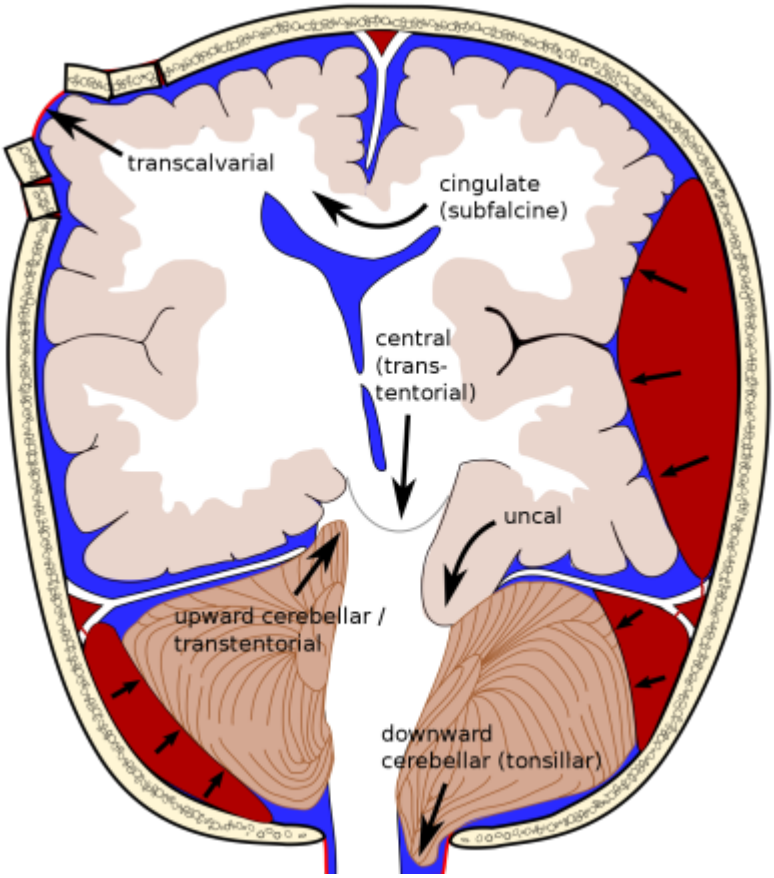
Central herniation



In the first phase of central herniation, the diencephalon and the medial parts of both temporal lobes are forced through a notch in the tentorium cerebelli.

Caused by diffuse brain edema as seen in patients with severe traumatic brain injury.

CT Scan shows effacement of the perimesencephalic cisterns and loss of Grey-white differentiation.

Stages

Early diencephalic stage (reversible)

Early. May be due to diffuse bilateral hemisphere dysfunction (e.g. from decreased blood flow from increased ICP) or (more likely) from bilateral diencephalic dysfunction due to downward displacement. This stage warns of impending (irreversible) midbrain damage but is frequently reversible if the cause is treated.

Midbrain—upper pons stage

When midbrain signs fully developed (in adults), prognosis is very poor (extreme ischemia of midbrain). Fewer than 5% of cases will have a good recovery if treatment is successfully undertaken

at this stage.

Consciousness: Altered alertness is first sign; usually lethargy, agitation in some. Later: stupor → coma.

Respiration: Sighs, yawns, occasional pauses. Later: Cheyne-Stokes.

Pupils: Small (1–3 mm), small range of contraction.

Oculomotor: Conjugate or slightly divergent roving eyes; if conjugate then brainstem intact.

Usually positive DOLL'S EYES and conjugate ipsilateral response to cold water calorics (CWC) in the caloric reflex test. Impaired upgaze due to compression of superior colliculus and diencephalic pretectum: Parinaud's syndrome

Motor: Early: appropriate response to noxious stimuli, bilateral Babinski, gegenhalten (paratonic resistance). If previously hemiparetic contralateral to lesion: may worsen. Later: motionlessness & grasp reflexes, then DECORTICATE (initially contralateral to lesion in most cases).

Lower pons—upper medullary stage

Respiration: Regular, shallow and rapid (20–40/min).

Respiration: Cheyne-Stokes → sustained tachypnea.

Pupils: Moderately dilated midposition (3–5mm), fixed. Note: in pontine hemorrhage pinpoint pupils appear because the loss of sympathetics leaves the parasympathetics unopposed, whereas in herniation, the parasympathetics are usually lost, too (3rd nerve injury).

Oculomotor: Doll's eyes & CWC impaired, may be dysconjugate. MLF lesion → internuclear ophthalmoplegia (when doll's or CWC elicited and dysconjugate, medially moving eye moves less than laterally moving eye).

Motor: Decorticate → bilaterally DECEREBRATE (occasionally spontaneously).

