Original Article

Outcome of vitreoretinal surgery for rhegmatogenous retinal detachment in X-linked juvenile retinoschisis

Parveen Sen, Ashutosh Agarwal, Pramod Bhende, Lingam Gopal, Muna Bhende, Pukhraj Rishi, Tarun Sharma, Mukesh Jain

Purpose: Rhegmatogenous retinal detachment (RRD) is a vision-threatening complication of X-linked juvenile retinoschisis (XLRS). The aim of this study is to report the anatomical and functional outcomes of vitreoretinal surgery for the treatment of RRD in X-linked juvenile retinoschisis (XLRS). **Methods:** This is a retrospective, interventional, consecutive case series in which case records of 34 eyes of 28 XLRS patients, who underwent surgery for RRD, were reviewed. Statistical analysis used is as follows: visual outcomes were categorized into three groups: improved, remained stable or deteriorated. Wilcoxon signed-rank test was used to determine the difference in visual acuity at baseline and at final visit. Univariate binary logistic regression analysis was used to determine the risk factors for retinal detachment. Any P value < 0.05 was considered as statistical significant. **Results:** Mean age of the boys at presentation was 9.2 ± 3.5 years. Indication for surgery was RRD in all eyes. Concurrent vitreous haemorrhage was present in four eyes (11.8%). The primary surgical intervention was scleral buckle in 12 eyes and pars plana vitrectomy in 22 eyes. Persistence of subretinal fluid/redetachment was seen in 15 eyes of which 11 eyes underwent additional surgeries. At final follow up, 27 eyes (79.4%) were noted to have an attached retina. Of the total eyes, in 20, 6 and 8 eyes the visual acuity improved, stabilized and deteriorated, respectively. **Conclusion:** Favourable outcomes are seen in a majority of eyes after vitreoretinal surgery for RRD in XLRS; however, multiple surgical interventions may be necessary.

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X-linked juvenile retinoschisis (XLRS), first described by Haas in 1898, is the most common juvenile onset retinal degeneration seen in males with a prevalence of 1 in 25,000 to 1 in 5000.[1,2] XLRS is due to a mutation in the RS1 gene (located on the Xp22.1 chromosome) which codes for retinoschisin, a protein that helps in cellular adhesion as well as in cell-to-cell interaction.[3,4] Females are rarely affected.[5]

It typically manifests in the first decade of life with diminution of vision resulting from schisis involving the fovea. There is no treatment to prevent the progression of the disease. However, complication like rhegmatogenous and/or tractional retinal detachment (RD), vitreous haemorrhage (VH), progressive schisis involving the macula and haemorrhage within large schisis cavity can occur suddenly during the course of the disease, requiring treatment in the form of surgical intervention.[6–8] In the literature, there are a few small case series reporting the management of vitreoretinal surgery for vision-threatening complications associated with XLRS.[9–14] Rhegmatogenous RD (RRD) in XLRS is seen in 10–22% of cases with XLRS.[15] In the present study, we reviewed the outcomes as well as the prognostic factors of surgery for RRD in a series of 34 eyes of 28 children with XLRS.

Medical Research Foundation, Sankara Nethralaya, Chennai, Tamil Nadu, India

Correspondence to: Dr. Parveen Sen, Medical Research Foundation, Sankara Nethralaya, Chennai - 600 006, Tamil Nadu, India. E-mail: parveensen@gmail.com

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Methods

Retrospective review of case record of 34 consecutive eyes of 28 children with XLRS, who underwent vitreoretinal surgery for RRD during the study period of 1995 to 2015, at a tertiary care hospital was done. The diagnosis of XLRS was made based on family history and characteristic findings on ophthalmic evaluation. In selected cases, it was further confirmed on electroretinogram (ERG) and/or optical coherence tomography (OCT). RRD was differentiated from retinoschisis clinically by the presence of detachment of the outer retina in addition to retinal splitting with full thickness retinal breaks seen in the outer retina, either pre-operatively or intra-operatively. Informed consent from patients was taken prior to the surgical intervention.

