Heart Disease, Congenital

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Risk

- CHD is the most common birth defect.
- Incidence: 1:25 live births.
- 85–90% of pts with CHD survive to adulthood in USA due to advances in medical care.

Perioperative Risks

- The highest risk factors of this complex disease include HLHS; poorly compensated physiology; presence of long-term complications (arrhythmia, pulm Htn, CHF); and emergency surgery.
- Intermediate risk factors include major surgery, age less than 2 y, preop hospital stay >10 d, ASA physical status IV or V.
- Cardiac failure.
- Pulm Htn defined as PAP >25 mm Hg at rest and >30 mm Hg during exercise.
- Arrhythmias.
- Cyanosis.
- Mortality: there is a twofold increase in mortality in children with congenital cardiac lesions compared to those without CHD who present for noncardiac surgery.
- POCA registry.
- Majority of cardiac arrests occurred in general OR (54%) in children undergoing noncardiac surgery.
- Out of all the children with heart disease that arrested, 75% of them were <2 y, often with unrepaired lesions.

Worry About

- Resource availability: Is this child's cardiac history too complex for this institution/periop team?
  - Send the following children to a specialist center: Cyanosis, neonate with CHD, Eisenmenger syndrome, pulm Htn, aortic stenosis, HLHS, single ventricle physiology (BT shunt/Sano, Glenn, Fontan).
  - If true emergency and cannot be transferred, then understanding anatomy, physiology, and shunting is key to management.
  - Use PICU for postop management, especially complex lesions.
- Maintain forward flow/cardiac output.
- Balance pulmonary and systemic blood flow.
- Maintain adequate tissue oxygen delivery.
- Prevent arrhythmia.
- Optimize fluid balance.

Overview

- How to group these children: There are multiple ways, but the most useful is by physiology.
- Normal "series" circulation: Most repaired pts; there can be a small amount of mixing.
  - ASD/VSD
    - L-to-R shunting: This increases pulmonary blood flow and potentially decreases systemic blood flow.
    - R-to-L shunting: Deoxygenated blood flows into systemic circulation and causes reduced pulmonary blood flow and increased cyanosis.
    - Changes in SVR and PVR during anesthesia have the greatest effect in pts with large, unrestrictive defects.
- Parallel "balanced" circulation:
  - Pts with large AV septal defect or VSD, BT or Sano shunt, Truncus arteriosus.
  - Mixing of systemic venous and pulmonary venous blood; potential for cyanosis.
  - Balance between SVR and PVR.
- Single-ventricle circulation:
  - Blood flows passively to the lungs down a pressure gradient from the pulmonary artery to left atrium in pts who have a Glenn shunt or Fontan circulation.
  - Changes in intrathoracic pressure or in PVR affect pulmonary blood flow, which then affects systemic blood flow.
  - BT or Sano shunts are usually the first stage of creating Fontan circulation (palliative, to supply blood flow to the lungs).
    - Graft is connected between the subclavian artery (BT) or right ventricle (Sano) and the pulmonary artery.
    - Complete mixing of systemic venous and pulmonary venous blood (normal SpO_2 75–85%).
    - Flow is determined by SVR and PVR ratio: These pts are sensitive to changes in PVR or SVR, which can be caused by increased FIO_2, changes in $PaCO_2$, and volatile anesthetics or other vasodilators.
  - Glenn shunt is second stage of Fontan repair:
    - Bidirectional superior cavopulmonary shunt.
    - Connects SVC to the right pulmonary artery.
    - IVC drains to right atrium.
    - Pulmonary venous and systemic venous blood mix, yielding SpO_2 75–85%; pt will have cyanosis after procedure.
    - Can tolerate FIO_2 100% usually without issues.
  - Fontan circulation:
    - Inferior vena cava connected to the right pulmonary artery.
    - Separates the pulmonary and systemic circulation.
    - Passive flow to pulmonary circulation.
    - Normalizes oxygenation (children are sensitive to increases in PVR; decreases blood return to the heart, leading to a reduction in cardiac output).
    - Pressure gradient from the pulmonary artery to LA is the force driving pulmonary blood flow.

