



Myocarditis Education for Emergency Room Nurses

Produced by:

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With the assistance of:

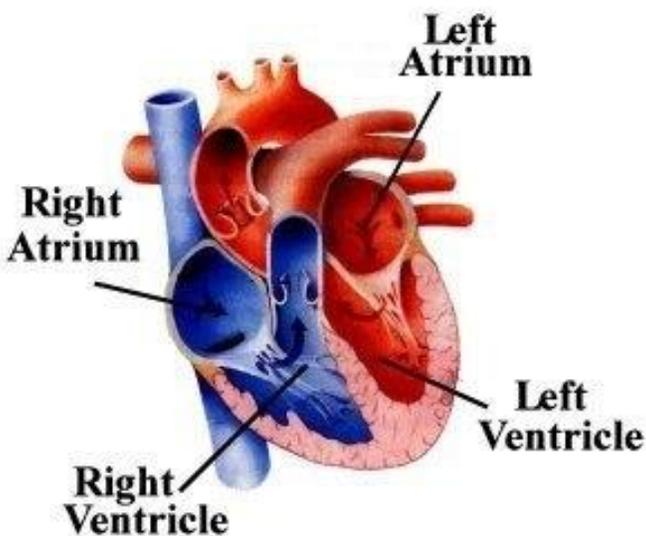
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The Heart

The heart is a four-chambered muscular pump, in an adult it is about the size of an adult fist and in children it is about the size of their fist.

Normally, the heart beats 60-100 times per minute, pumping blood throughout your body with each beat.



Two upper chambers called the right and left atria (each is called an atrium), receive blood that returns to the heart from the body. Veins carry this returning blood to the atria.

When the muscles of the atria contract, blood is squeezed into the two larger, lower chambers called the right and left ventricles. When the muscles of the ventricles contract,

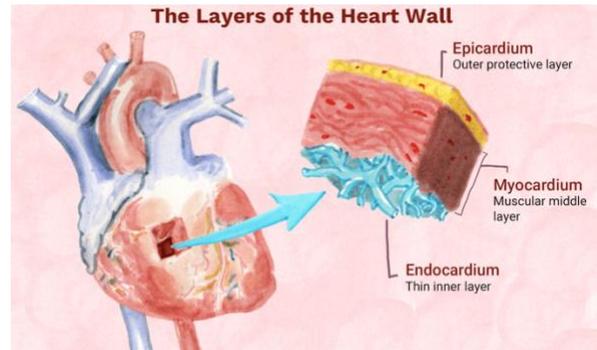
blood is propelled throughout the arteries to the entire body. The pumping of the ventricles creates the pulse that you feel in your wrist or your neck.

An Ejection Fraction (EF) refers to the percentage of how much blood is pumped out of a filled ventricle with each heartbeat. A normal Ejection Fraction is between 55% and 75%, which means that over half of the blood volume is pumped out of the heart with each beat. When your heart relaxes, the ventricles refill with blood. No matter how forceful the contraction, it is never able to pump all of the blood out of a ventricle. One reason for a decreased EF, (Less than 55%) is dilated cardiomyopathy, which is often caused by Myocarditis.

What is Myocarditis?

(The term Myocarditis will refer to Viral Myocarditis unless otherwise specified)

The Myocardium, the middle layer of the heart wall, is composed of heart muscle. The term, Myocarditis, was first designated in Germany in 1837 after inflammation of the heart muscle was found on autopsy. Not much was spoken again about this for the next 100+ years. Over the past 25 years, there has been an increase in interest and research into Myocarditis.



There are over 7,000 rare diseases globally. Myocarditis is considered a rare disease, but many believe that Viral Myocarditis is more common than is estimated.

Key figures for a disease to be defined as a rare disease:

In Europe – when a disease affects fewer than 1 in 2,000.

In USA – when it is diagnosed less than 200,000 times in any given year.

In 2016, there were 2.2 million cases of Myocarditis correctly diagnosed globally.

In 2017, there were 3.1 million cases of Myocarditis correctly diagnosed globally.

The increase in the numbers is not because the numbers are worsening, but because it is correctly getting diagnosed due to increased awareness and education on the disease.

Myocarditis is the 20th leading cause of death worldwide

In the past, Myocarditis was never taught in medical school, and as such is not readily diagnosed. Only recently, with the help of Dr. Leslie Cooper, of the Mayo Clinic and co-founder of the Myocarditis Foundation, Myocarditis is being taught in medical schools around the world.



Myocarditis usually attacks otherwise healthy children and young adults. It can attack anyone of any age. There are many causes, including viral infections, environmental toxins (chemicals), parasites, and adverse reactions to medications. The most common cause of Myocarditis in the developed world is viruses.

