Assessment of Inconsolability in Children with Neurodevelopmental Delay

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Neuro-palliative Care

• Lars is a 3-year old boy with severe neurological impairment (NI), history of infantile spasms, gastroesophageal reflux, constipation

• 4 months in and out of hospital: “continuous crying”, “abdominal pain”, “not tolerating feeds”, “toning and arching”

Sources of Somatic Pain

• Dental
• GER disease
• Cholecystitis
• Pancreatitis
• UTI
• Renal stones

• Fracture
• Hip subluxation
• Other

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• Interactivity: withdrawn, less active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


Pain Assessment Tools

• Revised-Face, Legs, Activity, Cry, Consolability (R-FLACC)
  • Allows addition of specific behaviors
• Individualized Numeric Rating Scale (INRS)
  • Parents identify behaviors that indicate no pain to the worst possible pain
• NCCPC-PV, NCCPC-R, PPP

### Sources of Pain Behaviors

- **Spasticity**
- **Autonomic Dysfunction**
- **Visceral Hyperalgesia**
- **Central Neuropathic Pain**

Blue boxes = impaired nervous system

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### Nociceptive Evaluation

- Initial nociceptive evaluation:
  - BMP, CBC, ALT, alk phos, t bili, lipase
  - UA/UCx
  - Gastric pH (patient with G-tube)
  - X-ray or bone scan if fracture suspected
  - Dental exam if no exam in 12 months

- Other studies to consider:
  - pH probe, abdominal U/S, endoscopy

### Acute versus chronic

- “Acute” symptoms
  - New somatic pain source
  - Increasing symptoms from neurological source
- Is your child typically calm or often “agitated”??

### Attitudes/Misconceptions

- Fear of missing source
- This is how such children look
- Need to know if it is pain to treat
- Focus on spasticity
- Language takes focus from pain
  - Agitation
  - Neuro-irritability
General Principles

- Features of neurological problems include pain behaviors
- Risk for several problems to coexist
- WHO pain ladder
  - Not studied in this group

<table>
<thead>
<tr>
<th>Cerebral injury</th>
<th>Loss of central inhibition</th>
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</thead>
<tbody>
<tr>
<td>Central pain</td>
<td>Abrupt onset of pain &quot;out of the blue,&quot; GI tract pain with distention</td>
</tr>
<tr>
<td>Visceral hyperalgesia</td>
<td>Sensitization of visceral afferent pathways, gut pain with distention</td>
</tr>
<tr>
<td>Dysautonomia, PAID, Storms</td>
<td>Facial flushing, sweating, hyperthermia, vomiting, gut pain</td>
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<tr>
<td>Dystonia, paroxysmal autonomic instability with dystonia (PAID)</td>
<td>Muscle contractions with twisting and repetitive movements, abnormal postures, or both</td>
</tr>
<tr>
<td>Spasticity, Muscle spasms</td>
<td>Often not painful, can be triggered by pain</td>
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</tbody>
</table>

Tone is Altered by Pain

- 21/22 with benefit for persistent pain behaviors
- 19/22 (86%) with intermittent increase in tone: muscle tensing, stiffening of extremities, back arching, abnormal movements
- 20/22 (91%) on drugs for spasticity

Hauer, Solodiuk 2015

General Principles

- Assess for pain behaviors
- Assess for somatic pain source
- Review length of symptoms
- Consider the CNS
- Prioritize problems
- Identify medications trials
- Consider triggers

Triggers

- Increased severity due to loss of central inhibition?
- Constipation and GI feedings
  - Trigger due to distention of GI tract
  - Central pain, visceral hyperalgesia
  - Impaired sleep
  - Illness, new nociceptive source
**Empirical Treatment**

<table>
<thead>
<tr>
<th>Medication</th>
<th>Indications</th>
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<tbody>
<tr>
<td>Gabapentinoids</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>Methadone</td>
<td>Central Neuropathic pain</td>
</tr>
<tr>
<td>Baclofen</td>
<td>Spasticity, Dystonia</td>
</tr>
<tr>
<td>PRN (opioid, benzo)</td>
<td>Autonomic storm</td>
</tr>
</tbody>
</table>

**Neuro-Pain Ladder**

- **Step 1**
  - Gabapentin OR Pregabalin +/- adjuvant

- **Step 2**
  - Tricyclic (TCA) OR Clonidine +/- adjuvant

- **Step 3**
  - Methadone +/- adjuvant

**Adjuvants:** opioid, benzodiazepine

**References**

**Thank you**

Be well  
Do good work  
And keep in touch

Garrison Keillor

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