

7.1 Introduction

Pediatric cataract surgery techniques have significantly improved over course of time with development of new microsurgical techniques. Despite this, pediatric eyes are more predisposed to development of media opacity following cataract surgery as compared to adult eyes. Visual axis opacification (VAO) includes any significant media opacity in the visual axis which hinders the transmission of light and thus, affect image formation. This defeats the purpose of early pediatric cataract surgery which is primarily to clear the media and prevent stimulus deprivation amblyopia.

There are various causes of media opacity, including early onset and late onset causes (Table 7.1). The most important and commonest cause is posterior capsular opacification (PCO) [1, 2]. It is the commonest complication requiring second surgical intervention following pedi-

Table 7.1 Causes of visual axis opacification [4]

Early media opacification	Late media opacification
Corneal edema	Posterior capsular opacification (Fig. 7.2)
HypHEMA	Hyaloid face opacification
Inflammatory or fibrinous membrane (Fig. 7.1)	IOL glistening (Fig. 7.3)
Cortical remnants	IOL debris or pigments
Vitreous hemorrhage	Late vitreous exudates
Vitreous exudates	



Fig. 7.1 Postoperative serial picture of 12-year-old boy with post traumatic cataract with fibrinous membrane in the anterior chamber on first postoperative day

Supplementary Information The online version of this chapter (https://doi.org/10.1007/978-981-16-0212-2_7) contains supplementary material, which is available to authorized users.

C. Dhull (✉)
Eye Q Hospital, Rohtak, Haryana, India

S. K. Khokhar
Professor Ophthalmology, Dr. RP Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

atric cataract [3]. Intraoperative and postoperative complications are discussed in the respective chapter. In this chapter, we will discuss VAO related to PCO and hyaloid face opacification. PCO in children is generally thicker than adults and most often would require surgical interven-

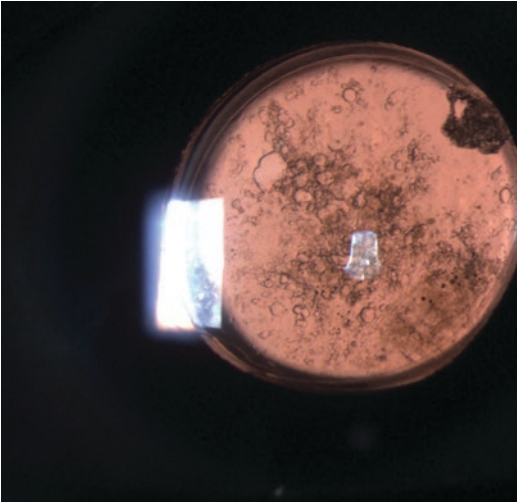


Fig. 7.2 Posterior capsular opacification seen 4 months post-surgery in 8-year-old child (Posterior capsule intact)

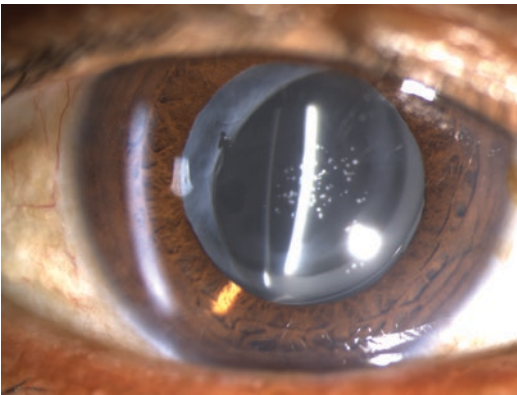


Fig. 7.3 IOL glistening seen with anterior capsular fibrosis 4 years after surgery

tion for removal. Understanding pathogenesis of PCO will help in understanding predisposing factors and prevention techniques for PCO formation. We would also discuss surgical approaches for successful PCO management.

7.2 Pathogenesis

After any extracapsular cataract surgery, lens epithelial cells (LECs) on the anterior capsule are invariably left behind. These LEC's are responsible for PCO formation [5]. The germinal equa-

torial E-type LECs show active mitosis and produce lens fibers throughout life. Some of the anterior or equatorial LECs left after surgery show the process of proliferation, migration and differentiation [6] as a healing response to surgery. There is epithelial to mesenchymal transition (EMT) along with collagen deposition and lens fiber [7–9].

There are two types of PCO: fibrotic and proliferative type. There may be contraction of fibroblasts deposited which may cause wrinkling and folds in the capsule. This leads to fibrotic variety of PCO [8, 9]. The lens fiber proliferation and regeneration from E-type LECs is responsible for proliferative type of PCO [6, 9]. Just like the posterior capsule, anterior hyaloid face can also act as the scaffold for this process of migration, proliferation and EMT.

