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Partial anomalous pulmonary venous return with dual drainage leading to pacemaker lead malposition

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The patient, a 76-year-old woman, with a history of hypertension, paroxysmal atrial fibrillation on oral anticoagulation therapy and New York Heart Association class II chronic heart failure. At age of 47, she underwent surgical closure of type 2 atrial septal defect. At 73, a dual-chamber pacemaker was implanted due to symptomatic tachycardia-bradycardia syndrome. Post-operative anteroposterior chest X-ray showed normal lead positioning. The electrocardiogram showed sequential pacing at 70 bpm, with a right bundle branch block (RBBB) morphology.

During subsequent outpatient follow-up, a transthoracic echocardiogram (TTE) detected one of the pacemaker leads within the left ventricle (LV) (Figure 1A). However, the TTE could not clarify the pathway of the lead into the LV. Although a patent foramen ovale was identified, the ventricular lead did not appear to traverse it. Transoesophageal echocardiography visualized the lead coursing through the superior vena cava (SVC) into right upper pulmonary vein (RUPV), then left atrium (LA), and ultimately LV. (Figure 1B–E). Nevertheless, the precise pathway of the lead from the SVC to the LA remained unclear. Based on echocardiographic findings, a fistula between the RUPV and SVC was suspected.

To further investigate, the patient underwent contrast-enhanced multi-slice computed tomography of the heart, which revealed a vascular anomaly — an abnormal connection between the

RUPV and SVC, measuring 8.5 × 7.0 mm. This finding was consistent with partial anomalous pulmonary venous return (PAPVR) with dual drainage to the SVC and LA (Figure 1 F–H).

Given the high surgical risk and the patient's decision to decline surgical intervention, she was scheduled for transvenous lead extraction. The procedure was performed without complications using a cerebral circulation protection system. The LV lead was successfully removed after screw retraction with manual traction. Subsequently, a new lead was implanted into the right ventricle. The patient remains under outpatient follow-up.

Inadvertent lead malposition in the left heart is a rare but recognized complication of pacemaker implantation, with an estimated incidence of 0.34% of procedures [1]. On electrocardiography, incorrect lead placement in the LV should be suspected when lead V1 exhibits a RBBB morphology. However, “pseudo-RBBB” has been reported in patients with correctly positioned leads in the right ventricle and can represent a desired QRS morphology in the era of conduction system pacing [2]. The definitive lead position can be confirmed through TTE.

The most common causes of incorrect lead placement in the left heart are congenital defects such as patent foramen ovale, atrial septal defect, or ventricular septal defect [3, 4]. Isolated PAPVR with a double connection of the RUPV to the SVC and LA is extremely rare congenital defect. To date, only three cases PAPVR with dual drainage have been reported [5]. This case represents the first description in the literature of a pacemaker lead pathway involving PAPVR into the left heart. The defect was undiagnosed preoperatively and remained untreated until 25 years later, when the lead malposition enabled a proper diagnosis. Based on this experience, malpositioned leads in the LV due to such anomalies can be safely and effectively removed using transvenous lead extraction.

