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Implantation of a Dual-Chamber Pacemaker in a Patient with Situs Inversus and Dextrocardia using a Novel Ultrasound Technique

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Abstract

We report a case of a 43-year-old man with situs inversus and dextrocardia who was admitted with syncope in the setting of complete atrioventricular block. The complex anatomy poses a considerable challenge to transvenous permanent pacemaker implantation. We employed a novel technique using vascular ultrasound and agitated saline solution to assist with lead positioning. This technique could be useful in pediatric populations or younger patients, in whom the use of ionizing radiation is an important issue.

Keywords: Bubble contrast, congenital cardiac disease, dextrocardia, pacemaker implantation, vascular ultrasound

Introduction

Situs inversus (mirror viscera arrangement) is a very rare occurrence. It occurs more frequently with dextrocardia (true mirror image) with an incidence estimated as 1 in 6000–1 in 12,000 live births;[1,2] the incidence of cardiac abnormalities in these patients is 2%–5%.[3]

There are very few reports in the literature describing complete atrioventricular (AV) block in patients with situs inversus.[4,5] In the setting of complete AV block, the complex extracardiac and intracardiac anatomy, as well as the reversed fluoroscopic orientation, constitutes a particular technical challenge to transvenous permanent pacemaker implantation (PPI). We describe a case of a patient with situs inversus and dextrocardia, who was admitted with complete AV block.

Case Report

We report a case of a 43-year-old male who presented to our institution with syncope and a 2-month history of dizziness. His medical history was remarkable for situs inversus with dextrocardia and surgical closure of a membranous ventricular septal defect (VSD) 30 years previously. On physical examination, holosystolic murmur was heard on the right sternal border. Twelve-lead electrocardiogram (ECG) at admission showed complete AV block, heart rate of 30 bpm, and wide-QRS complexes (±160 ms); there were right-axis deviation, inverted P, QRS, and T waves in lead I, and absence of R-wave progression in

chest leads, suggesting dextrocardia [Figure 1a]. Transthoracic echocardiogram (TTE) showed preserved biventricular systolic function as well as AV and ventriculoarterial concordance. There was a postoperative restrictive membranous VSD with a left-to-right shunt and a maximum gradient of at least 70 mmHg. A temporary transvenous pacemaker was placed through the right femoral vein into the apex of the morphological right ventricle. A left inferior vena cava course of the lead was noted under fluoroscopy guidance [Figure 1b]. PPI was scheduled for the following day.

To evaluate the venous anatomy and minimize radiation exposure (by avoiding computed tomography angiography or venography), vascular ultrasound of the left subclavian vessels was performed before PPI. The ultrasound probe was placed inferior to the left clavicle with the indicator pointing toward the clavicle, in the midclavicular line [Figure 2a and b]. Doppler-color ultrasound showed left subclavian vein in blue color [Figure 2c]. A 90° counterclockwise rotation of the probe allowed longitudinal visualization of the subclavian vessels [Figure 3a and b]. Visualization of either subclavian artery or subclavian vein was possible with superior or inferior tilting of the probe, respectively. Bubble contrast ultrasound with agitated saline solution using a left forearm vein showed normal left subclavian vein patency and proximal drainage [Figure 3c]. On TTE, bubble contrast also showed normal drainage from left superior vena cava into the morphological right atrium, with no right-to-left shunt [Figure 3d]. After confirmation of these findings, the team agreed on proceeding with PPI without further tests, given the low likelihood of intracardiac or extracardiac abnormalities that could interfere with PPI.

Venous access was gained via the left cephalic vein. Two active fixation leads were implanted in the morphological right ventricular septum and morphological right atrial appendage using an inverted fluoroscopic image (left-to-right and opposite angulated views as needed to simulate normal anatomy). The leads were connected to a dual-chamber pacemaker. At the end of the procedure, satisfactory lead parameters were achieved. Twelve-lead ECG at the end of the procedure showed a paced rhythm at 75 beats/min [Figure 4a]. Anteroposterior and right lateral chest X-ray after permanent PPI revealed a correct placement of the right atrial and right ventricular leads [Figure 4b and c]. At 6-month follow-up, the patient was asymptomatic and pacemaker had normal sense and pace thresholds.

Discussion

We described a novel approach to PPI in a patient with situs inversus and dextrocardia, which obviates the need for more expensive and less innocuous procedures. The use of agitated saline solution and vascular ultrasound to assess venous drainage before lead positioning has not been previously described and may be useful in pediatric populations or younger patients.

