Chapter

Cataract Surgery in Microphthalmic Eyes

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Abstract

Microphthalmos is a congenital ocular abnormality that mainly manifests as a significant reduction in the size of the eye and is often associated with cataracts and other eye diseases. Due to its special anatomical features, cataract surgery in microphthalmos has a higher risk of intraoperative and postoperative complications and impaired visual prognosis and is associated with reduced intraocular lens (IOL) calculation accuracy. This chapter describes the characteristics of microphthalmic cataract surgery, the incidence of complications, classic and additional surgical procedures (e.g., phacoemulsification combined with prophylactic anterior lamellar sclerostomy, laser peripheral iridotomy, anterior segment vitrectomy, piggyback IOLs), and selection of IOL calculation formula.

Keywords: microphthalmos, nanophthalmos, cataract surgery, complications, prevention

1. Introduction

Microphthalmos is a congenital ocular dysplasia characterized by a small eye volume with or without dysplastic ocular structures. It has been reported that there are one to three patients with microphthalmia in every 10,000 newborns [1]. The relevant ocular malformations including cataract congenital corneal opacity chorioretinal coloboma retinal dysplasia and so on can cause visual impairment to various degrees and lead to amblyopia and even blindness [1]. Common complications of microphthalmos are congenital cataracts in adolescents and age-related cataracts in seniors. In general cataract surgery for patients with microphthalmos is challenging with poor prognosis due not only to abnormal ocular development but also to the great extent of surgical difficulty and complication rate. Possible complications such as uveal effusion or suprachoroidal hemorrhage glaucoma and corneal decompensation may seriously harm the visual outcomes by resulting in exudative retinal detachment optic nerve impairment and corneal opacity. In 1982 Singh et al. first reported cataract surgery in six patients with microphthalmia and concluded that intraocular surgery in microphthalmos can have disastrous consequences [2]. With gradual maturation of phacoemulsification (PE) and other additional surgical procedures the safety and effectiveness of cataract surgery for microphthalmos have improved [3, 4]. This chapter mainly describes the surgical effects of cataract surgery for microphthalmos different additional surgical procedures methods for preventing surgical complications and selection of intraocular lens (IOL) formulas.

2. Clinical features and diagnosis of microphthalmos

Microphthalmos is divided into three clinical subtypes based on the axial length (AL) and the anterior chamber depth (ACD) [5, 6]. (i) Small eye with a proportionally decreasing volume is characterized by short ACD and short AL and can be further divided into (a) simple microphthalmos or nanophthalmos with a normal morphology structure, and (b) complex microphthalmos associated with other congenital ocular anomalies. The diagnostic criterion is generally defined as AL < 20–21 mm, which is less than two standard deviations of the average AL of healthy people [7–10]. Wu et al. suggest that increased retinal-choroidal-scleral thickening (greater than 1.7 mm) should also be considered when diagnosing microphthalmos because the increased retinal-choroidal-scleral thickness can account for potential risks of glaucoma, uveal effusion, and other complications in microphthalmos [11]. (ii) A normal AL eye with a disproportionately narrow anterior segment is called relative anterior microphthalmos (RAM). The diagnostic criteria are corneal diameter (CD) <11 mm and ACD <2.2 mm [12]. (ii) Short AL eye with a normal anterior segment is diagnosed as axial high hyperopia eye characterized by normal ACD and short AL.

3. Cataract surgeries in microphthalmos

Cataracts are a common complication in microphthalmos. For instance, complex microphthalmos is often associated with congenital cataracts, and simple microphthalmos with age-related cataracts can occur in senior patients. Current surgical treatments for cataracts include extracapsular cataract extraction (ECCE), small-incision cataract surgery (SICS), and PE. Regarding which approach is the best choice for patients with microphthalmos, it is generally currently believed that PE is much safer. However, some studies have noted that the lower complication rate and better prognosis of PE are due to earlier surgical intervention [4, 7, 9, 10, 13]. Kohli et al. suggested that surgical procedures should be selected according to the patient's biometric measurements and the cataract severity. For patients with microphthalmia, PE is recommended for soft cataracts in eyes with CD >8 mm and SICS for hard cataracts in eyes with CD ranging between 6 mm and 8 mm [13]. In conclusion, as long as the CD and rigidity of cataract nuclei permit, PE is considered the preferred surgical procedure for cataracts in microphthalmos.

