital walls.
19. The surplus of skin in the nasal and forehead region is excised, preferably with the creation of a straight scar in the middle of the dorsum and the forehead (nasal reconstruction). When necessary, a forehead flap is harvested for construction of the columella and nasal tip.
20. Other nasal corrections are performed to achieve the best possible initial result (see Fig. 27–21*G,H*).
21. Two subgaleal Redon drains are placed under the temporal muscles on each side and led out through the *contralateral* retroauricular area.
22. Subcutaneous scalp stitches are placed, and the outer incision is closed by stapling.
23. The oral incision is closed with a continuous Supramid suture after additional miniplating or wiring of the infraorbital rim to its new position in relation to the maxilla.

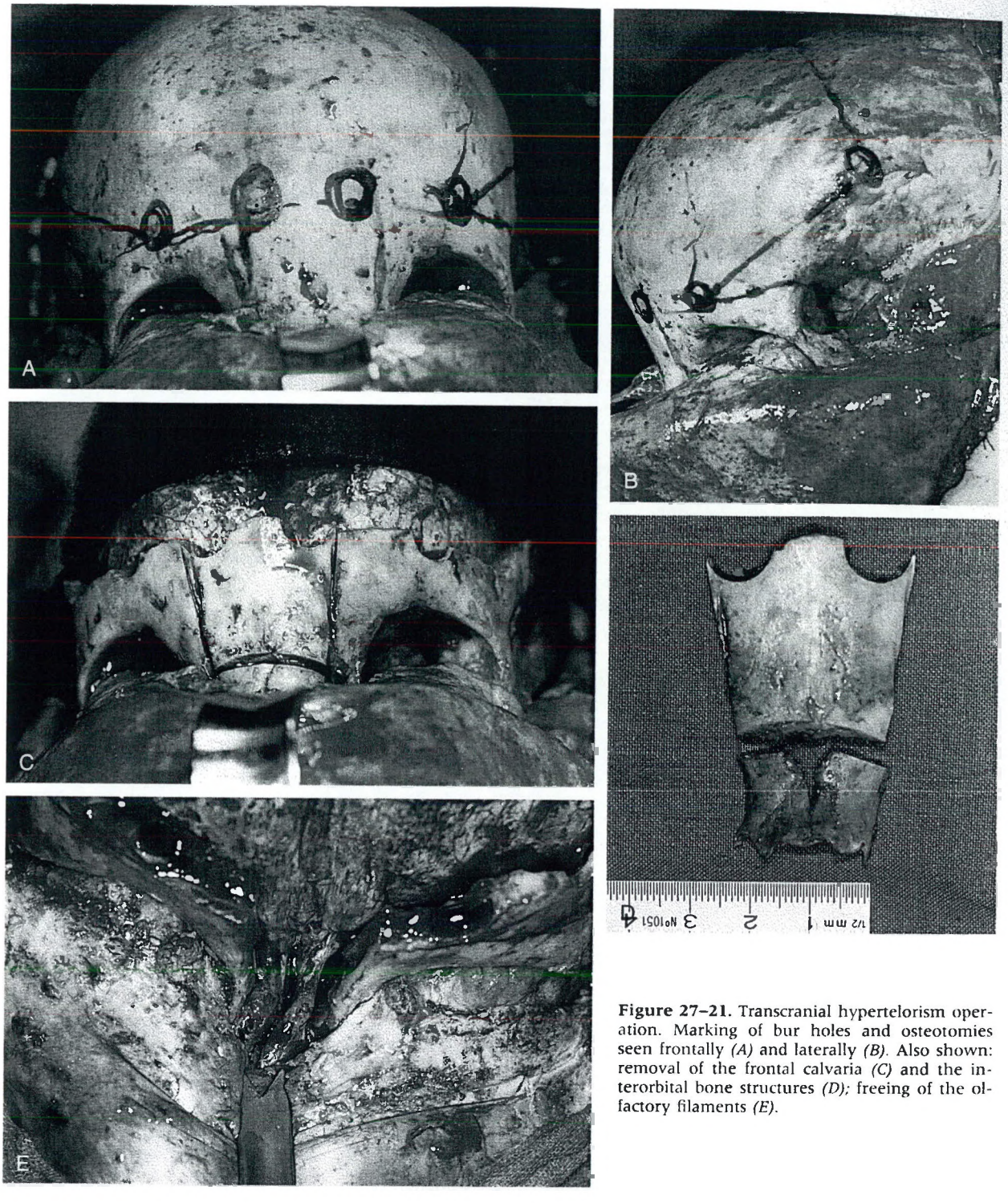


Figure 27–21. Transcranial hypertelorism operation. Marking of bur holes and osteotomies seen frontally *(A)* and laterally *(B)*. Also shown: removal of the frontal calvaria *(C)* and the interorbital bone structures *(D)*; freeing of the olfactory filaments *(E)*.

