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Proceeding

Advances in Cardiology and Cardiovascular Disorders

Genetic basis of arrhythmogenic right ventricular dysplasia and brugada syndrome

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Received: November 10, 2017 | **Published:** November 14, 2017

Sudden cardiac death is increasingly being recognized as a major contributor to mortality in young individuals. However, a significant number of them are unexplained. These are believed to be arrhythmogenic in origin. Sudden death may be the first manifestation of an inherited cardiac disorder. This is difficult to diagnose and genetic testing helps to improve its recognition. There has been considerable progress in the understanding about genetics of sudden cardiac death.

In arrhythmogenic right ventricular dysplasia (ARVD), five causative genes have been identified. However only 30% to 50% of the patients with ARVD are affected with one of these gene abnormalities. It is probable that other genes have not been identified. Frequently, patients have more than 1 genetic defect in the same gene or in a second complementary gene. A family member may have the gene defect with development of disease or have no manifestations. Brugada syndrome is a hereditary disease responsible for ventricular fibrillation and sudden cardiac death in the young. This disease exhibits an autosomal dominant pattern of transmission with variable penetrance. The mean age of onset of events is around 40 years. Sudden death can affect individuals of any age; particularly men. 20–50% of cases have a family history of sudden death. Around 20-25% of cases have mutations in the SCN5A gene.

Clinical genetic testing is now commercially available. It is beneficial for first-degree family members to have genetic testing but only if there is a known genetic abnormality in the affected person. Time has now come for advising genetic counseling to the family members of the affected persons.

Citation: Shankar KJ. Genetic basis of arrhythmogenic right ventricular dysplasia and brugada syndrome. *Adv Cardiol Cardiovasc Disorders*. (2018);1(1):20.