Primary headaches are symptom-based

- Arising of their own accord and organized according to symptomology or phenomenology
- Migraine
- Tension type headaches
- Cluster headaches

Secondary headaches are etiology-based

- Headaches caused by something else.
- They are classified by what is causing them
- Tumor
- Meningitis
- Giant Cell Arteritis.

90% of headaches presenting to primary care are primary headaches

Migraine is the most common headache type to present to healthcare settings although it is not the most prevalent (tension type headache is less severe)

3 Question Screen

High specificity and sensitivity for migraine

- During the last 3 months did you have the following with your headaches?
  - You felt nauseous or sick to your stomach
  - Light bothered you a lot more than when you don’t have headache
  - Your headaches limit your ability to work, study or do what you need to do for at least 1 day
- Nausea, photophobia and disability

Lipton et al 2003
Testing

- Primary headaches are a clinical diagnosis
- Testing is useful to rule out other disorders in a planned and purposeful way
- If criteria for migraine are met and the neuro exam is normal, there is a 0.18% chance that imaging will change management

When would we consider additional imaging?

- Unusual, persistent or prolonged aura
- Change in clinical features, severity or frequency
- First or worst migraine

Red flags

- Systemic symptoms (fever, weight loss) or secondary risk factors (HRV, systemic cancer)
- Neurologic symptoms or abnormal signs (confusion)
- Onset: Sudden, abrupt or split second
- Older: New onset and persistent headache in middle age >50y/o (giant cell arteritis)
- Previous headache history: Change in character, frequency, clinical features, severity
- Progressive: Headache getting worse over time and is not responding
- Positions: Better lying down and worse standing up?

What tests to get

- MRI is better than CT.
  - Less radiation which is not negligible
  - Can visualize the posterior fossa better
  - Can visualize the vessels better
- CT when there is a concern for bleed or skull-based fracture
- Contrast for neoplasm, infection or intracranial hypotension
- May also benefit from a C-spine

Migraine Aura vs TIA

Aura

- Positive symptoms (lights)
- Gradual evolution
- Sequential evolution
- Repetitive attacks which are similar
- Headache follows ~50%
- 95% of people with aura also have visual aura

TIA

- Negative symptoms (numbness)
- Abrupt onset
- Simultaneous onset
- Different, rarely in the same vascular distribution
- Headache is uncommon

Treatment of Migraine

- Almost everyone needs an abortive
- Only a subset needs preventative treatment, but probably more than we do currently
Lifestyle modifications

- Regular meals
- Adequate and scheduled sleep
- Regular exercise
- Normal body weight prevents chronification
- Limit caffeine - somewhat controversial
- Alcohol – consistent trigger for almost all patients
- No good evidence for dietary restrictions, but may be individual per patient

Acute treatments

Tension-type headaches

- NSAIDS and mild analgesics

Migraine

- Effervescent ASA
- NSAIDS and mild analgesics
- Triptans (try a few, not always contraindicated with SSRIs)
- Ergit derivatives
- Antimetics

Alcohol

- Consistent trigger for almost all patients

Limit caffeine

- Somewhat controversial

No good evidence for dietary restrictions, but may be individual per patient

Prophylactic treatment

- HA more than 2/week
- Symptoms are particularly bothersome
- Cannot tolerate SE of acute abortive treatments
- Overuse of acute abortive treatments

- Start Magnesium 500mg QHx
- CoQ10 200mg BID
- Riboflavin
- Butterbur

How to choose a treatment

An effective trial is 2 months at target dose

Side Effect concerns

- Weight gain
- Hypertension
- Cognitive
- Possibility of pregnancy

Choices

- Amiodarone
- Propranolol
- Venlafaxine
- Topamax and venlafaxine
- Avoid BP meds
- Avoid antiepileptics and antihistamines
- Avoid Depakote, Topamax, Lisinopril and candesartan

Triptans can cause...