Scleral buckling (SB) along with an encircling band was done in eyes with peripheral outer retinal breaks leading to RRD, with clear ocular media and without significant proliferative vitreoretinopathy (PVR). Pars plana vitrectomy (PPV) was done in eyes with posterior breaks or in eyes with PVR more than

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Grade 2 as per the International PVR Classification.[16] In all eyes, PPV was combined with an encirclage using a 240 band, which was placed 2.5 mm posterior to the ora serrata. PPV consisted of complete vitrectomy with induction of posterior vitreous detachment (PVD). Pre-retinal membranes and subretinal gliotic bands were removed to the extent possible to adequately relieve traction. Perfluorocarbon liquids were used in some eyes to facilitate retinal reattachment. Cautery of fragile retinal vessels over the schitic area was done followed by inner retinal layer retinectomy, wherever necessary. Fluid-air exchange and internal subretinal fluid drainage was done through the outer retinal layer break followed by retinopexy. Silicone oil/perfluoropropane gas (14% C3F8) was used as a tamponading agent, as and when necessary.

Retrospective chart review was done to collect information regarding age at presentation, history of preceding trauma, refractive state, baseline visual acuity, detailed anterior and posterior segment findings, primary surgical procedures, additional surgical procedures if required and outcomes (both anatomical and functional) at final visit. Institutional review board approval was obtained and the tenets of the declaration of Helsinki were followed.

Statistical analysis: Snellen's visual acuity was converted to equivalent logMAR units. Visual outcomes were categorized into three groups: improved, remained stable or deteriorated. Wilcoxon signed-rank test was used to determine the difference in visual acuity at baseline and at final visit. Univariate binary logistic regression analysis was used to determine the risk factors for poor anatomic outcome. Any P-value < 0.05 was considered as statistical significant. All statistical tests are performed using the SPSS V 14.0.

Results

The median age was 9 years (range 3–16 years) at the time of surgery. All patients were male. Of 34 eyes, there was involvement of the right eye in 19 cases and the left eye in 15 cases. Six patients presented with RRD in both eyes. Presenting complaints were sudden dimness of vision (n = 15), gradual dimness of vision (n = 13), incidental finding on routine ophthalmic check-up (n = 4), deviation of eye (n = 1) and abnormal head posture (n = 1). A history of minor trauma was elicited in 9 (26.5%) eyes. Twenty eyes (58.8%) and two eyes (5.9%) were hypermetropic and myopic, respectively. Refractive error ranged from a spherical equivalent of −6 to +20 dioptres. Early lenticular opacity was seen in five (14.7%) eyes at presentation.

Twenty one (61.8%) eyes had profound visual impairment at baseline, whereas ten eyes (29.4%) had moderate, and two eyes (5.9%) had mild visual impairment at baseline. The mean best-corrected visual acuity (BCVA) at baseline was 1.52 (±0.67) logMAR. One patient was 3 years old and could not be assessed by the Snellen's acuity, but could fix and follow torch light and was able to reach out for near objects. At presentation, eight patients had squint and six patients had nystagmus.

On fundus examination both peripheral and foveal retinoschisis was seen in all eyes. Location of retinal breaks was inferotemporal quadrant (n = 13), temporal half of the retina (n = 11), superotemporal quadrant (n = 6), inferior half (n = 4) and superonasal quadrant (n = 1). ERG was done in 12 eyes (35.3%). All 12 eyes showed the characteristic negative waveform in the combined scotopic response (3.0 scotopic ERG).

Table 1 shows the baseline characteristics, surgical intervention and outcomes in 34 eyes of 28 children. The indication for surgical intervention was RRD in all eyes. Concurrent VH was present in four eyes (11.8%). Three of these eyes underwent primary PPV while in one eye SB was performed, since VH at the time of surgery was limited to the inferior quadrants; thus allowing visualization of the outer retinal breaks. Total and subtotal RRD was seen in 10 (29.4%) and 24 (70.6%) eyes, respectively. Macular detachment (macula-off RD) was noted in 28 (82.4%) eyes. PVR was seen at presentation in 12 eyes (35.3%), of which 5 eyes had Grade 2 PVR and 7 eyes had Grade 3 PVR. Mean number of retinal quadrants involved in RD was 2.79, with the inferotemporal quadrant involved in all cases and the inferonasal quadrant being the second most commonly involved quadrant [31 eyes (91.2%)].

