

A case of cardiac strangulation following epicardial pacemaker implantation in an adult: a case report

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Received 10 August 2021; first decision 9 November 2021; accepted 9 February 2022; online publish-ahead-of-print 23 February 2022

Background

Cardiac strangulation (CS) is a rare but potentially devastating complication caused by the leads of an epicardial pacemaker (EP). Most cases have been reported in paediatric patients, and there has been no report wherein the diagnosis was made in a living, adult patient, and treated successfully.

Case summary

A 31-year-old woman with a history of atrial septal defect (ASD) patch closure and EP implantation for congenital atrial stand-still presented with dyspnoea on exertion. The blood investigation of the patient showed liver dysfunction, chest radiography showed pulmonary artery dilatation, and transthoracic echocardiography showed right chambers dysfunction. Right heart catheterization showed haemodynamics similar to those of constrictive pericarditis, eventually leading to the diagnosis of CS due to EP leads. The patient was successfully operated upon.

Discussion

We reported the first case where CS was diagnosed in adulthood and successfully treated with surgical intervention. Cardiac strangulation is challenging to diagnose because of the small number of cases reported and the lack of definitive diagnostic algorithms or criteria. Surgical EP lead removal should be performed without hesitation in cases where CS is considered the primary aetiology of critical symptoms or complications because surgical removal is the only fundamental treatment for CS. In addition, paediatric patients undergoing EP implantation need for close follow-up.

Keywords

Cardiac strangulation • Epicardial pacemaker • Adult • Case report

ESC Curriculum

5.9 Pacemakers • 6.3 Heart failure with preserved ejection fraction • 6.6 Pericardial disease • 7.5 Cardiac surgery

Learning points

- Cardiac strangulation is a mechanical complication that occurs when the leads of an epicardial pacemaker tighten around the heart and coronary vessels, impairing cardiac growth, and function.
- Most cases of cardiac strangulation have been reported in the paediatric age group so far. This case stands out because it was diagnosed in an adult and treated successfully.
- Surgical removal of the epicardial leads forms the cornerstone of management of this rare but life-threatening condition.

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Handling Editor: Diego Araiza-Garaygordobil

Peer-reviewers: Edin Begic; Diego Araiza-Garaygordobil

Compliance Editor: Sara Monosilio

Supplementary Material Editor: Jonathan Senior

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Introduction

Cardiac strangulation (CS) is a mechanical complication that occurs when the leads of an epicardial pacemaker (EP) cause a discrepancy in cardiac growth by tightening around the heart and coronary vessels.^{1–4} To date, CS lacks consistent diagnostic and therapeutic algorithms because of its rarity, thereby delaying its diagnosis and treatment. Most reports of CS have been on paediatric patients, and few cases have been reported in adults.³ Moreover, reports wherein an adult patient was promptly diagnosed and successfully treated are absent from the literature. Here, we report the first case where CS was diagnosed in adulthood and successfully treated with surgical intervention.

Timeline

Time	Event
At birth	Diagnosis of congenital atrial stand-still and atrial septal defect (ASD).
6 years old	ASD patch closure and epicardial pacemaker (EP) implantation.
17 years old	1st generator exchange.
23 years old	2nd generator exchange.
30 years old	EP generator was extracted and replaced with a transvenous endocardial pacemaker.
31 years old (25 years after the EP lead implantation)	Complaints of dyspnoea on exertion. Haemodynamic findings similar to constrictive pericarditis and the existence of EP led to a diagnosis of cardiac strangulation. Surgical EP lead removal.
33 years old (2 years after the EP lead removal)	The patient remained asymptomatic.

Case presentation

A 31-year-old woman with complaints of mild dyspnoea on exertion over the last several years was referred to our institution for liver dysfunction and cirrhosis. She had a significant medical history of congenital atrial stand-still and atrial septal defect (ASD) at birth. She had undergone no interventions immediately after the birth and ASD patch closure and EP implantation at the age of 6 years. Her generator was exchanged at the ages of 17 and 23 years. Although the EP generator was extracted and replaced by a transvenous endocardial pacemaker (ventricular sensing and pacing) at the age of 30 years, EP leads had been abandoned after the transvenous endocardial pacemaker implantation. The patient had no relevant family history.

