European Heart Journal - Case Reports (2023) **7**, 1–5
https://doi.org/10.1093/ehjcr/ytad382

CASE REPORT
Electrophysiology

Cardiac resynchronization therapy using conduction system pacing after double-switch surgery for congenitally corrected transposition of the great arteries: a case report

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Received 3 March 2023; revised 12 July 2023; accepted 8 August 2023; online publish-ahead-of-print 11 August 2023

Background Patients with congenitally corrected transposition of the great arteries (ccTGA) are at risk of developing conduction disease and complete atrio-ventricular block and this risk increases after corrective cardiac surgery. However, the optimum pacing modality remains controversial.

Case summary Twelve years after a double-switch surgery with ventricular septal defect correction, a 16-year-old ccTGA female was referred with an indication for cardiac resynchronization therapy. In the absence of coronary sinus (CS) or direct access to the conduction system, several therapeutic options were considered. Finally, using a three-dimensional navigation system and customized sheaths, a left bundle branch area pacing (LBBAP) lead was successfully implanted. The implantation resulted in stable pacing parameters and positive haemodynamic changes. At 9-month follow-up, pacing parameters were stable and the patient reported a significant improvement in quality of life.

Discussion Cardiac resynchronization therapy in adults with repaired congenital heart disease remains challenging, especially in the absence of CS or direct access to the conduction system. In such a situation, LBBAP appears as an attractive alternative pacing modality. However, pre-operative management is critical to the success of the implantation.

Keywords Case report • Left bundle branch area pacing • Conduction system pacing • Cardiac resynchronization • Transposition of the great arteries • Pacemaker • ccTGA • Congenital heart disease

ESC curriculum 5.11 Cardiac resynchronization therapy devices • 9.7 Adult congenital heart disease

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Handling Editor: Michael Spartalis
Peer-reviewers: Linh Ngo; Luis Antonio Moreno-Ruiz
Compliance Editor: William Crawford

Learning points

- Benefit of physiological pacing in congenitally corrected transposition of the great arteries (ccTGA) patients with double-switch surgery by means of left bundle branch area pacing (LBBAP).
- In ccTGA patients who have undergone double-switch surgery, there is usually no direct access to the coronary sinus or the conduction system. Therefore, LBBAP represents a unique opportunity to deliver physiological pacing or cardiac resynchronization therapy (CRT).
- Compared with traditional pacing methods that can result in abnormal ventricular activation sequence and desynchrony, LBBAP preserves a physiological ventricular activation and may improve cardiac function in patients with CRT indication.
- Importance of detailed anatomical and electrical description before LBBAP implantation.
- In patients with congenital heart disease, venous access must be guaranteed before the procedure and venous stenting (if needed) should be performed before lead implantation.
- Mapping-guided approach to identify the best region for initial lead deployment and avoid scar/patch area greatly facilitates the implantation.
- Reshaping of commercially available sheaths is often mandatory. In the future, the use of personalized tools and/or custom-made three-dimensional models may help to optimize LBBAP implant procedure in these patients.

Introduction

Patients with congenitally corrected transposition of the great arteries (ccTGA) often develop conduction disease and complete atrio-ventricular (AV) block because of developmental anatomical anomalies. AV block also frequently occurs after corrective cardiac surgery, particularly in the setting of ventricular septal defect (VSD) closure.[1,2] The optimum pacing modality for permanent pacemaker implantation in such cases remains controversial. Although pacing-induced ventricular dyssynchrony may play an important role in the functional deterioration of ccTGA patients,[3] cardiac resynchronization therapy (CRT) by means of biventricular pacing or His bundle pacing remains challenging in this population, particularly after surgical anatomical correction (double-switch surgery) when there is typically no direct access to the conduction system or to the distal coronary sinus (CS). Recently, left bundle branch area pacing (LBBAP) has emerged as a promising alternative to deliver physiological pacing.[4,5] In this case report, we describe the implantation of a new endocardial lead in the LBBAP position in a 16-year-old ccTGA female referred for CRT.