Enteroviruses, most frequently Coxsackie-Virus B, are historically implicated as a common cause of this disease. The difficulty with the diagnosis is that it so often presents as a viral illness. In a majority of cases, the symptoms of Myocarditis are preceded a few days to weeks by a flu-like illness. Thus, it is extremely important to obtain a good medical history from the patient/parent to support the possibility of a diagnosis of Myocarditis.

Many infants and children with Myocarditis present with rapid and labored breathing, wheezing and grunting, often mimicking more common childhood diseases such as Asthma, Bronchiolitis, and Gastroenteritis.

Older children and young adults may complain of fatigue, fever, cough, nausea/vomiting, weakness, heaviness of legs and muscle aches a few days before developing more severe symptoms such as shortness of breath and exercise intolerance. Often Flu, Gastroenteritis and Asthma are the diagnoses given.

More advanced symptoms include rapid heart rate, chest pain (often a tightness or squeezing in the chest), back pain, erratic and weakened pulse, pallor, cardiac arrhythmias, diaphoresis and lightheadedness/dizziness. Dehydration is often considered the cause.

Myocarditis can be found in people of all ages, but the most common high-risk population brackets are young children with underdeveloped immune systems and those puberty/teenage through young adulthood up to age 40.

Twice as many men as women are affected by Myocarditis.

Myocarditis is the 3rd leading cause of Sudden Death in children and young adults.

20% of Sudden Deaths in the United States are a result of Myocarditis.



Myocarditis often can mimic many viral diseases and is often misdiagnosed. Many times, a patient is examined two or more times before a correct diagnosis is made.

A study conducted at Texas Children's Hospital of 193 consecutive children admitted with Heart Failure (50% were seen by their PCP and 50% by ER staff) proved that 52% had a missed diagnosis and an average encounter of 2 or more times before a correct diagnosis was made.

In another study, of 171 children hospitalized with Myocarditis:

- 24% of them were less than 2 years of age and 46% of them were between the ages of 14 to 18 years of age
- 58% were male
- 99% had respiratory, gastrointestinal or cardiac symptoms
- Average heart size was within normal limits on Echocardiogram
- Average BNP level: 1102 pg/ml (normal <50)
- Average Troponin I Level: 4.9 ng/ml (normal <0.2)

Butts RJ, et al. ISHLT abstract 2016



Including Myocarditis as a possibility of non-specific symptoms can be lifesaving. Most with Myocarditis recover with treatment, but a substantial percentage may progress to the long-term side effect of cardiomyopathy and progressive heart failure, leading to cardiac transplantation or death.

Subclinical Myocarditis (a mild case where the patient has mild symptoms or is asymptomatic) can progress to Dilated Cardiomyopathy and Chronic Heart Failure. Thus, obtaining a detailed history of recent complaints no matter how minor, can impact the outcomes to the disease. Prompt diagnosis is imperative to allow for rapid and appropriate treatment.

Idiopathic Giant Cell Myocarditis (IGCM) is a rapidly fatal disorder that may respond to certain immunosuppressive drugs or heart transplantation. From 1905 (when it was first discovered) until 1987, all cases of IGCM were described at autopsy, with survival usually less than 3 months from the initial onset of symptoms. We now know that with early diagnosis by heart biopsy and prompt immunosuppressive treatment, 90% of IGCM patients survive at least one year. IGCM can only be diagnosed by a heart biopsy.

How to Possibly Diagnose Myocarditis?

Although there is no specific tool to pinpoint that someone has Myocarditis, there are a number of ways that could raise red flags and potentially point to Myocarditis.

Myocarditis is classified based upon the type and severity of presenting symptoms.



Fulminant Myocarditis appears to be preceded by a viral syndrome followed by acute onset of cardio-pulmonary signs and symptoms consistent with impending shock.

Acute Myocarditis often has a milder, less distinct presentation but more often progresses to dilated cardiomyopathy and heart failure.

Chronic Myocarditis, as the name suggests, is persistent and may be latent or progressive with possible recurrences requiring ongoing medical therapy.

Idiopathic Giant Cell Myocarditis (IGCM), as the name suggests, has no known specific cause. To perform a cardiac biopsy, which is the only way it can be diagnosed, there must be some evidence of heart failure. IGCM is the deadliest form of Myocarditis.

Myocarditis should be suspected in people who have recent onset cardiac symptoms, such as chest pain or trouble breathing, and who have no evidence of more common disorders such as coronary artery disease, heart valve damage, or severe high blood pressure.

