

Intensive Care Nursery House Staff Manual

Congenital Heart Disease

INTRODUCTION: Congenital heart disease (CHD) affects ~1% of newborn infants and accounts for ~10% of all congenital anomalies. Factors that ↑ risk for CHD include maternal diabetes mellitus, familial presence of genetic syndromes (*e.g.*, Noonan syndrome), history of previous infant with CHD, and genetic factors that are just now starting to be identified. The most common types of CHD are ventricular septal defect, pulmonic stenosis, endocardial cushion defect, atrial septal defect and tetralogy of Fallot. Although some infants with CHD do not have signs or symptoms in the newborn period, others will need immediate intervention because of the severity of their disease. The following discussion of CHD is not exhaustive. It is intended as a guide to the initial management (diagnostic and therapeutic) of infants presenting with clinical findings indicative or suggestive of CHD.

SOME GENERAL GUIDELINES:

- Not all newborns with murmurs have CHD.
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- Not all newborns with CHD present in the newborn period.
- Not all newborns with clinical findings of CHD need immediate intervention.
- It is often difficult to differentiate CHD from pulmonary and other diseases (especially sepsis) in a newborn infant.
- Findings that should alert one to the possibility of serious CHD in a newborn include:
 - Cyanosis
 - Respiratory distress
 - ↓ pulses
 - Single 2nd heart sound
 - Prominent heart murmur
 - Hyperactive precordium
 - Metabolic acidosis (unexplained)
 - Poor peripheral perfusion
 - Difference in pulses (arm vs. leg)
 - Abnormally loud 2nd heart sound
 - Abnormally prominent pulses
 - Shock
- Obtain Cardiology Consult as soon as the diagnosis of CHD is suspected.**

EVALUATION OF AN INFANT WITH SUSPECTED CHD:

1. History:

- A. Family:** hereditary diseases, previous sibling with CHD
- B. Pregnancy and perinatal:** Viral infections, maternal medicines, maternal illness
- C. Postnatal:** Poor feeding, cyanosis, tachypnea, tachycardia, edema

2. Physical examination:

- A. Auscultation:** Do this first, before infant starts crying.
 - Heart rate, rhythm
 - Heart sounds (especially 2nd sound): intensity, split (presence, character)
 - Murmur: quality (systolic, diastolic, continuous), intensity, timing, location
 - Other sounds: gallop, click
- B. Cardiovascular system (remainder):** cyanosis, precordial activity, thrill, pulses, blood pressure (both arms vs. leg), peripheral perfusion (capillary refill time)

C. Other: respiratory distress (improving or worsening), hepatosplenomegaly, dysmorphic features, urine output

3. **Laboratory:**

- A. Chest radiograph:** heart size, pulmonary vascularity, pulmonary edema, lung parenchyma
- B. Electrocardiogram:** rate, rhythm, axes of P and QRS, voltages in precordial leads
- C. Monitoring:** O₂ saturation by pulse oximetry, both pre-ductal (right hand) and post-ductal (foot)
- D. Arterial pH and blood gas tensions:** Evaluate mainly for hypoxemia and metabolic acidosis. In contrast to older children and adults, CO₂ may be elevated in newborn infants with congestive heart failure.

COMMON MODES OF PRESENTATION of CHD include:

1. **Respiratory distress:**

- A. Likely lesions include:**
 - VSD, PDA, ASD: These rarely cause symptoms in first few days of life in term infants unless more than one lesion is present.
 - AV canal, aortico-pulmonary window, total anomalous pulmonary venous return (TAPVR) without obstruction
 - TAPVR with obstruction: Respiratory signs occur immediately after birth, may be very severe and worsen progressively. This lesion requires emergency intervention.
 - Truncus arteriosus
- B. Management:**
 - O₂ is rarely helpful and may worsen shunting by decreasing pulmonary vascular resistance.
 - Correct anemia (if present), limit sodium and fluid intake, administer diuretics, optimize nutrition.
 - Early surgery may be necessary for some.

2. **Cyanosis with normal or ↑ pulmonary blood flow:**

- A. Likely lesions include:**
 - Transposition of great arteries (TGA): the commonest form of CHD causing cyanosis in the newborn period. (post-ductal saturation > pre-ductal)
 - Truncus arteriosus (may present with respiratory distress)
 - Double outlet right ventricle
 - Ebstein's anomaly of the tricuspid valve (prominent RV impulse, cardiomegaly)
- B. Management:**
 - TGA without VSD: Cyanosis is prominent. In order to ↑ mixing between pulmonary and systemic circulations and ↑ systemic oxygenation:
 - Start prostaglandin E₁ (PGE₁) at 0.05 mcg/kg/min IV to dilate the ductus arteriosus (see section on PGE₁ at end of this chapter).
 - Balloon atrial septostomy (Rashkind procedure) to allow atrial mixing
 - Surgical repair is arterial switch procedure. If this is not feasible (*e.g.*, abnormal coronary arteries), an atrial baffle procedure can correct the physiological abnormality, but complications are common.
 - Congestive heart failure (CHF) is managed with diuretics and/or digoxin.

