

EP News: Allied Professionals

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Follow-up of patients with Brugada syndrome treated with ICDs

In this single-center registry of patients with Brugada syndrome (BrS), Hernandez-Ojeda et al (J Am Coll Cardiol 2017;70:1991, PMID 29025556) described long-term follow-up including incidence of appropriate and inappropriate shocks and complications associated with implantable cardioverter-defibrillators (ICDs). The study group was composed of 104 patients with BrS who subsequently underwent ICD implantation. All patients had either a spontaneous or a provoked (by intravenous flecainide or ajmaline) type 1 (coved-type) electrocardiographic pattern and had no overt structural heart disease on testing. The authors defined secondary prevention devices as those implanted for aborted sudden death or documented ventricular tachycardia (VT) or ventricular fibrillation (VF). All other devices were considered primary prevention, including those placed in patients with a history of syncope or inducible VT/VF. All but one of the ICDs implanted were transvenous systems (1 subcutaneous ICD), and all were programmed with a cutoff of ≥ 200 beats/min, a long detection time, and preshock antitachycardia pacing when available. Patients were followed routinely in cardiogenetic clinic, in ICD clinics, and with remote monitoring when available. No patient was lost to follow-up.

Indication for ICD implantation include secondary prevention in 10 (9.6%), syncope in 49 (47%), and inducible VT/VF in 35 (34%). During 9.3 ± 5.1 years of follow-up, 21 patients (20.2%) received appropriate ICD therapy for VT or VF, including 2 patients, one of whom was previously asymptomatic, with their first shock >10 years after initial implantation. The average time to first appropriate shock was 48.8 ± 54 months. In univariate and multivariate analyses, ICDs implanted for a secondary prevention indication and patients with a history of syncope had a higher risk of appropriate ICD therapy. Of note, 4 of 45 asymptomatic patients (8.9%) received appropriate therapy. All 4 had inducible VT/VF with programmed stimulation. A total of 4.5% of patients received therapy for monomorphic VT, and the remainder had polymorphic VT or VF. Inappropriate

shocks occurred in 9 patients (8.7%): 2 for atrial fibrillation, 1 for sinus tachycardia, 1 for T-wave oversensing, and 5 for lead noise. The incidence of inappropriate shocks in asymptomatic patients is 6.7%. Device-related complications occurred in a total of 21 patients (20.2%), including infection ($n = 7$ [6.7%]), cardiac perforation requiring pericardiocentesis ($n = 1$ [1%]), and lead dysfunction requiring revision ($n = 12$ [11.5%]). During follow-up, 3 patients (2.9%) died, including 1 who died of VT storm during lead extraction. *The authors conclude that ICDs are effective in high-risk people with BrS but can be associated with device-related problems including inappropriate shocks and other complications, some of which can be mitigated by appropriate device programming and follow-up.*

Brugada syndrome

Brugada syndrome (BrS) is an inherited arrhythmia syndrome characterized by ST-segment elevation in leads V₁-V₂. ST elevation can occur spontaneously or with provocation with certain medications or even during fever. Patients with BrS are predisposed to ventricular tachycardia/ventricular fibrillation (VT/VF), and sudden death can be the first manifestation. There are genes associated with BrS, in particular mutations in *SCN5A*; however, only approximately one-third of patients with BrS have a pathologic genetic abnormality identified. Most agree that an implantable cardioverter-defibrillator (ICD) is recommended in patients with aborted sudden death or documented VT/VF as well as patients with syncope and type 1 pattern. The role of programmed stimulation and drug infusion testing in risk stratification is less clear. Other treatments for BrS include quinidine and ablation, but neither of these supplants ICD at this time. There are important lifestyle modifications that can be made, including avoiding excessive alcohol and cannabis, aggressively treat fever, and avoiding certain drugs that can provoke the ST-segment elevation. A listing of these medications can be found at BrugadaDrugs.org. Important information on the diagnosis and treatment of BrS can be found in the HRS/EHRA/APHRS expert consensus statement on inherited arrhythmia disorders (Priori et al, Heart Rhythm 2013;10:1932, PMID 24011539).

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