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An ARVCs Overview: Right ventricular cardiomyopathy

Arrhythmogenic Right Ventricular Dysplasia (ARVD) most frequent form is due to PKP2 desmosomal mutation with increased RV size with thinness of the free wall and segmental anomalies of contraction. This is in contrast with increased trabeculations at the RV apex. It is the result of the presence of apoptosis, fat and interstitial fibrosis mostly observed in the RV free wall and LV apex. This disease is frequent in the general population but become clinically apparent in a small number of cases. Disease starts in the embryo in the mediomural layers and has been detected in one case by Echo sonography. It is also the result of a unique PLN mutation. Clinical presentation is mostly ventricular arrhythmias which can lead to unexpected sudden cardiac death especially in young people and during endurance sports. Biventricular form is less frequent but caries a poorer prognosis. Some of these patients seen at a late stage of the disease can be misclassified as IDCM. Pathology of Heart Tx will re-establish the diagnosis. Fatty non-compaction (FNC) is common in the general population (60%) due to the presence of an excessive amount of fat in the RV free wall up to typical ARVD but without fibrosis. It is a determinant of prognosis in case of LV failure. Mutation is unknown. Brugada syndrome (BrS) has a unique ECG pattern of coved type observed only in lead V1. Structural changes are sometimes suggesting ARVD. However, BrS and ARVD are two different entities with some degree overlap both phenotypically and genotypically in a small number of cases. The vast majority in each entity has its own unique features. RVOT VT is generally benign but one personal case of SD with pathologic documentation demonstrated a localized infundibular anomaly suggesting ARVD with fragmentation of potentials by needle infundibular transmural electrode in another case. Uhl's anomaly shows a huge RV with apposition of epicardium against endocardium. However, this can be a differential diagnosis with an extreme form of ARVD. Naxos disease originally detected by Echocardiography is the most impressive form of these RVCs because of its genotype in which both parents are affected by the same mutation of a desmosomal protein called Plakoglobin. However, the heterozygous form shows only minor ECG changes but no clinically significant disease. All of these cardiomyopathies can be affected by a wide clinical spectrum of superimposed myocarditis (which has also in some cases) a genetic background, frequently the determinant of prognosis. However, in some rare patients, the disease can stop completely spontaneously its progression.

Biography

Guy Hugues Fontaine has made 15 original contributions at the inception of cardiac pacemakers in the mid-60s. He has published more than 900 scientific papers including 201 book chapters. He is included in the *Profiles in Cardiology* (W Hurst 2003) book of the 216 individuals who have made a significant contribution to the study of cardiovascular diseases since 14th century. He has been included in the book "*500 greatest Geniuses of the 21st century*" of the American Biographical Institute (ABI 2005). He was the Reviewer of 17 journals both in clinical and basic science. He has served for 5 years as a Member of the Editorial Board of *Circulation*. He has been invited to give 11 master lectures of 90 minutes each during three weeks in the top universities of China (2014).

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