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Transvenous pacing through the pulmonary valve in a patient with cyanotic congenital heart disease after Glenn shunt – A case report

Srinivas Bhyravavajhala, Bharathi Vanaparty, Sreekanth Yerram*

Department of Cardiology, Nizam's Institute of Medical Sciences, Hyderabad, India



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ABSTRACT

Bradyarrhythmia requiring pacing is infrequently encountered in patients with complex cyanotic congenital heart disease. Even though epicardial pacing is the preferred mode, rarely, a need for endocardial lead implantation arises.

Patients with cavopulmonary shunts limit access to the venous atria and ventricles, necessitating alternate methods of pacemaker implantation. We report transvenous endocardial lead implantation by an unconventional method in a patient with congenitally corrected transposition of great arteries after a bidirectional Glenn shunt.

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1. Introduction

Congenitally corrected transposition of the great arteries (CC TGA) has a high incidence of complete heart block due to the abnormal position of the atrioventricular node (AV) and an abnormal course of the AV conduction system [1]. Patients with palliative cavopulmonary shunts provide a challenge for endocardial pacing. We report a patient with CC TGA, ventricular septal defect (VSD), severe pulmonary stenosis (PS), and atrioventricular block (AV) who had undergone bidirectional Glenn shunt (BDG). Endocardial pacing lead was inserted by a novel method through the Glenn shunt and the pulmonary valve.

2. Case

A 16-year-old girl with CC TGA, large inlet VSD, severe PS, and AV block not suitable for complete repair had undergone bidirectional Glenn shunt two years back. Permanent pacemaker implantation (Sensia, Medtronic Inc., Minneapolis, MN, USA) with a single epicardial lead was done during the same procedure. The

implantation threshold was 0.6 mV, which gradually increased to more than 5 mV over two years and four months with battery depletion requiring elective pulse generator replacement.

On examination, she was cyanotic with a prominent ejection systolic murmur in the second right intercostal space. Her electrocardiogram showed complete heart block with a narrow QRS ventricular escape rate of 65 bpm. (Fig. 1). Echocardiogram showed situs solitus, levocardia, atrioventricular and ventriculoarterial discordance with L-posed aorta (Fig. 2a, Supplementary Video 1). There was a large inlet ventricular septal defect with a bidirectional shunt and severe valvular and subvalvular pulmonary stenosis (Fig. 2b, video 2). A catheterization study demonstrated the patency of the Glenn shunt (video 3, 4, 5) and the right ventricular outflow tract (RVOT). The patient was not willing for a repeat surgical procedure, including Fontan completion. As there was a need for pacing, endocardial lead implantation was planned.

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Under local anesthesia, the extrathoracic left axillary vein was

* Corresponding author. Department of Cardiology Nizam's Institute of Medical Sciences Hyderabad, 500082, India.

E-mail address: sreeyerram@gmail.com (S. Yerram).

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Fig. 1. Electrocardiogram showing complete AV dissociation, right axis deviation, q waves in the anterior chest leads with their absence in the lateral leads suggestive of altered septal depolarization [AV- atrioventricular].

accessed, and a 6F sheath was inserted. The pulmonary valve was crossed with a glidewire (Terumo, New Jersey, USA) and a Judkins’ right catheter into the right-sided ventricle. A coronary sinus sheath (CPS Direct PL, St Jude Medical, USA) was positioned in the ventricle after confirming with a contrast injection (Fig. 3) on which a 58 cm screw-in MR conditional lead (Medtronic Inc., Minneapolis, MN, USA) was delivered to the desired position in the apex. The lead was screwed after achieving acceptable pacing parameters, and the coronary sheath was peeled away (Fig. 4). The pacing

threshold was 0.5 mV, R wave amplitude was 12 mV with an impedance of 720 Ω. The pulse generator (Sensia, Medtronic Inc., Minneapolis, USA) was placed in the subcutaneous pocket, and it was closed in layers (Fig. 5). The fluoroscopy time was 16 min, and the total procedure duration was 90 min. The device was programmed to a lower rate of 70 bpm in VVIR mode with an output of 3.5 mV. The post-procedure ECG showed consistent ventricular pacing with a relatively narrow QRS complex (Fig. 6). The patient was started on Vitamin K antagonist with a goal INR of 2.5, with a

