Introduction to Fetal Echocardiography

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ABSTRACT

The abstract should be informative and completely self-explanatory, briefly present the topic, state the scope of the experiments, indicate significant data, and point out major findings and conclusions. The abstract should summarize the chapter content in 300 words or less. Standard nomenclature should be used and abbreviations should be avoided. The preferable format should accommodate a description of the study background, methods, results and conclusion.

Keywords: Fetal echocardiography; Anomaly scan; Congenital heart disease; Prenatal diagnostic; Neonatal echocardiography.

Abbreviations: CHD = Congenital Heart Disease; FECG = Fetal Echocardiography; BCEE = Basic Cardiac Echocardiographic Examination; ECEE = Extended Cardiac Echocardiographic Examination; 4CV = Four Chambers View; LVOT = Left Ventricular Outflow Tract, RVOT = Right Ventricular Outflow Tract, 3VTV = Three Vessel and Trachea View.
GENERAL CONSIDERATIONS

Congenital Heart Defects (CHD) are defined as gross structural anomalies of the heart or intrathoracic vessels that are actually or potentially of functional significance. CHD can be diagnosed prenatally by a relatively brief but skilled ultrasound examination of the fetal heart. Fetal Echocardiography (FECG) is a detailed sonographic examination of the fetal heart that is used to identify and characterize fetal cardiac anomalies from the late first trimester to term. FECG is the most important prenatal diagnostic tool for the CHD detection [1-5], which represent the most common malformations in the developing fetus.

Although the examination of the fetal heart is part of the standard fetal scan, FECG is still considered a challenge even for experienced sonographers. The procedure is more difficult to perform than adult echocardiography. First, because of the complexity, small dimensions of the fetal heart and the prenatal circulation particularities regarding the physiology and anatomy. Second, many conditions may impair the fetal heart visibility: maternal obesity, abdominal or uterine scars, myomas, fetal unfavourable position and movements, oligohydramnios etc. Consequently, FECG has not been widely implemented, and the prenatal diagnosis of CHD varies considerably.

FECG has evolved rapidly in the last decades, favoured by the need for a proper prenatal CHD diagnosis and the advances in image quality of the ultrasound equipment. Still, despite the efforts to implement in the prenatal diagnosis field the necessary skills for fetal cardiac scan, the CHD detection of remains at the bottom of the list of identifiable abnormalities in fetal anomaly scan, as less than 50% of major CHD is diagnosed prenatally.

EPIDEMIOLOGY OF CHD

Incidence

CHD are the most common congenital anomalies in the human fetus, affecting about 5 to 12 per 1000 live births and account for the majority of all neonatal and infant deaths due to congenital abnormalities [6,7]. They have become the leading cause of infant mortality, responsible for 42% of infant deaths [8].

Etiology, Risk Factors

CHD may be the result of multiple underlying causes: genetic abnormalities, environmental factors and teratogenic exposures [9-11]. However, over 90% of heart malformations have no known cause and the etiology is multifactorial, as a result of the interaction between hereditary and environmental factors.

The environmental causes are represented by maternal diseases (see the indications below), alcohol and drug intake [12-25] and maternal intense stress [26-27]. In turn, periconceptional folic acid intake which may reduce the risk for CHD [28-29].

**INDICATIONS FOR FECG**

FECG has traditionally been reserved for pregnancies at increased risk for CHD. The following lists of situations were associated with an increased risk for CHD and represent important indications for FECG referral [30]. Usually the patients are referred for FECG at 18 to 24 weeks of gestation. However, in such cases an earlier cardiac should be considered in the late first-trimester between 12 and 14 weeks of gestation.

**Maternal indications for FECG:**

- Family history of CHD: a previous child with CHD increases the risk to 1 in 20 - 1 in 100; previous two children with CHD increases the risk to 1 in 10 - 1 in 20; maternal CHD increases the risk to 1 in 5 - 1 in 20, paternal CHD increases the risk to 1 in 30;
- Familial inherited disorders that involves potential CHD;
- Infectious disease: influenza, rubella, parvovirus B19, coxsackie virus;
- Autoimmune disease: sytemic lupus erythematousus, Sjogren’s, anti-Ro (SSA) / anti-La (SSB);
- Metabolic disorders (e.g. diabetes, phenylketonuria);
- Exposure to drugs and teratogens: halucinogenes (coca and marijuana), lithium, retinoids, phenytoin, trimethadione, valproic acid, coumadin, thalidomide, angiotensin-converting enzyme inhibitors, prostaglandin synthetase inhibitors (e.g. ibuprofen, salicylic acid, indomethacin), sulfalazine, trimethoprim-sulfonamide, metronidazole;
- Advanced maternal age (>35).

