Neuroimaging of the main Cerebrospinal Fluid Disorders

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**Learning objectives**

- To make an outline of the different cerebrospinal fluid (CSF) disorders and their associated radiological semiology.

- Recognize the signs of hydrocephalus, intracranial hypotension syndrome (IHS) and pseudotumor cerebri syndrome (PTCS).
The dynamics of the CSF have a great importance, among other reasons, due to the impact it has on intracranial pressure. This dynamic depends mainly on the formation, circulation and reabsorption of the CSF.

The alterations produced in any of the elements of the circuit that the LCR performs will cause defined entities that can mostly be identified by means of image tests. The main entities are hydrocephalus, CSF hypotension syndrome and cerebral pseudotumour:

- Hydrocephalus results from the imbalance between the formation (always constant) and drainage (hampered by defects in circulation or absorption) of CSF which leads to a net accumulation of fluid within the ventricles with the consequent increase in intracranial pressure. There are a great variety of etiopathogenic classifications of this entity (see table).
- IHS is caused by cerebrospinal fluid leaking from the subarachnoid space into the epidural space due to a defect in the dura.
- Cerebral pseudotumour syndrome (PTC) consists of a clinical picture of intracranial hypertension (ICH) with no demonstrable cause. It can be really idiopathic or secondary to certain conditions.

Broadly speaking, the clinical characteristics of each entity are:

- CSF hypotension syndrome: it is characterized by the appearance of a holocranial headache with a clear postural relationIHSp (orthostatic headache). The headache can be associated with vertigo, nausea, vomiting, profuse sweating, blurred vision, tinnitus.
- Hydrocephalus: Bifrontal headache with sometimes papilledema and optical atrophy, paresis of VI pair and Parinaud sign (Impaired vertical gaze but intact horizontal gaze), endocrine alterations, hypothalamus-hypophysis affectation, mental alterations, gait ataxia, sphincter decontrol and altered level of consciousness in acute cases.
- Pseudotumor cerebri syndrome: Headache (present in almost all patients), pulsating tinnitus, visual acuity impairment, transient loss of vision, photopsies, alterations of campimetry, paralysis of the cranial nerve VI. The main morbidity factor of this disease is the decrease in visual acuity, which in extreme cases can lead to blindness.

Computerized axial tomography is useful in the urgent diagnosis of hydrocephalus and cerebral hypotension syndrome. However, magnetic resonance imaging is the technique of choice for evaluating disorders of the dynamics of the CSF.
Findings and procedure details

IMAGING FINDINGS IN MAJOR CIRCULATION DISORDERS OF CSF

INTRACRANIAL HYPOTENSION SYNDROME

Although CT may give some clues, the technique of choice for IHS image diagnosis is MRI. However, the findings alone are not sufficient for diagnosis; they must be accompanied by a concordant clinical context.

Many of the radiological signs observed are due to the fact that the decrease in the volume of the CSF (in the presence of closed sutures) generates compensatory mechanisms such as the dilation of venous and arterial structures with the intention of keeping the total encephalic volume constant, according to the Monro-Kellie rule.

• **Sign of venous distension**: characterized by a convex morphology of the lower margin of the transverse sinus, which is usually slightly concave or straight. This sign is easily seen in the middle portion of the transverse sinus in sagittal T1 MRI sequences without intravenous contrast. The sign of venous distension has a sensitivity and specificity close to 94%.

• **Vasodilation of the Galen vein and the bridging veins** due to an increase in pressure in the cavernous sinus

• **Meningeal enhancement**: To compensate for hypotension there is a secondary dilation of the meningeal vessels which results in an increase in contrast uptake.

• **Diffuse paimeningeal thickening and enhancement** Fig. 1 on page 10: This is the classic finding of this entity, it is present in 83% of cases, however, this sign is nonspecific and also occurs in other dural processes. It is produced by the hyperemia of the dura mater as a compensatory mechanism of the low volume of the CSF, which manifests as hyperintensity of dural signal in the enhanced sequences in T2, this usually disappears after recovery of the picture. This sign may be absent when the CSF volume and pressure values are insufficiently low to cause significant venous congestion, so its absence does not exclude the diagnosis of IHS.

• **Subdural effusions, hygromas or subdural haematomas**: The traction caused by the encephalic descent causes the rupture of the small lymphatic vessels and bridge veins located in the thickness of the dura-arachnoid interface and bridge veins causing exit from the CSF to the subdural space and forming hygromas and, rarely, true subdural haematomas. These collections are usually bilateral and located in the parietal convexity. Their diagnosis is difficult because they tend to be thin (typically 2-7 mm) and
easier to recognise in T1 enhanced axial sequences after intravenous contrast administration.

