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CASE REPORT
Adult congenital heart disease

A case report of upgrading to cardiac resynchronization therapy in a patient with congenitally corrected transposition of great arteries and dextrocardia

Jakub Šimka *, Kamil Sedláček, Rudolf Praus, and Petr Pařízek

First Department of Internal Medicine, Cardiology and Angiology, University Hospital Hradec Králové and Charles University Faculty of Medicine in Hradec Králové, Czech Republic

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Background Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital heart anomaly. Physiological correction may be associated with a long pre-symptomatic period in many patients and delayed accidental diagnosis. Additional related congenital malformations may increase the complexity of cardiac interventions.

Case summary A 59-year-old man with known dextrocardia, situs viscerum inversus, and CCTGA was scheduled for upgrading of a dual-chamber pacemaker to cardiac resynchronization therapy to treat heart failure related to a progressive systolic dysfunction of the systemic right ventricle (RV). Because of the specific anatomy of this patient, the therapeutic procedure was complicated by the cannulation of the Marshall vein. Nevertheless, the left ventricular lead was successfully implanted into the coronary sinus lateral branch. At the 3-month follow-up, the patient remarkably reported a significant functional improvement, despite no favourable reverse remodelling of the systemic RV.

Discussion Upgrade of a pacemaker to biventricular pacing was feasible in this patient, who had CCTGA and dextrocardia, which resulted in symptomatic improvement at follow-up. Pre-implant contrast cardiac computed tomography angiography was essential for visualizing the venous-specific anatomy in this patient, who suffered from congenital heart disease. Conduction system pacing represents a potential alternative for the patient to prevent or treat pacing-related heart failure.

Keywords Cardiac resynchronization therapy • Heart failure • Congenitally corrected transposition of the great arteries • Dextrocardia • Case report

ESC curriculum 2.4 Cardiac computed tomography • 5.9 Pacemakers • 9.7 Adult congenital heart disease • 5.11 Cardiac resynchronization therapy devices

Learning points

- To understand the anatomy, conduction system abnormalities, and indications for cardiac resynchronization therapy (CRT) in patients with congenitally corrected transposition of the great arteries.
- To recognize some of the challenging technical aspects of CRT in patients with structural heart abnormalities.
- To recognize electrocardiogram interpretation in patients with dextrocardia.

* Corresponding author. Tel: +420 495 83 3249, Email: jakub.simka@fnhk.cz
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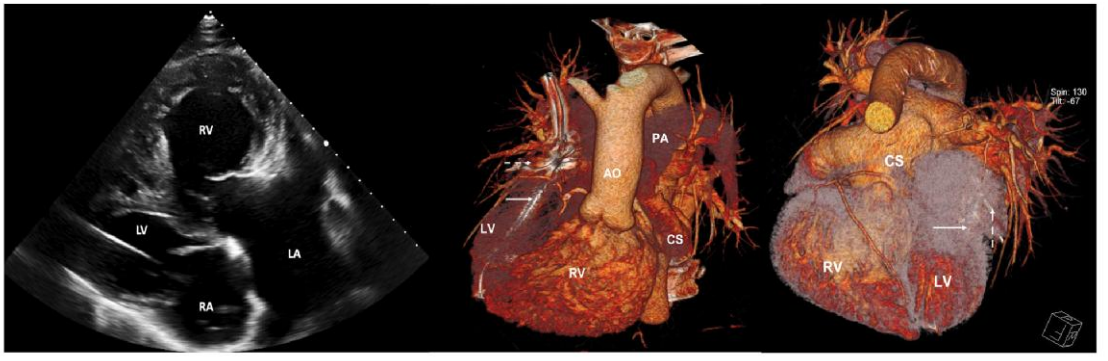
Introduction

Congenitally corrected transposition of the great arteries (CCTGA), also known as L-transposition, is a rare congenital heart defect (CHD) occurring in <1% of all CHD patients, with a prevalence of ~0.03 per 1000 live births.[1] CCTGA incorrectly connects the aorta to the right ventricle (RV), the pulmonary artery to the left ventricle (LV) (ventriculoarterial discordance), and the atria to the ventricles [atrioventricular (AV) discordance]. CCTGA is mostly accompanied by other congenital anomalies such as ventricular septal defect (70%), pulmonary valvular stenosis (40%), morphological tricuspid valve abnormalities (33%), dextro-/mesocardia (17%), or complete AV block (13%).[2] Patients with CCTGA may be asymptomatic for a long period in their lifetime and its detection may be accidental. CCTGA commonly manifests as arrhythmia, requiring pacing, or as systemic RV failure by the third or fourth decade of life.

