Heart Disease in Children

Richard U. Garcia, MD*, Stacie B. Peddy, MD

KEYWORDS

• Congenital heart disease • Children • Primary care • Cyanosis • Chest pain
• Heart murmur • Infective endocarditis

KEY POINTS

- Fetal and neonatal diagnosis of congenital heart disease (CHD) has improved the outcomes for children born with critical CHD.
- Treatment and management of CHD has improved significantly over the past 2 decades, allowing more children with CHD to grow into adulthood.
- Appropriate diagnosis and treatment of group A pharyngitis and Kawasaki disease in pediatric patients mitigate late complications.
- Chest pain, syncope, and irregular heart rhythm are common presentations in primary care. Although typically benign, red flag symptoms/signs should prompt a referral to cardiology for further evaluation.

INTRODUCTION

The modern incidence of congenital heart disease (CHD) has been reported at 6 to 11.1 per 1000 live births.[1,2] The true incidence is likely higher because many miscarriages are due to heart conditions incompatible with life. The unique physiology of CHD, the constantly developing nature of children, the differing presenting signs and symptoms, the multiple palliative or corrective surgeries, and the constant development of new strategies directed toward improving care in this population make pediatric cardiology an exciting field in modern medicine.

THE FETAL CIRCULATION AND TRANSITION TO NEONATAL LIFE

Cardiovascular morphogenesis is a complex process that transforms an initial single-tube heart to a 4-chamber heart with 2 separate outflow tracts. Multiple and

Disclosure Statement: All Authors take responsibility for all aspects of the reliability and freedom from bias of the information presented and their discussed interpretation. No conflict of interest, grants, or other financial support was received for this article.
Division of Cardiac Critical Care Medicine, Departments of Pediatrics and Critical Care Medicine, The University of Pennsylvania and the Children's Hospital of Philadelphia, 34th Street, Civic Center Boulevard, Philadelphia, PA 19104, USA
* Corresponding author.
E-mail address: rugsal25@gmail.com

Prim Care Clin Office Pract 45 (2018) 143–154
https://doi.org/10.1016/j.pop.2017.10.005
primarycare.theclinics.com

overlapping signaling events make this possible and when the process deviates from normal, CHD occurs.[3] The fetal circulation occurs in a parallel circuit. Because the source of oxygenation in fetal life is the placenta, only 10% of the combined cardiac output reaches the fetal lungs.[4] Because of a mechanism of preferential blood flow streaming, the oxygenated blood coming back from the placenta bypasses the liver through the ductus venosum, enters the right atrium by the inferior vena cavae, and is streamed through the patent foramen ovale to the left atria, providing the brain with highly oxygenated blood. Only approximately 10% of the combined cardiac output goes through the aortic isthmus to the descending aorta. Fetal systemic venous return enters the right atrium through the superior and inferior vena cavae and streams toward the tricuspid valve to reach the pulmonary artery, subsequently crossing the patent ductus arteriosus almost in its entirety (approximately 50% of the combined cardiac output) to reach the placenta through the descending aorta.[4]

These unique characteristics of the fetal circulation allow for tolerance of complex heart disease like hypoplastic left heart syndrome in utero with minimal hemodynamic consequences. The extra utero circulation, however, occurs in parallel. Once a fetus takes that important first breath, a cascade of events leads to significant hemodynamic changes, making tolerance of complex CHD difficult.

SCREENING FOR CONGENITAL HEART DISEASE: WHEN AND WHO?

The etiology of CHD is multifactorial. Prenatal diagnosis of critical neonatal CHD affects neonatal morbidity and (to a lesser extent), mortality.[5] Accurate prenatal diagnosis also contributes to preservation of long-term neurocognitive function and outcome,[6] highlighting the importance of accurate prenatal screening and prompt diagnosis. The risk for CHD in the general population is less than 1% and in this setting, fetal echocardiography screening is not recommended. If, however, the risk for CHD is greater than 3% (**Table 1**), fetal echocardiography should be performed. When the risk for CHD is estimated at 1% to 2%, fetal echocardiography can be considered, although the relative benefit of such additional testing in this population is not clear. In all cases, fetuses with an abnormal screening ultrasound of the heart should have a detailed fetal echocardiogram read by a trained examiner.[5]

Kemper and colleagues[7] made recommendations for universal neonatal screening to diagnose critical CHD (CCHD). These recommendations included measurement of the percutaneous oxygen saturation in the right hand and feet between 24 hours to 48 hours of birth or just before discharge if a neonate leaves the hospital before 24 hours of life.

A screen is positive screening if

- The oxygen saturation is below 90% in the right hand *OR* feet during 3 consecutive measurements separated by 1 hour.
- The oxygen saturation is below 95% and above 90% in the right hand *AND* feet *OR* more than a 3% difference between the right hand and feet saturation during 3 consecutive measurements separated by 1 hour.

The screen is negative if

- The oxygen saturation is above 95% in the right hand *OR* feet *AND* less than a 3% difference between the right hand and feet saturation at *any point* during 3 consecutive measurements separated by 1 hour.

These recommendations were endorsed by American Academy of Pediatrics, the American College of Cardiology Foundation, and the American Heart Association