TRIGONOCEPHALY

R Roddi

Trigonocephaly is the term used to describe the craniofacial malformation caused by the premature closure of the metopic suture. It was first used by Welcker in 1862.

At the level of the cranial vault, the keel shape of the forehead is characteristic (6.1). It is especially apparent in axial views, providing a contrast with the normal biparietal diameter. The mediofrontal angulation extends back as far as the anterior fontanel in the form of a median ridge. This ridge, sometimes visible and palpable, corresponds to the fused metopic suture.

The frontal area becomes triangular, giving the face of the patient its typical clinical feature. The upper part of the face is greatly affected by the underdevelopment of the frontal eminences. As a result in patients with involvement of the lower portion of the suture, the orbital arches seem to project and the eyes appear abnormally close because of a varying degree of hypotelorism.

In almost all cases, the craniostenosis is the primary and single event. Most associated malformations (observed in 9% of cases according to Shillito and Matson) appear to be incidental. In spite of this, mental retardation is occasionally reported even in the absence of a raised intracranial pressure. Infants with severe microcephaly may present with trigonocephaly which is probably due to lack of brainpush and early closure of multiple sutures. A combination of trigonocephaly, hypotelorism, and atrhinencephaly, first reported by Kundrat in 1882 may also be observed.

The radiological findings merely reflect the morphological abnormalities. The axial views bring out the triangular shape of the frontal region. The fused metopic suture is seen as a dense rectilinear ridge. It is important to differentiate true metopic synostosis with trigonocephaly from a prominent metopic ridge without trigonocephaly and orbital involvement. The latter condition, quite common and often familial, does not require an operation.

Narrowing of the ethmoidal labyrinth is associated with verticalization of the medial orbital wall, producing a typical form of malorbitism. The orbital shape is ovoid sloping downward and out-
ward in contrast with the 'Mephistophelian' deformity of brachycephaly (6.2).

**Surgical Technique**

Exposure of the skull is performed through a bicoronal incision. Dissection is initially carried in a subgaleal layer, and then, 2 cm above the eyebrows, continued in a subperiosteal plane, allowing dissection of the upper, lateral, and inferior orbital margins and of the zygomatic arch. A frontobiparietal bone flap is elevated, and a supraorbital Tessier osteotomy is performed. The metopic suture is resected towards and up to the frontonasal suture. Frontal remodelling may be achieved in several ways – by lateral translation or rotation of the two frontal halves, or, as was suggested by Marchac in 1978, by transposition of a parietal segment with a curvature somewhat different from that of the forehead. This last procedure is the one most frequently utilized by us.

This procedure makes it possible to achieve the transformation of the anterior angulation into a more harmonious curvature together with restoration of the orbital contour. Correction of the V-shaped angulation is obtained by differential propulsion of the fronto-orbital band (6.3). Splitting of the internal cortex by multiple cuts along its length permits forward displacement of the lateral extremities of the band. Downward displacement of these parts is made possible by the formation of a greenstick fracture in the upper part of the midline. The median defect thus formed must be closed with a triangular bone graft. Downward displacement may produce a discrepancy between the vertical dimensions of the lateral and medial orbital wall, as the latter remains too high.

This inadequacy may be corrected by removing a small part of the medial wall and by lowering the fronto-orbitonasal band as a whole.

Aware of this problem, Marchac advises the advancement of the lateral orbital walls in conjunction with the bar. The resulting triangular defect is closed with a graft. Sometimes it is necessary to reinforce the superior orbital band with a bone graft taken from the skull behind the proximal line of the craniotomy, at the level of the coronal suture. This graft, fixed with titanium wire or titanium screws at the supraorbital rims, reinforces the fragile band and permits a good stabilization. The resultant reshaping of the cranio-orbital region is generally very good, although it sometimes falls short of expectations owing to the residual transverse shortness of the forehead, which is accentuated by the flat lateral parts and the presence of hypotelorism.

