HEART

- arterial line should be calibrated at external auditory meatus - to reflect intracranial blood pressure.
- only vasopressor which reduces CSF production (→ ICP↓) is NOREPINEPHRINE.
- PLASMA-LYTE A - **isotonic** 294 mOsmol/L, **140 mEq sodium** (also 5 mEq potassium, 3 mEq magnesium, 98 mEq chloride)

LUNGS

INTUBATION

Rapid sequence induction [RSI] (with adequate sedation and paralysis!!!) is recommended to avoid ICP↑.

1) *SEDATION* - cerebroprotective agent ETOMIDATE (maintains BP, lowers ICP and brain metabolism, has rapid onset and brief duration);
2) LIDOCAINE IV (1-2 mg/kg)
3) *PARALYSIS* - SUCCINYLCHOLINE 1-2 mg/kg.
4) airway irritation (→ ICP↑) is blunted by FENTANYL.

- goal - end tidal CO_2 ($ETCO_2$) 25-30 mmHg with correlating $PaCO_2$ of 30-35 mmHg.

NEUROGENIC PULMONARY EDEMA

(form of ARDS with increase in interstitial and alveolar fluid): suddenly raised ICP → medullary ischemia → *increased sympathetic tone* → sudden shift of intravascular volume from systemic to pulmonary circulation → alveoli fill with fluid as they would in congestive heart failure, but left ventricular end-diastolic pressure (measured by PCWP) is normal.

- etiology: acute central nervous system injury (well documented in SAH but prevalent in ICH).
- radiographically, it is indistinguishable from cardiogenic pulmonary edema.
- treatment: lower ICP + mechanical ventilator.

DVT PROPHYLAXIS

- SCD
- prophylactic heparin (some delay until 24 hours postop or TBI)
  A) HEPARIN 5000 units subQ q8h (head problems); if < 60 kg use q12h
  B) ENOXAPARIN 30 mg subQ q12h (SCI, trauma with long bone fx)
  C) ENOXAPARIN 40 mg subQ daily (ischemic stroke); if morbidly obese use 40 mg q12h

SKIN

Stevens-Johnson syndrome (spectrum of toxic epidermal necrolysis) – reaction to drug (e.g. Dilantin): flu-like with fever, peeling blisters on skin and mucosa – fulminant or worsens up to 2 weeks – self-limited but may be fatal.
Treat in burn unit! (steroids not indicated)

ANESTHESIA

NITROUS OXIDE (N_2O s. "LAUGHING GAS")

- major component of general anesthesia - **minimally influences respiration & hemodynamics**.
- opposite to other gases, minimally ***increases cerebral metabolism***

- most important clinical problem - ***nitrous oxide is 34 times more soluble than nitrogen*** and diffuses into closed gas spaces faster than nitrogen diffuses out - contraindicated in presence of ***closed gas spaces***: air embolism, pneumocephalus - may convert to "tension pneumocephalus"

KETAMINE – does not affect MAP (and CPP) – good inducing agent in hemodynamic instability.

BRAIN EDEMA

BRAIN EDEMA - brain volume↑ due to increase in ***extravascular*** brain water.
– it is general reaction to insults.

N.B. differentiate from **BRAIN ENGORGEMENT** - brain volume↑ due to increase in ***intravascular*** volume (e.g. obstruction of cerebral veins, arterial vasodilatation).

N.B. vasogenic edema is present always! (but steroids are not recommended in strokes & traumas)

- in tumors – VASOGENIC

  N.B. in brain tumor, clinical signs are often caused more by surrounding edema than by tumor mass itself (so deficits maybe reversible with steroids)!

- in infection (abscess, meningitis, encephalitis) – VASOGENIC
- in trauma – VASOGENIC & CYTOTOXIC (reaches maximum at 48-72 hours)
- in ischemic stroke – CYTOTOXIC → VASOGENIC (progressively worsens for 3-4 days)

	Cerebral Edema		
	Vasogenic	**Cytotoxic**	**Hydrocephalic (s. interstitial)**
Pathogenesis	Capillary permeability↑ ***= BBB disruption***	Cellular swelling due to ***membrane pump failure***	Intraventricular fluid↑
Location	White matter, exception is corpus callosum	Gray & white matter	White matter (periventricular)
Edema fluid	Plasma filtrate	Intracellular H_2O & Na	CSF

Extracellular fluid volume	↑	compensatory ↓	↑
Contrast enhancement	+	-	-

DEXAMETHASONE – for vasogenic edema – decreases BBB permeability.
MANNITOL – for cytotoxic edema (mannitol in vasogenic edema is not effective - leaky capillaries do not form osmotic barrier!)
ACETAZOLAMIDE – for interstitial edema.

MALIGNANT BRAIN EDEMA INTRAOP

- see 00. Condensed (last minute) read >>

ICP

5-15 mmHg (50-195 mmH_2O) with zero at midbrain level / foramen of Monroe (***external acoustic meatus*** is anatomic landmark).

N.B. intracranial compensatory mechanisms can accommodate 50-100 ml (rate of volume change is very important!).

- when these buffering systems are **exhausted**, even small increase in volume (e.g. vasodilatation) → dramatic ICP↑↑↑
  Spatial compensation → → Spatial decompensation

- when ICP↑ compromises CPP → autoregulation is lost (**vasoparalysis**)

Early morning **projectile vomiting without nausea** (precedes appearance of headache by weeks) is especially suggestive – due to direct pressure on vomiting centers in brainstem.

Papilledema - most reliable sign of ICP↑ (good specificity; sensitivity observer dependent)

- possible **other ocular symptoms** - visual obscurations, visual loss (typically - blind spot enlargement, visual field constriction), diplopia (CN6 palsy due to stretching)

CUSHING reflex (= acute life-threatening ICP↑↑↑ → medullary hypoxia):

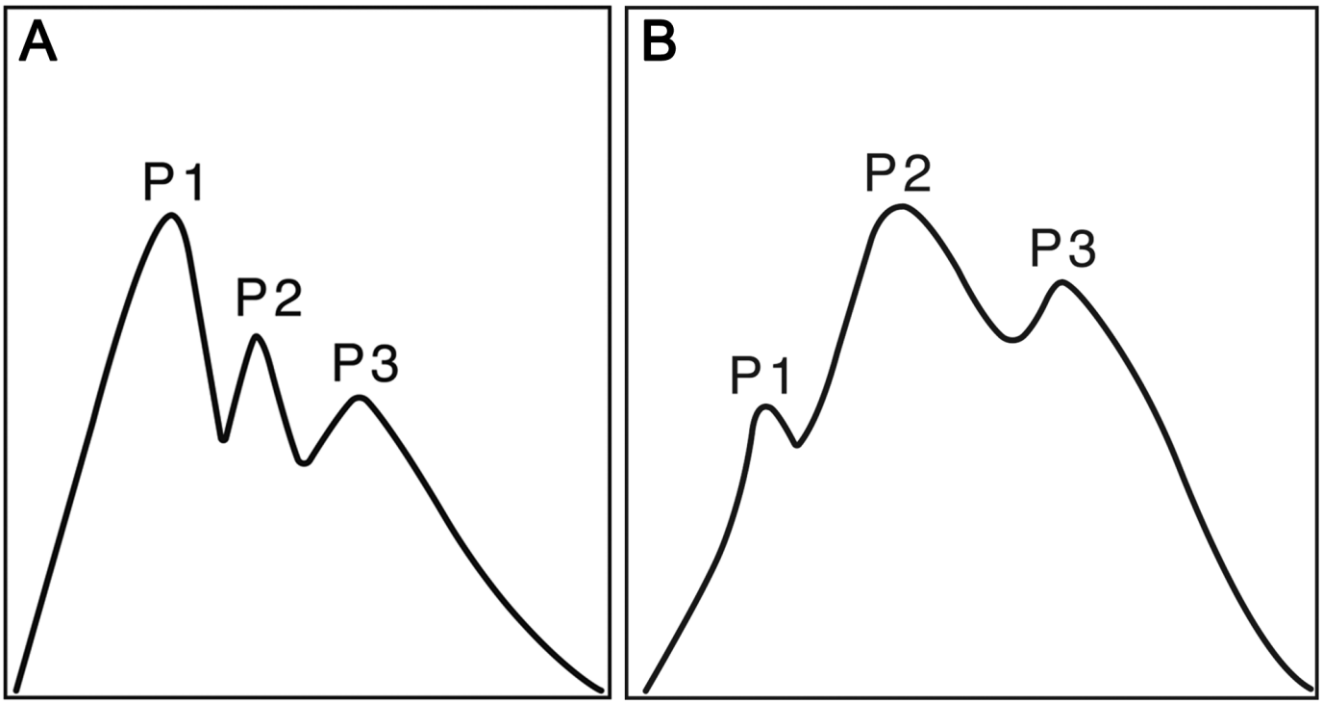
1) **hypertension** – compensatory attempt to restore brainstem perfusion.
   N.B. antihypertensive therapy → critical cerebral ischemia!
2) reflex **bradycardia**
3) **respiratory irregularity** (tachypnea, hyperpnea, Cheyne-Stokes respiration) → neurogenic pulmonary edema.

Cushing ulcer (s. Rokitansky–Cushing syndrome) - gastric ulcer associated with elevated ICP; ulcers may also develop in the proximal duodenum and distal esophagus.

ICP WAVEFORMS

A. Normal ICP pulse waveform:
   P1 (percussion wave) – derives from pulsations of large intracranial arteries.
   P2 (tidal wave) – derives from **cerebral elastance**.
   P3 (dicrotic wave) – corresponds to dicrotic notch in normal arterial pulse waveform.

B. Impaired intracranial compliance results in P2 having the greatest amplitude (increased conductance of pressure waves through "tightening" brain):



LUNDBERG A WAVES (S. PLATEAU WAVES) - acute ICP elevations (**> 50 mmHg**); last **5-20 minutes** and then spontaneously return to baseline level that is slightly higher than when waves began.

- plateau waves are accompanied by dilatation of cerebral arteries and, at the same time, a decrease of the cerebral blood flow.
- these findings indicate that during the plateau waves there is an increase of the intracranial blood volume.

- *poor prognosis*! (ICPs > 30 mmHg are rarely associated with intact survival).
- represent autoregulatory response to insufficient cerebral blood flow (secondary to elevated ICP or systemic hypotension) that produces vasodilatation → increased cerebral blood flow → further ICP elevations.

  N.B. mechanism – ***vasodilation***; proposed treatment – vasopressors (but they actually may worsen lesion and edema formation).
- may be precipitated by iatrogenic maneuvers (suctioning, physical therapy, excess fluid administration, pain).

IMAGING

- *erosion of lamina dura of dorsum sellae* '**J-shaped**' **sella** (in children - *separation of sutures*, "**beaten-silver appearance**").
- small ventricles
- flattened pituitary → empty sella
- passively narrowed dural venous sinuses.
- Gosling index↑ (> 1) on TCD.

TREATMENT

- management of increased ICP is indication for care in NEUROINTENSIVE CARE UNIT.
- **LIDOCAINE** 1% 1-1.5 mg/kg IV to blunt ICP elevation during airway manipulation (e.g. suctioning, endotracheal intubation).

HYPERVENTILATION - to achieve mild ***hypocarbia*** ($PaCO_2$ 30-35 mmHg) and ***avoid hypoxemia*** (PaO_2 80-100 mmHg).

$PaCO_2$ is most potent regulator of cerebral vessel size! **PaO_2** is also important!

Between 20 mmHg and 80 mmHg, CBF is linearly responsive to PaCO2

1-mmHg change in $PaCO_2$ is associated with 3% change in CBF

$PaCO_2$ ↓ to 20-25 mmHg reduces CBF by 40-45% → adult cerebral blood volume reduces from 50 mL to 35 mL (only 15 mL intracranial volume decrease may have tremendous beneficial effect!)

Fortunately, even if pressure autoregulation is frequently lost in TBI, CO_2 vasoreactivity remains!

- $PaCO_2$ < 20-25 mmHg → significant vasoconstriction → cerebral ischemia (especially hazardous after few hours after brain trauma when cerebral blood flow is already diminished to almost ischemic levels)
- when hyperventilation is discontinued, $PaCO_2$ should be tapered over 24-48 hours (to avoid rebound vasodilatation)!
- PEEP can ***increase intrathoracic pressure*** → ICP↑.

Although effect of PEEP on ICP is complex, PEEP should not be withheld if necessary for oxygenation!

OSMOTIC AGENT:

20% MANNITOL 1.0 (0.25-2.0) g/kg IV bolus over 15-30 minutes* q3-6 h (practically, one bag of 20% 500 mL = 100 g of MANNITOL); renal losses must be replaced with isotonic saline

***if IV drip** → MANNITOL accumulation in brain (esp. injured areas with damaged BBB) → reverse osmotic effect; if **more rapid bolus** → transient CBF increase (→ ICP↑)

N.B. the risk of acute tubular necrosis and renal failure has been suggested with mannitol administration with serum osmolarity > 320 mOsm in adults (however, the literature supporting this finding is limited in scope and was generated at a time when dehydration therapy was common).

23.4% NACL 30 mL via central line over 15-30 minutes

- mannitol eventually causes hypotension vs. 23.4% NaCl causes increase of intravascular volume.
- hypertonic saline may be hazardous for a hyponatremic patient (too rapid rise in [Na] may cause central pontine myelinolysis).
- much higher levels of serum osmolarity (approx. 360 mOsm) may be tolerated in children when induced with hypertonic saline vs. mannitol.

N.B. world TBI experts (***Seattle Conference 2019***) recommend same limits (osmolality 320, Na 155) regardless if mannitol or hypertonic NaCl are used!

- mechanisms of action:
  - hypertonicity and osmotic diuresis (**brain dehydration**); effect starts after 15-30 minutes (intact BBB is needed) and lasts 6 hours.
  - immediate plasma expansion → immediate blood viscosity↓ (**rheological effect** – most important mechanism of action!!!; effect lasts < 75 minutes. MANNITOL increases bleeding during surgery for the first 15-30 minutes (rheological effect?) – give during scalp incision.

ICP PROTOCOL (indication: requires mannitol < q12hrs): sedation **FENTANYL** IVI + **PROPOFOL** → paralytic (e.g. **VECURONIUM** or **CISATRACURIUM** IVI – titrate to 1-2 TOF) → moderate therapeutic **hypothermia** (up to 32-34°C).

Rewarming – by 0.5°/day

LUND protocol:

1) **lowering CBV** by ***lowering mean arterial blood pressure***
2) alleviating brain edema by ***reducing hydrostatic forces in damaged capillary beds*** (by **precapillary vasoconstriction** with DIHYDROERGOTAMINE) + ***increasing plasma oncotic pressure*** (ALBUMIN infusion).

- initial clinical trials of this approach yielded outcomes that were no worse than those achieved with more conventional techniques.
- this protocol is in stark contrast to principles of CPP management, which state that low CPP stimulates arteriolar vasodilatation, causing increases in both CBV and ICP.

BRAIN HERNIATION

1. **Lower ICP**:
   1) **hyperventilation**.
   2) MANNITOL.
   3) DEXAMETHASONE.
2. **Neurosurgical intervention** (evacuation of mass).

CENTRAL (S. DOWNWARD TRANSTENTORIAL) herniation

N.B. coma may develop before eye signs appear!

progression of transtentorial herniation is often accompanied by **DURET hemorrhages**

UNCAL (S. LATERAL MASS) herniation

Early CN3 palsy! → coma

Hemiplegia is not as accurate lateralizing sign as is mydriasis (50% vs. 80-85%)

Best diagnosed by evaluating CSF spaces around midbrain (**MESENCEPHALIC CISTERNS**) - absence or occlusion of mesencephalic cisterns usually indicates dislocation of supratentorial structures below tentorial notch!

- movement of more posterior aspects of medial temporal lobe may *compress posterior cerebral artery*.

CINGULATE (S. SUBFALCINE) herniation

- does not affect consciousness!
- ischemia in *anterior cerebral arteries* (loss of function in opposite **leg**, loss of **bladder** control).

CEREBELLAR TONSILLAR herniation

- medulla compression:
  1) stiff neck
  2) flaccid quadriplegia
  3) ***apnea and circulatory collapse*** → secondary coma (medullary RF has little direct role in arousal!!!)

Posterior fossa mass - strong contraindication to lumbar puncture!

UPWARD TRANSTENTORIAL herniation

ETIOLOGY - **posterior fossa mass** + **(overdraining) ventriculostomy**.

- midbrain compression: AMS + ocular signs
- prepare for *emergent posterior fossa decompression*.

CSF LEAK

RING (HALO) TEST
glucose concentration: in CSF ≥ 30 mg/dl (in lacrimal secretions / nasal mucus < 5 mg/dl)
β2-transferrin assay (present in CSF) - most accurate diagnostic test for CSF!

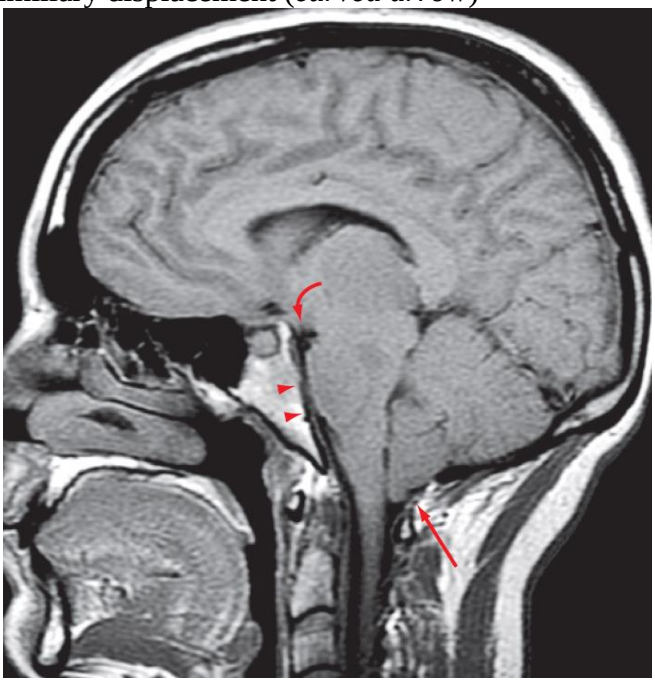
significant CSF loss may cause brain sag with subdural hematomas/hygromas, venous infarcts, brain herniation, meningitis

CT bone window – fluid in paranasal sinuses, skull fracture.

overpressure CT cisternography - instillation (via LP) of water-soluble contrast into CSF → temporarily occlude both jugular veins for 4-5 min to encourage active leakage → **CT with patient placed prone*** → contrast medium in sinuses or nasal cavity.
*leaking is likely to be maximal in this position

MRI with intrathecal gadolinium (CT is less sensitive – bone and intrathecal contrast look the same and obscure each other).

MRI with systemic gadolinium - ***signs of intracranial hypotension***:
1) striking enhancement of the thickened dura
2) globular pituitary (as opposed to empty sella in intracranial hypertension)
3) cerebellar tonsillar descent (*arrow*)
4) pons "flat tire sign" (*arrowheads*)
5) retroclival mammillary displacement (*curved arrow*)



Intrathecal fluorescein (via LP) → ENT performs endoscopy.

TREATMENT
Keep flat / HOB up
Oversew, pressure dressing
Lumbar drain

Epidural blood patch - forms gelatinous tamponade, stopping CSF leak

- 10-20 mL of autologous blood slowly injected (1-2 mL every 10 seconds) into epidural space at site of CSF leak
- in patch failures (15-20%), second patch is often successful.

Operative repair

Shunt – last resort (in case of coexisting hydrocephalus / pseudotumor cerebri)

POSTSURGICAL CSF LEAK

Spinal

Keep flat + hydrate

Blood patch – mainly for post-LP headache

Open direct repair is the best! add glue, lumbar or cervical CSF drain.

- thoracic CFS leak into pleural cavity (hard to control due to negative pressure) – definitely need lumbar or cervical CSF drain

Cranial (CSF leaks into mastoid air cells, frontal sinus)

- HOB up
- Diamox

A. **Early leak** – from poor closure H: **resuture / explore** - **direct dural repair with vascularized flap** (e.g. galeal)
B. **Late leak** (> 2-3 weeks postop) - do LP first:
   a) infection → **treat meningitis** (CSF leak will stop)
   b) CSF contaminated with blood, bone dust, necrotic debris → inflammatory & mechanical interference at arachnoid villi → CSF pressure↑ / hydrocephalus H: **lumbar drain** at 10 cc/hr (if leak recurs → **shunting**)

CSF from frontal sinus ("failed pericranial flap"): 3-5 days LD → endoscopic repair → open exploration

Persistent CSF leak – likely due to ICP↑→ **VPS**

CONSCIOUSNESS

AWAKE = AROUSED = ALERT

CONSCIOUSNESS = AWARENESS

AWARENESS is *not modality specific* - responds to various stimuli

ATTENTION - ability to respond to particular types of stimuli (*modality specific*) – depends also on *specific sensory pathways*

AWARENESS is primarily function of **cerebral cortex** (vs. **AROUSAL** - **brainstem**).

DELIRIUM - state of ***awareness*** (may be even↑) ***without attentiveness***

COMA - ***cannot be aroused*** - profound unconsciousness: ***does not make attempt to avoid noxious stimuli, eyes are closed***

STUPOR - only ***continual intense stimulation*** arouses patient.

OBTUNDATION, **LETHARGY**, **SOPOR** - ***unnaturally deep sleep***

DROWSINESS - ***simulates light sleep*** - can be easily aroused.

VEGETATIVE STATE – state of ***arousal without awareness*** - loss of cortex with preservation of ARAS – eyes open*, does not respond to any stimuli.

*N.B. spontaneous eye opening is sign of arousal, not awareness! *spontaneous roving eye movements are particularly characteristic*

If some response is preserved - **MINIMALLY RESPONSIVE STATE (MINIMALLY CONSCIOUS STATE)**

Persistent (diagnostic category) - vegetative state *for ≥ 1 month*.

Permanent (prognostic category) - vegetative state *for ≥ 3 months* (if brain injury was medical) or *> 12 months* (if brain injury was traumatic) - further improvement is unlikely!

Jei *gilioje* komoje vyzdžiai *simetriški & išlikusi r-ja į šviesą* – komos priežastis ***metabolinė*** (džn. barbiturate poisoning; but in pentobarbital coma pupils become paralyzed).

Intact reflex lateral eye movements = intact brainstem, no mass lesion in posterior fossa!
Lack of reflex lateral eye movements + preserved pupillary reactivity = metabolic / drug toxicity.

BRAIN DEATH

= legal death - neither ***cerebrum*** nor ***brain stem*** is functioning (single exception is ***osmolar control*** - diabetes insipidus is not required for BD diagnosis).

Purely spinal reflexes may be present! incl. sitting up (**"Lazarus" sign**)

- BD rarely lasts more than few days (always followed by circulatory collapse* even if ventilatory support is continued); mean = 4 days.

*progressive hypotension that becomes increasingly unresponsive to catecholamines

no response to painful *CENTRAL stimuli* (*PERIPHERAL stimuli* may elicit spinal reflex movements and may confuse family)
temperature > 36°C
SBP > 100
electrolytes must be WNL + no known **endocrine** disturbances + *absence of drug intoxication*

N.B. for patients coming out of **pentobarbital coma**, wait until level < 10 mcg/mL

known *structural* cause – at least **6 hours** observation (absent brain function)
others (incl. anoxic-ischemic brain damage, children) – at least **24 hours** observation
Absence of cephalic reflexes, incl. pupillary (pupils should not be constricted!), corneal, oculocephalic, oculovestibular (caloric), gag, cough

60-100 ml ice water into one ear with HOB at 30° - wait at least 1 minute for response, and 5 min before testing the opposite side

Not triggering the ventilator = "not breathing over the vent"
Apnea off ventilator (with ongoing oxygenation) for duration sufficient to produce hypercarbic respiratory drive ($PaCO_2 \geq 60$ mmHg for 2 minutes or > 20 above baseline).
Optional confirmatory studies – if unable to tolerate apnea test (hypotension, arrhythmias), if high cervical cord injury

1. **EEG** - isoelectric for 30 minutes at maximal gain.
2. Absent **evoked responses** (BAER and SSEP)
3. Absent **cerebral circulation** demonstrated by CTA - most definitive confirmatory tests

- when BD criteria are met, it is legal time of death – *artificial ventilation* and *blood pressure support* are no longer an option (unless organ harvesting is intended).

NEUROLOGIC EXAMINATION

See "Intro (neuro - nerves, muscles).pdf"

DIAGNOSTICS

IMAGING

CT signal is dependent on ***electron*** density; MRI signal – ***proton*** density.

N.B. *dural enhancement* and *pial enhancement* have clearly different appearances - never use term "meningeal enhancement"!

PLAIN X-RAY

– nonroutine; indications:

SKULL – congenital anomalies, osteolytic / osteoblastic disorders, some trauma cases.
SPINE – trauma F/U, degenerative conditions, evaluation of instability.

CALDWELL - orbital structures
TOWNE - foramen magnum, entire occipital bone
SUBMENTOVERTICAL – skull base

Temporal:

STENVERS – internal ear, internal auditory canal
SCHÜLLER - mastoid air cells

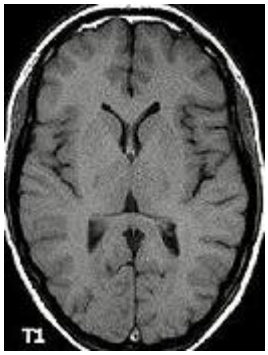
Facial:

WATERS

MRI

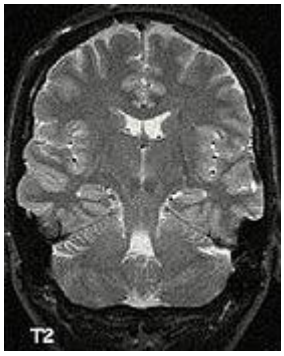
T1 (**fat** is bright, water is dark - white matter [more fat] is brighter than grey matter) - used to obtain high-resolution anatomic detail.

- can be used with gadolinium (in spine may add fat-suppression).



T2 (**fat** is bright, **water** is bright - grey matter [more water] is brighter than white matter*) - more sensitive in detecting focal abnormality *same as CT

FLAIR – T2 with nullified water – used in brain
STIR – T2 with nullified fat ("T2 fat saturation") – used in spine



Screen all patients for:

1) **implanted devices** (even if previously removed - portions of leads often remain).
2) **metallic foreign bodies** (esp. history of metalwork)

Normal tissues: - intermediate signal; ↑ small increase; ↑↑ bright; ↓ small decrease; ↓↓ dark.

Tissue	T1	T2
Fat	↑↑	↑
Bone, calcifications*	↓↓	↓↓
CSF, edema (water)	↓	↑↑
Gray matter (more water)	-	-
White matter (more lipid)	↑	↓
Flow void in vessels**	↓↓	↓↓

*bone (calcium), air has *no protons* – always dark

***moving protons* in vessels demonstrate signal (s. flow) void (↓↓↓ signal of both T1 and T2); CSF flow is more difficult to see (except at aqueduct - fastest CSF flow).

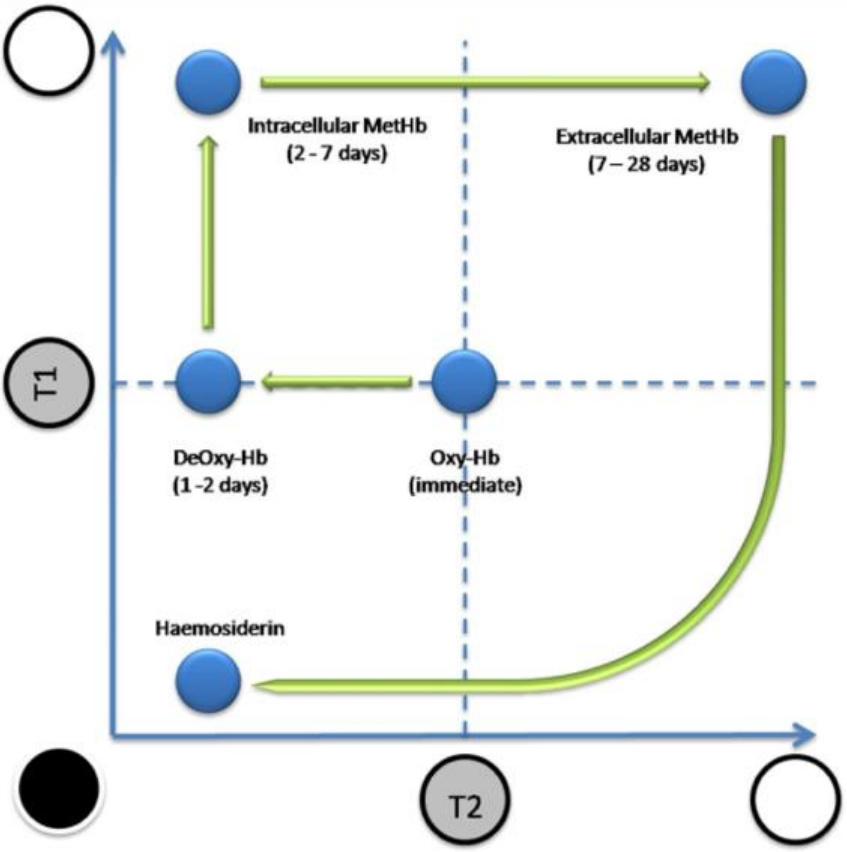
Abnormal tissues / processes:

Material	T1	T2
Air	↓	↓
Acute / subacute infarct	↓	↑
Melanin	↑	↑ or ↓
Demyelination		↑

Only a few naturally occurring substances appear bright on T1:

1. **Lipid** (also bright on T2 and "empty area" on CT) – lipoma, dermoid cyst
2. **Melanin** (likely dark on T2) – **melanoma**!
3. **Protein** – colloid cyst, Rathke cleft cyst, cholesterol granuloma
4. **Methemoglobin**

Blood



- **GRE (gradient-recalled echo), SWI (susceptibility-weighted imaging)** – most sensitive MRI sequence for *acute/subacute blood* breakdown products.

CT

(without contrast) – only for acute head / spinal trauma, acute stroke (esp. hemorrhagic), bone disease, spinal column disease; for other cases – **MRI**

Tissue	Hounsfield Unit	Gray Scale
Air	-1000	very black
Fat	-100	black
Water*	0	black
Cerebrospinal fluid	4-10	black
White matter	22-36	gray
Gray matter	32-46	gray
Extravasated blood	50-90	white
Contrast medium**	100	white
Bone	1000	very white

*edema (water content↑) is seen as lucency

**in any case of suspected acute hemorrhage, contrast medium should not be administered (similar attenuation characteristics to acute extravascular blood)

N.B. ***acute hematoma*** has high attenuation owing to clot retraction with separation of high-density erythrocytes from lower density plasma;

N.B. ***unclotted blood*** (coagulopathy or hyperacute active bleeding) is seen as relative lucency!

INTRAVENOUS CONTRAST ENHANCEMENT

Four questions before contrast:

1. Allergy to contrast
2. Nephropathy? (GFR)
3. Diabetic? (metformin – for CT)
4. Pregnant – counsel for:

   CT:

   low risk of childhood cancer - meta-analysis showed 6% increase in risk of childhood cancer per 100 rad

   doubled risk of congenital defects above baseline population risk (5% → 10%) with 5-10 rad

   - CT of maternal abdomen is 3 rad, head or cervical spine < 0.01 rad
   - shielding of abdomen (lead vest), does not significantly reduce minimal fetal radiation exposure but may help to alleviate maternal anxiety!

   MRI - gadolinium *is contraindicated*

Plus, two questions for MRI – metallic foreign bodies, neuroimplants.

- incidence of reaction is much lower with MRI contrast agents (vs. CT contrasts) - *MRI is modality of choice when contrast-enhanced CNS examination is indicated*.
- maximum dose with normal renal function – 90 gm of *IODINE* in 24 hour period (CTA uses ≈ 21 gm of iodine) vs. intrathecally (myelography) – max 3 gm of iodine
- free gadolinium is toxic to tissues (so MRI contrast is chelated); gadolinium deposits several months later in brains – FDA found no evidence it is harmful

Clinical situations in which contrast is recommended:

1. Infection
2. Inflammation
3. Neoplasia
4. Process thought to involve leptomeninges, nerve roots
5. Seizures
6. Spinal:
   1) intramedullary lesions
   2) subarachnoid lesions
   3) extradural malignant lesions
   4) **postoperative spine** (to separate scar [enhances] from recurrent disk [does not enhance])

Clinical situations in which contrast is not recommended:

1. Hemorrhagic event
2. Ischemic event
3. Congenital anomaly
4. Head trauma
5. Neurodegenerative disease (dementias, etc)
6. Hydrocephalus
7. Spinal cord – trauma, degenerative disease (not operated)

NORMALLY ENHANCING STRUCTURES

1. **Lack of BBB** - dural structures (falx and tentorium), pituitary gland, pineal gland.
2. **Blood (contains contrast material)** - vessels (esp. slowly flowing blood within cavernous sinus or cortical veins), choroid plexus.

ALLERGY TO CONTRAST

e.g. patient allergic to shellfish → use MRI or premedication:

1. PREDNISONE (50 mg oral) – three doses: 13, 7, and 1 hour before study
2. DIPHENHYDRAMINE (50 mg oral) 1 hour before study

N.B. history of laryngospasm / hypotension with previous use of contrast → anesthesiologist should be present during contrast administration

KIDNEY FAILURE

After ***iodinated contrast*** – **hemodialysis** on patient's regular schedule / soon after study.
After ***gadolinium*** – **hemodialysis** for three consecutive days (start immediately after MRI).

GFR has to be:
> 45 for iodine, else risk of *CONTRAST NEPHROPATHY* (rise in serum [creatinine] ≥ 1 mg/dL within 48 h); prophylaxis – **good hydration (± bicarbonates, acetylcysteine)**

N.B. avoid of iodine contrast in *diabetics* - iodinated contrast may delay excretion of METFORMIN → **lactic acidosis** (H: withhold metformin 48 hrs prior to and following contrast administration)
Acute renal failure - absolute contraindication!

> 30 for gadolinium, else risk of *NEPHROGENIC SYSTEMIC FIBROSIS*

INTRATHECAL contrast enhancement (CT cisternography)

Primary approved agent - IOHEXOL (Omnipaque®)
10 ml of Iohexol (concentration of 240 mg/ml) is usually more than adequate

N.B. any myelography should be avoided when arachnoiditis is suspected!

Inadvertent intrathecal injection of ionic contrast*
*i.e. agents not specifically indicated for intrathecal use.

Clinical Features (significant fatality rate): uncontrollable seizures, intracerebral hemorrhage, cerebral edema, coma, paralysis, arachnoiditis, hyperthermia, and respiratory compromise
Management:

1. **Elevate head of bed 45°**
2. **Immediately remove CSF** through myelography needle → **lumbar drain**
3. IV **steroids**, **antihistamines**
4. **AED**

Contrast medium injection into spinal cord – commonest major complication in cervical myelography; *deaths have been reported*

FETAL NEUROIMAGING

- early detection of congenital malformations / destructive lesions → termination of pregnancy.

a) ***early pregnancy*** – **ultrasound**; ***ventriculomegaly***.
N.B. *ventricles are normally large* in fetus < 20 weeks!
N.B. fetal *brain is smooth* with few if any developed sulci - migrational malformations (e.g. agyria) are impossible to detect prior to 18 weeks' gestation.

b) ***late pregnancy*** – **MRI**.

NEONATAL NEUROIMAGING

- bedside **sonography** – can detect ***periventricular*** pathology, ***dysraphisms**** (→ **MRI**).
  *possible because posterior elements are membranous rather than bony in < 6 mos old
- **CT** could wait until at least 6 (preferably 12) months of age (e.g. to give abnormal calcifications time to develop).
- normal ultrasound + normal CT = most major malformations and acquired lesions are excluded → **MRI** (wait until brain is fully mature at ≈ 18 months) - to assess detailed ***cortical*** anatomy, ***myelination***.

MRS

- lesion has to be **≥ 1 cm**.
- MRS results may be distorted if there is hemorrhage (e.g. in tumor)
- three PEAKS representing:
  1) creatine (CR) - cellular energy metabolism; present in much higher concentrations in glia than in neurons.
  2) choline (CHO) - cell membranes; present in much higher concentrations in glia than in neurons.
     CHO↑ - abnormal membrane metabolism: myelin breakdown, inflammation, neoplasia.
  3) *N*-acetyl aspartate (NAA) is neuronal marker - found primarily within neurons and precursor cells; NAA is marker of neuronal integrity.
     NAA↓ - neuron loss.
- additional peaks (not detectable in MRS of normal brain):
  4) lactate – products of anaerobic glycolysis: inflammation, infarction, abscess.
  5) lipids – products of brain destruction: radiation necrosis

Tumor – lots of membranes (choline) and anaerobic metabolism (lactate)
Necrosis – everything is down except dead lipids↑
Stroke – everything is down except anaerobic metabolism (lactate)↑
Abscess – atypical peaks
MS - normal

ULTRASOUND

Higher beam frequency - better axial resolution, but less tissue penetration.

Sonographic "window" - ANTERIOR FONTANELLE also other fontanelles

- sensitivity for hypoxic–ischemic lesions is poor (normal sonogram does *not* exclude this pathology)

CSF

- CSF is inside BBB.
- CSF freely communicates with brain interstitial fluid.
- BUOYANCY reduces in situ weight of brain to ≈ 50 gm.
- CSF removal during lumbar puncture → brain weight↑ → tension on arachnoid trabeculae, nerve roots and blood vessels → headache.

- CHOROID PLEXUS is present in:
  1) ***4th ventricle ependymal roof*** – blood supply from *PICA*.

2) ***3rd ventricle ependymal roof*** – blood supply from *branches of PCA*.
3) ***medial wall of lateral ventricles*** – main mass! – blood supply from *anterior and posterior choroidal arteries*.

- **TOTAL CSF VOLUME** 150 ml; 7.5-70.5 ml in ventricles.
- **CSF PRODUCTION RATE** ≈ 500 ml/d = 20 ml/hr. CSF production is independent to ICP
- CSF volume removed at lumbar puncture is regenerated in 1 hour.
- ACETAZOLAMIDE (carbonic anhydrase inhibitor) can reduce CSF production significantly.
- ***adrenergic*** stimulation → diminished CSF production (e.g. norepinephrine)
- ***cholinergic*** stimulation may double normal CSF production rate.
- ARACHNOID VILLI – serve as ***one-way valve*** – prevent blood reflux into CSF; open at Δp = 5 mmHg; CSF reabsorption ceases at ICP < 5 mmHg

CSF production is ***independent*** of ICP;
CSF absorption is ***proportionate*** to ICP and dural venous sinus pressure (CSF reabsorption is especially highly dependent on dural venous sinus pressure);

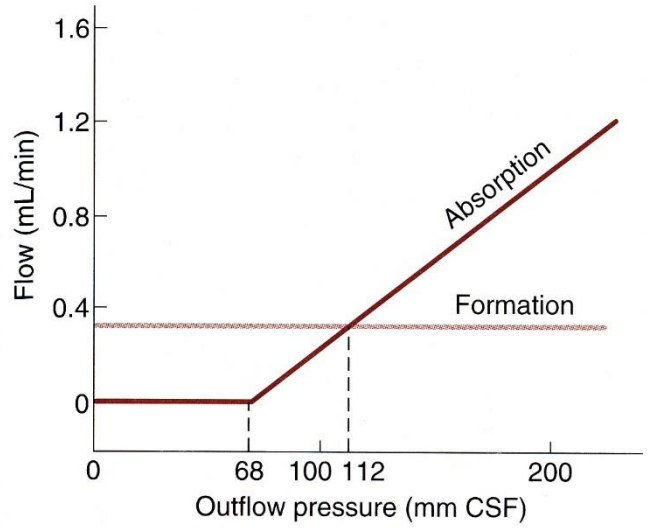


Figure 32–3. CSF formation and absorption in humans at various CSF pressures. Note that at 112 mm CSF, formation and absorption are equal, and at 68 mm CSF, absorption is zero.

NORMAL

1. **Color**

- **XANTHOCHROMIA** (literally, yellow color) = presence of any color.

Yellowish - ***increased protein*** (> 100-200 mg/dl).

Yellow / pink - ***hemoglobin***:

1) ***oxyhemoglobin*** (released with lysis of red cells).
   - disappears over next 7-14 days.
2) ***bilirubin*** (produced by leptomeningeal cells) is yellow.
   - may persist for 2-4 weeks.
3) ***methemoglobin*** (produced in old hematomas)

Yellow - severe jaundice, rifampin.

Brownish / gray - CNS melanoma.

Greenish - leukemic meningeal infiltration, pseudomonal meningitis.

2. **Opening pressure** 65-200 mmH_2O* (5-15 mmHg) with patient lying down (or at level of foramen magnum in sitting position).

*50 mmH_2O in neonates, 250 mmH_2O in extremely obese

Falsely elevated pressure:

1) marked obesity
2) tense patient (crying child)
3) head elevated above plane of needle

N.B. opening pressure is artificially elevated with patient in sitting position!

3. Few **cellular** components (≤ 5 lymphocytes or mononuclears / mm^3); *polymorphonuclear (PMN) cells* & *RBCs* are always abnormal (1 PMN is still normal if total cell count ≤ 5).

N.B. *normal newborn* - up to 19 lymphocytes/mm^3 (up to 60% cells may be PMNs);

N.B. many organic CNS diseases produce mild pleocytosis! (incl. seizures)

General rule:

> 100 WBC = ***infectious*** cause
< 100 WBC = ***noninfectious*** cause (carcinomatosis, sarcoid, etc)

Neutrophilic pleocytosis is indication for thorough **BACTERIOLOGIC INVESTIGATION**.

- cytopathological identification requires *large CSF volumes* (> 20 ml).

N.B. initial tap may be negative → serial LPs

At least 3 negative cytologic evaluations (i.e. 3 separate samplings) are required to rule out leptomeningeal malignancy!

4. **Protein** < 60 mg/dL*; mainly *albumin*.

*up to 150-170 mg/dL in neonates, esp. prematures (immature leaky BBB)

CSF albumin : serum albumin = 1:200

- majority of CSF protein (esp. **albumins**) is *derived from serum*.
- CSF proteins that *arise within intrathecal compartment*:
  1) **immunoglobulin G** (produced by CNS lymphocytes):
  2) **transthyretin** (produced by choroid plexus)
  3) structural proteins (**glial fibrillary acidic**, **tau**, **myelin basic protein**) found in brain tissue.
- CSF protein concentration increases from cephalad to caudal levels (different capillary permeability).

FROIN'S syndrome (s. loculation syndrome) – yellowish CSF *coagulates spontaneously* in few seconds – due to ***protein***↑↑↑; such CSF forms in loculated portions of subarachnoid space isolated from spinal fluid circulation by obstruction.

Intrathecal immunoglobulin synthesis (to support diagnosis of ***multiple sclerosis***) is determined by:

a) **IgG index** - intrathecal IgG synthesis rate↑ (vs. serum IgG that entered CNS passively across disrupted BBB):

$$\text{IgG index} = [IgG_{CSF} / albumin_{CSF}] / [IgG_{serum} / albumin_{serum}]$$

– normal IgG index is < 0.65-0.77.

b) **oligoclonal bands**; > 1 oligoclonal band in CSF (and absent in serum) is abnormal.

5. **Glucose** (> 60% of plasma amount, i.e. 50-100 mg/dl); values *< 50% (40-45 mg/dl)* are usually abnormal, and values ***< 40% (40 mg/dl)*** are invariably so (↑ ***anaerobic glycolysis*** + ↑ ***PMN leukocytes*** = CSF lactate also ↑)

- ratio *changes proportionately* in response to plasma glucose with 4-hour lag time (obtain concomitant serum glucose level at time of CSF sample!).

- if 50 ml ampule of 50% glucose has been given, 30 minutes is required to influence CSF glucose concentration.

6. **Ions**: *salt + Mg*
   a) concentration same or greater than in serum - **Na, Cl, Mg**.
   b) concentrations lower than in serum - K, Ca, bicarbonate, phosphate.

7. **Acid-base status**: *acidic due to CO2*
   - higher pCO_2 → slightly lower pH (than arterial blood).
   - bicarbonate levels are equal to arterial blood.

Common CSF studies:
1) direct observation for color, viscosity & turbidity.
2) cell count and differential
3) Gram's stain and culture
4) glucose
5) protein

If **cell count**, **protein**, and **glucose** are all normal, it is highly unlikely that additional studies will be useful.

BLOOD IN CSF

- bloody CSF should be collected in at least *three separate tubes* ("THREE-TUBE TEST").
- sample of bloody CSF should be *centrifuged* immediately (within 1 hour) and supernatant fluid *compared with tap water* (to exclude xanthochromia).

TRAUMATIC TAP
1) **CSF clears** as sequential amounts are collected (should be *confirmed by cell count* in first and last tubes);
2) **no xanthochromia**; causes of xanthochromia in traumatic tap:
   a) severely traumatic tap (RBC > 150,000-200,000/mm^3) - xanthochromia is due to serum *protein*.
   b) *oxyhemoglobin* - starts to appear if tube is tested > 1-2 hour after tap (RBCs lysis).
3) presence of **clot** in one of tubes strongly favors traumatic tap!
4) immediate repeat puncture at higher interspace yields clear CSF.

SAH
1) **CSF does not clear** with sequentially collected tubes;
   N.B. occasional declining cell count may represent layering of cells in recumbent patient!
2) **xanthochromia** (only if bleeding occurred before ≥ 2-4 hours); if ≥ 12 hours passed, virtually all patients' CSF will demonstrate xanthochromia!
3) blood **does not clot** (blood is defibrinated at site of hemorrhage).

Entered blood adds cells and protein to CSF - for every 1000 RBCs:
1) add 1 **WBC**
2) raise **protein** by 1 mg/dl.

Bacteriologic exam

Larger amounts of CSF (≥ 10 mL) improve chances of detecting pathogens (esp. tbc, fungi).

Gram stain is performed in ***all cases*** when CSF WBC count is elevated!

- *ongoing a/b therapy* – 25-33% positive tests are lost per day in setting of antimicrobial therapy (it does not significantly affect WBC counts, glucose, protein values)
- **antigen tests**:

Bacterial antigens persist in CSF for several days after antibiotic therapy.

- **PCR** – for viruses

LUMBAR PUNCTURE

Suspected meningitis overrides any contraindications - CSF examination is always indicated (imaging before LP is advisable); if suspected pressure > 350 mmH_20:

1) use 24G needle.
2) minimum required sample is obtained
3) administer IV bolus of MANNITOL 1 g/kg (ideally 20 min before LP)
4) DEXAMETHASONE (unless contraindicated).

