Curr Treat Options Cardio Med (2017) 19: 5
DOI 10.1007/s11936-017-0502-1

Prenatal Counseling of Fetal Congenital Heart Disease

Caroline K. Lee, MD

Address
Pediatric Cardiology, Washington University School of Medicine, One Children's Place, NWT Box 8116, St. Louis, MO, 63110, USA
Email: lee_c@kids.wustl.edu

Published online: 14 February 2017

This article is part of the Topical Collection on *Pediatric Congenital Heart Disease*

Keywords Fetal cardiology · Fetal echocardiography · Congenital heart disease · Prenatal counseling

Opinion statement

The field of fetal cardiology has advanced greatly over the last two decades and congenital heart defects can now be identified in utero with a high level of accuracy. Prenatal counseling of parents given the news of a fetal cardiac defect is an important role of the fetal cardiologist. Prenatal counseling is a complex task that requires skill to perform and interpret fetal echocardiograms, an understanding of fetal and postnatal cardiovascular physiology, knowledge of therapeutic and surgical options, and of long-term outcomes including quality of life. Just as important is the manner in which the information is conveyed and the support offered to the parents as these affect parental understanding, influence decision-making, and may impact the parents' emotional and psychological coping.

Introduction

Congenital heart defects (CHDs) are the most common fetal structural malformation and are the leading cause of neonatal mortality among birth defects. The estimated incidence of moderate and severe forms of CHD is 6 per 1000 live births [1]. With advances in the fields of fetal cardiology and ultrasound technology over the last two decades, most heart defects can now be well-defined by fetal echocardiography in the second trimester.

The fetal cardiologist, a pediatric cardiologist specializing in fetal medicine, plays a critical role in not only making an accurate diagnosis but also providing prenatal counseling to help parents understand the diagnosis, guide and support them in decision-making, and provide them with information about the implications of their child's heart disease, both medically and on the expected quality of life (QOL). This article will review the

benefits and aims of prenatal counseling, the issues encountered and topics to be covered, and considerations on providing family support during the prenatal period.

Prenatal detection rates

Despite the widespread availability and routine use of prenatal ultrasound, the current prenatal detection rate (PDR) of CHDs is suboptimal. Contemporary studies report PDRs of significant CHD ranging from 25 to 53% in North America and Europe [2–5]. Single ventricle heart defects, wherein one ventricle is hypoplastic, have the highest rate of prenatal detection [3]. In the largest study to date on PDRs, of the 31,374 infants in the Society of Thoracic Surgeons database undergoing congenital heart surgery at 6 months of age or younger during 2006–2012, only 34% were diagnosed prenatally [6]. There were higher rates of detection for lesions identifiable on four-chamber fetal ultrasound view than for those that require visualization of outflow tracts [6]. Studies have demonstrated improved prenatal detection when evaluation of outflow tracts is included [4, 7].

In 2013, the American Institute of Ultrasound in Medicine published revised guidelines for the performance of obstetrical ultrasound examinations [8]. In addition to the prior standard of evaluating the fetal heart with the four-chamber view, the addition of right and left ventricular outflow tracts was recommended. This important new recommendation should increase the PDR, as critical cyanotic lesions such as tetralogy of Fallot (TOF) and d-transposition of the great arteries (dTGA) may be missed by four-chamber view alone. However, many obstetric providers and sonographers have not been trained on how to obtain or interpret these outflow tract views, and so, there remains an important knowledge gap that must be addressed by the obstetric and fetal cardiology communities [7, 9, 10]. As most CHDs occur in low-risk pregnancies with no known risk factors, effective obstetric screening is critical [11–13].

Benefits of prenatal diagnosis

There is lack of consensus in the literature as to whether a prenatal diagnosis of CHD is associated with improved surgical outcomes and less neonatal mortality, with some studies reporting a positive impact on outcomes, and others showing no difference in outcomes compared to babies diagnosed with CHD after birth [14–31]. Confounding the evaluation of surgical outcomes is that fetuses with severe CHDs are more likely to be detected prenatally and they are generally more high risk; elective pregnancy termination or comfort care may be chosen by families with a prenatal diagnosis; and there may be incomplete ascertainment of postnatally diagnosed CHD cases if they died prior to transfer to a tertiary care center or before undergoing surgery. A recent meta-analysis evaluating the differences in pre-operative mortality rates between newborns with and without a prenatal diagnosis found that prenatal diagnosis of critical CHD improves neonatal pre-operative survival, and newborns with a postnatal diagnosis were more likely to die of cardiovascular compromise prior to planned cardiac surgery [32].

