WriteClick: Editor's Choice

Editors' Note: This week's WriteClick considers 2 diagnostic disagreements. Dr. Selmonosky and authors Simon et al. discuss the controversy around the diagnosis of thoracic outlet syndrome. Drs. Wall and Corbett argue against the need for new criteria for the diagnosis of idiopathic intracranial hypertension and include the modified Dandy criteria used in the Intracranial Hypertension Treatment Trial. Authors Friedman et al. support the revised criteria described in their original article.

—Megan Alcauskas, MD, and Robert C. Griggs, MD

SONOGRAPHIC DIAGNOSIS OF TRUE NEUROGENIC THORACIC OUTLET SYNDROME

Carlos A. Selmonosky, Falls Church, VA: The report by Simon et al.[1] contributes to the confusion about the diagnosis of thoracic outlet syndrome (TOS). They described a complication of a predominant uncomplicated neurogenic form that went undiagnosed for a long time. Early diagnosis of TOS will hopefully prevent the complications that are easily diagnosed but often too late. A new classification of TOS is well-described.[2] Knowledge of the forms and types—especially the uncomplicated form—will aid clinicians in the diagnosis of TOS before complications occur. Almost all cases present with mixed symptoms and signs of neurogenic, arterial, and venous compression.

Author Response: Neil G. Simon, Jeffery W. Ralph, Michel Kliot, San Francisco: The authors thank Dr. Selmonosky for emphasizing the ongoing debate on TOS diagnosis. We disagree that our recent study[1] contributes to the confusion. We believe that our findings highlight an emerging technology for the anatomical diagnosis of nerve injury and compression, including that in neurogenic TOS.

True neurogenic TOS has a characteristic presentation and pattern of abnormalities on electrodiagnostic studies.[3] However, vascular TOS is rare and evidence of positionally induced vascular compromise (i.e., Adson sign) may be seen in a majority of healthy subjects.[4] In addition, presentations characterized as "disputed TOS" are nonspecific and cannot be objectively verified,[5] and overlap with other musculoskeletal pathologies involving the neck and shoulder region.

Neurologic diagnoses must be supported by rigorous, peer-reviewed data. This is vital because patients with erroneous diagnoses of TOS may seek surgical intervention that may result in further morbidity and health care costs.

1. Simon NG, Ralph JW, Chin C, Kliot M. Sonographic diagnosis of true neurogenic thoracic outlet syndrome. Neurology 2013;81:1965.
2. Welcome to thoracic outlet syndrome. Available at: www.tos-syndrome.com. Accessed December 9, 2013.
3. Tsao BE, Ferrante MA, Wilbourn AJ, Shields RW Jr. The electrodiagnostic features of true neurogenic thoracic outlet syndrome. Muscle Nerve 2014;49:724–727.
4. Sanders RJ, Hammond SL, Rao NM. Diagnosis of thoracic outlet syndrome. J Vasc Surg 2007;46:601–604.
5. Wilbourn AJ. The thoracic outlet syndrome is overdiagnosed. Arch Neurol 1990;47:328–330.

REVISED DIAGNOSTIC CRITERIA FOR THE PSEUDOTUMOR CEREBRI SYNDROME IN ADULTS AND CHILDREN

Michael Wall, Iowa City; James J. Corbett, Jackson, MS: We read with interest the suggestion for new criteria for the diagnosis of pseudotumor cerebri syndrome and idiopathic intracranial hypertension written by our esteemed colleagues.[1] We disagree with the need for this exercise since the nomenclature we have used over the years and for the Idiopathic Intracranial Hypertension Treatment Trial (IIHTT), we believe, is simpler, is accurate, describes the condition, and is easily modified as new information surfaces.

Diseases or syndromes should be named for what they are—"idiopathic intracranial hypertension"—rather than what they are not—"pseudotumor cerebri," "primary pseudotumor cerebri" or "pseudotumor cerebri syndrome (PTCS)." This was impressed on us during a visit to the National Eye Institute many years ago when we were told "we are not going to fund a pseudo anything." We do not believe there is need for the term PTCS. Secondary causes of intracranial hypertension should also be called what they are: e.g., vitamin A-induced intracranial hypertension, tetracycline-induced intracranial hypertension, steroid withdrawal-related intracranial hypertension, rather than subsuming them with the PTCS acronym. When criteria for IIH

are not met and no secondary cause is found, "intracranial hypertension of unknown cause" should be used. The naming convention we propose is unambiguous; PTCS could mean any of the above diagnoses.

As regards criteria, Walter Dandy[2] suggested requirements to separate IIH from other causes of raised intracranial pressure, in particular brain tumor, that were codified in an editorial by J.L. Smith in 1985. He called them the modified Dandy criteria (even though Dandy did not directly specify criteria for the diagnosis of IIH).[3] These modified Dandy criteria have been used successfully for many years.

We agree with our colleagues that as we learn more about IIH and secondary forms of intracranial hypertension, criteria for the diagnosis of the various causes of intracranial hypertension should be amended to more clearly separate IIH from secondary causes. We have updated the modified Dandy criteria for the IIHTT (table). Once the results of this clinical trial are published, any treatment recommendations will apply most directly to patients that meet these criteria and not another set of criteria. We suggest that idiopathic intracranial hypertension is the most appropriate name for this disease and that the modified Dandy criteria continue to be used and updated when appropriate as they remain excellent diagnostic criteria.