(may last 30 min and often resolves with HA resolution)

- Tingling
- Warm
- Flushing
- Chest discomfort
- Dizziness

Optimizing abortive treatments

- Adequate dose, no reason to use sub-maximal dose except for SE control
- Early to mid stage of HA, before central sensitization sets in (cutaneous allodynia)
- Avoid overuse, more than 10 days per month
- Avoid combination treatments for first line therapy
When to refer

- Diagnosis is unclear
- If the diagnosis is challenging to manage e.g. Idiopathic Intracranial Hypertension
- Multiple acute or preventative strategies have failed
- Advanced treatments are required: Botox, Nerve blocks and infusions
- (Significant psychiatric comorbidities need a referral to psych not to headache)
- A multidisciplinary approach is optimal for refractory or challenging migraine cases

Case #1

- Lisa is a 38 year old woman with a long history of migraines. Her migraines have always characterized as unilateral throbbing head pain with scintillations which may or may not be associated with the headache. Today she presents to your office with complaints of increased migraine frequency and an aura which in addition to the scintillations, caused paresthesias in the left side of her face.

- What do you do?

- A patient presents to your office complaining of frequent disabling headaches. The diagnosis highest on the differential is...?

- Other strong diagnoses to consider include

- When should we consider preventative medications?

- You would be well advised to avoid which medications when treating headache?
SEIZURES AND SPELLS

Epilepsy is two or more unprovoked events
Or one event with specific biomarkers that increase the chance of future seizures

Focal seizure
Generalized seizure

Risk factors for seizures
- head trauma,
- febrile seizures,
- meningitis,
- encephalitis,
- status epilepticus,
- stroke,
- intellectual disability/developmental delay,
- CNS hemorrhage,
- brain tumor, and
- family history of seizures,
- reports product of normal birth with normal development.

Work-up
- Labs
  - CMP
  - CBC
  - A1C
  - TSH and free T4
  - Magnesium
- MRI with contrast
- EEG (1 hour, sleep-deprived with activating techniques)

No seizures
No side effects
No lifestyle limitations (participating and productive members of society)

Choosing a medication
- Identify the seizure type
- Age of patient
- Comorbid disorders
- Potential adverse effects
- Cost of therapy
- Patient compliance
The first medication is always the best (57% respond)
- The second medication is second best (7.3% respond)
- The response to the first 2 drugs is highly predictive of medical outcome
- About 1/3 of patients will have difficult to treat epilepsy
- Newer drugs are not necessarily better or more efficacious

Generic drugs
- FDA requires "bioequivalence"
- Same active ingredient
- Bioavailability 80-120%
- Not all generics are identical
- In most cases generics are adequate

Intractable epilepsy
- Surgery: Lesion resection or focal cortical resection
- Electronic stimulation: Vagus nerve stimulation or deep brain stimulation
- Ketogenic diet
- Clinical trials

Holistic management
- Evaluate and treat the patient with a seizure disorder
- Educate the patient, family members and caregivers
- Discuss issues of daily living e.g. safety in the home, driving, employment, birth control, EtOH and AED compliance
- Map a strategy of care (what to do when, when to go to the ED, doc call routine and urgent)

Consult
- Consultation is advised if diagnosis is uncertain
- If not controlled on 2 or more drugs

Case #2
- Latisha is an 18 y/o woman with epilepsy, who is maintained on Keppra and Trileptal. She has had 3 seizures since her last office visit with you 8 weeks ago.
- What is the first question that you have for this patient?
- What other factors can contribute to breakthrough seizures?
- What labs are you going to want to run
If someone has pseudoseizures does that mean that you have ruled out seizures? How do we manage pseudoseizures?

- When should you send a seizure patient to the ED?

Neuropathies
- Length dependent – (DM II)
- Single nerve – (compression i.e. carpal tunnel)
- Diffuse – (Guillain-Barre/ CIDP)
- Multifocal – (Guillain-Barre, paraneoplastic)
- Neuropathies with prominent dysautonomia
- Neuropathies with prominent ataxia

Describing Sensory Neuropathies
- Hypoesthesia – Decreased sensation
- Hyperesthesia – Increased sensation, no pain
- Paresthesia – Pins-and-needles, tingling, burning, electrical, stabbing, stringing
- Allodynia – Pain from non-noxious stimuli

Intermittent or Continuous?
- Most neuropathies are continuous
- Intermittent include compression neuropathies and psychiatric symptoms
Length dependent sensory > motor

- Diabetic ~1/3
- Alcoholic ~1/3
- Migraine ~1/3
- These seldom become seriously disabling as there is not a lot of motor involvement

Other neuropathies in this class include Vitamin B12, amyloidosis, paraprotein related confluent vasculitis, HIV and tox (chemotherapy and HIV)

Simple Neuropathy

- >50 y/o
- Started in toes of feet
- Symmetric
- Slowly progressive
- FBS or HbA1C
- B12
- SPEP and immunofixation
- Thyroid function tests yield <1% if no other Sx of hypothyroid

