Original Article

Juvenile Rhegmatogenous Retinal Detachment

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Purpose: To review the clinical features, evaluate visual, and anatomical outcomes and potential complications following surgery for rhegmatogenous retinal detachment (RRD) in juveniles.
Methods: Retrospective, consecutive case series of children and young adults (birth through 18 years) who underwent surgerys for RRD between February 1999 and January 2002.
Results: The authors reviewed a consecutive series of 111 eyes of 105 juveniles [86 (77.47%) eyes belonged to male and 25 (22.52%) to female subjects] operated for RRD. The mean age of patients was 13.62 years. Bilateral retinal detachment was present in 12 (10.8%); 51 (46%) patients had some form of bilateral ocular pathology at initial presentation. The two most common aetiologies were non-penetrating trauma (45.04%) and myopia (41.44%). Decreased vision was the most frequent symptom. The mean duration of symptoms was 165.36 days. The commonest retinal break was a retinal hole (34.23%). Late diagnosis was common, evidenced by high frequency of macular detachment (97.29%) and proliferative vitreoretinopathy (PVR) (45.94%) at initial presentation. The most commonly performed primary surgery was scleral buckle (61.26%). The average postoperative follow-up after the first procedure was 10 months (range 8 -19 months). Final retinal reattachment was accomplished in 78.37% (87/111) with a mean of 1.29 surgeries per eye. Improvement, no change and decline in vision was seen in 50 (48%), 32 (31%) and 22 (21%) eyes respectively.
Conclusion: Non-penetrating injury and myopia were the most common cause for RRD in juveniles. Fellow eyes commonly had vision-threatening abnormalities. Final anatomical and visual recovery rates were encouraging despite late initial presentation and high rates of macular detachment, and PVR at initial presentation.

Key Words: Paediatric, juveniles, rhegmatogenous retinal detachment, myopia, trauma, vitrectomy, scleral buckling

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The annual incidence of rhegmatogenous retinal detachment (RRD) has been estimated as approximately 12.4 cases per 100,000 population.[1] RRD in childhood and adolescence (birth to 18 years) has been estimated at 2.9 – 5.6%, a small fraction of all patients treated for RRD. [2] This low incidence may be responsible for paucity of large series of RRD in this age group in recent published literature.

Despite its low prevalence, RRD in juveniles is an important and distinct subset of retinal detachment. Unilateral retinal detachment in children is often an incidental finding due to lack of symptomatic complaints. Delayed diagnosis, possible association with other complicating factors including ambloyopia can worsen the prognosis. Entities causing retinal detachment in childhood possess extremely aggressive proliferative and cicatricial pathoanatomy, less amenable to surgical treatment. A large majority of these children have abnormal findings in the fellow eye, often requiring medical or surgical intervention.[3] Pre and postoperative evaluation is challenging and time consuming. Furthermore, the social and economic implications of childhood blindness are extremely severe. These factors merit a deeper scrutiny of the subject.

In one of the largest series so far, Winslow et al reported surgical results of 109 children with RD.[2] However, this and several other studies[4-9] were conducted in the pre-vitrectomy era. Recently Scott et al[10] published results of a prospective, multi-centre study of 205 patients (211 eyes) 16 years of age or younger for complex retinal detachments associated with trauma, proliferative vitreoretinopathy (PVR), giant retinal tear (GRT), or retinopathy of prematurity

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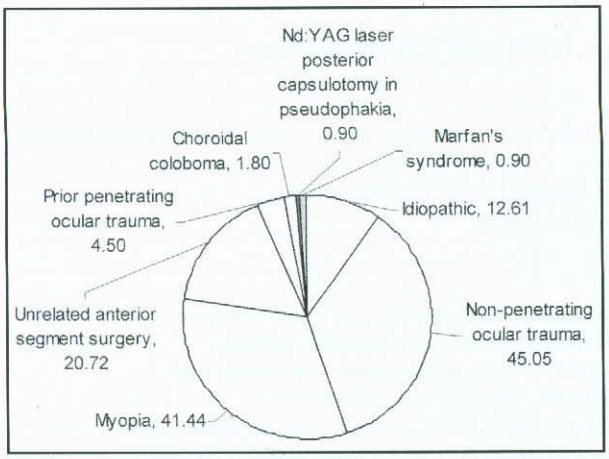

Figure 1. Aetiological factors (%);

(ROP). All patients underwent vitrectomy with silicone oil retinal tamponade. Some other recent studies[3,11-13] have fewer subjects. Inclusion of limited aetiological or complicating factors and selective surgical procedures (scleral buckling or vitrectomy) further limit their scope.