**Pregnancy-related indications for FECG**

- Fetal birth defects identified during the current pregnancy;
- Increased first trimester nuchal translucency;
- Chromosomal abnormality and increased combined or maternal serum risk for Down syndrome and other chromosomal defects;
- Arrhythmia;
- Non-immune hydrops;
- Multiple gestation – monochorionic twins;
- In vitro fertilization.
Most research into CHD screening was directed at high-risk groups or selected populations [31-37]. Still, we should be aware that most of the neonates born with CHD are not associated with known risk factors [38-45]. Thus, in order to improve the prenatal detection of CHD, a screening test, which can be offered to all pregnancies, is required.

**IMPORTANCE OF CHD PRENATAL DETECTION**

The importance of prenatal detection of cardiac disorders derives from several major facts:

- Epidemiologic reasons presented above: CHD is the most common severe congenital abnormality, and accounts for over half of the deaths from congenital abnormalities [46].
- CHD results in the most costly hospital admissions for birth defects.
- Early diagnosticof severe CHD provides the parents with the option to terminate the pregnancy [47-48]. This reduces the CHD rate at birth, as half of the prenatally diagnosed CHD are terminated [24].
- CHD accurate diagnostic permits in certain anomalies in utero treatment. Prenatal pharmacological transplacental treatment for fetal arrhythmias has become widely used [49-50]. Also, invasive procedures as laser coagulation of placental an astomoses in twin to twin transfusion syndrome, atrial and ballon valvuloplasties, cardiocentesis etc [51-68] has been described and successfully used.
- In certain situations necessitates rapid intervention at birth, a proper CHD diagnosis allows planning of the appropriate time and place of delivery in a referral centre, which offers a better neonatal and / or surgery care and optimise the chances of survival after postnatal treatment. Thus, prenatal diagnosis of CHD offers potential clinical benefit with regard to infant outcome in certain cardiac lesions, especially in those cases that are likely to require prostaglandin infusion to maintain patency of the ductus arteriosus as transposition of great arteries, aortic coarctation and hypoplastic left heart syndrome [69-75].
- In utero transportation and competition among centers may change the area of referral in favor of the best centers [76].
- Prenatal diagnosis allows counselling of families which are better prepared for the foreseeable management and outcome of the fetus.

**QUALIFICATIONS OF PERSONNEL INVOLVED IN CHD DETECTION**

Performance of the ultrasound scan and interpretation of the findings with regard to the FECG require specialized skills and knowledge. Appropriately trained maternal-fetal medicine specialists, pediatric cardiologists, obstetricians and radiologists with special expertise in fetal imaging may perform FECG.

Guidelines, training requirements and maintenance of competency parameters in the field have been developed [1,77-82] to ensure appropriate knowledge base and skills for the professionals.
EQUIPMENT SPECIFICATIONS

FECG should be conducted with real-time scanners, using a trans abdominal and/or transvaginal approach. The choice of transducer frequency is a trade-off between beam penetration and resolution. With modern equipment, 3- to 5-MHz abdominal transducers allow sufficient penetration in most patients while providing adequate resolution. A lower-frequency transducer (2–2.25 MHz) may be needed to provide adequate penetration for abdominal imaging in an obese patient. During early pregnancy, a 5-MHz abdominal transducer or a 5- to 10-MHz or higher vaginal transducer may provide superior resolution while still allowing adequate penetration [1,82].

Harmonic imaging may provide improved images, especially for patients with increased maternal abdominal wall thickness during the third trimester of pregnancy [83].

DOCUMENTATION

Adequate documentation is essential for high-quality patient care. The standard views of the fetal heart both normal and abnormal should be stored as images or digital videoclips routinely. Images should be labelled with the patient identification and the examination date. An interpretation of the ultrasound scan should be included in the patient’s medical record [1,3].

THE EXAMINATION OF THE FETAL HEART

Settings

For a satisfactory cardiac examination is imperative to obtain adequate visualisation of the heart. Technical limitations (e.g. maternal obesity, abdominal or uterine scars, uterine myomas, oligohydramnios, anterior placentation, prone fetal position, and late gestation) can make a proper detailed heart evaluation very difficult due to acoustic shadowing, especially in late pregnancy. Thus, FECG is commonly performed at 18 to 22 gestational weeks. In some settings, the majority of major CHD may even be recognized at the time of the genetic scan, in late first trimester – see below: First trimester detection of CHD. An early diagnosis is needed especially in high-risk pregnancies, e.g. when increased nuchal translucency thickness is identified.

