

SPONTANEOUS INTRACRANIAL HYPOTENSION

Introduction –

The production, absorption, and flow of cerebrospinal fluid (CSF) play key roles in the dynamics of intracranial pressure. Alterations in CSF pressure can lead to neurologic symptoms, the most common being headache.

Terminology -

- ▶ Spontaneous (or idiopathic) low CSF pressure headache
- ▶ Low CSF volume headache
- ▶ Hypoliquorrhoeic headache
- ▶ Aliquorrhea
- ▶ CSF leak headache
- ▶ CSF hypovolemia
- ▶ CSF volume depletion

Key Features -

- ▶ Orthostatic headache
- ▶ Low cerebrospinal fluid (CSF) pressure
- ▶ Diffuse meningeal enhancement on brain MRI

Etiology & Pathophysiology -

CSF leak \longrightarrow \downarrow CSF pressure \longrightarrow \downarrow buoyancy of the brain's
supportive cushion \longrightarrow sagging of the brain in the cranial cavity, causing
traction on the anchoring and supporting structures of the brain

Headache -

- ▶ Traction on sensory nerves and bridging veins which is exaggerated in the upright position, hence the postural component of the headache.
- ▶ Secondary vasodilation of the cerebral vessels to compensate for the low CSF pressure
- ▶ CSF hypovolemia, rather than CSF hypotension has been proposed as the underlying cause of the headache syndrome

Low Venous Pressure -

- ▶ Lowering of venous pressure within the inferior vena cava system due to the activation of leg muscles during standing and walking.
- ▶ This hypothesis has been challenged by experts.

CSF LEAK -

- ▶ CT disorders - Marfan syndrome with Meningeal Diverticula
- ▶ Minor trauma like fall, sudden twist or stretch, sexual intercourse or orgasm, a sudden sneeze, sports activity → rupture of spinal epidural cysts, or may cause a tear in a dural nerve sheath
- ▶ Degenerative disc disease, osseous spurs, and microspurs. Another uncommon cause is the spontaneous development of a
- ▶ Spinal CSF venous fistula, which enables CSF to drain from the subarachnoid space directly into adjacent spinal epidural veins in the absence of a dural defect.
- ▶ Site of CSF leak - almost exclusively spinal; most occur at the thoracic or cervicothoracic junction

Epidemiology -

- ▶ Annual incidence is 5 per 100,000
- ▶ The peak incidence is around age 40, but children and older adults are also affected
- ▶ Female : Male ratio of 2:1

Clinical Features -

► **Headache** —

- ❑ **Location** – Frontal pain is reported by patients as often as occipital and diffuse pain
- ❑ **Onset** - sudden or gradual onset.
- ❑ **Character** – throbbing or dull pain
- ❑ **Aggrating & Relieving factors** - Headache relief is typically obtained with recumbency, usually within minutes. The headache is seldom relieved with analgesics. Exacerbating factors include erect posture, head movement, coughing, straining, sneezing, jugular venous compression, and high altitude
- ❑ **Timing** - The headache ordinarily develops within two hours, and in most cases within 15 minutes, of sitting or standing
- ❑ **Resolution** - Spontaneously within two weeks . In some cases, it lasts months or rarely years
- ❑ **Severity** - The headache severity is widely variable and ranges from mild to incapacitating

Clinical Features -

- ▶ Other clinical features include neck pain or stiffness, nausea, vomiting
- ▶ The neurologic examination is often normal

Clinical Features Contd. -

- ▶ Change in hearing (eg, hyperacusis, echoing, or tinnitus)
- ▶ Anorexia
- ▶ Vertigo
- ▶ Dizziness
- ▶ Diaphoresis
- ▶ Blurred vision
- ▶ Diplopia
- ▶ Transient visual obscurations
- ▶ Photophobia
- ▶ Unsteadiness or staggering gait
- ▶ Hiccups
- ▶ Dysgeusia