This study was designed to present a composite clinical description of RRD in the juvenile age group. The authors aimed to evaluate incidence, aetiological factors, clinical presentation and functional and anatomical results in these cases. Subjects represented a wider range of aetiological and complicating factors, relatively larger sample size and numerous surgical options such as scleral buckling and/or three port pars plana vitrectomy (PPV), buckle revision, etc.

Materials and Methods

Case records of all subjects (aged 0-18 years) with RRD examined between February 1999 and January 2002 were reviewed retrospectively. We excluded patients who did not undergo recommended surgical treatment, eyes with surgical prognosis considered too poor to be operated upon and those who lacked the minimum follow-up period of 8 months after the first surgery. Eyes with RD secondary to retinopathy of prematurity (ROP) or recent globe disruptive trauma were excluded because of the tractional element in the former and potentially uncertain aetiology of detachment in the latter, respectively. RD in association with Coat's disease was also excluded. However, eyes with RRD in cases of old healed penetrating trauma, with structurally intact anterior and posterior segments, were included. Eyes with recurrent or persisting RD following previous buckling and/or PPV surgeries at our centre or elsewhere were also included. In these cases, the first surgery performed by us during the study interval was considered as the first surgery for the purpose of our study.

Patient data was gathered from standardised individual case forms. Records of both eyes were carefully gathered from these forms. Data collected included date of initial presentation, age at presentation, gender, affected eye, nature and duration of presenting symptom(s), aetiologic factor(s), relevant past history, medical history and positive ocular history in family members. Presenting Snellen visual acuity and refractive status (wherever possible to ascertain), and anterior segment examination details including lens status were noted. Refractive status of the other eye was carefully noted. Other details gathered included type and extent of RD; type, site and number of retinal breaks; macular status (off versus on); presence and severity of PVR; type of primary surgical procedure (scleral buckling or pars plana vitrectomy); subsequent surgeries; interval between surgeries; use of intraocular tamponade; complications and follow-up record. Postoperative refractive status, best-corrected visual acuity and steps taken to prevent or treat amblyopia were documented. Anatomical outcome was determined by noting intra- and postoperative retinal attachment status. The functional outcome was determined by change in visual acuity (where possible to establish).

All patients included in the study had a minimum of 8 months' follow-up after the first surgical procedure. They had usually been seen at the first, second, and third day postoperatively, followed by examinations at one week and 1, 2, 5 and 8 months postoperatively. Subsequently, they were scheduled for 6 monthly follow-ups. Patients with persistent re-detachment of retina, or those who developed complications following surgery were followed up with greater frequency – as clinically indicated in each case. Interventions such as repeat surgeries, etc had been carried out as and when necessary. Consecutive IRB approval was not necessary due to the retrospective nature of the study.

Table 1. Abnormal ocular findings among fellow eyes

Fellow eye status	Number of eyes
RD (inoperable)	6
RD (operated)	6
Moderate to high myopia (≥ - 4D sphere)	32
Peripheral retinal breaks	11
History of cataract surgery	3
Phthisical	3
History of PPV for RD	1
History of scleral buckling for RD, with retina settled	1
History of trabeculectomy	3
History of Cryo-prophylaxis	2
Subluxated crystalline lens in a case of Marfan's syndrome	1
Congenital cataract	6
Choroidal coloboma	2
History of laser prophylaxis	1
Absolute eye due to neo-vascular glaucoma	1

Results

Incidence

This study spanned three years - February 1999 to January 2002. During this period, 2374 eyes with RRD were seen at our institute. Of these, 137 eyes belonged to the 0 – 18 years age group, accounting to 5.77% of total eyes with RRD.

Patient characteristics

Eleven eyes (of 11 subjects) were excluded from the study because they did not undergo surgery despite clinical advice. A further 9 eyes (of 9 subjects) did not complete the minimum follow up of 8 months following the first surgery and hence were excluded. Six eyes (all contra-lateral eyes in cases with bilateral detachments) had long-standing detachments, considered inoperable and thus excluded. Thus total of 111 eyes of 105 patients were finally included. Six patients had bilateral RRDs and both eyes were enrolled in the study. Of these, 5 had detachments in both eyes at presentation and one patient developed RRD in the second eye within one month of surgery in the first eye. Eighty-one subjects (86 eyes; 77.4%) were males and 24 (25 eyes; 22.5%) females. The youngest patient was 40 months and oldest 18 years old. The mean age of male patients at presentation was 13.67 ± 3.47 years, and that of females, 13.46 ± 3.58 years. Six children were below 8 years of age. Postoperative follow-up ranged from 8–19 months (average 10 months) following the first procedure.

