Sir,

Emergency use of pilocarpine and pupil block glaucoma in ectopia lentis

Pupillary block glaucoma secondary to ectopia lentis is well described, as is its potential exacerbation with the application of topical miotics.[1]

However, miotics can still be used in association with ectopia lentis in two situations: to either confine a dislocated or severely subluxated lens to the posterior chamber pending vitreolensectomy or to confine a dislocated lens to the anterior chamber pending urgent lens extraction via an anterior approach.[2] Both indications are not without risk. We would like to present a short case series to demonstrate the risks of miosing patients with ectopia lentis in emergency situations and to recommend alternative strategies to pilocarpine.

Case reports

Case 1

A male patient with Marfan's syndrome and inferiorly subluxated microspherophakic lenses presented to eye casualty in 1994 with the microspherophakic left lens incarcerated in the pupil, endothelial touch, and an intraocular pressure of 62 mmHg secondary to pupil block. The lens returned to the posterior segment with posturing, oral acetazolamide, and tropicamide mydriasis. The patient was commenced on pilocarpine 4% qds to the left eye awaiting surgical intervention. The pupil was noted to be well miosed. However, 2 days later he reported increasing pain. Examination revealed the microspherophakic lens again incarcerated in the pupil and an intraocular pressure of 48 mmHg. The lens returned to the posterior pole with mydriasis and posturing, and a vitreo-lensectomy was performed the following day. The eye settled and his vision was corrected to 6/9 with an aphakic contact lens.

Case 2

A 27-year-old female with Marfan's syndrome and bilateral inferiorly subluxated lenses complained of a change of vision in her left eye. She had long experienced an aberration in her left eye due to the margin of her lens encroaching upon the visual axis and consequently wore aphakic spectacles over homatropine 2% mydriasis. The aberration disappeared 5 days previously, and on examination it was noted that the lens had spontaneously dislocated onto the posterior pole. Over the next 3 days, however, the aberration returned and was more prominent. Examination of her left eye revealed that the lens had completely dislocated into the anterior chamber (Figure 1). The intraocular pressure in the left eye was 16 mmHg. She was immediately commenced on topical pilocarpine 4% qds, in order to prevent the lens prolapsing back into the posterior segment prior to surgical removal.

The following morning, the patient complained of severe pain in her left eye. On examination, the eye was injected and the lens was completely incarcerated within the pupil. The intraocular pressure was recorded at 50 mmHg. A diagnosis of secondary pupil-block glaucoma was made. The cornea was markedly oedematous and the view was too compromised to perform laser iridotomy. Topical atropine 1% and intravenous mannitol were administered, which normalized the intraocular pressure and the patient was taken to theatre. The lens was removed using a bimanual lensectomy technique with a limited anterior vitrectomy and a peripheral iridectomy. No intraocular lens implant was inserted, as the patient was happy using her aphakic spectacles. At 1 week postoperatively, the vision in the left eye was 6/6.

Case 3

A 51-year-old male was referred to a local eye clinic by his optometrist who noted deteriorating vision and increasing myopic astigmatism. As part of the examination, his pupils were dilated with tropicamide 1% at which point, inferior subluxation of the right lens and marked left phacodonesis was noted. He was therefore, given pilocarpine 4% to both eyes to reverse the mydriasis and sent home pending referral for management of his subluxated lenses and systemic investigations.

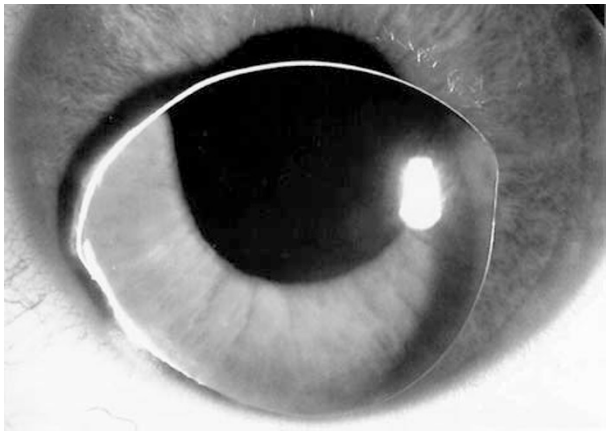

Figure 1 Case 2: Dislocated lens in anterior chamber prior to administration of topical pilocarpine and subsequent pupil block glaucoma.