In dextrocardia with situs inversus, the left subclavian vein has a more angulated distal course, which constitutes one of the main challenges of implanting pacemaker leads. In addition, fluoroscopy times during PPI are expected to be higher given the abnormal arrangement of cardiac chambers and great vessels. Using an inverted fluoroscopy image, we were able to minimize fluoroscopy times; this technique has been previously described elsewhere.[5]

Dextrocardia with situs inversus is less associated with other cardiovascular abnormalities than dextrocardia with situs solitus.[2] Thus, the chances of having concomitant venous return abnormalities in our patient were lower. A less invasive approach could then be justified as an alternative to more conventional methods (e.g., venography or computed tomography angiography). Vascular ultrasound allowed visualization of the normal subclavian vein anatomy and course. As venous drainage could still be anomalous in nonvisualized segments of the venous system, we performed bubble contrast during both vascular ultrasound and TTE. Bubble contrast confirmed normal proximal drainage and distal drainage into the morphological right atrium, without right-to-left shunts or persistence of right superior vena cava. Overall, this technique can be easily performed with just a short period of training. However, there are some caveats: (1) vascular ultrasound was only performed before PPI and not during PPI, as it would increase the intraprocedural workload; (2) vascular ultrasound only allows visualization of a portion of the venous system; nonetheless, bubble contrast can help in the indirect assessment of the nonvisualized segments.

To the best of our knowledge, the use of vascular ultrasound and agitated saline solution to assist with lead positioning in patients with dextrocardia and situs inversus has not been previously described. Vascular ultrasound shows the proximal and middle course of the subclavian vein. The addition of agitated saline solution allows better visualization of the vein and confirms correct drainage into the morphological right atrium.

This technique could be useful in pediatric populations, in whom the use of ionizing radiation is an important issue. This is a novel tool, simple, innocuous, and inexpensive, which provides guidance to the operator and obviates the need for additional examinations.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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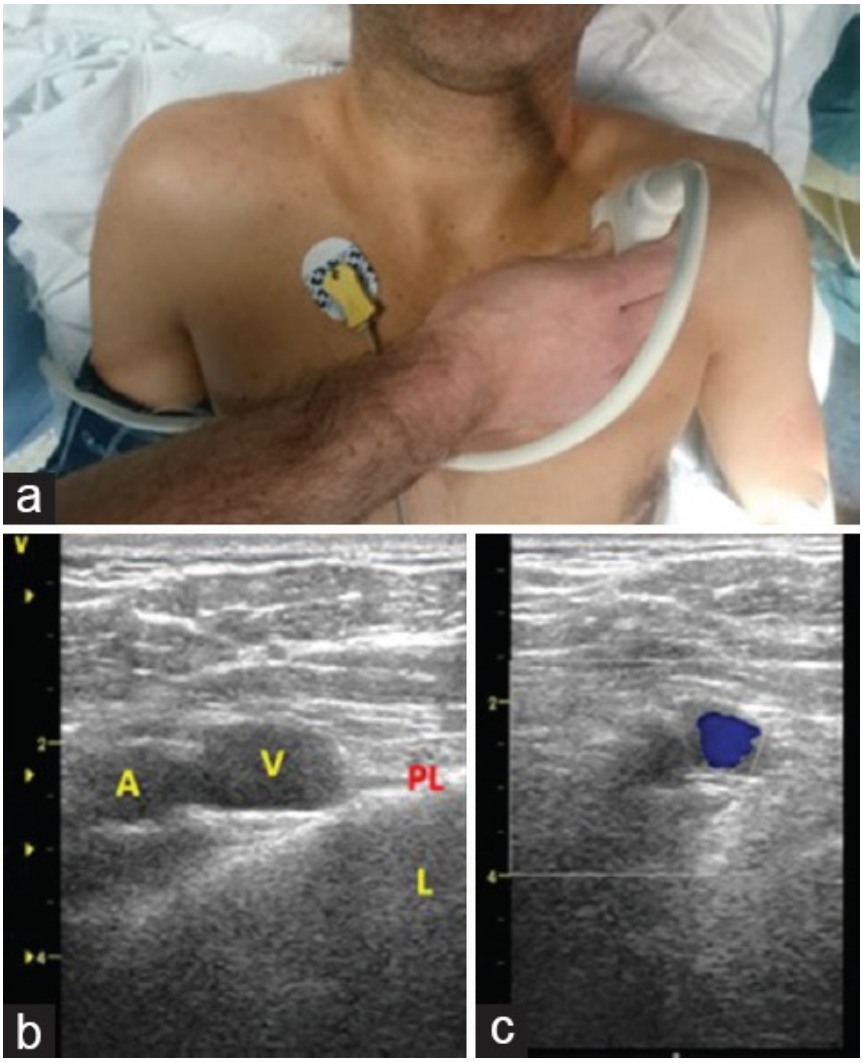
Figures and Tables

