Abnormal movements, when are they important?

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• Hyperkinetic movements
• Hypokinetic movements
• Clinical Course

• Jerky movements
• Twisting movements
• Lack of movements
• Episodic movements
  - Eyes
  - Others
JERKY MOVEMENTS
BENIGN NEONATAL MYOCLONUS – NOT INFREQUENTLY TREATED AS EPILEPTIC SEIZURES

- Clinical Reliability in diagnosis of neonatal seizures 30%
- Request EEG – at least 1 hr
Benign neonatal Myoclonus

- Classically in sleep / around onset or offset of sleep
  - Neonatal – 3 mths
- Can last for several minutes & several months (1/3)*
- Does not need to be bilateral or synchronous
- Can even worsen on holding the limbs
- Self limiting forms can also occur in the awake state
- Maternal drug withdrawal
  - SSRI
  - antipsychotics

Benign Neonatal Myoclonus

<table>
<thead>
<tr>
<th></th>
<th>Classic</th>
<th>Maternal Drug withdrawal*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset</td>
<td>1st month</td>
<td>Up to 2 mths</td>
</tr>
<tr>
<td>Total duration</td>
<td>97% up to 12 months</td>
<td>Up to 3 months</td>
</tr>
<tr>
<td>Sleep association</td>
<td>All phases</td>
<td>Not necessary</td>
</tr>
</tbody>
</table>

*sometimes provoked on treatment with morphine or chloropromazine
Shuddering episodes

- Usually begin in infancy and remit by 4 years of age
- Few seconds long
- Can happen up to 100/day

- Triggering factors include feeding or eating, head movements, and selective tasks such as pressing toys together or sticking a fork into a piece of bread
JERKY MOVEMENTS

TWISTING MOVEMENTS
Torticollis

- Benign Paroxysmal Torticollis
- Spasmus nutans
- Genetic Dystonia with normal MRI
- Sandifer syndrome
Torticollis

• Can be secondary to extra ocular paresis particularly the IVth nerve
• Can be seen with intermittent squint – null point
• In pre-school children new onset torticollis may signify a brain tumor

• In practice – physiotherapy often helps in transient disorders
Dystonia

- Term HIE
- Kernicterus
- Metabolic – mitochondrial disorders, glutaric aciduria
- Neurodegenerative disorders – brain mineralisation
- Genetic dystonias
- Consider treatable disorders
  - Biotin
  - Dopamine
  - Autoimmune
TWISTING REPETITIVE MOVEMENTS
Chorea

- Genetic – some can have a benign course, some may not – Huntington’s disease
- Metabolic – consider Wilson’s disease
- Autoimmune
  - Sydenham’s chorea
  - SLE
  - Encephalitis
- Injury to basal ganglia
- Post cardiac bypass
- Drug induced
- Psychogenic
Stereotypies

- Normal children
- Accompaniment with neuro-developmental syndromes
- Sometimes quite violent – as in autoimmune encephalopathies
- Ebb with age, possibly don’t remit altogether
- In most normal children – reassurance and education are underestimated and under utilised
Tic disorders

• Very common
• It is very important to determine what problem, if any, is a DISORDER
• Identify and treat the comorbidity (ADHD, OCD, Anxiety, Sensory processing disorders, executive dysfunction)
• Behavioural therapy is limited in younger children and when certain comorbidities are over powering
• We medicate few children with tics and of those the medication is rarely for tics themselves
Tremor – causes to think of

- Is there associated dystonia – think of DDx for dystonia
- Has there been drug exposure – anti emetics, stimulants, valproate.
- Is there a family history – Dystonic syndrome, Essential tremor
- Could this be psychogenic?
- Is it tremor – Fast myoclonus, seizure.
SLEEP RELATED MOVEMENTS
SOME RHYTHMIC AND SOME BIZARRE


• 82% of seizures occurred during stage 1 or 2 sleep, with 100% of parasomnias occurring from stage 3 or 4 sleep
### NREM parasomnias v/s Nocturnal Frontal Lobe Epilepsy

