ABSTRACT. The primary objective of preparticipation screening is the detection of intrinsic structural or electrical cardiovascular disorders that predispose an athlete to sudden cardiac death. Both Brugada syndrome (BrS) and arrhythmogenic right ventricle cardiomyopathy (ARVC) can cause repolarization abnormalities in right precordial leads and predispose to sudden cardiac death due to ventricular arrhythmias. Although there is controversy over whether BrS is distinct from ARVC, it is believed that both are different clinical entities with respect to both the clinical presentation and the genetic predisposition. In clinical practice, there may be cases where the dividing line is not so clear, especially in asymptomatic patients or preparticipation individuals. We present a challenging electrocardiogram in a healthy person referred for preparticipation evaluation.

KEYWORDS. Brugada phenocopy, electrocardiogram, epsilon wave, preparticipation screening.

Introduction

The appropriate screening strategy to prevent sudden cardiac death (SCD) in athletes remains a challenging and a highly debated issue. Both the 1996 American Heart Association (AHA) and the 2005 European Society of Cardiology (ESC) consensus panel recommendations agree that cardiovascular screening for young competitive athletes is justifiable and compelling on ethical, legal, and medical grounds. However, there is disagreement about the best approach. The AHA recommends history and physical examination; this approach is pragmatic and relatively inexpensive but has poor sensitivity because most athletes are asymptomatic and physical examination identifies only a minority of those at risk of sudden cardiac death. The inclusion of the electrocardiogram (ECG) in accordance with the recommendations of the ESC improves sensitivity for detection of serious cardiac disease but is associated with an unacceptably high false-positive rate, in part because of the overlap between the electrical manifestations of athletic training and the cardiomyopathies. For young athletes with normal ECG results, echocardiography contributes minimally to the diagnosis of serious cardiac diseases.\(^1\)

The primary objective of preparticipation screening is the detection of intrinsic structural or electrical cardiovascular disorders that predispose an athlete to SCD. Arrhythmogenic disorders of genetic origin include structural cardiomyopathies and inherited arrhythmic syndromes.\(^2\) Arrhythmogenic right ventricle cardiomyopathy (ARVC) is a myocardial disorder characterized by fibro-fatty replacement of the myocardium and ventricular arrhythmias. In contrast, Brugada syndrome (BrS) has long been considered a functional cardiac disorder: no gross structural abnormalities can be identified in the majority of patients. Although there is controversy over whether BrS is distinct from ARVC, it is believed that both are different clinical entities with respect to both the clinical presentation and the genetic predisposition. However, there is a subpopulation with a clinical and electrocardiographic pattern similar to that of BrS among patients with ARVC. The coexistence of these two relatively rare clinical entities is also reported, but some hypothesized that it is more possible that disease of the right ventricular muscle might accentuate the Brugada electrocardiographic pattern. In clinical practice there may be cases where the
Figure 1: The 12-lead electrocardiogram showing the Brugada-like pattern with epsilon wave in V1–V2 positioned in the fourth intercostal space.
dividing line is not so clear. Moreover, an asymptomatic population with Brugada-type ECG exists in the community, and SCD risk stratification of the individual can be problematic.

Case report

A 23-year-old asymptomatic male patient was admitted to the cardiology clinic for preparticipation cardiovascular evaluation before physical activity and participation in sports. He did not have a history of syncope or a family history of SCD, and the results of the physical examination and routine blood tests were unremarkable. He was taking no medications at the time of the evaluation. An echocardiogram showed that the left ventricle appears to function normally, but there was mild right ventricular dilatation without microbubbles in the right atrium or ventricle on contrast study. His first ECG showed a Brugada-like pattern with an epsilon-like wave in leads V1–V2 (Figure 1). The second ECG of the intercostal space showed a Brugada-like wave resembling coved-type ST-segment elevation (Figure 2); however, a drug challenge test with ajmaline (1 mg/kg intravenous infusion over 5 min) showed no diagnostic changes (Figure 3). We planned cardiac magnetic resonance imaging (MRI), which showed no structural cardiac abnormality. According to his asymptomatic status and negative family history, we decided to follow up this patient without any medication or device implantation. At present, this patient is being followed asymptotically at the outpatient clinic.

