

CASE REPORT

Non-Cardiac Sudden Death in a Patient with Arrhythmogenic Right Ventricular Cardiomyopathy

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ABSTRACT

We herein present the case of a 70-year old lady with probable arrhythmogenic right ventricular cardiomyopathy (ARVC) and recurrent episodes of syncope. She was referred to our department due to an episode of sustained ventricular tachycardia (VT) which caused hemodynamic collapse and was converted electrically. During diagnostic investigation, echocardiography revealed evidence of right ventricular dysfunction. She underwent risk stratification and sustained monomorphic VT was easily induced during an electrophysiology study. An implantable cardioverter-defibrillator (ICD) device was subsequently implanted, but she died suddenly three months later. Interrogation of the ICD device did not reveal any arrhythmic event and death was attributed to carbon monoxide poisoning.

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KEY WORDS: *Sudden death; arrhythmogenic right ventricular cardiomyopathy; implantable cardioverter defibrillator*

ABBREVIATIONS

ARVC = arrhythmogenic right ventricular cardiomyopathy
CAD = coronary artery disease
CMRI = cardiac magnetic resonance imaging
ECG = electrocardiogram
EPS = electrophysiology study
ICD = implantable cardioverter defibrillator
LBBB = left bundle branch block
SAECG = signal averaged ECG
SCD = sudden cardiac death
VT = ventricular tachycardia

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INTRODUCTION

Sudden cardiac death (SCD) refers to the sudden cessation of cardiac activity with hemodynamic collapse occurring in a short time period (generally within 1 hour of symptom onset) in a person with known or unknown cardiac disease. Most cases of SCD are related to cardiac arrhythmias. SCD accounts for approximately 15% of the total mortality in industrialized countries.¹

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an underrecognized clinical entity characterized by ventricular arrhythmias and a specific ventricular pathology. It is a leading cause of sudden death among young athletes, although the condition can affect people of all ages and activity levels. Its prevalence in the general population is estimated to be approximately 1:1000.²

CASE REPORT

A 70-year-old Caucasian female was hospitalized for acute dyspnea, associated with

a hypertensive crisis, in a local hospital. Upon discharge she experienced an episode of sustained ventricular tachycardia (VT) and subsequent hemodynamic collapse. The tachycardia was converted with a direct current shock of 360 joules delivered via an external defibrillator. Subsequently, she was readmitted and then transferred to our tertiary cardiology department for further investigation. Six years earlier she had suffered a similar episode and during that hospitalization she had undergone coronary catheterization with no signs of coronary artery disease (CAD).

Her past medical history was remarkable for hypertension, diabetes mellitus, dyslipidemia and thyroidectomy, receiving

respective therapies (Table 1). She was also taking carvedilol, lisinopril, and furosemide for hypertension and was recently placed on amiodarone, right after the episode of sustained VT. The patient had a strong family history of SCD, as all three of her siblings had died suddenly at ages varying from 32 to 55 years old.

On physical examination, the patient was afebrile, with a blood pressure of 120/80 mmHg and a heart rate of 60 beats per minute. Her clinical examination and chest x-ray were unremarkable. ECG showed sinus rhythm and prolonged QRS duration, with left bundle branch block (LBBB) pattern (Figure 1).

No particular findings were noted on her basic laboratory testing, which included complete cell blood count, glucose and electrolyte levels, kidney and liver function tests and lipid profile. Thyroid-stimulating hormone levels were within normal range and cardiac troponin-I was only traceable (0.09 ng/ml).

Echocardiography revealed mild dilatation of the left atrium and mild concentric hypertrophy of the left ventricle, along with mild diastolic dysfunction. Evidence of systolic and diastolic dysfunction of the right ventricle were present and the right ventricular free wall was found to be hypertrophic and echodense. Ejection fraction was measured to be approximately 45% by cardiac magnetic resonance imaging (CMRI), but the exam did not elicit any other particular findings. More specifically, other than a moderate left atrial dilatation and a mild left ventricular hypertrophy, no areas of abnormal signal or gadolinium enhancement were spotted in either the left or the right ventricle, and no diffuse or segmental wall motion

