

# Two-dimensional fetal echocardiography: where we are

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## Summary

**Congenital Heart Disease (CHD) is the most common severe congenital abnormality in the newborn and the cause of over half the deaths from congenital anomalies in childhood. Prenatal diagnosis, possible as early as 15 weeks of gestation, allows physicians and families the greatest number of therapeutic options, and can improve the postnatal outcome. There are several potential indications for performing such examination. Evaluation of the heart in the setting of restricted fetal growth or fetal distress is often recommended. Whenever extracardiac anomalies are detected during fetal ultrasound examination or in presence of chromosomal abnormalities detected with amniocentesis, cardiac assessment is mandatory. The test should also be performed as part of the assessment of fetal arrhythmias. Finally, whenever congenital heart disease is suspected for other reasons, such as maternal exposure to teratogenic substances or a parental history of previous children with congenital lesions, the examination should be considered.**

**The performance of a fetal echocardiogram requires experience and a systematic approach. Guidelines for training have been formulated, and only qualified individuals should perform this highly specialized examination. A description of the techniques of heart examination is presented below.**

*KEY WORDS: fetal, echocardiography, congenital, cardiopathy.*

## Introduction

Considerable advances in ultrasound technology and a close collaboration between the specialties of paediatric

cardiology and fetal medicine have resulted over the last three decades in the increasing ability to diagnose congenital heart disease before birth.

CHD at present time show an incidence of about 4-13 per 1000 live births (1-3), thus representing one of the most frequently detected congenital malformations (4). They are responsible for about half of the deaths caused by lethal malformations during postnatal age (5).

The percent probabilities to recognize CC in prenatal age are quite varying (6), being influenced by several factors such as the experience of the operator, maternal obesity, the type of transducer used, gestational age, the volume of amniotic liquid and the fetal position (7, 8).

Echography proves to be a well established exam overall, non-invasive, simple to carry out, and shows sensitivity and specificity which are strictly dependent on the capacities of the individual operator (9-11). The diagnosis of CHD during prenatal age should never be performed without a careful exploration of other eventual genetic and polymalformative syndromes (that may concern other organs). Importantly, outcomes for specific lesions may differ as extracardiac abnormalities and chromosomal defects may alter the prognosis of otherwise straightforward cardiac lesions such as tetralogy of Fallot.

## Timing of fetal echocardiography

The echocardiographic study of the fetal heart is optimally performed between 18 and 22 weeks of gestational age, a time window that enables the evaluation of most details of fetal cardiac anatomy. Some alterations may be identified starting from the first trimester of gestation, occasionally with the aid of the transvaginal probe, particularly when an increased thickness of nuchal translucency is detected during the screening for chromosomal abnormalities (12-15) conducted between the 11<sup>th</sup> + 6 days and the 14<sup>th</sup> week of gestation. The use of the color Doppler is of utmost importance in the early echocardiography as it helps in the recognition of the large vessels.

The undertaking of fetal heart screening towards mid second trimester can be very useful in addition to a previous fetal echocardiography performed a few weeks earlier, since several cardiac pathologies tend to show a later onset (for example, ventricular hypoplasia due to obstructive process).

If fetal echocardiography examination conducted in the first trimester or during pre-morphologic age (16-18 weeks of gestation) cannot exclude the development of late onset pathologies, the undertaking of such exam around the 28<sup>th</sup>-30<sup>th</sup> week of gestation can face some obstacles provided by the fetal position and the rib bones.

### Indications for performing the examination

The indications for performing such examination can be divided into two main categories: maternal and fetal (16).

#### Maternal indications

- Familiar anamnesis positive for CHD.
- Metabolic disorders such as Insulin-Dependent Diabetes Mellitus (IDDM), especially if not compensated during pregnancy, and phenylketonuria, due to fetal exposition during organogenesis to values of maternal phenylalanine > 15mg/dL.
- Exposition to teratogenic agents such as: steroids, anticonvulsants, alcohol, lithium, but most of all derivatives of vitamin A (retinoic acid and derivatives).
- Exposition to inhibitors of prostaglandin synthesis (ibuprofen, salicylic acid, indometacine).
- Infections from rubella, CMV, Coxsackie e Parvovirus B19.
- Autoimmune diseases such as LES and Sjogren's syndrome.
- Hereditary familiar disorders such as, for example, Marfan's syndrome.
- Medically Assisted Reproduction (PMA).