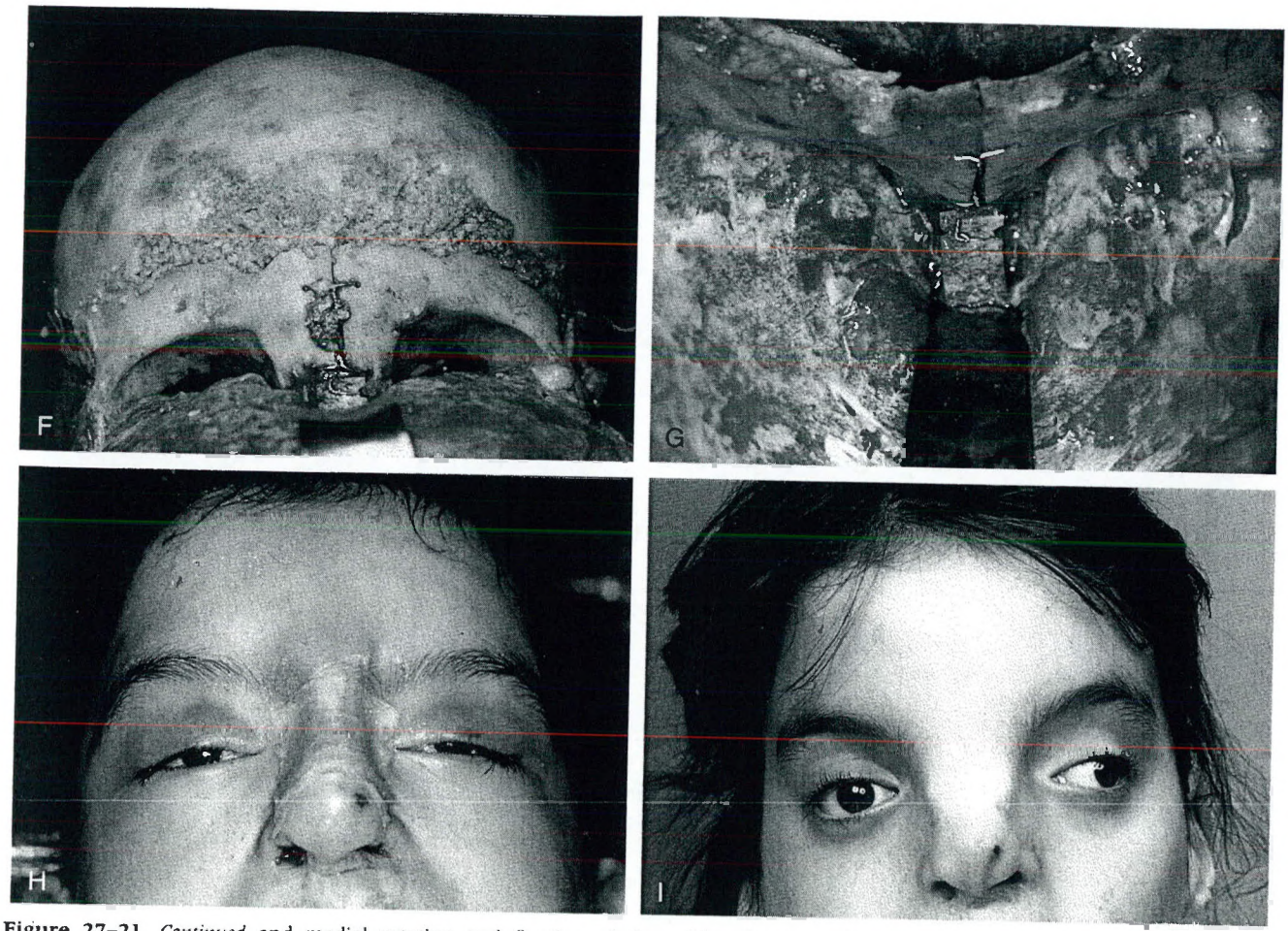


Figure 27–21. *Continued* and medial rotation and fixation of the orbits; the intraorbital osteotomies are clearly seen *(F)*. Nasal correction is by means of bone or lyocartilage struts *(G)* fixed by wires; at the end, a nasal acrylic stabilization dressing is applied *(H)*. *I*, preoperative condition of the same patient.

Remarks

When possible, the medial canthal ligaments are left in place in spite of the fact that this makes the intraorbital circular osteotomy more difficult. If the medial canthal ligaments require special movements, they are detached and placed into the correct position by twisted wires pulled through the approximated medial orbital walls to the opposite side and fixed there in the routine way.

The Facial Bipartition Procedure (Fig. 27–22)

This procedure is indicated in cases of cranial clefts No. 14 to 10 according to Tessier's classification[14] and also in cases of hypertelorism with "antimongoloid" orbital positions and an anterior open bite.