Etiology

- Genetics/syndromes:
  - Chromosomal abnormalities: Down syndrome (up to 30% can have heart defect), trisomy 18 and 13, Turner syndrome, Cri-du-chat syndrome, Wolf-Hirschhorn syndrome, DiGeorge syndrome
  - Associations: VACTERL
  - Syndromes: William syndrome, Goldenhar syndrome, Marfan syndrome, Noonan syndrome, Smith-Lemli-Opitz syndrome
- Family history of congenital cardiac disease
- Maternal factors/medications:
  - Untreated maternal PKU has a sixfold increased risk.
  - Preexisting maternal diabetes has a fivefold increased risk.
  - Medications: Lithium, thalidomide, isotretinoin, and bactrim.
  - Rubella.
  - Febrile infection, especially in the first trimester.

Usual Management

- Management is dependent on specific anatomy and physiology.
- Attempt to maintain same SpO_2 and other vital signs, as when the pt is at their baseline.
- L-to-R shunts:
  - Avoid increases in SVR, which will increase shunt.
  - Avoid decreases in PVR, which will increase shunt.
  - Avoid negative inotropes.
  - Avoid hypervolemia, which can lead to congestion.
  - In the event of desaturation, consider whether cause could be reversal of shunt.
- R-to-L shunts:
  - Maintain high SVR, to decrease shunt: Ketamine, phenylephrine.
  - Avoid increases in PVR.
  - Minimize intrathoracic pressure.
  - Avoid air bubbles in IV.
- Single ventricle physiology:
  - Common outpatient medications: Diuretics, antiarrhythmics, anticoagulants, antihypertensives
  - Corrective/palliative procedures (see overview for more detailed anatomy/pathophysiology of the following):
    - BT or Sano shunt: Systemic and pulmonary blood flow is determined by SVR and PVR ratio; avoid changes to PVR, as this leads to changes in systemic flow; use caution with volatile agents and vasodilators.
    - Glenn shunt: This can typically tolerate FIO_2 100%.
    - Fontan circulation: Avoid increases in PVR; maintain normal SpO_2.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
RESP	Pulm edema Cyanosis Pulm Htn Decreased lung compliance	Baseline SpO_2 when healthy Dyspnea TET spells Previous surgeries: rib resection, RLN injury	Crackles Blue/gray skin Clubbing in extremities	CXR SpO_2 ABG
CV	New murmur Heart failure Arrhythmia ASD/VSD Shunting Mixing	Dyspnea Lethargy Syncope Low functional capacity Poor feeding Sweating while eating	Tachycardia Tachypnea Hypotension Cool extremities Hepatomegaly Peripheral edema S_3 gallop Diaphoresis Elevated JVP	ECG ECHO (TTE) CXR Cardiac cath BP in upper and lower extremities
GI	Poor weight gain Protein losing enteropathy in post-Fontan patients (particularly in adolescence)	Poor feeding Poor weight gain Nausea Ascites	Hepatomegaly	LFTs Synthetic function of liver (coagulation panel)
CNS	CVA Syncope	Syncope Fatigue Headache Seizures	Neurologic deficits Blurred vision	ECHO (TTE) CBC (hyperviscosity)
HEME	Polycythemia	Low SpO_2 CVA	Neurologic deficits Blurred vision	CBC, peripheral smear Coagulation studies
METAB	Lyte abnormalities	Medication history Diuretics	Peripheral edema	Lytes Ca^{2+}, Mg^{2+}, phosphate

Key References: Thomas J: Anaesthesia for the child with congenital heart disease: pointers and pitfalls, *CME* 29(11):463–466, 2011; Cannesson M, Earing M, Collange V, Kersten JR: Anesthesia for noncardiac surgery in adults with congenital heart disease, *Anesthesiology* 111(2):432–440, 2009.

