

## Editorial

### Cataracts in children with uveitis

Kanski, who has contributed significantly to the management of children with uveitis, now focuses on their 'complicated' cataracts, giving his authoritative views in this issue of *BJO*.

The frequency of uveitis in children is relatively low, approximately 8% of all cases of uveitis occurring in children under the age of 16.<sup>1</sup> Large series of patients with uveitis have been described by Kimura and Hogan,<sup>2</sup> by Perkins,<sup>3</sup> and by Jütte *et al.*<sup>4</sup>

In children, juvenile rheumatoid arthritis (JRA), also known as Still's disease, is the most common systemic association of chronic uveitis. Of 340 cases of systemic uveitis in childhood Kanski and Shun-Shin<sup>5</sup> found that 277 cases were associated with JRA. Juvenile ankylosing spondylitis (46 patients), juvenile psoriatic arthritis (nine patients), sarcoidosis (three patients), tuberculosis, Reiter's syndrome (one patient) and the Voght-Koyanagi-Harada syndrome (one patient) were the other underlying systemic disorders. Specific uveitis entities unassociated with a systemic disorder include pars planitis, Fuch's heterochromic cyclitis, and juvenile chronic iridocyclitis.<sup>6</sup>

In chronic anterior uveitis the anterior chamber reaction is often not gross but can persist for years. In most cases the onset is insidious and the disease is only discovered when the child is noted to have a difference in colour between the two eyes, a difference in the shape of the pupil, the onset of strabismus, or a white mark on the pupil or the cornea. Some are detected when being examined routinely after the diagnosis of arthritis is made.<sup>7</sup> Seventy per cent are bilaterally affected and 75% of the patients with JRA and uveitis are girls.<sup>8</sup>

Chronic uveitis in children often results in band keratopathy, cataracts, vitreous opacities, macular oedema and pucker, and glaucoma which, alone or in concert, progressively and insidiously impair vision.

Before starting treatment an underlying disorder should be excluded. It is preferable not to start systemic steroids before any immunological investigations are undertaken especially where tuberculosis and sarcoidosis may be suspected.

Steroids are still the mainstay of the management of uveitis. Though the chronic anterior uveitis can persist for many years in most cases it can be at least partly controlled by topical, periocular, or systemic steroids. Mydriatics may prevent but will rarely break down the posterior synechiae. It is usually best to give short acting mydriatic drops; they can, especially before synechiae have formed, be used at night causing fewer problems from the cycloplegia and mydriasis. The severe side effects of prolonged systemic steroids have often been the reason for using cytotoxic drugs in particularly severe cases of uveitis in adults: Dinning<sup>9</sup> however felt they should not be used in children. Martenet<sup>10</sup> on the other hand was impressed with the results of cyclophosphamide in the treatment of severe anterior uveitis in children.

One of the most frequent complications of a chronic anterior uveitis is the formation of cataracts. The term 'complicated' cataract has been used for many years<sup>11</sup> to describe the cataract which develops initially at the posterior pole of the lens as a polychromatic lustre surrounded by a cloudy haze. Later there is more extensive posterior and anterior subcapsular opacification, then the cortex is progressively involved until the cataract becomes mature.<sup>12</sup> The

posterior synechiae may stimulate local cataract formation. The prolonged use of systemic or topical steroids can be partly responsible.

The management of complicated cataracts represents a surgical challenge which has been partly met by technological developments with improvement in the visual prognosis.

Intracapsular surgery and techniques which leave the posterior capsule intact are too prone to complications to be used routinely when lensectomy can be performed. Smiley<sup>13</sup> in 1974 reported no light perception in seven of 18 eyes within 2 months of cataract extraction by needling, intracapsular extraction, or aspiration techniques. Half of the JRA cataract patients treated by aspiration, extracapsular, or intracapsular extraction by Key and Kimura<sup>14</sup> had postoperative visual acuities of less than or equal to 20/200. Diamond and Kaplan conclude that it is advisable to minimise the preoperative inflammatory reaction by steroids or indomethacin.

The technique of choice today, as described by Kanski in this issue, is probably a lensectomy with lens fragmentation/aspiration and anterior vitrectomy, either by a limbal or pars plana approach. Diamond and Kaplan<sup>15</sup> performed pars plana lensectomy and vitrectomy on 13 patients (15 eyes) with complicated cataracts and preoperative visual acuities of 10/200 or less: postoperative acuities with a mean follow-up period of 10.5 months were 20/25 or better in eight eyes, 20/70 in two eyes and 20/100 to 20/400 in five eyes.

