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 predominantly 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.

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

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, “intrascan-
mal 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 re-
quirements 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 hyper-
tension, 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 idio-
pathic intracranial hypertension is the most appro-
riate 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 over-
weight 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 patho-
physiologic basis of IIH.

Our recent criteria were expanded to be able to diag-
nose 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 crite-
ria, but the title of his article was “Whence pseudotumor
cerebri?” and did not refer to IIH. He specified an open-
ing 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

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### 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 obstructions 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.

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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.
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