

### DEFINITION: What is Myocarditis?

Myocarditis is a rare, sometimes fatal disease characterized by inflammation of the heart muscle. The disease can affect the muscle cells of the heart (myocytes), the heart valves and blood vessels, and the specialized electrical conduction pathways within the heart.

Most patients recover completely from acute or sudden myocarditis. In newborns, myocarditis can be severe due to an immature immune system. Other organ systems may also be affected in this group of patients. Myocarditis is an important cause of sudden death in young athletes.

When the muscle cells of the heart weaken from myocarditis, the heart chambers may enlarge. This form of heart disease is called dilated cardiomyopathy. Children with dilated cardiomyopathy may develop symptoms of heart failure, sometimes necessitating a heart transplant. Heart failure resulting from myocarditis-induced dilated cardiomyopathy is a significant cause of disability and death in children.



### DEFINITION: What is Pediatric Myocarditis?

Myocarditis is inflammation of the heart muscle. When myocarditis affects patients younger than 18 years of age, it is called pediatric myocarditis.

In newborns the disease can be especially severe because of an underdeveloped immune system. Myocarditis can present in young athletes as sudden death. The incidence of dilated cardiomyopathy in children ranges from 0.57 to 0.76 cases per 100,000 children. Myocarditis is associated with dilated cardiomyopathy in 27% to 46% of cases. Dilated cardiomyopathy is the primary indication for childhood heart transplantation. Therefore, heart failure resulting from myocarditis-induced dilated cardiomyopathy is a significant cause of disability and death in children as well as adults.

### ETIOLOGY: What Causes Pediatric Myocarditis

Myocarditis may be caused by infections in the heart, autoimmune disorders, hypersensitivity reactions to drugs and toxin exposure. Viruses are the most common cause of myocarditis in children. Less common infectious agents include bacteria and protozoa. Coxsackie B was the first virus associated with myocarditis. Many other viruses have also been implicated in children including Adenovirus, Parvovirus B19, hepatitis B and C viruses and HIV. The heart can be damaged by the virus itself or by the body's immune defense against the virus. Drugs including some antibiotics, antipsychotics and anticonvulsants may rarely cause hypersensitivity reactions resulting in inflammation of the heart. Scorpion bites have been associated with myocarditis primarily in India. Autoimmune disorders associated with myocarditis include: systemic lupus erythematosus, celiac disease, and sarcoidosis. Finally, Giant Cell Myocarditis is a rare but serious cause of acute dilated cardiomyopathy and heart failure carrying a high risk of death unless heart transplantation is performed.



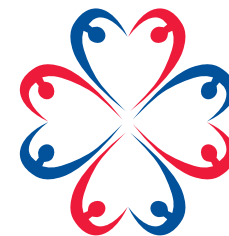
### SIGNS AND SYMPTOMS: When to Suspect Pediatric Myocarditis

Clinical manifestations of acute viral myocarditis are usually nonspecific and highly variable. They range from mild flu-like symptoms to sudden death. Many infants and children with myocarditis present with fulminant features such as rapid and labored breathing, wheezing, grunting, low blood pressure, cool extremities and decreased urine output. Older children may complain of fatigue, fever, vomiting and muscle aches a few days before developing more severe symptoms such as shortness of breath and exercise intolerance. More advanced symptoms include rapid heart rate, erratic and weakened pulse, pale color, sweating and dizziness.

### DIAGNOSIS: How is Pediatric Myocarditis Detected?

A physical examination may reveal signs of respiratory distress or decreased cardiac function. Clinical findings may include: rapid respiratory or heart rate, retracting chest wall respiratory muscles, nasal flaring, distended neck veins, weakened pulse, irregular heart rhythm, cool extremities, enlarged liver, low blood pressure. Some patients may develop a change in their mental status, becoming confused, disoriented or non-interactive. Blood tests for cardiac injury may help establish a diagnosis of myocarditis. Blood levels of troponin I and B-type natriuretic peptide (BNP or NT-pro BNP) are frequently elevated. A chest x-ray may reveal lung congestion or an enlarged heart. The electrocardiogram (ECG) is abnormal in about half of cases and may show decreased electrical voltages or an irregular heart rhythm. An echocardiogram (Echo) is usually nonspecific but may reveal decreased left and/or right ventricular pump function. Magnetic resonance imaging (MRI) may detect inflammation of the heart muscle and give additional information about the size and function of the heart.

Once a diagnosis of myocarditis is suspected and other common causes of cardiomyopathy have been excluded, a cardiologist may perform a cardiac catheterization to retrieve tissue samples from the inner lining of the heart. This is called an endomyocardial biopsy. The biopsy is the gold standard for diagnosis of myocarditis but its use is limited by risks associated with the procedure. Tissue obtained by this procedure may also yield clues about the identity of the causative agent. The Pediatric Cardiomyopathy Registry, a study funded by the National Heart, Lung and Blood Institute, has reported that only one-third of children have a defined cause of their cardiomyopathy.



### THERAPY: What are the Treatment Options for Myocarditis?

The primary treatment of myocarditis is supportive care based on guidelines and recommendations published by major cardiovascular organizations in North America and Europe. There are specific guidelines for the management of heart failure in children and standards of care for heart transplantation and pediatric cardiomyopathies. Children with a new diagnosis of myocarditis usually require hospitalization for treatment of heart failure and arrhythmias. Administration of intravenous cardiac medications or insertion of a temporary pacemaker may be necessary. In severe cases, extracorporeal membrane oxygenation (ECMO) or a ventricular assist device (VAD) may be necessary in the acute phase to allow the heart to recover or to serve as a bridge to transplantation. Immunoglobulin (IVIG) or corticosteroids have been used in some acute cases to inhibit the immune response. Following the acute phase, surviving patients may recover completely or have long-term deficits in cardiac function. Some patients may develop a slow and progressive course of dilated cardiomyopathy with diminishing cardiac function and heart failure symptoms. In severe cases cardiac transplantation may offer the best chance for long-term survival.

### EXPECTED OUTCOMES: What is the Prognosis for Myocarditis Patients?

Although many children recover from myocarditis with no serious consequences, severe forms of myocarditis are associated with significant morbidity and mortality worldwide. Children with fulminant myocarditis are more likely to achieve a complete recovery if they survive the acute phase. Children who develop cardiomyopathy and progress to end-stage heart failure, despite optimal medical therapy, may be candidates for cardiac transplantation.

Transplant-free survival rates are highly variable, ranging from 40%-80% over 5 years. A recent study demonstrated improved 1 and 5 year survival rates of 90% and 83%, respectively. Predictors of outcome in children with dilated cardiomyopathy include severity of symptoms at presentation, the presence or absence of arrhythmias and the rate of clinical improvement. After therapy, post-transplantation survival rates are 80% at 1 year and 70% at 5 years.