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REVIEW ARTICLE

Screening of Fetal Heart for the Congenital Heart Diseases

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ABSTRACT

Screening of the congenital heart disease (CHD) is one of the most important techniques in prenatal ultrasonographic examination. Step by step screening methods for taking account for the level of screener, especially in the area with poor detection rate, is needed for starting effective fetal CHD screening. In this review, the fetal cardiac screening is divided into two methods accounting for steps for learning screening technique; the basic screening and the advanced screening. Basic screening is a simple method even for the one who is not familiar to the cardiac anatomy. The goal of this basic screening is to detect most of the ductal dependent lesions including transposition of the great arteries. For the basic screening, 'location' and 'size' of the heart and vessels are checked in standard four-chamber view and three-vessel view. Advanced screening is a screening for detecting all fetal CHDs, including total anomalous pulmonary venous return. For the advanced screening, the side of the heart is defined, and then the 'detail anatomy' and the 'function and blood flow' are assessed in all standard screening views, including one fetal abdominal transverse view and three fetal chest transverse views, such as four-chamber, three-vessel and three-vessel and trachea view.

Keywords: Fetal echocardiography, Fetal screening, Congenital heart disease.

INTRODUCTION

Screening of the congenital heart disease (CHD) is one of the most important techniques in prenatal ultrasonographic examination. Recent conventional ultrasound equipment can visualize cardiac anatomy *in utero* from 16 weeks of gestation even by the transabdominal approach.[1-3] Referral to the tertiary care center after effective screening of fetal CHD can make it possible to diagnose detail anatomical abnormality and to plan appropriate perinatal management. Prenatal diagnosis of CHD can improve the outcome of the fetuses with CHD.[4]

For the screening, detail anatomical assessment of the CHD is not needed. The purpose of the screening is to detect the fetuses with possibility of having CHD, and to refer to the fetal cardiac center. Several methods with simple technique have been proposed for effective screening. However, learning the technique for obtaining the images in sufficient quality still requires certain training, hence, the detection rate of fetal CHD is varied between the areas.[5] Different condition of the region and country, such as medical system and political environment, may be one of the causes of difficulty for establishing effective fetal CHD screening program. An issue for poor detection rate in some regions seems to be caused by the feeling of difficulty for starting fetal cardiac screening for the screeners who are not familiar to the fetal echocardiography. Hence, step by step screening methods for taking account for the level of screener in certain area is needed for increasing the area for starting fetal CHD screening.

In this review, we present the fetal cardiac screening dividing into two methods account for step for learning screening technique; the basic screening and the advanced screening. The basic screening is a very simple method even for the screener who is not familiar to the somewhat complicated cardiac anatomy. The advanced screening is the method of goal for all fetal sonographic screener who is trying to detect all major CHD.

INDICATIONS OF THE SCREENING

Appropriate ultrasonographic screening of fetal CHD is indicated for all pregnant women.[1-3] Unlike in the newborn period, the presence of severe cardiac disease in the fetus, such as heart murmur and cyanosis, cannot be detected by physical examination. Therefore, fetal ultrasonographic screening of a completely healthy mother with an uneventful pregnancy is the only method of identifying the majority of cases of congenital heart disease *in utero*.

The first screening ultrasound should be performed at approximately 20 weeks of gestation, if termination of pregnancy is to be considered as an option in complex cardiac problems, and there is some restriction in gestational age by law for termination. Screening has to be performed sufficiently early to make it possible to refer the case to a fetal cardiac center and to allow the parents to make their decision. The second screening for fetal CHD should be performed at approximately 30 weeks of gestation because some cardiac abnormalities develop more obvious structural abnormality in later gestation.

CONCEPT OF BASIC AND ADVANCED SCREENING

Basic Screening

Basic screening is a simple method even for the one who is not familiar to the cardiac anatomy. This screening method can be applied when the detection rate of fetal CHD is low in that region. The goal of this basic screening is to detect most of the ductal dependent lesions including transposition of the great arteries (TGA). Although, standard four-chamber view and three-vessel view are used in this basic screening, the check point is only limited to the location and size of the heart and vessels. Checking the detail anatomy of the heart is not required if the purpose of the screening is limited to detect major CHD, except total anomalous pulmonary venous drainage (TAPVR).

