## Title

The Cerebral Venous Disorders Severity Scale



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# The Cerebral Venous Disorders Severity Scale

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Exciting new insights, much of it featured in the recent pages of *JNIS*, have provided important evidence expanding our understanding of cerebral venous outflow impairment diseases and therapies.[1–8] In fact, with each passing year, *JNIS* features more and more manuscripts addressing this subject domain.[9] The relationship between idiopathic intracranial hypertension (IIH), cerebral venous hypertension, and venous sinus stenosis is well established, so much so that the term "idiopathic" has largely become a misnomer[10] and the disorder, or at least substantial subgroups, may be more aptly referred to as "venopathic" intracranial hypertension (VIH). While the role of intracranial venous stenosis in VIH is inarguable, the role of extracranial venous stenosis or congestion as pathophysiologic drivers of chronic neurological disease is much less certain. Importantly, studies have identified similar clinical manifestations to VIH from a variety of extracranial sites of venous stenosis or congestion, or both,[11] and a potential contribution of dynamic extracranial venous stenosis in disease.[1 7] Recently, we have been considering disorders related to cerebral venous outflow insufficiency as a spectrum of conditions, including both intracranial and extracranial venous etiologies, that can collectively be referred to as cerebral venous outflow disorders (CVD), with VIH representing the flagship CVD condition. Recent publications highlight potential benefits to surgical jugular decompression, jugular stenting, or brachiocephalic interventions in select patients with a suspected venous etiology of their symptoms,[12 13] but surgical candidacy is poorly defined. These treatments carry risk, and while anecdotally many patients note symptomatic improvement after intervention, published evidence supporting their use is quite limited. This has led prominent authors to question the existence of these conditions, with some considering the protean CVD symptoms to be either too non-specific or failing to meet criteria as a distinct clinical entity, and have criticized that no recognizable outcome metrics exist.[14]

It is evident that much works needs to be done in terms of establishing rigorous methods for diagnosis and measuring disease severity for these conditions. For classic VIH (IIH; a universally accepted pathologic entity), most patients are diagnosed using established modified Dandy criteria and undergo evaluation by ophthalmology, with visual severity assessed through Frisen papilledema grade, optical coherence tomography, and nerve fiber layer thickness metrics. Overall clinical severity is usually evaluated through intracranial pressure (ICP) measures on lumbar puncture and headache scores, such as the Headache Impact Test (HIT-6). These three assessments (ophthalmologic assessment, ICP, and headache scores) have been the mainstay of understanding the severity of disease in patients, assessing candidacy for surgical treatment, and the response to interventions. However, many of these patients have additional disabling symptoms that are marginally or wholly unrepresented by these assessments, such as tinnitus, severe cognitive dysfunction, or dizziness. As our understanding of CVD expand, it is becoming clear that the usual VIH disease metrics are no longer sufficient, especially for extracranial pathologies that manifest as severe neurological symptoms in the absence of papilledema or significant elevations in ICP on lumbar puncture. A patient-by- patient review of the original English language paper by Walter Dandy reveals a highly heterogenous set of symptoms that greatly exceed the simplified cardinal symptoms that have come to be the sina qua non of "IIH".[15] The VIH rubric provides a reset of this ancient disease by widening the scope of symptoms that affect these patients.

At the second Society of Neurointerventional Surgery (SNIS) Cerebral Venous and CSF Disorders Summit held in Honolulu in early 2024, over 70 in-person participants (and over 140 total attendees, including virtual) from neurointervention, neurosurgery, headache neurology, radiology, otolaryngology, cardiothoracic surgery, vascular surgery, and others, assembled and engaged in multidisciplinary discussions of CVD and challenges facing these patients. The spectrum of CVD,[11] including conditions such as VIH, transverse sinus stenosis, CSF leak, primary pulsatile tinnitus, internal jugular vein stenosis, brachiocephalic vein stenosis, and central venous hypertension, were presented. Patients with these conditions have a collection of protean but fairly conserved symptoms of differing severities, including pressure headache, neck pain, visual abnormalities, cognitive dysfunction and "brain fog," dizziness, disequilibrium, dysautonomia, ringing and pulsatile tinnitus, barometric pressure sensitivity, cranial neuropathies, positional intolerance, sleep disorders, choking sensations or swallowing discomfort and, in rare cases, involuntary motor movements, among others. Some patients experience debilitating symptoms, such as profound cognitive impairment, loud and constant pulsatile tinnitus, or disabling dizziness in the absence of significant headache, significant ICP elevation, or objective ophthalmologic abnormalities. To fully understand the individual and collective impacts of these symptoms on a given patient, a battery of tests would be required, such as HIT-6, ophthalmologic metrics, ICP, tinnitus severity scores, dizziness scales, cognitive impairment tests, and sleep disorder screens, as well as global quality of life/functional capacity scales. Using all of these individual tests is impractical and burdensome, using none or a few established metrics, such as HIT-6, ICP, and papilledema grade, does a substantial disservice to many affected patients, and using global quality of life scales (while helpful in capturing disease impact) does not allow for differentiation of individual symptoms and their response to therapies.

