
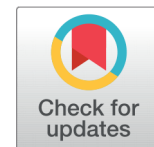


# CAN 'SOVAC' -A SET OF BRAIN MAGNETIC RESONANCE IMAGING FINDINGS-DETECT IDIOPATHIC INTRACRANIAL HYPERTENSION?



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## ABSTRACT

### Objective

Intracranial hypertension is a condition of raised intracranial cerebral spinal fluid pressure and can be primary (idiopathic) or secondary (due to an underlying cause). If left untreated, it can lead to problems like severe headaches and permanent loss of vision. Therefore, we sought to determine the diagnostic ability of brain magnetic resonance imaging in the detection of idiopathic intracranial hypertension by considering certain findings under the SOVAC approach.

### Methods

This descriptive observational study was conducted in the radiology department at King Fahad Military hospital in Dhahran from January 2018 to January 2021. Evaluating MR imaging data of all adult patients (n=107) with clinical presentations of headaches. Patients with space-occupying lesions, obstructive hydrocephalus, and cerebral venous sinus thrombosis were excluded.

Brain MRI findings of (i) an empty sella, (ii) tortuosity of optic nerves with perioptic CSF prominence, (iii) non-dilated lateral ventricles, (iv) acquired cerebellar ectopia, and (v) cerebral venous sinus stenosis was documented based on our 'SOVAC' (Sella, Optic nerve, Ventricular, Acquired cerebellar ectopia, Cerebral venous sinus stenosis) approach. The presence of all first four findings was considered positive for IIH. Imaging was reviewed by two neuroradiologists and consensus reporting was made. Findings of IIH were confirmed by the presence of high lumbar opening cerebrospinal fluid (CSF) pressure or symptomatic response towards specific medical treatment. Outcomes of brain MRIs were presented on a 2x2 contingency table.

### Results

Sensitivity and specificity of routine brain MRI findings in detection of IIH were calculated as 76.4% [95% confidence interval (CI), 64.6-85.9%], and 84.6% (95% CI, 69.4-94.1%) respectively.

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## Conclusion

Brain MRI findings under the 'SOVAC' approach can predict Intracranial hypertension with greater sensitivity and specificity.

### المخلص

#### الهدف

ارتفاع ضغط الدم داخل الجمجمة هو حالة من ارتفاع ضغط السائل الدماغي النخاعي داخل الجمجمة ويمكن أن يكون أولياً (مجهول السبب) أو ثانوياً (بمسبب أساسي). عند تركه دون علاج يمكن أن يؤدي إلى مشاكل مثل الصداع الشديد والفقان الدائم للبصر. لذلك سعينا إلى تحديد القدرة التشخيصية للتصوير بالرنين المغناطيسي للدماغ في الكشف عن ارتفاع ضغط الدم داخل الجمجمة مجهول السبب من خلال النظر في بعض النتائج في باستخدام طريقة سوفاك.

#### الطريقة

أجريت هذه الدراسة الوصفية في قسم الأشعة بمستشفى الملك فهد العسكري في الظهران من يناير ٢٠١٨ إلى يناير ٢٠٢١. تم تقييم بيانات التصوير بالرنين المغناطيسي لجميع المرضى البالغين (العدد = ١٠٧) الذين يعانون من الصداع خلال الكشف السريري. تم استبعاد المرضى الذين يعانون من الأورام واستسقاء الرأس الانسدادي وتجلط الجيوب الوريدية للدماغ. تم توثيق نتائج التصوير بالرنين المغناطيسي للدماغ بطريقة سوفاك بالنسبة للتالي: (١) منطقة السرج التركي، (٢) تعرج الأعصاب البصرية مع بروز السائل الدماغي الشوكي حول العين، (٣) البطنات الجانبية غير المتوسعة، (٤) الانتباز المخيخي المكتسب (٥) تضيق الجيوب الوريدية. اعتبر وجود جميع النتائج الأربعة الأولى إيجابياً لارتفاع ضغط الدم داخل الجمجمة. تمت مراجعة التصوير من قبل اثنين من أخصائيي الأشعة العصبية وتم تقديم تقرير نهائي بالإجماع بينهما. تم تأكيد نتائج ارتفاع ضغط الدم داخل الجمجمة من خلال وجود ارتفاع ضغط السائل النخاعي من خلال الفتحة القطنية أو من خلال استجابة الأعراض نحو علاج طبي محدد. تم تقديم نتائج التصوير بالرنين المغناطيسي للدماغ في على شكل جدول ثنائي الأضلاع.

