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CASE REPORT
Arrhythmias/electrophysiology

Case report of a patient with congenital long QT syndrome Type 2 presenting with electrical storm: do not judge a book by its cover!

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Background Patients with congenital long QT syndrome (LQTS) are at high risk for sudden cardiac death (SCD). Although several triggers can provoke ventricular fibrillation (VF) in patients suffering from LQTS acquired heart disease in addition to LQTS should not be overlooked.

Case summary We present a case of a 47-year-old female patient who was diagnosed with congenital LQTS Type 2 at the age of 23 after surviving SCD. At that time, she underwent implantable cardioverter-defibrillator (ICD) implantation and was free of events for 24 years. Recently, the patient was referred to our institution after suffering from an ICD shock during sleep. Upon arrival she developed electrical storm and received overall six ICD shocks. The initial electrocardiogram (ECG) showed atrially triggered ventricular pacing. However, distinct ST segment elevations in the inferior leads could be observed. Thus, coronary angiography was immediately performed and showed subtotal occlusion of the right coronary artery, which was treated by drug-eluting stent implantation. Atrioventricular conduction immediately resumed after revascularization and the following non-paced ECG revealed a prolonged QT interval. Laboratory measurements confirmed acute myocardial infarction with elevated cardiac enzymes. The patient was put on betablockers, dual antiplatelet therapy, and statins and discharged in good condition.

Discussion This case report highlights that diagnostic work-up in patients with LQTS presenting with VF should always include the search for additional acquired heart disease such as myocardial infarction, as a potential trigger for electrical storm. Moreover, signs of ischaemia can be discerned even in a paced ECG which should lead to immediate cardiac catheterization.

Keywords Cardiovascular disease • Case report • Electrical storm • Long QT syndrome • Myocardial infarction

ESC Curriculum 3.2 Acute coronary syndrome • 3.4 Coronary angiography • 5.6 Ventricular arrhythmia • 5.8 Cardiac ion channel dysfunction • 5.10 Implantable cardioverter defibrillators

Learning points

- Diagnostic investigation in patients with an inherited primary arrhythmia syndrome presenting with ventricular tachyarrhythmias should always include the search for additional acquired heart disease.
- Signs of ischaemia can be discerned even in a paced electrocardiogram, which should lead to immediate cardiac catheterization and revascularization if necessary.

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Introduction

QT interval prolongation has been found to be associated with sudden cardiac death (SCD) more than five decades ago.[1] Later, long QT syndrome (LQTS) has been identified as an inherited primary arrhythmia syndrome caused by mutations of ion channels that are responsible for cardiac repolarisation.[2] Its therapy usually includes the avoidance of QT-prolonging drugs and genotype-specific triggers for arrhythmias, betablocker treatment, and the implantation of an implantable cardioverter-defibrillator (ICD) in patients at high risk for SCD.[3] Genotype-specific triggers have since been identified to cause Torsades de Pointes tachycardias and ventricular fibrillation (VF).[4,5]

Myocardial ischaemia constitutes a common aetiology for ventricular arrhythmias. As patients with LQTS are not resilient to suffer from electrical storm associated with myocardial ischaemia, acquired heart disease in addition to LQTS should not be overlooked as highlighted in the case presented.

Timeline

Date	Event	Treatment
October 1996	Circulatory arrest due to VF	Successful resuscitation by defibrillation. Genetic testing revealed LQTS Type 2. Thus, an ICD was implanted
November 2010 and December 2017	Battery depletion of the ICD	Battery replacement
June 2021	ICD shock during sleep. Occurrence of electrical storm with overall 6 ICD shocks in the emergency department. The paced electrocardiogram (ECG) revealed inferior wall ST-elevation	Infusion of magnesium sulfate intravenously. Coronary angiography revealed subtotal occlusion of the right coronary artery which was successfully treated by drug-eluting stent implantation

