CORRESPONDENCE

The South Asian cataract management study

EDITOR,—In reading the preliminary report evaluating the clinical outcome of intraocular cataract extraction (ICCE) with aphakic correction either via anterior chamber intraocular lens (AC IOL) implants or aphakic spectacles in a developing country setting I was given cause for alarm with regard to the assessment of the differences in visual outcome.

While the study is ongoing, I think it is important to recognise that a varying percentage (16–25%) of standard ICCE patients in a developing country setting will lose their spectacles and have problems adapting to the magnified image, forcing some patients not to wear them at all.1,2 By the same token, the quality of vision and life enjoyed by those patients with AC IOL implants is likely to be superior to those patients relying on aphakic spectacles, or none at all. However, the report’s stress on the stringent use of visual acuity measures alone and its conclusion that ‘There was no significant difference in visual outcome between study groups’, fails to address adequately the variation attributable to spectacle and non-spectacle using aphakic patients in determining the overall differences in visual outcomes between the two study groups.3 In regard, recent instruments have been developed to measure both the preoperative and postoperative visual function and quality of life aspects of cataract patients within the context of a developing country setting offering an avenue for further exploration of the relation between visual acuity and overall patient satisfaction with the visual outcome of cataract surgery.4

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Reply

EDITOR,—The south Asian cataract management study (SACMS) was not designed to address the issue of quality of life. There is certainly no longer any controversy that the introduction of an IOL improves the quality of vision compared with aphakic spectacle correction. This is reflected in the significant increase of IOL implantations being performed in most developing countries every year.

The authors of the SACMS are well aware of the excellent work being undertaken at the Aravind Eye Hospital (Madurai Intraocular Lens Study—MIOLS) addressing the quality of life issue in cataract surgery with and without IOL.

The SACMS group has chosen to focus on documentation of the clinical aspects of surgery with and without AC IOL implantation survival with a well proved surgical technique after implantation of a modern generation AC IOL. We are confident that the outcome of the SACMS, MIOLS, and other similar randomised prospective studies will give comprehensive conclusive data to guide surgeons and clinical centres on approaches to cataract surgery in these and other settings.

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Peters’ anomaly

EDITOR,—I read with interest the recent letter by Saitoh et al describing an infant with both Peters’ anomaly and anterior chamber dysgenesis.1 Unfortunately the discussion which follows the case description is marred by a failure to mention some significant discoveries which have been made in this field over the past few years. I would like to bring to their attention the finding of a deletion of the PAX6 gene in a child with aniridia and Peters’ anomaly, and an intragenic mutation of the same gene in a family with heterogeneous anterior segment malformations including Peters’ anomaly.2,3 Deletions and mutations of the PAX6 gene were sought in these patients for the following reasons. The PAX6 gene was known to be deleted or mutated in aniridia, and cases of aniridia with Peters’ anomaly had been described.4,5 Furthermore, deletions and mutations of Pax-6, the mouse homologue of PAX6, had been found in the Small eye mouse which phenotypically resembles aniridia and Peters’ anomaly.6,7 The PAX6 gene plays a crucial role in development as illustrated by the discovery of mutations in homologue genes in humans, zebrafish, Drosophila, Caenorhabditis elegans, and sea urchin. Recent experiments using targeted expression in Drosophila led to the production of supernumerary ectopic eyes giving support to the idea that PAX6 is the ‘master control’ gene for eye formation.8

The expression pattern of PAX6 has been investigated in both human and mouse.9–11 These studies found expression in ocular progenitors derived from neuroectoderm and head surface ectoderm which give rise to the corneal epithelium, the lens, the posterior layers of the iris, the ciliary body, the retina, and the optic nerve. The primary defect in aniridia and Peters’ anomaly caused by PAX6 mutations must therefore lie within these tissues and fits well with the spectrum of clinical abnormality seen in these disorders—that is, corneal opacification at the subepithelial level, incomplete separation of the lens, persistent lens stalk, central corneal leucoma, iridocorneal adhesions, iridocorneal adhesions, iris hypoplasia, cataract, macula dysplasia, and convergent strabismus. The line of migration of midbrain neural crest cells has been found in the Uchida rat which has an intragenic mutation of the rat homologue of PAX6.12 However, this does not affect the arrival of neural crest cells at the eye and since the PAX6 gene is not known to be expressed in this tissue it is unlikely to be the site of primary pathology.

