brachioradialis deep tendon reflexes, increased left patellar reflex, bilateral increase of Achilles tendon reflexes, left spontaneous Babinski, and shorter distance between the knee and ankle cutaneous creases on the left compared to the right leg.

Case 2, a 12-month-old boy with right brachial plexus palsy presented for evaluation of ipsilateral leg weakness, first noted by the mother when the infant attempted to walk. Neurological examination uncovered a tight right heel cord. Brain MRI revealed diffuse cortical dysplasia of the left hemisphere. (Alfonso I, Alfonso DT, Price AE, Grossman JAI. Cortical dysplasia and obstetrical brachial plexus palsy. **J Child Neurol** Dec 2008;23:1477-1480). (Respond: Israel Alfonso MD, Department of Neurology, Miami Children's Hospital, 3200 SW 60 Court, Suite 302, Miami, Fl 33155. E-mail: ialfonso@pediatricneuro.com).

COMMENT. The authors found no previous reports of an association of brachial plexus palsy and cortical dysplasia. They propose that this association helps explain the pathophysiology of brachial palsy in these patients by 2 mechanisms: prenatal shoulder girdle weakness and an abnormal arm position that increase the vulnerability of the plexus to stretch injury during delivery. Case 1 emphasizes the importance of attention to the length of the lower limbs and asymmetry in a neonatal neurological examination. MRI examination to exclude associated brain pathology should be considered in neonates with severe or complicated brachial plexus palsy.

Small Focal Cortical Dysplasia (FCD) lesions overlooked by routine MRI are visualized by high-resolution MRI, in a study at Montreal Neurological Institute, Canada (Besson P et al. **Brain** Dec 2008;131:3246-3255). Of 21 patients with small FCD, 17 (81%) were not identified initially, and 18 (86%) were located at the bottom of a sulcus. The knowledge that small FCD lesions are preferentially located at the bottom of an abnormally deep sulcus should aid the search for developmental cerebral lesions by routine MRI.

Outcome of Obstetric Brachial Plexus Injury correlates with force of downward traction of the fetal head in a study of 98 affected children at Goteborg University, Sweden (Mollberg M et al. **J Child Neurol** 2008;23:1424-1432). At 18 months follow-up, 82% had recovered completely and 18% had persistent functional neurological deficits.

DEMYELINATING DISEASES

RELAPSE RATE IN PEDIATRIC-ONSET MULTIPLE SCLEROSIS

Relapse rates were compared during 12 months or longer follow-up between 21 pediatric onset cases of multiple sclerosis (MS) seen at the Massachusetts General and 110 patients with adult-onset MS at the Brigham and Women's Hospitals, Boston, MA. Pediatric-onset patients had a 1.13 annualized relapse rate that was significantly higher than that in the adult-onset group (0.40) ($P<0.001$). The adjusted rate ratio was 2.81. The increased relapse rate in pediatric-onset MS remained highly significant when controlled for disease-modifying treatment time, and when age at onset was treated as a continuous variable. Pediatric-onset MS has a more inflammatory disease course than adult onset MS. (Gorman MP, Healy BC, Polgar-Turcsanyi M, Chitnis T. Increased relapse rate in pediatric-onset compared with adult-onset multiple sclerosis. **Arch Neurol** January 2009;66:54-59). (Respond: Tanuja

Chitnis MD, Partners Pediatric Multiple Sclerosis Center, Massachusetts General Hospital, ACC-708, 55 Fruit St, Boston, MA 02114. E-mail: tchitnis@partners.org).

COMMENT. Pediatric-onset MS has a slower rate of progression than adult-onset disease, according to several reports. The discrepancy between higher relapse rate and slower long-term progression of pediatric-onset MS is unexplained.

FAMILIAL HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS TYPE 3 WITH DEMYELINATING CNS DISEASE

A case of familial hemophagocytic lymphohistiocytosis type 3 (FHLH3) presenting in a 3-year-old boy with fulminant demyelinating neurological disease is reported by researchers at Kravis Children's Hospital at Mount Sinai Medical Center, New York. Clinical examination 4 days after onset of neurological symptoms revealed an alert, active child with massive splenomegaly, and broad-based gait. MRI showed diffuse small demyelinating foci in subcortical and perivascular white matter. CSF protein was 55 mg/dL, and myelin basic protein was elevated (7.2 ng/mL, normal 0-4). EEG showed bilateral slowing. He had presented 6 weeks earlier at another hospital with irritability, abdominal pain, head tilt, neck and back pain, and inability to walk. Laboratory tests showed anemia, neutropenia and thrombocytopenia; serum ferritin was normal. Having recently returned from a trip to Honduras, he had received a course of amphotericin B for clinical suspicion of visceral leishmaniasis, later confirmed negative.

His neurological status deteriorated in hospital, with fever, progressive loss of head control, side-to-side head movements, right hemiparesis with tremor, and generalized hypotonia. MRI showed progressive demyelination. On suspicion of ADEM, he was treated with methylprednisolone without benefit. He developed seizures refractory to anticonvulsants. At 40 days after onset, typical diagnostic criteria for HLH were absent. Brain biopsy was consistent with ADEM. At 50 days after onset, soluble interleukin-2-receptor antibody levels were elevated, with increased expression of perforin-granzyme B. Sequence DNA analysis of blood showed a mutation in intron 10 of the Munc13-4 gene, diagnostic of FHLH3. The disease was too advanced for standard treatment with chemotherapy and stem cell transplantation; the patient continued to have seizures and died of sepsis 173 days after initial presentation. The family was offered genetic screening. (Weisfeld-Adams JD, Frank Y, Havalad V, et al. Diagnostic challenges in a child with familial hemophagocytic lymphohistiocytosis type 3 (FHLH3) presenting with fulminant neurological disease. **Childs Nerv Syst** February 2009;25:153-159). (Dr JD Weisfeld-Adams, Division of Medical Genetics, Mount Sinai School of Medicine, One Gustave L Levy Place, PO Box 1497, New York NY 10029. E-mail:james.weisfeld-adams@mssm.edu).

COMMENT. Familial HLH is an autosomal recessive multisystem disease characterized by fever, rash, splenomegaly, cytopenias, hyperferritinemia, and variable CNS manifestations with demyelination. This case report illustrates the difficulties encountered in diagnosis of HLH, and the need for a high index of suspicion and early molecular testing in cases of undiagnosed inflammatory CNS disease presenting as ADEM or pediatric MS. Leishmaniasis, considered as a possible cause of the symptoms in this case, shares similar features and is reported in 12% of HLH cases.