

## MINI-FOCUS ISSUE: ARRHYTHMIAS AND EP

ADVANCED

## CASE REPORT: CLINICAL CASE

# Left Pericardiophrenic Vein Pacing for Tachy-Brady Syndrome Due to an Obstructing Cardiac Angiosarcoma



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## ABSTRACT

We describe a case of a permanent pacemaker lead placement via the left pericardiophrenic vein for the treatment of tachy-brady syndrome due to a primary cardiac angiosarcoma. (**Level of Difficulty: Advanced.**)

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## PRESENTATION

A 56-year-old woman with a history of hypothyroidism was transferred to the authors' hospital after presenting to an outside institution with a 3- to 4-month history of progressive dyspnea and left-sided rib pain, as well as a 30-lb unintentional weight loss over the preceding 6 months. A right atrial filling defect was seen on her transthoracic echocardiogram at the outside

institution, which was initially thought to be a thrombus. On admission, the patient was hemodynamically stable with a blood pressure of 134/61 mm Hg, a heart rate of 54 beats/min, a respiratory rate of 16 breaths/min, and oxygen saturation of 98% on room air. A physical examination was significant only for point tenderness of the rib under her left breast with no appreciable cardiac murmurs. There were no initial laboratory abnormalities. During her hospitalization, she had frequent paroxysms of atrial fibrillation and atrial tachycardia with ventricular rates ranging from 180 to 200 beats/min followed by prolonged, symptomatic conversion pauses of 4 to 5 s, as well as junctional bradycardia with rates of 30 to 40 s (**Figures 1A to 1C**). She was initially treated with low-dose metoprolol and amiodarone for rate control and potential suppression of episodes of atrial arrhythmia, however, this further exacerbated the symptomatic conversion pauses and episodic bradycardia.

