Bilateral implantation of +56 and +58 diopter custom-made intraocular lenses in patient with extreme nanophthalmos

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A B S T R A C T

Purpose: To present the case of a 60-year-old patient with severe nanophthalmic eyes, who underwent cataract surgery with a bilateral implantation of custom-made high-power intraocular lenses (IOLs).
Observations: The axial length was 14.94 and 15.05 mm of the right and the left eye, respectively. The preoperative corrected distance visual acuity (CDVA) was +0.46 logMAR (20/63) in the right eye and +0.58 logMAR (20/80) in the left eye with rigid contact lenses of +17.5 D bilaterally. The calculated IOL power for emmetropia with different formulas ranged from +55.28 to +70.09 D. The IOL power selection was based on the average value from four formulas (Haigis, Holladay 1, Holladay 2, SRK/T) with the target refraction of emmetropia. Custom-made +56.0 and + 58.0 D Aspira-aAY IOLs (HumanOptics AG, Erlangen, Germany) were implanted without any complications. The postoperative CDVA was +0.40 logMAR (20/50) and +0.60 logMAR (20/80). The manifest refraction spherical equivalents were +0.625 D and −0.375 D.
Conclusions and importance: Even in eyes with the axial length of only 15 mm, cataract surgery can be successfully performed after adequate preparation. High-power customized IOLs allow complete correction of hyperopia but caution is required with the results from different IOL power calculation formulas, which can be misleading.

1. Introduction

The term nanophthalmos refers to a proportionally small eye without malformations. It is a rare congenital condition and the current definition mainly relies on axial length (AL).[1] Most recent studies define the eyes with an AL of <20.0 mm or <20.5 mm as nanophthalmic.[1–5] In contrast, the eyes with normal AL but disproportionally small anterior segment are described by the term relative anterior microphthalmos.[6,7] In both nanophthalmos and relative anterior microphthalmos, cataract surgery is difficult due to the small anterior segment.

Early attempts of cataract surgery in nanophthalmic eyes were associated with a high risk of complications and poor outcomes.[8] Although modern surgical techniques allow a safer procedure with better outcomes, cataract surgery in nanophthalmic eyes remains challenging.[3] As the current intraocular lens (IOL) power calculation formulas are considered less accurate in nanophthalmic eyes, the IOL power selection presents a problem for the surgeon.[2] In addition, patients with nanophthalmos require high-power IOLs that are not mass produced. In Germany, it is possible to order customized high-power

IOLs directly from manufacturers. In a lot of other countries, however, only IOLs with powers up to +30.0 to 35.0 D are available. Another challenge is the cataract surgery itself. It is more difficult to perform, carries a higher risk of complications and the visual prognosis is markedly worse, compared to normal eyes.[2–4] We present a case of a patient with extremely short eyes, who underwent cataract surgery with a bilateral implantation of custom-made high-power IOLs.

2. Case report

The 60-year-old male patient with bilateral nanophthalmos was referred for the first time to our clinic for cataract surgery in 2014. The patient had been wearing rigid contact lenses since the age of 14 because of high hyperopia. Before that, spectacles had been used since the age of 5. According to the patient, the vision had never been good. At that time, in 2014, the patient had cataracts but, on being informed of his options, decided to postpone the surgery.

Five years later, the patient presented to our clinic complaining of increasing glare sensitivity. CDVA with rigid contact lenses of +17.5 D

was +0.46 logMAR (20/63) in the right eye and +0.58 logMAR (20/80) in the left eye. No improvement could be achieved with additional lenses. IOP was 12 mm Hg in both eyes. Slit lamp examination of the anterior segment revealed progression of cataracts (Fig. 1). Fundoscopy and optical coherence tomography findings were unremarkable.

