Cite as: Bravo-Valenzuela NJ, Peixoto AB, Araujo Júnior E: Prenatal diagnosis of congenitally corrected transposition of the great arteries. J Ultrason 2019; 19: 78: 314–317. doi: 10.15557/JoU.2019.0048

Submitted: 04.02.2019
Accepted: 01.06.2019
Published: 31.12.2019

Prenatal diagnosis of congenitally corrected transposition of the great arteries

Nathalie Jeanne Bravo-Valenzuela, Alberto Borges Peixoto, Edward Araujo Júnior

Department of Obstetrics, Paulista School of Medicine – Federal University of São Paulo (EPM-UNIFESP), São Paulo-SP, Brazil

Correspondence: Prof. Edward Araujo Júnior, PhD, Rua Belchior de Azevedo, 156 apto. 111 Torre Vitoria, São Paulo-SP, Brazil, CEP 05089-030; tel.: +55-11-37965944, e-mail: araujojred@terra.com.br

DOI: 10.15557/JoU.2019.0048

Keywords
congenitally corrected transposition of the great arteries, fetal, prenatal diagnosis, echocardiography

Abstract

Congenitally corrected transposition of the great arteries is a rare and complex congenital heart disease. It essentially occurs with atrioventricular and ventriculoarterial discordance, in which the great vessels become parallel. Cases of corrected transposition of the great arteries are more frequently associated with other cardiac anomalies. In the fetus, corrected transposition of the great arteries may not be diagnosed on obstetric cardiac ultrasound, especially in the absence of other cardiac malformations (isolated corrected transposition of the great arteries). In this manuscript, we describe a case of isolated corrected transposition of the great arteries diagnosed in utero, and highlight the clues to make this diagnosis.

Introduction

Congenitally corrected transposition of the great arteries, a rare congenital heart disease, is characterized by discordant ventriculoarterial and atrioventricular connections. It is considered a conotruncal anomaly that accounts for approximately 0.05% of congenital heart diseases after birth[1]. Although corrected transposition of the great arteries is rarely associated with chromosomal or extracardiac anomalies, it is frequently associated with cardiac anomalies, such as ventricular septal defect, left ventricular outflow tract obstruction, anomalies of the left-sided tricuspid valve, and complete heart block[2].

During heart morphogenesis, an abnormal left looping of the primitive cardiac tube brings the morphological left ventricle to the right even as the morphological right ventricle veers to the left. This ventricular inversion is generally accompanied by transposition of the great arteries. Accordingly, in corrected transposition of the great arteries, a double discordance enables the physiological correction of the transposed great arteries. However, the issue of the morphological right ventricle as the systemic ventricle, and cardiac arrhythmia are associated with clinical symptoms in adulthood. Indeed, depending on the degree of corrected transposition of the great arteries, associated cardiac lesions may occur as clinical manifestations, even during the neonatal period[3].

In this manuscript, we describe a case of isolated corrected transposition of the great arteries diagnosed *in utero*, focusing on the clues necessary to make the prenatal diagnosis during fetal cardiac ultrasound/echocardiography.

Case report

A 21-year-old secundigravida at 37 weeks of gestation was referred to a fetal cardiologist due to suspected transposition of the great arteries. She was a healthy pregnant woman, and there was no familial history of a congenital heart disease, infectious diseases or consumption of any drugs. Fetal echocardiography showed

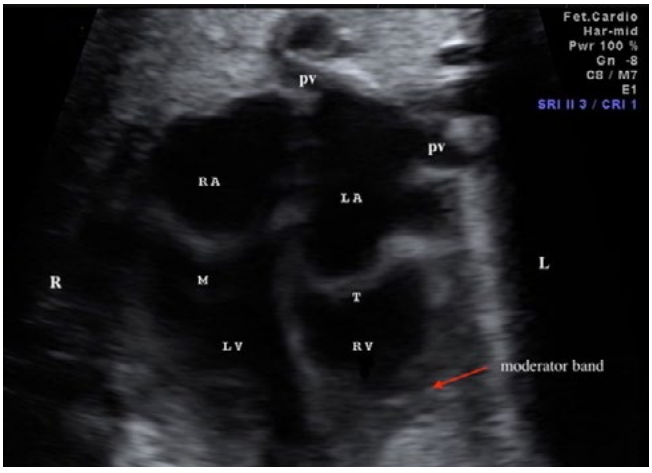

Fig. 1. *Four-chamber view at 37 weeks of gestation. The left atrium, which is located most posteriorly and contains the foramen oval flap, is connected to the right-shaped left ventricle; the right atrium is also connected to the left-shaped right ventricle. Note that the left-sided ventricle contains the moderator band (red arrow) and tricuspid valve (left-sided AV valve displays greater apical displacement than the right-sided one). RA – right atrium; LA – left atrium; RV – right morphological ventricle; LV – left morphological ventricle; R – right side; L – left side; T – tricuspid valve; M – mitral valve; pv – pulmonary vein*

