

## A standardized approach to idiopathic pain in children with neurological conditions

Hal Siden<sup>1</sup> Tim Oberlander<sup>1</sup> Ashley Wilson<sup>2</sup> Shalu Duggal<sup>1</sup>  
1. University of British Columbia, Vancouver, BC; 2. Canuck Place Children's Hospice, Vancouver, BC

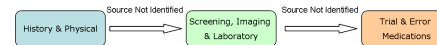
### BACKGROUND

Children with rare diseases affecting the nervous system often experience pain due to unknown causes.<sup>1</sup> The diseases in question include rare neurodegenerative diseases, metabolic diseases, and other conditions due to single gene defects. These children frequently have language delay and cannot localize pain.

Some sources of pain can be identified through careful physical examination, laboratory and imaging studies. However, many times careful examination reveals no obviously correctable sources for pain and irritability. At that stage, empiric therapies are undertaken to reduce neuropathic and central nervous system pain signals and reduce irritability and agitation.

Unfortunately, there is no standard or common approach to treating this idiopathic pain.<sup>2</sup> Parents and caregivers often use physical-behavioural techniques such as rocking, swaddling, and environmental calming. Medications commonly used by experienced clinicians include non-steroidal anti-inflammatory drugs (NSAIDs, acetaminophen), tricyclic antidepressants, anti-convulsants (gabapentin), anti-psychotics (risperidone), and opiates.<sup>3</sup>

Because we do not understand the neurotransmitter disruptions in children with rare diseases that lead to idiopathic pain, medication choices and the sequence in which they are introduced are highly empiric. The sequence differs from one physician to another, and at times a physician will use different sequences in different patients without specific rationale. There is, however, evidence that standardized approaches developed by experts and revised in a continuous quality improvement (CQI) process can result in better care.<sup>4</sup>



### METHODS

6 physicians specializing in pediatric pain and/or palliative medicine were recruited to pilot a targeted approach to pain management. To initiate the study, these physicians were presented with a case study of a child with a neurological condition displaying pain-like behaviors. Physicians were asked to list their top 5 medications and the order in which they would trial them.

#### Case Study:

*You are presented with a child with a genetic and/or metabolic and/or neurological condition who is displaying "pain"-like behaviors. The child is non-verbal and cannot localize the pain for you, nor can he/she provide a reliable signal as to the degree of discomfort. The behaviors that are pain-like include vocalizations with crying out, grimacing, and wrinkling the forehead. There are tears occasionally but not always. There is arching and stiffening, especially of the extensor muscles. The episodes are not predictable, can last from a few minutes to 1/2 hour, and will disrupt sleep.*

*Having initiated and completed an extensive work-up you are not able to determine an obvious or treatable nociceptive/inflammatory cause of the pain-like behavior. The work up includes a careful history which reveals no specific pattern. The physical examination does not identify a cause; the child has many handicaps but none of these have changed recently. Spasticity and hypertonicity are present but not increased and the child is on baclofen. The seating system has been reviewed. Gastrostomy feeding is tolerated. Constipation is treated. A urinalysis is normal. X-rays of the hips and spine are unchanged. An abdominal ultrasound is not revealing. Treatment with acetaminophen, ibuprofen, positioning and cuddling has not made a difference.*

*You are left to consider either a nociceptive/inflammatory cause for this pain which you cannot identify, or a case of CNS "irritability" (which might also be called neuropathic pain or central pain). You decide at this point in time that an empiric trial of medication is warranted. There are many classes of medication to choose from - all of them have central nervous system effects, but only some are traditional analgesics, while others are considered "adjuvants" for pain control and span a number of classes.*

**In this exercise you will choose 5 medications you include in your trial, and a sequence for the trial.**

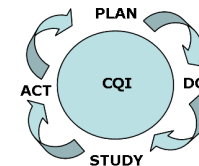
### RESULTS

The physicians are identified only by letter, A-F. One physician provided three different sequences in response to the case vignette; those sequences are shown as physician A1,A2,A3.

PHYSICIAN	1 <sup>st</sup> Drug	2 <sup>nd</sup> Drug	3 <sup>rd</sup> Drug	4 <sup>th</sup> Drug	5 <sup>th</sup> Drug
A1	Opiate	Benzodiazepine	Methadone	Gabapentin	TCA
A2	Opiate	Gabapentin	TCA	SRI	Atypical Antipsychotic
A3	Opiate	TCA	Benzodiazepine	Atypical Antipsychotic	Methadone
B	Atypical Antipsychotic	Gabapentin	Benzodiazepine	Tramadol	Opiate
C	Gabapentin	TCA	Atypical Antipsychotic	Benzodiazepine	Tramadol
D	Gabapentin	Opiate	Atypical Antipsychotic	Benzodiazepine	Methadone
E	Benzodiazepine	Gabapentin	Atypical Antipsychotic	Opiate	n/a
F	Benzodiazepine	Opiate	Atypical Antipsychotic	Methadone	Gabapentin

### DISCUSSION

Because we do not fully understand the disruptions resulting in pain in children with rare neurological conditions, medication choices and sequences are highly empiric, as demonstrated in the study. The lack of consensus leads to ineffectiveness and frustration for families and professionals alike. There is evidence that standardized approaches revised in a continuous quality improvement process can result in better care. We have recently undertaken a pilot study of a CQI approach to pain management in this population. Results will be presented in the near future.



### REFERENCES AND ACKNOWLEDGEMENTS