Clinical Features Contd. -

- ▶ Galactorrhea and hyperprolactinemia (pituitary stalk)
- ▶ Ataxia (posterior fossa)
- ▶ Quadriparesis (brainstem and upper cervical spinal cord)
- ▶ Cerebellar hemorrhage (cerebellar bridging veins)
- ▶ Posterior circulation infarction (deformation of cerebral arteries)
- ▶ Movement disorders including parkinsonism, tremor, chorea, and dystonia (deep midline structures)
- ▶ Hypoactive, hypo alert behavior (pons and midbrain)
- ▶ Decreased level of consciousness, stupor, and coma (diencephalon)

Evaluation & Diagnosis -

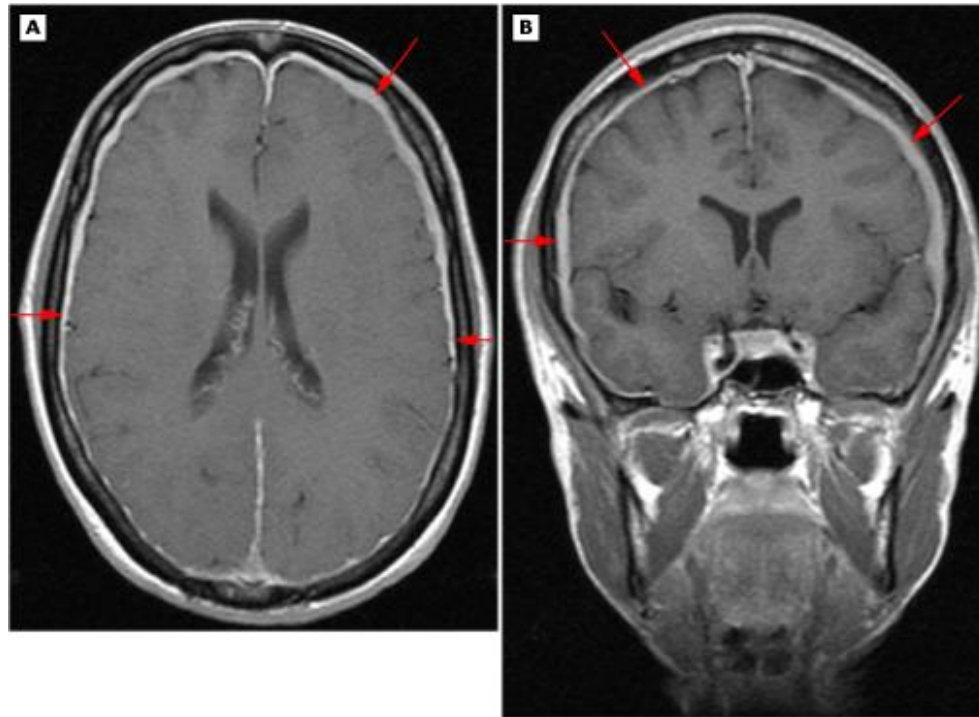
▶ Brain MRI –

- ❑ Diffuse meningeal enhancement (DME)
- ❑ Subdural hematomas or hygromas, presumably from rupture of the bridging veins as the CSF volume decreases
- ❑ "Sagging" of the brain, with cerebellar tonsillar herniation and descent of the brainstem
- ❑ Engorgement of cerebral venous sinuses
- ❑ Pituitary enlargement, flattening of the optic chiasm, and increased anteroposterior diameter of the brainstem
- ❑ Decrease in the size of cisterns and ventricles

