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Special considerations for pediatric vitreoretinal surgery

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Abstract:

In this review, the authors present special considerations a vitreoretinal surgeon should take into account before embarking on surgery in a pediatric eye. First, the anatomy of a pediatric eye is different from an adult and changes as the child grows. This is important especially in relation to the placement of transconjunctival ports. The structural characteristics of the sclera are also different, with lower scleral rigidity found in pediatric eyes. When considering vitrectomy, a posterior pars plicata lens-sparing technique should be considered. However, this may not be possible in complicated total detachments where anterior translimbal vitrectomy may be the method of choice. Scleral buckles are preferred for certain cases, and division of the encirclage is advocated in children below the age of 2 years, once the retina has stabilized. Enzymatic vitreolysis has been described as a preoperative adjunct to enhance complete detachment of the posterior hyaloid and reduce iatrogenic retinal breaks. However, its use in pediatric eyes has been limited, and larger studies are warranted. Finally, postoperative visual rehabilitation and treatment of amblyopia are key to maximizing functional outcomes in the pediatric patient. Co-management with a pediatric ophthalmologist and enlisting the co-operation of the parents are essential.

Keywords:

Endoscopic vitrectomy, pediatric, scleral buckling, vitrectomy, vitreoretinal surgery

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Introduction

Pediatric vitreoretinal surgery has its unique set of challenges compared to surgery in the adult eye. Before performing surgery, the vitreoretinal surgeon should be familiar with the anatomical and structural characteristics of the pediatric eye and the changes with age. Both vitrectomy and scleral buckling techniques may be used, but each has its advantages and disadvantages peculiar to the pediatric population. Endoscopic vitrectomy is another useful technique to have in the surgeon's armamentarium, especially in complicated eyes with a poor view of the posterior segment. Enzymatic vitreolysis has been used as an adjunct to aid in complete vitreous separation and reducing the risk of intra-operative retinal breaks. Finally, postoperative monitoring and visual rehabilitation are essential in maximizing functional outcomes.

Anatomical Considerations

Infants have smaller eyes with different surgical landmarks compared to adult eyes. These anatomical considerations must be taken into account when performing a vitrectomy in newborns or in infants. Surgical instrumentation for a vitrectomy can be approached via two-ways, either anterior (translimbal) or posterior (trans pars plicata/pars plana).

The pars plana, through which vitreoretinal surgical instruments are usually safely introduced in adults, is not fully formed until approximately the age of 8–9 months.[1] As a rule of thumb, the sclerotomies are placed at 4 mm from the limbus only in children aged 4 years and older. The distance of the sclerotomies from the

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limbus is dependent on age, and different practices may recommend different cutoffs [Table 1]. Our practice is illustrated in the table below and can be easily remembered as adding 1 mm to the distance from the limbus with each year of age after the 1st year of life. Our sclerotomy approach is conservative and is applicable for a variety of eye sizes. For eyes with unusual anatomy such as microphthalmos in persistent fetal vasculature (PFV), or eyes with severe anterior segment abnormalities, trans-scleral illumination is another useful method to determine the location of the pars plana for safe sclerotomy entry.[2]

As babies under 2 years of age have very thin sclera, trocar-cannula systems are utilized only at ≥2 years of age. Trocars are usually inserted perpendicular through the sclera as the oblique placement of the trocars is associated with an increased risk of accidental trocar dislocation and lens trauma. This is because the trocars have a shorter path through the pediatric sclera compared to that in the adult sclera.[3] Therefore, sutureless sclerotomies are not advisable in pediatric patients. In addition, an oblique entry together with the relatively thicker dimension of the children's lens increases the risk of accidental injury to the lens. Thus, insertion of intraocular instruments should always be parallel to the visual axis to spare the lens.

