Management of the Preterm Infant with Congenital Heart Disease

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KEYWORDS
- Prematurity
- Low birth weight
- Congenital heart disease
- Congenital heart surgery
- Cardiopulmonary bypass
- Neonate

KEY POINTS
- Premature neonates are more likely to be born with congenital heart disease (CHD), and neonates with CHD are more likely to be born premature.
- Prematurity imparts significant morbidity and mortality risk in the neonate with CHD.
- Premature neonates with CHD may encounter hemodynamic instability during fetal transition as well as in the perioperative period.
- Management of premature neonates with CHD requires the collaboration of highly specialized providers from multiple disciplines.

INTRODUCTION

Premature newborns with CHD require unique considerations for optimal management. Despite obstetric advances and improvements in antenatal care, the rate of preterm delivery in the United States has increased in the past 20 years, and premature neonates have a more than 2-fold higher risk of cardiovascular abnormalities. As antenatal diagnosis of CHD improves and fetal interventions expand, preterm infants with CHD will be a growing population.

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The standard definitions of prematurity and low birth weight (LBW) are described in Table 1. Infants may be LBW (due to prematurity alone) or small for gestational age (SGA). Cardiovascular abnormalities are associated with SGA and a 2-fold risk of prematurity (<37 weeks’ gestational age [GA]). Furthermore, congenital anomalies or genetic or chromosomal abnormalities are frequently associated with both CHD and intrauterine growth restriction. Both gestational age and birth weight are important factors with regard to timing of delivery and timing of surgical repair.

TIMING OF DELIVERY AND OBSTETRIC CONSIDERATIONS

Although preterm delivery of infants with CHD may be indicated due to maternal or fetal issues, many cases of known CHD are scheduled to deliver at term gestation to facilitate coordination of care. The notion that all term babies with CHD have equivalent outcomes, however, was recently challenged by a large study of the Society of Thoracic Surgeons Congenital Heart Surgery Database. This study found that outcomes were worse for neonates born at 37 to 38 weeks’ gestation compared with those born at 39.5 weeks’ gestation. These early-term infants had higher in-hospital mortality (adjusted odds ratio for mortality of 1.34 [1.05–1.71]), more postoperative complications, and prolonged length of stay. Other studies have confirmed a higher mortality rate and greater resource utilization with delivery of CHD infants at 39 to 40 weeks’ gestation. Furthermore, Goff and colleagues found that adjusted neurodevelopmental outcomes at age 4 years were significantly improved for infants with CHD born between 39 and 40 weeks’ gestation compared with those born between 36 and 38 weeks’ gestation. Delayed brain maturation by up to 1 month in infants with CHD has been demonstrated by fetal and postnatal MRI and may contribute to the poorer neurodevelopment seen in even late preterm infants or early-term infants compared with those born at 39 to 40 weeks’ gestation.

American College of Obstetricians and Gynecologists guidelines recommend a gestational age of at least 39 weeks for all elective deliveries, even in infants without CHD, due to the increased risk for mortality and morbidity. 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.

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Categories for prematurity and low birth weight</th>
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<tr>
<td></td>
<td>Gestational Age</td>
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<tr>
<td>Preterm</td>
<td>&lt;37 wk</td>
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<tr>
<td>Extremely preterm</td>
<td>&lt;28 wk</td>
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<tr>
<td>Late preterm</td>
<td>34–36 6/7 wk</td>
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<tr>
<td>Early term</td>
<td>37–38 6/7 wk</td>
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<td></td>
<td>Birth Weight</td>
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<tr>
<td>LBW</td>
<td>&lt;2500 g</td>
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<tr>
<td>VLBW</td>
<td>&lt;1500 g</td>
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<tr>
<td>ELBW</td>
<td>&lt;1000 g</td>
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PREOPERATIVE CONSIDERATIONS

Abnormal hemodynamics may create a vulnerable period for preoperative, preterm infants with CHD. 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. Specific preoperative considerations in the preterm infant are outlined in Table 2.

Timing of Cardiac Surgery

Having considered the important transition from fetal to postnatal physiology, the clinician must then consider the implications of surgical palliation or repair of the congenital heart defect. Historically, pediatric cardiovascular surgeons were unable to repair even simple cardiac defects in small or preterm neonates. As appropriate-sized instruments were developed and surgical techniques for CHD improved, surgeons began to consider the elective palliation or repair of these challenging patients.16,17 Today, although a surgical approach to most congenital heart defects is technically possible, debate remains regarding the appropriate timing of referral for congenital heart surgery in the preterm neonate.

