

NON-EPILEPTIC “SPELLS”



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NON-EPILEPTIC SPELLS



- Motor, behavioral, or sensory phenomenon that is not due to abnormal neuronal activity originating in the cerebral cortex
- Also called Paroxysmal Non-Epileptic Events (PNE's)
- 20% - 30% of children suspected to have seizures who were admitted to epilepsy monitoring units
- Extensive evaluations often performed
- Occasionally started on anticonvulsants
- Diagnosis requires familiarity with these conditions and a *good clinical history*

Remember...



- A seizure is the result of an abnormal electrical event in the brain
- Discharges may be focal or generalized
- Seizures are usually *random* events
- Often *stereotyped* and *interrupt an activity*
- Usually are infrequent (except absence)
- Usually cannot be triggered (except absence)
- Cannot be aborted with verbal or tactile stim.

EVALUATION OF “SPELLS”



- DETAILED HISTORY
- Have parents videotape the episode

HISTORY OF “SPELL”



- Describe evolution of event in detail
- Time of day
- If with sleep...during transition or later ?
- Duration
- Level of consciousness
- Effect of distraction and restraint
- Recall of event
- Post-ictal ?
- Bothered by the episode ?

HISTORY OF “SPELL” (cont.)



- Precipitating event ?
 - change in posture
 - exertion
 - pain, fear, tantrum, trauma
- Feeding history
- Last meal
- Palpitations, chest pain, SOB
- Headache
- Pallor or cyanosis
- Medications

HISTORY OF “SPELL” (cont.)



- Past Medical History
- Family History
- Neurodevelopmental status
- Psychosocial stressors

History that might help

SEIZURE ??

- Interrupts an activity
- Not provoked
- Duration (> 30 seconds)
- Altered responsiveness
- Automatisms
- Post-ictal drowsiness
- Bothers child
- Not aborted
- Incontinence
- Amnesia for event
- Focal head and eye deviation
- Occurs out of sleep

NON-EPILEPTIC SPELL ??

- Low stim environment
- Provoked
- Brief (seconds)
- Preserved consciousness despite bilateral limb movements
- Not post-ictal (esp. if prolonged)
- Not bothersome to child
- Can be aborted
- Only seen by the teacher
- Associated with exertion
- Atypical movements
- Frequent during day but never in sleep

Videotape



6 month old observed to have episodes of hand trembling. Primarily seen when excited or after he eats. Normal development.

Is this a seizure? Why or why not ?
What work-up would you do ?

