Successful implantation of a leadless pacemaker in a patient with complete atrioventricular block and congenital absence of superior vena cava: a case report

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Background
Congenital absence of superior vena cava (CASVC) is an extremely rare vascular anomaly often associated with conduction disturbances which makes implantation of a pacemaker difficult. We report a case of pacemaker implantation in a patient presenting with complete atrioventricular block (c-AVB) with bilateral absence of the SVC.

Case summary
A 68-year-old man who had experienced dyspnoea on exertion by c-AVB was admitted to our hospital for treatment and management. Permanent pacemaker insertion was initially planned; however, an endocardial pacemaker lead could not be implanted in the right atrium. Computed tomography scan with contrast revealed that the venous blood from the upper half of the body flowed into the inferior vena cava via the azygos vein. Due to the difficulty of inserting an endocardial lead from the subclavian vein, a leadless pacemaker (LP) was implanted instead via the femoral vein.

Discussion
This is the first case of an LP implantation in a patient presenting with c-AVB with CASVC. Confirmation of blood vessel anatomy to rule out CASVC is necessary prior to pacemaker implantation when abnormal venous anatomy is suspected.

Keywords
Case report • Congenital absence of superior vena cava • Systemic venous anomalies • Persistent left superior vena cava • Complete atrioventricular block • Leadless pacemaker

Learning points
• Congenital absence of superior vena cava (CASVC) is a rare vascular anomaly that presents a challenge during pacemaker implantation.
• Leadless pacemaker is a potential alternative to transvenous pacing system and should be considered when confronted with challenging vascular access such as CASVC.
Introduction

Congenital absence of superior vena cava (CASVC) is a very rare vascular anomaly. CASVC is usually asymptomatic, so many patients remain undiagnosed and live a normal quality of life. However, this condition may be associated with congenital cardiac anomalies and/or conduction disturbances.

Symptomatic atrioventricular block (AVB) is commonly treated using a permanent pacemaker with endocardial lead implantation via the subclavian vein. In patients with CASVC, endocardial lead implantation can be difficult. Instead, epicardial lead implantation via thoracostomy is commonly utilized despite its high invasiveness. Although leadless pacemakers (LPs) provide only ventricular pacing, a patient with CASVC may benefit from them as they can be less invasively implanted via a transfemoral vein approach. To the best of our knowledge, this is the first reported case of LP implantation in a patient with CASVC who presented with a complete AVB (c-AVB).

Timeline

| Time                        | Events                                                                 |
|-----------------------------|------------------------------------------------------------------------|
| 1 month before hospitalization | Patient had been aware of dyspnoea on exertion                         |
| Day 1                       | Patient was transported to our hospital due to dyspnoea on exertion. An electrocardiogram revealed complete atrioventricular block. |
| Day 3                       | Insertion of a permanent pacemaker was performed. However, we could not place an endocardial electrode lead. Instead, a temporary stimulation electrode was inserted through the femoral vein. Contrast-enhanced computed tomography revealed that the superior vena cava was absent. |
| Day 9                       | A leadless pacemaker (LP) was implanted.                               |
| Day 11                      | The patient had no subjective complaints, and no complications were noted. The patient was discharged from our hospital. |
| 6 months after discharge    | The patient has been asymptomatic since LP implantation. Left ventricular ejection fraction of 54%, brain natriuretic peptide level of 66.7 pg/mL, and percentage of right ventricular pacing of 99.9%. |

Case presentation

A 68-year-old man with an unremarkable medical history had experienced dyspnoea on exertion for 1 month. After c-AVB was found on an electrocardiogram in the outpatient clinic, he was immediately transferred by ambulance to our hospital. The vital signs revealed a blood pressure of 157/83 mmHg and a heart rate of 44 b.p.m. The electrocardiogram showed a sinus rhythm at a rate of 105 b.p.m. with c-AVB and an escape rhythm of 40 b.p.m. in the left bundle branch block configuration (Figure 1). The chest X-ray did not reveal pleural effusion, pulmonary congestion, or cardiac enlargement. The echocardiographic findings did not show any structural abnormality, and the left ventricular function was preserved. Thus, the patient was considered to be a good candidate for a DDD pacemaker, and the decision for permanent pacemaker implantation was made.

The preoperative left subclavian vein angiography showed the persistent left superior vena cava (PLSVC) and the left brachiocephalic vein, which appeared to drain into the right atrium via the superior vena cava (SVC) (Video 1). Via the left subclavian vein approach, pacemaker implantation was performed. However, a guidewire and electrode lead could not be guided into the right ventricle although they seemed to take the brachiocephalic SVC route (Figure 2). The vein angiography was repeated, this time from the level above the right atrium, but the contrast medium did not drain directly into the right atrium (Video 2). Although an attempt to insert the electrode lead through the PLSVC was made, the tortuous nature of the blood vessel prevented the insertion of the electrode lead (Figure 2). Instead, a temporary stimulation electrode was inserted by puncturing the femoral vein. Reconsidering the procedure, contrast-enhanced computed tomography (CT) scan was performed, which revealed the absence of the SVC and showed that the bilateral brachiocephalic veins drained into the inferior vena cava by way of the azygos vein (Figure 3). Furthermore, the PLSVC drained into both the right and left atria. Additional transthoracic and oesophageal echocardiography was performed and did not show any other congenital vascular or coronary defects.

