



REVIEW

Physiological Pacing in Congenitally Corrected Transposition of the Great Arteries with Atrioventricular Block

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ABSTRACT: Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital heart disease characterized by atrioventricular, ventriculoarterial, and conduction system discordance, commonly accompanied by atrioventricular block (AVB). Pacing in patients with CCTGA and AVB (both pediatric and adult) poses challenges in strategy selection, procedural complexity, and clinical decision-making due to limited evidence. Conventional morphological left ventricular pacing is widely adopted but may induce ventricular dyssynchrony, heart failure, and tricuspid valve dysfunction. While cardiac resynchronization therapy serves as an upgrade for pacing-induced cardiomyopathy and heart failure, its application may be limited by coronary sinus anatomical variations and uncertain clinical outcomes. His bundle pacing is rarely reported due to the variation of the His bundle and high pacing threshold. The superficial, wide, multi-branched left bundle branch favors left bundle branch pacing, though delayed systemic right ventricle (sRV) activation may cause ventricular dyssynchrony and impair sRV function. Right bundle branch pacing offers a novel alternative for pacing therapy. Conduction system pacing-optimized cardiac resynchronization therapy is preferred in those with evidence of intrinsic ventricular conduction dysfunction. This narrative review synthesizes current evidence on pacing strategies for CCTGA with AVB, integrating anatomical and pathophysiological insights to evaluate physiological pacing strategies, while highlighting critical knowledge gaps to guide future research.

KEYWORDS: Congenitally corrected transposition of the great arteries; arrhythmia; atrioventricular block; pacemaker; cardiac resynchronization therapy; cardiac physiologic pacing

1 Introduction

Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital heart disease characterized by atrioventricular and ventriculoarterial discordance, with the morphological left ventricle (mLV) connected to the pulmonary trunk and the morphological right ventricle (systemic right ventricle, sRV) connected to the aorta. CCTGA accounts for less than 1% of all congenital heart defects and approximately 0.03‰ of all live births [1]. Commonly associated anomalies include ventricular septal defects (VSD), pulmonary stenosis, tricuspid valve (TV) abnormalities, and abnormal cardiac position (dextrocardia/mesocardia) [2]. 7% to 40% of CCTGA cases present with rhythm abnormalities, particularly atrioventricular block (AVB) [1,3,4] due to a tenuous atrioventricular bundle.

For young patients who undergo postnatal interventions, surgical heart block raises concerns about long-term outcomes [5,6]. Among adults, 57% reportedly develop complete AVB by the average age of



40, with an annual progression risk of 2% [7,8]. Both scenarios consequently heighten the likelihood of requiring a permanent pacemaker.

Conventional mLV pacing, involving lead placement at the ventricular apex or septum myocardium, is a proven, reliable, and safe technique. However, it may result in sRV dysfunction and progressive TV regurgitation [9–11]. Cardiac resynchronization therapy (CRT) is beneficial in mitigating or preventing heart failure from mLV pacing [10], while the anatomical variation of the coronary sinus (CS) brings difficulties in the implantation process [12]. Recently, conduction system pacing (CSP) has been demonstrated to promote physiologic activation of the sRV through direct stimulation of the His bundle, left bundle branch (LBB) [13] or the right bundle branch (RBB) [14]. His bundle pacing (HBP) shows promise as an alternative to CRT, but the unique positioning of the atrioventricular node (AVN) and His bundle in CCTGA introduces uncertainty in lead positioning [15]. Left bundle branch pacing (LBBP) effectively delivers physiological pacing and maintains electrical synchrony, demonstrating high implant success rates and low procedural complication rates [16,17]. Notably, the fan-like LBB just beneath the endocardium of the mLV septum may offer a broad target for LBBP in CCTGA [15,18]. Still, long-term and high-burden pacing in the mLV may deteriorate the sRV function. In contrast, pacing in the cord-like RBB can replicate LBBP activation patterns, potentially improving sRV function [19]. Conduction system pacing-optimized cardiac resynchronization therapy (CSP-optimized CRT) combines CSP and biventricular resynchronization to address distal conduction abnormalities in CCTGA, particularly for patients with sRV delay or wide QRS duration [20,21].

