



A Case of Microspherophakia and Congenital Glaucoma with a Touch of High Myopia

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Background

Microspherophakia (MSP) is a rare bilateral condition in which the crystalline lens assumes a spherical shape with an increased anteroposterior diameter and reduced equatorial diameter. It can occur as an isolated anomaly that is familial or associated with a systemic disease such as Weill-Marchesani syndrome and Marfans syndrome. Other ocular manifestations of MSP include glaucoma, shallow anterior chamber, and high myopia.

Case Presentation

19-year-old female presents with constant blurry vision OU at all distances. She currently wears single vision glasses but due to her high myopia, she is interested in being fit with contact lenses in order to obtain clearer acuity. She was diagnosed with MSP and congenital glaucoma at the age of five and is currently being managed with topical medication. She also has a history of bilateral narrow angles, which have been treated in the past with laser peripheral iridotomies and are currently patent and stable.

Medical History

Heart defect: surgically repaired shortly after birth

Ocular History

High myopia, narrow angles, elevated pressure, congenital glaucoma, MSP

Medications

Cosopt BID, Latanoprost QHS

Exam Findings

Entering VA's (cc): OD 20/30⁻¹ PHNI OS 20/40⁻¹ PHNI

Entrance testing: Unremarkable

Lensometry:

- OD: -23.00-1.00x180
- OS: -22.00-1.00x180

Slit Lamp Exam: See table 1

OD		OS
Clear	Adnexa/Lids/Lashes	Clear
All layers clear	Cornea	All layers clear
Clear/White and Quiet	Conjunctiva/Sclera	Clear/White and Quiet
Superior patent LPI	Iris	Superior patent LPI
Shallow (1.6 mm)	Anterior Chamber	Shallow (0.58 mm)
Grade 2 nasal and temporal	Angles	Closed nasally, grade 1 temporally



Figure 1 (07/2019): Note the shallow anterior chambers and narrow angles causing our patient to experience elevated IOP. The left eye, shown in the image on the right, is significantly more shallow than the right eye.

References

- Chan, RTY., et al. Microspherophakia. *Clin Exp Optom* 2002; 85: 5: 294-299.
- Senthil, S., et al. Glaucoma in Microspherophakia: Presenting Features and Treatment Outcomes. *J Glaucoma* 2014; 23: 4: 262-267
- Bitar, M., et al. Challenges in Diagnosing Microspherophakia in a Pediatric Patient. *JSM Ophthalmol* 2016; 4 (1): 1040
- Muralidhar, R., et al. Visual Outcome and Incidence of Glaucoma in Patients with Microspherophakia. *Eye* 2015; 29: 350-355

