NEUROSURGERY

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INTRACRANIAL MASS LESIONS

INTRACRANIAL DYNAMICS

Intracranial Pressure/Volume Relationship

- ❑ intracranial volume is constant in the adult
  - $V_{brain} + V_{blood} + V_{csf} = V_{skull}$ = constant (Monro-Kellie hypothesis)
- ❑ as lesion expands, Intracranial pressure (ICP) does not rise initially
  - CSF, blood, some brain water displaced out of the head
  - brain tissue may shift into compartments under less pressure (herniation)
- ❑ ICP then rises exponentially

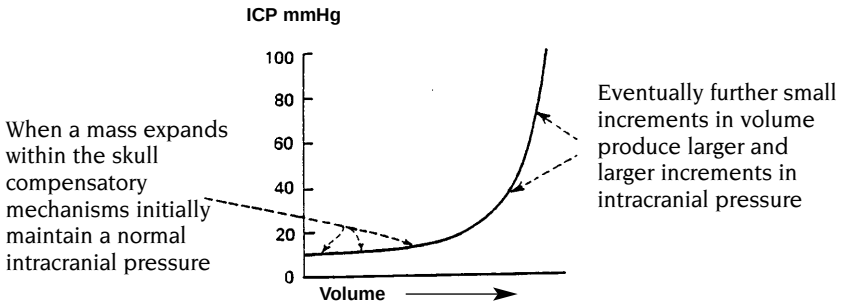

Figure 1. ICP-Volume Curve

Adapted from Lindsay KW, Bone I, Callander, R: Neurology and Neurosurgery Illustrated

- ❑ normal ICP ~ 6-15 mm Hg (8-18 cm H_2O) for adult, 3-7 mm Hg (4-9.5 cm H_2O) for child and varies with patient position
- ❑ consider therapy for high ICP when ICP > 20-25 mm Hg

ICP Measurement

- lumbar puncture (contraindicated with known/suspected intracranial mass lesion)
- ventricular catheter ("gold standard", also permits therapeutic drainage of CSF to decrease ICP)
- intraparenchymal monitor
- subdural/subarachnoid monitor (Richmond bolt)

Cerebral Blood Flow (CBF)

- ❑ CBF depends on cerebral perfusion pressure (CPP) and cerebral vascular resistance (CVR)
- ❑ CPP = MAP (mean arterial pressure) – ICP (intracranial pressure)
  - Normal CPP > 50 mm Hg in adults
- ❑ cerebral autoregulation maintains constant CBF by compensating for changes in CPP, unless
  - high ICP such that CPP < 40 mm Hg
  - MAP > 160 mm Hg or MAP < 60 mm Hg
  - brain injury: i.e. subarachnoid hemorrhage (SAH), severe trauma

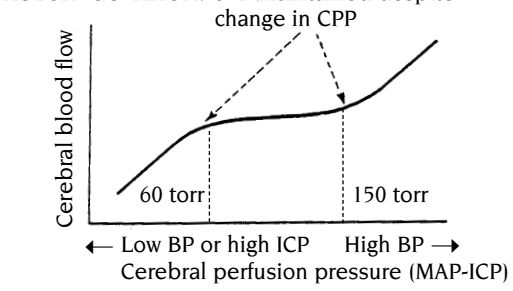

Figure 2. Cerebral Autoregulation Curve

Adapted from Lindsay et al: Neurology and Neurosurgery Illustrated

❑ causes of raised ICP
- increased intracranial blood volume
  - hypoventilation —> increased pCO_2 —> vasodilatation —> decreased pO_2 —> (< 60) —> vasodilatation
  - decreased venous drainage
  - venous sinus thrombosis
  - superior vena cava (SVC) syndrome
  - cranial dependency
  - valsalva
- cerebral edema
- hydrocephalus
- intracranial mass lesion (tumour, pus, blood – see section below)
- status epilepticus
- systemic hypertension

HERNIATION SYNDROMES

1. Subfalcine
2. Central
3. Uncal
4. Upward
5. Tonsillar

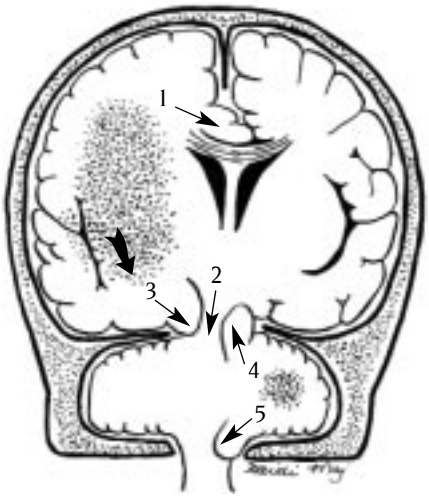

Figure 3. Herniation Types

Illustration by Heidi Maj

1. Subfalcine (Cingulate) Herniation
❑ definition: cingulate gyrus herniates under falx
❑ cause: lateral supratentorial lesion
❑ clinical presentation
- usually asymptomatic
- pathological/radiological observation
- warns of impending transtentorial herniation
- rarely, frontal infarct due to kinked anterior cerebral artery (ACA)

2. Central Tentorial (Axial) Herniation
❑ definition: displacement of diencephalon and midbrain through tentorial notch (often gradual)
❑ cause: supratentorial midline lesion, diffuse cerebral swelling, late uncal herniation
❑ clinical presentation
- rostral to caudal deterioration (sequential failure of diencephalon, midbrain, pons, then medulla)
- decreased LOC (midbrain compressed)
- EOM/upward gaze impairment ("sunset eyes", pressure on superior colliculus in midbrain compresses 3rd nerve nucleus)
- brainstem hemorrhage ("Duret's", secondary to shearing of basilar artery perforating vessels)
- diabetes insipidus (traction on pituitary stalk and hypothalamus) - this is an end stage sign

3. Lateral Tentorial (Uncal) Herniation
- ❑ definition: uncus of temporal lobe herniates down through tentorial notch
- ❑ cause: lateral supratentorial lesion (often rapidly expanding traumatic hematoma)
- ❑ clinical presentation
  - unilateral dilated pupil (earliest, most reliable sign), followed by extraocular muscle (EOM) paralysis (ipsilateral cranial nerve III compressed)
  - decreased level of consciousness (LOC) (midbrain compressed)
  - contralateral hemiplegia, +/– extensor plantar response
  - +/– "Kernohan's notch": contralateral cerebral peduncle compressed due to shift of brain —> ipsilateral hemiplegia (a false localizing sign)

4. Upward Herniation
- ❑ definition: cerebellar vermis herniates through tentorial incisura, causing midbrain compression
- ❑ cause: posterior fossa mass
- ❑ clinical presentation
  - SCA compression —> cerebellar infarct
  - Compression of cerebral aqueduct —> hydrocephalus

5. Tonsillar Herniation ("Coning")
- ❑ definition: cerebellar tonsils herniate through foramen magnum
- ❑ cause: infratentorial lesion or following central tentorial herniation
- ❑ clinical presentation
  - rapidly fatal (compression of cardiovascular and respiratory centers in medulla)
  - may be precipitated by lumbar puncture (LP) in presence of space occupying lesion (particularly in the posterior fossa)

CLINICAL FEATURES

Acute Raised ICP
- ❑ headache
- ❑ nausea and vomiting (N/V)
- ❑ decreased LOC
- ❑ change in Glasgow Coma Scale (GCS) best index to monitor progress and predict outcome of acute intracranial process (see Neurology Chapter)
- ❑ papilledema **(see Colour Atlas OP21)**
  - may take 24-48 hours to develop
- ❑ abnormal EOM
  - CN VI palsy
    - longest intracranial course
    - causative mass may be remote from nerve root, i.e. CN VI palsy can be a false localizing sign
    - upward gaze palsy (especially in children with obstructive hydrocephalus)
- ❑ Cushing's Triad (full triad seen in 1/3 of cases)
  - increased blood pressure (BP), decreased HR (late finding), abnormal respiratory pattern (Cheyne Stokes, apneustic, ataxic)
- ❑ signs/symptoms of herniation syndromes
- ❑ focal signs/symptoms due to responsible lesion

Chronic Raised ICP
- ❑ headache
  - postural: worsened by coughing, straining, bending over (Valsalva)
  - morning headache (H/A): worse on waking in the morning (increased CO_2) or waking with headache in night
- ❑ visual changes
  - enlarged blind spot, preserved vision (until extremely advanced, then episodic constrictions of visual fields, i.e. "gray-outs")
  - long standing papilledema (not necessarily present) may produce optic atrophy and blindness
- ❑ differentiate from papillitis (usually unilateral with decreased visual acuity)

Imaging Features
- ❑ CT and MRI: key diagnostic investigation
  - enlarged ventricles - hydrocephalus
  - compressed ventricles with midline shift - mass lesion
- ❑ skull x-rays (academic) in chronic ICP may show
  - separation (diastasis) of sutures in infants
  - digital markings in skull vault from compression of brain matter against bone ("beaten copper cranium")
  - thinning of dorsum sellae

MANAGEMENT

- ❑ goals
  - keep ICP < 20-25 mm Hg
  - keep CPP > 70 mm Hg
- ❑ elevate head
  - head of bed at 30-45 degrees —> decreases intracranial venous pressure
- ❑ ventilate/hyperventilate (pCO_2 30 ± 2 mm Hg)
  - decreases pCO_2, increases pO_2, decreases venous pressure
- ❑ mannitol (20% IV solution, 1 gm/kg)
  - can give rapidly, effects in 30 minutes, (see Drugs section)
  - maintain sBP > 90 mm Hg
- ❑ corticosteroids
  - decreases vasogenic edema around brain tumour, abscess
  - no proven value in head injury or stroke
  - works slowly (days)
- ❑ identify etiology (CT, MRI)
- ❑ surgery
  - remove mass lesion
  - remove CSF by external ventricular catheter drain (if acute) or shunt
  - Note: lumbar puncture contraindicated when known/suspected intracranial mass lesion

BENIGN INTRACRANIAL HYPERTENSION (PSEUDOTUMOUR CEREBRI)

- ❑ incidence ~0.5/100,000/year
- ❑ raised intracranial pressure and papilledema without evidence of any "mass" lesion, hydrocephalus, infection or hypertensive encephalopathy
- ❑ diagnosis of exclusion

Etiology

- ❑ unknown (majority), but associated with
  - diet: obesity, hyper/hypovitaminosis A
  - endocrine: menarche, menstrual irregularities, Addison's disease
  - hematological: iron deficiency anemia, polycythemia vera
  - drug: steroid withdrawal, tetracycline, nalidixic acid

Clinical Features

- ❑ usually in 3rd and 4th decade (F>M)
- ❑ symptoms and signs of raised ICP (headache in > 90%), except no decreased LOC
- ❑ radiological (CT or MRI) and CSF studies - normal (ventricles may be smaller)
- ❑ usually self-limited, recurrence is common, chronic in some patients
- ❑ a preventable cause of (often permanent) blindness from optic atrophy
- ❑ risk of blindness is not reliably correlated to duration of symptoms, papilledema, headache, visual acuity or number of recurrences

Differential Diagnosis

- ❑ true mass lesions (see Intracranial Mass section)
- ❑ venous outflow obstruction to CSF absorption
  - dural sinus thrombosis, jugular vein or sigmoid sinus obstruction
  - intrathoracic mass lesion
  - superior vena cava syndrome
  - congestive heart failure
  - hyperviscosity syndromes
- ❑ infections
- ❑ inflammatory conditions: e.g. neurosarcoidosis, SLE
- ❑ vasculitis
- ❑ metabolic conditions: e.g. lead poisoning
- ❑ pseudopapilledema associated with hyperopia and drusen
- ❑ meningeal carcinomatosis
- ❑ Guillain-Barre syndrome
- ❑ following head trauma

Management

- ❑ R/O conditions that may mimic benign intracranial hypertension
- ❑ D/C offending medications, weight loss, fluid/salt restriction
- ❑ drugs - acetazolamide (decreased CSF production)
  - thiazide diuretic or furosemide
- ❑ if above fail —> serial LPs, lumboperitoneal shunt, VP shunt
- ❑ optic nerve sheath fenestration - if progressive impairment of visual acuity despite treatment
- ❑ 2 year follow-up with imaging studies to rule out occult tumour

HYDROCEPHALUS

❑ **see Colour Atlas NS1**
❑ definition: increased CSF volume, decreased CSF absorption
❑ normal CSF volume = 100 - 150 mL (50 in ventricles, 25 around brain, 75 around spinal cord)
❑ CSF production is constant at 0.4-0.6 cc/hr

