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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 a side work at the beginning of anti-arrhythmic surgery. Genetic background has been discovered mostly due to PKP2 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 anomaly 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 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 recognised 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 and may have a genetic background which can be the same as the trouble in development.

Biography

Guy Hugues Fontaine has made 15 original contributions at the inception of cardiac pacemakers in the mid-60s. He has identified ARVD by serendipity in the late 70s, published 900 scientific papers including 201 book chapters. He is got placed in 3 books: 216 Profiles in Cardiology since the 14th century (Hurst 2003), 500 greatest Geniuses of the 21st century (ABI) 2005 USA, the 100 Life time of Achievement (IBC) 2005 Cambridge UK. He is Reviewer of 17 journals both in clinical and basic science. He has given 11 master lectures in China (2014). He is also working on brain and heart protection in cardiac arrest and stroke by therapeutic hypothermia.

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