Is it amblyopia? How scleral lens can aid in determining potential visual acuity in the presence of concurrent strabismus and keratoconus

STATE UNIVERSITY OF NEW YORK
COLLEGE OF OPTOMETRY

Franklin Bui OD, Eva Duchnowski OD

BACKGROUND

This case explores a patient who presents with a large angle strabismus and advanced keratoconus. It is important to know whether the patient's reduced visual acuity is due to amblyopia, irregular astigmatism, or another etiology in order to address the chief complaint properly and effectively.

Keratoconus is a chronic, bilateral, ectatic condition that is characterized by corneal thinning and associated progressive steepening, apical scarring, and visual distortion.[1]

The traditional treatment of visual rehabilitation in keratoconus includes spectacle correction and contact lenses. Correction using contact lenses includes corneal gas permeable (GP) lenses, soft lenses, scleral lenses, hybrid contact lenses, and piggyback systems.[1] Treatment of keratoconus also includes collagen cross-linking, lamellar and penetrating keratoplasty, and intrastromal corneal ring segments.[2] Treatments are designed to improve visual potential, delay progression of keratoconus, or both.

Amblyopia is decreased vision in one or both eyes due to abnormal visual development during infancy or childhood. A common cause of amblyopia is strabismus, or eye misalignment, which can lead to the brain ignoring or turning off the misaligned eye and can lead to reduced vision. Treatment for strabismus should include patching and vision therapy to enhance eye alignment, binocularity, and visual processing. With large angle strabismus and in adults past their critical period for recovery of vision, another consideration is surgical correction of extraocular muscles.[3, 4]

The goal of surgical correction of extraocular muscles is to straighten misaligned eyes and potentially allow the eyes to work together as a team and maintain binocular function.[3] For adults with strabismic amblyopia, surgery can help improve psychosocial difficulties like self-image and interpersonal relationships.[5]

PATIENT HISTORY

45-year-old African American female presents for contact lens fitting and to determine the visual potential of her strabismic right eye prior to pursuing surgical intervention.

Ocular history: Longstanding history of strabismus of the right eye, keratoconus, reduced vision in both eyes; no previous eye surgery. Previous history of piggyback system with poor stability and visual function in the left eye. History of unsuccessful corneal lens fits in the right eye.
Medical history: Unremarkable
Ocular medications: None
Systemic medications: None

CLINICAL FINDINGS

Unaided visual acuity:
OD: 20/200, PH no improvement
OS: 20/200, PH no improvement

Motility:
Distance cover test: >40 prism diopter constant right exotropia
Duction: full OD and OS

Manifest refraction:
OD: balance 20/200
OS: -8.50 -2.50 x 090 20/80+

Anterior segment

Adnexa/ Lids/ Conjunctiva/ Iris/ Lens:
OD: Unremarkable
OS: Unremarkable

Cornea:
OD: Munson's sign, inferior ectasia, Fleischer ring, 2mm paracentral stromal scarring
OS: Munson's sign, inferior ectasia, Fleischer ring, 2mm paracentral stromal scarring, mild superficial punctate keratitis

Posterior segment

OD: unremarkable
OS: unremarkable

OCULUS PENTACAM IMAGES

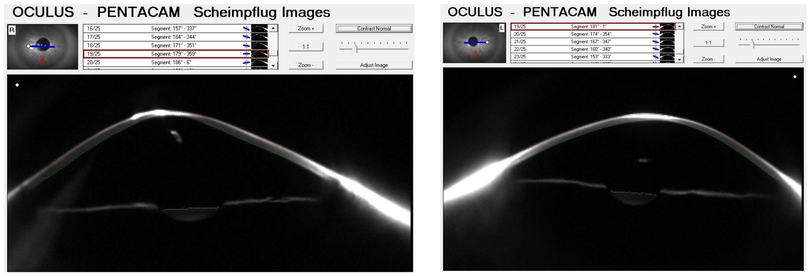

Figure 1. Scheimpflug images show corneal thinning of both the right eye (left) and left eye (right)

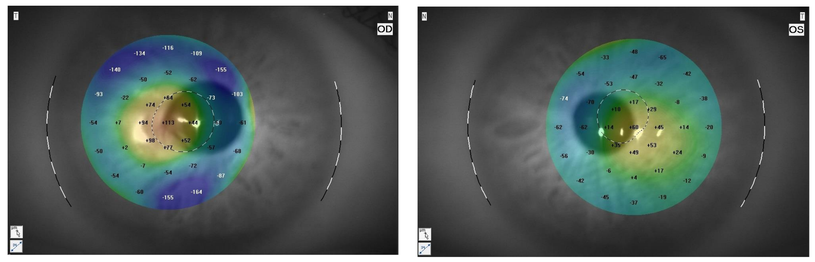

Figure 2. Anterior Elevation map

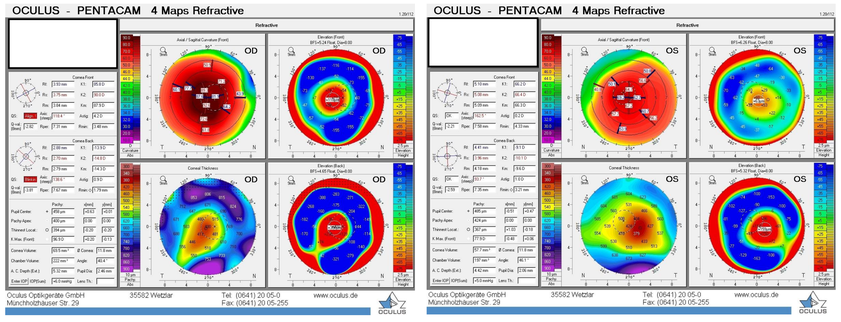

Figure 3. Topography map reveals apical corneal steepening, steep curvature, and thinning consistent with severe keratoconus in both eyes