<table>
<thead>
<tr>
<th>Features Strongly suggestive of Parasomnia</th>
<th>Moderately favouring parasomnia</th>
<th>Non discriminatory</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yawning</td>
<td>Tremor</td>
<td>Brevity</td>
</tr>
<tr>
<td>Scratching and nose rubbing</td>
<td>Myoclonic jerks</td>
<td>Sitting</td>
</tr>
<tr>
<td>Internal / External trigger</td>
<td>Coughing</td>
<td>Standing or walking</td>
</tr>
<tr>
<td>Waxing and Waning</td>
<td>Semipurposeful behaviour</td>
<td>Preceding “normal” arousal</td>
</tr>
<tr>
<td>Physical/Verbal Interaction</td>
<td>Variability / non stereotypic</td>
<td>Brief arousals</td>
</tr>
<tr>
<td>Sobbing / emotional behaviour</td>
<td>No events on 1st night of monitoring</td>
<td>Fearful emotional behaviour</td>
</tr>
<tr>
<td>Indistinct offset</td>
<td>&lt; 3 events recorded in total</td>
<td></td>
</tr>
<tr>
<td>Failure to fully arouse</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt;2 min</td>
<td></td>
<td></td>
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<tr>
<td>Clinical Feature</td>
<td>Score</td>
<td></td>
</tr>
<tr>
<td>---------------------------------------------------------------------------------</td>
<td>-------</td>
<td></td>
</tr>
<tr>
<td>Age at onset</td>
<td></td>
<td></td>
</tr>
<tr>
<td>At what age did the patient have their first clinical event?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;55 y</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>≥55 y</td>
<td>-1</td>
<td></td>
</tr>
<tr>
<td>Duration</td>
<td></td>
<td></td>
</tr>
<tr>
<td>What is the duration of a typical event?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;2 min</td>
<td>+1</td>
<td></td>
</tr>
<tr>
<td>2-10 min</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>&gt;10 min</td>
<td>-2</td>
<td></td>
</tr>
<tr>
<td>Clustering</td>
<td></td>
<td></td>
</tr>
<tr>
<td>What is the typical number of events to occur in a single night?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 or 2</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>3-5</td>
<td>+1</td>
<td></td>
</tr>
<tr>
<td>&gt;5</td>
<td>+2</td>
<td></td>
</tr>
<tr>
<td>Timing</td>
<td></td>
<td></td>
</tr>
<tr>
<td>At what time of night do the events most commonly occur?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Within 30 min of sleep onset</td>
<td>+1</td>
<td></td>
</tr>
<tr>
<td>Other times (including if no clear pattern identified)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Symptoms</td>
<td></td>
<td></td>
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<tr>
<td>Are the events associated with a definite aura?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>+2</td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Does the patient ever wander outside the bedroom during the events?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>-2</td>
<td></td>
</tr>
<tr>
<td>No (or certain)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Does the patient perform complex, directed behaviors (eg, picking up objects, dressing) during events?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>-2</td>
<td></td>
</tr>
<tr>
<td>No (or uncertain)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Is there a clear history of prominent dystonic posturing, tonic limb extension, or cramping during events?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>+1</td>
<td></td>
</tr>
<tr>
<td>No (or uncertain)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Stereotypy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Are the events highly stereotyped or variable in nature?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Highly stereotyped</td>
<td>+1</td>
<td></td>
</tr>
<tr>
<td>Some variability/uncertain</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Highly variable</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Recall</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Does the patient recall the events?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes, lucid recall</td>
<td>+1</td>
<td></td>
</tr>
<tr>
<td>No or vague recollection only</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Vocalization</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Does the patient speak during the events and, if so, is there subsequent recollection of this speech?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Yes, sounds only or single words</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Yes, coherent speech with incomplete or no recall</td>
<td>-2</td>
<td></td>
</tr>
<tr>
<td>Yes, coherent speech with recall</td>
<td>+2</td>
<td></td>
</tr>
<tr>
<td>Total score</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
ACUTE & EPISODIC MOVEMENT DISORDERS
ABNORMAL EYE MOVEMENTS
Tonic Upgaze/Downgaze

