Catecholaminergic Polymorphic Ventricular Tachycardia
Activity as Tolerated?*

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“Children very quickly learned how to avoid the stress that might trigger the light-headedness.”

—From Leenhardt’s seminal report on the first pediatric CPVT cohort (1)

The practice of wise physicians recommending activity restriction in the setting of heart disease is woven into the fabric of cardiology. Until quite recently, “bed rest” was standard care for many common conditions, including myocardial infarction, rheumatic fever, and Kawasaki disease. Three decades ago we learned that patients with heart failure were best off with beta-blocker–induced “bed rest” after inotropic and adrenergic stimulation in heart failure not only failed to improve outcomes but also placed patients in harm’s way. In the realm of adrenergically triggered arrhythmias, immense emphasis has been placed on ultracautious prevention of triggers that lead to events (2), arguably more so than on the development of rigorous evidence to titrate and deliver effective medical and interventional therapies to protect patients and liberate them to be more active.

It has been just more than a decade since a group of experts came together to deliberate on exercise recommendations across a broad range of cardiac conditions, armed with very little data on which to base understandably conservative recommendations (2). Dr. Ackerman coauthored the Bethesda recommendations stating that “Symptomatic patients (with catecholaminergic polymorphic ventricular tachycardia [CPVT]) have a poor prognosis unless treated with an implantable cardioverter-defibrillator (ICD), and all such patients are restricted from competitive sports with the possible exception of minimal contact, Class 1A activities” (2). Since then, Ackerman and his team have been working to temper this edict with data. In this issue of JACC: Clinical Electrophysiology, Ostby et al. (3) report their experience with a cohort of patients diagnosed with CPVT, one-third of whom chose to continue with athletic participation after consultation at the renowned Mayo Clinic’s specialized inherited heart rhythm clinic. We do not have all the details we would like about the population, such as whether the patient is the proband or a symptomatic relative, whether this is a “reassigned” diagnosis, whether the patient is seeking a review of athletic exclusion, or what is meant by “clinical risk profile.” However, we can glean that the population is primarily referral based with a consensus statement–supported diagnosis of CPVT at a mean age of 16 years. At the time of the retrospective analysis, 79% of the group was highly treated with beta-blockers, and 35% were treated with flecainide; 32% had an ICD and 30% had previous sympathetic denervation. The temporal management of medical therapy is not described in response to intake or subsequent clinical status, but we are provided with helpful specifics about drug selection and titration.

SEE PAGE 253

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Ostby et al. (3) focus on the patients’ self-declared athletic status. A diversity of activities is represented, none in the “officially sanctioned” 1A zone of the Bethesda guidelines (billiards, cricket, bowling, golf, curling and riflery). Importantly, the level of participation represents the spectrum from primary school to collegiate and professional athletes. Even allowing for recall bias, patients identified as “never athletes” were 10 years older at the time of diagnosis and had fewer events. This is consistent with the literature on other inherited heart rhythm disorders in which athletes are more likely to present earlier (4). After a comprehensive assessment and discussion, 3 of 24 athletes chose to discontinue their sports, whereas the remainder continued, with 8 competing at a collegiate or adult level. In the context of a 14% CPVT-related breakthrough event rate during follow-up, 3 events occurred in athletes (mean 9-year follow-up) and 7 occurred in 6 nonathletes (mean 5.5-year follow-up). The events were primarily appropriate ICD shocks, 2 with electrical storms, and syncope. There was no difference in event rates between groups, although if anything, a trend suggested fewer events in athletes. Eight of 9 events did not occur during athletic competition. Additional contributors included medication noncompliance, a missed dose, or use of metoprolol, which has recently been shown to be inferior to nadolol in a population who received both medications (5). Simply put, no events would have been prevented by activity restriction, and 4 of 9 events are not readily explicable, especially without the granularity of timing of medication dose, intercurrent illness, and other modulators of autonomic tone.

