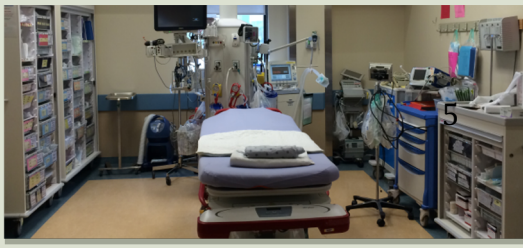



Section VII: Supporting Documents, Laboratory Results, & Multimedia

| Laboratory Results | | | | | | |
|---|---|---|---|---|---|---|
| Na: 148 mmol/L | K: 6 mmol/L | Cl: 115 mmol/L | $HCO_3$: 26 mmol/L | BUN: 25 | Cr: 0.3 | Glu: 4.0 mmol/L |
| Ca: 2.5 mmol/L | Mg: 0.8 mmol/L | $PO_4$: 2.5 mmol/L | Albumin: 3.5 g/dL | | | |
| | | | | | | |
| VBG | pH: 7.1035 | $PCO_2$: 28 mmHg | $PO_2$: 40 mmHg | $HCO_3$: 13 | Lactate: 4 mmol/L | |
| | | | | | | |
| WBC: 10 | Hg: 20 g/dL | Hct: 60% | Plt: 300 | | | |
| ***Labs indicate dehydration, metabolic acidosis.*** | | | | | | |

| Images (ECGs, CXRs, etc.) |
|---|
| None |
| Ultrasound Video Files (if applicable) |
| None |

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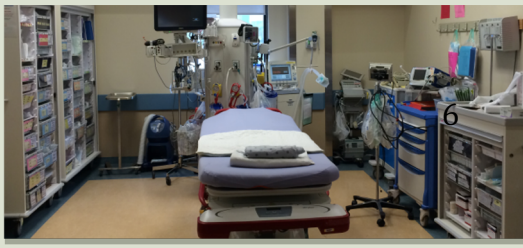
# Pediatric Cardiac Emergency



Section VIII: Debriefing Guide

| General Debriefing Plan | | | |
|---|---|---|---|
| ☐ Individual | ☒ Group | ☐ With Video | ☒ Without Video |

| Objectives | |
|---|---|
| Educational Goal: | To review the management of cardiovascular shock in neonates and develop a differential diagnosis for it |
| CRM Objectives: | 1) Effectively lead a newborn resuscitation and seek input from others<br>2) Use effective communication strategies, including closed loop communication and summarizing events for team |
| Medical Objectives: | 1) Recognize cardiovascular shock in the newborn<br>2) Develop differential diagnosis for cyanosis and cardiovascular shock in the pediatric population<br>3) Develop differential diagnosis for non-cardiac causes of shock in neonates } sepsis, infection, metabolic disturbances, inborn errors of metabolism, non-accidental trauma<br>4) Work-up for cyanosis and cardiovascular shock<br>5) Management of pediatric patient in cardiovascular shock<br>6) Developing understanding of coarctation of the aorta and how a patent ductus arteriosus maintains peripheral circulation in utero and in the setting of cardiovascular shock<br>7) Becoming aware of "pulmonary stealing" phenomenon in neonate and how high oxygen exposure can actually worsen shock In children with ductal-dependent heart disease |

| Sample Questions for Debriefing |
|---|
| 1) Did you feel that your team leader had control over the room? Did he or she ask for input?<br>2) Do you feel you communicated well as a team?<br>3) What were your priorities in managing this patient? How did they change as the case progressed?<br>4) What was the cause of this patient's abnormal vitals signs and level of consciousness?<br>5) How do you get vascular access in a newborn?<br>6) How much fluid do you give a child with suspected cardiac disease?<br>7) What were some delays in arranging transport for this patient and how could this be expedited? What are your nearby neonatal cardiac centres and how does your centre arrange transport to them? |

| Key Moments |
|---|
| Recognition of coarctation of the aorta in neonate and the immediate need to give prostaglandins to maintain PDA |

**BACKGROUND INFORMATION**

- 6 common pediatric cardiac presentations in emergency medicine:
  - Cyanosis
  - Shock
  - Congestive heart failure
  - Pathologic murmur
  - Hypotension
  - Syncope
  - Poor feeding

