

IOL Power Calculation in the Short Eye 64

David Flikier

Introduction

Cataract extraction surgery with the intraocular lens implant in a short or small eye is one of the most complex interventions for the anterior segment surgeon, [\[1,](#page-9-0) [2](#page-9-1)] and its biometrical difficulty is inversely proportional to the axial length [[3](#page-9-2)].

With the improvement of surgical techniques, instruments, equipment, design, and intraocular lens material, both outcome optimization and patients' expectations have increased for these difficult cases. The success of the final visual outcome, obtained with emmetropia, is still one of the critical issues to resolve. The great variability of dimensions of the internal ocular structures and the diffculty in the estimation of the effective lens position are the culprits for the errors in the calculation formulas, even for the latest generation ones.

Defnition

What constitutes a short eye? The diagnostic parameters include: **the axial length, the corneal diameter, and the anterior chamber depth.** The existence of different eye patterns, which include variable corneal diameters and

normal or narrow anterior chambers, can aid us in classifying a short eye and to anticipate modifcations in their pre, trans, and postoperatory management, in order to avoid complications.

The median axial length oscillates between 22.76–23.55 mm \pm 1.17–1.49 mm [[4–](#page-9-3)[9\]](#page-9-4), which is why a standard deviation below that threshold would get closer to 22.0 D and two standard deviations, would remain within the 20.5 D range. These values have generally been used as starting points in order to describe short eye classifcations. In Melles et al.'s [\[10](#page-9-5)] work, they studied 27,191 eyes and considered small eyes whose axial length was shorter than 22.5 mm, where they included the lowest 10% of the population under study.

Short Eye Classifcation

The axial hyperopic eye is a short eye, which has a length shorter than 22.0 mm and up to 20.5 mm, placing it outside of the frst standard deviation, but within the second one; they are considered normal short eyes, due to their anatomical characteristics and diffculties in their calculation of the intraocular eye. (see Fig. [64.1.](#page-1-0))

The clinical spectrum of the short eye varies in a phenotypical range, according to the relative sizes for the anterior and posterior segments, and D. Flikier (\boxtimes)
it is classified in [[2,](#page-9-1) [12–](#page-9-6)[16\]](#page-9-7) (see Fig. [64.2\)](#page-1-1):

Instituto de Cirugía Ocular, San José, Costa Rica

 \degree The Author(s) 2024 893

J. Aramberri et al. (eds.), *Intraocular Lens Calculations*, Essentials in Ophthalmology, [https://doi.org/10.1007/978-3-031-50666-6_64](https://doi.org/10.1007/978-3-031-50666-6_64#DOI)

Fig. 64.1 Eye classification according to anatomical characteristics, axial length vs. size of the anterior segment. (Modifed from Holladay's diagram [[11](#page-9-8)])

Fig. 64.2 Representation of short eye variants, comparing the axial length, anterior chamber depth, corneal diameter, crystalline lens thickness, and scleral thickness. (**a**) Eye with normal parameters, (**b**) Microphthalmos, with axial length reduction, (**c**) Nanophthalmos, with

axial length reduction, anterior segment, crystalline lens thickness, and an increase in scleral thickness, (**d**) relative anterior Microphthalmos, with its reduced anterior segment, (**e**) Posterior Microphthalmos with axial length reduction, by a reduction of the posterior pole

- (a) Simple Microphthalmos.
- (b) Complex Microphthalmos.
- (c) Nanophthalmos.
- (d) Relative anterior Microphthalmos.
- (e) Posterior Microphthalmos.

Microphthalmos corresponds to an eye with short axial length and is classifed into two types: *Simpl*e and *Complex*, based on the presence of ocular anatomical malformations.

- 1. **Simple Microphthalmos:** is an eye with short axial length, without ocular malformations. As short, we mean two standard deviations (2 SD) shorter than normal for the age group. Historically, it has been reported as shorter than 20.5 mm in adults and shorter than 17.8 mm in children up to one year of age. Other epidemiological studies have defned this value at 21.0 mm for the adult [\[17\]](#page-9-9), present in 0.046– 0.11% of ophthalmological patients. These eyes are hyperopic, but they have a normal anterior chamber and normal scleral thickness. They are not at risk for angle-closure glaucoma.
- 2. Complex **Microphthalmos:** It is an eye with small axial length and anatomical malformations. As in the simple microphthalmos, the axial length is more than two standard deviations shorter than its age group. Besides, they can present with marked ocular anatomical malformations, such as coloboma of the iris, chorioretinal coloboma, persistent fetal vasculature, and retinal dysplasia. They also have normal scleral thickness.
- 3. **Nanophthalmos:** It is a condition where there is also a short eye, with a small anterior segment, and a thick choroid and sclerotic [[2\]](#page-9-1). There is no consensus for the axial length, which defnes the nanophthalmos, but there are reports that range from less than 20.5 mm [\[18](#page-9-10)], 20.0 mm [[19\]](#page-9-11), 18.0 mm [[16\]](#page-9-7), and 17.0 mm [[20\]](#page-9-12), though accepting those at 20.0 and 20.5 mm as the most recent ones [[1,](#page-9-0) [3](#page-9-2), [21](#page-9-13)[–24](#page-10-0)]. These eyes are constituted by:
	- (a) **Anterior chambers** that keep narrowing, as the crystalline grows with age.
	- (b) **Convex iris** with propension towards painless angle-closure chronic glaucoma.

(c) **Scleral and choroid thickness increase**, larger than 1.7 mm [[18,](#page-9-10) [25](#page-10-1)] predisposing to uveal effusion.