We believe the solution lies in developing a validated, concise, and reliable scale, designed specifically for patients with these disorders, that captures global disease severity for afflicted patients while also allowing for assessment of core symptom components. In stroke care, the National Institutes of Health Stroke Scale (NIHSS) is a universally accepted means of measuring stroke symptoms and allowing for precise communication of disease severity in a standardized manner. The SNIS Cerebral Venous and CSF Disorders section is engaging in a concerted effort to develop the Cerebral Venous Disorders Symptom Severity scale, or CVDSS scale, which would aim to represent a

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**CEREBRAL VENOUS DISORDER SEVERITY SCALE (v1.1)**

| Symptom | SYMPTOMS ARE NOT PRESENT OR RARELY PRESENT | MILD SYMPTOMS PRESENT (interferes slightly with activities of daily living but still able to function) | MODERATE SYMPTOMS PRESENT (interferes with ability to work or carry out normal ADLs, but still able to function most days) | SEVERE OR DEBILITATIING SYMPTOMS PRESENT (interferes with ability to function/work most days) |
|---|---|---|---|---|
| **HEADACHE** (includes head pressure, eye pressure, base of head pain) | O 0 | O 1 | O 2 | O 3 |
| **COGNITIVE DYSFUNCTION** (includes brain fog, poor memory, difficulty thinking, or word finding difficulty) | O 0 | O 1 | O 2 | O 3 |
| **TINNITUS** (sound in the ear, includes whooshing, high pitched ring) | O 0 | O 1 | O 2 | O 3 |
| **DIZZINESS** (includes balance difficulties, vertigo, disequilibrium) | O 0 | O 1 | O 2 | O 3 |
| **VISUAL SYMPTOMS** (includes blurry or cloudy vision, visual spotting, loss of vision) | O 0 | O 1 | O 2 | O 3 |

| | NOT PRESENT | OCCURS OCCASIONALLY | OCCURS FREQUENTLY |
|---|---|---|---|
| **INVOLUNTARY MOTOR EPISODES** (includes catatonia, seizures, locked in, or shaking spells) | O 0 | O 2 | O 4 |

| | NO | YES |
|---|---|---|
| **ARE YOU DISABLED BECAUSE OF YOUR SYMPTOMS?** (includes receiving disability benefits, inability to work or attend school) | O 0 | O 4 |

**TOTAL SCORE** (add the number under each selected box to obtain a total value): _____

MILD 0-5, MODERATE 6-12, SEVERE > 12

**Figure 1** Cerebral Venous Disorders Symptom Severity (CVDSS) scale inventory for standardization of disease severity and monitoring outcomes following interventions.

NIHSS-type score except specifically for CVD and its related conditions. A preliminary, non-validated scoring system has been developed by CVD experts and stakeholders and is being used in clinical practice (figure 1). At the time of this writing, a rigorously validated form of this scale is currently being developed using individual patient quantitative symptom inventory responses adjudicated with scores on validated headache, cognitive, and quality of life scales. This process will take some time to complete, but in the meantime, the presented CVDSS scale is a first step in providing clinicians with a scale to assess and monitor disease severity. We encourage readers to incorporate this scale into their practices.

As evidenced by recent publications featured in *JNIS*, our understanding of disorders of the cerebral venous system is escalating at a tremendous pace. New and exciting research and discussion topics will be highlighted at the third annual SNIS Cerebral Venous and CSF Disorders Summit held in Honolulu in March of 2025. We hope to see you there!

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