Brain Herniation

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1 Introduction

A wide range of abnormal imaging characteristics are evaluated daily, representing various intracranial pathologies like tumour, oedema, haemorrhage, or brain herniation. Brain herniation represents a shift of brain parenchyma across the various anatomical boundaries formed by intracranial bony ridges and dural folds. This usually occurs when an expanding mass lesion including tumours, hematomas, infarctions, infection, or haemorrhage exhausts the limited capacity of intracranial cavity (Andrews et al., 1986; Cuneo et al., 1979; Cruz et al., 2001; Dagnew et al., 2002; Reich et al., 1993; Ropper, 1986). Any insult to the blood supply or the ventricular system can also result in rise of intracranial pressure (ICP), with consequent mass effect upon adjacent compartments and subsequent herniation. Furthermore, other causes of cerebral herniation such as acute hydrocephalus, hepatic encephalopathy, regional or diffuse cerebral oedema by ischemia or infarction and therapeutic lumbar cerebrospinal fluid (CSF) drainage are also common (Cuneo et al., 1979; Lidofsky et al., 1992; Reich et al., 1993; Ropper, 1986; Muhonen & Zunkeler, 1994; Weinberg et al., 2003).

Development of herniation due to mass effect depends upon many factors including patient age, size/location/progression rate of mass lesion, and pressure gradient between mean blood pressure, CSF, and brain tissue (Andrews et al., 1986; Cuneo et al., 1979; Cruz et al., 2001; Dagnew et al., 2002; Reich et al., 1993; Ropper, 1986). Although it may affect any age group, elderly patients are at a higher risk of insult. This is due to more space for mobility of cerebral tissue owing to increased CSF content, large ventricles and cerebral atrophy. However, initial neurological findings are minimal in older individuals, due to less compression of brain tissue. Young patients have small, closely apposed basilar cisterns and ventricles, allowing less compensation for any mass effect. Therefore, neurological manifestations of brain herniation are most severe in younger individuals. Herniation can be a slow process resulting from a gradually expanding mass lesion such as subdural hematoma/tumour, or may develop rapidly after an acute traumatic event like epidural/subdural haemorrhage (Andrews et al., 1986; Cuneo et al., 1979; Cruz et al., 2001; Dagnew et al., 2002; Reich et al., 1993; Ropper, 1986). Chronic lesions usually present as severe anatomic herniation with few initial neurological manifestations. In contrast, a rapidly enhancing mass lesion may result in profound and dreaded neurological deficits.

Anatomically, four varieties of cerebral herniation are described. These include: subfalcine, transtentorial (ascending or descending), tonsillar and transphenoidal herniation (Figure 1) (Reich et al., 1993; Meyer, 1920; Scheinker, 1945). A fifth variety, transcalvarial herniation, has also been described. The most common sites are transtentorial, either upward or downward, and subfalcine herniation. The key to gain complete knowledge of brain herniation requires a thorough understanding of anatomic characteristics of brain, intracranial compartments, and dural folds.

2 Anatomical Considerations

The cranium is a rigid bony structure occupying the brain tissue, CSF, blood vessels and meninges. The meninges are protective coverings of the brain that lines the internal cavity to the cranium. Meninges consist of three membranous connective tissue layers: dura mater, arachnoid mater and pia mater. The dura mater is a tough, thick external fibrous membrane consisting of two layers. The outer periosteal layer is intimately adhered to the skull; the inner visceral layer projects inwards to form dural reflections. It divides the cranial cavity into various compartments and protects the brain against excessive movement.
Figure 1: Brain Herniation. 1: Uncal; 2: Descending; 3: Subfalcine; 4: transcalvarial 5: Ascending; 6: Tonsillar.

These dural reflections are known as falx cerebri, tentorium cerebelli, falx cerebelli and the diaphragm sellae.

The falx cerebri is a sickle-shaped inward reflection that separates the cerebral hemispheres. The falx cerebri is adhered in the midline to the inner surface of the calvaria, extending anteriorly from crista galli to the internal occipital protuberance posteriorly (Gray, 1974). Further posteriorly, falx cerebri continuous as the tentorium cerebelli. Falx cerebri is narrow anteriorly but broadens posteriorly, which makes it more resistant to any pressure effect in the posterior portion. For this reason, subfalcine herniations are more common anteriorly. The falx cerebri encloses the superior sagittal sinus along its upper margin and the inferior sagittal sinus along its lower margin. The superior sagittal sinus drains directly into the sinus confluence while inferior sagittal sinus drains into the straight sinus (Gray, 1974). Falx cerebri is in close relationship with anterior cerebral artery (ACA) anteriorly, cingulate gyrus inferiormedially, and internal cerebral veins, vein of Galen and deep subependymal veins posteriorly. Any mass effect in this region can lead of brain ischemia/infarction as a result of arterial compression or brain parenchymal congestion/raised ICP due to compression of venous system (Gray, 1974).

The tentorium cerebelli is a crescent shaped lamina that separates the occipital lobes of cerebral hemispheres from the cerebellum. It resides in the cerebrocerebellar fissure, attached anteriorly to the clinoid processes of the sphenoid bone, anterolaterally to the petrous part of the temporal bone, and posterolaterally to the internal surface of occipital and the parietal bone (Gray, 1974). Superiomedially, tentorium cerebelli continues as falx cerebri; giving it a tent-like appearance. The concave anterioimedial margin of the tentorium between the two clinoid processes is free, leaving a semiovular opening known as tentorial notch or incisura through which the brainstem extends into the infratentorial compartment.
Tentorium cerebelli encloses the transverse sinus, and straight sinus which empties into the sinus confluence (Gray, 1974).

The space between the free margin of incisura and the brain stem varies considerably in size, from virtually no space to as much as 7 mm on either side, among the individuals (Sunderland, 1958). The dimensions of tentorial notch have been categorized into eight distinct varieties. Alder and Milhorat (2001) have observed wide inter-individual variations in the exposed portions of cerebellar parenchyma within the incisura and poor correlation between brain stem position and the tentorial edge. This pattern potentially alters the susceptibility for upward or downward transtentorial herniation among the individuals.