Also, in 2016, Guo et al. [\[25](#page-10-1)] described the characteristic features of ciliary body ultrasonic biomicroscopy, iris, and the eye angle with Nanophthalmos, both for chronic primary angle-closure glaucoma and for chronic secondary angle-closure glaucoma. The typical feature for Nanophthalmos is a small eye, with a narrow anterior chamber, and where the growth of the crystalline lens is the cause for the development of chronic secondary angle-closure glaucoma, with symptoms comparable to primary angle-closure.

They may have associated microcorneas with shorter diameters than 11 mm [\[26](#page-10-2)]. The microcornea is a distortion that can be observed in any of the short eyes: *simple microphthalmos, complex microphthalmos, nanophthalmos, and anterior relative microphthalmos.*

- 4. **Relative anterior Microphthalmos**: name coined by Naumann [[27\]](#page-10-3), it is an eye with a normal axial length, but with a small anterior segment, with an axial length longer than 20.5 mm, but with an ACD equal or lesser than 2.2 mm and a corneal diameter shorter than 11 mm [\[12](#page-9-6), [28\]](#page-10-4). They have no other ocular anatomical malformations, nor any associated increase in scleral growth. It is sub-diagnosed before cataract surgery due to its normal axial length. However, it is crucial to differentiate, due to the high incidence of angle-closure glaucoma, cornea guttata, and pseudoexfoliation association.
- 5. **Posterior Microphthalmos:** It is an extremely rare condition, typically recessive, with an anterior segment at normal dimensions, but with a posterior shortening, from a reduction of the growth of the posterior seg-ment that results in high hyperopia [[29–](#page-10-5)[31\]](#page-10-6). Due to the scleral thickening, the choroidal and pigment epithelial growth is limited, but with normal neuroretinal development, inducing the development of papillomacular folds [\[32](#page-10-7)] including the retina without pigment epithelium or choroidal tissue. They may be

associated to several pathologies such as: esotropia, peripheral avascular zones without vascular crest formation, uveal effusion syndrome [[28\]](#page-10-4), pigment retinopathy, retinoschisis, and retinal dialysis [[14,](#page-9-14) [33–](#page-10-8)[36\]](#page-10-9).

Kaderli et al. [[37\]](#page-10-10) reported in 2018, in a normal and short eye study, using an EDI-OCT test, that the thickness of the choroids and the diameter of large-sized choroid vessels in the posterior pole increases are inversely proportional to the axial length, regardless of sex or patient's gender.

It is important to understand that this complex of anatomical characteristics for nanophthalmos and posterior microphthalmos is associated with a symmetrical reduction of the axial length and high hyperopia, such as a variable phenotypical spectrum, but could also be the expression of the same genetic mutation, usually variable biallelic in MFRP [\[38](#page-10-11)] and PRSS56 [[39,](#page-10-12) [40\]](#page-10-13) and rare monoallelic in TMEM98 [\[41](#page-10-14), [42](#page-10-15)] and MYRF. Variable expressions of the gene can be found in a same family, with nanophthalmos in some members and microphthalmos of different magnitude in others. In the case of PRSS56, the production of a soluble protease stimulating the axial length was found, through a function gain mechanism, also implicated in the development of myopia [[43\]](#page-10-16). In a near future, we could think about treatments through protease inhibitors or monoclonal antibodies.

The genetic origin of these entities is polygenic; and the axial length is raised as associated to the degree of involvement by deep intronic or regulatory variants in the four known genes [\[43,](#page-10-16) [44\]](#page-10-17).

Characteristic Features of Short Eyes

Achieving a precise refractive outcome in the short eye is a real challenge, and often enough, simple axial length, keratometry, and ACD parameters become insufficient.

The best calculation formulas are those that can predict the *effective lens position (ELPo)* in

a more exact manner, but even so, the standard deviation is high.

The three main variables in the calculation of the intraocular lens are:

- 1. **The power of the cornea.**
- 2. **The axial length.**
- 3. **The effective lens position**.

In any eye, the main challenge will always be the estimation of the fnal lens position, based on preoperatory measurements. In the large eye with high myopia and a low power intraocular lens, the ELPo is **not** critical; a small anteroposterior movement would produce a very small refractive deviation. In contrast, the short eye, with the combination of certain variables such as narrow chambers, thick crystalline, steep corneas, and higher powers, produces the ideal mix to derail the prediction algorithms for the effective lens position, severely affecting the fnal refraction.

Third-generation formulas use only two variables for ELPo prediction: *Keratometry and axial length*. They assume that the greater corneal curvatures have a deeper chamber, but in reality, in short eyes, the anterior chambers tend to be narrow, and therefore the outcome estimates a lens position more posterior than the desired one.

With the formulas that use ACD (anterior chamber depth) to calculate the effective lens position, such as the Haigis formula, the opposite happens, by neglecting to take the thickness of the crystalline lens into account. In the short eye, chambers are narrow and therefore Haigis formula estimates a very anterior position, but since the thickness of the crystalline is large, the position ends up being a little more posterior.

Due to the introduction of new variables, in *fourth-generation formulas such as*: **Corneal Diameter: CD, Lens thickness: LT, and the Anterior Chamber Depth: ACD** associated to the keratometry and axial length, the estimation of the effective lens has improved, and therefore the calculation power for the lens.