Case summary

This case report is about a Caucasian woman who was treated for the first time in our institution in October 1996 at the age of 23 years. At that time, she suffered from a circulatory arrest in the presence of her boyfriend who immediately started cardiopulmonary resuscitation. Ventricular fibrillation was detected which was successfully terminated by defibrillation. The patient survived this episode without sequelae. The QT interval was found to be significantly prolonged (QTc 640 ms) in the ECG and, thus, LQTS was diagnosed. Subsequently, a dual-chamber ICD was implanted and betablocker treatment was initiated. Genetic testing revealed LQTS Type 2 (amino acid deletion in the subunit of the I_{Kr}-channel, which was not further specified at that time). Moreover, the patient's father and sister were genetically diagnosed for LQTS, too, but they had never experienced any cardiac events. The ICD battery was replaced twice in November 2010 and in December 2017 but no episodes of ventricular tachyarrhythmias were detected. After 24 years free of events, the patient, at the age of 47 years, was again referred to our emergency department in June 2021 after experiencing an ICD shock during sleep. Upon arrival, she presented with an electrical storm necessitating six ICD shocks for recurrent VF (*Figure 1*). Magnesium sulfate (2 g) was administered intravenously, which successfully suppressed the VF. Moreover, 10 mg of diazepam was infused to suppress anxiety. The patient felt no typical angina pectoris but rather some paraesthesia in both arms and the chest after the ICD discharges. Cardiovascular examination did not reveal any remarkable findings. Her heart rate was 90 beats/min and blood pressure 105/66 mmHg during sinus rhythm. The patient reported hypothyroidism and depressive episodes as accompanying diseases. Medication at admission included thyroid hormone replacement (75 µg levothyroxine once a day) and metoprolol (47.5 mg twice a day). The patient had a history of smoking (25 pack-years) but no other cardiovascular risk factors. The body mass index was 26 kg/m^2. The initial ECG showed atrially triggered ventricular pacing. However, despite pacing-related alterations of the QRS complex, distinct ST segment elevations were noted in the inferior leads (*Figure 2*). The patient was treated with 250 mg aspirin and 5000 I.U. heparin intravenously and primary coronary angiography was immediately performed. The angiography revealed coronary artery disease with a short-segment low-grade stenosis (20%, Type A lesion) in the middle third of the left anterior descending artery, an ostial short-segment high-grade stenosis (90%, Type B lesion) of a diagonal branch (*Figure 3A*), and a 10 mm long subtotal occlusion (99%, Type A lesion) of the middle right coronary artery (*Figure 3B*). The right coronary artery was treated with balloon dilatation followed by drug-eluting stent implantation (*Figure 3C*) resulting in successful elimination of the culprit lesion (*Figure 3D*). A loading dose of 180 mg ticagrelor was administered orally during the intervention and, additionally, glycoprotein IIb/IIIa inhibitors were infused. Pre-treatment with ticagrelor before coronary angiography was waived due to inconclusive data and the potential need for urgent surgical revascularization.[6]

Immediately after revascularization, normal atrioventricular conduction resumed (*Figure 4*). The following non-paced ECG revealed a prolonged QT interval (QTc 581–628 ms using Bazett's formula; *Figure 4*). Laboratory measurements confirmed an acute myocardial infarction (MI) with elevated cardiac enzymes (troponin initial 0.030 ng/mL, peak 2.170 ng/mL, upper limit of normal <0.015 ng/mL) and creatine kinase (initial 119 U/L, peak 2535 U/L, upper limit of normal <145 U/L). Blood lipids were not elevated (cholesterol 154 mg/dL, low density lipoprotein 58 mg/dL; Lipoprotein(a) 5.4 nmol/L). The HbA1c value was in the normal range (5.0%, upper limit of normal <5.9%). Echocardiography 2 days later showed a preserved right and left ventricular ejection fraction (60%) and no discernible wall motion abnormalities. Dual antiplatelet therapy (aspirin 100 mg daily and ticagrelor 90 mg twice a day) and a daily 40 mg dose of atorvastatin were administered. The pre-existing medications metoprolol (47.5 mg twice daily) and thyroid hormone (75 µg once daily) were continued. The patient was discharged in good condition a few days after the initial event, and she has remained free of angina and arrhythmias for 5 months afterwards.

