Fetal Echocardiography

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ABSTRACT

Congenital heart disease is the most common birth defect. Although most of the affected newborns do well, babies with severe restrictions of systemic or pulmonary blood flow are at risk for decompensation from cyanosis and acidosis, and usually require early cardiac interventions. Some babies have sustained arrhythmias in utero that can alter cardiac output and lead to fetal hydrops. All these complex issues can be assessed by targeted fetal echocardiography, allowing families to be advised about the implications of their child's cardiac abnormality, its potential clinical course, and the options for management. When there is a likely need for urgent stabilization after birth, a prenatal diagnosis allows planning for a delivery in circumstances that permit rapid postnatal evaluation and management, thus minimizing the risk of hemodynamic decompensation and end-organ damage in a fragile neonate.

This article discusses the general scope of fetal echocardiography and perinatal cardiology.

INTRODUCTION

As the most common congenital defect, congenital heart disease (CHD) affects 8 of every 1,000 live neonates. Fetal echocardiography has become an important component of perinatal evaluation for CHD over the past two decades. Cardiac structure, function, and rhythm can be assessed, with reasonably accurate results in the hands of experts in this aspect of obstetric care. Families can be counseled about their unborn child's potential outcome, and both prenatal and perinatal management strategies can be implemented. The ultimate goal is to optimize postnatal outcomes for children with serious cardiac abnormalities.

THE FETAL CARDIOVASCULAR SYSTEM

Since the placenta is the site of fetal oxygenation and ventilation, the right ventricle is the dominant ventricle, pumping approximately 55-60% of the combined cardiac output.[1] Fig. 1 provides a review of the fetal circulation, in which oxygenated blood from the placenta is transported via the umbilical vein, through the ductus venosus into the inferior vena cava and right atrium, across the patent foramen ovale (PFO) into the left atrium and left ventricle, preferentially perfusing the coronary arteries and the cerebral vasculature. Concurrently, deoxygenated blood from the superior vena cava enters the right heart and main pulmonary artery. The elevated pulmonary vascular resistance in the unexpanded fetal lungs preferentially shunts deoxygenated blood across the patent ductus arteriosus (PDA), and towards the low resistance placenta.

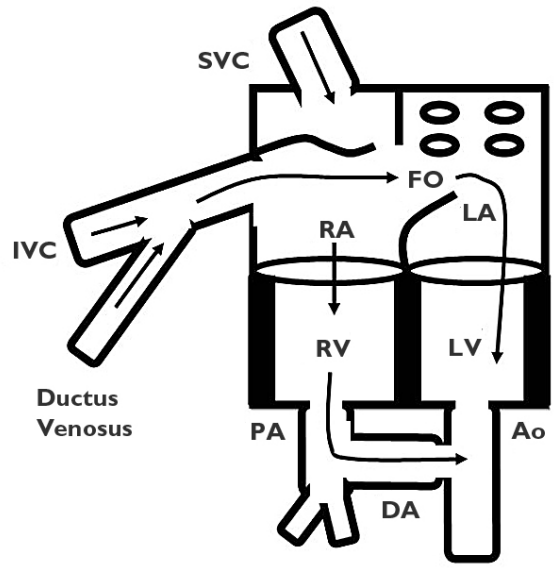

Fig. 1. The Fetal Circulation: The ductus venosus carries the most highly oxygenated blood sequentially across the foramen ovale (FO) to the left atrium (LA), the left ventricle (LV), and then out of the aorta (Ao) to supply the coronary arteries and brain. The superior vena cava directs the most deoxygenated blood across the tricuspid valve into the right ventricle (RV) and then out the pulmonary artery and ductus arteriosus to the lower half of the body and the placenta. (With permission from Gleason M, ed. Pediatric Practice: Cardiology. New York: McGraw-Hill Medical; 2012. Chapter 3. Fetal Echocardiography.)

It is important to note that the fetal myocardium differs from the adult myocardium; relaxation is impaired, so ventricular filling is accomplished predominantly by active atrial contraction rather than passive ventricular filling. Disease processes that cause

Table 1. Indications for Fetal Echocardiography
Maternal Indications:
■ Family history of congenital cardiac disease
■ Metabolic disorders, such as diabetes
■ Exposure to teratogens
■ Infection with rubella or other viruses
■ Autoimmune disease, such as systemic lupus erythematosus or Sjögren's syndrome
■ Familial inherited disorders, such as Marfan's, DiGeorge, or Noonan's syndrome
■ Artificial fertilization
Fetal Indications:
■ Abnormal obstetrical ultrasound screening
■ Two-vessel umbilical cord
■ Extracardiac abnormality
■ Chromosomal abnormality (known or suspected)
■ Arrhythmia
■ Hydrops fetalis
■ Increased nuchal translucency in first trimester (greater than 3 mm between 10-14 weeks gestation[3])
■ Multiple gestation and twin-twin transfusion syndrome

increased atrial pressure or affect atrial emptying can lead to *hydrops fetalis* (fluid in any two extra-vascular spaces, such as ascites, peripheral or scalp edema, pleural and pericardial effusions). Yet, despite severe cardiac structural deformities, if the PDA is non-restrictive in utero and placental function is good, most gestations go to full term.

