PEDIATRIC VITREORETINAL SURGERY
ROP, FEVR, COATS, PFV....

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PEDIATRIC VITREORETINAL SURGERY

- Its own rules
- Its own complications
- Does not forgive faults
- Does not accept apologies
Very small eye: Narrow space for maneuvers (15-19mm)

Lens is relatively large

Vitreous-Retina relation!

Pars plana has not developed.

Sclera is thin and elastic
PEDIATRIC VRS Features

- Retinal breaks usually ends up with surgical failure
- PFCL and Silicon oil are rarely used.
- Children do not obey the head positioning!
- PVR is frequent and severe!
- Amblyopia problem!
VITRECTOMY
Sclerotomy Construction

- Pars plicata, Trans iris-root, Limbal
- 2-3 port entry
- 20G-23G-25G-27G surgeries
- Lens sparing vitrectomy (LSV) if possible!
- Lensectomy-Vitrectomy
# Sclerotomy

## Data Used to Plan Sclerotomy Location

<table>
<thead>
<tr>
<th>Age</th>
<th>Minimum Ciliary Body Length (Aiello et al.(^1))</th>
<th>Minimum Limbus-to-Ora Serrata Distance</th>
<th>Limbus-to-Sclerotomy Distance</th>
</tr>
</thead>
<tbody>
<tr>
<td>0–6 mo</td>
<td>2.60 mm</td>
<td>2.95 mm</td>
<td>1.45 mm, 1.5 mm(^\dagger)</td>
</tr>
<tr>
<td>6–12 mo</td>
<td>2.86 mm</td>
<td>3.21 mm</td>
<td>1.71 mm, 2.0 mm</td>
</tr>
<tr>
<td>1–2 yr</td>
<td>3.28 mm</td>
<td>3.63 mm</td>
<td>2.13 mm, 2.5 mm</td>
</tr>
<tr>
<td>2–3 yr</td>
<td>3.75 mm</td>
<td>4.10 mm</td>
<td>2.60 mm, 3.0 mm(^\dagger)</td>
</tr>
<tr>
<td>Adult</td>
<td>4.60 mm</td>
<td>4.95 mm</td>
<td>3.45 mm, 3.5 mm(^\mathsection)</td>
</tr>
</tbody>
</table>

At the very beginning...

Narrow space can be widened with Canthothomy
When Lenscetomy is Planned

1. Iris root entry: Iris problems

2. Limbal entry:
   - Corneal distortion, endothelial damage
   - 10/0 sutur needed (EUGA for removal)

Advantages:
   - Chantotomy not needed
   - Conjunctiva is protected
   - Peripheral retinal damage is avoided
Choosing Sclerotomy Sites

- Conjunctival peritomy
- Avoid damage to anterior fibrovascular tissue and retina
- Suture all sclerotomies
• Adult type instrumentation, with/without trockars
• Infusion cannula: 23G sutured (4mm)
ROP

STAGE 4A

STAGE 4B

STAGE 5

SCATRICIAL
23G-with cannula
Stage 4a ROP

Prof. Dr. Şengül Özdek
Stage 4A-without trockars
Stage 4B
(Fellow eye stage 5)
• This is the most typical surgery where the surgeon should know where to stop!

• “Perfect” is the enemy of “good”

• Do not let any iatrogenic retinal break!
If a retinal break occurs...

• In a silent area... continue with standard techniques

• In the most proliferative area... abandon the surgery
SURGERY FOR STAGE 5

• SURGERY?
• VERY LOW VISUAL EXPECTATIONS
• PRESERVATION OF LIGHT PERCEPTION/AMBULATORY VISION???
• ANATOMICAL PURPOSES
• IF FELLOW EYE IS GOOD…..MOSTLY NOT OPERATED
Stage 5
SURGERY FOR ROP
To decrease risk of post op hemorrhage

- Preop avastin to eyes with plus disease?
- Do not let hypotony at the end of surgery.
- F-A-X
- Fill with Viscoelastics?
### Surgery for Cicatricial ROP