Table 2. Retinal break characteristics

Retinal break characteristics	No. of eyes
Single	54
Multiple	45
Obscured view due to:	
Vitreous haemorrhage	2
Cataract	2
PVR	2
PCO	1
Choroidal effusion	1
Hole	38
HST	22
Dialysis	22
Large tear (>1/2 clock hour)	15
GRT	11
Lattice with hole	13
Lattice	11
Operculum	6
Hole with operculum	5
Erosions	3
Coloboma	2
Peripheral retinal thinning	1

Table 3. Second procedures in eyes following successful reattachment of retina after the first surgery

Second procedure in eyes with retina settled by the first surgery	Number of eyes
Lensectomy, Silicone oil removal, Endolaser	1
Silicone oil removal, reinjection	2
Silicone oil removal, phaco, IOL	2
Plomb removal	1

Aetiological factors

The contributing aetiologic factor/s could be determined from history or ocular examination records in 97 (87.38%) of 111 eyes (Figure 1). The remaining 14 (12.61%) eyes where no aetiological factor could be construed were labelled idiopathic. Non-penetrating trauma was the main aetiologic factor, observed in 50 (45.04%) eyes. Leading causes for blunt trauma in this age group (in descending order) were – ball injury (while playing cricket), injury while playing tipkat (*gilli-danda)*, injury with stone, fist or hand injury and firecracker injury. The second most common aetiological factor was moderate to high myopia (≥ 4.0 D) in 46 (41.44%) eyes. Twenty-three (20.72%) eyes had a history of anterior segment surgery cataract extraction for congenital (8 eyes) or traumatic cataract (10 eyes) or surgery for childhood glaucoma (5 eyes). One patient was a known case of Marfan's syndrome. He was myopic with bilateral supero-temporal subluxation of the clear crystalline lenses. Other factors associated with RRD were old, healed penetrating ocular trauma, choroidal coloboma and following Nd:YAG laser posterior capsulotomy in pseudophakia. These factors were not mutually exclusive and more than one was often found in the same eye.

Three patients developed RRD despite previous prophylactic treatment to offending retinal lesions – one following prophylactic laser treatment 7 months prior and two following cryo-prophylaxis at 3 and 12 months prior respectively. The child with cryo prophylaxis 3 months prior presented with total RRD with GRT. Twelve eyes had recurrent or persisting retinal detachments following previous scleral buckling and/or PPV surgeries performed at our centre or elsewhere. One child previously operated for scleral buckle (two and a half months prior) elsewhere presented with settled retina and plomb infection and protrusion. Following plomb removal, he developed re-detachment, on the third day after removal.

Fellow eyes

Fifty four (51.42%) of 105 patients had normal fellow eyes, and 51 (48.57%) had some form of bilateral ocular pathology, seen either at initial presentation, or during

Table 4. Postoperative complications

Complication	Number of eyes
Silicone oil related: Secondary glaucoma	7
Cataract	6
Keratopathy	5
Emulsified silicone oil	3
Silicone oil globules in AC	3
Retro silicone oil membrane	2
Subretinal silicone oil	1
Optic atrophy	1
Macular pucker	1
Plomb infection	2

follow up examinations (Table 1) (not mutually exclusive).

RD characteristics

Decreased vision – as reported by patient or parent – was the chief presenting complaint in 103 eyes, followed by complaints of deviation in 14, floaters in 2, flashes in 1 and field loss in 1 eye. One fellow eye was discovered to have RD on routine examination and another fellow eye developed detachment during the course of follow-up of the first eye operated for scleral buckle.

Of the 68 patients/parents who could quantify duration of chief symptoms, it ranged between 1 day and 7 years. Average time of presentation following onset of symptoms was 165.36 days. Seven patients had positive family history of RRD and nine had family history of myopia. Retinal detachment involved the macula in 108 (97.29%) of 111 eyes. Proliferative vitreoretinopathy was present at initial presentation in 51 (45.94%) of 111 eyes.

