Pulmonary Vein Stenosis

Yun, Tae-Jin

Asan Medical Center, University of Ulsan
DEPARTMENT OF CARDIOVASCULAR SURGERY – July 1, 2002 to June 30, 2003

Left to Right: Dr. Tae-Jin Yun, Dr. Randall Fortuna, Dr. Julia Ritter, Dr. Nilton Carias De Oliveira, Dr. Glen Van Arsdell, Dr. John Coles, Dr. Harold Burkhart, Dr. William Williams, Dr. David Ashburn
Pulmonary Vein Stenosis

• Etiology: Acquired vs. Congenital
• Classification
• Indications for intervention
  : For individual vein (diffuse disease, atresia..)
  : For patients
• Surgical techniques
  : Excision vs. Incision
    Sutureless Repair vs. Conventional technique
Excision vs. Incision

A: PVS

B: Incision of PV

C: Excision of PV
Sutureless technique for repair of pulmonary vein stenosis
- Extension of indications -

Yun TJ, Coles JG, Konstantinov IE, Wald RM, Guerra V, Van Arsdell GS, Williams WG, Smallhorn J, Caldarone CA

The Hospital for Sick Children in Toronto
Sutureless Repair

A

B

C

SVC
AO
IVC
Sutureless repair (SR)
- Theoretical advantage -

- Avoid geometric distortion
- No suture material on PV
- Avoid restriction of ostial growth
Objectives

Evaluate Sutureless Repair (SR):

- Post-repair PVS (After repair of TAPVD)
- Efficacy of SR for other indications
- Safety of SR without retrocardiac adhesion
SR without Retro-cardiac adhesion
SR without Retro-cardiac adhesion
SR without Retro-cardiac adhesion
Methods

60 PVS patients over 20 years
73 procedures (40 Sutureless Repairs)
Age: 7 d – 38 m (4.4 m)

Follow-up: 1 m – 18.6 yrs (2.9 yrs)
Complete: 88%
Statistical Analysis

Retrospective analysis

Cox Proportional Hazard Survival Model

Endpoint: Re-operation or death

Variables:
- Age
- Indication category
- Extent of disease (PVS score)
- Heterotaxy syndrome
- Types of intervention
- Previous cardiac operation

Model validation with bootstrap analysis
Individual pulmonary vein stenosis graded

**PVS score**

Summation of four PV grades

0: No PVS
1: Mild-Moderate PVS
2: Severe PVS
3: Obstruction

Possible range: 0 - 12
PVS score

![Histogram of PVS scores with bars representing the frequency of scores ranging from 0 to 10. The histogram shows a peak at score 2, with other scores having varying frequencies.]
60 Patients
60 Patients

Post Repair (PR)-PVS (n=17)
PR-PVS (n=17)

- Sutureless repair (SR) 5 (0)*
- Patch pulmonary venoplasty (PVP) 6 (5)*
- Ostial endovenectomy (OE) 2 (0)*
- Stent 3 (2)*
- Atrial PVP 1 (1)*

* Reop or death
60 Patients

‘Naïve’ PVS (n=36)

PR-PVS (n=17)
60 Patients

‘Naïve’ PVS (n=36)

PR-PVS

Postop-PVS (n=10) Unoperated-PVS (n=26)
60 Patients

‘Naïve’ PVS (n=36)

PR-PVS (n=17)  Postop-PVS (n=10)  Unoperated-PVS (n=26)

Sutureless repair (SR)  5 (2)*
Patch pulmonary venoplasty (PVP)  3 (1)
Ostial endovenectmy (OE)  1 (0)
Stent  1 (0)
Atrial PVP  0 (0)

* Reop or death
60 Patients

'Naïve' PVS (n=36)

PR-PVS (n=17)  PO-PVS (n=10)  Unoperated-PVS (n=26)

Sutureless repair (SR)  18 (6)*
Patch pulmonary venoplasty (PVP)  1 (1)
Ostial endovenectomy (OE)  0 (0)
Stent  3 (2)
Atrial PVP  4 (1)

* Reop or death
60 Patients

- 'Naïve' PVS (n=36)
  - PR-PVS (n=17)
  - PO-PVS (n=10)
  - Unoperated-PVS (n=10)
  - Small PV (n=7)
60 Patients

‘Naïve’ PVS (n=36)

- PR-PVS (n=17)
- PO-PVS (n=10)
- Unoperated-PVS (n=26)
- Small PV (n=7)

- Sutureless repair (SR) 7 (3)*
- Patch pulmonary venoplasty (PVP) 0 (0)
- Ostial endovenectomy (OE) 0 (0)
- Stent 0 (0)
- Atrial PVP 0 (0)

* Reop or death
73 Procedures
73 Procedures

1st PV procedure

- SR (n=35)
- Patch PVP (n=10)
- OE (n=3)
- Stent (n=7)
- Atrial PVP (n=5)
73 Procedures

