

**Version 21: 28**

**Doc tutorial PPT(NEET SS Surgery)**

**prepared under**

**RRM NEET SS Team of Faculties**



**Next Offline  
Face to Face  
NEET SS Class**

**From**

**Dec 2024 Onwards**

**For Details 6384111333**



**RRM'S NEET SS**  
CHENNAI - DELHI

# NEW PATTERN **GENERAL SURGERY**

OFFLINE FACE TO FACE CRASH COURSE CLASS MATERIALS

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AUTHOR.....

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October 2023

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from Dec 19 to 24<sup>th</sup> of 2023  
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# Bailey based Neurosurgery

*Dr. R SSK*

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Raised ICP

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## High pressure headaches

- nausea and vomiting,
- blurred vision and double vision;
- **cranial nerve compression** can result in eye movement and pupil abnormalities.
- Fundoscopy can detect **papilloedema**, but this takes time to develop so may be absent in the acute phase!!!

- High pressure headache, worse on **coughing or bending forward**
- Low pressure headaches, encountered after excessive cerebrospinal fluid (CSF) drainage, are typically **worse on standing**.

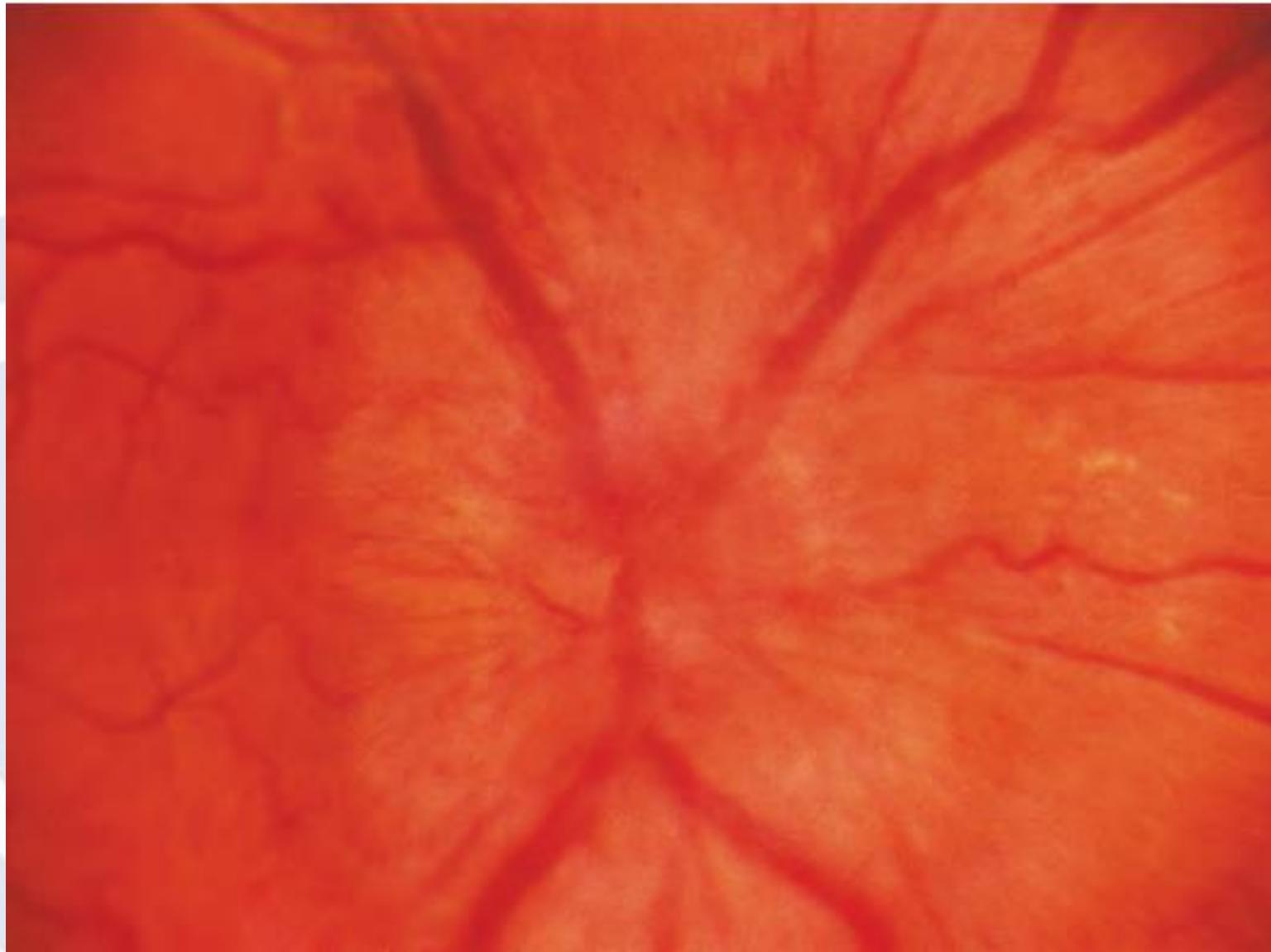
## Infants

- Fontanelle is tense and bulging,
- increase in head circumference
- **bulging scalp veins.**
- As pressure rises, **conscious level is impaired.**
- In children **Parinaud's syndrome** results from dorsal midbrain compression, with a loss of upgaze known as **sunsetting**



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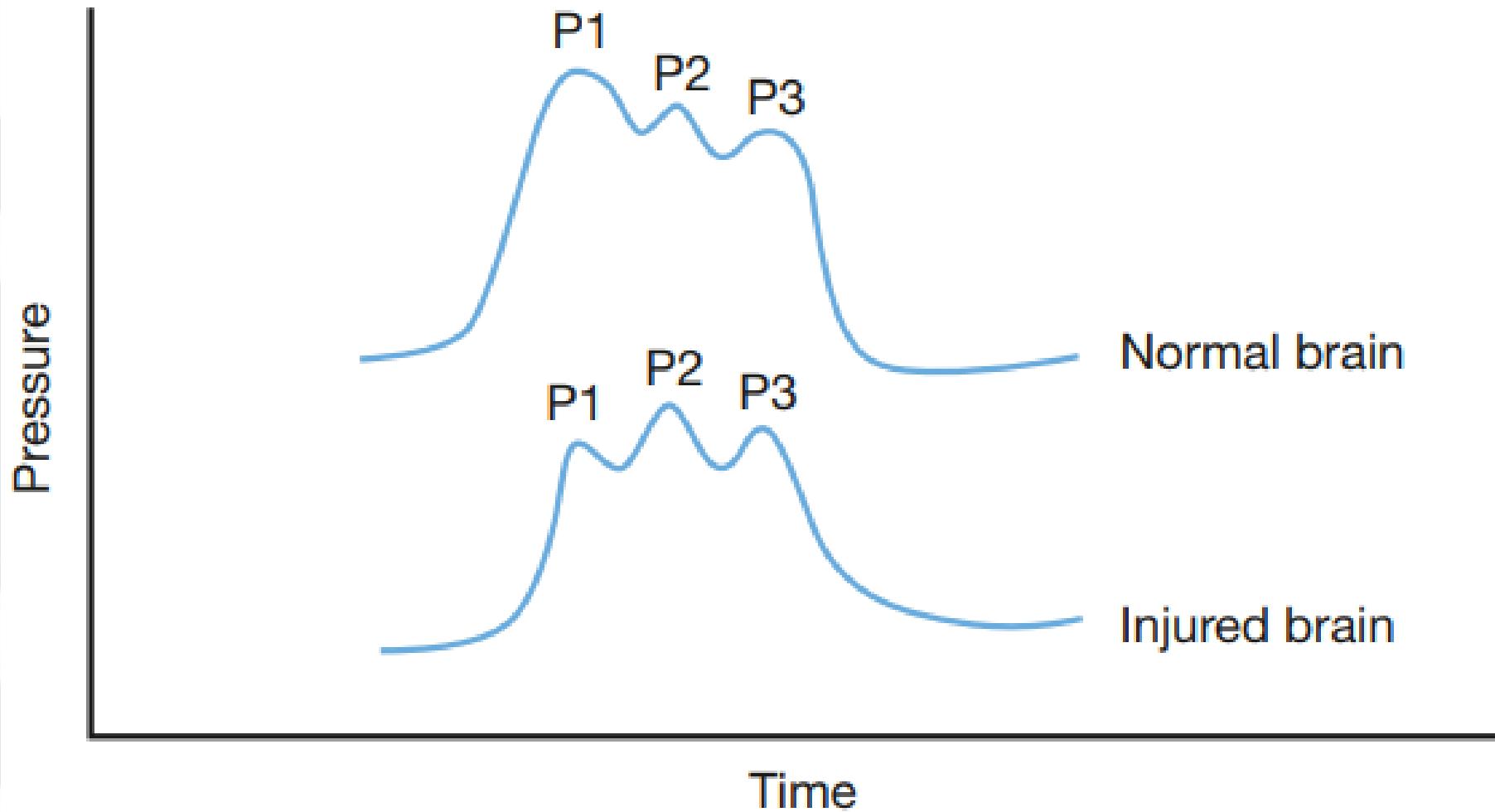
TEST



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- The P1 percussion wave corresponds to arterial pulsation.
- Reduced brain compliance in the setting of traumatic brain injury among others is associated with a prominent P2 tidal wave.
- The P3 dicrotic wave represents venous pulsation.



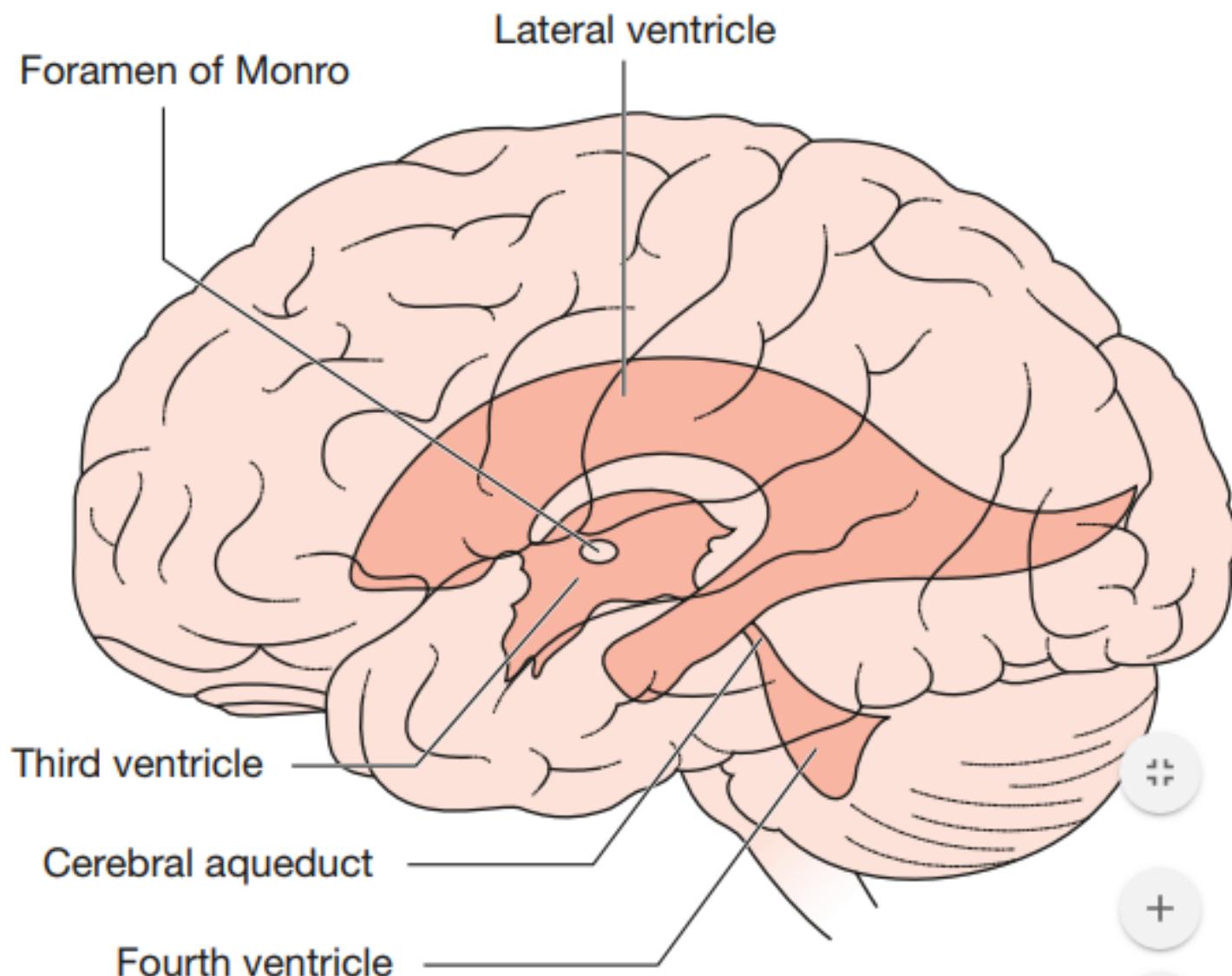
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Hydrocephalus

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- The total CSF volume is normally about **150mL**.
- Production from the walls of the ventricles and the choroid plexus is about **20 mL per hour**.
- Hydrocephalus refers to an increase in CSF volume with ventricular enlargement, often presenting symptoms of raised ICP.



- CSF flows from the **lateral ventricles** through the **foramen of Monro** to the **third ventricle**, then down the **cerebral aqueduct** to the **fourth ventricle**, where it exits to the subarachnoid space via the midline foramen of Magendie and the lateral foramina of Luschka.
- CSF is **reabsorbed** into the arachnoid villi along the **superior sagittal sinus**.

- Hydrocephalus almost always reflects obstruction to circulation (an obstructive hydrocephalus) or failure of reabsorption (a communicating hydrocephalus)
- *The distinction is important since obstructive hydrocephalus especially can cause very sudden deterioration with coma and death, and because lumbar puncture in this context carries a risk of herniation of the brainstem and cerebellar tonsils due to the resulting differential pressure changes (sometimes termed 'coning').*

# Hydrocephalus

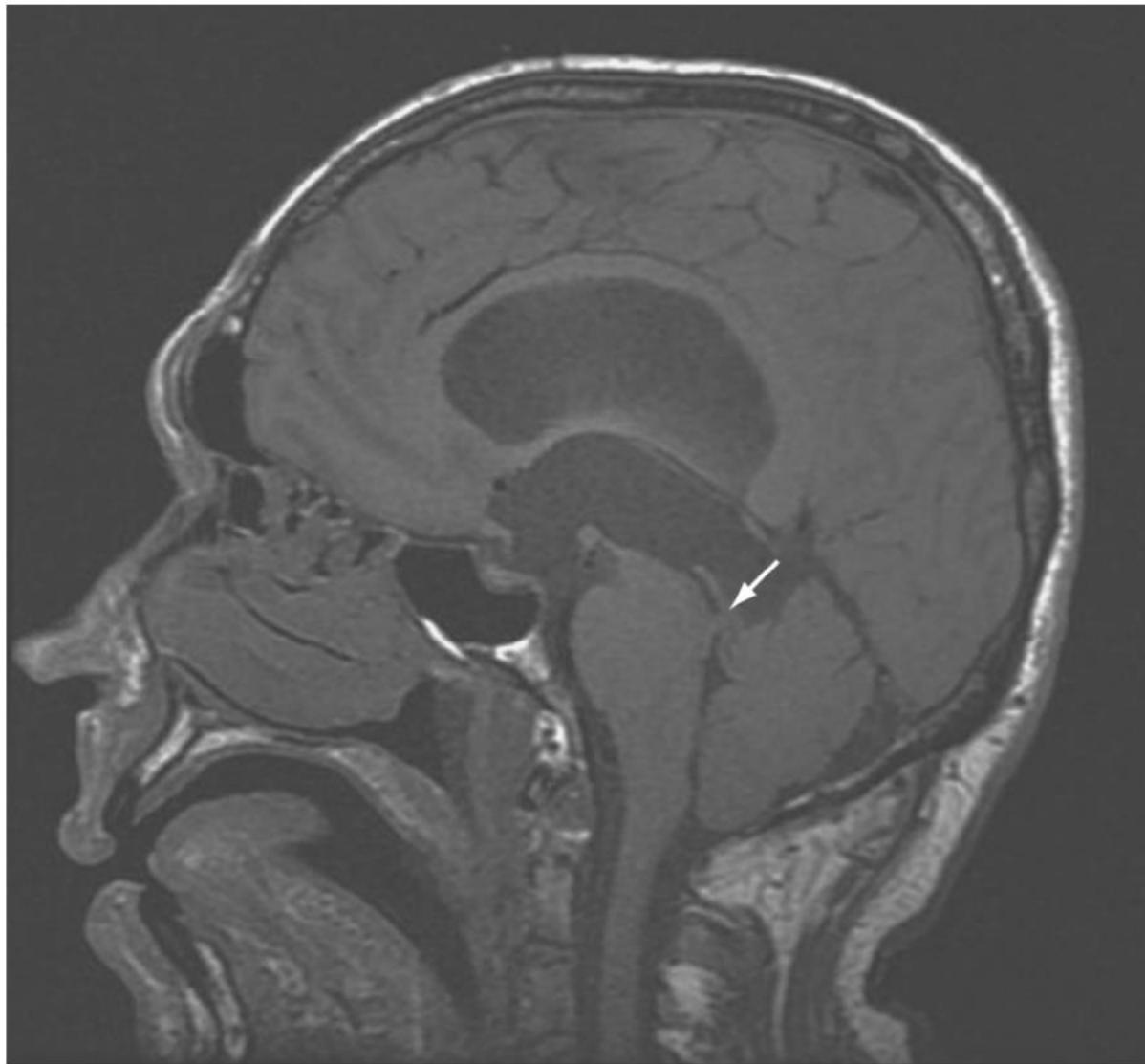
- In obstructive hydrocephalus, the obstruction is at or proximal to the fourth ventricular outlet foramina (foramen of Magendie and Luschka).
- If the obstruction is beyond the fourth ventricular outlet foramina (**cisterns** or arachnoid granulations), it is classified as **communicating** hydrocephalus.
- Common examples of obstructive hydrocephalus are aqueductal stenosis and hydrocephalus associated with tumors

# Acute and chronic hydrocephalus

- Hydrocephalus developing within days or a few weeks (e.g., hydrocephalus due to tumor) is manifested with **rapid** progression of symptoms known as acute hydrocephalus.
- CSF accumulation during months (or even years) presents with subtle signs of **memory impairment, walking difficulty, or urinary incontinence** and is termed chronic hydrocephalus.
- A classic example of chronic hydrocephalus is normal-pressure hydrocephalus.

# Congenital and acquired hydrocephalus

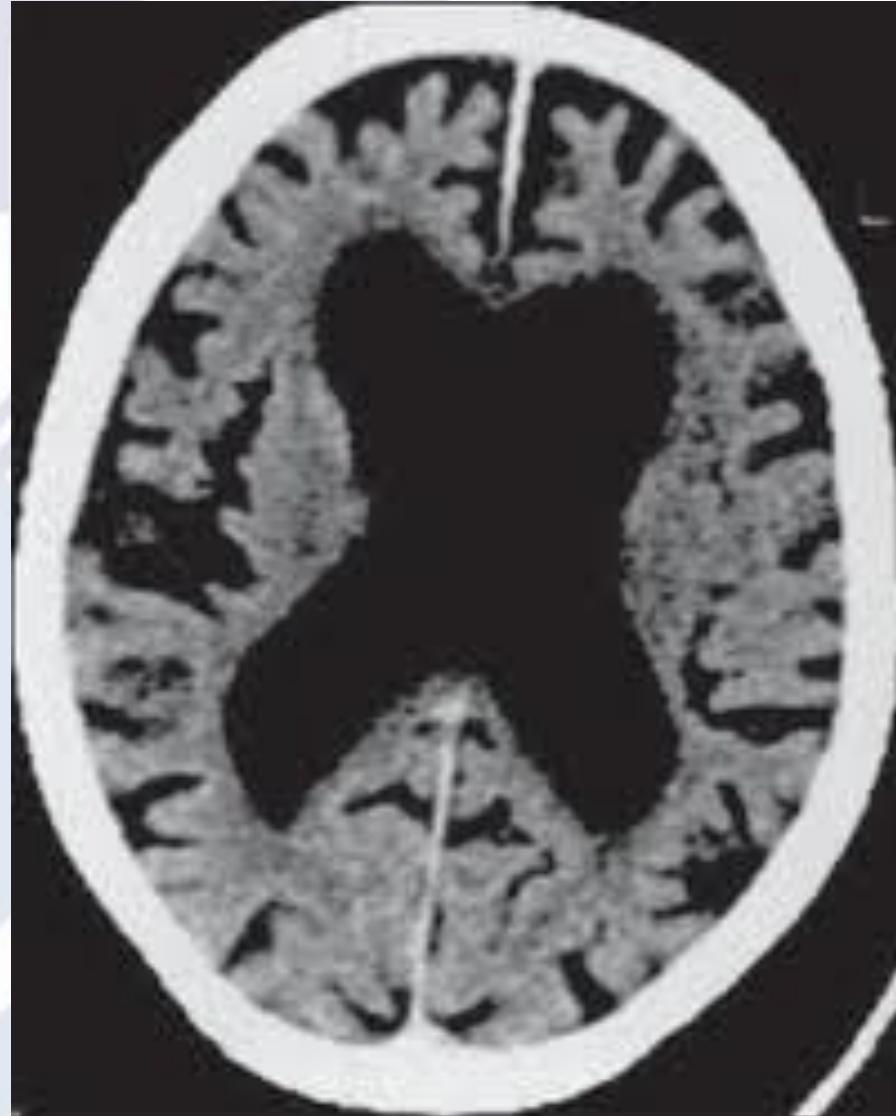
- Hydrocephalus present at birth is known as congenital hydrocephalus. Although congenital hydrocephalus is commonly obstructive in nature, it can be *communicating, as seen in intrauterine toxoplasmosis or cytomegalovirus infections.*
- In acquired hydrocephalus, the pathologic process starts after birth and includes post-traumatic hydrocephalus, hydrocephalus associated with tumors, and normal-pressure hydrocephalus.



**FIGURE 67-25** T1-weighted sagittal MRI scan of patient with gross obstructive hydrocephalus caused by aqueductal stenosis (arrow).

# Hydrocephalus ex vacuo

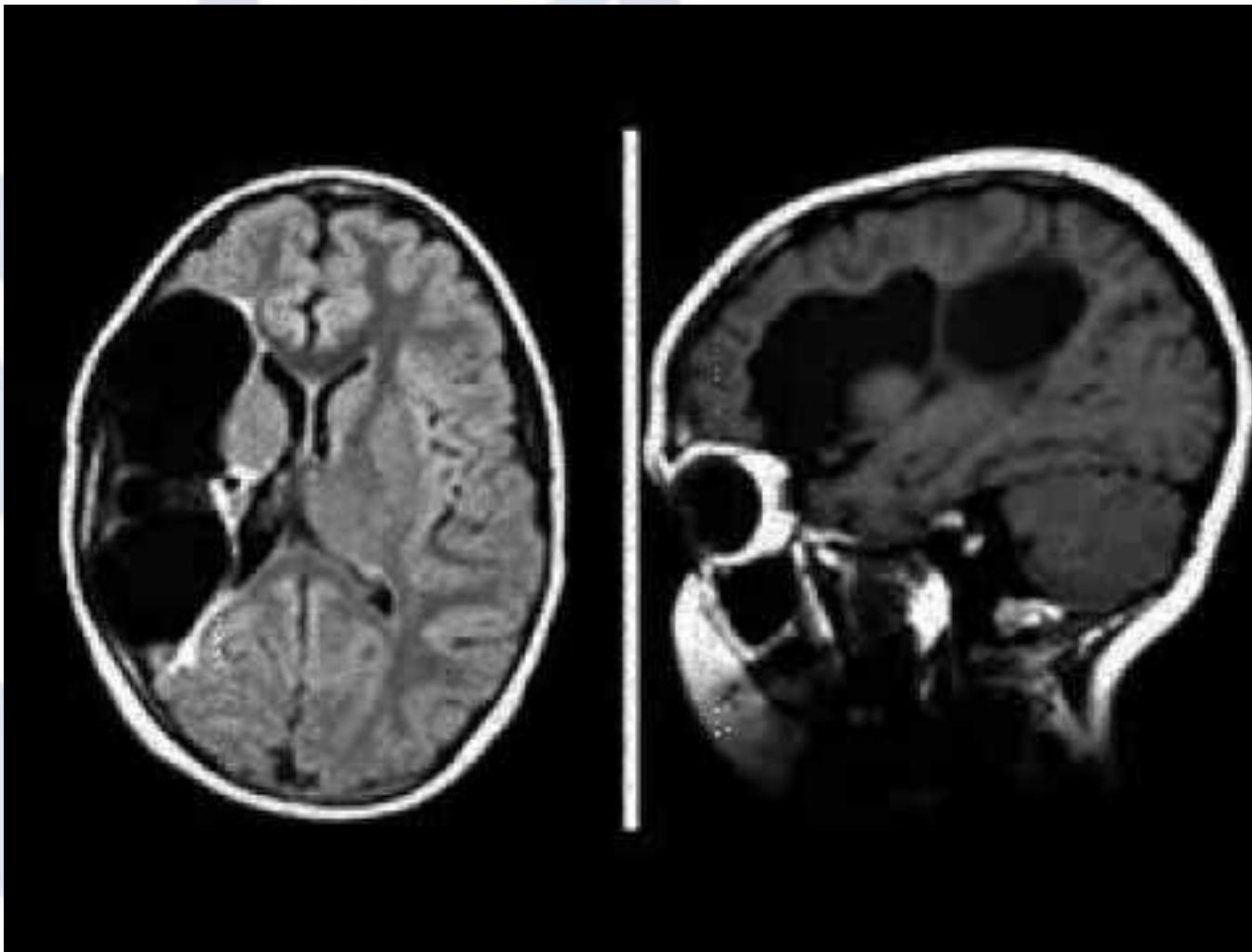
- Ventricles enlarge **compensatory** to overall shrinking of the brain tissue. Really the **enlargement** of the ventricles is due to the shrinkage of the brain tissue.
- Seen in advanced age with **brain atrophy**, after **diffuse head injury or stroke, and with various neurodegenerative conditions.**



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# Porencephaly

- a focal brain substance has suffered some loss of volume (e.g., stroke, postsurgical change in volume) leading to collection of CSF in the cavity.
- **Porencephalic cyst** is usually differentiated from hydrocephalus ex vacuo by its localized nature.



