



Published in final edited form as:

J Neuroophthalmol. 2021 September 01; 41(3): 379–384. doi:10.1097/WNO.0000000000001373.

Should lumbar puncture be required to diagnose every patient with Idiopathic Intracranial Hypertension?

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Introduction- Drs. Lee and Van Stavern

Idiopathic Intracranial Hypertension (IIH) is a disorder uniquely within the purview of neuro-ophthalmologists. Although overall a rare condition, patients with IIH are commonly diagnosed and managed primarily by neuro ophthalmologists. Although the criteria for IIH has changed over the years, a constant feature of the modified Dandy criteria for IIH has been the requirement for lumbar puncture to confirm the diagnosis and to exclude alternative etiologies. Given the advancements in neuroimaging technology, and better understanding of the range of intracranial pressure, two experts debate whether LP is still necessary for diagnosis of IIH.

Should lumbar puncture be required to diagnose IIH in every patient – Yes!

Heather E. Moss, MD, PhD

High intracranial pressure (ICP) causes IIH's major morbidities of vision loss and headache. Accordingly, ICP is the target of both medical and surgical therapies for IIH and measurement of ICP is the cornerstone of diagnosis. Though what constitutes abnormally high ICP has evolved over the years, all past and current diagnostic criteria require lumbar puncture (LP) for measurement of opening pressure. Furthermore, LP enables cerebrospinal fluid (CSF) analysis in order to exclude secondary causes of high ICP. Some criteria allow for diagnosis of possible or probable IIH without LP, but none allow for definitive diagnosis of IIH without LP and CSF analysis. Thus, LP is necessary in every patient in order to diagnose definitive IIH by current guidelines.

LP is an invasive procedure, one that is increasingly harder to arrange and not enjoyed by patients. Thus, it has been proposed that LP may not be necessary to diagnose IIH in every patient. As with any clinical decision risk:benefit analysis can be helpful (table 1). By deferring LP, procedure complications and false results are avoided which may increase both doctor and patient satisfaction. However, by deferring LP the diagnosis is not confirmed and alternative diagnoses are not excluded. Specifically, measurement of opening pressure, CSF analysis and evaluating patient response to short term ICP lowering are not accomplished.

There are some patients who have high pre-test probability for IIH based on weight, papilledema and neuro-imaging findings suggestive of high intracranial pressure, and some diagnostic criteria allow for diagnosis of probable IIH on this basis. In these patients it has been proposed that LP is unlikely to change management and that they can be treated presumptively. My debate opponent has compiled a series of 68 patients with mild papilledema from presumed IIH who, despite lack of LP, were managed without negative outcomes.¹ Similarly, a multicenter study reports 156 patients with probable IIH in whom LP did not change management.² However, both of these studies suffer from selection bias in that the subjects were all seen by expert neuro-ophthalmologists. Scaling their success to other practitioners less expert in optic nerve assessment is fraught with the risk of missed diagnoses and over diagnosis. Even in expert hands, the pre-test probability that a patient has IIH never reaches 100%. A risk of not doing LP in these patients is failure to diagnose and treat a condition other than IIH that is causing their symptoms and signs. For example, compression of the optic nerve in the posterior orbit or canal can cause bilateral optic nerve edema similar to papilledema but without high ICP. An LP with normal opening pressure would lead to consideration of this and further anatomical evaluation of the posterior orbit. Additionally, some secondary causes of high ICP require CSF analysis for diagnosis. Multiple studies in the literature illustrate the non-zero likelihood of conditions with substantial morbidity being overlooked in the setting of presumed IIH diagnosis. A single center study of 86 previously diagnosed cases of IIH included 1 with cerebral venous sinus thrombosis.³ In multi-center study of 496 consecutive new neuro-ophthalmology referrals (102 referred for papilledema or abnormal optic disc appearance) one of these (0.2% of all patients, 1% of papilledema patients) was a previously missed diagnosis of secondary elevated ICP.⁴ Another multi-center study of 206 patients referred to neuro-ophthalmology for assessment of papilledema included 4 who were found to have non-IIH diagnoses requiring directed therapy including uveitis, meningitis and leptomenigeal carcinomatosis⁵ (personal communication). All of these had positive neuro-imaging findings suggesting high ICP and no neuro-imaging findings suggesting a secondary cause.