The principal structures coursing through the incisura include the midbrain, oculomotor nerve, posterior communicating artery, posterior cerebral artery (PCA) and superior cerebellar artery (SCA). The medial margin of uncus, parahippocampal gyrus, hippocampal formation and first six cranial nerves are also intimately related to the incisura. The length, course and anatomical relationship of third nerve to the skull base vary widely in the population (Sunderland & Hughes, 1946). The pupillo-constrictor fibres of third nerve are placed along its outer aspect and are exceedingly sensitive to external pressure. Thus, any mass effect in this region pressurizes the third nerve, resulting in loss of pupillary constriction. For this reason pupillary dilation is considered as a symbol of transtentorial herniation (Adler & Milhorat, 2001; Cuneo et al., 1979; Ropper, 1986; Sunderland & Hughes, 1946). The PCA and anterior choroidal artery (ACHA) are vulnerable to occlusion by downward transtentorial herniation. In contrast, SCA is sensitive to upward herniation from the infratentorial compartment. Occlusion of the ACHA results in infarction of vital structures including the optic tract, temporal lobe, basal ganglia, cerebral peduncles, and midbrain (Grossman & Yousem, 1994). Occlusion of the SCA results in cerebellar infarction while PCA occlusion results in occipital lobe infarction (Komaki & Handel, 1974). Histological changes include neuronal swelling with lipid vacuolization and peripherally placed nuclei. Surviving neurons become pyknotic and fibrous gliosis ensues. Anatomically, midbrain consists of four components: tectum, tegmentum, ventricular mesocoelia and cerebral peduncles. The mid brain contains many vital structures including third and fourth cranial nerve nuclei, the red nuclei, substantia nigra, periaqeductal gray matter, neurons of reticular activating system, aqueduct of sylvius and numerous interneuronal connections between the cerebral cortex and the lower brain stem or spinal cord. Thus any mass effect in this region leads to profound neurological deficits. The subarachnoid space inside the incisura is divided into various basilar cisterns including quadrigeminal cistern posteriorly, and the interpeduncular and suprasellar cisterns anteriorly (Sunderland & Hughes, 1946; Nguyen et al., 1989). They act as hydraulic buffers to prevent any pressure effect upon the midbrain. Compression of these cisterns is regarded as a hallmark sign of transtentorial herniation. It is also considered as a negative prognostic sign in condition of intracerebral hematoma or contusion (Ross et al., 1989; Toutant et al., 1984).

3 Pathophysiology

3.1 Subfalcine Herniation

The subfalcine herniation is considered as one of the most common variety of brain herniation. It occurs when a supratentorial mass causes a midline shift of brain tissue under the falx cerebri together with lateral displacement of the falx cerebri (Osborn, 1996). Though, subfalcine herniation can occur in isolation, the usual presentation is in conjunction with transtentorial herniation (Taveras, 1996). This could be due to generalized effect of clinical pathology, but does not necessarily signify severe clinical presentation.
Table 1: Aetiology of brain herniation

<table>
<thead>
<tr>
<th>S. No.</th>
<th>Type</th>
<th>Causes</th>
</tr>
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</table>
| 1.     | Developmental | Malformed posterior fossa  
               | CSF loss through open spinal defect        |
| 2.     | Traumatic   | Pneumocephalus  
               | Epidural/subdural                          |
|        |            | Intracerebral haemorrhage                    |
|        |            | Brain contusion                              |
| 3.     | Infectious  | Brain abscess                               |
|        |            | Empyema                                     |
|        |            | Meningitis                                  |
|        |            | Encephalitis                                |
| 4.     | Metabolic   | Hepatic encephalopathy                       |
|        |            | Toxic-metabolic encephalopathy              |
| 5.     | Vascular    | Aneurysm                                    |
|        |            | Ischemic stroke                             |
|        |            | Hydrocephalus                               |
|        |            | Hypertensive encephalopathy                 |
| 6.     | Neoplastic  | Supratentorial/infratentorial tumours        |
| 7.     | Therapeutic | Over-drainage of CSF through ventriculo-peritoneal shunting  
               | Graft herniation during translabyrinthine craniotomy |

Due to its inferiomedial location, cingulate gyrus is the most commonly involved portion in subfalcine herniation. The severity of herniation may vary from mild form with minimal midline shift to complete herniation of the cingulate gyrus and the adjoining parenchyma. Continued mass effect upon the cingulate gyrus can lead to brain parenchymal ischemia or necrosis (Loevner, 1999). Owing to its rigid fibrous assembly, falx cerebri protects the brain against any movement; so even a few millimetres of midline shift is considered significant. Further rise in mass effect compresses the ventricular system including ipsilateral lateral ventricles and foramina of Monro. This results in enlargement of contralateral lateral ventricles with consequent obstructive hydrocephalus (Osborn, 1996). The ACA and its tributaries (the callosomarginal, pericallosal, and frontopolar arteries) course between the falx cerebri and the adjacent frontal and parietal lobes; they are vulnerable to compression by any mass effect in the anterior portion of supratentorial compartment (Osborn, 1996; Loevner, 1999). As the cingulate gyrus extends under the falx, distal ACA gets occluded and consequently ischemia/infarction develops in the supplied portion, leading to contralateral leg weakness (Taveras, 1996; Hassler, 1967; Sohn & Levine, 1967; Barr & Gean, 1994). Complications such as vascular aneurysms have also been reported due to compression of the ACA in subfalcine herniation (Nakstad, 1986). When the lesion is located posteriorly, the internal cerebral veins, vein of Galen, or the deep subependymal veins may get involved, leading to brain congestion and rise in ICP (Sohn & Levine, 1967; Ecker, 1948).

