CASE REPORT

10.2478/AMB-2026-0073

CARDIAC RESYNCHRONIZATION THERAPY IN A PATIENT WITH TOTAL CORRECTION OF TETRALOGY OF FALLOT AND RIGHT VENTRICULAR LEAD DYSFUNCTION

S. Yovev[1], I. N. Dimitrova[1], P. Simeonov[1], V. Groudeva[2*]

[1]Department of Cardiology, "Sv. Ekaterina" University Hospital – Sofia, Medical University – Sofia, Bulgaria
[2]Diagnostic Imaging Department, "Sv. Ekaterina" University Hospital – Sofia, Medical University – Sofia, Bulgaria

Abstract. *Adults with congenital heart lesions constitute a rapidly growing group of patients with cardiovascular disease, with patients with surgically corrected Tetralogy of Fallot forming one of the most complex groups in terms of long-term therapeutic management. Although it can be surgically repaired, Tetralogy of Fallot remains a progressive condition in which heart failure becomes the main determinant of long-term clinical outcomes and mortality. Cardiac resynchronization therapy has a well-established role in the treatment of left ventricular systolic dysfunction with interventricular dyssynchrony, either due to left bundle branch block or due to chronic right ventricular pacing. Here, we present a case of a 52-year-old male patient with Tetralogy of Fallot who underwent radical surgical correction. The patient experienced the typical progression of conduction abnormalities associated with this defect, requiring permanent pacemaker implantation. Because of confirmed septal dyskinesia and borderline left ventricular function (Left Ventricular Ejection Fraction of 45%), the patient was referred for cardiac resynchronization therapy pacemaker. Precise anatomical assessment of the new pacing system and postoperative status was assessed by multislice cardiac computed tomography. This case illustrates the critical transition from conventional pacing to cardiac resynchronization therapy pacemaker. During the 8-month follow-up, achieving electromechanical synchrony in both the left ventricle and right ventricle was confirmed, resulting in significant clinical improvement.*

Key words: *adults with congenital heart disease, cardiac resynchronization therapy, cardiac computed tomography*

Corresponding author: *Violeta Groudeva, MD, PhD, Diagnostic Imaging Department, "Sv. Ekaterina" University Hospital, Medical University – Sofia, Bulgaria, email: violetagroudeva@gmail.com*

ORCID: *0000-0001-7416-1550*

Received: *14 March 2026;* ***Accepted:*** *01 June 2026*

INTRODUCTION

In modern cardiology, the population of adults with congenital heart disease (ACHD) represents a growing proportion, with patients who have undergone correction of Tetralogy of Fallot (ToF) forming one of the most complex groups in terms of long-term clinical and therapeutic management. Despite its possible surgical treatment, Tetralogy of Fallot remains a progressive condition in which sudden death and progressive heart failure are predominant causes of death [1].

Epidemiological data indicate that between 20% and 25% of patients with repaired ToF develop clinical manifestations of heart failure by the fifth decade of life. This process results from a complex hemodynamic evolution dominated by right ventricular (RV) dilation and dysfunction, which is observed in more than 40% of patients in the long run. Progressive decompensation often also affects the left ventricle (LV) (10-15% of cases), as a consequence of unfavourable mechanical interactions between the two ventricles (interventricular dependence). In this phenomenon, severe RV dilation leads to paradoxical displacement of the interventricular septum, disrupting the geometry and filling of the left ventricular cavity [2, 3].

In parallel with the hemodynamic changes, electrical remodeling is an almost universal finding in ACHD-ToF. Complete right bundle branch block (RBBB) occurs in 80% to 90% of operated patients as a direct result of surgical resections in the RV outflow tract or stretching of the conductive system. A critical moment in the therapeutic pathway is the development of indications for cardiac pacing, with around 10% of patients becoming pacemaker-dependent due to AV block or sinus node dysfunction. However, prolonged conventional pacing may further exacerbate mechanical dyssynchrony [4]. Development of LV dysfunction after permanent pacemaker implantation is a known effect. Studies show that early activation of collagen synthesis after permanent pacemaker implantation is a prerequisite for myocardial collagen deposition, resulting in LV remodeling [5].

Recent studies show that 5% to 8% of adults with ToF require transition to cardiac resynchronization therapy (CRT). Although CRT is traditionally associated with correction of left bundle branch block (LBBB), in this specific population, it is applied as a "bi-ventricle-oriented resynchronization". The primary goal is to overcome the extreme electrical delay in the right ventricle and optimize global cardiac performance [6].