Summary figure



AO = aorta, CCTGA = congenitally corrected transposition of the great arteries, CRT = cardiac resynchronization therapy, CS = coronary sinus, EF = ejection fraction, LA = left atrium, LV = left ventricle, PA = pulmonary artery, RA = right atrium, RV = right ventricle, white solid arrow = ventricular electrode, white dashed arrow = atrial electrode.

Case presentation

A 59-year-old man with known dextrocardia, situs viscerum inversus, and CCTGA was scheduled for upgrading of a dual-chamber pacemaker to cardiac resynchronization therapy (CRT) to treat heart failure related to a progressive severe systolic dysfunction of the systemic RV. In 1994, a dual-chamber pacemaker was implanted for a symptomatic Type 1 second-degree AV block. Subsequently, over the years, the patient suffered a heart failure as the AV block progressed and the ventricular pacing percentage increased above 95. At the time of upgrade, the systemic RV ejection fraction (EF) was 25%, as assessed by transthoracic echocardiography (TTE), and the patient was grouped under New York Heart Association (NYHA) functional Class III. The TTE findings included systemic RV hypertrophy, moderate aortic valve and morphological tricuspid valve regurgitation, and preserved subpulmonary LV EF. Coronary angiography excluded a coronary artery disease. Pre-implantation pharmacological treatment included perindopril, metoprolol, digoxin, eplerenone, and furosemide. On physical examination, the patient was haemodynamically stable, a systolic murmur Grade II/VI and a diastolic murmur were present above the right hemithorax, and no signs of systemic congestion were found. A computed tomography (CT) was performed to obtain an accurate anatomical CHD description and localization of the coronary sinus (CS) (*Figure 1*).

Upgrading to biventricular pacing was performed from the right side. The CS was cannulated using a decapolar catheter Livewire 6F deca XLRG (Abbott) and a right CPS Direct Universal RS slittable guide catheter (Abbott) in the posteroseptal right atrium where it is usually located. The first cannulated structure was the Marshall vein presenting with atrial electrograms, and the guidewire visibly penetrated beyond it into the pericardium with a small pericardial contrast leakage (*Figure 2A*). The guidewire was removed without sequelae. The CS guiding catheter was retracted to the CS ostium and a hydrophilic Guide Wire M (Terumo) was used to safely recannulate the CS and reposition the guiding CPS Direct catheter. An occlusive CS angiography revealed only one lateral branch with a short accessible segment for LV lead implantation (*Figure 2B and C*). With no transvenous alternatives available, we implanted a bipolar LV pacing electrode QuickFlex 1258T/86 (Abbott) into this branch with good pacing parameters. The QRS complex narrowed from 220 to 160 ms on biventricular pacing (*Figure 3C*).

At the three months follow-up visit, the patient reported a significant performance improvement and was in the NYHA class I-II. The narrowing of the QRS complex to 160 ms remained stable. However, systemic RV dysfunction persisted (EF 25%), which is illustrated in *Figure 4* and in Supplementary material (Supplementary material online, *Video S1* represents echocardiography pre-CRT, Supplementary material online, *Video S2* is post-CRT). The follow-up interrogation showed correct functioning of the CRT device, with a biventricular pacing of >99%

