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Non-invasive isthmus identification of complex arrhythmias in congenital heart disease

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1 | CLINICAL PRESENTATION

A 30-year-old man with the previous history of surgical repaired Tetralogy of Fallot (rTOF) was referred to our arrhythmia unit for sudden onset palpitations, associated with light-headedness and presyncope.

Clinical history revealed surgical rTOF with pulmonary valvotomy, closure with a direct suture of ventricular septal defect performed and subsequent implantation of valvulated homograft biological prosthesis. Due to stenosis of the biological conduit implantation of a percutaneous pulmonary valve stented prosthesis implant (Melody™) was performed, at the of age 22 years old.

A previous 24-hour EKG monitoring recorded brief runs of supraventricular tachycardia and a low burden of ventricular premature beats without symptoms during recording.

A treadmill test revealed reduced functional capability, without exertional arrhythmias.

2 | NO INVASIVE SUBSTRATE AND ISTHMUS IDENTIFICATION

A Cardiac angio-MRI showed mild-moderate stenosis of the main pulmonary artery, adjacent to the stent, mild-moderate regurgitation of the biological prosthesis pulmonary valve, dilated and hypertrophic RV, with hypertrabeculation and moderate systolic dysfunction.

There was late gadolinium enhancement (LGE) of the anterior and inferior insertion points, but also of the RV outflow tract, basal portion of the interventricular septum and mid to apical free wall of the RV. The right atrium was also analyzed in MRI with LGE in the lateral wall and posterolateral wall. Both atrium and ventricular cardiac MRI LGE areas were suggestive of post-surgical changes producing extensive replacement fibrosis.

All LGE-MRI images were segmented with ADAS3D software (ADAS3D Medical, Barcelona, Catalonia, Spain). The atrial and the ventricular walls were manually traced in axial slices and the blood pool was automatically calculated. Color-coded pixel signal intensity maps were projected to the 3D shell of the atrium and ventriculi. (Figure 1)

The resulting processed 3D model of the RA and RV LGE-MRI was than integrated with the electroanatomical map of the navigation system.

3 | ELECTROPHYSIOLOGICAL STUDY

Due to the patient's symptoms and to stratify the risk of sudden cardiac death an electrophysiological study with an ultra-high density electro-anatomical mapping system (RHYTHMIA HDx™ Mapping System, Boston Scientific, USA) was planned.

First substrate mapping reconstruction of right atrium was quickly mapped by collecting electroanatomical points (voltage) with the IntellaMap™ Orion (Boston Scientific, USA) ultra-high density mapping catheter. Areas of low voltage suggestive of fibrosis