Discussion

The primary objective of preparticipation screening is the detection of intrinsic structural or electrical cardiovascular disorders that predispose an athlete to SCD.\textsuperscript{3} Moreover, the risk stratification is still challenging, especially in cases of asymptomatic BrS patients or patients with Brugada phenocopy. Management continues to be challenging, with a lack of drug therapy and high complication rates from implantable cardioverter defibrillators (ICDs).\textsuperscript{4} As subsequent registry data were published, it became apparent that the spectrum of risk is wide, with the majority of patients classified as low risk. The low risk of arrhythmic events that is exceeded by ICD-related adverse effects should inform discussions with patients who do not have a prior history of cardiac arrest.

Epsilon wave is said to be pathognomonic of ARVC, but coexistence of the epsilon and Brugada waves on the same tracing has been reported in patients with ARVC.\textsuperscript{5}
Corrado et al.\textsuperscript{5} presented data in support of the hypothesis that a subpopulation of patients with ARVC, referred to as “concealed forms,” present with the typical clinical and electrocardiographic features of BrS, including the pre-sence of type 1 ST-segment elevation and polymorphic Ventricular Tachycardia (VT). The concealed forms of ARVC might be a cause of so-called “idiopathic” ventricular arrhythmias in subjects with apparently “normal hearts.” These findings show an overlap in clinical manifestation and mechanisms of ventricular arrhythmias between patients with ARVC and BrS.\textsuperscript{5} Specific ECG markers that reflect ventricular conduction delay in ARVC are commonly observed in subjects with spontaneous or drug-induced type 1 ECG pattern of BrS.

\textbf{Figure 3:} Ajmaline provocative testing. Negative ajmaline provocative testing showing no ST-segment elevation in leads V1–V3 positioned in the fourth intercostal space.
Moreover, provocable coved-type ST-segment elevation in the right precordial leads is observed in approximately 16% of patients with typical ARVC. The structural right ventricular abnormalities in ARVC therefore most likely predispose patients to develop Brugada phenocopy and associated ventricular arrhythmias. Moreover, it has recently been speculated that there are some discriminating ECG criteria between incomplete right bundle branch block (RBBB) and Brugada types 2 and 3 ECG patterns. Our patient’s ECG was compatible with RBBB (Figures 4 and 5).

In the current case, there was no known family history of SCD or personal history of arrhythmic symptomatology, and therefore there was a low clinical pretest probability of true BrS. The patient underwent ajmaline provocative testing, with a negative result ruling out the diagnosis of BrS. Clinical diagnosis of ARVC is often difficult because of the non-specific nature of disease features and the broad spectrum of phenotypic manifestations. He had a prominent epsilon wave, but the cardiac MRI was negative. Although negative MRI findings do not rule out ARVC, positive MRI findings should be used as important additional criteria in the clinical diagnosis of ARVC. Furthermore, ARVC is a disease that may have a temporal progression, and the disease may manifest differently according to the time of patient presentation. However, we decided that these electrocardiographic findings were normal in an asymptomatic individual without family history of SCD; we followed him up with periodic outpatient visits with regard to arrhythmic symptoms without restricting competitive sports activity. The inclusion of the ECG in accordance with the recommendations of the ESC improves sensitivity for detection of serious cardiac disease, but is associated with an unacceptably high false-positive rate. Further studies are required to define what constitutes a normal ECG in athletes.

Figure 4: Discriminating electrocardiogram (ECG) criteria between incomplete right bundle branch block and the Brugada ECG patterns.
References


Figure 5: Discriminating electrocardiogram (ECG) criteria between incomplete right bundle branch block and the Brugada ECG patterns.