Figure 1

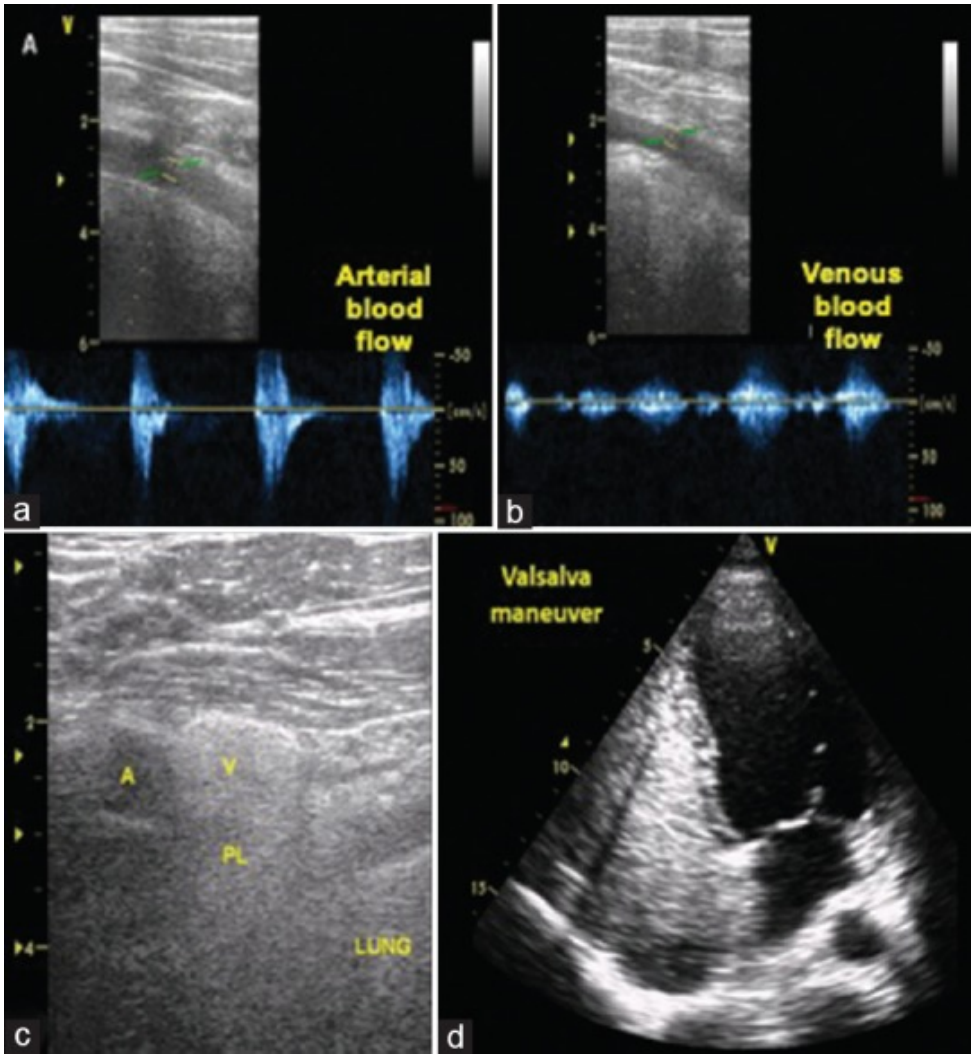
Standard left 12-lead electrocardiogram (a) and chest X-ray (b) at admission. electrocardiogram showed a complete atrioventricular block, heart rate = 32 bpm, wide QRS complexes; there was also right axis deviation, inverted P, QRS, and T waves in lead I, and absence of R-wave progression in chest lead, suggesting dextrocardia. Chest X-ray after temporary pacemaker implantation confirmed dextrocardia and showed a left inferior vena cava course of the pacemaker lead

