Surgical Treatment of Pulmonary Vein Stenosis

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Surgery of the Pulmonary Veins
Repair of

- PAPVR / Scimitar syndromes
- Total anomalous pulmonary venous return
- Pulmonary vein stenosis, hypoplasia

TAPVR

A • Supracardiac
  • Infracardiac
  • Mixed
  • Cardiac

B • Obstructed, unobstructed

C • Biventricular heart
  • Single ventricle (RAI)

Repair of TAPVR - Pulmonary Vein Stenosis

- Repair by connecting pulmonary venous confluence to left atrium
- Pulmonary vein stenosis
- Unilateral, bilateral
- 1-4 veins
- Causes:
  1. Technical
  2. Exuberant inflammatory fibrotic response
  3. Primary developmental process
**Pulmonary Vein Stenosis**

- Following repair of TAPVR
  - 5-15% of repaired TAPVR
    (references: Lacour-Gayet, Zoghbi, Serraf, et al., 1998; Devaney, Chang, Olpe, Bove, 2006; Karamlou, Gurofskyk, Al Sukhni, et al., 2006)
  - Mortality – up to 50%
  - High morbidity
  - Increased risk in single ventricle, RAI/heterotaxy, infracardiac type, hypoplastic veins

**Pulmonary Vein Stenosis - Pathology**

- Intimal hyperplasia
- Medial hypertrophy
- Periadventitial fibrosis
- Intraparenchymal hypoplasia
- Intraparenchymal obstruction
- Lymphatic ectasia

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**Medial hypertrophy**

**Intimal proliferation**
Post-Repair Pulmonary Vein Stenosis

- **Repair strategies**
  - Stenting
  - Patch repair
  - Revision
  - High morbidity, mortality, failure

Sutureless Repair

- Introduced by Lacour-Gayet, Coles
- Incision / excision of stenotic region
- Marsupialization with pericardial flaps
- Posterior mediastinal scarring prevents “bleeding” into pleural pericardium
- Care taken around phrenic nerve

Post-Repair Pulmonary Vein Stenosis

- **Sutureless repair**
  - Better outcomes
  - Small number of cases
    - Michigan: \( n = 11 \)
    - Toronto: \( n = 35 \) (25 primary)
    - Denver/Paris: \( n = 23 \)
Risk Factors for Recurrent / Persistent Pulmonary Vein Stenosis
1. Single ventricle – RAI, heterotaxy syndrome
2. Orificial stenosis – discrete
3. Pulmonary vein hypoplasia
4. Nonsutureless repair technique

TAPVR
Post-repair of TAPVR
Primary (developmental, programmed)
Post-operative (non-pulmonary vein surgery, inflammatory, fibrosis)

Primary Sutureless Repair
Adopted as a strategy for patients where the risk of recurrent stenosis is higher:
- Single ventricle, RAI
- TAPVR infracardiac Christmas tree patterns
- Primary pulmonary vein hypoplasia, stenosis
- “Danger” bleeding into pleural space because of lack of posterior mediastinal adhesions

Primary Sutureless Repair for TAPVR / Pulmonary Vein Stenosis – Technique 1
- Incision into pulmonary veins, beyond stenosis, up to parenchyma / pleural reflection
- Sew left atrial cuff or pericardial flap posterior to mediastinum / pleura
Primary Sutureless Repair for Primary Pulmonary Vein Stenosis – Technique 2

UCSF Experience – Primary Sutureless Repair of Pulmonary Veins (n = 13)

- 6 RAL, single ventricle, pulmonary atresia
- 3 Primary pulmonary vein stenosis
- 4 Hypoplasia of 1 or more pulmonary veins in TAPVR
- No operative/surgical mortality
- Median follow-up 15 months (4-30 months)
- 3 Single ventricle patients progressed, 2nd stage palliation
- Out of 52 veins – recurrence at 3 veins of 2 patients

Primary Sutureless Repair: Primary Pulmonary Vein Stenosis

- 6 month old infant
- Premature, low birth weight
- Primary pulmonary vein stenosis
- Ventricular septal defect, patent ductus arteriosus

[video]
Primary Sutureless Repair: Post Surgical - Case Study

- 6 year old
- Pulmonary atresia, intact ventricular septum
- s/p Blalock-Taussig shunt, s/p bidirectional cavopulmonary anastomosis
- Cath/MRI/echo: hypoplastic left pulmonary veins, orificial stenosis
- Primary sutureless repair (pericardial flap) with Blalock-Taussig shunt
- Saturations 60’s -> 80’s postoperatively

References


Primary Sutureless Repair: Post Surgical - Case Study

- TEE of left pulmonary vein flow pattern:

  Pre-operative

  Post-operative

\[ \text{\text{Image of TEE}} \]