

Imaging and interventions in idiopathic intracranial hypertension: A pictorial essay

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Abstract

Intracranial hypertension is a syndrome of elevated intracranial pressure that can be primary or secondary. The primary form, now termed idiopathic intracranial hypertension (IIH), was in the past a disease of exclusion and imaging played a limited role of excluding organic causes of raised intracranial pressure. However imaging markers have been described with patients with IIH at the orbit, sella and cerebral venous system. We wish to reiterate the characteristic imaging features of this poorly understood disease and also emphasise that stenting of the transverse sinus in select cases of IIH is an efficacious option.

Key words: Cerebrospinal fluid; Cranial venous outflow obstruction; Idiopathic intracranial hypertension; Intracranial pressure; MR venography; Optic nerve sheath; Optic nerve sheath diameter; Pseudotumor cerebri; Short tau inversion recovery; Superior sagittal sinus; Transverse sinuses; Transverse sinus stenosis (TSS)

Introduction

Intracranial hypertension is a syndrome of elevated intracranial pressure that can be primary or secondary. The primary form is characterized by normal CSF composition and no other identifiable cause. It is termed Idiopathic intracranial hypertension (IIH) and has now replaced the older entities *pseudotumor cerebri* or *benign intracranial hypertension*.

In the past, IIH was a diagnosis of exclusion and imaging played a limited role of excluding lesions producing intracranial hypertension, like "obstructive hydrocephalus, tumour, chronic meningitis, arteriovenous fistula, internal

jugular vein stenosis, and dural sinus thrombosis".^[1] Of late, few imaging markers have been described in patients with IIH at orbit, sella and cerebral venous system. Transverse sinus stenosis is now an important treatable entity in select cases of IIH.

Terminology

To understand IIH as an entity, few terminology and issues have evolved and are summarised in Table 1.

Clinical features

The typical IIH patient is an obese woman of childbearing age, with a body mass index >25. The reported incidence of IIH is 19/100,000 in this population.^[2] IIH is clinically characterised by headache, symptoms of increased intracranial pressure, normal cerebrospinal fluid, without ventriculomegaly or mass lesion. Other presentations include retro-orbital pain, pulsatile tinnitus, visual disturbance (acuity and/or field loss) and blindness.

Among the visual disturbances, the most described is papilledema which may be bilateral, asymmetrical, or

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even unilateral; IIH can however occur in the absence of papilledema.^[1] In view of the fact, that few cases of IIH develop blindness due to severe papilledema, the very nature of it being termed a benign entity is under scrutiny.

Treatment options in IIH, aim to reduce the CSF pressure. They include weight reduction, acetazolamide, surgical procedures like CSF shunt insertion, optic nerve sheath fenestration or subtemporal decompression.^[3,4] Lately, stenting of transverse sinus stenosis is emerging as a therapeutic option. There exists a wide variety of conditions and medications which mimic IIH, as well as diseases involving the venous sinuses either primarily or secondarily.^[5] It is imperative to exclude them before labelling a case as idiopathic.

Criteria

Dandy Criteria has been devised for attributing raised intracranial pressure as IIH. It includes neurological symptoms and signs (often non-specific) and measurement of intracranial or lumbar CSF pressure (invasive procedure).^[6] To make the criteria more objective, they have been revised and modified [Table 2]. It is important to note that CT, MRI and MRV are components in this updated version.

Table 1: Timelines in intracranial hypertension^[5,20,21]

| Term/event | Author/Worker | Year |
|--------------------------------------|--------------------------|------|
| Meningitis serosa | Heinrich quincke | 1893 |
| Pseudotumor cerebri | Nonne | 1904 |
| Otitic hydrocephalus | Symonds | 1931 |
| Criteria/diagnostic features | Walter Dandy | 1937 |
| Benign intracranial hypertension | Foley | 1955 |
| Idiopathic intracranial hypertension | Corbett <i>et al.</i> | 1982 |
| Modified dandy criteria | Smith | 1985 |
| Cranial venous outflow obstruction | Karahalios <i>et al.</i> | 1996 |
| Revised modified Dandy criteria | Friedman, Jacobson | 2002 |
| Transverse sinus venous stenting | Higgins, Owler, Cousins | 2002 |

