Contact Lens Management of a KID Syndrome Patient

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Background
Keratitis-ichthyosis-deafness (KID) Syndrome is a rare genetic multi-system disorder that causes dry, scaly skin, severe ocular surface disease, and profound hearing loss. Approximately 100 cases have been reported. KID Syndrome patients present to eye care providers with ocular pain, photophobia, corneal neovascularization, and cicatrization leading to progressive vision loss and eventual blindness. Scleral contact lenses can be used to improve comfort and vision.

Case: 04/2017

- 10-year-old Hispanic male presents for a scleral contact lens consult due to severe dry eyes OU. Patient's mother also notes rubbing of eyes and significant light sensitivity. Mother also reports frequent and copious use of artificial tears and warm compress with little to no relief. She was told by the patient's ophthalmologist that the patient would be blind by the age of 12. He attends a school for the blind and deaf.
- Medical history:
  - KID Syndrome: complete hair loss, widespread ichthyosis, severe deafness
- Ocular history:
  - Severe keratitis with severe corneal neovascularization and corneal scarring OU
- Medications:
  - Terbinafine HCl (anti-fungal cream)
- Visual acuities through glasses:

Eye	Lensometry	Visual Acuity
OD	-5.25	20/50-2
OS	-5.00 -0.50 x 176	20/40

  - **Patient had great difficulty holding eyes open for any period of time. Hard, frequent blinks or eye closing with eye rubbing noted.
- Pupils, EOMs, confrontation visual fields: grossly normal
- Slit lamp examination (see figure 1):

OD		OS
Scaling, dryness; no eyebrows	Adnexa	Scaling, dryness; no eyebrows
Extensive loss of lashes (two lashes seen); 2+ MGD	Eyelids	Extensive loss of lashes (one lash seen); 2+ MGD
White and quiet	Sclera	White and quiet
Severe perilimbal injection; 3+ injection of palpebral conjunctiva	Conjunctiva	Severe perilimbal injection; 3+ injection of palpebral conjunctiva
3+ PEE centrally; 3.5mm neovascularization 360, central nebular scarring	Cornea	3+ PEE centrally; 3.5mm neovascularization 360, central nebular scarring
Deep and quiet	AC	Deep and quiet
Flat, intact	Iris	Flat, intact
Clear	Lens	Clear

- Given patient's extreme photophobia, unable to acquire a reliable corneal topography

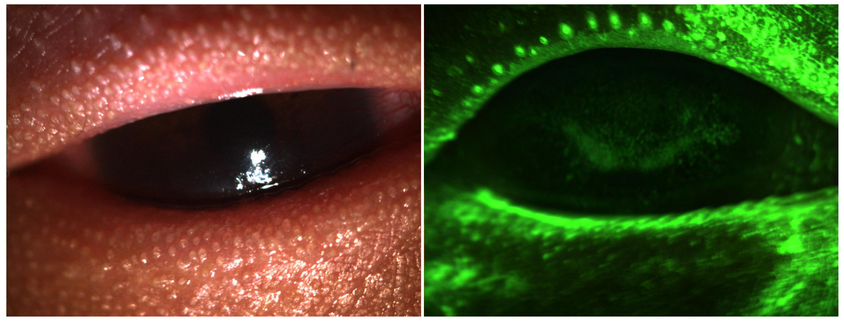

Figure 1: 04/2017. Left photo: Patient's right eye at his first consultation visit. Note the texture of the patient's skin and extensive madarosis. Note the dulled corneal reflex indicating corneal dryness and irregularity. Right photo: Same eye with fluorescein staining. Note the significant punctate staining present. The patient's left eye was very similar in presentation.

References
1. NIH US National Library of Medicine, Genetics Home Reference: https://ghr.nlm.nih.gov/condition/keratitis-ichthyosis-deafness-syndrome#diagnosis
2. Strul S, Straughn P. Successfully improving visual acuity in keratitis-ichthyosis-deafness syndrome utilizing gas-permeable contact lenses: a case report. Eye & Contact Lens 2018; 44:S330-S332.
3. Shanker V, Gupta M, Prashar A. Keratitis-Ichthyosis-Deafness syndrome: a rare congenital disorder. Indian Dermatol Online J 2012; 3(1):48-50.

