Hypertrophic Cardiomyopathy in a Young Athlete at Risk for Sudden Cardiac Death

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32.1 Case Summary

A 15 year-old male presented to the electrophysiology clinic for evaluation for primary prevention implantable cardioverter defibrillator (ICD) after being diagnosed with hypertrophic cardiomyopathy (HCM). The patient was first diagnosed at age 13 on screening transthoracic echocardiogram (TTE) and diagnosis was confirmed on cardiac magnetic resonance imaging (MRI), which measured a septal thickness of 1.9 cm. The patient was asymptomatic and at first diagnosis had no risk factors for sudden death, including no nonsustained ventricular tachycardia, family history of sudden cardiac death (SCD), syncope, or hypotensive response on exercise tolerance testing. Follow-up cardiac MRI at nearly 15 years of age demonstrated a marked increase of septal thickness, which had increased to 3.4 cm, as well as left ventricular scarring quantified at 14%, prompting referral for implantable cardioverter defibrillator (ICD) evaluation. Additional testing revealed no additional new risk factors for SCD.

The patient and his family were offered the option of proceeding with ICD implantation, understanding that the patient had not reached adulthood and wished to continue participating in approved sports. After an extensive discussion of the risks, benefits, and alternatives to ICD implantation, the patient and his family elected to proceed with an ICD. What options for ICD implantation can be offered to this young patient to best tolerate physical activity and maximize lead longevity, and how can programming of his device be optimized to ensure successful termination of ventricular fibrillation while minimizing inappropriate shocks, both of which he will be at risk for during exercise?

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32.2 Case Discussion

The patient is at increased risk for SCD due to massive left ventricular hypertrophy and therefore has a Class IIa AHA 2011 HCM Guideline indication for primary prevention ICD implantation [1]. By the ESC HCM calculator he had a 2.78% 5-year SCD risk. Additionally, the significant amount of left ventricular scar, which was quantified at 14%, may elevate his risk. Recent prospective data has shown that scar involving ≥15% of the myocardium is an independent risk factor for SCD and in otherwise asymptomatic patients without typical risk factors, likely increased the risk of SCD twofold [2].

The subcutaneous ICD (S-ICD), FDA approved in 2012, utilizes a lead positioned in the subcutaneous layer of the thoracic cage to sense and terminate malignant arrhythmias. The subcutaneous location of the lead reduces potential complications related to wear, making this an attractive option for patients with HCM, who are typically younger and may wish to exercise [3]. While the device does not have pacing capabilities, many HCM patients, including the young patient presented, do not have any pacing indications. For these reasons, the S-ICD was selected by the patient and his family.

Despite the potential physical advantages of the S-ICD in this patient population, further screening is required to ensure appropriate sensing. There are also specific challenges that require particular attention in selection and programming. First, QRS and T-wave oversensing and myopotential oversensing may be more common in HCM and may be further exacerbated during exercise, resulting in inappropriate shocks [3, 4]. Second, patients with HCM may have higher defibrillation thresholds (DFTs) due to increased myocardial mass, which may progress over time.

Patients are initially screened for S-ICD using a manufacturer-provided tool that analyzes surface ECG recordings via three vectors taken while lying, standing, and immediately after running in place. While manufacturer directives do not require post-exercise screening we feel that