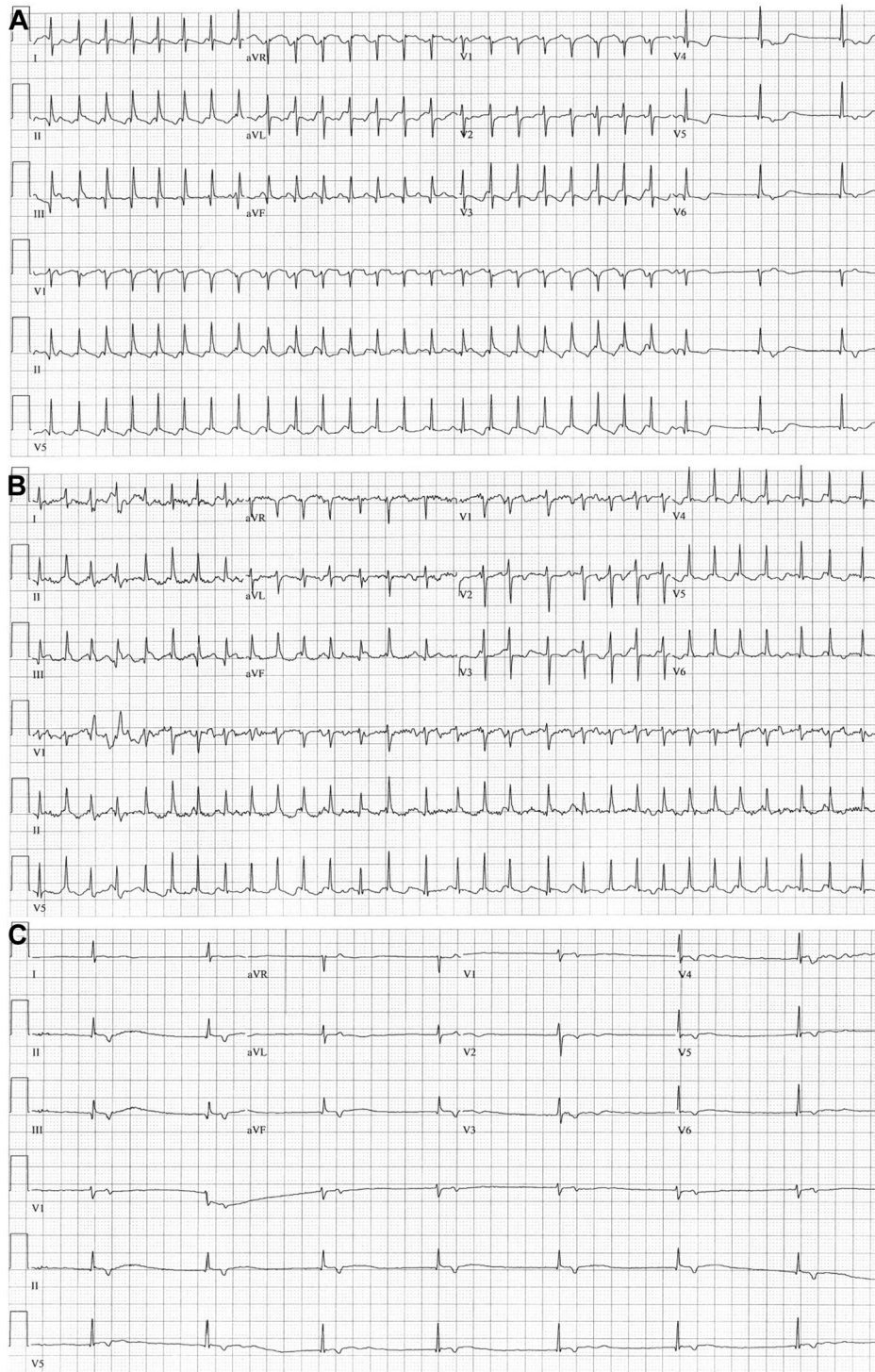
## LEARNING OBJECTIVES

- To recognize cardiac tumors as a potential cause of both tachyarrhythmias and bradyarrhythmias.
- To highlight cardiac vein anatomy as it relates to alternative positions for pacemaker lead placement.

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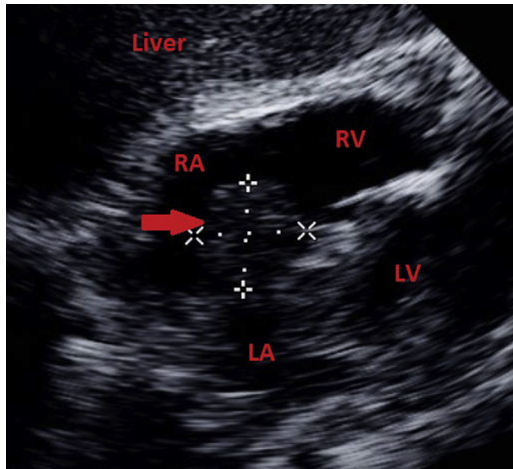
The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* [author instructions page](#).

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**FIGURE 1** Multiple Electrocardiograms Demonstrating the Patient's Various Arrhythmia Episodes

**(A)** Supraventricular tachycardia (SVT) with conversion to junctional bradycardia. **(B)** Atrial fibrillation with rapid ventricular response. **(C)** Junctional bradycardia.

**FIGURE 2** Transthoracic Echocardiogram (Subcostal View) Showing the Right Atrial Mass



The red arrow indicates the right atrial mass. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

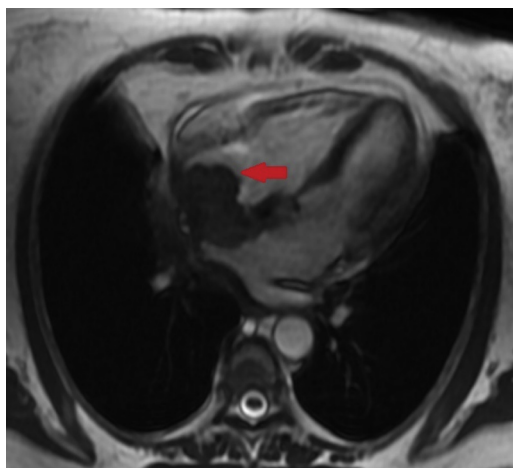
### MEDICAL HISTORY

The patient had a medical history of hypothyroidism on levothyroxine with a family history of malignancy including lung cancer (mother) and melanoma (father).

