Undersensing of Spontaneous Ventricular Fibrillation as a Cause of Sudden Death in a Child

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Received: 12/21/2007
Accepted: 08/31/2008

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ABSTRACT

In few cases implantable cardiodefibrillators have been reported to undersense ventricular fibrillation, and most of them happened during the measurement of defibrillation thresholds. This type of undersensing has been suggested as a potential cause of sudden death. In this case report we describe a 4-year old boy with hypertrophic myocardiopathy who suffered a spontaneous episode of ventricular fibrillation with a great alternation in the amplitude of local ventricular electrograms. The episode was undersensed by the device and therefore it was not treated, causing an episode of sudden death.

Rev Argent Cardiol 2008;76:317-320.

Key words > Implantable Cardioverter-Defibrillators - Ventricular Fibrillation - Sudden Death

Abbreviations >

ICD	Automatic implantable cardioverter-defibrillator	**VT**	Ventricular tachycardia
VF	Ventricular fibrillation	**SPVT**	Sustained polymorphic ventricular tachycardia
SD	Sudden death		

BACKGROUND

Automatic implantable cardioverter-defibrillator (ICD) has demonstrated to improve survival of patients at high risk of sudden death (SD) due to malignant ventricular arrhythmias. (1) Nevertheless, the presence of SD has been reported as a consequence of failure of electrode leads, device malfunction or accidental deactivation of the ICD. (2)

Since the last years different sensing algorithms have been developed in order to detect alternating low-amplitude ventricular signals which frequently occur during a ventricular tachycardia or ventricular fibrillation. At the same time, these algorithms are programmed to avoid T-wave oversensing.

While oversensing leads to delivery of an inadequate therapy, undersensing of a lethal arrhythmia may provoke death.

Few cases of undersensing of ventricular fibrillation (VF) have been published; however, most of them occurred during defibrillation thresholds measurement while the device was placed or at short-term follow-up during hospitalization. This type of undersensing is likely to cause SD. (3-8)

In this case report we describe a 4-year old boy with hypertrophic myocardiopathy who suffered a spontaneous episode of ventricular fibrillation that was undersensed by the device and therefore was not treated, causing an episode of sudden death.

CASE REPORT

A four-year old boy was referred to our institution in June 2004.

He had suffered two episodes of SD due to documented VF that was reverted with external defibrillation.

His brother had died suddenly two years before at the age of 9.

A diagnosis of hypertrophic myocardiopathy was made by echo-Doppler with an intraventricular septum diameter of 23 mm.

A VVI Vitality-VR 1870 (Guidant Inc, St.Paul, MN, USA) cardioverter-defibrillator was implanted in the apex of the right ventricle from the left suclavian approach, with a double-coil active fixation bipolar lead (interelectrode spacing > 6 mm).

The following normal measurements were obtained during implantation: intraoperative stimulation threshold 0.4 V to 0.5 msec, R wave amplitude 6 mV and stimulation and shock impedance 438 and 33 ohms, respectively.

The defibrillation threshold was established with a minimum sensitivity setting of the ICD to ensure appropriate sensing of low-amplitude ventricular fibrillation signals. The device detection was programmed for a zone of ventricular fibrillation of 180 beats per minute, with a minimum sensitivity of 0.43 mV. Ventricular fibrillation was induced thereafter with an electrical shock on the T wave; the arrhyth-

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mia was properly sensed and reverted with a shock delivery of 11 Joules in two opportunities.

During the immediate postoperative, ventricular fibrillation zone detection was programmed at 220 beats per minute (to avoid inappropriate therapies due to sinus tachycardia) with a first shock of 21 Joules and subsequent shocks of 31 Joules. A nominal sensitivity was programmed at 0.27 mV as usual to avoid undersensing of ventricular arrhythmias.

The boy was discharged on 10 mg/kg/day of oral amiodarone.

In January 2006 the child suffered two episodes of sustained polymorphic ventricular tachycardia (SPVT), which were properly sensed, detected and treated by the device (Figure 1).

Amiodarone was replaced by sotalol (75 mg/day).

The device was controlled every 3 months; data obtained with the intervalometer were normal (R wave amplitude > 7 mV with normal stimulation impedance).

In February 2006 the child presented an episode of VF with SD. The ICD failed to deliver an electrical shock. His father started cardiopulmonary resuscitation until the child was assisted by a medical team; 10 minutes after the onset of the episode, he was successfully rescued with an external defibrillator.

The boy was hospitalized. The chest X-Ray showed that the lead was correctly placed.

R-wave amplitude was 9.3 mV. Stimulation impedance and shock impedance were 447 ohms and 32 ohms, respectively.

A great alternation in the amplitude of the stored local ventricular electrograms was observed during the event. The device failed to sense low-amplitude electrograms which followed high-amplitude electrograms. In this way, the arrhythmia was not properly detected (Figures 2 and 3).