Most studies of preterm, LBW, or SGA newborns with CHD have demonstrated higher morbidity and mortality after congenital heart surgery.18–22 One study, however, of 102 patients found that emergency surgery and low cardiac output syndrome (but not prematurity or SGA, among other factors) increased the risk of mortality,23 and the importance of LBW alone in relation to mortality has been questioned.24 Retrospective outcome analyses have influenced clinicians to delay cardiac surgery until a newborn matures, allowing for weight gain and organ maturity. Delaying surgery, however (to achieve weight gain, for example), has not improved outcomes and in most cases exposes the neonate to a significant hemodynamic burden. Although individual centers have reported success with a delayed surgical approach to the management of preterm congenital heart defects,25,26 no randomized study has demonstrated superior results to early primary repair or palliation. Hickey and colleagues20 identified 2 kg as an “inflection point,” below which mortality from CHD significantly increases. Although their study found no mortality difference between a delayed approach to allow maturation compared with an immediate surgical approach, the complication rate was significantly higher in the group with a delayed surgical repair. Because delayed repair or initial palliation may impose unacceptable hemodynamics and equal risk of mortality,20 many centers advocate for early primary repair of premature or LBW neonates, despite the inherent risks.27–29 The appropriate timing for intervention remains unclear due to the heterogeneous patient population, the center-specific surgical approaches, and the unclear impact that timing plays on neurodevelopmental outcome.

The specific anatomic diagnosis has a significant impact on outcomes in preterm neonates with CHD.30 In a meta-analysis of 6 studies, including 356 LBW neonates, cardiac diagnosis was the most important predictor of mortality, and a complete physiological repair demonstrated slightly improved survival compared with surgical palliation.30 Selected individual congenital cardiac lesions have been retrospectively studied to determine the optimal approach for each lesion in the preterm (or LBW) infant.
<table>
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<tr>
<th>System</th>
<th>Physiologic Challenges</th>
<th>Considerations for Preoperative Management</th>
</tr>
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<tbody>
<tr>
<td>Cardiac development</td>
<td>• Postnatal adjustment to physiology of congenital heart defect&lt;sup&gt;16,31&lt;/sup&gt;</td>
<td>• Anticipate significant hemodynamic changes in initial 24 h of life.</td>
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<tr>
<td></td>
<td>• Neonatal sarcoplasmic reticulum is poorly developed.</td>
<td>• Ensure adequate ionized calcium levels for optimal cardiac contractility.</td>
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<td></td>
<td>• Neonatal myocardium uses glucose for energy (compared with fat in older children).</td>
<td>• Avoid hypoglycemia because low hepatic glycogen stores and impaired gluconeogenesis may lead to cardiac dysfunction.</td>
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<td></td>
<td>• Neonatal ventricular muscle poorly tolerant of pressure/volume load</td>
<td>• Cardiac output reliant on heart rate; may not tolerate pressure/volume load.</td>
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<tr>
<td>Fluids and electrolytes</td>
<td>• Increased fluid/caloric requirements and excessive heat loss from thin stratum corneum</td>
<td>• Maintain adequate fluid and caloric intake; consider careful diuretic use to reduce pulmonary edema/volume overload.</td>
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<td></td>
<td>• Immature renal function</td>
<td>• Monitor closely for electrolyte derangement (especially hyponatremia).</td>
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<td></td>
<td>• Acidosis due to elevated lactate or renal bicarbonate loss</td>
<td>• Careful consideration of bicarbonate use to correct acidosis and avoid cardiac dysfunction (increased risk of IVH in preterm neonates)&lt;sup&gt;32,33&lt;/sup&gt;</td>
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<tr>
<td>Cerebral blood flow and hemodynamics</td>
<td>• Adequate blood pressure for organ perfusion is necessary.</td>
<td>• Goal mean arterial pressure above a value equivalent to the gestational age (or &gt;30 mm Hg) has been utilized&lt;sup&gt;35,36&lt;/sup&gt;; CHD may warrant different blood pressure thresholds.</td>
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<td>• Preterm infants have impaired cerebral autoregulation&lt;sup&gt;34&lt;/sup&gt; and are at greater risk for IVH/hypoxic brain injury.</td>
<td>• Preserve cerebral blood flow by minimizing hypotension, hypocarbia, or anemia.</td>
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<td>• Near-infrared spectroscopy monitoring&lt;sup&gt;37&lt;/sup&gt; may help with assessment of end organ perfusion.</td>
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<td>Feeding and nutrition</td>
<td>• Higher-energy expenditure and greater risk for failure to thrive</td>
<td>• Consider early initiation of trophic feeds; monitor tolerance closely given increased risk for NEC with CHD.&lt;sup&gt;38,39&lt;/sup&gt;</td>
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<td>• Increased risk of NEC due to immature gut, altered intestinal microbiome, decreased gut perfusion, infection/inflammation</td>
<td>• Data lacking for preoperative enteral feeding in preterm infant, especially when requiring prostaglandins or pressors&lt;sup&gt;40,41&lt;/sup&gt;</td>
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Specific Lesions in the Premature Neonate

