Brain herniation is a potentially fatal condition that may present as a rise in intracranial pressure. The following text describes the various clinical signs to recognize this condition and the possible measures one can take to treat it.

Introduction

The cranium, or cranial vault, is shaped like a box formed by bones. It is fixed and nonexpandible. The cranium contains the brain tissue, cerebrospinal fluid (CSF), and blood vessels, all of which occupy precise and unwavering proportions within the cranium (see image). Pressure is exerted by the cranial vault on these internal structures. This pressure is known as intracranial pressure (ICP). At rest, in a normal adult, ICP is 7-15 mm Hg. ICP continuously changes due to various physical activities such as coughing, exercise, other strenuous activity, or even the respiratory cycle.
The **brain parenchyma makes up 80% of the brain’s volume, while CSF and blood vessels each account for 10%**. All 3 structures—the brain tissue, CSF, and blood vessels—have a unique and fixed relationship, because of which even the slightest change in the volume of any 1 of them leads to an increase in ICP.

These unique relationships are explained by the Monro-Kellie doctrine. According to this doctrine, **a self-regulating mechanism allows for the maintenance of a normal intracranial pressure when an injury occurs inside the cranium**. For example, if an individual suffers an intracranial hemorrhage, intracranial pressure increases and must be compensated for by a decrease in cerebrospinal fluid and venous blood to maintain the total intracranial pressure between 100 and 120 mL.

**Etiology**

Intracranial hypertension is defined as a **sustained (> 5 min) elevation of ICP of > 20 mm Hg**.
Herniation syndromes occur as a result of intracranial compartmental pressure gradients. There is displacement of the parenchymal tissues that lead to compression or displacement of the cranial nerves, brainstem, or cerebral vasculature. Edema and resultant deterioration occur in patient compliance as a result of ischemia or infarction from vascular compression.

When the self-regulating capacity of the cranium is exceeded and intracranial pressure exceeds normal limits, brain tissue is displaced (because of mass effect) from its normal location to adjacent spaces. This is called intracranial or brain herniation:

- **Mass effect**: space-occupying lesions such as a brain tumor, brain edema, abscess, contusion, and hematoma lead to the deformation of adjacent brain structures.
- Various medical conditions can lead to generalized swelling or edema of the brain. These include **acute liver failure**, hypertensive encephalopathy, and a state of ischemic-anoxia.
- An increase in venous pressure can lead to an increase in ICP. **Heart failure**, the obstruction of superior mediastinal or jugular veins, or venous sinus thrombosis can lead to increased venous pressure.
- Certain conditions can cause **obstruction of the CSF flow**, including **hydrocephalus** and extensive meningeal disease.

**Classification**

Brain herniation syndromes are classified into the following types according to the structure through which the tissue is herniated: **subfalcine**, **transtentorial**, **transcalvarial**, and **cerebellar**.
Six types of brain herniation: 4 supratentorial types, uncal, central (transtentorial), cingulate (subfalcine), and transcalvarial; and 2 infratentorial types, upward (upward cerebellar or upward transtentorial) and tonsillar (downward cerebellar). By: Delldot derivative work: RupertMillard. License: CC BY-SA 3.0

Subfalcine or Cingulate Gyrus Herniation (Midline Shift)
Subfalcine herniation is believed to be the most common of the 3 types of herniation (see image). It refers to cingulate gyrus displacement under the falx cerebri due to an increase in pressure in any of the cerebral hemispheres. This ultimately leads to widespread infarction involving the frontal and parietal lobes due to compression of the major blood vessel (anterior cerebral artery).

There are no associated specific clinical signs and symptoms and, when seen over a long duration, the effects are less severe compared with other types of herniation. Clinically, patients present with a decreased level of consciousness that is directly proportional to the degree of midline shift.

**Transtentorial Herniation (Downward, Central, or Uncal)**

When edema of the brain occurs, cerebral hemispheres are enlarged; thus, brain parenchymal tissue that is present in the supratentorial compartment is displaced caudally into the infratentorial compartment.

Various causes give rise to this displacement. These include supratentorial mass lesions, diffuse edema of the brain (seen in patients with traumatic injury to the brain), focal brain edema, or acute hydrocephalus.

The downward displacement of brain tissue causes compression of certain vital structures, including the 3rd cranial nerve, the upper brainstem, and the cerebral peduncles. Compression of the caudal cerebral artery causes ischemia and infarction of the occipital lobe of the brain. In some patients, distortion or traction of the superior portion of the basilar artery is also seen. Clinically, patients are usually in a state of coma. There is ipsilateral dilation of the pupil (mydriasis) due to stretching of the oculomotor nerve. Parinaud’s syndrome is present, (ie, failure to gaze upward, mid-dilated or pseudo-Argyll Robertson pupils, eyelid retraction, and conjugate downgaze [sun-setting sign]). In many patients, diabetes insipidus is also present.