This narrative review evaluates current pacing strategies for CCTGA with AVB, with particular focus on the anatomical and pathophysiological considerations of physiological pacing approaches, while identifying key knowledge gaps requiring further investigation.

2 Anatomy and Pathophysiology

2.1 Atria and Ventricles

Most CCTGA cases occur in situs solitus, though pediatric series report situs anomalies in 30–40% of patients [22,23]. In situs solitus CCTGA, the right atrium drains into the mLV through the mitral valve (MV), and the mLV supplies the pulmonary trunk. The left atrium empties into the sRV through TV, with the sRV connected to the aorta. Unlike the normal spiral great artery configuration, the aorta typically lies left-anterior to the pulmonary trunk. Thus, systemic venous return still reaches the lungs (pumped by a subpulmonary mLV), and oxygenated pulmonary venous blood is directed to the body (pumped by sRV). In situs inversus, the mLV is left-sided, and the aorta is usually right-positioned [4,24].

2.2 Coronary Vasculature

A mirror image distribution of coronary vasculature is observed in CCTGA. In situs solitus, the morphological left coronary artery originates from the anterior aortic sinus, with its short main stem dividing into the anterior interventricular and circumflex branches. The circumflex artery encircles the MV orifice. The morphological right coronary artery arises from the posterior aortic sinus and courses through the left atrioventricular groove to become the exclusive blood supply for the sRV [4]. This distinctive coronary anatomy predisposes to sRV hypoperfusion, potentially leading to myocardial ischemia and subsequent ventricular arrhythmias or complete heart block [25,26].

Coronary venous anatomy also demonstrates significant variability in CCTGA, thereby presenting challenges during the process of venous cannulation. While 88% of cases in a postmortem study of 51 specimens exhibited normal CS anatomy [27], common variations including separate or dual ostia and persistence of the vein of Marshall frequently complicate venous cannulation during CRT implantation [12,28].

2.3 Conduction System

The malalignment of the atrioventricular (AV) septum alters the conduction system course, resulting in an elongated and aberrant pathway that predisposes to AVB [18,29].

In situs solitus, the anterior node is situated anteriorly within the right atrial septum, giving rise to the penetrating AV bundle. The posterior node is located at the typical AVN position, with the posterior bundle making almost no contact with the ventricular musculature (Fig. 1). Anatomical abnormalities are strongly associated with AVB development. Hudson proposed that the discontinuity between the posterior node and bundle branches could lead to complete heart block [30]. Supporting this, Anderson et al. identified fibrous tissue disruption of the anterior bundle in histopathological analyses of 11 CCTGA cases, further implicating conduction system abnormalities in AVB pathogenesis [18]. These degenerative changes, particularly in older patients, significantly increase AVB susceptibility.

It is particularly noteworthy that in situs inversus, the cardiac anatomy demonstrates more optimal alignment of the atrial and ventricular septa. The His bundle originates from a normally positioned AVN and connects to the conduction bundle in a manner resembling AV-concordant hearts (Fig. 1). This anatomical preservation may explain the lower incidence of AVB in situs inversus—only 1 of 18 adult CCTGA patients developed AVB in a clinical series [23].

The elongated non-branching bundle courses along the subpulmonary outflow tract before dividing into: (1) a cord-like RBB extending to the sRV; (2) a broad, fan-like LBB along the mLV septal endocardium—providing an optimal target for LBBP.