3.2 Transtentorial Herniation

Transtentorial herniation is a dreaded complication, which has an acute presentation, rapid progression and lethal outcome. Transtentorial herniation was first described by Hill in 1896. It is the upward or downward shift of neural parenchyma across the tentorium at the level of incisura (Elguera, 1962). Due to
minimal space within the tentorial notch, only a few millimetres midline shift of brain stem is sufficient to cause significant compression (Elguera, 1962; Schwarz & Rosner, 1941; Smyth & Henderson, 1938). Further increase in pressure upon the brainstem, compresses the aqueduct of sylvius and cardiorespiratory centres, which can be fatal (Schwarz & Rosner, 1941; Taveras, 1996). Transtentorial herniation can be further subcategorized into ascending, descending and uncal herniation. Early or unilateral herniation is more accurately called uncal herniation. The terms cranio-caudal, rostro-caudal, central or bilateral descending transtentorial herniation, are more appropriately termed as "descending transtentorial herniation".

3.3 **Uncal Herniation**

Uncal herniation, a subtype of descending transtentorial herniation, was first described by James Collier in 1904. It occurs when innermost portion of temporal lobe, the uncus is pushed inferiomedially across the free edge of the tentorium cerebella. Furthermore, uncus obliterates the supracellular cistern, compresses the oculomotor nerve and cerebral peduncles, and obliterates the aqueduct of sylvius (Meyer, 1920; Jefferson, 1938; Howell, 1959; Kernohan & Woltman, 1929; Marshall et al., 1983). The stalk of pituitary gland may also get stretched across the sellar diaphragm, leading to pituitary infarction (Howell, 1959). Pressure effect upon third nerve causes loss of parasympathetic innervation, resulting in pupillary dilation and loss of light reflex. Pupillary dilation is followed by the motor effects of third nerve compression which precipitates as downward and outward deviation of eye, due to loss of innervation to all ocular muscles except those supplied by fourth and sixth cranial nerve (Barr, 1994; Kernohan & Woltman, 1929). The neurological deficit occurs in this sequence because the parasympathetic fibres are located outer to somatomotor fibres in third nerve, hence compressed first. Further increase in mass effect, compresses the midbrain leading to loss of both parasympathetic and sympathetic innervation bilaterally. This results in mid-positioned (4-5 mm) pupils not reacting to light. Uncal herniation can also compress the PCA resulting in ischemia of ipsilateral primary visual cortex, with consequent contralateral visual field defects in both eyes (Homonymous hemianopia). Pressure effect upon the ipsilateral cerebral peduncle (comprises corticospinal and corticobulbar fibres) results in contralateral hemiparesis. In contrast, uncus may also compress contralateral cerebral peduncle against the tentorium cerebelli, resulting in ipsilateral hemiparesis (false localizing sign), and known as Kernohan’s notch (Kernohan & Woltman, 1929; Marshman et al., 2001). Uncal herniation may also precipitate duret haemorrhage in midbrain and pons, which can be fatal.

3.4 **Descending Herniation**

Descending transtentorial herniation occurs when a midline/bilateral supratentorial mass pushes the diencephalon and/or the parahippocampal gyrus inferiomedially from the supratentorial space, into the incisura (Stovring, 1977; Sheinker, 1945). Jennett and stern (1960) showed that enlargement of supratentorial mass results in a pressure gradient between the supra and infratentorial compartment; maximum over the tentorium, intermediate under the tentorium and minimum in the spinal subarachnoid space. This pressure gradient is responsible for inferiomedial displacement of brain tissue (Hahn & Gurney, 1985). Imaging studies have documented that nearly 50% of patients with transtentorial herniation also develop downward shift of brain stem and concurrent cerebellar tonsillar herniation (Reich, 1993). Typical pathological manifestations include inferiomedial displacement of parahippocampal gyrus and diencephalon, obliteration of basilar cisterns, and compression of oculomotor nerve, midbrain and posterior cerebral/ACHA (Sheinker, 1945; Stovring, 1977, Blinkov et al., 1992). With further increase in herniation, pons and me-
dulla are also compressed (Feldmenn, 1988). In severe cases, occlusion of small perforating branches supplying the upper brain stem results in brain stem haemorrhage (Duret haemorrhage) (Howell, 1959; Kernohan & Woltman, 1929; Thompson & Salzman, 1988). Haemorrhage may also occur due to reperfusion injury after initial ischemia. Haemorrhage may further compromise the cardiorespiratory centres, which may be fatal (Sheinker, 1945).

3.5 Ascending herniation

Ascending transtentorial herniation results from upward migration of brain parenchyma through the incisura due to an expanding mass lesion within in the posterior fossa (Ecker, 1948). Common causes are hematoma, infarction, infection or tumour. It may however, rarely be seen after graft herniation into posterior fossa during translabyrinthine craniotomy. Herniated brain tissue includes central lobule, culmen, and cerebellar vermis, which compresses the dorsal mid brain, fourth ventricle, prectectum and the cerebral aqueduct (Sheinker, 1945). Pretectal compression leads to loss of vertical eye movements (Reich et al., 1993; Sunderland, 1958; Osborn, 1996). Increasing pressure effect compresses the pons, leading to loss of sympathetic innervation descending through the pontomedullary region. Thus, the unopposed parasympathetic papillary supply to the eye leads to small minimally reactive pupil, termed as ‘pontine pupil’ (Cuneo et al., 1979; Sunderland, 1958; Alder & Milhorat, 2002; Ross et al, 1989). However, coexistent compression of midbrain and third nerve may also cut the parasympathetic supply, leading to midpositioned or even dilated fixed pupil (Cuneo et al., 1979). With increasing upward herniation obstructive hydrocephalus develops, and the quadrigeminal cistern and superior cerebellar cistern becomes obliterated (Osborn, 1991; Osborn, 1996; Speigelman, 1989). The distal part of SCA and PCA may get compressed, resulting in infarction of cerebellar hemisphere and occipital lobe. Increasing mass effect also compresses the vein of Galen and the basal vein of Rosenthal, which further exacerbates the neuronal congestion, resulting in rise in ICP and secondary haemorrhagic infarction of diencephalon (Reich et al., 1993; Cuneo et al., 1979).

