

The man in the mirror: Biventricular device implantation in a patient with dextrocardia with situs inversus totalis



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Introduction

The population of individuals with adult congenital heart disease is expanding as advances in surgical and medical management allow patients to live into adulthood. Consequently, these patients are developing other cardiovascular diseases, such as arrhythmias and heart failure, later in life. Procedures involving congenital anatomic variations are becoming more commonplace, and operators must become familiar with them. One such congenital abnormality, dextrocardia, involves the reversal of the base–apex axis of the heart caudally and to the right. It may occur in isolation or, in approximately one-third of patients, involve other visceral organs of the body, which is referred to as situs.¹ Here we present a case of biventricular (BiV) device implantation in a patient with dextrocardia and situs inversus totalis and the techniques used to facilitate the procedure. Verbal consent was obtained from the patient before writing this case report.

Case report

A 63-year-old male with dextrocardia with situs inversus totalis, alcoholic cardiomyopathy with reduced ejection fraction, history of nonobstructive coronary artery disease, and atrial fibrillation and atrial flutter was seen at our cardiology clinic for management of heart failure. The patient had experienced multiple episodes of acute decompensated heart failure despite receiving goal-directed medical therapy for more than 2 years. The patient's management and follow-up had been interrupted by multiple heart failure admissions, and his ejection fraction remained low despite medi-

KEY TEACHING POINTS

- Anatomic variations of the cardiac chambers and visceral organs in patients with dextrocardia can occur along a spectrum and must be properly identified.
- Preprocedural planning with multimodal imaging strategy is vital for procedural success and efficiency.
- Once the preprocedural roadmap is established, intraprocedural confirmation of anatomic position using mapping catheter is an effective strategy to confirm accurate lead position.

cal therapy. The patient was referred to the cardiac electrophysiology clinic for primary prevention device implantation and cardiac synchronization. He endorsed episodes of syncope that became more frequent. Baseline electrocardiogram showed first-degree atrioventricular block with right bundle branch block and left anterior block resulting in QRS duration of 164 ms. Given the patient's heart failure status and unexplained syncope, the decision was made to implant a BiV device for cardiac resynchronization therapy.

In preparation for the procedure, coronary computed tomographic study was performed to delineate the patient's coronary venous anatomy (Figure 1). The study confirmed the diagnosis of dextrocardia with no additional variants in cardiac anatomy and was used to guide our procedural approach. At the start of the procedure, a decapolar deflectable catheter (steerable, 6F, decapolar, 270° standard, 2.5-mm spacing, 105 cm, Polaris [Boston Scientific, Marlborough, MA]) was introduced from the right femoral vein and was used to try to cannulate the coronary sinus (CS). Given the difficulty in locating the CS, the “mirror” mode

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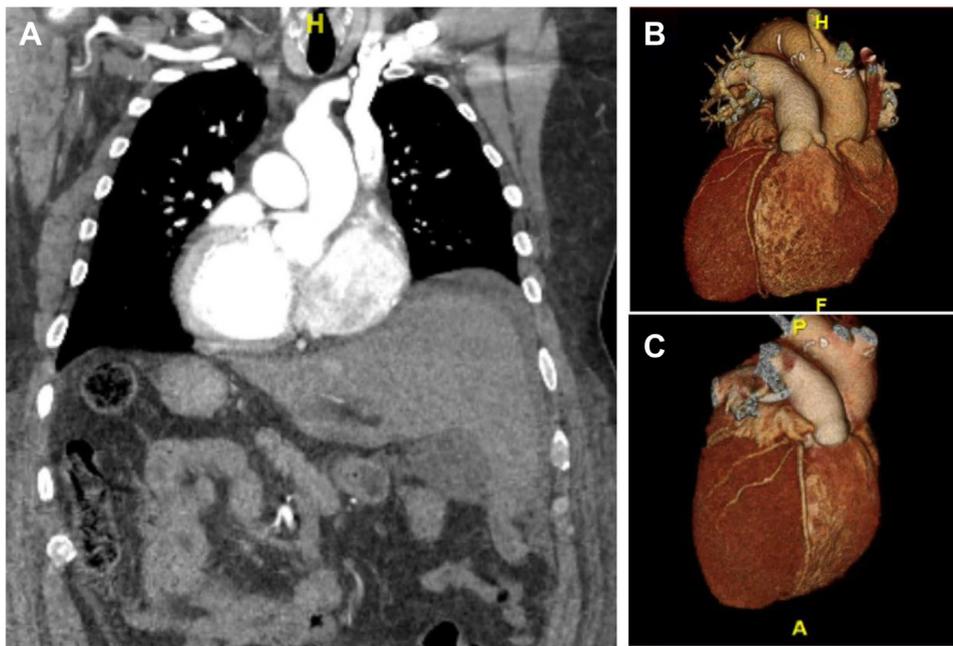


