Pediatric retina surgery is not for the faint of heart. The stakes could not be higher for the youngest of our patients, and the margin between success and failure is razor thin. In surgery, as in medicine, small innovations and stepwise refinements to technique can, over time, cumulatively create tremendous progress.

In this edition of Practical Retina, **Yoshi Yonekawa, MD,** provides a comprehensive approach to the pediatric retinal surgical patient: from differential diagnosis, through diagnostic imaging, and ultimately to surgical decision-making with a compendium of surgical pearls.

There is an adage among pediatricians that children are not small adults. Dr. Yonekawa discusses the importance of stratifying pediatric patients by age and stresses the anatomical changes that dictate sclerotomy placement as the eye grows. The impact of technologic advances has translated well to pediatric retina, from widefield fluorescein angiography to small gauge instrumentation. The classic debate of scleral buckle versus vitrectomy surgery is addressed. Appropriate goal setting is important in pediatric retinal surgery to achieve the surgical objective while minimizing risk.

Howard F. Fine
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Readers of *OSLI Retina* are certain to find Dr. Yonekawa's approach to pediatric retinal surgery enlightening and instructive.

Practical Pearls in Pediatric Vitreoretinal Surgery

by Yoshihiro Yonekawa, MD

Yoshihiro Yonekawa

Victories in pediatric vitreoretinal surgery can have profoundly meaningful improvements in the quality of life of our young patients for their many years to come. However, children with retinal pathology often pose unique, humbling challenges. Our lives would be infinitely easier if pediatric eyes were simply small adult eyes. But they are not, and they constantly challenge our understanding of ocular development and how the eye stays together, requiring us to go beyond the conventional teachings of adult vitreoretinal surgery.

Knowing how the rulebooks differ between adult and pediatric retina surgery can maximize the outcomes and help stay clear of disasters. In this Practical Retina piece, I hope to share some of the key elements of evaluating and operating on children that I have learned from my many mentors, colleagues, and patients.

RULE OUT RETINOBLASTOMA

Retinoblastoma is the No. 1 diagnosis we all need to have in the back of our minds for any child with any presentation. Since ophthalmoscopy may not be well-tolerated in young children, having B-scan ultrasonography in our offices is helpful.[1] We all must have low thresholds for an examination under anesthesia (EUA) or referral to a retinoblastoma specialist if there is any degree of suspicion.[2]

EXAMINE BOTH EYES

Many pediatric pathologies are bilateral in nature, and the fellow eye can provide important diagnostic clues. For example, a child can present with dense vitreous hemorrhage in one eye, but the fellow eye may provide you with the diagnosis of familial exudative vitreoretinopathy (FEVR).[1,3-5] We rarely have opportunities for children to tolerate complete ophthalmoscopies, but they tolerate it very well under general anesthesia.

EXAMINING FAMILY MEMBERS

Along similar lines, examining parents and siblings can provide clues to a familial diagnosis. This can be the most cost-