Cytokines and growth factors such as transforming growth factor β (TGF- β), fibroblast growth factor 2 (FGF-2), Interleukins 1 and 6 also have a significant role in cellular response in PCO [10–12]. The inflammatory response in children following cataract surgery is significantly greater than adults. This explains thicker and more frequent PCO is younger children. Additionally, in conditions associated with more severe inflammatory response such as uveitis, trauma, etc., PCO may be more frequent.

7.3 Predisposing Factor

The incidence of PCO is variable and can be as high as 95% [13] without adequate protective measures. The incidence depends various predisposing factors: age at the time of surgery, etiology of cataract, surgical technique, IOL implantation and position, ocular comorbidities such as persistent fetal vasculature (PFV), systemic comorbidities such as juvenile idiopathic arthritis.

7.3.1 Age

Age is one of the most important factors in determining the predisposition to PCO. Younger age group is significantly more predisposed to PCO

formation [14–19]. Peterseim et al. reported higher incidence of PCO in children <2 months of age when compared with older children [19].

Hosal and Biglan found the risk of secondary cataract formation to be 4.76-fold (odd's ratio) when operated at <1 year of age compared to children >1 year of age.

7.3.2 Etiology of Cataract

Traumatic cataracts may have higher risk of VAO formation compared to developmental or congenital cataract of non-infective origin [20]. This may be explained by higher inflammation in traumatic cases before and after surgery.

Systemic association such as JIA may significantly increase incidence of PCO. Incidence of PCO has been reported in as high as 70–80% eyes despite posterior capsulotomy with or without anterior vitrectomy [21, 22]. PCO formed may be very thick and vascularized (Fig. 7.4). In other cause of uveitis, there is increased incidence of PCO as well. Infective cataracts such as TORCH infections (Toxoplasma, others, rubella, cytomegalovirus, and herpes simplex virus) may also show more inflammation and hence greater risk of PCO.

Ocular association such as PFV has been associated with increased incidence of PCO [1, 2]. In addition to PCO, they may develop hyphema or more commonly vitreous hemorrhage which may be the cause of media opacity.

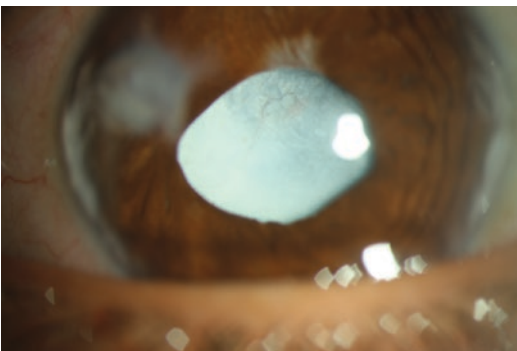


Fig. 7.4 8-year-old JIA patient with aphakia with vascularized visual axis opacification 1 year after surgery. Band shaped keratopathy is also seen

7.3.3 Surgical Technique

The importance of posterior capsulorhexis (PCCC) or capsulotomy with or without anterior vitrectomy (AV) in pediatric cataract surgery has been clearly established in the last 30 years. Incidence of PCO depend on the presence or absence of PCCC as well as AV. Posterior capsule or anterior hyaloid face act as a scaffold for PCO to form.

Hosal and Biglan clearly established the importance of PCCC and AV in their study of 152 patients with a mean follow of 6 years. PCO occurred 78.6% of eyes without PCCC, 42.9% with PCCC, and 22.5% with combined PCCC and AV [16].

7.3.4 Size of Capsulorhexis

A 5 mm opening in anterior capsulorhexis is considered as ideal. Smaller capsulorhexis may be associated with anterior capsular phimosis.

7.3.5 Aphakia or Pseudophakia

It has remained somewhat controversial over time whether the presence of IOL increases VAO formation. In our experience, we have not observed drastic difference between the two groups. However, a review of five studies including 597 eyes was published by J. Chen et al. where they found primary IOL implantation increased risk of VAO compared to contact lens fitted group [23]. There may be associated confounders such as surgical technique, surgeon's experience, and use of steroids which have been identified in the study.

7.3.6 IOL Position and Type

Incidence of VAO in the case of IOL in bag and IOL in sulcus in pediatric traumatic cataract was compared [24]. Complications including VAO and pupillary capture were observed more often in sulcus group. In other studies, no difference in

PCO was found based on IOL position [25]. IOL in sulcus with optic capture has been used to reduce PCO formation [26]. Besides these, poorly centered anterior capsulorhexis in the bag may also accelerate PCO formation if it does not cover all the edges of IOL.

Studies comparing the acyclic material prove that hydrophilic acrylic material accelerates PCO formation more than hydrophobic material [27]. Heparin surface coating on polymethyl methacrylate (PMMA) i.e., heparin surface modified IOLs (HSM-IOLs) also cause less PCO formation and are used for uveitic cataracts [22]. (Hydrophilic acrylic IOL > PMMA IOL > hydrophobic IOL). A capsular bend with sharp and square optic edge induce contact inhibition to migrating LECs, thus reducing PCO formation [28].

