No Disclosures
Goals

- Review the embryology and anatomy
- Review Surgical Strategies for repair
- Discuss recent results for surgical repair
• Total anomalous pulmonary venous connection (TAPVC) develops when the primordial pulmonary vein fails to unite with the plexus of veins surrounding the lung buds. This results in return of pulmonary venous blood to the heart via a systemic vein, and subsequently to the right atrium.
Embryology

• Theories regarding the development of the pulmonary veins are still controversial. The common pulmonary veins form through canalization of the mid-pharyngeal esophageal strand [15], a network of endothelial precursors which lies between the splanchnic plexus around the lungs and the sinus venosus.

• At first the primitive pulmonary venous plexus connects to the sinus venosus [16] and drains into the precursor of the right atrium.

• Later, the common pulmonary vein drains into the left atrium. It is not clear whether the common pulmonary vein forms as an outpouching of the left atrium, as proposed by Neill (1956) [17], or whether the pulmonary venous ostium is secondarily transferred to the left atrium by differential growth and sino-atrial septation as proposed by Kutsche and Mierop (1988) [16].

• In either case, failure of the common pulmonary vein to attach to the left atrium results in the persistence of the initial connection to the right atrium or one of its venous tributaries.

• The exact type of anomalous venous connection is probably a secondary event, a fact supported by the variability in anatomic site of connection in both sporadic and familial cases of TAPVR [2-8].
Historical Perspective

- 1798 First reported by Wilson
- 1942 First Series of Autopsies by Brody
- 1951 First Attempt at surgical Repair by Muller UCLA
- 1956 Lewis & Varco first successful attempt
- 1956 Kirkland series
- 1970's Barrt-Boyes Improved technique with Circ Arrest
Definition

• Total Anomalous Pulmonary Venous Connection TAPVC is a congenital heart defect in which the pulmonary veins do not connect normally to the left atrium. Instead they connect to the right atrium, often by way of a systemic vein.

• Relatively rare, it occurs in about 1 in 17,000 live births. (Fulton, 2008).

• ASD or foramen ovale always present.

• Right to left shunt.

• Accounts for only 1-2% of CHD.

• The presence of an atrial septal defect is necessary to allow partially oxygenated blood to reach the left side of the heart.
Classification

- TAPVC is classified according to the site of connection

- Supracardiac includes connections to the left innominate vein, the SVC, or the azygous

- Cardiac includes connections to the coronary sinus or directly to the right atrium

- Infracardiac includes connections below the diaphragm to the IVC, the portal vein, hepatic veins, or ductus venosus

- Mixed type involves connections of two or more of these types; at least one of the main lobar pulmonary veins is draining differently from the others
Types Of TAPVR

• Supracardiac (type 1): The pulmonary veins drain into the right atrium through the superior venacava. (50% of cases)

• Cardiac (type 2): There are two types: The pulmonary veins can directly enter into right atrium. Or the pulmonary veins can drain into the coronary sinus. (30% of cases)

• Infracardiac (type 3): The pulmonary veins drain into the right atrium through the liver (hepatic) veins and the inferior vena cava. (15% of cases).

• Mixed (type 4): The pulmonary veins split up and drain partially to more than one of these options. (5% of cases).
Pathophysiology

• Oxygenated blood that would normally enter the left atrium now enters the right atrium and passes to the right ventricle.

• As a result the pressure on the right side of the heart increases, leading to hypertrophy.

• TAPVC is incompatible with life unless there is an associated defect present that allows for shunting of blood from the highly pressured right side of the heart.

• A patent foramen ovale or an ASD is usually present.
Pathophysiology

• Since none of the pulmonary veins connect normally to the left atrium, the only source of blood to the left atrium is blood that is shunted from the right atrium across the defect to the left side of the heart.

• The highly oxygenated blood from the lungs completely mixes with the poorly oxygenated blood returning from the systemic circulation. This causes an overload of the right atrium and right ventricle.

• The increased blood volume going into the lungs can lead to pulmonary hypertension and pulmonary edema.
Signs, Symptoms And Clinical Manifestations

• Depends on the presence of obstruction to pulmonary venous drainage and size of ASD.

