

Recognition of Undiagnosed Neonatal Heart Disease



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KEYWORDS

- Aortic transposition • Left-to-right shunt • Cyanosis • Left heart obstructive lesions
- Foramen ovale • Pulmonary hypertension of the newborn • Hypoperfusion
- Right-to-left shunt

KEY POINTS

- Symptomatic congenital heart disease usually presents in early infancy and may present as 1 of 3 clinical presentations: cyanosis, hypoperfusion, or respiratory distress/failure to thrive (without hypoperfusion).
- Cyanotic heart disease can be subdivided into lesions with decreased pulmonary blood flow and those with the aorta receiving blood from the venous ventricle (aortic transposition in which the circulations are mostly separated).
- Hypoperfusion is caused either by obstruction to the inflow or outflow of the left ventricle or by the inability of the left ventricle to deliver an adequate systemic blood flow in the absence of obstruction.
- Respiratory distress/failure to thrive caused by congenital cardiac defects has the common pathophysiological problem of a left-to-right shunt causing excessive pulmonary blood flow, with or without an associated right-to-left shunt.
- The initial evaluation of the newborn with symptomatic heart disease should be directed at defining the presenting pathophysiological process, and the initial management should be similarly focused.

INTRODUCTION

Heart defects are the most common congenital malformation. In the general population, the incidence is about 7 to 10 per 1000 live births (excluding bicuspid aortic valve and hemodynamically insignificant lesions such as very small secundum atrial and muscular ventricular septal defects [VSDs]), and the incidence is much higher in

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stillborn infants and abortuses.^{1,2} About 60% of newborns with heart defects require an intervention, either surgery or catheter-based, at some time in their lives. About half of those require intervention in the first year of life, and two-thirds of this latter group (20% of all heart defects) are critical, defined as a defect that requires an intervention within the first month of life or serious morbidity or mortality is likely. At the current birth rate in the United States, this means that there are approximately 40,000 infants born each year with congenital heart disease and that approximately 8000 require diagnosis and intervention in the newborn period to avoid severe injury or death. Thus, early recognition of symptomatic heart disease is essential to prevent morbidity in the many infants born with critical heart disease each year.³⁻⁵

The following sections elucidate the physiology, presentation, and general approach to the treatment of symptomatic heart disease in the newborn.

CONGENITAL HEART DEFECTS

The number of distinct congenital heart defects is sufficiently large that pediatric cardiologists with more than 30 years' experience still see congenital heart defects that they have never seen before. Thus, it is necessary to take a logical approach to understanding congenital heart disease rather than an approach in which the various lesions are memorized. Moreover, treatment of cardiac defects in the newborn requires a physiology-based approach, to ameliorate the cardiovascular derangements that are caused by the transition from the fetal to newborn circulation. It is noteworthy that, because of the presence of 3 interconnected circulations in the fetus (the systemic, pulmonary, and placental circulation, connected by the ductus venosus, the ductus arteriosus, and the foramen ovale), even the most complex cardiac defect rarely causes symptoms in the fetus. Birth is associated with abolition of the placental circulation and functional closure of the ductus arteriosus and foramen ovale, leading to 2 circulations in series, both requiring normal vascular connections and biventricular function for hemodynamic stability and adequate oxygen uptake and delivery. Thus, it is not surprising that symptomatic heart disease frequently occurs in the newborn period.

The most effective and logical approach is to consider the physiologic consequences of congenital heart defects that cause symptoms in the newborn. Using this approach, heart defects can be separated into 3 symptomatic modes of presentation.⁶ They are

1. Cyanosis (decreased arterial oxygen saturation causing central cyanosis, which impairs systemic oxygen delivery)
2. Hypoperfusion (decreased systemic perfusion of either the upper or the lower body circulation or both)
3. Respiratory distress/failure to thrive (visible distress or subclinical, leading to poor growth; this is in the presence of normal perfusion)

Within each of the 3 modes of presentation, there are 2 pathophysiologic processes that may cause that presentation. These modes of presentation are

1. Cyanosis
 - a. Decreased pulmonary blood flow
 - b. Separated systemic and pulmonary circulations (variants of d-transposition of the great arteries in which the aorta is malposed over the systemic venous [SV] ventricle)
2. Hypoperfusion
 - a. Left-sided obstruction

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