

# Myotonic Dystrophy and Bundle-Branch Re-Entrant Tachycardia

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We report the case of a 37-year-old man diagnosed with myotonic dystrophy who presented atrial fibrillation with high ventricular rate. While being treated with amiodarone, he suffered cardiac arrest. The electrophysiological study disclosed bundle-branch reentrant ventricular tachycardia and ventricular fibrillation. Catheter ablation of the right bundle branch was performed and a bicameral defibrillator was implanted. The mechanisms and treatment of arrhythmias in these patients are discussed.

**Key words:** *Dystrophy. Tachycardia. Re-entry. Ablation. Defibrillator.*

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## Distrofia miotónica y taquicardia ventricular por reentrada rama-rama

Presentamos el caso de un varón de 37 años con distrofia muscular miotónica que ingresó por fibrilación auricular con alta respuesta ventricular. Durante la administración de amiodarona presentó un episodio de muerte súbita intrahospitalaria. En el estudio electrofisiológico se evidenció una taquicardia ventricular por reentrada rama-rama y una fibrilación ventricular. Se realizó, con éxito, ablación mediante energía de radiofrecuencia de la rama derecha del haz de His y se implantó un cardiodesfibrilador bicameral. Se discuten los mecanismos y tratamiento de las arritmias en estos pacientes.

**Palabras clave:** *Distrofia. Taquicardia. Reentrada. Ablación. Desfibrilador.*

## INTRODUCTION

Myotonic muscular dystrophy is the most frequent type of muscular dystrophy in adults, with a prevalence of 1 in 8000.<sup>1-3</sup> It is an autosomal dominant hereditary familial disease characterized by unstable amplification of the cytosine-thymine-guanine trinucleotide in the long arm of chromosome 19.<sup>1,4</sup> It affects the neuromuscular system and can also affect other systems.<sup>1,3</sup> Cardiac involvement is frequent and disturbances in the His-Purkinje conduction system predominate.<sup>2,4,5</sup> In post-mortem studies, fibrosis, fat infiltration, and atrophy of the conduction system are found.<sup>6,7</sup> Myocardial involvement is rare, although echocardiography shows disturbances in the ventricular diastolic function<sup>8,9</sup> and, occasionally, systolic function as well.<sup>10</sup> Atrial and ventricular

arrhythmias are present in up to 50% of patients, with sinus bradycardia, atrioventricular block, atrial and ventricular premature beats, atrial fibrillation and flutter, and ventricular tachycardia being reported.<sup>4,11</sup> The frequency of sudden death varies in different series and is caused by complete atrioventricular block and ventricular tachycardia or fibrillation. Exceptionally, sustained ventricular tachycardia is the cardiac form of presentation of this disease<sup>4</sup> and the most likely trigger mechanism is bundle-branch re-entry, as demonstrated in recent publications.<sup>2,13</sup>

## PRESENTATION OF CLINICAL CASE

A 37-year-old man, who had been diagnosed two years earlier as myotonic muscular dystrophy by a genetic study and had a family history of the disease (his mother, brother, and maternal uncles and cousins had died), was seen in the emergency service for chest pain, palpitations, diaphoresis, and dyspnea. The physical examination disclosed an arterial pressure of 120/80 mm Hg and heart rate of 110 beats/min. On auscultation, the heart sounds were arrhythmic and tachycardic, without murmurs. The pulmonary fields were well ventilated. Other typical findings of the

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**Fig. 1.** Twelve-lead electrocardiogram showing atrial fibrillation with a ventricular response of 110 beats/min and complete block of the left bundle branch.