At initial presentation, her blood pressure was 131/80 mmHg, pulse rate was 60 beats per minute (b.p.m.), and her oxygen saturation was 98% at rest. Physical examination was normal, save for mild peripheral oedema. An electrocardiogram showed a ventricular

pacing rhythm with a heart rate of 60 b.p.m. Blood investigations revealed thrombocytopenia with a platelet count of 98 000/ μ L (reference range 158 000–348 000/ μ L); mild liver dysfunction; and an elevated B-type natriuretic peptide level of 41.9 pg/mL (reference range <18.4 pg/mL). Coagulation capacity was within the normal range. Antibody titres related to autoimmune disease, hepatitis viruses, and human immunodeficiency virus were negative. Tumour markers and interferon-gamma release assay results were negative. Chest radiography revealed bilateral enlargement of the pulmonary artery with cardiomegaly, EP leads with the loop placed on the anterior surface of the heart, and the endocardial pacemaker (Figure 1). Computed tomography (CT) scan of the thorax showed calcified EP leads on the anterior surface of the heart; CT scan of the abdomen revealed surface nodularity of the liver suggesting cirrhosis and mild ascites, with no evidence of spontaneous portosystemic shunts other than the splenorenal shunt (Figure 2). Transthoracic echocardiography demonstrated diffusely reduced left ventricular wall motion with an ejection fraction of 49%, bi-atrial enlargement, moderate mitral regurgitation due to left atrial enlargement, and dilatation of the inferior vena cava. There was no evidence of ventricular septal diastolic shudder, respiration-related ventricular septal shift, intracardiac shunt including residual ASD, or pericardial effusion. On pulsed-wave tissue Doppler imaging, the lateral and medial velocities of the mitral annulus in early diastole (e') were 16.5 cm/s and 9.5 cm/s, respectively. The tricuspid annular plane systolic excursion was 10.9 mm and the tricuspid annular velocity in systole (s') was 6.6 cm/s. Coronary angiography revealed no evidence of coronary stenosis. Right heart catheterization (RHC) showed a right atrial pressure of 14 mmHg, pulmonary artery pressure of 36/16 mmHg, mean pulmonary artery pressure of 24 mmHg, pulmonary capillary wedge pressure of 18 mmHg, cardiac index of 2.48 L/min/m², pulmonary artery saturation of 78%, and waveforms similar to those of constrictive pericarditis (CP) (Figure 3). These results suggested that she had developed cardiac strangulation by the EP leads with haemodynamics similar to those of CP, leading to congestive hepatopathy with cirrhosis. Upon arriving at this diagnosis, complete EP lead removal and mitral valve repair were successfully performed (Figure 4).

After surgical removal, her dyspnoea on exertion was improved. Further, RHC 1 year after surgery showed a right atrial pressure of 6 mmHg, pulmonary artery pressure of 19/12 mmHg, mean pulmonary artery pressure of 15 mmHg, pulmonary capillary wedge pressure of 13 mmHg, cardiac index of 3.46 L/min/m², and pulmonary artery saturation of 80%. The waveforms of RHC after surgery also changed compared with those before surgery (Figure 5). The medication administered immediately after surgery included a beta-blocker, angiotensin-converting enzyme inhibitor, mineralocorticoid receptor antagonist, loop diuretic, and vitamin K antagonist. However, all medication has since been stopped because of pregnancy. The patient has shown no worsening of heart failure or other unanticipated events until the present time.

Discussion

We present a rare case of CS diagnosed in an adult patient, who was treated successfully by removal of the offending EP leads. Cardiac strangulation is caused when EP leads are implanted in childhood and