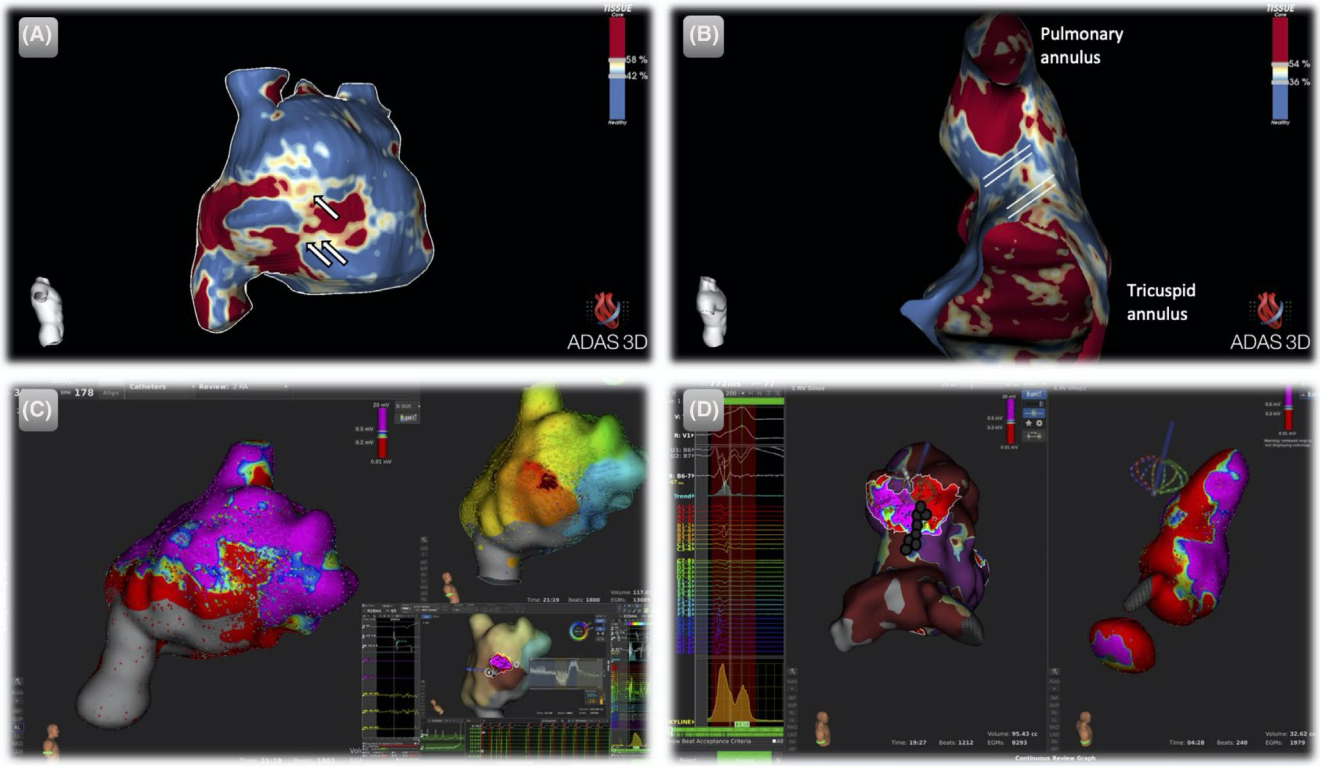

FIGURE 1 LGE-MRI 3D reconstruction (panels A and B) compared with Electroanatomic findings (panels C and D). Both atrium and ventricular cardiac MRI LGE areas were suggestive of post-surgical changes producing extensive replacement fibrosis, and suggest possible slow conducting isthmuses that could lead to re-entrant arrhythmias (Fig. A and B, white arrows and bars). Areas of slow conduction were localized during electroanatomic mapping and confirmed the presence of arrhythmia isthmus with adequate coincidence with the LGE-MRI. Right ventricle voltage and activation map was performed and identified areas of extensive and areas of slow conduction between both scars, concordant with CMR reconstruction (D). An ablation line between both low voltage areas was performed (black dots) to achieve non inducibility of VT

replacement were localized in the lateral wall of right atrium with adequate concordance with the LGE-MRI localization.

After a brief atrial continuous stimulation (at 400 ms) a sustained atypical atrial flutter was induced (TCL 240 ms). The activation map showed a clockwise circuit in the right atrium lateral wall. (Figure 2).

The isthmus of the flutter on the lateral side of the right atrium wall in an area of heterogeneous tissue adjacent to the dense scar, probably in relation to atrial incision of previous cardiac surgery, was localized (Figure 2) in the same area where the cardiac MRI identified the possible arrhythmia isthmus.

The ablation catheter was moved up to region suggested. Entrainment showed a post-pacing interval identical to the flutter cycle at that point. A radiofrequency (45°C, 30W) application was delivered and converted the patient to sinus rhythm (Figure 3). An ablation line was then completed from the area of intermediate scar to the dense scar that extended to IVC. Subsequently, a protocol of atrial induction was performed without inducing atrial flutter or other atrial arrhythmias.

Finally, a ventricular tachycardia induction with programmed electrical stimulation was performed. While stimulating apex of RV with train of stimulus followed by up to 3 extra stimuli, a sustained monomorphic ventricular tachycardia was induced. The morphology was right bundle branch block with inferior axis, 260 bpm, which required external cardioversion due to hemodynamic instability. VT morphology was suggestive of a conductive isthmus between VSD direct suture and pulmonary valve (so-called isthmus 3) activated in clockwise direction; heterogeneous tissue isthmus was previously identified by the cardiac MRI reconstruction in the same zone.

Using the high-density mapping catheter, during sinus rhythm, we performed a right ventricle voltage and activation map. We identified two areas of low voltage zone compatible with extensive fibrosis (anterior part of the RV outflow tract and basal areas of interventricular septum in relation to previous surgeries) and areas of intermediate tissue with potential slow conduction between both scars (Figure 4) concordant with ventricular LGE at the cardiac MRI (Figure 1) that we had into the navigation system side-by-side from the beginning of the procedure.

