Doose Delphi Round 2

Please complete the survey below.

Thank you!

The following statements are made on basis of results of round 1. We would like to test strength of consensus now. Pls answer all questions. Follow up questions will come up depending on your answers- please write in answers for all subsequent questions.
Clinical presentation (Age at presentation and prior history)

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
In the majority of patients with EMAS a diagnosis of EMAS is strongly suspected by 6-12 months AFTER the first afebrile seizure	○	○	○	○	○
EMAS is highly unlikely when the age at first afebrile seizure is < 12 months.	○	○	○	○	○
Diagnosis of EMAS is highly unlikely when the age at first afebrile seizure is >6 years.	○	○	○	○	○
A minority of patients with EMAS (up to 30%) could have suspected delay prior to seizure onset.	○	○	○	○	○
A minority of patients with EMAS (20-30%) could have mild developmental delay prior to the first seizure.	○	○	○	○	○
Moderate to severe developmental delay preceding the first seizure is highly unlikely in patients with EMAs and suggests alternative diagnoses.	○	○	○	○	○
Less than 50% patient s with EMAS have a personal history of prior febrile seizures .	○	○	○	○	○
A family history of febrile seizures is noted is < 25% cases of EMAS.	○	○	○	○	○
A family history of epilepsy is noted in < 50% cases with EMAS.	○	○	○	○	○

Reason: In the majority of patients with EMAS a diagnosis of EMAS is strongly suspected by 6-12 months AFTER the first afebrile seizure _____

Reason: EMAS is highly unlikely when the age at first afebrile seizure is < 12 months.	_____
Reason: Diagnosis of EMAS is highly unlikely when the age at first afebrile seizure is >6 years.	_____
Reason: A minority of patients with EMAS (up to 30%) could have suspected delay prior to seizure onset.	_____
Reason: A minority of patients with EMAS (20-30%) could have mild developmental delay prior to the first seizure.	_____
Reason: Moderate to severe developmental delay preceding the first seizure is highly unlikely in patients with EMAs and suggests alternative diagnoses.	_____
Reason: Less than 50% patient s with EMAS have a personal history of prior febrile seizures .	_____
Reason: A family history of febrile seizures is noted is < 25% cases of EMAS.	_____
Reason: A family history of epilepsy is noted in < 50% cases with EMAS.	_____

Clinical presentation (seizure types and when seen)

We would like to test strength of consensus now based on your previous answers regarding seizure types and when they are seen. Pls answer all questions. Follow up questions will come up depending on your answers- please write in answers for all subsequent questions.

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
Myoclonic seizures are NOT mandatory for a diagnosis of EMAS	○	○	○	○	○
Myoclonic seizures are seen in a majority of cases with EMAS	○	○	○	○	○
Myoclonic seizures most commonly present within the first year after the first afebrile seizure	○	○	○	○	○
Atypical absence seizures are NOT mandatory for a diagnosis of EMAS	○	○	○	○	○
Atypical absence seizures are seen in 10-25 % cases with EMAS	○	○	○	○	○

Of patients with EMAS who have atypical absences, these are seen in a minority of patients within the first year after the first afebrile seizure.	○	○	○	○	○
Generalized tonic clonic seizures are NOT mandatory for a diagnosis of EMAS	○	○	○	○	○
Generalized tonic clonic seizures are seen in less than half of cases with EMAS	○	○	○	○	○
When present, generalized tonic clonic seizures most commonly begin within the first year after the first afebrile seizure	○	○	○	○	○
Atonic seizures are NOT mandatory for a diagnosis of EMAS	○	○	○	○	○
Atonic seizures are seen in upto 50% of cases with EMAS	○	○	○	○	○
When present, atonic seizures most commonly begin within the first year	○	○	○	○	○
NCSE is seen in a minority of patients with EMAS (less than 50%)	○	○	○	○	○
NCSE, when present, usually starts within 12 months after first afebrile seizure.	○	○	○	○	○
Tonic seizures are seen in a minority of patients with EMAS	○	○	○	○	○
Tonic seizures are seen in < 25% cases within first year .	○	○	○	○	○
Tonic seizures do not exclude diagnosis of EMAS	○	○	○	○	○