Summary figure

2008	Congenitally corrected transposition of the great artery—double-switch surgery
2020	Paroxysmal complete atrio-ventricular block—implantation of dual chamber pacemaker connected to epicardial leads (atrial lead on the anterior wall of the atrium, ventricular lead on morphologically right ventricle). Ventricular pacing was associated with a paced QRS of 220 ms and very poorly tolerated
10 February 2020	Deterioration in functional status and exercise capacity with a marked impact on activities of daily living. New York Heart Association (NYHA) Class III. Reprogramming of the device in VVI back-up 40′ (ventricular pacing 0.1%)
15 December 2021	Patient referred for cardiac resynchronization therapy

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30 March 2022	Pre-operative evaluation identifies a significant stenosis of the superior vena cava at the junction with the baffle. Implantation of a stent.
16 May 2022	Implantation of a new endocardial lead in the left bundle branch area. After procedure, paced QRS was 130 ms
30 June 2022	Significant functional improvement. Patient back to school and to fitness activities
15 February 2023	The 9-month follow-up demonstrates maintenance of a small-paced QRS (120 ms), ventricular pacing of 44%, and improved quality of life (NYHA Class I)

Clinical vignette

We report a 16-year-old ccTGA female who presented with symptomatic episodes of intermittent complete AV block 12 years after a double-switch surgery with VSD correction. Because of the complexity of the anatomy, she was initially implanted with a dual-chamber pacemaker connected to epicardial leads. Unfortunately, right ventricular (RV) pacing was extremely poorly tolerated and associated with a significant drop in mean blood pressure (>30 mmHg). This severely impacted her quality of life with a cessation in all physical activity in the year post implantation [ventricular pacing 100%, New York Heart Association (NYHA) Class III], necessitating reprogramming of the device in VVI back-up mode 40 b.p.m. Ultimately, given the paced QRS duration and echocardiographic indices of pacing-induced dyssynchrony, she was referred for CRT upgrade. Several options were considered, including (i) the addition of a new left ventricular epicardial lead for biventricular epicardial pacing, (ii) the addition of a new endocardial lead positioned deep in the middle cardiac vein to allow for more septal epicardial pacing, and (iii) the addition of a new endocardial lead in LBBAP position.

The patient was extremely reluctant to undergo a new thoracotomy, and a pre-procedural contrast-enhanced cardiac computed tomography demonstrated no suitable middle cardiac vein or CS for lead implantation as well as a mild stenosis (with a luminal diameter of 7 × 9 mm) at the junction of the superior vena cava and the baffle. Accordingly, implantation of a new endocardial lead in the LBBAP position was planned 1 month after stenting of the stenosis. The patient had an additional diagnosis of primary ciliary dyskinesia for which she was taking antibiotic prophylaxis, along with 100 mg aspirin post