The previous paragraph discussed the problem of over diagnosis of IIH leading to missed diagnosis of other conditions with high morbidity needing directed treatment. The corollary of this is over diagnosis of IIH leading to un-necessary diagnostic testing and treatment. In the Fisayo study 16/86 patients previously diagnosed with IIH had pseudopapilledema,³ which is a clinical diagnosis not necessitating any additional work up. In the Stunkel study, 15/89 patients with suspected papilledema due to IIH were re-diagnosed to have pseudopapilledema.⁴ While I do not advocate for LP to be performed for work up of pseudopapilledema, in cases where there is diagnostic uncertainty due to non-expert or equivocal optic nerve exam, LP would prevent a false diagnosis of IIH and reduce harm due

to unnecessary medical or even surgical treatment, which was proposed for 4/34 patients found to be mis-diagnosed with IIH in the Fisayo study.³

In conclusion, lumbar puncture is an important component of IIH diagnostic criteria because it confirms high intracranial pressure and excludes secondary causes of high intracranial pressure. Presumptive diagnosis of IIH on the basis of high BMI, headache, bilateral optic disc abnormalities and/or neuro-imaging findings of high intracranial pressure leads to overdiagnosis of IIH due to failure to confirm the diagnosis and exclude other diagnoses. The consequences are inadequate treatment for a missed diagnosis or inappropriate testing and/or treatment for IIH. It is therefore important to follow the accepted diagnostic criteria including LP for diagnosing this condition.

Is Lumbar Puncture Necessary for the Diagnosis of Idiopathic Intracranial Hypertension? Not always.

Edward Margolin, MD

Lumbar puncture (LP) is part of the modified Dandy criteria for establishing the diagnosis of idiopathic intracranial hypertension (IIH) and is considered by many to be the cornerstone for making the diagnosis. The purpose of LP is two-fold: to confirm increased opening pressure (OP) of cerebrospinal fluid (CSF) during the procedure which is an indirect indication of intra-cranial pressure (ICP), and to check its composition to rule out other, more ominous causes of increased ICP, either an infection or, very rarely, a neoplasm. The main disadvantage of obtaining an LP is of course the pain and discomfort to the patient during the procedure as well as intra- and post-procedural side effects of which the most common is severe headache due to persistent CSF leak from the site of dural puncture⁶. Lumbar puncture can also be difficult to arrange for practitioners who are non-neurologists and the technical difficulties in performing an LP are often exacerbated by the fact that most patients with IIH have increased body-mass index (BMI). As more and more LPs are performed through fluoroscopic guidance, radiation exposure from fluoroscopy is also not trivial.

As most neuro-ophthalmologists are aware though, the LP in a patient who is in a typical demographic group for IIH (women of childbearing age with increased BMI) will demonstrate expected results: elevated OP and normal CSF composition. We know that OP during LP does not correlate with the severity of the disease nor with visual prognosis^{7,8}. Neuro-imaging, which would have been performed prior to an LP, should demonstrate indirect neuro-imaging signs in nearly all patients suspected of having IIH. As experienced neuro-ophthalmologists are also aware, the composition of CSF in patients who are in a typical demographic group for IIH, have typical symptoms and are systemically well, is nearly always normal. Thus, the question of this point-counterpoint manuscript: “Does one really need to obtain an LP for patients who are in a typical demographic group for IIH, have typical symptoms and are otherwise systemically well?”

The incidence of side effects of LP is not trivial. Post-LP headache occurs in up to 25% of patients⁶ and requires placement of blood patch in a significant minority of patients. Rarer

side effects include meningitis, nerve root pain, infection at the skin site, and very rarely an epidural abscess⁹.

One should also be aware that the OP number obtained during LP is not always an “absolute truth” and is prone to error. Multiple factors can influence OP on LP including positioning of the patient, inadvertent Valsalva maneuver during the procedure, needle positioning, and characteristics of the needle being some of the confounders^{10,11}. Thus, if the OP is under 25 cm of water in a patient who is in a typical demographic group for IIH and has bilateral optic nerve head edema with indirect neuro-imaging signs of ICP, one would have to assume that either the OP is erroneous or that the normal value is not the same for all individuals and proceed with treating the patient for IIH.

There are definitely many situations when obtaining OP and CSF analysis is very useful: it should be done in all patients who are not in a typical demographic group for IIH, those who have symptoms not typical for IIH, and where pseudo-papilledema is difficult to rule out and the clinician is suspecting increased ICP as an additional cause for optic nerve head edema. LP is also needed for those patients who have severe disease which is typically defined by abnormal visual function (decreased central acuity and/or decreased mean deviation (MD) on Humphrey perimetry, previously defined as MD worse than -7.0 decibels) and for those patients in whose surgical treatments are planned¹².

