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CASE REPORT
Heart failure

A case report: upgrade to cardiac resynchronization therapy with a blocked persistent left-sided superior vena cava

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Background Pacemaker-induced cardiomyopathy (PICM) can occur in up to 9% of patients having a pacemaker. Pacemaker-induced cardiomyopathy can be treated by upgrade to a biventricular pacemaker with a left ventricular (LV) lead implantation. The procedure can be technically challenging in patients with persistent left-sided superior vena cava (PLSVC).

Case summary We report the case of a 72-year-old gentleman with a PLSVC, who had a dual-chamber pacemaker implanted 15 years ago for complete heart block. After 12 years of good health, the gentleman developed breathlessness due to PICM. At upgrade to biventricular pacemaker, his coronary sinus was found to be occluded and a collateral branch was used to successfully position an LV lead. Marked clinical improvement was seen before representation with syncope after 2 years due to simultaneous failure of both LV and right ventricular leads. Subsequently, a right-sided *de novo* biventricular pacemaker was implanted. In this instance, the PLSVC was beneficial because it isolated the existing leads from the new implant, thereby reducing the risk of SVC obstruction.

Discussion Although implantation of pacemaker leads through a PLSVC constitutes a challenging procedure due to manoeuvring difficulties of the pacing leads into the cardiac chambers, in this particular case, the presence of PLSVC was beneficial because it meant that no leads were present in the true SVC, reducing the risk of occlusion and avoiding the need for lead extraction.

Keywords Pacemaker • Cardiac resynchronization therapy • Left-sided superior vena cava • Pacemaker-induced cardiomyopathy • Case report

Learning points

- Persistent left-sided superior vena cava (PLSVC) affects about 0.5% of the general population. Persistent left-sided superior vena cava is often asymptomatic but can be associated with other cardiac anomalies, increased risk of arrhythmias or right to left shunt.
- Persistent left-sided superior vena cava causes challenges in implantation of permanent pacemaker leads. During left ventricular (LV) lead placement, it is difficult to image the coronary sinus (use subselection catheter) and difficult to leave sufficient LV lead slack (use active fixation lead).
- Extraction of left-sided cardiac leads in the presence of PLSVC is high-risk procedure because of the angulation of the right ventricular lead.

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Introduction

Pacemakers can cause undesirable complications during and after implantation. Challenges faced with the implantation procedure are not unusual and may include anatomical variations, such as a persistent left-sided superior vena cava (PLSVC) which can be present in ~0.5% of the general population.[1]

Though uncommon, pacemaker problems can occur long after the implantation procedure. Examples of these late complications include pacemaker-induced cardiomyopathy (PICM) which affects up to 9% of patients, vein occlusion and lead failure. We report a case in which all the above common issues combine to create a challenging scenario in which conflicting clinical prerogatives needed to be balanced and the lesson learnt through a series of pacemaker procedures over a 15 year period.

Timeline

November 2003	Age 58	Dual-chamber pacemaker implanted through his persistent left-sided superior vena cava prior to atrioventricular node ablation. Transthoracic echocardiogram showed left ventricular ejection fraction (LVEF) 60–65%.
October 2015	Age 71	Referred back due to symptoms of heart failure, transthoracic echocardiogram showed LVEF 15–20%. A 100% paced with QRS duration of 238 ms.
January 2016	Age 71	Upgraded to biventricular pacemaker. During the procedure, the coronary sinus was found to be occluded. Serendipitously a collateral vein arising from the subclavian vein was seen to pass close to the heart. A quadripolar left ventricular (LV) lead was passed to a posterolateral position and cardiac resynchronization therapy was achieved.
April 2016 - June 2017	Age 71	Follow-up clinic assessment and repeat transthoracic echocardiography showed improvement of symptoms from New York Heart Association (NYHA) III to NYHA II and improvement in LVEF 35–40%.
January 2018	Age 72	Admitted with syncope, pacemaker interrogation showed failure of both LV and right ventricular leads with long pauses. Successful implant of right-sided CRT-P with good symptomatic benefit.
April 2019	Age 73	No symptoms of breathlessness. Further improvement in LVEF 45–50%.