MECHANISMS

❑ increased CSF production
  - e.g. choroid plexus papilloma (0.4-1% of intracranial tumours)

❑ decreased CSF absorption (see below)

CLASSIFICATION

Obstructive (Non-Communicating) Hydrocephalus

❑ absorption is blocked within ventricular system proximal to the arachnoid granulations
❑ causes/location of block
  - intraventricular hemorrhage
  - ventricular tumours (e.g. 3rd ventricle colloid cyst)
  - supratentorial mass causing tentorial herniation and aqueduct compression
  - infratentorial mass causing 4th ventricle or aqueduct obstruction
  - congenital e.g. aqueductal stenosis, Dandy-Walker malformation, or Chiari malformation (see Pediatric Neurosurgery section)

❑ CT findings
  - lateral and 3rd ventricles dilated
  - normal 4th ventricle (e.g. aqueduct stenosis) or deviated/absent 4th ventricle (e.g. posterior fossa mass)

Communicating (Non-Obstructive) Hydrocephalus

❑ absorption is blocked at some part of extraventricular pathway, such as arachnoid granulations
❑ causes
  - meningitis
  - SAH
  - trauma

❑ CT findings
  - all ventricles dilated

Normal Pressure Hydrocephalus (NPH)

❑ gradual onset of classic triad
  - incontinence
  - gait apraxia/ataxia
  - dementia

❑ CSF pressure often within clinically "normal" range
❑ usually communicating

Hydrocephalus Ex Vacuo

❑ enlargement of ventricles (and sulci) secondary to diffuse brain atrophy
❑ usually a function of normal aging
❑ not true hydrocephalus

CLINICAL FEATURES

Acute Hydrocephalus

❑ signs and symptoms of acute raised ICP
❑ usually obstructive type

Chronic Hydrocephalus

❑ similar to NPH

INVESTIGATIONS

CT

❑ ventricular enlargement, may see prominent temporal horns
❑ periventricular lucency (CSF forced into extracellular space)
❑ narrow/absent sulci, +/– 4th ventricular enlargement

Ultrasound (through anterior fontanelle in infants)

❑ ventricular enlargement

MANAGEMENT

- ❑ spinal taps (for transient, communicating hydrocephalus)
- ❑ remove obstruction (if possible)
- ❑ excision of choroid plexus papilloma
- ❑ third ventriculostomy (for obstructive hydrocephalus)
- ❑ shunts
    - ventriculoperitoneal (VP) = ventricle to peritoneum
    - ventriculopleural = ventricle to pleura
    - ventriculo-atrial (VA) = ventricle to right atrium
    - lumboperitoneal = lumbar spine to peritoneum (for communicating hydrocephalus and pseudotumour cerebri)

Shunt Complications

- ❑ obstruction
    - etiology: infection, obstruction by choroid plexus, buildup of proteinaceous accretions, blood, cells (inflammatory or tumour)
    - signs and symptoms of acute hydrocephalus or increased ICP
    - radiographic evaluation: "shunt series" (plain x-rays which only show disconnection of tube system), CT, isotope shunt study (nuclear medicine)
- ❑ infection (3-4%)
    - etiology: S. epidermidis, S. *aureus*, gram-negative bacilli
    - presentation: fever, nausea and vomiting, anorexia, irritability; signs and symptoms of shunt obstruction; shunt nephritis (antibodies generated against bacteria in shunt leads to kidney damage)
    - investigation: CBC, blood culture, shunt tap (lumbar puncture (LP) usually NOT recommended in obstructive hydrocephalus)
- ❑ overshunting
    - slit ventricle syndrome (collapse of ventricles leading to shunt catheter occlusion by ependymal lining)
    - subdural effusion, hygroma, hematoma
    - secondary craniosynostosis (children)
    - low pressure headache
- ❑ seizures

INTRACRANIAL MASS

- ❑ **see Colour Atlas NS15**
- ❑ differential diagnosis: "tumour, pus or blood", cyst
- ❑ history important for localizing and differentiating mass lesions
- ❑ important features on CT (with and without contrast enhancement)
    - lesions (+/– edema, necrosis, hemorrhage)
    - midline shifts and herniations
    - effacement of ventricles and sulci (often ipsilateral), basal cisterns

TUMOUR (see Colour Atlas NS16 and NS17)

- ❑ primary versus metastatic
- ❑ primary tumours (benign or malignant) rarely metastasize
- ❑ presenting symptoms
    - local effects
        - dependent on site: focal deficits, lobe syndromes, seizures, headaches
    - raised ICP
        - acute or chronic depending on tumour growth rate (see Raised ICP section)
    - sudden onset of symptoms after hemorrhage (5-10%)
- ❑ consider by
    - location (supratentorial vs. infratentorial)
    - age (adult vs. child)

Table 1. Tumour Types: Age, Location and Clinical Features

	Supratentorial	Infratentorial
Children (< 15 years, primarily infratentorial - 80%)	Astrocytoma - all grades Craniopharyngioma Ependymoma Other: dermoid/epidermoid, pineal tumours, primitive neuroectodermal tumours	**Cerebellar astrocytoma** **Medulloblastoma** Ependymoma Choroid plexus papilloma Brain stem astrocytoma
Adult (> 15 years, primarily supratentorial - 80%)	**Astrocytoma** (40-50%) **Metastatic** (20-30%) **Meningioma** (15%) Pituitary ademona (5%) Oligodendroglioma (5%)	**Metastatic** (20-30%) Schwannoma (6%) e.g. vestibular schwannoma Medulloblastoma (5%) Hemangioblastoma
Signs and Symptoms	Raised ICP Focal or lobar effects Seizures Mental status changes Personality changes Visual field deficits Endocrine disturbances (with pituitary tumour) Speech, motor and sensory deficits	Raised ICP Local effects in posterior fossa Extremity ataxia Truncal ataxia CN palsy - often multiple Nystagmus Gait disturbance Vertigo

Investigations
- ❑ CT, MRI, stereotactic biopsy (tissue diagnosis)

Management
- ❑ conservative
  - serial history, physical, imaging for slow growing/benign lesions
- ❑ medical
  - corticosteroids to reduce vasogenic cerebral edema
  - pharmacological treatment for pituitary tumours (see Pituitary Adenoma section)
- ❑ surgical
  - excisional: total, partial, decompressive, palliative
  - shunt if CSF flow is blocked
- ❑ radiotherapy - stereotactic radiosurgery (Gamma-knife, Linear Accelerator)
- ❑ chemotherapy – e.g. alkylating agents (temozolomide)

Metastatic Tumours (see Colour Atlas NS19)
- ❑ most common brain tumour seen clinically
  - 15% of cancer patients present with cerebral mets
  - source
    - Lung 44% (especially bronchogenic cancer)
    - Breast 10%
    - Kidney 7% (renal cell carcinoma (RCC))
    - GI 6%
    - Melanoma 3%
- ❑ route of spread – hematogenous
- ❑ location – 3/4 are supratentorial, often at grey-white matter junction
- ❑ diagnosis: metastatic work-up (CXR, CT chest/abdo, abdominal U/S)
  - CT with contrast (round, well-circumscribed uniformly lesion)
  - consider biopsy (as up to 10% may not be cerebral met in patient with cancer history) and patient may not have a cancer history
- ❑ prognosis: median survival with optimal Rx 26-32 weeks but varies depending on primary
- ❑ treatment: palliative
  - single accessible lesion —> surgical excision + radiation
  - multiple lesions —> whole brain radiation

Astrocytoma
❑ most common primary brain tumour (45-50%)

Table 2. Grading (Multiple Systems)

Kernohan Grade	National Brain	CT Findings Tumour Study Group	MRI Findings	Survival
I	Low grade	Low density	Abnormal signal	8-10 yrs
II	Low grade	+ mass effect	+ mass effect	7-8 yrs
III	Anaplastic	Complex enhancement	Complex enhancement	2 yrs
IV	GBM	Necrosis (ring enhancement)	Necrosis (ring enhancement)	< 1 yr

• GBM: anaplastic astro: low grade = 5:3:2

❑ "cystic cerebellar" astrocytoma
- pediatric population, infratentorial
- potentially curable

❑ clinical presentation: middle aged, recent onset of new, worsening H/A, N/V, +/– focal deficits or symptoms of increased ICP
❑ diagnosis: CT (see Figure 4), MRI with contrast +/– biopsy
❑ therapy:
- surgery: not curative, aim to prolong "quality" survival
- radiotherapy prolongs survival (retrospective evidence)
- chemotherapy (alkylating agents)

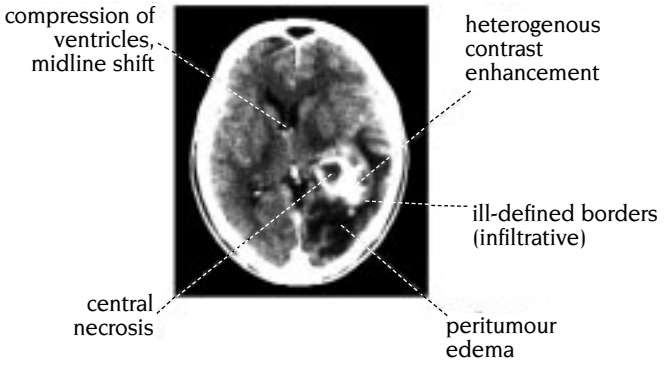

Figure 4. Malignant Astrocytoma on CT

Meningioma (see Colour Atlas NS16)
❑ mostly benign (1% malignant), slow-growing, non-infiltrative
❑ common locations: parasagittal convexity, sphenoid wing, falx
❑ presentation: middle aged, symptoms of increased ICP, focal symptoms depend on location
❑ diagnosis: MRI, CT with contrast (see Figure 5)
❑ therapy
- conservative management for slow-growing lesions
- surgery is treatment of choice (curative if complete resection)
- radiotherapy – ineffective

❑ prognosis: > 90% 5-yr survival

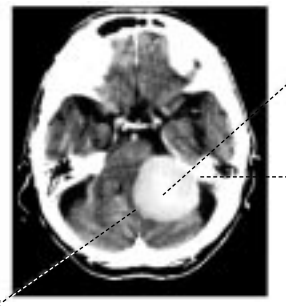

Figure 5. Meningioma on CT

Vestibular Schwannoma ("Acoustic Neuroma")

- ❑ progressive unilateral deafness = acoustic neuroma until proven otherwise
- ❑ slow-growing (average of 1-10 mm/yr), benign posterior fossa tumour
- ❑ arises from vestibular component of CN VIII at cerebello-pontine angle (CPA)
- ❑ clinical presentation: compression of structures in CPA
  - often CN VIII symptoms, then V, then VII
  - CN VIII: unilateral sensorineural deafness, tinnitus, dysequilibrium
  - CN V: facial numbness, loss of corneal reflex
  - CN VII: facial weakness (uncommon pre-operatively)
  - cerebellum: ataxia, nystagmus
- ❑ diagnosis
  - MRI, CT (contrast enhancing mass in CPA)
  - audiogram, BAEP (brainstem auditory evoked potentials), caloric tests
  - if bilateral: neurofibromatosis type II
- ❑ management
  - conservative: serial imaging
  - surgery: several routes, curable if complete resection (almost always possible)
  - stereotactic radiosurgery: gamma-knife, linear accelerator
  - significant post-therapy morbidity: CN VII, VIII dysfunction (only significant disability if bilateral), CSF leak

Pituitary Adenomas (see Colour Atlas NS18)

- ❑ primarily from anterior pituitary, 3rd-4th decade, M=F
- ❑ may be functional (secretory) or non-functional
- ❑ clinical presentation
  - a) mass effects
    - H/A
    - bitemporal hemianopsia (compression of optic chiasm)
    - CN III, IV, V1, V2, VI palsy (compression of cavernous sinus)
  - b) endocrine effects
    - hyperprolactinemia —> infertility, amenorrhea, galactorrhea, impotence
    - ACTH production —> Cushing's disease
    - GH production —> acromegaly
    - panhypopituitarism (hypothyroidism, hypoadrenalism, hypogonadism)
  - c) apoplexy (abrupt onset H/A, visual disturbances, ophthalmoplegia, and reduced mental status) and CSF rhinorrhea (rare presenting signs of pituitary tumour)
- ❑ diagnosis: formal visual fields, endocrine tests (PRL level, TSH, cortisol, fasting glucose, FSH/LH, IGF-1), imaging (MRI)
- ❑ differential: parasellar tumours (e.g. craniopharyngioma, tuberculum sellae meningioma), carotid aneurysm
- ❑ treatment
  - medical
    - dopamine agonists (e.g. bromocriptine) for prolactinoma
    - serotonin antagonist (cyproheptadine), inhibition of cortisol production (ketoconazole) for Cushing's
    - somatostatin analogue (octreotide) +/– bromocriptine for acromegaly
    - endocrine replacement therapy
  - surgical
    - trans-sphenoidal, transethmoidal, transcranial approaches

