Perinatal and Delivery Management of Infants with Congenital Heart Disease

Laura Sanapo, MD[a], Anita J. Moon-Grady, MD[b], Mary T. Donofrio, MD[a,c,*]

KEYWORDS

• Fetal echocardiography • Fetal cardiology • Congenital heart disease
• Neurodevelopment • Biophysical profile • Obstetric management
• Antenatal surveillance

KEY POINTS

- Prenatal diagnosis has improved neonatal outcomes of congenital heart disease (CHD) but perinatal morbidity and mortality are still significant in some cases.
- Fetal echocardiography can facilitate delivery and perinatal planning for infants with CHD.
- Antenatal surveillance of fetuses with CHD can identify prenatal progression of the lesion and decompensation, and may improve perinatal and postnatal outcomes.
- Successful perinatal management of neonates with a prenatal diagnosis of CHD requires close collaboration between obstetric, neonatal, and cardiology services.
- Delivery of infants prenatally diagnosed with CHD in most cases should not be scheduled before 39 weeks unless there is an obstetric indication or concern regarding fetal well-being.

Video content accompanies this article at http://www.perinatology.theclinics.com/

INTRODUCTION

Advances in prenatal imaging and increasing experience in fetal cardiology have improved the examination of the fetal cardiovascular system.[1] Fetal echocardiography

Disclosure: The authors have nothing to disclose.
[a] Division of Fetal and Transitional Medicine, Children's National Health System, 111 Michigan Avenue, Northwest, Suite M3-118, Washington, DC 20010, USA; [b] Fetal Cardiovascular Program, UCSF Benioff Children's Hospitals, University of California San Francisco, 550 16th Street, 5th Floor, Box 0544, San Francisco, CA 94158, USA; [c] Fetal Heart Program, Division of Cardiology, Children's National Health System, 111 Michigan Avenue, Northwest, Washington, DC 20010, USA
* Corresponding author. Fetal Heart Program, Division of Cardiology, Children's National Health System, 111 Michigan Avenue, Northwest, Washington, DC 20010.
E-mail address: MDonofri@childrensnational.org

Clin Perinatol 43 (2016) 55–71
http://dx.doi.org/10.1016/j.clp.2015.11.004 perinatology.theclinics.com
0095-5108/16/$ – see front matter

is able to obtain precise details of cardiac structural and hemodynamic alterations in fetuses with congenital heart disease (CHD). Sequential examinations through gestation can predict the evolution of disease in utero and during the transition to postnatal circulation at delivery. This approach allows detailed prenatal counseling and enables planning to define perinatal management, selecting the fetuses at risk of postnatal hemodynamic instability who might require a specialized delivery plan.[2,3] The prenatal diagnosis and management of critical neonatal CHD has been shown to play an important role in improving the outcome of newborns with these conditions, allowing timely stabilization of the disease before cardiac surgery[4] and reducing the risk of perioperative morbidity,[4–9] including the risk of perioperative neurologic insults.[10] Despite evidence that fetal diagnosis has improved the outcome of some CHDs, critical forms may still be associated with significant morbidity and mortality caused by hemodynamic instability that occurs after birth, often shortly after separation from the placental circulation.[11] Therefore, prenatal assessment of the severity of the lesion and disease-specific delivery recommendations has been suggested to ensure the best care and avoid delays in treatment.[11–14] Perinatal management of neonates with a prenatal diagnosis of CHD requires a close collaboration between obstetric, neonatal, and cardiology services, and a well-delineated network with communication between the adult hospital and pediatric tertiary care center.[3]

This article reviews the most recent recommendations for the perinatal and delivery management of infants with a prenatal diagnosis of CHD.

FETAL ECHOCARDIOGRAPHY AND RISK OF HEMODYNAMIC INSTABILITY AT BIRTH

Most CHD is well tolerated in utero, does not present a risk of hemodynamic instability at birth or in the first days of life, and does not require specialized delivery care. However, some critical CHDs have an increased risk of hemodynamic instability after delivery and may require maintenance of patency of the fetal shunts and/or immediate intervention.[2] In order to identify the fetuses with CHD at risk of hemodynamic instability at birth, it is important to understand the physiology of the fetal circulation and the transition to the extrauterine circulation and how this process is compromised in newborns with a cardiac defect.

Fetal and Transitional Circulation

The fetal circulation is a highly efficient system that provides blood to the fetal body and the placenta. Fetal shunts allow the more highly oxygenated and nutrient-rich blood from the umbilical vein to be preferentially delivered to the left ventricle, therefore entering the systemic circulation. The remainder of the umbilical vein blood mixes with less oxygenated blood from the fetal body and passes to the right heart, and via the ductus arteriosus is directed into the descending aorta supplying the lower body and the placenta.[15] The fetoplacental circulation is characterized by low resistance, whereas the circulation to the fetal lungs is limited by high resistance. At delivery, multiple important changes occur. Cord clamping interrupts the low-resistance placental circulation, whereas initiation of respiration decreases the pulmonary vascular resistance and increases pulmonary blood flow and ultimately the blood volume returning to the left atrium through the pulmonary veins. Consequently, the left atrial pressure increases and functional closure of the foramen ovale occurs. With the closure of the ductus venosus (within the first week of life) and the ductus arteriosus (usually within 12–72 hours),[16] the fetal circulation transitions to the postnatal circulation in series.[17]

In specific cases of CHD, the presence of in utero cardiac shunting permits redistribution of blood flow to maintain cardiac output and adequate oxygen delivery to