Coarctation of the aorta

Aortic coarctation represents a surgically correctable lesion in the premature or LBW neonate. Although balloon angioplasty remains an option for certain populations, the high rate of restenosis remains an unacceptable outcome in most native aortic coarctation. Small studies of newborns weighing less than 2 kg with uncomplicated, isolated coarctation of the aorta have demonstrated a high surgical success rate with acceptable mortality rates, but mortality increases with addition of associated left-sided cardiac abnormalities. Recoarctation has been reported in 15% to 40% of patients, although contemporary results may reflect a lower recurrence rate (Reddy VM, unpublished data, 2015), patients must be closely followed for growth of the aortic arch and coarctation site. Recoarctation may not relate to LBW but rather the anatomy of the aortic arch and coarctation site. Based on these studies, there seems little indication for delaying surgical repair for the premature patient with an isolated coarctation. For a complicated premature or LBW newborn or for patients with significant

<table>
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<td><strong>System</strong></td>
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<td>RDS, apnea of prematurity</td>
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<td>Preoperative genetic testing</td>
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<td>Neurologic Imaging</td>
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surgical contraindications (such as extracardiac complications), a hybrid approach with initial stenting of the aortic coarctation followed by subsequent surgical repair has been reported.\textsuperscript{56}

**Transposition of the great arteries**
Premature neonates with transposition of the great arteries (TGA) require surgical correction with an arterial switch operation, which includes transfer of the coronary arteries to the neoaortic (native pulmonary) root. Historically, surgery was delayed with continuous prostaglandin infusion to maintain ductal patency, in conjunction with a balloon atrial septostomy (Rashkind procedure) to promote atrial mixing. With the advent of neonatal surgery, however, repair within the first week of life prevents muscular involution of the left ventricle and is now the standard of care. Data suggest that longer time to surgery relates to preoperative brain injury for a neonate with TGA.\textsuperscript{57} A premature neonate with TGA, however, is likely to experience a prolonged length of stay in the postoperative ICU\textsuperscript{58} and still has a higher risk of early postoperative mortality compared with a normal birth weight neonate.\textsuperscript{59,60} Although the operative mortality may approximate 15% to 20% for premature or LBW neonates after arterial switch operation, Roussin and colleagues\textsuperscript{60} found that both surgical palliation (not repair) and age greater than 30 days at the time of surgery increased the mortality risk. Therefore, data suggest that early primary repair for TGA represents a high-risk but optimal surgical approach.

**Hypoplastic left heart syndrome**
Hypoplastic left heart syndrome (HLHS) represents one of the most complex and challenging congenital heart lesions to manage, even in term neonates with normal birth weight. With a mortality rate of 25% to 30% in the first year after staged single-ventricle palliation, pediatric cardiologists and cardiothoracic surgeons continue to study optimal surgical approaches.\textsuperscript{61} In premature neonates, the important morbidity and mortality of HLHS are amplified, especially after stage I palliation (Norwood procedure). Even late preterm birth imposes significant risk of death for newborns with HLHS; compared with term neonates, late preterm birth was associated with an almost 3-fold odds of hospital death in HLHS patients.\textsuperscript{62} Newborns with HLHS are also more likely to be born premature and LBW, both factors that increase the risk of early death.\textsuperscript{63} A recent evaluation of 47 patients after surgical palliation for HLHS reported 49% mortality, with SGA and noncardiac abnormalities representing significant risk factors for death.\textsuperscript{64} An earlier study of 27 neonates with HLHS reported similar mortality but found that pulmonary vein and coronary artery anomalies were related to poor patient outcome (not prematurity, weight, or SGA).\textsuperscript{65}