Advanced Screening

Advanced screening is a screening for detecting all fetal CHDs as much as possible, including TAPVR. First, the side of the heart is defined. Then, all standard screening views including one fetal abdominal transverse view and three fetal chest transverse views are checked. In all view, detail anatomy and function and blood flow of the heart and vessels, in addition to the location and size, are checked.

FETAL HEART SECTIONS FOR SCREENING CHD

For the fetal heart screening, defining the side of the heart is needed (advanced screening). The other views are four transverse images of the fetus. First one is an abdominal transverse image of the fetus, and remaining is three simple transverse images of the fetal chest, such as the four-chamber, the three-vessel, and the three-vessel and trachea view (Figs 1 and 2). In each view, checking points are divided into location, size, detail anatomy and function and blood flow.

Defining the Side of the Heart

Since conventional ultrasound image is two-dimensional in nature, the side of the image depends on the probe direction, and cannot be interpreted from the 2D image itself. Hence, the side of the fetal heart has to be defined at the beginning of the examination (Fig. 3).[6] A longitudinal section of the fetal chest or abdomen is imaged with the fetal head positioned at the right side of the screen. Then, a fetal transverse section is imaged by rotating the probe by 90° clockwise. Regardless of fetal position,

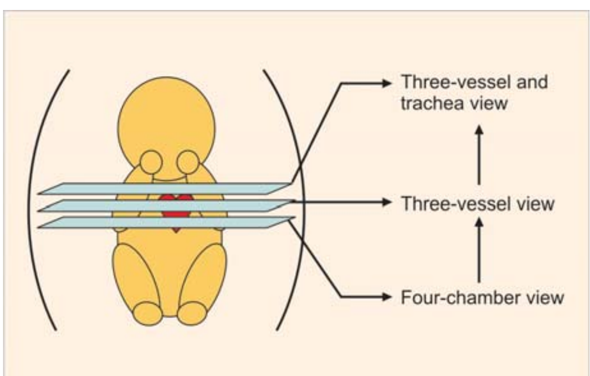

Fig. 1: Three transverse sections of the fetal chest, four-chamber, three-vessel, and three-vessel and trachea view

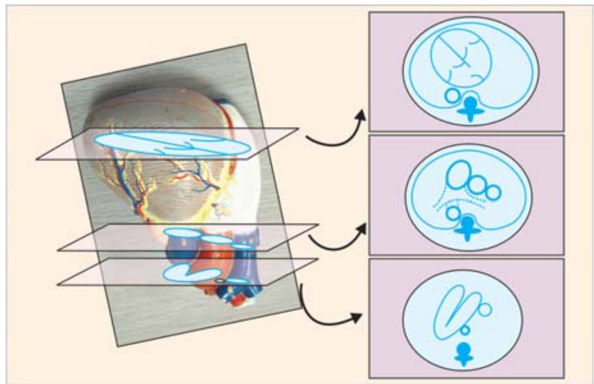

Fig. 2: Location of the each transverse section of the fetal heart

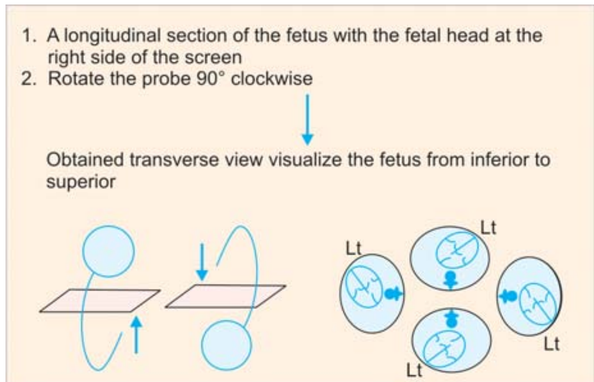

Fig. 3: Step for deciding the side of the fetal heart

the obtained image should be used to visualize the fetus from inferior to superior. In this view, the side of the heart and stomach should be defined whether it is left side (normal) or right side (abnormal). The easy way to interpret the side of the heart is that the heart is rotated clockwise after the probe is rotated clockwise.