There are many benefits to the parents and newborn with a prenatal diagnosis of CHD. It allows opportunities to counsel the parents about the diagnosis and

increases parental understanding of their child's CHD [33]. It allows time for parental decision-making regarding continuation or termination of pregnancy, prenatal genetic testing, site of delivery and postnatal cardiac care, and discussion about the option of palliative care if the CHD is severe and the prognosis poor. If indicated, delivery can be planned at a tertiary care facility where pediatric cardiology and surgical services are available, thus avoiding a transfer and potential long-distance separation of the mother and baby. For ductal-dependent lesions, availability of prostaglandins can be ensured so that profound cyanosis or shock can be avoided.

Having a prenatal diagnosis allows time for parents to process the emotions of shock, grief, and disappointment and allows them to be mentally and emotionally prepared for the postnatal hospitalization and medical needs of their baby. It allows them time to prepare for their family's needs, such as arranging parental leave from work, relocating if necessary to be near a cardiac center, and making plans regarding care for their other children.

Referral for fetal echocardiogram and cardiology consultation

When there is a suspected CHD on screening fetal ultrasound, there should be minimal time delay in referring the mother for a fetal echocardiogram and cardiology consultation. Women and their partners value a short waiting time from the first suspicion of fetal heart disease to the cardiac evaluation and confirmation of diagnosis [34]. Delays in referral for further evaluation increase parental distress [35] and may preclude the option of pregnancy termination. The fetal cardiologist should be flexible and willing to accommodate scheduling these consultations as soon as feasible.

Pregnant women presenting for fetal echocardiograms report high anxiety levels [36]. It is therefore ideal if the consulting physician meets with the patient prior to the fetal echocardiogram being performed, as patient anxiety may be allayed by explaining to the patient what to expect for the echocardiogram and consultation. This discussion should include a review of the mother's understanding of the indication for the fetal echocardiogram; a review of the patient's past medical, family, and social histories; and a review of the pregnancy to date including whether other fetal anomalies have been suspected and if genetic screening or testing has been performed. This allows the entirety of the discussion after the scanning to be focused on the fetal cardiac findings.

The patient should be told that the duration of the scanning may be long, that there may be long silences during the study, and that a full explanation of the findings will occur after the scan is completed. Any trainees present during the scan should be properly introduced, and advised to refrain from making comments or asking questions during the scan, as this can be distracting and anxiety-provoking to the mother.

Prenatal counseling

Once the fetal echocardiogram is complete, counseling should occur shortly following. In addition to the physician, a fetal nurse coordinator is a valuable addition to the counseling session, as they can take notes for the family, offer

support, and be an important ongoing resource for follow-up with the family [37].

There is little research on performing prenatal counseling for CHD or determining the most effective strategies for providing family support. While counseling styles vary by providers, it is important that counselors have good communication skills, show empathy, and be perceptive in assessing how the information is being received. The counselor must assess parental understanding and emotional status throughout the discussion. Particularly at the first visit, the initial shock and grief reactions to an abnormal finding may inhibit the parents' ability to retain information. The initial consultation is particularly challenging, as it is necessary to explain potentially complex anatomical and medical information to a family when stress levels are very high. Ideally, there will be an opportunity for follow-up consultation(s) in order to adequately complete the counseling and reinforce points that may have been forgotten in the midst of an emotional first meeting. Parents report that oral information needs to be repeated several times at different occasions [34]. Indeed, an important benefit of early prenatal diagnosis is that it allows time during the remainder of the pregnancy for multiple opportunities to review information with the parents.

Elements of counseling

A checklist of information to be covered during fetal consultations may be helpful to ensure that all important topics are addressed (see Table 1).

The diagnosis

Once confirmation of CHD has been made, discussion of the diagnosis with the parents is significantly aided by the use of diagrams comparing normal cardiac anatomy with a depiction of the specific fetal cardiac findings. This helps the patient have a visual understanding of the differences from normal. Other written materials and surgical diagrams are helpful to provide as the parents can refer back to them after the visit [35]. Having a fetal nurse coordinator write down the diagnoses, surgical procedure names, and other notes allows the parents to better focus on the discussion without worrying that information may be forgotten.

