

## Nanophthalmos with angle closure glaucoma, a challenging case



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### ABSTRACT

**Introduction:** Nanophthalmos basically is a simple microphthalmos with clinical characteristics of short axial length (AL), shallow anterior chamber (AC), high lens to eye volume ratio, small cornea and thick sclera, associated with angle closure glaucoma. We report a case of nanophthalmos with angle closure glaucoma and cataract who underwent trabeculectomy and end up with malignant glaucoma. Pars plana vitrectomy (PPV) and phacoemulsification with posterior chamber intraocular lens (PC IOL) implantation was done to manage the case.

**Case Description:** A 41-year-old man came with blurred vision in the right eye with redness and headache for 8 weeks. Intraocular pressure (IOP) was 46 mmHg, visual acuity was counting fingers at 2 m with nystagmus. Slit lamp examination revealed short axial length (AL), shallow anterior chamber (AC), small corneal diameter and high lens to eye volume. Trabeculectomy with 5FU was done with complication of malignant glaucoma. After PPV combined with phacoemulsification and PC IOL implantation, IOP was 14 mmHg, visual acuity 0.125 with deep AC.

**Conclusion:** Surgical intervention in nanophthalmos with angle closure glaucoma must be managed with good preparation. Malignant glaucoma in this case can be managed with pars plana vitrectomy and phacoemulsification without scleral procedure.

**Keywords:** Nanophthalmos, angle closure glaucoma, malignant glaucoma, case report.

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## INTRODUCTION

Microphthalmos is an ocular defect with an axial length less than 2 SD below the normal age group.<sup>1</sup> Nanophthalmos basically is a simple microphthalmos with clinical characteristic of short axial length (AL), shallow anterior chamber (AC), high lens to eye volume ratio, small cornea and thick sclera.<sup>2</sup> Nanophthalmos is often associated with varying degree of angle closure glaucoma. Angle closure glaucoma is often present between the age of 40-60 years, due to progressive shallowing of AC and narrowing of the angle due to thickening of the lens with age.<sup>3</sup> Surgical intervention in these eyes is challenging due to high risk of intraoperative and postoperative complication. Sudden decompression during surgery may trigger uveal effusion in thickened and weak sclera. Nanophthalmos has a high risk of serious complication such as suprachoroidal hemorrhage, non-rhegmatogenous retinal detachment, and malignant glaucoma.<sup>4</sup>

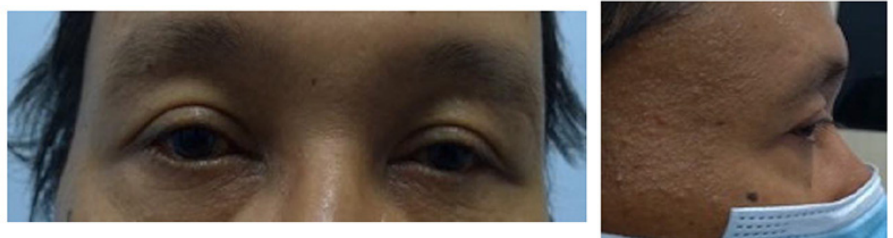
We report a case of nanophthalmos that came with angle closure glaucoma and cataract who underwent trabeculectomy without scleral procedure and lead to malignant glaucoma.

## CASE PRESENTATION

A 41-year-old man, works as a public transportation driver, was referred to tertiary eye hospital with gradually decreased vision in the right eye for 3 months and significantly since 8 weeks accompanied by redness, pain and headache. The patient has been treated by a local ophthalmologist for 6 weeks with topical and oral anti glaucoma. History

of thick glasses since childhood with Sph +18.00 D since 7 years old, now the patient wears Sph +13.50 D in both eyes. No history of eye trauma. No family history of glaucoma, no other family member wears thick glasses. Systemic condition was unremarkable.

Patient has a small eye from external examination (**Figure 1**). Best corrected visual acuity in the right eye was counting fingers at 2 m and 0.08 in the left eye, with nystagmus. Intraocular pressure measured by Goldmann applanation tonometer was 44 mmHg in the right eye and 14 mmHg in left eye. Slit lamp examination in the right eye revealed ciliary injection, microcystic



**Figure 1.** External examination.

corneal edema, pigment deposition in corneal endothelium, shallow AC with grade 2 Van Herick, fixed mid dilated pupil and lens opacity with grade 3 nuclear sclerosis. Funduscopy was deferred because of edema cornea.