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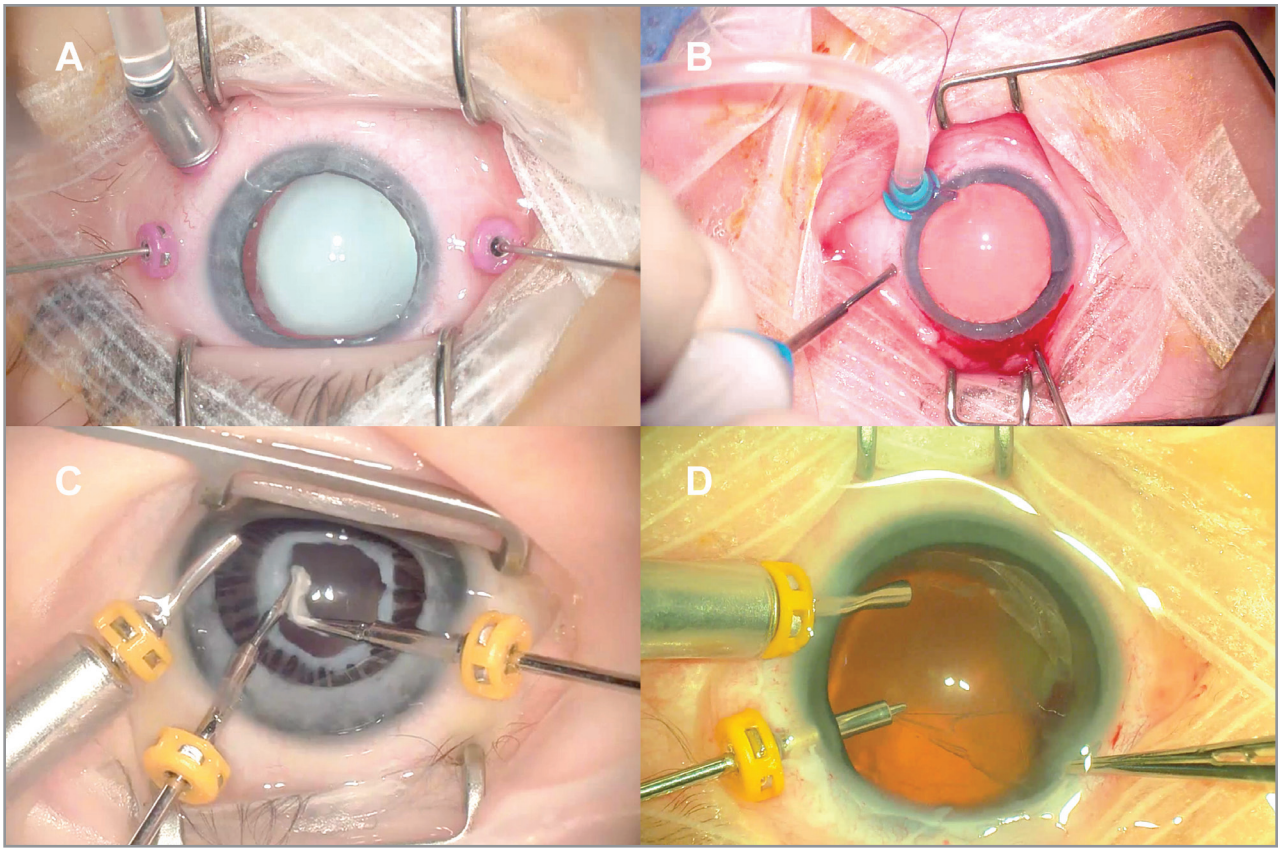

Figure. Various sclerotomy locations based on age and pathology. (A) 27-gauge vitrectomy for traumatic cataract in an 8-year-old child. All transconjunctival cannulas are placed 3 mm posterior to the limbus in a standard fashion. (B) 25-gauge short vitrectomy for retinopathy of prematurity stage 4A. There is an anterior temporal retinal detachment, so the infusion is sutured nasally, 1 mm posterior to the limbus. The other two sclerotomies are made away from the temporal detachment. All incisions were made after a peritomy, without cannulas. (C) 23-gauge vitrectomy for anterior persistent fetal vasculature (PFV). The infusion is placed in the anterior chamber via the trocar/cannula system. Limbal cannulas are then placed (can also be iris root incisions without cannulas after focal peritomies). PFV can be associated with fingers of retina drawn anteriorly, so unless the pars plana can be confirmed during the exam under anesthesia, staying anterior is recommended. (D) 23-gauge vitrectomy in Norrie disease in a 1-month-old. There is a nasal/inferonasal retinal detachment pulled up to and making contact with the crystalline lens. The temporal infusion is in the anterior chamber, away from the detachment. The superotemporal cannula is through the iris root. A limbal incision is made without a cannula, nasally since space is limited.

effective genetic testing. For suspicions of retinal vascular diseases, widefield fluorescein angiography (WFA) may uncover subtle findings and clues in family members.[1,5,6]

WIDEFIELD FLUORESCEIN ANGIOGRAPHY

Many pediatric retinal diagnoses, from sickle cell retinopathy and Coats' disease to FEVR and incontinentia pigmenti, harbor peripheral vascular changes that may not be fully appreciated without WFA.[3-11] This can be accomplished by non-contact wide-angle cameras in the outpatient setting, and contact wide-angle cameras during EUAs. Intravenous pediatric dosing can vary from 5 mg/kg to 8 mg/kg, but in our practice at Massachusetts Eye & Ear and Boston Children's, we use 7.0 mg/kg to 7.7 mg/kg.