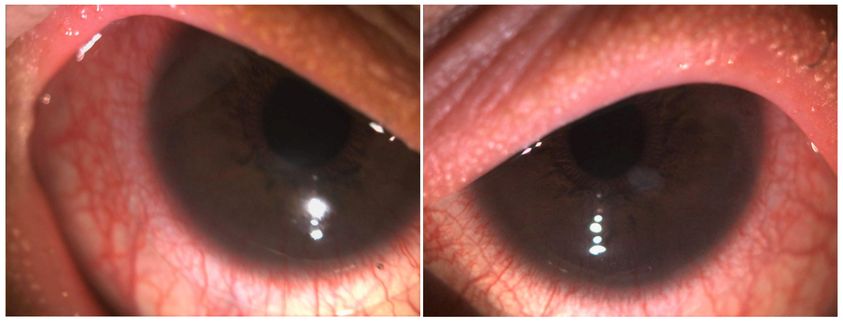

Figure 2: 08/2017. Given the patient's age and dexterity, the patient was fit into a 14.8mm diameter scleral contact lens. Left photo: Patient's right eye with the finalized scleral lens in place. Right photo: Patient's left eye with the finalized scleral lens in place. Note the amount of corneal neovascularization. The details of the finalized 2017 lens parameters are below.

Eye	Material	BC (mm)	CLP (D)	DIA (mm)	Sag (mm)	CT (mm)	Limbal Clearance (µm)	APS
OD	Boston XO2 Clear (dot)	8.13	-4.50	14.8	3.950	0.25	+200	Flat 5
OS	Boston XO2 Clear	8.13	-4.50	14.8	4.050	0.25	+200	Horizontal: Flat 9 Vertical: Flat 2

He initially had a lot of difficulty with lens application; however, improved by the finalized pair. The patient was able to see 20/25-3 OD and 20/25+1 OS. He appeared to be pleased with the comfort and vision through both lenses as his mother stated that he is always eager to put the lenses on in the morning.

- Average wear time: 14 hours
- Solution: ClearCare, filling with Lacripure

OD lens fit:
- Centrally: 75um
- Limbal vault: 25um 360º
- Trace impingement nasally

OS lens fit:
- Centrally: 125um
- Limbal vault: 25um 360º
- Very mild blanching superiorly, trace impingement nasally

Plan: RTC 6 months for anterior segment check.

Case: 08/2018

- Patient, now 11-years-old, returns for a comprehensive eye exam and anterior segment check. (Patient had been doing well and lens had fit well at 6-month anterior segment check OU.) He notes that his eyes have been more red recently and notes pain after removing his scleral contact lenses. He also notes that his vision is blurry when he is looking straight ahead for long periods of time.
  - Average wear time: 16 hours
  - Patient naps in lenses, approximately 3 days/week
  - Comfort while wearing lenses: OD 10/10, OS 10/10
  - Vision while wearing lenses: OD 7/10, OS 7/10
  - Solution: Not sure, described as blue and white bottle
- Medical history and ocular history unchanged
- No medications
- Visual acuities through lenses: OD 20/25+2, OS 20/25+2
  - **Able to hold eyes open for longer periods of time but still very light sensitive. No longer squinting.
- Pupils, EOMs, confrontation visual fields: normal
- Slit lamp examination (see figure 3):

OD		OS
Scaling, dryness; no eyebrows	Adnexa	Scaling, dryness; no eyebrows
Madarosis, 2+ MGD	Eyelids	Madarosis, 2+ MGD
White and quiet	Sclera	White and quiet
1+ perilimbal injection	Conjunctiva	1+ perilimbal injection
4.5x4mm central clear zone; conjunctivalization and neovascularization of rest of cornea	Cornea	3x4.5mm central clear zone; conjunctivalization and neovascularization of rest of cornea

