Protocol: Proactive Cardiac Care in LMNA-CMD

Cardiac findings in congenital LMNA related dystrophy (L-CMD)

Short runs of atrial tachycardia may begin in the first decade of life and become more persistent over time, resulting in chronic atrial fibrillation. Anticoagulation may be needed.

Progressive AV block may also begin early, and get worse over time. Some patients require pacemakers.

Asymptomatic non-sustained ventricular tachycardia (VT) may be seen on Holter. Sudden death, while rare, has been reported, even with a prior normal ECHO and Holter.

Ventricular systolic dysfunction tends to be a late finding, and may deteriorate rapidly. Diarrhea, edema may be early symptoms of cardiac dysfunction before a noticeable decline in ejection fraction on ECHO. BNP levels may be a more sensitive indicator of heart dysfunction than ejection fraction.

Physiologic stress (surgery, infection) may precipitate a rapid decline in heart function.

Recommendations

Baseline EKG and ECHO should be performed as soon as diagnosis is suspected or confirmed, and at least yearly thereafter, even when asymptomatic. Pay careful attention to right heart function and diastolic function on ECHO.

Check yearly BNP levels after significant arrhythmias develop.

Periodic Holter monitoring should begin as early as age 5, even in asymptomatic patients. Implantation of a loop recorder is an option for extended rhythm surveillance.

Consider ICD if there is sustained or non-sustained VT on Holter/Loop, especially if left ventricular ejection fraction <45% or if a pacemaker is needed for heart block.

Aggressive heart failure management should be initiated early, using diuretics, β-blockers, aldosterone antagonists, and angiotensin-converting enzyme inhibitors, although it is unclear whether this alters prognosis.

Steroids may be helpful for GI symptoms (especially if there is protein losing enteropathy). It is unknown if steroids have other cardiac benefits.

Optimize pulmonary management and treat night time hypoventilation which can impact the heart.

Involve palliative care team throughout child's course, to optimize quality of life and assist family with care planning.

Excerpted from: Heller, et al. Cardiac manifestations of congenital LMNA-related muscular dystrophy in children: three case reports and recommendations for care. Cardiology in the Young, Dec 2016

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