Neurological Symptoms (Non-Cancer)

Julie Hauer, MD 2nd Conference on Paediatric Palliative Care: a Global Gathering November 20, 2014



Minnesota Or

Massachusetts?

Neurological Symptoms

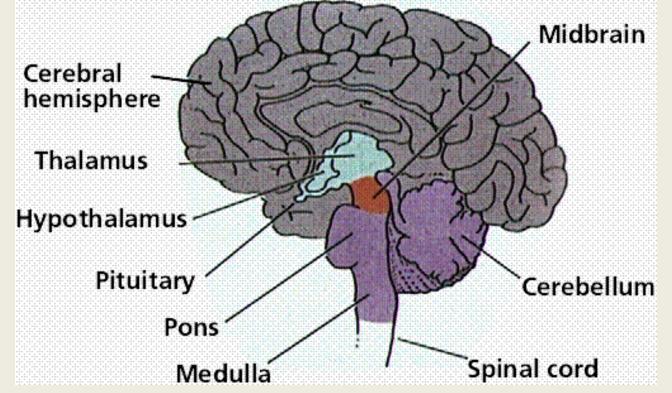
- Neurological impairment = NI
 - Seizures
 - Spasticity
 - Autonomic dysfunction
 - Dystonia
 - Myoclonus
 - Central neuropathic pain

General Principles

- Different neurological symptoms have similar features
- Risk for several problems to coexist
- Pain can worsen any neurological symptom



| Spasticity Muscle spasm | Velocity dependent Intermittent |
|-------------------------------|--|
| Myoclonus | Brief, abrupt, sudden contraction of one or more muscles |
| Dystonia | Muscle contractions with twisting and repetitive movements, abnormal postures, or both |
| Dysautonomia, PAID, Storms | Facial flushing, sweating, hyperthermia, vomiting, gut pain |
| Central Pain | Abrupt onset of pain "out of the blue," gut pain |



<u>Cerebrum</u>: inhibition
Thalmus:

somatosensory transmission (somatic, visceral), arousal and sleep

- <u>Basal ganglia</u>: control of movement
- <u>Hypothalamus</u>: regulation of heart rate, blood pressure, temperature, sleep

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- Interactivity: withdrawn, less active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

Hunt 2004, Breau 2002, Malviya 2006

General Principles

- Consider involvement from more than one area of the CNS
- Prioritize problems
- Is it bothering the child?
- Assess for pain behaviors
- Identify medications for each problem

Empirical Treatment

| Gabapentin | Central neuropathic pain, Dysautonomia, Spasticity | |
|-----------------|--|--|
| Clonidine | Dysautonomia, Spasticity | |
| Tricyclic (TCA) | Central Neuropathicpain | |
| Baclofen | Spasticity, Dystonia | |
| Benzo | Myoclonus, Dystonia | |
| Beta blocker | Autonomicdysfunction | |
| Benzo, opioid | Autonomic storm | |

Empirical Treatment

| Anticholinergics: Trihexyphenidyl Benztropine | Dystonia | |
|---|-------------------------------|--|
| Dopamine agonist Bromocriptine | Dystonia | |
| Dopamine depletor Tetrabenazine | Chorea, Dystonia | |
| Cannabinoids | Central neuropathic pain | |
| Phenobarbital | Seizures, irritability at EOL | |

Integrative/Supportive Management

- Rocking, massage, repositioning, music
- Vibratory stimulation (mats, pillows)
- Supportive equipment (seating, pillows)
- Calm environment, sleep
- Pools, massage, aromatherapy

Case #1

- 3 year old with severe NI
- Intractable focal epilepsy on 4 antiepileptic drugs
- Severe daily episodes with facial flushing, sweating, tachycardia, hypertension, arching and posturing
- Some with focal seizures



Case #1 – Care Plan

- Gabapentin and clonidine: 75% improved
- Events with arching and/or tremors
 - Reposition
 - If no stool
 - Ibuprofen
 - Calm, dark room



Case #1 – Care Plan

- Event with flushing and agitation
 - As needed clonidine
- If movement suggests seizure
 - Rectal diazepam
- Not critical to determine if event is seizure or if discomfort is triggering tremors, allow experience when not possible to know with certainty

Case #2

- 3 year old with hypoxic/hypotensive event, MRI basal ganglia
- Dystonic movement and daily irritability
- Meds: clonazepam, lorazepam gabapentin, methadone, baclofen (trihexyphenidyl stopped due to side effects)

| Drug | Problems treated | Patient mg/kg/day (14.6 kg) | Typical dose mg/kg/day |
|------------|--|-----------------------------------|------------------------------|
| Baclofen | Spasticity | 1.8 | 1.9* |
| Clonazepam | Dystonia, Spasticity | 0.05 | 0.015-0.03 |
| Lorazepam | Dystonia, Spasticity | 0.2 | 0.08-0.2 |
| Gabapentin | Autonomic dysfunction, Neuropathic pain, Spasticity | 10 | 35-45** |

*Lubsch 2006, **Korn-Merker 2000



- Potential for return of symptoms
 - Unable to remove source of problem
 - Hoping and preparing
 - Transient, new plateau, intractable with ongoing decline

The Face of Palliative Care





- Discussion: it is permissible to discontinue any technology that is prolonging suffering
- Introduced with no need for decision

Summary

- Overlap in features of each problem
- More than one neurologic symptom can co-exist in a child with global involvement of the CNS
- Treatment is empiric
- Neuro symptoms can be intractable

Prognosis



You Betcha'

Be well

Do good work And keep in touch

Garrison Keillor

Julie Hauer

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References

- Hauer J, Wical B, Charnas L. Gabapentin successfully manages chronic unexplained irritability in children with severe neurologic impairment. Pediatrics. 2007;119(2):e519-22.
- Hauer J. Improving Comfort in Children with Severe Neurological Impairment. ProgPalliat. 2012; 20(6):349-56
- Hauer J. Caring for Children who have Severe Neurological Impairment: A Life with Grace.
 Baltimore, Maryland: Johns Hopkins University Press, 2013