PUS

Brain Abscess (see Colour Atlas NS8)

❑ etiology
- local spread (adjacent infection)
  - otitis media, mastoiditis, sinusitis
  - osteomyelitis
  - dental abscess
- hematogenous spread
  - adults: lung abscess, bronchiectasis, empyema
  - children: cyanotic heart disease with R to L shunt (blood is shunted away from lungs preventing filtration of bacteria)
  - immunosuppression (AIDS - toxoplasmosis)
- dural disruption
  - surgery, trauma
  - congenital defect, e.g. dermal sinus
- pathogens
  - *Streptococci* (most common), often anaerobic or microaerophillic
  - *Staphylococci* (penetrating injury)
  - Gram negatives, anaerobes
  - Toxoplasmosis and Nocardia in immunocompromised hosts

❑ diagnosis
- focal neurological signs and symptoms
- mass effect, increased ICP and sequelae
- seizures
- +/– signs of systemic infection (mild fever, leukocytosis)
- blood cultures rarely helpful, LP not helpful and contraindicated
- CT scan (see Figure 6)

❑ management
- multiple aspiration of abscess and/or excision, and send for C&S
- excision is preferable if location suitable
- antibiotics
  - empirically: vancomycin plus ceftazidime plus metronidazole or chloramphenicol or rifampin
  - after sensitivity results return, revise antibiotics
- anti-convulsants x 1-2 years

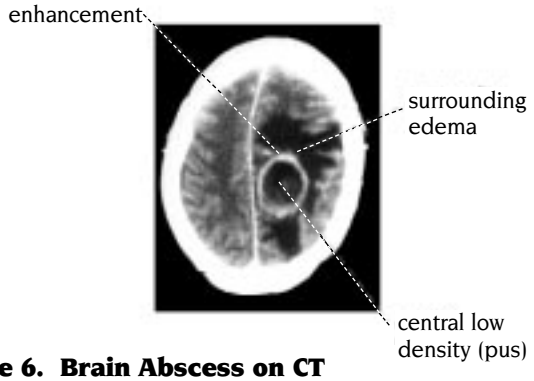

Figure 6. Brain Abscess on CT

Other Causes of Pus

❑ subdural empyema (from sinusitis, mastoiditis - rare, 20% mortality)
❑ meningitis, encephalitis, AIDS, toxoplasmosis (see Neurology Chapter)
❑ osteomyelitis of skull (Pott's puffy tumour), usually seen with sinusitis
❑ granuloma (TB, sarcoid)

BLOOD

Hematoma/hemorrhage

❑ epidural, subdural hematoma (see Trauma section)
❑ intracerebral, intraventricular hemorrhage, SAH (see Cerebrovascular Disease section)

Vascular Abnormality

❑ aneurysm, AVM (see Cerebrovascular Disease section)

- ❑ ischemic cerebral infarction (80%)
  - embolic (heart, carotid artery, aorta) or thrombosis of intracerebral arteries (see Neurology Chapter)
- ❑ intracranial hemorrhage (20%)
  - subarachnoid hemorrhage (SAH), spontaneous intracerebral hemorrhage (ICH), intraventricular hemorrhage (IVH)

SUBARACHNOID HEMORRHAGE (SAH) (see Colour Atlas NS3)

Etiology
- ❑ trauma (most common)
- ❑ spontaneous
  - aneurysms (75-80%)
  - idiopathic (14-22%)
  - AVMs (5%)

Risk Factors
- ❑ pregnancy/parturition in patients with pre-existing AVMs
- ❑ hypertension
- ❑ oral contraceptives (OCP)
- ❑ substance abuse (cigarette smoking; cocaine; alcohol (debatable))
- ❑ diurnal variation in BP
- ❑ slight increased risk with advancing age, and with LP/cerebral angiogram in patients with cerebral aneurysm

Clinical Features
- ❑ sudden onset severe headache: "worst headache of my life"
- ❑ sentinel/warning leaks
  - small SAH with sudden severe H/A +/– transient focal neurological deficit
  - blood on CT or LP
  - 30-60% of patients with full blown SAH give history suggestive of a warning leak
    - diagnosis must be made or excluded the first time it is suspected
- ❑ vomiting, nausea (increased ICP)
- ❑ meningismus (neck stiffness, positive Kernig's and Brudzinski's sign)
- ❑ photophobia
- ❑ decreased level of consciousness
- ❑ focal deficits: cranial nerve palsy (e.g. III, IV), hemiparesis
- ❑ ocular hemorrhage in 20-40% (due to sudden increase in ICP)
- ❑ occasionally exertional (straining, intercourse)

Clinical Course/Natural History
- ❑ 10-15% die before reaching hospital
- ❑ overall mortality 50-60% in first 30 days
- ❑ major cause of mortality is rebleeding
  - risk of rebleeding: 4% on first day, 15-20% within 2 weeks, 50% by 6 months
  - if no rebleed by 6 months chance of rebleeding decreases to same incidence of unruptured aneurysm (2%)

Diagnosis (see Figure 7)
- ❑ differential diagnosis: migraine, tension H/A, meningitis, stroke, flu
- ❑ CT without contrast (see Figure 8)
  - 90% sensitivity, 100% specificity
  - may be negative if small bleed or presentation delayed several days
  - positive history for SAH with negative CT - **MUST do LP**
  - hydrocephalus, IVH, ICH, infarct or large aneurysm may be present
  - CT may also suggest site of aneurysm that has bled
- ❑ lumbar puncture (LP)
  - contraindications
    - known or suspected intracranial mass
    - non-communicating (obstructive) hydrocephalus
    - decreased LOC, focal neurological deficit (hemiparesis), papilledema
    - coagulopathy (platelets < 50, anticoagulants, etc.)
      - correctable if no alternative to LP
      - change site - e.g. cisternal or C_1-C_2
    - infection at site desired for LP (e.g. epidural abscess)
  - CSF colour: bloody initially —> xanthochromic supernatant (yellow) by 12-48 hours
  - high sensitivity
  - traumatic tap (false positive): if bloody MUST centrifuge and observe the supernatant, clear supernatant means traumatic tap and xanthochromia means SAH
- ❑ cerebral angiography
  - demonstrates source of SAH in 80-85% of cases

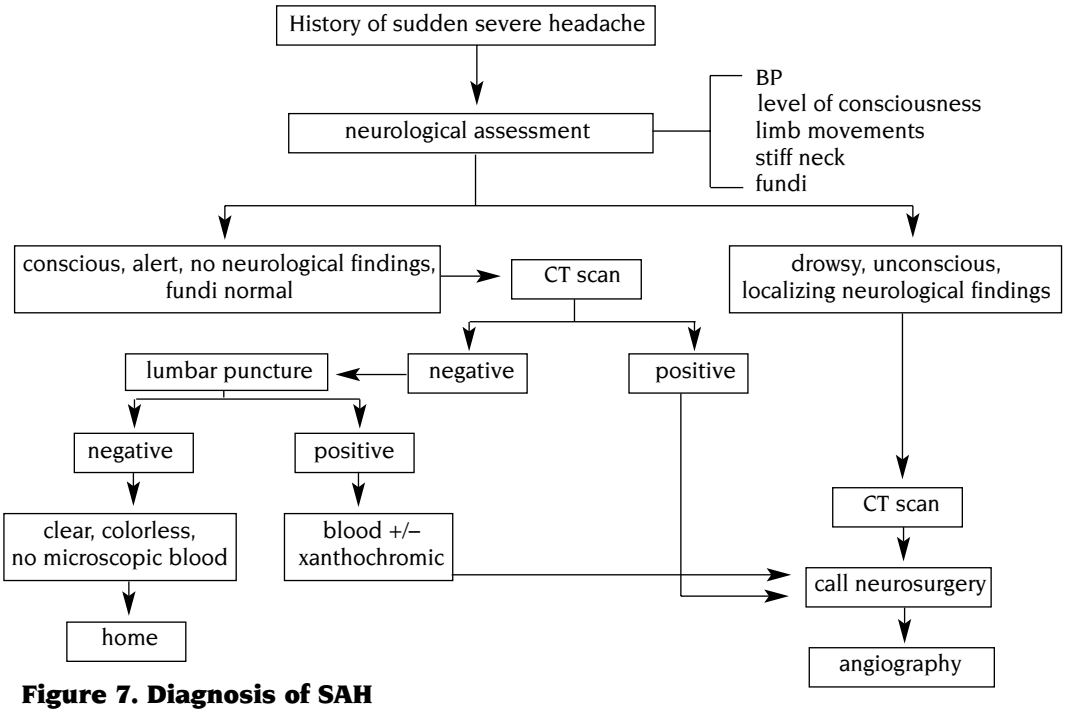

Figure 7. Diagnosis of SAH

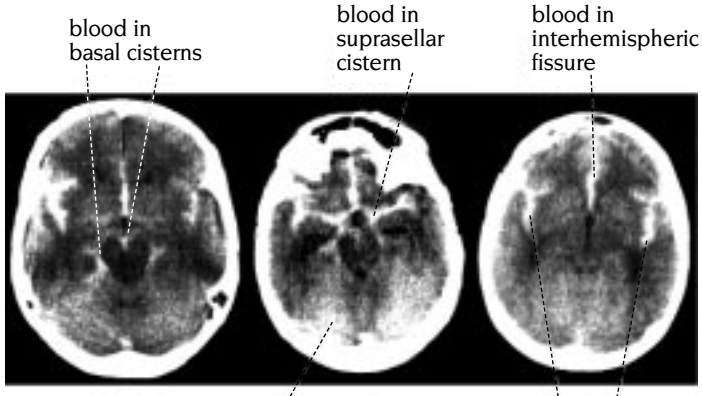

Figure 8. SAH on CT

Complications

- ❑ vasospasm
  - constriction of blood vessels in response to high pressure arterial blood outside vessels in the subarachnoid space (starting day 4 after SAH)
  - confusion, decreased LOC, focal neurodeficit (speech or motor)
  - detect clinically and/or with angiogram (decreased vessel caliber) or transcranial doppler (increased blood velocity)
  - radiographic evidence seen in 30-70% of arteriograms performed 7 days following SAH (peak incidence)
  - symptomatic only in 20-30% of patients with SAH
  - onset: 4-14 days post SAH (if patient deteriorates within first 3 days, MUST look for another cause)
  - can produce permanent infarcts and death
  - a major cause of morbidity and mortality
- ❑ hydrocephalus (15-20%)
  - can be acute or chronic - requiring shunt or drain
- ❑ neurogenic pulmonary edema
- ❑ hyponatremia (SIADH, cerebral salt wasting)
- ❑ diabetes insipidus
- ❑ cardiac - arrhythmia, MI, CHF

Management
- ❑ bed rest, elevate head (30 degrees), minimal external stimulation
- ❑ control HTN, avoid hypotension since CBF autoregulation impaired by SAH
- ❑ prophylactic anticonvulsant: short course of Dilantin (one week)
- ❑ neuroprotective agent: nimodipine (for vasospasm)
- ❑ early surgery to prevent rebleed
- ❑ intraventricular catheter if acute hyrdocephalus present
- ❑ "**Triple H**" therapy for vasospasm: **h**ypertension, **h**ypervolemia, **h**emodilution
- ❑ angioplasty for refractory vasospasm

SPONTANEOUS INTRACEREBRAL HEMORRHAGE (ICH)

Definition
- ❑ bleeding into brain parenchyma without accompanying trauma
- ❑ can dissect into ventricular system (IVH) or through cortical surface (SAH)

Etiology
- ❑ hypertension
- ❑ vascular anomalies
    - aneurysm
    - AVM's and other vascular malformations
    - cerebral amyloid angiopathy
    - vasculitis
- ❑ coagulopathies
- ❑ tumours (1%) – often malignant (eg. glioblastoma multiforme (GBM), lymphoma, metastases)
- ❑ hemorrhagic transformation of previously ischemic area (infarct)
- ❑ drugs (amphetamines, cocaine, etc.)
- ❑ anticoagulants (coumadin, tPA, streptokinase)
- ❑ idiopathic