The ICD was reprogrammed at a sensitivity of 0.18 mV (maximum sensitivity) for a better detection of VF. The detection zones for ventricular tachycardia and ventricular fibrillation were programmed at 160 and 180 beats per minute, respectively.

Finally, "Committed Shock" was turned on in order to skip the reconfirmation algorithm prior to therapy once the arrhythmia is detected. In this way, the shock is released at once when VT or VF are detected in the programmed zone.

Sotalol was replaced by atenolol (0.75 mg/kg/day) and amiodarone (10 mg/kg/day).

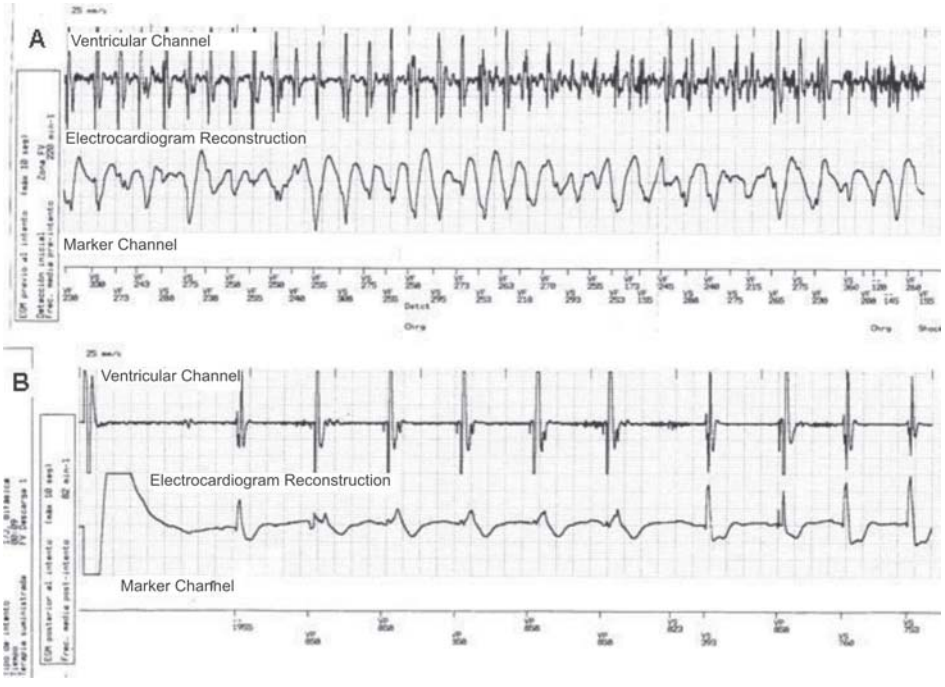

Fig. 1. Record stored by the ICD, showing adequate sensing (**A**) and shock delivery (**B**)

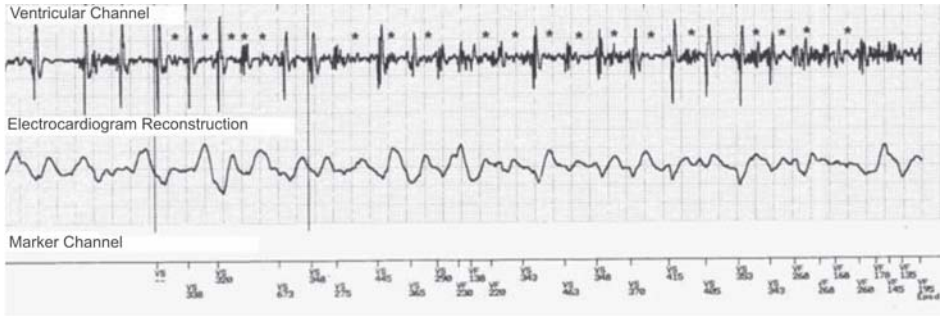

Fig. 2. Record stored by the ICD, Asterisk (*) indicates absence of sensing of local ventricular electrograms producing a failure in the detection of the arrhythmia. Note that the device failed to sense those electrograms of low amplitude which occurred after electrograms of high amplitude.