Puncture above L_3 is absolutely contraindicated – in 1% people (esp. short) spinal cord ends at L_{2-3} interspace!
spinal needle with stylet (to avoid introducing epidermal cells→ iatrogenic epidermoid tumor): 20G 3.5-in. – for adults

Collect four tubes (extra tubes may be required for additional studies if indicated, e.g. cytology) – 5 ml each; N.B. if ***CSF is bloody***, cells are counted in both 1st and 4th (clearest) tubes

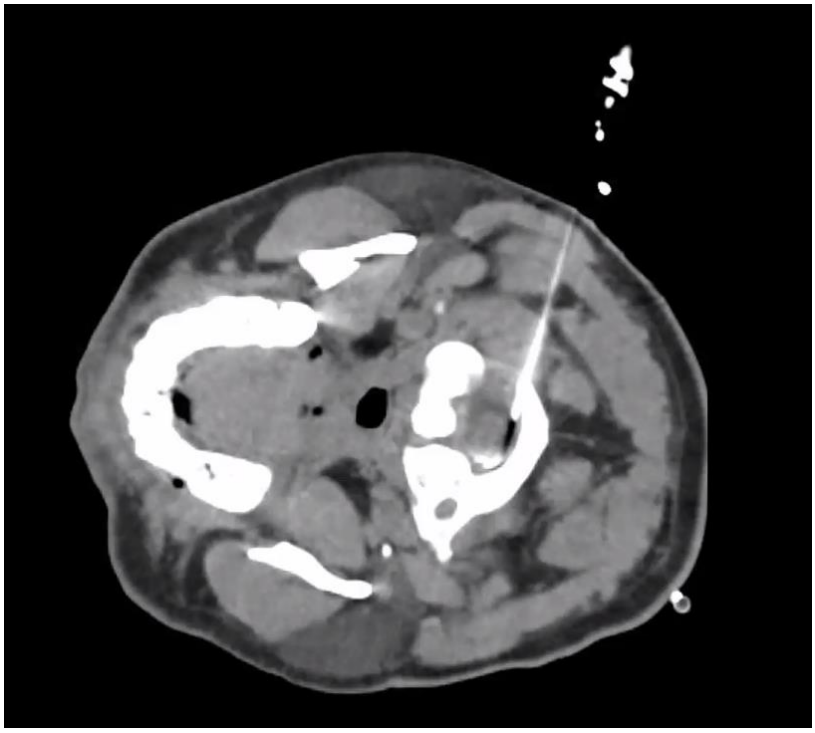
- routinely ***observe*** for cord / spinal nerve compression from developing hematoma after traumatic taps!

QUECKENSTEDT test (seldom performed today – replaced by myelography and MRI) - demonstrates **SPINAL BLOCK**:

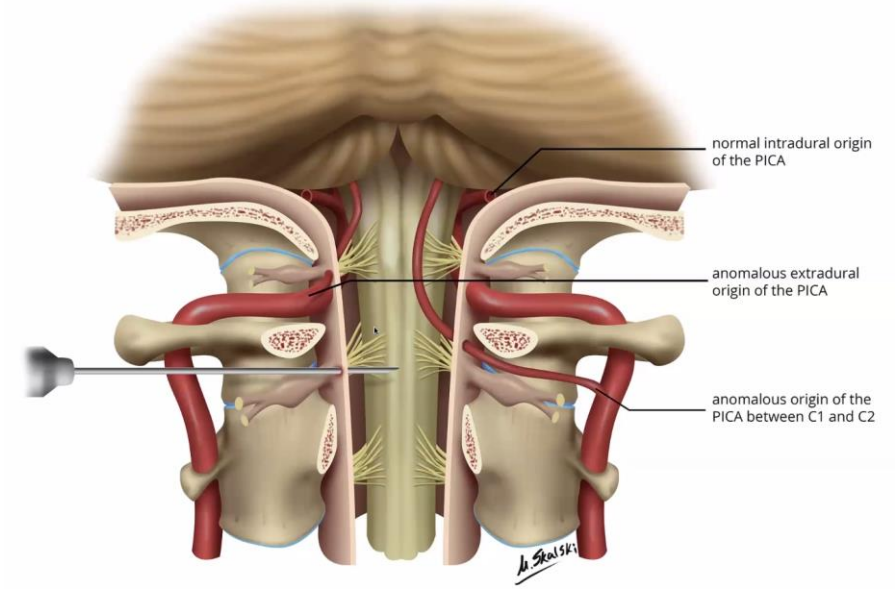
- patient in lateral recumbent position.
- 10-12 seconds of ***bilateral internal jugular vein compression*** (by assistant) → decreased venous return to heart → ICP rise → **CSF pressure**↑ in norm.

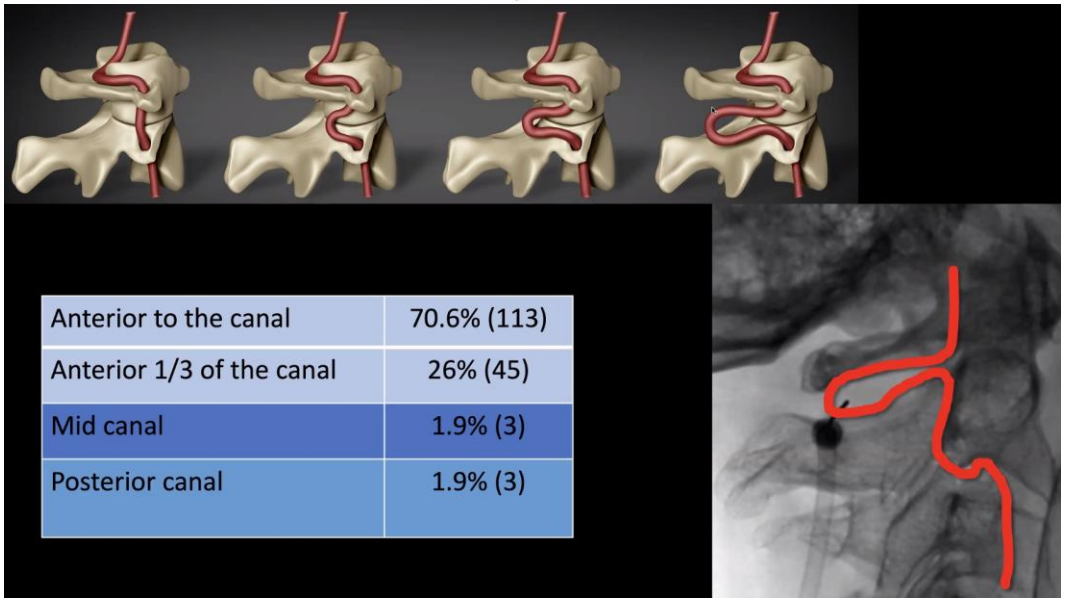
Herniation – **TRANSTENTORIAL** or **CEREBELLAR TONSILLAR**: *inject 5 mL of NS* and *withdraw needle immediately* → intubate & hyperventilate, mannitol, steroids.

Alternatives
Cisternal (s. suboccipital) Puncture - puncture to CISTERNA MAGNA *in midline halfway* between **C_2 spinous process** and **inferior occiput**
Lateral Cervical (C1-2) Puncture - puncture in C_{1-2} interspace from posterolateral approach – fluoro or CT guidance:



Beware of anomalous PICA or VA loop:





Anterior to the canal	70.6% (113)
Anterior 1/3 of the canal	26% (45)
Mid canal	1.9% (3)
Posterior canal	1.9% (3)

EVD

Frontal Approach – Kocher's point: 10-12 cm in sagittal line posteriorly from nasion (it should be 1-2 cm anterior to coronal suture) then 3-4 cm laterally – mark entry point here (coincide with sagittal plane going through midpupil) – aim 2 cm anterior to **tragus** and ipsilateral **nasion / glabella / medial canthus** → depth of 6 cm

Posterior (Occipital) Approach

Adults:

a) 4-5 fingerbreadths above and 4-5 posterior to auricle tip.

b) Dandy's point: 3 cm superior to inion and 3 cm off midline – higher risk of visual pathway damage.

c) Frazier burr hole: 6-7 cm superior to inion (might be too difficult to locate it) and 3-4 cm off midline - this places burr hole approximately 1 cm anterior to lambdoid suture and allows insertion of catheter down length of body of lateral ventricle.

Kids: 3 fingerbreadths above and 3 posterior to auricle tip.

aim at ipsilateral medial canthus + middle of forehead above eyebrows
with stylet to 6 cm, then soft pass to depth of 10 cm (8 cm for kids)

Transorbital approach

For herniating kids in ED: 18G 3.5 inch spinal needle – enter at the superior medial orbital roof corner → aim towards the opposite parietal bossing.

According to pathology drainage is set using external auditory meatus as reference point

Do not overdrain! Clamp EVD while positioning in OR (e.g. prone)

SAH (unsecured aneurysm) – drain at 10.
SAH (secured aneurysm) – drain at 0.
Trauma – "20 pop down to 10" – i.e. monitor ICP and, if ICP goes above 20, open and drain at 10 until ICP drops.
IVH – may drain at 0 to encourage CSF clearance.
posterior fossa mass – drain at 15-20 (some say in pediatric patients – 25).

CSF output relevance to challenging

When **< 100/8hr shift** – may start challenging (i.e. rising); don't challenge EVD until > 6-7 days post SAH

When **< 100/24hr** – may clamp → CT after 24 hours – if ventricles of normal size (can be slightly larger than when EVD was open), may D/C EVD.

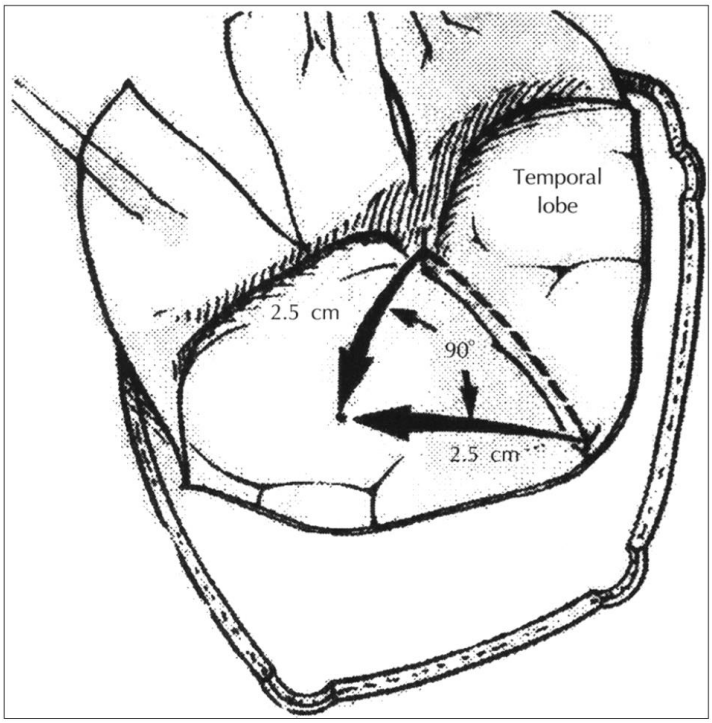
Dr. JRC challenges: 0 → 10 → 20

INTRAOPERATIVE ventriculostomy

PAINE's point (during pterional craniotomy) - creation of 2.5-cm isosceles right triangle:

hypotenuse (3.5 cm) is along sylvian fissure.
anterior limb starts on dura overlying sphenoid ridge (lateral orbital roof) and goes superiorly 2.5 cm;
posterior limb starts from sylvian fissure and goes anteriorly 2.5 cm;

- catheter enters frontal cortex perpendicularly at vertex of triangle:



CSF ACCESS DEVICES (IMPLANTABLE)

OMMAYA® reservoir
LEROY device.

- ensure that all catheter ***perforations lie within ventricular compartment***
  N.B. do not advance too far into 3rd ventricle (chemotherapy injection → severe nausea)
- reservoir can be used immediately;

Reservoir Puncture - **25G non-coring needle** at oblique angle.

EVOKED POTENTIALS

EVOKED POTENTIAL (EP) - electrical response *recorded* from CNS, *elicited* by external stimulus.

- may reveal abnormalities missed by MRI, and vice versa.
- changes produced by disease states:
  1) ***delayed responses*** - reflect conduction delays in responsible pathways.
  2) ***attenuation*** / ***loss of component waveforms*** – reflect conduction block or dysfunction of responsible generator.

SOMATOSENSORY EVOKED POTENTIALS (SSEP)

Stimulation of sensory systems leads to generation of **CORTICAL EVOKED POTENTIALS** - can be recorded with exploring electrode:

a) over scalp (surface electrode)
b) over pial surface of cortex
c) over cervical spine, Erb point (for median nerve stimulation at wrist)
d) over lumbar spine, popliteal fossa (for peroneal or posterior tibial nerve stimulation at ankle)

- not apparent in ordinary EEG; SSEP can be demonstrated by superimposing multiple traces - **signal averaging technique**
- 3-5 Hz ELECTRICAL STIMULATION of peripheral nerve - sufficient to produce ***slight muscle twitch*** (when mixed nerve is stimulated)
- response is small - necessary to average ***2000 responses in arm*** or ***4000 responses in leg***.

N.B. *physiological transmission* must be distinguished from *electrical conduction*!

MOTOR EVOKED POTENTIALS (MEP)

- stimulation of brain / spine elicits **motor evoked potentials** (i.e. compound muscle action potential over appropriate target muscle)

a) **Magnetic stimulation**
b) **Electrical stimulation** (painful in alert patients)

VISUAL EVOKED POTENTIALS (VEP)

- cortical activity (best recorded over midoccipital region) in response to ***monocular visual stimuli***:

- VEPs are useful in evaluating **anterior visual pathways**;

N.B. VEPs are *not useful in evaluating lesions posterior to optic chiasm*! (e.g. in cortical blindness, VEP may be normal!!!); retrochiasmatic lesions can be evaluated using *MONOCULAR HEMIFIELD STIMULATION*.

BAER

- stimulus - monaural click 65 dB above patient's hearing threshold. *audiometry is recommended before BAER!*

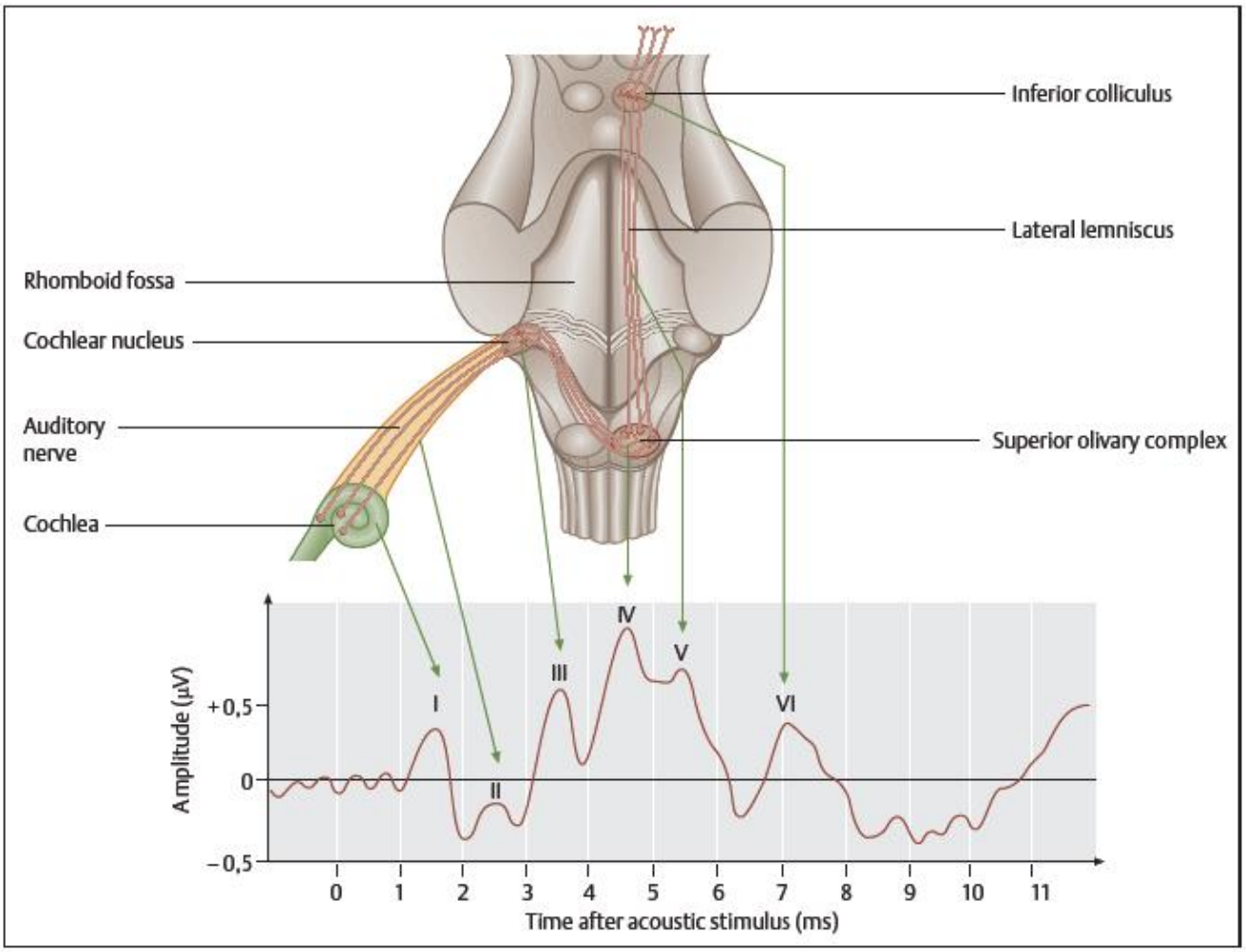
Series of up to seven components that occur within 10 msec of click stimulus:

- **wave I** – *cochlea*.
- **wave I** and early part of **wave II** – *auditory nerve* action potential.
- **wave II** - *cochlear nucleus*.
- **wave III** - *superior olive*.
- **wave IV** - *lateral lemniscus*.*
- **wave V** - *inferior colliculus*.
- **waves VI and VIII** are inconsistent and of *uncertain origin* - little clinical utility.

N.B. most consistent are **waves I, III, V** (CN8, superior olive, inferior colliculus).

*because **lateral lemniscus** contains *second order* neurons from cochlea and *third and fourth order* neurons from superior olive, it contributes to three waves.

Alarm: if BAER > 1 msec



BAER is normal in coma due to ***metabolic / toxic disorders*** or ***bihemispheric disease*** but abnormal in presence of ***brainstem pathology***

INTRAOPERATIVE MONITORING (IOM)

IOMs can reliably detect and predict neurological damage but there are 2 major problems:

a) it is ***too late*** (damage is done)
b) IOM is not useful if ***corrective action is not available*** (e.g. operating on spinal tumor – still have to resect tumor despite change in IOM signals; operating on brain AVM – still have to complete resection)

– monitoring *CN7 and CN8* (brain stem auditory evoked potentials) during posterior fossa surgery.
– monitoring *spinal cord* (SSEP, MEP) during scoliosis* / myelomeningocele (?***) / intramedullary tumor / degenerative** cervical spine surgery, repair of coarctation of aorta.

*reduces complications rate 10-fold (because effective corrective action exists if IOM signal changes – popping rod)
**most likely no benefit at all (studies show, IOM does not prevent complications)
***for kids < 4 years old, white matter long tracts are immature, cortex less excitable – EPs are unreliable.

- check baseline and then after positioning.

Practice guideline: Intraoperative electrophysiologic monitoring during surgery for cervical spondylotic myelopathy or radiculopathy

Use of intra-op EP monitoring during routine surgery for cervical myelopathy/radiculopathy is not recommended as indication to alter surgical plan or administer steroids since this paradigm has not been observed to reduce incidence of neurologic injury (***Level D Class III***)

SSEP

- use dorsal column pathway.

- frequent stimulation of bilateral **median / ulnar** or **posterior tibial** nerves → response measurement via contralateral cortical electrodes.
- averages signal over 50 seconds – gives warning too late
- what changes in SSEP should trigger concern:
    a) increased signal LATENCY (typically **> 10% prolongation**)
    b) decreased signal AMPLITUDE (typically **> 50% reduction**)
- ***irreversible*** – if fails to be returned to baseline before end of procedure.
- false-positive SSEP signal change may be caused by:
    1) **blood pressure** (mean and diastolic)
        – amplitudes of SSEPs are very sensitive to changes in ***mean arterial pressure***, making them useful for detecting ischemia!
    2) heart rate, temperature, partial pressure of alveolar carbon dioxide
    3) anesthetic drugs
        – all ***volatile anesthetics*** produce dose-dependent reduction in SSEP peak amplitude and increase in peak latency.
            - if inhalational anesthetic agents are required: use < 1 MAC (maximal allowable concentration), ideally < 0.5 MAC (i.e. inhalation anesthetics are kept at concentrations below 0.4%)
        – **PROPOFOL** has a mild effect on EP: total anesthesia with propofol causes less EP depression than inhalational agents at the same depth of anesthesia.
            - total intravenous anesthesia (TIVA) - propofol, fentanyl, and etomidate - is ideal; nitrous/narcotic technique is a second choice
        – ***hypocapnia*** (down to end tidal CO_2 = 21) causes minimal reduction in peak latencies
        – ***antiepileptic drugs*** (phenytoin, carbamazepine, phenobarbital) do not affect SSEP.

MEP

N.B. **MEP (motor evoked potentials)** is gold standard but are highly affected by anesthesia and muscle relaxation in particular

- checking MEP causes patient motion - need to pause surgery to run stimulation (bite block is necessary!) – gives warning to surgeon too late!

DIRECT (D)-WAVE

- epidural MEP (epidural electrode), resistant to anesthesia mishaps

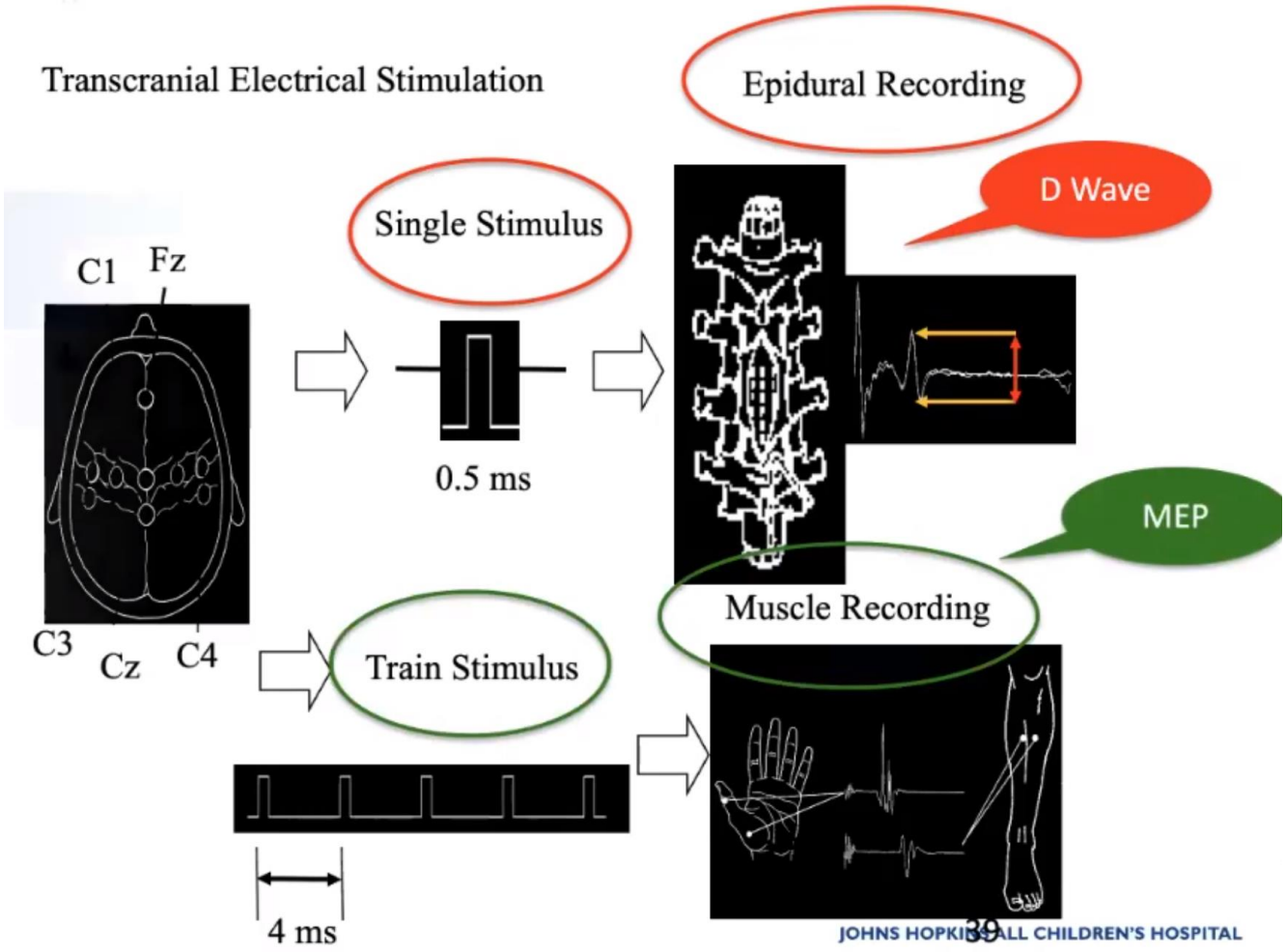
The direct (D) wave is a direct measure of the number of functioning fast-conducting fibers in the corticospinal tract (CST).

The use of D waves is limited in the cord up to T10–11 (fibers numerically decrease craniocaudally and are absent in the lumbosacral region)

In contrast to mMEPs and SSEPs, the **D wave is not in influenced by blood pressure, heart rate, temperature, and anesthesia drugs**

D wave monitoring needs midline recording.

A warning is a decrease of more than 50% of the baseline amplitude



Interpretation of D-wave:

D Wave	Muscle MEP		Expected Outcome
↓ < 50%	No Change	→	No Change
↓ < 50%	↓ Unilateral or Bilateral	→	Transient motor deficit
↓↓ > 50%	Bilateral loss	→	Prolonged or permanent motor deficit

SENSITIVITY, SPECIFICITY

– SSEP sensitivity 99%, specificity only 27%.
– MEP sensitivity 90-100%, specificity 90-100%.
– combined SSEP+MEP: sensitivity 83% and specificity 99%

In order of importance: MEP > SSEP latency > SSEP amplitude

STAGNARA TEST

– awakening patient during surgery (e.g. under remifentanil balanced anaesthesia) and performing neuro exam.

INTRAOP ALTERATIONS IN EVOKED POTENTIALS

a) increased SSEP LATENCY (**> 10% prolongation**)
b) decreased SSEP / MEP / D-wave AMPLITUDE (**> 50% reduction**)

Just legs or arms & legs?
If signals changed after positioning on the table – consider different table.

- check **mechanical factors**:
  - prompt cessation of dissection / manipulation (until potentials recover) - changes are mostly transient and are not predictive for postoperative neurologic outcome.
  - any retractors should be loosened; look at the entire operative field to verify that there are no impinging factors on the spinal cord.
  - reverse last maneuver, e.g. removal of any oversized graft.
  - perform further decompression if stenosis is present
- check ***electrodes***
- verify the **depth of anesthesia** - decrease gases to MAC (maximal allowable concentration) < 0.5 or switch to TIVA (propofol, fentanyl, and etomidate) [add KETAMINE], check TOF)
- check for presence of **hypotension** or **hypothermia** or **anemia**.
  - increase **blood pressure** (MAP > 85) - use an arterial line!, may press to MAP > 100
  - increase ***oxygen*** concentration
  - **transfuse** blood if needed.
- give additional **steroids**.
- cord **irrigated** with **warm** normal saline ± **papaverine**.
- consider CALCIUM CHANNEL BLOCKER (topical, IV)
- if nothing helps:
  a) do **Stagnara wake up test**

b) **terminate surgery** (consider expansile duraplasty + additional decompression to allow for cord swelling).
c) go for immediate **postop MRI** (but keep OR sterile – in case need to come back!)

EEG decline → reposition aneurysm clip, ICG angio

IF THE PATIENT EMERGES FROM SURGERY WITH A NEW NEUROLOGIC DEFICIT

a) immediately **reoperate** if the deficit is believed to be a result of hematoma or screw malpositioning.
b) proceed directly to **MRI or CT myelography** to determine the cause.

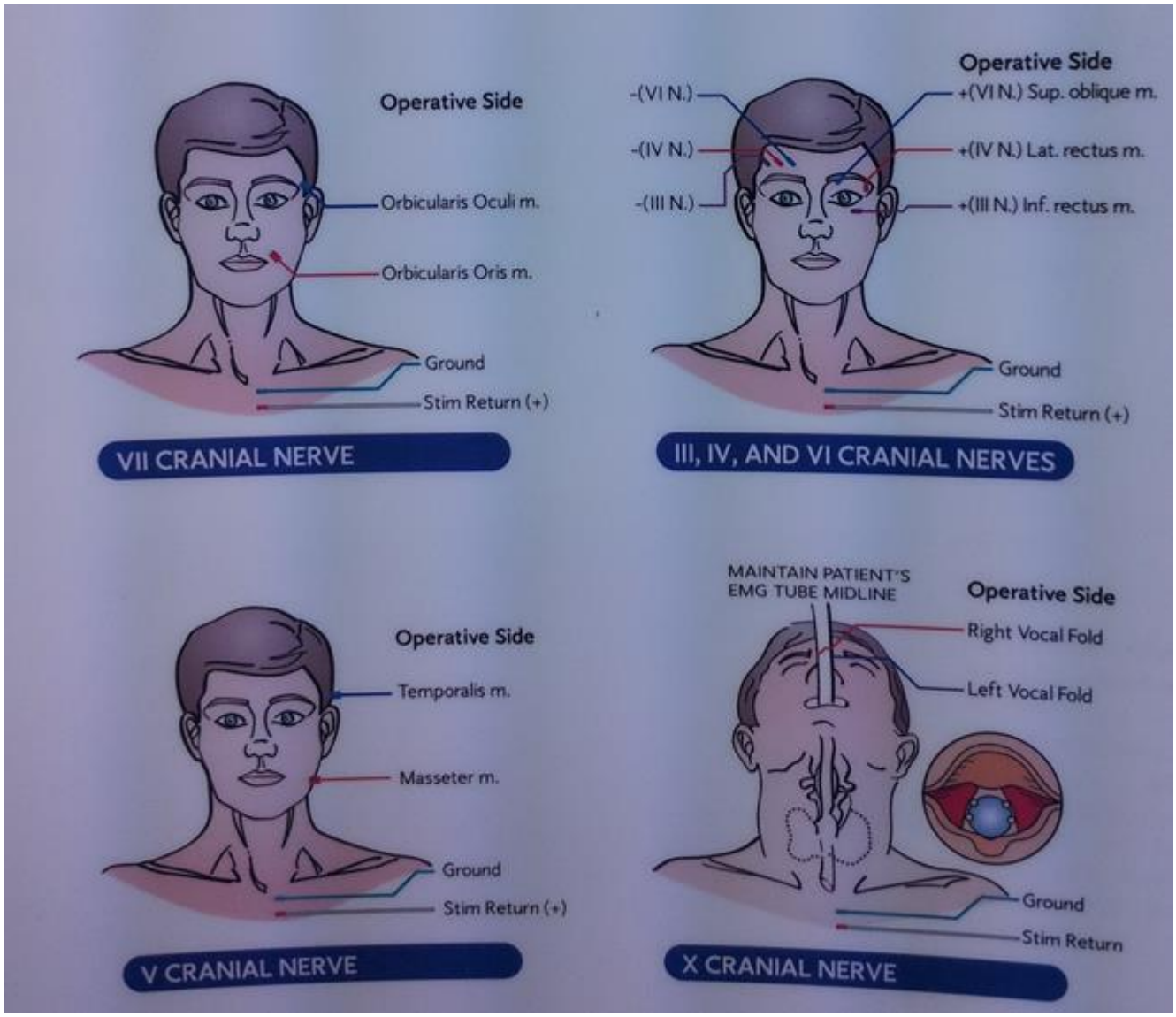
GU MONITORING

- for spinal cord tumor (esp. conus)

- **anus EMG + bulbocavernosus reflex** – only "lost or retained"

CRANIAL NERVE ION

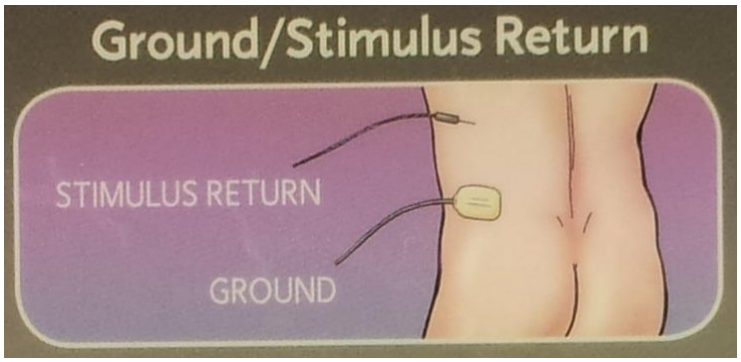
VEP – for CN2
BAER – for CN8



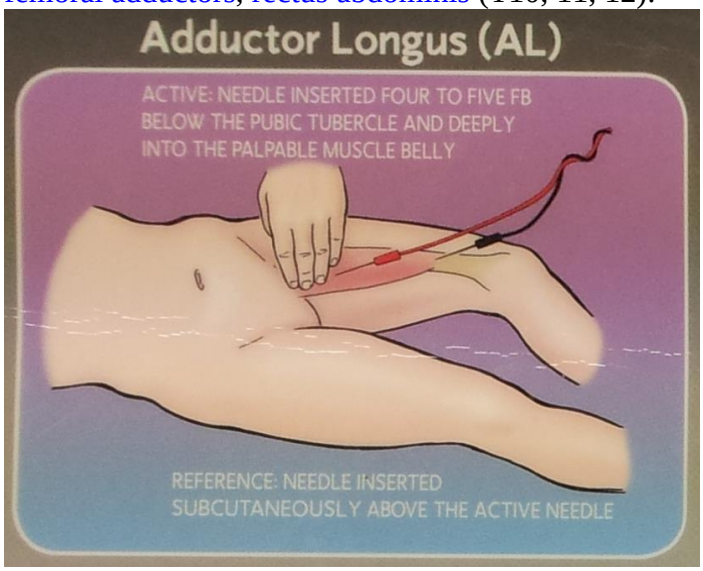
TRIGEMINAL EVOKED RESPONSES

- **infraorbital nerve** is stimulated with electrode inserted into infraorbital foramen.
- absence of waves 2 and 3 is after successful surgery for trigeminal neuralgia.

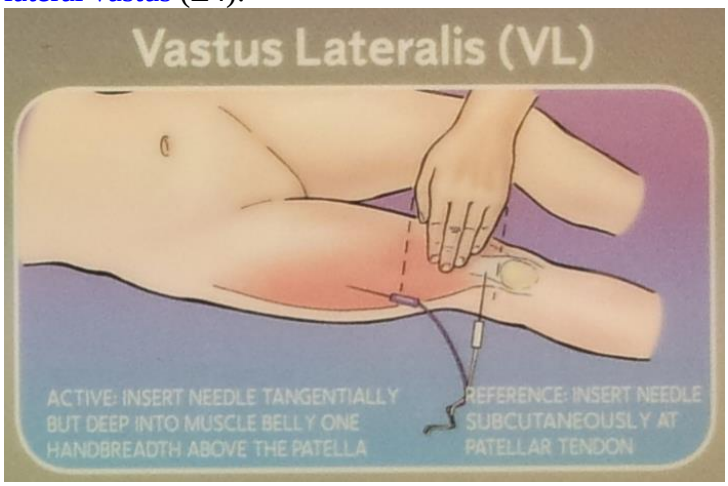
FREE RUNNING EMG (S. NIM, NEURAL INTEGRITY MONITORING)



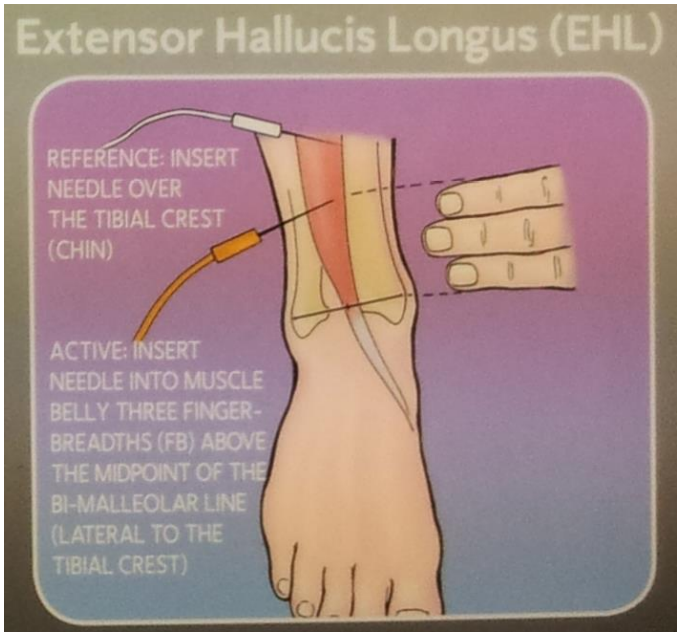
femoral adductors, rectus abdominis (T10, 11, 12):



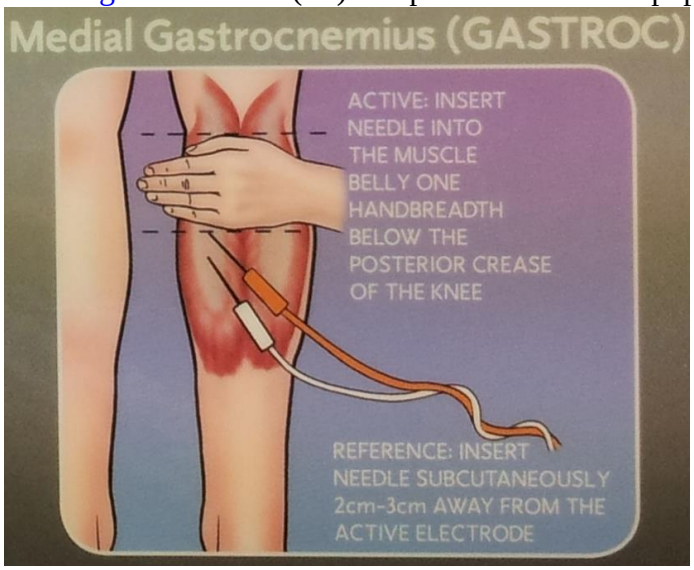
lateral vastus (L4):



extensor hallucis longus (L5) one palm-width above ankle, close to tibia:



medial gastrocnemius (S1) one palm-width below popliteal fossa:



OUTCOMES

GLASGOW OUTCOME SCALE (GOS) (Jennett, Bond in 1975)

5 – **GOOD RECOVERY** - normal life despite minor deficits

4 - **MODERATE DISABILITY** - disabled but independent; can work in sheltered setting

3 - **SEVERE DISABILITY** - conscious but disabled; dependent on others for daily support

2 - **VEGETATIVE**

1 - **DEAD**

GOS can be divided further into:

good outcomes (**independent**): 5 and 4
poor outcomes: 1-3

MODIFIED RANKIN SCALE (mRS) (1957 by *Rankin*; modified by *Lindley et al* in 1994) - degree of disability / dependence in daily activities:

0 - No symptoms.
1 - No significant disability. Able to carry out all usual activities, despite some symptoms.
2 - Slight disability (functionally still independent). Able to look after own affairs without assistance, but unable to carry out all previous activities.
3 - Moderate disability. Requires some help, but able to walk unassisted.
4 - Moderately severe disability. Unable to attend to own bodily needs without assistance, and unable to walk unassisted.
5 - Severe disability. Requires constant nursing care and attention, bedridden, incontinent.
6 - Dead.

mRS ≤ 2 means independent

MEMORY

Working memory – PREFRONTAL CORTEX
Short-term memory – PARAHIPPOCAMPAL GYRUS, HIPPOCAMPUS
Long-term memory – various parts of ASSOCIATION NEOCORTEX

AMNESIA - syndrome with **disturbance of RECENT MEMORY** (but not immediate memory!) – i.e. **disturbance of learning new information**.

ANTEROGRADE amnesia - inability to LEARN NEW information (i.e. acquire new declarative memories).
RETROGRADE amnesia - loss of ALREADY LEARNED information (i.e. memories acquired prior to amnesia onset).

AMNESTIC SYNDROME - disturbance of RECENT DECLARATIVE MEMORY (but not immediate memory!) (pvz. parodžius "7", po to liepus pakartoti, jis pasakys "7"; bet jei nors trumpam nukreipiamas dėmesys, pacientas skaičiaus nebeprisimena); **REMOTE memory** (previously learned information) **is more or less preserved;** amnesia by definition is **anterograde** amnesia, but retrograde loss is frequently present.

Transient global amnesia – idiopathic sudden severe anterograde amnesia (typically lasts for hours); ***total recovery is the rule***!
Factitious [psychogenic] amnesia - **RETROGRADE amnesia** (remote ≥ recent!); forget **identity & personal history*** (maximal for emotional crises), but remember public events; **no ANTEGRADE amnesia!**
*vs. *ORGANIC RETROGRADE AMNESIA* patients remember their identity (never disoriented to self!)

DEMENTIA, DELIRIUM

Organic brain syndrome - **global cognitive impairment states** whose unifying and defining feature is **CONFUSION**:

acute organic brain syndrome with clouding of consciousness = DELIRIUM
chronic organic brain syndrome with clear consciousness = DEMENTIA

DEMENTIA - ***acquired**** *chronic progressive encephalopathy* - chronic and substantial decline in ≥ 2 areas of cognition (i.e. AMNESIA + at least one of following: APHASIA, AGNOSIA, APRAXIA, EXECUTIVE FUNCTION DISTURBANCE**) - sufficient to interfere with previously successful daily activities.

* vs. ***mental retardation*** - ***developmental*** (present since early childhood).

N.B. **consciousness & perception are intact**!!!
Jei yra dementia, tai jau papildomai amnesia neberašoma!
**abstraction, judgment, complex problem solving, concept formation, planning, use of feedback to guide ongoing performance
Of all dementias, 20% are potentially reversible! Main role of **neuroimaging** is to exclude treatable causes

CORTICAL DEMENTIA SYNDROME – global declarative **memory loss** + elements of **aphasia**, **apraxia**, **agnosia, acalculia**.

SUBCORTICAL DEMENTIA SYNDROME: PD, Huntington, AIDS-dementia complex

1) movement disorders (e.g. **bradykinesia**)
2) slowed thought (**bradyphrenia**)
3) **disproportionate memory problems**
   - severely affected *working* memory, *reasoning*, *procedural* memory, and *strategic* memory (e.g. recall);
   - deficits in nondeclarative memory (vs. remain intact in cortical dementia).

Alzheimer's disease – slowly steadily progressive **CORTICAL DEMENTIA** with ***memory loss*** (earliest and cardinal clinical sign) → + ***aphasia*** (anomia, Wernicke), ***apraxia*** (difficulties with activities of daily living), ***agnosias*** (anosognosia, tendency to get lost), ***psychiatric symptoms*** (depression, paranoid delusions, hallucinations), ***sleep-wake cycle disturbances, incontinence;*** death ensues after 5-10 years

- generalized **CORTICAL ATROPHY**; ***hippocampal formation*** is involved early.
- MICROSCOPY - widespread cortical:
  1) intracellular **neurofibrillary tangles (NFT)** – "flames" (contain TAU protein)
  2) extracellular **neuritic (senile) plaques (NP)** (may also be found in normal aged persons!) - focal, spherical collections of dilated, tortuous axonal endings (***DYSTROPHIC NEURITES***) surrounding central ***AMYLOID CORE****
  3) **amyloid* angiopathy**

  *both contain amyloid beta peptide (Aβ); mutations in PRESENILINS ***increase Aβ production***
- one allele (**ε4**) of *ApoE gene* increases AD risk and lowers age at onset.
- ***neuronal loss in BASAL FOREBRAIN*** (esp. nucleus basalis of Meynert) → profound reductions in **acetylcholine** and **choline acetyltransferase**
- 5-10% cases are familial! – **AUTOSOMAL DOMINANT**
- most important risk factors: age↑, **ε4** allele
- treatment:
  1. Centrally acting ACETYLCHOLINESTERASE INHIBITORS: TACRINE (used rarely), DONEPEZIL, RIVASTIGMINE, GALANTAMINE
  2. NMDA antagonist MEMANTINE
  3. **ADUCANUMAB** (Aduhelm®) - FDA approved monoclonal antibody targeting amyloid ß (Aß) aggregates

Pick disease (frontotemporal dementia) - severe FRONTAL and TEMPORAL **lobar atrophy**

- surviving neurons are chromatolytic (**Pick cells**) and contain pathognomonic **Pick bodies** (cytoplasmic, round-oval, filamentous inclusions).
- dementia with frontal & temporal features - **abulia, apathy, language disturbances**; ***relative memory sparing***

Lewy body dementia – ***dementia*** + mild÷moderate ***parkinsonism***
Vascular dementia – abrupt onset with step-wise deterioration; focal signs; history of TIAs:

1. **Cortical syndrome** (***multi-infarct dementia***)
2. **Subcortical syndrome** in *état lacunaire* (***Binswanger disease***)

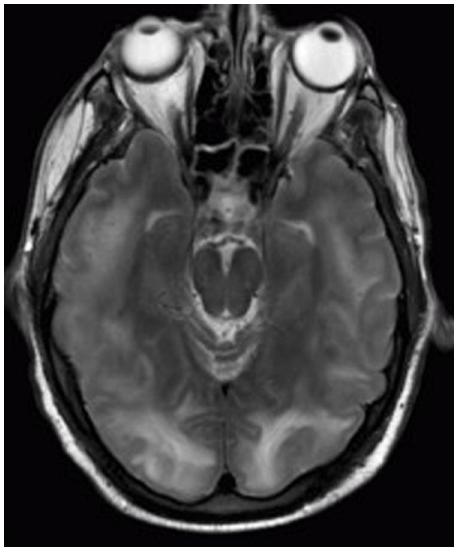
Normal pressure hydrocephalus – person > 60 yrs: dementia + incontinence + gait dyspraxia

Delirium (**toxic / metabolic encephalopathy**): changed level of consciousness + clouding of consciousness (confusion) → ***inability to maintain attention**** → global change in cognition; sympathic tonus↑; visual ± auditory hallucinations.

*main difference from dementia

- disproportionate (!) difficulty with immediate recall (e.g. of list of three items) suggests *DEPRESSION*, while difficulty with recalling items 5 min later suggests *DEMENTIA*
- normal EEG is incompatible with severe delirium - diffuse symmetric slowing of background EEG rhythm
- **Delirium is medical emergency** - nonpharmacologic approaches should be used; **HALOPERIDOL** is drug of choice; **BENZODIAZEPINES** are drugs of choice for ***withdrawal from alcohol or sedative hypnotics***

Posterior reversible encephalopathy syndrome (PRES)
headache, confusion, seizures and visual loss
etiology - malignant hypertension, eclampsia and some medical treatments (tacrolimus and cyclosporine) – treatment directed at etiology
MRI - areas of edema



APRAXIA

- acquired inability (of previously normal patient) to execute **skilled (s. learned) movements** - **association cortex**, dalyvaujančios in MOTOR PLANNING (**prefrontal cortex, posterior parietal cortex**) pažeidimas

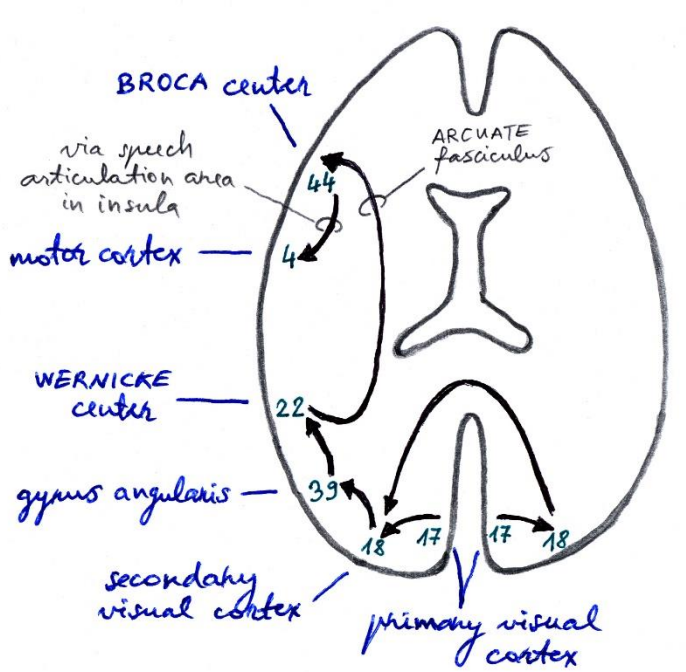
- ***lost motor templates for skilled movements***.
- APRAXIAS are body-movement equivalents of APHASIAS.