TABLE 1. Oral Medications

Medication	Dose
Lisinopril	20 mg qd
Furosemide	40 mg qd
Carvedilol	6.25 mg bid
Amiodarone	200 mg qd
Aspirin	100 mg qd
Simvastatin	20 mg qd
Metformin	850 mg qd
Glimepiride	4 mg qd
Levothyroxine	0.1 µg qd

bid = twice daily; qd = once daily

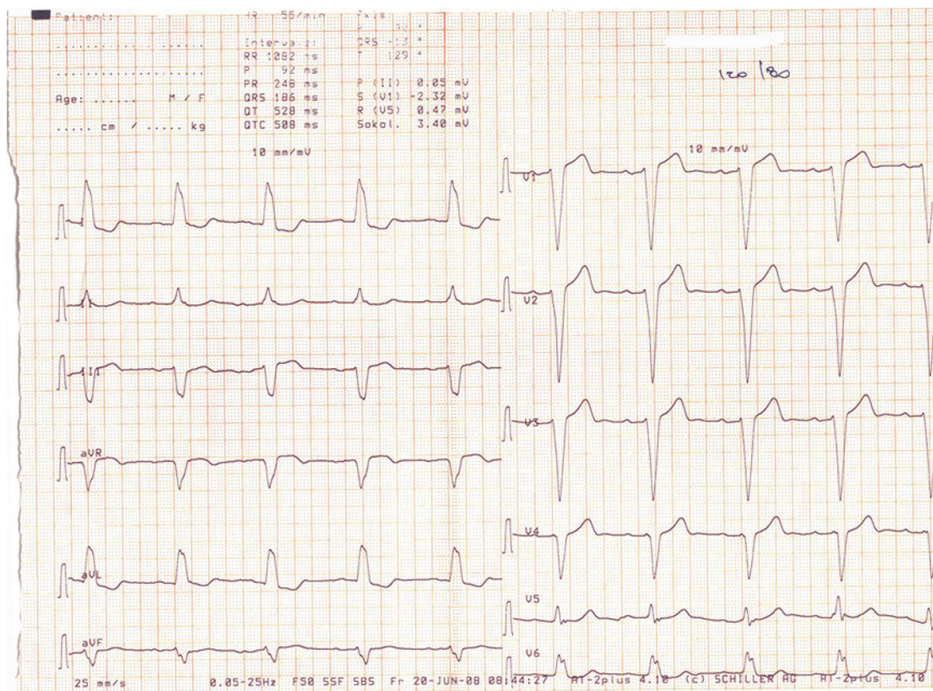


FIGURE 1. Patient's baseline electrocardiogram (ECG) showing a left bundle branch block (LBBB).

abnormalities were noted in the left ventricular myocardium.

Due to the history of sustained VT, further investigation and risk stratification for SCD was deemed necessary, as there was a discrepancy between echocardiographic and CMRI findings. One episode of sinus pause was identified on a 24-hour holter monitor with duration of 1.8 seconds along with multiple premature ventricular complexes (>1500) (Figure 2). Signal averaged ECG (SAECG) was positive for late potentials, but with limited value due to underlying LBBB (Figure 3). Further investigation included interventional testing. Coronary angiography revealed patent coronary arteries. During an electrophysiology study (EPS), monomorphic sustained VT was induced with a single premature stimulus delivered at

the right ventricular apex and was reverted with overdrive pacing (Figures 4a, 4b and 4c). Interestingly, right after the conversion of the arrhythmia, the patient had evidence of 2:1 atrioventricular block (Figure 4d) causing hemodynamic instability and requiring admission to the intensive care unit and the insertion of a temporary pacemaker. Based on these findings, a decision was then made to implant a cardioverter defibrillator (ICD) device, which was performed successfully on the following day (Figure 5).