It is important to keep Acute Myocarditis in the differential diagnosis of a patient who presents with new signs or symptoms of heart failure (which can initially look like a viral syndrome with vomiting, diarrhea, fever, cough, shortness of breath, fatigue...) especially in children and young adults.

Before you diagnose a flu or other viral illness, do a thorough physical exam. Do not just place the symptoms in a box marked “viral syndrome, flu, gastroenteritis, etc.” and go by that for a diagnosis.

A patient who has been to see you once or even twice with viral syndrome complaints in the past few days and does not seem to be improving or in fact getting worse is in itself a red flag for Myocarditis. It is extremely important to maintain a high degree of suspicion for potential Myocarditis when assessing patients.

Findings on a patient history may include:



- A complaint of chest tightness, back pain, weakness, heavy feeling in their legs, palpitations, feeling of lightheadedness/dizziness, shortness of breath, fatigue, and no history of Cardiac Disease

- Shortness of breath and the inability to lie flat
- Rales, wheezing, shortness of breath, grunting
- No history of Asthma
- Usually an otherwise healthy, often athletic, individual
- Decreased exercise tolerance
- Recent history of a recent viral infection
- Even a recent slight cold, runny nose, viral type illness, that often doesn't stop a person's normal activity
- Abdominal discomfort, vomiting and diarrhea

Findings on physical exam may include:

- Hepatomegaly (From fluid overload with backup to the liver)
- Peripheral edema
- Abnormal heart sounds (murmur of mitral regurgitation) or a gallop rhythm
- Tachycardia (the body tries to compensate for inadequate tissue oxygenation secondary to diminished cardiac output); rhythm disturbances
- Delayed capillary refill
- Elevated jugular venous distention (JVP)
- Crackles or rales on auscultation of lung fields
- Wheezing, grunting, tachypnea
- Tachycardia
- Evidence of poor perfusion (altered mental status, low blood pressure, end-organ dysfunction)



Diagnostic testing can help to discriminate heart failure symptoms from other common illnesses that Myocarditis mimics.

Diagnostic Testing should include:

- B-type natriuretic peptide concentration (BNP), is a non-specific hormonal marker of stress or volume loading of the ventricle. **BNPs will be elevated in Heart Failure patients, such as those with Myocarditis**, but cannot distinguish Myocarditis from other causes of heart failure.



- **Troponin Levels (Troponin I, Troponin-C, Troponin-T concentration)**, a marker of myocardial injury that **may or may not be elevated in children with Myocarditis**
- **Myoglobin** is released during cardiac injury and increases with severity of myocarditis. Since myoglobin is available in most emergency rooms as a routine laboratory test it will add value in helping to potentially diagnose myocarditis.



- **CBC with differential, WBC, ESR, CRP, urea, creatinine, liver enzymes, and CK-MB elevated levels may be indicative**
- Potential findings on an **Electrocardiograph (EKG)** can be: Sinus Tachycardia, Ventricular Hypertrophy, ST segment and T wave abnormalities, Arrhythmia, Bundle Branch Block, Prolonged QTc, AV Block, Abnormal QRS axis, Low voltage QRS

- **Echocardiography is of great importance in the diagnosis of myocarditis**, especially to rule out other causes of heart failure and to look for depressed left ventricular systolic function, mitral regurgitation, atrioventricular valve regurgitation, dilated left ventricle, regional wall motion abnormalities, thickened ventricular wall or septum, or thrombus.
- An Ejection Fraction (EF) of 55-75% is considered within normal limits; an EF of 36-54% considers the heart's pumping ability to be Below Normal; and an EF of below 35% considers the heart's pumping ability is low.
- **Chest Radiograph (X-Ray) to look for cardiomegaly, pulmonary vascular congestion, and pleural effusion.** Cardiomegaly and pulmonary vascular congestion are important findings on a chest radiograph in the myocarditis patients **and can help to distinguish a diagnosis of Myocarditis from more common respiratory ailments.** However, these radiographic features may not be evident in cases presenting as Fulminant Myocarditis. In patients presenting with shock or impending shock of unknown cause without cardiomegaly or pulmonary vascular congestion on chest radiograph, a diagnosis of Myocarditis cannot be excluded.
- A **Cardiac MRI** can reveal myocardial edema and ischemia that often occur in Myocarditis, though specificity is lacking. Even though it is the best test to diagnose Myocarditis, even at its best it is still only 80% correct. 20% can be falsely negative. The further out from the initial episode of Myocarditis, the less specific the MRI will be. More studies are being done on the prognostic value of Cardiac MRI.