As it was previously mentioned, according to the classifcation of the short eye, each type of eye will have its own specifc characteristics, but as a general rule, and as an average data (taken from the Kane et al. study $[45]$ $[45]$, we find:

- 1. A reduction of the axial length.
- 2. A relative increase in the percentage of the ratio between the anterior and posterior segments, due to an **increase in the thickness of the crystalline lens**; in average 4.7 mm and ± 0.42 mm SD.
- 3. Narrow anterior chambers, with a median 2.61 mm and ±0.39 mm SD.
- 4. Steep keratometries, with a median 43.81 D ±1.76.
- 5. Some smaller corneas.

In the year 2008, Erdol et al. [[34\]](#page-10-19) reported a couple of cases of posterior microphthalmos with an apparent normal anterior segment, where ultrasonic biomicroscopy was performed, demonstrating a thickened and anteriorized ciliary body [\[34](#page-10-19)]. These data increase the possibility that the equator of the crystalline lens is not in normal relationship with the other measurements (Axial length, Keratometry, ACD, LT, CD, age, etc.) usually considered in order to estimate the position of the intraocular lens. Therefore, the theory is introduced; when dealing with short eyes, the placement of the equator of the crystalline lens, the position of the ciliary body and sulcus, could be of greater value to improve the estimation for the effective positioning of the intraocular lens.

In the year 2016, Goto et al. [\[46](#page-10-20)] conducted a study in search for other variables in order to improve the estimation of the postoperative ACD. They found that the depth of the angle to angle, measured with AS OCT from the angular recess, and introduced as a regression coefficient in a prediction formula for the ELPo for the postoperatory ACD, along with the preoperatory ACD and the axial length, using Haigis and SRK/T formulas, increased predictions for the IOL power in a signifcant manner and reduced the residual postoperatory defect (98.7% \pm 0.50 D). As interesting data, the study demonstrated that the anterior segment in the nanophthalmos is

Fig. 64.3 Representation of the normal eye vs. variants of short eyes, reduction of the ciliary ring (diameter of the ciliary body, CBD), the anterior rotation of the ciliary processes against the equator of the crystalline lens, and the vault of the crystalline lens (in a more anterior position)

Fig. 64.4 Representation for short eyes, reduction of the ciliary body (Ciliary Body Diameter, CBD), anterior chamber (ACD), lens-ciliary body-lenticular vault (LCLV), angle to angle distance (ATA)

more crowded, *due to the reduction of the ciliary ring (ciliary body diameter, CBD), the anterior rotation of the ciliary processes against the equator of the crystalline, and the vault of the crystalline (in a more anterior position);* all of them risk factors for developing malignant glaucoma.

For the topic at hand in this chapter, the proposal rests on the fact where the crystalline lens anteriorization, the modifcation of the position of the equator, and the ciliary body, with regard to the anatomic positions used as baselines to calculate the ELPo, are the origin of the calculation error for the IOL power. Therefore, these three variables: the position of the ciliary body, the position of the equator, and the vault of the crystalline lens, must be considered in order to help defne the effective lens' position (see Figs. [64.3](#page-4-0), [64.4](#page-4-1), and [64.5\)](#page-5-0).

Fig. 64.5 Representation of short eyes, reduction of the ciliary ring (Ciliary Body Diameter, CBD), crystalline lens with greater thickness, with reduction of the anterior chamber (ACD), lens-ciliary body-lenticular vault (LCLV)

Biometry in the Short Eye

A small error on the axial length, ACD, and crystalline lens thickness measurements will result in a greater refractive error in the patient with short eye. This is why we require equipment to measure the axial length and the different intraocular structures in a very precise manner.

From the beginnings of optical biometry by partial coherence interferometry (PCI), its manufacturers decided to use a group refractive index and a calibration function to calculate the geometrical axial length from the measured optical path length (OPL) in spite of the differences in the refraction indexes for each of the structures in the eye and the travel speed difference when crossing them [[47–](#page-10-21)[50\]](#page-11-0). Using this calibration, the axial length measurements are accurate only for average eyes. The problem arises in extreme eyes, and especially in the case of short eyes, where proportionally, the crystalline lens occupies a greater percentage of the anterior segment and the eye.

Wang et al. [\[50](#page-11-0)] reported higher axial length measurements in short eyes, when segmentation with correction for refractive indexes for each segment was done. This difference with the real axial length, measured with the sum of the segments, versus the measurements by the biometers, explains in part the myopization observed with the calculation of the majority of the thirdgeneration short eye formulas [[47,](#page-10-21) [50\]](#page-11-0).

Cooke et al. [[47\]](#page-10-21) described the method to modify the axial length, with a regression formula (Cooke-modifed AL), in the Hoffer Q, SRK/T and Holladay I and 2 formulas, noticeably improving the results, both for long eyes and for short eyes. The separation of the segments can be achieved through automatic detection with the Spike Finder developed by David Cooke. Being able to measure each structure with its real refractive index and obtaining the precise measurements leads to better results with the IOL power calculation, specifcally in short eyes with the Holladay 1 and 2, Hoffer Q, and SRK/T; with the Haigis formula only if the constants were optimized. The results did not improve or were worse with Barrett and Olsen formulas; and with OKULIX, only improved in the study of short eyes [\[47](#page-10-21)[–50](#page-11-0)].

Since the internal limiting membrane is hardly identifed by the optic biometers, detecting the interface between the retina and the pigment epithelium, the length of the optic trajectory of the vitreous really becomes the vitreous and retinal one. In order to determine the length of the vitreous, the retina is given a theoretical thickness, which will then be subtracted from the vitreousretinal thickness in the segmentation or sum of segments [[49\]](#page-10-22). This retinal theoretic value has been generally calculated according to the axial length, accepting that long eyes have a thinner thickness and short eyes a thicker one. With the emergence of OCT, a precise measurement of the foveal thickness will be accomplished to incorporate this real measurement in calculation programs.

