

## The review of neuroimaging findings of 139 patients with idiopathic intracranial hypertension: A clinical retrospective experience

Neuroimaging findings of idiopathic hypertension

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### Abstract

**Aim:** Idiopathic intracranial hypertension (IIH), or pseudotumor cerebri is a challenging condition with raised intracranial pressure (ICP) in the absence of identifiable cause. Typical clinical manifestations can be a headache, tinnitus, papilledema, and decreased vision. The diagnosis is established by the Modified Dandy Criteria. Several brain Magnetic Resonance Imaging (MRI) findings have been associated with the diagnosis of IIH. The aim of this study is to determine the characteristics of IIH and establish whether there are characteristic appearances on MRI and MRV that are being routinely overlooked in our clinical practice. **Material and Method:** All patients with a diagnosis of IIH between January 2010 and January 2018 at Bakırköy Mental Health Education and Training Hospital of Neurology, Neurosurgery, and Psychiatry were enrolled. The diagnosis of IIH was established according to the Modified Dandy Criteria (Table 1). Only patients who had available images of MRV examination and/or MR imaging examination during the period of review were included. Electronic medical records were reviewed for clinical parameters such as symptom profile, age, gender, opening pressure at lumbar puncture and neuroimaging. Each case was reviewed independently by a neuroradiologist and a neurologist. **Results:** One hundred thirty-nine patients were included in the study. The patient group consisted of 114 females and 25 males, ages 17-72 years ( $38.13 \pm 11.51$ ). The most common symptom was a headache, which was noted in 84 %. Papilledema was found in 118 patients (86.33 %). In the neurological examination, 7 patients had abducens nerve palsy (Table 2). Opening pressure on lumbar puncture was available in the hospital chart for 130 of the 139 IIH patients (93.5 %). Opening pressure ranged from 160 to 800 mm H<sub>2</sub>O (mean: 365 mm H<sub>2</sub>O; 1 standard deviation=130 mm H<sub>2</sub>O). One hundred twenty-two of 139 patients had abnormal opening pressure. Eight of the 139 patients had normal opening pressure (<250 mm H<sub>2</sub>O). The MRI was normal in 66 patients (47.5 %). The MRI disclosed flattening of the posterior eyeballs and vertical tortuosity of the orbital optic nerve in 6.5 %, optic nerve hyperintensity in 2.2 %, empty sella in 24.5 %, Chiari malformation in 4.3 %, and optic nerve hyperintensity together with empty sella in 9.4 % of patients (Table 3). There was no evidence of sinus thrombosis on the static images of the patients. Unilateral transverse sinus hypoplasia was perceived in 19.4 % of patients. Bilateral TS stenosis was perceived categorically in 20.9 % of patients with IIH. **Discussion:** The study detects that empty sella, optic nerve enhancement on MRI and bilateral transverse sinus stenosis on MRV are mostly seen neuroimaging findings in IIH patients. The presence of Chiari Malformation on MRI is also the supportive clue of IIH in the absence of sinus thrombosis. **Conclusion:** MRI is an important imaging technic to support the diagnosis of IIH. With further investigations, MRI can be enough to diagnose the disease in the future and it will also provide cost-effectiveness.

### Keywords

Idiopathic Intracranial Hypertension; Magnetic Resonance Imaging; Magnetic Resonance Venography

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## Introduction

Idiopathic intracranial hypertension (IIH), or pseudotumor cerebri is a challenging condition with raised intracranial pressure (ICP) in the absence of identifiable cause [1]. Typical clinical manifestations can be intermittent and include headaches, transient visual obscurations, pulse-synchronous tinnitus, papilledema, and decreased vision or visual field. The diagnosis is established by the Modified Dandy Criteria, which, in essence, means an opening pressure at lumbar puncture of >250 mm of water in adults, with no definable etiology [2]. Several brain Magnetic Resonance Imaging (MRI) findings have been associated with the diagnosis of IIH. These include an appearance of an empty sella (>50% vacancy of the sella with a concave upper surface of the pituitary gland), optic nerve sheath dilation, vascular distension and protrusion of the optic papillae with advanced papilledema, “slit ventricles,” and a bright spot at the optic nerve head on diffusion-weighted imaging. When a Magnetic Resonance Venography (MRV) sequence is available, bilateral transverse sinus (TS) stenosis of >50% degree is seen in 93% of the patients with IIH, suggesting that this is the most sensitive imaging characteristic of this condition [3,4].

