Cataract formation is the most common vision-threatening complication resulting from uveitis that is associated with juvenile idiopathic arthritis (JIA). Surgical management is especially challenging, as cataract formation frequently occurs in young children with chronically active inflammation. Implantation of an intraocular lens (IOL) has frequently been associated with poor visual outcome as a consequence of the development of posterior synechiae, fibrous membranes, IOL deposits, persistent inflammation and hypotony. Therefore, the widely used surgical technique has been lensectomy and anterior vitrectomy, and many uveitis authorities discourage implantation of an IOL at the time of cataract extraction in these children [1, 2].

However, it is well known that visual rehabilitation in aphakia has several major disadvantages, particularly in children with chronic uveitis. Intolerance to contact lenses has been reported in 17–35% of children [1, 3]. The limiting factors include poor compliance, cost of contact lenses, pre-existent band keratopathy, and need for long-term topical steroid medication. Further problems are the high weight of spectacles in bilateral aphakia, and anisometropia in unilateral aphakia. Amblyopia is a limiting factor in vision improvement in up to 15% of the patients [4]. The surgical management of glaucoma may also be more complicated in aphakia.

Recently, a few retrospective studies have reported on positive results of cataract extraction with primary IOL implantation in selected patients with JIA-associated uveitis. Probst and Holland [5] reported on the results in 7 patients with juvenile rheumatoid arthritis (JRA)-associated uveitis who underwent phacoemulsification with IOL implantation. Although visual acuity (VA) of 20/40 or better was obtained in all eyes, the 2 patients younger than 10 years developed posterior synechiae, and 1 had persistent inflammation. Notably, no immunosuppressive drugs other than corticosteroids were given.

BenEzra and Cohen 2000 described the experience with phacoemulsification and IOL implantation in 5 JRA uveitis patients aged between 4 and 8 years. Postoperatively, retrolental membranes developed in 4 patients, and posterior synechiae in another 3. Besides corticosteroids, no additional immunosuppression was used.

In another study by Lundvall and Zetterstrom [6], cataract surgery with IOL implantation was performed in 7 children (10 eyes) with JRA-associated uveitis. All of the patients were 10 years old or younger at the time of surgery. Corticosteroids and methotrexate were inconsistently used. While VA of 20/40 or better was obtained in 7 eyes, loss of vision occurred in the only child that did not receive systemic corticosteroids or immunosuppressive drugs.

Lam et al. [7] had observed VA of 20/40 or better in 5 patients with JRA-associated uveitis of 12 years or younger after cataract surgery with posterior chamber IOL implantation. The authors concluded that the key factor for success was the long-term preoperative and postoperative control of inflammation with the use of systemic corticosteroids and additional topical corticosteroids.

The retrospective study by Kotaniemi and Penttilä published in this issue of Ophthalmic Research now analyzes the outcome after phacoemulsification with IOL implantation in 36 eyes out of 25 JIA patients in a 15-year period. The mean age at onset of uveitis was 5.8 years. Cataract developed at a mean of 2.3 years after diagnosis of uveitis, and cataract extraction was performed at a mean of 4.5 years, which reflects the typical time course in patients with JIA-associated iridocyclitis. The major information from this interesting study is that favorable visual results, defined as VA of 0.5 or better, were noted in 64%, and moderate VA results (0.3 to <0.5) were obtained in another...
er 11%. The interval VA results or details on rates of acuity changes have not been analyzed further. Rather unexpectedly, the outcome differed neither between uni- and bilateral cataract cases, nor between young children and adults.

Ten of the patients included in the study belonged to the HLA-B27-positive JIA subgroup. HLA-B27-positive JIA typically occurs with recurrent acute anterior uveitis and has a more favorable outcome after surgery as compared to the chronic type uveitis in ANA-positive oligoarthritis children. Interestingly, the visual outcome after cataract surgery with IOL implantation did not differ between the HLA-B27-positive and ANA-positive patients. Studies performed in the future may preferentially be performed with the selected subgroup of ANA-positive oligoarthritis or seronegative polyarthritis patients, known for their persistent type of complicated uveitis.

Also, few patients in this study were in adulthood already at the time of surgery, and probably with burned-out disease. Previously, more favorable results have been seen in this subgroup of patients as compared to children [5, 8].

The authors state that arthritis and uveitis were carefully treated with systemic immunosuppression and topical corticosteroids. However, uveitis in their patients continued to be active in the pre- and postoperative period. This is a critical issue, as the age of 12 years or younger, perioperative activity of uveitis and inadequate use of immunosuppressive drugs were the major limiting factors for postoperative outcome in the previous studies on IOL implantation in patients with JIA-associated uveitis [5–8]. Although anterior uveitis remained active in all of the 25 patients in this study, the authors report on VA improvement in 30 out of the 36 operated eyes. However, details on the level of VA improvement, the perioperative immunosuppression and grading of anterior chamber cells, and of the postoperative development of posterior synechiae as a consequence of inflammation are not provided. Taking into consideration the observations of the previous and current publications, the editors of Ophthalnic Research suggest that IOL implantation in patients with JIA-associated uveitis may be restricted to individuals with long-term control of inflammation before the time of surgery.

Previous studies on cataract extraction with primary IOL implantation in patients with JIA-associated uveitis pointed out that surgery is most often (80–100%) complicated by secondary capsular opacification [5–8]. As a further drawback, many of the patients required repeated Nd: YAG capsulotomies and also surgical posterior capsulotomy and vitrectomy. The observations in the study by Kotaniemi and Penttilä now suggest that primary capsulotomy should be combined with core vitrectomy in order to prevent secondary cataract development. However, previous reports have shown that secondary capsular opacification and retrolental membrane formation may develop despite the primary capsulotomy and vitrectomy [6, 8].

Although the study by Kotaniemi and Penttilä included quite a large group of patients, it is limited by its retrospective design and the lack of a control group. Several important issues remain to be answered in the future by prospective controlled studies. The most important question to be answered is in whom the IOL implantation might be safe. The level of VA deterioration that is indicating surgery in uveitis children with either uni- or bilateral cataract should be better defined, and herein the development of amblyopia must be considered. How long should uveitis be quiet before surgery? What is the optimal perioperative anti-inflammatory medical regimen? What is the optimal IOL design and material? The extent of iris manipulation during surgery, which has a significant impact on the outcome, must be considered. The longitudinal changes of the refractive error, the extent of capsular opacification, the IOL deposits and cystoid macular edema (including a more sensitive measure such as optical coherence tomography) also need to be analyzed.

Arnd Heiligenhaus, Münster

References