Transverse Section of the Fetal Abdomen

Assessment of location is sufficient in this view. The stomach is left side, same as the fetal heart. In front of the fetal spine, the descending aorta (dAo) is positioned left side, and the superior vena cava (SVC) is positioned right and anterior to the dAo.

Four-Chamber View

A four-chamber view of the fetal heart is obtained by a transverse section of the lower part of the fetal chest (Figs 2, 4 and 5). This four-chamber view can detect more than 50% of CHDs, such as single ventricle, hypoplastic left ventricle (Fig. 6) and Ebstein's anomaly (Figs 7 and 8).

The location and size in this four-chamber view should be assessed for basic screening (Figs 4 and 5). There are three check points in each of location and size assessment in four-chamber view. For the location assessment, the fetal heart is positioned at left side of the chest, and the heart axis is about 45%. The dAo is positioned at the left anterior to the spine. For

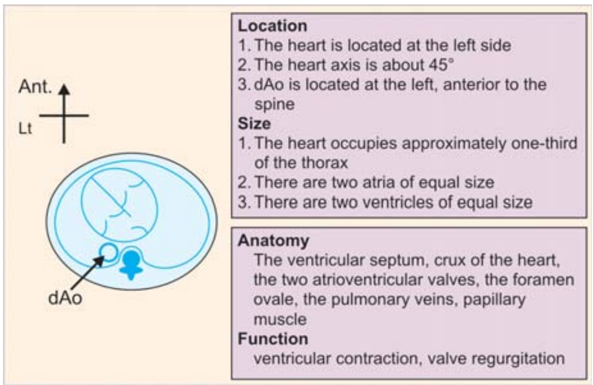

Fig. 4: Check list of four-chamber view

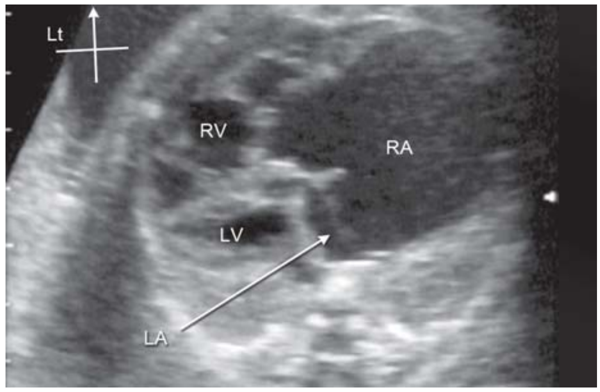

Fig. 7: A four-chamber view of a fetus with Ebstein's anomaly. The heart is quite enlarged and fills almost the entire chest. The right atrium (RA) is enlarged because of severe tricuspid valve regurgitation. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle