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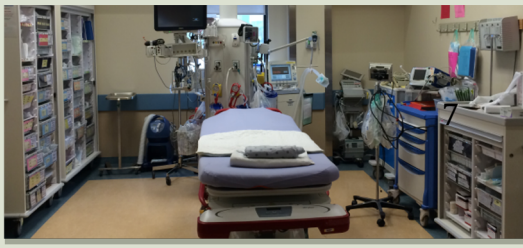

**TABLE 72-1** Clinical Presentations of Congenital Heart Disease

| Clinical Presentation | Causative Conditions in Neonates | Causative Conditions in Infants and Children |
|---|---|---|
| Cyanosis | Transposition of the great arteries, TOF, tricuspid atresia, truncus arteriosus, total anomalous pulmonary venous return | TOF, Eisenmenger's complex |
| Cardiovascular shock | Critical AS, coarctation of the aorta, HLHS | Coarctation of the aorta (infants) |
| Congestive heart failure | Rare: PDA, HLHS | PDA, VSD, ASD, atrioventricular canal |
| Murmur | PDA, valvular defects (AS, PS) | VSD, ASD, PDA, outflow obstructions, valvular defects (AS, PS) |
| Syncope | – | AS, PS, Eisenmenger's complex |
| Hypertension | – | Coarctation of the aorta |
| Dysrhythmias | – | ASD, Ebstein's anomaly, postsurgical complication after repair of congenital heart defect |

*Abbreviations:* AS = aortic stenosis; ASD = atrial septal defect; HLHS = hypoplastic left heart syndrome; PDA = patent ductus arteriosus; PS = pulmonic stenosis; TOF = tetralogy of Fallot; VSD = ventricular septal defect.

- **Cyanosis and shock** usually seen in the *first few weeks* of life and infants are VERY ill
- **CHF** can be more insidious and presents with respiratory distress, feeding intolerance
- **Asymptomatic murmurs** can be evaluated on a non-emergent basis as an outpatient
- **Innocent murmurs** tend to be low intensity, are brief, and occur during SYSTOLE
- In contrast, ***pathologic murmurs*** *tend to be harsh, holosystolic, continuous/ diastolic, and radiate*

**CYANOSIS & CARDIOVASCULAR SHOCK**

- Cardiac causes of cyanosis and shock are usually within the **first 2 weeks** of life and the neonate is critically ill
- **The Ddx** for cyanosis and shock is broad→ congenital heart disease, sepsis, infection, metabolic disturbances, non-accidental trauma
- If there is cyanosis, use the **hyperoxia test.** In the hyperoxia test, the infant is given 100% oxygen. If there is no improvement in $PaO_2$ (ABG) or if unavailable, pulse oximetry, then cyanosis and shock is cardiac in nature. If pulse oximetry or $PaO_2$ improves, then it is respiratory in nature
- **Presentation:**
  - **Acral cyanosis**: blue in the distal extremities. This can be normal
  - **Central cyanosis**: blue in the mucus membranes. This is very suggestive of cyanotic congenital heart disease
  - Cyanosis + murmur is very suggestive of **congenital heart disease**. But if the murmur is not present, there can still be a structural heart lesion
  - Shock +/- cyanosis within the first 2 weeks of life = possible **ductal-dependent congenital heart disease** where the systemic (shock) or pulmonary (cyanosis) blood flow is dependent on the circulation thorugh the patent ductus arteriosus
  - **Shock in neonate**= pallor/ grey, mottling, cyanosis, and changes in mental status (apathy, irritability, lethargy, tachypnea, tachycardia

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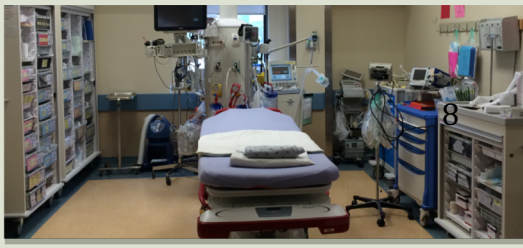