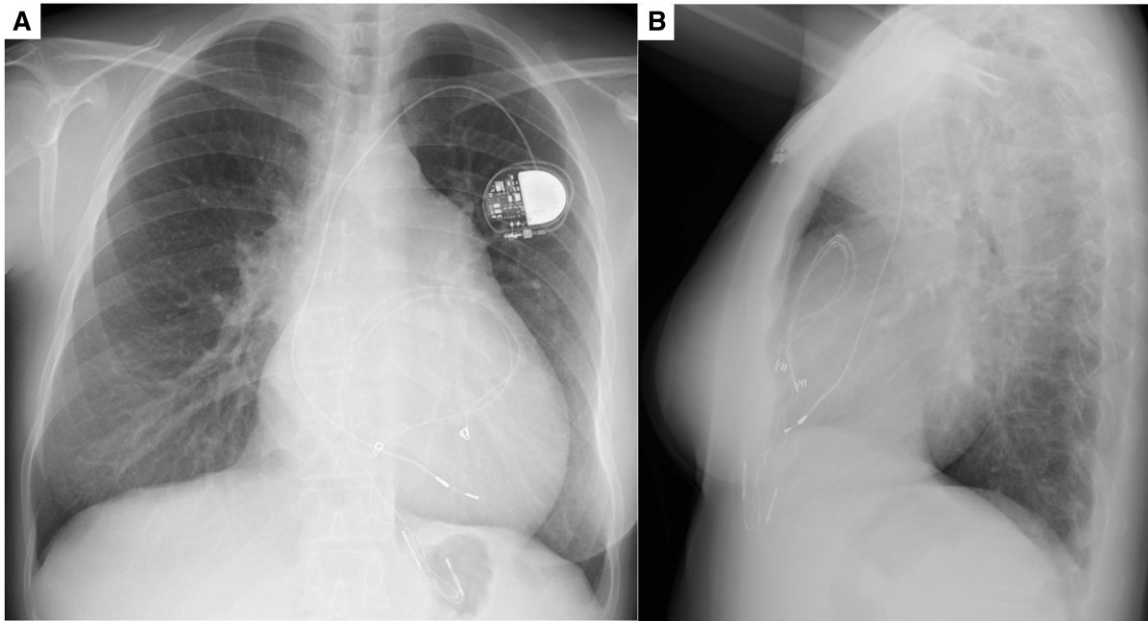


Figure 1 Chest X-ray before epicardial pacemaker lead removal. Posteroanterior and lateral chest radiographs (A and B, respectively) showing bi-lateral enlargement of the pulmonary artery with cardiomegaly, epicardial pacemaker leads with the loop placed on the anterior surface of the heart, and the endocardial pacemaker.

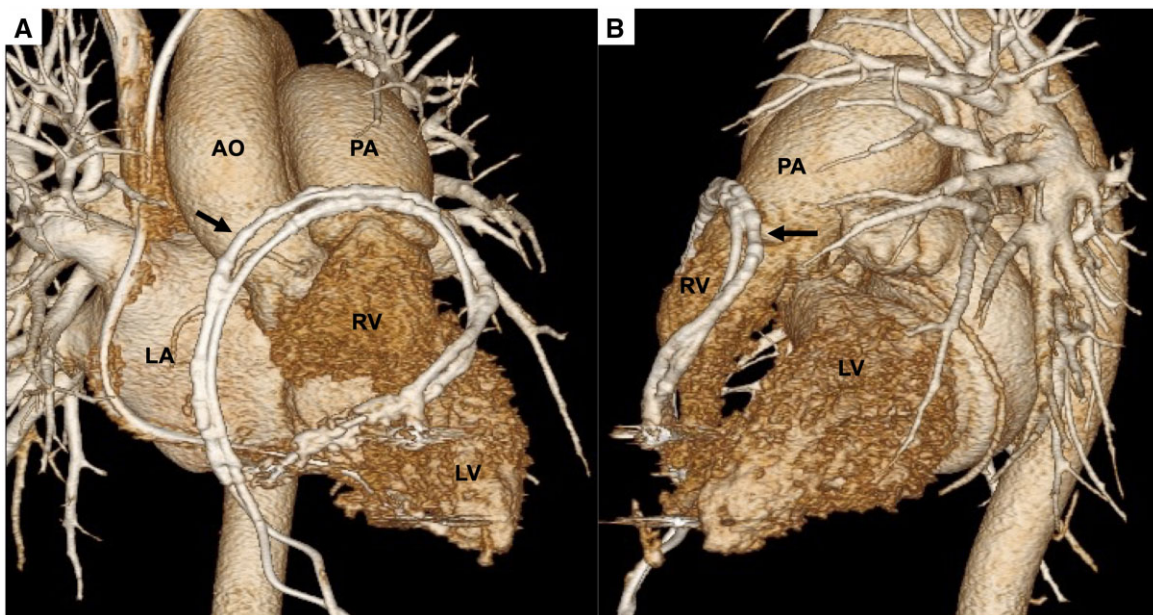


Figure 2 Three-dimensional computed tomography before removal of the epicardial pacemaker. The frontal view of the three-dimensional computed tomography scan revealing that the epicardial pacemaker lead loop (arrow) was placed on the anterior surface of the heart (A). The lateral view revealing that the epicardial pacemaker lead strangled the heart, especially the right ventricle and main pulmonary artery (B). AO, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RV, right ventricle.

