Ventricular tachycardia in a patient with repaired d-transposition of the great arteries

Philipp Krisai, MD,* Konstantinos Vlachos, MD, PhD,* Nadir Tafer, MD,† Hubert Cochet, MD,‡ Xavier Iriart, MD,§ Frédéric Sacher, MD, PhD*

From the *Department of Cardiac Electrophysiology, IHU Liryc, Electrophysiology and Heart Modeling Institute, Univ. Bordeaux, Bordeaux University Hospital (CHU), Pessac-Bordeaux, France, †Department of Congenital Heart Disease and Anesthesiology, Centre hospitalier universitaire de Bordeaux, Pessac-Bordeaux, France, ‡Department of Cardiovascular Imaging, Centre hospitalier universitaire de Bordeaux, Pessac-Bordeaux, France, and §Department of Pediatric and Adult Congenital Cardiology, Centre hospitalier universitaire de Bordeaux, Pessac-Bordeaux, France.

Introduction
The risk for life-threatening arrhythmia in patients with surgically corrected congenital heart disease is very low and is most commonly owing to scar in the area of a repaired ventricular septum defect or ischemia.1 We present a rare case of scar-related ventricular tachycardia (VT) in the area of the sinus of Valsalva (SV) late after surgically corrected dextro-transposition of the great arteries (d-TGA).

Case report
A 17-year-old male patient with a history of repaired transposition of the great arteries presented to the emergency department with a suspected panic attack. His congenital heart defect consisted of dextro-transposition of the aorta anterior to the pulmonary artery, resulting in 2 parallel circulatory circuits instead of a single serial circulation. Blood oxygenation after birth was maintained by a patent foramen ovale and a persistent ductus arteriosus (Supplemental Figure S1A). The surgical correction included an arterial switch of the pulmonary artery and the ascending aorta distal to the coronary arteries with the LeCompte maneuver, a reinsertion of the coronary arteries at the SV of the neoaortic valve, closure of the patent foramen ovale, and ligation of the persistent ductus arteriosus (Supplemental Figure S1B).

In the emergency department, the electrocardiogram revealed a rapid VT with a rate of 240 beats per minute with 2 slightly different QRS morphologies (both inferior axis and negative in I and aVL; the first with a precordial transition in V3, the second with positive precordial concordance, Supplemental Figure S2). Both morphologies suggested a VT origin from the area of the left ventricular summit with different exits. The first morphology with an abrupt precordial transition in V3 and a precordial pattern break supported a more rightward exit. The second morphology with positive precordial concordance was more in line with a leftward exit. The patient was successfully cardioverted and scheduled for cardiac ablation.

Preinterventional cardiac magnetic resonance imaging and multidetector computed tomography (CT) scans were merged on a submillimetric level using MUSIC software.3 Myocardial and vascular structures were segmented using the CT images, whereas scar areas were defined based on cardiac magnetic resonance imaging. The scar threshold was defined as 50%–100% of the maximal myocardial signal. The segmented images were then used to generate 3D surface meshes of each relevant cardiac structure (endo- and epicardium, coronary arteries, coronary sinus, and scar) and were imported into the 3D mapping system (CARTO 3; Biosense Webster, Diamond Bar, CA) for real-time guidance during the procedure. With this

KEY TEACHING POINTS
• Patients with repaired d-transposition of the great arteries might experience scar-related ventricular tachycardia originating from the aortic root.
• Detailed preinterventional imaging is necessary to identify the scar area.
• Preinterventional imaging allows to choose the optimal ablation approach from different anatomical sites.

KEYWORDS
Ablation; Adult congenital heart disease; Cardiac imaging; Transposition of the great arteries; Ventricular tachycardia

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approach, we could identify circumscribed scar in the area of the aortic root below the left coronary cusp (LCC). This substrate’s location was compatible with the origin of the VT morphologies and was likely due to a surgical incision or trauma below the reinsertion of the coronary arteries during his cardiac repair, which was not reported in the surgical notes (Figure 1 and Supplemental Video). His cardiac imaging and coronary angiography additionally revealed an anomalous origin of the circumflex artery from the right coronary artery without evidence of stenosis.