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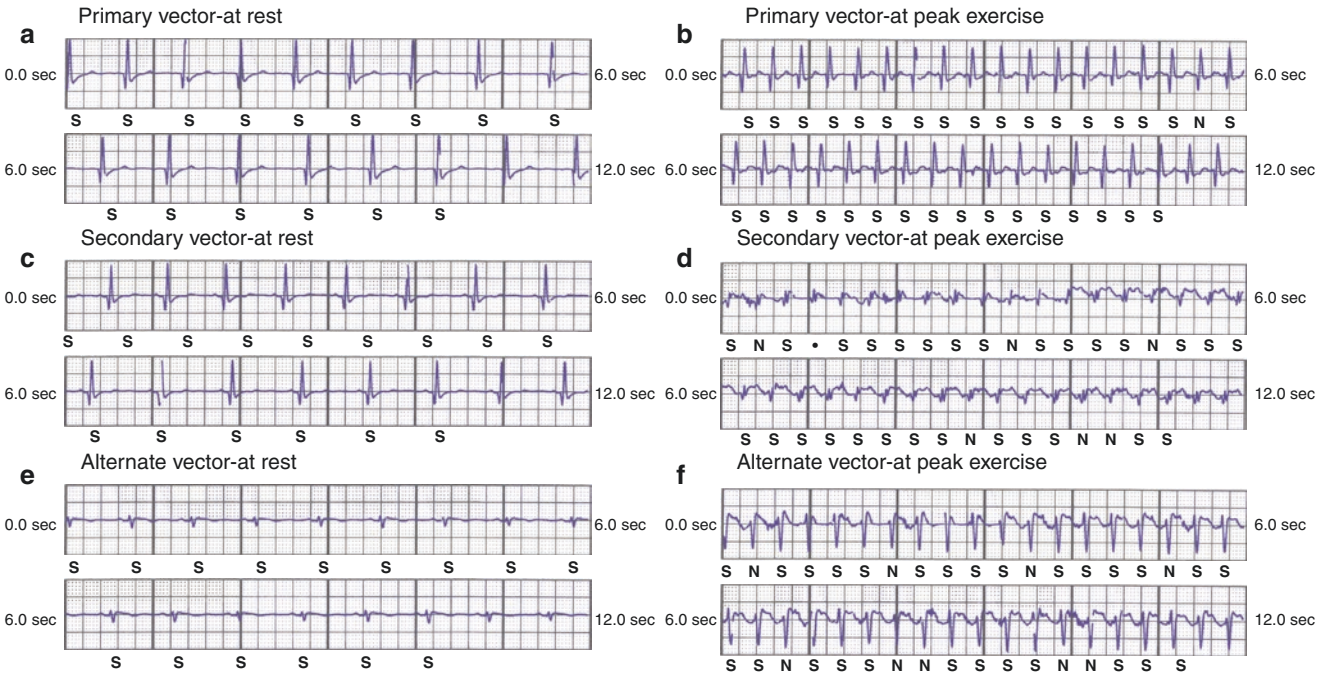

Fig. 32.1 At rest the both primary and secondary vectors had excellent sensing and discrimination between R waves and T waves. The alternate vector had low amplitude R waves. At peak exercise the secondary vector R waves were reduced in amplitude and the T-waves increased, rendering this a suboptimal sensing channel. However, the R waves remained ideal in the primary vector. Interestingly the R waves improved in the alternate channel with exercise. (**a**) Primary vector-at rest. (**b**) Primary vector-at peak exercise. (**c**) Secondary vector-at rest. (**d**) Secondary vector-at peak exercise. (**e**) Alternate vector-at rest. (**f**) Alternate vector-at peak exercise

this is particularly necessary for young HCM patients. Screening is typically deemed to be successful if two ECG vectors do not exhibit T-wave oversensing while lying, standing, and after exercise [3]. The patient passed screening on two of three vectors and underwent successful implantation of a S-ICD in 2017. Step down DFTs (65, 50, and 35 J) all successfully terminated ventricular fibrillation. His ventricular fibrillation and ventricular tachycardia zones were set at 240 and 220 beats/min, respectively. Tachytherapies for each zone were programmed to defibrillation at 80 J × 5 to provide an adequate safety margin, which is particularly important in HCM patients in whom the DFT may increase with time [3].

The importance of assessing for T-wave oversensing during adequate exercise testing, not only with screening, but as well after implantation, particularly in exercising athletes, should be stressed. As discussed, T-wave amplitude and morphology may significantly change with exercise, thereby increasing the risk for inappropriate shocks [3]. Six weeks following implantation, the patient was evaluated during exercise testing, in which his tachytherapies were temporarily disabled and he exercised to 100% of his maximum age-predicted heart rate without issue. Interrogation of his device vectors revealed that his primary vector had a more ideal sensing profile with less T-wave sensing than the secondary vector in which he was initially programmed. Consequently, his primary vector was chosen as his sensing channel. See Fig. 32.1a–f.

