Perinatal and Delivery Management of Infants with Congenital Heart Disease

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Mary T. Donofrio, MD\textsuperscript{a,c,*}

\section*{INTRODUCTION}

Advances in prenatal imaging and increasing experience in fetal cardiology have improved the examination of the fetal cardiovascular system.\textsuperscript{1} Fetal echocardiography

\section*{KEYWORDS}
\begin{itemize}
\item Fetal echocardiography
\item Fetal cardiology
\item Congenital heart disease
\item Neurodevelopment
\item Biophysical profile
\item Obstetric management
\item Antenatal surveillance
\end{itemize}

\section*{KEY POINTS}
\begin{itemize}
\item Prenatal diagnosis has improved neonatal outcomes of congenital heart disease (CHD) but perinatal morbidity and mortality are still significant in some cases.
\item Fetal echocardiography can facilitate delivery and perinatal planning for infants with CHD.
\item Antenatal surveillance of fetuses with CHD can identify prenatal progression of the lesion and decompensation, and may improve perinatal and postnatal outcomes.
\item Successful perinatal management of neonates with a prenatal diagnosis of CHD requires close collaboration between obstetric, neonatal, and cardiology services.
\item 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.
\end{itemize}

\textbf{Video content accompanies this article at http://www.perinatology.theclinics.com/}

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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. 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 and reducing the risk of perioperative morbidity, including the risk of perioperative neurologic insults. 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. 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. 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.

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. 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. 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), the fetal circulation transitions to the postnatal circulation in series.

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
the fetal body to maintain a normal fetoplacental circulation. For this reason, most CHDs are well tolerated in utero, although the altered circulatory pattern may impair systemic oxygenation and affect fetal growth and brain development. The risk of hemodynamic instability after birth depends on 3 factors: (1) the type of CHD, including whether the defect is shunt dependent; (2) whether there is cardiac dysfunction either from a primary cardiomyopathy, structural anomaly leading to an excessive pressure or volume load to the heart, or a cardiac rhythm abnormality; and (3) whether there is an associated abnormality of the respiratory system.

Models of Risk Assessment of Hemodynamic Instability at Birth

Several postnatal risk stratification protocols for babies diagnosed in utero with CHD have been proposed and more recently summarized as part of the American Heart Association Statement on Fetal Cardiology. The risk of potential compromise at birth is most often determined prenatally by the fetal cardiologist, taking into account the specific CHD as well as patient-specific findings noted on fetal echocardiogram. A recent evaluation of a risk assessment protocol applied prospectively to a patient population at the Children's National Health System showed high accuracy with a sensitivity ranging from 0.83 to 0.99 for prediction of postnatal care and need for specialized intervention at birth.

CHDs can be divided into 3 main categories, according to the predicted risk of hemodynamic instability at birth: (1) CHD without risk of hemodynamic instability during or immediately after delivery, or in the neonatal period; (2) CHD with minimal risk of hemodynamic instability; and (3) CHD with high risk of instability.

Congenital heart disease without predicted risk of hemodynamic instability at birth

This group includes left-to-right shunt lesions such as ventricular septal defects, atrial septal defects, atrioventricular septal defects, and mild valve abnormalities. Left-to-right shunt lesions most often become hemodynamically unstable weeks after birth when the decrease in the pulmonary vascular resistance causes significant left-to-right shunting and associated pulmonary overcirculation. Similarly, infants prenatally diagnosed with a mild isolated valve abnormality and normal cardiac function are usually stable after birth. These conditions do not require specialized care in the delivery room, and babies often can be delivered at local hospitals and be evaluated either in the nursery or as an outpatient.

Congenital heart disease with minimal risk of hemodynamic instability at birth

This group mainly includes CHDs that depend on the patency of the ductus arteriosus for maintenance of the systemic or pulmonary circulation after birth. The ductus arteriosus does not generally close immediately at birth, but after 12 to 72 hours, and therefore these babies are not expected to be compromised in the delivery room or immediate perinatal period. In these cases, babies may be delivered at a location that can institute therapy with prostaglandin E1 for maintenance of ductal patency. After initial stabilization, transport of the newborn to the tertiary care cardiac center for anticipated intervention and/or surgery should be arranged.