#### النتائج

تم حساب حساسية المعدل الموجب الحقيقي ونوعية المعدل السالب الحقيقي لنهج سوفاك في الكشف عن ارتفاع ضغط الدم داخل الجمجمة على أنها ٧٦.٤٪ (مستوى ثقة بين ٦٤.٦ - ٨٥.٩) و ٨٤.٦٪ (مستوى ثقة بين ٦٩.٤ و ٩٤.١) على التوالي.

#### الخاتمة

يمكن لتصوير الرنين المغناطيسي للدماغ بطريقة سوفاك أن يتنبأ بارتفاع ضغط الدم داخل الجمجمة بدقة ونوعية أكبر

**Keywords:** Intracranial hypertension, SOVAC, cerebral spinal fluid, magnetic resonance imaging

## 1. INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a clinical syndrome with signs and symptoms of raised intracranial pressure (ICP) without any evident cause [1]. Mechanisms like increased CSF production, decreased CSF absorption, increased intravascular volume, and increased intracranial venous pressures have been proposed for the pathogenesis of this condition [2, 3]The usual presentation is with headaches, visual problems (photop-

sia, diplopia, visual loss), tinnitus, and eye pain. Papilledema may not always be present and can be unilateral or bilateral [4, 5]. Computed tomography (CT) and magnetic resonance imaging (MRI) of the brain help to exclude brain tumors, dural sinus thrombosis, and hydrocephalus [6]. A normal CSF fluid analysis with raised lumbar puncture opening pressure aid in establishing a diagnosis [7, 8].

In the absence of any cause of ICP, certain brain imaging features support the diagnosis of IIH [9]. MRI is superior to CT in demonstrating those features with its high inherent contrast resolution. Features pertaining to optic nerves such as; a) prominent subarachnoid space around the optic nerve, b) vertical or coronal tortuosity of the optic nerve, c) flattening of the posterior sclera, d) intraocular protrusion of optic nerve head, and e) enlarged intraocular protrusion of optic nerve head, besides enlarged arachnoid CSF outpouchings like empty sella, enlarged Meckel cave, prominent arachnoid pits or small meningoceles within the temporal bone, and enlarged CSF around oculomotor nerve within the cavernous sinus, venous sinus stenosis particularly lateral segment of the transverse sinus, slit-like ventricles and acquired tonsillar ectopia have all been suggested to indicate IIH [9–11].

Although few studies have discussed management options and prognosis of IIH in Saudi Arabia [12, 13], no recent or specific studies are available in Kingdom to emphasize the potential diagnostic capability of routine brain MRI based using a set criterion or approach towards detection of IIH, that remains a diagnostic challenge for the neuroradiologists [14]. Therefore, we aim to estimate the diagnostic capability of brain MRI to detect IIH.

## **2. METHOD**

This descriptive observational study was conducted in the radiology department at our hospital over three years, from Jan. 2018 to Jan. 2021.

### **2.1 Subjects**

All adult patients for whom routine brain MRIs were performed with clinical indications of headaches with or without visual symptoms were retrospectively reviewed. Pregnant patients, those with space-occupying lesions, trauma patients, obstructive hydrocephalus, and known cerebral venous sinus thrombosis were excluded.

## **2.2 Ethical approval**

The research protocol was approved by the Hospital Research and Ethics Committee. The study was conducted by the Helsinki Declaration.

## **2.3 Data acquisition**

Clinical information was obtained from patients' medical records via the hospital information system (HIS), while MR imaging findings were reviewed through the radiology information system/ picture archiving and communication system (RIS/ PACS). All clinical and radiological data were kept strictly confidential.

