

Lensectomy for complicated cataract in juvenile chronic iridocyclitis

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Abstract

Experience with the removal of complicated cataract by lensectomy in patients with juvenile chronic iridocyclitis (JCI) has so far been limited. The results of lensectomy were reviewed retrospectively in 131 patients with JCI (187 eyes). The mean follow up period was 5 years 4 months. The main operative complication was accidental loss of lens material into the vitreous cavity. The postoperative complications were glaucoma (23 eyes, 15%), phthisis (14 eyes, 8%), secondary pupillary membranes (11 eyes, 6%), and retinal detachment (six eyes, 3%). The incidence of postoperative phthisis was related to the level of intraocular pressure (IOP) at the time of surgery. Twenty four per cent of hypotonous eyes and 4% of eyes with normal or elevated IOP became phthisical. Visual acuity was improved in 77%, was worse in 13%, and unchanged in 10% of eyes. The main causes of a postoperative visual acuity of 6/60 or less were glaucoma, amblyopia, and phthisis. Lensectomy did not appear to alter the course of uveitis.

Cataract is a common complication of chronic iridocyclitis particularly in patients with associated juvenile chronic arthritis (JCA) where the reported incidence is about 30%.^{1,2} In the past the prognosis of lens extraction was poor mainly due to technical difficulties which resulted in incomplete removal of lens cortex and capsule. The need for re-operation for secondary pupillary membranes frequently resulted in exacerbation of the uveitis and the subsequent development of chronic hypotony and blindness.³⁻⁶ Most of these problems can be overcome by using an automated cutting instrument. Because juvenile chronic iridocyclitis (JCI) is relatively rare the literature contains a few sporadic cases treated by this technique. The follow up in most series is also relatively short. This communication presents the long term results of a large series of patients.

Patients and methods

During early 1991 the records of 136 consecutive patients treated for cataracts secondary to JCI by lensectomy were reviewed. All were managed during a 15-year period (1976-90). Follow up data of five patients (six eyes) were unavailable. The remaining 131 patients (187 eyes) were reviewed retrospectively and the following data recorded: sex, presence of arthritis, age at the time of surgery, length of follow up, operative and postoperative complications, and visual results.

Juvenile chronic iridocyclitis was defined as a specific type of intraocular inflammation which developed before age 16 years. The uveitis was non-granulomatous, frequently bilateral, and usually had an insidious and asymptomatic onset. Band keratopathy and posterior synechiae were frequently seen in longstanding cases. Of the 131 patients (82 female and 49 male) with JCI, 100 had associated JCA, three had juvenile spondylitis, and 28 had no associated systemic disease (Table 1). The diagnosis of arthritis was made by a paediatric rheumatologist following appropriate investigations. The average age at the time of cataract surgery was 12 years 7 months (range: 3-40 years). The intraocular pressure at the time of surgery was more than 21 mm Hg in 37 (20%) and less than 10 mm Hg in 29 (16%) of the 187 eyes. Preoperative visual acuities ranged from 6/12 to hand movements.

PREOPERATIVE PREPARATION

Eyes with active uveitis were treated with dexamethasone drops at 2-hourly intervals for 2 days prior to surgery. Systemic corticosteroids were not used. An attempt was made to dilate the pupil with atropine 1% the day before surgery and phenylephrine 10% every half hour for 2 hours immediately prior to surgery. All operations were performed under general anaesthesia.

SURGICAL TECHNIQUES

Lensectomy was performed through one incision with the infusion sleeve placed over the probe either at the limbus (97 eyes) or pars plana (90 eyes). The pars plana technique was as follows. A stab incision was made with a knife into the lens 3.5 mm posterior to the limbus. The probe was introduced into the lens and small pupils were enlarged. The lens was excised from within the capsular bag. Soft lens matter was removed by suction alone while harder pieces were excised by activating the cutting mode. The cutting port was then positioned posteriorly to excise the posterior capsule and the anterior third of the vitreous gel. Finally the anterior capsule was excised. The sclerotomy was sutured and an inferior sub-Tenon's injection was given of

Table 1 Patient characteristics

Aetiology of uveitis	Number of patients		Number of eyes
	Female	Male	
Juvenile chronic arthritis	62	38	139
Juvenile spondylitis	0	3	5
Idiopathic	20	8	43
Subtotals	82	49	187

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Accepted for publication
20 June 1991

gentamicin (20 mg) and either methylprednisolone acetate (20 mg) or triamcinolone acetonide (20 mg). The technique of lensectomy via the limbus was similar except that the incision was made in peripheral clear cornea.