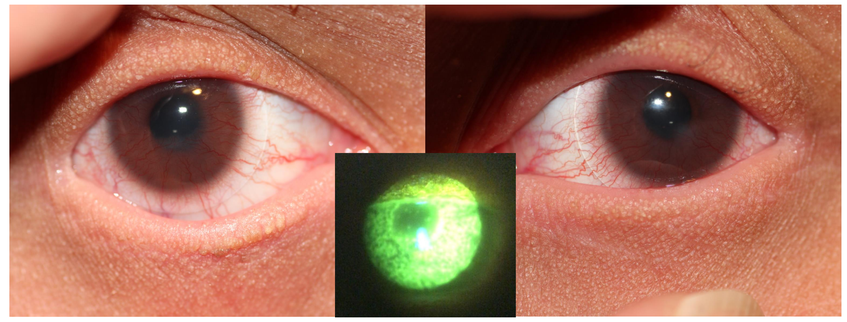

Figure 3. 08/2018. 2017 lenses seen on the right (left photo) and left (right photo) eye. Note the worsened conjunctivalization (center photo) and neovascularization of the cornea, but improvement in the conjunctival injection. These lenses were no longer fitting well as outlined below.

OD lens fit:
- Centrally: 25um
- Limbal vault: touch 360º
- No blanching or impingement

OS lens fit:
- Centrally: 50um
- Limbal vault: touch 360º
- No blanching or impingement

Given the patient's improved application and removal techniques, the patient was re-fit into a larger lens diameter to achieve better limbal clearance. Additionally, the patient was prescribed erythromycin 500mg PO TID and fluorometholone 0.1% BID OU per the recommendation of the corneal specialist to decrease the amount of inflammation at the ocular surface. Restasis was previously trialed but became a financial burden upon the family.

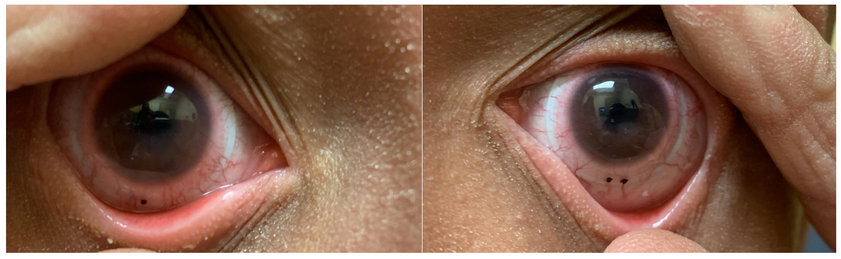

Figure 4: 09/2018. One of the initial trial lenses in the larger diameter demonstrating that significant flattening of the peripheral curves was needed in the horizontal meridian.

Material	BC (mm)	CLP (D)	DIA (mm)	OZD (mm)	CT (mm)	PC1r/w (mm)	PC2r/w (mm)	PC3r/w (mm)	PC4r/w (mm)	PC5r/w (mm)
Optimum Extreme	7.60	-8.12	16.6	8.00	0.20	7.73/1.0	9.71/8.65 /1.0	8.37/8.47 /1.0	15.56/12.2 /0.9	18.52/14.5 /0.4
Optimum Extreme	7.60	-8.00	16.6	8.00	0.20	7.73/1.0	9.71/8.65 /1.0	8.37/8.47 /1.0	15.56/12.2 /0.9	18.52/14.5 /0.4

The final lens parameters are listed above for his 2018 scleral lenses. He was able to see 20/20-1 OD and 20/20-2 OS. He stated his vision was an 8/10 OU and his comfort was 10/10 OU.

- Average wear time: 14 hours
- Solution: Unique PH, filling with Lacripure

OD lens fit:
- Centrally: 225um
- Limbal vault: 25um N/T/S, 50um I
- Trace blanching temporally

OS lens fit:
- Centrally: 250um
- Limbal vault: 25um N/T/S, 50 I
- Moderate heel blanching nasally

He returned in November 2019 and is currently undergoing another refitting process due to limbal touch 360º with these 2018 lenses.

Conclusion
KID Syndrome is a rare cause of limbal stem cell deficiency and in severe cases, regardless of management options, can lead to complete cicatrization of the cornea and blindness. Scleral lenses can be used to help manage the patient's symptoms of decreased visual acuity and light sensitivity. Patients with KID Syndrome should be co-managed with a corneal specialist and need to be monitored closely.