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**Purkinje-related ventricular fibrillation associated with a homozygous H558R polymorphism in the sodium channel SCN5A gene**

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A 36-year-old patient was admitted after successful resuscitation due to ventricular fibrillation. The patient recovered without neurological sequelae. Cardiological examination including QTc interval, ajmaline testing, echocardiography, coronary angiography, and cardiac magnetic resonance imaging revealed no abnormal findings.

The patient received a dual-chamber ICD and was discharged on bisoprolol therapy. After 6 weeks, he was re-admitted due to recurrent ICD shocks for ventricular fibrillation. Frequent monomorphic premature ventricular complexes (PVCs) were recorded on Holter ECG. Recurrent torsades de pointe tachycardia initiated by PVCs with the same QRS morphology (arrows) led to almost daily ICD shocks despite DDDR-70 pacing (Figure 1A). Ablation targeting the earliest electrical activity in the distal Purkinje network during the PVCs (arrow) in the left ventricular septum eliminated the monomorphic PVCs (Figure 1B) and no further ventricular tachyarrhythmias occurred during the 6-month follow-up. Genetic evaluation revealed the polymorphism c.1673A > G (H558R) in the sodium channel α-subunit gene (SCN5A) in a homozygous form.

Successful ablation of monomorphic Purkinje-related PVCs as a focal trigger eliminated recurrent torsades de pointes tachycardias in a patient with the common H558R polymorphism in homozygous form. The case highlights the role of otherwise ‘benign’ frequent PVCs in the context of a relatively common inherited arrhythmogenic substrate.

The full-length version of this report can be viewed at: http://www.escardio.org/Guidelines-&-Education/E%E2%80%93learning/Clinical-cases/Electrophysiology/EP-Case-Reports.

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