SCREENING WITH FETAL ECHOCARDIOGRAPHY

Echocardiographic detection of fetal congenital heart defects has improved with increased experience. Targeted fetal echocardiograms are ideally performed between 18 and 24 weeks gestation, when there is adequate amniotic fluid for good visualization of the cardiac structures and vasculature. After 30 weeks of gestation the increase in fetal body mass and bony shadowing can make image acquisition challenging.

In high-risk pregnancies, including fetal aneuploidy (an abnormal number of chromosomes), early transabdominal and transvaginal fetal echocardiography may be attempted as early as 11 to 14 weeks of gestation. Current maternal and fetal indications for fetal echocardiography, as recommended by the American Society of Fetal Echocardiography, are outlined in Table 1.[2]

Some maternal medical conditions or exposure to specific medications are associated with the development of specific congenital cardiac abnormalities. (Table 2)

COMPONENTS OF THE FETAL ECHOCARDIOGRAM

Comprehensive fetal echocardiography includes two-dimensional imaging for anatomy, as well as color and pulsed Doppler interrogation of blood flow for fetal cardiac physiology.

Two-dimensional cardiac imaging:

Thorough anatomic assessment should include an apical 4-chamber view of the heart, an apical 5-chamber view, views of the left ventricular and right ventricular outflow tracts, a view to show orientation of the great arteries, visualization of the systemic and pulmonary venous returns, a view of the ductal arch, and a

Table 2. Maternal Medical Conditions and Medication Exposures, and Associated Congenital Heart Lesions	
Maternal Condition / Medication Exposure	**CHD Lesions**
■ Diabetes	■ Conotruncal defects, VSD, hypertrophic cardiomyopathy
■ Phenyketonuria (PKU)	■ TOF, VSD, PDA, LVOTO
■ Autoimmune-Lupus	■ Complete heart block
■ Rubella Infection	■ PS, PDA
■ Lithium	■ Ebstein's Anomaly
■ Phenytoin, Hydantoin	■ VSD, PS
■ Retinoic Acid	■ Severe and complex CHD, Conotruncal defects

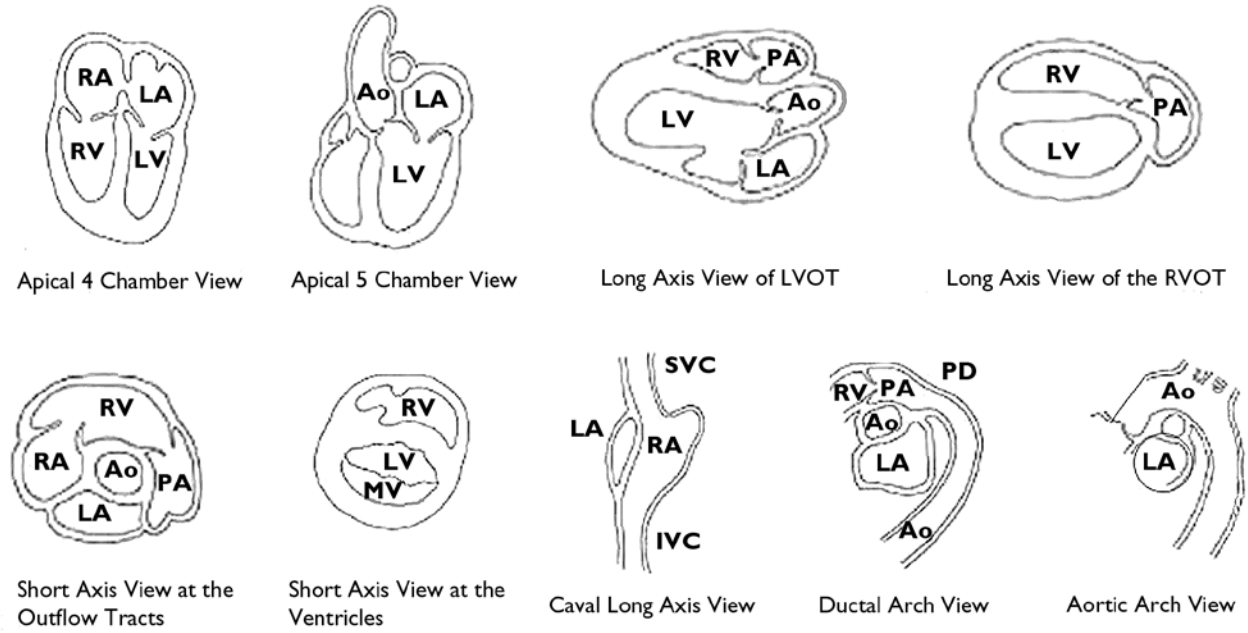

Fig. 2. The Fetal Imaging Views: (RA) right atrium, (LA) left atrium, (RV) right ventricle, (LV) left ventricle, (Ao) aorta, (PA) pulmonary artery, (MV) mitral valve, (SVC) superior vena cava, (IVC) inferior vena cava, (PDA) patent ductus arteriosus. (With permission from Gleason M, ed. Pediatric Practice: Cardiology. New York: McGraw-Hill Medical; 2012. Chapter 3. Fetal Echocardiography)