#### Fetal indications

- Morphostructural exam in another gestation age which suggests the presence of factors possibly indicating CC.
- The presence of other alterations which refer to other fetal organs and/or structures.
- Chromosomal abnormalities.
- Cardiac arrhythmia (persistent tachycardia, persistent bradycardia, persistent irregular fetal heart rate).
- Fetal hydrops.
- Monocorial twin pregnancy and suspected TTTS (*Twin to Twin Transfusion Syndrome*).
- Increased values of nuchal translucency during the first trimester of gestation (>3.5 mm).
- Early fetal growth restriction (that appears in the II trimester); in these cases, CC are most frequently associated with aneuploidy or with complex syndromes.

### Technical execution of fetal echocardiography

The successful outcome of the procedure depends upon the ability to establish a correct frame for the heart using the appropriate ecographic window. Since the fetal position is not fixed in time, but is rather continuously moving during the echographic examination, it is important to have an optimal knowledge of the cardiac scans and of their characteristics.

#### The cardiac situs

The first step to be undertaken in the study of fetal heart is certainly the establishment of the "*Situs*" and, thus, of the position of the heart and of its relationships with the superior abdominal organs. Under physiological conditions, we often refer to "*Situs Solitus*" if the apex of the

heart is on the left (levocardia), the gastric air bubble are on the left side, the abdominal aorta is posterior and on the left of the spinal chord, the inferior vena cava is anterior and on the right of the spinal chord, the liver on the right.

Whenever the cardiac apex is to facing right then we use the term *Destrocardia*; the term *Destroposition* refers to a heart that is displaced from its normal position due to extracardiac reasons (for example, diaphragmatic hernia, pulmonary congenital cystic adenomatoid malformation, pleuric effusion, etc.).

We use the term "*Situs Inversus*" when all the thoracic and abdominal organs are positioned in a perfect mirror image reversal of the normal situs solitus (apex and the gastric air bubble are on the right, the abdominal aorta is posterior and on the right of the spinal chord, the inferior vena cava is anterior and on the left of the spinal chord, the liver on the left).

#### Cardiac axis (Fig. 1)

Besides the position, particular attention needs to be placed also on the cardiac axis, which represents an element that can be easily evaluated even though the projection of 4 chambers cannot be visualized clearly (17). The cardiac axis originates from the axis passing through the interventricular septum and the anteroposterior axis of the abdomen, passing from the breastbone to the spinal chord. The angle defined by these two lines is around 45° (normal range 22°-75°). Deviations towards right or left from this normal range represent signs of cardiac anomalies, that can affect particularly the outflow tracts.

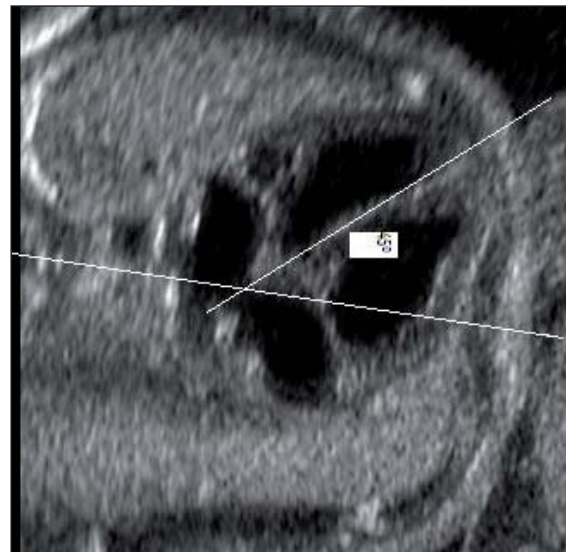


Figure 1

#### Projection of four cardiac chambers (Fig. 2)

The exam of the fetal heart proceeds further on with the projection of the four cardiac chambers (18, 19) (five-chambers in the case the image comprises also the origin of the ascending aorta).

