# A Scleral Lens as an Alternative to Tarsorrhaphy for Ocular Rehabilitation in a Toddler

Amanda Dieu OD, Derek Louie, MSc, OD, Brooke Harkness, MS, OD

Casey Eye Institute, Oregon Health & Science University, Portland, OR

## INTRODUCTION

Central nervous system tumors are the most common solid neoplasm in children, occurring most commonly in the posterior fossa. Surgical resection followed by craniospinal irradiation and/or chemotherapy is the gold standard treatment.[1] Direct compression from the tumor or surgical interventions can disrupt cranial nerves that affect visual function. Visual abnormalities, found in 40% of children with posterior fossa tumors, manifest as papilledema, strabismus, lagophthalmos, neurotrophic keratopathy, and others.[1] This case highlights the use of a scleral lens for the management of exposure and neurotrophic keratopathy secondary to a high-grade posterior fossa neuroepithelial tumor in a toddler.

## CASE PRESENTATION

A 20-month-old female was referred for a medical contact lens evaluation for recurrent corneal abrasion and exposure keratopathy OS secondary to posterior fossa tumor and/or intracranial surgery. The mother of the patient hoped to seek other options to avoid or delay the need for a tarsorrhaphy. The patient experienced recurrent corneal abrasions several times a month for most of her life.

## MEDICAL AND OCULAR HISTORY

The patient's medical history was positive for a large posterior fossa hemorrhagic mass at 4 months of age, with a recurrence of the tumor one year later. She underwent two craniotomies for tumor resections, two treatments of chemotherapy with stem cell infusion, and one treatment of radiation. She otherwise had an uncomplicated non-oncologic history with a normal and full-term pregnancy. There is no family history of childhood or adult malignancies.

The patient's ocular history is complicated by multiple ophthalmic manifestations secondary to the tumor. She had full nerve palsies of the left CNIV through CNVII, and as a result suffers from strabismic and deprivational amblyopia OS. Previous treatments included bland ointment OS q2h, erythromycin ointment OS TID+ prn, a tape patch OS qhs for night exposure, and amblyopia patching OD 2 hours/day.

## CLINICAL FINDINGS

| | |
|---|---|
| Visual Acuity | CSM OD; CSUM OS |
| Cover Test | LET >50PD |
| Lids/Lashes | Normal OD; Lagophthalmos OS |
| Conjunctiva/Sclera | Normal OD; 1+ temporal sectoral injection OS |
| Cornea | Clear OD; 1+ interpalpebral PEE with central 1mm round epi defect inferotemporal to pupil with mild subepi haze OS |

## TREATMENT & MANAGEMENT

To reduce corneal exposure and protect the ocular surface, a 11.0 diameter corneal gas-permeable lens was ordered to function as a bandage contact lens.

However, at the follow up visit, the patient's mom reported that the lens regularly displaced from the ocular surface and spontaneously ejected. In attempt for better centration and coverage, a Valley Contax Custom Stable Elite scleral lens was ordered. The patient's mother was instructed to fill the scleral lens with Refresh Celluvisc preservative-free lubrication drop to help reduce bubbles upon application and allow for a thicker lubrication coverage during wear.

After four months of scleral lens wear, the patient's epithelial defect resolved and subepithelial haze significantly improved. Her mom reported that the patient tolerated the lens very well and had not had a recurrent erosion since using the lens. The tarsorrhaphy was postponed indefinitely in consultation with the cornea specialist and pediatric ophthalmologist. The final scleral lens design is described below.

| | Lens Design | BC | Diameter | CCZ | Lite | SLZ | Power |
|---|---|---|---|---|---|---|---|
| OS | Custom Stable Elite | 8.04 | 15.8 | STD | +6 | +2/-5 | +1.00 sphere |