## **2.4 Clinical measures**

The age and gender of patients were taken. Body mass index (BMI) was expressed as within normal (18.5- 24.9), over-weight (25.0- 29.0), and obese (30.0 and above).

## **2.5 Image acquisition**

All MRI brain studies were performed on a 1.5 T machine (GE Machine). Routine brain imaging included T1W (T1-Weighted) axial (Ax.) and sagittal (Sag.) Fast Spin Echo (FSE) sequences, Ax. T2WI (T2-Weighted Imaging), Ax. FLAIR (Fluid Attenuation and Inversion Recovery), DWI (Diffusion-Weighted Imaging) and ADC (Apparent Diffusion Coefficient), and SWAN (Susceptibility Weighted Imaging) sequences. Thin slice sagittal 3D CUBE (GE) T2WI, 3 mm Ax. And Cor. T2-W steady-state (FIESTA) was also acquired for orbits and skull base (in cases of visual symptoms). MR venographies (either contrast-enhanced or time of flight) were reviewed if available. Image parameters were not standardized.

## **2.6 Image interpretation**

The presence of all brain MRI findings of (i) an empty sella (with or without sellar enlargement), (ii) tortuosity of optic nerves (with or without perioptic cerebrospinal fluid/ CSF prominence), (iii) non-dilated (or slit-like) ventricles, (iv) cerebellar ectopia, and (v) transverse cerebral sinus stenosis was considered positive for IIH. Empty sella was labeled when the pituitary gland was found shrunken or flattened, filling the sella turcica with cerebrospinal fluid instead of the normal pituitary. More than 50% sellar vacancy with the concave surface of the pituitary gland was taken as a quantitative measure to label empty sella. Optic nerve tortuosity was defined as lack of optic nerve congruity in >1 coronal section. Prominent CSF space along the optic nerve sheaths was taken if the thickness of

surrounding CSF exceeded 1 mm. In cases where MR venographies were acquired, more than 50% luminal narrowing of cerebral venous sinuses was taken as stenosis. Cerebellar tonsillar ectopia was labeled if the cerebellar tonsillar position was found under 5 mm below the base of the skull. The absence of all or any one of these findings was taken as negative for IIH. Final documentation of findings was mutually agreed upon between the two radiologists.

## 2.7 Outcome analysis

Opening CSF pressures of more than 250 mm H<sub>2</sub>O (by estimating the value of pressure in mm Hg x 13.56) under lumbar punctures were taken as high intracranial pressure. Clinical diagnoses of IIH were established (that partly include Dandy criteria) on documentation of signs of raised ICP (papilledema), MRI findings (negative for structural causes or hydrocephalus), opening CSF pressures of more than 250 mm H<sub>2</sub>O, and relief of symptoms by medical management. Outcomes of brain MRI imaging were presented as true positive (TP; brain MRI correctly identified IIH), true negative (TN; brain MRI correctly excluded IIH), false positive (FP; brain MRI incorrectly identified IIH), or false negative (FN; brain MRI incorrectly excluded IIH). A fourfold contingency table represented the outcomes.

## 2.8 Statistical analysis

Data was collected and analyzed using Statistical Package for Social Sciences (SPSS Statistics for Windows, version 22; IBM Corp., Armonk, NY, USA). The sensitivity, specificity, and diagnostic accuracy of brain MRI were determined.