ORAL FLUORESCEIN ANGIOGRAPHY

For children who cannot tolerate intravenous access but are old enough to cooperate for outpatient imaging, oral fluorescein administration is a great option.[12,13] We mix fluorescein with 4 to 6 ounces of fruit juice and encourage the children to quickly drink the mixture. Images are taken 15 to 20 minutes later and will provide late-frame angiograms. Dosing again is not standardized, but in our practice, we use 25 mg/kg for children under 20 kg, one bottle (500 mg) for those between 21 kg and 45 kg, and two bottles (1,000 mg) for children heavier than 45 kg.

OPTICAL COHERENCE TOMOGRAPHY

We are finding an increasing utility for optical coherence tomography for pediatric indications.[7,14-17]

The scans are well-tolerated and can supplement ophthalmoscopy, which may not be well-tolerated. Quick single-line scans can be used for children with limited cooperation.

PEDIATRIC RHEGMATOGENOUS RETINAL DETACHMENT = SCLERAL BUCKLE

The vitreous is densely formed and the posterior hyaloid is brutally adherent in children.[18] Complete hyaloidal separation during vitrectomy is nearly impossible in infants and very challenging in younger children. Therefore "just a quick vit" to repair rhegmatogenous retinal detachment (RRD) in children can be the start of a downhill battle with relentless proliferative vitreoretinopathy (PVR), because residual vitreous will inevitably be present. We therefore avoid entering young eyes if possible, and typically repair pediatric RRDs with scleral buckling.[19-22]

Even if children present with PVR, a primary buckle is often my modality of choice for treatment-naive eyes. PVR in pediatric eyes that have not been previously vitrectomized tends to be subretinal, and subretinal bands often do not exert substantial traction. Draining or not is up to the surgeon, but we should keep in mind that the majority of retinal detachments in children are chronic.[19]

Another pearl is that many "secondary" RRDs, such as in older children with histories of prematurity, will often have broad vitreous insertions, so wider buckles may be recommended if encircling. Finally, careful examination of the fellow eye is paramount due to high likelihood of pathology.[19]

BELIEVING IN THE RETINAL PIGMENT EPITHELIUM PUMP

Patience is key after scleral buckles in children. It is not unusual for the subretinal fluid to take months to resolve, and we must tame the urge to jump into recommending vitrectomy. The retinal pigment epithelium (RPE) is a powerful pump that we partner with and should believe in.

BITING THE BULLET: VITRECTOMY

Vitrectomy becomes necessary if scleral buckling fails. One of the keys to success, similar to adult surgery, is to induce a complete posterior vitreous detachment (PVD) and extend it past the break. The younger the child, the more difficult this will be, and we may also be fooled by vitreoschisis.

Triamcinolone staining and re-staining can be helpful to ensure that the hyaloid is separated. Additional instrumentation such as picks, forceps, needles, scrapers, and blades may often be necessary to peel the hyaloid, particularly in reoperations, where the hyaloid can become a dense membrane.

Meticulous peeling is key so that we can avoid retinectomies if possible, because extensive PVR often develops along retinectomy edges in young eyes. Although surgeons have individual preferences regarding 1,000 versus 5,000 centistokes (cs) silicone oil, I have come to appreciate 5,000 cs for pediatric cases.

TRACTION RETINAL DETACHMENTS: CANNOT MAKE BREAKS

Pediatric traction retinal detachment (TRD) is usually approached via vitrectomy, and again, requires patience and reliance on the RPE pump. From retinopathy of prematurity (ROP), FEVR, persistent fetal vasculature, to Norrie disease, the goal of surgery is to peel membranes and / or cut tractional vectors to release the traction, and watch the retina gradually reattach over time.[7,23]

In adults, we can resort to retinotomies, retinectomies, perfluorocarbon liquids, and other elegant techniques to flatten the retina by brute force. In general, we cannot do this in children because it is often not possible to completely flatten the retina, nor do we want to, because retinal defects will lead to explosive PVR.

