Treatment

Treatment of RCM focuses on alleviating the symptoms of heart failure. Diuretics are used for decongestion, but intravascular depletion may compromise ventricular filling and lead to reduced cardiac output and hypotension. Supraventricular tachyarrhythmias are poorly tolerated. In patients with conduction system disease such as advanced atrioventricular block, a permanent pacemaker may be indicated. Specific therapies for underlying disorders include chemotherapy in amyloidosis, phlebotomy and iron chelation therapy in hemochromatosis, and steroids in sarcoidosis and endomyocardial fibrosis.

Prognosis

The course of RCM depends on the pathology, and treatment is often unsatisfactory. In the adult population, the prognosis usually is poor, with progressive deterioration and death due to low-output heart failure.

Arrhythmogenic Right Ventricular Cardiomyopathy

Definition and Epidemiology

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an autosomal dominant disease characterized by specific myocardial pathology. The estimated prevalence of ARVC is about 1 case in 2000 to 5000 people, and it has a male predominance.

Pathology

The myocardium of the right ventricular free wall is progressively replaced by fibrous and adipose tissue. Right ventricular function is abnormal, with regional akinesis or dyskinesis or global right ventricular dilation and dysfunction.

Clinical Presentation

The disease typically manifests in young adults as palpitations, dizziness or syncope, or sudden cardiac death. Symptoms of right ventricular failure are rare, despite evidence of right ventricular dysfunction on imaging studies.

Diagnosis

The clinical diagnosis of ARVC is suggested by integration of the information from the clinical presentation (e.g., arrhythmias), electrocardiogram, family history, and imaging studies. When available, histologic examination of the right ventricle confirms the diagnosis. The resting electrocardiogram may be normal, but common abnormalities include incomplete or complete right bundle branch block, the so-called epsilon waves that follow the QRS complex, and inverted T waves in the precordial leads. Right ventricular dilation and systolic dysfunction can be seen with echocardiography and MRI. The latter modality can also show myocardial fat.

Treatment

Treatment consists of ICD therapy to prevent sudden cardiac death, but the indications for implantation are not well defined. Antiarrhythmics and radiofrequency ablation of ventricular tachycardia are used in patients with frequent arrhythmias, but they have not been shown to reduce the risk of sudden cardiac death. Patients with a probable or definite diagnosis of ARVC should be excluded from competitive sports.

Prognosis

The prognosis for these patients remains uncertain.

Unclassified Cardiomyopathies

Some cardiomyopathies that do not fit the current categories are described in Table 10-2.

For a deeper discussion of this topic, please see Chapter 60, "Diseases of the Myocardium and Endocardium," in Goldman-Cecil Medicine, *25th Edition.*

SUGGESTED READINGS

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