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Advances in the understanding of ARVCs

ARVCs is covering a spectrum of mostly inherited cardiomyopathies of increasing interest because of a relatively small number of mutations have been identified in 60% of patients. The mechanisms of ECG anomalies are better understood as well as their long term prognosis leading to CHF. 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 3.7% 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 producing a Phenotype suggesting ARVD. However, these diseases 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. Naxos disease identified in the Greek eponym disease is the homozygous form with associated palmoplantar keratosis. This led to the identification of the first mutation Plakoglobin leading to the discovery of multiple candidate genes and finally other mutations. UHL's anomaly is rare form which suggests major early apoptosis creating an arrhythmogenic substrate which proved important to demonstrate the re-entrant mechanism of ventricular arrhythmias. These cardiomyopathies can be affected by a genetically superimposed myocarditis which is frequently the determinant of prognosis. Experimental ablation of the disease has been demonstrated on the Zebra fish and the mouse opening new vistas for its treatment and prevention.

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 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", one of the "500 Greatest Geniuses of the 21st century" (USA Books), one of the "100 Life Time Achievement" awardees (UK Book). He has 900 publications including 201 book chapters. Reviewer of 23 scientific journals both in basic and clinical science. He has served as a member of the Editorial Board of Circulation during 5 years after reviewing during decades papers for this Journal. He has given 11 master lectures of 90' each in inland China in 2014. He has developed new techniques of hypothermia for neurologic brain protection in OHCA, stroke and spinal cord injury. He is the first to have resuscitated his wife at home with an external defibrillator (Schiller) still working after 30 years. He has also invented a high-tech device which can be considered as the ultimate in palliative care.

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