Clinical Features
- ❑ 30 day mortality rate is 44%, mostly due to cerebral herniation
- ❑ gradual onset of symptoms over minutes to hours (unlike ischemic stroke)
- ❑ H/A, vomiting, decreased LOC are common
- ❑ specific symptoms depend on location of ICH
    - putamen
        - contralateral hemiparesis progressing to hemiplegia, coma or death
    - thalamus
        - contralateral hemisensory loss
        - contralateral hemiparesis with internal capsule involvement
    - cerebellum
        - sudden severe vertigo and vomiting
        - ataxia, nystagmus, dysmetria, incoordination
        - preserved consciousness until late then sudden death, "talk 'til death"
        - mass effect (tonsillar herniation) —> surgical emergency
        - headache (occipital)
    - pons
        - quadriplegia
        - sudden decreased LOC
        - "pinpoint pontine pupils", disconjugate extraocular movements
        - respiratory abnormalities
        - hyperthermia
        - rapid death
    - lobar
        - frontal lobe: frontal H/A with contralateral hemiparesis
        - parietal lobe: contralateral hemisensory loss and mild hemiparesis
        - occipital lobe: ipsilateral eye pain and contralateral homonymous hemianopsia
        - temporal lobe: on dominant side, fluent dysplasia with receptive aphasia

Diagnosis
- ❑ high density blood on CT without contrast
- ❑ MRI does not show blood immediately - not procedure of choice

Management
- ❑ medical
    - correct HTN, coagulopathy
    - control ICP (mannitol, hyperventilate, elevate head of bed)
    - anticonvulsants

❑ surgical
- craniotomy with evacuation of clot under direct vision, resection of source of ICH (i.e. AVM, tumour, cavernoma), ventriculostomy to treat hydrocephalus
- indications
  - symptomatic
  - marked mass effect, raised ICP - evacuate clot, decompress
  - rapid deterioration (especially with signs of brainstem compression)
  - favorable location, e.g. cerebellar
  - young patient (< 50)
  - if tumour, AVM, aneurysm, or cavernoma suspected (resection or clip to decrease risk of rebleed)
- contraindications
  - small bleed: minimal symptoms, high GCS (not necessary)
  - massive hemorrhage (especially dominant lobe), low GCS/coma, brainstem lost (poor prognosis)
  - medical reasons, e.g. very elderly, severe coagulopathy, difficult location, e.g. basal ganglia, thalamus (poor surgical candidate)

INTRACRANIAL ANEURYSMS (see Colour Atlas NS12, NS13 and NS14)

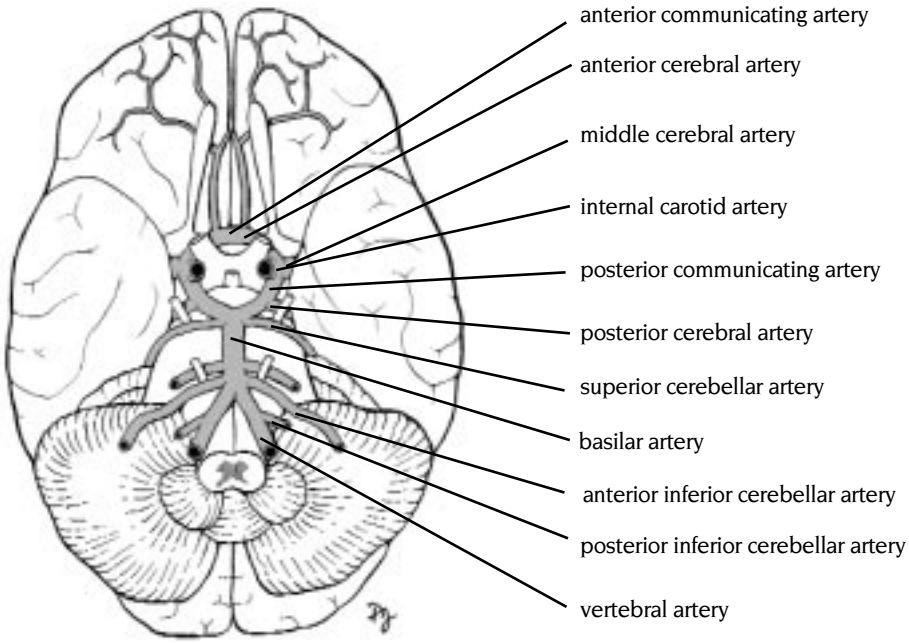

Figure 9. Circle of Willis

Illustration by Andree Jenks

Epidemiology
❑ prevalence of about 5%
❑ female > male
❑ 20% multiple aneurysms
❑ age 35-65 years

Types
❑ saccular (berry)
- most common type of aneurysm
- located at branch points of major cerebral arteries (Circle of Willis)
- 85-95% located in the carotid system:anterior communicating artery/anterior cerebral artery (30%), posterior communicating artery (25%), middle cerebral artery (20%)
- 5-15% in posterior circulation (vertebrobasilar)

❑ fusiform
- atherosclerotic
- more common in vertebrobasilar system
- rarely rupture

❑ mycotic
- secondary to any infection of vessel wall
- most commonly *Streptococcus* and *Staphylococcus* (associated with spontaneous bacterial endocarditis (SBE))

Clinical Presentation
- ❑ rupture (SAH, ICH, IVH, subdural blood) – 90%
- ❑ mass effect (giant aneurysms)
  - internal carotid or anterior communicating aneurysm may compress
    1) the pituitary stalk or hypothalamus causing hypopituitarism.
    2) the optic nerve or chiasma producing a visual field defect
  - basilar artery aneurysm may compress the midbrain, pons (limb weakness), or CN III (impaired eye movements)
  - posterior communicating artery aneurysm may produce a CNIII palsy
  - intracavernous aneurysms may compress CN's III, IV, VI, and V1 producing ophthalmoplegia and facial pain
- ❑ small infarcts due to distal embolization
- ❑ seizures
- ❑ headache without hemorrhage
- ❑ incidental CT or angiography finding (asymptomatic)

Management
- ❑ imaging: CT, magnetic resonance angiography (MRA), angiogram
- ❑ ruptured aneurysms
  - initial management of SAH/ICH
  - overall trend towards better outcome with early surgery (48-96 hours after SAH)
  - surgical clipping is the optimal treatment
  - other treatment options: trapping(clipping of proximal and distal vessels), thrombosing (endovascular technique), wrapping, proximal ligation
- ❑ unruptured aneurysms
  - 1-3% annual risk of rupture: risk dependent on size of aneurysm
  - no clear evidence on when to operate: need to weigh life expectancy, risk of hemorrhage and mortality/morbidity of SAH vs. that of aneurysm surgery (age, medical risk, etc.)
  - treat unruptured aneurysms >10 mm; consider treating when aneurysm 7-9 mm in middle-aged or younger patients
  - follow smaller aneurysms with serial angiography

VASCULAR MALFORMATIONS OF THE NERVOUS SYSTEM
- ❑ types
  - arteriovenous malformations (AVMs)
  - cavernous malformations (cavernoma, cavernous hemangioma, angiographically occult vascular malformation)
  - venous angioma
  - capillary telangiectasias
  - arterio-venous fistula
- ❑ clinical significance
  - principally AVMs and cavernous malformations produce intracranial hemorrhages and seizures

Arteriovenous Malformations (AVMs) (see Colour Atlas NS9, NS10 and NS11)
- ❑ description
  - tangle of abnormal vessels, arteriovenous shunts, with no intervening capillary beds or brain parenchyma
  - congenital, tends to enlarge with age
  - male:female = 2:1
  - present in younger age group than aneurysms (average age at diagnosis ~ 33 years)
- ❑ presentation
  - ICH (40-60%)
    - risk of major bleed: 4% per year
    - 10% mortality (versus 50-60% for aneurysmal SAH) per bleed
    - 30-50% morbidity (serious neurological deficit) per bleed
  - seizures (50%)
  - mass effect (e.g. Tic Douloureux 2º to CPA AVM)
  - focal neurological signs secondary to ischemia (high flow —> "steal phenomena")
  - localized headache (infrequent)
  - bruit (especially with dural AVMs)
  - increased ICP
  - may be silent
- ❑ diagnosis
  - MRI (flow void), MRA
  - angiography

- ❑ management
  - decreases risk of future hemorrhage and seizure
    - surgical excision
    - endovascular embolisation (glue, balloon)
    - stereotactic radiotherapy (for small AVMs; i.e. ≤ 3 cm in diameter)
  - conservative (seizure control if necessary)

Cavernous Malformations

- ❑ benign vascular hamartoma consisting of irregular thick and thin walled sinusoidal vascular channels located within the brain
- ❑ symptoms: H/A, seizure, neurological deficit, ICH
- ❑ prevalence: 0.3-0.5%
- ❑ hemorrhage risk may be up to 3.6% per year
- ❑ diagnosis: MRI usually not seen with angiography
- ❑ treatment: surgical excision - depending on presentation and location (most are observed)

SPINE

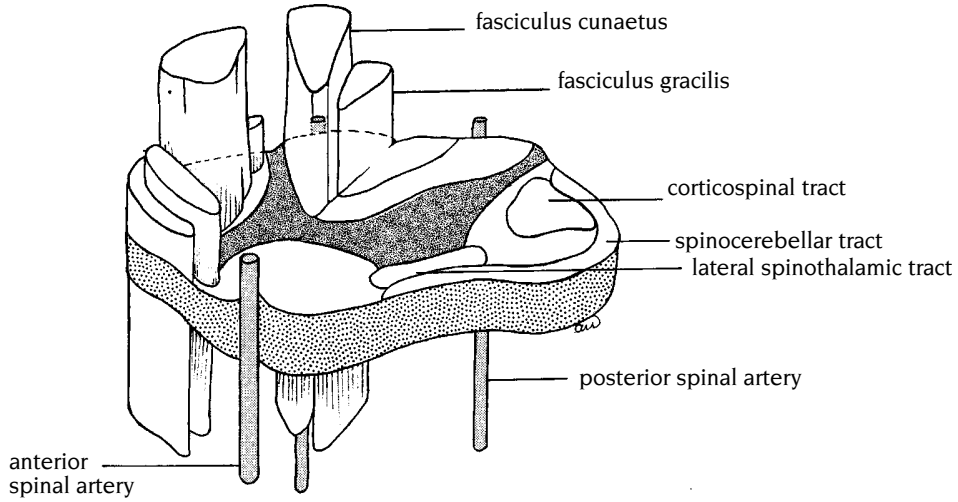

Figure 10. Anatomy of the Spinal Cord

Illustration by Aimée Warrell

CORD AND ROOT COMPRESSION

Etiology

- ❑ congenital
  - Chiari malformation
- ❑ acquired
  - trauma —> hematoma, vertebral fracture, subluxation
  - herniated disk (nucleus pulposus of disk herniates through ruptured annulus fibrosus)
  - spondylosis (degenerative process of the spine which may result in spinal stenosis)
  - spondylolisthesis (anterior subluxation of one vertebral body on another)
  - infectious (abscess)
  - vascular (AVM=rare)
  - neoplastic
    - extradural (lymphoma, metastases from prostate, lung, breast, kidney)
    - intradural
      - extramedullary (schwannoma, meningioma, neurofibroma)
      - intramedullary (ependymoma, astrocytoma, hemagioblastoma)

Clinical Features
- ❑ local pain at site of lesion
- ❑ radiculopathy
  - Motor: weakness, wasting, decreased deep tendon reflex in root distribution
  - Sensory: dermatomal decreased pinprick sensation, numbness, paresthesiae, pain
  - Trophic changes: eg. dry skin (if long-standing radiculopathy)
- ❑ myelopathy
  - LMN signs/symptoms at level of lesion
  - UMN signs/symptoms below lesion
    - motor: proximal weakness and spasticity of lower extremities, increased reflexes, clonus, Babinski sign (extenser plantar response), sphincter disturbance
    - sensory: findings may be minimal (reduced vibration, proprioception), +/– Lhermitte sign

	LMN (Lower Motor Neuron)	UMN (Upper Motor Neuron)
Tone	Flaccid	Spastic
Reflexes	Decreased	Increased
Plantar Response	Extensor	Extensor
Muscles	Atrophy, fasciculations	Atrophy arms flexed, legs extended

