spinal cord dysfunction or atrophy; none had macrocephaly. (van der Knaap MS, Ramesh V, Schiffmann R, et al. Alexander disease. Ventricular garlands and abnormalities of the medulla and spinal cord. **Neurology** February (2 of 2) 2006;66:494-498). (Reprints: Dr Marje van der Knaap, Department of Child Neurology, VU University Medical Center, De Boelelaan 1117, 1081 HV Amsterdam, The Netherlands).

COMMENT. Alexander disease, a rare leukoencephalopathy caused by mutations in the *GFAP* gene, is characterized by white matter abnormalities, predominantly affecting the frontal lobes. The disease typically occurs in infants with macrocephaly and delayed development. Since DNA confirmation of the diagnosis, the clinical and MRI phenotype has widened to include cases with predominant brainstem and spinal cord involvement, usually of late onset. Signal abnormalities or atrophy of these locations should warrant DNA analysis for Alexander disease. An MRI appearance of ventricular garlands, a relatively new sign, is also an indication for DNA confirmation.

MUSCLE DISORDERS

BRAIN MRI FINDINGS IN CONGENITAL MUSCULAR DYSTROPHY

Brain magnetic resonance imaging (MRI) findings in 13 patients with congenital muscular dystrophy (MDCIC) and Fukutin-related protein (*FKRP*) gene mutations were retrospectively reviewed in a study at Hammersmith Hospital, London, UK, and European centers. MRIs were obtained between 22 months and 11 years of age. Five patients had normal MRIs and normal cognitive abilities. Three had cerebellar cysts and cognitive impairment (IQ 50, 56, and 70); 1 was microcephalic. Five had cerebellar cysts associated with other structural abnormalities, including nodular heterotopia, frontal pachygyria, pontine hypoplasia, microcephaly, lissencephaly, Dandy-Walkerlike malformation, and absence of cerebellar vermis. One patient had Walker-Warburg syndrome (WWS) with involvement of muscle, eye and brain. The severity of CNS involvement reflected the severity of disruption of a-dystroglycan glycosylation (DGG). DGG was almost absent in the patient with WWS and less severely reduced in patients with MDCIC with or without cerebellar cysts. (Mercuri E, Topaloglu H, Brockington M, et al. Spectrum of brain changes in patients with congenital muscular dystrophy and *FKRP* gene mutations. **Arch Neurol** Feb 2006;63:251-257). (Respond: Francesco Muntoni MD, Dubowitz Neuromuscular Centre, Department of Paediatrics, Imperial College, Hammersmith Hospital Campus, Du Cane Road, London WI2 OHN, UK).

COMMENT. Only 5 of 13 patients with congenital muscular dystrophy and *FKRP* gene mutations had normal brain MRIs and normal IQs. Eight patients had structural brain anomalies consisting of cerebellar cysts with or without other structural changes and cognitive deficits of various degrees of severity. Cerebellar cysts, either alone or associated with vermis hypoplasia and white matter abnormalities, were the most common MRI findings with *FKRP* mutations and MDCIC. The degree of abnormal glycosylation of a-dystroglycan correlates with disease severity, MDCIC having a more severe depletion compared with later onset limb girdle MD. Congenital MDs are called dystroglycanopathies and include Fukuyama CMD, muscle-eye-brain disease, and Walker-Warburg syndrome.