In seven eyes the lens nucleus was too hard to be removed with the cutter. In these cases a large corneal incision was made and the nucleus was expressed manually. Residual soft lens matter together with the capsule and anterior vitreous were excised with the cutter. In eight eyes the anterior capsule was covered by a dense rubbery plaque attached to the surrounding iris which could not be excised with the cutter. In these cases the plaque was first cut with scissors into several pieces and then each piece was removed separately with forceps or the cutter. In one eye a dense retrolenticular cyclitic membrane was excised in a similar manner. In some eyes calcified lenticular flakes that could not be aspirated into the cutter were removed with forceps. In two eyes intraocular diathermy was used to control bleeding from the iris. Combined procedures were performed in 17 eyes (trabeculodialysis 11, chelation five, trabeculodialysis combined with chelation one).

POSTOPERATIVE MANAGEMENT

During the immediate postoperative period all eyes showed a severe surgically-induced exacerbation of uveitis which was treated with the frequent use of dexamethasone drops. The rate of instillation was then adjusted according to the severity of inflammation. With the passage of time the aqueous flare gradually became less dense but in the majority of cases some degree of flare persisted indefinitely. At follow up only four eyes had a longlasting and complete resolution of uveitis.

Results

A total of 187 eyes in 131 patients were available for analysis. The mean follow up period was 5 years 4 months (range: 1 month–15 years).

OPERATIVE COMPLICATIONS

In 14 eyes lens fragments were accidentally dislodged into the vitreous. Small fragments in nine eyes were left. In five eyes the fragments were large; in three of these they were left and in two they were retrieved with the cutting probe. The three large lens fragments which were left subsequently appeared in the anterior chamber during the postoperative period. In one case the lens material was removed with the cutter without complications. One of the two remaining eyes developed corneal decompensation due to endothelial damage which later necessitated penetrating keratoplasty and the other eye

developed intractable glaucoma and was enucleated.

POSTOPERATIVE COMPLICATIONS

Details of postoperative complications are shown in Table 2.

1. Secondary glaucoma

Fifteen per cent (23) of the 150 eyes that did not have glaucoma preoperatively subsequently developed persistently raised intraocular pressure. The mean interval between cataract surgery and the diagnosis of glaucoma was 3 years (range: 3 months–10 years). When last examined four of these eyes had no light perception and two had been enucleated.

2. Phthisis bulbi

Twenty four per cent (seven) of the 29 eyes with an intraocular pressure of less than 10 mm Hg at the time of surgery became phthisical, usually within a few weeks of surgery. Of the remaining 22 eyes 12 regained a normal intraocular pressure, one developed glaucoma, and nine remained soft. Four per cent (seven) of the 158 eyes that were not hypotonous at the time of surgery subsequently developed phthisis. The overall incidence of postoperative phthisis was 8% (14) of the 187 eyes.

3. Secondary pupillary membranes

Six per cent (11) of the 187 eyes developed secondary pupillary membranes. In seven eyes the membranes developed within the first few postoperative days either as a result of very severe surgically induced exacerbation of uveitis (four) or the presence of blood in the pupil (three). In the remaining four eyes the membranes consisted of residual opacified posterior capsule that had been inadequately excised during lensectomy. In 10 eyes the membranes were subsequently excised surgically, and in one eye the membrane was treated with a Nd:YAG laser.

4. Retinal detachment

Three per cent (six) of the 187 eyes developed retinal detachment. The mean interval between cataract surgery and the diagnosis of retinal detachment was 2 years 5 months (range: 2 months–7 years 6 months). In two eyes retinal surgery was not performed because of poor visual prognosis. Successful retinal reattachment was achieved in three of the remaining four eyes.

