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Case Report

Cardiac MR imaging reveals L-type transposition of the great vessels and failing right heart[☆]

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ABSTRACT

L-type transposition of the great vessels is a rare congenital heart disease in which both the great arteries and the ventricular chambers are reversed. Because this condition preserves a physiologic circulatory pathway, it can be challenging to diagnose in infants with no concurrent cardiac abnormalities. Early detection is essential, however, because these patients will eventually experience severe complications, as the structural right ventricle is unable to function long-term in the systemic position. We report a rare case of L-type transposition of the great vessels in a 32-year-old male who presented in adulthood with tachycardia and palpitations. The initial echocardiogram was inconclusive. Further imaging (cardiac MRI & transesophageal echocardiogram) revealed the inverted anatomy due to the presence of key morphological features, such as the malposed great vessels along with the moderator band and prominent trabeculae within the right ventricle, which was functioning systemically.

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Introduction

Transposition of the great vessels (TGV) is a congenital heart disease (CHD) typically diagnosed in neonates. It most commonly occurs as *dextrorotation* (D-TGV) characterized by ventriculoarterial (VA) discordance, wherein the aorta arises from the right ventricle (RV) and the pulmonary artery arises from the left ventricle (LV). In the setting of proper atrioventricular (AV) concordance, this establishes a parallel circula-

tion in which deoxygenated blood returns to the systemic vasculature. Therefore, these newborns are cyanotic at birth [1].

Alternatively, patients with *levorotation* of the great vessels (L-TGV) display both VA and AV discordance. Systemic venous return to the right atrium (RA) is normal. However, due to ventricular inversion, the RA empties into the morphologic LV. This ventricle gives rise to the pulmonary artery and will be referred to as the *subpulmonic* ventricle in this report. Pulmonary venous return to the left atrium (LA) is normal. The LA empties into the morphologic RV, which gives rise to the

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aorta. This leaves the weaker, morphologic RV in the systemic position; it will be referred to as the *subsystemic* ventricle in this report.

Despite malposition of both the great arteries and the ventricles, the anatomy maintains proper oxygenation of the systemic circulation, resulting in a CHD that is clinically silent at birth [1]. Most infants with L-TGV present in infancy due to other heart defects, such as large ventricular septal defects (VSD) or valvular stenosis. Because the atypical arterial-chamber connections are physiologically adequate during infancy, heart failure is not present initially. Diagnostic imaging of L-TGV is focused on identifying the subtle morphologic differences between left- and right-sided cardiac anatomy and the malposition of the great arteries [2]. We discuss a case of a patient with congenital L-TGV and an associated VSD presenting in adulthood with chronic, systolic heart failure from the morphologic RV supporting the systemic circulation.

Case report

A 32-year-old male presented to an outpatient cardiologist for tachycardia, palpitations, and chest pain. He had been evaluated for a murmur as a child, and a small membranous VSD was found. The patient was lost to follow-up. At presentation, 2D echocardiography displayed systemic ventricular remodeling with a mildly reduced ejection fraction. Other findings included an aneurysmal appearance at the basal to mid portion of the septum; the LA was mildly enlarged with associated significant AV valve regurgitation. Due to the limitations of echocardiography in evaluation of 3D cardiac structure and anatomy, a follow-up cardiac MRI was recommended for better visualization.

Subsequent MRI revealed that the patient's actual pathology involved AV and VA discordance, consistent with L-TGV. The aorta, arising from the subsystemic RV, was positioned anterior and to the left of the pulmonary artery which arose from the subpulmonic ventricle (Fig. 1). There was also a small, perimembranous VSD with shunting from the systemic to pulmonary circulation. The subpulmonic LV displayed normal structure and function. The moderator band was identified in the left-sided, subsystemic ventricle (Figs. 2A and 3A). There were coarse trabeculations as well, thus labeling the subsystemic ventricle as the morphologic RV (Figs. 2A and 3B). Further assessment of the subsystemic RV revealed significant dilation and consequent systolic dysfunction due to its unsuited role as the systemic pump. The pulmonary veins identified the LA which was seen emptying into the trabeculated subsystemic RV (Fig. 2B). A ventricular septal aneurysm and septal dyskinesia were also noted at the base.