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# Arrested hydrocephalus

- Ventricles are large with the patient having no significant symptoms to require a surgical procedure.



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# Clinical Features of Hydrocephalus

- The symptoms differ considerably in **different age groups**.
- In infants, a thin and relatively **nonrigid skull allows an overall cranial expansion**
- In older children and adults, the rigid fused skull prevents its enlargement

- Infantile hydrocephalus, either the infant is born with a large head or the head abnormally grows during the first few months of life.
- **Anterior fontanelle** is usually full
- In extreme cases, a relatively higher ICP causes the blood to be diverted from the intracranial to the extracranial compartment, resulting in prominent and dilated scalp veins.

- A late feature is the classic “sunset sign” manifested with **downward deviation of the eyeballs (like a setting sun)**. This is due to compression of the **midbrain tectum** by the posterior part of the dilated third ventricle.
- In later stages, the child will be **irritable** and fussy and may not accept feeds. There may be associated **vomiting**.
- Lethargy, drowsiness, and lapsing into the **comatose** state will follow if the child remains untreated.

- In older children and adults, fusion of the skull bones no longer permits the cranium to enlarge.
- This result in raised ICP and cause **compression** of the adjacent brain.
- Two common modes of presentation: **rapidly progressive hydrocephalus** and **chronic hydrocephalus**.

- Rapidly progressive hydrocephalus
- new-onset **headache** and vomiting
- blurring of **vision**
- **Papilledema** can result in secondary optic atrophy
- **Drowsiness** and progression to coma follow

- Chronic hydrocephalus, the CSF accumulates more slowly, thus gradually compressing the brain.
- The patient becomes progressively *dull*, apathetic, and uninvolved with the surroundings
- ***Memory*** impairment for recent events is commonly seen; the remote memory is usually well preserved.
- short stepped *gait* with a wide stance with unsteadiness
- *Urinary incontinence and urgency*

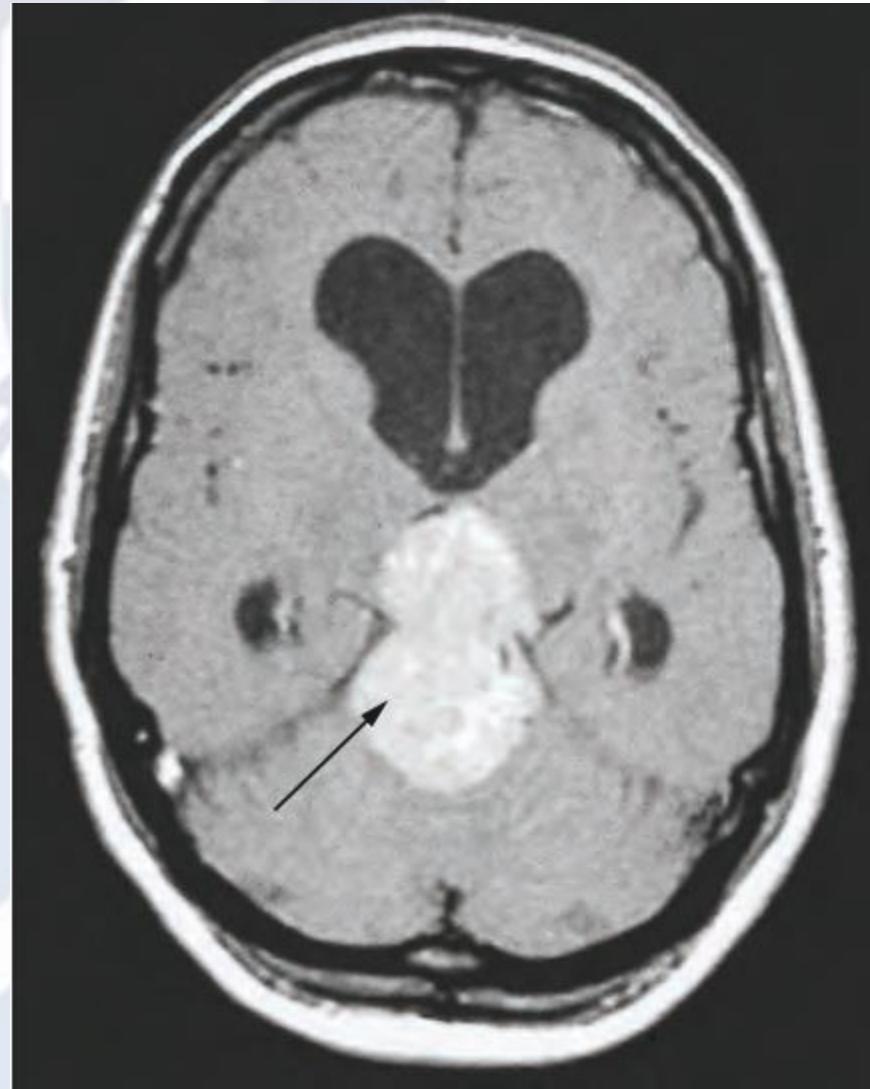
- For communicating hydrocephalus, lumbar puncture is of diagnostic value, deriving an opening pressure and assessment of the CSF contents.
- It is also therapeutic; drainage of typically between **10 and 30 mL of CSF**, with the goal of halving the opening pressure, can relieve the hydrocephalus at least temporarily.

## Normal pressure hydrocephalus

- Reversible dementia
- Triad of **gait disturbance, incontinence and cognitive decline**
- Occur de novo or on a background of previous brain insults including *subarachnoid haemorrhage (SAH), head injury, meningitis and tumour.*
- Ventriculomegaly + cortical atrophy on CT.
- CSF pressure at lumbar puncture (LP) is normal, but it is believed that intermittent elevations in pressure may be involved in the aetiology.
- **Lumbar infusion testing involves insertion of a fine drain at LP, followed by measurement of the CSF pressure changes associated with a fluid challenge administered through this????**

# Investigations

- Cranial **ultrasound** evaluation has been used predominantly in the newborn and infants with open fontanelle.
- The **CT** scan shows dilated ventricles and often indicates the pathologic process and the site of obstruction.
- **MRI** has been the imaging modality of choice in newly diagnosed hydrocephalus.
- MRI is considered essential before considering endoscopic third ventriculostomy and aqueductoplasty, and in assessing the effectiveness of endoscopic third ventriculostomy during the follow-up.



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# Treatment

- **Acetazolamide** - reduces the CSF production.
- High doses of the drug causing metabolic acidosis are required to achieve the effect.
- CSF shunts are tubes with valves that drain the CSF out from one compartment to another.
- The shunt contains three parts: the ventricular end, the valve complex, and the distal end.
- **Shunt malfunction, infection, overdrainage, brain injury, seizures,** and distal complications are the major complications associated with shunts.



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- **High pressure valves may fail to allow adequate CSF drainage,** whereas low pressure valves can overdrain.
- An anti-siphon system may be incorporated to prevent excessive ***drainage in the standing position.***
- Programmable valves offer variable opening pressures, adjusted magnetically using a device applied externally over the valve.
- The valve system will also typically incorporate a CSF reservoir, which allows for percutaneous sampling.

## Shunt complications

- **Overdrainage = low-pressure headaches**, which are typically worse on standing = subdural hygroma or subdural haematoma.
- **Slit ventricle syndrome** = in children treated with shunts, whose ventricles and subarachnoid spaces are underdeveloped, resulting in poor brain compliance.
- Shunts are **vulnerable to disconnection**, infection and blockage, with the result that **15–20% require replacement within 3 years**.

- Shunt malfunction is the predominant complication of shunt procedures.
- **29% of the shunts failed in the first year, requiring reoperation.**
- The incidence of shunt infection, the second significant complication, ranges between 4% and 7%.

- The common organisms include **Staphylococcus epidermidis (50% to 60%)**, **Staphylococcus aureus (20% to 30%)**, **gram-negative bacilli**, and **Propionibacterium species**.
- Usually occur within 3 months of insertion.
- *S. epidermidis* forms a **biofilm** and adheres to the shunt tube, which protects the bacteria against orally or intravenously administered antibiotics. The colonization permits the bacteria to stay quiescent for weeks or sometimes a few months before the infection is manifested.

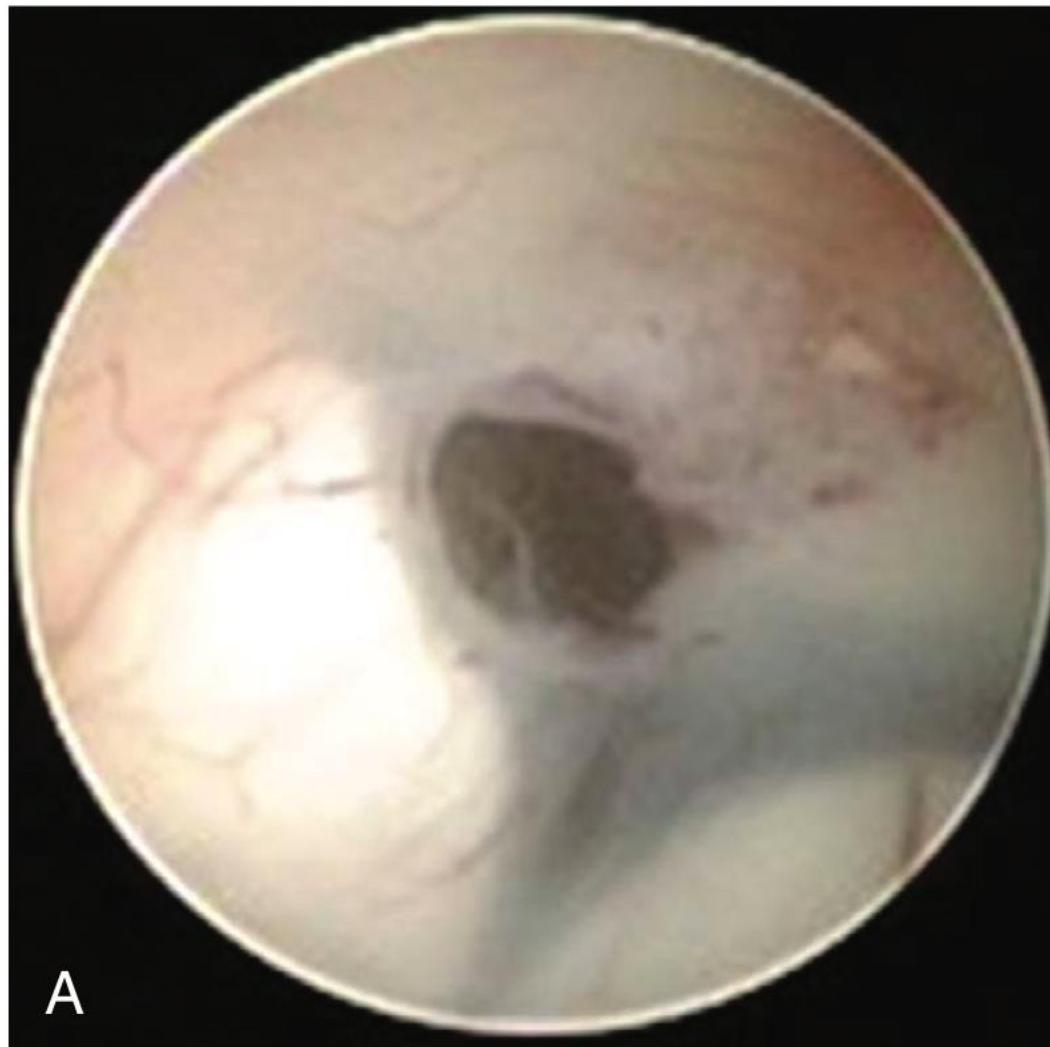
- Shunt infections can be infection of the shunt tube either in its subcutaneous track or in the wound (**wound infection**), infection of the CSF spaces (**meningitis**) or ventricles (ventriculitis), or infection of the abdominal space (**peritonitis**).

- Early subcutaneous infections are manifested with **low-grade fever, redness along the shunt tube, and purulent discharge from the incision.**
- Wound breakage and exposure of the shunt tube can occur.
- Later, as the infection involves the CSF and ventricles, it may be associated with decreased sensorium, seizures, and neurologic deficits.
- If the infection involves the abdominal cavity, it can present with features of **peritonitis**.

- The diagnosis is confirmed by shunt tap and CSF culture.
- Complete removal of the shunt tube is recommended with reinsertion of a new shunt once the infection clears.
- The incidence of shunt infection is reduced by use of catheters **impregnated with rifampicin and clindamycin, which are effective against the gram-positive bacteria.**

## Endoscopic third ventriculostomy

- *Obstructive hydrocephalus due to aqueduct stenosis, tumors of fourth ventricle, and fourth ventricular outlet obstruction*
- The floor of the ventricle is then opened between the mammillary bodies and the pituitary recess.
- Free drainage between the third ventricle and the adjacent subarachnoid cisterns is then possible, without the infection risk posed by implanted tubing.
- Complications include damage to the **basilar artery, or fornacial damage resulting in permanent memory impairment.**



A



B

**FIGURE 67-26** **A**, Endoscopic view of the third ventricular floor after the third ventriculostomy. **B**, Follow-up MRI scan 4 years later demonstrating good flow at the fenestration site (*arrow*).

- **Aqueductoplasty**, the obstructed aqueduct is recanalized with the help of a 3 Fr Fogarty catheter under direct endoscopic vision
- **Aqueductal stenting**, a stent is placed in the aqueduct to prevent further reclosure. The stent is usually attached to a subcutaneous reservoir to prevent its migration.

# Shunts and Appendicitis

- Uncomplicated appendicitis often can be effectively managed by the protocols followed conventionally. If the shunt tube is seen during the appendectomy, this often can be managed by replacing the catheter away from the operative site. These patients need to be followed up closely to assess for any chronic abdominal infection that may present several weeks after the initial surgery.
- Patients with ruptured appendicitis most often need their shunt to be externalized with intravenous antibiotics, and once the peritoneal infection is settled, another site in the peritoneal cavity can be chosen to insert the shunt.

# Hernia, Hydrocele, and Shunts

- Persistence of the **peritoneovaginal** canal causes the CSF to track from the peritoneal cavity into the scrotum, thus causing **hydrocele**.
- If the communication is large, bowel loops can migrate into the scrotal sac, which results in **inguinal hernia**. The collection is usually lax and supple. Uncommonly, the distal end of the shunt tube can migrate into the sac.
- In most cases, these spontaneously reduce in size and do not need any surgical intervention. However, tense or growing collections need a repositioning of the catheter with correction of the defect.

## Idiopathic intracranial hypertension

- Features of **raised ICP without an underlying tumour or HCP**
- IIH can progress **rapidly to blindness**.
- a young **overweight female**, describes a headache typical of raised pressure, and visual deterioration.
- **Papilloedema + cranial nerve palsies**.
- Imaging is unremarkable, but **LP demonstrates a raised opening pressure  $>25$  mm Hg**.

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- **Impaired CSF resorption may reflect raised venous pressure**, either as a result of sinus thrombosis or secondary to raised intra-abdominal pressure in obese patients.
- Weight loss and cessation of certain medications including the oral contraceptive pill is often effective. This is combined with medical therapy using **acetazolamide** to reduce CSF production.
- **Lumboperitoneal or ventriculoperitoneal shunting**
- **Optic nerve sheath fenestration** by an ophthalmologist may also be employed.



Infections

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- **Intracranial infections and spinal infections.**
- The intracranial infections can occur in the **epidural** space (epidural abscess), in the **subdural** space (subdural empyema), in the **subarachnoid** space (meningitis), **intracerebrally** (cerebral abscess), or in the **ventricles** (ventriculitis).

## Meningitis

- Inflammation of the meninges of the brain and spinal cord, most commonly and most seriously due to bacterial infection.
- **Fever, headache, neck stiffness and photophobia.**
- **Complications = subpial encephalopathy, venous thrombosis, cerebral oedema and death.**
- Meningitis after head injury is common, affecting 25% of patients with base of skull fracture and CSF leak.

- The causative organism varies with the patient's age.
- **Neonatal** meningitis is caused by group B **streptococcus**, *Escherichia coli*, or *Listeria* spp. infection.
- **Late neonatal** meningitis can be caused by any of these organisms as well as by **staphylococci** or ***Pseudomonas aeruginosa***.
- In **children**, *Streptococcus pneumoniae* (**pneumococcus**) and *Neisseria meningitidis* (**meningococcus**) are the most common causative organisms.
- **Pneumococci and meningococci are the most common causative organisms in adults.**

- Treatment consists of prompt **CSF culture and immediate IV administration of antibiotics.**
- Altered mental status secondary to communicating hydrocephalus may necessitate placement of an external ventricular drain and eventual placement of a **ventriculoperitoneal shunt** once the CSF is sterilized.
- Recurrent episodes of bacterial meningitis prompt investigation into abnormal communication between the CNS and the exterior environment (**dermal sinus or CSF fistula**).

## **Brain abscess and empyema**

- Abscesses (usually bacterial) arise when the brain is exposed directly, for example as a **result of fracture or infection of an air sinus**, or at surgery.
- **Haematogenous spread**, typically in association with respiratory and dental infections, or endocarditis.
- **In 25% of cases, no underlying primary infection is found.**
- Low-grade fever, confusion, seizures and focal deficits

## Common causative organisms.

- ***Sinus/mastoid infection:*** aerobic and anaerobic streptococci; bacteroides; enterobacteria; staphylococci; Pseudomonas
- ***Haematogenous spread:*** bacteroides; streptococci
- ***Penetrating trauma:*** ***Staphylococcus aureus;*** clostridia; bacillus; enterobacteria
- ***Food contamination:*** Toxoplasma, pork tapeworm (producing neurocysticercosis)
- ***Immunocompromise*** (e.g. HIV/AIDS): protozoal (e.g. Toxoplasma), fungal (e.g. Cryptococcus), viral (e.g. JC virus producing multifocal leukoencephalopathy) and mycobacterial abscesses

- CT scan with contrast is the initial imaging modality of choice.
- Hypodense oedematous brain representing early cerebritis is visible in the first few days
- The classic appearances of a **smooth-walled, well-defined, ring-enhancing mass** develop as the abscess matures



**FIGURE 67-31** Brain abscess (arrow), with an area of frontal subdural empyema in the convexity (arrowheads).



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- The distinction between abscess and tumour can be difficult and has important management implications, since abscesses generally require urgent drainage.
- ***Diffusion-weighted MRI*** is a valuable tool in this context
- Intravenous antibiotic therapy should last **at least 6 weeks**
- Up to **50%** of patients with brain abscess will **develop seizures** at some stage, therefore prophylactic anticonvulsants should be considered.

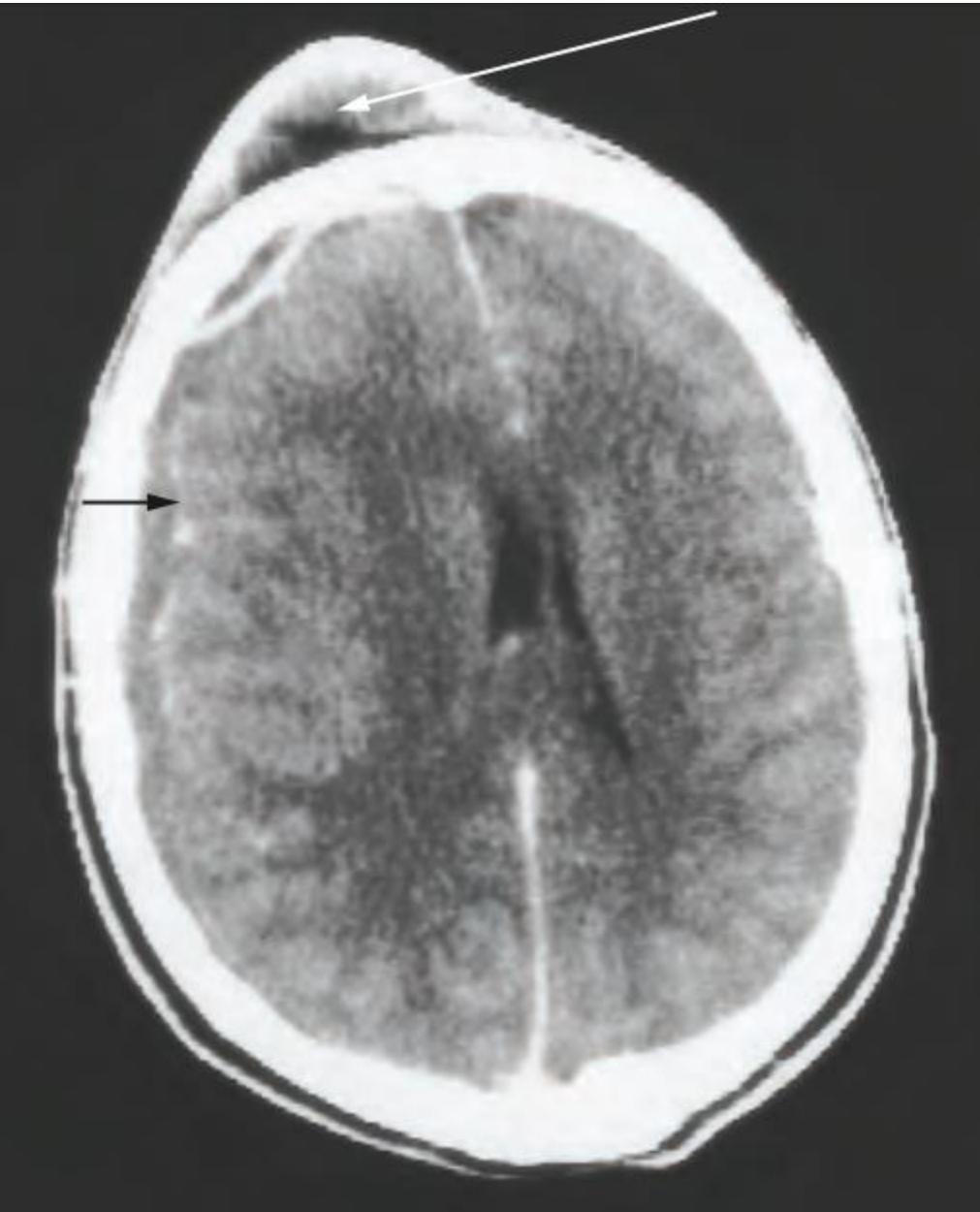
- Cranial CT scan usually shows the infection with osteomyelitis.
- The treatment is **surgical evacuation, débridement of the osteomyelitic bone, drainage of the adjacent infected sinuses, and prolonged antibiotic therapy.**

# Ventriculitis

- Infection of the ventricles is usually a result of spread of infection from either a ruptured cerebral abscess or other intracranial infection.
- **Shunt infections are the most common cause of ventriculitis.**
- Systemically administered antibiotics often do not penetrate significantly into the ventricle spaces.
- Treatment - Drainage of infected CSF with intraventricular instillation of vancomycin, gentamicin, or amikacin.

## Subdural empyema

- Infective collection in the subdural space
- Result of sinusitis, mastoiditis or meningitis, and can complicate trauma or surgery.
- *Subdural empyema associated with osteomyelitis of the frontal bone and associated scalp swelling (Pott's puffy tumour).*
- Mortality of 8–12%.
- CT appearances are of hypodense or isodense subdural collection, with contrast enhancement at the margins, and a degree of swelling and midline shift.



