NEUROLOGY CLERKSHIP REVIEW

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Review Approach/Principles

Not "Pattern Recognition" but a top-down approach, which helps in clinical neurology.

Will help all clinicians who interact with neurologic patients, conditions, or consults.

Clinical decision-making involves looking at the:

1. Localization and time course, to arrive at likely categories under the VITAMNDEC organizing principles, so we can choose....
2. Diagnostic studies that can help rule in or out categories, then going into specific diseases.
3. When we discuss treatments, we'll go into disease-modifying (acute management and secondary prevention) and symptomatic treatments.

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So let's define some terms.

Localization: First, look at symptoms and signs – so do a good history and neuro exam.
Where is the lesion most likely to be, and where else could it be?

Neurologic lesions can be localized to three categories:
- Focal – a single, well-defined anatomic lesion
- Multifocal – multiple well-defined anatomic lesions
- Diffuse – broader, affecting a functional or anatomic part of the NS that is selectively vulnerable

Time Course:
Looking at time course of onset
- Acute instantaneous, or over minutes to maybe an hour or two
- Subacute development over many hours-days (short subacute) to weeks/months (long subacute)
- Chronic development over many months to years.

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VITAMN DEC is what we use to consider a differential in neuro problem solving. Vascular, Infectious, Trauma, Autoimmune/Immunologic, Metabolic/Electrolytes, Neoplastic, Degenerative, Episodic/Paroxysmal, and Congenital

Vascular – TIA or stroke. Aneurysm or aberrant vessel causing mass effect don't count. Acute and focal.

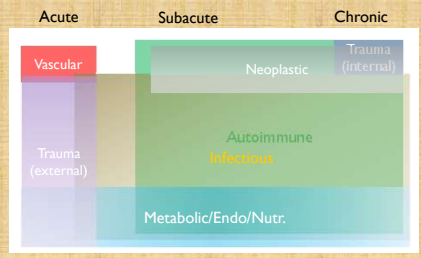
Infectious – Viral, bacterial, fungal etc. May be fever, WBC count. Often rapid subacute, but can be any time course/localization

Traumatic – Two types: External – usu w/ history of head/neck/back/extremity trauma and/or serious soft-tissue injury.
Internal – mass effect from structures, or nerve rubbing against bone.

Autoimmune/immun. – (e.g. MS, GB, SLE)
Any localization, onset subacute or chronic
Most follow a "subacute exacerbation, static defects, & subacute partial/full resolution.
Some, like GBS are monophasic.
MS is odd, with serial lesions, disseminated in time and space.

Metabolic/toxic/endo/nutritional
Usu. Diffuse & subacute, but may be any time course

Neoplastic: Focal lesions. Subacute (if very malignant tumor or metastasis) or chronic



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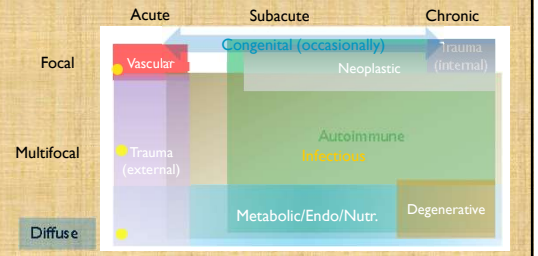
Degenerative: Alzheimer's, Parkinson's, ALS, muscular dystrophies. Typically diffuse and chronic.

- Episodic/Paroxysmal: A series of acute episodes – things like headaches, seizures, vertigo, narcolepsy.
- Timing: Periods of spontaneous change in neurologic function, interspersed with periods of normal function. These may recur over a series of years. May be primary or secondary to another neurological category (e.g. seizures d/t neoplasm)

Congenital/Developmental – (e.g. Chromosomal abnormalities, CNS malformations) Often diffuse and stable, but can occasionally present as focal lesions with any time course.



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Confirmatory Tests

Know your tests and their functions – important to help support or eliminate possible differentials.

Physiologic studies: EEG, EMG, nerve conduction
confirm normal or abnormal function

Imaging Studies: CT, MRI, SPECT, PET, fMRI

confirm presence or absence of lesions in specific locations

Lab studies: LP, blood labs, urinalysis, special studies, etc.

confirm or deny hypotheses about types of disease categories

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Syndromes

Recognizing Patterns with distinct anatomic associations

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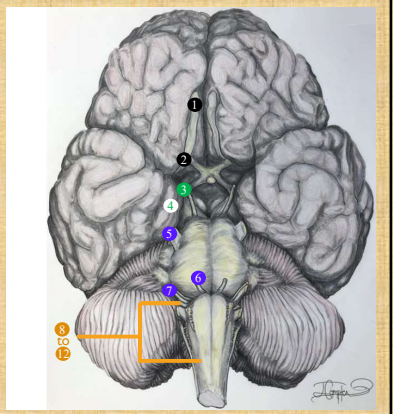
Cranial Nerves

1 & 2	above Midbrain
3 & 4	Midbrain
5, 6, 7	Pons
8, 9, 10, 11, 12	Medulla

Will mix up order so it's not totally linear and predictable, but will save 3,5,7,8 for last since the discussion is a bit more involved for those



Art by Jason Compton jasoncomptonart.com

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Patient has difficulty swallowing, she's been having bitter coffee and lots of lemon in tea lately. On neurologic exam, she does not have a gag reflex.

Glossopharyngeal CN9 lesion. Loss of stylopharyngeus (swallowing food bolus), and special sensation of taste, with innervation to posterior third of tongue (bitter and sour). Will lose gag reflex, especially if CN 10 also injured.

Pt with hoarse/nasal voice, and difficulty swallowing. Hasn't been feeling as hungry. On physical exam, you notice the uvula deviates to the L side.

R Vagus nerve CN10 injury. Uvula points to the normal side.

Can also see delayed gastric emptying, and can lose gag reflex, especially if CN9 also injured.

CN IX carries the ipsilateral gag reflex afferent limb,

CN X carries out the bilateral efferent limb.

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Patient says things don't taste as good, and upon further assessment, has lost the ability to smell.

CN 1: Olfactory Nerve

Pt concerned about double vision when looking to the left. On visual field testing, you notice the R eye follows your finger to the patient's left, but the L eye stays in the center.

Left CN 6: Abducens – affected eye cannot abduct

Can also be seen in person with increased ICP. e.g. IIH/pseudotumor

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L eye with blurry vision and "washed out" colors. On exam, when light is shone into R eye, L eye constricts, but when light moved to L eye, it appears to dilate.

L CN 2 (Optic Nerve) lesion.
Causes optic neuritis and afferent pupillary defect.

Patient presents with some pain in her neck, and double vision. On physical exam, her head is slightly tilted to the left, and when head is straightened, her right eye is slightly deviated up and to the right.

R CN4 (Trochlear) lesion. Lose innervation (and tone) for R superior oblique. (Head tilt is an attempt to compensate for this)

CN4 has a very long course, so multiple possible areas of injury.

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Weakness rotating head to the R and turning chin to R. Has a left shoulder droop.

L Accessory nerve CN 11 injury. Trouble turning head or chin to opposite side.

Patient with trouble speaking, and tongue deviates to the left when they stick it out.

L CN 12 Hypoglossal nerve injury. L LMN (nucleus or nerve fibers).

Lower motor neuron "Lick your wounds"

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Decreased facial sensation, unable to chew, and no corneal reflex.

Trigeminal nerve CN5 lesion. Chewing difficulty due to muscle atrophy, also controls afferent limb of corneal reflex. Look for other signs in the pons area.

Classic peripheral nerve disorder?

Trigeminal neuralgia (tic doloreux) – excruciating pain even while brushing teeth or applying makeup to face.