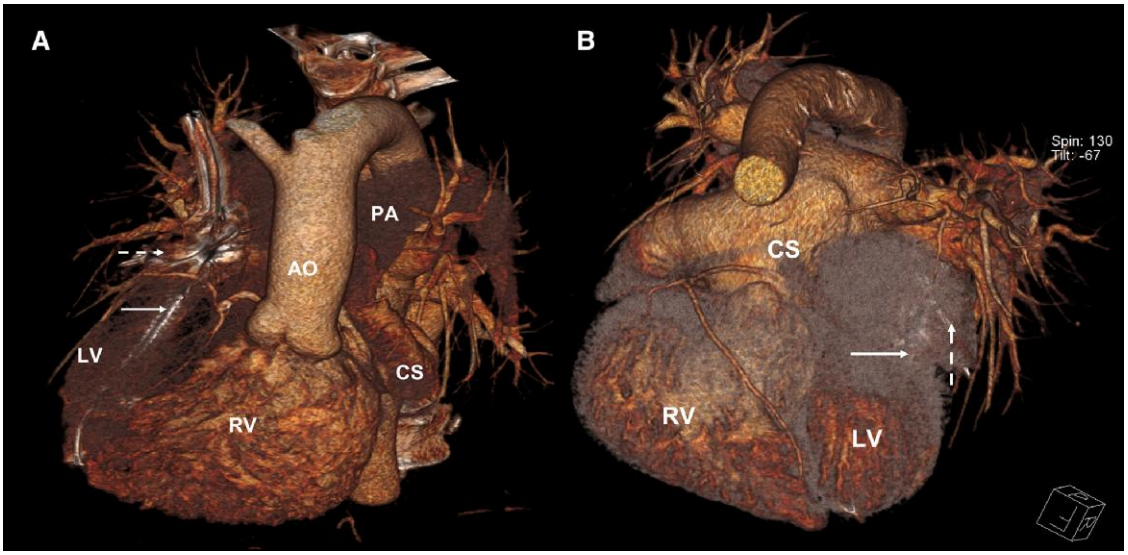


Figure 1 Three-dimensional reconstruction rendered from the electrocardiogram-gated contrast computed tomography of the heart in the arterial phase, demonstrating the anatomical condition in the patient with congenitally corrected transposition of the great arteries and dextrocardia. (*A*) The anterior position rotated by 28° up and by 23° to the left. (*B*) The posterior position rotated by 67° down and by 130° to the right. AO, aorta; CS, coronary sinus; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; white solid arrow, ventricular electrode; white dashed arrow, atrial electrode.

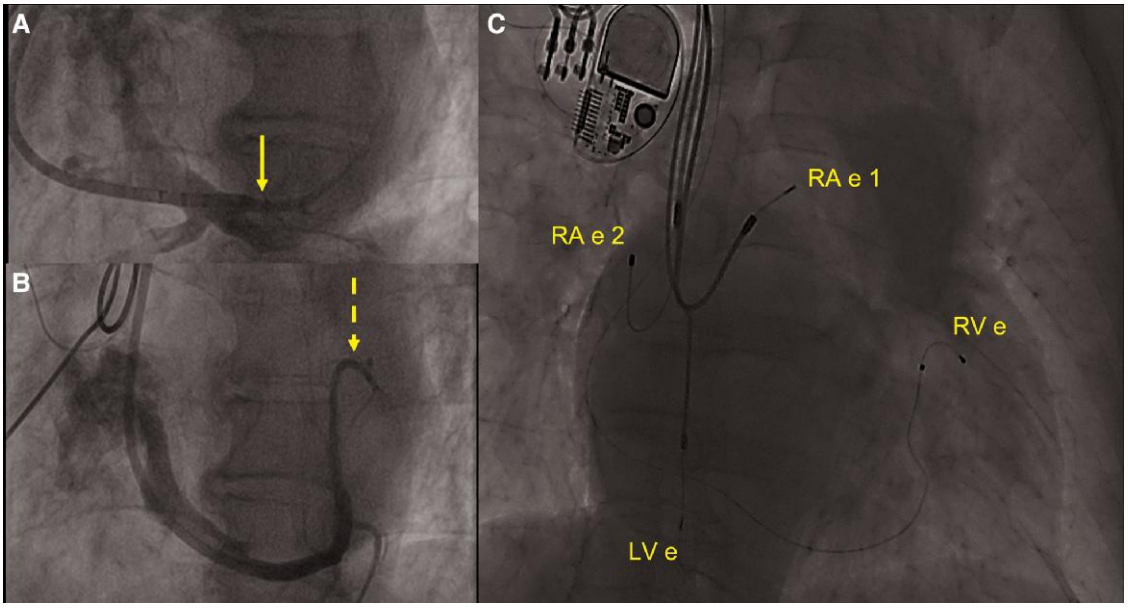


Figure 2 Implantation of a cardiac resynchronization therapy system. (*A*) Coronary sinus cannulation followed by occlusive venogram that clarifies the position in the Marshall vein (yellow solid arrow). (*B*) Venogram of the coronary sinus. Only one lateral branch is suitable for implanting the left ventricular electrode (yellow dashed arrow). (*C*) RAO projection after the completion of the procedure, a new bipolar systemic right ventricle electrode in the coronary sinus (RV e), and previously implanted devices: an original right atrium electrode implanted in 1994 (RA e 1), a right atrium electrode implanted in 2003 (RA e 2), and a subpulmonary left ventricle electrode implanted in 1994 (LV e).

