conus. We, however, feel this is highly unlikely, as the donor cornea in that eye came from a 51-year-old white female who had no known history of any ocular disease. Also following surgery the donor cornea was crystal clear with minimal astigmatism and 20/20 vision with spectacle correction.

Eighteen years following the corneal grafting the cornea started showing changes typical of keratoconus, including increase in myopic oblique astigmatism, corneal protrusion and thinning, and the development of subepithelial and stromal scarring with a reduction in best corrected visual acuity to 20/400. The histopathological changes also showed changes consistent with keratoconus, including abnormalities in the basal epithelium, breaks in basement membrane, duplication and thickening of Bowman’s layer, and abnormal stromal kerocytes with accumulation of granular intra- and extracellular material. It seems unlikely that the donor cornea, which was grafted at age 51, would not have shown keratoconus changes at that time and started showing changes 22 years later.

Findings similar to ours have been reported before. Unfortunately we do not have any information on the recipient of the fellow donor eye, as the surgery occurred 22 years ago and such information was not available. It is also not unusual for abnormal host factors to affect donor tissue, resulting in the recurrence of original pathology, as is seen in lattice macular and granular dystrophy.⁴

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References

Book reviews


As expected, this book reflects the competence, enthusiasm, and integrity of its editor. The whole book is manageable in size and is largely up to date—always difficult in a textbook because of the time taken in publication. The comments on intraocular lenses show a careful balance of views. There is no reference to the use of viscoelastic substances because this development has been so recent.

Many of the specialised chapters have been written by invitation, but each has been reviewed by another author before being passed for publication.

There is a thoughtful introduction which reflects the editor’s own experience and contains much useful advice. The emphasis in other chapters is not always directly applicable worldwide. The text is aimed at the US ophthalmologist. Proprietary names which are used are not always known elsewhere. A statement that ‘Local anaesthesia remains the favorite method of most ophthalmic surgeons for cataract surgery’ would not be so readily accepted in Europe. Few intraocular lens implantations would ‘last between 1 and 2 hours in duration’ on this side of the Atlantic. It continues to surprise me that in the United States local anaesthesia is so often advised as the method of choice, particularly when operations of 1–2 hours are being described. The quality of general anaesthesia must be very different from that in the United Kingdom, if one of the authors lists among the advantages of local anaesthesia that total operating room time is decreased. In my experience induction and recovery from general anaesthesia is rapid and take place outside the operating room. The patient comes into the operating room ready for surgery with an uncongested soft eye.

The chapter on fundamental surgical principles is excellent and provokes thought. There is a welcome economy of words, which helps towards clear understanding. Seidel’s test is well described, but would seem better placed in a chapter on postoperative management. A useful table on the relationship between magnification, diameter, and depth of field of a Galilean microscope seems misplaced in the middle of this piece of text.

Instruments and sutures are described in another excellent chapter. Intraocular infections are also well covered. The chapter on ophthalmic conditions requiring prompt care is valuable, but many of the conditions are not surgical.

The quality and style of the chapters on surgical disorders shows some variety as is to be expected in a multi-author text. In some chapters the references are sequential, following the text, but in others they have been listed alphabetically. When some of the writing is so good it is annoying to come across loose comments such as ‘even after the patient is 35 to 40 years of age or older’ or ‘globe of formed vitreous’. There are many ophthalmic surgeons who will not know what a ‘frisbee’ is and they would not find its description in a medical dictionary. The ‘Sheets glide’ is mentioned but not described or illustrated. I cannot agree that ‘one of the most important advances in cataract surgery has been the introduction of anterior vitrectomy for vitreous prolapse’.

The first 90 pages in the chapter on the surgery of the orbit consist of clinical manifestations and are rather heavy reading. The last 25 pages of this chapter give only a broad guide to surgical method. The proportion seems wrong and this chapter is not as useful to a surgeon needing technical advice.