Figure 2



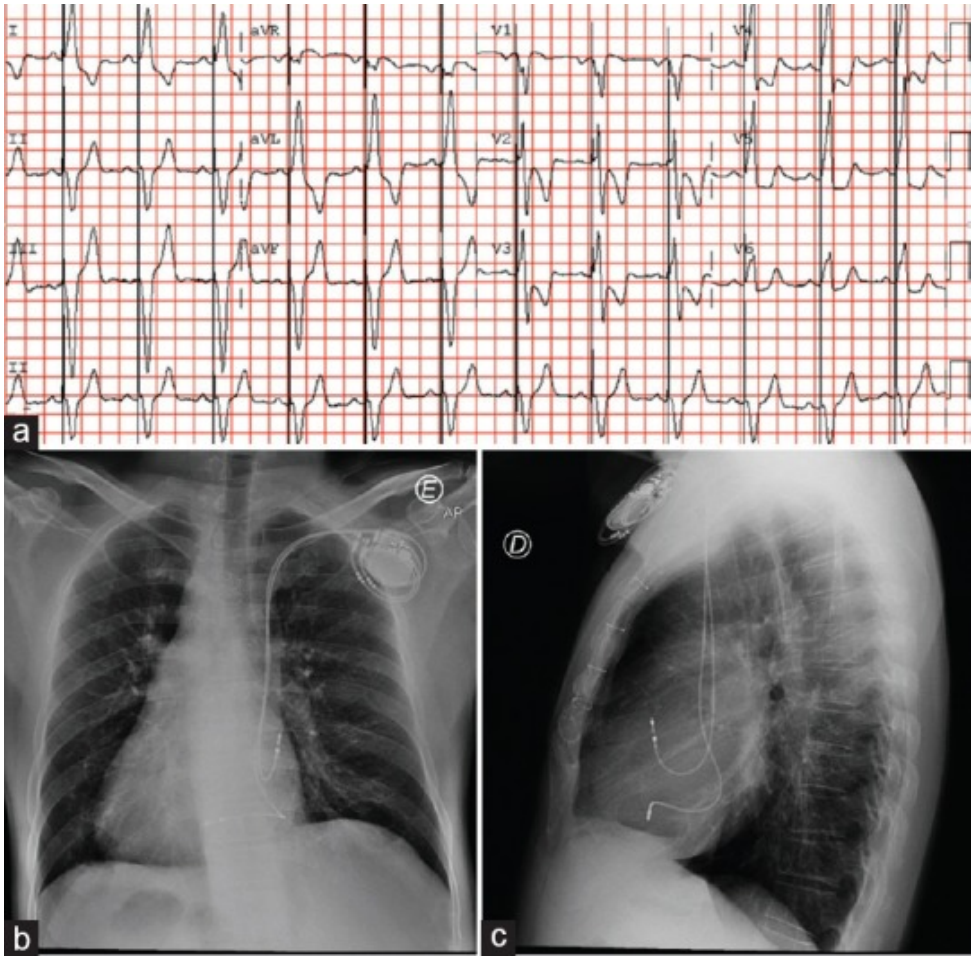
Left infraclavicular vascular ultrasound was performed before permanent pacemaker implantation. The ultrasound probe was positioned inferior to the clavicle in the midclavicular line (a). Main anatomical landmarks in this view (b) are as follows: A – Left subclavian artery, V – Left subclavian vein, PL – Pleura, L – Lung. Doppler-color ultrasound (c) showing left subclavian vein in blue color

Figure 3



Starting in the same position described as in Figure 2a, a 90° counterclockwise rotation of the probe allows longitudinal visualization of the subclavian vessels. Superior and inferior tilting of the probe allows visualization of either subclavian artery (a) or subclavian vein (b), respectively. This was further confirmed using pulsed wave doppler. Bubble contrast ultrasound showed opacification of left subclavian vein (c) and drainage from left superior vena cava without right-to-left shunt (d)

Figure 4



Twelve-lead electrocardiogram with right leads (a) showed a paced rhythm at 75 beats/min. Anteroposterior (b) and right lateral (c) chest X-ray after permanent pacemaker implantation revealed a correct placement of right atrial and right ventricular leads. Gastric bubble is also noted on the right side