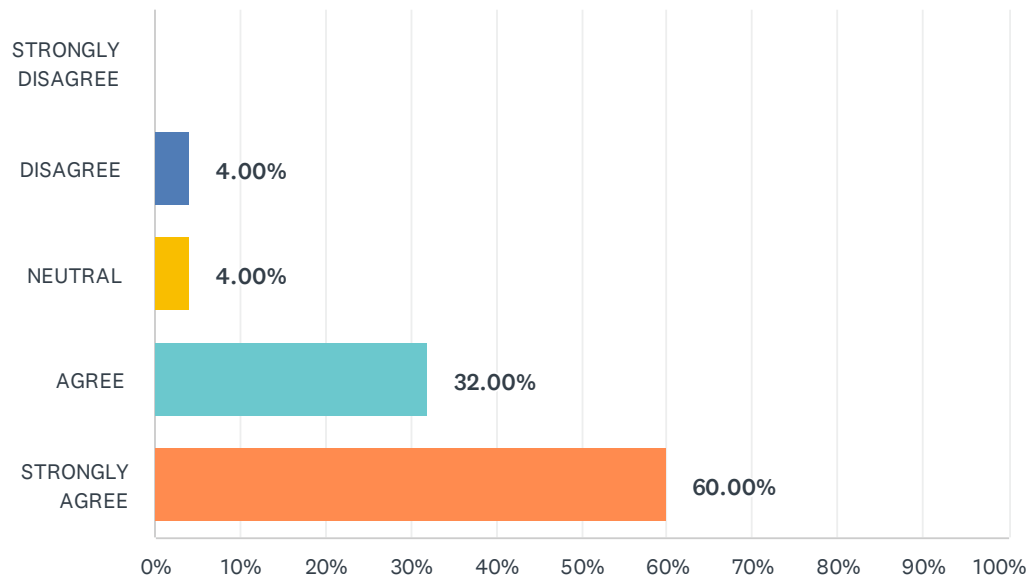


The acronym MELAS (Mitochondrial Encephalomyopathy, Lactic Acidosis, Stroke like episodes) is still useful but should be limited to the clinical syndrome and not to the m.3243A>G variant in MT-TL1 gene.

Answered: 25 Skipped: 0



**STRONG CONSENSUS**

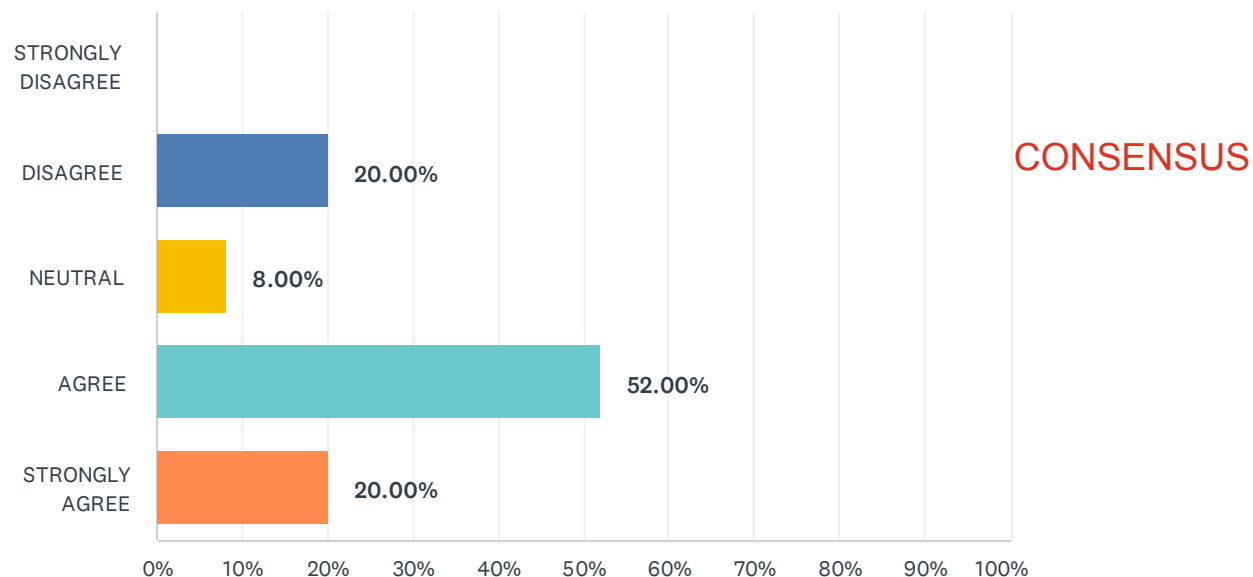
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	4.00%	1
NEUTRAL (3)	4.00%	1
AGREE (4)	32.00%	8
STRONGLY AGREE (5)	60.00%	15
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	5.00	4.48	0.75

The definition of MELAS syndrome proposed by NMDAC (Emmanuele et al. 2022, PMID: 35606253) is appropriate and should be endorsed:

“Mitochondrial Encephalomyopathy, Lactic Acidosis and Stroke-like episodes (MELAS): Both A and B. A) Stroke-like episodes (sudden-onset focal neurological deficit with brain MRI or CT showing a cerebral lesion that does not conform to a large vessel territory and typically affects cortex and adjacent white matter) B) Encephalomyopathy” (i.e. clinical myopathy).