view of the aortic arch (Fig. 2). The fetal heart rate is documented, and arrhythmias are assessed by pulsed Doppler and M-mode imaging. Cardiac dysfunction may be implied if there are ascites, pleural or pericardial effusions, skin edema, or cardiomegaly [ratio of cardiac to thoracic areas of greater than 0.36[4]].

Doppler Flow Evaluation:

Color Doppler imaging allows rapid qualitative assessment of the direction and velocity of blood flow through the cardiovascular system. Abnormal color Doppler patterns help identify valve stenosis or regurgitation and septal defects. Mild tricuspid regurgitation may be benign,[5,6] but mitral, pulmonic, or aortic valve leakage usually suggests structural or functional heart disease. The presence of left to right shunting at either the atrial or ductal levels, instead of the usual right to left pattern, is always abnormal and suggests cardiac disease. Left-to-right flow at the PFO may indicate inadequacy of the left heart or aortic arch, whereas reversed flow through the ductus suggests right heart obstruction.[7]

Pulsed Doppler echocardiography assesses the direction and velocity of blood flow quantitatively. Expected Doppler flow patterns for the umbilical artery, umbilical vein, middle cerebral artery, ductus venosus, atrioventricular valve inflows, and ductus arteriosus, have been documented for each gestational age. Doppler echocardiography may help quantify the degree of cardiac compromise in fetuses with altered hemodynamics secondary to congenital heart disease, intrauterine growth retardation, twin-to-twin transfusion syndrome, or extra-cardiac anomalies that are known to impact the fetal cardiovascular system.

Since the umbilical artery connects to the low resistance placenta, its flow is normally continuous in systole and diastole. Diminished diastolic flow in the umbilical artery can be seen with intrauterine growth retardation and/or placental insufficiency. Diastolic flow reversal in the umbilical artery indicates a high risk for in utero demise. (Fig. 3) Doppler assessment of the umbilical vein normally shows continuous flow. If fetal central venous pressure rises, notching of the umbilical vein Doppler pattern occurs; with severe compromise, venous pulsations may be seen.

It is important to recognize abnormal Doppler patterns in the ductus venosus, which carries oxygenated blood from the umbilical vein to the heart. Normally, after 14 weeks gestation, ductus venosus Doppler flow is phasic and antegrade, with no reversal during atrial contraction. In fetuses with elevated central venous pressure, such as complete