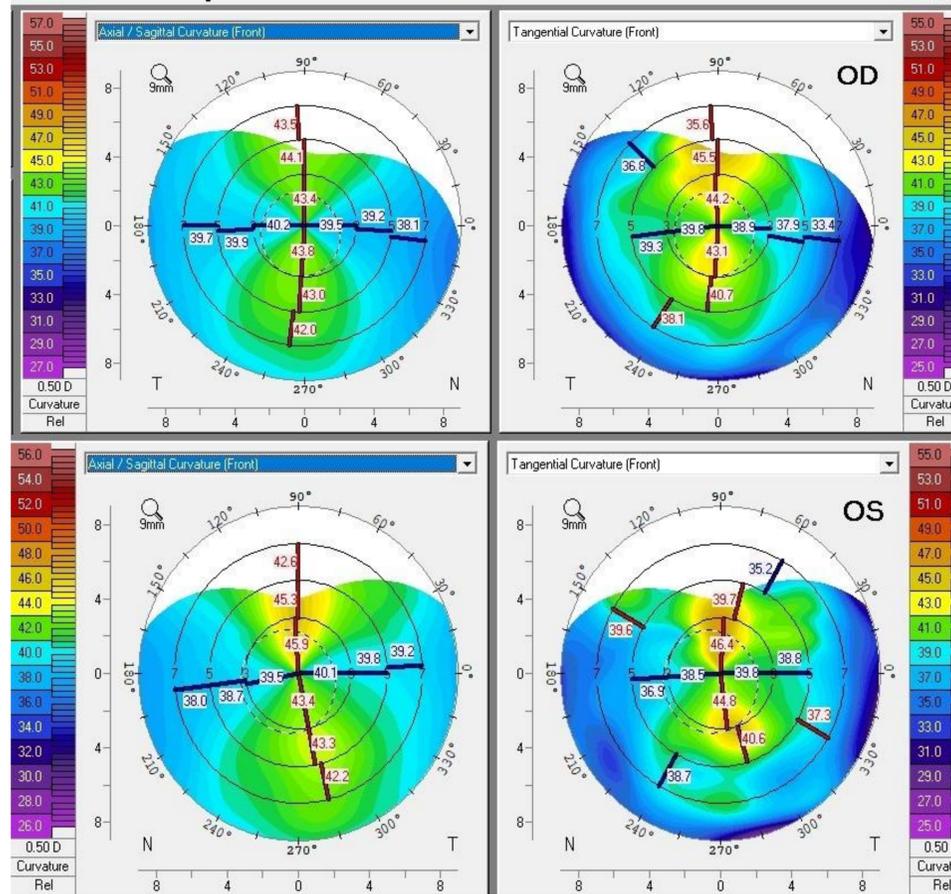


Figure 2 (07/2019): Corneal topography maps. Note regular WTR astigmatism OD and OS, ruling out any corneal ectasias. Confirmed with poster elevation maps not shown above. Keratometry values resulted as the following: **OD 39.8/43.6@86.4 OS 39.7/44.4@95.3**

Case Continued

After obtaining corneal topography, keratometry results revealed corneal astigmatism greater than 2.50 diopters. Due to this finding, gas permeable lenses were empirically ordered in the form of bitoric lenses. Table 2 contains the initial lens parameters ordered for the patient. Patient scheduled to RTC when lenses arrive for a dispense.

	Manufacturer	Brand	Base Curve	Diameter	Power	Notes
OD	Art Optical	GP Bitoric	8.49/7.91	9.6	-18.00/-19.00	(+) lenticulation
OS	Art Optical	GP Bitoric	8.49/7.78	9.6	-17.50/-18.50	(+) lenticulation

Initial Dispense Appointment (7/2019)

Patient presented to clinic for a contact lens dispense with no changes noted in vision or health. After cleaning and rinsing, lenses were inserted and the following was observed behind the slit lamp:

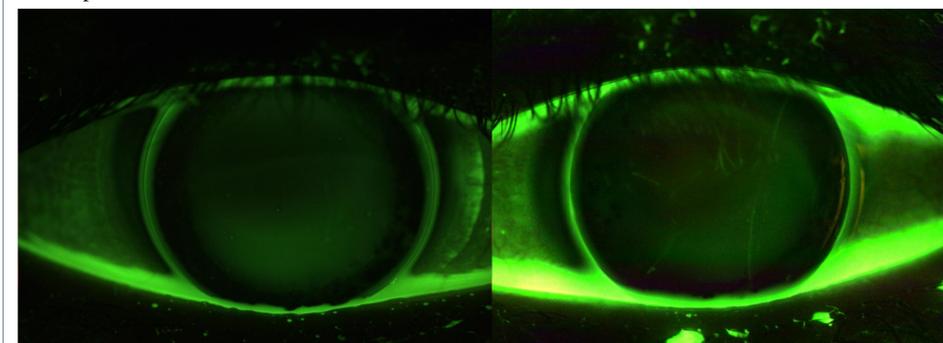


Figure 3: Note moderate apical pooling with moderate midperipheral bearing OD and OS. OD presents with moderate edge lift and minimal edge lift noted in OS. Both lenses rested slightly superior with acceptable movement. Vision was rated a 7/10 while comfort was rated 5/10.

