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CASE REPORT

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Thoracoscopic AF ablation in situs inversus dextrocardia with interrupted inferior vena cava continuation in azygos vein

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Abstract

Introduction: Situs inversus totalis, dextrocardia with interrupted inferior vena cava, and azygos vein continuation concomitant with symptomatic atrial fibrillation requiring ablation. This case was deemed not suitable for percutaneous ablation due to anatomic variations and the lack of case reports in the literature.

Methods and Results: We performed bilateral thoracoscopic epicardial ablation and epicardial left atrial appendage exclusion. The direct vision allowed for a complete box lesion set with bipolar radiofrequency device. Patient remained in sinus rhythm at the 12-months follow-up.

Conclusion: Surgical thoracoscopic epicardial ablation is safe and effective also in congenital defects. Multidisciplinary expertise can offer minimally invasive ablation treatments.

KEYWORDS

atrial fibrillation, dextrocardia, interrupted inferior vena cava, left atrial appendage, left atrial appendage exclusion, minimally invasive, radiofrequency ablation, surgical thoracoscopic ablation, situs inversus totalis

1 | INTRODUCTION

Intrahepatic interruption of the inferior vena cava (IVC) with azygos vein continuation is a rare congenital aberration, occurring in 0.6% of patients with other cardiac defects as situs abnormalities and dextrocardia.[1] In most instances, patients are asymptomatic. They might be noticed as incidental findings following imaging investigations. We present a case of situs inversus totalis and dextrocardia associated with interrupted IVC and azygos vein continuation in a patient with concomitant long-persistent atrial fibrillation (AF). The treatment of AF has been considered at high risk for percutaneous interventions and thus referred to our institute for surgical bilateral thoracoscopic epicardial ablation and left atrial appendage (LAA) exclusion.

The patient is a 45-years old male with symptomatic long-standing persistent atrial fibrillation (LsP-AF) who received multiple electrical cardioversions for AF recurrences since 2003. Class I antiarrhythmic drugs failed to restore sinus rhythm thus, according to the current ESC 2020 Guidelines for AF treatment,[2] the patient was scheduled for transcatheter pulmonary veins (PVs) isolation.[3] Preoperative chest X-Ray showed the presence of a complete situs inversus dextrocardia (SID) (Figure 1A).

Then, thoraco-abdominal computed tomography (CT) scan was performed to rule out other anatomical abnormalities. A concomitant

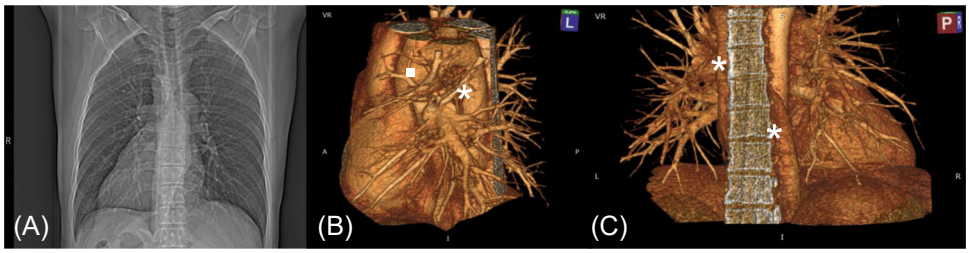

FIGURE 1 (A) Chest X-Ray showing complete situs inversus dextrocardia. (B) Left-sided view showing azygos vein (white asterisk) continuation draining into the posterior aspect of the superior vena cava (white dot). (C) Posterior view of the azygos vein (white asterisk) crossing the midline.