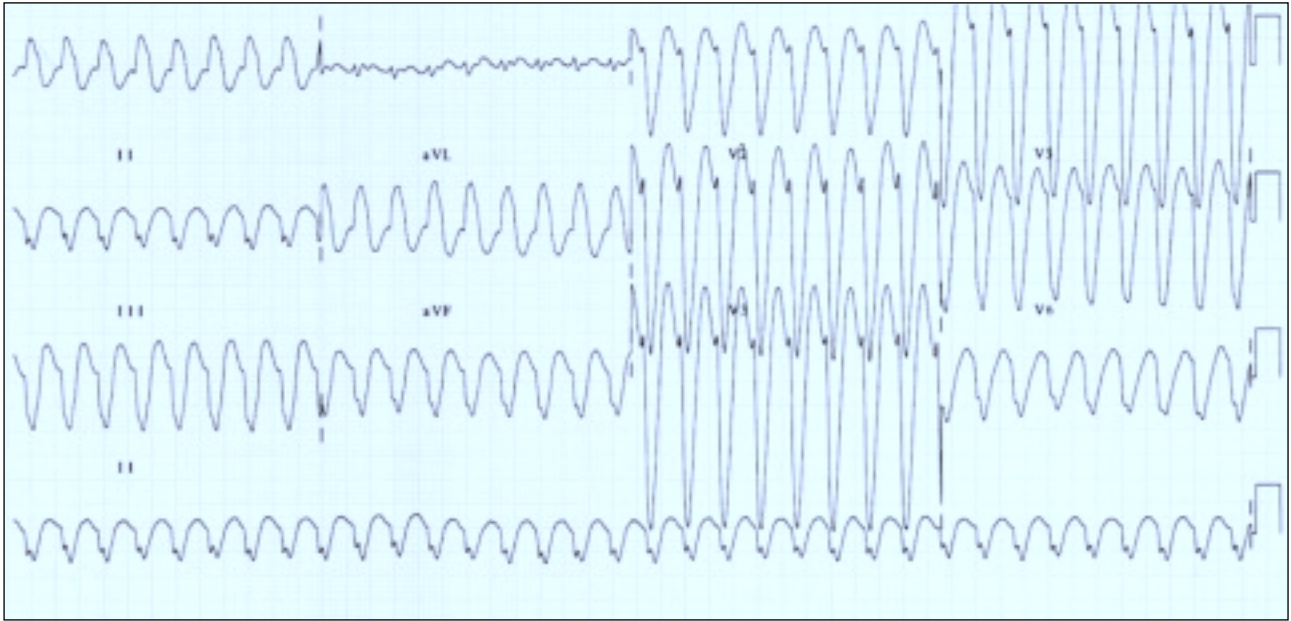
disease were frontal alopecia, bilateral palpebral ptosis, testicular atrophy, muscular weakness with distal predominance, generalized hyporeflexia, and moderate mental retardation.

The electrocardiogram showed atrial fibrillation with a high ventricular response and complete left bundle-branch block (Figure 1). In the chest X-ray was appreciated an increased cardiothoracic index with left ventricular enlargement and incipient signs of pulmonary congestion. In the echocardiogram, biventricular systolic and diastolic function was normal. No signs of pulmonary hypertension or valvular defects were found. The differential blood count, coagulation parameters, general biochemistry, and electrolytes were normal.