Answered: 25 Skipped: 0

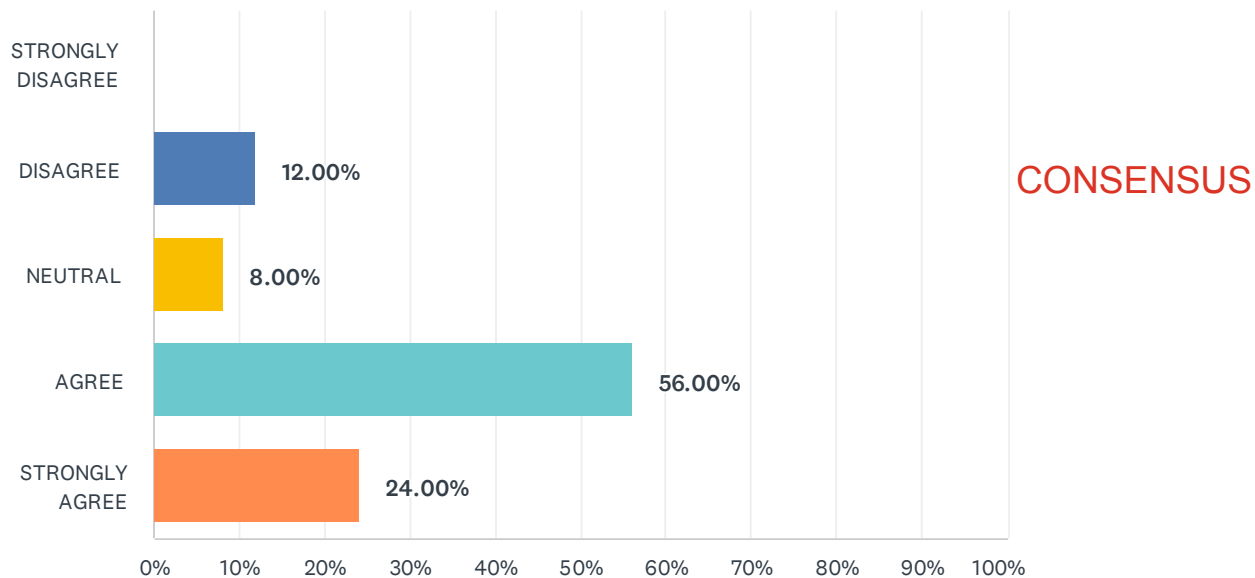


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	20.00%	5
NEUTRAL (3)	8.00%	2
AGREE (4)	52.00%	13
STRONGLY AGREE (5)	20.00%	5
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	3.72	1.00

The following definition of stroke-like episodes proposed by NMDAC (Emmanuele et al. 2022, PMID 35606253) is appropriate and should be endorsed: “sudden-onset focal neurological deficit with brain MRI or CT showing a cerebral lesion that does not conform to a large vessel territory and typically affects cortex and adjacent white matter”

