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CASE REPORT

Use of a subcutaneous ICD in a patient with short QT syndrome

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Key clinical message

Short QT syndrome carries the risk for inappropriate therapies using transvenous ICD because of overdetection of the ample T wave. SQT syndrome may also benefit from subcutaneous ICD, although additional cases are needed to affirm the safety of such device in this setting.

Keywords

Short QT syndrome, subcutaneous ICD.

Introduction

Subcutaneous ICD may be proposed to patients with channelopathies presenting with significant risk for sudden cardiac death. Short QT syndrome is a rare channelopathy carrying the risk for inappropriate therapies with current transvenous ICD because of overdetection of the ample T wave. If subcutaneous ICD are safe in this setting is unknown. We report on the uneventful follow-up after implantation of a subcutaneous ICD in a patient with a short QT syndrome.

Case presentation

Because patients with short QT syndrome (SQT) may have tall peak T waves with short QRS-T intervals, inappropriate ICD therapies may occur due to the possibility of double counting of both the QRS and the T wave [1], leading to a succession of short cycle lengths, as realized for example during exertion. Inappropriate ICD therapies have been described earlier in SQT patients with transvenous ICD, which should be prevented by tailoring the detection window characteristics [2]. If such an hazard may happen with subcutaneous ICD – whose ability in avoiding T wave detection is crucial – is unknown, since no SQT case seems to have been reported with such a device as yet.

A 30 year-old man with SQT syndrome implanted for 9 years with a transvenous ICD for primary prevention (familial sudden death due to SQT and induction of ventricular fibrillation) was explanted because of battery depletion and lead failure. His ECG showed ample T waves with 280 ms QT interval (corrected QT 330 ms). We decided to reimplant the ICD, using a subcutaneous ICD (S-ICDTM System, Boston Scientific, St. Paul, MN, USA). After assuring an acceptable R/T ratio using the screening ruler (Fig. 1), the pulse generator was placed subcutaneously in the anterior axillary line with the electrode inserted subcutaneously parallel to the sternal midline. Ventricular fibrillation was induced and terminated by a 65 J shock (15 J safety margin).

Testing of the device at baseline, during treadmill or during the follow-up never displayed T wave over sensing in any detection configuration ("primary" between the pulse generator and the proximal electrode, "secondary" between the generator and distal electrode, or "alternate" between both electrodes). The device adequately sensed the R wave (Fig. 2) even if there are a few missed R waves at exertion in some configuration (arrows), especially when the R wave amplitude slightly decreases (the subcutaneous ICD uses a two peak average to adjust the detection profile dynamically, but in the presence of varying amplitudes such as during exertion, a few beats may be missed while

