Myoclonus

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What is it?

• A twitch, a jerk, a seizure, a shake
• “A brief shock-like involuntary muscle jerk caused by neuronal discharges”
  – A single muscle discharge, but can be repetitive
  – Types:
    • Action
    • Positive
    • Negative
    • Spontaneous

What could it be?

• Tics
  – Brief, stereotyped movements
  – Can be suppressed!
  – Rarely disrupt motor tasks
  – Somesthetic sensation/aura
  – Persist during sleep
• Chorea
  – Fragmentary and flowing movements
• Postural/Action Tremor
• Dystonia
  – Slower, sustained, twisting postures

Classification

• Clinical
  – Presentation
    • Spontaneous
    • Action
    • Reflex
  – Distribution
    • Generalized
    • Multifocal
    • Segmental
    • Focal
• Neurophysiologic Origin
  • Cortical
  • Subcortical (brainstem)
  • Spinal
• Etiology
  – Physiological
  – Essential
  – Symptomatic
  – Associated with epilepsy
  – Other causes
Types of Myoclonus

- Reflex (Stimulus Induced) Myoclonus
  - Triggered by somesthetic, visual, or auditory stim
  - May become self perpetuated or even look like a tremor
  - Have to distinguish from hyperekplexia

- Action
  - Occurs during active muscular contraction
  - Can be focal or segmental or generalized
  - Abnormality probably lies within same pathways that control normal motor control mechanisms
  - Can be very disabling

- Negative
  - More freq. in hospital setting
  - Difficult to treat
  - Brief loss of muscle tone in agonist mm followed by a compensatory jerk of antagonist muscles
    - Asterixis, Postural lapses with postanoxic myoclonus or stiff person syndrome

- Positive
  - More common in outpatient setting
  - Responds better to treatment
  - Rapid, active contractions of muscles

Types of myoclonus

- Cortical (or epileptic) myoclonus
- Subcortical myoclonus
  - Thalamic
  - Startle
  - Palatal
- Spinal myoclonus
  - Segmental
  - Propriospinal
- Peripheral

Where does it come from?

Neurophysiological Origin

- Cortical (or epileptic) myoclonus
- Subcortical myoclonus
  - Thalamic
  - Startle
  - Palatal
- Spinal myoclonus
  - Segmental
  - Propriospinal
- Peripheral
Cortical Myoclonus

- **Cause:**
  - Abnormal activity arising in sensorimotor cortex and spreading down via the corticospinal pathway
  - Considered a focal (or partial) epilepsy
  - EEG: focal positive-negative event over the sensorimotor cortex contralateral to the jerk

Cortical Myoclonus

- **Primary generalized epileptic myoclonus**
  - Fragment of generalized epilepsy
  - Clinical presentations:
    - 1. Small focal jerks, often of the fingers
    - 2. Brief full body jerks (appear similar to reticular reflex myoclonus)
  - EEG: See a slow bilaterally frontocentrally predominant negativity similar to that of a primary generalized discharge

Subcortical Myoclonus

- **Cause:**
  - Neuronal discharge originates in structures between cortex and spinal cord
  - Example is “Reticular Reflex Myoclonus”
    - Generalized jerks induced by sensory stim
    - Can appear very similar to “cortical myoclonus” as can be seen in generalized epilepsies

Subcortical Myoclonus

- **Palatal Myoclonus (tremor)**
  - Essential Palatal Tremor
    - Activation of tensor veli palatini
  - Symptomatic
    - Cerebellar lesions or Mollaret Triangle
    - Levator veli palatini
  - Can also see activation of adjacent muscles
Spinal Myoclonus

• Cause:
  – Originates from abnormal discharges within the spinal cord
• Spinal Segmental
  – Frequently rhythmic rather than stim induced
  – Usually restricted to a few segments of the spinal cord (cervical, thoracic)
• Propriospinal
  – Spontaneous or stim induced
  – Trunk and abdominal muscles

Peripheral Myoclonus

• Some disagree on this definition
  – ? Fasiculation
  – ? Myokymia
• See signs of acute deervation in involved muscles
• Reported with lesions of a nerve, plexus, or root
• Example: hemifacial spasm

Etiology

• Physiologic
  – Hypnic jerks
  – Hiccups
  – Benign Infantile
• Myoclonus-Dystonia
  – Epsilon-sarcoglycan
  – Chromosome 18
  – Sporadic
• Epileptic
  – Epilepsia Partialis Continua
  – Infantile spasms
  – Juvenile Myoclonic Epilepsy
• Symptomatic
  – Postanoxic (Lance Adams)
  – Posttraumatic
  – Myoclonic Dementias
    • Alzheimer’s
    • JCD
  – Basal ganglia disorders
    • CBGD
    • Parkinson’s
    • Huntington’s
    • MSA
  – Drug Induced
  – Post-infectious
  – Inflammatory
  – Infectious (SSPE)
  – Metabolic
  – Toxin Induced