We concluded that the transvenous endocardial pacemaker lead implantation was not feasible. Thus, we decided that the LP implantation was the best option for the patient due to its less invasive nature, and we successfully implanted the LP via a femoral vein approach (Figure 4).

The patient has been asymptomatic and after 6 months from implantation, he has never experienced heart failure or other complications such as infection and pacemaker dislodgement. Although the right ventricular (RV) pacing was 99.9%, the patient’s left ventricular ejection fraction was preserved at 54% and the brain natriuretic peptide level remained low (66.7 pg/mL).
An electrocardiogram reveals a complete atrioventricular block upon arrival of the patient to the hospital.

Preoperative left subclavian vein angiography revealed the persistence of left superior vena cava and the left brachiocephalic vein, which appeared to drain into the right atrium via the superior vena cava.

The guidewire (arrow) could not be guided into the right ventricle although it seemed to take the brachiocephalic superior vena cava route. Selective persistent left superior vena cava vein angiography revealed a tortuous route, and persistent left superior vena cava (arrowhead with dotted line) drained into both the right and left atria.
Discussion

We report a case of LP implantation in a patient with CASVC who presented with c-AVB. To the best of our knowledge, this is the first case of its kind to be reported. In congenital heart disease, systemic venous anomalies are common variants. However, CASVC is a rare congenital disorder, and it only accounts for 0.1% of all congenital cardiovascular malformations along with the presence of PLSVC. When the bilateral SVC is absent, a more severe and extremely rare form of CASVC occurs. Patients with this condition are usually asymptomatic, but it is often associated with other cardiac anomalies. Moreover, most congenital heart diseases are also known to be associated with conduction abnormalities. One report suggested that rhythm abnormalities associated with congenital absence of the right SVC have been reported to be as high as 36% and occurs in 70% of patients with congenital absence of the right SVC over 60 years old. However, whether CASVC is correlated with conduction abnormality remains unclear. In this case, although CASVC was only diagnosed when the patient developed c-AVB, a 68-year-old man may have the possibility of developing degenerative conduction abnormalities, and it was difficult to predict any vascular anomaly. Before the electrode lead implantation, subclavian vein angiography was performed, but it was difficult to distinguish whether the SVC was absent using this technique. Consequently, this procedure made it seem that the left subclavian vein drained into the right atrium via the SVC. Thus, the CT examination should be performed when abnormal venous anatomy is suspected before attempting transvenous device implantation.

Pacemaker implantation in patients with AVB with vascular anomalies has not been thoroughly documented. However, two cases of pacemaker implantation in patients with CASVC have been reported. One case was implanted with an epicardial lead, while the other case was implanted by venotomy of the epigastric vein. Additionally, one report utilized a transthoracic transtrial approach using a parasternal extrapleural route through the mediastinum to directly puncture the right atrium, which is a useful technique in patients with upper vein obstruction.

LP is not the first choice for AVB because treatment of AVB requires atrioventricular (AV)-synchronous pacing. Although...
different techniques to implant a pacemaker system in patients with CASVC have been described, the majority of them are highly invasive. According to the MICRA™ (Medtronic, USA) Transcatheter Pacing Study, the leadless device was successfully placed 99% of the time. Besides, 96% of patients who received an LP had no major device-related complications, and major complications were 48% lower compared to patients who received a transvenous pacemaker. Thus, the decision of implanting an LP via a minimally invasive technique was considered. However, one must keep in mind that several problems should be considered before the procedure. RV pacing can induce left ventricular dyssynchrony and systolic dysfunction as well as contributing to tricuspid regurgitation. Furthermore, the risk of developing a pacemaker syndrome in patients in sinus rhythm receiving AV-asynchronous pacing should not be overlooked. As a result of this, patients can become symptomatic due to the poorly timed atrial kick and the reduction of effective diastolic filling and stroke volumes. Although AV-synchronous LPs have been marketed, these are unfortunately not yet available for clinical practice in Japan. Thus, a VVI LP was implanted in this patient. AV-synchronous LPs should be considered in similar cases in patients in sinus rhythm when available.

Conclusion

We reported an extremely rare case in which c-AVB was reported in a patient with CASVC and was successfully managed by implantation of an LP with a favourable outcome.

Figure 4 Leadless pacemaker was successfully implanted in the right ventricle. (A) Left anterior oblique 0° and caudal 0° view. (B) Left anterior oblique 45° and caudal 0° view.

Lead author biography

Taiyo Kawaguchi received his MD degree from Yamanashi University, Yamanashi, Japan in 2019. Currently, he is a resident at Juntendo University Shizuoka Hospital, Shizuoka, Japan. He gained experience in research while in medical school, serving as an assistant at a neurophysiologic research project. He used two-photon microscopy, optogenetics, and electrophysiology to implement real-time monitoring of neural activities in the cerebellum.

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: Written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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