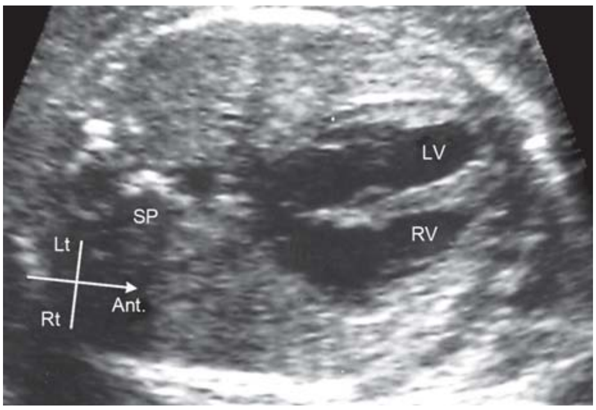

Fig. 5: Fetal echocardiographic image of four-chamber view

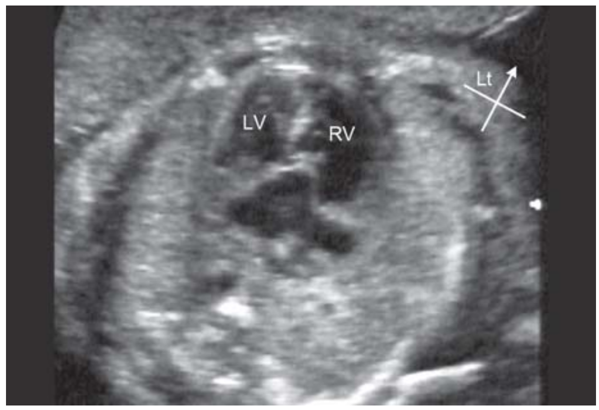

Fig. 8: A four-chamber view of a fetus with atrioventricular septal defect. A large one common atrioventricular valve is revealed, and the crux of the heart is not formed. LV, left ventricle; RV, right ventricle

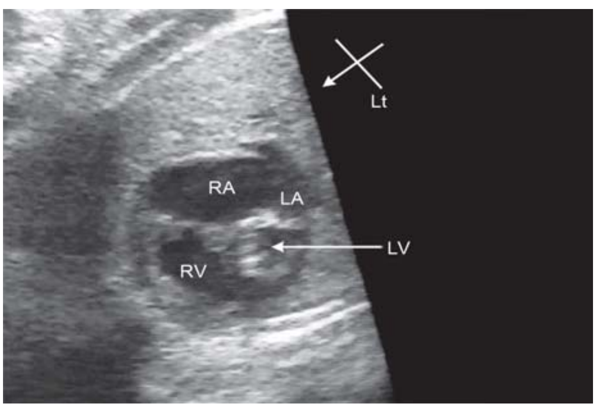

Fig. 6: A four-chamber view of a fetus with hypoplastic left heart syndrome. The left ventricle (LV) is small and endocardium of LV is high echogenic due to endocardial fibroelastosis. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle

the size assessment, the fetal heart occupied approximately 1/3rd of the fetal chest. There are two atria of equal size, and two ventricles of equal size.

The assessment of the detail anatomy and the function and blood flow is included for advanced screening. Sufficient knowledge of cardiac anatomical feature and usage of color Doppler are required for these assessments. For the assessment of the detail anatomy, the crux of the heart, the ventricular septum, flap of the foramen ovale, the atrioventricular valves, the pulmonary veins and the papillary muscles should be checked. Next, function and blood flow should be assessed. Good contraction of each ventricle is carefully checked. By the color Doppler flow, location of the pulmonary venous return can be confirmed. In addition, regurgitant jet at the mitral valve and tricuspid valve can also be detected.

Three-Vessel View

The three-vessel view can be obtained by sweeping superior from the four-chamber view (Figs 2, 9 and 10).[7] This view can detect abnormal connections between the ventricles and the great arteries, such as TGA (Fig. 11) and tetralogy of Fallot.

The location and the size in this three-vessel view should be assessed for basic screening (Fig. 9). There is a just one check point in each of location and size assessment in three-vessel view. For the location assessment, three vessels are straightly aligned from left anterior to the right posterior. In another ward, the most anterior vessel is located at the most left side. For the size assessment, the vessel size is aligned from the largest to the smallest when the straightly aligned vessels are followed from left anterior to right posterior. The most left anterior side vessel, the main pulmonary artery, is of the largest size. The central vessel, the ascending aorta (aAo), is medium in size. The most right posterior side vessel, the superior vena cava, is the smallest.