Caused by *compression* of trigeminal nerve root.

Treatment

NaCh-blocking AED's and neuropathic pain meds.
Classically Carbamazepine

– but beware of N/V, leukopenia and Stevens-Johnson syndrome

(SJS association: patients with HLA B1502 haplotype, MC in SE Asians)

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One week of facial weakness on L side, straw falls out while drinking, and mouth droops.

CN 7 Facial nerve – controls muscles of facial expression.

What do you want to know next?

Is forehead able to move? If yes, UMN lesion. Perhaps 2/2 to stroke or tumor.

If no, LMN (Bell's) palsy. Can be temporary and resolve. Consider time course and onset.

If forehead is involved (LMN), what else might you see?

Loss of:
- Taste in anterior 2/3 of tongue
- Sensation on skin of ear
- Stapedius muscle (watch out for hyperacusis)
- Lacrimal and salivary gland function Give artificial tears!
- Ability to close eye

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Patient with recent hearing loss in L ear. When tuning fork placed on forehead, it's louder on normal (right) side.

Sensorineural hearing loss due to CN8 (Vestibulocochlear) lesion. Because it's unilateral, lesion is in cochlear nuclei or cochlear part of CN8, not the nucleus

Classic neoplastic cause of LMN syndrome in CN VII, VIII

Cerebellopontine angle tumor (acoustic neuroma/vestibular schwannoma) – rare in USA that it gets big enough to cause significant facial weakness, but can see loss of balance early.

If bilateral?

Consider NF2

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Non-CN Visual Syndromes

Pinpoint Pupils

Lose descending sympathetic tract, in medulla

Sluggish, dilated pupil nonresponsive to light, acute headache, nausea, blurry vision and halos around lights.

Angle-closure glaucoma

Can also see injected eye, tearing, vomiting.
Can be triggered by anticholinergics used for Parkinson's (like trihexyphenidyl) or tolterodine.
Also triggered by dim light, which dilates the pupil.
If not treated with IOP-lowering drugs, patients can develop permanent vision loss within 2-5 hours.

Difference from open angle glaucoma?

Open-angle develops more slowly, and the angle between iris and cornea isn't rapidly closing, but its trabecular meshwork is slowly getting clogged.

Dilated pupils react sluggishly to light, and reduced neck and arm muscle strength

Foodborne botulism. Treat with equine antitoxin

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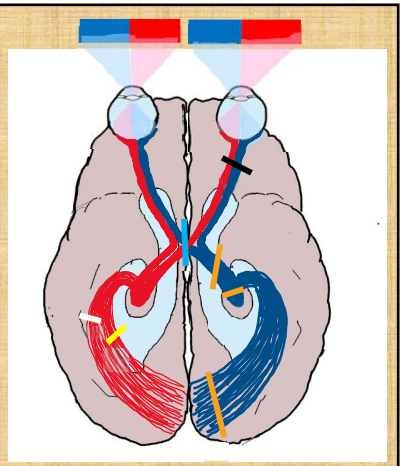
Visual Field Syndromes

Optic nerves-> chiasm -> Optic tracts -> LGN
-> Optic radiations -> Occipital lobe



Right monocular vision loss
R optic nerve

Bitemporal hemianopsia
Optic chiasm (think pituitary tumor)

Left Homonymous hemianopsia
MC: Post. Cerebral Artery infarct in R occipital lobe
Possible: optic tract or lateral geniculate of thalamus

Right upper quadrantanopia
Left optic radiations in left temporal lobe

Right lower quadrantanopia
Left optic radiations in left parietal lobe

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Acute onset monocular blindness

Painless, for several hours

Central Retinal Artery Occlusion. Will see cherry red spot, and a whitened retina (ischemia)

Painless, often gone within minutes, less than 30 mins.

Amaurosis Fugax (a TIA of the eye)

Work up with?

Carotid imaging (duplex ultrasound of neck), and also some cardiac imaging

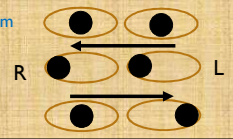
Pupil constricts with accommodation, but not with light:

Argyll-Robertson pupil of neurosyphilis: affects efferent pupillary fibers of the Edinger-Westphal nucleus (which responds to light), but preserves the ones that respond to near vision

Patient can abduct their eyes to the side but can't adduct one of them

Internuclear ophthalmoplegia. MLF lesion

The eye that can't adduct is the side of the MLF lesion (here, it's right MLF)



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Auditory

Progressive bilateral symmetric hearing loss with subjective tinnitus, and absence of other focal neurologic deficits

Presbycusis. Seen more in old age. Will be high frequency hearing loss.

Rinne and Weber tests

To find etiology: sensorineural vs. conductive hearing loss in a "bad ear"

Weber on top of head, louder in affected ear if conductive.

Rinne – fork over mastoid bone until stops, then put near ear to see if they can hear bone > air if conductive. *Key: travels better through bone if it's conductive hearing loss.

Pay attention to history/time course for causes of

Sensorineural	CN8 lesion, ototoxicity, loud noise
Conductive	cerumen impaction, foreign object, allergies/fluid, cholesteatoma, ruptured TM

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Patient says "I'm dizzy" -what do they mean?

Vertigo (spinning/unsteady)
Presyncope/syncope

Vertigo with associated hearing loss or tinnitus.

Peripheral vertigo – a problem with the inner ear vestibular apparatus or CN8, which is considered peripheral. Can have hearing issues due to inner ear involvement, or loss of CN8

If recovered from a viral URI in the past few weeks?

Labyrinthitis (will have hearing loss), or
Vestibular neuritis (no hearing loss)

Steroids can help reduce duration.
Meclizine can make them feel better.

Vertigo, tinnitus and hearing loss, and in episodes 40 mins-1 day.

Meniere's Disease. Diagnose clinically, treat with salt restriction and thiazide diuretics, and/or meclizine

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You don't need hearing issues for peripheral vertigo.
What if it's position-dependent?

Benign Paroxysmal Positional Vertigo (BPPV)

Diagnose with Dix-Hallpike maneuver
Treat with Epley maneuver

If vertigo plus other focal neurologic deficits localizable to the brain stem or cerebellum?

Central vertigo. In brainstem's vestibular apparatus or its connections, most notably to the cerebellum.

The lesion will be a posterior fossa insult (MS, stroke, tumor, abscess, complex migraine) – look at time frame

Can see cerebellar signs because cerebellum is in posterior fossa, which is rigidly bound

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Intracranial Syndromes: Cerebellar

Patient can't stand steady while walking, and cannot perform a heel-to-shin test.
Cerebellar injury.

Cerebellum – coordinates skeletal muscle contraction, balance, and gait
Receives and projects peduncles from/to Pons, Midbrain and Medulla
Often presents with ataxia

Can see signs like: an intention tremor with movement
dysdiadochokinesia (alternating hand movements)
"scanning" dysarthria (pitch & rhythm affected, but not grammar or word choice.

The balance /posture/equilibrium issues are from the vestibulospinal and reticulospinal projections to axial and proximal limb muscles.

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Intracranial Syndromes
Cerebral Lobes

Apathy, inattention, disinhibition, labile affect:
Frontal lobes. Lose executive function.

Person starts ignoring one side of body, trouble getting dressed.
Hemi-neglect due to contralateral hemisphere, typically vignettes say parietal lobe.

If they also have inability to read, write, name things or do math…
Dominant hemisphere (perhaps parietal lobe). Aphasia can make it hard to identify neglect.
Most people are left- language dominant.