- **Diffuse pituitary enlargement**: caused by dilation of the pituitary veins and portal pituitary system. The convexity of the upper edge of the gland is considered a positive finding, while a flat or concave appearance is considered negative. This sign is unspecific and generally does not correlate well with the size of the gland. On the other hand, the protrusion of the pituitary gland on the Turkish chair seems to have a better specificity and could be considered as an auxiliary sign for the diagnosis of IHS. The pituitary returns to normal size after clinical improvement.

- **Signs of brain descent or sagging brain**: Fig. 2 on page 10 With chronicity there is a decrease in the volume of the CSF causes a descent of the brain through a suction mechanism. This can be represented by:

  1. A location of the cerebellar tonsils below the foramen magno (pseudomalformation of Arnold-Chiari type I)
  2. Decreased subarachnoid space
  3. Decreased ventricular size
  4. Decrease in preponderant cistern size
  5. Decrease in periquiasmatic cisternal size
  6. Slimming of the optic chiasm
  7. Iter descent (The iter is the opening of Silvio's aqueduct and must be identified in middle-agital cuts)

- **Spinal findings**: Dural spinal reinforcement, dilation of epidural veins (flow void), meningeal diverticula, liquid collection in soft tissues after C1-C2 or epidural or subdural collections. The epidural space represents a gutter within which the CSF rises or falls, so that the location of the epidural collection does not necessarily correlate with the site of the fistula, it can be mobilized and manifest several segments away from the site of the leak.

**Additional considerations:**

- Diffuse paquimeningeal hyperintensity in FLAIR images is slightly less sensitive than diffuse paquimeningeal enhancement in T1 with gadolinium. However, FLAIR sequences, because they do not require contrast administration, are useful in patients in whom the diagnosis of IHS is not initially suspected, in patients with renal failure, or in the follow-up of patients with IHS to avoid the administration of additional contrast.
- The sign of venous distension is the first sign to disappear after improvement of symptoms.
- Sometimes a marked decrease in the intensity of the signal of the cerebral white substance with respect to the grey substance can be observed, in particular in the subcortical white substance of the frontal and parietal lobes, in the FLAIR images, with recovery of this after the resolution of the picture.
This alteration in the intensity of the subcortical white substance signal could be related to a greater amount of deoxyhemoglobin, secondary to venous stasis.

**Differential diagnosis by image:**

The enhancement of the duration in the SHI is, in a characteristic way, linear and continuous, of diffuse and symmetrical distribution, most evident in the convexity, more evident in convexity, interhemispheric fissure and tentorium, and respecting the basal meninges.

Typically, unlike meningitis, the arachnoid is preserved, since it is not associated with the alteration of the hematomeningeal barrier (key data for diagnosis).

Likewise, evidence of epidural venous dilation as an indicator of hypovolemia of CSF can be considered to rule out other causes of diffuse paquimeningeal enhancement.

Diffuse dural thickening manifests as a diffuse dural hyperintensity in enhanced sequences in T2. This radiological data is interesting to differentiate IHS from idiopathic intracranial paquimeningitis, in which the dura mater appears hypointense or slightly hyperintense.

**HYDROCEPHALIA**

In the case of clinical suspicion of hydrocephalus, images play a central role in confirming the diagnosis, identifying the cause, and planning treatment.

In patients with the acute clinical picture, CT is the indicated technique. Otherwise, the mode of choice for examination is MRI.

The MR protocol for diagnosing the underlying cause in patients with confirmed hydrocephalus should always include high-resolution fluid-sensitive sagittal T2weight sequences.

Findings of hydrocephalus in the image:

- **Tetraventricular dilation** Fig. 3 on page 11
1. The dilation of the temporal horns is a very sensitive sign. Although there are no standard values for this in the literature, a diameter of > 2 mm in adults is considered pathological.
2. The third ventricle is balloon-shaped or rounded.
3. The posterior horns, which are usually slit-shaped, are also rounded.
4. Compared to the dilated ventricular system, the external space of the CSF is disproportionately thin, observing decreased visibility of furrows and cracks in the upper cranial convexity.

• **Evans´s Index**
  1. It is the quotient between the diameter of the front antlers and the maximum biparietal diameter.
  2. A value of > 0.3 is considered pathological.

• **Ratio between the diameter of the frontal horns and the distance between internal tables** measured both at the same level greater than 50%. Fig. 3 on page 11
• **Transependimal extravasation** of the CSF caused by increased pressure.
  1. It appears on cranial CT as hypodense changes in the region of the frontal and posterior horns.
  2. In MRI, these changes can be detected in T2-weighted FLAIR scans as signal hyperintensity of the periventricular white matter.
  3. Extravasation of CSF should be differentiated from changes related to periventricular white matter. These changes are usually less than 10 mm in diameter in the images of the axial cross section and their thickness decreases from anterior to posterior. In the case of clinical suspicion of acute hydrocephalus, FLAIR scans are sufficient to rule out an alteration in the circulation of the CSF and to detect or rule out extravasation of the CSF as an indirect sign of increased intracranial pressure.