Cine MRI showed a regurgitant jet into the dilated LA from the subsystemic ventricle, indicating tricuspid valve regurgitation. Global hypokinesia of the subsystemic RV and significant tricuspid insufficiency with resultant LA enlargement account for the patient's RV failure and reduced ejection fraction of 21.3%. Ultimately, the patient was referred to an adult congenital heart clinic for a transesophageal echocardiogram (TEE) and further management recommendations. TEE aided in shunt evaluation through the patient's VSD and verified

that the subsystemic RV was failing due to chronically working against systemic vascular resistance.

Discussion

L-TGV is also known as “congenitally corrected” TGV and makes up less than 1% of all CHDs [3]. In general, transposition is more common in males compared to females, although there is no explanation for this discrepancy at present [4]. Unlike other CHDs, patients with L-TGV may remain hemodynamically stable until their subsystemic RV fails, a common manifestation during the fourth and fifth decades [5]. However, L-TGV patients are often born with concomitant cardiac anomalies, including VSD, pulmonic stenosis, tricuspid regurgitation, Ebstein anomaly, and heart block. VSD accompanies 70% of L-TGV cases, with its size determining the severity of clinical symptoms at birth; for example, the patient discussed here was an asymptomatic newborn, likely due to the small size of his VSD [1].

Many imaging modalities can be employed to capture the AV and VA discordance that characterizes L-TGV, including echocardiography, cardiac MRI, and TEE. The distinction in the morphology of the 2 ventricles is made by the presence of the moderator band and prominent trabeculae within the subsystemic RV. The morphologic RV demonstrates marked hypertrophy, contrasted by an abnormally thin-walled LV [1]. The AV valves are displaced in accordance with their normally associated ventricles, placing the tricuspid valve in the systemic left position with the morphologic RV [6,7]. The coronary arteries arise from the aorta but may have aberrant origins and course. Further imaging of the outflow tracts displays an aorta arising slightly anterior and to the left of the pulmonary trunk, which often leads to pulmonary stenosis/atresia in these patients [2].

Unfortunately, many of these characteristics are not clearly distinguishable and can easily be mistaken for other common cardiac malformations if L-TGV is not a considered differential. In the case of the patient discussed here, the initial echocardiogram was interpreted as traditional left heart failure with a remodeled/weakened systemic ventricle and a septal aneurysm; it was only upon follow-up imaging in the form of cardiac MRI that the correct diagnosis was made. Without conclusive analysis from echocardiography, 3D visualization of the anatomy was required via MRI. Thus, prior clinical indication, cardiac MRI, and TEE confirmed the final diagnosis of L-TGV.

Historically, treatment of L-TGV focused solely on repairing associated anomalies, since patients were otherwise hemodynamically stable. However, disregarding the AV and VA discordances during surgical correction of L-TGV is no longer recommended due to subsequent failure of the RV in the systemic role and other adverse cardiac sequelae, as demonstrated in our case [8]. A procedure known as *Double Switch* is now preferred in these patients to reestablish AV and VA concordance and restore the LV to the systemic position. Other defects, such as VSD and valvular stenosis, can be addressed at this time.

This more aggressive approach has improved survival rates compared to physiologic correction of concomitant anoma-