Gruss has solved this problem by including the lateral orbit down to the junction with the inferior orbital rim and by leaving a wide and long temporal extension on the frontal bandeau. When the bandeau is expanded centrally and then bent at the lateral orbital region, the temporal extension will widen the bitemporal width and prevent hollowing.

External canthopexy completes the operation and in order to fix the periosteal flaps and the temporal muscles the human fibrin sealing Tissucol (Immuno, Vienna) is used. The skin is sutured in two layers, after positioning of two mini-aspirative subcutaneous drains.

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6.2 Malorbitism in trigonocephaly: 
- a causative mechanism of malorbitism; 
- b malorbitism due to trigonocephaly in a patient with Pfeiffer's syndrome.

6.3 Correction of trigonocephaly.
Plagiocephaly

R Roddi

Plagiocephaly is the term used to refer to an asymmetric malformation secondary to the fusion of one half of the coronal suture. This term, coined by Virchow in 1851, means literally cranial asymmetry (from the Greek: plágio - oblique or aslant, and kephalé - head). Unfortunately the word has been applied to dysmorphic skulls that look similar but have different aetiologies. The word plagiocephaly is in general used to define a unilateral coronal synostosis, affecting the sphenotemporal suture (Seeger, 1971). Many authors employ the same term to describe head asymmetry when there is no evidence of sutural synostosis. Marsh and Vannier define this abnormal morphology as 'plagiocephaly without synostosis'; others (see Clarren) call it 'deformational plagiocephaly'.

Such variable use of the term plagiocephaly perpetuates confusion regarding the aetiology and pathogenesis, and the diagnosis and treatment of this anomaly. It is therefore important to have a clinical categorization and definition of plagiocephaly.

Simple plagiocephaly (category I)

Simple plagiocephaly (category I) covers unilateral synostosis of the ipsilateral coronal suture as well as the frontoethmoidal suture.

Clinically these patients are characterized by frontal flattening on the affected side (6.4). Bulging of the ipsilateral temporal bone is also seen. Palpation sometimes reveals a ridge corresponding to the obliterated suture. Often it is possible to note a compensatory expansion on the opposite side with frontal bossing and abnormal protrusion of the supraorbital ridge or bulging in the parietal region. At the facial level, there is a typical retrusion of the orbital arch on the affected side with steepening of the lesser wing of the sphenoid and ovalization of the orbital shape in a superolateral direction. This produces the typical 'harlequin' shape (6.5).

6.4 Simple plagiocephaly:

a preoperative view; b postoperative view.

6.5 Malorbitism in plagiocephaly:

a causative mechanism; b 3-d image showing unilateral teleorbitism (left), superomedial flattening, and superolateral retrusion of the left orbit.
The eyebrow and eyeball appear raised, and strabismus, usually convergent, may be observed in about 25–35% of patients. The nose itself, the maxilla and dentofacial midline are deviated to the contralateral side due to expansion of the middle fossa. Anterior enlargement pushes the zygoma forward and causes deviation of the face to the opposite side. Unilateral teleorbitism associated with malorbitism due to interorbital hyperelorism is quite common.

The ear on the affected side is sited further forward and downward than the opposite ear. The hypoplasia sometimes extends to the middle third of the face and particularly to the malar bone. The maxillary bone may also be affected with abnormalities of dental apposition (Rougerie and Tessier, 1972).

**Simple plagiocephaly (category II)**

Simple plagiocephaly (category II) includes the clinical features of an asymmetrical head that result from premature fusion of the contralateral lambdoidal suture. In these patients there is bulging of the forehead on the opposite side of the lambdoidal synostosis due to compensatory growth within the contralateral coronal suture. This clinical situation has been defined as occipital plagiocephaly; we take exception to this term because of the frontofacial compensatory distortion.

**Complex plagiocephaly (category III)**

Complex plagiocephaly (category III) includes the type of plagiocephaly characterized by the association of a unilateral synostosis of the coronal or lambdoidal suture and the synostosis of at least one other suture of the cranial vault.

