EPICANTHUS

Congenital epicanthal folds are a normal racial characteristic in Asia and the Americas. In the western part of the world they are considered an abnormality. Two explanations can be offered.

The first is laxity of skin over the nasal roof. In support of this are the facts that some of the folds disappear before the age of 6 years and that obliteration of the folds is observed by insertion of an implant or by pinching up of the nasal skin.

The second is that the folds are caused by an intrinsic shortage of skin, for which evidence can be found in the facts that different types of epicanthal folds have been distinguished and that correction of these can be obtained by redistribution of skin using one of the various methods that have been advocated (8.1).

8.1 Different techniques for correction of epicanthus.
Unfortunately, some of these procedures produce more scar tissue than others because of difficulty in planning the direction of the incisions and consequently the orientation of the scars. Careful selection and execution of the technique is therefore imperative (8.2, 8.3).

8.2 Correction of epicanthus. a Design of V-Y plasty. b Result of V-Y advancement. c Preoperative appearance. d Postoperative appearance.

8.3 Correction of epicanthus. a Design of double Z-plasty. b Result of Z-plasty procedure. c Preoperative appearance. d Postoperative appearance.
**TELECANTHUS**

An increase in width between the medial canthi as a solitary phenomenon is rare. The anomaly is one of the characteristics of Waardenburg's syndrome and is a dominant feature in blepharophimosis. The normal intercanthal width is almost exactly half of the interpupillary distance, and telecanthus is caused by an abnormal insertion of the medial canthal tendons or by abnormal length of these tendons.

If telecanthus appears to be the only abnormality present, it should be distinguished from teleorbitism by careful measurements of the interorbital distance. Since there are no complicating epicanthal folds, correction is straightforward and can always be achieved by means of a transnasal canthopexy (8.4, 8.5).

**BLEPHAROPHIMOSIS**

Blepharophimosis is a rare congenital anomaly. The word phimosis or stenosis refers to the short and narrow, almost slit-like palpebral fissure or meatus, which forms a sharp contrast with the general flatness of the orbital region. This flatness is due to the absence of distinct convexities formed by the nasal roof and orbital roof, or prominent concavities such as a palpebral fold or canthal fossa. The palpebral aperture usually shows an antimongoloid slant, though occasionally the slant is mongolid.

Epicanthus inversus, telecanthus, and ptosis of the upper eyelid form the triad that is most characteristic of this striking anomaly. However, abnormalities of the lateral canthus and the lower eyelid may also be observed. Closer inspection of the orbital region reveals the signs described below.

**UPPER EYELID**

A palpebral fold is virtually non-existent and ptosis is always present. The dimensions of the eyelid are short in every direction. The skin seems to be somewhat thicker than normal, and hirsutism is occasionally observed.

**THE MEDIAL CANTHUS**

Telecanthus is invariably observed, but the canthal drift is frequently masked by an epicanthal fold, which differs from a simple epicanthus in that its direction is more obliquely oriented. An epicanthus inversus is thus produced, rounding out the commissure, hiding the caruncle, and blending with the medial part of the lower eyelid, where it forms an epiblepharon.

**THE LOWER EYELID**

In contrast with the medial part of the lid, where an epiblepharon may create the impression of congenital entropion, the lateral half is frequently somewhat everted. Indeed the whole lid has the appearance of having shifted somewhat laterally, causing the canaliculi to lengthen, the lacrimal punctum to drift laterally, and the eyelid rim
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to lose contact with the globe. As a result, epiphora may occur. These phenomena can be explained by the uninhibited pull of the orbicularis muscle in a lateral direction, made possible by the medial canthal drift. Shortage of skin, particularly in the lateral part of the lid, and hypoplasia of the orbital septum may also play a role, however.

THE LATERAL CANTHUS

Laxity of the lateral canthus or canthal dystopia is occasionally observed. These contribute to the formation of an antimongoloid slant and to the distortion of the lower lid and shortening of the palpebral fissure.

TREATMENT

The birth of a child with blepharophimosis is a traumatic experience for the parents, because the impression of mental retardation is easily created. It often takes some time before this suspicion disappears. The anomaly is rare and few physicians are familiar with it. Advice to the parents is frequently confusing or controversial.

Once treatment is suggested, the surgeon is compelled to confess that complete success can never be guaranteed, because the causative mechanisms of the different abnormalities are still not sufficiently understood. There seem to be skeletal abnormalities, including hypoplasia of the orbital roof and the lateral orbital wall, but three-dimensional CT reconstruction of the orbit has failed to provide evidence for this hypothesis, and the impression of enophthalmus even seems to contradict it. The soft tissue abnormalities are, however, better understood, making it possible to improve the static and dynamic conditions in the orbital region to some degree.

