Clinical Guidance

Paediatric Critical Care:
Total Anomalous Pulmonary Venous Drainage (TAPVD)

Summary
This is for staff to use to provide guidance regarding the diagnosis, management and perioperative care of TAPVD.

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Glossary:
MAS = meconium aspiration syndrome
PPHN = persistent pulmonary hypertension of the newborn
PEEP = positive end expiratory pressure
CXR = chest x-ray
CVS = cardiovascular system
RAI = right atrial isomerism

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Paediatric Critical Care
Total Anomalous Pulmonary Venous Drainage. (TAPVD also referred to as TAPVR/C)

**Presentation:**
Obstructed TAPVD may present in 1st hrs of life.
- Frequently undiagnosed lesion on antenatal scans
- Baby may be compromised in utero, pass meconium during labour and be mis-diagnosed as MAS/PPHN
- Profound cyanosis, tachypnoea, tachycardia, poor volume pulses, hepatomegaly.
- Murmur not usually present. Gallop rhythm
- Early recognition/ consideration essential
- Unobstructed symptoms less severe.
- Present first weeks/ months of life
- Failure to thrive, tachypnoea, recurrent chest infections, mild cyanosis, hepatomegaly.

**Emergency management:**

Obstruction is surgical emergency: Urgent cardiac referral & transfer to surgical centre
- Intravenous/ umbilical/ intraosseous access
- Intubate & ventilate for hypoxia & cardiac support
  - Sat's often do not improve
- May require inotropes to support blood pressure
- Cautious aliquots of anaesthetic induction agents.
- Peak pressure to achieve good chest movement. PEEP 5-10cm H2O.
- Dinoprostone (Prostaglandin E2): if no improvement, do not escalate>10 nanogram/kg/min
- Standard sedation with Morphine
- Unobstructed baby: Rarely a clinical emergency. May need ventilation, CVS support & diuretics.

**Special investigations** (RV is volume loaded)
- CXR: heart size = small to normal if obstructed as left ventricle empty
- SVC widening in supracardiac TAPVD; “snowman”. Diffuse pulmonary oedema. Intersitial oedema identified by diffuse reticular pattern; “Snowstorm”. Pleural effusion/s may be present.
- ECG: RV enlargement /R axis deviation. Tall, peaked p in L2.
- $\uparrow$PO2 in UVC gas diagnostic of infracardiac TAPVD

**Peri-Operative management**
- **Obstructed:** (Low cardiac output state/shock)
  - Proceed to definitive surgery. (ECMO may be considered on case by case discussion)
- **High risk surgical categories** Wt <2.5kg. Evidence of in-utero pulmonary vein obstruction with restricted ASD (pleural effusions, ascites); Small left atrium, neonatal presentation with end organ dysfunction & profound cyanosis, hypoplastic pulmonary veins
- **Unobstructed:**
  - Cardiac failure management.
  - Accurate delineation of pulmonary veins & abdominal anatomy.
  - Delayed surgical intervention with surveillance for obstruction.
- **Post op:** Routine post cardiac surgical care in PICU.
  - Risk of pulmonary hypertensive crises ~50% so atrial communication should be left. Signs include ↓ETCO2, desaturation, hypotension & $\uparrow$CVP. Causes RV failure. Prevent & treat with high fio2 (50-70%), minimal handling, sedation +/- muscle relaxation. Prime nitric oxide into ventilator circuit before return from theatre. Short term sildenafil may be required during NO wean. Potential for atrial arrhythmia due to surgical atriotomy
  - Increased risk of organ dysfunction in obstructed group. May require organ support- renal (dialysis), liver (glucose & coagulation) and delayed feeding or TPN

**Outcome/ long term prognosis**
- Influenced by early recognition, timely intervention & severity of pulmonary hypertension post op.
- Long term morbidity due to progressive or recurrent pulmonary vein obstruction. Recurrence risk 15-20%
  - Infracardiac & mixed at greatest risk. Incidence at anastomotic site may be reduced by surgical 'sutureless' technique.
  - Vessel hypoplasia difficult to diagnose pre op. Intrinsic lack of venous growth remains challenging with 3 yr survival ~60%

**Classification:**
- **Supracardiac 49%** PV connect to Superior vena cava or innominate
- **Infracardiac 26%** PV connect to hepatic or inferior vena cava
- **Cardiac 16%** PV to coronary sinus to right atrium
- **Mixed 9%**: Combination of above.

Obstruction seen in 75% infra-cardiac & in 50% supra-cardiac lesions.
Obstruction may develop over time
1/3 patients have no other cardiac anatomical defect.
Frequently occurs with asplenia, right atrial isomerism & heterotaxy.
89% RAI associated with TAPVD

Obstructed infracardiac TAPVD
Small heart shadow, interstitial and alveolar oedema (plethoric, "snowstorm") with pleural effusion. R>L

Supracardiac TAPVD
Dominant right sided cardiac shadow. Enlarged mediastinum due to supracardiac draining vein & enlarged SVC. "Snowman". If obstructed, "snowman in snowstorm", Plethoric lungs fields.