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CLINICAL STUDY

Retinal detachment following surgery for congenital cataract: presentation and outcomes

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Abstract

Aims **To review the current management and outcomes of retinal detachment following cataract surgery in childhood.**

Methods **A retrospective review of 59 eyes of 52 patients.**

Results **In 37% of patients, the fellow eye was blind (<3/60). The macula was detached at presentation in 81% of eyes. The detachment was obscured by residual lens matter, or a miotic pupil in 67% of eyes that had cataract surgery before 1970, but in only 30% of eyes operated after 1970. The retina was reattached with one operation in 59% of eyes, and final reattachment was achieved in 81%. No retina was reattached by scleral buckling alone. Visual outcomes were slightly better in eyes that had retinal detachment repair after 1 January 1998. Nine patients were blind in both eyes at the latest follow-up.**

Conclusions **Retinal detachment following childhood cataract surgery remains a serious condition. There has been some improvement in the prognosis due to a combination of advances in childhood cataract surgery as well as to improvements in the management of retinal detachment. Early recognition and prompt surgery, using an internal approach, may prevent blindness in most cases.**

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Introduction

Between one and six children out of every 10 000 live births have congenital cataract.[1] Many of them will develop normal vision, following cataract surgery, but others will become blind, either from the cataract, or from complications following surgery. These complications can be devastating,[2,3] and include glaucoma, and retinal detachment.

Retinal detachment following surgery for congenital cataract has a poor prognosis.[4–6] This is partly because the detachment is obscured by miotic pupils, nystagmus, and residual lens material. However, there may also be persistent traction caused by incarceration of vitreous strands in corneal wounds; and pathological elongation of the globe.[7]

In the last decade, the prognosis may have improved, partly as a result of advances in detachment surgery, such as vitrectomy[6] and wide angle viewing systems, and also from developments in paediatric cataract surgery, which allow complete clearance of the lens remnants.[8]

This study was carried out to examine the presenting features and postoperative outcomes in a series of eyes that had retinal detachment following surgery for congenital cataract.

Patients and methods

This was a retrospective case series that included 59 eyes of 52 patients.

Cases were identified through two databases. The first covered the period from 1 March 1988 to 30 September 1991 (27 cases). The second covered the period from 30 March 1998 to 31 February 2002 (25 cases). Seven cases were identified outside these date ranges; most of these were patients in whom one eye was included in the databases, and a detachment in the other eye was found on reviewing the notes.

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Criteria for inclusion in the study were: a history of surgery for cataract before the age of 16 years, and surgery for rhegmatogenous retinal detachment carried out at Moorfields Eye Hospital. Patients were excluded if there was a history of trauma, or if lens extraction had been carried out for a dislocated lens.

Following identification of the cases, case notes were retrieved, and eligibility confirmed. Patient data were then entered into a database for analysis. Continuous variables were analysed by *t*-test, and categorical variables by χ^2 and Mann–Whitney U-test.

Results

Of the 52 patients, 33 (63.5%) were male. Eyes were divided into two groups: Early (first operation before 1 January 1998), and Late (first operation after 1 January 1998). The characteristics of the two groups are shown in Table 1.

In all, 12 patients (23.1%) had nystagmus. The cause of the cataract was unknown in 45 patients (86.5%). Three patients (5.8%) had autosomal dominant cataract, and one each had cataract related to uveitis, steroid use, lenticonus, and rubella. Details of the cataract surgery were unavailable in most cases. The types of detachment identified are given in Table 2.

Preoperatively, nine eyes (15.3%) had a visual acuity of 6/18 or better, 15 (25.4%) were 6/24–6/60, and 35 (59.3%) were less than 6/60. Visual acuities in the fellow eye were better in patients that had cataract surgery after 1970 (Mann–Whitney U-test, $z = 2.38$, $P = 0.017$). In 22 cases (37.3%), the fellow eye was blind (<3/60).

