

CASE REPORT

Documented exercise-induced cardiac arrest in a paediatric patient with hypertrophic cardiomyopathy

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A paediatric patient with hypertrophic cardiomyopathy (HCM) presented cardiac arrest due to ventricular fibrillation. Ventricular arrhythmias were not induced in an electrophysiological study, but an implantable cardioverter defibrillator (ICD) was implanted. Nine months later, the child experienced a recurrence of cardiac arrest during exercise, which was successfully treated with a defibrillator shock from the device. Analysis of the stored electrograms demonstrated ventricular fibrillation of abrupt onset following sinus tachycardia. The risk factors and the potential mechanism leading to recurrent cardiac arrest in this case are discussed. This report supports implantation of an ICD as a life-saving therapeutic approach not only for adults but also for children with HCM at high risk.

Sudden death is a well-known complication in patients with hypertrophic cardiomyopathy (HCM). Whereas, it is usually associated with ventricular arrhythmias in adult patients, the mechanism responsible for cardiac arrest in children with HCM is unclear.¹ Only a few reports of aborted sudden death in children and adolescent patients, and limited information about the treatment of these patients with automatic implantable cardioverter defibrillators (ICD) is available.^{2–6} We report a case of a 12-year-old boy with HCM and aborted sudden death caused by ventricular fibrillation, who underwent ICD implantation. Very shortly thereafter, he experienced an effective defibrillator shock, and the stored electrogram showed ventricular fibrillation following sinus tachycardia during exercise. In this child with HCM, the benefit of an ICD is clearly demonstrated through stored intracardiac electrogram analysis.

Case report

A 12-year-old male was diagnosed with HCM during the investigation of a systolic murmur when he was three. At that time the echocardiogram showed asymmetrical left ventricular hypertrophy, with a 15 mm thick septum, and absence of intraventricular gradient. A 24-h ambulatory Holter monitoring did not show ventricular tachyarrhythmias. His brother was found to have HCM when his family was studied. The patient remained asymptomatic under

treatment with propranolol until 9 years later when he suffered a cardiac arrest while playing football. Resuscitation was initiated by his father, and afterwards by the emergency squad. On the monitor ventricular fibrillation was observed, and sinus rhythm was restored with a 200-J external shock.

During his hospital admission he underwent an echocardiogram (*Figure 1*); septal thickness was 36 mm, and an intraventricular gradient of 80 mmHg was measured. The electrocardiogram showed left ventricular strain pattern. Again 24-h Holter monitoring did not register any ventricular arrhythmias. Late potentials were not present on a signal-averaged electrocardiogram. No perfusion defects were found on exercise-thallium scintigraphy; of note, a 20 mmHg decrease in blood pressure was detected during exercise. An electrophysiological study was performed with three extrastimuli (S2, S3, S4) from two stimulation points in right ventricle (apex and outflow tract) using three basic cycle lengths (600, 400 and 350 ms), but no sustained ventricular arrhythmias were induced. The preferred therapeutic option was to implant an ICD (Ventak MINI, Cardiac Pacemakers Inc., MN, USA) with a transvenous lead system. No complications related to the implantation procedure were observed. The patient was discharged on propranolol (80 mg three times a day).

Nine months later, the child experienced a syncope while playing football at school. It was followed by an ICD shock. This episode was not preceded by chest pain or any other symptom. At the hospital, the electrogram related to the ICD shock was obtained by means of the ICD programmer, showing abrupt-onset ventricular fibrillation preceded by

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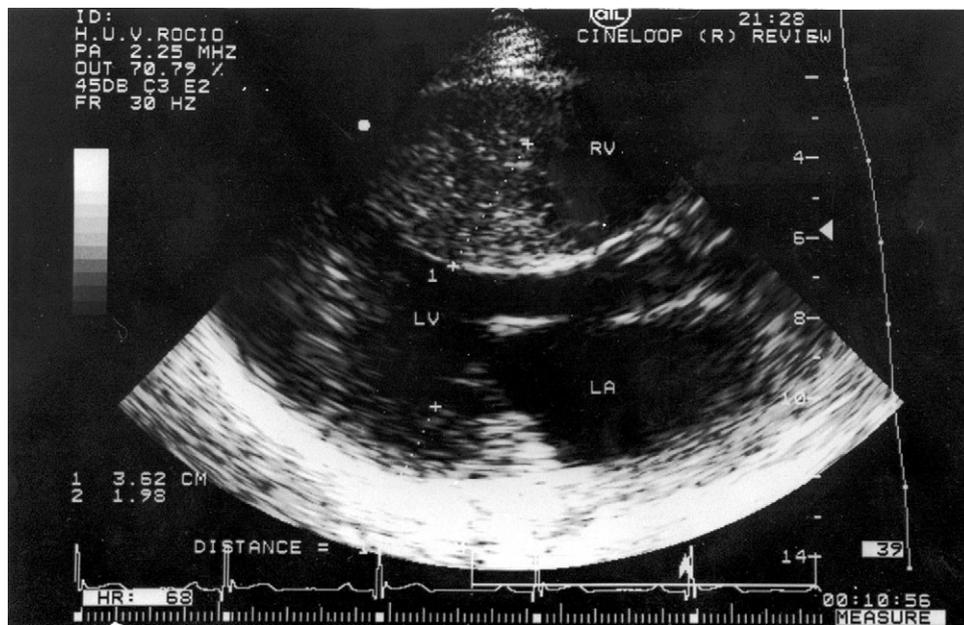


