Surgery Related to the Correction of Hypertelorism

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Tessier et al. have more than once stressed the fact that "hypertelorism has its real difficulties, not so much in reducing an exaggerated interorbital distance as in correcting the other malformations which increase it or are associated with it."

Our experience with the intracranial correction of orbital hypertelorism, which is now based on the treatment of 30 patients, has convinced us of the correctness of this statement. Stigmata indeed may remain following an adequate reduction of the interorbital distance (I.O.D.). Some of these are obviously produced by the operation itself, such as canthal drift, enophthalmus, ptosis, shortening or lengthening of the palpebral fissure, and temporal depression. Others have been present since birth, but become more conspicuous after the correction of the orbital hypertelorism. These malformations may involve the forehead (widow’s peak and eyebrow dystopia), the orbit (abnormal orientation and configuration), the canthi (medial and lateral canthal dystopia), the eyelids (colobomata, ptosis), the eye (anophthalmia, microphthalmia), the eye muscles (strabismus), the nasolacrimal apparatus (obstruction), the nose (shortness, bifidity, alar clefting, choanal atresia), or the maxilla (sagittal arching, clefting).

So far the management of these deformities has been discussed by few authors.\textsuperscript{3-5} This paper, therefore, is devoted to an analysis of the factors that cause new deformities to develop and to a discussion of the measures that have been taken for their prevention. Methods used for the correction of associated deformities in as far as they were present in our patients are also dealt with.

Acquired Stigmata

A discussion of the stigmata following the correction of hypertelorism requires first of all an answer to the following questions:

1. What is orbital hypertelorism or teleorbitism?

Tessier\textsuperscript{3} defines orbital hypertelorism as a congenital condition in which the interorbital distance increases from the apex to the orbital rim and distinguishes between three degrees of severity. Objective measurements are only taken between the anterior lacrimal crests (intercanthal distance, I.C.D.). Tessier confesses that this classification is artificial because it is not possible to define exactly when the orbit becomes lateralized. In his opinion, a description of the malformation is difficult because it is always secondary to other malformations, such as clefts.

Converse et al.\textsuperscript{7} differentiate between ocular hypertelorism, which is characterized by an increased divergence of the orbital axis from the midsagittal line, and pseudohypertelorism. The latter name refers to patients in whom the interpupillary distance and interorbital distance are normal and an increase of the intercanthal distance is found, such as in traumatic telecanthus, blepharophimosis, and so forth.

Munro and Das\textsuperscript{8} seem to include trauma or fibrous dysplasia as a cause of hypertelorism and distinguishes between four types of deformities that can be observed along the length of the medial orbital walls in an anteroposterior direction. In one of these (type IV), an increase of the...
interorbital distance in the posterior part of the interorbital space appears to be associated with a normal intercanthal distance.

Classifying hypertelorism with its different etiologies and varied appearances is not as simple as one might believe. Not only is it possible to differentiate between deformities along the length of the medial orbital wall in an anteroposterior direction, as Munro does, but differences in a craniocaudal direction also exist. A normal intercanthal distance may be associated with an increased interorbital distance in the superior part of the interorbital space in malformations such as craniosynostosis, encephaloceles, pneumatoceles, and fibrous dysplasias, while an abnormal intercanthal distance without further increase of the interorbital distance is frequently encountered after trauma. An overall increase of the interorbital distance without lateralization of the orbit may finally be observed in patients with bone deformities such as craniofacial dysplasia.

The application of the name pseudohypertelorism to all patients with an increase of the interorbital distance but without lateralization of the orbits tends to obscure the fact that the distance between the orbits is indeed increased. From a clinical point of view, however, differentiation between different types of hypertelorism is indicated because correction of hypertelorism is less difficult when there is no lateralization of the orbits (Fig. 1).

We therefore suggest that distinction is made between orbital hypertelorism, in which there is true lateralization of the orbits, and interorbital hypertelorism, in which the malformation is limited to the interorbital space and no lateralization of the orbits is observed.

Lateralization is present when the distance measured between the lateral orbital walls exceeds the values found by Jöhr and Laestadius et al. for corresponding age groups or when the angle between the lateral orbital walls on a computed tomogram is more than 90 degrees.