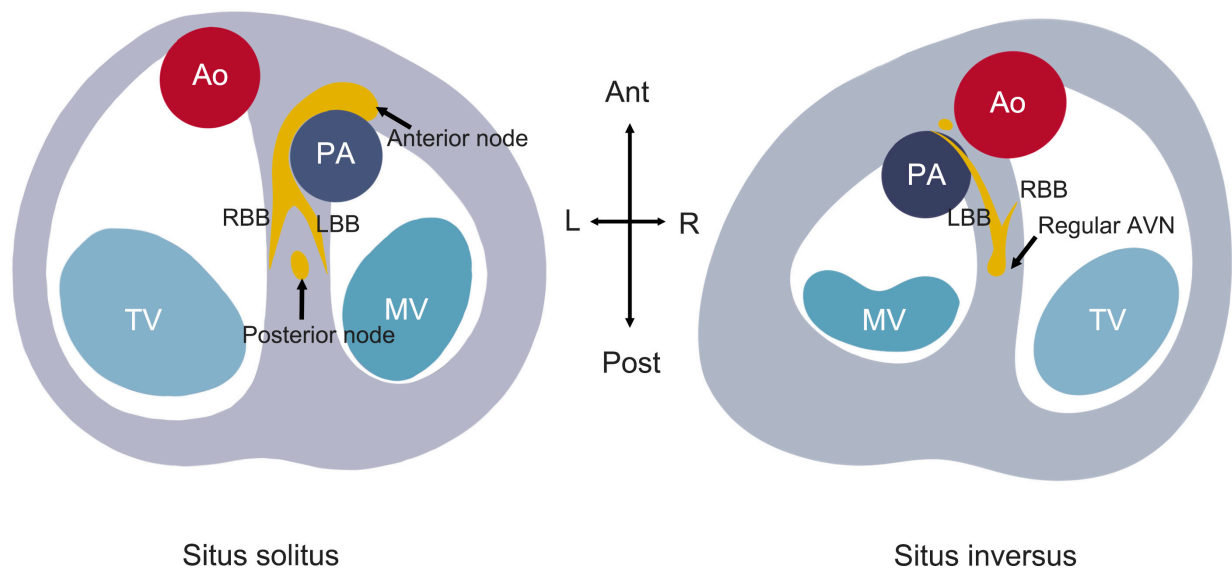


Figure 1: Schematic diagram of the conduction system in CCTGA, illustrating the anatomical differences between situs solitus and situs inversus configurations.

3 Surgically Induced Atrioventricular Block

Approximately 64.7% of CCTGA patients require surgical intervention for associated defects during childhood [6], which further elevates AVB risk through iatrogenic injury to the already vulnerable conduction system.

Tricuspid regurgitation (TR) frequently complicates CCTGA, affecting 50% of situs solitus patients but rarely occurring in situs inversus [23]. TV replacement or repair is the most commonly performed surgery in CCTGA with moderate or severe TR [31]. While TV surgery improves sRV function, it carries a 41%

risk of pacemaker-requiring AVB within 10 years, with associated QRS prolongation and worsening heart failure [32]. Novel transcatheter techniques (transcatheter TV replacement, transcatheter edge-to-edge repair) offer alternatives for high-risk CCTGA patients, showing superior outcomes to medical therapy alone [9,11,33]. However, long-term outcomes remain limited regarding: (1) cardiac implantable electronic device-related TR management; (2) lead jailing safety/prognosis; (3) post-procedural mitral regurgitation risk under chronic pacing. Transseptal access further complicates the intervention process in CCTGA.

Postoperative AVB occurs in 1–3% of isolated VSD closure for non-CCTGA patients [34–37], with a higher risk in infants <4 kg [38]. Anatomic proximity of the AVN to the VSD's posterior-inferior margin predisposes to conduction system injury during VSD repair, increasing pacemaker dependence [39]. In children <12 years, epicardial pacing shows higher lead failure rates [40], while long-term ventricular pacing reduces battery longevity and may impair ventricular function [41]. Therefore, the aberrant conduction anatomy in CCTGA itself warrants particular clinical attention to the risk of postoperative AVB during VSD closure.

4 Pacing Strategies in Congenitally Corrected Transposition of the Great Arteries

4.1 Conventional Morphological Left Ventricular Pacing

Conventional mLV pacing remains widely used in CCTGA, with the ventricular lead typically positioned at the mLV apex [5]. Evidence has demonstrated that conventional mLV pacing induces ventricular dyssynchrony and pacemaker-induced cardiomyopathy, which progressively impairs sRV function, leading to ventricular dilatation and deterioration [42–45]. These adverse hemodynamic consequences necessitate alternative pacing strategies in this sRV failure-prone population.

