An overview of arrhythmogenic cardiomyopathy

Understanding conditions that can cause this disorder will help nurses educate and care for patients.

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CNE
1.5 contact hours

LEARNING OBJECTIVES

1. Compare the causes of arrhythmogenic cardiomyopathy (ACM) as to pathophysiology, clinical course, and management.

2. Discuss nursing implications for patients with ACM.

ARAN*, *an otherwise healthy 16-year-old South Asian male, arrives at the emergency department with paramedics after having a syncopal episode while watching a soccer game. The initial ECG shows polymorphic ventricular tachycardia, which requires cardioversion in the field. Slight ST-elevations in the right precordial leads are noted but no T-wave inversions. Aran denies any cardiac history, but a cardiology consultation with his parents reveals a family history of sudden cardiac death (SCD) and that the father has had peri-syncopal and syncopal episodes.*

An electrophysiologic study shows an easily inducible Type 1 ST-elevation, and programmed electrical stimulation induces ventricular tachycardia. Aran is diagnosed with Brugada syndrome (BrS). An implantable cardiac monitor is placed with plans for an implantable cardioverter defibrillator (ICD), prophylactic quinidine, regular follow-ups, and genetic testing.

Our understanding of the major risk factors of cardiovascular disease—such as diet, elevated blood pressure, high body mass index, increased total cholesterol, high fasting glucose level, smoking, and sedentary lifestyle—has increased, but little was known about arrhythmogenic cardiomyopathy (ACM) until recently.

ACM is defined as an arrhythmogenic disorder of the myocardium not secondary to ischemic, hypertensive, or valvular heart disease. Because of the low prevalence of conditions that lead to ACM, many healthcare providers may not be familiar with managing the disease or identifying those at risk. In 2019, the Heart Rhythm Society (HRS) issued a comprehensive consensus report on ACM management. This article provides an overview of selected genetic (BrS, left ventricular noncompaction), systemic (sarcoidosis, cardiac amyloidosis), infiltrative (cardiac amyloidosis), and infectious (Chagas disease) disorders that can lead to ACM. Although these conditions may result in a host of systemic complications, the discussion will focus on cardiac pathology and the role of nurses.

Brugada syndrome

BrS, which is named after the cardiologist Brugada brothers in Spain, is a genetic disease of the cardiac ion channel function. The syndrome causes dysfunction in the ion channels via up or down regulations, depending on the genetic mutation or variant involved, which can lead to arrhythmias. Patients with the disease can experience syncope, ventricular tachycardia (VT), ventricular fibrillation (VF), cardiac arrest, or SCD. The most common ECG manifestations are right bundle branch block (RBBB) and ST-segment elevations in the right precordial leads. BrS affects more men than women and especially those of South Asian or Japanese descent. El Sayed and colleagues report that overall prevalence is 5:10,000. The disease is the leading cause of SCD in men under 40 years and accounts for 12% of all SCD worldwide. (**Web resource: SADS.org**)

Pathophysiology

No clear pathophysiology for BrS exists, but genetic links are well established. When the disease was initially discovered, and up until a decade ago, most thought it was a condition of an otherwise structurally normal heart. Recent advances in higher resolution–computed tomography (CT) and cardiac magnetic resonance imaging (MRI) have revealed some structural abnormalities. However, because no pathognomonic pattern exists, the debate lingers as to whether the abnormalities cause BrS or are a result of it.

Genetically, an SCN5A mutation affecting the sodium channel flow is the most common (up to 30%) BrS variant. The mutation causes a decrease in the early peak current in the cardiac muscle and slows or blocks the conduction rate in parts of the heart, allowing for wave-break, which leads to arrhythmias. According to Brugada and colleagues, other genetic variants affecting the cardiac sodium, potassium, or calcium channel subunits account for 2% to 5% of inherited BrS cases. Estimates show that in up to 70% of patients with the inherited disease, no genetic variant has been identified.