Another important consideration in biometric variants is the relative size of the anterior chamber in comparison with the axial length. The calculation for the power of the IOL tends to be more precise in a short eye with a proportionally smaller anterior chamber than with a deep cham-ber. Holladay et al. [[11,](#page-9-8) [51\]](#page-11-1) discovered that approximately 20% of eyes with short axial length have a small anterior segment and are classifed as nanophthalmics and the remaining 80% have a normal anterior segment (see Fig. 64.3). Eyes with a fat ACD tend to require IOLs with +30.0 D or less, whereas those with normal ACDs require IOLs with more than +40.0 D [\[13](#page-9-15)], which entails Piggy-back lens systems and wider anterior segments.

The closer to the retina the IOL is, the greater the transcendence will be for a small change on ELPo and in its refractive result. The A constant used in the IOL power calculations depends on multiple factors, including: the type of lens used, the refractive index of the material, the geometry, the variance of the biometric equipment, the surgical technique, and factors affecting ELPo. This is why in a small eye, the A constant must be **personalized.**

Results in Short Eyes

In normal eyes, 90–98% of the cases reach their final refraction between ± 1.00 D, whereas in eyes with nanophthalmos, with shorter lengths than 20.5 mm, only 46–66% achieve theirs [\[3](#page-9-2)].

Third-generation formulas only use the axial length and the corneal curvature (keratometry). **Fourth-generation** formulas such as Haigis and Holladay 2 and other more modern ones like Barrett Universal II and others include a greater number of parameters [\[52](#page-11-2)], mainly the **depth of the anterior chamber ACD** [[21,](#page-9-13) [53\]](#page-11-3), increasing the quality of the results. Eom et al. found that Haigis formula has better results than the Hoffer Q formula, in short eyes with narrow chambers, $ACD < 2.4$ mm $[54]$ $[54]$.

It should be made clear that in the studies the median axial length for short eyes is very variable and oscillates between 19.53 and 21.69 mm. However, for the very small eye with simple microphthalmos group, there is no comparative statistic study for formulas, rather isolated case reports [[22–](#page-10-23)[24\]](#page-10-0).

In the Melles and colleagues study [[10\]](#page-9-5) also with short or small eyes, shorter than 22.5 mm (between 21.0 and 22.5 mm), several interesting conclusions were found:

- 1. Barrett and Olsen's formulas had the best behavior.
- 2. Hoffer Q tends towards a myopic outcome, by reducing the axial length.
- 3. Haigis and SRK/T tend towards the hyperopic defect in very fat anterior chambers.
- 4. Hoffer Q and Holladay 1 tend towards myopia.
- 5. Olsen and Haigis tend towards hyperopia.

Other more recent studies such as Shivastava and colleagues [[55\]](#page-11-5), also in short eyes, but not as small, between 20.76 and 21.96 mm, found no statistically signifcant differences when comparing the Barrett Universal, the Hill RBF method, Haigis, Hoffer Q, and Holladay 2, with an outcome that coincides with two previous studies by Kane and cols. [\[56](#page-11-6)] and by Gokce et al. [\[57](#page-11-7)]. It is interesting to highlight that in the study by Shivastava [\[57](#page-11-7)], the median absolute error found within the ± 0.50 D range oscillates between 46 and 56%, and ± 1.00 D between 76 and 80%.

In the year 2018, Wang et al. [[58\]](#page-11-8) conducted a meta-analysis with 1161 cases, in order to compare Haigis, Holladay 2, Hoffer Q, SRK/T, and SRK II formulas. In short eyes, a frank superiority was found for the Haigis formula over the other ones. More recently, Melles et al. [\[59](#page-11-9)] found better performances in the formulas by Kane, Olsen (with 4 factors), and Barrett, followed by EVO and Hill RBF 2, over Holladay 2, Haigis, Hoffer Q, and SRK/T.

Sudhakar et al. [\[55\]](#page-11-5) in 2019, in a study of hyperopic eyes (19.77–22.06 mm), compared the intra-operatory aberrometry, the Hill RBF method, and several formulas: Barrett Universal, Holladay, Haigis, and Hoffer Q. Among their conclusions, it is interesting to see that they didn't fnd the aberrometer to be superior to the studied formulas, in the cases where the difference in the predictions was higher than 0.50 D. Taking the value of the aberrometer as fnal value, they only estimated it as adequately in half of the cases, and none of the methods obtained a result superior to ± 0.50 D in more than 60% of patients.

The most recent report, from Kane and Melles [\[45](#page-10-18)], for 270 eyes from 182 patients including smaller eyes, with axial lengths starting at 18.86 mm up to 22.46 mm, intraocular lenses with 30 D or more, found mean absolute errors oscillating between 0.838 and 0.533, with SD $\pm 0.812 - 0.707$ and median absolute errors oscil-

Short axial length sub analysis (≤22.0 mm)

Fig. 64.6 Studies in eyes with short axial lengths, showing the date when it was conducted, and the formula presenting the best results. In blue, you can observe that on

lating between 0.696 and 0.371. Continues with less than 60% of patients having a fnal refraction between ± 0.50 D, with better statistic results in Kane and EVO 2.0 formulas, followed by Haigis, Holladay 2, Olsen, and Hill RBF 2 and fnally Barrett y Hoffer Q.

Confrming these latest data, Hipólito-Fernandes et al. [[60\]](#page-11-11) conducted a study where they compared 13 formulas in several eye sizes. In the short eye group (20.82–22.0 mm), they found the best results with the VRF-G, EVO 2.0, and Kane formulas.