• In supracardiac type without obstruction: mild cyanosis, CHF, S2 wide and splitted.

• Infracardiac type is always obstructive.

• Infants are always symptomatic, Pulmonary hypertension, cyanosis, tachycardia etc.
Diagnosis

- Chest x-ray:
  - normal or mildly cardiomegaly
  - Varying degrees of pulmonary edema

- ECG:
  - With obstruction:
    - RV volume overload
    - Right axis deviation RVH
Diagnosis

• Echocardiography: confirms the diagnosis. Right heart volume overloaded ASD if present. R to L atrial level shunting PulmonaryVein (site of drainage and degree of obstruction)

• Cardiac catheterization : To visualize the abnormal connection of pulmonary veins particularly if an obstruction is present.

• CTA

  • 3D reconstructions

  • 3D Model Printing
Risk Factors

- Poor pre-operative status (acidosis)
- Pulmonary venous obstruction
- High pulmonary vascular resistance
- Young age
- Small left ventricle
- Major associated anomalies
- Infracardiac or mixed type (see below)
Problems With Mixed TAPVR

- No pulmonary venous confluence
- Requires 2 or more anastomoses
- Smaller anastomoses predispose to pulmonary venous obstruction
- Risk of mortality and late pulmonary venous obstruction is increased
- Operative repair probably not curative
Operative Technique

• Cardiopulmonary bypass with hypothermic circulatory arrest is the preferred approach in critically ill infants

• Surgical goals are eliminating all anomalous connections, draining the pulmonary veins into the left atrium, and closing intracardiac shunts

• The PV-LA anastomosis must be large and undistorted
Operative Technique
Supracardiac Type

• Left SVC connection
  • Ligate left-sided vertical vein at junction with innominate vein
  • Open left atrium and incise the posterior wall
  • Find and incise the anterior wall of the confluence
  • Anastomose the pulmonary venous confluence to the left atrium
  • Close PFO or ASD through left atrium or through separate right atrial incision
Operative Technique

Supracardiac Type

- Right SVC connection
  - Expose pulmonary venous confluence and anastomose to left atrium as above
  - Ligate anomalous connections or patch from within SVC
  - A baffle may be used instead to channel flow from the right SVC through an enlarged interatrial connection
Operative Technique

Supracardiac Type

• Azygous connection

• Ligate anomalous connection

• Anastomose confluence to left atrium as above
Operative Technique

Cardiac Type

Coronary sinus connection

- Create common large interatrial connection by incising coronary sinus septum and septum primum
- Close this new defect with a single patch; all pulmonary venous return and coronary sinus return now drains into the left atrium

Right Atrial Connection

- Enlarge interatrial connection
- Create baffle to direct flow from pulmonary venous opening across interatrial connection
OP Infra-Cardiac Type

- Ligate PDA once CPB is established
- Identify and ligate anomalous descending vertical vein at the diaphragm
- Initiate circulatory arrest
- Open left atrium and incise the posterior wall
- Find and incise the anterior wall of the confluence
- Anastomose the pulmonary venous confluence to the left atrium
- Close the interatrial communication
Complications

Early

- Pulmonary Edema
- Pulmonary Hypertensive Crisis
- Phrenic Nerve Damage

Late

- Pulmonary Venous Obstruction
- Anastomotic Stricture
- Pulmonary Venous Stenosis
Total Anomalous Pulmonary Venous Connection: An Analysis of Current Management Strategies in a Single Institution

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Table 1. Mortality According to Anatomic Subtype/Obstruction for Patients with Two Ventricle