Author Response: Deborah I. Friedman, Dallas; Grant Liu, Philadelphia; Kathleen Digre, Salt Lake City: The authors thank Drs. Wall and Corbett for their comments. We suggest calling the syndrome, inclusive of all etiologies, the "pseudotumor cerebri syndrome" (PTCS)—a condition of increased intracranial pressure and papilledema due to all causes as long as the brain parenchyma is normal. This reflects the most common term used globally.[4]

Neuro-ophthalmologists and other physicians still refer to the condition as "pseudotumor" and patients are also familiar with this term. We retained the term "idiopathic intracranial hypertension" to refer to the specific condition that most frequently occurs in overweight women of childbearing age where no specific etiology is apparent. IIH was the term used in the previous criteria proposed by Friedman and Jacobson[5] in 2002, which has been cited almost 500 times. Thus, there is no conflict regarding terminology used for the IIH Treatment Trial and we are hopeful that the trial will provide us greater understanding of the pathophysiologic basis of IIH.

Our recent criteria were expanded to be able to diagnose patients with the idiopathic form and those with a secondary cause. Most importantly, there are now official guidelines for children, a population that was not included in any of the previous criteria because of lack of high-quality normative data for CSF pressure in this population.

To clarify the term "the Dandy criteria," we remind readers that Dr. Dandy[2] reported a series of cases of patients seen in the 1930s, when the only diagnostic techniques available were pneumoencephalography ("trephine and air injection") and lumbar puncture. It is possible that some of his cases did not actually have IIH, as some had atypical manifestations, such as CSF pleocytosis, preceding head trauma, drowsiness, and transient hemiplegia. He summarized his findings but did not propose diagnostic criteria. It should also be noted that Smith[3] suggested the modified Dandy criteria, but the title of his article was "Whence pseudotumor cerebri?" and did not refer to IIH. He specified an opening pressure of 200 mm CSF, which subsequent studies have shown to be too low. All previous criteria have accepted symptoms and signs. Headache and tinnitus

Table Idiopathic Intracranial Hypertension Treatment Trial Modified Dandy Criteria

1. Signs and symptoms of increased intracranial pressure
2. Absence of localizing findings on neurologic examination
3. Absence of deformity, displacement, or obstruction of the ventricular system and otherwise normal neurodiagnostic studies, except for evidence of increased CSF pressure (>200 mm water); abnormal neuroimaging except for empty sella turcica, optic nerve sheath with filled out CSF spaces, and smooth-walled non-flow-related venous sinus stenosis or collapse should lead to another diagnosis
4. Awake and alert
5. No other cause of increased intracranial pressure present
For CSF opening pressure of 200–250 mm water required at least one of the following:
Pulse synchronous tinnitus
VI palsy
Frisen grade II papilledema
Echography for drusen-negative and no other disc anomalies mimicking disc edema present
Magnetic resonance venography with lateral sinus collapse/stenosis preferably using auto-triggered elliptic centric-ordered technique
Partially empty sella on coronal or sagittal views and optic nerve sheaths with filled out CSF spaces next to the globe on T2-weighted axial scans

are common and nonspecific symptoms. True transient obscurations are more indicative of intracranial hypertension than another condition but may also be confused with transient visual loss occurring with migraine by clinicians who are not familiar with the description of this symptom.

The validity of diagnosing the syndrome of intracranial hypertension without papilledema has been contentious as long as we have been in the field of neuro-ophthalmology. The previous criteria do not directly address the diagnosis of IIH without papilledema, which has become pervasive in the world of headache medicine with unintended consequences. Allowing a diagnosis based on headache and elevated CSF pressure alone leads to false-positive and erroneous diagnoses and potentially unnecessary surgical interventions and incorrect medical treatments. Finally, the older criteria do not address the common scenario of an obese female patient with optic disc swelling, normal imaging, but an opening pressure of 190 mm of CSF.

Most experienced clinicians would consider the measured opening pressure in this case to be falsely low, given the characteristic clinical presentation, and treat the patient as if she had elevated intracranial pressure. The newly proposed criteria allow for a "probable" diagnosis of IIH syndrome in such instances.

1. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. Neurology 2013;81:1159–1165.
2. Dandy WE. Intracranial pressure without brain tumor. Ann Surg 1937;106:492–513.
3. Smith JL. Whence pseudotumor cerebri? J Clin Neuroophthalmol 1985;5:55–56.
4. Johnson I, Owler B, Pickard J, eds. The Pseudotumor Cerebri Syndrome. New York: Cambridge University Press; 2007.
5. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. Neurology 2002;59:1492–1495.

CORRECTION

Hemodynamic Changes Associated with Interictal Epileptiform Activities Using Simultaneous Video Electro-encephalography (EEG)/Near Infrared Spectroscopy (NIRS) in Patient Self Control Study (P4.330)

In the abstract "Hemodynamic Changes Associated with Interictal Epileptiform Activities Using Simultaneous Video Electro-encephalography (EEG)/Near Infrared Spectroscopy (NIRS) in Patient Self Control Study (P4.330)" by K. Sannagowdara (*Neurology*® 2014;82:P4.330), the author list is incomplete. The byline should read "Kumar Sannagowdara, MD, Sugandha Kirankumar, MD, Pyria Monrad, MD, Kurt Hecox, MD, Michael Schwabe, MD, Michael Meyer, MD, Jenna Prigge, NP, Russ Lemke, BS, Briana Horn, CRC, Harry Whelan, MD." The AAN staff regrets the omission.

Author disclosures are available upon request (journal@neurology.org).

Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children

Michael Wall, Deborah I. Friedman, James J. Corbett, et al.

Neurology 2014;83;198-200
DOI 10.1212/01.wnl.0000452039.32455.3e