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Screamin' for No Reason

An Approach to Severely Neurologically Impaired Children with
Unexplained Irritability.



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- I have no financial relationships to disclose





- Will go over some cases
- Differential diagnosis-search for causes of pain
 - Common
 - Less common but important
- Autonomic storming detour
- Suggested initial lab work up.



Case 1

- 12 year old girl with HIE after a pulmonary embolus post renal transplant
 - Admitted with : 2day hx of severe irritability
 - PHM
 - GJ fed due to feeding intolerance
 - Recurrent aspiration pneumonia
 - Recurrent periods of crying, agitation, Temp to 40 degrees C, Exam: agitated, arching back, crying, tachycardia to 190, marked tachypnea with increased work of breathing and increased oxygen requirement, Elevated BP to 170/120). No vomiting, BM's a bit loose, occur every other day



12 year old girl with HIE after a pulmonary embolus post renal transplant

Labs:

WBC 17.4 70% neutrophils. Plts, Hgb normal.

CXR: no infiltrates. Some chronic changes

U/A normal

Electrolytes, LFT's lipase normal, creatinine normal.



Abd X ray: marked constipation



Golytely clean out. Mom had stopped miralax due to concerns on the internet about it's safety

Calmed down and returned to baseline.

Gave Mom information on miralax and resumed Rx after clean out.



Take Home message

- Constipation is a major cause of irritability/autonomic storming



12 year old girl with HIE after a pulmonary embolus post renal transplant

Returned 1 month later with similar symptoms except now vomiting
Now having fevers, agitation, tachypnea, increased oxygen requirement to 5L/min by face mask. Heart rate up to 175.
No hx of constipation. Taking miralax with daily BM's



Labs:

WBC 20.3. 75% neutrophils
CRP 15 mg/L
Creat increased to 1.14 (normal for him 0.71).



- CXR: GJ tube coiled up in esophagus



Take Home message

- Look for sources of pain/discomfort



- 4 year old female with laryngomalacia, tracheostomy, arthrogryposis, trigonocephaly, absence of corpus callosum, feeding dysfunction s/p GJ tube and recurrent ICU admissions for fever admitted with severe irritability that is worse with feedings, fever to 40.5, increased work of breathing. No cough/cold symptoms, vomiting, or diarrhea. BM's normal. Note: hx of recurrent admissions for "urosepsis"
- Exam:
- GENERAL: Irritable, agitated. Arches back.
- HEAD: severe microcephaly Marked retrognathia.
- Neck: tracheostomy with trach collar
- RESPIRATORY: increased RR
- GI: GJ tube in place.
- MUSCULOSKELETAL: diffuse contractures most noticeable on hands, feet.
- NEUROLOGIC: profoundly delayed No eye contact with me.



- CXR, KUB neg
- WBC 29.2 64 N, 20 L 16 M
- CRP 1.0 mg/L



- Admitted, given bolus of fluids, iv ceftriaxone and vancomycin
- B/C neg, U/A , U/C neg
- 1/2L/min oxygen. Later weaned
- Rx'd propranolol. clonidine in G tube and improved. Required prn morphine for agitation
- Feeds slowly advanced.



Take home message

- Autonomic storming can mimic sepsis



5 year old boy with HIE due to arrest after episode of Guillian Barre syndrome

- New patient to our system with hx of multiple admissions to treat episodes of irritability, agitation, tachycardia and marked spasticity. Multiple courses of antibiotics. Has had two documented positive urine cultures. Intermittent elevations of BP
- Admitted with increased posturing, fever to 41.2, concern for seizures and infection



- ALT 290 AST 159 Bili 0.7
- Blood, urine, CSF cultures neg.
- Other tests?



- CPK 8405 (upper normal 168)



- Treated with clonidine but BPs dropped
- Propranolol added and improved. Also given valium prn with marked improvement.
- Started on balcofen



Take Home message

- Severe spasticity with rigidity/muscle spasms can cause pain



Severe irritability/agitation in the severely neurologically impaired patient

- Patient who are subjects of this talk
 - Nonverbal
 - Non-ambulatory
 - Totally dependent on others for care
 - CC: persistent crying/screaming for no obvious reason
- Problem causes Frequent ED visits and sometimes admissions.