Unformed visual hallucinations (lights, shapes etc.)
Occipital lobes

If formed, like little people or animals.
Superior brainstem, but also can be seen with PD or dopaminergic drugs

Memory impairment, hyperaggression, hypersexuality:
Temporal lobes

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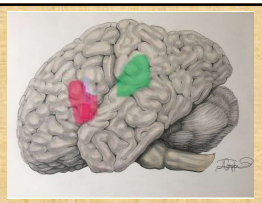
Intracranial Syndromes



Can comprehend speech but has trouble naming things.
Broca's lesion (frontal lobe)

Person can produce speech fluidly, but it doesn't make much sense, and they have trouble comprehending.
"Sensory aphasia"
Wernicke's lesion (posterior superior temporal lobe

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Intracranial Syndromes

Person is in a coma.

Reticular activating system in rostral Pons and Midbrain.

If pinpoint pupils?
Pontine injury

Fixed, medium-sized pupil?
Midbrain injury

Hemiparesis and hemisensory loss. Eyes deviate toward hemiparesis.
Thalamus (eyes *T*oward hemiparesis, which is contralateral to lesion)

R hemiparesis, eyes deviate to left, high incidence of seizures.
L frontal lobe injury. Cerebral cortex has eyes *C*ontra from hemiparesis (but toward lesion).

Can also see with contralateral hemisensory loss (parietal lobe) or ipsilateral homonymous hemianopsia (occipital lobe).

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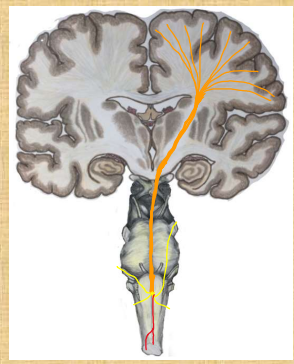
Pyramidal Tract

When do you see a pronator drift?
With subtle upper motor neuron disorders. Can see in pyramidal tract lesions, which may include MS.

Other Pyramidal Tract signs
Focal weakness, spasticity, hyperreflexia, Babinski sign

If pure motor hemiparesis (upper & lower limbs, even face)
Internal capsule. Will lose corticobulbar and corticospinal

All these starting acutely and all simultaneously
consider lacunar infarct (from small vessel hyalinosis)



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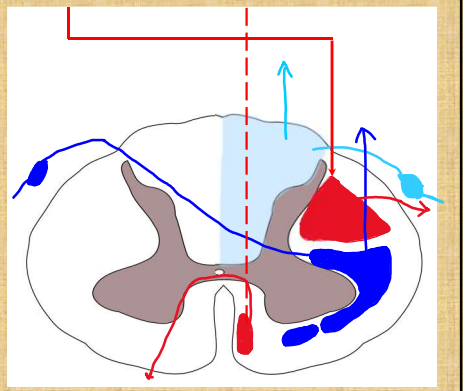
Spinal Pathways

Lateral Corticospinal Crossed in medulla
Carries fibers for ipsilateral motor

Anterior corticospinal
Decussates at spinal level,
but for proximal/postural muscles

Dorsal Column
Fine touch and proprioception for ipsilateral body
Will decussate into Medial Lemniscus in brainstem

Spinothalamic Pain and temperature
Incoming info crosses at spinal level,
Then carries this info to thalamus



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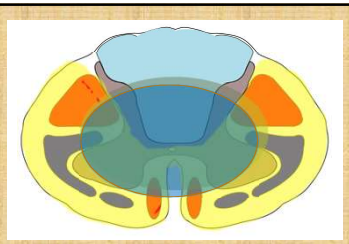
Spinal Syndromes

Weakness and decreased pain/temp in arms, spares legs

Central cord syndrome
After hyperextension (fall, whiplash) often in elderly with underlying spondylosis
(Lose central corticospinal and decussating spinothalamic)



Loss of pain/temp and some motor function in shoulders, back of neck, and arms
Syringomyelia (fluid-filled cavity in central spinal cord)
"Cape-like distribution". Similar to central cord lesion.

Loss of everything except proprioception and/or discriminative touch
Anterior cord syndrome (anterior spinal artery occlusion).
Loss of pain/temperature sensation and motor.
DCML is preserved so proprioception and discriminative touch are intact.

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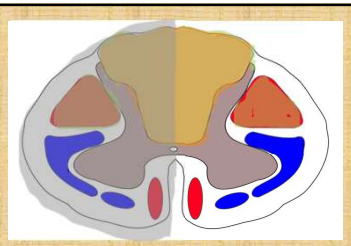
Loss of proprioception, vibration, and discriminative touch.

Loss of dorsal column system – perhaps Tabes Dorsalis



Numbness, paresthesias, impaired proprioception/vibration, and subsequent gait ataxia

Subacute combined degeneration. "Combined" because dorsal columns and lateral corticospinal tracts are lost. Will have upper motor neuron syndrome in legs as well.

Person can't feel tuning fork or move right side, and can't feel pinprick or cold on left.

Brown-Sequard (hemisection) Loss of Ipsilateral vibe, Ipsilateral motor, and Contralateral Pain/Temp
Often due to a stab-type injury

Cancers that metastasize to spine

Breast, Lung, Prostate, Renal, Multiple Myeloma

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Herniation Syndromes

Fixed and dilated pupil, may progress to homonymous hemianopsia

Uncal transtentorial herniation.
The uncus of the temporal lobe is pushed down through the tentorium

Causes Oculomotor CN3 palsy, and affects parasympathetic fibers ipsilaterally (so blown pupil dt unopposed sympathetic tone) and can compress Posterior Cerebellar artery flow to occipital lobe, causing homonymous hemianopia

Also see decreasing LOC due to distortion of ascending arousal systems passing from pons through midbrain

Progression of Uncal herniation?

First, patient gets sleepy, then see blown pupil, then UMN signs (sometimes hemiparesis, sometimes decorticate posturing). It compresses the ipsilateral cerebral peduncle, and then (since the corticospinal tracts decussate below midbrain), you get contralateral hemiparesis

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Herniation Syndromes

Dilated, fixed pupil with paralyzed upgaze (Sunset sign)

Central (transtentorial) herniation
Midline lesion causes diencephalon to herniate through tentorium cerebelli, causing decreased LOC and difficulty concentrating, agitation, drowsiness, and breathing problems. Will see dilated, fixed pupils with upgaze paralysis

As it progresses, will see decorticate or decerebrate posturing

Neck stiffness, headache, cardiac, respiratory difficulties

Tonsillar herniation (downward cerebellar herniation)
Compression of medulla's cardio-respiratory center → respiratory and possibly cardiac arrest
If RAS affected, will also see coma
Very difficult prognosis

Child or young adult with headache, balance problems, or peripheral nerve problem (fine motor issues or neuropathy)

Chiari Malformation – can present for the first time in 20's-40's. Occurs with a fluid-filled cyst (syrinx) and headache will worsen with increased ICP (vagal maneuvers, sneezing)

A syrinx can also be acquired due to trauma, tumor, meningitis and other causes.

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Cerebral Vasculature/Circle of Willis – best to study with strokes:

Feet and legs affected by stroke, greater than face and arms.

Anterior cerebral artery occlusion:
Supplies primary motor and somatosensory, but more medial areas, so legs and feet are affected. Will see contralateral loss of sensation and motor to lower legs>upper extremities.

Contralateral loss of sensation & motor to the upper part of body (e.g. face and upper limbs), AND Broca's area.

Middle Cerebral artery:
Can be the most devastating stroke.

Homonymous hemianopia

Posterior cerebral artery stroke

Locked in syndrome

Basilar artery:
Lose the corticospinal tracts in the pons – bilateral loss of corticospinal tracts. Lose everything except vertical eye movements.
Can also see in central pontine myelinolysis, due to rapid correction of hyponatremia.