LANGUAGE DISORDER

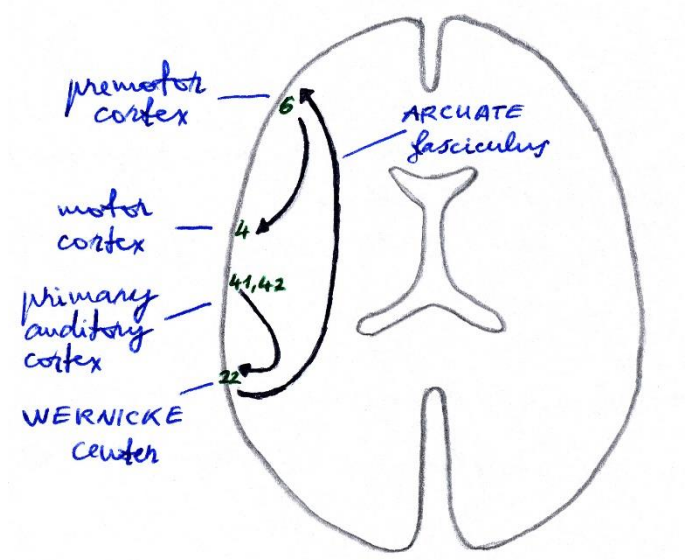
N.B. *SPEECH* is vocal expression of language.

WERNICKE-GESCHWIND model for language & gestures

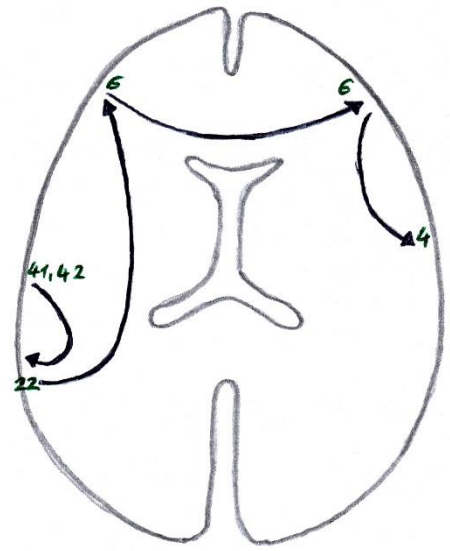
Person **names visual object**:



Person is asked to **raise right hand**:



Person is asked to **raise left hand**:



Aphasia:

1) *verbal fluency* (nonfluent - **motor aphasia**)
2) *comprehension* (impaired - **sensory aphasia**)
3) *repetition* (impaired - **conduction aphasia**)
4) *naming* (impaired - **anomic aphasia**)
5) *reading* (impaired - **alexia**)
6) *writing* (impaired - **agraphia**)
7) **aphemia** - disturbance in ***verbal output*** (BUCCOFACIAL APRAXIA) with preserved ***written language*** (i.e. nonfluent aphasia without agraphia)
8) **aprosody** – kalbos *emocinio atspalvio* sutrikimas
9) **auditory aphasia** (s. pure word deafness) – nesuvokia tik *girdimų* žodžių prasmės.

N.B. **NAMING, WRITING*** are impaired in ***all*** aphasias!
REPETITION impaired only in ***perisylvian*** aphasias

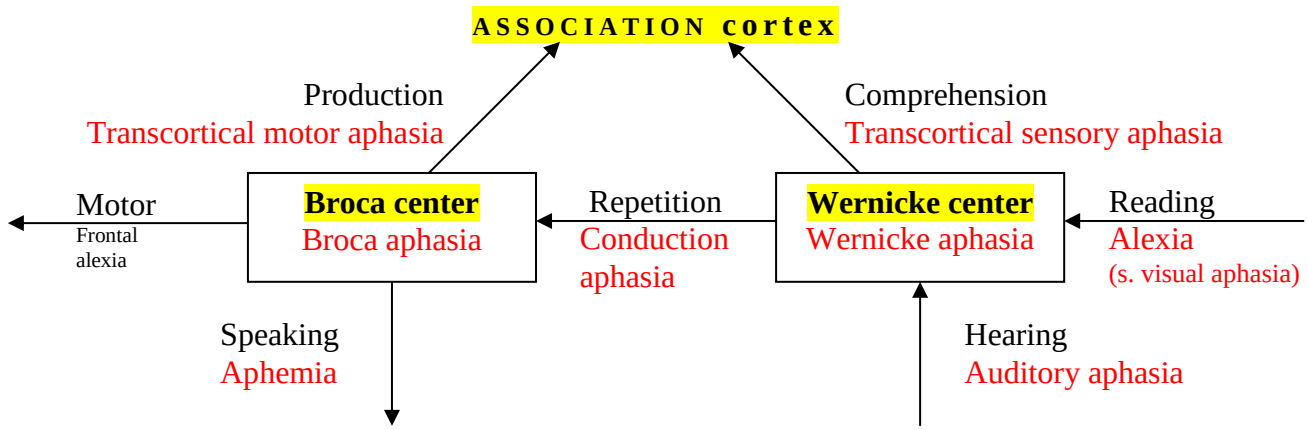
*writing errors typically parallel errors in spoken language

Transcortical aphasia - **left parasylvian area** is intact but **isolated** from rest of hemispheric cortex.

- dažniausia priežastis – ***infarction in watershed area***.
- **preserved REPETITION**!!!

APHASIA	Verbal output	Comprehension	Repetition	Naming	**Reading** aloud / comprehension	Associated signs
Broca	**nonfluent**	normal	impaired	marginally impaired	poor / good	RHP (esp. lower face)
Wernicke	fluent (paraphasic)	**impaired**	impaired	impaired	poor / poor	± RHH
Conduction	fluent (paraphasic)	normal	**impaired**	impaired (paraphasic)	poor / good	± RHS
Global	**nonfluent**	**impaired**	**impaired**	**impaired**	poor / poor	RHP, RHS, RHH
Anomic	fluent	normal	normal	**impaired**	variable	-
Transcortical:						
MOTOR	nonfluent	normal	**normal**	impaired	poor / good	RHP
SENSORY	fluent	impaired	**normal**	impaired	poor / poor	± RHH
MIXED	nonfluent	impaired	**normal**	impaired	poor / poor	RHP, RHS

Wernicke-Geschwind model of language and language disorders:



Speech-Related Disorders

1. **Vocal tics**
2. **Echolalia** - tendency to **repeat what has just been said by examiner**.
3. **Palilalia** - involuntary **repetition of words / phrases during verbal output**.
4. **Stuttering** - multiple rapid **iteration of uttered partial-word** (not whole word!)
5. **Logoclonia** - tendency to **repeat final syllable of word**.

SPEECH DISORDERS

N.B. *SPEECH* is vocal expression of language.

Mutism - total loss of speech.

DYSPHONIA

1. Vocal cord muscle paralysis / fatigability
2. Spasmodic dysphonia

DYSARTHRIA

1. Lower motor neuron (flaccid dysarthria)
2. Upper motor neuron (spastic dysarthria)
3. Cerebellar (ataxic dysarthria)
4. Extrapyramidal (hypo- / hyper-kinetic dysarthria)
5. Mixed dysarthria

SPEECH TESTING

CN7 – pronounce **LABIAL SOUNDS** – M, B, P. "Say 'baby hippopotamus'"
CN12 – pronounce **LINGUAL SOUNDS** – T, D, L. "Say 'yellow lorry'"
CN10 – ask patient to cough and to say 'Aaah' (**PALATAL SOUNDS**)

SLEEP (PHYSIOLOGY, DISORDERS)

PHYSIOLOGY

- **suprachiasmatic nucleus (SCN)** - internal circadian rhythm generator (pacemaker)!!!
- ***melatonin*** has phase-shifting properties opposite to bright light effects.
- ≈ 80% sleep time is **NREM sleep**; remaining ≈ 20% is **REM sleep**.

Stages 1+2 = light sleep
Stage 2 – **K complexes, SLEEP SPINDLES**

Stages 3+4 = deep sleep (rhythmic slow large waves)
Stage 3 – **theta rhythm**
Stage 4 – ***maximum slowing*** with ***largest waves*** (**delta rhythm**)

REM sleep - ***rapid, low-voltage EEG activity*** generated primarily in **pons tegmentum (RF)**!!!
(resembles waking state, but individual is clearly asleep, threshold for arousal is even elevated; muscle atonia in all but respiratory and ocular muscles; fluctuations in respiratory rate, heart rate, and BP; dreaming occurs).
Toward morning deep sleep↓ (stage 3 & 4 sleep↓) and REM sleep↑ (frequency and duration of REM episodes↑).

DISORDERS

insomnia - subjective sense that sleep is inadequate, insufficient, or interrupted (despite adequate opportunity for sleep) + ***impairment in daytime functioning***
N.B. excessive daytime sleepiness is rare in insomnia!

1) sleep-onset insomnia
2) sleep maintenance insomnia
3) sleep offset insomnia

rebound insomnia – due to withdrawal of sedatives.
adjustment sleep disorder - due to ***acute situational stress*** (e.g. new job, upcoming deadline, exam)
psychophysiological insomnia (most common primary insomnia) - patient is preoccupied with perceived inability to sleep - more patient tries to sleep, less successful attempts become; BEHAVIORAL THERAPY is mainstay

excessive daytime somnolence - impaired *performance* + **SLEEP ATTACKS (DOZINGS)**

1. **Narcolepsy** - incurable lifelong genetic disorder: involuntary **DAYTIME SLEEP EPISODES ± CATAPLEXY** (muscle atonia for seconds without altered consciousness; responds to tricyclic antidepressants) ± episodes of complete paralysis at beginning or at end of sleep, hypnagogic & hypnopompic hallucinations
   N.B. narcoleptics do not sleep more, but *need to sleep more frequently*!
   Dx: **multiple sleep latency test** - shortened latency, early REM

Rx: DEXTROAMPHETAMINE / METHYLPHENIDATE / MODAFINIL + rational scheduling of ***daytime naps***

2. **Sleep Apnea** – apnea > 10 sec, sukelianti hipoksiją (→ cardiac ischemia, pulmonary hypertension, stroke!) → awakening (gasping for breath); at least 5 episodes/hour (→ excessive daytime sleepiness!!!); Dx: **polysomnography**
   a) **OBSTRUCTIVE SLEEP APNEA** – in obese persons with loud snoring; Rx: **CPAP** by nasal mask
   b) **CENTRAL SLEEP APNEA** up to **ONDINE curse**; Rx: **BPAP** by nasal mask
3. **Periodic Limb Movement Disorder** - stereotyped periodic **LEG MOVEMENTS DURING SLEEP → sleep-maintenance insomnia**; if also **UNPLEASANT SENSATIONS IN LEGS** (urge to move, begin / worsen at rest, relieved by movement) - **Restless Legs Syndrome (→ sleep-onset insomnia)**; Rx: dopaminergics (ROPINIROLE)

parasomnias - undesirable behavioral (physical & mental) phenomena during sleep:
nightmares (during REM – vivid recall), sleep terrors (during NREM – no recall), sleepwalking (s. somnambulism), sleep talking (s. somniloquy), hypnic jerks (s. sleep starts), head / body rocking, nocturnal leg cramps, REM sleep behavioral disorder (DREAM-ENACTING BEHAVIOR – CLONAZEPAM universally effective!!!), sleep bruxism, sleep enuresis (normal before age 5-6 yrs; Rx: behavioral ± IMIPRAMINE / oral DESMOPRESSIN / OXYBUTYNIN)

NEUROCUTANEOUS DISORDERS (PHACOMATOSES)

PHACOMATOSES - heterogeneous genetic neurocutaneous disorders characterized by ECTODERM-based ***dysplasia***, ***hamartomas*** and ***neoplasia***.

- CNS, optic system, and skin are primarily involved – *both originate from ectoderm* (other organ systems can also be affected) – most correct term ***NEUROECTODERMAL DYSPLASIAS***.

AD INHERITANCE

Feature	NF1	NF2
Proportion	85-90%	10%
Gene - product	NF1 (17q11.2) – **NEUROFIBROMIN** **Constitutive Ras activation** → increased cell proliferation and survival.	NF2 (22q12) - **MERLIN**
Skin	*frequent cutaneous findings ("external NF")*: cafe-au-lait, axillary freckles	*relative paucity of cutaneous findings*
Tumor type	primarily *NEUROFIBROMAS*	primarily *SCHWANNOMAS*
Malignization	3-10% to MPNSTs	almost unheard
CNS	*lower incidence of CNS tumors* Optic nerve, brainstem, cerebellar gliomas! + unidentified bright objects (UBOs)	*higher incidence of CNS tumors* Bilateral CN8 schwannomas! Multiple meningiomas!
Eye	*Lisch nodules* in iris (90-95%)	*Posterior subcapsular (juvenile) cataracts*
Prognosis	better	worse

Tests: genetic testing → annual head MRI + ophthalmology + BEAR (in NF2)
- ***spinal MRI*** only for symptomatic cases.

NF 1 (VON RECKLINGHAUSEN'S DISEASE)

INCIDENCE - 1 in 3000 - one of most common autosomal dominant genetic disorders in humans!
- ½ cases appear sporadically (new mutations)

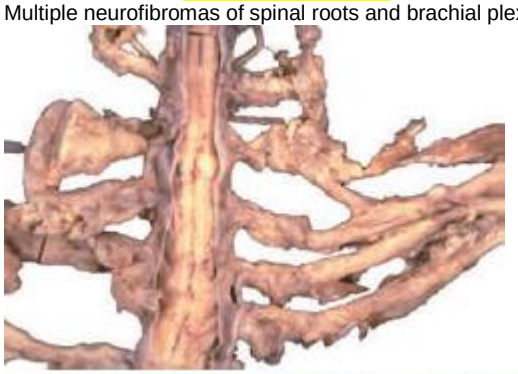
Mutation rate in NF1 gene (1 case per 10,000 population) is among highest known for any human gene!

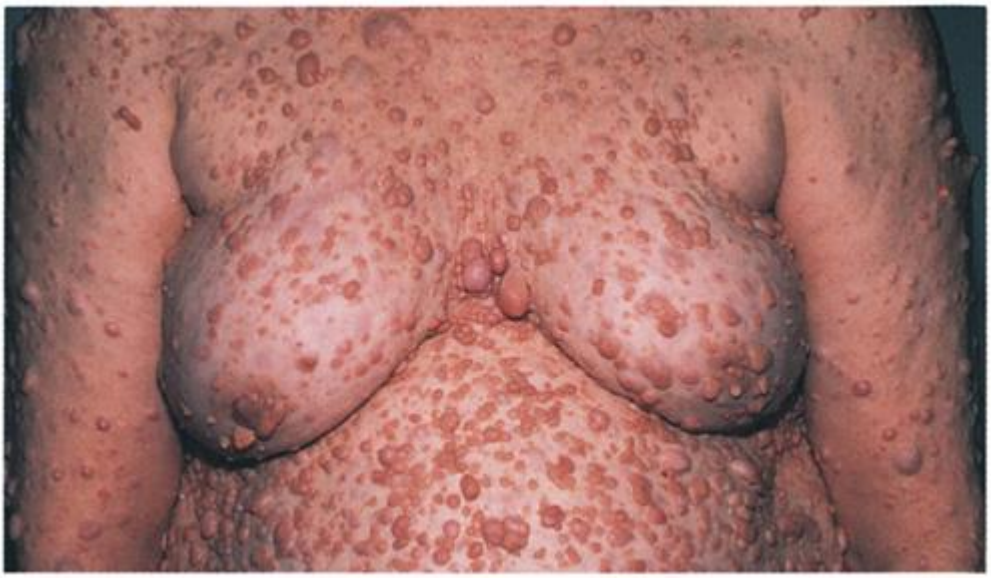
Many patients are functionally indistinguishable from normal!
Overall LIFE EXPECTANCY is reduced by 15 years – malignancy, hypertension, sequelae of spinal cord lesions.

Clinically – skin > CNS:

multiple skin *and any organ* ***NEUROFIBROMAS***

Multiple neurofibromas of spinal roots and brachial plexus in patient with NF1:



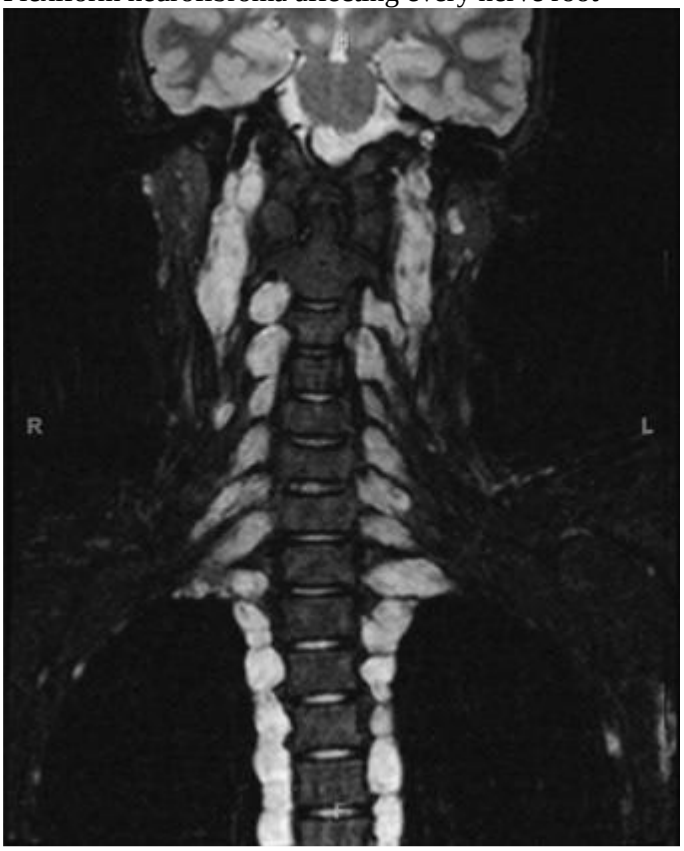


- may undergo *malignant degeneration* (esp. *PLEXIFORM NEUROFIBROMAS*) to malignant peripheral nerve sheath tumors (MPNSTs) – (lifetime risk 10%).
- management - **careful observation**; surgical intervention only for symptomatic cases (no routine spinal imaging because symptoms, not imaging characteristics, ultimately determine surgical management)
- ***PLEXIFORM NEUROFIBROMAS*** - almost pathognomonic of NF1: large and deep, invasive; extend across length of nerve and involve multiple nerve fascicles or multiple branches of large nerve → sizable ropelike mass of diffusely thickened nervous tissue
  - frequently involve nerve roots at multiple levels → extensive compression myelopathy; not amenable to cure (H: multilevel laminectomies* for debulking;

loss of neurological function is rare after surgery; regrowth → secondary reoperation)

*progressive kyphosis may require subsequent spinal fusion; most authors, do not advocate fusion at time of initial resection (rather use osteoplastic laminotomy)

Plexiform neurofibroma affecting every nerve root



multiple café-au-lait spots (pigment↑; ***earliest clinical finding***)

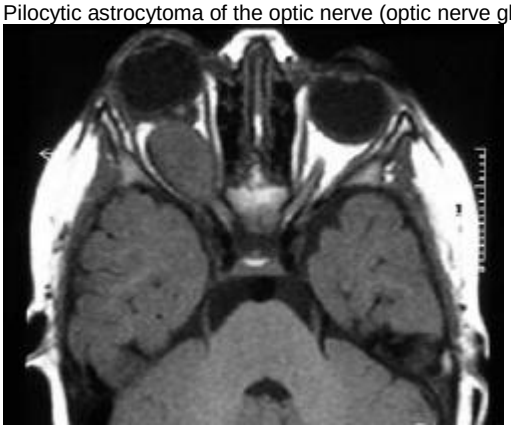
axillary or inguinal **freckles** (*Crowe sign*)

low-grade gliomas (esp. bilateral intraorbital CN2 pilocytic astrocytomas – tumors may regress*!!!, indolent brainstem gliomas**) – wait-and-see approach

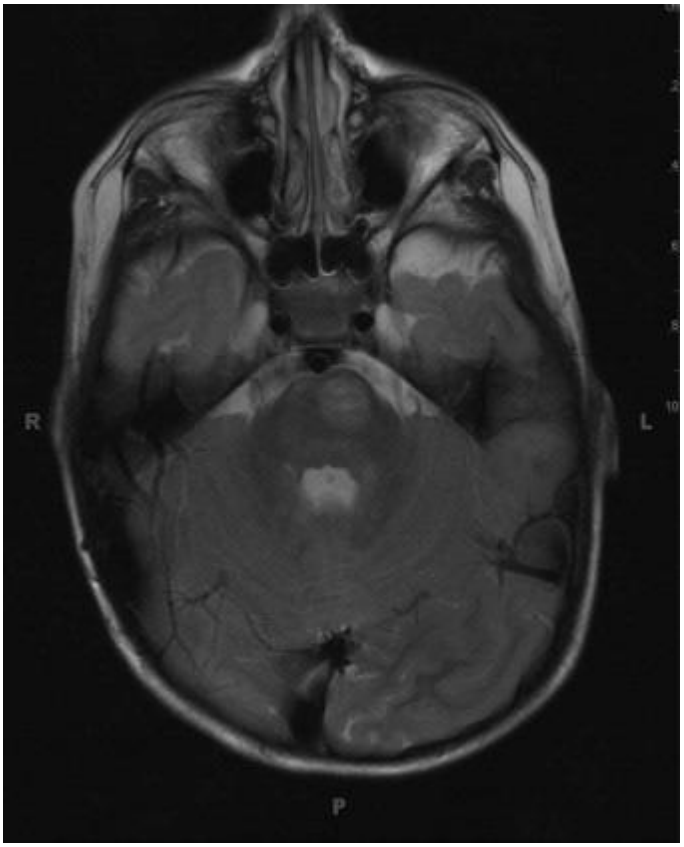
* **conservative monitoring** until:

a) **MRI progression / atypical appearance** → biopsy
b) **symptomatic** → debulking → adjuvant chemotherapy

Pilocytic astrocytoma of the optic nerve (optic nerve glioma):



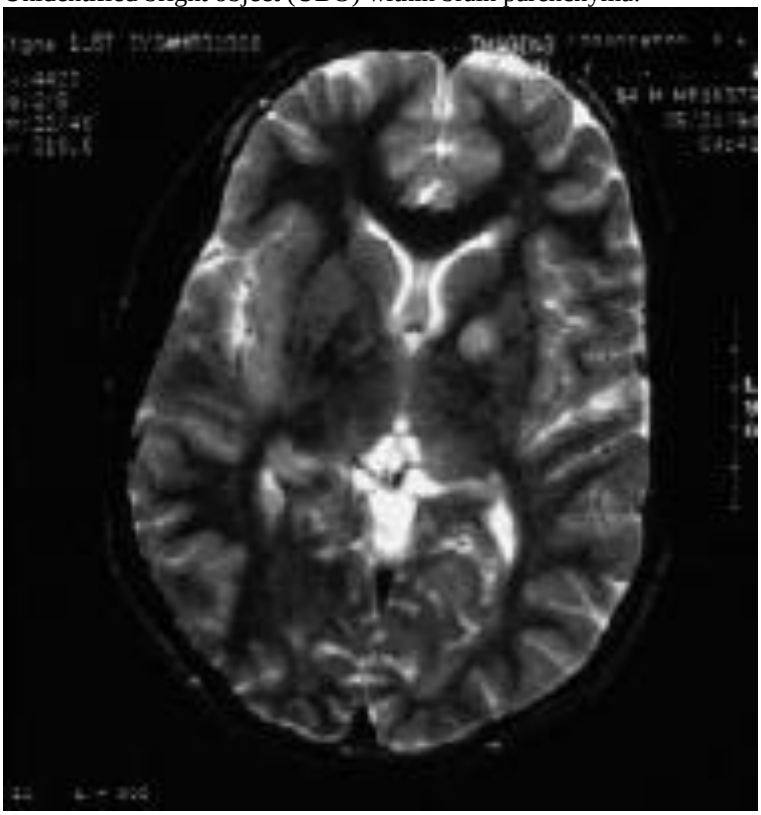
**management: serial MRIs → treat only clear progression.



- T2-MRI frequently detects unidentified bright objects (UBOs) – in cerebellar white matter, dentate nucleus, basal ganglia, periventricular white matter, optic nerve and pathways;
  - isointense on T1.
  - ***do not enhance, cause no mass effect*** (do not expand gyri, vs. tubers in tuberous sclerosis).
  - occur in > 50% NF1 patients.

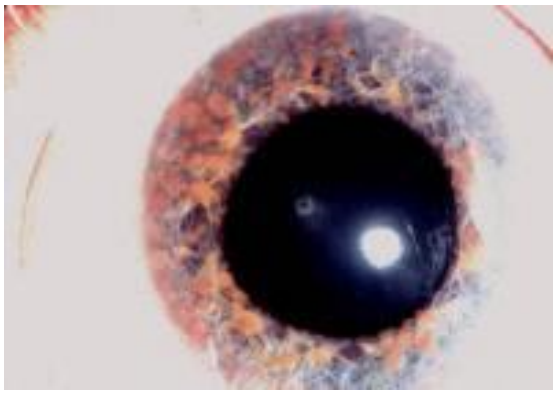
    UBOs ar the most common imaging findings in NF1 patients!
  - resolve as individual gets older (almost never seen at age > 20 years).

Unidentified bright object (UBO) within brain parenchyma:



bony dysplasia (sphenoid wing dysplasia/absence, bowing/thinning of long-bone cortex ± pseudarthrosis)

Lisch nodules in iris:



NF2

– skin <<< CNS:

- annual monitoring with ***head MRIs*** begins in teens → through late 50s.

SCHWANNOMAS - bilateral CN8 (> 95%, **hallmark of NF2**), any cranial and spinal nerve (skin, spine)
N.B. not neurofibromas!!!!

- patients > 60 years with bilateral CN8 masses are unlikely to have NF2 (NF2 usually presents before age of 40 years).
- management:
  Large tumors causing brainstem compression → resect microsurgically.
  Smaller asymptomatic tumors (or only audiologic symptoms) - natural history somewhat unclear (growth rates tend to decrease with increasing age); refer patient and family for education in sign language.
- *careful with SRS - has additional risks*!
  N.B. be aware of risk associated with stereotactic radiosurgery for treating benign lesion in patients genetically predisposed to malignancy
- ERLOTINIB - therapeutic activity for progressive vestibular schwannoma in NF2.

MENINGIOMAS (**second hallmark of NF2**) - multiple, high mitotic index, earlier in life.

SCHWANNOMATOSIS (NEURILEMOMATOSIS)

- multiple schwannomas (spinal, cutaneous and cranial nerve) without vestibular schwannomas or other manifestations of NF2 or NF1.

- associated with inactivation of NF2 gene in tumours but not in germline.

TUBEROUS SCLEROSIS (BOURNEVILLE'S DISEASE)

- variety of hamartomas that may affect ***every organ system***.

VOGT triad: ***Seizures***, ***Mental Retardation***, and ***Adenoma Sebaceum***

1. Brain: 30-50% patients have normal intelligence!

1) **hamartomas**: tubers, subependymal nodules ("candle-dripping") – both may enhance (do not necessarily imply malignant transformation!!!).

SENs → migration streaks in white matter → cortical tubers

TSC is one of the leading causes of genetic epilepsy!!! *infantile spasms* are characteristic; seizures may disappear in adult life

N.B. close relationship: onset of seizures at younger age → more severe mental retardation!; mental retardation rarely occurs without seizures, but intellect may be normal, despite seizures.

2) **grade 1 tumors**: subependymal giant cell astrocytoma (old name - SEGA);

- SGCT is continuum of SENs (histopathologically) - serial imaging of SENs is strongly recommended
  - any lesion exhibiting growth on serial imaging or causing hydrocephalus is called SGCT

SEGA exhibits growth vs. hamartomas (SENs, tubers) do not grow

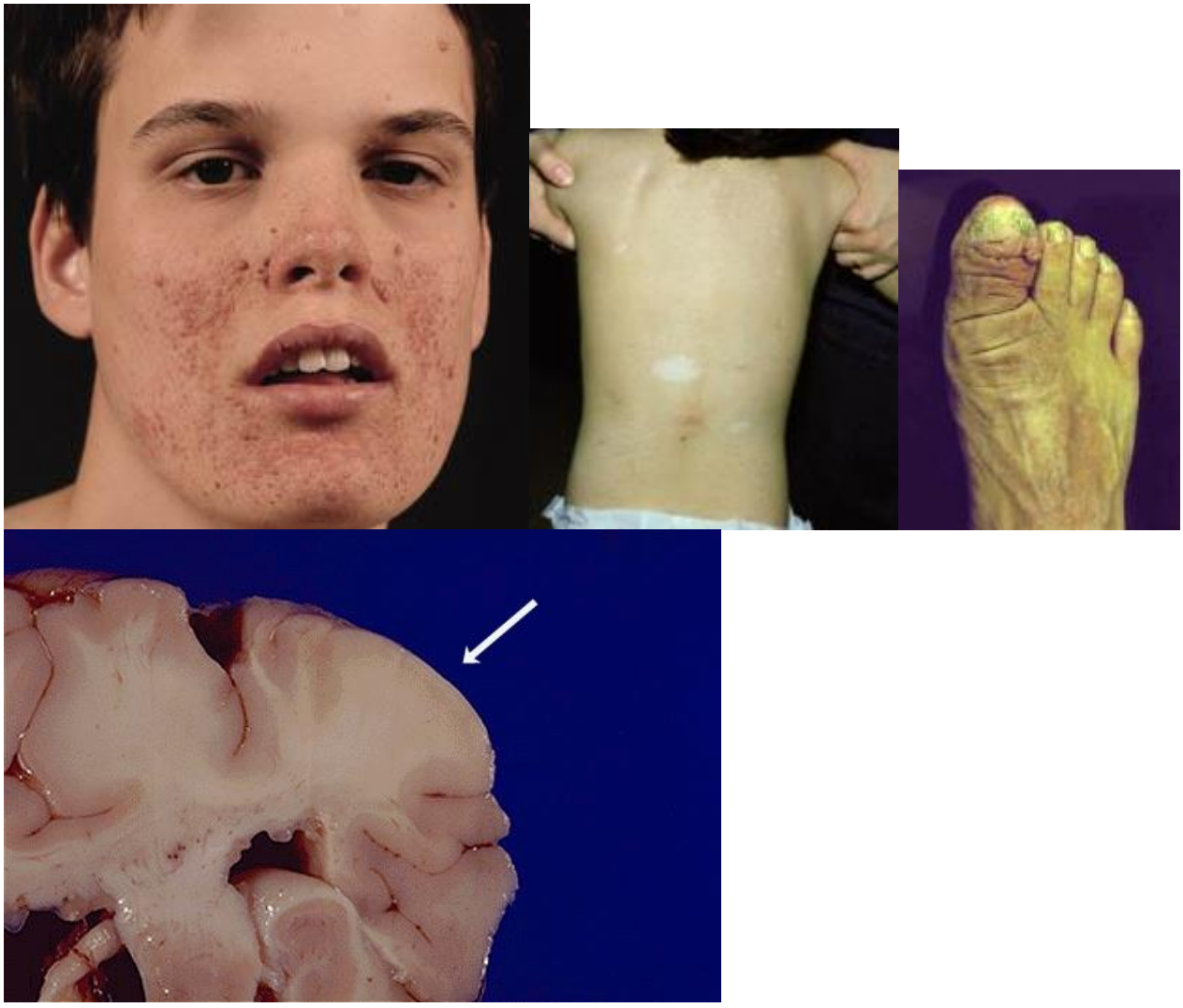
2. Skin - adenoma sebaceum in face (in ***butterfly distribution***), "ash leaf" (pigment↓) macules; fibromas (e.g. periungual)
3. Heart rhabdomyomas - spontaneous regression in first few years
4. Eyes - retinal hamartomas (**phakomas**)
5. Lung lymphangioleiomyomatosis;
6. Renal angiomyolipomata → life-threatening *retroperitoneal hemorrhage*.

a) 10-20% - TSC1 gene (9q34) - product (**HAMARTIN**) is tumor suppressor.

b) 80-90% - TSC2 gene (16p13) - product (**TUBERIN**) is tumor suppressor.

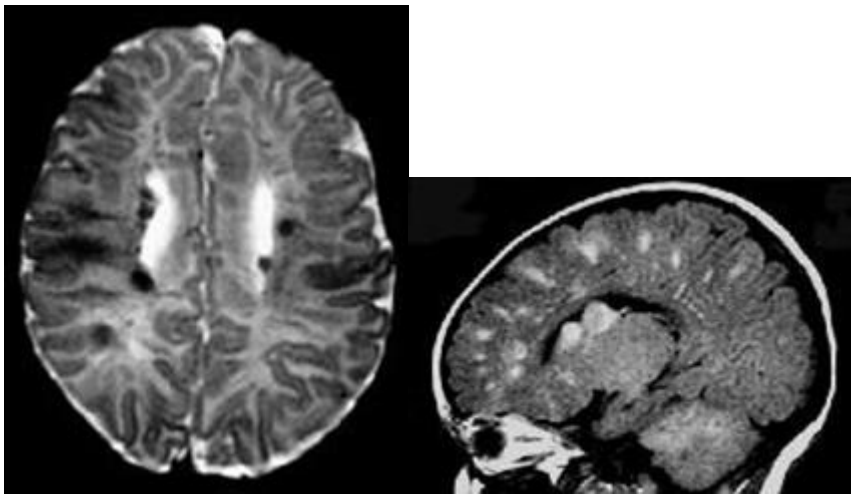
N.B. clinical-pathologic features caused by these different genes are indistinguishable!

- *hamartin and tuberin form tumor suppressor complex* that inhibits protein complex mTOR (mammalian target of rapamycin)

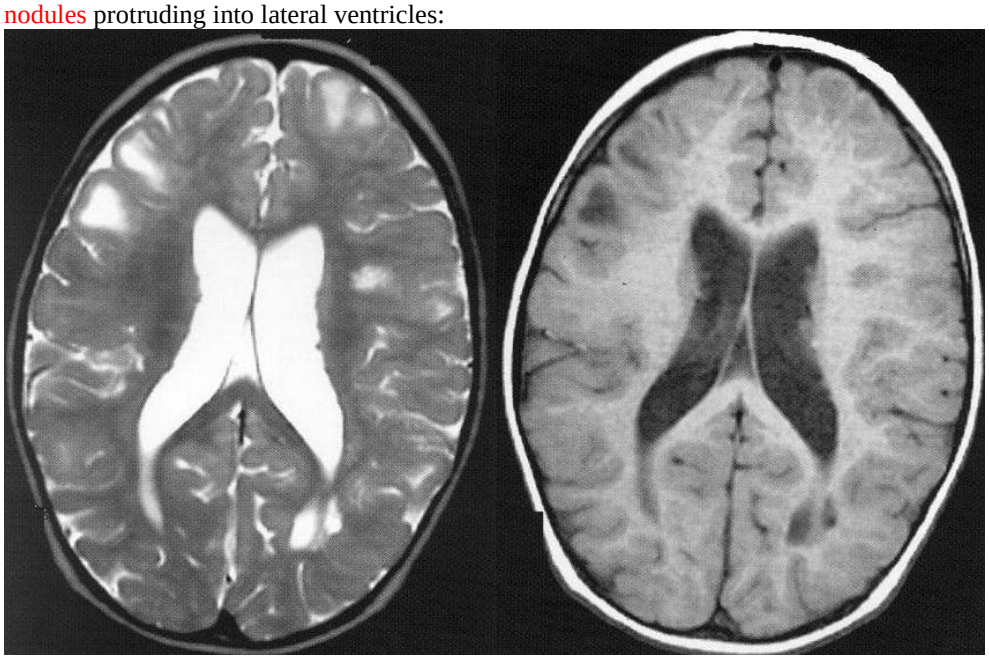


- **tubers** and **SENs** are:
  in neonates (unmyelinated white matter) - T1 hyperintense and T2 hypointense;
  older children - T1 hypointense and T2 hyperintense.

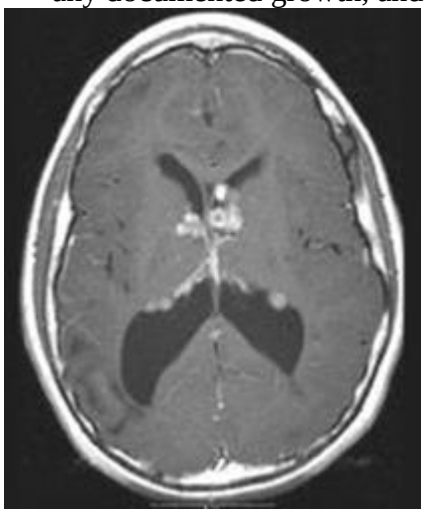
T2: T1:



(A) T2-MRI and (B) T1-MRI - multiple subcortical areas (tubers) with abnormal signal; small irregular subependymal nodules protruding into lateral ventricles:



- **SEGAs** – diagnostic criteria: tumor location near foramen of Monro, > 0.5 cm in diameter, with any documented growth, and intense gadolinium enhancement



mTOR inhibitors:

1. SIROLIMUS (S. RAPAMYCIN) - *SEGA* may regress!!!
2. TEMSIROLIMUS
3. EVEROLIMUS

Antiepileptics are mainstay of therapy for most patients (VIGABATRIN is first choice; FDA approved CANNABIDIOL).

Oncologic surgery

- no need to remove SGTC if it is asymptomatic - may stabilize or stop growing spontaneously after puberty!
- SGTC often large and difficult to resect by time they produce clinical symptoms, so lesions fulfilling criteria for SEGA should be removed as soon as growth has been confirmed (transcallosal or transcortical or endoscopic or LITT).

N.B. *early SEGA resection at first symptoms or documented growth* (modern approach)

Epileptic surgery - consider surgery immediately after failure of 2 medications! (no age restrictions – early surgery improves developmental trajectory); SEEG is indicated - even when multiple tubers are present, epileptogenic activity can often be localized to 1 or 2 tubers (*brain is loaded with TSC-mutated cells* – sometimes removing half of tuber helps, sometimes even lobectomy is not enough).

- tubers have more defined margin than focal cortical dysplasias.
- epileptogenic zone may include cortex surrounding tuber - **resection larger than tuberectomy (tuberectomy plus)**
- ***seizure often recur*** – palliative result (paradigm shift from cure to disease modification)
  N.B. prepare patient for the need of multiple surgeries! (incl. invasive monitoring)

Age-dependent SEGA monitoring: every 2 years before the age of 20 years; from the third decade onwards (SEGA is less frequent in adults and there is a lower potential for existing SEGA growth):

stable SEGA - no monitoring
growing SEGA - continued monitoring

PROTEUS SYNDROME

PROTEUS - Greek god who appeared in different forms

Macrocephaly, mental deficiency, seizures, **hemihypertrophy** (asymmetrical arms or legs), large flat feet ("moccasin feet"), thickened skin, hyperpigmented areas, hemangiomata and lipomata (subcutaneous and abdominal), bony defects, macrodactyly, hypocalcemia

SPORADIC

STURGE-WEBER SYNDROME

(encephalotrigeminal angiomatosis)

Gene	Main Features
unknown - congenital SPORADIC phacomatosis	ipsilateral capillary venous ANGIOMAS in leptomeninges, skin of face ("port-wine stain" s. nevus flammeus), eye (may lead to glaucoma → buphthalmos)

- absent superficial cortical veins are replaced by countless small cortical veins that become enlarged and calcified due to chronic venous hypertension.

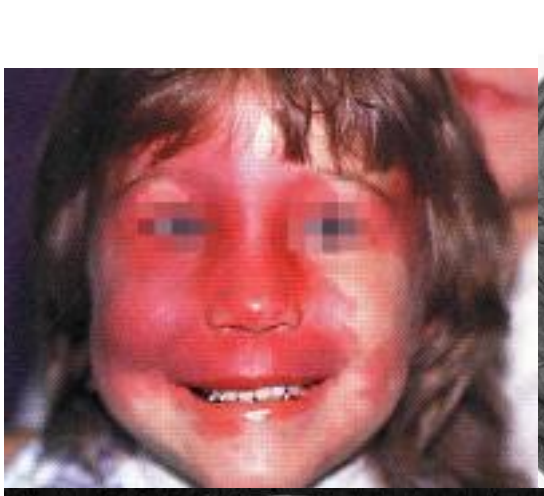
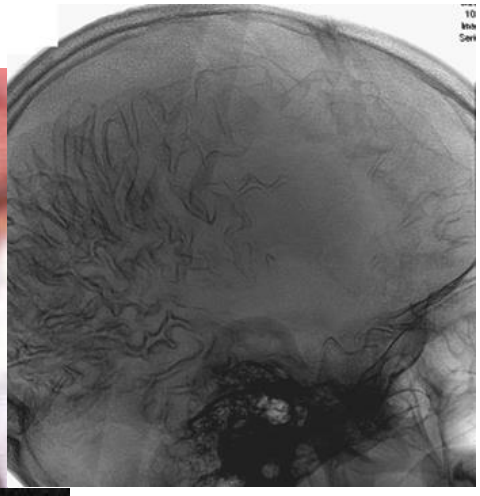
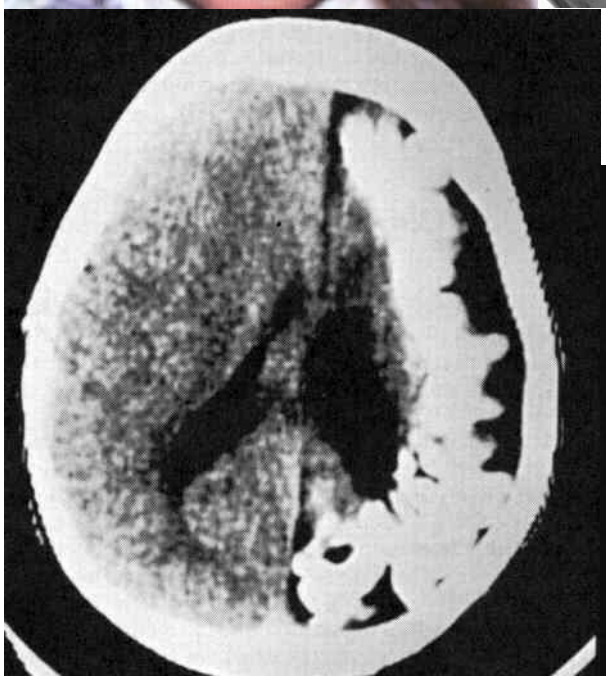
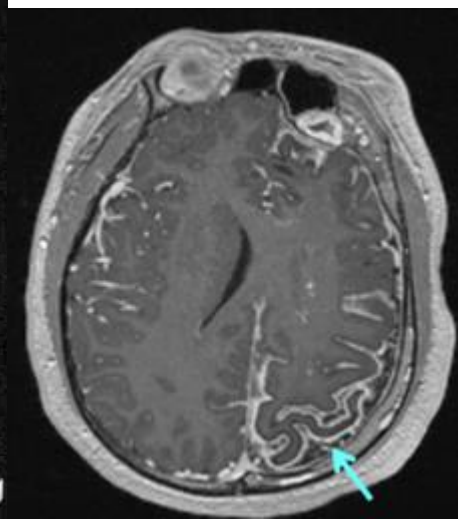
- developmental delay, vascular headaches
- *recurrent seizures* → progressive dystrophic **calcification** (in cerebral substance rather than in vessel walls), gliosis, brain atrophy → neurologic deterioration (focal deficits), seizures↑.
- CNS is not affected if port-wine stain does not involve V_1 area!

N.B. most patients with facial port-wine stains do not have SWS!

- **X-ray** – pathognomonic subcortical *"tram-track" calcifications* in gyriform pattern.
- **MRI:** atrophy + calcification + leptomeningeal thickening with gyral enhancement (appears as enhancement of subarachnoid space)!
- shrunken cerebral lobe with calcified cortex
- epilepsy surgery should not be delayed (ideally – during infancy)! - epileptogenic region is located in cortex adjacent to angioma → **focal cortical resection / hemispherectomy / corpus callosotomy / VNS**
- daily low-dose ASPIRIN - for headaches, stroke-like events (may be result of progressive venous thrombosis).

No increased propensity for cancer!!!

N.B. major intracranial hemorrhage is rare!

HYDROCEPHALUS

1) *dilation of cerebral ventricles* (at expense of periventricular white matter but with relative preservation of gray matter) – the only invariable sign!
    - temporal horns – first part to dilate > 2 mm!; in young children - occipital horns
    - transependymal flow phenomenon

- **Evans' index** ≥ 0.3
- role of suture status:
    a) ***open sutures*** - even moderate increases in pressure can *expand ventricles enormously* and surrounding brain may appear compressed to thin band (dramatic reconstitution of cerebral mantle may be seen after shunting, sometimes with reversion of some preexisting neurological deficits).
    b) ***fixed-volume skull*** - even enormously elevated intraventricular pressures may *only moderately expand ventricular size*.

2) *raised ICP*; active CSF secretion continues even though ICP increases; ICP frequently is normal in chronic hydrocephalus (e.g. NPH)
3) ***bulging fontanelles***, **"setting sun" sign**, ***increasing head circumference*** (!!!) with cranial *sutural diastasis*, "beaten-silver appearance" + no papilledema (infants)
4) CN6 palsy – long intracranial course
5) empty sella
6) seizures (infants)

LUMBAR PUNCTURE - perform only after imaging rules out obstructive hydrocephalus.

Any event resulting in RBCs in CSF may result in communicating hydrocephalus!
CSF protein > 500 mg/dl also may interfere with CSF absorption.

EXTERNAL HYDROCEPHALUS – CSF accumulation in **subarachnoid spaces** with *macrocrania* and *normal ÷ mildly dilated ventricular system*.
- caused by ***immaturity of CSF absorption system*** (at level of arachnoid villi)
- resolves in virtually every case.

SHUNTING

- parietal catheters are directed into frontal horn* by aiming toward ipsilateral medial eye canthus.
*ideally, use endoscope to guide catheter anterior to foramen of Monro

VENTRICULAR LOCULATIONS – use **multiple ventricular catheters** connected to single-valve system (to equalize pressures in various compartments and avoid dangerous brain shifts).

Torkildsen (s. internal) shunts – straight tubes that communicate ventricles to CSF spaces without valve (e.g. occipital horn with cisterna magna)

V-pleural shunt is a choice after VA shunt.

firm, noncompressible dome = *proximal and distal occlusion*.

Assessment of shunt – **palpation, CT (US for kids), XR, tap**
Assessment of ETV and Torkildsen shunt - **MRI flow studies**

COMPLICATIONS

In pediatric population, 90-day shunt complication rate leading to surgery is 16.9% (35% of those cases are preventable)

1. **Infection** - most feared complication (usually due to *Staphylococcus epidermidis & aureus*):
    - transient bacteremia has not been shown to cause shunt infection.

- 70% are diagnosed within first month after surgery and 90% within 6 months.
  N.B. chance of shunt infection after 6-9 months postop is almost zero!

