# Endocardial pacing in a single-ventricle patient with tricuspid atresia- a case report

Ahmad Yamini-Sharif<sup>1</sup>, Ramin Yaghoubian<sup>1</sup>, Homa Ghaderian<sup>1</sup>, and Najme-Sadat Moosavi<sup>1</sup>

<sup>1</sup>Tehran Heart Center

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#### Abstract

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#### Title Page

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- B) Authors:
- 1) Ahmad Yamini-Sharif<sup>1</sup> (first author), ahmadyaminisharif@yahoo.com
- 2) Ramin Yaghoubian<sup>1</sup> (corresponding author), ramin.yaghoubian@gmail.com
- 3) Homa Ghaderian<sup>1</sup>, homaghaderian@gmail.com
- 4) Najme-Sadat Moosavi<sup>1</sup>, ns.moosavi@yahoo.com

<sup>1</sup>Cardiology Department, Tehran Heart Center, Tehran University of Medical Sciences, Tehran, Iran

C) Correspondence to: Ramin Yaghoubian. Address: Department of Cardiology, Tehran Heart Center, Tehran University of Medical Sciences, North Kargar Avenue, Tehran, Iran.

Email address: ramin.yaghoubian@gmail.com / https://orcid.org/0000-0003-1723-157X

#### ABSTRACT

The use of endocardial pacing in patients with univentricular hearts is limited. However, it may be the better option when the risk of surgery is high. We report a case of complete heart block who underwent left ventricular endocardial pacing due to the high risk of epicardial lead placement.

# **KEYWORDS**

Univentricular hearts, Transvenous pacing, Epicardial pacing, Tricuspid atresia, Congenital heart disease, Perirenal fluid

## 1. INTRODUCTION

Multiple congenital heart diseases have progressive conduction disorders, either intrinsic or developed through reparative surgeries.<sup>1</sup> In particular, univentricular hearts are more associated with bradyarrhythmias and complete heart block. Many of these patients need a permanent pacemaker.<sup>2</sup> In univentricular hearts, epicardial pacing has been the first option because of the limited venous access. Also, in the presence of intracardiac shunts, it is the preferred method since there is an increased risk of systemic thromboembolism with the endocardial route.<sup>1-3</sup> However, epicardial leads have considerable disadvantages. One of them is that they have higher chronic pacing thresholds and reduced generator longevity, although this may be improved with the usage of steroid-eluting epicardial leads. Another problem is invasive surgeries used for implanting these leads.<sup>4,5</sup> The present report explains the implantation of a permanent endocardial pacemaker in an adult patient with tricuspid atresia type IIc with severe pulmonary hypertension and complete heart block.

#### 2. CASE PRESENTATION

A 42 years old man from Iraq with frequent episodes of syncope since one month ago was admitted to the Tehran heart center hospital. He had a history of tricuspid atresia type 2c without reparative surgery. On his admission, the patient had fatigue. On physical examination, he had nail clubbing and cyanosis with an O2 saturation of 75%. His electrocardiogram revealed a complete heart block with a right bundle branch block (figure 1). Echocardiography showed tricuspid atresia with transposition of the great arteries (TGA type), severe pulmonary hypertension (mean of PAP:85mmHg), and dominant left ventricle with LVEF:35%. Cardiothoracic CT was done to evaluate better the cardiac structure, which revealed a large pulmonary trunk, hypoplastic ascending aorta, aortic arch with large PDA, and interrupted aorta. Detailed findings of the Cardiothoracic CT scan are shown in figures 2 and 3. In abdominal CT, an accidental finding was bilateral perirenal fluid accumulation (figure 4).

