

PLEASANT SURPRISES IN THE PEDIATRIC OR



Lessons in never giving up, even when the presentation seems hopeless.

BY ŞENGÜL ÖZDEK, MD, FEBO, FASRS, AND ECE ÖZDEMİR ZEYDANLI, MD, FEBO, FICO

Perseverance is one of the most important qualities a medical professional possesses, particularly when treating children. In challenging cases—such as when a child's ocular presentation is highly disconcerting or surgical intervention seems unlikely to succeed—there may be a temptation to abandon efforts. However, it is crucial to remember that during surgery, unexpected outcomes and pleasant surprises are possible. Even eyes that appear inoperable can, in some instances, be salvaged.

Our work with pediatric eye conditions has presented many instances where an eye was deemed inoperable and beyond hope—only for us to discover that the condition was less severe than initially thought. With appropriate treatment and surgical intervention, some eyes not only avoid significant complications but even demonstrate functional improvement. In this article, we review two such cases.

CASE NO. 1

A 7-month-old boy presented with unilateral leukocoria and retrolental and vitreous hemorrhage. This eye was previously considered inoperable due to a presumed closed-funnel retinal detachment (RD) detected with ultrasonography for an unknown etiology (Figure 1). The fellow eye was normal.

A detailed history revealed that the visual impairment had been noticed very early in the first weeks of life. Careful examination and ultrasonography provided additional insights that led us to a diagnosis of persistent fetal vasculature (PFV) and tractional RD with extensive retrolental and vitreous hemorrhage. After explaining the very low expectations of the surgical outcome in detail to the family, we decided to proceed with surgery. The corneal diameters were 11.5 mm x 11.0 mm for both eyes.

We performed a limbal lensectomy and vitrectomy, revealing a multirooted hyaloidal stalk and a tent-like tractional RD. Peeling the membranes resulted in an attached retina, including the macular area, with extramacular folds in the nasal retina. As was true in this case, the presence of dense hemorrhages accompanying a hyaloid stalk may resemble the appearance of a closed-funnel RD.^{1,2} Given their

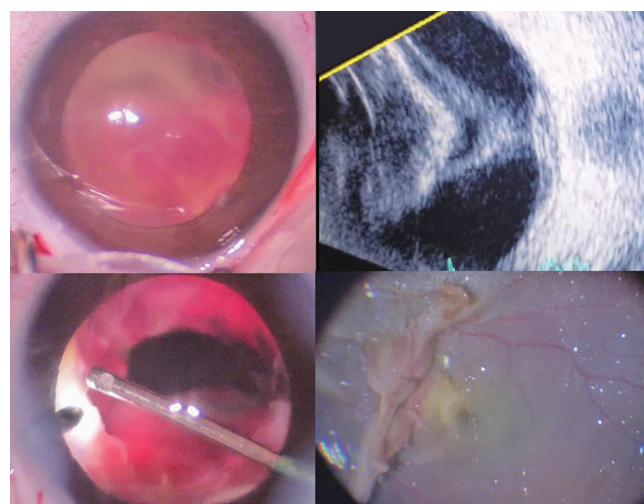


Figure 1. This patient with posterior PFV presented with retrolental and vitreous hemorrhage, mimicking a closed-funnel RD on B-scan ultrasonography. A limbal lensectomy and vitrectomy, followed by layer-by-layer removal of dense hemorrhagic membranes, uncovered a hyaloid stalk extending from the optic nerve head and forming retinal folds in all directions. The surgery resulted in an attached retina including the macula except from the narrow retinal folds in the nasal area.

mostly unilateral nature, these cases can be deemed inoperable. However, surgical intervention proved successful in this case, resulting in good anatomy, release of tractions, an attached macula, and the ability to fix and follow light, as we would expect in tent-like posterior PFV cases.³

CASE NO. 2

A 10-month-old boy presented with bilateral leukocoria, severe anterior segment dysgenesis, glaucoma, and no light reaction in either eye (Figure 2; Video). Several practices had considered these eyes inoperable. The patient had high IOP (35 mm Hg OD and 40 mm Hg OS), a cloudy cornea, and buphthalmia with corneal diameters of 16 mm OD and 17 mm OS. He also exhibited dysplastic iris and angle structures, rubeosis iridis and lentis with extensive peripheral anterior synechiae, total iridocorneal apposition causing partial corneal opacities, and fibrovascular tissue covering



Figure 2. This 10-month-old boy presented with severe bilateral anterior segment dysgenesis and glaucoma (A, B). Ultrasonography revealed bilateral multifocal tractional RD and vitreous opacities. The patient underwent vitrectomy in each eye, which included extensive synechiolysis, pupilloplasty, removal of tractional membranes, and laser photocoagulation of the avascular peripheral retina (C, D, and E). Following the surgery, the patient achieved ambulatory vision in each eye (F). Although the cornea was cloudy, he maintained ambulatory vision during the 5-year follow-up period (G).

the pupillary area. Ultrasonography revealed vitreous opacities and membranes causing local tractional RDs at multiple points of the posterior pole in each eye.

Cases with severe anterior segment dysgenesis tend to be considered inoperable when accompanied by posterior segment pathology.^{4,5} Without treatment, they usually end up with total corneal opacification with no visual function or good cosmetic appearance. After discussing the prognosis in detail and explaining the very low expectations of the surgery with the family, we performed a trial for surgery.

The surgical steps included limbal incisions, separation of the iridocorneal apposition with a spatula, formation of the anterior chamber, pupillary reconstruction with the extraction of the pupillary fibrovascular membrane, vitrectomy to clear the hemorrhage, extensive epiretinal membrane peeling to relieve traction on the retina, and laser photocoagulation

of the avascular peripheral retina. The eye was left in air. In this case, a presumed diagnosis of familial exudative vitreo-retinopathy was made during the surgery.

The same surgery was performed on the fellow eye 10 days later. The surgery resulted in the normalization of IOP in each eye, and the patient maintained ambulatory vision despite the corneal opacities, which improved slightly during the 5-year follow-up.

DISCUSSION

Not every surgical intervention will yield outcomes as favorable as these. But even when the outcome is not as desired, there may still be value in giving a trial for surgery, provided that the eye is not severely microphthalmic, disorganized, or already phthisical. Additionally, pursuing surgical intervention can give the family a sense of satisfaction, knowing that they've tried everything possible to gain vision.

Even when the surgery does not result in significant functional gain, it can still enhance the child's quality of life. For instance, the ability to perceive light is crucial for the diurnal rhythm of hormones, regulation of sleep-wake cycles, and orienting oneself in space and time. Maintaining a cosmetically acceptable globe can also improve the child's self-esteem and reduce bullying. Furthermore, surgery can pave the way for potential restorative technologies in the future.

Thorough counseling and active involvement of the family in the decision-making process are vital parts of this journey. Families might be eager to proceed without being fully aware of realistic outcomes, and surgeons must discuss the goals, risks, and benefits of surgery and set realistic expectations.

Ultimately, when it comes to pediatric retina surgery, surgeons must never give up. With skill, innovation, and perseverance, even the most challenging cases can sometimes have pleasantly surprising outcomes. ■

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ŞENGÜL ÖZDEK, MD, FEBO, FASRS

- Professor in Ophthalmology, Gazi University, Ankara, Turkey
- sengulozdek@gmail.com
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ECE ÖZDEMİR ZEYDANLI, MD, FEBO, FICO

- Retina Fellow, Ankara Retina Clinic, Ankara, Turkey
- ece.ozdemir@gmail.com
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