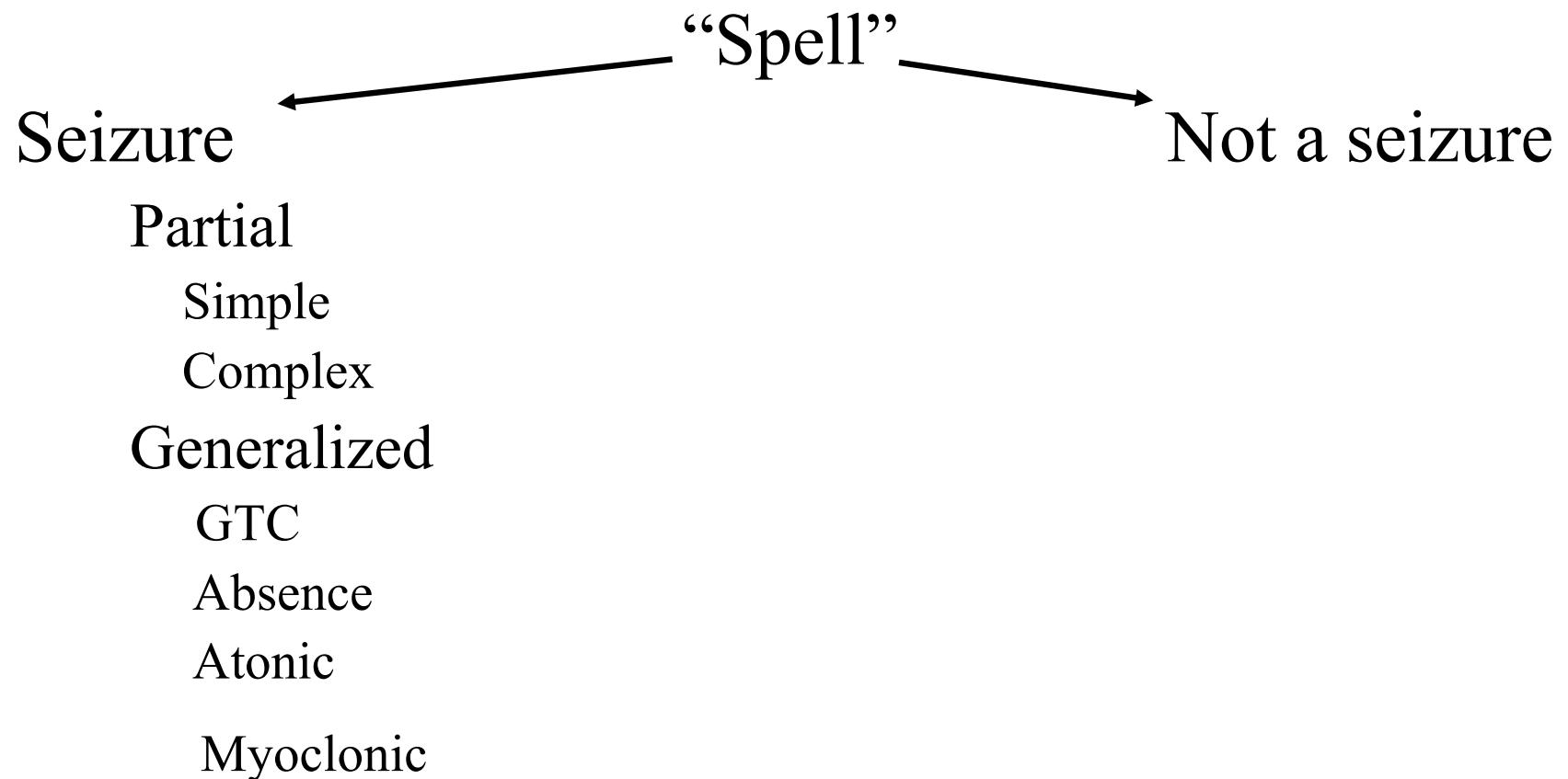


Just a few non-epileptic spells...



- Jitteriness
- Startle
- Clonus
- Sleep myoclonus
- Reflux
- Shuddering attacks
- Stereotypy
- Benign paroxysmal vertigo
- Benign paroxysmal torticollis
- Daydreaming
- Breath-holding spell
- Sleep paralysis
- Rage attacks
- Syncope
- Convulsive syncope
- Acephalic migraine
- Complicated migraine
- Hyperventilation/Panic attacks
- Gratification disorder
- Rhythmic movement disorder of sleep
- Pseudoseizures
- Narcolepsy
- Cataplexy...

The first decision point



How to narrow down ‘spells’



- Age of child
- Was it sleep related or did it occur during wakefulness?
- Family history of similar episodes

“NOT A SEIZURE”

Daytime

Jitteriness
Sandifer
Shuddering attack
BHS
BPV
Stereotypies
Gratification disorder
Daydreaming
Syncope
Migraine
Drug reaction
Narcolepsy
Pseudoseizure

Night / Sleep

Neonatal sleep myoclonus
Rhythmic mvt disorder
Pavor nocturnus
Sleep myoclonus
Somnambulism
Sleep paralysis
Confusional arousal

Infant

Adolescent

“SPELLS”



NEONATES (Birth - 8 weeks)

Wakefulness

- Jitteriness
- Startle or Moro
- Breath-holding spell
- Clonus

Sleep

- Benign neonatal sleep myoclonus

“SPELLS”



INFANTS (2 mos - 2 years)

Wakefulness

- Shuddering attacks
- Breath-holding spells
- Sandifer syndrome
- Stereotypies
- Benign Paroxysmal Vertigo
- Benign Paroxysmal Torticollis
- Gratification disorder
- Benign Myoclonus of Infancy

Sleep

- Rhythmic movement disorder
- Hypnic jerks

“SPELLS”



CHILDHOOD (2 years - 12 years)

