David Greig, physician and conservator of the Edinburgh Museum, coined the term hypertelorism, erroneously thinking it to be a specific syndrome. Tessier, however, was the first to stress the difference between ocular hypertelorism (teleorbitism), characterized by an increased divergence of the orbital axis from the midsagittal line, and hypertelorism, caused by craniosynostosis. Indeed, an abnormally wide interorbital distance is not automatically associated with such a divergence.

If the malformation and its correction are to be matched, distinction must be made between orbital hypertelorism, in which there is true lateralization of the orbits, and interorbital hypertelorism, in which no lateralization is observed. The distance between the dacrya (IOD - intraorbital distance), the distance between the ecctocanthia, and the angle between the greater wings of the sphenoid are the criteria by which assessment can be made (see chapter 3). There is orbital hypertelorism or teleorbitism when both these distances are increased.

Interorbital hypertelorism (pseudohypertelorism according to Converse) is present when the IOD is increased and the distance between the ecctocanthi is normal. As the angle between the greater wings of the sphenoid increases, the cranial width measured at the pteria as well as the IOD should also increase. It is therefore not surprising that a relationship between the IOD and the cranial width has been established in all types of teleorbitism.

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°.

**PATHOMORPHOLOGY**

Teleorbitism may be the only sign of a developmental abnormality, but more often it is part of a syndrome. At the time of their formation (7mm crown-rump length - CRL) the optic cups are wide apart, but this situation changes rapidly. The nasal capsule develops in a frontocaudal direction and 3 weeks later at the 28m CRL stage interorbital distance has already been reduced to normal.

Persistent divergence of the orbital axes must therefore be due to an earlier developmental arrest. Several mechanisms may be envisaged:

- Deficient growth affecting the brain and the eyes.
- Deficient closure of the rostral neuropore, resulting in the formation of frontoethmoidal encephalocoeles and the prevention of movement of the eyes towards the midline.
- Deficient differentiation of the nasal capsule, causing the frontonasal sphenoid complex to freeze in its fetal form.
- Deficient ossification of the sphenoid and frontal bones, resulting in abnormalities in the position and configuration of the orbits.
- Premature fusion of the sphenofrontal suture.
- The compensatory accommodation of the cranial base to the growing brain, resulting in downward deflection of the cribriform plate.

Costaras et al. have suggested that distinction should be made between a static and a dynamic form of arrest. The first, observed in facial clefting, represents a singular event. The second, seen in craniosynostosis, is an ongoing process.

A singular event disturbing growth in a complex area at a critical stage must have an important adverse effect on normal development. Testimony to this is the wide spectrum of anomalies that has been observed in combination with teleorbitism.

These anomalies may involve:

- The interorbital area – frontoethmoidal encephalocoeles or lipomas (5.1).
- The orbit – abnormal orientation, configuration, and occasionally dimension (micro-orbitism).
- The nose – various types of clefting.
- The maxilla – sagittal arching.
- The forehead – widow’s peaks.
- The eyelid – colobomata and ptosis.
- The eye muscles – strabismus.
- The nasolacrimal apparatus.

Each of these abnormalities may require correction and therefore add to the complexity of treatment.

By contrast, an ongoing process observed in patients with craniosynostosis will, in addition, have a significant effect on the morphology of the interorbital area and the orbit itself. Anomalies mainly involve widening of the frontal processes of the maxilla and of the nasal bones, widening of the ethmoidal labyrinth, lowering of the cribriform plate with inward bulging of the orbital roof, and upward slanting of the lesser wings of the sphenoid. Teleorbitism in these patients is therefore always associated with malorbitism.
CORRECTION

The correction of teleorbitism is always preceded by the removal of interorbital obstacles. This may involve the resection of bone in the glabellar area, brain tissue in patients with encephaloceles, ethmoidal cells, the perpendicular plate of the ethmoid bone, the septum, and the turbinates. The integrity of the cribiform plate and the nasal mucosa should of course be respected. Following exenteration of the interorbital area, the orbital osteotomies are performed.

Teleorbitism is not corrected by linear translation of both orbits. Rather, a more complex movement is required because the abnormal orientation involves anteroposterior divergence of the orbit in a horizontal plane and caudocranial divergence in a frontal plane.

Correction of the first abnormality is achieved by rotation of the orbit around a caudocranial axis in the centre of the orbit (5.2), while correction of the second is obtained by rotation of the orbitomaxillary unit around an anteroposterior axis through the centre of the maxilla (see 5.2).

These objectives can be achieved by the use of a classical orbital osteotomy or a more extensive orbitomaxillary osteotomy (facial split). The choice between the two procedures is dictated by the absence or presence of maxillary arching.

Teleorbitism in craniosynostosis mainly involves the upper part of the orbit. In less severe cases and in young children, the use of an orbitofrontonasal osteotomy will permit narrowing of the interorbital distance and remodelling of the orbital roof by changing the medial convexity into a concavity. However, more extensive orbital or orbitomaxillary osteotomies may also be indicated.

COMPLICATIONS

Enophthalmus and canthal drift are the most important deformities that may remain after correction of orbital dystopia.

Enophthalmus is caused by excessive projection of the lateral orbital wall and can be avoided if rotation of the orbit around a central axis is performed with minimal displacement of the medial and lateral orbital walls (5.3).

Canthal drift is another problem that is occasionally observed. Several explanations for it have been offered, such as:
• Inadequate interorbital exenteration and fixation.
• Excessive, laterally directed contracting forces.
• Apposition of bone and scar tissue in the medial canthal area.

5.1 Severe teleorbitism due to lipoma in interorbital area.
Complications

5.2 Correction of teleorbitism: 

- a by rotation of the orbit around caudocranial axis;  
- b by rotation of the orbit around an anteroposterior axis through the maxilla.

5.3 Prevention of enophthalmus in correction of teleorbitism: 

- a original preoperative situation in a 6-month-old patient with internasal dysplasia associated with a midline lipoma;  
- b the rotation axis is placed in the medial wall, resulting in anterior displacement of the lateral wall associated with orbital volume increase;  
- c the rotation axis is placed in the lateral wall, resulting in posterior displacement of the medial wall that requires partial resection and that is associated with the decrease in orbital volume;  
- d the rotation axis is placed in the centre of the orbit, resulting in minimum displacement of the medial and lateral walls and no change in orbital volume.  

(Re redrawn with permission from van der Meulen JC, Zonneveld FW. Discussion of enophthalmos following orbital transposition for craniofacial malformations. Plast Reconstr Surg 1993; 91.1:423-428.)
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