• **Decreased mamilopontine distance**
• **Prominent "flow void" signal in the aqueduct** in T2W images (sign of communicating hydrocephalus).
• **Thinning of the corpus callosum**, with elevation and rectification of its contour (corpus callosum angle < 90°: Hydrocephalus > 90°: atrophy), as well as appearance of hyperintense punctation in its endoventricular face. These signs occur in cases of persistent hydrocephalus. Fig. 6 on page 14
• **The floor of the third ventricle** is usually inclined upwards. However, in the case of hydrocephalus, it becomes thinner or even tilts downwards.
• **The infundibular recess dilates** with respect to the pituitary gland.

**PSEUDOTUMOR CEREBRI SYNDROME (PTCS)**
The imaging is necessary to be able to rule out secondary causes of intracranial hypertension.

There are a series of radiological signs that guide us towards the diagnosis of HII.

**Brain Magnetic Resonance Imaging (MRI)**

Is the study of choice for parenchymal assessment.

Discard intracranial expandssive processes with certainty.

It rules out the main differential diagnoses linked to structural causes.

The signs of PTC that can be observed in MRI are:

*(The first four are part of the diagnostic criteria of Friedman et al. that are currently used)*

- **Anterior posterior flattening of the eyeballs** → resulting from the direct relationHSp between elevated ICP and intra-orbital pressure through the suarachnoid space, extending through the optic nerve sheath.
- **Empty sella turcica** → is associated with a long period of increased intracranial pressure and is believed to be secondary to an arachnocele that has herniated inferiorly through the selar diaphragm. Present in 70% of patients. [Fig. 7 on page 15](#)
- **Transverse sinus stenosis.** Compression due to increased ICP. Not to be confused with breast thrombosis. [Fig. 8 on page 16](#)
- **Optic nerve changes**
  1. Increased optic nerve sheath diameter.
  2. Increased CSF around the optic nerve or enhancement of the optic nerve as a result of damage to the blood-retinal barrier secondary to increased ICP.
  3. Tortuosity of the optic nerve (40%) with the horizontal device being more specific than the vertical one.
  4. Distension of the perioptic subarachnoid space bilaterally, which simulates compression of the same in coronal sequences and thinning in axial sequences (train rail appearance).
- **Decreased ventricular size**
- **Increase the size of trigeminal cisterns** with bone remodeling of the Meckel cavum secondary to the distension of the subarachnoid space.

**Cranial Tomography**
It's less useful than MRI to rule out small expansive processes.
It assesses the presence of hydrocephalus with acceptable sensitivity.
Angio CT with venous time has an acceptable performance to mislead TVC.
Indicated in cases of urgency or MR contraindication.

**Digital angiography**

Indicated in cases of atypical clinical picture with signs that raise the differential diagnosis with a CTV (consciousness disorder, epileptic seizures, alterations to the examination that exceed the affection of cranial pairs, etc.).

Another usefulness of this technique is if there are doubts in MRI Angio about asymmetries of the venous sinuses (especially the transverse ones) are congenital (frequent occurrence) or secondary to partial thrombosis or stenosis of the same. In these cases the angiography will be conclusive.
Fig. 1: Axial T2-Flair MR showing diffuse paimeningeal thickening

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Fig. 2: Sagittal T1-Flair MRI showing brain descent.

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Fig. 3: CT scan showing tetraventricular dilation with a history of neurinoma and subependymoma operated on years ago, which presents the clinical triad of gait apraxia, incontinence and dementia compatible with normotensive hydrocephalus of the adult. Note the marked dilation of temporal ventricles.
**Fig. 4:** Evans´ Index. > 0.3 is considered pathological.
Fig. 5: Ratio between the diameter of the frontal horns and the distance between internal tables measured both at the same level greater than 50%.

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Fig. 6

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Fig. 7: sagittal T1-weighted MRI showing empty sella turcica.

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**Fig. 8:** In the cerebral venous vascular assessment it is objective to decrease the caliber of the ventral portion of right transverse sinus.

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Conclusion

Imaging studies are important in the suspicion and identification CSF system disorders because they present definite radiological signs that can help to make the differential diagnosis. For this reason, the radiologist must be able to handle the radiological semiology that corresponds to each entity with ease, and thus be able to offer an early diagnosis that allows an opportune and opportune treatment.
References

• Incidencia: