

# Symptom management in children with severe neurological impairment

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**Ireland's 4<sup>th</sup> International Children's**

**Palliative Care Conference**

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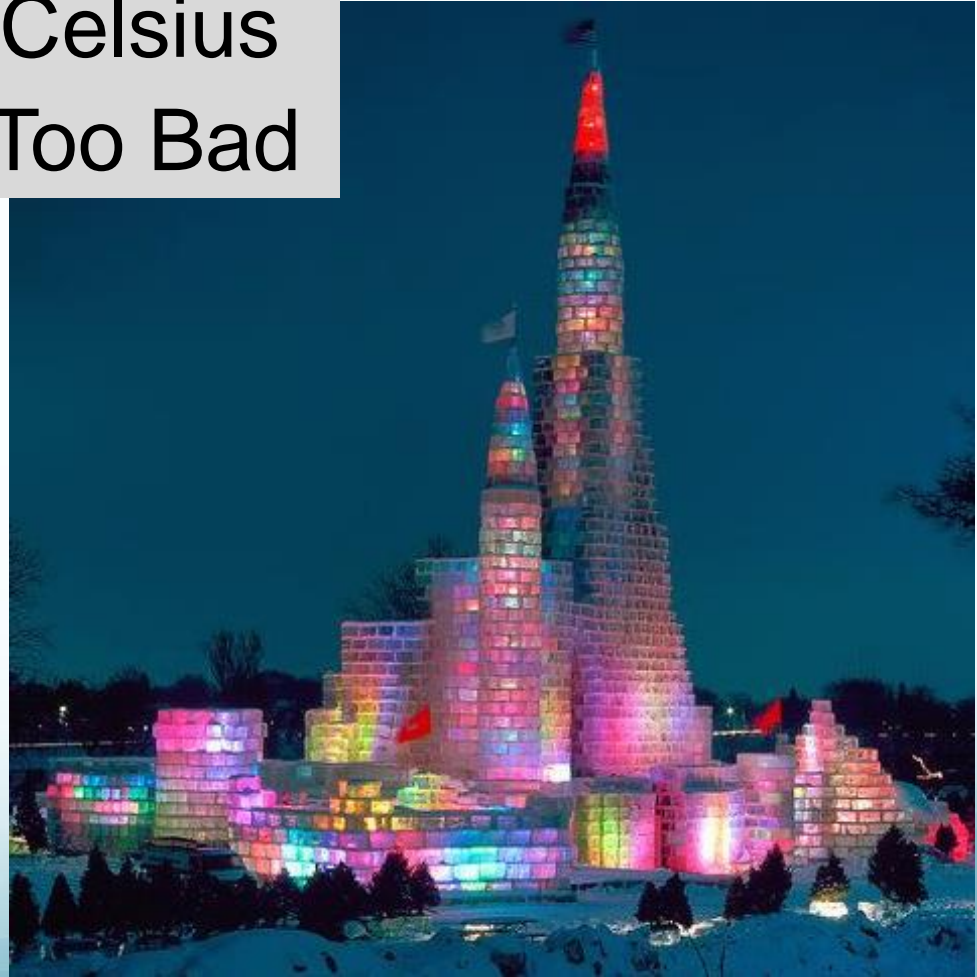


Boston Children's Hospital



HARVARD MEDICAL SCHOOL  
TEACHING HOSPITAL

-25 Celsius  
Not Too Bad



# Severe Neurological Impairment

- Neurodevelopment disabilities
- Intellectual disability, cerebral palsy
- **Diagnosis:** anoxic or traumatic brain injury, in-utero infection, metabolic, genetic, congenital, neuro-structural malformations



- ❑ Eliminate symptoms: pain, agitation, dystonia, muscle spasms
- ❑ Goal: not possible for many with severe neurological impairment (SNI)
- ❑ How good is good enough?



# Pain Assessment and Treatment in Children With Significant Impairment of the Central Nervous System

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Significant problem for children with impairment of the central nervous system, with the highest frequency and severity among children with the greatest impairment. Despite the significance of this population, this population remains vulnerable to underrecognition and undertreatment of pain. Barriers to treatment may include uncertainty about diagnosis, limited resources, and lack of information about

abstract

FREE

# Communicating a Need

- ❑ **Agitation:** Unpleasant state of arousal manifesting as **irritability**, restlessness, and increased motor activity
- ❑ **Irritability:** abnormal response to stimuli or physiological arousal
- ❑ **Causes:** **Pain**, anxiety, acute illness, drugs

# Pain Behaviors

- ❑ Vocalizations: crying, moaning
- ❑ Facial expression: grimacing
- ❑ Consolability
- ❑ Interactivity: withdrawn, less interactive
- ❑ Physiological responses: pale, sweating
- ❑ **Movement: pulls legs up, restless**
- ❑ **Tone and posture: arching, stiffening**



# Sources of Symptoms

## ○ Nociceptive:

### *Acute and Acute on Chronic*

- Fracture
- UTI
- Pancreatitis
- GERD

## ○ Neuro-Pain:

### *Chronic*

- Central pain
- Visceral hyperalgesia
- Dysautonomia
- Spasticity



# Chronic Neuro-Pain

- ❑ > 3 months
- ❑ Impacts QOL and health outcomes
- ❑ Highest risk:
  - Severe to profound intellectual disability
  - GMFCS level 5
  - Multiple co-morbidities

Hauer & Houtrow 2017, Friedrichsdorf et al 2017, Steele et al 2014,  
WHO & IASP (International Association for the Study of Pain)