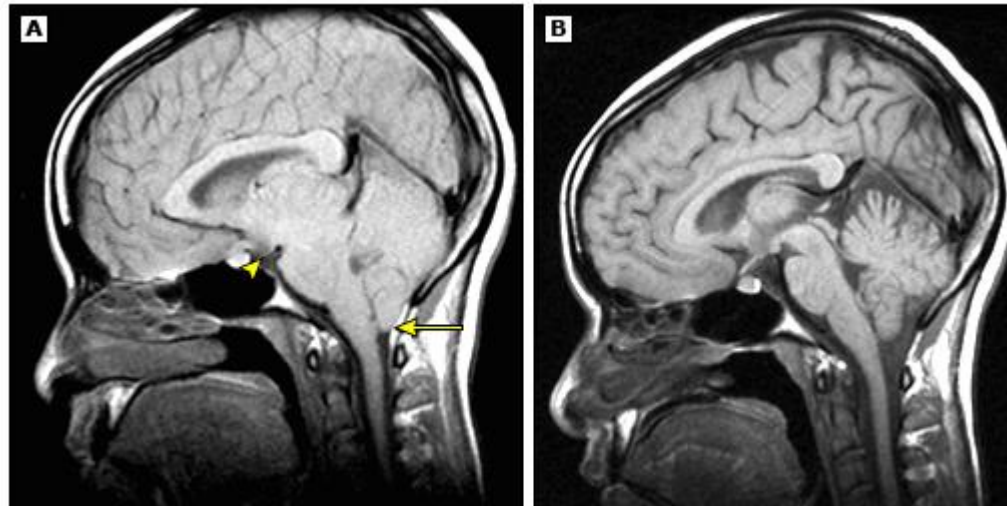
The acronym SEEPS (for Subdural fluid collections, Enhancement of the pachymeninges, Engorgement of the venous structures, Pituitary enlargement, and Sagging of the brain)

- Lumbar puncture – Used if MRI is not available

Diffuse meningeal enhancement



Sagging of the brain -



- ▶ **Spine MRI** - for confirming the diagnosis and for identifying the exact location of the CSF leakage
- ▶ **Radioisotope cisternography** — useful if MRI is normal or nondiagnostic.
- ▶ **CT myelography** — CT myelography is the best test to identify the exact site of the CSF leak caused by a dural defect when treatment beyond epidural blood patch is contemplated.
- ▶ **Dynamic CT myelography** — detecting rapid or high-flow CSF leaks.
- ▶ **MR myelography** - reserved for patients with debilitating symptoms of spontaneous intracranial hypotension when the site of the CSF leak has not been identified by CT myelography and other techniques.
- ▶ **Digital subtraction myelography** – to detect CSF leaks due to direct spinal venous fistulae

Complications and Misdiagnosis

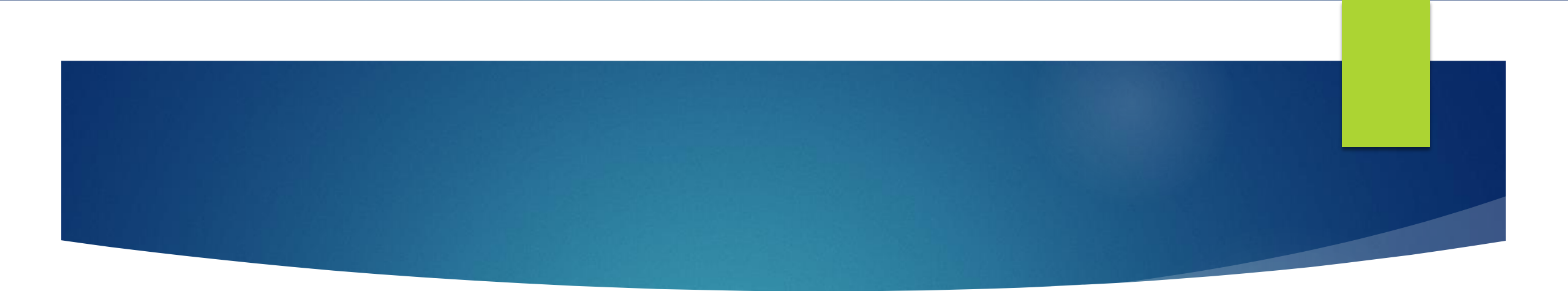
- ▶ **Superficial Siderosis** - hemosiderin deposition in the leptomeninges and subpial layer, often years after the onset of spontaneous intracranial hypotension
- ▶ **Frank dementia** – Frontotemporal Dementia
- ▶ **Misdiagnosis**

