Early Treatment Improves Cardiac Function in MD

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NEW ORLEANS — Treatment of newly diagnosed dilated cardiomyopathy in children with muscular dystrophy can improve the heart's size and function in many cases, John L. Jefferies, M.D., said in a poster at the annual scientific sessions of the American Heart Association.

"The goal is to preserve normal cardiac function. If we catch a patient at the start of ventricular dysfunction, we can start treatment before the ejection fraction is substantially depressed" and symptoms from ventricular dysfunction appear, said Dr. Jefferies, of Texas Children's Hospital, Houston.

Dilated cardiomyopathy, common in muscular dystrophy, responds to therapy. Thus, annual screening by echocardiography should start when a child with muscular dystrophy is about 5 years old, Dr. Jefferies told CLINICAL NEUROLOGY NEWS.

The study involved 69 boys with muscular dystrophy who were referred to the Cardiovascular Genetics Clinic of the Texas Heart Center. Seven had Becker's muscular dystrophy (BMD) and were first seen at an average age of 12.9 years; 62 boys had Duchenne's muscular dystrophy (DMD) and were first examined at an average age of 13.7 years. These boys underwent annual echocardiography, and dilated cardiomyopathy was eventually diagnosed in 4 of the patients with BMD (57%) at an average age of 14 years, and in 27 of those with DMD (44%) at an average age of 16 years. All 31 patients were started on drug therapy. At the time of initial diagnosis, the 31 patients with dilated cardiomyopathy had an average left ventricular ejection fraction (LVEF) of 36% and an average myocardial performance index of 0.53.

Either liquid enalapril or captopril was given to younger children, with a tablet formulation of lisinopril an option for adolescents. If a patient's LVEF failed to improve after 3 months with this treatment, the physicians added a β-blocker, either metoprolol or carvedilol, which both come in liquid formulations

Clinical follow-up was possible for 29 of the 31 treated patients. During an average follow-up of 3.3 years, 19 patients (3 with BMD and 16 with DMD) had complete normalization of their cardiac size and function. Another eight patients, all with DMD, had improvements in cardiac size and function, and three patients, all with DMD, had stabilization of their cardiac size and function, Dr. Jefferies reported. After an average of 3.3 years of treatment, the average LVEF in these 29 patients had improved to 53%, and the average myocardial performance index had improved to 0.38. (Less than 0.41 is considered normal.) ■

Screens Fail to Find Diabetic Neuropathy

QUEBEC CITY — Noninvasive methods for identifying pediatric diabetic neuropathy are not as sensitive as conventional nerve conduction studies and should not be considered for screening purposes, Daniele Pacaud, M.D., said at the joint annual meeting of the Canadian Diabetes Association and the Canadian Society of Endocrinology and Metabolism.

Her study compared vibration perception thresholds (VPT) and tactile perception thresholds (TPT) with nerve conduction studies (NCS) in 73 children, mean age 13 years, with type 1 diabetes.

For VPT testing, subjects were asked to touch a box with their big toe and to indicate if it is vibrating. The amplitude of the vibrations is steadily decreased until they become imperceptible.

For TPT, subjects were asked to indicate when they feel microfilaments that are applied to the plantar surface of the foot.

All children in the study completed a neurologic questionnaire, underwent NCS, and received a neurologic exam. Based on two abnormalities on NCS, diabetic neuropathy was present in 42 (57%) of the 73 subjects.

Of these, 37 were picked up by VPT, 26 by neurologic exam alone, and 19 by TPT, said Dr. Pacaud, of Alberta Children's Hospital and the University of Calgary in Calgary (Alta.). The neurologic symptoms questionnaire was not useful, she added.

—Kate Johnson