Pupils: Midposition (3–5 mm), fixed.

Oculomotor: Doll's eyes and CWC unelicitable.

Motor: Flaccid. Bilateral Babinski. Occasionally LE flexion to pain.

Late diencephalic stage

Patient becomes more difficult to arouse

Localizing motor responses to pain disappear and decorticate posture appears with eventual progression to decerebrate posturing

Progressive diencephalic impairment is thought to be the result of stretching of the small penetrating vessels of the posterior cerebral and communicating arteries which supply the hypothalamus and thalamus

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As herniation progresses to the midbrain stage signs of oculomotor failure appear The pupils become irregular and then fixed at midposition Oculocephalic movements become more difficult to elicit Extensor posturing appears spontaneously Motor tone is increased and plantar responses are extensor

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The progression of symptoms indicates irreversible ischemia and therefore intervention must occur before the midbrain stage to prevent permanent deficits from central herniation

Medullary stage (terminal stage)

Respiration: Slow, irregular rate and depth, sighs/gasps. Occasionally hyperpnea alternating with apnea

Pupils: Dilate widely with hypoxia.

Clinical characteristics differentiating uncal from central herniation

- decreased consciousness occurs early in central herniation, late in uncal herniation
- uncal herniation syndrome rarely gives rise to decorticate posturing

Outcome

In a series of 153 patients with signs of central herniation (altered level of consciousness, anisocoria or fixed pupils, abnormal motor findings) 9% had good recovery, 18% had functional outcome, 10% were severely disabled, and 60% died [1)].

Factors associated with a better result were young age (especially age≤17 yrs), anisocoria with deteriorating Glasgow Coma Score and nonflaccid motor function. Factors associated with poor outcome were bilaterally fixed pupils, with only 3.5% of these patients having a functional recovery.

Case reports

A 32-year-old female presented with a history of hydrocephalus and ventriculoperitoneal shunt placement at 9 months of age. She began experiencing significant headaches in college, later

accompanied by stereotypical 5- to 25-minute episodes of unresponsiveness, posturing and pupillary dilatation, and failing anticonvulsant therapy. No neurosurgical evaluation was sought because of small ventricles on brain imaging. Episodes became progressively more frequent over a 10-year period, eventually occurring daily. On presentation, 5 clinical events were captured on EEG over 12 hours of monitoring. With each episode, she became unresponsive and hypertensive, with fixed, dilated pupils and flexor posturing. Between events, she was awake and alert, without confusion or postictal state. She had papilledema and limited extraocular movements, with normal pupils and vital signs. Computed tomography scanning showed small ventricles. A shunt tap revealed no flow. With each episode onset, an EEG revealed an abrupt background rhythm slowing to 2-3 Hz delta range without epileptiform discharges. Between events, EEGs displayed normal waveform activity. Emergent ventriculoperitoneal shunt revision resulted in no further episodes in a 4-year follow-up period.

Slit Ventricle Syndrome can lead to severe intermittent brainstem herniation syndrome in the setting of shunt malfunction. Seizure diagnosis should be reserved for cases with proven functional shunt and EEG confirmation of epileptiform activity [2)]

A 62-year-old man with gait disturbance due to subdural fluid collection (SDFC) who underwent burr hole irrigation and additional craniotomy, in which postoperative deterioration resulted from rapidly progressing central herniation with a large amount of air accumulation. Epidural blood patch with saline infusion in the thoracic spine finally resolved central herniation.

SDFC deteriorating after surgery has never been reported. SDFC has communication with CSF differing from mature CSDH composed of closed cavity surrounded by neomembrane. Under situations of CSF hypovolemia due to spinal dural tear, opening the cranium can prompt air replacement in the CSF space, which might represent a substantial risk for central herniation caused by a rapid loss of buoyancy force [3)]

1)
Andrews BT, Pitts LH. Functional Recovery After Traumatic Transtentorial Herniation. Neurosurgery. 1991; 29:227–231
2)
Hamilton K, Koueik J, Maganti R, Iskandar B. Slit Ventricle Syndrome Leads to 10-Year History of Repetitive Transient Central Herniation Masquerading as Seizures: Hydrocephalus Case Report. World Neurosurg. 2019 Jun;126:134-138. doi: 10.1016/j.wneu.2019.02.106. Epub 2019 Mar 1. PMID: 30831286.
3)
Haku T, Motoyama Y, Takamura Y, Yamada S, Nakagawa I, Park YS, Nakase H. Central Herniation Induced by Craniotomy Prompting Air Replacement for Subdural Fluid Collection Due to Cerebrospinal Fluid Hypovolemia. World Neurosurg. 2018 Sep;117:182-185. doi: 10.1016/j.wneu.2018.06.059. Epub 2018 Jun 18. PMID: 29929033.