Patients with myotonic dystrophy type 1 (DM1) typically develop atrioventricular blocks, atrial, and ventricular arrhythmias.\textsuperscript{1–4} The incidence of sudden cardiac death is increased compared with the general population.\textsuperscript{1} The identification of patients at risk for sudden cardiac death is still challenging, even though larger cytosine-thymine-guanine trinucleotide repeat expansions and ECG abnormalities (rhythm other than sinus, PR interval $\geq$ 240 ms, second-third degree AV block, QRS $\geq$ 120 ms) were found to be associated with an increased risk of major cardiac events.\textsuperscript{1,2,4}

\section*{Introduction}

Patients with myotonic dystrophy type 1 (DM1) typically develop atrioventricular blocks, atrial, and ventricular arrhythmias.\textsuperscript{1–4} The incidence of sudden cardiac death is increased compared with the general population.\textsuperscript{1} The identification of patients at risk for sudden cardiac death is still challenging, even though larger cytosine-thymine-guanine trinucleotide repeat expansions and ECG abnormalities (rhythm other than sinus, PR interval $\geq$ 240 ms, second-third degree AV block, QRS $\geq$ 120 ms) were found to be associated with an increased risk of major cardiac events.\textsuperscript{1,2,4}

\section*{Case report}

At age 16, the patient presented a brief loss of consciousness while playing soccer. No investigations were performed at that time. He continued to play soccer until age 25 when, again during a soccer game, he had a syncope associated with an episode of paroxysmal atrial fibrillation. ECG, echocardiogram, stress test, Holter recording, and EEG were within normal limits. He was discharged with no specific diagnosis. Two years later, he had a cardiac arrest (CA) while playing soccer caused by ventricular fibrillation (VF). He was resuscitated on the field with basic life support measures and DC-shock and admitted to the hospital, without neurological impairment. No major arrhythmias were subsequently detected and ECG was normal. Echocardiogram showed marked right ventricular trabeculae. Coronary angiography did not reveal coronary artery anomalies. At right ventricular angiography, areas of wall motion abnormalities of mild extent were noted with evidence of apical trabeculae. Right ventricle myocardial biopsy revealed mild fibrosis and myocyte hypertrophy, but no fatty infiltrations. Cardiac magnetic resonance imaging (MRI) was reported to show non-specific findings. Electrophysiological study (EPS) showed: normal sinus node function and AV conduction; programmed ventricular stimulation did not induce ventricular tachycardia (VT) or VF. The exercise test did neither provoke major atrial and/or ventricular arrhythmias nor repolarization abnormalities. The patient was discharged on nadolol.

Eight years after the CA, the patient was referred to our Institution (out-patient Neuromuscular Clinic) to rule out the diagnosis of DM1, which had been made in his twin brother at that time. Since the CA he stopped playing soccer and did not complain any cardiac symptoms except occasional well-tolerated short episodes of palpitations. Cardiac assessment (ECG, echocardiogram, 24-h Holter monitoring) did not reveal abnormalities. Myotonia, distal muscle weakness, and cataracts fulfilled clinical diagnostic criteria for DM1, which was confirmed by genetic testing [cytosine-thymine-guanine repeat (CTGn $= 5/812$)].

During the subsequent 4 years follow-up, the clinical status of the patient did not substantially change. However, considering the history of previous CA a cardiac re-evaluation was advised. ECG, echocardiogram, and Holter monitoring were within normal limits. Repeat EPS showed: normal sinus node function and A-V conduction times (AH interval $= 92$ ms; HV $= 42$ ms); atrial stimulation induced a typical clockwise atrial flutter (cavo-tricuspid isthmus-dependent), which was reproducible; programmed ventricular stimulation was unable to induce VT or VF. Cardiac MRI imaging showed normal size and function of both ventricles.

\section*{Discussion}

Our case had two not well-defined syncopal episodes and one resuscitated CA due to VF, most likely related to DM1 predisposing arrhythmia occurrence. For these reasons, if recent international guidelines\textsuperscript{5} had been applied at the time of CA, the patient would probably have had a cardioverter defibrillator (ICD) implanted. On the other hand, no further episode occurred during the 12-year follow-up period. In addition, except for the occurrence of atrial fibrillation at age 25, ECG-predictive factors for sudden cardiac death

\begin{center}
\textbf{SHORT COMMUNICATION}
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Long-term follow-up free of ventricular fibrillation recurrence after resuscitated cardiac arrest in a myotonic dystrophy type 1 patient
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in DM1 subjects, recently reported by Groh et al.\(^2\) were not present in our patient. EPS was negative in two occasions and did not provide specific indications regarding the need for more invasive preventive treatments like pacemaker or ICD implantation. Nevertheless, considering the recent guidelines\(^3\) that recommend ICD implantation in patients who are survivors of CA owing to VF, and the fact that DM1 patients, particularly with large number of CTG repeats, as our patient, have a higher rate of ventricular ectopy and of conduction defect progression, we decided to implant an ICD. However, in the proband’s twin, who had more severe muscle weakness, but no cardiac involvement (no ECG abnormalities, normal echocardiogram and MRI, negative EPS, which was performed considering the family history of CA), and never presented syncope, CA, or major arrhythmia, our treatment strategy was to follow-up the patient, monitoring the appearance of cardiac symptoms and ECG abnormalities.

In conclusion, our report confirms that CA is a possible event in adult patients with DM1 even in the absence of structural cardiac involvement. On the other hand, patients, as our proband, may remain asymptomatic after a resuscitated CA for very long period of time, yet bearing the potential risk of life-threatening arrhythmias, which justifies ICD implantation.

**Conflict of interest:** none declared.

**References**


