REVIEW ARTICLE

Small Eyes Big Problems: Is Cataract Surgery the Best Option for the Nanophthalmic Eyes?

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Abstract

Nanophthalmos refers to an eyeball of short axial length, usually less than 20 mm which leads to angle closure glaucoma due to relatively large lens. Intra-ocular lens extraction relieves the angle closure in nanophthalmos. Cataract surgery in a nanophthalmic eye is technically difficult with high risk of complications such as posterior capsular rupture, uveal effusion, choroidal haemorrhage, vitreous haemorrhage, malignant glaucoma, retinal detachment and aqueous misdirection. Various options are explained in the literature to perform cataract surgery in nanophthalmos, like extracapsular cataract extraction with or without sclerostomy; small-incision cataract extraction by phacoemulsification which not only helps maintain the anterior chamber during surgery but also reduces the incidence of complications due to less fluctuation of intraocular pressure (IOP) during the surgery. Cataract surgery deepens and widens the anterior chamber angle in nanophthalmic eyes and has beneficial effects on IOP in eyes with nanophthalmos but is associated with a high incidence of complications.

Key Words: *Nanophthalmos. Cataract surgery. Choroidal effusion. Angle closure glaucoma.*

INTRODUCTION

Nanophthalmos is a rare congenital anomaly characterized by a short axial length of the eyeball, which is usually less than 20 mm.[1,2] 54 – 77% of nanophthalmic patients develop angle-closure glaucoma because the lens is relatively large for a small eye.[3-6] Multiple mechanisms are involved in angle closure in nanophthalmos like pupillary block, plateau iris configuration, choroidal expansion and forward lens movement.[4,5]

There is a strong evidence that suggests that the lens extraction relieves the angle closure in nanophthalmos. However, cataract surgery in a nanophthalmic eye is technically difficult with high risk of complications such as posterior capsular rupture, uveal effusion, choroidal haemorrhage, vitreous haemorrhage, malignant glaucoma[6,7] retinal detachment and aqueous misdirection.[2]

Despite recent advancements in cataract surgery, visual outcomes after cataract surgery in nanophthalmic eyes are highly unpredictable due to high refractive surprises and macular changes after the surgery.[7,8-11]

METHODOLOGY

Review of the literature and journal articles was conducted to find out the success of cataract surgery in nanophthalmic patients and to suggest ways to avoid complications in nanophthalmic eyes while performing cataract surgery.

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DISCUSSION

Challenges of intervention: The size of the crystalline lens in nanophthalmic eyes is within the normal range; therefore, the crystalline lens/globe volumetric ratio, which is 4% for normal eyes, increases upto the pathological level of 10 – 30% in nanophthalmics leading to chronic, painless glaucoma in these patients at an early age due to the natural increase in the size of the lens with age.[12-14]

The intraocular pressure (IOP) lowering response to conventional medical treatment in nanophthalmic glaucoma is usually poor due to the structural change within the drainage mechanism.[15,16] Miotics may make the raised IOP worse by relaxing the lens zonule and producing relative pupillary block.

Laser iridotomy and iridoplasty are moderately successful initially but later due to peripheral anterior synechia; it becomes ineffective in keeping the IOP lower. Fistulizing glaucoma surgery like trabeculectomy could lead to malignant glaucoma due to postoperative ciliary block and choroidal effusion, due to changes in the dynamic of IOP during and after the surgery.[15,16]

Removal of the intraocular lens could relieve the obstruction of the filtration meshwork and keep the angles open, avoiding potential glaucoma and helps to have better control of intra-ocular pressure post-operatively.[15,16]

Eyes with nanophthalmos seemed to pose a significant challenge during cataract extraction because of the

reduced working space, due to shallow anterior chamber and a small corneal diameter.