Subsequently, the ablation catheter was ascended to the region of interest, performing ablation line (45°C, 40W) between both low voltage areas. We checked and confirmed the conduction block in the line with a fast activation map the Orion catheter, with no viable tissue in the ablation line (Figure 4). Finally, the same induction protocol (including pacing from apex and RVOT close to the line with

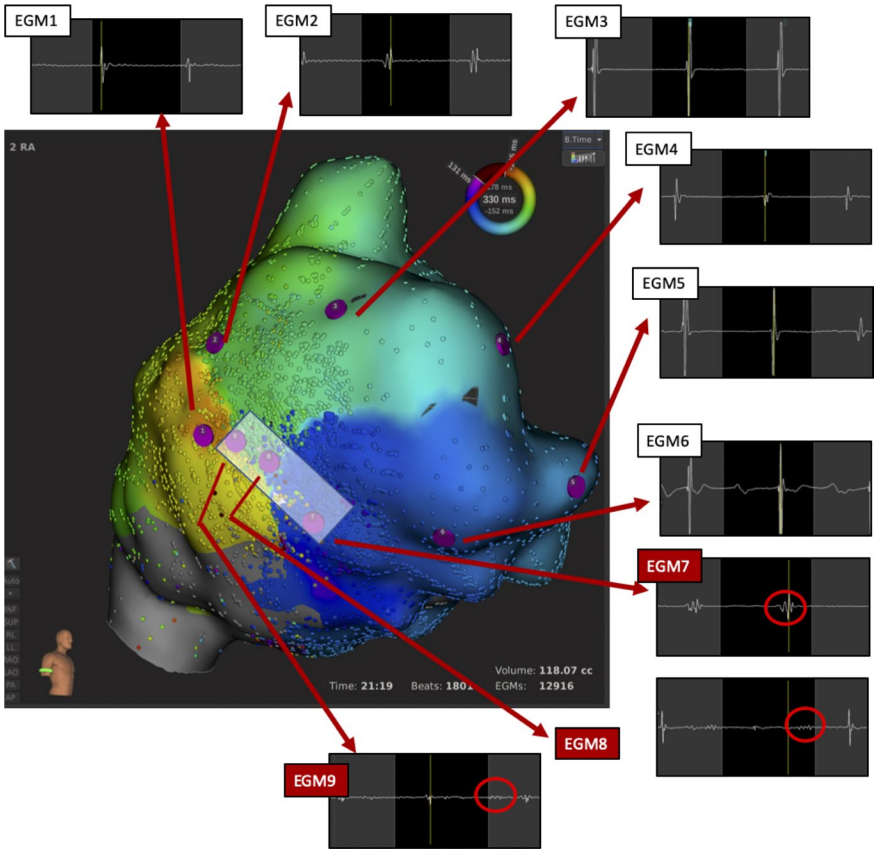

FIGURE 2 Atrial flutter electrophysiologic study and activation electroanatomic maps. The activation map shows a clockwise flutter in the lateral right atrium wall with area of slow conduction (electrograms 7-9) that correspond to the area of the heterogenous tissue of the scar

