Managing Keratoconus in a Pediatric Patient with Corneal Scarring Secondary to Hydrops: A Case Report

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Abstract: A case of a pseudoophakic pediatric patient with keratoconus and scarring due to hydrops who was fit with a piggyback system with a vast improvement in vision

I. Case History

- Patient demographics:
  - 9 year old Caucasian female

- Chief complaint
  - Wants improvement in vision due to continuing decline in vision

- Ocular
  - Seen in clinic 14 months ago for contact lens fit for keratoconus, fit discontinued
  - S/P cataract extraction for congenital cataracts (psuedophakic) OU, age 3
  - Keratoconus OU, diagnosed age 6
    - History of apical scarring OD
  - S/P Hydrops OD 1 year and 4 months ago, OS 8 months ago

- Medical
  - Unremarkable

- Medications
  - None

- Family History
  - Father-keratoconus

II. Pertinent findings

- Clinical
  - VA (uncorrected) OD 20/100, OS 20/400, OU 20/100
    PH OD 20/80, PH OS 20/300
  - Refraction: Scissoring on Retinoscopy
    OD -0.75-4.25x086 20/80- OS +2.75-4.50x084 20/300
  - VF: FTFC OD, OS EOMS: FROM OU, Pupils: Slightly irregular pupillary border OU (likely due to surgery), reactive to light, -APD OU
  - Slit lamp exam (pertinent findings):
    - Cornea: OD: central scar ~ 2x2mm, +Munson’s Sign, large area of stromal thinning (centrally); OS: central scar ~1x1mm, +Munson’s Sign, large area of stromal thinning (central/inferior), faint Fleischer’s Ring (concentrated inferiorly)
    - Lens: IOL with PCO with clear visual axis OU
    - Very small palpebral aperture and small corneal size observed
      - OD HVID 9mm, IPA <7mm
      - OS HVID 9mm, IPA <7mm
Topography (Medmount)

- Simulated K’s: (14 months ago) OD 41.60@178/42.80@088, OS 60.00@155/55.60@065; Topography shows large steep central cone area with shallow central area OU
- Simulated K’s: (Current visit) Only able to obtain right eye. OD 57.40@161/70.80@071, OS unobtainable with Medmount Topographer; Topography OD shows large steep central cone area with more shallow central area (less flat centrally than previous topography)

Diagnostic fit of corneal gas permeable lenses with hydrogel carrier lens (piggyback system) for increased comfort

- Vast improvement in visual acuities with contact lenses,
  - VA with contact lenses 20/50 OD, 20/50 OS, 20/50 OU with over-refraction
- Small diameter custom hydrogel contact lenses of OD 12.5 mm and OS 13.0 mm were ordered to make insertion less difficult. Steeper base curve of carrier lens indicated due to unacceptable diagnostic lens fitting.
- Corneal gas permeable lenses ordered with over refraction.
- Bifocal polycarbonate spectacles with no distance correction and a +2.50 add were prescribed to be worn over contact lenses for protection.

III. Diagnoses

- Primary/leading
  - Keratoconus, OU
  - Central corneal scarring secondary to Acute Hydrops OD, OS
  - Irregular Astigmatism, OU
  - Pseudophakia, OU

- Differential Diagnoses
  - Keratoglobus
  - Progressive Myopia
  - S/P Infectious Keratitis, OU
  - Amblyopia, OU

IV. Diagnosis and discussion

- Condition
  - Keratoconus is a thinning, or ectasia, of the cornea that results in a conical shape and induces irregular astigmatism. It is usually bilateral and often asymmetric. It is the most common corneal ectasia. The prevalence of this disease ranges from 50 to 230 per 100,000. In the United states it is reported to be about 1/2000 persons. It is non-inflammatory in nature and can occur intrinsically or secondary to surgical procedures. Keratoconus in young children is relatively rare because the typical onset of keratoconus is in the second decade of life, usually coinciding with puberty. In multiple studies pediatric keratoconus has been shown to progress faster and is more severe at diagnosis than in adults. One study found 27.8% of children with keratoconus reached Stage 4, in contrast to only 7.8% of adults. Diagnosis of keratoconus in children is often delayed. This is concerning considering the progressive nature of the condition and possible impact of visual impairment on the child’s development and future quality of life. An indication of this under-diagnosis is evident in studies showing that corneal hydrops, usually a late complication of keratoconus, is often the presenting sign that leads to the diagnosis of keratoconus in children. Corneal hydrops is a condition characterized by swelling of the cornea and can lead to scarring and further decrease of vision.
Unique attributes
- This case presents a unique presentation of keratoconus
  - Early age of onset and severity of keratoconus.
  - History of acute hydrops in both eyes.
  - Coexistence of congenital cataracts and Psuedophakia OU
  - Small eye size and palpebral aperture and the role it plays in management
  - Genetic Component

IV. Treatment, management

- Treatment
  - Treatment of keratoconus in early stages involves glasses and soft contact lenses in early stages when vision is not greatly compromised. As the condition progresses, gas permeable corneal or scleral contact lenses may provide improvement in vision correction due to irregular astigmatism.
  - In late stages or when vision is severely reduced by scarring, Keratoplasty may be warranted. Approximately 10 to 15 percent of patients with keratoconus will require Keratoplasty. Penetrating Keratoplasty (full thickness corneal transplant) is the most common procedure, but Deep Lamellar Keratoplasty is utilized as well.
  - Future indications of referral for Corneal Cross-linking procedure. The treatment involves instillation of a solution of Riboflavin and UVA irradiation. Its goal is to strengthen the corneal collagen and slow down the progression of Keratoconus. Many laboratory and clinical studies have shown promising results. It is currently being performed in Europe and is going though FDA approval in the United States.
  - Continued close monitoring of patient and progression of condition is important.

- Bibliography
VI. Conclusion

- When fitting children with keratoconus with contact lenses, different lens options should be considered. The piggyback lens system was a good option for this patient because it allowed for increased comfort and easier insertion and removal. Proper instruction of insertion and removal is critical to success with contact lenses in children. In this case, the patient had an extremely positive response to the improvement in vision and remarked she felt as if she was seeing her father’s face clearly for the first time.

- Clinical pearls
  - Detecting keratoconus early in children may be more imperative due to evidence of more severe stage at onset and faster progression. Closer monitoring may be indicated as well.
  - Management of keratoconus in children may call for different considerations than in adults due to smaller palpebral aperture and HVID.
  - Contact lenses should be considered a viable option in children due to vast improvements in vision and ability to delay surgical intervention.