While there are only a few studies^{1,2} evaluating the management of IIH patients without an LP, several IIH cohorts reported on the findings of the LP. One study evaluating 53 patients with IIH in Kuwait reported normal CSF composition in all¹³. In the IIH Treatment Trial, while the explicit data regarding the number of patients who were found to have abnormal CSF composition at the study entry was not provided, Dr. Michael Wall, one of the study’s founders, kindly provided the spreadsheet with the details of each excluded patient for the purposes of this article. Abnormal CSF composition is not listed for any of them¹⁴. My colleagues at the University of Toronto and myself followed a cohort of 68 patients with the mild presumed IIH without an LP for at least 63 weeks, with no alternate diagnosis made in any of the patients and all of the patients remaining well (this manuscript is currently under consideration), again providing data about the reasonableness of following patients with mild presumed IIH without an LP.

Modern neuro-imaging has allowed us to be able to presume a diagnosis of IIH with high degree of certainty as most patients will demonstrate several indirect neuro-imaging features associated with increased ICP with transverse sinus stenosis been reported to be present in nearly all patients with IIH¹⁵. Adequate neuro imaging (MRI with venography and contrast administration) can also effectively rule out many alternate diagnoses. Thus, if the LP is not performed, high quality MR scan of the brain with venography sequences and contrast administration is paramount.

When searching prior reported cases of misdiagnosis of IIH when LP was not performed, no definitive reports were found where the patient was in a typical demographic category for IIH who did not have “red-flag” symptoms at presentation. One study retrospectively reviewed all patients who were diagnosed with aseptic meningitis and who also had

increased opening pressure on LP¹⁶. Seven patients met both criteria and had an increased number of white blood cells in CSF with lymphocytic predominance. However, all were under the age of 19, 6 were male, papilledema was documented in 3 patients only, two of whom had neck stiffness with only patient without other constitutional symptoms at presentation who was 10 years old. This study highlights the rarity of aseptic meningitis presenting with isolated papilledema and no other red flags on history or examination. Another study reported three patients with cryptococcal meningitis all of whom had papilledema, however, two were immunocompromised and the third patient had neuroretinitis-like picture at presentation¹⁷. One report highlighted a 21-year-old male patient who presented with papilledema which was eventually linked to aseptic meningitis due to acute HIV infection; the patient had decreased central visual acuity at presentation and a low grade fever¹⁸. Another report described a 9-year-old boy who presented with symptoms of increased ICP and papilledema and was eventually diagnosed with neuroborreliosis¹⁹. Another manuscript described a 32-year-old woman who had papilledema and increased OP on LP and was found to have Human Herpes Virus-6 in the CSF. She had a BMI of 48 kg/m², however, she had a new onset of thunderclap headaches associated with photophobia, nausea and vomiting which were not responsive to oral analgesics and sumatriptans²⁰. Several reports described spinal cord tumors misdiagnosed as IIH: in one the patient was a 41 year-old woman who was not obese (exact BMI was not reported), had bilaterally decreased vision and a “left nasal hemianopia” with mildly dilated ventricles on MRI; in the second report the patient was a 48-year-old man with normal neuro-imaging whose BMI was not reported; in the third report a 41-year-old man complained of distortion of peripheral vision and was found to have papilledema, his OP on LP was normal but because of the increased protein in CSF he was eventually diagnosed with spinal cord plasmacytoma; in the fourth report a morbidly obese 41-year-old woman experienced progressive visual loss, headaches, nausea and vomiting and was found to have papilledema and anisocoria and was eventually diagnosed with intramedullary astrocytoma as a culprit producing elevated CSF protein^{20–24}. Another report highlighted a young man who presented with papilledema and was eventually diagnosed with carcinomatous meningitis due to adenocarcinoma of the lung; the patient was a very thin 30-year-old man²⁵. Two cases of spinal leptomeningeal lymphoma were reported that mimicked IIH: a 43-year-old woman whose BMI was not reported had symptoms of headaches and dizziness for a year, was found to have papilledema and elevated CSF protein and eventually abnormal spinal MRI led to the diagnosis, the second case was that of a 49-year-old man whose BMI was also not reported and who also had symptoms of increased ICP, was found to have papilledema while MRI of the brain was normal, eventually LP was performed demonstrating increased protein and MRI of the spine led to the final diagnosis²⁶. These reports emphasize that while very rarely patients with underlying CNS infection or malignancy can present with isolated symptoms of increased IIH and normal brain imaging, they practically never fit all of the criteria outlined below that sum up the characteristics of a patient with presumed IIH where LP can be safely deferred.