Article information

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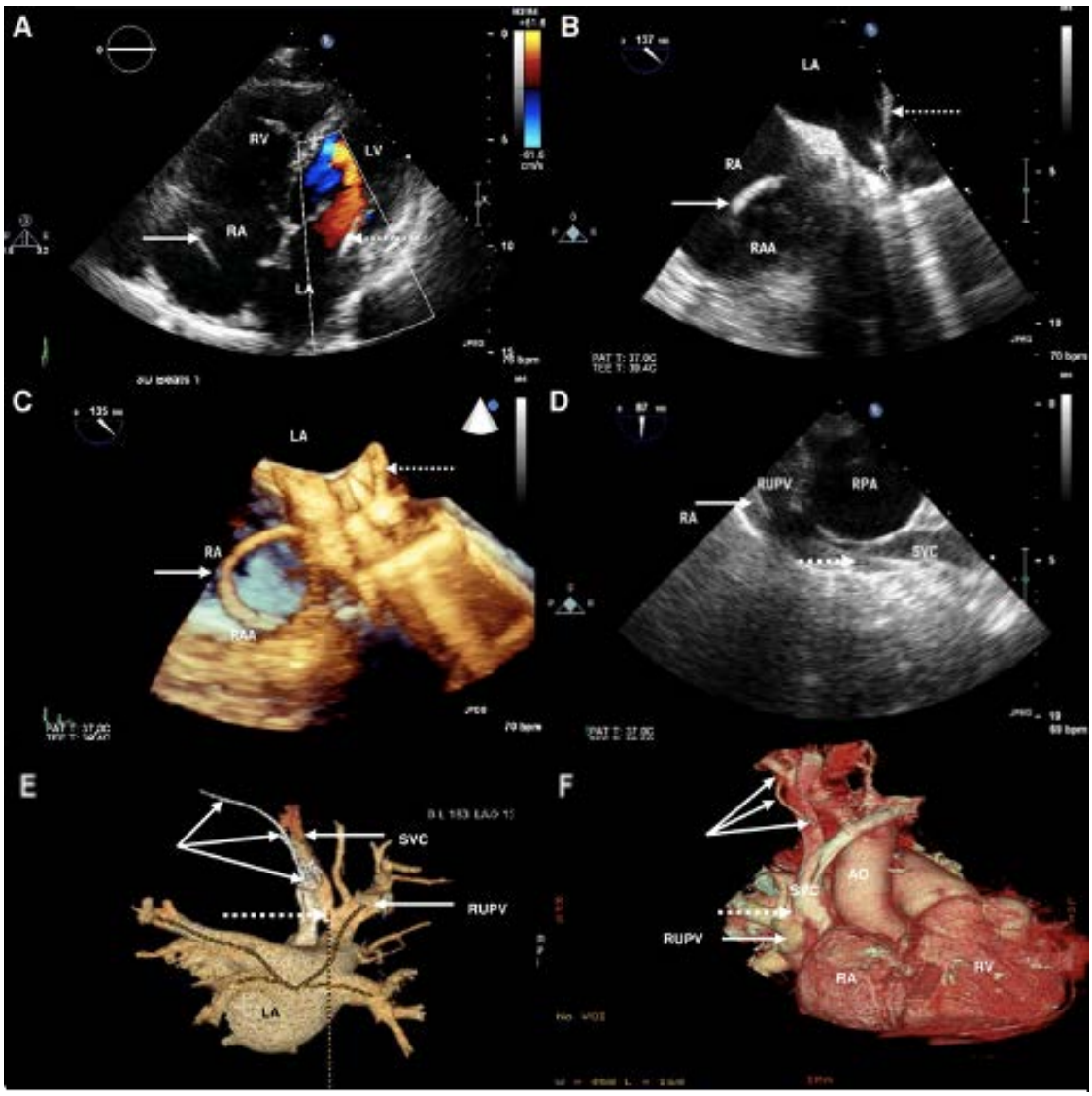

Figure 1. A. Echocardiography — TTE 2D modified 4C: visible one lead in RA (solid arrow) and other lead penetrating from LA to LV through the mitral valve (dashed arrow). **B.** Echocardiography — TEE 2D: visible one lead in RA (solid arrow) and other in LA (dashed arrow). **C.** Echocardiography — TEE 3D: visible one lead in RA (solid arrow) and other in LA (dashed arrow). **D.** Echocardiography — TEE 2D: visible lead in SVC (dashed arrow) and in RUPV (solid arrow). **E.** Cardiac tomography — MSCT VRR: visible lead (triple solid arrows) and anomalous connection of RUPV to SVC size 8.5 × 7.0 mm (dashed arrow). **F.** Cardiac tomography — MSCT VRR: visible lead (triple solid arrows) and anomalous connection of RUPV to SVC size 8.5 × 7.0 mm (dashed arrow)

Abbreviations: AO, aorta; 4C, four-chamber apical view; LA, left atrium; LV, left ventricle; MSCT MPR, multi-slice computed tomography multiplanar reconstruction; MSCT VRR, multi-slice computed tomography volume rendering reconstruction; RA, right atrium; RAA, right atrium appendage; RPA, right pulmonary artery; RUPV, right upper pulmonary vein; RV, right ventricle; SVC, superior vena cava; TEE, transoesophageal echocardiography; TTE, transthoracic echocardiography; 3D, tree-dimensional; 2D, two-dimensional