The sonographer should optimize the image by appropriate adjustment of technical settings, such as image magnification, signal gain, acoustic focus, frequency selection, harmonic imaging, and Doppler settings. Images should be magnified until the heart fills at least a third to half of the display screen. System settings should emphasize a high frame rate, with increased contrast and high resolution. Low persistence, a single acoustic focal zone and a relatively narrow image field should also be used. The cine-loop feature should be used to assist the real-time evaluation of normal cardiac structures, for example, to confirm movement of heart valve leaflets throughout the cardiac cycle [1].
Optimal views of the fetal heart are obtained when the cardiac apex is orientated toward the anterior maternal wall. If the fetal position impairs satisfactory visualisation of the heart anatomy, then it is necessary to await for spontaneous fetal movements, to reschedule the scan, or to undertake such measures as getting the mother to fill or empty her bladder, rotating or tilting her abdomen, or gently manipulating the maternal abdomen to change the fetal relative position.

**Approach to fetal cardiac scanning**

Regarding the specifications of the fetal cardiac examination, several organizations have published formal practice guidelines [84-86]. The fetal cardiac anatomy is evaluated using a sequential segmental analysis of 3 basic areas starting from the atria, then ventricles, and finishing at the great arteries pole and their connections [87-89].

During the examination it is important to positively demonstrate the normality of these fetal heart segments/anatomic planes. The difficulty or impossibility to demonstrate the standard cardiac views should alert for the presence of CHD. No single plane will demonstrate all abnormalities and a comprehensive evaluation of the heart is necessary to accurately describe the vast majority of CHD.

In practice, a segmental analysis initially includes an assessment of fetal right/left orientation. Then each region is evaluated independently, following the direction of blood flow: systemic and pulmonary veins, atria, atrioventricular valves, ventricles and right ventricular outflow tract, semilunar valves, and great arteries. In a systematic manner, right-sided and left-sided structures at each level are evaluated according to their morphology, their positions and connections to segments [90].

**Fetal cardiac sweep**

Five transverse and three longitudinal views of the heart and vasculature are key scanning planes that provide useful information to confirm fetal heart normality or to suspect CHD [1,80,91-96]. The examination should include these planes with their features, noting the abnormalities. The transversal planes include the upper abdomen view, the four-chamber view, the left outflow tract view, the right outflow view and the three-vessel and trachea view (Figure 1, Clip1). The three longitudinal planes are the aortic arch view, the pulmonary arch view and the inferior and superior vena cava plane.
Figure 1: Fetal cardiac sweep. During the standardized transverse scanning planes for fetal echocardiography the sonographer is sweeping the transducer beam in a transverse plane from the level of the four-chamber view towards the fetal neck as presented in the left of the image. By doing so, the following views become apparent: Four-Chamber View (4CV), arterial outflow tracts: Left Ventricular Outflow Tract (LVOT), Right Ventricular Outflow Tract (RVOT) and the Three-Vessel and Trachea View (3VTV).

- Upper abdominal view - abdominal situs.

The normal image of the fetal stomach should be identified on the left side of the fetus and just below the diaphragm. It is important to localize the spine, the descending aorta (on the left side) and inferior vena cava (anteriorly, on the right side) (Figure 2).
Figure 2: Upper abdominal view, used to determine the abdominal situs. This can be made when the Abdominal Circumference (AC) is measured. L, left; R, right; St, stomach; UV, umbilical vein; Dao, descending aorta; IVC, Inferior Vena Cava, Sp, spine.

- **Four-chamber view (4CV)**

  This was the first screening sonographic marker for CHD detection, proposed 30 years ago [97]. The plane is situated the lower half of the fetal chest (Figure 1). In the correct plane, the abdominal organs and the left ventricular outflow tract are not visible, a complete rib is present and the cruxcordis occupies the central portion of the heart (Figure 3). The optimal incidence is the apical insonation (Clip 2), but the information are sometimes well visible from lateral (Clip 3) and back-up views (Clip 4).

  The correct visualization of the 4CV, facilitates the evaluation of the following normal features:

  - **Situs**- levocardia, the heart is left-sided.
  - **Axis**– apex points toward the left side of the fetus at 45 ± 20° (2 standard deviations). Cardiac situs and axis may be abnormal because of cardiac malformations such as isomerism but also the heart may be displaced because of abnormal adjacent structures: diaphragmatic hernia with thoracic herniation of the abdominal organs, or pulmonary hypoplasia, or cystic adenomatoid malformation.
  - **Area** of no more than 1/3 of the area of the thorax.
  - **Atria** - morphology and symmetry. Right and left atria are of the same size and thickness.
It is important to note the atrial situs solitus" with the pulmonary veins entering the left atrium, in order to confirm that the left-sided atrium is indeed the morfo-functional left atrium. The right atrium is anteror and the left posterior. Lack of the pulmonary venous connections to the back of the left atrium suggests anomalous venous return.

- **Ventricles** - morphology of the. Right and left ventricles are of the same width and contract equally.