        - Tachypnea and tachycardia suggest impending cardiovascular collapse
    - Discrepancies between the pre-ductal (right brachial) and post-ductal (femoral) blood pressures and pulses are classic for **ductal-dependent lesions** (e.g. coarctation of the aorta)
- **Work up:**
    - Distal pulses → check for quality, amplitude and duration of the pulses
    - Vitals
    - Pre-ductal and post-ductal pulse oximetry (right arm and right/ left leg)
    - 4 extremity blood pressures
    - Chest X-ray
        - heart size
        - shape
        - pulmonary blood flow→ increased pulmonary vascularity can be seen in left-to-right shunting and left-side CHF. In right-sided outflow issues (e.g. pulmonic stenosis), there is decreased pulmonary vascularity
        - position of the aortic arch → abnormal right position of the aortic arch suggests congenital cardiac lesion
    - ECG with pediatric analysis
        - **Cyanotic heart lesions** will often have right axis deviation and right ventricular hypertrophy
        - **Left outflow lesions** (i.e. coarctation of the aorta) can show left ventricular hypertrophy
    - Echocardiogram would be used for the definitive diagnosis
- Differential diagnosis for cyanosis/ shock from congenital heart disease:
    - **Cyanotic lesions**→ transposition of the great vessels, tetralogy of Fallot, other forms of right ventricular outflow tract obstruction, abnormalities of right heart formation
    - **Acyanotic lesions**→ severe coarctation of the aorta, critical aortic stenosis, hypoplastic left ventricle
- **Transposition of the great vessels:**
    - Most common cause of *cyanosis in the FIRST week of life*
    - Often missed because there is NO cardiomegaly or murmur unless there is also a VSD
    - Prior to the stage of shock, it presents with central cyanosis, tachypnea, and feeding difficulty
    - Loud and single S2
    - CXR looks like "**egg on a string"** shaped heart with narrow mediastinum and increased pulmonary vasculature
    - ECG shows *right-axis deviation* and *right ventricular hypertrophy*
- **Tetralogy of Fallot**:
    - Most common *cyanotic congenital heart disease*
    - Cyanosis later on in infancy/ childhood
    - Physical exam→ holosystolic murmur of VSD, diamond-shaped murmur of pulmonary stenosis and cyanosis
    - Cyanotic spells leading to the **child squatting**
    - CXR reveals **boot-shaped heart** with decreased pulmonary vascular markings or right-sided aortic arch
    - ECG shows right ventricular hypertrophy and right axis deviation
    - **"tet spells**": hyper-cyanotic episodes in children with Tetralogy of Fallot. The patients present with paroxysmal dyspnea, labored respirations, increase cyanosis, and syncope. These are often triggered by feeding, crying, or straining while toileting. They can last from minutes to hours.
- **Left ventricular outflow obstruction syndromes**
    - When it is the cause of shock, it can present with or without cyanosis
    - Several congenital lesions belong to this group. ALL of them are dependent on having blood flow thorugh a **patent ductus arteriosus (PDA)**. If the ductus arteriosus closes, then the infant will have decreased/ absent perfusion, pallor/ ashen colour, hypotension, tachypnea, and severe lactic acidosis
    - **Coarctation of the aorta** present with decreased lower extremity pulses & BP (relative to the right brachial pulse & BP)

**Management of Cyanosis & Cardiovascular Shock**

1. Cyanosis and respiratory distress are first managed with high flow oxygen, cardiac and oxygen monitoring, and a stable IV/ IO line
    - Neonates have a fetal hemoglobin, which has a higher affinity for binding oxygen. Thus, they can handle lower oxygen saturations. Oxygen is a potent pulmonary VASODILATOR and can lead to "**pulmonary stealing**" of systemic blood flow. Thus

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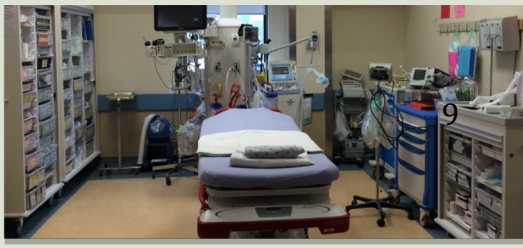

children with ductal-dependent heart disease (i.e. coarctation of the aorta) can experience systemic shock with high oxygen exposure. In these cases, give p**rostaglandins** to maintain the PDA

