THE
MYOCARDITIS
FOUNDATION

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The Foundation board is comprised of medical professionals with experience in myocarditis and lay persons who have been touched by the disease.

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Myocarditis Foundation

You Can Help, Please Donate:

By Mail: **The Myocarditis Foundation**
3518 Echo Mountain Drive
Kingwood, Texas 77345

Online: **www.myocarditisfoundation.org**
Click DONATE Link

The Myocarditis Foundation (MF) seeks to increase awareness and hasten progress in understanding myocarditis by awarding grants to help guarantee that new and innovative research avenues are thoroughly funded and explored. Please donate now.

The MF is a private, non-profit organization that exists to educate physicians and the public about this rare disease and support the patients and their families who have been affected by the disease. Copies of our materials will be available without charge. All of the money donated to MF will go directly to programs and services.

For more information:
info@myocarditisfoundation.org

MyocarditisFoundation.org
Website Resources

- Up-to-Date Medical information
- Resources for Medical Professionals
- Real-Life Stories
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Be Alert to Myocarditis in Children: A Guide for Physicians

MYOCARDITIS
FOUNDATION

Knowledge Nurtures Hope. . .

Your journey is just beginning

The Myocarditis Foundation is here to help.

www.myocarditisfoundation.org

DEFINITION: What is Myocarditis?

Myocarditis is a rare, potentially life-threatening inflammatory disorder of the myocardium. It is a common cause of heart failure in otherwise healthy children and accounts for up to one-third of the cases of pediatric dilated cardiomyopathy. The true incidence of myocarditis in children is unknown. Some cases are subclinical, making it difficult to make a timely and accurate diagnosis. Recognition is made difficult in very young children because they cannot clearly describe their symptoms. Moreover, the signs and symptoms often mimic those of more common childhood diseases such as asthma, bronchiolitis, and gastroenteritis. Including myocarditis in the differential diagnosis of children presenting with nonspecific symptomatology can be lifesaving. Most children with myocarditis recover with treatment, but a substantial percentage may develop progressive heart failure leading to cardiac transplantation or death. Prompt diagnosis is imperative to allow for rapid and appropriate treatment of these children.

ETIOLOGY

In the developed world, the most common causes of pediatric myocarditis are viral infections. Enteroviruses, most frequently coxsackievirus B, were historically implicated as a common cause of this disease in children, although many other infectious agents have since been identified. Other viruses such as influenza, adenovirus, and parvovirus B19 as well as bacteria, fungi, protozoa, rickettsiae and parasites may also cause myocardial inflammation. Immune-mediated diseases such as collagen vascular diseases, venoms, toxins, and some chemotherapeutic agents can also lead to myocarditis.

EPIDEMIOLOGY

Myocarditis may occur more frequently during seasonal influenza epidemics and during the summer and fall, a time of increased prevalence of coxsackievirus B in the general population. Neonatal myocarditis usually presents acutely and severely with a mortality rate reported around 75%. Infants are also quite susceptible to viral myocarditis but the incidence decreases during the toddler and early childhood years. Around the time of puberty and adolescence, the incidence increases again. In this older population, myocarditis is a significant contributor to sudden cardiac death, as verified on post-mortem examination. Gender differences have also been observed. Approximately two-thirds to three-quarters of coxsackievirus B cases occur in males.

CLINICAL PRESENTATION

Signs and symptoms of myocarditis are highly variable in the pediatric population. Children may present with complaints of mild, non-specific flu-like symptoms or with evidence suggestive cardiac involvement such as acute chest pain. A subset of patients recall a history of a viral prodrome followed by the sudden onset of symptoms consistent with cardio-pulmonary disease such as chest pain, tachypnea, dyspnea, fatigue, pallor, lethargy or cyanosis, suggesting hemodynamic compromise.

A three-tiered classification of myocarditis is based upon the type and severity of presenting symptoms coupled with the clinical course and outcomes. Fulminant myocarditis appears to be preceded by a viral prodrome in children followed by acute onset of cardio-pulmonary signs and symptoms consistent with impending shock. Despite the serious nature of the initial illness, the prognosis for recovery and long-term survival is excellent if the patients survive the acute episode. Acute myocarditis 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.

