The first recommendations and guidelines for physicians training in fetal echocardiography were created in 1990 and later updated by multiple medical associations and journals in Europe and the United States. This time advanced fetal cardiac ultrasound recommendations focused more on the organizational and logistical aspects of FE, to better define the fetal echo guidelines for practitioners in tertiary centers. Underlined is FE in 3rd trimester, with special attention to the direction of flow across the foramen ovale and ductus arteriosus.

AHA classification of heart defects in prenatal cardiology into seven major groups (from 2014) is presented as well as the Polish classification into four groups (from 2012) related to the urgency of required time to postnatal treatment/intervention based on FE findings in the 3rd trimester of pregnancy.

Current definition of fetal cardiologist in 2015 is also presented.

Key words: fetal heart classifications, 3rd trimester, fetal cardiologist

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How to cite this article:

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Submitted: 2015-06-14; accepted: 2015-07-02

Abstract
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North American guidelines for low risk pregnancies initially focused on the inclusion of the four-chamber view with an attempt to visualize the outflow tracts only if technically feasible. However, more recent publications have emphasized the importance of
visualizing the outflow tracts to improve identification of conotruncal cardiac anomalies. Also, the previous AIUM guidelines for fetal echocardiography published in 2011 recommended neither storage of digital clips nor the use of color flow Doppler, but these recommendations have been revised in more recent North American guidelines for fetal echocardiography (echo) published in 2013 [11-13].

In the May, 2014 edition of Circulation, a major document on fetal cardiology, “Diagnosis and Treatment of Fetal Cardiac Disease: A Scientific Statement From the American Heart Association”, provided a thorough evaluation of the current practice of fetal cardiology. This paper laid the framework for the sub-specialty of fetal cardiovascular medicine, including a detailed review on prenatal diagnosis and management of congenital heart disease (CHD), assessment of fetal cardiac function and the fetal cardiovascular system, and current in-utero fetal therapy options. Recommendations include the timing of referral for study, indications for referral, and the experience required for the performance and interpretation of fetal echocardiograms. Complementary modalities for fetal cardiac assessment are reviewed. Recommendations for parental counseling are described. Fetal therapies, including medical management of arrhythmias and heart failure and closed or open intervention for diseases affecting the cardiovascular system are discussed. Experimental catheter-based fetal cardiac interventions to prevent the progression of valvular disease in utero are also discussed. Recommendations for delivery planning strategies for fetuses with CHD including models based on classification of disease severity and delivery room treatment are also defined.

This article in Prenatal Cardiology will compliment previous published guidelines and recommendations, while focusing more on the organizational and logistical aspects of fetal echocardiography. Our aim is to better define the fetal echo guidelines for practitioners in tertiary centers, encompassing early diagnoses and emphasizing the importance of appropriate parental counseling for prenatal diagnosis. The guidelines are intended to address patients that have already been diagnosed with suspected fetal CHD and the family has now been referred to a tertiary care/higher level center that routinely manages these types of patients.

This position paper is not specific for screening the fetal heart as there is a separate protocol for basic fetal heart examination in this issue of Prenatal Cardiology. In this prior article we recommend that the evaluation (pages 24,25,26) of the fetal heart during routine anatomy scans include sweeps and clips in both transverse and sagittal planes for obtaining 5 cardiac views which are abdominal situs, 4 chamber view, aorta from LV, pulmonary artery from RV including crossover of arteries and the 3 vessel tracheal view; both with and without color Doppler. While there is generally agreement on the appropriate fetal heart views that should be part of a basic fetal echo examination, achieving a complete examination of the fetal heart remains difficult. We believe that, when heart images cannot be obtained during routine anatomical scans, a repeat or targeted fetal echo should be offered. This is especially true for outflow tract anomalies such as transposition of the great arteries where the identification rate in utero is less that 50%. Another issue that has been increasing over time is difficulty of obtaining diagnostic grade images in obese gravidas.

These guidelines will focus on the performance of a complete fetal echocardiogram, performed by a dedicated team of specialists (fetal and pediatric cardiologists, MFM specialists or radiologists).