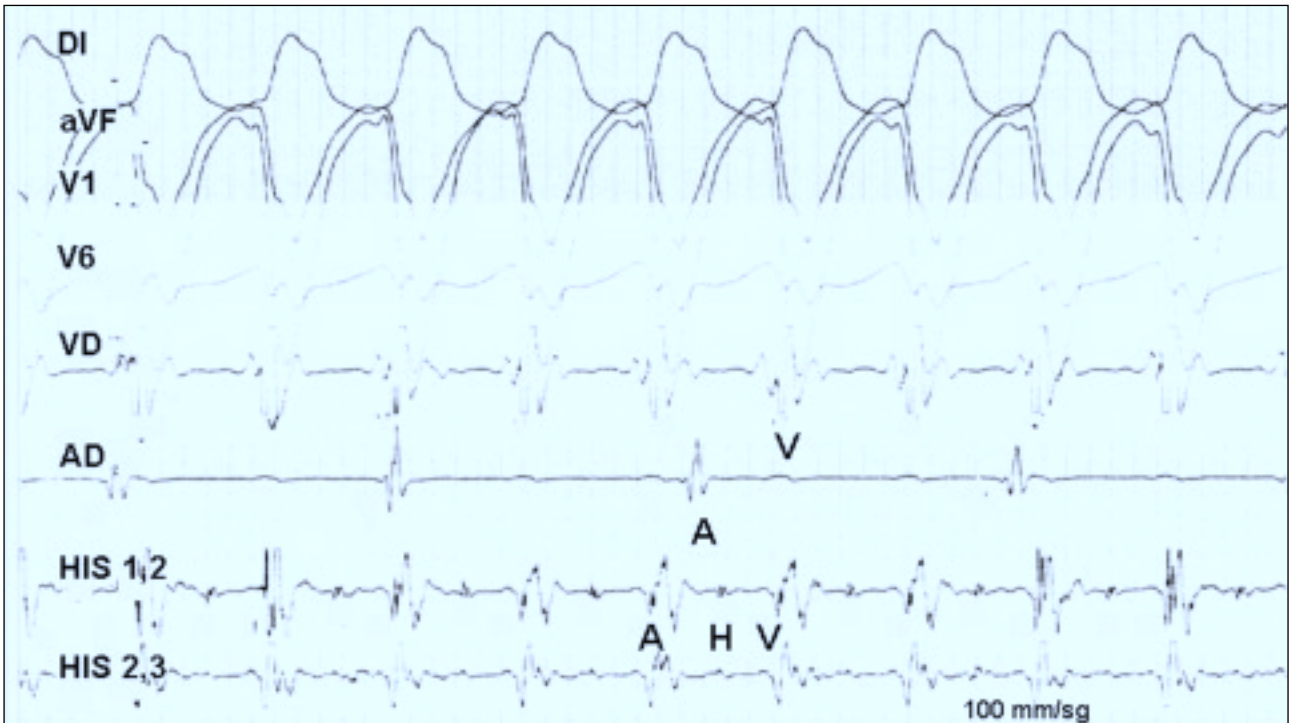
The patient was hospitalized in the cardiology department to begin treatment for atrial fibrillation. Pharmacological treatment with intravenous amiodarone began with an initial dose of 5 mg/kg and continued with a 24-hour infusion of 10 mg/kg. The day after admission, the patient presented an episode of regular tachycardia with a wide QRS complex, heart rate of 160 beats/min (Figure 2), and hemodynamic collapse (blood pressure [BP] 50/0 mm Hg) that required electrical cardioversion with a 200-joule discharge. The patient came out of atrial fibrillation, showing an electrocardiogram similar to the one seen at admission (until then, the patient had received 750 mg of amiodarone intravenously). The patient was transferred to the coronary care unit, where a wide QRS complex, regular tachycardia appeared again without hemodynamic impairment. A

pattern of left bundle-branch block with a left axis was evident. Procainamide, 300 mg, was given intravenously, which resulted in a conversion to sinus rhythm with first-degree atrioventricular block.

After discontinuing amiodarone 48 h earlier, an electrophysiological study was made via the right femoral vein with quadripolar catheters placed in the upper right atrium, bundle of His region, and right ventricular apex. The intervals of atrioventricular conduction showed first-degree infrahisian atrioventricular block with the following intervals: PA 30 ms, AH 80 ms, and HV 100 ms. Sinus function and Wenckebach AV point were normal. No hidden accessory pathways were appreciated in sinus rhythm. With atrial stimulation at 280 ms, common atrial flutter was induced with a cycle length of 240 ms and 2:1 atrioventricular conduction that required atrial pacing at 200 ms for conversion. During baseline programmed ventricular pacing at 600 ms, ventricular fibrillation was induced with three extrastimuli (S2, 250 ms; S3, 240 ms, and S4, 210 ms) that required defibrillation by the application of a 200-joule discharge. With ventricular pacing at 500 ms and 2 extrastimuli (S2, 260 ms, and S3, 330 ms), a regular sustained monomorphic ventricular tachycardia was induced. It was tolerated and had a pattern of left bundle-branch block and a cycle length of 280 ms. Atrioventricular dissociation and a stable His potential were appreciated before the ventricular electrogram with a constant HV interval (Figure 3). The tachycardia was terminated by a ventricular extrastimulus at 250 ms. A cardiac electrophysiological study was made and the



**Fig. 2.** Twelve-lead electrocardiogram showing regular tachycardia with a wide QRS complex, left axis, and a pattern of complete left bundle-branch block. The heart rate is 160 beats/min.