Two months later, the patient's husband called their children and asked for help, as the patient had a new episode of syncope. Upon their arrival, both parents were found dead, with a cherry-red discoloration in their skin. Device interro-

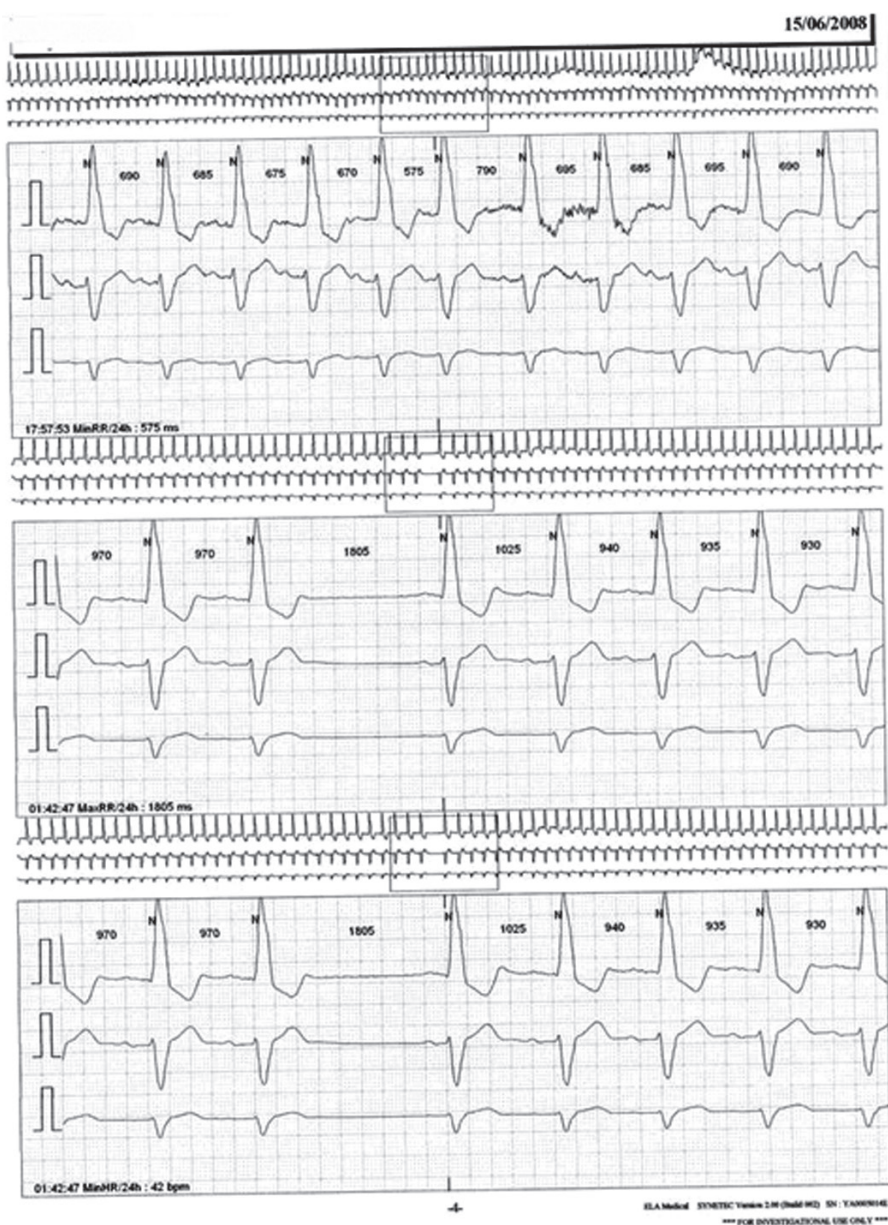


FIGURE 2. Holter monitoring revealed only a non-significant pause of 1.8 sec during sleep.

