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# IMAGES in PAEDIATRIC CARDIOLOGY

Wittekind SG,<sup>1</sup> Salerno JC,<sup>2</sup> Rubio AE.<sup>2</sup> Pacemaker-associated cyanosis in an adolescent: The answer hiding behind shadows. *Images Paediatr Cardiol* 2012;14(4):6-10.

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## Abstract

Lead thrombosis is a recognized complication of permanent transvenous pacemaker (PM) implantation. We present the interesting case of an adolescent with a dual-chamber PM presenting with fatigue and hypoxemia. Due to limitations of various imaging modalities, the diagnosis was difficult. She was eventually diagnosed with intracardiac PM lead thrombi obstructing tricuspid valve inflow. The pediatric literature on PM lead thrombosis is also briefly reviewed.

## Introduction

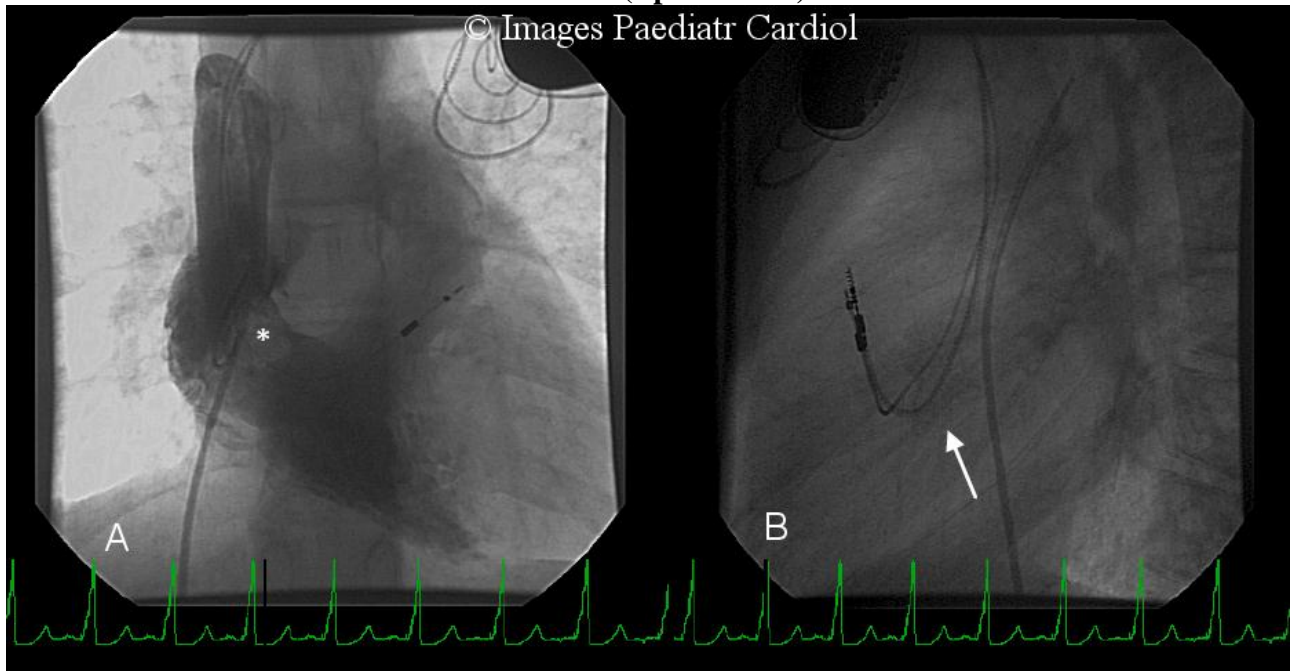
Most PM lead-associated venous thrombi are asymptomatic and screening studies suggest an incidence between 20-30%.<sup>1</sup> Intracardiac thrombi are less common than venous thrombi and can be missed with standard venography or transthoracic echocardiography (TTE). The reader should be alerted to the significant incidence, difficulty in diagnosis, and potential morbidity of this complication.