Treatment for cataracts in microphthalmos has been a major challenge in ophthalmic surgery. The unsatisfactory prognosis is due not only to abnormal ocular development but also to the narrowed operating space. A shallow anterior chamber and small cornea can lead to more difficult operative procedures and a higher incidence of complications. In 1982, Singh et al. first reported cataract extraction in six microphthalmic eyes, with disastrous outcomes [2]. However, with the gradual development of PE and other surgical techniques, the safety and effectiveness of microphthalmic cataract surgery have improved significantly [3, 4]. Multiple studies have found that the current routine surgery, namely, PE combined with intraocular lens (IOL) implantation, achieves satisfactory therapeutic effects in patients with microphthalmia and cataracts [9, 14–16]. However, including patients with relatively long AL (inclusion criteria: AL < 20.0–21.0 mm) in these studies may be the actual reason for the good surgical outcomes, as shorter AL is a significant risk factor for surgical complications [9, 14–16]. It has been reported that the total complication rate of cataract surgery is 15 times higher in eyes with AL < 20.00 mm than in those with AL < 20.00 ~ 20.99 mm [8, 9].

The author of this chapter investigated outcomes of cataract surgery in patients with extreme microphthalmos (eyes with extremely short AL or extremely small CD) [6], including microphthalmia patients with AL < 18 mm and CD < 8 mm and found that the outcome of routine PE alone was not ideal. The median best-corrected visual acuity (BCVA) score merely increased from preoperative baseline at 20/800 (equal to 0.025 in decimal visual acuity) to 20/160 (equal to 0.125 in decimal visual acuity) postoperatively. Indeed, 75% of patients still had low vision, with visual acuity lower than 20/60 (equal to 0.3 in decimal visual acuity), and 25% remained blind, with only postoperative hand motion (HM). In particular, the vision of patients with complex microphthalmia did not improve at all. The high incidence of complications is an important reason for the poor postoperative outcome of cataract surgery in extreme microphthalmos. The incidence of severe complications reaches 16.7%, much higher than that in normal eye cataract surgery ("severe complications" include uveal effusion syndrome (UES), suprachoroidal hemorrhage (SCH), retinal detachment, corneal endothelial decompensation, intraoperative posterior capsule rupture, etc.). Therefore, lowering the rate of complications in extreme microphthalmic cataract surgery is an urgent goal.

4. Prevention of complications in microphthalmos cataract surgery

With modern cataract surgical techniques, microphthalmic eyes are no longer a restriction for cataract surgery. Nonetheless, intraocular surgery in microphthalmos involves a complication rate of nearly 40–60% [17]. Common complications include UES (incidence rate: 2.63–15.79%) [7, 10, 16, 18, 19], SCH (incidence rate: 3.33–12.50%) [3, 4, 6, 11], glaucoma (incidence rate: 10.64–40.91%) [6, 20–23], and corneal endothelial edema (incidence rate: 0.87–75%) [6, 7, 18, 22, 24], among others. In general, appropriate additional surgical procedures can help prevent the occurrence of complications.

4.1 UES and SCH

UES and SCH are among the most severe complications of cataract surgery for microphthalmos and may lead to permanent vision impairment. UES is characterized by a series of fundus changes, such as subchoroidal fluid accumulation and exudative retinal detachment. The primary mechanism is congestion of choroidal veins due to compression of the vortex vein via a thickened sclera or a sudden drop in intraocular pressure (IOP) during surgery [25]. Moreover, UES may occur in unoperated eyes as well as during or after cataract surgery. Postoperative UES may also result from latent uveal effusion that existed before surgery [26]. With the development of modern surgical techniques, the incidence of UES has decreased owing to less intraoperative IOP fluctuation (incidence rate: 2.63–15.79%) [7, 10, 16, 18, 19]. Day et al. retrospectively analyzed outcomes of cataract surgery in 103 microphthalmic eyes, with only three eyes developing choroidal effusion (mean AL = 20.05 ± 1.55 mm, 14.60 –20.99 mm) [9]. Lu et al. performed PE and IOL implantation in 38 microphthalmic eyes, and only

1 eye developed exudative ciliochoroidal detachment (mean AL = 16.87 ± 1.02 mm, 15.32–18.49 mm) [19].