The importance of fetal echocardiography in 3rd trimester has not been well defined so far. However, some forms of both structural and functional congenital heart disease may progress during gestation and may not be clinically evident at the usual time period that routine fetal screening ultrasound examination is performed. Examples include diagnoses such as rhabdomyomas associated with tuberous sclerosis causing intracardiac obstruction; vein of Galen arterio-venous malformation causing fetal heart failure due to volume loading and diabetic fetal cardiomyopathy with obstruction. In this new set of guidelines, a team of colleagues from Europe and the United States share their experience as fetal cardiologists with other specialists in perinatology, in order to find a common language and better understanding which may improve the perinatal care for fetuses with CHD. The detailed fetal cardiovascular ultrasound evaluation, or fetal echocardiography, is now established as the standard of care around the world in selected pregnancies at risk for fetal heart disease. The indications, timing and approach to fetal echocardiography have been detailed by various organizations [6,7,11].

General Concepts: Fetal echocardiography (FE) at any point in gestation must minimize the thermal index and concentrated exposure to prolonged use of color/pulsed Doppler. All consultative scans should be thoroughly documented with still frame images and clips or continuous video recordings (on DVD).

13-17 weeks: We recommended FE for increased nuchal translucency seen at the 11th to 13th week dating scan and to clarify abnormal cardiac findings at obstetrical evaluation. Early studies are particularly useful for diabetic mothers with elevated HgbA1c, parents with previous child that has complex CHD, and suspected genetic anomaly and also after infertility treatment. This study should include structural and functional cardiovascular evaluation that would be checked again at the 20 to 26
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week gestation fetal echocardiogram. Early FE scans are limited due to the small size of the heart making accurate measurements challenging with larger chance for error. This early FE study may confirm normal heart anatomy or may detect major CHD. This early FE study usually would not detect mild stenosis of semilunar valves or abnormal pulmonary venous connections. All early FE studies should be followed by a second trimester study to rule out potential changes in diagnosis, chamber size and cardiovascular function.

18-28 weeks: This FE study will typically include a complete ultrasound examination for suspected abnormalities of the heart, or other high risk factors for fetal CHD. The FE evaluation should begin with fetal biometry to assess fetal age and estimated weight. Placental location, AFI and maternal BMI or weight should be included in the report. Initial scanning may also include assessment of the placenta for chorioangiomas, cord insertion, number of cord vessels and to rule out vasa previa. Establishing fetal position of the head and spine, putting body marker on the screen, confirming cardiac and abdominal situs should all be performed prior to evaluation the fetal cardiovascular system. Peripheral Doppler measurements such as measuring the middle cerebral artery Doppler pulsatility index, umbilical artery Doppler pulsatility index and assessing the ductus venosus can be performed at any point during the exam. Find the abdominal cord insertion and follow the umbilical arteries around the fetal bladder then continuing into the abdomen to follow the intra-abdominal course of the umbilical vein to look for pulsatility. Show that the portal extension of this vessel continues to the right and opposite of the fetal stomach to rule out persistent right umbilical vein. At the top of the curve of this vein lies the ductus venosus which narrows and enters the left hepatic vein just before joining the inferior vena cava. The ductus venosus (DV) should be centrally located just below the diaphragm and at the level of the fundus of the left sided stomach. The DV pulsatility should be biphasic and always showing positive flow toward the heart throughout the cardiac cycle in order to assure normal central venous pressure. The heart should then be imaged to show its location within the central left thorax above the stomach. This insures normal situs. Situs also involves looking at relative position of descending aorta and IVC in abdomen and position of abdominal organs such as stomach. Any variation from normal situs is a flag for CHD or a mass effect on the fetal heart. The relative positions and function of all systemic and pulmonary veins, relative chamber sizes and morphologies, all valves, and both the aortic and ductal arches should be imaged both statically as a saved freeze frame and dynamically in a video clip.

Fetal echocardiography represents a detailed evaluation of fetal cardiovascular position, axis, size (HA / CA and C/T ratio)*, structure, function and rhythm, Color Doppler of valves and outflow tracts, crossing of outflow tracts, PW Doppler of valves and vessels, and cardiac biometry. As such, this examination should include multiple modalities, including 2D analysis, color, pulsed, and basic spectral Doppler, and M-mode. In cases of potential heart

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*HA/CA = heart area / chest area ratio. C/T = chest / thorax circumference ratio

Figure 1: The comparison of fetal, newborn and infant circulation; pay attention on the direction of blood flow at the atria level: right-left in healthy fetus, left-right in healthy newborn, no flow in infant.
failure, additional detailed Doppler evaluation (such as cardiovascular profile score, tissue Doppler) or 3D/4D fetal cardiac imaging may be indicated. In most cases, continued surveillance of shunting patterns across the foramen ovale and ductus arteriosus should be performed, including assessment for ductal constriction.