Parents should be made aware of any limitations of the study and type and degree of uncertainty.

A common question from expectant parents who are given a diagnosis of a fetal anomaly is "How did this defect occur?" There are several known maternal, familial, and fetal risk factors for the development of CHDs. These include maternal pregestational diabetes, maternal phenylketonuria, medication exposure (e.g., retinoic acid, angiotensin-converting enzyme inhibitors), CHD in a first degree relative of the fetus, inherited syndromes associated with CHD, increased fetal nuchal translucency, assisted reproductive technology, monochorionic twinning, known or suspected chromosomal abnormality, and extracardiac anomalies such as omphalocele or duodenal atresia [38]. In most cases, however, CHDs occur in low-risk pregnancies and the etiology is unknown. Reassurance should be provided to alleviate the parental guilt that is commonly felt when a malformation is detected prenatally.

Table 1. Elements of prenatal counseling of congenital heart disease

□ ***Diagnosis***: Provide description of diagnosis and diagram depicting fetal echocardiogram findings compared with normal heart anatomy; discuss the important physiologic alterations from normal
□ ***Diagnostic limitations and uncertainties***: May be affected by gestational age, fetal position, maternal habitus
□ ***Known causes or risk factors for CHD***: Discuss if risk factors are present and provide reassurance to allay any parental guilt that CHD is their fault
□ ***Potential for progression of disease or fetal demise***: Discuss what may evolve and the anticipated follow-up for remainder of gestation
□ ***Extracardiac anomalies***: In collaboration with maternal fetal medicine, it is important to assess for other anomalies and discuss the possible effect on postnatal management and prognosis
□ ***Genetic associations and testing***: Discuss possible genetic syndromes associated with the CHD; review prenatal screening results and limitations; discuss consideration of amniocentesis, and if declined, the postnatal genetic testing to be performed
□ ***Increased risks to CHD outcome***: May occur with associated prematurity, growth restriction, extracardiac anomalies, heterotaxy syndrome, genetic syndrome
□ ***Pregnancy options***: Discuss if parents are considering continuation or termination of pregnancy
□ ***Fetal cardiac intervention options***: Discuss if applicable
□ ***Delivery planning***: Timing, induction of delivery, site of delivery, site of postnatal cardiac care
□ ***Expected postnatal management***: ICU admission, intravenous lines, prostaglandins (PGE1), diagnostic testing (e.g., echocardiogram, angiography, CT, MRI)
□ ***Expected neonatal cardiac surgery or catheterization procedure***: Nature of procedure(s), survival rates, potential complications
□ ***Expected hospital course and length of stay***: Mechanical ventilation and vasoactive medication support, sedation, feeding issues (e.g., NPO, need for nasogastric feeding tube, pumping breast milk), medications, therapy involvement (physical, occupational, speech)
□ ***What to expect at hospital discharge***: Medications, possible need for feeding tube, possible home monitoring of weight and oxygen saturations, home nursing visits, outpatient therapies, cardiology follow-up
□ ***Long-term issues***: Anticipated or possible surgical or catheterization interventions needed in the future, physical limitations, neurodevelopmental issues, cardiology follow-up into adulthood, possible development of heart failure and need for heart transplant
□ ***Family issues***: Planning for parental leave from work, housing needs if from out of town, financial considerations and insurance issues, psychological support, sibling support
□ ***Other prenatal consultations***: Cardiothoracic surgery, neonatology, genetics, palliative care, social work, etc.
□ ***Provide tours*** of labor & delivery, newborn and/or cardiac intensive care units
□ ***Offer resources for parents***: Provide written materials on diagnosis and cardiac operations, websites for CHD information, information on support groups; offer to connect parents with other CHD families with a similar diagnosis; provide contact information for fetal nurse coordinator and fetal cardiology physician
□ Prior to delivery, parents should ***identify a primary care pediatrician*** comfortable following a baby with CHD

The possibility of disease progression during the remainder of gestation should be discussed. Semilunar valve stenosis may progress in severity and evolve into functional atresia. Ventricular hypoplasia, hypertrophy, and dysfunction may result from outflow tract obstruction. Atrioventricular valve regurgitation such as tricuspid regurgitation in Ebstein's anomaly may progress, resulting in progressive cardiomegaly, hydrops, and intrauterine death [39].

