

S. Winder ✉  
Department of Ophthalmology  
Royal Hallamshire Hospital  
Glossop Road  
Sheffield S10 2JF, UK

Sir,

We thank Winder and colleagues for their interest in our paper.<sup>1</sup> 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.<sup>2</sup> 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

1. Briggs MC, Beck SA, Esakowitz L. Sub-Tenon's versus peribulbar anaesthesia for cataract surgery. *Eye* 1997;11:639-43.
2. Redmond RM, Dallas NL. Extracapsular cataract extraction under local anaesthesia without retrobulbar anaesthesia. *Br J Ophthalmol* 1990;74:203-4.

L. Esakowitz  
M.C. Briggs  
S.A. Beck  
Eye Department  
Royal Alexandra Hospital NHS Trust  
Corsebar Road  
Paisley PA2 9PN, UK

M.C. Briggs ✉  
St Paul's Eye Unit  
Royal Liverpool University Hospital  
Prescot Street  
Liverpool L7 8XP, UK

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.*<sup>1</sup> 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<sup>2-4</sup> 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,<sup>3,4</sup> but this was not mentioned in Reck *et al.*'s paper. Identical FAP mutations in unrelated patients can also demonstrate marked variability.<sup>5</sup>

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<sup>6</sup> 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 variability 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.

#### References

1. Reck AC, Bunyan D, Eccles D, Humphry R. The presence of congenital hypertrophy of the retinal pigment epithelium in a subgroup of patients with adenomatous polyposis coli mutations. *Eye* 1997;11:298-300.
2. Bunyan DJ, Shea-Simonds J, Reck AC, Finnis D, Eccles DM. Genotype-phenotype correlations of new causative APC mutations in patients with familial adenomatous polyposis. *J Med Genet* 1995;32:728-31.

3. Olschwang S, Turet A, Laurent-Puig P, *et al.* Restriction of ocular fundus lesions to a specific subgroup of APC mutations in adenomatous polyposis coli patients. *Cell* 1993;75:959-68.
4. Caspari R, Olschwang S, Friedl W, *et al.* Familial adenomatous polyposis: desmoid tumours and lack of ophthalmic lesions (CHRPE) associated with APC mutations beyond codon 1444. *Hum Mol Genet* 1995;4:337-40.
5. Groden J, Gelbert L, Thiveris A, Nelson L. Mutational analysis of patients with adenomatous polyposis: identical inactivating mutations in unrelated individuals. *Am J Hum Genet* 1993;52:263-72.
6. Hickey-Dwyer MU, Willoughby CE. Assessment of the value of congenital hypertrophy of the retinal pigment epithelium as an ocular marker for familial adenomatous polyposis coli. *Eye* 1993;7:562-4.

C.E. Willoughby ✉  
M.U. Hickey-Dwyer  
St Paul's Eye Unit  
Royal Liverpool University Hospitals  
Prescot Street  
Liverpool L7 8XP, UK

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.<sup>1</sup> 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.

## Reference

1. Bunyan DJ, Shea-Simonds J, Reck AC, Finnis D, Eccles D. Genotype-phenotype correlations of novel causative APC gene mutations in patients with familial adenomatous polyposis. *J Med Genet* 1995;32:924-31.

Anne C. Reck ✉  
David Bunyan  
Diana Eccles  
R. Humphry  
Human Genetics  
Level G  
Princess Anne Hospital  
Coxford Road  
Southampton SO16 5YA, UK

Tel: +44 (0)1703 794172  
Fax: +44 (0)1703 798416  
e-mail: de1@soton.ac.uk

Sir,

We have very mixed feelings about The National Survey of Local Anaesthesia for Ocular Surgery: Early Report from the Royal College of Ophthalmologists.<sup>1</sup>

We are pleased that the survey was able to show (as have others<sup>2</sup>) that 'routine' pre-operative investigations before local anaesthesia are unnecessary, and feel that the Royal College of Ophthalmologists should now revise their original guidelines<sup>3</sup> to reflect this.

We have major reservations about methodology, which is seriously flawed from at least two aspects:

1. The lack of standardised definition of method of anaesthesia, in particular failing to define peribulbar and retrobulbar injections, will have caused confusion. A peribulbar injection is defined as a deliberate extraconal injection<sup>4</sup> and a retrobulbar as a deliberate intraconal injection.<sup>5</sup> We are aware that many doctors who administer ophthalmic local anaesthesia do not follow these definitions, and that much of the outcome data comparing these two techniques is therefore suspect.
2. There is a large variation in the incidence of adverse effects between the two phases of the survey. The survey reports the incidence of systemic adverse events at 0.9% (0.1% severe) in the first week when all cases were to be reported and 0.19% (0.06% severe) over the remainder of the 3 months when only adverse events were to be reported. We feel that this can only be explained on the basis of under-reporting and that the data from the second 3 month period cannot be relied on.