**Metabolic or dysmetabolic plagiocephaly (category IV)**

Metabolic or dysmetabolic plagiocephaly (category IV) defines a craniosynostosis that is due to vitamin D deficiency, polycythemia vera, thalassaemia, sickle cell anaemia, or mucopolysaccharidosis.

**Pseudoplagiocephaly (category V)**

Particular attention must be paid finally to the asymmetrical head without synostosis of the cranial suture. This situation, which we call pseudoplagiocephaly (category V), is secondary to birth injuries or to postural abnormalities such as congenital torticollis or postnatal molding of the skull. The incidence in N. America of the latter deformity has increased dramatically since it was recommended that infants lie on their back because of the risk of SIDS (sudden infant death syndrome). However, hypoplasia of one cerebral hemisphere or hyperplasia in abnormalities such as arachnoid cysts, Sturge-Weber syndrome, neonatal hydrocephalus, and chronic subdural effusion in the neonate can also be responsible for frontal asymmetries. The radiographic features, in particular the disappearance or sclerosis of a coronal hemisuture, are thus of great importance in the correct diagnosis in these patients.

**INCIDENCE**

The incidence of craniosynostosis is estimated to be one per 1000 to one per 2500 live births. Between 18% and 29% have coronal synostosis, of which 50% are unilateral (Shellto, 1968; Tessier, 1971; Hunter, 1977). The prevalence of unilateral coronal synostosis is estimated to be one per 10 000 live births. Compensatory frontal plagiocephaly (category II) may be under-reported because posterior cranial asymmetry is not as obvious as frontal asymmetry.

**SURGICAL CORRECTION**

Surgical correction (6.6) is essentially based on:

- Resection of the fused suture to decompress the brain and allow its expansion.
- Prevention of premature reossification by alteration or reduction of the osteogenic potential of the external dural layer.
- Restoration of a normal morphology by cranial and facial remodelling. Instant correction may be achieved by the mobilization and transposition of bony segments. Long-term improvement may be obtained by dynamic forces of unrestricted cerebral growth which guarantees the normal development.

Surgery may be performed from the age of 3 months onwards when it will draw maximum benefit from the cerebral expansion that increases the cerebral volume to 75% of its adult volume at the end of the first year. Early operation is therefore advocated in all cases, and appropriate measures must be taken to prevent rapid reossification of the bony defect, such as a wide resection of the synostosed suture. Synostosis of a hemilambdoidal suture, in the absence of important compensational fronto-orbital malformations, is not treated in the absence of a clinical finding of intracranial hypertension.
BRACHYCEPHALY

Complex forms (category III) and dysmetabolic forms (category IV) require complementary correction of the skull and the facial skeleton. Pseudoplaigiocephaly is not treated surgically in most cases. Simple plagiocephaly (category I) is therefore the main indication for surgery.

Surgical technique
Three fields of surgical action may be distinguished: the cranial vault, the fronto-orbitonasal junction, and the face.

The cranial vault
Exposure of the skull is performed through a bicoronal incision placed behind the hairline and extended to the supraauricular region. Dissection is initially carried out in a subgaleal layer, and then, 2 cm above the eyebrows, it continues in a subperiosteal plane, allowing dissection of the upper, lateral, and inferior orbital margins and of the zygomatic arch. Two periosteal flaps are dissected, one anteriorly and one posteriorly.

Correction of the frontal region is performed with the 'floating forehead' technique of Marchac. This technique consists of a combination of a 2 cm advancement of the frontal bone fixed to the face with titanium plates, and coronal and orbital craniectomies, thereby dissociating as far as possible the forehead and the face from the base of the cranium and the cranial vault.