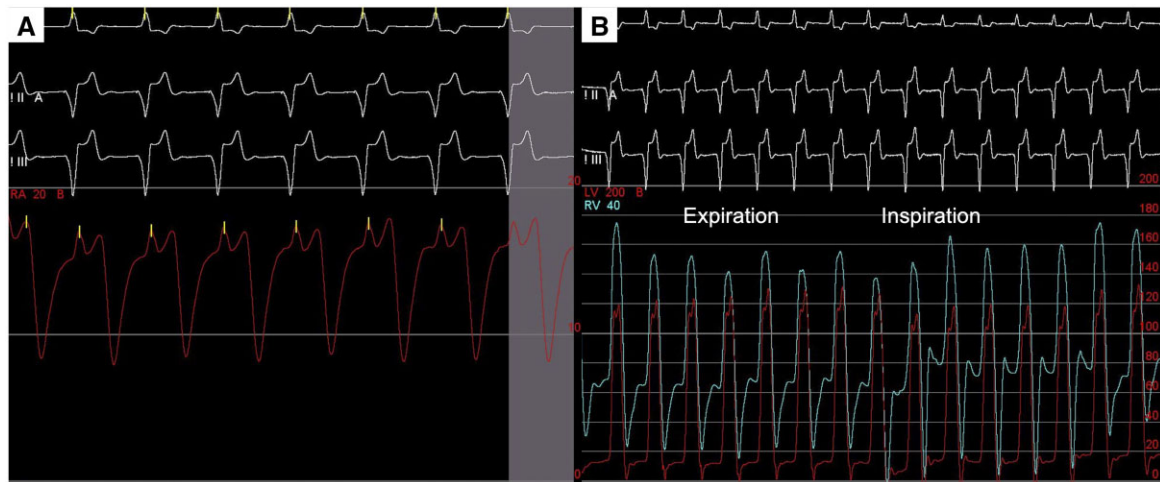


Figure 3 Haemodynamic tracings before epicardial pacemaker leads removal. Right heart catheterization before the surgery showing a steep y descent waveform with a pressure of 14 mmHg in the right atrium (A), a 'dip and plateau' waveform in the right ventricular pressure, and dysynchrony between the right and left ventricular systolic pressure during the respiratory cycle (B). LV, left ventricle; RA, right atrium; RV, right ventricle.

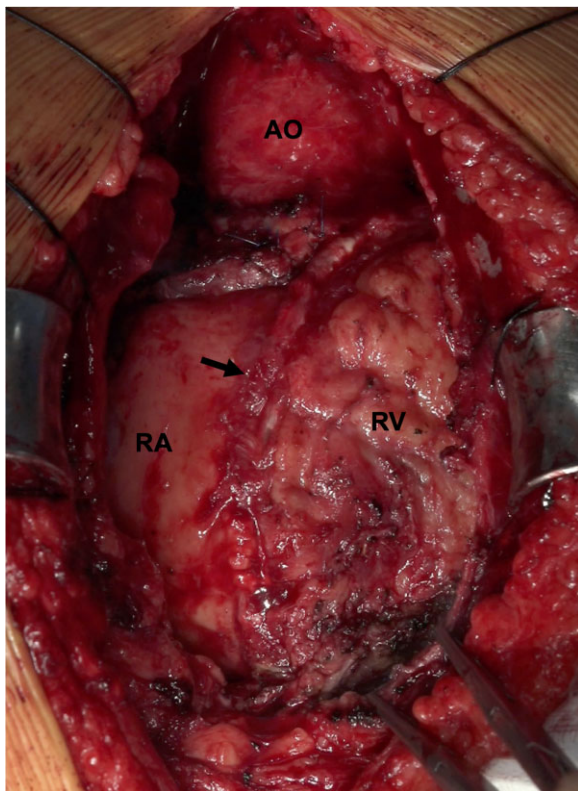


Figure 4 Intraoperative findings. Intraoperative findings showing the epicardial pacemaker lead (arrow) adherent to the anterior surface of the right ventricle. AO, aorta; RA, right atrium; RV, right ventricle.

was first reported by Brenner *et al.* in 1988.⁵ As the child grows, the EP leads strangle the heart, causing dizziness, syncope, arrhythmia, angina pectoris, myocardial infarction, heart failure, valvular diseases, and sudden cardiac death, but non-specific or asymptomatic cases have also been reported.^{2,6} Cardiac strangulation is difficult to diagnose because of the small number of cases reported and the lack of definitive diagnostic algorithms or criteria, which may contribute to the variability in its reported prevalence.

