Fetal Provider Information Sheet
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Pulmonary Atresia with Intact Ventricular Septum (PA/IVS)

Incidence

- 0.45-0.83 per 10,000 live births, 1-3% of CHD
- PA-IVS can be seen in the recipient twin in twin-twin transfusion

Subtypes:

- Unipartite (8%), bipartite (34%) or tripartite RV (remainder)
- Membranous pulmonary atresia (75%) versus muscular or long segmemt (25%)
- RV ventriculo-coronary connections (VCC)
- RV dependent coronary circulation (RVDCC)

Available Fetal Interventions[1]

Some centers have attempted/offer fetal intervention for some fetuses with PA/IVS

Rationale: In utero balloon dilation of PV may promote RV growth

- Qualifying factors based on imaging
  - Membranous pulmonary valve (identifiable leaflets or membrane)
  - Intact or highly restrictive ventricular septum
  - TV <-2.5 and small RV
- Type – Fetal pulmonary valvuloplasty
  - Goal to decompress hypertensive RV and improve RV inflow and outflow
  - Less experience than with HLHS and criteria for good candidacy evolving
- Outcomes
  - Data on impact on outcomes and impact on natural course is limited and depends on patient characteristics[2,3]

Fetal Imaging Predictors of Postnatal Interventions/Outcomes

Predictors of postnatal Biventricular repair –

Study	Predictors
Lowenthal et al.[4] (n=15, 2V=9, SV=6)	• TV Z score >-4 (sens 90%, spec 83%) • TV/MV ratio ≥0.63 (sens 78%, spec 100%) • >mild TR not seen in any infant that

	did not undergo 2V
Gomez-Montes et al[5] (n=16, 9 PA)	• TV/MV ratio: >0.83 • PA/Ao ratio >0.75 • TR inflow duration >36.5% • RV/LV ratio >0.64 • (3/4 – 100% sens, 92% spec)
Roman et al[6] (n=27, 8 SV, 19 BV)	• TV/MV >0.7 • RV/LV>0.6 • TV inflow >31.5% • No VCC • 3 out of 4 (100 sensitive, 75% specific)
Gardiner 2008[7]	• PV Z score >−1 or TV Z score >−3.4 before 23 weeks • Median TV Z score >−3.95, before 26 weeks • Median PV Z score >−2.8 and medium TV/MV ratio >0.7 at 26–31 weeks • Median TV Z score >−3.9 and medium TV/MV ratio >0.59 after 31 weeks
Predictor of ventricular-coronary connections (n=17)[8]	• Absence of TR and TV/MV<0.56

Rate of growth of TV from 2nd to 3rd trimester is also a predictor (0.012±0.008 cm per week vs 0.028±0.014 cm per week for SV versus 2V outcome)[9]

Prognosis

Therapies: Available therapies and resulting outcomes depends on the favorability of anatomy/subtype of PA/IVS. **This summarizes recently published data but interventions and outcomes may vary significantly by center.**

1. RV dependent coronary circulation:
   - Available therapies: Single ventricle staged surgical palliation with a first stage BT shunt or PDA stent or primary transplant. The latter may be

considered with there is coronary ostial atresia or significant coronary artery stenosis.

2. No RVDCC but unipartite RV and muscular PA
   - SV palliation with BT shunt as first procedure

3. No RVDCC and membranous pulmonary valve atresia
   - Pulmonary RFA balloon valvuloplasty (for eventual 1.5 or 2V repair) – 97% success
     - Pulmonary blood flow can be augmented (required in 50-60%) with a BT shunt or PDA stent
       - Similar complications, mortality, but higher reinterventions in PDA stent group[10]
     - 72% require some type of reintervention on RVOT before 3 years[11]
     - Most achieved 2V circulation, 17% 1.5V or SV (associated with ≤mild TR)[11]

Survival:

- Overall survival 1 month 80%, 1 year 68%, 5 year 60%[12], more recent data 100% 2V, 80% SV)[13]
  - 2V survival rates from 90-91% survival at 20-25 years follow up.
- RVDCC – 40% survival (with most mortality occurring in the first 6 months of life)[14]
  - At risk for sudden death even after Fontan[15]

Neurodevelopment:

- No specific data on ND outcomes in PA/IVS

QOL:
Lower, FHS scores compared to population norms particularly in physicial function, but similar FHS and VO2 in 1V, 1.5V and 2V patients[16] (CHSS n=271)
No significant difference in exercise capacity between two.

<u>Associated Problems</u>

1. Ebstein's anomaly or significant tricuspid valve dysplasia – 10%

2. Uhl's anomaly - extremely rare
3. Associated defects/syndromes – Extracardiac anomalies in up to 13%[17]

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