Surface fragmented QRS in a patient with hypertrophic cardiomyopathy and malignant arrhythmias: Is there an association?

Francisco Femenía, Mauricio Arce, Martín Arrieta, Adrian Baranchuk[1]

Unidad de Arritmias. Departamento de Cardiología. Hospital Español de Mendoza. Argentina,
[1]Heart Rhythm Service, Queen's University, Kingston, Ontario, Canada
***Address for correspondence:** Dr. Francisco Femenía, Av. San Martín 965. Godoy Cruz. Mendoza CP:5501, Argentina. E-mail: femeniafavier@hotmail.com*

ABSTRACT

An 18- year old woman with hypertrophic cardiomyopathy, aborted sudden cardiac death and implanted with an implantable cardioverter defibrillator (ICD), developed progressive fragmentation of her surface 12-lead electrocardiogram (ECG). During the follow-up, she presented with multiple appropriate ICD discharges. Here, we discuss the possible association between surface fragmented ECG and the risk of ventricular arrhythmias in patients with hypertrophic cardiomyopathy.

Key words: Fragmented QRS complex, hypertrophic cardiomyopathy, sudden cardiac death

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a genetically heterogeneous heart muscle disorder characterized by myocardial hypertrophy in the absence of abnormal loading conditions. While it has been recognized for many decades that some patients with the disease die suddenly from ventricular arrhythmia, data from contemporary studies suggest that the annual risk of sudden cardiac death (SCD) is in the range of 1%.[1,2] Some risk factors have been shown to be associated with an increased risk of SCD in patients with HCM.[1] The challenge for clinicians is to identify the small cohort of patients who are at a higher risk in order to target potentially lifesaving therapy such as implantable cardioverter defibrillators (ICD).[2] Fragmented surface electrocardiogram (ECG fQRS) has been identified as a useful risk marker in other clinical conditions;[3] however, its use in arrhythmic events in patients with HCM has not been suggested. We report a case of an 18-year-old girl with HCM and aborted SCD and implanted with an ICD. Her surface 12-lead ECG evolved from normal to fQRS complex in less than 7 years. During a 2-year follow-up, the ICD delivered several appropriate therapies to the malignant ventricular arrhythmias.

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CASE REPORT

An 18-year-old female was diagnosed with HCM at the age of nine. She presented with recurrent syncope. At that time, her 12-lead electrocardiogram (ECG) showed sinus rhythm, Q-waves and negative T-waves in leads V5 and V6 with a QRS duration < 120 ms. [Figure 1, left panel]. Her Echocardiogram depicted an asymmetrical left ventricular hypertrophy with an interventricular septum measuring 22 mm and no intraventricular gradient. During the screening, secondary causes of left ventricular hypertrophy such as systemic hypertension, congenital disease and valvular diseases were ruled out. A 24-hour ambulatory Holter monitoring showed frequent monomorphic premature ventricular contractions. Her first degree relatives were screened with ECG and echocardiogram, however, no phenotypic HCM was found. The patient remained asymptomatic under treatment with atenolol 50 mg/day for 7 years, when she suffered a cardiac arrest following an episode of emotional stress. Resuscitation was