We are most concerned about the results of the survey. If the adverse event data from the first week are accurate then local anaesthesia for ocular surgery as currently practised in

the UK is an unsafe procedure. In the first week 3.5% of patients had either an 'orbital' (2.6%) or a 'systemic' (0.9%) adverse effect; 0.28% of patients had a severe adverse event. Taken at face value this 3.5% risk makes local anaesthesia the single highest risk to the patient's health or sight, comparable to the risk of vitreous loss or endophthalmitis, in cataract surgery.<sup>6</sup> A risk of this magnitude must be disclosed and discussed with the patient, and it is our belief that no sensible patient would choose to run this risk unless general anaesthesia was absolutely contraindicated. This being said, these figures do not accord with our own experience, nor with that of many other surgeons who also electively perform cataract surgery under local anaesthesia, nor with other published results.

It is clear that rather than settling issues, the survey may have actually raised more serious issues. Careful thought needs to be given to whether this is a valid survey and ought to be accepted, or repeated if flaws can be identified and addressed. If the survey is valid it is necessary to identify the reasons for and remedy the high rate of adverse effects. We look to the Royal College of Ophthalmologists to undertake this.

There is a further issue, which is probably the main issue in ophthalmic anaesthesia from the patients' perspective. This is 'What is the safest anaesthetic for the procedure?' This can only be answered by comparing local anaesthesia with general anaesthesia and comparing the various techniques of local anaesthesia. We would hope that any future survey could be structured so as to answer this question.

## References

1. Eke T, Thompson JR. National survey of local anaesthesia for ocular surgery: early report. London: Audit Committee, Royal College of Ophthalmologists, 1997.
2. Walters G, McKibbin M. The value of pre-operative investigations in local anaesthetic ophthalmic surgery. *Eye* 1997;11:847-9.
3. Report of the Joint Working Party on Anaesthesia in Ophthalmic Surgery. London: Royal College of Anaesthetists and College of Ophthalmologists, March 1993.
4. Davis DB, Mandel MR. Posterior peribulbar anaesthesia: an alternative to retrobulbar anaesthesia. *J Cataract Refract Surg* 1986;12:182-4.
5. Hamilton RC. Techniques of orbital regional anaesthesia. In: Smith GB, Hamilton RC, Carr CA, editors. *Ophthalmic anaesthesia: a practical handbook*. 2nd ed. London: Arnold, 1996:112-3.

6. Powe NR, Schein OD, Gieser SC, et al. Synthesis of the literature on visual acuity and complications following cataract extraction with intraocular lens implantation. The Cataract Patient Outcomes Research Team. *Arch Ophthalmol* 1994;112:239-52.

Girish Kamath ✉  
Somdutt Prasad  
Louis Clearkin  
Department of Ophthalmology  
Arrowe Park Hospital  
Wirral  
Merseyside L49 5PE, UK

Sir,

I read with interest the paper by Claoué and colleagues on the relative frequencies of ophthalmic disease in Moorfields Eye Hospital and one of its outreach clinics.<sup>1</sup> Whilst it may be useful for an individual department to examine its referrals in terms of proportion percentages, caution should be used before assuming similar proportions would exist in other hospital populations as there are clearly many factors that play a part in whether a patient is referred to a particular unit.

In order to plan services appropriately for a population, a combination of epidemiological prevalence studies and demand incidence work is required. Such an example of the latter type of study was performed at Nottingham in 1989/90,<sup>2</sup> which included all presentations of eye disease in a balanced population of 36 000 utilising verified data from GP attendances and Eye Casualty.

It is interesting to compare some 'proportions'. In Nottingham, the demand incidence for cataract (at 1.9 per 1000 population per year) was approximately twice that of glaucoma and suspect glaucoma, whereas in the Moorfields series referral for cataract was 3.3 times as common in 1991 and 3.6 times as common in 1993. However, the ratio of glaucoma to age-related macular degeneration was similar in the two studies, at 1.27 and 1.16 in the Moorfields series versus 1.29 in the Nottingham series. This suggests, for whatever reasons, a 'bias' towards cataract in the Moorfields patient population. This bias may be greater than it appears as some of the Nottingham patients presenting to their GPs may not have been referred to the hospital service. Indeed, only 29% had an acuity less than 6/12 in both eyes and 33% had 6/12 or better in both eyes.

The purchaser/provider split renders recent data from many Units suspect and changes in the pattern of disease presenting to ophthalmologists must be identified by the use of the appropriate methodology, i.e. by demand incidence studies.