



Case Report

Transvenous lead extraction in a patient with polysplenia and inferior vena cava defect



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ARTICLE INFO

Article history:

Received 17 July 2020

Received in revised form 26 August 2020

Accepted 30 August 2020

Keywords:

Lead extraction

Polysplenia

Congenital heart disease

Pacemaker

Inferior vena cava defect

ABSTRACT

A 28-year-old woman with polysplenia was referred to our hospital for atrial lead failure. She had undergone an intracardiac repair (ICR) for incomplete atrioventricular septal defect and the implantation of epicardial pacing leads due to complete atrioventricular block at the age of 1 year. When she was 13 years old, an endocardial dual-chamber pacemaker was implanted via the right subclavian vein because of epicardial lead failure. The contrast-enhanced computed tomography scan revealed an inferior vena cava defect with an azygos vein connection to the superior vena cava, occlusion of the right brachiocephalic vein, a defect of the left brachiocephalic vein, and a persistent left superior vena cava ligated at the ICR. Therefore, lead exchange was indicated. During the operation, the temporary pacing lead and the guidewire for emergent deployment of the Bridge Occlusion Balloon[®] were advanced through the azygos vein and placed at the right ventricle and the hepatic vein, respectively. Both 11-Fr and 13-Fr mechanical rotational dilator sheaths were needed for the lead extraction owing to dense calcification and tight adhesions. The atrial lead was successfully extracted without any complications despite extremely restricted venous access. A new atrial lead was inserted through the space created by the 13-Fr sheath.

<Learning objective: Transvenous lead extraction in patients with polysplenia is technically challenging. These patients often undergo pacemaker implantation in childhood, which results in tight adhesions and dense calcifications on the leads, and venous access is extremely restricted. It may be impossible to use a snare and deploy the endovascular balloon to prevent a catastrophic complication from the right femoral vein to the superior vena cava in cases of the inferior vena cava defect.>

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Introduction

Transvenous lead extraction (TLE) for patients with a structurally normal heart is an established procedure, and its safety and efficacy have been previously reported [1]. In contrast, TLE in adult patients with congenital heart disease is challenging. There are many anatomical variations of the heart and vessels in such patients. The number of adult patients with congenital heart

disease who require lead extraction is increasing for the following reasons [2]. First, more patients with congenital heart disease are surviving into adulthood. Additionally, some patients with congenital heart disease are requiring pacemaker implantation at a younger age because of congenital or acquired bradycardia, which results in a higher rate of lead failure compared to those with structurally normal hearts [3,4].

Polysplenia, known as left isomerism, is a rare congenital subtype of heterotaxy syndrome associated with heart and organ malformations and with inferior vena cava (IVC) defect and azygos or hemi-azygos connection [5]. Herein, we report the rare case of a successful TLE in a patient with polysplenia.

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Case report

A 28-year-old woman with polysplenia was referred to our hospital for endocardial atrial lead failure. At the age of 1 year, she had undergone intracardiac repair (ICR) for incomplete atrioventricular septal defect, ligation of a persistent left superior vena cava (SVC), and implantation of an epicardial pacemaker due to complete atrioventricular block. The epicardial pacemaker was switched to an endocardial DDD pacemaker due to epicardial pacing lead failure at the age of 13 years. Transvenous pacing leads were implanted via the right subclavian vein. The transvenous pacemaker system was composed of a Reply pulse generator (Sorin, Mirandola, Italy), a 45-cm Capsurefix (model 5068) active fixation lead (Medtronic, Minneapolis, MN, USA) at the right atrial appendage, and a 52-cm Capsurefix (model 5068) at the right ventricle.

