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TECHNICAL ISSUES

Three-dimensional electroanatomical mapping guided right bundle branch pacing in congenitally corrected transposition of great arteries

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Aims The ideal pacing strategy has been the Achilles' heel for patients with congenitally corrected transposition of great arteries (ccTGA) with bradycardia. Various pacing modalities were documented in the literature. This article describes a novel pacing strategy and its feasibility in ccTGA with an intact ventricular septum.

Methods and results We prospectively recruited three patients with ccTGA who presented with symptomatic complete heart block to our institute and were evaluated. All patients were planned for conduction system pacing. Those who had more than moderate or severe systemic atrioventricular regurgitation and systemic ventricular dysfunction were planned for conduction system pacing with an additional lead in the coronary sinus (CS) tributary, i.e. bundle branch pacing optimized cardiac resynchronization therapy with the intention to achieve incremental benefit. Since right bundle pacing is not described previously and in view of anatomical complexity in location, three-dimensional (3D) anatomical mapping was done with the EnSite system and later right bundle capture is identified conventionally as that of a left bundle in a normal heart. All three patients have stable lead positions and adequate thresholds at short-term follow-up.

Conclusion In this report, we demonstrated the feasibility of permanent physiological pacing of the systemic ventricle by capturing the right bundle with 3D anatomical mapping guidance, which results in physiological activation of the systemic ventricle.

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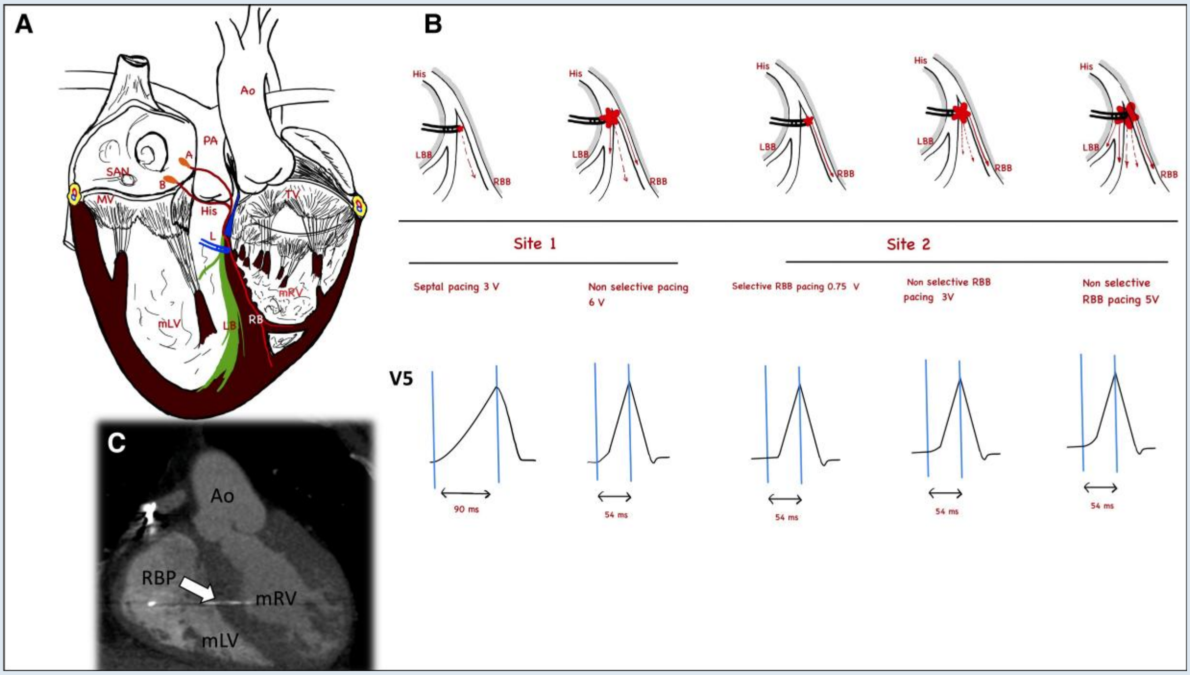
Graphical Abstract