In order to assess the location of the three vessels with confidence, sweeping movement of images from the four-chamber view to the three-vessel view is useful. Using this moving image with sweeping, connection from both the ventricle to the vessels is visualized, so that the location of the vessel is clearly assessed. The ascending aorta and the main pulmonary arteries have cross relationship in normal heart (Fig. 12). By this sweeping movement, the connection from the left ventricle to the ascending aorta is first visualized. The ascending aorta is located posteriorly to the right ventricle and running from left to the right side according to the sweeping movement of the images. After this, the connection from the right ventricle to the main pulmonary artery is visualized. The main pulmonary artery is located anteriorly to the aorta just posterior to the chest-wall, and running from right to left side according to the sweeping movement of the images. The most obvious abnormality of this cross relationship of the two vessels is TGA. For the fetus with TGA, two vessels are aligned to parallel and straight rather than cross (Fig. 13), making the most left-side vessel, the main pulmonary artery, located posteriorly to the right sided ascending aorta (Fig. 11).

The assessment of the detail anatomy and the function and blood flow are required for advanced screening. For the detail anatomy assessment, the most left side vessel, the main pulmonary artery, is branching pulmonary arteries. It also connected to descending aorta via the ductus arteriosus at a slightly superior slice of three-vessel view. For the blood flow assessment, color Doppler flow reveals laminar flow in both the main pulmonary artery and the ascending aorta, directed from anterior to the posterior.

Three-Vessel and Trachea View

The last screening view, three-vessel and trachea view (Figs 14 and 15), can be obtained by farther superior sweep from three-vessel view.[8] This view may be included to the advanced

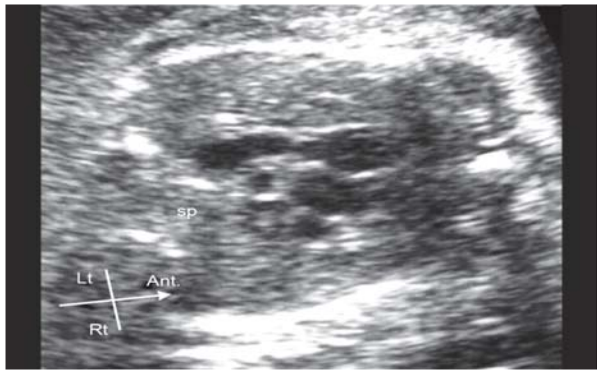

Fig. 10: A fetal echocardiographic image of three-vessel view

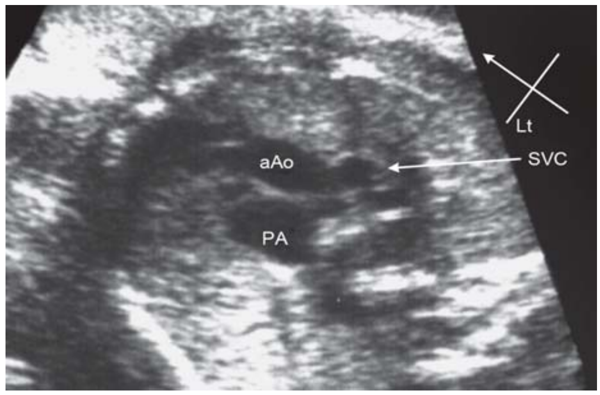

Fig. 11: A three-vessel view reveals a right anterior ascending aorta (aAo) and left posterior main pulmonary artery (PA). ant, anterior; SVC, superior vena cava

