D[illegible] RCM

Restrictive cardiomyopathy is among the rarest of childhood cardiomyopathies. Its diagnosis is difficult to establish early in the clinical course due to the lack of symptoms. Therefore, in many cases, this diagnosis is made only after presentation with symptoms such as decreased exercise tolerance, new heart sound (gallop), syncope (passing out) or chest pain with exercise.

Once suspected, there are certain tests that can help confirm this diagnosis. An electrocardiogram, or EKG, which records the electrical conduction through the heart, can be very helpful. This can show abnormally large electrical forces from enlargement of the atria (upper chambers) of the heart. An echocardiogram, or ultrasound of the heart, can provide additional clues to help make this diagnosis. Generally, in children with RCM, the echocardiogram shows marked enlargement of the atria (upper chambers), normal sized ventricles (lower chambers) and normal heart function. In more advanced disease states, pulmonary artery pressure (blood pressure in the lungs) will be increased and can often be estimated during the echocardiogram.

Cardiac catheterization is usually the next procedure done to confirm the diagnosis. During this procedure, a catheter (thin plastic tube) will be slowly advanced through an artery or vein into the heart (while watching its course on a TV monitor) so that pressures within the heart chambers can be measured. These measurements often show significantly elevated pressures during the relaxation period of the heart (when it fills with blood before the next beat) and varying degrees of increased pulmonary artery pressure (which can confirm the echo estimates) in the absence of any other structural heart disease. In very rare cases, based on clinical symptoms and prior laboratory evaluation, a cardiac biopsy may be performed. This involves removing tiny pieces of heart muscle for inspection under the microscope to search for potential causes of this condition (such as amyloidosis or sarcoidosis, which are common causes of RCM in adults but rarely in pediatric patients).

Finally, since childhood RCM is often genetic and in many cases will be inherited, once this diagnosis is established, your doctor will likely request that parents, siblings of the patient and sometimes other close relatives be screened with an echocardiogram to rule out the presence of this disease in other family members.

C[illegible] RCM

Although the cause of RCM is not known in most pediatric cases, there is some scientific evidence suggesting that individual genetic “mutations” may be a cause in some cases of RCM in children. For a greater understanding of the basics of human inheritance patterns and a more detailed discussion of the potential genetic causes of RCM, the reader is encouraged to read separate sections entitled “Overview of Inheritance” and “Genetics of Cardiomyopathies” printed elsewhere in this brochure.

C[illegible] T[illegible]

Currently, there are no therapies that can “cure” RCM; however, some treatments are available that can improve symptoms in children with RCM. The choice of a specific therapy depends on the clinical condition of the child, the risk of dangerous events and the ability of the child to tolerate the therapy.

M[illegible] T[illegible] T[illegible] RCM [illegible] A[illegible] H[illegible] F[illegible]

Some children with RCM have signs and symptoms of heart failure due to the abnormal relaxation properties of the heart muscle. The most common types of medications used to treat heart failure under these circumstances include diuretics, beta-blockers and occasionally afterload reducing agents.

Diuretics, sometimes called “water pills,” reduce excess fluid in the lungs or other organs by increasing urine production. Diuretics can be given either orally or intravenously. Common diuretics include furosemide, spironolactone, bumetanide and metolazone. Common side effects of diuretics include dehydration and abnormalities in the blood chemistries (particularly potassium loss). In patients with RCM, diuretics must be used very carefully and given only in doses to treat extra lung and abdominal fluid without inducing excessive fluid loss as this may cause symptomatically low blood pressure.

Beta-blockers slow the heartbeat and increase the relaxation time of the heart. This may allow the heart to fill better with blood before each heart beat and decrease some of the symptoms created by the stiff pumping chambers. Common beta-blockers (taken by mouth) include carvedilol, metoprolol, propanolol and atenolol. Side effects include dizziness, low heart rate, low blood pressure, and, in some cases, fluid retention, fatigue, impaired school performance and depression.

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S

The intellectual, psychological and social bene□ts of attending school cannot be overestimated in the child with RCM. Adjusting medication schedules so they do not interfere with school activities, discussing safe activity levels with school personnel, and providing tutoring to maintain academic performance are important interventions that can help a child to stay in school and keep up with their peers. Often close communication between the parents, medical care team, and the school nurse can help to keep a child up to date in school.

F

Every effort should be made to allow a child with RCM to spend time with friends. The child should also be allowed to participate in recreational activities whenever possible. However, an effort should be made to avoid contact with those who are acutely ill with fever, even though many children with RCM are able to tolerate upper respiratory tract illnesses (common colds) well.

P

Adjusting to having a chronic illness is stressful for the child and the family. The child's reaction to having RCM often depends on the stage of the child's development. Discussions about the disease should be tailored to the speci□c concerns of the child. Child-life professionals and pediatric psychiatrists are important resources to help children cope, and their services are often available thro spe(i)5(n)5(g)5hil ro5(i)5(s)5()5(i)5(c)5(a)5(n)5(v)5(e)l.

Dr[illegible] F[illegible] H[illegible] RCM?