**Recommendations:**

Follow up studies for established CHD to assess functional changes and plan the timing and location of delivery can be scheduled at approximately 4 week intervals or earlier when considering in utero treatment of heart failure or persistent arrhythmias. In cases of maternal pharmacological treatment, particularly if the treatment includes substances known to cause ductal constriction, special attention should be paid to ductus arteriosus flow. Should tricuspid valve regurgitation be detected, especially with constriction of the ductus arteriosus, the mother should be questioned about her use of NSAIDs, acetaminophen or other drugs.

Schematic drawings are recommended for parental counseling sessions, with the abnormal variations found during the fetal echocardiogram.

In cases of established CHD, it is recommended to pay special attention to the direction of flow across the foramen ovale and ductus arteriosus: right-left? bidirectional? or evidence of left-to-right shunting which may be crucial for delivery and post-partum management (Fig.1).

Recording the umbilical cord Doppler at slow sweep speed is important in detecting the frequency of ectopic beats or the duration of tachycardia or bradycardia in patients referred for fetal arrhythmias. Abnormal Doppler tracing should be differentiated from fetal hiccups. The Doppler recording of the hepatic vein will prove the atrial origin of single ectopic beats. M-mode recordings of the of the atrial wall, the septum primum movement, semilunar valve and ventricular wall can help delineate atrial from ventricular tachycardias and also confirm complete heart block. Doppler measurements through the LVOT including mitral inflow and aortic outflow can define the PR and AV interval and isovolumetric contraction and relaxation times, which may quantitated with a myocardial performance index (MPI or Tei Index).

**29 to 39 weeks:** We recommend the same comprehensive FE be performed at 20 to 27 weeks as described above to look for changes in cardiac size and function or progression of disease. Cardiomegaly can develop in the presence of structural CHD, pericardial effusion, AV valve regurgitation, fetal anemia or volume loading lesions such as arterio-venous malformations. Another cardiac anomaly that may not be seen before 25 weeks gestation is rhabdomyoma associated with familial tuberous sclerosis.

Close serial monitoring of should be performed for certain cases, optimally starting at 4 week intervals, through 3, 2 and 1 week intervals up to the time of delivery in order to identify those fetuses at risk for restriction at the

<table>
<thead>
<tr>
<th>Status Level</th>
<th>Definition</th>
<th>Treatment</th>
<th>Prognosis</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Cardiovascular findings with minimal postnatal impact</td>
<td>None</td>
<td>Excellent, normal</td>
<td>No significant disease</td>
</tr>
<tr>
<td>2</td>
<td>Postnatal follow required with possible treatment</td>
<td>Postnatal evaluation required for treatment assessment</td>
<td>Excellent, normal</td>
<td>Mild diseases with possible need for surgery or catheter treatment</td>
</tr>
<tr>
<td>3</td>
<td>Simple cardiovascular findings requiring surgery (2 ventricular repair)</td>
<td>Surgery or catheter treatment required</td>
<td>Excellent, normal</td>
<td>One intervention required</td>
</tr>
<tr>
<td>4</td>
<td>Complex cardiovascular findings requiring surgery (2 ventricular repair)</td>
<td>Surgery and likely further treatment required</td>
<td>Excellent, normal</td>
<td>One intervention required</td>
</tr>
<tr>
<td>5</td>
<td>Complex cardiovascular findings requiring surgery (1 and later possibly 2 ventricular repair)</td>
<td>Fontan surgical strategy required</td>
<td>Good, close to normal</td>
<td>Single ventricle: low risk. 2 ventricles high risk. One intervention required. Fair to good survival</td>
</tr>
<tr>
<td>6</td>
<td>High risk complex cardiovascular findings requiring surgery (2 ventricular repair)</td>
<td>Fontan surgical strategy required but at very high risk</td>
<td>Fair to poor</td>
<td>Survival beyond childhood is possible but unlikely</td>
</tr>
<tr>
<td>7</td>
<td>Poor outcome complex cardiovascular findings requiring intervention</td>
<td>Intervention may be offered with poor prognosis</td>
<td>Demise likely</td>
<td>Survival not expected</td>
</tr>
</tbody>
</table>

Table 1: Cardiac defects classification according to American Heart Association in 2014
foramen ovale and/or constriction of the ductus arteriosus.