UES can progress to severe SCH, which often predicts poor visual prognosis. Specifically, choroidal effusion stretches the wall of ciliary arteries and leads to their rupture and bleeding into the suprachoroidal space. Influenced by the fluctuation in IOP, SCH may occur intraoperatively or postoperatively, with the latter being delayed SCH [27]. SCH can occur in all intraocular procedures, including cataract surgery, but is more commonly encountered in microphthalmos (incidence rate in microphthalmia patients: 3.33–12.50% [3, 4, 6, 11]; incidence rate in the healthy population: 0.03–0.06% [27]). In 2016, Lemos et al. reported that 1 of 14 nanophthalmic eyes (mean AL = 18.72 ± 2.23 mm, 14.00–20.45 mm) that underwent cataract surgery developed transient choroidal hemorrhage, recovering with a final BCVA of 0.15 logMAR [3]. In 2017, the author of this chapter reported cataract surgery in 30 microphthalmic eyes. In the simple microphthalmos group, SCH occurred in 1 of 11 eyes and resolved after two months, with the BCVA recovering from early postoperative HM to 20/100 (mean AL = 16.4 ± 0.8 mm) [6].

Some researchers recommend anterior lamellar sclerectomy and vortex vein decompression for UES prevention and treatment. These two procedures can create and maintain an outflow pathway for the fluid accumulated in the suprachoroidal cavity during or after surgery, and the latter can directly relieve compression of the thickened sclera on the vortex vein. Anterior lamellar sclerectomy refers to making a two-thirds thick scleral flap in size from 4 × 4 mm to 6 × 6 mm, 2–8 mm behind the limbus, with the posterior margin not beyond the equator of the eyeball. At the bottom of it, a "V"-shaped sclerotomy is generated, with the tip toward the limbus and 4 mm away from it; the length of each arm of the "V" is 1.5 mm. Additionally, the tip of the "V" (approximately 5 mm) is excised, and the sclerotomy is left open [28]. Wax et al. reported a 38-year-old female patient with bilateral nanophthalmos with acute angle-closure glaucoma (AL = 16 mm in both eyes) [28]. After pars plana vitrectomy combined with trabeculectomy, UES occurred in the left eye and disappeared after drainage by the anterior lamellar sclerectomy mentioned above. Prophylactic anterior lamellar sclerectomy was performed in her right eye before subsequent procedures. As a result, the IOP of the right eye was well controlled, and no UES occurred. Wang et al. reported a 29-year-old female patient with bilateral microphthalmos and angle-closure glaucoma (AL = 16.4 mm in both eyes). Her left eye developed UES after cataract extraction combined with trabeculectomy and peripheral iridectomy and recovered after anterior lamellar sclerectomy. In her right eye, prophylactic anterior lamellar sclerectomy followed by trabeculectomy, peripheral iridectomy, cataract extraction, and IOL implantation was performed. With stable postoperative IOP and the absence of UES in her right eye, this quadruple-combined operation had a credible preventive effect.

