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Case Report

Congenitally Corrected Transposition of the Great Arteries Identified Through Atrioventricular Block: A Case Report

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Abstract: Congenitally corrected transposition of the great arteries (CCTGA), is a complex and extremely rare congenital heart defect characterized by atrioventricular and ventriculoarterial discordance due to abnormal ventricular positioning. Isolated forms may remain asymptomatic for years, unlike cases associated with other cardiac anomalies. We report the case of an 18-year-old patient with no known medical history, in whom a first-degree atrioventricular block was incidentally detected during routine evaluation. Echocardiography performed revealed double discordance. Given the absence of symptoms, a conservative approach with regular clinical, ECG, and echocardiographic monitoring was adopted.

Keywords: Congenitally corrected transposition of the great arteries, Atrioventricular block, Echocardiography, Congenital heart disease.

1. INTRODUCTION

Congenitally corrected transposition of the great arteries is a complex and extremely rare congenital anomaly, representing less than 0.5% of all congenital heart diseases [1]. This anomaly results from a defect in the rotation of the cardiac loop during embryonic development [2]. It involves both atrioventricular and ventriculoarterial discordance, resulting from abnormal ventricular positioning. The ventricles are malposed such that the morphologically right ventricle is located on the left and receives blood from the left atrium, while the morphologically left ventricle is on the right and receives blood from the right atrium. The aorta arises from the right ventricle now situated on the left, and the pulmonary artery emerges from the left ventricle on the right. This configuration leads to a physiologically corrected but anatomically discordant circulation [3,4]. The etiology remains unknown; however, several risk factors have been reported in the literature, including maternal exposure to pesticides and the use of certain antiepileptic drugs during pregnancy [2]. A recently published study has also suggested a possible genetic link [5]. In nearly 90% of cases, this congenital heart defect is associated with additional anomalies such as ventricular septal defect, pulmonary stenosis, conduction and rhythm disturbances, coronary anomalies, and Ebstein's anomaly, all of which worsen the prognosis [6,7]. While simple forms may allow for an acceptable quality of life into adulthood, the majority of cases progress to global heart failure [8,9]. Prenatal diagnosis is possible but is often made late, typically in adulthood, either due to the presence of clinical signs or incidentally during echocardiography performed for unrelated reasons [10]. In addition to echocardiography, cardiac magnetic resonance imaging (MRI) is an essential tool for the diagnosis and follow-up of this condition [6,10,11]. The optimal treatment involves surgical correction aimed at restoring functional anatomy. However, this approach remains complex and is not systematically indicated [12]. In resource-limited settings, such as sub-Saharan Africa, access to specialized cardiac surgery is limited, making medical follow-up and clinical assessment all the more critical. In this context, we report the case of an asymptomatic young patient in whom double discordance was incidentally discovered during the evaluation of a conduction abnormality.

2. CASE PRESENTATION

An 18-year-old male with no known medical history was referred for evaluation of an atrioventricular block identified during a routine check-up. Physical examination revealed a grade 4/6 systolic murmur at the apex, with no other abnormalities. Hemodynamic and respiratory status were stable. Vital signs were as follows: blood pressure 125/75 mmHg, heart rate 95 bpm, respiratory rate 20 breaths per minute, oxygen saturation 93% on room air (tolerated desaturation),

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temperature 37.2°C, weight 55 kg, height 165 cm, and BMI 20 kg/m². Laboratory investigations were within normal limits. Electrocardiography showed a regular sinus rhythm, normal axis, first-degree atrioventricular (AV) block, and repolarization abnormalities in the apicolateral leads (Figure 1). Holter ECG confirmed a persistent first-degree AV block. Echocardiography demonstrated a morphologically right ventricle positioned on the left, featuring a low-inserting atrioventricular valve consistent with a left-sided right ventricle and a tricuspid valve inserted at a lower level than the mitral valve. The aorta arose from the morphologically right ventricle located on the left, while the pulmonary artery originated from the morphologically left ventricle on the right. A trabeculated interventricular communication with bidirectional shunting was observed, accompanied by moderate tricuspid regurgitation leading to left atrial dilation (Figure 2). The systemic ventricular systolic ejection fraction was visually preserved at approximately 55%. Given the asymptomatic presentation, regular clinical, electrical, and echocardiographic follow-up was recommended.