Case presentation

In 2003, our then 58-year-old gentleman had a dual-chamber pacemaker and atrioventricular node ablation for ongoing highly symptomatic paroxysmal atrial fibrillation despite two ablations. During the implant, he was found to have a PLSVC (*Figure 1*). Other past medical history included: body mass index 32 kg/m^2, hypertension, and a gastrointestinal bleed. After 12 years of good health, the gentleman represented age 71 years with progressive exertional dyspnoea and peripheral oedema. A transthoracic echocardiogram demonstrated left ventricular ejection fraction (LVEF) <30% with moderate secondary mitral regurgitation. He was diagnosed with PICM in view of 100% right ventricular (RV) pacing with a QRS duration of 238 ms. After commencing appropriate heart failure therapy (daily: bisoprolol 5 mg, losartan 100 mg, spironolactone 25 mg, and furosemide 20 mg), the gentleman consented for upgrade to cardiac resynchronization therapy pacemaker (CRT-P). A CRT-P was favoured over cardiac resynchronization therapy defibrillator (CRT-D) in view of the anticipated difficulties and risk from either a new right-sided system or a new left-sided implantable cardioverter-defibrillator lead and the potential for the cardiomyopathy to improve with resynchronization.

During the CRT-P upgrade procedure under local anaesthetic, a venogram was performed but it was too lateral to identify occlusion of the PLSVC at the junction with the coronary sinus (*Figure 2*) (Supplementary material online, *Video S1*). Serendipitously, a small collateral branch was seen originating 5 cm above the occlusion and passing to the posterolateral aspect of the heart, offering a potential route for the left ventricular (LV) lead (Supplementary material online, *Video S2*). The options were to implant: a right-sided LV lead and tunnel to the left-sided device, a full new right-sided system or to use the collateral vein.

The collateral vein was accessed using a St. Jude CPS 115 slittable outer guide catheter and a St. Jude 90° CPS Aim SL inner catheter with a St. Jude Courier medium wire. Despite causing a small dissection a St. Jude Medical Quartet 1458Q LV lead was easily passed to an excellent posterolateral position but it could not be wedged (Supplementary material online, *Video S3*). The quadripolar LV lead was selected to give flexibility in pacing parameters given the use of a collateral vessel and anticipated proximity to the phrenic nerve (*Figure 3*). Initial parameters were good with a threshold of 1.6 V at 0.5 ms but during stability testing the threshold varied and was 2.5 V at 0.5 ms. In hindsight, an active fixation lead such the Attain Stability Bipolar MRI 4796 lead [the Medtronic Attain Stability Quad MRI SureScan (Model 4798) was not available at the time of implant] would have been preferable but in view of the dissection the lead was not changed (Supplementary material online, *Video S4*). Patient's symptoms started to improve after 3 months, and over the next 18 months following the CRT-P upgrade, the patient was able to walk up hill without breathlessness, lost weight (body mass index 26 kg/m^2) and had better energy levels. On serial transthoracic echocardiography, the LVEF improved from 15–20% to 45–50%. A particular difficulty in LV lead placement with a PLSVC is the inability to leave slack, exacerbated in this case by the LV lead not being wedged. This

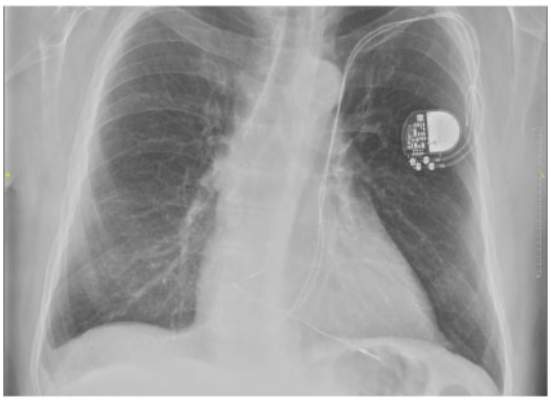

Figure 1 Dual-chamber pacemaker *in situ* through persistent left superior vena cava.