The posterior trans pars plicata lens-sparing technique can only be considered if the surgeon can safely introduce the instruments without causing an iatrogenic retinal break. Creating an iatrogenic retinal break in pediatric cases is best avoided as a massive proliferative response often follows.[4] This trans pars plicata approach may not be possible in patients with advanced retinal detachment (e.g., Stage 5 retinopathy of prematurity [ROP] with extensive anterior traction, or severe anterior and posterior persistent fetal vasculature) or with a poor view posterior to the lens (e.g., in PFV with diffuse retrolental plaques). In these cases, the risk of instruments entering the subretinal space outweigh the benefits of performing lens-sparing surgery, and an anterior trans-limbal approach with lensectomy and vitrectomy is usually preferable.

In patients with advanced retinal detachment, lens removal necessitates a total capsulectomy. Residual capsule serves as a scaffold for preretinal membrane proliferation, and secondary circumferential vitreo-retinal contraction can result in ciliary body detachment and hypotony. Once formed, this contractile anterior ring is usually difficult to visualize and dissect safely. In cases with lenticulo-retinal apposition, the capsular material can generally be dissected away from the retina without causing retinal breaks. Anterior dissection of preretinal proliferative membranes can then proceed.[5]

Table 1: Recommendations for the distance of the sclerotomy from the limbus

Authors		Age (months)				
		<3	>3-6	>6-12	>12-24	>24
Meier and Wiedemann[3]	Distance of sclerotomy from limbus (mm)	1.5	2.0	2.5	3.0	3.5
Gan NY, Lam WC	Distance of sclerotomy from limbus (mm)	1.0	1.0	1.0	2.0	3.0

As pediatric sclera has decreased scleral stiffness and increased elasticity, suturing of sclerotomies in children is also recommended to ensure a water-tight closure. Closure of the sclera and conjunctiva may be done with absorbable 8–0 vicryl sutures.

Surgical Techniques

In pediatric eyes, the surgical method of choice should take into account the anatomical differences compared to adult eyes, in addition to increased vitreoretinal adhesion and increased propensity for membrane proliferation.

Vitrectomy

Today, vitrectomies are mostly performed with wide-angle viewing systems. There are wide-field vitrectomy contact and noncontact lenses that are available and designed for use specifically in pediatric vitreoretinal surgery.

Babies and infants have lower systolic blood pressure compared to adults and surgeons must bear in mind that iatrogenic occlusion of the central retinal artery can be induced if the infusion pressure is too high or with prolonged scleral depression. Therefore, the optic nerve must be observed at all times to ensure patency of the central retinal artery.[6] In the anterior trans-limbal approach, the infusion is usually supplied via a self-retaining anterior chamber maintainer. A shelving corneal wound is made with a 20G MVR blade and the anterior chamber maintainer anchored in the corneal wound via grooves on its side [Figure 1]. The infusion pressure should be optimized to reduce the risk of corneal clouding and retinal incarceration during withdrawal of instruments. The surgeon should bear in mind that occasionally the anterior chamber may be very shallow and iris may be dragged while creating the sclerotomies, increasing the risk of iridodialysis, hemorrhage, and traction on the peripheral retina. To avoid this, the anterior chamber should be deepened with the infusion before creating the remaining sclerotomies.

Creation of a posterior vitreous detachment (PVD) is an important step in the successful management of a retinal detachment. This may be essential in managing rhegmatogenous retinal detachments in older children.

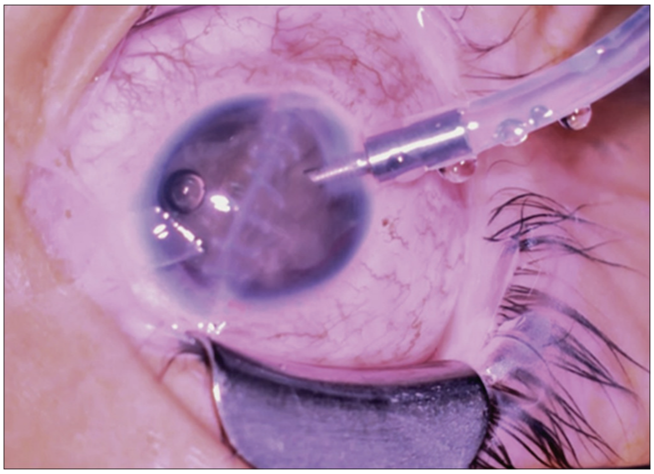

Figure 1: Trans-limbal self-retaining anterior chamber maintainer in a patient with previous blunt trauma, corneal laceration, total RD and 12 clock hours of anterior Grade C proliferative vitreoretinopathy

