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Sir,

We thank Winder and colleagues for their interest in our paper.[1] We used a 10-point visual analogue score in our study, which was completed by each patient with the help of a nurse on the day following surgery. This did not cause any difficulty. A 5-point score is a reasonable suggestion; however, patients do occasionally experience extreme pain on administration of anaesthetic that may not be adequately illustrated by a shorter scale.

It would be very interesting to compare the discomfort and surgical complications associated with cataract surgery under what we believe to be the best injectable method of local anaesthesia, i.e. the sub-Tenon's technique, with those under topical anaesthesia that provides no akinesia. A previous study comparing retrobulbar anaesthesia with sub-conjunctival anaesthesia found no difference in the complication rate between these two techniques.[2] Our study, and that of Winder and colleagues, has shown that it is possible to provide pain-free cataract surgery under local anaesthesia. The onus is therefore on all ophthalmologists to ensure that their patients do not suffer discomfort during cataract surgery, whatever the chosen method of anaesthesia. An ideal subject for clinical audit!

References

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Sir,

We would like to comment on some points raised by the interesting paper on congenital hypertrophy of the retinal pigment epithelium (CHRPE) and familial adenomatous polyposis (FAP) by Reck *et al*.[1] The aim of their study was to correlate mutation site with CHRPE status in a group of patients with FAP. We feel their discussion of the previous literature on CHRPE and mutation site simplistic and thus misleading, making their conclusions more secure. Previous studies[2–4] have supported a CHRPE-positive phenotype with mutations upstream of exon 9 to codon 1387 but not with mutations before exon 9. However, mutations in exon 9 can result in either a CRPHE-positive or -negative individual within the same family,[3,4] but this was not mentioned in Reck *et al*.'s paper. Identical FAP mutations in unrelated patients can also demonstrate marked variability.[5]

Reck *et al*. (p. 300) make the point that the 'CHRPE status provides a guide to the likely position of the causative mutation', but we feel the CHRPE status of the family should be determined. In our paper[6] on the value of CHRPE in screening for FAP it was found that there was a large intrafamilial variation in the incidence of CHRPE in individuals with the disease. Reck *et al*.'s study findings and conclusions were based on isolated cases. Readers should be aware of intrafamilial variation and that exon 9 mutations can result in a CHRPE-negative or -positive individual within the same family. 'Considerable reassurance' (Reck *et al*., p. 300) would not be gained from a CHRPE-negative member in these families.

We therefore feel that the role of CHRPE in screening individuals at risk of FAP needs careful consideration. We do not feel on the basis of ocular screening that individuals at risk of FAP should be excluded from colonoscopic screening unless the CHPRE status of the family has been firmly established. Mutational analysis, although expensive and time-consuming, will hopefully provide the most valuable tool for screening.

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Sir,

The comments made by Willoughby and Hickey-Dwyer are perfectly valid. Our discussion was deliberately simplistic because we have published a more detailed paper with the molecular genetic analysis and refer to this paper in the text.[1] In our discussion we make the observation that in a family with CHRPE, an at-risk person negative for CHRPE has 'a reduced risk of carrying the defective gene'. We also make the point that 'the only test which is 100% certain to exclude an individual from carrying the gene is mutation analysis'. Since the advent of molecular genetic testing in FAP has allowed the carrier status to be determined to a high degree of certainty in the vast majority of families, the clinical value of an ophthalmic examination is less clear. It is still interesting, however, to observe correlations between genotypes and phenotypes. The variation between family members and between the two eyes of a single individual presumably indicates that the development of CHRPE is not solely dependent on the underlying constitutional mutation but on a second somatic event in the retinal pigment epithelial cells, like many of the other extra colonic manifestations of this disease.