The procedure was initially described as facial bipartition[15–17] because both full halves of the face are mobilized, the orbits are tilted medially, and the dental arches are rotated downward. As a result of the downward rotation of the anterior maxilla, closure of the open bite occurs. The disadvantage is that the amount of the open bite influences the amount of the medial movements of the orbits. The same type of procedure had already been performed in the Department of Maxillofacial Surgery Zurich in 1970 in the famous case of Antonio, who presented with a double formation of the midface with two premaxillae and extreme hypertelorism.[18]

In the facial bipartition procedure, there is no infraorbital horizontal osteotomy. Instead, an osteotomy runs from the inferior orbital fissure through the posterior wall of the maxillary sinus into the pterygomaxillary junction, which is also separated. In addition, the zygomatic arch is dissected. The alveolar process is divided between both first incisors (if this is necessary), as is the whole palatal bone in the sagittal plane.

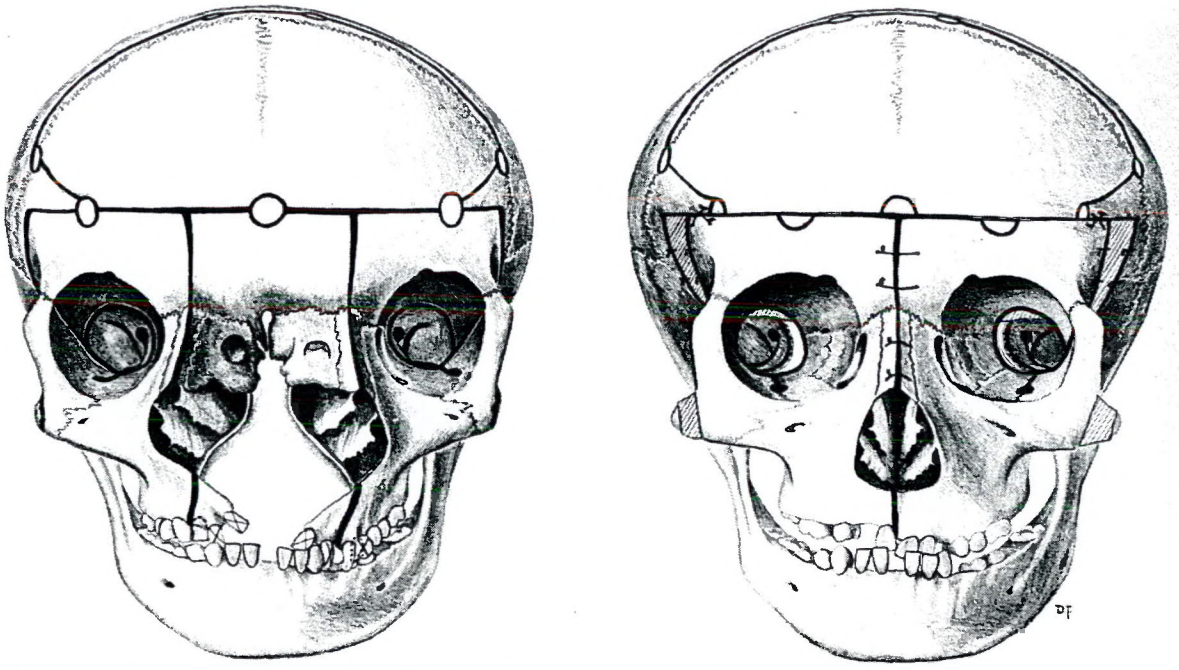


Figure 27–22. Facial bipartition, as it was performed in Zurich by Obwegeser in 1970 in the case of Antonio. No infraorbital horizontal osteotomy is performed; instead, an osteotomy is running from the infraorbital fissure through the posterior wall of the maxillary sinus to the pterygomaxillary junction, which is separated. (From Obwegeser HL, Weber G, Freihofer H-P, Sailer HF: Facial duplication—The unique case of Antonio. J Maxillofac Surg 1978; 6:179–198.)

After this, both halves of the face, including the orbits and the dentition, can be mobilized.

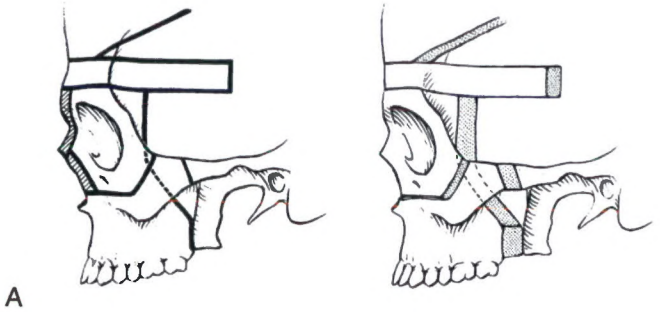
Combined Hypertelorism Operation and LeFort III Osteotomy
(Fig. 27–23)

Patients with some of the craniofacial syndromes (e.g., Crouzon and Apert syndromes) present with varying degrees of hypertelorism. Full correction of these syndromes therefore includes frontofacial advancement (forehead, orbital rims, zygomatic complex, and maxilla for occlusal correction) in combination with the normalization of the IOD.