7.4 Clinical Presentation

Detection of VAO is challenging in pediatric patients. Symptoms may be nonspecific and parents need to be counselled to look for anything out of the ordinary. In bilateral cataracts, if only one eye is affected, parents may not notice anything. Hence, the importance of regular follow up and EUA needs to be explained beforehand.

Sometimes, presence of whitish opacity or squint is observed by the caregivers. In unilateral cataracts, where patching is being followed with good compliance, the child may now be noted to resist occlusion of the good eye more than usual.

Most often, it is picked up on routine follow-up. Clinician may find poor visual acuity on Cardiff's tests and a poor glow (Bruckner reflex) through direct ophthalmoscope. A child presenting to clinic with a poor glow should be assessed through the corrective spectacles as high refractive error may show dull glow specially in aphakia.

VAO may be predominantly posterior to lens, anterior to lens or combination of both. It may be visual insignificant when central visual axis is clear. PCO is classified as:

- Fibrotic type: In fibrotic variety, capsule is thickened and fibrosed with folds and wrinkling in the capsule (Fig. 7.5).
- Proliferative type: Include both Elschnig pearls and Sommering ring. Elschnig pearls are round, clear pearl like clusters of residual LECs which are clearly demarcated on retroillumination (Fig. 7.6). Sommering ring forms in the periphery generally between anterior capsular edge and posterior capsule, it may

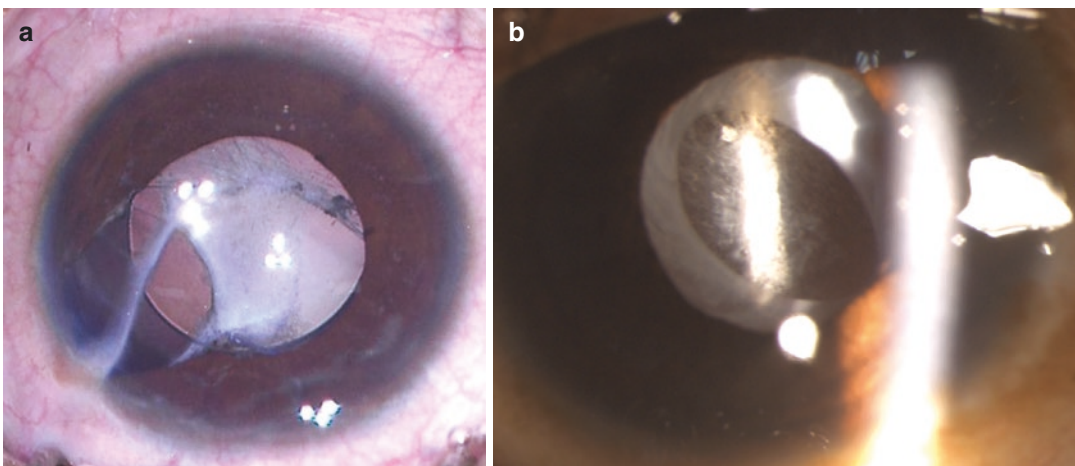


Fig. 7.5 Fibrotic type of VAO. (a) Anterior fibrotic VAO seen 4 months after surgery for post traumatic cataract. (b) Posterior fibrotic VAO seen 1 year after surgery for intermediate uveitis

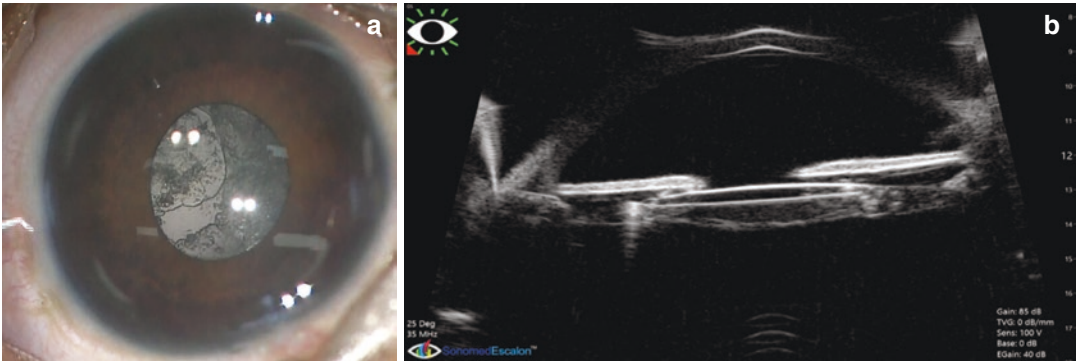


Fig. 7.6 Proliferative VAO with Elschnig pearl formation. (a) Clinical picture. (b) UBM of the same showing VAO posterior to IOL and intact posterior capsule