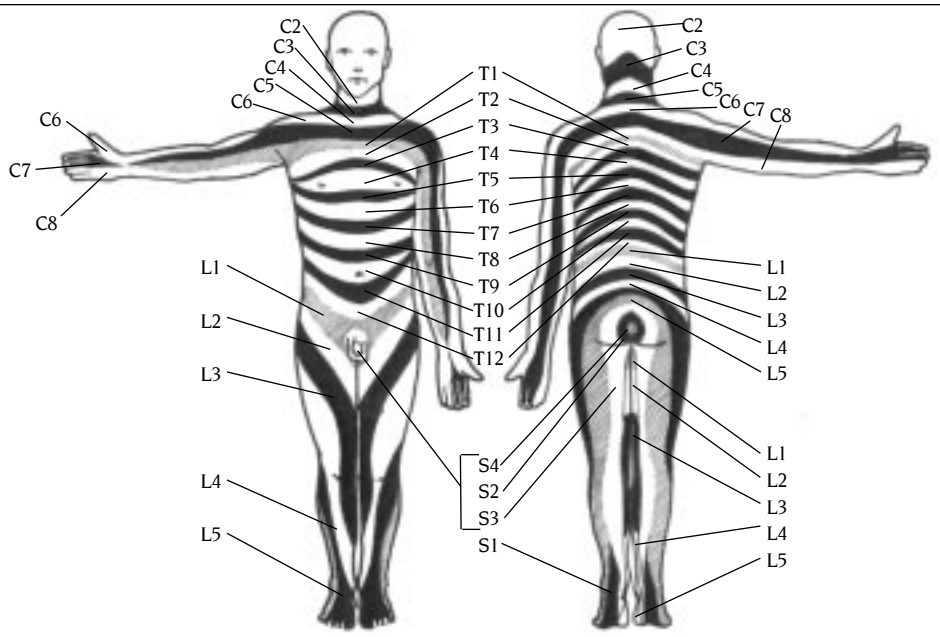

Figure 11. Dermatomes

Illustration by Glen Oomen

Investigations
- ❑ plain x-ray of spine
- ❑ CT, MRI
- ❑ myelogram
- ❑ electromyography (EMG), electrophysiology

Management
- ❑ for disc herniation see Lumbar Disk Syndrome
- ❑ unstable fractures may require surgical intervention
- ❑ neoplasms are treated with a combination of surgery and radiation therapy

SPINAL CORD SYNDROMES (see Neurology Chapter)

- ❑ complete spinal cord lesion
  - no preservation of motor/sensory function at > 3 segments below lesion/injury
- ❑ incomplete spinal cord lesion
  - any residual function at > 3 segments below lesion
  - signs include sensory/motor function in lower limbs and "sacral sparing" (perianal sensation, voluntary rectal sphincter contraction)
  - syndromes include Brown-Sequard's, central cord, anterior cord and posterior cord syndrome

Brown-Sequard's Syndrome (Hemisection of cord)

- ❑ causes include
  - penetrating trauma
  - extrinsic compression
- ❑ clinical features
  - ipsilateral weakness (UMN lesion) below lesion
  - contralateral pain and temperature sensory deficits (deficits are 1 to 2 levels below injury)
  - ipsilateral reduction in proprioception and vibration sense below lesion
  - light touch preserved
- ❑ best prognosis of cord injuries (90% independently ambulate and have good sphincter control)

Central Cord Syndrome

- ❑ most common incomplete spinal cord injury syndrome
- ❑ cause: spinal extension injury, particularly with pre-existing cervical spondylosis
- ❑ clinical features
  - weakness upper (LMN lesion) > lower (UMN lesion) extremities; more pronounced in the hands
  - dissociated sensory loss
    - "vest" or bilateral suspended pain and temperature deficit with sacral sparing
    - spared touch, joint position and vibration sensation
  - sphincter dysfunction (usually urinary retention)
- ❑ 50% recover enough LE function to ambulate
- ❑ hand recovery variable

Anterior Cord Syndrome

- ❑ causes
  - anterior cord compression or anterior spinal artery occlusion
- ❑ clinical features
  - dissociated sensory loss
    - bilateral pain and temperature deficit below lesion
    - spared touch, joint position and vibration sensation
  - bilateral paraplegia (UMN below and LMN at level of the lesion)
  - sphincter dysfunction (urinary retention)
- ❑ worst prognosis, only 10-20% recover functional motor control

Posterior Cord Syndrome (~rare)

- ❑ causes
  - trauma
  - posterior spinal artery infarct
- ❑ clinical features
  - joint position and vibration sensation loss
  - pain and paresthesias in neck, upper arms, torso
  - mild paresis of upper extremities

SYRINGOMYELIA (see Colour Atlas NS22)

- ❑ "syrinx", cavitation of spinal cord substance

Etiology

- ❑ idiopathic
- ❑ post-traumatic
- ❑ associated with
  - craniovertebral anomalies (congenital) e.g. Chiari malformation or myelomeningocele
  - intramedullary tumours
  - arachnoiditis (traumatic)

Presentation

- ❑ suspended, dissociated sensory loss
  - pain and temperature loss in a cape-like distribution at level of cervical syrinx
  - preserved light touch and other modalities
- ❑ LMN arm/hand weakness or wasting
- ❑ may have spastic weakness of legs
- ❑ may have hydrocephalus, often asymptomatic
- ❑ painless arthropathies (Charcot's joints)

Investigations
❑ MRI is best method
❑ myelogram with delayed CT

Management
❑ conservative if NOT progressing
❑ shunt (syringosubarachnoid, synringoperitoneal or syringopleural)
❑ if associated with Chiari malformation
  • first decompress posterior fossa, if not successful then shunt
❑ progressive deterioration in >1/3 despite therapy

CERVICAL DISC SYNDROME

Etiology
❑ most common levels
  • C5-C6 (C6 root) more common vs. C6-C7 (C7 root)
❑ less common, but important with respect to activities of daily living
  • C4-5 (C5 root), C7-T1 (C8 root)

Table 3. Lateral Cervical Disc Syndrome

	C4-5	C5-6	C6-7	C7-T1
Root Involved	C5	C6	C7	C8
Motor	Deltoid Supraspinatus Biceps	Biceps	Triceps	Digital flexors Intrinsics
Reflex	No change	Biceps	Triceps	Finger jerk
Sensory	Shoulder	Thumb	Middle finger	Ring finger, little finger

Clinical Features
❑ lateral disc protrusion compresses nerve root
  • pain down arm in nerve root distribution, worse with neck extension
  • referred parascapular pain
  • +/– nerve conduction velocity abnormalities
❑ central cervical disc protrusion compresses spinal cord as well as nerve roots

Differential Diagnosis
❑ shoulder lesion
❑ thoracic outlet syndrome (including Pancoast tumour)
❑ cervical spine tumour
❑ peripheral nerve lesion (e.g. carpal tunnel)
❑ acute brachial neuritis
❑ myocardial infarction (MI) (left C6 radiculopathy)

Investigations
❑ C-spine x-ray
❑ CT, MRI (procedure of choice)
❑ EMG, nerve conduction studies

Management
❑ conservative (recovery in 95%)
  • NSAIDs, collar, traction may help
  • most patients get better spontaneously in 4 to 8 weeks
❑ surgical indications
  • intractable pain despite adequate conservative treatment for > 3 months
  • progressive neurological deficit
  • anterior cervical discectomy is usual surgical choice

LUMBAR DISC SYNDROME (see Colour Atlas NS20)

Etiology
❑ protrusion/herniation of nucleus pulposus
- laterally: compressing nerve root
- centrally: compressing cauda equina

❑ common: (>95% of herniated lumbar disks): L4-5 (L5 root), L5-S1 (S1 root)
❑ uncommon: L3-4 (L4 root)

Table 4. Lateral Lumbar Disk Syndromes

	L3-4	L4-5	L5-S1
root Involved	L4	L5	S1
Pain	Femoral pattern	Sciatic pattern	Sciatic pattern
Motor	Quads (knee extension)	Tibialis anterior(dorsiflexion), Extensor hallusis longus (hallux extension)	Gastrocnemius, soleus (plantar flexion)
Reflex	Knee jerk	Medial Hamstrings	Ankle jerk
Sensory	Medial leg	Dorsal foot to hallux, lateral leg	Lateral foot
Relative Incidence	< 10%	45%	45%

Clinical Features
❑ leg pain > back pain
❑ limited back movement (especially forward flexion)
❑ symptoms and signs of radiculopathy
- pain in root distribution (worse with movement, valsalva)
- dermatomal sensory deficit
- LMN weakness
- reduced deep tendon reflex
- +/– reflex paravertebral muscle spasm (functional scoliosis, loss of lordosis)

❑ nerve root tension signs:
- straight leg raise (SLR: Lasegue's test), crossed SLR —> L5, S1 roots
- femoral stretch —> L4 root

Differential Diagnosis
❑ spinal: stenosis, tumour, spondylolisthesis **(see Colour Atlas NS21)**
❑ leg: spinal stenosis, arthritic hip, sciatic nerve lesion (e.g. tumour)
❑ pelvic bones: tumour
❑ functional /nonorganic

Investigations
❑ X-ray spine (only to rule out other lesions)
❑ CT
❑ MRI
❑ myelogram and post-myelogram CT (if surgery contemplated and plain CT not conclusive)

Management
❑ conservative
- no bedrest unless severe radicular symptoms
- activity modification (reduce sitting, lifting)
- physiotherapy (PT), exercise programs
- analgesics (acetominophen, NSAIDs)
- patient education
- 95% improve spontaneously within 4 to 8 weeks

❑ surgical indications
- intractable leg pain despite adequate conservative treatment for > 3 months
- disabling neurological deficit
- progressive neurological deficit
- cauda equina syndrome

CAUDA EQUINA SYNDROME

Etiology
- ❑ secondary to compression of lumbosacral nerve roots below conus medullaris
- ❑ extrinsic tumour, carcinomatous meningitis, arachnoiditis, spinal stenosis, central posterior lumbar disc herniation

Clinical Features
- ❑ motor
  - weakness/paraparesis in multiple root distribution
  - reduced reflexes (usually achilles reflex)
  - sphincter disturbace (urinary retention which can lead to overflow incontinence, and fecal incontinence due to loss of anal sphincter tone)
- ❑ sensory
  - multiple dermatome, bilateral sensory loss or pain
  - saddle anesthesia (most common sensory deficit)
- ❑ other
  - sexual dysfunction (late finding)

Management
- ❑ requires urgent investigation and decompression to preserve bowel and bladder function

LUMBAR SPINAL STENOSIS
- ❑ etiology
  - congenital narrowing of spinal canal combined with degenerative changes (herniated disk, hypertrophied facet joints and ligaments)
- ❑ clinical presentation
  - neurogenic claudication ("pseudoclaudication")
  - neurologic exam may be normal, including straight leg raise test
  - symptoms relieved only by changing position (leaning forward, sitting down)
- ❑ investigations
  - MRI, CT
- ❑ treatment
  - conservative (NSAIDS, PT)
  - surgical (laminectomy with root decompression)

See RED FLAGS FOR BACKPAIN (see Family Medicine Chapter)
- ❑ the presence of any or several of these red flags warrants thorough investigations to determine the cause of the back pain as it may require immediate intervention (eg. cauda equina syndrome)
- ❑ conditions to keep in mind
  - neoplasm
  - infection
  - fracture
  - Cauda Equina Syndrome

TRAUMA (BRAIN AND SPINAL CORD)

HEAD INJURY

Scalp Injury
- ❑ rich blood supply
- ❑ considerable blood loss (vessels contract poorly when ruptured)
- ❑ minimal risk of infection due to rich vascularity

Skull Fractures
- ❑ depressed fractures —> double density on skull x-ray
  - simple fractures
  - compound fractures —> increased risk of infection
- ❑ internal fractures into sinus —> meningitis, pneumocranium
- ❑ basal skull fractures —> not readily seen on x-ray, rely on clinical signs
  - retroauricular ecchymoses (Battle's sign)
  - periorbital ecchymoses (raccoon eyes)
  - hemotympanum
  - CSF rhinorrhea, otorrhea
  - suspect with Lefort II or III midface fracture

Cranial Nerve Injury
- ❑ most commonly olfactory

Arterial Injury
- ❑ e.g. carotid-cavernous (C-C) fistula, carotid/vertebral artery dissection

INTRACRANIAL BLEEDING

Extradural ("Epidural") Hematoma (see Figure 12) **(see Colour Atlas NS2)**
- ❑ young adult, male > female
- ❑ temporal-parietal skull fracture —> ruptured middle meningeal artery
- ❑ symptoms: lateral transtentorial herniation, classically there is lucid interval between concussion and coma
- ❑ prognosis: good with optimal prompt management, since the brain is often not damaged
- ❑ CT without contrast: high density biconvex mass against skull, usually with uniform density and sharp margins "lens-shaped"
- ❑ management: head elevation, mannitol pre-operatively, evacuation with small craniotomy