## Case Report

A 13 year-old female with congenital complete atrioventricular (AV) block presented to cardiology clinic with progressive exertional fatigue. She had a permanent dual-chamber transvenous PM that was implanted at 5 years of age, and the generator was replaced 5 years later. On TTE there was mild tricuspid valve inflow obstruction deep in the RA at the level of the atrial lead with mean Doppler gradient of 3.9 mmHg. The tricuspid valve appeared normal and there was no valvular stenosis or regurgitation. There was no atrial shunt detected. The estimated RV systolic pressure was 20.4 mmHg above RA pressure. A cardiopulmonary stress test demonstrated decreased exercise capacity with 94% oxygen saturation at rest, decreasing to 77% with exercise, but was otherwise unremarkable. Chest CT with contrasts did not reveal evidence of pulmonary arteriovenous malformations or thromboembolism. The chest CT was confounded by a significant amount of metal artifact caused by the transvenous pacing leads. A stress echocardiogram was performed to further assess the hypoxemia with exercise. No evidence of pulmonary hypertension during exercise was noted, but there was a right-to-left shunt through a small intra-atrial communication that only became evident at peak exercise. Acoustic shadowing from the pacing leads again obscured any detailed assessment of the tricuspid inflow obstruction.

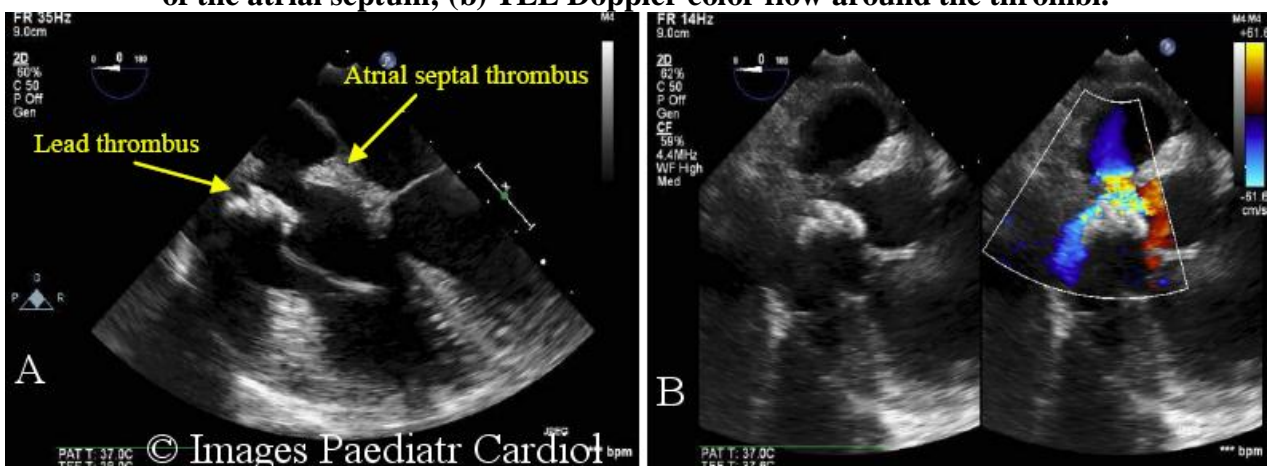
During the next few months she experienced an interval worsening of fatigue at rest as well as dizziness and headaches. She was scheduled for cardiac catheterization to measure right heart and pulmonary artery (PA) hemodynamics and to determine candidacy for PFO closure. At the onset of the study angiography was performed and a large calcified mass at the superior vena cava-right atrium (SVC-RA) junction created a filling defect (Figure 1).

**Figure 1: (a) Angiographic evaluation of the right SVC and the right atrial chamber delineating a filling defect (\*) within the RA around the leads; (b) Straight lateral fluoroscopic projection demonstrating a calcified area encompassing the leads, representative of a calcified thrombus (tip of arrow).**



PA pressures were normal. There were no filling defects in the pulmonary segments. A transesophageal echocardiogram (TEE) confirmed two atrial thrombi; one at the SVC-RA junction associated with the pacing leads, measuring 1.9 x 0.6 cm, and the other at the inferior atrial septum, measuring 1.7 x 0.8 cm (Figure 2).

**Figure 2: (a) Two dimensional imaging via transesophageal echocardiogram (TEE) of a hyperechoic thrombus on the atrial lead and a second thrombus located on the inferior aspect of the atrial septum; (b) TEE Doppler color flow around the thrombi.**



The mean gradient across the SVC-RA junction was 3 mmHg. Closure of the PFO was delayed and the patient was started on enoxaparin. A hypercoagulability work-up was negative.

