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Advances in the understanding of inherited cardiomyopathies

An increasing number of genetic mutations can explain the mechanism of inherited cardiomyopathies which can lead to arrhythmias and risk of sudden death as well as irreversible heart failure in the end stage of the disease. Arrhythmogenic Right Ventricular Dysplasia (ARVD) has been identified by the presenter in 1977 as side work at the beginning of anti-arrhythmic surgery. Genetic background has been discovered mostly due to PKP_2 desmosomal mutation with increased RV size, presence of large amount of fatty tissue mostly located on the right ventricle with apoptotic thinness of the free wall and segmental anomalies of contraction. Based on systematic analysis of histology of right ventricle in patients who died of a non-cardiac cause it was found that this disease is frequent in the general population (4%) 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 in whom heart transplantation is the only effective treatment. However, in some rare patients, the disease can stop completely its progression. An important marker of the disease is the presence of Epsilon wave on the ECG. Naxos disease, Uhl's anomalies are rare but important forms. They have initiated the discovery of the first mutation and help in the understanding of arrhythmogenicity as well as advanced forms of treatment including drugs, ablation and implantation of implanted cardiac defibrillator. Brugada syndrome (BrS) has a unique ECG pattern of coved type of the T wave of the ECG 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. Both of them can be controlled by antiarrhythmic drugs, ablation of ventricular tachycardia and implanted cardiac defibrillator. Right Ventricular Outflow Tract Ventricular Tachycardia (ROVT VT) is generally benign but one personal case of SD with pathologic documentation demonstrated a localized infundibular anomaly suggesting localized ARVD. Hypertrophic Cardiomyopathy (HCM) is produced by a genetic mutation in the contractile molecules of the heart producing hypertrophy of myocardial fibers with disarray. It is also a major cause of SD during sports recognized as the most frequent. 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.

Biography

Guy Hugues Fontaine has made 16 original contributions in the design and the use of the first cardiac pacemakers in the early 60s. He has serendipitously identified arrhythmogenic right ventricular dysplasia (ARVD) during his contributions to antiarrhythmic surgery in the early 70s. He has developed the technique of fulguration to replace surgery in the early 80s. He has been one of the 216 individuals who have made a significant contribution to the study of cardiovascular disease since the 14th century and one of the 500 greatest geniuses of the 21st century. He has 900+ publications including 201 book chapters. He is a Reviewer of 21 scientific journals both in basic and clinical science. He has developed new techniques of hypothermia for neurologic brain protection in out-of-hospital cardiac arrest (OHCA), stroke and spinal cord injury. He has recently invented a high-tech device which can be considered as the ultimate in palliative care.

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