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FOCUS ISSUE ON SPORTS CARDIOLOGY

INTERMEDIATE

CASE REPORT: CLINICAL CASE

Idiopathic Ventricular Fibrillation in a Previously Healthy Recreational Athlete

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ABSTRACT

There are several clinical challenges in the survivor of sudden cardiac arrest (SCA), including ensuring that a comprehensive diagnostic evaluation has been performed and providing counseling on return to activity. We report a case of a highly conditioned athlete who presented following aborted SCA during exercise with a diagnosis of idiopathic ventricular fibrillation arrest. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:1129–1133)

HISTORY OF PRESENT ILLNESS

A 40-year-old previously healthy man presented to our clinic 3 weeks after an aborted sudden cardiac arrest (SCA) episode. While running a 5-km race, he collapsed, and an automated external defibrillator delivered a shock, leading to prompt return of spontaneous circulation. The patient was highly active overall, and 2 months before this current episode he had performed a 26-mile marathon race. He ran several miles on multiple days per week and was also an avid mountain biker.

LEARNING OBJECTIVES

- To discuss the differential diagnosis for the origin of SCA in the athlete and the importance of performing a comprehensive evaluation to determine the cause.
- To discuss the current available data on return to activity in patients who have experienced SCA and to use this information in shared decision making.

PAST MEDICAL HISTORY

The patient had neither any known medical diagnosis nor daily medication or supplement use. He denied a history of alcohol use, illicit drug use, or tobacco use. He had no first- or second-degree relatives with unexplained death.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for the origin of SCA is broad, and it can be categorized into primarily structural, electrical, or acquired cardiac abnormalities. The most common substructural abnormality manifesting with SCA is coronary artery disease with acute coronary syndrome (ACS), and it is critical to evaluate patients with a postresuscitation electrocardiogram rapidly.[1] Other common causes of SCA are listed in **Figure 1**.

INVESTIGATIONS

The patient was awake, alert, and following commands after resuscitation. His initial post-resuscitation electrocardiogram (ECG) was reported

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ABBREVIATIONS AND ACRONYMS

ACS = acute coronary syndrome

ECG = electrocardiogram

ICD = implantable cardioverter-defibrillator

SCA = sudden cardiac arrest

VF = ventricular fibrillation

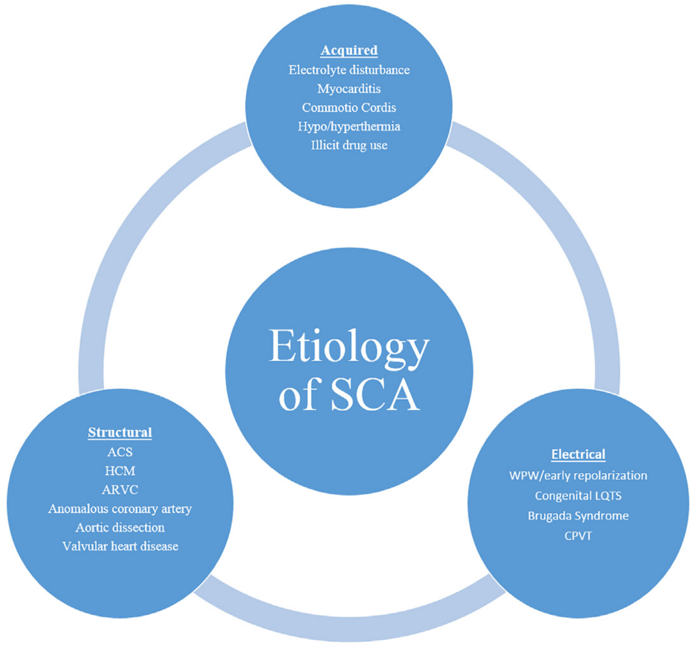
as sinus rhythm without evidence of ST-segment changes. He underwent additional extensive evaluation for the origin of his SCA, including echocardiography, invasive coronary angiography, and cardiac magnetic resonance, without a clear cause identified. His testing results and initial laboratory results are summarized in Tables 1 and 2.

MANAGEMENT

The patient was referred for transvenous implantable cardioverter-defibrillator (ICD) placement. He was discharged home with a plan for follow-up at our center after discharge.