Nanophthalmic eyes have a high incidence of intra-operative and postoperative complications, including uveal effusion, retinal detachment, vitreous haemorrhage, or malignant glaucoma, suprachoroidal haemorrhage, iris prolapse, persistent corneal oedema, and cystoid maculopathy.[7,16-19]

Surgical manipulation in a narrow and crowded anterior chamber with increased vitreous pressure, tends to cause papillary blockage which is the most challenging situation to manage in the nanophthalmic patients.[17-19]

The reduction of intra-ocular lens size after cataract surgery not only opens-up the drainage angle[20] but also helps to reduce the intraocular pressure in eyes with nanophthalmos.

Pentacam and ultrasound biomicroscopy have shown an increase in anterior chamber volume, depth, and opening of the angles after extraction of cataracts from nanophthalmic patients; strongly suggesting the morphological change following cataract extraction.[20]

Pre-operative workout: Before planning any surgery in nanophthalmics, it is prudent to do careful pre-operative workout.
Ultrasound B scan to assess retinal-choroidal-scleral thickness in nanophthalmic eyes allow physicians to do careful pre-operative assessment and plan appropriate operative procedures to prevent subsequent complications like choroidal effusion.[7]

Accurate intraocular lens (IOL) power calculation is the most important aspect of cataract surgery outcome to avoid unnecessary refractive surprises. Calculation of IOL powers remains the major challenge in nanophthalmic eyes and has an impact on poor postoperative refractive outcome due to shallow anterior chamber depth, the short axial length (AL), the small corneal diameter and relatively high power IOL.[1,21] No formula for calculation of IOL power is accurate; Inatomi and associates suggested Hoffer Q and Holladay-II formulas are equally good to calculate IOL power for small eyes.[22] On the contrary, Narváez and associates reported that the Hoffer Q, Holladay-1, Holladay-II, and SRK/T IOL power formulas in the eyes with an AL less than 22.0 mm showed similar accuracy in mean absolute error.[23] The least accurate method of IOL power calculation proved to be SRK-II due to myopic shift.[24]

Hence, no single formula is accurate to calculate IOL power in nanophthalmic eye, therefore, clinicians are advised to discuss the refractive outcome with the patients and consider the un-predictability of IOL power calculation before operation.

It is strongly suggested to keep the cataract surgical option only in patients with un-controlled glaucoma and symptomatic cataract.

Is extracapsular cataract extraction (ECCE) the best option? Extracapsular cataract extraction with or without sclerostomy could be an option for management of nanophthalmos where cataract surgery with phaco-emulsification is not possible due to logistic reasons and very shallow anterior chamber in the nanophthalmic patients. Thickened lens and crowded anterior chamber could make phacoemulsification difficult. Classical extracapsular cataract extraction (ECCE) with primary posterior chamber intraocular lens implant (PCIOL) could be the best option in such cases. It is important to reduce the intraocular pressure pre-operatively with medical therapy in cases of nanophthalmos.[14]

ECCE can prevent the serious complication of posterior capsule rupture as compared to phacoemulsification. PCIOL acts as a barrier against enormous vitreous pressure, prevents the pushing of the anterior chamber structures and keeps the drainage angles open in this way.[25]

Role of prophylactic sclerostomy to avoid the uveal effusion syndrome is always controversial. However, Jin *et al*. reported that nanophthalmic uveal effusion can be prevented or treated with an unsutured sclerotomy or sclerectomy before ECCE.[26]

Is phacoemulsification the best option? Small-incision cataract extraction by phacoemulsification allows the surgeon to perform well controlled surgery and helps maintain the anterior chamber during surgery which could help to reduce the incidence of complications due to less fluctuation of IOP during the surgery. The best approach in such patients for cataract surgery with phacoemulsifcation is the biggest dilemma faced by the ophthalmic surgeons today. Few available options are discussed below keeping in mind one or none could work for such a patient and the outcome of surgery is higly unpredictable.