In the year 2020, Kane along with Chang [\[61](#page-11-10)] conducted a very complex review of the literature of the last 10 years and concluded that currently the best results are obtained with Kane's formula, followed by good results with the Olsen formulas (4-factors), Haigis, and Hill-RBF (see Fig. [64.6](#page-7-0).).

Poly-pseudophakia: Piggy-Back Lens

The piggy-back lens option was described for the frst time by Gayton and Sanders [\[62](#page-11-12)] in the year 1990, in a 31-year patient with microphthalmos, requiring an approximate lens with 46 D for both eyes. For the surgery in the frst eye, the second lens implanted in the sulcus left a residual hyperopic defect at +8.00 D, requiring its replacement. For the second eye, an empirical calculation allowed for a closer emmetropic result.

the most recent studies, with the greatest number of patients (table sizes), the predominant formula is Kane's [[61](#page-11-10)]

This report was the beginning of the correction of high refractive defects with two lenses, where the residual correction for the second lens can be left for a second intervention according to the residual defect, and not to the biometrical characteristics.

Due to the possibility of opacifcation and inter-lenticular membrane formation as a result of the implantation of two lenses in the capsular bag, it is recommended to place the greater power IOL in the capsular bag and the second IOL in the ciliary sulcus. Ideally, this lens must be lowpowered, angulated, with rounded edges in order to reduce the risk for iris touch and UGH syndrome or pigment dispersion. The sulcus lens could be implanted on a second surgical time, after the stability of the second postoperatory graduation, so as to increase the refractive success possibilities, even though the additional risks for a new intervention must be considered.

In order to calculate the power of the Piggyback lens, there are different options:

(a) Primary poly-pseudophakia, when the implantation of the IOL will be made both in the capsular bag and in the sulcus in the same act. The total calculation for the value of the lens to be placed in the plane of the capsular bag, according to the ELPo, trying to correct the greatest amount of the defect as possible at this level, according to the lenses available by the various commercial companies. The remaining defect will be corrected by the second lens, which will be implanted at the ciliary sulcus level, adjusting the power due to its more anterior position. When a lens is placed more anteriorly, it requires less power in order to have the same effect, and this reduction is proportional to the power of the intraocular lens. The lens at the sulcus must be adjusted, according to the Holladay 1997 table [\[11](#page-9-8)] for poly-pseudophakia in hypermetropy, in the following way:

- 1. From $+1.00$ to $+8.00$ does not require adjustment.
- 2. From +8.50 to +15.00 subtract 0.5 D to the value of the IOL.
- 3. From +15.5 to +25.5 D subtract 1.0 D to the value of the IOL.
- 4. From +25.5 D to +30.0 D subtract 1.5 D to the value of the IOL.
- (b) The second option is used when the lens will be implanted in another surgical act, once the residual defect is stable. To calculate it, this residual hyperopic defect is multiplied by 1.5, in lower defects smaller than $+6.00$ D $[63]$ $[63]$ $[63]$.

For other cases, it is advisable to use optical vergence formulas which take the keratometry into account, the ELPo for the sulcus, and the residual defect, used as ACD value, the value of the manufacturer −0.65 in order to adapt it to the sulcus.

The new lenses that have surfaced, specifc for placement on the sulcus, and for the correction of residual defects, have specifc optic vergency programs in the web pages for their companies [[64\]](#page-11-14). We are attaching some examples. Sulcoflex: [https://www.ray](https://www.raytrace.rayner.com/)[trace.rayner.com/](https://www.raytrace.rayner.com/), Add-On: [https://](https://www.1stq.de/en/34-addoncalculator) [www.1stq.de/en/34-addoncalculator,](https://www.1stq.de/en/34-addoncalculator) ICL: <https://ocos.starag.ch/>.

High-Power Intraocular Lenses

The ideal in short eyes is to achieve optical correction with a high-power intraocular lens which will allow for poly-pseudophakia in case of an unexpected high residual defect. These are hard to get and sometimes do require special orders.

Even with the highest standards, and lots of care in the biometry, the biometric results may be affected by the variability in tolerance when manufacturing intraocular lenses [[54\]](#page-11-4). In high power ranges of IOLs (>30.0 D) that are usually required in this high hyperopic eyes, the real dioptric power may vary as much as ± 1.0 D, according to the International Organization of Standards [\[65](#page-11-15)].

The three problems with high power lenses are:

- 1. Values for the International Organization for Standardization allowing for a tolerance of ± 1.0 D in IOL at $> +30.0$ D, and ± 0.50 in IOL $\leq +30$ D [[65,](#page-11-15) [66\]](#page-11-16)
- 2. Increase in spherical aberration by increasing the power for the IOL [[64\]](#page-11-14). Important in high power lenses free from spherical aberration such as Aspira-aAY
- 3. The rarity of this cases makes lens manufacturers to lose the appeal for their mass production and therefore there is scarce global availability.

Visual Acuities Obtained

Corrected visual acuities in nanophthalmic eyes tend to be considerably worse in normal eyes, with a range of $+0.55 \log MAR$ to $+0.41 \log MAR$ [\[1](#page-9-0), [3](#page-9-2), [21](#page-9-13), [24](#page-10-0)].

This is in part due to the relative effect of optic minifcation or reduced image magnifcation in comparison with distant correction for glasses or contact lenses in such high hyperopic cases and for amblyopia inherent to the refractive defect.