The patient was evaluated 3 weeks after his SCA presentation. Other than occasional palpitations, he did not report any symptoms or new medications since his hospital discharge. His ECG from the clinic is presented in Figure 2A (Figures 2B to 2G are comparison pathologic ECG examples). One of his main concerns involved when he could return to regular physical activity. He was noted on initial device interrogation to have no episodes of sustained or nonsustained ventricular arrhythmias. He was scheduled for an invasive electrophysiology study with a procainamide infusion to assess for ECG changes suggestive of Brugada syndrome, and the results are summarized in Table 1.

FIGURE 1 Differential Diagnosis of Causes of SCA



ACS = acute coronary syndrome; ARVC = arrhythmogenic right ventricular cardiomyopathy; CPVT = catecholaminergic polymorphic ventricular tachycardia; HCM = hypertrophic cardiomyopathy; LQTS = long QT syndrome, SCA = sudden cardiac arrest; WPW = Wolff-Parkinson-White syndrome. Adapted with permission from Chandra N, Bastiaenen R, Papadakis M, Sharma S. Sudden cardiac death in young athletes: practical challenges and diagnostic dilemmas. *J Am Coll Cardiol.* 2013;61(10):1027-1040.

DISCUSSION

There are multiple challenges in managing patients after a diagnosis of idiopathic ventricular fibrillation (VF). It is imperative that a comprehensive investigation is performed to determine the cause of SCA. In patients without ACS or apparent structural heart disease, a structured approach consisting of tiered diagnostic testing should be performed before a diagnosis of idiopathic VF arrest is made (Figure 3).[1-3] The patient in this case appropriately underwent ICD implantation for secondary prevention before hospital discharge.[4] The decision to proceed with transvenous ICD placement was made before the patient received care at our center. Strong consideration should have been made for placement of a subcutaneous ICD system, given the risk of long-term complications associated with a transvenous system in a relatively young patient without anticipated pacing requirements.

Among the patient's primary concerns were recommendations on returning to regular physical activity. The initial recommendations, which were made predominantly on the basis of expert opinions, restricted all moderate to high-intensity exercise in patients with prior SCA with secondary prevention ICD in place.[5] However, data subsequently emerged that challenged these recommendations.[6] A study was published in 2013 that evaluated the outcome of 372 patients with an ICD who were competing in high-risk sports and competitive athletics over a median 31-month follow-up period.[7] There were no episodes of tachyarrhythmic death, resuscitated tachyarrhythmias during or after sports activity, or severe injury secondary to syncope or shock during sports in this group.[7]

Restricting patients from exercise may also carry unintended morbidity. In a study published in 2018, 366 athletes were retrospectively identified with genetic cardiac disease and were previously restricted from further competitive sports activity, with 44 patients self-disqualifying and 322 patients continuing to participate, without statistically significant differences in the groups.[8] Interestingly, only 9 of the 322 (3%) athletes who continued to participate in sports experienced nonlethal cardiac events (4 occurred outside of athletics) in 961 combined athlete-years,

TABLE 1 Diagnostic Testing Performed in Patient

Test	Result
Transthoracic echocardiogram (hospital day 1)	Normal LV size and systolic function with ejection fraction 55%-60%; mild concentric LVH; normal right ventricular size and systolic function; no significant valve disease
Left-sided heart catheterization (hospital day 1)	Normal coronary arteries without evidence of coronary artery disease; LV ejection fraction estimated at 55%-60% by left ventriculogram
Cardiac magnetic resonance (hospital day 5)	Normal biventricular size and systolic function; no evidence of LVH; no evidence of late gadolinium enhancement; no regional wall motion abnormalities or scarring; no criteria for arrhythmogenic right ventricular cardiomyopathy; LV end-diastolic volume index measured at 89 mL/m^2; end-systolic volume index measured at 38 mL/m^2
Exercise treadmill testing (hospital day 6)	Performed estimated 14.8 METs; reached peak heart rate of 152 beats/min (85% of maximum predicted heart rate); no evidence of exercise-induced ectopy or arrhythmias; appropriate shortening of the QT interval with exercise.
Holter monitor (~3 mo post-discharge)	72-h Holter monitor worn, showing no significant premature ventricular contraction burden; premature atrial contractions with aberrant conduction (estimated at ~1% of overall burden); 3 episodes of atrial fibrillation, which appeared to regularize into atrial flutter with the fastest ventricular rate of 163 beats/min (longest duration, ~2 h)
Electrophysiology study (~6 mo post-discharge)	1. Normal baseline intervals, including normal HV interval (43 ms) 2. No accessory pathways; no dual AV nodal physiology 3. No spontaneous or inducible ventricular arrhythmias under our standard protocol for ventricular arrhythmia induction, which includes the following: administering ventricular extrastimuli (up to 3 extrastimuli) at 2 separate anatomical locations (typically right ventricular outflow tract and right ventricular apex) at 2 different pacing drive train lengths; performance of 10- to 20-ms decrements in the ventricular extrastimuli until the ventricular effective refractory period or until 200-ms coupling interval reached; isoproterenol then initiated at a dose of 1 µg/min and titrated until heart rate response achieved (up to dose of 5 µg/min); protocol then repeated during the administration of isoproterenol 4. Inducible atrial flutter prompting CTI ablation; nonsustained inducible atrial fibrillation 5. Procainamide challenge failing to induce type 1 Brugada pattern on ECG