Answered: 25 Skipped: 0

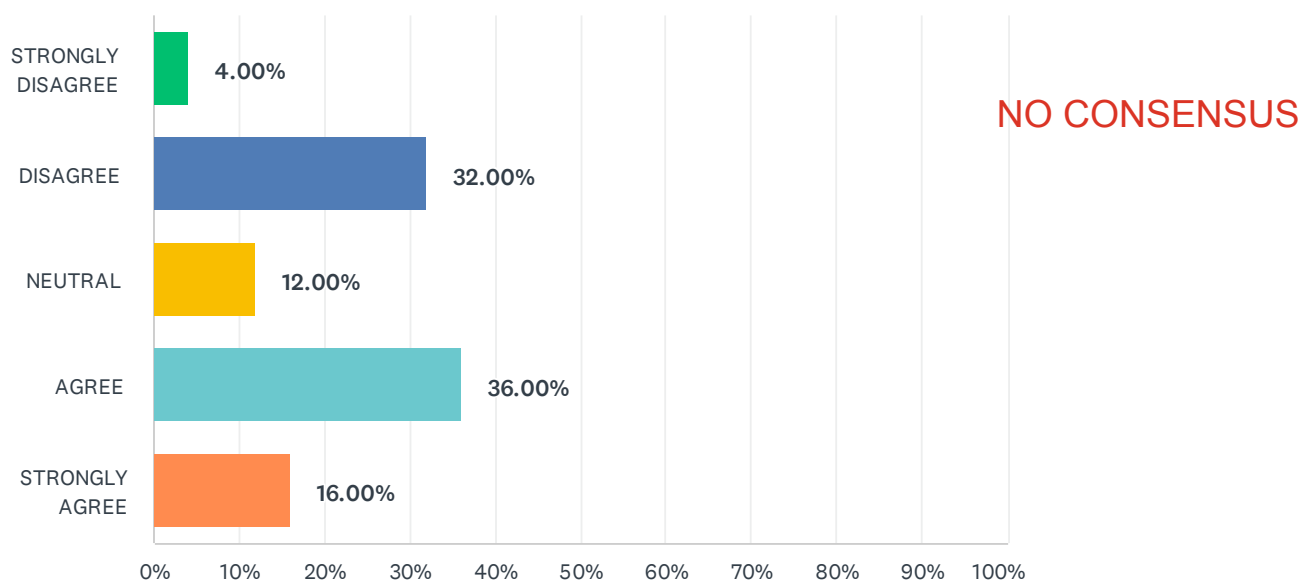


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	12.00%	3
NEUTRAL (3)	8.00%	2
AGREE (4)	56.00%	14
STRONGLY AGREE (5)	24.00%	6
TOTAL		25

BASIC STATISTICS				
Minimum 2.00	Maximum 5.00	Median 4.00	Mean 3.92	Standard Deviation 0.89

The following definition of stroke-like episodes proposed by the European consortium (Yi et al. 2019, PMID: 32090171) is appropriate and clinically relevant: “A mitochondrial stroke-like episode is a subacute, evolving brain syndrome driven by seizure activity in genetically determined mitochondrial disease. These potentially treatable encephalopathic episodes can present at any age with neurological and/or psychiatric symptoms typically associated with cortical/subcortical MRI changes and EEG abnormalities.”

Answered: 25 Skipped: 0

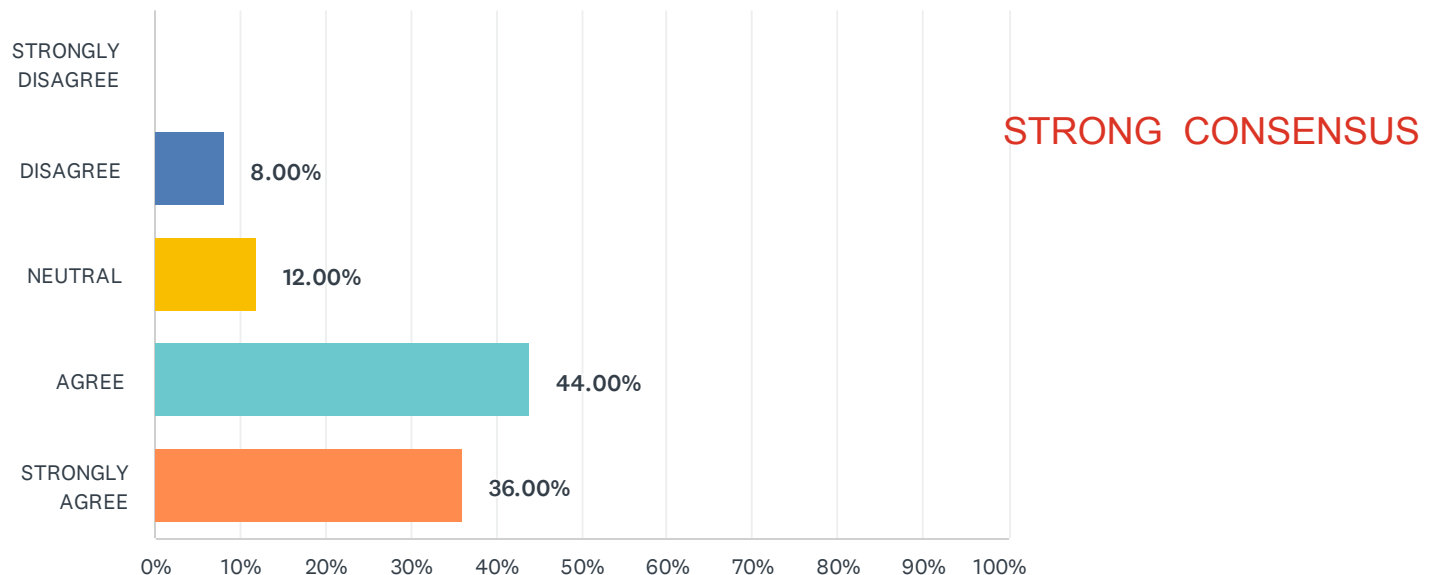


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	4.00%	1
DISAGREE (2)	32.00%	8
NEUTRAL (3)	12.00%	3
AGREE (4)	36.00%	9
STRONGLY AGREE (5)	16.00%	4
TOTAL		25

BASIC STATISTICS				
Minimum 1.00	Maximum 5.00	Median 4.00	Mean 3.28	Standard Deviation 1.18

The following modified (from NMDAC, PMID: 35606253, from the European consortium, PMID: 32090171 and from the latest Newcastle cohort study, PMID: 34927673) definition of stroke-like episode should be adopted: "A mitochondrial stroke-like episode is a acute/subacute, evolving brain syndrome that can manifest at any age with neurological and/or psychiatric symptoms typically occurring in association with cortical/subcortical MRI changes and/or pathological EEG abnormalities."

