**Tips / Tricks for Operating on Adult Patients with Pediatric Issues**

- Adult ROP
- FEVR
- Coats
- Norries
- Sticklers
- Marfan's
- Dislocated/subluxed lens
- VHL
- X-linked retinoschisis

- Trauma
  - Cycodialysis cleft
  - IOFB
  - Macular hole
- Congenital optic nerve head disorders
  - Morning Glory detachment
  - Coloboma detachment
  - Optic Nerve Pit
- ARN

**Possible Scenarios**

**RD and ROP in Adults**

- 108 patients over 30 years, 216 eyes
  - 30/216 developed RD (13%)
  - 7/30 RD repair failed (initially SB)
  - Characteristics included posterior break, oval holes, horseshoe tears
  - 60% retained initial Va of 20/60 or better
    - Kaiser, RS, Trese, MT et al. 2001

**RD and ROP in Adults**

- Adult ROP
  - Typical detachment is temporal, extramacular, associated with multiple breaks with vitreous traction and no PVD
  - Best treated with broad scleral buckle (276, 287), good imbrication beneath breaks, extender if necessary, cryo, drainage, and no vitrectomy and no gas.
  - If there is a posterior break, likely primary vitrectomy is of benefit
**ROP in Adults**

- 25% RD
- 90% myopia
- 87% cataract
  - Tear or detachment in 23%
  - 1/3 improved, stayed the same or worsened after cataract surgery
  - Smith and Tasman 2005

**Cataract and ROP in Adults**

- Adult ROP
  - Cataract induced retinal complications
  - Average age 40 yo
  - Roughly 1/3 improve, stay the same or get worse
  - Retinal tear or detachment in 25%
  - Kaiser RS et al 2008

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**Adult ROP**

- 18 yo with fovea involving low lying rhegmatogenous retinal detachment
- Va RE = 20/150 LE = 20/30  Rx -9.50 sph ou
- SRF 2:00-10:00 RE (clockwise) no breaks visible
- Management?
Adult ROP

- 56 yo white male
- BCVA: 20/25 OD, 20/60 OS
- History of premature birth
- CE/IOL/PPV/MP OD for ERM/VMT in 1999
- CE/IOL/PPV/MP OS for ERM/VMT in 2008
  - IVK OS 4/09
  - RT: 546→335
  - Va: 20/60 → 20/50

Case assembled by Nathan Steinle
OD OD
SB alone if you can, obvious traction PPV
Cataract surgery always a risk for complication
Foveoschisis like condition can be both tractional and ischemic in origin
Watch for NV!!!
Coats’ Disease

- Aneurysmal dilatation
- Retinal capillary non-perfusion
- Light bulb or clustered aneurysms
- Sheathed vessels

Coats’ Disease

- Localized lipid deposition
- Subretinal exudate

Coats’ Disease

- Localized lipid deposition
- Subretinal exudate
Coats’ Disease

- Exudative retinal detachment
- Subretinal cholesterol crystals
Coats’ Disease

- 18 yo male dx’d upon referral with bilateral Coats disease s/p remote retinal procedure RE and multiple cryoretinopexy LE complicated by total retinal detachment
- Va RE= NLP LE = 40/400
- RE phytsis bulbi, LE normal anterior segment, DFE demonstrates RRD with large inferior break and PVR C2,3 and 4

Coats’ Disease

- Can have RRD as an adult, SB/PPV with peripheral thermal treatment
- Laser or Cryo ischemic areas
- With exudative RD, IVK not a big help
- Trend towards anti-VEGF plus thermal treatment
- Central lipid often CNV confuser

FEVR

- Recessive, dominant, and X-linked
- Congenital bilateral but assymetric
- Temporal traction, heme, peripheral avscularity, exudation
- Spectrum of disease involving Norries, PHPV, Coats
FEVR

- Recessive, dominant, and X-linked
- Congenital bilateral but assymetric
- Temporal traction, heme, peripheral avascularity, exudation
- Spectrum of disease involving Norries, PHPV, Coats

