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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 colleagues¹ 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

pupillae, homocystinuria, Weill–Marchesani syndrome, sulfite oxidase deficiency, and hyperlysinemia. In addition to the investigations included in our report, she reported no family history of Marfan syndrome, nor of any other family members with her condition, including her two school-aged children. In addition, no clinical evidence of pseudoexfoliation, retinitis pigmentosa, buphthalmos, megalocornea, chronic uveitis (other than that associated with her condition), or ocular syphilis, nor aniridia was seen.

To our regret, we did not perform any genetic studies, and our patient may have separate mutations resulting in both SWS and isolated ectopia lentis. Indeed, that is likely, given the paucity of reports associating ectopia lentis and SWS. However, reports of unilateral isolated ectopia lentis are similarly rare;⁴ this entity is usually familial and bilateral, at least those due to known genetic mutations.⁵ We welcome further research into the pathogenetic mechanisms of ectopia lentis.

Conflict of interest

The authors declare no conflict of interest.

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Sir, Endophthalmitis following vitrectomy

We read with interest the article by Patel and Rahman,¹ whose study involved an online survey aimed at members of the Britain & Eire Association of Vitreoretinal Surgeons (BEAVRS) to disclose their experience with endophthalmitis following smaller gauge vitrectomy surgery. Two cases of endophthalmitis following 4944 transconjunctival 23G vitrectomy operations were noted, providing an estimate for the incidence of 1 in 2472.

As the authors highlight, this remains an estimate due to the low response rate (31%) and lack of documentary evidence. However, it is reassuring that this study has not revealed a dangerously high rate for this severe complication as sutureless vitrectomy has now become the mainstay in an increasing number of vitreo-retinal centers.

Previous studies have differed greatly with respect to the estimated incidence of endophthalmitis following sutureless vitrectomy—exemplified by the study at Wills Eye Hospital² finding the rate to be more than 12 times higher than that following sutured vitrectomy and contrasting with another American collaborative study³ that recently found no increased rate of endophthalmitis following sutureless surgery. These studies are limited by their retrospective nature and coverage of a relatively small catchment area.

In order to establish a more reliable incidence for endophthalmitis following vitrectomy in the United Kingdom, we have currently completed 15 months of prospective, national surveillance (in association with the British Ophthalmic Surveillance Unit, BOSU), and received 18 reports of endophthalmitis following vitrectomy—14 of which meet our case definition. Given that data from the Hospital Episode Statistics disclose that ~20 000 pars plana vitrectomies are performed each year, this provides an approximate incidence of 1 in 1800 before adjusting for underreporting (yet to be established with the use of validation centers). This framework that BOSU helps to provide is well suited to rare complications providing prospective surveillance across a large geographical area. National surveillance for this complication terminates in May 2012 and we urge all UK ophthalmologists to report cases to us via BOSU or directly to jonathanpark@nhs.net in order to investigate this disastrous complication thoroughly.

Conflict of interest

The authors declare no conflict of interest.

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