PATIENT PRESENTATION

A 47-year-old Caucasian male with a past medical history of alcohol use disorder, polysubstance use disorder, major depressive disorder, bipolar disorder, and PTSD presented to the emergency department with cardiopulmonary arrest secondary to fentanyl overdose.

ACLS measures were initiated ROSC achieved after eight minutes.

Five days after resuscitation, the patient began shaking vigorously when attempting to sit or ambulate. The patient demonstrated action myoclonic jerks more pronounced in the lower extremities than the upper extremities on examination. The action myoclonus intensified the longer the patient sustained active contraction of the muscles in the lower extremities.

The patient also demonstrated truncal myoclonus impairing his ability to maintain upright posture without support.

Cognition and swallowing remained intact however he did demonstrate speech impairments including hyperkinetic dysarthria.

LABORATORY ANALYSIS

- Demonstrated electrolytes, liver studies, creatinine kinase, ammonia, cobalamin, blood counts, folate, mercury, arsenic and ceruloplasmin were all within normal limits.
- Imaging obtained including a head CT, brain MRI and full spine MRI were negative for acute pathology.
- EEG demonstrated background discontinuity and slowing correlating with moderate/severe encephalopathy; no seizure activity correlate was identified even during the episodes of observable jerking.
- Neurosurgery and Neurology ruled out any other potential neurological explanation and thus the diagnosis of Lance Adams Syndrome was made.

WORKUP

MANAGEMENT AND OUTCOMES

- Patient was admitted to inpatient rehabilitation for intensive multidisciplinary therapies.
- The patient was treated initially with propranolol with no significant improvement noted.
- He was then treated with clonazepam and levetiracetam with pronounced subjective and objective improvement in action myoclonus which translated into a large improvement in function.
- Patient improved with therapies in all modalities eventually reaching supervision level for all bed mobility, transfers, ambulation with rolling walker and activities of daily living.

DISCUSSION

- Lance-Adams Syndrome (LAS) is characterized by severe and debilitating action myoclonus presenting days to weeks following a global cerebral hypoxic event.
- The disease pathogenesis and treatment remain poorly understood with a scarcity of described cases despite being first discovered over seventy years ago in the 1960’s.
- There is a void of sufficient literature focusing on the definitive successful medical management, specifically cases that incorporate inpatient rehabilitation therapies.
- We describe for the first time a successful treatment of LAS in a middle-aged male. The patient’s action myoclonus was nearly silenced with combination therapy of levetiracetam and clonazepam alongside intensive integrative therapies in an inpatient rehabilitation (IPR) setting.
- It is important to recognize therapies as well as clonazepam and levetiracetam are important tools to rehabilitating a patient with Lance Adams Syndrome.

CONCLUSIONS

- LAS is a rare disease that manifests as action myoclonus days to weeks after a global cerebral hypoxic event.
- Studies focusing on inpatient integration of interdisciplinary therapies alongside medication management have yet to be properly investigated.
- Reduction of debilitating myoclonus with levetiracetam and clonazepam in LAS can drastically increase the patient’s functional outcome and improve quality of life.

REFERENCES