An approach to dealing with these patients

- Explain to parent that this is a common and challenging problem
- May take time to figure out
- Usually a satisfactory approach is found



- History and Physical Exam





- How long has pain/irritability been present?
- Any triggers or precipitating events?
- Does anything make crying better or worse?
- Was onset associated with any changes at home?
- Trauma?
- Changes in medications?
- Any other significant changes?
- Any signs of acute illness/infection?



Differential Diagnosis-looking for causes of pain

- Easily diagnosed conditions (ear infections, acute infections such as pneumonia, or other acute problem causing distress excluded)
- What follows is focused on problems to which neurologically impaired children are particularly susceptible
- Most common discussed first*, followed by other but equally important conditions listed in somewhat arbitrary order



Constipation



- Easily overlooked
- Overflow stools
- KUB is useful screen
- Constipation extremely common in non-ambulatory neurologically impaired patients
- Rx: polyethylene glycol/electrolyte solution, senna, bisacodyl, lactulose, Mg citrate.



Other GI causes

- Esophagitis
 - Reflux very common
 - Consider empirical antacids to eliminate pain from esophagitis
 - Can d/c later if no improvement
 - Gastric pH (can check in G tube)
 - Goal 4.5 over greater



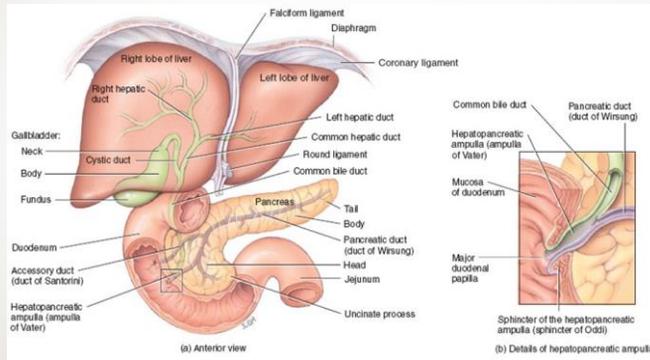
Visceral hyperalgesia

- pain associated with feeding
- If bloating, consider venting the G tube
- Gabapentin may be helpful
- Cyproheptadine if retching
 - Erythromycin



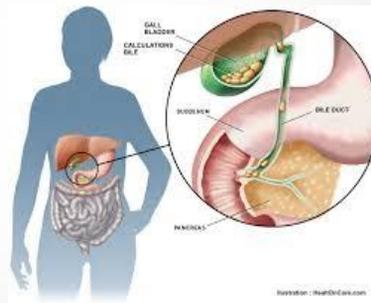
Pancreatitis

- Associated with valproic acid, gallstones
- Lipase, amylase



Gallstones

- Increased risk with tube feeding?
- Increased with hemolysis, hx TPN
- Elevated LFT's
- Abdominal Ultrasound





Fractures

- Osteopenia
 - Can occur after PT
 - Usually can detect on exam. Consider skeletal survey
 - Parents may be the best at detecting location



Hip Subluxation/dislocation

- Common in spastic cerebral palsy
- Does not always cause pain
- Subluxation more likely to cause pain
- Especially with abduction
 - Diaper changes
- AP pelvic radiographs





Kidney Stones

- Increased Risk due to calcium mobilization in non-ambulatory patient
- KUB screen
- U/A for blood
- CT scan
- Must have obstruction to be causing pain



Urinary retention

- Large volumes with infrequent urination
- Bladder distention
- May not be as painful as you would predict
- Catheterization should relieve pain





Muscle Spasms

- Spasticity itself not supposed to be painful
 - Adults with CP may disagree
- However, some patients become very rigid
- Elevated CPK can be a clue this can be a problem
 - Will elevate LFT's if present-can lead one to think that liver problem is present
- Rx: balcofen, tizanidine
- Prn diazepam (preferred over lorazepam)