Surgical outcomes

Primary management was in the form of SB in 12 (35.3%) eyes. A broad buckling element in form of solid silicone tyre (MIRA Inc., Uxbridge, MA) [276 (n = 3), 277 (n = 1), 279 (n = 7), 280 (n = 1)] was used after taking anchor sutures with 5-0 polypropylene suture (Ethicon Inc., Somerville, NJ). Cryopexy was done to outer retinal holes. SRF drainage was necessary in all but two patients. Extent of buckle ranged from one-fourth to three-fourth quadrants. Ten scleral buckles were explants and two were implants. Of the 12 eyes that had SB as the primary surgery, eight eyes (66.7%) had an attached retina after a single surgery. Of the remaining four eyes, two eyes had persistence of subretinal fluid, whereas two eyes had recurrence of RD. A PPV with silicone oil tamponade was done in three cases. One patient refused to undergo repeat surgical intervention. One patient had retinal reattachment after two surgeries and another after three surgeries. Overall, in the group that underwent SB as primary surgery (n = 12), anatomic success was seen in 10 eyes (83.3%).

Primary PPV was done in 22 (64.7%) eyes. PPV was combined with an encirclage in all eyes. In six (27.3%) eyes, a combined lensectomy with PPV was done. Silicone oil was used as the endotamponade agent in 21 eyes (95.5%) and C3F8 gas was used in one eye (4.5%). Inner retinal layer retinectomy was done in 9 of 22 cases (40.9%) undergoing primary vitrectomy. Location of retinectomy was inferotemporal quadrant in four eyes and temporal periphery in the remaining eyes. This was usually limited to within the area of the schisis and further laser was done to the edge of the retinoschisis. The success rate of primary surgery by PPV was 50% (11 out of 22 eyes). In the PPV group, of the 11 eyes that had recurrent RD after the first procedure; second surgery was done in eight of these cases and the remaining three were inoperable because of advanced PVR. Anatomic success could be achieved in five of these eight cases. For the three cases that could not be anatomically reattached after the second surgery, one case underwent a third surgery following which retina was attached; one case was inoperable and one patient refused for another procedure. In the PPV group (n = 22), final anatomic success was achieved in 17 eyes (77.3%).

Hence, of a total of 34 eyes, anatomical success was achieved in 19 eyes (55.9%) after the primary surgery and 27 eyes (79.4%)