Figure 1 Echocardiogram showing an asymmetrical left ventricular hypertrophy. Septal thickness is 36 mm.

sinus tachycardia (155 bpm) and followed by a successful 27-J biphasic shock (Figure 2). Verapamil (80 mg three times a day) was started, and abstinence from vigorous physical activity was emphasized. No recurrences have been detected after 36-month follow-up.

Discussion

HCM is characterized by ventricular hypertrophy not caused by known diseases or conditions, and it has been related to heterogeneous genetic mutations.⁷ It can be diagnosed at any age and its clinical course is variable, ranging from asymptomatic patients to those presenting sudden death. Among children, HCM is associated with especially increased mortality, and it has been suggested that half of the child population who are diagnosed at an age between 1 and 14 years die suddenly in the following 9 years.⁸

In the few reported cases of children with HCM resuscitated after cardiac arrest, the underlying arrhythmia is usually a polymorphic ventricular tachycardia or ventricular fibrillation.⁹ The mechanisms leading to sudden death in these patients can be varied, and may interact among them; it has been suggested that they could differ in young and adult patients.¹⁰ It is possible that, in younger patients, sudden death is more frequently related to myocardial ischaemia rather than to a primary arrhythmogenic ventricular substrate.⁸ In the present case, the abrupt onset of ventricular fibrillation and the absence of late potentials or inducible ventricular arrhythmias on programmed stimulation do not support the hypothesis of an established arrhythmic ventricular substrate other than presumed myocardial disarray. Cardiac arrest can be triggered by atrial fibrillation in adolescent patients with HCM,¹¹ but it was not registered in this child anytime. Despite the absence of regional perfusion defects in the stress scintigraphy images, myocardial ischaemia cannot be excluded as the precipitor of this event. It is known that some patients

with HCM may have an abnormal vascular response to exercise, as has this child.¹² In the present case, one potential explanation could be that, during strenuous exercise, sinus tachycardia combines with severe hypotension and decreased myocardial perfusion to trigger ventricular fibrillation. In this setting the presence of moderate to severe outflow tract obstruction may have contributed to the event in this child.

The identification of individual patients with HCM at high risk for sudden death remains controversial. Several indicators have been identified but have poor positive predictive value and a high negative predictive accuracy.^{13,14} This child had two risk factors for sudden death at the time of his cardiac arrest, the degree of wall thickness, and the abnormal blood pressure response to exercise. A recent study reported that the magnitude of left ventricular hypertrophy (≥ 3 cm) is directly related to a risk of sudden death and justifies ICD use, particularly in the young.¹⁵ However, severe cardiac hypertrophy was not associated with increased rate of therapeutic ICD interventions in another study.¹⁶ It is possible that this risk factor alone has insufficient predictive accuracy to guide decisions regarding prophylactic treatment. An abnormal blood pressure response during exercise can be detected in 25% of HCM patients. It is a more sensitive indicator of risk in younger patients and is associated with sudden death, although the relative risk is low.¹⁴ Therefore, a positive result should be used in conjunction with other risk factors. It is interesting to note that some of the most mentioned markers of high risk, such as the detection of non-sustained ventricular tachycardias (nSVT) on Holter monitoring or the inducibility of sustained ventricular arrhythmias on electrophysiological study in high-risk population, were absent in this case despite suffering recurrent cardiac arrest. The low prevalence of nSVT in young patients with HCM is well recognized; current data suggest that the risk associated with nSVT is higher in the young.¹⁴ We agree that invasive electrophysiological investigations

