Aortopulmonary Window and Associated Anomalies

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Disclosures

- Financial disclosures
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Aortopulmonary Window

- The result of failure of fusion of the two opposing conotruncal ridges that are responsible for separating the truncus arteriosus into the aorta and pulmonary artery.
- APW occurs between the ascending aorta and the main pulmonary artery and may be found just above the semilunar valves or between the more distal ascending aorta and main pulmonary artery.
Classification

Mori, K et al British Heart Journal 1978
Classification

- Perhaps more a continuum?
Classification – APW with IAA

- Embryology of aortopulmonary window including interrupted aortic arch

Berry, Tet al American Journal Cardiology 1982
Classification – APW with IAA

Konstantinov et al JTCVS 2006
Antenatal Diagnosis

• First reported by Collinet et al 2002.
  – Simple aortopulmonary window may not be identified by fetal echocardiography because equal pressure in the ascending aorta and pulmonary root.

• Antenatal diagnosis of aortopulmonary window with interrupted aortic arch first reported in 2009 by Hayashi et al
  – Posterior deviation of the outlet septum is absent.
Presentation

- Similar to that of other patients with left-to-right shunts, such as PDA or VSD and depends on the size of the defect.
- Congestive heart failure
  - Tachypnea
  - Diaphoresis
  - Poor feeding
  - Inadequate weight gain are common. With large defects, bidirectional shunting can produce systemic desaturation.
Presentation

• Physical exam
  – Tachypneic infant with accessory respiratory muscle use.
  – Cardiac examination reveals an enlarged heart, and like patients with patent ductus arteriosus, the pulses are bounding.
  – A systolic murmur can be heard along the left sternal border; however, unlike patients with a patent ductus arteriosus, a diastolic component to the murmur is rare.
  – Patients with associated arch abnormalities frequently present with shock coinciding with closing of the ductus arteriosus.
Presentation

- The diagnosis is routinely made with echocardiography.
- The location and size of the communication as well as associated anomalies are carefully identified.
- Cardiac catheterization is occasionally indicated for the patient who presents after early infancy to assess pulmonary vascular resistance.
Presentation

- MRI

Presentation

- Associated cardiac lesions >50% (other than ASD and PDA)
  - Arch anomalies; arch hypoplasia/coarctation → arch interruption ~ one-third
  - VSD
  - RVOT anomalies; TOF, PS, PA/IVS, aortic origin of RPA
  - LVOTO
  - Transposition
  - Coronary anomalies
Indications for intervention

- Presence of an aortopulmonary window is an indication for treatment
  - Very small window could be followed and ultimately closed using catheter based techniques
- Small to moderate intermediate APW may be suitable for device closure
  - Access in a small infant may be difficult
- Repair is best accomplished using CPB
Preoperative management

- Resuscitation - limiting excessive pulmonary blood flow
  - requires intubation and mechanical
  - sedation ± neuromuscular blockade
  - hypercapnea and minimizing $\text{FiO}_2$ will increase the pulmonary vascular resistance, decrease left-to-right shunting, and improve systemic oxygen delivery.
  - Inotropic support may be required.

- Prostaglandin infusion is necessary in patients with Interrupted aortic arch
<table>
<thead>
<tr>
<th>Year</th>
<th>Surgeon</th>
<th>Method</th>
</tr>
</thead>
<tbody>
<tr>
<td>1948</td>
<td>Gross</td>
<td>Ligation</td>
</tr>
<tr>
<td>1953</td>
<td>Scott and Sabiston</td>
<td>Division without cardiopulmonary bypass</td>
</tr>
<tr>
<td>1957</td>
<td>Cooley et al</td>
<td>Division with cardiopulmonary bypass</td>
</tr>
<tr>
<td>1966</td>
<td>Putnam and Gross</td>
<td>Transpulmonary patch closure</td>
</tr>
<tr>
<td>1967-1968</td>
<td>Negre et al</td>
<td>Anterior sandwich patch closure</td>
</tr>
<tr>
<td>1967-1968</td>
<td>Bircks</td>
<td></td>
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<tr>
<td>1967-1968</td>
<td>Johansson et al</td>
<td></td>
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<tr>
<td>1968</td>
<td>Wright et al</td>
<td>Transaortic suture closure</td>
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<tr>
<td>1969</td>
<td>Deverall et al</td>
<td>Transaortic patch closure</td>
</tr>
<tr>
<td>1987</td>
<td>Shatapathy et al</td>
<td>Pulmonary artery flap closure</td>
</tr>
<tr>
<td>1992</td>
<td>Matsuki et al</td>
<td>Anterior pulmonary artery flap closure</td>
</tr>
</tbody>
</table>