However, in later gestation, the right ventricle becomes slightly larger and could be confused with right ventricular dilation secondary to CHD [23,24]. Reference diameters and Z-scores for the atrio-ventricular valve annuli, cavitary lengths and thickness of the ventricular walls are available [98-99].

The right ventricle is the most anterior structure and closest to the anterior chest wall. The main imagistic structural particularities of the ventricles are that the insertion of the tricuspid valve along the septum is more apical than the insertion of the mitral valve, the right ventricular apex should contain the moderator band, causing the apex to appear “filled in”, and the apex of the heart is formed by the left ventricle. Also, there are slight differences in lining of the two ventricles with more coarse lining of the right ventricle due to a coarser trabeculation.

- **Ventricular septum**. The integrity is better evaluated from a lateral cardiac incidence, perpendicular to the septum.

In case of an apical insonation of the fetal heart, or in case of insufficient lateral resolution small septal defects may be missed. On the other hand, in similar conditions the ventricular septum will appear thinner than its actual thickness and an area of dropout may been seen in the membranous portion, an artefact that can be confused with a ventricular septal defect.

- **Atrial septum** primumpresence, at the crux of the heart. It is important to note that there is a small amount of offsetting towards the apex of the tricuspid valve septal hinge point relative to the mitral valve.

- **Foramen ovale occupying** about one third of the atrial septum and the flap bulging toward the left atrium.

- **Atrioventricular valves** with their insertion and opening.

- **Pericardial effusion** should be absent, or less than 2-4 mm.

- **Heart rate and the regularity** of the rhythm has to be assessed.

- **Coronary sinus**, may be optionally demonstrated by fine sweeping posteriorly from the 4CV.

Occasionally, echogenic lines may be seen inside the right atrium: the **Eustachian valve** located close to the inferior vena cava and the **Chiari network** that appears as lace-like strands attached to the Eustachian valve and crista terminalis, as a result of the incomplete reabsorption of the septum spurium (1% of patients).
From the extensive description above of this important cardiac plane, it is evident that the 4CV is much more than a simple count of cardiac chambers. It offers a great deal of information regarding the heart chambers, but specific types of abnormalities (e.g. great vessels malformations) may not be evident from the 4CV plane alone [100]. The reason for this is because the 4CV plane is situated at the level of the ventricles and atria, which is caudally from the levels of the outflow / great arteries tracts. Thus, the CHD detection rates can be optimized by visualizing routinely the outflow tract views which are described below.

The sonographic technique to visualize all these cardiac planes was described as the sweep technique [101-102] and involves sweeping the transducer beam in a transverse plane from the 4CV level towards the fetal neck. By doing so, progressively the following views are obtained: 4CV, left ventricular outflow tract, right ventricular outflow tract and three-vessel and trachea view.

**Figure 3:** Schematic presentation of the four-chamber view and sonographic assessment in duplex mode (Gray-scale and color Doppler) in Diastole (D) and Systole (S). Note that the mitral valve has a slightly more apical insertion at the crux cordis. The atrio-ventricular Doppler flow is red because of the direction toward the direction during diastole. When the atrio-ventricular are closed, during systole, the atrio-ventricular flow is absent.

**LV, Left Ventricle; RV, Right Ventricle; LA, Left Atrium; RA, Right Atrium; mb, Moderator Band; mv, Mitral Valve; tv, Tricuspid Valve; vi, Valve Insertion; pv, Pulmonary Veins; sp, Septum Primum; fo, Foramen Ovale, DAO, Descendent Aorta; Sp, Spine.**
• **Left ventricular outflow tract (LVOT)– five-chamber view / long axis view;**

This view is found cranially from the 4CV plane and directed towards the fetal right shoulder. Therefore, from the standard 4CV the transducer is angulated towards the fetal head and rotated clockwise. In this view, along the cardiac chambers, the aortic root (ascending aorta) appears *arising from the posterior* (left) ventricle, giving the aspect of five cardiac chambers (Figure 4, Clip 5).

In the correct plane the aortic root is clearly visible and the *septo-aortic continuity* should be confirmed: the ventricular septum forms a continuous line with the aortic wall without any gap. The continuity between the left ventricle and the aorta resembles the ballerina feet aspect. A break in this line may indicate the presence of a ventricular *septal defect*.

A sufficient length of the ascending aorta that leaves the left ventricle should be visualized in order to confirm its *long sweeping course* and the *absence of proximal transversal branching*. These aspects differentiate the ascending aorta from the main pulmonary artery that may be abnormally connected to the left ventricle.

Also, a sufficient length of the ascending aorta is necessary to confirm its uniform *diameter* and the *direction* towards the right shoulder, typical for a normal crossing of the great vessels.

The *aortic valve cusps* opening freely, disappearing in systole and not thickened should be noted, in order to rule out a dysplastic aspect.

