International Journal of General Medicine
and Pharmacy (IJGMP)
ISSN(P): 2319-3999; ISSN(E): 2319-4006
Vol. 3, Issue 4, July 2014, 31-40

International Academy of Science,
Engineering and Technology
Connecting Researchers; Nurturing Innovations

UPDATE ON ARRHYTHMOGENIC VENTRICULAR DYSPLASIA.

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ABSTRACT

The Arrhythmogenic Ventricular Dysplasia (AVD) is a rare hereditary primary cardiomyopathy causing many diagnostic problems by the limits of current knowledges and instrumental opportunities.

The authors present a review about the most recent advances in the field of pathogenesis and diagnosis.

The pathogenesis relates to mutations in desmosomal genes, proteins involved in intercellular adhesion and in intracellular signal transduction, but also in no-desmosomal genes mutations have been described.

ECG, echocardiography, angiocardiography and RM have diagnostic limitations, such as endomyocardial biopsy, due to defects in sensitivity or specificity.

The AVD is a complex disease and there are still several and significative limitations both in the knowledge of pathogenesis than in diagnosis.

KEYWORDS: Arrhythmogenic Ventricular Dysplasia, Diagnosis, Pathogenesis