

PEDIATRIC RHEGMATOGENOUS RETINAL DETACHMENT

GEORGE D. FIVGAS, MD, ANTONIO CAPONE, JR, MD

Purpose: To review the clinical features and surgical and visual outcomes of pediatric rhegmatogenous retinal detachment (RRD) as seen in a tertiary referral center.

Methods: Retrospective case series spanning 6 years from January 1, 1991 to January 1, 1997. Exclusionary criteria were trauma disrupting the globe and acute retinopathy of prematurity.

Results: The authors reviewed a series of 29 eyes in 27 pediatric patients (birth to 18 years of age) with RRD. Seventy percent of the patients were male. The mean patient age was 9.6 years. Bilateral RRD was present in 22% of patients; 89% of patients had some form of bilateral ocular pathology at initial presentation. The two most common etiologies (34% each) were myopia and eyes that had undergone surgery for another ocular disorder with subsequent development of RRD. The most common presentation was decreased vision, with a mean duration of 52 days. At presentation, 75% of the affected eyes and 48% of the fellow eyes had visual acuity worse than 20/800. The most common type of retinal break was a horseshoe tear. Late diagnosis was a common problem, evidenced by the frequency of macular detachment (79%) and proliferative vitreoretinopathy (45%) at initial presentation. The most common primary repair was a scleral buckle. Anatomic reattachment was ultimately accomplished in 72% of cases with a mean of 2.2 surgeries per eye. Average postoperative follow-up time was 21.4 months (range 4 to 61 months). At final follow-up, 41% of the affected eyes had visual acuity 20/800 or better. Thirty-eight percent of the affected eyes had a final visual acuity better than or equal to the fellow eye.

Conclusion: In this series, pediatric RRD occurred most commonly in association with myopia (Stickler's syndrome and adolescent retinopathy of prematurity) and prior intraocular surgery. Most eyes were anatomically reattached after multiple surgeries. Forty-one percent of eyes retained vision of 20/800 or better. Preserving vision in children with RRD is of great importance, particularly given the 89% frequency of vision-threatening abnormalities in fellow eyes.

RETINA 21:101–106, 2001

From the Department of Ophthalmology, Emory Eye Center, Emory University School of Medicine, Atlanta, Georgia.

Dr. Fivgas is currently in practice at the Vitreoretinal Institute, Baton Rouge, Louisiana. Dr. Capone is currently in practice at Associated Retinal Consultants, Royal Oak, Michigan.

Supported in part by a departmental grant from Research to Prevent Blindness, Inc., New York, New York.

The authors have no proprietary interest in the material presented.

Reprint requests: George D. Fivgas, MD, Vitreoretinal Institute, 7698 Goodwood Blvd., Baton Rouge, LA 70806; e-mail: gfivgas1@home.com

Rhegmatogenous retinal detachment (RRD) has an annual incidence of approximately 12.4 cases per 100,000 population.¹ Rhegmatogenous RD occurring in the pediatric age group (birth to 18 years of age) accounts for only 3.2% to 5.6% of the total (approximately 0.38–0.69 per 100,000 population), with over 40% of cases secondary to ocular trauma.² There are few reported series of RRD in children, most likely due to the low incidence in this age group. Herein, we

review pediatric detachments unrelated to globe-disrupting trauma or acute retinopathy of prematurity (ROP) with cicatricial changes in infancy.

Methods

We retrospectively reviewed charts from the Emory Eye Center, Emory University Hospital, and Henrietta Egleston Children's Hospital for the period spanning January 1, 1991 to January 1, 1997. Data gathered included age at presentation, gender, affected eye, nature and duration of presenting symptom(s), medical and ocular history, fellow eye pathology and acuity, family ocular history, presenting visual acuity (VA), refractive status, type of detachment, macular status (off versus on), presence of proliferative vitreoretinopathy (PVR), initial lens status, number and type of surgeries performed, success of intraoperative repair, postoperative retinal attachment status, use of intraocular tamponade, and follow-up interval.

Results

Sixty patients under 18 years of age underwent surgery for RRD within the study interval. Thirty-three of these were excluded: 25 owing to globe-disrupting trauma, 6 owing to acute ROP in infancy (associated tractional component secondary to fibrovascular proliferation), and 2 who carried the diagnosis of RRD but had incomplete hospital charts and insufficient follow-up data. The 31 traumatic and acute ROP patients were excluded because the mechanism of detachment was decidedly different from the remainder of the group.