Wakefulness

- Breath-holding spells
- Benign Paroxysmal Vertigo
- Stereotypies
- Rage attacks / Episodic Dyscontrol
- Daydreaming
- Dystonic drug reaction
- Tic
- Migraine
- Syncope

Sleep

- Headbanging
- Pavor nocturnus
- Rhythmic mvt disorder
- Confusional arousal
- Nightmares
- Somnambulism
- Hypnic jerks
- Nocturnal enuresis

“SPELLS”



ADOLESCENCE (>12 years)

Wakefulness

- Syncope / Convulsive syncope
- Acute confusional migraine
- Basilar artery migraine
- Pseudoseizures
- Hyperventilation syndrome
- Narcolepsy

Sleep

- Hypnic jerks
- Sleep paralysis
- Confusional arousal
- Somnambulism

Spells during wakefulness



- Age ?
- Circumstance under which it occurs
- Provoked ?

Jitteriness



- Low amplitude, rapid, tremulous movements
- Often seen 1st 3 days of life
- Stopped by gentle flexion or restraint of limb or sucking on examiner's finger
- NO associated apnea or eye deviation
- Stimulus sensitive (increases with crying)
- Seen with HIE, hypoglycemia, hypocalcemia, ICH, drug withdrawal (tremor may be more 'coarse')
- May also occur in first few months of life

Case (Videotape)



Parents are concerned about jerking leg movements.

Was this a seizure ? Why or why not ?
What work-up would you do?
Consult to neurology?



Clonus



- Rapid, rhythmic movement of extremity
- Often noted at ankle or leg in infant
- Might be seen when foot is resting against an object
- Reproduced with quick dorsiflexion of ankle
- R/O CNS pathology if sustained

Case (videotape)



26 year old male with static encephalopathy, spastic quadriplegia and seizure disorder. Recurrent pneumonia due to aspiration. Intern notices frequent “jerks.” “Do we need to increase his seizure medication?”



Startle



- Moro reflex – typically lost by 4 months
- Startle may persist in children who have not lost primitive reflexes
 - Cerebral palsy
- Triggered by sudden noise or stimulus
- Does not cluster in series!
- Reproducible

Case (Videotape)



Neonate born at term after an uncomplicated pregnancy. In the nursery was noted to have tonic extension movements concerning for seizures. Despite therapeutic PB the infant continued to have jitteriness, increased tone, and exaggerated startle.

Was this a seizure ? Why or why not ?

What work-up would you do?

Consult to neurology?



Hyperekplexia



- Rare
- “Startle disease” “Stiff baby syndrome”
- Excessive startle that does not habituate
 - Triggered by nose tapping
- Episodic stiffening, at times leading to apnea
 - Can be aborted with forced flexion of head and legs toward the trunk
- Feeding difficulties
- Developmentally normal
- At risk for frequent falls due to hypertonicity and startle
- AD or sporadic
- Glycine receptor gene mutation (GLRA1)
- Rx: Clonazepam

What is your diagnosis?



Sandifer Syndrome



- Intermittent, unusual postures with GER during infancy
- Arching or turning of trunk
- Extension or sideways bending of neck
- Doesn't tend to involve limbs
- Can last for minutes
- ± apnea
- ± history of vomiting
- ± relationship with meals
- Dx: posturing associated with esophageal pH < 4
- Improves with anti-reflux Rx

Case (videotape)



Healthy infant with abnormal eye movements.

Occurs multiple times per day with no associated head, trunk, or arm movements. No associated alteration of consciousness.



Paroxysmal Tonic Downgaze of Infancy



- Sudden downward eye deviation
- Very brief (secs)
- Can recur multiple times per day
- Triggers: stimulation, movement, feeding, bathing
- Remission by six months in majority
- Normal exam and development
- *Diagnosis of exclusion:* EEG, MRI

Paroxysmal Tonic UpGaze

- Onset: 4 - 10 months
- May start after illness or immunization
- Transient upgaze with associated neck flexion
“eyes up, chin down”
- Downbeating saccades on attempted down gaze
- May last seconds to hours
- Normal consciousness
- Not seen in sleep
- Resolves by 1-2 years
- A channelopathy ? CACNA1A mutation ?

Ozbay Ped Neuro 2012; 47(4): 306-308



Case (videotape)



Parents are very concerned about their six-month old boy with recurrent episodes of head “twitching” and arm trembling, only seen when awake. No apparent alteration of consciousness. Increasing in frequency.