A single break was found in 54 (48.64%) and multiple in 45 (40.54%) eyes. Preoperatively, breaks could not be detected in 12 eyes (in eight eyes due to obscure view of fundus). The most common type of retinal break was a hole – in 38 (34.23%) eyes. Both horseshoe tear (HST) and retinal dialysis were found in 22 (19.81%) eyes each. A large tear (> half clock hour) was present in 15 (13.51%) and GRT in 11 (9.90%) eyes. Lattice with holes were seen in 12 (10.81%) and without holes in 10 (9%) eyes. Floating opercula were seen in 6 (5.40%), and hole with operculum in 5 (4.50%) eyes. Other associated lesions (not mutually exclusive) are listed in Table 2.

Surgical procedures

All 111 eyes underwent surgical repair for retinal detachment. Of these, 68 (61.26%) eyes underwent scleral buckling surgery alone. Fifty-five, 12 and 1 (previously operated for scleral buckling) of these 68 eyes underwent encirclage, segmental buckle and buckle revision respectively. Forty-three (38.73%) eyes underwent standard three port pars plana vitrectomy (PPV). Of these, 25 underwent encirclage and one eye, segmental buckle along with vitrectomy.

Retina settled in 88 (79.27%) eyes with a single procedure. Of these, 6 eyes (with settled retinas) required a subsequent procedure as mentioned in Table 3. Twenty-three eyes required a second procedure for retinal reattachment. The second surgery performed in all eyes with failure of retinal attachment following primary procedure was vitrectomy. Along with vitrectomy, phacoemulsification was done in six eyes and silicon oil removal with reinjection in five. Among these 23 eyes, retinas settled in 14 eyes with the second procedure, and 9 eyes remained unsettled. Thus a total of 29 eyes underwent second surgeries. Four of 9 eyes where the retina failed to settle after second procedure required a third surgery for retinal reattachment. Five were considered inoperable with retinas damaged beyond repair.

Initial surgical outcome

A total of 144 surgeries were carried out in 111 eyes, an average of 1.29 procedures per eye. Retinas in 106 (95.5%) of 111 eyes were settled per-operatively, 88 following one, 14 following two and another four following three surgeries.

Final surgical outcome

Of the 111 eyes, retina was attached in 87 (78.37%) eyes at last follow-up. Of the other 24 eyes (with detached retinas), 23 had PVR changes. Nine of these had retinas damaged beyond repair and the rest were advised trial of further surgeries.

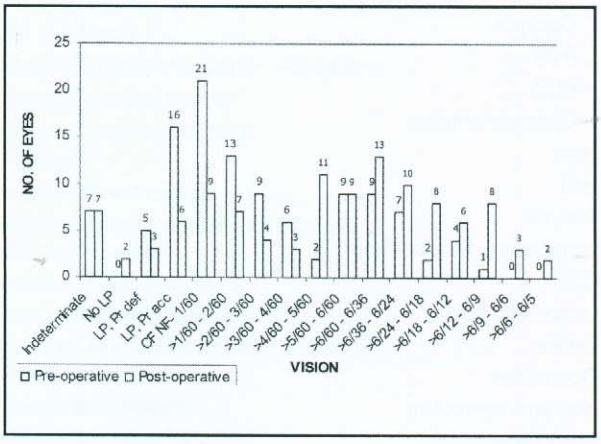

Figure 2. Comparison between pre- and postoperative vision

Table 5. Comparison with recent similar studies

Authors	Number of eyes	Mean RD duration days	Macula involved (at presentation) (%)	PVR (at presentation (%)	Type of primary surgery (%)	Final Post-op anatomic reattachment (%)	Mean surgeries/ eye	Final visual result (Improvement/ unchanged/ worsening)
Present study	111	165.36	97.29	45.94	SB:61.26 PPV: 38.73	78.37	1.29	98% / 21% / 31%
Fivgas Capone[3]	29	52	79	45	SB:28 PPV:72	72	2.2	NA
Butler Kiel Orr[11]	15	90	67		SB:80	86.6	1.46	53.3% / 33.3% / 13.3%
Haring Wiechens[12]	33	33	45	Excluded from study	SB:100	1.12	1.12	NA
Weinberg Lyon, Greenwald, J, Mets MB[14]	39		74	31	SB:41 PPV: 13 SB+PPV 46	79	1.6	NA

NA – Not available; SB – Scleral buckling; PPV – Pars Plana vitrectomy

Visual outcome

In seven eyes, vision remained indeterminate either due to small age or mental retardation. For the rest an increase or decrease of vision by ≥ 2 lines of Snellen's acuity was considered improvement or decline respectively and any change < 2 lines was considered no change. In 104 eyes where vision could be determined, improvement, no change and decline was noted in 50 (48%), 32 (31%) and 22 (21%) eyes respectively (Figure 2). Forty-two and 20 eyes in the pre- and postoperative group respectively had extremely poor vision (≤ 1/60). Thus, the relative chances of extremely poor vision were much more statistically significant in the pre-operative group as compared to postoperative group (P <.008, odds ratio 2.85). Five and 19 eyes in the pre- and post-operative group respectively had vision better than 6/18. Thus relative chances of attaining good visual acuity following surgery were highly significant (P < 0.002, odds ratio: 4.43). Maximum eyes in the pre-operative group were in the cohort of counting fingers near face to 1/60, while those in the postoperative group were in the cohort of 6/36. Some patients were still undergoing amblyopia treatment at final follow up.