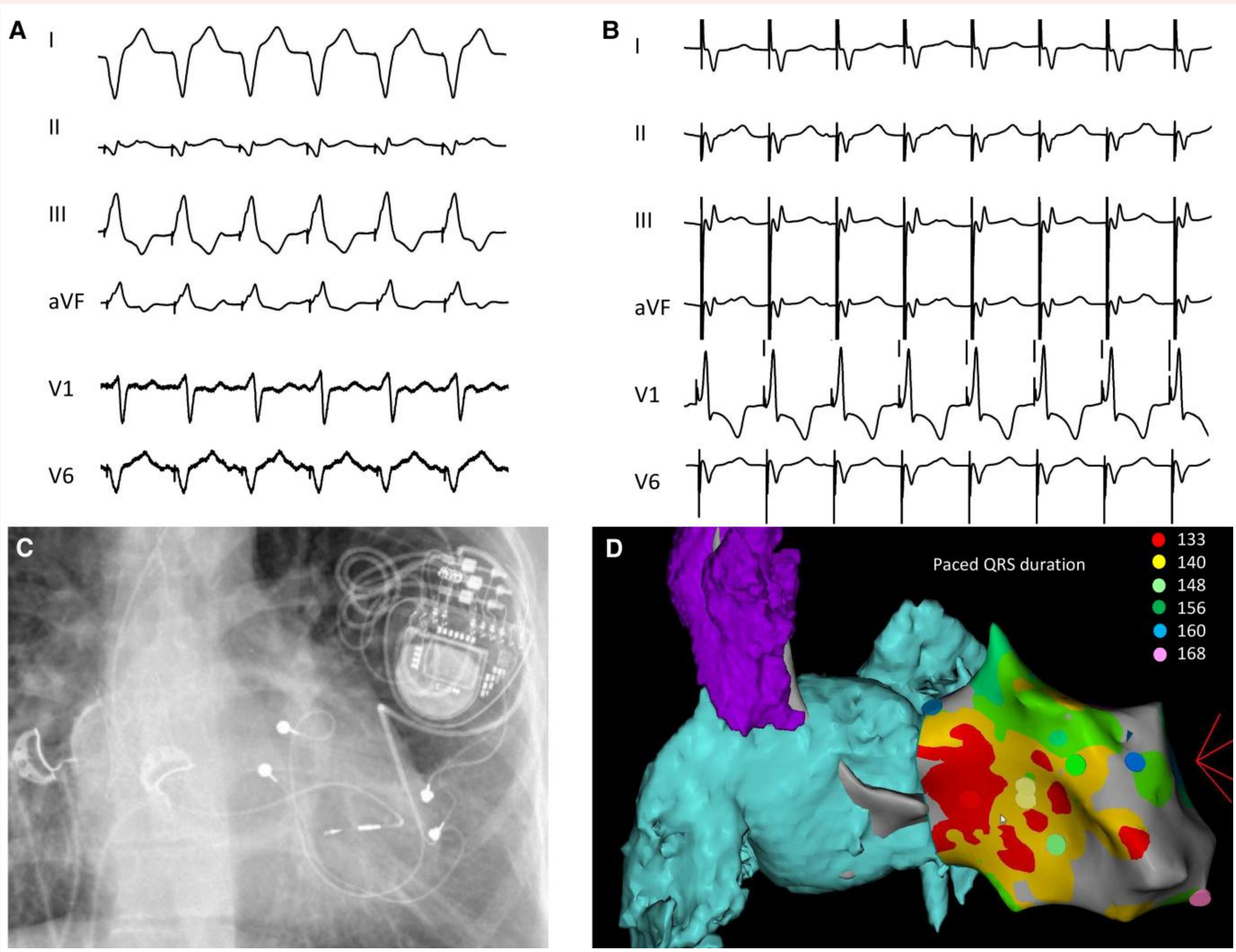

Figure 1 Electrocardiogram recordings before (*A*) and after left bundle branch area pacing (*B*). Chest X-rays (*C*) and electroanatomic map (*D*). Baseline electrocardiogram (*A*) during ventricular pacing (dual chamber mode) from the epicardial electrode. Electrocardiogram (*B*) obtained immediately after left bundle branch area pacing implant (VVI—unipolar). Note the significant reduction of the paced QRS duration (from 220 to 130 ms) and the short V6 S-wave peak time (80 ms) mirroring left ventricular activation time. (*C*) Antero-posterior chest X-ray after the implant procedure showing the pacing lead implanted in the middle of the interventricular septum. (*D*) Electroanatomical three-dimensional maps of the morphological right ventricle (in grey), the superior vena cava (purple), and the baffle (light blue). The colour scale in the right ventricle represents paced QRS duration according to lead position. The red area represents the targeted implant site with the shortest QRS. LBBAP, left bundle branch area pacing.

implantation of the stent. Upon physical examination on admission, no abnormalities were detected. Pre-procedure echocardiography revealed a status post Senning's correction and arterial switch procedure (morphological RV in the left position and morphological left ventricle in the right position), normal ventricular function, a VSD patch, mitral regurgitation (2/4), and a superior vena cava stent.