3.6 Tonsillar herniation

In Tonsillar herniation, increased pressure gradient within the posterior fossa pushes the cerebellar tonsils through the foramen magnum, which compresses the lower part of brain stem and upper spinal cord, having life threatening consequences (Reich et al., 1993; Cuneo et al., 1979; Ross et al., 1989; Meyer, 1920; Kernohan & Woltman, 1929). Common causes are again the same, but it has also been observed in condition of malformed posterior fossa. It occurs usually in conjunction with ascending or descending transtentorial herniation in majority of cases (Meyer, 1920). The lumber puncture in presence of supratentorial mass causes sudden decline in CSF pressure below the level of foramen magnum. This augments the cephalocaudal pressure gradient, which results in tonsillar herniation with consequent sudden decline in vital parameters of patient. For this reason, lumber puncture is contraindicated in presence of intracranial tumour (Ramsey, 1994). Acute tonsillar herniation can cause significant compression upon the medulla, posterior inferior cerebellar arteries and fourth ventricle which can be fatal (Cuneo et al., 1979). Ischemia and infarction of cerebellar tonsil, lower cerebellum and entire lower brain stem and upper spinal cord may occur due occlusion of vertebral arteries, their branches and the origin of anterior spinal artery (Reich et al., 1993; Cuneo et al., 1979). Tonsillar herniation of cerebellum is also termed as chairi malformation. Clinical symptomatology may be subtle or patient may exhibit Lhermitte’s phenomenon, defined as dyesthesia in arms and legs upon forward bending of head (Reich et al., 1993; Cuneo et al., 1979).
1979). It is considered to be due to pressure upon anterior spinal cord tracts. Compression of ventricular system may result in obstructive hydrocephalus (Cuneo et al., 1979).

3.7 Transphenoid (transalar) herniation

Transphenoidal herniation occurs when mass lesion shifts brain tissue across the sphenoid wing. There are two types of transphenoidal herniation, anterior/ascending and posterior/descending. Posterior herniation occurs when the frontal lobe extends posteriorly and inferiorly over the greater sphenoid ridge, causing posterior displacement of the sylvian fissure and the middle cerebral artery (Taveras, 1996). Anterior herniation occurs when the temporal lobe, sylvian fissure, and middle cerebral artery are shifted anterosuperiorly over the sphenoid wing. Alar herniations are difficult to demonstrate on imaging and clinical symptomatology is also subtle.

3.8 Transcalvarial herniation

It is the herniation of oedematous brain tissue through a bony defect, either due to a congenital bony anomaly, trauma, otitis media or following decompressive craniectomy (Figure 2). For external herniation to occur, three pre-requisites have been described: presence of a bony defect, a dural defect, and increased intracranial pressure (Ramsden et al., 1985). The oedema following decompressive craniectomy is known to result from hyperperfusion of brain parenchyma or increased capillary permeability resulting from decrease in interstitial hydrostatic pressure (Stiver, 2009). This may lead to pinching of cerebral vessels, epidural haematoma or injury to the herniating brain parenchyma at the level of defect edge, resulting in ischemia and necrosis of the affected portion. This manifestation is more common in cases where decompression is performed through small sized craniectomy defect (Yang et al., 2008). For this reason, it is always advised to perform sufficient sized craniectomy and optimal decompression, allowing brain to expand outward without constriction.

4 Radiological Aspects

Under the emergent condition, due to easy availability and speed of imaging, cranial computed tomography (CT) should be obtained without any delay to identify the underlying primary pathology that requires immediate medical attention and/or surgical intervention. However, magnetic resonance imaging (MRI) may subsequently be needed for more accurate categorization of mass lesion and associated complications.

4.1 Subfalcine herniation

Imaging signs include unilateral mass effect such as haemorrhage or tumour causing subfalcine shift of brain to the contralateral side, asymmetry of the anterior falx with a widened CSF space on the contralateral anterior falx (Osborn, 1996; Loevner, 1999) (Figure 3). There may be ipsilateral lateral ventricle compression with contralateral lateral ventricle and atria dilation. Other finding includes truncation of anterior aspect of ipsilateral frontal horn and widening of contralateral frontal horn (Osborn, 1996; Loevner, 1999).
Figure 2: Transcalvarial Herniation

Figure 3: Subfalcine Herniation
4.2 Transtentorial herniation (Uncal)

Imaging findings include inferiomedial displacement of uncus across the tentorial edge, widening of ipsilateral cerebellopontine angle cistern, distortion of suprasellar cistern, compression of cerebral peduncle (ipsilateral or contralateral) and midbrain, aqueductal compression with consequent obstructive hydrocephalus, and PCA compression with occipital infarction (Taveras, 1996; Osborn, 1976) (Figure 4). Duret haemorrhages may also be seen.

![Figure 4: Uncal herniation](image)

4.3 Transtentorial herniation (Descending)

Imaging findings include the downward displacement of the parahippocampal gyrus through the incisura, and widening of ipsilateral and obliteration of contralateral ambient and prepontine cisterns (Feldmann et al., 1988; Osborn, 1976; Osborn, 1996). The contralateral temporal horn is also widened. There may be ipsilateral lateral ventricle compression with subsequent dilatation of the contralateral ventricle. Midbrain is twisted, and the cerebral peduncles become elongated and flattened (Feldmann et al., 1988; Osborn, 1976). There is downward shift of pineal calcification and midline shift of brain parenchyma. Occipital lobe infarction and Duret haemorrhage can also be seen.

4.4 Transtentorial herniation (Ascending)

MR imaging is useful in early identification of upward herniation, prior to the development of severe neurological complications and can be used to monitor the course of progression or recovery of ascending herniation (Figure 5). Imaging findings include upward migration of cerebellar tissue through the tentorium cerebelli, flattening of smile shaped quadrigeminal cistern (termed as crooked smile), obliteration of
quadrigeminal cistern (termed as frown shape) and superior cerebellar cistern and in severe cases, ‘spinning top appearance’ of midbrain due to bilateral compression of posteriolateral aspect of midbrain (Osborn, 1978). It may be associated with infarct in the territory of PCA and SCA, obstructive hydrocephalus as a result of pressure upon aqueduct (Reich et al., 1993). Rostral migration of the vermis into the tentorial apex is confirmed angiographically by elevation of superior vermian arteries and anterosuperior displacement of cistern segment of the ACHA (Osborn, 1978).