Table 1: Details of the surgical procedure

Age (years)	Eye	Baseline BCVA*	Diagnosis	Extent of RD†; macular status	PVR‡ grade	Primary surgery	1st additional surgery	2nd additional surgery	Anatomical outcome	Final BCVA, cause of poor BCVA
11	OD§	6/24	RRD‖	Partial; Off	2	SB + BB¶	-	-	Stable	6/60
6	OD	PL+**	RRD	Total	3	SB	VIT†† + PFCL‡‡ + SOI§§	Left alone	Detached	PL+
6	OS¶¶	3/60	RRD	Partial; Off	0	SB	Refused re-surgery	-	Detached	HMCF***
15	OD	6/36	RRD	Partial; Off	3	SB	-	-	Stable	6/36
10	OD	6/36	RRD	Partial; Off	2	V + BB + SOI	-	-	Stable	3/60; Foveal atrophy
11	OS	6/18	RRD	Partial; Off	2	V + BB + SOI	Re-VIT††† + Re-SOI‡‡‡	-	Stable	6/18
14	OD	1/60	RRD	Partial; Off	3	V + SB + PFCL + SOI	Re-VIT + L§§§ + RR‖‖‖ + PFCL + Re-SOI	Refused re-surgery	Detached	PL+
5	OS	2/60	RRD	Partial; Off	2	V + SB + PFCL + SOI	Re-VIT + Re-SOI	-	Stable	6/36
7	OS	PL +	RRD	Total	0	L + V + BB + SOI	Nil intervention advised	-	Detached	PL+
11	OD	6/12	RRD	Partial; On	0	SB	-	-	Stable	6/18
8	OS	PL+	RRD	Total	3	L + V + BB + SOI	-	-	Stable	1/60; ERM¶¶¶ + disc pallor
9	OS	3/60	RRD	Partial; On	0	V + BB + C3F8****	-	-	Stable	4/60; ERM + pale disc
9	OD	1/60	RRD	Partial; Off	0	SB	-	-	Stable	4/60; foveal atrophy
9	OS	6/60	RRD	Partial; On	0	SB	VIT + SOI	-	Stable	3/60; Foveal atrophy + cataract
6	OD	PL+	RRD + VH††††	Total	0	V + BB + PFCL + SOI	Nil intervention advised	-	Detached	No PL
16	OS	1/60	RRD	Total	0	V + BB + SOI	-	-	Stable	6/36
12	OD	PL+	RRD	Total	3	V + BB + SIO	ReVIT + L + MP‡‡‡‡ +RR + PFCL + Re-SOI	ReVIT + MP + ReSOI	Stable	2/60; pale disc
3	OS	F & F§§§§	RRD	Total	0	L + V + BB + SOI	Re-VIT + SOR + C3F8	-	Stable	F & F
6	OS	1/60	RRD	Partial; Off	0	V + SB + SOI	Nil intervention advised	-	Detached	1/60
11	OD	6/60	RRD	Partial; Off	0	V + BB + SOI	-	-	Stable	6/24
7	OD	1/60	RRD	Partial; On	2	V + BB + SOI	-	-	Stable	3/60; Foveal atrophy
4	OD	6/60	RRD + VH	Partial; On	0	SB	-	-	Stable	6/36
13	OS	1/60	RRD	Partial; Off	0	SB	VIT + SOI	ReVIT + MP + RR ++ ReSOI	Stable	6/24
13	OD	1/60	RRD	Partial; Off	0	V + BB + SOI	-	-	Stable	1/60; cataract
4	OS	1/60	RRD + VH	Total	0	L + V + BB + PFCL + SOI	-	-	Stable	2/60; foveal atrophy

Contd...

Table 1: Contd...

Age (years)	Eye	Baseline BCVA*	Diagnosis	Extent of RD†; macular status	PVR‡ grade	Primary surgery	1st additional surgery	2nd additional surgery	Anatomical outcome	Final BCVA, cause of poor BCVA
9	OD	1/60	RRD	Total	3	L + V + BB + SOI	Re-VIT + Re-SOI	-	Stable	3/60; pale disc + foveal atrophy
10	OS	6/60	RRD	Partial; Off	0	V + BB + SOI	-	-	Stable	6/36
12	OD	6/36	RRD + VH	Partial; On	0	V + BB + SOI	-	-	Stable	6/24
6	OD	6/60	RRD	Partial; Off	0	V + BB + SOI	Re-VIT + Re-SOI	-	Detached	PL+
6	OD	6/60	RRD	Partial; Off	0	V + BB + SOI	Re-VIT + Re-SOI	-	Stable	6/45
5	OS	PL +	RRD	Total	3	L + V + PFCL + SOI	-	-	Stable	5/60; pale disc
14	OD	2/60	RRD	Partial; Off	0	SB	-	-	Stable	6/24
14	OS	2/60	RRD	Partial; Off	0	SB	-	-	Stable	3/60
10	OD	5/60	RRD	Partial; Off	0	SB	-	-	Stable	6/36

