Commentary: Nuances in management of rhegmatogenous retinal detachment in X-linked juvenile retinoschisis

X-linked juvenile retinoschisis (XLRS) is an inherited retinal degeneration characterized by formation of schitic cavities in the superficial retinal layers due to abnormal Muller's cell pillars.[1] Prenner *et al.* devised a new classification based on clinical and optical coherence tomography (OCT) findings.[2] Type 1 has only clinically evident foveal schisis, type 2 has additional macular lamellar schisis seen on OCT, type 3 has additional peripheral schisis, whereas type 4 is a combination of types 1 and 4.

XLRS-associated rhegmatogenous retinal detachment (RRD) accounts for nearly 4% of pediatric RDs.[3] RRD in case of XLRS develops secondary to either a full-thickness retinal break or to an outer retinal break with concurrent inner retinal break, in the area of peripheral retinoschisis. While peripheral retinoschisis is present in around 50% of cases, RRD develops in nearly 20% of cases.[1]

As the authors of the article "Outcome of vitreoretinal surgery for rhegmatogenous retinal detachment in X-linked juvenile retinoschisis" have correctly highlighted, there is no treatment to prevent the progression of the disease.[4] Progressive XLRS has been defined as a case with progressively decreasing vision, in the absence of any complication, along with progressive expansion of macular schisis or peripheral schisis cavity so as to threaten the fovea.[5] Honghua *et al.* advocated vitrectomy for such cases even in the absence of retinal detachment, pointing out that once a complication has occurred, the prognosis remains poor even after surgical intervention.[5] Studies have shown that performing vitrectomy in such cases not only decreases the chances of retinal detachment but also causes resolution of macular schisis.[5,6] However, performing vitrectomy in cases without any complications still remains debatable.

Surgical intervention is recommended in cases of complications such as dense nonclearing vitreous hemorrhage with a large schisis cavity hemorrhage and schisis combined with RRD or tractional RD.[5,6] As correctly pointed out by the authors, in case of minimal proliferative vitreoretinopathy (PVR) changes, anterior outer retinal breaks, and a clear media, scleral buckling is the first choice of treatment.[4] The principle is to seal and cover only the outer retinal breaks in addition to supporting the vitreous base.

Vitrectomy should be performed in case of presence of significant PVR changes or a failed buckling surgery. The authors correctly mention that the success of vitrectomy in such cases depend on the ability to completely relieve the retina of traction and complete induction of posterior vitreous detachment (PVD), which is very difficult to achieve in pediatric eyes.[4] In a number of cases, the inner wall of the retinoschisis has to be removed to relieve the vitreous traction.[5] As the area of retinoschisis corresponds to an absolute scotoma on the visual field, removing the inner wall of retinoschisis has no effect on the patient's visual field.[7] However, this eliminates any possible chance of future schisis cavity adhesion due to the loss of retinal ganglion cells and interneurons. Some authors describe inner wall retinotomy and fluid drainage. However, these retinotomies are prone to PVR changes along the anterior leaflet of the schisis.[8] Trese *et al.* recommended the use of autologous plasmin for PVD induction.[6] In pediatric cases, we have a different technique for PVD induction. We perform PVD by stroking the cortical vitreous over the posterior pole, using a diamond dusted membrane scraper in a circumferential manner, till a defect in the vitreous sheet is noted. Stroking of the vitreous sheet around this defect is continued in centrifugal fashion to enlarge the defect and form a ring of size 4–5 disc diameter (DD), with rolled out vitreous sheet with thick edges. The entire layer of vitreous sheet is then removed *in toto*, by lifting the edges of the ring using an internal limiting membrane (ILM) peeling forceps. Once the edges of the ring are lifted, perfluorocarbon liquid naturally slides into the potential space created between the vitreous sheet and retina, causing gradual extension of PVD in all quadrants. We are able to remove the vitreous easily and completely with this technique. Silicone oil (SO) tamponade should be provided in young children. We prefer using 5000 centistokes SO and giving a long-term tamponade.

The authors again correctly point out that performing ILM peeling in such cases is controversial.[4] ILM peeling in RRD has been proved to prevent the formation of postoperative macular epiretinal membrane and reduce the incidence of RD recurrence.[9] However, in case of XLRS, ILM peeling can lead to an iatrogenic macular hole, due to the weakened retinal structure of the retinoschisis area. Some authors advocate that ILM peeling should be avoided over the weakened retinoschisis area, but peeled 2–3 DD around this area.[5]

The authors in their retrospective study have given a detailed account of presentation, management, and prognosis of 34 patients presenting with XLRS-associated RRD.[4] Since it is a very rare condition, with a prevalence of 1 in 15,000–30,000, performing studies is difficult.[1] However, this study design had few fallacies, which should be kept in mind while planning future studies.[4] This study included cases from 1995 to 2015 and involved multiple surgeons. There has been a plethora of changes in the surgical technique in the past two decades, especially vitrectomy. A minimum follow-up of 6 months should be there to comment on the success of retinal detachment surgery. All these facts can introduce a possible bias in the study and should be carefully avoided in future studies.

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