CALMING THE STORMS: COPING WITH COMPLICATIONS

Shih-Ning Liaw, MD

Glenda McDonald, MDiv, MS

February 21, 2014

UTHealth
The University of Texas

Texas PEDIATRIC Palliative Care Consortium

Children's Memorial Hermann Hospital

OBJECTIVES

- To identify common sources of suffering when families are faced with unintended complications
- To develop strategies for the treatment of irritability in a neurologically impaired child
- To discuss interdisciplinary modalities to provide support for family members with differing hopes and goals of care

JOHNNY'S SERIES OF UNFORTUNATE EVENTS

- 13 month old boy with complex congenital heart disease
  - Term infant with cardiac disease identified prenatally
  - Double outlet right ventricle, transposition of the great arteries, ventricular septal defect, hypoplastic aortic arch
  - s/p 2 cardiac surgeries with uncomplicated post-operative courses
  - On routine aspirin prophylaxis
  - Making good developmental progress
- 2 weeks after second cardiac procedure, presented with left facial droop and left-sided weakness
  - Initial CT showed right middle cerebral artery infarct

- Overnight in PICU, developed seizures and right-sided weakness
  - Intubated
  - Antiepileptics started for seizures, confirmed by EEG
  - MRI showed large left middle cerebral artery infarct
  - Thrombus found on MRA
    - Stroke team recommended changing aspirin to clopidogrel
    - Cardiology recommended treating with both aspirin and clopidogrel
    - Hematology recommended enoxaparin

WHAT SHOULD THE PICU DO?

1. Follow stroke team recommendations (clopidogrel only)
2. Follow cardiology team recommendations (clopidogrel and aspirin)
3. Follow hematology team recommendations (enoxaparin only)
4. Request a family meeting with above consultants to discuss these treatment options

- Treated with aspirin and clopidogrel, while low-dose heparin was started due to presence of central venous catheters
- MRI showed hemorrhagic conversion of left infarct (hospital day #3)
  - Aspirin and heparin discontinued per stroke team recommendations
  - Also showed Chiari 1 malformation with stenosis of foramen magnum
  - Neurosurgery recommended to monitor clinically
- After diagnostic procedures re: Chiari malformation, extubated and transferred out of PICU (hospital day #17)
- Discharge planning initiated to discuss options for rehabilitation

- During this time, became increasingly more agitated and spastic
- Treated with acetaminophen, simethicone, and lorazepam prn
- After one week of increased irritability, scheduled diazepam was added by neurology with some relief
- PM&R recommended changing diazepam to baclofen
- Reluctance by family and health care team to use sedating medications; tendency to minimize symptoms
- Neurosurgery performed posterior fossa decompression (hospital day #24)
- Chronic And Palliative Services (CAPS) team consulted due to family distress (hospital day #30)

WHAT IS CONTRIBUTING MOST TO THE FAMILY'S DISTRESS OR SUFFERING?

1. Johnny's physical suffering (pain, irritability, medical complications)
2. Poor communication amongst family members and health care team
3. Increasing despair and grief due to a catastrophic neurological prognosis
4. Bad hospital food

- Neurological status did not improve after decompression surgery
- Feeding intolerance, increased agitation, and respiratory distress
- Acutely decompensated and readmitted to PICU (hospital day #32)
- Found to have acute parainfluenza infection, placed on BiPAP
- Developed CSF leak from surgical incision site and underwent surgical repair (hospital day #36)
- CSF culture positive for Staphylococcus non-aureus and treated for meningitis
- Remained intubated for 10 days (hospital day #46)

- While intubated, remained on PICU drips for sedation
  - Fentanyl, midazolam, dexmedetomidine
- After extubation, IV drips changed to enteral formulations
  - Methadone, lorazepam, clonidine
- Worsening episodic agitation, posturing, tachycardia, and hypertension lasting up to 2 hours despite increases in dosing
  - Methadone, lorazepam, clonidine, baclofen, dantrolene
- Episodes escalated to unremitting opisthotonus
- Emergent brain imaging revealed massive hydrocephalus
- External ventricular drain placed (hospital day #56)

WHICH OF THE FOLLOWING IS THE GREATEST CONTRIBUTOR TO HIS NEUROLOGICAL STORMS?

