

## EP CASE REPORT

# Unusual response to ajmaline test in Brugada syndrome patient leads to extracorporeal membrane oxygenator support

Giuseppe Ciconte, Michelle M. Monasky, Gabriele Vicedomini, Valeria Borrelli, Luigi Giannelli, and Carlo Pappone \*

Department of Arrhythmology, IRCCS Policlinico San Donato, Piazza E. Malan 1, 20097 San Donato Milanese, Milano, Italy

\* Corresponding author. Tel: +39 02 5277 4260; fax: +39 02 5277 4306. E-mail address: carlo.pappone@af-ablation.org

A 41-year-old man without family history of sudden death or Brugada syndrome (BrS) experienced aborted cardiac arrest due to ventricular fibrillation (VF) while resting. Coronary angiography revealed a severe stenosis of the proximal left anterior descending artery, successfully treated by percutaneous coronary intervention. While in intensive care, VF recurred in the absence of coronary abnormalities, as demonstrated by a repeated angiogram. A subcutaneous defibrillator was implanted.

Months later, VF relapsed and myocardial ischaemia was ruled out by coronary angiogram. Ajmaline challenge revealed BrS ([Supplementary material online, Figure S1](#)), but the patient refused to undergo genetic testing.

Right ventricular epicardial ablation<sup>1</sup> was performed. Ajmaline provoked a Type 1 pattern with 0.7 mg/kg. Immediately after electrophysiological mapping, the QRS complex suddenly widened and a rapidly progressive ST elevation developed in almost all leads ([Supplementary material online, Figure S2A–D](#)). Then, a premature ventricular ectopic beat triggered the initiation of a monomorphic ventricular tachycardia degenerating into VF ([Supplementary material online, Figure S2E](#)), resistant to external cardioversion ([Supplementary material online, Figure S2F](#)). Cardiopulmonary resuscitation (CPR) was immediately started, along with the infusion of a high-dose of isoproterenol. After approximately 15 min, VF self-terminated with a progressive and complete recovery ([Supplementary material online, Figure S3](#)).

Since clinical conditions were stable, mapping was completed ([Supplementary material online, Figure S4](#)) and ablation performed. To evaluate residual areas of abnormal electrograms, post-ablation mapping was performed without ajmaline because of concerns related to the previous spontaneous ventricular arrhythmia (VA). During mapping, a huge ST-segment elevation developed in most leads, followed a few minutes later by spontaneous initiation of VF ([Figure 1A](#)) refractory to external cardioversion ([Figure 1B](#)). Along with CPR and isoproterenol infusion, veno-arterial extracorporeal membrane oxygenator (ECMO) was placed for cardiovascular support, with a flow of 5.5 L/min, resulting in prompt haemodynamic improvement. After 21 min, a DC shock effectively restored sinus rhythm, and ST-segment elevation slowly reduced. A coronary angiography showed, besides intra-stent restenosis, a slight focal coronary spasm ([Supplementary material](#)



**Figure 1** Second refractory ventricular tachycardia/fibrillation. (A) ST-segment elevation development in most leads, followed by the spontaneous initiation of ventricular fibrillation, which was (B) again refractory to external cardioversion.

online, [Figure S5](#)). Within hours, the clinical condition improved, ECMO assistance was gradually weaned and then removed, and the patient completely recovered clinically, although negative T waves were observed, as a consequence of myocardial ischaemia ([Supplementary material online, Figure S6](#)).

Sustained VAs during ajmaline challenge occurs in roughly 2% of patients.<sup>2</sup> Moreover, catheter manipulation may mechanically induce ectopic beats, which can trigger arrhythmias. Possibly, myocardial ischaemia and the presence of intra-stent restenosis<sup>3</sup> may have played a role in such an adverse reaction. All these factors, alone or in combination, may have contributed to refractory VF.

In patients with BrS, coronary artery disease might further increase the risk of VAs. Therefore, sodium channel blockers should be managed very carefully in this subset of patients, even though they represent the ideal drug to fully unmask the BrS arrhythmogenic substrate. Extended intra- and periprocedural ECMO support is important in cases of arrhythmic emergencies as a bridge to recovery.

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

**Conflict of interest:** none declared.

## References

1. Pappone C, Brugada J, Vicedomini G, Ciconte G, Manguso F, Saviano M et al. Electrical substrate elimination in 135 consecutive patients with Brugada syndrome. *Circ Arrhythm Electrophysiol* 2017;**10**:e005053.
2. Conte G, Sieira J, Sarkozy A, de Asmundis C, Di Giovanni G, Chierchia GB et al. Life-threatening ventricular arrhythmias during ajmaline challenge in patients with Brugada syndrome: incidence, clinical features, and prognosis. *Heart Rhythm* 2013;**10**:1869–74.
3. Spar DS, Silver ES, Hordof AJ, Torres A, Liberman L. Coronary artery spasm during radiofrequency ablation of a left lateral accessory pathway. *Pediatr Cardiol* 2010;**31**:724–7.