Vortex vein decompression refers to producing a rectangular scleral flap of 6 mm × 4 mm with long edges parallel to the equator at 4 mm in front of the outlet of the vortex vein. On this premise, the vortex vein is gradually separated backward, and a 2 mm × 2 mm deeper scleral incision is made in front of the vortex vein near the long edge of the scleral flap. Skipping the eyeball's 3 and 9 o'clock direction is vital to avoid injury to the long posterior ciliary artery and nerves [10, 29]. Recent clinical studies have confirmed the safety and effectiveness of vortex vein decompression in cataract surgery for microphthalmos. In 2017, Rajendrababu et al. conducted a randomized controlled trial to compare differences in surgical complications and visual outcomes between a sclerostomy group (cataract surgery combined with prophylactic vortex vein decompression) and control group (cataract surgery alone) (mean AL of the sclerostomy group = 17.50 ± 1.31 mm; mean AL of the control group = 18.71 ± 1.33 mm). Multivariate model analysis of 60 nanophthalmic eyes showed that prophylactic use of vortex vein decompression was associated with a lower risk of complications than cataract surgery alone (odds ratio: 0.20) but without a significant effect on the prognosis of vision and postoperative IOP [10]. In 2021, the same research team reported a retrospective study of cataract surgery in 114 nanophthalmic patients (mean AL = 17.64 ± 1.74 mm, 14.5 mm – 20.5 mm). The incidence of UES in the sclerostomy group (cataract surgery with prophylactic vortex vein decompression) was significantly lower than that in the control group (cataract surgery alone) (incidence rate of UES in sclerostomy group: 7.84%; incidence rate of UES in control group: 22.22%), though the difference in visual outcome between the two groups was not analyzed [7].

It should be noted that the scleral incision may result in new complications, including iatrogenic retinal tears, fibrovascular in-growth into the sclerostomy site, and vitreous incarceration [10, 30]. Hoffman et al. suggested that the combination of sclerectomy and cataract surgery should be considered cautiously. For existing UES, sclerostomy should be performed several weeks before cataract surgery. To prevent intraoperative and postoperative UES, prophylactic application of mannitol or acetazolamide 30 minutes before surgery to reduce vitreous volume may be a safer alternative to additional surgical procedures [31].

4.2 Glaucoma

Glaucoma is a comorbidity of microphthalmos and a common postoperative complication after cataract surgery. In microphthalmic eyes, the anatomical basis for angle-closure glaucoma is a short AL, short ACD, and narrowed anterior chamber angle. The lens is disproportionally larger and thicker compared with the volume of microphthalmos, and the area where the lens is close to the iris increases, leading to a higher risk of pupillary block. In microphthalmic eyes, angle-closure glaucoma is common before surgery, but both open-angle and angle-closure glaucoma after surgery have been reported [19, 32]. The onset of postoperative open-angle glaucoma is relatively late, usually several years after surgery and may not be directly related to the operation but to anterior segment dysplasia, including malformation of the trabecular meshwork and the Schlemm canal [23, 32].

Cataract extraction combined with IOL implantation can relieve lens-derived narrow anterior chamber angle conditions and achieve the therapeutic effect of deepening the anterior chamber. However, one study found that compared with normal eyes, patients with microphthalmos are more likely to have peripheral anterior synechiae (PAS) [19]. As a result, the postoperative effect of angle opening is often poor, and the IOP even increases slightly in microphthalmic eyes. Additionally, the success rate of combined goniosynechialysis with cataract surgery is low [19]. The author of this chapter found AL, ACD, and the degree of angle closure before surgery to be closely related to the likelihood of developing postoperative glaucoma [6]. In infants with microphthalmos, extremely early surgical intervention may be an important trigger of postoperative glaucoma [20, 21]. Praveen et al. suggested that to minimize the complication rate without compromising the final potential visual outcomes, cataract surgery in microphthalmic eyes should be postponed until the end of the first month of life [21].

Peripheral iridectomy (PI) is a common prophylactic surgical procedure used to prevent angle-closure glaucoma in microphthalmos. The dilemma between the crowded anterior segment and the normal-sized lens leads to an extremely narrowed aqueous passage on the iris's posterior surface and the lens's anterior surface, resulting in a high risk of pupillary block in microphthalmia patients. PI eliminates the pressure difference between the anterior and posterior chambers and is a standard treatment for correcting pupillary block. To date, the combination of PI and cataract surgery has achieved high recognition among ophthalmic specialists for treating microphthalmic cataracts. Prasad et al. applied PI, posterior capsulotomy, and anterior vitrectomy in combination with phacoaspiration in 37 microphthalmic infant eyes and indicated that prophylactic PI and adequate vitrectomy can reduce glaucoma incidence (mean AL = 15.76 ± 0.56 mm, 14.66–16.41 mm) [33]. Furthermore, Day and Seki et al. suggested that combined usage of PI and cataract surgery in patients at high risk of malignant glaucoma (microphthalmic eye with AL < 20 mm) may facilitate future laser zonulotomy or hyaloidotomy if aqueous misdirection occurs [9 34]. Nevertheless, Steijns et al. conducted retrospective analysis of 43 nanophthalmic eyes and found no significant correlation between PI and occurrence of postoperative angle-closure glaucoma. These authors believed that the decision to perform intraoperative PI should be based on the degree of angle closure and the individual risk of postoperative angle-closure glaucoma (average AL = 20.01 mm, 15.47–20.48 mm) [16]. The discrepant conclusions of the retrospective studies by Prasad and Steijns may be associated with differences in the mean AL of the included patients. The possibility that PI has a more significant anti-glaucoma effect in eyes with shorter ALs cannot be ruled out and needs to be verified.

