Appreciate central pain as a trigger for severe movement and posturing (Attitude)

Discuss treatment options for dystonia (Knowledge)

Practice developing a home based plan for prolonged seizures (Skill)

Objectives

Neurological Impairment (NI)

- Indicated by developmental label
  - cerebral palsy
- A result of a CNS insult
  - hypoxic ischemic encephalopathy (HIE), anoxic encephalopathy, traumatic brain injury (TBI)
- Associated with a specific diagnosis
  - genetic disorder, congenital anomaly, structural brain malformation, or metabolic disorder
Neurological Symptoms

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

General Principles

- Different neurological problems and symptoms have similar features
- Children with global CNS impairment: if 1 exists, risk for others to coexist
- Central neuropathic pain can worsen any neurological symptom
CNS Problems
- Seizures
- Spasticity/Muscle spasms
- Autonomic dysfunction/P A I D
- Dystonia
- Central neuropathic pain

Hauer 2012, 2015

Anti-seizure drugs
  - Benzodiazepines
  - Baclofen
  - Dantrolene
  - Alpha-2 agonists
  - Gabapentinoids
  - Opioids
  - Tricyclic (TCA)
  - Beta blockers
  - Trihexyphenidyl
  - Antipsychotics
  - Cannabinoids

Dystonia
- Involuntary muscle contractions with slow twisting and repetitive movements, abnormal postures, or both
- Can result in pain
- Pain from other sources can worsen dystonic movement

Primary versus Secondary
- Diagnosis versus a label
- Primary Dystonia
- Secondary dystonia
  - Global dystonia
    - Global impairment, microcephaly
  - Risk for co-existing problems (spasticity, seizures, dysautonomia, central neuropathic pain)
Treatment: Goals

- Maximize function
- Decrease pain
- Enhance ease of care-giving
- Interdisciplinary team:
  - Therapists, Neurologists, PM&R
  - Palliative medicine (severe NI)

https://www.aacpdm.org/publications/care-pathways/dystonia

Treatment: Medications

- General indications
  - First line  Baclofen
  - Second line  Trihexphenidyl
- Painful dystonia
  - Gabapentin
- Dystonia with disturbed sleep
  - Clonidine
Empirical Treatment

<table>
<thead>
<tr>
<th>Anticholinergics:</th>
<th>Dystonia</th>
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<tbody>
<tr>
<td>Trihexyphenidyl</td>
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<tr>
<td>Benztropine</td>
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<td>Dopamine depletor</td>
<td>Dystonia, Chorea</td>
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<td>Tetrabenazine</td>
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<td>Gabapentin</td>
<td>Neuropathic pain</td>
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<tr>
<td>Baclofen</td>
<td>Dystonia, Spasticity</td>
</tr>
<tr>
<td>L-Dopa</td>
<td>Dopamine responsive</td>
</tr>
</tbody>
</table>

Treatment: Surgical

- Intrathecal baclofen (ITB) pump
- Deep brain stimulator (DBS)
- Dystonia with pain behaviors: drug trials before surgical interventions
  - Medications for dystonia and pain
  - Gabapentinoid + Tricyclic antidepressant
  - Gabapentinoid + Clonidine

Case – Dystonia

- 3 year old with hypoxic/hypotensive event, MRI – basal ganglia
- Severe dystonic movement and irritability daily
- Meds: clonazepam, lorazepam, gabapentin, methadone, baclofen (trihexyphenidyl stopped due to side effects)
Before Next Intervention

- Consider nociceptive sources
- Maximize drug doses
- Frequency/duration of episodes
- Effectiveness of breakthrough symptom care plan
- Manage possible triggers

Hauer J, Houtrow A, AAP clinical report, June 2017

Integrative Management

- Rocking, massage, repositioning, fan, cool air, music, water, aromatherapy
- Vibratory stimulation (mats, pillows)
- Supportive equipment (seating, pillows)
- Calm environment
- Sleep, constipation