The definitions of athletes as “never,” “ever,” and “yes” or “no” are somewhat stochastic for the real world. For this study, “competitive” was defined as physical demands greater than perceived of Class 1A; it is ironic that many would consider most of the Class 1A activities as competitive (i.e., officiated with score keeping and producing winners and losers) but not remotely adrenergic. However, we know that athletics is not a uniformly stressful endeavor. Training makes exercise safer with fewer cardiovascular adverse outcomes in those who regularly prepare for physical exertion. The “weekend warrior” is at higher risk because the stress of the exertion is greater for the infrequent participant. Is it possible that the regularly exercising patient with CPVT is at lower risk than the deconditioned burst exercise patient? Could regular intermediate intensity exercise be protective, suggesting a pilot study of exercise in CPVT?

The metabolic equivalents may be important determinants of stress to the system. Gow et al. (6) demonstrated that the metabolic equivalents associated with the usual activities of children and adolescents far exceed what the guidelines condone. Although we have made great progress in understanding the mechanisms of arrhythmias in CPVT, this study again confirms that there is more to the lethality than adrenaline and exertion. Perhaps athlete and nonathlete classifications are less the issue than the largely unmeasured adrenergic state of say a 2-year-old child having a temper tantrum in the middle of a shopping mall or an adolescent playing video games!

One of the most important aspects of this study was relegated to Appendix status: the systematic overview of the discussion with the family. The patient- and family-centered approaches to exercise prescription are bold developments for a historically paternalistic profession (7). In the channelpathy chapter in the 2015 update to the Bethesda guidelines, section chair Ackerman did foreshadow this work (8). The guidelines are permissive in the genotype-positive, phenotype-negative patient. He also built in some welcome flexibility with the statement that “Exceptions to this limitation should be made only after consultation with a CPVT specialist.” However, because of variability in the stress test in CPVT, this concept of unleashing the athlete with a negative stress test who is receiving medical therapy may be challenging to put into practice.

What then can we conclude? These patients are complex and are best cared for in specialized multidisciplinary clinics (9). The nuanced nature of these conditions does exceed the skill set of most health care providers (10) and may be difficult for families to fully comprehend at the initial appointment; an ongoing dialogue and relationship is recommended (9). Shared decision-making supported by evidence-guided medical therapy and incremental interventions lays the foundation for a more permissive approach to not only allowing but also potentially encouraging participation in physical activity, including competitive sports.

An automated external defibrillator is recommended as part of the athlete’s equipment. We direct this responsibility to the entire circle of sport, including the team, trainer, spectator (including family), and teammates. Thus, the emergency plan is important to review, and we encourage schools, teams, and recreational clubs to embrace cardiopulmonary resuscitation training. Despite its relative absence in this series, we also think that swimming retains special status, not only because of the specific association with CPVT but also because of the potential for a worse outcome in water.

Can we relax about sports in CPVT now? This paper is part of a growing body of evidence that sports can
be undertaken safely in the presence of previously limiting conditions, including long QT syndrome and implantable defibrillator recipients (11–14). That said, there are a few provisos to consider. The first is the paucity of data to base decisions on, which is particularly concerning when errors in judgment may lead to sudden death. Although data are mounting, there is immense work to be performed to assemble compelling data to inform safety of activity recommendations. Ostby et al. (3) rightfully acknowledge the greater lethality of CPVT compared with long QT syndrome, and we should anticipate some events, even in well-treated patients.

The principle of a unanimous family plan is laudable in theory. Family dynamics are not always simple, especially after the diagnosis of a chronic condition or the death of an affected relative. The coach and the other parents are seldom as motivated or enlightened as the family and health care team (“You want him to have what on the bench at all times?”), and liability is a major concern for many practitioners. The importance of compliance with the correct medication at the optimized dose is a consistent theme in CPVT. The emphasis of this part of the patient/family/community/health care team contract is key. We often make participation in sports conditional on the patient’s compliance with medication, as verified by the parent who is usually sponsoring, disclosing, and transporting to the events. This may align with the postulate of better compliance in athletes. Finally, the addition of flecainide appears to provide compelling evidence of a short-term incremental benefit with beta-blockers, albeit the size and long-term outcome of the population treated requires expansion (15,16).

In conclusion, the study by Ostby et al. (3) is good news for patients with CPVT and their care providers. Although highly publicized, sudden death in the young, particularly during athletic competition, is rare, even in CPVT. None of the events in this series would be prevented by activity restriction, and our bias should be toward health promotion and athletic participation. Further studies and adoption of thoughtful decision-making guided by the scripts of the leaders in the field will help us all improve the care of patients with CPVT.

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