- Basal ganglia:  
control of movement

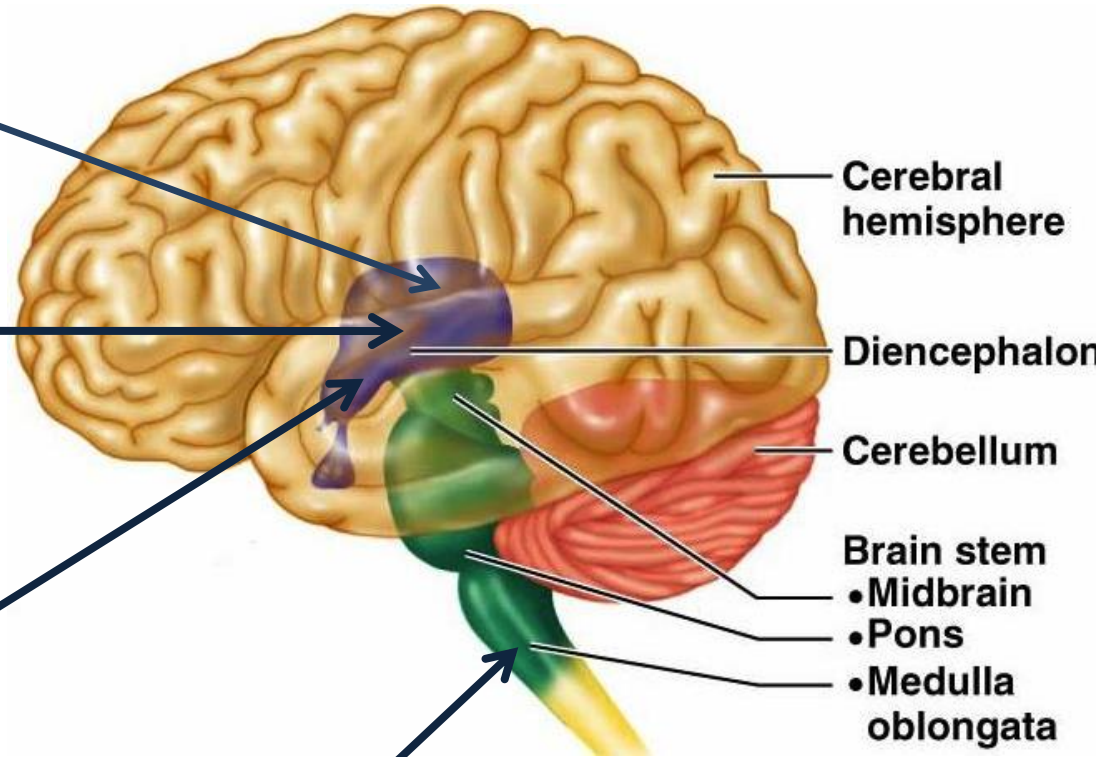
***Dystonia***

- Thalamus:  
somatic/visceral  
sensory signals

***Central Pain***

- Hypothalamus:  
autonomic regulation

***Dysautonomia***



Visceral afferents:  
***Visceral hyperalgesia***

## **Presenting features**

## **Causes due to altered nervous system**

### **Pain Behaviors**

Central pain, Visceral hyperalgesia, Dysautonomia, Dystonia, Spasticity

### **Changes in tone and posture**

Central pain, Visceral hyperalgesia, Dysautonomia, Dystonia, Spasticity

### **Intestinal tract pain, emesis**


Central pain, Visceral hyperalgesia, Dysautonomia



# Medication Trials, Figure 2

**First Line Scheduled**

Gabapentinoid



**Second Line Scheduled**

Clonidine

Tricyclic antidepressant



**Third Line Scheduled**

Methadone

Cannabinoid

**As needed:** opioid, clonidine, benzo

# Empirical Treatment

<b>Gabapentinoids</b>	Central neuropathic pain, Dysautonomia, Visceral hyperalgesia, spasticity
<b>Clonidine</b>	Dysautonomia, Spasticity, Sleep
<b>Tricyclic (TCA)</b>	Central neuropathic pain, Sleep
<b>Methadone</b>	Central neuropathic pain
<b>Cannabinoids</b>	Central neuropathic pain