These possible optic results alongside the risks for complications must be discussed with the patients ahead of time in the preoperatory, as well as the limitations in the calculation prediction for the IOL. Even with these risks, the potential for improvement is signifcant for the quality of life of these nanophthalmos cases, by reducing the preoperatory refractive error and by eliminating dependence in glasses or contact lenses.

Conclusions

The short eye is a phenotypical spectrum for genetic abnormalities, leading to anatomical conditions of the ocular structures, producing not only clinical-pathological consequences, which increase pre, trans, and postoperatory comorbidities in cataract surgery, but they also pose a signifcant challenge for the physician when calculating the intraocular lens. There is a marked variability for the estimate of the effective lens position, even when using the new calculation formulas, with unprecise and unexpected refractive results in some cases.

According to the latest reports, the recommendation is to use multiple formulas and to compare at least three formulas, such as Kane, Olsen, EVO2, Haigis, and Hill RBF v3.0, especially in eyes with lengths shorter than 21.5 mm. Even so, you must explain to the patients the possibility of getting unexpected results, with >40% outside ±0.50 D, and visual acuities, according to preoperative amblyopia and a possible optic effect from the degree of image reduced magnifcation, in the case of prior corrections with glasses or contact lenses.

References

- 1. Lemos JA, Rodrigues P, Resende RA, Menezes C, Goncalves RS, Coelho P. Cataract surgery in patients with nanophthalmos: results and complications. Eur J Ophthalmol. 2016;26(2):103–6.
- 2. Singh OS, Simmons RJ, Brockhurst RJ, Trempe CL. Nanophthalmos: a perspective on identifcation and therapy. Ophthalmology. 1982;89(9):1006–12.
- 3. Jung KI, Yang JW, Lee YC, Kim SY. Cataract surgery in eyes with nanophthalmos and relative anterior microphthalmos. Am J Ophthalmol. 2012;153(6):1161–8.e1.
- 4. Cao X, Hou X, Bao Y. The ocular biometry of adult cataract patients on lifeline express hospital eye-train in rural China. J Ophthalmol. 2015;2015:171564.
- 5. Fotedar R, Wang JJ, Burlutsky G, Morgan IG, Rose K, Wong TY, et al. Distribution of axial length and ocular biometry measured using partial coherence laser interferometry (IOL Master) in an older white population. Ophthalmology. 2010;117(3):417–23.
- 6. Lim LS, Saw SM, Jeganathan VS, Tay WT, Aung T, Tong L, et al. Distribution and determinants of ocular biometric parameters in an Asian population: the

Singapore Malay Eye Study. Invest Ophthalmol Vis Sci. 2010;51(1):103–9.

- 7. Pan CW, Wong TY, Chang L, Lin XY, Lavanya R, Zheng YF, et al. Ocular biometry in an urban Indian population: the Singapore Indian Eye Study (SINDI). Invest Ophthalmol Vis Sci. 2011;52(9):6636–42.
- 8. Shufelt C, Fraser-Bell S, Ying-Lai M, Torres M, Varma R, Los Angeles Latino Eye Study Group. Refractive error, ocular biometry, and lens opalescence in an adult population: the Los Angeles Latino Eye Study. Invest Ophthalmol Vis Sci. 2005;46(12):4450–60.
- 9. Warrier S, Wu HM, Newland HS, Muecke J, Selva D, Aung T, et al. Ocular biometry and determinants of refractive error in rural Myanmar: the Meiktila Eye Study. Br J Ophthalmol. 2008;92(12):1591–4.
- 10. Melles RB, Holladay JT, Chang WJ. Accuracy of intraocular lens calculation formulas. Ophthalmology. 2018;125(2):169–78.
- 11. Holladay JT. Standardizing constants for ultrasonic biometry, keratometry, and intraocular lens power calculations. J Cataract Refract Surg. 1997;23(9):1356–70.
- 12. Auffarth GU, Blum M, Faller U, Tetz MR, Volcker HE. Relative anterior microphthalmos: morphometric analysis and its implications for cataract surgery. Ophthalmology. 2000;107(8):1555–60.
- 13. Hoffman RS, Vasavada AR, Allen QB, Snyder ME, Devgan U, Braga-Mele R, et al. Cataract surgery in the small eye. J Cataract Refract Surg. 2015;41(11):2565–75.
- 14. Khairallah M, Messaoud R, Zaouali S, Ben Yahia S, Ladjimi A, Jenzri S. Posterior segment changes associated with posterior microphthalmos. Ophthalmology. 2002;109(3):569–74.
- 15. Vingolo EM, Steindl K, Forte R, Zompatori L, Iannaccone A, Sciarra A, et al. Autosomal dominant simple microphthalmos. J Med Genet. 1994;31(9):721–5.
- 16. Weiss AH, Kousseff BG, Ross EA, Longbottom J. Simple microphthalmos. Arch Ophthalmol. 1989;107(11):1625–30.
- 17. Foster PJ, Broadway DC, Hayat S, Luben R, Dalzell N, Bingham S, et al. Refractive error, axial length and anterior chamber depth of the eye in British adults: the EPIC-Norfolk Eye Study. Br J Ophthalmol. 2010;94(7):827–30.
- 18. Wu W, Dawson DG, Sugar A, Elner SG, Meyer KA, McKey JB, et al. Cataract surgery in patients with nanophthalmos: results and complications. J Cataract Refract Surg. 2004;30(3):584–90.
- 19. Yuzbasioglu E, Artunay O, Agachan A, Bilen H. Phacoemulsifcation in patients with nanophthalmos. Can J Ophthalmol. 2009;44(5):534–9.
- 20. Faucher A, Hasanee K, Rootman DS. Phacoemulsifcation and intraocular lens implantation in nanophthalmic eyes: report of a medium-size series. J Cataract Refract Surg. 2002;28(5):837–42.
- 21. Day AC, MacLaren RE, Bunce C, Stevens JD, Foster PJ. Outcomes of phacoemulsifcation and intraocular

lens implantation in microphthalmos and nanophthalmos. J Cataract Refract Surg. 2013;39(1):87–96.