*BCVA: Best-corrected visual acuity, †RD: Retinal detachment, ‡PVR: Proliferative vitreoretinopathy, §OD: Right eye ||RRD: Rhegmatogenous retinal detachment, ¶SB: Scleral buckle, BB: Belt buckle, **PL+: Perception of light ††V or Vit: Vitrectomy, ‡‡PFCL: Perfluorocarbon liquid, §§SOI: Silicone oil infusion, ¶¶OS: Left eye, ***HMCF: Hand motions close to face, †††Re-Vit: Repeat vitrectomy, ‡‡‡Re-SOI: Repeat silicone oil infusion, §§§L: Lensectomy, ||||RR: Relaxing retinotomy, ¶¶¶ERM: Epiretinal membrane, ****C3F8: Pefluoro propane, ††††VH: Vitreous haemorrhage, ‡‡‡‡MP: Membrane peeling, §§§§F and F: Fixates and Follows light

after multiple surgeries. PVR was the cause for a detached retina at final follow up in all seven eyes with recurrent RD in our case series. Patients were followed up post-operatively for a mean duration of 48 months, with a range of 1.2 months to 17.2 years. A mean number of 1.38 surgeries were performed in these eyes. Silicone oil removal (SOR) was done in 14 of 21 eyes which underwent primary vitrectomy with silicone oil insertion. 1300 centistoke oil used in all cases except one, in which 5000 centistoke oil was used. Of these 14 eyes, the retina was attached in 13 eyes at final follow-up visit. Of seven eyes in which SOR was not done, four had a redetachment at final visit, and oil was left *in situ*. Of the remaining three eyes with an attached retina, oil was not removed because of hypotony. All these cases had multiple surgeries for PVR and recurrent RD which resulted in hypotony in spite of successful reattachment of the retina. In the primary vitrectomy group, overall mean duration of oil *in situ* was 24.6 months (SD 32.9 months, range 4–132 months). Complications of long-term oil tamponade included band-shaped keratopathy (n = 3). In the SB group, SOR was done in all three cases which had undergone a subsequent vitrectomy.

Visual outcomes

Table 2 shows the visual acuity at presentation and at final visit. Of the total eyes, in 20, 6 and 8 eyes the visual acuity improved, stabilized and deteriorated, respectively. In the 20 eyes (58.8%) with visual improvement, the improvement was significant with a mean change of 0.56 units of logMAR units ($P < 0.001$). However, in eight eyes despite surgical intervention, there was a significant drop in visual acuity with a mean change

Table 2: Best corrected visual acuity (BCVA) outcomes for 34 eyes

Groups	Baseline BCVA	Final BCVA	Mean change in BCVA	*P*
Improved (*n*=20)	1.59±0.54	1.03±0.35	-0.56±0.41	<0.001
Stable (*n*=6)	1.64±0.85	1.64±0.85	Nil	-
Deteriorated (*n*=8)	1.25±0.83	1.91±1.0	0.66±0.52	0.012
Overall (*n*=34)	1.52±0.67	1.34±0.74	-0.17±0.65	0.094

*BCVA: Best corrected visual acuity

Table 3: Binary logistic regression analysis for a detached retina at final visit

Parameter	Odds Ratio	95%CI* Lower	95% CI Upper	*P*
Age	0.801	0.605	1.061	0.121
Total RD†	2.143	0.381	12.048	0.387
Absence of PVR‡	2.267	0.419	12.265	0.342
Squint	7.667	1.223	48.046	0.03
Nystagmus	2.3	0.326	16.224	0.403
Baseline BCVA§	3.34	0.823	13.56	0.092
Hypermetropia	0.333	0.017	6.374	0.466
Myopia	3	0.157	57.365	0.466
PPV‖	1.471	0.239	9.043	0.677

*CI: Confidence interval, †RD: Retinal detachment, ‡PVR: Proliferative vitreoretinopathy, §BCVA: Best-corrected visual acuity, ||PPV: Pars plana vitrectomy, ¶OR: Odds ratio

of 0.66 units of logMAR units (P = 0.01). The cause of drop in vision was recurrent RD (n = 4) and foveal atrophy (n = 4). Of the 27 eyes with attached retina at final follow-up, 19 eyes (70.4%) showed visual improvement, whereas four (14.8%) eyes had stabilization of BCVA. The remaining four eyes (14.8%) had worsening of BCVA due to foveal atrophy.