After performing biometry measurements with IOLMaster 700 (Carl Zeiss Meditec, Jena, Germany), the IOL power for emmetropia for the foldable acrylic Aspira-aAY IOL (HumanOptics AG, Erlangen, Germany) was calculated with different formulas. The suggested power to achieve emmetropia ranged from +55.28 to +70.09 D. Based on our past experience of selecting high-power IOLs, we had a certain preference for the Haigis formula in achieving a reliable result but we tended to choose the average value taken from four formulas (Haigis, Holladay 1, Holladay 2, SRK/T). The IOL power of +56.0 D was selected for the right eye and one of +58.0 D for the left eye. The biometry values and IOL power calculation results are presented in Table 1.

With the patient under general anesthesia, both surgeries were performed by an experienced surgeon (GUA). LenSx® femtosecond laser (Alcon Laboratories, Inc., Fort Worth, TX, USA) was used for capsulotomy and lens fragmentation. The docking of the patient interface was more complicated, due to the nanophthalmos, but was completed successfully. Considerable artefacts were visible in the LenSx® optical coherence tomography images so the automatic detection of structures had to be manually overridden (Fig. 2). The femtosecond laser was in this case capable of creating only a partial capsulotomy and it was safely completed with a manual technique. However, it should be noted that incomplete capsulotomy, if unrecognized, could have resulted in a radial tear of the capsule, potentially jeopardizing the implantation of the IOL in the capsular bag. The nucleus fragmentation was also only partial. We applied the "pizza cut" pattern but only a few lines could be seen intraoperatively. Despite these technical difficulties, a round capsulotomy could be achieved, which we think is advantageous when implanting a high power IOL (+56.0 and + 58.0 D) into the capsular bag. A temporal position for the main incision was chosen in order to facilitate access. The incisions were created manually since we do not use the femtosecond laser for the corneal incisions in our clinic, based on our past experiences. The IOL was top-loaded using forceps into the single-use syringe-type ACCUJECT™ IOL injection system (Medicel AG,

Table 1

Biometry data, IOL power calculation results, and prediction error using Hoffer Q, Haigis, SRK/T, Holladay 1 and Holladay 2 formulas.

Biometry Data		
Parameter	Right Eye	Left Eye
AL	14.94 mm	15.05 mm
R	6.70 mm	7.04 mm
R1	6.75 mm @ 132°	7.10 mm @ 64°
R2	6.65 mm @ 42°	6.97 mm @ 154°
WTW	11.5 mm	11.3 mm
ACD	2.24 mm	2.33 mm
LT	5.96 mm	5.90 mm
Calculated IOL Power for Emmetropia (Aspira-aAY)		
Formula	Right Eye	Left Eye
Hoffer Q	+70.09 D	+69.96 D
Haigis	+55.28 D	+57.47 D
SRK/T	+56.04 D	+57.09 D
Holladay 1	+57.07 D	+59.20 D
Holladay 2	+57.43 D	+59.05 D
Prediction Error (Postoperative Spherical Equivalent - Target Refraction)		
Formula	Right Eye	Left Eye
Hoffer Q	−7.57 D	−7.75 D
Haigis	+1.21 D	+0.06 D
SRK/T	+0.60 D	+0.34 D
Holladay 1	−0.19 D	−1.29 D
Holladay 2	−0.45 D	−1.18 D

ACD: anterior chamber depth, AL: axial length, IOL: intraocular lens, LT: lens thickness, R: corneal radius, WTW: white-to-white distance.

Altenhein, Switzerland), which has the cartridge integrated in the injector. Due to the thickness of the IOL, the 3.0–1P version of the injector was chosen and the incision size of slightly more than 3.0 mm was used (Fig. 3, Fig. 4). Despite small dimensions of the eyes, both surgeries were uneventful.