Slit lamp examination in the left eye revealed shallow AC with grade 2 Van Herick, lens opacity with grade 2 nuclear sclerosis. Funduscopy revealed cup to disc ratio 0.5. Gonioscopy with Sussman four mirror was deferred in the right eye because of hazy media, closed angle in the left eye with peripheral anterior synechiae in 3 quadrants after indentation. Short axial length was revealed from B-scan ultrasonography (Nidec echoscan US-4000) with sclerochoroidal complex thickness 1.60 mm in the right eye and 1.68 mm in the left eye (Figure 2). Optical biometry was done using IOL master-700 (Carl Zeiss Meditec AG, Jena, Germany) showed axial length 15.74 mm, anterior chamber depth 2.17 mm, lens thickness 1.16 mm, white to white corneal diameter 10.9 mm in the right eye. Axial length 15.47 mm, anterior chamber depth 2.02 mm, lens thickness 1.36 mm, white to white corneal diameter 10.9 mm in the left eye. Calculated IOL Power for the right eye was +46.7 D. Anterior OCT (Zeiss Cirrus 4000 HD-OCT) showed convex iris configuration and closed angle in both eyes (Figure 3).

Glaucoma medical therapy with acetazolamide tablets and topical beta blocker 0.5% were given. Combined phacotrabeculectomy was planned. Intraocular pressure before surgery was 46 mmHg, 20% mannitol 5cc/kg BW intravenous was given. After mannitol, IOP was 42 mmHg then the surgery switched to trabeculectomy with 5FU without inferior sclerostomy. Preplaced scleral flap suture, slow decompression through paracentesis track and tight suture closure were done. In the end of surgery, AC cannot be reformed, and the palpable intraocular pressure was high. After trabeculectomy, bleb was formed, the IOP range between 32-46 mmHg with persistent shallow anterior chamber, patent iridectomy, mid dilated pupil with posterior synechiae (Figure 4). Ultrasonography revealed no choroidal effusion nor suprachoroidal hemorrhage.

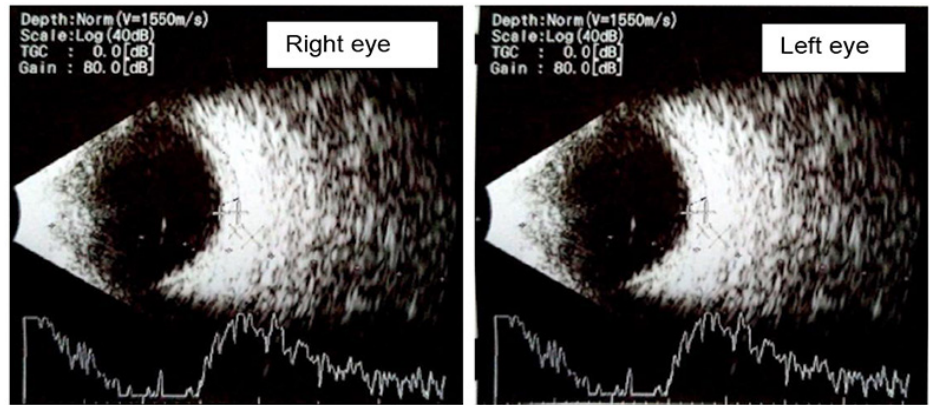


Figure 2. Ultrasound b scan revealed short axial length in the right eye and left eye.