Figure 1 A: Computed tomography showing dextrocardia and situs inversus totalis. B: Three-dimensional reconstruction (cranio-caudal orientation). C: Three-dimensional reconstruction (anterior-posterior orientation).

on fluoroscopy was used, thus flipping the images obtained in the right anterior oblique (RAO) and left anterior oblique (LAO) views horizontally (ie, the image shot under RAO projection was equivalent to an image of a normal situs in LAO) (Figure 2). After locating the CS, we advanced the catheter distally into the CS main trunk and observed the typical atrial and ventricular signals. The catheter was left in place and served as a landmark for left ventricular lead placement. The right chest was chosen for device placement, and a right arm venogram confirmed axillary and subclavian vein patency and drainage into the superior vena cava. The right ventricular lead was placed successfully at the right ventricular apex. The CS ostium was cannulated with the CS sheath and deflectable Polaris mapping catheter. A quadripolar active-fixation left ventricular lead was placed in a lateral branch with excellent pacing threshold without diaphragmatic stimulation. Postprocedural chest radiography showed appropriate lead positions and no pneumothorax (Figure 3).

Discussion

With advancements in the medical and surgical management of congenital heart disease, cardiologists are encountering anatomic variants in the procedural setting more frequently. Dextrocardia creates technical challenges with regard to identification of cardiac chambers and other anatomic landmarks. Thus, manipulation of catheters and leads becomes increasingly difficult. When associated with abnormal orientation of visceral organs, 3 scenarios exist: (1) situs solitus, in which orientation is normal; (2) situs inversus, in which orientation is a mirror image of normal; and (3) situs ambiguous, in which orientation is inconsistent. Other congenital valvular or vascular abnormalities can coexist with dextrocardia. Thus, appropriate preprocedural imaging is necessary to establish the location of anatomic landmarks and select compatible equipment. Two major anatomic associations are most useful in determining the identity and position of cardiac chambers in dextrocardia. First is the identification of atria based on the entry point of the inferior and superior

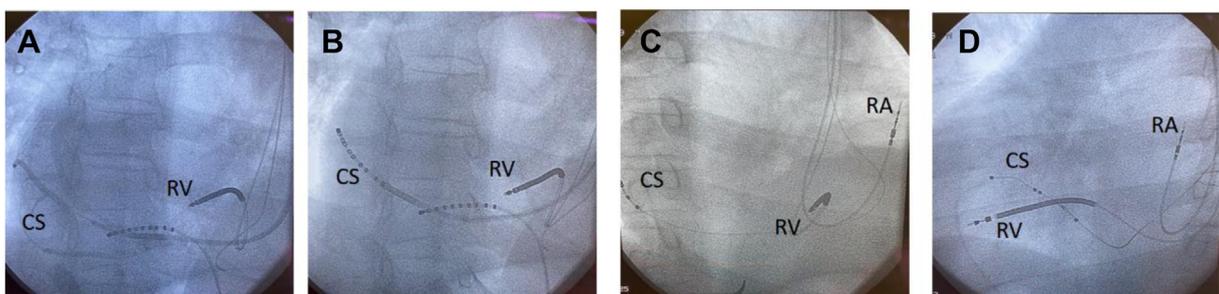


Figure 2 A: Coronary sinus (CS) venogram (right anterior oblique [RAO]) view. B: Mapping catheter in the CS (RAO view). C: RA, RV, and LV lead (RAO view). D: RA, RV, and LV lead (left anterior oblique view). RA = right atrium; RV = right ventricle.