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Anterior Inferior Cerebellar:

Lateral pontine syndrome:
CN7 palsy (Bell's), Vestibular artery (vertigo & nystagmus), perhaps even deafness/tinnitus, and middle cerebellar peduncle – ataxia, These will be ipsilateral!
However, will be contralateral loss of pain/temperature (due to decussation in spinal cord)

Posterior Inferior Cerebellar:

Poor coordination/muscle tone/balance (cerebellum), (dysphagia (CN10 palsy), maybe – gag, and the descending sympathetic fibers (Horners syndrome – miosis, anhidrosis, ptosis)

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Classic Lacunar Strokes

Unilateral numbness, that can progress to burning, tingling, other unpleasant sensations on that side

Pure sensory stroke
Contralateral thalamus (Ventral Posterolateral Nucleus)
Can then progress to thalamic pain syndrome (allodynia on light touch)

Hemiparesis or hemiplegia on one side of the body, affecting face, arm or leg. May see dysphagia/dysarthria or transient sensory symptoms

Pure motor stroke, from the contralateral posterior internal capsule

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Other Stroke Syndromes

Abnormal facial sensation and pain, vertigo falling to side of lesion, ipsilateral Horner's syndrome, intractable hiccups, dysarthria, nystagmus, ipsilateral

Wallenburg syndrome. Lateral medulla lesion. Nucleus ambiguus and autonomic systems involved.

L oculomotor nerve palsy, ataxia, and R hemiparesis

L Midbrain stroke

Putaminal hemorrhage

Contralateral sensory loss and contralateral hemiplegia

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Peripheral Syndromes/Reflexes

Sensory loss of 4th and 5th digits, and weakness of grip

Ulnar Nerve Lesion — Perhaps entrapment in the epicondylar groove of the elbow from leaning on elbows over desk.

Wrist Drop, thumb sensory, and can't extend thumbs

Radial nerve palsy — "Saturday night palsy" or "honeymoon palsy" From arm over back of chair

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(Transiently) Impaired foot dorsiflexion (can't walk on heels), impaired great toe extension, and sensory changes over dorsal foot and lateral shin

Common fibular neuropathy Result of leg immobilization, leg crossing, or protracted squatting

MC Cause of foot drop

Common peroneal nerve injury Can be secondary to habitual crossing of legs

Reflex	Nerve	Spinal Root(s)
Biceps	Musculocutaneous	C5 (C6)
Brachioradialis	Radial	C6 (C5)
Triceps	Radial	C7
Patellar	Femoral	L3-L4
Achilles	Tibial	S1-S2

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Numbness, tingling in feet/hands that spreads medially. Sharp, jabbing, throbbing, freezing, burning pain. May also see incoordination.

"Stocking glove pattern"

Peripheral neuropathy.

Has many causes. Think of time course and VITAMNDEC categories:

Infectious: Lyme, diphtheria, HIV, leprosy
Trauma: Carpal tunnel, fractures
AI: GBS, SLE, Polyarteritis Nodosa, Scleroderma, Sarcoidosis, Amyloidosis
Metabolic/Endo: DM, hypothyroid, uremia, deficient B12, B6, Thiamine, Vit E

What if it's a cancer patient?

Chemo drugs -> peripheral neuropathy:

Vincristine, platinum-based (-platins), and taxanes (paclitaxel)

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AMS

Someone comes in to ED in a confused or cognitively impaired state Altered mental status

If confused, with fluctuating level of consciousness

Delirium *"waxing and waning"* pattern

What part of Mental Status Exam is significantly affected?

Impaired attention Serial 7's, days of week backward, months forward, or spell WORLD backwards

Predisposing factors for delirium: various neurological impairments (Dementia, prior stroke, advanced age, Parkinson's, sensory impairments)

Precipitating factors drugs, infection, electrolyte/metabolic issues, and systemic illnesses

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Delirium treatment

Best way to treat delirium

Fix underlying problem

and Provide Careful Supportive Care

- Constant reorientation
- Day: Lights on, and encourage regular activity (perhaps structured like OOB to chair)
- Night: Lights out, reduce nighttime disturbances
- As a last resort, low-dose antipsychotic (e.g. haloperidol)

(But black box warning against use in elderly/demented because of behavior problems)

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Dementia

Patient has been forgetting where they live/forgetting people/not as interactive, it has been subacute to chronic in onset, and doesn't "wax and wane"

Consider dementia

Narrowing it down:

Reduced mood, lower energy, guilt, concentration issues, appetite changes, suicidality.

Pseudodementia, seen in MDD. Look out for SIGECAPS. Treat the depression.

Pt has been growing increasingly forgetful in steps, and has had multiple unwitnessed falls

Vascular dementia – multiple strokes, vascular risk factors, stepwise decline

Hypersexual and personality changes

Pick's (Frontotemporal) – will have frontal and temporal lobe lesion/atrophy
Personality will go first, opposite of Alzheimer's

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Dementia

Person was having movement problems (rigidity, etc) before dementia came

Parkinson's

Person has had dementia and has now developed lower limb rigidity and bilateral hand tremor … or has visual hallucinations

Lewy Body dementia

Both can be described as "reduced substantia nigra volume and Lewy bodies"

But it is more likely to refer to Parkinson's

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Dementia

Rapidly progressive over weeks or months, perhaps in younger patient 30's-40's

Creutzfeldt-Jakob

Sporadic mutation, although some get Mad Cow disease from undercooked meat
Tx is supportive - may die in 6 months

Has been having balance problems/ataxia and experiencing urinary incontinence not explained by other causes

NPH (normal pressure hydrocephalus) – caused by *decreased absorption* of CSF

Enlarged lateral and third ventricles, or "hydrocephalus"

LP will improve these patients (can't be NPH if this is untrue)
Tx with VP shunt

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Dementia - Alzheimer's Disease

Progressive dementia over years, with generalized cortical atrophy and reduced hippocampal volume

Alzheimer's disease

Clinical course of Alzheimer's Disease

Chronic, progressive over years,
Diffuse dysfunction of bilat medial temporal lobes → anterograde amnesia
Language and executive dysfunction → speech apraxia, impaired judgment
Psychosis – paranoid delusions common).

Apo E4 association

Treatment Minor response to Donepezil or memantine
(improve cognition slightly, delay SNF admission, but no dramatic improvement)

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Coma & Brain Death

If the patient is not interactive or responsive at all at all.

Coma – loss of pontine reticular activating system function

ER Coma cocktail

Naloxone to reverse opiate-induced coma
Thiamine & D50 to correct hypoglycemia

What confirms brain death?

Absence of function of the whole brain, including the brainstem.
Absence of brainstem reflexes

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Brain Death

Brainstem Reflexes

Corneal, Cold-water Calorics, Oculocephalic (Doll's Eyes)

Cold Water Calorics

Vestibulocochlear nerve causes eyes to go toward cold, away from warm water. "Eyes to the cold"

Normally, an intact Vestibular Nucleus in Medulla will try to correct this with fast nystagmus movements toward center

"COWS" – Cold Opposite, Warm Same – refers to the fast, *correcting* nystagmus movements

No movement/no nystagmus → lack of brainstem function

Will DTRs be present?

Yes, because they're spinally mediated.

Can heart speed up after atropine administration?

No, because vagal (and all cranial nerve) tone is lost

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Infectious Disease - Meningoencephalitis

Patient comes in with neurologic findings, plus a fever/elevated WBC count

Consider infectious disease

Headache, fever, other focal neurologic deficits like seizures, light sensitivity

Encephalitis – inflammation of the brain

Most commonly viral but can be other infectious or autoimmune
Enteroviruses are a common cause

HSV-1 is a particularly morbid one – will see hyperintense area in temporal lobe

What if there's neck stiffness?

