clumsiness to severely disabling generalized dystonia. Mild to moderate cognitive dysfunction was present in 31 (67%). Maladaptive behaviors included onychophagia in 6 patients, and impulsivity, OCD, aggression, OCD and anxiety in isolated cases. Together with 78 prior reports of 127 Lesch-Nyhan disease variants, a spectrum of clinical features includes patients with a full phenotype of classical L-N disease and patients with uric acid overproduction but no neurological or behavioral deficits. Between classical and asymptomatic variants are patients with varying degrees of motor, cognitive, or behavioral abnormalities. Of 47 previously reported cases of L-N variants, 18 (38%) had extrapyramidal features, 19 (40%) hyperreflexia and spasticity, 19 (40%) dysarthria, and 7 (15%) seizures. (Jinnah HA, Ceballos-Picot I, Torres RJ, et al. Attenuated variants of Lesch-Nyhan disease. **Brain** March 2010;133(3):671-689). (Respond: Dr HA Jinnah, Department of Neurology, Emory University School of Medicine, Atlanta, GA 30322. E-mail: hjinnah@emory.edu).

COMMENT. The diagnosis of classical Lesch-Nyhan disease is not difficult when a young child presents with self-injurious behavior, symptoms of hyperuricemia, and early-onset dystonia or clumsiness. In patients with attenuated variants of L-N disease, diagnosis should be suspected when a child has nephrolithiasis, gout, and motor or cognitive abnormalities. Serum uric acid may be normal or mildly elevated initially, and molecular testing for mutations of *HPRT* gene sometimes unavailable. A knowledge of the spectrum of neurological variants of L-N disease should lead to more prompt diagnosis, treatment, and carrier identification for family counseling.

ACUTE ENCEPHALITIS

ACUTE ENCEPHALITIS WITH REFRACTORY, REPETITIVE PARTIAL SEIZURES

Clinical characteristics and outcome of 29 children (19 male; 10 female) with acute encephalitis with refractory, repetitive partial seizures (AERRPS) were analyzed in a retrospective, multicenter nationwide questionnaire-based study in Japan. Age of disease onset ranged from 1 to 14 years (6.8 +/- 4.0). Four had a history of febrile seizures but none had preceding neurologic abnormalities. The acute phase characterized by persistent fever (>39C) and persistent seizures ranged from 15 to 312 days. Seizures were partial and repetitive, and consisted of eye deviation or facial twitching. They were refractory to conventional anticonvulsants and were suppressed by high-dose IV barbiturate. Impairment of consciousness was common. Pre-treatment EEGs in the first 14 days of illness showed high voltage slowing and in later stages, interictal epileptiform discharges. Ictal discharges were periodically repeated every 5-10 min. Early MRI, within 7 days of onset, showed mild brain edema in 2 of 14 cases, and later, 6 showed hippocampal or amygdaloid hyperintensities on FLAIR, without development of epileptic foci. Serial MRI revealed diffuse brain atrophy after a month or more. Laboratory studies showed high serum ferritin (221-2370 mg/dl) in 4/4 and positive anti-GluRe2 antibodies in 6/9 blood and CSF specimens. Other CSF abnormalities included pleocytosis in 19/29, high protein in 5/29, and high neopterin in 4/4. IV methylprednisolone in 12 was

effective only in 2; IV immunoglobulin was ineffective in all 13 patients treated. Follow-up ranged from 8-194 months (mean 60.9 months), and outcome was uniformly poor. All had residual epilepsy without a latent period, and the majority had cognitive impairment. WISC scores were <70 in 16 and <20 in 10 patients. All with GluRe2 antibodies were cognitively impaired. Other neurologic deficits included memory impairment, autistic tendency, hyperkinesis, learning disability, and emotional instability, each occurring in 15-22% cases. One patient died of unknown cause at 9 year after onset. (Sakuma H, Awaya Y, Shiomi M, et al. Acute encephalitis with refractory, repetitive partial seizures (AERRPS): a peculiar form of childhood encephalitis. **Acta Neurol Scand** April 2010;121:251-256). (Respond: Dr H Sakuma, National Center of Neurology and Psychiatry, Tokyo, Japan. E-mail: sakumh@ncnp.go.jp).

COMMENT. AERRPS differs from most types of childhood encephalitis with post-encephalitic epilepsy in that intractable partial seizures develop without a latent period between the acute febrile phase and the onset of epilepsy. Awaya Y and Fukuyama Y (1986) first described this unusual form of encephalitis and post-encephalitic epilepsy in 5 cases. More than 30 cases have now been reported in Japan (Awaya et al. 2007). Similar cases have also been reported worldwide. The cause is unknown but an autoimmune process related to anti-GluRe2 antibodies may be contributory.

ANTI-N-METHYL-D-ASPARTATE RECEPTOR ENCEPHALITIS: RESPONSE TO EARLY TREATMENT

A 5-year-old girl with anti-N-methyl-D-aspartate (NMDA) receptor encephalitis and near-complete recovery following early diagnosis and treatment with immune globulin and steroids is reported from Indiana University, University of Pennsylvania, and Mayo Clinic. She presented with episodic headache, paresthesias and weakness of left hand, agitation, and slurred speech. CT scan, MRI, and video-EEG were initially normal. Symptoms rapidly progressed with choreoathetoid movements, urinary incontinence, mutism, rhythmic tongue thrusting, and dysphagia. EEG on day 3 showed bilateral slowing and occipital epileptiform discharges. Methylprednisolone (IV, 40 mg/kg/day) for 4 days was followed by improvement that plateaued. IV Octagam (0.4 g/kg/day) was given for 5 days, and oral steroids were tapered over 12 weeks. At 5-week follow-up, recovery was almost complete. Immunohistochemical analysis (Mayo Clinic) of CSF obtained on admission revealed anti-NMDA antibodies, with molecular confirmation at University of Pennsylvania. Pelvic MRI was unremarkable, but close observation was advised to rule out relapse and occult ovarian teratoma. (Breese EH, Dalmau J, Lennon VA, Apiwattanakul M, Sokol DK. Anti-N-methyl-D-aspartate receptor encephalitis: early treatment is beneficial. **Pediatr Neurol** 2010;42:213-214). (Respond: Dr Sokol, Riley Hospital for Children, Indiana University School of Medicine, 575 West Drive, Indianapolis, IN 46202. E-mail: dksokol@iupui.edu).

COMMENT. Specific antibody detection is indicated in young children presenting with acute psychiatric and movement disorders such as agitation and dyskinesias. Ovarian teratoma associated with NMDAR encephalitis in young female