The risk of intrauterine death is low in fetuses with CHD who are in sinus rhythm with good myocardial function and no or trivial atrioventricular valve regurgitation. In addition to severe atrioventricular valve regurgitation, other risk factors for intrauterine death are concomitant

bradycardia or tachycardia in the setting of CHD or the presence of extracardiac or chromosomal abnormalities [37, 40].

Associated anomalies

The possible association of genetic, chromosomal, or syndromic anomalies should be discussed and how these may affect management and outcomes of the cardiac disease. Extracardiac anomalies not uncommonly coexist with CHD [13]. In a recent cohort of over 15,000 neonates (<30 days) in the Society of Thoracic Surgeons Congenital Heart Surgery Database undergoing cardiac surgery, nearly 19% had associated extracardiac abnormalities, genetic abnormalities, or syndromes [41].

There are known associations of specific heart defects with genetic syndromes. The finding of a conotruncal defect such as TOF, truncus arteriosus, or interrupted aortic arch should prompt a discussion of the possible association with chromosome 22q11 microdeletion syndrome. In a fetus with suspected trisomy 21, there is a 40–60% chance of associated CHD, most commonly a complete atrioventricular canal defect (CAVC) [42]. Conversely, 80% of patients with CAVC have trisomy 21 [43, 44]. In a female fetus, left heart obstructive lesions such as aortic stenosis, coarctation of the aorta, or hypoplastic left heart syndrome (HLHS) may be associated with Turner syndrome (monosomy X). dTGA has the lowest prevalence of associated abnormalities [41].

Since its introduction in 2011, non-invasive cell-free fetal DNA screening in high-risk pregnancies, such as those with suspected CHD, has become widespread in practice. It is important to remember, however, that these are screening tests that assess risk for fetal aneuploidy (trisomies 13, 18, and 21) and they do not have the diagnostic accuracy of amniocentesis. When the indication for amniocentesis is presence of a fetal structural abnormality, chromosomal microarray testing (which identifies duplicated or deleted chromosomal segments) is recommended; targeted testing may be performed based on the heart defect (e.g., FISH for suspected chromosome 22q11 microdeletion or karyotype for suspected trisomy 21) [45]. Therefore, counseling should include discussion of consideration of amniocentesis, as this knowledge may help determine prognosis and inform postnatal care. The finding of an associated chromosomal abnormality may also strongly influence decisions about pregnancy termination [34, 46].

Parental decision-making

Parents given the news that their baby has a CHD must make critical decisions including whether or not to continue pregnancy. Often there is only a short time available to make the decision regarding pregnancy termination, and this time constraint adds to parental stress [46]. The counselor must know the time constraints on termination relevant to their state or region. Amniocentesis and additional high-level obstetric ultrasound imaging may need to be arranged expeditiously to evaluate for other genetic or extracardiac abnormalities.

Counselors should appreciate the emotional turmoil parents are feeling, provide families with information to come to a decision that is best for them,

and provide support regardless of the choice parents make [47]. Parents and cardiologists generally agreed that QOL of the child was the most important factor that would affect their decision about whether or not to continue pregnancy, but compared with cardiologists, parents were more likely to select religious/moral beliefs as an important factor in their decision-making [48]. Other factors influencing this parental decision include severity of CHD, gestational age at diagnosis, and chromosomal abnormalities [46, 49].

If the decision concerning pregnancy continuation is compromised by an incomplete or suboptimal study, or an uncertain diagnosis, then the fetal echocardiogram should be repeated within a short time period or the patient referred to a more experienced fetal cardiologist [47, 50]. Delays in further evaluation and defining a diagnosis should be avoided, as they increase parental anxiety and may preclude the option of pregnancy termination.

Delivery planning

Delivery planning is an important component of the counseling. Fetal cardiologists and maternal fetal medicine specialists work collaboratively as caregivers to the mother and fetus with heart disease. Delivery at a tertiary care facility with access to pediatric cardiac care is recommended for ductal-dependent lesions and any heart defect that is expected to require neonatal intervention. For cardiac defects such as CAVC or TOF with mild pulmonary stenosis, delivery at a local hospital is possible, depending on the nursery's level of comfort managing a baby with CHD. Counseling should be given in close cooperation with the obstetrical team to provide consistent information to the parents and optimal care for the fetus.