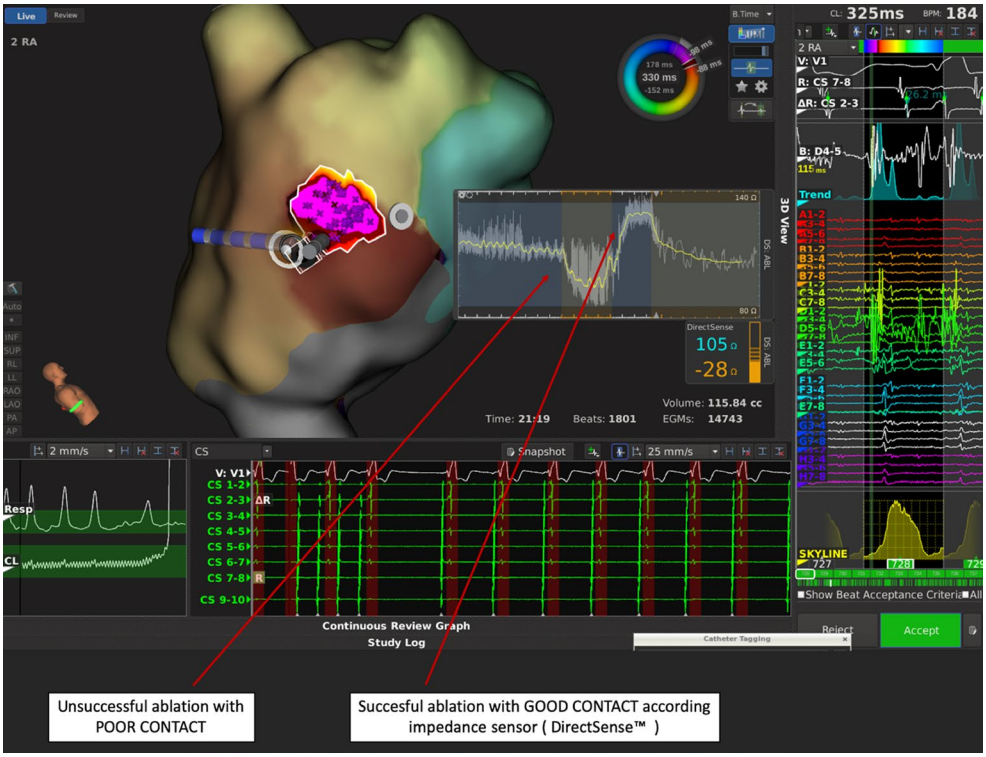

FIGURE 3 Atrial flutter ablation. Clockwise flutter in the lateral right atrium wall with area of slow conduction in the heterogenous tissue of the scar. Two RF applications are shown, first one with poor contact and a second one (effective in sinus rhythm restoration) with good contact (according Direct Sense™ algorithm)

