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First histologic demonstration of atrial dysplasia in two ARVD patients

Since the reproduction of the disease-in-the-dish by the group of San Diego, the original term of ARVD is now back in Force. ARVD has been discovered by its disorder in ventricular abnormal activation leading to premature reactivation of myocardium called an extrasystole or Premature Ventricular Contraction (PVC). In a most severe form several consecutive PVCs are observed, are poorly tolerated (especially by women) when they are frequent. In the worst situation this abnormal fast cardiac activity may result to desynchronization of activation of myocardium leading to Ventricular Fibrillation (VF) and in sudden unexpected death. The disease is the result of a genetic anomaly mostly affecting the RV in which the myocardium is occupied by fat and fibrosis. In addition, presence of lymphocytes is the marker of poor prognosis. In anecdotal cases it was observed that patients later confirmed as typical ARVD had atrial arrhythmias as the first presentation of the disease suggesting that the disease can start by the atrium before the ventricle and that atrium was also involved. This concept has been recently confirmed. However, I am the first to study the histology of the atrium in two patients with known ARVD who died suddenly of a non-cardiac cause and in whom I performed myself the extraction of the heart immediately after death and in whom samples were taken from both ventricles and atria giving perfect gross pathology before immersion in formalin. Special histologic staining (HPS) was performed to clearly identify fibrosis. In the first patient only severe interstitial fibrosis was observed all over right and left ventricle. In the second patient a less severe interstitial fibrosis was observed but was associated to replacement fibrosis with some lymphocytes suggesting superimposed myocarditis. The systematic study of the right ventricle of 82 individuals who died of non-cardiac cause in a general hospital showed that 3.7% had the histologic pattern of RVD and not ARVD since those individuals had non arrhythmias. Therefore, these cases represent the quiescent form of ARVD. It is therefore possible to consider that the same situation exists in the general population as far as atrial dysplasia is concerned. This situation may lead to atrial fibrillation spontaneously because of the anatomic creation of an anatomic substrate or it could be a more stable form which becomes arrhythmogenic in case of superimposed myocarditis.

Biography

Guy Hugues Fontaine has written over 990 scientific publications, including over 200 book chapters (number in "text books" Americans). He is one of the 126 greatest cardiologists since the 14th century and one of the 500 Greatest Geniuses of the 21st century. He was the only invitee to a three-week trip in inland China and has attended 11 conferences. He has received Prof. Pierre Rijnant Prize (Brussels) which is equivalent to Nobel Prize in Cardiac Electrophysiology. He is the only Clinician who received it.

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