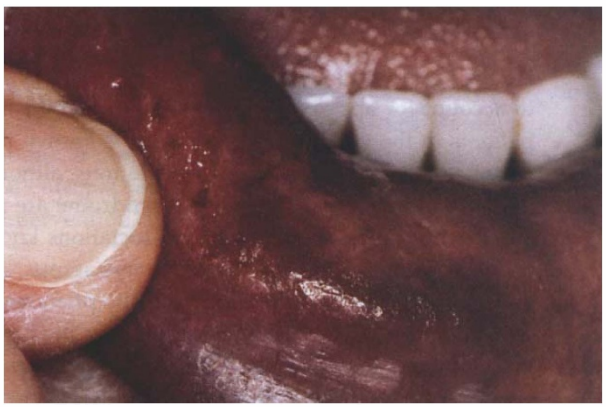

Fig. 2. *Large mucosal defect seen on lower lip eversion.*

planes to the lumbar plexus, anaesthetising the obturator nerve and lateral cutaneous nerve of the thigh as well.[3] There is one reported case of immediate contralateral amaurosis following a retrobulbar anaesthetic, one theory being that the anaesthetic tracked under the optic nerve sheath to the chiasm,[4] presumably the mechanism of rare intrathecal spread and respiratory depression, anaesthetic being recovered from the cerebrospinal fluid after one such event.[5]

This rare complication has not been previously reported and was not observed in a large series of 16 224 consecutive peribulbar blocks,[6] but may be added to the list of needle-related complications, supporting further the argument in favour of the alternative local anaesthetic techniques for cataract surgery – sub-Tenon, topical and intracameral – on safety grounds. These techniques are justifiably becoming more popular with ophthalmic surgeons, with the suggestion that most intraocular procedures can be completed without use of retrobulbar or peribulbar blocks.[7,8] It would seem wise for staff to enquire about lip and tongue anaesthesia before offering food and hot drinks to post-operative patients following use of a peribulbar or retrobulbar block.

References

1. Bron AJ, Tripathi RC, Tripathi BJ. Wolff's anatomy of the eye and orbit, 8th ed. London: Chapman and Hall Medical, 1997:8.
2. Gills JP, Hustead RF, Sanders DR, editors. Ophthalmic anesthesia. Thorofare, NJ: Slack, 1993:146.
3. Brown DL. Atlas of regional anaesthesia. Philadelphia: WB Saunders, 1992:7.
4. Bishara SA, Estrin I, Rand WJ. Immediate contralateral amaurosis after retrobulbar anesthesia. Ann Ophthalmol 1990;22:63–5.
5. Kobet KA. Cerebral spinal fluid recovery of lidocaine and bupivacaine following respiratory arrest subsequent to retrobulbar block. Ophthalmic Surg 1987;18:11.
6. Davis DB, Mandel MR. Efficacy and complication rate of 16 224 consecutive peribulbar blocks. J Cataract Refract Surg 1994;20:327–37.
7. Greenbaum S, editor. Ocular anesthesia. Philadelphia: WB Saunders, 1997:30–40.
8. Manners TD, Burton RL. Randomised trial of topical versus sub-Tenon's local anaesthesia for small incision cataract surgery. Eye 1996;10:367–70.

Wayne Birchall ✉
Manchester Royal Eye Hospital
Oxford Road
Manchester M13 9WH, UK
Tel: +44 (0)161 276 1234
e-mail: waynebirchall@yahoo.co.uk

Sir,

Primary congenital upper eyelid entropion in association with Duane's retraction syndrome

Congenital primary upper eyelid entropion is a very rare eyelid anomaly.[1] Corneal complications are common.[2] The diagnosis may easily be overlooked in the crying infant with secondary blepharospasm.

Duane's syndrome usually presents as failure of abduction of the affected eye with co-contraction of the horizontal recti muscles, resulting in narrowing of the palpebral fissure on adduction. It is known to be associated with various ocular and non-ocular anomalies, including skeletal, neural and auricular defects.[3,4]

We report a case of congenital primary right upper eyelid entropion in association with a right-sided Duane's syndrome. To the best of our knowledge, this association has never previously been described in the literature. Furthermore, spontaneous resolution of congenital primary upper eyelid entropion has also never previously been reported in the literature to the best of our knowledge.