Clinical course

BrS is diagnosed through analysis of an ECG. Three ECG patterns have been identified as hallmarks of the disease, all having ST elevations in the precordial leads; the most common is type 1 coved ST elevation. Under the

HRS guidelines, BrS is confirmed when a type 1 ST elevation is observed either spontaneously or after I.V. administration of a sodium channel blocker in at least one right precordial lead (V1 and V2) combined with at least one of the following: documented VF or polymorphic VT, inducibility of ventricular arrhythmias with programmed electrical stimulation, family history of SCD before age 45, history of non-vasovagal syncope, or nocturnal agonal breathing.

Patients with BrS can remain asymptomatic for life, but those who have symptoms, which can vary from syncope to SCD, frequently start to experience them close to the mean age of 40 years. Certain factors, such as medication or fever, can induce arrhythmias associated with BrS.

Management
Treatment options for patients with BrS are limited and guided by risk stratification. The recommended pharmacologic intervention is quinidine, which suppresses arrhythmias. Definitive treatment is an ICD. New research and current medical investigations are showing promising results in using ablation to stop frequent arrhythmic storms.

Left ventricular noncompaction

Left ventricular noncompaction (LVNC), also known as honeycomb myocardium, is a heart muscle disorder in which the left ventricle becomes thick and spongy, leaving the heart weak and unable to contract or relax properly. The National Institute of Health estimates that LVNC affects eight to 12 patients per 1 million annually; however, because of the sometimes-hidden clinical manifestations, its true prevalence may be underreported. Epidemiology is also hampered by the lack of consensus on a diagnostic standard. The condition usually affects the left ventricle, but in 22% to 38% of patients, the pathology is found in both ventricles. (**Web resource: cardiomyopathy.org/left-ventricular-non compaction/intro**)

Pathophysiology
Although the exact cause of LVNC is unknown, it's thought to result from a disruption of the compaction process during left ventricle myocardium development. Normally, the myocardium gradually compacts from the epicardium inward. When this fails, LVNC can result. Mutations in MYH7 and MYBPC3 genes have been found in 30% of patients with LVNC. These genes play a role in the function of sarcomeres, the structure within muscle fibers that causes contraction.

Clinical course
LVNC diagnosis requires echocardiography or cardiovascular MRI to measure the ratio of thickness, mass, or volume of the noncompacted layer of the left ventricle to the compacted layers. If the ratio of X (distance from epicardial surface to trough of trabecular recess) to Y (distance from epicardial surface to peak of trabeculation) is <0.5, the patient meets diagnostic criteria. Although some patients are asymptomatic, others may experience SCD, blood clots, frequent premature ventricular complexes, ventricular tachycardia, palpitation, and exercise intolerance because of the ventricle's inability to contract and relax normally. According to Towbin and Jefferies, two-thirds of individuals with LVNC develop heart failure (HF). Except for dilated and hypertrophic cardiomyopathy, LVNC frequently presents with features similar to those found in pulmonary atresia and intramyocardial hematoma.

Management
For the most part, LVNC treatment is symptom-specific, and HF is the most common reason for hospitalization. Treatment guidelines for HF should be applied on the basis of the patient's clinical scenario; treatment may include beta-blockers, ACE inhibitors, angiotensin-II receptor blockers, diuretics, and aldosterone antagonists. Antidysrhythmic therapy, such as ICD implantation and ablation, is recommended for patients with life-threatening dysrhythmia, especially those with nonsustained VT. In patients with atrial fibrillation or presence of atrial or ventricular clots, oral anticoagulation is recommended. The risks and benefits of heart transplant also should be explored.

Sarcoidosis

Sarcoidosis is a multisystem, inflammatory disease that primarily targets the lymph nodes and lungs. The hallmark manifestation is the accumulation of noncaseating granulomas

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(nonsolidifying areas of inflammation that consist of macrophages, epithelioid cells, mononuclear cells, and CD4+ T cells with a few peripheral CD8+ T cells) in these organs. The Mayo Clinic reported a sarcoidosis prevalence of approximately 4.7 to 64 per 100,000 with the highest rates among northern Europeans and Blacks.