AV = atrioventricular; CTI = cavotricuspid isthmus; ECG = electrocardiogram; LV = left ventricular; LVH = left ventricular hypertrophy.

whereas 6 of the 44 (14%) former athletes experienced cardiac events (P = 0.03 on Kaplan-Meier analysis).

On the basis of the results from these studies, updated guidelines recommend that patients with ICDs inserted for secondary prevention may consider a return to higher-intensity activity if they have been free of ventricular tachycardia or VF for 3 months.[9] The importance of shared decision making with patients on return to play is emphasized in updated guidelines.

FOLLOW-UP

The patient presented for a follow-up visit approximately 6 weeks following his electrophysiology study. He reported resolution of palpitations, with no episodes of ventricular arrhythmias or ICD shocks. The patient declined genetic testing to evaluate for genetic cardiomyopathy and arrhythmia syndromes. We engaged in shared decision making regarding his return to regular physical activity. We advised against returning to mountain biking, given the risk of serious injury with syncope. We also recommended exercising in a semisupervised environment with an available automated external defibrillator and with a partner trained in cardiopulmonary resuscitation.

CONCLUSIONS

We present the case of a patient who experienced aborted SCA while running in a 5-km race. The patient underwent extensive evaluation for the cause of his SCA without a clear origin, and he received a diagnosis of idiopathic VF arrest. It can be challenging to provide return to activity recommendations for the

TABLE 2 Initial Laboratory Evaluation Results From Presentation[a]

Test	Result	Reference Range
Sodium	137 mmol/L	130-143 mmol/L
Potassium	3.7 mmol/L	3.2-4.8 mmol/L
Magnesium	2.0 mg/dL:	1.7-2.5 mg/dL
Chloride	100 mmol/L	95-108 mmol/L
Bicarbonate	**20 mmol/L**	23-33 mmol/L
Glucose	**247 mg/dL**	80-115 mg/dL
Blood urea nitrogen	16 mg/dL	7-24 mg/dL
Creatinine	1.4 mg/dL	0.5-1.5 mg/dL
Total protein	6.5 g/dL	6.4-8.2 g/dL
Total bilirubin	4.2 g/dL	2.8-4.9 g/dL
Aspartate transaminase	**139 IU/L**	15-53 IU/L
Alanine transaminase	**176 IU/L**	15-57 IU/L
Alkaline phosphatase	89 U/L	30-140 U/L
Lactate	**8.9 mmol/L**	0.5-1.6 mmol/L
High-sensitivity troponin I	**21 pg/mL**	0-20 pg/mL
B-type natriuretic peptide	32 pg/mL	<100 pg/mL
Thyroid-stimulating hormone	3.67 µIU/mL	0.45-5.33 µIU/mL
Arterial blood gas (pH/P_{CO_2} [mm Hg]/P_{O_2} [mm Hg])	7.44/36/64	7.38-7.45/35-45/83-108
White blood cell count	**14,700/µL**	4,000-10,000/µL
Hemoglobin	16.6 mg/dL	12.5-17 mg/dL
Hematocrit	48.3%	36%-50%
Platelet count	278,000/µL	140,000-450,000/µL

[a]All abnormal results highlighted in **bold**. All abnormalities were attributed to postarrest status and normalized after admission.

