

thrombosis in 6 patients, upper respiratory tract infections in 4, iron deficiency anemia in 3, steroid withdrawal in 3 epilepsy patients, risperidone or cyclosporine usage in 3, Brucella infection, post-traumatic and slit ventricle syndrome in one each of the secondary PTC group. Comorbid disorders in the idiopathic group were related to obesity (hypertension, diabetes), and in the symptomatic group, epilepsy, and vitamin D deficiency. Papilledema was lessened by acetazolamide in 72%. (Degerliyurt A, Teber S, Karakaya G, et al. Pseudotumor cerebri/idiopathic intracranial hypertension in children: An experience of a tertiary care hospital. **Brain Dev** 2014 Sep;36(8):690-9).

COMMENTARY. Idiopathic intracranial hypertension (IIH) is defined as intracranial pressure increase with no intracranial pathology and a normal CSF content [1]. The term, “pseudotumor cerebri,” is used where an etiology for intracranial hypertension is identified or suspected [2]. Olfactory impairment, an under-recognized complication of idiopathic intracranial hypertension, is studied in relation to astronauts in head-down tilt positions [3]. Many long-duration astronauts develop signs of elevated intracranial pressure and have olfactory threshold dysfunction.

**Controversy regarding efficacy of acetazolamide in IIH.** The efficacy of acetazolamide in IIH is questioned since a randomized controlled trial failed to show a significant difference in lumbar puncture pressure, headache disability and visual acuity in the acetazolamide vs placebo groups [4]. A prospective cohort study found that weight loss is an effective therapy in IIH [5][6], leading to the proposition that weight loss may be the reason for the small improvement in the acetazolamide cohort in a most recent IIH Treatment Trial [6]. Proponents of a positive effect of acetazolamide in IIH argue that the effect of acetazolamide on visual field function is independent of its effect on weight loss and does not relate to the anorexigenic effect of acetazolamide [7].

#### References.

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3. Bershada EM, et al. J Neurol Neurosurg Psychiatry. 2014 Sep;85(9):959-64.
4. Ball AK, et al. J Neurol. 2011 May;258(5):874-81.
5. Sinclair AJ, et al. BMJ. 2010 Jul 7;341:c2701.
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## **HEADACHE DISORDERS**

### **SYMPTOMS AND ETIOLOGIES OF ALICE IN WONDERLAND SYNDROME**

Investigators from Children’s Hospital of Philadelphia, PA, conducted a retrospective chart review of 48 children (average age at presentation 8.1 yrs, range 5-14 yrs; 73% male) diagnosed with “Alice in Wonderland” syndrome (AWS) or “Alice in Wonderland”-like syndrome between 1993 and 2013. Micropsia occurred in 69%, teleopsia in 50%, macropsia 25%, metamorphopsia 15%, and pelopsia (objects appear nearer) in 10%. MRI and EEG were normal. Etiology was infection (viral or

streptococcal sore-throat) in 33% of patients, migraine in 6%, and head trauma in 6%. A family history of migraine was elicited in 46%; 5 parents (33%) of affected patients had experienced AWS symptoms. Of 15 patients with follow-up by telephone interview, 20% had occasional recurrences, 40% had no further attacks, 40% were still having symptoms, 4 (27%) developed migraine, and 1 patient (7%) had seizures. The interval between initial diagnosis and telephone contact was an average of 6.5 yrs (range, 2.1-13.53 yrs). (Liu AM, Liu JG, Liu Alm GW, Liu GT. "Alice in Wonderland" syndrome: Presenting and follow-up characteristics. **Pediatr Neurol** 2014 Sep;51(3):317-20).

COMMENTARY. AWS is a disorder of childhood that affects boys more often than girls, and may subsequently 'metamorphose' and develop into migraine in one quarter of patients.

**Headache metamorphosing into AWS.** Patients with a diagnosis of childhood headache in 1983 were interviewed by telephone in 1993, 2003, and 2013. Of 28 patients monitored, headaches were ongoing in 71%, and distortions of time and space were experienced by >25% and ~20%, respectively. There was no clear correlation with migraine, and patients with tension-type headaches also reported the AWS symptoms. Distortions of space and time persist into the fifth decade for many patients initially observed with headaches in childhood [1].

#### References.

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## NUTRITIONAL DISORDERS

### THIAMINE DEFICIENCY IN INFANCY

Investigators at Tel Aviv University, Loewenstein Rehabilitation Hospital, Schneider Children's Medical Center, and other centers in Israel report the clinical presentation of acute encephalopathy in 11 children and the long-term sequelae of 8 who initially survived an episode of thiamine deficiency. In 2003, 20 Israeli infants were seriously affected after being fed an international brand of soy-based formula later found to contain no thiamine. In the acute phase, 6 had bulbar signs, 5 had ophthalmologic signs and 2 had phrenic neuropathy. MRI, the best test for diagnosis of thiamine deficiency in the acute phase, showed symmetric involvement of frontal, temporal and parietal lobes, lesions in the periaqueductal region, thalami, and the mammillary bodies, findings similar to sequelae of hypoxic-ischemic injury. Of 5 patients with cardiac involvement, 3 had cardiomyopathy and died in the acute phase, and one presented with a complete atrioventricular block. Lactic acidosis was present in 10 patients. In long-term follow-up, one patient in a chronic vegetative state died after 6 years, 7 children were mentally retarded and had motor abnormalities, 6 developed severe epilepsy, 3 with West syndrome, 2 had kyphoscoliosis, and one remained in complete A-V block. (Mimouni-Bloch A, Goldberg-Stern H, Strausberg R, et al. Thiamine deficiency in infancy: Long-term follow-up. **Pediatr Neurol** 2014 Sep;51(3):311-6).