Of course, an important caveat for the conclusion that LP is not necessary for the diagnosis of IIH in patients who are in a typical demographic category is that the involvement of

an experienced neuro-ophthalmologist is required- someone who can recognize atypical features of the disease at presentation or during follow up.

In conclusion, the purpose of performing LP before diagnosing a patient with IIH is two-fold: to confirm elevated OP and to rule out mimickers of IIH by establishing its normal composition. In a large cohort of patients with typical IIH reported in this issue of JNO where LP was performed at diagnosis, it did not change the management in any patient and an alternate diagnosis was not established in any patient². Several large cohorts of typical IIH patients reported in the literature echo this finding. Review of individual reports describing misdiagnosis of IIH in the absence of LP did not yield any results where the patient was both in the typical demographic group for IIH and did not have any red flags on their presentation. Thus, one can conclude that in a patient with presumed papilledema who is in a typical demographic group for IIH (women 18–45 years of age with increased body-mass index), has no red-flag symptoms at presentation and is systemically well, who had adequate neuro-imaging that confirmed expected indirect neuro-imaging findings of increased ICP and no other pathology, and who has mild to moderate IIH where surgical treatment is not planned, LP may not be required for the diagnosis if the followed up by an experienced neuro-ophthalmologist is available.

Rebuttal-Dr. Moss

Dr. Margolin and I agree about many things.

First, we agree on the facts, namely that current diagnostic criteria for IIH require lumbar puncture, that lumbar puncture is helpful to measure ICP and evaluate for secondary causes of high ICP, that patients don't like lumbar punctures, that there is a risk of procedural complications and that false positive and negative results can complicate management. Second, we share the opinion that there are situations in which lumbar puncture for work up of suspected IIH is critically important. These include situations in which the pre-test probability for a disease other than IIH is higher such as for patients with atypical demographics or with "red flag" symptoms. These also include situations in which the risk of incorrect IIH diagnosis and resultant incorrect management is higher such as for patients with severe vision loss or in whom a surgical intervention is planned. Third, we agree on the importance of an experienced neuro-ophthalmologist being involved in the care of patients with suspected IIH so that features suggesting the diagnosis is not IIH can be identified, and appropriate management instituted.

Dr. Margolin makes the excellent point that there are many patients for whom final outcome is not impacted by lack of LP. Though I do not routinely recommend against LP in my own practice, I have managed plenty of patients with likely IIH without LP due to their active refusal to get the test, or more passive test avoidance by not attending the appointment or even cancelling it. I do not recall any bad outcomes as a result. However, I do not think we should go so far as to routinely recommend against LP, even for specific subpopulations. The pre-test probability may approach but never reaches 100%. A low risk of a severe disease bears consideration and discussion with the patient.

My other concern is that it is not practical to have a neuro-ophthalmologist involved in every case of suspected IIH because we are in short supply²⁷. Non-experts will be involved and it is incumbent upon us to provide guidance on diagnosis and management. Any message that LP is optional is at risk of misinterpretation and misapplication. So, let's keep it simple and follow the diagnostic criteria, which require LP. If a patient declines to get the test, we can still manage them, after appropriate education about what information is missing and the diagnostic uncertainty that that creates.

Rebuttal, Dr. Margolin:

I thank Dr. Moss for writing an elegant piece about why LP is important as a part of diagnostic process in patients with IIH. Her main argument is that an LP allows confirmation of diagnosis by measuring OP of CSF and allows ruling out of alternate diagnosis by evaluating CSF content.

Dr. Moss argues that both papers published in this issue of JNO suffer from selection bias as evaluation and management of patients was done by expert neuro-ophthalmologists^{1,2}. I agree with this and thus the recommendation for avoiding the LP in a select group of patient includes availability of expert neuro-ophthalmological opinion and follow up.