Drug Problems treated Patient mg/kg/day (14.6 kg) Typical dose mg/kg/day

<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>
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</thead>
<tbody>
<tr>
<td>Baclofen</td>
<td>Spasticity</td>
<td>1.8</td>
<td>1.9*</td>
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<tr>
<td>Clonazepam</td>
<td>Dystonia, Spasticity</td>
<td>0.05</td>
<td>0.015-0.03</td>
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<tr>
<td>Lorazepam</td>
<td>Dystonia, Spasticity</td>
<td>0.2</td>
<td>0.08-0.2</td>
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<tr>
<td>Gabapentin</td>
<td>Autonomic dysfunction, Neuropathic pain, Spasticity</td>
<td>16</td>
<td>35-55**</td>
</tr>
</tbody>
</table>

*Lubsch 2006, **Korn-Merker 2000
Case – Dystonia
- Acute illness
- Increase in movement and agitation
- Risk for status dystonicus
- Acute care plan
  - Home
  - Hospital

Status Dystonicus
- Increasingly frequent or continuous severe episodes of generalized dystonic spasms
- Trigger often identified
- Infection, medication toxicity or withdrawal, pain, constipation, puberty, ITB pump failure

Home Care Plan
- **Presenting features:** grimacing, arching, twisting movement, stiffening
- **Routine interventions:** reposition, personal care
- **Non-pharmacologic strategies:**
- **GI tract distention as a trigger:** as-needed supp or enema, vent G-tube
- **As-needed medications:** ibuprofen, morphine, clonidine, or benzodiazepine

Hauer and Houtrow 2017, Hauer 2017
Hospital Care Plan

- Rhabdomyolysis, Respiratory
- Medications
  - Clonidine starting dose 0.004 mg/kg every 6 hours
  - Midazolam continuous infusion

References – Dystonia

- AACPDM Dystonia in Cerebral Palsy Care Pathway, updated June 4, 2018.
  https://www.aacpdm.org/publications/care-pathways/dystonia

References – Dystonia

Barry

- 15 year old, intractable epilepsy, Bi-pap, inpatient every 1-2 months past 6 months, hospitalization with mechanical ventilation
- "Far from his baseline"
- Goals: comfort, quality of life, intubate if "reasonable" chance for recovery
- 3 months later: 3 hospitalizations in 6-weeks for prolonged seizures

Hypothetical Trajectory

- **Fixable, Modifiable, or Irreversible**
- **Preserving health, Re-goaling, Preserving Comfort**
- **Improve**
- **Progressive**

**Intractable**: not easily relieved or cured

- Test and “Fix”
  - Fracture
  - Renal stones
  - Bladder infection
  - Respiratory infection
- Modify with risk for intractable
  - Seizures
  - Dysautonomia
  - GI motility
  - Central pain
  - Mucus clearance
Barry: 15 year old, intractable seizures

- “I wish he could be seizure free, I worry…”
- **What we may not want to do:** “It might make sense not to use (intubation, IVs) given that it doesn’t improve his (seizures, GI), what are your thoughts...?”
- **What we can do: Offering options**
  - Location of Care
  - Non-invasive management (home or hospital)
  - Use at home, Trial in the hospital

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**Home Treatment Plan**

- Diazepam rectal
- Lorazepam
  - 0.1mg/kg (4mg) buccal/rectal
- Midazolam
  - 0.2mg/kg (10mg) buccal/intranasal
- Rectal AEDs: carbamazepine, phenobarbital, lamotrigine, and valproic

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**Home Treatment Plan**

- For seizure that persists for 5 minutes, give
  - Diastat 25 mg rectal
  - Lorazepam 4 mg buccal
- For seizure that persists another 10 minutes (20 min total) give lorazepam 4 mg buccal, may repeat 10 minutes later (30 min total)
- 4 hours after last dose of lorazepam:
  - Lorazepam 2 mg every 8 hours VGT x 3, then
  - 1 mg every 8 hours VGT x 3, then
  - 1 mg every 12 hours x 2, then stop

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Barry: 15 year old, intractable seizures

- 6 months later: Bipap discontinued
- 1 ½ years later, acute ileus: “One option is to send him to the hospital to place an IV. I think the best plan is to use a trial of pedialyte by feeding tube.”

Thank you!

Keep in touch!

julie.hauer@childrens.harvard.edu