Fronto-orbitonasal junction
The orbital anomaly is characterized by retrusion and ovalization of the superolateral quadrant. Prior to the bifrontal craniotomy, a template is used to outline a cranial segment, generally from the non-affected side, to be transposed as the new forehead, selecting the part that will provide the best curvature in two directions.

Correction by advancement of the retracted area on a median axis is performed by an osteotomy of the entire supraorbital bar using a tenon-shaped extension. Multiple vertical osteotomies of the inner cortex are performed, allowing the bone to be bent in a suitable contour. The superior orbital band is reinforced with a bone graft taken from the skull behind the proximal line of the craniotomy, at the level of the coronal suture. This graft, fixed with titanium wire or titanium screws at the supraorbital rims, reinforces the fragile band and permits a good stabilization. The fronto-orbital band, advanced on the affected side, is fixed to the skull in a similar manner. The inward protruding convexity of the upper medial orbital quadrant is corrected by reshaping of the roof.

Facial correction
In order to correct facial deformities, some authors have advocated osteotomies of the roof of the nose and the maxillomalar complex. Justified as this approach may be in adolescents and adults, we consider that it is not indicated for corrections performed in the first year of life, where we can obtain a normalization of the facial shape following normal growth.

Reinsertion of the temporal muscle is extremely important. It will serve to maintain the anteroposterior enlargement of the temporal fossa and to promote mandibular symmetry.

BRACHYCEPHALY

JC van der Meulen

Brachycephaly is most commonly seen as a single anomaly caused by bilateral coronal synostosis. It may, however, also be part of a syndrome involving a complex system of synchondroses (sphenopapillary) and sutures (sphenofrontal, sphenethmoidal, sphenotemporal, temporozygomatic).

Best known of these syndromes are:
- Craniofrontonasal dysplasia.
- Acrocephalosyndactyly (Type I-IV), e.g. Apert's syndrome, Saethre-Chotzen syndrome, etc.
- Craniofacial dysostosis (Crouzon's syndrome).

Discussion of malorbitism in patients with brachycephaly will therefore be limited to these entities.

Brachycephaly simplex
Brachycephaly simplex (6.7) is primarily characterized by frontal retrusion, bitemporal protrusion, and typical harlequin-shaped orbits. There are no acral anomalies.

6.7 Brachycephaly simplex: a frontal view of patient; b 3-D image of the orbits showing superomedial flattening and superolateral retrusion.
Craniofrontonasal dysplasia
Craniofrontonasal dysplasia (6.8) is characterized by frontal retrusion (a widow’s peak may be observed), teleorbitism, nasal shortening (bifidity may be present) and maxillary arching. Acral anomalies are frequently associated. They can involve the fingers and toes (syndactyly or clinodactyly), the clavicles, and the scapulae.

Apert’s syndrome (6.9)
Acrocephalosyndactyly has frontal retrusion and teleorbitism in common with craniofrontonasal dysplasia. The coronal sutures are prematurely closed and a widely patent midline fontanelle extending from the glabella posteriorly is commonly observed. In addition to these stigmata, the syndrome features severe bitemporal protrusion, anti-mongoloid slanting of the palpebral fissures, ptosis of the
upper eyelids, a bulbous nose, maxillary retrusion associated with arching of the alveolar process, and a typical cross-bow appearance of the upper lip.

Severe exophthalmus may be observed. Acral anomalies involving fusion of digits and toes are pathognomonic.

Crouzon's syndrome (6.10)
The craniofacial features of this syndrome resemble those of Apert's syndrome only to a certain degree. Crouzon's syndrome is characterised by multiple suture synostoses, fused synchondroses and digital markings. Radiological evidence of this, however, only becomes manifest after 1–2 years. Frontal and maxillary retrusion are the most striking stigmata. Bitemporal protrusion, anti-mongoloid slanting of the palpebral fissures, and teleorbitism may be observed, but are less prominent. Acral anomalies are never present.