The prediction of the need for patency of ductus arteriosus after birth may be difficult but there are several criteria available (see Table 2). In case of pulmonary outflow tract obstruction, such as critical pulmonary stenosis or atresia, severe tricuspid valve stenosis or atresia without or with a small ventricular septal defect, or severe tetralogy of Fallot (TOF), the following findings have been shown to be fairly reliable predictors of
<table>
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<td>CHD in which palliative care is planned</td>
<td>CHD with severe/fatal chromosome abnormality or multisystem disease</td>
<td>Arrange for family support/ palliative care services Normal delivery at local hospital</td>
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<td>CHD without predicted risk of hemodynamic instability in the DR of first days of life</td>
<td>VSD, AVSD, mild TOF</td>
<td>Arrange cardiology consultation or outpatient evaluation Normal delivery at local hospital</td>
<td>Routine DR care Neonatal evaluation</td>
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<td>CHD with minimal risk of hemodynamic instability in DR requiring postnatal catheterization/surgery</td>
<td>Ductal-dependent lesions, including HLHS, critical coarctation, severe AS, IAA, PA/IVS, severe TOF</td>
<td>Consider planned induction, usually near term Delivery at hospital with neonatologist and accessible cardiology consultation</td>
<td>Neonatologist in DR Routine DR care, initiate PGE if indicated Transport for catheterization/surgery</td>
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<td>CHD with likely hemodynamic instability in DR requiring immediate specialty care for stabilization</td>
<td>d-TGA with concerning atrial septum primum (it is reasonable to consider all d-TGA fetuses without an ASD at risk) Uncontrolled arrhythmias CHB with heart failure</td>
<td>Planned induction at 38–39 wk; consider CS if necessary to coordinate services Delivery at hospital that can execute rapid care, including necessary stabilizing/lifesaving procedures</td>
<td>Neonatologist and cardiac specialist in DR, including all necessary equipment Plan for intervention as indicated by diagnosis Plan for urgent transport if indicated</td>
</tr>
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<td>CHD with expected hemodynamic instability with placental separation requiring immediate catheterization/surgery in DR to improve chance of survival</td>
<td>HLHS/severely RFO or IAS d-TGA/severely RFO or IAS and abnormal DA Obstructed TAPVR Ebstein anomaly with hydrops TOF with APV and severe airway obstruction Uncontrolled arrhythmias with hydrops CHB with low ventricular rate, EFE, and/or hydrops</td>
<td>CS in cardiac facility with necessary specialists in the DR usually at 38–39 wk</td>
<td>Specialized cardiac care team in DR Plan for intervention as indicated by diagnosis; may include catheterization, surgery, or ECMO</td>
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**Abbreviations:** APV, absent pulmonary valve; AS, aortic stenosis; ASD, atrial septal defect; AVSD, atrioventricular septal defect; CHB, complete heart block; CS, cesarean section; DA, ductus arteriosus; DR, delivery room; d-TGA, d-transposition of the great arteries; ECMO, extracorporeal membrane oxygenation; EFE, endocardial fibroelastosis; HLHS, hypoplastic left heart syndrome; IAA, interrupted aortic arch; IAS, intact atrial septum; PA/IVS, pulmonary atresia/intact ventricular septum; PGE, prostaglandin; RFO, restrictive foramen ovale; TAPVR, total anomalous pulmonary venous return; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

**Source:** American Heart Association, Inc.
<table>
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<th>CHD</th>
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<td>Left-to-right atrial flow across the foramen ovale</td>
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<td>HLHS with RFO or IAS</td>
<td>Ratio of pulmonary vein forward to reversed velocity-time integral &lt;3</td>
<td>Plan for possible urgent intervention to decompress left atrium (catheterization balloon or stent; surgery)</td>
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<td>Maternal hyperoxygenation in third trimester with no change in fetal branch pulmonary artery pulsatility index</td>
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<td>d-TGA</td>
<td>Reported FO findings predictive of restriction:</td>
<td>Plan for urgent balloon atrial septostomy, on site if possible in the DR or ICU</td>
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<td>Angle of septum primum &lt;30° to the atrial septum</td>
<td>Initiation of prostaglandin E1</td>
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<td>Accelerated flow in decompressing vein</td>
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<td>Tachyarrhythmias</td>
<td>Rapid heart rate</td>
<td>Consider early delivery if appropriate gestational age</td>
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<td>Decreased heart function</td>
<td>Urgent cardioversion or medical therapy in DR if possible</td>
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<td></td>
<td>Pericardial effusion/hydrops fetalis</td>
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<tr>
<td>CHB</td>
<td>Decreasing CVP score (to &lt;7)</td>
<td>Consider early delivery</td>
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<td></td>
<td>Very low ventricular rate</td>
<td>Consider medical chronotrope or temporary pacing in DR if possible</td>
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<td></td>
<td>Decreased heart function/EFE</td>
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<td></td>
<td>Hydrops fetalis</td>
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**Abbreviations:** CHB, complete heart block; CVP, cardiovascular profile; d-TGA, transposition of the great arteries; DA, ductus arteriosus; ECMO, extracorporeal membrane oxygenation; EFE, endocardial fibroelastosis; FO, foramen ovale; HLHS, hypoplastic left heart syndrome; IAS, intact atrial septum; ICU, intensive care unit; MRI, magnetic resonance imaging; RFO, restrictive foramen ovale; TAPVR, total anomalous pulmonary venous return; TOF with APV, Tetralogy of Fallot with absent pulmonary valve.