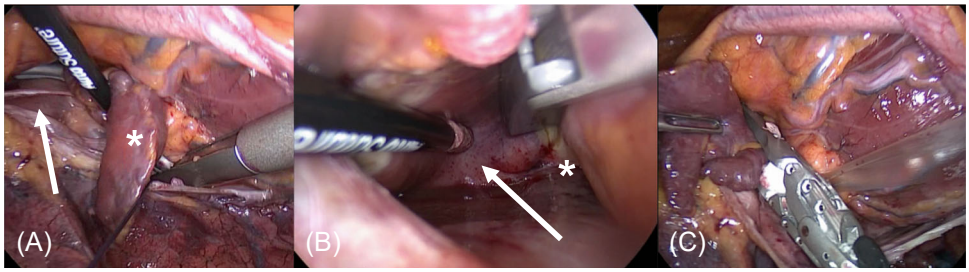

FIGURE 2 (A) Right curved bipolar ablation device encircling the right-sided pulmonary veins (white arrow: right superior pulmonary vein; white asterisk: left atrial appendage). (B) Linear radiofrequency device performing connecting lesion at the level of the floor of the box lesion (white arrow: left inferior pulmonary vein; white asterisk: confluence of the hepatic veins). (C) Left atrial appendage exclusion device positioning.

intrahepatic interruption of the IVC was described with the renal veins draining into the azygos vein which was directly collecting blood from the lower body and draining it posteriorly into the superior vena cava (SVC). In adjunct, the hepatic veins were draining directly into the right atrium (RA). Nonetheless, regular anatomy of the left atrium (LA) with two right and two left PVs was depicted (Figure 1B,C).

The patient was deemed not suitable for percutaneous ablation (PA) because of the complex anatomy and was then referred to our Institution for surgical thoracoscopic ablation. Transesophageal echocardiogram was performed to rule out the presence of thrombus in the LAA before surgery.

2 | METHODS

Thoracoscopic procedure was performed starting from the chest side in which LAA was promptly approachable: the right side. Two 10-mm ports and one 5-mm port were placed along the right anterior axillary (3th and 5th intercostal space) and midaxillary line (4th intercostal space) as previously described for classic anatomy procedures.[3-5] Briefly, once the pericardium was opened, the LAA was visualized as a reference. A blunt dissection at the level of the roof of the LA was performed to allow the right curved bipolar radiofrequency (RF) ablation device (Isolator Synergy Clamp, EMR2; AtriCure Inc) to encircle the PV on the right side (Figure 2A). The device followed the route previously prepared by means of a track-light dissector (Lumitip, AtriCure Inc). Connecting lesions were then performed by using a linear bipolar RF device (Coolrail linear pen, AtriCure Inc) at the level of the inferior side of the Box Lesion (Figure 2B). LAA occlusion was performed by means of AtriClip Pro 2 device 40 mm (AtriCure Inc). We then moved ports to the contralateral side to complete the thoracoscopic surgical ablation (Figure 2C).

A blunt dissection of the pericardial reflection below the confluence of the two sovra-hepatic veins (draining into the RA) was performed to gain access to the oblique sinus (Figure 3A). The access to the transverse sinus was obtained by gently dissecting the pericardial reflection below the SVC. PVs at the left side were encircled and RF energy was delivered by means of a left curved bipolar RF ablation device (Isolator Synergy Clamp, EML2 AtriCure Inc) (Figure 3B). Once completed, connecting lesions between right and left PVs were performed at the level of the roof of the LA (Coolrail, AtriCure Inc). Exit block was confirmed at the end of the procedure with a total surgical time of 200 min.

3 | RESULTS

No complications occurred intra-operatively and during hospitalization. The patient was discharged in sinus rhythm after 4 days. At 3 and 12 months follow-up, the patient was in stable sinus rhythm without experiencing recurrent symptomatic episodes of AF, as confirmed by means of holter-ECG evaluation.

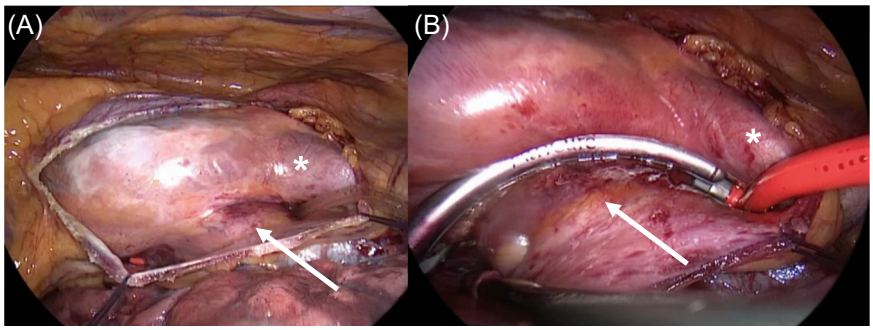

FIGURE 3 (A) Left anatomy after pericardial opening. (B) Left-curved bipolar ablation device encircling the left-sided pulmonary vein. (white arrow: left pulmonary vein; white asterisk: superior vena cava).

