When Double Discordance Hides Behind a Slow Heartbeat: A Case of ccTGA and Complete AV Block

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Résumé

Introduction: La transposition corrigée congénitale des gros vaisseaux (ccTGV) est une cardiopathie congénitale rare, représentant moins de 0,05 % des malformations cardiaques congénitales. Elle se caractérise par une double discordance atrioventriculaire et ventriculoartérielle, conférant une configuration anatomique particulière avec un ventricule droit morphologique assurant la fonction systémique. Cette anomalie est fréquemment associée à des troubles de conduction, notamment des blocs auriculo-ventriculaires (BAV), en raison de la position anormale du nœud AV et du trajet atypique du système de conduction. Nous rapportons le cas d'un patient de 41 ans, asymptomatique, chez qui un BAV complet a été découvert fortuitement. L'échocardiographie a confirmé le diagnostic de ccTGV avec fonction conservée du ventricule droit systémique. Bien que le patient remplissait les critères pour une stratégie conservatrice, une implantation prophylactique d'un pacemaker double chambre a été décidée, au vu des données de la littérature suggérant un risque non négligeable d'événements asystoliques soudains même en l'absence de symptômes. Nous discutons les défis diagnostiques et thérapeutiques posés par cette malformation rare et l'importance des stratégies de stimulation physiologique pour préserver la fonction ventriculaire systémique à long terme.

Mots-clés

congenital corrected transposition of the great vessels; atrioventricular blocks; echocardiography

Summary

Congenital corrected transposition of the great arteries (ccTGV) is a rare congenital heart disease, representing less than 0.05% of congenital heart defects. It is characterized by a double atrioventricular and ventriculoarterial discordance, conferring a particular anatomical configuration with a morphological right ventricle ensuring systemic function. This anomaly is frequently associated with conduction disorders, including atrioventricular blocks (AVB), due to the abnormal position of the AV node and the atypical course of the conduction system. We report the case of a 41-year-old asymptomatic patient in whom a complete AVB was discovered incidentally. Echocardiography confirmed the diagnosis of ccTGV with preserved systemic right ventricular function. Although the patient met the criteria for a conservative strategy, prophylactic implantation of a dual-chamber pacemaker was decided, given the literature data suggesting a significant risk of sudden asystolic events even in the absence of symptoms. We discuss the diagnostic and therapeutic challenges posed by this rare malformation and the importance of physiological pacing strategies to preserve long-term systemic ventricular function.

Keywords

congenital corrected transposition of the great vessels; atrioventricular blocks; echocardiography

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INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital cardiac malformation, accounting for less than 0.05% of all congenital heart diseases(1). Its incidence is estimated at approximately 1 in 33,000 live births. While the precise etiology remains unclear, the occurrence of familial cases suggests a possible genetic component.

Due to the abnormal anatomical positioning of the atrioventricular (AV) node and the atypical course of the conduction system, cardiac conduction disturbances-particularly AV blocks- are frequently observed in ccTGA (2). As survival rates of patients with congenital heart disease continue to improve, it is essential for clinicians to recognize ccTGA and anticipate its long-term complications.

This report aims to illustrate the clinical and imaging features of ccTGA complicated by complete AV block and to discuss the diagnostic and therapeutic challenges in light of current literature.

CASE PRESENTATION

A 41-year-old male with no significant medical history presented with epigastric discomfort.

On physical examination, he was hemodynamically and respiratorily stable, with a blood pressure of 120/68 mmHg, a bradycardic pulse of 45 beats per minute, and oxygen saturation of 98% on room air.

The electrocardiogram (EKG) revealed complete atrioventricular block with narrow QRS complexes and a ventricular rate of 45 bpm.

Blood tests were unremarkable except for elevated C-reactive protein (CRP) at 200 mg/L and leukocytosis with a white blood cell count of 13,000 /mm³.

Transthoracic echocardiography showed situs solitus with mesocardia (figure 1) and confirmed atrioventricular and ventriculoarterial discordance consistent with ccTGA. Specifically, the left atrium was connected via a tricuspid valve to a left-sided, trabeculated morphological right ventricle, which gave rise to the aorta—indicating a systemic right ventricle.

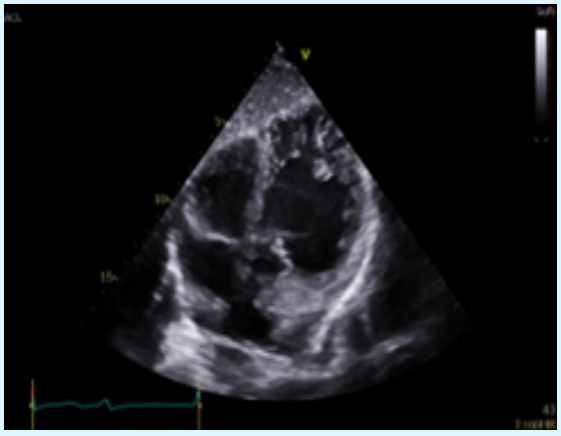

Figure 1. Mesocardia and double discordance, consistent with ccTGA

Systemic right ventricular function was preserved, with a TAPSE of 22 mm and a peak systolic velocity (S') at the tricuspid annulus of 15 cm/s (figure2).