The remaining 27 patients comprised the study group. One patient presented with bilateral detachments and another developed a consecutive detachment during the course of follow-up, yielding a total of 29 eyes for inclusion and evaluation. The etiology of rhegmatogenous detachment for each of the 29 eyes is listed in Table 1. Myopia (34%, 10/29 eyes) and RRD occurring subsequent to unrelated intraocular surgery (34%, 10/29 eyes) were the two most commonly noted predisposing conditions. Of the 10 myopic eyes, five carried a clinical diagnosis of Stickler's syndrome, four had ROP-related detachments in adolescence, and one had unqualified high myopia. Of the 10 eyes developing RRD postoperatively, six were aphakic at presentation. Table 2 lists the operative procedures performed before the development of RRD in the postoperative group.

Twenty-five of the 29 (86%) eyes underwent vitreoretinal surgery at Emory University. Four eyes (14%) underwent initial attempt at RRD repair else-

Table 1. Etiology of Pediatric Rhegmatogenous Retinal Detachment (RD)

Ocular Condition	No. (%) of Eyes
Myopia (≥ -4.00 diopters)*	10 (34)
Postoperative	10 (34)
No previous ocular disease	3 (11)
Subluxed lens	1 (3.5)
Hereditary RD syndrome	1 (3.5)
History of contralateral RD	1 (3.5)
Persistent fetal vasculature	1 (3.5)
Congenital cytomegalovirus	1 (3.5)
Ocular toxoplasmosis	1 (3.5)
Total eyes	29 (100)

* The myopia etiology consists of three subclasses: 1) Stickler's syndrome (5 eyes), 2) retinopathy of prematurity-related detachment in adolescence (4 eyes), and 3) unqualified high myopia (1 eye).

where and subsequent surgeries at Emory for persistent or recurrent RRD. All patients had at least 4 months of postoperative follow-up (mean 21.4 months, range 4 to 61 months).

The mean age of the 27 patients included in the study group was 9.6 years (range 0.1–17.8 years). Fifteen (56%) of the patients were white and 12 (44%) were black. Nineteen (70%) of the children were boys and 8 (30%) were girls. There were 16 (55%) right eyes and 13 (45%) left eyes. Six (22%) patients had bilateral RRD: 4 (15%) presented with chronic detachments in the contralateral eye, 1 (4%) presented with bilateral involvement at initial presentation, and 1 (4%) became bilateral during the course of follow-up. The four long-standing/chronic RD in fellow eyes were not operated on or included in the case series described, other than elucidating the status of the fellow eye and the overall incidence of RRD bilaterality in this study group.

Medical history was remarkable for prematurity in 5 patients (19%), mental retardation in 2 (7%), and congenital cytomegalovirus infection in 1 (3.5%). Family ocular history was remarkable for RD in family members of 5 of the 27 children (19%); four were

Table 2. Surgical Procedure Performed Before the Development of Rhegmatogenous Retinal Detachment

Procedure	No. (%) of Eyes
Pars plana vitrectomy	5 (50)
Molteno tube shunt	2 (20)
Trabeculectomy	1 (10)
Suprachoroidal drainage	1 (10)
Penetrating keratoplasty	1 (10)
Total eyes	10 (100)

Table 3. Abnormal Ocular Findings Among Fellow Eyes

Ocular Condition	No. of Patients*
High myopia (≥ -4.00 diopters)	10
Congenital cataract	5
Retinopathy of prematurity	4
Unrepaired chronic RD	4
Juvenile glaucoma	3
Peter's anomaly	3
Microphthalmos	2
Persistent fetal vasculature	2
Subluxed lens	1
Retinoschisis	1
Repaired RD†	1

* Each patient's fellow eye could have more than one significant ocular abnormality. Approximately 59% of eyes had two or more ocular abnormalities.

† This patient presented with a consecutive rhegmatogenous retinal detachment (RRD) during the course of follow-up.

attributed to myopia secondary to Stickler's syndrome and one had no ocular disease other than a family history of RD. Of the 27 children in the study group, only 3 (11%) had normal fellow eyes (no pathologic findings). The remaining 24 patients (89%) had at least one of the following abnormal ocular findings in the fellow eye (Table 3): high myopia (≥ -4.00 diopters), ROP, repaired RD, chronic RD, persistent fetal vasculature, congenital cataracts, juvenile glaucoma, Peter's anomaly, microphthalmos, retinoschisis, and subluxed lens.