  N.B. tap shunt only if other causes of fever are excluded!!! - 25G butterfly noncoring needle angle of puncture 20-30°

- treatment: **remove** infected shunt system + **abx*** (cefepime + vancomycin; may add rifampin especially if shunt is not removed) → place **external ventricular drain** to control CSF flow → **sterilize CSF***→ place **new shunt** (when CSF culture confirms eradicated infection)**.
  *antibiotics same as meningitis/ventriculitis; continue 7 – 14 days after hardware removal (usually after 2 CSF cultures are negative)
  **reshunt:
  S. aureus, GNR – 10-14 days of negative cultures.
  CoNS - may re-shunt in 3 days if no CSF abnormalities and follow-up cultures negative
  CoNS – if CSF abnormal and positive culture, may re-shunt in 7 days (10 days after last negative culture if follow up culture positive *after* shunt removal)

CNS Systematic Review and Evidence-Based Guidelines on the Treatment of Pediatric Hydrocephalus:
Level 2 recommendation: partial (externalization) or complete shunt hardware removal is an option in the management of CSF shunt infection.
Insufficient evidence to recommend either shunt externalization or complete shunt removal as a preferred surgical strategy.
Insufficient evidence to recommend the combination of intrathecal + systemic antibiotics when infected shunt hardware cannot be fully removed or must be removed and immediately replaced.

2. **Shunt failure** - most common complication!
   80% proximal, 10% valve, 10% distal
   Pediatric patient with shunt comes to ED with headache (and nothing else) → admit for observation!
   N.B. acute obstruction with deterioration is neurosurgical emergency; forced pumping may be attempted but provides only temporary relief in a minority of cases!
   N.B. if using occipital horn for catheterization, advance ventricular catheter > 10 cm to reach foramen of Monro (i.e. past choroid plexus); therefore, occipital approach is not recommended!
   Abdominal pseudocysts are synonymous with low-grade shunt infection
   Treatment: emergency high volume shunt tap → emergency shunt revision in OR.

3. **Slit ventricle syndrome**
   - mostly occurs after *ventriculitis* or *shunt infection* → subependymal gliosis → unusually low brain compliance ("unresponsive ventricles") - patient develops *high ICP without ventricular dilatation*.
     N.B. slit ventricle syndrome ≠ overdrainage; symptoms are those of high pressure rather than low pressure.
   - imaging findings falsely reassuring!
   - slit ventricles predispose to ventricular catheter failure (coapted ventricular wall) – how to diagnose:
     1) check for papilledema
     2) shuntogram / ShuntCheck
     3) place ICP monitor
   - **replacing shunt** in slit ventricles: Bugbee (to take out old catheter), navigation, Neuropen (to verify), consider valve with higher settings.

4. **Shuntodynia** – tenderness and pain along shunt.

5. **Hydrocele** (in boys).

VPS

CNS Systematic Review and Evidence-Based Guidelines on the Treatment of Pediatric Hydrocephalus (2021 Update):
Level 1 recommendation: **antibiotic-impregnated** shunt tubing reduces the risk of shunt infection (compared with conventional silicone hardware).
two antibiotics: **rifampicin and clindamycin**
Level 1 recommendation: There is insufficient evidence to demonstrate an advantage for one shunt hardware design over another in pediatric hydrocephalus.
Level 2 recommendation: There is insufficient evidence to recommend the use of a programmable versus a nonprogrammable valve for pediatric hydrocephalus.

Best valve:
NPH: Aesculap ProGAV + Pro ShuntAssist ← does not regulate flow, only prevents siphoning.

Small babies: Codman Certas with SiphonGuard ← regulates flow independent of position.

Bloody CSF (e.g. after SAH) – need the simplest design (most patients don't need shunt after several months): fixed medium pressure valve.

> **Target** – catheter tip above foramen of Monro (*choroid plexus* travels through foramen – may block catheter if it is placed inside foramen).

- for newborns, insert shunt via lateral corner of open anterior fontanel
- place redundant tubing into abdomen to allow for future child growth (may trim tubing in adults)

N.B. for difficult anatomy / revisions for malpositions – consider intraop verification (air ventriculogram, endoscope, or O-arm).

- may inject "shunt meds" into valve reservoir using 25G needle: 10 mg of VANCOMYCIN + 4 mg of TOBRAMYCIN solution.

N.B. if placing shunt because of unable to wean EVD, pull EVD at the end of OR but think twice – if patient was ***very sensitive to EVD clamping*** during wean trial (or if ***ventricles dilate significantly even at low level of EVD drainage***), may leave EVD catheter in and clamped and watch patient postop – EVD will work as a safety valve and can be pulled on POD1-2 after postop CT shows good position and no ventricle increase; may also think of installing Rickham reservoir at bur hole (will allow to aspirate CSF and flush with alteplase if system clots off).

- monitor children every 6-12 months:
  1) ***head growth*** in infants (occipitofrontal head circumference)
  2) detailed ***funduscopy***
  3) ***distal tubing length*** (with plain radiographs) when child grows.
  4) ***neuropsychological testing*** or ***developmental assessment*** (in younger children)

VAS

- consider preop cardioECHO.

N.B. any condition that causes ***significant elevation in central venous pressure*** can hinder the function of VA shunt.

fluoroscopic guidance - to prevent catheter thrombosis (short distal catheter) or cardiac arrhythmias (long distal catheter)

Open vascular cutdown technique into **transverse FACIAL VEIN** (esp. for small children – percutaneous technique is more difficult) - incision parallel to SCM medial border and 2 fingerbreadths below mandible (at the angle)

Percutaneous Seldinger technique into **INTERNAL JUGULAR*** as if placing central line – use US and standard central venous catheter kit: **20-22G needle** is used to puncture the skin 1-3 fingerbreadths (depending on the size of the patient) above the clavicle, between the heads of the sternocleidomastoid muscle - needle is inserted at an approximately 45° angle → **flexible J guide wire** is passed through the needle → needle is then removed, and a ***nick incision*** → load the **peel-away sheath** on the **dilator** and pass them together over guide wire

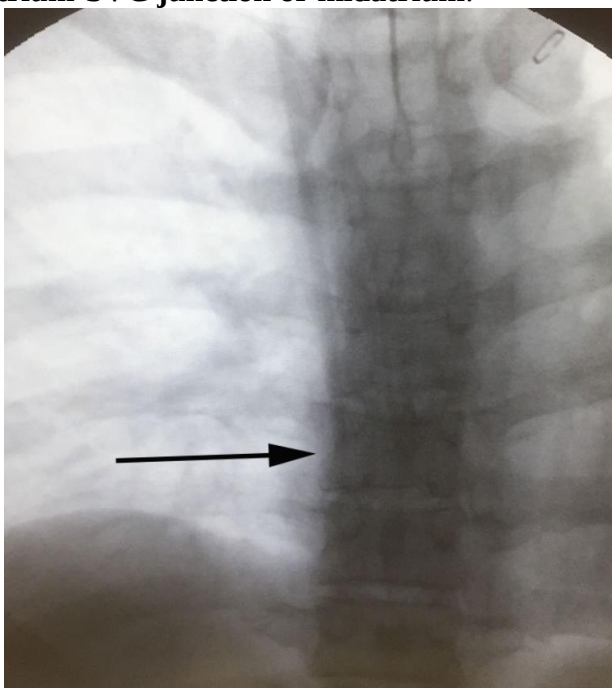
– tunnel distal catheter from cranium into neck and exit at guidewire skin entry site → flush entire shunt system with heparinized saline
– dilator and guide wire are then removed, and ***distal catheter is passed down the peel-away sheath*** into vein until it is at ***target position*** (pass the distal shunt well beyond the lower atrium before splitting and removing the peel-away sheath; it is not always possible to advance the shunt tubing once the sheath is removed; catheter is then pulled back to an appropriate final position, flushed with heparinized saline, and connected to the proximal shunt system).

N.B. heparinize catheter before placing it into vessel!

  - veins of most adults and larger children are large enough to accept a standard distal shunt catheter (outer diameter 2.2 mm - fits through a 7- or 9-French sheath – but test size of the sheath with the distal catheter before inserting it!).

*lateral or anterior to ICA, is compressible, and dilates with a Valsalva maneuver; placing in a head-down position often helps dilate and define the IJV

- ***target position*** - **atrium-SVC junction or midatrium**:



- **additional tubing** cannot be inserted to **allow for growth** – anticipate electively scheduled VAS lengthening procedures (educate patient parents about it!).
- complications are serious - renal failure (shunt nephritis), great vein thrombosis & pulmonary embolism, septicemia (shunt gets exposed at any bacteremia).

REMOVAL OF VAS

- if catheter is stuck, may leave in place but need to suture to pericranium or other tissues – to prevent distal intravascular migration.

VENTRICULO-GALLBLADDER SHUNT

- check **HIDA scan** to make sure bladder empties.
- purse string suture on fundus.
- cut catheter and use **straight connector** where catheter goes through bladder wall – prevents dislodgement (→ bile peritonitis).

VENTRICULO-SUBGALEAL SHUNT

- temporary measure until newborn reaches 2 kg for a permanent VPS.
- repeated trans-fontanel taps – too high risk of infection

ETV

INDICATIONS

Obstructive hydrocephalus
Persistent shunt infections (ETV allows to avoid hardware)

CONTRAINDICATIONS

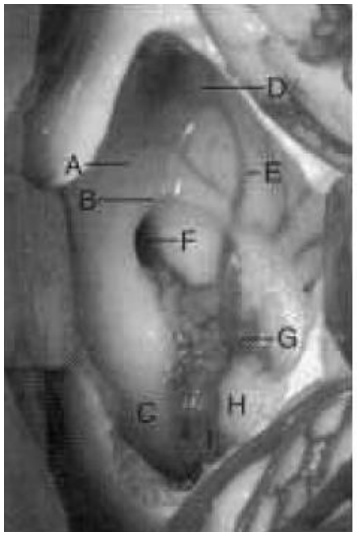
ETV may not work if patient has extensive metastatic deposits in subarachnoid spaces (absorptive capacity↓) H: regular VP shunt.
Low success rate of ETV – after previous shunt, WBRT

Check size of 3rd ventricle floor on volumetric MRI – if distance between dorsum sellae and basilar artery is too small, think twice if shunt is not safer option!

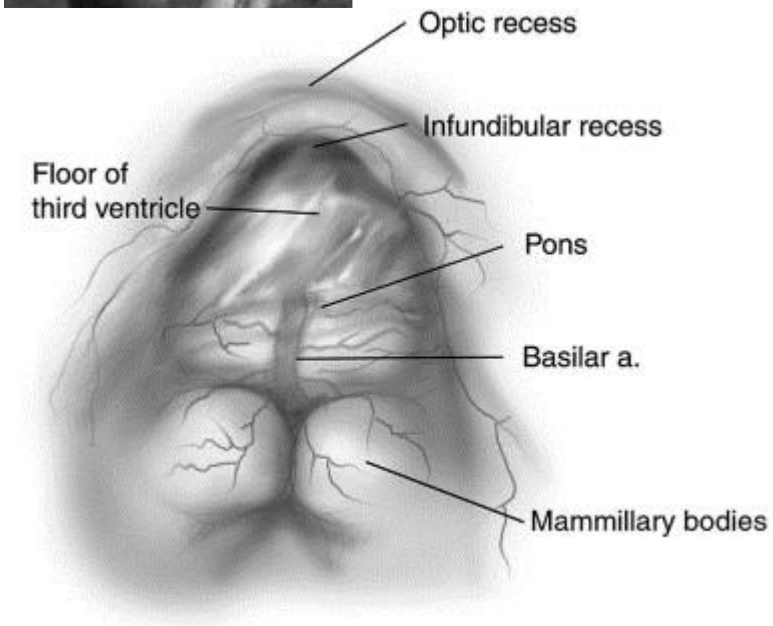
TECHNIQUE

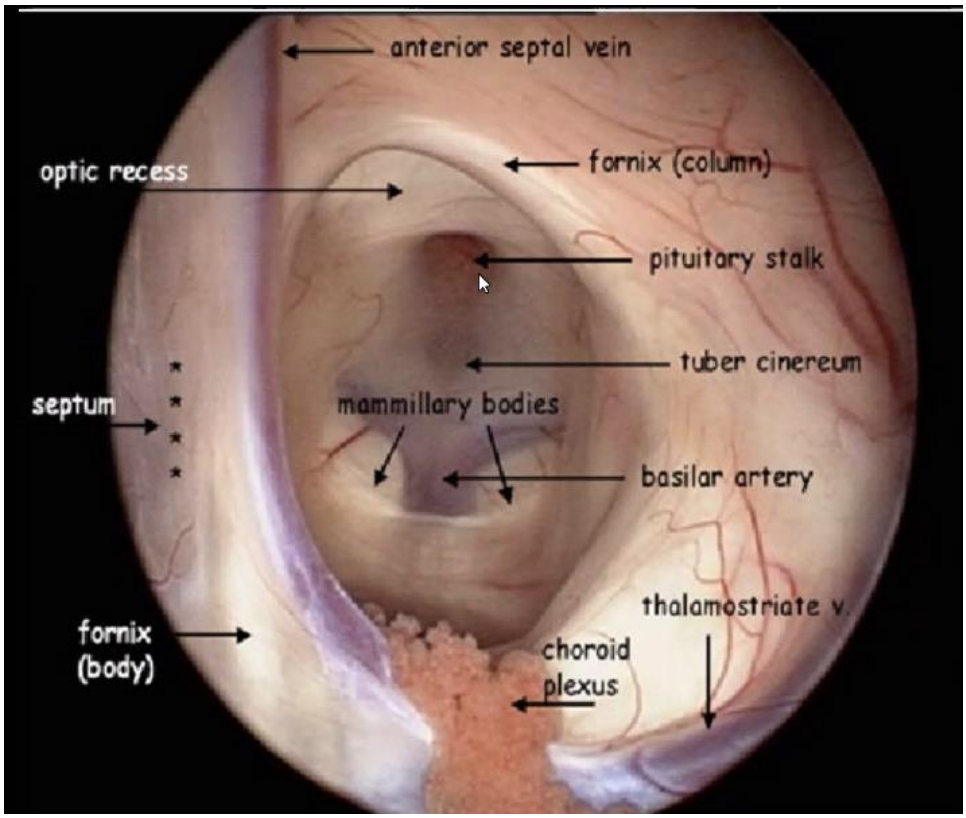
Classical Kocher bur hole site –straight trajectory via right lateral ventricle to foramen of Monro

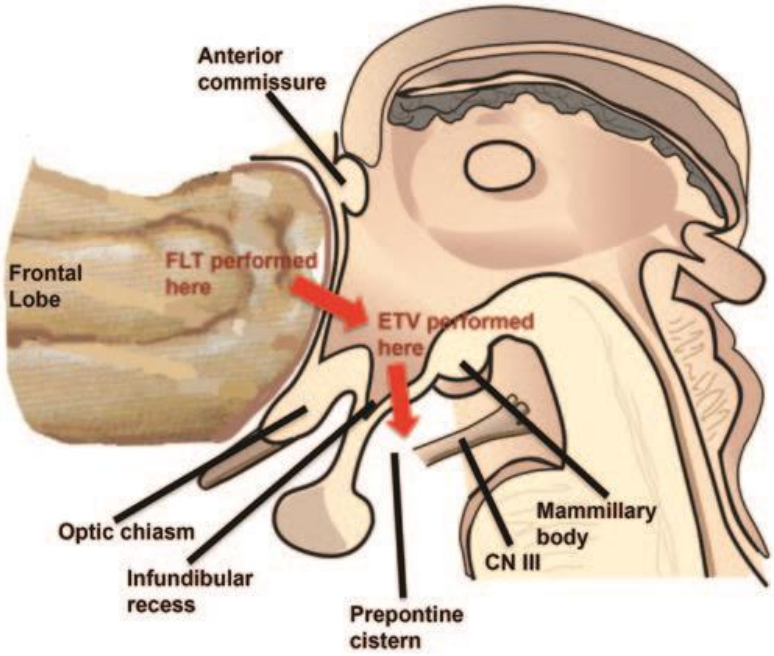
- use **rigid** endoscope.
- choroid plexus is the best guidance towards foramen of Monro.
- floor of third ventricle must be *thin and translucent enough to permit visualization of the basilar artery and mammillary bodies* - if these structures cannot be visualized then procedure is aborted.
- pierce mechanically with **blunt Bugbee tip without cautery current** (no sharps, no electricity! – basilar tip!) to perforate 3rd ventricular floor in several spots just anterior to mamillary bodies (through tuber cinereum) → keep **2-3F Fogarty catheter ballon** inflated a little bit for hemostasis and tissue hole expansion (use 0.2 mL then may add 0.75 mL balloons).
  N.B. do not go too anterior – will damage pituitary stalk and cause DI!
- then advance Fogarty catheter – need to perforate Liliequist arachnoid membrane below 3rd ventricle floor – should clearly see dorsum sellae and basilar artery – prepontine cistern!
- may use monopolar cautery (Bugbee) to coagulate choroid plexus.
- may leave **Ommaya reservoir** with *catheter in ventriculostomy orifice* – keeps orifice open and also gives access to CSF in case ETV fails; alt - may leave **clamped EVD** in place postoperatively.
- postop MRI will show drop out of T2 signal at stoma of ETV.



A. Septum pellucidum
B. Column of fornix
C. Body of fornix
D. Caudate nucleus
E. Anterior caudate vein
F. Foramen of Monro
G. Thalamostriate vein
H. Thalamus
I. Choroid plexus







OUTCOME

Chance of an ETV lasting 6 months without failure:

Table 25.1 ETV Success Score

Category	Description	Value	Score
Age	<1 month	0%	___%
	1 to<6 months	10%	
	6 months to<1 year	30%	
	1 to>10 years	40%	
	≥10 years	50%	
Etiology	• post-infectious	0%	___%
	• myelomeningocele • post IVH • non-tectal brain tumor	20%	
	• aqueductal stenosis • tectal tumor • other	30%	
Shunt history	• previous shunt	0%	___%
	• no previous shunt	10%	
		Total (range 0–90%)	___%

SEPTOSTOMY

Need more lateral bur hole than for Kocher approach (classically – **lateral eye canthus**); use endoscope and Fogarty balloon.
avoid injury to fornices!

ARRESTED HYDROCEPHALUS, s. long-standing overt ventriculomegaly of adulthood (LOVA)

- stable ventriculomegaly (in absence of functioning shunt) with stable neurologic status.

N.B. it is not benign - may be associated with cognitive decline and even sudden death.

Arrested hydrocephalus - stable ventriculomegaly (in absence of functioning shunt) with stable neurologic status.

- careful follow-up (esp. neuropsychological testing) is still required, particularly in children - reported cases of *sudden death*, sometimes years after initial diagnosis.

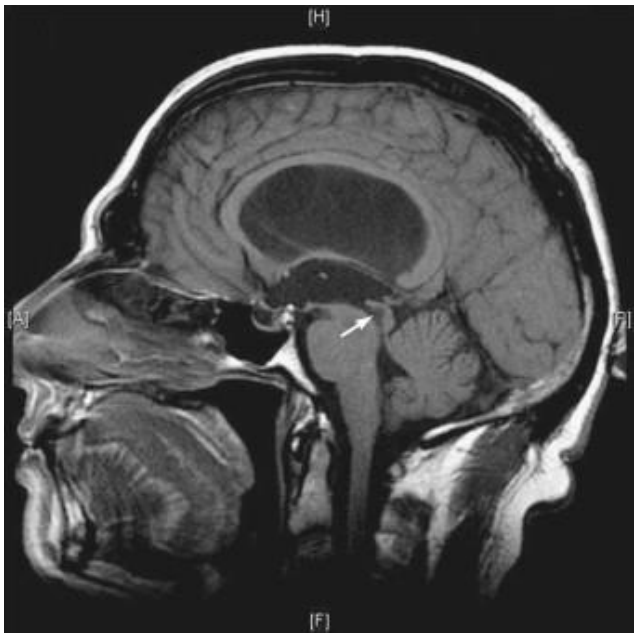
TREATMENT

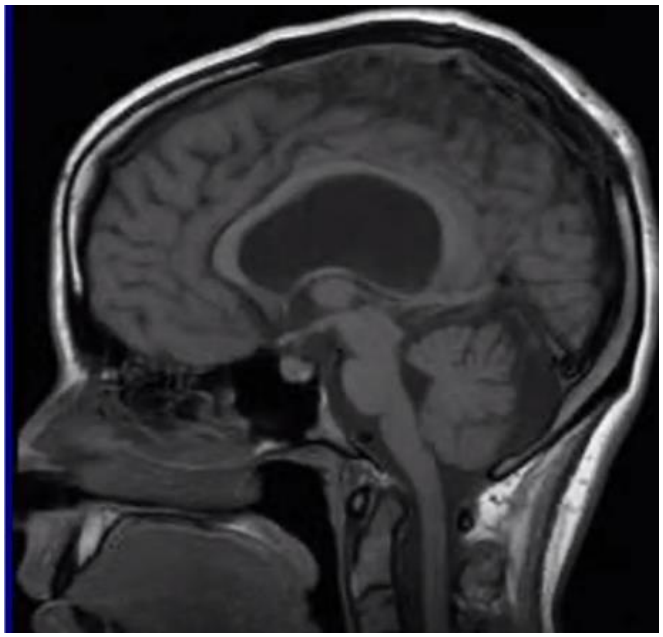
- should not be treated if asymptomatic or stable neurological exam (i.e. it important to ascertain before operating that hydrocephalus is progressive)
- careful follow-up (esp. scheduled interval neuropsychological testing) is still required, particularly in children.

  N.B. children can present with very subtle neurological deterioration (e.g. slipping school performance).
  Decompensation of arrested hydrocephalus can be sudden and fatal.
- ETV is a treatment of choice (VPS may cause overdrainage and SDH).

AQUEDUCTAL STENOSIS

- ≈ 1/3 congenital hydrocephalus cases
  - may be inherited - **X-linked aqueductal stenosis**
- HA, upward gaze palsy
- normal or small 4th ventricle!
- normal aqueduct of Sylvius is 3 mm length and 2 mm diameter in child
- ***aqueductal gliosis*** may be result of neonatal meningitis or SAH in premature infant→ brisk glial response → complete aqueduct obstruction





- ETV is a treatment of choice

See Case P3 >>

NORMAL-PRESSURE HYDROCEPHALUS

- impaired CSF absorption in the absence of prior illness or injury - ***communicating hydrocephalus*** with *increased resistance to CSF outflow* (at *subarachnoid space* or *arachnoid villi* or *glymphatics*) and **normal ICP** (absence of headache & papilledema).

- > 60 yrs.
- dilated ventricles without brain parenchymal loss
- **GAIT APRAXIA** **wide-based "magnetic gait" with small steps** → **DEMENTIA** ***subcortical frontal dysexecutive syndrome*** (mild ÷ moderate; accounts for ***5% of demented patients*** in older age group) → **URINARY INCONTINENCE** ***detrusor hyperactivity***
  - legs are bradykinetic (vs. Alzheimer disease – normal gait).
  - gait disturbance should precede cognitive decline!!!
- normal motor force, tone, and reflexes.

  N.B. upper motor neuron signs or lower limb weakness may be indicative of ***cervical myelopathy*** and ***lumbar canal stenosis***, respectively!
- *discrepancy between **walking** and **simulated walking*** (eliminates pyramidal lesion) - can move legs well and imitate walking while in chair, but becomes awkward and severely impaired as soon as attempts to walk.
- differentiation from parkinsonism:
  - Parkinson's patients are able to increase their stride length and walking cadence with aid of external cueing such as counting, command lines / landmarks; vs. patients with NPH have gait apraxia that does not respond to aids.
  - patients with NPH mobilize with relatively preserved arm swing.
- differentiation from Alzheimer's disease: NPH has milder disorientation and memory impairment, and greater frontal lobe dysfunction (attention impairment, declined psychomotor speed, impaired verbal fluency, dysexecutive syndrome).

Evans' index > 0.3
Callosal angle < 90 degrees
Disproportionately enlarged subarachnoid-space hydrocephalus (DESH) - imaging hallmark of iNPH: ventricular enlargement accompanied by shrinkage of the subarachnoid space at cerebral high convexities ("high-convexity / midline tightness") + Sylvian fissure enlargement.

Ventriculomegaly + absence of full triad = generally not NPH

No any single component of clinical triad = unlikely NPH

No ventricular enlargement + some triad symptoms = unlikely NPH

Papilledema or ICP↑ = unlikely NPH

- **removal of 30-50 ml CSF** → transient clinical *improvement* → **VENTRICULAR SHUNTING**

Guidelines for Management of Idiopathic NPH (3rd Edition 2021, endorsed by the Japanese Society of NPH): CSF drainage test (tap test) is useful and is evaluated within 24 hours after CSF removal and multiple evaluations should be done for up to a week: gait improvement → cognitive and urinary improvement.

- may have 50% placebo effect!

Prolonged external lumbar drainage (3 days at 5 mL/hr) – highest sensitivity (50-100%), specificity (80%), and positive predictive value (80-100%).

Practically:

a) if LP helps or if opening pressure > 15-20 cmH2O (in old patient it is too much) → **shunt**.
b) otherwise → **lumbar drain trial**.

- advance stages (> 2 years' duration) tend to be less responsive to treatment - *early treatment can be instrumental*

Guidelines for Management of Idiopathic NPH (3rd Edition 2021, endorsed by the Japanese Society of NPH):
A programmable-pressure valve is recommended.
If intracranial hypotension is detected and valve is set low, anti-siphon/gravity device should be added.
Initial high pressure setting with gradual decrease

- while valve adjustments are made, keep patient off anticoagulants.

IDIOPATHIC (BENIGN) INTRACRANIAL HYPERTENSION (PSEUDOTUMOR CEREBRI)

Typical patient - ***obese woman of childbearing age*** (15-44 yrs) with chronic daily ***headaches*** ± pulsatile tinnitus, normal neurological examination (except for ***papilledema**** and/or CN6 palsy), and LP with opening pressure > 25 cm H_2O

*best objective reproducible exam - Ocular coherence tomography (OCT).

Chronically elevated ICP → papilledema → progressive optic atrophy → blindness.

Secondary causes: venous sinus occlusion, vitamin A intoxication, minocycline, steroid withdrawal

- **constriction of visual fields**; progressive **vision loss** can occur.

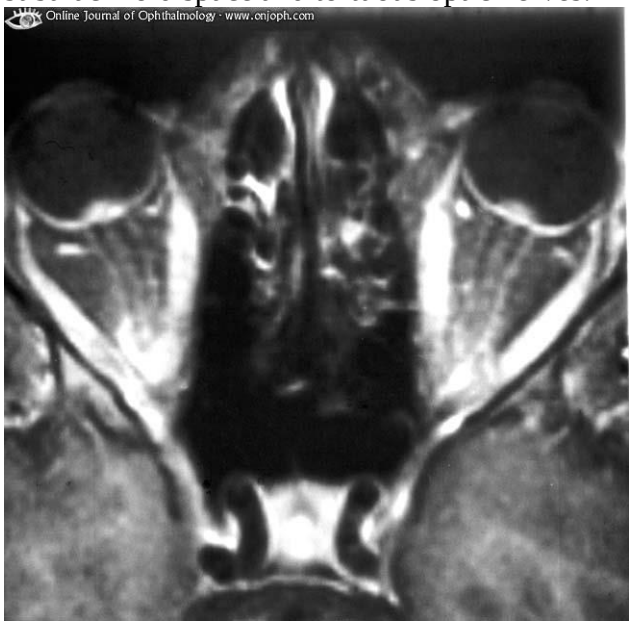
N.B. *visual loss related to optic nerve dysfunction causes major morbidity* (term *benign intracranial hypertension* is improper)!

Perimetry is the best means to detect and follow visual loss!

- high rate of spontaneous remission within some months (usually 1 year).

Send **CSF** - do not miss chronic fungal meningitis!

MRI: ***empty sella***, normal to slit ventricles, flattened posterior eye globes, distention of perioptic subarachnoid space and tortuous optic nerves:



MRV – to rule out stenosis / thrombosis

Trial of **carbonic anhydrase inhibitor**: ACETAZOLAMIDE (teratogenicity!), TOPIRAMATE - anticonvulsant with secondary inhibition of CA
In acute cases – repeat LP large gauge needle (e.g. 18 G) + short course of **high dose corticosteroids** DEXAMETHASONE 12 mg/d

Weight loss should be attempted in all!

shunt (**lumboperitoneal** vs **ventriculoperitoneal** – equal in shunt failure and complication rates!)
- indicated even if there is ***optic atrophy*** but still ***elevated opening pressure*** on LP (shunting prevents complete blindness).

optic nerve sheath fenestration (ONSF) - relative CI – optic atrophy (nerve is scarred)
dural venous sinus angioplasty and stenting – for endovascularly measured **gradient > 8 mmHg**; poststenting - aspirin and Plavix (same as post arterial stenting).

NEUROSARCOIDOSIS

– part of systemic granulomatous disease (lungs, skin, lymph nodes, bones, eyes, muscles, parotid glands) – all patients need CXR

- primarily involves **basal leptomeninges** (parenchymal invasion also often occurs) - ***adhesive*** arachnoiditis with ***nodule*** formation.
- ***noncaseating granulomas with lymphocytic infiltrates*** - *intra- or extraparenchymal, periventricular*, in *basal cisterns*

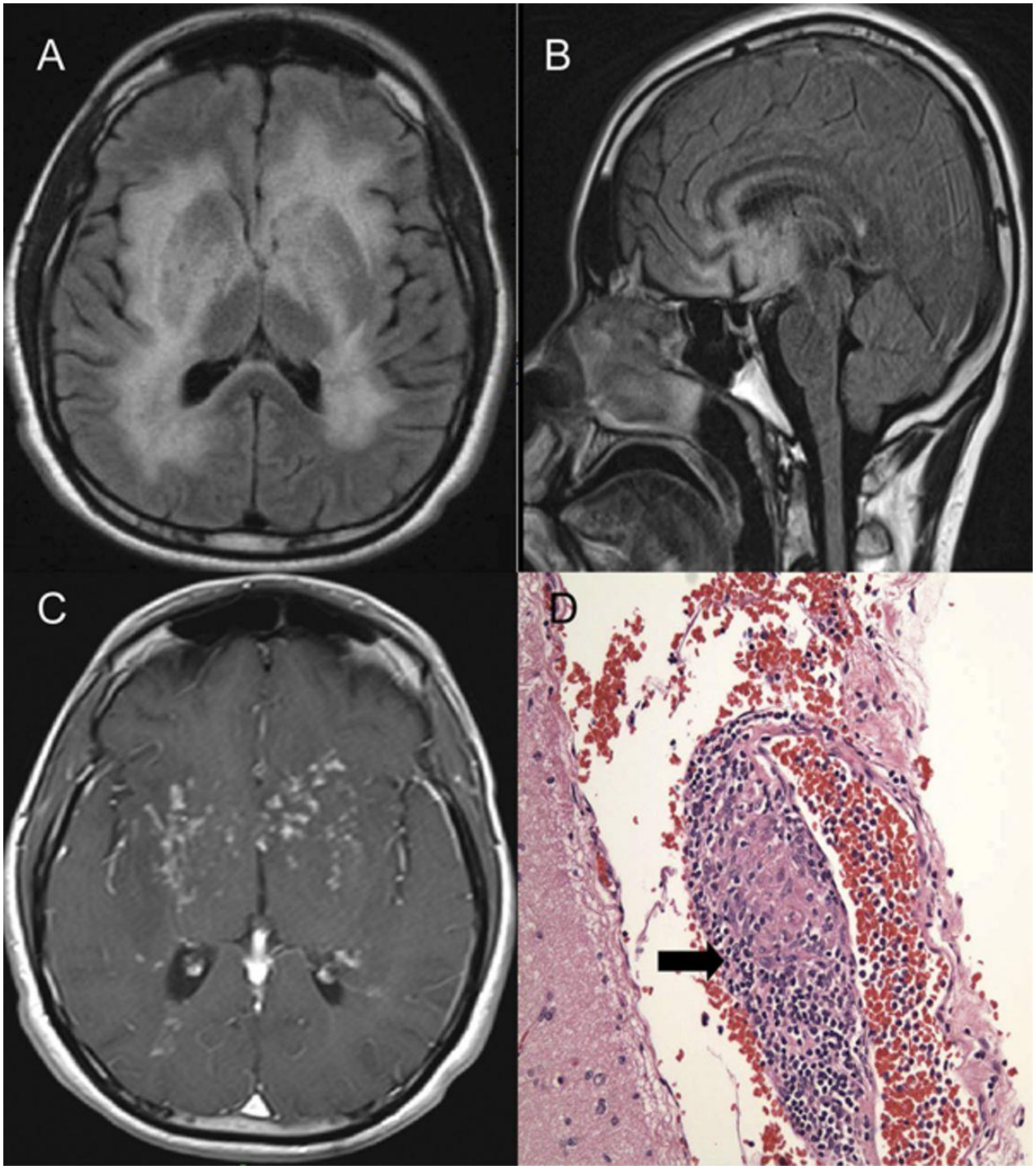
1. Diabetes insipidus (hypothalamic involvement)!
2. Multiple cranial nerve palsies (esp. facial diplegia, CN2)
3. Hydrocephalus - from adhesive basal arachnoiditis
4. Seizures (15%)

1. Mild **leukocytosis** and **eosinophilia**.
2. ↑**Angiotensin-converting enzyme (ACE)** in serum
3. **CSF** (similar to any *subacute meningitis*).
4. Characteristic findings on CXR - hilar adenopathy

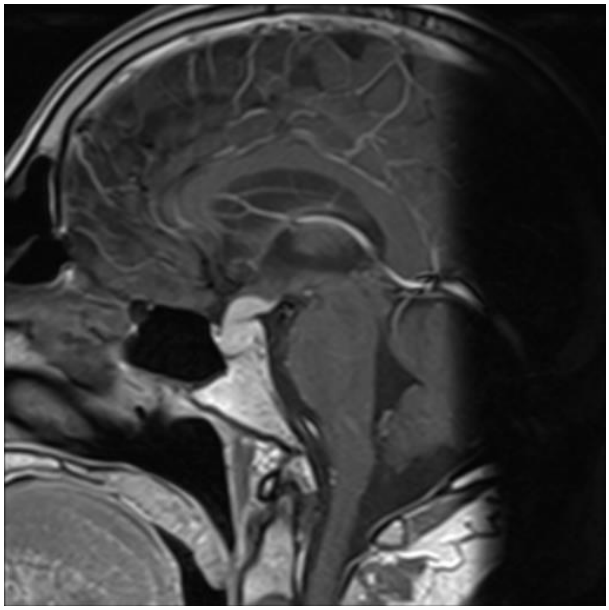
MRI

- FLAIR + gadolinium enhancement of **pachymeninges**, **leptomeninges, cranial nerves, parenchymal** (most common finding – can mimic MS)

Diffuse cerebral neurosarcoidosis mimicking gliomatosis cerebri:



Pituitary involvement:



BIOPSY

- in uncertain cases.
- include all layers of meninges and cerebral cortex.
- + ***fungus*** and ***acid-fast bacteria (TB)***.

TREATMENT

Corticosteroids - PREDNISONE 60 mg PO qd → tapered based on response.

- METHOTREXATE sometimes used as a second line agent
- CYCLOSPORINE may allow a reduction in steroid dosage in refractory cases.

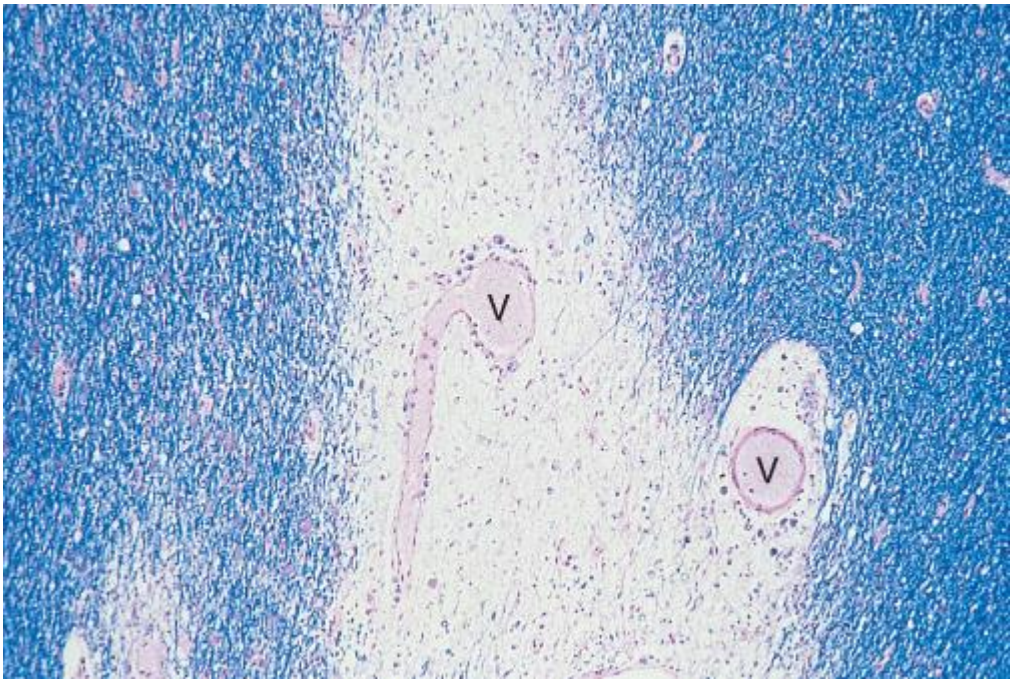
DEMYELINATING

MS

– anti ***MBP (myelin basic protein)*** → slowly progressive CNS inflammation (monocytic and lymphocytic perivascular cuffing) → demyelination → severe gliosis (glial scars)

- MS plaques found throughout ***white matter of neuraxis*** esp. in ***periventricular regions*** ("Dawson's fingers").
- remyelination is aberrant and incomplete
- in severe lesions, axons may be entirely destroyed → secondary sclerotic degeneration of long tracts.

Chronic plaque - myelin loss (appears pale) containing fibrillary astrocytes; few lymphocytes and **macrophages** around blood vessels (V); normal myelinated white matter appears blue:



Cx: distinct acute, but *not apoplectic* *EPISODES OF NEUROLOGIC DEFICITS* - separated in time + separated in space multiple (≥ 2) white tract symptoms (esp. eye) in young patient; chronic *relapsing-remitting* disease of **young adults**; true exacerbation lasts > 24 hours (vs. Uhthoff phenomenon)

Any symptoms from **any part of neuraxis** from spinal cord to cerebral cortex

Most commonly affected systems are **optic nerves**, **pyramidal tracts**, **posterior columns**, **cerebellum**, **central vestibular system**, **medial longitudinal fasciculus** – visual field cuts, diplopia (INO!!!), scanning speech, spastic paresis, paresthesias, bladder dysfunction

Fatigue - pervasive symptom among MS patients!!!
Optic neuritis - acute visual loss in one or both eyes with mild pain (often on eye movement).

Dx: **No specific test for MS is available** - MS remains **CLINICAL DIAGNOSIS**

1. MRI - ovoid perivenular plaques anywhere in CNS: ↑T2, active lesions enhance, periventricular Dawson's fingers (oriented perpendicular to ependymal surface), dissemination of lesions in time and space.

T2 hyperintense, enhancing (active plaque*) lesions with Central Vein Sign

*i.e. enhancement is not always present

   – DWI should be normal; however, plaques can sometimes exhibit "shine through" (ADC map must be checked to rule out infarct)

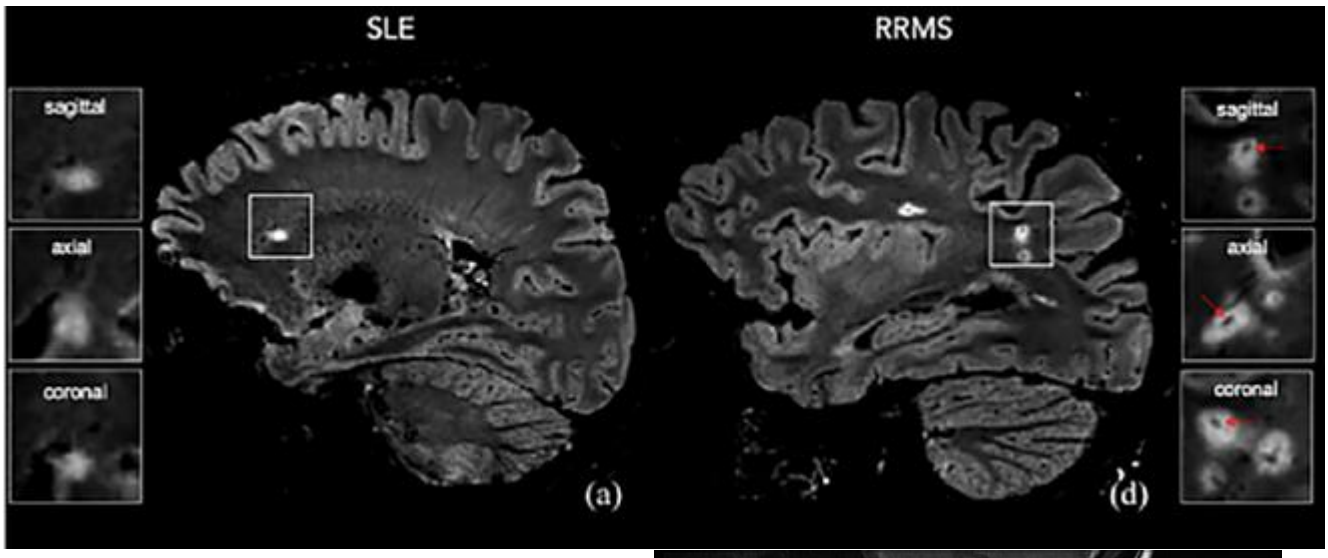
N.B. T2 lesion in spinal cord (esp. with little or no swelling and no enough stenosis to explain it) – ask for MRI with contrast! (MS lesions are < 2 vertebral segments, occupy < ½ of cord cross-section - peripherally located in dorsal and lateral columns) → CSF study for MS + serum aquaporin-4 antibodies

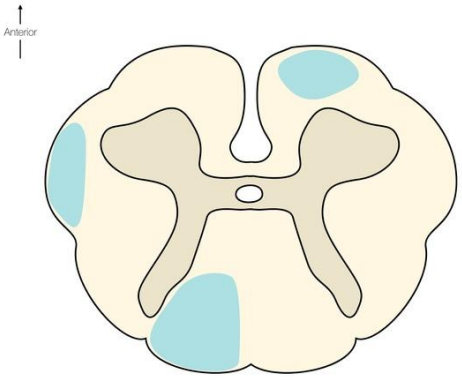
2. CSF: IgG index↑, oligoclonal IgG bands [must not be present in serum], MBP

IgG index= $[IgG_{CSF} / albumin_{CSF}] / [IgG_{serum} / albumin_{serum}]$

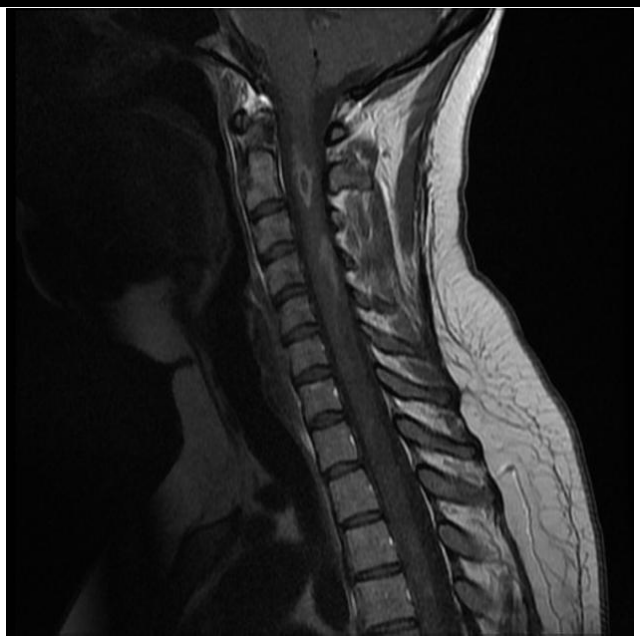
   – normal IgG index is < 0.7
   – most patients have IgG index > 1.7
   – if protein level > 100 mg/dL – look for other diagnoses!
   – if clinical suspicion for MS is high bur CSF equivocal → repeat LP.

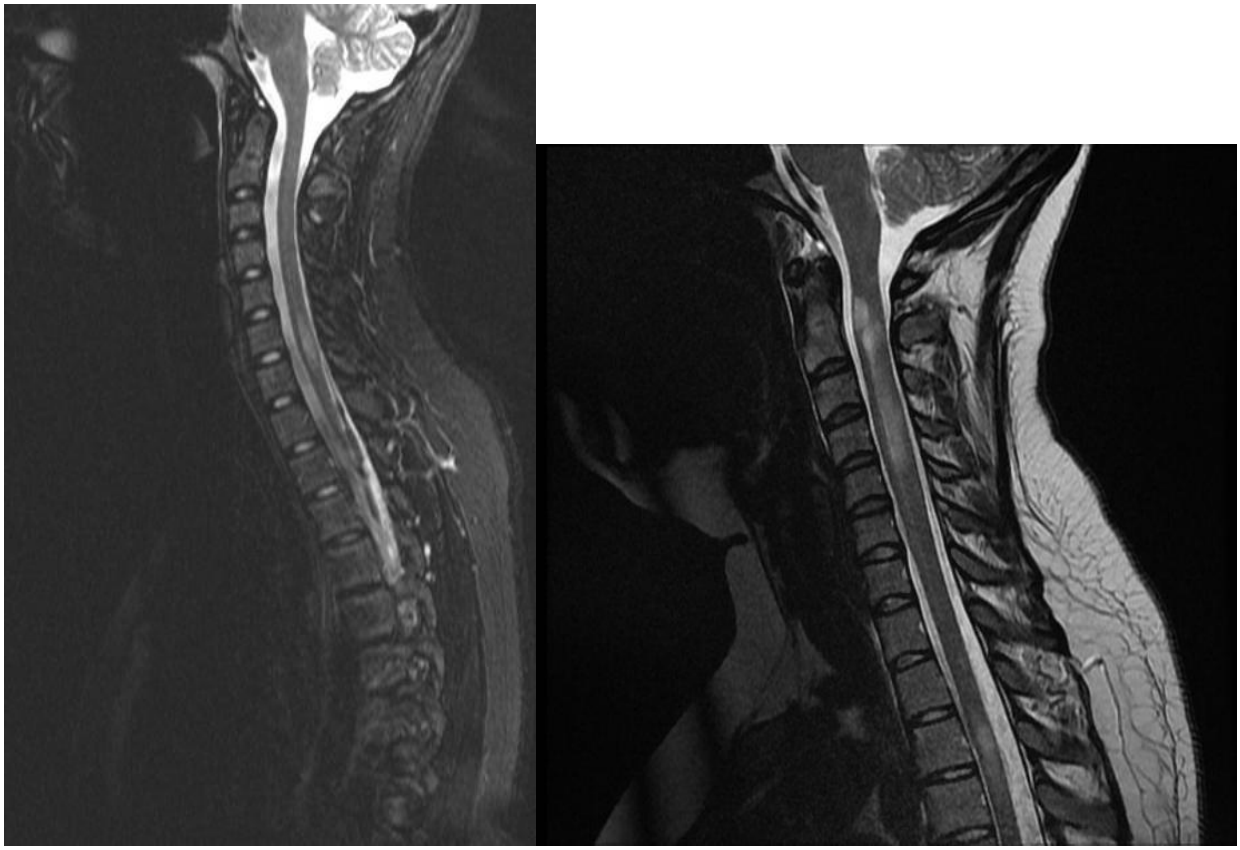
3. Evoked potentials (abnormally delayed, esp. visual EP).





