Variables (General clinical)	**Finding through Delphi**	**Strength**
Time of diagnosis of EMAS	In the majority of patients EMAS is suspected by 6-12 months after first afebrile seizure	Strong
Clinical presentation		
Age at onset	EMAS is highly unlikely when age at first afebrile seizure is < 1 or > 6 years	Strong
Sex predisposition	Boys are more commonly affected than girls	Modest
Preceding development	• 25-30% patients have mild developmental delay preceding first afebrile seizure • Preceding moderate to severe delay is highly unlikely	• Strong • Strong
Prior febrile seizures	Less than half of patients have a history of febrile seizures	Strong
Family history	• A family history of febrile seizures is found in less than one quarter of cases • A family history of epilepsy is found in less than half of cases	• Modest • Modest
Seizure types		
Myoclonic atonic seizures	• Myoclonic atonic seizures are mandatory for diagnosis • Myoclonic atonic seizures usually appear in the first year after seizure onset	• Strong • Strong
Myoclonic seizures	• Myoclonic seizures are **NOT** mandatory for diagnosis • Myoclonic seizures are seen in the majority of patients • Myoclonic seizures usually appear in the first year after seizure onset	• No consensus • Strong • Strong
Generalized tonic clonic seizures	• Generalized tonic-clonic seizures are **NOT** mandatory for diagnosis • Generalized tonic-clonic seizures are seen in more than half of cases	• Strong • No consensus

	• Generalized tonic-clonic seizures usually appear within the first year after seizure onset	• Strong
Atypical absence seizures	• Atypical absence seizures are **NOT** mandatory for diagnosis • Atypical absences are seen in <50% of cases within the first year	• Strong • Modest
Atonic seizures	• Atonic seizures are **NOT** mandatory for diagnosis • Atonic seizures are seen in up to half of cases • When present, atonic seizures typically begin in the first year	• No consensus • Strong • Strong
Tonic seizures	• Tonic seizures are seen in a minority of patients with EMAS • When present, fewer than one quarter of patients have tonic seizures in the first year • Tonic seizures do not exclude a diagnosis of EMAS	• Strong • Modest • Strong
Non convulsive status epilepticus	• NCSE is seen in < 50% of cases • When present, NCSE usually is seen in the first year after seizure onset	• Strong • Strong
Stormy Phase		
Stormy Phase	• A stormy phase is seen in up to 50% patients • If present, the stormy phase usually starts within the first year, • In a minority stormy phase starts in the first 3 months after seizure onset • The stormy phase typically lasts less than 12 months • A stormy phase lasting >18 months should suggest an alternate diagnosis	• Strong • Strong • Strong • Strong • Modest
Development during active epilepsy period		
Development	• Developmental plateauing can be seen in up to 50% patients	• Strong • Strong

	• Plateauing can occur even without the occurrence of a stormy phase • Developmental regression can be seen in up to 50 % of patients in association with stormy phase	• Strong
Hyperactivity and behavior problems	• Hyperactivity and behavior problems can be seen in up to 25% • Hyperactivity and behavior problems can be seen in the absence of a stormy phase	• Strong • Strong
Ataxia	• Ataxia may be seen unrelated to stormy phase as it may be seen due to medications	• Modest
Factors that would lead to reconsideration of a diagnosis of EMAS		
This factor alone would lead to diagnosis reconsideration	No factors identified	NA
This factor may contribute to diagnosis reconsideration but only if other atypical features were also present	• Tonic seizures and vibratory tonic seizures • EEG showing generalized paroxysmal fast activity • Age at onset <2 years • Age at onset >6 years	• Strong • Modest • Strong • Strong
These factors would not have a significant impact on diagnosis reconsideration	• Greater number of NCSE episodes • Longer duration of NCSE episodes • Near-continuous generalized discharge on EEG • Slow spike-wave on EEG • Focal Spikes • Persistence of epilepsy beyond 48 months of age • Lack of response to the ketogenic diet	• No consensus • No consensus • No consensus • No consensus • No consensus • No consensus • No consensus
Reclassification to a diagnosis of LGS	Once initially diagnosed with EMAS, It is inappropriate to reclassify patients into the diagnosis of LGS because they have continued to have seizures > 48 months	• Modest consensus