VISUAL RESULTS

Visual acuity was improved in 77% (144), it was worse in 13% (24), and unchanged in 10% (19) of the 187 eyes. Table 3 shows the pre- and postoperative visual acuities in relation to the follow up period. Table 4 shows the causes of a postoperative visual acuity of 6/60 or less. The mean age at the time of surgery of patients with amblyopic eyes was 5 years.

Table 2 Postoperative complications

Complication	Number of eyes (%)
Glaucoma	23/150 (15)
Phthisis	14/187 (8)
Secondary membranes	11/187 (6)
Retinal detachment	6/187 (3)

Table 3 Visual results in relation to follow up

Visual acuity	Follow up (months)					Number of eyes (%)
	1-35	36-71	72-107	108-143	144-180	
HM-no PL	9	6	5	10	7	37 (20)
CF-6/60	17	7	5	6	2	37 (20)
6/36-6/24	6	1	2	1	0	10 (5)
6/18-6/12	10	4	2	3	1	20 (10)
6/9-6/6	37	15	9	15	7	83 (45)
Subtotals	79	33	23	35	17	187 (100)

HM=hand movements.
PL=perception of light.
CF=counting fingers.

Discussion

In analysing the results of this study it is not possible to apply the same criteria as for senile cataracts because of the completely different problems encountered in eyes with chronic iridocyclitis. For example 36% of eyes had either secondary glaucoma or hypotony at the time of surgery and the majority had persistent inflammation following cataract extraction. In addition the poor visual results in some cases were caused by amblyopia.

Secondary glaucoma developed postoperatively in 15% of the eyes with frequently devastating effects. It can be speculated that some eyes would have developed glaucoma had cataract extraction not been performed. However it is probable that in the majority of cases the development of glaucoma was either accelerated by or directly caused by cataract surgery. It seems likely that the postoperative surgically induced exacerbation of intraocular inflammation enhanced the development of secondary angle closure by the formation of peripheral anterior synechiae.

The incidence of phthisis bulbi was in part related to the level of intraocular pressure at the time of cataract surgery (that is 4% in eyes that were not hypotonous and 24% in soft eyes). In the past hypotony was considered to be a contraindication to cataract surgery. However this study shows that the presence of hypotony at the time of lensectomy did not necessarily mean that postoperative phthisis was inevitable because 45% of soft eyes either regained a normal intraocular pressure or became glaucomatous during the postoperative period. It therefore appears that if lensectomy is performed in a 'pre-phthisical' eye the progressive downhill course to complete phthisis may be halted or even reversed. The exact mechanism by which lensectomy combined with anterior vitrectomy may result in a return to normal of secretion of

aqueous by the ciliary epithelium is speculative. It can be postulated that the excision of an invisible 'cyclitic membrane' which is causing ciliary shutdown secondary to detachment of the ciliary body is a possible mechanism in some cases. However the author was able to identify a true cyclitic membrane at the time of surgery in only one eye.

One of the main theoretical advantages of lensectomy over other techniques is its ability to eliminate the 'scaffold' (that is posterior capsule and anterior vitreous gel) along which secondary membranes can grow postoperatively. Despite this 6% of the eyes still developed secondary pupillary membranes. In four eyes secondary membrane formation could have been prevented by taking greater care to perform a more complete excision of the posterior capsule at the time of lensectomy. In the remaining seven cases secondary membrane formation was probably unavoidable as it occurred during the immediate postoperative period as a result of either a severe exacerbation of intraocular inflammation or the presence of organised blood within the pupil.

Analysis of the visual results should be interpreted with caution because, even following a perfectly performed lensectomy, the visual acuity may be poor due to other complications of uveitis. It is however important to point out that several aphakic eyes remained unscathed and maintained excellent visual acuity for several years. For example 50% of aphakic eyes maintained a visual acuity of 6/18 or better after a follow up period of 9 years or more (Table 3).