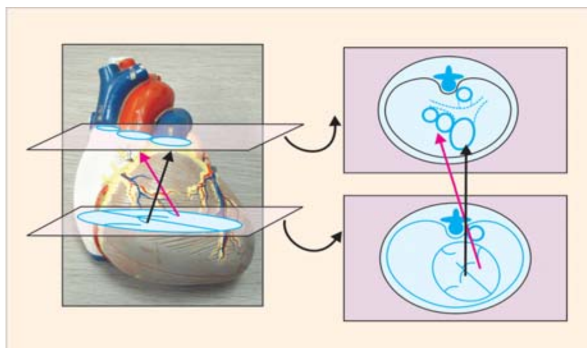

Fig. 12: Normal cross connection of the ventricles and the great vessels

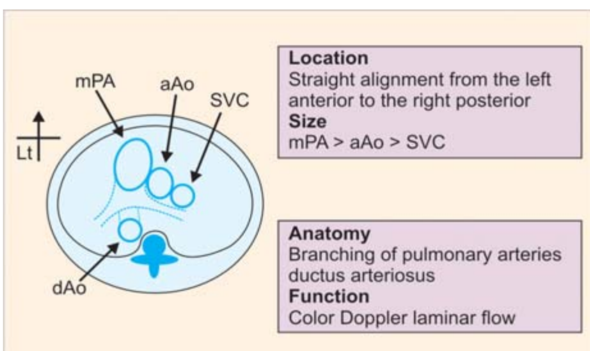

Fig. 9: Check list of three-vessel view

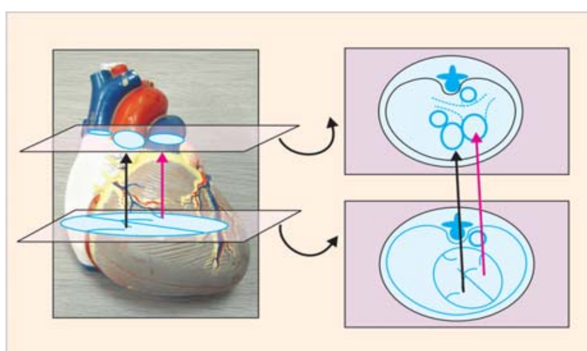

Fig. 13: Abnormal parallel connection of the ventricles and the great vessels

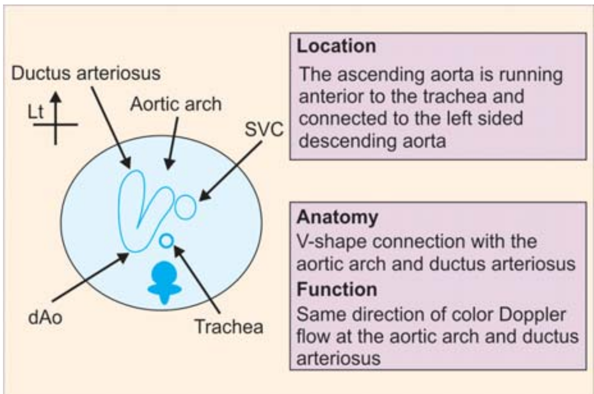

Fig. 14: Check list of the three-vessel and trachea view

Fig. 15: A fetal echocardiographic image of three-vessel and trachea view

screening. This three-vessel and trachea view is useful for detecting variable vascular rings, which sometimes cause marked airway problems postnatally (Fig. 16).

For the location assessment, the aortic arch crosses anterior side of the trachea from right anterior to the left posterior, and connected to the descending aorta. There is no 'size' assessment in this view. For the detail anatomy assessment, the ascending aorta is connected to the descending aorta via the aortic arch and isthmus, and the aortic arch and the ductus arteriosus makes V shape at the left side of the trachea. For the function and blood flow assessment, the color Doppler flow reveals the flow to the same direction at both the ascending aorta and the ductus arteriosus.

ROLE OF THREE-DIMENSIONAL ECHOCARDIOGRAPHY TO THE SCREENING

Three-dimensional (3D) echocardiography may have important role for the both the basic and advanced screening of abnormal fetal heart.[9,10] The information of the side of the heart is already included to the 3D data, and the four simple transverse views for screening can be obtained from the 3D data set (Fig. 17). For the conventional two-dimensional echocardiography, fetal movement and limited window due to the fetal position often makes it difficult to do fetal heart screening. Whereas in the 3D echocardiography, any optimal screening image can be obtained without fetal movement once the 3D data set of the fetal heart is acquired and saved into the hard disk.

For only the basic screening, still 3D data set is sufficient to assess the location and the size using the four-chamber view and the three-vessel view (Fig. 18). For the advanced screening, STIC method is needed to assess the detail anatomy and the function and blood flow. The STIC method, 3D images with heart beat, can be created by automatic calculation of fetal heart rate from the acquired data set.