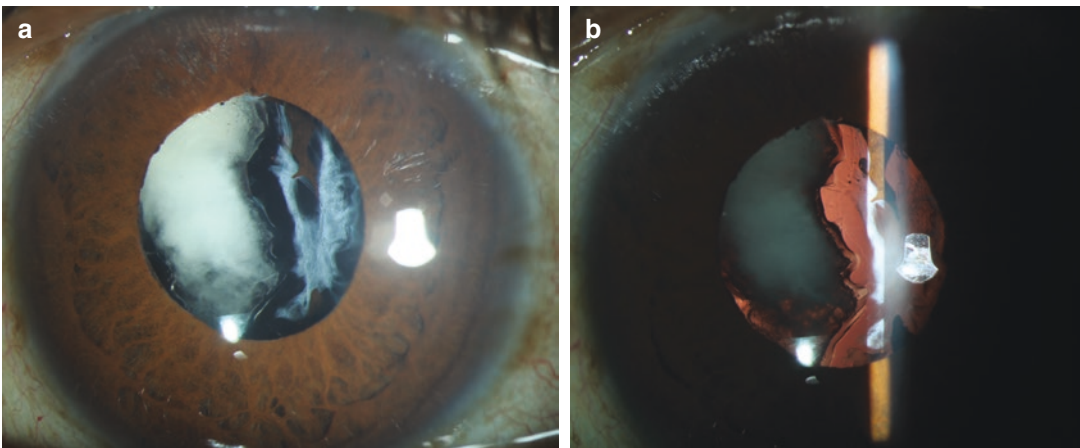


Fig. 7.7 Proliferative VAO with Soemmering ring seen in aphakic patient 20 months after surgery

proliferate further, contract and involve the visual axis (Fig. 7.7).

7.5 Investigations

Ultrasonography should be performed preoperatively to rule out posterior segment pathology such as retinal detachment. In uveitis cases, USG should be performed to evaluate activity in the posterior segment as well.

Ultrasound biomicroscopy (UBM) can be performed in cases of severe VAO or non-dilating pupil. It provides detailed information related to the location and severity of VAO. Anterior, poste-

rior and mixed variety can be evaluated and surgery can be planned preoperatively (Fig. 7.8), it can also provide information regarding IOL position and associated pathology such as persistent fetal vasculature.

7.6 Prevention

7.6.1 Medical Preventive Measures

Use of topical steroids and cycloplegics post-surgery helps in control of inflammation and hence VAO formation. Slow taper of steroids is recommended.

In etiologies associated with excess inflammation such as uveitis, prophylactic use of systemic steroids or immunosuppressants is recommended.

7.6.2 Surgical Preventive Measures

ACCC: 5 mm circular central ACCC covering IOL 360° should be made as there are greater chances of PCO formation in eccentric IOLs (Fig. 7.9).

Hydrodissection and cortical clean up: We recommend performing good cortical clean up especially the equatorial fibers and cells. Residual cortical matter can promote VAO formation.

PCCC and AV: Studies have well established the role of primary PCCC with anterior vitrectomy to effectively delay the secondary cataract formation in infants and children [29].

Anterior vitrectomy breaks the scaffold for the LECs that are actively proliferating and prevents deposition of metaplastic cells, thus preventing PCO formation [30]. Posterior capsulotomy is must for all patients <6 years of age [18]. Vitrectomy can be deferred after 5 years of age [30].

The authors routinely perform PCCC with/without AV for all children operated for cataract at the age of less than 6 years and in special cases of older children with mental retardation, nystagmus or who are unable to follow-up frequently.

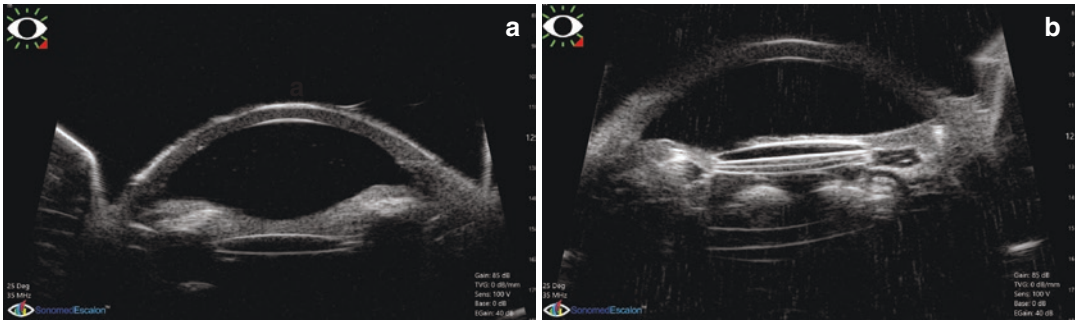


Fig. 7.8 UBM of dense VAO patients. (a) Anterior VAO seen, no VAO behind the lens. (b) Combined anterior and posterior VAO