Delivery of CHD babies at later gestation has been shown to be associated with improved survival [51]. High morbidity and mortality have been reported in CHD babies born before 36 weeks, particularly in those with extracardiac and genetic abnormalities [52]. Even delivery at an early term (37–38 weeks gestation) is associated with worse morbidity (complications, length of stay) and mortality after neonatal cardiac surgery than delivery at 39 weeks gestation [53, 54]. For this reason, delivery after 39 weeks is typically recommended.

Prenatal diagnosis of CHD is associated with a threefold risk of developing intrauterine growth restriction (IUGR) [55], and therefore, monitoring fetal growth is important. Parents should be counseled that IUGR is associated with increased morbidity after cardiac surgery [56]. Similarly, lower weight (<2.5 kg) is associated with higher mortality after cardiac surgery [57].

Mode of delivery is not typically altered in the setting of fetal CHD, and high rates of vaginal delivery can be achieved [58]. There are rare circumstances when the fetus may benefit from exact timing of delivery with a Cesarean section, when immediate surgical or cardiac catheterization intervention after birth is anticipated for a critically ill infant. An example is if an atrial septostomy is required urgently after birth for a restrictive or intact atrial septum in a fetus with HLHS or dTGA. Most critical heart defects, however, can be managed with prostaglandin infusion for days

before cardiac intervention, and so, precise timing of delivery is not needed. Many of the more common CHDs—shunt lesions and mild valve abnormalities—may be delivered without anticipated specialized delivery room care [59].

Anticipated neonatal course and interventions

The range of possible treatment options for the heart defect should be discussed. In rare instances, fetal cardiac intervention may be considered as in severe aortic stenosis with evolving HLHS or HLHS with restrictive atrial septum [60–62]. This type of care is available only at a few specialized centers.

In most instances, intervention for the heart defect will occur after birth. This can include medical, surgical, or cardiac catheterization interventions. Prostaglandin is made available for ductal-dependent lesions—those that require patency of the ductus arteriosus for either pulmonary blood flow (e.g., severe TOF or pulmonary atresia) or systemic blood flow (e.g., HLHS, interrupted aortic arch).

Parents should be counseled on what to expect for delivery room management, as it differs from a normal delivery experience. Various teams and personnel may be present in the delivery room. Parents should be told whether they will be able to hold their new baby and how much time they may have before the infant is transferred from the delivery room. Discussion should include which unit the baby is expected to be admitted, whether nursery (for defects without anticipated significant hemodynamic or clinical effects) or an intensive care unit. Offering parents a tour of these units allows them an opportunity to visualize where their care and their infant's care will take place and may help lessen anxiety about what to expect [63].

Parents should know that modifications to the fetal diagnosis may be made after birth and that surgical plans may change depending on postnatal findings. Nevertheless, the anticipated type of cardiac intervention should be discussed as well as expected hospital course and length of stay. Parents should be made aware of the invasive lines and tubes to expect (e.g., central venous lines, endotracheal tube, nasogastric tube), as the sight of these in their infant may be upsetting. Feeding issues should be discussed, including whether the infant will be allowed to feed pre-operatively, and the mother should be counseled that at least initially, she may have to pump breast milk instead of directly breastfeeding.

The expected course for follow-up after hospital discharge may be touched upon, including medications, the possibility of going home with a feeding tube, and what follow-up care (with the cardiologist, home nursing, and therapies) is anticipated. Helping the parents understand what they might expect when going home will help them prepare for childcare and work decisions.

After the initial consultation, follow-up visits may include consultations with a pediatric cardiac surgeon, neonatologist, geneticist, or social worker.

Outcomes

An honest appraisal of outcomes in general for the diagnosis and procedures, and specific to the provider's cardiac center, should be discussed as much as is

known. While early mortality rates are often the focus of prenatal counseling, longer term issues about need for reoperations or other cardiac interventions, and QOL, are important to discuss as well. While cardiologists may be conscious of not wanting to provide too much information and overwhelm parents, parents report they prefer to receive more information than cardiologists provide during the prenatal period [48, 64].

Approximately 85% of babies born with CHD are expected to reach adulthood [65]. While outcomes vary significantly depending on the type of heart defect, in general, long-term survival (>20 years) rates are estimated to be 95% for simple CHD (e.g., atrial and ventricular septal defects, isolated semilunar valve disease), 90% for moderate severity CHD (e.g., TOF, coarctation of the aorta, atrioventricular canal, Ebstein's anomaly), and 80% for complex CHD (e.g., single ventricle, dTGA, truncus arteriosus) [65]. As long-term outcomes are evolving, particularly for more complex heart defects such as single ventricle lesions, it is important that the fetal consultant be aware of the most contemporary outcomes data.