2. What are the skeletal changes produced by medial transposition of the orbits?

Medial dislocation and correction of the position of the orbital aperture is not so much obtained by a linear as by a rotatory movement. The nature of this movement depends on the degree to which the medial walls of the orbit can be displaced medially. If these walls can be moved over the same distance, the orbital transposition will consist of a single rotatory movement around an imaginary craniocaudal axis, which corresponds with the line of osteotomy through these walls. This movement will inevitably result in an anterior projection of the lateral wall (Fig. 2). Orbital transposition becomes more complex when the anteroposterior divergence of the orbits is associated with caudocranial divergence.

Fig. 1. (Left) Severe frontal encephalocele. (Center) Skeletal malformation observed during surgery. The interorbital distance has increased; the intercanthal distance is normal. (Right) Two months following correction.
these patients, the superior interorbital distance is greater than the inferior interorbital distance, and orbital approximation will be associated with a second rotatory movement around an anteroposterior axis through the medial wall of the maxillary sinus (Fig. 3). The latter movement will obviously be followed by a cranial displacement of the lateral wall of the orbit, and Tessier therefore has very wisely suggested the preservation of a frontal crown as a safeguard against the projection of the orbital frames.

**Canthal Drift**

The most important of all the deformities that may be produced by the operation is probably canthal drift. It is extremely difficult to correct, in spite of effective medial canthopexies, and its presence after the correction of orbital hypertelorism may mar the end result. The major reason for its development seems to be the stress that may be created following the medial dislocation of the orbit and the anterior projection of its lateral wall. This lateral pull may be attributed to residual tension in the different structures, to postoperative scar contraction, and to the action of the orbicularis muscle. As a result, the following phenomena may be observed:

1. Relaxation of the canthal ligament: stretching of a tendon suture or tenodesis subject to stress is well known in hand surgery. In a canthopexy, this may also occur following strangulation and dehiscence of the canthal tendon.
2. The production of epicanthal folds as a result of relaxation of skin over the glabella and in the medial canthal region.
3. The formation of an abundance of scar tissue between periorbita and nasolacrimal skeleton.
4. The apposition of new bone after mobilization of the periosteum in young children.

It should be emphasized that canthal drift is not to be confused with bony relapse, which, according to Tessier, may be explained by pneumatization of the ethmoid or frontal bone. However, growth of ethmoid air cells is impossible, while outgrowth of remaining cells is a slow process that can hardly be held responsible for a rapid increase of the interorbital distance. More likely, partial relapse is due to technical failures, such as inadequate exenteration of ethmoidal air cells and intranasal structures or insufficient mobilization and translocation of the true functional orbital volume.

Attempts to prevent canthal drift by transcaneous and transnasal fixation of the skin and bone using lead plates may cause ulceration of this area and have little effect because some lateral pull remains in spite of a periosteal release over the lateral wall of the orbit. Stricker therefore also frees the orbicularis from its origin and insertion to reduce the effect of these laterally directed forces.

Overcorrection has also been advised by several

**Fig. 3.** Position of the orbital cone following two rotatory movements, one around the cranio-caudal axis (seen in Fig. 2), the other around an imaginary anteroposterior axis, which corresponds with the line of infracture in the medial wall of the maxillary sinus.
CORRECTION OF HYPERTELORISM

It can usually be obtained by the removal of a thick or deformed cartilaginous septum and resection of a hypertrophic concha. Its importance may be questioned, however, because overcorrection will inevitably be associated with increased lateral pull. The first author (van der Meulen) has applied the following modifications to the present technique because he was impressed by the tendency of the canthi to drift.

The first modification concerns the canthal ligaments. Early in 1975 it was decided to preserve the insertion whenever the situation made this possible, as Mustardé has done. Not only is it quite simple to perform the osteotomy behind an intact ligament after careful mobilization of the periorbita, but the advantages of this technique are also considerable, since canthal drift resulting from relaxation of the ligament can be avoided. Adaptation of the skin by transnasal fixation and a plaster of Paris splint may help to prevent hematoma formation between skin and bone. The splint is fixed by means of an Opsite sheet, which has a function similar to that of a bandage and in addition offers the advantage that the area can be continuously inspected.

The second modification aims at reducing the lateral pull by recessing the medial wall of the orbit. This recession is obtained by the removal of a small, wedge-shaped fragment of bone from the roof and the medial wall itself. As a result, the lateral outward rotation of the orbital aperture is corrected with a minimum of forward projection of the lateral wall (Fig. 4). The temporal fossa and orbit are not significantly enlarged, and traction on both canthi should logically be diminished.