Complications

Iatrogenic breaks occurred in 3 eyes, all during PPV. Laser retinopexy was done to these. Silicone oil related complications occurred in 23 (20.72%) eyes. Other post-operative complications are listed in Table 4.

Discussion

In our three-year study, 5.77% belonged to subjects 18 years or younger. This figure is close to the upper limit of the 2.9% – 5.6%[2] fraction reported in literature. The male:female ratio was 3.4:1. Blunt trauma due to ball injury (while playing cricket) was the foremost aetiological factor in our study. Non-penetrating trauma was observed in 45.04% eyes and old (healed) penetrating trauma in another 4.5%. The comparative data with recent publications are shown in Table 5.

Moderate to high myopia (≥ 4.0 D) was observed in 41.44% of our cases. Comparable, high incidence of trauma and myopia among subjects with paediatric RRD has been found by Winslow et al[2] (44% and 15% respectively), Haring et al[12] (24% and 42% respectively), Butler et al[11](40% cases with trauma), and Fivgas et al[3] (34% cases with myopia). In our series, 20.72% eyes had history of prior anterior segment surgery. 18 (16.2%) eyes had been operated for cataracts (for congenital cataract in 8 eyes and traumatic cataract in 10 eyes) and 5 (4.5%) for glaucoma. Other aetiological factors found less frequently in our study included Marfan's syndrome, retino-choroidal coloboma and following Nd:YAG laser posterior capsulotomy in pseudophakia. Three patients developed RRD despite previous

prophylactic treatment to offending retinal lesions. One child who underwent previous cryo-prophylaxis presented with total RD with GRT within 3 months. Close follow-up of children with such history is therefore mandatory.

Another remarkable feature in these children was a high incidence (in 48.57%) of bilateral ocular pathology, noted either at initial presentation or during follow up. Moderate to high myopia was the most common finding in the fellow eye. Twelve patients had bilateral detachments, of which six were inoperable. Other grave or vision threatening ocular pathologies in fellow eyes included phthisis bulbi, congenital cataract, glaucoma, lens subluxation, retino-choroidal coloboma and peripheral retinal breaks. Interventions carried out in fellow eyes (prior to or during course of study) included laser or cryo prophylaxis, scleral buckling or PPV for RD, cataract surgery, trabeculectomy and radial keratotomy.

Sixty-eight (61.26%) eyes in our series underwent scleral buckling surgery and 43 (38.73%) PPV. We reserved primary vitrectomy for cases with GRT, large breaks, posterior breaks not amenable to conventional surgery with buckling or old detachments with PVR changes. Strong vitreoretinal adhesion, occurrence of localised areas of PVD and a high incidence of post-vitrectomy cataract formation complicates vitrectomy surgery in children. Hence an external approach was preferred by us whenever feasible.

We achieved a per-operative success rate of 95.5% (106/111), with 88, 14 and 4 eyes following one, two and three surgeries respectively. The average number of procedures per eye was 1.29, comparing favourably with mean of 2.2 surgeries/eye reported by Fivgas and Capone.[3] Final anatomic success achieved by us was 78.37% (87/111 eyes), similar to the results attained in recent studies (Table 5). This was less than the 100% retinal re-attachment rate reported by Haring and coworkers.[12] The difference in average symptom duration between their subjects and ours (33 versus 165 days) and exclusion of cases with PVR in their series may explain this difference. In addition, we included 13 (11.71%) cases of previously failed retina reattachment surgeries (both scleral buckling and PPV).

Persistent retinal detachment with PVR changes was seen in 23 cases at the final follow-up. In eyes where vision could be determined, improvement, no change and decline was noted in 48%, 31% and 21% respectively. However, some patients were still undergoing amblyopia treatment at final follow up.

Thus, anatomic success and visual stabilisation to improvement was achieved in more than two-thirds of subjects below 18 years with RRD operated by us, which is encouraging despite the limitations.

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