Superior vena cava obstruction and cardiovascular implantable electronic devices—a new era of leadless devices

Tomoyuki Kabutoya^

Division of Cardiovascular Medicine, Department of Medicine, and Adult Congenital Heart Disease Center, Jichi Medical University School of Medicine, Shimotsuke, Tochigi, Japan

Correspondence to: Tomoyuki Kabutoya, MD, PhD. Division of Cardiovascular Medicine, Department of Medicine, and Adult Congenital Heart Disease Center, Jichi Medical University School of Medicine, 3311-1 Yakushiji, Shimotsuke, Tochigi 329-0498, Japan. Email: kabu@jichi.ac.jp.

Abstract: Cardiovascular implantable electronic devices (CIEDs) such as pacemakers and implantable cardioverter defibrillators require the placement of a transvenous lead through the superior vena cava (SVC), which can be difficult if there is stenosis or obstruction of the SVC. Moreover, SVC syndrome may occur after the lead is inserted even if the SVC was intact before the implantation. Therefore, there is need of an appropriate strategy for handling stenosis or obstruction of SVC during lead placement. In addition, advances are being made in CIEDs that do not require transvenous leads, and thus CIEDs without a transvenous lead should be considered depending on the indications and urgency of the particular case. This manuscript is divided into (I) device therapy for patients with SVC obstruction and (II) therapeutic strategy for SVC obstruction after lead implantation. In patients with SVC syndrome, treatment of the SVC occlusion should be based on the individual pathophysiology, and depending on the indications and urgency of the case, treatment with CIEDs that do not require transvenous leads should be considered. Further data must be accumulated to clarify the long-term prognosis of device implantation after treatment of SVC occlusion. In addition, transvenous lead extraction is now widely used for device-related SVC obstruction, and this procedure also merits further accumulation of data.

Keywords: Superior vena cava syndrome (SVC syndrome); pacemaker; implantable cardioverter defibrillator (ICD); mediastinum

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Introduction

Cardiovascular implantable electronic devices (CIEDs), such as pacemakers for bradycardia and implantable cardioverter defibrillators (ICDs) for tachycardia, are usually implanted by inserting a lead from the subclavian vein into the cardiac cavity via the superior vena cava (SVC) and connecting it to a generator that is implanted above (or below or in other locations) the pectoralis major muscle. However, transvenous lead placement may be difficult if the SVC is obstructed by a tumor or for some other reason. Moreover, SVC syndrome may occur after the lead is inserted even if the SVC was intact before the implantation. This article outlines these problems.

Device therapy for patients with SVC obstruction

SVC syndrome is a syndrome in which severe stenosis or obstruction of the SVC causes impaired venous blood return from the upper body, resulting in congestion and edema of the head, face, neck, and upper body. The most common presenting symptoms include facial and neck edema, distended neck and chest veins, watering eyes,

^ ORCID: 0000-0002-5439-4303.

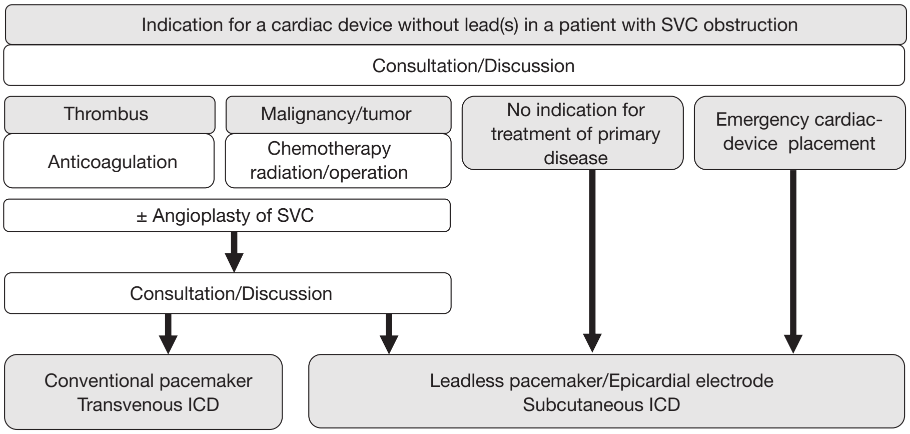

Figure 1 Device therapy for patients with SVC obstruction. SVC, superior vena cava; ICD, implantable cardioverter defibrillator.

and dizziness, particularly when leaning forward (1). The clinical presentation varies depending on the severity, location, and rapidity of the onset of the obstruction and the establishment of collateral veins. The diagnosis of SVC syndrome is based on the clinical symptoms and imaging. Imaging modalities include chest radiography, contrast-enhanced computed tomography (CT), duplex ultrasound, conventional catheter-based digital subtraction venography, and magnetic resonance venography (1). If a patient with SVC obstruction requires CIED therapy, two strategies should be considered: (I) treatment of the SVC obstruction, and (II) placement of a CIED in which the lead line does not pass through the SVC.