PLAY

Shuddering Attacks

- Usually begin 4-6 months of age
- Arrest of activity
- Stiffening and flexion of trunk and extremities
- Shivering type movements of head and arms
- Brief (seconds)
- Multiple times per day (not seen during sleep)
- No LOC or post-ictal state
- Increase with excitement
- Pathophysiology ??
- Early manifestation of essential tremor ?
- May respond to Inderal

Case (videotape)



Stereotypies



- Usually begin in first few years of life
- Repetitive, stereotyped, purposeless movements
- Not distressing to the child
- Increased during excitement, concentration, preoccupation, boredom, drowsiness
- Do not occur when asleep
- Body rocking, head banging, arm flapping
- Head shaking, head rolling, head to shoulder mvt
- Can be distracted from the activity
- Common in children with MR, autism, blindness
- Pleasurable ?? Relieve tension or boredom??

Consult (video)



6 month old female with hypertonic episodes of increasing frequency over the past 3 weeks. Infant was initially felt to be constipated and straining so was prescribed lactulose. Episodes consist of tensing and crossing legs, flexing hips, grunting, and clasping hands. Episodes last 5 to 30 seconds and occur multiple times per day but do not seem to be bothersome to the patient.



Gratification Disorder



- Masturbatory behavior
- Onset 3 months – 3 years
- Girls >> boys
- Infancy to early elementary school
- Repeated adduction of thighs
- Arm extension
- Grunting, sweating, facial flushing
- Rocking
- Staring
- May be followed by fatigue
- Normal consciousness
- Can be stopped or interrupted



Stool Withholding



- Episodes of leg stiffening
- Staring, moaning, decreased responsiveness
- May c/o stomachache
- Variable age of onset
- Cease with Rx of constipation

Case (videotape)



Six-month-old with 2 week history of “jerking” up to three times per day. Usually when tired. Flexion of neck and trunk with either limb extension or flexion. No postictal state. Normal neuro exam and development.



Case



- Diagnosis: Infantile Spasms
- Evaluation: MRI, EEG, Woods lamp, Labs
- Treatment: ACTH, Prednisone, Vigabatrin, Topiramate
- A neurologic urgency!

Benign Myoclonus of Infancy



- Uncommon
- Onset 3-9 months
- Clusters of myoclonic jerking resembling IS
- Occurs when awake
- Normal EEG
- Normal MRI
- Normal development
- Resolves by 12 months
- *A diagnosis of exclusion!!*

Case (videotape)



Infant admitted to the hospital after several ALTE episodes and possible seizure activity. Continues to experience these episodes in the hospital despite a negative evaluation. Mother is a nurse.



AM 6:08
JAN. 29 1996

Breath-Holding Spells



- Simple BHS in ~ 25% of all children
- Severe BHS in ~ 4%
- Onset: neonate - 18 months
- Rare onset after 2 years
- Typically resolve by 7 years
- Multiple per day to once a year
- Vagal excitation in response to noxious event
 - Trauma, tantrum, emotional upset, frustration
- Family history in 25%
- May precipitate a seizure or status epilepticus

Cyanotic Breath-Holding Spells



Precipitating event (frustration, fear, trauma, anger)



Cry followed by prolonged expiration ('quiet cry')



Cyanosis and apnea



Loss of consciousness



± Posturing and clonic movements



Gradual return of consciousness

Pallid Breath-Holding Spells

Precipitating event (minor trauma, fear)

↓
Minimal to no crying

↓
Limp, unresponsive, and pale

↓
± Posturing or clonic movements

↓
Gradual return of consciousness

- hypersensitive cardioinhibitory reflex with resultant asystole

Breath-Holding Spells



- CYANOTIC (60%)
 - PALLID (20%)
 - MIXED (20%)
 - SIMPLE - color change
 - SEVERE - LOC and change in tone, \pm seizure
-
- No sequelae except increased risk of syncope
 - Look for underlying iron deficiency (low ferritin)
 - EKG to r/o prolonged QTc
 - Rx: FeSO₄ and reassurance
 - Severe cases: piracetam, clonidine, fluoxetine, tetrabenazine, scopolamine, atropine, pacemaker

Case (Video)



2 1/2 year old girl with recurrent episodes of coarse nystagmus associated with distress, poor head control, imbalance, inability to ambulate, and frightened appearance. Mental status appears to be normal during the “spells” as she is still able to communicate. Episodes resolve within one minute with return to full activity. Normal exam.



