

EP CASE REPORT

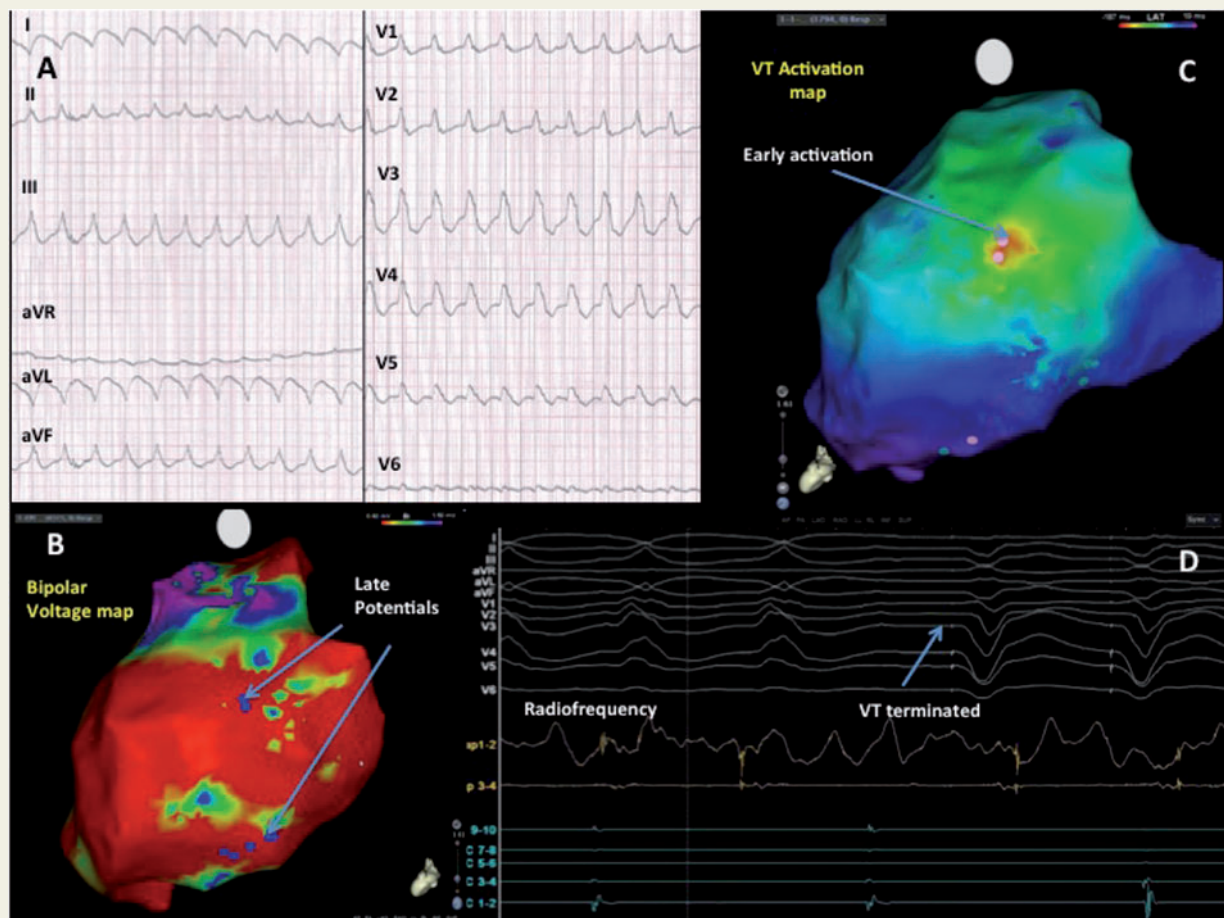
Epicardial ablation for ventricular tachycardia in Friedreich's ataxia cardiomyopathy

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A 55-year-old male patient with Friedreich's ataxia (FA), with advanced neurological involvement patient, was referred to our centre due to severe electrical storm caused by recurrent monomorphic ventricular tachycardia (MVT; *Panel A*). Four months earlier, he was admitted for an MVT episode of the same morphology which was treated with electrical cardioversion, amiodarone, and an implantable cardioverter-defibrillator (ICD). He was already on bisoprolol, sacubitril-valsartan, and spironolactone. In this new admission, tachycardia ablation was planned, as the patient continued receiving multiple shocks for new MVT recurrences despite mechanical ventilation, amines, and intra-aortic balloon pump. We decided to perform an epicardial approach to the tachycardia since an apical thrombus was seen on the echocardiogram and the QRS morphology in tachycardia suggested an epicardial origin (q wave in lead I, pseudo-delta wave >75 ms¹; *Panel A*). After gaining access to the pericardial space with the usual subxiphoid puncture technique, epicardial electroanatomic mapping with



CARTO 3[®] in sinus rhythm with a multi-electrode mapping catheter (PentaRay[®]) showed diffuse fibrosis and two areas of late potentials at basal infero-lateral and apical lateral wall of the left ventricle (*Panel B*, blue dots). During the study, the documented clinical MVT started spontaneously and a propagation map revealed presystolic potentials (>30 ms before QRS), but not epicardial mid-diastolic potentials, in the same basal infero-lateral site that showed late potentials in sinus rhythm (*Panel C*; see supplementary material online, *Video S1*). At this location, radiofrequency ablation with a 3.5-mm-cooled-tip catheter finished the VT (*Panel D*). Subsequently, ablation was extended, eliminating all late potentials. Finally, a programmed ventricular pacing protocol with up to three extrastimuli was performed, without VT induction. Overall, it was most likely a focal tachycardia.

After the procedure, life-sustaining measures were withdrawn, and the patient was discharged a week later. Follow-up at 1 month revealed no new arrhythmic episodes on ICD interrogation. Cardiac involvement is the main cause of mortality in FA.² Arrhythmias are less common than in other inherited heart conditions.³ To the best of our knowledge, this is the first case of MVT treated successfully by epicardial ablation in a patient with FA.

[Supplementary material](#) is available at *Europace* online.

Conflict of interest: none declared.

References

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2. Tsou AY, Paulsen EK, Lagedrost SJ, Perlman SL, Mathews KD, Wilmot GR *et al*. Mortality in Friedreich ataxia. *J Neural Sci* 2011;**307**:46–9.
3. Weidemann F, Störk S, Liu D, Hu K, Herrmann S, Ertl G *et al*. Cardiomyopathy of Friedreich ataxia. *J Neurochem* 2013;**126**:88–93.