Postoperative malignant glaucoma is one of the most serious complications of cataract surgery for microphthalmos. The pathogenesis involves backward flows of the aqueous humor, becoming trapped in the anterior vitreous cavity and resulting in an elevation in posterior segment pressure and a forward movement of the lens-iris diaphragm, which further obstruct the aqueous humor flow. Once a vicious cycle forms, the anterior chamber will become progressively shallow, and the IOP will increase continuously [35, 36]. Despite the development of surgical techniques, malignant glaucoma is still frequently reported after microphthalmic cataract surgery (incidence rate: 1.85–9.52%) [6, 9, 18, 19, 37, 38]. In 2013, Day et al. reported seven cases of malignant glaucoma among 103 postcataract surgery microphthalmic eyes (mean AL = 20.05 ± 1.55, 14.60–20.99 mm) [9]. In 2015, Ye et al. reported two cases of malignant glaucoma among 89 nanophthalmic eyes after cataract surgery (mean AL = 19.24 ± 1.20 mm, 15.82–20.97 mm) [18], and Rajendrababu et al. diagnosed one case of secondary malignant glaucoma after cataract surgery among 19 nanophthalmic eyes (mean AL = 18.6 ± 1.8 mm, 17.7–19.5 mm) in 2021 [39].

The principle for treating malignant glaucoma is to communicate the anterior chamber to the vitreous cavity to relieve obstruction at the interface between the ciliary body and vitreous body [40]. Iridazolulohyaloid vitrectomy (IZHV), which was first proposed by Lois in 2001 [41], has been proven to be a safe and effective technique. This procedure creates a direct aqueous humor channel between the anterior chamber and the vitreous cavity, with a lower recurrence rate than for other operations (such as laser posterior capsulotomy or anterior vitrectomy). It is an applicable procedure for surgeons specializing in cataract surgery and is a good choice for treating or preventing malignant glaucoma intraoperatively and postoperatively [42]. For example, Zarnowski et al. used IZHV to treat 10 patients with pseudophakic malignant glaucoma and achieved a cure rate of 100% (mean AL = 21.30 ± 1.06 mm, 20.15–22.31 mm) [43]. The author of this chapter has also adopted this procedure and successfully treated a case of malignant glaucoma in complex microphthalmos with congenital zonular abnormality [6]. In summary, combining IZHV and cataract surgery for microphthalmia patients with a high risk of malignant glaucoma may achieve an ideal outcome, but further investigation is needed.

4.3 Corneal edema and endothelial decompensation

An impaired corneal endothelium is an important factor affecting the visual prognosis of microphthalmia patients after cataract surgery. Corneal endothelial injury presents as corneal edema in the early stage, and in severe cases, it may gradually progress to corneal endothelial decompensation.

In intraocular surgery for microphthalmos, transient corneal edema is the most common early postoperative complication attributed to the reduced surgical space in a shallow anterior chamber and a small cornea. Both the proximity of the phaco probe to the cornea and the generation of excess heat energy result in mechanical and thermal damage to the corneal endothelium [39]. With the improvement in surgical technology, the incidence of corneal endothelial decompensation after microphthalmic cataract surgery has gradually decreased, despite high occurrence of corneal edema. Matalia et al. reported transient corneal edema in 22 of 47 eyes with microcornea after cataract surgery, with an incidence rate of 46.8% (mean CD = 8.6 ± 0.72 mm, 7–9.5 mm) [22]. Additionally, the author of this chapter reported that the incidence of postoperative transient corneal edema reaches 73% in all microphthalmia patients, with 100% in the RAM group (a total of 11 eyes, CD = 7.3 ± 1.0 mm, ACD = 1.15 ± 0.85 mm) [6]. However, except for two eyes in the RAM group that developed chronic corneal endothelial dysfunction, corneal edema in the other patients disappeared within one to two weeks.

