Transposition of the Great Arteries

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Risk

- Incidence: 2.7-3.2:10,000 live births
- Relatively common; represents >5% of all congenital cardiac malformations
- One of the most common causes of newborn cyanosis
- Male predominance of 2.25-fold increased risk
- Relatively low risk (12%) of coexisting syndromes/noncardiac malformations

Perioperative Risks

- Newborn: Hypoxemia, acidemia, low cardiac output syndrome, and death.
- Risk of inadequate mixing of pulm and systemic circulations to allow oxygen uptake and delivery.
- Closure of ductus arteriosus can be fatal.
- Myocardial ischemia secondary to increased myocardial workload, aortic hypotension, and severe hypoxemia.

Worry About

- Adequate mixing of pulm and systemic circulations
- Adequate cardiac output, aortic root diastolic pressure, systemic perfusion, and systemic oxygenation
- Adequate, but not excessive, pulm blood flow
- Rapid changes in PVR in first 72 h of life and impact on adequate flow in pulm and systemic circulation
- Embolic stroke in setting of shunt-dependent physiology

Overview

- Aorta arises from RV, and pulm artery arises from LV, resulting in futile cycle of blood flow in two parallel circulations; saturated blood continuously cycles through lungs, and desaturated blood continuously cycles through body.
- Adequate mixing (shunting) between these two parallel circulations is required to prevent imminent death.
- Intracardiac and extracardiac mixing of parallel circulations may occur at ductus arteriosus, PFO, and/or VSD.
- Three primary subgroups of TGA:
  - TGA with intact ventricular septum (70% incidence) usually present with poor circulatory mixing and profound hypoxemia and acidemia.
  - TGA with ventricular septal defect (25% incidence) usually present with adequate mixing and short-term cardiopulmonary stability but may have severe pulm overcirculation and systemic hypotension.
  - TGA with ventricular septal defect and left ventricular outflow tract obstruction (5–10% incidence) usually present with poor pulm flow and severe hypoxemia and acidemia.

Etiology

- Embryologically, TGA results from abnormal rotation and septation of the truncus arteriosus, resulting in ventriculoarterial discordance.
- No known fetal environmental risk factors for TGA.

Usual Treatment

- Surgical correction is required for survival; medical management is crucial prior to surgery.
- Preop medical management:
  - Ensure adequate mixing as needed: prostaglandin E_1 for ductal patency and/or balloon atrial septostomy for atrial mixing.
  - Supplemental oxygen, ventilation, and/or paralysis as needed in setting of severe hypoxemia and acidemia.
  - Inotropic support as needed for support of systemic blood pressure.
- Surgical options, typically within first wk of life:
  - The arterial switch operation (ASO) is fully corrective and the procedure of choice for most neonates. Aorta and pulm artery are transected and transposed to location above proper ventricles. The ASO has a 97% 30-day survival rate.
  - Surgical banding of the pulm artery may be required for those unable to tolerate early complete repair.
  - The Rastelli, Nikaidoh, or REV procedure may be required for TGA with ventricular septal defect and left ventricular outflow tract obstruction.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
RESP	Hypoxemia/acidemia	Dyspnea, tachypnea, poor feeding	Cyanosis	CXR, ECHO, pulse oximetry, NIRS, ABG
	CHF	Dyspnea, tachypnea, poor feeding	Lung auscultation	CXR, ECHO, pulse oximetry, ABG
	Apnea	Receiving PGE_1		
CV	RV (systemic ventricle) failure, ischemia, hypotension	Nonvigorous, shock	Cool, mottled, cap refill	ECHO, ECG, NIRS, ABG
	Inadequate mixing of systemic and pulm circulations leads to hypoxemia/acidemia	See above	See above	ECHO
CNS	Cerebral stroke/intracranial hemorrhage	Irritability, hypotonia, seizure	Hypotonia, neurologic signs (rare)	US, CT, or MRI of head

Key References: Latham GJ, Joffe DC, Eisses MJ, et al.: Anesthetic considerations and management of transposition of the great arteries, *Semin Cardiothorac Vasc Anesth* 19(3):233–242, 2015; Warnes CA: Transposition of the great arteries, *Circulation* 114(24):2699–2709, 2006.

Perioperative Implications

Preoperative Preparation

- Optimize oxygenation; ventilation and paralysis or RBC transfusion may be required preop.
- Optimize systemic perfusion; inotropic support may be required preop.
- Order blood products and anticipate severe coagulopathy after neonatal hypothermic CPB.
- Review ECHO to understand complex shunt physiology that will dictate intraop management.
- Review electrolytes, complete blood count, coagulation labs, ECG, and CXR.

Monitoring

- Arterial line and central venous access are required if not already present.
- Adequate IV access for transfusion.
- Five-lead ECG, NIRS, core temperature, TEE, and preductal and postductal saturations.

Airway

- Oral or nasal endotracheal intubation.

Preinduction/Induction

- Induction technique should consider impact on SVR, PVR, cardiac output, and shunt physiology.
- IV induction with a combination of an opiate, muscle relaxant, and inhalational agent.
- Inotropic support as needed to maintain cardiac output.
- Short-term hyperoxygenation prior to intubation is acceptable; adjust FIO_2 down thereafter.

Maintenance

- High-dose opiate has advantage of reduced stress response, less coagulopathy, less transfusion requirements, and less postop morbidity in this age.
- SaO_2 of 75–80% is acceptable and may not improve significantly with increased FiO_2.
- Cool pt in preparation for CPB; surround head in ice if using deep hypothermic circulatory arrest.
- Utilize TEE findings to guide therapy in setting of cardiopulmonary instability.
- Manage CPB per institutional protocol.
- Provide inotropic support prior to separation from CPB; observe ECG and TEE for signs of coronary ischemia.
- Prepare for extensive coagulopathy post-CPB and transfuse as indicated.

Extubation

- Postop ventilation is required; extubation is not recommended.

Postoperative Period

- Be vigilant for signs of coronary insufficiency/ischemia due to mechanical kinking of coronary arteries during translocation.
- Continue inotropic support in anticipation of late-onset low cardiac output syndrome from myocardial edema and imperfect protection on CPB.
- Depending on surgical procedure, postop arrhythmias may occur.

Anticipated Problems/Concerns

- After CPB, the LV is now handling the systemic rather than pulm circulation; this increased pressure load can lead to LV failure.
- Postop low cardiac output syndrome.
- Ischemia and arrhythmias.