Benign Paroxysmal Vertigo (BPV)



- Recurrent attacks of vertigo without provocation
- Duration: seconds to minutes
- Toddlers and young children (onset usually 1 - 3 years)
- Sudden onset, unprovoked
- Loss of balance, unable to walk, may cling to parent or fall
- Distressed, frightened
- Pallor, nystagmus, nausea, emesis
- No alteration of consciousness
- No postictal state
- Normal between attacks
- Migraine equivalent (CACNA1A gene mutation ??)
- Resolves by 6 years in most

Consult (Video)



Monthly spells

- Unprovoked
- Head tilt and unsteadiness
- Normal consciousness

Benign

VIDEO CALIBRATION

Paroxysmal

Torticollis

Benign Paroxysmal Torticollis



- Recurrent episodes of torticollis
- Associated pallor, emesis, ataxia, irritability
- Duration: days
- No altered consciousness
- No precipitating factor
- Onset: 6-12 months of life
- *Diagnosis of exclusion: Nor*, EEG, audiology
- Resolve by 3 years
- Migraine equivalent (CACNA1A gene mutation?)

Rage Attacks / Episodic Dyscontrol



- May begin in childhood or adolescence
- Episodic, severe, directed aggression
- Out of proportion to preceding trivial provocation
- Physically or verbally abusive
- Inconsolable, often require restraint
- Duration: minutes - hour
- May have change in awareness
- Exhausted and quiet afterwards
- May be amnestic for event
- Rx: Psychology consult, Tegretol, VPA, SSRI's

Daydreaming



- Staring and decreased responsiveness
- Occur in low-stimulation environments (e.g. school)
- Does not interrupt an activity
- Resolves with touch or tickle vs voice
- No associated automatisms
- No post-ictal state



Staring Spells



- Frequency?
- When and where are they seen?
- Does it interrupt an activity?
- Focal head or eye movements?
- Automatisms?
- Post-ictal?
- What stimulus was used to abort?
- Reproduced in office with HV?

	Absence seizures	Complex partial	Non-epileptic staring
Age	3-8	Any	3-100
Frequency	Multiple/day	Infrequent	Infrequent
Duration	< 30 sec	Sec - minutes	Variable
Situation	Random, interrupts activity	Random, interrupts activity	School, tired, bored, TV, computer
Automatisms	+/-	++	--
Postictal	never	often	Never
Abort with touch	no	no	Yes
HV	provoked	--	--

Case (Videotape)



Consult (Video)



- 5 year old with several months of abnormal movements
- Unaware that this is happening
- Not distressing
- No alteration of consciousness



Tics



- Sudden
- Brief, intermittent
- Recurrent
- Non-rhythmic
- Stereotyped
- INVOLUNTARY
- Movement or vocalization
- Usually starts with motor tic(s)
- Child is usually unaware of them

Tics



- 4 – 25 % of children
- M:F (3:1)
- Onset usually between 3 and 8 years
- 96 % manifest symptoms by 11 years
- Eye blinking most common
- Phonic tics usually emerge 1-2 yrs after motor tics
- Most severe: 10 – 15 years ?
- Improve late adolescence or early adulthood

Consult



10 1/2 year old girl felt funny while getting her hair brushed after coming out of the shower. Mother noted that she didn't look right and caught her before she fell. While mother helped her to the next room to lay down, she developed generalized rhythmic shaking lasting for a minute. Normal exam. Remembers feeling warm and "dizzy".