FOR IIT/JEE ENTRANCES

- Clinical features **fever, meningeal signs, headache, seizures, focal neurologic deficits, and altered mental status.**
- The most significant complication is **cortical venous thrombosis leading to cerebral infarction.** Followed by seizures and rapid clinical deterioration.

- Surgical drainage with antibiotic therapy.
- **Anticonvulsants** are indicated even in the absence of seizures as there is high risk of seizures.
- **Steroids** are often administered with antibiotic to cover life-threatening situations.
- **Craniotomy** with evacuation of the pus has better overall outcome than burr hole evacuation.

# Post-Traumatic Meningitis

- Meningeal infection after head injury is typically related to CSF fistula.
- Most post-traumatic fistulas stop spontaneously within days of injury.
- The incidence of meningitis increases if a leak persists for longer than 7 days. Clinically obvious leaks are manifested as CSF rhinorrhea or otorrhea.
- A persistent post-traumatic CSF fistula is addressed surgically to prevent the risks associated with recurrent bouts of meningitis.

# Spinal Infections

- Spinal infections can be grouped into those affecting the bone (vertebral osteomyelitis), disc space (discitis), and epidural space (spinal epidural abscess).

# Vertebral Osteomyelitis

- Osteomyelitis of the bone is generally seen in ***IV drug users, diabetic patients, hemodialysis patients***, and older adults.
- The causative organism is usually ***S. aureus***, and spread is hematogenous, although postoperative infections are also seen. These infections can and do affect the **integrity of the bone, resulting in collapse**. This in turn can result in pain and neurologic compromise.
- Treatment consists of appropriate long-term antibiotics, and maintenance of anatomic spinal alignment, with or without surgical intervention.



**FIGURE 67-32** MRI with gadolinium short T1 inversion recovery sequence revealing disc osteomyelitis in the L4-5 and L5-S1 intervertebral spaces suggestive of an infectious process.

# Discitis

- Infection of the disc (discitis) often occurs concomitantly with osteomyelitis and is seen in the same population of patients.
- Fever, back pain, and an elevated sedimentation rate or C-reactive protein level are often seen. The white blood cell count may or may not be elevated. It may occur spontaneously or postoperatively.
- Long-term antibiotic therapy is usually indicated.

# Spinal Epidural Abscess

- This usually occurs in the setting of an infectious process elsewhere in the body. Spread occurs hematogenously or by direct extension.
- Clinical features - localized back pain and possible radiculopathy.
- Spinal cord compromise can follow rapidly, with paraplegia or quadriplegia. Diagnosis is made with contrast-enhanced MRI.
- When spinal cord compression is evident, surgery is usually performed for decompression and diagnosis.

# Acquired Immunodeficiency Syndrome

- The most common CNS opportunistic infection in patients with AIDS is toxoplasmosis caused by **Toxoplasma gondii**.
- The lesions usually present with ***ring enhancement on contrast-enhanced*** imaging studies and are usually in the **basal ganglia**.
- Primary CNS **lymphoma** occurs in approximately 10% of AIDS patients and presents as an irregularly enhancing mass (target lesion).
- **Progressive multifocal leukoencephalopathy** presents with hypodense, nonenhancing white matter lesions.
- **Fungal abscess and viral encephalopathy**



# Hemorrhages

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## Subarachnoid haemorrhage

- From a **ruptured aneurysm** (~80% of SAH) or an arteriovenous malformation (AVM).
- Most ruptured aneurysms are located in the circle of Willis, at branch points in the arterial tree associated with turbulent blood flow
- **Pseudoaneurysms** may also develop after trauma or after surgery.

- A distinct subgroup of SAH patients suffer bleeds confined to the basal cisterns anterior to the midbrain and pons, without an underlying lesion evident on angiogram. This is termed **perimesencephalic SAH**, is believed to **represent venous bleeding** and has an excellent prognosis.



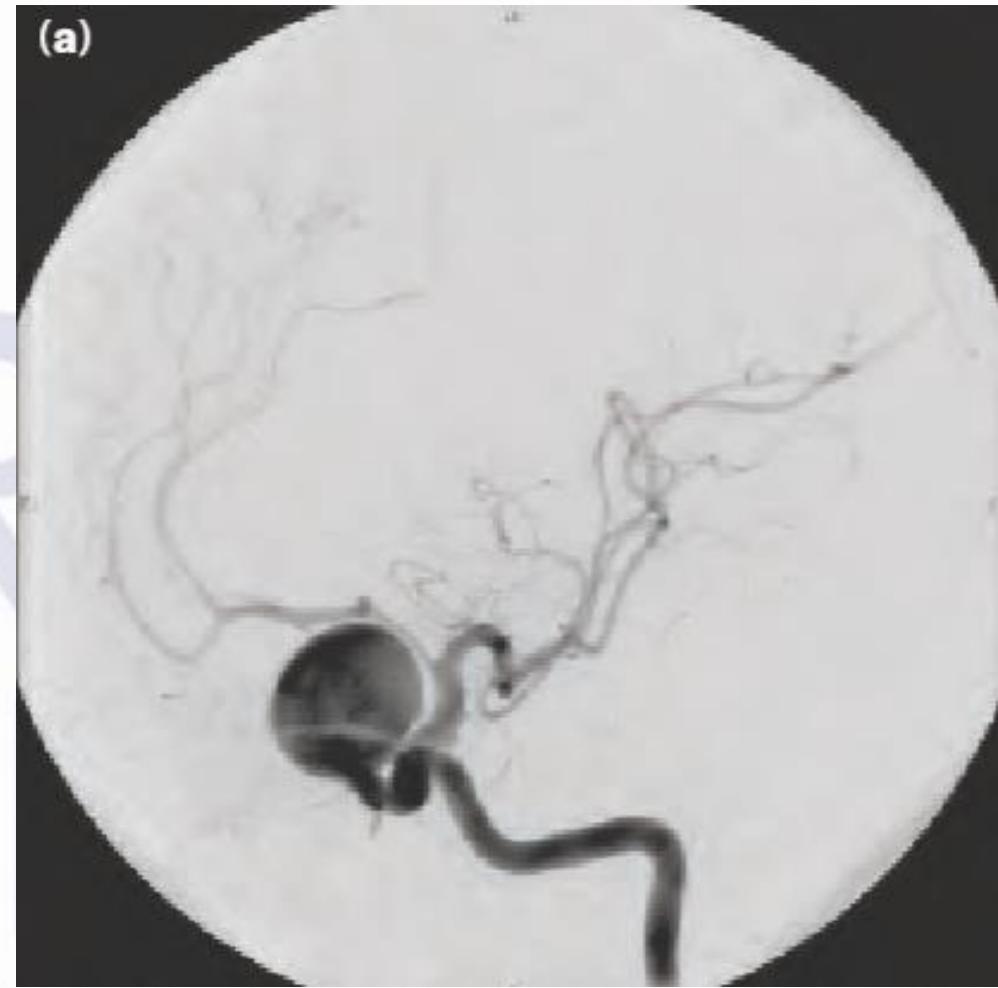
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# Aneurysms

- Aneurysms are an excessive localized enlargement of a vessel due to a weakening and subsequent defect in the wall of the artery.
- There is an adult prevalence of 2% (this varies according to the study) with an annual incidence of aneurysmal subarachnoid hemorrhage of 6 to 8/100,000 with peak age at 50 years.
- Modifiable risk factors for subarachnoid hemorrhage include **hypertension, smoking, and excessive alcohol**.
- Aneurysms may be further divided into saccular, fusiform, dissecting, infectious, and traumatic aneurysms.

- Aneurysms may also develop as a result of **infective infiltration of arterial walls** in the context of **bacteraemia (mycotic aneurysm)**, often in the setting of **intravenous drug use or infective endocarditis**.



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## Genetic disorders

- adult polycystic kidney disease,
- fibromuscular dysplasia,
- neurofibromatosis type 1,
- Ehlers–Danlos and
- Marfan's syndromes

- **Thunderclap' headache**, which is both sudden and severe and is outside the patient's normal experience.
- May be associated with **seizure (10%)**, **unresponsiveness (50%)** and **vomiting (70%)**.
- Sometimes it is difficult to establish whether SAH has caused a fall, or whether a fall with head injury is responsible for the SAH.
- At high risk of succumbing to early complications, especially a rebleed.

- A **painful third nerve palsy** is typically the result of compression from a posterior communicating artery aneurysm.
- Meningitic features of neck stiffness and photophobia often develop over hours.
- Intraocular haemorrhages, classically subhyaloid, on fundoscopy.
- The combination of SAH and vitreous haemorrhage is known as Terson's Syndrome and occurs in 15–20% of patients.

- A **rebleed risk of 4% in the first 24 hours**, then 1.5% per day thereafter is quoted for aneurysms
- **Delayed ischaemic neurological deficit (DNID)**, which commonly develops **3 to 10 days after aneurysmal haemorrhage** and can progress rapidly to infarction.
- The process is attributed to **cerebral vasospasm** in response to, and correlating with, the blood load.

**TABLE 67-4 Hunt and Hess Clinical Grading Scale**

<b>DESCRIPTION</b>	<b>GRADE</b>	<b>GOOD OUTCOME</b>
Asymptomatic or minimal headache and slight nuchal rigidity	1	≈70
Moderate to severe headache, nuchal rigidity, ± cranial nerve palsy only	2	≈70
Drowsy, confusion, or mild focal deficit	3	≈15
Stupor, moderate to severe hemiparesis, possibly early decerebrate rigidity	4	≈15
Deep coma, decerebrate rigidity, moribund appearance	5	≈0

**TABLE 67-5 World Federation of Neurological Surgeons Clinical Grading Scale**

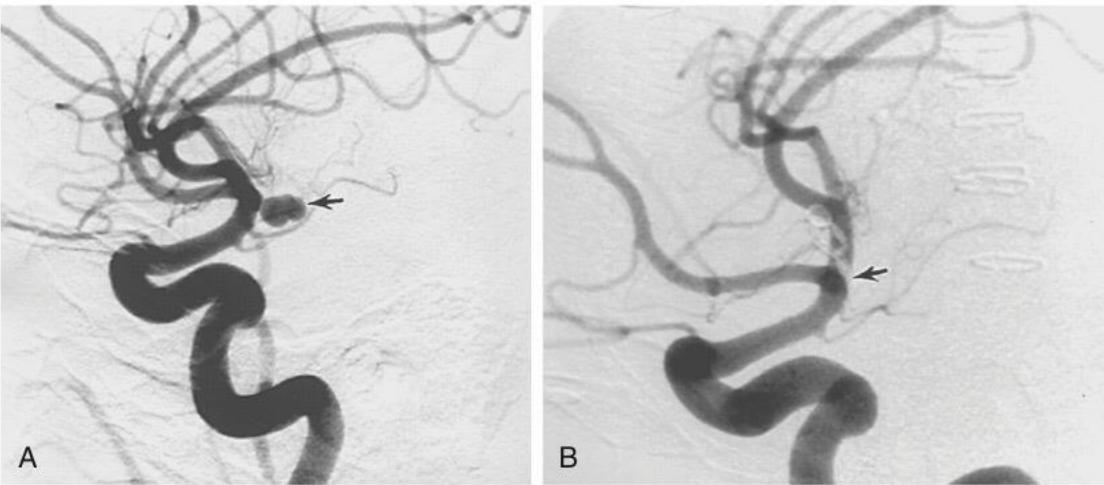
<b>GRADE</b>	<b>GCS SCORE</b>	<b>MOTOR DEFICIT</b>
I	15	No
II	13-14	No
III	13-14	Yes
IV	7-12	Yes or no
V	3-6	Yes or no

GCS, Glasgow Coma Scale.

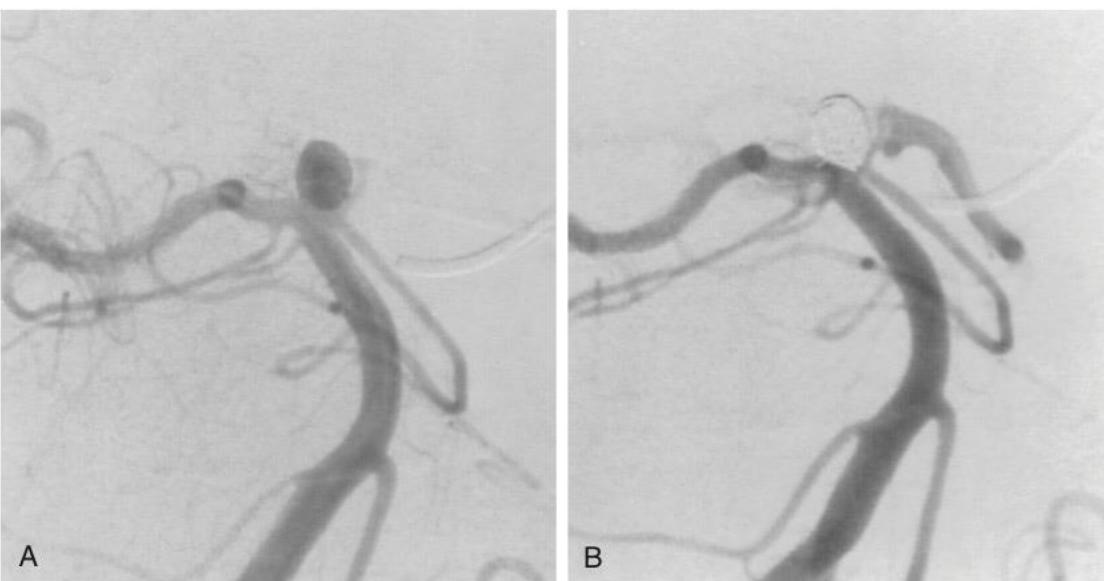
**TABLE 67-6 Fisher Grading for Appearance of SAH on Computed Tomography**

<b>GRADE</b>	<b>CT FINDINGS</b>
I	No hemorrhage evident
II	Diffuse SAH with vertical layers < 1 mm thick
III	Localized clots and/or vertical layers of SAH > 1 mm thick
IV	Diffuse or no SAH, but with intracerebral or intraventricular hemorrhage

SAH, subarachnoid hemorrhage.



**FIGURE 67-7** **A**, Subtraction carotid angiogram shows a 4 x 6-mm berry aneurysm (arrow) originating from the distal internal carotid artery. **B**, Postoperative carotid angiogram shows clip placement (arrow), with total obliteration of the aneurysm.



**FIGURE 67-8** **A**, Subtraction vertebral angiogram shows a basilar tip aneurysm. **B**, Subtracted vertebral angiogram after the placement of coils demonstrates excellent obliteration of the aneurysm and preservation of adjacent vessels.

- The major complications of subarachnoid hemorrhage include rebleeding, hydrocephalus (which is observed in around 15% to 20% of cases), cardiac events in around 50% of patients, vasospasm, hyponatremia, and seizures.
- Vasospasm is a narrowing of the cerebral arteries thought to be caused by smooth muscle dysfunction related to blood breakdown products in the spinal fluid. Leads on to significant ischemia of the brain and, as such, be a significant cause of morbidity.

- Treatment of symptomatic vasospasm is typically multimodality and includes the use of hypervolemia and induced hypertension, endovascular procedures, and a variety of pharmacologic agents

## BOX 67-2 Treatment of Vasospasm

### **Prevention of Arterial Narrowing**

Subarachnoid blood removal

Prevention of dehydration and hypotension

Calcium channel blockers (nimodipine)

### **Reversal of Arterial Narrowing**

Intra-arterial calcium channel blockers

Transluminal balloon angioplasty

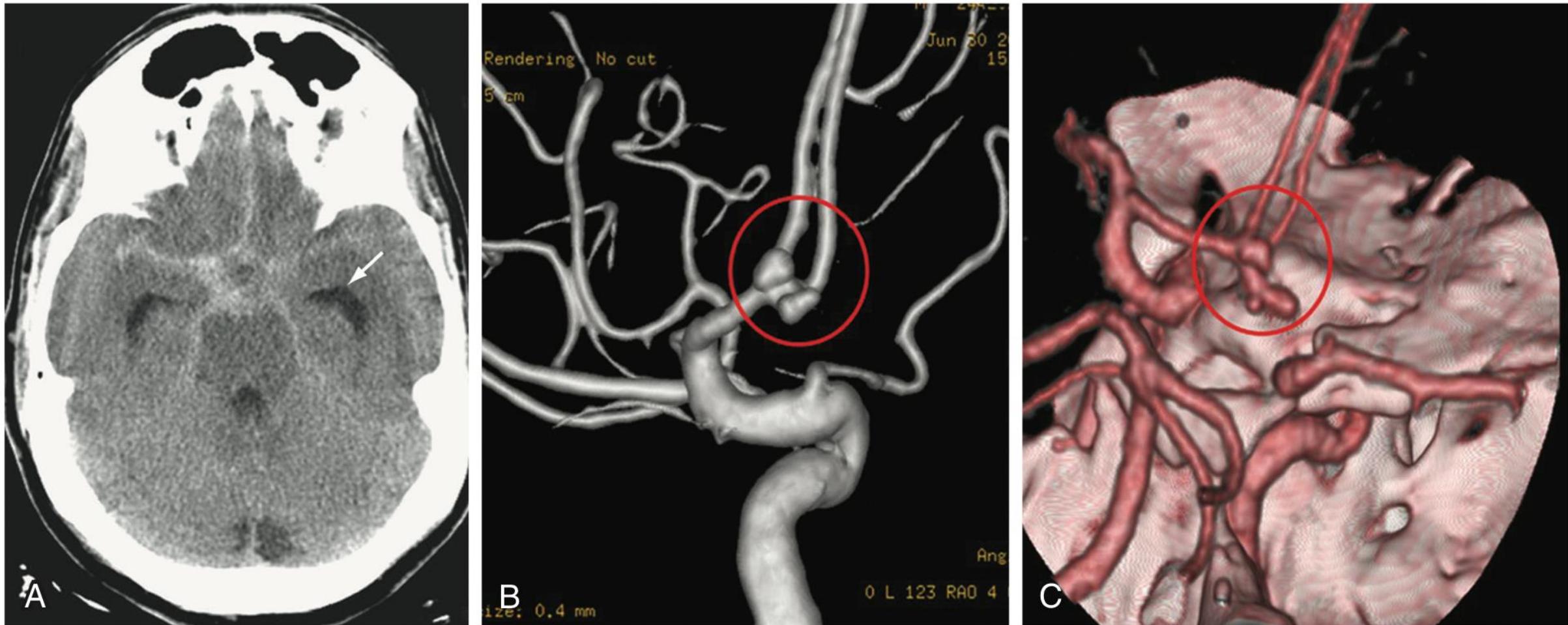
### **Prevention and Reversal of Ischemic Neurologic Deficit**

Hypertension, hypervolemia, hemodilution

# Saccular Aneurysms

- Also referred to as **berry** aneurysms, are usually **saccular** in form and come off the vessel wall or at a bifurcation.
- The classic presentation of **subarachnoid hemorrhage** due to a cerebral aneurysm is that of sudden onset of a **headache**, described as the “worst headache of my life.”
- Workup typically includes a CT scan, demonstrating a typical distribution of blood. Between 10% and 15% of subarachnoid hemorrhages from saccular aneurysms are fatal before the hospital is even reached.

- Treatment of saccular aneurysms consists of coiling, coiling through a stent, or surgical clipping with subsequent intensive critical care unit management of potential vasospasm and comorbidities.



**FIGURE 67-6** **A**, CT scan of brain showing subarachnoid blood in the basal cisterns. Dilated temporal horns (arrow) indicate the presence of hydrocephalus. **B**, Cerebral angiogram shows two aneurysms located at the junction of the A1 and A2 segments of the anterior cerebral artery (circle). **C**, CT angiography with three-dimensional reconstruction showing the relationship of the aneurysms (circle) with the skull base.

# Mycotic Aneurysms

- Associated with a **systemic infection** capable of showering small particles of bacteria-infected material into the cerebral vascular bed.
- **Subacute bacterial endocarditis** and pulmonary infections
- A distinguishing feature of these aneurysms is that they are generally found more **distal** in the cerebral vascular bed, as opposed to berry aneurysms, which are usually found on larger vessels near the circle of Willis.

- Maximal **antibiotic** treatment is essential at the outset.
- Attempts to dissect and to define a neck are frustrated by a lack of developed fibrous tissues, and intraoperative hemorrhage is then common.
- Typically, the diseased arterial segment must be occluded and resected when it is operated on in this early stage.
- If the mycotic aneurysms are discovered or treated at some later stage, a fibrous wall to the aneurysm may have had time to develop, and clipping can then be a possibility.

- **Spontaneous intracerebral haemorrhage** accounts for **10–15%** of strokes and has a **mortality of 40% at 1 year**.
- The majority occur in the **context of hypertension or amyloid angiopathy**, or as a complication of ischaemic stroke.
- Coagulation disorders, especially patients being treated with warfarin, are a major risk factor.

- They can occur anywhere in the cerebral circulation or brainstem but are classically described in association with **small degenerative aneurysms (microaneurysms; also known as Charcot- Bouchard aneurysms)** at the junctions of the perforating vessels and larger vessels at the skull base
- They are typically on the middle cerebral artery junctions with the small perforating **lenticulostriate** vessels, leading to hemorrhage into the **putamen**.
- Clinical presentation **stroke** pattern of sudden-onset neurologic signs and symptoms that depend on the area of brain affected.