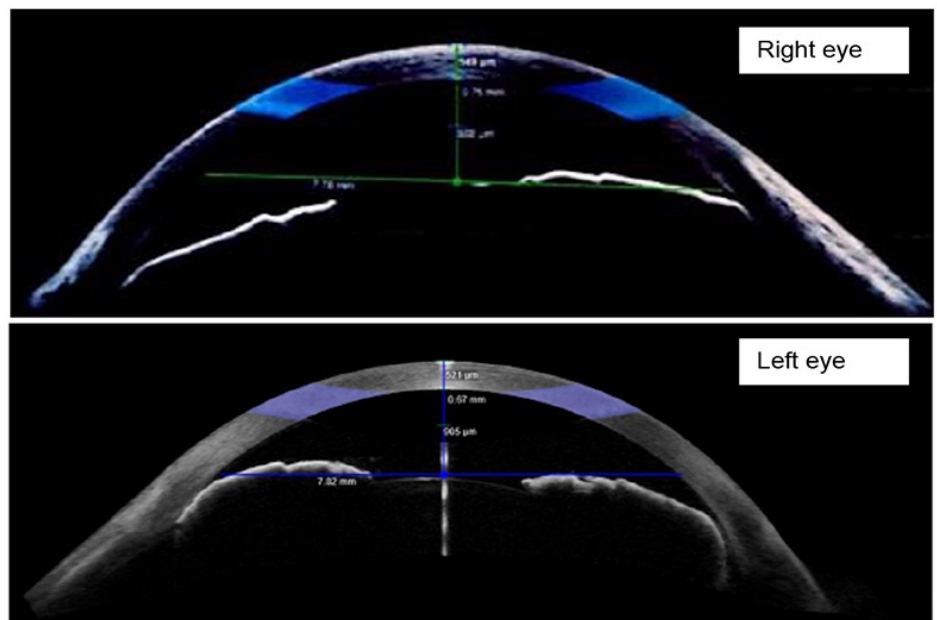


Figure 3. Oct showed convex iris and shallow anterior chamber in the right eye and left eye.

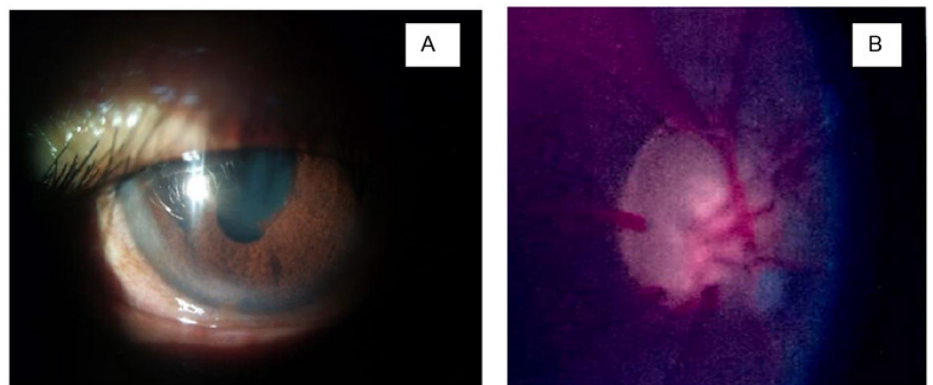


Figure 4. After trabeculectomy. A). Shallow anterior chamber and posterior synechiae; b). Funduscopy showed advanced cupping.

Malignant glaucoma post trabeculectomy was suspected, acetazolamide tablet, topical beta blocker, cycloplegic and mydriaticum given.

Intraocular pressure remains high in the range of 32-46 mmHg with persistent shallow AC, cup to disc ratio 0.9. Two months after trabeculectomy, pars plana



vitrectomy and phacoemulsification was planned under general anesthesia. Intraocular pressure was 46 mmHg, 20% mannitol 5cc/kgBW intravenous given. Stage procedure, combined pars plana vitrectomy and phacoemulsification was done. Retinal surgeon performed pars plana vitrectomy (Alcon® constellation vision system) with 23 G system, trocar was inserted 2-2.5 mm behind the limbus. Core vitrectomy proceeds until the eye softens and AC deepens. Posterior synechiae released then phacoemulsification proceeded in usual manner (Alcon® constellation vision system), hydrophobic acrylic foldable lens inserted in the bag with maximal power available +30.00 D. Finally pars plana vitrectomy proceeded to take the vitreous in vitreous base and hyaloid membrane (Figure 5). Zonulectomy was not performed in this case. The pressure was stable during surgery, we tried not to make any movement to disrupt the stability of the anterior chamber. Sclerostomy was not done and there was no uveal effusion detected.