<table>
<thead>
<tr>
<th>Olgu/Cinsiyet/Göz</th>
<th>Doğum Haftası/Kilosu (hft/gr)</th>
<th>Cerrahi Yaşı (ay)</th>
<th>Preop GKY</th>
<th>Preop Tedavi</th>
<th>Cerrahi</th>
<th>Komplikasyon</th>
<th>Takip Süresi (ay)</th>
<th>Anatomik Başarı</th>
<th>Maküler katlanı Preop/Postop</th>
<th>Postop GKY</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/E/Sol</td>
<td>28/1060</td>
<td>6</td>
<td>-</td>
<td>-</td>
<td>PPV+PPL</td>
<td>-</td>
<td>24</td>
<td>Kismi Başarı</td>
<td>+/-</td>
<td>-</td>
</tr>
<tr>
<td>2/K/Sol</td>
<td>25/650</td>
<td>38</td>
<td>IOT(-)</td>
<td>LFK</td>
<td>PPV</td>
<td>-</td>
<td>5</td>
<td>+</td>
<td>-/-</td>
<td>1mps</td>
</tr>
<tr>
<td>3/E/Sol</td>
<td>29/1100</td>
<td>59</td>
<td>PPEH</td>
<td>LFK</td>
<td>PPV+PPL</td>
<td>İatrojenik delik</td>
<td>1</td>
<td>-</td>
<td>-/-</td>
<td>PPEH</td>
</tr>
<tr>
<td>4/K/Sol</td>
<td>30/1370</td>
<td>10</td>
<td>IOT(-)</td>
<td>-</td>
<td>PPV</td>
<td>-</td>
<td>3</td>
<td>Kismi Başarı</td>
<td>+/+</td>
<td>IOT(+)</td>
</tr>
<tr>
<td>5/K/Sol</td>
<td>28/1500</td>
<td>11</td>
<td>IOT(-)</td>
<td>-</td>
<td>PPV+PPL</td>
<td>-</td>
<td>20</td>
<td>+</td>
<td>+/-</td>
<td>0.1</td>
</tr>
<tr>
<td>6/E/Sağ</td>
<td>28/850</td>
<td>79</td>
<td>0.2</td>
<td>-</td>
<td>PPV</td>
<td>-</td>
<td>40</td>
<td>+</td>
<td>+/-</td>
<td>0.7</td>
</tr>
<tr>
<td>6/E/Sol</td>
<td>28/850</td>
<td>81</td>
<td>0.05</td>
<td>-</td>
<td>PPV</td>
<td>-</td>
<td>40</td>
<td>+</td>
<td>+/-</td>
<td>0.3</td>
</tr>
<tr>
<td>7/K/Sağ</td>
<td>28/1800</td>
<td>4</td>
<td>IOT(-)</td>
<td>-</td>
<td>PPV</td>
<td>-</td>
<td>24</td>
<td>+</td>
<td>+/-</td>
<td>0.05</td>
</tr>
<tr>
<td>8/K/Sağ</td>
<td>30/1290</td>
<td>104</td>
<td>1mps</td>
<td>-</td>
<td>PPV</td>
<td>-</td>
<td>6</td>
<td>Kismi Başarı</td>
<td>+/+</td>
<td>0.05</td>
</tr>
<tr>
<td>9/K/Sağ</td>
<td>28/990</td>
<td>2</td>
<td>IOT(-)</td>
<td>LFK+IVB</td>
<td>PPV+PPL</td>
<td>-</td>
<td>19</td>
<td>Kismi Başarı</td>
<td>-/-</td>
<td>IOT(-)</td>
</tr>
<tr>
<td>9/K/Sol</td>
<td>28/990</td>
<td>2</td>
<td>IOT(-)</td>
<td>LFK+IVB</td>
<td>PPV+PPL</td>
<td>Geç Vit Hem/RRD</td>
<td>19</td>
<td>-</td>
<td>-/-</td>
<td>IOT(-)</td>
</tr>
<tr>
<td>10/K/Sağ</td>
<td>28/900</td>
<td>213</td>
<td>1mps</td>
<td>-</td>
<td>PPV+PPL</td>
<td>-</td>
<td>12</td>
<td>Kismi Başarı</td>
<td>+/+</td>
<td>1mps</td>
</tr>
</tbody>
</table>
6.5 y, M, VA: 0.1/0.15
28 wk 1300 gr, No treatment before
Cicatricial ROP
Postop 3rd year
VA: 0.7
Postop 3rd year: VA: 0.3
Cicatricial ROP Surgery

- VRS may lead to very good anatomical and functional results in some of scatricial ROP cases
Mostly AD inheritance

NDP, FZD4, LRP5, TSPAN12, ZNF408: (Wnt-NORRIN signal pathway) defective genes

Retinal angiogenesis is defective: Vascular differentiation is insufficient

Peripheral retinal vascularization is incomplete

21-64% RD: Tractional or exudative
FEVR-Treatment

- LASER
  - STAGE 1: Peripheral avascular retina
  - STAGE 2: Retinal NVE
- SURGERY
  - STAGE 2: Retinal NVE-Vit Hem
  - STAGE 3: Macula on RD (macula ectopic)
  - STAGE 4: Macula off RD (macular fold)
  - STAGE 5: Total RD
FEVR-Treatment
6y old, F, bilat TRD, VA: 0.05
FEVR-VRS
VA: Preop: 0.05 Postop: 0.4
FEVR-Treatment
VA: Preop: 0.05 Postop: 0.3
COATS’ DISEASE
COATS DISEASE