3. Cyanosis with ↓ pulmonary blood flow: The most common causes are **right-sided obstructive lesions** that result in inadequate blood flow to the lungs. Age of presentation varies depending on the lesion. Whereas tricuspid atresia presents rapidly immediately after birth, mild forms of Ebstein's anomaly may not present until later in life. With some lesions, cyanosis decreases as pulmonary vascular resistance drops.

A. Likely lesions include:

- Tetralogy of Fallot (with or without pulmonic atresia). In mild cases, cyanosis may not be present at birth (acyanotic Fallot, "pink tet").
- Tricuspid atresia, pulmonic atresia with intact ventricular septum, severe pulmonic stenosis
- Ebstein's anomaly of the tricuspid valve

B. Management: In most cases, infusion of PGE₁ increases pulmonary blood flow and stabilizes the infant until surgical intervention is undertaken. Assisted ventilation and increased environmental oxygen are often necessary. For these infants, oxygen saturations in the 75-85% range are adequate.

4. Shock: The differential diagnosis of shock is discussed in the section on Shock (P. 101). The most common types of CHD that cause shock are **left-sided obstructive lesions**. The mechanism of shock is inadequate systemic blood flow. The onset of signs relates to closure of the ductus arteriosus which may not occur until a few days after birth.

A. Likely lesions include:

- TAPVR with obstruction may present with both respiratory distress and shock.
- Mitral atresia or severe mitral stenosis (type of hypoplastic left heart complex)
- Aortic atresia or severe aortic stenosis (type of hypoplastic left heart complex)
- Interrupted aortic arch (often associated with DiGeorge Syndrome)
- Coarctation of aorta (usually presents several days after birth)

B. Management:

- PGE₁ to allow right→left shunting through ductus arteriosus
- Assisted ventilation to treat pulmonary edema (as well as apnea from PGE₁)
- Correct metabolic acidosis (see sections on Resuscitation, P. 1, and Acid Base Balance, P. 62)
- Inotropic agents to improve myocardial function
- Be cautious with use of oxygen in setting of a single ventricle. Discuss use of oxygen with Fellow or Attending. **Oxygen will accelerate the closure of the ductus arteriosus and worsen infant's condition by decreasing systemic output. Therefore, do not increase environmental oxygen until after PGE₁ has been started.**
- Fluid restriction if there is renal failure
- In some cases of hypoplastic left heart, there is excessive pulmonary blood flow and inadequate systemic output, even with PGE₁. In such cases, the following measures may be used in an attempt to decrease pulmonary blood flow and increase systemic output. These infants require mechanical ventilation and it is usually necessary to administer muscle relaxants (*i.e.*, pancuronium bromide) to prevent the infant from hyperventilating:
 - Decrease inspired oxygen to ~18% by adding N₂ to inspired gas (to ↓ alveolar PO₂)

- Add CO₂ to inspired gas (to ↓ arterial pH)
- ↑ PEEP to try to impede pulmonary blood flow

PROSTAGLANDIN E₁ (PGE₁) is used to dilate the ductus arteriosus in infants with inadequate mixing between systemic and pulmonary circulations (*i.e.*, inadequate pulmonary blood flow or inadequate systemic output).

A. Starting dose of PGE₁ is 0.05 mcg/kg/min IV. If no improvement, ↑ dose to 0.1 mcg/kg/min. Discuss with Cardiology before increasing dose further. After the infant's condition has stabilized, the usual maintenance dose of PGE₁ is 0.025 mcg/kg/min.

B. Complications of PGE₁:

- **Apnea** is the most common complication and is due to a direct effect of PGE₁ on the CNS. The vast majority of infants who require PGE₁ will also need assisted ventilation, either for the severity of their CHD or because of apnea. Therefore, **do not start PGE₁ unless:**
 - (a) Infant is intubated and receiving assisted ventilation *or*
 - (b) You are prepared to intubate the infant and start assisted ventilation immediately after PGE₁ has been started.
- **Hypotension**, due to vasodilatation of peripheral circulation and decreased cardiac output, occurs most commonly at higher doses. Be sure to have arterial access with continuous measurement of arterial blood pressure when PGE₁ is started.
- **Fever**, which makes evaluation for infection difficult. Some cardiologists will administer antibiotics to an infant on PGE₁.
- **Irritability, abnormal EEG and seizures** due to a direct CNS effect and more common at higher doses.
- **Diarrhea.** Although this may occur, infants who require PGE₁ for several days may be tried on enteral feedings, if their condition otherwise permits.
- Longer term effects include **hypertrophy of gastric mucosa** mimicking congenital pyloric stenosis and **periosteitis**.