The present clinical case illustrates precisely this critical phase in the life of an ACHD patient. Given borderline LV function and progressive electromechanical dyssynchrony, upgrading to CRT-P is a strategy to prevent terminal heart failure. Achieving synchrony addresses both the residual hemodynamic consequences of ToF and the negative effects of long-term conventional pacing.

CASE PRESENTATION

We present the case of a 52-year-old man admitted for a scheduled intervention on his permanent cardiac pacing system. The patient belongs to the growing cohort of adults with ACHD, with a clinical history marked by several complex surgical interventions. His first intervention was performed at the age of 1- relief of right ventricular outflow tract (RVOT) obstruction, followed by radical total correction of ToF at the age of 26. Because of intermittent complete AV block and syncopal symptoms, a permanent dual-chamber pacemaker (DDD mode) was implanted at the age of 32. The current hospitalization is prompted by the telemetric detection of a high-pacing threshold in the right ventricular lead. The patient reported feeling generally well, without chest pain or resting dyspnoea upon admission. Electrocardiogram (ECG) on admission – P-synchronized DDD pacing rhythm – was recorded over an underlying persistent third-degree AV block (native rate 30 bpm). The ECG showed intrinsic atrial activity with signs of P-pulmonale, consistent with right atrial overload. The ventricular paced complex was markedly prolonged (> 160 ms), with a morphology resembling a complete LBBB, typical of long-term right ventricular apical pacing. These findings demonstrated severe electromechanical dyssynchrony, serving as the primary pathophysiological substrate for the patient's septal dyskinesia and reduced left ventricular function. Echocardiography revealed septal dyskinesia induced by chronic pacing. The LV had normal volumes (end diastolic volume – EDV 125 mL), but borderline ejection fraction (EF 45%). The RV shows a residual outflow tract gradient (32 mmHg) after prior RVOT repairs. A small, hemodynamically insignificant residual ventricular septal defect (VSD) was also described. The patient was referred for CRT. The clinical decision to upgrade to CRT-P in this patient is based on consolidated recommendations as of 2025, emphasizing prevention of further ventricular remodelling. The interventional procedure was performed under local anesthesia. Due to multiple prior procedures, an ipsilateral right-sided venous access was selected for upgrading the system. Pre-procedural venography demonstrated venous occlusive disease, manifested as subtotal occlusion of the right brachiocephalic vein (Figure 1).

Successful passage through the stenotic segment was achieved using a hydrophilic guidewire, followed by the placement of an introducer system for coronary sinus (CS) cannulation. The coronary sinus CS was catheterized, and retrograde venography visualized a scarce lateral venous network with a single small posterolateral branch (Figure 2).

Due to the small calibre of the posterolateral vessel, a bipolar LV lead (Quickflex micro 86) was successfully implanted with optimal parameters: threshold 2.0 V/0.5 ms and sensing 10 mV (Figure 3).

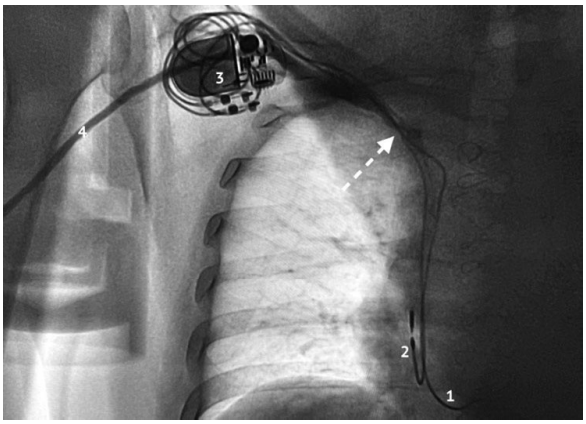

Fig. 1. Fluoroscopy, ipsilateral access. The components of the existing cardiac implantable electronic device and the venous pathology. White dotted arrow – occlusion of v. brachiocephalica dextra. 1) position of RV lead passing through the stenotic venous segment toward the right heart; 2) position of RA lead; 3) old two-chamber dual chamber pulse generator in the left peritoneal area; 4) proximal segment of the venous access via v.cephalica