- **Endomyocardial (Cardiac) Biopsy** has been considered the “gold standard” for confirming a diagnosis of Acute Myocarditis. However, because of the patchy distribution of inflammation in the myocardium, the cardiac biopsy in acute myocarditis is fraught with a high false negative rate. The invasive nature of obtaining a myocardial tissue sample poses a significant risk to the patient and should only be utilized when the clinical suspicion is high for a cardiac disorder that is amenable for treatment.

In the case of trying to diagnose Giant Cell Myocarditis however, a Cardiac Biopsy is needed to look for the Giant Cells that it presents with.

CBC = Complete Blood Count; WBC = white blood count; CK-MB = Creatine kinase muscle, brain; CRP = C-Reactive Protein; ESR = erythrocyte sedimentation rate; QTc = corrected QT interval;
AV = atrioventricular

Treatment and Outcome:

If Myocarditis is suspected, immediate referral to a cardiologist is highly recommended.

Clinical deterioration may occur rapidly, especially in small children. Most children and adults with Myocarditis will recover with supportive care but a substantial percentage may develop progressive heart failure leading to death or need for cardiac transplantation.



Myocarditis accounts for 45% of Heart Transplants in the United States.

The mainstay of therapy for acute myocarditis is supportive therapy for left ventricular dysfunction. Most patients will improve with a standard heart failure regime according to the current American Heart Association and the Heart Failure Society of America Guidelines. In patients with acute myocarditis, therapy for arrhythmias is also supportive, since such arrhythmias usually resolve after the acute phase of the disease, which can last several weeks. As well, temporary pacemakers may be required for patients with symptomatic bradycardia or complete heart block. Patients with symptomatic or sustained ventricular arrhythmias may require an implantable pacemaker-defibrillator, even if active inflammation is still present.

In patients whose condition deteriorates despite optimal medical management, in those with shock or impending shock, case scenarios suggest a role for mechanical circulatory support, such as Ventricular Assist Devices or Extracorporeal Membrane Oxygenation (ECMO). Immediate transfer to a facility that can support this is necessary.

In some situations, death is sudden and unexpected. A diagnosis of Myocarditis is not made until a post-mortem exam is performed in these cases.

In Giant Cell Myocarditis (IGCM), diagnosis can only be confirmed by Cardiac Biopsy.

Typically, when a patient exhibits heart failure this disease is attacking the heart very aggressively and the only option is immediate treatment. One treatment is to give the patient a variety of immunosuppressive drugs combined with steroids. Another option is a heart transplant.

The goal of immunosuppression therapy is to delay heart transplantation. The need for a transplant may be delayed by months or even years. Since each case is unique there is not a typical time.



Patients recovering from acute myocarditis should refrain from aerobic activity for a period of months after the clinical onset of the disease. In a study conducted on sports-related sudden death in the general population, it is noted that it is more common than previously noted.

Athletes with probable or definitive myocarditis should not participate in competitive sports while active inflammation is present. This recommendation is independent of age, gender and Left Ventricular function. **In the latest guidelines from JAMA Cardiology, 5/13/2020, risk stratification may occur after 3 to 6 months of exercise restrictions and is based on extensive testing including echocardiography, stress testing, and rhythm monitoring. Return to competitive sports and exertional activities after myocarditis, is predicated on normalization of ventricular function, absence of biomarker evidence of inflammation, and absence of inducible arrhythmias.**

There is no sensitive or specific test that can determine when the inflammatory process ends.

Patients in whom the findings of acute inflammation have resolved may still have a risk of arrhythmias related to the resultant myocardial scar which still may be very irritable.

Dilated Cardiomyopathy (DCM) associated with acute myocarditis often resolves over 6-12 months.

References:

Price, J.F., Myocarditis and Heart Failure in Children: How to Make the Diagnosis, March 2017

Cooper, Leslie T. Jr, Myocarditis, New England J of Medicine April 9, 2009 p 40-52

Maron et al, Competitive Athletes: Hypertrophic Cardiomyopathy, ARVC, and Myocarditis
AHA Journal of Medicine download by Dr. L. Cooper at Mayo Clinic Libraries,
January 9, 2016
p e276-e277

Marijon et al, Sports-Related Sudden Death in the General Population,
Circulation, AHA Journals, June 2011, p 50-59

Myocarditis Foundation Pamphlet on: Myocarditis and Giant Cell
Myocarditis, updated 2021

Myocarditis Foundation Pamphlet on: Be Alert to Myocarditis in Children: A
Guide for Physicians, updated 2021

“Myoglobin for Detection of High-Risk Patients with Acute Myocarditis”
The Journal of Cardiovascular Translational Research, 31 January 2020