Unique Contact Lens Fitting Challenges in Microphthalmia

Resident’s Day, American Academy of Optometry, 2016
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ABSTRACT
A 28 year old male with microphthalmia presents for a contact lens fitting. The multiple lens trials attempted before achieving a successful fit illustrate the specific challenges in fitting lenses on microphthalmic eyes.

CASE HISTORY
- Patient demographics
  o 28 year old Hispanic male
- Chief complaint
  o Longstanding blurred vision in the left eye, not improved with glasses
  o Referred to the Therapeutic Contact Lens Clinic at Pacific University for a contact lens fitting
- Ocular history
  o OD: anophthalmia, currently has prosthesis
  o OS: microphthalmia, aniridia
  o Congenital nystagmus
- Medical history
  o Unremarkable systemic history
- Medications
  o Multivitamins
- Family ocular history
  o Older brother: anophthalmia, OU
  o Younger sister: normal ocular health

PERTINENT FINDINGS
- Clinical/physical
  o Visual acuity (Snellen): Uncorrected NLP OD; 20/100 OS
    ▪ VA with habitual spectacles: no improvement; frames obstructed vision with gaze in the direction of null point
  o Pupils: n/a due to prosthesis OD and aniridia OS
  o EOMs: grossly full OS; nystagmus with null point in far right gaze
  o CF: difficult to assess due to poor fixation
  o IOP: 21mmHg OS (tonopen)
  o Biomicroscopy: OD prosthesis clean and in place, flakes in lashes; OS aniridia, shallow anterior chamber, pigment on posterior lens capsule
  o Posterior segment evaluation: OS staphyloma, retinal coloboma, peripheral retinal atrophy
  o Corneal topography (Medmont): OS simulated K values: 37.2/55.5@088; high irregular astigmatism
  o Horizontal visible iris diameter OS: 7.9mm
DIFFERENTIAL DIAGNOSIS

- Primary: Microphthalmia
  - The diagnosis of microphthalmia was previously made in this case but can be confirmed by measuring an axial length of less than 20mm via ultrasound or CT scan (1). Although not performed on this patient, the appearance of the globe is significantly smaller than average, and the retinal findings match those often associated with microphthalmia.

- Others:
  - Microcornea without microphthalmia: corneal diameter less than 11.0mm but with a normal sized globe (2)
  - Synophthalmia: congenital defect involving the fusion of both orbits during development, resulting in one central eye (1)
    - This patient was born with two separate orbits, with the left globe forming smaller than average and the right not forming at all.

DIAGNOSIS AND DISCUSSION

Microphthalmia and anophthalmia appear to present along a continuum, with anophthalmia at the severe end of the spectrum. Many etiologies can contribute to the development of these conditions, including genetic mutations and environmental insults during fetal development (1). They are often associated with other ocular findings, most commonly anterior and posterior segment colobomas (3). The definition of these conditions accepted by the International Clearinghouse for Birth Defects Monitoring Systems is as follows (1):

- Microphthalmia: congenitally small eye with axial length less than 20.0mm and corneal diameter less than 10.0mm
- Anophthalmia: absence of ocular tissue

Approximately one third of cases are associated with syndromes involving cardiac, neuropsychiatric, and craniofacial abnormalities (1,3). The patient discussed above was affected significantly by the ocular condition, but he did not present with any systemic findings. His visual acuity was presumed to be limited by multiple factors including nystagmus, incomplete retinal development, and high irregular astigmatism. The patient was referred to our clinic for a contact lens fitting to determine best corrected visual acuity by neutralizing the irregular astigmatism.

TREATMENT AND MANAGEMENT

Management of patients with microphthalmia and anophthalmia typically includes visual correction, if possible, and socket expansion to stimulate orbital development (4,5). The patient discussed above was unsure if he received socket expansion as a child in either eye. Contact lens use in patients with microphthalmia is not extensively documented in the literature. This patient was referred to our clinic for a contact lens fitting; although spectacle correction was preferable for protection due to his monocular status, the frames obstructed his view in his null point gaze direction. With his high amount of irregular corneal astigmatism, the optics of a rigid contact lens were preferred to that of a soft lens. Several lenses were fit and dispensed before finding a successful lens. Each lens trialed created certain complications at follow-up appointments:

- Lens #1: ICD scleral lens/3200um sag/14.5mm dia/8.88mm BC/-6.75D pwr
Adequate apical clearance, limbal clearance, centration; no blanching or impingement; VA 20/60+
Difficulty with application and removal; bubble formation on application presumed to be further complicated by nystagmus
Patient was unable to apply lens without bubbles after three separate training sessions
Goal for next lens: decrease diameter to improve ease of handling and decrease thickness of tear reservoir to prevent bubble formation

- Lens #2: Aspheric 0.65 eccentricity/10.5mm dia/7.50mm BC/-14.00D pwr
  - Fit similar to a scleral lens, landing outside the limbus but with low apical clearance (25 microns), decentered inferiorly; VA 20/50-
  - Goal for next lens: decrease diameter and flatten base curve to improve centration

- Lens #3: CAD 9.0mm dia/8.00mm BC/-11.00D power
  - Proper centration, slight apical clearance, adequate vertical movement
  - VA 20/50; patient reported noticeable improvement in vision

Lens #3 was dispensed and on follow-up patient reported acceptable comfort and vision with the lens. He successfully performed application and removal of the lens. Monocular precautions and the risk associated with contact lens wear were discussed in detail with the patient.

CONCLUSION
This case suggests that contact lenses can provide functional visual improvement for some patients with microphthalmia. It also highlights some of the unique challenges involved in fitting lenses on these patients. For patients with only one functioning eye, the concern of monocular precautions is an important consideration when fitting contact lenses. Centration of the lens is often difficult in microphthalmia due to the small size and irregularity of the cornea, and non-standard fitting relationships are often required. Due to the wide variety of manifestations of this condition, each patient must be addressed with a unique approach, and multiple trial lenses may be required to achieve the best fit.

REFERENCES