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Guy Hugues Fontaine

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Biography:

Guy Hugues Fontaine has made many original contributions in the field of Cardiac Arrhythmias and has received the Prestigious Award of "Pioneer in Pacing and Electrophysiology" from the Heart Rhythm Society in 2005. He has been also classified by three famous American Cardiologists as one of the 216 individuals who have made a significant contribution to the study of cardiovascular disease since the 14th century. This was because he was the first to identify Arrhythmogenic Ventricular Dysplasia in 1977 currently included in the Right Ventricular Cardiomyopathies.

Advances in the understanding of right ventricular cardiomyopathies

An increasing number of genetic mutations can explain the mechanism of inherited cardiomyopathies Arrhythmogenic Right Ventricular Dysplasia (ARVD) is mostly due to PKP2 desmosomal mutation with increased RV size with apoptotic thinness of the free wall and segmental anomalies of contraction. This is also due to the presence of 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. Clinical presentation is mostly ventricular arrhythmias which can lead to unexpected sudden cardiac death especially in young people and during endurance sports. Some of these patients seen at a late stage of the disease can be misclassified as IDCM. However, in some rare patients, the disease can stop completely its progression. 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. Right Ventricular Outflow Tract Ventricular Tachycardia (ROVT VT) is generally benign but one personal case of SD with pathologic documentation demonstrated a localised infundibular anomaly suggesting localised ARVD. Hypertrophic Cardiomyopathy (HCM) is produced by a genetic mutation in the contractile molecules of the heart producing hypertrophy of myocardial fibres with disarray. It is also a major cause of SD during sports Idiopathic Dilated Cardiomyopathy (IDCM) is mostly due to multiple genetic mutations lamin and myosin affecting myocardial force of contraction. All of these cardiomyopathies can be affected by superimposed myocarditis which is frequently the determinant of prognosis.

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