Poor VA was the presenting symptom in 18 of 29 eyes (62%). Of the remaining eyes, RD was detected on routine examination in 8 (28%), and following parental awareness of leukocoria in 3 (10%) (Table 4). Rhegmatogenous RD was detected on routine follow-up in an additional 6 of the 10 patients presenting pursuant to a prior surgical procedure (the postoperative etiology). For those patients who could quantify the duration of vision loss in the affected eye ($n = 17$ eyes), the mean duration of vision loss before repair was 52 days (range 1–180 days).

The RRD included the macular area in 23 of 29 eyes (79%). Proliferative vitreoretinopathy was present at initial presentation in 13 of 29 eyes (45%). In these 13 eyes, fixed retinal folds were present in at

Table 4. Presentation of Pediatric Rhegmatogenous Retinal Detachment

Ocular Condition	No. (%) of Eyes
Visual loss	18 (62)
Routine examination	8 (28)
Leukocoria	3 (10)
Total	29 (100)

Table 5. Type of Retinal Break in Pediatric Rhegmatogenous Retinal Detachment

Ocular Condition	No. (%) of Eyes
Horseshoe tear	16 (55)
Dialysis	6 (21)
Giant retinal tear	4 (14)
Not stated in chart	3 (10)
Total	29 (100)

least one quadrant. A single horseshoe break was the cause of the RRD in 16 of 29 eyes (55%) (Table 5). Of the four giant retinal tears, three were associated with Stickler's syndrome (grouped within the high myopia etiologic subgroup). Of the six dialyses, three occurred in the postoperative etiology.

All eyes in this series were brought to the operating suite for attempted surgical repair. Four of the 29 eyes (14%) were deemed irreparable once the visual axis had been cleared; as such, the surgical procedure was aborted. Of the 25 repairable eyes, the initial surgical procedure performed was a scleral buckle in 7 (28%) and pars plana vitrectomy (either alone or in combination with other procedures) in 18 (72%). In all, 23 (92%) ultimately underwent vitrectomy. Silicone oil was used at the time of initial surgery in 8 of 18 (44%) primarily vitrectomized eyes, and in at least one phase of repair in 18 of 25 eyes (72%).

Most repairable eyes required more than one surgical procedure to effect retinal reattachment, with a mean of 2.2 surgeries (range 1–5) per eye. Ultimate anatomic reattachment was accomplished in 18 of the 25 eyes (72%) over the follow-up period. The final reattachment rate was slightly higher in the myopic subgroup (8 of 10 eyes), and drastically lower in the postoperative subgroup (4 of 9 eyes).

Table 6 lists the etiology of the affected eye, presenting VA of both eyes, and final VA of the affected eye.

Discussion

We retrospectively reviewed a series of 29 eyes in 27 pediatric patients with RRD. Although the incidence of RRD in childhood is quite low, this entity merits scrutiny for at least two reasons: the high rate of vision-threatening pathology in the fellow eye and the considerable financial and emotional investment incurred when the decision is made to attempt to repair such eyes.

Ocular trauma was the most common etiology for pediatric RRD in the initial research for this series, accounting for 25 (42%) of the surgical cases for pediatric RRD at Emory University between 1991 and

Table 6. Visual Acuities (VA)

Eyes Evaluated	Etiology	Affected Eye, Presenting VA	Fellow Eye, Presenting VA	Final VA of Affected Eye	Comparison of Affected Eye
1	Subluxed lens	LP	20/40	20/400	Improved
2	Hereditary RD syndrome	20/400	20/1000	20/30	Improved
3	None	20/1333	*	Unable†	‡
4	None	LP	*	Unable†	‡
5	None	HM	20/20	20/200	Improved
6	PFV	LP	No LP	No LP	Declined
7	CMV retinitis	Unable§	Unable§	Surgery aborted	‡
8	Toxoplasmosis	20/40	20/20	20/20	Improved
9	Myopia, Stickler's	Unable§	Unable§	HM	‡
10	Myopia, Stickler's	LP	20/100	20/60	Improved
11	Myopia, Stickler's	20/400	20/100	20/100	Improved
12	Myopia, Stickler's	20/4000	20/60	20/4000	No change
13	Myopia, Stickler's	LP	20/400	Surgery aborted	‡
14	Myopia	20/30	20/30	20/400	Declined
15	Myopia, ROP	HM	20/40	20/4000	‡
16	Myopia, ROP	HM	20/40	LP	Declined
17	Myopia, ROP	20/60	LP	20/40	Improved
18	Myopia, ROP	20/200	20/4000	HM	Declined
19	H/O contralateral RD	Unable§	Unable§	Surgery aborted	‡
20	Postoperative	20/1333	20/100	20/1333	No change
21	Postoperative	Unable§	Unable§	Unable†	‡
22	Postoperative	LP	Unable§	No LP	Declined
23	Postoperative	20/4000	20/20	HM	Declined
24	Postoperative	HM	20/400	No LP	Declined
25	Postoperative	LP	LP	No LP	Declined
26	Postoperative	HM	HM	HM	No change
27	Postoperative	LP	LP	20/600	Improved
28	Postoperative	LP	No LP	Surgery aborted	‡
29	Postoperative	20/2000	20/1333	No LP	Declined
Summary of Presenting VA		Affected Eyes (%)		Fellow Eyes (%)	
20/800 or better		6 (24)¶		13 (59)**	
Worse than 20/800		19 (76)¶		9 (41)**	