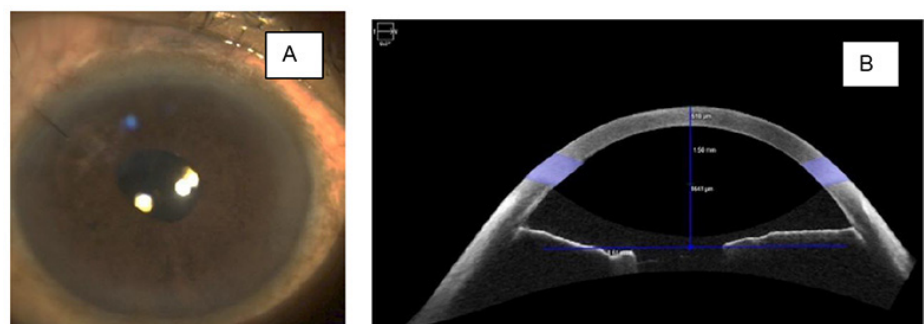
One month after surgery, visual acuity of the right eye was 0.125, IOP 14 mmHg with topical timolol maleate 0.5%, deep anterior chamber, PC IOL in place (Figure 6). Laser peripheral iridectomy done in the left eye.

## DISCUSSION

Microphthalmos is an array of ocular malformation that may appear as congenital or acquired disorders. These developmental arrest result from a various of genetic defect that induce abnormalities during early ocular embryogenesis in 7-8 weeks of gestation.<sup>1,5</sup> It characterized by AL less than 2 SD below the mean for age (< 20,9 mm in adult).<sup>1</sup> Microphthalmos can be classified into simple if the eyeball is small but otherwise normal or complex (complicated) if associated with other ocular or systemic malformation either isolated or as part of a syndromes.<sup>1,6</sup> Clinical manifestation range from complete microphthalmos in which anterior and posterior segment both reduced in size or partial microphthalmos with shortening of either anterior (relative anterior microphthalmos) or posterior segment (posterior microphthalmos).<sup>7,8</sup>



**Figure 5.** Pars plana vitrectomy combined with phacoemulsification.



**Figure 6.** One month after surgery. A). Slit lamp examination; b). Oct shows deep anterior chamber.

Nanophthalmos basically is a simple and complete microphthalmos, often defines as a dwarf eye.<sup>5</sup> Nanophthalmos is a rare disease, with prevalence of 0.0009-0.017%, but potentially blinding disease if not treated properly.<sup>9</sup> Its characteristic clinical findings include short AL,<sup>1</sup> high hyperopia (+8.00D - +25.00D or higher),<sup>6</sup> deep set orbit, narrow palpebral fissure, microcornea (<11.00 mm),<sup>6</sup> steep corneal curvature (> 46 D),<sup>8</sup> shallow AC (<3mm),<sup>6,10</sup> angle closure, high lens to eye volume ratio (10-30%),<sup>10</sup> thickened sclera (scleralchoroid complex > 1.7 mm)<sup>2</sup> with abnormal collagen fibrils. Fundus abnormality that can be found are yellow macular pigmentation, chorioretinal folds, crowded optic disc, retinal striae, wrinkling of internal limiting membrane, recurrent uveal effusions and non-rhegmatogenous retinal detachment.<sup>7</sup> These clinical condition may lead to glaucoma and retinal complication.<sup>2</sup>

Nanophthalmos is often associated with varying degree of angle closure, incidence of angle closure disease and angle closure glaucoma is around 70% and 35.7% respectively.<sup>4,7</sup> Angle closure glaucoma is often present between the age of 40-60 years due to thickening of the lens

with age. There are multiple mechanisms of angle closure in nanophthalmos, crowding anterior chamber secondary to high lens to eye volume ratio causing chronically appositional iris and trabecular meshwork that lead to progressive peripheral anterior synechiae. The large lens pushes iris forward to the already crowded anterior chamber lead to pupillary block.<sup>3</sup> Abnormal collagen arrangement lead to thickened and weak sclera which impairs venous drainage through vortex veins that may lead to ciliochoroidal effusion which cause displacement of peripheral iris by anteriorly rotated ciliary processes. Uveal effusion also can induced relative pupillary block by relaxed zonulae that allowing anterior displacement of lens and increasing iridolenticular contact.<sup>9,11</sup>