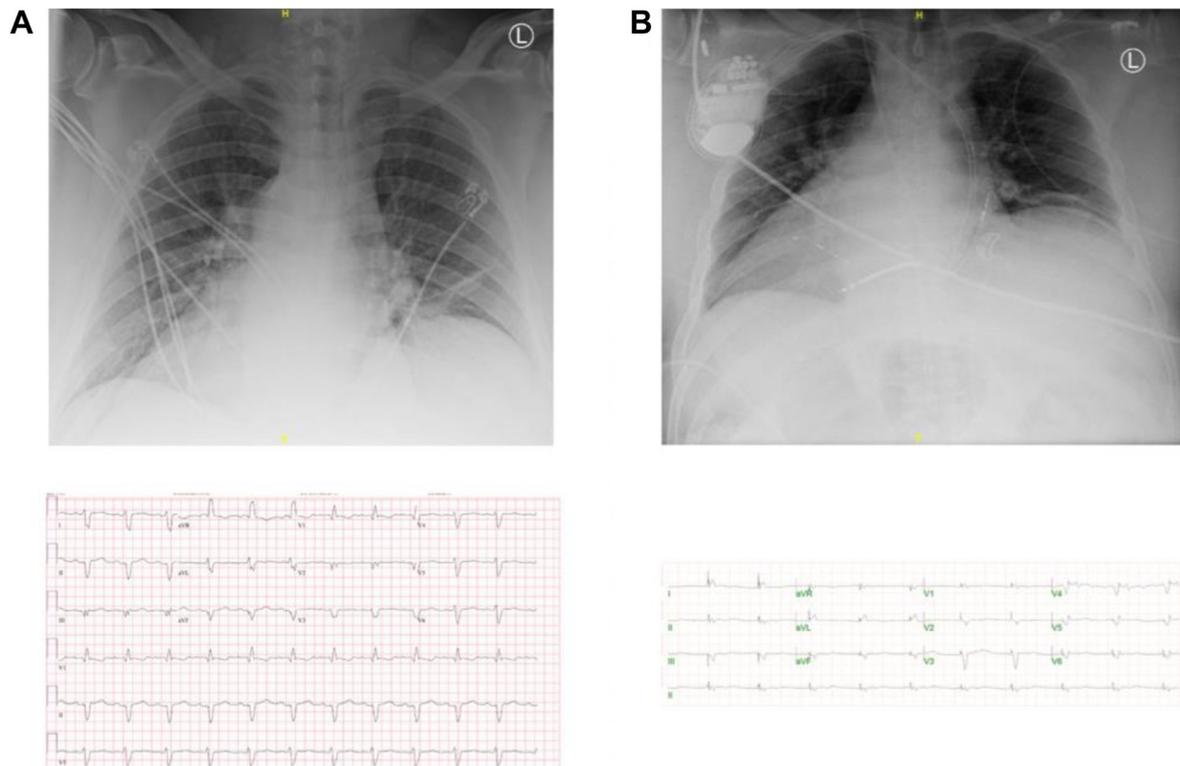


Figure 3 Chest radiography and electrocardiography before (A) and after (B) the procedure.

venae cavae rather than the position of visceral organs. Second is the identification of ventricles based on the course of the left anterior descending coronary artery, pulmonary trunk, and position of aortic and pulmonary annuli in patients without transposition.² Once these 2 key anatomic associations have been established and any other congenital abnormalities are accounted for, procedural planning can begin.² Preprocedural planning included cardiac computed tomography with contrast enhancement in the venous phase, similar to a pulmonary embolus protocol, which enabled us to view the course of the venous drainage into the right atrium and right ventricle.

During the implant procedure, several potential strategies can help identify the established landmarks. We chose to use CS venography and electrical mapping to identify anatomic landmarks, as reported by other investigators.³ Another potential strategy is to perform a digital horizontal flip of the fluoroscopic screen projection to have the heart in a normal orientation. This allows the operator to overcome the technically difficult aspects of the procedure that would otherwise require unfamiliar hand–eye coordination and catheter manipulation.⁴ Although our case utilized mapping catheter and CS venography, locating and cannulating the CS ostium can be difficult. An alternative strategy when unable to locate the CS is venous-phase coronary angiography to determine appropriate lead placement.⁵

Regardless of intraprocedural strategy, the success of device implantation in patients with dextrocardia with situs in-

versus relies on adequate preprocedural imaging to establish key anatomic associations and to account for other potential congenital abnormalities associated with dextrocardia. At 6-month follow-up, the patient had significant improvement in dyspnea and fatigue, with no further hospitalizations for decompensated heart failure.

Conclusion

Implanting a BiV device in a patient with dextrocardia is challenging. Our case presents multiple strategies that we used in preparation of the procedure and during the implant. Given that cardiologists may encounter adult patients with congenital anomalies, being aware of potential strategies is crucial for procedural success.

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