Since the beginning of this century ophthalmologists have made major contributions to clinical genetics. The papers presented at the symposium held in Jerusalem in September 1981 give some indication of the areas of particular interest at the present time, interest that is not surprising when, as stated by François in the first paper, 50% of blindness before the age of 6 years is inherited, and of the entire blind population (in a developed country) about one-third has a genetic basis.

The symposium was divided into several sections, covering the following topics, in each of which a number of papers were presented: retinal pigment epithelium and experimental retinal degeneration, retinitis pigmentosa, therapy of gyrate atrophy, retinal degenerations, albinism, lysosomal storage disease, corneal storage diseases and dystrophies, connective tissue diseases, syndromes involving the eye, epidemiology of blindness, and retinoblastoma. Certain of these topics deserve further comment.

Mette Warburg, of Copenhagen, gave the first Franceschetti Memorial Lecture on 'Diagnostic precision in microphthalmos and coloboma of heterogeneous origin,' and produced a precise diagnosis in almost half of 86 cases of microphthalmos and coloboma in mentally retarded patients. This and other papers published elsewhere by the same author throw considerable light on this difficult and heterogeneous group of ocular abnormalities.

Five papers are concerned with the retinal pigment epithelium in tissue culture, a relatively recent and fruitful method of studying this important cell layer, and one which could elucidate the role of this layer in retinitis pigmentosa and related disorders. The frequencies of the different genetic (and possibly nongenetic) forms of retinitis pigmentosa are analysed in 4 papers, and a comparison of the results indicates the difficulties inherent in this type of analysis, the results of which are of considerable practical value in genetic counselling. It would appear that about half the patients with this common group of disorders have no family history; however, this finding is open to differing interpretations.

The optic and otic neurological abnormalities in albinism are reviewed by Witkop and his coworkers, and their paper is followed by one on the diagnoses in a series of 201 albinos. This series includes 3 families where a man with X-linked ocular albinism has married a woman with autosomal recessive ocular albinism, tyrosinase-negative ocular albinism, and yellow-mutant ocularalbinism respectively.

Two papers on the Stickler syndrome emphasise the frequent occurrence of retinal detachment and other ocular changes in this common autosomal dominant progressive arthro-ophtalmopathy, with its variable expressivity and incomplete penetrance. Ophthalmologists should be aware of this syndrome with its high incidence of retinal detachment that is technically difficult to treat successfully. In the last section there were 3 papers on the genetics and treatment of retinoblastoma. The gene for the 13 deletion form of retinoblastoma (on region 13q14) is closely linked to that for esterase D, and estimation of esterase D activity in blood and in fetal cells is a useful indicator of the deletion form of the tumour. A preliminary report suggests that the gene for nondeletion dominantly inherited retinoblastoma segregates with one of the 2 esterase D alleles.

These proceedings can be highly recommended to anyone interested in reading about recent advances in ophthalmic genetics. Every fellowship candidate is advised to have glanced through the contents and to have read selected papers.

Barrie Jay


Premature fusion of the skull sutures can occur with involvement of only one suture (the so-called simple craniosynostoses) or with involvement of multiple sutures, in which case it usually represents one element of a more complex congenital condition such as Crouzon's, Apert's, or Pfeifer's syndromes. These cases are almost invariably associated with bicornal synostosis, but with the addition of other skull abnormalities and with hydrocephalus to provide a driving force that exceeds that of normal brain growth, grotesque abnormalities of head shape, culminating in the clover-leaf skull, can be produced.

Craniosynostosis is of interest to the ophthalmologist for several reasons. The maxillary recession and the failure of advancement of the supraorbital ridge that is seen in bicornal synostosis may leave the patient with shallow orbits, the eyes like frog's eyes, scarcely contained within them. Defects of conjugate gaze are frequent in such cases, and where the exposure is severe there may be failure of the lids to cover the globes. Raised intracranial pressure due