Table 2: Revised modified Dandy criteria for diagnosing IIH^[7,22]

| Criteria | Salient features |
|--|--|
| Symptoms if present reflect raised ICP | Important symptom is headache and/or papilledema |
| Signs attributed to raised ICP | Important sign is papilledema (swelling of optic disc) ^[23] |
| Elevation of CSF opening pressure | Measured in lateral decubitus position > 20 cm H ₂ O in non-obese patients ^[3] > 25 cm H ₂ O in Obese patients ^[3] > 28 cm H ₂ O paediatric patients ^[7,16,22,23] |
| Normal CSF composition | No evidence of meningitis or tumour |
| No underlying structural cause | No hydrocephalus, mass, structural, or vascular lesion on CECT, MRI, MRV |
| No other cause of intracranial hypertension identified | No hypoparathyroidism, Addison disease, Chronic obstructive pulmonary disease or polycystic ovary syndrome ^[5] No medications like tetracycline, steroids, Vitamin A or Amiodarone ^[5] |

MRV: MR venography

Imaging and interventional findings

MRI brain and MR venography are the modalities of choice in diagnosing this condition using imaging.

Sequences

A study is considered complete after contrast enhanced scans of brain, orbit and evaluation with MRV. The sequences among others should include a) T2 TSE axial with fat suppression, 3 mm section thickness, b) STIR coronal, 3 mm, covering orbit and pituitary gland; and c) 3D phase-contrast MRV with velocity encoding at 15 cm/s.

Orbital findings

Fat-saturated T2-weighted images is recommended for analysis of orbits. A typical case variably reveals distension of the perioptic subarachnoid space, which causes enlarged optic nerve sheath [Figure 1], flattening of the posterior sclera [Figure 2], vertical tortuosity and elongation of orbital optic nerves [Figure 3].^[5]

An enlarged optic nerve sheath diame (ONSD), is hypothesised to occur as a result of raised intracranial pressure, since subarachnoid space underneath the optic nerve sheath is in direct anatomical connection with CSF surrounding the brain. Increased intracranial pressure causes direct transmission of pressure resulting indistention of optic nerve sheath (ONS).^[7]

Sellar findings

Partial or empty sella is a well-documented imaging marker of IIH [Figure 4]. It has a sensitivity of 53 to 80% and specificity of 75 to 92%.^[5] An empty sella reflects a chronic increase in intracranial pressure (ICP) and is caused by downward herniation of CSF through the diaphragm sella.

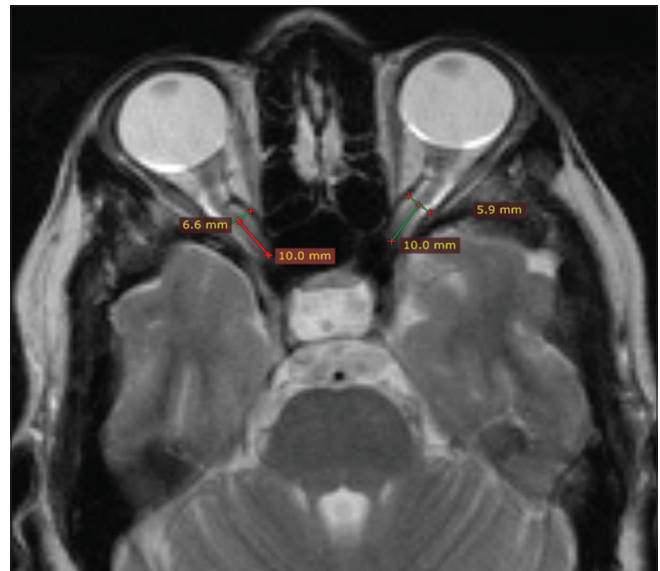


Figure 1: Orbital findings: Axial T2-wtd FS image (4000/100/2) of both optic nerves reveals flattening of posterior sclera and a distended perioptic subarachnoid space. A distension of optic nerve sheath >2 mm is significant. The ONSD is measured 10 mm anterior to optic foramen

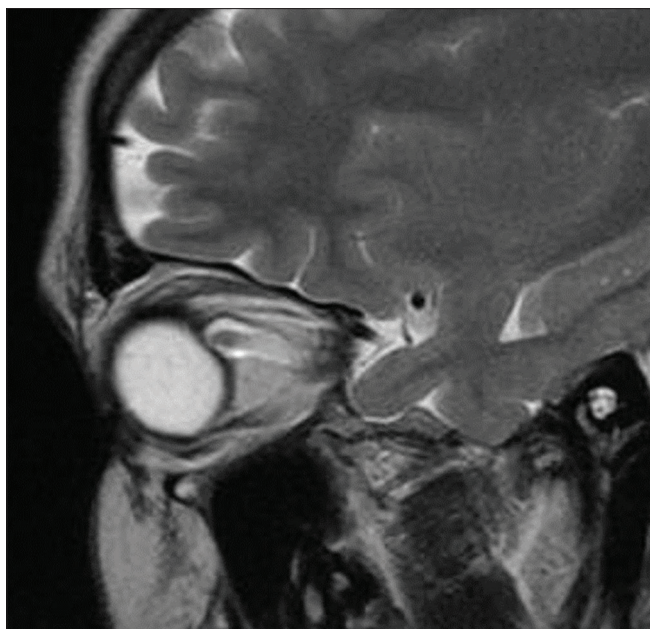


Figure 2: Orbital findings: Sagittal T2-wtd FS image (4000/100/2) of right optic nerve shows flattened optic nerve head (ONH) with protrusion of intra-ocular portion of ONH

