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REVIEW ARTICLE

Diagnosis of Transposition of the Great Arteries in the Fetus

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Transposition of the great arteries (TGA) is a group of congenital cardiac defects characterized by ventriculoarterial discordance. It is one of the most common cyanotic heart diseases, and most affected neonates are undiagnosed as fetuses. The sequelae of the complex congenital heart diseases could be severe if undiagnosed before birth. However, the outcome of fetuses with TGA is improved with prenatal detection of this condition. The purpose of this article is to review the gold standard and additional signs for the diagnosis of fetal TGA.

Introduction

Transposition of the great arteries (TGA) was first described by Mathew Baillie in 1797 [1]. The term transposition was first used by Farre in 1814, in which "trans" means "across" and "position" means "placement." Transposition means that the aorta and pulmonary artery are placed across the ventricular septum. A few decades ago, Van Praagh et al suggested that the aberrations in conotruncal development could result in a continuum of malpositions of the great arteries [2]. TGA is simply one subtype of these malpositions. Other subcategories of these malpositions include double-outlet right ventricle, double-outlet left ventricle, and anatomically corrected malposition [3].

The reported prevalence of congenital heart disease (CHD) varies between four and 10 per 1000 live births [4—6]. Due to advances in fetal ultrasound resolution and techniques, obstetricians have the increasing ability to detect fetal CHD prenatally, thereby prompting timely referral for adequate perinatal management. Based on

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a review of the literature, the antenatal diagnosis and transfer of neonates with CHD could improve both short-term and long-term outcomes, especially in those with cyanotic congenital heart diseases [6,7].

In a population-based review, cardiac defects account for almost half of the neonatal deaths attributed to congenital malformations [8]. Tetralogy of Fallot and TGA are the most common cyanotic heart defects, each constituting approximately 10% of fetal congenital heart defects [9—13]. However, in most fetuses, TGA remains undiagnosed before birth. According to a population-based review [14], TGA is diagnosed prenatally in only 17% of affected neonates. In contrast, approximately one-half of cases of tetralogy of Fallot can be correctly diagnosed *in utero*. We conducted a comprehensive review for the diagnosis of TGA in the fetus because of this low detection rate.

Terminology

TGA as well as other CHD are best diagnosed using the segmental approach of the fetal heart [15—17]. To enhance understanding of the terminology used in the segmental approach for the diagnosis of TGA, the following terms should be clarified in advance [2,3,16,18—20].

Atrioventricular (A-V) discordance: The morphologic right atrium (RA) is erroneously connected to the morphologic left ventricle (LV); and the morphologic left atrium (LA) is incorrectly drained to the morphologic right ventricle (RV).

Ventriculoarterial (V-A) discordance: The pulmonary artery (PA) arises from a morphologic LV, and the aorta (Ao) arises from a morphologic RV.

Complete transposition of the great arteries (TGA): Indicates the conditions of ventriculoarterial discordance. The Ao arises from a morphologic RV, and the PA arises from a morphologic LV. The A-V connection is correct.

Congenitally corrected transposition of great arteries (ccTGA): Refers to the condition of A-V discordance plus V-A discordance. In brief, the RA enters the LV, which gives rise to the PA. The LA connects the RV, which gives rise to the Ao. Thus, the circulation becomes physiologically corrected although double errors occur.

d-loop: Denotes the normal rightward (*dextro*, *d*) loop of the embryonic cardiac tube. The inflow portion of the RV is to the right of the morphologic LV.

l-loop: Denotes the cardiac tube bending leftward (*levo*, *l*) during embryogenesis. The inflow portion of the morphologic RV is to the left of the morphologic LV.

Detecting transposition of the great arteries in the fetus

According to the policy of the Bureau of Health Promotion in the Department of Health of Taiwan, general practitioners should perform routine ultrasound to evaluate fetal growth at 20 weeks' gestation. The diagnosis of TGA can be made by carefully and appropriately evaluating the anatomic locations of cardiac chambers and the connections between the atria, ventricles, and great arteries at this gestational age with high-resolution ultrasound. However, it must be emphasized that diagnostic accuracy is only half based on the literature [21—23]. The gold standard and the additional signs of ultrasound in the diagnosis of fetal TGA are discussed in the following paragraphs.

The localization of cardiac chambers and great arteries

The first step in the diagnosis of fetal TGA is the localization of the cardiac chambers and their connections with the great arteries [22]. This is best done by the segmental approach as described in the literature [15,24—26]. These cardiac structures can be identified on the basis of their specific morphologic features.

Anatomically, the differentiation of the atrial chamber is based on the morphologic aspect of the atrial appendages. The atrial appendages are the earlike extensions of the atria. Typically, the RA appendage is triangular in shape, whereas the LA appendage is fingerlike [15]. However, differentiation of atrial appendages is difficult during the antenatal period. In addition, systemic and pulmonary venous connections may provide important clues to define situs of the atria in the fetus. The supradiaphragmatic portion of the inferior vena cava (IVC) provides a reliable landmark to identify the anatomic RA (because of the rule of venoatrial concordance) [15], and the drainages of four pulmonary veins usually define the location of anatomic LA (Fig. 1A and B).

Certain features that could help differentiate the right and left ventricles are the texture and distribution of internal trabeculae. The trabeculae are coarse in the RV, but thin and delicate in the LV (Fig. 1B and 1C). Moreover, the papillary muscles of the RV are attached to both the interventricular septum and the lateral wall, yet the two papillary muscles of the LV are attached only to the lateral wall of myocardium [15,27,28] (Fig. 1C). Nevertheless, these anatomic features may not be apparent in the fetus. The insertion of the tricuspid valve to the ventricular septum is lower than that of the mitral valve, and the location of the moderator band in the apical area identifies the location of the RV (Fig. 1B). Both characteristics are valuable in defining the morphologic RV.

The major characteristic of the PA is that the three branches (right and left PA, ductus arteriosus) immediately emerge from the main PA when it arises from the RV (Fig. 1D). In contrast, the Ao does not branch into the ascending portion except for the coronary arteries. The aortic arch can be seen by positioning the transducer to the left parasagittal plane. In the view, the Ao is seen to arise from the middle of the fetal thorax with acute curvature (usually described as having a 'candy cane' appearance). Three branches from the transverse arch can be identified as the innominate, the left common carotid, and the left subclavian arteries (Fig. 1E). The ductal arch can be seen by sliding the transducer to the left from the aortic arch (with a slight tilt of the transducer). In the normal fetus, the ductal arch arises from the anterior thorax with a wide curvature (usually described as having a 'hockey stick' appearance). There is no branch in the