On the first day postoperatively, CDVA in the right eye was +0.38 logMAR (20/50) with the manifest refraction of +0.50 –1.00 × 80°. CDVA in the left eye was +0.64 logMAR (20/80) with the manifest refraction of −0.50 –0.50 × 65°. The early postoperative period was complication-free. Two months after the surgery, CDVA was +0.40 logMAR (20/50) and +0.60 logMAR (20/80) with the manifest refraction of +1.00 –0.75 × 135° and +0.50 –1.75 × 45° in the right and the
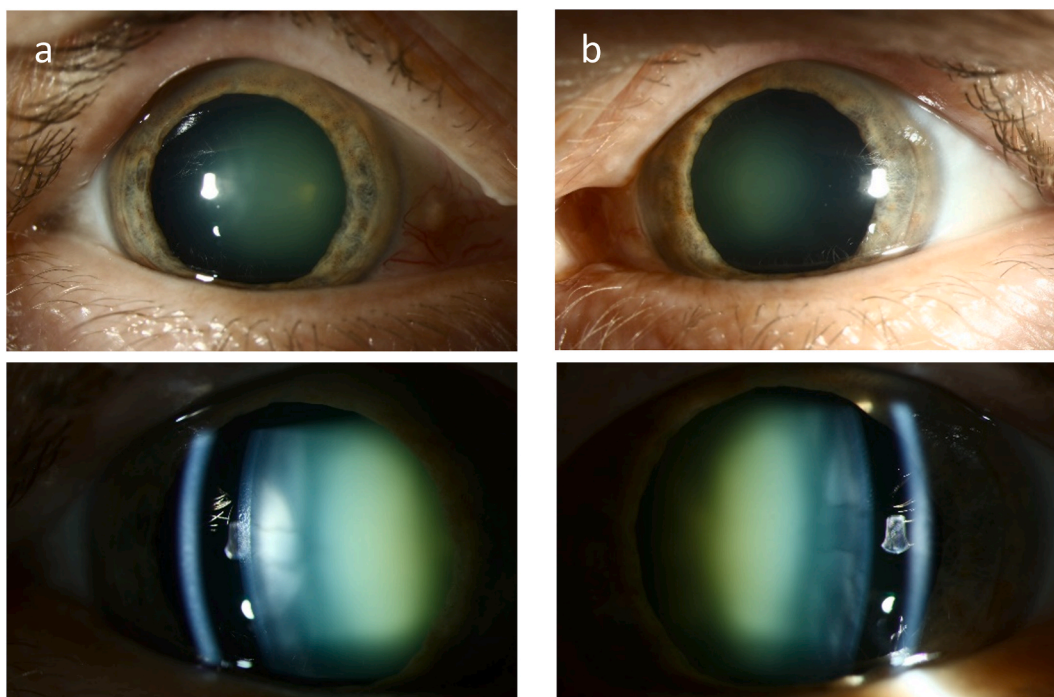