Syncope



- Transient LOC and decreased tone due to decreased cerebral perfusion
- Provoking factors: prolonged standing, heat, pain, prolonged recumbency, pregnancy, emotional stress, injection, change in posture, trauma, hair grooming, urination, stretching, cold liquids, coughing
- **PRESYNCOPE**: lightheaded, dizzy, blurred vision, “greying” of vision, tinnitus, parasthesia, diaphoresis, pallor, nausea, muffled hearing
- **SYNCOPE**: loss of tone, eyes roll, usually brief, minimal trauma, PALLOR, rapid return of consciousness usu. within 1 minute

Syncope



- Most common: neurocardiogenic syncope (vasovagal)
 - peripheral vasodilation with poor venous return
 - increased cardiac contractility with poor filling
 - increased vagal output and bradycardia
- **CONVULSIVE SYNCOPES**: generalized tonic or clonic seizure after syncope (anoxic seizure)
- More likely if head elevated by caretaker
- Post-ictal afterwards
- **Not** indicative of predisposition for epilepsy

Syncope



- Check EKG
- Tx: recognize and respond to pre-syncope, avoid triggers
- Rx: Fluids, salt, Florinef, pseudoephedrine, beta blocker, midodrine
- WARNING SXS: (r/o cardiac disease !)
 - recumbent syncope
 - palpitations or chest pain
 - exertional syncope
 - child < 8 years old
 - familial syncope, sudden death or deafness
 - Trigger: loud noise, fright
 - Absence of pre-syncopal symptoms

Consult



16 year old girl with acute onset of disorientation (time and place) and amnesia. Didn't know names of friends, what classes to attend, how to get to cafeteria. Seemed inappropriate and confused to her classmates. Normal neuro status otherwise. Mild headache earlier that morning. In ER, normal chemistry, HCG, drug screen. No seizure history. On Wellbutrin for ADHD.

Complicated Migraines



Acute Confusional Migraine

- Onset in late childhood or adolescence
- Abrupt onset of confusion and disorientation
- Trouble forming new memories
- May not have associated headache
- Duration < 12 hours
- Usually terminates in sleep
- May be recurrent in 25%
- May follow trivial head trauma
- Often positive history of migraine
- Typical migraine headaches eventually develop

Other Complicated Migraines...



Migraine with Brainstem Sxs (Basilar Migraine)

- Onset in adolescence or late childhood
- Females > males
- Brainstem sxs: vertigo, diplopia, ataxia, tinnitus
- \pm Loss of consciousness
- Fall to ground
- Followed by an occipital HA
- MRI to rule out posterior fossa lesion

Acephalic Migraine



- Aura without headache
- Predominantly visual auras
 - scotomas
 - colorful photopsias
 - micropsia
 - Dizziness / vertigo ?
- Episodic and stereotyped
- Evolves over minutes (seconds for seizures)
- Duration < 60 minutes (usually < 10 minutes)
- Strong history of HA or family history of migraine
- Normal EEG and imaging

Seizures?



Psychogenic Paroxysmal Non-Epileptic Events

- aka ‘pseudoseizures’
- 5 - 20% of referrals for intractable epilepsy
- “Spells” that imitate true epileptic seizures
- Adolescents and adults females >> males
- Often significant psychosocial stressors (abuse, family discord, depression, drug abuse, school)
- Often occur in presence of others during stress
- No tongue biting or incontinence
- Prompt recovery
- May be induced by suggestion
- May also have true seizures !

Pseudoseizures



- Unresponsiveness or staring (younger patients)
- Kicking and thrashing movements
- Out-of-phase limb movements
- Side-to-side head movements
- Pelvic thrusting
- Awareness despite generalized convulsive activity
- Uncommon to be injured
- Atypical exam:
 - noxious stimulation (corneal, tickling, sternal rub...)
 - avoidance maneuvers (lift arm over face and release)
 - Babinski
 - passive eye opening
- Normal prolactin ??

When to suspect PNE's...



- Frequent episodes despite Rx
- Atypical clinical features
- Multiple normal EEG's
- Tend not to occur during sleep
- Often with audience
- “Florid” positive ROS
- Swoon-type events lasting more than 1 minute

Case (Videotape)



22 year-old on FP service with new onset
Seizures triggered by head turning. No
improvement with anticonvulsant therapy.



Case



8 year old girl with 5 month history of excessive sleepiness. Suddenly falls asleep in school, during karate class, watching TV, riding in car. Sleeps well at night. No snoring. Episodes increasing in frequency. Normal TFT's, CT, EEG, monospot, CBC, Hep/renal panel. Air evac from Korea : r/o seizures and post-ictal drowsiness.

