The eye in Marfan's syndrome

Sir,

In their review of Marfan's syndrome (Vol 62(3), 2001, p. 153), Aburawi et al understandably focus on cardiac compliations but somewhat to the neglect of the effects on other systems of this multisystem disease. The ocular complications of Marfan's syndrome are not life-threatening (unlike aortic dissection) but prompt recognition may enable improvement and preservation of sight. Since the initial observations of Borger (1914), the list of ocular complications of Marfan's syndrome has been found to include: ectopia lentis, cataracts, glaucoma, strabismus, myopia, reduced miosis, retinal detachment, megalocornea, corneal clouding and scleral defects.

The best known complication of ectopia lentis, and a major criterion for diagnosis of Marfan's syndrome, arises from changes in fibrillin (in the epidermal growth factor-like regions), the major constituent of the suspensory ligaments of the lens (Dietz et al, 1991). This results in the ligaments being attenuated or even broken. Interestingly, the lower ligaments are more severely affected, giving rise to upward dislocation, in contrast to homocystinuria (where there tends to be inferionasal dislocation) and Weill–Marchesani syndrome (inferioanterior dislocation).

The patient may describe blurring or even diplopia. Signs include iridodonesis (the tremulousness of the iris because of loss of support), an apparent deepening of the anterior chamber and, sometimes very helpfully, the edge (equator) of the lens may be seen in the aperture of the pupil. It occurs in approximately 80% of patients with Marfan's syndrome, and in 50%, it is apparent by the age of 5 years. Interventions vary from simple optical correction by spectacles or contact lenses to medical manipulation of the iris diaphragm to surgical removal of the lens.

Most seriously, changes in the lens position and possibly in the trabecular meshwork may result in sight-threatening glaucoma. Medical therapy may be used to temporize, but definitive surgical treatment, such as prophylactic iridectomy, may be required. Assessment and treatment may be complicated if the onset of glaucoma occurs in childhood.

Marfan's syndrome is truly a multisystem disease. The ocular findings are not merely useful diagnostic footnotes, but potentially serious complications in their own right. Prompt recognition, monitoring and treatment may help to prevent sight-threatening amblyopia or glaucoma.

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Dietz HC, Cutting GR, Pyeritz RE et al (1991) Marfan syndrome caused by a recurrent de novo missense mutation in the fibrillin gene. *Nature* **352:** 337–9

Further reading

Buckley EG (1998) Paediatric cataracts and lens anomalies. In: Nelson LB, ed. *Harley's Paediatric Ophthalmology*. 4th edn. WB Saunders, Philadelphia
Pennock CA (1999) Congenital disease — inherited disorders of metabolism. In: Easty DL, Sparrow JM, eds. *Oxford Textbook of Ophthalmology.* 1st edn. Oxford University Press, Oxford

New standards for public health

Sir,

Increasingly, it is realized that public health is an autonomous and mainstream speciality, which, for example, is neither clinical medicine nor anthropology. At the moment, public health is thought to be a branch of clinical medicine, and yet anthropology is a core component. Many complex disciplines are core to public health, of course — depending on what aspect of wellbeing, among whom and when it is to be protected or improved. To enable public health to be effective, it has to have a strong and dedicated workforce in various sectors acting together for a common purpose, led by enthusiastic lifetime experts from whatever relevant discipline.

In his editorial (vol 62(4), 2001, p. 198), Dr Golding describes well the processes that have been occurring in this country to enable such a workforce (and an appropriate infrastructure) to be properly developed. One of the basic problems is that the existing vocabulary is all wrong; for a start, there is no descriptive noun that adequately describes these experts — apart from public healthist, which is just clumsy.

Nor can the degree of competence be properly described, since specialist in public health now means something to do with becoming only competent in the basics of the broad science of public health. A real public health specialist needs that and considerable depth in a particular facet of the subject, and likewise there is no acceptable term for that either.

So there is a long way to go even now, but the work is clearly moving forward now that we might have unified national standards for the entry to the specialist public health career paths. Together the various experts can then tackle the horrendous threats to public health and work together to improve the wellbeing of all of our society. The opportunities are only now being fully realized.

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