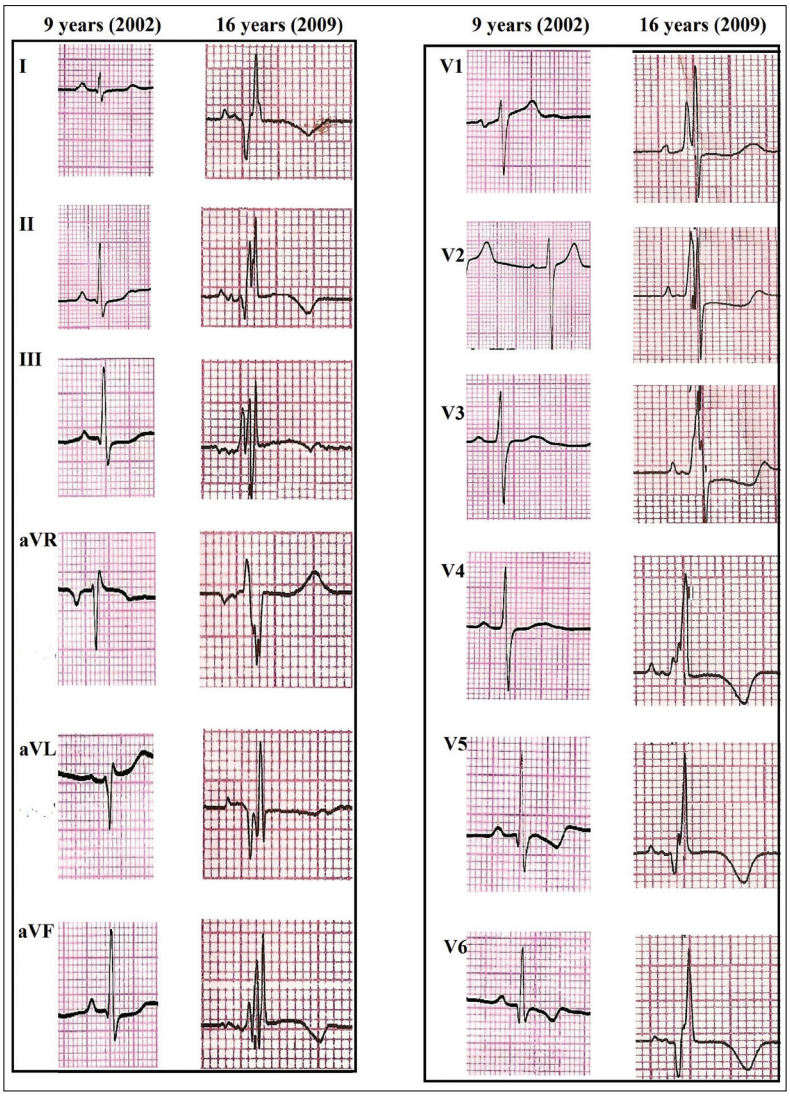

Figure 1: In the left panels, 12-lead electrocardiogram (ECG)at the age of nine (2002), depicting sinus rhythm, PR interval 160 ms, QTc 380 ms and Q-waves in leads V5, V6 with negative T-waves. In the right panel, 12-lead ECG on admission at the age of sixteen (2009), depicting sinus rhythm; PR interval 130 ms and delayed left atrial depolarization (the second vector of the P-wave is delayed, most likely by fibrosis of the interatrial septum); deep Q-waves in leads I, aVL, V5, and V6, diffuse T-wave inversion, QRS duration 130 ms and different morphologies of fQRS, including various RSR´ patterns

initiated by her mother, and continued by paramedics. Ventricular fibrillation (VF) was detected and terminated by an external electrical shock. During hospitalization, an echocardiogram showed massive left and right ventricular hypertrophy with a septal thickness of 38 mm and absence of intraventricular gradient [Figure 2]. The surface 12 lead ECG showed Q-waves in leads I, aVL, V5, V6, left ventricular strain pattern, inverted T-waves, and a QRS duration of 130 ms with fQRS in all leads. [Figure 1, right panel] A dual chamber ICD (Maximo DR 7278, Medtronic, Inc, MN, USA) was implanted. The patient was discharged and treated with atenolol 50 mg twice a day. During the two-year follow-up, the patient presented with sustained Ventricular tachycardia (VT) requiring ICD shocks. Anti-tachycardia pacing was effective in numerous occasions to convert VT into a normal sinus rhythm; however, on 2 occasions, shocks were necessary to resolve the arrhythmia. Atenolol dose was increased and the patient has remained asymptomatic since the last six months.

DISCUSSION

SCD is one of the most perplexing issues in the management of HMC. The stratification of risk in HCM in clinically stable patients is predicated on generally accepted noninvasive markers. The accepted risk factors in HCM include family history of ≥ 1 HCM-related SCDs, ≥ 1 episode of unexplained syncope, massive Left ventricular (LV) hypertrophy (thickness ≥ 30 mm); nonsustained VT on serial ambulatory 24-hour Holter,

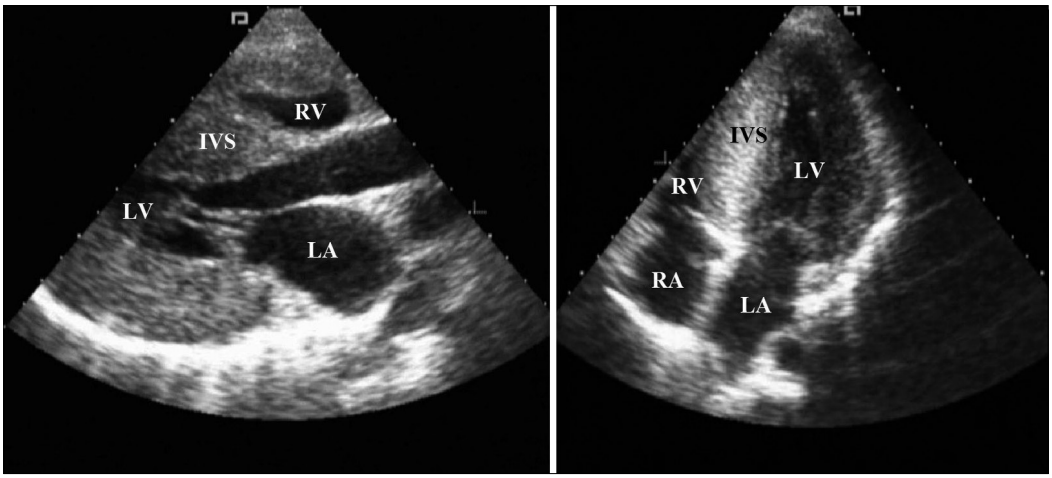

Figure 2: Parasternal large axis view (left) and apical four-chamber view (right) depicting global thickened of the left ventricular walls and right ventricle and interatrial septum involvement. IVS: interventricular septum; LA: left atria; LV: Left ventricle; RA: right atria; RV: right ventricle