atrioventricular and ventriculoarterial discordance (Fig. 1 and Fig. 2). The four-chamber view demonstrated that the left-sided ventricle was more trabeculated than the right-sided ventricle and displayed greater apical displacement of its atrioventricular valve. Thereafter, the anatomic right ventricle was on the left, and the anatomic left ventricle was on the contralateral side (Fig. 1). The great arteries ran in parallel and showed a different fashion in the three-vessel view, in which the ascending aorta was located on the left side of the main pulmonary artery. The male newborn weighing 3070 g was born by vaginal delivery at 38 weeks of gestation with Apgar scores of 9 and 10 at 1st and 5th min, respectively. Postnatal echocardiography confirmed the transposition of the great arteries with inverted ventricles (Fig. 3). There was no association with other cardiac anomalies. The electrocardiogram demonstrated regular sinus rhythm. The neonate was discharged in a stable condition two days after birth. He is currently two years old and remains asymptomatic.

Discussion

Cases of corrected transposition of the great arteries without associated cardiac anomalies (isolated corrected transposition of the great arteries) are rare, and the diagnosis may not easily be made by cardiac screening ultrasound. In this setting, corrected transposition of the great arteries is a unique conotruncal anomaly, in which the four-chamber view is abnormal. The left-sided ventricle containing the moderator band and tricuspid valve (left-sided AV- atrioventricular ventricle valve displays greater apical displacement than the right-sided one) is the morphological right ventricle, and the right-sided ventricle is the morphological left ventricle. Furthermore, the left atrium, which is located more posteriorly and contains the foramen ovale flap, is connected to the right-shaped left ventricle, where the right atrium is also connected to the left-shaped right ventricle. Therefore, the ventricular inversion aided by a transposed relationship of the great arteries is an important clue to enable the diagnosis of corrected transposition of the great arteries by fetal cardiac screening ultrasound[4].

In cases of corrected transposition of the great arteries, the atrial situs is generally solitus with levocardia. However, dextrocardia and mesocardia are common in the cases of corrected transposition of the great arteries. Approximately 5% of patients with corrected transposition of the great arteries have atrial situs inversus, which includes significantly fewer complications than those with situs solitus[3]. In the cases of corrected transposition of the great arteries with situs solitus, there is a higher prevalence of Ebstein-like anomalies and congenital heart