Avoidance of SVC obstruction

The maximum number of leads that can be implanted in a vein with an acceptably low risk of complications is a controversial topic (2). There are few data on the lead burden that results in venous access issues and SVC syndrome, and consensus documents are based on expert opinion as to the numbers of abandoned leads that justify extraction; i.e., a total of more than four leads on one side or five leads through the SVC (3). Performing a venography examination prior to the puncture is important in cases of SVC occlusion or pre-existing venous thrombosis.

Treatment of SVC obstruction

Malignancy accounts for about 70% of the cases of SVC syndrome. The most common benign causes are cardiac device therapy and catheterization, such as in patients on dialysis (1). The strategy of treatment for patients with SVC obstruction requiring device therapy is illustrated in *Figure 1*. Treatment of SVC occlusion is based on treatment of the underlying disease causing the occlusion and the mechanism of occlusion (thrombotic or non-thrombotic). If treatment of the underlying disease is difficult or the need for device therapy is urgent, a CIED that does not require a transvenous lead should be considered.

Symptomatic therapies for SVC occlusion include thrombolytic therapy in cases with a thrombotic cause of occlusion and balloon dilation, stenting, and surgical SVC angioplasty in the non-thrombotic cases. Pothineni *et al.* described four cases of angioplasty and implanted transvenous lead placement in patients with SVC syndrome (4). Although stenting for SVC syndrome had a favorable outcome in a meta-analysis (5), there is insufficient prognostic data for transvenous lead placement after angioplasty. Discussions should be held again after the SVC has been unobstructed, and the indications for and methods of CIED should be thoroughly discussed.

For patients with tumors, chemotherapy, radiation therapy, and surgery should be considered as appropriate; if there is no indication for treatment of the underlying disease causing the SVC obstruction or if urgent cardiac device therapy is needed, use of a CIED that does not require a transvenous lead will be necessary.

CIED for SVC obstruction

CIEDs can be divided into two categories: (I) pacemakers for bradycardia and (II) ICDs for defibrillation of tachycardia. Devices that do not require a transvenous lead