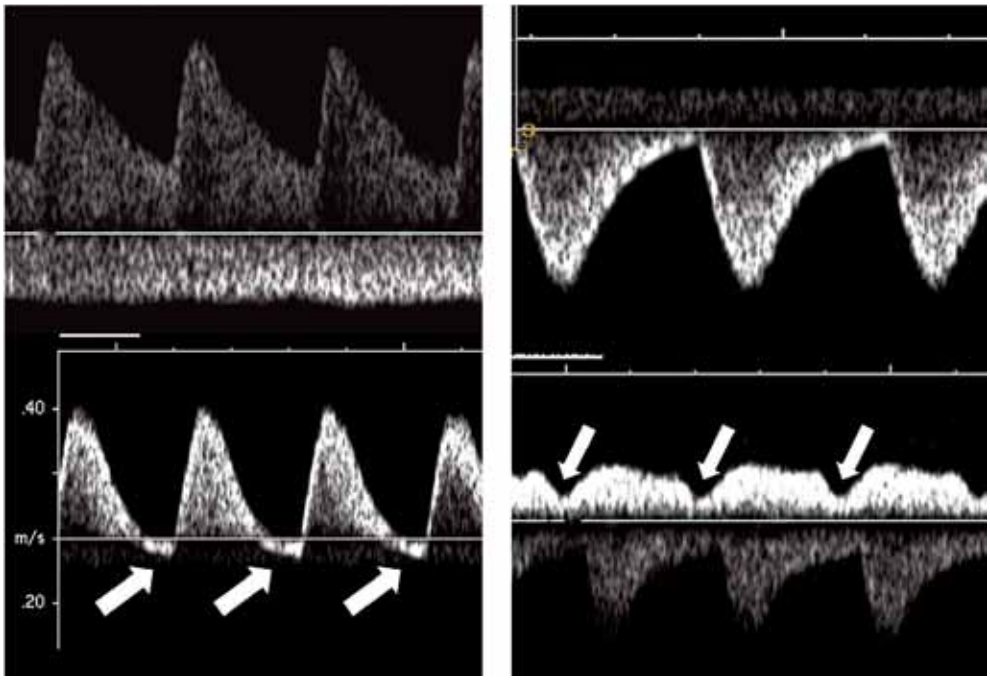

Fig. 3. Umbilical artery and vein Doppler tracings: The top left-hand panel demonstrates normal umbilical artery (UA) and umbilical vein (UV) Doppler flow patterns, the top right-hand panel demonstrates diminished diastolic flow which returns to baseline in the UA. (This panel appears inverted because of the baby's position when the echo was obtained.) The bottom left panel demonstrates reversal of flow in the UA with atrial contraction, as illustrated with the arrows. The bottom right panel demonstrates venous pulsations, as illustrated with the arrows. (With permission from Gleason M, ed. Pediatric Practice: Cardiology. New York: McGraw-Hill Medical; 2012. Chapter 3. Fetal Echocardiography)

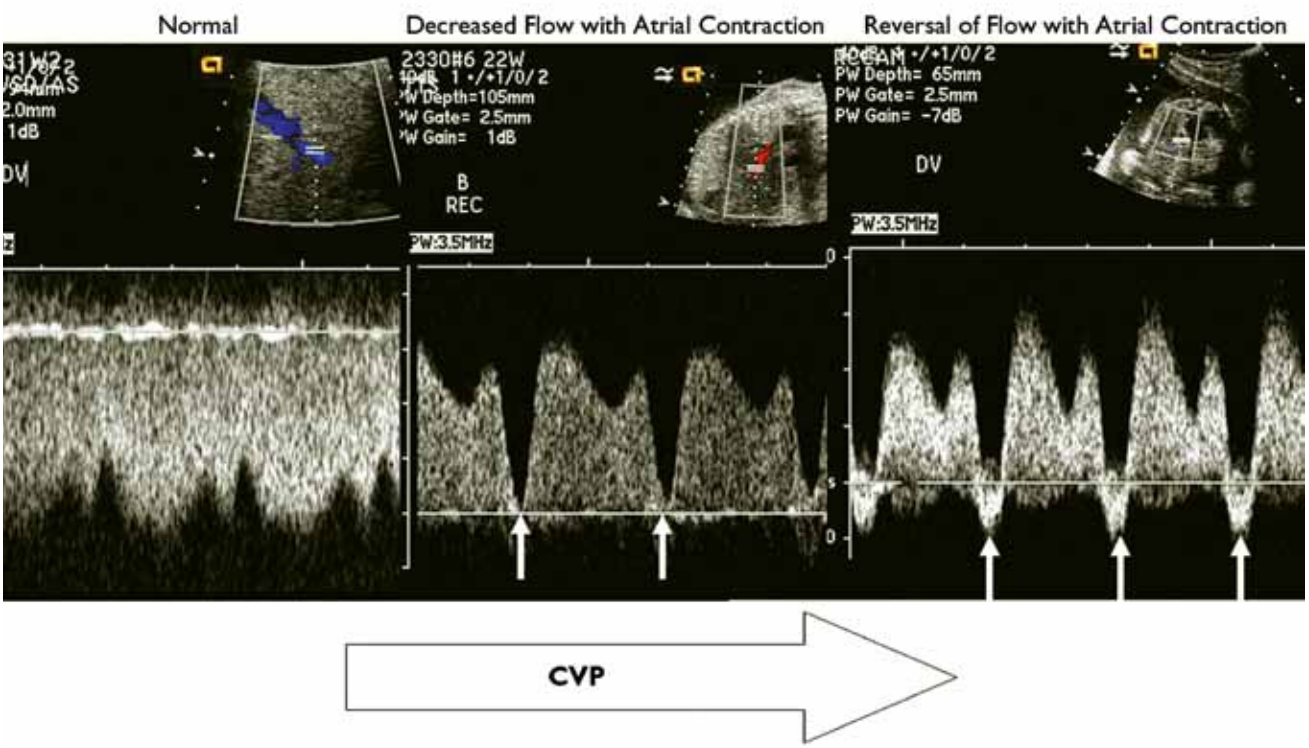

Fig. 4. Ductus venosus (DV) Doppler flow patterns: the left-hand panel demonstrates the normal flow pattern. (This panel appears inverted because of the baby's position when the echo was obtained.) As central venous pressure increases, flow first returns to baseline with atrial contraction. Then there may be reversal of flow with atrial contraction. (With permission from Gleason M, ed. Pediatric Practice: Cardiology. New York: McGraw-Hill Medical; 2012. Chapter 3. Fetal Echocardiography.)

atrioventricular block and severe tricuspid regurgitation, the ductus venosus flow is reversed during atrial contraction, and may occur before changes are seen in the umbilical vein Doppler pattern (Fig. 4).