4.5 Tonsillar herniation

Radiological findings include downward displacement of cerebellar tonsil up to the level of odontoid process and lesser amount of CSF in the foramen magnum, on axial CT images (Grossman & Yousem, 1994). A sagittal MRI is however, more accurate method for determining the herniation (Reich et al., 1993). Tonsillar herniation should be differentiated from low lying normal cerebellar tonsils. Radiologically, cerebellar tonsil should lie at least five millimetres below the level of foramen magnum in adults or seven millimetres in children, to categorize it as herniation (Reimer et al., 2010). Cisterna magna obliteration with associated PCA infarction and hydrocephalus are also common (Reimer et al., 2010).

4.6 Transphenoidal (transalar) herniation

Imaging characteristics include anterior or posterior displacement of middle cerebral artery in anterior or posterior alar herniation, respectively (Taveras, 1996).
## 5 Clinical Symptomatology

<table>
<thead>
<tr>
<th>Type</th>
<th>Early features</th>
<th>Late Features</th>
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<tbody>
<tr>
<td>Subfalcine</td>
<td>• Headache</td>
<td>• Ipsilateral anterior cerebral artery infarction</td>
</tr>
<tr>
<td></td>
<td>• Contralateral leg weakness</td>
<td>• Contralateral hydrocephalus</td>
</tr>
<tr>
<td>Transtentorial (Uncal)</td>
<td>• Anisocoria</td>
<td>• Midpositioned fixed pupil</td>
</tr>
<tr>
<td></td>
<td>• Ipsilateral dilated pupil</td>
<td>• Ptosis</td>
</tr>
<tr>
<td></td>
<td>• Loss of light reflex</td>
<td>• Downward and outward deviation of eye</td>
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<tr>
<td></td>
<td>• Altered consciousness</td>
<td>• Homonymous hemianopia</td>
</tr>
<tr>
<td></td>
<td>• Contralateral hemiparesis</td>
<td>• Pituitary infarction</td>
</tr>
<tr>
<td></td>
<td>• Ipsilateral hemiparesis</td>
<td>• Hydrocephalus</td>
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<tr>
<td></td>
<td></td>
<td>• Hemiplegia</td>
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<tr>
<td></td>
<td></td>
<td>• Coma</td>
</tr>
<tr>
<td>Transtentorial (descending)</td>
<td>• Constricted reactive pupils</td>
<td>• Midpositioned fixed pupils</td>
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<tr>
<td></td>
<td>• Cheyne stokes breathing</td>
<td>• Loss of oculocephalic reflex</td>
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<td></td>
<td>• Decorticate posturing</td>
<td>• Decerebrate posturing</td>
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<td></td>
<td>• Altered consciousness</td>
<td>• Coma</td>
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<tr>
<td></td>
<td></td>
<td>• Apnoea, cardiac arrest</td>
</tr>
<tr>
<td>Transtentorial (ascending)</td>
<td>• Nausea, vomiting</td>
<td>• Hydrocephalus</td>
</tr>
<tr>
<td></td>
<td>• Constricted minimal reactive pupil (pontine pupils)</td>
<td>• Coma</td>
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<tr>
<td></td>
<td>• Loss of vertical eye movement</td>
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<td></td>
<td>• Altered consciousness</td>
<td></td>
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<tr>
<td>Sphenoidal</td>
<td>• Subtle</td>
<td>• Subtle</td>
</tr>
<tr>
<td>Tonsillar</td>
<td>• Bilateral arm dysesthesia</td>
<td>• Hydrocephalus</td>
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<td></td>
<td>• Pontine pupils</td>
<td>• Coma</td>
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<tr>
<td></td>
<td>• Loss of lateral eye movement (Ocular bobbing)</td>
<td>• Apnoea, cardiac arrest</td>
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<tr>
<td></td>
<td>• Altered consciousness</td>
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Table 2: Symptoms of brain herniation

## 6 Management

Management of brain herniation begins with initial resuscitative measures like airway, breathing and hemodynamic concerns, followed by therapies aimed at reduction of ICP while maintaining cerebral perfusion pressure (CPP), minimizing oxygen demand while maximizing oxygen delivery and prevention of hypercarbia and acidosis/alkalosis (Figure 4) (Lidofsky et al., 1992; Stevens et al., 2012; Andrews, 2003). If the initiating cause is uncertain, a cranial CT scan should be performed as an emergency measure just after initial resuscitation, for early identification of a treatable mass lesion. Prolonged unattended herniation may lead to permanent brain injury, having devastating consequences in the form of permanent neurological sequel or death. These interventions help to tolerate the pressure effect of an expanding intracranial mass lesion temporarily, until final diagnosis and treatment is initiated.
6.1 Initial Resuscitation and Management