Fifteen years after transvenous pacemaker implantation, atrial lead failure such as noises, high impedance, and high pacing threshold occurred. The contrast-enhanced computed tomography scan revealed an IVC defect with an azygos vein connection to the SVC, occlusion of the right brachiocephalic vein, a defect of the left brachiocephalic vein, and a persistent left SVC ligated at the ICR site (Fig. 1). Venography also revealed total occlusion of the right brachiocephalic vein (Fig. 2A). Because there was no venous access for a new atrial lead, TLE was indicated. A temporary pacing lead was inserted into the right ventricle through the azygos vein connection prior to lead extraction (Fig. 2B). We attempted to extract only the atrial lead since the ventricular lead was functioning normally. An 11-Fr mechanical rotational dilator sheath (Evolution®, Cook Medical, Bloomington, IN, USA) was advanced from the entry site (Fig. 2C) and was switched to a 13-Fr Evolution® because of dense calcifications and tight adhesions. An outer sheath (SteadySheath®, Cook Medical) was also utilized to protect a collateral lead and tissue (Fig. 2D). Finally, the atrial lead was successfully extracted without damage to the right ventricular lead or any other complications, and a new atrial lead was inserted

through the space created by the 13-Fr sheath. A calcified shell, dense calcifications, and fibrotic tissues were seen on the extracted lead (Fig. 3).

Discussion

Polysplenia (left atrial isomerism) is usually accompanied by a complex cardiac anomaly, vascular anomalies, and bradycardia arrhythmias such as sick sinus syndrome or atrioventricular conduction disturbance. Therefore, patients with polysplenia often require pacemaker implantation at a young age, and they are likely to encounter lead-related troubles during a long implantation period. In some cases, lead extraction is required due to cardiac implantable electronic device infection or lead failure. Some previous reports have shown the safety and efficacy of lead extraction in adult congenital heart disease (ACHD) patients [6,7], but these reports have included a heterogeneous population with various congenital heart diseases, and lead extraction in polysplenia was extremely rare.

This case presented endocardial lead failure, total occlusion of the right brachiocephalic vein caused by transvenous leads implanted 15 years previously, a defect of the left brachiocephalic vein, and the ligation of the left SVC from the past ICR operation, which suggested a class II a indication for TLE according to the current guidelines in Japan and the guidelines of the Heart Rhythm Society [8]. However, TLE in patients with polysplenia and IVC defect is technically challenging for the following reasons. First, patients with complex congenital heart diseases usually undergo several generator exchanges and lead revisions, and they often retain abandoned leads [7]. In addition, leads implanted in childhood tend to have tight adhesions with calcification, as in the present case. Second, it is difficult to use a snaring technique by the femoral approach for extracting leads, due to the IVC defect with an azygos connection to the SVC. According to previous reports, femoral snares have been utilized in approximately 10% of ACHD patients who have undergone TLE, which was significantly

