

Editorial

Cardiomyopathy: The Importance of Recognizing the Uncommon Diagnosis

The distinction between a sound cardiologist and an excellent cardiologist is the ability of the latter to recognize and diagnose the unusual. In this issue, a group of experts reviews a spectrum of unusual cardiomyopathies, any or all of which may present to the general cardiologist. Myocarditis, reviewed by Blauwet and Cooper, runs the gamut of severity from mild to life-threatening and may mimic a whole spectrum of cardiovascular disease. Although the classic presentation with acute onset heart failure, electrocardiographic changes, and arrhythmia suggests an immediately apparent diagnosis, the authors clearly describe the more subtle ways in which myocarditis may present, as well as the difficulties in precisely defining a diagnosis.

Chagas cardiomyopathy is a disorder that may not be on the diagnostic horizon of cardiologists in the United States and Europe. However, ignoring its existence is a mistake, particularly in an age when travel is so prevalent and migration so common. As Clausell and her coauthors point out in their detailed review, it is estimated that there are more than 100 000 persons in the United States infected with the parasite. Clearly, this is a cardiomyopathy to be reckoned with, wherever one practices.

Cardiomyopathy induced either by cocaine or alcohol has long been recognized, but it is very likely that it is underappreciated. Drug and alcohol abuse is not limited to underprivileged populations, but the more sophisticated abuser may be skilled at hiding his or her addiction. Substance abuse is an important cause of cardiomyopathy in all social strata, and Awtry and Philippides' contribution to this issue discusses the features and subtleties of this potentially reversible cardiac disease.

A particularly difficult diagnosis is that of a disease that mimics a commoner condition. Such is the case with Anderson-Fabry disease, which may be confused with hypertrophic cardiomyopathy. The advent of enzyme replacement therapy with recombinant α -galactosidase has the potential to alter the prognosis of this uncommon condition, which may be present in up to 3% of patients seen in centers specializing in hypertrophic cardiomyop-

athy. O'Mahoney and Elliott draw upon their extensive experience to describe the features, diagnosis, and management of this intriguing condition.

Cardiac amyloidosis is a rapidly progressive disease with several forms. Recent advances in diagnosis and therapy have changed it from a universally fatal condition to one in which long-term survival is possible for many patients. Early diagnosis is critical to obtain the best outcomes. Falk and Dubrey describe the various manifestations of this disease and the importance of precisely determining the type of amyloid in order to appropriately target therapy. In another article in this issue, they address cardiac involvement in sarcoidosis. This is a disease that poses particular challenges in terms of a high risk of sudden death and one in which no randomized trials have addressed the benefit of therapy despite the empiric use of corticosteroids.

Left ventricular noncompaction is a disease in search of a clear definition. An increased awareness of this condition should lead to more frequent diagnosis. More information in a prospective fashion will be required to fully understand the prognostic and therapeutic implications of this condition. Sarma et al provide the most detailed review to date of left ventricular noncompaction and describe the diagnostic challenges as well as therapeutic implications.

Peripartum cardiomyopathy is a form of idiopathic left ventricular dysfunction that occurs during pregnancy or a few months postpartum. Differences in the phenotypic presentation of this disease in different parts of the world add to the complexity of this condition. The article by Tibazarwa and Sliwa provides a detailed review of peripartum cardiomyopathy in Africa where the disease is 3 to 4 times more prevalent than in the United States. Because of the difficulties to diagnose heart failure during pregnancy or in the early postpartum period, diagnosis is often missed or delayed. There is therefore a need for increased awareness and knowledge of this condition among physicians to allow timely diagnosis, early treatment, and prevention of potential complications.

Tibazarwa and Sliwa provide detailed information regarding the challenges of diagnosis and prognosis and highlight some of the exciting therapeutic possibilities recently developed.

We hope that this selection of articles will engender the interest that each diagnosis deserves, will serve to heighten

the awareness of these uncommon but important diseases, and will lead to earlier and more frequent diagnosis to the satisfaction of the patient and cardiologist alike.

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