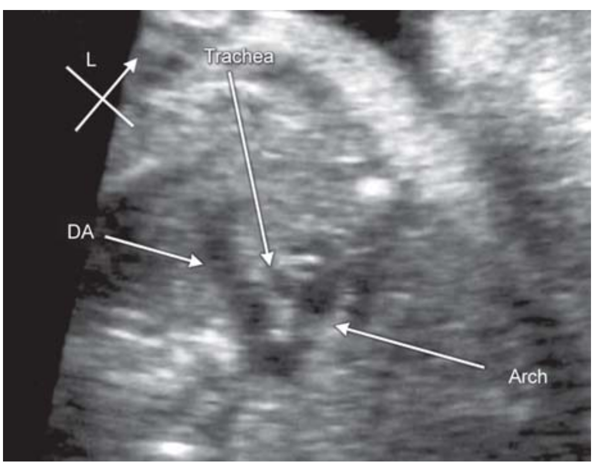

Fig. 16: A three-vessel and trachea view of the fetus with vascular ring caused by right sided aortic arch. The aortic arch is running right side of the trachea, and the ductus arteriosus (DA) is running left side of the trachea which making vascular ring

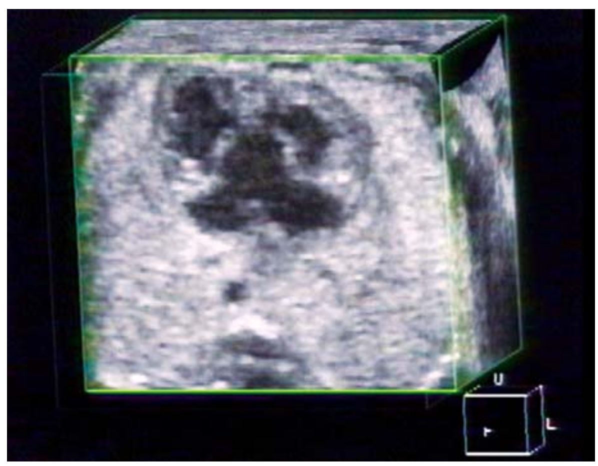

Fig. 17: Three-dimensional echocardiography in a case with common atrioventricular valve (asplenic syndrome). Optimal 2D cutting plane can be created from the 3D data set and demonstrated from optimal angle

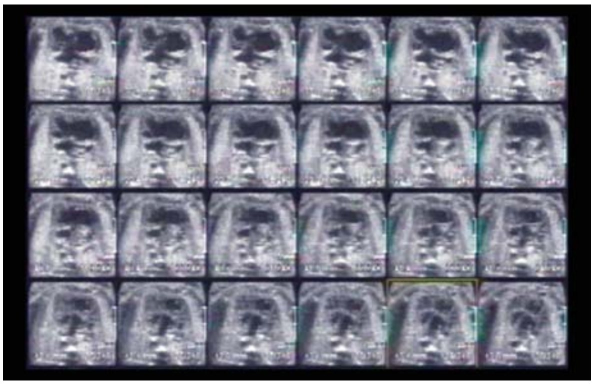

Fig. 18: Three-dimensional echocardiography in a case with common atrioventricular valve. Multiple parallel cutting planes obtained from 3D data set demonstrate the connection from the cardiac chambers to the vessels

CONCLUSIONS

Since the recent development of the fetal echocardiography and development of the perinatal management, the issue of the prenatal diagnosis of the fetal CHD is the screening system. In order to start fetal cardiac screening system in certain region, where the screeners are not familiar to the screening of CHD, step by step screening methods for taking account for the level of screener in certain area is needed. We present the fetal cardiac screening divided into two methods accounting for steps for learning screening technique; the basic screening and the advanced screening. The basic screening using only four-chamber view and three-vessel view with the assessment of location and size does not require complex knowledge of cardiac structures. Using this basic screening method, we believe more region start the systematic fetal screening program, and more fetuses have benefit for recent advanced perinatal management of CHD.

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