Reason: Myoclonic seizures are NOT mandatory for a diagnosis of EMAS	_____
Reason: Myoclonic seizures are seen in a majority of cases with EMAS	_____
Reason: Myoclonic seizures most commonly present within the first year after the first afebrile seizure	_____
Reason: Atypical absence seizures are NOT mandatory for a diagnosis of EMAS	_____

Reason: Atypical absence seizures are seen in 10-25 % cases with EMAS	_____
Reason: Of patients with EMAS who have atypical absences, these are seen in a minority of patients within the first year after the first afebrile seizure.	_____
Reason: Generalized tonic clonic seizures are NOT mandatory for a diagnosis of EMAS	_____
Reason: Generalized tonic clonic seizures are seen in less than half of cases with EMAS	_____
Reason: When present, generalized tonic clonic seizures most commonly begin within the first year after the first afebrile seizure	_____
Reason: Atonic seizures are NOT mandatory for a diagnosis of EMAS	_____
Reason: Atonic seizures are seen in upto 50% of cases with EMAS	_____
Reason: When present, atonic seizures most commonly begin within the first year	_____
Reason: NCSE is seen in a minority of patients with EMAS (less than 50%)	_____
Reason: NCSE, when present, usually starts within 12 months after first afebrile seizure.	_____
Reason: Tonic seizures are seen in a minority of patients with EMAS	_____
Reason: Tonic seizures are seen in < 25% cases within first year .	_____
Reason: Tonic seizures do not exclude diagnosis of EMAS	_____

Questions related to the STORMY COURSE. In round 1 we found some variability in the experts' opinion about what this is and we'd like to get some further details

Stormy course can be seen in up to 50% patients with EMAS	◯ Strongly agree ◯ Agree ◯ Neutral ◯ Disagree ◯ Strongly disagree
Reason: Stormy course can be seen in up to 50% patients with EMAS	_____

In patients who HAVE a stormy course:

	Yes	No
it usually begins within the first year after first afebrile seizure	○	○
a. It may begin in a minority of patients within 3 months of first afebrile seizure	○	○

In patients who HAVE a stormy course:

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
Stormy course typically lasts less than 12 months	○	○	○	○	○
Stormy course lasting > 18 months should suggest a diagnosis other than EMAS	○	○	○	○	○

Reason: Reason: Stormy course typically lasts less than 12 months _____

Reason: Reason: Stormy course lasting > 18 months should suggest a diagnosis other than EMAS _____

Questions related to course of EMAS- these questions have to do with patients you have already diagnosed with EMAS

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
Upto 50% patients with EMAS can have developmental plateauing	○	○	○	○	○
Developmental plateauing may be seen in patients with EMAS even without stormy course	○	○	○	○	○
Up to half the children with EMAS may develop regression during the course of EMAS	○	○	○	○	○
Hyperactivity and behavioral problems can be seen in at least 25% patients with EMAS	○	○	○	○	○
Hyperactivity and behavioral problems may be seen in patients with EMAS even without stormy course (strongly agree, agree, neutral, disagree, strongly disagree)	○	○	○	○	○

Ataxia will be seen in upto 50% patients with EMAS	○	○	○	○	○
Ataxia may be seen in patients with EMAS even without stormy course	○	○	○	○	○

Reason: Upto 50% patients with EMAS can have developmental plateauing _____

Reason: Developmental plateauing may be seen in patients with EMAS even without stormy course _____

Reason: Up to half the children with EMAS may develop regression during the course of EMAS _____

Reason: Hyperactivity and behavioral problems can be seen in at least 25% patients with EMAS _____

Reason: Hyperactivity and behavioral problems may be seen in patients with EMAS even without stormy course (strongly agree, agree, neutral, disagree, strongly disagree) _____

