Tricuspid Atresia

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Risk

- Uncommon; occurs in 0.056:1000 live births.

Perioperative Risks

- Hypoxia caused by limited pulm blood flow.
- Reliable systemic and pulm blood flow in these pts depends on existence of an unobstructed atrial level right to left shunt, an unobstructed left to right ventricular septal defect, and intact pulm artery.
- There is obligatory mixing of systemic venous blood return to the heart from the vena cavas (lower O_2 sat) and blood return to the heart from the pulm veins (higher O_2 sat).

Worry About

- Inadequate ability of systemic venous and pulm venous blood to mix caused by restrictive atrial septal defect (rare additional problem, but vital).
- Inadequate pulm blood flow caused by restrictive ventricular septal defect, pulm artery stenosis, pulm subvalvular obstruction, or pulm atresia.
- Less common is that the pt that presents with too much pulm blood flow and CHF (completely unobstructed pulm blood flow).

Overview

- Defined by the lack of a connection between the right atrium and hypoplastic (could be practically nonexistent) right ventricle.
- The tricuspid valve may be completely absent, or there may be a rudimentary valve-like structure on the floor of the right atrium that is not patent.
- Basically, there are three major types:
  - Tricuspid atresia with normally related pulm artery and aorta (70–80%). There are three subtypes:
    - Ia Tricuspid atresia with normally related great vessels, pulm atresia, and no ventricular septal defect (pulm blood flow completely dependent on the maintenance of a patent ductus arteriosus in the immediate period after birth)
    - Ib Tricuspid atresia with normally related great vessels, hypoplasia of the pulm artery, and a small ventricular septal defect
    - Ic Tricuspid atresia with normally related great vessels, no hypoplasia of the pulm artery, and a large ventricular septal defect
  - Tricuspid atresia with transposition of the great arteries (pulm artery arising from the left ventricle and the aorta arising from the hypoplastic right ventricle—20–30%). There are three subtypes:
    - IIa tricuspid atresia with transposed great arteries, atresia of the pulm artery arising from the left ventricle, and a ventricular septal defect allowing systemic blood flow to occur through the aorta arising from the hypoplastic right ventricle (pulm blood flow completely dependent on the maintenance of a patent ductus arteriosus in the neonatal period)
    - IIb tricuspid atresia with transposed great arteries, hypoplasia of the pulm artery arising from the left ventricle and a ventricular septal defect
    - IIc tricuspid atresia with transposed great arteries, no hypoplasia of the pulm artery, and a ventricular septal defect
  - Tricuspid atresia with congenitally corrected transposition of the great arteries. The pt can have varying degrees of pulm, subpulmonary, or subaortic stenosis. Also, can be assoc with other lesions like atrioventricular septal defect.

Etiology

- Cause is unknown.
- Although specific genetic causes of the malformation have not been determined in humans, data indicate that the FOG2 gene may be involved in the process.

Usual Treatment

- Two phases of treatment: The first stage is geared to allow the pt to survive the immediate postnatal period. The goals of treatment being to provide adequate but not a plethora of pulm blood flow and sufficient systematic blood flow. Ideally unity would be achieved in blood flow to the pulm and systemic vasculature. Another goal is attain satisfactory mixing of the relatively desaturated blood from the venous return and fully saturated blood from the pulm venous return.
- Limited pulm blood flow makes the pt temporarily dependent on Prostin (PGE_1) to maintain the patency of the ductus arteriosus. This is followed by the creation of a modified Blalock-Taussig-Thomas shunt to create a reliable source of pulm blood flow in the stage prior to the superior caval pulm shunt (Bidirectional Glenn procedure).
- Rarely, if the atrial level communication is limited, then adequate mixing of blood return from the vena cavae and the pulm veins is impeded. An atrial balloon septostomy may be required.
- Rarely, if pulm blood flow is unrestricted, then a surgical banding of the pulm artery may be needed.
- Long-term palliation of these pts involves two more operations intended to separate the blood returning to the heart via the vena cavas (with lower O_2 sat) from the blood returning to the heart via the pulm veins (with higher O_2 sat). This will mimic normal series physiology. A pathway is first created directly from the superior vena cava in the right pulm artery (bidirectional Glenn procedure). Then a pathway is created from the inferior vena cava to the right pulm artery (Fontan procedure).

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV/RESP	More common, limited pulm blood flow causing hypoxia and possibly resp acidosis	Possibly requiring intubation or supplemental O_2	Cyanosis, holosystolic murmur, possible thrill	SpO_2 ABG ECHO
	Rare, over-circulation of pulm vessels causing unacceptably high PO_2	Signs of poor systemic arterial blood flow like metabolic acidosis, necrotizing enterocolitis, poor wt gain	Tachypnea, tachycardia, hepatomegaly	CXR SpO_2 ABG
CNS	Stroke due to single ventricle physiology	Pt may have received a balloon septostomy	Varying levels of consciousness Varying degrees of hemiplegia	CT MRI

Key References: Brown ML, DiNardo JA, Odegard KC: Patients with single ventricle physiology undergoing noncardiac surgery are at high risk for adverse events, *Paediatr Anaesth* 25(8):846–851, 2015; Holtby HM: Anesthetic considerations for neonates undergoing modified Blalock-Taussig shunt and variations, *Paediatr Anaesth* 24(1):114–119, 2014.

Perioperative Implications

Preoperative Preparation

- Maintain PGE_1 preop if needed.
- Have drugs and blood products available to maintain acceptable BP, preload, and contractility to maintain pulm blood flow in ductal or systemic to pulm shunt–dependent pt.

Monitoring

- In addition to routine monitors, implement continuous arterial and central venous pressure monitoring for procedure to create systemic arterial to pulm arterial shunt is recommended.
- If available, monitoring of central venous saturation (SvO_2) is also recommended.

Airway

- Keep the pt intubated postop after systemic arterial to pulm shunt creation. This is usually the period when the Prostin is weaned off, and the transition is made to pulm flow being purely maintained by the newly created shunt.

Preinduction/induction

- Maintain adequate afterload, preload, contractility, heart rate, and heart rhythm.
- Careful titration of benzodiazepines and narcotics help maintain hemodynamic goals.

Maintenance

- Maintain preload and a hemoglobin level of approximately 13.5–15 g/dL. This will maintain adequate tissue O_2 delivery.
- Maintain adequate sedation to prevent rises in PVR.
- This anesthetic involves a careful balance of resistances (PVR and SVR).

Adjuvants

- Heparin, usually 100 units/kg (more may be necessary), to maintain an activated clotting time of at least 200 sec during the creation of the surgical shunt. May require redosing, over the length of the procedure. Remeasure ACT every 20–30 min.
- Inotropes to maintain adequate contractility and SVR.

Anticipated Problems/Concerns

- Thorough understanding of parallel physiology and maintenance of equal pulm arterial and systemic arterial blood flow from a single ventricle is required.