* Eyes 3 and 4 came from the same patient who presented with bilateral retinal detachments (RD); therefore, there is no companion eye.

† Unable means that a VA was not attainable due to the patient's inability to respond to a VA test at final follow-up.

‡ No comparison could be made.

§ Unable means that the patient was too young or mentally retarded and unable to respond to a VA test in that eye.

|| Eyes 16 and 17 came from the same patient who presented with consecutive RD in both eyes that were 6 months apart. As such, this patient's eyes were counted as two pairs.

¶ N = 25 eyes because the eyes for which vision was not available or not able to be recorded totaled five; these were not included in the calculation.

** N = 22 eyes for the same reasons as the presenting eyes.

LP, light perception; HM, hand motions; PFV, persistent fetal vasculature; CMV, cytomegalovirus; ROP, retinopathy of prematurity; H/O, history of.

1997. Winslow and Tasman² noted a similar incidence of RRD attributed to trauma (44%) in their review of juvenile RRD in 1978. When considered across all age groups, RRD occurs following trauma in approximately 11% of cases.^{1,3} Excluding trauma and acute cicatricial ROP, pediatric RRD in the current series of 29 eyes was most commonly associated with myopia in 10 (34%) and antecedent intraocular surgery in 10 (34%). Winslow and Tasman² noted a similar frequency of association with myopia, accounting for 28

of 102 eyes (28%) with RRD without globe-disrupting trauma.

The postoperative subgroup included all eyes with RRD and known antecedent intraocular surgery. This group included eyes with juvenile glaucoma, congenital cataracts, Peter's anomaly, juvenile retinoschisis, and the sequelae of these disorders, and accounted for 34% (10/29) of eyes in this series. We qualified them by the procedure performed immediately before the development of the RRD. None of the eyes developed

a recognized RD intraoperatively. Of the 10 eyes in the postoperative group, six were aphakic, five had undergone pars plana vitrectomy, two had Molteno tube placement, one had trabeculectomy, one had suprachoroidal drainage, and one had penetrating keratoplasty. Waterhouse et al⁴ cited a 5.0% risk for RRD after Molteno glaucoma implant surgery, with a median patient age of 9 years. Retinal detachment following other ophthalmic surgeries is not well documented except for extracapsular cataract extraction for which the incidence of RD is estimated to be 0.02% to 3.6%.⁵

The most common presenting complaint was decreased vision in 18 of 29 eyes (62%). Of the patients who could quantify the duration of vision loss in the affected eye ($n = 17$ eyes), the mean duration of vision loss was 52 days (range 1–180 days) from initial symptoms to presentation. In studies including all age groups, the mean time to presentation is usually much less than 30 days.³ The tendency to late diagnosis at presentation is an important feature of pediatric RD. Given this, it is not surprising that 79% (23/29) of eyes in this series presented with macula-off RRD. Winslow and Tasman² and Hilton and Norton⁶ each reported a similar frequency of macular detachment at the time of presentation among children with RRD (77% and 80%, respectively). In Laatikainen and Tolppanen's³ study of 342 eyes with RRD, wherein the mean age was 52.8 years, the macula was detached in only 56.5% of the eyes. Delay in diagnosis is further evidenced by the observation that 45% (13/29) of eyes had PVR at initial presentation. This finding is in line with reports by Sternberg et al⁷ and Waterhouse et al,⁴ wherein PVR was observed at initial presentation of a RD after previous ocular surgery in 39% and 41% of adult patients, respectively.