Resuscitative measures began with assessment of airway patency, ventilation and adequate circulation. Loss of consciousness diminishes the normal neuromuscular tone and reflexes of upper airway (cough and gag), endangering the airway further through aspiration and its sequel (Colquhoun et al., 2004). This may be corrected by various maneuvers including head tilt with chin lift or jaw thrust technique or use of adjuncts, such as an oropharyngeal or nasopharyngeal airway. Though, the use of head tilt alone will itself relieve the obstruction in 80% of patients, it should be avoided in cases of suspected cervical spine injury. If injuries are visible on upper half of the body, trauma to cervical spine should be considered by default, unless excluded by its radiography (Colquhoun et al., 2004). Such cases should be managed by chin lift and jaw thrust alone, if possible. Furthermore, head and neck should be maintained in a neutral position by manual inline immobilization, semi-rigid collars, sandbags, spinal board, and securing straps. If the patient needs to be turned, “log rolled” rotation should be there into a true lateral position, avoiding any movement in the spine (Colquhoun et al., 2004). In the field, non-invasive ventilation with 100% oxygen through an integrated mask and reservoir bag, may act as a temporary maneuver to prevent hypoxemia. However, endotracheal intubation remains the gold standard for securing an airway, so it should be provided as early as possible. Regardless of a negative cervical radiograph, excessive movement over the cervical spine should be avoided during intubation, as there are 20% chances of significant cervical injury despite a normal screening radiograph (Bivins, 1988). This could be better achieved by use of flexible fibreoptic bronchoscope, McCoy laryngoscope, supraglottic airway or combitube etc. Surgical approaches like needle cricothyroidotomy or tracheostomy may be necessary, if other means of securing an airway fail. Once the airway has been secured, controlled ventilation should be started at the earliest in order to maximize oxygen delivery, reverse hypercarbia and to prevent acidosis/alkalosis (Andrews, 2003).

Once the airway, oxygenation and ventilation are adequate, circulation should be assessed and supportive measures should be rapidly initiated. Isolated head injury rarely causes hypotension, although associated scalp lacerations or multisystem injuries may cause significant blood losses (Colquhoun et al., 2004; Haddad & Arabi, 2012). Hypotension may also be due to systemic vasodilation due to spinal cord injury. Wherever possible, any on-going bleeding should be controlled and fluid resuscitation should be started, if the losses appear to be significant. If the initial blood pressure is normal, fluid resuscitation should be modest, as overhydration may aggravate cerebral/pulmonary oedema (Colquhoun et al., 2004; Haddad & Arabi, 2012). Most vital parameters change little in adults until more than 30% blood volume has been lost; children compensate even more effectively. Any patient who is hypotensive through blood loss should be, therefore, considered to have lost a significant volume. Choice of resuscitative fluid is usually balanced salt solution such as ringer lactate, although hypertonic saline has also been tried in such situations (Prougn, 2005). Though hypertonic saline could additionally reduce ICP, there is associated risk of severe hypernatremia; it should be considered in refractory cases only, who no longer respond to mannitol or developing uremia (Prougn, 2005; De Vivo et al., 2001).

6.2 Control of Intracranial pressure

Various studies indicate that sustained rise in ICP over 20 mm Hg results in a poor outcome (Marshall et al., 1983; Howard et al., 1988; Narayan et al., 1982). Jiang JY et al (2002) showed that mortality rates were 14% if ICP rise was less than 20 mm Hg, but 34% if ICP was greater than 30 mm Hg at 48 hrs. For this reason, the brain trauma foundation recommends for initiating decrease in ICP, at a threshold rise of
Figure 6: Protocol for management of intracranial herniation

20 mm Hg with level II recommendation (Bratton et al., 2007). Various researches have documented that CPP levels of less than 60-70 mm Hg adversely affect the cerebral oxygenation and metabolism; what is the safe range of CPP still remains controversial, with the consensus that it should be 60-70 (Unterberg et al., 1997; Vespa et al., 1998, Bullock et al., 1996). However, CPP levels of less than 50 mm Hg should always be avoided, as recommended by brain trauma foundation (Bratton et al., 2007). Various manoeuvres used for lowering of ICP include head end elevation of bed, hyperventilation, and use of osmotic agents, diuretics, barbiturates, CSF drainage and hypothermia. Other agents like procaine derivatives, indomethacin, propofol, hyperbaric oxygen and tromethamine have been tried in the past, but in absence of convincing results, not used in routine clinical practice. Role of sedatives still remain controversial, with no clear advantage in absence of anxiety or agitation. Moreover, use of sedatives or paralytics is associated with increased ICU stay and added complications (Hsiang et al., 1994).

6.2.1 Hyperventilation

Hyperventilation is known to lower arterial carbon dioxide tension, resulting in diffuse cerebral vasoconstriction (approximately 3% for every 1 mm Hg decrease in PaCO2), decreasing cerebral vascular volume
and lowering ICP (Robertson, 2004; Kochanek et al., 2012). Thus, hyperventilation can temporarily revert the constellation of symptoms observed in acute rise in ICP, until investigations are performed and the definitive treatment is initiated (Robertson, 2004; Kochanek et al., 2012). However, cerebral vasoconstriction is only effective in unaffected areas of the brain; therefore hyperventilation may be less effective in patients with diffuse brain injury than in those with focal lesions. Cerebral vasoconstriction in hyperventilation could result in brain ischemia (Andrews, 2003). Therefore, hyperventilation should only be instituted for short periods where immediate control of ICP is necessary and the target PaCO2 levels should be maintained between 30-35 mm Hg (Cruz, 1993). In some situations, hyperventilation may be necessary for longer periods in patients with persistently high ICPs, who do not respond to other treatment modalities. However, these patients should be regularly monitored for adequate cerebral oxygenation and tissue perfusion by utilizing various neuromonitoring modalities such as jugular bulb venous oxygen saturation, brain tissue oxygen tension, transcranial doppler ultrasonography and near infrared spectroscopy assessments, with level III recommendation (Cruz, 1993; Robertson, 1995; Lewis, 1996; Cruz, 1998; Dosemeci et al., 2004; Robertson, 2004; Stiefel et al., 2005; De Freitas & Andre, 2006; Rosenthal et al., 2008; Kochanek et al., 2012). Hyperventilation is highly controversial, with the 2012 Cochrane database concluding that hyperventilation should not be used in the first 24 hours after TBI when cerebral blood flow (CBF) is generally lowest, with level III recommendation and prophylactic hyperventilation (PaCO2<25 mm Hg) is not recommended, with level II recommendations (Kochanek et al., 2012).