1. Evolution of ischemic and hemorrhagic stroke
2. Opioid and benzodiazepine withdrawal
3. Constipation
4. Hydrocephalus

NEUROLOGICAL IRRITABILITY (ACUTE PHASE)

- Paroxysmal Autonomic Instability with Dystonia (PAID)
  - Intermittent, centrally mediated bursts of sympathetic activity
  - Agitation, diaphoresis, hyperthermia, hypertension, tachycardia, tachypnea, extensor posturing
  - Develops in acute phase of severe brain injury, lasting days to months
  - May be related to noxious trigger, so identify and treat inciting stimulus
  - Often no triggers identified
  - Aggressive symptom management for comfort
  - Other names
    - Autonomic dysfunction, dysautonomia
    - Neurostorming, sympathetic storms
- Differential diagnosis
  - Autonomic dysreflexia (spinal cord injury above T6)
  - Neuroleptic malignant syndrome

Blackman *et al*. Paroxysmal autonomic instability with dystonia after brain injury. *Arch Neurol*. 2004;61:321-28.

NEUROLOGICAL IRRITABILITY (CHRONIC PHASE)

- Persistent pain behaviors and/or agitation in children with neurologically impairment
- No nociceptive pain source identified or persistence of symptoms despite appropriate treatment of potential sources (constipation, spasticity, GERD)
- Suggested pathophysiology
  - Central nervous system
  - Peripheral nervous system (neuropathy)
  - Visceral nervous system (visceral hyperalgesia)

Hauer *et al*. Pediatric pain and symptom management guidelines. *Dana Farber Cancer Institute/Children's Hospital Boston*. 2011.

WHICH CLASS OF DRUGS IS THE MOST EFFECTIVE TREATMENT FOR NEUROLOGICAL IRRITABILITY?

1. Alpha agonists (clonidine, dexmedetomidine)
2. Benzodiazepines (lorazepam, clonazepam)
3. Beta blockers (propranolol, labetalol)
4. Dopamine agonists (bromocriptine, levodopa/carbidopa)
5. Muscle relaxants (baclofen, dantrolene)
6. Neuropathic agents (gabapentin, amitryptilene)
7. Opioids (morphine, methadone)
8. I don't know

- After placement of EVD, his neurological storms lessened in severity but persisted as PICU attempted to wean down his sedation
- Because goal of PICU (and mom) was to wean off medication, he only received acetaminophen or ibuprofen for breakthrough events
- Significant family distress due to poor control of symptoms and breakdown in communication
- CAPS became more directly involved to provide symptom management, counseling, and coordination of care

PALLIATIVE CARE INTERVENTIONS

- Established deeper therapeutic relationship
  - "Tell us about Johnny before this hospitalization"
- Shifted goal from weaning off medications to providing comfort over a longer time frame
  - Recommended adding gabapentin, increasing methadone, changing lorazepam to diazepam, changing clonidine to patch
  - Recommended IV morphine and lorazepam for breakthrough
- Determined parents' understanding and goals of care
  - Both parents were good historians about major events
  - Mom still very hopeful for neurological recovery and viewed medications as a barrier
  - Dad very distressed about symptoms and wanted comfort

- Coordinated family meeting with PICU, Neurology, and PM&R
  - Reestablished lines of communication
  - Reviewed progression of imaging
    - Infarcts → encephalomalacia
    - Chiari 1 → myelomalacia
    - Intraventricular hemorrhage/debris → hydrocephalus
  - Demonstrated consensus in palliative treatment plan
  - Established goals for discharge home
    - Internalize ventricular shunt
    - Defer gastrostomy
    - Continue aggressive symptom management
  - Discussed code status and introduced hospice to provide in-home support
- EVD internalized, and Johnny did well through procedure
- Parents agreed to DNR (no cardiac code)

IN YOUR INSTITUTION, WHO IS IN THE BEST POSITION TO SUPPORT A FAMILY WITH DIVERGENT HOPES?

1. Chaplaincy
2. Child life
3. Social work
4. Palliative care team

HOSPICE COURSE

- Neurological storms resolved at home, but still had neuro-irritability
  - Able to wean off methadone, diazepam and gabapentin
  - Remained on baclofen, and clonidine
  - Mom was very proactive in weaning his medications
- Continued family struggles
  - "If dad had his way, he'd still be on all these medications and wouldn't be doing anything"
  - Excellent family support from grandparents
- Discharged from hospice and transitioned care to local PCP
- Goals to place gastrostomy, reestablish subspecialty care, and start outpatient therapies

SUMMARY OF COMPLICATIONS

- Neurological: 4 surgeries
  - Bilateral middle cerebral artery infarcts
  - Chiari 1 malformation with foramen magnum stenosis
  - Neurological storming/irritability
  - Wound dehiscence with CSF drainage (pseudomeningocele)
  - Encephalomalacia, myelomalacia
  - Intraventricular hemorrhage, chronic subdural hematomas
  - Hydrocephalus
- Hematological:
  - Left middle cerebral artery thrombus
  - Hemorrhagic conversion of stroke
  - Deep venous thrombosis of left internal jugular and subclavian veins
- Respiratory: intubated 5 times
- Infectious:
  - Parainfluenza infection
  - S. non-aureus meningitis