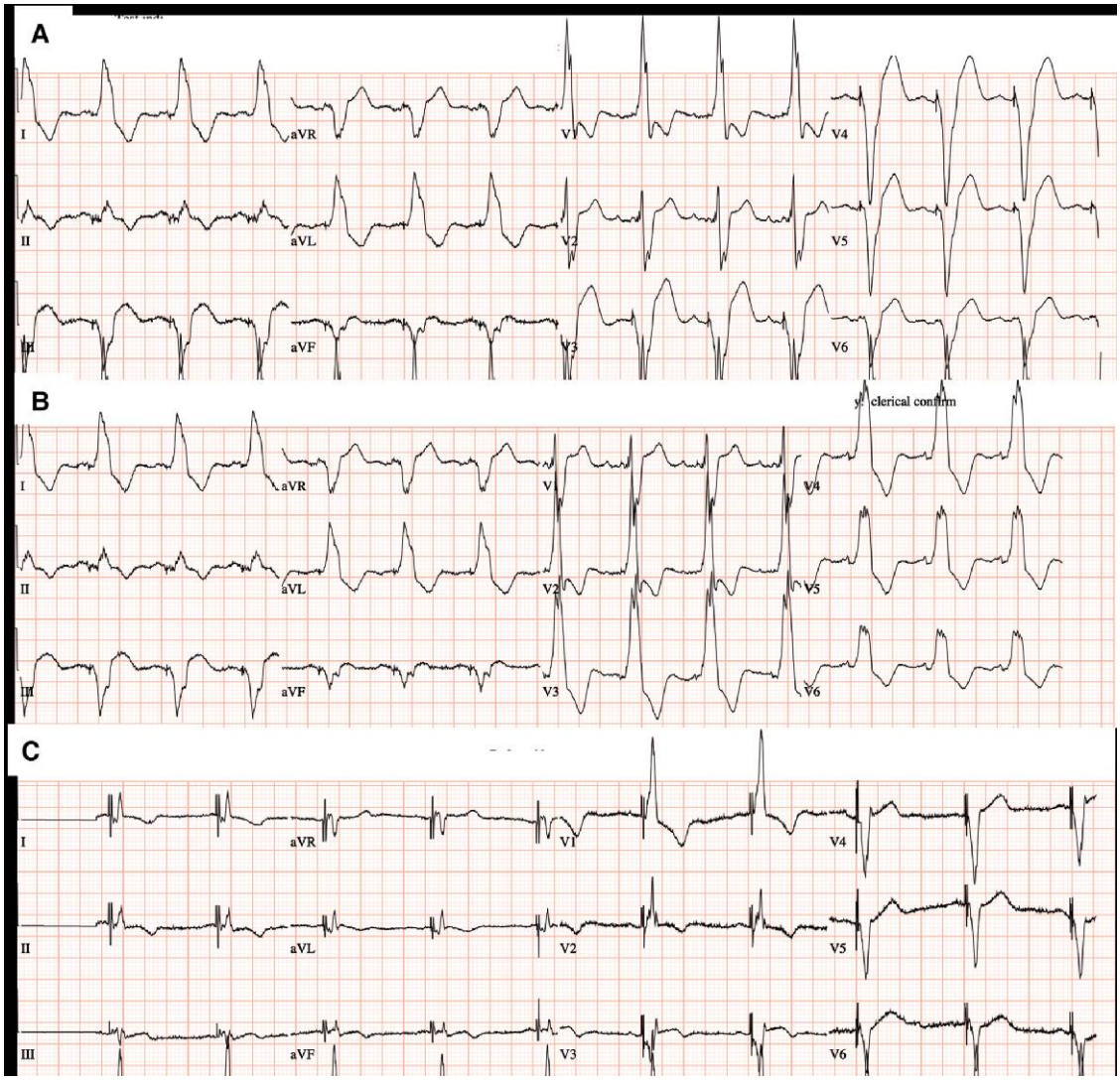


Figure 3 (*A*) Electrocardiogram levels recorded during pacing of the morphological left ventricle, standard placement of limb and chest leads, and a QRS duration of 220 ms. (*B*) Electrocardiogram levels recorded during pacing of the morphological left ventricle, standard placement of limb leads, chest leads placed inversely (V_1R–V_6R), and a QRS duration of 205 ms. (*C*) Electrocardiogram taken during biventricular pacing, standard placement of limb leads, reversed placement of chest leads, and a QRS duration of 160 ms.