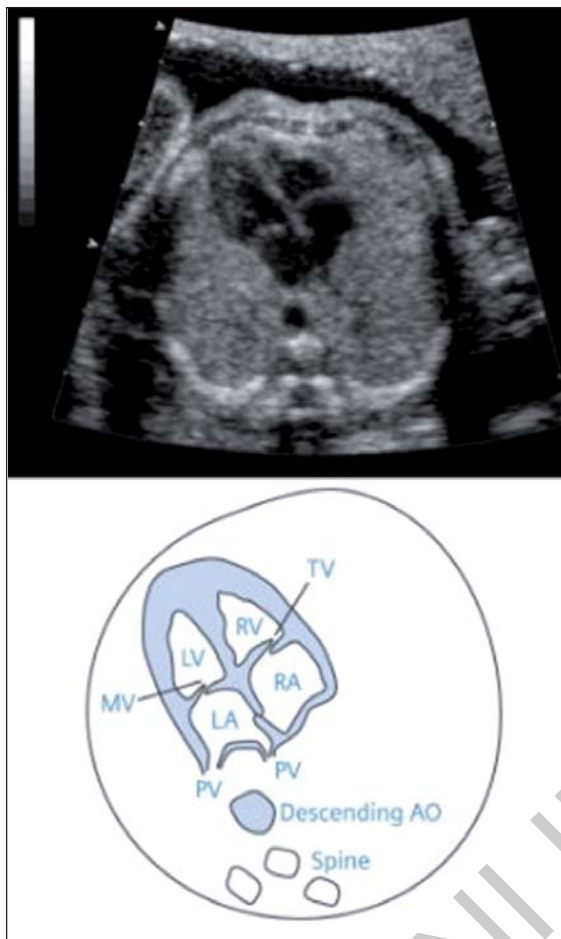


Figure 2

Under physiological conditions, the heart tends to fill almost a third of the fetal thorax with the right apex heading towards the left anterior thoracic wall.

The atrial chambers appear, under physiological conditions, similar for dimensions. The opening of the oval foramen is towards the left atrium since the blood flux is carried through it, always from right towards left.

The thin, crescent fold of endocardium, known as the septum primum should be identified. The pulmonary veins should be visualized on the posterior wall of the left atrium.

Both the ventricles appear similar in terms of dimensions. Sometimes it is possible to recognize a slight ventricular disproportion as a normal variant. Whenever such disproportion should be more evident then it most probably refers to pathologic conditions such as hypoplastic left heart syndrome and aortic coarctation (20, 21).

The four chambers projection is also needed to identify the landmarks that allow the recognition of the atriums and the ventricles.

Two distinct atrio-ventricular (A-V) valves (the tricuspid on the right and the mitral on the left) should be seen opening separately and freely. The tricuspid valve is normally placed in a more apical position with respect to the mitral valve (normal valve *off-set* distance). An unusual

alignment of the atrioventricular valves can be a key element in the identification of atrial-ventricular septal defects (AVSD).

The interventricular septum should be examined entirely in all of its length. The evaluation of ventricular septal defects (VSD) can be quite difficult when the angle of insonation of the transducer is directed parallel towards the ventricular wall. In such case, a false positive defect can be suspected due to the appearance of an acoustic shadow. Minor septal defects (1-2 mm) can be hard to recognize if the echographic machine is not equipped with a sufficient degree of lateral resolution, especially if the dimension and the position of the fetus are not favourable. In synthesis, the four chamber projection is the most frequently used scan and employed in 90% of cases. However it shows a low sensitivity for the recognition of CHD (22).

Such a scan can be able to identify pathologic conditions such as ventricular hypoplasia, defects of the atrio-ventricular septum or atresia to one or both A-V valves, nonetheless we must regard as absolutely unsatisfactory its capacity to recognize other pathologic conditions such as: subaortic ventricular defects, Fallo's tetralogy, transposition of large vessels, double outlet right ventricle and many other complex defects. For this reason it appears clear the need to use other types of scan of the fetal heart that can show the relationship between fetal heart and large vessels.

#### *Projection of the cardiac outflow tracts*

The evaluation of the outflow tracts can increase the percent probability to identify cardiac malformations with respect to the diagnosis provided by the four cardiac chamber projection alone (23, 24). Under physiological conditions, the large vessels present approximately the same dimensions, crossing over at their origins when they exit their respective ventricular chambers.

The outflow tracts can usually be obtained by moving the transducer towards the fetal head, starting from the four chamber projection, when the interventricular septum is tangential to the ultrasonic beam.

Another method to evaluate the outflow tracts has also been described for the fetus when the interventricular septum is perpendicular to the ultrasonic beam (25). This approach requires a four cardiac chamber projection where the probe is rotated in order to visualize the left ventricular outflow tract. Once obtained this position, the transducer is shifted in the cephalic sense in order to observe the outflow tract of the pulmonary artery on a plane which is perpendicular to the aorta.