EXTRACARDIAC ANOMALIES AFFECTING THE FETAL CARDIOVASCULAR SYSTEM

Extra-cardiac anomalies may alter circulating blood volume, compress the heart, or cause other

Table 3. Extracardiac Anomalies Impacting the Fetal Heart

Condition	Impact on the Fetal Heart
■ SCT: Sacrococcygeal Teratoma	Fluid Overload: causes cardiomegaly, potential hydrops, high output heart failure, AV valve leakage
■ C-CAM: Congenital Cystic Adenomatoid Malformation	Space occupying lesion in the lung: depending on size, could lead to mediastinal shift, cardiac compression, lung hypoplasia, hydrops
■ d-hernia: Congenital Diaphragmatic Hernia	Lung hypoplasia, potential mediastinal shift and cardiac compression. Pulmonary artery hypoplasia, pulmonary hypertension, increased risk for congenital heart disease
■ TTTS: Twin-Twin Transfusion Syndrome (Monochorionic Twins)	Recipient twin (larger): fluid overload, polyhydramnios, potential hydrops, RV dysfunction, acquired right heart obstructive lesions (pulmonary stenosis or atresia) Donor twin (smaller): oligohydramnios and growth retardation
■ Tuberous Sclerosis	Intracardiac rhabdomyomas, fetal arrhythmias

problems.[8-12] Table 3 lists such lesions and their consequences.

It is important to recognize that fetal echocardiography has limitations, and not all forms of congenital heart disease can be "ruled out" on a fetal echocardiogram. Atrial septal communications and the patent ductus arteriosus are mandatory in utero, and statistically the vast majority of these close within the first year of life, but we cannot predict which will fail to close. Less severe lesions, such as well-functioning bicuspid aortic valves or small to moderate ventricular septal defects, may be too minor to detect before birth. Additionally, the postnatal development of aortic coarctation cannot be fully ruled out by this testing. These limitations are explained to mothers, so their expectations are appropriate.

The goal of fetal echocardiography is to identify babies with severe congenital heart lesions that may depend upon a patent ductus arteriosus after birth for either systemic or pulmonary blood flow. The PDA closes in a variable time frame after birth, anywhere from 24 hours to a week, so newborns with serious congenital heart disease may be discharged from the hospital with an open ductus, only to present within the first two weeks of life in shock or with severe hypoxia after ductal constriction occurs. Most states, including Pennsylvania, require newborns to undergo screening for critical congenital heart disease within 24 hours of hospital discharge, including pulse oximetry of the right arm and leg.[13] A baby "fails" this screen if either oxygen saturation is less than 95%, or pulse oximetry in the leg is more than 3%-5% lower than in the arm. A "failed" screen requires cardiac assessment including echocardiography.

TYPES OF CHD DIAGNOSED OR SUSPECTED ON FETAL ECHOCARDIOGRAPHY.

I. Left-Sided Obstructive Lesions

Hypoplastic Left Heart Syndrome (HLHS) is a common and fatal syndrome which, like other single ventricle defects, is readily diagnosed with fetal echocardiography. The left heart structures are underdeveloped, with stenosis or atresia of the mitral and aortic valves and varying degrees of left ventricular hypoplasia. (Fig. 5) Babies with HLHS are followed serially in utero to look for changes in right ventricular function, tricuspid valve leakage, or a restrictive atrial septum (which can lead to a poor postnatal prognosis). If the family chooses to continue the pregnancy, or gestation is beyond 24 weeks at diagnosis, it is recommended that the baby be delivered at a tertiary care medical center when possible. Babies are sent to an Intensive Care Unit for complete assessment of the heart, as well as overall evaluation of the baby, including neurological and genetic assessment, as needed. Ductal patency is maintained with prostaglandin E1 infusion after birth, thus avoiding systemic hypoperfusion from ductal constriction, and minimizing end organ damage. This measure may improve overall outcome.[14] Although a few families choose palliative care or listing for cardiac transplantation, many babies are referred for staged surgical palliations towards a Fontan procedure.

