



ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA

Arrhythmogenic right ventricular dysplasia (ARVD) is a rare disorder affecting the right ventricle, one of the two lower pumping chambers of the heart. In ARVD, some of the normal heart muscle cells are replaced by fatty material and scar tissue. Advanced cases may also affect the left ventricle. ARVD is important because it can cause potentially dangerous abnormal heart rhythms called “arrhythmias.” These arrhythmias are often dangerous and account for some episodes of sudden cardiac death in adolescents and young adults.

Although most cases of ARVD occur sporadically throughout the general population, it appears that 20-30% of cases are due to a genetic abnormality in a gene called plakophilin-2. This is a protein that binds the heart muscle cells together. When this protein is defective, the muscle cells lose contact with one another and are replaced by fat tissue.

Symptoms in affected persons are caused by the arrhythmias, which originate as abnormal electrical impulses from the right ventricle. These “ventricular arrhythmias” range from isolated extra heart beats which may cause palpitations but are not dangerous, to life-threatening arrhythmias called ventricular tachycardia and ventricular fibrillation. Sudden death often occurs during exercise.

ARVD usually becomes evident in adolescence or early adulthood. Although some patients have stable and well-tolerated symptoms for many years, others present with life-threatening arrhythmias as the initial manifestation of the disease. Unfortunately, it is often not possible to determine, based on the initial event, how dangerous the condition will be in the future.

The diagnosis of ARVD may be difficult at times, but it is usually possible if the disease is considered. Clues are sometimes found by recognizing abnormalities in the electrocardiogram (EKG), but the diagnosis needs to be confirmed by ultrasound examination of the heart (echocardiography), magnetic resonance imaging (MRI), or injection of contrast dye into the right ventricle during an angiogram. Occasionally a heart muscle biopsy is required. ARVD should be suspected in all patients with ventricular arrhythmias that appear to originate from the right ventricle, especially in young patients without known heart disease. The condition should be considered with unexplained arrhythmias, palpitations, or fainting episodes. Treatment depends on the severity of the symptoms and assessment of future risk. The latter is sometimes aided by an electrical study of the heart called an electrophysiologic study. Some patients may be treated effectively with “anti-arrhythmia” medications, while those at high risk may require the surgical placement of a device called an “implantable” cardioverter defibrillator.

For more information about ARVD, ask your physician and visit www.arvd.org and www.arvd.com.

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