



Prematurity & Congenital Heart Disease

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Epidemiology

- CHD is the most commonly reported birth defect (6–10 per 1,000 live births) in newborns and is associated with more than a 2-fold increased risk of preterm birth (PTB), defined as birth before 37 weeks' completed gestation.
- Population-based studies attribute this risk of PTB largely to spontaneous preterm delivery, rather than medically induced labor.
- Preterm prelabor rupture of membranes mediates more than 50% of these cases, with additional cases related to extracardiac and genetic anomalies.
- Maternal factors may also play a role in the relationship between CHD and PTB, as complications such as maternal diabetes carry increased risk for CHD and polyhydramnios, which may lead to PTB.

Timing of Delivery

- In the past, it was common for fetuses with critical CHD to have a scheduled delivery at 37 to 38 weeks' gestation, with the intention of facilitating postnatal care at a congenital heart center.
- However, recent literature has consistently demonstrated that delivery of neonates with critical CHD in the “early term” period between 37 and 38 weeks' gestation is associated with greater in-hospital morbidity and mortality.

Timing of Delivery

- Therefore, unless obstetric or fetal concerns, such as preterm premature rupture of membranes, oligohydramnios, preeclampsia, hydrops, placental abnormalities, or nonreassuring fetal status exist, then elective delivery for infants with CHD ideally should be targeted for 39 to 40 weeks' gestation.
- Antenatal care coordination should include relocation of expectant mothers (at approximately 37 weeks' gestation) near a facility with specialized pediatric cardiac care to avoid unexpected spontaneous delivery at a more remote hospital.

Timing of Delivery

- Placental/uterine conditions, fetal growth restriction, multiple gestations, maternal hypertensive disorders including preeclampsia, poorly controlled pregestational or gestational diabetes, and premature rupture of membranes are examples of medical indications for delivery before 39 weeks' gestation.
- Decisions about timing of delivery should be individualized to the needs of the expectant mother and her fetus.
- Multidisciplinary planning with close communication among obstetric, neonatology, and cardiology teams is essential.

Timing of Delivery

- Earlier planned delivery at 38 to 39 weeks' gestation may be considered in circumstances that warrant delivery at a specific cardiac center and extensive coordination of multiple interventionalists and specialists.
- Certain critical CHDs require immediate postnatal intervention, these lesions include Ductal dependent physiology, uncontrolled tachyarrhythmias with hydrops, Ebstein anomaly with hydrops, obstructed total anomalous pulmonary venous return, and tetralogy of Fallot with absent pulmonary valve with severe airway compression.

Postnatal Cardiac Evaluation

- The relative immaturity of the myocardial structure and function in term neonates relative to older infants and children is well known.
- These issues are amplified in premature neonates.
- In addition, in premature neonates with the potential for left-to-right shunting, the underdeveloped pulmonary arterioles may allow for a more rapid decrease in pulmonary vascular resistance and increase in pulmonary overcirculation postnatally in comparison to term neonates with similar cardiac lesions.

Postnatal Cardiac Evaluation

- Postnatal cardiac imaging evaluation of infants with suspected CHD typically begins with echocardiography, whereas other imaging modalities, such as computed tomography angiography (CTA), cardiac magnetic resonance imaging (MRI), and cardiac catheterization, can be considered for specific lesions.
- Risks and benefits of each mode should be considered, especially in premature infants.

Preoperative Cardiac Management

- The first few breaths after birth are typically accompanied by a dramatic hemodynamic shift involving changes in systemic and pulmonary preload and afterload.
- The physiologic stress of this process is magnified in preterm infants with immature organ systems and is further complicated by the presence of CHD.
- The myocardium of preterm neonates has fewer contractile elements, higher water content, and increased reliance on L-type calcium channels, which depend on extracellular calcium instead of calcium stores in the sarcoplasmic reticulum.
- Premature myocardium relies on dextrose for energy (rather than fat).
- For these reasons, preterm neonates with CHD may be more susceptible to circulatory compromise in the transitional phase.