Under general anaesthesia, using the CARTO XP system and a deflectable EP catheter (Biosense Webster Navistar 4 mm, Johnson & Johnson Medical, New Brunswick, NJ, USA), a comprehensive three-dimensional (3D) electroanatomical map of the RV was created from the subclavian vein. Sites of earliest activation during conducted sinus rhythm and an area of low-voltage corresponding to the region of VSD closure were identified. In addition, a map of the QRS duration during RV endocardial pacing was created. The initial site for transseptal screwing of the lead was determined using the following criteria: (i) local voltage >5 V, (ii) within a region of early activation and (iii) associated with a relatively short-paced QRS duration. Two customized CS delivery sheaths [(i) outer sheath, multipurpose long, and (ii) 130°—curved inner sheath] shortened to accommodate the length of the Medtronic select secure 3830 lead were used to facilitate close contact with the septum of the morphological RV. After identification of the site of implantation, the lead was implanted by rapid clockwise rotation to allow for deep penetration of the helix into the myocardial septum. Rotations were interrupted after the occurrence of relatively narrow premature QRS complexes on the surface electrocardiogram. Conduction pacing capture was further suggested by the presence of a transition from non-selective to selective LBBAP during threshold testing by a relatively narrow paced QRS duration (130 ms) and by a short S-wave peak time (mirroring the conventional R-wave peak time in structurally normal hearts) in leads V5 and V6 (<90 ms) (*Figure 1*). Immediately after implant, continuous ventricular pacing was well tolerated and was associated with positive haemodynamic changes. No new treatments were initiated at the time of discharge. At 9-month follow-up, pacing parameters were stable, with only 44% burden of ventricular pacing and a paced QRS of