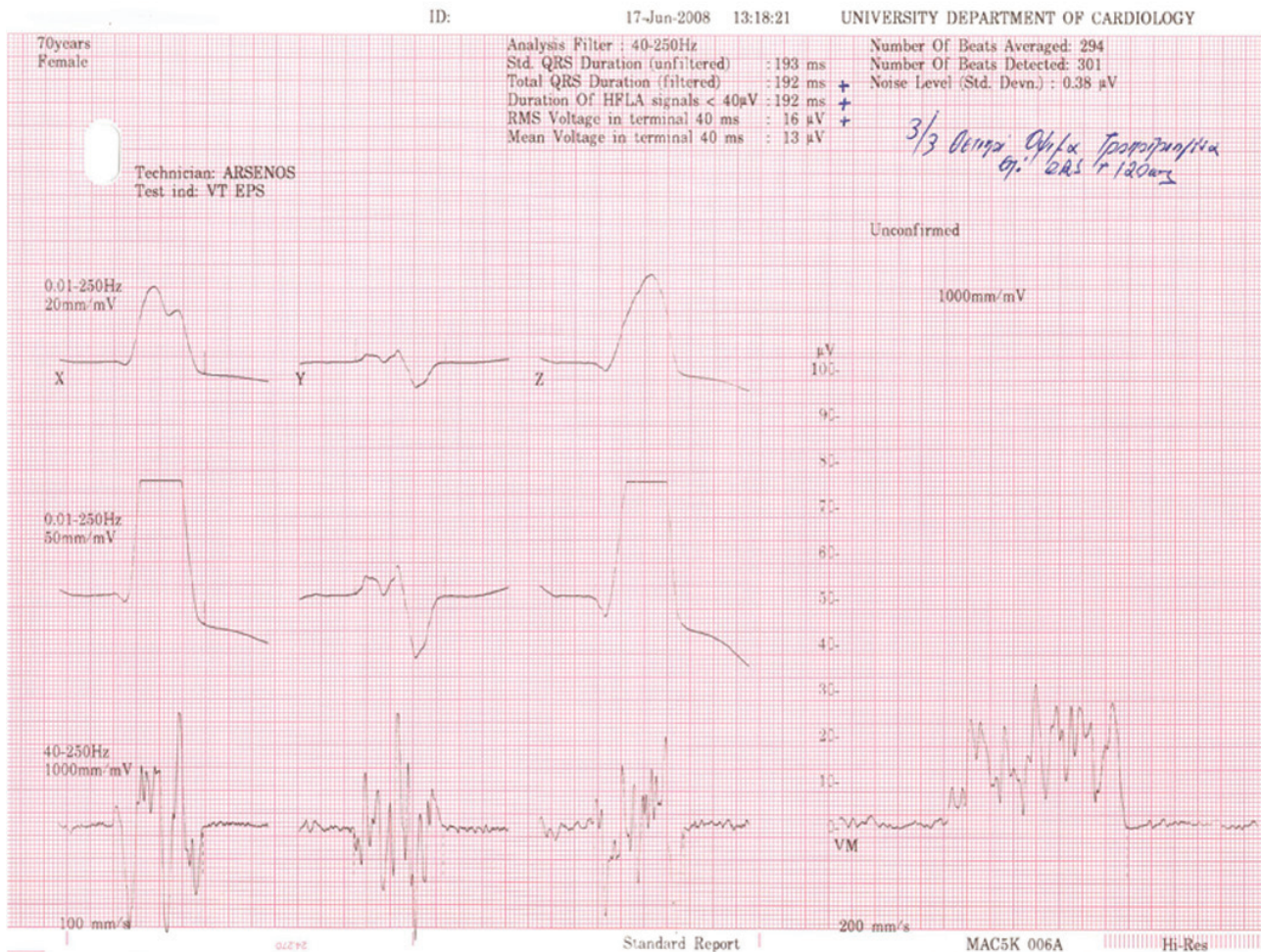


FIGURE 3. Signal averaged ECG was positive for late potentials with use of modified criteria, which though remain questionable due to underlying LBBB.

gation in the patient did not reveal any arrhythmic event and the ICD had not delivered any mode of therapy (Figure 6). This incident occurred just one week after the patient's last follow up visit in the outpatient device clinic, where sensing parameters of the ICD were found normal. The simultaneous death of the patient and her husband was finally attributed to carbon monoxide poisoning, as the couple were using a brazier for heating.

