FETAL ECHOCARDIOGRAPHY: IN UTERO ACOUSTIC WINDOW ON THE DEVELOPING HEART

MOHAMEL LA...SELIEM, MI; RICHARD H. SAN MIGUEL, MD; MAE P ALILEO, BSMT

The purpose of this study is to evaluate prospectively the accuracy of fetal echocardiography performed at a tertiary referral center (Aramco Dhahran Health Center) in defining the exact anatomic features of various cardiac lesions in fetuses with congenital heart disease. Fetal echocardiography has been used to plan the appropriate course of care for high-risk pregnancies and in the immediate neonatal period. This includes a team of obstetrician, radiologist, pediatric cardiologist, and neonatologist. A sample of 65 consecutive fetal echocardiograms and their matching neonatal echo/Doppler studies were compared. The referral sequence was initiated by the obstetrician to the radiologist, then to the pediatric cardiologist, and the neonatologist. Four groups of fetuses/neonates were identified in this study based on their fetal cardiac examination: (I) normal cardiac anatomy and performance (n = 30): none had any significant cardiac lesion on postnatal echocardiogram; (2) fetal arrhythmia (n = 12): premature atrial beats and bigeminy were found in 9 fetuses and disappeared in the immediate neonatal period. Atrial tachycardia (atrial rate 400's – ventricular rate 200's) was seen in 3 fetuses and required maternal and neonatal treatment with digoxin in one; (3) nonimmune hydrops fetalis was diagnosed in 8 fetuses: it was mild and resolved in 7 patients shortly after birth. One fetus died in utero with severe hydrops; and (4) complex congenital cardiac lesions were found in 11 fetuses (18%). Some minor anatomic findings could not be defined prenatally in 7 of them. Four cases were excluded due to lack of correlation between prenatal and postnatal studies. We conclude that fetal echocardiography, as an intimate part of a comprehensive fetal assessment, is highly sensitive and specific in detecting congenital heart disease in the fetuses; (2) appropriate management of high-risk pregnancies and neonates can be significantly improved when the exact pathophysiology of the unborn fetus is known by fetal echo/Doppler; and (3) fetal echocardiography is extremely safe and reliable throughout the three trimesters.

TECHNOLOGICAL ADVANCES in echocardiography have revolutionized the field of prenatal diagnosis over the last two decades.1-5 The availability of high resolution ultrasound systems has enabled us to recognize congenital cardiac lesions in the fetus as early as the first trimester of pregnancy, to understand the pathophysiology of the more complex anomalies, to intervene with different therapeutic modalities in utero, and to plan the appropriate course of care for the prospective newborn with congenital heart disease.4

Care for the unborn fetus with congenital disease requires the collaborative work of the obstetrician, radiologist, neonatologist, and the pediatric cardiologist.1-3 The purpose of this study is to evaluate prospectively the accuracy of fetal echocardiography performed at a tertiary referral center (Aramco Dhahran Health Center) over the last three years (1992-1994).

Methods

Patients

A sample of 65 consecutive fetal echocardiograms and their matching neonatal echo/Doppler
PETAL ECHO

studies were included in this prospective study. Four cases were excluded because the fetal studies were not followed by a neonatal examination (these four neonates were discharged from the hospital prior to the echocardiographic examination; they had normal fetal echocardiograms and normal physical examination at birth).

The referral sequence is initiated by the obstetrician to the radiologist if the fetus is suspected to have any congenital malformation, including those of the cardiovascular system on the initial obstetric scan. The radiologist will then examine the fetal heart (four-chamber view), and if a cardiac abnormality is suspected, the pregnant woman is referred to the pediatric cardiologist for fetal echocardiogram. If the latter is positive for congenital cardiac anomalies, the neonatologist is also informed.

Fetal Echocardiographic Doppler Examination

The entire examination is performed by a pediatric cardiologist with special experience in pediatric and fetal echocardiography. The average examination lasts for 30 minutes (range, 20 to 60 minutes) with the mother in the supine position and her head slightly elevated. A HP-sonos 1500 ultrasound system with 5 MHz (short and medium focus), 3.5 and 2.5 MHz transducers was used.

The examination begins by using the lowest possible transducer frequency. The position of the fetus relative to the mother is first identified and then the situs of the heart within the fetal chest cavity is determined (the heart is i:1 normal situs when it lies on the same side of the fetal trunk as the fetal stomach).

Four-chamber views and long- and short-axis parasternal views are usually obtained easily. All four chambers are identified as well as all four valves. M-mode recording of cardiac chamber dimensions and wall motions (in arrhythmias) is done routinely. Doppler examination of all valves, patent foramen ovale, and patent ductus arteriosus is also recorded. Color-flow mapping is routinely used. Continuous-wave Doppler is limited to cases where the degree of valve stenosis and/or shunt is an important part of the examination.

M-mode and/or Doppler tracings are recorded on paper format to calculate the fetal-heart rate and to determine the atrioventricular rate relationship. The results of the fetal examination are then reported to the referring obstetrician or radiologist.