Narcolepsy

- Intrusion of REM sleep onto wakefulness
 - Irresistible and excessive daytime sleepiness, esp. when sedentary (e.g classroom, watching TV, driving)
 - HLA DR2 and HLA DQB1*0602
 - Hypocretin disorder
 - Lifelong
-
- NARCOLEPSY (Rx Modafanil, naps)
 - CATAPLEXY
 - SLEEP PARALYSIS
 - HYPNAGOGIC HALLUCINATIONS



Cataplexy



- Abrupt loss of tone
 - Droop of eyelids, relaxing of mouth, head drop, fall to ground
- Provoked by emotion: laughter, fright
- Preserved consciousness
- Rx: sodium oxybate, clomipramine, venlafaxine

Panic Attacks



- Adolescents: females > males
- Parasthesias of face and extremities
- Dizzy, lightheaded
- \pm LOC
- SOB, sensation of choking
- Chest pain / palpitations
- Sense of anxiety or panic
- Psychological disturbances or chronic physical sxs
- DX: voluntary HV 2-3 minutes: reproduce sxs
- Rx: relaxation, reassurance, propranolol ?

“Hyper-vigilance”



- Over interpretation of benign or non-specific symptoms and events
 - Dizziness, numbness, head sensations, brief involuntary movements
- Anxious parents
- \pm ICU setting
- Ask for home video
- May require video EEG

Case (videotape)



New onset seizures while watching TV. Noted to fall to the ground and violently shake all over. Upon further history, all episodes have occurred while watching Entertainment Tonight with Mary Hart.





“Delay in diagnosing epilepsy is far less serious than diagnosing it erroneously!”

M. Prasad

Arch Dis Child Educ Pract Ed 2016



Cases

Case 1 (videotape)



Parents are very concerned about their six-month old boy with recurrent episodes of head “twitching.” Only seen when awake. No apparent alteration of consciousness. Increasing in frequency.



Case 1



- Diagnosis: Stereotypy (Head Shaking)
- Evaluation: None
- Treatment: Reassurance

Case 2 (videotape)



8 1/2 month old girl with 2 week history of “spells” occurring multiple times per day. Sudden arrest of activity with flexion and stiffening of extremities and trunk, grimacing, and quick trembling movements. Last For several seconds. Only seen while awake.
Family history of epilepsy.



Case 2



- Diagnosis: Shuddering Attacks
- Evaluation: None
- Treatment: Reassurance
Increased risk of essential tremor?

Case 3 (Videotape)



10 year-old developmentally normal girl who shakes her arms and flaps her hands multiple times per day. Sometimes accompanied by exaggerated mouth opening. More common when excited or nervous (e.g. playing volleyball). No alteration of consciousness and it is preceded by a ‘tingling’ sensation. She can suppress the movement briefly but prefers not to. Has been noted since she was two years old.



Case 3



- Diagnosis: **Stereotypy**
- Evaluation: None
- Treatment: Reassurance

Case 4 (videotape)



Young girl is reported to have recurrent “spells” at night or during naps. Unresponsive to her parents and not consolable. Falls back asleep after several minutes. No memory of events.
Please r/o nocturnal seizure disorder



Case 4



- Diagnosis: Night terrors
- Evaluation: None
- Treatment: Reassurance

Benzodiazapines if severe

Case 5 (videotape)



6 1/2 year old boy with rhythmic head banging at the onset of sleep since infancy. Increasing in frequency and severity as he has gotten older. No unusual postures or noises. Present only in the prone position. Stops when he is aroused. He is aware of the movements as he is going to sleep. No history of apnea or snoring. Twin sister unaffected.



Case 5



- Diagnosis: Rhythmic movement disorder
(Jactatio Capitis Nocturna)
- Evaluation: None
- Treatment: Reassurance
Benzodiazepines or imipramine ??

Case 6 (Video)



- 5 yr old from Palau
- 2 months of unusual eye movements
- No automatisms or limb movements
- No altered consciousness
- Normal head CT



Case 6



- Diagnosis: Simple motor tics
- Evaluation: none
- Treatment: reassurance

Case 7 (videotape)



3 year old with history of febrile seizures and ? afebrile seizures. Several month history of jerking while falling asleep, lasting up to 30 minutes. Distressing and interfering with sleep. Increasing in frequency and severity. Normal exam and MRI in the past.