• Progressive Myoclonic Epilepsy
  – Lafora Body Disease
  – Gangliosidosis
  – Gaucher’s
  – Krabbe
  – Sialidosis
  – Baltic Myoclonus
  – MERRF
  – Mitochondrial
    • MERRF
    • MELAS
  – Ceroid lipofuscinosis

Anti-myoclonic meds

• Clonazepam
  – 0.5, 1, or 2 mg tabs
  – Start low
  – Titrate to effect, SE
  – Drowsiness
  – LFTs
  – Wean if needed

• Valproic Acid
  – Can start low (125 BID) up to >1000 mg/day
  – LFTs
  – Fetal malformations

• Levetiracetam
  – 250 BID up to 4000 mg/day
  – Well-tolerated
  – Few med interactions
  – Psychosis
  – Best for cortical myoclonus

• Others:
  – Sodium Oxybate
  – Narcolepsy
  – Acetazolamide
  – Primidone
  – Zonisamide
Specific Myoclonic Syndromes

Startle Syndrome

- Exaggerated startle to sudden unexpected stimuli
  - Hyperekplexia (Familial Startle Disease)
    - AD
    - Mediated by pontine reticular nucleus
    - Can progress into attacks of generalized stiffness

Lance-Adams (post-anoxic)

- Overall, a poor prognostic indicator
- In survivors, can get severe action or intention myoclonus
  - LEV
  - Alcohol

Some studies: there are a bazillion

- The significance of myoclonic status epilepticus in postanoxic coma.
- Young GB, Gilbert LJ, Zochodne DW
- Department of Clinical Neurological Sciences, Victoria Hospital, London, ON, Canada.
- We report 11 adults who exhibited myoclonic status epilepticus (MSE) after cardiac arrest. Based on pathologic, electroencephalographic, and clinical evidence, we conclude that our patients died from the initial anoxic-ischemic insult rather than as a result of MSE. We suggest that the seizures in these nonsurvivors were self-limited events arising from lethal damage to neurons. Thus, in patients with bilaterally synchronous facial myoclonus, bilateral loss of pupillary or oculovestibular reflexes, and suppression and burst-suppression on EEG, it is not warranted to use anesthetic barbiturates to treat MSE.
Outcome from coma after cardiopulmonary resuscitation: relation to seizures and myoclonus.

- Krumholz A, Stern BJ, Weiss HD.

- We studied the effect of seizures and myoclonus following cardiopulmonary resuscitation (CPR) on the outcome of all comatose adult survivors of CPR over an 8-year period. Either seizures or myoclonus occurred in 50 of 114 patients (44%): seizures in 41 patients (36%) and myoclonus in 40 (35%). Status epilepticus or status myoclonus occurred in 36 patients (32%), and 19 (17%) had myoclonic status epilepticus (MSE). Seizures and myoclonus per se were not significantly related to outcome, but status epilepticus, status myoclonus, and, particularly, MSE were predictive of poor outcome as judged by survival and recovery of consciousness.

Myoclonus and Dementia

- Alzheimer’s
  - Triad of myoclonus, dementia, parkinsonism
- CBGD
  - Limb apraxia, alien limb, dysarthria, aphasia, limb myoclonus
- CJD
  - Triphasics/periodic discharges on EEG, often associated with myoclonus

Myoclonus Dystonia

- Intractible Hiccups
  - Make sure there is no structural cause
  - Treatment options:
    - Baclofen
    - Amitriptyline
    - VPA
    - Neuroleptics last choice
Palatal Myoclonus
• A lesion of the ______ can lead to this condition. . .
• If you do not know, pay attention!!! We already discussed this.
• Rhythmic lesions of the palate
• Ear clicking (LTG)
• Carbamazepine, Phenytoin, barbs, botox

Opsoclonus-Myoclonus
• Paraneoplastic or postinfectious (strep)
  – What child tumor commonly causes this?
  – Adult cancers?
  – Infections?
  – Can try IVIG, steroids, plasma exchange
  – Treat cancer

Neurophysiology

Evoked Potentials
• Trivial fact often on boards:
  • Patients with myoclonus often have giant SEPs (N20)
Case Studies

Disclaimer: I ran outta time and borrowed these from Continuum, Movement Disorders, 2007

• 37 y/o woman suffered cardiac arrest after a routine surgery. You are called to see her because she is not waking up but it “twitching”
  - What tests do you want?
  - Treatments?
• She survives (yeah, in real life this hardly ever happens) but continues to have severe disabling myoclonus. When she extends her arms with wrists extended, she has jerking. When she attempts to stand, she has jerks in her legs, producing a bouncing pattern.
  - What is the name of this condition?
  - What kind of myoclonus is she having?

• 72 yr old woman is having involuntary movements of her trunk and thighs for the past year. These occur mainly when she walks. She has normal strength and sensation but tapping her patella elicits the jerks.
  - What type of myoclonus does she have?
  - What test should you order?
  - What treatment options are available?

• A 14 yr old comes to your office with a new onset seizure. In the past year, he has also been told he was “Daydreaming” at school a couple of times. He has also been dropping his orange juice in the morning because his arm flings and he throws the glass.
  - What do you think he has?
  - Treatment options?
  - What would EEG show?