IMPORTANCE OF PRENATAL DIAGNOSIS OF CONGENITAL HEART DISEASE

Major cardiac defects are found in about 5 per 1,000 births and they account for about 50% of all neonatal and infant deaths from congenital abnormalities. Cardiac defects may be an isolated finding or may occur in association with other defects, chromosomal abnormalities and genetic syndromes.

Most major cardiac defects can be diagnosed prenatally by a relatively brief but skilled examination of the heart during the routine mid-trimester scan. Prenatal diagnosis allows planning of the appropriate time and place of delivery to optimise the chances of survival after postnatal treatment. In a few cases, such as arrhythmias, it is also possible to carry out prenatal treatment. In cases of severe defects prenatal diagnosis provides the parents with the option to terminate the pregnancy.

APPROACH TO FETAL CARDIAC SCANNING

Fetal echocardiography is not particularly difficult and can be quite easily learnt by somebody already competent at general fetal scanning. It is far preferable, and easier in the long run, to invest in trying to understand the underlying anatomy rather than simply learning the appearance of the standard views by rote. This is helpful in both obtaining the standard views and in interpreting them.

A good cardiac examination is absolutely dependent on obtaining adequate views of the heart. During the examination it is important to positively demonstrate that the fetal heart is normal rather than suspect that the heart may be normal because of mere failure 'to notice something wrong'. The presence of a major abnormality may itself be a cause of difficulty in obtaining standard views. If the fetal position precludes adequate views, then it is essential to either await for spontaneous fetal movements or undertake such measures as getting the mother to fill or empty her bladder, rotating or tilting her abdomen, or gently manipulating the fetus by appropriate pressure on the maternal abdomen.

ANATOMY OF THE NORMAL HEART

An understanding of the anatomy of the normal heart is essential in obtaining the appropriate views for fetal echocardiography. The heart is three-dimensionally complex and the standard four-chamber view represents only one thin slice through it. Adjacent slices appear very different. It is important to appreciate that firstly, the left atrium is not really on the left but it is situated posteriorly near the midline of the chest, secondly, the right atrium lies in front of, below and to the right of the left atrium, and thirdly, the left ventricle lies posterior as well as to the left of the right ventricle. The right ventricle and its outflow tract wrap around the left ventricle and outflow tract.
ULTRASOUND ANATOMY OF THE HEART

Post-natally, imaging planes are limited because of interposed air-filled lung or substantial bone. Pre-natally, when the lungs are fluid filled and the bones less dense, images may be obtained in almost any plane, though the relative positions of the spine, limbs, placenta and maternal tissues to the heart may influence image quality. No single plane will demonstrate all cardiac abnormalities, but the vast majority of major abnormalities will be readily demonstrated by one or more standard plane views of the heart. The skilled sonographer does not perform a random search for recognisable standard planes but works methodically and deliberately to establish orientation and position and moves from one plane to the next by understanding their relative positions in a mental three-dimensional model of the heart.

The four-chamber view
The four-chamber view is obtained by a transverse section through the lower half of the fetal chest (Figures 1 and 2). The best way to achieve this is by firstly, obtaining a mid-sagittal section of the fetal chest, secondly, rotating the transducer through 90 degrees, and then making fine adjustments according to the image obtained. In the correct plane, the stomach or other abdominal organs are not visible, a complete rib is present and none of the left ventricular outflow tract should be visible, the crux occupying the central portion of the heart. The standard four-chamber plane implies a specific section through the fetal heart that fulfills these criteria, rather than any view which includes all four chambers of the heart. A precise 4-chamber view is essential for correct diagnosis.

Figure 1. Plane for examination of the four-chamber view of the fetal heart. Ao aorta, PA pulmonary artery, RA right atrium, LA left atrium, RV right ventricle, LV left ventricle.
The main features to confirm from the four-chamber view are that:
1) The heart occupies no more than 1/3 of the area of the thorax.
2) The cardiac apex points to the fetal left.
3) There is no pericardial effusion.
4) The right and left atria are of the same width.
5) The right and left ventricles are of the same width and contract equally.
6) The ventricular septum is intact.
7) The atrioventricular septum, at the crux of the heart, is intact. 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 septal hinge point. See Figure 15.
8) The pulmonary veins enter the left atrium.
9) The heart rate and rhythm are normal.

In order to demonstrate these features it is often necessary to obtain the four-chamber view from two different angles so that the ventricular septum appears both vertically and horizontally. In 'Spine-up' views heart structures may be obscured by acoustic shadows.