In 32 eyes, the surgeon noted poor visualisation of the retina before surgery, usually because of a combination of lens remnants and poor mydriasis. In one patient, the diagnosis of retinal detachment could only be made with B-scan ultrasonography. The retina was obscured in 26 eyes (66.7%) in which cataract surgery was carried out before 1970, and in six eyes (30.0%) in which surgery was carried out in 1970 or later ($\chi^2 = 5.76$, $P = 0.02$).

The macula was detached in 39 (92.9%) eyes that had been symptomatic for 3 days or longer, and in nine (52.9%) eyes that had symptoms for less than 3 days ($\chi^2 = 10.2$, $P = 0.001$). The extent of the detachment was limited to one quadrant in eight eyes (13.6%), and the retina was totally detached in 23 eyes (38.9%).

In all, 13 patients (25%) had bilateral detachments.

Seven eyes (11.9%) had a scleral buckle as their primary procedure. In total, 52 eyes (88.1%) had a vitrectomy.

In all, 24 eyes (40.6%) needed a second reattachment procedure. Of these, four (16.7%) had a scleral buckle, and 20 required vitrectomy.

Five eyes required a third operation to reattach the retina. One (20%) required a gas/fluid exchange, and four (80%) required vitrectomy and silicone oil.

Silicone oil was used in 19 (32.2%) eyes. The oil was removed in 11 eyes. At least one vitrectomy was required in 56 (94.9%) eyes. Nine eyes (15.3%) required a retinectomy.

In eyes that had cataract surgery before 1970, 25 (64.1%) needed anterior segment revision, compared to five (25%) of the eyes that were operated in 1970 or later ($\chi^2 = 6.6$, $P = 0.01$). In the Early group, 22 (64.7%) had anterior segment revision, compared to eight (32%) of the Late group ($\chi^2 = 4.93$, $P = 0.03$).

Table 2 Type of detachment

Type of detachment	*No. of eyes*
U-tear and PVD	39 (66.1%)
Atrophic holes, no PVD	9 (15.3%)
Dialysis	2 (3.4%)
Giant retinal tear	2 (3.4%)
No break found	7 (11.8%)

Table 1 Characteristics of Early and Late groups

	Early	*Late*	P
Number	34	25	
Male	20 (58.8%)	18 (72%)	0.44
Mean age at retinal detachment (years) (SD)	37.4 (11.5)	33.4 (13.5)	0.09
Mean age at cataract operation (years) (SD)	4.3 (4.3)	6.9 (5.1)	0.04*
Mean interval from cataract surgery (years) (SD)	33.1 (11.5)	25.3 (14.9)	0.03*
Mean duration (days) (SD)	69.6 (162)	29.3 (45)	0.23
Macula-off	30 (88.2%)	18 (72%)	0.22
Blind (<3/60) in fellow eye	16 (47.1%)	6 (24%)	0.12
Blind (<3/60) patient	6 (17.6%)	2 (8%)	0.49
PVR (C1 or worse) at presentation	7 (29.1%)	3 (12%)	0.21
Poor visibility at presentation	22 (64.7%)	10 (40%)	0.11

*P < 0.05.

The retina was reattached after one operation in 35 (59.3%) eyes. Primary reattachment was more likely if the initial operation used an internal approach (35 eyes, 67.3%) rather than a scleral buckle (0 eyes) ($\chi^2 = 8.96$, $P = 0.003$). In the Early group, 50% of retinas were reattached after one operation, compared to 72% of the Late group ($\chi^2 = 2.05$, $P = 0.15$).

Primary reattachment was achieved in 10 (90.9%) eyes that were macula-on at presentation, compared to 25 eyes (52.1%) in which the macula was detached ($\chi^2 = 4.1$, $P = 0.04$).

There was no significant association between primary reattachment and age at cataract surgery, grade of surgeon, duration of symptoms, visibility of retina, extent of detachment, presence of proliferative vitreoretinopathy (Grade C1 or worse), or anterior segment revision.