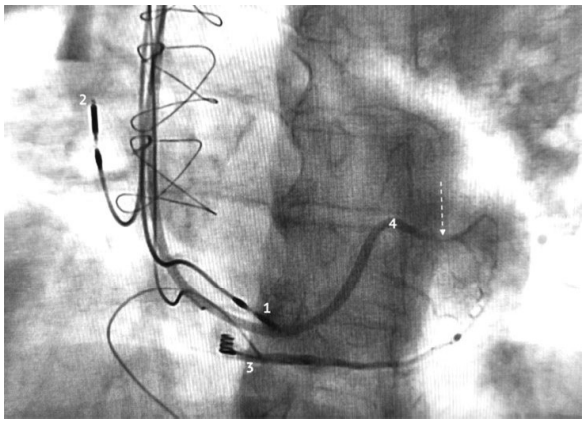

Fig. 2. Retrograde venography of the coronary sinus: 1) position of RV lead; 2) position of RA lead; 3) old epimyocardial lead from prior surgical correction of ToF; 4) introducing system positioned in the coronary sinus and subselector (Selectra) intubating posterolateral venous branch; white dotted arrow showing target posterolateral venous branch with small caliber and absence of larger lateral branches

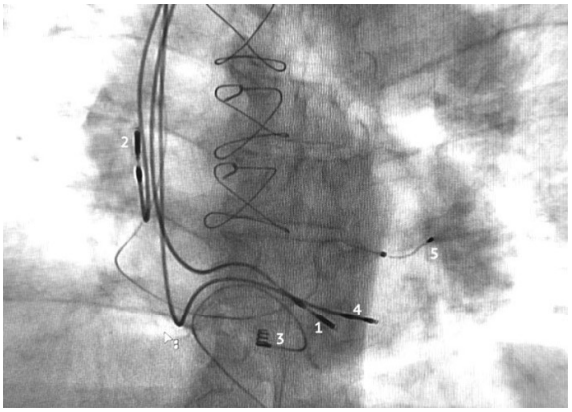

Fig. 3. Final configuration of the CRT-P system. Follow-up chest x-ray demonstrating upgrade and positioning of all components: 1) new RV lead with active fixation; 2) RA lead; 3) old epimyocardial lead; 4) old RV lead, abandoned in situ due to high threshold of stimulation and capped off with silicon cap; 5) bipolar LV lead placed in the posterolateral venous branch

Due to dysfunction of the old RV lead, a new active-fixation RV lead was implanted and stably positioned with optimal parameters (Figure 3). The system was upgraded to CRT-P using a Biotronik Enitra 8 HF-T pulse generator. Effective biventricular pacing was achieved, with significant narrowing of the QRS complex and correction of the previously documented electromechanical dyssynchrony. Regular follow-ups demonstrated sustained improvement in clinical condition and stability of the implanted system. The achieved QRS narrowing was maintained (from 180 ms to 130 ms). Biventricular pacing remained effective with no episodes of loss of capture or lead displacement, including the bipolar LV lead positioned in the small posterolateral branch. Left ventricular ejection fraction (EF) remained stable at 45-48%, with slight improvement and complete resolution of the initial septal dyskinesia. Clinically, the patient remained stable, in NYHA functional class I–II. At 8-month telemetry follow-up, all three active leads demonstrated stable stimulation thresholds. High-frequency ventricular pacing was well tolerated without inducing arrhythmias. For precise anatomical assessment of the new pacing system and postoperative status, a cardiac multislice computed tomography (MSCT) with ECG-gating was performed. Post-procedural MSCT confirmed the technical success of the intervention and detailed the complex anatomy of the patient. Three endocardial leads were visualized traversing the superior vena cava. The LV lead was clearly identified in the target venous branch, adjacent to the circumflex artery (Figures 4–6).

Old epicardial leads remained in situ. The interventricular septum was intact, with calcifications in the region of the prior patch. The RV outflow tract measured 28 mm at its narrowest point. Typical post-ToF repair findings were present: dilation of the aortic root (44 mm) and of the left pulmonary artery (37 mm). The right-sided chambers were not dilated. LV myocardial thickness measured 8.5 mm with preserved wall motion and no perfusion defects. CT coronary angiography showed a proximal mixed LAD plaque causing borderline stenosis (50–60%), with no significant disease in the remaining coronary vessels.