**Source:** American Heart Association, Inc.
the necessity of patency of the ductus arteriosus or significant cyanosis and need for neonatal intervention to improve pulmonary blood flow after birth:

- Reversed flow in the ductus arteriosus, described as flow from the aorta to the pulmonary artery through the ductus (Fig. 1, Video 1)\(^{23,24}\)
- Reversed orientation of the ductus arteriosus, defined as the angle of junction between the ductus arteriosus and the aorta being less than 90°\(^{25}\)
- Pulmonary valve z score less than −3 measured after 16 weeks of gestation (in cases of TOF)\(^{26}\)

In cases of obstruction to the systemic circulation, inadequacy of the left heart to maintain systemic output after birth can be predicted based on the following:

- Reversed flow across the foramen ovale, defined as flow directed from the left atrium to the right atrium (Fig. 2)\(^{23,27}\)
- Systolic flow reversal in the distal transverse aortic arch, implying perfusion of the aortic arch via the ductus arteriosus (Fig. 3, Video 2)\(^{23,27}\)

### Congenital heart disease with high risk of hemodynamic instability at birth

This group includes cardiac defects that require immediate stabilization after birth with intervention in the immediate perinatal period. Fetuses with these conditions should be delivered at a hospital with neonatology and pediatric cardiology preferably on site, with rapid access to an interventional cardiac catheterization service and cardiac surgery. Examples of CHD in this category include hypoplastic left heart syndrome (HLHS) with a restrictive or closed foramen ovale, d-transposition of the great arteries (d-TGA), uncontrolled arrhythmias, complete heart block, TOF with absent pulmonary valve and concern for airway obstruction or with hydrops, severe Ebstein anomaly with hydrops, and obstructed total anomalous pulmonary venous return.

Fetal echocardiographic findings that have been shown to be predictive of the need for intervention to open the foramen ovale in cases of HLHS and d-TGA have been reported (see Table 2). In HLHS, the presence of the following findings increases the likelihood of need for intervention to open the atrial septum at birth:

- A ratio of forward pulmonary vein flow to reversed flow less than 3 (where the pulmonary flow is expressed as velocity-time integral) (Fig. 4)\(^5\)
- Lack of vasoreactivity in the fetal branch of the pulmonary artery during the maternal hyperoxygenation testing performed in the third trimester\(^{28}\)

**Fig. 1.** Fetus with TOF. (A) Sagittal two-dimensional imaging of the aortic/ductal arches. (B) Color Doppler shows reversed flow (red) in the ductus arteriosus. Ao, aorta; DA, ductus arteriosus; DAo, descending aorta; LA, left atrium.
In fetuses with d-TGA, specific echocardiographic features of the septum primum have been shown to have a high specificity to predict the need for atrial septostomy, although specificity is not adequate:

- Angle of septum primum less than 30° to the atrial septum
- Bowing of the septum primum into the left atrium greater than 50% (Fig. 5)
- Lack of normal swinging motion of septum primum
- Hypermobility of septum primum (Video 3)

Fig. 2. Fetus with hypoplastic left heart syndrome (HLHS). (A) Axial 4-chamber view. Color Doppler shows reversed flow (red) across the foramen ovale. (B) Pulmonary vein flow with a velocity-time integral forward/reversed flow ratio greater than 5 indicating a nonrestrictive atrial septum. The asterisk indicates the foramen ovale. f, forward flow; LV, left ventricle; r, reversed flow; RA, right atrium; RV, right ventricle.