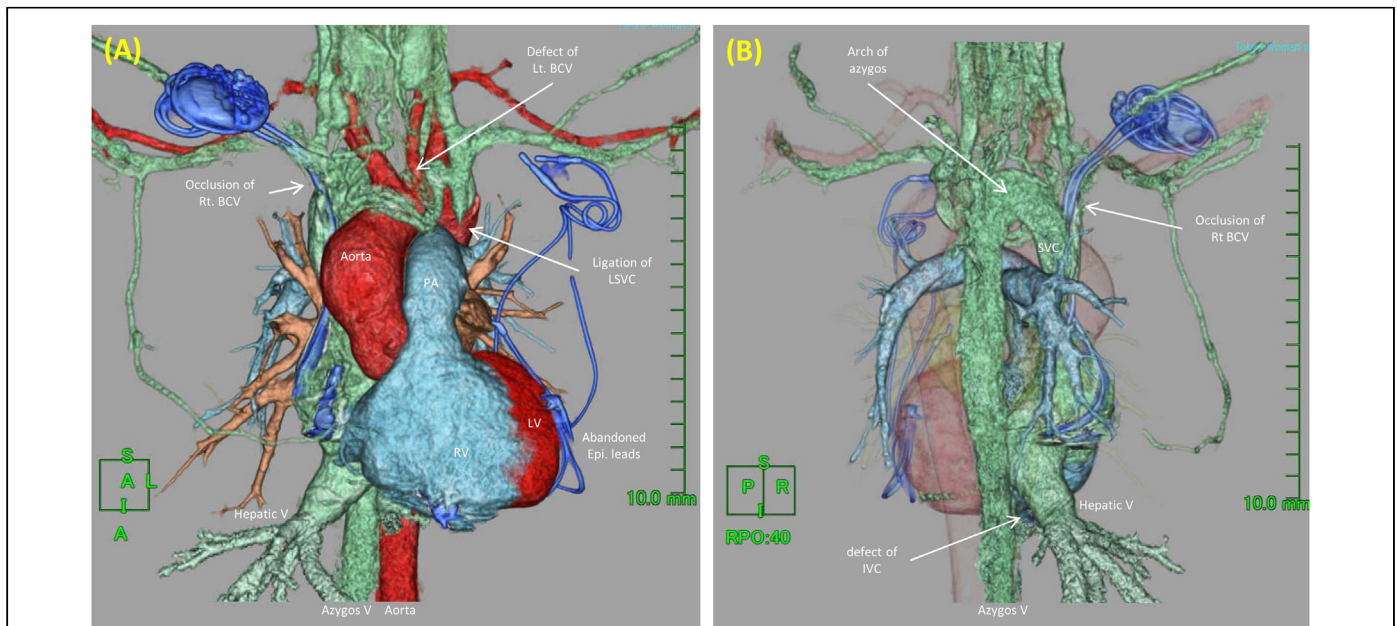
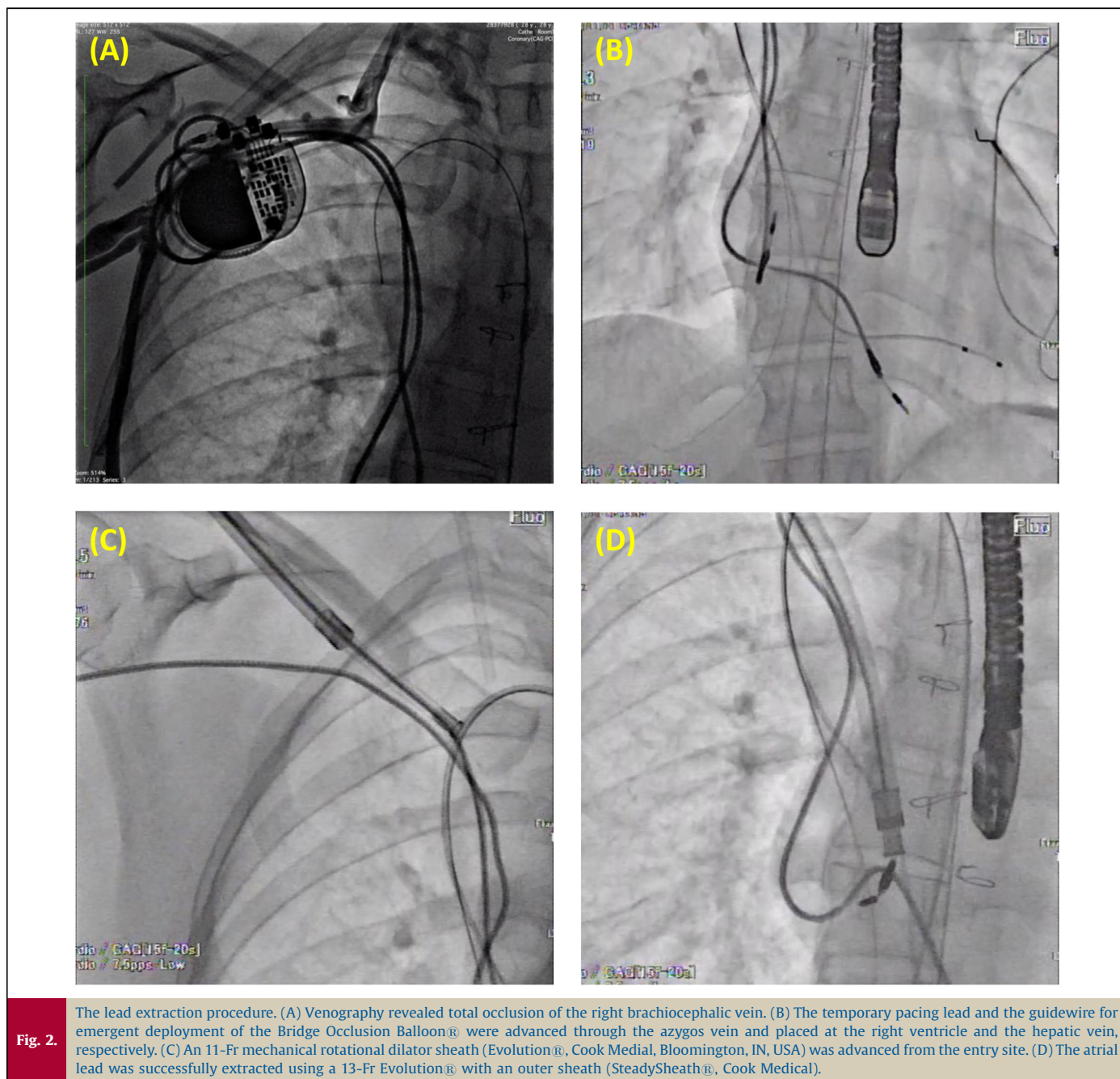