Prognostic factors

In eyes with retinal detachment, the presence of squint at baseline was found to predict a poor anatomical outcome (P = 0.03) [Table 3]. Favourable anatomical outcome was seen in 7 of 10 eyes (70%) presenting with total RD, in contrast to 20 of 24 eyes (83.3%) with subtotal RD (P = 0.387). Similarly, favourable anatomical outcome was seen in 21 of 28 patients (75%) with baseline macular involvement in contrast to 6 of 6 (100%) patients with no macular involvement (P = 0.169). Children having PVR Grade 3 and above were associated with less favourable anatomical outcome, 55.6% (5 of 9) in contrast to 88% (22 of 25) of those with lower grades of PVR (P = 0.039). Prognostic factors for visual outcome in eyes with an attached retina after single surgery (binary logistic regression analysis.) showed that absence of PVR at baseline and vitrectomy as the primary surgery had 6 times [odds ratio (OR) =6, P = 0.19] and 3.3 times (OR = 3.3, P = 0.19) higher chance of achieving either improvement or stabilization of post-operative BCVA. Although, due to a relatively small sample size, P value was insignificant.

Discussion

The RS 1 gene encodes for retinoschisin, a 224-amino acid soluble secretory protein with a discoidin like domain secreted from bipolar cells and photoreceptors.[17] It functions primarily in cellular adhesion, cell-to-cell interaction and retinal differentiation. Absence of retinoschisin leads to cell-to-cell dehiscence. Vitreous tractional forces result in progressive splitting up of the weakened retina in patients with XLRS, causing progressive visual loss. Retinal complications due to XLRS have been documented in the very first year of life.[18] More commonly these complications are seen in the first decade; VH and RD can occur suddenly anytime during the course of the disease, but are more commonly seen in the first decade.[7] Eyes with peripheral schisis are more prone to develop these complications.[6] All eyes in our series had peripheral schisis. All cases belonged to type 3/ type 4 (presence of both foveal as well as peripheral schisis) XLRS according to the new system of classification.[19]

RRD is the most common cause of surgery in patients with XLRS. Rosenfeld *et al*. reported RRD to be cause for surgery in 12 of 16 eyes (75%) in their series of eyes undergoing vitreoretinal surgery in XLRS.[10]

Management of RD in these cases is complicated by its presentation in a younger age group as well as by the underlying pathology. Before the era of advanced vitreoretinal instrumentation and techniques, only SB was recommended for retinoschisis with RD.[9] SB is the preferred choice of the surgeon only when the outer retinal breaks are peripheral and can be supported by a peripheral buckling element. It is less invasive; less wrought with complications and avoids the necessity to induce PVD which can be particularly difficult in very young eyes. The goal is to achieve reattachment of the outer retinal breaks to the underlying retinal pigment epithelium (RPE). Yokoyama *et al*. reported retinal reattachment in 90% of eyes that underwent the initial SB procedure.[20] In our study, SB was done in 12 eyes as a primary surgical intervention, of which 8 (66.7%) eyes had a successful outcome. Unlike the inner retinal breaks that are large, identifying the outer retinal breaks can be difficult because they can be posteriorly placed, very small and can be easily missed leading to recurrent RD. Also, since SB does not relieve all the vitreous traction in patients with XLRS, redetachment can be common, as was seen in four (33.3%) eyes after primary SB in the present study. Once PVR sets in, PPV is necessary.