Venous sinus morphological findings

Bilateral transverse venous sinus narrowing can be either a cause or an effect of IIH. The causative role is supported by the fact that transverse venous stenting controls IIH in patients with venous stenosis. The notion of it being an effect is reinforced by a lowering of intracranial pressure that results in normalization of venous morphology, suggesting a form of secondary cranial venous outflow obstruction (CVOO).^[2,6]

Venogenic causes of transverse sinus narrowing is seen best on sagittal and axial MR and delineated well on MR venography [Figure 5A and B]. The findings range from bilateral, smooth, short segment areas of focal narrowing to well-defined signal gaps at transverse-sigmoid junctions.^[6,8-12]

The word 'Idiopathic' implies there is no underlying cause on any form of imaging. A knowledge of normal variants is important, especially regarding a superior sagittal sinus that is uniformly narrowed in its anterior third and transverse sinus that is uniformly narrow.^[12] Mimics of narrowing on MRV include non-thrombotic *extrinsic* (venous compression) or *intrinsic* (arachnoid granulations/fibrous septa/intraluminal partition) and thrombotic causes ([cerebral venous thrombosis (CVT)] etc).^[4,13] The tapering of transverse sinuses in IIH is attributed to its compression by raised CSF pressure.

Venous sinus pressure findings

Venous sinus pressure is measured during neurointerventional procedures, by attaching a pressure transducer to the microcatheter [Figure 6A and B]. Manometry features of transverse sinus stenosis is featured

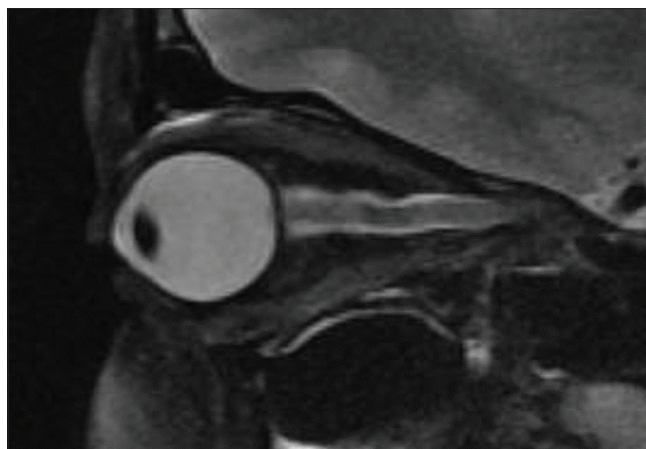


Figure 3: Orbital findings Sagittal Axial T2-wtd FS image (4000/100/2) of right optic nerve shows vertical tortuosity of ONS. The distal and proximal points of optic nerve are fixed. Elongation and kinking in its course due to raised intracranial pressure

by high superior sagittal sinus pressure above and a pressure gradient across the stenosis.^[4]

Under normal conditions "a pressure gradient of only 0-3 mm Hg exists between the superior sagittal sinus and internal jugular vein.^[14] In IIH, a pressure difference of at least 10 mm Hg is highly suggestive of underlying stenosis. The increased pressure difference across the transverse sinuses is hypothesised to be caused by increased resistance from external compression and an increased blood flow.^[11]

Venous stenting

Stent placement at transverse sinus in IIH, was first reported in 2002.^[15] Stent placement is ideally indicated for patients with fixed transverse sinus stenosis having a significant pressure gradient (>8 mm Hg) [Figure 7A-C]. It improves CSF clearance, thereby reducing intracranial (CSF) pressure and papilledema.^[4] One author has recently suggested that long term outcome of venous stenting in young IIH patients is still not yet established, and therefore venous stenting should be limited to "selected patients with bilateral TSS or with a hypoplastic transverse sinus on one side and TSS on the other, and refractory symptoms and signs of increased ICP, who cannot undergo more conventional surgical treatments".^[16]

Stent placement procedures are performed under general anaesthesia because guide-catheter access and dural stretching during stent insertion can cause severe bradycardia.^[4] Commonly, self expanding stents are used. However, in those cases of venous narrowing resulting from raised ICP, endovascular treatment is not considered a therapeutic approach, due to the fact that re-stenosis can occur.^[17]