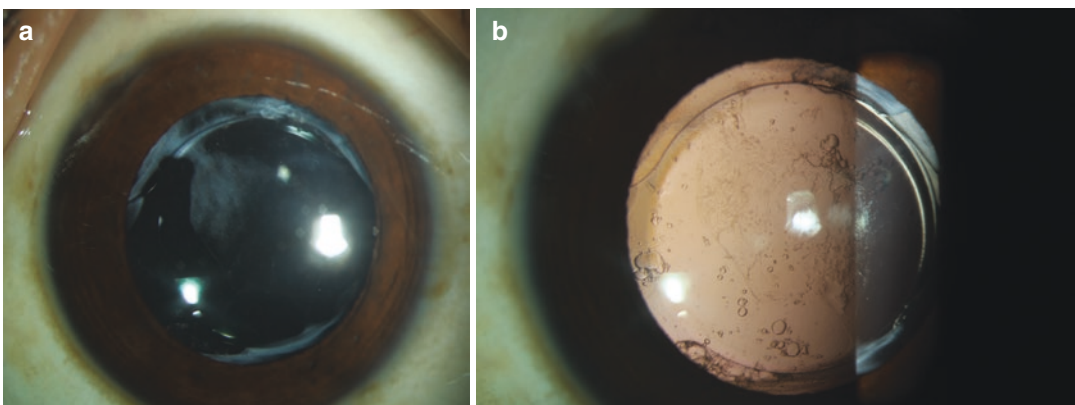


Fig. 7.9 PCO formation in case of larger capsulorhexis not covering IOL completely. (a) Diffuse illumination picture. (b) Retroillumination picture. Note both fibrotic and pearl like PCO formation

IOL: We recommend in the bag IOL implantation with square edge haptic IOL or in the sulcus IOL with optic capture with both anterior and posterior capsule depending on suitability, to reduce VAO.

7.7 Management

PCO is amblyogenic in the critical period of visual development and causes stimulus deprivation in a child, thereby interfering with the goal of a successful cataract surgery. If PCO is visually significant, it requires removal via laser or surgery.

7.7.1 Neodymium Yttrium Aluminum Garnet (Nd:YAG) Capsulotomy

Nd:YAG laser is a relatively noninvasive non-surgical procedure to remove PCO in a day care setting. Since the procedure requires significant cooperation from the patient, it is used in older children with sufficient understanding.

The procedure is performed under topical anesthesia. An Abraham's Nd YAG laser lens is used. Energy requirement is generally low from 1–2 mJ/pulse. It can be titrated based on the thickness of PCO. Laser shots are directed at taut capsule to create a 4 to 5 mm circular opening [31]. Cruciate opening can also be used but generally not preferred due to risk of extension. A posterior offset may be required to avoid hitting the IOL. It is not preferred in cases of PMMA IOL.

However, with the use of Nd:YAG capsulotomy, few limitations remain: (1) the anterior hyaloid still remains intact, which may again act as scaffold for recurrent PCO formation; and (2) a very thick fibrous PCO may not be easily cut with laser. (3) Risks of treatment with Nd:YAG laser include: cystoid macular edema (CME) [32, 33] and low risk of retinal detachment [33]. IOL

pitting can happen in uncooperative children especially with thick PCO.

7.7.2 Surgical Management

Most children especially under 6 years of age require surgical intervention for treatment of VAO. Surgical approach depends on the location of PCO, location of IOL and surgeon's experience. VAO must be managed promptly and child has to be taken up for resurgery under general anesthesia. An informed consent must be obtained from caretakers/parents in these cases, with clinician explaining them the risk of recurrent PCO and associated complications.

7.7.2.1 Limbal Route

Most anterior segment surgeons prefer membranectomy from anterior route or limbal route (Fig. 7.10). Two paracentesis incisions are made at the limbus nearly 180° apart. Anterior chamber is filled with viscoelastic and vitrectomy cutter and irrigator can be used for making an adequate opening in the PCO in aphakic eyes. If PCO is very thick intravitreal scissors and forceps may be required to cut and remove the membranes especially in cases such as uveitis, PFV, etc. Limited localized anterior vitrectomy is performed. If IOL is placed in the sulcus, a slight nudge can help to place the probe underneath it for membranectomy. If IOL is in the bag or fibrosed strongly with the capsule, excessive manipulation may weaken the zonules. We prefer a pars plana approach in such cases.