**Fig. 3.** Induction of ventricular tachycardia due to bundle-branch re-entry by programmed ventricular pacing with a cycle length of 280 ms. Observe the His potential preceding the ventricular electrogram and atrioventricular dissociation. H indicates His potential; V, right ventricular potential; A, right atrial potential.

right bundle branch was ablated with a catheter with a 4-mm distal electrode and application of radiofrequency energy for 2 min at a temperature of

60°C. After ablation, atrioventricular conduction was maintained with a PR interval of 240 ms and HV of 130 ms. A new programmed ventricular pacing

protocol was begun and tachycardia was not again induced.

## DISCUSSION

Bundle-branch re-entry is the typical mechanism of sustained monomorphic ventricular tachycardia in patients with myotonic muscular dystrophy.<sup>2,13</sup> Merino et al<sup>2</sup> made a genetic study of 6 patients with confirmed myotonic dystrophy who presented wide QRS complex tachycardia due to a bundle-branch re-entrant mechanism.<sup>2</sup>

Macroreentry in the His-Purkinje system is the cause of 6% of ventricular tachycardias. It is usually associated with dilated cardiomyopathies and severe valvular dysfunction.<sup>14</sup> The requisite condition for triggering this arrhythmia is a delay in conduction through the His-Purkinje system, which is a frequent finding in myotonic dystrophy.<sup>2,5,8,15</sup> The involvement of this system is manifested in the surface electrocardiogram by intraventricular conduction defects or bundle-branch block and in the electrophysiological study by prolongation of the HV interval.<sup>5,7,13,15</sup>

The tachycardia that appeared in this patient was undoubtedly due to a bundle-branch re-entrant mechanism, as demonstrated by the suppression of inducibility by ablation of the right bundle branch.

It is interesting to note that at admission, atrial fibrillation with complete block of the left bundle branch was documented in this patient and that recurrent sustained ventricular tachycardia appeared after the intravenous administration of 750 mg of amiodarone. This finding could be due to a proarrhythmic effect of amiodarone, in view of the fact that no episodes of ventricular tachycardia had been detected previously and that the delay in atrioventricular conduction favors the appearance of bundle-branch re-entry with these antiarrhythmic agents.<sup>16</sup> Nevertheless, the diagnostic electrophysiological study was made after amiodarone was discontinued (48 h) and confirmed the abnormalities in the His-Purkinje conduction system.

It is important to diagnose ventricular tachycardia due to bundle-branch re-entry because it can cause sudden death in these patients.<sup>17</sup> The response to pharmacological treatment is poor<sup>2,12,14</sup> but ablation of one of the branches of the His system, usually the right branch, followed by permanent pacemaker implantation has been shown to be the treatment of choice, with very good results.<sup>2,13,14</sup>

Patients with muscular dystrophy have an incidence of sudden death that varies widely in published series, reaching 33%.<sup>4,6,8,15</sup> Atrioventricular block, ventricular tachycardia, and ventricular fibrillation have been identified as the cause of these episodes.<sup>2,4,12,15,17</sup> Some reports have shown that patients with myotonic

dystrophy who have been implanted a definitive pacemaker can die suddenly from ventricular tachyarrhythmias.<sup>2,8,17</sup> Lazarus et al<sup>8</sup> have proposed the prophylactic implantation of a definitive pacemaker in patients with significant abnormalities in the His-Purkinje conduction system to prevent episodes of sudden death due to atrioventricular block. The same author has found an incidence of 18% of ventricular arrhythmias (polymorphic ventricular tachycardia and ventricular fibrillation) induced in the electrophysiological study in a heterogeneous population of these patients.<sup>8</sup> On the other hand, patients with bundle-branch re-entrant ventricular tachycardia with structural heart disease have a higher incidence of sudden death determined by the clinical characteristics of each condition. Saliba and Natale have discussed whether these patients should routinely be implanted a cardiodefibrillator, since up to 6.5% of these patients die suddenly from arrhythmias in spite of successful ablation.<sup>13</sup> A defibrillator was implanted in this patient because a pacemaker had to be implanted for his conduction disorder and it was judged that he might be at risk of sudden death in spite of ablation of the right bundle branch, for the reasons described above.

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