Although the similarity of patients with this syndrome is striking, particularly when they belong to the same family, important variations in appearance may be observed. The degree in severity of each of the symptoms may differ, creating a wide spectrum of abnormalities, which becomes even more complex when there is disturbance of other oculomotor functions, thereby causing underaction of the superior or inferior rectus muscles, exotropia, esotropia, or nystagmus.

Better surgical techniques and the fact that growth of the nasal dorsum and increased activity of the frontalis muscle will support the surgeon in his efforts justify the view that, with time, substantial improvement can be obtained by correction of the telecanthus first and the ptosis secondarily.

The medial canthus

Correction of the canthal drift has priority and it can be achieved only by a transnasal canthopexy. Some surgeon perform this procedure in combination with one of the many techniques that can be used to correct an epicanthus. If this approach is preferred, redistribution of skin should be achieved with a minimum of incisions to avoid conspicuous scarring. The various methods that may be used are shown in 8.1.

Blair's technique

Blair's technique involves the use of one or two unequal Z-plasties. It has given good results in our hands (8.6), but we now believe that, in the majority of cases, better results with less scarring can be obtained if no redistribution of skin is performed and all efforts are concentrated on reconstruction of the canthal depression by means of the transnasal canthopexy.

8.6 Correction of epicanthus. a Design of flaps. b Result of transposition. c Preoperative appearance. d Postoperative appearance.
Author's technique

This technique is illustrated in 8.7 and 8.8. A transverse incision is first made over the lax medial canthal tendon. This incision is extended over a small distance into the medial part of the lower eyelid, where the epiblepharon is seen. All the fat and muscle between skin and peristeum in the canthal area are then carefully removed. This step is important because resection of this surplus of tissue allows for accurate and intimate fixation of the skin to the underlying skeleton once a deep canthopexy has been performed in the manner described in Chapter 3.

The canthal depression can thus be reproduced and the need for extensive redistribution of skin is eliminated, because the epicanthal convexity is turned into an endocanthal concavity. The procedure also tends to correct eversion of the lower eyelid and to narrow the palpebral fissure temporarily.

The lateral canthus

Laxity of the lateral canthal insertion or canthal dystopia is best treated by means of a canthopexy.

Canthopexy (see Chapter 3)

A short transverse incision is made just lateral to the outer canthus. Through this incision, the lateral orbital wall is first exposed. The peristeum overlying the skeletal rim is then incised in a cranial and caudal direction over some distance allowing for adequate release of the lateral canthus together with the periorbita and orbital region.

8.7 Correction of epicanthus and telecanthus by canthopexy. a Design of transverse incision in medial canthal region with lateral extension parallel to the rim of the eyelid. b Resection of fat in medial canthal region. c Traction on transnasal sutures in order to turn a canthal convexity into a concavity. d Result of canthopexy and linear closure.

8.8 a Preoperative appearance of a patient with blepharophimosis. b Postoperative appearance of a patient on whom the author's technique was used.
septum. Once sufficient mobility is obtained, the lateral canthus is raised and a canthopexy is performed in the manner described in Chapter 3.

The lower eyelid
Correction of lower lid eversion is usually obtained by adequate medial and lateral canthopexies. If necessary, transposition of a small laterally based V-flap from the upper to the lower eyelid is used in conjunction with the canthopexies. Eversion may, however, be so extreme that its correction by means of a skin graft is considered. It should be remembered that the colour match of a retroauricular graft, although good, never equals the eyelid skin itself.

The upper eyelid
Attempts to correct the ptosis by means of one of the various levator shortening procedures are frequently doomed to fail, from a functional point of view, because the levator muscle is almost nonexistent or hypoplastic. Nevertheless cosmetic improvement is occasionally seen when the tenodesis effect of the procedure results in the restoration of a palpebral fold or the widening of the palpebral fissure.

In the majority of patients however it will be necessary to perform a frontalis suspension. It is true that the direction of pull by this muscle is abnormal and that the eyelid initially may even lose its contact with the globe, but definite functional and cosmetic improvement is almost always obtained.

Operative technique
The procedure used (8.9) is a personal modification from the many techniques that have been advocated in the past. Two short incisions are first made, one at the site of the upper rim of the tarsus, the other immediately above the eyebrow. A tunnel is then made connecting the two incisions. Through the upper incision the surface of the frontalis is carefully dissected over some distance in all directions.

The inferior edge of the frontalis muscle, beneath the eyebrow, is then incised transversely over a distance of approximately 1.5 cm and dissection is continued between the muscle and the periosteum in a cranial direction. A rectangular muscle flap is finally formed by means of two parallel, cranially directed incisions that start at the exposed inferior edge.