6.2.2 CSF Drainage

Ventricular catheterization should be done wherever possible, if the ventricles are compressed or displaced. Drainage of even small amounts of CSF is sufficient to produce required fall in ICP and a rise in CPP (Gahjar et al., 1993). However if the systolic blood pressure is dependent on continued stimulus by the Cushing response, then sudden decrease of ICP may precipitate hypotension. This can be precluded by IV fluid preloading and setting the CSF drainage pressure to about 20 cm H2O (Gahjar et al., 1993). However, continuous CSF drainage is not recommended as it may lead to collapse of the ventricles and catheter malfunction.

6.2.3 Osmotic agents and Renal diuretics

From many decades, osmotic agents remained the primary means of controlling the raised ICP, yet their exact mechanism of action is still debated. Mannitol, a sugar alcohol solution, is an osmotic diuretic which causes increase in intravascular volume and reduction in blood viscosity (Muizelaar et al., 1983; Burke et al., 1981; Schrot & Muizelaar, 2005). To maintain a constant CBF, this leads to acute cerebral vasoconstriction with consequent rapid reduction in ICP (Muizelaar et al., 1984, Leech & Miller, 1974). More prolonged reduction in ICP is due to osmotic effect, by drawing fluid into the vascular compartment. Mannitol reduces red blood cell rigidity, allowing them to pass through small blood vessels and areas of marginal perfusion (Burke et al., 1981). It also acts as a free radical scavenger, which have been implicated in ischemic brain injury. Mannitol is not metabolized and remains in intravascular space, unless high concentrations or continuous infusion breaches the normal blood–brain barrier, allowing it to enter the interstitial space, taking fluid with it and causing a ‘rebound’ rise in ICP (Kaufmann & Cardoso, 1992). Conventionally, it is administered as a 20% solution in a bolus dose of 0.25–1.0 g/kg given over 10–20 min, to avoid the risk of hypotension from rapid and continuous infusion. ICP falls within 5–10 minutes. The maximum effect occurs in about 60 minutes and the total effect last for 3–4 hours (James et
al., 1977). Previous studies show that higher doses of mannitol (1.4 g/kg) are more effective in obtaining brain relaxation than conventional doses, in patients with brain herniation (Cruz et al., 2001; Andrews, 2003). These observations support the use of higher dosage mannitol in patients with transtentorial herniation. However, it also has some limitations including progressive dehydration, hypotension, prerenal failure and expansion of an intracranial hemorrhage by brain shrinkage (Cottrell et al., 1977; Feig & McCurdy, 1977). To avoid the risk of hypotension, its use is prohibited in condition of haemorrhagic shock (Andrews, 2003). Central venous pressure, urine output, serum electrolytes and blood osmolality need to be carefully monitored, particularly if it is combined with the renal loop diuretics. In combination with mannitol, furosemide produces a synergistic effect in reduction in ICP and also increases the duration of its effect (Pollay et al., 1983; Wilkinson & Rosenfeld, 1983). However, the considering the added risk of hypovolemia and renal failure, their combined use should be restricted for patients with associated cardiac failure or pulmonary edema.

6.2.4 Barbiturates

Barbiturate causes a decrease in cerebral metabolic rate; reduces CBF, decreases production of CSF, increases absorption of CSF, and causes burst suppression of EEG. This leads to a parallel fall in ICP (Michenfelder, 1974). To observe significant reduction in ICP, anesthetic equivalent doses are required and therefore cardiorespiratory and ICP monitoring is compulsory. Considering the risk of hypotension and respiratory depression, barbiturate therapy should be reserved as a step-down protocol therapy for ICP control, once other means of medical management have failed (Ward et al, 1985).

6.2.5 Corticosteroids

Corticosteroids lower ICP by decreasing vasogenic edema through inhibition of phospholipase A2, stabilization of lysosomal membranes, reduction of edema producing vascular endothelial growth factor expression, and improvement in microcirculation (Yamada et al., 1989; Heiss et al., 1996; Machein et al., 1999). Corticosteroids play an important role in decreasing edema secondary to brain tumors, however not recommended for cytotoxic edema secondary to stroke or head injury (Yamada et al, 1989; Ruderman & Hall, 1965; Alderson & Roberts, 2005). In fact, systemic complications of steroids like gastrointestinal bleeding, and glucose intolerance (50% incidence) can further worsen the patient’s condition (Gotzsche, 1994; Meyer, 2003; Conn & Poynard, 1994). Patients treated with systemic steroids are also at risk of infection with Pneumocystis carinii pneumonia; the reported incidence is about 2%. Prophylactic control with trimethoprim–sulfamethoxazole may prevent this complication, but is mainly utilized for immunocompromised patients (Kovacs, 2001). Patients with extensive brain edema may also develop withdrawal symptoms such as headache, lethargy, low-grade fever and adrenal insufficiency after rapid tapering of corticosteroids, thus leading to increased morbidity and mortality (Amatruda, 1965). The results of the large CRASH trial suggest that steroids should not be used in head injury, as they appear to increase mortality. However, the exact mechanism of this effect still remains unclear (CRASH Trial Collaborators, 2003). Current Cochrane review recommends for need to reassess the pathophysiologic understanding of the traumatic brain injury (TBI) and no need to start such future trials (Alderson & Roberts, 2005).

6.2.6 Hypothermia

Recent studies suggest that moderate hypothermia (32-34°C) can reduce ICP and is cytoprotective in cases of global ischemic injury. It is also known to prevent glutamate release and depletion of high-energy
phosphate compounds (Chopp et al., 1991; Busto et al., 1989; Marion et al., 1993). Although known to cause coagulopathy, there have been no reported significant complications with moderate hypothermia in the studies reported so far (Metz et al., 1996; Resnick et al., 1994). Thus, hypothermia could be a safe additional treatment in patients with high ICP.

6.3 Monitoring

Monitoring is a vital component in the optimal management of patient. It allows for early detection of intracranial mass lesion, guidance for therapy, early diagnosis of secondary complications, and determination of prognosis. Therefore, monitoring of patients with brain herniation, either traumatic or non-traumatic must comprise both general and specific neurologic monitoring.

