Pain and Symptom Management: Children with Severe Neurological Impairment
Presenters

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Objectives

• Identify the symptoms of pain and discomfort in children with SNI
• Recognize the importance of the parent history and relationship with their child in identifying and treating pain
• Describe the common etiologies of pain in children with SNI
• Discuss both pharmaceutical and non-pharmaceutical pain treatment modalities
Severe Neurological Impairment

- Children with severe physical and developmental impairment
- Limitation in communication
- Wide range of diagnoses (although cerebral palsy, TBI, HIE researched)
- Wide range of prognoses: but cannot be “cured” and lifespan is limited
Is Pain a Problem?

- Pain in children with severe neurological impairment is under-recognized, under-treated and negatively affects quality of life.

- Communication challenges and multiple pain etiologies complicate diagnosis and treatment.

- Most studies done in cerebral palsy population.
Pain in Children with Cerebral Palsy

• One time questionnaire, 252 respondents

• Over 55% of children had pain on a daily basis

• For 25% that pain was severe enough to affect activities and impair mobility/function

(Penner et. al. Pediatrics Vol. 132 No. 2 August 1, 2013)
Frequency of Pain Symptoms

- A prospective study over a 4 week period with moderate to severe cerebral palsy
  - untreated pain “once or twice” in 44%
  - untreated pain “fairly often” to “every/almost every other day” in 21% of children

(Houlehan et.al. Dev Med Child Neuro 4 p305-10. 2004)
Pain and Level of Impairment

• Pain occurred weekly in 44% of children with moderate to profound cognitive impairment

• Pain occurred almost daily in 41% of children with greater impairment

Recognition of Pain Symptoms

- Detailed history/physical/observation required.
- Children with SNI have behaviors that physicians may not recognize as pain.
- Parents often recognize symptoms but challenging to verbalize it as pain or discomfort.
- The pain behaviors often become a behavioral/personality trait or “the way they are” and dismissed by both parents and physicians.
Cerebral Palsy “Colic”

- Uncontrollable bouts of crying in children with SNI in the first year of life

- Sometimes pain is associated with another symptom (constipation, formula) by parents

- Family may be so overwhelmed with all other medical issues that this may get lost, or only brought up if associated to a medical concern
Case 1

- Baby ML is a 10 month old with history of chromosomal abnormality, severe hypotonia and developmental disability, congenital heart disease s/p repair, FTT, feeding issues, and infantile spasms.

- He was always “crabby” and hard to console. He was hospitalized for several months, and was never observed to smile.
Audiology exam showed OME, parents became focused on that as cause for pain (multiple ER/clinic/specialty visits)

- Finally articulated chronic pain symptoms
- Started neuropathic pain medication
Case 1

- Parents reported, smiling, laughing and much more interactive

- Making some developmental gains in therapy

- Tolerating feeds better, weight gain, decreased seizures
Case 1

- Was this acute pain? No.
  - The acute issue brought to light the chronic pain

- Acute issues in children with SNI often reflect increasing severity of chronic symptoms
- Once parents are taught to observe for pain behaviors, they are often able to vocalize and advocate.
Pain Behaviors In SNI

- Vocalization, crying, moaning
- Facial expression, grimacing
- Consolability (cerebral palsy “colic”)
- Interactivity: withdrawn, less active
Pain Behaviors in SNI

- Movement: pulls legs up, restless
- Tone and posture: arching, stiffening
- Physiological responses: pale, sweating, tachycardia, tachypnea, increased salivation/secretions
Idiosyncratic Pain Behaviors

- Laughing with pain

- Withdrawal: lack of facial expression (if usually animated), sudden loss of skills/milestones

- Increased aggression or self-injurious behaviors. Atypical antipsychotic vs. pain medication?
Parents as Advocates

• Many parents worry about the difficulty of recognizing their children’s pain

• When asked, they often say that they do not know

• Ask them to track troubling behaviors (arching, crying, moaning, withdrawal, agitation)
Types of Pain

- Nociceptive pain
- Neuropathic pain:
  - Central pain
  - Peripheral Neuropathic pain
  - Autonomic instability (sympathetic storming)
  - Visceral hyperalgesia
Sources of Pain