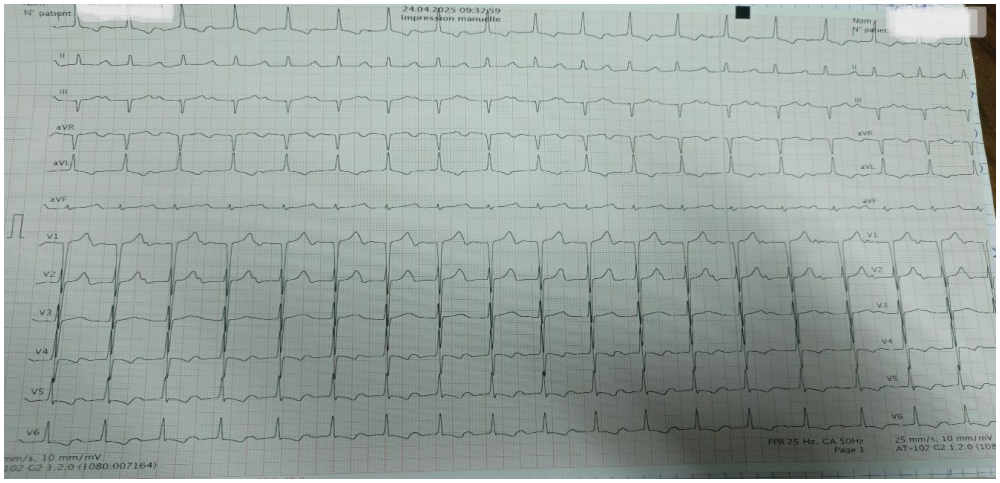

Figure 1: Electrocardiogram (ECG): Sinus rhythm, normal electrical axis, left atrial enlargement, first-degree atrioventricular block, and repolarization abnormalities in the apicolateral leads

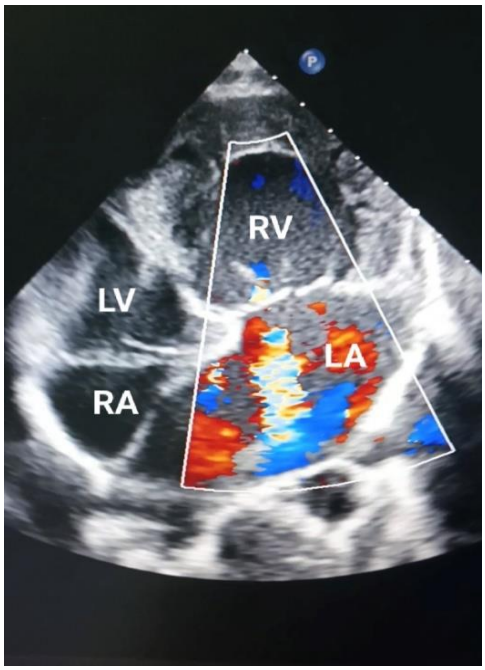

Figure 2a. Transthoracic echocardiography in the apical four-chamber view shows a morphologically right ventricle in the systemic position, with a mitral–tricuspid valve offset of 9 mm, moderate tricuspid regurgitation, and left atrial dilation