Multiple sclerosis

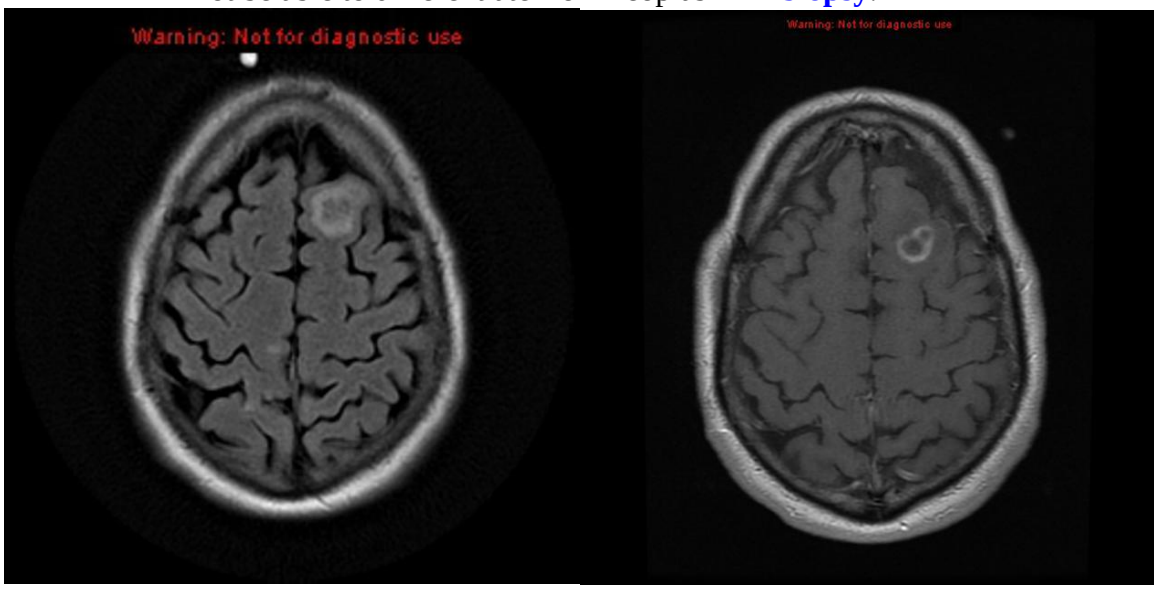




Rx: **attacks** - high-dose METHYLPREDNISOLONE or high-dose PREDNISONE
+ disease-modifying drugs (ASAP to stop irreversible axon loss, continued indefinitely): IFN-β (**A**vonex, **B**etaseron, **R**ebif), glatiramer (**C**opaxone), natalizumab (Tysabri), mitoxantrone

Focal tumefactive demyelinating lesions (TDL) - large aggressive demyelinating lesions - may represent an intermediate position between MS and ADEM.

- tend to be symmetric.
- may enhance, and show perilesional edema - mistaken for neoplasms; even **MRS** may not be able to differentiate from neoplasm → **biopsy**.

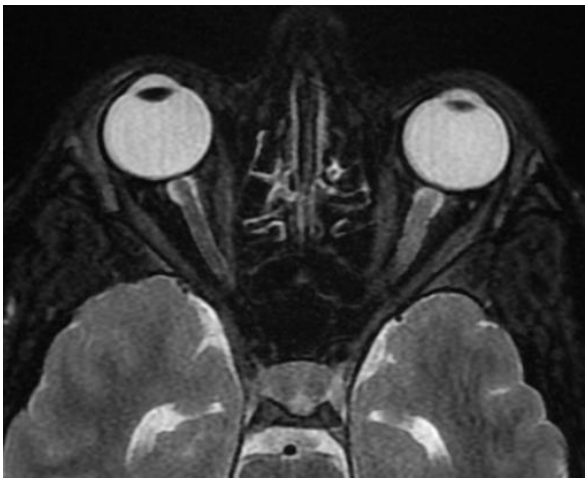
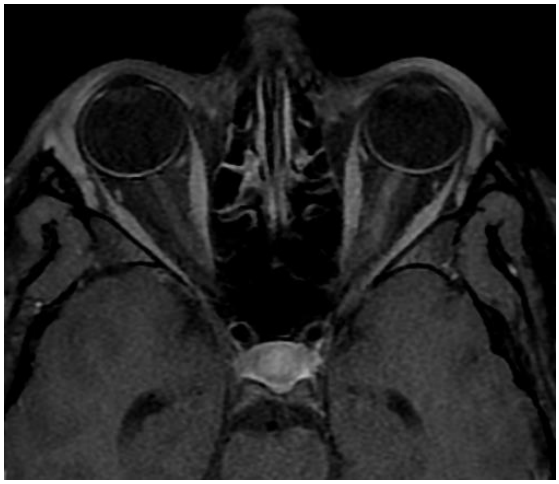


See N6 case >>

NEUROMYELITIS OPTICA (DEVIC'S DISEASE)

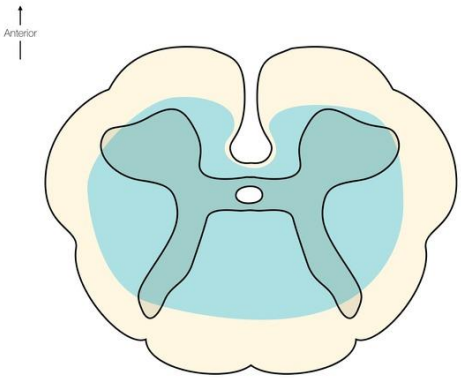
- acute optic neuritis and transverse myelitis in close temporal relationship (interval < 1 month).

1) serum **autoantibody** to aquaporin-4 water channel (70%) (i.e. it is not MS variant)
   + ***marked CSF pleocytosis*** (**PMNs** may predominate), ***protein***↑↑↑, oligoclonal bands are conspicuously absent
2) **optic neuritis** (concurrently with myelitis but one may precede the other by up to several weeks) – MRI: optic nerves (up to chiasm) hyperintense and swollen on T2 and enhancing on T1 (in chronic stages – atrophy):

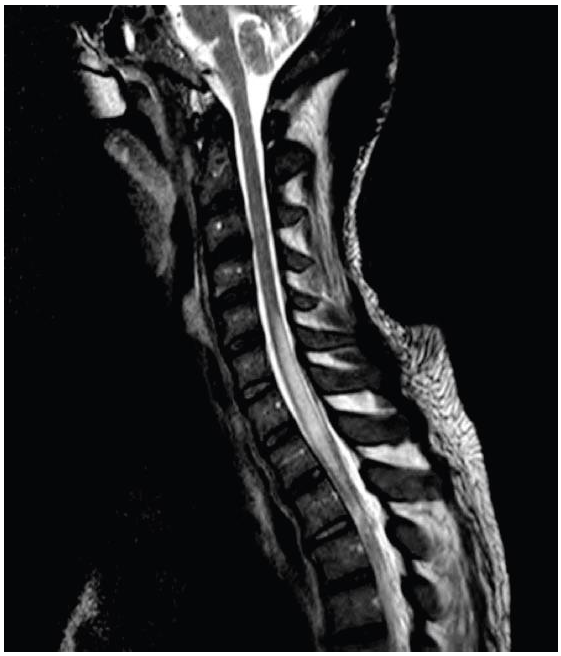



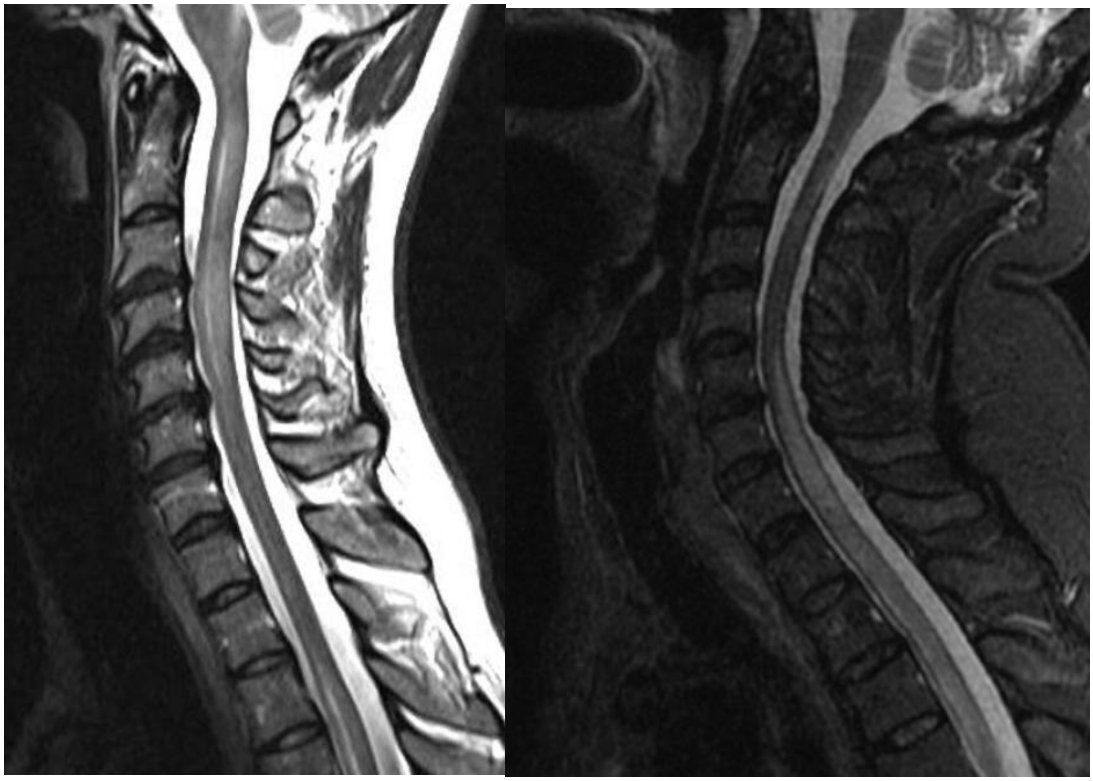
3) longitudinally extensive **myelitis** – preferential involvement of central cord rather than peripheral cord; variable patterns of enhancement: patchy, ring, lens, ependymal.

- diffuse spinal cord ***swelling & softening*** (extends over several levels ÷ nearly entire cord in continuous or patchy distribution) - appearance may resemble spinal cord tumor → biopsy



NMO





Rx: IV METHYLPREDNISOLONE or **plasma exchange**

- **secondary prophylaxis –** RITUXIMAB, NEWER MEDICINES

N.B. MS-specific therapies (e.g. beta-interferon or natalizumab) can actually lead to its exacerbation

MONOPHASIC DEMYELINATION

(may be first clinical episode of multiple sclerosis!):

1. Optic neuritis
2. Acute transverse myelitis
3. Acute disseminated encephalomyelitis (ADEM)

- **multifocal** at onset (i.e. monophasic vs. MS) autoimmune demyelinating disorder (**PERIVENOUS DEMYELINATION !!!**) that begins ***within 6 weeks*** of antigenic challenge (infection or immunization); *monophasic* disease of **prepubertal children**

Rx: METHYLPREDNISOLONE / plasmapheresis / IVIG

ADEM is typically *monophasic* disease of **prepubertal children**;
vs. MS is chronic *relapsing-remitting* disease of **young adults**.

(ACUTE) TRANSVERSE MYELITIS

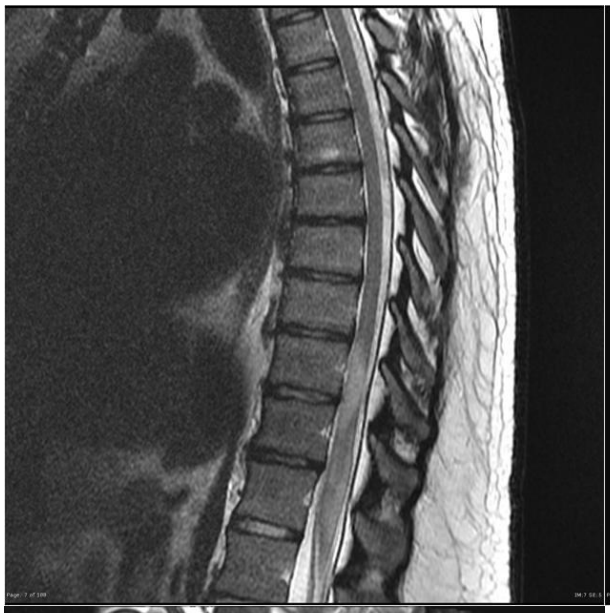
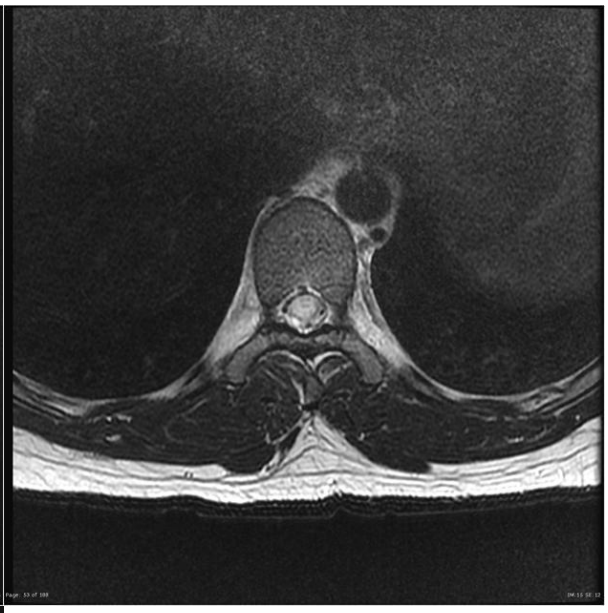
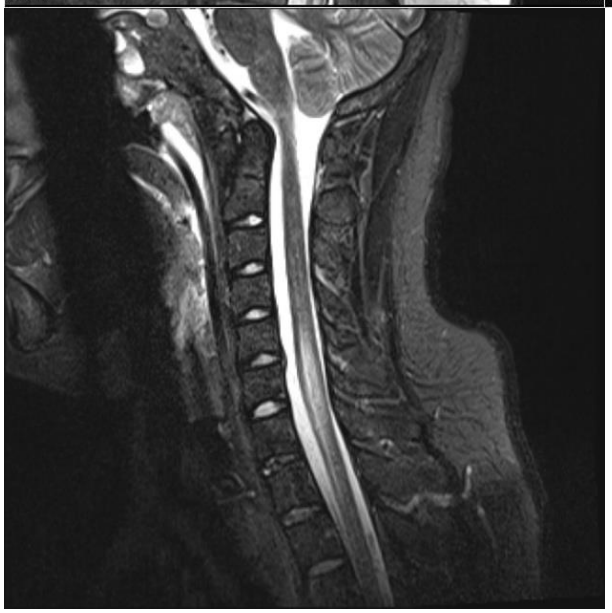
(rapidly* [24 hours] progressive pain + motor, sensory, and autonomic dysfunction):
*tumors – slowly progressive

Immunologic response against CNS (most likely cell mediated)

MRI:

- long segment (≥ 3-4 segments, vs. < 2 in MS).
- > 2/3 of cord cross-sectional area (vs. < ½ in MS)
- variable enlargement of spinal cord
- T2↑
- variable pattern of enhancement (none, diffuse, patchy, peripheral)

- no diffusion restriction ← main dif from cord infarct!

CSF: pleocytosis (lymph and/or PMN) or increased IgG index

N.B. CSF studies always go after MRI ruled out spinal block!

Corticosteroids (e.g. IV METHYLPREDNISOLONE for 3-5 days → MRI – if no response → **plasma exchange**, CYCLOPHOSPHAMIDE).

- in cases of focal spinal cord enlargement, surgical decompression may be considered.
- prognosis variable

LEUKODYSTROPHY

– AR inherited, progressive encephalopathy (at age newborn ÷ 20 yrs)

a) **DYSMYELINATION** (abnormal lipids incorporated into defective myelin are ***metachromatic***)
b) **DEMYELINATION** (e.g. many ***sudanophilic*** leukodystrophies)

1. ALEXANDER'S disease
2. CANAVAN'S disease
3. COCKAYNE'S syndrome
4. KRABBE'S disease (Globoid Cell Leukodystrophy)

5. PELIZAEUS-MERZBACHER disease
6. Metachromatic Leukodystrophy
7. **Adrenoleukodystrophy** - PEROXISOMAL leukodystrophies: **adrenal atrophy**, extensive diffuse **demyelination** (***CNS & PNS***); Dx: **very long chain fatty acids (VLCFA)**↑ in plasma; Rx: **Lorenzo's oil + Steroid replacement**

GUILLAIN-BARRÉ SYNDROME (GBS), ACUTE INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY (AIDP)

- autoimmune (anti myelin sheath) **PNS demyelination** - *rapidly evolving (reaches maximum in 3 days – 3 weeks) sensorimotor polyradiculoneuropathy*: ascending symmetric flaccid WEAKNESS, AREFLEXIA!!!!!, PARESTHESIAS with little sensory loss + CARDIOVASCULAR DYSAUTONOMIA + may involve CRANIAL NERVES

Unlike most neuropathies, proximal muscles are affected more than distal!

Triger - viral URI, immunization, Campylobacter jejuni enteritis, or surgery

CSF analysis – protein↑ with normal cell count (***albuminocytological dissociation***).

EDX examination – segmental demyelination

MRI (with IV gadolinium) - ***enhancement of roots***

Serum - antibodies to gangliosides and glycolipids in peripheral myelin

Rx: plasmapheresis or IVIG

- immunosuppressants, steroids, ACTH are not effective, vs. CIDP!

  Steroids are harmful (GBS is the only autoimmune nerve disorder that steroids are contraindicated!)

See N4 case >>

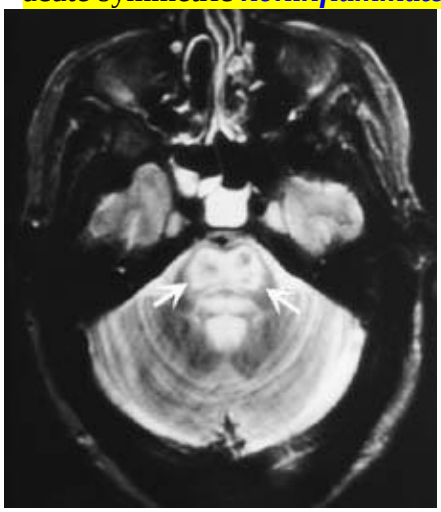
CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY (CIDP)

- chronic course with hypertrophic nerve thickening (“onion bulbs”); may be secondary to ***lymphoproliferative disorders, HIV.***

Rx: plasmapheresis / IVIG + immunosuppression (corticosteroids, azathioprine, cyclophosphamide)

CENTRAL PONTINE MYELINOLYSIS

- acute symmetric ***noninflammatory demyelination*** in central BASIS PONTIS:



Cause - ***too rapidly corrected*** severe and prolonged (< 120 mEq/L for > 48 hours) ***hyponatremia*** (OSMOTIC MYELINOLYSIS). H: correct hyponatremia at max 10 mmol/L/24 h

Cx - **Locked-in**

- *death* is common within days or weeks; full recovery has been reported

PSYCHIATRY

TREATMENT-RESISTANT DEPRESSION

1. Vagus nerve stimulation (VNS) - FDA approved for adults who have failed to respond to at least 4 medications and/or ECT treatment regimens.
2. DBS to subcallosal cingulate gyrus region (64.3% response rate 3-6 years later) - multisite, randomised, sham-controlled trial: study did not show statistically significant antidepressant efficacy in a 6-month trial; 6 additional months of DBS did not increase the proportion of patients who responded to DBS or who achieved remission.
3. Electroconvulsive therapy (ECT) - highly effective
4. Transcranial magnetic stimulation - long-term efficacy

OBSESSIVE-COMPULSIVE DISORDER (OCD)

- severity is scored with **Yale and Brown OCD Scale (YBOCS)**

  0–7 -subclinical symptoms

  8–15 -mild symptoms

  16–23 -moderate symptoms

  24–31 -severe symptoms

  32–40- extreme symptoms.

- surgery (indicated for YBOCS ≥ 28):
  (1) **Anterior cingulotomy**– risk of akinetic mutism (anterior cingulate syndrome).
  (2) **Anterior capsulotomy** – established and most effective procedure; may be done with SRS, RF, LITT.
  (3) **Bilateral DBS**
    a) bilateral *anterior limb of internal capsule (ALIC)* is the only FDA approved (2009) DBS application for psychiatric disorders; FDA approved OCD as an alternative to anterior capsulotomy for treatment of chronic, severe, treatment-resistant OCD in adults who have failed at least 3 SSRIs
    b) bilateral *subthalamic nucleus* (Level I evidence – recommended by CNS/ASSFN 2020 guidelines); target 2 mm anterior and 1 mm medial to target commonly used in Parkinson disease.
    c) bilateral *nucleus accumbens s. VC/VS (ventral capsule/ventral striatum)* (Level II evidence) – favored in worldwide clinical practice

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1. **Form of though**:
   i. circumstantiality: slowed thinking, loaded down with unnecessary detail and digression, but maintaining the goal of the thought.
   ii. flights of ideas: where ideas flow rapidly but remain connected although sometimes by unusual associations, e.g. clang words (associated with mania)
   iii. loosening of association: when the logical sequence of ideas is lost; specific abnormalities include thought blocking when thinking suddenly ceases and there is

a pause before a different thought emerges, and word salad which is the extreme case when words emerge as a jumble

iv. perseveration: inability to change theme, resulting in inappropriate repetition of response.
v. confabulation - bizarre and incorrect responses
vi. neologisms: invented words, or established words with a novel meaning.

2. **Thought content**:
   1) preoccupations: ruminations (ideas of guilt, unworthiness, burden and blame, dwelling on past losses, failures and disappointments, suicidal or homicidal ruminations), somatic preoccupations, hypochondriasis, obsessional thoughts, phobias.
      Specifically state presence or absence of suicidal ideation!
   2) abnormal beliefs: overvalued ideas, delusions (delusions of persecution, grandiose delusions, delusions of reference, delusions of influence, thought broadcasting, somatic delusions)

DRUGS

BUSPIRONE - partial agonist at 5-HT1A receptors – anxiolytic

BENZODIAZEPINES

BENZODIAZEPINE RECEPTORS - *separate from but adjacent* to $GABA_A$ receptor
Overdosage – CNS depression (EEG - widespread *high-voltage beta activity*)
Any benzodiazepine can be used to treat **INSOMNIA** as well as **ANXIETY**!!!

SHORT-ACTING **benzodiazepines** (e.g. TRIAZOLAM) - for **sleep induction**.

INTERMEDIATE-ACTING **benzodiazepines** (e.g. TEMAZEPAM) - for frequent **awakenings**.

LONG-ACTING **benzodiazepines** (e.g. FLURAZEPAM) – for **sleep induction** and frequent **awakenings**; also increase **sleep duration**; may result in daytime sedation.

BARBITURATES

LONG-ACTING barbiturates ($T_{1/2}$ – 1-2 days) – used as **antiepileptics**.
PHENOBARBITAL - duration of action greater than day.

SHORT-ACTING barbiturates ($T_{1/2}$ – 2-8 hours) – used rarely as **sedative-hypnotics** or **anxiolytics**.
PENTOBARBITAL – used to induce **therapeutic coma** in refractory status epilepticus / ICP↑

ULTRA SHORT-ACTING barbiturates ($T_{1/2}$ – 20 minutes) – used for **anesthesia induction**.
THIOPENTAL - acts within seconds; duration of action ≈ 30 minutes.

Withdrawal is much more severe than opiate withdrawal and can result in **DEATH**!!!

NEUROLEPTICS

ANTIPSYCHOTIC EFFECTS depend on blocking of D_2 receptors.
Atypical (2nd generation) neuroleptics (modestly greater efficacy + reduced adverse effects) - more selective D_2 blockade + blockade of serotonin 5-HT_2 receptors
HALOPERIDOL (prototypic *high potency drug*) - drug of choice for acute psychosis
CHLORPROMAZINE is prototypic *low potency drug*
Atypical neuroleptics: **CLOZAPINE, RISPERIDONE, OLANZAPINE, QUETIAPINE, ZIPRASIDONE, ARIPIPRAZOLE, PALIPERIDONE**

ANTIDEPRESSANTS

- MAOI and TCA cause **weight gain**; SSRI - **weight loss, libido↓**
- not suitable for persons < 18 years – ↑risk of suicidality (esp. SSRI).

Class	Reuptake blockage	Receptor blockage
SSRI	serotonin	-
TCAs	norepinephrine-serotonin	muscarinic, H_1, α
SNRI	norepinephrine-serotonin	-
BUPROPION	serotonin-norepinephrine-dopamine	

SSRI - selectively inhibit SEROTONIN reuptake
- SSRI and MAOI or SSRI and triptan concurrent use is contraindicated → "serotonin syndrome"

TCAs block NOREPINEPHRINE and SEROTONIN reuptake (+ block muscarinic, H_1, α receptors) → down-regulation of monoamine receptor densities - onset of antidepressant activity.

SNRI (Selective Serotonin-Norepinephrine Reuptake Inhibitors) - block NOREPINEPHRINE and SEROTONIN reuptake: **VENLAFAXINE**, **DULOXETINE**

BUPROPION – inhibitor of **serotonin-norepinephrine-dopamine** reuptake

MAO-A - norepinephrine, serotonin catabolism (important for depression).
MAO-B - dopamine catabolism (important for Parkinson disease).
- tyramine causes release of stored catecholamines → **hypertensive crisis;** H: **PHENTOLAMINE**, **SODIUM NITROPRUSSIDE**; for less severe cases - **CHLORPROMAZINE**, **PRAZOSIN**.

LITHIUM CARBONATE

- for mania.

N.B. lithium has specific ANTISUICIDE effect!

- extremely low therapeutic index (comparable to *digitalis*)

CHILDHOOD

STRANGER ANXIETY - begins at 7-9 months;
infant separated from mother between 6 and 36 months of age → *anaclitic depression* (apathy, emotional withdrawal, and diminished developmental quotient)

SEPARATION ANXIETY - peak at 18 months.

- **"terrible twos"** at 18-36 months - child attempts to ***separate psychologically from mother*** - noncompliant behavior and resists parental authority - may refuse to eat, sleep, or eliminate at parental request.
- **temper tantrum** - violent emotional outburst (shouting, screaming, crying, thrashing about, rolling on floor, stomping)

DEATH

Elisabeth Kübler-Ross stages before death occurs:

1. **Denial** - patient cannot believe that he is dying.
2. **Anger** - range of reactions (rage, bitterness, confusion).
3. **Bargaining** - search for meaning in life, return to religious institutions, belief that some magical power will intervene.

4. **Depression**
5. **Acceptance** - emotional detachment or neutrality or calm (even euphoric) state.

DISORDERS

Hallucinosis - hallucinations in otherwise *normal mental state* (without confusion, disorientation, or psychosis) - sensations are quickly interpreted as false (vs. hallucinations are experienced as real).

Psychoses:

schizophrenia – for > 6 months: delusions (of persecution), hallucinations (voices speaking to patient), disorganized speech (loosening of associations, thought blocking), grossly disorganized or catatonic behavior (activity↓ or ↑), negative symptoms (apathy, flat affect, social withdrawal up to autism, anhedonia, poverty of thought and content of speech)

brief psychotic disorder – as schizophrenia but < 1 month duration

schizophreniform disorder – as schizophrenia but 1-6 month duration

shared psychotic disorder (s. folie deux) - patient who did not originally have psychotic disorder, develops shared delusion because of close contact with psychotic person (such as parent or spouse).

delusional disorder (paranoia) – just nonbizarre delusions (for > 1 month) - delusions well systemized, well-organized, relatively consistent.

schizoaffective disorder - mood episode (manic or major depressive) with acute psychotic symptoms

Mood (s. Affective) Disorders:

major depressive disorder (MDD) - ≥ 1 episode of > 2 week duration (usually lasts months): depressed mood, diminished interest / pleasure, weight loss / gain, insomnia / hypersomnia, psychomotor agitation / retardation, loss of energy, feelings of worthlessness / guilt, diminished ability to think / concentrate, recurrent suicidal ideation

melancholia - *more severe subtype* of major depression

adjustment disorder - like MDE, but in context of recent life stressor (i.e. reactive depression)

normal bereavement - after loss of a loved one, no severe impairment/suicidality.

seasonal affective disorder – MDD during autumn ÷ winter

Rx: **bright light therapy** ± antidepressant

manic episode - > 1 week duration (usually lasts months): elevated mood, inflated self-esteem or grandiosity, need for sleep↓, talkative (pressured speech), flight of ideas, distractibility, goal-directed activity↑, involvement in pleasurable activities that have high potential for painful consequences

Rx: for acute mania – **VALPROATE** ± **atypical antipsychotic**
Maintenance - **LITHIUM**

bipolar I disorder (s. classic manic-depression) – at least one manic episode ± anything else (e.g. (mild) depression)

bipolar II disorder – depression + hypomania

for depressive episodes in bipolar disorders - **mood stabilizer** (**LITHIUM**, **LAMOTRIGINE**) ± **antidepressant**
(antidepressants alone may propel patient into manic episode!)

cyclothymic disorder – mild depression + hypomania

dysthymic disorder – chronic (> 2 yrs) mild depression

Sexual Disorders:

Dopaminergic medications - improve both sexual drive and performance in men and women; ***Serotonergic medications*** – priešingai

sexual dysfunction: hypoactive sexual desire disorder (most common type of sexual dysfunction in women!!!; severe form - sexual aversion disorder), male erectile disorder (impotence), female sexual arousal disorder, inhibited orgasm, premature ejaculation, dyspareunia (genital pain associated with sexual intercourse), vaginismus (spasm of muscles that surround outer portion of vagina)

paraphilias

gender identity disorders (most severe form - transsexualism)

premenstrual dysphoric disorder: Rx - **SSRI** – specifically beneficial effects

Anxiety Disorders (for short treatment – benzodiazepines; for chronic treatment – antidepressants; also β-blockers):

panic attack - sudden unexpected unprovoked (spontaneous) episode of intense anxiety (*sympathetic hyperactivity*); not medically dangerous

panic disorder - at least one panic attack followed by ≥ 1 month worries about it

generalized anxiety disorder - > 6 months of almost daily excessive worry about many actual life circumstances (i.e. fear of specific but multiple circumstances).

obsessive-compulsive disorder - obsessions (unwanted and bothersome recurrent ideas, that intrude upon patient and cannot be pushed out of consciousness), compulsions - irresistible need to perform activity; patient realizes that thoughts or behaviors are irrational (vs. in psychosis)

phobic disorders - persistent, unreasonable (irrational), intense fear:

social phobia

agoraphobia (as adjunct to panic disorder)

specific phobia

posttraumatic stress disorder (PTSD) – begins any time after extreme traumatic event and lasts > 1 month: reexperiencing initial trauma (nightmares), avoidance of stimuli associated with trauma; emotional numbing (detachment), hyperarousal (startle↑, insomnia)

acute stress disorder (ASD) – as PTSD but begins within 1 month of event and lasts < 1 month

situational anxiety

Personality Disorders - personality traits (patient > 18 yrs) that are enduring, inflexible, immature, maladaptive, causing significant *impairment in* interpersonal (!!!) *functioning* - affect *interpersonal relationships!!!* **Psychotherapy** is at core of treatment

Type A personality (twice risk for coronary artery disease)

Cluster A ("weird"): paranoid, schizoid, schizotypal

paranoid - pervasive distrust and suspiciousness

schizoid – voluntary social withdrawal

schizotypal – schizoid + odd thinking

vs. schizophrenia – greater odd thinking than schizotypal

Cluster B ("wild"): borderline, histrionic, narcissistic, antisocial

histrionic - excessive emotionality and attention-seeking behavior

narcissistic - grandiose and require admiration from others

Cluster C: obsessive-compulsive, dependent, avoidant

avoidant - very shy

obsessive-compulsive - preoccupation with orderliness, perfectionism, and control.

Somatoform (s. Psychosomatic) Disorders - chronic disabling mental disorders with presenting features suggesting physical illnesses:

somatization disorder - complaints in ***multiple*** (!!!!) body parts

conversion disorder (s. hysteria) - unconscious acute brief symptoms of sensory / voluntary motor function, which suggests ***neurologic disease***.

body dysmorphic disorder (s. dysmorphophobia) - preoccupation with imagined or slight ***visible defect*** in body (usually of face).

hypochondriasis - chronic preoccupation with fears of having ***serious illness***

(somatoform, s. psychogenic) pain disorder

factitious disorder – individual intentionally produces symptoms of illness (i.e. it is entirely mental disorder) *just to be in patient role*; in health care workers - Münchhausen syndrome.

malingering – as factitious disorder, but it is not mental illness.

Eating Disorders:

anorexia nervosa (underweight) - only concern is body shape and weight → obsessional weight loss (fasting, binge eating → purging) + amenorrhea!

bulimia nervosa (normal body weight) - recurrent binge eating followed by guilt, depression, and anger at oneself for doing so → caloric restriction or exercise, purging; no significant weight loss!

binge eating disorder (overweight) - binge eating not followed by inappropriate compensatory behavior

Impulse Control Disorders: intermittent explosive disorder (s. episodic dyscontrol syndrome - violent physical behavior with minimal provocation → sincere remorse), kleptomania (secondary gain plays no role), pyromania, pyrolagnia, trichotillomania, trichophagy, pathological gambling

Dissociative Disorders: dissociative amnesia, depersonalization disorder, dissociative fugue, dissociative identity disorder (s. multiple personality disorder)

SUBSTANCE USE

Abuse – during driving, legal problems
Addiction – overinvolvement
Physical dependence – tolerance, abstinence
Physical signs (blackouts, weight loss, fatigue, chronic cough)

Substance-induced disorders - symptoms that begin during or < 1 mo after intoxication with or withdrawal from implicated substance:

Substance-induced *psychotic disorder*
Substance-induced *mood disorder* with major depressive-like episode / with manic features
Substance-induced *delirium / dementia / amnestic disorder / anxiety disorder / sexual dysfunction / sleep disorder*
Substance INTOXICATION
Substance WITHDRAWAL

Substance use disorder

1. (Recreational) Substance use
2. Substance abuse – social problems
3. Substance physical dependence – tolerance, withdrawal
4. Substance psychic dependence (addiction) - overwhelming involvement (intrusion into person's life)

Opioid withdrawal:

1. Partial agonist substitution **METHADONE** / **BUPRENORPHINE-NALOXONE**
2. + **CLONIDINE** / **LOFEXIDINE** (can halt signs of opioid withdrawal)
   Maintenance of abstinence - **NALTREXONE**

Tobacco withdrawal:
NICOTINE replacement / **VARENICLINE** (agonist at α4β2 neuronal nicotinic acetylcholine receptors) / **BUPROPION**

Cocaine / amphetamine withdrawal: antidepressants that enhance DOPAMINERGIC transmission in reward centers: **BUPROPION**, **DESIPRAMINE** (!!!), **MAPROTILINE**, **VENLAFAXINE**

Benzodiazepines / Barbiturates withdrawal: **PENTOBARBITAL** / **PHENOBARBITAL** → gradual taper

ALCOHOL

binge drinking (≥ 5 alcoholic drinks for men [≥ 4 for women] on one occasion, binges lasting ≥ 2 days)
hazardous drinking (e.g. before or during situations requiring attention or skill, e.g. driving, arrests for driving under influence)

Alcohol abuse - maladaptive episodic drinking
Alcohol dependence - frequent consumption of large amounts of alcohol over time, resulting in tolerance, psychologic and physical dependence and dangerous withdrawal syndrome; when results in significant clinical toxicity and tissue damage - **chronic alcoholism**
Alcohol Intoxication (Drunkenness)

- **FRUCTOSE** might accelerate ethanol elimination; for agitation - low doses of high-potency **antipsychotic** (e.g. **HALOPERIDOL** IV, **RISPERIDONE**); severe cases - **hemodialysis**

Alcohol Abstinence (Withdrawal) Syndromes: minor alcohol withdrawal (occurs in anyone after brief but excessive drinking), delirium tremens (psychic & sympathetic hyperactivity 48-96 hours* after withdrawal of long-term drinking)

*later than other withdrawal syndromes

- ethanol inhibits **glutamate** receptors (long-term ingestion → synthesis of more glutamate receptors; withdrawal → CNS excitability↑).
- acute agitation of alcohol withdrawal may be aggravated by NEUROLEPTICS!
- Rx: ***sedative**** with cross-tolerance with ethanol in loading dose to cause mild intoxication (→ gradually tapered) + **β-blockers / CLONIDINE**

*examples: **benzodiazepines, barbiturates, PARALDEHYDE**

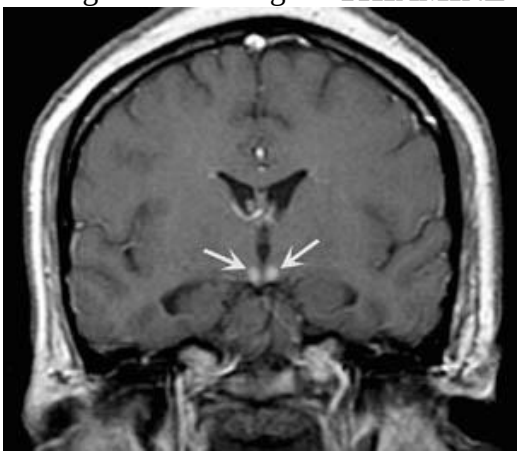
- nsgy patient with signs of alcohol abuse: thiamine 100 mg IV STAT, "goody" bag IV, CIWA scale with phenobarbital PRN, substance abuse consult.

Maintenance of sobriety:
Disulfiram inhibits ALDEHYDE DEHYDROGENASE
OPIATE ANTAGONISTS (safe to give to patients who are still drinking): Naltrexone, Nalmefene
GLUTAMATE RECEPTOR blockers + stimulate GABA transmission: Acamprosate, Topiramate

Wernicke encephalopathy (alcoholics, short-gut syndrome) - acute onset:

1) **Ataxia**
2) **Ophthalmoplegia** (esp. lateral gaze palsy)
3) **Quiet global confusional state** - profoundly disoriented, indifferent, and inattentive

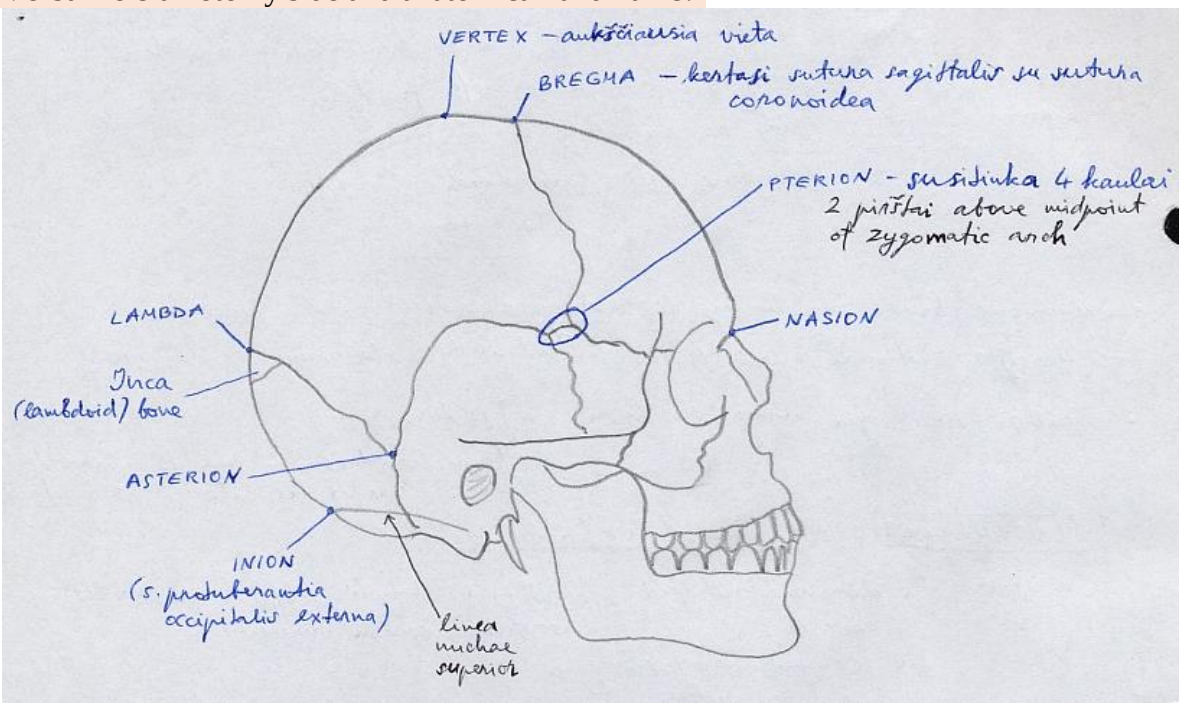
Rx: high dose 100 mg IV **THIAMINE** - untreated progresses to death!

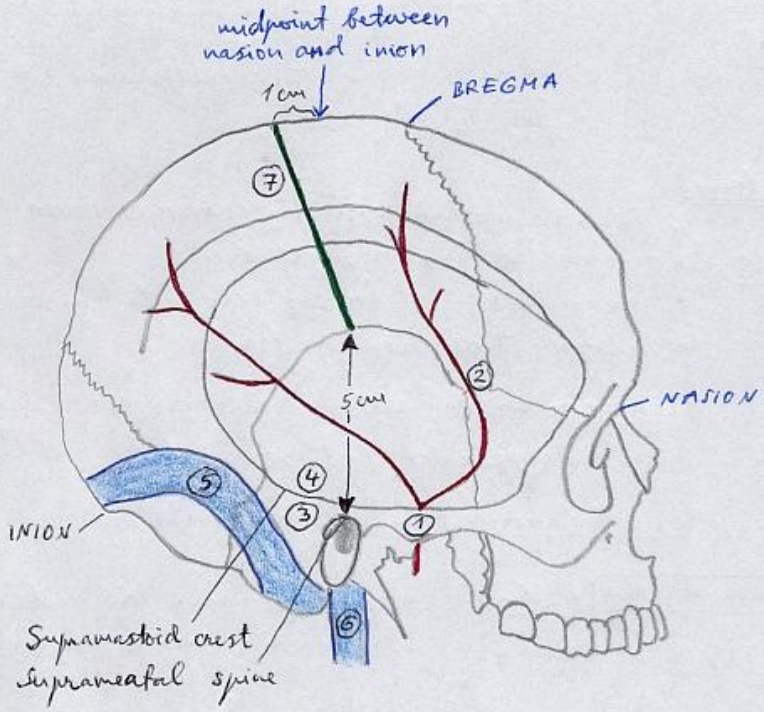


Korsakoff psychosis (chronic sequela of Wernicke encephalopathy) - irreversible very severe anterograde and retrograde (for recent events) amnesia → confabulation, confusion.

CRANIOTOMY

Verbalize craniotomy side and anatomical landmarks!