Nolthenius and Deutman<sup>16</sup> performed lens fragmentation/aspiration and pars plana vitrectomy on 21 patients (23 eyes) with chronic uveitis and cataracts. This resulted in all cases in an improved visual acuity with a mean follow-up time of 14 months. In four eyes the visual acuity only improved to 0.3-0.4. The primary cause of decreased vision postoperatively in both series was cystoid macular oedema. Flynn *et al.*<sup>17</sup> retrospectively reviewed the results of the same technique in 10 eyes from seven JRA patients with secondary cataracts. Visual acuities improved from 20/100 or worse preoperatively to 20/60 or better postoperatively with a mean follow-up of 62 months.

From these and Kanski's series with a mean follow-up period of 5.4 years it is evident that this surgical approach results in an improved vision and can treat or prevent a number of complications.

Postoperative topical steroid treatment is necessary in most patients and a hard contact lens is often the best optical solution postoperatively. Because of the risk of synechiae and secondary membrane formation intraocular lens implantation is contraindicated.

In very small children amblyopia should not be overlooked. Failure of occlusion to improve vision can also be due to pupillary membranes, band keratopathy, cystoid macular oedema, or retinal detachment.

Kanski's paper will be read with interest by anyone involved in the management of these most challenging patients. The surgical techniques for management of complicated cataracts in uveitis have improved the visual prognosis for those patients whose vision is lost by cataract and it may also reduce the incidence of glaucoma, macular oedema, and perhaps of irreversible hypotony, and phthisis bulbi.

I CASTEELS  
D TAYLOR

- 1 Schaegel TF. *Essentials of uveitis*. Boston: Churchill, Little, Brown, 1969.
- 2 Kimura SJ, Hogan MJ. Uveitis in children. Analysis of 274 cases. *Trans Am Ophthalmol Soc* 1964; **62**: 173.
- 3 Perkins ES. Pattern of uveitis in children. *Br J Ophthalmol* 1960; **50**: 169-85.
- 4 Jutte A, Lemke L, Opitz J. Chronische Zyklitis im Kindesalter *Ophthalmologica* 1969; **157**: 169-77.
- 5 Kanski JJ, Shun-Shin GA. Systemic uveitis syndromes in childhood: an analysis of 340 cases. *Ophthalmology* 1984; **91**: 1247-52.
- 6 Kanski JJ. *Uveitis. A colour manual of diagnosis and treatment*. London: Butterworths, 1987.
- 7 Casteels I, Leys A, Missotten L. Risico op uveitis bij juveniele reumatoïde artritis en frequentie van oftalmologische controle. *Tijdschr voor Geneeskunde* 1989; **45**: 973-8.
- 8 Chylack LT, Bienvang DC, Bellows AR. Ocular manifestations of juvenile rheumatoid arthritis. *Am J Ophthalmol* 1975; **79**: 1026-33.
- 9 Dinning WJ. Treatment of uveitis. *Trans Ophthalmol Soc UK* 1981; **101**: 391-3.
- 10 Martenet AC. Résultats de l'immunodépression par cytostatiques en ophtalmologie. *Schweiz Ophthalmol* 1976; **172**: 106-15.
- 11 Becker O. *Handbuch der gesamten Augenheilkunde*. Leipzig: Engelmann, 1877.
- 12 Fisher RF. The lens in uveitis. *Trans Ophthalmol Soc UK* 1981; **101**: 317-20.
- 13 Smiley WK. The eye in juvenile rheumatoid arthritis. *Trans Ophthalmol Soc UK* 1974; **94**: 817-29.
- 14 Key SN, Kimura SJ. Iridocyclitis associated with juvenile rheumatoid arthritis. *Am J Ophthalmol* 1975; **80**: 425-9.
- 15 Diamond JG, Kaplan HJ. Lensectomy and vitrectomy for complicated cataract secondary to uveitis. *Arch Ophthalmol* 1978; **96**: 1798-1804.
- 16 Tutein Nolthenius PA, Deutman AF. Surgical treatment of the complications of chronic uveitis. *Ophthalmologica* 1983; **186**: 11-6.
- 17 Flynn HW, Davis JL, Culbertson WW. Pars plana lensectomy and vitrectomy for complicated cataracts in juvenile rheumatoid arthritis. *Ophthalmology* 1988; **95**: 1114-9.