

# Multidisciplinary Approach to Neurofibromatosis 2 Related Paralytic Lagophthalmos and Scleral Lens Wear



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## Introduction

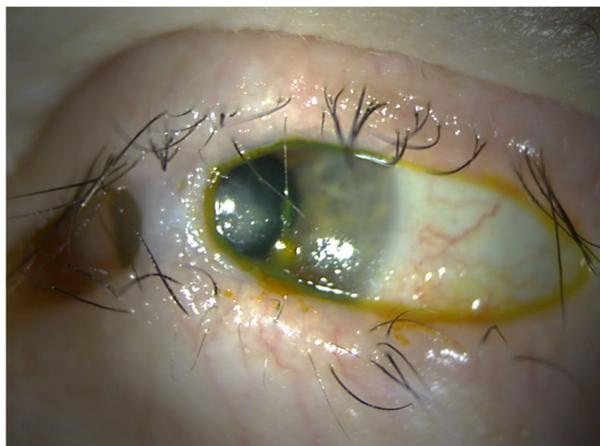
Neurofibromatosis refers to a group of genetic disorders that involve tumors of nerve sheath, including schwannomas and meningiomas. It can be classified as Neurofibromatosis 1 (NF1), Neurofibromatosis 2 (NF2), or schwannomatosis. NF2 commonly affects the eye as it includes tumors on the cranial nerves. Ocular manifestations range in severity based on the genetic expression and can include optic nerve sheath meningiomas, retinal hamartomas, optic nerve atrophy secondary to raised intracranial pressure or external tumor compression. Anterior segment findings can include neurotrophic keratitis, corneal scarring and ocular surface desiccation secondary to paralytic exposure. Other cranial nerves tumors will result in hearing loss, facial paralysis and binocular diplopia secondary to cranial nerve palsies. Scleral contact lenses are a well documented therapy to aid in ocular surface protection secondary to paralytic lagophthalmos and neurotrophic keratitis.

## Case Report

A 36-year-old Caucasian female presented for a scleral contact lens evaluation referred by the neuro-ophthalmology clinic. She had reported first experiencing severe headaches 20 years prior, leading to neurology evaluation and the diagnosis of bilateral acoustic schwannomas, and subsequently diagnosis of NF2. Consequent radiation and removal of the schwannomas resulted in complete left side facial paralysis, and partial right side paralysis. A complete tarsorrhaphy was placed over the left eye to aid in corneal and ocular surface protection. Due to the left sided facial paralysis tension on the lower eyelid the tarsorrhaphy frequently opened and resulted in a partial lateral tarsorrhaphy and central partial ankyloblepharon over the central cornea in the left eye (Figure 1).

To aid in the patient's left sided facial paralysis she had completed a procedure with a facial reconstructive surgeon. The goal of this procedure was to provide reanimation to the left side of the face and aid in improved lower lid apposition, which would also avoid surgical reconstruction to the lower lid and orbital region. A successful reanimation procedure was completed using the gracilis muscle and right sural nerve transposition. Due to the position of the ankyloblepharon and tarsorrhaphy, a scleral lens could not be applied to the left eye, she was then referred to the oculoplastics service for evaluation of the left eye and removal of the ankyloblepharon.

Figure 1. External Photography OS



Following removal of the ankyloblepharon, the patient reported current use of artificial tears every 2 hours and lubrication ointment at nighty. The partial lateral tarsorrhaphy was kept in place. Following the procedure, the patient's ocular surface OS was now completely visible with corneal scarring, neovascularization, and significant ocular surface desiccation (Figure 2). The patient demonstrated incomplete lid closure in both eyes, more significant on the left eye.

Figure 2. External Photography OS after ankyloblepharon removal



## Scleral Contact Lens Evaluation

The patient returned after her oculoplastic procedure for a diagnostic scleral contact lens evaluation. Her ocular surface was stable without epithelial defects OU but significant corneal staining OS>OD (Figure 3). A large diameter diagnostic scleral lens was chosen that had the ability to design the haptic in a quadrant specific manner and incorporate design features to alleviate any suction to the ocular surface.