1st PV procedure:
- SR (n=35)
- Patch PVP (n=10)
- OE (n=3)
- Stent (n=7)
- Atrial PVP (n=5)

2nd PV procedure:
- SR (n=3)
- Death (n=7)
- Stent (n=2)
73 Procedures

1st PV procedure

- SR (n=35)
- Patch PVP (n=10)
- OE (n=3)
- Stent (n=7)
- Atrial PVP (n=5)

2nd PV procedure

- SR (n=1)
- Death (n=6)
- Stent (n=2)
73 Procedures

1st PV procedure
- SR (n=35)
- Patch PVP (n=10)
- OE (n=3)
- Stent (n=7)
- Auricle PVP (n=5)

2nd PV procedure
- Death (n=4)
- Stent ballooning (n=2)

3rd PV procedure
- Stent ballooning (n=1)
73 Procedures

1st PV procedure

- SR (n=35)
- Patch PVP (n=10)
- OE (n=3)
- Stent (n=7)
- Auricle PVP (n=5)

Death (n=1)

2nd PV procedure

- SR (n=1)
- Death (n=1)
- Stent (n=1)
PV Stenosis Repair; \{n=60; HSC\}

Freedom from death

Freedom from reop or death

Time (yrs)
Statistical Analysis

By multivariable analysis

Higher PVS score was associated with increased risk of re-operation or death

HR 12.9 for PVS score from 2-7   p=0.0001

Model validation failed bootstrap analysis
PVS score

PVS score: 0-3

PVS score: 4-7

PVS score: 8-12

P<0.0001
Statistical Analysis

After adjustment for PVS score, the sutureless repair was associated with decreased risk of re-operation or death

HR 0.47  p=0.12

This effect was greatest in the patients with PVS after repair of TAPVD
Post-Repair PVS

Time (yrs)

Freedom from reop or death

Sutureless repair

Conventional

p=0.04
SR without Retro-cardiac adhesion
SR without Retro-cardiac adhesion
Summary (1)

Sutureless repair (SR) for Post-repair PVS is associated with excellent mid-term results.

PVS score is a significant risk factor for re-operation or death
Sutureless repair without retrocardiac adhesion appears safe with development of a simple hemostatic maneuver.

The efficacy of the sutureless repair in patients with indications other than PR-PVS is not well defined due to the small number of patients in the present report.

Consequently, a registry-based approach to evaluation would be expedient.
SR for TAPVD with small PV/CPVC
Right isomerism with TAPVD

Yun TJ, Osman Al-Radi, Caldarone CA, Coles JG, Williams WG, Smallhorn J, Van Arsdell GS

The Hospital for Sick Children in Toronto
Right atrial isomerism (RAI)

- Failure of lateralization with bilateral right-sidedness
- Centrally squeezed primary PV with abnormal PV drainage
- Dismal prognosis
Management and Outcome of RAI: A 26 year experience
(Hashmi et al, JACC 1998)

- The Hospital for Sick Children in Toronto
- 91 RAI patients between 1970 and 1996
- Cardiac abnormality
  AVSD (81%), FSV (73%), PS (84%)
  TAPVD (87%, 1/3 obstructive)
- No Tx in 24% of patients (with 95% mortality)
- Overall mortality: 69%
- Surgical mortality for TAPVD repair: 95%
Surgical management of TAPVD: Impact of coexisting cardiac anomalies (Caldarone CA et al, ATT 1998)

- The Hospital for Sick Children in Toronto
- 170 TAPVD pts between 1982 and 1996
- 44 complex TAPVD
- Op. mortality of complex TAPVD: 52%
- Risk factors for Op. mortalities:
  - FSV (P=0.03)
  - Associated complex anomalies (P<0.01)
TAPVD association in RAI

Incidence: 60% - 100%
(Varies according to the definition of TAPVD)

Outcome variables:

1. Drainage site obstruction
2. Individual PVS
3. Presence or absence of TAPVD
4. Types of TAPVD: Cardiac type?
Objective

Premise:
RAI associated with TAPVD is fatal

Question:
Can we modify the outcome by aggressive Tx of TAPVD?
Methods

55 Patients of RAI over 10 years

Patient enrolled: All patients of RAI
Including
1) Patients without Surgical Tx
2) Patients without TAPVD repair
3) Patients with TAPVD repair
Statistical Analysis

Retrospective analysis

Cox Proportional Hazard Survival Model

Observation starting point / end point:
  : Birth / Re-operation or death

Variables:
  Age
  Types of TAPVD
  Obstructive vs. non-obstructive
  FSV vs. BVR candidate
  PBF: \( \uparrow \) or \( \downarrow \) or \( \leftrightarrow \)
  Types of intervention (SR vs. Others)