**Tonsillar Herniation**

Tonsillar herniation occurs when downward pressure forces the cerebellar tonsils into the foramen magnum. When pushed in a downward direction, the cerebellar tonsils cause compression of the medulla oblongata and the upper cervical spinal cord. Clinical features include coma, apnea, hypertension, and neck stiffness.

**Symptoms**

The symptoms of brain herniation differ depending on the cause. In non-traumatic cases, a persistent headache can lead to nocturnal awakening. Headache is usually localized and its frequency, as well as severity, can gradually increase. There can be a worsening of headache during coughing, micturition, or defecation.

In patients with traumatic head injury, the main features of head injury are seen along with supplementary features including the presence of a low level of consciousness or unconsciousness, abnormalities in vital parameters, visual changes, and unbearable pain that waxes and wanes.

The presence of neurocutaneous syndrome, macrocephaly, hormonal abnormalities, lethargy, and/or personality change are risk factors for the development of intracranial pathology and thus must be examined regularly. Certain other factors, such as constant vomiting, abnormalities in growth patterns, focal
neurologic deficit, and nuchal rigidity, may be presenting symptoms of underlying pathology.

Both early and late signs of herniation are often present. Early signs include decreased level of consciousness, confusion, difficulty with memory and thinking abilities, restlessness, and lethargy. A deterioration in motor function, along with papillary dysfunction and vision changes, is also seen, as well as persistent headache. Certain personality changes may also occur. Finally, there is a decreasing Glasgow coma score.

Among the late signs, if papilledema is present, brain herniation is a confirmatory diagnosis. The pupils become dilated and do not react to light. There is a further decrease in the level of consciousness leading to a stuporous or comatose state. Other late signs include bradycardia, hyperthermia, and progressive hemiplegia.

Radiographic Features

Initial resuscitative measures, hemodynamic stabilization of the patient, securing the airway, circulation, and respiratory systems of the patient and early hyperosmolar therapy should be performed before a cranial computed tomography (CT) scan is performed.

Imaging is essential to finding the underlying cause for elevated ICP (see image). Generally, CT scan is used to rule out the lesions that cause a mass effect, such as a tumor or edema due to hemorrhage. It is also helpful in the identification of physical findings, such as midline shift and lateral displacement of the midbrain.

CT is preferred over magnetic resonance imaging (MRI) due to its wide availability and speed of imaging (see image below).

Imaging of patients with subfalcine herniation shows a shift in the septum pellucidum at the midline. There is effacement of sulci and ventricle on the affected side, as well as hydrocephalus.
In patients with transtentorial herniation, if it is central, there is **total obliteration of the basal subarachnoid cisterns** and small lateral ventricles suggestive of brain edema. There is an increase in the sagittal diameter of the brain stem and the basilar artery is displaced inferiorly.

In patients with tonsillar herniation, the **midbrain is effaced and displaced laterally along with effacement of suprasellar cisterns**. The uncus and medial temporal lobes are displaced medially.

![CT scan images](image)

Image: CT scan images after clinical signs of brain herniation developed. (A) Basal ganglia hemorrhage with perifocal edema. (B) Left-sided transtentorial herniation of the parahippocampal gyrus. (C) Edema of the left hemisphere with normally appearing right hemisphere. By: Dahlqvist MB, Andres RH, Raabe A, Jakob SM, Takala J, Dünser MW: Brain herniation in a patient with apparently normal intracranial pressure: A case report. License: CC-BY 2.0

**Treatment: General**

Correct diagnosis of the type of herniation and its underlying etiology are important parameters in its treatment as it helps the clinician take definitive steps to reduce the chances of the herniation entering into other compartments and the resultant further complications. It is therefore very important to understand all the general measures that can be taken if brain herniation is suspected so that prompt intervention can be instituted.

Among the general measures of treatment, the most important include a **constant control of vital signs**, **maintaining the head of the bed elevated to more than 45° and avoiding neck compression**, and using (if needed) an **endotracheal tube** to secure the airway (see image below).
Hyperventilation is a known method of lowering ICP, due to the association between cerebral blood flow and PaCO2. Hyperventilation causes decreased PaCO2; levels between 25 and 30 mm Hg lead to arterial vasoconstriction thus lowering cerebral blood flow (CBF), cerebral blood volume, and ICP. Endotracheal intubation should also maintain an adequate supply of oxygen.

