NOT EVERYTHING THAT SHAKES IS A SEIZURE: Non-Epileptic Movements

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As a result of attending this lecture at the AZ AAP course, attendees will make the following change in their practice:

1. Recognize disorders frequently mistaken as seizures
2. Become very comfortable with management of tics
3. Perform a basic evaluation for dizziness before referring a child to a specialist
A 2-year-old girl presents to the emergency department 10 minutes after the sudden onset of ataxia and refusal to walk. She has vomited twice. According to her parents, she experienced similar episodes three times in the past 6 months. On a previous evaluation, results of oto-acoustic emission testing and computed tomography of the head were normal.
QUESTION 1

Of the following, the MOST likely diagnosis is

A. benign paroxysmal vertigo
B. complex partial epilepsy
C. medulloblastoma
D. Ménière disease
E. perilymphatic fistula
Answer: A, benign paroxysmal vertigo

- Abrupt, brief episodes of vertigo with ataxia in children ages 2-6
- Child appears frightened, with pallor, and may indicate feeling dizzy
- Rapid eye movements or nystagmus may be observed if the eyes are open.
- Vomiting may be prominent
- No loss of consciousness
- Migraine variant
- Treatment is supportive
Your diagnosis?

Sandifer syndrome - episodic extension and lateral flexion of the head that occurs 4 months - 14 years, usually in association with feeding
PAROXYSMAL EVENTS

Not Everything that Shakes is a Seizure!

- Neonatal apnea
- Breath-holding spells
- Syncope
- Dizziness
- Vertigo
- Migraine
- Acute confusional state
- Nightmares (REM)
- Night terrors (nREM)
- Somnambulism (nREM)
- Narcolepsy
- Cataplexy
- Tic
- Dystonia
- Myoclonus

- Shuddering attacks
- Hyperekplexia
- Spasmus nutans
- Daydreaming/attentional disorder
- Hyperventilation
- Prolonged QT syndrome
- Wolff-Parkinson-White syndrome
- Brugada syndrome
- Hypertrophic cardiomyopathy
- Sandifer syndrome
- Transient ischemic attack
- Depression/fugue state
- Panic attack
- Paroxysmal behavior outburst
- Masturbation
- Masturbation
- Pseudoseizure
PAROXYSMAL EVENTS

Evaluation

- Complete history and physical exam
- Hyperventilation
- Electrolytes (with Ca$^{+2}$, Mg$^{+}$), BUN, liver function tests, CBC
- Toxicology screen
- Pregnancy test
- Head CT acutely; MRI is the preferred modality for nonurgent neuroimaging (Neurology 2000;55:616)
- EKG
- pH probe study?
- EEG, with hyperventilation and photic stimulation, after first seizure (Neurology 2000;55:616)
- Video-EEG monitoring
PAROXYSMAL EVENTS

Pearls

• For episodes that occur frequently, smart phone video is an excellent, cost-effective, first step.

• Eyes are open, not closed, during a seizure. Consider instead a psychogenic (i.e., non-electrical or behavioral) seizure.

• Patients with new-onset, daily seizures rarely have completely normal interictal EEGs.

• Seizures rarely produce “negative phenomena,” such as pallor, cold, apnea, and bradycardia. Consider instead syncope, breath holding spells, or prematurity.

• Directed acts of violence are not a feature of epilepsy. Consider instead rage attacks or other behavioral disorders.

• Conversion disorder and malingering, which can manifest as psychogenic seizures, are uncommon in the first decade.
• Onset 1-2 years of age, extinguish by 6
• Apnea with cyanosis, leading to limpness and then alteration of consciousness, often unresponsiveness, typically provoked by crying
• Pallid breath-holding spells (syncope) may be reflex asystole, provoked by a sudden event
• Gaze may deviate superiorly
• Last no longer than 1 minute
• Rapid recovery and arousal
• Normal EEG
• Family history in 1/4
BREATH HOLDING SPELLS
Management

- 20% children experience at least one episode of syncope in later life
- **Iron therapy** (5 mg/kg ferrous sulfate) appears beneficial in more than half of children with cyanotic spells, particularly those with iron deficiency anemia (*Arch Dis Child* 2002;86:77, *J Pediatr* 1997;130:547)
- Pacing in pallid spells?
“I’M DIZZY”

**Syncope vs. Dizziness vs. Vertigo**

- **Syncope** - sudden, reversible loss of consciousness due to failure to meet brain energy needs. Synonymous with fainting. Vasovagal reflex, may be stimulated by Valsalva, neck hyperextension.
- **Dizziness**
  - sensation of impending faint or loss of consciousness
  - dysequilibrium or loss of balance without head sensation
  - light-headedness
- **Vertigo**
  - illusion of motion
  - definite rotational sensation
- **Out of body, floating, spinning inside**
  - psychophysiological
DIZZINESS
Differential Diagnosis - Dizziness