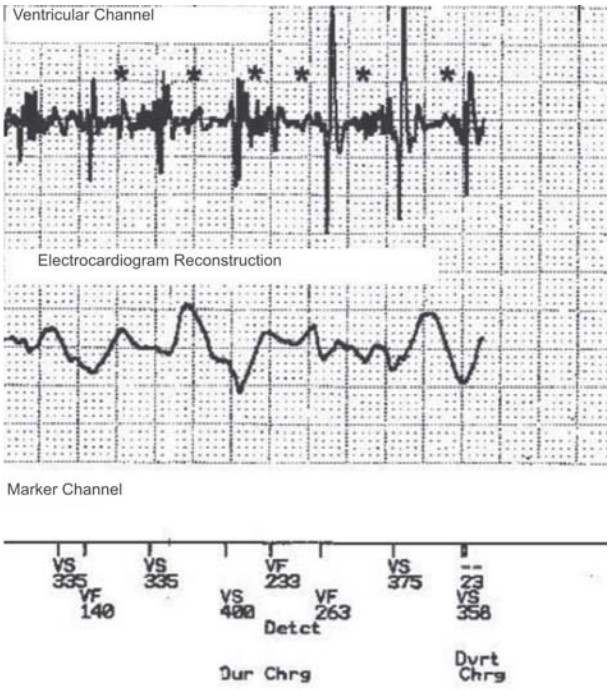

Fig. 3. Record stored by the ICD, Asterisk (*) indicates absence of sensing of local ventricular electrograms producing a failure in the detection of the arrhythmia. ICD deverts the therapy (Dvrt Chrg) while the patient remains in VF.

Under general anesthesia, SPVT and VF were induced. Both arrhythmias were properly sensed, detected and reverted by the device.

On November 20, 2007, he suffered another episode of VF that was correctly treated by the ICD.

DISCUSSION

The incidence of sudden death in patients with hypertrophic myocardiopathy is 2% to 4% in tertiary health care centers, while in unselected populations it is less than 1%. (9) Implant of ICD is indicated for secondary prevention in patients resuscitated from SD and in subjects with symptoms due to ventricular tachycardia and/or syncope related to ventricular arrhythmias. (10, 11)

The incidence of inappropriate therapies is about 20-30%.

Implantable dual-chamber cardioverter-defibrillators may be better for detecting supraventricular arrhythmias and thus may reduce inadequate shocks. Amiodarone and antitachycardia pacing have been effective for shock reduction in randomized trials. (12, 13)

The different sensing algorithms have been designed to detect and treat VT or VF in a rapid and efficient fashion and to prevent oversensing of intracardiac or extracardiac signals which might lead to inappropriate therapies. Automatic gain control algorithms try to fulfill these requirements.

Implantable cardioverter-defibrillators should be programmed with special care and their normal operation should be carefully checked due to the great variability of the amplitude of ventricular signals detected during VT or VF.

Although a few cases of undersensing of VF have been published, most of them took place while intraoperative thresholds were measured or after changing sensitivity adjustment during in-hospital check-ups. (3-8)

In this case, undersensing occurred during a spontaneous VF in an outpatient basis with the subsequent absence of therapy that led to an episode of sudden death.

The implanted device has an "automatic gain control" and a dynamic sensing algorithm. It starts at the 75% of amplitude of the R-wave and decreases until it reaches a minimum value with a predetermined time constant.

We suppose that the ICD did not sense the VF due to the great alternation in the amplitude of local ventricular electrograms. After a large-amplitude electrogram, subsequent electrograms were smaller and remained below the dynamic level of sensing. In this sense, the heart rate detected was slower than the programmed detection rate.

For that reason we decided to reprogram the device reducing the detection rate and setting "Committed Shock" to ON; in this way, shock would be delivered once the arrhythmia has been detected skipping the reconfirmation algorithm prior to therapy.

Antiarrhythmic drugs might modify defibrillation thresholds, and amiodarone as well as sotalol might predispose to polymorphic ventricular tachycardia ("torsade de point") due to a proarrhythmic effect; however, this was not the case and the absence of antiarrhythmic therapy is clearly related to undersensing of low-amplitude signals which follow large-amplitude electrograms. Alternation between local consecutive electrograms has not been described as an adverse event related to antiarrhythmic drugs. (14)

A similar clinical case as the one here described has been previously published, but defibrillation tests have not been performed prior to the event, and although the ICD failed to sense the ventricular arrhythmia, it could complete the detection of the event and treat it successfully. (8)

The present case describes undersensing of a spontaneous VF in an outpatient basis which resulted in absence of adequate therapy and required external defibrillation to avoid the death of a child.

The problem was solved when the ICD was adequately reprogrammed.

Competing Interests
None declared.

RESUMEN

Subsensado de fibrilación ventricular espontánea como causa de muerte súbita en un niño

Se han comunicado casos aislados de subsensado de fibrilación ventricular por el cardiodesfibrilador implantable. La mayoría de ellos ocurrieron durante la medición de umbrales desfibrilatorios. Se ha sugerido que este tipo de subsensado podría ser una causa potencial de muerte súbita.
En esta presentación se describe a un paciente de 4 años con miocardiopatía hipertrófica que sufrió un episodio espontáneo de fibrilación ventricular, con una gran alternancia en la amplitud de los electrogramas ventriculares locales, que fue subsensado y no tratado por el dispositivo y provocó muerte súbita.

Palabras clave > Desfibriladores implantables - Fibrilación ventricular - Muerte súbita

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