Discussion
Normally the lateral walls of the orbit form an angle of 90°. The orbital axis and the plane of the superior and inferior rectus muscle, which correspond with the line of fixation when the eyes are in the primary position (i.e. looking straight ahead) (6.11) show an angle of approximately 23° with the sagittal axis of the skull.

Proper alignment of the eyes relies on several conditions involving normal vision of the eyes, normal position of the eyes, and normal function of the muscles. Abnormal vision of the eye due to optic atrophy, abnormal position of the eye related to malorbitism and abnormal function of the muscles due to a disturbance in the relationship between origin and insertion of the muscles or to deficiencies in their structure may be responsible for misalignment and produce exophoria, esophoria, hyperphoria or hypophoria.

Exophoria is frequently seen in teleorbitism when the angle between the orbital axis and the sagittal axis of the skull may increase from normal (23°) to 40°. A combination of exophoria in upward gaze and esophoria in downward gaze (V-pattern) may be observed in teleorbitism associated with exorotation of the orbit, anti-mongoloid slanting of the palpebral fissures, and pseudoeuctopia of the macula. This pattern is thought to be caused by a discrepancy between the angles formed by the superior and inferior oblique muscle with the axis of the eye. Normally the planes of the superior and inferior oblique muscles form an angle of approximately 51° with the sagittal axis of the skull.

Diamond et al. reported extraocular anomalies (primarily of superior and inferior rectus muscle) in 42% of patients with craniofacial dysostosis. Abnormalities in the horizontal position of the orbit and exorotation produce disturbances in fusion of the eyesight less easily than abnormalities in the vertical position.

Exophoria can be improved by correction of teleorbitism.
In contrast esophoria, hyperphoria and hypophoria are not affected by surgery; perhaps because they are due to primary structural or myoneural deficiencies. Normally also, the length of orbital roof and floor is such that the globe is adequately protected. The height of the orbit represented by the rim tangent in sagittal CTs has a mean of 2.4 cm. and the corneal plane is level with the tangent. Proper closure of the eyes will depend on a normal length of the orbital roof, floor, and rim tangent.

Abnormalities in length of the orbital roof, floor and rim tangent are due to either frontal or maxillary retrusion. The presence of exophthalmos and a shallow orbit, however, do not always mean that the orbital volume is reduced. A reduction in length of the floor and the roof may in fact be compensated for by an increase in height of the orbit, explaining our observation of exophthalmus in an Apert patient with a normal orbital volume.

**SURGICAL CORRECTION**

In patients with simple brachycephaly correction is relatively easy. Adequate correction of the frontal retrusion is obtained by forward and downward advancement of the frontonasal base, using Marchac's floating forehead technique.

In patients with complex brachycephaly, solutions to the problem are not so straightforward, and they may even be conflicting. Priorities in the treatment of these patients are first of all dictated
by the severity of the composing anomalies. Brain compression, ocular exposure and respiratory distress may require early frontal or frontofacial (monobloc) advancement, tempting the craniofacial surgeon to treat the orbital anomalies simultaneously. Posnick, however, has shown that a standard procedure, carried out on young children, has little long-term effect.

In general, we prefer to perform an intermediate correction of malorbitism and teleorbitism at the age of four. A late repair is usually indicated for any remaining maxillary or maxillofacial anomalies. Correction of malorbitism and teleorbitism in craniofrontonasal dysplasia requires frontal advancement, reduction of the interorbital distance, and orbital roof remodelling.

A facial bipartition procedure will achieve the first two objectives only if the orbital segment of the hemifacial is advanced to some extent. The correction of malorbitism and teleorbitism in Apert's syndrome requires frontal advancement, maxillary advancement, reduction of the interorbital distance, reduction of the orbital height, which may be significantly increased, and orbital roof remodelling. A facial bipartition procedure will achieve the first three objectives but it does not reduce the height of the orbit. As a result it will produce enophthalmus in Apert patients who have a normal orbital volume. To prevent this, the orbital volume increase required for the correction of the anomaly would have to be measured preoperatively. If the volume is normal to start with, its shape must be corrected while the volume is maintained. The circumference of the orbit at the site of the osteotomy must be known as the radius becomes larger if the osteotomy is made more anteriorly. Differential correction of the forehead, orbital, and maxillary anomalies should be considered where a monobloc procedure was performed previously.

