increased risk of complications. The gene for LD, located on the long arm of chromosome 16, is that encoding FOXC2, a forkhead family transcription factor involved in numerous developmental pathways.[13] In all three members of the family presented in this report, a 19 bp insertion in the FOXC2 gene was identified. To our knowledge this mutation has not been previously reported.

Distichiasis is the most consistently inherited feature of LD syndrome. Lymphoedema in this condition is highly penetrant; however, it is rarely recognised before puberty. While examining young children with distichiasis, the likelihood of them developing lymphoedema subsequently should be borne in mind and parents counselled accordingly. As there are many interesting and unanswered questions regarding the connection between various associated features in LD, further research may give us the answers. Every ophthalmologist, on recognising a case of distichiasis should take a clear family history asking about lymphoedema of the lower limbs and the other features listed above, and arrange molecular genetic investigations to look for mutations in the FOXC2 gene. The family should also be referred for genetic counselling as there can be associated cleft palate and/or congenital heart disease. Any families where distichiasis but no lymphoedema occur are also of great interest, as it is not clear whether mutations in the FOXC2 gene would account for this symptom alone.

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BB Patil[1], R Bell[2], G Brice[2], S Jeffery[2] and SP Desai[1] and the Lymphoedema Research Group[2]

[1]Department of Ophthalmology
Doncaster Royal Infirmary, Doncaster UK

[2]Medical Genetics Unit
St George's Medical School, London, UK

Correspondence: SP Desai
Tel: +44 1302 366666 Ext: 3325
E-mail: desaisp@hotmail.com

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Sir,

CJD and intraocular surgery

We read with interest the clinical study by Leslie *et al*,[1] in the May (2003) issue of Eye. The authors eloquently showed the contamination results before and after the use of an automated rinsing system. They hypothesized that the contamination may be the cause of

endophthalmitis found in their unit, but another observation can be made from their results. The contaminants noted by the authors included organisms, lens capsule and cells. The question of whether these organisms were viable or not is certainly significant but the finding of lens material may pose a risk of transmission of Creutzfeldt–Jakob disease (CJD).

CJD is a transmissible human spongioform encephalopathy characterized by the presence of abnormal prion protein. Different forms of the disease exists, for example, sCJD (sporadic), iCJD (following iatrogenic spread), fCJD (familial), and vCJD (variant, following the consumption of infected beef). Several authors have commented on CJD transmission from ocular transplants, for example, cornea[2] or sclera.[3] However, intraocular surgical transmission is of concern due to the sheer volume of cataract surgery performed. Hogan *et al* showed a 10-fold increase in prion titres in the lens in scrapie-infected hamsters once they had become neurologically symptomatic. In fact, levels in the lens were similar to levels in the cornea prior to the onset of neurological symptoms but became significantly higher than those in the cornea after the onset of symptoms, retina having the highest titres.[4] Intraocular 'inoculation' by phacoemulsification and irrigation-aspiration handpieces contaminated by lens material certainly would provide a route for transmission.

The recent steps taken by the Department of Health in reviewing sterilizing units probably triggered by the outbreak of vCJD is certainly welcomed.[5] The risk of intraocular transmission in anterior segment surgery is probably greater from sCJD than vCJD due to the age of the patients affected by the respective conditions. Tissue distribution of prion protein from human eyes has as yet to show levels of prion in the cornea or lens,[6,7] but the authors concluded that the inability to detect prion protein in the cornea or lens could not be taken as evidence for the absence of infectivity in these tissues.[7]

Since prions adhere strongly to metal surfaces and are resistant to many sterilization processes,[8] the reduction of contaminants as illustrated by this report in patients with known or suspected CJD[9] and the use of single-use equipment[5] (eg disposable simcoe) will reduce risks of transmission further.

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JS Mehta and R Osborne

The Western Eye Hospital
Marylebone Road, London, UK

Correspondence: JS Mehta
9 Sandringham Court
King & Queen Wharf
Rotherhithe Street
London SE16 5SQ, UK
Tel: +44 7980691396
Fax: +44 8701316622
E-mail: jodmehta@hotmail.com

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Sir,

Radiation retinopathy secondary to radiotherapy for a lower lid BCC

Basal cell carcinoma (BCC) is the most common malignant eyelid tumour. Although it may be treated by external beam radiotherapy, severe radiation retinopathy in this setting is extremely rare.[1]

Case report

A 39-year-old lady was successfully treated with 45 Gray (Gy) of external beam radiotherapy in nine equal fractions for a lateral lower lid BCC. An internal eye shield was used to protect the globe. After 8 years, her optometrist referred her to the local eye clinic after finding abnormal peripheral retinal blood vessels. After excluding diabetes and hypertension, radiation