However, this is not recommended in eyes with ROP because of the very firmly adherent posterior vitreous. Forceful creation of a PVD when there is adherent vitreous is not only challenging but also carries a high risk of inducing retinal tears. With formed vitreous in pediatric eyes, some authors advocate 20G instrumentation as the larger port size and lumen allow for higher flow rates and a better ability to engage and hold the vitreous.[7] However, with current advances in vitrectomy instrumentation, 23G or 25G instruments are preferred as they can be used with equal efficiency with the advantage of smaller sclerotomies.

In eyes with proliferative vitreoretinopathy, it is preferable to perform segmentation instead of delamination when removing preretinal membranes due to the firm vitreoretinal attachments in children.

When using heavy liquids intra-operatively, perfluorodecalin ($C_{10}F_{18}$) is the preferred tool of choice in pediatric retinal detachments as it has the highest specific gravity of 1.93g/cm^3 and is ideal for flattening the retina which is usually thicker in babies and children. This is especially so in eyes with extensive proliferative vitreoretinopathy. Posterior drainage retinotomies are best avoided as extensive fibrous proliferation can occur postoperatively, leading to retinal redetachment. If a retinotomy is unavoidable, it is usually recommended to create one as close to the ora serrata as possible.

For vitreous tamponade, silicone oil should be avoided in cases of advanced retinal detachment such as stage 5 ROP or advanced posterior PFV with retinal detachment. Incomplete dissection of all the tractional components increases the risk of subretinal migration of silicone oil. Similarly, in the case of retinal detachment associated with coloboma, there is also an increased risk of subretinal migration of silicone oil through the colobomatous defect. Therefore, use of silicone oil should be avoided.

Scleral buckling

A scleral buckle may be used as a primary cerclage or as an encircling band in combination with vitrectomy. When combined with a vitrectomy, the element of our choice for an encircling band in infants is a number 40 (2 mm) or 240 (2.5 mm) silicone band, and the band is usually placed just anterior to the equator. If additional height is needed, number 20 segmental element can be added.

In ROP-related detachments, the band is placed to support the ridge, akin to supporting a retinal tear, with the encircling element supporting the ridge along the anterior portion of the element. Sutures are imbricated to provide height.[8] Scleral belt loops are avoided in children as the sclera is thinner than in adults. In pediatric rhegmatogenous retinal detachments, we usually recommend a broad, low buckle with precise localization of the retinal break on the buckle. However, due to the more vigorous reproliferation in children, a slightly higher indent may be preferred by some, with care not to cause significant distortion of the globe and further opening of the retinal break.[9]

Postoperative complications of scleral buckling in children include limitation of eye growth, development of amblyopia and loss of vision from cycloplegic eyedrops. Some authors recommend dividing the encircling band approximately 3 months after the operation in children <2 years of age or in those whose eye growth is retarded. The band is preferably divided rather than removed as continued support may be provided by the encapsulated explant. In children with good visual potential in both eyes, atropine 1% eye drops should be avoided, instead a short-acting cycloplegic such as cyclopentolate 0.5%–1% may be prescribed to reduce the risk of developing amblyopia.[9] Refractive errors are also treated aggressively in the postoperative period to maximize visual outcomes.