Answered: 25 Skipped: 0

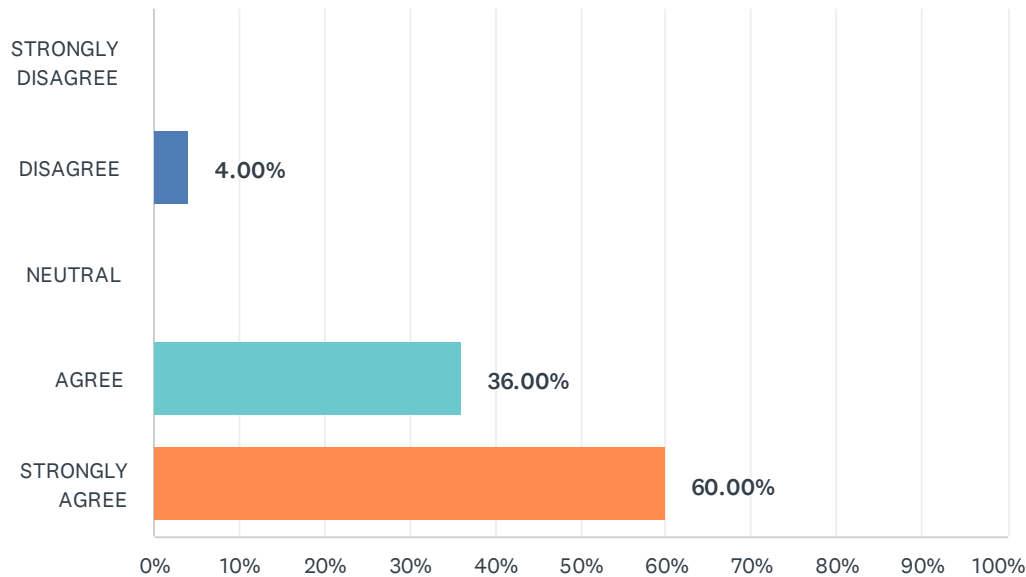


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	8.00%	2
NEUTRAL (3)	12.00%	3
AGREE (4)	44.00%	11
STRONGLY AGREE (5)	36.00%	9
TOTAL		25

BASIC STATISTICS				
Minimum 2.00	Maximum 5.00	Median 4.00	Mean 4.08	Standard Deviation 0.89

## Stroke-like episodes represent the core clinical feature required for the definition of MELAS syndrome.

Answered: 25 Skipped: 0



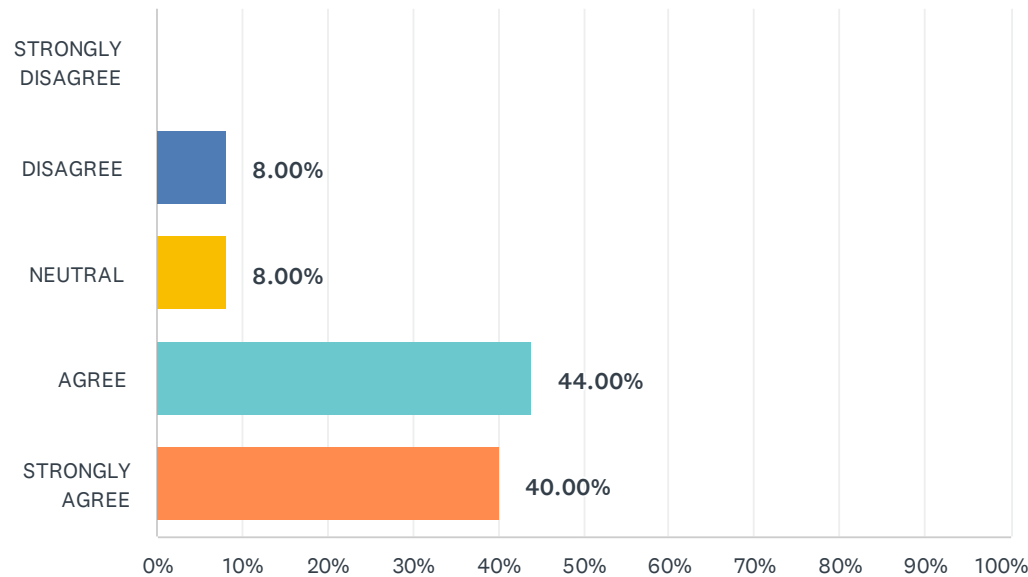
**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	4.00%	1
NEUTRAL (3)	0.00%	0
AGREE (4)	36.00%	9
STRONGLY AGREE (5)	60.00%	15
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	5.00	4.52	0.70

MELAS diagnosis without the occurrence of stroke-like episode, despite molecular confirmation of a pathogenic variant, is inappropriate.