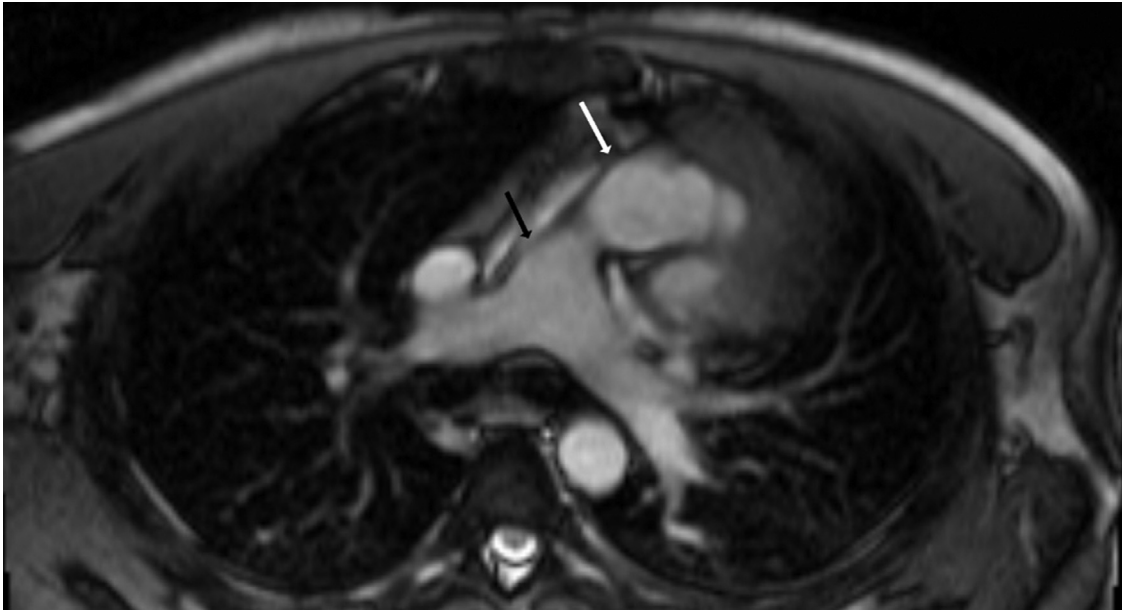


Fig. 1 – Great Vessel Orientation. Axial steady-state free precession MRI sequence demonstrating the inverted relationship of the great vessels. Typically, the aorta arises anteriorly and to the right of the pulmonary artery. In this patient, the aorta (white arrow) is positioned anteriorly and to the left of the pulmonary artery (black arrow).

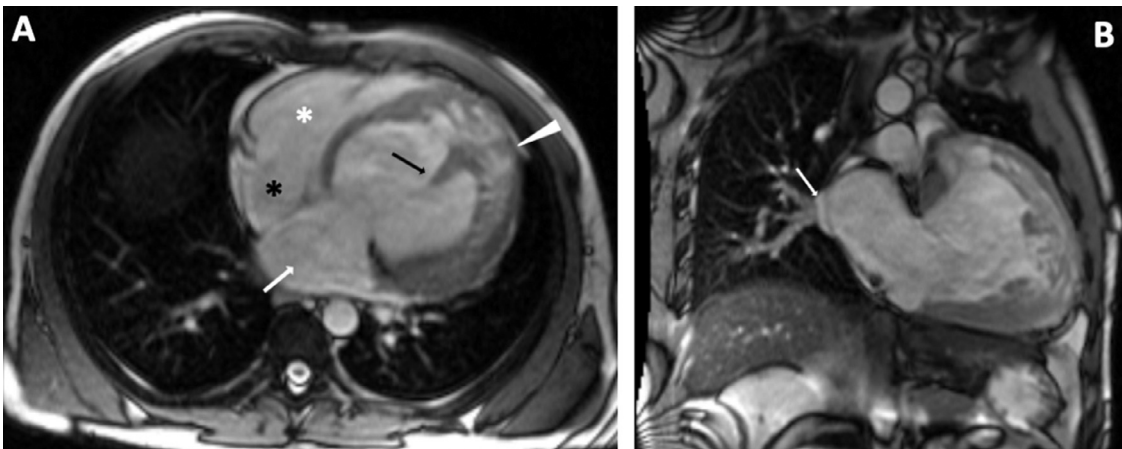


Fig. 2 – Atrial & Ventricular Architecture. (A) Four-chamber bright blood sequence demonstrating a subsystemic, morphologic RV containing a moderator band (black arrow). As a result of its systemic function, the morphologic RV shows a hyper-trabeculated wall with dilated ventricular chamber (white arrowhead). The hyper trabeculation is a normal RV finding and the dilation is due to subsystemic failure. This panel also demonstrates the discordant atrioventricular connections observed in L-TGV with the left atrium (white arrow) filling the morphologic RV and the right atrium (black asterisk) filling the subpulmonic, morphologic LV (white asterisk). (B) Two-chamber steady-state free precession MR sequence image demonstrating the pulmonary veins (white arrow) draining into the left atrium. The left atrium is seen emptying into a hyper-trabeculated morphologic RV.

lies alone [9]. Such anatomic corrections are typically made in the neonatal or infantile periods but may be performed further into adolescence. The most common long-term complications in symptomatic patients who did not undergo early surgical intervention are AV valve insufficiency and congestive heart failure, both of which are present in the patient discussed here [7,10]. Additionally, those with L-TGV are at an increased risk of developing complete heart block and arrhyth-

mias over time; such complications are often the presenting symptoms in these adult patients [10].