Investigations in EMAS – at presentation		
EEG	An EEG should be performed at baseline	• Strong
Prolonged video EEG	• A prolonged VEEG should be performed to capture and confirm different seizure types • A prolonged VEEG should be performed to exclude features of LGS, if the diagnosis is in question	• Strong • Strong
Neuroimaging	• An MRI should be performed at baseline • PET is not indicated	• Strong • Strong
Testing for GLUT1DS	• GLUT1DS should be ruled out in all patients • An LP should be pursued if SLC2A1 gene testing is negative	• Strong • No consensus
Metabolic	A basic metabolic panel including serum amino acids, urine organic acids, lactate, pyruvate, chemistries should be performed	Modest
Genetic studies	• A karyotype is not needed except in select cases due to clinical concerns • A chromosomal microarray is not indicated at baseline except if other clinical concerns • An epilepsy gene panel should be performed • If the epilepsy gene panel is negative, WES should be considered in patients with EMAS phenotype especially those who are drug resistant	• Strong • Strong • Strong • Strong
Investigations during the course of EMAS		
EEG	Routine EEG should be performed to confirm seizure freedom	Strong
Prolonged EEG	• A prolonged VEEG should be performed to confirm seizure freedom • A prolonged VEEG should be performed if there is a suspicion for NCSE	• No consensus • Strong

Genetic studies	In drug resistant cases, a chromosomal microarray and whole exome sequencing should be considered	Strong
Neuropsychological testing Neuropsychological testing contd	• Should be obtained prior to school entry in all patients with suspected or known developmental delay • Should be obtained annually if delay is present	• Strong • Strong
Recommended therapies		
First tier medications	• Valproic acid • Clobazam • Clonazepam • Levetiracetam	• Strong • Strong • Modest • Modest
Second tier medications	Ethosuximide	Strong
Meds not either first or second tier	CBD is not first or second tier medication	Strong
Later medications	• Topiramate • Zonisamide • Lamotrigine • Perampanel • Rufinamide • Lacosamide • Felbamate	• Strong • Strong • Strong • Modest • Modest • No consensus • No consensus
Dietary therapy	A ketogenic diet should be considered after failure of a first line therapy	Strong
Contraindicated treatments	All sodium channel blockers except lamotrigine should be avoided (lamotrigine may be useful in some cases). Vigabatrin should be avoided	Modest Strong
Treatment of stormy course		
Recommended treatments	• Ketogenic diet • Valproic acid • Benzodiazepines	• Strong • Strong • Strong
Other medications during stormy phase	Steroids can be used in the stormy phase	Modest
Role of surgical therapies		
VNS	• VNS can be considered after failure of 4-5 ASM	Strong

	• VNS can be considered after failure of ketogenic diet in patients who remain drug resistant for more than one year	Strong
Corpus callosotomy	Corpus callosotomy should only be considered after if patients have been drug-resistant for longer than one year, have failed 4-5 ASM and the ketogenic diet, and have frequent drop seizures	Strong
Remission of EMAS		
EMAS remission	• Remission occurs in at least 50% of patients • Of those who remit, the majority will remit by 24 months • Patients who continue to have seizures 5 years after the first afebrile seizure are highly unlikely to achieve remission.	• Strong • No consensus • Strong
Long-term developmental outcomes		
Development	• In patients who experience complete remission, more than 50% will be developmentally normal at follow up • Learning disorder without ID is seen in at least 25% who attain complete remission • Less than one quarter of patients who achieve complete remission will be left with moderate or greater intellectual disability • Mild to moderate intellectual disability is seen in the majority of patients who do not have complete remission • Severe intellectual disability is rare, even in those without remission • Drug-resistant EMAS patients who do not remit have more favorable cognitive outcomes than those with Lennox-Gastaut syndrome	• Strong • Strong • Strong • Strong • Strong • Strong
Prognostic factors		
Seizure types	• Tonic seizures and vibratory tonic seizures are at least moderately predictive of poor seizure outcome • Greater numbers of GTCS in the first 2 years are predictive or poorer outcome	• Strong • No consensus • No consensus

	• High frequency of drop seizures is predictive of poorer outcome	
Non convulsive status epilepticus (NCSE)	• Longer duration of NCSE is at least moderately predictive or poor seizure outcome • Greater number of NCSE is at least mildly to moderately predictive of poor seizure outcome	• Modest • Modest
EEG	• Paroxysmal fast activity on EEG is mildly predictive of poor seizure outcome • Focal spikes are predictive of poorer outcome • Slow spike-wave discharge is predictive of poorer outcome	• Strong • No consensus • No consensus
Age at onset	• Younger (<1 year) at seizure onset is predictive of poorer outcome • Older (>6 years) age at seizure onset is predictive of poorer outcome	• Modest consensus • No consensus
Family history	• Family history of epilepsy is not predictive of outcome	• No consensus

Sodium channel blockers asked about were phenobarbital, phenytoin, carbamazepine, Oxcarbazepine);

VNS: Vagus nerve stimulator; ASM: anti-seizure medication