In follow-up the patient was doing well and was able to exercise without sustaining any inappropriate shocks. In 2019 at age 17, and 2 years after S-ICD implantation, the patient was running on a treadmill and developed lightheadedness that culminated in syncope. Device interrogation revealed an episode of ventricular fibrillation following sinus tachycardia that was appropriately sensed and successfully treated with defibrillation by his S-ICD (Fig. 32.2). The successful therapy for this unfortunate event highlights the importance of risk stratification and ICD implantation in HCM patients at risk for SCD. This further demonstrates the effectiveness of a S-ICD device achieved after a meticulous screening process and device optimization with exercise testing to ensure appropriate device therapy for patients with HCM while minimizing inappropriate shocks.

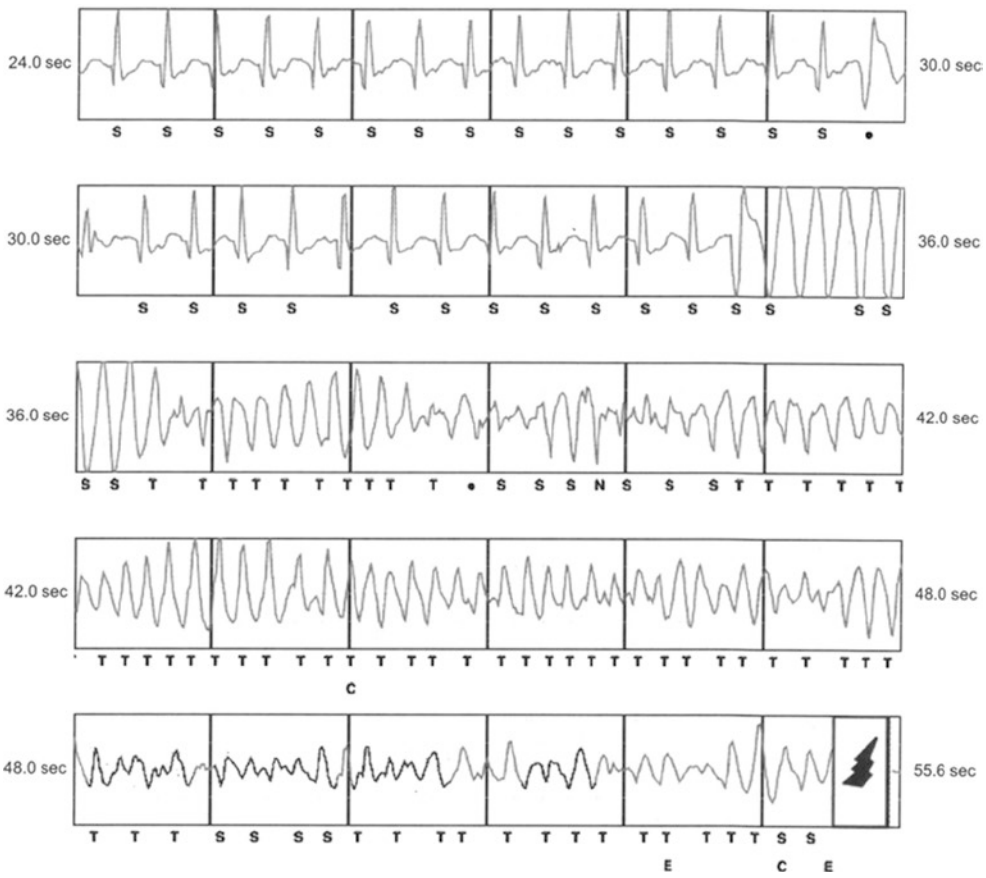

Fig. 32.2 Development of ventricular fibrillation following sinus tachycardia during exercise, successfully terminated with defibrillation administered by the patient's subcutaneous implantable cardioverter defibrillator

References

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