Initial Dispense Continued

Over-refraction was obtained after fit assessment was complete and the results are shown below in table 3 along with the vision results.

	OD	OS
	-0.50-0.50x143	20/30-1
	-0.75-1.50x165	20/20-3

With these over-refraction results and the fit assessment, new lenses were ordered through the lab and the following parameters were obtained (table 3):

	Manufacturer	Brand	Base Curve	Diameter	Power	Notes
OD	Art Optical	GP Bitoric	8.54/7.96	9.6	-18.50/-19.50	(+) lenticulation
OS	Art Optical	GP Bitoric	8.54/7.82	9.6	-18.00/-20.50	(+) lenticulation

Patient went through insertion and removal training and was taught and educated about proper care of the lenses. Patient was sent home with a sample of Boston Simplus cleaning solution. Patient was also instructed to apply her glaucoma drops 5 minutes prior to inserting her lenses. Patient told to begin with 2 hours of wear time and increase each day to allow for adaptation. Patient understood and scheduled to RTC for a contact lens check and dispense of new ordered lenses.

Contact lens check and second dispense (8/2019)

Patient presented to clinic for a contact lens check and second lens dispense. Patient reported no changes in vision from previous visit and states that comfort of lenses has mildly improved. Patient states that she has been compliant with cleaning of lenses and overall care. Patient reports an average wear time of 4 hours and has worn the lenses for 7 hours prior to her appointment. After assessment of initial lenses was complete and new lenses were cleaned and rinsed, new lenses were inserted. The following was observed behind the slit lamp:

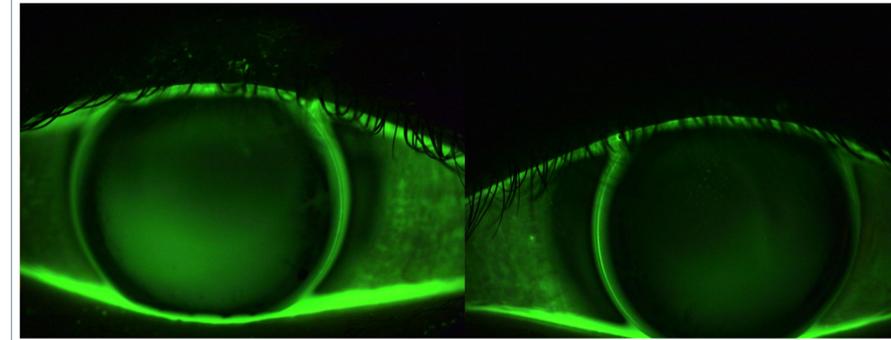


Figure 4: After flattening the base curve of the lenses by 0.25D, fluorescein pattern of the second set of lenses was similar to the initial lenses, however, the patient did notice an improvement in vision with the change in power. Note the foreign body tracking of the left eye (right image) has resolved.

Contact lens check and second dispense continued

New lenses were dispensed to patient and initial lenses were returned to the lab. The patient rated both vision and comfort 8/10 for the new lenses. Patient is scheduled to return for a final contact lens follow up and a dilated eye exam.

Conclusion

The cause of MSP is unknown, however, it is theorized that underdeveloped zonules of Zinn do not exert enough force on the lens to form the typical oval shape. Due to zonular instability, the lens can become subluxated and lead to secondary angle closure glaucoma which is the primary cause of permanent vision loss in MSP. Studies have reported up to 89% of individuals with glaucoma secondary to MSP being 30 years of age or younger with the prevalence increasing with age. Treatment of MSP mainly consists of the management of intraocular pressure to prevent visual complications. It has been described that a PI may be beneficial in the presence of closed angles with lens subluxation or rise in IOP secondary to pupillary block. The limited success of laser iridotomy in MSP, however, shows that pupillary block is not the only mechanism leading to IOP rise. The alternative treatment for these patients consist of surgical management of glaucoma by lensectomy, trabeculectomy, or shunt surgeries. A few reports have shown that lens extraction alone is sufficient to manage glaucoma in MSP.