Reason: Ataxia will be seen in upto 50% patients with EMAS _____

Reason: Ataxia may be seen in patients with EMAS even without stormy course _____

Questions about remission (define remission as no seizures off meds)

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
Complete remission (ability to achieve seizure freedom off meds) occurs in at least 50% patients with EMAS	○	○	○	○	○
In cases who remit- the majority will do so within 24 months after the first afebrile seizure	○	○	○	○	○
Patients who continue to have seizures 5 years after the first afebrile seizure are highly unlikely to achieve remission	○	○	○	○	○
In children who experience complete remission, more than half will be developmentally normal at follow up	○	○	○	○	○

In children who experience complete remission, at least 25% will have a learning disorder without intellectual disability	○	○	○	○	○
In children who experience complete remission, < 25% will be left with moderate to severe intellectual disability	○	○	○	○	○
In children who DO NOT experience complete remission, the majority will have mild to moderate ID	○	○	○	○	○
It is rare to see severe ID even in children who DO NOT experience complete remission	○	○	○	○	○

Reason: Complete remission (ability to achieve seizure freedom off meds) occurs in at least 50% patients with EMAS	_____
Reason: In cases who remit- the majority will do so within 24 months after the first afebrile seizure	_____
Reason: Patients who continue to have seizures 5 years after the first afebrile seizure are highly unlikely to achieve remission	_____
Reason: In children who experience complete remission, more than half will be developmentally normal at follow up	_____
Reason: In children who experience complete remission, at least 25% will have a learning disorder without intellectual disability	_____
Reason: In children who experience complete remission, < 25% will be left with moderate to severe intellectual disability	_____
Reason: In children who DO NOT experience complete remission, the majority will have mild to moderate ID	_____
Reason: It is rare to see severe ID even in children who DO NOT experience complete remission	_____

Questions about prognostic factors and their influence (how predictive are these) on eventual outcome					
	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
Tonic seizures are at least moderately predictive of poor seizure outcome	○	○	○	○	○
Vibratory Tonic seizures are at least moderately predictive of poor seizure outcome	○	○	○	○	○
Number of NCSE episodes are at least mildly- moderately predictive of poor seizure outcome	○	○	○	○	○
Duration of NCSE is moderately to highly predictive of poor seizure outcome	○	○	○	○	○
Interictal EEG showing slow spike wave outside of stormy phase is mildly predictive of poor seizure outcome	○	○	○	○	○
Interictal EEG showing PFA is mildly predictive of poor seizure outcome	○	○	○	○	○
Focal spikes on EEG are mildly predictive of poor seizure outcome	○	○	○	○	○
High frequency of drops/ myoclonus is mildly predictive of poor seizure outcome	○	○	○	○	○
Generalized tonic clonic seizures in the first two years is mildly predictive of poor seizure outcome	○	○	○	○	○
Family history of epilepsy is mildly predictive of poor seizure outcome	○	○	○	○	○
Earlier age at seizure onset is mildly predictive of poor seizure outcome	○	○	○	○	○
Later age at onset is mildly predictive of poor seizure outcome	○	○	○	○	○

Reason: Tonic seizures are at least moderately predictive of poor seizure outcome _____

Reason: Vibratory Tonic seizures are at least moderately predictive of poor seizure outcome	_____
Reason: Number of NCSE episodes are at least mildly-moderately predictive of poor seizure outcome	_____
Reason: Duration of NCSE is moderately to highly predictive of poor seizure outcome	_____
Reason: Interictal EEG showing slow spike wave outside of stormy phase is mildly predictive of poor seizure outcome	_____
Reason: Interictal EEG showing PFA is mildly predictive of poor seizure outcome	_____
Reason: Focal spikes on EEG are mildly predictive of poor seizure outcome	_____
Reason: High frequency of drops/ myoclonus is mildly predictive of poor seizure outcome	_____
Reason: Generalized tonic clonic seizures in the first two years is mildly predictive of poor seizure outcome	_____
Reason: Family history of epilepsy is mildly predictive of poor seizure outcome	_____
Reason: Earlier age at seizure onset is mildly predictive of poor seizure outcome	_____

