A 36-year-old patient equipped with an implantable cardioverter-defibrillator had frequent nocturnal episodes of torsades de pointe tachycardia initiated by monomorphic premature ventricular complexes. After successful ablation of the initiating Purkinje-related ventricular extrasystoles, the patient had no recurrence of ventricular fibrillation. Genetic evaluation revealed the common polymorphism H558R in the SCN5A gene in homozygous form.

A 36-year-old patient of Egyptian origin was admitted to our institution after successful out-of-hospital resuscitation due to ventricular fibrillation. During the previous months, he had undergone cardiological and neurological examinations due to recurrent syncope in another hospital without abnormal findings. A sister had died due to sudden unexpected death at the age of 2. The QTc interval, determined once daily, was normal (QTc 420 ms) except during the 24 h interval of therapeutic hypothermia (33 °C) immediately after the cardiac resuscitation (QTc 530 ms). The patient recovered without neurological sequelae. Non-invasive and invasive cardiological examination including echocardiography, coronary angiography, and cardiac magnetic resonance imaging revealed no abnormal findings. Ajmaline testing was negative, and there were no criteria on the surface ECG that orient for an Early repolarization Syndrome. During Holter ECG recordings, mean heart rate was 65/min (minimum 50/min, maximum 102/min). During bicycle exercise, there was an adequate frequency-dependent decrease of the QT interval from 380 ms at baseline (heart rate 75/min, QTc 425 ms) to 290 ms at 120 W (heart rate 121/min, QTc 412 ms).

The patient received a dual-chamber implantable cardioverter-defibrillator (ICD) and was discharged from the hospital on β-blocker therapy (bisoprolol 10 mg daily), potassium and magnesium. After 6 weeks, he was re-admitted due to recurrent adequate ICD shocks for ventricular fibrillation. Frequent monomorphic premature ventricular complexes (PVCs), partially as 2:1 ventricular extrasystoles, with a total of ~10 000 monomorphic PVCs per 24 h, were recorded on Holter ECG (Figure 1A). Recurrent nocturnal torsades de pointe tachycardia initiated by PVCs with the same QRS morphology, without QT prolongation during sinus rhythm, led to almost daily ICD shocks despite DDDR-70 pacing and continued bisoprolol therapy (Figure 1B). 12-Lead ECGs showed frequent monomorphic PVCs with a bifascicular block morphology with left superior axis and right bundle branch block configuration and a relatively narrow QRS complex (QRS 130 ms) (Figure 1C) suggesting an origin of the PVCs in the Purkinje network of the left posterior fascicle. Using programmed right ventricular stimulation, no ventricular tachyarrhythmias were inducible. Ablation using a standard irrigated ablation catheter targeting the earliest electrical activity in the distal Purkinje network during the PVCs in the left ventricular septum preceding the QRS complexes by ~40 ms (30 W, total 400 s radiofrequency energy) eliminated the monomorphic PVCs (Figure 1D, E) and no further torsades de pointe tachycardia occurred. During the 6-month follow-up, the patient remained free of further ventricular tachyarrhythmias and ICD interventions. Genetic evaluation had revealed the polymorphism c.1673A > G (H558R) in the sodium channel α-subunit gene (SCN5A) in a homozygous form. No other variants in the genes KCNQ1 KCNH2, KCNE1, KCNE2, and SCN5A were detected. The DPP-6 variant (c.340C > T) was excluded by genetic testing. Genetic evaluation and cardiological screening of family members were not available.

Recent reports point out to focal ablation of triggering beats from the Purkinje system as a potentially valuable tool in elimination of malignant arrhythmias in patients with idiopathic ventricular fibrillation and long-QT syndrome. The polymorphism H558R in the SCN5A gene involving a substitution of histidine (H) with arginine (R) is present with a reported homozygous frequency of 5–7% in the population. A recent report has suggested that the polymorphism H558R is associated with arrhythmic storm developing during myocardial infarction. We describe the elimination of recurrent torsades de pointe tachycardias by successful ablation of monomorphic Purkinje-related PVCs as a focal trigger 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.
Conflict of interest: none declared.

References

Figure 1  (A) Holter ECG showing frequent monomorphic PVCs as 2:1 ventricular extrasystoles. (B) Initiation of torsades de pointe tachycardia by a ventricular extrasystole with the same QRS morphology and termination by the ICD. (C) 12-Lead ECG showing a typical PVC during sinus rhythm with left superior axis, right bundle branch block configuration, and a relatively narrow QRS complex (130 ms) suggesting an origin in the Purkinje system of the left posterior fascicle. Sinus rhythm cycle length 840 ms, PR 180 ms, QRS 90 ms, QT 380 ms, and QTc 414 ms. (D) Intracardiac recordings from the ablation catheter in the left ventricular septum (ABL), the high right atrium (HRA), the right ventricular apex (RV), and surface leads aVF, V1, and V6. The Purkinje potential preceding the ventricular potential at the successful ablation site is indicated by arrow. (E) Fluoroscopic projection of the electrode catheters in a left anterior oblique projection. CS, coronary sinus.