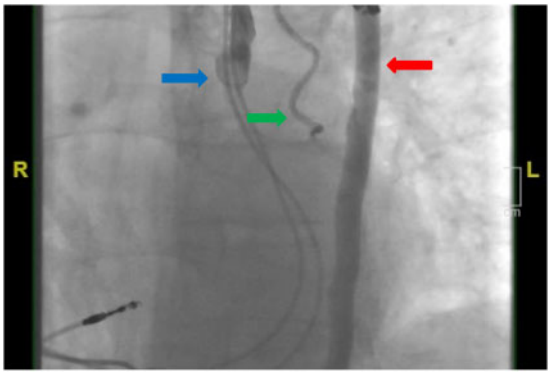

Figure 2 Venography demonstrating occlusion of a left-sided superior vena cava 13 years after dual-chamber pacemaker implantation (blue arrow). A prominent azygos vein is seen (red arrow), and a collateral vessel draining from the left ventricle which was used to upgrade to biventricular pacing (green arrow).

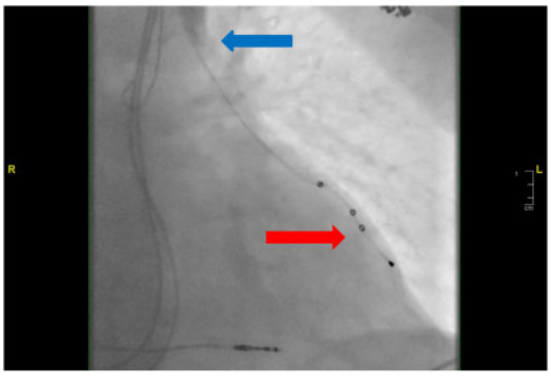

Figure 3 Fluoroscopy during PPM upgrade to cardiac resynchronization therapy pacemaker showing the Quadripolar left ventricular lead passing through a branch origination outside the heart but tracking to the posterolateral cardiac wall (red arrow). Coronary sinus occlusion can also be seen in the top (blue arrow).

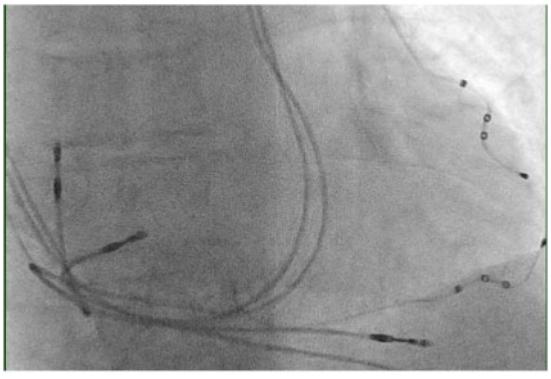

Figure 4 Fluoroscopy during biventricular pacemaker re-implantation via the right subclavian venous route, showing new and existing leads.

meant that the LV lead was not stable on inspiration and the threshold had to be set to a high level. Predicted device longevity was 4.7 years; this was felt to be acceptable given the complexities of the case. In January 2018, now aged 72 years, the patient was hospitalized due to syncope. He was found to be in intermittent complete heart block and device interrogation found that intermittently both LV and RV leads were simultaneously failing to capture. A decision was made to implant a right-sided CRT-P; the presence of the PLSVC was beneficial because it meant that no leads were present in the true superior vena cava (SVC), reducing the risk of occlusion and avoiding the need for extraction. The right-sided LV lead can be seen close to the left side LV lead though in a completely separate venous system (*Figure 4*; Supplementary material online, *Videos S5–S7*). The left-sided pulse generator and 2015 LV lead were removed, and the left RA and RV leads were capped and buried (*Figure 5*). Device checks 1-month post-procedure were satisfactory. At regular ongoing follow-up, the gentleman continues to be symptomatically well.

Discussion

A PLSVC is the most common variant of systemic venous drainage. It is present in ~0.5% of the general population and 1.5–10% of patients with congenital heart disease.[1] Failure of obliteration in utero of the left common cardinal vein means that the left subclavian and jugular veins drain into the right atrium via the coronary sinus (CS). Typically, it is discovered incidentally.[2] About 40% of cases are associated with atrial septal defect, bicuspid aortic valve, coarctation of aorta, coronary sinus ostial atresia,[3,4] and an unroofed coronary sinus which can rarely lead to right to left shunting.[5,6] In this case, transthoracic echocardiogram and chest CT scan excluded these features. Persistent left-sided SVC conveys an increased risk of atrial fibrillation.[7] A PLSVC makes standard pacemaker implantation challenging; the lead enters the RA through the CS and the acute angle between the CS ostium and tricuspid valve necessitates a 360° loop to enter the