- 22. Naujokaitis T, Scharf D, Baur I, Khoramnia R, Auffarth GU. Bilateral implantation of +56 and +58 diopter custom-made intraocular lenses in patient with extreme nanophthalmos. Am J Ophthalmol Case Rep. 2020;20:100963.
- 23. Singh H, Wang JC, Desjardins DC, Baig K, Gagne S, Ahmed II. Refractive results in nanophthalmic eyes after phacoemulsifcation and implantation of a highrefractive-power foldable intraocular lens. J Cataract Refract Surg. 2015;41(11):2394–402.
- 24. Zheng T, Chen Z, Xu J, Tang Y, Fan Q, Lu Y. Outcomes and prognostic factors of cataract surgery in adult extreme microphthalmos with axial length <18 mm or corneal diameter <8 mm. Am J Ophthalmol. 2017;184:84–96.
- 25. 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. Invest Ophthalmol Vis Sci. 2019;60(6):2248–56.
- 26. Jackson TL, Hussain A, Salisbury J, Sherwood R, Sullivan PM, Marshall J. Transscleral albumin diffusion and suprachoroidal albumin concentration in uveal effusion syndrome. Retina. 2012;32(1):177–82.
- 27. Bartke TU, Auffarth GU, Uhl JC, Volcker HE. Reliability of intraocular lens power calculation after cataract surgery in patients with relative anterior microphthalmos. Graefes Arch Clin Exp Ophthalmol. 2000;238(2):138–42.
- 28. Nihalani BR, Jani UD, Vasavada AR, Auffarth GU. Cataract surgery in relative anterior microphthalmos. Ophthalmology. 2005;112(8):1360–7.
- 29. Khan AO. Recognizing posterior microphthalmos. Ophthalmology. 2006;113(4):718.
- 30. Khan AO. Posterior microphthalmos versus nanophthalmos. Ophthalmic Genet. 2008;29(4):189.
- 31. Spitznas M, Gerke E, Bateman JB. Hereditary posterior microphthalmos with papillomacular fold and high hyperopia. Arch Ophthalmol. 1983;101(3):413–7.
- 32. Alkin Z, Ozkaya A, Karakucuk Y, Demirok A. Detailed ophthalmologic evaluation of posterior microphthalmos. Middle East Afr J Ophthalmol. 2014;21(2):186–8.
- 33. Boynton JR, Purnell EW. Bilateral microphthalmos without microcornea associated with unusual papillomacular retinal folds and high hyperopia. Am J Ophthalmol. 1975;79(5):820–6.
- 34. Erdol H, Kola M, Turk A, Akyol N. Ultrasound biomicroscopy and OCT fndings in posterior microphthalmos. Eur J Ophthalmol. 2008;18(3):479–82.
- 35. Kim JW, Boes DA, Kinyoun JL. Optical coherence tomography of bilateral posterior microphthalmos with papillomacular fold and novel features of retinoschisis and dialysis. Am J Ophthalmol. 2004;138(3):480–1.
- 36. Kiratli H, Tumer B, Kadayifcilar S. Bilateral papillomacular retinal folds and posterior microphthal-

mus: new features of a recently established disease. Ophthalmic Genet. 2000;21(3):181–4.

