

and upper and lower extremities. Funduscopic examination revealed cherry red spot of the retina, consistent with a diagnosis of the lysosomal disease, GM1 gangliosidosis type 1. (Hackbart BA, Arita JH, Pinho RS, Masruha MR, Vilanova LCP. Mongolian spots are not always a benign sign. **J Pediatr** 2013 May;162(5):1070). (Response: Dr Barbara A Hackbart, Division of Child Neurology, Federal University of Sao Paulo, Brazil).

COMMENT. The Mongolian spots result from entrapment of melanocytes in the dermis because of arrested transdermal migration from the neural crest into the epidermis. Hurler syndrome and GM1 gangliosidosis type 1 are diseases associated with generalized Mongolian spots. Infants with GM1 gangliosidosis type 1, also known as Pseudo-Hurler's disease, show facial abnormalities that include frontal bossing, depressed nasal bridge, macroglossia, large low-set ears, and marked hirsutism. About 50% have cherry-red spots. The association of Mongolian spots with the lysosomal disease GM1 gangliosidosis type 1 was not recorded in older neurology textbooks, but a PubMed search found 10 references in the last 30 years (Weissbluth M, et al. **Br J Dermatol** 1981 Feb;104(2):195-200) (Ashrafi MR, et al. **Pediatr Neurol** 2006 Feb;34(2):143-5). Mongolian spots when unusually numerous should prompt an examination for the lysosomal disease, GM1 gangliosidosis type 1.

## DEMYELINATING DISEASES

### **DIAGNOSTIC CRITERIA FOR PEDIATRIC MS**

Investigators at Northwestern University Feinberg School of Medicine and Ann & Robert H. Lurie Children's Hospital of Chicago review the diagnostic criteria for pediatric multiple sclerosis, the differential diagnosis, the 2010 McDonald criteria, and Callen criteria. Of all persons with MS, 2% to 5% have onset before 16 years of age. The diagnosis is clinical, requiring recurrent episodes of CNS demyelination, serial changes in MRI lesions, and CSF oligoclonal bands or elevated IgG index. MS must be differentiated from ADEM, neuromyelitis optica and other inflammatory, infectious or metabolic conditions. These include mitochondrial disorders, leukodystrophy, Alexander's disease, MELAS, Kearns-Sayre syndrome, Behcet and Sjogren syndromes, sarcoidosis, Hashimoto's encephalitis, HIV, herpes virus, neuroborreliosis, mycoplasma, the arteriopathy CADASIL, and CNS vasculitis. (Rubin JP, Kuntz NL. Diagnostic criteria for pediatric multiple sclerosis. **Curr Neurol Neurosci Rep** [Section Editor, Nordli DR Jr] 2013 Jun;13(6):354).

COMMENT. In this excellent and comprehensive review, the differentiation of MS from ADEM and other inflammatory or infectious conditions is stressed. Transient demyelinating events must be distinguished from a life-long diagnosis of MS.

**Cerebral venous thrombosis (CVT) after LP and steroids in childhood MS.** The association between CVT and MS is reported in a 13-year-old girl admitted with left hemiparesis, ataxia, and headache following vaccination against meningococcal group C and hepatitis A. LP and high dose corticosteroids for MS may have contributed to the CVT (Presicci A, et al. **Brain Dev** 2013 Jun;35(6):602-5).