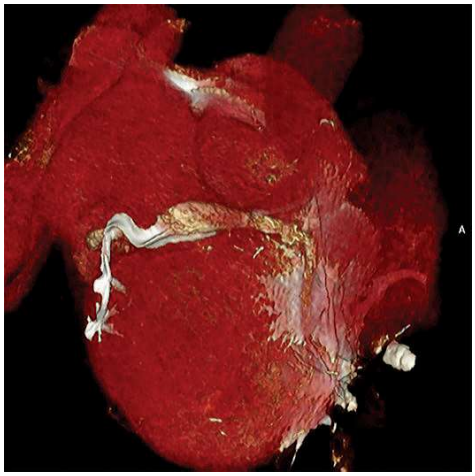

Fig. 4. Cardiac CT with ECG gating, volume-rendered images demonstrating 3D anatomy of the coronary sinus and venous vessels

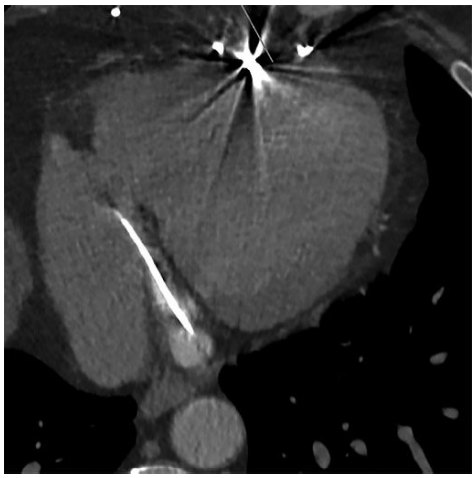

Fig. 5. Cardiac CT with ECG gating, image through the base of the heart demonstrating the lead in the coronary sinus

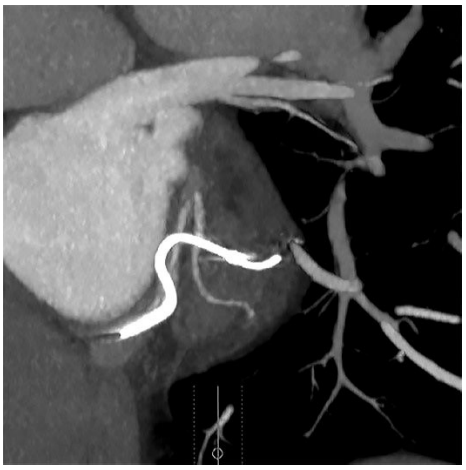

Fig. 6. Cardiac CT with ECG gating, reformatted image demonstrating the lead placed through the coronary sinus in the posterolateral branch and its position in relation to the left circumflex artery and OM coronary arterial branch

DISCUSSION

The management of ACHD, particularly those after repair of ToF, represents an increasing challenge in modern cardiology. CRT in the CHD population and ToF, in particular, continues to be a challenge [7]. The literature is scarce regarding reported cases of CRT implantation in ACHD and, particularly, regarding surgically repaired Fallot. Predominantly, single clinical cases are reported [8]. A few studies with a small number of patients have also been published. Gatzuolis et al in 1995 identified 9 patients with sustained ventricular arrhythmias and 4 with Sudden Cardiac Death (SCD) who all had a QRS measurement ≥ 180 ms, and therefore suggested "a potentially causative relation between RV dilation (represented by pronounced intraventricular conduction delay) and life-threatening arrhythmias." [9].

The primary indications for CRT are well established in the ESC guidelines [10]. The main indication is severe electromechanical dyssynchrony (QRS > 150 ms). The presence of a wide-paced QRS complex with LBBB-like morphology and echocardiographic septal dyskinesia is a strong predictor of positive hemodynamic response to resynchronization therapy. Also, an upgrade for pacing-induced cardiomyopathy is a Class I, Level B indication. According to ESC guidelines [10], in patients with a high percentage of ventricular pacing (> 20-40%) who develop left ventricular dysfunction with EF ≤ 45%, as in our case, transition to CRT is indicated. The aim is to reduce the risk of heart failure hospitalizations. Also, there are ACHD-specific considerations. According to recommendations, indications for CRT in ToF extend beyond those for idiopathic or ischemic cardiomyopathy. In adults with repaired Tetralogy of Fallot (ACHD-ToF), biventricular pacing

is increasingly recognized as the “gold standard” for optimizing interventricular interaction. Achieving ventricular synchrony is crucial for compensating for right ventricular dysfunction. In this patient, the leading indications were extreme QRS prolongation (160 ms), symptomatic heart failure (NYHA III), and the need for system upgrade due to ventricular dyssynchrony. Literature recognizes QRS ≥ 160 ms in ToF as a strong predictor of SCD and RV dysfunction, making resynchronization a pathophysiology-grounded intervention [6]. And, last but not least, technical necessity and system optimization are another indication. The documented dysfunction of the RV lead (high pacing threshold) mandates surgical revision of the system. In the context of 2025 standards, a routine generator/lead replacement without addressing dyssynchrony would be considered suboptimal clinical practice.