Case report

A 5-month-old female infant was referred to the oculoplastic service of the Manchester Royal Eye Hospital with an entropion of the right upper eyelid. She had been a full-term normal delivery and was normal apart from jaundice at birth, treated successfully by phototherapy. Examination revealed a complete right upper eyelid entropion with the eyelashes directed against the cornea (Fig. 1). The tarsal plate structure was normal with no evidence of any kinking. There was also a bilateral lower eyelid epiblepharon. There was no

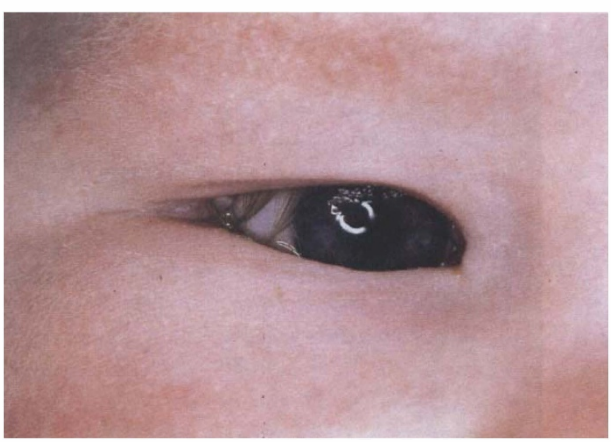

Fig. 1. *Photograph of the infant's right eye showing upper eyelid entropion with the lashes directly opposed to the globe.*

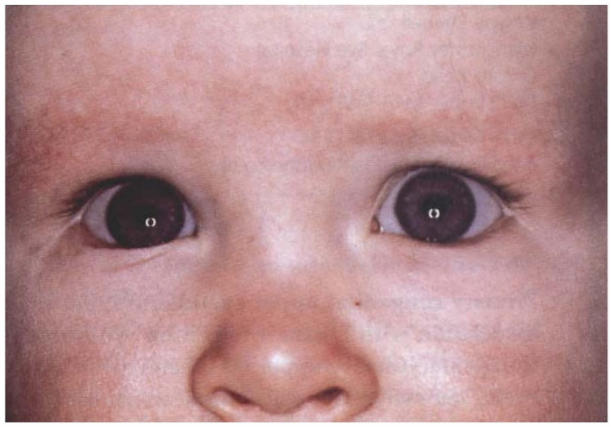

Fig. 2. *Photograph of the infant 2 months later. The right primary upper eyelid entropion has resolved.*

evidence of any corneal abrasion or ulceration and the rest of the ocular examination was unremarkable. A diagnosis of true congenital primary upper eyelid entropion was made. It was decided to keep the infant under regular observation, given the lack of symptoms and the absence of secondary corneal defects, and to defer any surgical intervention.

The infant was seen 2 months later. The parents reported that over the preceding few weeks the infant appeared to have developed a squint. On examination, the right upper eyelid entropion had completely resolved with the lid now in an entirely normal position (Fig. 2). In addition, the bilateral lower lid epiblepharon had resolved. However, the infant appeared to have developed an abnormal head posture with a slight face turn to the right. Examination of her ocular movements revealed an absence of abduction of the right eye beyond the mid-line and significant narrowing of the palpebral fissure on laevoversion, supporting a diagnosis of Duane's syndrome type 1 (Fig. 3). Her visual acuity was recorded as 3.8 cycles per degree in each eye. There was no significant refractive error in either eye and she had good binocular function.

She continues to enjoy good binocular vision in both eyes, and remains under review.

