PALLIATIVE CARE CASE OF THE MONTH

"Neurosurgical intervention as palliation: Complex management of post-hypoxic myoclonus"

by
Molly Mack, MD MS
& Carol May RN, MSN, MBA, CHPPN

Volume 26. No. 153

Case: Case summary: A 17-year-old female suffered cardiac arrest secondary to anaphylaxis after accidental peanut ingestion resulting in severe hypoxic-ischemic encephalopathy (HIE) with bilateral thalamic and basal ganglia injury. Her post-arrest course required prolonged PICU admission and intubation following aspiration pneumonia. As a result of HIE, she developed spasticity complicated by bruxism and autonomic dysfunction. Symptoms of agitation required multiple pharmacologic agents during prolonged intubation. After transition to inpatient rehabilitation, storming and autonomic dysfunction were refractory to multiple agents.

About six weeks after injury, the patient developed mixed hyperkinetic movement disorder. Movement was described as multisegmented dystonia, semirhythmic upper extremity flexion posturing, and myoclonus. EEG captured during events did not show epileptiform correlate. Movements were exacerbated by pain. These movements progressed to new morphology with semirhythmic upper extremity movements consistent with intermittent dystonic posturing. These movements were very distressing to the patient as well as for family members and providers to witness. Multiple pharmacologic agents were trialed to manage symptoms, and diphenhydramine, carbidopa/levodopa, gabapentin, and fluoxetine were all discontinued after each failed to show a discernable benefit. Baclofen and dantrolene were helpful for the dystonic component but did not improve myoclonus. Clonidine, dexmedetomidine, and ketamine as needed were noted to provide some benefit. Levetiracetam initially led to a good response with decreased movement burden; however, the patient's symptoms recurred on high-dose therapy. Trihexyphenidyl was discontinued due to anticholinergic burden, and her symptoms were worse upon discontinuation. Continuous midazolam was found to be quite helpful, though the dose that was needed for a therapeutic effect led to deep sedation and ventilator dependence.

About 10 weeks after injury, the patient's symptoms progressed despite adequate medication trials, with worsening dystonic movements.

In consultation with neurology and neurosurgery, the decision was made to proceed with bilateral Globus Pallidus Internus Deep Brain Stimulation (GPi DBS). Post-implantation course was complicated by respiratory failure secondary to aspiration pneumonia and status myoclonus, requiring intubation. After the acute complications were resolved, DBS programming led to initial improvement in dystonia, and the patient was able to proceed to extubation. A post-extubation exam revealed mild residual upper extremity spasms, markedly improved from prior to device placement. The patient's current medication regimen with reasonably well controlled symptoms includes baclofen, clonazepam, dantrolene, clonidine, and methadone.

Discussion: Dystonia is recognized as a major source of suffering in children with life-limiting conditions [1]. In this case, the dystonic and myoclonic movements were a cause of severe distress for the patient and her family in addition to causing respiratory compromise and posing an aspiration risk. Given the devastating neurologic injury and altered level of consciousness, discerning the relationship to pain and triggers for exacerbation of movements was difficult. The patient had a decrease in movement symptoms and fewer outward signs of distress with the initiation of ketamine and methadone, both NMDA receptor antagonists. It has been suggested that modulating the central sensitization was effective beyond traditional analgesia.

This case highlights the framing of the use of DBS as a palliative measure. Early introduction of palliative care services in this patient's care allowed for the development of a longitudinal relationship and thoughtful goals of care. At a family meeting with multidisciplinary teams about eight weeks into her course, the static nature of her injury was discussed. This was new and difficult information for the family, who had had hopes of a meaningful recovery. Continued discussions at this time focused on optimization of pain and symptom management, and shift in strategy to manage her post-hypoxic myoclonus while working on weaning medications that had not been helpful.

Discussion (Continued)

Generally, DBS is presented as a therapeutic intervention with the goal of reducing dystonia severity and improving neurologic function. In this case, given the transition of goals with new prognostic information, proceeding with DBS placement was indicated as a palliative procedure to reduce symptom burden, improve comfort, and enhance quality of life rather than restore neurologic function. This was particularly important as the movements were distressing and improvements in contractures, spasms, and comfort were aligned with the patient's goals of care. This case also highlights the pharmacologic challenges in post-hypoxic movement disorders. With largely empiric evidence for treatment [2, 3], determining an appropriate medication regimen was difficult and led to periods of polypharmacy. At its peak, this patient required a midazolam drip alongside six scheduled agents (baclofen, dantrolene, gabapentin, carbidopa/levodopa, clonidine, Keppra) as well as multiple as-needed agents. This led to difficulties in balancing the goals to manage pain and relieve suffering against the risks of sedation and respiratory depression. Recent expert opinion in management of posthypoxic myoclonus recommends multimodal therapy – most commonly levetiracetam with clonazepam and/or valproic acid – noting that monotherapy is often insufficient [4].

Table 1: Medications used

Agent	Effect	Side effects/Notes
Trihexyphenidyl	Improvement in symptoms	Discontinued due to anticholinergic burden Dystonia worsened upon withdrawal
Diphenhydramine	No clear benefit as monotherapy	
Carbidopa-levodopa	No meaningful response	
Benzodiazepines	Most consistently helpful	Midazolam infusion at 10-15mg/hr provided most reliable relief but at the cost of deep sedation and ventilator dependence
Gabapentin	Helpful for dystonia No effect on myoclonus	
Dantrolene	Helpful for dystonia No effect on myoclonus	
Levetiracetam	Initial dramatic response with decreased movement burden	Symptoms returned on high-dose therapy (consistent with known limitations of levetiracetam monotherapy for post-hypoxic myoclonus
Clonidine and dexmedetomidine	Benefit consistent with known effects on dystonia and autonomic storming	
Ketamine	PRN doses helpful	
Fluoxetine	No discernable benefit	
Methadone		

References:

1. Mercante, A., et al., Towards new perspectives: International consensus guidance on dystonia in pediatric palliative care. Eur J Paediatr Neurol, 2025. 56: p. 24-37.
2. Lungu, C., et al., Defining research priorities in dystonia. Neurology, 2020. 94(12): p. 526-537.
3. Koy, A., et al., Advances in management of movement disorders in children. Lancet Neurol, 2016. 15(7): p. 719-735.
4. Romozzi, M., et al., Treatment approaches in posthypoxic myoclonus: A narrative review with expert opinion. Epilepsia, 2026. 67(3): p. 1049-1065.