In infant TRD surgery, a single iatrogenic break usually makes the eye inoperable. Therefore, the first goal of pediatric TRD surgery is not to make any iatrogenic breaks, and the second is to release the traction. As long as a break has not been made, the eye will live to fight another day for another round of membrane dissection.

STAGED SURGERY

Intentionally staged surgery is, therefore, a strong option for severe TRDs.[7] For example, in an eye with stage 5 ROP or FEVR with lens-cornea touch, the first surgery may be to reform the anterior chamber, lensectomy, capsulectomy, potentially iridectomy, to improve the corneal edema, and revive the eye. The second surgery may be to initiate the plaque dissection, carefully peeling to not create any central or pars plana breaks. The third surgery may be to complete the membrane dissection of the central stalk to open up the funnel and then wait for the subretinal fluid to resorb over time.

NO SUCH THING AS 'STANDARD PARS PLANA VITRECTOMY'

For older children and young adults, entering 3 mm posterior to the limbus reliably allows safe sclerotomy creation through the pars plana. However, in younger children, this distance may result in an iatrogenic break through the anterior retina. For neonates, we generally enter the eye 1 mm posterior to the limbus to perform pars plicata vitrec-

tomy. Two mm tends to be a safe distance for 2-year-olds, and 3 mm for 3-year-olds.[24,25]

However, we cannot rely on these estimates alone. Thorough EUAs need to be conducted to assure that there is no detached retina drawn up in the areas of intended sclerotomies. Some incisions may therefore have to be created in unconventional locations, and the surgeon may have to occasionally sit temporally (Figure). We also need to remember that the crystalline lens makes up a large proportion of the eye, so the trocars need to be angled appropriately.

When there is no space in the pars plicata/plana, such as in stage 5 detachments, or poor views in high-risk cases such as anterior segment dysgenesis or severe PFV, corneal limbal or iris root entry is an essential technique.[7,23] Infusion is accomplished by an anterior chamber infusion (Figure).

LESS IS MORE

"Perfect is the enemy of good" could not be truer for pediatric vitreoretinal surgery. The best surgeries achieve the anatomic goals and stop before iatrogenic complications arise. For example, in ROP surgery, the goal is to cut the tractional vectors and allow the retina to settle on its own — not a complete vitrectomy.[7] In PFV surgery, the goal is to cut the stalk, allowing the eye to snap open and the TRD to settle over time.[26] In endophthalmitis, the goal is to debulk without aggressive shaving that may result in iatrogenic breaks.[27]

BILATERAL SURGERY

For sick children with bilateral pathology (such as ROP, or children with syndromic detachments), operating on both eyes during the same anesthesia session may not only be vision-saving, but also a life-saving surgery. The risk of death with a second general anesthesia session is exponentially higher than the risk for bilateral endophthalmitis. We recently published recommendations for performing immediate sequential bilateral vitreoretinal surgery in children.[28,29]

INSTRUMENTATION AND VIEWING SYSTEMS

Smaller eyes force us to hold intraocular instruments closer together, and we are making the sclerotomies closer to the limbus as discussed above. This means that the instruments are more likely to hit the viewing lens. If instruments hit the lens, the lens moves, and if the lens moves, there is no view, and if there is no view, we cannot operate. Some viewing systems are more amenable to surgeries for small eyes, and there are certain instruments designed for pediatric surgery.

GODSPEED

There is no better feeling than telling parents that their child will have vision. To maximize the surgical outcomes, the differences between adult and pediatric eyes need to be kept in mind. When in doubt, we have a great community of pediatric retina surgeons that is always happy to offer its thoughts. I hope this introduction to some of the nuances in pediatric retina surgery was helpful, and I appreciate the editors for allowing such a piece to be published in this journal.

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