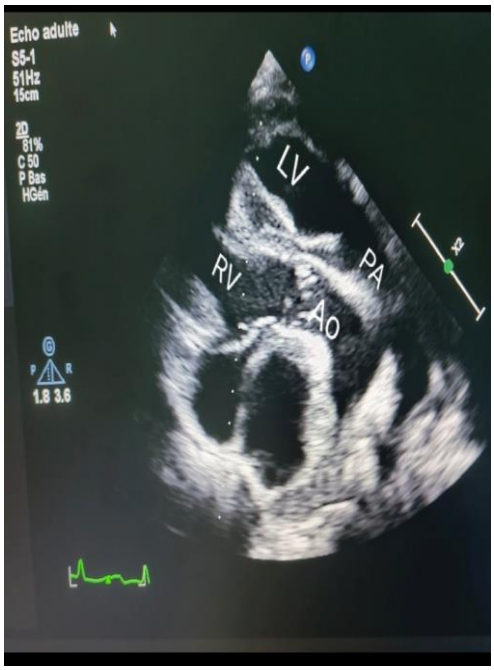

Figure 2b. Transthoracic echocardiography in the parasternal long-axis view demonstrates the aorta (Ao) arising from the systemic right ventricle (RV) and the pulmonary artery (PA) from the left ventricle (LV)

RV: right ventricle; LV: left ventricle; RA: right atrium; LA: left atrium.

3. DISCUSSION

Congenitally corrected transposition of the great arteries (CCTGA), also referred to anatomically as double discordance, is an extremely rare and complex congenital heart defect characterized by both atrioventricular and ventriculoarterial discordance. Despite the anatomical malalignment, the circulation remains physiologically corrected, which often delays diagnosis until adulthood, particularly in cases without associated anomalies [9].

Wissocque L *et al.,* [8] reported a case of a 92-year-old patient diagnosed at the age of 70, who remained clinically stable under medical management. The onset of heart failure and severe tricuspid regurgitation are key prognostic indicators that may significantly impact patient outcomes [13]. The right ventricle is more compliant than the left but less contractile due to a lower density of myocardial fibers. When positioned on the left and exposed to systemic circulation, it initially undergoes hypertrophy to overcome systemic afterload. However, over time, poor adaptation to systemic pressures leads to progressive dilation and gradual deterioration of its function. Tricuspid valve insufficiency may develop either due to annular dilation or inadequate adaptation to systemic circulation [9,13]. Transthoracic echocardiography is the first-line imaging modality for establishing the diagnosis [10]. However, due to the complex geometry of the right ventricle, systolic ejection fraction calculations based on volumetric methods such as Simpson's rule are unreliable. In such cases, cardiac magnetic resonance imaging (MRI) remains the gold standard [6,10]. When MRI is unavailable, the Tei index measured by echocardiography serves as a valuable alternative, offering a more accurate assessment of right ventricular function than Simpson's method [11]. Management of congenitally corrected transposition of the great arteries (CCTGA) is complex and often surgical, involving a double switch procedure, atrial and arterial preceded by pulmonary artery banding to train the left ventricle [6]. Due to suboptimal long-term outcomes, this approach is not routinely indicated [12]. In symptomatic patients with severe tricuspid regurgitation, early valve replacement is recommended before systemic ventricular function deteriorates [6]. In our patient, systemic ventricular function was preserved and mitral regurgitation was moderate. Management should also address associated complications such as conduction disorders and heart failure [7,9]. Medical therapy mirrors that for left ventricular failure, though its efficacy in right ventricular dysfunction remains uncertain; small studies suggest benefit, but randomized data are lacking [6,14]. Prognosis depends on systemic ventricular function, tricuspid valve integrity, and associated anomalies [6,15]. Even asymptomatic patients may experience progressive dysfunction. Graham et al. [15] found that 25% of patients over age 45 developed heart failure. Helsen *et al.,* [12] suggested that pulmonary outflow obstruction may protect against heart failure by reducing systemic AV valve regurgitation. In pediatric cases, Silvetti *et al.,* [7] showed that 3D-guided pacing can preserve systemic right ventricular function in patients with AV block. Prognosis worsens with advanced right ventricular dysfunction, severe tricuspid regurgitation, and complex congenital anomalies [15].

4. CONCLUSION

Although rare, congenitally corrected transposition of the great arteries (CCTGA) is a congenital heart defect with significant anatomical and therapeutic complexity. Diagnosis relies primarily on Doppler echocardiography, often delayed due to the absence of symptoms, particularly when systemic right ventricular function is preserved. In resource-limited settings, where access to advanced imaging and cardiac surgery is restricted, management is largely based on clinical and paraclinical follow-up, highlighting the importance of early detection and specialized referral.

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