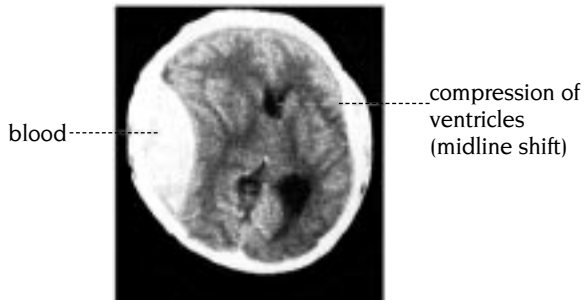

Figure 12. Extradural Hematoma on CT

Subdural Hematoma (see Figure 13) **(see Colour Atlas NS4)**
- ❑ acute
  - arise from rupture of a vessels that bridge the surface of the cerebral hemisphere and the skull (e.g. cortical artery, large vein, or venous sinus) due to violent trauma
  - prognosis: poor overall, since the brain is often injured
  - CT: high density concave mass usually less uniform, less dense and more diffuse than extradural hematoma
  - management: craniotomy

❑ chronic **(see Colour Atlas NS5 and NS6)**
- often due to minor injuries or no history of injury
- several weeks after injury
- the clot from the original bleed liquifies and becomes bounded by a thick, friable, vascularized outer membrane and a thin, lucent inner membrane.
- symptoms of raised ICP and sometimes seizures, progressive dementia, gait problem, obtundation out of proportion to focal neurological deficit, "the great imitator" (of dementia, tumours, etc.), normal pressure hyrocephalus (NPH) (see Neurology Chapter)
- risk factors: older, alcoholic, patients with CSF shunts, anticoagulants
- expands due to repeated bleeding
- prognosis: brain usually undamaged, however, recurrent bleeding from the outer membrane leads to expansion of the hematoma, increased ICP, shift of the cerebral hemispheres, transtentorial herniation, and death
- CT: low density (liquefied clot) concave mass
- management: burr hole drainage, craniotomy if recurrent

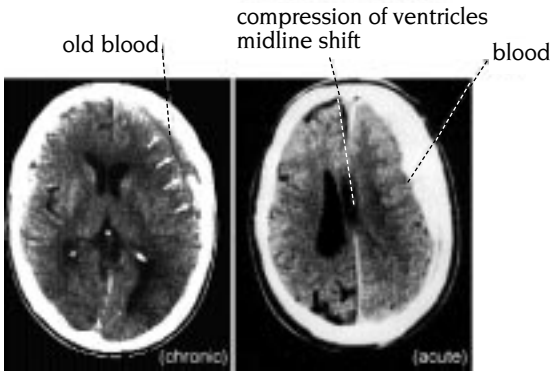

Figure 13. Subdural Hematoma on CT

Traumatic Intracerebral Hemorrhage
❑ any size, any part of brain, may be multiple
❑ immediate or delayed
❑ frontal and temporal lobes most commonly injured (by coup/contre-coup mechanism)

BRAIN INJURY

Primary Impact Injury
❑ mechanism of injury determines pathology: i.e. with penetrating injuries, gun shot wounds
- low velocity —> local damage
- high velocity —> distant damage possible (due to wave of compression)

❑ concussion
- American Academy of Neurology (AAN) definition: "a trauma-induced alteration in mental status that may or may not involve loss of consciousness"
- AAN Classification:
  Grade 1: altered mental status <15 min
  Grade 2: altered mental status >15 min
  Grade 3: any loss of consciousness
- no parenchymal abnormalities on CT

❑ coup (damage at site of blow)
❑ contre-coup (damage at opposite site of blow)
- acute decompression causes cavitation
- followed by a wave of acute compression

❑ contusion (hemorrhagic) **(see Colour Atlas NS7)**
- high density areas on CT with little mass effect
- commonly occurs with brain impact on bony prominences (falx, sphenoid wing, floor of frontal and temporal fossae)

❑ diffuse axonal injury (diffuse axonal shearing)
- may tear blood vessels->hemorrhagic foci (may not be proportionate to axonal injury)
- wide variety of damage results
- all brain injury causes shear
- often the cause of decreased LOC if no space occupying lesion on CT

Secondary Pathologic Processes

- ❑ 1/3 who die in hospital after head injury were able to talk after the injury
- ❑ delayed and progressive injury to the brain due to:
  - edema
  - intracranial hemorrhages
  - ischemia/infarction
  - raised ICP (which leads to (decreased cerebral perfusion pressure (CPP) and herniation)

Extracranial Conditions

- ❑ hypoxemia
  - trauma: chest, upper airway, brainstem
  - exceptionally damaging to traumatized brain cells
  - leads to ischemia, raised ICP
- ❑ hypercarbia
  - leads to raised ICP (secondary to vasodilation)
- ❑ systemic hypotension
  - caused by blood loss, not by head injury (e.g. ruptured spleen)
  - cerebral autoregulation lost in trauma
  - leads to decreased CPP, ischemia
- ❑ hyperpyrexia
  - leads to increased brain metabolic demands
- ❑ fluid and electrolyte imbalance
  - iatrogenic (most common)
  - syndrome of inappropriate antidiuretic hormone (SIADH) secretion (from head injury)
  - diabetes insipidus (DI) (from head injury)
  - may lead to cerebral edema and raised ICP
- ❑ fat embolism
  - multiple trauma
  - long bone fractures
  - petechiae and edema
  - hypoxia a key feature due to pulmonary effects
  - decreased LOC, seizures
- ❑ coagulopathy
- ❑ post-traumatic carotid artery dissection

Intracranial Conditions

- ❑ raised ICP due to
  - traumatic cerebral edema OR traumatic intracranial hemorrhage
- ❑ raised ICP results in
  - decreased cerebral perfusion (CPP = MAP – ICP)
  - +/– herniation

LATE COMPLICATIONS OF HEAD INJURY

Seizures

- ❑ 5% of head injury patients develop seizures
- ❑ incidence related to severity and location of injury —> increased with local brain damage or intracranial hemorrhage
- ❑ post-traumatic seizure may be immediate, early, or late
- ❑ presence of early (within first week) post traumatic seizure, incidence of later seizures rises to 25%

Meningitis

- ❑ associated with CSF leak from nose or ear

Hydrocephalus

- ❑ acute hydrocephalus or delayed normal pressure hydrocephalus (NPH)

SPINE INJURY

Vertebral Column (bone, discs, ligaments)

- ❑ stable fracture
  - compression fracture
- ❑ unstable fracture
  - burst fracture (note: not all burst fractures are unstable)
  - dislocation
- ❑ "special" fractures
  - Odontoid (Type I, II, III): Type II is unstable and most require fixation
  - Jefferson (fractures in ring of C1): due to axial loading (C1)
  - Hangman's (fractured C2 pedicles at pars interarticularis): due to hyper-extension (C2)

Spinal Cord
❑ cord injury with initial bony or ligamentous trauma or after moving an unstable vertebral column
❑ complete
- no preservation of sensory/motor function below lesion
- no recovery

❑ incomplete lesions (see Spinal Cord Syndromes section)

Nerve Roots
❑ avulsion, e.g. brachial plexus in motorcycle accident

TRAUMA MANAGEMENT (see Emergency Medicine Chapter)

Aims
❑ recovery from primary injury
❑ prevent further damage from secondary pathologic processes

Initial Management
❑ ABC's of trauma management take priority

A - airway
- ensure that there is no airway obstruction caused by local trauma to the larynx or trachea
- C-spine immobilization to prevent further spinal cord injury (immobilize with collar, sandbags, fracture board, skull tong or halo traction)
- if intubation is indicated emergently and cervical fracture cannot be ruled out, orotracheal intubation with in-line neck stabilization is used

B - breathing
- ensure adequate oxygen supply by monitoring with a pulse oximeter
- respiration is often depressed after brain injury when consciousness is depressed
- ventilate with oxygen (100%) if necessary

C - circulation
- differentiate hypotensive shock (BP low, HR high) from neurogenic shock (BP low, HR low)
- ensure adequate perfusion of spinal cord and manage neurogenic shock (dopamine, IV fluids, MAST)

❑ suspect spinal cord injury with weakness, numbness, spine pain, head injury, high energy injury or multisystem injuries
❑ rule out spinal fracture (cervical, thoracic, lumbar)
❑ 5-10% of patients with spinal injuries have injuries at other levels

Neurological Assessment
❑ mini history
- period of LOC
- post traumatic amnesia
- loss of sensation/function

❑ neurological exam
- head and neck (lacerations, bruises, basal skull fracture signs, facial fractures, foreign bodies)
- spine (palpable deformity, midline pain/tenderness)
- Glasgow Coma Scale (GCS) (see Emergency Medicine and Neurology Chapters)
- eyes (pupillary size and reactivity)
- brainstem (breathing pattern, CN palsies)
- motor exam, sensory exam (only if GCS is 15), reflexes
- cranial nerve exam
- sphincter tone
- record and repeat neurological exam at regular intervals

Initial Investigations
❑ CT head and upper C-spine (whole C-spine if patient unconscious)
❑ ABG, CBC, drug screen (especially alcohol)
❑ C,T,L-spine x-rays
- AP, lateral, odontoid views for C-spine
- must see C1 to C7 and C7-T1 interspace (swimmer's view if necessary) or CT
- ABCS - Alignment, Bone, Cartilage, Soft tissues (see Orthopedics Chapter)

❑ chest and abdominal x-ray as indicated

Late Management
❑ treatment for minor head injury
- observation over 24-48 hours
  - wake every hour
  - judicious use of sedatives or pain killers during this monitoring period

❑ treatment for severe head injury
- clear airway and ensure breathing (intubate if necessary)
- secure C-spine
- maintain adequate BP
- monitor to detect complications (Glasgow Coma Scale, CT, ICP)
- manage increased ICP if present
  - elevation of head
  - hyperventilation (target PCO_2 32-35 torr)
  - mannitol (temporary preoperative measure)
- remove hematoma if present

- ❑ treatment of spinal injury
  - reduce dislocation if present by traction or surgery
  - stabilize spine if unstable (halo vest, fusion, etc.)
  - further investigations (CT, tomogram, myelogram, MRI) to rule out cord compression
  - emergent surgical decompression and/or fusion if necessary; i.e. patient with neurological deterioration
  - more likely to be beneficial in incomplete cord injury
  - emergent surgery contraindicated for: complete spinal cord injury > 24 hours, medically unstable patient, and central cord syndrome
- ❑ which patients should be admitted to hospital?
  - skull fracture
  - indirect signs of basal skull fracture
  - confusion, impaired consciousness
  - focal neurological signs
  - extreme headache, vomiting
  - seizures
  - concussion with > 5 minutes amnesia
  - unstable spine
  - use of alcohol
  - social (i.e. no friend/relative to monitor for next 24 hours)
  - if there is any doubt, especially with children
- ❑ which patients need CT head or transfer to a neurosurgical center?
  - remains unconscious after resuscitation
  - focal neurological signs
  - deteriorating

KEY POINTS

- ❑ never do lumbar puncture in head injury
- ❑ all patients with head injury have C-spine injury until proven otherwise
- ❑ don't blame coma on alcohol - there may also be a hematoma
- ❑ low BP after head injury means injury elsewhere
- ❑ must clear spine both radiologically AND clinically

PERIPHERAL NERVES

INJURY

Classification and Clinical Course

- ❑ neuropraxia: nerve intact but fails to function, recovery within hours to days
- ❑ axonotomesis: axon disrupted but nerve sheath intact —> Wallerian degeneration (of axon segment distal to injury) —> recovery 1 mm/day
- ❑ neurotmesis: nerve completely severed, need surgical repair for recovery

Management

- ❑ electrophysiological studies (EMG, nerve conduction velocities (NCV)) may be helpful in assessing nerve integrity
- ❑ surgical repair unless nerve is known to be intact
- ❑ delay surgical repair for a few weeks (unless first 2 conditions met) to allow
  - clean wound
  - optimal surgical facilities
  - optimal cell metabolism
  - possible spontaneous recovery/regeneration
- ❑ microsurgery: suture nerve sheaths +/– nerve graft

ENTRAPMENT

General

- ❑ nerve compressed by nearby anatomic structures
- ❑ often secondary to localized, repetitive mechanical trauma with additional vascular injury to nerve
- ❑ consider systemic causes
  - rheumatoid arthritis
  - diabetes mellitus
  - hypothyroid
  - acromegaly
  - vasculitis
  - amyloidosis
  - pregnancy
- ❑ symptoms
  - pain distal (occasional proximal) to lesion
  - burning paresthesia/dysesthesia
  - sensory loss in nerve distribution
  - muscle weakness/wasting (advanced cases)

Carpal Tunnel Syndrome (CTS)