**Aortic arch obstruction (2-ventricle surgical repair)**
The complexity of the single-ventricle reconstruction inherent in the stage I palliation is underscored by the contrasting outcomes in premature neonates with aortic arch obstruction and ventricular septal defect who undergo a 2-ventricle repair. Haas and colleagues\textsuperscript{66} studied 21 newborns with variants of aortic arch obstruction/ventricular septal defect and found a mortality of only 14%; these data suggest that early complete anatomic repair can significantly reduce mortality in preterm and LBW newborns. Underlying anatomy influences both surgical decision making and patient outcome. Neonates with all types of single-ventricle physiology (including HLHS, unbalanced atroventricular canal, and tricuspid atresia) all experience higher rates of preterm birth, low birth rate, and SGA,\textsuperscript{67} emphasizing the need to further study and address this important preoperative risk factor for this complex population.
**Tetralogy of Fallot**

Tetralogy of Fallot (TOF) is the most common cyanotic heart defect and was the first cyanotic lesion palliated with congenital heart surgery. Historically, TOF was palliated with a surgical aortopulmonary shunt to augment pulmonary blood flow, followed by complete intracardiac repair before 1 year of age. With advances in neonatal surgical and cardiopulmonary bypass (CPB) techniques, however, many centers advocate for early primary repair. Referral for surgery typically occurs at approximately 3 to 6 months of age, before right ventricular and infundibular hypertrophy worsens enough to induce a hypercyanotic episode. Hypercyanotic episodes (tet spells) define an indication for immediate surgery. Therefore, for symptomatic neonates, some centers perform a complete intracardiac repair of TOF.

Unlike other congenital cardiac surgeries, however, repair of TOF increases the combined ventricular volume compared with the preoperative state. Neonatal repair also may lead to a relatively larger right ventriculotomy scar and more frequent use of a transannular patch (leading to more severe pulmonary insufficiency). Neonates have noncompliant ventricular muscle that is resistant to volume loading and, therefore, may struggle to recover from early primary repair. Studies of the surgical approach to TOF indicate that early primary repair (at 6–8 weeks of age) is technically feasible and may not be associated with increased morbidity or mortality. Even though prematurity and LBW increase the time of mechanical ventilation, length of stay, and cost, surgical repair (with maintenance of a postoperative patent foramen ovale to allow decompression of the noncompliant right ventricle) may be a better option than the alternative, a surgically created aortopulmonary shunt. Systemic to pulmonary shunts are difficult to size appropriately in LBW and premature neonates, and may pose a higher mortality risk in neonates in general. One study of surgical aortopulmonary shunts in patients less than 3 kg described acceptable overall survival but increased reintervention with smaller (3-mm) shunts required for the LBW and premature patients. One case series described interventional stenting of the right ventricular outflow tract in patients who were not surgical candidates; this approach may be attractive in specific subpopulations (eg, LBW neonates with contraindications to cardiopulmonary bypass).

**Postoperative Considerations**

After cardiac surgery, premature neonates are at risk for complications inherent to the intraoperative course. CPB imposes significant hemodynamic and inflammatory insults that have been well described in varied populations. Strategies to mitigate the inflammatory cascade, such as intraoperative ultrafiltration and steroid administration, are common at most institutions. Systemic heparinization is essential during CPB, and any preoperative bleeding concerns may be magnified in the postoperative period. Although heparinization can be safely performed without significant morbidity in this population, clinicians should consider the risks in patients with preoperative intraventricular or gastrointestinal bleeding.

Optimal management of postsurgical cardiac neonates requires a dedicated team approach. 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; it is not uncommon for neonatologists at the authors’ institution to provide postoperative guidance in the cardiovascular ICU.
OUTCOMES: MORTALITY AND COMMON COMPLICATIONS OF PREMATURITY

Several multicenter studies have confirmed a mortality rate between 1.5 and 4 times higher in LBW, very LBW (VLBW), and extremely LBW (ELBW) infants with CHD compared with their counterparts of similar birth weight without CHD. A description of the most common and important morbidities after surgical repair of CHD follows.