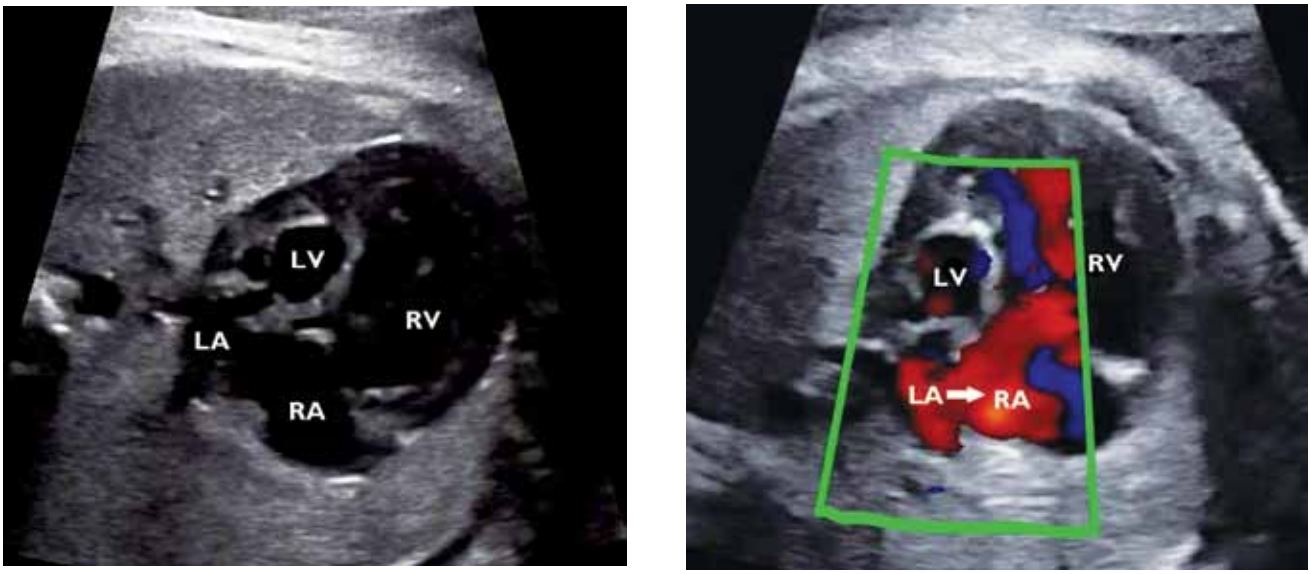

Fig. 5. At left: Four chamber view of hypoplastic left heart syndrome (HLHS): The left ventricle (LV) and left atrium (LA) are significantly smaller than the right ventricle (RV) and right atrium (RA). The LV is echobright consistent with endocardial fibroelastosis. At right: Color flow imaging of HLHS: the arrow shows blood flow going from the left atrium to the right atrium, which is the opposite of normal flow through the foramen ovale.

II. Right-Sided Obstructive Lesions

When there is significant obstruction to blood flow across the pulmonary valve in utero, the PDA shunts bi-directionally instead of purely right to left, in order to perfuse the pulmonary artery bed. By the time of delivery, the branch pulmonary arteries may be normal or hypoplastic, resulting in varying degrees of cyanosis and hypoxia in these neonates. Assessment of the size of the branch pulmonary arteries, and the direction of PDA flow, help in predicting whether pulmonary blood flow will depend on a patent ductus after birth.

Tetralogy of Fallot:

Hallmarks of Tetralogy of Fallot (TOF) include a large ventricular septal defect (VSD) with an overriding aorta that is larger than the pulmonary artery. If the pulmonary artery is <50% of the size of the aorta, with left to right flow in the PDA, prostaglandin E1 will likely be needed after birth to support adequate oxygenation. Echocardiography in conjunction with oxygen saturation levels and differentiation of anatomical variants will assist postnatal choice of palliation with an aorto-pulmonary shunt or complete repair.

Critical Pulmonic Stenosis (PS), Pulmonary Valve Atresia (PA):

When a baby has pulmonic valve stenosis or atresia, the pulmonary artery appears smaller than the aorta; if the pulmonic valve is "critically" stenotic or atretic, the PDA will shunt bi-directionally on the fetal echocardiogram. Infusion of prostaglandin E1 after birth is indicated to maintain a level of oxygenation that allows stability. If the pulmonary valve is patent, with an accelerated Doppler flow pattern, a neonatal interventional cardiac catheterization with balloon dilation of the pulmonary valve will be the likely choice.

Pulmonary atresia with intact ventricular septum (PA/IVS) is a complex congenital heart lesion. The atretic pulmonary valve is accompanied by hypoplasia of the tricuspid valve and right ventricle in many babies. The fetal tricuspid valve (TV) z score has been used as a predictor of right ventricular adequacy, with a TV z score of < -3.5 identified as a predictor of RV hypoplasia.[15,16] If the right ventricle is near normal, then babies may undergo cardiac catheterization with radiofrequency perforation and balloon dilation of the pulmonary valve, but for babies with hypoplastic right ventricles and/or fistulous connections from the right ventricle to one or more coronary arteries, families are counseled about a pathway of staged palliation leading to a Fontan procedure.

