Surgical Interventions for Pulmonary Vein Stenosis

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Clinical problem

Surgical repair of TAPV

TAPV repair:
Strong era effect – We are getting better!

Karamlou, Circulation 2007
TAPV repair: Still a problem in ‘unfavorable patients’

Karamlou, Circulation 2007

TAPV repair: Early hazard phase for reoperation

Karamlou, Circulation 2007

TAPV repair: Risk Factors for Reoperation

High risk population
Small PV confluence
Mixed type
Small patients (<2 kg)
Right atrial isomerism

Yun, JTCVS 2005

Residual stenosis predicts reop
Diagnosis: Post-repair pulmonary vein stenosis

Post-repair Pulmonary Vein Stenosis
Three varieties
• Anastomotic stenosis
• Fibrosis of confluence
• Retrograde ‘UPSTREAM’ progression into lungs
  – Unilateral or bilateral
  – Often relentless

Surgical evaluation: Post-repair pulmonary vein stenosis

Who is operable?
Extent of disease can predict survival:
PVS Score

- Grading of PVS in each pulmonary vein
  0: None
  1: Mild-Moderate
  2: Severe
  3: Occlusion

- Summation of PVS grades of 4 PVs
- Possible range: 0 - 12

PVS Score

PVS Score

‘Upstream’ pulmonary veins

(Not here)
Operate HERE!
Use a clinical surveillance protocol
to detect recurrence early! 1, 6, 12 months (at least)
Congenital PVS

PR-PVS and congenital PVS

Surgical treatment for PR-PVS: Sutureless repairs
John Coles’ Hypothesis

Irregular contour >> geometric distortion which is ‘fixed’ by a sutureline

- Distortion
- Flow disturbance
- Local injury
- Upstream propagation

John Coles’ Hypothesis

- ‘Sutureless repair’
- Atrium to pericardium
  - No direct anastomotic suture
  - “Controlled bleed” into neoatrium
- No geometric constraints on tissue
  >>>> Tissue conforms to local flow patterns

Sutureless Repairs: A Controlled Bleed
Post-repair PVS

- Sutureless repair
- Conventional

\[ p = 0.04 \]

Time (yrs)

Survival without re-repair (%)

- Technique
- Other
- SPM

Months

Post-repair PVS

Mixed TAPVD

- Simplifies complex geometry

Survival

Log-rank: \( p = 0.07 \)

- Sutureless
- Overall
- Conventional

Months from surgery

If beneficial for PR-PVS, should we use sutureless repairs for all TAPV patients?

> Prophylactic procedure
Primary sutureless repair for TAPV

Positive Attributes:
- Less precision required >> EASY
- Short learning curve
- Easier to teach
- No circulatory arrest

Negative Attributes:
- Broncho-atrial fistula?
- Pleural disruption?
- Embolism?
- >>> Uncommon
Primary sutureless repair for TAPV

- Shi et al
- n=768
- Lower restenosis in patients with preop PVS

Conclusions: Sutureless repairs

- Sutureless repairs for treatment of post-repair pulmonary vein stenosis
  >> ‘superior’ midterm results

- Sutureless repairs for prevention of PVS
  - Less clearly supported, but little downside
  - Recommended for:
    • patients at high risk of recurrence
    • mixed TAPV

Primary sutureless repair for TAPV

Can we prove there is a benefit?

Sample size requirement
- Assume 50% decrease PR-PVS
- 150 patients (10 years of accrual) needed

- High risk populations >> smaller sample size
- Registry (e.g. PVS Network) will enable
- Low risk of adverse events makes it attractive

Surgical treatment for PR-PVS: Lung transplant

TAPVD → Surgery → PR-PVS?

10-15%

No

Yes

Congenital PVS

?
Lung transplant and PVS

Lung transplant is a viable treatment option for patients with congenital and acquired pulmonary vein stenosis

Ankit Bharat, MD,6 Deirdre J. Epstein, RN,5 Mark Grady, MD,5 Albert Faro, MD,2 Peter Michelson, MD,2 Stuart C. Sweet, MD,6 and Charles B. Huddleston, MD6

- 12 congenital PVS + 8 acquired PVS
- Median wait: 26d
- Bilateral lung transplants on CPB

5 year survival: 60%
BO-free survival ~50%
Preop ECMO is risk factor*

Waiting for transplant

- Mechanical Pulmonary Support
  - ECMO

- Rapidly evolving techniques
  - RA>LA (pump + oxygenator, special cannula)
  - PA>LA (oxygenator)

Dual lumen ECMO catheter
RA > LA
Paracorporeal lung assist device (PLAD)  
PA > LA

- Stabilize awaiting transplant
- Enable therapies in non-hypertensive lungs
- Enable surgical interventions

Conclusion

- Surgery is not enough

- Immediately achievable:
  - Match surveillance to velocity of disease
    - Detect while still treatable
    - Use Clinical Surveillance Protocol (1,6,12 months)
    - Use dedicated "PVS Team" to manage surveillance and treatment

- Our future
  - Combination therapy offers opportunities
    - Integrate catheter-based <> surgical interventions
    - Mechanical pulmonary support <> transplant <> MPS-enabled therapies
    - Medical adjuncts (losartan, gleevec, avastin)