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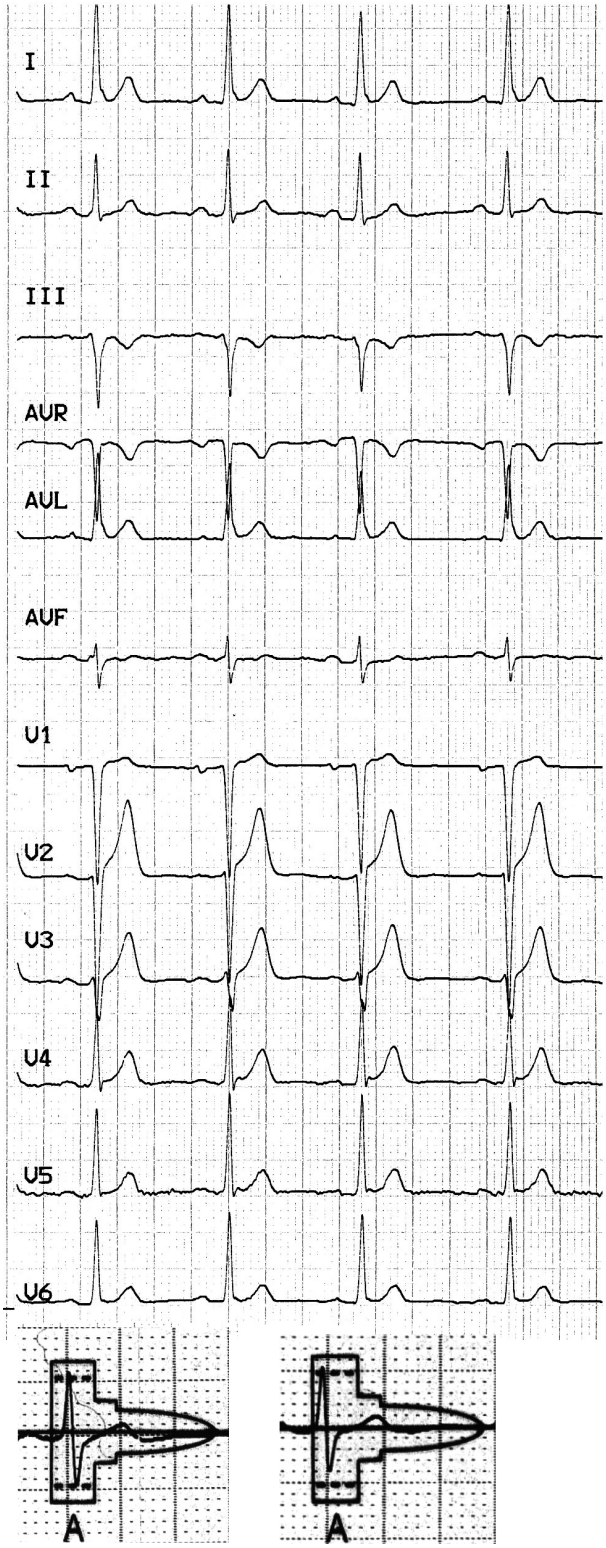

Figure 1. 12-lead ECG of the patient with short QT. Below are shown the acceptable R/T ratio using the screening ruler both at implant (left) and at 1 year (right).

adjusting to the underlying amplitudes). The latency sometimes observed after the R wave on markers (red circle) in the alternate configuration is an artifact of programmer telemetry's ability to process increased marker activity, for example, exertion scenario at follow-up (such a behavior is not observed in the ambulatory setting).

At 1 year follow-up, the R wave was 1.1 mV while T wave was 0.2 mV on ECG (R/T ratio = 5.5) (fig). The R/T ratio was higher at implant during treadmill and during the follow-up in the primary configuration (Table 1).

The patient was discharged in the primary configuration and no appropriate or inappropriate detection (Fig. 2) or therapy happened over a follow-up of 1 year.

Discussion

Short QT syndrome is a very exceptional channelopathy carrying a significant risk of sudden cardiac death due to malignant ventricular arrhythmias [3]. SQT is caused by an excessive abbreviation of repolarization duration, mainly due to some inherited gain of function in potassium channels [3]. SQT is diagnosed if corrected QT is ≤330 ms or 360 ms together with clinical events [4]. Risk stratification to date only refers to spontaneous clinical events such as unexplained syncope or a previous cardiac arrest [4]. ICD is the only recommended therapy, although medical treatment with quinidine may be used in selected cases [4].

Patients with SQT syndrome often present with tall peak T waves together with the short QT interval [1]. Thus, inappropriate ICD therapies have been described, using transvenous ICD because of double counting of both the QRS and the T wave [1], especially during exertion. Of note, this may be prevented by tailoring the detection window characteristics [2]. If such hazard may happen with subcutaneous ICD – whose ability in avoiding T wave detection is crucial – is unknown, since no SQT implanted with subcutaneous ICD seems to have been reported as yet. This case highlights the fact that patients with SQT syndrome may also benefit from subcutaneous ICD.

Conclusion

Due to morbidity and complications inherent to transvenous ICDs, SQT syndrome may also benefit from subcutaneous ICD. However, additional cases are needed to affirm the safety of such device in this setting, especially in patients with ampler T waves.

Conflict of Interest

None declared.