and previously undetected repeated paroxysms of atrial fibrillation. Despite a CHA_2DS_2VASc score of 1 and HAS-BLED score of 1, anticoagulation therapy with apixaban at a twice daily full dose of 5 mg was started immediately, because the patient's weight was 86 kg and he had a normal creatinine range. The patient developed a severe ischaemic stroke on the same day because of the embolic occlusion of the right internal carotid artery and middle cerebral artery. The treatment for this included endovascular recanalization; despite this, he developed a moderate paresis of the left upper limb and mild paresis of the left lower limb. Amiodarone was added as part of the rhythm control strategy. A scheduled electrical cardioversion after transoesophageal echocardiography was performed by administering a synchronized shock of 300 J in the anteroposterior paddle placement with sinus rhythm restoration. The patient continues to be monitored by an expert for adult CHD, and perindopril was replaced with sacubitril/valsartan because of the persistence of systemic RV dysfunction. Depending on the development of his condition, the possibility of referral to the centre of heart transplant and placement of a ventricular assist device will be considered.

Discussion

We describe successful upgrading from a dual-chamber pacemaker to CRT in a patient with a combination of conditions such as CCTGA, dextrocardia, and systemic RV failure. To the best of our knowledge, only one report of a patient with similar conditions is available in the literature.[3]

In CCTGA, the conduction system is inverted and descends along the anterior part of the atrial septum into an elongated (and at times duplicated) AV node and then around the anterolateral segment of the morphological LV outflow tract diving into the anterior part of the interventricular septum, where it lies subendocardially on the right side.[4] Anatomical abnormalities may be responsible for a higher incidence of conduction disorders. The condition of 10–50% of patients

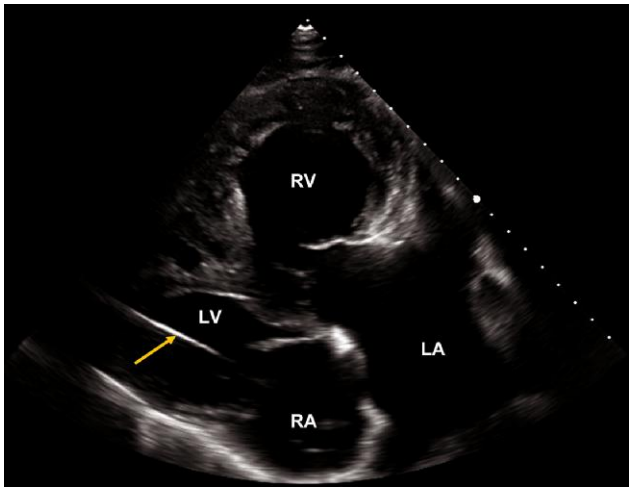


Figure 4 Transthoracic echocardiography after upgrading to cardiac resynchronization therapy and a modified apical four-chamber view from the right hemithorax. The objective parameters persisted despite the therapy. Thickness of the systemic right ventricle free wall 13 mm, right ventricle ejection fraction 25%, left ventricle ejection fraction 50%, moderate regurgitation of the morphological tricuspid valve, and mild regurgitation of the morphological mitral valve. LA, left atrium; LV, subpulmonary left ventricle; RA, right atrium; RV, systemic right ventricle; solid arrow, left ventricle electrode.

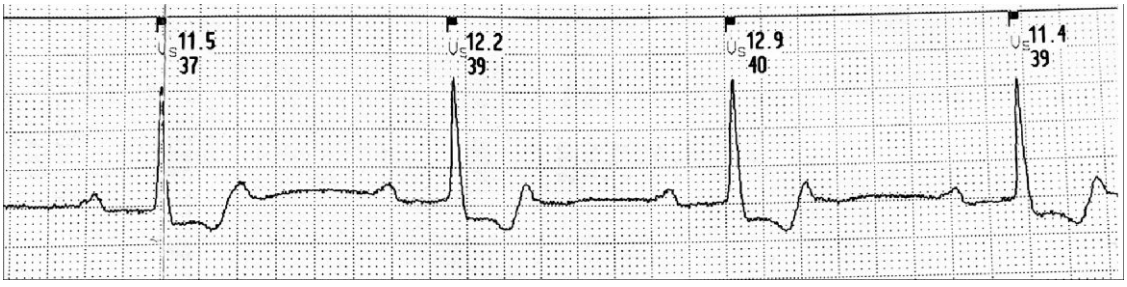


Figure 5 Electrocardiogram strip obtained during device pre-implant integration presenting a 2:1 second-degree atrioventricular block and a native narrow QRS complex.