While the effect of this modification on the lateral pull is difficult to evaluate, it seems certain that the decision not to detach the canthal tendon has been particularly helpful because it has removed the principal cause of canthal drift and by this has made it possible to improve our results in the majority of cases. However, the problem of canthal drift is not so readily solved in all patients, as the following example will illustrate.

During the operation on a young girl with hypertelorism in September of 1977, the interorbital distance, as measured at the site of insertion of the canthal tendon, was found to be 35 mm (grade 1 hypertelorism) (Fig. 5). This distance was reduced to 22 mm, but although the above-mentioned modifications were carried out, the end result was not as good as expected. It was therefore decided to proceed with a second correction in October of 1979. The canthal ligaments were freed and the interorbital distance of 22 mm was reduced to 16 mm by removal of bone in the medial region. A transnasal canthopexy was then performed, and a fairly thick piece of fibrotic tissue was removed. The result was satisfactory.

A reduction from 35 to 22 mm in the presence of an intact canthal insertion should have been adequate, since the average interorbital distance at age 6 is 21 mm. Canthal drift in the presence of an intact tendon insertion may, however, be explained in the following way. Medial to the caruncle, the canthal tendon bifurcates into a superficial anterior and a deep posterior limb. The anterior limb has its insertion on the frontal process of the maxilla. The direction of its course is lateromedial as well as posteroanterior. The posterior limb is associated with Horner's muscle, which, together with the orbicularis muscle, is said to maintain the position and shape of the medial canthus. Dissection of the periorbita with the posterior limb of the tendon attached to it allows for the accumulation of a fibrous substrate behind the lacrimal sac and anterior limb. This results in obliteration of the naso-orbital valley. The anterior limb will be pushed forward, changing its course into one that is purely lateromedial (Fig. 6).

![Fig. 4. (a) Horizontal section of orbit at the equator. Reduction of the interorbital distance in combination with minimal recession of the medial orbital wall corrects the position of the orbital aperture and produces minimal anterior projection of the lateral wall. (b) The orbit and temporal fossa are not enlarged, and less bone is needed to close the defects in the lateral wall of the orbit.](image-url)
ENOPHTHALMUS
The amount of sclera shown plays an important role in our evaluation of a human face. Too much, as in exorbitism, or too little, as in microphthalmia or enophthalmia, arrests ones attention. Postoperative enophthalmus was found bilaterally in one of our patients and monolaterally in a second one (Fig. 7). The deformity is produced by the anterior rotation of the lateral wall, which enlarges the orbit and thus predisposes to enophthalmus. In addition, the dead space may be filled with periocular fat that is inadvertently released, and the tendency of the globe to retract after release of the medial and canthal cheek ligaments may be increased. These complications may be prevented by the preservation of a frontal crown, recession of the medial orbital wall, and closure of residual skeletal defects.

PTOSIS
Transient ptosis is a frequent finding after the reduction of orbital hypertelorism. It is probably caused by the lateral pull that results from the dislocation of the orbital wall. This pull lengthens the palpebral fissure and, by doing so, produces a caudal force that counteracts the levator function. This kind of ptosis usually disappears after a short period. Permanent ptosis, however, may also occur.

Munro and Das\textsuperscript{8} consider lateral ptosis to be a frequent problem, especially in major cases. In a retrospective study of 31 patients with orbital hypertelorism, Choy et al.\textsuperscript{26} found 6 patients with bilateral postoperative ptosis who did not have
this deformity preoperatively. In our series there was one patient in whom bilateral ptosis became more severe following the operation and a second patient with ptosis resulting from the operation (Fig. 7). Choy et al. suggest that postoperative ptosis may be caused by mechanical damage to the levator muscle or to Whitnall’s ligament. We find it difficult to accept this theory since the periorbit is usually not entered. In addition, it should be noted that ptosis was bilateral in the six patients reported by Choy et al. This latter observation almost automatically excluded direct trauma as a possible cause and suggested a different mechanism. One possibility is that the anterior projection of the orbital frame causes the upper eyelid to lose its global support. This phenomenon can be observed in the patient with the pseudoptosis and enophthalmos shown in Figure 7. Caudal dislocation of the lateral part of the upper eyelid following disinsertion of the lateral canthus may provide a second explanation.