In the electrophysiology laboratory, we used an irrigated, bidirectional contact force catheter (ThermoCool Smart-Touch SF; Biosense Webster, Diamond Bar, CA) for all mapping and ablation and a decapolar steerable diagnostic catheter (Dynamic Tip; Boston Scientific, Marlborough, MA) as a reference in the coronary sinus. First, we mapped the coronary sinus and the right ventricle as well as the aorta via a retrograde approach to facilitate a merge with the MUSIC images. We then recorded fractioned potentials in the LCC (Figure 2A), with pacemapping at this location showing a good match with the first VT morphology (Figure 3A). No clearly fractioned potentials were recorded below the LCC (Figure 2B) and a slightly worse pacemap was recorded from the great cardiac vein (Figure 3A). Programmed stimulation with up to 3 extrastimuli (coupling intervals down to 200 ms, drivetrain of 5 beats) under resting conditions failed to induce VT. However, after administration of isoproterenol, a single extrastimulus at a coupling interval of 230 ms induced a hemodynamically tolerated VT with a cycle length of 250 ms and similar morphology to the clinical VT (Figure 3A). Activation mapping confirmed the earliest activation in the LCC with termination of the tachycardia after 15 seconds of radiofrequency ablation at 35 watts (Figure 3B and C) at a safe distance from the coronary artery ostium, as shown on the integrated CT scan. To consolidate the ablation of the target scar, we additionally ablated from below the LCC via a retrograde approach with 35 watts for up to 90 seconds (Figure 2B) and from inside the great cardiac vein with 20 watts and a higher flow rate of 20 mL/min for up to 60 seconds. After ablation, the VT was noninducible under isoproterenol and the patient remained free of VT during a follow-up of 3 months.

**Discussion**

The current case is the first to relate a scar area in the SV to sustained VT in this patient population. This highlights the importance of detailed cardiac imaging studies to define the etiology and to choose the optimal ablation approach from different endocardial and epicardial sites.

Sustained VT after arterial switch operation for d-TGA is a rare but potentially life-threatening complication.1 Most commonly, VTs in this patient population originate from areas of myocardial ischemia or from the repaired ventricular septal defect.1 To the best of our knowledge, only 2 prior cases have reported a VT origin and successful ablation in the area of the SV independent of ischemia or a ventricular septal defect.4,5 Maury and colleagues4 described a case of sustained, monomorphic VT in a 15-year-old boy that was only inducible with atropine and isoproterenol, similar to our case. The successful ablation site was in the posterior and in the left-sided noncoronary SV, where postsystolic, fractionated potentials were found.4 Bhaskaran and colleagues5 reported on a 30-year-old man with sustained VT that was successfully ablated at the left noncoronary cusp junction also in an area of fractioned electrograms. Although late and fractionated potentials supported surgical scar reentry as the underlying cause in both cases, imaging studies did not reveal any scar tissue. Thus, idiopathic or triggered outflow tract VT could not be excluded.4,5

Our case is the first to directly relate scar in the area of aortic root, most probably postsurgical, with sustained VT in a patient after arterial switch operation. Detailed preinterventional cardiac imaging helped us to plan a comprehensive,
sandwich” ablation approach from different sites. The detection of scar supports reentry as the most likely VT mechanism in this patient.

**Figure 2** Catheter positions shown in the 3D mapping system (left) and in fluoroscopic anterior-posterior views (middle) inside the left coronary cusp (A) and below the left coronary cusp (B), with 1 surface electrocardiogram and intracardiac electrograms from the ablation catheter (right). Fractionated potentials were noted inside the left coronary cusp (A, right), but not below the cusp. Arrows indicate the amplitude (0.5 mV) and sweep speed (100 ms) of the intracardiac tracings.

**Figure 3** A: Pacemap from the left coronary cusp (left), pacemap from the great cardiac vein (middle), and induction of ventricular tachycardia with isoproterenol (right). B: Bipolar and unipolar electrograms in the left coronary cusp during ventricular tachycardia at the successful ablation site. C: Termination of the tachycardia after 15 seconds of radiofrequency ablation. GCV = great cardiac vein; LCC = left coronary cusp.

**Conclusion**
Patients with repaired d-TGA might experience scar-related VT originating from the aortic root. Detailed
preinterventional imaging is beneficial to identify the scar area and to choose the optimal ablation approach.

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Appendix
Supplementary data
Supplementary data associated with this article can be found in the online version at https://10.1016/j.hrcr.2020.10.006.

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