Discussion

This is an unusual case of a patient presenting with an electrical storm that has apparently been caused by a combination of different triggers. Taking the patient's history of congenital LQTS Type 2 with aborted SCD in early adulthood into account, an electrical storm due to impaired ventricular repolarization may be considered the most likely

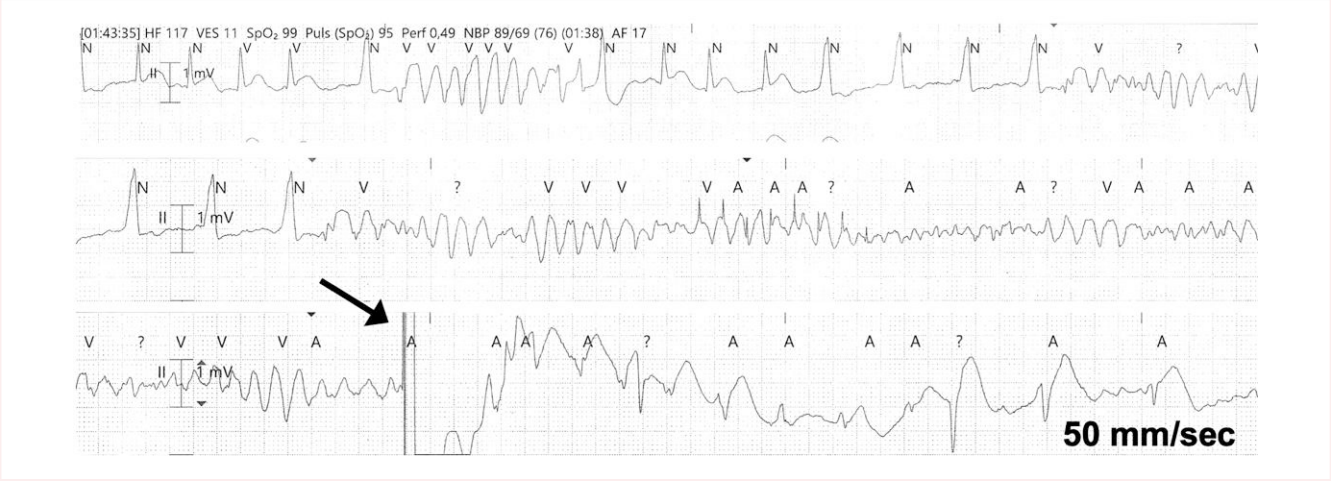


Figure 1 Extract from the rhythm monitoring of the 47-year-old patient immediately after admission to the emergency department. The rhythm strip shows polymorphous ventricular tachycardias similar to Torsades de Pointes tachycardias. The first two episodes were self-limiting. The third episode converted into ventricular fibrillation and was successfully terminated by the implantable cardioverter-defibrillator (arrow). In the following minutes, the patient developed electric storm and received overall six ICD shocks.

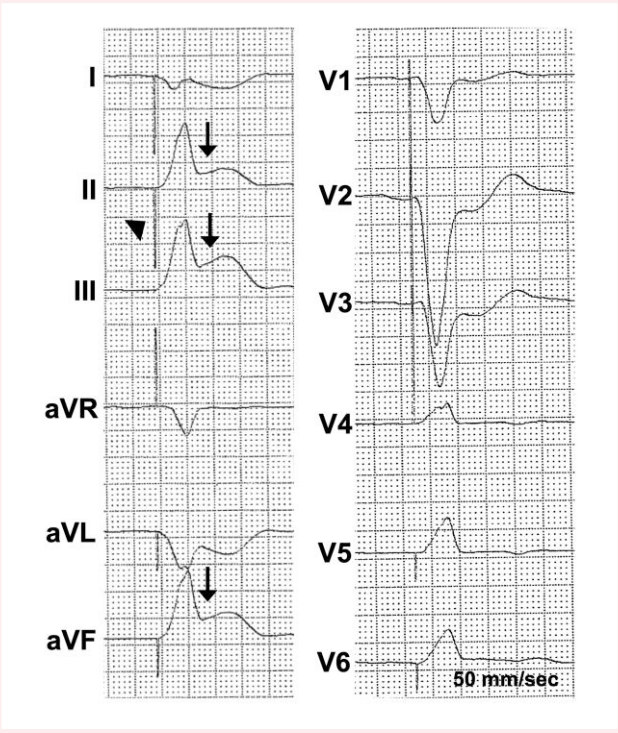


Figure 2 Initial 12-channel-electrocardiogram revealed atrially triggered ventricular pacing by the dual-chamber implantable cardioverter-defibrillator (arrow tip). Despite ventricular pacing distinct ST segment deviation concordant with QRS polarity was seen in the inferior Leads II, III, aVF (arrows).

primary diagnosis. However, this case report highlights that during diagnostic work-up, even in patients with confirmed LQTS presenting with VF, the possibility of additionally acquired heart disease should always be considered. Other causes for VF, e.g. MI, myocarditis, electrolyte imbalance, drugs, and electrical accident, must be taken into account for differential diagnosis. In fact, we diagnosed an inferior MI in this 47-year-old woman necessitating immediate coronary revascularization by percutaneous coronary intervention.