The flap thus formed is then pulled through the tunnel and attached to the tarsus by means of several interrupted sutures. The immediate effect may be that the eyelid loses contact with the globe. This rapidly improves, however, owing to the balancing action of the orbicularis muscle (8.10, 8.11).

8.9 Correction of ptosis by frontalis plasty. a Dissection of frontalis muscle flap. b Delivery of frontalis muscle flap through tunnel. c Preoperative appearance. d Postoperative appearance.
BLEPHAROSCHIZIS (COLOBOMA)

The term coloboma refers to full-thickness defects of the eyelids. The anomaly is rare. It can be unilateral or bilateral, and all degrees of severity are seen. The configuration of the defect may be triangular or quadrilateral. The cornea is often exposed, which causes severe risks for normal vision, owing to corneal opacities or ulcerations. These risks are particularly great when the condition involves the centre of the eyelid and normal mobility of the eye is restricted by the presence of tethering bands. Urgent treatment is then indicated.

Colobomas of the eyelid may occur:
• As a solitary anomaly.
• As part of a syndrome that includes a complex variety of skeletal and soft tissue anomalies.

These complex colobomas may be typical or atypical for the syndrome involved.

TREATMENT OF SOLITARY COLOBOMAS

Solitary colobomas are almost exclusively confined to the central and medial parts of the upper eyelid. The reason for this is not known. The size of these defects can be difficult to estimate because the edges are pulled in opposite directions by the separated parts of the orbicularis muscle. Careful examination under anaesthetic is indicated before the best method of treatment can be selected. For this purpose, the edges of the colobomas are approximated with skin hooks to define the true size of the defect. In the colobomas involving less than one-third of the eyelid rim, correction is performed by excision of the edges followed by direct apposition of the two segments in separate layers. Since straight line closure may result in considerable contraction, the incorporation of an additional Z-plasty is advocated.

8.10 Correction of severe blepharophimosis by medial canthopexy and frontalis suspension. a Preoperative appearance. b Postoperative appearance.

8.11 Correction of ptosis following adequate canthopexy. a Preoperative appearance. b Postoperative appearance. Note the strong frontalis muscle action.
Colobomas involving one-third or more of the eyelid rim require a different approach. Several principles have been advocated by various authorities, but the technique that seems simplest and most effective is the transfer of a skin flap lined with a mucosal graft. Unfortunately, the selection of flaps is limited, because the integrity of the lower eyelid deserves to remain intact and the quantity of available skin in the upper eyelid may be small. Nevertheless restoration of the eyelid is feasible in the majority of patients (8.12, 8.13).

8.12 a Coloboma of the upper eyelid. b Dissection of edges and closure of conjunctival defect with buccal graft. c Design of transposition flap. d Resurfacing of buccal graft by transposition of the flap.

8.13 a Coloboma of the upper eyelid. b Reconstruction of the eyelid by closure of conjunctival defect with buccal graft and resurfacing of graft by transposition of flap. Long-term result. Note that the appearance of an eyelid following correction of a coloboma can be improved by secondary resection of a hairless eyelid rim or by 'make-believe' cilia using dermatography.
TREATMENT OF COMPLEX COLOBOMAS

In the correction of complex colobomas, two alternatives based on slightly different principles can be considered (8.14).

The first principle concerns the transfer of a composite flap from the lateral part of the upper eyelid to close a defect in the central or medial part. The lateral segment is first mobilized by a canthotomy and then transferred in conjunction with a temporal extension. This transfer is facilitated by the inclusion of a Z-plasty in the extended skin incision. Closure of the conjunctival defect that has been produced by the canthotomy may require the insertion of a mucosal graft. The straight line closure of the coloboma may demand incorporation of a Z-plasty to prevent or correct contracture.

The second principle concerns the transfer of a semicircular or semiquadrilateral skin flap raised in the lateral part of the eyelid and the palpbraltemporal area. Medial rotation of the flap is facilitated by the inclusion of a Z-plasty in the lateral extension of the incision. The coloboma is first closed with a mucosal graft, inserted under some tension, and the skin flap is then used to cover the graft.

Complex colobomas are usually associated with an underlying skeletal anomaly or cleft. These clefts were described and numbered by Tessier. Circling the orbit in different degrees of severity a distinct pattern can be observed.

An explanation for this was offered by van der Meulen et al., who postulated that the skeletal anomalies have their origin in dysostoses of the various ossification centres, resulting in typical hourglass deformities of the soft tissues (hairline, eyebrow, and eyelids). The size of the associated eyelid colobomas may be of such magnitude that more extensive transfers of tissue involving adjacent areas of skin are involved.

