Tetralogy of Fallot with anomalous left pulmonary artery arising from the ascending aorta: Case Reports

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Case 1

* Patient TM:
  * 3 months old female patient

* An incidental murmur was discovered during her admission for the treatment of bronchopneumonia

* Referred for cardiac assessment
On Examination

* Acyanotic

* Tachypnoeic

* 3/6 ESM at ULSB

* Cardiomegaly

* CCF
Special investigations

CXR
- Dextroposition
- Oligaemic right lung
- Plethoric left lung

ECG
- RVH

ECHOCARDIOGRAPHY
- Features in keeping with TOF
- Anomalous LPA arising from ascending aorta
Angiograms

Right Ventriculogram

Ascending Aortogram
## Haemodynamics

<table>
<thead>
<tr>
<th>Site</th>
<th>Saturations 30% O2</th>
<th>Pressures (s/d/m)</th>
<th>Qp:Qs</th>
<th>PVR Wood unit</th>
</tr>
</thead>
<tbody>
<tr>
<td>MV</td>
<td>70%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RV</td>
<td>76%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>MPA</td>
<td>70%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RPA</td>
<td>70%</td>
<td>25/10/17</td>
<td>1:1</td>
<td>1.1</td>
</tr>
<tr>
<td>LPA</td>
<td>90%</td>
<td>50/20/38</td>
<td>5:1</td>
<td>0.8</td>
</tr>
<tr>
<td>Asc Ao</td>
<td>94%</td>
<td>55/2538</td>
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</tbody>
</table>
Diagnosis:
* TOF with anomalous LPA arising from the ascending aorta.

Management:
* Surgical repair with implantation of the LPA to the MPA.
**Case 2**

Patient AB

* 2 year and 7 months old female.

* Presented at 6/12 old with bronchiolitis and a cardiac murmur.

* Diagnosed with TOF with absent pulmonary valve syndrome and managed conservatively.

* Diagnostic cardiac catheterization done in preparation for surgical repair.
On Examination

- Acyanotic. No clubbing. Sats = 97%.

- Cardiomegaly with features in keeping with increased pulmonary blood flow.

- Normal heart sounds.

- 3/6 ESM ULSB and a 2/4 EDM ULSB.

- No gallop. Not in CCF
Investigations
Angiograms

MPA Angiogram

Right Ventriculogram
Angiograms

Aortogram
# Haemodynamics

<table>
<thead>
<tr>
<th>Site</th>
<th>Saturations 25% O2</th>
<th>Pressures (s/d/m)</th>
<th>Qp:Qs</th>
<th>PVR Wood unit</th>
</tr>
</thead>
<tbody>
<tr>
<td>MV</td>
<td>71%</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>RV</td>
<td>72%</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>MPA</td>
<td>83%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RPA</td>
<td>83%</td>
<td>41/18/30</td>
<td>1.6:1</td>
<td>1.8</td>
</tr>
<tr>
<td>LPA</td>
<td>92%</td>
<td>58/30/45</td>
<td>3.7:1</td>
<td>1.4</td>
</tr>
<tr>
<td>Asc Ao</td>
<td>94%</td>
<td>76/40/58</td>
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</tr>
</tbody>
</table>
**Diagnosis & Management**

**Diagnosis:**

* TOF with absent PV syndrome.
* Incidental finding of anomalous LPA arising from the ascending aorta.

**Management:**

* On diuretic therapy.
* Awaiting surgical repair.
TOF is rarely associated with an anomalous PA.

It's also rarely (3-6%) association with absent pulmonary valve syndrome.

Coexistence of TOF and absent PV syndrome an anomalous PA with is extremely rare.
ANOMALOUS PA FROM AORTA (PAAo):

* Rare congenital cardiac defect.

* Comprise 0.12% of all CHD’s.

* Solitary defect or in association with:
  * TOF
  * ASD
  * AP window
  * DiGeorge Syndrome
Characteristics PAAo:

- Anomalous origin of one of the branch PA’s from the aorta.
- Contralateral PA arises from RVOT.
- 2 separate semilunar valves.

Origin:

- Majority - The anomalous PA originates on the postero-lateral wall of the ascending aorta, near the aortic valve. **Case 1**

- 15% - The origin is distal, near the base of the innominate artery. **Case 2**
The natural history of PAAo:

- Rapid development of PHT progressing to PVOD.
- Subsequent death.
- A resultant 1 year survival of a mere 30%.
AN ANOMALOUS LPA:

- More common than an anomalous RPA in TOF.

- Occurs due to failure of development of the left 6\textsuperscript{th} arch, leading to failure of fusion of LPA to MPA.

- The aortic sac persists, which is the site from where the LPA then arises.
• STRIKING FEATURE ANOMALOUS LPA and TOF,:
  • Differential pulmonary blood flow.

• The right lung is oligaemic because RVOTO.

• The left lung has increased PBF from unrestricted shunting from the aorta at high systemic pressures.

• It (left lung) is also at risk for Eisenmenger Syndrome.
**DIAGNOSIS:**

- Suspected in patients with TOF, when CXR showing differential pulmonary vascularity, owing to the difference in PBF.

- Can be detected on echocardiography, but was missed in our 2\textsuperscript{nd} patient.

- Cardiac catheterization and angiograms are confirmatory, and also estimate PVR.
* MANAGEMENT:
* Early surgical intervention.

* Preferably before 12 months of age is crucial for the prevention of PVOD.
Complication

Repeat Angiogram case 1

❖ After 27 months

❖ LPA smaller with origin stenosis

❖ This procedure further complicated into a CVI as evidenced by a hyperacute infarct on CTbrain.

❖ Demised soon thereafter.
References


END

THANK YOU