**ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY: DISEASE OF THE DESMOSOME WITH MULTIPLE CLINICAL PRESENTATIONS**

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Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a polygenic myocardial disease usually with an autosomal-dominant inheritance pattern. Patients with ARVC usually present with ventricular arrhythmias arising from the right ventricle (RV), and in later stages, progressive heart failure may occur due to involvement of the left ventricle (LV). However, sudden cardiac death (SCD) may be the initial presentation or may occur at any time during the course of the disease. Genetic studies have identified causative mutations in genes encoding proteins of the intercalated disk, mainly desmosomal proteins that lead to a reduced electrical and mechanical stability of the myocardium. The diagnosis is established on a point score basis according to the modified Task Force criteria utilizing imaging techniques demonstrating functional and structural changes of the right ventricle, fibrous replacement of myocardium through biopsy, electrocardiogram depolarisation and repolarisation abnormalities, ventricular arrhythmias and a positive family history including identification of causative genetic mutations. Although several risk factors for SCD such as previous cardiac arrest, syncope, documented ventricular tachycardia or ventricular fibrillation, severe RV dysfunction, LV involvement and young age at manifestation have been identified, risk stratification still needs improvement, especially in asymptomatic patients. Therapeutic interventions include restriction from strenuous physical exercise, antiarrhythmic drugs such as beta-blockers, amiodarone and sotalol and implantable cardioverter defibrillator therapy. Long-term follow-up is warranted in asymptomatic carriers of pathologic mutations due to differences in age-related penetrance of the disease.