2. In infants who are suspected to have shunt-dependent lesions and are in severe shock, give **prostaglandin E1 (PGE1)** to try to reopen the ductus. Start with **PGE1 0.05 ug/kg/min** and taper down to the lowest effective dose. Can go up to 0.2 ug/kg/min. Side effects include: fever, skin flushing, diarrhea, and periodic apnea
3. These are *very* sick kids. Consults a **pediatric cardiologist** ASAP. For the children in shock, consult a **pediatric intensivist.**
4. Children having hyper-cyanotic spells are first managed with putting them into the **knee-to-chest position** and giving them **morphine sulfate 0.2 mg/kg SC/ IM/ IO**. If refractory and (persistent hypotension and tachycardia), consult pediatric cardiologist for using **phenylephrine** (for hypotension) and **propranolol (**for tachycardia)
5. Also think about the **NON-cardiac cause of shock** → fluid challenge with NS 10-20 mL/kg and empiric antibiotics if indicated. Fluids should be given more cautiously in neonates with congenital heart disease (i.e. use 10 mL/kg boluses)
6. For hypotension, start with **epinephrine 0.05-0.5 ug/kg/min infusion** and titrate to desired blood pressure.

**CONGESTIVE HEART FAILURE**

- Can result from congenital or acquired heart disease
- Usually occurs AFTER the neonatal period
  - Congenital causes → 2nd – 3rd month of life
  - Acquired causes→ Later in childhood
- **Clinical presentation of CHF** = poor feeding, diaphoresis, irritability, lethargy with feeding, weak cry, and in severe cases, grunting, nasal flaring, and respiratory distress
  - *Early tachypnea* of CHF in infants is usually "effortless" and is the first sign of decompensation. It then progresses to increased work of breathing and rales
- **DDx**:
  - With the exception of constrictive pericarditis, **cardiomegaly** will be seen in the other causes of CHF
  - **Cardiomegaly =** on CXR, the AP view will show a cardiothoracic index >0.6; on lateral views, there will be a lack of retrosternal air space due to the direct abutment of the heart against the sternum
  - **Early onset CHF (neonatal period)** is associated with ductal-dependent lesions like coarctation of the aorta, persistent patent ductus arteriosus (PDA), sustained tachyarrhythmias
  - **2nd-3rd month of life, CHF** causes include ASD and VSD that leads to pulmonary over-circulation
  - **After 3 months of life,** the causes of CHF are usually acquired cardiomyopathy or myocarditis.
  - Other causes include pneumonia, endocarditis or another complication that exacerbates a congenital heart condition
  - **Cardiomyopathy presentation=** rales; feeding difficulty; S3 +/- S4; organomegaly and pulmonary vascular congestion on CXR
  - **Myocarditis presentation:**
    - usually after a viral respiratory illness and more common in school-aged children
    - shortness of breath; vomiting; poor feeing; lethargy; fever; poor perfusion; organomegaly; tachypnea; tachycardia, gallop rhythm if in CHF
    - **ECG** → diffuse ST changes, dysrhythmias or ectopic beats on ECG are associated with increased risk of death. A prolonged QT interval is associate with poor outcomes
    - **cardiac troponin T** is highly sensitive but not specific and if <0.01 ng/mL, myocarditis can be ruled out
    - **CXR**→ cloudy lung field from inflammation or pulmonary edema
    - **Cardiomegaly with poor distal pulses** and **prolonged cap refill** rule out pneumonia (can be hard to differentiate pneumonia from myocarditis)
  - **Pericarditis** presents with:
    - pleuritic pain, positional chest pain, muffle heart sounds and friction rub
    - **CXR** → cardiomegaly
    - **Echocardiogram** is done on an urgent basis to differentiate it pericardial effusion vs dilated cardiomyopathy vs hypertrophic cardiomyopathy. Also determines the need for pericardiocentesis
    - **Pure right-sided CHF** causes are usually pulmonary (i.e. cor pulmonale). In the early stages, the first sign can be **periorbital edema.** This then progresses towards hepatomegaly, jugular venous distention, peripheral edema, and anasarca

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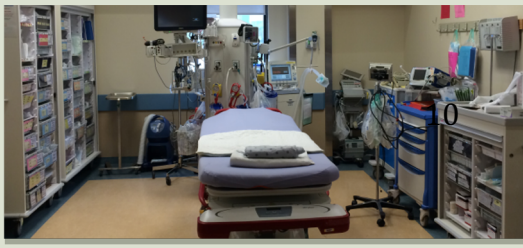