Case 7



- Diagnosis: Sleep myoclonus
- Evaluation: EEG during sleep onset?
- Treatment: Reassurance

Additional information regarding
sleep associated ‘spells’...



Sleep-related spells



Sleep-wake transition disorder

- Transitioning to sleep ?
- Awakening from sleep ?

Arousal disorder

- Several hours into sleep ?
- Awakened from sleep ?

Videotape



7 month old with several episodes of “shaking” of arms.
Most recent episode videotaped by parents and
reviewed by pediatrician, who sent them from
Yokosuka, Japan for evaluation of “movements
concerning for seizures.”

Was this a seizure ? Why or why not ?

What work-up would you do?

Consult to neurology?



AUG 20 99

Sleep Myoclonus (Hypnic Jerks)



- Sleep-wake transition “disorder”
- Sudden, random, brief jerks of limbs
- Subjective perception of falling
- Occur at sleep onset
- Neonates through adulthood
- In children, may be prolonged and semi-rhythmic
- Increase with prior physical activity, stress, caffeine, or sleep deprivation
- May be provoked in neonate with head to toe crib rocking
- Stops with arousal

Videotape



Mother awoke to a noise and noted this movement on the baby monitor while her toddler was in his crib.

Was this a seizure ? Why or why not ?

What work-up would you do?

Consult to neurology?



Rhythmic Movement Disorders of Sleep



- Sleep-wake transition disorders
- Stereotyped, rhythmic movements before sleep onset
- May continue into light sleep
- Head banging (Jactatio Capitis Nocturna)
- Head rolling, leg rolling
- Body rocking (hands and knees) or rolling
- Last minutes, rarely hours
- Stops with arousal, distraction, or deep sleep
- Onset: 1st year of life resolve spontaneously by 5 yr
- More prevalent in children with MR, autism, delays

“Binkie Flutter”



- Rapid, fine tremor of pacifier
- During pause from vigorous sucking
- Puckering of lips
- Peaceful, motionless, eyes closed

Sleep Paralysis



- At sleep onset (hypnagogic) or awakening (hypnopompic) from sleep
- Paralysis (except EOM's and diaphragm)
- Unable to speak
- Aware of event and frightened
- \pm sensation of inability to breathe
- Terminates with touch or noise
- Isolated or associated with narcolepsy
- Intrusion of REM atonia in awake state

Arousal Disorders



- Partial arousal from SWS
- First half of sleep
- Inc. with stress, sleep deprivation, fever



Pavor Nocturnus (Night Terrors)

- Impaired arousal from stage III/IV non-REM sleep
- Incidence: 1-3 %
- Onset: 4 - 12 years
- Usually 1 - 2 hours after falling asleep
- May be precipitated by illness, sleep deprivation, stress
- Suddenly sit upright, eyes open
- Inconsolable screaming
- Dilated pupils, tachycardia, tachypnea, sweating
- Fall back asleep, amnesia for event
- Duration: up to 30 minutes
- Benign, outgrown
- Rx: reassurance; benzodiazepines, imipramine

Somnambulism (Sleepwalking)

- Onset in childhood, usually terminates by adolescence
- Incomplete arousal from stage III/IV sleep
- Usually 1-2 hours after falling asleep
- Walking with eyes open, blank expression
- Slow or absent responsiveness
- Usually less than 10 min, followed by return to sleep
- No memory of event
- Family predisposition
- May coexist with night terrors
- Rx: safety precautions; benzodiazepines





Confusional Arousals



- Provoked by sudden or forced arousal
- Usually awakened from deep non-REM sleep
- Disoriented
- Slow speech and mentation
- Poor memory
- May display some aggression
- Duration: minutes
- Attempts to “wake them up” often unsuccessful
- No recollection of event

Periodic Limb Mvts of Sleep

Restless Legs Syndrome



- Urge to move the legs
- Accompanied or caused by unpleasant sensn in legs
- Worse during rest or inactivity
- Worse at night
- Improved with movement
- Insomnia
- Daytime drowsiness
- Autosomal dominant
- Assoc with ADHD and iron deficiency
- Rx dopamine agonist, iron, clonazepam