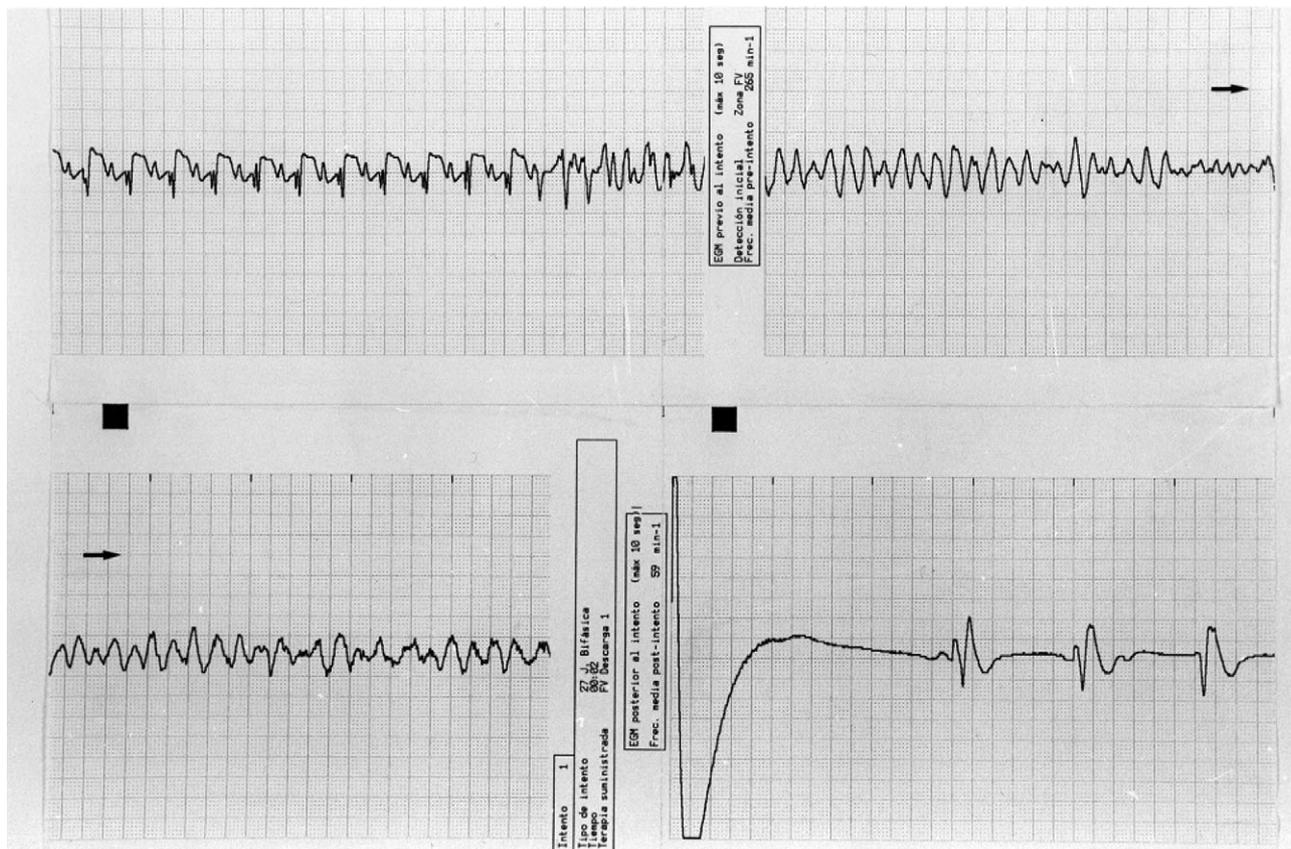


Figure 2 Stored intracardiac electrogram from the ICD register/monitor. A sinus tachycardia during exercise is followed by an abrupt onset of ventricular fibrillation, which is terminated by a 27-J shock from the device.

result in a low predictive positive accuracy and are not useful in most patients with HCM when assessing risk.¹⁷ However, there seems to be agreement that aborted sudden death is a strong predictor of recurrent cardiac arrest in patients with HCM.^{10,13,14}

It has been suggested that, in a selected adult population with HCM, the ICD seems to have a less important impact on prognosis than it has in adult patients with other aetiologies of cardiac arrest.^{18,19} Moreover, the information available in the literature on ICD treatment in children with HCM is limited.²⁻⁶ This case offers some confirmation that ICD therapy is effective in young patients with HCM and previous cardiac arrest. The ICD may become established as a therapeutic option in patients with HCM at high risk.^{16,19,20} These devices have not been evaluated in HCM as part of prophylactic trials because it is a relatively uncommon disease and difficulties in identifying patients who are at risk of sudden death. In two recent studies with long-term follow-up,^{16,20} the range of therapeutic ICD interventions is lower in patients who received devices for primary prevention (3–4.5%) that for secondary prevention (7–11%). We must consider, before implantation of an ICD in young patients, a high incidence of serious side effects, the most common being inappropriate ICD discharges, infections and lead related problems.^{4,5}

This fully documented case of a child with HCM and recurrent cardiac arrest in whom sinus tachycardia precedes abrupt-onset ventricular fibrillation may help to highlight the unclear mechanisms leading to sudden death in these

patients. Here the episode has been faithfully registered in the stored electrograms of a latest generation ICD. As sudden death has a particularly high incidence in children and young patients with HCM, especially when there is a history of previous cardiac arrest, ICD use can be an effective alternative treatment for these high-risk patients.

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