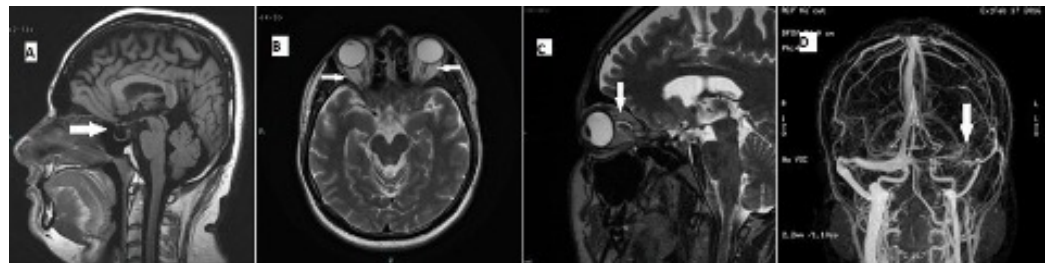
## 3. RESULTS

Of 107 patients, majority were females (n= 88, 82.2%), and the mean age was 44.78 years (range 29- 68 years, standard deviation/ SD- 9.1). Papilledema was seen positive in 52 out of 68 positive patients of IIH. Sensitivity, and specificity of routine brain MRI in detection of IIH were calculated as 76.4% [95% confidence interval (CI), 64.62-85.91%], and 84.6% (95% CI, 69.47-94.14%) respectively (P-value, 0.0005) [Table 1].

Out of 52 true positive cases of IIH, the majority (43/52; 82%) were either overweight (n=29) or obese (n=14), while the majority of true negative cases of IIH (26/33; 78%) were within normal limits of their body mass indices. MR venographies were found in 47 patients (n= 47), and 35 showed either unilateral (22/35) or bilateral (13/35) transverse cerebral venous sinus stenosis. Acquired cerebellar ectopia was seen in 7 patients.

**Table 1** Brain MRI findings and their outcome in the detection of IIH.

MRI Findings	Clinical / CSF pressure		Total
	Positive	Negative	
<b>Positive</b>	52	6	58
<b>Negative</b>	16	33	49
<b>Total</b>	68	39	107



**Figure 1** Selected MRI brain images of a 43-year-old male patient diagnosed with IIH showing A) empty sella on sagittal T1W image, B & C) tortuosity of optic nerve and prominence of perioptic CSF on axial and sagittal T2W image, and D) MRV TOF showing unilateral stenoses of left-sided transverse and sigmoid cerebral venous sinuses.

Opening CSF pressures were measured in 31 patients and found to be high in 23 patients with a mean of 261 mm H<sub>2</sub>O (SD- 4.6 mmH<sub>2</sub>O). For the remaining TP patients for whom opening CSF pressure measurements were not made, clinical improvement with medical treatment was observed. Opening CSF pressures were measured in 31 patients and found to be high in 23 patients with a mean of 261 mm H<sub>2</sub>O (SD- 4.6 mmH<sub>2</sub>O).

#### 4. DISCUSSION

Idiopathic intracranial hypertension (IIH) is a disorder related to raised intracranial pressure with unknown etiology. It is also sometimes called benign intracranial hypertension or pseudotumor cerebri. High pressure around the brain causes symptoms like headaches and vision changes. The role of clinical neuroradiology is to exclude causes of secondary intracranial hypertension (e.g., space-occupying lesions, obstructive hydrocephalus, infection, and cerebral venous sinus thrombosis) and to identify imaging findings that support the diagnosis of IIH [15]. Our study exploited the role of routine brain MRI in the detection of such findings by adopting 'SOVAC' approach. Although some studies have highlighted different sensitivities of various individual imaging findings, we found 'SOVAC' approach a more practical and easy way to identify such condition in a methodical way.

Partially empty sella sign can occur in 70-80% of IIH cases and is produced by downward herniation of arachnocele through the diaphragmatic sellae and results in subsequent flattening or thinning of pituitary gland tissue [5]. This sign may be a normal appearance in the older population and is best depicted on sagittal T1- or T2-weighted images. This was one of the findings in false-positive cases in our study. Optic nerve sheath (ONS) is composed of CSF within perioptic subarachnoid space and dura. Unilateral or bilateral enlargement of ONS diameter (to more than 5 mm) and vertical tortuosity may be seen in 45-65% of IIH cases [10]. Orbital abnormalities seen in IIH may be explained by elevated intracranial and CSF pressure through subarachnoid space surrounding the optic nerves. However, it is worth stating that their appearances do not always correlate with the clinical symptoms of papilledema. We found the presence of optic nerve tortuosity with concomitant prominence of CSF along the optic nerve sheath to be more commonly associated with papilledema rather than only tortuosity without perioptic CSF prominence.