- Nociceptive pain - pain associated with tissue damage or inflammation:
  - GERD, constipation, pancreatitis, gallstones
  - kidney stones, UTI
  - fracture/dislocation
  - Surgical pain
Nociceptive Pain

• PLEASE REMEMBER
• When doing work up for nociceptive pain (pancreatitis) there is no contraindication for treating the pain
• Patients with SNI get less pain medication for nociceptive pain (injury/surgery) than verbal patients
Neuropathic Pain

• Changes in the nervous system that result in abnormal transmission of pain signals. Often accompanied by
  
  – Hyperalgesia: increased response to a painful stimulus
  
  – Allodynia: pain due to a stimulus that does not usually cause pain (Sickle cell)
Peripheral Neuropathic Pain

- Diabetes, PVD, trigeminal neuralgia
- Phantom limb pain
- More than 20% of children with cerebral palsy will exhibit peripheral neuropathic pain after orthopedic surgical procedures (sciatica)
Central Pain

- Neuropathic pain resulting from impairment in the CNS in the region of the spinothalamic tract
- Associated with hypoxic ischemic encephalopathy, post stroke, brain trauma, also Rett syndrome
- In adults—degenerative brain disorders
Autonomic Dysfunction

• Also called dysautonomia or autonomic or sympathetic “storming”

• Altered heart rate, blood pressure, temperature, flushing, sweating, retching, vomiting, increased salivation

• Same vital sign changes as if pain
Case 2

- NC is a 4 year old with metachromatic leukodystrophy
- Has had progressive loss of function. No seizures. She is very spastic and tight. Arches and cries with movement/transfers.
- Started baclofen, diazepam and morphine with some benefit
Case 2

• Started gabapentin and after 2 months
  – Was able to discontinue the morphine
  – Mother reports she is much more comfortable, will smile and respond positively to touch
  – Is not crying constantly with transfers and movement
Case 2

- After 6 months of comfort, was hospitalized for pneumonia for 3 weeks
- After this admission mother prioritized comfort and QOL over aggressive care
- Discharged on oxygen, and was again irritable, spastic and arching, with some new autonomic storming
- Added methadone and morphine and was able to keep her comfortable until EOL
Treatment of Dysautonomia

- Bromocriptine (studies show best usage after acute injury—TBI, HIE)
- Gabapentin
- Clonidine
- Beta blockers
- Benzodiazepines
- Morphine, methadone,
Visceral Hyperalgesia

• Increased pain response to GERD and constipation, despite medications for these (often accompanied by retching/vomiting)

• Feeding intolerance

• Pain behaviors associated with “normal gut behaviors” passing gas, tube feeding and bowel movements
Treatment of Neuropathic Pain

- Central pain /visceral hyperalgesia/
  - Step 1: Gabapentin /pregabalin +/- adjuvant
  - Step 2: Clonidine or TCA +/- adjuvant
  - Step 3: Methadone +/- adjuvant
Gabapentinoids

- Mechanism: binds to alpha 2 delta subunit of voltage dependent calcium channels in brain and spinal cord
- Weak Antiepileptic activity
- Minimal drug interactions, side effects sedation, nystagmus
- Gabapentin (liquid, capsule) and pregabalin (Lyrica)
Gabapentin Dosing

- May start as young as 2 month

- Start with low dose (2-3/Kg/dose) and titrate up to 20/kg/dose

- May take 2 month for maximal effect

- Max dose 2400-3600 mg/day or 50-70mg /kg/day
Clonidine

- Central acting alpha 2 adrenergic agonist
- Reduces sympathetic outflow
- Useful for dysautonomia and also possible anti nociceptive effects
Clonidine

• Comes in liquid, pill or patch

• Start with liquid or pill—transition to patch after oral dose is determined.

