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Review article

Prenatal diagnosis of congenital heart disease: A review of current knowledge

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ABSTRACT

This article reviews important features to improve the diagnosis of congenital heart disease (CHD) by applying ultrasound in prenatal cardiac screening. As low and high-risk pregnancies for CHD are subject to routine obstetric ultrasound, the diagnosis of structural heart defects represents a challenge that involves a team of specialists and subspecialists on fetal ultrasonography. In this review, the images highlight normal anatomy of the heart as well as pathologic cases consistent with cardiac malposition and isomerism, septal defects, pulmonary stenosis/atresia, aortic malformations, hypoplastic left ventricle, conotruncal anomalies, tricuspid dysplasia, and Ebstein's anomaly, and univentricular heart, among other congenital cardiovascular defects. Anatomical details of most CHD in fetuses were provided by two-dimensional (2D) ultrasound with higher quality imaging, enhancing diagnostic accuracy in a variety of CHD. Moreover, the accuracy of the cardiac defects in obstetrics ultrasound improves the outcome of most CHD, providing planned delivery, aided genetic counseling, and perinatal management.

Contents

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1. Introduction

Congenital heart disease (CHD) are the most common form of birth defects. The incidence of CHD is about eight to 10 per 1000 (0.8%–1%) live-born, full-term births, and it could be 10 times higher in preterm infants (8.3%).[1,2] Furthermore, in early gestation, this incidence is even higher as certain CHDs are complex and have been show to result in fetal demise. In fact, 50%–60% of the CHD will require surgical correction and of these, 25% are critical with CHD a leading cause of infant mortality.[3,4] In this setting, the survival, extensive medical care, and developmental disabilities depend on the time of the diagnosis, on the delay of the treatment, and on the severity of the CHD. Therefore, early fetal diagnosis of a treatable CHD has been shown to reduce the risk of perinatal morbidity and mortality.[5]

Cardiovascular development involves a complex process in which genetic and environmental factors are involved. Considering that approximately 49% of pregnancies are unplanned, women may not take precautionary actions against environmental factors.[6] The detection of CHD by fetal echocardiography when referred by a suspicion of cardiac abnormality on routine obstetric ultrasound is up to 40% in low-risk populations. However, risk factors are identified in only 10% of CHD. In this scenario, the heart should be examined in detail on a routine sonographic scanning.

In the sonographic prenatal diagnosis of CHD, the fetal heart remains a challenge that involves sonographers, obstetricians, radiologists, and fetal medicine subspecialists. High risk for cardiac defects and the suspicion of a cardiac abnormality on obstetric ultrasound, even in low-risk populations, are indications for referral for performance of a detailed fetal echocardiogram. This manuscript reviews important aspects to improve the prenatal screening for CHD focusing on ultrasound clues to enable the diagnosis of the cardiac defects; drawing the management of cardiac defects in utero and the delivery plan strategies were also approached in this study.

2. Prenatal screening

2.1. How to screen the fetal heart

The fetal cardiac screening by ultrasound can detect a high proportion of cases of CHD. However, when the prenatal screening was based on the visualization of the four-chamber view, it was inadequate to detect many cases of CHD, especially conotruncal and outflow defects (ex: transposition of the great vessels, tetralogy of Fallot, double-outlet right ventricle, truncus arteriosus, and outlet septal defects). When the evaluation of outflow tracts was added to the four-chamber view, the sensitivity of ultrasound screening for CHD increased from approximately 30% to 69%–83%.[7] Currently, the three vessels (3 V) and 3 V with trachea (3VT) views were added to the standard four-chamber and outflows views in order to improve the detection of CHD.[7,8] The latter one enabled the detection of lesions such as coarctation of the aorta, right aortic arch, double aortic arch, and vascular rings, achieving a prenatal detection rate of congenital heart disease to up 90%. The average time to obtain the cardiac views was just over 2 min, but in approximately one third of cases, the cardiac examination was postponed by 15–20 min due to unfavorable fetal lie (anterior spine).[9] A fetal echocardiogram should be performed if the CHD is suspected on the obstetric cardiac of screening, or if there is a recognized increased risk (maternal, fetal and/or familial factors) for CHD >2% to 3%. Fetal echocardiography may be considered when risk is estimated at 1% to 2%, and when risk approaches that of the general population (≤ 1%), this exam is not indicated.[10]

2.2. Upper abdomen and four-chamber views

The examination of the upper abdomen (cross-sectional plane) of the fetus by echocardiography provides the distinction between the left and the right sides of the fetus. When the situs is normal (solitus), the aorta and stomach are located on the left side and the inferior vena cava and liver are placed on the right. Therefore, situs solitus is the normal arrangement of thoracic and abdominal organs (Fig. 1). In general, more complex CHD are associated with abnormalities of the situs. Furthermore, the umbilical vein and the hepatic veins can be visualized in upper abdomen view.

The four-chamber view is the most important plane. This approach enables the evaluation of the main cardiac structures, the position, the size (1/3 of the thorax), the contractility and the rhythm of the heart. In normal levocardia, 2/3 of the heart is left-

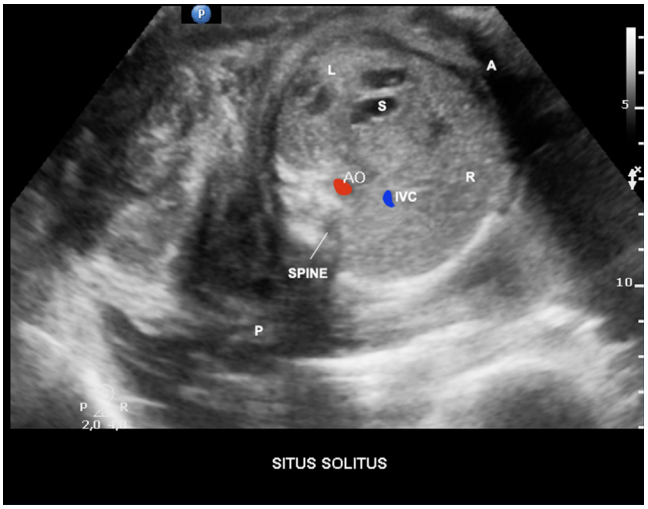

Fig. 1. Upper abdomen view showing situs solitus. AO, aorta; IVC, inferior vena cava; L, fetal left; R, fetal right; P, fetal posterior; A, fetal anterior; S, stomach.

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