Cardiac strangulation in adults is extremely rare; only two cases have been reported, both of which resulted in death.^{3,7} In this regard, it is interesting to note that a favourable outcome in an adult-onset CS was obtained in this case. Furthermore, our patient developed heart failure with RHC findings similar to those of CP, but the possibility of collagen diseases and cancers as the cause of CP was ruled out. Although we could not rule out the possibility of CP due to the history of cardiac surgery in childhood, we diagnosed CS because of the history of EP lead implantation in childhood. There have been no previous reports of CS demonstrating haemodynamics similar to those of CP, as was seen in this case.

The only approach to treat CS is to remove the EP leads surgically, so as to release the physical strangulation. All patients with CS underwent surgical EP lead removal in previous reports,^{2-5,8} and there have been no reports of conservative treatment. The optimal medical therapy for patients with CS has not been established. The only fundamental treatment for CS is surgical epicardial pacemaker lead removal. In the current case, the patient received a beta-blocker, angiotensin-converting enzyme inhibitor, mineralocorticoid receptor antagonist, loop diuretic, and vitamin K antagonist immediately after surgery. She did not receive aspirin or non-steroidal anti-inflammatory drugs because there was no evidence of pericarditis. She has since stopped all of the medication due to pregnancy. In a report on 10 CS cases, surgical EP

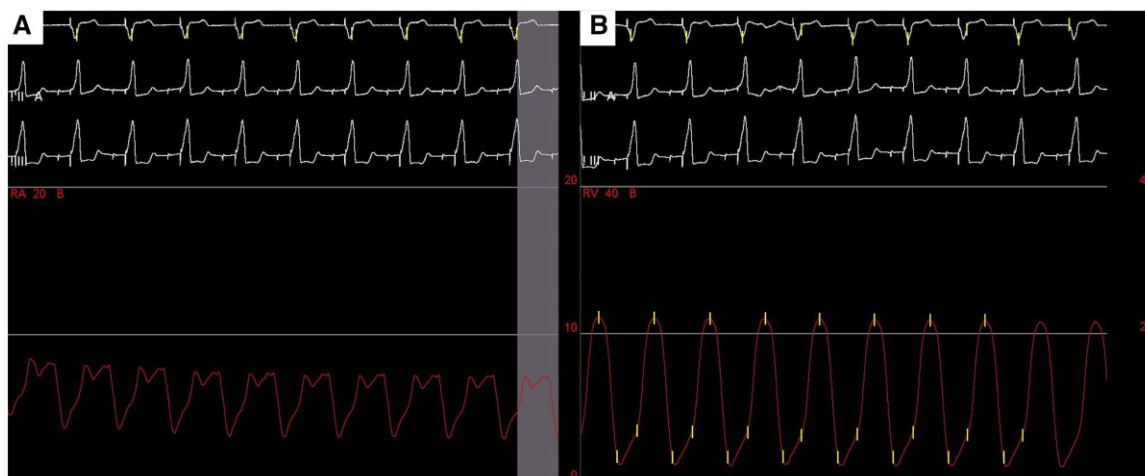


Figure 5 Haemodynamic tracings after epicardial pacemaker lead removal. Right heart catheterization after the surgery showing a persistent steep y descent waveform with a pressure of 6 mmHg in the right atrium (A) and disappearance of the 'dip and plateau' waveform in the right ventricular pressure (B). RA, right atrium; RV, right ventricle.

lead removal was performed in 9 cases, except for a case of sudden cardiac death, and 7 of them had a good outcome.² The above report recommends performing surgical EP lead removal as soon as possible after the diagnosis of CS. In the present case, surgical EP lead removal was performed as described in previous reports.

Although the optimal method of implanting EP to prevent CS has not yet been established, it has been suggested that lead loops should not be placed on the anterior surface of the pericardium, surrounding the heart, or crossing the main coronary arteries.^{2,8,9}

Despite its rarity, CS can cause critical symptoms and complications. Cardiac strangulation may be diagnosed in the late stage of EP implantation, as in this case. Accordingly, patients with EP implantation should be carefully monitored for the development of CS, even in adulthood. Moreover, surgical EP lead removal should be performed without hesitation in cases where CS is considered the primary aetiology of critical symptoms or complications, because surgical removal is the fundamental treatment for CS.

Lead author biography



Dr Ryeonshi Kang was graduated from The Jikei University School of Medicine and received PhD in 2021. He belongs to a cardiology heart team performing a variety of TAVI and PCI operations and managing, on a daily basis, patients in the coronary care unit.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent has been obtained from the patient for the submission and publication of this report including images, in line with the COPE guidelines.

Funding: None declared.

Conflict of interest: None declared.

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