III. d-Transposition of the Great Arteries

Dextro-transposition of the great arteries (d-TGA) is a common form of cyanotic CHD. In simple d-TGA (no VSD), the basic cardiac structure and function is normal, but the aorta arises from the anatomic right ventricle ("d" designation) and the pulmonary artery arises from the anatomic left ventricle. As a result, the

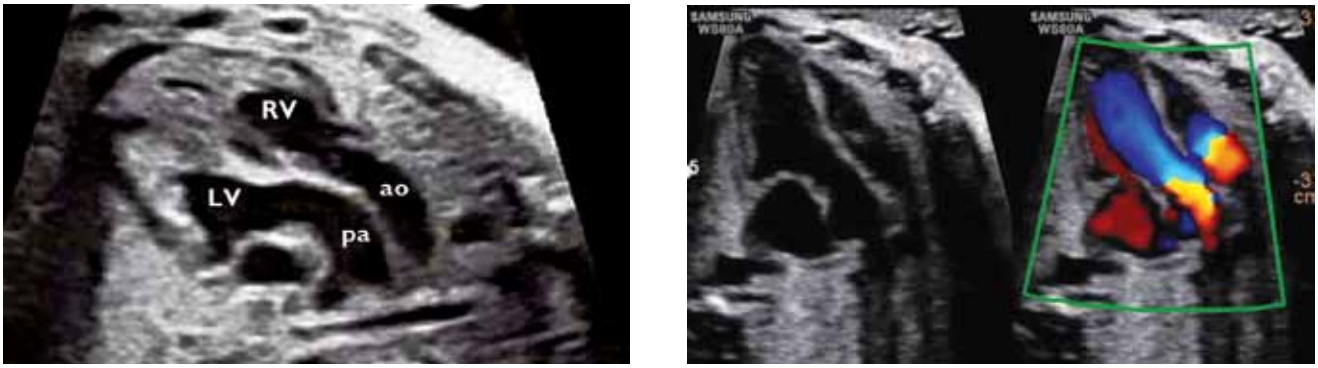

Fig. 6. At left: Apical cardiac imaging including the outflow tracts shows d-transposition of the great arteries (d-TGA). The pulmonary artery (pa) arises from the left ventricle (LV) and the aorta (ao) from the right ventricle. At right: Color flow imaging of d-TGA shows that both outflow tracts are virtually side-by-side and not crossing.

great arteries parallel each other, rather than the normal crossing of the pulmonary artery over the aorta from right to left. If the four-chamber view of the heart is not supplemented by views of the alignment of the outflow tracts and great arteries, it may not be accurately diagnosed in utero. Parallel outflow tracts (Fig. 6) are highly suspicious for d-TGA. Newborns with d-TGA have significant cyanosis, sometimes of a critical nature, and without treatment this form of CHD is incompatible with life.

When a prenatal diagnosis of d-TGA is made, planning the postnatal management is key. Severe hypoxia and a small PFO on postnatal echocardiography usually indicate the need for emergency balloon atrial septostomy (BAS). This procedure tears the PFO flap and allows more oxygenated blood from the left atrium to enter the right heart, thus improving systemic oxygenation. After initial stabilization, the usual course is subsequent referral for an arterial switch (ASO) operation within the first week of life.

It is important to realize that in d-TGA, the fetal brain is perfused by blood with less than normal oxygen content. The greater than expected incidence of attention deficit disorder and learning disabilities in these children may be related to altered prenatal cerebral perfusion. In general, there are many concerns about fetal brain development in babies with CHD.[17,18]

FETAL ARRHYTHMIAS

Abnormal fetal heart rates are often noted on routine obstetric screenings, and may be too slow, too fast, or irregular. The normal fetal heart rate range is 120 to 160 beats/minute, and should vary with fetal activity. Most fetal arrhythmias are benign, with a minority being life-threatening, leading to fetal hydrops.

Fetal arrhythmias are a common indication for two-dimensional and Doppler echocardiography. In addition to assessing structural abnormalities and dysfunction, the cause of the arrhythmia can be evaluated by pulsed wave Doppler flows at the atrial and ventricular levels, as well as M-mode imaging of atrial and ventricular wall movement. Arrhythmias in the fetus include premature atrial or ventricular contractions (PACs, PVCs), tachyarrhythmias, and abnormal atrioventricular (AV) conduction.

I. Atrial ectopy

Premature atrial contractions (PACs) are the most common arrhythmia seen in the fetus and are generally benign. Doppler interrogation of mitral inflow and aortic outflow will show a premature mitral deflection from the premature atrial beat which may be conducted (a small aortic deflection also occurs), or not (no aortic deflection). Since non-conducted PACs are not "heard" on obstetric Doppler, the effective heart rate will seem slower and irregular. The estimated mechanical PR interval (time from atrial contraction to aortic outflow on pulsed Doppler) is measured to assess atrioventricular (AV) conduction, and should normally be </= 120-130 msec. If this value is prolonged, it suggests that the irregular beat is due to a dropped beat from second degree atrioventricular block (AVB). Very frequent PACs may be a precursor of supraventricular tachycardia (SVT) or atrial flutter.