4.2 Cardiac Resynchronization Therapy

CRT serves as a therapeutic option for CCTGA patients with AVB, particularly for pacing-induced cardiomyopathy and heart failure. CRT indications for CCTGA generally follow standard criteria but require additional anatomical and functional considerations. For patients in sinus rhythm, CRT carries a Class I recommendation when QRS duration is ≥ 150 ms with left bundle branch block (LBBB) pattern and left ventricular ejection fraction (LVEF) $\leq 35\%$. For pacemaker-dependent patients with LVEF $\leq 35\%$ and high right ventricular pacing burden ($>40\%$), CRT upgrade may be considered [46–48].

While CRT improves ventricular function [49] and transplant-free survival [50] in congenital heart disease with systolic dysfunction and electrical dyssynchrony [51], its application in CCTGA remains limited due to anatomical complexities. Key uncertainties persist regarding CS lead implantation techniques, optimal ventricular lead positioning [52], and long-term outcomes. CS angiography or cardiac computed tomography (CT) helps identify venous variants, facilitating transvenous lead placement in most cases [12]. The optimal ventricular lead configuration and determinants of long-term CRT efficacy in CCTGA remain poorly characterized [53–55], compounded by frequent concomitant hemodynamic and arrhythmic comorbidities. Large-scale studies are required to establish evidence-based implantation strategies.

4.3 Conduction System Pacing

The 2023 CSP guideline recommends HBP/LBBP for CCTGA with AVB (Class IIb, Level of evidence C) [10]. A schematic diagram summarizing the currently reported physiological pacing strategies of HSP, LBBP, and RBBP is provided in Fig. 2. However, key aspects remain undefined, including implantation techniques, His bundle/LBB/RBB anatomy variations, optimal lead positioning, and long-term outcomes.

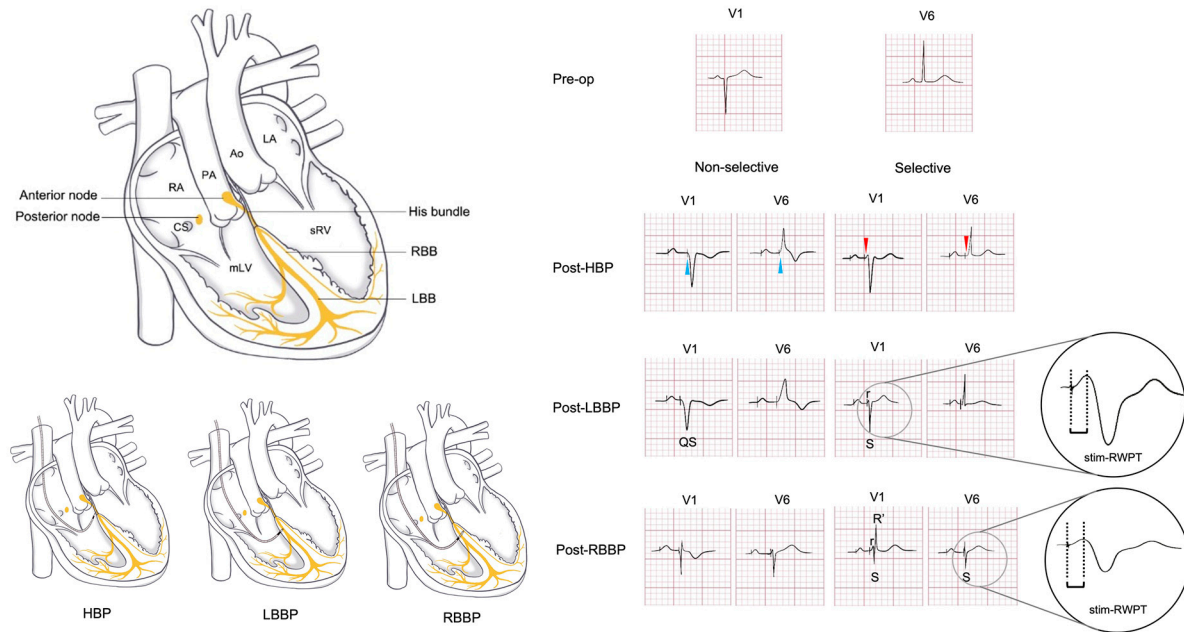


Figure 2: Preoperative and postoperative ECG patterns in CCTGA with situs solitus. The preoperative ECG of CCTGA patients without pulmonary stenosis or VSD typically shows a QS pattern in the V1 lead. This schematic illustrates the cardiac anatomy and corresponding ECG changes following CSP, delineating the distinctions between selective and non-selective capture. The depicted waveforms are derived from established literature and the authors' clinical observations.