Answered: 25 Skipped: 0



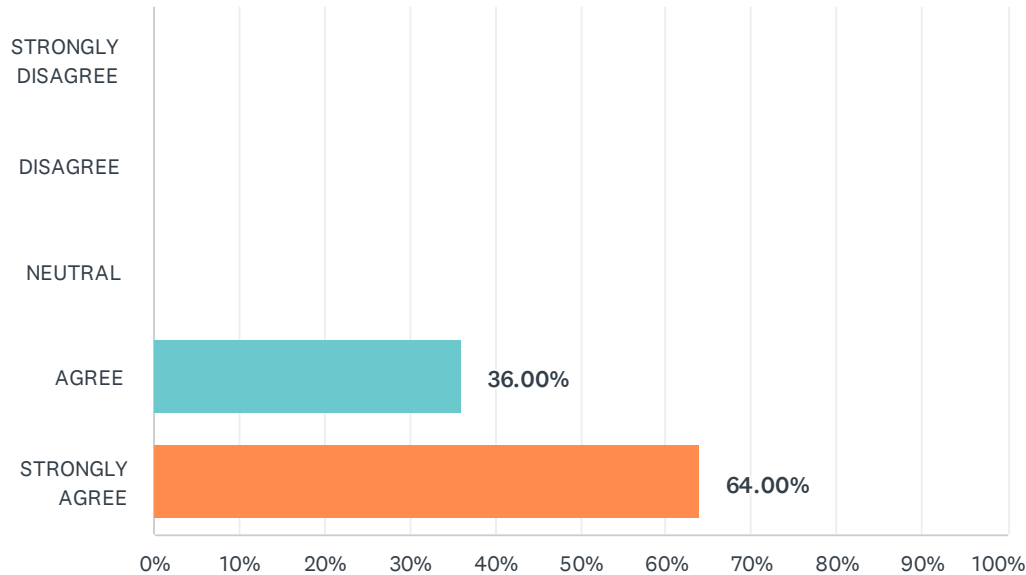
STRONG CONSENSUS

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	8.00%	2
NEUTRAL (3)	8.00%	2
AGREE (4)	44.00%	11
STRONGLY AGREE (5)	40.00%	10
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	4.16	0.88

## Stroke-like episodes can occur at any age, including late-onset cases (>40 years old).

Answered: 25 Skipped: 0



**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	36.00%	9
STRONGLY AGREE (5)	64.00%	16
TOTAL		25

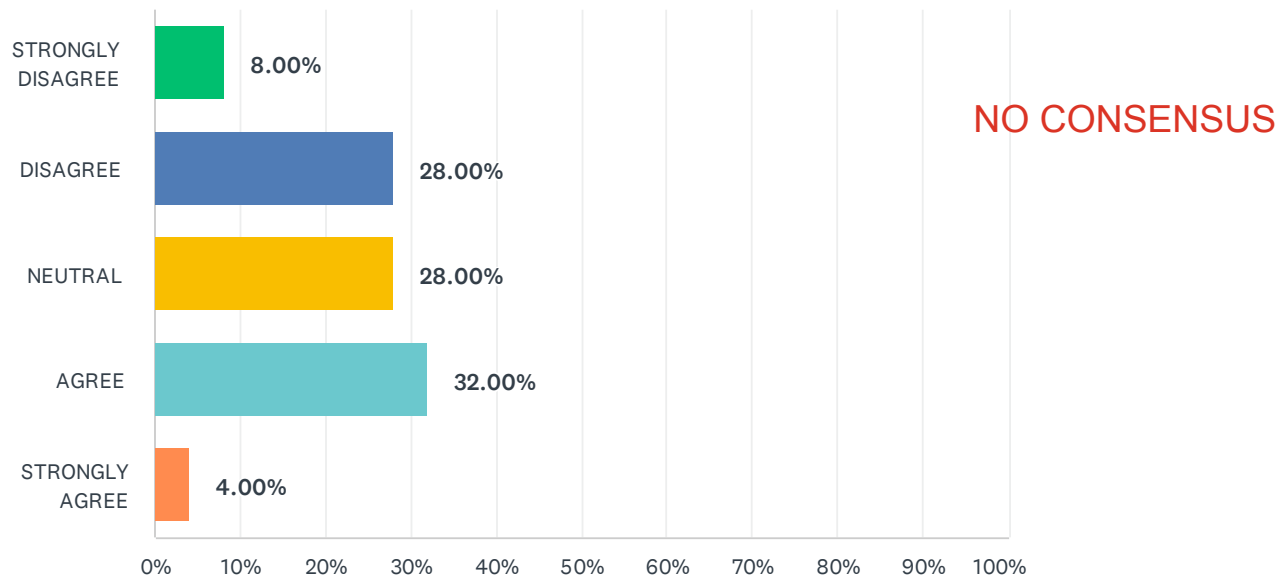
BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
4.00	5.00	5.00	4.64	0.48





The m.3243A>G variant in the MT-TL1 gene is associated with a specific clinical phenotype, distinguishable within the MELAS syndrome spectrum ("3243-related MELAS").

