A rare combination of correct transposition of the great arteries and isolated levocardia: Imaging characterization

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Abstract
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Abstract
Congenitally correct transposition of the great arteries (cc-TGA) is an extremely rare congenital cardiac malposition. They are usually detected by echocardiography in the fetal period. This case report describes a 58-year-old female patient who presented with tachycardia. The combination of cc-TGA and isolated levocardia is incidentally diagnosed by transthoracic echocardiography and cardiac magnetic resonance imaging.

KEYWORDS
Congenitally correct transposition of the great arteries, Isolated levocardia, Echocardiography, Magnetic resonance imaging

A 58-year-old female was referred to our hospital complaining of tachycardia for about half a month. There was no history of hypertension, diabetes, thyroid disease or drug abuse. Physical examination revealed a grade 3/6 systolic murmur in the fourth left intercostal space. Transthoracic echocardiography (TTE) suspected that right heart had enlarged and left heart had decreased. However, there was no atrioventricular septal defect. Further evaluation demonstrated the liver and inferior vena cava (IVC) were located on the left side of the abdomen, while the spleen and aorta (AO) were on the right side (Fig.1A-C). The right atrium
(RA) received IVC blood and the left atrium (LA) received pulmonary venous (PV) blood (Fig.1D-E). The atrial situs inversus was existed. Thus, the position of the heart chambers was reconsidered. In the 3-vessel-trachea view, the abnormally spatial relationship of great arteries was indicated. The AO was located right in front of the main pulmonary artery (PA), which was also known as D-positioned aorta (Fig.1F). Based on the morphological character of moderate band, atrioventricular valve and ventricular trabecular, the spatial relationship of ventricular loop was normal, which was meant D-loop. The enlargements of LA and right ventricle (RV) were also seen on two-dimensional echocardiography (Fig.1G). Color Doppler flow imaging (CDFI) showed the tricuspid valve had severe regurgitation with the systolic peak velocity and pressure gradient were 7.0m/s and 195mmHg, respectively (Fig.1H-I). Owing to her ultrasonic features, a cardiac magnetic resonance imaging (MRI) was then performed and confirmed the presence of situs inversus and cc-TGA (Figure 2). Thus, based on the above images, a combination of cc-TGA (IDD) and isolated levocardia was verified. The patient refused further medical treatment. Upon outpatient follow-up on three months, the tricuspid regurgitation had no significant changes.

Isolated levocardia is an extremely rare disease with an incidence of approximately 1/2,000,000.1 It is commonly accompanied by congenital heart defect. Cc-TGA is characterized by atrioventricular and ventriculoarterial discordance. According to the position of vicoatrial situs, heart ventricle and the great arteries, cc-TGA was divided into two types: SLL: atrial situs solitus, L-loop, L-positioned aorta; IDD: atrial situs inversus, D-loop, D-positioned aorta. IDD accounts for roughly 5% of cc-TGA.2 Echocardiography is the method of choice for early detection of cc-TGA and can be used to assess the chamber size, the spatial relationship of great arteries, and the hemodynamic changes.3 According to the position of liver and IVC, RA can be confirmed. The position of atroventricular valve is in accordance with the ventricles. Moderate band is the typical symbol of RV. Besides, TTE is a valuable tool for the evaluation of valvular stenosis and regurgitation severity.

Due to the RV replaces LV as a systemic pumping chamber in patients with cc-TGA, ventricular structure and interventricular interaction becomes abnormal.4 The pathophysiology of ccTGA mainly depends on the heart malformation involved. Palliative care is the treatment of choice for patients without severe regurgitation and coexist of other heart malformations.

Reference

FIGURE 1 Echocardiographic images of cc-TGA (IDD) and isolated levocardia. (A, B, C) Transthoracic echocardiography and transabdominal ultrasound indicated situs inversus. (D, E) The RA received IVC blood (blue arrow) and the LA received PV blood (red arrow). (F) The spatial relationship of AO and the main PA was displayed and it was D-positioned aorta. (G) 4-chamber view showed the enlargements of LA and RV. (H, I) CDFI revealed tricuspid incompetence and regurgitation. AO, aorta; IVC, inferior vena cava; LA, left atrium; PA, pulmonary artery; PV, pulmonary vein; RA, right atrium; RV, right ventricle.
**FIGURE 2** (A) The longitudinal section showed situs inversus and the cardiac apex pointed towards the left side of the chest. (B) The cross section revealed four heart chambers. PV blood flowed to LA (red arrow). (C, D) RV blood flowed to ascending aorta (AAO, red arrow) and LV blood flowed to PA (blue arrow). Superior vena cava (SVC) and IVC blood flowed to RA (blue arrow). IVC, inferior vena cava; LA, left atrium; LV, left ventricle; PA, pulmonary artery; PV, pulmonary vein; RA, right atrium.

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**Ethical approval** Not applicable. Since this case report only used figures, no approval from our institutional review board was required.

**Informed consent** Written consent was obtained from our patient for publication.