 Bronchopulmonary Dysplasia

VLBW infants with CHD were found more than 4 times as likely to develop bronchopulmonary dysplasia (BPD) compared with those without CHD, likely due to prolonged mechanical ventilation and pulmonary edema. BPD has significant impact on the morbidity of neonates with CHD, especially in patients with single-ventricle physiology. Clinicians must pay careful attention to the goals of mechanical ventilation, to avoid the development of ventilator-induced lung injury. The goal of low tidal volume (4–6 mL per kilogram), shorter inspiratory time, and permissive hypercapnia can be achieved in some neonates, but hypercapnia may alter cardiopulmonary hemodynamics by elevating pulmonary vascular resistance, causing dilation of the systemic vascular bed, and impairing cardiac contractility. Nitric oxide may mitigate the effects of hypercapnia and elevated pulmonary vascular resistance, but data in premature neonates with CHD are lacking.

Postoperative management of premature or LBW neonates with CHD may also present a unique challenge when titrating fraction of inspired oxygen concentration (FiO₂). The detrimental effects of hyperoxegenation should be emphasized in managing the ventilator, because reactive oxygen species have been implicated in retinopathy of prematurity (ROP) and also BPD. Therefore, the authors’ postoperative management includes administering the minimum FiO₂ to achieve the goals for the cardiac repair. Patients with single-ventricle disease are rapidly titrated to minimal oxygen levels but may still require supplemental oxygen (FiO₂ approximately 0.3) to overcome pulmonary venous desaturation common after CPB. Patients with fully septated intracardiac anatomy should be maintained at an oxygen level adequate to oxygenate tissues and allow organ recovery; neonates rarely require a PaO₂ greater than 100, even in the setting of right ventricular dysfunction or pulmonary hypertension. The authors’ general approach is to target a PaO₂ and PacO₂ of 50 mm Hg to optimize postoperative ventilation (rather than increasing ventilator pressure or respiratory rates to achieve a lower PaCO₂ or higher PaO₂). In select patients, high-frequency oscillation has been used in an attempt to minimize of ventilator-induced lung injury.

 Sepsis

Small studies have demonstrated a greater than 2-times higher incidence of sepsis in preterm infants with CHD compared with those without CHD as well as higher rates of nosocomial infections in preterm infants with CHD compared with those born at 39 weeks’ gestation. These findings may be attributable to longer durations of hospital stay, mechanical ventilation, and central venous catheter placement in preterm infants with CHD. Premature and LBW newborns are also at significantly increased risk of invasive fungal infections. Fungal infections in neonates with CHD impart a mortality risk as high as 21%; some clinicians opt for early antifungal prophylaxis in a high-risk premature neonate (eg, delayed sternal closure). Many patients require stable preoperative vascular access, such as a peripherally inserted central catheter or an umbilical venous catheter for medication administration (such as prostaglandin). These catheters put patients at risk for central line-associated blood stream infections and should be attentively managed with bundled care protocols. The authors’ preference
is to place a right atrial Broviac catheter at the time of chest closure in the operating room or with subsequent sternal closure.

**Necrotizing Enterocolitis**

NEC is a known complication of prematurity, and CHD is a risk factor for development of NEC. A large study of VLBW infants from the Vermont Oxford Network (VON) found a higher incidence of NEC when CHD was also present (adjusted odds ratio 1.8; \( P < .0001 \)). Although lower birth weight typically is a predictor of higher mortality in VLBW infants with NEC, this study did not demonstrate an effect of birth weight on mortality for patients with both CHD and NEC. Although splanchnic ischemia and associated intestinal tissue hypoxia are contributing risks, a different pathophysiologic mechanism for NEC in preterm infants with CHD may predominate. Different specific cardiac lesions, including HLHS\(^{85}\) and atrioventricular canal defect,\(^{38}\) have been associated with increased risk for NEC, but no consistent cardiac pathophysiology has been identified (See Karpen HE: Nutrition in the Cardiac Newborn Evidence-based Nutrition Guidelines for the Cardiac Newborn, in this issue).