Questions that address diagnostic switching: factors that would lead to reconsidering the diagnosis of EMAS

Please indicate how impactful each of these factors is, in leading you to reconsider the diagnosis of EMAS

	High Impact - this factor alone, regardless of the rest of the clinical picture, would cause me to reconsider the diagnosis	Moderate impact - this factor may contribute to me reconsidering the diagnosis, but only if other atypical features were also present	Low Impact - this factor would have no impact on me reconsidering the diagnosis
Tonic seizures	○	○	○
Vibratory tonic seizures	○	○	○
Greater number of NCSE episodes	○	○	○
Longer duration of NCSE episodes	○	○	○
EEG showing frequent or near continuous irregular generalized spike wave activity	○	○	○

EEG showing slow spike wave activity	◯	◯	◯
EEG showing generalized paroxysmal fast activity:	◯	◯	◯
EEG showing generalized paroxysmal fast activity	◯	◯	◯
EEG showing focal spikes	◯	◯	◯
Younger age (< 2 years) at seizure onset	◯	◯	◯
Older age (>6 years) at seizure onset	◯	◯	◯
Persistence of epilepsy beyond:48 months	◯	◯	◯
Lack of response to ketogenic diet	◯	◯	◯

For ongoing care of patients with EMAS in clinic- what tests do you perform? When and Why? Following statements are designed based on consensus from round 1

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
You would order an EEG every 1 month	◯	◯	◯	◯	◯
You would order an EEG every 3 months	◯	◯	◯	◯	◯
You would order an EEG every 6 months	◯	◯	◯	◯	◯
You would order an EEG every 12 months	◯	◯	◯	◯	◯
You would order an EEG only if clinical concerns	◯	◯	◯	◯	◯

Reason: You would order an EEG every 1 month _____

Reason: You would order an EEG every 3 months _____

Reason: You would order an EEG every 6 months _____

Reason: You would order an EEG every 12 months _____

Reason: You would order an EEG only if clinical concerns _____

An EEG should be done to confirm remission if a child achieves seizure freedom?
◯ Yes
◯ No

If yes, how long after seizure freedom is achieved should an EEG be done (months)? _____

When should an overnight - prolonged video-EEG be done ? Please explain in terms of time after diagnosis and during course of treatment	_____
The reason to order a video EEG is: What are you looking for on this EEG?	_____
In what proportion of your EMAS patients do you order neuropsychological testing?	◯ always ◯ usually (>75-99%) ◯ sometimes (25-74%) ◯ rarely (1-24%) ◯ never
I would order neuropsych testing If delay is suspected.	◯ Yes ◯ No
How often should neuropsychological testing be performed?	◯ every 3 months ◯ every 6 months ◯ yearly

Questions related to treatment of EMAS: Following statements are based on consensus from round 1- we are now testing strength of that consensus.
Please answer all additional questions that pop up at the end based on your choices in for following questions.

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
All sodium channel blockers should be avoided in treatment of EMAS	◯	◯	◯	◯	◯
VPA would be your first choice to treat EMAS	◯	◯	◯	◯	◯
LEV can be considered a first tier antiseizure medicine to treat EMAS	◯	◯	◯	◯	◯
Benzos can be considered a first tier ASM to treat EMAS	◯	◯	◯	◯	◯
KD should be considered as second tier to treat EMAS	◯	◯	◯	◯	◯
ETX is considered can be considered as second tier for EMAS	◯	◯	◯	◯	◯
You would choose to use ETX only if frequent atypical absence or myoclonic seizures are noted	◯	◯	◯	◯	◯

You would choose to use ETX in EMAS irrespective of prominent seizure type	○	○	○	○	○

Reason: All sodium channel blockers should be avoided in treatment of EMAS _____

Reason: VPA would be your first choice to treat EMAS _____

Reason: LEV can be considered a first tier antiseizure medicine to treat EMAS _____