Model validation with bootstrap analysis
Withdrawal (n=11)

Decision making of withdrawal

- by parents: 7 / 11
- by co-existing medical problem: 4/11
  - Chromosomal anomaly (n=1)
  - Delayed Dx with MOF (n=1)
  - Complex obstructive TAPVD (n=2)

High withdrawal rate by specific cardiologist

<table>
<thead>
<tr>
<th></th>
<th>Cardiologist A</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tx</td>
<td>11</td>
<td>33</td>
</tr>
<tr>
<td>No Tx</td>
<td>7</td>
<td>4</td>
</tr>
<tr>
<td>Total</td>
<td>18</td>
<td>37</td>
</tr>
</tbody>
</table>

(P=0.000326, OR=12.2)
Annual changes in withdrawal
# Types of TAPVD (n=55)

<table>
<thead>
<tr>
<th>Group</th>
<th>No TAPVD</th>
<th>SupraC</th>
<th>Cardiac</th>
<th>InfraC</th>
<th>Mixed</th>
</tr>
</thead>
<tbody>
<tr>
<td>No Tx</td>
<td>0</td>
<td>5(2)</td>
<td>1(0)</td>
<td>1(1)</td>
<td>4(3)</td>
</tr>
<tr>
<td>TAPVD repair (-)</td>
<td>4(0)</td>
<td>3(1)</td>
<td>3(0)</td>
<td>0</td>
<td>3(0)</td>
</tr>
<tr>
<td>TAPVD repair (+)</td>
<td>0</td>
<td>19(7)</td>
<td>5(4)</td>
<td>5(3)</td>
<td>2(1)</td>
</tr>
</tbody>
</table>

TAPVD, Total anomalous pulmonary venous drainage; Tx, Treatment; ( ), Draining site obstruction
## Operative procedures

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Without TAPVD repair</th>
<th>With TAPVD repair</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systemic-pulmonary shunt</td>
<td>7</td>
<td>7</td>
</tr>
<tr>
<td>PA banding</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>BCPS</td>
<td>1</td>
<td>9</td>
</tr>
<tr>
<td>Fontan operation</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Cardiac Transplantation</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>PA angioplasty</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>Isolated TAPVD repair</td>
<td>0</td>
<td>4</td>
</tr>
<tr>
<td>Norwood operation</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Miscellaneous*</td>
<td>0</td>
<td>3</td>
</tr>
</tbody>
</table>
## Ventricular Morphology and Surgical Strategies

<table>
<thead>
<tr>
<th></th>
<th>No Tx</th>
<th>BVR</th>
<th>OVR</th>
<th>HTx</th>
<th>Undetermined*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Balanced ventricles</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(n=20)</td>
<td>2</td>
<td>2</td>
<td>5</td>
<td>1</td>
<td>10</td>
</tr>
<tr>
<td>Unbalanced ventricles</td>
<td>9</td>
<td>0</td>
<td>16</td>
<td>1</td>
<td>9</td>
</tr>
</tbody>
</table>

(n=35)

Tx, treatment; BVR, Biventricular repair; OVR, One ventricle repair; HTx, Heart Transplantation

* Undetermined: operative death or treatment withdrawal before determination of surgical strategy
55 patients

- Withdrawal of Tx (n=11)
- No TAPVD repair (n=16)
- TAPVD repair (n=28)

10

- Death (n=10)
- Survival without further Tx (n=1)
55 patients

Withdrawal of Tx (n=11)

No TAPVD repair (n=16)

TAPVD repair (n=28)

10

BCPS (n=5)

Death (n=12)

Fontan (n=5)

5

1

10

1

1

4
55 patients

- Withdrawal of Tx (n=11)
- No TAPVD repair (n=16)
- TAPVD repair (n=28)
  - Death (n=13)
    - Heart TPL (n=2)
    - Fontan (n=6)
    - BCPS (n=12)
  - Biventricular repair (n=1)
  - Waiting (n=1)
Survival after birth (Entire cohort, n=55)
Survival by groups

Survival Probability

Age in months

- TAPVD repair (+)
- TAPVD repair (-)
- No Tx
Summary

- **Withdrawal** from treatment has poor results with 95% mortality within a year.
- **Risk factors** for death in TAPVD repair group:
  1. Obstructive TAPVR
  2. Mixed and infracardiac type TAPVR
- Adjusted with these risk factors, **primary sutureless repair for TAPVD** provide survival benefit (HR:0.43)
Conclusions

• Most patients with PVS are Surgical candidates
• Sutureless repair for PR-PVS shows excellent long term outcome
• Surgical Tx of Congenital PVS is still challenging, and Sutureless repair might be an option
• Primary Sutureless repair for TAPVD with small PV needs to be defined
• Registry based approach for PVS might be expedient to come up with a conclusion