Younger patient/ Not so straight forward

- FBS or HbA1C
- B12
- SPEP and immunofixation
- Thyroid function tests yield <1% if no other Sx of hypothyroid
- FBS or HbA1C
- B12
- SPEP and immunofixation
- Thyroid function tests yield <1% if no other Sx of hypothyroid
- FBS or HbA1C
- B12
- SPEP and immunofixation
- Thyroid function tests yield <1% if no other Sx of hypothyroid
- FBS or HbA1C
- B12
- SPEP and immunofixation
- Thyroid function tests yield <1% if no other Sx of hypothyroid

Also...
- DM/NC1
- Creatinine
- FSR, ANA, rheumatoid factor, LFTs
- Hepatitis (HBV and HCV serology)
- HIV (if positive then CMV)
- Vitamin E

Neuropathic pain medication

- Neuropathic pain is caused by a heterogenous group of conditions
- Current drugs do not address specific mechanistic etiologies or clinical phenotypes
- Few head to head comparisons, variable outcomes and much extrapolation

Medical Interventions

- Medications for pain:
  - Gabapentin
  - Lyrica
  - Cymbalta
  - Amalgam

NOT NARCOTICS for nerve pain
- B12 supplementation if low

When to refer

- Unclear etiology or diagnosis
- Complicated polypharmacy
- If you find yourself ordering tests that you are unable to interpret then someone else should probably be following the patient
Case #3

- Jack is a 72 year old gentleman with a medical Hx significant for cardiovascular disease, stroke and obesity. He presents to your clinic with complaints of imbalance and frequent falls. He has no other complaints. His neurological examination is intact with the exception of a profound sensory neuropathy in his distal lower extremities.

- What would explain Jack’s falls?
- What workup would be appropriate?
- What medication would you suggest?

**DEMENTIA**

**Initial approach**

- Speed of onset
- Nature of symptoms (fluctuating awareness, cueable memory, language)
- Associated symptoms (hallucinations)
- Precipitating event
- Lifestyle and medical Hx (alcohol, Hx of chemo or cancer, diabetes, psych Dx)

**Differential if broad**

**Rapid onset (<2 years)**
- Prion
- Metabolic
- Depression
- Rapid onset Alzheimer’s
- Infectious

**Insidious onset**
- Alzheimer’s
- Vascular
- DLB

**Initial workup**

**Labs**
- CMP
- CBC/diff
- THS and free T4
- B12 and methylmalonic acid
- HIV

**Other Studies**
- EEG
- MRI without contrast
- PET of questionable value
- Neuropsychometric testing

Clock: Parkinson’s with dementia

Clock: CBD
PET scan for
dementia

Right hemisphere dysfunction
- visuospatial
- expression of emotion
- social
- memory
- judgment
- reasoning

Medical interventions
- Treat reversible causes
- Acetyl- and butyrylcholinesterase inhibitors: donepezil, rivastigmine and galantamine
- Memantine (modest glutamatergic, but many other neurotransmitters)
- B3, new data shows that this may be of some benefit 500-1000mg daily
- Titrated over time
- Response is subtle
- Maximum dose is often required to see benefits
- No data out from 1 year

My father is demented ....
- Please can you write a letter saying that he does not have the capacity to make good medical decisions so that I can care for him.

When to refer
- Suspicion for dementia other than Alzheimer's or reversible cause dementia
- The diagnosis is in question or advanced work-up is desired
- Clinical trials

Case #4
- Joe is a 72-year-old gentleman with a history significant for stroke, HTN and DMII. He is not certain why he is seeing you in clinic today but states that things are going well and he has no problems. His family reports that he is becoming more forgetful and having trouble with processing speed. His family note that he has visual hallucinations of fully formed people in their house, who are non-threatening and non-interactive. He gets upset when his wife does not lay a place at the table for them at suppertime. Some days are better than others. He acts out his dreams.
- His neurological examination is significant for 2/5 word recall with 5/5 on cuing. He has a slow shuffling walk with shortened stride length and en bloc turning. There is some rigidity in his upper extremities.
- What is the most likely diagnosis?