Schulman *et al*. in 1985 for the first time performed PPV for retinal complications in XLRS.[12] In the present study of the 25 eyes that underwent PPV, either primary (22 eyes) or after failed SB (3 eyes), favourable anatomical outcome was seen in 18 (72%) eyes. PPV in XLRS involves complete induction of PVD. Inner retinal layer retinectomy ensures complete vitreous removal and relieve of traction. It has been shown to be of benefit in eyes undergoing surgery for XLRS by Trese and Ferrone.[14] Removal of the inner retinal layer which is mostly incomplete, thin and in shreds, allows better visualization of the outer retinal layer breaks as well as complete fill up with silicone oil at the end of the procedure. This is followed by laser to the outer retinal breaks as well as the edge of the schitic cavity [Fig. 1]. However, inner retinal layer retinectomy was not done in all our cases. In some eyes the inner wall was thicker; hence internal drainage through the "retinotomy in the inner layer" was done to flatten the schitic cavity followed by laser photocoagulation to the inner and outer layer breaks. We found no statistically significant difference on the anatomical outcome in eyes undergoing inner layer retinectomy as compared to eyes not undergoing the same (P = 0.59).

Many authors have reported flattening of the foveal schisis following vitrectomy with or without internal limiting membrane (ILM) peeling. Yu *et al*. noted that ILM peeling is a risk factor for retinal breaks and macular hole due to the weakened retinal architecture in the schitic area in XLRS patients.[21] The role of ILM peeling at present remains controversial. ILM peeling was not done in any of our cases. Repeat OCT in post-operative period in one of our cases showed flattening of schisis due to foveal atrophy while in other case flattening of the foveal schisis was seen under oil, which again showed recurrence after SOR [Fig. 2]. This has been previously reported by other authors as well.[22]

Given the complexity of surgery and young age of this group of cases silicone oil was necessary as a tamponading agent in all but one case. (Gas was used in this case due to the presence of a single break in the superotemporal quadrant.)

Rosenfeld *et al*. reported a success rate of about 87% after an average of 1.2 surgeries per eye with visual improvement in about 50% of eyes.[10] In our series, overall success rate was 79% with an average of 1.38 surgeries. Ferrone *et al*. reported a favourable anatomical and functional outcome in 88.8% (8 of 9) and 77.8% (7 of 9) eyes, respectively, after surgical intervention for complications in XLRS patients.[11] Comparison with other studies is shown in Table 4.

Rosenfeld *et al*. and Regillo *et al*. also found PVR as the most common pathology complicating the outcome necessitating repeat surgery.[9,10] PVR has been found to be the common cause of recurrent RD in these eyes by various authors.[9] The probable reasons for high incidence of PVR in XLRS could be delayed presentation as well as increased cellular activity and proliferation is seen in paediatric age group.[23]

Table 4: Comparison with other studies

Study	Year	Median Age (years)	Number of eyes		Type of surgery		Overall anatomical success (%)
			Surgery	RD*	SB†	PPV‡	
Regillo *et al.*[9]	1993	5	6	4	2	2	83.3
Ferrone *et al.*[11]	1997	6	9	4 RRD§ + 5 TRD‖	-	9	88
Rosenfeld *et al.*[10]	1998	11.5	16	12 RRD	5	7	88
Wu *et al.*[24]	2007	3.5	22	7 RRD + 4 TRD	-	11	91
Yu *et al.*[21]	2012	13.2	10	5 RRD	1	4	100
Present study	2017	9	34	34 RRD	12	22	79.4

*RD: Retinal Detachment, †SB: Scleral Buckle, ‡PPV: Pars plana vitrectomy, §RRD: Rhegmatogenous retinal detachment, ‖: Tractional retinal detachment

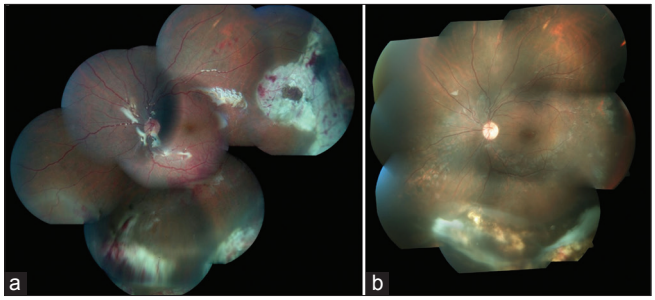

Figure 1: (a) Laser to the outer retinal hole; (b) laser to the border of the attached retina and the schitic cavity