<b>PRN (opioid, benzo, clonidine)</b>	Breakthrough pain, Spasms, Autonomic storm

Dosing guidelines in AAP Clinical Report Table 8

# Communication

## Mindful Communication and Empathy

- Parental information:
  - Nociceptive vs neuropathic
  - No test to confirm
  - “your son is at risk for this type of pain”
- Manage worries:
  - Pain treatment will not stop pain from new source
  - “sedation means the drug is working”
  - Expectations: time for trial, breakthrough pain



# Communication

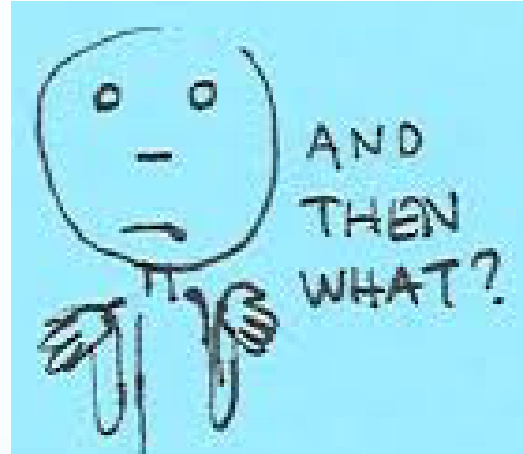


**COURAGEOUS**  
**PARENTS**  
**NETWORK**

- ❑ Courageous Parent's Network chronic neuropathic pain parental information sheet
- ❑ On site, click on "Guides", then click on "Neuropathic Pain"
- ❑ [https://courageousparentsnetwork.org/guides/neuropathicpainmanagementguide?blm\\_aid=3434497](https://courageousparentsnetwork.org/guides/neuropathicpainmanagementguide?blm_aid=3434497)

# Before Next Drug Trial

- ❑ Consider nociceptive sources
- ❑ Manage co-morbid problems
  - Spasticity, dysautonomia
- ❑ Manage triggers: GI tract distention
- ❑ Event frequency, breakthrough care plan
- ❑ Drug dose(s) maximized