Yoo et al. have further described the "three vessel view" (Fig. 3) to evaluate the pulmonary artery, the ascending aorta and the superior vena cava taking into account their relative dimensions and relationships (26, 27).

#### *Projection of the left ventricular outflow tract*

The projection of the left ventricular outflow tract (Fig. 4) confirms the presence of a large vessel that originates from the left ventricle and is needed to ascertain the continuity from the anterior wall of the aorta to the ventricular septum.





Figure 3



Figure 5



Figure 4

The aortic valve moves freely without showing any sign of thickness within its context.

The identification of the aorta should be done along with the evaluation of the aortic arch that provides the origin for three vessels: the right brachiocephalic artery, the left common carotid artery, the left subclavian artery.

The aortic arch then folds down on the left of the spinal chord (Fig. 5) showing a typical “candy-cane” shaped structure, which then becomes the descending aorta that is placed in a slightly more medial position.

The projection of the left ventricular efflux tract can help in the identification of defects arising in the ventricular septum and of anomalies affecting the trunks, that are not recognizable with a simple four cardiac chamber projection.

#### Projection of the right ventricular outflow tract

The right ventricular efflux tract shows the presence of a large vessel that departs from the morphologically right ventricle. The pulmonary artery, normally, originates from the right ventricle and runs on the left of the more posterior ascending aorta. The former is slightly larger than the aorta during fetal life and crosses the ascending aorta at an angle of about 70° just above its origin. The valves of the pulmonary artery move freely and

should not be thickened. Such a projection can be confirmed when the pulmonary artery at its distal extremity shows a bifurcation of its left and right branches, even though such a bifurcation can usually be observed only in a few fetal positions.

#### Echocardiography for the evaluation of the fetal heart rate (FHR)

Echocardiography is very useful for the diagnosis and the management of fetal arrhythmia. The fetal ECG can be obtained by combining the use of 2D imaging and the M-mode registration during the atrial and ventricular contractions.

The incidence of fetal arrhythmia is 1-2%, 90% of which are due to isolated atrial or ventricular ectopic beats, and are in most cases of benign origin.

More severe types of arrhythmia include:

- supraventricular tachycardia;
- atrial fibrillation/flutter;
- ventricular tachycardia;
- complete A-V block.

Some of these arrhythmias can be associated with structural cardiopathies.

**Supraventricular tachycardia:** the cardiac beats show a fast rate, normally around 180 beats per minute (28). In most cases there are no underlying cardiac pathologies. Ebstein’s anomaly of the tricuspid valve and rhabdomyomas can be responsible for a low number of cases.

Supraventricular tachycardia is generally well tolerated for short periods, however if should it be continuous it can cause myocardial dysfunction, tricuspid valve reflux and cardiac insufficiency that manifest echographically through the appearance of hydrops. Echocardiography can identify all of these complications.

**Complete A-V block:** echocardiography allows the definition of a differential diagnosis between complete A-V block and sinus bradycardia by showing a normal cardiac rhythm, a slow ventricular rhythm and the presence of an A-V dissociation. The complete A-V block can be an isolated anomaly, in those cases where the mother shows a disorder affecting the connective tissue such as systemic lupus erythematosus, even on a subclinical scale.

In such cases the A-V blockade is caused by circulating

antibodies, especially anti-Ro and anti-La antibodies. These antibodies cross the placenta and damage the fetal conduction tissue hence producing a complete cardiac blockade.

Bradycardia is generally well tolerated, even though a small number of fetuses experience an intrauterine cardiac insufficiency.

In the other half of cases of complete cardiac blockade, there is a basic cardiac malformation such as the correct complete transposition of great vessels or left isomerism (29). Death rates are generally quite high in the latter group, revealing a substantial number of deaths during prenatal life.

### Benefits of fetal echocardiography

Fetal echocardiography has many benefits; some of these are still under investigation. The diagnosis of CHD during prenatal age is fundamental since it helps the counseling and enables parents to be informed and be prepared psychologically for the moment of birth or, alternatively, offers them the free choice of termination of pregnancy.

The newborn affected by CC should be born in a III level centre, equipped with a Unit of Intensive Neonatal Care, so that in the transition from pre- to post-natal life, the baby does not face periods of hypoxia or acidosis, and can be given immediate care.

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