Following the birth of the fetus, the neonate undergoes comprehensive echocardiographic examination within the first 24 hours after birth. The results of the fetal echocardiographic examination are then compared with those of the neonatal examination.

Results

Sixty-one cases were included in the final analysis. Fetal age at which screening was performed ranged from 16 weeks to full term (40 weeks gestation). Four categories of fetal echocardiographic findings were identified: (1) normal cardiac anatomy and performance, (2) fetal arrhythmia, (3) hydrops fetalis, and (4) congenital cardiac disease.

Normal Cardiac Anatomy and Performance

Thirty fetuses had normal cardiac anatomy and performance. The indications for the fetal examination included the following: suspicion of a structural heart disease on obstetric examination of the fetus (12 cases), fetal arrhythmia (10 cases), polyhydramnios (2 cases), single umbilical artery (1 case), intrauterine growth retardation (1 case), and history of congenital heart disease in previous siblings (4 cases). All cases with suspected fetal arrhythmia were sinus arrhythmias. All matching neonatal examinations were normal.

Fetal Arrhythmia

Twelve fetuses had an arrhythmia during the fetal examination. Three types of arrhythmias were identified: (1) premature beats (7 cases): all were presumed to be of atrial origin. These were also seen during fetal cardiac monitor in the immediate neonatal period and all disappeared during the neonatal period (first month of life); (2) atrial bigeminy and trigeminy (2 cases): these were also confirmed postnatally and ran the same course like the premature beats; and (3) atrial tachycardia (3 cases) (Figure I): in one fetus (37 weeks gestation) the atrial rate was 420 beats/min while the ventricular rate was 210 beats/min (i.e., 2:1 atrioventricular relationship). This fetus had mildly dilated heart with mildly mitral and tricuspid regurgitations but no hydrops. The mother was treated with digoxin, but she delivered after two
Indications for fetal echocardiography in our institution generally fall into one of three categories5,7,9: (1) maternal indications: including congenital heart disease in the mother, systemic disease known to affect the cardiovascular system of the fetus (e.g., collagen vascular disease, diabetes, etc), previous pregnancies with congenital heart disease, oligo- or polyhydramnios, and exposure of the mother to certain teratogenic drugs (e.g., alcohol, lithium, indocin, etc); (2) fetal indications: multiple congenital anomalies, abnormal four-chamber view or suspected fetal arrhythmias on obstetric scans, intrauterine growth retardation, abnormal fetal karyotype, and hydrops fetalis; and (3)

Figure 1. (A) M-mode recording of the atrial wall contractions (arrow heads) in a fetus with atrial tachycardia – atrial rate is approximately 420 beats/min. (B) Doppler signals (arrow heads) of the pulmonary artery in the same fetus in (A). Note that the number of semilunar valve openings (and hence ventricular rate) is approximately 210 beats/min, i.e., 2:1 atrioventricular rate relationship. (C) EKG tracing from the same fetus following birth. The neonate was still having atrial tachycardia.

Fetal echocardiography has become a standard procedure to screen for congenital cardiac anomalies in the developing fetus over two decades.1-5 The technique has proved to be highly sensitive and specific in detecting and defining different congenital cardiac anomalies including complex ones. In addition, there have been no significant reports to date to indicate any adverse side effects in the developing fetus even when performed as early as the first trimester of pregnancy. The theoretical effect of prolonged exposure of biological tissues to ultrasound waves (especially during Doppler examinations)
Table I. Spectrum of complex congenital cardiac disease diagnosed by fetal echocardiography with matching neonatal examination.

<table>
<thead>
<tr>
<th>No.</th>
<th>Fetal echocardiographic diagnosis</th>
<th>Postnatal echocardiographic diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Patent of Cantrell with severe hypoplastic right heart syndrome (Figure 3)</td>
<td>Same</td>
</tr>
<tr>
<td>2</td>
<td>Patent of Cantrell with tetralogy of Fallot</td>
<td>Same (the congenital cardiac defect was more consistent with hypoplastic right heart syndrome)</td>
</tr>
<tr>
<td>3</td>
<td>Situs solitus, double inlet single ventricle (RV), pulmonary atresia</td>
<td>Same + dextrocardia and bilateral SVC with LSVC-CS</td>
</tr>
<tr>
<td>4</td>
<td>Unbalanced right-sided AV, hypoplastic LV truncus arteriosus, severe AVV regurgitation</td>
<td>Same + right aortic arch</td>
</tr>
<tr>
<td>5</td>
<td>Common AVC with two AV ostices, unbalanced towards the RV with mild hypoplasia of LV and aortic valve</td>
<td>Same + anomalous attachments of MV to LVOT</td>
</tr>
<tr>
<td>6</td>
<td>Hypoplastic left heart syndrome with mitral and aortic atresia</td>
<td>Same</td>
</tr>
<tr>
<td>7</td>
<td>Hypoplastic left heart syndrome with mitral and aortic atresia</td>
<td>Same + LSVC-CS</td>
</tr>
<tr>
<td>8</td>
<td>Hypoplastic left heart syndrome with mitral and aortic atresia</td>
<td>Same</td>
</tr>
<tr>
<td>9</td>
<td>Situs solitus, D-TGA, mitral atresia, hypoplastic LV</td>
<td>Same + pulmonary stenosis</td>
</tr>
</tbody>
</table>
| 10  | Double outlet right ventricle, D\text{}

(Note: more anatomical details were detected during the postnatal echocardiographic examination (following the [+] sign under postnatal echocardiographic diagnosis).

