Etiology was unidentified in 30 patients (71%) and termed primary PTC-HH. Secondary causes were detected in 12 (28.6%), defined as secondary PTC, and these included familial Mediterranean fever in 2, preceding trauma (2), and one of each of the following: mycophenolate mofetil-induced PTC, hypervitaminosis A, corticosteroid withdrawal with nephrotic syndrome, oral contraceptives, Guillain-Barre syndrome, urinary tract infection, varicella-zoster virus infection and dural venous sinus thrombosis with otitis media. Treatment included LP, acetazolamide (effective in 14 (37.8%)), and topiramate (effective in 13 of 17 patients (82.4%)). Mean duration of medical treatment was 9 months (range 1-48 months). Ventricular peritoneal shunt was beneficial in 3 patients with impaired visual fields, and visual acuity was normal in all patients at follow-up. (Per H, Canpolat M, Gumus H, et al. Clinical spectrum of the pseudotumor cerebri in children: Etiological, clinical features, treatment and prognosis. Brain Dev 2013 Jun;35(6):561-8). (Respond: Huseyin Per, Erciyes University, Division of Pediatric Neurology, Talas, Kayseri 38039, Turkey. E-mail: hper@erciyes.edu.tr).

COMMENT. Criteria for the diagnosis of idiopathic intracranial hypertension (HH) or pseudotumor cerebri (PTC) are as follows: 1) symptoms and signs of increased intracranial pressure or papilledema, 2) elevated CSF pressure at LP, 3) normal CSF composition, and 4) normal brain imaging (Per H et al. Brain Dev 2013 Jun;35:561-8). Treatable associated disorders should be excluded or treated. Topiramate appeared more effective than acetazolamide in this study and may be used as the drug of choice. Prompt diagnosis and management are important to prevent loss of visual field and acuity.

SLEEP DISORDERS

THALAMIC GLUTAMATE/GLUTAMINE IN RESTLESS LEGS SYNDROME

Investigators at Johns Hopkins University, Baltimore, MD, studied glutaminergic activity and arousal in 28 adults with restless legs syndrome (RLS) and 20 matched controls, using proton magnetic resonance spectroscopy. The thalamic glutamate/glutamine/creatine ratio was higher in patients with RLS than controls (p=0.016) and correlated significantly with the wake time during the sleep period (p=0.007) and all other RLS-related polysomnographic sleep variables (p<0.05) except for periodic leg movements during sleep (PLMS/hour). Glutamate metabolism is strongly related to arousal sleep disturbance but not to PLMS motor features of RLS. This finding contrasts with the reverse for dopamine that shows a limited relation to arousal sleep disturbance but strong relation to PLMS. (Allen RP, Barker PB, Horska A, Earley C J. Thalamic glutamate/glutamine in restless legs syndrome. Neurology 2013 May 28;80(22):2028-34). (Response: Dr RP Allen, E-mail: richardjhu@mac.com).

COMMENT. An increased glutaminergic activity in RLS demonstrated in this study represents a new RLS abnormality involving thalamocortical activation in a major nondopaminergic neurologic system. The authors (Allen RP, et al) conclude that the combination of glutaminergic (sleep disturbance) and dopaminergic (sensory symptoms, PLMS) abnormalities are involved in the full RLS symptomatology. The elevated
glutamate levels are considered a reflection of “hyperarousal” of RLS, which leads to sleep disturbance at night.

In an editorial (Neurology 2013 May 28;80(22):2006-7), Winkelman JW asks the question, is RLS a sleep disorder, a movement disorder, or a chronic pain disorder? He concludes that individual patients should be subtyped into biologically based phenotypes, with or without sleep disturbance, PLMS, or painful RLS. Although the current Hopkins study was confined to older subjects, RLS is also a pediatric problem and is closely associated with brain iron insufficiency and dopaminergic dysfunction. (Connor JR et al. Brain 2011 Apr;134(Pt 4):959-68). (Dosman C. et al. Paediatr Child Health 2012 Apr;17(4):193-7).

**Oral iron and RLS.** Oral iron treatment is initiated for RLS if serum ferritin is below 50 ng/mL. In a study of 22 children referred because of sleep disturbances, median age at onset of RLS symptoms was 7.5 months (range, 0-40 months). In addition to kicking or hitting the legs, the most striking symptoms were awakening after 1-3 hours of sleep followed by screaming and crying. Oral iron supplementation had a positive ferritin-concentration-dependent clinical effect. A relation between high PLMS index and low ferritin levels was demonstrated. An increased awareness of RLS in early childhood is recommended (Tilma J, et al. Acta Paediatr 2013 May;102(5):e221-6).

**NARCOLEPSY AND H1N1 INFLUENZA VACCINATION**

The incidence of narcolepsy between January 2000 and December 2010 in children in western Sweden and its relation to the Pandemrix H1N1 influenza vaccination were assessed by collection of data from hospital and clinic medical records and by parent telephone interviews. Of 37 children identified with narcolepsy, 9 had onset of symptoms before the H1N1 vaccination and 28 had onset of symptoms within 12 weeks postvaccination. Median age at onset was 10 years. All patients in the postvaccination group were positive for human leukocyte antigen (HLA)-DQB1*0602. The incidence of narcolepsy was 25 times higher after the vaccination compared with the time period before. Pandemrix H1N1 vaccination is a precipitating factor for narcolepsy. Postvaccination narcolepsy has a lower age at onset and more sudden onset than generally seen. (Szakacs A, Darin N, Hallbook T. Increased childhood incidence of narcolepsy in western Sweden after H1N1 influenza vaccination. Neurology 2013 Apr 2;80(14):1315-21). (Response: Dr A Szakacs, County Hospital, Halmstad, Sweden).

**COMMENT.** An abrupt increase in the incidence of childhood narcolepsy also followed an adjuvanted AH1N1 vaccine in Finland. The incidence of narcolepsy was 9.0 in the vaccinated as compared to 0.7/100,000 person years in unvaccinated individuals, the rate ratio being 12.7 (Nohynek H, et al. PLoS One 2012;7(3):e33536). A similar increased risk of narcolepsy followed vaccination with adjuvanted pandemic A/H1N1 2009 vaccine in England (Miller E, et al. BMJ 2013 Feb 26;346:f794). In contrast, no increase in narcolepsy diagnoses followed the H1N1 pandemic and vaccination campaign in Denmark, in South Korea (Choe YJ, et al, Vaccine 2012 Dec 14;30(52):7439-42), or in China (Han F, et al, Ann Neurol 2011 Sep;70(3):410-7). An autoimmune process is considered the most likely mechanism for the narcolepsy following influenza vaccination (Kornum BR, et al. Curr Opin Neurobiol 2011 Dec;21(6):897-903).