with CCTGA may progress to a complete AV block with an incidence rate of 2% per year.[5] Many CCTGA patients, therefore, need ventricular pacing; however, this may contribute to the development of systemic RV dysfunction and heart failure by producing electro-mechanical dyssynchrony. Early electrical activation and mechanical contraction causes an initial stretching of late-activated segments, and then these segments contract against the early relaxed parts.[6] Thus, subpulmonary LV pacing may cause early septal activation, the contraction of which is not involved in systemic RV ejection, and secondary dilation of the systemic AV annulus with subsequent morphological tricuspid valve regurgitation.[4] The consensus statement of the Pediatric and Congenital Electrophysiology Society and the Heart Rhythm Society recommends upgrade to a CRT device when the systemic RV EF drops below 35% and ventricular pacing exceeds 40% for patients grouped under NYHA functional Classes I–IV.[7] The patient in this study reported a significant functional improvement 3 months after upgrading to CRT therapy despite no favourable reverse remodelling of the systemic RV. Similar improvements in the symptoms of at least one patient group in NYHA classification have been reported in previously published cases.[3,5] The reasons for the absence of echocardiographic remodelling in our patient were the presence of severe organic heart disease, long duration of RV dysfunction, and short procedural follow-up.

Cardiac resynchronization therapy implantations or upgrades are challenging procedures in patients with CCTGA. In ~25% of patients, the CS may be absent or unidentifiable and the coronary veins may have separate entries to the right or left atrium. Therefore, it is vital to perform an imaging study to determine the complete anatomy before starting the procedures. Both contrast cardiac CT and coronary angiography with late venous phase imaging of the CS provide useful overall cardiac and CS anatomy visualization.[8] The largest available study of patients with CCTGA demonstrated that the CS ostium was located in its normal position in 88% of the patients, and they had a normal anatomical structure.[9] Coronary sinus cannulation may be (and was in this case) complicated by the presence of the Marshall vein, and this complication occurs in ~23% of CCTGA patients.[9]

Nevertheless, a 95% success rate of transvenous CRT implantation has been previously documented.[10] In patients in whom it is impossible to perform CS cannulation or in those in whom the CS venous anatomy is unsuitable for such a procedure, surgical implantation of the systemic ventricular lead has been recommended.[8] Recent data demonstrated the feasibility of a permanent physiological pacing of the systemic ventricle by capturing the right bundle as a potential alternative to prevent or treat pacing-related heart failure, which would also be possible in our patient due to the presence of the narrow native QRS complex (*Figure 5*).[11] Despite adequate therapy to treat heart failure, there has been a reduction in the life expectancy of patients with CCTGA without any associated lesions over the years, and only ~50% of patients have survived at the age of 60 years.[12] Patients die from congestive heart failure or die suddenly, presumably due to ventricular tachycardia or ventricular fibrillation, regardless of the presence of the condition of advanced heart failure. Nevertheless, the benefit of implantable cardioverter-defibrillator therapy in primary prevention for single or systemic RVs has yet to be well established.[12]

Congenitally corrected transposition of the great arteries, in combination with dextrocardia, requires carrying out modifications of an electrocardiogram (ECG) recording and its interpretation. It is recommended to use reverse right-sided precordial leads from V1R to V6R in such patients.[13] The placement of limb leads is not unanimously defined, but reverse placement may also be preferable.[14] On a standard 12-lead ECG, CCTGA is associated with complex changes. Ventricular and conduction system inversion is associated with a reverse septal activation, *Q* waves in the right precordial leads, and absent *Q* waves in the left precordial leads. These features may be misinterpreted as anterior myocardial infarction.[15] The LV shift to the right side leads the reversal of the bundle branch block and paced QRS morphology (*Figure 3A* and *B*).

Conclusion

Despite the anatomical complexity and various technical challenges that we encountered in a patient with CCTGA and dextrocardia, who developed heart failure associated with systemic RV dysfunction, we successfully implanted the CRT device using standard tools and techniques.

Lead author biography

Dr Jakub Šimka graduated in 2018 at Charles University, Faculty of Medicine, in Hradec Králové. Currently, he is a resident cardiologist at University Hospital in Hradec Králové. His research focus is on arrhythmias, electrophysiology, and cardiac devices.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

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Consent: The authors confirm that written consent for submission and publication of this case, including images and associated text, has been obtained from the patient in accordance with COPE guidelines.

Conflict of interest: None declared.

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Data availability

The data underlying this article will be shared on reasonable request to the corresponding author.

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