Meningitis – inflammation of the meninges (pia, arachnoid, dura)

MC Bacterial meningitis in adults

Strep pneumoniae

Empiric coverage for bacterial meningitis

Vanc & Ceftriaxone (Except older adults also get ampicillin for enterococcus)
Rifampin PPX for close contacts

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Infectious Disease

Fever, headache, periorbital edema (uni or bilateral) and inability to move eyes

Cavernous sinus thrombosis. Affects CNs 3, 4, V1, V2, and 6

MC from infection in face (corners of mouth to nose bridge or dental procedures)

Can be due to non-septic cause, e.g. trauma, surgery, pregnancy *Valveless venous supply

40M with DM comes in with 2 days of smelly yellow drainage from ear, and recently has developed hearing loss, pain with swallowing, and voice sounds hoarse

Malignant otitis externa – MC pseudomonas (but maybe staph) and diabetes is predisposing factor. Can move to pharynx via eustachian tube.

Brief shooting or burning pain in back, face or extremities; plus sensory ataxia (reduced proprioception)

Tabes dorsalis: Lancinating pains, sensory ataxia.

Can also have Argyll-Robertson pupils. Treat with penicillin.

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Infectious Disease

Stiffness, muscle pains, unable to open mouth completely

Tetanus. Toxin-mediated blockage of inhibitory Glycine and GABA at NMJ
Use equine antitoxin if unvaccinated or unknown vaccination status

Patient had cellulitis, but now they're having leg weakness and lumbar spinal tenderness.

Spinal epidural abscess (65% are S Aureus)

Most common brain abscess bugs — S. Aureus, Strep viridans, or Gram Negatives

Person with shingles 3 months ago now has pain and sensitivity there.

Postherpetic neuralgia

Tx? gabapentin or neuropathic pain meds (pregabalin, duloxetine)

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Infectious Disease

Subacute or acute onset of droopy eyes, bilateral facial weakness, N/V, trouble breathing, and the vignette talks about something they ate

Foodborne Botulism – honey or home-canned food are classic, but can have other exposures

Treatment: Botulinum antitoxin approved 2013, but can monitor respiratory status as inpatient. Spontaneous recovery within 1 week

Animal exposure, nonspecific symptoms, then neuropathy, agitation, pharyngeal spasms while drinking water

Rabies, a rhabdovirus

By the time it gets symptomatic, it's inherently fatal, so the boards will want you to recognize animal exposure – raccoon, possum or bat bite, or any time alone in a room with a bat, even if no clear bite.

Treatment: People at risk can have prophylactic pre-exposure vaccine series
After suspected exposure, clean the wound, give rabies vaccination, and rabies immune globulin.
People who were already vaccinated won't need the IG.

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Infectious Disease

CD4 count for AIDS <200

HIV patient, MRI shows solitary, weakly ring-enhancing lesion in periventricular area

First consider toxo and treat with Bactrim (TMP SMX)

What if no response to Bactrim?

Primary CNS Lymphoma. Will see EBV DNA in CSF

Treatment of CNS lymphoma — Chemo (MTX) and radiation

Subacute onset of altered mentation and focal neuro deficits in HIV or transplant patient

Consider Progressive Multifocal Leukencephalopathy

JC virus affecting immunocompromised

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Disorders of Motor Function

Terminology

Movement disorder: Disorder of basal ganglia motor function
Too many and/or too few motor movements

Weakness: Inability to generate power due to a lesion of UMN, LMN, NMJ or Muscle

-paresis = weakness
-plegia = no movement

Decreased or no reflexes, fasciculations, atrophy:
LMN (or muscle)

Hyperreflexia, clonus, increased muscle tone
UMN (Cord or brain)

Some NMJ/Muscle findings?
Weakness, possible fatigability or improvement with use

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Chorea Hyperkinetic, inappropriate, "purposeless" movements

Athetosis Slow, writhing movements typically affecting hands and feet.
Seen in HD and CP (more often "choreoathetosis in CP)

Akathisia Feelings of inner restlessness and inability to stay still

Ataxia "The presence of abnormal, uncoordinated movements"

Dystonia involuntary contraction of muscles

Meds that cause it

Typical antipsychotics, metoclopramide, and prochlorperazine (All dopaminergic)

Torticollis Neck Dystonia

Treatment of Dystonia Anticholinergics (Benztropine or trihexyphenidyl)
Diphenhydramine (antihistamine)

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Tremors

Tremor that you see while arms are dangling while walking, or sitting with hands in lap

Resting tremor Parkinson's disease vs Secondary Parkinsonism
This is due to lesions in the basal ganglia circuitry
Can treat with dopaminergics or anticholinergics

Dysmetria: over- or under-reaches while aiming for a target – with a compensatory tremor trying to get it right

Intention tremor Due to a cerebellar circuitry lesion
Treated with support and education
Cannot be treated with meds

40-60 yo patient has a tremor with movement, but it worsens as they approach target

Essential tremor Fine tremor, suppressed at rest; exacerbated by goal-directed movement
Treat with Low-dose BB, barbs, gabapentin, benzos (as well as alcohol)

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Parkinson's Disease/Parkinsonism

How do you diagnose Parkinson's? Clinically, just like you diagnose Parkinsonism

4 Features of Parkinson's/Parkinsonism

1. Bradykinesia (slowness/poverty of movement, mask-like facies)
2. Rigidity ("cogwheel," "lead pipe")
3. Resting tremor ("pill rolling")
4. Gait/postural instability ("hypokinetic")

Can also see depression in PD

Pathophys of Parkinson's

Loss of dopaminergic neurons in basal ganglia (esp. substantia nigra)

Mean age of onset 60

When do you see dementia in idiopathic Parkinson's disease? Late in the course

When can you see dementia before Parkinsonism?

Lewy Body Dementia, Progressive Supranuclear Palsy, Multisystem Atrophy

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Treatment options for Parkinson's

- Dopamine precursors (levodopa with carbidopa)
- Anticholinergic (benztropine, trihexiphenidyl), use on young people
- COMT inhibitors (-capones)
- MAOb-I (selegiline) – rarely used anymore. Serotonin syndrome or hypertensive crisis
- Dopamine agonists (Ropinirole, pramipexole)

What is the function of carbidopa?

In periphery, it prevents conversion of L-dopa into dopamine

Common AEs of Sinemet (Levodopa/Carbidopa)

Somnolence, confusion, hallucinations, dyskinesias

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Pt presents with signs/symptoms of parkinson's, how do you treat…

If <70 and/or not showing decreased function?

Start dopamine agonists (ropinirole or pramipexole).
Hold off on L-dopa/carbidopa until absolutely necessary

If >70 or showing decreased function?

Start with L-dopa and carbidopa, then bring in the COMT-I or (maybe) MAOB-i

When to use Trihexiphenidyl or Benztropine?

Use anticholinergics for younger Parkinson's patients who predominantly have tremor or…

To treat Parkinsonism s/p antipsychotics (or can use diphenhydramine)

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Weakness – "paresis"

Can be localized to UMN, LMN, NMJ or Muscle

Tests we can use to work these up?

EMG
Nerve conduction studies
Muscle biopsy
CK, myoglobin
special labs

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Guillain Barre

H/o mild URI 1 week prior to the onset of symmetric feet and lower leg weakness.