① A. meningea media kaminas už articular tubercle (projekcijoje)

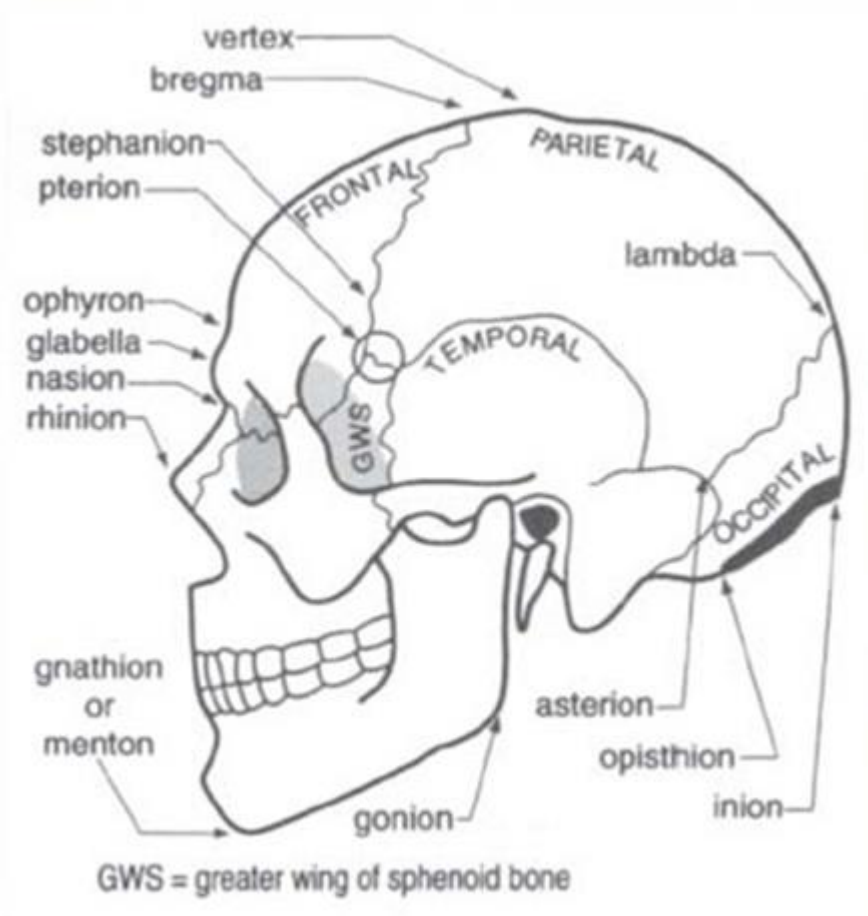
② R. anterior a. meningeae med. eina per PTERION

③ TRIGONUM SUPRAMEATALE (tarp suprameatal spine ir supramastoid crest) - - pateikimas į MASTOID ANTRUM

④ pateikimas į MIDDLE CRANIAL FOSSA (virš supramastoid crest)

⑤ TRANSVERSE + SIGMOID sinus - eina nuo inion iki ≤2cm už external meatus - pereina į ⑥ v. jugularis int. (deep and anterior to mastoid process)

⑦ SULCUS CENTRALIS cerebri (projekcija)



10 (green dot), **superior Rolandic point** 1-2 cm posterior to the midpoint of the nasion–inion line / 4-5 cm behind coronal suture

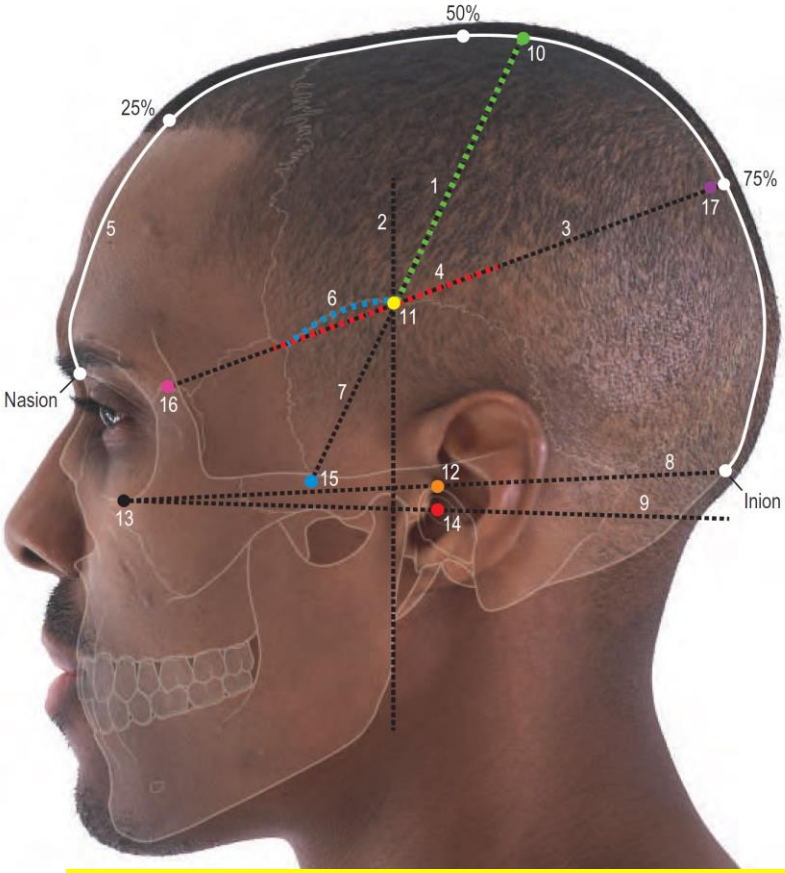
15 (blue dot), midpoint of zygomatic arch

7 line from the superior Rolandic point (green dot) to the midpoint of the zygomatic arch (15, blue dot)

3 Sylvian line passing from frontozygomatic suture [lateral canthus] (16) to point 75% of the way along nasion–inion midsagittal line

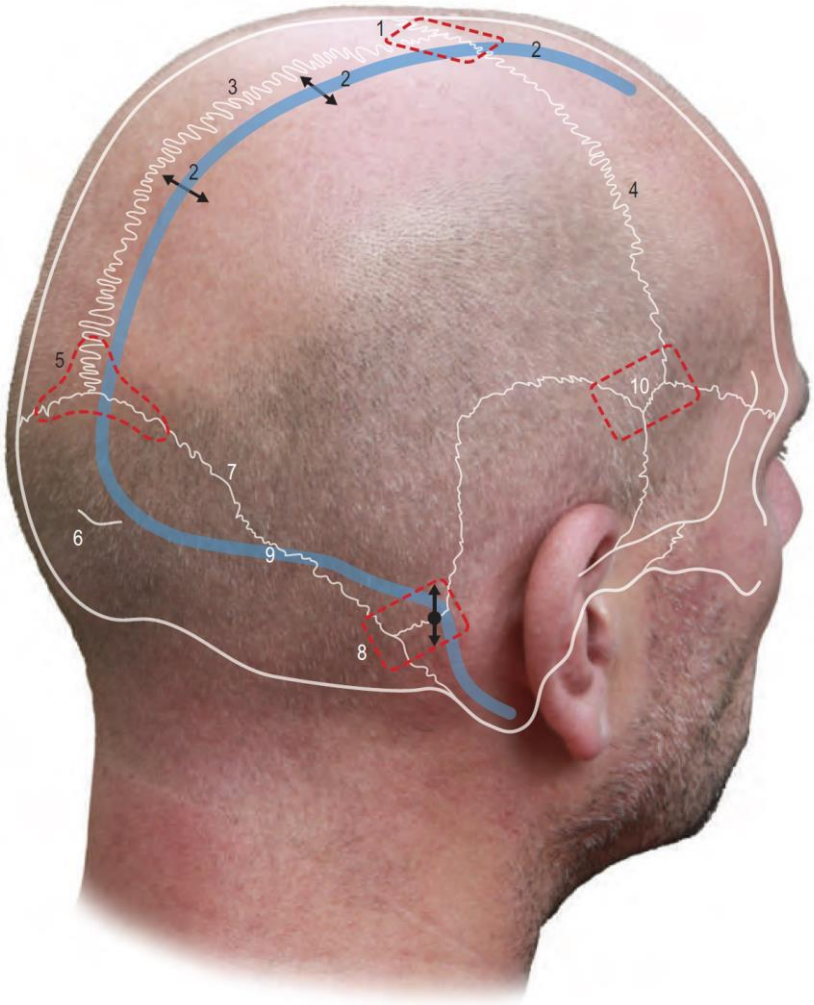
1 Rolandic line/central sulcus (green line)

11 (yellow dot), **inferior Rolandic point** (5 cm above root of zygoma)

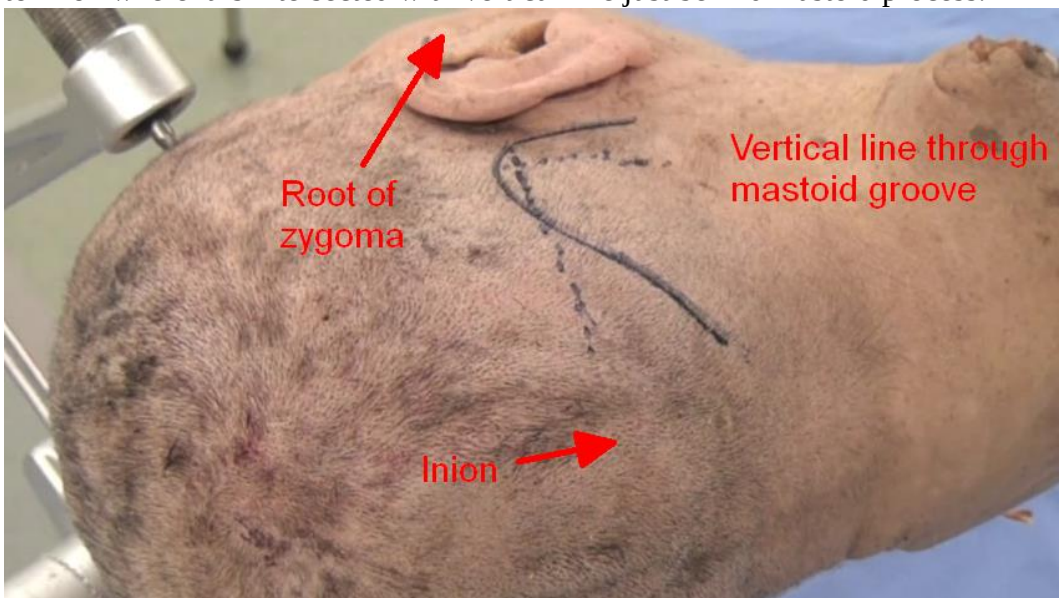


Sylvian point – skull point on nearest the Sylvian fissure and is located about 3 cm behind zygomatic process of frontal bone

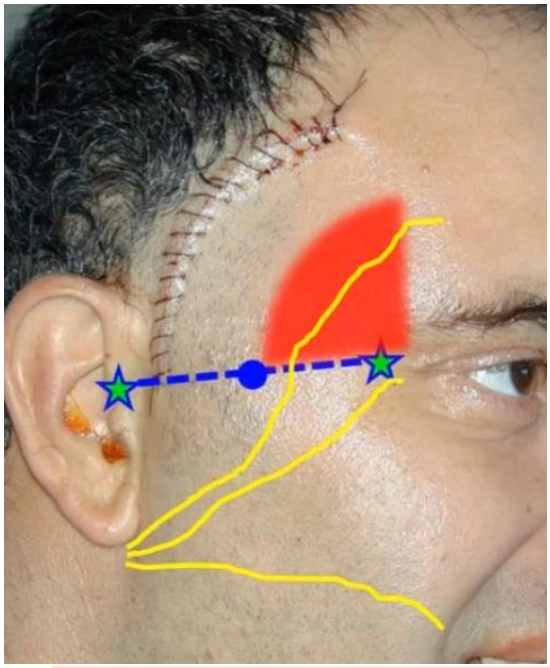
2 superior sagittal sinus: commonly deviates to the right of the sagittal suture by up to 11 mm (black arrows)
6 inion: normally sits below and left of the confluence of sinuses and below the level of the internal occipital protuberance
8 asterion: sits over (in 81%), just below (in 15%) or just above (in 4%) transverse–sigmoid sinus junction
9 transverse sinus: variable course makes accurate surface localization difficult



Asterion – sigmoid-transverse sinus junction most commonly (but not always) hugs it anteriorly-superiorly (so it is safe to drill here if targeting venous sinus angle) – on the line from root of zygoma to inion where it is intersected with vertical line just behind mastoid process.

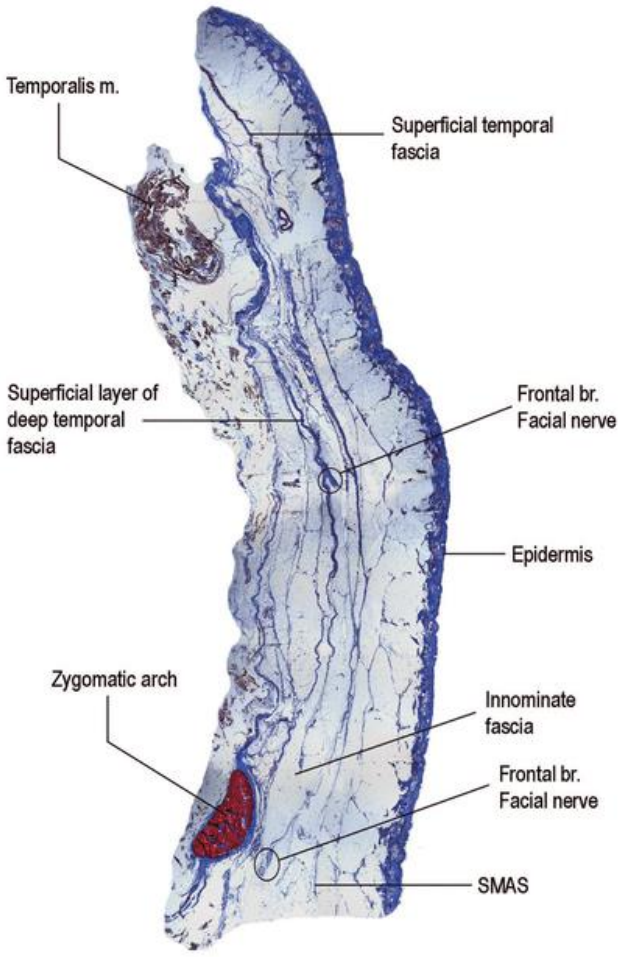


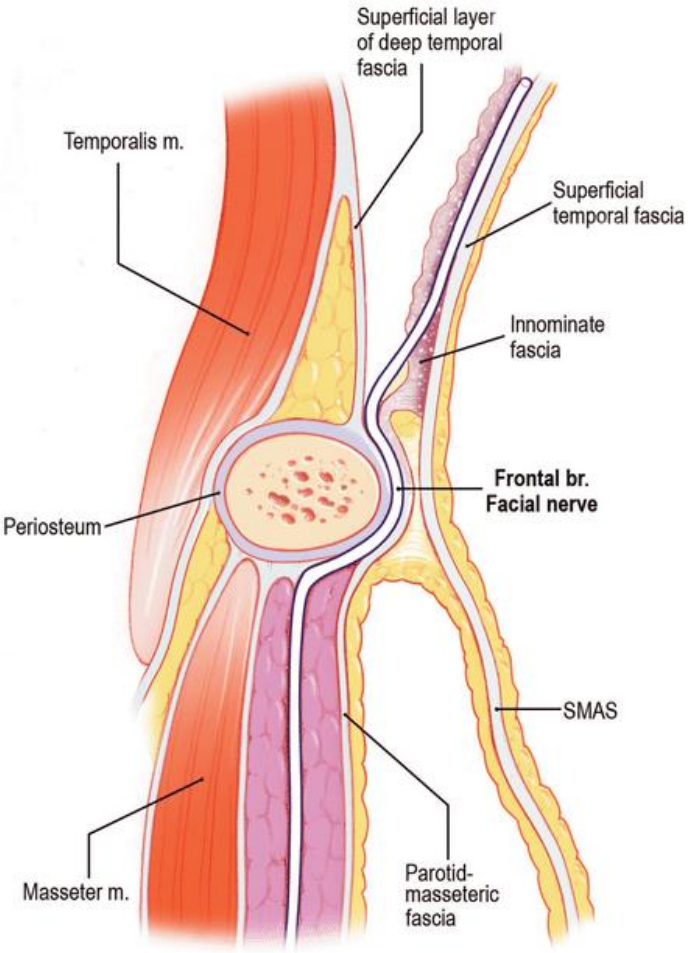
Frontalis branches of CN7 - line between tragus and lateral canthus; frontalis branches are in front of midpoint of this line ("no go" zone – red):

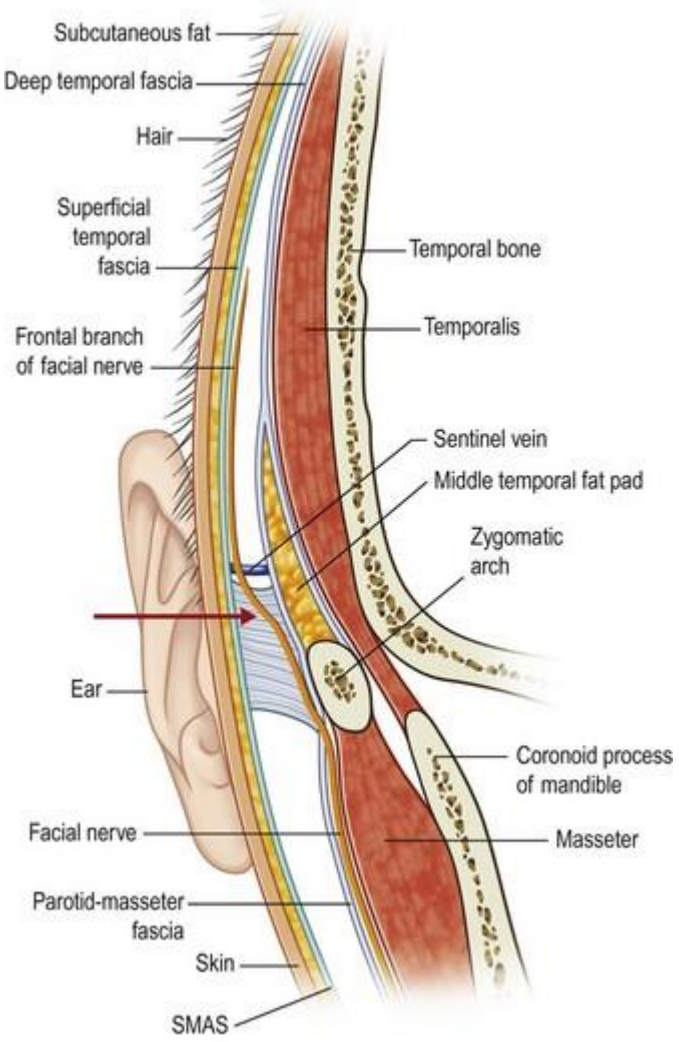


- CN7 frontalis branch(es) are located in the superficial fascia of the fat pad, not within the fat pad - two techniques for reflecting the fat pad

  A. *Interfascial technique*: superficial temporal fascia is reflected anteriorly along with the fat pad via dissection underneath the fat pad but superficial to the deep temporal fascia.
  B. *Subfascial technique* (safer): superficial temporal fascia + fat pad + deep temporal fascia reflected anteriorly all as one layer.


Temporalis m.
Superficial temporal fascia
Superficial layer of deep temporal fascia
Frontal br. Facial nerve
Epidermis
Zygomatic arch
Innominate fascia
Frontal br. Facial nerve
SMAS

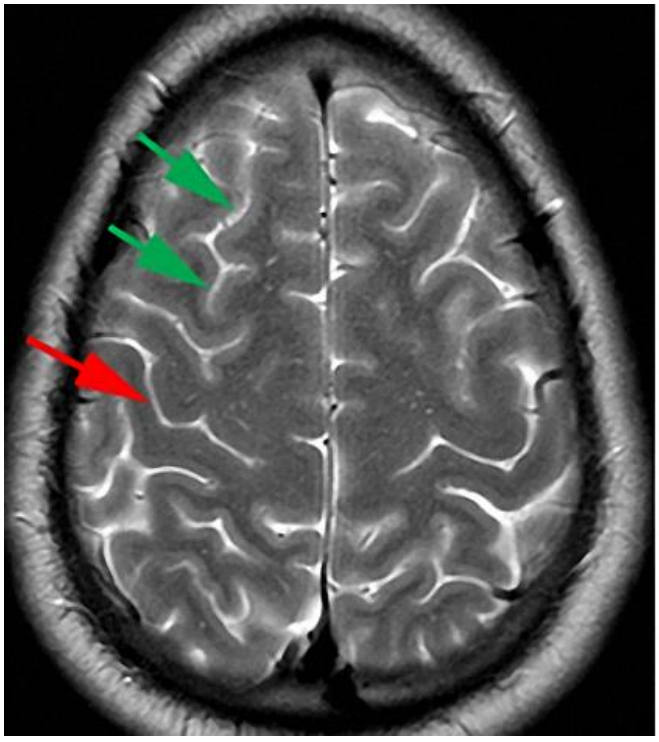

Superficial layer of deep temporal fascia
Temporalis m.
Superficial temporal fascia
Innominate fascia
Frontal br. Facial nerve
Periosteum
SMAS
Masseter m.
Parotid-masseteric fascia

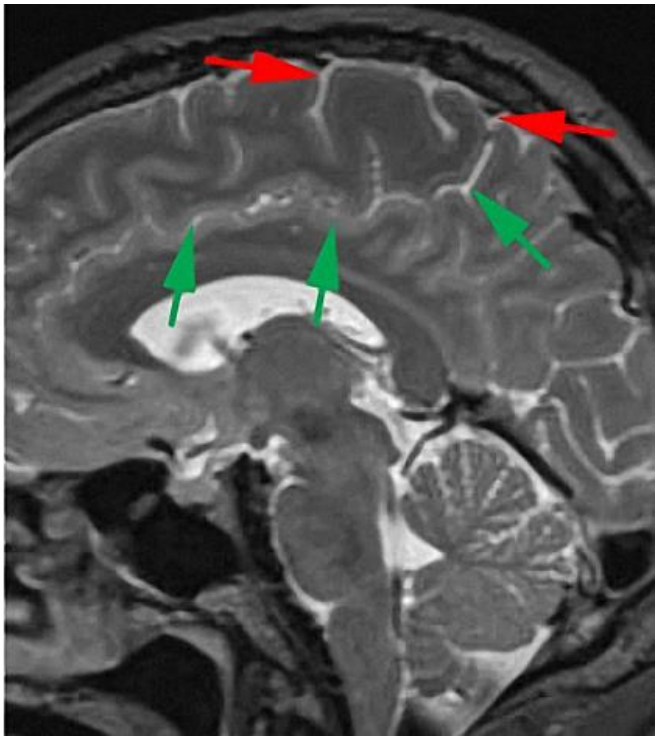


central lobule = sensorimotor cortices

Left image: central sulcus (*red arrow*) is second, more posterior vertical sulcus, from end of superior frontal sulcus (*green arrows*).

Right image: trace cingulate sulcus (*green arrows*) on midsagittal MRI images posteriorly, then superiorly (marginal sulcus) to its end - marginal sulcus is just posterior to central lobule bounded by red arrows.

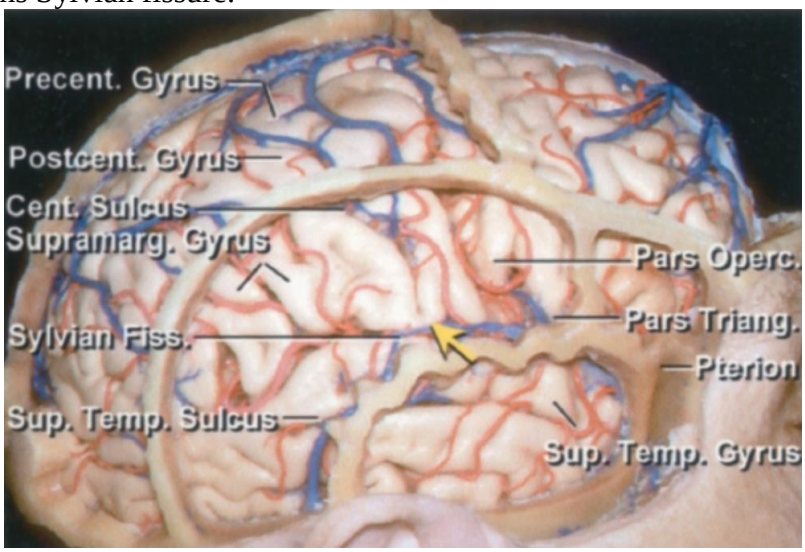




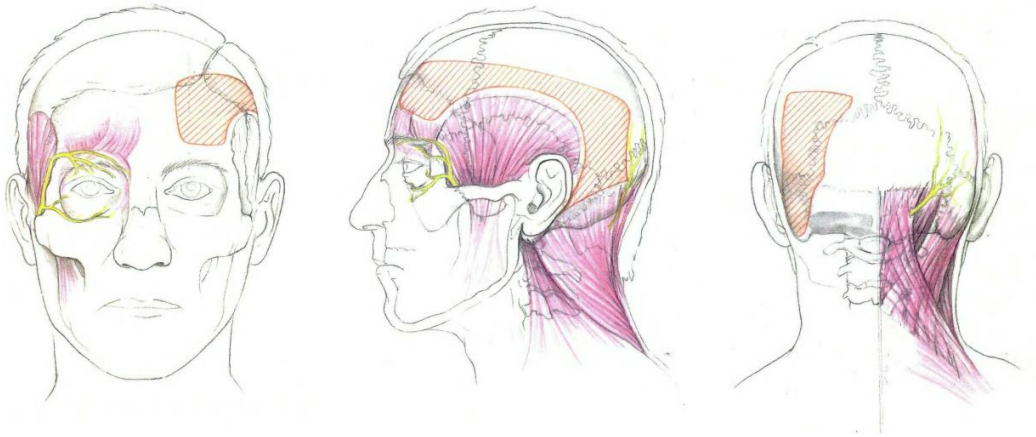
On convexity surface:

lower portion of precentral gyrus is located just posterior to pars opercularis;
lower portion of postcentral gyrus is just anterior to supramarginal gyrus

- gyral ridge (*yellow arrow*) connects precentral and postcentral gyri so that central sulcus never directly joins Sylvian fissure:



Safe areas for pinning:



MAYFIELD INFINITY PEDIATRIC HEAD HOLDER

- has integrated skull clamp and horseshoe headrest – adjustable height so head weight is distributed between pins (mainly serve as motion stabilizers) and head rest.
- uses lower (max. 18 lbs vs 80 lbs for adults) torque screw:

80-lbs Torque Screw

18-lbs Reduced Load Torque Screw

PEDIATRIC PINS

- have low profile not to perforate skull; suitable for kids ≤ 3 years (for older patients with thicker scalp, pin rim will press and necrose skin).

TUBULAR BRAIN RETRACTORS

Vycor

BrainPath

- has 13.5 mm diameter:
    a. does not disrupt white matter
    b. unable to use microscope (for binocular vision, channel must be at least 22 mm); thus, use exoscope
- do not use mannitol – brain turgor stabilizes retractor and pushed lesion into retractor's lumen.
- very limited dural opening – cruciate incision (each limb 1 cm)
- incise arachnoid over sulcus; push BrainPath (with regular Stealth probe inside obturator) through sulcus towards target, remove obturator.

May combine with **Myriad (NICO)** - Side-Cutting Aspiration Device: side mouth cutting (oscillating guillotine) and aspiration aperture.

ENDOSCOPE

10F peel away introducer sheath
continuous irrigation with warm (!!!) Lactated Ringer solution

Rigid:

Little Lotta (Karl Storz) – diameter 3 mm
Lotta (Karl Storz) – diameter 6 mm

- hemostasis may be achieved with monopolar cautery and vigorous irrigation (to verify hemostasis, pinch irrigation hose to stop irrigation – watch for blood wisp in CSF).

RHOTON

Rhoton Style Micro Dissection Set Components

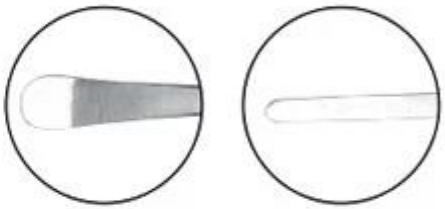
7½" (190mm) length

Dissectors

Catalog#	Tip Diameter	Style
MA-2626	1mm	#1
MA-2627	2mm	#2
MA-2628	3mm	#3

Elevators

Catalog#	Description	Tip Width	Style
MA-2629	Curved	2.6mm	#4
MA-2630	Angled	1.3mm	#5

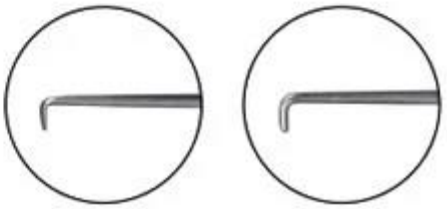


Spatula Dissectors

Catalog#	Description	Tip Width	Style
MA-2631	Small	1mm	#6
MA-2632	Medium	1.5mm	#7
MA-2633	Large	2mm	#8

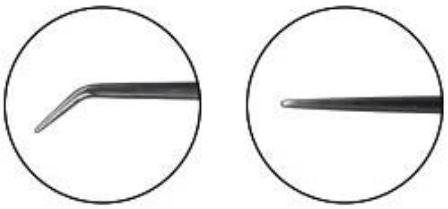
Right Angle Hooks

Catalog#	Description	Hook Length	Style
MA-2634	Semi-Sharp	2mm	#9
MA-2635	Blunt	3mm	#10



Needle Dissectors

Catalog#	Description	Cup Size	Style
MA-2636	Semi-Sharp, 45° Angle	3mm	#11
MA-2637	Semi-Sharp, Straight	5mm	#12

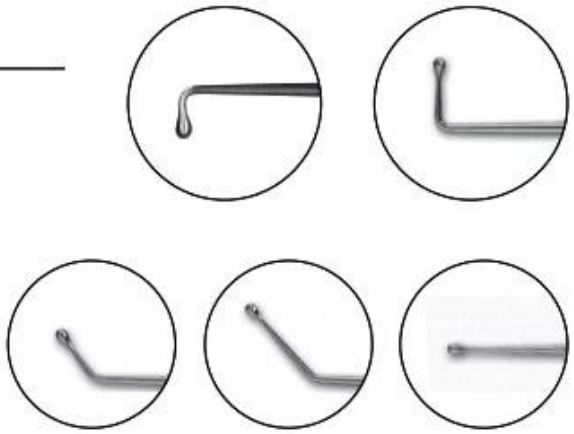


Micro Curettes

Catalog#	Description	Cup Size	Style
MA-2638	Straight	1 x 2mm	#13
MA-2639	Angled	1 x 2mm	#14

Ball Dissectors

Catalog#	Description	Tip Length	Style
MA-2640	90° Angle	3mm	#16
MA-2641	90° Angle	5mm	#17
MA-2642	45° Angle	4mm	#18
MA-2643	45° Angle	8mm	#19
MA-2644	Straight		#15



MALIGNANT HYPERTHERMIA

See >>

BRAIN BIOPSY

Nashold Biopsy Needle (Integra)

- side cutting window – 10 mm

Use CTA / MRA whenever available to plan trajectories away from vessels.

- possibility of tissue heterogeneity! - multiple biopsies along needle trajectory through entire tumor thickness (to the farthest tumor edge).

  N.B. sampling only central area (e.g. complete tissue necrosis in tumors or abscesses) may not yield diagnostic tissue!

major **hemorrhage** - risk 0-3% (0-12% in AIDS)

FORAMEN OVALE

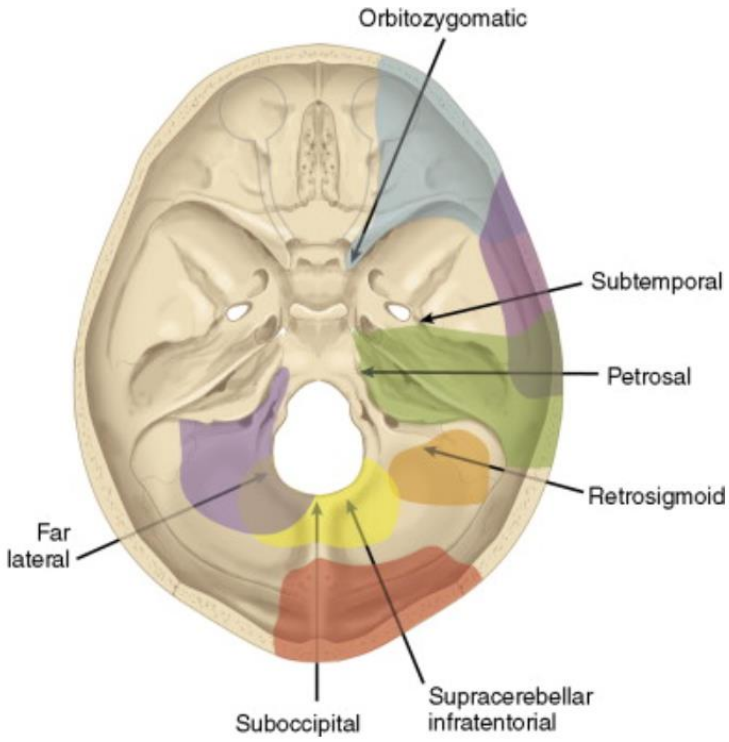
- 18 G 3.5 inch foramen ovale (vs. spinal) needle for cannulation
- second needle is 22G 5 inch spinal needle – wrap SteriStrip on shaft so tip protrudes 1.5* cm from first needle tip (*or whatever distance to tumor)
- guidance:

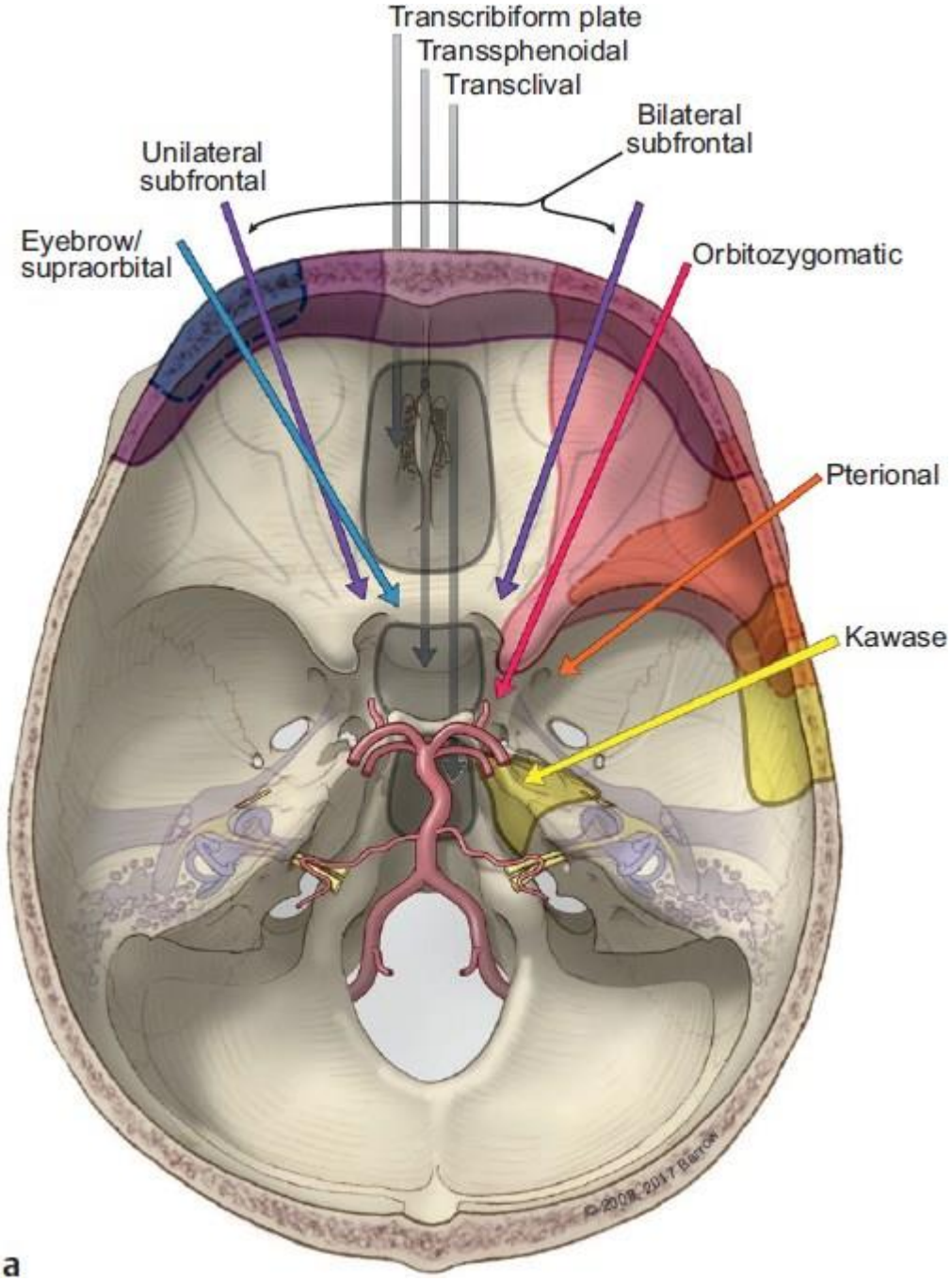
  a) frameless navigation, e.g. Stryker mask

b) fluoroscopy (C-arm) – work under **AP** with beam aligned along trajectory head rotated away from operated side – should clearly see foramen ovale (where top of petrous bone meets clivus)

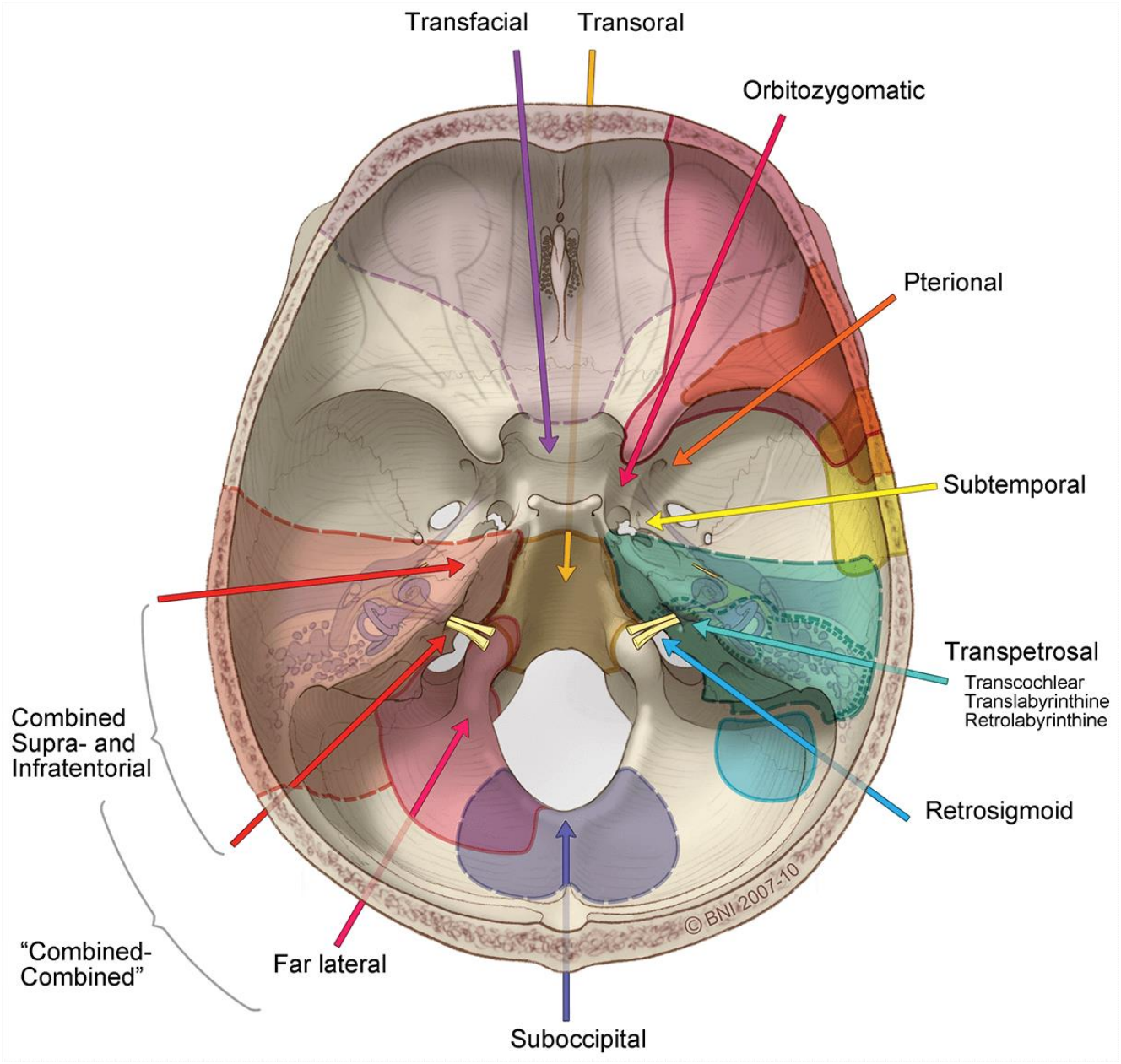
- **Härtel's landmarks**:
  - needle entry point – 2-3 cm lateral to mouth corner.
  - surgeon inserts **index finger in patient's mouth**; keep needle medial to coronoid process of mandible.
  - needle is aimed at inner aspect of ipsilateral pupil + at point 3.5 cm anterior to external ear canal at level of zygoma (practically, it is inverse EVD target).
- may feel ***mandible jerk*** when needle irritates CNV_3
- may cause ***bradycardia*** (have atropine ready), HTN
- change fluoroscopy to **lateral** to verify – needle tip has to be just posterior to clivus

SKULL BASE




Transcribiform plate
Transsphenoidal
Transclival
Bilateral subfrontal
Unilateral subfrontal
Eyebrow/ supraorbital
Orbitozygomatic
Pterional
Kawase

a


Transfacial
Transoral
Orbitozygomatic
Pterional
Subtemporal
Transpetrosal
Transcochlear
Translabyrinthine
Retrolabyrinthine
Retrosigmoid
Combined Supra- and Infratentorial
"Combined-Combined"
Far lateral
Suboccipital
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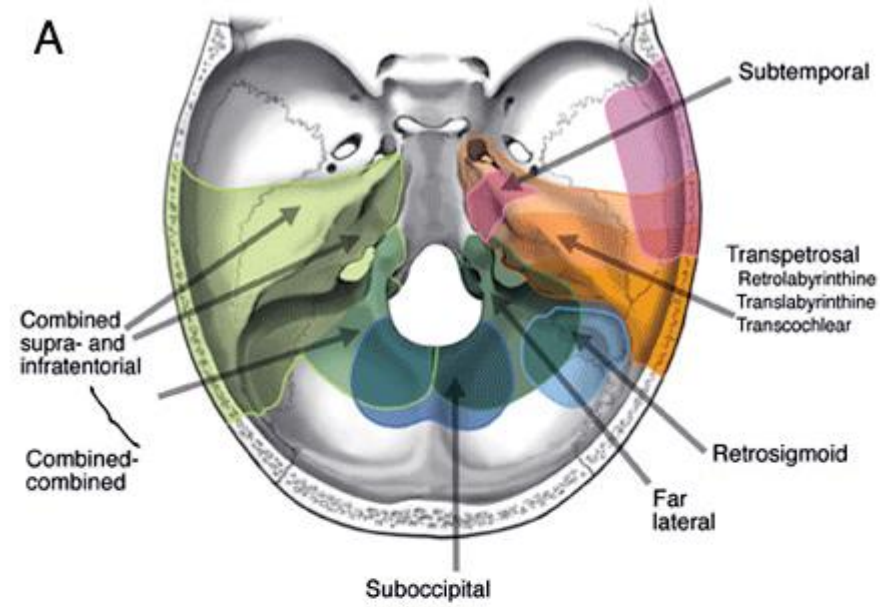
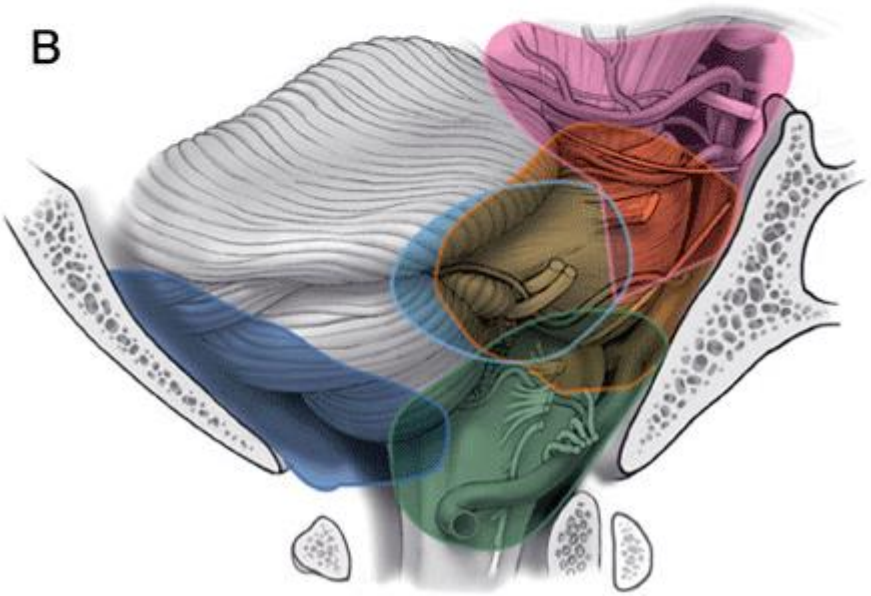
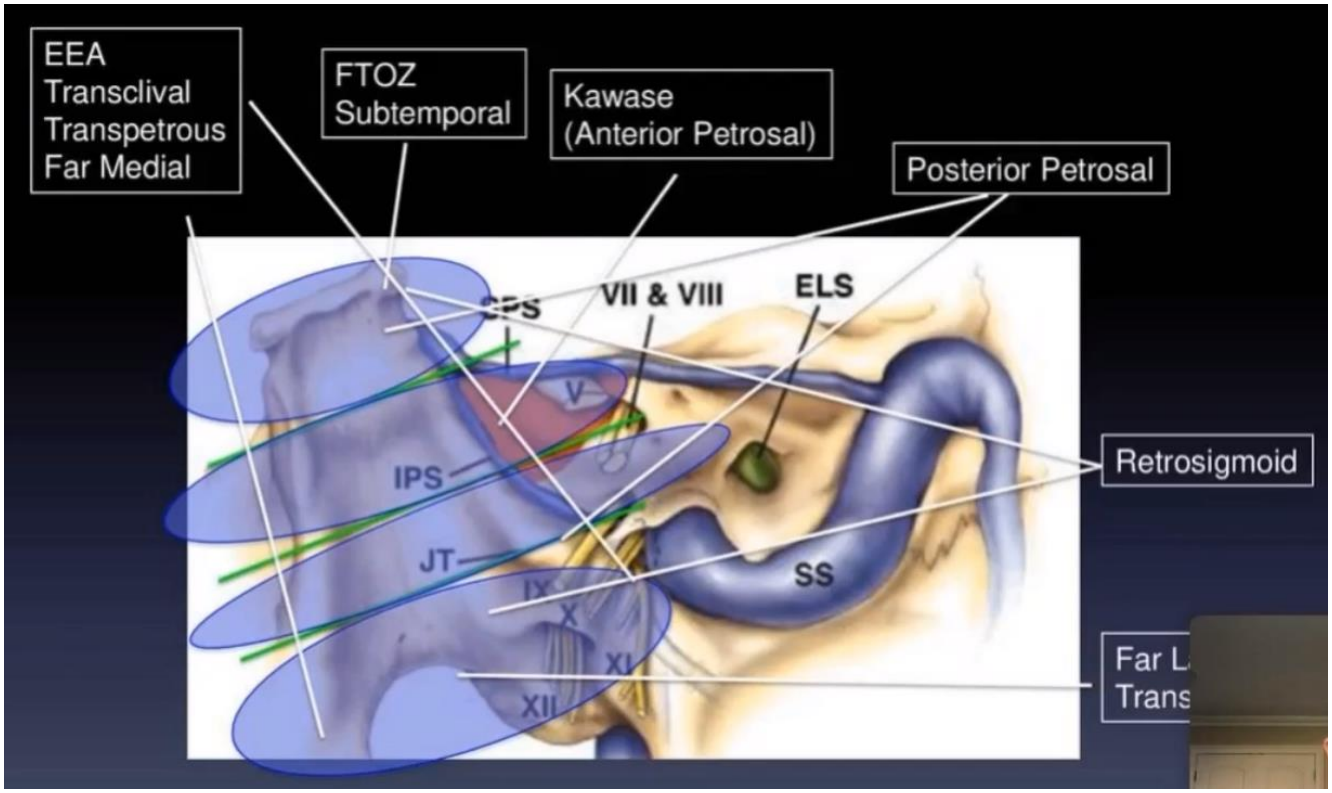

A
Subtemporal
Transpetrosal
Retrolabyrinthine
Translabyrinthine
Transcochlear
Combined
supra- and
infratentorial
Combined-
combined
Retrosigmoid
Far
lateral
Suboccipital


B



- for anterior skull base repair may use titanium mesh, pericranium flap, galea flap, temporalis muscle rotated on vascular pedicle; postop may use "airflow diversion" (tracheostomy, intubation, or at least nasal airway for 3 days – all help to prevent soft tissue, used for reconstruction, movement when patient is breathing / coughing / sneezing).

MESIAL TEMPORAL REGION

- divided into 3 areas:

1. **Anterior** - transsylvian-transinsular approach
2. **Middle** - transtemporal approach
3. **Posterior** - supracerebellar-transtentorial approach

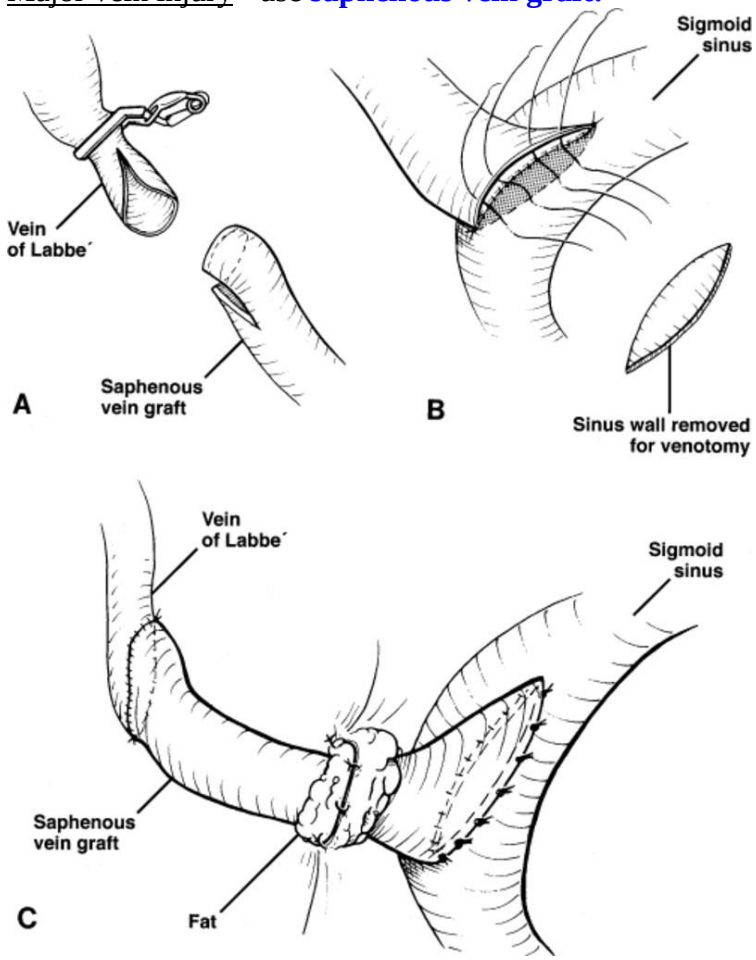
POSTERIOR FOSSA

A. Suboccipital craniotomy >>
   Supracerebellar approach >>
B. Retrosigmoid (retromastoid) >>
   Far Lateral approach >>
C. Presigmoid (in order of increasing temporal bone drilling):
   a) retrolabyrinthine
   b) translabyrinthine
   c) transcochlear
D. Transoral Clival approach >>

DURAL VENOUS SINUS INJURY

- before elevating fracture over sinus ***notify anesthesia*** (risk of bleeding + air embolism) and have large piece of Gelfoam and **rapid infuser** ready.
- if bleeding is too brisk and finger pressure blocks view for repair → proximal sinotomy to allow temporary placement of inflatable **Fogarty balloon catheter**.
- if interposed graft is necessary - ***autogenous saphenous vein graft*** may be utilized after temporarily shunting blood through interposed shunt to allow time to sew graft into place.
- anterior 1/3 (or 25%) of SSS can be sacrificed!