Mannitol infusion in a dose of 2 g/kg (if it is not possible to use furosemide) helps reduce elevated ICP, as it decreases cerebral edema. A 30 cc bolus of hypertonic saline serum can be given. If necessary, in order to decrease cerebral metabolism, the patient can be sedated.
An **external ventricular drain** can be instituted to decrease the amount of cerebrospinal fluid amount and, thus, ICP.

It is essential to find and eliminate the injury responsible for the mass effect. In the case of a tumor, it is important to use **intravenous steroids** to decrease the size of the tumor as much as possible. If the injury corresponds to a hematoma or abscess, it must be drained as soon as possible (ie, decompressive craniectomy).

Finally, if the intracranial mass is a tumor, it can be diagnosed **using MRI and CT.** The tumor mass must be aggressively treated and removed immediately after diagnosis to prevent further complications and morbidity.

**In children, treatment depends on the clinical presentation and the cause of increased ICP.** A proper airway must be established. Blood pressure and ventilation must be maintained at normal levels, as there is a **risk of cerebral ischemia** if blood pressure is not maintained, and sufficient ventilation is necessary to avoid vasodilation due to hypercapnia. As per the standard **Pediatric Advanced Life Support (PALS),** the child’s cardiopulmonary status must also be stabilized so that further investigations, such as advanced imaging (head CT without contrast), can be performed.

In children with elevated ICP, **endotracheal intubation** should be done. Indications include refractory hypoxia, hypoventilation, or a loss of airway protective reflexes. In acute herniation, endotracheal intubation is necessary, as controlled hyperventilation is
required, and resuscitation medications can also be administered endotracheally.

Emergency Neurological Life Support Guidelines

Emergency neurological life support guidelines are helpful in managing a patient with brain herniation, as they classify the treatment plan into 4 tiers.

**Tier 0:**
- Brain-code resuscitation is initiated.
- Assessment of circulation, airway patency, and ventilation must be carried out.
- The head of the bed should be elevated to more than 30 degrees.
- Tracheal suctioning should be minimized.
- Body temperature should be normalized.
- Hyperosmotic fluids such as hyperosmotic saline should be used during resuscitation.
- High-dose corticosteroid therapy should be started for patients with vasogenic brain edema.
- Once the patient is stabilized, non-contrast cranial CT should be performed.

**Tier 1 (initiate only if Tier 0 standard measures do not correct ICP and brain herniation):**
- Hyperosmolar therapy with mannitol or hypertonic saline is initiated.
- Brief course, less than 2 hours in duration, of induced hypo-capnia, is attempted; PaCO$_2$ between 30 and 35 mm Hg.
- If the patient does not improve, decompressive surgery should be considered.
- If surgery is not possible, Tier 2 treatments should be initiated. Cranial CT scan should be repeated if the patient’s condition deteriorates.

**Tier 2:**
- An induced state of hypernatremia should be achieved, but sodium levels should not be > 160 mmol/L.
- Propofol sedation is recommended in this tier.
- If Tier 2 measures do not correct the state of ICP, rescue decompressive surgery should be considered.
- If the patient is not a surgical candidate, Tier 3 interventions should be initiated.

**Tier 3 (consider only if Tier 2 interventions fail and rescue decompressive surgery is not an option):**
- The administration of pentobarbital should be considered.
- Continuous electroencephalographic monitoring is indicated while the patient is receiving pentobarbital.
- Moderate hypothermia should be induced (target core temperature between 32 and 34ºC).
- Moderate hypocapnia with a PaCO$_2$ between 25 and 35 mm Hg should be induced.
- Hyperventilation should not be attempted for more than 6 hours.

**Decompressive Surgery**

Decompressive surgical interventions include the following:
- Placement of a ventricular drain
- Evacuation of an epidural hematoma in patients with traumatic brain injury if it is the cause of brain herniation
- Resection of the intracerebral lesion causing the mass effect responsible for brain herniation
- Removal of brain parenchyma
- Unilateral or bilateral craniectomy

Rescue decompressive surgery usually entails performing a decompressive craniectomy. It is usually required in patients with diffuse brain swelling due to traumatic brain injury; these patients usually do not respond to Tier 0–2 interventions.

Summary

Cerebral herniation can be the result of different kinds of injuries that cause an increase in ICP and exceed the brain's auto-compensatory mechanisms. Treatment should be initiated as early as possible in order to prevent complications. The symptoms of different types of cerebral herniation are different and are indicative of the possible brain structure that received the trauma, leading to change in its position or compression. Thus, careful observation of symptoms helps in early diagnosis and treatment.

References


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