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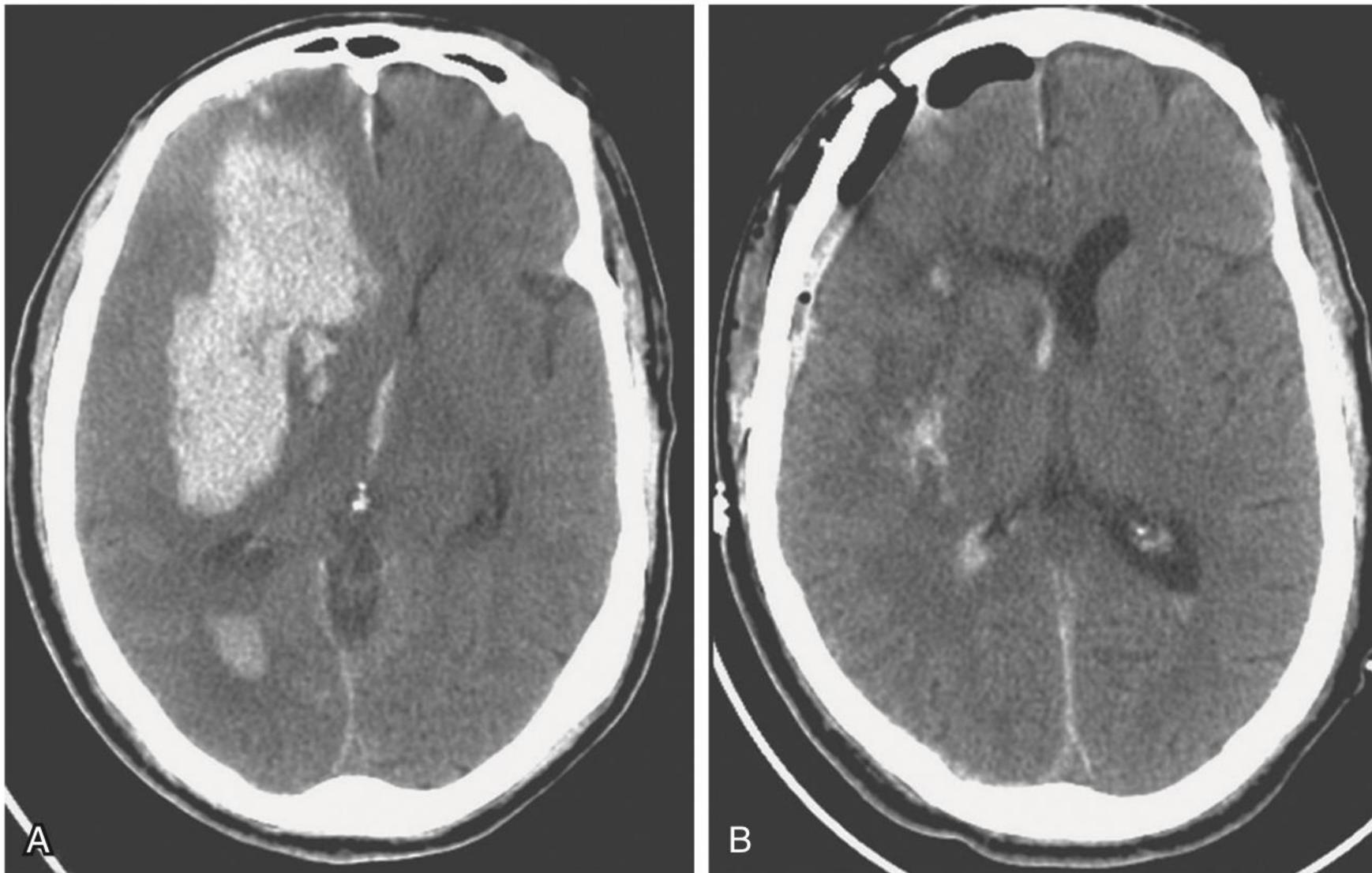
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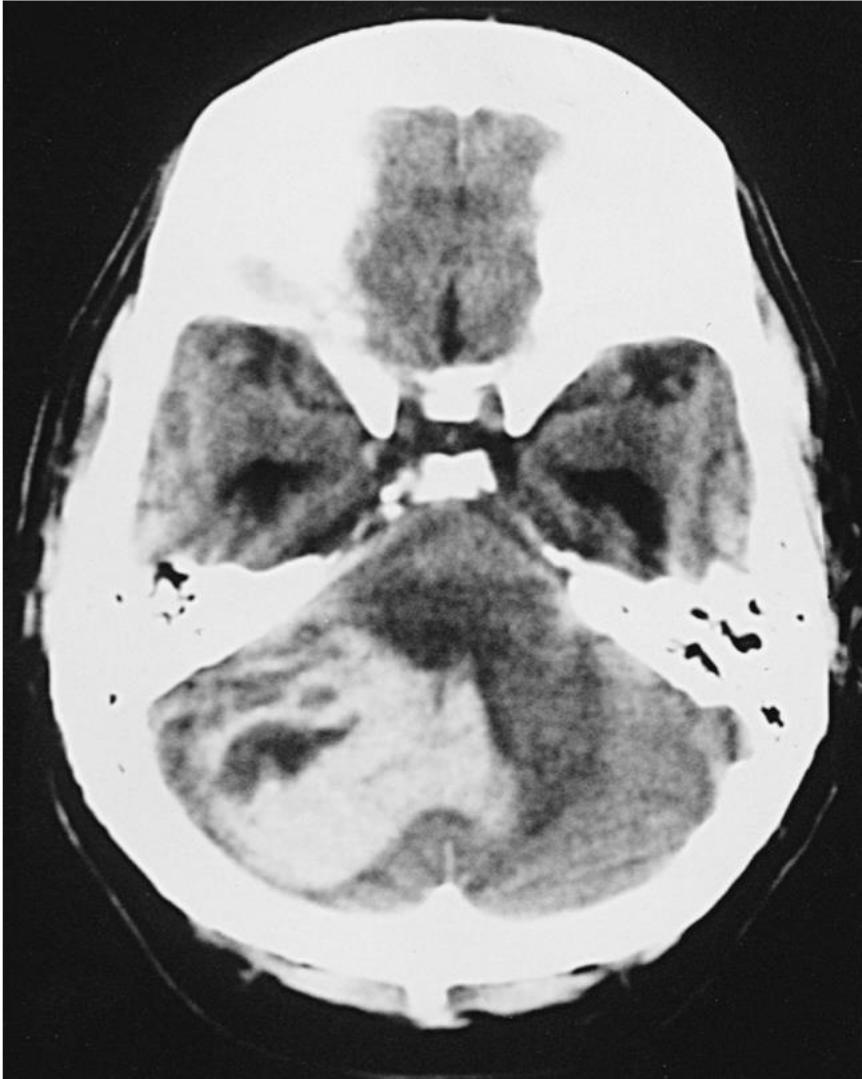


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**FIGURE 67-9** Nonenhanced CT scan of the head. **A**, Spontaneous hypertensive intracerebral hematoma in the right basal ganglia, with extension to the frontal and temporal lobes. **B**, Immediate postoperative CT scan shows near-total removal of the intracerebral hematoma.

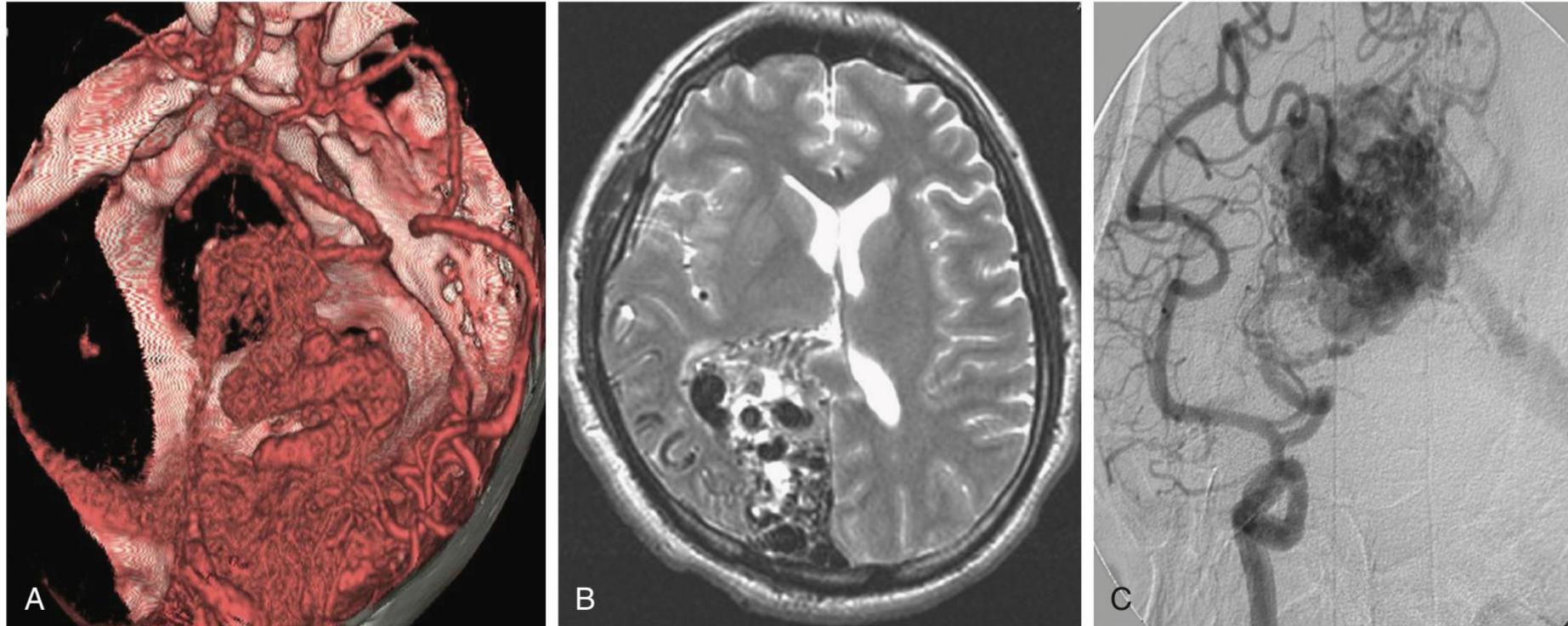


**FIGURE 67-10** Nonenhanced CT scan of the brain shows a large, hypertensive, intracerebellar hematoma with obstruction of the fourth ventricle and enlargement of the temporal horns, indicating obstructive hydrocephalus.

# Arteriovenous Malformations

- An AVM is an abnormal collection of blood vessels wherein arterial blood flows directly into draining veins without the normal interposed capillary beds.
- They present with **hemorrhage, ischemia** of the brain parenchyma around the lesion due to a vascular “steal” phenomenon, or seizures

# Arteriovenous Malformations



**FIGURE 67-4** CT angiography with three-dimensional reconstruction **(A)**, MRI **(B)**, and conventional angiogram **(C)** of a large AVM with supply from the middle and posterior cerebral arteries and ill-defined nidus. Complex deep and superficial venous drainage is present.

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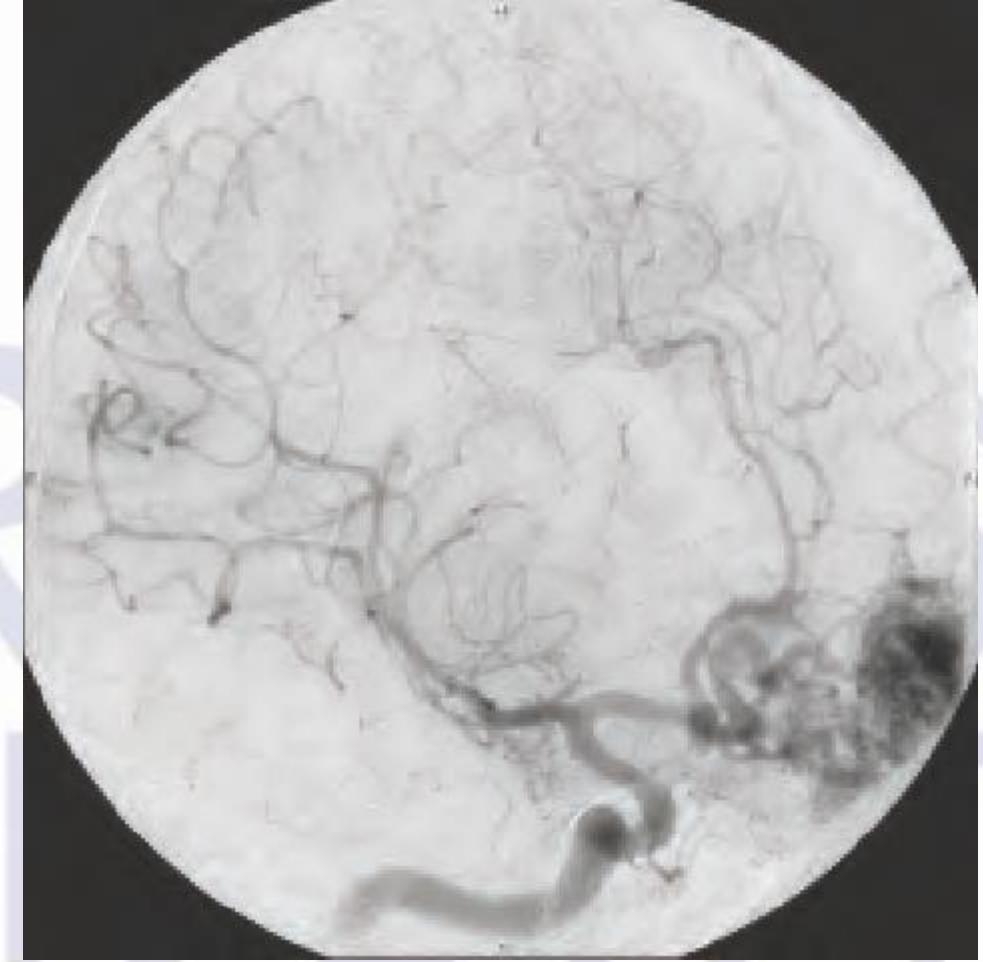
**TABLE 67-2 Spetzler-Martin Grading System**

<b>FEATURE</b>	<b>POINTS</b>
Nidus size (cm)	
Small (<3)	1
Medium (3-6)	2
Large (>6)	3
Eloquence of adjacent brain	
Noneloquent	0
Eloquent (sensorimotor, language, visual, thalamus, hypothalamus, internal capsule, brainstem, cerebellar peduncles, deep cerebellar nuclei)	1
Pattern of venous drainage	
Superficially only	0
Deep	1

# Arteriovenous Malformations

- Clinical features - *headaches, neurologic deficit, seizures*, or varying combinations of the three.
- **Catheter angiography** is then performed to define the vascular anatomy of the lesion and is used for treatment planning.

- **4% risk of rebleed per annum.**
- The risk is particularly high in the first **6 weeks** and where the bleed is from an aneurysm related to the AVM.
- Surgery (not always), Radiosurgery and endovascular embolisation with tissue glue are treatment options
- SRS is usually reserved for compact lesions less than 2.5 cm in diameter. It can take up to 3 years for irradiated AVMs to shut down after SRS.

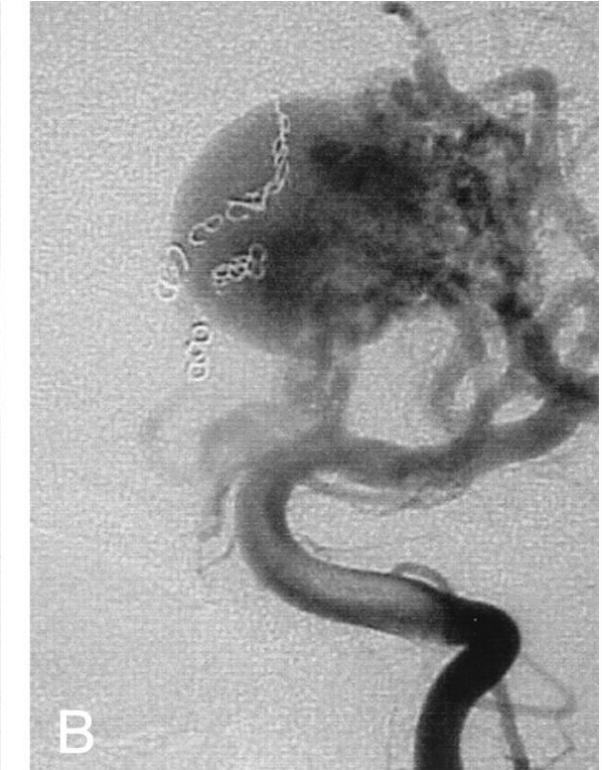
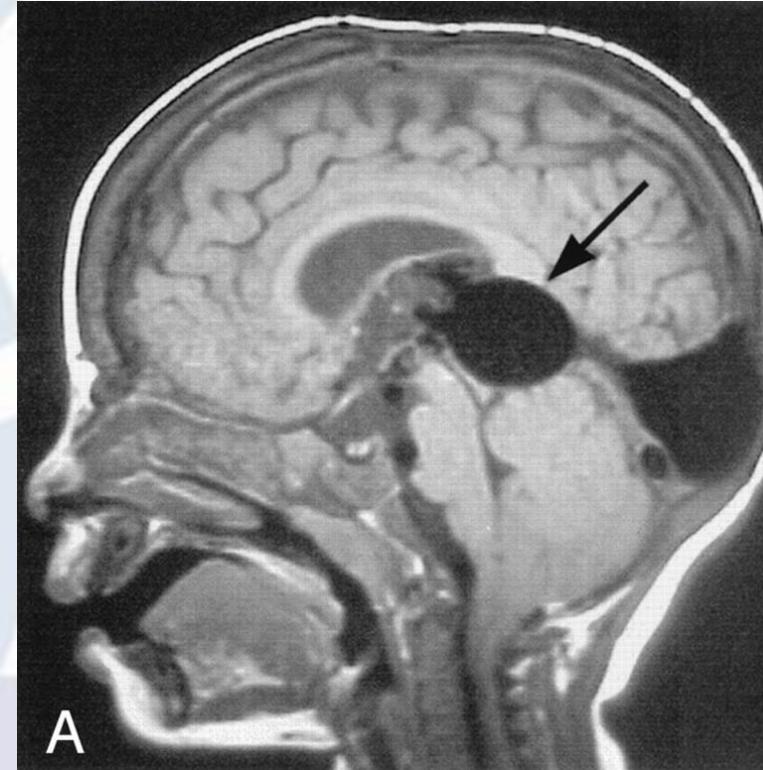


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- Vein of Galen malformations

AVMs feeding into an embryological venous remnant dorsal to the **brainstem** presenting in childhood.

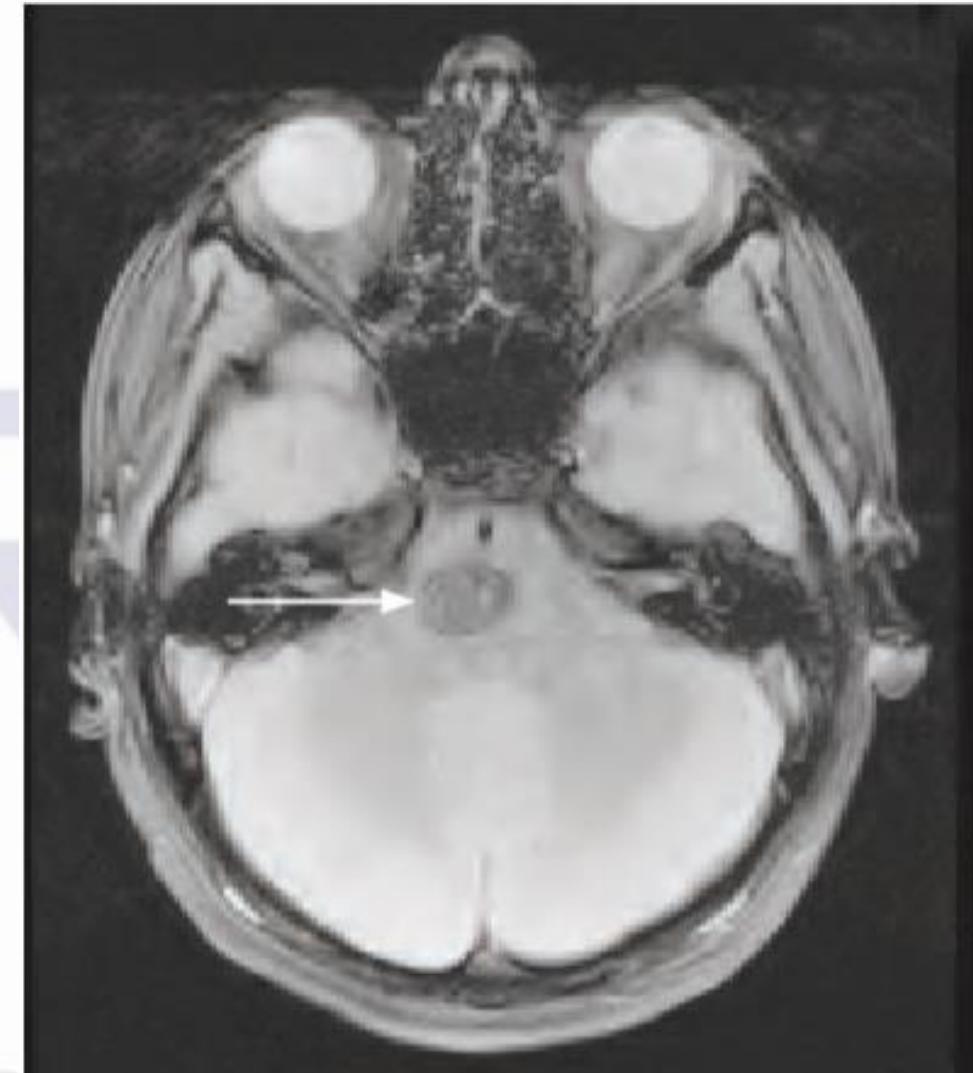
- High-flow malformations may cause cardiac failure.
- They may be treated by embolisation.



# Cavernous Malformations

- A well-circumscribed, benign vascular lesion consisting of irregular thin-walled sinusoidal vascular channels located within the brain but lacking intervening neural parenchyma, large feeding arteries, or large draining veins.
- Because these are low-pressure, low-flow lesions, hemorrhage is typically not catastrophic unless it is in a highly eloquent area of the brain.
- Patients usually present with hemorrhage or headaches with or without a history of new-onset seizures.

- **Cavernomas are venous anomalies**, demonstrated on MRI but ***not with angiography***, which may require operation if they cause progressive deficits, intractable epilepsy or recurrent bleeding.



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# Capillary Telangiectasia

- Composed of vascular channels with extremely thin walls similar to those of dilated capillaries. They are often clinically silent and generally do not appear on imaging studies.
- They are not evident on conventional catheter angiography unless they are large, and then only in the capillary venous phase.

# Developmental Venous Anomaly: Venous Angioma

- Abnormally configured venous drainage system converging on a single, enlarged venous outflow channel.
- The typical appearance is that of a hydra, with radially converging veins.
- The venous anomaly represents the only venous drainage available to that area of brain, and therefore removal of the venous anomaly is not recommended. Doing so could lead to a venous infarction with swelling and hemorrhage, the consequences of which are particularly dangerous in the posterior fossa.

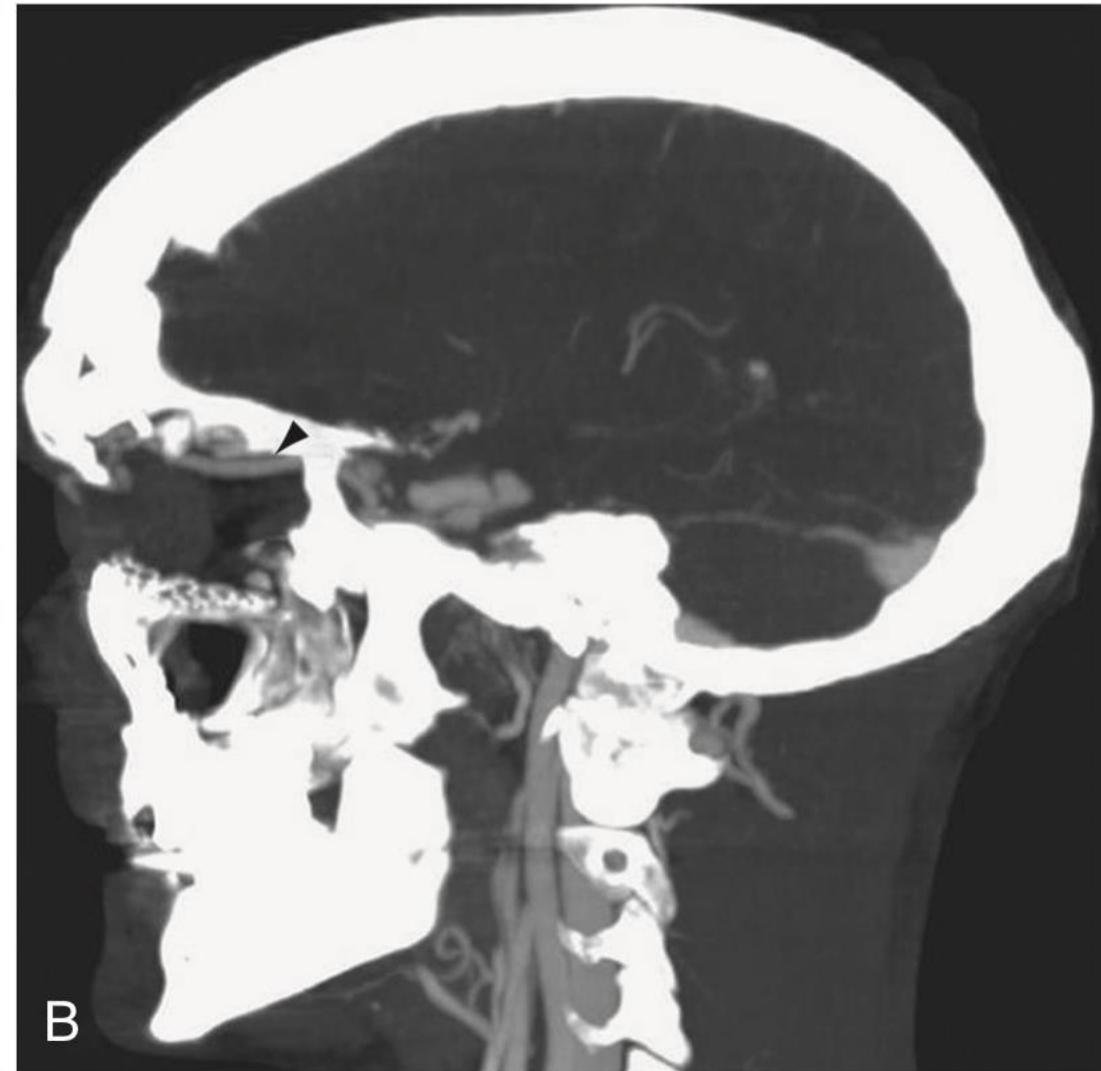
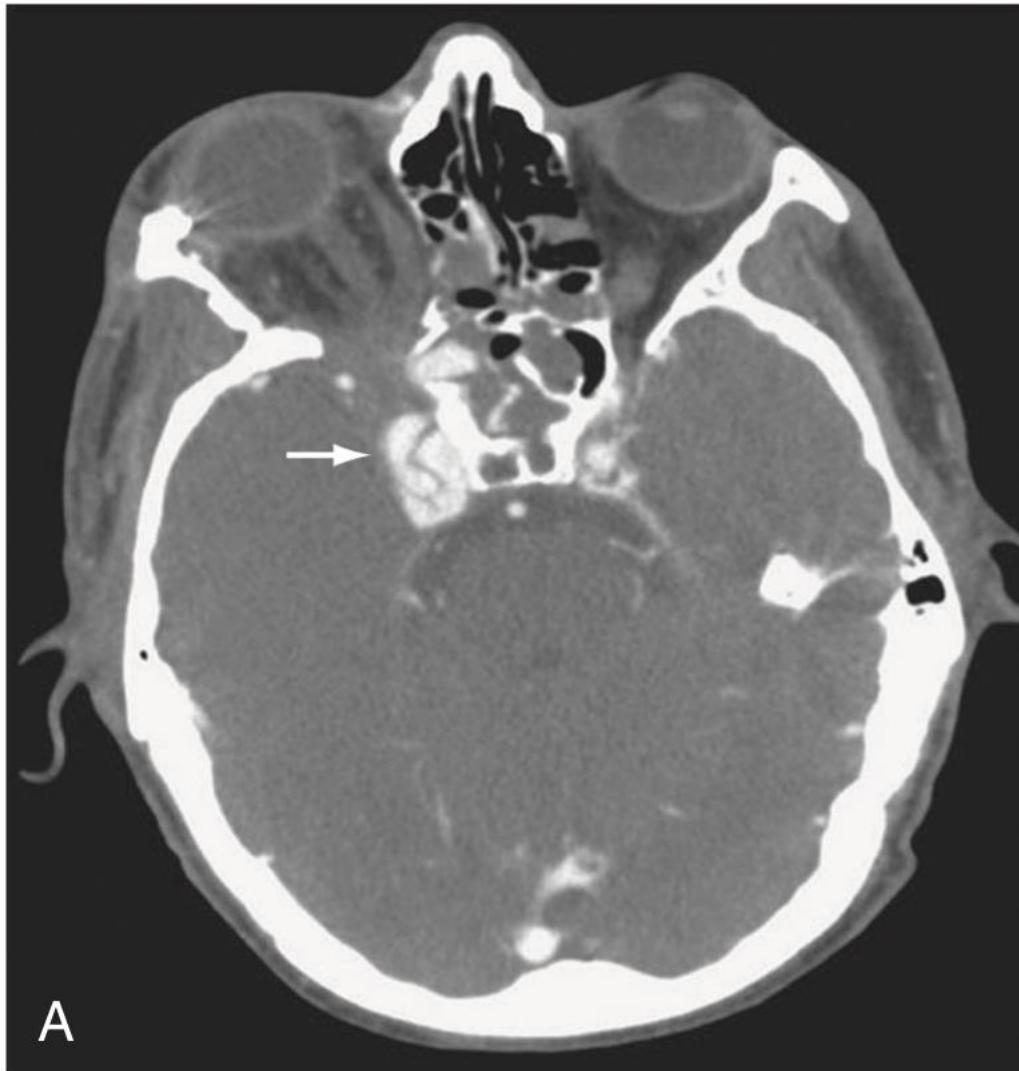
- **Dural arteriovenous fistulae (DAVs)** are shunts between dural arteries and veins or sinuses.
- They are proposed to arise as a result of vessel remodelling in response to **dural sinus thrombosis and subsequent recanalisation**.
- They may present with subarachnoid, intracerebral or subdural bleeding, or with headache and pulsatile tinnitus.
- A carotid cavernous fistula is a spontaneous or traumatic DAVF between the **internal carotid artery and surrounding cavernous sinus**, typically producing **eye pain, ocular muscle palsies and exophthalmos**.