4.3.1 His Bundle Pacing

His-bundle localization is paramount for successful His-bundle pacing (HBP). In most non-congenital patients, His bundle can be mapped using the pacing lead by positioning the electrode tip near the tricuspid annulus under intracardiac electrogram guidance. Alternative approaches include: (1) fluoroscopic-guided positioning in the right anterior oblique (RAO) 30° projection followed by confirmation via high-output, high-frequency unipolar pacing with analysis of QRS morphology; (2) advanced imaging techniques such as three-dimensional electroanatomic mapping (3D-EAM), intracardiac echocardiography (ICE), transesophageal echocardiography (TEE), transthoracic echocardiography (TTE), and tricuspid annular angiography [10,56].

However, these standard techniques face unique challenges in CCTGA due to the abnormal conduction system anatomy. The elongated His bundle in CCTGA courses anterior to the pulmonary annulus, frequently posing capture challenges. Limited successful cases have been documented: Takemoto et al. achieved HBP in an 8-year-old, demonstrating QRS narrowing (198 ms to 94 ms) with clinical improvement [57]. Vijayaraman et al. utilized 3D-EAM to localize the His bundle and LBB, achieving selective capture (0.5 V/1 ms) from initial nonselective HBP (1.5 V, 132 ms QRS) with 8.5 mV R-wave amplitude and 609 Ω impedance. As schematically illustrated in Fig. 2, selective and non-selective HBP exhibit distinct electrocardiographic features. Selective capture (red arrows) is identified by a consistent paced/native QRS morphology and a clear isoelectric line between the stimulus and QRS. Non-selective capture (blue arrows) demonstrates fusion at the stimulus site, resulting in the loss of the isoelectric line, a wider paced QRS, and T-wave discordance. Echocardiography confirmed lead tip position anterior to the pulmonic valve [58]. While HBP shows therapeutic potential for AVB, concerns remain regarding chronic threshold elevation and battery longevity. Large-scale trials are warranted to establish long-term efficacy.

4.3.2 Left Bundle Branch Pacing

In CCTGA patients, LBBP is typically performed via axillary or subclavian venous access. The procedure employs the widely adopted selected secure pacing lead (model 3830; Medtronic Inc., Minneapolis, MN, USA) delivered through a fixed curve sheath (C315 HIS; Medtronic Inc.) under fluoroscopic guidance [13].

Crucially, the precise LBB location is critical for successful lead placement. As demonstrated in prior cadaveric studies, the LBB in CCTGA patients is characteristically superficial within the subendocardium of the mLV, exhibiting a broad, interconnected network that merges with the Purkinje system [15]. Intraoperative localization of the LBB can be achieved through several approaches. 3D-EAM provides optimal guidance, with initial experience in 7 pediatric CCTGA cases confirming the LBB's accessible position and favorable morphology for pacing [19,59]. In the absence of 3D-EAM, fluoroscopic guidance (RAO 30° with 9-partition method) offers a practical alternative, though it demands substantial operator expertise in LBBP [60]. Also, TTE further aids lead positioning, with the target zone identified 2–3 cm from the tricuspid septal leaflet within the interventricular septum in apical four-chamber or subxiphoid views [61].