Characteristics of the conduction system and right bundle branch pacing in ccTGA (*A*). Anatomical aspects of ccTGA; lead position at the right bundle. (*B*) Pacing at Site 1 showed mRVAT of 90 ms with high output which was reduced to 54 ms with low output. Pacing at Site 2, reached upon advancing two more turns, resulted in a constant mRVAT of 54 ms at high and low outputs indicating RB capture. The isoelectric line after the pacing spike suggested selective RB capture. (*C*) Cardiac CT of Patient 1 post-procedure showed the heavily trabeculated mRV giving raise to the aorta on the left and less trabeculated mLV on the right with RB lead (arrow). A, anteriorly located Atrio ventricular node; Ao, aorta; AVN, atrioventricular node; B, posteriorly located atrioventricular node; L, lead; LB, left bundle; SAN, sinoatrial node; MV, mitral valve; mLV, morphological left ventricle; mRV, morphological right ventricle; mRVAT, morphological right ventricular activation time; PA, pulmonary artery; RB, right bundle; TV, tricuspid valve.

Keywords ccTGA • Conduction system pacing • Complete heart block • Right bundle pacing

What's new?

- This report shows the feasibility of pacing the right bundle under 3D electroanatomical mapping guidance preserves the natural physiological activation sequence of the systemic ventricle to a greater extent and corrects the residual dysynchrony with additive benefit to the cardiac resynchronization therapy.

Introduction

Congenitally corrected transposition of great arteries (ccTGA) is a rare cardiac malformation accounting for 0.05% of congenital heart diseases and is characterized by discordant atrioventricular (AV) and ventriculoarterial connections.[1] It is associated with a 2% annual risk for spontaneous AV block.[2] Due to inherent problems associated with ventricular inversion, these patients are more susceptible to pacing-induced cardiomyopathy. Approximately 25% of isolated ccTGA patients and 67% of patients with concomitant structural heart disease develop systemic ventricular failure by the age of 45 years.[3,4] We demonstrate a novel pacing technique in ccTGA guided by three-dimensional (3D) electroanatomical mapping.

Methods

This novel technique of pacing was used in three patients with ccTGA intact ventricular septum whose baseline characteristics are summarized in *Table 1*. In two patients with moderate to severe systemic atrioventricular valvar regurgitation (AVVR), biventricular pacing with additional coronary sinus (CS) lead was implanted for bundle branch pacing optimized cardiac resynchronization therapy with the intention to achieve incremental benefit (in Patients 2 and 3).

After the baseline electrophysiological study, a 3D anatomical map was created in both ventricles by using the EnSite Precision mapping system (Abbott cardiovascular, Plymouth, MN) using an Advisor HD Grid mapping catheter. The course of His bundle (HB), right bundle (RB), and left bundle (LB) branches was tagged. Subsequently, the pacing lead (Select secure 3830, Medtronic Inc., Minneapolis, MN) was advanced into the morphological left ventricular (mLV) septum using a delivery catheter (C135 His, Medtronic Inc., Minneapolis, MN). The entry point of the screwing lead into the septum was selected as corresponding to the proximal-most RB signals on the left side of the septum, which is related to the morphological right ventricle (mRV) (*Figure 1A*). During transseptal entry as in any standard LB pacing,

Table 1 Baseline and procedural characteristics

	Patient 1	Patient 2	Patient 3
Baseline characteristics			
Baseline QRS duration (ms)	114	108	130
Systemic AVVR (grade)	2+	3–4+	3–4+
Systemic ventricular function (EF) (%)	Good (57%)	Fair (50%)	Fair (51%)
Septal thickness at the implantation site (mm) (measured in diastole 1.5–2 cm below the left AV valve)	10.5	10.8	9.8
NT pro-BNP (pg/mL)	1902	1658	2060
Procedural characteristics			
Procedure time (min)	70	75	50
Fluoroscopy time (min)	20	18	18
Number of attempts	2	1	1
Indication for periprocedural lead repositioning	Wrong path	—	—
Septal perforation	nil	nil	nil
Pericardial effusion	nil	nil	nil
Development of RBBB	nil	nil	nil
Induction of VT	nil	nil	nil

the RB potentials were recorded via lead tip in a unipolar fashion and confirmed[2]. Non-selective capture was noted and ventricular activation time (VAT) was recorded. With gentle counterclockwise torque on the sheath, the pacing lead was advanced and screwed in, and the selective RB capture was confirmed. Later, the depth of lead in the septum was confirmed with 3D mapping, contrast injection, and transthoracic echocardiography. In the other two patients, another CS lead was implanted along with the right bundle (RB) lead to attain RB-optimized cardiac resynchronization to gain incremental benefit. Cardiac CT done in one patient post-implantation demonstrated a stable lead position.