4 | DISCUSSION

The incidence of dextrocardia with situs inversus totalis is 1/10,0000–50,000 births.[6] In such patients, the IVC might be interrupted or stenotic in 8%–18% of patients, with azygos continuation present in only 0.6% of cases.[7] Unless severe concomitant congenital defects occur, patients with SID have life expectancy similar to that of the general population.[8] In the literature, only few case reports described percutaneous ablation (PA) strategies in this specific subset of patients, in which PVs isolation can be extremely challenging. The major limitations are considered: small caliber of the entrance vessel, often complicated by tortuosity especially at the level of the azygos vein;[9] difficulty in obtaining trans-septal puncture; limited maneuverability of the ablation/mapping system.[6] For these reasons, "unconventional methods" were explored and described in literature[10,11]: so far, three different PA approaches have been reported: trans-septal puncture via trans-jugular approach through the SVC[8]; transaortic retrograde approach,[9] and transhepatic vein approach.[11]

Trans-septal puncture via internal jugular/subclavian vein and SVC was described by Masumoto et al.[9] PVs isolation was effectively performed under 3D navigation system. However, the procedure was reported as extremely long despite authors simplified the procedure by omitting electrophysiological mapping of the LA (over 360 min of procedural time with more than 60 min of pulmonary vein isolation time). Major concerns raised mainly while performing the trans-septal puncture with a procedure generally defined as "not smooth". In this case, operators avoided to approaching the IVC because of the tortuosity.[8]

An intriguing trans-aortic solution was reported by Okajima and colleagues[10] in this specific subset of patients. Via the left femoral artery authors gained access to the LA in a retrograde fashion and under magnetic navigation system. However, authors were unable to perform a complete isolation of the four PVs: the right inferior PV isolation was aborted since the ablation catheter repeatedly dropped into the left ventricle because of an unfavorable angle and short distance between mitral annulus and PV orifice. Moreover, to simplify a such complex procedure, authors avoided to evaluate entrance/exit block as well as the electro-anatomical mapping.[9,10]

A percutaneous trans-hepatic vein approach was described by Tandon et al.[10] in a patient with dextrocardia and IVC continuation in the azygos vein. LA access was gained via hepatic vein puncture under ultrasound guidance and fluoroscopy. The procedure was reported as safe, however, only addressed the LAA.[11]

Noninvasive percutaneous solutions provided a stable restoration of the sinus rhythm, however, they were reported as particularly challenging. In most instances, the major drawbacks were the necessity to use different types of catheters, the complex catheter guidance and maneuverability, thus leading to exceedingly long and often incomplete procedures.

Surgical thoracoscopic AF ablation had the advantage of a direct vision of the complex anatomy of the patient, allowing the surgeons to promptly recognize cardiac structures despite the complete SID. Effective lesions were safely performed and confirmed by testing the presence of the exit block, thus without affecting the completeness of the ablation lesion set and the procedural time. Moreover, the LAA was successfully excluded by epicardial access with no additional risk.

5 | CONCLUSION

In conclusion, patients with complex anatomy should be carefully evaluated with a multidisciplinary approach in specialistic facilities with a wide expertise in minimally invasive arrhythmia surgery. A patient-tailored approach was guaranteed once risks and benefits of the surgical procedure over percutaneous strategy were wisely balanced, thus providing the best option in terms of safety, efficacy, and patient satisfaction.

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CONFLICTS OF INTEREST

Stefano Benussi is consultant/proctor for Atricure, Allergan, and Cryolife. The remaining authors declare no conflict of interest.

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