**ENCEPHALOCELE**

*R Roddi  JC van der Meulen*

**DEFINITION**

Encephalocele has been defined as a protrusion of cranial content beyond the normal confines of the skull (French, 1982). By this definition the term encephalocele includes meningocele (herniation of meninges and cerebrospinal fluid), encephalomeningocele (herniation of brain tissue and meninges), and hydroencephalo-meningocele (herniation of a portion of the ventricle, brain tissue, and meninges). The first observation of this anomaly of the central nerve system seems to have been reported by Forestus in 1590 and later in 1749 by Corvinus. Early theories about its nature were proposed by Spring in 1853.

Patients with this anomaly demonstrate swellings of varying size in the glabellar region. These swellings may be sessile or pedunculated. On palpation, the mass may be solid and firm or soft and cystic. The contents of the coele mostly consists of glial tissue, often infiltrate with fibrous trabeculae (David *et al.*, 1984). The ventricular system may be involved in craniofacial clefting.

The skin over the mass may be thin and shiny or thick and wrinkled. Hyperpigmentation or hypertrichosis may be noted. Telecanthus is usually pronounced (6.12).

Downward dislocation of the insertion of the medial canthal tendon is frequently observed, producing a mongoloid slant, but the lateral orbital walls are usually in a normal position (6.13).
Elongation of the nose is a cardinal feature of this condition. The anomaly may be associated with abnormalities involving the brain, anterior angulation of the optic nerves and pituitary stalk, and partial or complete occlusion of the foramen of Monro, which may result in hydrocephalus, ventricular dilatation, agenesis of the corpus callosum, cortical atrophy, mental retardation, or generalized convulsions.

Visual acuity may be decreased. Strabismus and lacrimal obstructions, resulting in epiphora and dacryocystitis have been reported.

**Epidemiology**

The incidence of this anomaly shows wide variations. It is quite common in Thailand, with an incidence of one per 5000 or 6000 live births (Charoensris and Suwanwela, 1974); relatively common in some African countries, including Ethiopia, Morocco (Acquavita et al., 1972), Nigeria (Onuigbo, 1977), Zimbabwe (Kalanga et al., 1990), and South Africa (Lipschitz et al., 1969); and extremely rare in other parts of the world. A genetic base has never been established.

These references, however, do not provide a clear picture of the incidence of encephaloceles in general. An inconsistent terminology has been used, which makes the comparisons between the various forms of encephaloceles less reliable.

**Pathogenesis**

Frontoethmoidal encephaloceles are probably due to a developmental disturbance in the separation of neural and surface ectoderm, occurring after closure of the rostral neuropore in the fourth week of gestation. Persistence of the strong connection between these layers at the site neuroporoticus seems to be responsible for the secondary mesodermal skull defects and consequently for the herniation of encephaloceles before bone centres have developed in the neurocranium and ossification proceeds.

Several important phenomena can be explained by this theory. First, the association of encephaloceles and facial clefting. This combination is always characterized by severe orbital hypertelorism or teleorbitism and is occasionally incompatible with life (split face). Teleorbitism has its origin in a period before the 28 mm CRL stage, before medialization of the orbits is complete.

Secondly, the variation between the different forms of encephaloceles that are characterized by interorbital hypertelorism or malorbitism. This anomaly must have its origin in a period that follows medialization of the orbits and is commonly associated with a normal distance between the ectocanthi.

The variations in morphology seem to be dependent on such factors as the extent of non-separation along the lines of fusion of the neural folds, the site of the developmental arrest, the shape of the resulting mesodermal defect, and the manner in which ossification of the mesoderm adapts to this process by lowering the cribriform plate and nasal septum and by widening the interorbital distance.