MDCT with ECG synchronization is emerging as a critical tool in the ACHD population. Advances in MDCT technologies allowed for high spatial resolution, rapid image acquisition, and, using standardized imaging protocols, high-quality images are obtained, which have contributed to the wide adoption of the modality into clinical practice in different clinical scenarios. Modern techniques include dual-source, wide-detector CT and dual-energy CT, and recently, the photon-counting technique has been introduced for cardiac imaging [11].

As a result, a high-image quality cardiac CT scan can provide detailed information not only on atrial and ventricular anatomy, cardiac function, and assessment of the coronary arteries, but also on myocardial tissue and myocardial perfusion [12].

The benefit of cardiac CT in CRT patients has been proven before [12, 13]. By assessing the functional and anatomical parameters of the cardiac chambers, MDCT can identify heart failure in patients. The most important advantage of MDCT, however, is the ability to demonstrate the coronary sinus and cardiac vein anatomy and thus to guide the procedure. Variations of the coronary venous system, such as anomalous coronary sinus ostium or course, venous anatomy of lateral branch, small caliber branches, or tortuous veins, can be analyzed, as well as the presence of valves. The accuracy of CT in demonstrating the venous branches is similar to that of retrograde balloon angiography [14].

In this case, MDCT provided essential information in three key areas: venous status verification, spatial orientation and stability, and coronary assessment and prognosis. MDCT confirmed the condition of the venous system after lead placement through the subtotal occlusion of the right brachiocephalic vein, demonstrating that the new leads did not further compromise lumen patency. Visualization of the LV lead adjacent to the circumflex artery alleviated concerns regarding potential mechanical conflict and confirmed optimal positional stability. Identification of borderline LAD stenosis (50-60%) added prognostic relevance, reinforcing the need for close ischemic risk monitoring while optimizing ventricular synchrony with CRT-P.

The limited venous anatomy and small calibre of the posterolateral branch required the use of a specialized sub-selection system. An alternative strategy – epicardial LV lead implantation via thoracotomy – was considered technically unfavourable due to severe pericardial adhesions from prior surgeries. The absence of phrenic nerve stimulation in a bipolar (ring-to-tip) configuration further confirmed the success of the chosen approach.

While CRT for ACHD and specifically for ToF repair is increasingly discussed in the literature, it remains technically challenging and relatively rare. CRT shows promise as a potential treatment option for patients with CHD and ventricular impairment, but larger clinical outcome studies are required before definitive guidance can be issued [7].

Although CRT in ToF is less common than in acquired cardiomyopathies, available data show significant improvement in hemodynamic and functional capacity. The study of Thambo et al. supports the benefit of CRT in improving the functional capacity of adult patients with ToF [15]. The 8-month follow-up in our case demonstrated sustained benefit – QRS narrowing to 130 ms and stable LV mechanics. The restored synchronous ventricular contraction prevented further cardiac remodelling – an important achievement in a patient with ToF within this 8-month window. This case is notable due to the combination of venous occlusive disease, which is a common complication in patients with long-standing intravascular leads, limited venous anatomy, and concurrent coronary atherosclerosis (LAD stenosis) confirmed postoperatively. Contemporary Europace publications support the feasibility of using modern sub-selection systems to avoid high-risk epicardial re-operation in heavily scarred pericardial environments.

MDCT is also used after implantation, with data suggesting assessment of CRT response by assessing the impact of LV wall remodelling [16]. Also, the use of MSCT for verifying the spatial relation between the LV lead and the circumflex artery represents an innovative method for ensuring long-term safety and myocardial perfusion in ACHD patients.

CONCLUSION

This clinical case illustrates the complex approach needed when implanting a CRT-P system in the setting of severe venous pathology and atypical anatomy and confirms that CRT-P upgrade is an effective and safe strategy in complex ACHD patients. Despite technical challenges related to the venous system and the presence of ischemic substrate, achieving electromechanical synchrony (QRS narrowing from 160 to 130 ms) resulted in significant clinical improvement. The case highlights the value of postoperative MSCT imaging as a standard for verification and risk management in complex electrophysiological interventions.

