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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This clinical guideline has been produced by the South Thames Retrieval Service (STRS) at Evelina London for nurses, doctors and ambulance staff to refer to in the emergency care of critically ill children. This guideline represents the views of STRS and was produced after careful consideration of available evidence in conjunction with clinical expertise and experience. The guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient.	

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

Change History		
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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
  - Sats 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 H_2O.
- 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.

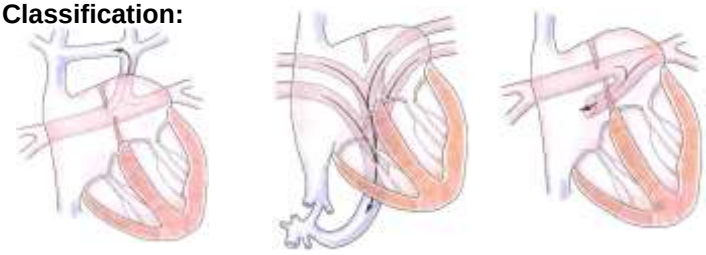
Special investigations (RV is volume loaded)

- **CXR:** heart size = small to normal if obstructed as left ventricle empty
  SVC widening in supracardiac TAPVD; "snowman".
  Diffuse pulmonary oedema. Interstitial oedema identified by diffuse reticular pattern; "Snowstorm".
  Pleural effusion/s may be present.
- **ECG:** RV enlargement /R axis deviation. Tall, peaked p in L2.
- ↑pO_2 in UVC gas diagnostic of infracardiac TAPVD

Description:

Pulmonary veins (PV) do not return to L atrium: blood drains into R side of heart via systemic veins and then back to the L side via ASD/ VSD. If this blood flow becomes obstructed it is surgical emergency.

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.

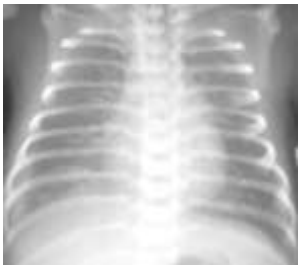
Obstruction seen in 75% infra-cardiac & in 50% supra-cardiac lesions.

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.

Peri-Operative management

- **Obstructed**: (Low cardiac output state/shock)
  - Needs urgent anatomical diagnosis. Confirm by ECHO prior to emergency surgery. Urgent cross match.
  - 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 & ↑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[1]. 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%[2]

References: 1: Rito , Cardiology 2015. 2: Seale l, Circulation 2010 3:Hashmi, J Am Coll Cardiol 1998