6.3.1 General Monitoring

General parameters that should be repeatedly monitored include electrocardiography, arterial oxygen saturation, capnography, arterial blood pressure, central venous pressure, systemic temperature, urine output, arterial blood gases, and serum electrolytes and osmolality. Invasive or non-invasive cardiac output monitoring may be required in haemodynamically unstable patients who do not respond to fluid resuscitation and vasopressors therapy.

6.3.2 Neurological Monitoring

The brain trauma foundation guidelines recommends for ICP monitoring in all salvageable patients with severe TBI and an abnormal CT scan, and in patients with severe TBI with a normal CT scan, if two or more of the following features are noted at admission: age over 40 years, unilateral or bilateral motor posturing, or systolic blood pressure < 90 mm Hg (Bullock, 2007). Indications of ICP monitoring in patients with brain tumours are not clearly defined. It is mostly employed in cases with extensive cerebral oedema having brain herniation. Currently available methods of ICP monitoring include ventricular catheters, subdural-subarachnoid bolts or catheters, epidural transducers and intraparenchymal fiberoptic devices. The ventricular ICP catheter is the gold standard and preferred for ICP monitoring whenever possible. It is the most accurate, low-cost, and reliable technique of monitoring ICP. Additional benefits include provision for continuous ICP monitoring and therapeutic CSF drainage in the event of raised ICP (Marshman, 2001). ICP monitoring should be continued till the patient remains on intensive treatment or there is risk of secondary deterioration. This period usually last up to two weeks after injury. Advantages of ICP monitoring include early detection of intracranial mass lesion, guidance for therapy and prevention of irrational use of therapies to control ICP, CSF drainage with reduction of ICP and improvement of CPP, and determination of prognosis. Although ICP measurements have become an integral component of monitoring in patients with TBI, there is contradictory literature regarding improved outcome with this manoeuvre in acute coma, whether traumatic or non-traumatic (Saul & Ducker, 1982; Eisenberg et al., 1988; Howells et al., 2005; Aarabi et al., 2006; Timofeev et al., 2006; Bulger et al., 2002; Lane et al., 2000; Mauritiz et al., 2008; Stocchetti, 2001; Cremer et al., 2005; Cremer, 2008; Shaﬁ et al., 2008; Randall et al., 2012). Potential complications of ICP monitoring include infection, haemorrhage, and catheter malfunction or malposition. However, routine administration of prophylactic antibiotic after ventricular catheter placement is not recommended to reduce infection (Bullock, 2007).
6.4 Surgical Management

As soon as initial steps are over, a diagnostic brain CT should be performed immediately to identify the clinical cause; if not done so far because of hemodynamic instability (Andrews, 2003). CT scan is a vital tool for identification of intracranial mass lesions such as tumour, oedema, haemorrhage, or hydrocephalus. After obtaining the definitive diagnosis, if the patient has remained haemodynamically stable, exploratory burr-hole may be performed as an emergency measure on the same side of lateralizing signs such as dilated pupil, to explore the expanding mass lesion like epidural/subdural haemorrhage and immediate surgical evacuation should be done, if feasible (Andrews et al., 1986; Chesnut et al., 1994). In case of non-lateralizing signs of herniation, this procedure has to be considered bilaterally. Intraoperative ultrasonic brain imaging can further improve the utility of exploratory burr-holes, allowing for accurate identification and management of intracranial hematomas (Andrews et al., 1990). In patients with haemodynamic instability, it is prudent to start ICP monitoring rather than performing exploratory burr holes, considering the lower probability of intracranial mass lesions in such patients. If the ICP is low, then no further intervention is required except for general stabilization of the patient. However, if the ICP is elevated, then further diagnostic imaging may be required, to decide for surgical exploration for such patients. If craniectomy is performed, it should be targeted to fully decompress the brain parenchyma. In case of non-traumatic mass lesion, immediate surgical decompressive craniotomy may be indicated for extensive cerebral oedema secondary to upward transtentorial or tonsillar herniation. For an external cerebral herniation, urgent surgical repair is mandatory to avoid any serious complications. The primary goal of surgery is to disconnect the encephalocele and perform dural repair. Various materials have been used for the dural repair includes fascia lata, temporalis fascia, lyophilised dura, or a biosorbable polyglycolic acid sheet (Jackson et al., 1997). Other potential management tools include insertion of vascular cushions adjacent to large draining veins at the bone margin to reduce risk of venous ischemia (Stiver, 2009). Repair of the bony defect with bone or cartilage has also been described (Feenstra et al, 1985). However, this is not always performed as removal of the herniated brain tissue and prevention of CSF leak will in itself allow the bony defect to heal and thereby preventing recurrence.

6.5 Prognosis

The prognosis of patients with brain herniation syndromes depends mainly upon the age of presentation, initial GCS scores, pupillary response and size, hypoxia, hyperthermia, high intracranial pressure, aetiology, site and severity of brain herniation and how soon it is being attended (Jiang et al., 2002). Recovery is better in patients of younger age group and with initial high GCS scores. In patients with a treatable cause like acute epidural or subdural hematoma, there are high chances of survival, if initial resuscitation and corrective management are initiated rapidly (Duffy, 1982; Andrews et al., 1986). However, patients with traumatic transtentorial herniation have an overall mortality rate of 70%. Patients with non-traumatic causes of herniation (acute hydrocephalus, tumour-related cerebral oedema, temporal lobar haemorrhage, hemispheric infarction or cerebello-tonsillar herniation from lumbar drainage) having intact brain functions carry better prognosis and functional recovery, if appropriate resuscitative measures and surgical decompression of mass effect are instituted in a timely fashion (Dagniew et al., 2002; Muhonen & Zunkeler, 1994; Weinberg et al., 2003; Ross et al., 1989; Andrews, 2005). The brain herniation can itself cause massive stroke which can lead to compression of cardiorespiratory centres in the brain stem, and consequent mortality. Brain herniation into temporal lobe or the cerebellum always carries a grave prognosis. However, outcome patterns are variable for other areas of the brain.
References