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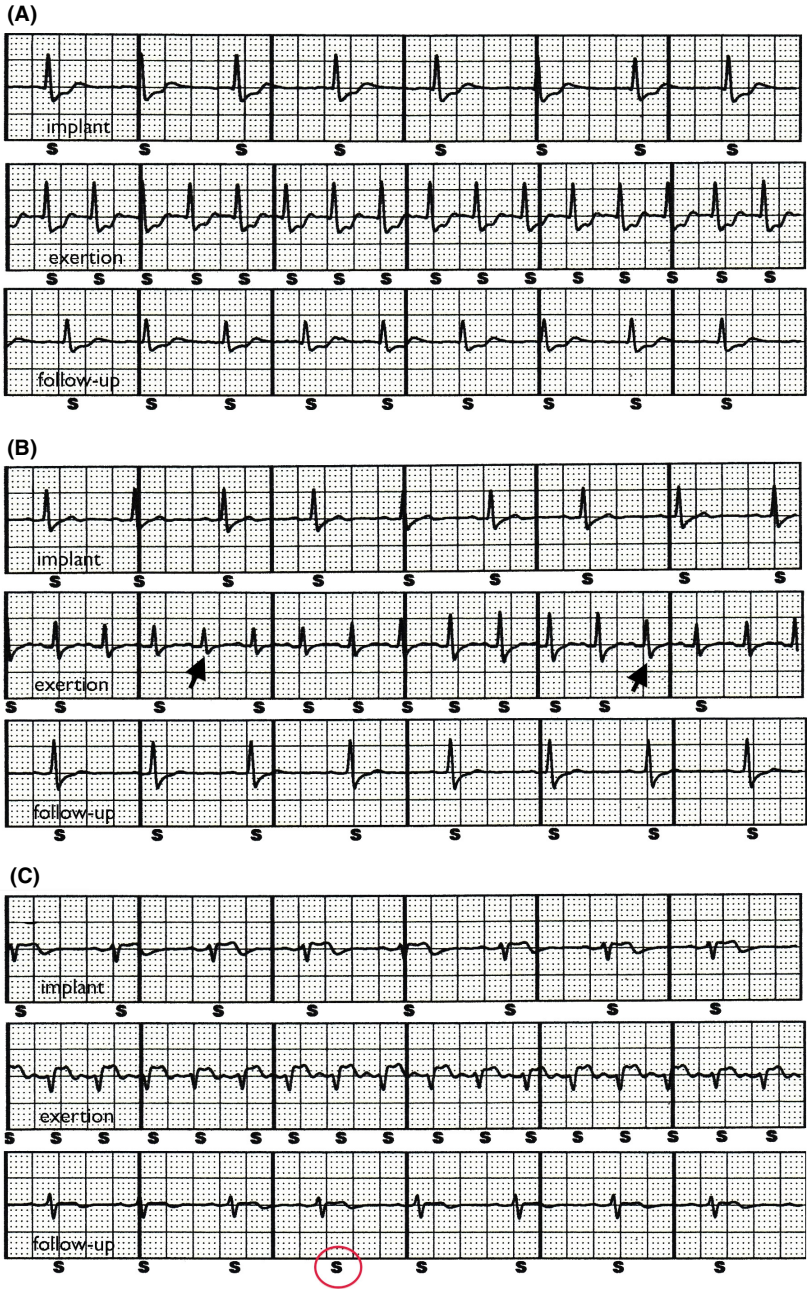

Figure 2. Programmer telemetry recordings in the primary (A), secondary (B) and alternate (C) detection configurations at implant, during exertion and at rest at 1 year follow-up. S are the markers of the QRS complexes as sensed by the device (see text for detailed explanation).

Table 1. Amplitude and QRS: T-wave Ratio measurements from three time points: Implant, Exertion and Follow-up.

	Implant			Exertion			1-yr Follow-up		
	R(mV)	T(mV)	R/T ratio	R(mV)	T(mV)	R/T ratio	R(mV)	T(mV)	R/T ratio
Primary	2.2	0.4	5.5	2.5	0.2	12.5	1.4	0.35	4.0
Secondary	2.2	0.2	11	1.1	0.2	5.5	2.2	0.2	11
Alternate	0.72	0.4	1.8	0.72	0.2	3.6	0.72	0.2	3.6

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