Fig. 3. Fetus with HLHS. Color Doppler shows reversed flow (red) in the transverse aortic arch.
Fig. 4. Fetus with HLHS and intact atrial septum. (A) Axial 4-chamber view. (B) Color Doppler shows no flow across the foramen ovale. (C) Pulmonary vein flow with a velocity-time integral forward/reversed flow ratio less than 3. The asterisk indicates the foramen ovale.

Fig. 5. Fetus with d-TGA and bowing of atrial septum primum. The asterisk indicates the atrial septum.
Given that the predictive value of these findings with regard to the need for urgent atrial septostomy in d-TGA is still limited, it is reasonable to consider all fetuses with d-TGA to be at risk of hemodynamic instability at birth and it is therefore recommended that all fetuses with d-TGA be delivered at a hospital with rapid access to a pediatric cardiologist able to perform a septostomy if needed.

At present there are limited data available to guide decisions regarding delivery management of complex rare diseases that are thought to be at risk of intrauterine demise and poor neonatal outcome because of pulmonary comorbidity, heart failure, and/or compromised cardiac output, such as TOF with absent pulmonary valve complex and severe Ebstein anomaly. However, because rapid deterioration in the perinatal period can be seen with these defects, delivery in a setting that affords immediate access to specialized teams including neonatology, pediatric cardiology, and cardiothoracic surgery is probably prudent if there is any antenatal evidence of fetal compromise (effusions, hydrops, poor growth, nonreassuring fetal monitoring).

**ANTENATAL FETAL SURVEILLANCE AFTER A DIAGNOSIS OF CONGENITAL HEART DISEASE**

There may be a role for increased antenatal surveillance of a fetus with CHD. Goals of increased monitoring through the pregnancy may include evaluation for progression of the severity of the lesion in utero (ie, from non–ductal dependent to ductal dependent) and the early recognition of fetal compromise, including impairment of the fetal growth, evidence of fetal hypoxemia with altered umbilical and/or cerebral Doppler blood flow, or the development of fetal hydrops (as a manifestation of fetal congestive heart failure). Findings on serial assessment may require adjustment of the plans for perinatal and delivery management in order to improve perinatal and postnatal outcome.

**Cardiotocography and Biophysical Profile**

Computerized cardiotocography (CTG) and biophysical profile are tests used to identify fetuses at risk of intrauterine hypoxia and acidosis and they are currently used for the fetal surveillance in high-risk pregnancies with the aim of selecting fetuses at risk of poor perinatal outcome. Recommendations regarding frequency and timing have been defined for specific obstetric complications such as advanced maternal age, diabetes, hypertension, and previous stillbirth, or specific fetal conditions such as intrauterine growth restriction and multiple gestations. No guidelines are available that support this testing in fetuses with isolated CHD. Nevertheless, it is prudent to consider their use in the CHD population, especially when additional comorbidities exist or the cardiac defect puts the fetus at risk for heart failure or significant arrhythmias.

**Cardiovascular Profile Score**

The cardiovascular profile (CVP) score may be used to assess the risk of congestive heart failure in fetuses with a diagnosis of CHD. It consists of 5 parameters: edema, effusions, or overt hydrops fetalis; heart size; cardiac function; and Doppler findings of the ductus venosus and umbilical veins and umbilical artery. Any variables may be rated from 0 to 2 with a final score ranging from a maximum of 10 to a minimum of 0 (Table 3).
The use of the CVP score has been evaluated in CHD and fetal rhythm abnormalities, with a score less than or equal to 7 correlating with a higher risk of perinatal compromise or death. Among the 5 parameters, the presence of hydrops fetalis and severe cardiomegaly, defined as heart/chest area ratio greater than 0.5, have showed the most statistically significant association with mortality. The presence of hydrops or CVP less than 7 may represent an indication for urgent delivery and planning for potential immediate postnatal intervention.

DELIVERY PLANNING FOR NEONATES PRENATALLY DIAGNOSED WITH CONGENITAL HEART DISEASE

Delivery planning should take into account 3 main factors: (1) the risk of hemodynamic instability at birth, (2) the resources of the region, (3) the presence of obstetric complications.

