*Fetal Counseling Provider Information*
*Last Updated:3/1/21*
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## Tricuspid atresia

Definition/Anatomy

- The tricuspid valve is absent
- Anatomic Subtypes:
    - Type 1: Tricuspid atresia with normally related great arteries (70%)[2]
    - Type 2: Tricuspid atresia with transposed great arteries
        - D-TGA (25%)
        - L-TGA (3-6%)
    - Then further subdivided in terms of size of VSD/bulboventricular foramen and restriction to pulmonary or systemic blood flow (atresia, stenosis, no obstruction).
- Associated defects such as ASD, VSD/bulboventricular foramen, or PDA are necessary for survival.

## Incidence

1-2 in 10,000 live births

1.6-2% of CHD (STSS database)[1]

20% of patients have other associated cardiac anomalies (right aortic arch, LSVC, coarctation, aortic arch hypoplasia)

## Associated problems

- Genetics may play a role in some cases:
    - Can be associated with 22q11 deletion, 4q31 deletion, 8p23 deletion, 3p deletion, trisomy 13, trisomy 18, trisomy 21
    - Mutations in ZFPM2 (FOG2), HEY2, NFATC1, NKX2.5, and MYH6 genes reported

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## Fetal Imaging Predictors of Postnatal Management/Outcome

- Management is affected by subtype of tricuspid atresia
- Size of VSD/bulboventricular foramen determines degree of shunting from LV to RV.
    - If ventriculoarterial connections are concordant, the VSD/bulboventricular foramen size affects the amount of flow across the pulmonary valve , determining pulmonary avlave annulus size and potential for ductal dependent pulmonary blood flow postnatally

    - If ventriculoarterial connections are discordant, the VSD/bulboventricular foramen size affects the amount of flow across the aortic valve, determining aortic valve annulus size, aortic arch development, and potential for ductal dependent systemic blood flow postnatally

## Single ventricle palliative surgeries

Timing and type of postnatal palliation are determined by the anatomy

- Restrictive pulmonary blood flow will require BTS vs ductal stenting
- Unrestrictive pulmonary blood flow will require a PA band
- Systemic outflow obstruction or arch hypoplasia will require a modified Norwood or arch repair and PA band.
- In a balanced circulation with concordant AV connections and pulmonary valve stenosis, their first surgery may be a bidirectional Glenn.

## Outcomes[3-5]

8 year Fontan survival 84% (Emory group)[6]

10 year Fontan survival 82% (Toronto group)[7]

All the long-term issues associated with Fontan physiology (PLE, plastic bronchitis) and possible need for heart transplant if left ventricular dysfunction develops

Neurodevelopmental outcomes

Much of the data focuses on HLHS patients, which can likely be correlated somewhat if a neonatal Norwood was needed

However, if the initial surgery is a PA band or no surgery until the Glenn neurodevelopmental outcomes are better[8]

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