Figure 2. Four-chamber view of normal fetal heart. RA right atrium, LA left atrium, RV right ventricle, MB moderator band, LV left ventricle, MV mitral valve, TV tricuspid valve, PV pulmonary veins, dAo descending aorta.
The left ventricular outflow view

The left ventricular outflow tract lies in a plane nearer to the head than the four-chamber view and is directed towards the right shoulder of the fetus. To examine this tract it is necessary to rotate the transducer clockwise from the four-chamber plane with some angulation towards the fetal head, regardless of fetal lie, provided that it is not spine up (Figures 4 and 5). In the correct view, the tricuspid valve should not be visible, being out of the plane of section, but the mitral valve should be demonstrated. In this view, the ventricular septum forms a continuous line with the aortic wall without any break or step (shown in red in Figure 3). A break or step in this line may indicate the presence of a ventricular septal defect. An ideal left ventricular outflow view demonstrates sufficient length of the vessel that leaves it to confirm its lack of proximal major branches and sweeping course out towards the right shoulder, typical of a normal aorta rather than an abnormally connected pulmonary artery.

Figure 3. Plane for examination of the left ventricular outflow tract view of the fetal heart. This plane, which cuts through the right shoulder of the fetus, may be reached by clockwise rotation of the transducer from the four-chamber plane. The red line shows the normal continuity between the ventricular septum and the aortic wall. Loss of this normal continuity may indicate a ventricular septal defect. Note that the right ventricular inflow lies below the level of this plane and thus the tricuspid valve is not seen in this view. aAo ascending aorta, dAo aorta, SVC superior vena cava, LA left atrium, RV right ventricle, LV left ventricle.
Figure 4. Left ventricular outflow tract view of normal fetal heart. aAo ascending aorta, Ao aortic valve, dAo descending aorta, MV mitral valve, LA left atrium, RV right ventricle, LV left ventricle.

The right ventricular outflow view

The right ventricular outflow tract is directed backwards towards the fetal spine. To examine this tract it is necessary to slide and angle the transducer upwards from the four-chamber plane (Figures 5 and 6). This is sometimes referred to as the three-vessel view, as it demonstrates the superior caval vein (SVC), the ascending aorta and the pulmonary artery in continuity with the arterial duct.

The right ventricular outflow view should be interpreted in conjunction with the left ventricular outflow view. Scanning back and forth between the two views helps establish the relationship of the pulmonary artery and aorta and their relative sizes (Figures 3-6).
Figure 5. Plane for examination of the right ventricular outflow tract view of the fetal heart. This plane is angled up towards the head relative to the four-chamber view. SVC superior vena cava, Ao aorta, PA pulmonary artery, RV right ventricle, LA left atrium, RV right ventricle, LV left ventricle.

Figure 6. Right ventricular outflow tract view of normal fetal heart. SVC superior vena cava, Ao aorta, PA main pulmonary artery, RPA right branch pulmonary artery, RV right ventricle, LA left atrium, RV right ventricle, PV pulmonary valve.
The main features to confirm from outflow tract views are that:

1. There are two separate arterial valves. Both arterial valves should be seen to open freely, the thin valve cusps disappearing from view as they approximate the vessel wall in systole.
2. The two arteries are of approximately equal width, the pulmonary artery being slightly larger.
3. The left and right ventricular outflow tracts are directed almost at right angles to each other at their origin.
4. The artery arising from the front (right) ventricle is the main pulmonary artery, which has a characteristic straight course back towards the spine. The main pulmonary artery is very short, soon dividing into three: the right and left pulmonary artery branches and the arterial duct, which is a direct extension of the main artery.
5. The artery arising from the posterior (left) ventricle is the aorta. In contrast to the pulmonary artery, the ascending aorta is relatively long, follows a sweeping course and the first main branches are those to the head and neck. The aorta forms the higher of two arches in the thorax. The lower arch, formed by the pulmonary artery in continuity with the arterial duct ("ductal arch") is devoid of branches to the head.

The transverse arch view

The transverse arch view is obtained in an oblique transverse plane high in the thorax (Figure 10). Because the aortic arch rises higher in the chest than the ‘ductal arch’, which lies to its left, the imaging plane has to be tilted down towards the fetal left side to take in both arches.

The main features to confirm from this view are that:
1. The vessels from fetal left to right are the pulmonary artery, aorta and superior caval vein, with the pulmonary artery being the more anterior vessel.
2. The aortic arch and pulmonary artery/ductal arch should be approximately equal in width at about 20 weeks. At later gestations, the pulmonary artery tends to be a little bigger than the aorta. A marked discrepancy in size (aorta smaller than pulmonary artery) may indicate the presence of coarctation of the aorta.

Figure 7. Plane for examination of the transverse arch view of the fetal heart.