Perioperative Implications

Preoperative Preparation

- Hx can include poor feeding and failure to gain weight.
- Physical examination:
  - New onset murmur: Examine heart first; ignore the murmur, define the nature of the first and second heart sounds; most systolic murmurs are benign; all pansystolic and diastolic murmurs are pathologic and should be worked up prior to elective procedures; all murmurs that radiate are pathologic (PDAs, aortic valve); if child has a murmur, have BP taken in upper and lower extremities (coarctation).
  - Pathologic murmurs: Obtain TTE prior to proceeding.
  - Pt color:
    - Pink: Normal or L-to-R shunt; pathology—ASD, VSD, PDA
    - Blue: R-to-L shunt or mixing lesions; pathology—TOF, TGA, single ventricle
    - Gray: Sick children, usually critically ill; decreased CO; pathology—severe coarctation, interrupted aortic arch
  - Examine for cyanosis/clubbing.
  - Labs/studies: Hematocrit—could have polycythemia -> compensating for hypoxemia; ECG; recent TTE or cardiac catheterization; CXR.

Monitoring

- Dependent on the surgical procedure.
- Arterial line.
- Consider CVP.
- TEE if indicated.
- If PDA is present, have pulse ox on right upper extremity (preductal oxygenation) and any other extremity (postductal oxygenation).

Airway

- Consideration for syndromes that include CHD that could lead to difficult airway: Down syndrome, Pierre Robin (10% have cardiac anomalies), Beckwith-Wiedemann, Goldenhar syndrome, mucopolysaccharidoses

Preinduction/Induction

- Consider IV preinduction.
- Premedication is acceptable (and often indicated): Consider monitoring premedicated pts.
- Propofol: Reduces SVR and MAP; no change in heart rate, PVR, and PAP; left-to-right shunting decreases; right-to-left shunting increases; propofol may cause reduction in oxygenation and PBF by increasing shunting.
- Ketamine: Well tolerated in children with CHD; increases MAP; minimal effect on SVR, PVR, and PAP.
- Inhalational induction: Induction may take longer than in children without CHD; volatile agents decrease SVR and myocardial contractility; use caution with higher levels of inhalational agent, as these children may not tolerate high concentrations as well as children without CHD.

Maintenance

- Inhalational agents usually can be used with minimal effects on myocardial contractility and shunting. Caution with sicker/younger children who have lesions sensitive to PVR: SVR. Studies are limited on use of desflurane in this population.
- Can use opioids to minimize activation of SNS.
- Remember endocarditis prophylaxis if appropriate.

Extubation

- Period with the highest oxygen demand

Postoperative Period

- Acute care or ICU level of care, unless child has fully repaired ASD, PDA, VSD, or other two-ventricle repairs without significant sequelae.
- Good pain control.
- Maintenance of normothermia.
- Avoid hypovolemia.
- Maintain normal acid/base balance.
- Continue to minimize physiologic changes that could affect shunt fraction.

Anticipated Problems/Concerns

- If at all possible, consider transferring pt to center equipped to handle complexity of anatomy and physiology; if unable to transfer, contact pediatric cardiac anesthesia specialists for advice if pt presents in an emergency and transfer is not an option.
- Who to transfer:
  - Children with complex lesions, such as single ventricles, Fontan circulation, BT shunts, bidirectional cavopulmonary shunt, and cyanosis, should be transferred if possible.
  - Repaired ASD, VSDs can typically be cared for safely in most institutions.
  - For adults presenting with CHD with complete anatomic repair, manage with conventional strategies.
  - For adults presenting with CHD with complex anatomy and physiology, transfer to a center that has subspecialty consultants and cardiac anesthesiologists experienced in caring for complex physiology. These pts should transfer when able: Fontan circulation, repaired TOF with pulmonic valve stenosis or right ventricular failure, cyanotic lesions.