# “Required” Calories

Resting energy expenditure (REE) X stress/activity factor	Comments
REE X 1.6	Ambulatory
REE X 1.1	Nonambulatory (CP)
REE X 0.8	Many with SNI
REE X 0.5-0.6	Hypothermia, hypotonia, limited movement

Hauer 2013, Gale R, et al 2016, Hauer 2017

# Breakthrough Care Plan

- ❑ Goals: decrease polypharmacy
- ❑ Presenting features
- ❑ Routine interventions
- ❑ Non-pharmacologic strategies
- ❑ GI tract distention as a trigger
- ❑ As-needed medications

Hauer and Houtrow 2017; Hauer 2017

# How Good is Good Enough?

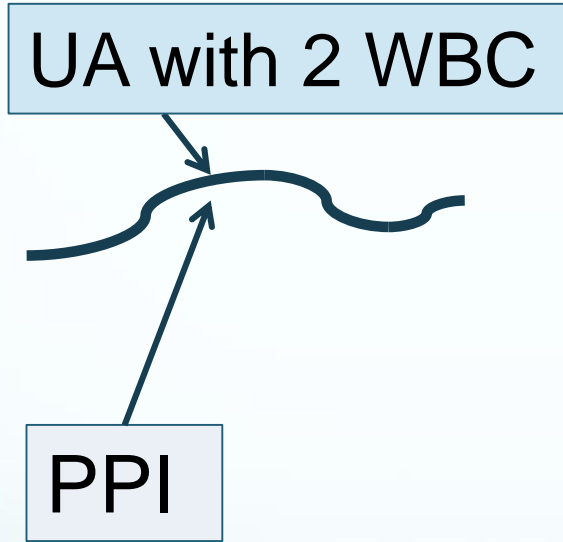
Meds	1	2	3
Pain	Frequent Episodes	Fewer severe episodes per week, benefit within 1 to 2 hours of breakthrough plan	Fewer Episodes
Sleep	Poor Sleep	Awake During Day, Asleep at Night	Sleepy During the Day

# Worries

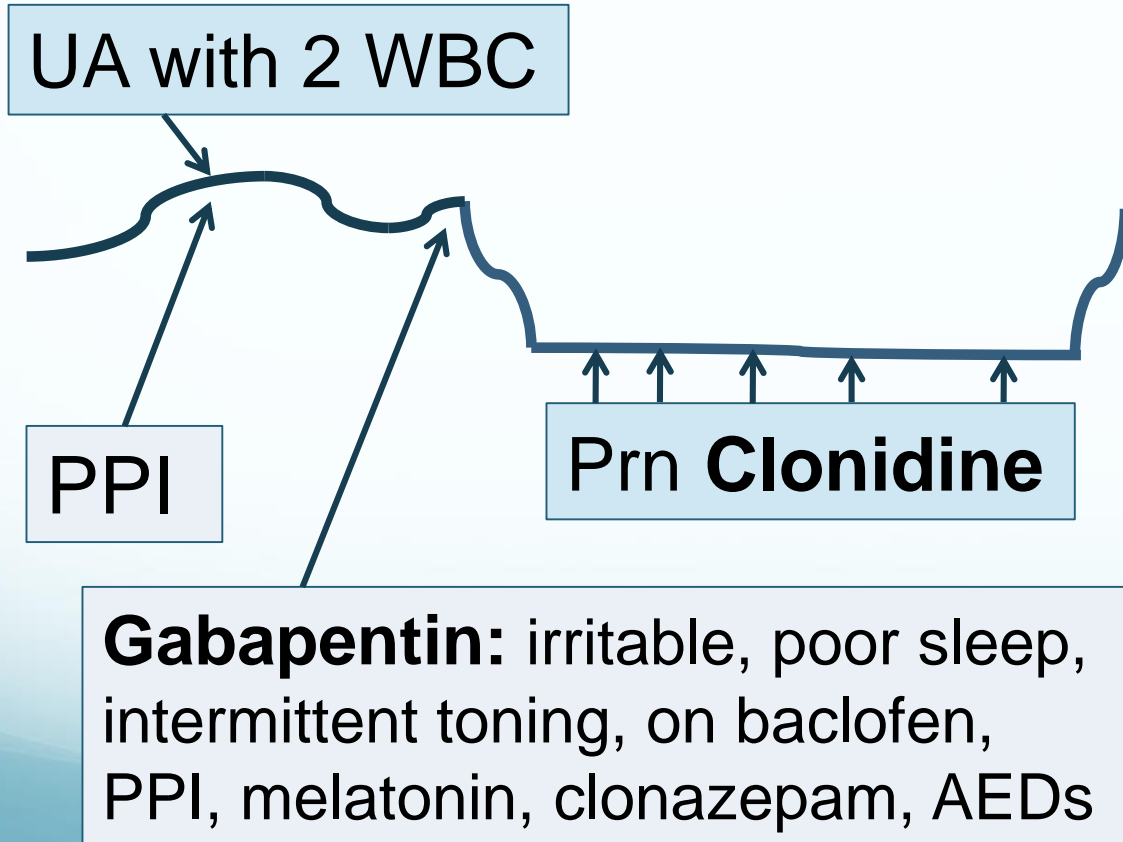
- ❑ Fear of missing a problem
- ❑ Fear of not trying hard enough
- ❑ How good is good enough