Anterior vitrectomy combined with cataract surgery is an effective method to solve the problem of anterior segment narrowness in microphthalmic eyes. When combined with posterior capsulotomy, anterior vitrectomy can prevent visual axis opacification, the most common complication of pediatric cataract surgery, caused by anterior vitreous fibrosis and posterior capsular opacification [44]. Moreover, anterior vitrectomy helps to reduce the volume of the vitreous body and deepen the anterior chamber while decreasing the difficulty of cataract surgery and the risk of corneal endothelial injury in microphthalmic eyes [45]. Recently, phacoaspiration with primary posterior capsulotomy and anterior vitrectomy has achieved satisfactory results in clinical treatment for microphthalmos with congenital cataract [21, 33, 45]. Praveen et al. applied cataract surgery combined with anterior vitrectomy in 72 congenital cataract eyes with microphthalmos and found only four cases of visual axis opacification and no corneal edema (mean AL of the right eye: 16.7 ± 1.5 mm; mean AL of the left eye: 16.6 ± 1.3 mm) [21]. Prasad et al. combined PE, PI, primary posterior capsulotomy, and anterior vitrectomy in 37 microphthalmic eyes of 20 infants with congenital cataracts; only two eyes developed visual axis opacification, and no corneal edema occurred postoperatively (mean AL = 15.76 ± 0.56 mm, 14.66–16.41 mm) [33].

In regard to treatment of cataracts in adult microphthalmic eyes, clinicians should balance the benefits of anterior vitrectomy with its risks. Anterior vitrectomy for microphthalmos can easily lead to posterior capsule rupture during cutting and penetrating procedures because the lens/eye volume ratio is approximately 25%, much larger than the 4% in the healthy population [16]. Additionally, a risk of retinal impairment exists during anterior vitrectomy due to the close proximity between the limbus and ora serrata region in microphthalmos [16]. Therefore, for adult microphthalmic cataract surgery, an individualized clinical decision on whether to combine anterior vitrectomy needs concrete analysis, including ACD, intraoperative visibility, and surgical techniques.

5. IOL implantation and formula selection in microphthalmic cataract surgery

Microphthalmic eyes usually present high hyperopia, which requires high IOL power. For adults, IOL implantation is necessary if the ocular condition permits. Regardless, several studies have noted that primary IOL implantation should be considered cautiously for microphthalmic children with congenital cataracts [20, 33]. First, implantation of an adult-sized IOL into the small eyeball of an infant with microphthalmia requires more precise surgical technique than routine cataract surgery. Second, owing to the growth of an infant's eyes, the change in postoperative refraction is more significant than in adults. The correction achieved with frame glasses or contact lenses for the aphakic eye may be more accurate than that with IOL implantation. In addition, primary IOL implantation may increase the incidence of postoperative complications such as secondary glaucoma and retinal detachment [20, 33, 46]. A study by Praveen et al. showed that even without IOL implantation, early surgical intervention and adequate amblyopia training can improve the visual acuity of children with microphthalmos and congenital cataracts [21].