FEVR: Peripheral avascularity
Premature arrest of retinal angiogenesis

### Pathogenesis

#### Genetics

- 100% penetrance, variable expressivity
- Sporadic cases: FZD4, NDP

<table>
<thead>
<tr>
<th>Gene locus</th>
<th>Gene</th>
<th>Inheritance</th>
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<tbody>
<tr>
<td>EVR1</td>
<td>11q13-q23</td>
<td>FZD4</td>
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<tr>
<td></td>
<td>11q13.14</td>
<td>LRP5</td>
</tr>
<tr>
<td>EVR2</td>
<td>Xp11.4</td>
<td>NDP</td>
</tr>
<tr>
<td>EVR3</td>
<td>11p13-p12</td>
<td>?</td>
</tr>
<tr>
<td>EVR4</td>
<td>11q</td>
<td>?</td>
</tr>
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</table>
Norrie disease
- 4th generation family, affected female
- Missense mutation in exon 3, transmitted through 3 generations
- Mutation in one allele: affected maternal grandmother and unaffected mother

FEVR
- Sporadic case, affected female
- Heterozygous missense mutation in exon 3
- Mutation undetected in unaffected family members

Due to nonrandom X-chromosome inactivation

Variable progression:
- Rapid progression
  - Total retinal detachment by 1st or 2nd decade
  - Significant cause of RD in patients younger than age 30
- Slow or no progression
  - Adulthood: appears to remain stable

**Classification and Management**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
<th>Management</th>
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<tbody>
<tr>
<td>1</td>
<td>Avascular retinal periphery without extraretinal vascularization</td>
<td>Observation/no progression</td>
</tr>
<tr>
<td>2</td>
<td>Avascular retinal periphery with extraretinal vascularization</td>
<td>Laser ablation/no progression</td>
</tr>
<tr>
<td>3</td>
<td>RD not involving fovea</td>
<td>Scleral buckle + vitrectomy</td>
</tr>
<tr>
<td>4</td>
<td>RD involving fovea</td>
<td>Scleral buckle + laser + vitrectomy</td>
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<tr>
<td>5</td>
<td>Total RD</td>
<td>Vitrectomy</td>
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</table>

**Management**

- Frequent ophthalmologic follow-up
- Amblyopia management
- Screen family members
- Genetic counseling
  - Genetic testing available: NDP
### Coats vs. FEVR

<table>
<thead>
<tr>
<th>Features</th>
<th>Coats</th>
<th>FEVR</th>
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<tbody>
<tr>
<td>Age of onset</td>
<td>6-8 yo (mean)</td>
<td>Birth</td>
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<tr>
<td>Gender</td>
<td>MF 10:1 (&gt;20 yo)</td>
<td>MF 1:1</td>
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<tr>
<td>Laterality</td>
<td>Unilateral</td>
<td>Bilateral</td>
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<td>Inheritance</td>
<td>Non-genetic/sporadic</td>
<td>AD, AR, XR</td>
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<td>Retinal vessels</td>
<td>Capillary non-perfusion</td>
<td>Avascular zone</td>
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<td>Telangiectasia</td>
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<td>Microneuromas</td>
<td>Neovascularization</td>
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<tr>
<td>Retinal exudates</td>
<td>Present</td>
<td>Present</td>
</tr>
<tr>
<td>Vitreoretinal traction</td>
<td>Absent</td>
<td>Present</td>
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</table>

### FEVR

- 2 yo male dx’d upon referral with FEVR
- Genotype showed X-linked NDP mutation
FEVR

- WFA essential
- Thermal treat ischemic areas
- Tractional and exudative detachmets
- Asymmetric bilateral

Post op 1 year

Norries

- Don't operate
**Stickler’s Syndrome**

- Hereditary Progressive arthro-ophthalmopathy
- Col2A1 (75% Type I)
- Col11A1 (ocular Type II) or Col11A2 (non-ocular, Type III)
- Col9A1 (recessive)
- Vitreous is composed of Type II, IX, and XI
- Marshall Syndrome
- Type I likely to have RRD, Type II can, Type 3 not. Hearing loss c/w Type II or III}

