Doose Delphi Round 3

Please complete the survey below.

Thank you!

The following statements are made on basis of results of rounds 1 and 2. We have just a few more questions to clarify CLASSIFICATION, INVESTIGATION, PROGNOSIS AND TREATEMENT .Pls answer all questions. Follow up questions will come up depending on your answers- please write in answers for all subsequent questions.

Questions regarding terminology and alternate diagnosis:

	Strongly Agree	Agree	Neutral	Disagree	Strongly Disagree
For the subset of patients who do not become seizure free and have residual seizures after 4-5 years- I believe it is appropriate to reclassify them as having LGS phenotype.	◯	◯	◯	◯	◯

"Reason: For the subset of patients who do not become seizure free and have residual seizures after 4-5 years- I believe it is appropriate to reclassify them as having LGS phenotype.
"

_____

The next 3 questions have to do with reasons to reclassify this subset of drug resistant EMAS: as LGS include

	Strongly Agree	Agree	Neutral	Disagree	Strongly Disagree
Reason to reclassify a subset of drug resistant EMAS as LGS includes -eligibility for LGS-Approved Drugs/Treatments	◯	◯	◯	◯	◯
Reason to reclassify a subset of drug resistant EMAS as LGS includes -eligibility for DRUG TRIALS aimed at LGS patients	◯	◯	◯	◯	◯
Reason to reclassify a subset of drug resistant EMAS as LGS includes -providing MORE DEFINED EXPECTATIONS REGARDING DRUG RESISTANCE AND COGNITIVE DELAYS- FOR FAMILY	◯	◯	◯	◯	◯

Reason: Reason to reclassify a subset of drug resistant EMAS as LGS includes -eligibility for LGS-approved drugs/treatments _____

Reason: Reason to reclassify a subset of drug resistant EMAS as LGS includes -eligibility for drug trials aimed at LGS patients _____

Reason: Reason to reclassify a subset of drug resistant EMAS as LGS includes -providing more defined expectations _____

The next two questions have to do with outcomes related to classification:

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
I believe that most drug resistant EMAS patients have a better seizure outcome than most LGS patients?	○	○	○	○	○
I believe that most drug-resistant EMAS patients have a better cognitive outcome than most LGS patients	○	○	○	○	○

Reason: I believe that most drug resistant EMAS patients have a better seizure outcome than most LGS patients? _____

Reason: I believe that most drug-resistant EMAS patients have a better cognitive outcome than most LGS patients _____

Questions regarding investigations in EMAS:

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
I believe that basic metabolic testing should be performed in the majority of patients presenting with a phenotype of EMAS (serum amino acids, urine organic acids, lactate etc)	○	○	○	○	○
Either an epilepsy gene panel OR Whole Exome Sequencing should be strongly considered in ALL patients presenting with an EMAS phenotype	○	○	○	○	○

If the epilepsy gene panel is negative, I believe WES should be strongly considered in all cases of EMAS phenotype	○	○	○	○	○
If the epilepsy gene panel is negative, I believe WES should be strongly considered in a patient with EMAS phenotype who remains with Drug Resistant Epilepsy (DRE) for longer than 4-5 years?	○	○	○	○	○
A karyotype is NOT required for ALL patients presenting with an EMAS phenotype, but could be ordered selectively if other clinical concerns	○	○	○	○	○
Re CMA: Chromosomal microarray (CMA) should be strongly considered in the majority of patients with an EMAS phenotype	○	○	○	○	○
Re CMA: CMA is not indicated in most cases of with an EMAS phenotype but may be ordered selectively if other clinical concerns	○	○	○	○	○
Re CMA: CMA should be strongly considered in Cases with EMAS who remain with DRE for longer than 4-5 years.	○	○	○	○	○
I believe that GLUT1 should be excluded in a patient presenting with an EMAS phenotype	○	○	○	○	○
If SLC2A1 testing is normal, I would pursue an LP to exclude GLUT 1 in most cases with EMAS phenotype	○	○	○	○	○

Reason: I believe that basic metabolic testing should be performed in the majority of patients presenting with a phenotype of EMAS (serum amino acids, urine organic acids, lactate etc)	_____
Reason: Either an epilepsy gene panel OR Whole Exome Sequencing should be strongly considered in ALL patients presenting with an EMAS phenotype	_____
Reason: If the epilepsy gene panel is negative, I believe WES should be strongly considered in all cases of EMAS phenotype	_____

Reason: If the epilepsy gene panel is negative, I believe WES should be strongly considered in a patient with EMAS phenotype who remains with Drug Resistant Epilepsy (DRE) for longer than 4-5 years? _____

Reason: A karyotype is NOT required for ALL patients presenting with an EMAS phenotype, but could be ordered selectively if other clinical concerns _____

Reason: Chromosomal microarray (CMA) should be strongly considered in the majority of patients with an EMAS phenotype _____

Reason: CMA is not indicated in most cases of with an EMAS phenotype but may be ordered selectively if other clinical concerns _____

Reason: CMA should be strongly considered in Cases with EMAS who remain with DRE for longer than 4-5 years. _____

