Morning Report

Daniel Corbett, PGY-3

Preceptor: Drs. Singh and Gelston
3 day old male with report of “poor red reflex and cloudy cornea” in both eyes
• OB Hx:
  – Born via SVD @ 39.5 weeks. No trauma/forceps during delivery.
  – MOC Rubella immune
  – Tested +Gonorrhea/Chlamydia 3 months prior to delivery that was successfully treated

• FHx: Unremarkable

• PMHx: No other systemic medical conditions. Pt reported to have failed his newborn hearing screening exam
EXAM

- Gen: pt alert and appropriate for age
- VA: BTL both
- IOP: STP both
- Pupils: Unable to be visualized
- EOM: appear full both by spontaneous eye movements
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<tbody>
<tr>
<td>L/L</td>
<td>WNL</td>
<td>WNL</td>
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<tr>
<td>C/S</td>
<td>White and Quiet</td>
<td>White and Quiet</td>
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<tr>
<td>K</td>
<td>Ring of central corneal clouting. Peripheral cornea clear. No epithelial defect</td>
<td>Ring of central corneal clounding. Peripheral cornea clear. No epithelial defect</td>
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<tr>
<td>AC</td>
<td>Not well visualized</td>
<td>Not well visualized</td>
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<tr>
<td>I</td>
<td>Not well visualized</td>
<td>Not well visualized</td>
</tr>
<tr>
<td>L</td>
<td>Unable to assess</td>
<td>Unable to assess</td>
</tr>
<tr>
<td>DFE</td>
<td>Unable to assess</td>
<td>Unable to assess</td>
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Differential?

- **S** - Sclerocornea
- **T** - Tears/Trauma (Congenital Glaucoma/Birth Trauma-forceps)
- **U** - Ulcer (TORCH/HSV/neurotrophic)
- **M** - Metabolic opacities (Mucopolysaccharidoses/Mucolipidoses/Sphingolipidosis)
- **P** - Posterior corneal defect (Posterior keratoconus/Peters’ Anomaly/Staphyloma)
- **E** - Endothelial dystrophy (CHED, Posterior polymorphous dystrophy, CHSD)
- **D** - Dermoid
Additional Studies?
• Referred to TCH for EUA and UBM
UBM demonstrates anterior segment dysgenesis with iris adherent to cornea in both eyes.
Exam Under Anesthesia

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<tr>
<td>IOP</td>
<td>13</td>
<td>13</td>
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<tr>
<td>K Diameters</td>
<td>10 mm (H), 9 mm (V)</td>
<td>10 mm (H), 9 mm (V)</td>
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<td>C/S</td>
<td>White &amp; Quiet</td>
<td>White &amp; Quiet</td>
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<tr>
<td>K</td>
<td>Central Opacity</td>
<td>Central Opacity</td>
</tr>
<tr>
<td>AC</td>
<td>Shallow at areas of I-C touch</td>
<td>Shallow at areas of I-C touch</td>
</tr>
<tr>
<td>I</td>
<td>Anterior synechiae</td>
<td>Anterior synechiae</td>
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<tr>
<td></td>
<td>(iridocorneal touch) from 4-7 oc</td>
<td>(iridocorneal touch) from 1-6 oc</td>
</tr>
<tr>
<td>L</td>
<td>Clear</td>
<td>Clear</td>
</tr>
<tr>
<td>DFE</td>
<td>No view but with small area nasally of poor red reflex</td>
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Pt did not have craniofacial dysmorphism, dental abnormalities, or redundant umbilical skin.
Plan for PKP Right
Peters’ Anomaly
Background

• Part of a spectrum of Anterior Segment Dysgenesis

• Most cases are sporadic

• Multiple causative gene loci: PAX6, PITX2, FOXC1, CYP1B1, MAF and MYOC

• Can be unilateral or bilateral (80%)

• Characterized by defects in the corneal stroma, Descemet’s membrane and endothelium in the area of opacification
Clinical Features

- **Cornea**
  - Opacity Spectrum: Faint stromal opacity to dense opaque central leukoma
  - Rarely Vascularized
  - Peripheral cornea clear
  - Limbus may be scleralized

- **Iris**
  - Strands of iris tissue extending to posterior border of the corneal leukoma (Type I)

- **Lens**
  - Central keratolenticular adherence along with other lenticular abnormalities (Type II)
Associated Conditions

• **Glaucoma**
  - may be present in < 50% of cases
  - 2/2 incomplete development of TM/Schlemm’s Canal and/or presence of shallow AC

• **Peters’-Plus Syndrome (Type III)**
  - Developmental Delay
  - Congential Cardiac Defects
  - Cleft Lip & Palate
  - Craniofacial Dysplasia
  - Skeletal Changes
Prognosis

- Associated with severe visual impairment

- Amblyopia from opacification of the central visual axis from central leukoma

- Glaucoma

- Spontaneous clearing may occur if central opacity mild
Management

• Large Peripheral Optical Iridectomy
  – Mild Corneal Involvement
  – No Lens Involvement

• PKP
  – Significant Corneal Involvement
  – Bilateral
Challenges of PK in Infants

• Reduced Scleral Rigidity/Increased Elasticity
• Risk of Spontaneous Lens Expulsion
Success of PKP?

- Retrospective review by Yang looked at 144 PKs for Peters Anomaly.
- Median age of first PKP 4.4 months with avg f/u of 11.1 years.
- Overall probability of maintaining a clear first graft:
  - 56% @ 6 months
  - 49% @ 12 months,
  - 44% @ 3 years
  - 35% @ 10 years.
Post Operative Challenges

- Accurate Refraction
- Occlusion Therapy Post-Op to Prevent Ambylophia
- Glaucoma
  - 50% result in significant visual loss
  - Often need filtering surgery or cyclocryotherapy for IOP control
Surgical Management of Glaucoma

• Notoriously challenging to control

• Study by Yang et al, looked at 34 eyes that underwent total of 126 glaucoma procedures

• 32% eyes had IOP <21

• 53% eyes had VA LP or NLP
Our Patient

- POM # 2 PKP right
- Doing well, graft clear at last visit 2 wks ago
- IOP grossly normal
- No note of nystagmus or occlusion therapy being initiated
- On Pred Forte QID right
- Plan for PKP left eye in next 2-3 weeks
References