DISCUSSION

In this case, the diagnosis of ARVC could not be established according to the task force criteria. In more detail, the patient had an episode of VT with LBBB morphology, according to the report from the hospital from which she was transferred to our department, and family history consistent

with SCD due to suspected ARVC. The regional right ventricular hypokinesia on echocardiography raises suspicion for right ventricular pathology but does not fulfill the diagnostic criteria. The presence of late potentials on SAECG could not be considered a criterion, due to the presence of LBBB.³⁻⁴

On the other hand, no alternative diagnosis seemed more appropriate for this case. Coronary artery disease had been ruled out by coronary angiography. There was no evidence of other types of cardiomyopathy, congenital heart disease, or of significant valvular disease on both imaging studies (echocardiography and CMRI). The findings from the ECG were not compatible with preexcitation syndrome or a channelopathy associated with SCD. Idiopathic right ventricular tachycardia could be an alternative diagnosis, but the history of resuscitation, the echocardiographic findings, the positive family history for SCD and the easy induction of VT during EPS, favor the diagnosis of ARVC over idiopathic right ven-

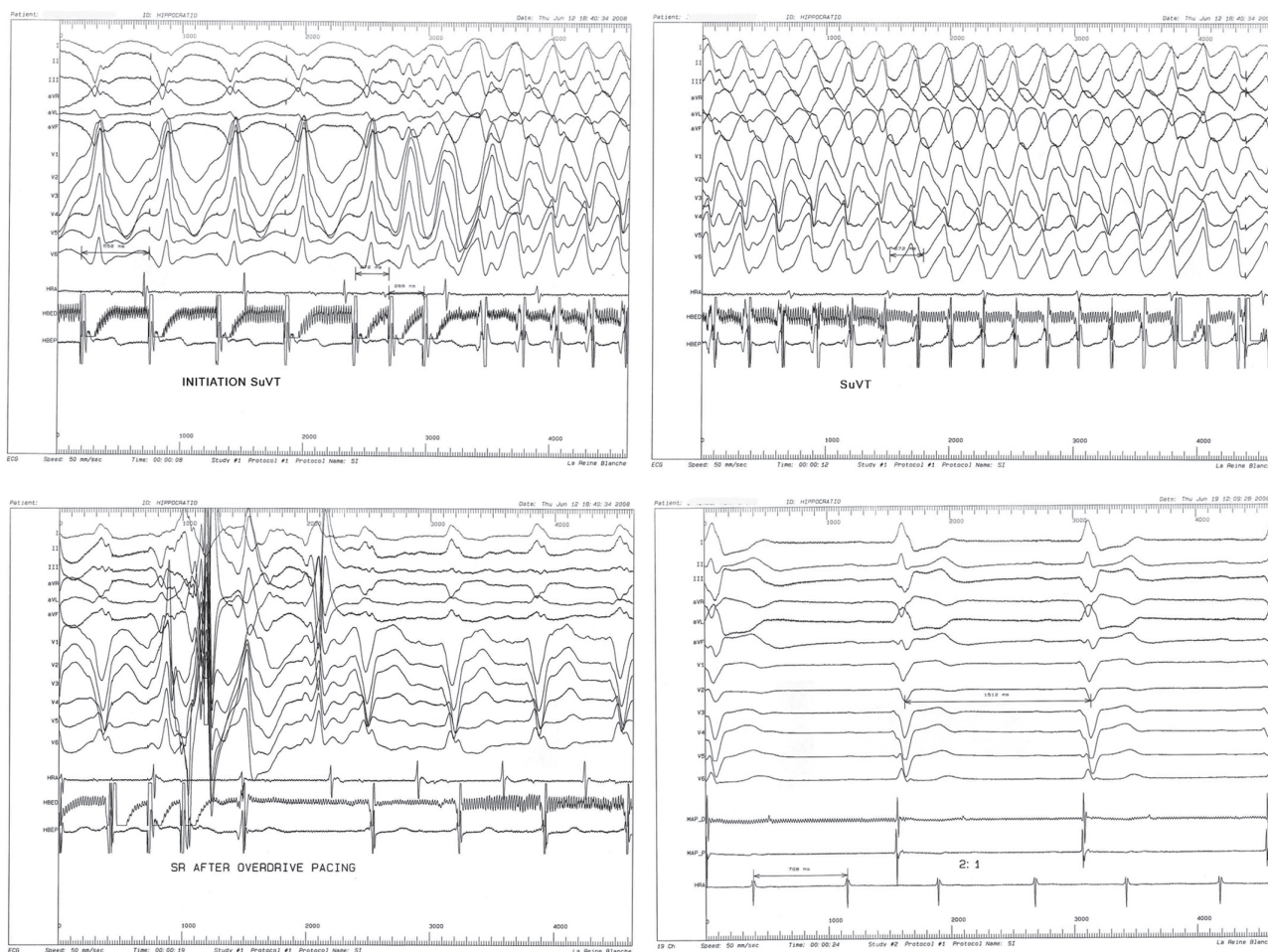


FIGURE 4. During the electrophysiology study (EPS), sustained monomorphic ventricular tachycardia (SuVT) was easily induced with the application of one ventricular extrastimulus at the right ventricular apex (**a** & **b**). The tachycardia was terminated with overdrive ventricular pacing and sinus rhythm (SR) was restored (**c**). However, shortly after sinus rhythm was restored, 2:1 atrioventricular block developed (**d**).