According to Birnie and colleagues, the prevalence of sarcoidosis is notably higher in females (the female-to-male prevalence ratio of almost 2:1) and 70% of those affected are between the ages of 25 to 60 years old. Although the etiology of sarcoidosis is unknown, studies suggest that genetic predisposition and environmental factors play a role in its development. (**Web resource: stopsarcoidosis.org/**)

Pathophysiology
In cardiac sarcoidosis (CS), heart tissue becomes infiltrated with sarcoid granulomas; the myocardium is the most affected. This condition leads to three stages of cardiac pathologic features—edema, granulomatous inflammation, and fibrosis—which results in post-inflammatory scarring. Noncaseating granulomas form in the heart, primarily in the left ventricle, making the left ventricular ejection fraction the most important mortality indicator. Hulten and peers estimated that 20% to 25% of those with pulmonary/systemic sarcoidosis have asymptomatic cardiac involvement.

CS is the leading cause of death (up to 85%) in Japanese patients with sarcoidosis. Gene HLA DQB1*0601 and tumor necrosis factor allele TNFA2 found in Japanese patients are shown to predispose them to CS. According to Birnie and colleagues, angiotensin-converting enzymes are elevated in 60% of those with sarcoidosis; however, this biomarker lacks sensitivity and specificity for diagnosing the condition.

Clinical course
Sarcoidosis with cardiac involvement has a poor prognosis (it's life-threatening in 5% of patients). CS manifestations largely depend on noncaseating granuloma location and severity, but some of the primary features include conduction abnormalities and arrhythmias (such as AV block), HF, and SCD.

No diagnostic gold standard for CS exists, but fluorodeoxyglucose positron emission tomography of the heart may reveal inflammatory lesions in the myocardium. ECG may show AV blocks, QRS complex fragmentation, ST and T wave changes, pathological Q waves, and sometimes epsilon waves (a small positive deflection "blip" buried at the end of the QRS complex). Echocardiography may reveal myocardial wall thickness and left ventricular hypertrophy, which resembles hypertrophic cardiomyopathy. Heart biopsies also are used to determine the spread of noncaseating granuloma.

Management
Limited randomized controlled trials on the management and treatment of CS exist. Birnie and colleagues developed a helpful treatment algorithm. Corticosteroids will help slow inflammation and fibrosis progression. Anecdotal reports of antimalarial, methotrexate, and azathioprine use for patients who don't respond to corticosteroids or as steroid-sparing alternatives have been documented in the literature. Permanent pacemaker and ICD also are considered for patients with CS.

Cardiac amyloidosis

Cardiac amyloidosis is a form of restrictive cardiomyopathy caused by extracellular deposition of proteins in the myocardium. The ventricles become stiff and inflexible, leading to reductions in filling and, ultimately, cardiac output. Two types of cardiac amyloidosis exist: light-chain amyloidosis (AL) and transthyretin amyloidosis (ATTR), also known as transthyretin amyloid cardiomyopathy (ATTR-CM). ATTR is an inherited autosomal dominant trait (needs only one mutated gene to be affected by the condition) caused by pathogenic variants in the transthyretin gene TTR (ATTRv). According to Siddiqi and Ruberg, the incidence of AL is 1 per 100,000, resulting in 2,500 to 5,000 new cases annually in the United States. ATTR prevalence increases with age: 25% of those over age 80 have some type of amyloid deposition related to ATTR. The condition is rare, but according to Gilstrap and colleagues, from 2000 to 2012, a notable increase in prevalence and incidence rates occurred in the United States, particularly among older Black men. (**Web resource: bit.ly/35mQGjh**)

Pathophysiology
Unstable proteins are deposited in the myocardium, where they misfold and aggregate to become amyloid fibrils. In cardiac amyloi-

dosis, the pathology is mainly misfolded monoclonal immunoglobulin light chains (ALs) from an abnormal clonal proliferation of plasma cells or transthyretin (TTR), a liver-synthesized protein involved in the transportation of thyroxine and retinol-binding protein. Left ventricular hypertrophy is a key feature of cardiac amyloidosis, so left ventricular wall thickness ≥14 mm usually warrants evaluation for the condition.

