We suggest that some unknown feature(s) of responders with high IOP enables steroids to reduce retinal edema.

Conflict of interest
The authors declare no conflict of interest.

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Sir,

The island of ischaemia: submacular choroidal nonperfusion in giant cell arteritis

Olali *et al*[1] presented a case of biopsy-proven giant cell arteritis (GCA) with atypical visual loss. The visual impairment had manifested as bilateral reduction of visual acuity but with sparing of peripheral field. Fluorescein angiography provided the *denouement*: there was focal perfusion defect localised to the submacular choroid. The angiographic curiosity was more pronounced in the right eye, with its central scotoma of greater eminence, as evidenced by a poorer acuity. The authors noted that this pattern—of isolated focal ischaemic choroidopathy in GCA patients—had been reported earlier.[2] However, as the literature burgeons we become ever remiss of treasures from the past.

Almost 40 years ago a monograph called 'Segmental nature of the choroidal vasculature' appeared in the *British Journal of Ophthalmology*, a masterclass by SS Hayreh on the anatomy and haemodynamics of the choroidal vascular system, with allusion to the superimposition of diseases including arteritis.[3] A homage to this contribution better explains the features observed by Olali *et al*[1] in their patient.

They wished specifically to report 'pigmentary disturbance' at the maculae in a case of GCA in whom there appeared no other ophthalmoscopic features. A reference to the oeuvre of Hayreh affords insight: acute submacular choroidal ischaemia causes pallor of the overlying retinal pigment epithelium. Eventually, the pallid zone acquires depigmentation, the precise ophthalmoscopic anomaly visible in the patient under discussion.

Indeed the description appended by Olali *et al*[1] to their case report can be elevated to a formulation *in dilgenter*: in their presented arteritic case, the occurrence of submacular ischaemia (with sparing of the optic nerve head) denoted hypoperfusion in the lateral posterior ciliary artery (LPCA), the vascular stalk that serves the temporal posterior choroid. As perfusion pressure decreased inside the LPCA—in this situation as consequence of arteritis—the submacular choroid was the first casualty of ischaemia because it is a watershed vascular territory. This is the explanatory nub of the clinical constellation detected in this patient.

Of further intrigue, the optic nerve head escaped infarction because it retained perfusion from the medial PCA and a meshwork of circumferential short PCAs. With unsuppressed disease progression, however, the inevitable course would have been *arteritic anterior ischaemic optic neuropathy*, as the operational nerve head feeders succumbed finally to arteritis. Through a marshalling of superb diagrams, the outstanding duo of McLeod and Hayreh,[4] both now emeritus professors, have earlier expounded the behaviour of the choroidal tree as observed after piecemeal cauterisation of its various feeder PCAs.

Also in this patient, the submacular choroidal ischaemia was cast into a sharp outline by the 19-s image from the angiogram. Later, at 52 s, the outermost fringe of the ischaemic zone had turned indistinct. Explanation for this observation again comes from the experiments of landmark stature, which hark back some 40 years. As ischaemic patches form within the choroid there is remedial response from neighbouring vascular systems, instanced among others by retrograde flux in the vortex veins. When such zones of choroidal ischaemia are captured over days and weeks via serial angiograms, the non-perfused zones are seen to contract in area. The speed of angiographic filling improves through brisk consolidation of collateral flow. Thus, late frames were found to show dye ingress into choroidal zones that showed stark ischaemia in the opening sequence.

The broader message for ophthalmologists is that, in its incipient stealthy phase, GCA can produce localised ischaemic patches in the choroid while entirely sparing the optic nerve and retinal arterial system. Ophthalmologists must be cognizant of this possibility when faced with an older patient who has seemingly inexplicable visual loss and biomicroscopic signs of minimal intensity.

Conflict of interest
The author declares no conflict of interest.

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Sir,
Alternative diagnoses with ectopia lentis

We read with great interest the case presented by Moore *et al*,[1] suggesting a correlation between Sturge–Weber syndrome and ectopia lentis. As the authors discuss, the ophthalmic features of Sturge–Weber syndrome do not classically include ectopia lentis, with only two previous reports of such an association.[2,3] The first of these reports is over 35-years old,[2] whereas the second may have been attributable to trauma.[3] Moore *et al* therefore seem to be presenting the first case of ectopia lentis associated with Sturge–Weber syndrome in the era of modern genetic analysis.

The most common cause of spontaneous ectopia lentis is Marfan syndrome, which the authors excluded on clinical grounds in this case, although no family history is discussed. Although normal mentation is mentioned, other syndromes associated with ectopia lentis have not been explicitly discussed by the authors. Nevertheless, if all associated syndromes are excluded, the condition of isolated ectopia lentis (IEL) must be considered. IEL has been established for over 30 years, with both dominant and recessive inheritance reported.[4,5] This can occur with marked asymmetry (Chandra A. ARVO Abstract (2766)(2011)) and indeed be unilateral.[6] Although estimates of IEL prevalence and incidence are yet to be established, up to 31% of congenital ectopia lentis in a national study were not associated with a nosological classification.[7] These may thus be termed IEL, suggesting that the condition of IEL is far more common than an association with Sturge–Weber syndrome, which Moore *et al* are reporting. We have previously shown that mutations in *FBN1* and *ADAMTSL4* have been associated with IEL.[8] Other genetic mutations have also been associated with ectopia lentis.[9]

Although we do not deny that the association suggested in this case by Moore *et al* may be true, it is important to exclude more common causes of ectopia lentis beyond Marfan syndrome, including IEL. Interrogation for known genetic mutations would be an important and crucial step before such an association can be suggested.

Conflict of interest
The authors declare no conflict of interest.

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Sir,
Response to 'Alternative diagnoses with ectopia lentis'

Chandra and colleagues[1] rightly wonder about other possible causes for ectopia lentis seen in our patient with Sturge–Weber syndrome (SWS).[2] Because of space constraints, we did not report on the full differential diagnosis of ectopia lentis.[3] As we noted, our patient denied even a remote history of trauma; her normal mentation and body habitus, and her ocular examination and the unilaterality of her condition made several possible diagnoses unlikely, including ectopia lentis and