**Classification of Encephaloceles**

The classification of congenital anomalies as a tool for better understanding is of little practical value if the surgical implications are neglected. Therefore, in the description of encephaloceles, distinction must be made between encephaloceles associated with teleorbitism and encephaloceles associated with malorbitism. The first category is characterized by an abnormal orientation and position of the orbits. The configuration is usually normal. In the second category the reverse is true. Orientation and position of the orbit are normal. The configuration however is abnormal, producing different types of malorbitism. A study of 3D-images (6.14) shows that an accurate diagnosis can be based on the location of the defect in the cranial base, on the site of the external orifice in the facial skeleton, or on a combination of both. Von Meyers in 1890 has classified encephaloceles on the basis that the cranial end of the defect is always at the junction of the frontal and ethmoidal bones (the foramen caecum), and he distinguishes between different types of frontoethmoidal dysplasia (nasofrontal, nasoethmoidal, naso-orbital).

The second author feels that further distinction between frontonasal and frontoethmoidal dysplasia is justified and has proposed the following classification:

- Frontonasal or supranasal encephalocele.
- Medial frontoethmoidal or nasal encephalocele.
- Lateral frontoethmoidal or orbital encephalocele.
- Ethmoidal or retronasal encephalocele.

**Frontonasal or supranasal encephalocele (6.15)**

The encephalocele leaves the cranial cavity through the fonsitculis frontalis between the developing frontal and nasal bones. The cribiform plate is usually tilted downwards, forming an angle of 45–50° with the orbitomeatal plane. The nasal bones are attached to the inferior margin of the defect.

**Medial frontoethmoidal or nasal encephalocele (6.16)**

The defect at the anterior cranial fossa is situated posterior to the frontonasal suture and anterior to the ethmoid. The mass protrudes between the nasal bones and the nasal cartilage, pushing the septum backwards and downwards. The pyriform aperture is displaced inferiorly. The lateral wall of the tunnel is formed by the intact medial wall of the orbit.

**Lateral frontoethmoidal or orbital encephalocele (6.17)**

The herniation in the cranial base is in the same position as that of the medial variation but the mass protrudes through holes in the medial wall of the orbit, in the frontal process of the maxilla, or in the lacrimal bones, forming a paranasal swelling.

**Ethmoidal or retronasal encephalocele**

The defect in this category is situated at the level of the ethmoidal bone. The cerebral mass remains intranasally and may even extend to the palate.
SURGICAL CORRECTIONS

Computed tomography, including 3D-imaging, provides accurate information concerning the exact site, size and shape of the entry and exit holes in the skeleton.

Accurate measurement of the distance between the ectocanthis allows for distinction between an encephalocele associated with teleorbitism or malorbitism. The treatment of teleorbitism has been discussed in Chapter 5. Correction of malorbitism involves the following steps:

- Frontal bone and herniating encephalocele are first exposed by a combination of coronal and (para)nasal incisions.
- A subperiosteal approach, a frontal access osteotomy, and the removal of a bony rectangle in the glabellar area permit extracranial and intracranial isolation and resection of the coele.
- Regulation of the intracranial pressure and prolonged ventricular drainage may be required in patients with hydrocephalus.
- Following repair of the dura the interorbital distance is reduced by single-quadrant, two-quadrant, or three-quadrant osteotomies (6.18).

The choice between these three osteotomies is dictated by such factors as the dimension of the orbit, the configuration of the orbit, the site or sites of the abnormality, and the level of the cribriform plate. Interorbital hypertelorism caused by encephaloceles is usually treated by mobilization and medialization of a superior skeletal quadrant. Remodelling of the medial orbital wall and closure of the defect in the anterior cranial fossa with a bone graft can, however, be sufficient in some patients with frontonasal and frontoethmoidal encephaloceles. The surgical protocol should be adapted to the specific requirement posed by each case.