**SHORTENING OF THE PALPEBRAL FISSURE**

In one of our patients (Fig. 8.), reduction of the interorbital distance was associated with shortening of the palpebral fissure. The reason for this is not clear. Loss of balance between levator and orbicularis action resulting from release of the lateral canthus may have been the cause.

**TEMPORAL DEPRESSION**

The occurrence of this deformity after orbital transposition has been well recognized in recent years. Anterior rotation of the lateral wall will enlarge the temporal fossa and create a depression. An hourglass deformity may be prevented, however, by advancing the temporal muscle as a whole or by advancing part of it after section of the aponeurosis.

**ASSOCIATED MALFORMATIONS**

At the time of their formation (7 mm C.R.L.), the optic cups are widely apart, but this situation changes rapidly. The nasal capsule develops in a fronto-caudal direction, and 3 weeks later, at the 28 mm C.R.L. stage, the interorbital distance has already been reduced to normal. Hypertelorism therefore is due to a developmental arrest occurring between the fifth and eighth weeks of gestation. It may be associated with a cleft or with craniosynostosis, but it is not due to these malformations. Since the arrest occurs during a critical period of development, it need hardly surprise us.
that hypertelorism is frequently found in combination with a variety of other malformations.

**THE FOREHEAD**

Resection of a widow's peak and other ectopic patches of hair, correction of irregularities of the eyebrows, and reduction of the intereyebrow distance may be indicated to improve the result. In general, these procedures are simple and straightforward.

**THE ORBIT**

*Abnormal Orientation*

Anteroposterior divergence of the orbit may be associated with caudocranial divergence. In these patients, the distance between the orbits in the upper part of the interorbital space is greater than in the lower part. The maxilla may show distinct sagittal arching and the nose is bifid. Correction of this caudocranial divergence can be obtained by rotation of the orbit around an anteroposterior axis, while it is moved to the midline, thus allowing for greater reduction of the interorbital distance in the upper part of the interorbital space than in the lower part.

Application of this principle by combining rotation of each orbit in continuity with the corresponding maxillary half in two of our patients made it possible to correct both the orbital divergence and the sagittal arching of the maxilla by a medial fasciotomy (Fig. 9). This procedure also allows one to combine the intra- or extracranial correction of hypertelorism with advancement of the midface in patients with craniofacial synostosis.

*Abnormal Configuration*

The fact that orbits differ widely in size and shape may have an important bearing on the result to be obtained, and severe abnormalities should therefore be corrected. When the orbit is too small, expansion is required. In a young girl with microorbitism and hypertelorism, correction was obtained in one operation by dislocation of the medial wall of the microorbit in combination with medial transposition of the contralateral normal orbit (Fig. 10). Bulging of the orbital roof in combination with an increase of the interorbital distance, although not necessarily of the intercanthal distance, may also need correction. This can be achieved by replacement of the protruding convexity with a concave bone graft or...
Correction of caudocranial divergence of orbits and sagittal arching of the maxilla by medial faciotomy.

In a child with Apert's syndrome and severe bulging of both the orbital roof and the temporal fossa, the use of this technique was much facilitated by removal of the wall of the middle cranial fossa, providing free access to the lateral wall of the orbit (Fig. 11) and at the same time creating an excellent opportunity to correct the temporal malformation.

The Canthi

Medial canthal dystopia after reduction of the interorbital distance is an indication for disinsertion of the ligament and canthopexy (Fig. 12) in a better position. The technique consists of boring a hole at the site of insertion of the canthal tendon with a diameter sufficiently wide to allow the tendon to enter it as deeply as possible. After identification of the tendon, it is secured with a...
monofilament Ethilon 3-0 suture, leaving both ends long. Then, parallel to and just below the bridge of the nose, two holes are bored on the contralateral side. The direction of the canals is such that the tip of the burr becomes visible in the hole that was previously made. Through these canals two hollow needles are passed, and these serve as guides for the ends of the sutures. The needles are then withdrawn and the procedure is concluded with the tying of a firm knot. Lateral canthal dystopia indicating correction was not observed in this group of patients.

THE EYELIDS

In our series, there were four patients with preoperative ptosis involving six eyelids. In one of these, ptosis became worse following the operation. This number is somewhat less than in the series of 31 patients reported by Choy et al. They found 7 patients with preoperative ptosis involving 10 lids. One of our patients was born with a coloboma of the upper eyelid, but this had been corrected before we first saw the patient.