RV = right ventricle; SVC = superior vena cava; LSVC-CS = left superior vena cava-Io-coronary sinus; AVC = common atrioventricular canal; LV = left ventricle; AV = atrioventricular valve; MV = mitral valve; LVOT = left ventricular outflow tract; D-TGA = D-transposition of the great arteries.

Hydrops Fetalis

Eight fetuses had evidence of nonimmune hydrops fetalis. Five had small-to-moderate pericardial effusions and three had pleural effusions only. In one fetus, there was marked hypoplasia of both lungs (Figure 2) and in utero pleural drainage was attempted, however, the fetus died in utero following the procedure. No arrhythmias were documented in any of these fetuses and all had normal cardiac anatomy. Postnatally, all pericardial effusions were found to be small and resolved spontaneously without consequences.

Congenital Cardiac Disease

Eleven fetuses had complex congenital cardiac disease. All were confirmed by postnatal examination (Table I). Minor details of the anatomical features were found on the neonatal examination, however, the diagnoses were essentially the same.

Discussion

Fetal echocardiography has become a standard procedure to screen for congenital cardiac anomalies in the developing fetus over two decades.1-5 The technique has proved to be highly sensitive and specific in detecting and defining different congenital cardiac anomalies including complex ones. In addition, there have been no significant reports to date to indicate any adverse side effects in the developing fetus even when performed as early as the first trimester of pregnancy. The theoretical effect of prolonged exposure of biological tissues to ultrasound waves (especially during Doppler examinations) has not been substantiated to date by any reports of clinical significance.6

Who Performs Fetal Echocardiography

Recognition and further management of fetal cardiac anomalies in the ideal set up is a collaborative effort of the obstetrician, radiologist, pediatric cardiologist, neonatologist, and pediatric cardiac surgeon. Such sequence of referral has been used in our institution for the past three years. The fetal cardiac examination is eventually performed by the pediatric cardiologist who has special experience with pediatric echocardiography.1-3.7
When Is Fetal Echocardiography Indicated

Indications for fetal echocardiography in our institution generally fall into one of three categories5.7.9: (1) maternal indications: including congenital heart disease in the mother, systemic disease known to affect the cardiovascular system of the fetus (e.g., collagen vascular disease, diabetes, etc), previous pregnancies with congenital heart disease, oligo- or polyhydramnios, and exposure of the mother to certain teratogenic drugs (e.g., alcohol, lithium, indocin, etc); (2) fetal indications: multiple congenital anomalies, abnormal four-chamber view or suspected fetal arrhythmias on obstetric scans, intrauterine growth retardation, abnormal fetal karyotype, and hydrops fetalis; and (3) familial risk factors: significant genetic syndromes or congenital heart disease in the family.

Spectrum of Fetal Cardiac Anomalies Detected By Echocardiography

The main objective of the present study is to evaluate the accuracy of fetal echocardiography performed at our institution in defining a wide spectrum of fetal congenital cardiac defects.

Among 65 cases included, 15 fetuses (23%) had significant disease (11 with congenital cardiac defect, 3 with atrial tachycardia, and 1 with severe nonimmune hydrops). All cases with complex congenital cardiac defect were accurately identified. However, minor anatomic details were identified only on the postnatal examinations (Table 1). Paradoxically, common and simple cardiac defects, e.g., isolated ventricular septal defects, atrial septal defects, and mild valvar pulmonary stenosis were underrepresented in this series. This may be related to our inability to define such details with the current ultrasound systems and also because complex lesions are more likely to manifest themselves in hemodynamic impairment prenatally.3

In addition to the anatomical lesions,3.10 fetal arrhythmias were well diagnosed and characterized using a combination of M-mode and Doppler techniques.4.11 The former was excellent in defining "the rate" of atrial wall contractions and the latter in defining "the rate" of the semilunar valve opening, i.e., the atrioventricular rate relationship (Figure 1).

Counseling of Prospective Parents

The results of this prospective echocardiography study are the first step in the process of counseling prospective parents regarding the prognosis and outcome of a fetus with certain congenital cardiac defect. With low false positive and negative diagnoses, the discussion about the prognosis of the prospective newborn is more objective and realistic. When the fetus has severe complex congenital cardiac lesion and other noncardiac anomalies, i.e., a lethal syndrome, it becomes an option to the parents to terminate the pregnancy during the first trimester.1-3

Acknowledgment

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References