- ❑ etiology
  - most common entrapment neuropathy
  - median nerve entrapment at wrist, usually bilateral
  - female:male = 4:1
- ❑ presentation
  - classically: patient awakened at night with numb/painful hand, relieved by shaking/dangling/rubbing
  - distribution: radial 3.5 fingers
  - decreased light touch, 2 point discrimination, especially finger tips
  - job/hobby related repetitive trauma, especially forced wrist flexion
  - advanced cases: wasting/weakness of thenar muscles, especially abductor pollicis brevis
- ❑ diagnosis
  - history, physical
  - +/– Tinel's sign (tingling sensation on percussion of nerve)
  - +/– Phalen's sign (wrist flexion)
  - confirm with nerve conduction velocities (NCV), EMG pre-operatively
- ❑ management
  - conservative
    - neutral wrist splints (bedtime)
    - NSAIDs
  - surgical
    - release of flexor retinaculum
    - indications for surgery: refractory pain, +++ sensory loss, muscle atrophy

Ulnar Nerve Entrapment at Elbow

- ❑ second most common entrapment neuropathy
- ❑ may be entrapped at several locations
  - behind medial epicondyle
  - at medial intermuscular septum
  - distal to elbow at cubital tunnel
- ❑ presentation
  - sensory: pain, numbness in ulnar 1.5 fingers
  - wasting of interossei (especially first dorsal interosseous —> thumb web space)
  - weakness (especially abduction of index finger)
- ❑ diagnosis
  - history, physical
  - NCV: conduction delay across elbow
- ❑ management
  - conservative: prevent repeated minor trauma (e.g. leaning on elbow or sleeping with hand under head), elbow pads, NSAIDs
  - surgical: nerve decompression and transposition to front of elbow

Less Common Entrapments

- ❑ common peroneal nerve
  - superficial and fixed behind fibular head
  - sensitive to trauma (e.g. fracture of fibular head)
  - motor: decreased foot and toe extension ("drop foot"), decreased ankle eversion
  - sensory: decreased lateral foot and dorsum (less common)
  - distinguish from L5 radiculopathy
- ❑ lateral cutaneous nerve of the thigh ("meralgia paraesthetica") – pain over anterior/lateral aspect of thigh (common in obese people,patients post-iliac bone grafts)
- ❑ motor branch of ulnar nerve at wrist (Guyon's canal)
- ❑ posterior tibial nerve ("Tarsal Tunnel")
- ❑ radial nerve ("Saturday Night Palsy") (more often a pressure palsy)
- ❑ thoracic outlet syndrome
  - compression of the lower portion of the brachial plexus (which supplies the ulnar nerve) as it emerges from the axilla, through a narrow passage beneath the clavicle and between the anterior and middle scalene muscles, while resting on the first rib
  - hard to diagnose
  - rule out Pancoast tumour (associated with Horner's Syndrome) as this may mimic thoracic outlet syndrome (do chest x-ray)

PAIN SYNDROMES

PHYSIOLOGY OF PAIN

- ❑ peripheral sensors: free nerve endings
- ❑ neurotransmitters: substance p, endorphins
- ❑ gate control theory:
  - summation of inhibitory and excitatory afferent input at the synapse to the second order neuron of the spinothalamic tract determines amount of pain felt
  - segmental and higher center influence the perception of pain
- ❑ perception
  - thalamus to cerebral cortex —> awareness
  - personality and mood —> intensity
  - spinothalamic tract, reticular formation and limbic system —> unpleasant, emotional aspect

MEDICAL TREATMENT

- ❑ acute pain (< 2-3 weeks duration): analgesics +/– tranquilizers
- ❑ benign chronic pain: antidepressants, anticonvulsants, topical (capsicin), NOT narcotics or sedatives
- ❑ malignant chronic pain: strong narcotics in frequent, small doses

SURGICAL TREATMENT

Central

- ❑ stereotactic thalamotomy
  - remove spinoreticular relay
  - indication: malignancy of head, neck or brachial plexus
- ❑ deep brain stimulation
  - stimulation of electrodes placed in periventricular gray matter, sensory relay nucleus of thalamus or internal capsule +/– radiocontrolled stimulator subcutaneously
- ❑ hypophysectomy (chemical: uses alcohol)
  - unknown mechanism
  - indication: metastatic disease
- ❑ dorsal root entry zone lesions
  - indication: deafferentation pain (brachial plexus avulsion, postherpetic neuralgia)
  - major complication: ipsilateral leg weakness
- ❑ percutaneous anterolateral cordotomy
  - lesion of spinothalamic tract giving pain relief contralaterally
  - 90% patients respond
  - complications: respiratory difficulties and ipsilateral limb weakness
- ❑ commisural myelotomy
  - division of decussating pain fibers for temporary pain relief
  - indication: terminal malignancy
- ❑ dorsal column stimulation
  - percutaneous electrodes in epidural space
  - indication: intractable chronic pain

Peripheral

- ❑ nerve blocks
  - dermatomal pain relief, loss of motor and sympathetic function
  - permanent: neurolytics (phenol, alcohol)
  - temporary: local anesthetics
  - paravertebral or peripheral: NOT neurolytics —> painful neuritis
- ❑ transcutaneous electrical nerve stimulation (TENS)
  - prolonged stimulation of large diameter fibers inhibiting ascending pain fibers or via higher centers
- ❑ dorsal rhizotomy
  - dorsal root division
  - infrequently done: high failure rate and short effect
- ❑ denervation of facet joints
  - cut posterior ramus of spinal nerves
  - temporary: relief until nerve regrows

TIC DOLOUREUX

Clinical Features
- ❑ older age
- ❑ location: V2 + V3 > V2 > V3 > V1 + V2 > V1 + V2 + V3 > V1
- ❑ R>L
- ❑ F:M=2:1
- ❑ short, sharp jabs in series, last a few seconds to a few minutes
- ❑ violent, terrible, "lightning", "electrical", lancinating pain, distinguish from burning pain
- ❑ may be weeks or months of remission
- ❑ neurological examination normal
- ❑ if other neurological findings - consider other diagnosis
- ❑ triggers: areas on face (especially around mouth), wind, eating, drinking, talking (proprioceptive fibers)

Diagnosis
- ❑ history
- ❑ rule out structural lesion affecting trigeminal nerve (tumour, aneurysm) - CT, MRI
- ❑ may be due to multiple sclerosis (especially in younger patients)

Management
- ❑ pharmacologic
  - drug of choice is carbamazepine 200 mg tabs, 3-5 per day
  - phenytoin is second choice
  - baclofen (potentiates carbamazepine effect)
  - other: sodium valproate, gabopentin, lomotrigine
  - response to medication is almost diagnostic
  - eventually becomes refractory
- ❑ procedures
  - percutaneous thermocoagulation of CN V
  - percutaneous balloon compression of CN VI ganglion
  - glycerol injection into Meckel's cave (trigeminal cistern)
  - division/avulsion of branches of CN V in face
  - microvascular decompression of CN V at pons

REFLEX SYMPATHETIC DYSTROPHY (CAUSALGIA)

Etiology
- ❑ incomplete peripheral nerve injury in nerve with sympathetic fibers

Clinical Features
- ❑ intense, continuous, burning pain
- ❑ touch worsens pain
- ❑ red, warm, dry and swollen skin initially (sympathetic overactivity)
- ❑ cool, clammy, glossy and atrophic skin in advanced stages

Treatment
- ❑ sympathetic nerve blockade: medical or surgical

POSTHERPETIC NEURALGIA

Etiology
- ❑ reactivation of latent varicella zoster virus that lay dormant in dorsal root or gasserian ganglion

Clinical Features
- ❑ burning, constant pain
- ❑ severe, sharp paroxysmal twinges over area of affected sensory neurons
- ❑ touch worsens pain

Treatment
- ❑ no specific treatment and is difficult to treat
- ❑ medical: antidepressants, carbamazepine, ethychloride spray (temporary relief), topical (capsicin, promising treatment that works by blocking substance p), steroid injection or topical in eye during acute eruptive phase (decreases severity of pain and decreases corneal scar), gabopentin
- ❑ surgical: percutaneous cordotomy, possible dorsal root entry zone lesion

THALAMIC PAIN

Clinical Features
- ❑ begins with hemianesthesia (due to thalamic infarction or hemorrhage)
- ❑ becomes diffuse, burning pain contralateral to lesion
- ❑ worse with light touch (e.g. clothing)
- ❑ may have prior history of thalamic stereotactic procedure for movement disorder

Treatment
- ❑ medical: poor response to medication
- ❑ surgical: stereotactic thalamic stimulation but may increase sensory deficit

PHANTOM LIMB PAIN

Etiology
❑ complication of 10 % amputation patients

Clinical Features
❑ continuous burning
❑ pain from some point on missing limb

Treatment (see Postherpetic Neuralgia section)

PEDIATRIC NEUROSURGERY

SPINA BIFIDA

Spina Bifida Occulta
❑ definition
- congenital absence of a spinous process and variable amounts of lamina
- no visible exposure of meninges or neural tissue

❑ epidemiology
- 20-30% of the general population

❑ etiology
- failure of fusion of the posterior arch

❑ clinical features
- no obvious external markings
- no obvious clinical signs
- presence of lumbosacral cutaneous abnormalities (dimple, sinus, port- wine stain, or hair tuft) should increase suspicion of an underlying anomaly (lipoma, dermoid, diastomatomyelia)

❑ investigations
- plain film: absence of the spinous process along with minor amounts of the neural arch
- most common at L5 or S1

❑ treatment and results
- requires no treatment

Meningocele (Spina Bifida Aperta)
❑ definition
- a defect consisting of a herniation of meningeal tissue and CSF through a defect in the spine

❑ etiology
- 2 theories
  - primary failure of neural tube closure
  - rupture of a previously closed neural tube due to overdistension (Gardner; unpopular theory)

❑ clinical features
- most common in lumbosacral area
- usually no disability
- low incidence of associated anomalies and hydrocephalus

❑ investigations
- plain films, CT, MRI, U/S, ECHO, genitourinary (GU) investigations
- MRI of entire spinal column because increased liklihood of additioal anomalies

❑ treatment and results
- surgical excision (excellent results)

Myelomeningocele
❑ definition
- a defect consisting of a herniation of meningeal tissue and CNS tissue through a defect in the spine

❑ etiology - same as meningocele

❑ clinical features
- sensory and motor changes distal to anatomic level producing varying degrees of weakness, anesthesia, urine and fecal incontinence
- 65-85% of patients with myelomeningocele have hydrocephalus
- most patients with myelomeningocele have Type II Chiari malformation

❑ investigations
- plain films, CT, MRI, U/S, ECHO, GU investigations

❑ surgical indications
- preserve intellectual, sensory and motor functions
- prevent CNS infections

❑ results
- operative mortality close to 0%
- 95% 2 year survival
- 80% have IQ in > 80 (but most are 80-95)
- 40-85% ambulatory
- 3-10% have normal urinary continence
- most common cause of early mortality are complications from chiari malformation (respiratory arrest and aspiration), whereas late mortality is due to shunt malfunction

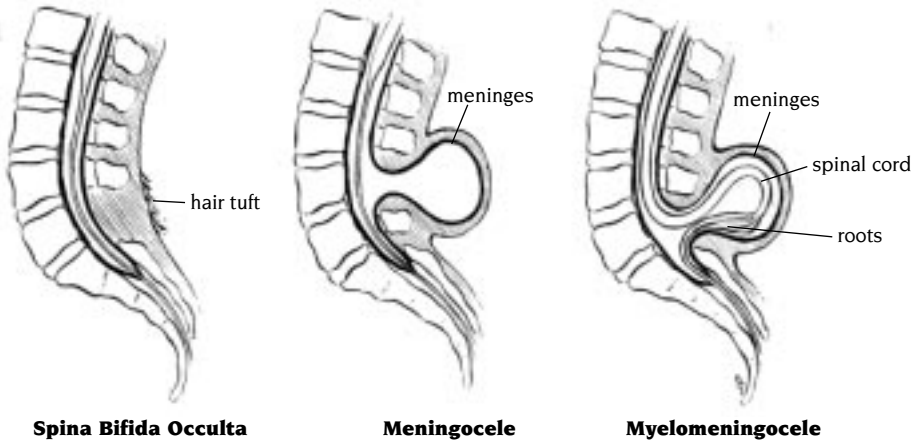

Figure 14. Diagram Showing Spina Bifida Occulta, Meningocele, Myelomeningocele

Illustration by Jen Polk

INTRAVENTRICULAR HEMORRHAGE (IVH)

Definition
- ❑ a disease of the premature, low-birth weight infant
- ❑ consists of hemorrhage into the germinal matrix of the developing brain

Classification (based on ultrasound or CT)
- ❑ Grade I: germinal matrix (subependymal) hemorrhage only
- ❑ Grade II: blood filling lateral ventricles without distention
- ❑ Grade III: blood filling and distending lateral ventricles
- ❑ Grade IV: hemorrhage with parenchymal involvement (ICH)