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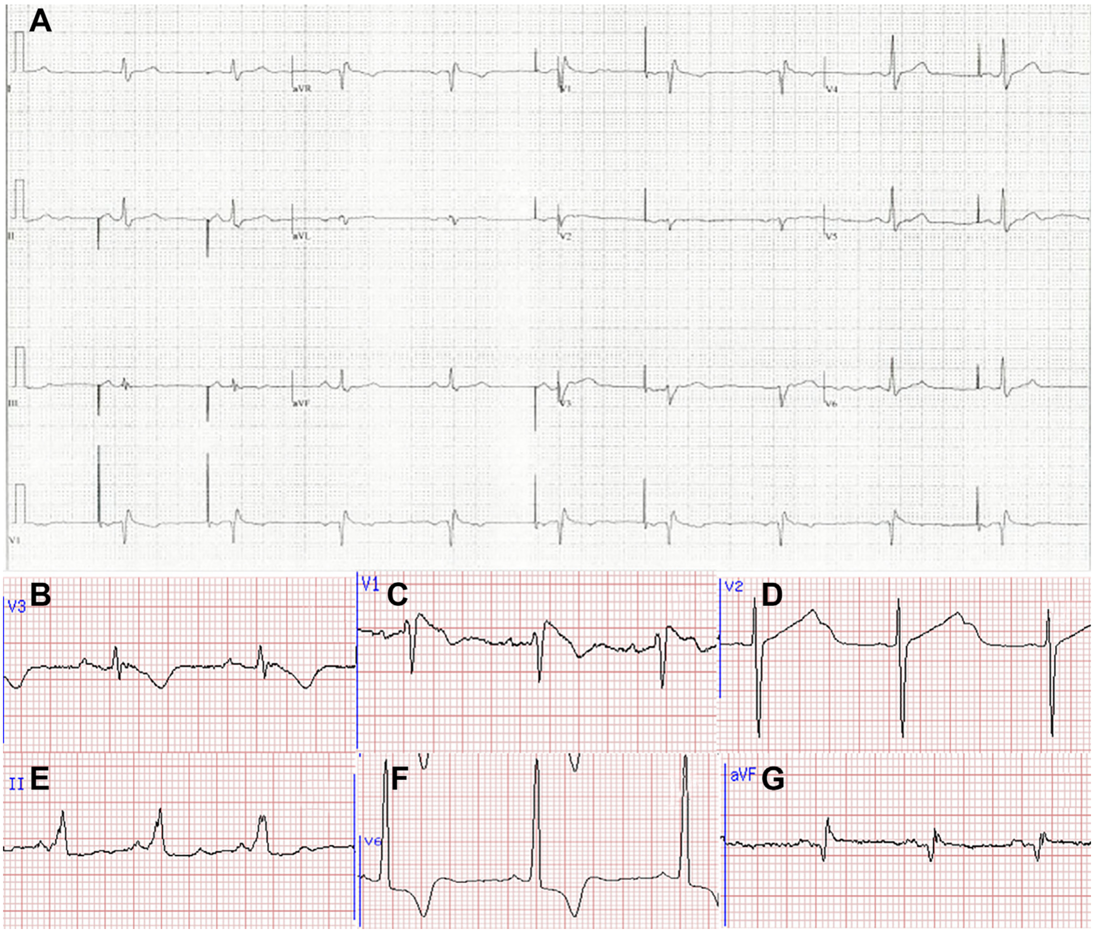
FIGURE 2 **Electrocardiographic Evaluation**



(A) The 12-lead electrocardiogram obtained from the patient's clinic evaluation, showing intermittent atrial pacing with incomplete right bundle branch block and down-sloping of the ST-segment in lead V_1. **(B to G)** Other important electrocardiographic patterns to recognize, including **(B)** arrhythmogenic right ventricular cardiomyopathy with epsilon waves, **(C)** type I Brugada pattern, **(D)** prolonged QT syndrome, **(E)** pre-excitation pattern, **(F)** hypertrophic cardiomyopathy with left ventricular hypertrophy and deep T-wave inversions, and **(G)** previous inferior infarction with pathologic Q waves. **(B to G)** Used with permission from Nathanson LA, McClennen S, Safran C, Goldberger AL. ECG wave-maven: self-assessment program for students and clinicians. Accessed March 13, 2022. http://ecg.bidmc.harvard.edu

athlete who has SCA that is diagnosed with idiopathic VF arrest. Although it is not necessary to disqualify the patient from returning to activity, it is imperative to engage in shared decision making with the patient.