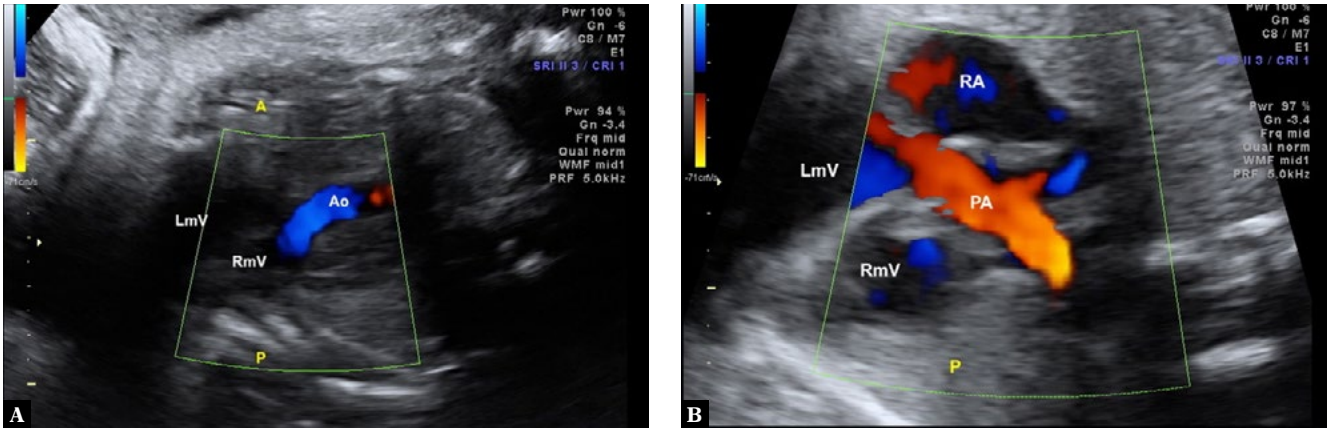

Fig. 2. *Fetal echocardiography at 37 weeks of gestation showing the ventricular outflow tracts:* **A.** *the aorta artery arising from the right morphologically ventricle (left-sided ventricle) and* **B.** *the pulmonary artery arising from the left-morphologically ventricle (right-sided ventricle). Ao – aorta; PA – pulmonary artery; RA – right atrium; RmV – right morphological ventricle; LmV – left morphological ventricle; A – anterior; P – posterior*

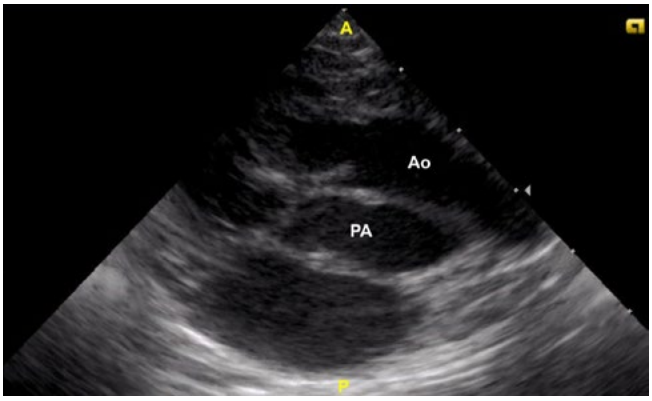

Fig. 3. *Postnatal transthoracic echocardiogram of the parasternal long axis with the great vessels running in parallel. Ao – aorta; PA – pulmonary artery; A – anterior; P – posterior*

block[5]. The ventricles occasionally display a superoinferior relationship. Classically, the aorta is anteriorly located to the left of the pulmonary artery in the three-vessel view[6].

Considering the outflow tract views, the diagnosis of transposed arteries is based on the identification of the origin of the aorta, which arises from the anterior ventricle. The pulmonary artery arises from the posterior ventricle and is aided by a parallel relationship of the great arteries. A rightward convexity of the anterior ventricle outflow tract known as "the reverse boomerang sign" is an easier marker of transposed great arteries during the first trimester[7]. It is important to differentiate between corrected transposition of the great arteries and transposition of the great arteries. In corrected transposition of the great arteries, the aorta arises from the morphologically right ventricle, which is the left-sided ventricle. Conversely, transposition of the great arteries displays normal spatial arrangement of the ventricles. Further, in corrected transposition of the great arteries, the left-sided atrioventricular valve (morphologically – the tricuspid valve) is not in fibrous continuity with the aortic valve because of the presence of the anatomically right ventricular infundibulum. Indeed, anatomically corrected malposition of the great arteries is a rare type of a congenital heart disease, in which the aorta and the pulmonary aorta are not transposed. However, the great arteries run parallelly. In transposition of the great arteries and corrected malposition of the great arteries, the four-chamber view appears normal, and in the outflow tract views, the great arteries are in a parallel relationship. However, in corrected malposition of the great arteries, the great arteries originate from the morphologically appropriate ventricle (ventriculoarterial concordance). Often misdiagnosed as transposition of the great arteries *in utero*, malposition of the great arteries results in a conotruncal inverted twist with normal ventricular looping (ventricular dextro-loop)[8].

In the three vessels and trachea-view, the "V shape" of the great arteries is not present when the great arteries are transposed because of the anterior course of the aorta in relation to the pulmonary artery. Further, the posterior pulmonary trunk is not visible in transposition of the great arteries or corrected transposition of the great arteries, and the three vessels and trachea-view is frequently a misnomer because only two vessels are identified ('2V finding')[9]. Furthermore, the appearance of the aortic arch in the three vessels and trachea view resembling the letter 'I' ('I-shaped sign') can be used as a sign to prenatal detection of transposition of great arteries[10].