Location of Delivery and Transportation of the Newborn

Most newborns with CHD do not need any specialized care in the perinatal period and recommendations are made to deliver at the local hospital and be followed as outpatients. However, if the presence of a specialized cardiac team after delivery is anticipated (see Table 1), the location of delivery should take into account these special needs. Determination of the site of delivery may depend on the availability of a pediatric cardiac unit in close proximity. In some regions where the number of specialized cardiac centers is limited, strategies have been developed to safely plan the delivery and perinatal management of infants prenatally diagnosed with CHD. These plans include either creation of highly specialized neonatal units locally or maternal-fetal transport with delivery nearer to a cardiac center in patients determined to be at high risk of hemodynamic instability. In order to improve the neonatal outcome of

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Abbreviations: AEDV, absent end-diastolic velocity; DV, ductus venosus; HA/CA, heart to chest area ratio; LV, left ventricle; MR, mitral valve regurgitation; MV, mitral valve; REDV, reversed end-diastolic velocity; RV, right ventricle; SF, ventricular shortening fraction; TR dP/dt, change in pressure over time of TR jet; TR, tricuspid valve regurgitation; TV, tricuspid valve; UV, umbilical vein.

the infants with CHD associated with hemodynamic instability with placental separation and avoid transportation of critically ill newborns, some freestanding children’s hospitals have planned deliveries in their facilities to accommodate these particularly high-risk patients, thus minimizing time to lifesaving intervention.11,38

**Timing of Delivery**

Recent studies have shown that infants prenatally diagnosed with critical forms of CHD tend to be delivered earlier than neonates in whom the diagnosis of CHD is made after birth.10,39,40 This finding is particularly worrisome given that otherwise healthy neonates born at 37 to 38 weeks have increased risk of worse outcomes compared with those born at later term (39–40 weeks).41 This finding has also been observed in babies with CHD. Studies have shown that babies with CHD have longer postoperative lengths of stay and higher mortality if delivered before 39 weeks.40,42,43 Therefore, in the absence of fetal or maternal indications for earlier delivery, the potential advantages of an elective delivery of fetuses with CHD early should be carefully considered.

In addition to an increased mortality, there is growing evidence that the decision about timing of delivery of infants with CHD should also consider the potential effect of the gestational age at birth on the neurologic outcome. It has been shown that fetuses with critical CHDs, such as d-TGA or single-ventricle physiology, have delays in brain maturation that manifest as alteration of brain growth, brain metabolism, and the microstructure of the white matter of the pyramidal tract44–47 and that suggest that brain development may lag by as much as 4 weeks in term neonates with CHD before heart surgery. Given these findings, it has been suggested that planning for the delivery of babies with CHD near or at full term may improve brain development and decrease susceptibility to injury postnatally16; however, further studies are needed to establish whether this will translate to improvement in long-term neurologic outcome.

Planning of the delivery of babies with CHD is also affected by the overall risk of preterm birth (occurring in 12.8% of all US births).48 To date, although there are effective predictors of preterm birth before 34 weeks, such as cervicovaginal fetal fibronectin and cervical length based on ultrasonography evaluations, there is no way to identify the exact timing of the delivery among women with risk factors or symptoms of preterm labor.49,50 For these reasons, close communication between the obstetrician and the pediatric cardiologist is necessary if spontaneous preterm birth seems likely.

**Mode of Delivery**

Data from retrospective studies show that prenatal diagnosis of a major CHD, such as HLHS, d-TGA, double-outlet right ventricle, or TOF, increases the likelihood for planned delivery and cesarean section. In fetuses with CHD, the mode of delivery has not been shown to affect the Apgar score, presurgical and postsurgical morbidity including the risk of hemodynamic instability, metabolic acidosis, and end-organ dysfunction, the length of hospitalization, or survival to surgery or discharge.51,52 Two retrospective studies have concluded that labor is safe for fetuses with CHD in most cases,53,54 but the impact on the long-term functional and neurodevelopmental outcomes is largely unknown.

**Fetal Surveillance During Labor**

The decision to perform a vaginal delivery in women with a prenatal diagnosis of fetal CHD opens a debate regarding how to monitor these fetuses during labor with the aim of promptly identifying and intervening for those at risk of hypoxemia and acidosis in
order to minimize risk of hypoxic ischemic encephalopathy and adverse long-term neurologic outcome.

