PEDIATRIC NEUROLOGY BRIEFS
A MONTHLY JOURNAL REVIEW

J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

Vol. 20, No. 7 July 2006

MUSCLE DISORDERS

CARDIAC COMPLICATIONS OF FUKUYAMA-TYPE CONGENITAL MUSCULAR DYSTROPHY

The course of left ventricular function was evaluated using M-mode and Doppler echocardiography in 34 patients with Fukuyama-type congenital muscular dystrophy (FCMD), in a study at the Tokyo Women's Medical University, Tokyo, Japan. Ages ranged from 6 months to 30 years (median, 6 years). Diagnosis of FCMD was based on haplotype analysis, neurologic examination and muscle biopsy. The haplotype was ancestral homozygous in 21 and heterozygous in 10 patients. The interval between echocardiographic examinations ranged from 12 months to 11 years (median, 4 years). ECG abnormalities were present in 20 (59%) patients: a tall R wave in 20, and a deep but narrow Q wave over lead V6 in 2 patients. The PG interval was normal. Decreased left ventricular systolic function (expressed as left ventricular fractional shortening [LVFS]) was present in 16 (47%) patients. The finding of LVFS was significantly correlated with age; LVFS was normal in patients <10 years and reduced in patients >15 years of age. LVFS in 16 patients with ancestral heterozygotes was not significantly different from that in 9 patients with homozygotes. LVFS was not significantly different in 3 groups with mild, moderate, and severe skeletal muscle involvement. All 5 patients who died during follow-up had decreased LVFS. They had not received heart-failure therapy. Three patients died of respiratory failure at 2, 27, and 31 years of age. Two patients died of heart failure at 16 and 17 years of age. Autopsy findings showed multifocal fibrosis throughout the LV wall, especially severe in those with heart failure. LVFS was increased significantly in 5 patients who received ACE inhibitors. (Nakanishi T, Sakauchi M, Kaneda Y, et al. Cardiac involvement in Fukuyama-type congenital muscular dystrophy. **Pediatrics** June 2006;117:1187-1192). (Respond: Toshio Nakanishi MD, Department of Pediatric Cardiology, Heart Institute of Japan, Tokyo Women's Medical University, 8-1 Kawada-cho, Shinjuku, Tokyo, 162-8666, Japan).

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COMMENT. Fukuyama-type congenital muscular dystrophy is frequently complicated by cardiac involvement after 10 years of age. Treatment with ACE inhibitors and, if needed, beta-blockers, when patients develop early signs of left ventricular dysfunction, may delay the onset of heart failure. Routine cardiac evaluation, including echocardiograms and follow-up, is recommended, especially in older children.

Treatment of the heart in Duchenne muscular dystrophy (DMD) is discussed in an editorial (Baxter P. **Dev Med Child Neurol** March 2006;48:163-163). Multicenter trials are recommended, since reports of treatment with ACE inhibitors and steroids are variable. Cardiomyopathy develops in most patients with DMD by age 18 years, and is the leading cause of death in 10-40%. Initiating therapy at the pre-symptomatic stage of LV dysfunction may delay the onset of cardiac failure and improve prognosis (Bourke JP. Cardiac monitoring and treatment for children and adolescents with neuromuscular disorders. **Dev Med Child Neurol** March 2006;48:164-164).

DIAGNOSIS AND TREATMENT OF DERMATOMYOSITIS

The impact of duration of untreated symptoms in children with juvenile dermatomyositis (JDM) on clinical and laboratory findings at diagnosis was studied in 166 patients enrolled in the National JDM Research Registry and examined at Children's Memorial Hospital, Northwestern University; Loyola University; and University of Chicago, Chicago, IL; and other centers in the US. The girl:boy ratio was 2:1; 74% white, 13% Hispanic, and 10% African-American. Duration of untreated disease ranged from .07 month to 98 months (median, 4.04 months). Age at diagnosis and duration of disease showed no significant differences with gender or race. Rash (heliotrope eyelids, malar erythema, Gottron's metacarpal papules) was the first symptom reported by 100 patients (65%), and muscle weakness was the first in 44 (29%). Nine (6%) had both rash and weakness at disease onset. The severity of muscle symptoms based on disease activity scores (DAS 0-11) was greater in nonwhite (6.77) cf white (4.71) children (P>.0005). Mean DAS total (skin rash and muscle weakness) was higher in nonwhite (12.52) cf white (10.45) children (P=.001). DAS skin rash (scale 0-9) ranged from 3-9 (mean, 6) and did not vary with duration of untreated disease, whereas DAS muscle weakness (scale 0-11) ranged from 0-11 and declined gradually with duration of untreated disease (P>.0005). More children with untreated JDM were in lower percentiles for height and weight than their healthy controls, irrespective of the duration of untreated disease. In addition to rash and muscle weakness, symptoms at diagnosis of JDM in order of frequency included capillary dilation and telangiectasia, followed by arthritis, dysphagia, and abdominal pain. Dysphagia and arthritis were more common in older children, whereas cutaneous calcifications were correlated with duration of untreated disease (P=.006). Levels of all 4 routine muscle enzymes (CK, aldolase, LDH, and SGOT/AST) were lower with longer duration of untreated disease; they were normal in 13-26% of children at the initial clinic visit. CK level was normal in 33 untreated JDM cases, despite muscle weakness. (Pachman LM, Abbott K, Sinacore JM, et al. Duration of illness is an important variable for untreated children with juvenile dermatomyositis. **J Pediatr** Feb 2006;148:247-253). (Reprints: Dr Lauren M Pachman, Children's Memorial Medical Center, Division of Immunology/Rheumatology, 2300 Children's Plaza, Box 50, Chicago, IL 60614).