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**Figure 4:** Schematic presentation of left ventricular outflow tract view and sonographic assessment in duplex mode (Gray-scale and color Doppler) in Diastole (D) and Systole (S). Note the continuity of the Ventricular Septum (VS) with the aortic wall, namely the Septo-Aortic Continuity (SAC). When the Aortic Valve (AoV) is closed, during dystole, the aortic flow is absent. LV, left Ventricle; RV, Right Ventricle; LA, Left Atrium; RA, Right Atrium; Ao, Aorta.
- Right Ventricular Outflow Tract (RVOT) and short axis view;

This view is found cranially from the LVOT plane, therefore during the cardiac sweep it is necessary to slide and angle the transducer upwards to the fetal neck (Figure 1).

The main pulmonary artery root arises from the anterior right ventricle and has a characteristic straight course back towards the spine. Its course is short, soon branching into a large vessel, the ductus arteriosus directed straight posteriorly toward the descending aorta as an extension of the main artery, and the smaller pulmonary arteries directed laterally (Figure 5, Clips 6,7).

Using a small rotational adjustment, the short-axis view is demonstrated, with the anterior right ventricle and pulmonary artery emergence, course and branching. In the middle of the heart, the transversal view of the aortic root and valves. This is the best section to demonstrate the pulmonary valve.

The pulmonary valve cusps should be seen, with a similar aspect as described for the aortic valves.

RVOT and LVOT views should be interpreted in conjunction by scanning back and forth between the two planes. This helps to establish two important features. First, the spatial relationship at the crossing of the two arterial arches that is almost at a right angle. Second, regarding their relative sizes, the two ventricular outflow tracts have approximately equal width, with the pulmonary artery being slightly larger, especially in late pregnancy.

![Figure 5: Schematic presentation of right ventricular outflow tract view (RVOT) and sonographic assessment in duplex mode (Gray-scale and color Doppler). Note that the short axis view offers a better image of the connection between the right ventricle and the main pulmonary artery. RV, Right Ventricle; MPA, Main Pulmonary Artery; DA, Ductus Arteriosus; RPA, Right Pulmonary Artery; SVC, Superior Vena Cava; DAO, Descending Aorta; pv, Pulmonary Valve.](image-url)
• Three-Vessel and Trachea View (3VTV);

The view is obtain sliding the ultrasound beam cranially in a transverse oblique plane in the upper thorax during the sweep technique described above. Also, to have a proper image of both arches, the scanning plane has to be slightly tilted down towards the fetal left side, because the aortic arch rises higher than the ductal arch (Figures 1, 6).

In this plane three vessels are visualized (Figure 6, Clip 8): the Superior Vena Cava (SVC) and the two arterial arches, namely the aortic arch and the ductal arch. The ductal arch lies to the left of the aortic arch and continues the more anterior vessel, pulmonary artery (Figure 10). The normal alignment of the vessels evidentiate the arterial ductus at the fetal left, then aorta and superior vena cava in the right side. The aortic arch course is now changed, toward the left of the spine, in front of the trachea. Both the aortic arch and the ductus arteriosus are visualized forming a „V“-shaped confluence at the descending aorta, on the left side of the spine. These view can be used to detect aortic arch abnormalities (right aortic arch, interrupted aortic arch) and for evaluation of the thymus [103-105].

The relative sizes of the two arterial arches should be approximately equal. In the second half of pregnancy the ductal arch tends to be a little bigger than the aortic arc. However, marked discrepancies in the two vessel's size may indicate the presence of arterial stenosis or aortic coarctation.

Figure 6: Schematic presentation Three Vessel and Trachea View (3VTV) and sonographic assessment in duplex mode (Gray-scale and color Doppler). In a slightly cranial plane of the fetal chest, we note that the ductus arteriosus is fading, as the aorta is still well-seen. This is because the aortic arch rises higher than the ductal arch, and this view was proposed by some authors as the final plane of the cardiac sweep – the aortic arch axial plane. DA, Ductus Arteriosus; Ao, Aorta; SVC, Superior Vena Cava; DAO, Descending Aorta, T, Trachea.
• **Aortic arch view;**

The aortic and ductal arches can be evaluated in both axial and longitudinal views. The axial view has the advantage that the two arterial arches can be directly compared in size.

The aortic arch view is obtained in a longitudinal thoracic sectional plane from anterior right to posterior left of the fetal chest, where the descendent aorta lies.

The normal aortic arch has the following features (Clip 9, Figure 7): its origin from the middle of the heat, the typical “hook” shape in longitudinal view, and the longitudinal branching form the aortic cross. The three vessels emerging are the neck vessels: brachiocephalic trunk, left common carotid artery and left subclavian artery.