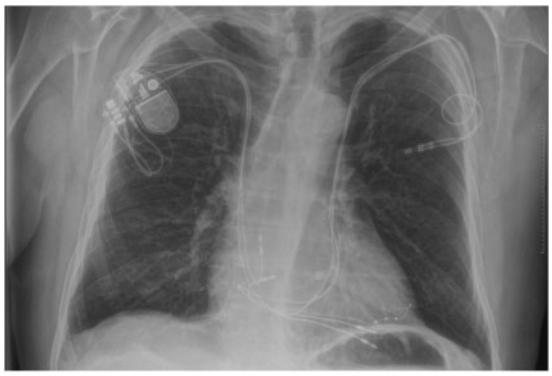

Figure 5 Chest radiograph showing the final lead configurations. Following the new right-sided cardiac resynchronization therapy pacemaker implantation, the old device in the left side along with its associated left ventricular lead was removed, and the right atrium and right ventricular leads were capped and buried.

RV.[2] There are additional complexities in implanting an LV lead, a situation that is not commonly faced. Firstly, it is challenging to image the coronary sinus because it cannot usually be occluded with a balloon due to its size. Our approach is to use 90° subselection catheter to blindly cannulate a branch; injecting contrast then frequently backfills other branches. Secondly, there is no reservoir to position lead slack when approaching from the left side because the lead does not pass through the right atrium. The LV lead is therefore at considerable risk of displacement with deep inspiration. A solution to this problem is to use an active fixation LV lead.

Pacemaker-induced cardiomyopathy is defined as a reduction in LVEF of >10% after pacemaker placement where paced beats comprise >20% of the total QRS complexes. Other causes of a decreased LVEF should be considered including ischaemia, valvular disease, and atrial arrhythmias before diagnosing PICM.[7,8] Pacemaker-induced cardiomyopathy is present in up to 9% of patients and is most prevalent within the first year after implantation.[9] Risk factors for PICM include male gender, frequent/sustained RV pacing, paced QRS duration ≥150 ms, and wider native QRS duration (particularly >115 ms).[7,8]

Left ventricular dyssynchrony caused by sustained RV pacing causes progressive molecular changes due to a decrease in systolic function, an increase in end-systolic volume and wall stress, and delayed relaxation. PICM is treated by upgrade to a biventricular pacemaker with an LV lead; stimulation of both RV and LV can reduce dyssynchrony and has been shown to increase exercise tolerance and reduce heart failure hospitalization.[10] The duration of the PICM is inversely associated with response to biventricular pacemaker upgrade. Occlusion of a PLSVC is likely to be a rare occurrence in view of it usually being a large vessel. The situation here is seen more frequently when upgrading a pacemaker with standard anatomy and an occluded subclavian vein. This case report demonstrates the potential to use collateral vessels that proximate the heart and the pitfalls of doing so. In the presence of anatomical variations, innovative strategies may be required during complex pacing such as highlighted in this case.

In this gentleman, while the LV lead was always considered to be at risk of failure due to instability, the RV lead failure came without warning with no significant change in parameters before his presentation with lead failure. Consideration was given to extraction of the old RA and RV leads, given that it is undesirable to have six leads in the heart due to risk of venous occlusion, future infection, and lead interaction. However, due to the angulation of the RV lead and 15-year duration of implant, extraction was considered to be extremely high risk. In addition, the PLSVC isolated the left-sided system from the right such that the true SVC would have three leads.

Conclusion

Patients with PLSVC represent a particularly challenging cohort for pacing due to the angulation between the coronary sinus os and the tricuspid valve. Left ventricular lead implant is made difficult in these cases due to the difficulties imaging coronary sinus branches and the lack of a natural place to store the slack needed for the lead to move with respiration and postural change. Simple solutions to these issues are available, particularly use of an LV lead with fixation.

Lead author biography

Kawan Fadhil Abdalwahid is a Kurdish-born medical doctor and graduate of Sulaimani school of Medicine. He completed his foundation and core training in Iraqi Kurdistan. He was also a health associate at the United Nations High Commissioner for Refugees (UNHCR) in 2016–17. Currently, he is a cardiology specialist registrar at Glenfield Hospital in Leicester, UK. Certificates: MBChB, MRCP (UK), DipUKMP.

Supplementary material

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

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

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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