Finally, a review of literature reveals there are no evidence-based data to guide therapy yet.^[18] Further there is a limited availability of meta-analysis studies with regard to cerebral venous stenting for IIH.^[19]

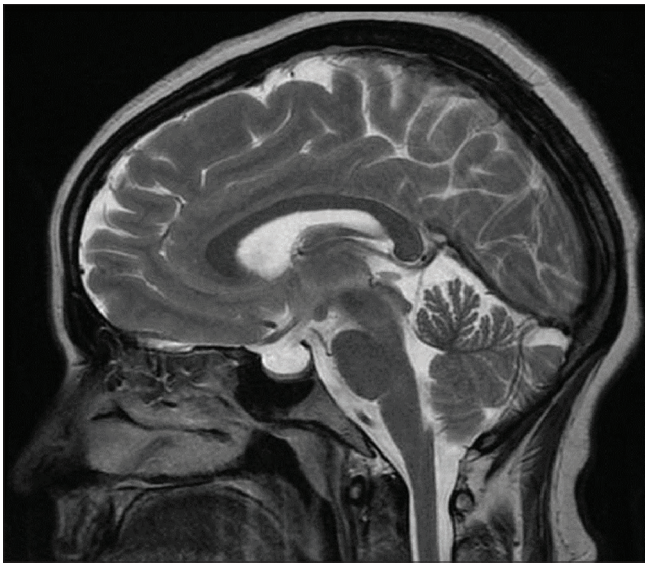


Figure 4: Sellar findings: T2W Sagittal midline image shows a partially empty sella. Mild form or partially empty sella is indicated by incomplete compression of pituitary gland. Severe form is an empty sella with non-visualised pituitary gland

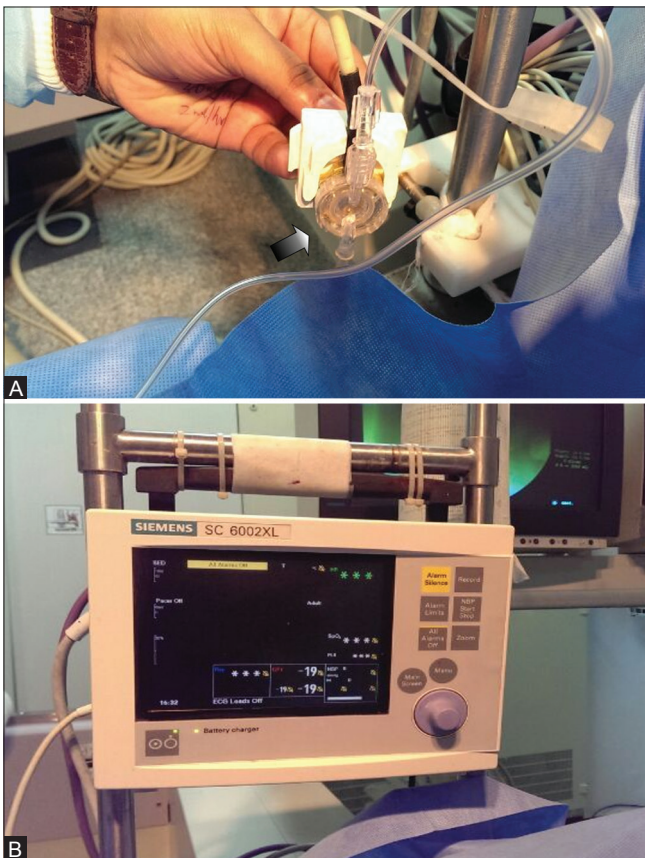


Figure 6 (A and B): (A) Arrow points to the pressure transducer which is connected to the intravenous microcatheter (B) The other end connects to the multifunction monitor which offers equalisation with axillary pressure and reflects the pressures within the sinuses

Table 3 summarises the markers useful to in radiology practice.