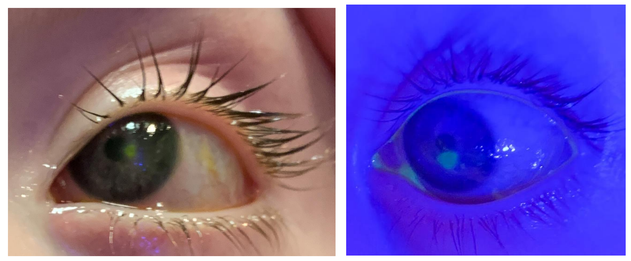
## DISCUSSION

The anatomical integrity, transparency, and function of the cornea relies on the health of the trigeminal and facial nerves. The ophthalmic division of the trigeminal nerve is responsible for providing sensitivity and trophic support to the cornea though the release of neurotrophic factors. Impairment of corneal sensory innervation impedes trophic tissue support, lacrimal reflex, and blinking. Corneal hypoasthesia or anaesthesia may result in neurotrophic keratopathy, characterized by spontaneous epithelial breakdown and poor healing.[2] Damage to the facial nerve, which normally controls the orbicularis oculi muscle for blinking, causes lid retraction, lagophthalmos and decreased tear production.[3]

Treatments of neurotrophic and exposure keratopathy focus on corneal wound healing and ulcer prevention. Therapies include aggressive preservative-free eye drops and ointments, topical antibiotics, topical autologous serum drops, amniotic membrane transplantation, topical recombinant human nerve growth factor, surgical tarsorrhaphy, and therapeutic contact lenses. Our patient has had limited success with topical lubrication alone. At this point, the patient's remaining options were to undergo a tarsorrhaphy, attempt autologous serum drops, or risk an infection and/or perforation. Mantelli *et al* suggested that tarsorrhaphy should be avoided in children as it comes with the risk of deprivational amblyopia. Thus, scleral lenses were left as a one of the few viable options for our patient. Scleral lenses are custom designed, gas-permeable, rigid lenses that retain a pool of fluid over the corneal surface. This "liquid corneal bandage" protects the surface from breakdown and optically neutralizes any irregularities of the surface, reducing the incidence and severity of amblyopia.[4] Prolonged therapeutic contact lens use may increase the risk of secondary infections. The use of prophylactic topical antibiotics is strongly recommended.[2]

## FIGURE 1

Epithelial defect OS at initial exam, under normal illumination (left) and Colbalt blue filter (right)



## FIGURE 2

Final therapeutic scleral lens OS showing aligned haptics in downgaze (left) and left gaze (right)

## CONCLUSION

- Posterior fossa tumors and surgical interventions can cause damage to cranial nerves, commonly resulting in vision-threatening ocular complications.[1]
- In children with corneal anesthesia, a prompt diagnosis and therapeutic intervention is important to prevent amblyopia and permanent visual damage.[2]
- Scleral lens wear is a valid long-term alternative to standard treatment options such as a tarsorrhaphy for pediatric patients with ocular surface disease as it provides effective protection of the ocular surface while optimizing vision.[3]

## REFERENCES

1) Gadgil N, Edmond J, Stormes K, Lam S, Shah V. Visual Complications of Pediatric Posterior Fossa Tumors: Analysis of Outcomes. Pediatr Neurol. 2019 Mar;92:48-54. doi: 10.1016/j.pediatrneurol.2018.09.016. Epub 2018 Oct 18. PMID: 30661932.
2) Mantelli F, Nardella C, Tiberi E, Sacchetti M, Bruscolini A, Lambiase A. Congenital Corneal Anesthesia and Neurotrophic Keratitis: Diagnosis and Management. Biomed Res Int. 2015;2015:805876. doi: 10.1155/2015/805876. Epub 2015 Sep 16. PMID: 26451380; PMCID: PMC4588028.
3) Lee S, Lew H. Ophthalmologic Clinical Features of Facial Nerve Palsy Patients. Korean J Ophthalmol. 2019 Feb;33(1):1-7. doi: 10.3341/kjo.2018.0010. PMID: 30746906; PMCID: PMC6372383.
4) Gungor I, Schor K, Rosenthal P, Jacobs DS. The Boston Scleral Lens in the treatment of pediatric patients. J AAPOS. 2008 Jun;12(3):263-7. doi: 10.1016/j.jaapos.2007.11.008. Epub 2008 Feb 7. PMID: 18258469.

The authors have no financial interest in any of the products referenced in this study, nor were the authors supported by any company referenced in this project. Casey Eye Institute is supported by unrestricted departmental funding from Research to Prevent Blindness (New York, NY) and by grant P30 EY010572 from the National Institutes of Health (Bethesda, MD).

Research to Prevent Blindness

CASEY EYE Institute