7.7.2.2 Pars Plana Route (Fig. 7.11)

A pars plana port is constructed first as described in chapter on surgical management. A limbal paracentesis is created for irrigation. Limited anterior vitrectomy is performed followed by membranectomy using vitrectomy cutter in Cut IA mode. Similar to limbal approach, in case of thick VAO, microincision (23 gauze) intravitreal

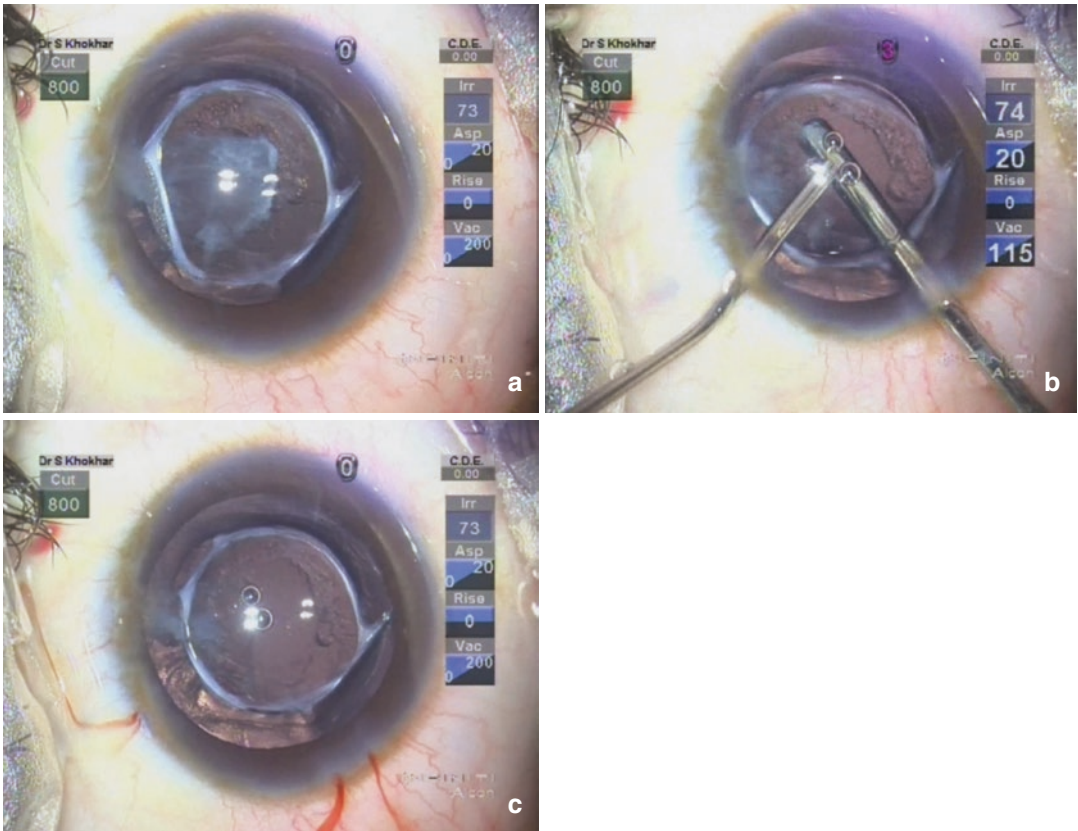


Fig. 7.10 Surgical management of PCO via anterior route. (a) Thick PCO behind IOL. (b) Membranectomy and anterior vitrectomy being performed by lifting the IOL slightly. (c) Surgery complete with clear 5 mm central zone

scissors and forceps can be used. There is minimal displacement of IOL and the irrigation probe can be used to support IOL throughout the procedure. Combined approach in cases of VAO both anterior and posterior to IOL can be used to achieve optimal outcome.

VAO associated with complicated surgery or mal-positioned IOL may require additional procedure to membranectomy and anterior vitrectomy.

Pupilloplasty, IOL exchange, repositioning or explant may be required. This may be a surgical challenge due to extensive fibrosis and/or vascularization. Complications associated with membranectomy are similar to any intraocular procedure such as pediatric cataract surgery. Refractive correction after surgery is a must for early visual rehabilitation. Amblyopia therapy should be initiated as soon as media is rendered clear.