The duration of vision loss for the 10 eyes in the postoperative subgroup could not be quantified because 60% (6/10) of these eyes were diagnosed with RRD on routine postoperative follow-up. Children who developed RRD in this series often had bilateral ocular abnormalities predisposing them to poor vision in both eyes. Consequently, it is difficult for them to compare vision in both eyes or appreciate a difference in vision, analogous to the scenario of an adult developing a RRD after penetrating keratoplasty.⁶

Eighty-six percent (25/29) of eyes in this series underwent attempted surgical repair. In 4 eyes (14%) surgery was aborted once clearance of the visual axis (via cataract removal or placement of a temporary keratoprosthesis) allowed assessment of the posterior segment by direct visualization, and

the retina was determined to be irreparable. For the 25 eyes in which repair was attempted, anatomic reattachment was accomplished in 18 (72%). In Winslow and Tasman's² study, anatomic reattachment was accomplished in 94 of 117 eyes (80.3%). Our population differs from Winslow and Tasman's series in the larger percentage of previously operated eyes (34% versus 10%). The difference is important as eyes that have undergone prior surgical intervention often fare considerably worse than eyes with no history of antecedent surgery. Of the 25 eyes for which repair was attempted, anatomic reattachment was achieved in 8 of 10 (80%) of the myopic eyes but only in 4 of 9 (44%) eyes that had undergone prior intraocular surgery.

In 1969, the previtrectomy era, Hilton and Norton⁶ reviewed juvenile RD. Their anatomic success rate was 89%. This reattachment rate is 17% higher than our current study's. Moreover, Hilton and Norton report a higher anatomic success rate than other similar studies. With respect to our work, the patients in the Hilton and Norton study had important differences. None of the patients had undergone surgery for other problems before RD surgery and fewer presented with proliferative retinopathy. Correcting for these findings would probably make the anatomic success rate similar, but nonetheless the authors were able to effect anatomic success at a sufficiently high rate in a time of much fewer surgical options.

The overall number of operations was 2.2 per eye. The mean is slightly higher for myopic eyes (2.8, range 1–5) and lower for eyes with postoperative RRD (1.6, range 1–3). The lower mean number of procedures per eye for postoperative eyes is misleading, however, as such eyes often could not tolerate further procedures.

Whereas anatomic reattachment is its necessary precedent, functional outcome is the ultimate measure of success. Visual acuity was measured in both the affected eye and the fellow eye at presentation and the end of follow-up. At presentation, 24% (6/25) of affected eyes and 59% (13/22) of fellow eyes had VA of 20/800 or better (Table 6). At final follow-up (Table 6), VA was measurable in 22 repaired eyes. Of these, 41% (9/22) had VA of 20/800 or better. Overall, the group of repaired eyes showed a substantial improvement in VA. Of the 21 children for whom final VA was available for both eyes ($n = 21$), in 8 (38%) the affected eye that had undergone surgical repair had equal or better final VA compared to the fellow eye after surgical repairs.

Pediatric RRD is rare, with an incidence of 0.28 to 0.69 per 100,000 population and over 40% of cases secondary to ocular trauma.² In the current series,

which excludes ocular trauma and acute ROP, the two most common etiologies were myopia and postoperative RRD. Eighty-nine percent of our patients had bilateral involvement of ocular abnormalities—RD or other ocular disorders limiting vision—with 75% of affected eyes and almost half of the fellow eyes presenting with vision worse than 20/800. The tendency to bilateral ocular disease, and the finding that nearly 40% of the repaired eyes have better or equal VA compared to the fellow eye at final follow-up, lead to the conclusion that in pediatric RD, the affected eye must be treated with great care.

Key words: myopia, pediatric, rhegmatogenous retinal detachment, silicone oil, vitrectomy, vitreoretinal surgery.

References

1. Haimann MH, Burton TC, Brown CK. Epidemiology of retinal detachment. *Arch Ophthalmol* 1982;100:289–292.
2. Winslow RL, Tasman W. Juvenile rhegmatogenous retinal detachment. *Ophthalmology* 1978;85:607–618.
3. Laatikainen L, Tolppanen EM. Characteristics of rhegmatogenous retinal detachment. *Acta Ophthalmol* 1985;63:146–154.
4. Waterhouse WJ, Lloyd ME, Dugel PU, et al. Rhegmatogenous retinal detachment after Molteno glaucoma implant surgery. *Ophthalmology* 1994;101:665–671.
5. Scheie HG, Morse PH, Aminlari A. Incidence of retinal detachment following cataract extraction. *Arch Ophthalmol* 1973;89:293–295.
6. Hilton GF, Norton EW. Juvenile retinal detachment. *Mod Probl Ophthalmol* 1969;8:325–341.
7. Sternberg P, Meredith TA, Stewart MA, et al. Retinal detachment in penetrating keratoplasty patients. *Am J Ophthalmol* 1990;109:5148–152.