The retina was finally reattached in 48 eyes (81.4%). Final reattachment could not be achieved in eight (23.5%) of the Early group, and in three (12%) of the Late group ($\chi^2 = 0.62$, $P = 0.43$). Among patients who had a retinectomy, only three (33.3%) were successfully reattached ($\chi^2 = 12.63$, $P = 0.0004$).

There was no significant association between final anatomical success and age at cataract surgery, grade of surgeon, duration of symptoms, visibility of retina, extent of detachment, presence of proliferative vitreoretinopathy, or anterior segment revision.

Visual acuities before and after surgery are shown in Figure 1. In all, 34 eyes (57.6%) achieved an acuity of 6/60 or better. A total of 20 eyes (33.9%) achieved 6/18 or better. The median final visual acuity was 6/60 in the Early group, and 6/24 in the Late group (Mann–Whitney U-test, $z = 1.70$, $P = 0.089$). Among eyes in which the macula was attached at presentation, 11 (100%) had a good visual outcome (6/60 or better) compared to 23 (47.9%) of the eyes in which the macula was detached ($\chi^2 = 7.92$, $P = 0.005$). In eyes with a vision of counting fingers or less at presentation, 20 (69.0%) had a poor visual outcome, as opposed to five (16.7%) of the eyes with a preoperative vision of better than counting fingers ($\chi^2 = 14.44$, $P = 0.0002$). Of the 19 eyes in which silicone oil was used, 15 (78.9%) had a poor visual outcome ($\chi^2 = 13.22$, $P = 0.0003$). In the Early group, 10 (29.4%) lost vision (latest visual acuity at least two lines worse than preoperative visual acuity), whereas in the Late group, only one (4%) eye lost vision ($\chi^2 = 4.57$, $P = 0.03$).

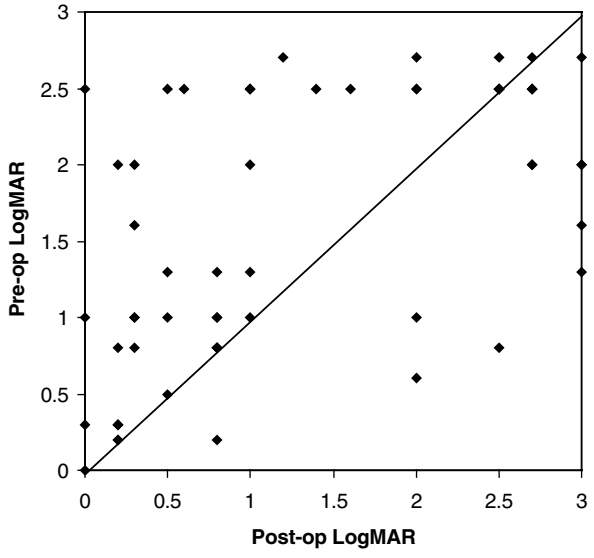

Figure 1 Preoperative and postoperative visual acuities (log-MAR equivalents).

There was no significant association between poor visual outcome and duration of symptoms, primary anatomical failure, presence of PVR, age at cataract surgery, or year of cataract surgery.

Preoperatively eight patients (15.4%) were blind (<3/60 in both eyes). At the latest follow-up nine patients (17.3%) were blind. Two patients recovered vision in the operated eye, and three patients had become blind in their better eye.

Discussion

Over 60% of the patients were male. This has been reported by other authors.[4–7] It is not clear whether males have an increased risk of congenital cataract, or an increased risk of retinal detachment following congenital cataract surgery.

The mean interval between cataract surgery and retinal detachment was 29.8 years. This is similar to other reports.[4–7,9] The prolonged interval makes it difficult to estimate accurately the risk of retinal detachment following surgery for congenital cataract, or the effect of changes in cataract surgical techniques. The incidence of retinal detachment following surgery for congenital cataract has been estimated at 1.5%;[3] however, the mean follow-up of this cohort was only 5 years, so the true incidence is likely to be higher. The mean interval between surgery and detachment was significantly shorter in the Late group. There were no lensectomies in the early group, but at least three patients in the Late group had lensectomy, and the mean interval to detachment was only 8.7 years in these eyes. Lensectomy may be associated with a more rapid progression to retinal detachment.