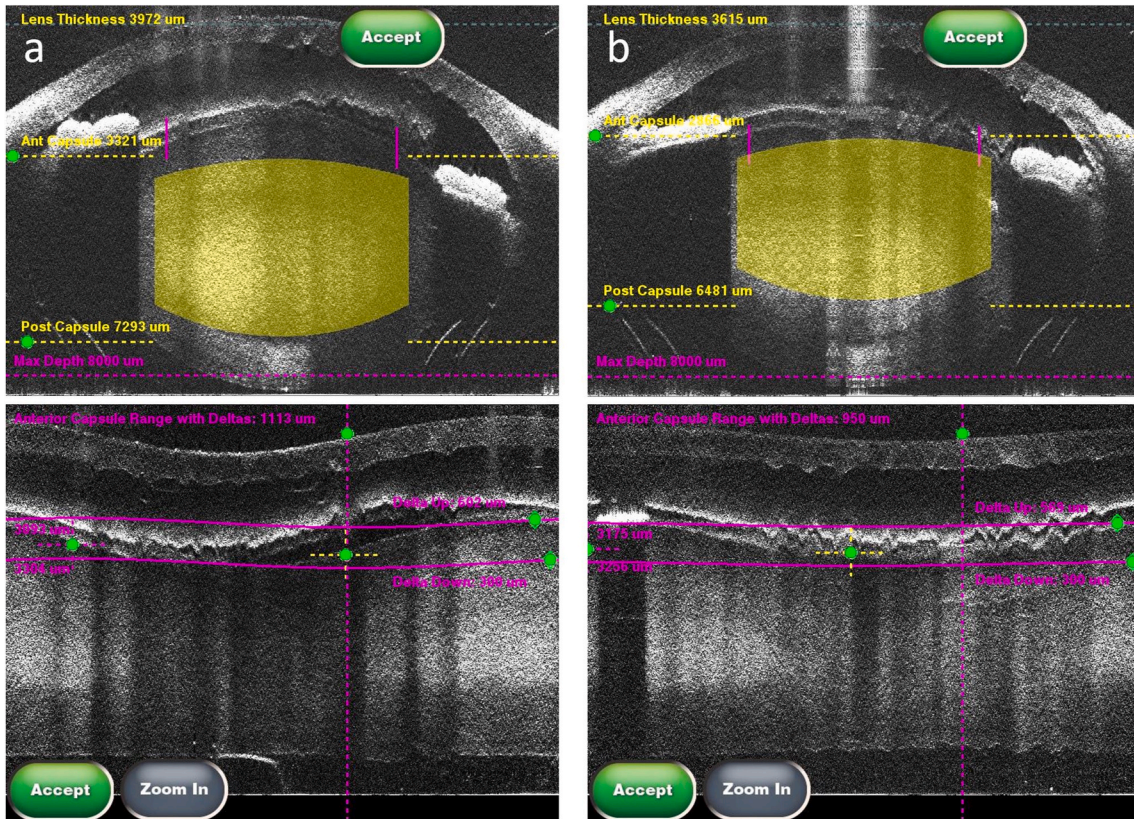**Fig. 1.** Preoperative images of the right (a) and the left (b) eye.

Fig. 2. Optical coherence tomography images of the right (a) and the left (b) eye from the LenSx® femtosecond laser.

Fig. 3. Aspira-aAY intraocular lens (IOL) of +56.0 D before loading it into the cartridge.

left eye, respectively. Spherical equivalent refractions were +0.625 D and −0.375 D in the right and the left eye, respectively. The difference from target refraction was the smallest with Holladay 1 formula for the right eye (−0.19 D) and Haigis formula for the left eye (+0.06 D). The IOP was 12 mmHg in the right eye and 14 mmHg in the left eye. The slit lamp examination revealed slight posterior capsule opacification in both eyes, currently not requiring treatment.

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3. Discussion

Cataract surgery in nanophthalmic eyes presents several challenges. The postoperative refraction is difficult to predict as IOL power calculation is less accurate in short eyes.[9] Several issues exist concerning the manufacturing and availability of high-power IOLs. The surgery itself is problematic because of the eye's reduced anatomical size and it carries a high risk of complications.[2,4] In addition, patient expectations need to be managed since the visual outcome tends to be worse than in routine
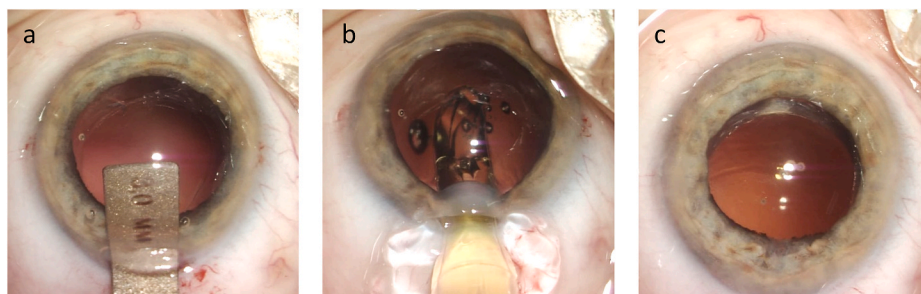

Fig. 4. Intraoperative images of the left eye: incision size measurement before IOL implantation (a), injection of +58.0 D Aspira-aAY IOL (b), and the eye after the IOL implantation (c).

cataract surgery.[4]