The authors' original procedure for these cases includes the correction of the exorbitism together with the hypertelorism. It is most important in these patients that the intraorbital osteotomy is positioned not too far posteriorly but in the area of the largest circumference of the ocular globe. This makes it possible to advance the orbital walls, thus correcting the exorbitism as well as shifting the globes medially by approximation of the mobilized orbits. The reconstruction of the orbital walls is of great importance for ensuring the position of the globes. In order to move the zygomatic complex forward together with the maxilla, a connecting osteotomy is performed between the infraorbital fissure and the pterygomaxillary junction. The pteryoid plates are separated from the maxillary tuberosity. The zygomatic arch is osteotomized. Special care must be taken not to fracture the zygomatic complex during mobilization.

Important

If in cases of frontofacial advancement in combination with THO the circular intraorbital osteotomies are placed posteriorly as described for the routine THO, the eye globes are pulled forward with the optic nerves, and the protrusion of the eyes is not corrected.



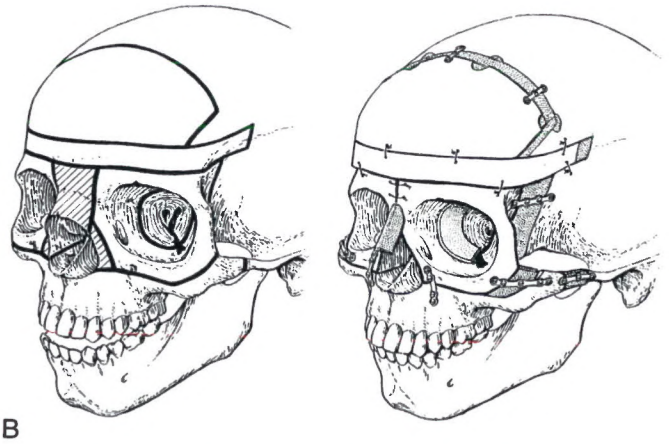




Figure 27–23. Combined THO and LeFort III osteotomy (frontofacial advancement). During this procedure, the forehead, the orbital rims, the zygomatic complex, and the dentition are advanced *(A)*, and the orbits are simultaneously approximated. It is necessary to place the circular intraorbital osteotomies slightly farther forward *(B)* than in the routine hypertelorism correction. The defects within the frontal and parietal bones, the zygomatic bone, and the zygomatic arch are bridged by bone grafts (lyobone or autologous calvarial bone). Fixation is performed by miniplates and wiring. The intraoperative photograph *(C)* shows the amount of forward movement *(arrows)* of the forehead and orbits and the closure of the skull defect with lyocartilage (LC); also clearly visible is the stabilization of the orbital rims with lyobone (LB). The patient, who had Apert syndrome, is shown before *(D)* and after *(E)* this procedure.