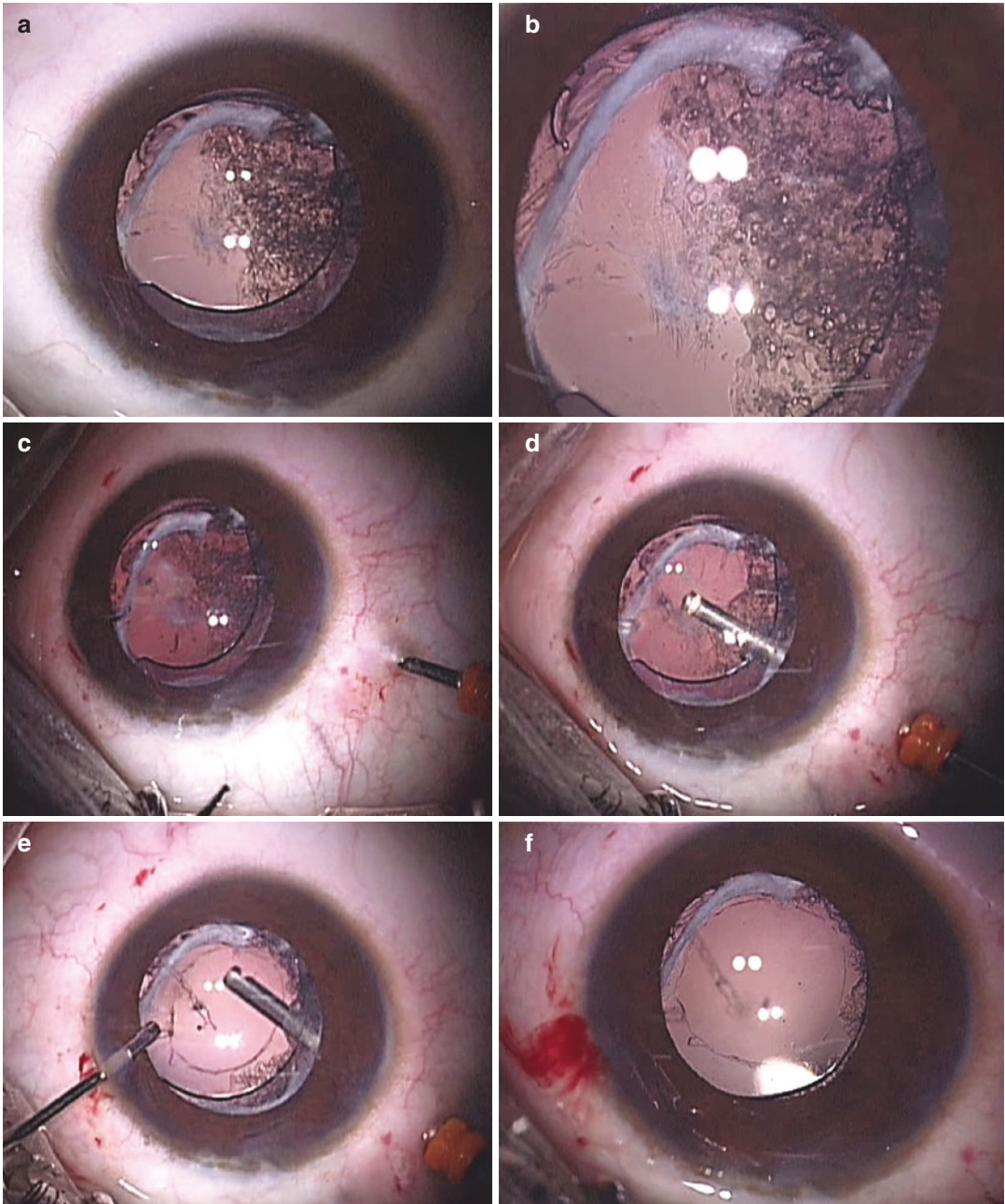


Fig. 7.11 Surgical management of PCO via pars plana route. (a) Clinical picture showing proliferative type of PCO. (b) Magnified view showing primarily posterior variety of VAO. (c) 23 g port being made. (d)

Membranectomy is performed in cut IA mode. (e) Limited anterior vitrectomy done. (f) Post-surgery central 5 mm opening

7.8 Conclusion

VAO remains the commonest postoperative complication that requires surgical intervention. Since incidence of VAO is extremely high in younger children without posterior capsulotomy, primary surgery should include meticulous technique with PCCC and AV. Prevention of PCO formation can help in reducing the rate of secondary surgical intervention in children. However, if PCO form, early remedy by the most suitable method is warranted. Refractive correction and amblyopia therapy should be continued after treatment of PCO.