- Onset from infancy
- Mostly <10 minutes; can be for hours
- Upto 10 times/day
- Normal horizontal movements, even during episodes
- Most cases transient and improve by 4 yr
- Sometimes reported with Episodic movement disorders
Opsoclonus-Myoclonus-Ataxia Syndrome

• Early childhood – often due to a neural crest tumour
• Late childhood – can be post-encephalitic
• Early aggressive treatment is essential
• Will often do poorly if untreated
  – 60-80% will have significant cognitive, behaviour, motor and developmental problems (an insidious ‘dementia’)
• Oral steroids, IVIg --- Rituximab
Episodic disorders

- Paroxysmal movement disorder
- Seizures
- Migraine
- Neurocognitive delay
- Paroxysmal Eye movements

Dystonia
- Chorea, Stereotypy

Ataxia
Episodic disorders

- Paroxysmal kinesigenic dystonia or chorea
- Infantile Seizures
- PRRT2
Episodic disorders

- Paroxysmal kinesigenic dystonia or chorea
- Infantile Seizures
- Paroxysmal torticollis
- Hemiplegic migraine

- Exquisitely respond to small dose Carbamazepine
Episodic disorders

Exercise induced dystonia

Paroxysmal eye movements

Dopa Responsive Dystonia
Neurotransmitter disorders

- Consider in infants with signs of dopamine deficiency
- High Phenylalanine on Guthrie card
- Encephalopathic infants without any other explanation
- Oculogyric crises, temperature instability
Pointers to dopamine dysfunction

- Dystonia / Tremor
- Daily fluctuation of symptoms
- Oculogyric crises
- Palpebral ptosis
- Axial hypotonia
- Swallowing/alimentation difficulties, Hypersalivation
- Excessive sweating
- Psychomotor/mental delay
- Unstable temperature
- High Phenylalanine on Guthrie card
- Encephalopathic infants without any other explanation
Episodic disorders

- Paroxysmal exercise induced dystonia
- Intellectual disability
- GLUT1-DS
Episodic disorders

Paroxysmal exercise induced dystonia

Intellectual disability

Early onset absence seizures

Hemiplegic migraine

Nystagmus

GLUT1-DS

Improve on Ketogenic Diet or Modified Atkins Diet

History of improvement with food
Episodic disorders

- Episodic ataxia
- Hemiplegic migraine (familial)
- Seizures

CACNA1a
Episodic disorders

- Episodic ataxia
- Hemiplegic migraine (familial)
- Seizures
- Tonic upgaze/nystagmus
- Paroxysmal torticollis

**CACNA1a**

- Progressive cerebellar atrophy
- May respond to acetazolamide
- 4-aminopyrididine could have a neuroprotective effect
- Caution with phenytoin
When are abnormal movements important?

- Always, but they are not always a disorder.
- Sometimes the movement phenomenology is an important clue to specific diagnoses.
- Sometimes the movements are just a signal to look for more disabling comorbidity.
- If you have one test to request – ask for a video.
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Review article

Paroxysmal tonic upgaze of childhood—a review

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bSave Sight Institute, GPO Box 4337, Sydney, NSW, Australia

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Benign Neonatal Sleep Myoclonus: A Review of the Literature
Valeria O. Maurer, Mattia Rizzi, Mario G. Bianchetti and Gian Paolo Ramelli
*Pediatrics* 2010;125;e919; originally published online March 29, 2010;
DOI: 10.1542/peds.2009-1839
Parasomnias of childhood
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Purpose of review
To enhance the ability of the practitioner to diagnose and manage children with parasomnias in the office setting.

Recent findings
Over 80% of preschool-age children experience parasomnia events. Instability in the regulation of sleep continuity might underlie sleep walking, sleep terrors, and confusional arousals. Catathemia or nocturnal groaning is a parasomnia that is recognized in adults but frequently has onset during childhood.
Review

Developmental and Benign Movement Disorders in Childhood

Cecilia Bonnet, MD, Agathe Roubertie, MD, PhD, Diane Doummar, MD, Nadia Bahi-Buisson, MD, PhD, Valérie Cochen de Cock, MD, PhD, and Emmanuel Roze, MD, PhD
Emilio Fernandez-Alvarez

Transient movement disorders in children