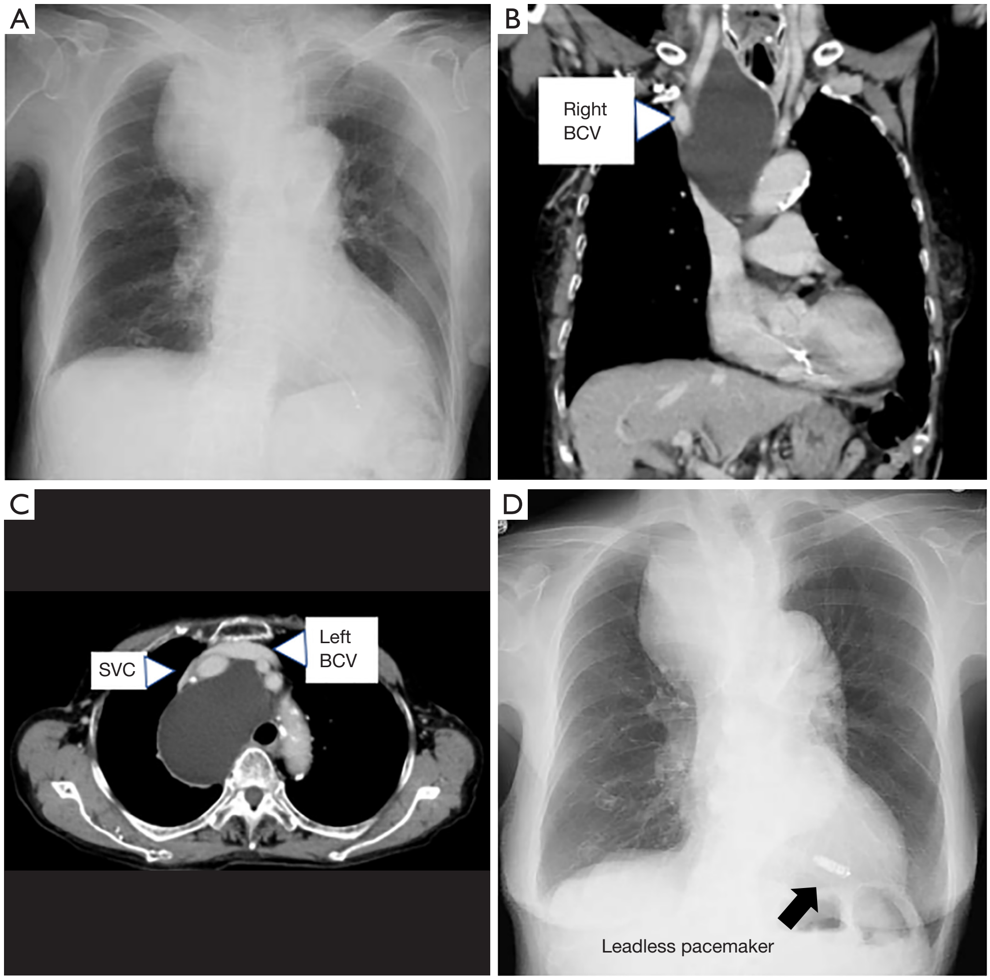

Figure 2 A case of complete atrioventricular block associated with a mediastinal tumor. (A) Chest X-ray, (B) coronal view (contrast computed tomography), and (C) axial view (contrast computed tomography); (D) chest X-ray after implantation of the leadless pacemaker. Reproduced with permission from Kabutoya *et al.* (11). BCV, brachiocephalic vein; SVC, superior vena cava.

have been developed (6). Cardiac resynchronization therapy (CRT) is also included in CIEDs with transvenous leads, but the potential issues that are associated with CRT are discussed below.

Pacemakers for bradycardia

Pacemakers that do not require a transvenous lead include open chest surgery with myocardial electrodes and leadless pacemakers.

Myocardial electrodes do not pass through the intravascular space. Ito *et al.* reported open chest biopsy and implantation of a myocardial electrode in a patient with primary cardiac lymphoma who had SVC syndrome and sinus failure (7). Maseda Uriza *et al.* (8) reported a case of SVC syndrome with a pacemaker lead in which the lead was removed, stents were placed, and a myocardial lead was implanted.

Leadless pacemakers have become widely used in recent years (9). Hirano *et al.* reported a case of complete atrioventricular block complicated with SVC syndrome due to malignant lymphoma (10). In this case, a leadless pacemaker was inserted during chemotherapy, and the atrioventricular block improved 74 days after chemotherapy. We experienced a case of complete atrioventricular block associated with a mediastinal tumor (11). The patient was very old and there was no indication for treatment of the tumor; thus, a leadless pacemaker was implanted (*Figure 2*).

ICDs for tachyarrhythmias

ICDs are implanted for the tachyarrhythmias (ventricular

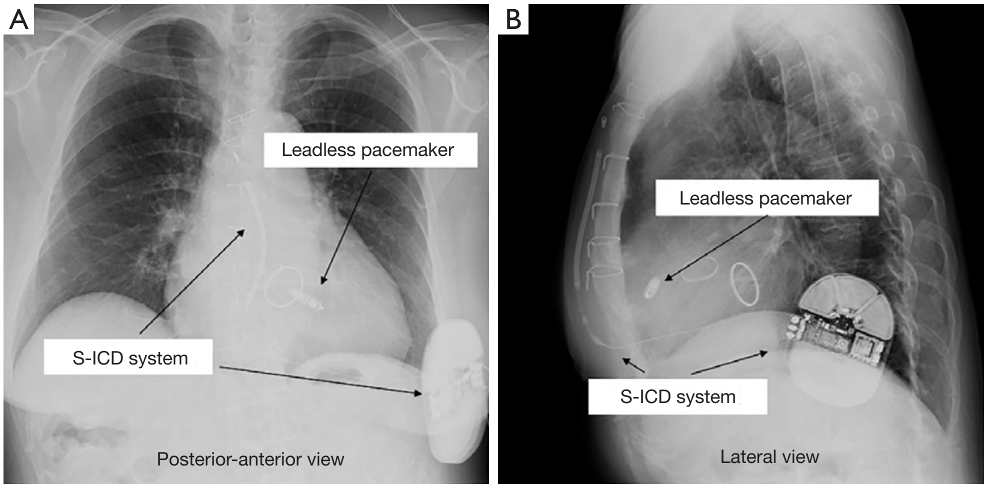

Figure 3 Posterior-anterior and lateral views of the leadless pacemaker and subcutaneous implantable cardioverter defibrillator systems. Reproduced with permission from Ito *et al.* (14). S-ICD, subcutaneous implantable cardioverter defibrillator.