Example of optic nerve compression in the posterior orbit or optic canal has been provided as one scenario where optic nerve head may be elevated but CSF opening pressure will be normal thus an alternate diagnosis will be considered based on the LP. I would counter that in this particular scenario one would expect central visual acuity to be decreased. In addition, optic nerve compression anywhere along its pathway should also be visible on an MRI of the brain and orbit with contrast enhancement, a study that should be performed in all patients with suspected IIH where LP is not performed. Regarding a referenced study where there was one case of dural venous sinus thrombosis in a cohort of 86 patients with IIH³, I would counter again that if the imaging study detailed above would have been performed, this misdiagnosis would have been avoided. Further, LP in the case of dural sinus thrombosis is not helpful in making the diagnosis as it would have indicated increased OP with normal constituents thus pointing one toward making the erroneous diagnosis of IIH if appropriate neuro-imaging has not been performed. It is difficult to comment on a recent paper by Stunkel, etc⁴ where 1 out of 102 patients referred for papilledema or abnormal optic disc appearance had a previously missed diagnosis of secondary elevated ICP, as no details of this case were provided in the study and we do not know whether LP was critical in making or changing the diagnosis. It is also difficult to comment on the study where 4 out of 206 patients who were referred for papilledema were found to have a non-IIH diagnosis⁵ (uveitis, meningitis and leptomenigeal carcinomatosis) as this study has not been published yet; however, uveitis would have been obvious on ophthalmic examination and a patient with meningitis is unlikely to have been systemically well. One patient with leptomenigeal carcinomatosis had reportedly normal neuro-imaging, we do not know whether the MRI was performed with contrast, and whether there were other clues to an alternate diagnosis. It is also unclear whether this patient had a known history of cancer, and whether all of these non-IIH patients fit into typical demographic for IIH.

I will now address the argument that not performing LP can lead to over-diagnosis of IIH in patients who have pseudopapilledema. In almost all cases of pseudopapilledema, an expert neuro-ophthalmic examination and appropriate diagnostic testing such as fundus autofluorescence, orbital ultrasound and enhanced depth imaging ocular coherence tomography would be sufficient to make a diagnosis; on top of that, it is unlikely that these patients would demonstrate indirect neuro-imaging signs of increased ICP: prevalence of transverse sinus stenosis is almost 100% in patients with IIH and, in contrast, only 2% of patients in one recent study evaluating incidental prevalence of radiological signs of increased ICP and no papilledema on clinical examination had evidence of transverse sinus stenosis^{28,29}. Thus, presence of transverse sinus stenosis is a very sensitive indirect radiological sign indicating likely presence of increased ICP and can serve as a substitute for OP on LP in these cases. Finally, patients with pseudopapilledema most of the time have normal or near-normal visual function and should not be treated aggressively even if suspected of having IIH. If they fit into a typical demographic for IIH, have normal visual function and are thus given a recommendation for weight loss, this would not have any harmful effect.

In summary, if the recommendation of avoiding LP is applied to the selected group of patients detailed below, LP can be safely avoided without the risk of missing an alternate diagnosis that would make a difference in the treatment. This select group of patients should: 1) be women of reproductive age with increased body-mass index; 2) have no red flag symptoms and be systemically well; 3) have normal or near-normal visual function (normal central acuity and mean deviation less than 7 dB on Humphrey visual fields; 4) have an MRI and MRV of the brain and orbits with contrast that is unremarkable other than presence of indirect imaging signs of increased ICP; and, finally, 5) be under the care of neuro-ophthalmologist experienced in diagnosing and treating patients with IIH.

Conclusion- Drs. Van Stavern and Lee:

The diagnostic criteria of IIH have been modified several times in the past, in response to changes in technology and our understanding of the disease (e.g., MR venography). However, lumbar puncture has been considered mandatory in each revision of the modified Dandy criteria for IIH. It is conceivable that in very select patients, with classic clinical and neuroimaging features, LP could be deferred unless the clinical course deviated from what is expected for IIH. The downside is the risk of over-diagnosing and over-treating patients without IIH, and missing and delaying diagnosis of patients with alternate, potentially more serious conditions. Further well designed, prospective longitudinal studies would help provide better guidance.

In my own practice (AGL) and as described by my esteemed colleagues, I have patients who did not meet the modified Dandy criteria for IIH. Some patients could not have an MRI (e.g., pacemaker or other ferromagnetic contraindication or severe obesity), while other patients could not or would not have a lumbar puncture. These patients I have documented as having “presumed IIH” in contrast to other patients who I can confidently document as having “IIH by modified Dandy criteria”. Both Dr. Van Stavern and I agree that the current state of literature is insufficient to provide a definitive evidence-based answer to

this controversy and it is our intention to generate discussion rather than to provide a firm opinion on the subject.

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Table 1:

Risks and Benefits of doing or not doing a lumbar puncture in suspected cases of IIH

	Risks	Benefits
Perform LP	<ul style="list-style-type: none"> • Procedure complications • False positive/negative results 	<ul style="list-style-type: none"> • Diagnostic confirmation • Short term treatment of ICP
Defer LP	<ul style="list-style-type: none"> • Failure to diagnose normal ICP (necessitating evaluation for other causes of symptoms) • Failure to diagnose secondary cause of high ICP • Inappropriate medical or surgical treatment due to incorrect diagnosis 	<ul style="list-style-type: none"> • No procedure complications • Money saved • Resources saved

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