Currently, there are no established clinical diagnostic criteria for LBBP in CCTGA. Based on our institutional experience and systematic evaluation, we propose the following diagnostic criteria: (1) pacing lead positioned in the LBB region with a recordable LBB potential; (2) pacing ECG demonstrates RBBB morphology in V1; (3) short and consistent stimulus to R-wave peak (stim-RWPT) in V1 (65–80 ms); (4) sudden shortening of the stim-RWPT in V1 (≥ 10 ms) with increasing output; (5) for selective LBB capture: V1 shows rS/RS pattern with QRS duration < 120 ms; (6) for non-selective LBB capture: non-selective LBB capture: V1 waveform less typical than above, with relatively narrow QRS duration (Fig. 2). We maintain that myocardial capture exhibits characteristics analogous to septal pacing, defined by: (1) paced QRS duration ≥ 120 ms, lacking typical bundle branch capture patterns; (2) V1 demonstrating a wide QS morphology; (3) stim-RWPT in V1 exceeding 85 ms [10,56].

Following the successful lead positioning, confirmation of adequate helical screw depth and stable fixation is essential. Fluoroscopic assessment of the tip-fulcrum distance helps evaluate septal penetration depth, while left anterior oblique (LAO) 30° sheath angiography and intraprocedural TTE provide additional confirmation. For selected secure pacing lead (model 3830; Medtronic Inc.), rebound testing should be avoided due to perforation risk. Instead, maintain gentle tension during sheath withdrawal, verify electrical parameters for stability, and ensure appropriate lead slack [56,61].

While LBBP demonstrates proven long-term benefits in non-congenital populations, its application in CCTGA raises unique concerns: pacing from the mLV subendocardium may delay transseptal activation, exacerbating ventricular dyssynchrony and potentially worsening sRV dysfunction. Current evidence remains limited, with intermediate-term (≤ 1 year) data showing preserved QRS duration and stable thresholds [13], though some report QRS widening post-LBBP in previously non-paced patients [19]. While preliminary findings suggest maintained sRV function [19,45], small cohort sizes and absent long-term follow-up necessitate cautious interpretation of these outcomes.

4.3.3 Right Bundle Branch Pacing

Right Bundle Branch Pacing (RBBP) represents an emerging physiological pacing strategy for CCTGA. In situs solitus, the RBB's anatomical position mirrors the LBB in normal hearts, allowing analogous implantation techniques. A case report confirmed its feasibility using 3D-EAM, demonstrating QRS narrowing, stable parameters, and clinical improvement. Notably, selective RBBP is characterized by V1 showing “M” or rsR’ morphology with notched R’ waves and deep notched S waves in lead I/V5/V6, with short stim-RWPT in V5/V6 (38–72 ms) (Fig. 2) [14].

Challenges include the RBB's cord-like morphology, which complicates transseptal capture, and its limited branching may reduce resynchronization efficacy versus LBBP. While 3D-EAM may address anatomical localization, further technical refinement is needed to establish RBBP's safety profile and standardized protocols. Nevertheless, it expands physiological pacing options for CCTGA with AVB.

4.4 Conduction System Pacing-Optimized Cardiac Resynchronization Therapy

Despite the successful implantation of CRT, failure can be found in improving ventricular function. Also, CSP may be ineffective in 10% of adult CCTGA patients with intrinsic ventricular conduction dysfunction [12]. Thus, CSP-optimized CRT has been described as a strategy to overcome distal Purkinje or myocardial disease by directly recruiting late-activated myocardial segments [20]. Case reports demonstrated that CSP-optimized CRT could benefit bi-ventricular resynchronization in CCTGA, resulting in an improved electrocardiographic and clinical response. An algorithmic approach to CRT for adult CCTGA is employed. For intact AV conduction with evidence of sRV electrical delay, CSP-optimized CRT is pursued. With narrow QRS, CSP targeting the distal His bundle or proximal LBB alone is expected to maintain electrical synchrony. With wide QRS, CSP-optimized CRT is preferred [21].

5 Discussion

CCTGA is a rare congenital heart defect characterized by the abnormal positioning of the heart's main chambers. AVB, often being intrinsic or caused by surgical impairment, is a common associated rhythm abnormality. Thus, permanent pacing therapy is often required. However, due to limited pacing experiences in such rare cases, the lack of clinical evidence greatly restricts the treatment of CCTGA with AVB. We hope this review on pacing therapy in patients of CCTGA with AVB can increase further clinical research and highlight the emerging role of physiological pacing.