5.1 Formula selection in microphthalmos cataract surgery

A unified standard for the most appropriate formula for microphthalmos has yet to be determined. Among conventional IOL formulas, the Hoffer Q formula is recommended by the Royal School of Ophthalmologists as the preferred formula for short eyes (AL ≤ 22 mm) [47]. However, several large-scale clinical studies have found that the Haigis formula outperforms the Hoffer Q formula [48–51]. Over the last decades, new-generation IOL formulas have emerged, as follows: formulas involving artificial intelligence (Hill-Radial Basis Function, Ladas Super Formula AI, Pearl-DGS formula); formulas using a combination of theoretical optics, thin lens formulas, and big data techniques (Kane formula); and formulas based on theoretical optics with regression and ray-tracing components (Olsen formula, Okulix formula). Among new-generation formulas, the Kane formula has been demonstrated to be relatively suitable for microphthalmic eyes in a few studies [50, 52, 53]. However, the update of the IOL formula is not equivalent to replacement. Luo et al.'s meta-analysis of 1476 microphthalmos (AL ≤22 mm) in 14 studies showed that the accuracy of new-generation formulas, such as Barrett Universal II and Kane, were generally superior to conventional IOL formulas (including the Haigis, Hoffer Q, SRK/T formulas) [54]. Moreover, Shrivastava and associates performed a meta-analysis of 15 studies involving 2395 short eyes (AL ≤22 mm) and concluded that there is no significant difference between the Barrett Universal II formula, the Olsen formula, and conventional IOL formulas (including the Haigis, Hoffer Q, SRK/T formulas) [55]. The author of this chapter also compared the accuracy of six IOL formulas (including the Haigis, Hoffer Q, Holladay I, SRK/T, Barrett Universal II, and Hoffer QST formulas) in nanophthalmos and RAM patients. When using the IOL Master 500 for biometric measurement (omitting the parameter of the lens thickness), the Haigis formula was the most accurate among the six IOL calculation formulas for cataract patients with nanophthalmos (mean AL = 16.84 ± 1.36 mm, 15.25–19.82 mm), whereas the Barrett Universal II formula showed the highest accuracy in cataract patients with RAM (mean CD = 8.41 ± 0.92 mm, 7.00–9.50 mm) [56]. Certain limitations in the new-generation formulas have been found, including requirements for more biometric parameters and specific instruments than traditional formulas and restrictions in calculating the IOL power of short eyes by the scope of the database [56]. As a result, application of new-generation formulas in nanophthalmos and RAM eyes needs further investigation.

5.2 Piggyback IOL implantation

Microphthalmos with high hyperopia usually requires high-power IOLs (generally greater than 40 diopters), which is generally out of the normal range and inaccessible in clinical practice [57]. In 1993, Gayton implanted two IOLs in the same eye for the first time, pioneering the procedure of piggyback IOL implantation [58]. However, this technique remains controversial among clinicians. Proponents believed that the piggyback technique has the advantage of a higher correction power and a greater postoperative benefit than traditional single-piece IOL implantation [57, 59], and opponents have concerns about its intraoperative safety. Because of the crowded anterior chamber and the fragile zonula, it is a challenge for surgeons to implant two IOLs in the same microphthalmos, which will certainly prolong the operation time and amplify the risk of intraoperative complications [8, 9]. This author's previous study reported that during piggyback IOL implantation, a patient with microphthalmos developed SCH after the first IOL was inserted into the capsule [6]. Moreover, with this approach, the unique postoperative complication interlenticular opacification severely impairs postoperative visual acuity [57].

In summary, implantation of a customized single IOL is the safest choice for microphthalmic eyes. If a high-power IOL cannot be obtained, experienced surgeons may adopt piggyback IOL implantation, placing two pieces of IOLs in the capsular bag and the sulcus, respectively [31, 57]. During the operation, attention should be given to thoroughly cleaning the capsule to minimize excessive proliferation of lens epithelial cells and avoid occurrence of interlenticular opacification [57].

6. Conclusion

Cataract surgery in microphthalmos is no longer forbidden, and PE is the first choice of surgical treatment. In microphthalmic eyes with a relatively long AL, PE can generally achieve satisfactory surgical outcomes. However, extreme microphthalmic eyes (with extremely short AL) still have a high risk of complications. Additional application of anterior lamellar sclerectomy or vortex vein decompression may be a good option to prevent UES and SCH. PI may reduce the incidence of postoperative angle-closure glaucoma, and the IZHV procedure can effectively treat or prevent malignant glaucoma. Anterior vitrectomy may be used to deepen the anterior chamber and reduce corneal endothelial damage. However, it should be noted that additional surgical procedures may bring new risks of complications.

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