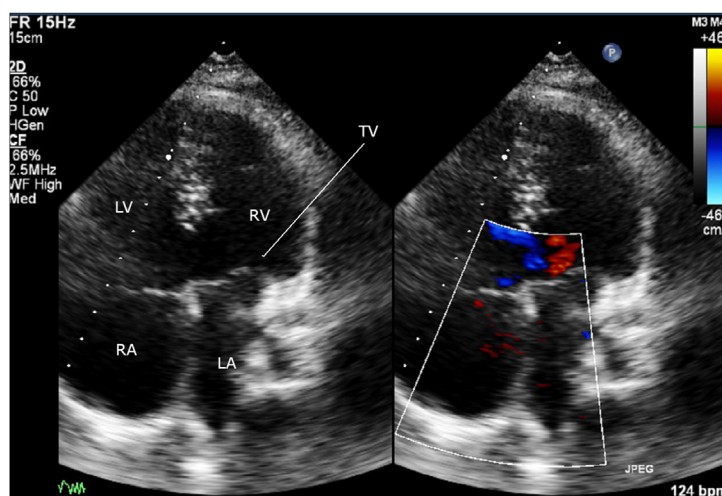


Fig. 2a. Echocardiogram in the four-chamber view demonstrates the morphological RV on the left and vice versa. The apical insertion of the tricuspid valve, a large inlet VSD, and an ostium secundum ASD can be seen. VSD-ventricular septal defect, ASD- Atrial septal defect LV- left ventricle, RV- right ventricle, TV- tricuspid valve, LA-left atrium, RA-right atrium.

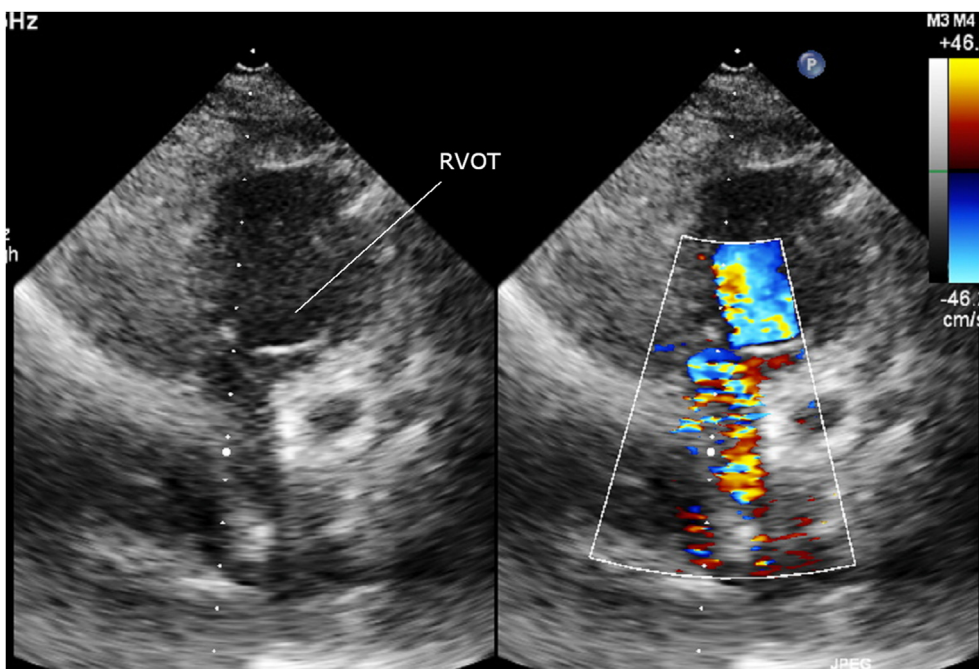


Fig. 2b. Echo with color doppler showing the thickened and stenotic pulmonary valve and turbulent jet across it. RVOT-right ventricular outflow tract.

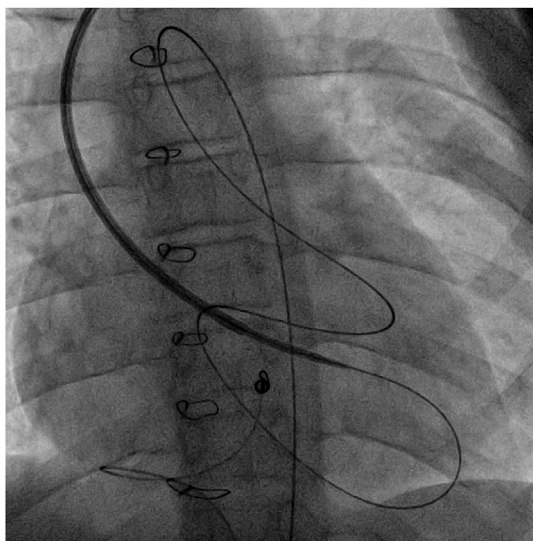


Fig. 3. The fluoroscopic image demonstrates the coronary sinus sheath with the dilator, delivered to the ventricle over the guidewire.

