Neurological Symptoms (Non-Cancer)

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Minnesota
Or
Massachusetts?
Neurological Symptoms

- Neurological impairment = NI
  - Seizures
  - Spasticity
  - Autonomic dysfunction
  - Dystonia
  - Myoclonus
  - Central neuropathic pain
General Principles

• Different neurological symptoms have similar features
• Risk for several problems to coexist
• Pain can worsen any neurological symptom
<table>
<thead>
<tr>
<th>Spasticity</th>
<th>Muscle spasm</th>
</tr>
</thead>
<tbody>
<tr>
<td>-- Velocity dependent</td>
<td>-- Intermittent</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Myoclonus</th>
<th>Brief, abrupt, sudden contraction of one or more muscles</th>
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<table>
<thead>
<tr>
<th>Dystonia</th>
<th>Muscle contractions with twisting and repetitive movements, abnormal postures, or both</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Dysautonomia, PAID, Storms</th>
<th>Facial flushing, sweating, hyperthermia, vomiting, gut pain</th>
</tr>
</thead>
</table>

| Central Pain               | Abrupt onset of pain “out of the blue,” gut pain          |
• **Cerebrum**: inhibition
• **Thalamus**: somatosensory transmission (somatic, visceral), arousal and sleep
• **Basal ganglia**: control of movement
• **Hypothalamus**: regulation of heart rate, blood pressure, temperature, and sleep
Pain Behaviors

- Vocalizations: crying, moaning
- Facial expression: grimacing
- Consolability
- Interactivity: withdrawn, less active
- Physiological responses: pale, sweating
- Movement: pulls legs up, restless
- Tone and posture: arching, stiffening
- Idiosyncratic behaviors: laughing

General Principles

• Consider involvement from more than one area of the CNS
• Prioritize problems
• Is it bothering the child?
• Assess for pain behaviors
• Identify medications for each problem
# Empirical Treatment

<table>
<thead>
<tr>
<th>Medication</th>
<th>Conditions</th>
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</thead>
<tbody>
<tr>
<td>Gabapentin</td>
<td>Central neuropathic pain, Dysautonomia, Spasticity</td>
</tr>
<tr>
<td>Clonidine</td>
<td>Dysautonomia, Spasticity</td>
</tr>
<tr>
<td>Tricyclic (TCA)</td>
<td>Central Neuropathic pain</td>
</tr>
<tr>
<td>Baclofen</td>
<td>Spasticity, Dystonia</td>
</tr>
<tr>
<td>Benzo</td>
<td>Myoclonus, Dystonia</td>
</tr>
<tr>
<td>Beta blocker</td>
<td>Autonomic dysfunction</td>
</tr>
<tr>
<td>Benzo, opioid</td>
<td>Autonomic storm</td>
</tr>
</tbody>
</table>
# Empirical Treatment

<table>
<thead>
<tr>
<th>Category</th>
<th>Effect</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Anticholinergics:</strong></td>
<td></td>
</tr>
<tr>
<td>--Trihexyphenidyl</td>
<td>Dystonia</td>
</tr>
<tr>
<td>--Benztropine</td>
<td></td>
</tr>
<tr>
<td><strong>Dopamine agonist:</strong></td>
<td></td>
</tr>
<tr>
<td>--Bromocriptine</td>
<td>Dystonia</td>
</tr>
<tr>
<td><strong>Dopamine depletor:</strong></td>
<td></td>
</tr>
<tr>
<td>--Tetrabenazine</td>
<td>Chorea, Dystonia</td>
</tr>
<tr>
<td><strong>Cannabinoids</strong></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Central neuropathic pain</td>
</tr>
<tr>
<td><strong>Phenobarbital</strong></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Seizures, irritability at EOL</td>
</tr>
</tbody>
</table>
Integrative/Supportive Management

- Rocking, massage, repositioning, music
- Vibratory stimulation (mats, pillows)
- Supportive equipment (seating, pillows)
- Calm environment, sleep
- Pools, massage, aromatherapy
Case #1

- 3 year old with severe NI
- Intractable focal epilepsy on 4 antiepileptic drugs
- Severe daily episodes with facial flushing, sweating, tachycardia, hypertension, arching and posturing
- Some with focal seizures
Case #1 – Care Plan

- Gabapentin and clonidine: 75% improved
- Events with arching and/or tremors
  - Reposition
  - If no stool
  - Ibuprofen
  - Calm, dark room
Case #1 – Care Plan

- Event with flushing and agitation
  - As needed clonidine
- If movement suggests seizure
  - Rectal diazepam
- Not critical to determine if event is seizure or if discomfort is triggering tremors, allow experience when not possible to know with certainty
Case #2

- 3 year old with hypoxic/hypotensive event, MRI basal ganglia
- Dystonic movement and daily irritability
- Meds: clonazepam, lorazepam, gabapentin, methadone, baclofen (trihexyphenidyl stopped due to side effects)
<table>
<thead>
<tr>
<th>Drug</th>
<th>Problems treated</th>
<th>Patient mg/kg/day (14.6 kg)</th>
<th>Typical dose mg/kg/day</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baclofen</td>
<td>Spasticity</td>
<td>1.8</td>
<td>1.9*</td>
</tr>
<tr>
<td>Clonazepam</td>
<td>Dystonia, Spasticity</td>
<td>0.05</td>
<td>0.015-0.03</td>
</tr>
<tr>
<td>Lorazepam</td>
<td>Dystonia, Spasticity</td>
<td>0.2</td>
<td>0.08-0.2</td>
</tr>
<tr>
<td>Gabapentin</td>
<td>Autonomic dysfunction, Neuropathic pain, Spasticity</td>
<td>10</td>
<td>35-45**</td>
</tr>
</tbody>
</table>

*Lubsch 2006, **Korn-Merker 2000
Case #3

- Potential for return of symptoms
- Unable to remove source of problem
- Hoping and preparing
- Transient, new plateau, intractable with ongoing decline
The Face of Palliative Care
Case #3

- Discussion: it is permissible to discontinue any technology that is prolonging suffering
- Introduced with no need for decision
Summary

• Overlap in features of each problem
• More than one neurologic symptom can co-exist in a child with global involvement of the CNS
• Treatment is empiric
• Neuro symptoms can be intractable
Prognosis
You Betcha’

Be well

Do good work

And keep in touch

Garrison Keillor

Julie Hauer

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References


• Hauer J. Caring for Children who have Severe Neurological Impairment: A Life with Grace. Baltimore, Maryland: Johns Hopkins University Press, 2013