DIAGNOSIS

The variable nature of the pediatric myocarditis patients' presenting signs and symptoms makes it difficult to diagnose accurately. In fact, the disease is often missed at the initial physician encounter. The consequences of a missed diagnosis can be dire, highlighting the need for physicians to maintain a high degree of suspicion for myocarditis when assessing these patients. Common findings on exam are: Tachycardia partially compensates for inadequate tissue oxygenation secondary to diminished cardiac output. Decreased peripheral perfusion manifests as cool extremities, weak pulses, pallor, increased time to capillary refill, and/or decreased urine output. Vasoconstriction initially maintains an age-normal blood pressure but hypotension may occur as a late finding of cardiac failure in children. A variety of abnormal heart sounds may be detected, including diminished heart sounds, murmurs, gallops and rhythm disturbances. Wheezing, coughing, grunting, nasal flaring, intercostal retractions, rales, dyspnea, tachypnea, cyanosis and hypoxia are evidence of respiratory distress. Hepatomegaly and peripheral edema suggest cardiac failure in severe cases. Non-specific findings include fever, malaise, anorexia, fatigue and lethargy.

FURTHER DIAGNOSTIC EVALUATION

Common findings on diagnostic evaluation in pediatric myocarditis are:

Chest radiography: 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 vascualr congestion on chest radiograph, a diagnosis of myocarditis cannot be excluded.

Electrocardiography: The electrocardiogram (ECG) is usually abnormal in pediatric myocarditis patients. Findings on ECG may include sinus tachycardia, low voltage QRS complexes, ST-T wave abnormalities, prlonged QT intervals and/or atrioventricular block. A variety of tachy- and brady-arrhythmias including ventricular tachycardia and third degree atrioventricular block may be observed in pediatric myocarditis.

Laboratory studies: Biomarkers of cardiac injury, including troponins, have variable diagnostic yeild. Viral serologies and peripheral blood and tissue cultures are frequently negative not helpful in achieving an early diagnosis. B-type natriuretic peptide (BNP) is a non-specific hormonal marker of stress or volume loading of the ventricle. Concentrations of BNP are elevated in heart failure patients but cannot distinguish myocarditis from other etiologies of heart failure.

The *endomyocardial biopsy* remains the gold standard for diagnosis of viral myocarditis despite the low sensitivity and high rate of false negative test results. Due to the focal nature of myocardial inflammation, biopsy samples may fail to include affected areas causing a missed diagnosis. Polymerase chain reaction (PCR) of myocardial tissue for viral genome can increase the sensitivity of the endomyocardial biopsy. 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 to treatment.

FURTHER DIAGNOSTIC EVALUATION Continued

Echocardiogram: The echocardiogram is a ueful tool for evaluating ventricular function and excluding more common causes of heart failure. Echocardiographic findings are variable and may include wall motion abnormalities, ventricular chamber dilation, atrial enlargement, atrioventricular valve regurgitation, and left ventricular or biventricular dysfunction. Fulminant myocarditis is often associated with an echocardiographic picture of reduced left ventricular systolic function without left ventricular chamber dilation.

Cardiac MRI: Contrast-enhanced cardiac MRI may assist in the diagnosis by identifying local sites of suspected inflammation in the myocardium. The prognostic value of MRI tissue characterization is an area of active research.

PROGNOSIS

Most children with myocarditis recover with supportive care, but a substantial percentage may develop progressive heart failure leading to cardiac transplantation or death. In some situations, death is sudden and unexpected and a diagnosis of myocarditis is not made until post-mortem examination is performed. Prompt diagnosis is imperative to allow for rapid and appropriate treatment of these children. There is no virus-specific therapy for myocarditis. Treatment is focused on correcting hemodynamic derangements, optimizing cardiac output, and providing symptomatic relief. In children with shock or impending shock, temporary mechanical circulatory support (ECMO or ventricular assist devices) can maintain circulation and end-organ perfusion, allowing some patients to bridge to myocardial recovery.