This study aims to determine the characteristics of IIH and establish whether there are characteristic appearances on MRI and MRV that are being routinely overlooked in our clinical practice.

## Material and Method

All patients coded with a diagnosis of IIH between January 2010 and January 2018 at Bakırköy Mental Health Education and Training Hospital of Neurology, Neurosurgery, and Psychiatry were identified. Patients were diagnosed as having IIH if they had presented with a syndrome of raised ICP usually manifest

by a headache and/or visual disturbance, without ventricular enlargement or an intracranial mass on imaging, with no evidence of venous sinus thrombosis, and with normal cerebrospinal fluid (CSF) constituents.

We conducted a retrospective study with institutional review board approval of images acquired during an 8-year period (2010–2018) in 134 adult patients in whom the diagnosis of IIH was established according to the Modified Dandy Criteria. Only patients who had available images of a gadolinium-bolused MRV examination and/or a pre-/postgadolinium MR imaging examination including at least a 3D T1 postcontrast sequence during the period of review were included. Electronic medical records were reviewed for clinical parameters such as symptom profile, age, gender, opening pressure at lumbar puncture and neuroimaging.

MR and MRV images were reviewed independently of one another. Each case was reviewed independently by a neuroradiologist and a neurologist.

## Results

One hundred and thirty-nine patients were included in the study. The patient group consisted of 114 females and 25 males, ages 17–72 years ( $38.13 \pm 11.51$ ).

The most common symptom in our patients was a headache, which was noted in 84%. The clinical symptoms of our patients are shown in Table 2. Papilledema was found in 118 patients (86.33%). In the neurological examination, 7 patients had abducens nerve palsy.

Opening pressure on lumbar puncture was available in the hospital chart for 130 of the 139 IIH patients (93.5%). Opening pressure ranged from 160 to 800 mm H<sub>2</sub>O (mean: 365 mm H<sub>2</sub>O; 1 standard deviation=130 mm H<sub>2</sub>O). One hundred twenty-two of 139 patients had abnormal opening pressure. Eight of the 139 patients had normal opening pressure (<250 mm H<sub>2</sub>O) as they were on diuretic therapy. Lumbar puncture could not be applied to nine patients because of obesity or no permission but they all had papilledema and fulfilled the neuroimaging criteria.

One hundred thirty-two patients had MRI, 115 patients had MRV imaging. One hundred seven patients had both MR and MRV imaging. The MRI was normal in 66 patients (47.5%). The MRI disclosed flattening of the posterior eyeballs and vertical tortuosity of the orbital optic nerve in 6.5%, optic nerve hyperintensity in 2.2%, empty sella in 24.5%, Chiari malformation in 4.3%, and optic nerve hyperintensity together with empty sella in 9.4% of patients. Seven patients did not have an MRI (Table 3).

There was no evidence of sinus thrombosis on the static images of the patients. The MRV findings were normal in 42.4% of the patients. Unilateral transverse sinus hypoplasia was perceived in 19.4% of patients. Bilateral TS stenosis was perceived categorically in 20.9% of patients with IIH. The nature of the TS stenosis was often a collapse or flattening of the expected contours of the dural sinus (Table 3).

Table 1. The Modified Dandy Criteria

1. Required for diagnosis of pseudotumor cerebri syndrome
A. Papilledema
B. Normal neurologic examination except for cranial nerve abnormalities
C. Neuroimaging: Normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and MRI, with and without gadolinium, and magnetic resonance venography for others; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used
D. Normal CSF composition
E. Elevated lumbar puncture opening pressure ( $\geq 250$ mm CSF in adults and $\geq 280$ mm CSF in children [ $250$ mm CSF if the child is not sedated and not obese]) in a properly performed lumbar puncture
2. Diagnosis of pseudotumor cerebri syndrome without papilloedema
In the absence of papilledema, a diagnosis of pseudotumor cerebri syndrome can be made if B–E from above are satisfied, and in addition, the patient has a unilateral or bilateral abducens nerve palsy.
In the absence of papilledema or sixth nerve palsy, a diagnosis of pseudotumor cerebri syndrome can be suggested but not made if B–E from above are satisfied, and in addition at least three of the following neuroimaging criteria are satisfied:
i. Empty sella
ii. Flattening of the posterior aspect of the globe
iii. Distention of the perioptic subarachnoid space with or without a tortuous optic nerve
iv. Transverse venous sinus stenosis

A diagnosis of pseudotumor cerebri syndrome is definite if the patient fulfills criteria A–E. The diagnosis is considered probable if criteria A–D are met but the measured CSF pressure is lower than specified for a definite diagnosis. CSF, cerebrospinal fluid; CT, computerized tomography; MRI, magnetic resonance imaging.