- Patients with vascular dementia typically present with
  A. Fluctuations and fluctuating levels of awareness
  B. Poor short-term memory which does not respond to cues
  C. Lower extremity parkinsonism and white matter changes on MRI
PARKINSON’S DISEASE

Motor symptoms and diagnosis

- 4 cardinal signs:
  - Slowness (always)
  - Stiffness
  - Tremor (sometimes)
  - Falls (LATER)

- Resting tremor is low frequency 4-6 Hz, at rest
- Postural tremor is more controversial to define, generally remitting

Parkinson-Plus Syndromes
Your positive DAT scan will include:

- Synucleinopathies
  - PD
  - DLB
  - MSA
- Tauopathy
  - PSP
  - CBD

PD is the primary care of neurology!
Non-motor symptoms

- Anosmia (loss of smell)
- Sialorrhea (drooling)
- Autonomic dysfunction
- Constipation/gastroparesis
- Neurogenic bladder
- Sweating
- NOH
- Erectile dysfunction
- Cognitive dysfunction with prominent executive dysfunction
- Hallucinations, generally visual but not always
- Depression and anxiety (may be wearing off)
- Aphasia
- REM Sleep Behavior Disorder (RBD)
- Tresnia and sleep consolidation issues
- RLS

Medication

Dopaminergic agents
- Carbidopa-levodopa
  - 25/100 IR
  - 50/200 CR (NOT double)
  - Duopa intestinal gel

- Dopamine agonists
  - Pramipexole
  - Ropinirole
  - Rotigotine patch
  - Apokyn SQ

Other agents
- Amantadine
- MAO-B inhibitors
  - Selegiline
  - Rasagiline
  - Xadago
- COMT inhibitors
  - Entacapone

Dosing carbidopa-levodopa

- Titrate up
  - More flexibility with Sinemet 25/100, but may have better tolerability in the older-old with Sinemet 50/200 CR
  - 3 times a day dosing starting from time of waking
  - Dose 4 hours apart (e.g. 6A, 10A and 2P)
  - Evening/nighttime doses generally not needed early on in the disease
  - No real upper limit
  - Doses higher than needed may cause dyskinesias, if not then suspect a P-Plus syndrome
  - Not reserved for the very old or very advanced
When to refer

- When confirmation of the diagnosis is needed
- Any Parkinson-plus syndrome
- Advanced PD
- Complex polypharmacy
- Consideration for Duopa treatment
- Consideration for DBS surgery

- There are many, many community resources be sure to hook your patients into a network that can serve them

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**ESSENTIAL TREMOR**

**Essential tremor**

- This is a med high frequency tremor with action
- Genetic basis, generally there is a strong family history
- Alcohol responsive
- Classic sinusoidal Archimedes spirals

- Etiology is unknown, but it does not appear to be dopaminergic
- We think there is a spectrum from ET to PD with 20% of ET patients developing PD
- None should (and few do) respond to dopaminergics

**Medications, in this order:**

- Propranolol (less or no data for other beta blockers)
- Primidone (dose at night? Maybe)
- Zonisamide (more s/e than primidone)
- Topamax
- Gabapentin
- Sinemet
- Artane

- DBS is an option after more than 2 medications

**Let’s talk DBS**

- Pre-surgery
- Final placement in the OR

**DBS is appropriate for:**

- Patients who have failed multiple medications after a fair trials or cannot tolerate the medications
- Patients who are cognitively intact (MCI is okay, but no dementia)
- Patients who can reliably follow through with follow-up appointments
DBS is FDA approved for...

Approved
- Parkinson’s disease
- Essential Tremor
- Seizures

Compassionate use
- OCD (not hoarding disorder)
- Dystonia

Other uses we consider
- Tourette’s syndrome
- Depression

When to refer
- After the first 2 medications
- When symptoms are uncontrolled and life-limiting
- For consideration of DBS

RESTLESS LEG SYNDROME

What is RLS?
It is not moving or jumping of legs
- Restless leg syndrome is the sensation of creepy-crawly feelings deep in the lower legs bilaterally.
- Discomfort not pain, although patients sometimes describe it as such.
- Sensations resolve with walking and return when sitting or lying down.
- Generally most troublesome at night before sleep and may interrupt sleep.
- As it progresses and with certain medications it can augment.