Answered: 25 Skipped: 0

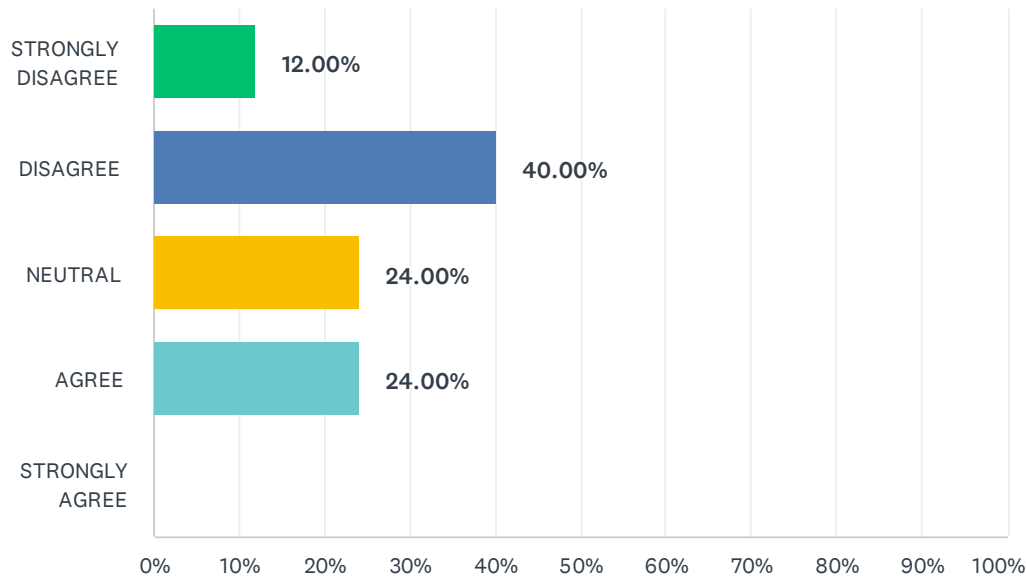


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	8.00%	2
DISAGREE (2)	28.00%	7
NEUTRAL (3)	28.00%	7
AGREE (4)	32.00%	8
STRONGLY AGREE (5)	4.00%	1
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	3.00	2.96	1.04

## Intravenous L-arginine should be administered in the acute phase of SLE.

Answered: 25 Skipped: 0



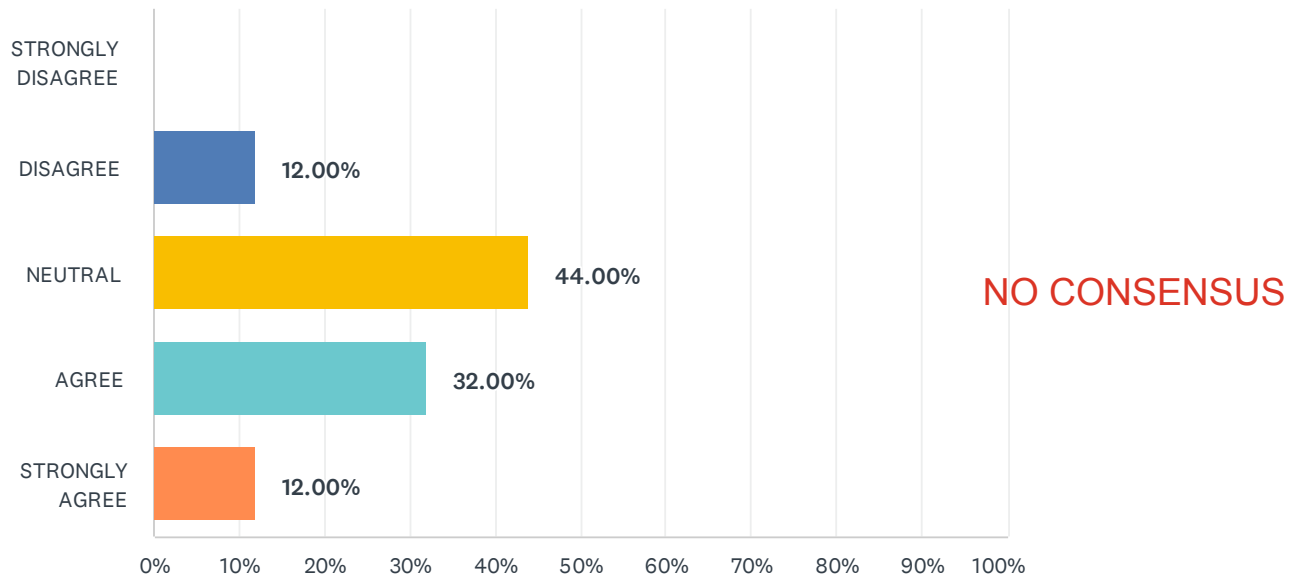
NO CONSENSUS

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	12.00%	3
DISAGREE (2)	40.00%	10
NEUTRAL (3)	24.00%	6
AGREE (4)	24.00%	6
STRONGLY AGREE (5)	0.00%	0
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	4.00	2.00	2.60	0.98