Amblyopia was the second most common cause of a final visual acuity of 6/60 or less. A poor visual result in a young child should not be attributed to mild band keratopathy or questionable macular oedema with the possibility of amblyopia being overlooked. It is also important not to delay cataract surgery unnecessarily because the longer the delay the worse will be the results of anti-amblyopia therapy.

It is difficult to draw meaningful comparisons between this series and other reports because of variations in instrumentation, technique of lens removal, extent of excision of the vitreous gel, and period of follow up. Flynn *et al*⁷ operated on seven patients (10 eyes) with JCI associated with JCA using a three incision pars plana approach with excision of the posterior vitreous in all cases. After a mean follow up of 2 years 4 months all 10 eyes had a visual acuity of 20/70 or better. The two major obstacles to good vision in eyes that had successful surgery were persistent macular oedema and poor control of pre-existing glaucoma.

Diamond and Kaplan⁸ performed lensectomy on 25 eyes of 20 patients for complicated cataracts due to various forms of uveitis. They used a pars plana approach and performed a subtotal vitrectomy. The two patients with JCI also had JCA. The postoperative visual acuities were 20/20 and 20/70, but no details were given of the period of follow up.

Dangel *et al*⁹ reported the results of lensectomy in eight eyes of five patients with various forms of uveitis. They used a two incision limbal approach with a separate incision for the infusion cannula and performed a partial anterior vitrec-

Table 4 Causes of poor postoperative visual acuity

Cause	Visual acuity (number of eyes)		
	No PL-HM	CF-6/60	Subtotals
Glaucoma	17	4	21
Amblyopia	1	14	15
Phthisis	14	0	14
Hypotony	0	8	8
Maculopathy	1	7	8
Retinal detachment	4	0	4
Keratopathy	0	3	3
Unknown	0	1	1
Subtotals	37	37	74

PL=perception of light.
HM=hand movement.
CF=counting fingers.

tomy. Of the two patients with JCI, one had juvenile spondylitis (two eyes) and the other JCA (one eye). The postoperative visual acuities were 20/40 (two eyes) and 20/80. No complications were encountered after a mean follow up of 11 months.

Tutein Nolthenius and Deutman¹⁰ performed lens removal through a pars plana incision of 19 cataracts in patients with various forms of uveitis. Although details of the age of the patients at the time of surgery and the types of uveitis were poorly documented it appears that in the two patients (two eyes) with JCI and JCA the postoperative visual acuities were 6/18 and 6/12. No complications were reported after a mean follow up of 1 year 8 months. Visual results of lensectomy in JCI in other isolated reports were 20/25 (two eyes) and 20/200,¹¹ 20/50 and 20/80,¹² 20/30 and counting fingers,¹³ and 20/70.¹⁴

Whether lensectomy-vitreotomy alters the pattern of uveitis is difficult to answer because only careful observation over a very long period of time is likely to reveal a significant change in pattern. Diamond and Kaplan⁸ noted an apparent decrease in the severity of recurrent episodes of uveitis after a mean follow up of 1 year 3 months. They speculated that lensectomy-vitreotomy may be beneficial by providing an unobstructed pathway that facilitates the removal of cellular debris from the vitreous cavity. It has also been postulated that the maintenance and/or relapse of intraocular inflammation may be associated with the presence of antigen-specific 'memory cells' in the vitreous cavity.¹⁰ The mechanical removal of these cells together with immune complexes may in some way modulate the course of the uveitis. In this study only 2% of the eyes had a complete and long lasting cessation of uveitis activity following lensectomy. The mean follow up in these four eyes was 9 years 6 months. It is

important to point out that whereas Diamond and Kaplan⁸ performed a subtotal vitrectomy in all eyes the author performed only an anterior vitrectomy. It is possible that a more extensive excision of vitreous gel might have been more beneficial.

In conclusion, it appears that lensectomy is a good technique for the removal of soft complicated cataracts in patients with JCI. The risk of postoperative phthisis is small provided the intraocular pressure is not low at the time of surgery. Lensectomy does not appear to alter the course of chronic iridocyclitis.

This work formed part of an MS thesis accepted by the University of London.

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