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CASE REPORT

Short QT syndrome presenting as syncope: How short is too short?

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KEYWORDS
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Abstract We report the case of a 52-year-old man who presented to our emergency department (ED) after three episodes of syncope in the seven hours before admission. During his stay in the ED he had recurrent ventricular tachycardia (VT) requiring external electrical cardioversion. A 12-lead electrocardiogram (ECG) showed a short QT (SQT) interval (270 ms, QTc 327 ms), with frequent R-on-T extrasystoles triggering sustained polymorphic VT. After exclusion of other precipitating causes, the patient was diagnosed as having SQT syndrome (SQTS) according to the Gollob criteria. To our knowledge, this is the first known documentation of an SQT-caused arrhythmic episode on a 12-lead ECG, as well as the first reported case of SQTS in Portugal. The patient received an implantable cardioverter-defibrillator and was discharged. At a follow-up assessment 14 months later he was symptom-free, interrogation of the device showed no arrhythmic events, and the ECG showed a QT interval of 320 ms (QTc 347 ms).

PALAVRAS-CHAVE
Síncope;
Síndrome QT curto;
Morte súbita cardíaca;
Arritmia ventricular

Quando o intervalo QT é curto demais: síncope causada por síndrome QT Curto

Resumo Os autores apresentam o caso de um homem com 52 anos que recorreu ao Serviço de Urgência (SU) por três episódios sincopais ocorridos nas últimas sete horas pré-admissão hospitalar. Durante a estadia no SU, objetivaram-se períodos de taquicardia ventricular (TV) recorrente, com necessidade de cardioversão elétrica externa, tendo o doente sido transferido para a unidade de cuidados intensivos cardíacos para monitorização e orientação clínica. O electrocardiograma de 12 derivações (ECG) registado no contexto de um dos episódios revelava um intervalo QT curto (260 ms, QTc 327 ms) em ritmo sinusal, com fenómeno de R-on-T desencadeando TV polimórfica mantida. Após exclusão de causas precipitantes, o doente foi

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classificado como tendo Síndrome QT Curto (SQTC), de acordo com os critérios de Gollob. Tanto quanto é do nosso conhecimento, esta é a primeira documentação no ECG de um episódio arrítmico causado por SQTC e a primeira descrição de um caso de SQTC em Portugal. Foi submetido a implantação de cardioversor-desfibrilhador implantável (CDI) monocameral e teve alta hospitalar. No seguimento aos 14 meses pós-implantação, o doente negava recorrência da sintomatologia, a interrogação do CDI não mostrou novos episódios arrítmicos, e no ECG persistia um intervalo QT de 320 ms (QTc 347 ms).

List of abbreviations

ECG	electrocardiogram
ED	emergency department
ICD	implantable cardioverter-defibrillator
PVC	premature ventricular contractions
PVT	polymorphic ventricular tachycardia
QTc	corrected QT
SQT	short QT
SQTS	short QT syndrome
VT	ventricular tachycardia

Case report

A 52-year-old man presented to our emergency department (ED) due to three episodes of sudden transient loss of consciousness in the seven hours before hospital admission. The first event occurred at 2:00 am, while the patient was lying in bed, recurring at 4:00 am and 7:00 am. He reported malaise, but denied prodromes, chest pain, palpitations or dyspnea. His wife, who witnessed the episodes, described them as lasting for about five minutes, with spontaneous and complete recovery. There was no evidence of seizure-like activity.

The patient had no history of known cardiovascular disease and no recollection of previous episodes of syncope. His father died at 38 years of age due to sudden death of undetermined origin, with no other relevant family history. His medical history was notable for depression (managed at an outpatient clinic), duodenal peptic ulcer (five years previously), and hypercholesterolemia. His regular medication consisted of mirtazapine 30 mg daily, alprazolam 0.5 mg daily, bupropion 300 mg daily and fenofibrate 200 mg daily. He had no clinical allergies and denied smoking, alcohol intake or any kind of addiction.

Physical examination showed a well-nourished man with no external signs of distress. At admission, he was afebrile, with blood pressure of 112/65 mmHg, heart rate of 76 bpm and oxygen saturation of 98% by pulse oximetry. Cardiac examination showed an irregular heart rhythm with no murmurs. The rest of the physical examination was unremarkable.

During his assessment in the ED, he suffered a new episode of syncope with spontaneous recovery, and cardiac monitoring during the event showed a polymorphic ventricular tachycardia (PVT). Despite intravenous infusion of amiodarone and magnesium sulphate, there was recurrence of the cardiac arrhythmia requiring a total of five external electrical cardioversions. A 12-lead electrocardiogram (ECG) recorded during one such event is presented in Figure 1.

A blood chemistry panel showed normal serum potassium and magnesium, with no hypocalcemia. Cardiac biomarkers were negative, and other laboratory investigations were unremarkable. The patient was transferred to our cardiac intensive care unit for further assessment and treatment.

Careful review of ECG records (Figures 1 and 2) showed a short QT interval (280 ms, QTc 329 ms according to the Bazett formula). Given the setting of a PVT storm coronary angiography was performed, which ruled out coronary artery disease or anomalous coronary origin (Figures 3 and 4), with normal wall motion and left ventricular ejection fraction (70%). Transthoracic echocardiographic examination showed a normal heart with good left ventricular systolic function and no signs of structural disease.

In the absence of other causes, a presumptive diagnosis of short QT syndrome (SQTS) was made, and quinidine tablets were requested from the hospital pharmacy. The patient was classified as high-probability SQTS according to the Gollob criteria,[1] with a score of 5 (Table 1).

In-patient 24-hour Holter monitoring was performed and showed infrequent dimorphic premature ventricular contractions (PVCs), with no repetitive patterns. An electrophysiologic study was performed and was notable for the induction of self-limited PVT with hemodynamic collapse (induction protocol: S1 400/S2 200 ms, pacing at the right ventricular apex). The measured right ventricular refractory period was shortened (200 ms). No attempt was made to induce atrial fibrillation. The measured atrial refractory period was 350 ms in the right atrium and 200 ms in the left atrium (as measured in the distal coronary sinus).

In accordance with the ACCF/AHA/HRS guidelines for device-based therapy,[2] the patient received an implantable cardioverter-defibrillator (ICD) as secondary prevention indicated for SQTS, repeated syncopes and inducible PVT. There were no procedural complications and he was discharged on his previous medication and quinidine.

At a follow-up visit 14 months later, the patient was feeling well and had had no new episodes of syncope or palpitations. Physical examination was unremarkable. ICD device interrogation showed no shocks or arrhythmic events,