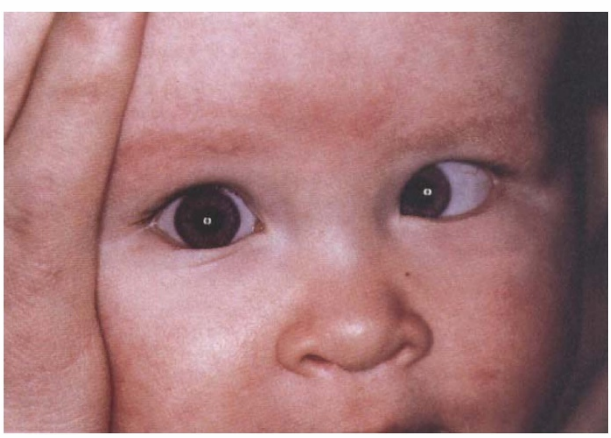

Fig. 3. *Photograph showing failure of abduction of the right eye beyond the mid-line.*

Comment

Duane's syndrome is relatively common and is believed to account for approximately 1–4% of all strabismus patients in the general population.[3] It is known to have various ocular and non-ocular associations, although the majority of patients with Duane's syndrome are free of other congenital defects.[3] Ocular associations known to occur in Duane's syndrome include nystagmus, epibulbar dermoids, colobomas and the phenomenon of crocodile tears. Eyelid anomalies that have been reported in association with Duane's syndrome include ptosis, congenital ectropion[5] and lagophthalmos.[6] Congenital upper eyelid entropion has never previously been described in association with Duane's syndrome.

It seems that the majority of Duane's-related anomalies that occur, do so at the embryological stage of between 4 and 8 weeks of gestation,[3] thus suggesting they may all be linked to this critical point of embryological development. It has previously been postulated that a teratogenic effect occurring during the first trimester of pregnancy may be why Duane's syndrome is frequently associated with other congenital defects.[5]

Primary congenital upper eyelid entropion is exceedingly rare and is usually, but not always, associated with a deformed tarsal plate.[7] It is frequently confused with epiblepharon, which is characterised by the presence of a fold of skin running horizontally across the upper or lower eyelid.[8] All previous reports of primary congenital upper eyelid entropion have shown no evidence of spontaneous resolution and were surgically corrected. This has been used as one of the distinguishing criteria between true congenital entropion and the more common epiblepharon, which usually resolves spontaneously.[8]

However, this case was one of true congenital entropion as characterised by the fact that the position of the lid margin was completely inverted along its entire length with the lashes directly opposed to the globe. This is in contrast to epiblepharon, where the lashes are merely pushed against the globe due to an excess fold of skin.

Due to the risk of developing corneal complications as a result of the entropion, the infant was being considered for early surgical intervention, only to discover at further review that the entropion had spontaneously resolved. It is possible that a proportion of upper eyelid entropia would resolve but the vast majority are surgically corrected before this can occur spontaneously.

It has been postulated that primary congenital upper eyelid entropion may be part of a syndrome involving many systemic anomalies[1,9] and this may be related to the same teratogenic event that occurs in Duane's syndrome. Indeed, it has previously been emphasised that a general systemic evaluation is mandatory in all cases of congenital upper eyelid entropion.[1]

In conclusion, we report a unique case of primary congenital upper eyelid entropion which spontaneously resolved and occurred in association with Duane's

retraction syndrome. It lends further evidence to the theory that the two have a common teratogenic aetiology in early gestation. The fact that the entropion resolved would suggest that, in the absence of any corneal or other complications, it may be possible to closely monitor congenital entropia rather than perform immediate surgical intervention, as they may resolve spontaneously.

References

1. Zak TA. Congenital primary upper eyelid entropion. J Pediatr Ophthalmol 1984;21:69–73.
2. Luchs JI, Laibson PR, Stefanyszyn MA, *et al*. Infantile ulcerative keratitis secondary to congenital entropion. Cornea 1997;16:32–4.
3. DeRespinis PA, Caputo AR, Wagner RS, *et al*. Duane's retraction syndrome. Surv Ophthalmol 1993;38:257–88.
4. Cross HE, Pfaffenbach DD. Duane's retraction syndrome and associated congenital malformations. Am J Ophthalmol 1972;73:442–50.
5. Pfaffenbach DD, Cross HE, Kearns TP. Congenital anomalies in Duane's retraction syndrome. Arch Ophthalmol 1972;88:635–9.
6. Ro A, Gummeson B, Orton RB. Duane's retraction syndrome: southwestern Ontario experience. Can J Ophthalmol 1989;24:200–3.
7. Bartley GB, Nerad JA, Kersten RC, *et al*. Congenital entropion with intact lower eyelid retractor insertion. Am J Ophthalmol 1991;112:437–41.
8. Duke-Elder S. System of ophthalmology, vol III, part 2. St Louis: CV Mosby, 1963:857.
9. Hiles DA, Wilder LW. Congenital entropion of the upper lids. J Pediatr Ophthalmol 1969;6:157–61.