**Intraventricular Hemorrhage and Periventricular Leukomalacia**

Both preterm infants without CHD and term infants with CHD are susceptible to white matter injury in the brain. The selective vulnerability of developing oligodendrocytes in an environment of both ischemia and inflammation may contribute to injury,\(^{86}\) but no studies have specifically focused on unique patterns of brain injury with the combined risks of prematurity and CHD. Literature also remains inconsistent regarding increased risk for IVH in preterm infants with CHD.\(^{21,77}\) Intraoperative variables may also contribute to the risk of neurologic injury. Specifically, hemodilution of hematocrit (caused by the priming volume of the CPB circuit) may contribute to lower oxygen delivery to the brain.\(^{87}\) Similarly, the hypothermia required to perform complex cardiac repairs may alter cerebral autoregulation and blood flow; many pediatric centers use a pH-stat management during CPB to correct for this alteration.\(^{88}\) Finally, the choice of anesthetic agent has become increasingly scrutinized, because animal models have demonstrated neurotoxicity with certain drug combinations.\(^{89}\) No human data have confirmed these findings, and there is consensus that inadequate analgesia during cardiac surgery results in important negative hemodynamic and behavioral changes. To optimize the treatment of premature neonates, further study of specific anesthetic agents is warranted.

**Retinopathy of Prematurity**

Disturbances in retinal perfusion, increased VEGF (vascular endothelial growth factor) expression from hypoxia, and ensuing neovascularization potentially lead to worsening ROP in preterm infants with CHD. A large study of ELBW infants found that the ROP rate was not significantly different between infants with and without CHD, but the infants with CHD had an increased risk of bilateral blindness (relative risk 7.8; CI, 2.5–23.9). It is unclear if this increased risk was due to central nervous system injury of the visual cortex or due to increased abnormalities specific to the lens or retina of the eyes.\(^{78}\) Limitation of \(\text{FiO}_2\) during mechanical ventilation (as described previously) may also prevent hypervascularization of the retina and ROP.

**Neurodevelopmental Outcomes and Follow-up Needs**

Close neurodevelopmental follow-up and engagement with early intervention services as needed for infants with CHD are recommended and particularly necessary in premature infants due to inherent neurodevelopmental risk associated with prematurity alone. A large study of ELBW infants in the National Institute of Child Health and Human
Development Neonatal Research Network centers found increased mortality and risk for neurodevelopmental impairment at 18 to 22 months of age in infants with isolated CHD compared with those without CHD (relative risk 1.43; CI, 1.29–1.58). This study found poorer growth parameters and longer length of hospital stay in the infants with CHD. Delayed brain maturation of infants with CHD may further predispose these infants to neurodevelopmental impairment. Specific evaluation of gross and fine motor skills, speech and language, executive functioning, and behavior is necessary with formal developmental testing and early referral for services.

SUMMARY

In summary, premature neonates with CHD require the collaborative effort of highly specialized teams to manage the perioperative course. Preoperative complications of prematurity are amplified in the setting of hemodynamic disturbances that are common with CHD. The decision to proceed with surgery and the surgical procedure performed are important determinants of postoperative hemodynamics. Postoperative complications may arise in virtually every organ system and require the close attention of clinicians. As more is learned about the management of these comorbidities, improved outcomes will follow for premature neonates with CHD.

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**What is the current practice?**

Best practice/guideline/care path objective(s):

In the absence of maternal or fetal indications for early delivery, the fetus with CHD should be delivered at term.

The premature neonate with CHD should be managed by a multidisciplinary team with experience in complex neonatal and cardiac care.

When technically possible (and in the absence of morbidities or contraindications), early primary surgical repair should be performed for congenital heart lesions with significant hemodynamic abnormalities; a delayed surgical approach may allow for weight gain in anticipation of subsequent repair but may lead to unacceptable cardiac pressure and volume load.

**What changes in current practice are likely to improve outcomes?**

The continued improvement in fetal diagnosis may provide an opportunity for optimal timing of delivery (39–40 weeks’ gestation).

Early surgical repair of hemodynamically significant cardiac lesions may lead to improved cardiac physiology.

The impact of CPB and congenital heart surgery on the premature developing brain and subsequent neurodevelopmental outcomes, however, is unclear. Further study of these outcomes is warranted.

Continued collaboration of multidisciplinary teams, including neonatologists, cardiologists, anesthesiologists, cardiac surgeons, intensivists, nurses, and advanced practice providers, will be critical in the management of postoperative complications in this fragile population.

**Summary statement**

Premature neonates with CHD represent a complex and high-risk population that requires highly specialized medical care. When unable to achieve term delivery, a neonate with CHD provides important challenges to the clinician. A collaborative approach to the surgical decision making and postoperative management of complications is required.
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