Idiopathic Ventricular Fibrillation in a Previously Healthy Recreational Athlete

Caleb Norton, MD, Daniel H. Cooper, MD, Gregory Ewald, MD, Mustafa Husaini, MD

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) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

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.

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 postresuscitation electrocardiogram (ECG) was reported...
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.

**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,
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 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/m²; end-systolic volume index measured at 38 mL/m² |
| 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 = cavo-tricuspid 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. 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

**TABLE 2 Initial Laboratory Evaluation Results From Presentation**

| 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 µU/mL | 0.45-3.33 µU/mL |
| Arterial blood gas (pH/PCO₂ [mm Hg]/PO₂ [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 |

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

**FUNDING SUPPORT AND AUTHOR DISCLOSURES**

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.

**ADDRESS FOR CORRESPONDENCE:** Dr Caleb Norton, Division of Cardiovascular Disease, Washington University in St Louis, 660 South Euclid Avenue, CB 8086, St Louis, Missouri 63110, USA. E-mail: calebn@wustl.edu. Twitter: @cardscalebMD.
REFERENCES

1. Marijon E, Uy-Evanado A, Reinier K, et al. Sudden cardiac arrest during sports activity in middle age. Circulation. 2015;131(16):1384–1391.

2. Krahn AD, Healey JS, Chauhan V, et al. Systematic assessment of patients with unexplained cardiac arrest: Cardiac Arrest Survivors With Preserved Ejection Fraction Registry (CASPER). Circulation. 2009;120(4):278–285.

3. 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.

4. Al-Khatib SM, Stevenson WG, Ackerman MJ, et al. 2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2018;72(14):e91–e220.

5. Maron BJ, Zipes DP. Introduction: eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities-general considerations. J Am Coll Cardiol. 2005;45(8):1318–1321.

6. Lampert R, Cannon D, Olshansky B. Safety of sports participation in patients with implantable cardioverter defibrillators: a survey of Heart Rhythm Society members. J Cardiovasc Electrophysiol. 2006;17(1):11-15.

7. Lampert R, Olshansky B, Heidbuchel H, et al. Safety of sports for athletes with implantable cardioverter-defibrillators: results of a prospective, multinational registry. Circulation. 2013;127(20):2031-2030.

8. Turkowski KL, Bos JM, Ackerman NC, Rohatgi RK, Ackerman MJ. Return-to-play for athletes with genetic heart diseases. Circulation. 2018;137(10):1086–1088.

9. Zipes DP, Link MS, Ackerman MJ, et al. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 9: Arrhythmias and Conduction Defects: a scientific statement from the American Heart Association and American College of Cardiology. J Am Coll Cardiol. 2015;66(21):2412–2423.

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