Prophylactic lamellar sclerostomy with decompression of the vortex veins at the time of cataract surgery by phacoemulsification with IOL implantation could be helpful to prevent choroidal and retinal detachment post-operatively.[17] However, advances in a small-incision cataract surgery technique (Phacoemulsification) are considered safe and sometimes could obviates the need for prophylactic sclerotomies in nanophthalmic patients.[18] Therefore, full thickness lamellar sclerectomy should be kept reserved for patients with established choroidal effusion.[27]

The poor refractive outcome and poor predictability is usually due to shallow anterior chamber depth, the short axial length, the small corneal diameter, relatively high power IOL[1] and high posterior capsular rupture rates with phacoemulsification in such patients.[24]

Nanophthalmics with intumescent lens and subsequent severe anterior chamber shallowing, could be benefited

from pars plana vitrectomy to create the space in the anterior chamber for lens extraction with phaco-emulsification.[19]

Recently, Varma *et al*. suggested to perform a planned pars plana vitrectomy tap to create additional working space in the anterior chamber intraoperatively followed by phacoemulsification, insertion of capsular tension ring to help draw the IOL-capsule complex posteriorly.[28] In the presence of Plateu iris and peripheral anterior synechiae: a modified endocyclophotocoagulation (ECP) to address Plateau iris and goniosynechialysis helped to release peripheral anterior synechiae and reported good postoperative refractive outcome.[28]

How to avoid intraoperative complications? Sudden lowering of IOP during the cataract surgery is a known risk factor to develop uveal effusion syndrome in small axial length eyes which could be prevented by oral steroids, intravenous acetazolamide, and mannitol pre-operatively and operative sclerotomies to release suprachoroidal fluid.[29]

It is very important to avoid hypotony during surgery and maintain AC volume throughout the procedure with abundant injection of viscoelastic materials.[30]

Various techniques to prevent choroidal effusion are described; like lens extraction through pars plana vitrectomy with gas exchange,[31] prophylactic lamellar scleral resection with decompression of the vortex veins.[29]

Due to small working space in the anterior chamber and high vitreous pressure; higher incidence of posterior capsular rupture is noted in patients with nano-phthalmos.[32,33] This can not only have a significant effect and postoperative refractive outcome but also on the long-term management of these patients for raised IOP. The reported incidence of posterior capsular rupture varies from 1 out of 8 (12.5%)[3] to 2 out of 17 (11.7%)[24] in nanophthalmic eyes.

Various methods can be adopted to prevent posterior capsular rupture like controlling the IOP fluctuation during phacoemulsification by creating water tight incisions, adjustment of bottle height with the depth of anterior chamber, peroperative pars plana vitrectomy to reduce the posterior vitreous pressure and create space within the anterior chamber.[19,28] Diuretics, intravenous mannitol or laser iridotomy could be helpful pre-operatively. Topical anaesthesia could be safe for performing surgery as it could better reduce the posterior vitreous pressure rather than peribulbar or retrobulbar anaesthesia.[24]

Endothelial cell loss is a major reason for postoperative persistent corneal oedema leading to visual disappoint-ment and poor refractive outcome. Nanophthalmic patients have greatest mean endothelial cell loss following cataract surgery as compared to micro-phthalmic and normal eyes.[24] The soft-shell technique with dispersive-cohesive viscoelastic could prevent the endothelial cell loss significantly during cataract operation.[24,33]

How many IOLs? The nanophthalmic IOL are high power IOLs due to small axial length and small cornea. The high power lenses are usually special order lenses and sometimes difficult to get hold of and are relatively expensive.

However, the current system of lens power calculation markedly underestimates the required lens power, resulting in a hyperopic refractive error after surgery. The benefits of in-the-bag placement of 2 or more foldable lenses are questionable,[34] due to high incidence of posterior synechia, interlenticular opacifi-cation, posterior lens opacification, IOL displace-ment, hyperopic refractive errors,[35,36] angle closure glaucoma[36,37] and pigment dispersion syndrome.[38]

Ideally one should try to use single IOL in the bag following cataract surgery if possible in all cases to achieve a better refractive outcome, less intraoperative and postoperative complications.[39] Single IOL also creates more deepening of anterior chamber as compared to multiple piggyback lenses.

CONCLUSION

Cataract surgery deepens the anterior chamber and widens the anterior chamber angle in nanophthalmic eyes which may have beneficial effects on IOP in eyes with nanophthalmos. However, it is associated with a high incidence of complications.

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