Although the concurrence of Peters’ anomalies and anterior chamber dysgenesis is an interesting finding it may be a small part of a larger picture that is the case with the usual genetic abnormalities and has hitherto remained undetected. I would like to suggest that an alternative explanation is that deletion in a single gene can cause a broad range of phenotypes with the primary abnormality residing within the expressing— that is, neuroectoderm and head ectoderm, and secondary abnormalities occurring within tissues with which PAX6 is expressed. This principle should still hold true for cases of Peters’ anomaly not due to abnormality of the PAX6 gene. Some of these may result from a primary disorder of neural crest cells but this has yet to be proved.

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Reply

EDITOR.—We also have an interest in the recent discoveries of the association between anterior segment anomalies and PAX6 locus. At present, we are analysing the PAX6 locus of an affected family in which the PAX6 gene is deleted. By using the polymerase chain reaction, single strand conformation polymorphism, and sequencing analysis methods we are able to locate the mutation in the PAX6 gene. Jordan’s alternative explanation is interesting, but it is difficult to accept.
but the reports he presented were about the case of Peters’ anomaly with posterior embryotoxon, small eye, or aniridia. Unfortunately there has been no report about PAX6 analysis of a case of Peters’ anomaly with Axenfeld’s anomaly yet. He pointed out that it might be premature to conclude that the concurrence of Peters’ anomaly and Axenfeld’s anomaly was the usual case. We have started a prospective study of gonioscopic examination of Peters’ anomaly as we first examined a case with this combination from the same standpoint as his. We have already examined three infants with Peters’ anomaly, under general anaesthesia, and one adult patient, under topical anaesthesia. We have found Axenfeld’s anomaly in two infants and one adult patient including our first case (Table 1). Two infants had two or three iris strands attached to the posterior embryotoxon, but without careful examination under general anaesthesia these strands would have been overlooked. Though our study is still in progress, our conclusion that careful examination of patients with Peters’ anomaly with a gonioscopy under general anaesthesia would reveal latent Axenfeld’s anomaly looks like being confirmed.

Table 1 Examined cases of Peters’ anomaly. Cases 1, 2, and 3 had Axenfeld’s anomaly.

<table>
<thead>
<tr>
<th>Case No</th>
<th>Age at first examination (months)</th>
<th>Sex</th>
<th>Ocular tension</th>
<th>Gonioscopic findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5</td>
<td>Male</td>
<td>R12</td>
<td>Several iris strands attached to the posterior embryotoxon in the temporal and upper parts of the right anterior chamber</td>
</tr>
<tr>
<td>2</td>
<td>3</td>
<td>Female</td>
<td>R15</td>
<td>Two iris strands attached to the posterior embryotoxon in the temporal lower parts of the right anterior chamber</td>
</tr>
<tr>
<td>3</td>
<td>22</td>
<td>Female</td>
<td>L15</td>
<td>Closed angle.</td>
</tr>
<tr>
<td>4</td>
<td>37</td>
<td>Male</td>
<td>Treated with β blocker</td>
<td>Typical Axenfeld’s anomaly</td>
</tr>
</tbody>
</table>

Other anomalies | Anesthesia
---|---
Microcornea of both eyes | General
Microcornea of the right eye, congenital cataracts of both eyes | General
Microcornea | General
None | Topical

This book is written by a patient who suffers from Stocker syndrome and is intended to provide an informative guide to fellow sufferers and their families in the light of the many difficulties this remarkable individual has faced. It covers the articular, auditory, and orofacial features in addition to the severe ophthalmic problems, and the appendix provides some useful references and contact addresses for patients. The recent subclassifications of this disorder are not alluded to, but with the current molecular genetic advances in this field, the prenatal and postnatal testing mentioned by the author will be available to the majority of these patients within a year.