- Endothelium and pericyte disease: Inner blood retina barrier defect
- Exudative (Tractional) RD
1. ONLY TELANGIECTASIS

2. TELANGIECTASIS & EXUDATION
   A. Extrafoveal Exudation
   B. Foveal Exudation

3. + EXUDATIVE RD
   A. subtotal
   B. total

4. + NVG

5. END STAGE DISEASE (Painful red eye)
COATS-Treatment

- Primary Treatment: Laser (Stage 1,2,3)
- Cryotherapy: Stage 3
- Surgery: Stage 3-4
  - External Drainage Cryo or Laser
  - Anti-VEGF or Ozurdex
  - PPV-External Drainage
  - Retinotomy for internal drainage should absolutely be avoided!
**COATS Treatment**

**End stage**

- Surgery to preserve the globe in advanced cases (for cosmesis and comfort)

- 16% of eyes in Coats disease are enucleated because of painful eye.
9 mo old boy, Stage 4 Coats Drainage Cryo
9 mo old boy, Stage 4 Coats Drainage Cryo
Stage 4 Coats

4y, girl, Stage 4, NVG, red eye

3 mo later
Persistent Fetal Vasculature
PFV

Anterior
Posterior
Mixed
Combined with other retinal pathologies
Radial iris vessels (hairpin-loops)
Goals of Surgery for PFV

To get acceptable anatomical and functional results

- Clear the media in order to prevent amblyopia
- Relieve tractional forces
  - To prevent TRD, glaucoma, Phthisis bulbi
  - To let the eye to have the opportunity to grow
Anatomical differences in Anterior PFV

- PP not developed,
- No zonules,
- Elongated ciliary processes

Anterior fibrovascular structure may be continuous with retina!
PFV-Bad surprise!
PFV-anterior retinal elongation
Tips: Pigmented plaques
Nasal and inferior location
Total excision vs leaving some part of FV tissue?

FV tissue contracts and causes pupillary obliteration or Peripheral RD within years leading total TRD
Leaving peripheral part of FV tissue....pupillary obliteration
Patients and Methods

- 29 eyes of 28 patients with PFV (last 6 years)
- 19 (67.1%) were male
- 14 eyes (47.7%): Anterior PFV
- 10 eyes (34.5%): Posterior PFV
- 5 eyes (17.2%): Mixed type
- Median follow-up: 14 months (6 months to 5.5 years)
Patients and Methods

- Anterior retinal elongation: 64.3% of anterior PFV.
- 84.6% of anterior PFV cases resulted in anatomic success.
- Final VA: 20/200 or better in 38.5% of anterior PFV cases.
# Complications

## Table 2. Complications

<table>
<thead>
<tr>
<th></th>
<th>Retinal complications</th>
<th>Anterior segment complications</th>
<th>p-value'</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Yes (%)</td>
<td>No (%)</td>
<td>Univariate</td>
</tr>
<tr>
<td>Extend of lens opacity (n=19)*</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Less than half (n=8)</td>
<td>0 (0)</td>
<td>8 (100)</td>
<td>0.018'</td>
</tr>
<tr>
<td>More than half (n=11)</td>
<td>6 (54.5)</td>
<td>5 (45.5)</td>
<td></td>
</tr>
</tbody>
</table>

*includes anterior and combined cases.

'Fischer exact test.

'OLS
PFV patients who have more extensive anterior disease tend to have a higher risk of overall complications than patients with localized fibrovascular tissue.
PFV-mixed
Anterior PFV-Take home messages

• Aware of peripheral retinal extensions through pars plana esp in Ant PFV.

• Do not leave peripheral vascular fibrotic tissue during surgery: May cause severe complications

• Be prepared for RD surgery.
CONGENITAL X-LINKED RETINOSCOPYSIS
CXLRS
CXL R

- Males
- Bilateral, peripheral+ foveal retinoscisis
- Recurrent vitreous hemorrhages
- RRD
8 mo old, M, Bilateral Vit Hem,
21y, M, VA: 0.3 (LE)
Bilat PPV 15 yr earlier, RE: FB
Postop 5 mo
VA: 0.4
PEDIATRIC TRAUMA
TRAUMA
Penetrating Injury

- Delayed diagnosis,
- Risk of Endophthalmitis high
- Wound healing: PVR risk high
- Follow up examination are really difficult, assessment is hard.
- Ambliopia!
SHAKEN BABY
RETINAL HEMANGIOBLASTOMA
14 y old, Cerebellar hemangioma (operated)
VA: HM
Fundus: Vit Hem-TRD
RETINAL HEMANGIOBLASTOMA
3 years postop
RETINAL HEMANGIOBLASTOMA
Follow ups with FA every 4-6 months
SURGERY IS AN ART

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