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Number</th>
<th>Obstructed</th>
<th>Early Mortality</th>
<th>Late Mortality</th>
<th>Total Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supracardiac</td>
<td>46</td>
<td>20</td>
<td>(4 (%)</td>
<td>1 (2%)</td>
<td>5 (11%)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(4 obstructed)</td>
<td>(1 obstructed)</td>
<td>(5 obstructed)</td>
</tr>
<tr>
<td>Infra-cardiac</td>
<td>26</td>
<td>24</td>
<td>1 (4%)</td>
<td>2 (8%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(obstructed)</td>
<td>(all)</td>
<td></td>
</tr>
<tr>
<td>Cardiac</td>
<td>7</td>
<td>0</td>
<td>0</td>
<td>2 (29%)</td>
<td>2 (29%)</td>
</tr>
<tr>
<td>Mixed</td>
<td>5</td>
<td>2</td>
<td>1 (20%)</td>
<td>1 (20%)</td>
<td>1 (20%)</td>
</tr>
<tr>
<td>Total</td>
<td>84</td>
<td>46 (55%)</td>
<td>0 (5%)</td>
<td>10 (12%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(5 obstructed)</td>
<td>(6 obstructed)</td>
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</tr>
</tbody>
</table>

Table 2. Mortality According to Anatomic Subtype/Obstruction for Patients with Single Ventricle

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Number</th>
<th>Obstructed</th>
<th>Early Mortality</th>
<th>Late Mortality</th>
<th>Total Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supracardiac</td>
<td>24</td>
<td>10</td>
<td>7 (28%)</td>
<td>2 (8%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(4 obstructed)</td>
<td>(1 obstructed)</td>
<td>(5 obstructed)</td>
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<tr>
<td>Infra-cardiac</td>
<td>9</td>
<td>8</td>
<td>5 (56%)</td>
<td>8 (89%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(5 obstructed)</td>
<td>(8 obstructed)</td>
<td></td>
</tr>
<tr>
<td>Cardiac</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Mixed</td>
<td>5</td>
<td>4</td>
<td>2 (40%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(1 obstructed)</td>
<td>(2 obstructed)</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>39</td>
<td>22 (56%)</td>
<td>14 (36%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(10 obstructed)</td>
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</table>

Table 3. Predictors of Death and Restenosis After the Repair of Total Anomalous Pulmonary Venous Connection (TAPVC)

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Variable</th>
<th>Univariable p Value</th>
<th>Cox Multivariable Analysis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Early death</td>
<td>Proop PVO</td>
<td>0.02</td>
<td>3.8 2.2-6.6 &lt;0.01</td>
</tr>
<tr>
<td></td>
<td>Complex operative technique</td>
<td>0.04</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>1V</td>
<td>&lt;0.001</td>
<td>5.4 2.7-12.0 &lt;0.001</td>
</tr>
<tr>
<td></td>
<td>Total pump time</td>
<td>0.01</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>Circulatory arrest time</td>
<td>0.01</td>
<td>NS</td>
</tr>
<tr>
<td>Total death</td>
<td>Proop PVO</td>
<td>0.04</td>
<td>2.6 1.4-5.5 0.02</td>
</tr>
<tr>
<td></td>
<td>Complex operative technique</td>
<td>&lt;0.01</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>1V</td>
<td>&lt;0.001</td>
<td>4.8 2.5-9.2 &lt;0.001</td>
</tr>
<tr>
<td></td>
<td>Heart in situ vs. everted</td>
<td>0.01</td>
<td>2.8 1.3-6.3 &lt;0.01</td>
</tr>
<tr>
<td></td>
<td>Total pump time</td>
<td>0.01</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>Circulatory arrest time</td>
<td>0.04</td>
<td>NS</td>
</tr>
<tr>
<td>PV restenosis</td>
<td></td>
<td>0.04</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>Nonabsorbable suture</td>
<td>&lt;0.001</td>
<td>NS</td>
</tr>
</tbody>
</table>
Results
Sutureless Vs Conventional

![Graph showing comparison between Sutureless and Conventional methods](image)

- Right Ventricular Systolic Pressure (mmHg)
- Time (Months)

- Sutureless
- Conventional

*P* = 0.6
Freedom from Re-operation

![Graph showing freedom from re-operation over time with two lines representing Sutureless and Conventional methods, and Log-Rank = 0.3]
Freedom From Re-operation or Death

![Graph showing freedom from re-operation or death over time with lines for Sutureless and Conventional methods, and Log-Rank statistic of 0.6]
Summary
100% FOR CHILDREN