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"Torsades de pointes" in a patient with history of epilepsy

Case report

A female Caucasian patient, born 1930, was admitted to hospital for reposition of a broken arm. During anaesthesia she developed "torsades de pointes" (fig. 1) and was successfully defibrillated. The 12-lead ECG after defibrillation is shown in figure 2.

(1.) What is your diagnosis? (2.) Do you suggest further steps, and if yes, which ones?

Explanatory answers

(1.) The 12-lead ECG shows bradycard sinus rhythm with left axis deviation, one SVES, borderline PQ interal and QTc prolongation of 508 ms. The T-wave is of low amplitude and double notch appearance. The ECG phenotype and the history of "torsades de pointes" are consistent with the long QT syndrome (LQTS) type 2 phenotype [1]. Subsequent ECG's confirmed the LQTS type 2 phenotype.

(2.) The patient was treated for epilepsy for more than 30 years. She reported that she suffered from witnessed (husband) syncope's usually in the morning, still lying in the bed, when the telephone rang, which is a typical trigger for ventricular arrhythmias in LQTS type 2 patients [2]. With the background of LQTS "epilepsy" must be considered as haemodynamically relevant ventricular arrhythmia, eg "torsades de pointes" in the context of LQTS. As a consequence phenytoine was stopped, beta-blocker therapy was started and the implantation of an ICD was discussed with the patient. Gene testing was performed and a heterozygous mutation was identified in the *KCNH2* gene.

Congenital long QT syndrome is an inherited disease characterised by prolonged ventricular repolarisation and a high risk for sudden cardiac death (SCD) due to complex ventricular arrhythmias and "torsades de pointes". Today eight LQTS phenotypes are known. The most common forms are type 1 and 2, caused by mutations in the potassium channel genes *KCNQ1* and *KCNH2*, respectively, and type 3, caused by mutations in the *SCN5A* gene encoding the cardiac sodium channel. These types can be distinguished on the surface ECG [1]. LQTS type 1 is characterised by the broadest T-wave, whereas type 2 shows low T-wave amplitude with a double notch appearance as shown in figure 2. Type 3 reveals the most delayed onset of a narrowly peaked T-wave which also can be biphasic, depending on

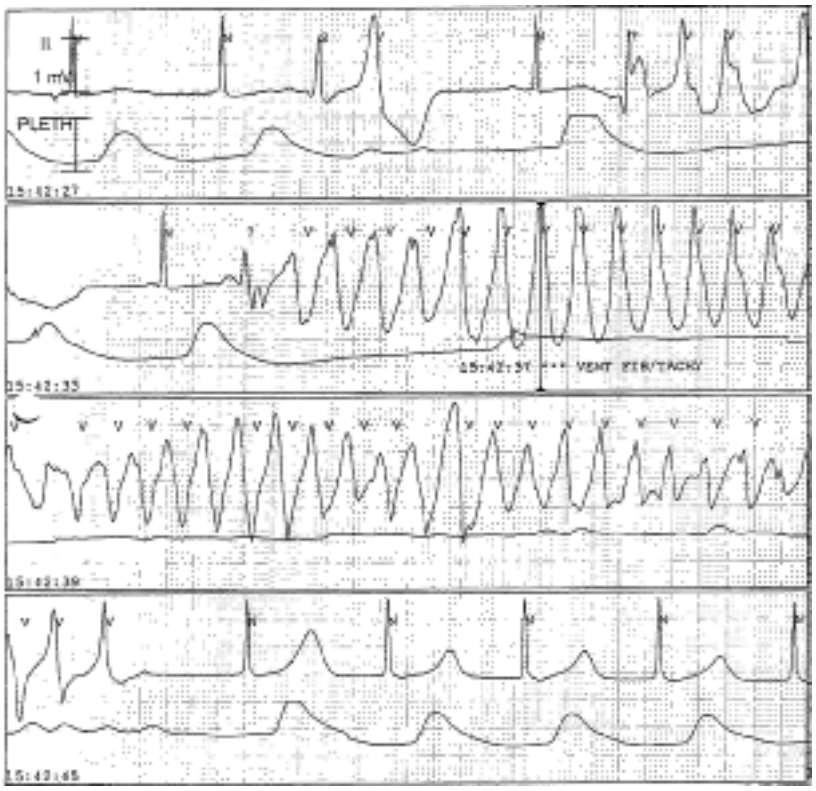

Figure 1
Intraoperative ECG monitoring with non-sustained "torsades de pointes".

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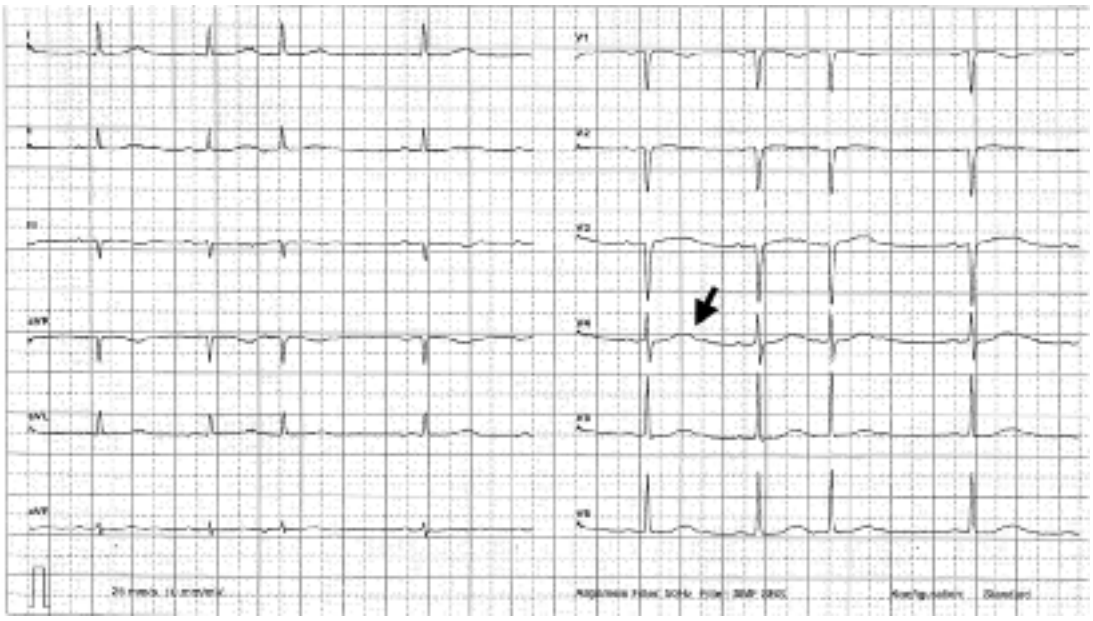

Figure 2
12-lead ECG showing QTc prolongation of 508 ms and a T-wave with low amplitude and double notch (arrow).

the preceding RR-interval. Triggers for ventricular arrhythmias are sports eg swimming in LQTS type 1, loud noise in LQTS type 2 and sleep/rest in LQTS type 3. The only tool to prevent LQTS patients from SCD is ICD implantation. Family screening (phenotype and genotype) in patients with LQTS is always indicated.

References

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2 Schwartz PJ, Priori SG, Spazzolini C, Moss AJ, Vincent GM, Napolitano C, et al. Genotype-phenotype correlation in the long-QT syndrome: gene-specific triggers for life-threatening arrhythmias. Circulation. 2001;103:89–95.