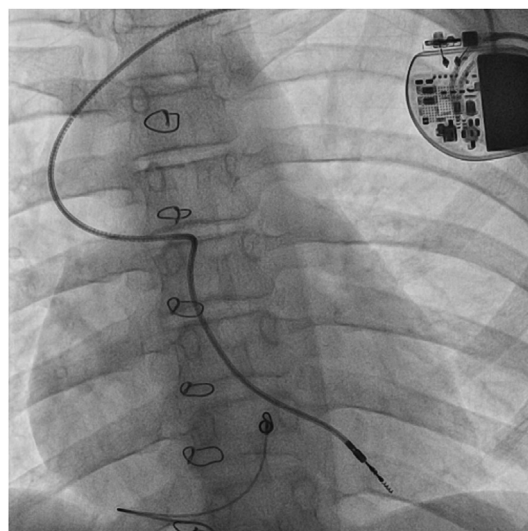


Fig. 4. Fluoroscopy in the AP view showing the screw-in lead implanted in the ventricular apex. The lead takes a tortuous course.

plan to continue indefinitely. She had no adverse events until the six-month follow-up visit.

3. Discussion

In the presence of cavopulmonary shunts, epicardial leads are the ideal choice for pacing, but they are prone to early failure [2].

Endocardial pacing needs unconventional ways to reach the venous atria and ventricles, like transhepatic, hybrid per atrial route, and through the shunts [3]. The femoral vein access could be another option for implantation but is known to have high lead displacement rates, and the transhepatic route has a higher risk of complications. There was antegrade flow across the RVOT (pulsatile

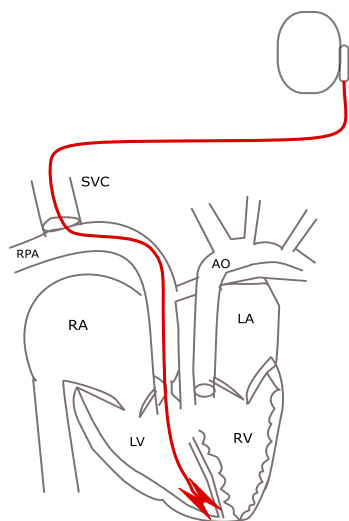


Fig. 5. Graphical representation depicting the lead course. RA-right atrium, LA-left atrium, RV- right ventricle, LV- left ventricle, RPA-right pulmonary artery, AO- aorta.

Glenn) in our patient, which provided a path for the lead from the systemic vein to reach the right-sided ventricle. Habib M et al. have reported a single such case, where they implanted a permanent pacemaker in a patient with single ventricle after Glenn shunt. The right subclavian access was used, the ventricle was reached with a glide wire and a 7 French peel-away sheath and a screw-in lead was

implanted [4].

The main difficulty encountered during the procedure was to guide the lead to the desired position in the ventricle. There was a tortuous course that the lead had to traverse, crossing the severely stenosed pulmonary valve in a retrograde manner. We used a glide wire to cross and exchanged to a coronary sinus lead delivery sheath. The lead could not be manipulated inside the ventricle and hence needed a delivery sheath, which takes it to the final desired position in the ventricle. The apical position was obtained without much effort.

The formation of thrombus on the endocardial lead with a potential for embolism is the primary concern, but the events are less if they are on adequate anticoagulation [5]. Another concern is the thrombosis and occlusion of the Glenn shunt due to the lead, which is a low flow shunt. Leadless pacemakers do not have the risk of thrombosis of the veins, but they are a bulkier device in the ventricle, and it is not known whether they have a higher risk of thrombus formation on the surface of them, which can cause embolization.

Even though dual-chamber pacing was ideal, it was not done due to logistic reasons. Nevertheless, the atrial lead also could have been steered into position using the same method.

4. Conclusions

Patent RVOT can provide access for endocardial lead implantation in patients with cavopulmonary shunts.



Fig. 6. Electrocardiogram after the procedure showing consistent pacing of the ventricle.

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Declaration of competing interest

Authors declare no conflict of interests for this article.

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