In the case of fetal well-being with CHD there is usually not a need to plan cesarean section in advance. The majority of newborns with severe CHD will be born in good clinical condition, at term, with good Apgar scores. Most of these babies will not require cardiac intervention immediately after delivery

However, FE should focus on those cases with potential restriction at the foramen ovale and/or ductus arteriosus and with increasing cardiomegaly and other symptoms suggesting deterioration. These ominous signs can be silent during routine obstetrical cardiotocography.

For those selected cases — fetuses with CHD that are ductal dependent and or critical who might be a candidate not only for neonatal prostaglandin infusion but also for balloon valvuloplasty after delivery or early open surgical approach.

It may be worth considering having a special delivery room and perinatal team of dedicated specialists including obstetrician, fetal cardiologist, pediatric invasive cardiologist, neonatologist, anesthesiologists (both for mother as well as for the newborn), and cardiac surgeon. That group of CHD are nowadays most important to select from all prenatally diagnosed CHD.

Consultations: A fetal echocardiogram is typically performed in conjunction with extensive consultation and counseling of the family. Before the delivery the likelihood of neonatal interventions, such as initiation of prostaglandin, balloon atrial septostomy, urgent balloon valvuloplasty, or urgent repair of obstructed pulmonary venous return should be considered and discussed with the perinatal team.

Discussion of the plan for delivery and neonatal interventions should be made with all members of the healthcare team. Parents should be offered the opportunity to tour the hospital where delivery is being planned, and to meet with the neonatologists and cardiovascular surgeons prenatally as well as key medical personnel anticipated to be involved following delivery. In cases where fetal cardiac diagnosis requires a change in the delivery location, appointments with the appropriate MFMs and obstetric providers should be facilitated. Resources for learning additional information about the diagnosis should be provided, as well.

Classification of heart defects in prenatal cardiology

Cardiac defects detected in utero can be classified by severity into seven major groups according to the 2014 classification17:

These severity levels are not specific; many diagnoses defined in utero may be seen in 2 or more of the categories based on other complicating factors, physician preference, and the location of the family and their ability to access high risk or tertiary level hospitals. Definition of cardiac structural anomalies must be verified after delivery.

Nonetheless, such a grading system may be helpful for consultation of the family and planning for the treatment of the affected fetus. Examples of these status levels:

Level 1. Occasional premature atrial contractions, small muscular ventricular septal defects, intracardiac echogenic foci, redundant and aneurysmal septum primum, 2 vessel umbilical cord, persistent right umbilical vein with otherwise normal central venous connections, isolated persistent left superior vena cava.

Level 2. Mid-muscular or conoventricular ventricular septal defect, mild pulmonary valve stenosis, mild tricuspid valve regurgitation, atrial bigeminy, absent ductus venosus, large ventricular septal defect, and congenitally corrected transposition of the Great Arteries (CCTGA or LTGA) with no associated defects.

Level 3. Tetralogy of Fallot with mild pulmonic stenosis, dextro-transposition of the great arteries with intact ventricular septum, double outlet right ventricle with mild pulmonic stenosis, hypoplastic left heart syndrome with non-restricted foramen ovale and without tricuspid valve regurgitation, RV > LV disproportion with suspected coarctation of the aorta.

Level 4. Complete AV Canal defect with balanced ventricles and no AV valve regurgitation, truncus arteriosus without stenosis or obstruction, double outlet right ventricle with anterior aorta or pulmonic atresia, pulmonary atresia with intact ventricular septum and tricuspid regurgitation.

Level 5. Tricuspid Atresia, VSD, mild Pulmonic Stenosis and normally related great arteries, Complete AV Canal defect with AV valve regurgitation, dextro-Transposition of the Great Arteries with VSD and PS, Interrupted Aortic Arch with VSD and mild sub-Aortic narrowing, Critical Aortic Stenosis with normal sized Left Ventricle,

Level 6. Tetralogy of Fallot with hypoplastic Pulmonary Arteries and Multiple Aortico-Pulmonary Collaterals, Pulmonary Atresia, intact ventricular septum with hypoplastic TV and RV without tricuspid valve regurgitation and coronary cameral fistulae, Heterotaxy with pulmonary atresia and Totally Anomalous Pulmonary Venous Return below the diaphragm, Hypoplastic Left Heart Syndrome with restricted or intact Foramen Ovale and mitral stenosis and aortic stenosis, Truncus Ateriosus with severe truncal valve insufficiency.