Endoscopic vitrectomy

The conventional surgeon's perspective is to look down an operating microscope through the patient's anterior segment and into the vitreous cavity and retina using contact or noncontact viewing systems. The endoscope, however, allows direct visualization of the vitreous cavity by capturing images through its internal tip. This results in a side-on intra-operative view that is up to 90° off the conventional viewing axis of microscope-based systems.

Endoscopic vitrectomy is thus complementary to conventional top-down microscope-based viewing systems as it can bypass anterior segment opacities and provide undistorted and unobstructed views of the space between the vitreous base and the posterior iris.[10] The surgeon performs heads-up surgery and looks at a display screen [Figure 2] to see the posterior segment and anterior structures including the vitreous base, pars plana, pars plicata, ciliary body, lens, posterior iris surface and the anterior hyaloid face. The on-screen image is, however, 2-dimensional rather than 3-dimensional. Thus, the surgeon needs to compensate by using nonstereoscopic clues such as shadows to judge distance.[11]

Conventional top-down microscope viewing systems dissociate the surgeon's visual axis and source of illumination. Light is transmitted through the patient's clear vitreous, resulting in the vitreous appearing mostly transparent. In contrast, the illumination and surgeon's view of the reflected light that bounces off the retina/vitreous is co-axial in endoscopic vitrectomy. Vitreous and membranes appear more opaque because of the co-axial viewing and illumination, which is a bonus in pediatric detachments where removing adherent vitreous and membranes from the retinal surface is already challenging.[10]

Endoscopy is particularly useful in advanced pediatric tractional retinal detachments in retinopathy of prematurity or familial exudative vitreoretinopathy, in which there is often a significant anteroposterior tractional component with the retinal detachment extending towards the anterior hyaloid and lens. The endoscope enables better visualization of the side profile of the retinal detachment, versus looking at the top edge of the retinal detachment with a conventional top-down view, thereby facilitating more direct and potentially more complete tissue dissection.[12]

In ROP and PFV, extensive retrolental plaques may occur, blocking direct visualization of the underlying retina. Avoiding iatrogenic retinal breaks is critical in these cases. In PFV, the retina is also often drawn up along the hyaloidal stalk. Differentiating the limit of the retina along the stalk to allow safe transection is challenging with a bird's eye view in conventional microscope-based systems. With endoscopy, direct visualization enables the entire side profile of the hyaloidal stalk and its relationship to the retina to be seen with greater ease [Figure 3].[13]

Enzymatic vitreolysis

In pediatric patients, the vitreous attachments to the retina are very strong, due to the absence of PVD. The strong vitreoretinal adhesions make vitrectomy and removing all the vitreous gel in pediatric cases challenging.

Pharmacologic vitreolysis has been attempted in pediatric eyes as a neo-adjuvant to vitrectomy surgery. The hypothesis is that enzymatic vitreolysis can weaken the vitreoretinal junction, resulting in a more complete dissection of the hyaloid from the retina with less trauma to the retina and less iatrogenic breaks. Plasmin-assisted vitrectomy has been described in patients with stage 5 ROP reducing iatrogenic breaks and improving anatomical outcomes in reoperations;[14] reducing the need for inner-wall retinectomies in congenital X-linked retinoschisis;[15] and improving the rates of complete PVD with successful closure of pediatric traumatic macular holes.[16,17] In these studies, autologous or maternal plasmin enzyme was used. Blood was drawn a few days before the surgery, centrifuged and purified followed by eluting of plasminogen and finally, conversion to plasmin with streptokinase.