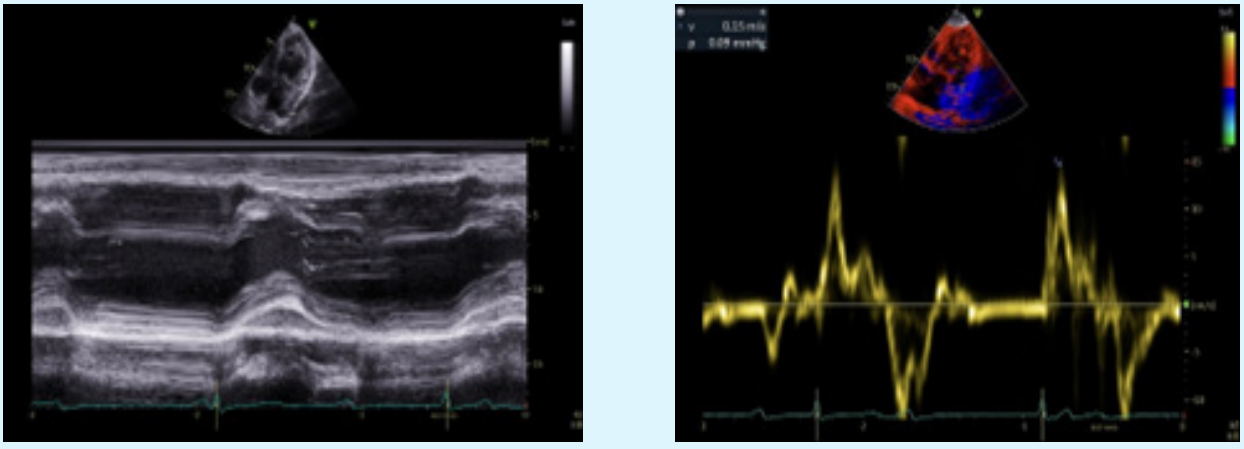

Figure 2. TAPSE measured at 21 mm and a peak systolic velocity (S') at the tricuspid annulus of 15 cm/s , confirming preserved systolic function of the systemic right ventricle

Global stain of 22.7% and a right ventricular fractional area change of 49.76% (figure3).

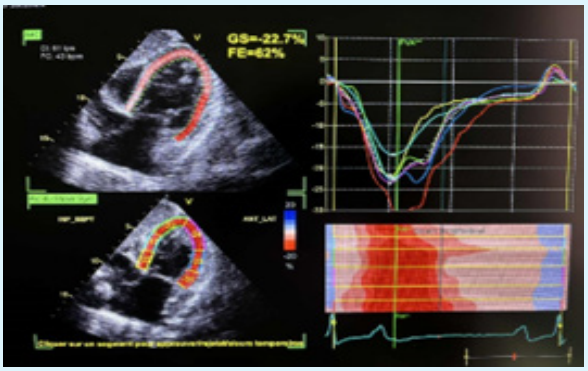

Figure 3. Global stain of 22.7% and a right ventricular fractional area change of 49.76%

The right atrium was connected via a mitral valve to a smooth-walled, right-sided morphological left ventricle, which supplied the pulmonary artery (figure 4).

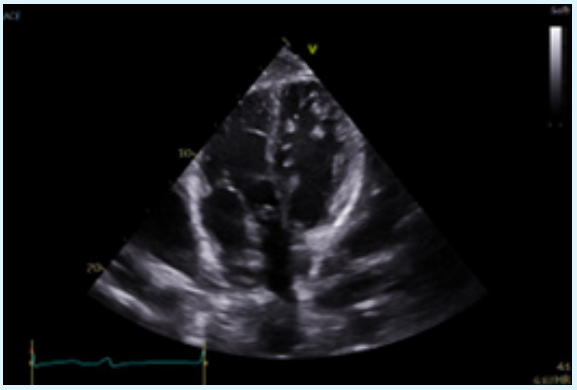

Figure 4. The pulmonary artery emerges from the morphological LVconfirming ventriculo-arterial discordance.

A mild form of Ebstein anomaly was noted, with apical displacement of the septal leaflet of the tricuspid valve measuring 12 mm (figure 5).

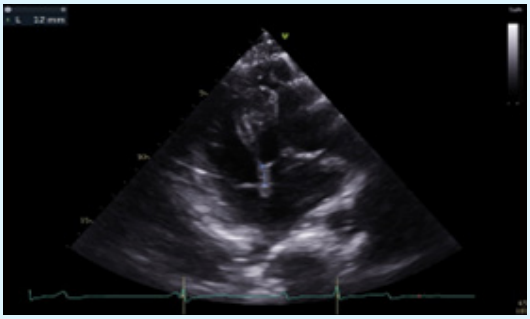

Figure 5. Apical displacement of the septal leaflet of the tricuspid valve, suggesting a mild Ebstein anomaly

Cardiac magnetic resonance imaging (MRI) was requested for a more accurate assessment of systemic right ventricular ejection fraction.