Single ventricle heart defects are often detected prenatally because severe hypoplasia of a ventricle is easily seen on a four-chamber view of the heart. The Fontan procedure is the final common pathway for all functional single ventricles, including HLHS, tricuspid atresia, pulmonary atresia with intact ventricular septum, double-inlet left ventricle, and unbalanced common atrioventricular canal. A recent follow-up of over 1000 Fontan patients in Australia and New Zealand showed 97% survival at 10 years for lateral tunnel and extracardiac conduit-type Fontans [66].

Despite this good survival, many Fontan patients experience long-term complications, including dysrhythmias, need for pacemaker, thromboembolic events, and failure of the Fontan circulation including protein-losing enteropathy and plastic bronchitis. The possible need for heart transplantation in the future may be discussed, depending on the CHD.

As survival for CHD has improved, there is increased recognition of the risk of neurodevelopmental delay in these children, particularly those with more severe defects [67]. Brain abnormalities and developmental delay were previously thought to be related to cardiac surgery; however, recent studies have demonstrated abnormal brain development and delayed brain maturation in fetuses with isolated CHD and newborns before CHD surgery [68–71]. Importantly, correlations of fetal brain findings with neurodevelopmental outcome have been inconclusive and more long-term data are needed. Because neurodevelopment is a significant factor affecting QOL for both the child and the family, prenatal counseling on this aspect of CHD may affect pregnancy decisions, and thus, the fetal counselor must be knowledgeable about this issue [72]. Parents should be informed that the American Heart Association recommends developmental surveillance, screening, and evaluation of children with CHD beginning in infancy [67].

Psychological stress

Receiving news of a CHD in their fetus is a stressful event for parents [73–75]. Mothers who have been given a prenatal diagnosis of CHD exhibit

posttraumatic stress, depression, and anxiety, and report lower partner satisfaction than normal pregnant women [73]. Parents experience grief at the loss of a "normal" pregnancy and imagined healthy child. As maternal depression and stress during pregnancy have been negatively associated with fetal somatic growth and neurocognitive development [76], it is important that attention be given to maternal psychological well-being. Psychological support and counseling should be offered and encouraged both during and after pregnancy [73–75].

Resources, support, and follow-up

It is expected that parents will seek further information and support on the internet [34, 77]. They should be cautioned about the difficulty in discerning accurate information on the internet, and that conflicting information may be confusing and discouraging. Providing a list of websites pre-approved and reviewed by the fetal cardiologist helps to guide families to the most accurate information relevant to their particular situation.

Parents report that they use the internet not only to seek medical information but also to learn from other parents of children with CHD about daily life, schooling, and QOL issues [34]. Parents have described the need for and importance of support from other parents who have gone through a similar situation [34, 64]. Blogs and social media are means by which parents connect and gain insight from others who have faced similar diagnoses and circumstances. Parents should be given information on support groups and offered to be placed in contact with other parents who have a child with a similar diagnosis. Connecting parents in these ways can reduce feelings of isolation, provide them with reassurance and hope, and help prepare them for what life may look like after the neonatal period.

The fetal nurse coordinator's role becomes increasingly important as pregnancy progresses. The nurse coordinator is available to resolve follow-up questions that arise after the visit and provide ongoing support and education. The nurse should be knowledgeable and experienced to be able to answer parents' questions about surgical and postnatal care issues and long-term issues as well.

Conclusion

"Counselling the parents following a diagnosis of fetal congenital heart disease is as important a task for the fetal cardiologist, as the skill involved in achieving an accurate diagnosis [50]." Prenatal counseling involves providing an accurate diagnosis of the CHD; clearly explaining complex medical information to parents who are in crisis, including management options and outcomes; preparing the parents for the delivery and course after birth; and supporting them through the process of decision-making and throughout the pregnancy. Improved prenatal detection of CHD is needed in order to provide parents this important opportunity for counseling and education and to optimize outcomes in infants with CHD.

Compliance with Ethical Standards

Conflict of Interest
Caroline K. Lee declares no potential conflicts of interest.

Human and Animal Rights and Informed Consent
This article does not contain any studies with human or animal subjects performed by any of the authors.

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