Guillain-Barre syndrome. Postinfectious polyneuropathy. Typically symmetric

Cause? Usu. follows minor infection (esp. URI) or an immunization

MC bug? Campylobacter jejuni (watery/bloody diarrhea)

Diagnosis? LP normal except lots of protein
Ig for Anti-Ganglioside (e.g. GM1) antibodies

Sensation? Intact or minimally impaired. Maybe mild paresthesias beginning in feet/legs.

Reflexes? Diminished or lost

Nerve conduction velocities: Slowed

Feared complication? Respiratory paralysis

Tx: Usually resolves spontaneously, Plasmapheresis (adults), IVIG (kids) to shorten duration
(DO NOT GIVE STEROIDS)

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Proximal Muscle Weakness

Can't rise from a chair, go up stair, comb hair, get things down from there

Proximal muscle weakness
Presents subacutely and painlessly

+Purple discoloration around eyes/Heliotropic rash –

Dermatomyositis

May also see Grotton's papules (red over knuckles)
or shawl sign (red over neck, upper back and/or upper chest)

Other myositis: Biopsy can differentiate dermato/pyo/inclusion body

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Proximal Muscle Weakness - Lambert Eaton

What proximal muscle weakness is associated with an antibody?

Lambert-Eaton myasthenic syndrome
The antibody is to a presynaptic CaCh controlling ACh release from the presynaptic membrane

Association? SCLC in 50-70% of LEMS patients

Classic age 50+

Dx: ABs. Also do a CT to look for SCLC

Tx If paraneoplastic, treat the cancer
SCLC will be chemo-sensitive, and can tx with Azathioprine or 8-Mercaptopurine

Other treatments? Steroids and IVIG can work, but not as well as for other conditions

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Myasthenia Gravis

70 yo man presents with double vision, and difficulty swallowing during dinner conversation?

Myasthenia Gravis Antibodies to AChR on *motor endplate*

Can contract initially because inhibitor is overcome, but when ACh vesicles run out, there's *fatigability* → worsening weakness with repeated use or near end of day

Age Range? Men get it in 6th to 8th decade. Women from 2nd to 4th decade.

Muscle groups affected Extraocular (ptosis, diplopia), nonpainful (15-17% have only ocular sx)
Bulbar (fatigable chewing, dysphagia, nasal speech)
Later: arm weakness
Respiratory muscles

Association? Thymoma. Can get dysphagia with oropharyngeal regurgitation

Dx? Anti-AChR AB, then EMG → Great amplitudes that fatigue and then stop contracting,
Then CT scan for a thymoma
(May mention an ice pack on closed eyelids reversing symptoms)
(*Edrophonium* or "Tensilon" tests (AChE inhibitor) no longer done d/t risk of asystole)

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Myasthenia Gravis

What's the feared complication?

Respiratory difficulty signifying Myasthenic crisis.
For some people, this can be an early manifestation of MG.

Precipitating factors for myasthenic crisis

Infection Surgery Pregnancy
Medications – Aminoglycosides (e.g. Azithro), fluoroquinolones (e.g. Cipro)

Tx: Increase ACh - AChE inhibitors (stigmines)
If stop responding to stigmines, use steroids (or mycophenolate, azathioprine, cyclosporine)
If in myasthenic crisis (can't swallow or breathe) - IVIG plasmapheresis
Intubate to protect airway
May also do thymectomy

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Muscular Dystrophies

MC type of muscular dystrophy? Duchenne
XLR mutation in the dystrophin gene Therefore more common in boys

Presentation Age 3-7
Muscle weakness (esp proximal leg) Calf hypertrophy
Gowers' sign (have to "walk" body up from squatting position)

Dx Muscle biopsy Labs? Increased CK

Tx Supportive (average life expectancy is mid-20's)

Other high-yield types of muscular dystrophies

Becker XLR Dystrophin, but milder presentation, later onset (10-20yo)

Myotonic AD, 20-30 yo, baldness, MR, testicular/ovarian atrophy,
Can't release grip or handshake (myotonia)

What class of inherited metabolic disorders affect muscle & may resemble muscular dystrophy?

Rare glycogen storage diseases (AR) esp McArdle
(glycogen phosphorylase def → weakness/cramping after exercise 2/2 lactic acid buildup)

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Other disease with motor manifestations

Areflexia, weakness, fasciculations… Also hyperreflexia, weakness

UMN and LMN lesions of ALS
Sensation will be spared
Dx and Tx? Dx: EMG
Tx: Supportive. Riluzole only extends life by 2-3 months.

Irresistible need to move legs, due to crawly, tingly feeling

Restless legs syndrome
Tx Dopamine agonists (ropinirole/pramipexole) or gabapentin

Someone has ataxia, seems to be confused, and has noticeable nystagmus

Wernicke Encephalopathy
Confusion, Ataxia, Nystagmus
Classically alcoholic patients, but can be seen in anorexic patients
Give glucose *with* (not before) thiamine

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Headache

Patient says "my head hurts" Consider headache :)

Red Flags Fever, focal neurologic deficit, thunderclap, new onset at age 50+,
Progressive nausea/vomiting, wakes pt from sleep

Ptosis, miosis, and eye pain that wakes them from sleep

Cluster headache

Can go asymptomatic for months, then maybe 1-2 HA/day with the same presentation
Can resemble Horner's syndrome (unilateral eye pain)
Can see rhinorrhea, lacrimation, conjunctival inflammation

Tx Improves with inhaled oxygen
Do brain imaging before prescribing verapamil

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Headache

Bilateral vise-like pain, starts in front of head and radiates to neck

Tension Headache "constant temporal/occipital pressure"
No photophobia, phonophobia or altered mentation
Exacerbated by noise and/or exercise

Tx OTC meds.

But it keeps getting worse, despite trying NSAIDS, triptans, even opioids every day?

Analgesic Rebound Headache

Regularly using analgesic and gets HA when stopping medication

Tx Let them withdraw/wean off, and the headache will resolve

Headache, somnolence, and confusion in elderly patient with multiple falls

Chronic subdural hematoma

No clearly defined treatment guidelines

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Headache

Unilateral pulsatile headache, maybe with photophobia, phonophobia, nausea, vomiting

Migraine Often has trigger: Menstrual cycle, chocolate, caffeine, MSG
Aborts with sleep

Dx:. Clinical

Acute Tx: NSAID, then can escalate to triptans
Avoid Ergots in CAD patients – can precipitate vasospasm
(Avoid fioricet or butalbital-containing compounds due to risk of tolerance, dependency and withdrawal seizures)

Prophylactic Tx: Avoid triggers
Beta blockers, anticonvulsants (Topamax, valproate, TCA)

Side effects of TCAs:
- Anticholinergic (dry mouth, constipation, urinary retention)
- Alpha adrenergic (orthostatic hypotension)
- Histamine (lethargy)

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Intracranial Hypertension

Headache worse at night or in mornings, nausea/vomiting, papilledema/blurry vision, Maybe FNDs. Vision can worsen when leaning forward.

Intracranial hypertension

Increased ICP from mass effect of Tumors
Hemorrhages
Abscesses
Inflammation (incl meningitis)
Idiopathic
Hydrocephalus

Other symptoms can include pulsatile tinnitus (whooshing in ears) or periorbital pain

What is a systemic sequela of increased ICP

Cushing's Triad bradycardia, hypotension, irregular breathing

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Intracranial Hypertension

If those symptoms, plus focal neurologic deficit, progressive N/V Consider Tumor

Most common intracerebral malignant tumor

Mets, either from very common tumors (breast and lung)
or rarer tumors with neurotropism (melanoma, RCC)

Description of brain mets
Discrete, well-circumscribed mass(es) at the gray-white junction

Treatment of single brain metastasis
Surgical resection

Treatment of pt with multiple brain metastases
Whole brain radiation

MC *Primary* intracerebral malignant tumors?

