Commentary: Short eyes and bigger challenges - Growing evidence in the management of pediatric nanophthalmos

Complete microphthalmos, also known as nanophthalmos, is a rare developmental ocular anomaly that is characterized by short axial length (≤20 mm), microcornea, high hypermetropia (+8D to +25D), relative anterior or posterior microphthalmos, and global reduction in ocular volume.[1] It is usually a bilateral symmetrical condition with high corneal curvature, high lens/eye volume ratio, narrow anterior chamber angle, and thickened sclera. The clinical appearance is small sunken deep, set eyes (relative enophthalmos), and narrow palpebral fissure height; occasionally, mild ptosis can be present.[2] The majority of cases are sporadic. Autosomal dominant and recessive modes have also been reported in the literature. NNO1 and NNO3 genetic loci are associated with autosomal dominant non-syndromic nanophthalmos, and NNO2 is associated with autosomal recessive non-syndromic nanophthalmos.[3] Nanophthalmos can present as an isolated entity or spectrum of various syndromes such as oculo-dento-digital (ODD) syndrome, foveoschisis, retinitis pigmentosa, optic drusen syndrome, Kenny–Caffey syndrome, and autosomal dominant vitreoretinochoroidopathy with nanophthalmos (ADVIRC).[4] Children with nanophthalmos are at high risk of developing strabismus, amblyopia, angle-closure glaucoma, retinal detachment, choroidal detachment, and uveal effusion syndrome. Various posterior segment pathologies such as cystoid macular edema, retinal and choroidal folds, crowded optic disc, sclerochoroidal thickening, and pigmentary retinal dystrophy have also been reported with nanophthalmos. In nanophthalmos, there is growth arrest due to rearrangement of scleral collagen and may not be associated with structural eye defects. The diagnosis in these cases is based on meticulous anterior and posterior segment examination, cycloplegic retinoscopy, applanation tonometry, gonioscopy, keratometry, A-scan, structural analysis of optic nerve head, B-scan, ultrasound biomicroscopy (UBM), and visual evoked potential (VEP) in required cases. The management options available are spectacles, strabismus surgery for non-refractive esotropia, laser peripheral iridectomy for angle closure, filtration surgery, and cataract surgery. Nanophthalmos management is still a therapeutic challenge for clinicians; thus, prompt diagnosis and meticulous management are warranted in each case to safeguard vision and prevent irreversible complications. The clinical and biometric parameters of nanophthalmos in children are less well understood, and the clinical features may overlap with relative anterior microphthalmos and simple, partial, complex, and posterior microphthalmos. There is limited literature available on pediatric nanophthalmos. Recently, some studies have offered promising insights into the clinical, morphological, biometric features, and management aspects.

Agarkar *et al*.[5] analyzed the clinical and morphometric characteristics of 75 children under 18 years with nanophthalmos. They reported that ametropic amblyopia was the major cause of visual impairment; 17 children had occludable angles, posterior segment pathology was reported in 28 children, and the mean axial length was 16.88 mm. They concluded that nanophthalmos patients had short axial length, high hyperopia, and shallow anterior chamber, and 20% of children had occludable angles. Wu *et al*.[6] assessed the outcomes and complications of cataract surgery in 12 eyes of eight nanophthalmic patients. They concluded that echography should be done to know the retinochoroidal thickness in hyperopic eyes with a shallow anterior chamber that are at risk for angle-closure glaucoma. They also found that phacoemulsification is safe in these eyes with or without scleral lamellar resection. All cases of nanophthalmos require a careful preoperative assessment to prevent intraoperative and postoperative mishaps. Steijns *et al*.[7] performed cataract surgery in 43 eyes of 32 nanophthalmic patients and achieved good outcomes in nearly 71% of the patients, with good BCVA in 70% of cases. Only 12 patients had complications; the most common complications were uveal effusion and cystoid macular edema.

The current study[8] compares the long-term clinical and biometric characteristics between the nanophthalmic children and age-matched controls, and the authors must be congratulated for this excellent comparative analysis. The major reasons for visual impairment were ametropic amblyopia, strabismus, angle-closure, and pigmentary retinopathy, which is in accordance with the previously available literature. The striking finding of the study was that 50% of the patients had angle closure, which is comparatively on the higher side probably to less sample size as compared to Agarkar *et al*.[5] Another interesting take-home message is that although 50% of children had occludable angles, the decision to perform laser peripheral iridectomy was based on multiple factors such as age, family history, IOP on the first visit, and cooperation of the patient rather than gonioscopic findings alone. This is critical while managing pediatric nanophthalmos. The authors had also reported an interesting observation that the presence of peripheral anterior synechiae in adults has 3.66 times higher odds for developing angle-closure glaucoma. Gonioscopy is not always possible in pediatric children; thus, another take-home message is that pediatric nanophthalmic children should undergo serial biometry and, whenever possible, gonioscopy to prevent the development of vision-threatening angle-closure glaucoma. The study is also unique because it presents the largest database of nanophthalmic eyes and age-matched controls. The authors also compared the biometry between eyes with an axial length of less than or more than 17 mm. To conclude, nanophthalmos always remains a clinical challenge for all ophthalmic surgeons, and pediatric nanophthalmos pose further challenges in management due to patient cooperation and the high risk of irreversible vision loss. The authors must be congratulated again for this novel, interesting, and rare analysis on pediatric nanophthalmos.

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