Level 7. Turner’s Syndrome with Hypoplastic Left, Heart Syndrome and severe hydrops, Ebstein’s Malformation with pulmonary Atresia and severe hydrops,Isoimmune anemia with severe hydrops, Heterotaxy Syndrome with complete heart block and hydrops, combined right and left ventricular outflow tract obstruction.

The above examples of the AHA classification system may not be easily understood or adopted by many centers and therefore we propose a much simpler classification scheme.
The Polish classification system has some similarities to the AHA by with some important differences, which are based on the FE findings in 3rd trimester made by a fetal cardiologist (Table 3). In our classification system we stress the urgency of required time to postnatal treatment/intervention and thus we define four groups of fetuses with CHD:

1. **Severest heart defects** - fetuses suffering from CHD whose treatment is not possible prenatally and/or postnatally. (For instance Ebstein’s anomaly with pulmonary hypoplasia due to severe cardiomegaly or critical aortic stenosis with giant left atrium and cardiomegaly.

2. **Severe urgent heart defects** - CHD that requires emergent invasive cardiac intervention by catheterization or surgery within the first 48 hours after birth, (ductal and foramen ovale dependent) such as d-TGA and HLHS with restricted foramen ovale, critical aortic or pulmonary stenosis for balloon valvuloplasty.

3. **Severe planned heart defects** - CHD that is ductal dependent requiring cardiac surgery during the first month after birth (for instance d-TGA with good size foramen ovale), also some cases of coarctation.

4. **Planned heart defects** - isolated CHD which does not typically require cardiac surgery within the first month after birth; (i.e. tetralogy of Fallot, complete atrioventricular canal defects, ventricular septal defects).

Our classification is based on FE findings in the 3rd trimester of pregnancy, preferable at 2-3 weeks prior to delivery.

This is due to relatively rare, however very important, possibility of prenatal progression of hemodynamic disturbances in the given congenital heart defect.

<table>
<thead>
<tr>
<th>Type of CHD</th>
<th>Prenatal</th>
<th>Delivery</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>The severest</td>
<td>For instance: Ebstein +</td>
<td>Vaginal</td>
<td>Conservative approach</td>
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<tr>
<td></td>
<td>cardiomegaly HA/CA &gt; 6</td>
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<td></td>
<td>Lung hypoplasia</td>
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<tr>
<td>Severe urgent</td>
<td>CHD with ducal and formane</td>
<td>Elective</td>
<td>Prostaglandin and</td>
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<tr>
<td></td>
<td>ovale dependent circulation</td>
<td>CS</td>
<td>cardiac catheherisation</td>
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<td></td>
<td>for instance critical AS</td>
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<tr>
<td>Severe planned</td>
<td>For instance d-TGA with good</td>
<td>Vaginal</td>
<td>Prostaglandin and</td>
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<td>surgery</td>
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<tr>
<td>Planned</td>
<td>For instance AVC</td>
<td>Vaginal</td>
<td>Waiting list for</td>
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<td>infants cardiac</td>
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<td></td>
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<td>surgery</td>
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</table>

**Table 1**: Classification of heart defects.

**Table 2**: Fetal echocardiography initial analysis in cardiac center (in addition to intracardiac flows, fetal heart rhythm and assessment of Cardiovascular Profile Score).

**Table 3**: Prenatal cardiac defects classification in Prenatal Cardiology Department Polish Mother’s Hospital Research Institute for 3rd trimester.
WHO IS A FETAL CARDIOLOGIST IN 2015

A physician with background in pediatric cardiology, neonatology, obstetrics (usually maternal-fetal medicine), and who has worked for about five years in a referral center for prenatal cardiology and independently performed and interpreted normal and abnormal fetal echocardiograms. In Poland, such physicians are certified by the Section of Fetal Echocardiography and Prenatal Cardiology Polish Ultrasound Society. In general, as described in the latest AHA statement on fetal cardiology, “only well-trained or experienced pediatric cardiologists, maternal-fetal medicine specialists, obstetricians, or radiologists who have acquired the appropriate knowledge base and skills should supervise and perform fetal echocardiograms.” Beside perform fetal echo prenatal cardiologist can also predict neonatal management and qualify CHD to appropriate group of one of the new CHD classification.

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