**TABLE 72-2** Differential Diagnosis of Congestive Heart Failure Based on Age at Presentation

| Cardiac Lesion | Chest Radiograph | Electrocardiogram |
|---|---|---|
| Tetralogy of Fallot | Boot-shaped heart, normal-sized heart, decreased pulmonary vascular markings | Right axis deviation, right ventricular hypertrophy |
| Transposition of the great arteries | Egg-shaped heart, narrow mediastinum, increased pulmonary vascular marking | Right axis deviation, right ventricular hypertrophy |
| Total anomalous pulmonary venous return | Snowman sign, significant cardiomegaly, increased pulmonary vascular markings | Right axis deviation, right ventricular hypertrophy, right atrial enlargement |
| Tricuspid atresia | Heart of normal to slightly increased size, decreased pulmonary vascular markings | Superior QRS axis with right atrial hypertrophy, left atrial hypertrophy, left ventricular hypertrophy |
| Truncus arteriosus | Cardiomegaly, increased pulmonary vascular markings | Biventricular hypertrophy |

**Management**

1. it is crucial to avoid metabolic stressors like hypothermia or hyperthermia in children that possibly have CHF. These patients will present with mild tachypnea, hepatomegaly, and cardiomegaly. Sit them comfortably upright, give oxygen, and keep them in a neutral thermal environment.
2. If work of breathing increases or CX shows CHF signs (e.g. pulmonary edema, cardiomegaly) → **furosemide 1-2 mg/kg IV**
3. For hypoxemia, give oxygen, fluid restrict and diurese. CPAP or BIPAP can also be used.
4. **Inotropic agents** can be used to stabilize and improve left ventricular function. For MILD CHF, the initial d**igoxin** dose is **20-30 µg/kg for neonates** an **30-50 µg/kg** for **ages 1 month-2 years**
5. If CHF progresses to cardiogenic shock (absent distal pulses, decreased end-organ perfusion), switch from digoxin to **dopamine OR dobutamine**
   - Initial starting dose for **dopamine** is 5-15 ug/kg/min
   - Initial starting dose for **dobutamine** is 2.5-15 ug/kg/min. however, dobutamine can cause significant tachycardia
   - **Milrinone** is given at 50 ug/kg IV over 10-60 minutes followed with continuous infusion of 0.25-0.75 ug/kg/min. Milrinone is a useful inotrope that improves diastolic relaxation and vasodilation without increasing myocardial oxygen demand.
6. **Secondary derangements** such as respiratory insufficiency, acute renal failure, lactic acidosis, disseminated intravascular coagulation, hypoglycemia, hypocalcemia, and fever should be aggressively managed
7. **Congenital defects** that cause the CHF need to be diagnosed and treated accordingly. This often involves cardiac catheterization and then surgical repair. Prostaglandin E1 may need to be given before surgery.

**COARCTATION OF THE AORTA (CoA)**

- **Coarctation of the Aorta:** narrowed aortic segment that consists of localized medial thickening with some infolding of the medial and superimposed neointimal tissue. The coarctation can be discrete (more common) or a long segment of the aorta is narrowed
- **Coarctation** is usually at the **proximal thoracic aorta** just after the left subclavian artery and across from the opening of the ductus arteriosus. Coarctation of the abdominal aorta is rare
- CoA can be the only heart defect but can also presents with **other congenital anomalies** (e.g. bicuspid aortic valve, ventricular septal defect, aortic stenosis, patent ductus arteriosus, mitral valve disorders, intracerebral aneurysms)
- **In utero,** most of the cardiac output bypasses the coarctation and enters the patent ductus arteriosus (PDA)
- **CoA** leads to 2 phenomena:

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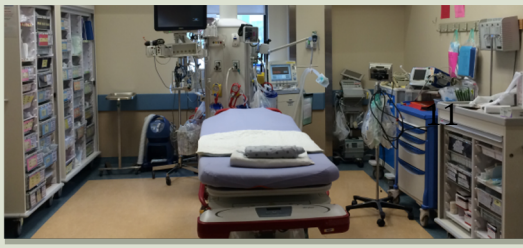