![Figure 7: Aortic arch longitudinal view, branching and continuity with the descending aorta.](image)

• **Ductal arch view**

The ductal arch view is obtained in a longitudinal thoracic sectional plane aligned with the right ventricular outflow tract and main pulmonary artery. In this view, the main pulmonary artery, the ductus arteriosus and the communication with the descending aorta, forms a flattened arch with the typical hockey stick” shape and without neck vessels arising from it.
Figure 8: Ductal and Aortic arches in longitudinal view. The differences mentioned in the text are obvious, regarding the origin, curvature and branching.

- Superior and inferior vena cava views / caval long-axis view / bicaval view;

The caval long-axis view is obtained in a longitudinal plane on the right of the spine, in line with the superior and inferior vena cava draining at the back of the right atrium (Clip 14, Figure 9).

Figure 9: Bicaval view. IVC, Inferior Vena Cava; SVC, Superior Vena Cava; RA, Right Atrium.
**Doppler Imaging**

In all the views mentioned above, *color Doppler* sonography should accompany the greyscale evaluation, to evaluate the normal flows in the respective cardio-vascular structures [106-108]. All the figures above present the cardiac features in both Color and gray-scale modes, in duplex mode.

In addition, *pulsed Doppler* sonography should be used as an adjunct to evaluate the blood flows at the level of atrioventricular valves, semilunar valves and optionally in ductus venosus, umbilical vein and artery. It is required in case of cardiac rhythm disturbances or cardio-vascular suspected abnormalities at the greyscale or color Doppler assessment.

B-flow, Power Doppler and High Definition Power Flow Doppler are not routinely used, although in some cases they may be useful, because each have unique capabilities that allow greater sensitivity in imaging cardio-vascular blood flow.

**Heart Rate and Rhythm Assessment**

Fetal Heart Rate (FHR) and rhythm should be evaluated based on the cardiac cycle length evaluated in pulsed Doppler or M-mode interrogation. The normal FHR in the second and third trimester is 120 to 180 beats per minute.

Abnormal rate and/or rhythm should trigger supplementary investigations to determine the underlying mechanism. M-mode and/or Doppler should evaluate simultaneously atrial and ventricular contraction, mitral and aortic outflows or superior vena cava and ascending aorta flows.

**Cardiac Biometry**

Cardio-vascular measurements should be considered in case of suspected structural or functional anomalies [1,77]. Specific measurements can be performed in 2D or M-mode images: fetal heart area and axis and cardiothoracic ratio, cardiac chambers, atrio-ventricular valves annulus in diastole, aortic and pulmonary valve annulus in systole, arterial arches diameters, ventricular walls and interventricular septum thickness.

Normal fetal ranges have been published as nomograms and z scores that are correlated with the gestational age or fetal biometry [98, 99,109-114].

**Cardiac Function Assessment**

Cardiac functional assessment is optional but should be considered for suspected structural or functional cardiac anomalies [1,77].

A certain degree of *qualitative* evaluation of the heart function should be part of standard scanning, as cardiomegaly, atrioventricular valve regurgitation, and hydrops should be recognized routinely.
The quantitative assessment of heart function include the study of myocardial movement such as tissue Doppler, myocardial / ventricular strain, strain rate imaging, fractional shortening and the myocardial performance index [115-121].

**Required Versus Optional and Basic versus Extended**

Previous guidelines stated a basic cardiac evaluation, included in the midtrimester standard fetal anomaly scan and an extended cardiac examination, used in selected pregnancies with increased risk for CHD.

The *basic cardiac screening examination* was basically the interpretation of the 4CV of the fetal heart. Evaluation of normal situs, area, axis, rhythm, pericardial effusion.

The extended basic cardiac examination added the ventricular outflow tracts views and optionally three-vessel and trachea view, in order to increase the detection rates for major CHD, especially regarding the conotruncal anomalies [122-123].

However, recent revised and updated guidelines and recommendations from several professional bodies [1, 124-127] plead for the routinely screening evaluation of the 4CV as well as the outflow tract views based on strong medical evidence regarding the prenatal detection of CHD [39, 95, 103, 128-131].

**Complementary Imaging Strategies. 4D Spatiotemporal Image Correlation (STIC)**

Volume datasets obtained with 4D STIC ultrasonography can be described as virtual blocks of pathological specimens of beating hearts, where all the anatomical information is contained inside. Various rendering techniques can be used to „cut” the block at any level, and interpret the structural and functional information of the fetal heart. Thus, an advantage offered by 4D sonography is to give access to virtual planes not available for direct visualization with 2D technique.

During the classic two-dimensional scan the operator has to find the key planes of the fetal heart and to realize a mental reconstruction of the spatial relationships between the cardiovascular structures and connections. 3/4D technology in conjunction with color / power Doppler, inversion or B-flow modes facilitates thereconstruction process using different rendering / display modalities of the volume datasets.