Table 2. Clinical Symptoms of the Patients

	Number (n)	%
Headache	117	84
Visual disturbance	74	53
Dizziness	3	0.2
Tinnitus	2	0.1

Table 3. Neuroimaging Findings of the Patients

	Number (n)	%
MRI		
Normal	66	47.5
Empty sella	47	34
Flattening of the posterior eyeballs/increased tortuosity of ON	9	6.5
Enhancement of ON	3	2.2
Chiary malformation	6	4.3
MRV		
Normal	59	42.4
Unilateral TS stenosis/hypoplasia	27	19.4
Bilateral TS stenosis	29	20.9

MRI: Magnetic Resonance Imaging, MRV: Magnetik Resonance Venography, ON: Optic nerve, TS: Transverse sinus

## Discussion

IIH is a clinical entity with a myriad of known and putative etiologies. The absence of a clear identifiable etiology for a clinical syndrome characterized by elevated ICP exists in nearly 90 % of cases, and this ambiguity inevitably has led to the replacement of the misnomer “benign” intracranial hypertension with IIH in light of the incidence of vision loss resulting from this condition [5].

IIH classically presents with a headache and, frequently, vision changes mostly in women. Headaches occur in nearly all (90%–94%) patients with IIH—they are characteristically pressure like, throbbing, and usually unremitting and occur with retro-ocular pain and may be accompanied by nausea. Vision loss is the most feared sequela of IIH, but most vision loss in this syndrome is transient in nature and occurs in approximately 68%–85% of patients. [6,7,8]. In our study, headache was also the most common symptom with the frequency of 84 % and the second symptom was the vision disturbance explained by the patients as blurred vision or diplopia. Pulse-synchronous tinnitus and dizziness were rarely reported symptoms by our patients. Although very common, tinnitus is often not reported by the patients unless specifically queried about it. More severe and more disturbing symptoms were more frequently encountered in the study like visual problems and headache.

Papilledema, or swelling of the optic nerves, is often considered a hallmark sign of IIH [9]. The ophthalmoscopic examination of most patients with increased ICP reveals papilledema as in our study, affecting 94 % of our patients. The absence of papilledema has been reported in many populations of patients with IIH, but its absence may be more suggestive of an alternative etiology for a headache and vision loss [6]. Cranial nerve palsies, usually of the abducens nerve (CN VI), may occur as well in many as 10 %–20 % of patients [10]. Rarely, facial nerve (CN VII) palsies may be associated with IIH; all of these CN palsies are thought to occur as a result of direct compression due to

elevated ICP (8). In our study, abducens nerve palsy was rarer compared to the literature rates (0.5 %) and there was no other cranial nerve palsy.

Another important clinical finding for IIH is the opening pressure on lumbar puncture. However, it may be normal or only mildly elevated above 200 mm H<sub>2</sub>O at a single time point due to a possible fluctuations. In our study, opening pressure ranged from 160 to 800 mm H<sub>2</sub>O (mean: 365 mm H<sub>2</sub>O; 1 standard deviation=130 mm H<sub>2</sub>O). One hundred twenty-two of 139 patients ( 93.5 %) had abnormal opening pressure. Opening pressure values above 250 mm H<sub>2</sub>O are considered abnormal [2]. We found opening pressure of the cerebrospinal liquid in normal ranges in 8 patients. We explained this result either by the fluctuation of the liquid or the treatment of diuretics. Also, it is only a point measurement and with high diurnal ICP variability repeated pressure measurements may be required in patients presenting with an atypical phenotype or with only marginally increased opening pressure. We could not perform lumbar puncture to nine patients as some of them did not give permission to the procedure and some of them had obesity. However, they all had papilledema and fulfilled the neuroimaging criteria of IIH.