Ultimately, an RV in the subsystemic position is intrinsically unfit to support the systemic circulation and puts the patient at significant risk for heart failure and arrhythmias [2]. The prognosis of patients with L-TGV depends on time of diagnosis, history of surgical intervention, and underlying comorbidities, all of which affect the rate of progression to

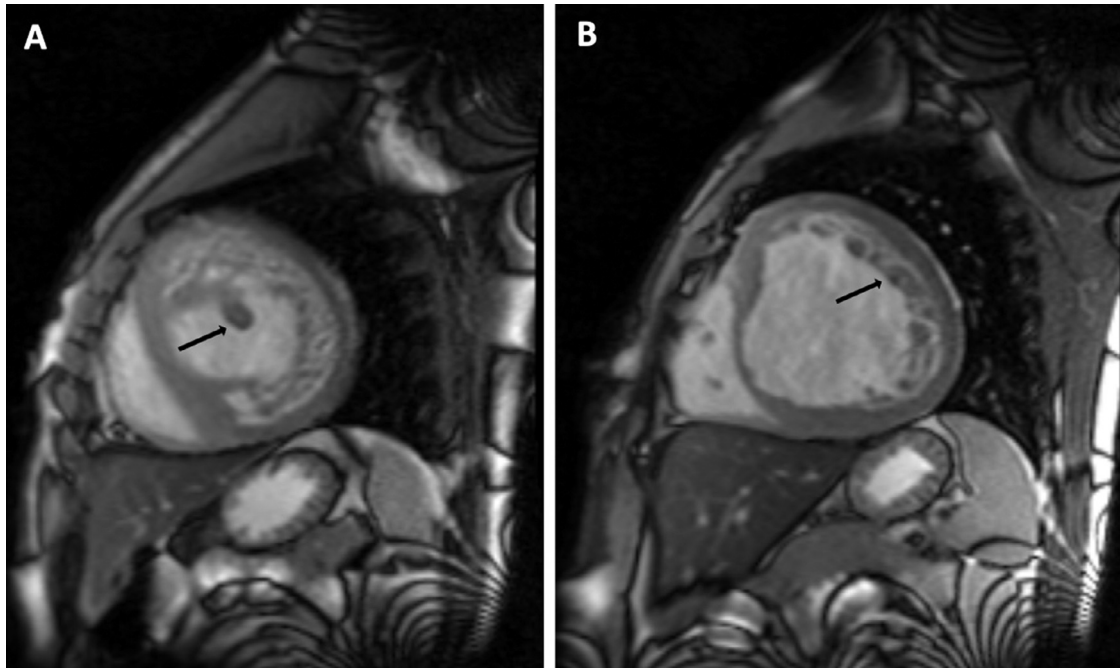


Fig. 3 – Morphologic Right Ventricle Anatomy. (A) Sagittal T2 bright blood short axis-weighted image of cardiac MRI showing the moderator band (black arrow) contained within the subsystemic morphologic RV. (B) Sagittal T2 bright blood short axis-weighted imaging of cardiac MR image showing the subsystemic morphologic RV with coarse trabeculations (black arrow) and dilated chamber.

heart failure and other cardiac dysfunctions. Follow-up imaging focuses on assessing AV valve insufficiency, as the rate of mortality significantly increases with greater degrees of regurgitation [10]. Due to the severity of these chronic complications and complexity of available treatment options, L-TGV requires prompt diagnosis by identifying subtle anatomical markers, namely the moderator band and prominent trabeculae in the wall of the RV, via appropriate imaging modalities. Young adults presenting with heart failure or spontaneous arrhythmia, particularly those with a known history of CHD, should undergo adequate imaging, including echocardiography and cardiac MRI, to rule out additional underlying congenital malformations, such as L-TGV. Although L-TGV is rare, it is vital to understand the condition's subtle presentation, assess a patient's cardiac symptoms without presumption of proper anatomy, and order advanced imaging for ambiguous echocardiogram results.

Patient consent

Written informed consent was obtained from the patient prior to reporting the case and findings.

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