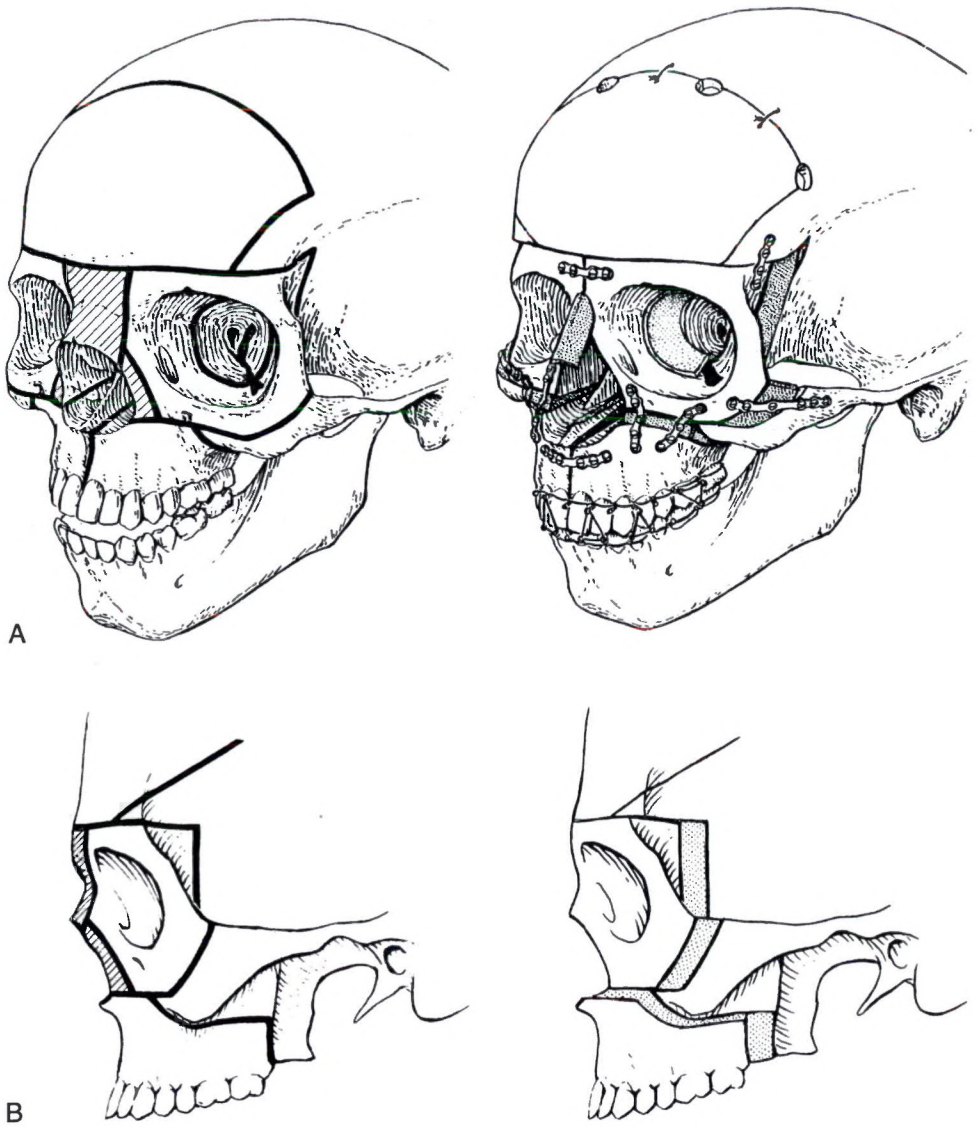


Figure 27–24. Combined THO and LeFort I osteotomy. The bone cut of the LeFort I osteotomy joins the infraorbital horizontal osteotomy. The upper jaw can be divided in the midline *(A)*. The procedure allows independent orbital and maxillary movements *(B)* and, at the same time, orbital approximation.

Combined Hypertelorism Operation and LeFort I Osteotomy
(Fig. 27–24)

The indication for this correction is hypertelorism and an occlusal disturbance, such as an open bite or class III occlusion, without the necessity to also advance the zygomatic complex (see Fig. 27–24*A*). A LeFort I osteotomy in one or more parts can be performed simultaneously with the THO. The whole zygomatic complex is then only "pedicled" to the zygomatic arch (see Fig. 27–24*A,B*).

If the orbits require advancement (see Fig. 27–25*A*), the circular intraorbital osteotomy should run more anteriorly than in the routine THO (as mentioned in the section "Combined Hypertelorism Operation and LeFort III Osteotomy"). Stabilization is accomplished by microplates and miniplates. Intermaxillary fixation is usually not necessary just during fixation of the segments by miniplates.

This procedure could also be used in cases of hypertelorism caused by a huge encephalocele extending to the oral cavity; in the patient shown in Figure 27–25, there was a palatal cleft with a width of 50 mm and a complete circular nonocclusion.

Nasal Reconstruction

Hypertelorism correction necessitates construction of the hard and soft nasal tissues. It is advisable to perform the best possible reconstruction of the nose simultaneously with the hypertelorism procedure; the patient will be grateful for this.