References

- Apple DJ, Solomon KD, Tetz MR, et al. Posterior capsule opacification. *Surv Ophthalmol*. 1992;37:73–116.
- Pandey SK, Apple DJ, Werner L, et al. Posterior capsule opacification: a review of the aetiopathogenesis, experimental and clinical studies and factors for prevention. *Indian J Ophthalmol*. 2004;52:99–112.
- Sachdeva V, Katukuri S, Ali MH, Kekunnaya R. Second intraocular surgery after primary pediatric cataract surgery: indications and outcomes during long-term follow-up at a tertiary eye care center. *Eye*. 2016;30:1260–5.
- Khokhar SK, Dhull C, editors. Atlas of pediatric cataract. Singapore: Springer Nature Singapore Pte Ltd; 2019. p. 145. https://doi.org/10.1007/978-981-13-6939-1_15.
- Lambert SR, Drack AV. Infantile cataract. *Surv Ophthalmol*. 1996;40:427–8.
- Marcantonio JM, Vrensen GF. Cell biology of posterior capsular opacification. *Eye (Lond)*. 1999;13(Pt 3b):484–8.
- McDonnell PJ, Zarbin MA, Green WR. Posterior capsule opacification in pseudophakic eyes. *Ophthalmology*. 1983;90(12):1548–53.
- Cobo LM, Ohsawa E, Chandler D, et al. Pathogenesis of capsular opacification after extracapsular cataract extraction: an animal model. *Ophthalmology*. 1984;91(7):857–63.
- Wormstone IM. Posterior capsule opacification: a cell biological perspective. *Exp Eye Res*. 2002;74(3):337–47.
- Meacock WR, Spalton DJ, Stanford MR. Role of cytokines in the pathogenesis of posterior capsule opacification. *Br J Ophthalmol*. 2000;84(3):332–6.
- Nishi O. Posterior capsule opacification, part 1: experimental investigations. *J Cataract Refract Surg*. 1999;25(1):106–17.
- Wallentin N, Wickström K, Lundberg C. Effect of cataract surgery on aqueous TGF- β and lens epithelial cell proliferation. *Invest Ophthalmol Vis Sci*. 1998;39(8):1410–8.
- Knight-Nanan D, O’Keefe M, Howell R. Outcome and complications of intraocular lenses in children with cataract. *J Cataract Refract Surg*. 1996;22:730–6.
- Morgan KS, Karcioğlu ZA. Secondary cataracts in infants after lensectomies. *J Pediatr Ophthalmol Strabismus*. 1987;24:45–8.
- Alexandrakis G, Peterseim MM, Wilson ME. Clinical outcomes of pars plana capsulotomy with anterior vitrectomy in pediatric cataract surgery. *J AAPOS*. 2002;6:163–7.
- Hosal BM, Biglan AB. Risk factors for secondary membrane formation after removal. *J Cataract Refract Surg*. 2002;28:302–9.
- Lambert SR, Buckley EG, Plager DA, et al. Unilateral intraocular lens implantation during the first six months of life. *J AAPOS*. 1999;3:344–9.
- Trivedi RH, Wilson E, Vasavada AR, et al. Visual axis opacification after cataract surgery and hydrophobic acrylic intraocular lens implantation in the first year of life. *J Cataract Refract Surg*. 2011;37:83–7.
- Peterseim MW, Wilson MW. Bilateral intraocular lens implantation in the pediatric population. *Ophthalmology*. 2000;107:1261–6.
- Gimbel HV, Ferensowicz M, Raanan M, et al. Implantation in children. *J Pediatr Ophthalmol Strabismus*. 1993;30:69–79.
- BenEzra B, Cohen E. Cataract surgery in children with chronic uveitis. *Ophthalmology*. 2000;107:1255–60.
- Lundvall A, Zetterstrom C. Cataract extraction and intraocular lens implantation in children with uveitis. *Br J Ophthalmol*. 2000;84:791–3.
- Chen J, Chen Y, Zhong Y, et al. Comparison of visual acuity and complications between primary IOL implantation and aphakia in patients with congenital cataract younger than 2 years: a meta-analysis. *Cataract Refract Surg*. 2020;46:465–73.
- Pandey SK, Ram J, Werner L, et al. Visual results and postoperative complications of capsular bag and ciliary sulcus fixation of posterior chamber intraocular lenses in children with traumatic cataracts. *J Cataract Refract Surg*. 1999;25:1576–84.
- Jensen AA, Basti S, Greenwald MJ, et al. When may the posterior capsule be preserved in pediatric intraocular. *Ophthalmology*. 2002;109:324–8.
- Raina UK, Gupta V, Arora R, Mehta DK. Posterior continuous curvilinear capsulorhexis with and without optic capture of the posterior chamber intraocular lens in the absence of vitrectomy. *J Pediatr Ophthalmol Strabismus*. 2002;39(5):278–87.

27. Heatley CJ, Spalton DJ, Kumar A, Jose R, Boyce J, Bender LE. Comparison of posterior capsule opacification rates between hydrophilic and hydrophobic single-piece acrylic intraocular lenses. *J Cataract Refract Surg.* 2005;31(4):718–24.
28. Nishi O, Nishi K, Sakanishi K. Inhibition of migrating lens epithelial cells at the capsular bend created by the rectangular optic edge of a posterior chamber intraocular lens. *Ophthalmic Surg Lasers.* 1998;29(7):587–94.
29. Brady KM, Atkinson CS, Kilty LA, Hiles DA. Cataract surgery and intraocular lens implantation in children. *Am J Ophthalmol.* 1995;120(1):1–9.
30. Khokhar SK, Pillay G, Dhull C, Agarwal E, Mahabir M, Aggarwal P. Pediatric cataract. *Indian J Ophthalmol.* 2017;65(12):1340–9.
31. Atkinson CS, Hiles DA. Treatment of secondary posterior capsular membranes with the Nd:YAG laser in a pediatric population. *Am J Ophthalmol.* 1994;118(4):496–501.
32. Gilbard SM, Peyman GA, Goldberg MF. Evaluation for cystoid maculopathy after pars plicata lensectomy-vitreotomy for congenital cataracts. *Ophthalmology.* 1983;90:1201–6.
33. Hoyt CS, Nickel B. Aphakic cystoid macular edema; occurrence in infants and children after transpupillary lensectomy and anterior vitrectomy. *Arch Ophthalmol.* 1982;100:746–9.