# Traumatic Fistula

- The internal carotid artery passes through the cavernous sinus, which communicates with the superior ophthalmic vein, petrosal sinus, and sphenoparietal sinus.
- The vertebral artery passes through a venous plexus at the occipital-C1 epidural space, which communicates with the jugular vein, epidural venous plexus, and paraspinal venous plexus.
- **Trauma leading to a tear in the carotid or vertebral artery at its tether point passing through the skull base can lead to fistula with the surrounding venous plexus.**

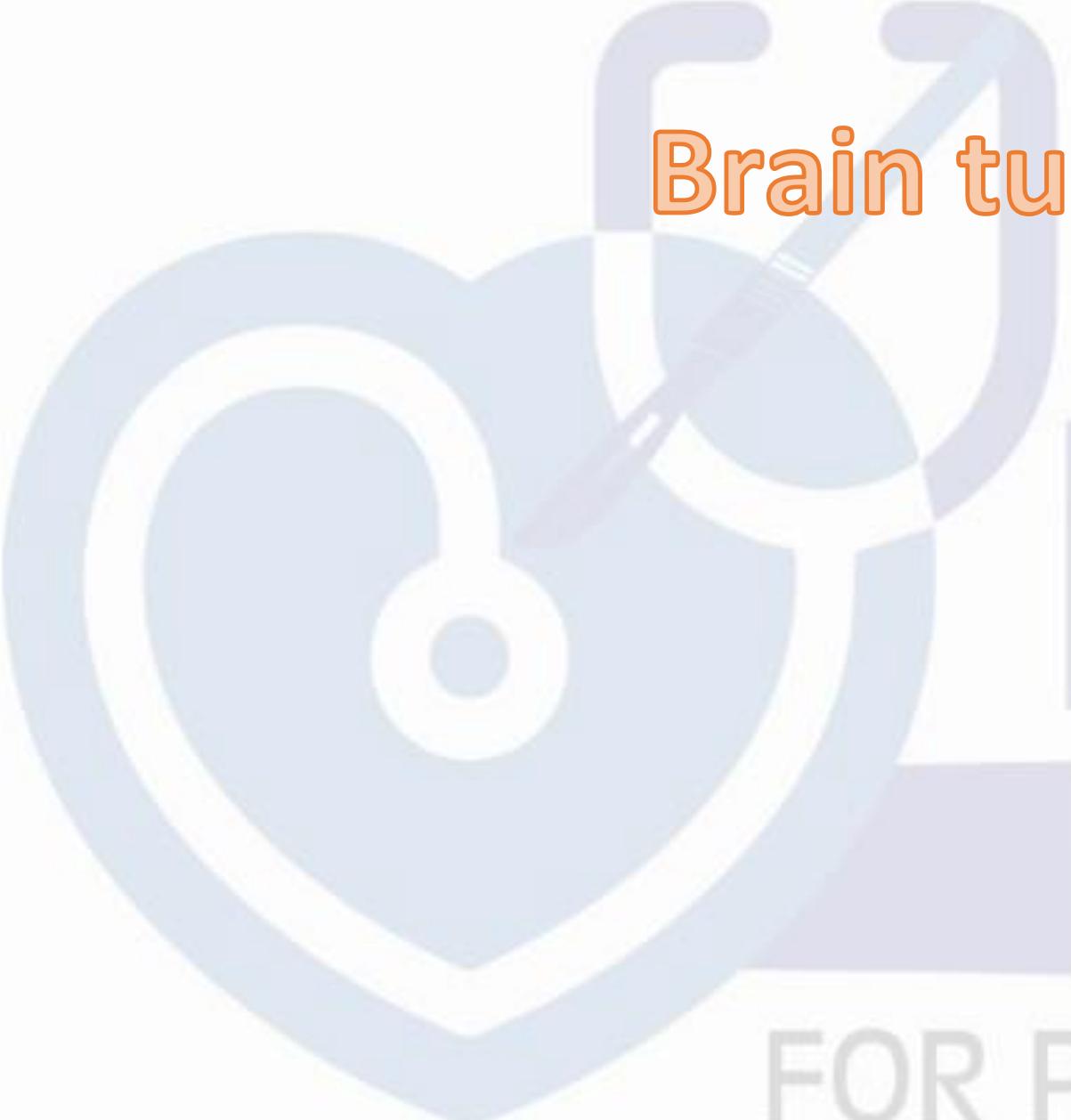
- The consequences may vary in severity and suddenness but typically include **periorbital swelling**, with **proptosis and scleral edema** in the case of the carotid-cavernous fistula (CCF) and prominent **pulsatile bruit** in the case of the vertebral-jugular fistula.
- On radiologic examination, **dilation of the superior ophthalmic vein** is characteristic. These lesions are usually treated by **endovascular** techniques. A catheter is advanced through the tear in the artery into the venous side of the fistula. The high flow and large fistulous channel facilitate this process.
- **Embolic material, a coil, or a detachable balloon** is then used to occlude the venous side of the fistula.





**FIGURE 67-5** Right internal carotid-cavernous sinus fistula (**A**, arrow) with dilation of the superior ophthalmic drain (**B**, arrowhead), a typical imaging finding of this pathologic process.

- **Moya moyo disease** - the progressive obliteration of one or both internal carotid arteries (*?autoimmune process*)
- The development of external carotid circulation collaterals produces the angiographic 'puff of smoke' appearance responsible for this Japanese-derived name.
- It presents in youth or early middle age with ischaemia or haemorrhage.
- Ischaemia may be addressed by a variety of bypass techniques, for example by anastomosing the superficial temporal artery (arising from the external carotid) to the middle cerebral artery.



Brain tumors

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## Brain tumours

- Most brain tumours will present with one or more feature related to the following triad:

Raised ICP

Seizures

Focal deficit

- Primary brain tumours represent **1.5% of all cancers**, with an incidence of 19 per 100 000 person years.
- Common adult primary brain tumours include gliomas, **meningiomas (15–20% of total)**, **pituitary adenomas (10–15% of total)** and vestibular schwannomas.

**TABLE 43.3** Chromosomal abnormalities associated with brain tumours.

Syndrome	Gene defect	Tumour
Neurofibromatosis type 1	<i>Neurofibromin</i> (Chr 17)	Astrocytomas; neurofibromas
Neurofibromatosis type 2	<i>Schwannomin</i> (Chr 22)	Acoustic neuromas (bilateral); meningiomas
Cowden's disease	<i>PTEN</i> (Chr 10)	Astrocytomas
Hereditary non-polyposis colorectal cancer	Multiple	Astrocytomas
Li-Fraumeni syndrome	<i>p53</i> (Chr 17)	Astrocytomas

**TABLE 43.4** Patterns of deficit generally associated with certain tumours.

Tumour location	Expected deficit
Pituitary (e.g. pituitary adenoma)	Bitemporal hemianopia; gaze palsies
Cerebellopontine angle (e.g. vestibular schwannoma)	Hearing loss; balance disturbance; tinnitus
Anterior skull base (e.g. olfactory groove meningioma)	Anosmia; ipsilateral optic atrophy; contralateral papilloedema (Foster-Kennedy syndrome)
Occipital (e.g. glioma, metastasis)	Homonymous hemianopia with central sparing
Parietal (dominant hemisphere)	Acalculia; agraphia; left-right disorientation; finger agnosia (Gerstmann syndrome)
Parietal (e.g. glioma)	Sensory inattention; dressing apraxia; astereognosis
Temporal (e.g. glioma)	Memory disturbance; contralateral superior quadrantanopia; dysphasia (dominant hemisphere)
Frontal (e.g. glioma)	Personality change; gait disturbance; urinary incontinence
Brainstem (e.g. brainstem glioma)	Multiple cranial nerve deficits; long tract signs; nystagmus
Posterior fossa (e.g. medulloblastoma)	Ataxia; hydrocephalus

**TABLE 43.5** Tissue of origin for brain metastases (approximate).

Origin	Percentage
Lung	40
Breast	15
Melanoma	10
Renal/genitourinary	10
Other/unknown	25

# Clinical Presentation

- The clinical manifestations of various brain tumors can be divided into those caused by focal compression and irritation by the tumor itself and those attributed to secondary consequences, namely, increased ICP, peritumoral edema, and hydrocephalus.

# Clinical Presentation

- Headache classically described as being worse in the morning, probably because of hypoventilation during sleep, with consequent elevation of the PCO<sub>2</sub> and cerebrovascular dilation.
- nausea and vomiting in 40% of patients and may be temporarily relieved by vomiting as a result of hyperventilation.
- Seizures may be the first symptom of a brain tumor. Patients older than 20 years presenting with a new-onset seizure are aggressively investigated for a brain tumor.

- **Infratentorial** lesions - headache, nausea and vomiting, gait disturbance and ataxia, vertigo, cranial nerve deficits leading to diplopia (abducens nerve), facial numbness and pain (trigeminal nerve), unilateral hearing deficit and tinnitus (vestibulocochlear nerve), facial weakness (facial nerve), dysphagia (glossopharyngeal and vagus nerves), and CSF obstruction causing hydrocephalus and papilledema.

- Supratentorial lesions may be manifested with different symptoms, depending on the location.
- Frontal lobe lesions are manifested as personality changes, dementia, hemiparesis, or dysphasia.
- Temporal lobe lesions may be manifested with memory changes, auditory or olfactory hallucinations, or contralateral quadrantanopia.
- Patients with parietal lobe lesions may develop contralateral motor or sensory impairment, apraxia, and homonymous hemianopia, whereas those with occipital lobe lesions may show contralateral visual field deficits and alexia.

# Imaging Studies

- CT scan of the brain provides a rapid means of evaluating changes in brain density, such as calcifications, hyperacute hemorrhages (<24 hours old), and skull lesions.
- MRI of the brain, however, is the “gold standard” modality for diagnosis, presurgical planning, and post-therapeutic monitoring of brain tumors. Gadolinium contrast enhancement with MRI is more sensitive in demonstrating defects in the blood-brain barrier and localizing small metastases (up to 5 mm).

- **Diffusion-weighted imaging** can help distinguish between gliomas and abscesses, and perfusion-weighted imaging can predict response to radiotherapy in low-grade gliomas.
- **Functional MRI** can be used in planning of surgery for tumors in eloquent areas of the brain to enable radical resection with less morbidity.
- **Diffusion tensor imaging** can demonstrate the effect of a tumor on white matter tracts.
- **Magnetic resonance angiography** is used more routinely as a noninvasive modality to evaluate the vascularity of a tumor or anatomic relationship of a tumor to normal cerebral vasculature.

## *Regarding phenytoin*

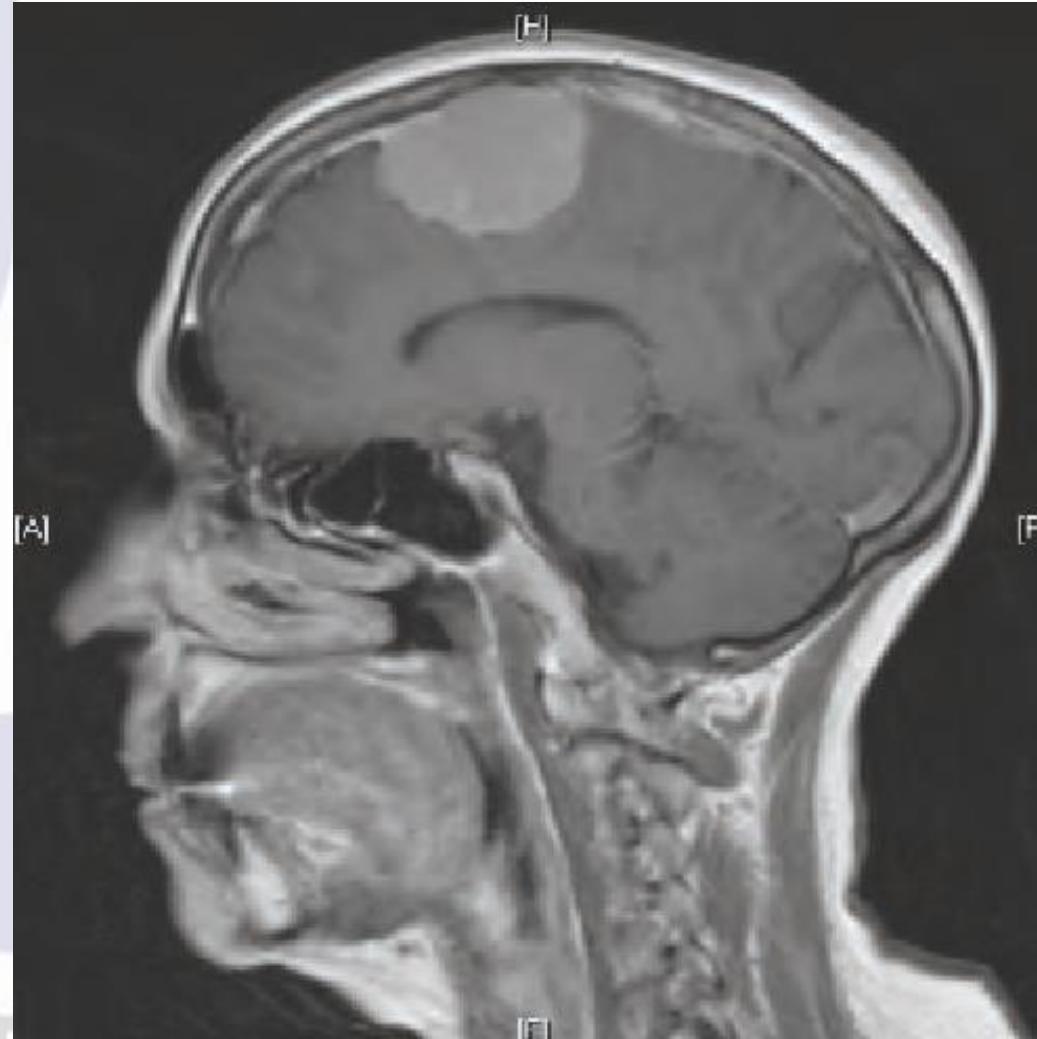
- Therapeutic levels of phenytoin can be achieved rapidly with intravenous loading, but its enzyme-inducing effect can complicate the administration of chemotherapy. **Routine prophylaxis in patients with tumours who have no history of seizures is not recommended**, although a short course at the time of craniotomy for tumour excision may be warranted.

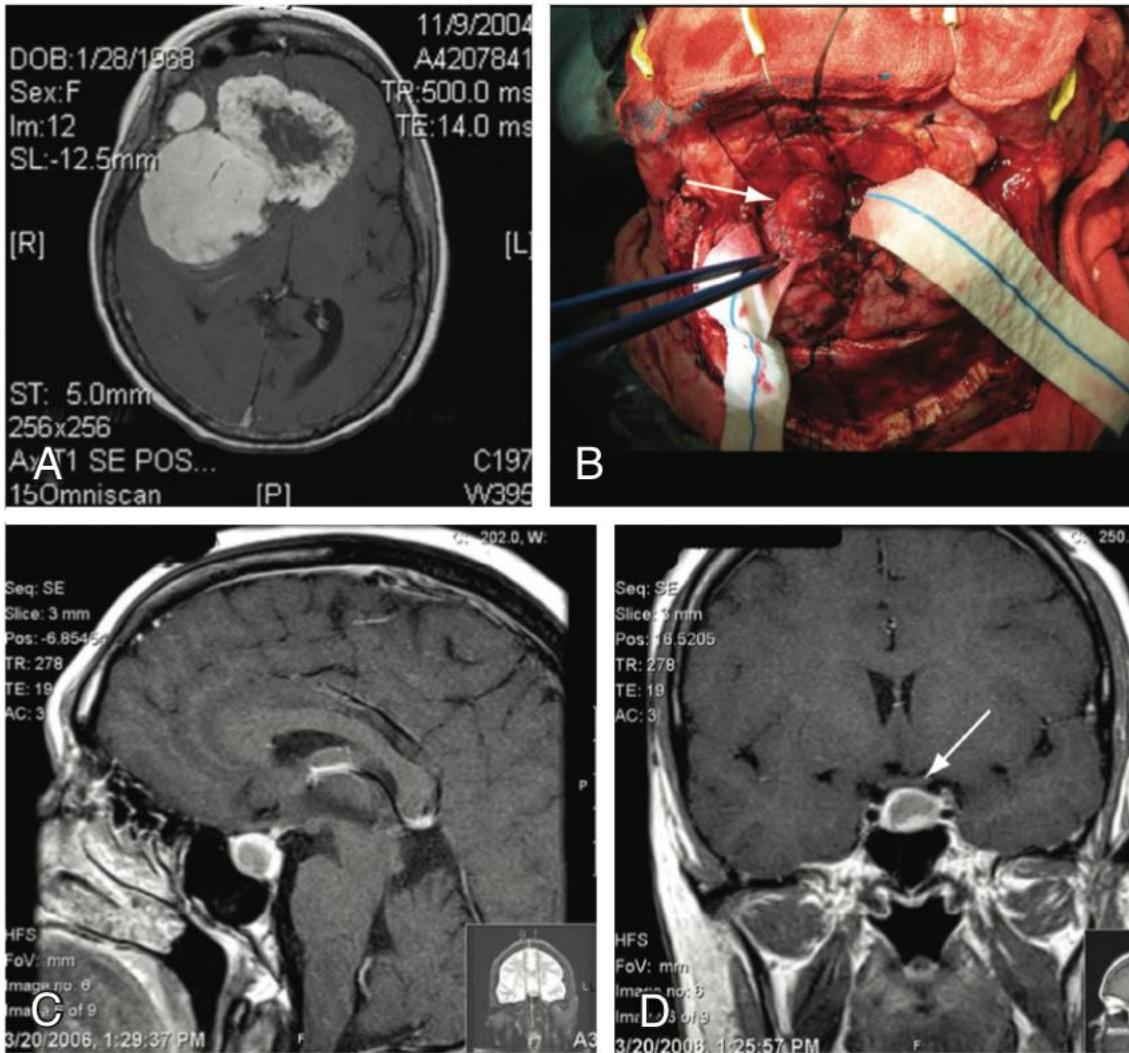
# Meningioma

- Meningiomas are observed in between 15% and 20% of all primary intracranial tumors, second only to glioblastoma multiforme.
- The prevalence is in **females** (2:1), and it is rare in childhood unless it is associated with neurofibromatosis type 1.
- The presentation is usually incidental in up to 50% of cases; they are typically slow growing, and overall 5-year survival is greater than 90%.
- Meningiomas can recur, depending on the resection obtained at surgery as described on the **Simpson** grading system for meningioma resection as well as the atypical histology.
- Surgical resection is the treatment of choice if the patient is neurologically symptomatic.
- **Asymptomatic tumors can be observed.**

# Meningioma

- The degree of resection predicts **recurrence, with rates of 10% at ten years for total excision with a clear dural margin and 30% at ten years for subtotal excision.**





**FIGURE 67-14** **A**, A large enhancing meningioma can be seen in this gadolinium-enhanced axial MRI study. **B**, Intraoperative picture showing dissection of the meningioma (arrow) from the surrounding gyri. Gadolinium-enhanced sagittal (**C**) and coronal (**D**) MRI scans of a patient with a pituitary macroadenoma show impingement on the optic chiasm (arrow).

**TABLE 67-7 Simpson Grading System for Meningioma Resection**

<b>GRADE</b>	<b>EXTENT OF RESECTION</b>	<b>RECURRENCE RATE*</b>
I	Complete including dural attachment and abnormal bone	10%
II	Complete with cauterization of dural attachment	15%
III	Complete without dural attachment	30%
IV	Incomplete resection	Up to 85%
V	Biopsy	100%

\*Length of follow-up varies around 5 years; numbers may increase with longer follow-up.

# Hemangioblastoma

- 25% and 40% are associated with von Hippel–Lindau syndrome (VHL).
- When they are associated with VHL, hemangioblastomas typically occur in young adults with a slight male predominance.
- Hemangioblastomas make up around 10% of posterior fossa tumors; when they are not associated with VHL, they have a sporadic peak at the age of 50 years.
- They tend to present with mass effect because of cyst expansion and typically are slow growing and histologically benign.



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# Primary Central Nervous System Lymphoma

- 2% to 6% of patients with AIDS.
- The mean age at presentation is **60 years** in **immunocompetent** patients and 35 years in patients with acquired immunodeficiency with a slight male predominance.
- The presentation can be with symptoms from a mass effect and, depending on location, sometimes with neuropsychiatric changes.
- There is a dramatic but short-lived response to steroids.
- Treatment consists of **stereotactic biopsy** followed by radiation therapy and **chemotherapy** because of their chemosensitivity to methotrexate.
- **Intrathecal methotrexate** will usually be advised for young patients.

# Germ Cell Tumors

- Germinomas compose 1% to 2% of all primary CNS tumors;
- 50% are found in the pineal region and have most frequently been described in the Japanese population.
- The peak age at presentation is around 10 years, with more than 90% being found in the population younger than 20 years.

# Germ Cell Tumors

- The male to female ratio is 10:1 for the pineal region, whereas suprasellar germinomas are more common in females.
- When located in the pineal region, they can become large and are present with hydrocephalus and Parinaud syndrome. This consists of paralysis of upward gaze, convergence, and accommodation and is associated with lid retraction, creating the so-called **setting sun sign**.

- When located in the suprasellar region, they may produce compression of the hypothalamus and cause hypothalamic- pituitary dysfunction with **diabetes insipidus and visual decline from compression of the optic tracts.**
- Tumor markers help confirm diagnosis and a favorable prognosis when low secretion of human chorionic gonadotropin is observed.
- The first line of treatment consists of biopsy, then radiation therapy plus chemotherapy and treatment of hydrocephalus with either placement of a ventricular peritoneal shunt or a third ventriculostomy.