THE EYES

Malformations of the eye were found in two of our patients. In one patient with right-sided microphthalmia, significant improvement could be achieved by the use of an episcleral prosthesis. In the second patient with unilateral anophthalmia and microorbitism (Fig. 10), the socket lining was gradually expanded after preliminary addition of mucosa. The appearance of the contralateral eye with minimal vision cannot be improved.

THE EYE MUSCLES

Orbital hypertelorism often goes hand in hand with strabismus. Usually there are abnormalities in the horizontal plane, so the exotropic form is found more frequently than the esotropic. Reducing the interorbital distance is generally followed by a reduction of the exotropic form and may even transform it into the esotropic variety. In our series, the remaining exotropic or esotropic strabismus has never been so serious that the postoperative appearance was dominated by it. However, malposition of the eye was especially noticeable in the patient with congenital shortening of the superior rectus muscle (Fig. 7). Choy et al. are of the opinion that the motility disturbance of the eye muscles in patients with orbital hypertelorism is caused by the abnormal anatomic relations in these patients. They apparently base their supposition on the fact that the extra-
ocular muscles have developed normally. Experience gained from the patient with motility disturbances in the vertical plane seems to indicate that dysfunction may also be caused by congenital malformations of the extraocular muscles.

THE NASOLACRIMAL APPARATUS

Whitaker et al. have emphasized that hypertelorism is frequently associated with abnormalities of the nasolacrimal apparatus. Considering the development of this structure by the merging of the lateronasal and maxillary processes, the time at which this process is completed (less than 17 mm C.R.L.), and the closeness of the nasoethmoidal and nasolacrimal structures, this is hardly surprising. In our patients the nasolacrimal apparatus has not been examined routinely.

THE NOSE

Shortness of the nose is a frequent problem that is usually overcome by wide dissection of skin and mucosa and implantation of an iliac bone graft to the nasal bridge. Such a bone graft is an absolute must in terms of aesthetic appearance following correction for hypertelorism. It restores the nasal contour, stretches the skin, and may help to prevent the formation of epicanthal folds resulting from relaxation of skin over the glabella and medial canthal region. It also suggests a decrease in the distance between the eyes. Tessier reconstructs the nasal bridge with a bone graft that is anchored to the frontal and nasal bones. If necessary, a columella strut is added. We have occasionally used a thin, keel-shaped bone graft that is inserted deep between the two nasal halves in the space created by the resected part of the septum. This graft is covered with two smaller grafts (split-rib) simulating the nasal bones.

In bifid noses and in nasal clefting, additional skin is always required. It is tempting to correct these malformations by a simple rearrangement of the available surplus of skin in the nose or in the glabellar area. Unfortunately, it is difficult to predict the behavior of scars running across the bridge of the nose. They may be scarcely visible or quite conspicuous. It is also tempting to preserve the skin of the intact ala, and in fact, this may even be necessary, as is demonstrated in the patient shown in Figure 10 with ectopic patches of hair dispersed over the forehead. However, texture and color of a forehead flap and that of the alar skin may differ widely, again arresting.

Fig. 13. (Left) One-year-old child with extreme hypertelorism and severe monolateral clefting of the nose. (Right) After reduction of the interorbital distance and reduction of the nose using frontonasal surplus of skin. Owing to excessive soft-tissue traction, some canthal drift has occurred on the right side.
our attention. We have therefore come to the conclusion that it may be wiser to remove the skin of the ala together with the skin covering the nasal tip or the groove between two nasal halves and close the resulting defects by economic use of the available nasal and frontal skin. This approach allows us to obtain a new skin cover with the final scars on the lateral aspect of the nose and in the alar rim, where they are least visible. 29 Recent experience with two patients, one of whom had a monolateral deformity (Fig. 13) and the other of whom had a bilateral deformity (Fig. 14), supports this view.

**SUMMARY**

The importance of skeletal reduction of the interorbital distance in the treatment of patients with teleorbitism is now well recognized. In spite of this, results of surgery are not always as good as one would hope. For this there are two reasons: (1) reduction of the interorbital distance may be followed by deformities such as canthal drift, enophthalmus, pseudoptosis, and so forth; and (2) hypertelorism is frequently associated with a variety of other malformations that become more conspicuous after reduction of the interorbital distance. In this paper attention is focused on the mechanisms responsible for the appearance of new stigmata, on their prevention, and also on the treatment of the associated malformations.

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