In this case, the patient has short axial length, shallow anterior chamber, small corneal diameter, high lens to eye volume ratio, convex iris configuration and closed angle. Sclerochoroidal complex is thicker than normal in both eyes, but not more than 1.7 mm. The clinical findings are characteristic of nanophthalmos. He has had high hyperopia and nystagmus since childhood, he has been using glasses since 7 years old with possibility of amblyopia.

He is 41 years old and has complained of blurred vision for 3 months that may be due to immature cataract and chronic angle closure glaucoma. He complained of redness, pain and headache for 8 weeks that may be due to acute on chronic angle closure glaucoma. There is a possibility that extensive peripheral anterior synechiae has developed and cataract increased pupillary block. Unrecognized and untreated high IOP in the right eye had occurred long enough to cause advanced glaucomatous optic neuropathy in the right eye.

Management of angle closure in nanophthalmos may be challenging. Laser peripheral iridectomy and or argon laser peripheral iridoplasty should be performed to relieve pupillary block components in the early stage of glaucoma. If extensive synechial angle closure has developed, management of elevated IOP with laser and medical treatment is not effective and surgical treatment will be required. Response to medical treatment is poor in nanophthalmos with angle closure glaucoma.<sup>11</sup> Surgery for glaucoma in nanophthalmos is often considered as a last resort due to high risk of intraoperative and postoperative complication. Sudden decompression during surgery in thickened and weak sclera may trigger uveal effusion, retinal detachment, suprachoroidal hemorrhage or malignant glaucoma with loss of vision. Post operative complication as shallow anterior chamber, increased anterior chamber reaction, malignant glaucoma and suprachoroidal hemorrhage often occur.<sup>4</sup> Glaucoma filtering surgery such as trabeculectomy and or goniosynechiolysis can be performed with several scleral procedures include anterior sclerostomy, posterior sclerostomy or vortex vein decompression, to prevent uveal effusion.<sup>12</sup>

Beside high risk of uveal effusion, conventional glaucoma filtering surgery is to be avoided because fear of malignant glaucoma.<sup>13</sup> Malignant glaucoma occurs 2-4% in eyes undergoing surgery for angle closure glaucoma. Thick choroid, crowded anterior chamber and small ciliary ring diameter has a high risk to develop ciliary block or malignant glaucoma especially in surgery with unstable anterior chamber.<sup>14</sup> A disproportionally large lens in nanophthalmos increased ciliolenticular

apposition and short axial length with small anterior chamber depth associated with more anterior location of ciliary body which might be a contributing factor for the occurrence of malignant glaucoma.<sup>5</sup>

If glaucoma filtering surgery is done, it should be performed with caution in early stage before permanent damage occurs and lens extraction must be anticipated.<sup>13,15</sup> Lens extraction has a role in the management of nanophthalmos, it may deepen anterior chamber, widen the angle, relief of pupillary block and prevent progressive angle closure.<sup>15</sup> Surgical manipulation in shallow, crowded anterior chamber with high IOP and increased vitreous pressure tends to cause pupillary block and challenging with poor visual outcome and potential complication such as uveal effusion, retinal detachment, corneal decompensation and glaucoma malignant. Cataract surgery and PC IOL implantation with prophylactic sclerostomy should be considered to avoid such complication.<sup>15</sup> Cataract surgery with small incision technique such as phacoemulsification, seemed to be safe and may obviate the need for prophylactic sclerotomies.<sup>15</sup> In intumescent lens and subsequent severe anterior chamber shallowing, the use of pars plana vitrectomy may successfully manage the problem.<sup>3,15</sup>

In this patient, laser or medical management will not be effective because of extensive peripheral anterior synechiae, advanced glaucoma and very high IOP. Surgical intervention is the last choice available in this challenging case. Combined trabeculectomy and cataract extraction was deferred because of high IOP even after intravenous mannitol and trabeculectomy was chosen instead. Unfortunately this was a wrong decision, because trabeculectomy alone can trigger malignant glaucoma and trabeculectomy without scleral procedure carry a high risk of uveal effusion. After surgery, malignant glaucoma was suspected because of persistent shallow anterior chamber, patent peripheral iridectomy, high IOP and no sign of abnormality in posterior segment from B-scan ultrasonography. Preplaced suture, careful slow decompression and tight suture cannot prevent the occurrence

of malignant glaucoma, maybe because of high risk of anatomical predisposing factor. Limitation of this case report is that we don't have UBM to confirm this condition.