## Glucocorticoids should be used in the acute phase of SLE

Answered: 25 Skipped: 0

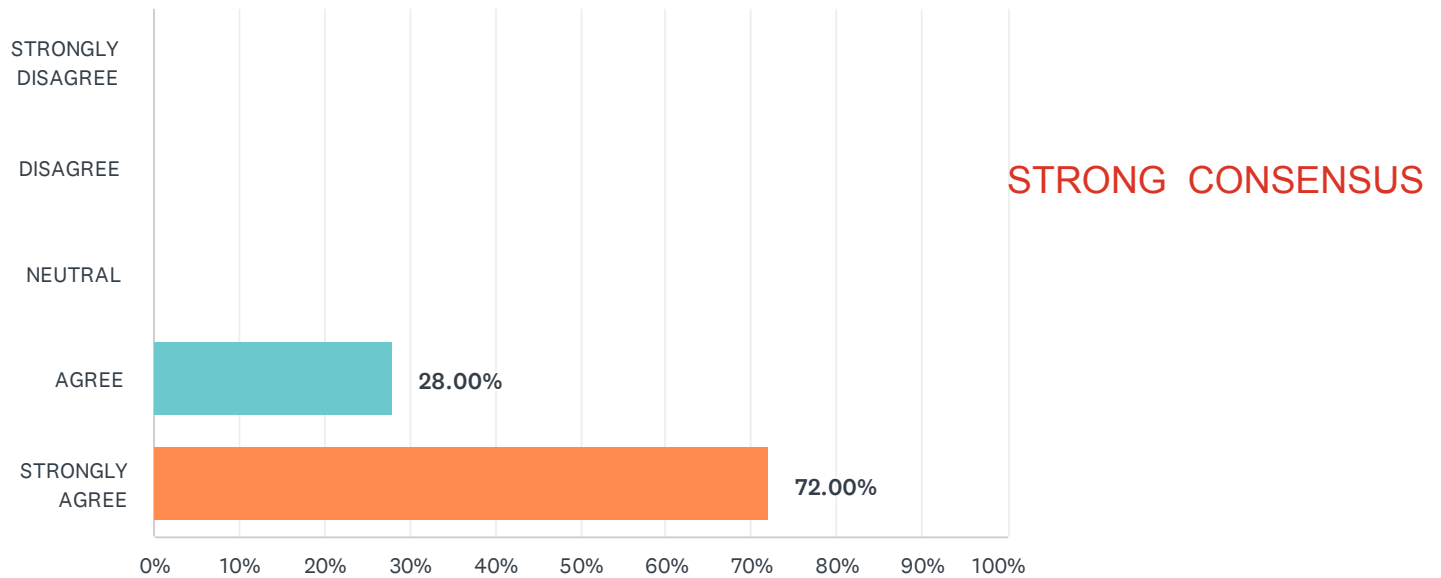


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	12.00%	3
NEUTRAL (3)	44.00%	11
AGREE (4)	32.00%	8
STRONGLY AGREE (5)	12.00%	3
TOTAL		25

BASIC STATISTICS				
Minimum 2.00	Maximum 5.00	Median 3.00	Mean 3.44	Standard Deviation 0.85