Several aspects in this case increase the likelihood for additionally acquired heart disease as an arrhythmia trigger: firstly, the sudden presentation with VF following a 24-year asymptomatic interval after the initial diagnosis of LQTS comes at an age at which acquired heart disease is more likely to occur. Secondly, the occurrence of VF during sleep is not a typical presentation in LQTS Type 2, which usually presents with VF upon emotional stress.[3,4] Thirdly, the presence of ventricular pacing was suggestive of some new conduction disturbance, which could later be well explained by ischaemically induced high-degree atrioventricular (AV)-block following occlusion of the right coronary artery that quickly resolved after revascularization. Finally, the initial ECG showed distinct ST segment elevation following the paced QRS complex indicating acute MI. The presence of ventricular conduction disturbances or a paced QRS complex may obscure the diagnosis of MI which may delay reperfusion therapy. Thus, based on the Sgarbossa criteria, algorithms to assess the likelihood of MI in patients with left bundle branch block (LBBB) have been developed[7–9] Systematic data for validating these algorithms in paced ECGs are scarce. However, some authors suggested that these criteria may also be helpful in patients with a right-ventricular paced ECG with a similar QRS morphology to LBBB.[10,11] The ventricular paced ECG in the case presented (*Figure 2*) is in accordance with the main criteria of the algorithms to diagnose MI.[7–9] Our report highlights the importance of a thorough diagnostic work-up in patients with VF to not miss any relevant comorbidities, even in the presence of a known underlying genetic disorder such as LQTS. In this case, the early diagnosis of a MI with ST-segment elevation allowed for rapid primary cardiac catheterization and revascularization of the culprit lesion. However, the decision process to take the patient to primary coronary catheterization was complex and required careful but urgent consideration of several findings as described above.

Moreover, several factors may have contributed to the occurrence of VF in this case. Firstly, QT prolongation has been found to be correlated with ventricular arrhythmias and in-hospital mortality in patients