Paroxysmal Sympathetic Hyperactivity (PSH) a.k.a. autonomic storming





Paroxysmal Sympathetic Hyperactivity a.k.a autonomic storming

- Consider in children /adolescent with severe brain injury
- Especially
 - HIE
 - TBI
- More common early and usually becomes less of a problem over time.
- Look for causes of pain as triggers (no different than items we are discussing)



Paroxysmal Sympathetic Hyperactivity a.k.a autonomic storming

- Severe agitation, discomfort
- Tachycardia
- Hypertension
- Fever
- Often severe muscle spasm
- Sweating



Paroxysmal Sympathetic Hyperactivity a.k.a autonomic storming

- Can be confused with sepsis
 - Acute phase reactants not markedly elevated
 - Fever (often very high) closely or clearly associated with obvious episodes of agitation, storming
 - Fever goes down when storming episode resolves



Paroxysmal Sympathetic Hyperactivity a.k.a autonomic storming

- Treatment: (limited evidence base)
- Select daily controller meds
- Use Prn meds
- Don't forget nonpharmacological treatment
 - Cuddling, repositioning, weighted blankets, massage, vibration



Paroxysmal Sympathetic Hyperactivity a.k.a autonomic storming

- **Daily meds**
 - If rigidity, severe muscle spasms, very high CPK
 - Baclofen
 - If primarily excess sympathetic symptoms
 - Clonidine or beta blockers (propranolol)
 - Gabapentin good to add to the mix
 - Bromocriptine has been used
- **Prn meds**
 - Morphine
 - Benzodiazepines
 - Diazepam
 - Clonazepam



Adverse Reactions to Medications or medication withdrawal



- Kevetiracetam (Keppra) may cause behavior change
- Antihistamines
- Serotonin syndrome
 - SSRI's
- Neuroleptic malignant syndrome
 - Dopamine antagonists
 - Haloperidol, risperidone, olanzepine, aripiprazole, metoclopramide, withdrawal of levodopa
- Withdrawal from baclofen, narcotics, benzodiazepines.



Other important but less common causes of pain to consider

- Ulcers, gastritis
- Debutitus ulcers
 - Often not as painful as one would think
- Ocular
 - Corneal abraision
- Dental
 - Reduced risk if no carbohydrates by mouth
- Environmental change
 - Family change, move, new nurse, etc
- Bacterial overgrowth
 - Abd distension, increased gas (may be seen on KUB)



Other important but less common causes of pain to consider(cont.)

- Adrenal dysfunction
 - Unexplained hypoglycemia, unexpected hypotension with surgical procedures or infections
- Seizures: unusual as cause of unexplained irritability
- Migraine headaches
 - Periodic, positive FH
- Increased ICP (if shunt)
- Post procedure
 - Osteotomies for dislocated hips
 - Post fundoplication



Idiopathic

- No cause can be found
- OK to treat empirically:
 - Gabapentin first line
 - Nortriptyline, amitriptyline, SNRI's.
 - Trial and error often required



Initial lab workup:

- CBC, CRP: screen for infection
- Urinalysis: check for UTI (WBC, nitrites), stones (blood).
 - Asymptomatic bacteriuria common and not a cause
- Comprehensive metabolic screen (liver disease, gallstones, dehydration)
- Lipase, amylase
- Abdominal radiograph (constipation, plus gallstones; renal stones if seen)
- Consider CPK if severe spasticity or rigidity, posturing
- Abdominal ultrasound if clinical suspicion (CT scan more sensitive for renal stones)



Summary

- Basic Hx and Physical looking for clues
- Basic lab work up as above
- Explanation to parents that problem is common but may take some time to figure out and treat successfully.
- Treat causes as found or suspected
- OK to treat even if no diagnosis found
- If storming:
 - Treat with daily meds (gabapentin, baclofen, clonidine, propranolol) +
 - Prn meds for flare ups: benzo's , narcotics
 - Non medical Rx



Handouts of Differential Diagnosis

- Differential Diagnosis
- Suggested initial work up



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