- 37. Kaderli A, Acar MA, Unlu N, Uney GO, Ornek F. The correlation of hyperopia and choroidal thickness, vessel diameter and area. Int Ophthalmol. 2018;38(2):645–53.
- 38. Sundin OH, Leppert GS, Silva ED, Yang JM, Dharmaraj S, Maumenee IH, et al. Extreme hyperopia is the result of null mutations in MFRP, which encodes a frizzled-related protein. Proc Natl Acad Sci U S A. 2005;102(27):9553–8.
- 39. Gal A, Rau I, El Matri L, Kreienkamp HJ, Fehr S, Baklouti K, et al. Autosomal-recessive posterior microphthalmos is caused by mutations in PRSS56, a gene encoding a trypsin-like serine protease. Am J Hum Genet. 2011;88(3):382–90.
- 40. Nair KS, Hmani-Aifa M, Ali Z, Kearney AL, Ben Salem S, Macalinao DG, et al. Alteration of the serine protease PRSS56 causes angle-closure glaucoma in mice and posterior microphthalmia in humans and mice. Nat Genet. 2011;43(6):579–84.
- 41. Awadalla MS, Burdon KP, Souzeau E, Landers J, Hewitt AW, Sharma S, et al. Mutation in TMEM98 in a large white kindred with autosomal dominant nanophthalmos linked to 17p12-q12. JAMA Ophthalmol. 2014;132(8):970–7.
- 42. Khorram D, Choi M, Roos BR, Stone EM, Kopel T, Allen R, et al. Novel TMEM98 mutations in pedigrees with autosomal dominant nanophthalmos. Mol Vis. 2015;21:1017–23.
- 43. Tedja MS, Wojciechowski R, Hysi PG, Eriksson N, Furlotte NA, Verhoeven VJM, et al. Genome-wide association meta-analysis highlights light-induced signaling as a driver for refractive error. Nat Genet. 2018;50(6):834–48.
- 44. Siggs OM, Awadalla MS, Souzeau E, Stafferi SE, Kearns LS, Laurie K, et al. The genetic and clinical landscape of nanophthalmos and posterior microphthalmos in an Australian cohort. Clin Genet. 2020;97(5):764–9.
- 45. Kane JX, Melles RB. Intraocular lens formula comparison in axial hyperopia with a high-power intraocular lens of 30 or more diopters. J Cataract Refract Surg. 2020;46(9):1236–9.
- 46. Goto S, Maeda N, Koh S, Ohnuma K, Hayashi K, Iehisa I, et al. Prediction of postoperative intraocular lens position with angle-to-angle depth using anterior segment optical coherence tomography. Ophthalmology. 2016;123(12):2474–80.
- 47. Cooke DL, Cooke TL. Approximating sum-ofsegments axial length from a traditional optical lowcoherence refectometry measurement. J Cataract Refract Surg. 2019;45(3):351–4.
- 48. Cooke DL, Cooke TL. A comparison of two methods to calculate axial length. J Cataract Refract Surg. 2019;45(3):284–92.
- 49. Cooke DL, Cooke TL, Suheimat M, Atchison DA. Standardizing sum-of-segments axial length using refractive index models. Biomed Opt Express. 2020;11(10):5860–70.
- 50. Wang L, Cao D, Weikert MP, Koch DD. Calculation of axial length using a single group refractive index versus using different refractive indices for each ocular segment: theoretical study and refractive outcomes. Ophthalmology. 2019;126(5): 663–70.
- 51. Holladay JT, Gills JP, Leidlein J, Cherchio M. Achieving emmetropia in extremely short eyes with two piggyback posterior chamber intraocular lenses. Ophthalmology. 1996;103(7):1118–23.
- 52. Gokce SE, Zeiter JH, Weikert MP, Koch DD, Hill W, Wang L. Intraocular lens power calculations in short eyes using 7 formulas. J Cataract Refract Surg. 2017;43(7):892–7.
- 53. Carif G, Aiello F, Zygoura V, Kopsachilis N, Maurino V. Accuracy of the refractive prediction determined by multiple currently available intraocular lens power calculation formulas in small eyes. Am J Ophthalmol. 2015;159(3):577–83.
- 54. Eom Y, Kang SY, Song JS, Kim YY, Kim HM. Comparison of Hoffer Q and Haigis formulae for intraocular lens power calculation according to the anterior chamber depth in short eyes. Am J Ophthalmol. 2014;157(4):818–24.e2.
- 55. Sudhakar S, Hill DC, King TS, Scott IU, Mishra G, Ernst BB, et al. Intraoperative aberrometry versus preoperative biometry for intraocular lens power selection in short eyes. J Cataract Refract Surg. 2019;45(6):719–24.
- 56. Kane JX, Van Heerden A, Atik A, Petsoglou C. Intraocular lens power formula accuracy: comparison of 7 formulas. J Cataract Refract Surg. 2016;42(10):1490–500.
- 57. Shrivastava AK, Behera P, Kumar B, Nanda S. Precision of intraocular lens power prediction in

eyes shorter than 22 mm: an analysis of 6 formulas. J Cataract Refract Surg. 2018;44(11):1317–20.

- 58. Wang Q, Jiang W, Lin T, Wu X, Lin H, Chen W. Metaanalysis of accuracy of intraocular lens power calculation formulas in short eyes. Clin Exp Ophthalmol. 2018;46(4):356–63.
- 59. Melles RB, Kane JX, Olsen T, Chang WJ. Update on intraocular lens calculation formulas. Ophthalmology. 2019;126(9):1334–5.
- 60. Hipolito-Fernandes D, Elisa Luis M, Gil P, Maduro V, Feijao J, Yeo TK, et al. VRF-G, a new intraocular lens power calculation formula: a 13-formulas comparison study. Clin Ophthalmol. 2020;14:4395–402.
- 61. Kane JX, Chang DF. Intraocular lens power formulas, biometry, and intraoperative aberrometry: a review. Ophthalmology. 2020;128:e94.
- 62. Gayton JL, Sanders VN. Implanting two posterior chamber intraocular lenses in a case of microphthalmos. J Cataract Refract Surg. 1993;19(6):776–7.
- 63. Levinger E, Mimouni M, Finkelman Y, Yatziv Y, Shahar J, Trivizki O. Outcomes of refractive error correction in pseudophakic patients using a sulcus piggyback intraocular lens. Eur J Ophthalmol. 2021;31(2):422.
- 64. Barbero S, Marcos S, Jimenez-Alfaro I. Optical aberrations of intraocular lenses measured in vivo and in vitro. J Opt Soc Am A Opt Image Sci Vis. 2003;20(10):1841–51.
- 65. Hoffer KJ, Calogero D, Faaland RW, Ilev IK. Testing the dioptric power accuracy of exact-powerlabeled intraocular lenses. J Cataract Refract Surg. 2009;35(11):1995–9.
- 66. Hoffer KJ, Savini G. IOL power calculation in short and long eyes. Asia Pac J Ophthalmol (Phila). 2017;6(4):330–1.

Open Access This chapter is licensed under the terms of the Creative Commons Attribution 4.0 International License ([http://creativecommons.org/licenses/by/4.0/\)](http://creativecommons.org/licenses/by/4.0/), which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons license and indicate if changes were made.

The images or other third party material in this chapter are included in the chapter's Creative Commons license, unless indicated otherwise in a credit line to the material. If material is not included in the chapter's Creative Commons license and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder.

