juvenile MG. Seven of 7 treated with PLEX alone responded, 5 of 10 treated with IVIG alone responded, and 9 of 10 patients who received both responded. Response rate to PLEX was significantly higher and more consistent than that of IVIG. Of 17 patients treated by thymectomy, 11 were significantly benefited. (Liew WKM, Powell CA, Sloan SR, et al. Comparison of plasmapheresis and intravenous immunoglobulin as maintenance therapies for juvenile myasthenia gravis. **JAMA Neurol** 2014 May;71(5):575-80).

COMMENTARY. PLEX and IVIG both have high response rates as maintenance therapies for juvenile MG, but PLEX response is superior. Nine of 11 patients with response graded as "B"-good improvement [1][2], had early thymectomies.

References.

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INFECTIOUS DISEASES

DIAGNOSIS OF NEUROLEPTOSPIROSIS

Investigators at University of California, San Francisco, and other centers, report a case of leptospirosis in a 14-year-old boy with severe combined immunodeficiency. He presented with headache and fever that lasted 6 days. In the previous 8 months he swam in a river in Puerto Rico and in a pool frequented by feral cats. A previous episode of fever and headache was complicated by conjunctivitis and followed by uveitis and thrombocytopenia. A recurrence of headache and fever was associated with increased CSF white cells (125/cmm) and protein (97 mg/dL), lowered glucose (24 mg/dL) and negative cultures, compatible with meningoencephalitis. He was readmitted with fever, headache, weakness, myalgias, nausea and vomiting. MRI showed T2-weighted hyperintensities in the basal ganglia and granulomatous leptomeningitis. A worsening hydrocephalus was treated with an extraventricular drain, and new-onset status epilepticus was controlled by a medically induced coma. CSF and serum samples sent for unbiased next-generation sequencing was positive for leptospira infection in the CSF but not in serum. Clinical assays for leptospirosis were negative. Treatment with high-dose intravenous penicillin G was followed by a gradual recovery over 7 days, resolution of status epilepticus, normal CSF, and resolution of leptomeningitis on serial MRI scans. PCR and serological testing at the CDC subsequently confirmed evidence of Leptospira santarosai infection. (Wilson MR, Naccache SN, Samayoa E, et al. Actionable diagnosis of neuroleptospirosis by next-generation sequencing. **N Engl J Med** 2014 Jun 19;370(25):2408-17).

COMMENTARY. Leptospirosis is caused by spirochetes contracted from the urine of infected wild or domestic animals, usually while swimming in contaminated water. The incubation period is 5 to 14 days (range, 2 to 30 days). The most characteristic clinical findings are conjunctival suffusion, uveitis, and myalgias of the calf and lumbar regions [1]. An initial septicemic phase is followed by an immune-mediated phase,

complicated by purpuric rash. Duration varies from less than 1 week to several months, with occasional episodes of apparent recovery. Severity ranges from self-limited systemic illness (90% patients) to life-threatening illness with jaundice, renal failure, and hemorrhagic pneumonitis (Weil syndrome).

References.
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HEADACHE DISORDERS

THALAMIC FUNCTION AND VESTIBULAR MIGRAINE

Investigators at Universities of Naples and Salerno, Italy; and Maastricht University, the Netherlands, explored the functional response of vestibular neural pathways using whole-brain blood oxygen level dependent (BOLD) fMRI during caloric vestibular stimulation in 12 patients (mean age 31.2 +/- 5 yrs) with vestibular migraine (VM), in 12 healthy controls, and in a group of age- and sex-matched patients with migraine without aura (MwoA). In all subjects, caloric vestibular stimulation elicited activation in bilateral insular cortex (right > left), right parietal cortex, right thalamus, brainstem, and cerebellum. While all participants demonstrated this general pattern of response, patients with VM showed a significantly increased left medio-dorsal thalamic activation in response to an ipsilateral vestibular stimulation, relative to both healthy controls and patients with MwoA. The magnitude of the thalamic activation was positively correlated with the frequency of migraine attacks in patients with VM. (Russo A, Marcelli V, Esposito F, et al. Abnormal thalamic function in patients with vestibular migraine. **Neurology** 2014 Jun 10;82(23):2120-6).

COMMENTARY. Patients with VM have abnormal thalamic functional response to vestibular stimulation. These findings are consistent with the current view of VM as a migraine subtype clinically characterized by vestibular symptoms and correlated with interictal dysfunctional central vestibulo-thalamocortical processing [1]. Both structural and functional thalamic abnormalities are documented in patients with migraine [2].

References.
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DEMYELINATING DISORDERS

EVALUATION OF REVISED DIAGNOSTIC DEFINITION OF MS

Investigators at Erasmus MC, Rotterdam, The Netherlands, evaluated the 2012 revised IPMSSG consensus definitions in a cohort of children with acquired demyelinating syndromes (ADS) prospectively followed for 2 years from Jan 2007. An MRI within 90 days after first disease onset was a criterion for inclusion. Of 82 children