Case description

Case 1

A 52-year-old man, a case of ccTGA with moderate systemic AVVR, presented with recurrent syncope. He had NYHA Class II functional capacity and his baseline ECG showed sinus rhythm with 2:1 AV block with a QRS duration of 114 ms and intermittent complete heart block (CHB) (*Figure 2A*). At presentation, he had good systemic ventricular function and mild systemic AVVR. An electrophysiological study showed an atrio-Hisian (AH) interval of 376 ms and intermittent AH dissociation, and a His-ventricular (HV) interval of 40 ms, suggesting supra-Hisian AV block. Non-selective capture was noted with a QRS duration of 128 ms and VAT of 63 ms. Selective RB capture was noted with a QRS duration of 108 ms with an activation time of 68 ms. He completed 14 months of follow-up with us with stable lead parameters (summarized in *Table 2*). His functional class improved to Class I and with good systemic ventricular function and mild systemic AVVR.

Case 2

A 42-year-old woman presented with a history of breathlessness on exertion and recent hospitalization for pulmonary oedema. On evaluation, she was diagnosed to have ccTGA with moderate to severe systemic AVVR, good biventricular function, and atrial fibrillation with CHB (QRS duration of 108 ms) (*Figure 2A*). In view of underlying moderate to severe systemic AVVR along with CHB, she was planned for conduction system pacing with an additional lead in the CS tributary, i.e. bundle branch pacing optimized cardiac resynchronization therapy with the intention to achieve incremental benefit. HV interval was recorded as 40 ms. CS was cannulated and lead was implanted in the posterolateral tributary. Following this, the protocol used in the previous case was followed to create 3D anatomy of the ventricles and conduction system, and lead placement through the septum. Selective and non-selective capture of the RB were demonstrated (*Figure 3B*). The anodal capture threshold was 3.5 V at 0.4 ms (*Figure 3A*). She completed 8 months of follow-up with stable lead parameters (summarized in *Table 2*). She had improvement in her functional class from NYHA Class II to Class I and also her systemic ventricular function and systemic AVVR improved.

Case 3

A 77-year-old woman had recurrent presyncope for which she was evaluated and diagnosed to have ccTGA with fair systemic ventricular function and moderate systemic AVVR and CHB (*Figure 2A*). Because of moderate systemic AVVR and fair systemic ventricular function, she was planned for bundle branch pacing optimized cardiac resynchronization therapy as in the second case. The baseline QRS duration was 130 ms. HV interval was 45 ms. CS lead was implanted into a good posterolateral vein. The protocols used in the above two cases were followed. Selective and non-selective RB capture were demonstrated. The anodal capture threshold was noted at 3.25 V@0.4 ms. The device was finally programmed at bundle branch pacing output of 3 V@0.4 ms with the RV–LV delay of 30 ms achieving a QRS duration of 100 ms. She completed 7-month follow-up with stable lead parameters (summarized in *Table 2*). Her AVVR and NYHA functional class improved to NYHA class I at the last follow-up.

Baseline, procedural, pacing, and follow-up characteristics were summarized below (*Tables 1* and *2*). None of the patients had any complications during and post-procedure. All patients had objective improvement in NT pro-BNP levels on follow-up.

Discussion

We report three cases of ccTGA in whom we have performed physiological pacing by activating the RB branch first. Conventional pacing of the venous ventricle, the mLV, in ccTGA increases the interventricular septal shift and the severity of systemic AVVR, dysynchrony, systemic ventricular dilatation, and ventricular dysfunction with corresponding deterioration in NYHA functional classification, as early as 4 years after pacing.[5]

Biventricular pacing is an alternative option, but almost 20% of ccTGA patients have major anatomical challenges of CS like ostial atresia, anomalous CS drainage, duplication, ectopic displacement, or absence of conventional ostium requiring a hybrid approach to pace. Furthermore, the venous system in ccTGA is partly inverted and mLV receives major epicardial venous drainage. The mRV is predominantly drained by epicardial thebesian veins that are smaller and shorter, making lead implantation more challenging in a few cases, though, in the majority of cases, biventricular pacing can be performed as in the conventional method.

