High-density mapping and ablation of an incessant ventricular tachycardia with endo-epicardial involvement in a patient with situs inversus totalis and left ventricular noncompaction

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INTRODUCTION
Dextrocardia with Situs Inversus (situs inversus totalis) is a rare heart condition characterized by abnormal positioning of the heart. In this condition, the tip of the heart (apex) is positioned on the right side of the chest. Additionally, the position of the heart chambers, as well as the visceral organs such as the liver and spleen, is reversed (situs inversus).

This condition can be associated with left ventricular noncompaction even though the real incidence of the very rare association of situs inversus totalis and left ventricular noncompaction is still not known. The arrhythmic substrate in patients with left ventricular noncompaction and sustained VT typically involves the left ventricle (LV) apex and midapical segments, but more basal and epicardial involvement has also been demonstrated.

CASE DESCRIPTION
A 48-year-old male with situs inversus totalis and left ventricular noncompaction, normal ejection fraction (55%) and dual chamber implantable cardioverter defibrillator implanted for secondary prevention (Figure 1 lower panel), was referred to our center after a failed endocardial ablation attempt of an incessant ventricular tachycardia (VT) in another institution. The VT Electrocardiogram (Figure 1 upper panel), recorded with the electrodes placed in reversed positions, exhibited right bundle branch block and superior axis QRS morphology. Due to the nonischemic nature of the substrate and the previous failed endocardial ablation, we opted for an endo-epicardial approach.

As far as the epicardial puncture is concerned, it was carried out in a mirror image to the standard approach (Figure 2): considering the position of the apical LV pointing to the right and the inverted position of the liver and other abdominal organs, a Tuohy needle was inserted in the subxiphoidal space with its tip pointing towards the right shoulder. Epicardial access was easily gained with a posterior approach and the right anterior projection was used to confirm the guidewire route going from left to right before inserting the epicardial sheath (Video S1). Epicardial mapping during VT was obtained by means of the Rhythmia mapping system and the Orion catheter (Boston Scientific) and revealed an incomplete reentrant activation with very low-amplitude presystolic electrograms (EGMs) in a very small area in the posterobasal wall, with lack of middiastolic activity. For this reason, the Orion catheter was inserted in the endocardial LV through the aorta (Video S5) but endocardial activation map of the arrhythmia did not cover the full cycle length: only very low-amplitude signals in the early and middiastolic phase could be recorded in the posterobasal wall, but no late-diastolic and presystolic activities were detected by the Orion catheter despite very careful mapping. The smallest EGM components were not detected by the worse signal resolution of the ablation catheter and entrainment attempts failed due to inability to capture both in the endo- and epicardial posterior wall. By combining endo- and epicardial maps a more complete activation...
spanning a greater percentage of the full cycle length could be obtained: early and middiastolic EGMs were recorded in the endocardial posterobasal wall, while presystolic EGMs were detected in the epicardial posterobasal wall. Late-diastolic activity was still missing in both maps and we inferred that it was located in the intramural midmyocardial posterobasal wall (Figure 3; Video S2). The IntellaNav MIFI OI catheter (Boston Scientific) was used to deliver high-power applications (50 W) both in the endocardial and epicardial posterior wall. Several endocardial radiofrequency ablation lesions interrupted the arrhythmia (Video S3), however it was easily reinducible after each application. Subsequent epicardial RF lesions first changed VT exit and morphology (Video S4) and finally terminated the arrhythmia after cycle length slowing, rendering it noninducible in the course of time. Three-dimensional circuit activation is defined by the presence of incomplete reentrant patterns on one or both endo-epicardial surfaces, where activation gaps represent propagation at a sufficient depth that cannot be sensed by surface recordings (inferred intramural midmyocardial activation). In this case, involvement of both endo- and epicardial surfaces in the reentry with evidence of activation gaps during distal isthmus activation, led us to deliver high-power ablation lesions in both surfaces that ended up in no VT inducibility. The challenges of this case were not exclusively related to the complexity of the reentrant pattern but also to the anatomy of the situs inversus totalis, since manipulating catheters required movements to be performed in the opposite direction of what would be normally done and epicardial access was carried out in a “mirror fashion” (with the needle tip pointing to the right shoulder).

3 | DISCUSSION

Recent research has shown how simultaneous endocardial and epicardial mapping with high-resolution technology improves the ability to characterize the 3D nature of scar-related reentrant VT. Epicardial mapping and ablation in patients with situs inversus totalis is feasible and safe. The very rare combination of this condition
with left ventricular noncompaction can be associated with complex reentrant VT circuits that require mapping and ablation in both endocardial and epicardial surfaces.

DISCLOSURE
Dr Francesco Solimene and Giuseppe Stabile have received speaker’s fees from Boston Scientific Corporation. Francesco Maddaluno is a Boston Scientific employee.

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FIGURE 3  Left panel: Representatives EGMs recorded from beginning of diastole to presystolic phase both in endocardial and epicardial posterobasal LV. Endocardial recordings show activities spanning from the beginning to the mid diastolic phase of the reentry with lack of late-diastolic and presystolic activities. Epicardial recordings in the same region show presystolic EGMs. The yellow brackets between endocardial and epicardial recordings indicate the late-diastolic phase where we couldn’t record any activity neither in the endocardium or epicardium. We inferred that these missing EGMs (about 10% of cycle length) were concealed intramural mid-myocardial activation. Right panel A: Proposed VT circuit involving a very small region of the postero-basal LV: the early and middiastolic signals are recorded in the endocardial wall (white arrows). In the late-diastolic phase (yellow arrow) the wavefront is mid-miocardially propagating to the epicardial wall where presystolic activities (VT exit site, blue arrow) are recorded. After epicardial activation, the propagation wavefront re-activates the endocardial wall (white star) in the systolic phase and the reentry continues. For a better visualization of this complex reentrant pattern see Video S2. Right panel B: White arrows showing endocardial propagation without epicardial activation (epicardial wall is shaded to show endocardial postero-basal wall). Endocardial activation starts at the white star location as a pseudofocal pattern in the systolic phase of the reentry. The activation wavefront curves around a functional line of block (black line) to enter the diastolic phase of the reentry (light blue, blue and purple color in the activation map). The reentrant activity can’t close the loop in the endocardium because the activation stops at the black line, requiring an epicardial bridge to come back at the star location. EGM, electrogram; LV, left ventricle; VT, ventricular tachycardia

SUPPORTING INFORMATION
Additional supporting information may be found online in the Supporting Information section.

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