In the management of malignant glaucoma, medical anti glaucoma therapy combined with topical cycloplegics and mydriatics should be given to tighten the lens zonules and pull lens backward. In aphakic and pseudophakic eyes, Nd-yag laser used to perform posterior capsulotomy and hyaloidotomy so direct communication between vitreous cavity and anterior chamber can be established.<sup>19</sup> If it refractory to medical and laser therapy, surgical intervention to remove the vitreous with pars plana vitrectomy need to be done.<sup>15</sup> The success rate of core vitrectomy in phakic eye is only 25-50% compare to 65-90% in pseudophakic eye. Core vitrectomy combined with cataract extraction can increase the success rate to 25-83%. Combining pars plana vitrectomy with cataract surgery was a strategy to reduce intraoperative risk in eyes with high IOP and shallow anterior chamber. Removal of anterior vitreous will facilitate posterior displacement of the lens, deepen the anterior chamber and decrease positive vitreous pressure. This will make the phacoemulsification maneuver easier and safe. The stage approach can be done, first core vitrectomy done to soften the eye and deepen the anterior chamber, followed by standard phacoemulsification and PC IOL implantation. Finally residual vitrectomy, hyaloidectomy and posterior capsulectomy. Some author also suggest to do anterior vitrectomy combined with zonulohyaloidectomy to make connection between anterior chamber and vitreous cavity.<sup>13</sup> Pars plana vitrectomy in nanophthalmos has a special challenge, trocar should be inserted more anterior because of short pars plana and care should be taken during insertion not to injure the opposite retina. Fluctuation of the fluid and marked hypotony should be avoided to prevent uveal effusion.<sup>13</sup>

In this case, maximal medical therapy, cycloplegic and mydriatic eye drops were given but the condition has not improved. Laser hyaloidotomy cannot be performed in this case since the patient is phakic. Now the problem is angle

closure glaucoma in nanophthalmos with malignant glaucoma after trabeculectomy. Advanced glaucomatous cupping was noted to make the urgency to do surgical intervention. Pars plana vitrectomy procedure will give double advantages, like killing two birds with one stone in this case. Pars plana vitrectomy will provide a deep anterior chamber and soften the eye for safe phacoemulsification as well as to manage ciliary block in malignant glaucoma. Surgical intervention was planned as soon as inflammation subsided. General anesthesia is chosen to lower the intraocular pressure because of high IOP even after intravenous mannitol. Stage pars plana vitrectomy combined with phacoemulsification and PC IOL implantation successfully done without any complication. We put maximal IOP power available +30.00 D which will make it under correction since the biometry result was + 46.70 D. We did not make prophylactic sclerostomy and there was no sign of uveal effusion after surgery. Trabeculectomy with prophylactic inferior sclerostomy, cataract extraction and PC IOL implantation, combined with pars plana core vitrectomy will be the best and safest option in the first place for this patient who has nanophthalmos with cataract and angle closure glaucoma.

There is a risk of long term failure of trabeculectomy as a consequence of additional procedure and prolonged high IOP with shallow anterior chamber. So close follow up is mandatory. Laser peripheral iridotomy done for the fellow eye. The fellow eye has visually significant cataract, normal IOP, shorter axial length, shallow anterior chamber, thicker lens, thicker sclerochoroidal complex and more convex iris configuration. The best management option for the fellow eye will be staging pars plana vitrectomy and cataract extraction to prevent progressive angle closure and glaucoma malignant complication. Longer observation is needed for this patient to see the effectiveness of the procedure.