Newer conduction system pacing modalities like HB pacing and LB pacing were tried. Although HB is readily accessible in ccTGA patients from the venous end, due to its abnormal long course along the cephalad margin of the outflow tract just below the pulmonary valve increases its tenuousness resulting in fibrosis that accelerates its degeneration.[4] Pacing of HB in patients with ccTGA has been so far described only in few case reports.[6] Though it activates ventricles physiologically, HB pacing has a risk of progression of AV block and chronic elevation of the lead threshold. In the largest series on conduction system pacing in ccTGA of 15 patients with systemic ventricular dysfunction, no significant difference in QRS duration was noted in patients with pre-existing AV block and junctional rhythm.[7]

Pacing of the anatomical LB is more accessible in ccTGA as the lead stays on the right side of the septum and deeper penetrations are not needed as in conventional LB pacing. But the delayed transseptal activation

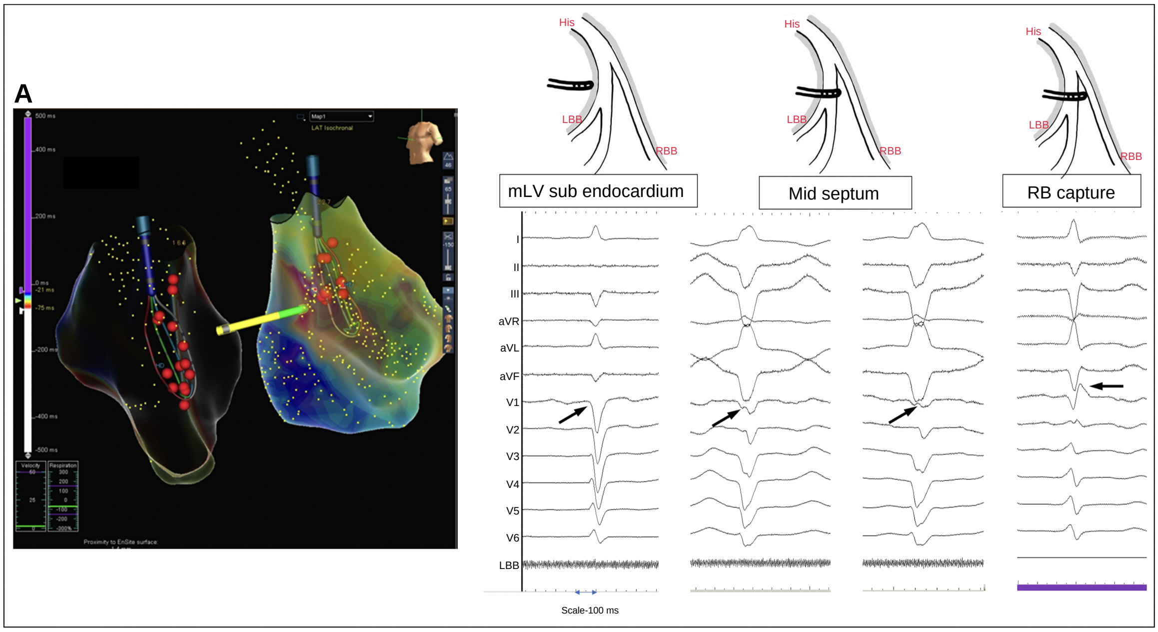


Figure 1 Procedural characteristics. (*A*) Bundle branch potentials on either side of ventricles are tagged and lead is placed transeptally into the right bundle (RB). Corresponding electrograms depicting transseptal entry. In lead V1, QRS morphology displays a QS pattern while pacing on the morphological LV (mLV) subendocardial aspect. As the lead progresses deep in the septum, an rSR' pattern gradually develops (black arrow).

