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Brugada Syndrome

State-of-the-Art Review

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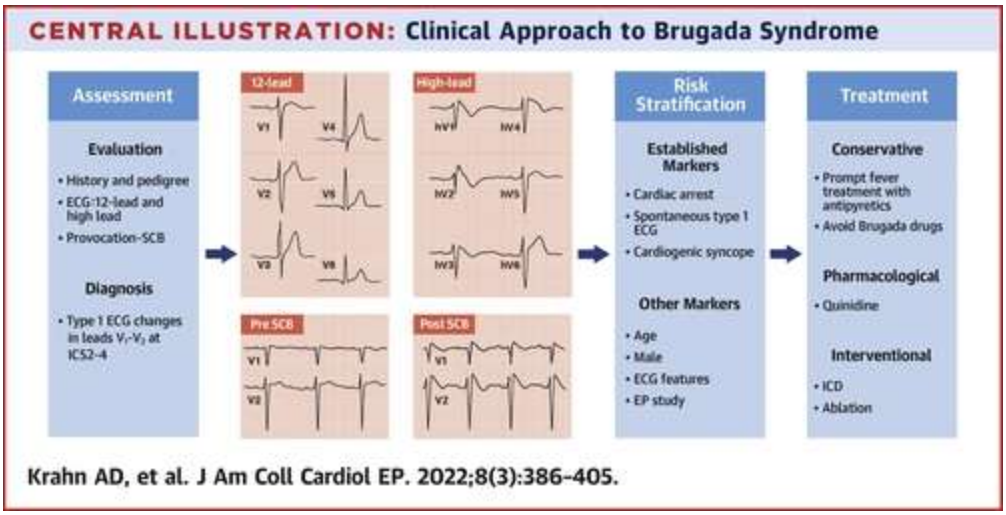
Central Illustration



Abstract

Brugada syndrome (BrS) is an “inherited” condition characterized by predisposition to syncope and cardiac arrest, predominantly during sleep. The prevalence is ∼1:2,000, and is more commonly diagnosed in young to middle-aged males, although patient sex does not appear to impact prognosis. Despite the perception of BrS being an inherited arrhythmia syndrome, most cases are not associated with a single causative gene variant. Electrocardiogram (ECG) findings support variable extent of depolarization and repolarization changes, with coved ST-segment elevation ≥2 mm and a negative T-wave in the right precordial leads. These ECG changes are often intermittent, and may be provoked by fever or sodium channel blocker challenge. Growing evidence from cardiac imaging, epicardial ablation, and pathology studies suggests the presence of an epicardial arrhythmic substrate within the right ventricular outflow tract. Risk stratification aims to identify those who are at increased risk of sudden cardiac death, with well-established factors being the presence of spontaneous ECG changes and a history of cardiac arrest or cardiogenic syncope. Current management involves conservative measures in asymptomatic patients, including fever management and drug avoidance. Symptomatic patients typically

undergo implantable cardioverter defibrillator insertion, with quinidine and epicardial ablation used for patients with recurrent arrhythmia. This review summarizes our current understanding of BrS and provides clinicians with a practical approach to diagnosis and management.

Highlights

- Recently, there have been significant advances in our understanding of BrS.
- This review provides practical recommendations for the diagnosis and treatment of BrS.
- Further research is required regarding the pathophysiological understanding and risk stratification in BrS.