Several formalized criteria for IIH exist in the literature and are subject to extensive debate. The Modified Dandy Criteria first incorporated the use of CT in the diagnosis of IIH, primarily as a means of excluding occult causes of IIH previously missed in the era before diagnostic imaging. Friedman and Jacobson updated these criteria (Table 1) to reflect the advances of MRI and the characterization of other etiologies of IIH such as venous thrombosis [11,12]. So the next step is neuroimaging, preferably MRI scan of the brain. Supplemental MRV to rule out sinus venous occlusions is essential, since sinus venous occlusions may manifest clinically as IIH syndrome in more than one-third of the patients [13]. According to the criteria in the absence of papilledema, MRI is essential to show at least three of the following neuroimaging criteria: empty sella, flattening of the posterior aspect of the globe, distention of the perioptic subarachnoid space with or without a tortuous optic nerve, and transverse venous sinus stenosis. In our study, MRI could be performed to 132 patients and 115 patients were carried out MRV. The MRI was normal in 66 patients ( 47.5 %). Empty sella was the most common finding (37.5 %) in our study. The “empty sella” sign is associated with the longstanding effects of increased ICP and is thought to result from a downward herniation of an arachnocele through the diaphragma sellae [14,15]. Its presence also supports the diagnosis when the optic nerve appearance of papilledema is equivocal or the opening pressure is within the normal range or mildly elevated. In our study, optic nerve abnormalities like hypertensity, enhancement of the nerve and nerve tortuosity, were also frequently reported findings ( 25 %). Optic nerve tortuosity has been associated with increased ICP; the distal and proximal points of fixation of the optic nerve enable it to kink freely in its course to the globe on the protrusion of the intracranial contents under the pressure [15]. Enhancement of the optic nerve is also thought to be reflective of the same pathology leading to papilledema [16]. In our study, Chiari malformation (CM) was found in 6 patients with a frequency of 4.3 % which is not apart of the IIH criteria. There are significant similarities between the CM and

IIH. They include similar demographics, clinical presentation, and response to treatment. There is an eightfold increase in the incidence of significant tonsillar herniation in patients with IIH. The physiopathology is that of cranioccephalic disproportion, a disproportion between the skull and the brain, due to a small skull or posterior fossa and/or an engorged brain. This will occasionally lead to tonsillar ectopia. The cranioccephalic disproportion will alter the brain compliance and lead to the symptomatology [17]. In the literature, Banik et al. reported that 24% of the patients with IIH had tonsillar ectopia [18]. The relationship between these two diagnoses has been established but is poorly understood. It is possible that elevated intracranial pressure in IIH may cause cerebellar tonsils to herniate through the foramen magnum, manifesting imaging criteria of Chiari I. Alternatively, it is possible that patients with Chiari I have abnormal CSF dynamics, which predispose to elevated ICP and IIH [18]. In the study, we found no sinus stenosis in patients with CM, and we thought that this condition was the result of IIH. So in the absence of sinus stenosis, the presence of CM on MRI can also support the diagnosis of IIH and it can be added as the fourth neuroimaging criteria in the absence of papilledema of the Modified Dandy Criteria.

Previously, it was thought that MRV should be reserved for use in atypical (eg, male, normal weight) patients to rule out sinovenous thrombosis [19]. Higgins et al. identified bilateral sinus flow gaps in 65% of patients with IIH [4]. Also, Farb et al. reported that stenosis of the transverse sinuses on gadolinium-bolused MRV is detectable in 94% of patients with IIH, making this a more sensitive diagnostic indicator in adults than previously established MRI signs [3].

Supplemental MRV to rule out sinus venous occlusions is essential, since sinus venous occlusions may manifest clinically as IIH in more than one-third of patients [13]. Bilateral transverse sinus stenosis, but not occlusions, are frequently reported in IIH patients [20,3,4]. Still, it remains unclear whether this plays a role in the pathogenesis of IIH or just serves as a radiological marker of raised ICP. In our study, the MRV findings were normal in 42.4 % of the patients and bilateral TS stenosis was perceived categorically in 20.9 % of patients with IIH.

In conclusion, our retrospective study is based on the neuroimaging findings of IIH seen on MRI and/or MRV. For the diagnosis of IIH MRI is needed to rule out secondary causes and needed in the absence of papilledema to support the diagnosis. As our study also detected, empty cella, optic nerve enhancement on MRI and bilateral transverse sinus stenosis on MRV are mostly seen neuroimaging findings. Our study also demonstrated the presence of CM can also be a supportive clue of IIH in the absence of sinus thrombosis. With further investigations and additive neuroimaging findings, MRI may be enough to diagnose the disease in the future and it will also provide cost-effective-ness.

#### Scientific Responsibility Statement

*The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.*

#### Animal and human rights statement

*All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.*

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#### Conflict of interest

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