Fig. 1. Contrast-enhanced computed tomography imaging. Computed tomography showed an inferior vena cava defect, azygos vein connection to the right superior vena cava, defect of the left brachiocephalic vein, occlusion of the right brachiocephalic vein, and disconnection of the left superior vena cava. (A) Anterior-posterior view. (B) Right-sided lateral view. BCV, brachiocephalic vein; Epi, epicardial; IVC, inferior vena cava; LSVC, left superior vena cava; Lt, left; LV, left ventricle; PA, pulmonary artery; Rt, right; RV, right ventricle; V, vein.



more frequent than in patients with a structurally normal heart [2,7]. The procedure to prevent lead breakage was absolutely required for TLE in this case. Finally, although the Bridge Occlusion Balloon® (Philips-Spectranetics, Colorado Springs, CO, USA) had shown remarkable reduction of mortality in the event of catastrophic complication such as SVC tears [9], we found it difficult to deploy through the ordinary route (i.e. from the right femoral vein to the right internal jugular vein). To prepare emergent deployment of the Bridge Occlusion Balloon®, we advanced a stiff guidewire from the right femoral vein to the SVC through the azygos connection and then inserted the guidewire to the hepatic vein through the right atrium (Fig. 2B). However, the efficacy of deploying the endovascular balloon through this unusual route is not yet established. Therefore, a careful and safe

procedure to avoid damage to the right brachiocephalic vein and the right SVC should be utilized for TLE in this special case.

Another consideration for our TLE procedure was that the functioning ventricular lead should be protected. We used an outer sheath to avoid collateral damage, which resulted in the confirmation of normal parameters of the ventricular lead after the atrial lead extraction. Then, we inserted a new atrial lead through the intravascular space, which opened the occluded right brachiocephalic vein. Although 15 years had passed since the transvenous lead implantation, dense calcification and tight adhesions were observed during the procedure. Owing to the previous ICR, the present case had less risk for cardiac tamponade, but more time might have been needed for an open chest operation. A safe TLE procedure was needed because it might have



taken a longer time to recover from catastrophic complications in TLE of polysplenia than in that of a normal heart.

Other surgical approaches, such as open chest epicardial lead placement with or without lead extraction, were alternative options for our patient. However, these are extremely invasive and would require a longer hospital stay compared to endovascular approaches. Although pacemaker implantation through the femoral vein had been previously reported in patients with restricted venous access [10], our patient presented with IVC defect and an azygos vein connection to the SVC. Therefore, endocardial pacing lead implantation via the femoral vein was impossible because of the long distance between the femoral vein and the heart. After careful discussion with our team and the patient, we decided to perform TLE with on-site cardiac surgery backup and a Bridge Occlusion Balloon® under general anesthesia in a hybrid operating room.

Conclusion

TLE of polysplenia is one of the most challenging cases because of extremely restricted venous access. Careful consideration of the

indications for lead extraction is needed before the procedure. In addition, the lead extraction procedure should be carefully planned and safely implemented.

Conflict of interest

The authors declare that there are no conflicts of interest.

Declaration of Competing Interest

The authors report no declarations of interest.

Acknowledgment

None.

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