Clinical course

Echocardiogram, cardiac MRI, and bone scintigraphy are used to diagnose ATTR-CM; Kittleson and colleagues developed a testing algorithm. Most patients have signs and symptoms of HF, such as dyspnea, fatigue, and peripheral edema. In patients with HF, clinical features that warrant evaluation for ATTR-CM include intolerance to antihypertensive or HF medication, persistent low-level elevation in serum troponin, discordance between QRS voltage on an ECG, wall thickness on imaging, unexplained AV block or left ventricular wall thickening, and family history of cardiomyopathy. On the other hand, AL tends to be more symptomatic before left ventricular wall thickening. In addition, heart murmurs might be detected because ATTR myocardial deposition has been noted in about a quarter of patients with degenerative aortic stenosis. Although imaging may create suspicion for amyloidosis, a cardiac biopsy is still the gold standard for diagnosis.

Management

Cardiac amyloidosis treatment focuses on managing HF and dysrhythmias and initiating disease-modifying agents. In symptomatic and asymptomatic individuals with cardiac amyloidosis and second-degree AV block type II, high-grade AV block or third-degree AV block, a permanent pacemaker is recommended. In patients with cardiac amyloidosis and symptomatic atrial arrhythmias, sotalol or amiodarone may be considered. Cardiac ablation is an option for those with cardiac amyloidosis and symptomatic atrial arrhythmias.

Disease-modifying agents are used as TTR stabilizers and silencers. *Stabilizers* are nonsteroidal anti-inflammatory agents (such as tafamidis) that help prevent misfolding and deposition of amyloid fibrils. *Silencers* (such as patisiran) target the hepatic synthesis of TTR, reducing TTR protein levels.

Experts suggest that cardiac amyloidosis should be considered in the differential diagnosis of patients ≥65 years with new onset HF. Consultation with a neurologist should be considered if neurologic involvement is evident or suspected.

Chagas disease

Named after the Brazilian physician Carlos Chagas, this zoonotic disease is caused by the protozoan parasite *Trypanosoma cruzi*, which is transmitted to humans by blood-sucking triatomine bugs. Less common modes of transmission are via blood product transfusion, organ transplantation, consumption of food or beverages contaminated by the vector or vector feces, and from mother to fetus. Chagas disease isn't endemic in the United States. According to Montgomery and colleagues, the approximately 300,000 chronic Chagas disease cases in the United States are among people originally from Latin American countries (Brazil, Bolivia, Argentina) where the disease is endemic. Between 1955 and 2015, only 28 confirmed cases of local transmission within the United States were documented. (**Web resource: who.int/health-topics/chagas-disease#tab=tab_1**)

Pathophysiology

How Chagas disease causes cardiomyopathy isn't well understood. Various explanations include antigens that persist in the heart, parasite-mediated myocytolysis, damage to cardiac microvasculature, antibody-mediated cytotoxicity, and nonspecific damage caused by eosinophils and neutrophils. A combination of these processes may lead to cardiomegaly, chronic myocarditis, and conduction system damage.

Notable rhythm abnormalities are RBBB and left anterior fascicular block. Years to decades after the acute infection, the patient might experience ventricular dysrhythmias, sinus node dysfunction and bradycardia, persistent or intermittent complete heart block, an apical aneurysm (usually in the left ventricle), thromboembolic events, and progressive dilated cardiomyopathy.

Clinical course

Although acute illness can last a few weeks or months, Chagas disease typically is undiag-

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nosed because symptoms are absent or mild and nonspecific (for example, fever and anorexia) even with a high degree of parasitemia. Diagnosis is confirmed during the acute phase via visualization of the parasite in a blood smear and during the chronic phase via immunoglobulin G serology.

In most cases, a prolonged chronic phase, in which the patient is asymptomatic and the parasitemia eventually resolves, follows the acute phase. About 20% to 30% of patients might develop life-threatening medical conditions, such as cardiomyopathy.

Management

Chagas disease can result in dilated cardiomyopathy, which ultimately can lead to HF. Treatment guidelines for HF are used for symptom management. Routine laboratory tests and cardiac biomarkers, ECG, and chest x-rays are integral to patient care. During the acute phase, the patient should be monitored for meningitis and encephalitis.

