Fetal Provider Information Sheet
Last Updated: 6/13/19
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Atrioventricular Septal Defect (AVSD)

Incidence [1]

- 2.4-3.1/10,000 live births, 4-5% of all CHD
- 57-92% prenatal detection rate

Fetal Interventions (None)

Fetal Imaging Predictors of Postnatal Management/Outcome

- Key predictor of outcome is balance of atrioventricular valve
    - RV/LV EDD ration between 2 and 4 SDs for gestational age suggests a borderline LV
    - Presence of an apex-forming LV is a predictor of biventricular repair

Prognosis [6, 7,8]

- Main prognostic factor based on associated genetic and extracardiac abnormalities and balance of atrioventricular valve
    - Postnatal TTE: Calculated AVVI < 0.67 + large VSD, recommend single ventricle approach
    - Postnatal TTE: To help determine unbalanced or not: AVVI (atrioventricular valve index) </= 0.4 (right dominant) or >/= 0.6 (left dominant)
- Low operative mortalities for balanced AVSD: 2.2%
- Higher mortality in cases with associated malformations (with the exception of T21)
- Most common reason for reoperation: left AVV regurgitation > subaortic stenosis > residual VSD > late onset CHB

Mortality, Risk and Indication for Reoperation in Balanced AVSD [9, 10, 11]

- Quoted reoperation rates from 10-25%
- Mortality and freedom from reoperation vary based on surgical era, and years of long-term follow-up

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Years Evaluated	Indications for Reoperation	Freedom from reoperation	Survival
1972-2007 [9]	LAVV regurgitation Subaortic stenosis LAVV stenosis, Residual ASD PA stenosis Aortic coarctation	63% at 5 yr 48% at 10 yr 42% at 15 yr	91% at 5 yr 91% at 10 year 86% at 15 year
1974-2000 [10]	LAVV regurgitation LVOTO	88% at 10 yr 83% at 20 yr 78% at 30 yr	85% at 10 yr 82% at 20 yr 71% at 30 yr
1975-2006 [11]	Residual ASD LAVV regurgitation	96% at 1 yr 89% at 5 yr 82% at 15 yr	91% at 1 yr 91% at 5 yr 89% at 15 yr

Associated Problems [1, 3, 4, 5]

- Chromosomal abnormalities, syndromes, extracardiac findings
    - 17-50% of Trisomy 21 have AVSD
    - Abnormal karyotype in 48-58% of those diagnosed with AVSD
- Associated defects:
    - Subaortic stenosis: due to subaortic shelf or fibromuscular tunnel, also evaluate for aortic valve hypoplasia and coarctation of aorta
    - Tetralogy of Fallot: 5% in patients with AVSD, more common in those with Trisomy 21
    - Atrial isomerism: right > left; If right, more likely univentricular heart with common atrium; If left, more likely biventricular heart and CHB common
    - Ventricular hypoplasia
        - Balanced vs. unbalanced will determine biventricular or univentricular repair
- AVV regurgitation – poorer prognosis, contributes to development of hydrops in utero

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