Myocardial Contractility

- Myocyte disorganization, fewer myofilaments, and ↑ noncontractile proteins
- Less contractile reserve and compliance

Energy Utilization

- Premature myocardium relies on dextrose for energy (rather than fat)
- Low hepatic glycogen stores and impaired gluconeogenesis leads to cardiac dysfunction

Calcium Utilization

- Functionally immature calcium regulatory proteins
- Cytosolic calcium concentration depends on transsarcolemmal influx

Sympathetic Nervous System and Catecholamine Response

- ↑ vagal tone and ↓ cortisol and ACTH levels
- Reduced cardiomyocyte β adrenergic receptors leads to blunted response to exogenous catecholamines

Persistent PDA

- Left to right shunting with systemic steal, diastolic hypotension, pulmonary overperfusion and heart failure

Hypotension

- Poor peripheral vasoregulation with myocardial dysfunction and volume

Myocardial Contractility

- CO reliant on heart rate
- Greater sensitivity to afterload, poor tolerance of ventricular pressure and volume load

Energy Utilization

- Avoid hypoglycemia

Calcium Utilization

- Ensure adequate ionized calcium levels for optimal cardiac contractility

Persistent PDA

- Evaluate and treat persistent PDA based on institutional specific guidelines

Hypotension

- Correct systemic hypotension using volume expansion, inotropic agents, and corticosteroids when appropriate
- Goal MAP above a value equivalent to the gestational age (or >30 mm Hg) has been utilized. CHD may warrant different blood pressure thresholds.

Preoperative Cardiac Management

- Some cardiac lesions are supported by fetal physiology but after birth, neonates with CHD are susceptible to significant hemodynamic compromise.
- PGE1 is a crucial medication used to maintain ductal patency in neonates with ductal-dependent CHD who require augmentation of either systemic or pulmonary blood flow, or enhanced mixing of oxygenated and deoxygenated blood.
- Its use is of particular significance in preterm neonates in whom the duration of PGE1 may be longer than in term infants, as cardiac surgery often is performed later.

Preoperative Cardiac Management

- Common complications of prematurity may have a further impact on infants with CHD.
- Timing (or even the utility) of surgical intervention for an infant's CHD may be altered based on the presence of intraventricular hemorrhage (IVH), necrotizing enterocolitis (NEC), or evolving respiratory distress syndrome (RDS).
- Increased risk of infection, anemia, and hyperbilirubinemia have a further impact on the typical management of an infant with CHD with regard to threshold for initiating antibiotics, transfusing blood products, or initiating phototherapy.

Surgical Consideration

- Experienced cardiac surgeons consistently have the impression that cardiovascular tissues are very delicate and relatively unfavorable for surgical manipulation in premature neonates relative to those of full-term neonates.
- Most studies of preterm, LBW, or SGA newborns with CHD have demonstrated higher morbidity and mortality after congenital heart surgery.
- Delaying surgery, however (to achieve weight gain), has not improved outcomes and in most cases exposes the neonate to a significant hemodynamic burden.

Postoperative Considerations

- These complex patients require the resources of multiple disciplines, and an organized handoff process can prevent errors.
- Specifically, neonatologists, cardiologists, (cardiac) intensivists, cardiothoracic surgeons, anesthesiologists, bedside nurses, and the frontline providers must all communicate the important preoperative clinical status (eg, duration of preoperative intubation and preoperative lung compliance) and intraoperative details (such as arrhythmias or residual lesions).
- Interdisciplinary consultation is appropriate in both the preoperative and postoperative settings.

Outcomes

- Operative mortality rates in preterm infants undergoing open cardiac surgery for CHD are higher in comparison to operative mortality rates of infants born at term.
- LBW (<2.5Kg), VLBW(<1.5Kg), and extremely LBW (<1Kg) infants with CHD carry a 1.5 to 4 times greater risk of mortality than those of comparable birthweight whose medical burden is prematurity alone.
- Current data also demonstrate significantly higher rates of postoperative complications and increased morbidity in preterm and early term infants with CHD including sepsis, NEC, seizure, neurodevelopmental disorders IVH, periventricular leukomalacia, retinopathy of prematurity, and BPD.
- Some proposed mechanisms of increased morbidity and mortality in this population include changes in the respiratory system that occur late in gestation, as well as immature energy stores, enzyme function, and immune systems.

Key Points

- Premature neonates with CHD require the collaborative effort of highly specialized teams (neonatologists, pediatric cardiologists, intensivists, cardiac surgeons, anesthesiologists, and bedside nurses) to manage the perioperative course.
- Preoperative complications of prematurity are amplified in the setting of hemodynamic disturbances that are common with CHD.
- Postoperative complications may arise in virtually every organ system and require the close attention of clinicians.