Heart transplant is an option for some patients with end-stage cardiomyopathy, although the patient should be monitored for signs of reactivation of the disease (such as acute myocarditis) in the transplanted heart. Reactivation of Chagas disease also is a risk among patients who are immunosuppressed (for example, those receiving chemotherapy) with the potential for cardiomyopathy and meningoencephalitis.

Public health efforts related to Chagas disease prevention focus on vector eradication in countries where the disease is endemic. Since 2007, blood donor screening for *T. cruzi* has become routine.

Nursing implications

RNs and nurse practitioners are integral members of interdisciplinary teams that care for patients with ACM. They're involved in comprehensive patient management using evidence-based interventions and protocols across all levels of care and transitions. In addition, they should be able to answer patient questions about exercise, exercise-induced arrhythmias, dietary modification (as they relate to overall health and medication interaction), sexual activity, pregnancy, and travel.

Patient history

A patient-centered interview is critical for di-

Nurses' role in testing

Nurses can help explain testing, interpret results, and provide support to anxious patients and families.

Standard lab testing. Complete blood count, metabolic panel, liver function tests, lipid profile, and diabetes and heart failure markers (for example, brain natriuretic peptide) establish a baseline and indicate treatment effectiveness trends. Nurses can explain the various tests and put results into context.

Genetic testing. Nurses can help patients understand procedures and results and explain how testing can be used for risk stratification or management.

Cardiac studies. Diagnosis by exclusion might require the patient to undergo several procedures—from noninvasive (ECG, echocardiogram, and cardiac imaging) to invasive (electrophysiology studies and endomyocardial biopsy). Nurses can explain pre- and post-testing expectations to patients and help allay their anxiety. With 12-lead ECGs and Holter monitoring, nurses can ensure correct lead placement to minimize artifacts and variability. In addition, nurses should have advanced ECG interpretation skills.

agnosing ACM. Obtain a detailed three-generation medical history looking for patterns of genetic and nongenetic causes (such as SCD) of ACM. Don't rush the history and use a medical interpreter as needed. A diagnostic algorithm (jamanetwork.com/journals/jama/full article/209410) can help exclude ischemic heart disease, hypertensive heart disease, and valvular heart disease as causes of cardiomyopathy. Some therapies, such as those for cardiac amyloidosis, are most effective when administered before significant cardiac dysfunction occurs, highlighting the importance of early identification.

Testing

Diagnostic testing includes standard lab tests, genetic tests, and cardiac studies. (See *Nurses' role in testing*.)

Pacemaker and ICD

If a patient with ACM (such as those with cardiac amyloidosis or BrS) requires a permanent pacemaker or ICD, implement best practices across the perioperative continuum and follow up. For example, after pacemaker or ICD placement, resume cardiac medications as prescribed, warn the patient to avoid heavy lifting on the same side as the device, and instruct the patient to notify their provider if they feel a "shock." If you care for patients with an ICD or pacemaker who have surgery that requires electrocautery, remember that the ground return pad should be placed as far from the device and as close to the surgical site as possible.

Symptom management and medications

Most patients with ACM take multiple medications to manage symptoms and prevent complications. Medications may include anticoagulants, steroids, non-steroidal anti-inflammatory agents, chemotherapy, disease-modifying agents, antihypertensives, diuretics, and antidysrhythmics. Beta-blocker therapy is recommended in patients experiencing inappropriate ICD shocks. Where applicable, the 2017 guidelines for heart failure management should be followed.

Medication reconciliation is key to preventing drug–drug interactions, prescription cascade, and polypharmacy. Teach patients and caregivers how to accurately monitor blood pressure and heart rate at home.

Referral

As a patient advocate, you can help navigate referrals to multiple providers with expertise in genetics, cardiology, rheumatology, neurology, and other modalities. Patients also might require physical and occupational therapy and home health assistance. Close collaboration with a case manager can make this care coordination more manageable.