Translocation of orbital segments sometimes causes these structures to override the cribriform plate; as a result the transnasal canthopexy becomes transcranial.

Additional bone grafting may be required to close residual defects in the orbit, to obliterate the interorbital dead space or to reconstruct the nasal dorsum.

The operation is completed by the excision and adaptation of skin to the bony frame.
TREACHER COLLINS SYNDROME

R Roddi  JC van der Meulen

The Treacher Collins syndrome or malar dysplasia is an uncommon autosomal dominant hereditary syndrome characterized by deficiency of the malar bone, flattening of the cheeks, anti-mongoloid eyelid malposition, lateral canthal dystopia, and notching or a coloboma of the lateral aspect of the lower eyelid (6.19, 6.20). The coloboma sometimes continues as a groove that may extend towards the angle of the mouth, or more laterally. Dystopic cilia along the edges of this groove, deficiency or absence of the eyelashes (madarosis) medial to the coloboma, and atresia of the lacrimal canaliculi may also be associated. The skeletal disruption seems to be responsible for the forward maxillary projection; malar hypoplasia, obliteration of the frontonasal angle, a beak-like nose, and retrogenia give the patient a typical convex bird-like aspect. In the orbitopalbral region there is often a defect of the inferolateral angle (6.21) and a caudal displacement of the superolateral angle.

The orbits assume an abnormal obliquity and narrowness and the inferior orbital fissure often appears enlarged.

A single temporomasseteric muscle is sometimes seen and various forms of ear malformations may be observed.

The Treacher Collins syndrome occurs in several degrees of severity, ranging from the complete form to the incomplete form, including the abortive form.

In the past, various materials have been used for the correction of the malar bone defect; these materials include cartilage, bone, dermis, and fat, either as autograft or as homograft.

The major disadvantages of alloplastic implants are dislocation, capsular contraction, and extrusion. Bone grafts have too high a resorption. As a result, children are submitted to too many operations with a high iatrogenic morbidity.

6.19 Patient with Treacher Collins syndrome treated by anterior osteoperiosteal flap (deep temporal artery): a preoperative appearance; b postoperative appearance. Secondary correction of left lower eyelid position is indicated. (Reproduced with permission from van der Meulen JCH, Hauben DJ, Birgenhagen-Frenkel DH. The use of a temporal osteo-periosteal flap for the reconstruction of malar hypoplasia in Treacher Collins syndrome. Plastic and Reconstructive Surgery 1984;74(5):687–693.)

6.20 Patient with Treacher Collins syndrome treated by posterior osteoperiosteal flap (superficial temporal artery): a preoperative appearance; b postoperative appearance.
Today, malar reconstruction is frequently performed with the help of a temporal bone flap or a rigidly-fixed full-thickness cranial bone graft. Two varieties of these flaps have been described: one anteriorly with a muscular pedicle vascularized by the deep temporal artery (van der Meulen et al., 1984), and one posteriorly with a galeal pedicle vascularized by the superficial temporal artery (McCarthy, 1984) (6.22, 6.23).

**Operative Technique**

The surgical approach is performed through a bicoronal incision, and dissection is carried downwards, exposing the frontal periosteum, the temporal muscle, the skeletal defect in the malar area, and the orbital contents on each side of the skull. The malar defect is first closed with a composite temporal bone flap or a full-thickness cranial bone graft. This bone is inserted into the infraorbital region and fixed to the maxillary periosteum through a subciliary incision giving the malar region its prominence.

The defective lateral orbital floor and wall are then reconstructed with another bone graft, lifting the eyeball and correcting the anti-mongoloid slant.

In addition, the superolateral orbital rim may be transposed medially to improve the shape of the upper orbit and create a superior orbital ridge.

Correction of the lower eyelid deficiency and of the lateral canthal dystopia is performed by transposition of a musculocutaneous superior palpebral V-flap and by an external canthopexy.
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