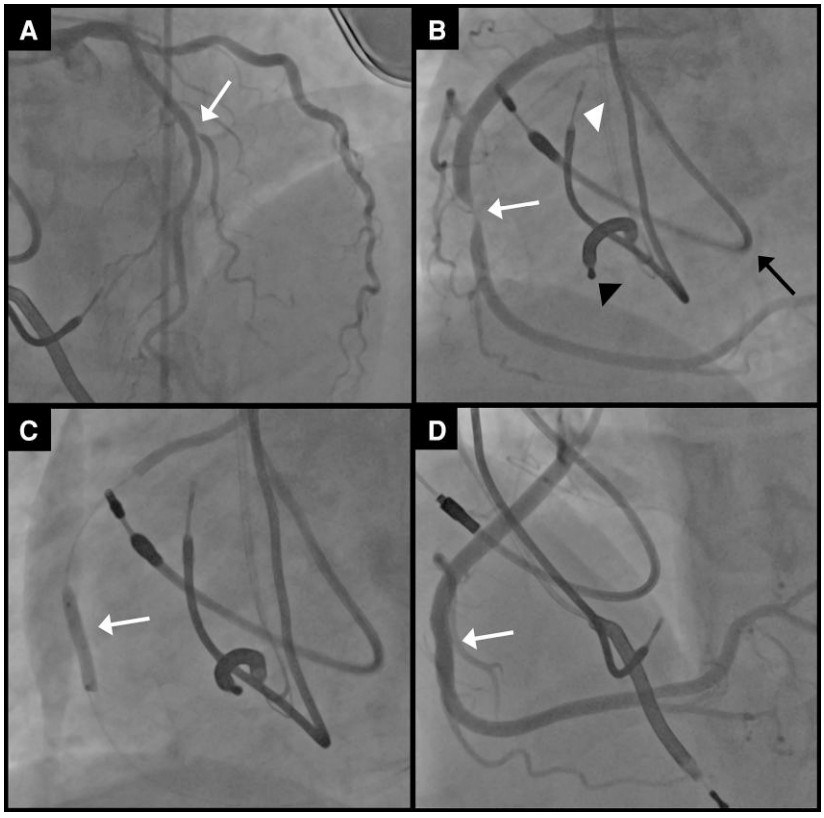


Figure 3 Sections from the coronary angiography (anterior–posterior projection with cranial angulation in (*A*), left anterior oblique projection in (*A–D*). (*A*) Angiography of the left coronary artery revealed a short-segment low-grade stenosis (20%, Type A lesion) in the middle third of the left anterior descending artery and an ostial short-segment high-grade stenosis (90%, Type B lesion) of a diagonal branch (white arrow). (*B*) 10 mm long subtotal occlusion (99%, Type A lesion) of the middle right coronary artery. The white arrowhead points the pace-sense lead fixed in the right-ventricular outflow tract (Membrane E, Pacesetter Systems Inc., Sylmar, CA, USA), the black arrowhead the dual-coil lead in the right-ventricular apex (Endotak C 0125, Cardiac Pacemakers Inc., St. Paul, MN, USA) and the black arrow the atrial lead (Type 4068, Medtronic, Minneapolis, MN, USA). (*C*) The right coronary artery was treated by pre-dilatation of the lesion and drug-eluting stent implantation (3.5 × 18 mm; white arrow). (*D*) The culprit lesion was reopened successfully (white arrow).

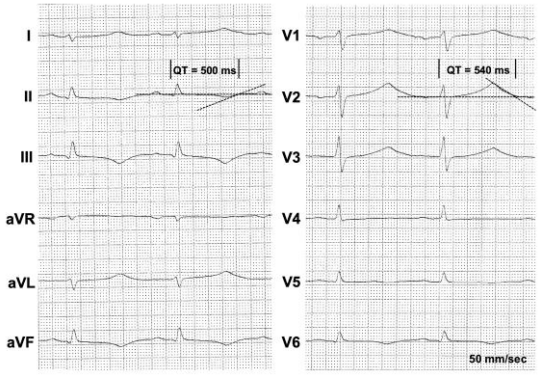


Figure 4 Twelve-channel-electrocardiogram registered immediately after revascularization. Spontaneous atrioventricular conduction resumed and ventricular pacing disappeared. Q-waves and T-wave-negations were observed in the inferior Leads II, III, and aVF and also in V6. The QT interval was substantially prolonged. QTc was calculated 581–628 ms using the Bazett formula.

with MI.[12] Thus, patients with inherited LQTS may have lower thresholds for VF in the presence of MI.[12] Secondly, female gender is associated with a higher rate of life-threatening cardiac events in a cohort of patients with LQTS Type 2.[13] Thirdly, bradycardia is known to precipitate VF in LQTS patients, which can be applied to our patient with bradycardia induced by ischaemic high-degree AV-block.[14]

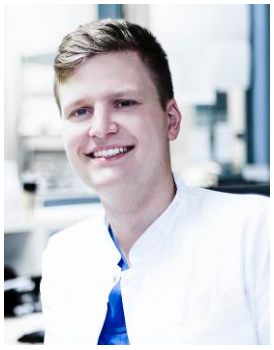
It is important to note, however, that the management of electrical storms with recurrent VF and ICD discharges is often challenging. In patients with LQTS, the intravenous administration of magnesium sulfate has been proven to be effective.[15] As indicated by this case, magnesium sulfate may also be effective in suppressing VF in patients with LQTS and an additional MI. In contrast, QT-prolonging antiarrhythmic drugs like amiodarone should be avoided in patients with confirmed LQTS to prevent deterioration of electrical storm.

Conclusion

The presented case highlights that in patients with an inherited primary arrhythmia syndrome and electrical storm one should always consider additionally acquired heart disease, such as an acute coronary syndrome, as a potential trigger for the electrical storm. Moreover, signs

of ischaemia may be discerned even in a ventricular paced ECG which should lead to immediate revascularization if necessary.

Lead author biography



Dennis Lawin achieved his licence to practice medicine at RTWH Aachen University, Germany. After internship at the university hospital of Bern (heart surgery), Switzerland, he finished his doctoral thesis at RWTH Aachen University, Germany. At present, he is a junior physician for cardiology and internal medicine at the University hospital OWL of Bielefeld University, Campus Klinikum Bielefeld, Germany, and also practices research as a Digital Clinician Scientist at Bielefeld University. His scientific interest is in electrophysiology, emergency care and digital medicine.

Supplementary material

Supplementary material is available at *European Heart Journal—Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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