The case was discussed at cardiac surgery conference and one week later she went to the operating room for a right atrial thrombectomy, primary closure of the PFO, and conversion to an epicardial pacing system. Intraoperative inspection within the RA showed the atrial and ventricular pacing leads surrounded by a calcified thrombus measuring 3 x 2 cm (Figure 3). Complete surgical extraction was performed along with successful placement of the epicardial pacing system. The patient's hospital course was complicated by a mild left-sided pleural effusion which resolved on diuretics. On follow-up 4 months post-operatively the patient had resolution of dizziness and fatigue. She had normal resting oxygen saturations and TTE showed normal tricuspid inflow and no recurrence of thrombus.

**Figure 3: Explanted lead along with the calcified thrombus.**



## Discussion

This case is unique in that intracardiac transvenous PM lead thrombosis causing hypoxemia has not been commonly reported in pediatric patients. There is a growing body of evidence demonstrating that the incidence of lead thrombosis as a complication of transvenous PMs may be underestimated. Much about the pathophysiology of lead thrombosis remains unknown.

An important study by Figa et al. in 1997 prospectively investigated the theory that a larger transvenous lead burden in a smaller patient might confer increased risk of venous thrombosis and occlusion.<sup>2</sup> Contrast venography was used as the gold standard to detect venous obstruction in 63 pediatric patients. An INDEX score, defined as the sum of the cross-sectional area of all leads indexed to body surface area at implant, greater than 6.6 mm<sup>2</sup>/m<sup>2</sup> predicted venous obstruction with a sensitivity of 90% and a specificity of 84%. For a decade this score was routinely used in the decision to implant a transvenous versus epicardial pacing system. The validity of the INDEX was challenged in 2006 by Bar-Cohen, et al.<sup>3</sup> Their retrospective analysis of 85 consecutive patients who underwent venography revealed a far less impressive sensitivity of 36% and specificity of 69%. Patient age and total lead duration were not associated with risk of thrombosis. The overall rate of venous occlusion was 25% (13% partial, 12% total). Adult studies have demonstrated similar rates of venous occlusion.<sup>4,5,6</sup> To date a single risk factor for lead thrombosis has not been identified. Current research is investigating hypercoagulability, inflammation, and relative low flow states

such as heart failure or pulmonary hypertension as potential risk factors. In a recent paper by Supple, et al. Intracardiac echocardiography (ICE) was used to evaluate the incidence of intracardiac lead thrombi in a cohort of 86 consecutive adults with implantable devices undergoing ablation procedures.<sup>7</sup> Notably, the majority had congestive heart failure due to cardiomyopathy. Lead thrombi were seen with ICE in 30% of patients but were seen on TTE in only 1 of these identified patients. Thrombi were mostly seen in the RA versus the RV. Of all patient characteristics measured only elevated PA systolic pressure (PASP), as estimated from the tricuspid regurgitation jet velocity, was significantly associated with the incidence of thrombi. Further investigation, particularly in smaller patients without heart failure, is needed to establish the clinical significance of this finding to the pediatric population.

In this present case, the lead thrombi were predominately intracardiac and were not easily identified on TTE. This is not surprising since studies have shown that TEE is the method of choice for diagnosis of such thrombi. A 2006 cohort study of 66 adult patients screened for lead thrombi at 6 months post-implant demonstrated that all lead thrombi in the RA, identified by TEE, could not be visualized by TTE.<sup>6</sup> However, TEE could not diagnose subclavian or innominate venous thrombi, which were far more common. This suggests that multiple imaging modalities including venography and TEE are needed to fully evaluate for vascular obstruction and intracardiac thrombus.

Due to the significant incidence of lead associated thrombi, investigation for the presence of an obstructive thrombosis should be undertaken when a patient with a transvenous PM presents with atypical symptoms including fatigue, presyncope, and dyspnea. This may be especially prudent in pediatric patients who undergo transvenous dual chamber pacemaker implantation at a young age. Since no risk factors for lead thrombosis have been validated, it is not currently possible to risk-stratify patients. A thorough history and physical, followed by ECG, PM interrogation, and chest radiography constitutes an appropriate initial evaluation. If this work-up is negative then imaging for thrombi should be done progressing from less to more invasive, beginning with TTE, then TEE, and finally venography.

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