Restrictive cardiomyopathy (RCM) is less common affecting 3 to 5 percent of children with cardiomyopathy. With RCM, the walls of the lower chambers of the heart (ventricles) are abnormally stiff but not necessarily thickened or enlarged. The heart’s rigid walls fail to relax and expand adequately, “restricting” the ability of the heart to fill with blood.

While the heart pumps normally, it is still unable to supply enough blood to the body. This puts pressure on the upper chambers of the heart (atria), and they become enlarged and out of proportion to the size of the lower chambers of the heart (ventricles). In advanced stages of the disease, the heart may not pump blood efficiently and blood may back up into the liver and lungs as a result of congestive heart failure.

**What is restrictive cardiomyopathy?**

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**How many children are affected?**

According to the Pediatric Cardiomyopathy Registry, RCM occurs at a rate of less than 1 per million children.

**Diagnosis**

Children’s Cardiomyopathy Foundation

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**What is the prognosis?**

RCM is a rare disease and there is limited information on the disease in children. Long-term survival increases for children who receive heart transplants. Irreversible and severe pulmonary hypertension has been the only risk factor associated with poor outcome for children with RCM.

The Children’s Cardiomyopathy Foundation (CCF) is a national non-profit organization dedicated to finding causes and cures for pediatric cardiomyopathy through the support of research, education, and increased awareness and advocacy.

Register with CCF:

childrenscardiomyopathy.org

**Understanding Restrictive Cardiomyopathy**

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This includes endomyocardial fibrosis, which may also form in the enlarged areas of the heart and travel to the brain or other parts of the body. As RCM advances, fluid builds up in the lungs as well as in the veins that carry blood back toward the heart. This may cause neck veins to bulge; an enlarged liver; and fluid in the abdomen, face and legs.

Some children may develop abnormal heart rhythms (arrhythmia) where the heart beats too slow (bradycardia) or too fast (tachycardia). With some forms of RCM, a slow heart rate may develop from heart block. This is due to the abnormal conduction of signals to the heart's pumping chambers. Pulmonary hypertension and blood clots are common complications of RCM. Pulmonary hypertension, defined as high blood pressure in the lungs, may occur when the arteries in the lungs are restricted and the heart must work harder to pump blood through the lungs. Blood clots may also form in the enlarged areas of the heart and travel to the brain or other parts of the body.

Other tests may be ordered to access the heart's condition and determine a treatment plan. These tests include a chest x-ray to check the heart's shape and size and to look for fluid in the lungs, computed tomography (CT scan) to observe the structure and function of the heart and blood vessels, magnetic resonance imaging (MRI) to evaluate heart and blood vessel function, and a Holter monitor to look for abnormal heartbeats. In order to get a more precise RCM diagnosis, more invasive tests may be necessary. A cardiac catheterization is used to measure heart and lung pressures, which are usually elevated with RCM. A heart (endomyocardial) biopsy, which involves removing a small piece of heart muscle for microscopic examination, is usually performed at the same time. The laboratory checks for infiltrating substances or abnormal deposits in the heart. It also looks for any indication of constrictive pericarditis, which resembles RCM in its signs and symptoms.

What are the common symptoms?
The onset of symptoms in RCM is often very subtle. Symptoms arise from the decreased filling of the heart and insufficient blood flow to the body. Infants and young children may show irritability, poor appetite and slow weight gain. Older children may experience fatigue, difficulty exercising or breathing (dyspnea), a persistent cough or wheezing, fainting (syncope), abnormal heart beat (palpitations), chest pain (angina) and an upset stomach. As RCM advances, fluid builds up in the lungs as well as in the veins that carry blood back toward the heart. This may cause neck veins to bulge; an enlarged liver; and fluid in the abdomen, face and legs.

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What is restrictive cardiomyopathy diagnosed?
In the early stages, RCM may be difficult to detect in a physical examination because of the absence of symptoms. Therefore, cardiologists rely on noninvasive cardiac tests such as the echocardiogram and the electrocardiogram (EGK) to diagnose the condition. An echocardiogram measures the size of the heart, how well the heart pumps and the severity of pulmonary hypertension if an issue. An ejection fraction can be calculated by measuring the percentage of blood ejected from the heart with each beat. Unlike other forms of cardiomyopathy, children with RCM typically have normal ejection fractions of 50 to 70 percent because the heart's pumping function is unaffected until the later stages of the disease. An EKG provides information on the heart's electrical activity and whether there is heart block or irregular heart rhythms. This test nearly always shows abnormal patterns associated with the enlargement of the heart's upper chamber (atria).

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