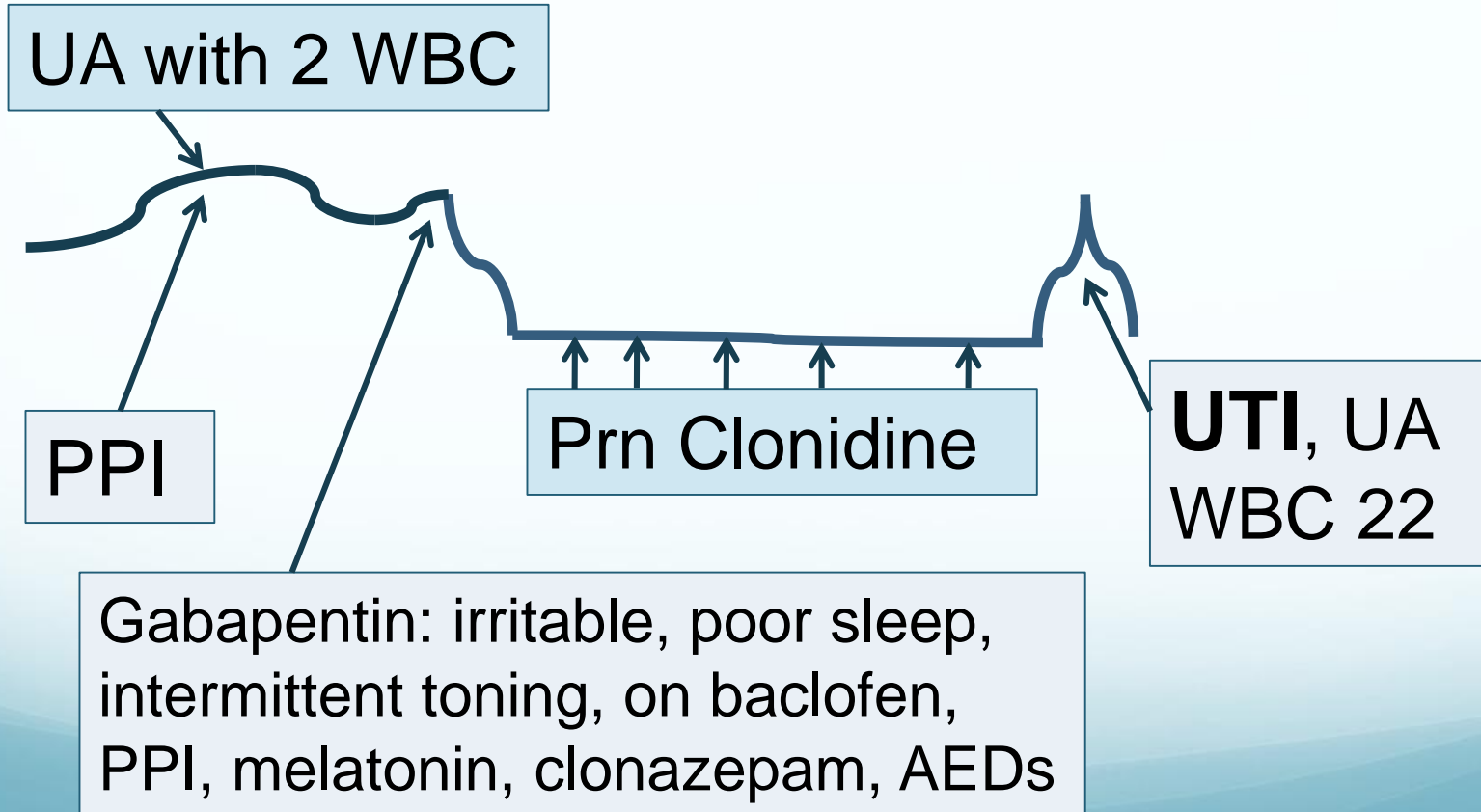
# Acute and Chronic Pain



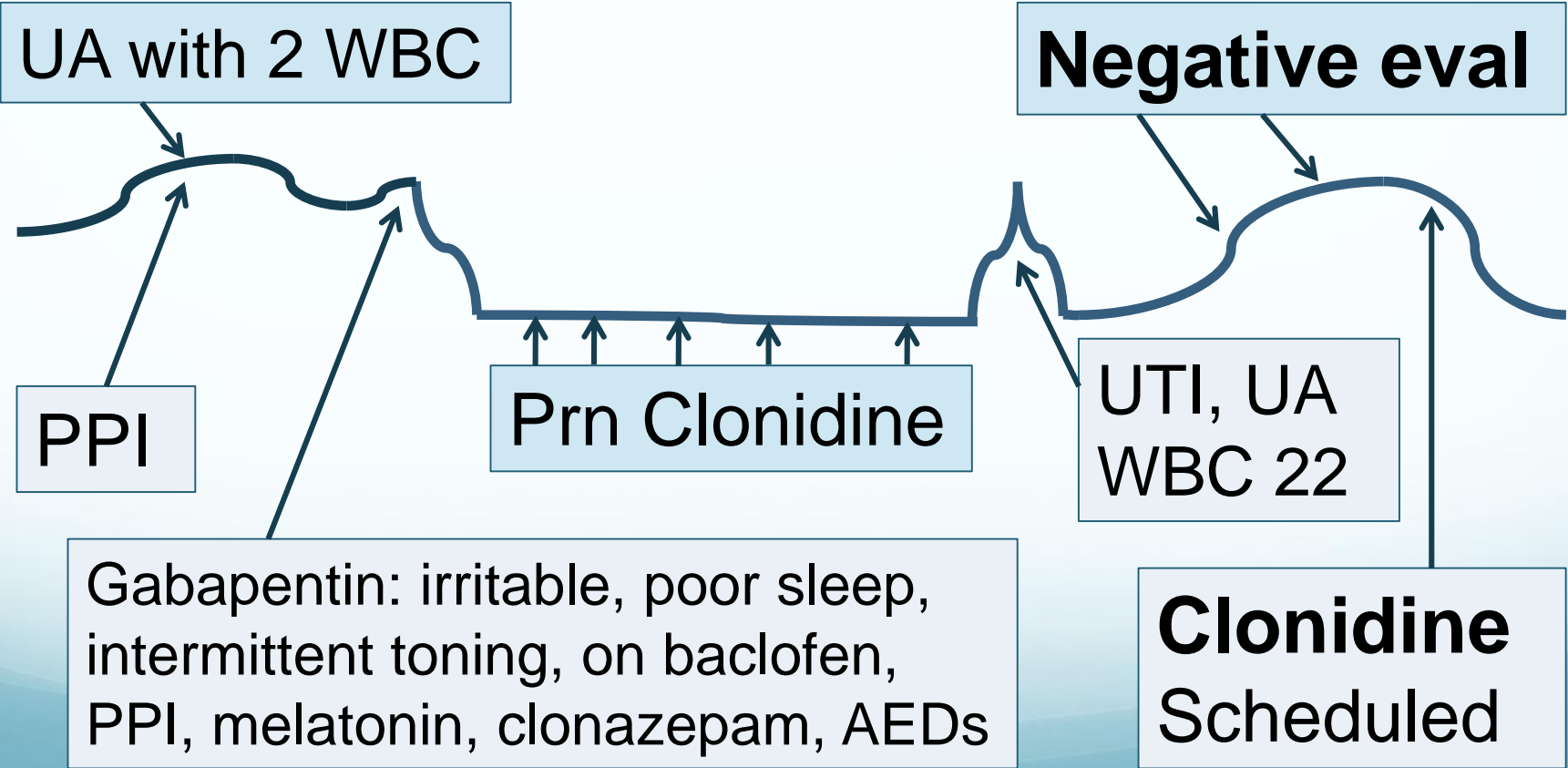
# Acute and Chronic Pain



# Acute and Chronic Pain



# Acute and Chronic Pain





# 28 kg

Day	TCA	Citalopram 20mg QHS	Gabapentin 400mg TID	Glycopyrrolate
1	5 mg	15 mg	500/500/500	0.5mg TID
5	10 mg	10 mg	500/500/600	0.5mg TID
9	15 mg	5 mg	500/600/600	0.5mg BID
13	20 mg	5 mg	600 TID	0.5mg BID
17	25 mg	Stop	600 TID	Stop

Significant improvement, smiling/giggling, sleeping, no daytime sedation, continued benefit 1-year later

# Clonidine Dosing

- Scheduled clonidine:
  - ✓ Starting dose 2 mcg/kg/dose
  - ✓ Average dose 0.02 mg/kg/day (6 mcg/kg/dose)
- As needed clonidine
  - ✓ Same dose every 3 or 4 hours prn
- Severe dystonia hospital plan
  - ✓ Starting dose 4 mcg/kg/dose every 6 hours

# Parental Fear and Anger

- ❑ I wish there were tests to tell us with certainty “why” and therefore “how” to treat
- ❑ I imagine this is hard as we talk about sources that might be improved but not fixed
- ❑ I am worried we can't remove all symptom episodes. What is most important to you?
- ❑ Previous tests have been negative, at this time I would add a 2<sup>nd</sup> medication, what are your thoughts?

# Persistent Symptoms

- **“I wish** we could eliminate all symptoms, **I worry..., I wonder...”**
- **What we may not want to do:** “It might make sense not to use (intubation, IVs) given his difficult symptoms, what are your thoughts...?”
- **What we can do / Offering options:**
  - Location of Care
  - Non-invasive management (home or hospital)

# Areas for Study

- ❑ Screening tool: chronic neuro-pain
- ❑ Criteria for first medication trial
- ❑ Script to assist with buy-in and fears
- ❑ Breakthrough care plan use
- ❑ Criteria to minimize polypharmacy

# Summary

- ❑ Treat testable sources
- ❑ Modify CNS sources (non-testable)
- ❑ Consider triggers (GI tract distention)
- ❑ Identify trials: medications, gut rest, breakthrough symptom care plans
- ❑ Modify as needed

**Thank You**  
**Go raibh maith agat**

**Keep in Touch!**

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