In our series, 27 (79.4%) eyes had a favourable anatomical outcome. About 20 (58.8%) eyes had visual improvement after surgical intervention, while another 6 (17.6%) eyes had stabilization of visual acuity. Therefore, surgical intervention helped to improve or stabilize vision in a total of 26 (76.5%) eyes. Presence of squint was associated with poor anatomical outcome in eyes with RD in the logistic regression analysis. Eyes undergoing SB as primary surgery had somewhat better outcomes than eyes undergoing PPV as primary surgical intervention (P = 0.68). But SB may not be possible in all cases especially those with posterior retinal breaks or PVR at presentation. PPV is the only choice in these cases.

Wu *et al*. performed plasmin-assisted vitreoretinal surgery in congenital XLRS in 22 eyes of 20 patients. About 91% eyes had anatomical success after a mean 1.3 procedures per eye with improvement of visual acuity in 53% eyes.[24]

The retrospective design, lack of long-term follow-up and lack of experience with plasmin are limitations of our study. Also, genetic confirmation of the disorder was not done in all patients. Although, we believe that the typical presentation further corroborated by the ERG and OCT does not necessitate a genetic diagnosis in all cases. Our study adds to the limited literature available, elaborating the clinical presentation, surgical management, favourable outcome and prognostic factors of a rare retinal disorder.

Conclusion

RRD is a vision threatening complication of XLRS. Surgical intervention consists of SB in selected uncomplicated cases. The remaining cases require PPV with PVD induction, inner retinal layer retinectomy and an endotamponade using silicone oil. Favourable outcomes can be achieved; however multiple surgical interventions may be necessary for ultimate success.

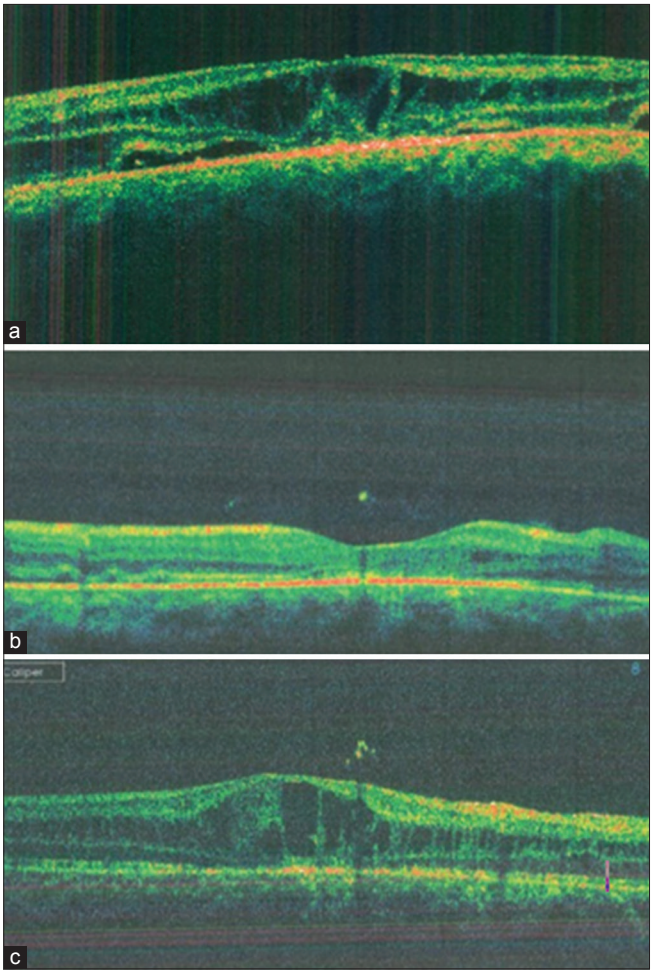

Figure 2: (a) Optical coherence tomography under silicone oil (2007); (b) optical coherence tomography just after silicone oil removal (2009); (c) optical coherence tomography after 6 months of silicone oil removal

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Conflicts of interest

There are no conflicts of interest.

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