- bleeding from **CAVERNOUS SINUS** – inject fibrin glue into it!
- Medium tears can be managed by placing Gelfoam+Surgicel, over opening.
  TACHOSIL® – fibrin sealant patch (sticks to sinus wall!)
- Larger tears (esp. if contralateral flow is not patent or nondominant on preoperative imaging):
  - ***drill bone around*** (keep sinus covered with patty + assistant may place fingers on sinus proximal and distal to sinus injury to apply pressure).
  - repair sinus wall using:
    a) **muscle piece** plug
    b) **pericranial flap**
    c) **dura flap** – cut C-shape adjacent dura flap (based on dural sinus) and reflect it over sinus wall defect – suture in place using interrupted 4-0 silk stitches, cover with Surgicel wrapping Gelfoam; may perform ICG angiography to confirm sinus patency
    d) **Gore-Tex graft**
    e) bypass
- Major vein injury - use **saphenous vein graft:**



TENSION PNEUMOCEPHALUS (POSTOP)

- uncommon but potentially serious complication (lumbar drain may be the etiology)

- progressive headache and mental status change.
- treatment:
  1) **head of bed** is kept flat
  2) **oxygen** by face mask.
  3) ***surgery*** is rarely necessary

AIR EMBOLISM

- results from non-collapsible vein (dural sinus, diploe) injury
  Detected 100% (when TEE is used) in cases in sitting position
- rarely may lead to pulmonary or cerebral air embolism.
- lethal amount – 200-300 mL of air (3-4 mL/kg)

PREPARATION

- when there is a risk of entering into venous sinus (fracture over sinus, supracerebellar-infratentorial approach):

Preop
- **transthoracic Doppler echocardiography** to detect any right-left shunt (e.g. patent foramen ovale) ← contraindication for the sitting craniotomy – risk of brain air embolism.
- **MRV** - check patency of contralateral venous sinuses.

Intraop (standard measures)
- **ECG** (ventricular arrhythmias)
- **end-tidal CO_2** monitoring (↓ along with BP↓, pO2↓, CVP↑)
  normal end-tidal CO_2 5-6% = 35-45 mmHg

Intraop (special measures)
- place **precordial Doppler ultrasonography** - early sensitive intraoperative detection of intracardiac air: AE is heralded by a change in sonic intensity and character ("machinery sounds").
- place **multichannel central venous catheter** (in Right atrium) to aspirate any large emboli.

MANAGEMENT

1) occlude site of air entry - soaked Raytec and ***flood with saline, wax bleeding bone edges***, ***occlude visual sources of venous bleeding***.
   Do not pack sinus – will cause thrombosis (maybe disastrous)
2) lower head if at all possible (30° or less from horizontal)
3) rotate patient LEFT side down (attempt to trap air in right atrium)
4) jugular venous compression by anesthesiologist (**bilateral** best; second choice: right only) – increases venous pressure
5) aspirate air from right atrium via CVP catheter
6) ventilate with 100% O2 + discontinue nitrous oxide (may expand AE)
7) if *patent foramen ovale* exists, stop PEEP (PEEP increases R-to-L intracardiac shunting)

- if nothing helps, terminate surgery.

ENTRY INTO FRONTAL SINUS

If entry is small, mucosa intact
- carefully remove sinus mucosa from ***bone flap*** pockets; leave mucosa intact in sinus → cover with vascularized pericranial flap → Tisseel / DuraSeal.

If mucosa violated, inflamed (sinus is no longer sterile)**, posterior wall fracture** – need **sinus cranialization- exenteration**: remove posterior wall of frontal sinus, pack with muscle plug, cover ostia with pericranial flap over, fibrin glue

AWAKE CRANIOTOMIES, BRAIN MAPPING

General anesthesia is preferred for most patients with ***minimal to moderate motor and/or sensory deficits*** and with lesions outside language regions.
Language function can only be assessed in awake patients.
Awake craniotomy is considered for cooperative patients with ***more severe motor or sensory deficits*** (must be motor at least 3/5)

cannot use paralytic agents!

awake craniotomy - currently reserved only for speech area testing with bipolar stimulation (while patient is counting or naming objects);

asleep (LMA)-awake-asleep craniotomy - *may compromise electrophysiological brain mapping* and thus endanger patient's neurological outcome

Alternative - subdural grid electrodes and extraoperative functional mapping.

Craniotomy - at least 2 cm of the cortex around tumor perimeter – to expose functional cortices.

INDICATIONS

Low-grade* gliomas in or near **motor**, **somatosensory**, **language** cortex, **thalamus**, **brainstem** – check MRI, fMRI, DTI (guide mapping to be more targeted):

1) Broca's and Wernicke's cortices, arcuate fasciculus
2) rolandic cortex, supplementary motor area, corona radiata, internal capsule
3) insula, uncinate fasciculus

*high-grade gliomas – only near (but not in) eloquent areas

- language mapping – preop fMRI to determine hemisphere dominance; object naming, at 4 sec per image, must be better than 75%.
- motor mapping requires (near) normal power (at least 4-/5 for mapping under general anesthesia)
- somatosensory mapping requires (near) normal sensation.

Lesions that do not need mapping* - well-defined (noninfiltrative) lesions: metastases, vascular.

*unless need to find a safe corticotomy site

Dr. Cohen-Gadol – strategy for HGGs:

asleep mapping - if the tumor *does not directly infiltrate* but is anatomically within millimeters of these vital structures.
awake mapping - for tumors (low-grade gliomas) that *directly infiltrate* these structures as shown on MRI + fMRI (esp. postcentral gyrus - in awake mapping leads to subjective paresthesias that are not detectable in anesthetized patient)

N.B. if tumor seems to involve eloquent areas but patient becomes asymptomatic on steroids – means symptoms were coming from edema and tumor is not invading – likely resectable! vs. high-grade infiltrative tumors in eloquent cortex – attempt to resect leads to STR (deficit and hematoma risk↑)

HGGs inside eloquent cortex – no surgery! (i.e. GBM not improving on steroids – beware surgery)

CONTRAINDICATIONS

1. Obesity, sleep apnea, airway problems (e.g. chronic cough, severe GERD)
2. History of anesthesia emergence delirium
3. Psychiatric issues (esp. anxiety, claustrophobia, cognitive impairment)
4. Uncontrolled seizures
5. Children < 6-10 years:
    1) not well tolerated

2) cortical stimulation mapping may not elicit motor responses (decreased cortical excitability).

6. Language baseline errors > 25% of objects named correctly at 4-second intervals.

Absolute contraindications - patient refusal and inability to cooperate (confusion, decreased level of consciousness, moderate to severe dysphasia or aphasia, language barrier) or inability to lay still.

Challenging pathologies: large lesions with marginal intracranial compliance, deep-seated tumors requiring significant surgical retraction, highly vascular lesions.

PREPARATION

REMIFENTANIL / PRECEDEX (helps with stressful shivering) / PROPOFOL
MANNITOL IV - maximum 0.5 g/kg → nausea and vomiting
good ***field block*** (**preauricular**, **postauricular**, **supraorbital**, and **occipital** nerves) + pin sites + incision

- Foley catheter is inserted.
- **Mayfield pin head holder** (*use 80 lbs for awake patients* to avoid pin slip); alternative – use **AxiEM** or **Neuro FrameLock** systems without pinning.
- patient comfort is paramount – should be able to swallow, pillows under knees and/or back, warming blanket, video monitors displaying operative field turned away from patient's line of sight.
  - **supine**, unless mapping Wernicke area – **lateral** position
- to facilitate any potential emergency airway rescue, mindful surgical positioning is important to avoid extreme rotation of the head and neck, and to leave adequate space for lower chin mobility to allow full mouth opening.
- before bone flap is elevated, ***local anesthetic is injected through burr holes*** to irrigate sensitive dura.
  - once bone flap is elevated, dura should be ***blocked around middle meningeal artery***.
  - subtemporal craniectomy using bone rongeurs should be avoided - can be very painful!
- *dura is not opened until the patient is completely awake and calm*.

TECHNIQUE

- Ojemann Cortical **bipolar Stimulator** + EEG machine + epileptologist
- paper tabs, 4-0 silk to outline cortex where lesion is – start stimulating the area in and around.
- seizures during cortical stimulation:
  1) mapping is aborted
  2) brain is irrigated with **iced irrigation fluid (e.g. Ringer)**
  3) increasing **supplementary oxygen** concentration;
  - if a seizure persists, small boluses of PROPOFOL (0.5-1.0 mg/kg) can be given;
  - ALTERNATIVELY, MIDAZOLAM* is administered in 1- to 2-mg increments until clinical seizure activity ceases; another alternative - METHOHEXITAL (Brevital).
  - it is very rare to evoke seizures that persist or become problematic for continued mapping.
  - if a seizure persists, **airway should be placed** and seizures stopped with **AED**.

*may not be optimal - slower clearance and possible interference on function testing

After Discharge = focal seizure

- strip electrode is placed nearby stimulation site (e.g. under craniotomy edge for the duration of mapping) - used to detect afterdischarge potentials.
- begin with 2 mA current → increase by 1-2 mA increments until ADs are elicited (this is AD threshold, usually never > 6 mA) or up to 6-10 mA (vs. asleep can go up to 16-20 mA).
- *current 1-2 mA below AD threshold is used for mapping*

- subcortical mapping needs 1-2 mA higher current.
- frequency of 1-**60** Hz (60-100 Hz for language mapping) at 1 msec pulses for 2-3 seconds
  HF stimulation - more versatile and safe approach in comparison to LF, with less risk of intraoperative seizures (< 2% even in difficult cases) and almost no chance of false negative mapping

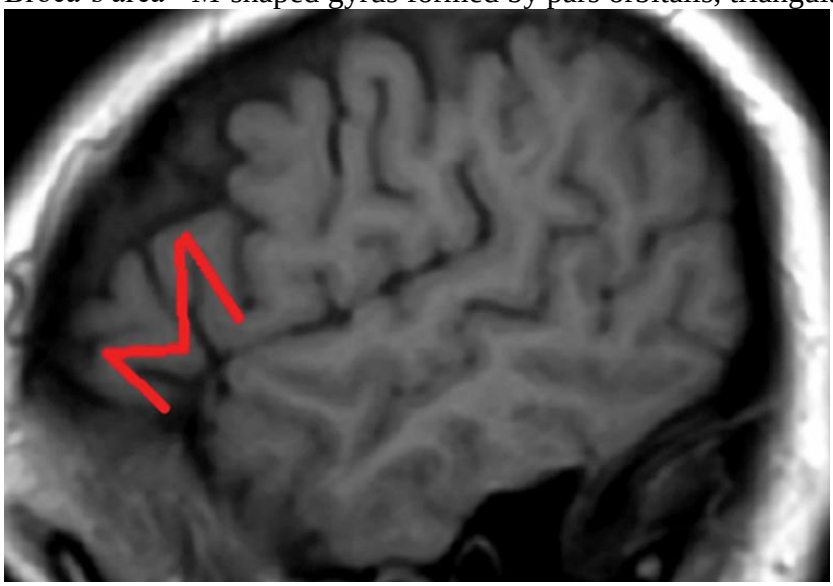
N.B. each cortical site is checked 3 times to ensure accuracy; avoid *stimulating the same area repeatedly* without a pause to prevent seizures.

+ place cottonoid pledget with 0.5% LIDOCAINE without epinephrine over cortex for 5 min - if there is no interference with task, area may be safely resected.

- corticotomy in safe area → subpial dissection with subcortical mapping.
- bipolar cautery is used sparingly, if it all, to avoid injuries to en passage small vessels (if vasospasm – apply PAPAVERINE on a small patty).

Language mapping

Broca's area - M-shaped gyrus formed by pars orbitalis, triangularis, and opercularis:



- only PROPOFOL and/or DEXMEDETOMIDINE - facilitate mild sedation during opening and closing, and maximum cooperation during mapping and resection phases; no narcotics or additional anesthetic medications!
- computer slide show with 50-100 object drawings, presented every 3-4 seconds.
- neuropsychologist to assist.
- while patient is performing language task, current is applied to cortex just prior to display of object, continuing until task is performed correctly or next task appears.
- stimulation sequence: face motor → Broca (speech arrest without any movement in oropharynx) → Wernicke.
- subdural grids are best - adjacent electrodes monitor afterdischarge potentials to exclude false positives.
- many people have **multiple language areas** - finding one language area does not mean that mapping is complete - the entire area at risk should be mapped.
- in **multilingual patients**, ***each language*** must be mapped.
- if surgeon cannot find language area, patient should continue object naming throughout resection.
- resections are kept a minimum of 1 cm from positive language sites (sulcal patterns may warrant modification).

Two ways of language mapping:

a) **fMRI** and/or **passive ECoG** – show critical and participating cortex

b) **ESM** (electrocortical stimulation mapping) – delineates critical cortex – gold standard to guide resections.

Language tasks:

a) naming objects presented for a 3-4-second duration (rehearsed preoperatively with the neurophysiologist) – for Wernicke
b) counting while intermittently stimulating – for Broca
c) following simple and complex commands – for Wernicke
d) repetition.

Sensory mapping

- **asleep** – strip or grid electrode recording SSEP (no seizure risk as cortex not stimulated)
- **awake** - perceived as tingling, vibration.
- cortical deficits improve significantly over time, but fine hand movement and gait will be permanent.

Motor mapping

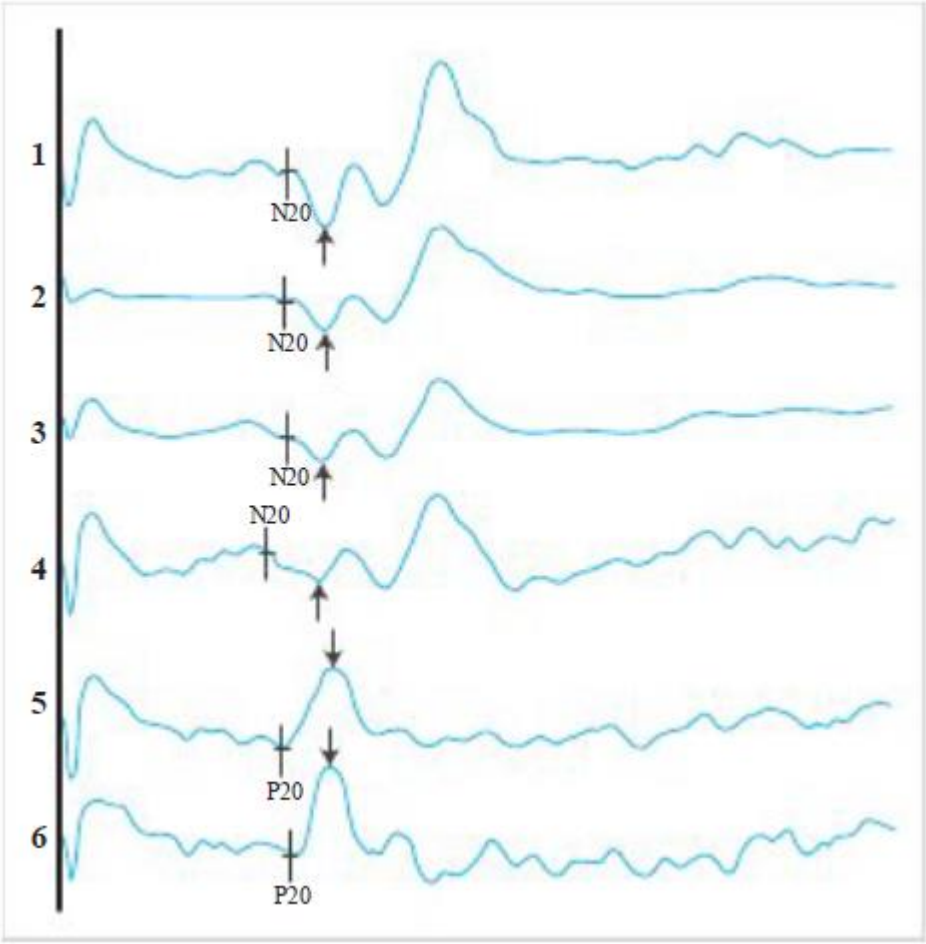
- **asleep** using EMG (check TOF) - evoked contractions of muscle groups (EMG - progressive motor unit recruitment).
- **awake** – evoked contractions of muscle groups → ask to move extremity, stop resection when begins difficult to move (slight deficit - "**BUSY LINE EFFECT**") – allows more radical resection (than asleep!)

  structures involved in a higher level of motor programming (eg, grasping networks) can be mapped only in awake setting - advanced awake mapping should be considered to fully maintain high skilled movements!
- resection just anterior to motor cortex → transient supplementary motor area (SMA) syndrome
- when resecting in motor area, resect lesion in motor cortex → SMA (not the opposite).

Dr. Komotar uses 5 mA bipolar for **cortical**, 5 mA monopolar for **subcortical** mapping

Nondominant face motor cortex may be removed → only temporary facial weakness → complete return of facial function as long as the underlying white matter tracts for the adjacent motor cortices were left intact.

Phase reversal method for localizing central sulcus under general anesthesia: 8-contact recording strip is placed perpendicular to the anticipated central sulcus → SSEP-type stimulation of contralateral median or tibial nerve: phase reversal of N20/P20 peak between a pair of electrodes (# 4 and 5 in example below) indicates that those electrodes straddle the central sulcus:



MULTISTAGE SURGICAL APPROACH

1) incomplete tumor resection during the first operation (to preserve a perfect QoL) → postoperative functional rehabilitation to induce neuroplasticity.
2) reoperation a few months or years later with increase EOR (so without any loss of chance from oncological perspective) while again avoiding any permanent neurological deterioration.

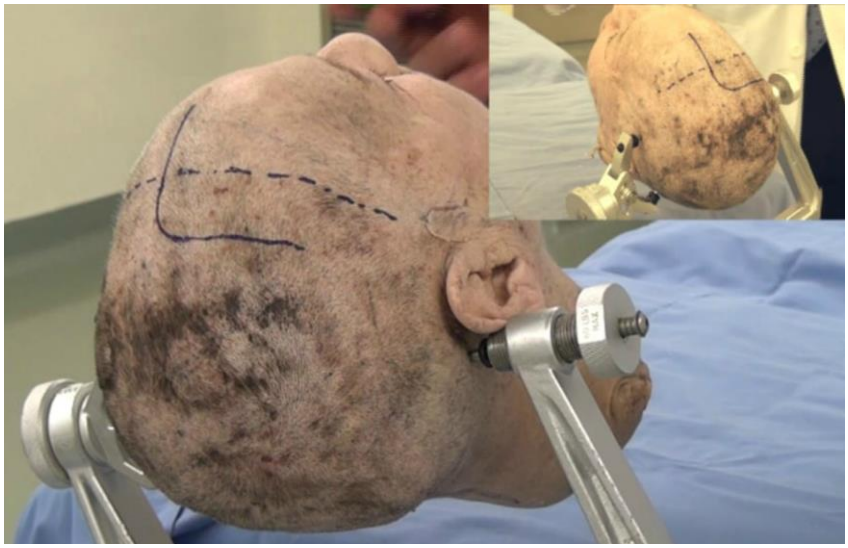
See Case F1 >>

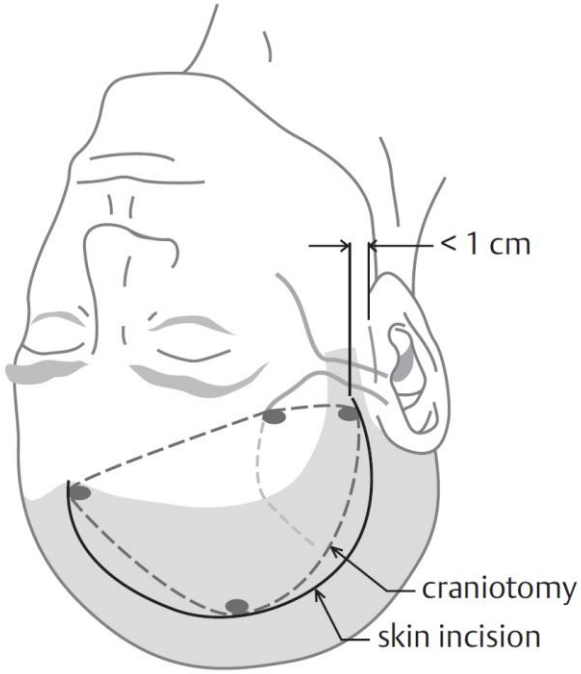
TRANSNASAL ENDOSCOPIC ACCESS TO ANTERIOR CRANIAL FLOOR

Modern – vascularized nasoseptal flap; fed by sphenopalatine artery.

- 48 hrs cefazolin IV (for extended flaps – triple antibiotics).
- **JANUS flap** – bilateral flaps (so each flap can be smaller).

FRONTAL (UNILATERAL) CRANIOTOMY



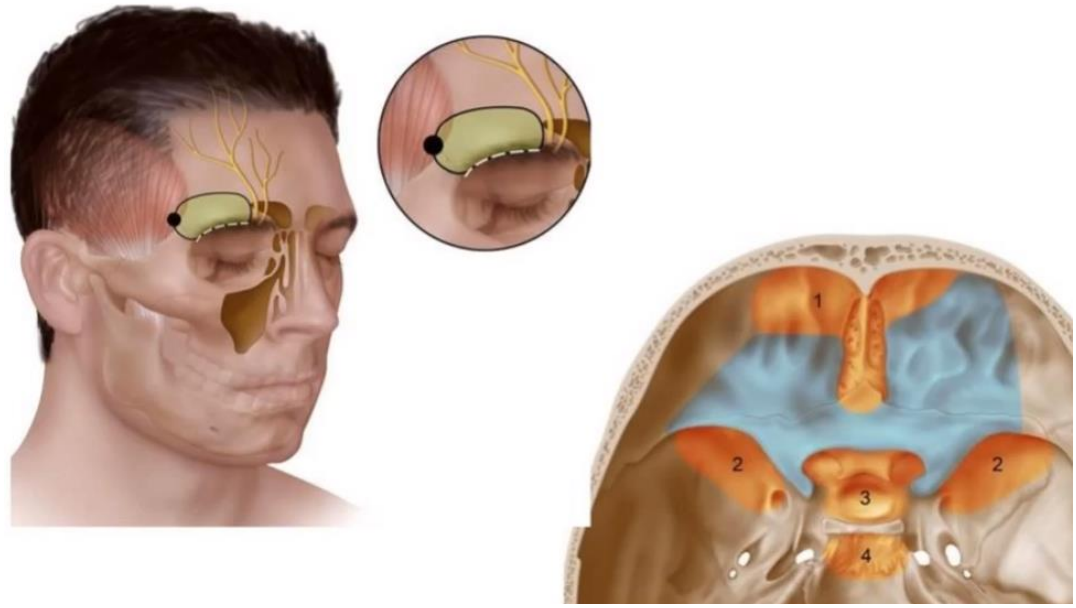


Incision:

A. L incision – for posterior lesions
B. Keyhole supraorbital incision
C. Incomplete (3/4) bicoronal incision

SUPRA-ORBITAL CRANIOTOMY (S. "EYEBROW" CRANIOTOMY)

- incision within the eyebrow (not above eyebrow!) – perfect cosmesis.

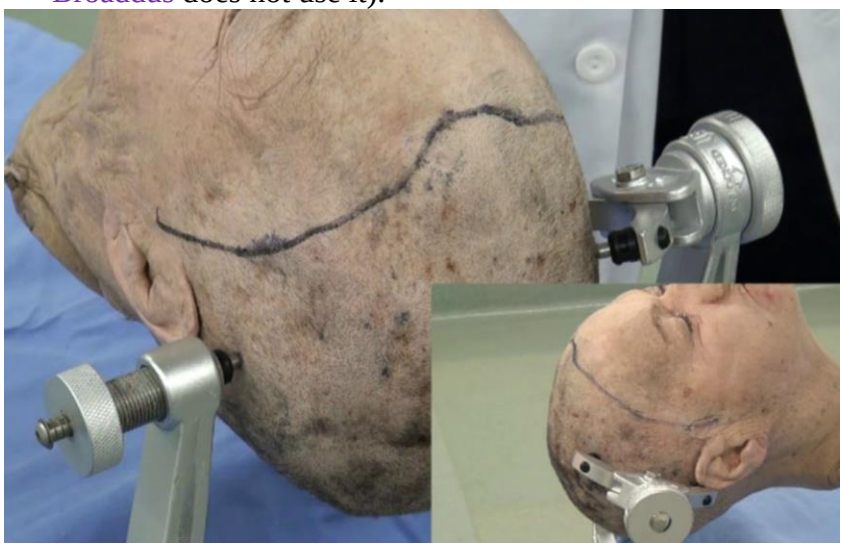


- areas in blue can be seen during craniotomy; areas in orange cannot (H: add endoscope).

BIFRONTAL CRANIOTOMY

Pin placement – as posterior as possible.
Incision:

- starts 1 cm anterior to tragus (in skin fold) at level of zygoma (or above)
- extends vertically up, curves anteriorly (always stays behind hair line); lazy omega at midline (Dr. Broaddus does not use it).



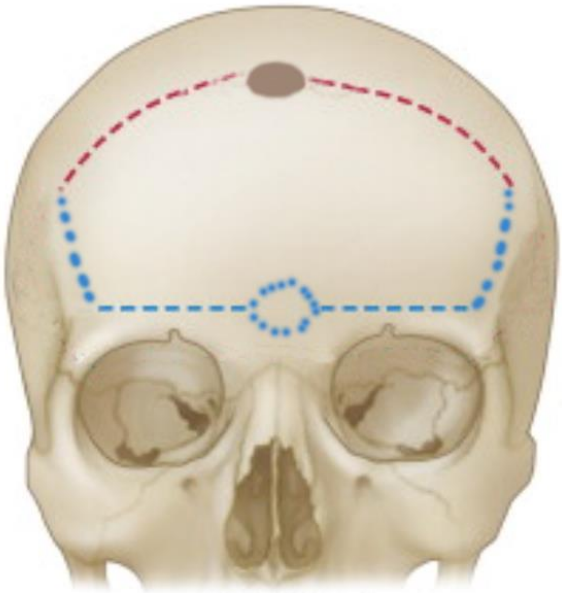
SUBFRONTAL CRANIOTOMY

- ideal for olfactory groove / planum sphenoidale meningiomas
- full or ¾ bicoronal incision
- make pericranial flap.
- need to incise temporalis fascia to lift it up together with scalp flap (in order to preserve frontalis branch); Dr. Broaddus leaves temporalis fascia untouched and only dissects scalp off.
- bur holes:
    a) ***posterior over sagittal sinus***: Dr. Broaddus - one bur hole right over sinus; Dr. JRC - two small bur holes posteriorly on the sides of SSS (uses matchstick to connect bur holes)

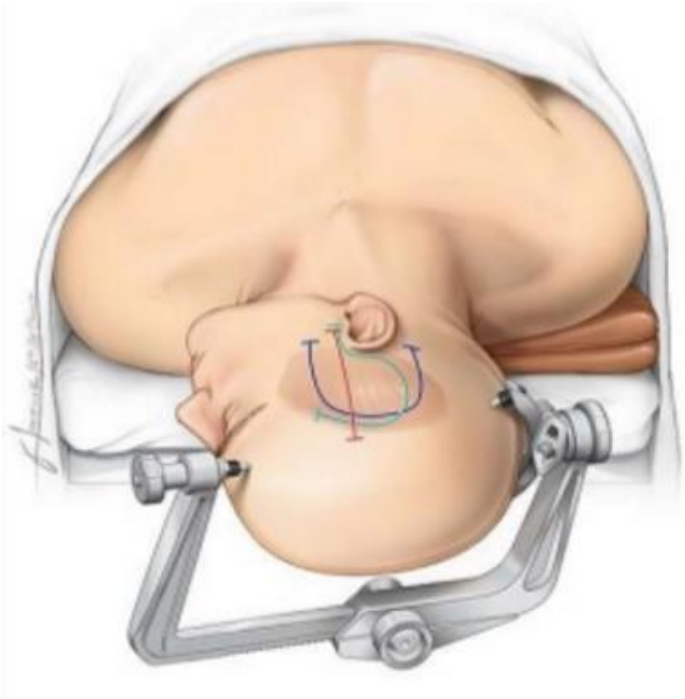
b) ***anterior midline burr hole*** (just above frontonasal suture); often frontal sinus is thick and impossible to cut with footplate, plus, risk of SSS damage, so Dr. Broaddus uses C drill bit to remove circle of bone in anterior wall of frontal sinus (at the end places bone circle back and fixes with dogbone) → use Acorn, Kerrison to go across posterior wall of frontal sinus.

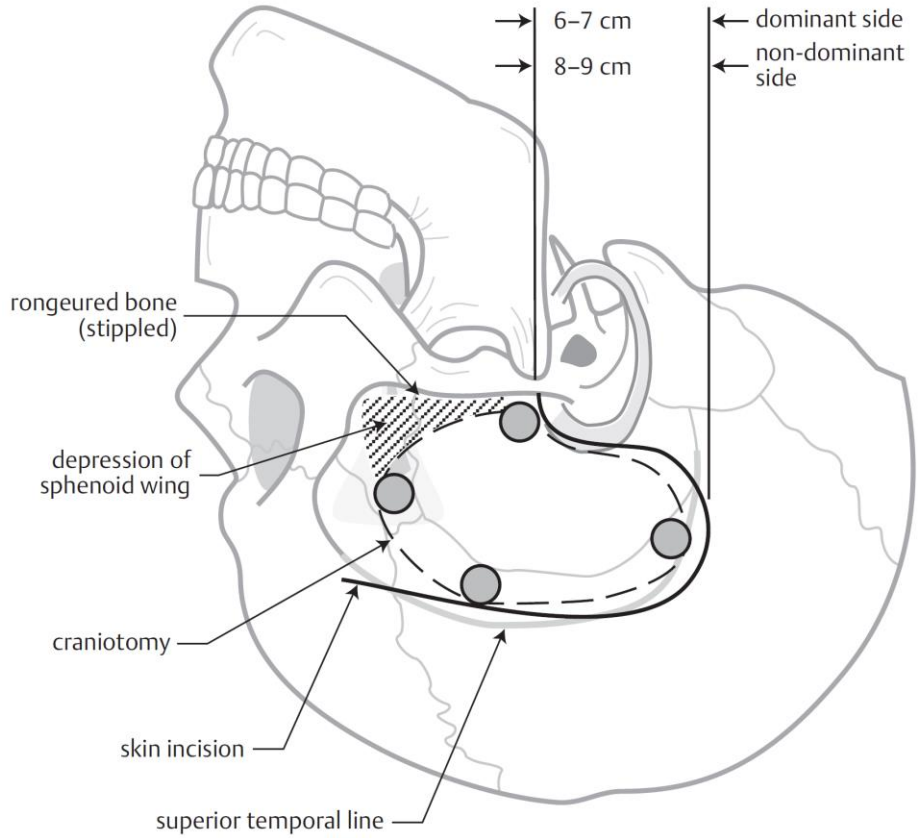
c) some experts place bur holes ***at keyholes*** – make large bone flap.

- sagittal sinus is 2-0 silk suture-ligated and transected (along with falx) at the base.
  - no need to reattach falx at the end of surgery
- anterior dura is stripped from remaining frontal bone bar and the bone is flattened.
- at the end lay pericranium on denuded anterior fossa floor.

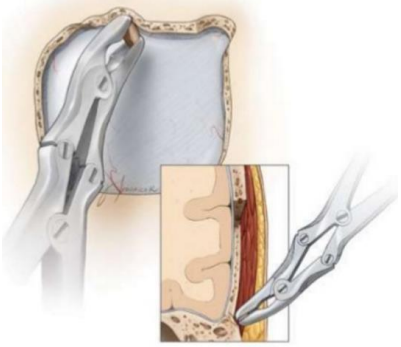


(SUB)TEMPORAL CRANIOTOMY



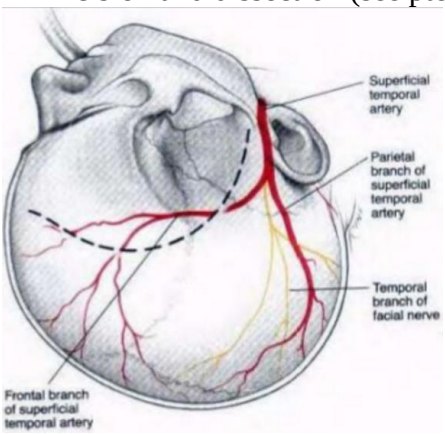


- biting off temporal bone:



ENCEPHALOCELE REPAIR (MIDDLE FOSSA)

- use navigation – mark location of bone defect + location of rotundum and ovale.
- incision and dissection (see pterional craniotomy):



- may use 3 cm craniectomy hole (instead of craniotomy) low on temporal squama.
- stay ***extradural*** and find bony defect (use navigation).

- amputate herniated brain fragment.
- repair dural defect: open slit of dura more proximal to you, slide intradurally dural matrix patch and cover dural defect from inside → suture close dural defect and then your durotomy.
  N.B. durotomy does not need to extend to the actual dural defect; it only serves to introduce dural matrix to inlay on dural defect (which is often small).
- close bone defect: (dural matrix patch) – bone graft (e.g. from craniotomy) – titanium (either mesh or snowflake) – dural matrix.
- mesh to cover craniectomy defect.

PTERIONAL CRANIOTOMY

- microscope: observer tube to operator's right for either right or left pterional crani.

INDICATIONS

1. Aneurysms
   a) all aneurysms of anterior circulation
   b) basilar tip aneurysms
2. Approach to cavernous sinus
3. Suprasellar tumors
   a) pituitary adenoma (large suprasellar component)
   b) craniopharyngioma

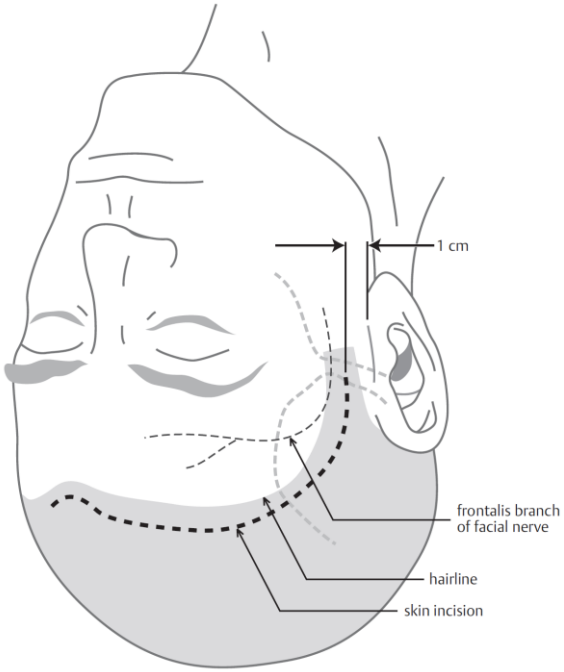
POSITION

- supine, ipsilateral shoulder roll if head rotated > 30°
- elevate thorax 10–15°: reduces venous distension
- flex knees
- neck extended 15°: allows gravity to retract frontal lobe away from skull base
- head rotated:

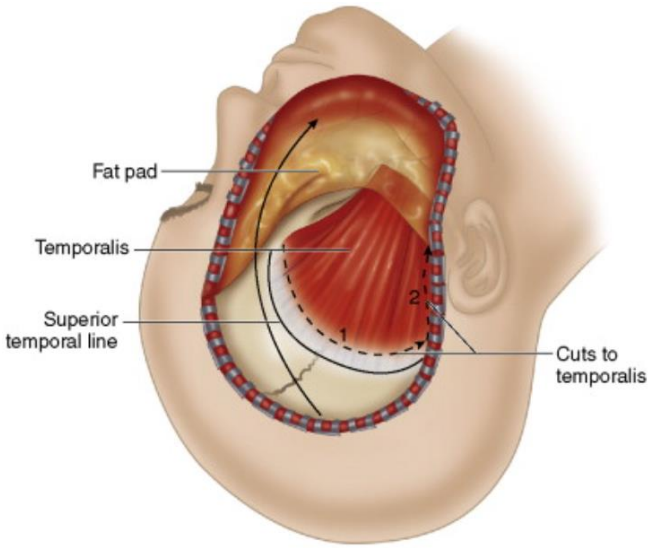
30° from vertical - for posterior exposure: PComA / carotid terminus / basilar bifurcation aneurysms
45° from vertical - for middle exposure: MCA aneurysms
60° from vertical - for anterior circle of Willis exposure: AComA aneurysms, suprasellar tumor

INCISION

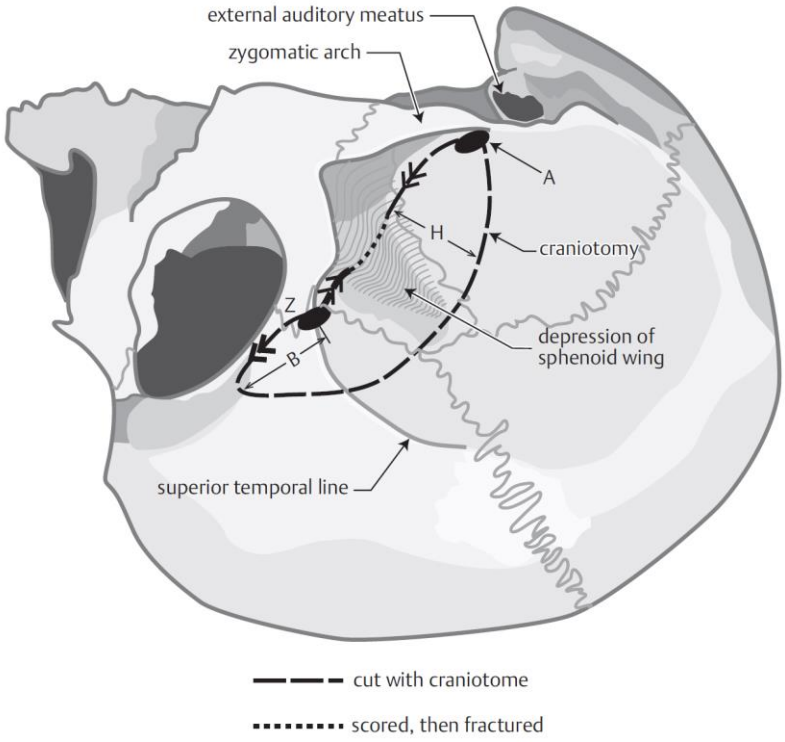
- from zygomatic arch 1cm in front of tragus (to avoid frontalis branch of facial nerve and frontal branch of superficial temporal artery), curving slightly anteriorly, staying behind hairline to widow's peak; optional additional curve beyond midline to aid in skin retraction.
- over temporalis muscle, incise skin down to but not through temporalis fascia.
- temporalis muscle incised in-line with the skin incision.



- once subgaleal fad pad is reached (that is where frontalis branch sits, usually in the anterior 1/3 or 1/2 of zygoma arch), incise superficial temporal fascia (interfascial dissection in the intrafascial fat pad) or also deep temporal fascia (subfascial dissection).
- some experts lift myocutaneous flap.
- when lifting temporalis muscle off the bone, do not use Bovie (although it bleeds) – innervation to muscle comes in the deepest layers; Dr. JRC uses retrograde fashion (Japanese technique)



CRANIOTOMY



- <u>two burr holes are sufficient</u>:
  - one burr hole is made at posterior insertion of zygomatic arch ("A"), as far caudally as possible to minimize amount of bone to be rongeured off to gain access to the floor of middle fossa.
  - second burr hole ("Z") is made at the intersection of zygomatic bone (near frontozygomatic suture), superior temporal line, and supraorbital ridge - hole should be as low as possible on the orbit (but aim drill slightly superiorly to avoid actually entering the orbit).
- bone flap is centered over ***depression of sphenoid ridge***.
- starting at frontal burr hole, craniotomy is taken anteriorly - across anterior margin of superior temporal line, staying as low as possible on the orbit:

  distance "B":
  a) anterior circulation aneurysms - 3 cm
  b) approaches to skull base (e.g. Dolenc) – larger, to ≈ mid orbit.
- from point "B," sharp superior turn is made and the opening is taken back to point "A."

  width of craniotomy ("H"):
  a) ≈ 3 cm for aneurysms of the Circle of Willis
  b) ≈ 5 cm for MCA aneurysms.
  c) tumors require large flaps ("H" is made larger to expose more temporal lobe).
- bone between two points is scored with craniotome → bone is fractured at this point.
- rongeur is used to remove as much sphenoid wing as possible.
- dural flap - curvilinear, centered over sphenoid wing, retracted inferiorly with dural stitch.

SYLVIAN FISSURE DISSECTION

- for MCA aneurysms, for Yasargil approach to basilar tip aneurysms, for insular tumors - Sylvian fissure needs to be split.

  N.B. prep neck for proximal control if going to split fissure!

EXTENDED PTERIONAL CRANIOTOMY

- aggressive drilling of roof of the orbit + lateral sphenoid wing + temporal bone down to floor.

ORBITOZYGOMATIC CRANIOTOMY (OZ)

- further expansion of **extended pterional approach** through osteotomy of various sections – removal of orbital rim, anterior orbital roof, and frontal process of zygoma → broadens the subfrontal trajectory and minimizes frontal lobe retraction to access the floor of the anterior and middle skull base as well as parasellar and interpeduncular spaces.

Add lumbar drain!

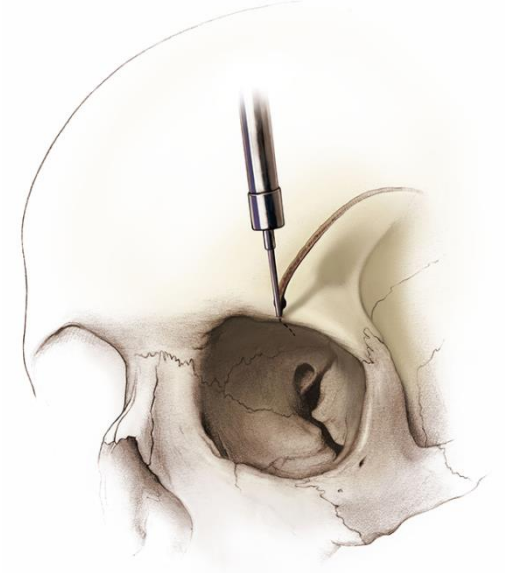
TWO-PIECE

first **traditional pterional** craniotomy → **supraorbital osteotomy**.

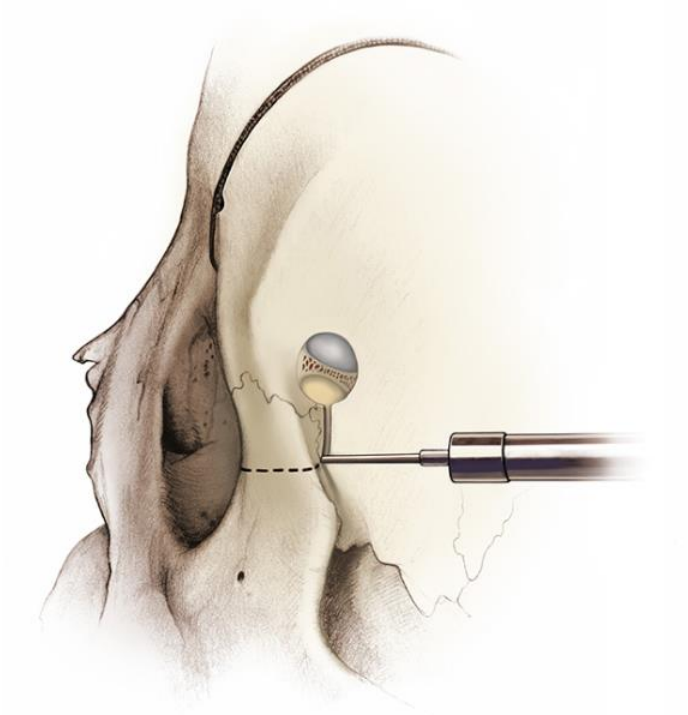
ONE-PIECE FRONTOTEMPORAL CRANIOTOMY AND SUPRAORBITAL OSTEOTOMY (MODIFIED OZ)

the least disruptive.