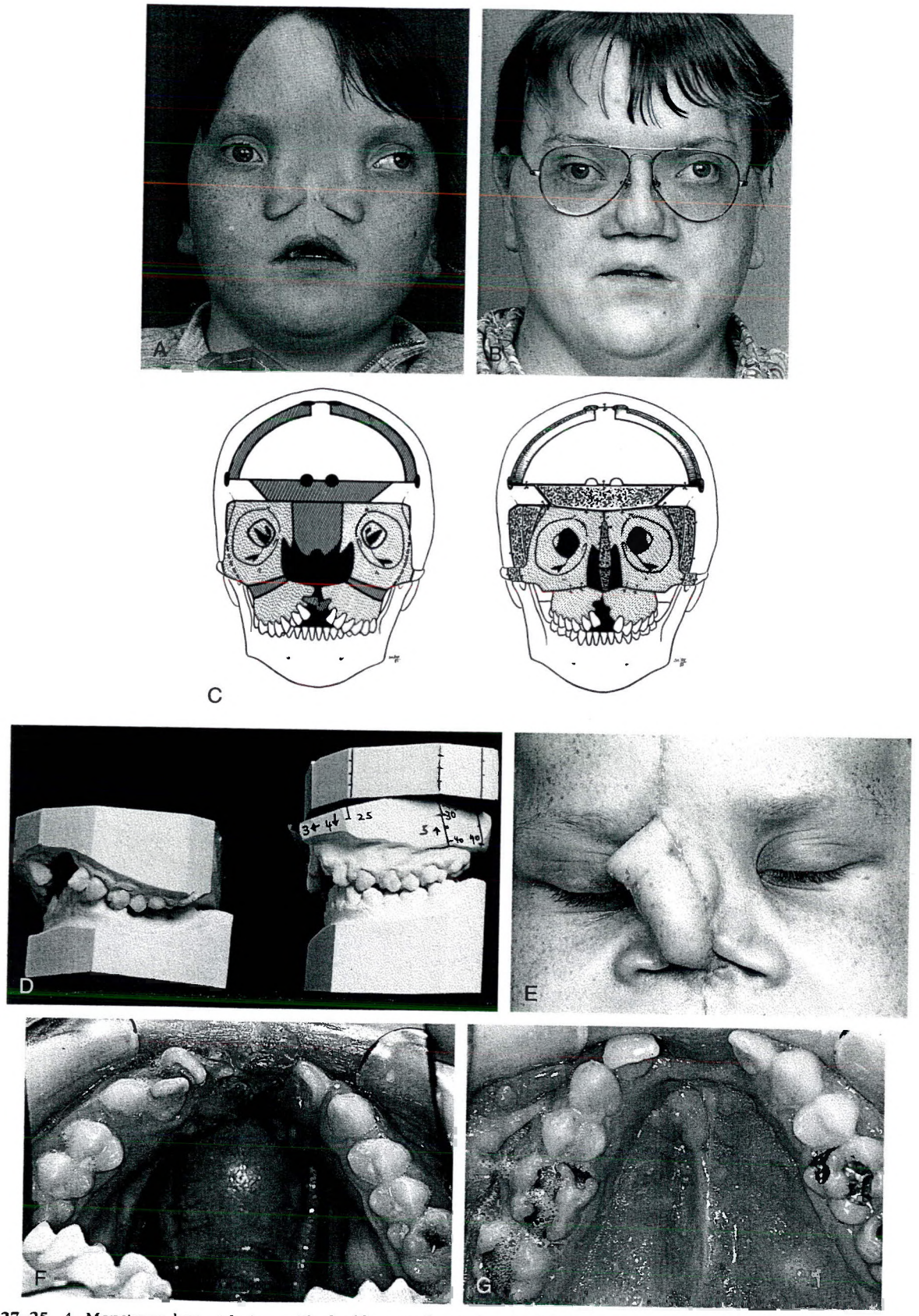


Figure 27–25. *A,* Monstrous hypertelorism with double nose formation caused by an encephalocele expanding into the oral cavity before correction; *B,* 10 years after correction. For correction of the nonocclusion, a bilateral LaFort I osteotomy was performed, and the maxillary segments were raised and narrowed according to the preoperative planning *(C)* and the model operation *(D).* Nasal reconstruction was achieved with a forehead flap *(E)* in combination with an L-shaped calvarial bone graft. The wide palatal cleft *(F)* that was narrowed by the LeFort I operation was closed in a second-stage procedure *(G)* with a cranially based pharyngeal flap. (From Sailer HF, Landolt AM: Hypertelorism with herniation of brain and pituitary gland into the oronasal cavity. *In* Marchac D [ed]: Craniofacial Surgery. Berlin: Springer, 1987.)