An abdominal CT scan, performed for the abdominal symptoms, revealed ileitis with no indication for urgent surgical intervention.

A 24-hour Holter monitor confirmed complete AV block with an average heart rate of 47 bpm, without significant ventricular ectopy or relevant pauses.

While the patient was initially asymptomatic with an adequate escape rhythm and no significant arrhythmias on Holter monitoring, the decision was ultimately made to proceed with permanent pacemaker implantation rather than pursue a conservative strategy and the patient was discharged with a plan for close clinical and echocardiographic monitoring, given the preserved function of the systemic right ventricle.

DISCUSSION

ccTGA is a rare congenital heart defect. Embryologically, it results from abnormal leftward looping of the primitive heart tube, leading to atrioventricular (AV) and ventriculoarterial (VA) discordance.

In this configuration of "double discordance," systemic venous return enters the right atrium and flows through the mitral valve into a morphological left ventricle, which ejects blood into the pulmonary artery. Pulmonary venous return then enters the left atrium, flows through the tricuspid valve into a morphological right ventricle, which acts as the systemic ventricle ejecting blood into the aorta.

This configuration may occur with either situs solitus or mirror-image atrial arrangements, and is frequently associated with other anomalies, including ventricular septal defects (70%), pulmonary stenosis (40%), and tricuspid valve abnormalities such as Ebstein-like malformations (3).

Our patient had situs solitus, mesocardia, and a mild form of Ebstein anomaly.

The diagnosis of ccTGA is often made incidentally in adulthood during investigations for unrelated symptoms or findings, such as murmurs or conduction disturbances.

Two major long-term complications may arise

1. Systemic right ventricular (RV) failure, as the morphological RV is not designed to sustain systemic pressure long-term

2. Cardiac conduction disorders

The abnormal course of the conduction system in ccTGA accounts for the high risk of conduction disturbances. In patients with situs solitus, the usual posterior AV node is hypoplastic due to septal misalignment. Instead, an anterior AV node develops near the orifice of the right atrial appendage, close to the fibrous continuity between the pulmonary and mitral valves.

This node gives rise to a long and vulnerable His bundle, which traverses the septum anteriorly beneath the pulmonary valve, making it highly susceptible to fibrosis and degeneration over time (2).

According to Huhta et al., the annual risk of progression to complete AV block is approximately 2%, reaching a prevalence of 30% in adulthood, as was the case in our 41-year-old patient (4).

The decision to implant a pacemaker in such cases depends on symptoms and escape rhythm characteristics. According to current guidelines, pacing is indicated in:

- Symptomatic complete AV block
- Mean daytime heart rate <50 bpm
- Pauses >3 times the basic cycle length
- Broad QRS escape rhythm
- Prolonged QT interval
- Complex ventricular ectopy (5)

In asymptomatic patients with stable escape rhythms (>40 bpm), no significant pauses, and preserved systemic RV function, a conservative strategy with close follow-up may be appropriate (6).

However, although our patient initially met these criteria — asymptomatic, stable escape rhythm at 47 bpm, no pauses or ectopy, and preserved systemic RV function — we elected to proceed with dual-chamber pacemaker implantation. This decision was supported by findings from a prospective study by Eriksson et al. (7), which demonstrated that even asymptomatic adults with congenital complete heart block remain at risk of sudden asystolic events, which can occur unpredictably and may be fatal from the first episode. The study also reported a progressive decline in ventricular rate over time, increased risk of morbidity, and the development of secondary mitral insufficiency. These findings support a prophylactic pacing approach in such patients, even in the absence of clinical symptoms.

However, if pacing becomes necessary, anatomical considerations specific to ccTGA must be addressed:

- The systemic ventricle is a morphological right ventricle located on the left side, making access appear reversed on imaging
- Coronary sinus anatomy may be atypical or absent
- Right ventricular apical pacing may worsen systemic RV function over time (2)

Physiological pacing strategies are increasingly being considered. Conduction system pacing (CSP) — including His bundle pacing (HBP) and left bundle branch area pacing (LBBAP) — can preserve ventricular synchrony and may help maintain long-term systemic ventricular function more effectively than traditional pacing (8).(9)

These techniques require specialized expertise and pre-procedural imaging due to anatomical variability.

Leadless pacemakers have also shown promise in ccTGA, especially when transvenous access is challenging or contraindicated (10) .

Although not yet widely implemented for all ccTGA patients, growing evidence supports their use in selected cases, particularly when preserving systemic RV function is a priority.

CONCLUSION

ccTGA is a rare but clinically significant congenital anomaly that may remain undiagnosed until adulthood, often revealed by conduction disturbances such as complete AV block. A thorough understanding of its unique anatomy and electrophysiological implications is essential for accurate diagnosis and optimal management. If pacing becomes necessary, physiological pacing strategies should be prioritized to preserve systemic ventricular function and improve long-term outcomes. Multimodal imaging and individualized follow-up are key to guiding management in this complex condition.

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