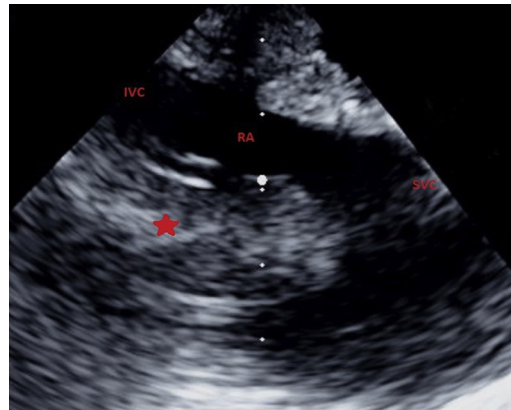
### DIFFERENTIAL DIAGNOSIS

Considering that the patient was a relatively healthy middle-aged woman, the differential diagnosis of her

**FIGURE 3** Cardiac Magnetic Resonance Showing Right Atrial Mass



**FIGURE 4** Transesophageal Echocardiogram (Bicaval View) During Biopsy

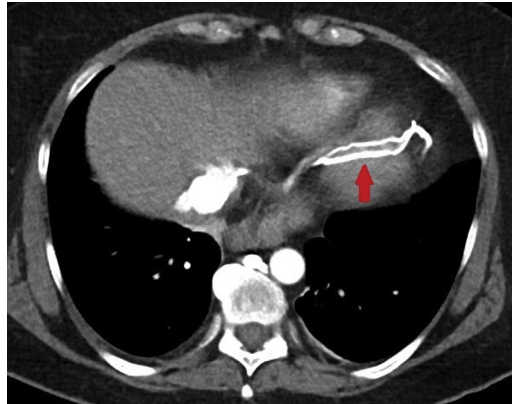


The red star indicates the right atrial mass. IVC = inferior vena cava; RA = right atrium; SVC = superior vena cava.

progressive dyspnea over a 3- to 4-month period included new-onset cardiomyopathy, however, her unintentional weight loss raised suspicion for malignancy.

### INVESTIGATIONS

Workup included a transthoracic echocardiogram, which showed a right atrial mass with poor definition of its borders (Figure 2). Cardiac magnetic resonance showed a large, solid, and nearly completely enhancing soft tissue mass, the bulk of which was located in the posterior septal wall of the right atrium measuring approximately 4.7 × 4.7 × 7.5 cm (Figure 3). The mass spanned the mid to lower superior vena cava (SVC) into the low, right atrium near the border with the inferior vena cava (IVC) but did not fully obstruct IVC inflow or the coronary sinus. There was extracardiac spread of the mass into the pericardial space surrounding the posterior right lateral circumference of the aorta as well as the upper lobe branches of the right pulmonary artery and at least 1 of the branches of the right upper pulmonary vein. The mass also extended around the right lateral aspect of the left atrium to the level of the incoming coronary sinus. In addition, there was complete atelectasis of the right middle lobe, likely due to mass effect, as well as extension around the right middle lobe bronchus. Preliminary diagnosis based on the cardiac magnetic resonance was that of an angiosarcoma. As a tissue diagnosis was required for treatment planning, the patient underwent transesophageal echocardiography-guided biopsy

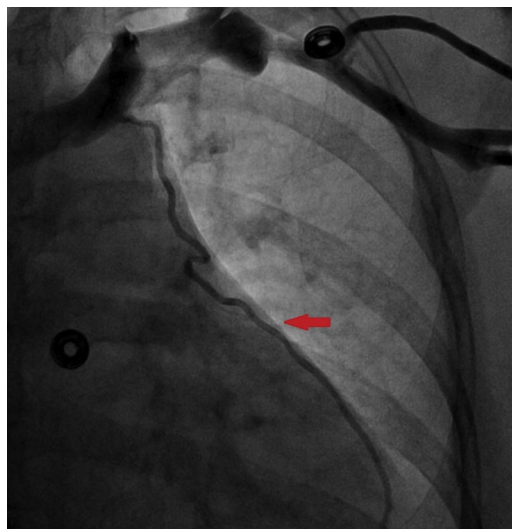
**FIGURE 5** Computed Tomography Venogram Showing Course of the Pericardiophrenic Vein

The **red arrow** indicates the left pericardiophrenic vein.

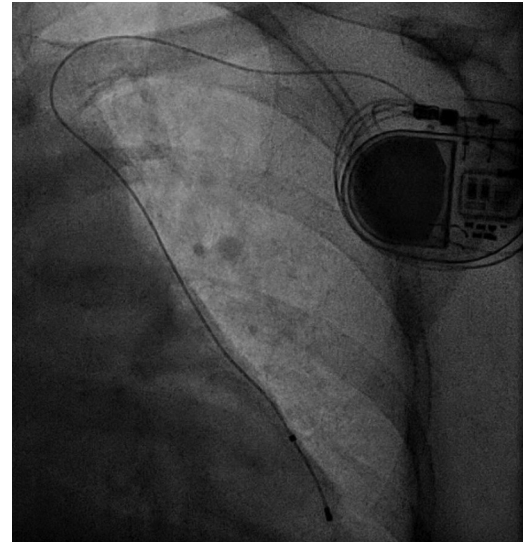
(Figure 4), which confirmed the diagnosis of an angiosarcoma.

