Fetal Provider Information Sheet
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Congenitally Corrected Transposition of the Great Arteries (ccTGA)

Definition/Anatomy: [1,2]

- Atrioventricular and Ventriculoarterial discordance (i.e., "physiologic correction").
- Other names: ventricular inversion, double discordance, corrected transposition, L-TGA
- Connections: RA-LV-PA and LA-RV-AO. The left ventricle is the subpulmonary ventricle. Right ventricle is the systemic ventricle.
- AO usually anterior and leftward to the PA (in atrial situs solitus).
- With atrial situs solitus: {S,L,L}. With situs inversus: {I,D,D} (3-5%).

Incidence: [3,4]

- 0.03/1000 live births. 1% of all congenital heart disease
- Recurrence risk ~5% for future siblings
- Fetal detection rate: ~50% (variable)

Associations: [2,5-7]

- Most common as isolated cardiac abnormalities. Less commonly in context of chromosomal abnormalities, genetic syndromes, extracardiac anomalies (though possible).
- Cardiac abnormalities: (90% have associated cardiac abnormalities)
    - Tricuspid valve abnormalities (up to 90%)
        - Can have Ebstein-like malformation of left-sided tricuspid valve with septal leaflet displaced apically below level of the annulus (~25%).
        - Leaflets can be thick, dysplastic, straddling across the VSD.
        - Tricuspid regurgitation
    - Ventricular septal defect (~75%)
        - Majority are membranous. Often large and extend anteriorly.
    - LVOTO/Pulmonary Stenosis (40-50%)
        - Usually subvalvar as a result of tissue protruding into outflow tract.
        - Pulmonary atresia rarely (~ 10-15%).
    - Conduction system abnormalities
        - Displaced AV node (vs dual AV node) due to septal malalignment; may result in AV block, other arrhythmias.
            - AV block may develop prenatally or postnatally
    - Others cardiac findings: dextrocardia/mesocardia (25-50%), mitral valve abnormalities, right ventricular (systemic) outflow obstruction with CoA.

Prenatal Physiology: [8,9]

- If no associated significant defects, physiology is similar to normal prenatal physiology.
- When VSD present, physiology is similar to prenatal VSD with bidirectional VSD shunting seen in utero, unless there is significant outflow obstruction, which will push VSD shunt to the opposite direction.
- Significant (R or L) outflow tract obstruction may result in postnatal ductal-dependence.
- Significant TR may produce cardiomegaly, pericardial and pleural effusions, arrhythmias, hydrops fetalis, or coarctation (decreased flow through the aorta).
- Key Imaging Predictors of Postnatal Outcomes:
    - Hypoplastic PAs suggests pulmonary obstruction.
    - Reverse flow in ductus arteriosus or aortic arch confers risk for postnatal ductal-dependent pulmonary or systemic circulation, respectively.

Fetal Imaging Checklist: [9]

- Transverse view, Abdomen
    - Situs determination
- Transverse view, 4-chamber + sweep to outflows
    - Cardiac position, segmental anatomy and connections A/AV/V(looping)/VA.
    - Tricuspid valve offset, abnormality, TR
    - VSDs
    - Outflow tract obstruction (valvar, subvalvar)
- 3 vessel view
    - Great artery relationship (e.g. aorta anterior/leftward) and size (e.g., pulmonary artery hypoplasia with LVOTO)
- Short axis
    - Ventricular size, position (e.g., RV posterior), and function, VSD shunting, overriding/straddling of AV valve tissue.
- Others
    - Bicaval view, aortic arch, ductal arch, foramen ovale, ductus venosus
    - Rhythm assessment – assess for AV block, rare SVT
    - Pleural/pericardial effusions, ascites, umbilical artery/vein.

Postnatal Physiology/Management: [10]

- Physiology and management depends on presence and severity of cardiac abnormalities:
    - TR, VSD, pulmonary outflow obstruction, aortic outflow obstruction/CoA, arrhythmia/bradycardia, presence of AV block.
- Difficult to predict degree of postnatal TR prenatally
    - If valve structurally normal with minimal TR, most likely to do well.

- If abnormal with prenatal ≥ mild TR, risk that degree of TR may increase postnatally (systemic AV valve, increased preload (increased pulmonary venous return), increased systemic afterload (removal of placenta).

- Possible management/interventions:
  - PGE1 for ductal dependent LVOTO or RVOTO
  - Aortopulmonary shunt (severe LVOTO)
  - VSD repair
  - Subpulmonary/pulmonary obstruction repair (+/- LV-PA conduit)
  - TV repair
  - CoA repair
  - Need for pacemaker (complete AV block)
- Possibility of "double switch" operation (atrial + arterial switch, vs atrial switch + Rastelli)

Prognosis and Outcomes: [8, 11-16]

- Long-term survival expected into adulthood.
- Outcome depends on severity of associated defects.
- AV block development anytime in life (infancy to adulthood, mean 18 yrs). General risk ~ 2%/year, increased risk post-surgery.
- Systemic right ventricular dysfunction.
  - Higher risk with TR, heart block, long-term ventricular pacing need, increasing age.
- Outcomes after surgical (*physiologic*) repair/
  - Risk for AV block; inevitable systemic RV failure.
  - Risk factor for worse outcome = Ebstein malformation, severe TR, need for TV replacement.
- Outcomes after surgical (*anatomic*) repair/"double switch":
  - Less risk for/improved TR and RV dysfunction, but risk remains for systemic LV dysfunction, especially if requiring pacing or "training" w/ PA band before surgery.
  - Long-term risks which apply to both atrial switch and arterial switch/Rastelli operations (baffle stenosis/leak, atrial arrhythmias, coronary insufficiency, supravalvar AS/PS, subAS, RV-PA conduit failure, need for reoperation).
  - Results promising in select patients, but variations in practice, patient selection, age and type of repair, make interpretation of long-term outcomes challenging.

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