## CONCLUSION

Management of eyes with nanophthalmos and glaucoma is always a challenge for ophthalmologist. Early diagnosis and proper management are very important. Surgical intervention must be plan properly. Malignant glaucoma is a common complication in nanophthalmos eye. Stage procedure pars plana vitrectomy and phacoemulsification without scleral procedure effectively manage this case. There is an improvement of management outcome in nanophthalmos eye with the progress of surgical technique in glaucoma, cataract and retinal surgery.

## ETHICAL STATEMENT

The patient received signed written informed consent regarding publication of medical data in scientific medical journals before any data collection with confidentiality of personal information.

## CONFLICT OF INTEREST

There is no conflict of interest or financial interest related to the manuscript.

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## AUTHORS CONTRIBUTION

EG responsible for concept and design of the study, definition of intellectual content, literature search, data acquisition, manuscript preparation, manuscript editing, manuscript review, and guarantor. AP responsible for manuscript review, and guarantor of the study. RMR responsible for concept and design of the study, definition of intellectual content, and manuscript review. SU responsible for manuscript preparation, manuscript editing.

## REFERENCES

- Elder MJ. Aetiology of severe visual impairment and blindness in microphthalmos. *Br J Ophthalmol.* 1994;78(5):332-4.
- Wu W, Dawson DG, Sugar A, Elnor SG, Meyer KA, McKey JB. Cataract surgery in patients with nanophthalmos: Results and complications. *J Cataract Refract Surg.* 2004;30(3):584-90.
- Yang N, Jin S, Ma L, Liu J, Shan C, Zhao J. The pathogenesis and treatment of complications in nanophthalmos. *J Ophthalmol.* 2020;2020.
- Rajendrababu S, Shroff S, Uduman MS, Babu N. Clinical spectrum and treatment outcomes of patients with nanophthalmos. *Eye.* 2021;35(3):825-30.
- Guo C, Zhao Z, Zhang D, Liu J, Li J, Zhang J, et al. Anterior segment features in nanophthalmos with secondary chronic angle closure glaucoma: An ultrasound biomicroscopy study. *Investig Ophthalmol Vis Sci.* 2019;60(6):2248-56.
- Carricondo PC, Andrade T, Prasov L, Ayres BM, Moroi SE. Nanophthalmos: A review of the clinical spectrum and genetics. *J Ophthalmol.* 2018;2018: 2735465.
- Relhan N, Jalali S, Pehre N, Rao HL, Manusani U, Bodduluri L. High-hyperopia database, part I: Clinical characterisation including morphometric (biometric) differentiation of posterior microphthalmos from nanophthalmos. *Eye.* 2016;30(1):120-6.
- Yang N, Zhao LL, Liu J, Ma LL, Zhao JS. Nanophthalmos: An update on the biological parameters and fundus abnormalities. *J Ophthalmol.* 2021;2021: 8853811.
- Yosar JC, Zagora SL, Grigg JR. Cataract surgery in short eyes, including nanophthalmos: visual outcomes, complications and refractive results. *Clin Ophthalmol.* 2021;15:4543-51.
- Yalvac IS, Satana B, Ozkan G, Eksioğlu U, Duman S. Management of glaucoma in patients with nanophthalmos. *Eye.* 2008;22(6):838-43.
- Burgoyne C, Tello C, New T, Eye Y. Cases in controversy nanophthalmia and chronic angle-closure glaucoma questions. *J Glaucoma.* 2002;11:525-8.
- Singh OS, Simmons RJ, Brockhurst RJ, Trempe CL. Nanophthalmos: A perspective on identification and therapy. *ophthalmology.* 1982;89(9):1006-12.
- Calhoun FP. The management of glaucoma in nanophthalmos. *Trans Am Ophthalmol Soc.* 1975;73:97-122.
- Chen X, Guo X, Xu X, Xiao H, Liu X. Is thicker choroid a risk factor for malignant glaucoma? *ophthalmic res.* 2018;60(3):161-8.
- Yuzbasioglu E, Artunay O, Agachan A, Bilen H. Phacoemulsification in patients with nanophthalmos. *Can J Ophthalmol.* 2009;44(5):534-9.



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