Ocriplasmin is a recombinant protease enzyme that can lyse fibronectin and laminin, components of

Figure 2: An endoscopy and laser unit used at the hospital for sick children, Toronto, housing a Xenon light source, diode laser, video camera and display screen, with the endoscope probe attached

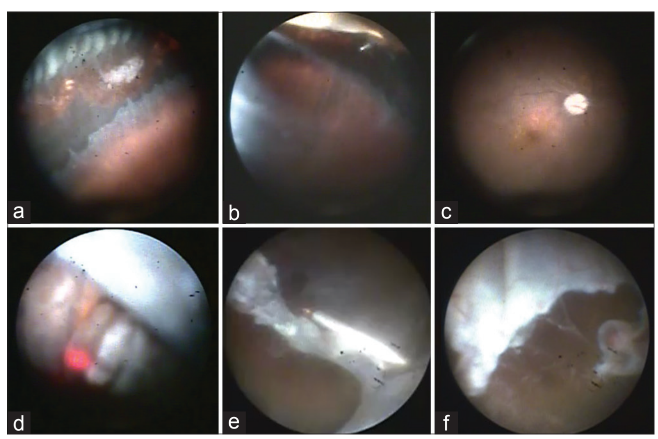

Figure 3: (a) Endoscopic view of ciliary body, pars plana and ora serrata, (b) wide angle view of a trans pars plana port and anterior retina, (c) endoscopic view of posterior pole, (d) endoscopic cyclophotocoagulation with laser beam aiming at a ciliary process with the adjacent ciliary process appearing white post ablation, (e) intraocular diathermy of vessels of anterior contracted retina with pre-equatorial band of proliferative vitreoretinopathy, (f) relaxing retinectomy

the vitreomacular interface. It is a stable, truncated form of plasmin and has several advantages over plasmin including its stability and smaller size, hence increased ability to penetrate tissues.[18] Its use as an intravitreous injection (125 μg) has been studied in two phase III randomized controlled trials for symptomatic vitreomacular adhesion (VMA) and vitreomacular traction (VMT)[19] and approved for use in more than 50 countries. The MIVI-TRUST study reported VMT release in 26.5% and closure of small full-thickness macular holes in 40.6%.[19] However, ellipsoid zone changes have been seen in some treated eyes, reportedly more common in those with a resolution of VMT but mainly transient.[20]

Drenser *et al*.[21] reported a Phase 2 single-center, prospective, randomized, placebo-controlled, double-masked study with 22 pediatric patients randomized 2:1 to receive 175 μg of ocriplasmin injection or placebo 30–60 min before vitrectomy. They found that ocriplasmin was generally well tolerated. However, the primary efficacy endpoint of total macular PVD either before vitrectomy or after suction was seen in 50% (8 of 16) of ocriplasmin-treated eyes and 62.5% (5 of 8) of placebo-treated eyes. The study sample size was small in this study. Therefore, the true efficacy of ocriplasmin as an adjunct cannot be adequately assessed.

As shown in the above small studies, enzymatic vitreolysis is a useful tool in complicated pediatric vitrectomies. However, large randomized controlled trials are still needed before pre-vitrectomy injection is an approved indication for ocriplasmin use.

Postoperative Care

In pediatric patients, postoperative care is an integral part of the treatment journey. Not only is compliance with the posturing and medication regimen important, visual rehabilitation also plays a large role in increasing functional success. In patients who have predisposing conditions that have caused deprivational amblyopia, visual potential may be guarded. Postoperatively, all refractive errors must be corrected and amblyopia treated aggressively so that maximal visual potential may be achieved. In children with baseline good vision and who undergo vitrectomy with long-acting gas tamponade or silicone oil, deprivational amblyopia may set in while the gas or silicone oil is in the eye. Children may be refracted to give the best correct visual acuity with silicone oil. Once the gas resolves or silicone oil is removed, any amblyopia present should be treated together with pediatric ophthalmologists.

In children with poor visual prognosis despite undergoing all treatment and surgery, ophthalmologists should be aware of the community resources that are available for the parents to ease their child's integration into society. Useful resources include parent support groups, schools for children with special needs, national societies for the visually handicapped for access to visual aids and learning of life-skills, for example, reading and writing braille.

For children with one good eye, parents must be taught the importance of avoiding contact sports, and to use polycarbonate lenses and sports goggles to avoid accidental trauma to the good eye.

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Conflicts of interest
The authors declare no conflicts of interest in writing this paper.

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