He returned to his local eye unit that same day with increasing discomfort in the right eye and deteriorating vision. The crystalline lens in the right eye had dislocated into the anterior chamber leading to pupil block glaucoma and an intraocular pressure of 62 mmHg. He was referred to the teaching unit where subconjunctival Mydricaine No.2 (Moorfields Eye Hospital Pharmacy), supine posturing, and intravenous diamox followed by intravenous mannitol failed to return the lens to the posterior segment. As with Case 2, marked corneal oedema prevented the safe placement of a patent iridotomy. Lens ballotment was attempted by corneal massage, but was abandoned due to patient discomfort. A minimal surgical intervention was therefore performed, in which 0.5 ml of fluid vitreous was aspirated via a 23-gauge needle through the pars plana. The lens was then balloted through the cornea until it reposited behind the iris.

At 1 week postoperatively, the visual acuity was still reduced at 1/60 in the right eye due to persistent corneal oedema. Intraocular pressure was 8 mmHg bilaterally. He was referred back to his local ophthalmic unit for continuing management and systemic investigation with regard to his ectopia lentis.

Comment

There are three types of lens-related pupillary block glaucomas in the context of ectopia lentis. The first is pupillary block by a subluxating lens posterior to the iris. The loose zonules allow the lens to move forward, increasing its contact with the iris. Iris bombé and angle closure glaucoma then develop.[1] Treatment with pilocarpine makes the block worse by stimulating ciliary muscle contraction, thereby loosening the zonules and further increasing anterior lens displacement. Mydriatics tighten the zonules and can break the attack. This has therefore been termed 'inverse glaucoma' and occurs only in eyes with some remaining zonular support to the lens.

The second type of pupil block is due to incarceration of the lens directly within the pupil. Increasing aqueous pressure behind a subluxated lens and increasing relative pupillary block leads to further lens movement anteriorly with eventual incarceration within the iris sphincter. Microspherophakic lenses are particularly prone to this complication, as in Case 1.

The third type follows complete dislocation of the lens into the anterior chamber. The posterior surface of the lens in contact with the iris then induces an anterior pupil block. Anterior dislocation can occur either following pupillary block with posterior aqueous pressure forcing the lens to dislocate into the anterior chamber, or following dilation, either physiological or iatrogenic, as in Cases 2 and 3.

The use of miotics has been suggested in the past for ectopia lentis to prevent lens dislocation into the anterior chamber but in association with iridectomies to reduce the risk of pupillary block and progressive anterior lens movement. However, Elkington *et al*[3] described a case series of 5 homocystinuria patients on long-term miotics (pilocarpine 4% or ecothiopate 0.125%), who continued to suffer lens dislocations and in which one of the patients already had a patent iridectomy. Harrison *et al*[4] concluded that iridectomies failed to prevent lens dislocation in their group of 45 patients with homocystinuria.

The use of miosis without an iridectomy either long term or while awaiting surgery will increase the risk of pupil block recurring with a subluxated lens and could thereby encourage lens dislocation anteriorly with subsequent recurrence of incarceration, as in Case 1. This may have been exacerbated by the use of 4% pilocarpine, which can lead to such zonular laxity as to aggravate pupil block in association with primary angle closure glaucoma.[5] Case 1 was noted to be well miosed, which would support this. However, this mechanism would be less pertinent for Cases 2 and 3 in who the lenses were completely free of zonular support.

Jaffe *et al*[2] recommends constricting the pupil with a lens dislocated into the anterior chamber, trapping the lens, and allowing extraction anteriorly. However, without an iridectomy the risk of inducing a pupil block in an otherwise quiet eye with the crystalline lens anterior is significant. As in Case 2, the lens had presumably been within the anterior chamber for 3 days, but pupil block glaucoma was precipitated only after the initiation of pilocarpine treatment.