• Cannot be stopped abruptly, needs to be weaned
Tricyclic Antidepressants

- Presynaptic reuptake inhibitors of serotonin/norepinephrine in CNS, inhibits pain transmission
- Nortryptiline has fewer drug-drug interactions than amitriptyline
- Need for EKG, dependent on goals of care
Nortryptiline

- May be used in combination with gabapentin (adult studies for neuropathic pain show greater benefit with both medications)

- Data in press: about 2/3 of children with SNI show benefit with Gabapentin alone, 1/3 will require second medication (Hauer)
Benzodiazepines

- Mechanism: increase affinity of GABA for GABA receptor
- Treatment of intermittent muscle spasms (mostly diazepam), anxiety and seizures
- Treatment of intermittent autonomic storms
Benzodiazepines

- Lorazepam
- Diazepam
- Midazolam (short, amnesia, pre-op)
- Clonazepam (longest duration)
Adjuvant to consider

- Cyproheptadine—mechanism: serotonin 2, histamine 1 and muscarinic receptor blocker

- Used for retching- and associated vomiting, also hypothermia and itching
Gastrointestinal Symptoms

Most common cause for pain/discomfort in children with severe neurological impairment.

• The enteric nervous system has been described as a "second brain"

• The enteric nervous system can operate autonomously

• It normally communicates with the CNS through the parasympathetic (vagal nerve) and the sympathetic (prevertebral ganglia)
80% of kids with CP have GERD
-retching is not characteristic of GERD

- Medications vs. surgery
- Issues with repetitive surgery
- Continuous and J tube feeds
Constipation

- Aggressive treatment of constipation (please)

- Neither spasticity nor immobility is p.r.n., so constipation cannot be treated p.r.n.

- May need both a softener and a stimulant
  - MUSH and PUSH
Constipation in SNI

- Untreated and undertreated constipation
  - exacerbates GERD (increases respiratory secretions)
  - can cause feeding intolerance
  - have caused FTT/malnutrition

- Can also be a hallmark of neurological decline
Retching and vomiting

- Long nociceptive differential for children with tube feeding:
  - GERD
  - Impaired gastric emptying,
  - Complication of Nissen fundoplication
  - Pancreatitis (think viral or drug induced)
  - G-tube complication
  - Malrotation
  - H. pylori
  - Untreated constipation
  - Elevated ICP
Retching

- Consider that retching with or without vomiting can be central (cyproheptadine)
- Consider visceral hyperalgesia
- Need a detailed history to differentiate VH from central pain
History for Retching/vomiting

- Timing of pain behaviors and vomiting
- Characteristic of the vomiting (with retching or not)
- Cough association: before or after the emesis?
- Use therapists and nursing in-hospital
- Coach parents to look “with new eyes” so that they can advocate for their children
Pharmacologic Treatments

• Treatment for GERD and constipation should always be maximized first!

• Treatment for visceral hyperalgesia
  – Gabapentinoid
  – Benzodiazepines
  – Clonidine
  – nortryptiline
Non Pharmacological Strategies

• For intestinal symptoms
  – Venting
  – Farrell bag usage
  – Modified timing of feeds
  – Reduced rate of feedings
  – Reduced amount of feedings (consistent with goals of care)
Non Pharmacological Treatments

• Comfort strategies: cuddling, rocking, massage, repositioning

• Warm baths/aqua therapy, swaddling, music with headphones, weighted blankets

• Vibratory stimulation-data to support this
  – vibration mats and CPT vests
Integrative Therapies

• Little data in our patient populations

• Experience is anecdotal

• Massage therapy and aromatherapy

• Be careful with herbal preparations
Consider an Empiric Trial of Neuropathic Pain Treatment

- Before considering a Nissen fundoplication (in a child already gastrostomy tube fed)

- Before considering moving to continuous or J-tube feeds

- For a child with SNI and “behavioral issues”
Palliative Care

- All care for children with SNI is palliative care, by definition

- Quality of Life is the priority

- Attention to symptoms, asking for pain history, will improve QOL
Role of the Caregivers

• We ask our parents to take on multiple professional roles in the care of their children: role of nurse, respiratory therapist, diagnostician, advocate

• Children with SNI have limited interaction skills, attending to pain improves them and can vastly improving quality of life

• Improving child’s pain behaviors and interaction can improve the parents quality of life
A true friend knows your weaknesses but shows you your strengths; feels your fears but fortifies your faith; sees your anxieties but frees your spirit; recognizes your disabilities but emphasizes your possibilities - “

William Arthur Ward
References

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