### MANAGEMENT

Given the patient's symptomatic tachy-brady syndrome, implantation of a permanent pacemaker (PPM) was the definitive treatment option. As there was an inability to place a conventional temporary or

**FIGURE 6** Fluoroscopic View of the Pericardiophrenic Vein

The **red arrow** indicates the left pericardiophrenic vein.

**FIGURE 7** Final Fluoroscopic View of the Permanent Pacemaker Implant

permanent pacing system due to SVC and IVC obstruction from the mass with deferment of a thoracotomy for epicardial lead placement due to the potential need for additional thoracic surgery in the future, an individualized approach to pacing was required to prevent symptomatic bradycardia and conversion pauses. Computed tomography of the chest with upper extremity venography revealed an accessory epicardial/pericardial vein extending from the left brachiocephalic vein along the left ventricle, consistent with the left pericardiophrenic vein (LPPV) (Figure 5).

The patient underwent implantation of a single-chamber PPM with placement of a bipolar left ventricular lead in the LPPV (Figures 6 and 7). Multiple pacing locations were mapped along the course of the LPPV, testing at various outputs and pulse widths; however, every location where there was ventricular capture, there was concomitant phrenic nerve capture. Because the patient's predominant symptoms were related to conversion pauses and because brief periods of phrenic nerve stimulation were not excessively uncomfortable for the patient, keeping in mind the desire to avoid an additional procedure (surgical epicardial lead placement), the decision was made to place the bipolar lead in the location with the lowest threshold and use it for demand ventricular pacing at a rate of 30 beats/min. After PPM implantation, antiarrhythmic therapy was increased with improvement in the patient's tachyarrhythmia episodes with only minimal ventricular pacing.

## DISCUSSION

Primary cardiac angiosarcoma is a rare cardiac malignancy with a typically poor prognosis (1). The age at diagnosis is generally in the third to fifth decade of life, and the median survival is <10 to 24 months with incomplete and complete resection, respectively (1-4). Presenting symptoms can include unexplained weight loss, dyspnea, thoracoabdominal pain, syncope, peripheral edema, heart failure, and palpitations (5). Given the tumor's predilection for the right atrium (3), mass effect can lead to vena caval obstruction, arrhythmia, and potential conduction disturbances (4). SVC and IVC obstructions can pose a unique problem when symptomatic conduction disturbances occur due to infiltration of a tumor into the cardiac conduction system. Without direct access to the right-sided cardiac chambers through the vena cavae for temporary or permanent pacing, an unconventional approach should be considered and sought, as was shown in the present case. The LPPV is a vessel located on the lateral border of the heart which courses adjacent to the pericardium and drains into the left brachiocephalic vein (6,7). As its name implies, the LPPV courses within the left pericardiophrenic bundle, adjacent to the phrenic nerve (8), which led to phrenic nerve stimulation with pacing in this location. It has been shown that phrenic stimulation after placement of a coronary sinus lead for cardiac resynchronization therapy is safe and effective (9,10), and in the present patient, this led to only

occasional episodes of mild discomfort. Pacemaker implantation using the LPPV in this patient was an appropriate temporizing measure which allowed her to undergo further treatment of her cardiac angiosarcoma.

## FOLLOW-UP

After 8 months of neoadjuvant chemotherapy, the patient underwent surgical resection of the angiosarcoma with reconstruction of the right atrium and SVC at another institution. Following this procedure, her burden of phrenic nerve stimulation increased. Pacemaker system revision was performed in which the LPPV lead was removed and replaced with a His bundle lead, a procedure facilitated by the newly reconstructed anatomy.

## CONCLUSIONS

Right atrial obstruction can complicate the treatment of symptomatic bradycardia. Without access to the right-sided cardiac chambers for pacing, unconventional access may be considered. Although it is not an ideal route for pacing given its proximity to the left phrenic nerve, the LPPV facilitated treatment of both bradycardia and atrial tachyarrhythmia.

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**KEY WORDS** bradycardia, cancer, cardiac magnetic resonance, cardiac pacemaker, computed tomography, electrophysiology, imaging, supraventricular arrhythmia, weight loss