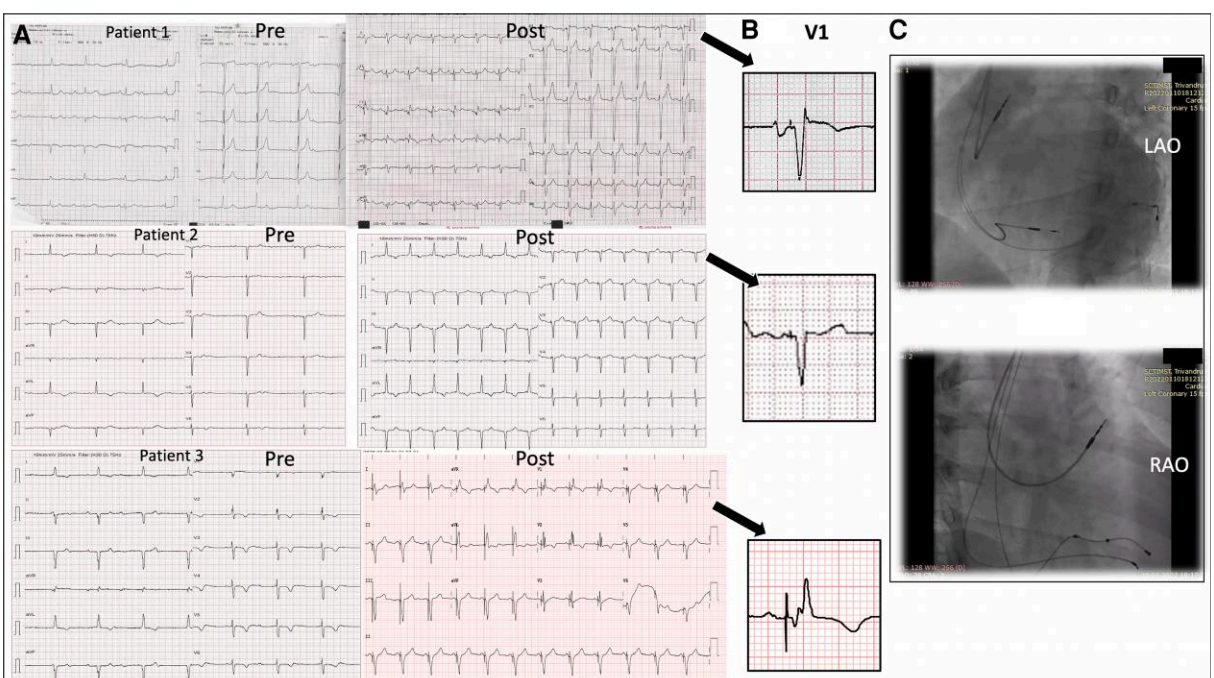


Figure 2 Post-procedure ECG; fluoroscopy and echocardiography. (*A*) ECGs of pre- and post-procedure in three patients; (*B*) zoomed view of V1 lead in three patients with QRSd of 110, 80, and 110 ms in patients 1, 2, and 3, respectively; (*C*) RAO and LAO views of final lead positions in Patient 2.

Table 2 Pacing characteristics and follow-up details

Basal intervals	Patient 1	Patient 2	Patient 3
QRS duration (ms)	114	108	130
AH interval (ms)	Intermittent AH dissociation + (376)	Patient was in atrial fibrillation AH interval could not be measured	340
HV interval (ms)	43	40	45
RB capture thresholds (@0.4 ms) (V)	0.7	1.2	0.4
Unipolar: Non-selective to selective capture transition (V)	1	1	1.25
Bipolar: Anodal capture threshold (@0.4 ms) (V)	3	3.5	3.25
R wave amplitude (mV)	15	7	10
Impedance (Ω)	960	1046	758
Right bundle potential present/absent	+	+	+
Right bundle potential-QRS duration (ms)	10	10	11
Stimulus-mRVAT in lead V5 (ms)	63	72	38
Paced QRS duration (ms)	110	92	110
Follow-up details			
QRS duration in ECG (ms) at latest follow-up	115	90	100
Device interrogation details at latest follow-up			
Pacing threshold (V)	0.5	0.5	1
Impedance (Ω)	532	633	559