This technology have the potential to decrease dependency on sonographer skills and experience, and knowing the fact that the FECG is the most difficult area of the fetal anomaly scan, it might be very useful for the settings with less experienced sonographers. Also, volume datasets from of fetal heart can be acquired in a high proportion of cases by properly trained non-expert operators and sent to an expert in FECG for offline evaluation via telemedicine. This may increase the detection rate of CHD.
The use of volumetric techniques in fetal heart assessment is optional and is not routinely used. However, in selected cases, it may offer important information. A valuable contribution of 4D fetal FECG was demonstrated in cases of complex heart defects, where a comprehensive evaluation may aid the diagnosis [132-135]. Another important application is the evaluation of cardiac function and quantification of fetal hemodynamic parameters, such as cardiac output [136-137].

**Figure 10:** STIC acquisition of the arterial arches with high definition direction power Doppler applied (A), and the manipulation of the volume in order to visualize the aortic arch (B) and the ductal arch and the connection with the descending aorta (C).

**Figure 11:** STIC acquisition of the arterial arches in conjunction with power Doppler. After the manipulation of the volume the aortic arch (A) and the ductal arch (B) are well seen.
Figure 12: STIC acquisition of the arterial arches in conjunction with B-flow. After the manipulation of the volume the aortic arch (A), the ductal arch (B) and the confluence of the two arterial arches (C) are well seen.

EFFICIENCY

Structural cardiac anomalies are the most frequent congenital malformations but also among the abnormalities most frequently missed at the prenatal anomaly scan. Isolated CHD are among the most challenging regarding the prenatal diagnosis [138-140], but are particularly important due to the need to prepare special conditions for delivery.

The efficiency of the cardiac scan is reported with great variation across different populations and registries [141], depending on the scanning protocol, examiner experience, equipment quality, gestational age, and the scanning conditions that may impair the proper visualization (maternal obesity, abdominal scars, amniotic fluid volume and fetal position) [142-143].

No other fetal organ is examined in so many modalities and with as many ultrasound techniques as the fetal heart including high resolution 2D imaging, M-mode, spectral, color, power, high definition Doppler, B flow, tissue Doppler, 3/4D STIC. Still, the screening programs, especially when limited to the study of the 4CV, reported low detection rates for CHD [90]. During the last decades, five scan protocols for fetal CHD diagnosis have been commonly used: Four Chamber View (4CV), 4CV + Outflow Tracts View (OTV) / Three Vessels and Trachea View (3VTV), 4CV+OTV+3TV, extended cardiac echography examination and 4D spatio-temporal image correlation (STIC). 4CV alone detects up to 77% of prenatally developed CHD, while OTV increases prenatal detection rate between 83-92% [144].
FIRST TRIMESTER (FT) EVALUATION OF THE FETAL HEART

The fetal heart is completely formed by eight gestational weeks. The fact that CHD heart defects happen during this crucial first eight weeks of the baby’s development, triggered studies aimed to investigate if a cardiac scan is feasible at the genetic scan, during 11-13 weeks of gestation.

Targeted Population

Some consider early screening for heart anomalies in unselected population not advisable yet due to high number of false negatives and costs in term of time and machines [145]. Completion of the heart visualization protocol differ between unselected population and referral population (47.5% vs 76.9%) [40]. Because the early heart evaluation is not easy, but the operator is constrained to finish the protocol of heart investigation when dealing with high-risk pregnancy for cardiac defects.

Timing

With improved technology it has become feasible to obtain images of the fetal heart in the FT, with visualization of the four heart chambers and outflow tracts of the great vessels from as early as 10 weeks [146-147].

However, the rate of a complete cardiac evaluation improves as gestational age increases: between 20% at 11 gestational weeks to 92% at 13 gestational weeks with a TV probe [148]. Other authors [35,149] reported similar success rates with 100% of visualization at 14-15 gestational weeks. The end-point is that comprehensive visualization of fetal cardiac anatomy is already possible at the end of the FT [33,41,42,152].

Imaging Technique

Grey scale is the basis of a reliable fetal cardiac scan in the ST. However, advanced sonographic techniques offer supplementary important information. For example, Doppler ultrasonography can imagine and measure blood flow velocity or identify abnormal flow patterns across valves and within heart chambers. M-mode echocardiography offers an important method for analyzing cardiac dysrhythmias, suspected ventricular dysfunction, and abnormal wall thickness. The routine use of color Doppler in low-risk populations remains controversial. For safety reasons routine use of pulsed color Doppler is advised against in the FT [153], although during the FT genetic and anomaly scan tricuspid and ductus venosus flow evaluations are routinely performed in the prenatal units and proved their importance for many years [36,144,154-160].

