The Critically Ill Infant with Congenital Heart Disease

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KEYWORDS

• Congenital heart disease • Cyanotic cardiac disease • Pediatric cardiology
• Critically ill neonate • Decompensated neonate

KEY POINTS

- Neonates presenting with acute and profound systemic hypoperfusion or cyanosis have a ductal-dependent cardiac lesion until proven otherwise.
- Ductal-dependent lesions typically present within the first 2 weeks of life, whereas shunting lesions with heart failure present within 1 to 6 months of life.
- Prostaglandin E1 is a life-saving medication. Be wary of its adverse effects of apnea and hypotension.
- Essential elements in the evaluation of a critically ill infant with congenital cardiac disease include (1) right upper and lower extremity blood pressure, (2) pulse oximetry, (3) brachial–femoral pulse differential, (4) electrocardiography, (5) chest radiography, (6) brain natriuretic peptide, and (7) bedside echocardiogram.

INTRODUCTION

Critically ill infants presenting to the emergency department (ED) inherently produce anxiety for emergency physicians (EPs). They often have nonspecific or subtle findings, making timely accurate diagnosis and implementation of life-saving interventions fraught with difficulty. The differential diagnosis for an ill neonate is best remembered by the mnemonic THE MISFITS (**Box 1**).[1] Children with congenital cardiac disease are especially challenging to diagnose and manage because of their complex physiology

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Box 1
THE MISFITS

- *T*rauma (accidental and nonaccidental)
- *H*eart disease and hypovolemia
- *E*ndocrine (congenital adrenal hyperplasia and thyrotoxicosis)
- *M*etabolic (hypocalcemia, hypoglycemia, etc)
- *I*nborn errors of metabolism
- *S*epsis
- *F*ormula dilution
- *I*ntestinal catastrophes (necrotizing enterocolitis, volvulus, intussusception)
- *T*oxins
- *S*eizures

and age-dependent variability. This article discusses the tools necessary for the identification and initial ED management of the infant with undifferentiated decompensated congenital heart disease (CHD).

Critical congenital cardiac lesions can be classified into 3 broad categories based on physiology: left-sided obstructive ductal dependent, right-sided obstructive ductal dependent, and shunting or mixing lesions. As with all pediatric cases, physiology is age dependent, so the child's age at presentation is among the most important variables to consider. Infants who present early—in the first month of life—most likely have a ductal-dependent lesion. They might be cyanotic from an obstructive right heart lesion or, more commonly, profoundly hypoperfused from an obstructive left heart lesion. After 1 month of age, infants most likely present in respiratory distress or congestive heart failure caused by a left-to-right shunt.[2] A rapid compilation of data obtained from the history and physical examination, focusing on essential elements (**Box 2**), can provide clues to the presence and physiology of the cardiac lesion. Subsequent interventions, such as use of supplemental oxygen, initiation of prostaglandin E1 (PGE1), or administration of fluid boluses can then be tailored to fit the patient's unique physiology.

Box 2
Toolkit for identification of a congenital cardiac disease

- Hyperoxia test
- Weight gain, murmur, hepatomegaly, brachial–femoral pulse differential
- Right upper and lower extremity blood pressure differential of greater than 10 mm Hg
- Pulse oximetry differential of greater than 3%, or less than 94% in lower extremity, or less than 90% in any extremity
- Electrocardiography (ECG)
- Chest radiography (CXR)
- Brain natriuretic peptide (BNP)
- Bedside limited echocardiography by the emergency physician (BLEEP)