N. Patton
B. Leatherbarrow
Department of Ophthalmology
Manchester Royal Eye Hospital
Manchester, UK

Mr B. Leatherbarrow ✉
Manchester Royal Eye Hospital
Oxford Road
Manchester M13 9WH, UK
Tel: +44 (0)161 276 5569
Fax: +44 (0)161 272 6618

Sir,

Necrosis of the lid due to *Pseudomonas aeruginosa*

Necrosis of the eyelids is rare and usually related to minor trauma such as electrical burns or spider bites, or, more dangerously, to infection.[1–7] In the absence of a history of trauma or infection a mitotic lesion such as a sebaceous cell carcinoma or a fungal lesion such as mucormycosis should be considered. Of infectious causes the commonest organisms are streptococci or staphylococci, but seven instances of *Pseudomonas* involvement have been reported, usually bilateral and associated with neutropenia.[8–14] We report such a case.

Case report

An 83-year-old woman with peripheral vascular disease and cardiac failure was admitted with pneumonia and a cellulitis affecting her lower leg, and treated with intravenous antibiotics. The cellulitis centred on a small punched-out ulcer on her left medial malleolus. Blood cultures revealed *Pseudomonas aeruginosa* and a coagulase-negative *Staphylococcus*, though neither were present in every bottle, and the question of contamination was raised. *Pseudomonas* was also found in the leg wound, however. Initial treatment with cefuroxime was therefore changed to gentamicin and piperacillin after sensitivity testing. The organism was sensitive to ciprofloxacin, pipericillin, gentamicin, ceftazidime, azatreonam, meropenem, neomycin and colistin. It was resistant to cefotaxime, trimethoprim, ampicillin/amoxycillin, cephadrine, chloramphenicol and tetracycline.

On the third day of her admission she developed a lesion on her right lower lid which was treated empirically with topical chloramphenicol and later with topical gentamicin as a swab revealed *Pseudomonas aeruginosa*, resistant to chloramphenicol. An ophthalmic review was requested on the tenth day. The patient was observed to have a grossly injected conjunctiva but, more interestingly, an area of black eschar covering the lateral third of the lid with yellowish discharge from the conjunctival surface which appeared to be frankly necrotic (Fig. 1a). Fungal involvement was felt unlikely on clinical grounds. Further swabs were taken for bacteria and fungi and topical medication changed to ciprofloxacin until a biopsy could be arranged. This was in part because it was felt the gentamicin might be exacerbating the conjunctival inflammation. The appearances began to improve. Three days later a formal full-thickness excisional wedge biopsy was performed. At biopsy the tissues looked relatively avascular. The defect was closed by primary intention with a good final result. However, the patient did not survive a developing gangrene of her leg for which she refused surgery.

The biopsy was examined at several levels (Fig. 1b). It showed an area of acute inflammation with surface ulceration and crusting and also inflammation related to occasional adnexal structures. No infecting fungus or bacteria was identified on PAS, Grocott or Gram staining and there was no evidence of tumour. Appearances were reported as non-specific but suggested the lesion may have originated within a hair follicle.

Comment

The lateral and medial palpebral arteries form two arcades in each eyelid which are thus richly supplied.[15] Healing, even after full-thickness excisions, is excellent. Infection rarely progresses because of the excellent blood supply and lymphatic drainage predominantly to the superficial parotid nodes, though debility can be permissive. Destruction of lid architecture may occur through a neoplastic process such as sebaceous cell carcinoma.

Increasingly recognised has been the occurrence of lid necrosis secondary to Group A streptococci, especially if the strain expresses pyrogenic exotoxins whilst expressing M surface protein, which facilitates invasion