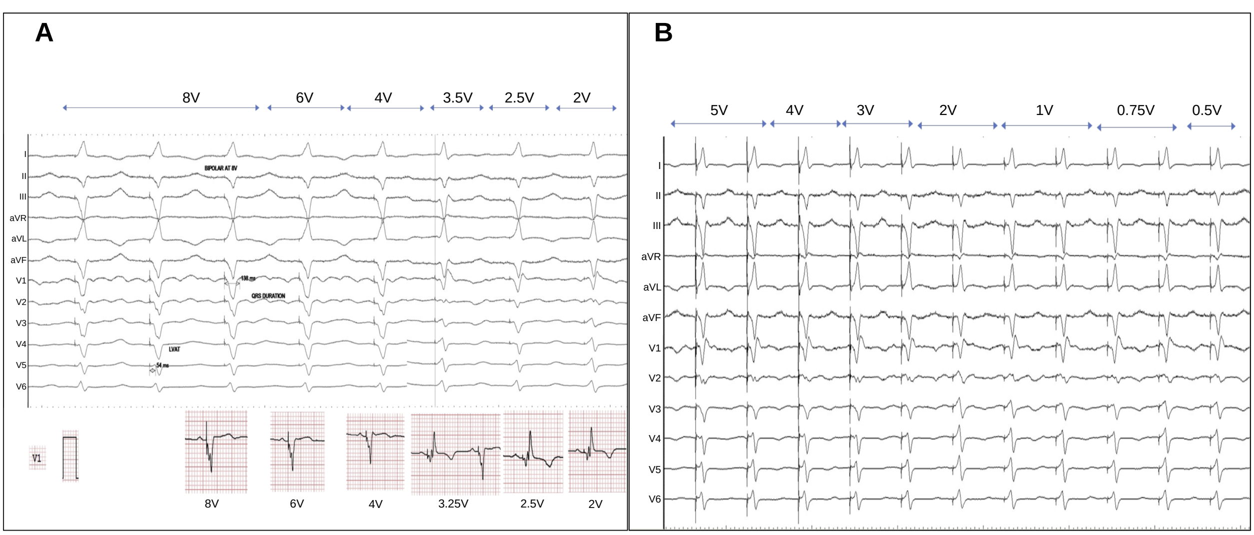


Figure 3 Unipolar and bipolar capture characteristics. (*A*) Anodal capture threshold of Patient 2. Once the anode loses capture with decreasing output, QRS morphology changes from QS pattern to QR pattern in lead V1. (*B*) Unipolar threshold testing in Patient 2. The transition from non-selective capture with initial slurring of the QRS complex to selective capture of the RB is identified by QRS preceded by an isoelectric line seen in V1 at 0.5 V.

and the subsequent ventricular dysynchrony that can result in the worsening of systemic AVVR and mRV dysfunction are still major concerns.

Ventricular inversion in ccTGA makes the distribution of the infra-atrial conduction system unique in these patients. The usual atrial arrangement, situs solitus, has the grossly abnormal disposition of the AV conduction system secondary to gross malalignment of the interatrial septum (IAS) and interventricular septum (IVS) which is filled by membranous interventricular septum or VSD. Due to these factors, the routinely placed posterior AV node in Koch's triangle is hypoplastic and may not establish contact with the distal conduction system. Instead, another anteriorly placed AV node is located near the os of the right atrial appendage which gives the penetrating bundle of His that courses anteriorly over the right ventricular outflow tract adjoining to pulmonary leaflets. As it reaches the interventricular septum, it turns inferiorly and divides into an LB supplying the mLV on the right and the RB supplying the mRV on the left[4] (see central illustration).

The unique features mentioned above suggest that the implications of RB-based conduction system pacing may not be similar to LB-based pacing in structurally normal hearts. In general, RB is a slender structure compared to LB, and capturing it with a transseptal lead may not be easy in all cases. Furthermore, limited ramifications of RB may not provide a substrate as effective as LB-based pacing to prevent ventricular dysynchrony in every case. However, the narrowing of QRS and reduction in ventricular dysynchrony noted in these cases were marked, and this pacing mode may serve as an ideal pacing strategy in ccTGA with bradycardia in the future.

Difficulties faced

(1) Identification of the thin right bundle on the septal aspect of the systemic ventricle—the target location of the pacing lead was cumbersome.
(2) The delineation of the conduction system and the subsequent lead placement had to be guided by 3D mapping.
(3) Defining the capture and characteristics of RB capture was not documented prior.

Limitations

- At present, left bundle optimized cardiac resynchronization therapy was demonstrated in a few studies for achieving incremental benefit in CRT non-responders and dilated cardiomyopathy. Knowledge of conduction system pacing in repaired or unrepaired congenital heart diseases like ccTGA is scarce.
- Long-term follow-up of these patients is needed.

Conclusion

The pacing of the sub-arterial or systemic ventricle by RB capture is true physiological pacing in ccTGA. 3D electroanatomical mapping and imaging guidance can help target the RB. For the very first time in literature, we have demonstrated the feasibility of permanent physiological pacing by RB of the systemic ventricle in ccTGA.

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Conflict of interest: No conflict of the interest among all authors.

Data availability
The authors confirm that the data supporting the findings of this study are available within the article.

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