The use of CTG to record the fetal heart rate and the uterine contractile activity can stratify the risk of intrapartum hypoxia and neonatal acidosis,55,56 but this test in practice has low positive predictive value for neonatal hypoxia and acidosis.57 The introduction of continuous CTG in labor has not been shown to decrease the incidence of cerebral palsy or infant death among low-risk or high-risk pregnancies but has been associated with a decrease in the incidence of neonatal seizures. However, it is also associated with a higher percentage of cesarean and instrumental vaginal deliveries.57

The fetal heart rate is determined by the interaction between the cardiovascular center in the medulla oblongata, the vagus nerve, and the heart. It has been hypothesized that fetal anomalies involving the central nervous system or the heart may alter the fetal heart rate patterns without correlating with a hypoxic or acidotic state.58 The few retrospective studies evaluating the use of CTG in labor of fetuses with CHD53,58,59 have shown that these fetuses show a higher percentage of nonreassuring fetal heart rate tracings, but no characteristic fetal heart rate patterns have been related to specific heart defects. As with normal fetuses, the use of continuous CTG in labor for fetuses with CHD has been associated with an increased rate of emergent cesarean delivery.58 In addition, CTG is limited in the assessment of the heart rate in fetuses with significant arrhythmias. To overcome these limitations, the fetal electrocardiogram, performed with scalp electrode, has been proposed in labor to monitor fetuses with a prenatal diagnosis of congenital heart block. Despite encouraging results, the only available data are based on case reports.60,61

Other types of fetal surveillance during labor, such as abdominal fetal electrocardiogram, pulse oximetry, and fetal scalp blood sampling for estimation of lactate, may serve as tools to select those fetuses with nonreassuring fetal heart rate tracings suggesting a risk of hypoxemia and acidosis; however, there are no data to support their routine use in fetuses with CHD.62 Given the lack of data, the CTG remains the main tool for routine surveillance during labor for fetuses with CHD, with the interpretation of the tracings presently based on the classification systems proposed for all pregnancies.55,56

**SUMMARY**

The prenatal diagnosis of severe CHD is associated with improvement in preoperative condition, with a reduction in morbidity, including neonatal hypoxemia, need for invasive respiratory support, and metabolic acidosis, and an increased survival in select defects, including HLHS and d-TGA. Furthermore, detection of CHD in utero allows better parental counseling and delivery planning, especially when the need for urgent postnatal intervention is anticipated based on available predictive models. Perinatal management should be tailored to the specific needs of the mother and fetus, and should include decisions regarding location, timing, and mode of delivery that in general minimize the risk of early or operative delivery. In selected cases, there may be compelling maternal or fetal indications for earlier delivery, including a variety of obstetric indications, such as spontaneous onset of labor, maternal comorbidities, pregnancy complications, or nonreassuring results of fetal testing. Collaboration between obstetric and pediatric specialty services and careful attention to perinatal management and delivery planning after a prenatal diagnosis of CHD is made can improve the perinatal status of newborns with potential for improvement in both survival and long-term functional and neurodevelopmental outcome.
**Best practices**

**What is the current practice?**

CHD recognized before delivery

Objective: to minimize perinatal morbidity and mortality for both infant and mother, optimize infant status before surgical intervention, and improve short-term and long-term outcomes

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

Prenatal diagnosis has the potential to confer improved survival and long-term outcome, specifically a neurodevelopmental advantage, if certain modifiable risk factors can be addressed during delivery planning

Where: triage of delivery to appropriate facility/level of care based on prediction of newborn hemodynamic status at birth

When: delivery as close to term as possible with avoidance of iatrogenic late preterm and early term delivery

How: avoidance of routine early induction of labor and of operative delivery in cases lacking clear obstetric indication

**Major recommendations**

- Perinatal management should emphasize maternal-fetal health and both short-term and long-term outcomes
- Most CHDs, such as shunt lesions or mild valve abnormality, with normal cardiac function may be delivered without anticipation of specialized delivery room care
- Delivery planning in a specialized center may be beneficial for certain high-risk lesions
- Vaginal delivery after spontaneous-onset labor is probably best for mother and baby in most cases; iatrogenic late-term and early term delivery should be discouraged
- Cardiotocography should be considered as a method of surveillance in labor for fetuses with heart malformations and the interpretation of the tracing is based on the same classification systems proposed for normal fetuses

**SUPPLEMENTARY DATA**

Supplementary data related to this article can be found at [http://dx.doi.org/10.1016/j.clp.2015.11.004](http://dx.doi.org/10.1016/j.clp.2015.11.004).

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