

# A Novel pacing option in patients with Right ventricular Endomyocardial Fibrosis: Report of 2 cases

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## Abstract

**Introduction:** Endomyocardial fibrosis (EMF) is characterized by patchy fibrotic thickening of the endocardial and myocardial layers of the heart. Although Sinus node dysfunction and tachyarrhythmia - atrial fibrillation, ventricular tachycardia, have been commonly reported, complete heart block complicating EMF is rare. Transvenous pacing is technically limited by fibrotic obliteration of the affected ventricle that results in poor lead parameters, and alternative pacing strategy like epicardial pacing may be required in many. **Methods and Results:** We present two cases of EMF with complete heart block that were managed by left ventricular pacing through the coronary sinus. **Conclusion:** EMF provides unique challenges to endocardial pacing. Transvenous epicardial pacing through CS tributary using an LV lead can provide a safe and effective alternative mode of pacing with optimal long-term pacing outcome.

## Title page:

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# Introduction

Endomyocardial fibrosis (EMF) is characterized by fibrous tissue deposition on endocardial surfaces leading to impaired filling of one or both ventricles, resulting in either right or left heart failure or both<sup>1</sup>. Chronic heart failure, thromboembolic disease and arrhythmia are some of the associated complications. Incidence of complete heart block (CHB) necessitating a pacemaker is rare in EMF<sup>1</sup>. We report two cases in which the patients had EMF with CHB, who were treated with an alternative pacing strategy.

## Case history

### Case 1:

A 75-year-old male presented with frequent syncopal episodes in the last 4 years, last episode being 20 days prior to the admission in our hospital. There was no history of angina or dyspnea on exertion. There was no history suggestive of any familial cardiac illness and sudden cardiac death.

On clinical examination, he was found to have a pulse rate of 40 bpm with blood pressure of 112/70 mmHg. Cardiomegaly was present with grade 2/6 pansystolic murmur at apex. ECG showed features of atrial flutter with CHB, narrow QRS escape with rate of 35-40 bpm, and incomplete right bundle branch block (Fig 1A). Chest- X- ray showed cardiomegaly. Transthoracic echocardiography (TTE) showed dilated right atrium (RA), apical obliteration of right ventricle (RV), moderate tricuspid regurgitation (TR) with good left ventricular (LV) function. RV angiogram (Figure 1C) in the RAO view shows bipartite RV, calcified and obliterated RV apex, partially obliterated RV body with normal RV inflow and outflow, mildly dilated RV outflow tract, irregularly shaped LV cavity and moderate diastolic mitral regurgitation.

A diagnosis of biventricular EMF with atrial flutter and CHB was made. The patient was planned for permanent pacemaker implantation. Lead parameters at multiple RV positions were suboptimal due to the obliterated RV body apex and body. Hence, coronary sinus was cannulated and LV active fixation lead (Attain Stability®, Model 4796-88 cm, Medtronic, Minneapolis, MN, USA) was positioned at the lateral tributary of coronary sinus. Lead parameters (bipolar threshold 1.3 V @ 0.5 ms, impedance 662 Ohms, R wave 17 mV) were found to be satisfactory. Post procedure fluoroscopy shows good lead position (Fig 1,D-G). Patient is symptom-free with no worsening of lead parameters at 4 years of follow up.

### Case 2:

A 54-year-old male was admitted with history of exertional dyspnea for 18 months. Clinical examination revealed an irregularly irregular pulse at rate of 40 bpm with blood pressure 160/70 mmHg and elevated JVP. Mild cardiomegaly was present with grade 2/6 pan systolic murmur at apex.

ECG (Fig 2A) revealed atrial fibrillation (AF) with ventricular rate of 40 bpm and normal axis. Echocardiography showed dilated RA, obliterated RV apex, moderate TR and mild mitral regurgitation. A 24-hour Holter showed evidence of high-grade AV block.

RV angiography (Fig 3, A and B) shows involvement of RV inflow with calcified and obliterated RV apex, dilated RV outflow and significant TR. Smooth endocardial border was visualized in the LV angiogram suggestive of LV involvement. A diagnosis of biventricular EMF with AF and CHB was made. Electrophysiological study showed high pacing threshold at RV endocardium along with poor R wave sensing. Coronary sinus was cannulated and LV lead (Corox ProMRI 0TW-S, Biotronik) was positioned at the middle cardiac vein distally. Post procedure chest X-ray (Fig 3 C and D) showed optimal lead position. Lead parameters remained stable at 4 years of follow up.

## Discussion

Obliteration of the apices of the affected ventricles due to fibrosis is the hallmark of EM. Endocardial calcification marks the burnt-out phase of EMF<sup>2</sup>. Though conduction abnormalities are common, pacing is rarely required in such patients<sup>1</sup>. Activation pattern of conduction system is impaired by fibrosis which provides substrate for wavelet breaks and reentry. Etiology of heart block in EMF has been much debated.

Subendocardial fibrosis frequently occurs due to the inflammatory reaction which affects the conduction system traversing within, leading to CHB.

These patients had atrial arrhythmia and CHB which demanded pacing. Atrial arrhythmia in RV restrictive physiology suggested significant underlying RA fibrosis, hence VVI was the only viable option and atrial-based pacemakers like AAI or DDD are not appropriate in these cases. For conventional RV endocardial pacing, obliteration of the RV cavity due to EMF is a major concern. In these patients, capture threshold is likely to be high and moreover, local sensing of R wave amplitude is likely to be unacceptably low. In addition, loss of trabeculations in RV will not allow a passive lead fixation as it is being practiced in many countries even now. Tricuspid regurgitation can also lead on to lead instability. Large RA and likely septal involvement on the endocardial aspect of the RV side could be concerns for conduction system (left bundle or His bundle pacing). Epicardial lead placement is an option; however, the high risk profile of these patients makes it not the primary option. Transvenous pacing through coronary sinus tributaries offer many advantages in this case.

There is ample experience with the LV pacing, and long term stability of the CS lead has already been established<sup>3</sup>. Not crossing the tricuspid valve gives an advantage of not worsening the pre-existing TR. Dilated CS tributaries make procedure relatively easier in these cases. Long term stability may demand anchoring of the lead in a large tributary and the use of an active fixation lead a safer option as we did in the first case.

In conclusion, EMF provides unique challenges to endocardial pacing. Transvenous epicardial pacing through CS tributary using an LV lead can provide a safe and effective alternative mode of pacing with optimal long-term pacing outcome.

1. **Author contributions:** Dr. C Sundaram: The principal author. Responsible for the preparation of case report and its editing.
2. Dr. Viswanatha, Kartik S: Responsible for selection of the case and drafting the case report and editing.
3. Dr. Narayanan Namboodiri: The corresponding author. Responsible for performing the procedure, idea, drafting the case report and revising it critically.
4. Dr. Valaparambil, Ajitkumar: Responsible for the performing the procedure mentioned in this case report..

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