Glioblastoma, followed by lower grade astrocytomas, followed by everything else

Tx Surgery, chemo, and radiation

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Intracranial Hypertension

If it's someone on doxycycline or isotretinoin A, or an obese young woman

Idiopathic Intracranial Hypertension (IIH) aka Pseudotumor Cerebri
Can also see with corticosteroid withdrawal.

Diagnosis Imaging first to rule out structural cause of ICH
LP (opening pressure >25cm H2O) can be diagnostic and therapeutic

Treatment: Depends on symptom severity:
Can do supportive (analgesia, encourage weight loss)
Acetazolamide +/- Furosemide
Serial LPs (should get relief with LP)
Corticosteroids can be used to bridge
Definitive treatment: VP shunt via surgery

MC complication if left untreated? Vision loss

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Headaches - Other

High acuity HA, rapid progression Meningitis Headache Consider headache :)

Will have fever, +CSF, likely WBC count and + Kernig's/Brudzinski's

"Worst Headache in My Life"

Subarachnoid Hemorrhage (SAH)

MCC Congenital berry aneurysm rupture or trauma

Diagnosis CT/MRI will show blood around brain or within sulci
LP will have grossly bloody CSF

Treatment Supportive, but any aneurysm will require surgical treatment

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Patient is Found Down

Use mnemonic SVncOPeS

Seizure (including nonconvulsive status)
Vasovagal *most common
Aka **N**euro**C**ardiogenic
Orthostatic
Pulmonary **E**mbolism
Stroke

If Nauseated, diaphoretic, pale, or bradycardic before the syncopal episode

Vasovagal (aka neurocardiogenic) syncope

Cause: sudden drop of BP to brain, due to neurocardiogenic factors/reflex

Possible triggers Neurogenic: exaggerated vagal response to micturition, pain, stress, emotions, needles
Cardiogenic: nonperfusing arrhythmias, prolonged standing → reduced cerebral bloodflow

Pt with clot risk, found down with tachycardia, tachypnea, "stabbing" chest pain

Consider **Pulmonary Embolism** – atypical presentation, but can present this way

Fainting after rising from bed or chair **Orthostatic** syncope

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Seizures

Causes of provoked seizures VITAMIN C

Vascular
Infection of CNS
Trauma (incl cerebral edema from severe/malignant HTN, eclampsia, pheochromocytoma)
Autoimmune – Limbic encephalitis incl. Anti-NMDA
Metabolic – Hypoglycemia, hypoxia, hyponatremia
Ingestion/Withdrawal - Ingestion of diphenhydramine, bupropion, cocaine, meth, tramadol, isoniazid
Withdrawal from booze, barbs, benzos or too rapidly stopping anticonvulsant
Neoplasm – Often due to the mass effect (same with some abscesses)
Congenital – Juvenile Myoclonic Epilepsy (JME), Lennox-Gastaut, West Syndrome

First seizure, no obvious provoked cause - what do you do next?

CBC, urine tox, and CT imaging If 2 unprovoked? Consider epilepsy diagnosis

Seizure and fever – next step

Brain imaging first – new onset seizure would likely be focal. Then can do Lumbar puncture

How are seizures induced?

Make them stay awake, use strobing lights, have them hyperventilate

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Seizures - Types

Pt gets really stiff and adopts an unusual posture, then begins shaking

Primary Tonic Clonic (formerly generalized or "grand mal")

Can last up to 2-5 minutes
Can see incontinence, tongue lacerations
Followed by postictal state: drowsiness, confusion, headache, sore muscles

Focal seizure Can be motor (Jacksonian march), sensory (hallucinations) or psychic (cognitive or affective changes)

No longer say "simple" or "complex"
Instead: "without (or with) cognitive impairment

Focal onset with spread to bilateral tonic clonic (formerly Secondary Generalized)

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Seizure >30 min long episode or 30 mins of episodes closely timed so person doesn't regain consciousness in between

Status epilepticus

Causes

Spontaneous, or withdrawing anticonvulsants too quickly

How do you treat it?

Benzo → Benzo → IV antiepileptic (e.g. fosphenytoin)
→ Anesthetic induction (intubate and sedate to the level of burst-suppression coma)

Major risk of prolonged status epilepticus

Cortical laminar necrosis

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3-year old… with a fever… who has tonic-clonic shaking

Febrile seizure
6mo-5yrs
-Tx underlying cause of the fever (acetaminophen to reduce fever – don't give aspirin!)

12-year old, excitedly talking about something and then stops for 5-10 seconds while blinking her eyes and not responding to voice, then resumes.

Absence seizure
Don't begin after 20 years old
5-10 seconds long
Will have LOC, but no posticta state
Tx Ethosuximide

Person with many unwitnessed seizures – explosive onset without apparent cause, negative workup, and not responsive to multiple medications, maybe wide variation in frequency

Psychogenic aka Pseudoseizures – do 24 hour video EEG inpatient

Atypical phenotype or atypical response to medications can also suggest psychogenic events

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Strokes

(Note: Stroke localizations were covered earlier)

Classes of stroke
Hemorrhagic (SAH, IPH)
Ischemic (can subdivide into thrombotic or embolic (with A Fib/valve issues))

Stroke code criteria: sudden onset of focal neurologic symptoms

Risk stratification for stroke: CHADS2 score
(CHF, HTN, Age >75, DM, Strokex2)

Most important risk factor for stroke
Hypertension

MC type of stroke: Ischemia due to atherosclerosis (atherothrombotic ischemia)

Exotic causes of stroke: Afib → clot & emboli to brain
Endocarditis → septic emboli
Sickle cell

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Strokes

Diagnosis of stroke: New onset of FND(s)
→ appropriate imaging (CT/ultrafast MRI) to exclude hemorrhage

Acute tx for all strokes: Supportive (Airway, oxygen, IV)

Tx - hemorrhagic stroke: Lower BP (esmolol or other BB)
Call neurosurg (clip, coil, craniotomy)
FFP to reverse anticoagulation if they're on it

Tx - ischemic stroke: tPA as soon as possible within guidelines
always within 3 to 4.5 hours of last known well, (depending on risk factors)

Contraindications to TPA: Surgery within 2 weeks, Intracerebral hemorrhage, on anticoagulation (risk of bleeding), BP>185/110 or if septic cerebral embolism (use abx instead)

Best treatment to reduce risk of recurrent stroke
Antiplatelet unless they have AFib, then anticoagulant

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Strokes

Berry/Saccular Aneurysm Risk Factors:
Chronic HTN or Connective Tissue Disorder
MC Location: anterior communicating artery, can be seen in MCA

Risk factors for lipohyalinosis/lacunar infarcts: HTN, HLD, diabetes and smoking

Treatment of Carotid Artery Stenosis: Medical: Long-term therapy (ASA, statin, BP control)
When do to do carotid endarterectomy?
if >70% or symptomatic (stroke or multiple TIAs)

Patient on Warfarin has hemorrhagic stroke. What do you give?
Prothrombin complex concentrate

Stroke that seems to be worsening - how do you diagnose?
CT - It's likely hemorrhagic (SAH or ICH) so diagnose it quickly

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Other Brain Bleeds

Crescent-shaped hyperdensity that crosses suture lines

Subdural hematoma

Cause? Rupture of the bridging veins

Risk factors? Brain atrophy
Alcohol
Anticoagulant use
Acute trauma

Tx Surgery

Biconvex/lens-shaped hyperdensity on cranial imaging

Epidural hematoma.
May see "talk and die" syndrome

MCC: Middle Meningeal Artery rupture d/t head trauma
Typically in temporal bone area

Tx: Urgent craniotomy or burr hole

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Back Pain

20-50 yo pt presents with back pain after heavy lifting, no specific neurologic symptoms.
(May also be described as "belt like")

MSK back pain.