- Hypotension
- Arrhythmia
- Anemia
- Vasovagal spell/paroxysmal orthostatic tachycardia syndrome (POTS)/?chronic fatigue syndrome (CFS)
- Hypoglycemia
- Hyperventilation
- Seizure
- Toxin - antiepileptic drugs, sedatives, ethanol, tobacco, tricyclic antidepressants, vestibular suppressants
- Mal de debarquement (perception of rocking as on boat)
- Psychiatric disorder
DIZZINESS

Differential Diagnosis - Vertigo

- Benign paroxysmal positional vertigo (BPPV) - seconds
- Benign paroxysmal vertigo/torticollis (migraine variant) - minutes to hours
- Basilar migraine - hours
- Vestibular neuronitis/acute labyrinthitis – days to weeks
- Herpes zoster - days
- Otitis media - days
- Post-traumatic vestibulopathy/perilymphatic fistula - days to weeks
- Ménière’s disease, endolymphatic hydrops - minutes to hours
- Cerebellopontine angle tumor - rare
- Cerebellar hemorrhage - abrupt onset
- Toxin - aminoglycosides, furosemide, cisplatin, ethanol, salicylates, isoniazid, phenytoin
- Psychiatric disorder
DIZZINESS

Evaluation I

• Complete history and physical exam. If true vertigo...
  – environment seems to move away from lesion
  – spontaneous horizontal nystagmus
    • fast phase away from side of the lesion
    • fast phase unchanged with direction of gaze
    • increases in intensity when gaze is in the direction of the fast phase
    • increases in intensity with removal of visual fixation
    • vertical nystagmus indicates a central lesion

• Romberg sign - fall toward lesion
• Pastpointing - toward side of lesion
• Head thrust test – patient makes corrective saccade to fixation target
BPPV is characterized by episodes of vertigo, lasting 5-30 seconds and provoked by changes in head position with respect to gravity.

The Nylen-Bárany maneuver for positional vertigo and nystagmus. The patient is moved abruptly from a seated [A] to a prone [B] position, with his head hanging 45° below the horizontal and rotated 45° to one side. He is observed for the development of nystagmus and vertigo.

The test is positive when the child develops vertical-torsional nystagmus to the downward ear.

- **Bárany maneuver**
DIZZINESS
Evaluation II

- Calorics
- Orthostatics
- Hyperventilation
- Audiometry
- EKG
- EEG
- MRI
- Tilt-table testing
DIZZINESS

Treatment

• Treat underlying cause
• Avoid provocative situations
• Syncope: beta blockers, phenylephrine (alpha-adrenergic agonist)
• POTS: fluids, salts, fludrocortisone 0.1 mg QD
• Menière’s/endolymphatic hydrops: eliminate caffeine, chocolate, alcohol; <2 g salt QD?; start acetazolamide
• Vestibular suppressants (e.g., meclizine, diazepam, dimenhydramine, diphenhydramine) beneficial only in acute vestibulopathy
• Mal de debarquement: benzodiazepines, exercise
SLEEP DISORDERS

Types

- Parasomnias - night terrors (nREM), somnambulism (nREM), nightmares (REM)
- Hypnagogic (nocturnal) myoclonus
- Insomnia
- Delayed-sleep-phase syndrome (DSPS) - circadian clock has free-running rate >24 hr
- Sleep-disordered breathing
- Narcolepsy
SLEEP DISORDERS

Night Terrors (Pavor Nocturnus)

- Onset age 4 to 6 years
- Partial arousal during nREM sleep
- Child sits up suddenly in bed, appears awake and agitated, may scream or struggle
- Sympathetic discharge is evident: diaphoresis, tachycardia, tachypnea, and mydriasis
- Usually terminates in a brief period of wakefulness with amnesia of the event and immediate return to sleep
- Lasts 5 to 15 minutes
- Responds to low-dose diazepam?
- May co-exist with sleepwalking (somnambulism)
SLEEP DISORDERS

Sleep-Disordered Breathing

• Obstructive sleep apnea and upper airway resistance syndrome

• Symptoms: excessive daytime sleepiness, hyperactivity, pathologic shyness, learning problems, morning headache, recurrent upper airway infection, enuresis, repetitive nightmares, failure to thrive/obesity

• Treatment: tonsillectomy and adenoidectomy, orthodontic treatment, nasal CPAP
SLEEP DISORDERS