tachycardia and ventricular fibrillation); ICDs can terminate tachyarrhythmias by delivering electric shock(s) between the generator and a shock lead implanted in the right ventricle. Subcutaneous implantable cardioverter defibrillator (S-ICD), in which the defibrillation lead is implanted subcutaneously rather than intravascularly, have also been developed (12), and have been shown to be as safe and effective as conventional ICDs (13). S-ICDs have the following two disadvantages: (I) antitachycardia pacing cannot be performed with an S-ICD, and (II) unlike ICDs with transvenous leads, S-ICDs do not have a pacemaker function for bradycardia except after shock. Ito *et al.* reported a case in which an S-ICD was implanted in a patient with ventricular tachycardia who had a history of mechanical valve infective endocarditis and a leadless pacemaker was placed for subsequent bradycardia (*Figure 3*) (14). In addition, attempts are being made to develop a novel modular cardiac rhythm management system consisting of a communicating antitachycardia pacing-enabled leadless pacemaker and an S-ICD (15). In the future, therefore, it should be possible to combine an S-ICD with a leadless pacemaker that has the same functionality as an ICD with transvenous leads. Problems with inappropriate therapy of the S-ICD have been reported (16), and attempts are being made to reduce the problems with these devices (17).

Management of terminally ill patients and others

In terminally ill patients, in justified cases, external endocavitary stimulation may be used. Indications for pacemaker or ICD implantation are included in the American College of Cardiology/American Heart Association guidelines for patients with indications for permanent pacing but also for those with significant comorbidities such that pacing therapy is unlikely to provide meaningful clinical benefit; if a patient's care goals strongly preclude pacemaker therapy, the implantation or replacement of a pacemaker should not be performed (18). In patients who are expected to have a shortened life span because of a terminal progressive illness, the benefits of pacing support may not be realized and are unlikely to positively impact the overall outcome (19). For patients with advanced cancer, in addition to a subcutaneous ICD, the possibility of implanting a defibrillating electrode into the azygos vein or pericardial sac retrosternally as well as a defibrillating vest might be considered an option.

Therapeutic strategy for SVC obstruction after lead implantation

The therapeutic strategy for SVC obstruction after lead implantation is depicted in *Figure 4*.

Pacemakers and ICDs account for 7.5% of all cases of thrombotic SVC syndrome, and the number of cases has been increasing over time (20). Thrombosis, mechanical stress, infection, and inflammation are possible mechanisms of lead-induced SVC occlusion (21). Riley *et al.* summarized information on the treatment of lead-related SVC syndrome (22). Medical treatment for SVC syndrome often results in recurrence (5–33%), while surgical treatment results in relatively few recurrences (11%), but the

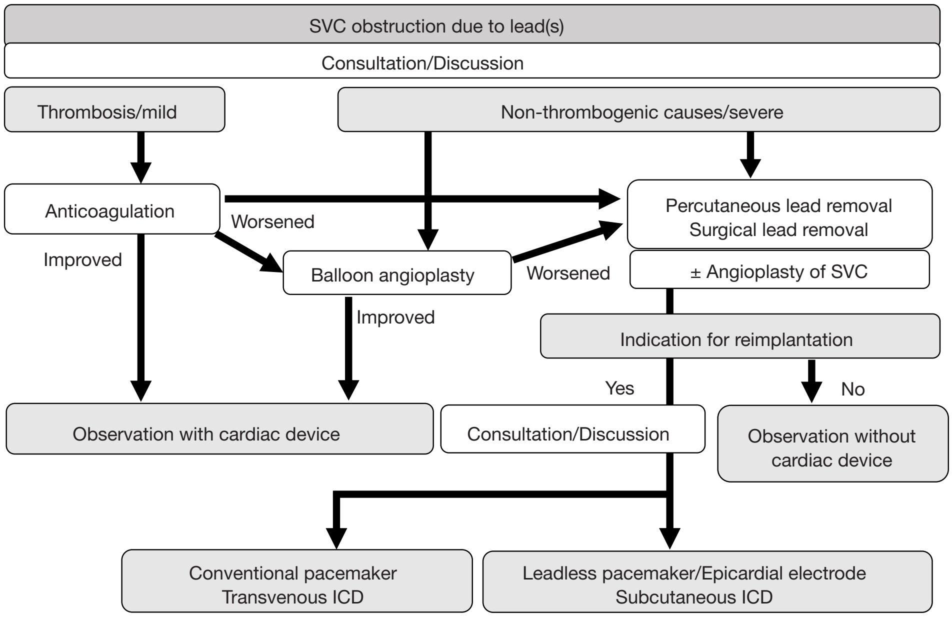

Figure 4 Therapeutic strategy for SVC occlusion after lead implantation. SVC, superior vena cava; ICD, implantable cardioverter defibrillator.

invasiveness of the procedure is problematic.