# **Nongerminomatous germ cell tumor**

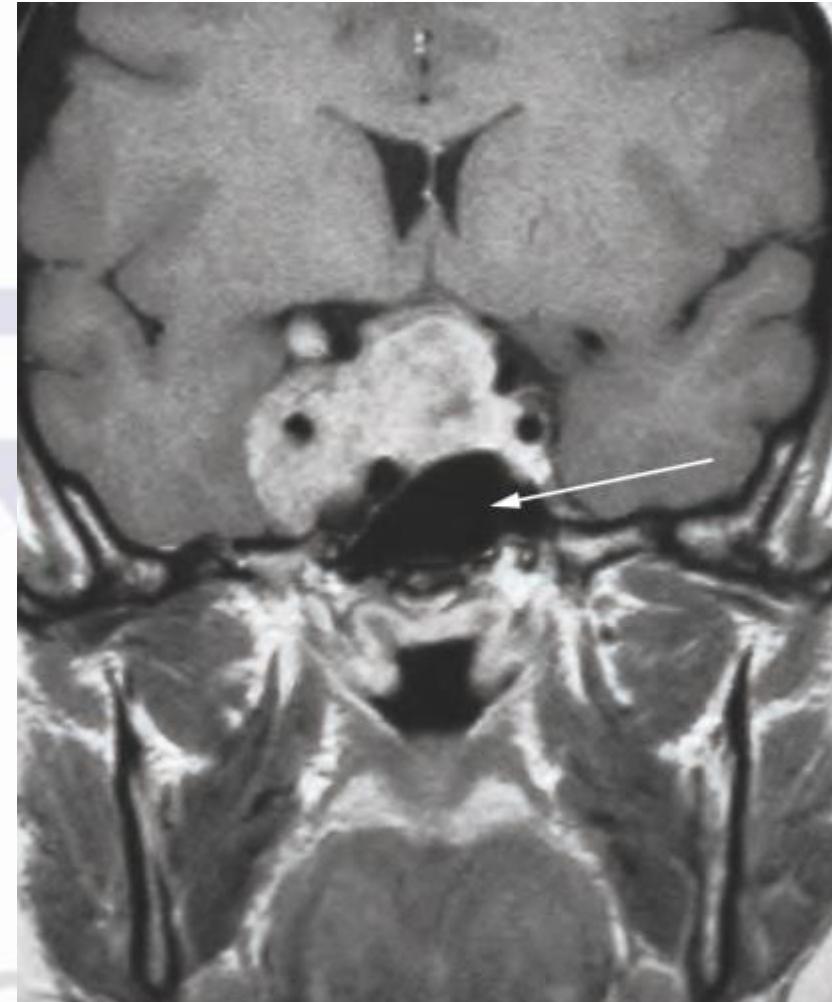
- These tumors are generally associated with a worse prognosis than germinomas are, with a 5-year survival rate of less than 50%.
- Embryonal carcinoma (malignant germ cell tumor) affects prepubertal children, it is associated with Klinefelter syndrome and is considered malignant and invasive.
- Yolk sac tumors are also known as endodermal sinus tumors; they are usually found in infants or adolescents and are aggressive and malignant.
- Choriocarcinomas, which are also malignant and highly hemorrhagic, are another variety.

# Pituitary adenoma

- Pituitary adenomas make up 10% of all intracranial tumors, with an equal male to female incidence; the peak incidence is in the third and fourth decades.
- Associated with multiple endocrine neoplasia syndromes. Around 50% present as macroadenomas that are larger than 1 cm in diameter.
- Symptoms develop from mass effect on the optic tract or hypothalamic-pituitary disturbance with endocrine abnormalities and rarely apoplexy.
- Typically, when it is a **hormone-producing tumor**, symptoms will appear at earlier stages in tumor growth than when nonfunctioning adenomas are found

# Pituitary adenoma

- Microadenomas = less than 10 mm usually present incidentally or with endocrine effects.
- Macroadenomas = larger than 10 mm - present with visual field deficits.
  - **30% are prolactinomas**
  - **20% are non-functioning,**
  - 15% secrete growth hormone and
  - 10% secrete ACTH.



- Prolactin levels of 25 ng/mL or less are considered normal; if the prolactin level is between 25 and 150 ng/mL, it is generally considered “stalk effect,” although levels above 100 ng/mL should be considered suspicious.
- However, when the level is higher than 150 ng/mL, it is considered diagnostic for prolactinoma.

# Classic presentation

- Prolactinoma - amenorrhea and galactorrhea in females and impotence in males. Infertility will be present in both. The treatment consists of dopamine agonist (e.g., bromocriptine) and generally provides complete control.

# Classic presentation

- Adrenocorticotropin adenoma - Cushing disease and classic hyperpigmentation of the skin and mucous membranes, ecchymoses, and purple striae, especially in the flanks, breast, and lower abdomen. Generalized muscle wasting with complaints of easy fatigability are among the other well documented signs and symptoms. The first line of treatment is surgery.

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- **Growth hormone secreting tumors** - acromegaly in adults and gigantism in prepubertal children.
- Surgery is the first line of treatment.
- Some patients may respond to octreotide, and others may show improvement with dopamine agonist.

- **Thyroid-stimulating hormone** secreting tumors may present as hyperthyroidism, anxiety, and palpitations (due to atrial fibrillation).
- Patients have heat intolerance, hyperhidrosis, and thyrotoxicosis, for which the treatment will require surgery.

- **gonadotropin-secreting and nonfunctional adenomas**, clinical presentations will be due to mass effect and stalk compression.
- If the tumor extends to the suprasellar region and compresses the optic chiasm, this will cause bitemporal hemianopia and may also have cranial nerve deficits.
- Treatment is surgical resection.

- After surgery, patients are at risk of CSF leak (3%), and pituitary insufficiency.
- Diabetes insipidus resulting from manipulation of the pituitary stalk is common in the immediate postoperative period and usually resolves spontaneously.

- **Pituitary apoplexy** is the syndrome associated with haemorrhagic infarction of a pituitary tumour. It presents with sudden headache, visual loss and ophthalmoplegia with or without impaired conscious level.
- **Endocrine resuscitation with intravenous steroids** is the priority, and surgical decompression may be required.

# Craniopharyngiomas

- Peak incidence between 5 and 10 years of age.
- Suprasellar masses with compression of the surrounding structures.
- The tumor is histologically benign but may sometimes have local aggressive and relentless behavior.



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# Craniopharyngiomas

- The most frequent postoperative complications include diabetes insipidus and hypothalamic injury with 5% to 10% mortality.
- Attempts to obtain total gross resection should be sought if appropriate.
- If subtotal resection, postoperative radiation therapy might be beneficial.

# Schwannoma

- 8% of all intracranial tumors.
- Vestibular schwannomas will usually have sensorineural hearing loss with tinnitus and dizziness and typically present at an age older than 30 years.
- Slow growing, with an average of 10% recurrence after total resection.
- When vestibular schwannomas are present bilaterally, the diagnosis of Nf 2 should be ruled out.

# *Vestibular schwannoma*

- These are nerve sheath tumours arising in the cerebellopontine angle, which present with **hearing loss, tinnitus and balance problems.** **Facial numbness and weakness are less common**, while large tumours may present with features of brainstem compression or hydrocephalus.
- The differential diagnosis includes meningioma, metastasis and epidermoid cyst

# Neurofibroma

- Neurofibromas are rarely found intracranially and can be associated with neurofibromatosis type 1.
- Along the spinal canal, they can develop into dumbbell-shaped masses as they exit the neuroforamina or on occasion into large peripheral nerve sheath tumors.
- **Painless mass** with slow growth that is histologically benign, but between 2% and 12% can degenerate into malignant peripheral nerve sheath tumor with a high recurrence rate.
- Treatment consists of surgical resection, most neurofibromas will encompass nerve fibers, and total resection results in nerve sacrifice as opposed to schwannoma resection, which usually can be achieved without nerve sacrifice.

# Intra-Axial Brain Tumors

- Intra-axial brain tumors develop from the glia, or supportive structures, of the neurons and are collectively called gliomas.
- Total surgical resection of gliomas is extremely rare because of their ability to infiltrate widely along the white matter tracts and to cross the corpus callosum into the contralateral hemisphere.
- Radiation therapy and chemotherapy options vary according to the histology of the brain tumor.
- Therapy involving surgically implanted carmustine-impregnated polymer combined with post- operative radiation therapy has a role in the treatment of de novo and recurrent high-grade gliomas.

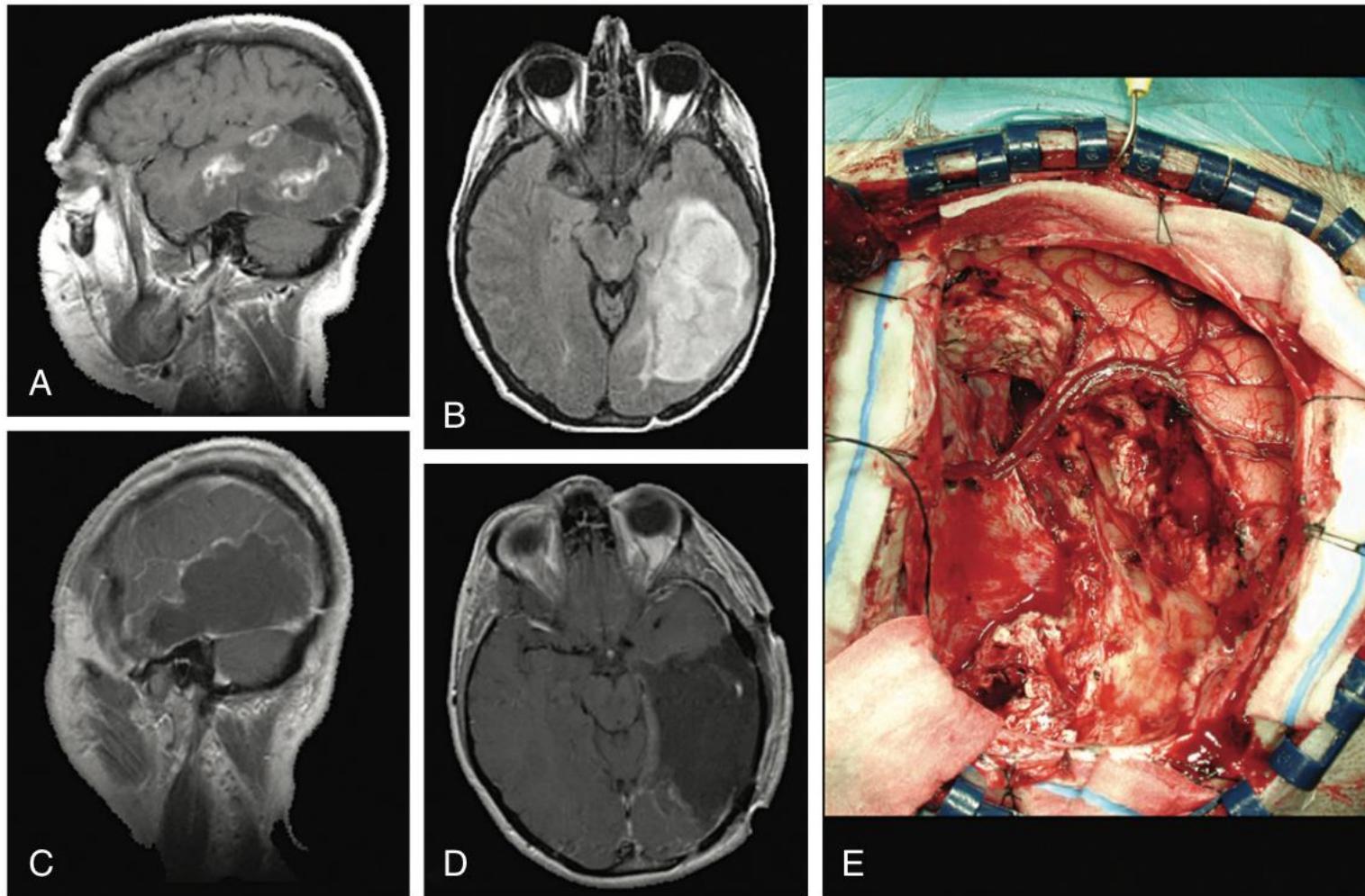
- Biologic therapies include dendritic cell vaccination, tyrosine kinase receptor inhibitors, farnesyltransferase inhibitors, virus-based gene therapy, and oncolytic viruses.
- WHO classifies intra-axial brain tumors by cell type and grades them on a scale of I to IV based on light microscopy characteristics that include the degree of cellularity, pleomorphism, mitotic figures, endothelial proliferation, and necrosis. The higher the grade, the more aggressive and malignant is the tumor.

# Astrocytic Tumors

- Pilocytic astrocytoma WHO grade I gliomas
- When arising in the posterior fossa, they can cause obstructive hydrocephalus and cerebellar signs on examination.
- Surgical resection is the treatment of choice for these posterior fossa lesions. However, for lesions in the hypothalamus or optic tract, biopsy and chemotherapy or radiation therapy should be considered.

# Astrocytic Tumors

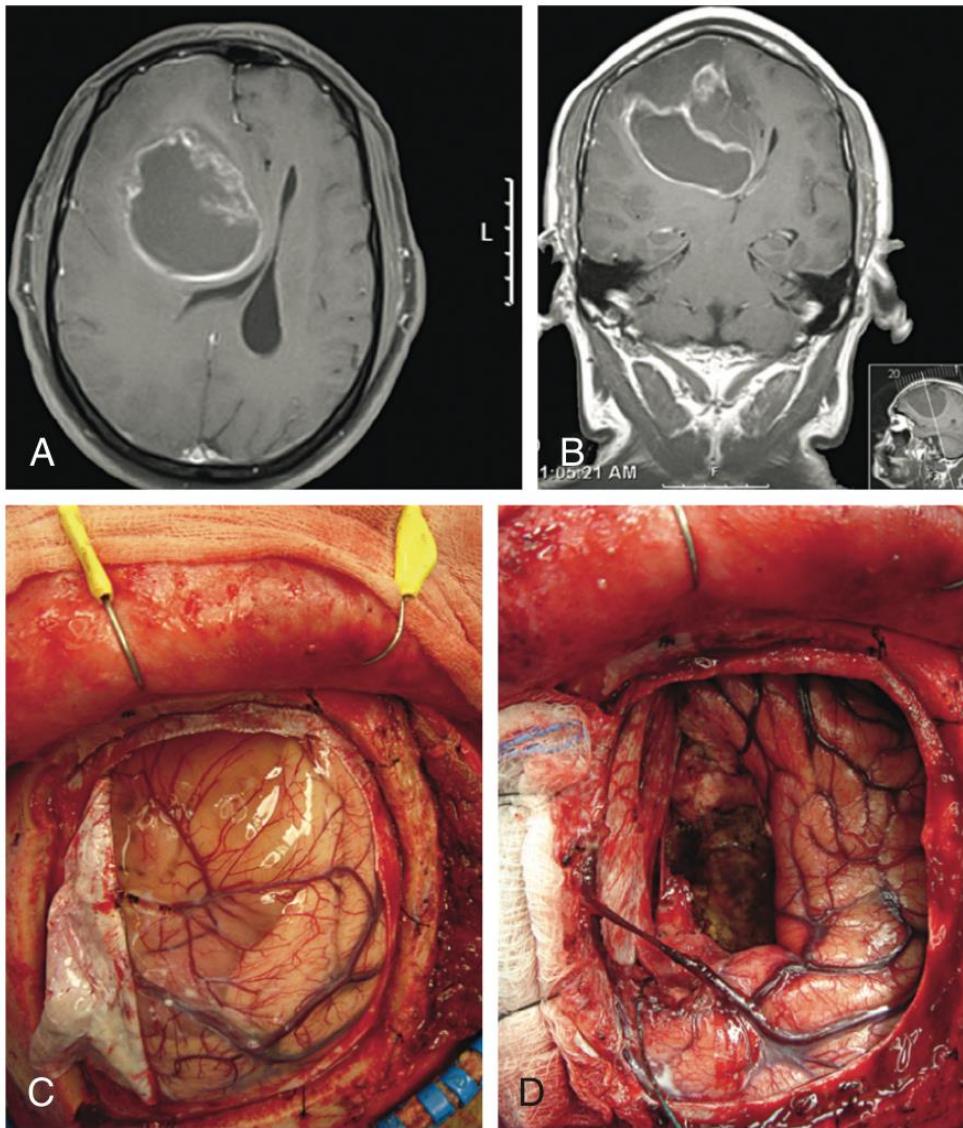
- Anaplastic astrocytoma WHO grade III tumor
- Slightly better long-term prognosis



**FIGURE 67-13** Radiographic and intraoperative images of a patient with a left temporal anaplastic astrocytoma. **A**, A partially enhancing tumor is noted in the left temporal lobe on this gadolinium-enhanced sagittal MRI study. **B**, Fluid-attenuated inversion recovery sequence axial MRI shows the extent of the tumor. The postoperative gadolinium-enhanced sagittal (**C**) and axial (**D**) MRI scans show near-total resection of the tumor. **E**, Intraoperative illustration of the surgical field after resection of the tumor.

# Astrocytic Tumors

- Glioblastoma multiforme WHO grade IV tumor, this is the most commonly encountered primary brain tumor in adults.
- Imaging usually demonstrates a ring enhancing lesion with surrounding edema and mass effect.
- Treatment of accessible lesions involves attempted gross total resection and postoperative radiation therapy and chemotherapy.
- Temozolomide is the current drug of choice.
- Recurrence is common, and repeated surgical resection is often deemed reasonable.



**FIGURE 67-12** MRI and intraoperative pictures of a patient with a glioblastoma multiforme. Gadolinium-enhanced axial **(A)** and coronal **(B)** MRI scans show a large tumor with ring enhancement causing a 1-cm subfalcine shift of midline structures. Intraoperative pictures show the yellowish tumor surrounded by normal brain gyri **(C)** and the surgical field after resection of the tumor **(D)**.

# Oligodendroglioma

- originate from oligodendroglial cells and represent
- 25% of all glial tumors. Another form known as oligoastrocytoma behaves like oligodendroglioma, and both have aggressive anaplastic forms.
- Treatment consists of surgical resection followed by chemotherapy.
- A particularly favorable response rate is associated with tumors that show allelic losses of chromosomes 1p and 19q.
- Radiation therapy is considered for tumors with anaplastic transformation.

# Ependymoma

- peak age at presentation is between 10 and 15 years.
- In children, ependymomas will typically be found in the fourth ventricle floor.
- An ependymoma typically is manifested as a slowly growing posterior fossa mass that may cause obstruction of CSF flow, leading to hydrocephalus and symptoms of increased ICP with nausea, vomiting, and intense headaches.
- Treatment consists of maximal possible resection because extent does affect survival, followed by fractionated radiation.
- Cauda equina lesions are of the myxopapillary variant.

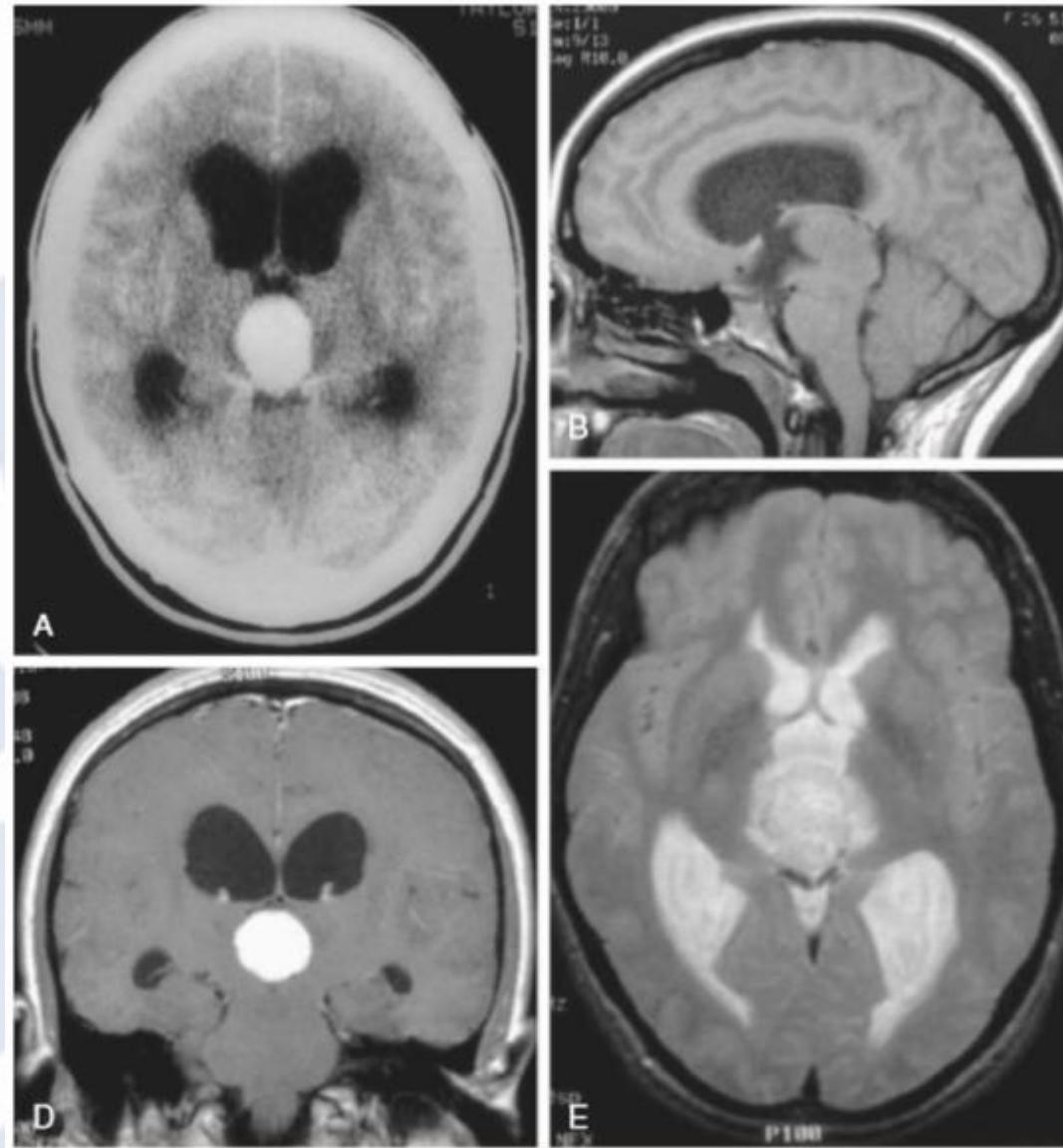
# Pediatric Brainstem Gliomas

- These tumors represent around 10% to 20% of all pediatric brain tumors; the mean age at presentation is 7 years.
- Midbrain gliomas (tectal and tegmental) usually have better survival rates than pontine gliomas.
- Tectal gliomas typically are manifested with hydrocephalus but have up to an 80% 5-year progression-free survival rate.
- Focal tegmental mesencephalic tumors may be manifested with hemiparesis that slowly progresses.
- The diffuse pontine glioma will usually present with multiple cranial nerve palsies and ataxia with increased ICP and have a poor overall median survival of less than 1 year.

- Treatment of tectal gliomas requires vigilant follow-up and frequently CSF diversion or shunting.
- Focal tegmental mesencephalic tumors might be surgically resected and require adjuvant chemotherapy and radiation therapy if they recur.
- In the case of diffuse pontine gliomas, treatment is with radiation with or without experimental chemotherapy or palliative care.

# Pineal Region Tumors

- Pineocytoma
- children and young adults
- C/F hydrocephalus, increased ICP, and Parinaud syndrome (which is a supranuclear vertical gaze disturbance caused by compression of the tectal plate).
- If symptomatic or enlarging, the treatment is surgical.



