Figure legends

Figure 1: Importance of diagnostic criteria for diagnosing EMAS. MAS- myoclonic atonic seizures.

History suggestive of MAS was critical criterion. Recorded MAS, parent/home video suggestive of MAS, generalized spike wave discharges on inter-ictal EEG, normal neuroimaging, and normal development prior to seizure onset were strong criteria. The presence of other generalized seizures was a moderate criterion. Neither a family history of seizures or epilepsy, nor diffuse theta with centroparietal predominance were important.

Figure 2: Importance of exclusionary criteria for diagnosing EMAS.

There were no critical exclusionary criteria. Epileptic spasms, abnormal neuroimaging, focal abnormal neurologic exam, and onset prior to age 6 months or after age 6 years were strong exclusionary criteria. Developmental delay prior to seizure onset, low CSF glucose, focal seizures, tonic seizures, and presence of focal abnormalities on EEG were moderate exclusionary criteria. A prior history of febrile status epilepticus was not exclusionary.

Figure 3: Importance of recommended investigations.

Routine and/or prolonged EEG and MRI brain were essential evaluations, recommended by >80% of respondents for all patients. Serum quantitative amino acids, urine organic acids, fatty acid oxidation/acylcarnitine profiles, microarray, genetic panel, lactate/pyruvic acid, and CSF/serum glucose and lactate were recommended for all patients or those with atypical features by >80% of respondents. Specific genetic testing, whole exome sequencing, electrolytes, and additional metabolic testing were recommended for all patients or those with atypical features by 50-79% of respondents.

Figure 4: EMAS recommended treatments

Valproic acid was the only essential therapy (first or second treatment recommended by >70% of respondents). Topiramate, zonisamide, levetiracetam, benzodiazepines, and dietary therapies were beneficial therapies (recommended as first 6 possible therapies by >70%). Acetazolamide, ethosuximide, felbamate, lamotrigine, rufinamide, and vagus nerve stimulator were of indeterminate benefit (recommended as therapy at some point, but not first 6 by 70%). Tiagabine, phenytoin, oxcarbazepine, eslicarbazepine, and carbamazepine contraindicated.