Truncus Arteriosus

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Risk

- Uncommon congenital heart defect; <3% of all congenital heart defects
- No gender predilection

Perioperative Risks

- CHF.
- Volatile agents may depress myocardial contractility and lower SVR.
- Inadvertent hyperventilation resulting in reduced PVR, excess PBF, and worsening CHF.
- Infective endocarditis.
- Risks of CPB.

Worry About

- Difficult intubation due to assoc with velocardiofacial syndrome (e.g., DiGeorge)
- Air embolus (VSD almost always present)
- Hyperoxia and hyperventilation resulting in pulm overcirculation
- CV collapse at induction due to diastolic runoff and assoc truncal regurgitation with resulting coronary steal and myocardial ischemia.
- Hypocalcemia due to parathyroid hormone dysfunction.

Overview

- There is a single great artery arising from heart, supplying the systemic, pulm, and coronary circulations.
- VSD almost always present; ASD present in two-thirds of pts.
- Abnormal truncal valve; 50% are regurgitant.
- Anomalies of coronary artery and aortic arch may be present.
- Pulm circulation arises directly from systemic circulation.
- Dominant physiology is a L-to-R shunt driven by the relatively lower resistance of the pulm vascular bed.
- Runoff from systemic circuit into the PA during diastole may compromise myocardial perfusion.
- Primary goal is to balance PVR and SVR so that Qp:Qs is close to unity.
- Pulm vascular obstructive disease due to excessive pulm blood flow develops early.
- Repair preferably done in early neonatal period before onset of pulm Htn.
- Uniformly fatal without surgical correction (50% of pts die by 1 mo and 80% within 1 y).
- Approx 30% have a deletion of 22Q11, resulting in phenotypic variants such as DiGeorge and Sphrintzen syndromes.

Etiology

- Conotruncal defect.
- Embryonic truncus arteriosus fails to separate into a pulm and aortic trunk.
- Partial or complete absence of conotruncal septum.
- A single arterial trunk arises from both ventricles.
- Classified into four types.
- Maternal diabetes mellitus predisposes to conotruncal abn.

Usual Treatment

- Medical therapy is temporizing (digoxin, loop diuretics, inotropes) to treat CHF and usually only to optimize status before surgery. Surgery should not be delayed for an extended period of time.
- Surgical repair in the neonatal period is the definitive treatment. On hypothermic CPB, the pulm trunk is separated from the truncal artery. The VSD is closed. A conduit from the RV to the PA (Rastelli) is placed to provide for pulm blood flow. The PFO is left open.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Difficult laryngoscopy and intubation		Small mandible, small mouth	
CV	CHF—truncal valve regurgitation Pulm Htn	Difficulty feeding Sweating during feeds FTT	Cyanosis with or without a single S_2 Murmur—systolic or diastolic	Pulse oximetry, ECG, ECHO, cardiac cath±
RESP	CHF, excessive pulm blood flow	Difficulty breathing	Tachypnea with retractions	CXR (increased pulm markings, cardiomegaly)
ENDO	Parathyroid hypoplasia	Seizures, tetany		Serum ionized Ca^{2+}, parathyroid hormone level
IMMUNE	Cellular immunodeficiency	Recurrent infections Chronic diarrhea	70% of 22q11 pts are immunosuppressed.	CBC, T-cell function
MS	Dysmorphic facies		Hypertelorism, low-set ears	

Key References: Jonas RA: Truncus arteriosus. In *Comprehensive surgical management of congenital heart disease,* ed 2, Boca Raton, FL, 2014, CRC Press, pp 571–584; Walker SG: Anesthesia for left to right shunt lesions. In Andropoulos DB, Stayer S, Mossad EB, et al. editors: *Anesthesia for congenital heart disease,* ed 3, Hoboken, NJ, 2015, Wiley, pp 486–489.

Perioperative Implications

Perioperative Preparation

- Treat CHF.
- If intubated, transport and ventilate pt with the FIO_2 at 21%, aiming for an SpO_2 of 75–80%, and appropriate hypoventilation to maintain the $PaCO_2$ at 45–50 mm Hg and pH at 7.25–7.35.
- Avoid hyperventilation and hyperoxia, which will lower PVR, increase PBF, and possibly compromise systemic and coronary perfusion.
- Check serum electrolytes, Ca^{2+}, and Hct.

Airway

- High index of suspicion for a difficult airway, with appropriate precautions if velocardiofacial syndrome present.
- Maintain FIO_2 at 21% but possibly give a few breaths at 100% just prior to intubation.
- Once pt is intubated, return FIO_2 to 21% and avoid hyperventilation.

Preinduction/Induction

- Meticulous air bubble exclusion
- Preop antibiotics.
- Consider inotropic support (e.g., dopamine at 3–5 µg/kg/min) if MAP is low prior to induction.
- Obtain a baseline ECG prior to induction. Monitor for myocardial ischemia (best detected with ECG leads II and V) due to PA runoff.
- Volume infusion is unlikely to increase diastolic BP unless the pt is significantly volume depleted. Consider a 1–2 µg/kg bolus of phenylephrine for low MAP.
- Pts are usually ventricular volume overloaded, and aggressive volume resuscitation will further elevate ventricular end-diastolic pressure, compromising subendocardial perfusion.
- Balance PVR and SVR so that Qp:Qs approaches unity. Key: Maintain SVR; keep SpO_2 below 90%.
- Surgeon must be in the OR prior to induction and prepared for sternotomy.
- For persistent hypotension and rapid sternotomy, snare the branch PAs to elevate MAP and reduce pulm overcirculation.

Monitoring

- Intra-arterial cath and CVP. May have in situ umbilical arterial and/or venous lines.
- TEE is valuable to assess truncal valve function, VSD patch leak, ventricular function, and PA pressure.
- Intraop LA line placement for postop management of preload.
- NIRS monitoring is standard, especially during low-flow selective cerebral perfusion or DHCA.

Maintenance

- Usually with fentanyl infusion 2–4 µg/kg/h.
- Inotropic drugs commonly used to facilitate weaning from CPB incl dopamine (3–5 µg/kg/min), milrinone (0.5–0.75 µg/kg/min) and low-dose epinephrine (0.03–0.05 µg/kg/min), calcium chloride (20–30 mg/kg/h).
- Inhaled NO should be available in the OR for the post-CPB period, as there is a high risk of pulm vasoreactivity.

Extubation

- Postop ventilation is usually required for at least 24 h, as pulm hypertensive crisis may occur. Not suitable for fast-tracking.

Postoperative Period

- Poor RV function (right ventriculotomy and Rastelli conduit placement): maintain appropriate inotropic support, afterload reduction, and adequate preload for RV. Mechanical ventilation with minimal mean airway pressure.
- LV dysfunction (circulatory arrest, long bypass time, myocardial ischemia, truncal valve abn).
- Increased PVR and pulm Htn (low CO and low SaO_2) responds to hyperventilation, metabolic alkalosis, vasodilators (milrinone, PGE_1, NO), sedation (analgesia, paralysis).
- If PA pressures are high, residual VSD and RV outflow tract obstruction must be excluded in addition to treating pulm Htn.
- R-to-L shunting across PFO facilitates systemic cardiac output at the expense of SaO_2 in the face of RV dysfunction and elevated PA pressures.
- AV block requiring temporary pacemaker.
- Bleeding.
- Cardiac tamponade.

Anticipated Problems/Concerns

- CHF
- Truncal valve regurgitation and/or stenosis
- Pulm Htn
- Infective endocarditis