Managing Pain in Children with Complex Chronic Conditions

*Irritation/agitation/pain, due to severe impairment of the central nervous system*

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Chronic pain is frequently seen in children with severe impairment of the central nervous system, often referred to as children with severe neurological impairment (SNI). In addition to discomfort for your child and distress for you and your family, chronic pain can result in poor sleep and worsened health. This summary is intended to increase your understanding of why children with SNI can have recurrent pain, and how your child’s medical team can improve your child's comfort. This is a very complex problem to understand. Take the time you need and use this information to identify questions for your child’s medical team.

**Points of Irritability, Agitation and Pain**

- **Irritability** is defined as an abnormal response to stimulus
- **Agitation** is defined as unpleasant state of arousal manifesting as irritability, restlessness, and increased motor activity
- **Pain** is a cause of irritability and agitation; other causes include an altered emotional state, medication toxicity, and acute illness

**There are 2 type of pain**

- **Tissue injury pain** alerts us to injury or inflammation of tissue in the body. This type typically comes on quickly and will resolve once the cause has been found and treated.
  - Examples of tissue pain include a bone fracture or bladder infection.

- **Nerve (neuropathic) pain** is a chronic form of pain, often with recurrent episodes of different intensity. This form of pain can occur in children with diseases or injury of the brain. Nerve pain can also develop after tissue injury pain has resolved, such as healing after surgery or healing of the intestinal tract. Episodes can occur suddenly, with no explanation or known trigger.
What you may see during episodes of pain

- Intermittent muscle tensing or tightening: increased spasticity, stiffening of legs, draws up arms, clenched fists, tremors or jerks, tense and still, kicking, thrashing
- Change in body position: back arching, rigid and stiff, head movement, thrashing
- Change in facial expression: grimace or frown, clenched jaw, distressed look, eyes wide open, or nonexpressive face
- Change in vocalizations: crying, soft moaning, grunting, gasping
- Child is difficult to console or comfort
- Change in interaction: withdrawn, less active
- Skin changes: pale or flushed skin, sweating

Reasons to consider medications

- Intermittent episodes occur without a clear or consistent cause
- These episodes have been occurring for more than three months
- Some episodes may have an explanation, yet symptoms keep returning after treatment of various problems
- The frequency and duration of episodes have a negative impact on your child’s life, such as:
  - Three or more episodes each week, with episodes lasting more than one hour
  - A cycle of daily episodes for one week out of every 3-4 weeks

Medications that your child’s doctors may consider (do not administer any of these without a prescribed plan from the doctor)

- Medications for neuropathic pain
  - gabapentin, pregabalin, nortriptyline, methadone
- Medications for autonomic dysfunction
  - clonidine, gabapentin, morphine
- Medications as needed for breakthrough symptoms
  - clonidine, lorazepam or clonazepam, morphine
- Medications for associated problems: such as treatment for spasticity or dystonia
  - baclofen, clonidine

Medication trial
Waiting for benefit and worrying about side effects can feel challenging for many parents. Here are some things to consider.

- Lessening your child’s sedation versus improving comfort: determine if it is more important to you to lessen sedation while monitoring for improvement in comfort, or if it is okay to be sleepier for the first 1-2 weeks. This information can help your child’s doctor determine how fast they will increase the medication dose
- Your child’s sedation can mean the drug is working because pain is exhausting improved pain control can result in the child catching up on lost sleep
• The sedation will likely get better. It is hard to be tolerant when you want your child both comfortable and awake; seek support and know it will get better, as your child becomes better rested through improved sleep and as your child’s doctor determines the dose that makes the best sense
• Although it is hard to wait, it can help to tolerate increased sleeping for the first two weeks, rather than decreasing the dose too soon: the dose that initially causes extra sleeping can often later be the best dose to help your child be comfortable awake during the day and sleep at night
• Plan on 2-4 weeks to determine this balance and if the medication will help. You may be exhausted by worry or poor sleep; it is hard to be patient through this process yet being prepared for this time frame may help you plan
• Pain treatment will not stop your child from experiencing and displaying pain from a new cause of pain: you will know if your child has a new cause of pain that needs to be identified, such as pain from a urinary tract infection

Non-medication strategies
• Comfort strategies: cuddling, rocking, massage, warm baths, music
• Repositioning and supportive seating systems: to minimize positional pain
• Pressure and vibration: weighted blankets, vibratory mats and pillows
• Complementary and integrative therapies: essential oils, Reiki
• Lessen distention of the intestinal tract: treat constipation, try a suppository during a pain episode to determine if symptoms decrease after a bowel movement, vent the gastrostomy feeding tube, assess for overfeeding
• Risk for overfeeding: calorie needs can be overestimated by 30% or greater in children with SNI who have a low body temperature (less than 95 F), limited movement of extremities, successful pain treatment with a reduction in intermittent muscle spasms, and general decline in activity

The goal is symptom free. This goal is not possible for all children when the altered nervous system is the cause of symptoms, a cause that cannot be fixed.

