Courageous Parents Network Guide

Managing Pain in Children with Complex Chronic Conditions

Chronic Irritability /agitation/ pain due to severe impairment of the central nervous system

Chronic pain occurs far too often 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, it can result in poor sleep and worsen health.

This summary is intended to increase your understanding of why such children 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 that alerts us to injury or inflammation of tissue in the body
- Nerve pain due to altered transmission of pain in the nervous system

Tissue injury pain causes sudden onset of pain

- Pain will resolve once the cause has been found and treated
- Examples 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 "out of the blue" 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: a grimace or frown, clenched jaw, distressed look, eyes wide open, or nonexpressive face
- Change in vocalizations: crying, soft moaning, grunting, gasping
- Difficult to console or comfort
- Change in interaction: withdrawn, less active
- Skin changes: pale or flushed skin, sweating

Reasons to consider medications for chronic pain in children with SNI:

- Intermittent episodes occur without a clear or consistent cause
- These episodes have been occurring for more than 3 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
    - 3 or more episodes each week, with episodes lasting more than 1-hour
    - A cycle of daily episodes for 1 week out of every 3 to 4 weeks

Medications that the doctors may consider

- 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

Medication trial: waiting for benefit and worrying about side effects can feel challenging for many parents

- Lessening 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 OK to be sleepier for the first 1 to 2 weeks; this information can help your child's doctor determine how fast they will increase the medication dose
- Sedation can mean the drug is working because pain is exhausting: improved pain control can result in a child catching up on lost sleep
- Sedation will 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 best sense for your child
- Wakeful and comfortable: though it is hard to wait, it can help to tolerate increased sleeping for the first 2 weeks rather than decreasing the dose too soon, as 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 to 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 2 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 2 or 3 medications for chronic pain
- For some children, 2 scheduled medications and an effective care plan for breakthrough symptoms is the "best" balance as depicted in the figure below
- Lessening 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).

Breakthrough care plan example

Care plan for events with back arching and/or muscle tremors, other features may include facial flushing, leg stiffening, appearing distressed

1. 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
1. Give ibuprofen and clonidine
2. Place in calm, quiet environment
3. If features are not starting to decrease after 30 to 45 minutes, give lorazepam
4. Call if new concerns 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?

Pain	Frequent episodes	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)	Fewer pain episodes
Wakefulness	Poor sleep	Awake during most of day, improved sleep	Sleepy during the day

These are general guidelines. This balance can help 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.

I hope this information has been helpful as you seek the best outcome possible for your child. 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 palliative care teams. You and your child deserve such expertise as you navigate problems that do not always respond as hoped for to the medical interventions available.

Examples of questions for your child's medical team, with information for consideration

Are there tests that will tell me that that this is nerve pain versus tissue injury pain?
Tests can identify causes of tissue injury. Because there are not medical tests to confirm nerve pain, the best "test" is a medication trial. What we do know is that your child is at risk for this type of pain.

If the medication for nerve pain does not help, does this mean my child doesn't have nerve pain?
Nerve pain can be difficult to treat in some because the damage to the nervous system cannot be fixed or cured. This means that for some children, the combination of 2 different medications can be more effective than either medication used alone. We will help support you throughout this process as we hope that your child has benefit with the 2nd medication we discussed adding.

Should we do all tests looking for causes of tissue injury before starting a medication for nerve pain?
The first tests we did have looked for most of the causes of pain we would want to identify and treat. Sometimes it makes sense to start a medication for nerve pain while considering the few other tests we would consider.

My child seems anxious. Do you think this is depression or anxiety?
We certainly want to consider all reasons why someone can appear anxious and agitated. Based on our best understanding of your child's developmental age, I

would anticipate that you could console her if this was more emotional. But lets discuss this further to see if there is a pattern of when these events happen. Are they on the days when she goes to school and not on the weekends? This could suggest she is tired or stressed by the long day at school and more comfortable when at home. If this is a consideration, we can work with therapy and school to give her time out of her chair in a reclined position that might be more comfortable.

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?

Children like your daughter are at risk for more than one cause of pain. Treating nerve pain can help us understand how much this might be an underlying reason 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. This can also decrease pain episodes that result in tests that can be difficult to interrupt. As an example, it can be hard to know if a positive culture from the trachea is a new infection versus bacteria that cling to the tracheostomy tube, called colonization. This is also true for some of the tests used to detect C. difficile.

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

If you are worried or uncertain, call the clinic during the day and the on call person at night. Overtime we will get a better sense of what features tend to respond to the breakthrough care plan and what features seem different. We will also learn more anytime we do tests. As an example, there has not been a bladder infection when we have checked. It makes it less likely to be a new problem the next time there is pain without other new features. If at another time there is a fever, it might make sense to assess for a bladder infection if the cause of both pain and fever is not clear. We are always here to support you along the way.

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.