- Pressure overload of the aorta and left ventricle proximal to the coarctation→ left ventricular hypertrophy and hypertension o the upper body including the brain
- Hypoperfusion distal to the coarctation→ poor circulation to the abdominal aorta and lower extremities. The poor abdominal organ circulation can also lead to increased risk of sepsis from the intestinal flora

- **Overtime, the complications of CoA** include:
  - Left ventricular hypertrophy
  - Heart failure
  - Collateral vessel formation
  - Bacterial endocarditis
  - Intracranial hemorrhage
  - Hypertensive encephalopathy
  - Hypertensive cardiovascular disease in adulthood
  - Later in life/ during pregnancy, there is an increased risk of aortic dissection or rupture aorta

**Epidemiology**

- M: F 2:1

**Presentation**

- **Significant CoA:**
  - Within the first 7-10 days of life, there will be circulatory shock with renal insufficiency (oliguria, anuria) and metabolic acidosis } can mimic sepsis
  - Acutely ill once the ductus arteriosus closes or constricts
- **less severe CoA:**
  - can be asymptomatic in infancy
  - CHF rare after neonatal period
  - Children can have subtle symptoms like headaches, chest pains, fatigue, or leg claudication while exercising
  - Upper extremity hypertension
  - Intracerebral aneurysms rupturing→ subarachnoid or intracerebral hemorrhage
- **Physical examination:**
  - Strong pulses and hypertension in upper extremities
  - Diminished/ delayed femoral pulses
  - **BP gradient** between upper and lower extremities. There is low/ unmeasurable arterial BP in lower extremities
  - **Grade 2-3/6 systolic ejection murmur** in the upper left sternal border, left axilla, and occasionally in the left interscapular area
  - If the child also has a bicuspid aortic valve, there can be an apical systolic ejection click
  - Dilated intercostal collateral arteries can cause a continuous murmur over the intercostal spaces
- Presents with upper extremity hypertension, left ventricular hypertrophy and malperfusion of the abdominal organs and lower extremities

**Management**

- **Symptomatic neonates** need to be treated immediately. **An infusion of prostaglandin E1** starting at 0.05-0.1 mcg/kg/min should be given to reopen the constricted ductus arteriosus. By opening the ductus arteriosus and its aortic ampulla, some of the pulmonary blood flow can bypass the coarctation and enter the PDA to the descending aorta. Form here, it can improve systemic perfusion and relieve metabolic acidosis
- **Be careful about supplemental oxygen due to *pulmonary stealing*!** Oxygen can cause pulmonary vasodilation and increased pulmonary blood flow, which impairs systemic blood flow
- For **symptoms of heart failure**, diuretics can be useful. IV milrinone can also be used for those with HF and LV dysfunction
- in **non-emergent situations:**
  - **patients with mild coarctation and NO signs of lower body hypoperfusion**→ monitor until definitive repair is done.
  - **hypertension**→ beta-blockers. *Avoid ACEi* because of potential renal damage
  - **hypertension** (that persists or lasts for a few years after repair) **AFTER repairing the coarctation**→ beta-blockers, ACEi, ARBs or CCBs

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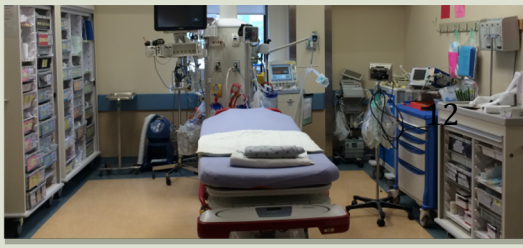



- **surgical management** depends on the centre. Options include:
  - balloon angioplasty with or without stents
  - surgical correction and later on, balloon procedure to re-coarctation
  - balloon procedure for primary treatment in older children and adults
  - resection and end-to-end anastomosis
  - patch aortoplasty
  - left subclavian lap aortoplasty
- **endocarditis prophylaxis** is needed for 6 months after surgery
- **periprocedural complications** include restenosis and aneurysm after **balloon angioplasty.** These are more common than with surgical intervention. However, these are still uncommon
- **complications of surgery** include paraplegia from cross-clamping the aorta during surgery. This is very rare

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