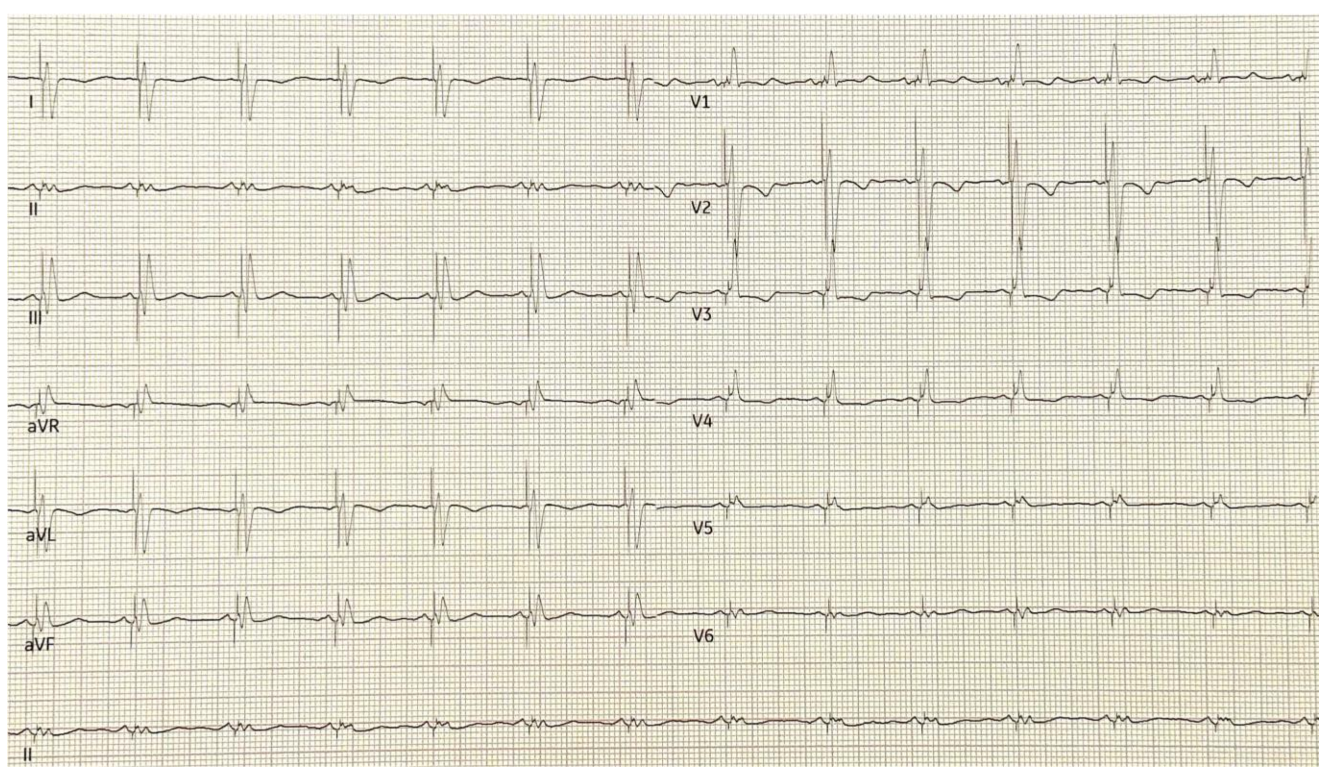

Figure 2 Electrocardiogram at 9 months follow-up. Paced QRS is 120 ms.

120 ms (*Figure 2*). The patient reported a significant improvement in quality of life with NYHA Class I and stable parameters on echocardiography.

Discussion

Although current AHA/ACC and ESC guidelines do not recommend CRT in patients with systemic RVs [dextro-transposition of the great arteries (D-TGA) with previous atrial switch repair and ccTGA], growing evidence from case series and individual reports suggest substantial benefits.[6] The present case report further demonstrates potential benefits from physiological pacing in this very specific population, even after corrective (double switch) surgery. As a result of recent technical development, conduction system pacing has attracted significant interest in patients with grown-up congenital heart disease (CHD). In a recent case series, O'Connor *et al.*[7] reported on 39 patients with CHD undergoing LBBAP (2 D-TGA patients with prior atrial switch). Despite inherent difficulties relating to anatomy, the overall success rate was 96%. Nevertheless, some methodological and technical aspects warrant consideration when performing LBBAP in the ccTGA population. First, as highlighted by our case, the patency of venous access must be ascertained pre-procedure (using venous angiography), and stenting, if needed, should be performed before any endovascular lead implantation. Secondly, a mapping-guided approach to identify the optimum region for lead deployment and to avoid zones of surgical correction (VSD in the present case) may greatly facilitate a successful implant. Finally, reshaping of commercially available sheaths is often mandatory to access the implant site. The use of personalized tools and custom-made 3D models generated from computed tomography may help optimize the procedure in the future. Some potential complications of LBBAP also exist. In CHD patients more specifically, the unpredictable course of the conduction system might increase the risk of procedural failure. Although speculative, the risk of lead failure might also be higher because of mechanical constraints resulting from the tortuous course of the lead. Finally, the risk of venous occlusion and/or thrombosis of the baffle should also be taken into consideration.

Conclusion

This case report is the first to document the use of LBBAP as a CRT bailout strategy in a ccTGA patient after double-switch surgery. In addition to the specific technical challenges posed, it highlights the potential benefits of physiological pacing in this unique population.

Lead author biography

Benjamin De Becker is an electrophysiology and pacing fellow in the Department of Cardiology, AZ Sint-Jan Hospital, Bruges, Belgium.

Consent: The authors confirm that written consent for submission and publication of this case report, including image(s) and associated text, has been obtained from the patient in line with COPE guidance.

Conflict of interest: Over the past 2 years, J.-B.L.P.D.W. received non-significant compensation for proctoring and teaching activities from Medtronic, Biotronik, Boston Scientific, and Johnson & Johnson. Other authors declare no conflict of interest.

Funding: None declared.

Data availability

The authors confirm that the data supporting the findings of this study are available upon request.

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