RARELY SEEN IN UNDERWRITING ARTICLE SERIES: ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY (ARVC)

Michelle Privett, RN, MS, FALU, FLMI, PCS
***OTR* Contributing Editor**
Hannover Re
Denver, CO
michelle.privett@hlramerica.com

Executive Summary *Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an under-recognized cardiac condition characterized by ventricular arrhythmias and fibrous or fibro-fatty replacement of the myocardium. The mean age of diagnosis is 30 years, and male gender has been associated with a more malignant course. Lack of diagnosis may be due to the disease itself; ARVC patients are often asymptomatic for decades. Principal symptoms include dizziness, palpitations and syncope. Approximately 30% of cases are genetic in origin. ARVC has been found as a cause of sudden cardiac death in young adults, and strenuous exercise should be avoided in these patients. In addition to exercise restrictions, patients with ARVC are treated with implantation of a cardioverter-defibrillator (ICD) and antiarrhythmic drugs. Due to the severity and progressive nature of ARVC, an underwriting offer is not likely to be made.*

Introduction to the Rarely Seen in Underwriting Article Series

It's happened to all of us sooner or later in underwriting: open up a file or a set of medical records, and you find a medical diagnosis that might as well be in a foreign language. It happened to me recently, and I've been a registered nurse since 1995 and an underwriter since 2000. I opened up a set of medical records and saw it: ARVD.

ARVD? You mean aardvark? What does that have to do with underwriting? In a flash I pulled up Google and began reading about an underdiagnosed condition that I had never seen before in all of my years of working at two large academic medical centers, as well as several busy direct companies and three large reinsurers.

While I was looking up the condition, I thought about how performing an internet search is vital to the work of today's underwriter. Numerous times I have utilized the internet to search for a medical condition that isn't in the underwriting manual. Underwriting manuals try to cover as many medical conditions as possible, but it's impossible to have them all. Sooner or later you will find yourself researching a condition that isn't in the manual.

While I was looking up ARVD, it struck me as such an interesting condition that I thought I would share it with you. We see cardiomyopathy affecting the left ventricle routinely, but what about right ventricular failure?

It also struck me that there is a need for short articles on lesser-known conditions that might not be in the typical underwriting manual. Hence, the *Rarely Seen in Underwriting* article series was created. I hope you enjoy it.

What Is ARVC?

Arrhythmogenic right ventricular dysplasia (ARVD) is actually now called ARVC, or arrhythmogenic right ventricular cardiomyopathy. ARVC is an under-recognized cardiac condition characterized by ventricular arrhythmias and fibrous or fibro-fatty replacement of the myocardium. Remember fatty liver? Well, now think of fatty heart.

The normal myocardium of the right ventricle is replaced with both fibrous and fatty tissue as well as patchy inflammatory infiltrates. This leads to a

scarred appearance, right ventricle (RV) dilation and myocardial thinning.

Historically, ARVC was thought to be mainly characterized by pathologic right ventricular changes called the triangle of dysplasia, involving the RV inflow tract, RV outflow tract and apex of the right ventricle.

However, recent research has discovered the left ventricle is also affected. Most patients will also have left ventricular (LV) myocyte loss and fibrosis involving the LV lateral and posterior walls. Interestingly, the septum is usually spared.

Prevalence and Presentation

The prevalence of ARVC in the general adult population is estimated to be approximately 1 in 2,000 to 1 in 5,000. It is virtually never diagnosed in infants or toddlers and is rarely seen prior to the age of 10. The mean age of diagnosis is approximately 30 years, and male gender has been associated with a more malignant course. Interestingly, the disease has been rarely diagnosed in the US, but this is thought to be due to under-recognition of the disease. Genetic variance may also contribute to this.

Lack of diagnosis may also be due to the disease itself; ARVC often remains clinically silent, and patients are asymptomatic for decades. Principal symptoms including dizziness, palpitations and syncope, but can also include symptoms such as chest pain and dyspnea.

The most common ventricular arrhythmia is sustained or nonsustained monomorphic ventricular tachycardia that originates in the right ventricle, giving it a left bundle branch block pattern.

Genetics

Sharing a commonality with hypertrophic cardiomyopathy, ARVC is an often inherited cardiomyopathy that affects a significant number of first-degree relatives. Approximately 30% of cases are familial in origin. Of these genetically linked cases, there are two kinds of familial right ventricular cardiomyopathies:

a) Autosomal dominant form (the most common).
b) Autosomal recessive form (the least common called Naxos disease or Carvajal syndrome).

Naxos disease has all of the typical features of ARVC plus a cardiocutaneous syndrome, which includes physical characteristics of woolly hair and hyperkeratosis of the palms and soles.

The Risk of Sudden Cardiac Death (SCD)

ARVC has been found as a cause of sudden cardiac death in young adults. Unfortunately, sudden cardiac death may be the first clinical presentation of the dis-

ease, being found on autopsy after no explanation can be provided on why a young person died so suddenly.

Histologic autopsy evaluations of those with sudden cardiac death showed an active inflammatory process surrounding necrotic myocytes (heart cells). This acute or "hot" phase of the disease is thought to disrupt intercellular connections, resulting in myocyte cell death.

Treatment

Since exercise increases stress on the right ventricle and can increase RV dilation, patients with ARVC, or those at risk of developing ARVC due to family history, should avoid competitive and endurance exercise. Research has shown that both ventricular tachycardia and sudden cardiac death can occur due to strenuous exercise in patients with ARVC.

In addition to the restriction from high intensity exercise and recreational activities, patients with ARVC are treated with implantation of a cardioverter-defibrillator (ICD) and antiarrhythmic drugs. Radiofrequency ablation is not a definitive therapy due to the patchy and progressive nature of ARVC. Lastly, cardiac transplantation has been performed in rare occasions.

Underwriting Bottom Line

Mortality is high for individuals with ARVC making pricing of the risk difficult to impossible.

References

Maron MS. Hypertrophic cardiomyopathy: Clinical manifestations, diagnosis, and evaluation. McKenna WJ, ed. UpToDate. Waltham, MA: UpToDate Inc. www.uptodate.com (Accessed on November 13, 2017).

McKenna WJ. Arrhythmogenic right ventricular cardiomyopathy: Anatomy, histology, and clinical manifestations. Calkins H, ed. UpToDate. Waltham, MA: UpToDate Inc. www.uptodate.com (Accessed on November 13, 2017).

McKenna WJ. Arrhythmogenic right ventricular cardiomyopathy: Pathogenesis and genetics. Calkins H & Raby BA, ed. UpToDate. Waltham, MA: UpToDate Inc. www.uptodate.com (Accessed on November 13, 2017).

McKenna WJ. Arrhythmogenic right ventricular cardiomyopathy: Treatment and prognosis. Calkins H, ed. UpToDate. Waltham, MA: UpToDate Inc. www.uptodate.com (Accessed on November 13, 2017).