Corneal topography values:

	OD	OS
Flat K	85.8 D	66.2 D
Steep K	90.0 D	66.4 D
Average K	87.9 D	66.3 D
K_{max} (Front)	96.9 D	77.9 D
Pachy at pupil center	458 microns	485 microns
Pachy at apex	400 microns	424 microns
Pachy at thinnest location	394 microns	376 microns

TREATMENT & MANAGEMENT

Scleral lens parameters:

	OD	OS
Lens	Alden Zenlens	Alden Zenlens
Sagittal depth (microns)	5600	5250
Base curve (mm)	6.23	7.30
Power (D)	-3.50	-10.00
Diameter (mm) and design	17.0 Prolate	17.0 Prolate
Center thickness (mm)	0.35	0.35
Advanced peripheral system (APS)	Standard	Toric 2 flat vertical/ standard horizontal

Assessment of fit after lens settling (~ 6 hours):

	OD	OS
Central vault	300 microns	350 microns
Apical Vault	100 microns	150 microns
Limbal vault	Adequate	Adequate
Centration	Centered	Centered
Edge	Good scleral alignment	Good scleral alignment
Visual acuity	20/50 with line isolation, 20/30 with letter isolation, PH NI	20/25+, PH NI

DISCUSSION

Amblyopia is a diagnosis of exclusion. It is important to rule out factors that can contribute to reduced visual acuity, including keratoconus. With the development of specialty contact lenses including rigid gas permeable lenses, clinicians are better equipped to manage and provide patients with keratoconus optimal visual outcome.

Spectacle and contact lens correction should be explored in patients with keratoconus in order to understand the extent on visual potential. For irregular corneas in which corneal gas permeable lenses do not provide adequate centration, visual acuity, or both, scleral contact lenses are a viable option.[1] Scleral lenses are fit to vault over the cornea and rest on the sclera. The fluid reservoir between the cornea and scleral lens masks corneal irregularity to provide visual improvement.

While both amblyopia and keratoconus can result in reduced vision, they are vastly different in their etiology and clinical presentation. Keratoconus often starts when individuals are in their late teens or early twenties, and tends to progress with time. Severity of progression varies between individuals, and can take years to progress from a mild to advanced stage. In contrast to keratoconus, amblyopia is decreased vision in one or both eyes due to abnormal development of vision in infancy or childhood.[4] Vision loss occurs because nerve pathways between the brain and the eye are not properly stimulated, which results in the brain favoring one eye over the other.[6] In strabismic amblyopia, one eye may turn in, out, up, or down, which leads to the brain ignoring that eye and resulting in reduced vision. However, not all patients with strabismus develop strabismic amblyopia. While patients with constant strabismus have poor prognosis for visual potential and binocularity, patients with alternating strabismus tend to have better vision.[4]

When refractive correction and vision therapy are insufficient in treating strabismus, surgical realignment of the eyes can potentially improve binocular function in a patient who demonstrates good visual potential of both eyes. For individuals with large angle exotropia, surgery can improve quality of life with positive psychosocial and functional effects.[7]

CONCLUSION

This case demonstrated the importance of exclusion of pathology prior to attributing visual reduction to strabismic amblyopia. For this patient with advanced keratoconus, a successful scleral lens fit showed significant improvement in visual acuity of both eyes. Furthermore, pursuing surgical ocular realignment may yield better binocular function and cosmesis for the patient moving forward.

REFERENCES

1. Moschos MM, Nitoda E, Georgoudis P, et al. Contact Lenses for Keratoconus- Current Practice. *The Open Ophthalmology Journal*. 2017;11:241-251. doi:10.2174/1874364101711010241.
2. Andreanos KD, Hashemi K, Petrelli M, et al. Keratoconus Treatment Algorithm. *Ophthalmology and Therapy*. 2017;6(2):245-262. doi:10.1007/s40123-017-0099-1.
3. Figueira EC, Hing S. Intermittent exotropia: comparison of treatments. Clin Experiment Ophthalmol. 2006;34:245–251. doi: 10.1111/j.1442-9071.2006.01199.x.
4. Levi DM, Knill DC, Bavelier D. Stereopsis and amblyopia: A mini-review. *Vision research*. 2015;114:17-30. doi:10.1016/j.visres.2015.01.002.
5. Menon V, Saha J, Tandon R, el al. Study of the psycho-social aspects of strabismus. J Pediatr Ophthalmol Strabismus. 2002;39(4):203–08
6. Asimellis G, Kaufman EJ. Keratoconus. In: StatPearls . Treasure Island (FL): StatPearls Publishing; 2017 Jun.
7. Sah SP, Sharma IP, Chaudhry M, el al. Health-Related Quality of Life (HRQoL) in Young Adults with Strabismus in India. *Journal of Clinical and Diagnostic Research : JCDR*. 2017;11(2):NC01-NC04. doi:10.7860/JCDR/2017/24541.9389.