From Gaynor in Pediatric Cardiac Surgery
Operative technique – APW

- Median sternotomy
- Inspection
  - Extent of APW
  - Evidence of anomalous coronary arteries
Operative technique – APW

- Preparation for CPB
  - Encircle branch pulmonary arteries
  - Arterial cannulation
  - Single venous cannulation unless associated intracardiac repair is necessary

- Once CPB initiated branch PAs are snared
  - Mild hypothermia
  - Antegrade cardioplegia
Repair of APW can be accomplished using an incision through the aorta, pulmonary artery or the aortopulmonary window.
Repair of APW best accomplished through an incision in the window itself
- Incision initiated in the superior aspect
- Identify the position of coronary origins before continuing
- Single patch of Gore-Tex
Operative technique – APW

- Single suture line
- Anteriorly suture line is used to complete closure of aorta and pulmonary artery
Operative technique – APW with IAA

- Aortopulmonary window with interrupted aortic arch

Diagram:
- PDA
- Type A IAA
- MPA
- RPA origin from ascending aorta
Operative technique – APW with IAA
Operative technique – APW with IAA
Operative technique – APW with IAA
Results of repair

- Outcome dependent on era of surgery and presence of additional anomalies
  - APW with IAA associated with increased time related mortality, hazard ratio = 5.87, p = 0.009

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<tbody>
<tr>
<td>Median age at presentation (range)</td>
<td>n = 12</td>
<td>n = 11</td>
<td>n = 19</td>
<td>0.61</td>
</tr>
<tr>
<td>3.7 months (3 days, 2.3 years)</td>
<td>0.6 month (3 days, 5.4 years)</td>
<td>2.4 months (birth, 6.1 years)</td>
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<tr>
<td>Presence of other cardiovascular anomalies</td>
<td>8 (67%)</td>
<td>10 (91%)</td>
<td>16 (84%)</td>
<td>0.29</td>
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<tr>
<td>Death before repair</td>
<td>2 (17%)</td>
<td>2 (18%)</td>
<td>1 (5%)</td>
<td>0.31</td>
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<tr>
<td>n = 8</td>
<td>n = 9</td>
<td>n = 18</td>
<td></td>
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<tr>
<td>Median age at repair (range)</td>
<td>5.0 months (1.3, 11.1 months)</td>
<td>2.2 months (27 days, 19.3 years)</td>
<td>3.3 months (3 days, 6.1 years)</td>
<td>0.63</td>
</tr>
<tr>
<td>Repair of other anomalies</td>
<td>At repair</td>
<td>2 (25%)</td>
<td>4 (44%)</td>
<td>13 (72%)</td>
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<tr>
<td>Early death after repair</td>
<td>0</td>
<td>1 (11%)</td>
<td>1 (6%)</td>
<td>0.67</td>
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<td>Total deaths</td>
<td>4 (33%)</td>
<td>3 (27%)</td>
<td>2 (11%)</td>
<td>0.13</td>
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Bagtharia et al Cardiol Young 2004
Children’s Hospital of Wisconsin experience

- Between 1983 and 2009 n = 24
- Simple APW (n = 11)
- Complex APW (n = 13)
  - IAA or coarctation of the aorta (n = 8),
  - VSD (n = 1),
  - PA/VSD and anomalous origin of the right coronary artery (n = 1),
  - PA/IVS and PAPVR (n = 1),
  - d-transposition of the great vessels (n = 1)
  - congenital absence of the left pulmonary artery with pulmonary artery hypertension (n = 1).
Children’s Hospital of Wisconsin experience

• Simple APW
  – No early or late mortality

• Complex APW
  – Early mortality = 1 APW, pulmonary atresia and intact ventricular septum and PAPVR.
  – Late mortality = 1 Following lung transplantation in the patient with APW, absent left pulmonary artery and pulmonary hypertension.
  – Morbidity – one patient underwent surgical repair of residual arch obstruction
Conclusion

• In the current era results with APW are excellent, mortality is rare and is associated with concomitant anomalies
• Long-term follow-up is indicated
  – Branch pulmonary artery stenosis
  – Recurrent arch obstruction
Thank you