Fortunately, complex colobomas of the upper or lower eyelid are extremely rare. The principles of correction are, however, similar. Redistribution of skin is extensive (8.15, 8.16) and the incorporation of skin is imperative if secondary contracture is to be avoided.

8.14 Principles for closure of complex coloboma. a Medial transposition of lateral eyelid segment using a lateral canthotomy and closure of the resulting conjunctival defect with a mucosal graft. The rotation is facilitated by the incorporation of a Z-plasty. (A1 and A2 show closure of a coloboma in the upper eyelid; A3 and A4 show closure of coloboma in the lower eyelid.)

b Closure of the conjunctival defect with a mucosal graft and medial transposition of lateral eyelid skin. This rotation is facilitated by the incorporation of a Z-plasty. (B1 and B2 show closure of coloboma in upper eyelid; B3 and B4 show closure of coloboma in lower eyelid.)
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8.15 a Severe coloboma of upper eyelid. b Dissection of edges. c Closure of conjunctival defect by insertion of buccal graft and design of transposition flap - the incorporation of a Z-plasty in the lateral part of the periphery of the flap permits rotation of the flap and closure of the small donor defect. d Resurfacing of the buccal graft by transposition of the flap and design of a V-flap that is used for a second Z-plasty in the medial part of the periphery of the flap directly over the buccal graft. e Result of second Z-plasty.

8.16 Severe facial and palpebral clefting (Tessier IV maxillary dysplasia). a Preoperative appearance. b Postoperative appearance.
EPIBLEPHARON

The term epiblepharon refers to the presence of an eyelid fold. Such a fold partially covering the rim of the eyelid may be observed in both eyelids. In spite of the similarity in appearance, the causative mechanism of each of these folds is different.

**UPPER EYELID EPIBLEPHARON**

In the upper eyelid, epiblepharon appears to be due to the protrusion of orbital fat over the levator aponeurosis. This occurs because of a more caudally situated fusion between orbital septum and levator aponeurosis (8.17). This condition is common in Orientals but it is rare in Caucasians, in whom fusion of levator aponeurosis and orbital septum is normally formed above the superior border of the tarsal plate (see 8.17).

Correction is relatively simple. The surplus of fat is resected through an incision at the site of the superior border of the tarsus and a fold is created by the insertion of some sutures that produce strong adhesions between skin and levator aponeurosis.

**LOWER EYELID EPIBLEPHARON**

In the lower eyelid, epiblepharon is caused by a shortage rather than by an excess of tissue. Testimony to this is the combination of epicanthus inversus and epiblepharon in patients with blepharophimosis and the fact that epiblepharon as a solitary phenomenon in the lower eyelid usually disappears with increasing laxity of skin. As a rule treatment is not required, but in some patients the fold may induce a congenital entropion that demands correction.

8.17 a Frontal view and b lateral view of an epiblepharon.
**EURYBLEPHARON**

The term euryblepharon refers to a congenitally long and wide palpebral fissure caused by extreme shortage of eyelid skin. The malformation may be associated with a lateral ectropion of the lower eyelid. Correction can be achieved only by the insertion of skin (8.18).

**CONGENITAL ENTROPION**

The combination of epiblepharon and congenital entropion is occasionally seen in children. It is rare in adults (8.19) because the malformation tends to disappear after a few years.

The entropion seems to be caused by a band of orbicularis muscle underlying the skinfold and overriding the tarsus. When there are no complaints of photophobia or recurrent conjunctivitis, conservative treatment with lubricating ointments is recommended.

There are some cases, however, that should be corrected by surgery. Release, relocation or resection of the muscular band, removal of a small crescent shaped segment of skin (less than 2 mm) and repositioning of the rim of the eyelid by the use of a few everting buried sutures is usually sufficient (8.20).

**CONGENITAL ECTROPION**

This anomaly is usually seen as part of blepharophimosis or euryblepharon. It is rare as a solitary malformation (8.21). Its cause is difficult to define. Factors that may play a role include:

- Laxity of the canthal tendons, in particular the medial tendon.
- Shortness of the orbital septum.
- Horizontal lengthening of the rim of the eyelid.
- Lack of skin.

Methods of correction involve canthopexies, lid shortening, and septal release. Serious effort should be made to avoid the insertion of skin grafts because the colour match is never satisfactory.
8.20 Congenital entropion associated with epiblepharon in a child. a Before correction. b After correction.

8.21 Congenital ectropion. a Right eye. b Left eye.

**BIBLIOGRAPHY**

**Congenital Anomalies**


EURYBLEPHARON


OTHER FURTHER READING