Narcolepsy

• Mean age onset 10 years
• Multifactorial mode of inheritance
• Excessive daytime sleepiness (EDS)
• **Cataplexy** - sudden loss of tone triggered by emotion; may enter REM sleep, 30-120 seconds. The motor inhibition is similar to that of normal individuals during REM sleep
• Hypnagogic hallucinations
• Sleep paralysis
• “Symptomatic narcolepsy” can be seen with diencephalic tumors and Niemann-Pick type C disease
SLEEP DISORDERS

Management

• DSPS - chronotherapy, light therapy
• Synthetic (not bovine) melatonin 0.3-3.0 mg may have direct hypnotic effects
• Sedative-hypnotics can be counterproductive in children with significant brain insults and DSPS
• EDS - methylphenidate 5 mg 3-4/day, modafinil 100-200 mg may help
• Cataplexy - clomipramine, fluoxetine
• Referral for specialty consultation
TICS

Diagnosis

• Typical onset age 6 to 7 years
• Simple or complex stereotyped movements (motor) or utterances (vocal)
• Simple tics can include eye blinking, grimacing, lip smacking
• Vocal tics can include snorting, coughing; coprolalia rare
• Can be suppressed briefly by voluntary effort
• Less frequent when children are out of school
• Effort is made to incorporate tics into voluntary movement
• Tourette’s syndrome defined by motor and vocal tics for over 3 months; may be associated with attention deficit disorder (ADHD) and obsessive-compulsive disorder (OCD)
• Most children outgrow their tics
TICS

Treatment

• Most children do not require treatment
• Initiate treatment only when tics are stigmatizing or interfering with daily activity
• Common medications: α-adrenergic agonists (e.g., clonidine 0.1 mg PO divided bid or patch; guanfacine); pimozide; haloperidol; risperidone; fluphenazine
• Tic reduction may not occur for up to 6 weeks
• Maybe habit reversal therapy, don’t know deep brain stimulation
• Often the accompanying ADHD or OCD merit closer scrutiny and treatment

TICS

PANDAS and CANS

• Is it real? Rare at best

• Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infection
  – Presence of OCD or a tic disorder
  – Abrupt onset between age 3 and puberty
  – Temporal relationship to Group A β-hemolytic streptococcus
  – Abnormal exam (i.e., hyperactivity, chorea, tics) during an exacerbation

• Insufficient evidence to support this hypothesis
• Neither routine testing for GABHS nor antibiotics, IVIG, or plasmapheresis warranted
• Is it Childhood Acute Neuropsychiatric Symptoms?
OTHER PAROXYSMAL EVENTS

- Neonatal sleep myoclonus
- Hyperekplexia (startle disease) - exaggerated startle in response to tactile, auditory, or visual stimuli; considered autosomal dominant; rare; normal EEG
- Shuddering attacks - 2 months to 11 years, shivering movements lasting 5-10 seconds, with flexion of the head, elbows, trunk and knees; appears as if “water were being poured down their backs”
- Spasmus nutans - triad of head nodding, torticollis, and nystagmus, with onset at end of first year and remission by age 2-3 years
A mother brings her 10-month-old son to the emergency department because he has been vomiting for the past 10 days. The child has not experienced any diarrhea. On physical examination, he is lethargic and has dry mucous membranes, reduced tears, a full anterior fontanelle, and 2-second capillary refill. After a second intravenous bolus of 20 mL/kg of normal saline, the boy extends his arms and legs forcefully for 10 seconds.
QUESTION 2

Of the following, the MOST appropriate next step in the management of this child is administration of

A. additional intravenous normal saline bolus of 20 mL/kg
B. intravenous dexamethasone of 1 mg/kg
C. intravenous fosphenytoin bolus at 20 mg/kg phenytoin equivalents over 10 minutes
D. intravenous prochlorperazine of 5 mg
E. rapid intravenous lorazepam of 0.05 mg/kg
Answer: B, intravenous dexamethasone of 1 mg/kg

• Beware of vomiting without diarrhea; don’t forget to consider an intracranial process
• Child is posturing, having “cerebellar fits,” (not a true seizure) because of increased intracranial pressure
• Antiemetics will simply mask the process
• Vigorous hydration will worsen symptoms
• Dexamethasone will decrease vasogenic edema from a mass lesion
Child likely has brain tumor

The movements are likely “cerebellar fits,” not seizures

Medulloblastoma, on coronal T-1 weighted magnetic resonance imaging with gadolinium
QUESTION 3
A 4-year-old girl is brought to the emergency department by her babysitter because the child has suddenly become clumsy, and her speech has become slurred over the last hour. On physical examination, the girl is afebrile and dysarthric. She has prominent vertical and horizontal nystagmus, along with truncal and appendicular ataxia. Deep tendon reflexes are normal, as are results of the remainder of the physical examination.
QUESTION 3

