

cingulate epilepsy confirmed by MRI and response to lesionectomy, 4 with lesions in the posterior cingulate location had electroclinical findings suggestive of a temporal origin of the epilepsy. Of 10 anterior cingulate cases, 6 in a typical (Bancaud) group had hypermotor/hyperkinetic seizures, rarely generalized, with fear, laughter, or severe interictal personality changes, and 4 were atypical, with simple motor seizures, frequently generalized, and a less favorable long-term surgical outcome. All atypical cases were associated with an underlying infiltrative astrocytoma. Posterior cingulate gyrus epilepsy is regarded as a pseudotemporal epilepsy. (Alkawadri R, So NK, Van Ness PC, Alexopoulos AV. Cingulate epilepsy: Report of 3 electroclinical subtypes with surgical outcomes. **JAMA Neurol** 2013 Aug 1;70(8):995-1002). (Response: Rafeed Alkawadri MD, Yale University School of Medicine, 15 York Street, LC17148, New Haven, CT 06510. E-mail: mhdrafeed.alkawadri@yale.edu).

COMMENT. Surface EEG is often inaccurate in localizing a deep-seated epileptogenic zone in a patient with cingulate epilepsy. Symptoms of cingulate epilepsy are heterogeneous and dependent on an anterior or posterior localization of the lesion. Anterior lesions are associated with hyperkinetic behavior, cycling and running and gelastic seizures, expressed by mirthless laughter. Posterior cingulate seizures resemble temporal lobe epilepsy. This report emphasizes the importance of an MRI-identifiable lesion in the diagnosis of cingulate epilepsy.

## **INTRACRANIAL PRESSURE DISORDERS**

### **SPONTANEOUS INTRACRANIAL HYPOTENSION**

Investigators at the Department of Neurosurgery, Cedars-Sinai Medical Center, Los Angeles, CA, evaluated 24 children (18 girls, 6 boys) with spontaneous intracranial hypotension, seen 2001-2012. Onset of symptoms was at mean age of 14.3 years (range, 2-19 years). The majority (23 patients) presented with orthostatic headaches, mainly occipital, and one with a non-positional headache. Additional symptoms included nausea, neck pain, dizziness, and hearing abnormalities. Precipitating headache factors included lifting, dance class, handstands, and football. Spinal MRI demonstrated a CSF leak in 12 (50%) patients, usually thoracic in location, and spinal meningeal diverticula without a leak in 10 (42%). Underlying connective tissue disorders in 13 patients (54%) included Marfan syndrome in 3, Marfan-like syndrome in 4, Ehlers-Danlos syndrome in 2, and hypomelanosis of Ito in 1. Dural ectasia with multiple meningeal diverticula were found in all 3 patients with Marfan syndrome.

Treatment consisted of bed rest and hydration, epidural blood patches in 23, with permanent resolution in 9 patients (39%). Injections of fibrin glue directed at the CSF leak were successful in 2 patients (25%). Surgical treatment had a good result in 10 patients (91%), with permanent resolution of symptoms. Acetazolamide for rebound high intracranial pressure headache was required in 5 patients. Overall, outcome was good in 22 patients (92%) and poor in 2 (8%). (Schievink WI, Maya MM, Louy C, Moser FG, Sloninsky L. Spontaneous intracranial hypotension in childhood and adolescence. **J Pediatr** 2013 Aug;163(2):504-10). (Response and reprints: Dr Wouter I Schievink. E-mail: SchievinkW@cshs.org).

COMMENT. **Intracranial hypotension and Marfan syndrome.** A PubMed search found several single case reports of spontaneous intracranial hypotension in children, two with Marfan syndrome. A 14-year-old girl followed for Marfan syndrome presented with a postural headache for a month, and MRI showed bilateral subdural hematomas. Large lumbosacral arachnoid diverticula were shown on spinal MRI. Headaches resolved after epidural blood patching. Dural ectasia is a common finding with Marfan syndrome. (Cheuret E, et al. **Childs Nerv Syst** 2008 Apr;24(4):509-13).

A 12-year-old girl with Marfan syndrome, sacral dural ectasia, and tonsillar herniation, presented with headache, unresponsive to surgical decompression of the foramen magnum. Further MR imaging demonstrated CSF leak at the level of the dural ectasia, and the headache resolved after blood patch of the leak. (Puget S, et al. **J Neurosurg** 2007 Jan;106(1 Suppl):48-52).

Skull thickening, paranasal sinus expansion, and sella turcica shrinkage are reported in a 29-year-old man with a history of VP shunt placement following traumatic brain injury at 9 years of age. MRI showed signs of chronic intracranial hypotension, and LP opening pressure was not recordable. Secondary installation of a valve to restore normal ICP is recommended. (Yoon MK, et al. **J Neurosurg Pediatr** 2013 Jun;11(6):667-72).

## **PSEUDOTUMOR CEREBRI SYNDROME**

Investigators from the University of Texas Southwestern Medical Center, Dallas, and centers in Philadelphia and Salt Lake City, propose updated criteria for the diagnosis of pseudotumor cerebri (PTCS) and its variations. Idiopathic intracranial hypertension (IIH) is an appropriate term for a subset of patients with primary IH of unclear etiology, but not for those precipitated by an identifiable secondary cause. The syndrome is best described using the umbrella term PTCS. Required criteria for the diagnosis of PTCS are as follows: a) Papilledema, b) Normal neurologic examination (except cranial nerves), c) Normal MRI (or CT), d) Normal CSF composition, and e) Elevated lumbar puncture opening pressure (>250 mm for adults; >280 mm for children [250 if not sedated and not obese]).

With no papilledema, diagnosis requires (b) to (e) satisfied plus abducens nerve palsy.

Uncommon manifestations include a facial nerve palsy, hemifacial spasm, or radicular pain. CSF rhinorrhea or otorrhea and confirmation of a CSF leak are highly suggestive of PTCS diagnosis. Papilledema may be absent in recurrent PTCS cases because of gliosis in the nerve or optic atrophy.

Neuroimaging abnormalities highly suggestive of PTCS are: 1) Empty sella. 2) Flattening of posterior aspect of the globe, 3) Distension of the perioptic subarachnoid space, and 4) Transverse venous sinus stenosis. Tonsillar ectopia is more frequent in cases of PTCS but is not specific; it may be a sign of low CSF pressure, and may indicate an increased risk for herniation with LP. (Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. **Neurology** 2013 Sep 24;81(13):1159-65). (Response: Dr Friedman, E-mail: Deborah.Friedman@UTSouthwestern.edu).