Reason: Benzos can be considered a first tier ASM to treat EMAS _____

Reason: KD should be considered as second tier to treat EMAS _____

Reason: ETX is considered can be considered as second tier for EMAS _____

Reason: You would choose to use ETX only if frequent atypical absence or myoclonic seizures are noted _____

Reason: You would choose to use ETX in EMAS irrespective of prominent seizure type _____

Following are once again based on consensus from round 1
In patients with EMAS during the stormy course when would you use each of the following treatments

	FIRST line	SECOND line	THIRD line or later	Would NOT use
Ketogenic diet	○	○	○	○
VPA bolus or increased VPA dose	○	○	○	○
Benzodiazepines	○	○	○	○
Steroids (ACTH/ Prednisone etc)	○	○	○	○

The next few questions related to VNS: Please DO NOT consider cost/ availability restrictions
Please answer all related questions as they pop up at the end based on your responses

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
VNS can be considered but only after failure of 3 ASMs	○	○	○	○	○
VNS can be considered but only after failure of 4 ASMs	○	○	○	○	○

VNS can be considered but only after failure of >5ASMs	○	○	○	○	○
The KD should be trialed prior to considering VNS	○	○	○	○	○
VNS should be considered as soon as intractability is present, regardless of epilepsy duration	○	○	○	○	○
VNS should only be considered if intractability has persisted longer than 1 year	○	○	○	○	○
VNS should only be considered if intractability longer than 2 years	○	○	○	○	○
VNS should only be considered if intractability has persisted longer than 3 years	○	○	○	○	○
I do not believe VNS is indicated for EMAS	○	○	○	○	○

Reason: VNS can be considered but only after failure of 3 ASMs	_____
Reason: VNS can be considered but only after failure of 4 ASMs	_____
Reason: VNS can be considered but only after failure of >5ASMs	_____
Reason: The KD should be trialed prior to considering VNS	_____
Reason: VNS should be considered as soon as intractability is present, regardless of epilepsy duration	_____
Reason: VNS should only be considered if intractability has persisted longer than 1 year	_____
Reason: VNS should only be considered if intractability longer than 2 years	_____
Reason: VNS should only be considered if intractability has persisted longer than 3 years	_____
Reason: I do not believe VNS is indicated for EMAS	_____

The next few questions related to Corpus callosotomy (CC) : Please DO NOT consider cost/ availability restrictions. Please answer all related questions as they pop up at the end based on your responses.

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
CC can be considered in an EMAS patient with drop seizures but only after failure of 3 ASMs	◯	◯	◯	◯	◯
CC can be considered in an EMAS patient with drop seizures but only after failure of 4 ASMs	◯	◯	◯	◯	◯
CC can be considered in an EMAS patient with drop seizures but only after failure of >5ASMs	◯	◯	◯	◯	◯
The KD should be trialed prior to considering CC	◯	◯	◯	◯	◯

Reason: CC can be considered in an EMAS patient with drop seizures but only after failure of 3 ASMs _____

Reason: CC can be considered in an EMAS patient with drop seizures but only after failure of 4 ASMs _____

Reason: CC can be considered in an EMAS patient with drop seizures but only after failure of >5ASMs _____

Reason: The KD should be trialed prior to considering CC _____

I would only offer CC irrespective of if drop seizures were problematic AND: Choose ONE BEST answer

- ◯ CC can be considered but only after failure of 3 ASMs
- ◯ CC can be considered but only after failure of 4 ASMs
- ◯ CC can be considered but only after failure of >5ASMs
- ◯ The KD should be trialed prior to considering CC

For DURATION of intractability: Choose ONE BEST answer

- ◯ CC should be considered as soon as intractability is present, regardless of epilepsy duration
- ◯ CC should only be considered if intractability has persisted longer than 1 year
- ◯ CC should only be considered if intractability longer than 2 years
- ◯ CC should only be considered if intractability has persisted longer than 3 years
- ◯ I would offer CC ONLY after VNS is tried first
- ◯ I do not believe that CC is indicated for EMAS