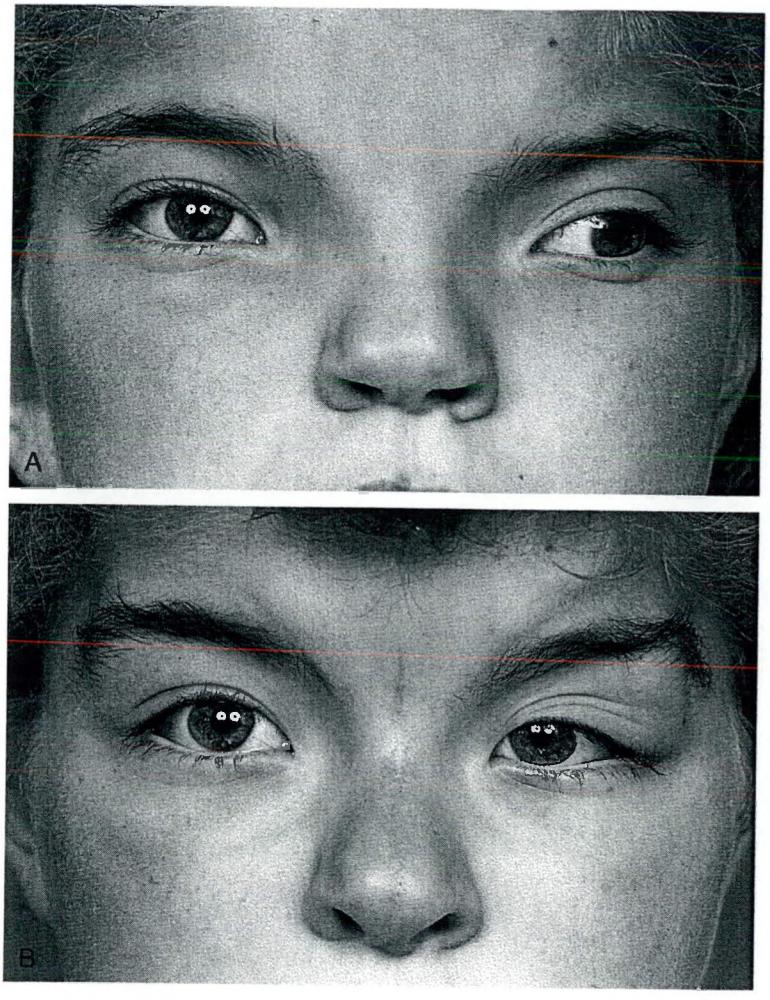


Figure 27–26. Patient with hypertelorism before *(A)* and after THO with median nasal scar *(B)*.

A new nasal dorsum is always constructed from homologous lyocartilage[19,20] or calvarial bone, either of which is fixed to the lacrimal bone by wires. The surplus of skin in the glabella and the nasal area is excised, and this results in a midline scar running from the forehead along the nasal dorsum to the nasal tip. This scar is in most cases barely visible after some time and presents the best esthetic result (Fig. 27–26*A,B*). Less desirable is a Z-plasty in the area of the frontonasal angle, because the resulting scars are easily visible even if they are very fine. In cases of hypertelorism in which the dorsum and the columella are too short, good results are achieved with Sailer's original method of a caterpillar flap and a forked flap (Fig. 27–27; see also Fig. 27–3) or with their original intracolumellar forked-flap procedure (Fig. 27–28).

When the nasal tip and the columella require construction, the interorbital surplus of skin is used for a forehead flap, as in the patient shown in Figure 27–25, who had an encephalocele extending into the oral cavity. In cases of complex forms of hypertelorism combined with facial clefts (Fig. 27–29*A*; see also Fig. 27–6), a variety of nasal hard and soft tissue corrections are made in order to achieve an acceptable result in the first operation (see Fig. 27–29*C,D*). Additional revision, however, may be necessary.