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Dr Cooper has reported consulting and advisory board membership for Medtronic and Boston Scientific; and has provided fellow lectures for Abbott. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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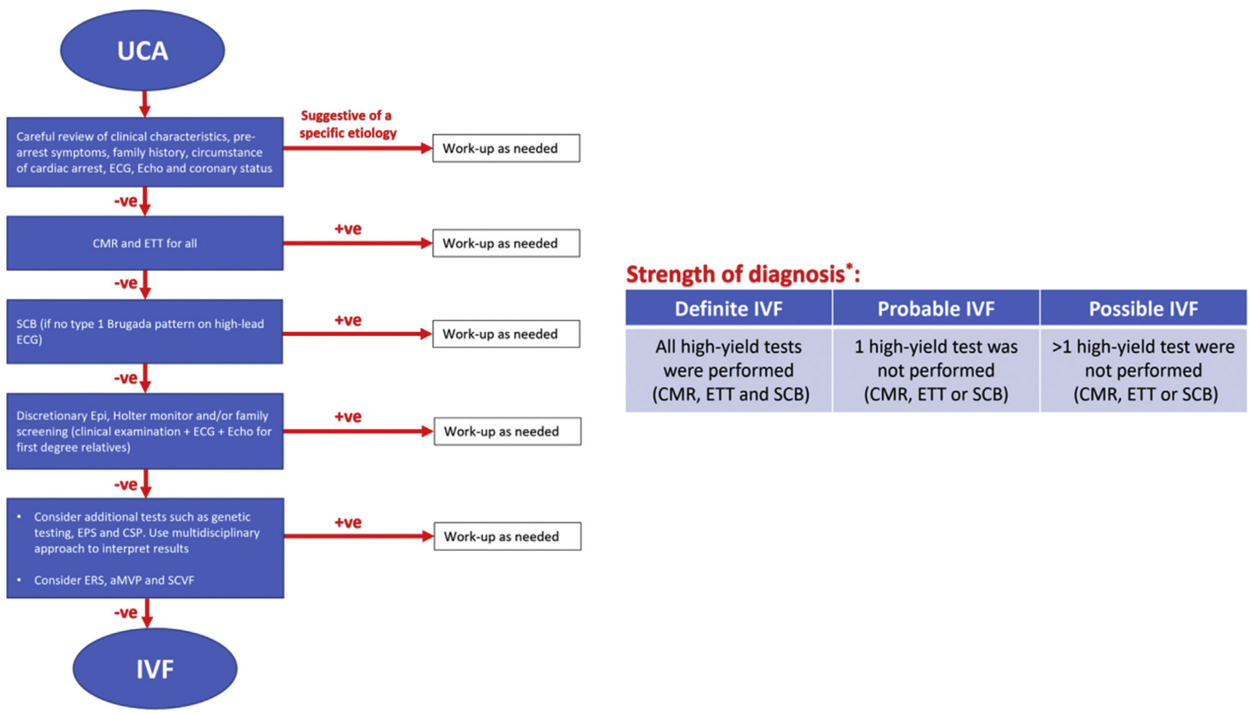
FIGURE 3 Diagnostic Algorithm to Approach the Patient After Sudden Cardiac Arrest, Organized by Highest-Yield Testing



aMVP = arrhythmic mitral valve prolapse; CMR = cardiac magnetic resonance imaging; CSP = coronary spasm provocation; ECG = electrocardiogram; Echo = echocardiogram; Epi = epinephrine challenge test; EPS = electrophysiology study; ERS = early repolarization syndrome; ETT = exercise treadmill testing; IVF = idiopathic ventricular fibrillation; SCB = sodium channel blockade; SCVF = short-coupled ventricular fibrillation; UCA = unexplained cardiac arrest; -ve = negative; +ve = positive. Reprinted with permission from Alqarawi W, Dewidar O, Tadros R, et al. Defining idiopathic ventricular fibrillation: a systematic review of diagnostic testing yield in apparently unexplained cardiac arrest. *Heart Rhythm.* 2021;18(7):1178-1185.

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KEY WORDS idiopathic ventricular fibrillation, implantable cardioverter-defibrillator, sudden cardiac arrest