Treatment

NSAIDS, exercise, and stretching, and reassess if improved in 4 weeks.

If not improved, or with neurologic symptoms
assess for more serious cause: X-ray and MRI

Still belt-like pain, but what's difference between presentation for MSK back pain vs. Herniated disk?

Disk has sciatica and positive straight leg test
X-ray can help diagnose

Initial Tx: Conservative therapy – will often improve on its own

Tx for herniated disk with symptoms Laminectomy and fusion

Lower back pain that also has pain/tingling/weakness going down leg, possibly to feet? (Can still be a/w heavy lifting)

Sciatica

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Older person who falls, has no sciatica, but has vertebral step-offs and pinpoint tenderness.

Compression fracture. Note it's a fracture, not cord compression!
More likely in a woman due to lower bone density).
Treat the osteoporosis, and can repair surgically.

Older person >50 with sciatica pain but not lower back pain, relieved by leaning forward.

Spinal stenosis. Presents with "pseudo-claudication" (usually buttocks and thighs), a positional claudication that is worse when they're upright (but not when bending forward). Worsens when walking downhill, and improved with walking up stairs.
Dx: MRI > Xray
Tx with laminectomy

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Back pain with saddle anesthesia, new neurologic deficits, bowel/bladder problems, or rapid progression
Often affects lower limbs – can see loss of sensation and UMN signs

Spinal cord compression (multiple etiologies)

Tx of cord compression

Steroids (dexamethasone, methylprednisolone)
Do X ray, MRI
Then decompress surgically (drain hematoma, I&D abscess, or irradiate cancer)

Low back pain, radicular pain, anesthesia in perianal area and maybe the inner upper thighs, and altered knee and/or ankle-jerk reflexes

Cauda equina syndrome or conus medullaris syndrome
These both occur in lumbar spine (cauda can go down to sacrum)

Key distinctions?

Conus medullaris syndrome has earlier sphincter dysfunction, and less overall pain.
Cauda equina (due to anatomy) more likely asymmetrical.
The cauda equina nerves are considered "spinal nerve roots"

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Multiple Sclerosis

Relapsing, remitting neurologic lesions occurring at different times

Common presenting symptoms

Optic neuritis (CN2) - often found in newly-diagnosed MS

Diagnosis

Periventricular white matter (perhaps in plaques).
Can also see oligoclonal bands of IgG
Evoked potentials

When do you do LP in MS?

When diagnosis is unclear

Exacerbating factor for MS

Moving to warm climate - Uhthoff Phenomenon (HT = Hot temperatures)

TIA vs MS

TIA resolves within 1 day. MS deficits last for days to weeks

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Multiple Sclerosis

MS Patient presents with new neurologic deficits:

MS Exacerbation

Acute Tx Steroids

Disease-modifying treatments

First generation: **Interferon beta, glatiramer acetate**
Biologics: monoclonal antibodies (-ab)
Oral small molecules: **fingolimod,** dimethylfumarate

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CSF Analysis

When is LP contraindicated?

Acute head trauma, signs of intracranial HTN, SAH suspicion, focal neurologic signs

Obtain a negative CT or MRI first. Otherwise → uncal herniation and death

Scenario	Protein mg/dL)	Glucose mg/dL	WBC/uL	Pressure cmH_2O	Other/Notes
Normal	20-45	50-100	0-3 (Lymph)	5-20	
Bacterial meningitis	↑ >100	↓ **<50**	100- 10K (L)	↑ >30cm	
Viral meningitis			100-2,000 (L)	NI or ↑	
Cerebral Hemorrhage	↑ >45		Bloody (RBC)	↑	
IIH/Pseudotumor Cerebri				↑ > 25	
Multiple Sclerosis	↑		0-50 (L)		Oligoclonal IgG bands
Guillain-Barre Syndrome	↑ >100		0-100 (L)		
Creutzfeldt-Jakob Disease					14-3-3 protein

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Classic Imaging Descriptions

Periventricular white matter lesions on MRI

MS most likely

"Blurring of the gray-white junction"?

Diffuse axonal injury
Due to serious deceleration trauma such as car accident

Atrophy of caudate nuclei due to loss of GABAergic neurons

Huntington's Disease

Diffuse atrophy which is disproportionately greater in parietal and temporal lobes

Alzheimer's Dementia

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Ring-Enhancing Lesions MAGIC DR mnemonic

Metastasis
Abscess MC S aureus, S viridans or GNs
GBM
Infarction (acute)
Contusion
Demyelination
Radiation Necrosis/Resolving Hematoma

What if it's an HIV patient?

Toxoplasmosis and CNS lymphoma
move way up list

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Miscellaneous Disease

Body temperature >40C (104F)

Severe heat exhaustion leading to heat stroke: either exertional or due to dehydration

Tx Rapid cooling

Best = immersion in ice-cold water
2nd line = Cool shower or rotating cold blankets

Dizziness/lightheadedness/N/V while doing work with arms overhead or playing tennis

Subclavian steal
Atherosclerosis of subclavian artery proximal to vertebral artery
Dx with doppler US or MRI
Left more affected than right d/t sharper curvature and more turbulent blood flow

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Younger patient with elevated liver enzymes, and some of: tremor, rigidity, depression, paranoia, catatonia

Wilson's disease. Dx with serum ceruloplasmin, urinary copper, and slit-lamp eye exam (Kayser-Fleischer rings)

Insensitivity to pain, BP and temperature lability, overflowing tears
Frequent pneumonia and poor growth

Familial dysautonomia (Riley Day syndrome)

Hypotonic @ birth (or at least by 6mo), Pos fam hx + long slowly progressive disease course;

Spinal Muscular Atrophy (Werdnig Hoffmann)
AR degeneration of ant horn cells in spinal cord & brainstem (LMN)
Treatment is supportive only

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Mental status change, rigidity, fever and autonomic dysregulation

Neuroleptic malignant syndrome.
Seen with dopamine antagonists and antipsychotics

Parkinsonism with orthostatic hypotension, impotence and/or autonomic dysfunction

Multiple system atrophy (Shy-Drager's syndrome)

Choreiform movements and intellectual deterioration in a 40-year old

Huntington's Disease
AD, presents btwn 35-50
Choreiform mvmts, progressive intellectual deterioration, dementia or psych disturbances
Dx with CT/MRI
Supportive treatment. Antipsychotics might help

5-15 year old with areflexia, loss of vibration/position sense, and cardiomyopathy

Friedreich Ataxia, AR mutation in FXN/Frataxin gene

Person with immune deficiency gets progressive cerebellar ataxia and you see vascular markings on eyes and skin

Ataxia telangiectasia, AR disease

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Neurocutaneous Diseases (all AD)

Kid with developmental delay, seizures, rough bumpy skin areas and a few little nodules, hypopigmented skin areas

Tuberous sclerosis. Bumpy skin = shagreen patches, hypopigmented skin = ash leaf spots, nodules = hamartomas

Person has light brown spots and fleshy growths under skin

Neurofibromatosis – café au lait spots, and neurofibromas

If they have hearing loss, dizziness, and signs of increased ICP or FND

NF2 – on Chromosome 22, can see acoustic schwannoma, meningiomas, ependymomas

If they have scoliosis, brownish 1-2mm spots on iris, and learning disability

NF1 – brown spots on iris are fatty lisch nodules, and 50-75% have learning disabilities

Sturge-Weber and Von-Hippel-Lindau are rare, and seem more likely for Step 1 than Shelf or Step 2

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