Transverse cerebral venous sinus narrowing (unilateral or bilateral) has also been reported in 65-90% IIH cases, usually involving lateral aspects [16]. Its role in the pathogenesis of IIH due to venous outflow obstruction or its compression secondarily from increased intracranial pressure is still unclear. We did not find MR venographies in all patients as these were not routinely requested for the clinical indications of headaches, but where available were reviewed to collaborate with the outcomes. Most positive IIH cases were having focal or diffuse, unilateral or bilateral stenoses, or atretic transverse or sigmoid sinuses. Slit-like ventricle may also be seen in IIH, though difficult to be objectively evaluated, and be a normal appearance in young subjects. We also considered non-dilated ventricles to be included in slit-like ventricles particularly in the older age group as mild age-related ventricular prominence could be seen due to atrophy. Acquired cerebellar ectopia may be seen in 10-20% of cases, which can be misinterpreted by Chiari 1 malformation [11].

IIH may be a self-limiting disorder, but it can become chronic requiring treatment. Weight loss is recommended for all obese patients. For all patients with severe headaches and papilledema, lumbar puncture is always performed after brain imaging. Medical treatment includes medications that reduce CSF production (e.g., acetazolamide or topiramate). CSF shunting (ventriculoperitoneal or lumboperitoneal) is reserved for the fulminant condition with the rapid visual loss [16]. Optic nerve sheath fenestration may be performed in fulminant cases (not responding to medicines) [17]. Endovascular stenting for transverse sinus stenoses has been considered in refractory cases [18]. The return to normal appearances of the pituitary gland and optic nerves indicates a positive response to therapy or treatment [10]. Other imaging sequences have also been reported in the literature to evaluate IIH. Sarica A et al found a significant diffusion tensor imaging (DTI) alteration in periventricular white matter microstructure of 35 patients with IIH suggesting

tissue compaction with higher CSF pressures [19]. Razek AAKA and colleagues found that susceptibility-weighted imaging (SWI) detected cerebral microbleeds (CMBs) in 16 out of 55 IIH patients especially those with a higher graphic rating scale (GRS) for headache [20].

We did not include the pediatric population in our study. However, Hartmann AJ et al found similar MRI findings as those for adults in the detection of IIH in children while reviewing data of 50 children aged under 17 years of age [21]. Gaier ED and Heidary G found that pediatric patients were more likely to be asymptomatic or present with atypical symptoms posing a diagnostic challenge in some patients [22]. Moreover, they observed no apparent association of IIH with gender or obesity in prepubertal patients. Aylward SC and Way AL found that untreated IIH in children could lead to poor quality of life and morbidity [23]. We did not include pregnant women in our study. However, Kesler A and Kupfermanc M suggested that IIH can occur in any trimester during the pregnancy and found no increase in visual outcome or change in morbidity in such patients [24].

Limitations to our study included its retrospective nature and the associated bias in excluding pregnant women and pediatric population which may show different disease course and pertinent findings. Various other MR imaging findings were not included that could have also suggested IIH like flattening of posterior sclera or globe flattening, intraocular protrusion of optic nerve head and enhancement, enlarged Meckel caves, and prominent arachnoid pits. High-resolution orbital imaging and contrast-enhanced MR venographies were not acquired for all the patients as routine studies were requested by the clinicians, however, authors recommend that these should be incorporated by the radiologists if clinical suspicion of IIH is raised or imaging findings suggestive of IIH on acquired images. Larger scale prospective studies with the inclusion of all these findings and subgroup analysis need to be considered for further validation of results.

## 5. CONCLUSION

IIH is a known cause of headaches. Detection of all three findings of empty sella, tortuosity of optic nerves and non-dilated ventricles on routine brain MRI can predict IIH with greater sensitivity and specificity.

## CONFLICT OF INTEREST

The authors declare that the research was conducted in absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

## ACKNOWLEDGEMENTS

N/A

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