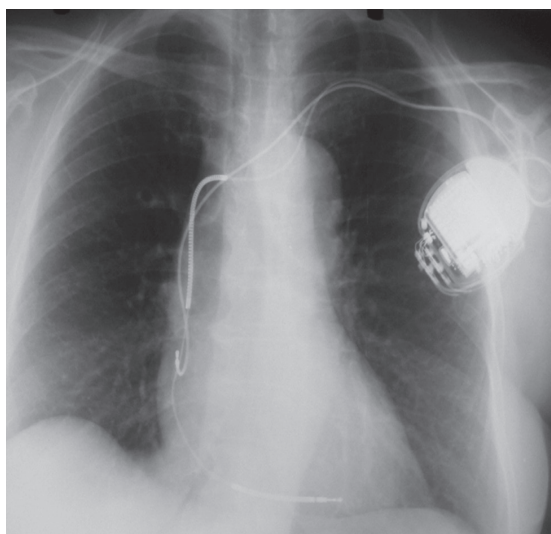


FIGURE 5. Chest X-ray after ICD implantation indicating two endocardial leads, one J-atrial pacing (placed at the right atrial appendage) and one pacing/defibrillating ventricular lead placed at the right ventricular apex. The ICD pulse generator is placed at the left infraclavicular area. ICD = implantable cardioverter defibrillator.

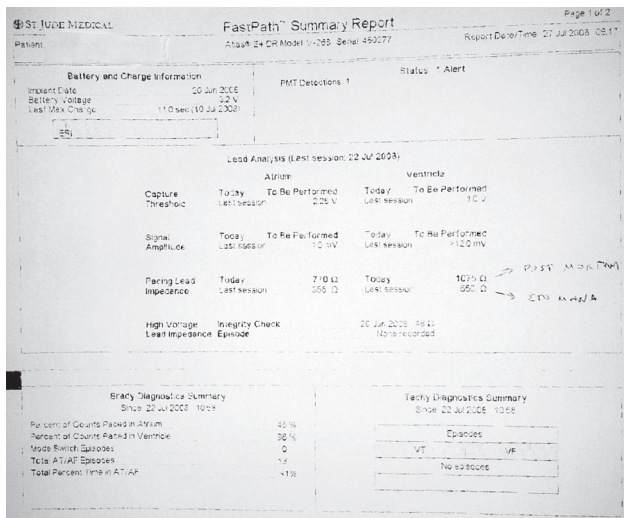


FIGURE 6. Post mortem ICD device interrogation indicated no detectable arrhythmias.

tricular tachycardia.⁵

The interesting fact is that, despite high clinical suspicion for cardiomyopathy, the diagnosis of ARVC could not be established according to current criteria and at the same time the patient was at high risk for SCD, according to risk stratification with EPS. This underlines the fact that even patients with only a few minor criteria could still be at high risk.

There is a high incidence of ventricular arrhythmias in ARVC patients, which is estimated to be 23% in those with mild disease, 82% with moderate and practically in all patients with severe disease.⁶ However, ARVC presents with syncope less frequently, with an overall incidence of 32%,⁷ and should be included in the differential diagnosis of syncope, especially in children and adolescents.

The 2006 ACC/AHA/ESC guidelines suggest the implantation of an ICD device in patients with documented sustained VT or ventricular fibrillation, for secondary prevention, just like in our case, and in those patients who are felt to be at

high risk for primary prevention.⁸ Nonetheless, what initially seemed to be a failure of an appropriately implanted ICD device to deliver the necessary therapy, it turned out to be the result of an unexpected environmental hazard.

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