The risk of the surgery for the epicardial lead placement was high due to pulmonary hypertension; on the other hand, the transvenous pacing would increase the risk of systemic thromboembolism. We discussed the risk associated with each procedure with the patient and recommended the endocardial route, which he accepted. Thus, endocardial pacemaker implantation was scheduled for the patient. After prep and drape, local anesthesia and light sedation were conducted. The left subclavian vein access was gained, and a dual-chamber rate-modulated (DDDR) permanent pacemaker was implanted using a peel-away introducer. Atrial lead was inserted in the right atrial auricle. Ventricular lead was advanced through the atrial septal defect and mitral valve and was placed in the common ventricle. The patient was on heparin during the procedure, and lifelong warfarin was initiated for the patient 24 hours after the surgery. On follow-up, the patient was well and had no complaints. The pacemaker was in the proper position without clot formation. The post-pacing electrocardiogram is shown in figure 1.

### 3. DISCUSSION

Epicardial pacing has been a regular option for patients with complex congenital heart disease. Epicardial leads may be placed during cardiac surgery for underlying congenital heart disease with a plan to replace them in the future with transvenous leads.<sup>4</sup>Endocardial pacing of the ventricle for patients with Fontan circulation was impractical due to the lack of transvenous access to the ventricle. With the advances in pacing techniques, ventricular pacing in these patients is possible. Hsieh et al. reported permanent pacing of the left ventricle via coronary sinus in a patient with Fontan circulation and complete atrioventricular (AV) block.<sup>6</sup> In another case report of a single ventricle patient using the endocardial method, DeWitt et al. described successfully placing a ventricular pacing lead through a trans-Fontan-baffle puncture.<sup>7</sup> These techniques may be good alternatives to high-risk epicardial pacing methods in this group of patients.

Endocardial ventricular pacing is technically possible in patients with univentricular hearts who have not had previous reconstructive surgery, but the higher risk for thromboembolism limits this procedure.<sup>2</sup> In a cohort study of 202 patients, transvenous pacemakers increased the risk of systemic thromboembolism in patients with intracardiac shunt by >2-fold, and anticoagulant therapy with warfarin or aspirin did not reduce the risk.<sup>3</sup> However, when the risk of the surgery for epicardial lead placement is high transvenous route may be the better choice. This case was a candidate for a Fontan procedure with epicardial lead placement, but this was a high-risk procedure considering his chronic high pulmonary artery pressure. As a result, we deemed that transvenous lead placement with a tight anticoagulant regimen is the option with the lower risk. According to our research, this is the second report of left ventricular endocardial pacing via the interatrial septum in a single ventricle patient. In a similar study, Scott et al. reported a case of tricuspid atresia with intracardiac shunt and symptomatic chronotropic incompetence. They implanted an endocardial pacemaker in his left ventricle through the atrial septal defect to improve his condition. The patient had no thromboembolic events or complications related to the procedure at the one-year follow-up.<sup>8</sup>

There are few studies comparing the outcomes of dual-chamber pacing versus single-chamber ventricular pacing in patients with single ventricle physiology. Nevertheless, dual-chamber pacemakers are generally considered in these patients because of atrioventricular synchrony, which enhances cardiac output and the capacity to interfere if atrial arrhythmia or sick sinus syndrome occurs.<sup>9</sup>

The bilateral perirenal fluid accumulation was a rare finding in the present case. It has been reported in cases with intracardiac shunt and pulmonary hypertension.<sup>10,11</sup> Pentimone et al. proposed that the underlying mechanism is that pulmonary hypertension increases local hydrostatic pressure in the perirenal veins, leading to fluid leakage to the renal subcapsular space. In their case repeated phlebotomies lowerd hematocrit and reduced perirenal fluid significantly.<sup>10</sup>

In conclusion, while endocardial pacing, in the presence of intracardiac shunts, has the risk of systemic thromboembolism, it may be the safer alternative for epicardial interventions in selected patients.

# AUTHOR CONTRIBUTIONS

AYS was involved in patient management and supervised the case report. RY undertook a comprehensive literature review and wrote the manuscript. HG was involved in patient management and manuscript preparation. NSM provided the images and contributed to their interpretation of them. All authors critically revised and approved the final manuscript.

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None

### CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest.

# DATA AVAILABILITY STATEMENT

Data sharing does not apply to this article as no new data were created or analyzed in this study.

## ETHICAL APPROVAL

Ethical approval for this case report was not required.

#### CONSENT

Written informed consent was obtained from the patient for the publication of this report.

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