**Stickler’s Syndrome**

- 19 yo male dx’d upon referral with RRD LE and Stickler’s syndrome s/p vitreoretinal procedure age 6 mos with CE in Kentucky RE
- Va RE=LP LE = 20/60
- Centered PCL RE IOP 8; clear lens LE IOP 18
- DFE attached retina RE macular scarring RRD LE HST x 2 at 2:00 and 7:00

**Stickler’s Giant Retinal Tear**

- GRT
- PFO
- SB optional, I use it whether phakic or pseudophakic
- Clean vitreous poster to retina, be careful of long posterior ciliary nerve and artery when lasering
- Gas or SO
Cavitary malformations of the optic nerve
- Coloboma
- Morning glory syndrome
- Peripapillary staphyloma
- Abnormalities in the optic nerve
  - optic disc hypoplasia
  - acquired pit
Congenital Optic Nerve Head Disorders

Slide by Baseer Ahmad
Coloboma

- 35 yo machinist c/o acute Va loss right eye
- Va RE = 20/80- LE = LP
- H/o bilateral colobomas
Coloboma associated RRD

- Incomplete closure of optic fissure
- 0.5:10,000
- RRD 8-40%
- CHARGE
- CES
- Patau Syndrome

See poster by Priyanka Kumar

Coloboma associated RRD

- Tears/holes/breaks are variable
- Iris coloboma think tear at 6:00
- Unless peripheral break obvious, laser around coloboma as there may be intercalary breaks
- Possible to SB alone if you identify peripheral breaks (but don't recommend that to friends)

Patient History

- “Referred for swelling, may need laser” OS
- 58 yo C man
- PMH:
  - HTN, arthritis

Examination

<table>
<thead>
<tr>
<th>VA</th>
<th>20/20</th>
<th>P</th>
<th>3</th>
<th>Ant Seg</th>
<th>2</th>
<th>IOP</th>
<th>15</th>
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<td>K</td>
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<tr>
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<td>Iris</td>
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<tr>
<td>D/O</td>
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<td>AC</td>
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<td></td>
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<tr>
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<td></td>
<td>Lens</td>
<td></td>
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</tbody>
</table>

Courtesy N. Steinle/D. Dhoot
Optic Nerve Pit

- Incomplete closure of superior edge of embryonic fissure
  - 1:10,000
  - 10-15% bilateral
  - Macular Schisis and/or SRF
  - Temporal but 1/3 central
Little doubt that SRF comes from vitreous cavity
- PPV with gas 80% effective
- Can lightly laser peripapillary region
- High rate of contralateral disc abnormality

Possibly incomplete closure of the embryonic fissure vs. mesenchymal failure
- Differs from disc colobma as there are radial emanation of blood vessels
- Periodically open or close (contractility)
Conclusion

- Likely see adult ROP
  - Cataract surgery not trivial
  - Can a SB work alone
  - NVI happens
- FEVR
  - Distinguish exudation from traction
  - NVI happens
- Coats
  - Prior cryo may necrose retina
- Coloboma
  - Mx break patterns
  - Iris coloboma think HST anterior edge of coloboma
  - Laser around the coloboma
- MGD
  - If rhegmatogenous consider hole on ONH

Morning Glory Disc

- Think hole on optic nerve head
- But remember RD can be exudative
- Moya moya

MGD associated RD

VHL
PPV

- PDT

VHL

Diode laser
Continued exudative RD
<table>
<thead>
<tr>
<th>VHL</th>
<th>VHL</th>
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<tbody>
<tr>
<td>SB, PPV, SO, EL</td>
<td>F/U 7 weeks</td>
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<tr>
<td>VA 20/400</td>
<td>Laser lesions when small, cryo peripheral lesions, PDT for posterior, larger exudative lesions but follow closely</td>
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<table>
<thead>
<tr>
<th>VHL</th>
<th>Hypotony</th>
</tr>
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<tbody>
<tr>
<td>Increased outflow</td>
<td>Decreased Secretion</td>
</tr>
<tr>
<td>Retinal detachment</td>
<td>Cyclic membrane</td>
</tr>
<tr>
<td>Retinectomy</td>
<td>Capsular contraction</td>
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<td>Glaucoma procedures</td>
<td>Congenital disorders</td>
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<tr>
<td>Cyclodialysis cleft</td>
<td>PVR</td>
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<tr>
<td>Trauma</td>
<td>PVR Surgery</td>
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<td>ACL explantation</td>
<td>Ischemia</td>
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<td>Surgery</td>
<td>Uveitis</td>
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<tr>
<td></td>
<td>infectious</td>
</tr>
<tr>
<td></td>
<td>noninfectious</td>
</tr>
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</table>
Causes of Hypotony

