the acute infarct with a remodeled ventricle and markedly depressed LV function. Even when there is biomarker evidence of acute MI, a preexisting scar from previous MI should be suspected as the cause of the VT. Infarct scars provide a durable substrate for sustained VT, and up to 70% of patients have a recurrence of the arrhythmia within 2 years. Scar-related reentry is not dependent on recurrent acute myocardial ischemia, so coronary revascularization cannot be anticipated to prevent recurrent VT, even when it may be appropriate for other indications. Depressed ventricular function, which is a risk factor for sudden death, is usually present. Implantation of an ICD is warranted for most patients provided that there is a reasonable expectation of survival with acceptable functional status for the next year after recovery from the VT episode. ICDs reduce annual mortality from 12.3% to 8.8% and lower arrhythmic deaths by 50% in patients with hemodynamically significant sustained VT or a history of cardiac arrest compared with pharmacologic therapy. Chronic amiodarone therapy may be considered for patients who are not candidates for or who decline ICD placement.

Following ICD implantation, patients remain at risk for heart failure, recurrent ischemic events, and recurrent VT, with a 5-year mortality that exceeds 30%. Attention to therapies with survival benefit, including β-adrenergic blocking agents, angiotensin-converting enzyme inhibitors, and statins, is important. Patients with frequent symptomatic recurrences of VT require antiarrhythmic drug therapy or catheter ablation.

NONISCHEMIC DILATED CARDIOMYOPATHY Sustained monomorphic VT associated with nonischemic cardiomyopathy is usually due to scar-related reentry. The etiology of scar is often unclear, but progressive replacement fibrosis is the likely cause. On cardiac MRI, scars are detectable as areas of delayed gadolinium enhancement and are more often intramural or subepicardial in location as compared with patients with prior MI. Scars that cause VT are often located adjacent to a valve annulus and can occur in either ventricle. Any cardiomyopathic process can cause scars and VT, but cardiac sarcoidosis **(Chap. 390)** and Chagas' disease **(Chap. 252)** are particularly associated with monomorphic VT **(Table 277-2)**. An ICD is usually indicated with additional drugs or ablation for control of recurrent VT.

MONOMORPHIC VT IN ARVC ARVC **(Chap. 287)** is a rare genetic disorder most commonly due to mutations in genes encoding for cardiac desmosomal proteins. Approximately 50% have a familial transmission with autosomal dominant inheritance. A less common, autosomal recessive form is associated with cardiocutaneous syndromes that include Naxos disease and Carvajal syndrome. Patients typically present between the second and fifth decade with palpitations, syncope, or cardiac arrest owing to sustained monomorphic VT, although polymorphic VT can also occur. Fibrosis and fibro-fatty replacement most commonly involve the right ventricular myocardium and provide the substrate for reentrant VT that usually has a left bundle branch block–like configuration, consistent with the right ventricular origin. The sinus rhythm ECG suggests the disease in more than 85% of patients, most often showing T-wave inversions in V_1–V_3 (Fig. 277-6). Delayed activation of the right ventricle may cause a widened QRS (≥110 ms) in the right precordial leads and a prolonged S-wave upstroke in those leads, and occasionally a deflection at the end of the QRS known as an *epsilon wave* (Fig. 277-6). Cardiac imaging may show right ventricular enlargement or areas of abnormal motion or reveal areas of scar on CMR imaging with gadolinium. The monomorphic VT of early ARVC can sometimes be difficult to differentiate from idiopathic right ventricular outflow tract VT.

LV involvement can occur and occasionally precede manifest right ventricular disease. Heart failure is rare except in late stages, and survival to advanced age can be anticipated provided that VT can be controlled. An ICD is recommended. When VT is exercise-induced, it may respond to β-adrenergic blockers and limiting exercise. Sotalol, amiodarone, and catheter ablation have been used to reduce recurrences. Ablation targets are often located in the subepicardium of the RV.

TETRALOGY OF FALLOT VT occurs in 3–14% of patients late after repair of tetralogy of Fallot **(Chap. 282)** and contributes to a 2% per decade risk of sudden death. Monomorphic VT is due to reentry around areas of surgically created scar in the RV (Table 277-2). Factors associated with VT risk include age >5 years at the time of repair, high-grade ventricular ectopy, inducible VT on an electrophysiologic study, abnormal right ventricular hemodynamics, and sinus rhythm QRS duration >180 ms. An ICD is usually warranted for patients who have a spontaneous episode of VT, but criteria for a prophylactic ICD in other patients have not been established. Catheter ablation is used to control recurrent episodes.

TABLE 277-2 VENTRICULAR ARRHYTHMIAS ASSOCIATED WITH DIFFERENT FORMS OF HEART DISEASE

- I. Idiopathic VT without structural heart disease
  - A. Outflow tract origin
    1. RV outflow tract: left bundle branch block pattern with inferior axis (tall QRS in inferior leads) and late transition in the precordial leads
    2. LV outflow tract: prominent R in V_1 with inferior axis
  - B. Left posterior fascicular VT
    1. Right bundle branch block pattern with left axis deviation (most common)
- II. Ischemic cardiomyopathy
  - A. Monomorphic VT is common with prior large myocardial infarction
  - B. Polymorphic VT and VF should prompt ischemia evaluation
- III. Nonischemic cardiomyopathy
  - A. Polymorphic VT and VF more common but fibrotic scars can cause monomorphic VT especially with sarcoidosis and Chagas' disease
- IV. Arrhythmogenic right ventricular cardiomyopathy
  - A. Monomorphic VT usually of right ventricular origin (left bundle branch morphology)
  - B. Polymorphic VT and VF can occur independently or through degeneration of monomorphic VT
- V. Repaired tetralogy of Fallot
  - A. Monomorphic VT of right ventricular origin (usually left bundle branch morphology)
- VI. Hypertrophic cardiomyopathy
  - A. Polymorphic VT or ventricular fibrillation
  - B. Less commonly, monomorphic VT associated with myocardial scars
- VIII. Genetic arrhythmia syndromes
  - A. Long QT syndrome: torsade de pointes VT
  - B. Brugada syndrome: VF
  - C. Catecholaminergic polymorphic VT: polymorphic VT or bidirectional VT
  - D. Short QT syndrome: ventricular fibrillation
  - E. Early repolarization syndrome: polymorphic VT or VF

Abbreviation: RV, right ventricle. See text for other abbreviations.

BUNDLE BRANCH REENTRY VT Reentry through the Purkinje system occurs in approximately 5% of patients with monomorphic VT in the presence of structural heart disease. The reentry circuit typically revolves retrograde via the left bundle and anterograde down the right bundle, thereby producing VT that has a left bundle branch block configuration. Catheter ablation of the right bundle branch abolishes this VT. Bundle branch reentry is usually associated with severe underlying heart disease. Other scar-related VTs are often present and often require additional therapy or ICD implantation.

IDIOPATHIC MONOMORPHIC VT Idiopathic VT in patients without structural heart disease usually presents with palpitations, lightheadedness, and occasionally syncope, often provoked by sympathetic stimulation during exercise or emotional upset. The QRS morphology of the arrhythmia suggests the diagnosis (see below). The sinus rhythm ECG is normal. Cardiac imaging shows normal ventricular function and no evidence of ventricular scar. Occasionally a patient with structural heart disease is found to have concomitant idiopathic VT, unrelated to the structural disease. Sudden death is rare.

Outflow tract VTs originate from a focus, usually with features consistent with triggered automaticity. The arrhythmia may present with