Visual Acuity with updated manifest refraction:

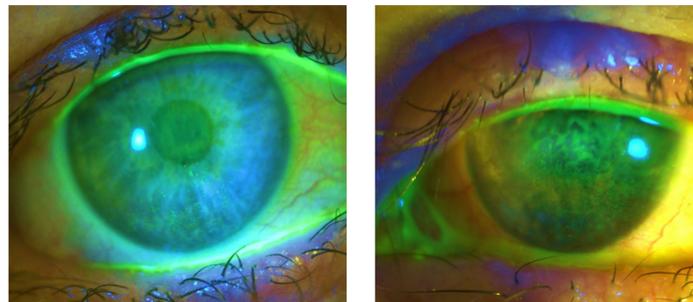
OD : 20/50+2

- 11.50 +1.00 x 105

OS: 20/300

- 7.75 + 1.25 x 093

Figure 3. External Photography OD, OS with NaFl Applied



## Diagnostic Scleral Lens Evaluation:

OD: BostonSight®SCLERAL 18.0 mm (R80-5e1 trial lens)

- Over-refraction: -4.50 DS → 20/20-2
- 300 μ vault centrally, adequate limbal clearance
- Haptic alignment other than mild lift-off inferior quadrant
- Plan: add 100μ centrally, steepen inferior quadrant 100μ, high Dk material without anterior surface coating (due to incomplete lid closure), add 2 SmartChannel™ areas to aid in decreasing lens suction

OS: BostonSight®SCLERAL 18.0 mm (L80-5e1 trial lens)

- Over-refraction: -3.75 DS → 20/60
- 350 μ vault centrally, adequate limbal clearance
- Haptic alignment other than mild blanching nasal
- Plan: add 150μ centrally, flatten nasal quadrant 150μ, high Dk material without anterior surface coating (due to incomplete lid closure), add 2 SmartChannel™ areas to aid in decreasing lens suction

The patient returned for scleral lens application and removal training, which was successful. She was advised to slowly increase wear time, continuing copious lubrication to the ocular surface with preservative free artificial tears and ocular ointment at night after lens removal.

## Scleral Lens Follow up

The patient returned 1 week after initiating lens wear with scleral lenses applied 4 hours prior to the appointment and reported significant improvement in visual acuity and ocular symptoms. Patient noted vision in the left eye fluctuates with lens surface deposits.

Visual Acuity OD: 20/20-1 OS: 20/60

## Fit Assessment OD

OD: 350 μ central vault, adequate limbal clearance, aligned haptic landing (Figure 4)

- Ocular surface after lens removal: no epithelial defect, rare PEE

Figure 4. External Photography of Scleral Lens OD



Final Scleral Lens Parameters OD:

- BostonSight®SCLERAL 18.0 mm (Contamac Optimum Infinite)
- BC: 8.30 mm
- Power: -2.98 DS
- Center Thickness: 370 μ
- SmartChannel™ nasal and temporal 20 degrees x 0.20mm depth

## Fit Assessment OS

OD: 500 μ central vault, adequate limbal clearance, aligned haptic landing (Figure 5)

- Surface wettability issues noted resulting in material change from Contamac Optimum Infinite to Boston EQ II
- Ocular surface after lens removal: no epithelial defect, 1+ PEE

Figure 5. External Photography of Scleral Lens OD



Final Scleral Lens Parameters OS:

- BostonSight®SCLERAL 18.0 mm (Boston EQ II)
- BC: 8.40 mm
- Power: -0.50 DS
- Center Thickness: 380 μ
- SmartChannel™ nasal, temporal, and inferior 20 degrees x 0.20mm depth

## Discussion

NF2 is characterized by bilateral schwannomas and CNS tumors which can cause a range of ocular sequelae. Paralytic lagophthalmos secondary to cranial nerve palsies can lead to severe desiccation of the ocular surface, blurred vision, corneal scarring, and corneal neovascularization. NF2 patients with ocular surface exposure secondary may undergo a variety of possible treatments, including surgical intervention. Scleral contact lenses are a treatment option for NF2 patients where other therapies have not provided adequate improvement in symptoms or protection. Scleral lens wear in this patient population should be closely monitored and co-managed with other specialties including neuro-ophthalmology, oculoplastics, and facial plastic surgeons. Due to the eyelid function abnormality, scleral contact lenses that are large in diameter and gently resting on the ocular surface are preferred. Scleral lens material selection and lab applied coatings should be carefully considered to optimize lens surface wettability and function.

## Conclusion

Scleral contact lenses can be considered as a treatment option for patients with paralytic lagophthalmos. Special considerations are needed due to the impaired eyelid function and ocular surface exposure. With a multidisciplinary approach, this patient was able to achieve an improved ocular surface and visual acuity, as well as improved facial sensation. The awareness of medical providers outside of the optometric profession being aware of scleral lenses as an option for this patient was paramount in this patient's outcome.

## References

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