Reason: I believe that GLUT1 should be excluded in a patient presenting with an EMAS phenotype _____

Reason: If SLC2A1 testing is normal, I would pursue an LP to exclude GLUT 1 in most cases with EMAS phenotype _____

Regarding Neuropsychological testing:

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
I believe every EMAS patient should have baseline formal developmental/ cognitive assessment prior to starting kindergarten, if one has not been done in the recent past.	○	○	○	○	○
If a patient with EMAS has clinical concerns for developmental delay prior to kindergarten entry, early referral for assessment is recommended	○	○	○	○	○

Reason: I believe every EMAS patient should have baseline formal developmental/ cognitive assessment prior to staring kindergarten, if one has not been done in the recent past. _____

Reason: If a patient with EMAS has clinical concerns for developmental delay prior to kindergarten entry, early referral for assessment is recommended _____

In rounds 1 and 2 - panelists opined about video EEG- Regarding prolonged VEEG- we would like to parse out more specific questions

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
I usually perform prolonged VEEG in patients presenting with an EMAS phenotype to confirm seizure types and exclude features which may suggest LGS.	○	○	○	○	○
I usually perform a prolonged VEEG in a patient with EMAS in whom I suspect NCSE	○	○	○	○	○
I usually perform a prolonged VEEG in a patient with EMAS with unexplained developmental regression	○	○	○	○	○
I usually perform a prolonged VEEG in a patient with EMAS to confirm seizure freedom	○	○	○	○	○
I usually perform a prolonged VEEG in a patient with EMAS who develop a new spell type, if I am not sure it is a seizure, or not sure what type of seizure it is.	○	○	○	○	○

Reason: I usually perform prolonged VEEG in patients presenting with an EMAS phenotype to confirm seizure types and exclude features which may suggest LGS.	_____
Reason: I usually perform a prolonged VEEG in a patient with EMAS in whom I suspect NCSE	_____
Reason: I usually perform a prolonged VEEG in a patient with EMAS with unexplained developmental regression	_____
Reason: I usually perform a prolonged VEEG in a patient with EMAS to confirm seizure freedom	_____
Reason: I usually perform a prolonged VEEG in a patient with EMAS who develop a new spell type, if I am not sure it is a seizure, or not sure what type of seizure it is.	_____

In rounds 1 and 2, we have consensus about tier 1 treatment and treatment of stormy phase- but want to know about tier 2 / other antiseizure meds (ASM) :
Questions regarding treatment: ASM
In a patient with EMAS WHO HAS FAILED THERAPY with VALPROIC ACID, CLOBAZAM,LEVETIRACETAM,ETHOSUXIMIDE AND KETOGENIC DIET ,the following agents are reasonable next therapeutic options.

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
rufinamide	◯	◯	◯	◯	◯
topiramate	◯	◯	◯	◯	◯
lamotrigine	◯	◯	◯	◯	◯
perampanel	◯	◯	◯	◯	◯
felbamate	◯	◯	◯	◯	◯
zonisamide	◯	◯	◯	◯	◯
lacosamide	◯	◯	◯	◯	◯

Reason: rufinamide

_____

Reason: topiramate

_____

Reason: lamotrigine

_____

Reason: perampanel

_____

Reason: felbamate

_____

Reason: zonisamide

_____

Reason: lacosamide

_____

Questions regarding treatment: VNS

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree

VNS is a reasonable consideration in a patient with EMAS who has failed multiple (>4-5) ASMs (including valproic acid, clobazam, levetiracetam, as well as 1-2 other agents) and the KD, and whose epilepsy has remained drug resistant for at least one year or longer.	○	○	○	○	○
I would consider VNS if a patient with EMAS has evolved to an LGS phenotype	○	○	○	○	○

Reason: VNS is a reasonable consideration in a patient with EMAS who has failed multiple (>4-5) ASMs (including valproic acid, clobazam, levetiracetam, as well as 1-2 other agents) and the KD, and whose epilepsy has remained drug resistant for at least one year or longer. _____

Reason:I would consider VNS if a patient with EMAS has evolved to an LGS phenotype _____

Questions regarding treatment: Corpus callosotomy (CC)

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
CC is a reasonable consideration in a patient with EMAS who has failed multiple (>4-5) ASMs (including valproic acid, clobazam, levetiracetam, as well as 1-2 other agents)and the ketogenic diet, whose epilepsy has remained drug resistant for more than one year, and who is having frequent drop seizures.	○	○	○	○	○
VNS should be strongly considered prior to CC	○	○	○	○	○
I would consider CC if a patient with EMAS has evolved to an LGS phenotype	○	○	○	○	○

Reason: CC is a reasonable consideration in a patient with EMAS who has failed multiple (>4-5) ASMs (including valproic acid, clobazam, levetiracetam, as well as 1-2 other agents)and the ketogenic diet, whose epilepsy has remained drug resistant for more than one year, and who is having frequent drop seizures. _____

Reason: VNS should be strongly considered prior to CC

_____

Reason: I would consider CC if a patient with EMAS has evolved to an LGS phenotype

_____