Conflict of Interest Statement: *The authors declare no conflicts of interest related to this work.*

Funding: *The authors did not receive any financial support from any organization for this research work.*

Ethical statement: *This study has been performed in accordance with the ethical standards as laid down in the Declaration of Helsinki.*

Consent for publication: *Consent form for publication was signed by the patient and collected.*

REFERENCES

1. Diller GP, Giardini A, Dimopoulos K, et al. Predictors of morbidity and mortality in contemporary Fontan patients: results from a multicenter study including cardiopulmonary exercise testing in 321 patients. Eur Heart J, 2010, 31(24):3073-83. doi: 10.1093/eurheartj/ehq356.
2. Davlouros PA, Kilner PJ, Hornung TS, et al. Right ventricular function in adults with repaired tetralogy of Fallot assessed with cardiovascular magnetic resonance imaging: detrimental role of right ventricular outflow aneurysms or akinesia and adverse right-to-left ventricular interaction. J Am Coll Cardiol, 2002, 40(11):2044-52. doi: 10.1016/s0735-1097(02)02566-4.
3. Geva T, Sandweiss BM, Gauvreau K, et al. Factors associated with impaired clinical status in long-term survivors of tetralogy of Fallot repair evaluated by magnetic resonance imaging. J Am Coll Cardiol, 2004, 43(6):1068-74. doi: 10.1016/j.jacc.2003.10.045.
4. Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease: developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD). Heart Rhythm, 2014, 11(10):e102-65. doi: 10.1016/j.hrthm.2014.05.009.
5. Yoncheva I, Biserov D, Negreva M. Permanent pacemaker implantation: early post-implantation data. Acta Med Bulg, 2024, 51(1):8-16. https://doi.org/10.2478/amb-2024-0002
6. Janousek J, Gebauer RA, Abdul-Khaliq H, et al. Working Group for Cardiac Dysrhythmias and Electrophysiology of the Association for European Paediatric Cardiology. Cardiac resynchronisation therapy in paediatric and congenital heart disease: differential effects in various anatomical and functional substrates. Heart, 2009, 95(14):1165-71. doi: 10.1136/hrt.2008.160465.
7. Merchant FM, Kella D, Book WM, et al. Cardiac resynchronization therapy in adult patients with repaired tetralogy of Fallot and left ventricular systolic dysfunction. Pacing Clin Electrophysiol, 2014, 37(3):321-8. doi: 10.1111/pace.12284.
8. Iovev S, Chilingirova N, Atzev B. CRT implantation in a patient with congenital heart malformation. Acta Med Bulg, 2019, 46(3):44-45. https://doi.org/10.2478/amb-2019-0029
9. Gatzoulis MA, Till JA, Somerville J, Redington AN. Mechanoelectrical interaction in tetralogy of Fallot. QRS prolongation relates to right ventricular size and predicts malignant ventricular arrhythmias and sudden death. Circulation, 1995, 92(2):231-7. doi: 10.1161/01.cir.92.2.231.
10. Glikson M, Nielsen JC, Kronborg MB, et al. ESC Scientific Document Group. 2021 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy. Eur Heart J, 2021, 42(35):3427-3520. doi: 10.1093/eurheartj/ehab364.
11. Si-Mohamed SA, Boccalini S, Lacombe H, et al. Coronary CT Angiography with Photon-counting CT: First-In-Human Results. Radiology, 2022,303(2):303-313. doi: 10.1148/radiol.211780.
12. Gerrits W, Danad I, Velthuis B, et al. Cardiac CT in CRT as a Singular Imaging Modality for Diagnosis and Patient-Tailored Management. J Clin Med, 2023, 12(19):6212. doi: 10.3390/jcm12196212.
13. Behar JM, Rajani R, Pourmorteza A, et al. Comprehensive use of cardiac computed tomography to guide left ventricular lead placement in cardiac resynchronization therapy. Heart Rhythm, 2017, 14(9):1364-1372. doi: 10.1016/j.hrthm.2017.04.041
14. Ricapito Mde L, Conde D, Theriault MM, et al. Multidetector cardiac tomography: a useful tool before cardiac resynchronization therapy. Cardiol J, 2015, 22(5):590-6. doi: 10.5603/CJ.a2015.0011.
15. Thambo JB, De Guillebon M, Xhaet O, et al. Biventricular pacing in patients with Tetralogy of Fallot: non-invasive epicardial mapping and clinical impact. Int J Cardiol, 2013, 163(2):170-4. doi: 10.1016/j.ijcard.2011.06.005.
16. Galand V, Ghoshhajra B, Szymonifka J, et al. Computed tomography-guided assessment of response to cardiac resynchronization therapy. JACC Clin Electrophysiol, 2019, 5(8):987-989. doi: 10.1016/j.jacep.2019.05.002.