

cytoplasmic filamentous inclusions. CT white matter hypodensities and MRI high signal intensities were compatible with leukodystrophy, yet the patients had no symptoms of white matter dysfunction. The authors conclude that this constellation of familial myopathy with muscle cytoplasmic inclusions and cerebral white matter changes represents a hitherto undescribed syndrome. (Cole AJ et al. Familial myopathy with changes resembling inclusion body myositis and periventricular leucoencephalopathy. Brain Oct 1988;111:1025-1037).

COMMENT. Familial cases of inclusion body myositis have been reported in younger patients but none associated with cerebral white matter changes as described above. In contrast to those myopathies sometimes associated with CNS dysfunction (eg. Duchenne muscular dystrophy, myotonic dystrophy, congenital muscular dystrophy of Fukuyama and others, and Kearns-Sayre-Shy syndrome) the white matter changes in the present cases were asymptomatic. The mode of inheritance could not be determined with certainty. Limb girdle dystrophy presents in yet another form and should prompt examination not only of blood platelets but also CT and MRI for white matter changes. CT findings were not included in the report from Manchester and blood platelet counts were not indicated in the Montreal syndrome.

CONGENITAL MYOPATHY, CLEFT PALATE, AND MALIGNANT HYPERTHERMIA

Six children with congenital ptosis, generalized weakness, hypotonia, cleft palate, and susceptibility to malignant hyperthermia with anesthesia, are reported in Lumbee Indians from Duke University Medical Center, Durham, North Carolina. All patients were members of the same ethnic group, 3 were related, and inheritance was probably autosomal recessive. Surgery for cleft palate at 14 months and for ptosis at 27 months, using halothane anesthesia, was complicated by malignant hyperthermia in one child. This syndrome showed some resemblance to King syndrome, characterized by multiple congenital facial and skeletal abnormalities along with slowly progressive myopathy and susceptibility to malignant hyperthermia. (Stewart CR et al. Congenital myopathy with cleft palate and increased susceptibility to malignant hyperthermia: King syndrome? Pediatr Neurol Nov/Dec 1988;4:371-4).

COMMENT. Clinicians, especially surgeons and anesthesiologists, should be aware of the risk of malignant hyperthermia in children with this syndrome and other myopathies, including Duchenne muscular dystrophy, myotonia congenita, and central core disease. Malignant hyperthermia is manifested by muscle rigidity, rapid elevation of temperature, metabolic acidosis, and rhabdomyolysis. Anesthetic agents most frequently invoked are halothane and succinylcholine. Screening of susceptible patients and their families by CK determinations is advised.

An index for PEDIATRIC NEUROLOGY BRIEFS, Vols 1 and 2, 1987-88 is in preparation.