- Decreased Secretion
  - Tractional forces on the anterior vitreous base leads to CB detachment
  - IOP reduction associated with two clock hours of ciliary body detachment.

- Increased Outflow
  - Cyclodialysis cleft
    - 4/17 eyes in prospective study of CBE delamination had a cyclodialysis cleft
    - Direct cyclopexy diagnosed with UBM

Cyclodialysis Clefs

Purpose: To determine the efficacy of UBM and radial cyclopexy.

Methods: Retrospective, interventional study of 18 consecutive patients with hypotony (IOP < 6mmHg) of unknown etiology who underwent ultrasound biomicroscopy (UBM).

Inclusion criteria: IOP <6 mmHg, record of UBM
Exclusion criteria: glaucoma surgery, concomitant RD, adjunctive intravitreal tamponade, cryocyclopexy, laser cyclopexy

Radial Sutures

Overlap edge of CDC with one suture
**Results:**

-55% (10/18) of patients were diagnosed with a CDC by UBM. Gonioscopy was helpful in only one of these patients. 5 pts had definite cleft, 5 questionable cleft.

-13 patients underwent surgery, 10 cyclopecty, 2 epipillary delamination, 1 IOL exchange.

-38% (5/13) increased intraocular pressure average 19 mmHg (range 6-28 mmHg) by cyclopecty alone.

-2/13 increased IOP (average 9 mmHg) but cannot directly attribute result to cyclopecty (IOL ex/CB MS).

-Average follow-up was 11.4 months (range 1-36 months).

---

**Table:**

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Hx</th>
<th>Etiology of Hypotony</th>
<th>Duration of Hypotony</th>
<th>Cleft by UB: y/n</th>
<th>Additional Findings by UB</th>
<th>Surgical Procedure</th>
<th>IOP preop</th>
<th>IOP postop</th>
<th>F/U (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>40</td>
<td>M</td>
<td>Blunt trauma</td>
<td>Traumatic</td>
<td>1 month</td>
<td>Yes</td>
<td>CB detachment and cleft at 11:00</td>
<td>Radial sutures and PCL</td>
<td>3</td>
<td>17</td>
<td>18 months</td>
</tr>
<tr>
<td>48</td>
<td>M</td>
<td>PDR</td>
<td>360</td>
<td>12 months</td>
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<td></td>
<td></td>
<td></td>
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<tr>
<td>60</td>
<td>F</td>
<td>RRD/PVR</td>
<td>C-4,5</td>
<td>11 months</td>
<td>No</td>
<td>CB detachment and possible Cleft</td>
<td>Radial sutures and PCL</td>
<td>0</td>
<td>15</td>
<td>6 months</td>
</tr>
</tbody>
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**Results:**

-Characteristics of successful outcomes:
  - definite identification of cleft (p=.02, .05).
  - hypotony less than 3 months (p=.006, .009).

-Characteristics of unfavorable outcomes:
  - h/o infectious uveitis (p=.02).
Conclusions:

- Hypotony is a bad problem with few solutions.
- Only definite identification of cleft by UBM warrants surgery.
- Patients with hypotony greater than 3 months and/or h/o of infectious uveitis may be poor candidates.
- Radial cyclopegy may be simple and effective.

Likely see adult ROP
- Cataract sx not routine
- Can a SB work alone?
- NVI happens
- FEVR
- Distinguish exuafion from traction
- Coats
- Prior cryo may necrose retina

Coloboma
- Mx break patterns
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