cardiomyopathy, suggesting that primary prevention with ICDs for patients with prior MI or nonischemic cardiomyopathy and heart failure was appropriate.

The risk of SCD after MI is highest in the few months after the index event. However, ICDs have not been effective when implanted immediately after MI or revascularization procedures. The reason for this is unclear; it may reflect the large percentage of patients who have improved cardiac function early on, which decreases the risk of SCD and therefore the benefit of an ICD. Alternatively, the mechanism for SCD in the early period after an MI or revascularization procedure may be recurrent ischemia rather than reentrant tachycardia and therefore less amenable to ICD therapy. The Defibrillator in Acute Myocardial Infarction Trial (DINAMIT) randomized 675 patients with low ejection fractions immediately after MI to ICD or medical therapy; no difference in mortality rates was seen. The current recommendations are to avoid primary prevention with ICDs within 40 days of an MI or 3 months of revascularization.

A significant challenge in modern medicine is identifying patients who are have an elevated risk of SCD to allow effective use of primary prevention interventions such as ICDs. Some known predictors of SCD after MI are shown in Table 9-7, but many are not specific or sensitive enough for practical use. Reduced ejection fraction has been the most successful noninvasive measure that can predict increased risk of SCD. An electrophysiologic study is a minimally invasive catheter procedure that with electrical stimulation can help to identify patients who are prone to VT. Electrophysiologic studies are most sensitive in patients with prior MI, but they may be less useful in other cardiac conditions. Cardiac magnetic resonance imaging (MRI), which can directly image cardiac function and cardiac scar or fibrosis, is showing great promise as a more sensitive and specific, noninvasive risk predictor of SCD.

Ventricular Tachycardia and Ventricular Fibrillation without Evident Heart Disease

Ventricular arrhythmias occurring in the absence of structural heart disease usually carry a benign prognosis but can be associated with SCD in patients with genetic arrhythmic syndromes predisposing to life-threatening polymorphic VT. Genetic screening for these syndromes is important to identify at-risk family members.

Idiopathic Ventricular Tachycardia

Idiopathic VT most commonly originates from the outflow tracts, with approximately 80% localized to the RVOT and the remainder originating in the left ventricular outflow tract (LVOT), the aortic sinuses of Valsalva, and the region of the aortomitral continuity. Idiopathic RVOT VT manifests with the characteristic electrocardiographic findings of left bundle branch block and inferior axis VT QRS morphology. Triggered activity is the mechanism underlying outflow tract tachycardias. This calcium-dependent mechanism explains why an outflow tract VT often terminates with adenosine, β-blockers, and calcium-channel blockers.

Patients in their third or fourth decade typically have palpitations, shortness of breath, and lightheadedness at presentation. Reports of cardiac arrest are rare, and treatment is directed at controlling symptoms. β-Blockers and calcium-channel blockers are often used initially, although some patients require catheter ablation or antiarrhythmic drug therapy. A subset of asymptomatic patients may develop tachycardia-mediated cardiomyopathy due to frequent ventricular ectopy. The PVC burden posing the greatest risk for producing left ventricular dysfunction is likely more than 10,000 PVCs daily. Fortunately, PVC suppression with catheter ablation usually improves ventricular function.

TABLE 9-7 PREDICTORS OF SUDDEN CARDIAC DEATH AFTER MYOCARDIAL INFARCTION

Decreased left ventricular ejection fraction
Residual ischemia
Delayed enhancement on cardiac MRI
Late potentials on signal-averaged electrocardiography
Decreased heart rate variability
Prolonged QT on ECG
Induction of sustained MMVT with programmed electrical stimulation
Complex ventricular ectopy (e.g., NSVT) on ambulatory monitoring

ECG, Electrocardiogram; MMVT, monomorphic ventricular tachycardia; MRI, magnetic resonance imaging; NSVT, nonsustained ventricular tachycardia.

Arrhythmogenic Right Ventricular Cardiomyopathy or Dysplasia

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited cardiomyopathy with typically autosomal dominant transmission. It is associated with mutations affecting desmosomes, which are molecular complexes of cell adhesion proteins that bind cardiac myocytes. Although morphologic changes in the RV free wall predominate, biventricular or primary left ventricular variants occur. Due to myocyte death, large portions of the right ventricle are replaced with adipose tissue, leading to wall motion abnormalities, cardiac dysfunction, and aneurysm formation. Structural changes spread from the epicardium to the endocardium. RV imaging classically demonstrates RV enlargement with focal wall motion abnormalities and RV hypokinesis. The RV free wall is not well imaged by routine cardiac echocardiography, and MRI has become the gold standard for the diagnosis of ARVC.

ARVC patients develop ventricular arrhythmias with associated symptoms, including palpitations, lightheadedness, syncope, and SCD. Given the typical RV origin of arrhythmias in ARVC, the ventricular arrhythmias have a left bundle branch morphology. The surface ECG during sinus rhythm may demonstrate inverted T waves in the V_1 to V_3 leads or epsilon waves, which are low-amplitude deflections at the end of the QRS complex in the right precordial leads resulting from slowed RV conduction.

Distinguishing ARVC from idiopathic RVOT VT is essential because of the different prognostic and therapeutic implications of the two diagnoses. The diagnosis of ARVC is established by the ARVC Task Force Criteria. Risk factors for SCD of ARVC patients include prior aborted episodes of SCD, syncope, young age, LV dysfunction, and markedly diminished RV function.

Patients with documented ARVC typically receive ICDs. Adjunctive therapy with antiarrhythmic drugs or ablation, particularly strategies incorporating combined epicardial and endocardial ablation, may be useful in treating symptomatic VT.