Nevertheless, the use of color Doppler in FT is necessary because of the low discrimination of the heart structures in gray-scale mode. Studies have demonstrated that colors improve the visualization of normal cardiac structures [161] and early detection of conotruncal anomalies [162]. Therefore, we should keep in mind to using Doppler technique always respecting the ALARA principle (As Low As Reasonably Achievable).
The examination protocol;

Depending on the operator skills and the scanning equipment, the FT protocol is similar to the second trimester [163-164] (Figure 13), using 2D gray-scale and Doppler techniques. Recent studies have shown that the use of 4D-STIC in the FT is feasible and is likely to improve the detection of CHD in expert hands [165-168] (109-112). It has been reported that the overall performance of pooled sensitivities of STIC, extended cardiac examination and the cardiac sweep (4CV + OTV + 3VTV) were around 0.90, which was significantly higher than that of 4CV+ OTV or 3VTV and 4CV alone [169].

A: Four-chamber view plane: gray-scale imaging shows four-chambered heart, crux cordis and one pulmonary vein entering left atrium; color Doppler imaging shows equal atrioventricular flow and no flow between ventricles (intact ventricular septum).

B: Left ventricular outflow tract plane: gray-scale imaging shows emergence of left ventricular outflow tract and septo-aortic continuity; color Doppler imaging shows the aortic flow.

C: Crossing of the great vessels.

D: Three vessels and trachea view plane: gray-scale imaging shows confluence of arterial arches on left of spine; color Doppler imaging shows confluence of arterial arches and normal direction and equal flow in both arches.

Figure 13: Cardiac sweep in the first trimester in duplex mode: gray-scale (left of each pair of images) and color Doppler (on the right).
In all the planes we note that the information provided by color Doppler is more confident regarding the fetal heart structure and function, especially in the upper thorax (C,D), where the cardio-vascular structures are smaller and difficult to imagine on gray-scale mode.

**Performance of the FT Cardiac Scan**

The detection rate of major CHD at the 11 to 13+6-week scan varies widely (5.6% - 90%) depending on the protocol used, studied population (high or low-risk), scan route (TV, TA or both), definition and prevalence of major CHD. There are reports of high early detection rates for major CHD even in unselected or low risk population 80% - 90% especially when using an extended standardized heart screening protocol [41,43,170-172].

A small number of studies report real accuracy of FT cardiac scan because of the lack of appropriate verification test. A systematic review of the literature [173] reported a pooled sensitivity and specificity were 85% (95% CI, 78– 90%) and 99% (95% CI, 98– 100%), respectively. Therefore, FT cardiac scan can diagnose major CHD with high accuracy a normal heart with reasonable accuracy.

We should be aware that normal results from echocardiographic examinations at any time of pregnancy do not exclude CHD, because some cardiac lesions may evolve in utero as gestational age advances or even occur later during pregnancy: hypoplastic left heart syndrome, coarctation of aorta, endocardial fibroelastosis, pulmonary stenosis, tetralogy of Fallot, cardiomiophaty or cardiac tumors [35,42,174-176]. Other heart defects such can evolve even after birth. Ventricular septal defects were the most missed lesions during prenatal echocardiographic evaluation because of limited resolution, the small size of the lesion and low flow velocities in the FT. It is very important to remember that a normal cardiac scan in the FT is not a replacement for the second trimester echocardiography.

**The Role of Nuchal Translucency (NT) Measurement, Ductus Venosus and Tricuspid Flows**

Increased NT was associated with cardiac dysfunction and abnormalities, even in chromosomally normal fetuses, but not obviously related to any particular type of cardiac anomaly [150,177-180]. The prevalence of CHD when NT is the 95th percentile is up to 20% [170,181]. and for NT ≥99th percentile is about six times higher than in unselected population [179,182-183]. However, it has been shown that NT measurement is not a reliable screening test for CHD during FT. Low detection rates for CHD (around 15%) are reported in studies where NT is measured in unselected or low-risk populations [184-185] and when septated hygromas cases are excluded [186].

The performance of early screening for CHD achieved by measurement of fetal NT is improved by the assessment of ductus venosus and tricuspid valve flow pattern. In fetuses with enlarged NT (above 95 centile) and absent or reversed a-wave in DV flow the risk for major CHD is tripled [158].
The finding of reversed a-wave in chromosomally normal fetuses increases by almost 10 times the risk of CHD, with a predominance of right-heart anomalies regardless of the measurement of NT [179].

Also, chromosomally normal fetuses with tricuspid valve regurgitation have an 8-fold increased risk for CHD [157, 187].

Figure 14: Normal ductus venosus (A) and tricuspid (B) flows. Reversed a-wave (C) and tricuspid regurgitation (D) in fetus with major CHD, atrio-ventricular septal defect (E).

References


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