There is evidence that the outcomes of congenital cataract surgery may have improved, as the visual acuity in the fellow eye was better in patients who had surgery after 1970. Visualisation of the retina was also easier in patients who had more recent surgery. Although in a retrospective study this is impossible to quantify,

significantly fewer patients required anterior segment revision in the later group. None of our patients were pseudophakic. The use of intraocular lenses in congenital cataract is controversial, but is becoming more widespread,[10–13] which may affect the visibility of the retinal periphery.

Bilateral detachment seems to be particularly common in these patients.[4,7] In our series, 25% had bilateral detachment. It is reported that in the entire detachment population, 10% have bilateral detachments;[14] however this rises to 16% in aphakic patients.

In the UK audit, total detachment occurred in only 12.5% of eyes;[15] however, in our patients, 38.9% had a total or complex detachment. The poor prognosis in our patients may be due to a higher prevalence of complex detachments as well as poor visualisation of the retina.

The macula was detached at presentation in 81.4%, which is higher than recent detachment audits,[15,16] but similar to the 72% found by Bonnet and Delage[6] In many patients, there was some delay before attending for surgery. Although there was no direct association between duration of symptoms and outcome, patients who were symptomatic for less than 3 days were more likely to have a macula-on detachment, and these eyes had a better outcome. It is possible that the greatest improvement in the prognosis of these detachments would come not from advances in re-attachment surgery, but from earlier recognition and referral.[17]

As other authors have suggested, an internal approach seems to improve the likelihood of anatomical success.[5,6] No eye that had an initial external approach was reattached with one operation. Although these patients are relatively young, we recommend an internal approach, as we believe this provides the best opportunity of inspecting the peripheral retina and identifying all breaks.

Anterior segment revision was required less frequently in the Late group. This may be a reflection of better cataract surgery, or of the benefits of a wide-angle viewing system. This system was used at Moorfields from 1998.

The anatomical success rate is lower for these eyes than the standard for the UK. In the national audit, 82% of eyes were reattached with one operation, and 93.3% were finally reattached.[18] In our series, 59.3% were reattached with one procedure, and 81.4% were finally reattached. This compares favourably with other published series of detachments following congenital cataract surgery (Table 3). The primary reattachment rate was higher in the Late group; however, as the numbers of eyes were small, this could have occurred by chance. Any improvement in outcomes is probably due to a combination of advances in surgery for congenital cataract and for retinal detachment.

Table 3 Anatomical success rates in previous reports of retinal detachment following surgery for congenital cataract

Author	*Year of publication*	*No. of eyes*	*Primary success*	*Final success*
Shapland	1934	16	0	0
Kanski	1974	34	14 (41.2%)	20 (58.8%)
Jagger	1983	62		39 (62.9%)
Bonnet	1994	25		22 (88.0%)
Current study	2003	59	35 (59.3%)	48 (81.4%)

Visual outcomes appeared to be slightly better in the Late group. Although there was no significant difference in the final visual acuity, fewer patients in the Late group lost vision following surgery. This suggests that the improved visual outcome is due to better management of the detachment. Final visual acuity depends largely on preoperative vision,[19–21] and eyes that could see better than counting fingers at presentation were more likely to regain useful vision.

Although there is evidence of improved outcomes, the prolonged interval between cataract surgery and retinal detachment means that it will be another 20 years before we can know if modern congenital cataract surgical techniques are less likely to lead to retinal detachment.

Despite surgical advances, nine patients (17.3%) remained blind at latest follow-up. It is still true that, as Shapland[9] wrote in 1934: 'of all cases of retinal detachment they are the most tragic, irretrievable blindness of both eyes being the lot of a disquietingly large proportion of them, and at an age when they are the greatest value to themselves and to the community'.

In conclusion, despite advances in the surgery of both congenital cataract and retinal detachment, these patients are at risk of blindness. Early diagnosis, followed by prompt vitrectomy, appears to offer the best hope for retaining useful vision.

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