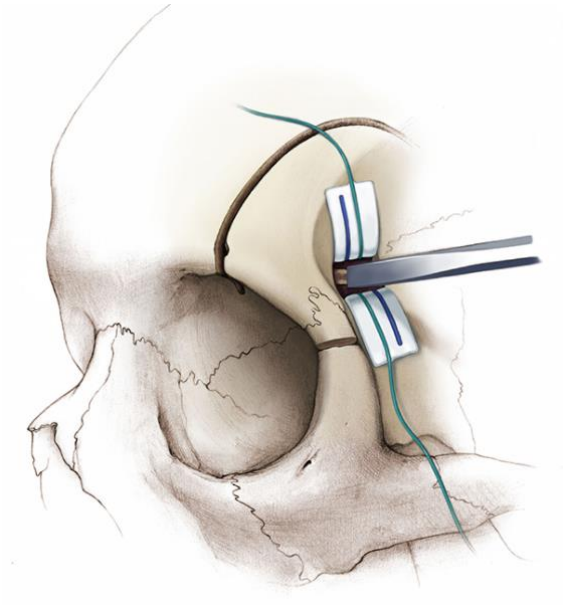
First osteotomy - cuts across the orbital rim:



Second osteotomy - disconnects the frontal process of the zygoma
(complete zygomatic osteotomy of the temporal portion of the zygomatic arch does not significantly add to the exposure)



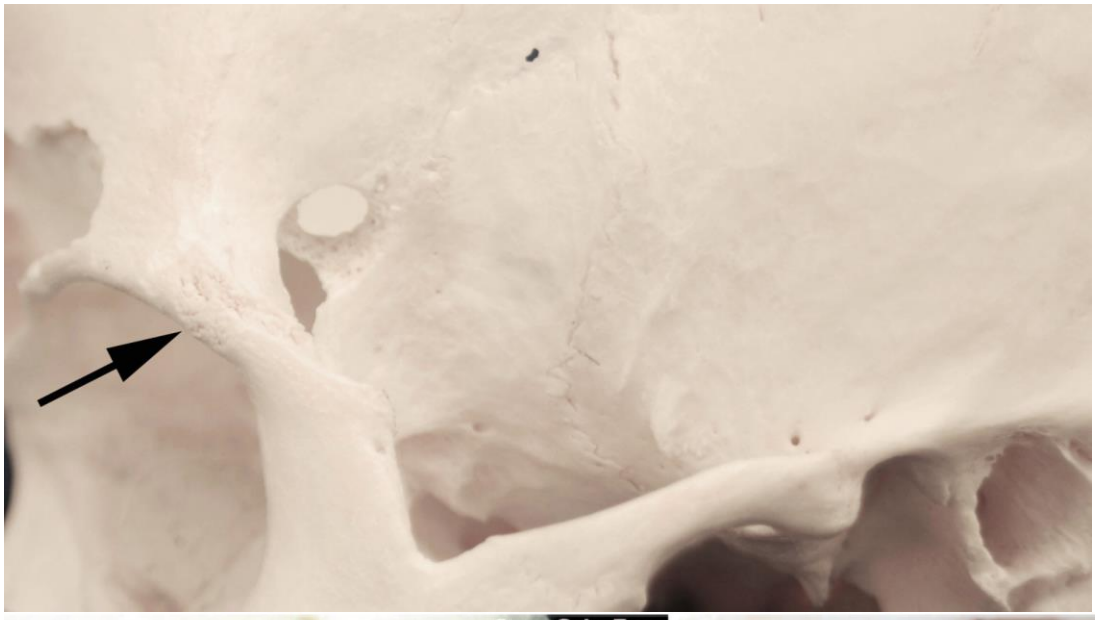
Last cut - across the roof of the orbit through an expanded keyhole (“key” location of keyhole for exposing the orbit and frontal dura is important) using protected (two patties) osteotome:

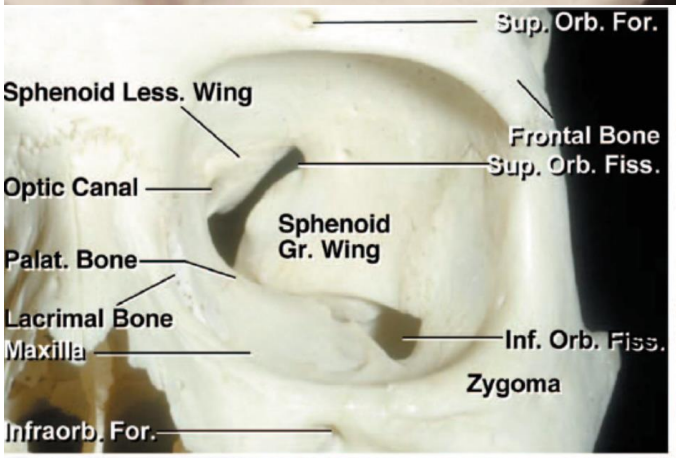


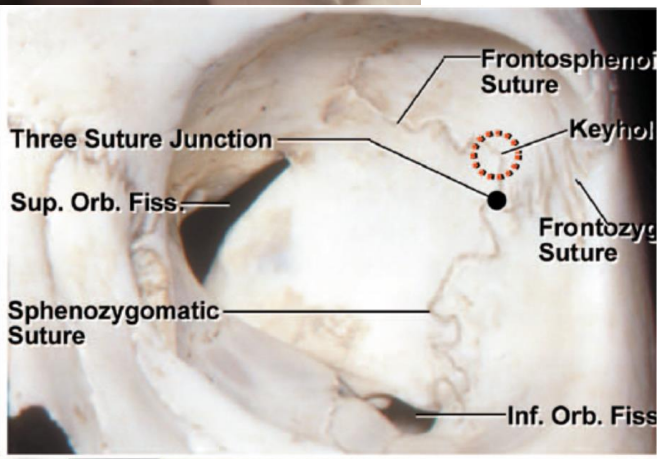
MCCARTY KEYHOLE

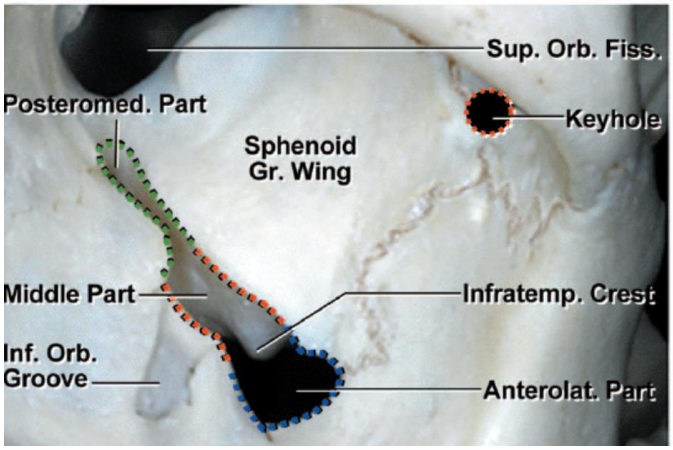
- access to orbita and frontal dura! (vs. pterional keyhole – made higher – only access to frontal dura)

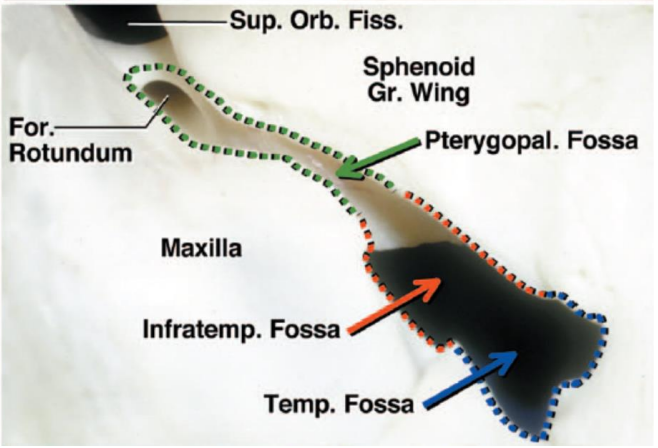
- keyhole (touch above where three sutures meet) - 7 mm superior and 5 mm posterior to frontozygomatic suture (black arrow):

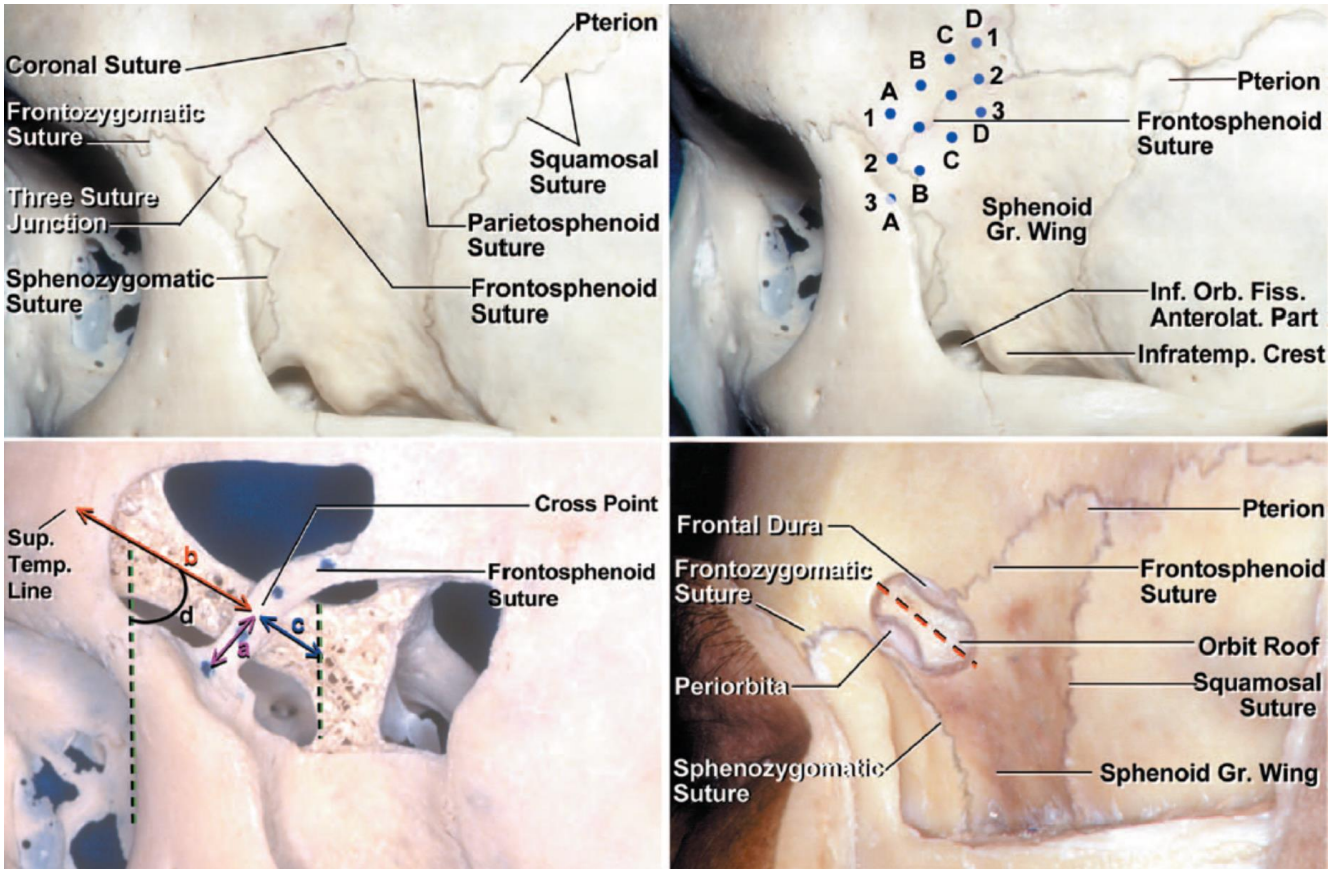



Sup. Orb. For.
Sphenoid Less. Wing
Frontal Bone
Sup. Orb. Fiss.
Optic Canal
Sphenoid
Gr. Wing
Palat. Bone
Lacrimal Bone
Maxilla
Inf. Orb. Fiss.
Zygoma
Infraorb. For.


Suture
Three Suture Junction
Sup. Orb. Fiss.
Suture
Sphenozygomatic
Suture
Inf. Orb. Fiss

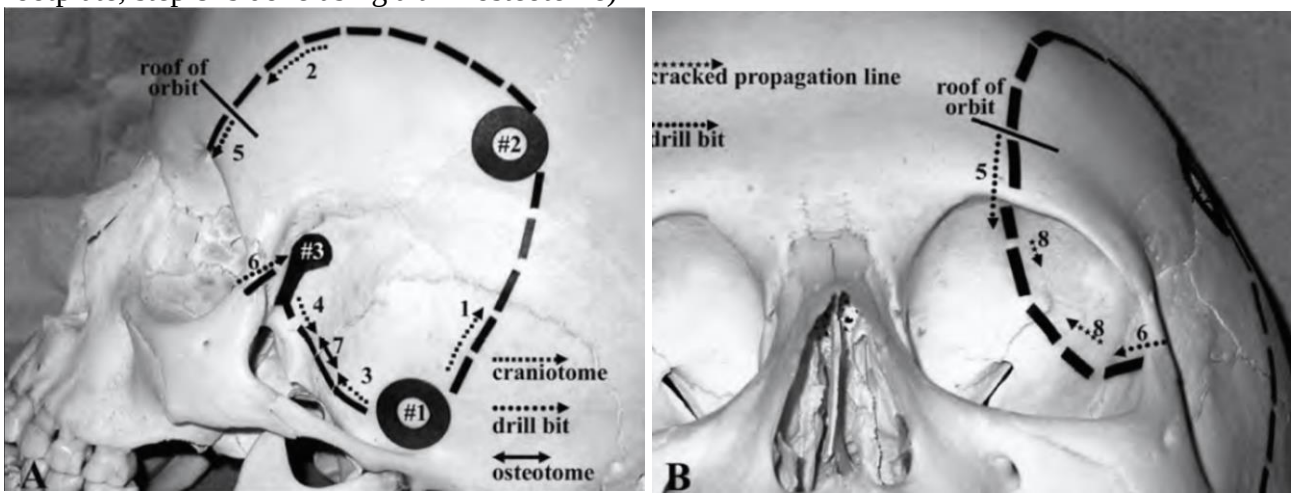

Sup. Orb. Fiss.
Posteromed. Part
Keyhole
Sphenoid
Gr. Wing
Middle Part
Infratemp. Crest
Inf. Orb.
Groove
Anterolat. Part


Sup. Orb. Fiss.
Sphenoid
Gr. Wing
For.
Rotundum
Pterygopal. Fossa
Maxilla
Infratemp. Fossa
Temp. Fossa



STEPS

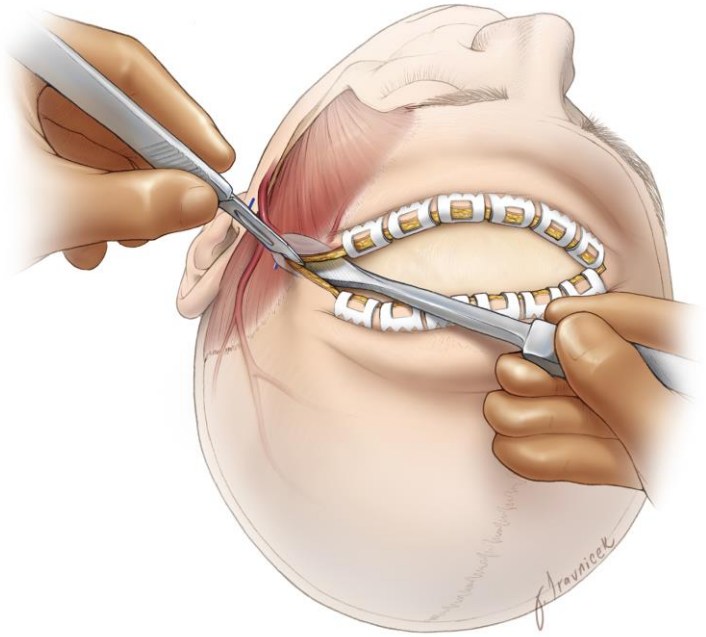
steps in execution of a one-piece modified OZ (steps 3-7 are performed using a B1 drill bit without a footplate; step 8 is done using a thin osteotome)



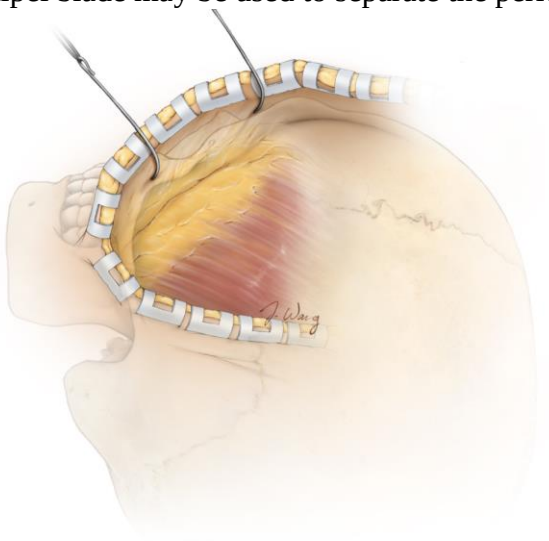
SURGERY

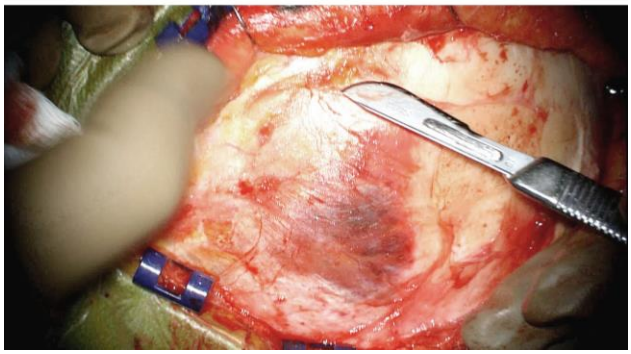
- supine with the head turned 20-40 degrees (closer lesion to midline, less head is turned)
- neck is slightly extended and the head is tilted toward the floor - malar eminence is the highest point (allows gravity retraction of frontal lobes away from the orbital roof).

- incision begins 1 cm anterior to the tragus at the level of the zygomatic arch → stays behind the hairline, curving forward across the midline to the point where the contralateral midpupillary line meets the hairline (flat dissector separates the galea from the pericranium):



- *plane between the galea and the pericranium* is developed in case a vascularized pericranial flap is needed at the time of closure.
- scalp flap is reflected anteriorly and is separated from the temporalis fascia; belly of the #10 scalpel blade may be used to separate the pericranium from the galea – until fat pad is exposed:



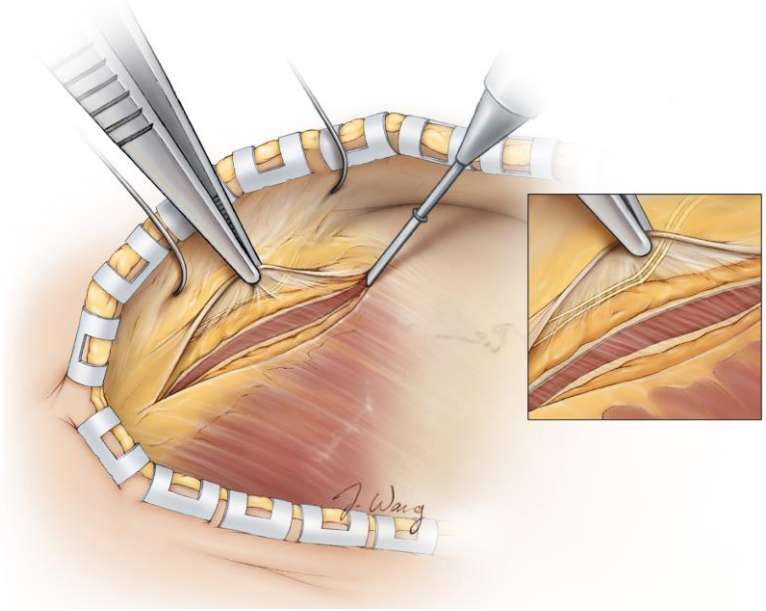


- CN7 frontalis branch(es) are located in the superficial fascia of the fat pad, not within the fat pad - two techniques for reflecting the fat pad

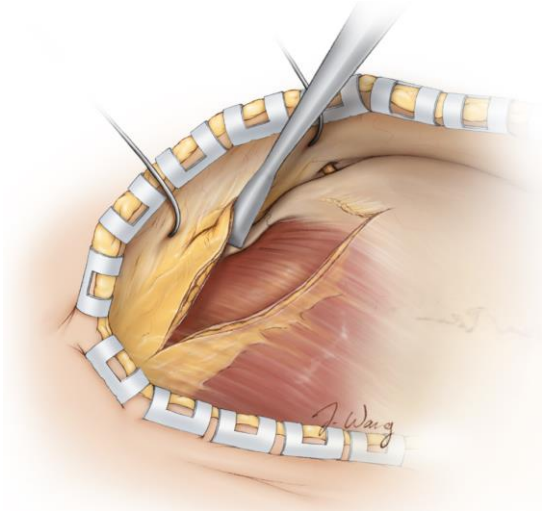
  A. Interfascial technique: superficial temporal fascia is reflected anteriorly along with the fat pad via dissection underneath the fat pad but superficial to the deep temporal fascia.

B. Subfascial technique (safer): superficial temporal fascia + fat pad + deep temporal fascia reflected anteriorly all as one layer.

- Bovie is used to cut deep temporal fascia and reflect the fat pad in the subfascial manner:



- subperiosteal zygoma exposure: deep temporalis fascia is fused along its anterior edge with the periosteum of the frontal zygomatic process; subfascial dissection is continued anteriorly with subperiosteal dissection over the frontal zygomatic bone to achieve full exposure of the superior orbital rim and frontal zygomatic process; dissection is continued until supraorbital nerve and notch are identified:



Further may see p. Op300 >>

ANTERIOR CLINOIDECTOMY

See p. Op 300 >>

TRANSPETROSAL APPROACHES TO POSTERIOR FOSSA

- resection of petrous temporal bone to various degrees provides different levels of access to lesions of posterior fossa (cerebellopontine angle, petroclival region).

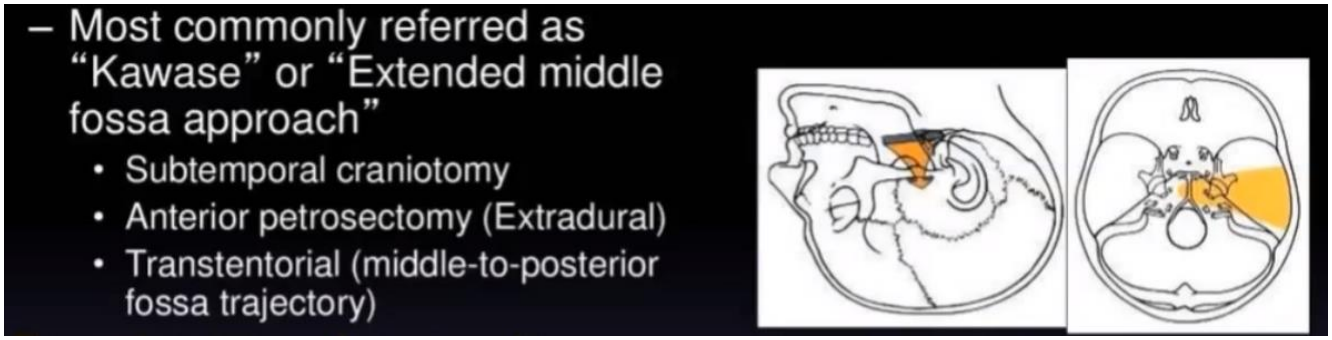
Variants of transpetrosal approaches can be classified:

A. Anterior transpetrosal approaches
B. Posterior transpetrosal approaches

ANTERIOR (MEDIAL) TRANSPETROSAL APPROACHES (KAWASE, S. ANTERIOR PETROSECTOMY)

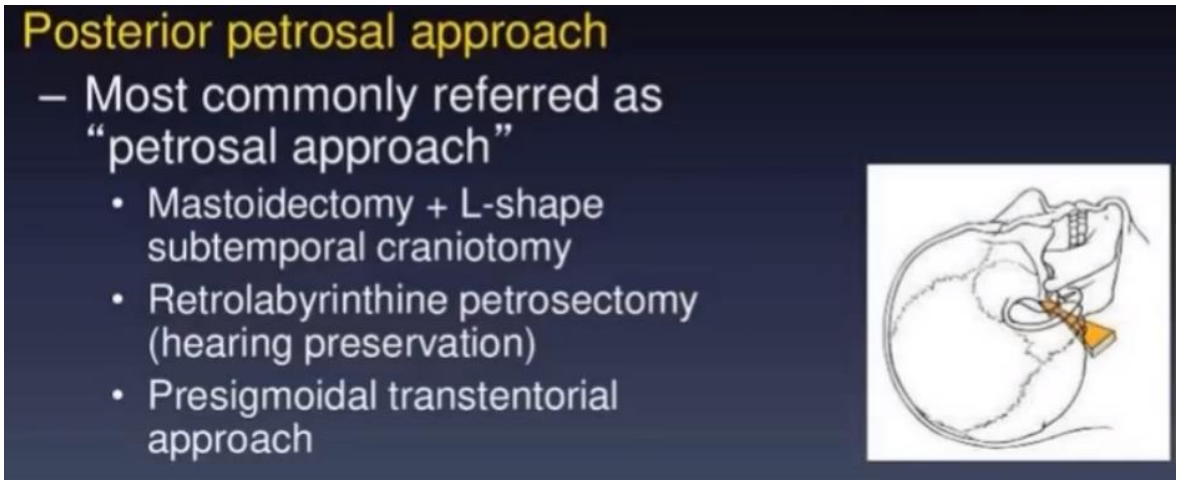
- extensions of basic middle fossa approach.

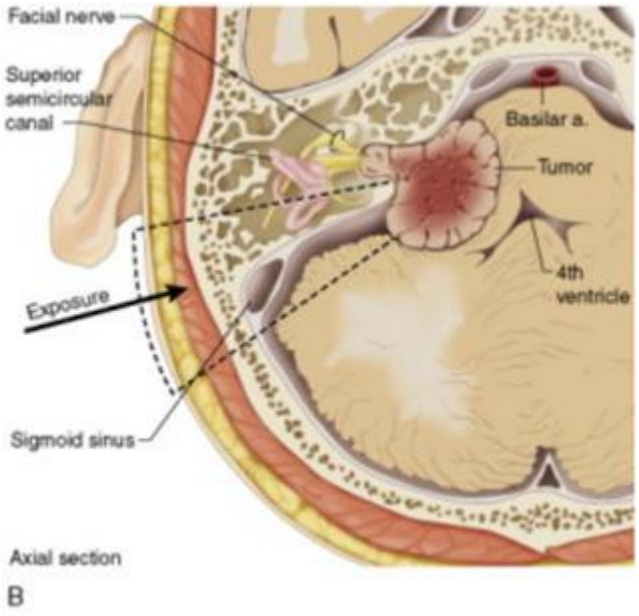
- designed to preserve hearing - spare lateral petrous bone.
- involve resection of medial petrous bone to various degrees.
- involve resection of bone within Kawase rhomboid and division of tentorium to provide exposure of posterior fossa.
- GSPN (easy to identify) is right above and parallel to petrous ICA

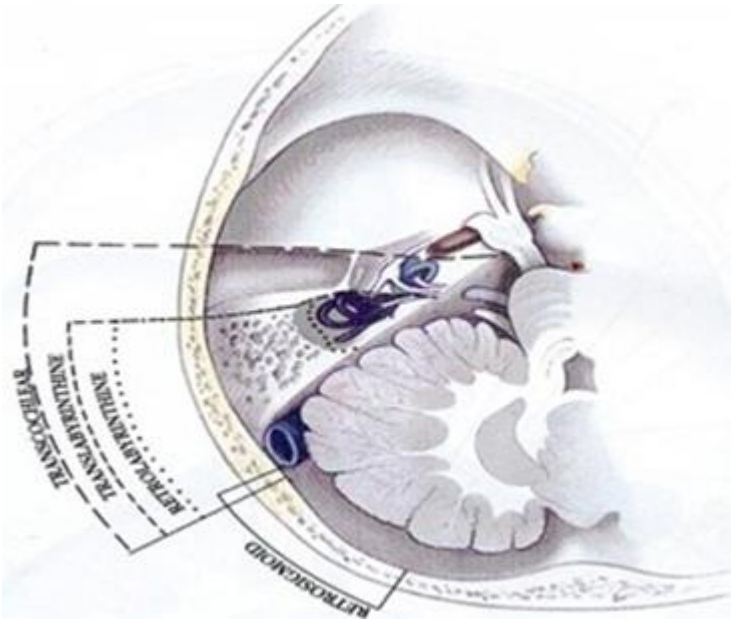


POSTERIOR TRANSPETROSAL (PRESIGMOID) APPROACHES

- standard **mastoidectomy** → resection of petrous bone to progressively increased exposure anteriorly: retrolabyrinthine → translabyrinthine → transcochlear
- comes at expense of hearing in **translabyrinthine** approach and of hearing and facial strength in **transcochlear** approach.

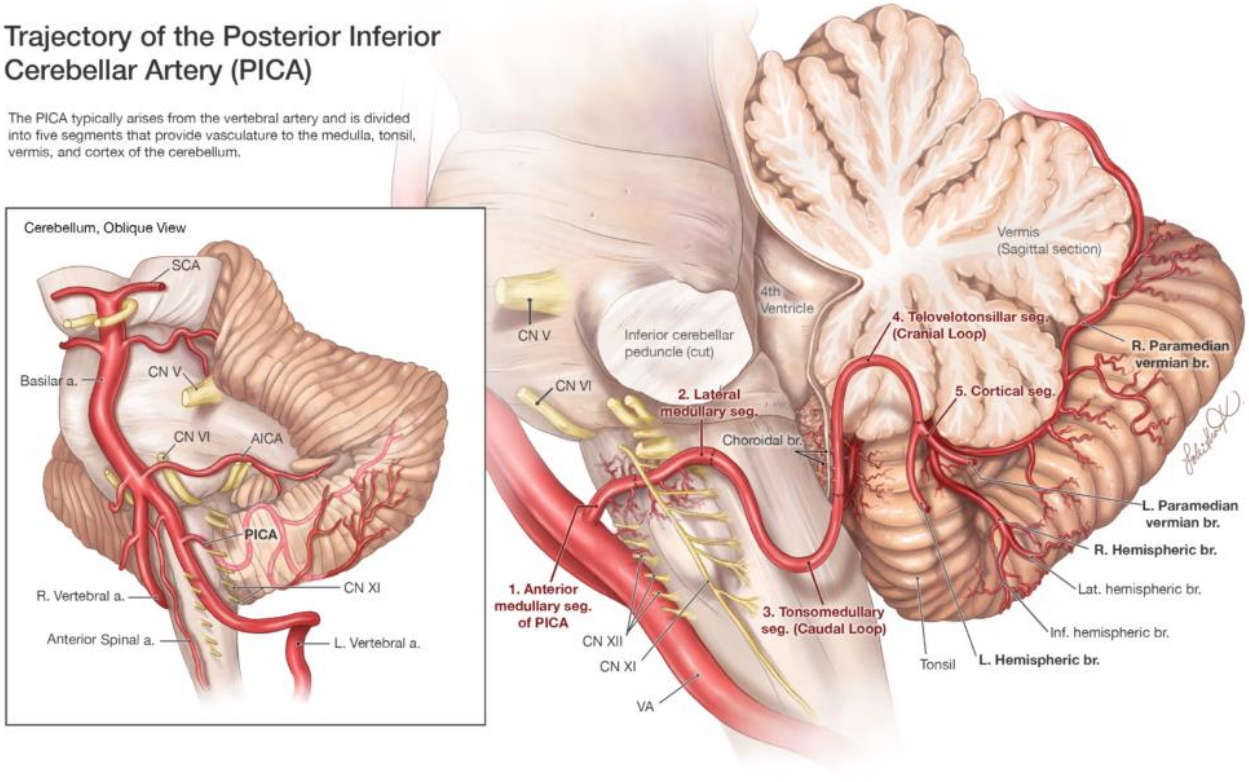






MIDLINE SUBOCCIPITAL CRANIOTOMY

- DEXAMETHASONE, MANNITOL (not for Chiari)
- consider ***neuromonitoring*** (SSEP, MEP, ± BAER) in severe cases.
- consider placing **lumbar drain / EVD** (e.g. occipital)
- N.B. translate head posteriorly and flex as much as possible to facilitate bone work!
- suboccipital decompression is approximately 3 × 3 cm
- N.B. in some posterior fossa cases, Dandy/Frazier burr hole* >> is included in field in case of intraoperative catastrophe or when preoperative hydrocephalus exists!
  *only bur hole (not actual EVD) – if needed (acute HCP), can be punctured percutaneously at bedside with spinal needle
- Dr. Tye uses intraoperative ultrasound to see if *cerebellar tonsils still pistoning* (despite craniectomy) – i.e. visibly moving up and down in sagittal plane (normally, tonsils remain still or gently pulsate with heart activity) → open dura.
- patent circular sinus (esp. in kids) around foramen magnum can be encountered → **Weck clips**.
- see PICA wrapping around tonsils.
- explore obex – to make sure no flow obstruction there (esp. if patient has syrinx).



EMERGENCY TREATMENT FOR P-FOSSA SWELLING

1. Rapid intubation
2. Ventricular tap (through previously placed Frazier burr hole)
3. Level 1 reoperation - wound should be opened immediately wherever patient is (recovery room, ICU, floor…) - CT scanning may cost valuable minutes!

RETROSIGMOID (RETROMASTOID) CRANIOTOMY

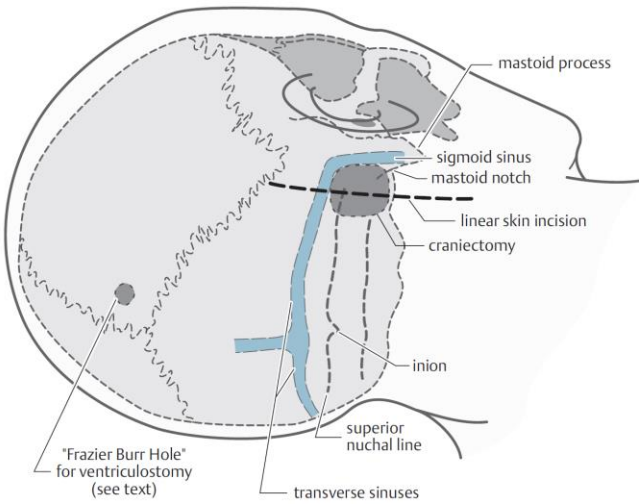
See p. Op300 >

Summary:
lateral decubitus ("park bench")

1) bilateral auditory potentials (BAER)
2) EMG of facial muscles.

Be prepared for venous sinus injury and air embolism!

Incision #1 vertical or slightly curvilinear (***lazy-S***) vertically behind auricle, 1-2 fingerbreadths behind hairline central incision third behind ear (1/3 of incision is above **transverse-sigmoid junction** and 2/3 are below)



- place bur hole just below transverse-sigmoid junction (i.e. not over asterion!)
- craniectomy is a safer option (e.g. in older patients with adherent dural).
- for MVD - small (3x3 cm) oval retrosigmoid craniectomy with Acorn drill bit and Kerrison rongeur, targeting corner of transverse-sigmoid venous sinus junction (neuronavigation!).
- for vestibular schwannoma – larger craniotomy, extend along sigmoid sinus and posteriorly to allow cerebellar retraction.

- sinus injury – suture piece of **muscle** over the defect!
- any exposed mastoid air cells are carefully waxed off (to prevent postoperative CSF leak).
- gentle retraction of cerebellum inferiorly and medially; arachnoid membrane fenestration with CSF cistern decompression to expose cerebellopontine angle.
- **superior petrosal (Dandy) vein** is coagulated and cut; however, it is possible to preserve it.
  N.B. coagulating and dividing the SPV is controversial (risks cerebellar infarction, midbrain and pontine infarction); if SPV is torn, the dural side is tamponaded (sometimes up to 30 minutes is needed) while the free end is coagulated.

Cerebellar retraction

A. Access to CN5 - retract cerebellum inferiorly
B. Access to porus acusticus (e.g. for vestibular schwannomas) - retract cerebellum medially
C. Access to the lower cranial nerves (e.g. for geniculate neuralgia) - retract cerebellum superiorly

Closure

- bone wax should be again applied
- craniectomy - cranioplasty with titanium mesh or with HydroSet.
- Valsalva is performed at every step of closure – make sure no CSF leak!

Postop

- **hypertension** should be avoided at all costs to prevent bleeding from tenuous vessels (sudden changes in BP may indicate elevated pressure in posterior fossa!)
- often keep intubated for 24-48 hrs (many complications may cause respiratory arrest)
- if early severe headache, STAT CT – if no bleed, no significant pneumocephalus → perform LP to relief CSF pressure;
- if severe headache 3-7 days postop → LP to diagnose aseptic meningitis (H: steroids).

SUPRACEREBELLAR INFRATENTORIAL APPROACH

Full text >>

Indications:

1) pineal region
2) posterior third ventricle
3) posterior mesencephalon

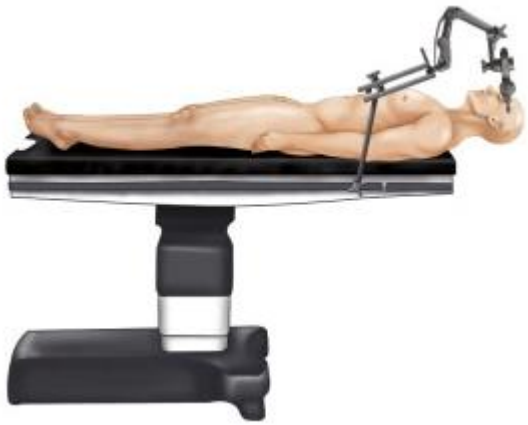
Contraindication - steeply angled tentorium (H: occipital transtentorial approach)

- preoperative **bubble cardiac Doppler study** (right-left shunt ← contraindication for the sitting position – risk of brain air embolism; H: prone)
- **MRV** - relationship of deep venous structures (vein of Galen, basal vein of Rosenthal, internal cerebral veins, and straight sinus) in relation to trajectory and tumor.

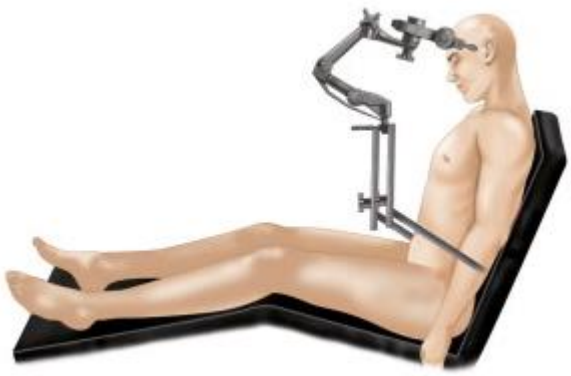
Sitting position – preferred – permits cerebellum to fall with gravity away from tentorium, prevents pooling of venous blood in operative field

N.B. it is the only position in neurosurgery when special preparation is needed for a high risk of air embolism!!!!

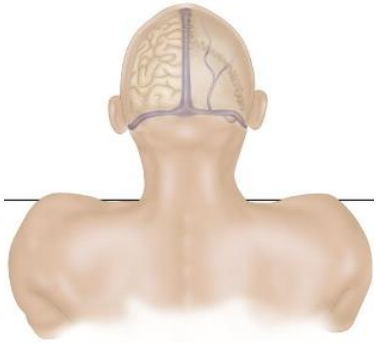
- first place supine on operative table (with reverse orientation):



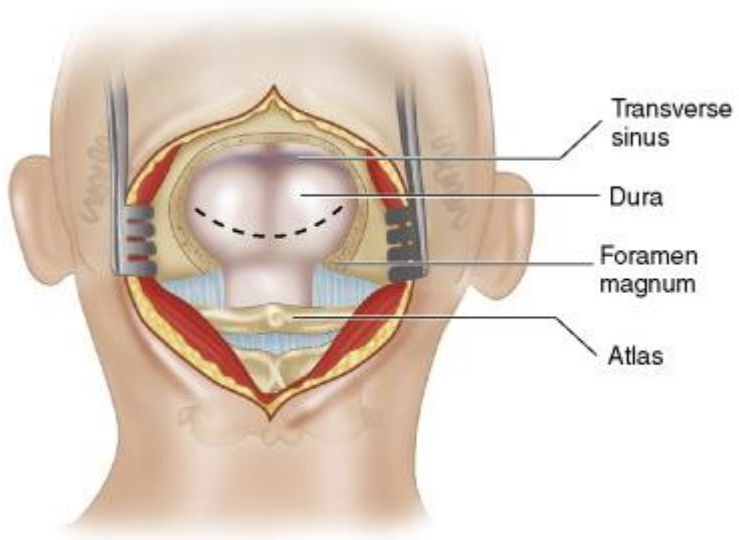
- after application of Mayfield holder, bed is maneuvered to raise patient's back and flex legs (elevate legs to promote venous return):



- head is flexed to place ***tentorium parallel to floor***:



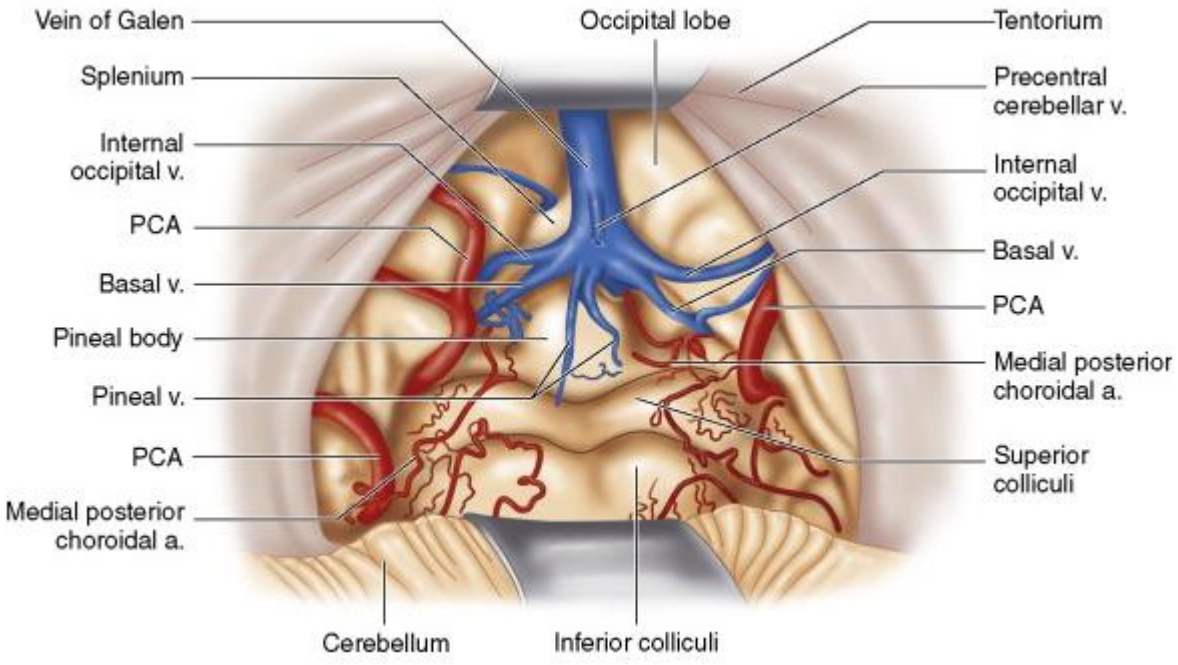
- skin <u>incision</u> from above inion down to approximately C2-4.
- <u>suboccipital craniotomy</u> (musculature is not detached from spinous processes of C1-2)
- burr holes are placed on each side of superior sagittal sinus (right above torcular Herophili) + superior and inferior to each transverse sinus, few centimeters distal to torcular Herophili.
- <u>dural incision</u> – semilunar



- *if angle of tentorium is too steep*, craniotomy can be extended for occipital transtentorial approach, or tentorium can be cut and retracted via supracerebellar approach.
- **arachnoid adhesions and bridging veins** between cerebellum and tentorium are divided to open supracerebellar infratentorial corridor

bridging veins should be divided close to cerebellum to prevent retraction of inaccessible bleeding sources back into tentorium

precentral cerebellar vein is visualized (draining into vein of Galen) – it is the only deep venous structure that should be cauterized and divided



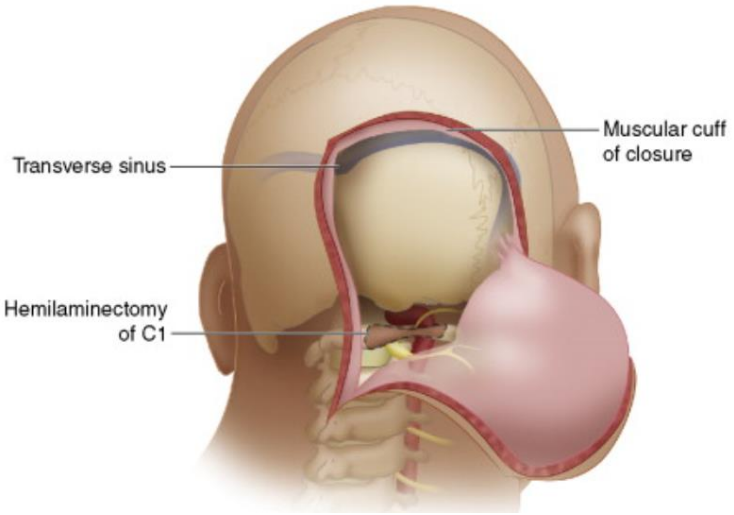
FAR-LATERAL SUBOCCIPITAL APPROACH

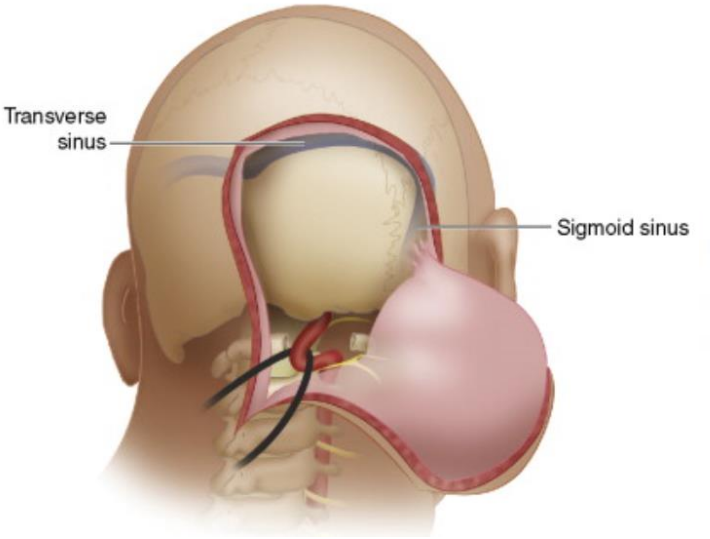
access to the 90 degrees anterior to the medulla

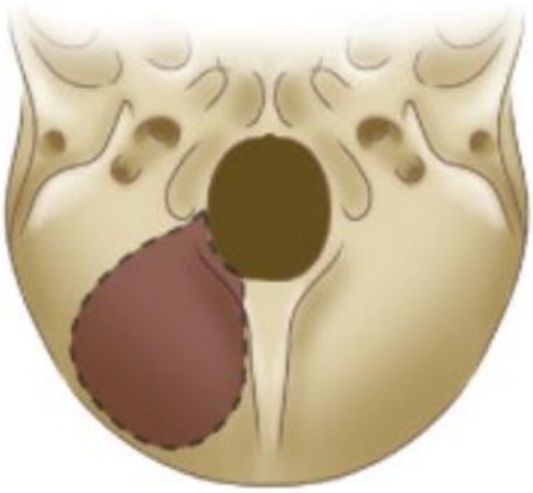
Roughly 50%* (8 mm) of condyle can be safely removed posteriorly before *occipitocervical fusion* should be considered. *some experts say 30%

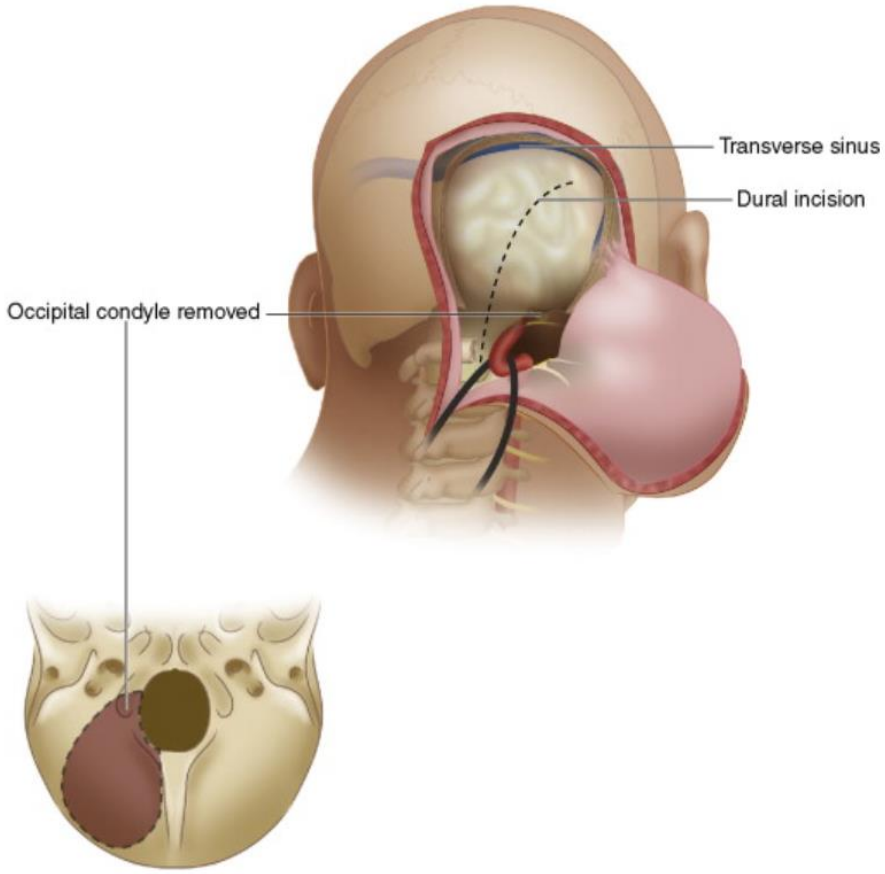
- following this rule, hypoglossal canal is rarely seen.
- usual stop landmark is condylar emissary vein.

- it is like retrosigmoid approach just more inferior - **three-quarter prone (s. park bench, lateral oblique) position** with the contralateral shoulder down.
- VA is mobilized medially.




Transverse sinus
Sigmoid sinus




Transverse sinus
Dural incision
Occipital condyle removed