The prediction of postoperative refraction in nanophthalmic eyes is considerably less accurate than in normal eyes. Jung et al. compared the refractive outcomes in nanophthalmic eyes to the outcomes in a normal control group. While 90%–98% of normal eyes achieved a refraction within ±1.00 D, this was the case in only 46%–66% of nanophthalmic eyes with AL <20.5 mm.[2] The prediction error increases with decreasing AL and this is in part due to the high optical power of IOLs implanted in such eyes, giving more weight to errors in the prediction of IOL position.[10,11] In our patient, the AL was extremely short and the power of IOLs implanted was particularly high, presenting a risk of significant postoperative refractive error.

Older third-generation formulas (Hoffer Q, Holladay 1, and SRK/T) use AL and corneal power to calculate effective lens position (ELP), while newer fourth-generation formulas such as Haigis and Holladay 2 and fifth-generation formulas such as Barrett Universal II include additional parameters to calculate ELP.[12] These newer formulas require the direct measurement of the anterior chamber depth (ACD) and therefore do not solely depend on assumptions.[10,13] In studies of short eyes, Haigis, Hoffer Q, Holladay 1 and Holladay 2 formulas were reported to be more accurate than SRK/T formula.[10,13,14] A study by Eom et al. compared Hoffer Q and Haigis formulas in short eyes and found Haigis formula to perform better in eyes with shallow anterior chamber (ACD <2.40 mm).[9] A meta-analysis by Wang et al. included 10 studies with 1161 short eyes in total and reported the superiority of Haigis formula over Hoffer Q and SRK/T formulas. However, there was not enough data available to evaluate the accuracy of Holladay 1 and Holladay 2 formulas.[15] The studies that included Barrett Universal II formula found no differences in accuracy of predicting the IOL power when comparing it with other formulas in short eyes.[11,12,18]

It should be noted that the mean AL in studies evaluating different IOL power calculation formulas in short eyes ranges from 19.53 mm to 21.69 mm.[2,9,10,12,14–18] However, there is limited data available regarding the IOL power calculation in extremely short eyes. In a study of 11 eyes with the mean AL of 16.4 mm (simple microphthalmos group), the only formula used was Hoffer Q and the mean absolute postoperative refractive error was 5.6 D. However, as this is an absolute value, it is not clear if there was a tendency toward myopia or hyperopia.[4] Another study of nanophthalmic eyes used a proprietary IOL calculation algorithm by Carl Zeiss Meditec AG which in most cases resulted in hyperopic outcomes.[1]

In our case, Haigis, Holladay 1, Holladay 2 and SRK/T formulas delivered similar results, with the prediction error ranging from −1.29 D to +1.21 D. The predictions from Haigis and SRK/T formulas were slightly hyperopic, while Holladay 1 and Holladay 2 formulas delivered slightly myopic results. In contrast, Hoffer Q formula returned highly myopic results (−7.57 D and −7.75 D for the right and the left eye, respectively). Barrett Universal II formula could not be used in this case of extreme nanophthalmos since the biometry values were out of the acceptable range for the formula and therefore an error message was displayed in the official calculator.

Several issues exist regarding the manufacturing of high-power IOLs. First of all, the International Organization for Standardization allows the tolerance of ±1.0 D for the IOLs of >30.0 D, while the tolerance for lower-powered IOLs is ±0.5 D or even less.[11,19] This is a concern when trying to achieve a good refractive outcome, especially as the IOL calculation is already less accurate in very short eyes.[2,9] Another issue is the increase of spherical aberration with increasing IOL power.[20] Some manufacturers now offer aspheric aberration-free IOLs, such as the Aspira-aAY implanted in our patient, which may be advantageous in these cases. The third issue is the rarity of cases when high-power IOLs are needed, which makes the production of such IOLs less attractive from a manufacturer's perspective. This results in limited availability of high-power IOLs.

