Fetal Provider Information Sheet
Last Updated: 02/2019
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Tetralogy of Fallot (TOF)

Incidence

- 3 to 4 per 10,000 live births.

Subtypes

- Tetralogy with varying degree of pulmonary stenosis
- Tetralogy with Pulmonary atresia (Pulmonary atresia with Ventricular Septal defect)
    - TOF/PA with major aortopulmonary collaterals
- Tetralogy with Complete Common Atrioventricular Canal
- *ToF with Absent Pulmonary Valve 3- 6% of all Tetralogy*
    - >30- 40% with 22q11 deletion; early respiratory failure
    - Fetal MRI may help Characterized by pulmonary regurgitation and stenosis and dilated branch pulmonary arteries and absence of patent ductus arteriosus
    - Poor ventricular function and pulmonary valve: aortic valve ratio>1 worse outcomes [1]
- *ToF with common atrioventricular canal (CAVC)*
    - 6% of ToF; 80-90% trisomies
    - Usually Rastelli Type C

Associations

- Nearly 1/3 have an associated extracardiac anomaly or chromosomal derangement.
- 22q11 has been identified in 16% to 18% of all cases of TOF, and in one-half of those patients with TOF and an associated right aortic arch.
- TOF with a common atrioventricular canal is commonly seen in Trisomy 21.
- Right aortic arch in 25% of patients. The ductus arteriosus may be tortuous or absent. This may be associated with a vascular ring in some cases.
- Persistent left superior vena cava in 11% of cases.
- Mirror image dextrocardia {I,L,I} or isolated infundibular inversion {S,D,I}

- Heterotaxy- interrupted inferior vena cava, polysplenia variety – usually associated with small left sided structures
- Branch pulmonary artery coarctation or isolation
- There are variations of coronary artery anatomy that needs to be confirmed postnatally.

Available Fetal Interventions - NONE

Fetal Imaging Predictors of Postnatal Interventions/Outcomes-

Echocardiographic measurements used for predicting severity of postnatal presentation include poor PA growth, smaller main PA, and PV z scores; ascending aorta or PV/aortic valve ratio; elevation of Doppler velocities in the right ventricular outflow tract; and retrograde flow in ductus arteriosus. *Caution – lack of reliability between prenatal and postnatal PV annulus Z scores* [5]

- If the PA is less than 50% of the size of the aorta on fetal, infants are more likely to have significant PA obstruction after birth, require postnatal PGE to maintain patency of DA, and are more likely to need a neonatal intervention.
- Retrograde flow in DA or absence of DA in the setting of severe RV outflow hypoplasia[6]
- PV Z score <-5 - Ductal dependent; PV Z < -3 with rev ductal flow [3]
- PV <- 3.5 Z, Surgery ~1 mo and 50% re intervention[2-4]
- PV Peak systolic velocity ≥ 87.5 cm/s at 19 to 22 weeks or >144.5cm/sec in third trimester – 93% sensitive and 100% specific for predicting early intervention[8]
- Progression to atresia RARE (Only 6% have reported decrease in PV Z score without progression to PA)[7]
- Aortic arch sidedness and presence of a vascular ring s should be identified as TOF with a right aortic arch has a higher association with 22q11 deletion.
- Absence of thymus is also suggestive of 22q11 deletion.
- ToF- with Absent PV: Left ventricular dysfunction and hydrops and aorta <50% the size of MPA poor prognostic factors

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Limitations of Fetal Echo for ToF

- Predicting Postnatal PV morphology and Z score
- Branch PA coarctation
- Coronary anatomy
- In ToF with Absent PV – unable to predict respiratory status
- ToF- MAPCAs - Distribution of pulmonary blood flow and surgical staging dependent on postnatal assessment. More diminutive or absent central branch PAs indicates more complex pulmonary blood flow

Neurodevelopmental Outcomes

- Variable based on associated syndromes and hypoxia prior to surgery and need for surgery in the first year of life.
- Include mild cognitive impairment; oral-motor dyscoordination, expressive speech and language abnormalities; impaired visual-spatial and visual-motor skills; attention-deficit/hyperactivity disorder (ADHD); motor delays; learning disabilities; and later problems with executive function and diminished health-related quality of life[9]
- As a group, children who require neonatal congenital heart surgery have lower mean IQs than age-matched controls, their IQs still fall within the normal range[10].
- Hypoxemia prior to surgery has not been shown to have a significant impact on neurodevelopmental outcome – but the study was small[11].

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