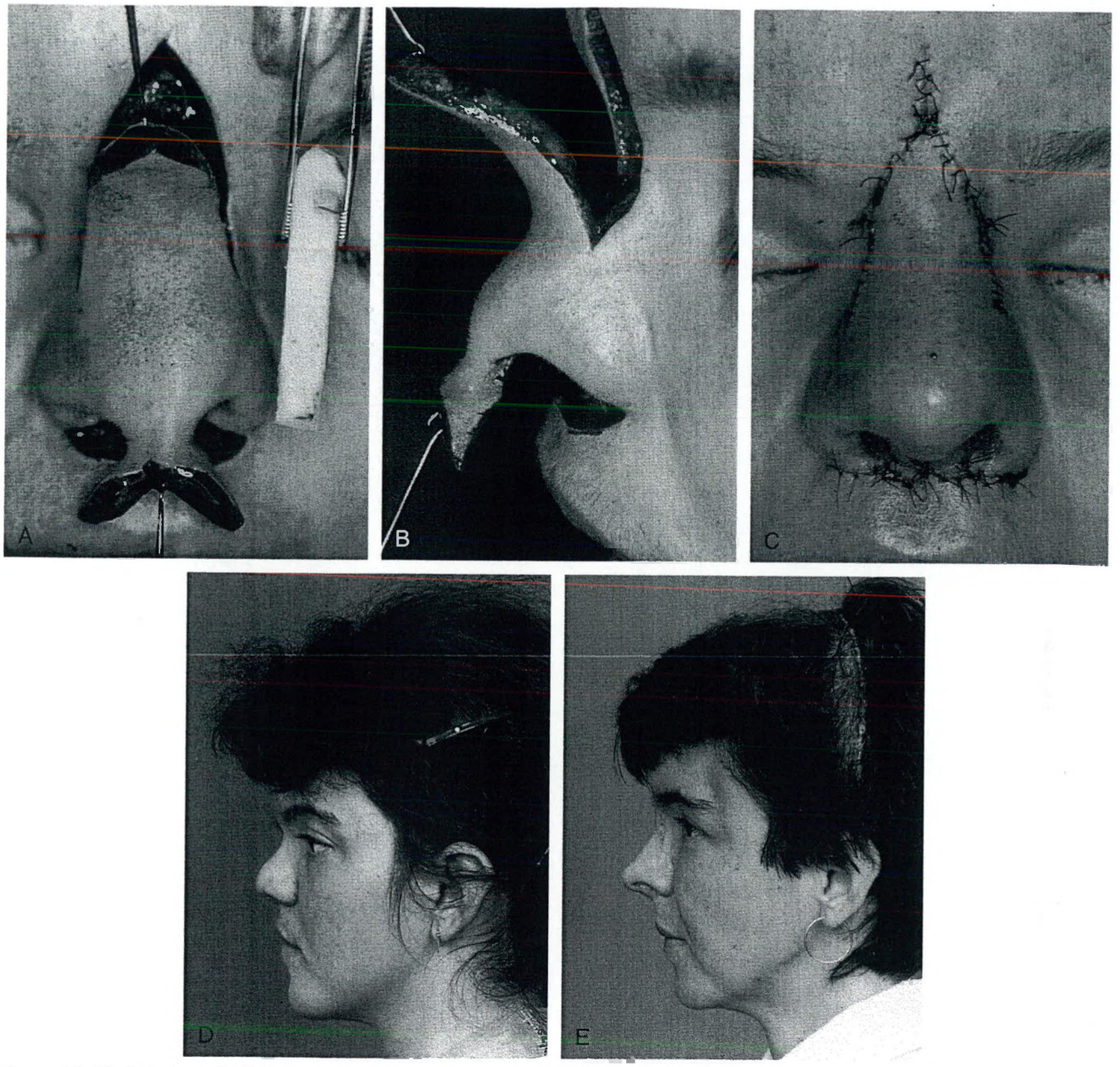


Figure 27–27. Sailer's method of increasing nasal length and projection. *A*, A caterpillar flap is designed along the dorsum, and a strut of lyocartilage is inserted. *B*, Blood supply is ensured over the nasal alae to the caterpillar flap and the columellar forked flap. *C*, Result after elongation of the dorsum and the columella. The result of this nasal and columellar elongation is seen in the profile of a patient with hypertelorism before *(D)* and after *(E)* surgery. The same patient is seen in Figure 27–3.



Figure 27–28. Intracolumellar forked-flap procedure (Sailer) in cases with a wide columella *(A)*. The central part of the columella stays in place *(B)*; the lateral parts plus the vestibular extensions move upward and give excellent tip projection *(C)*.



Figure 27–29. Asymmetric hypertelorism with several facial clefts *(A,B)*, presenting a complex reconstructive nasal problem; the result *(C,D)* achieved in the first operation is acceptable, but revision is required.

RECONSTRUCTION MATERIAL IN CRANIOFACIAL SURGERY

All types of craniofacial surgery result in defects, and these must be filled or bridged by autologous or homologous hard tissue. Autologous split-thickness cranial bone is usually used.[2] Splitting of calvarial bone is possible only in older children and adults, not in infants. Splitting and forming the brittle calvarial bone is time consuming and prolongs surgery. In the authors' unit, the use of autologous bone in craniofacial surgery is obsolete. They have introduced and used for many years lyophilized homologous bone from different regions (sternum, calvaria, mandible, ribs, femur) as well as lyophilized homologous cartilage in craniomaxillofacial surgery.[19,20] Since 1991, these materials have been combined with purified bovine bone morphogenetic proteins, which accelerate the incorporation of homologous lyophilized bone and cartilage.[21–23] Lyocartilage is the most suitable reconstructive material in craniofacial surgery. The lyocartilage from ribs is cut into slices by a dermatome. These slices are ideal for the reconstruction of the orbital walls as well as for the reconstruction of the cranial base (Fig. 27–30; see also Fig. 27–23*C*). The bridging of the defects within the zygomatic bone is also performed with lyophilized homologous calvarial bone (Fig. 27–31). Defects of the midface after the LeFort I operation are closed with

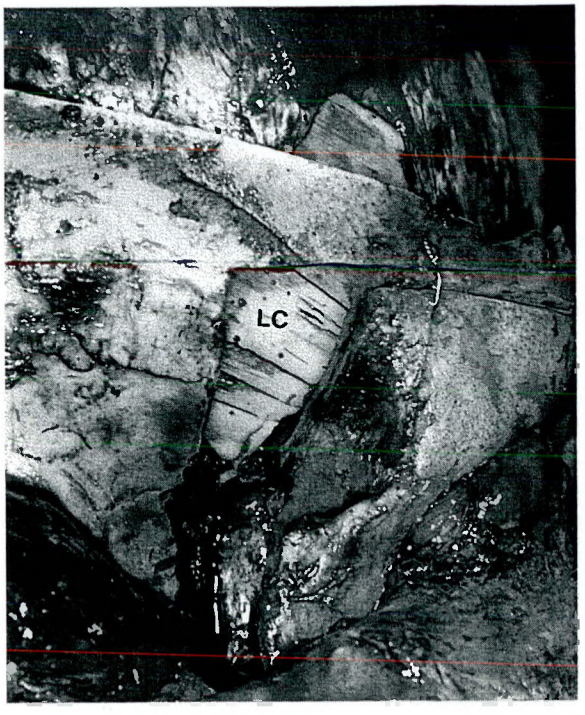


Figure 27–30. Reconstruction of the lateral anterior cranial base, using a slice of lyocartilage (LC).



Figure 27–31. Bridging a defect within the zygomatic complex after medial orbital rotation, using calvarial lyobone (LB).

lyophilized sternum or iliac bone. The lyobones and the lyocartilage are loaded with bone morphogenic protein.

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