Of the following, the MOST likely diagnosis is

A. brain tumor
B. cerebellar hemorrhage
C. Guillain-Barré syndrome
D. meningoencephalitis
E. toxic ingestion
Answer: E, toxic ingestion

- Vertical plus horizontal nystagmus is typical of phenytoin ingestion
- Vertical nystagmus can be seen with central processes, such as hemorrhage or Chiari malformation, but also is suggestive of phencyclidine, lithium, and phenytoin poisoning
- Lack of fever and obtundation make infection and hemorrhage, respectively, unlikely
- Deep tendon reflexes are diminished to absent in Guillain-Barré syndrome
A mother brings her 12-month-old boy to you because he holds his head tilted to the right. She tells you that he periodically draws up both his legs and cries. The child has developed normally, but does not yet walk or cruise. On physical examination, he has mildly increased deep tendon reflexes in the upper and lower extremities, but other findings are normal.
QUESTION 4
Of the following, the study MOST likely to establish this boy’s diagnosis is

A. audiometry
B. electroencephalography
C. magnetic resonance imaging of the brain and cervical spine
D. pH probe of the distal esophagus
E. radiographs of the cervical spine
Answer: C, magnetic resonance imaging of the brain and cervical spine

- Periodic withdrawal of legs could represent reflux, infantile spasms, or cervical spine pain
- Increased deep tendon reflexes → upper motor neuron sign
- Brain and cervical spine MRI is indicated
Differential diagnosis of head tilt

- Muscular torticollis
- Posterior fossa/cervical spine tumor
- Syringomyelia
- Dystonia
- Visual disturbance (IV > VI, III nerve palsy)
- Hearing loss
- Croup and epiglottitis

- Hemivertebrae
- Klippel-Feil syndrome
- Atlantoaxial rotary subluxation
- Lymphadenitis
- Sandifer syndrome (gastroesophageal reflux)
- Paroxysmal torticollis
- Spasmus nutans
QUESTION 5

A father brings his 2-year-old daughter to you because he has noticed “funny eye movements” over the past 2 weeks. Upon physical examination, you find that the child has pendular nystagmus, her head is tilted to the right, and she nods her head. Other findings on the physical examination are normal.
QUESTION 5

Of the following, the MOST likely diagnosis is

A. congenital nystagmus
B. optic glioma
C. phenytoin intoxication
D. retinoblastoma
E. spasmus nutans
Answer: E, spasmus nutans

- **Nystagmus** = involuntary, rhythmic oscillation of both eyes (rarely one) in which at least one phase is slow. Nystagmus reflects a deficit in gaze-holding mechanisms.
- **Jerk nystagmus** = slow phase in one direction, usually back to central position + fast, compensatory movement in the opposite direction.
  - Think drug toxicity, vestibular dysfunction, tumor, infarction, Chiari malformation, or normal if not sustained.
- **Pendular nystagmus** = slow movements in both phases, with the eyes moving “to and fro”.
  - Brainstem infarction, spinocerebellar degeneration, multiple sclerosis, or rarely seizure.
- **Spasmus nutans** = pendular nystagmus + intermittent head tilt + nodding.
A 3-year-old girl presents to the emergency department with the sudden onset of weakness of the left arm and leg. The babysitter who brought her to the hospital knows only that the child is followed by an ophthalmologist for myopia and some other eye problems. Physical examination reveals a tall, thin child who has a fair complexion, blue eyes, a malar rash, and long, slender fingers. Neurologic examination reveals 2/5 strength in the left upper extremity and 3/5 in the left lower extremity.
QUESTION 6

Of the following, the laboratory test MOST likely to establish this child’s underlying diagnosis is

A. collagen studies from skin fibroblasts
B. genetic testing for factor V Leiden
C. hemoglobin electrophoresis
D. lupus anticoagulant serum assay
E. urine homocystine level
Answer: E, urine homocystine level

• Sudden neurologic events
  – Stroke
  – Seizure
  – Complicated migraine

• Stroke pathophysiology: sudden interruption of blood flow in the central nervous system from thrombosis, embolism, or hemorrhage because of
  – Cardiac disease, e.g., congenital heart disease, endocarditis, cardiomyopathy
  – Vasculopathy, e.g., carotid dissection, aneurysm, arteriovenous malformation
  – Hematologic disease, e.g., sickle cell disease; lupus anticoagulant; antiphospholipid antibody syndrome; procoagulable states: deficiency of factor V Leiden, protein S or C, or antithrombin III;
  – Metabolic disorder, e.g., mitochondrial disorders, Fabry disease, homocystinuria

• The Marfanlike body habitus, fair complexion, myopia, blue eyes, malar rash, and abrupt hemiparesis suggest the possibility of homocystinuria
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