Epidemiology
- ❑ occurs in 45% of infants born with a birth weight of 1500 g or less
- ❑ uncommon after 32 weeks of gestation
- ❑ essentially nonexistent in full-term infants

Predisposing Factors
- ❑ low gestational age
- ❑ high cerebral blood flow and cerebral perfusion pressure
    - birth asphyxia, resuscitation
    - respiratory distress syndrome
    - rapid volume re-expansion
    - hypoxemia, hypercarbia, acidosis
    - seizure, pneumothorax

Investigations
- ❑ ultrasound is the method of choice to screen for ICH/IVH
- ❑ should be done routinely to screen preterm babies < 24 weeks gestation or < 1,500 gm
- ❑ CT scan will also show ICH and IVH as described above

Treatment
- ❑ best to withhold tapping ventricles, ventriculostomies, and shunting until blood has cleared
- ❑ if progressive hydrocephalus develops, then
    - serial LP
    - acetazolamide (25-100 mg/kg/day) and Lasix (2 mg/kg/day)
    - ventriculostomy
    - shunt (low pressure)

Results
- ❑ grade I-III hemorrhages can do as well as children without hemorrhages
- ❑ grade IV: only 50% chance of attaining normal life status
- ❑ prognosis more dependent on the degree of asphyxia than on hydrocephalus

HYDROCEPHALUS IN PEDIATRICS

Etiology
❑ congenital
- aqueductal anomalies
  - primary aqueductal stenosis in infancy
  - secondary gliosis due to intrauterine viral infections (mumps, varicella, TORCH) or germinal plate hemorrhage
- Dandy Walker (2-4%)
- Chiari malformation, especially Type II
- myelomeningocele

❑ acquired
- post meningitis
- post hemorrhage (SAH, IVH)
- masses (vascular malformation, neoplastic)

Clinical Features
❑ symptoms and signs of hydrocephalus are age related in pediatrics
❑ increased head circumference (HC)
❑ irritability, lethargy, poor feeding and vomiting
❑ bulging anterior fontanelle
❑ widened cranial sutures
❑ "cracked pot" sound on cranial percussion
❑ scalp vein dilation (increased collateral venous drainage)
❑ sunset sign - forced downward deviation of eyes
❑ episodic bradycardia and apnea

Management
❑ similar to adults (see Hydrocephalus section)

DANDY-WALKER MALFORMATION

Definition
❑ atresia of foramina of Magendie and Luschka, resulting in
- complete or incomplete agenesis of the cerebellar vermis with widely separated, hypoplastic cerebellar hemisphere
- posterior fossa cyst
- dilatation of 4th ventricle
- enlarged posterior fossa

❑ associated anomalies
- hydrocephalus (90%)
- agenesis of corpus callosum (17%)
- occipital encephalocele (7%)

Epidemiology
❑ 2-4% of pediatric hydrocephalus

Clinical Features
❑ 20% are asymptomatic
❑ only 50% have normal IQ
❑ symptoms and signs of hydrocephalus combined with a prominent occiput in infancy
❑ ataxia, spasticity, poor fine motor control common in childhood
❑ seizures occur 15%

Treatment
❑ asymptomatic patients require no treatment
❑ associated hydrocephalus requires surgical treatment

CHIARI MALFORMATIONS

Definition
❑ malformations at the medullary-spinal junction

Clinical Features
❑ Type I (cerebellar ectopia): cerebellar tonsils lie below the level of the foramen magnum
- average age at presentation 41 years
- brain compression: suboccipital headache, nystagmus, ataxia, spastic quadraparesis
  - foramen magnum compression syndrome (22%): ataxia, corticospinal and sensory deficits, cerebellar signs, lower cranial nerve palsies
  - central cord syndrome (65%): dissociated sensory loss, occasional segmental weakness and long tract signs
  - cerebellar syndrome (11%): truncal/limb ataxia, nystagmus, dysarthria
- hydrocephalus (10%)
- syringomyelia (50%)

❑ Type II: part of cerebellar vermis, medulla and 4th ventricle extend through the foramen magnum often to midcervical region
- present in infancy
- findings due to brain stem and lower cranial nerve dysfunction: swallowing difficulties, apneic spells, stridor, aspiration, arm weakness
- syringomyelia, hydrocephalus in > 80%

❑ Type I and Type II: if symptomatic

Investigations
❑ MRI or CT myelography

Treatment
❑ surgical decompression - indications
- Type I: symptomatic patients (early surgery recommended)
- Type II: neurogenic dysphagia, stridor, apneic spells

CRANIOSYNOSTOSIS

Definition
❑ premature closure of the cranial suture(s)

Classification
❑ sagittal (most common)
- long narrow head with ridging sagittal suture (scaphocephaly)

❑ coronal
- expansion in superior and lateral direction (brachiocephaly)
- bilateral coronal craniosynostosis often associated with Crouzon's and Apert's syndrome

❑ lambdoid - least common
❑ metopic (forehead)
❑ multiple suture synostosis or pansynostosis

Epidemiology
❑ 0.4/1,000 live births
❑ most cases are sporadic
❑ familial incidence is 2% of sagittal and 8% of coronal synostosis

Clinical Features
❑ skull deformity
❑ raised ICP
❑ ophthalmologic problems
- due to increased ICP or bony abnormalities of the orbit
- strabismus most common

❑ hydrocephalus may accompany multiple craniosynostoses

Investigations
❑ plain radiographs, CT scan (3D)
❑ bone scan: increased activity during active phase of union, decreased once union has occurred

Management
❑ parental counseling about
- nature of deformity
- difficulty growing up as "cone head"
- associated neurological symptoms

❑ surgery for cosmetic purposes, except in cases of elevated ICP

PEDIATRIC BRAIN TUMOURS

❑ 20% of all pediatric cancers (second only to leukemia)
❑ 80% of pediatric tumours are infratentorial (see Intracranial Mass section)
❑ most common manifestations
- vomiting
- arrest or regression of developmental milestones
- macrocrania
- poor feeding, failure to thrive
- hydrocephalus
- seizures

CHILD ABUSE (Shaken Baby Syndrome) (see Pediatrics Chapter)

❑ subdural hematomas of various ages
❑ retinal hemorrhages
❑ skull fractures

DRUGS

❑ the following are guidelines ONLY; follow clinical judgment and up-to-date prescription recommendations in practice; dosages refer to adults unless otherwise specified

Carbamazepine (Tegretol)
❑ Tic Douloureux
- dosage: 100 mg PO bid, increase by 200 mg/day up to a maximum of 1,200 mg/day divided tid
- usual optimum dosage: 200 mg tid

❑ seizures
- dosage: 600-2,000 mg/day, start low and increase in small increments (inpatient: every 3 days; outpatient: every week)
- usual optimum dosage: 800-1,200 mg/day
- monitor CBC (potential hematological toxicity)

Dexamethasone (Decadron)
❑ cerebral edema (e.g. secondary to tumour, head injury, pseudotumor cerebri)
❑ preoperative preparation for patients with increased ICP secondary to brain neoplasms
❑ palliation in recurrent inoperable brain neoplasms
- dosage: loading: 10-20 mg IV maintenance: 4-6 mg IV/day divided qid (may be given PO)

Lorazepam (Ativan)
❑ status epilepticus
- dosage: 4 mg IV over 2 minutes, q5 minutes
- start phenytoin loading simultaneously

Mannitol
❑ raised ICP
- dosage: 1 gm/kg IV rapid infusion (350 mL of 20% solution) followed by 0.25 g/kg IV q6h
- effect occurs in 1-5 minutes, maximal at 20-60 minutes
- often alternated with furosemide (Lasix) 10-20 mg IV q6h
- indwelling urinary catheter

Nimodipine (Nimotop)
❑ vasospasm in SAH
- dosage: 60 mg PO/NG q4h x 21 days, started within 96 hours of SAH
- causes vasodilation
- only calcium channel blocker (CCB) to cross BBB (blood brain barrier)
- use half the normal dose for liver failure
- monitor BP

Phenytoin (Dilantin)
❑ seizures
- dosage: loading: 18 mg/kg IV
  maintenance: 200-500 mg IV/day
  or
  rate: 40-50 mg
  loading: 300-600 mg PO/day divided bid/tid
  maintenance: 300 mg PO q4h until 17 mg/kg given
  (average maintenance dose: 300 mg/day PO)
- important to give over time to prevent causing a cardiac arrest

❑ status epilepticus
- dosage: 1200 mg IV over 30 minutes (~ 20 mg/kg) (if patient not on phenytoin regularly)
  500 mg IV over 10 minutes (if already on phenytoin)

SURGICAL PROCEDURES

Lumbar Puncture

❑ objective
- to enter the subdural space to measure or reduce cerebrospinal fluid (CSF) pressure or obtain CSF for analysis

❑ indications
- meningitis
- encephalitis
- meningeal carcinomatosis
- subarachnoid hemorrhage
- pseudotumour cerbri

❑ anatomical landmarks
- conus medullaris (spinal cord termination) is usually located around L1/L2
- superior border of posterior iliac crests aligns with the spinous process of L4 (may be variable)
- insert needle with a slight cephalad inclination into the L4-5 interspace until the subdural space is entered

❑ complications
- tonsillar herniation
- infection
- headache ("low pressure")
- spinal epidural hematoma
- nerve root trauma
- vestibulocochlear dysfunction
- ocular abnormalities
- dural sinus thrombosis

Burr Hole

❑ objective
- to decompress brain by removing a compressive fluid lesion through a small opening in the skull (called a burr hole)
- because it is small, cranioplasty is not generally used to fill-in the burr hole

❑ indications
- subdural hematoma
- subdural hygroma
- brain abscess
- ventriculostomy
- biopsy

❑ anatomical landmarks
- varies according to location of trauma or CT/MRI-identified lesion

❑ complications
- seizures
- intracerebral hemorrhage (0.7-5%)
- failure of brain to re-expand
- re-accumulation of compressive fluid
- tension pneumocephalus
- subdural empyema
- infection
- biopsy

Craniotomy

❑ objective
- to gain exposure to any structural lesion in the brain by removing a section of skull (called a bone flap) so that the lesion may be eliminated without harming intact brain
- once the lesion is eliminated the bone flap is fixed back in place

❑ indications
- brain tumour
- brain abscess
- intracranial aneurysm
- hematoma
- lobectomy
- biopsy

❑ anatomical landmarks
- numerous approaches depending on the site of the lesion (e.g. frontal, frontotemporal, temporal, subtemporal, pterional, petrosal, suboccipital)
- depending on the approach, important landmarks include the midsagittal line (overlying the superior sagittal sinus and falx cerebri), coronal suture (often palpable through the scalp; anterior to the precentral gyrus or motor cortex), glabella, nasion

❑ complications
- intracerebral hemorrhage (0.8-1.1%)
- hematoma
- infection
- cerebral infarction
- seizure
- acute hydrocephalus
- pneumocephalus
- cerebral edema
- vasospasm

Transphenoidal Tumour Resection

❑ objective
- to access suprasellar lesions and remove them without disturbing surrounding cerebral, neural, or vascular structures

❑ indications
- pituitary adenoma
- craniopharyngioma
- Rathke's cleft cyst
- meningioma
- germinoma
- epidermoid tumour

❑ anatomical landmarks
- the sella turcica is approached anteriorly through the nasal cavities to the midline nasopharynx which forms the anterior wall of the sphenoid sinus
- the posterior wall of the sphenoid sinus is the anterior wall of the sella turcica
- maintaining a midline approach is essential to avoid injury to the internal carotid arteries

❑ complications
- CSF leakage
- infection
- hemorrhage
- midbrain or pontine injury
- diabetes insipidus
- panhypopituitarism
- sinusitis

Spinal Decompression

❑ objective
- to remove or repair structural abnormalities compressing the spinal cord or nerve roots

❑ indications
- interverterbral disc herniation
- spondylolisthesis
- spinal stenosis
- spinal fracture or dislocation
- spinal tumour
- cauda equina syndrome

❑ anatomical landmarks
- various approaches depending on the location of the lesion (e.g. anterior, anterolateral, posterior, postolateral, cervical, throacic, lumbosacral)
- depending on the lesion site, important landmarks include
  - the transverse processes of C1 (palpable between the mastoid processes and the angles of the jaw)
  - vetebra prominens (indicating the spinous process of C7)
  - spinous process of L4 (indicated by an imaginary line between the superior margins of the posterior iliac crests)
  - spinous process of S2

❑ complications
- infection
- hemorrhage
- incidental durotomy
- neural injury
- great vessel injury
- arachnoiditis
- reflex sympathetic dystrophy (RSD)
- bowel perforation

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