In contrast to transposition of the great arteries and similarly to the anatomically corrected malposition of the great arteries, a specialized delivery room care team is not necessary in the cases of isolated corrected transposition of the great arteries. In the latter case, the physiology of this defect is congenitally corrected by double discordance (atrioventricular and ventriculoarterial), and in malposition of the great arteries, the great arteries arise from the respective ventricles with a parallel relationship (double concordance). In transposition of the great arteries, (atrioventricular concordance and ventriculoarterial discordance), the delivery should be planned at a specialized center that can manage the hypoxia and hemodynamic compromise. In cases of simple transposition of the great arteries, an urgent balloon atrial septostomy and a surgical repair, such as an arterial switch operation (the Jatene procedure), will be required during the first weeks of life[11]. Indeed, in the cases of corrected transposition of the great arteries and corrected malposition of the great arteries, a neonatal surgical approach will be required depending on the cardiac anomalies that can be associated with these conditions. Further, a significant proportion of patients with corrected transposition of the great arteries will develop congenital heart block during their life. However, complete heart block is much less common *in utero*. In the cases of isolated corrected transposition of the great arteries, the short-term outcome is favorable with symptoms in adulthood, and the surgical management is controversial. Considering potential progressive right ventricle dysfunction, the surgical approach of anatomical surgical repair or double-switch (combined atrial and arterial switch) has been proposed[12].

Conclusion

In summary, this case describes the clues to enable the prenatal diagnosis of corrected transposition of the great arteries by ultrasound/echocardiography. Although the physiology of this defect is congenitally corrected by the double discordance, the right ventricle is the systemic ventricle with an increased risk of deterioration in adulthood or earlier depending on the degree of associated cardiac defect.

Conflict of interest

Authors do not report any financial or personal connections with other persons or organizations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

References

1. Wallis GA, Debich-Spicer D, Anderson RH: Congenitally corrected transposition. Orphanet J Rare Dis 2011; 6: 22.
2. Presbitero P, Somerville J, Rabajoli F, Stone S, Conte MR: Corrected transposition of the great arteries without associated defects in adult patients: clinical profile and follow up. Br Heart J 1995; 74: 57–59.
3. Oliver JM, Gallego P, Gonzalez AE, Sanchez-Recalde A, Brett M, Polo L *et al*.: Comparison of outcomes in adults with congenitally corrected transposition with situs inversus versus situs solitus. Am J Cardiol 2012; 110: 1687–1691.
4. Sharland G, Tingay R, Jones A, Simpson J: Atrioventricular and ventriculoarterial discordance (congenitally corrected transposition of the great arteries): echocardiographic features, associations, and outcome in 34 fetuses. Heart 2005; 91: 1453–1458.
5. Mah K, Friedberg MK: Congenitally corrected transposition of the great arteries situs solitus or inversus. Circ Cardiovasc Imaging 2014; 7: 849–851.
6. Paladini D, Volpe P, Marasini M, Russo MG, Vassallo M, Gentile M *et al*.: Diagnosis, characterization and outcome of congenitally corrected transposition of the great arteries in the fetus: A multicenter series of 30 cases. Ultrasound Obstet Gynecol 2006; 27: 281–285.
7. Bravo-Valenzuela NJ, Peixoto AB, Araujo Júnior E, Da Silva Costa F, Meagher S: The reverse boomerang sign: a marker for first-trimester transposition of great arteries. J Matern Fetal Neonatal Med 2017; 32: 677–680.
8. Van Praagh R: What determines whether the great arteries are normally or abnormally related? Am J Cardiol 2016; 118: 1390–1398.
9. Yagel S, Arbel R, Anteby EY, Raveh D, Achiron R: The three vessels and trachea view (3VT) in fetal cardiac scanning. Ultrasound Obstet Gynecol 2002; 20: 340–345.
10. Ishii Y, Inamura N, Kawazu Y, Kayatani F, Arakawa H: 'I-shaped' sign in the upper mediastinum: A novel potential marker for antenatal diagnosis of d-transposition of the great arteries. Ultrasound Obstet Gynecol 2013; 41: 667–671.
11. Donofrio MT, Moon-Grady AJ, Hornberger LK, Copel JA, Sklansky MS, Abuhamad A *et al*.: Diagnosis and treatment of fetal cardiac disease: a scientific statement from the American Heart Association. Circulation 2014; 129: 2183–2242.
12. Said SM, Burkhart HM, Schaff HV, Dearani JA: Congenitally corrected transposition of great arteries: surgical options for the failing right ventricle and/or severe tricuspid regurgitation. World J Pediatr Congenit Heart Surg 2011; 2: 64–79.