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# Pinealoblastoma

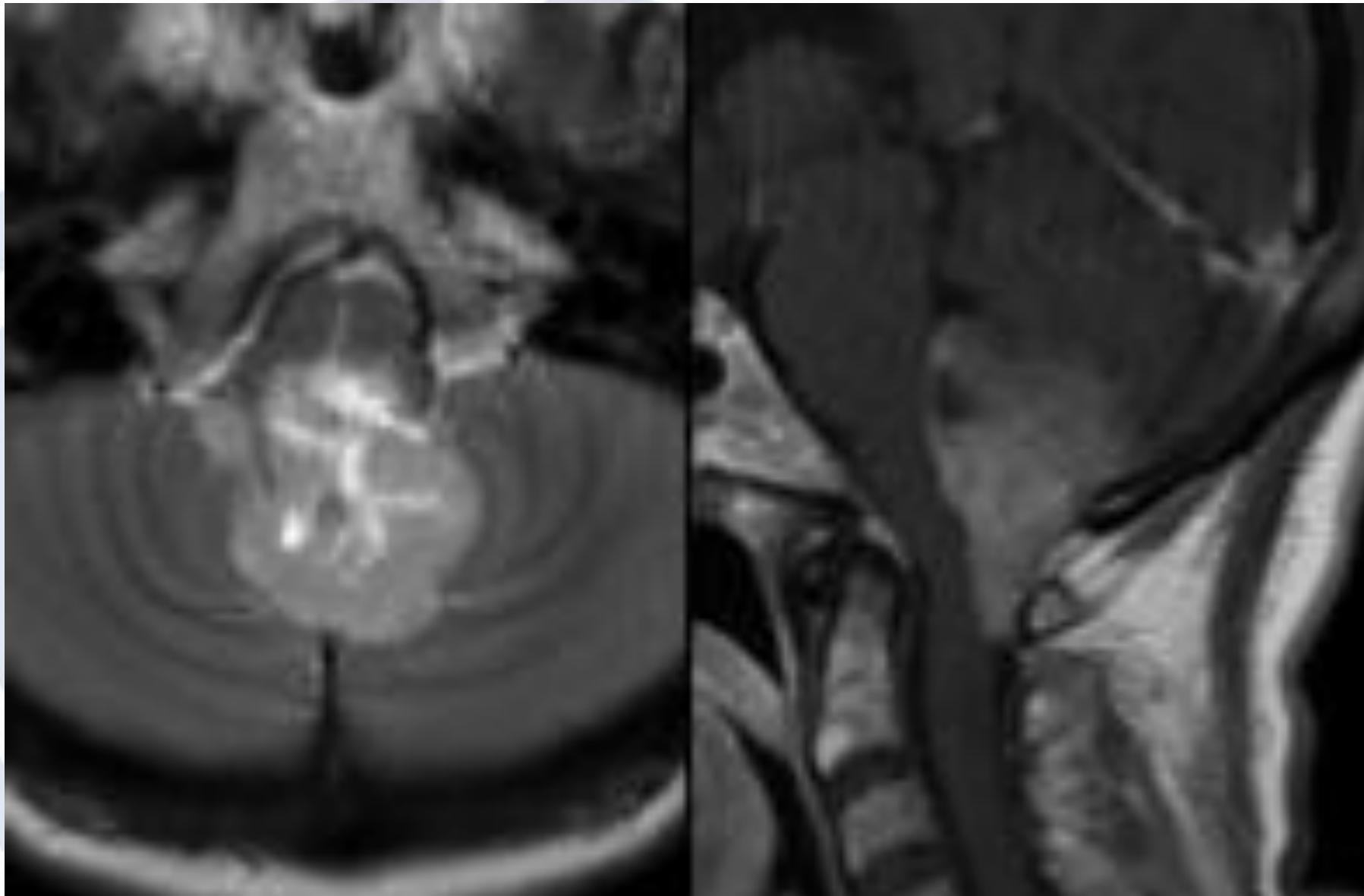
- Present with increased ICP, hydrocephalus, and Parinaud syndrome.
- Up to 50% will have CSF seeding, giving a median survival of 2 years from the time of diagnosis.
- Treatment should consist of surgical resection plus irradiation of the cranial vault and entire spinal axis.
- If the patient is older than 3 years, chemotherapy should also be considered.

# Papillary tumours of pineal region

- They will behave like a grade II or grade III tumor according to the WHO classification. It can recur and require surgical resection followed by focal irradiation.

# Primitive Neuroectodermal Tumors

- Medulloblastomas are found to be 15% to 20% of all brain masses and up to one third of all posterior fossa tumors in the pediatric population.
- Usually diagnosed by the age of 5 years, with a male to female ratio of 3:1.
- They tend to have a rapid presentation with hydrocephalus, increased ICP, and cerebellar signs.
- These tumors tend to disseminate through the CSF and are often found to involve the spinal subarachnoid space in a sizable number of patients at the time of diagnosis.
- Treatment consists of attempted gross total resection followed by adjuvant chemotherapy and radiation therapy if the child is older than 3 years.



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# Central Nervous System Metastasis

- Cerebral metastases are the most common brain tumor in adults and make up more than 50% of all brain tumors across all ages.
- Approximately 20% to 40% of patients with cancer develop brain metastases during the course of their illness.

# Central Nervous System Metastasis

- Most brain metastases arise from lung, breast, and renal cell tumors; however, melanoma, followed by lung, breast, and renal cell carcinoma, has the greatest propensity to develop brain metastasis.
- Breast and renal cell carcinoma tends to present as a single metastasis, melanoma and lung cancers have an increased incidence of multiplicity.

# Central Nervous System Metastasis

- Lung cancer is the most common source of brain metastasis in men, and breast carcinomas are the most common source of metastases in women.
- Men with melanoma are more likely to develop brain metastasis than are women.
- **Breast cancer generally exhibits the longest interval (mean, 3 years) and lung cancer the shortest (mean, 4 to 10 months) for time duration for mets.**
- Metastatic lesions tend to cause significant brain edema that initially will respond well to steroids (Dexamethasone).

- When the lesion is initially encountered and no primary tumor is known, recommendations for stereotactic biopsy or excision should be given.
- A solitary metastasis, total surgical excision should be attempted, followed by whole brain radiotherapy. SRS generally will be recommended if surgery is not feasible.
- When multiple metastases are encountered, consideration should be given to excision of the symptomatic lesion or multiple lesions (but this is controversial), followed by brain therapy or radiotherapy alone, and SRS will usually be considered if surgery is not feasible.



# Pediatric Neurosurgery

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# Cystic lesions

- **arachnoid cyst**: typically middle fossa, CSF enclosed in an envelope of arachnoid mater;
- **colloid cyst**: occur in the roof of the third ventricle, believed to represent embryonic endoderm remnants;
- **dermoid and epidermoid cysts**: epithelial lined structures arising from displaced ectodermal remnants, typically in the posterior fossa (midline) and cerebellopontine angle respectively;
- **porencephalic cysts**: brain cavities lined with gliotic white matter, containing CSF in communication with the ventricles or subarachnoid space.

# Spinal Dysraphism

- The neural tube forms from the neural placode at approximately 21 days of gestation.
- Failure of the neural tube to form results in neural tube defects, such as spinal dysraphism.
- Neural tube defects have already formed by the time pregnancy is diagnosed; thus, prevention of these defects by the administration of folic acid has to commence before 21 days of gestation.

# Spinal Dysraphism

- The spinal dysraphic state can be classified as spina bifida aperta (open defects, usually apparent) and spina bifida occulta (closed defects).
- Spina bifida aperta = myelomeningocele and meningocele.
- Spina bifida occulta = simple spina bifida occulta, spinal dermal sinus, lipomyelomeningocele, diastematomyelia, and tethered spinal cord.

# Myelomeningocele

- There is protrusion of a varying amount of spinal neural tissue outside the spinal canal confines.
- It has been associated with **folate deficiency** in the mother; intake of folate during pregnancy has reduced the incidence considerably.
- There is a deficiency of the skin, muscle, and bone elements, with the open neural placode exposed anywhere from the thoracic to the sacral level.
- **Hydrocephalus** is also present in 80% of patients and sometimes is manifested after surgical closure of the defect.
- **Chiari II malformation**, which occurs in 90% to 95% of cases. Associated brain anomalies include **corpus callosal anomalies, fused tectal plates, and thalamic fusion**.

# Myelomeningocele

- **Surgical closure of the myelomeningocele is undertaken within 24 to 48 hours of birth to avoid CNS infection (e.g., meningitis, ventriculitis).**
- All exposed neural tissue is considered viable unless otherwise proven.
- During the closure, adequate care is taken to separate the neural tissue (placode) from the cutaneous element to prevent an inclusion dermoid.
- Of children with myelomeningocele, 60% to 70% will ultimately require a shunt insertion, whereas only 15% to 30% of children will require a Chiari decompression.

# Myelomeningocele

- Serum and amniotic fluid  $\alpha$ -fetoprotein screening and prenatal ultrasound have been significantly helpful in diagnosing open neural tube defects in the prenatal period.
- A 20% to 65% incidence of latex allergies in this population has led to universal latex allergy precautions for this group of children.



FOR PREGNANCIES

# Meningocele

- a protrusion of dura and arachnoid outside the confines of the spinal canal, with neural tissue remaining within the spinal canal confines. Because no neural elements are present, there are no associated neural deficits and repair is simpler.
- Meningoceles are most common in the lumbar region.

# Spina Bifida Occulta

- A posterior lumbar bone defect is often present in 5% to 10% of the normal population, without any symptoms or deficits.
- However, association of other markers, such as a tuft of hair, cutaneous hemangioma, or sinus track, should be viewed with suspicion and warrants further investigation.



**FIGURE 67-27** Child with lumbar cutaneous hemangioma. This often accompanies an underlying spina bifida (spina bifida occulta) during the clinical evaluation.



**FIGURE 67-28** Myelomeningocele in a neonate. Note the deformity of the lower limbs.

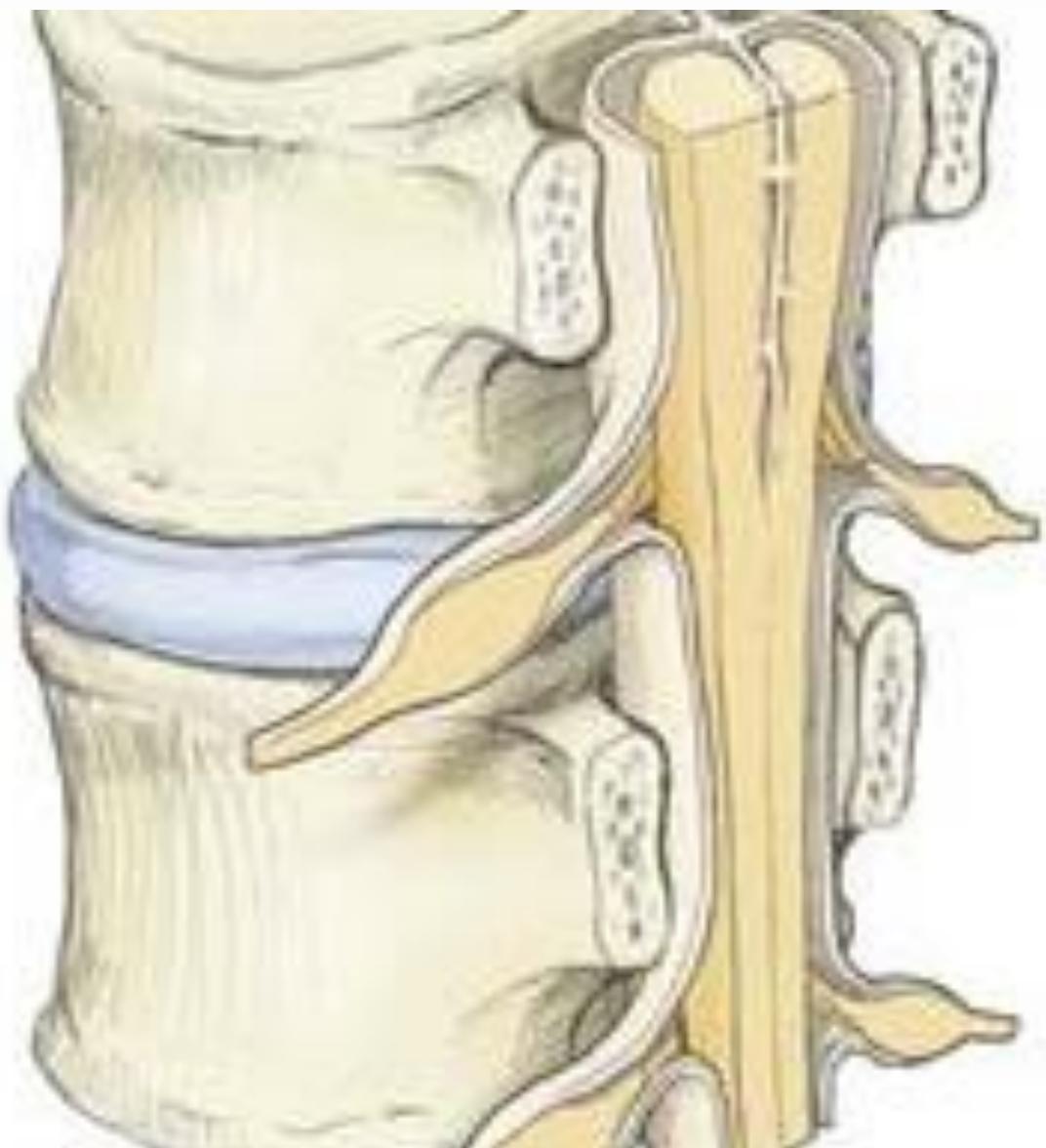
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# Dermal sinus

- Occurrence of a dermal sinus track from the skin to the spinal subarachnoid space is often associated with a cutaneous dimple or pit.
- These are most common in the lumbo-sacral region.
- Ascending infection or be symptomatic, with tethering of the cord.
- It may be associated with intraspinal inclusion tumors, such as dermoids.
- MRI is helpful for assessing the course of the track and its termination.
- The track is usually excised surgically, with care taken to untether the cord.

# Diastematomyelia

- The spinal cord is split into two hemicords, often by a bone or fibrous band that tethers the cord, preventing its free movement and ascent.
- It is often associated with a hairy patch on the back at the defect level.
- This needs to be repaired surgically.



FOR MAGNUM  
FOR SPINOSA

# Lipomyelomeningocele

- Varying amount of fatty tissue in the spinal cord and in the spinal canal tethering the cord.
- Often associated with a large dural defect, associated neurologic deficits, although uncommon at birth, usually develop later because of the tethering.
- Almost all lipomyelomeningoceles have a well-developed skin cover, which allows these children to be operated on electively at a later date.

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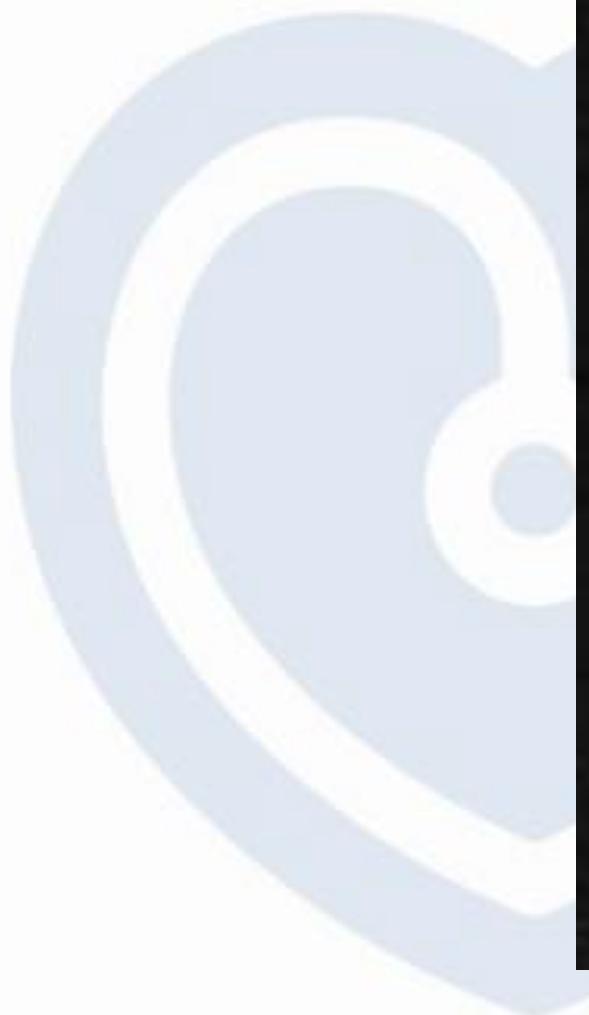
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# Cranial Dysraphism

- Cranial dysraphism includes encephalocele, meningocele, and cranial dermal sinus.
- The encephalocele can be in the cranial vault or cranial base.
- The occipital encephalocele is the most common, followed by anterior encephalocele and then basal encephalocele.
- Encephaloceles may be associated with other developmental anomalies, such as polydactyly, retinal dysplasia, microphthalmia, and orofacial clefts.
- Basal encephaloceles present with a CSF leak from the nose or ear or as a polyp.

# *Chiari malformations*

Chiari malformations - cerebellar herniation through foramen magnum

- An isolated descent of the tonsils below the rim of the foramen magnum without any spina bifida is known as Chiari I malformation.
- Chiari II is invariably associated with open spina bifida and has several other diagnostic features, such as descent of the brainstem and fourth ventricle into the upper spinal canal.
- Chiari type III malformation, there is an associated high cervical encephalocele containing herniated cerebellar and brainstem tissue.

# Chiari I Malformation

- Descent of the cerebellar tonsils more than 5 mm below the rim of the foramen magnum
- The descended tonsils are usually peg shaped, and the descent is associated with crowding of the soft tissue, obstructing CSF flow = Occipital headache
- Associated tingling or numbness in the extremities and impairment of joint position.
- Cavitation of the spinal cord (syringomyelia)
- Wasting and weakness of the extremities, scoliosis, and varying degrees of sensory impairment.



**FIGURE 67-29** T2-weighted sagittal MRI scan of a child with a significant type I Chiari malformation. Note the tonsillar descent below the rim of foramen magnum.

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vi

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# Chiari I Malformation

- Coexistent hydrocephalus is seen in 10% of cases.
- Removal of the rim of the foramen magnum and posterior arch of C1 and duraplasty are the most commonly performed procedures.
- Associated hydrocephalus requires ventriculoperitoneal shunt placement or an endoscopic third ventriculostomy.

# Chiari II Malformation

- Chiari II malformation is characterized by elongation and caudal displacement of the brainstem and cerebellar tonsils and by association with myelomeningocele. Hydrocephalus is common and syrinx occurs frequently.
- Although it is a common accompaniment of myelomeningocele, surgery is reserved for children who are symptomatic with lower cranial nerve paresis, weakness, respiratory distress, or syrinx.



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# Craniosynostosis

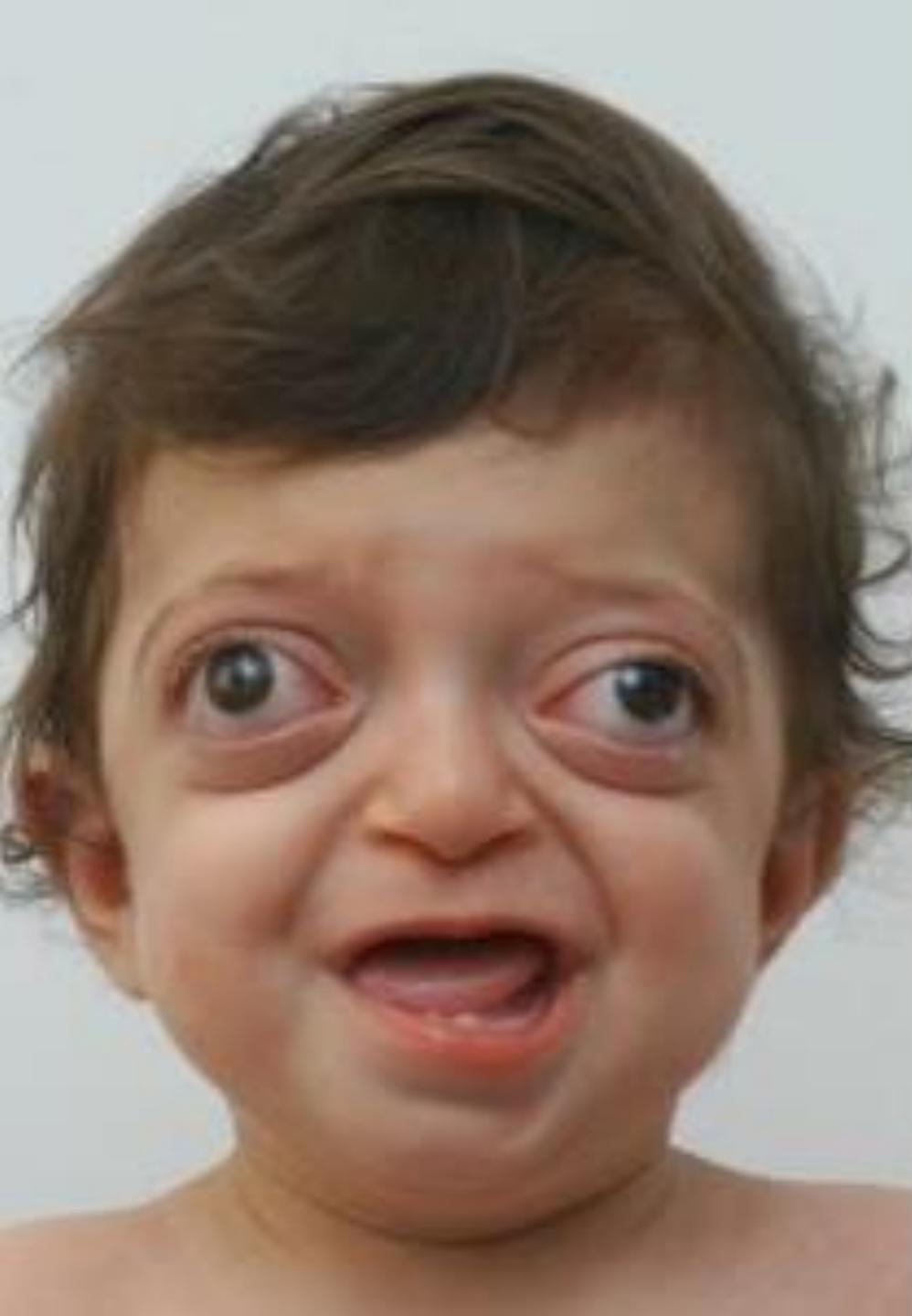
- Craniosynostosis involves premature fusion of the cranial sutures.
- This results in restricted growth of the skull bones at the involved suture and compensatory growth at the adjacent patent sutures, causing disfigurement of the cranial shape.
- In multisutural synostosis, restriction of the cranial growth at various sutures can cause impairment of growth of the developing brain.
- The most common suture involved is sagittal suture (50% to 60%), followed by coronal suture (30% to 35%), metopic suture (5%), and lambdoid suture (2%).

# Craniosynostosis

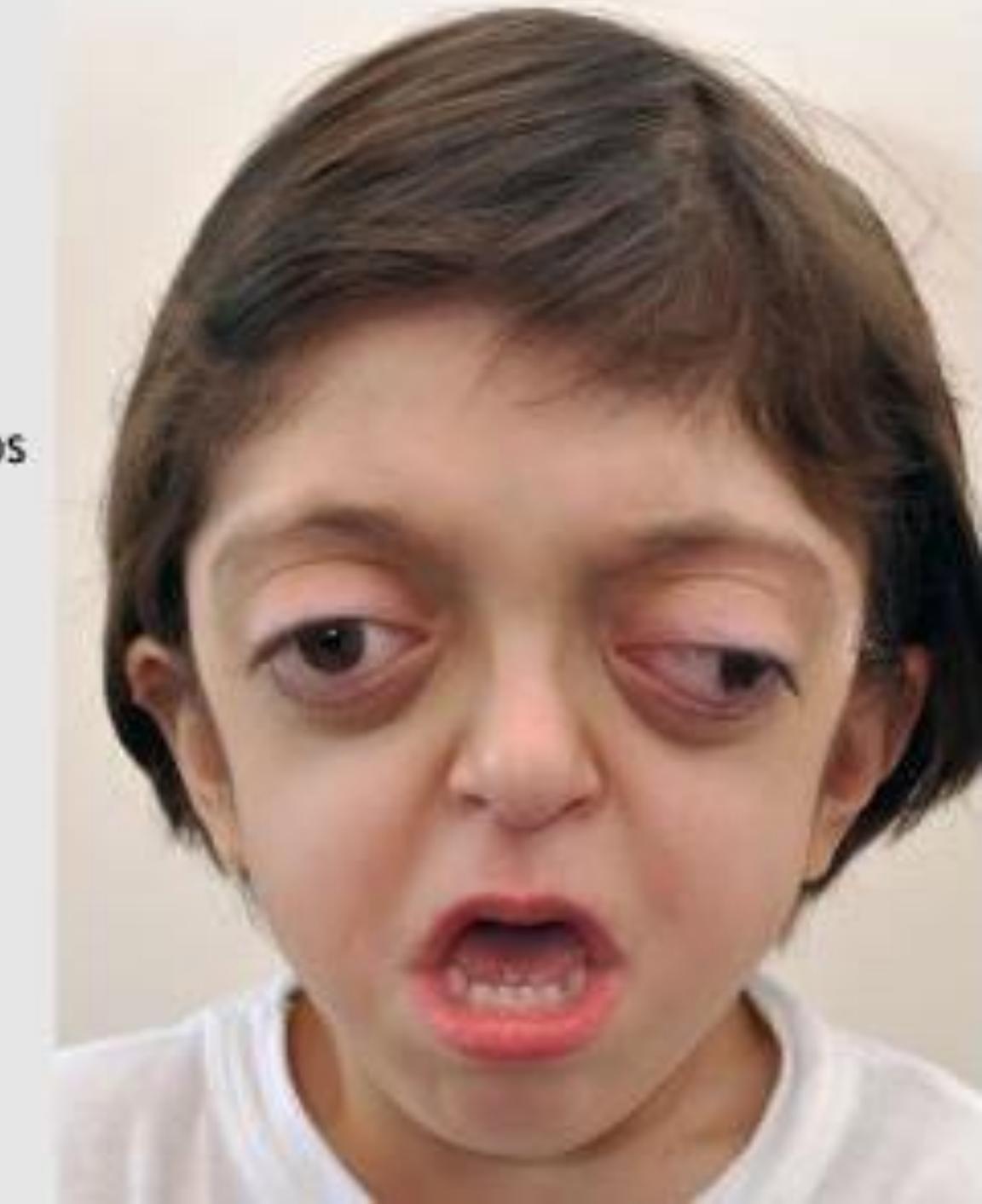
- Lambdoid suture synostosis has to be distinguished from positional plagiocephaly, which is common and does not require surgical intervention.
- More complex disorders, such as Crouzon, Apert, and Pfeiffer syndromes, have a genetic predisposition.
- The clinical picture is recognized by the abnormal skull shape associated with each sutural fusion

# Nomenclature

- sagittal, elongated skull, or **scaphocephaly**;
- coronal, **brachycephaly**; and
- metopic, **trigonocephaly**



Crouzon  
syndrome  
with  
exophthalmos



# Craniosynostosis

- Surgical correction involves a wide suturectomy and placement in a cranial remodeling helmet in children younger than 4 months and a craniotomy and cranial vault reconstruction in older children.
- In recent years, simple suturectomies have been performed in children younger than 6 months under endoscopic guidance with a small incision.
- Patients with coronal craniosynostosis will usually require advancement of the orbital rim in addition to the cranial remodeling.