and hypotensive or attenuated blood pressure response to exercise.[1,2] The ECG has traditionally been a part of the non-invasive evaluation for patients with HCM and though various electrocardiographic abnormalities were described, most of them correlated with the magnitude and extent of ventricular hypertrophy; however, did not reflecte increased risk *per se*. No specific criterion has hitherto been reported to be of value for risk stratification. Some investigators have attributed the disturbed myocardial electrical properties in HCM to the distinctive anatomical substrate of hypertrophied and disorganized myocytes.[4] Indeed, ventricular electrogram fractionation (delayed myocardial activation) has been documented by programmed ventricular extrastimulation in patients with HCM and induced VF; however, was absent in patients without VF.[4] Recently, Das *et al.*[3] described the presence of fQRS in patients with coronary artery disease (CAD). Patients with CAD and fQRS had more cardiovascular events and worse prognosis. The fQRS was defined in the 12-lead ECG as the presence of an additional R-wave (R') or notching in the nadir of the R-wave or the S-wave, or the presence of >1 R' (fragmentation) in 2 contiguous leads, corresponding to a major coronary artery territory. Fragmented QRS in the presence of wide QRS complexes (≥ 120 ms), such as bundle branch block (BBB), premature ventricular complexes (PVCs) and paced QRS complexes (pQRS), was defined by the presence > 2 notches in the R-wave or the S-wave, since BBB already have two notches or peaks present in two contiguous leads (3). fQRS has been associated with inhomogeneous activation of the ventricles due to myocardial scar and/or ischemia, which could predict arrhythmic events as well as death.[3] The underlying mechanisms of fragmentation have been supported by autopsy studies of patients with Myocardial infarction (MI) and left ventricular aneurysm. These studies showed that the presence of fQRS was associated with a significant myocardial necrosis alternating with viable myocardial tissue and interspersed in abundant fibrous tissue.[3,4] Distribution and pattern of the scar depends on the disease states. In CAD, myocardial scars are segmental and subendocardial or transmural, whereas in nonischemic cardiomyopathy the scars are patchy and mid myocardial or subepicardial, predominantly in the perivalvular areas.

In the presented case, myocardial scars are probably the cause of localized conduction block leading to an additional R´ or notching of the R – wave or S - wave. Endocardial and epicardial mapping in patients with CAD or dilated cardiomyopathy with ventricular arrhythmias have revealed fractionated electrograms over a wide area surrounding the myocardial scar.[3] fQRS is not specific for CAD, and was also observed in other myocardial diseases such as dilated cardiomyopathy, some congenital heart diseases, arrhythmogenic right ventricular cardiomyopathy and Brugada syndrome.[5,6]

To the best of our knowledge, this is the first report of a patient with HCM and rapidly progressive fQRS, with documented malignant ventricular arrhythmias treated by an ICD. The progressive fragmentation of the QRS and the presence of malignant ventricular arrhythmias, lead us to speculate on the value of fQRS as a predictor of ventricular arrhythmias in patients with HCM. Recently, Sherrid *et al.*,[7] in a long series of 330 patients with HCM and high-risk criteria of SCD with ICD implantation, compared the relationship of electrocardiographic findings and appropriate ICD therapies. During a follow-up of 3.7 ± 3.0 years, 17% of the patients had appropriate ICD discharges. The authors found no distinctive ECG

characteristics in patients with and without appropriate ICD interventions. Bongioanni *et al.*[8] found in 241 patients with HCM that a QRS duration ≥ 120 ms had an odds ratio of 5.2 for all-cause cardiovascular death; however, was not found to predict SCD. Hypothetically, the lack of correlation of malignant ventricular arrhythmias with ECG abnormalities supports the notion that SCD likely depends on transient electrophysiologic status and by interacting with the underlying substrate, such as vascular ischemia, neuroendocrine or electrolyte imbalance. Such transient injuries are consistent with the unpredictable nature of HCM-related SCD that may occur years after diagnosis, and often without any warning. In the presented case, the fQRS probably reflects a more severe myocardial disease, manifested by severe progression of ventricular hypertrophy. fQRS should be investigated in larger series of patients as a possible new risk factor and predictor of arrhythmic events in patients with HCM.

CONCLUSION

fQRS should be tested in a larger series of patients as a possible new risk factor and predictor of arrhythmic events in patients with HCM.

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