Supraventricular tachycardia, when sustained, can lead to fetal congestive heart failure, hydrops and even fetal demise. Usual re-entrant SVT rates are 180 to 280 bpm with a 1:1 ventricular response on Doppler or M-mode echo. When atrial flutter is present, the atrial rate is faster, generally near 400 bpm,

with a slower ventricular response.

Treatment of fetal SVT varies. If the baby is near term, the SVT is non-sustained, and there is no hydrops, close follow up is indicated. When SVT is sustained, hydrops is present, or if gestational age would not allow survival after delivery, prenatal medical management may be indicated. Successful treatment usually results from administering anti-arrhythmic medications to the mother in a hospital setting, in conjunction with our colleagues in Maternal Fetal Medicine and adult cardiology. Digoxin, sotalol, flecainide and amiodarone are well tolerated, but mothers need monitoring for pro-arrhythmic effects of certain drugs. Digoxin has the longest track record for success. Some medications are delivered directly to the fetus via umbilical cord injection. If a baby shows compromise from SVT, with the development of hydrops, then early delivery may be warranted, depending upon the fetal gestation at the time.

II. Abnormal A-V conduction

Congenital complete heart block (CCHB) is rare and occurs in ~ 1 in 20,000 newborns. Fetal bradycardia due to lack of atrio-ventricular conduction is usually associated with complex CHD or neonatal lupus (due to trans-placental transfer of maternal auto-antibodies; anti-Ro/anti-SSA and anti-La/anti-SSB). Less often it is idiopathic. Cardiac output depends on the underlying ventricular rate, which varies, but rates below 40 to 50 bpm can lead to heart failure. Fetal echocardiography can document that atrial wall motion is faster than, and independent of, ventricular wall motion. Fortunately, treatment of CCHB in utero is rarely required, though steroids and beta agonists have been given to mothers. If however, there is fetal hydrops, early delivery and postnatal transvenous pacing may be indicated. The decision to place a pacemaker depends upon the clinical cardiac output and the presence or absence of CHD.

CHD AND GENETIC ABNORMALITIES

During routine prenatal care, screening for fetal anomalies of all types is performed. When prenatal imaging shows certain structural abnormalities, there may be a suspicion for a genetic syndrome, and hence, there will also be a high suspicion for associated CHD (Table 4). Advances in non-invasive prenatal testing (NIPT) allow greater sensitivity in detecting some chromosomal anomalies in-utero. In addition to first trimester ultrasound measurement of nuchal skin thickness, pregnant women often undergo screening for genetic abnormalities, including NIPT blood sampling (i.e. Harmony test, cell free DNA, MaterniT21) and Quad screens. An abnormal result is an indication for fetal echocardiography. On the other hand, when certain types of CHD are recognized on prenatal imaging, then an evaluation for genetic syndromes is pursued. The most accurate genetic information comes from chorionic villous sampling (CVS) and amniocentesis, but these are

Table 4. Congenital Heart Defects and Associated Genetic Syndromes[19]

CHD Defect	Chrom / Gen Defect	Syndrome	Extracardiac Findings
Conotruncal: TOF Truncus	22q11 deletion	DiGeorge / VCF	Cleft palate, hypocalcemia, immunological-T cell dysfunction, feeding/speech issues, neurodevelopmental issues, schizophrenia
Branch pulmonary stenosis (PPS)	JAG-1	Alagille	Bile duct paucity, butterfly vertebrae
Coarction Bicuspid Aortic Valve and Other Left Heart Outflow Lesions	XO	Turner's	Cervical cystic hygroma, lymphedema-pedal edema, IUGR, horseshoe kidney, infertility, variable mental and developmental problems
PS Hypertrophic Cardiomyopathy	PTPN11 KRAS SO1	Noonan	Posterior nuchal cystic hygroma, polyhyradmnios, special facial features (low set ears, depressed nasal bridge, large head)
AV Canal Defect VSD	T21	Down's	Increase NT, macroglossia, hypoplastic 5th metacarpal, duodenal atresia
VSD, PS	T18	Edward's	IUGR, clindactyly, rockerbottom feet, esophageal atresia
VSD	T13	Patau	Holoprosencephaly, cleft lip / palate
Interrupted IVC		Heterotaxy	Biliary atresia, polysplenia, gut malrotation
ASD	TBX5	Holt Oram	Upper limb abnormalities-absence of thumb, club hand

invasive and carry small, but real, risks to the pregnancy.

CONCLUSION

Fetal echocardiography is a valuable tool in managing pregnancies in which there is an increased risk for the development of congenital heart disease or concern about genetic abnormalities, or in cases where arrhythmias or extracardiac malformations might adversely affect an otherwise normal fetal heart. It is critical for perinatologists and pediatric cardiologists to work together to get the most accurate information about fetal hemodynamics, which can guide appropriate management paths, and hopefully result in better outcomes for both mothers and babies.

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