Small eyes—big problems

Intellectuals solve problems, geniuses prevent them.
Albert Einstein

Nanophthalmos is a rare condition included in the spectrum of developmental eye diseases in which the ocular growth is compromised after the closure of the embryonic fissure. This compromised ocular growth can range from anophthalmos to nanophthalmos and microphthalmos. Nanophthalmos is derived from the Greek word nano, meaning dwarf. These eyes typically exhibit very high to extreme axial hyperopia and lack overt structural defects.1,2 Historically, this condition has had many synonyms, including pure microphthalmos, simple microphthalmos, and complex microphthalmos, depending on the presence of other associated ocular or systemic abnormalities.3-6

Epidemiologic data regarding the prevalence of nanophthalmos/microphthalmos in adults are few. Birth prevalence of microphthalmos varies from 0.002 to 0.017 in a British cohort7 to 0.0009% in a Chinese population.8

Data from a hospital sample in the United States reported a rate of 0.05% to 0.11% for simple microphthalmos.9 In the recently published European Prospective Investigation of Cancer (EPIC)—Norfolk Eye Study,10 the prevalence of an axial length (AL) of less than 21.0 mm was 1.20% and of less than 20.0 mm was 0.27%. This suggests that the prevalence of small eyes is more common than previously reported.

There has been great heterogeneity in the definition of nanophthalmos in the literature. Weiss et al.3 suggested an AL of less than 18.0 mm as a definition; however, it is unclear why and how this value was chosen. Axial length varies not only with age and sex but also with height, weight, education, and occupational class, as clearly shown in a multivariate analysis.11 With the current data, I think it would be reasonable to define microphthalmos as eyes with an AL of less than 21.0 mm (2 standard deviations [SDs] below the mean value) and nanophthalmos as eyes with an AL of less than 20.0 mm (3 SDs below the mean value), as suggested by Day et al.12

The clinical phenotypic characteristics of nanophthalmos include narrow palpebral fissures, deep-set small eyes in the orbit, extreme hyperopia, steep corneas, a shallow anterior chamber, and a normal-sized lens occupying a disproportionately large percentage of the intraocular volume. Histopathologically, these eyes have unusually thick sclera with irregular collagen, reduced levels of glycosaminoglycans, and elevated levels of fibronectin.13 It is likely that the reduced scleral permeability to proteins by the thickened sclera combined with compression of the vortex veins by the dense abnormal collagen might contribute to the high incidence of postoperative uveal effusions in these abnormal eyes.14

Cataract/lens-based surgery in nanophthalmic eyes is fraught with complications. Compared with the extremely low complication rate in normal eyes, the reported incidence of intraoperative and postoperative complications is 15.5% (16 of 103 cases) in microphthalmic eyes and nanophthalmic eyes.15 The complication rate is higher in eyes with an AL of less than 20.0 mm than in eyes with an AL between 20.0 mm and 21.0 mm. In a multivariate analysis, the AL and intraocular pressure (IOP) emerged as significant predictors, implying that the shorter the AL and higher the preoperative IOP, the greater the chances of intraoperative and postoperative complications after lens-based surgeries.12

In this issue, Singh et al. (pages 2394–2402) report a retrospective multicenter study of the use of a new high-powered diffractive intraocular lens (IOL) during lens-based surgery in nanophthalmic eyes (AL <20.0 mm). Although not approved by the U.S. Food and Drug Administration, the availability of this new custom-made high-powered single-piece hydrophilic IOL (CT Xtreme D, Carl Zeiss Meditec AG) will help avoid the use of piggyback IOLs in these very small eyes. However, in their cohort, the incidence of postoperative complications was 42.9% (9 of 21 eyes), with severe complications in 23.8% (5 of 21 eyes). Also in this issue, Hoffman et al. (pages 2565–2575), on behalf of the ASCRS Cataract Clinical Committee, provide a review of and update on cataract surgery in small eyes, highlighting issues such as preoperative assessment, challenges in IOL power calculation, and postoperative complications and their management.

Performing cataract/lens-based surgery in nanophthalmic eyes continues to be one of the greatest surgical challenges for the anterior segment surgeon. Technological advances in hardware and software with regard to micromanipulation and computerized vitrectomy systems have certainly helped surgeons to use minimally invasive surgical techniques. The availability of this new single-piece high-powered custom-made injectable IOL will further help us reduce the morbidity and improve
postoperative refractive outcomes with the use of modern fourth-generation IOL formulas in these challenging eyes.

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REFERENCES