**FIGURE 67-30** Three-dimensional reconstruction CT scan of a child with sagittal synostosis. The coronal and lambdoid sutures are well visualized.

# *Craniosynostosis*

Syndromic craniosynostosis, often associated with abnormalities of the **fibroblast growth factor receptor genes**, is accompanied by developmental delay and other abnormalities

**TABLE 43.6** Types of craniosynostosis.

Type	Suture involved	Clinical features
Scaphocephaly	Sagittal suture	Narrow boat-shaped head
Brachycephaly	Coronal suture	Shortened/broad forehead
Microcephaly	All sutures involved	Small head
Plagiocephaly	Unilateral coronal/lambdoid suture	Asymmetric skull
Trigonocephaly	Metopic suture	Pointed narrow forehead

# *Dandy walker malformations*

infancy with macrocephaly, developmental delay and hydrocephalus

Imaging demonstrates a **hypoplastic cerebellar vermis**, with the posterior fossa occupied by a large thinwalled cyst.

Treatment usually involves ***shunt placement***.

## *Wada test – cerebral dominance*

**Sodium amytal** is injected into each internal carotid artery in turn, with simultaneous speech and memory **testing to localise function**.

The aim is to confirm language laterality that resection on the side of the lesion will not significantly impair verbal memory functions.

# Epilepsy surgery

Disconnection procedures = **corpus callosotomy**, used for patients suffering drop attacks, and **subpial transections** to isolate a seizure focus in eloquent brain from the surrounding cortex.

Mesial temporal epilepsy – amygdalohippocampectomy

Anatomical hemispherectomy - specific epilepsy syndromes associated with hemiplegia, such as **infantile hemiplegia syndrome**

# Epilepsy surgery

Vagal nerve stimulators can be implanted in severe drug refractory epilepsy, with electrodes applied to the **vagus nerve in the carotid sheath in the neck**. This option can achieve effective seizure control, especially in children, although the mechanism is not clear

Non eloquent lobe – resection of seizure focus (lesionectomy)

# *Movement disorders - DBS*

## ***Condition – Target site***

Parkinson's – Subthalamic nucleus

Dystonia – Globus pallidus interna

Essential tremor – Vim Nucleus (thalamus)

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# Trigeminal neuralgia

***Paroxysmal lancinating pain in the distribution of one or more divisions of the trigeminal nerve. No other neurological deficit***

***Triggered by eating/brushing teeth***

The pain is often attributable to impingement on the nerve by the superior cerebellar artery or other vessels, as first postulated by Walter Dandy.

Bilateral trigeminal neuralgia in younger patients - multiple sclerosis.

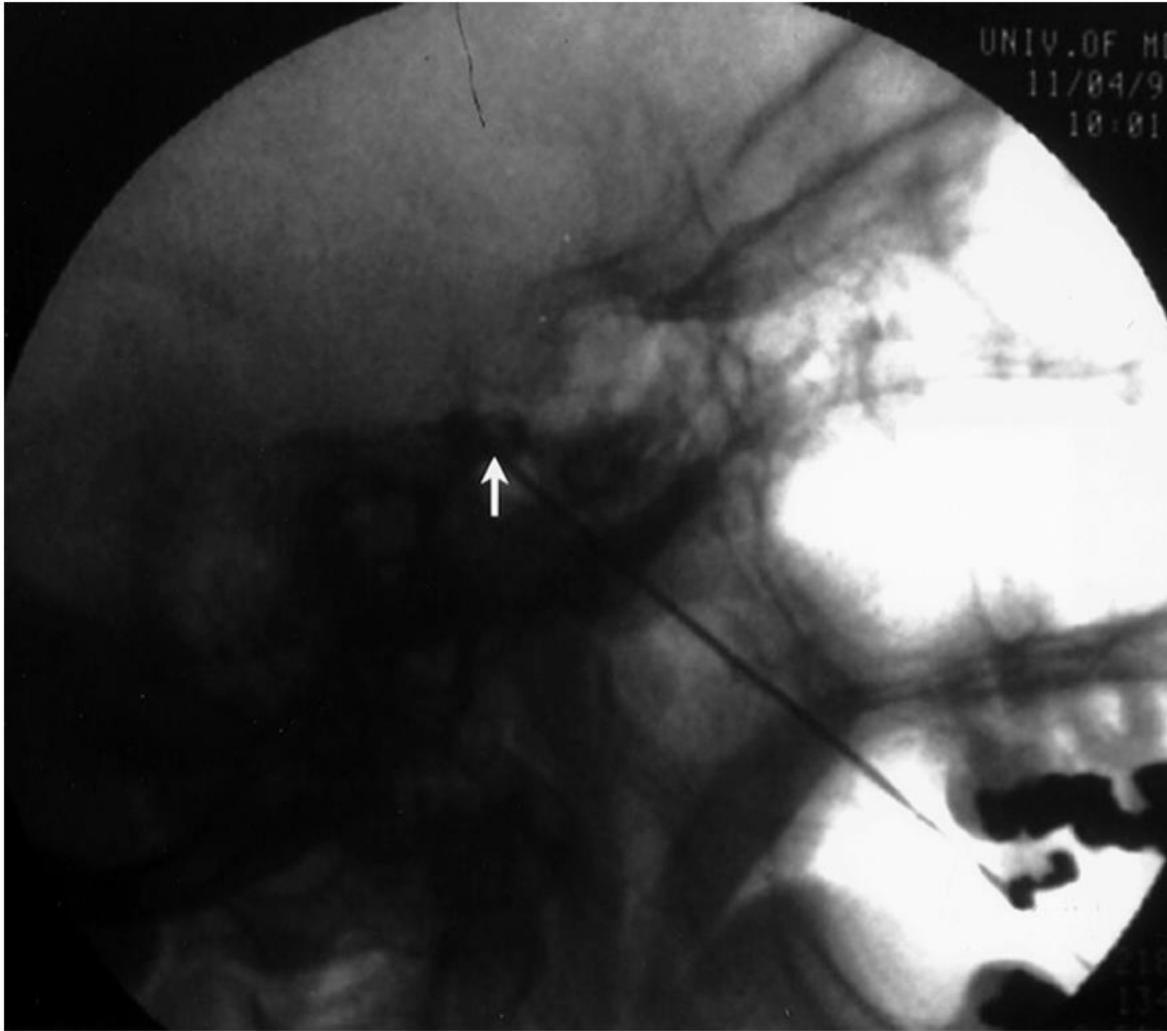
# *Trigeminal neuralgia – surgery*

Microvascular decompression

Peripheral nerve injections

Percutaneous Gasserian rhizolysis - needle placement under radiological guidance at the Gasserian ganglion in Meckel's cave

Lesioning of the ganglion by **glycerol injection, radiofrequency thermocoagulation or balloon compression**, with the aim of disrupting aberrant pain transmission. A similar effect can also be achieved using stereotactic radiosurgery!!!



**FIGURE 67-24** Lateral skull film in a patient undergoing glycerol rhizotomy for typical trigeminal neuralgia. A 20-gauge spinal needle is directed to the foramen ovale and nonionic contrast agent is injected to outline the trigeminal ganglion (arrow).

# Creutzfeldt–Jakob disease

Rare transmissible spongiform encephalopathy

Rapidly progressive dementia, and is uniformly **fatal**

**Prion**, which is not destroyed by conventional sterilisation techniques.

UK practice involves undertaking preoperative checks to exclude any risk factors for CJD infection. These include **family history**, receipt of pituitary-derived human growth hormone, cadaveric dura mater grafts and previous brain or spinal surgery prior to 1997.

# *Risks of craniotomy*

**infection (5%)** and wound breakdown

intracerebral haemorrhage;

seizures;

CSF leak;

permanent neurological deficit;

death (1%)

- 49) Target site for essential tremor is
- 50) Target site for DBS in Parkinson`s disease is?
- 51) Target site for DBS in dystonia is?
- 52) B/L trigeminal neuralgia is suggestive of \_\_\_\_?
- 53) Brain or spine surgery done before \_\_\_\_\_ year is considered as a risk factor for CJD
- 54) Incidence of infection after craniotomy is \_\_\_\_?

# *Brainstem death*

Irreversible loss of cerebral and brainstem function.

**Brainstem death is legally equivalent to death, and is a precondition for the harvesting of organs for transplant from heart-beating donors.**

Diagnosis requires:

1. identification of the cause of irreversible coma;
2. exclusion of reversible causes of coma;
3. clinical demonstration of the absence of brainstem function

# *Brainstem death*

In the UK, this entails testing twice, by two clinicians, to demonstrate the absence of:

response to pain;

respiratory drive (**apnoea despite a  $\text{pCO}_2 >6.7 \text{ kPa}$** );

pupillary light reflex; ● corneal reflex; ● vestibulo-ocular reflex;

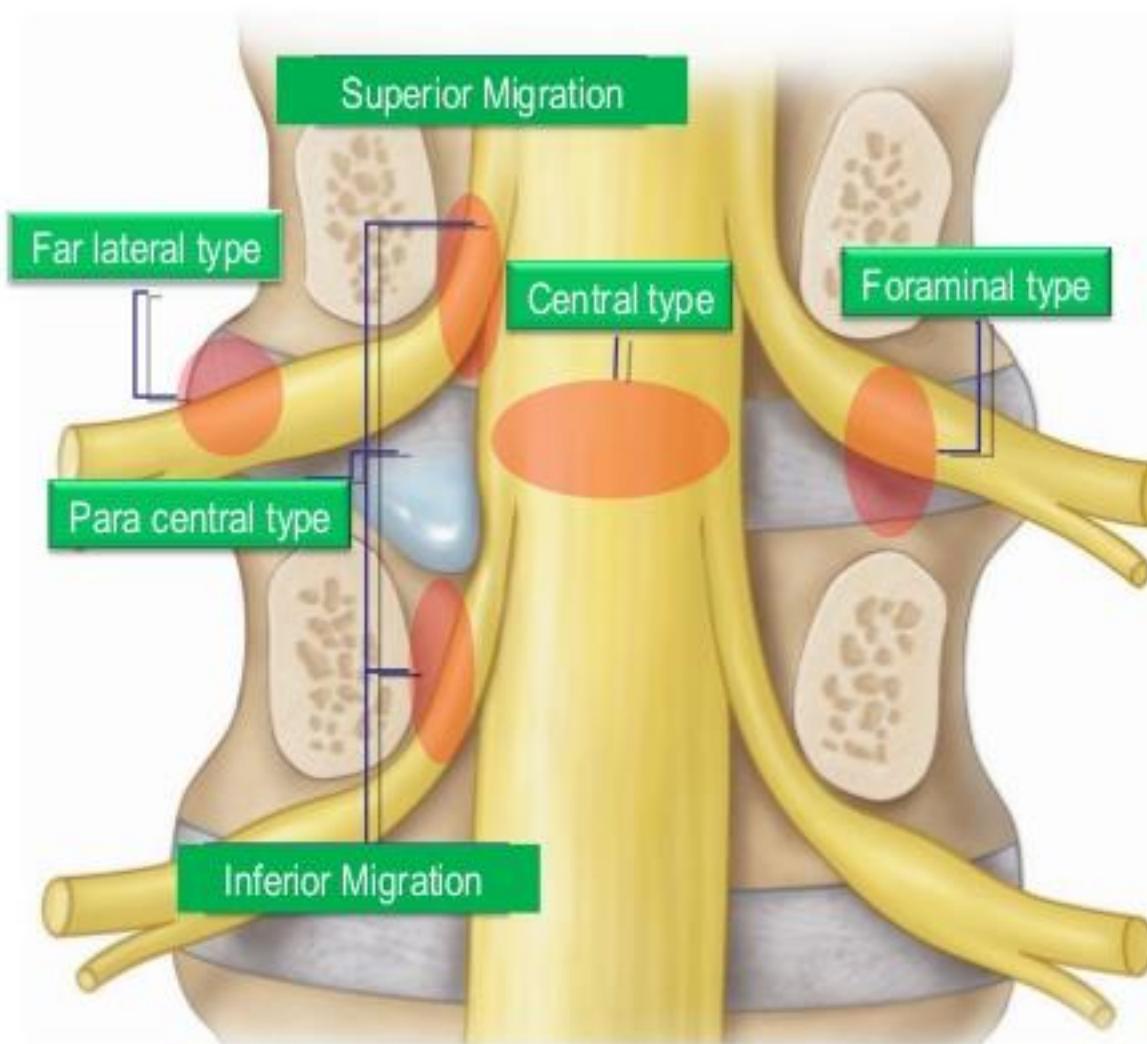
oculo-cephalic reflex; ● gag reflex.

# Relevant spine anatomy

- In adults, the spinal cord terminates at the lower border of L1.
- filum terminale, a relatively fibrous structure, extends from the lower border of the spinal cord to attach at the S2 level.
- The first spinal nerve roots (C1) exit above the atlas, the C2 roots exit between C1 and C2, and the C8 nerve exists between C7 and T1 (thus, there are only seven cervical vertebrae, whereas there are eight pairs of cervical nerve roots).
- This is significant in localization of the spinal nerve involvement by a prolapsed disc as a C5-6 disc will involve the C6 nerve root.

- In the thoracic and lumbar region, the corresponding thoracic nerve root exits below the corresponding vertebra; thus, the L4 nerve root exits between the L4 and L5. However, in the lumbar region, an extreme lateral course of the nerve root while exiting predisposes the next root to be involved in disc prolapse at the corresponding level.
- For example, in L4-5 disc prolapse, the L5 nerve root is commonly involved as the L4 root crosses the L4-5 disc space at the extreme lateral edge, thus escaping compression.
- The L5 root courses across the L4-5 disc space more medially, thus being involved in the process.

## Various types of lumbar disc herniation

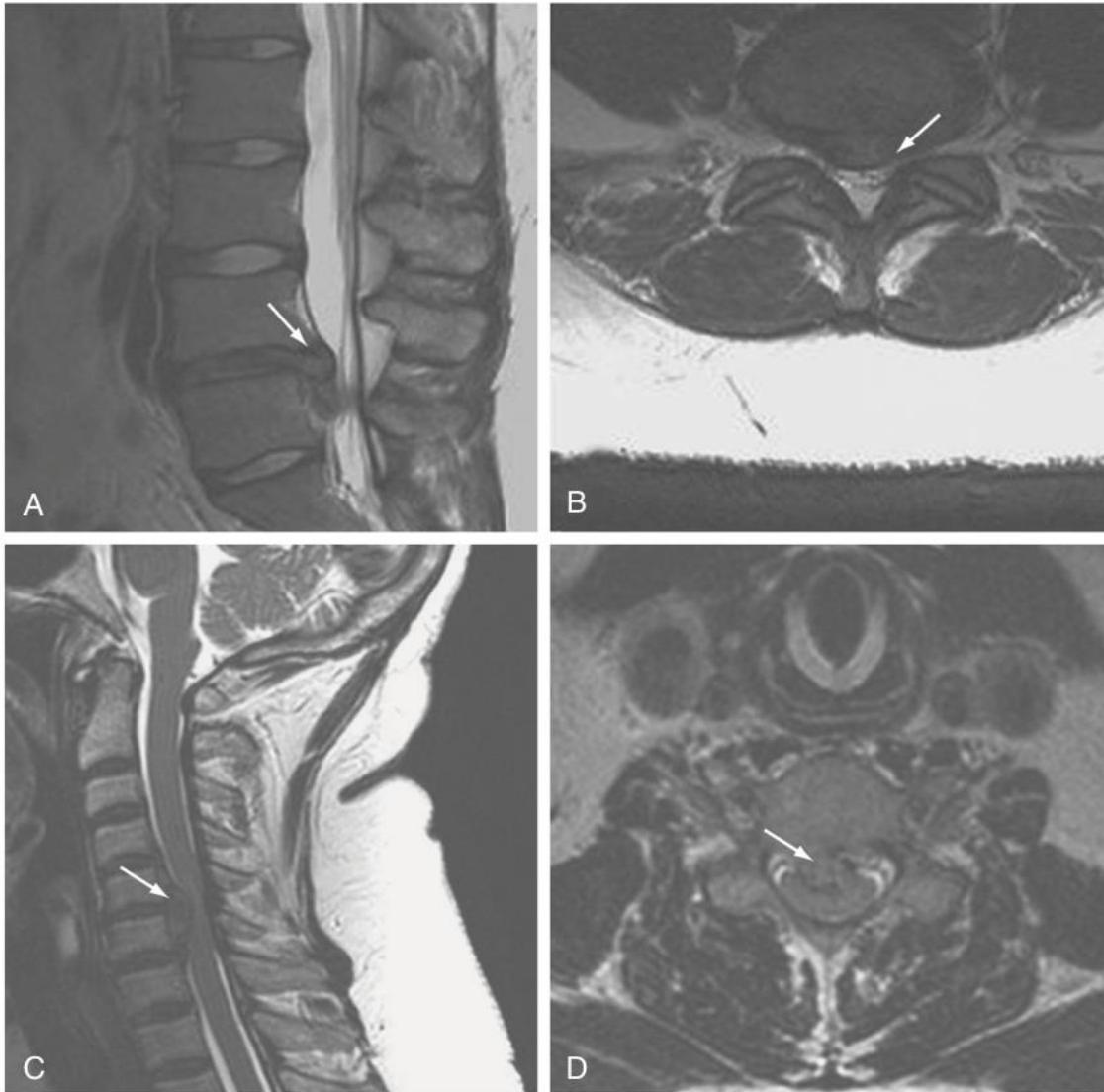


# Cervical Disc Prolapse

- 90% at C5-6 and C6-7 levels.
- An acutely herniated disc typically presents with local pain and tenderness, with the pain radiating in the distribution of the affected nerve root.
- Features of nerve root dysfunction in the form of **motor weakness** or sensory numbness usually follow the initial pain.
- Large disc prolapse can cause spinal cord compression, resulting in **spasticity** and weakness of the lower limbs.

# Lumbar Disc Prolapse

- Mostly L4-5 or L5-S1 level.
- **Acute radiculopathy**, acute back pain precipitated by an episode of lumbar strain, such as bending forward and lifting heavy objects.
- Low back pain radiating to one of the lower limbs + aggravated by coughing or straining.
- Restricted straight leg raising
- Numbness in the dermatomal distribution and associated weakness that may appear a few days later.
- **severe cases = bladder dysfunction.**



**FIGURE 67-17** **A**, T2-weighted sagittal MRI study showing a herniated lumbar disc fragment (arrow) at the L4-5 level. **B**, T2-weighted axial MRI showing the same fragment (arrow) compressing the thecal sac. **C**, T2-weighted sagittal MRI of a patient with a large anterior disc prolapse at C5-6 level (arrow). **D**, Axial images showing the disc compressing the spinal cord (arrow).

# Lumbar Canal Stenosis

- Congenitally (primary lumbar canal stenosis, congenital lumbar canal stenosis) or be secondary to degenerative changes (secondary lumbar canal stenosis).
- Advanced degenerative changes leading to **arthrosis, facet hypertrophy, and ligamentum hypertrophy** result in canal stenosis.
- Presents as **claudication** pain in both the legs that is precipitated by walking and occasionally even by standing for a prolonged period.
- This **neurogenic claudication** is relieved by maneuvers causing **flexion of the spine (e.g., sitting down or bending forward)** in addition to stopping the activity, as opposed to vascular claudication, which is relieved by stopping of the activity.



**Thank you!!!**

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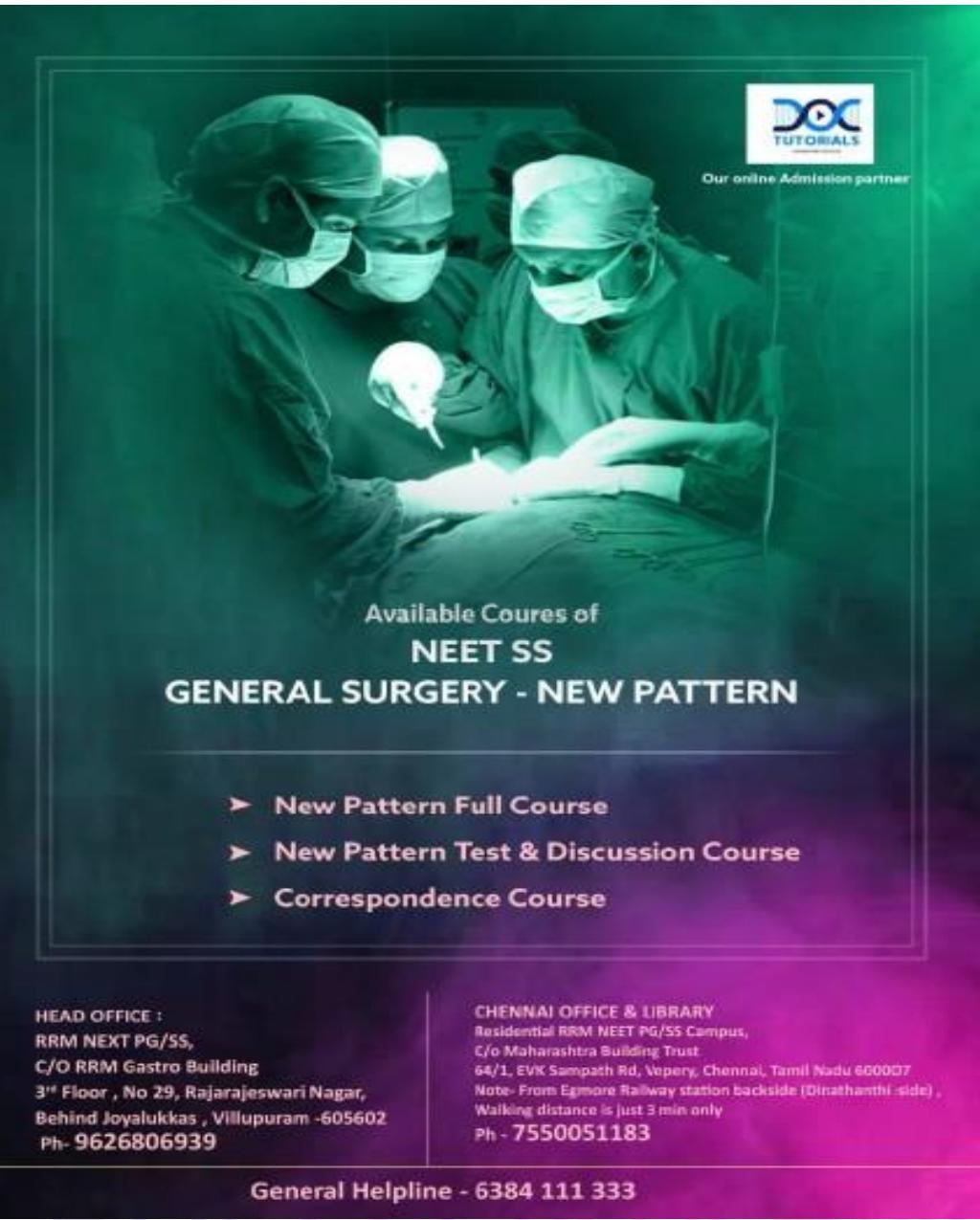
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