What is Myocarditis?

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

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. As awareness and education surrounding this disease are improved, we are noticing that the numbers of those being diagnosed are increasing – not because there are more cases, but rather since symptoms are being recognised more readily and more correct subsequent diagnosis are being made.

Myocarditis is the 20th leading cause of death worldwide

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 in the developed world for myocarditis 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 most 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 laboured 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 light-headedness/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.

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.

How to Possibly Diagnose Myocarditis?

Although there is no specific tool to pinpoint that someone has Myocarditis, there are several 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 light headedness/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 diarrhoea

Findings on physical exam may include:

- Hepatomegaly (From fluid overload with backup to the liver)
- Peripheral oedema
- 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, tachypnoea
- 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.

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.

Most patients will improve with a standard heart failure regime. 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 some situations, death is sudden and unexpected. A diagnosis of Myocarditis is not made until a post-mortem exam is performed in these cases.

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.

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Alexander Jansons Myocarditis UK would like to thank the www.myocarditisfoundation.org For compiling this data and allowing us to use it to further the awareness and education of medical professionals in the UK