Where does RLS come from?
- Mostly idiopathic
- Can coexist with PD
- Low iron (Ferritin >50) Some patients still improve with iron supplementation
- Sleep apnea
- Renal failure

Describe the differences in symptoms between neuropathy and RLS

What is RLS?
- Restless leg syndrome is the sensation of creepy-crawly feelings deep in the lower legs bilaterally.
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Where does RLS come from?
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- Low iron (Ferritin >50) Some patients still improve with iron supplementation
- Sleep apnea
- Renal failure
Medications for RLS, in this order

- Dopamine agonists
- Gabapentin
- Horizant (better but costlier)
- Sinemet (caution)
- Narcotics best choice is low dose methadone, but any works

When to refer

- Uncontrolled symptoms
- Patients require progressively increased doses of medication
- Patient is complaining of daytime symptoms
- Complex comorbid conditions (e.g., renal failure)

STROKE AND BLEED

Acute Ischemic Stroke
CT is immediate, but MRI is definitive
(also need MRA head and neck or CTA)

- New strokes and bleeds are ALWAYS a medical emergency and minutes matter
- For every minute that a patient is untreated 1.9 million neurons are lost and irrecoverable (large vessel ischemic stroke)
- 3.6 years of aging per hour untreated - AHA

F - Facial droop
A - Arm weakness
S - Speech
T - Time

Acute Bleed
CT is your best friend

- Stroke-like symptoms (FAST)
- Headache
- Vision changes
- Changes in gait
- Cognitive changes
- Sometimesparesthesias for a thalamic stroke

- Have a high suspicion:
  - Older patients
  - On anticoagulant therapy or anticoagulation (even ASA 81mg counts)
  - Recent fall (especially with head injury or second fall

Strength Testing

- Test strength across joints which is seldom a single muscle group

MRC Grading of Strength

Grade 0: No contraction
1. Flicker or trace of contraction
2. Active movement, with gravity eliminated
3. Active movement against gravity
4. Active movement against gravity and resistance
5. Normal power

Grades 4-4.5 may be used to indicate movement against slight, moderate and strong resistance respectively.
Weakness testing

- Deltoids
- Biceps
- Triceps
- Wrist flexors and extensors
- Finger flexors and extensors
- Hip abductors
- Hip flexors
- Hand grip
- Dorsi
- Foot everters
- E. flexor

Stoke Follow-up
Secondary prevention

- ASA 81mg (seldom value in higher doses)
- Systolic blood pressure less than 130/80
- Triglycerides <150 (generally high-dose statin e.g. Lipitor 80mg)
- LDL <70
- HbA1C <6.8
- Smoking
- Lifestyle changes
- Physical therapy and rehabilitation for deficits

Remember...

CT lights up

- Blood
- Bullets
- Bone

MRI Acute stroke buddies

- DWI (bright)
- DWI_ADC (dark)

Imaging pointers

- Neuroimaging should be hypothesis driven
- Localize the problem from the history and the physical exam
- Think about the possible etiology of the deficit
- Contrast for demyelination, tumor or infection, including recent surgery
What is Bell's Palsy?

- Unilateral facial weakness from CN VII lesion
  - Blaseness, drooling, dry eyes or tearing, pain around the ear; tinnitus, loss of taste, dizziness, difficulty eating
- Acute onset, over hours to a few days
- Peaks at 48 hours – 3 weeks
- May be related to viral infection. Mostly H. simplex but also H. zoster
- May or may not resolve completely (prompt treatment is important)
- Recovery 2 weeks to 6 months. Some never recover completely. But if there has been absolutely no recovery at 4 months then you need to doubt the diagnosis and do some imaging

Diagnosis

- Diagnosis is clinical based on history and examination
- Diffuse paralysis on the facial muscles, generally unilateral. May or may not affect taste, lacrimation and salivation
- The sparing of the forehead may be indicative of a central lesion, although one should be aware that this may also be due to a partial nerve palsy. Also, just to be confusing, central lesions can also take out the forehead area too, by taking out the facial nerve nucleus or facial nerve tract in the pons.

Imaging

At this point you may want to refer...

Imaging is warranted if:
- the physical signs are atypical
- there is slow progression beyond three weeks, or if
- there is no improvement at four months.
- History of a facial twitch or spasm that precedes facial weakness suggests nerve irritation from tumor and should also prompt imaging.

Treatment

Steroids

- Prednisone
  - 60mg x 7 days
  - 50mg x 1 day
  - 40mg x 1 day
  - 30mg x 1 day
  - 20mg x 1 day
  - 10mg x 1 day
  - STOP

Antiviral

- Acyclovir 1000mg TID x 7 days

ALSO...

- Lubricating eye drops
- Eye protection
- OTC analgesics for pain