Psychosocial support

Living with the prospect of SCD can be immensely unsettling. In addition, disability, lack of sufficient health insurance coverage, and other socioeconomic challenges can increase the burden of ACM. As a member of the care team, offer emotional support to patients and their caregivers and provide referrals to support groups.

Aran

After the initial episode that led to his diagnosis with BrS, Aran remains symptom-free for 8 years. At 24, he has influenza and fever, which induces a syncopal episode with polymorphic VT that requires cardioversion. Genetic testing confirms a TRPM4 mutation. Nocturnal bradycardia is noted during two hospitalizations. After ICD placement at age 28, Aran has a single shockable event but otherwise remains healthy with occasional asymptomatic bradycardia at night. After a non-cardiac related surgery, Aran develops type 1 BrS ECG changes (coved ST elevation), most likely due to the use of propofol. He's now 30 years old and remains free of any BrS-associated complications and has regular outpatient EP follow-up.

Increased understanding

ACMs are low-frequency, high-stakes conditions that include a host of genetic, systemic, infectious, and inflammatory disorders. With advances in science and technology, understanding of ACM continues to increase, leading to the development of consensus statements and treatment algorithms to improve outcomes. Because the conditions associated with ACM are relatively rare, patients are most likely seen in specialty cardiac service lines. Nurses who work in acute care, coronary care, and primary care settings should develop clinical skills to comprehensively manage and support these patients and their caregivers.   **AN**

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*Name is fictitious.

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Please mark the correct answer online.

1. Your patient's ECG shows right bundle branch block and ST-segment elevations in leads V1 and V2. This could indicate
- a. Chagas disease.
- b. sarcoidosis.
- c. left ventricular noncompaction (LVNC).
- d. Brugada syndrome (BrS).

2. Which of the following is caused by a protozoan parasite?
- a. Chagas disease
- b. Sarcoidosis
- c. LVNC
- d. BrS

3. What is the most common reason for patients with LVNC to require hospitalization?
- a. Ventricular tachycardia
- b. Atrial fibrillation
- c. Heart failure
- d. Stroke

4. Which statement about the pathophysiology of cardiac amyloidosis is correct?
- a. Amyloid granulomas form in the myocardium tissue, primarily in the left ventricle.
- b. Amyloid granulomas form in the myocardium tissue, primarily in the right ventricle.
- c. Unstable carbohydrates in the myocardium misfold and aggregate to become amyloid fibrils.
- d. Unstable proteins in the myocardium misfold and aggregate to become amyloid fibrils.

5. Which cause of arrythmogenic cardiomyopathy (ACM) primarily targets the lungs and lymph nodes?
- a. Chagas disease
- b. Sarcoidosis
- c. LVNC
- d. BrS

6. Which of the following would prompt you to suggest your patient with heart failure be evaluated for transthyretin cardiac amyloidosis (ATTR-CM)?
- a. Intolerance to antihypertensive medications
- b. Decreased troponin levels
- c. Left anterior fascicular block on ECG
- d. Family history of stroke

7. Your patient's ECG shows epsilon waves, which may indicate
- a. Chagas disease.
- b. cardiac amyloidosis.
- c. sarcoidosis.
- d. BrS.

8. Which statement about the treatment of ATTR-CM cardiac amyloidosis is correct?
- a. Cardiac ablation is contraindicated.
- b. Patisiran is used to manage ventricular arrhythmias.
- c. Anti-inflammatory agents are used to target hepatic synthesis of TTR.
- d. Treatment includes both stabilizers and silencers.

9. Nurses caring for patients with suspected or diagnosed ACM should do all of the following *except:*
- a. obtain a two-generation medical history.
- b. explain how genetic testing can be used.
- c. perform medication reconciliation.
- d. provide referrals to support groups.

10. The definitive treatment for BrS is
- a. a pacemaker.
- b. an aldosterone antagonist.
- c. an implantable cardioverter defibrillator.
- d. a beta-blocker.

11. Mutations in MYH7 and MYBPC3 genes have been found in 30% of patients with
- a. sarcoidosis.
- b. Chagas disease.
- c. LVNC.
- d. BrS.