Frontotemporal brain sagging syndrome" (FBSS)

- ▶ Progressive behavioral symptoms and cognitive dysfunction suggestive of behavioral variant frontotemporal dementia.
- ▶ Clinical Features - headache and daytime somnolence.
- ▶ Diagnostics - MRI in all cases revealed sagging of the frontal and temporal lobes with downward displacement of the cerebellar tonsils, swelling of the midbrain, flattening of the ventral pons, and effacement of the basal cisterns.

Treatment -

- ▶ Conservative treatment – for acute uncomplicated headache of mild to moderate severity. Includes a combination of –
 - ❑ Avoidance of the upright position,
 - ❑ Bed rest
 - ❑ Analgesics
 - ❑ Oral or intravenous hydration
 - ❑ High oral caffeine intake
 - ❑ High salt intake
 - ❑ Abdominal binder

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- ▶ Epidural blood patch – Following patient require EBP
 - ❑ Acute, mild to moderate headache unresponsive to a reasonable period of conservative treatment
 - ❑ Severe headache or other disabling symptoms
 - ❑ An aggressive precipitating injury (eg, a water skiing accident) as compared with a minor or "trivial" trauma (eg, a sudden twist or stretch)
 - ❑ A history of connective tissue disease or joint hypermobility

▶ EBP regimen —

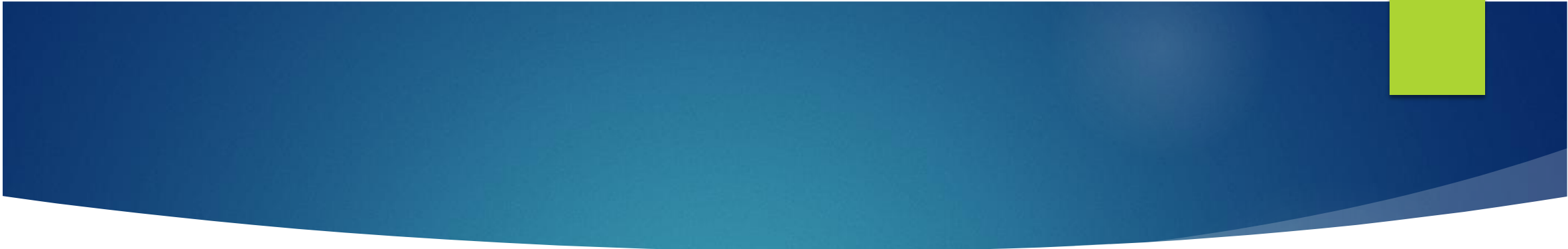
- ❑ The initial EBP treatment typically involves the infusion of 10 to 20 cc of autologous blood into the epidural space.
- ❑ A larger volume (20 to 100 mL) infusion is suggested if the initial blood patch is unsuccessful.

▶ Side Effects –

- ❑ Back pain
- ❑ Radiculopathy
- ❑ Leg paresthesias
- ❑ Fever

▶ Prevention of complications –

- ❑ Minimum of five days between blood patches
- ❑ Use of different injection sites for repeated blood patches

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- ▶ Epidural fibrin glue
 - ▶ Surgical repair – using suture or metallic clips. This method is used in patients who have failed all the above treatment.

Prognosis -

- ▶ Resolve spontaneously within two weeks. In some cases, it may last months, or in rare cases, years.
- ▶ Intermittent headaches have been reported at intervals of weeks, months, or years, probably caused by intermittent cerebrospinal fluid (CSF) leaks.
- ▶ Reoccurrence in approximately 10 percent of patients regardless of treatment

References -

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- ▶ Horton JC, Fishman RA. Neurovisual findings in the syndrome of spontaneous intracranial hypotension from dural cerebrospinal fluid leak. Ophthalmology 1994; 101:244.
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QUESTION??