Cardiac resynchronization therapy-defibrillator implantation guided by electroanatomic mapping in a young adult patient with congenital heart disease

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1. Introduction

Despite advances in medical therapy, heart failure (HF) continues to be the main cause of morbidity, hospitalizations and mortality in the adult congenital heart disease (ACHD) population [1]. Cardiac resynchronization therapy-defibrillator (CRT-D) implantation has become an established therapeutic option for adult patients with congenital heart disease (CHD), bundle branch block, reduced ejection fraction and symptoms of heart failure. A new implantation approach guided by the electroanatomic mapping (EAM) has been developed to overcome some issues of the standard technique: non-responder patients, high x-ray exposure and use of iodinated contrast medium for coronary sinus angiography. This approach has not been previously described in the CHD population. We report a case of EAM-guided approach for CRT-D implantation in a young adult patient with CHD.

2. Case report

A 19-year-old young man was admitted to our ACHD ward because of exertional dyspnea. He was diagnosed prenatally with transposition of the great arteries (TGA) and ventricular septal defect (VSD) by foetal echocardiography. After birth, Rashkind procedure was performed within the first 24 hours of life. At 7 days of age, he underwent arterial switch operation with atrial septal defect (ASD) and VSD closure. Periodic outpatient follow-up after surgical repair was unremarkable. At the age of 4, he developed a LV enlargement with LBBB and reduced contractility. Therefore, he was admitted again to the Paediatric Cardiology Unit to undergo cardiac catheterization, that showed increased LV dimensions, reduced LV ejection fraction (LVEF 43%), no residual shunts and coronary arteries without anomalies of ostia and course. In the following years, he was asymptomatic on therapy with carvedilol, enalapril and diuretics. At the last clinical assessment at our ACHD outpatient clinic, he complained of fatigue and moderate exertional dyspnea, consequently he was admitted to our ACHD ward for further medical testing. Physical examination and laboratory test
values on admission were unremarkable. ECG showed sinus rhythm with PR interval near the upper range (180 ms), wide QRS (160 ms) and LBBB (Fig. 1A). Transthoracic echocardiogram (TTE) revealed a severe LV dilation with severe systolic dysfunction and mechanical dyssynchrony (left ventricular end-diastolic diameter [LVEDD] 70 mm, left ventricular end-diastolic volume [LVEDV] 327 ml, left ventricular end-systolic volume [LVESV] 236 ml, LVEF by Simpson’s biplane method 28%). Aortic root was dilated (46 mm) with mild aortic regurgitation. Right ventricular (RV) function and diameters were normal. No residual shunts were visualized. Invasive coronary angiography showed no significant stenosis or other anomalies.

Cardiac magnetic resonance (CMR) confirmed TTE findings (LVEDV 318 ml, LVESV 223 ml, LVEF 30%). No significant abnormalities of tissue characterization suggestive of myocardial infiltration, prior myocardial infarction, or fibrosis, were found before and after administration of gadolinium-based contrast. Considering the symptoms and the reduced EF despite best tolerated medical therapy, the ECG features and the other findings, we decided to implant a CRT-D.

The procedure was performed with the support of the EnSite Precision mapping system (Abbott). First, the RV active-fixation lead (Plexa ProMRI, Biotronik) was positioned from the left subclavian vein on the interventricular septum, to avoid risk of atrioventricular block caused by mechanical bump on the atrioventricular node during catheter placement into the CS. A fixed-curve decapolar diagnostic catheter was used to create a 3D anatomic reconstruction of the right atrium, to cannulate the CS ostium and to guide the delivery system insertion. Then, the catheter was pulled and an insulated guidewire (connected in a unipolar fashion to the EAM system) was introduced to start navigation into the vein and the collateral branches, reconstructing 3D anatomy and recording local unipolar electrical signals (Fig. 2). EAM allowed us to avoid contrast dye injection and fluoroscopy during CS cannulation and exploration. The most delayed activation time was recorded in a posterior branch, where the electrical delay was 130 ms from the onset of spontaneous QRS, and there the LV lead (Sentus ProMRI, Biotronik) was placed over the wire. The final lead position was verified using a few seconds of fluoroscopy. The atrial active-fixation lead (Solia S53, Biotronik) was positioned in the right atrium. We used short time of fluoroscopy for atrial and RV lead positioning, to reduce total procedure time (138 minutes, with 8 minutes of fluoroscopy). Finally, the three leads were connected to the device (Acticor 7 HF-T, Biotronik) in the infraclavicular pocket. Electrical parameters (sensing, pacing threshold and impedance) were satisfactory. Chest x-ray demonstrated good placement of the leads and the device without complications (Fig. 1B). Post-procedural ECG (Fig. 1C) showed QRS narrowing from 160 ms to 120 ms. At 6-month follow-up, the patient was asymptomatic. We repeated TTE: an improvement of LV contractile function and dyssynchrony (LVEDD 63 mm, LVEDV 314 ml, LVESV 165 ml, LVEF by biplane method 48%) was evident.

3. Discussion

CRT-D implantation may not significantly improve the ventricular function in about one-third of patients, and this proportion may be even higher in patients with CHD [1]. Several authors have demonstrated that LV lead position is one of the most important determinants of failure or success of resynchronization [2–4]. Electrical pacing from the latest activation site of the left ventricle has been related to a greater ventricular contractility improvement and reverse remodeling [6]. The latest activation site in patients with HF and LBBB is often the LV posterior-lateral region, which is therefore the preferred site to place the LV catheter. Unfortunately, LV lead positioning is the most challenging step during device implantation. Standard implantation technique is guided by angiography with contrast medium infusion, choosing the best site for the LV catheter according to fluoroscopic anatomy of the CS. The new non-angiographic technique by using EAM may be slower than conventional approach during first procedures but allows electrophysiologists to choose not only the best anatomic position but also the best electrical site, to reach the highest electrical resynchronization from the LV pacing [2–5].

We have reported a case of CRT-D implantation guided by EAM in a young patient with corrected CHD and systemic left ventricle. EAM allowed us to place the LV lead at the site where the latest electrical delay from spontaneous QRS onset was recorded (130 ms). LV electrical delay >50% of the QRS duration has been associated with improved outcomes in patients with HFpEF and electrical dyssynchrony [7]. We obtained in this case an electrical delay >50% (81%) from QRS onset and CRT resulted in QRS narrowing with a significant improvement of LV function at 6-month follow-up. Other factors can influence the probability of response after CRT implantation in ACHD patients, such as the amount of surgical scars, fibrosis or necrosis of the myocardial tissue, the underlying anatomy and pathophysiology (systemic left ventricle, systemic right ventricle, functionally single systemic ventricle) and the type of electrical conduction delay. An initial LV remodeling was

Fig. 1. (A) ECG on admission. Sinus rhythm with wide QRS and LBBB. (B) Chest x-ray. Final position of the leads and the device. (C) QRS narrowing after CRT-D implantation. Evidence of q wave in lead I and r wave in V1.
observed in our patient, but longer follow-up is warranted to demonstrate a significant remodeling using this approach.

EAM can help to mitigate the anatomical difficulties encountered in patients with complex CHD. The ventricular relationship (consequently, the CS location) and the atrioventricular relationship are normal in D-TGA, so EAM was not used to overcome specific anatomic issues in this case. Even if our patient did not have an unusual CS anatomy or complex atrial anatomy, ACHD patients with systemic left ventricle are known to fare better than other types of congenital patients and they might benefit from this approach not only to obtain a higher response from CRT but also to reduce radiation exposure and to avoid contrast-induced kidney injury [2−5]: ACHD patients have often impaired renal function (up to 30–50% of patients [8]) and undergo several radiologic examinations and cardiac catheterizations using contrast media over the course of their life.

This approach might have some potential limitations. A complete reconstruction of the CS anatomy could not be obtained because the guidewire could not show the presence of further branches, in addition to main ones, with better features. Another limitation could be the increased cost of the procedure than the conventional technique. However, the avoidance of contrast medium and the reduction in x-ray exposure, with the possibility to optimize resynchronization may justify this approach. Comparative studies are necessary to determine the benefits of this technique over the standard approach in the congenital population.

In conclusion, EAM-guided approach for CRT-D device implantation can provide a feasible option in adult patients with CHD, HFrEF and LBBB, in order to avoid contrast dye injection, to reduce x-ray exposure to the patients and to the operators and to increase probability of high response from resynchronization.

**CRediT statement**

G.D. Ciriello: writing, review and editing. D. Colonna, E. Romeo and B. Sarubbi: review.

**Declaration of competing interest**

All authors have no funding, competing interests or financial relationships to disclose.

**Acknowledgments**

We thank Dr. Valerio Langella for his valuable assistance.

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