Anticoagulation

A high D-dimer level may indicate that thrombosis is a contributing factor in the mechanism of SVC occlusion, and anticoagulation may be indicated.

Mumoli *et al.* treated a patient with SVC syndrome 2 years after pacemaker implantation with a week of full dose subcutaneous enoxaparin and subsequent anticoagulation with edoxaban 60 mg, and after 3 months, the patient was free of symptoms and a chest CT angiography revealed complete resolution of the thrombosis, and the anticoagulant was interrupted (23). The thrombosis was not acute, and Mumoli *et al.* hypothesized that anticoagulation can restore a favorable balance between thrombosis persistence and physiologic fibrinolysis, leading to thrombus resolution (23). Fukui *et al.* treated a patient with SVC syndrome 2 years after pacemaker implantation and administered rivaroxaban; they observed no recurrence of thrombosis over 1 year of rivaroxaban therapy (24).

Balloon angioplasty, stenting

If symptoms are severe, catheterization is preferred for symptom improvement; Eberhardt *et al.* performed vasodilation for SVC syndrome in a patient more than 10 years after pacemaker implantation and reported no problems after 1 year of treatment with rivaroxaban 20 mg (25). Pham also reported a case of balloon dilation for SVC obstruction and insertion of a new lead in a patient with SVC occlusion after 5 years (26). Stenting without lead removal has also been performed, but can potentially cause failure of the lead (27).

Surgical angioplasty

Surgical angioplasty is invasive but has the advantage of reliably treating SVC obstruction, such as when a wire cannot pass through the obstruction. Hodges *et al.* reported a case of SVC syndrome 3 years after pacemaker implantation in which a leadless pacemaker was implanted under direct vision after surgical SVC repair (28).

Percutaneous lead removal

In recent years, percutaneous lead removal has evolved and is recommended in guidelines (2). Arora *et al.* performed lead removal in 13 of 17 patients with SVC syndrome. All patients underwent revascularization (of the remaining four, three patients were treated with venoplasty alone, and 1 patient underwent surgical SVC reconstruction), and all were confirmed to have no recurrence at 1 year (29). Gabriels *et al.* summarized results over a longer follow-up (median of 5.5 years) in 16 patients with SVC syndrome who underwent transvenous lead extractions (30). In addition to transvenous lead extraction and percutaneous treatment, 13 patients (81.3%) were managed with long-term anticoagulation therapy, but 4 patients (25%) had recurrent symptoms between 5 months and 2 years after extraction. These patients were managed either conservatively or with further balloon angioplasty and stenting. One patient required an emergent repair with a surgical reconstruction due to an SVC tear that occurred during the extraction and placement of an epicardial lead. Manual lead removal may cause venous or cardiac perforation, and it is safer to remove the lead using a device such as a laser sheath. Anticoagulation, venoplasty, and surgical interventions alone have been abandoned due to the high risk of recurrences, and thus transvenous lead extraction followed by venoplasty with or without stenting are a reasonable first-choice approach for SVC syndrome (27).

In a retrospective analysis of 3,002 venograms from patients awaiting transvenous lead extraction, Czajkowski *et al.* observed that SVC occlusion was rare (31). Their research group also described risk factors for lead-related venous obstruction and the influence of lead-related venous obstruction on the complexity and outcomes of lead extraction (32,33). Clinicians need to be familiar with these risk factors and review patients' venography findings to ensure safe lead removal.

Perspectives

Further data must be accumulated to clarify the long-term prognosis of device implantation after treatment of SVC occlusion. In addition, transvenous lead extraction is now widely used for device-related SVC obstruction, and this procedure also merits further accumulation of data. Ventricular single chamber pacing (VVI), single lead atrial synchronous ventricular pacing mode (VDD) leadless pacemakers are currently available, but dual chamber pacing (DDD) is also being developed (34). CRT for patients with heart failure that does not require a left ventricular lead is also being studied (35), but currently this strategy requires a right ventricular lead. The development of CRT without leads is eagerly anticipated.

Conclusions

In patients with SVC syndrome, treatment of the SVC occlusion should be based on the individual pathophysiology, and depending on the indications and urgency of the case, treatment with CIEDs that do not require transvenous leads should be considered.

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Footnote

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Ethical Statement: The author is accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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