Nonsurgical management of an anteriorly dislocated lens includes mydriasis with the patient supine. If this fails, central corneal pressure with a cotton tip swab to reposition the lens manually is recommended.[4] Although in both Cases 2 and 3 the cornea was already too oedematous at presentation to allow attempts at iridotomy, the corneal ballotment required by Case 3 (as per cited protocol) probably contributed to the persistent corneal oedema at discharge.

Although a lens completely free of zonular support should have a reduced risk of inducing pupillary block, the lens can still dislocate into the anterior chamber with the patient on pilocarpine, as in Elkington's series. And a miosed pupil will not only increase the risk of pupillary block from an anteriorly dislocated lens but also complicate any attempt to reposit the lens back to the posterior pole, as in Case 3.

We would therefore like to recommend that if the lens is to be kept anterior or posterior pending surgery, that

posturing the patient either face down or supine, respectively, is a much safer and more effective technique than the use of miosis. Although we have not reviewed surgical techniques for lens extraction in ectopia lentis, we would suggest that repositioning of the lens posteriorly is associated with less endothelial trauma. The risks of a posterior approach with a pars plana vitreolensectomy can be reduced in Marfan's patients with intraoperative laser retinopexy to areas of lattice.[6]

If miosis is to be attempted, then a laser iridotomy is also advisable not only to prevent pupil block from subluxating lenses in the posterior chamber but also to protect against pupil block if the lens dislocates anteriorly.

Our case series demonstrates the significant risk of precipitating pupil block glaucoma and potentially delaying surgery for ectopia lentis by the emergency use of pilocarpine without iridotomy.

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Sir,

Optical coherence tomography findings in acute macular neuroretinopathy

Acute macular neuroretinopathy (AMNR) is a rare condition characterized by a sudden onset of mild visual impairment in one or both eyes of young patients, predominantly women. Typically, the neuroretinopathy consists of reddish-brown wedge-shaped macular lesions, pointing towards the centre of the fovea.[1] The diagnosis is clinical, aided by the exact correspondence of the lesion with the scotoma identified on the Amsler grid. The precise location of the lesions is uncertain.[1] We evaluated a case of AMNR with optical coherence tomography (OCT) and fundus fluorescein angiography (FFA).

Case report

A 30-year-old man presented with blurring of vision in the left eye for 6 months. His best-corrected visual acuity was 6/6 in the right eye and 6/9 in the left. There was no history of headache, flu-like illness, trauma, or administration of intravenous epinephrine/contrast agents. Anterior segment examination was normal in both eyes. Slit-lamp biomicroscopy of the left fundus showed a subtle red wedge-shaped lesion above the fovea pointing towards its centre (Figure 1(a)). The right fundus was normal. Amsler grid examination of the left eye showed a scotoma corresponding exactly to the macular lesion. FFA revealed no hypofluorescence of the affected area. Optical coherence tomography of the left eye revealed distinct retinal thinning in the area of the lesion, more prominent in the inner retinal layers (Figure 1(b)).

Comment

The dark-red appearance of the lesions in AMNR is attributed to a focal atrophic thinning of retina.[1] Opinion is divided on the location of the lesions. Biomicroscopic interpretation is variable.[1,2] The striking correspondence of the lesions to the paracentral scotomata on Amsler grid and documentation of an abnormality of the early receptor potential point to the involvement of the outer retinal layers.[1,3] The retinal haemorrhages, acute oedematous appearance of the macula, suspected retinal capillary dilation, and an association with epinephrine administration indicate inner retinal involvement.[1,2,4] To the best of our knowledge, there is only one earlier report of OCT findings in AMNR: Feigl and Hass[5] observed a band of high reflectivity over the retinal pigment epithelium, and implicated an inflammatory process in the outer retina.They found no retinal thinning in their