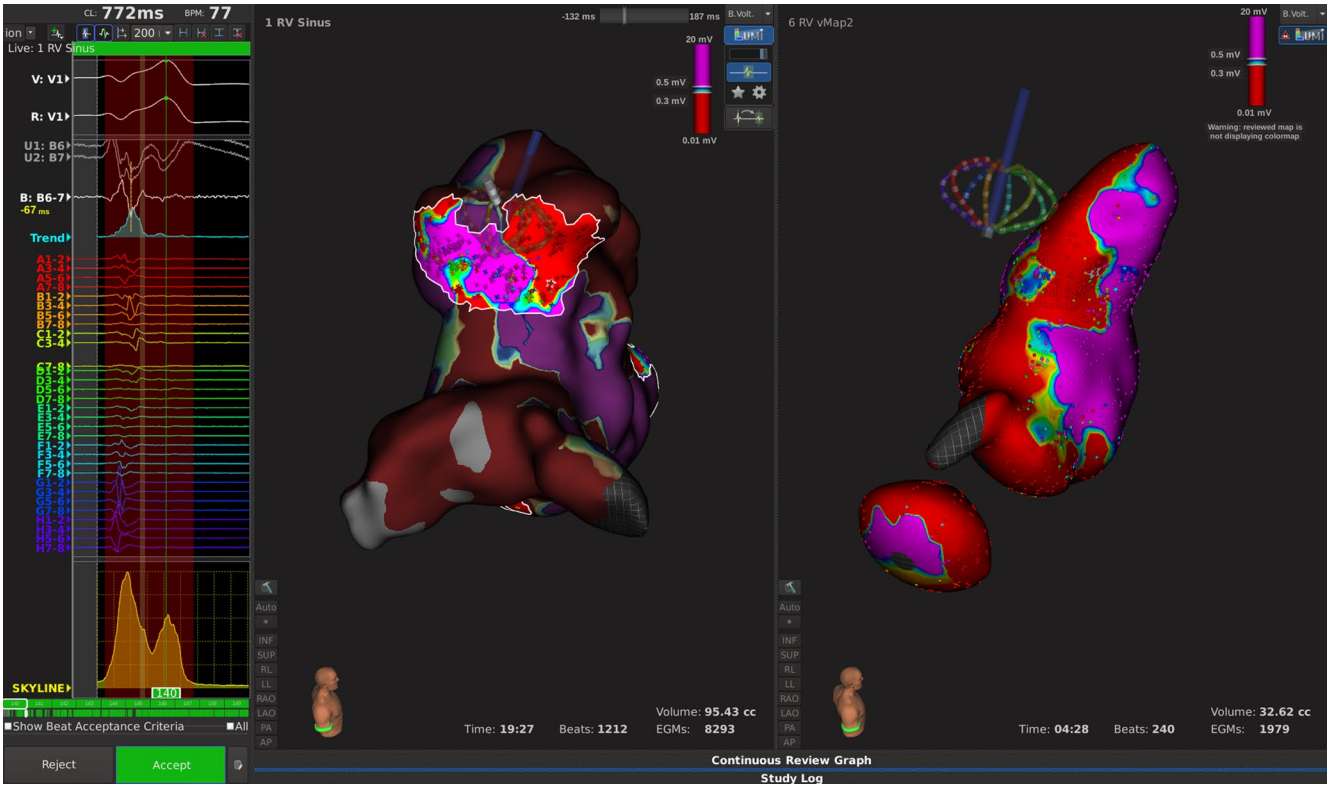

FIGURE 4 Right ventricle electroanatomic map. Left panel. Right lateral view of RVOT voltage map showing voltage isthmus between VSD patch scar and RVOT scar with slow conduction at that area according Lumipoint™ algorithm. Right panel. Voltage map after ablation with no viable tissue in the ablation line suggestive of isthmus elimination

up to 3 extrastimulus) was repeated without inducing ventricular tachycardia.

The patient was subsequently subjected to a subcutaneous automated cardioverter defibrillator implantation (S-ICD) due to SCD risk. One year after the procedure, he did not complain of new episodes of palpitation, nor arrhythmias were detected by S-ICD.

4 | DISCUSSION

We presented a case of rTOF with symptoms related to cardiac arrhythmias. Both atrial and ventricular substrate lead to develop atrial flutter and ventricular tachycardia in the same patient. These arrhythmias were attributable to re-entry circuits generated around scars of previous reparative surgery and/or fibrotic areas. The use of a preprocedural LGE MRI with postprocessing model integrated in ultra-high definition mapping system that analyses electrograms characteristics on top of timing and voltage amplitude allowed us to rapidly recognize the circuit with subsequent immediate tailored treatment with radiofrequency ablation in the same procedure.

Mechanisms of these arrhythmias are related to general mechanisms of re-entry in relation with surgical scars or surgical patches and fibrosis of the atrial wall with areas of slow conduction and sites of the unidirectional block.

Because of this slow intra-atrial conduction, IART is more frequent in CHD than in the general population.

To the best of our knowledge, there are no data about the possible application of tissue characterization and preprocedural imaging integration with MRI reconstruction in this type of patients, especially for the right atrial chamber.

In parallel, in our patient a VT was induced. VT in TOF is the main cause of sudden death in this population. Reentry is the most common pathophysiologic mechanism due to surgical scars and patches as in RVOT infundibulum and VSD suture or patch.

In our patient, despite the poor hemodynamic VT tolerance, a fast and ultra-high density mapping catheter and the substrate depiction by the MRI helped us to depict the arrhythmogenic isthmus and to confirm conduction block across the line after ablation.

By the way, perfect concordance between the two compared maps cannot be demonstrated.

The major pitfall in the LGE-MRI reconstruction (focused on this case) could be referred to different times of acquisition (with consequent different volemic status) that can produce differences in dimension and shape of LGE-MRI reconstruction, especially for the right chambers, and to the wall thickness considered in the LGE-MRI could differ in the PSI model analysis from the endocardial electroanatomic mapping.

Lastly, LGE localization highlight possible zone of scar or border zone (not yet demonstrated in the right atrial chamber). Different nature of tissue (border zone, scar or “normal” myocardium) of the different zone could have different and various electrical characteristic.

However, we considered the LGE-MRI reconstruction very helpful during this procedure, considering the complex nature of the anatomy of the patients who underwent multiple cardiac interventions (such as surgical and interventional procedures).

5 | CONCLUSION

Congenital heart disease patients can present with both ventricular and atrial arrhythmias due to re-entry related with surgical sutures and fibrosis. Due to the complexity of the circuits and poor hemodynamic tolerance the use of imaging to understand the substrate before ablation and a high-density mapping system to perform detailed and fast maps can help us to increase the success of ablation in these patients.

CONFLICT OF INTEREST

I.RL. and L.M. have served as a consultant for Biosense-Webster, Boston Scientific, and Abbot Medical. Other authors have no conflict of interest to declare.

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