The high incidence of mitral valve prolapse referred to has not been confirmed by later studies and the routine screening for this has now been abandoned in our unit. Similarly, urinary hydroxyproline measurement has not been accepted as a reliable diagnostic or screening test.

This book is clearly written, easily read, and fulfills its role admirably. The chapter dealing with a sighted person’s interaction with the visually impaired is excellent.

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BOOK REVIEW


Wendy Hughes is a co-founder of the Sticker syndrome patient support group, the inaugural meeting of which was held in 1994 and attended by Gunnar Stickler. Thereafter, annual meetings have been coordinated by the author.

This book is written by a patient who suffers from Stickler syndrome and is intended to provide an informative guide to fellow sufferers and their families in the light of the many difficulties this remarkable individual has faced. It covers the articular, auditory, and orofacial features in addition to the severe ophthalmic problems, and the appendix provides some useful references and contact addresses for patients. The recent subclassifications of this disorder are not alluded to, but with the current molecular genetic advances in this field, the prenatal and postnatal testing mentioned by the author will be available to the majority of these patients within a year.

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MARTIN SNEAD

OBITUARY

A G Cross

One of the leading figures in British ophthalmology during the three decades following the second world war, AG Cross died recently aged 87. Legendary for his voracious appetite for work, both clinical and administrative, he enjoyed the rare distinction of having served both as dean of St Mary’s Medical School and then later of the Institute of Ophthalmology. He was a quiet and unassuming man, but behind his mild exterior appearance there was a determined and well organised character with a talent for committee work. This was facilitated by a remarkable ability to recall what had been said at a previous meeting—a gift not possessed by the average member.

Alexander Galbraith Cross (Alex, to his family) was born on 29 March 1908 in Wimbledon where he was to spend the whole of his life except for war service. He was educated at King’s College School, Wimbledon and Gonville and Caius College, Cambridge. He entered St Mary’s Medical School in 1931 having obtained his BA Cambridge and qualified in 1933. After residencies in the professorial medical unit and then the surgical unit (under Professor Aubrey Pannett, the uncle of his future bride), he passed his primary FRCS and later the final in general surgery, there being no ophthalmological FRCS in those days. There followed a residency at Moorfields Eye Hospital (then the Royal London Ophthalmic Hospital).

In 1939 he married Eileen Longman Currie, one of identical twin daughters of Dr HB Currie of Lis, Hants.

During the war he served as a wing commander in the RAFVR 1941–6, being Adviser in Ophthalmology South East Asia Air Forces. When peace came he was appointed to St Mary’s and Moorfields, which hospitals he served till his retirement in 1973.

He held numerous other consultant hospital appointments during his career and was also consultant ophthalmologist to the Royal Navy, to St Dunstan’s, and to the Royal National Institute for the Blind.

In 1975–7 he was President of the Ophthalmological Society of the United Kingdom.

He was a busy man yet he was always courteous and helpful to all, especially his juniors to whom he gave much valuable advice. His clinical work was soundly based on his training and a spell as pathologist at Moorfields. His surgery was straightforward, competent, and of legendary speed; residents, entering the theatre having just scrubbed up, were known to encounter him leaving the theatre having done the first case.

Always keen on sport, he played rugby at first class level as a young man and later enjoyed squash, tennis, and golf. But his principal hobby was gardening, and an unexpected secondary accomplishment was tapestry as visitors to his home were bound to notice.

He is survived by his devoted wife, daughter, and grandchildren.

REDMOND SMITH