• Medications used for nerve pain can benefit many children: the goal with a scheduled medication is to decrease the frequency and severity of episodes; some have better symptom control with two medications that treat nerve pain
• Be prepared for breakthrough symptoms due to the inability to fix the nervous system; some children continue to have breakthrough symptoms even with two or three medications for chronic pain
• For some children, two scheduled medications and an effective care plan for breakthrough symptoms is the “best” balance as depicted in the figure below
• Decreasing too many medications: development of a breakthrough symptom plan –including use of as needed medications –and decreasing intestinal tract distention can help lessen too many scheduled medications.

The AAP link below has further information (pages e17-19) and care plan examples (e21-e22).
Example of a care plan in the event of breakthrough pain

With symptoms such as back arching and/or muscle tremors, facial flushing, leg stiffening, appearing distressed

Start with the following interventions:
- Reposition and review for any personal care need
- Hold feeds if being given and vent gastrostomy tube
- If no stool during the day
  - give suppository if not yet given that day
  - give as needed enema if suppository already given
- Give ibuprofen and clonidine
- Place in calm, quiet environment
- If features are not starting to decrease after 30 to 45 minutes, give lorazepam
- Call if new concerns arise or if symptoms persist

Steps your child’s doctor can consider before adding a 2nd or 3rd medication
- Maximize the dose of other medications being used for chronic pain
- Review and modify with your input the care plan for breakthrough symptoms
- Review constipation management and risk for overfeeding
- Review and manage other contributing problems

Resource for your child’s medical team (AAP – American Academy of Pediatrics)
https://pediatrics.aappublications.org/content/pediatrics/139/6/e20171002.full.pdf

How good is good enough?

<table>
<thead>
<tr>
<th>Pain</th>
<th>Frequent episodes</th>
<th>3 or fewer severe episodes per week that require a drug (other than Tylenol or Ibuprofen) and benefit within 1 to 2 hours to interventions for breakthrough episodes (pages e19 and e21-e22)</th>
<th>Fewer pain episodes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wakefulness</td>
<td>Poor sleep</td>
<td>Awake during most of day, improved sleep</td>
<td>Sleepy during the day</td>
</tr>
</tbody>
</table>

These are general guidelines. This balance can help you and your child’s medical team guide decisions to increase a medication dose or add another medication if the episodes are too frequent or not responding well to the care plan for breakthrough episodes of discomfort / agitation.

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You and your child’s medical team will always be the best experts in navigating how to use this information. Seek experts if your child’s symptoms are not easily improved, such as the expertise of a palliative care provider. You and your child deserve such expertise as you navigate problems that do not always respond as hoped for to the medical interventions available.

Questions you may ask your child’s medical team, with information for consideration (per Julie Hauer, MD)

Q: Are there tests that will tell me that this is nerve pain versus tissue injury pain?
Information: Tests can identify causes of tissue injury. Because there are not medical tests to confirm nerve pain, the best “test” is a medication trial.

Q: If the medication for nerve pain does not help, does this mean my child doesn’t have nerve pain?
Information: Nerve pain can be difficult to treat in some cases because the damage to the nervous system cannot be fixed or cured. This means that for some children, the combination of two different medications can be more effective than either medication used alone.

Q: Should we do all tests looking for causes of tissue injury before starting a medication for nerve pain?
Information: The first tests typically look for the most likely causes of pain to treat. Sometimes it makes sense to start a medication for nerve pain while considering some other tests as well.

Q: My child seems anxious. Do you think this is depression or anxiety?
Information: You and your child’s medical team certainly want to consider all reasons why your child appears anxious and agitated. Is there a pattern of when these events happen? For example, do they occur on days when your child goes to school but not on the weekends? This could suggest she is tired or stressed by the long day at school and more comfortable when at home. In that instance, you could work with therapy and school to give her time out of her chair in a reclined position that might be more comfortable.

Q: My daughter has other problems that cause pain. How will I know when this is due to her spasticity (gastroesophageal reflux, recurrent clostridium difficile, tracheitis) versus nerve pain?
Information: Children with SNI are at risk for more than one cause of pain. Treating nerve pain can help the team understand how much the nerve pain
may be responsible for some of her pain. If a medication decreases nerve pain, this can help decrease muscle spasms given that the body tenses when it is in pain.

Q: How will I know when a breakthrough symptom is due to the nervous system versus a new cause of pain?

Information: If you are worried or uncertain, call the clinic during the day and the on-call person at night. Over time, you and the team will get a better sense of which features tend to respond to the breakthrough care plan and which features seem different. For example, if there is a fever, it might make sense to assess for a bladder infection if the cause of fever with pain seems different or is not clear.

In closing: Be assured, you and your child’s medical team will always be the best guide to the care plan that makes best sense for your child, and how to approach decisions each step along the way.