In cases when high-power IOLs are not available, piggyback IOLs are an option to minimize the postoperative refractive error in nanophthalmic eyes. As the primary implantation of two IOLs in the capsular bag can result in interlenticular membranes and opacifications, late hyperopic shift and reduction of visual acuity, it is recommended to implant one IOL in the capsular bag and the other IOL in the ciliary sulcus.[21] In order to avoid the sulcus IOL scraping the posterior surface of the iris with the resulting pigment dispersion, the selection of a sulcus IOL with angulated haptics and a rounded edge has been suggested.[21] A potential benefit of piggyback IOLs is that the lower-powered sulcus IOL can be implanted later as a secondary procedure. The postoperative refraction, once stable, can be used to calculate the power of the sulcus IOL and potentially improve the refractive outcome.[21] However, the risks of an additional surgery also need to be considered.

The difficulty of IOL calculation in nanophthalmic eyes is matched by the cataract surgery itself. In 1982, Singh et al. reported "an extremely high complication rate with disastrous results" of intraocular surgery in nanophthalmic eyes.[8] Although the modern cataract surgery technique is safer, complications still occur often in nanophthalmic surgery and include posterior capsule rupture, vitreous loss, suprachoroidal hemorrhage, iris prolapse, iritis, persistent corneal edema, cystoid macular edema and phthisis.[2,4,22,23] Uveal effusion is another well-known complication in nanophthalmic eyes, which can lead to secondary retinal detachment, vitreous hemorrhage, malignant glaucoma and loss of the eye.[4,24–26] This latter complication should be considered when choosing the type of anesthesia because retrobulbar and peribulbar anesthesia increase posterior pressure that may lead to vortex vein congestion. Instead, topical or general anesthesia is preferred.[21,27] Small ocular dimensions and suboptimal access are other aspects to be considered. In our case, the docking of the femtosecond laser interface was more difficult than usual and only a partial capsulotomy was created. The nucleus fragmentation was also only partial. The surgeries were performed from temporal position to facilitate access and were complication-free. Although the incidence of glaucoma following a cataract surgery in nanophthalmic eyes is high, raised IOP was not observed in our patient.[21] During the postoperative follow-up, ultrasound biomicroscopy is a useful technique to evaluate the status and the proximities of the capsular bag, the IOL and the iris, although it has not been performed in our case yet.

Visual outcomes after cataract surgery in nanophthalmic eyes are considerably worse than in normal eyes. Studies report the mean postoperative CDVA to range from +0.55 logMAR to +0.41 logMAR.[2–5] The residual refractive error is also often high because of the limited accuracy of IOL power calculations. In our case, the CDVA was similar to the mean values reported by other authors. The postoperative CDVA remained similar to the preoperative values but the manifest refraction spherical equivalent was reduced from +17.5 D preoperatively to less than ±1.0 D postoperatively. Since in our case the preoperative hyperopic refractive error was corrected with rigid contact lenses and not with spectacles, the less-than-expected postoperative CDVA cannot be solely attributed to the effect of relative optical minification. Given the large preoperative hyperopic refractive error, the presence of bilateral ametropic amblyopia cannot be ruled out, which could have also contributed to the lack of improvement in CDVA.

The increased risk of complications and worse visual prognosis should be discussed with patients before the surgery. They should also understand the limitations of the IOL power prediction accuracy and be ready to wear spectacles to correct residual refractive error. Even though an improvement in visual acuity is not always achieved, cataract surgery in nanophthalmic eyes still has the potential to significantly improve the quality of life. By reducing the preoperative refractive error, it enables patients to be less dependent on spectacles and contact lenses.

4. Conclusions

Cataract surgery in nanophthalmic eyes is challenging but can be successfully performed after adequate preparation. In our case, even though the AL was extremely short, no complications occurred and the

prediction of postoperative refraction was relatively accurate.

Patient consent

The patient gave verbal consent to publication of the case. This report does not contain any information that could lead to identification of the patient. Retrospective review of this case was done in accordance with the Declaration of Helsinki.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

No conflict of interest (TN, DS, IB, RK, GUA).

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