In pediatric populations, surgical intervention can fundamentally address the problem, reducing the implantation rate of pacemaker devices in minors and directly altering the prognosis. However, surgical procedures often lead to serious AVB due to damage to the conduction system. While transient postoperative conduction disturbances may resolve spontaneously, permanent pacemaker implantation introduces long-term device-related complications, including lead dislodgement, lead fracture, and pacing-induced cardiomyopathy—particularly concerning given the anticipated decades of device dependency. We advocate for refined surgical techniques that minimize conduction tissue trauma, thereby reducing permanent pacing requirements in this vulnerable population.

For adult patients, congenital anatomical abnormalities of the conduction system are often caused by the formation of adipose tissue and fibrous tissue with age. While conventional mLV pacing remains widely employed, long-term mLV pacing carries the risk of pacing-induced cardiomyopathy and subsequent heart failure. CRT serves as the primary upgrade strategy for such cases, though its efficacy may be limited by CCTGA's unique CS anatomy. Recent clinical experience with CSP in CCTGA reveals important anatomical and technical considerations. There are few cases for HBP, probably due to the variation of His bundle (especially when combined with VSD) and high pacing threshold. In contrast, the superficial, wide, and multi-branched LBB in the mLV may provide a proper anatomical basis for pacing assisted by 3D-EAM. However, chronic LBB capture may still induce intraventricular dyssynchrony, resulting in delayed activation of the sRV and subsequent sRV functional deterioration. RBBP in the deeper surface of the ventricular septum (near the sub-endocardium of the sRV) provides new ideas for pacing strategy for such patients. Importantly, these techniques show limited efficacy in patients with extensive distal conduction disease, where a hybrid approach combining CSP with CRT may better address both proximal and distal conduction abnormalities.

Our analysis suggests that while physiological pacing approaches show theoretical promise, future investigations should focus on: (1) technical refinements for CS lead placement in the context of coronary venous anomalies, as well as the optimal ventricular lead position in CRT; (2) improved electrophysiological mapping modalities for precise LBB and RBB localization; (3) development of evidence-based guidelines for physiological pacing in CCTGA. These advancements will require multicenter collaboration to overcome the challenges posed by the rarity and anatomical complexity of this condition.

6 Conclusion

This review highlights the anatomical and technical considerations for pacing in CCTGA with AVB, emphasizing the potential of physiological pacing. Current evidence remains limited by small cohorts and technical challenges, particularly in lead placement and long-term stability. Future studies should prioritize refining lead positioning techniques and multicenter collaboration to establish standardized approaches.

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Abbreviations

All abbreviations are defined at first use in the text and summarized alphabetically below.

3D-EAM	Three-Dimensional Electroanatomical Mapping
Ao	Aorta
AV	Atrioventricular
AVB	Atrioventricular Block
AVN	Atrioventricular Node
CCTGA	Congenitally Corrected Transposition of the Great Arteries
CRT	Cardiac Resynchronization Therapy
CS	Coronary Sinus
CSP	Conduction System Pacing
CSP-optimized CRT	Conduction System Pacing-Optimized Cardiac Resynchronization Therapy
HBP	His Bundle Pacing
ICE	Intracardiac Echocardiography
LA	Left Atrium
LBB	Left Bundle Branch
LBBB	Left Bundle Branch Block
LBBP	Left Bundle Branch Pacing
LV	Left Ventricle
LVEF	Left Ventricular Ejection Fraction
mLV	Morphological Left Ventricle
MV	Mitral Valve
PA	Pulmonary Artery

Pre-op	Pre-Operation
Post-op	Post-Operation
RA	Right Atrium
RBB	Right Bundle Branch
RBBP	Right Bundle Branch Pacing
sRV	Systemic Right Ventricle
stim-RWPT	Stimulus to R-Wave Peak
TEE	Transesophageal Echocardiography
TTE	Transthoracic Echocardiography
TV	Tricuspid Valve
TR	Tricuspid Regurgitation
VSD	Ventricular Septal Defect

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