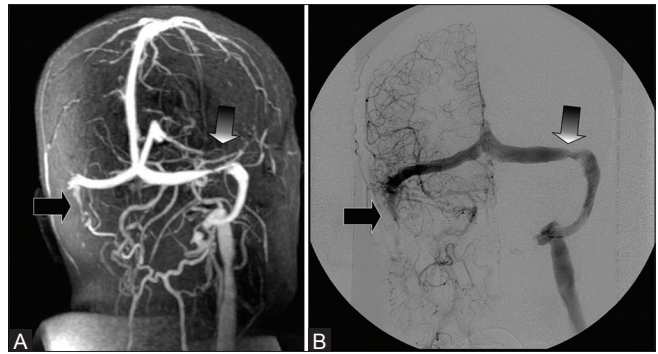


Figure 5 (A and B): (A and B) MRV and venous phase DSA images in a case of right transverse sigmoid dural AV Fistula shows short segment pseudo-stenosis of the left transverse sinus (arrow) due to increased intracranial pressures. Arrowhead points to the occluded right sigmoid sinus

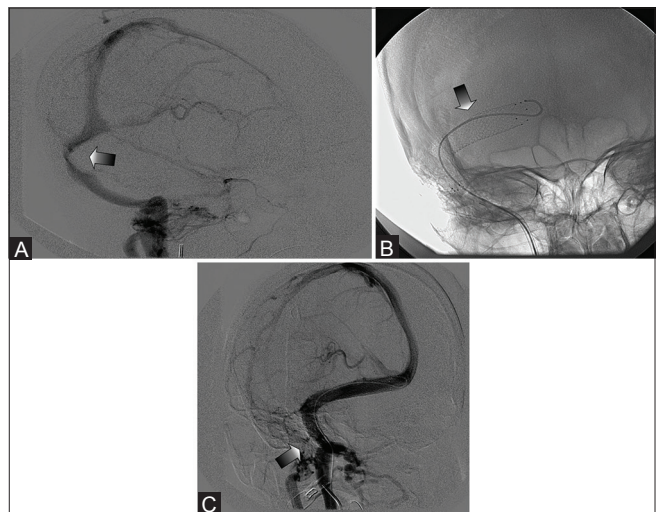


Figure 7 (A-C): (A) Venous phase of ICA angiogram shows a high grade stenosis of the right lateral sinus (arrow). Stenosis are characterised for the following: Intrinsic/Extrinsic; Unilateral/Bilateral and Dominant and/or Hypoplastic Stenosis (B) 8 x 80 mm self-expanding stent deployed across the stenosed segment of the right lateral sinus (C) Post stenting angiogram shows good calibre and filling of the prior stenosed segment. [Image courtesy Professor Dr Uday S Limaye, Consultant Interventional Neuroradiology, Mumbai]

Conclusion

This pictorial essay reviews briefly the terminology and criteria, while illustratively describing the imaging and interventional findings in patients with IIH. Imaging is an integral part of the modified and revised Dandy criteria to diagnose IIH, with imaging markers well established in literature now.

To summarise, MRI and MRV is recommended to analyse orbit, sellar and venographic findings in IIH. Specifically venogenic causes are sought either in symptomatic patients with headache or in asymptomatic patients with incidental findings of orbital hydrops and empty sella. Transverse sinus stenting has emerged as a successful alternative to CSF shunt surgery, particularly in patients with fixed transverse sinus stenosis with a gradient.

Table 3: Markers of IIH in imaging and interventional radiology

| Structure | Imaging finding | Salient features |
|---------------|--|---|
| Orbit | Flattening of posterior sclera | Commonest imaging finding in orbits ^[24] Strongest diagnostic value. Specificity approaches 100% ^[5] |
| | Distension of perioptic subarachnoid space | ONSD measured 3, 6, 10 mm anterior to optic foramina ^[7] Distension of optic nerve sheath (> 2 mm) is significant ^[17] |
| | Enhancement of prelaminar optic nerve | Raised pressure causes breakdown of blood-retinal barrier |
| | Vertical tortuosity of the orbital optic nerve | Tortuosity occurs in vertical or horizontal planes ^[7] Distal and proximal points of optic nerve are fixed; Elongation and kinking in its course due to raised intracranial pressure |
| | Intraocular protrusion of prelaminar optic nerve | Transmission of increased CSF pressure through subarachnoid space of optic nerve sheath ^[7] |
| Sella | Empty sella | Mild form has partially empty sella with compressed pituitary gland. Severe form is empty sella with non-visualised pituitary gland |
| | Type of stenosis | Categorised as Intrinsic or Extrinsic, Unilateral or Bilateral, Dominant and/or Hypoplastic |
| Venous system | Cranial venous outflow obstruction | Graded as Grade 0: Diameter of sinus <50%; Grade 1: CV00 lesion with TS lesions (type I), mid or dorsal SSS (type II), or combination of above 2 (type III). ^[6,25] |
| | Lower mean flow in superior sagittal sinus | Normal pressure gradient 0-3 mm Hg between SSS and IJV Normal mean blood flow in SSS ranges 400 mL/min, ^[11] 418 mL/min, ^[26] 457 mL/min, ^[27] 282 mL/min ^[28] |

ONSD: Optic nerve sheath diameter

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