## EEG is an important early investigation of acute SLE

Answered: 25   Skipped: 0

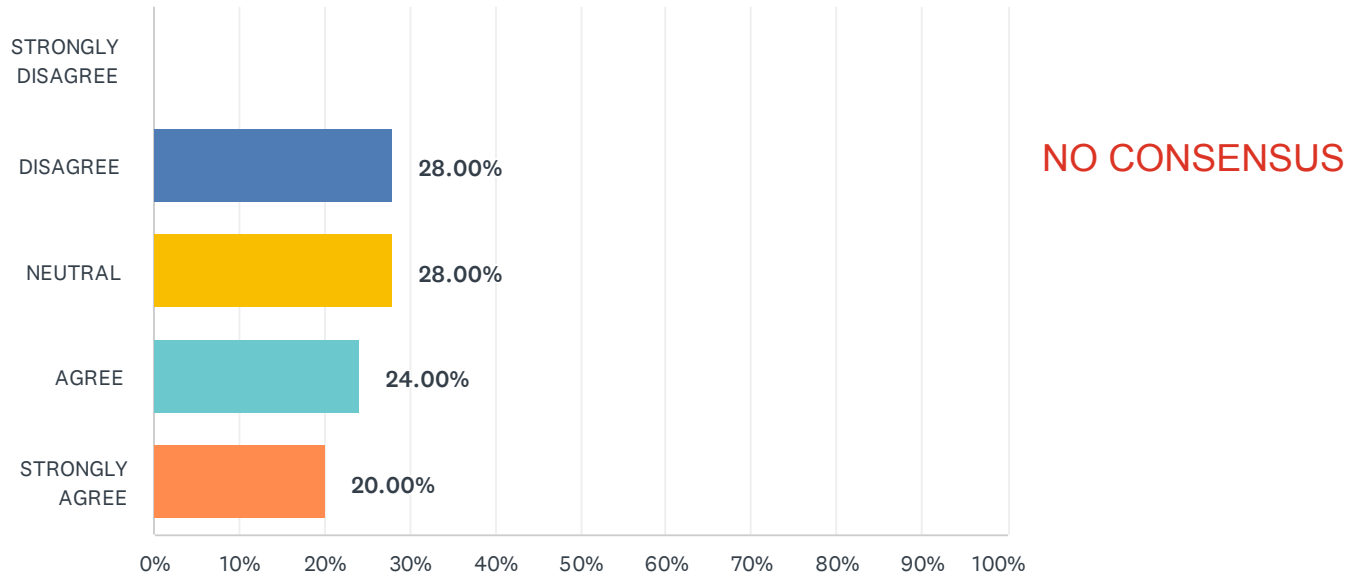


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	28.00%	7
STRONGLY AGREE (5)	72.00%	18
TOTAL		25

BASIC STATISTICS				
Minimum 4.00	Maximum 5.00	Median 5.00	Mean 4.72	Standard Deviation 0.45

## The administration of antiepileptic drugs should be routine management of an acute SLE irrespective of EEG appearance

Answered: 25 Skipped: 0



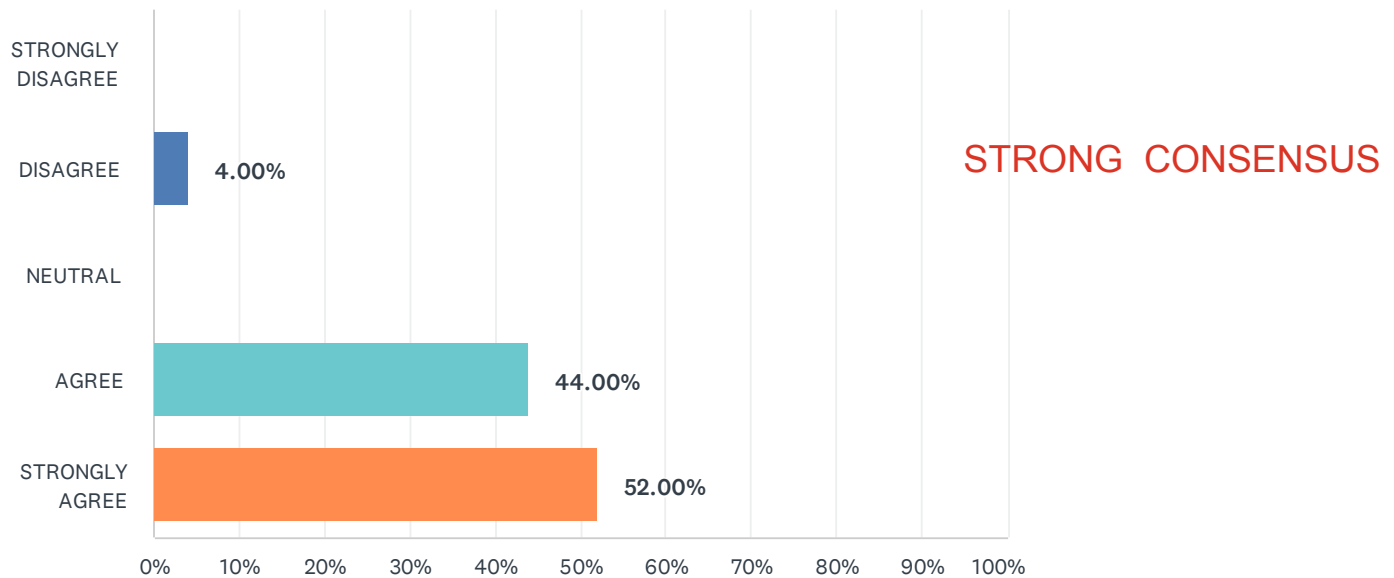
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	28.00%	7
NEUTRAL (3)	28.00%	7
AGREE (4)	24.00%	6
STRONGLY AGREE (5)	20.00%	5
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	3.00	3.36	1.09



## The consensus published in 2024 (PMID: 38576261) regarding the management of status epilepticus in association with acute SLE in PMD patients should be followed

Answered: 25 Skipped: 0



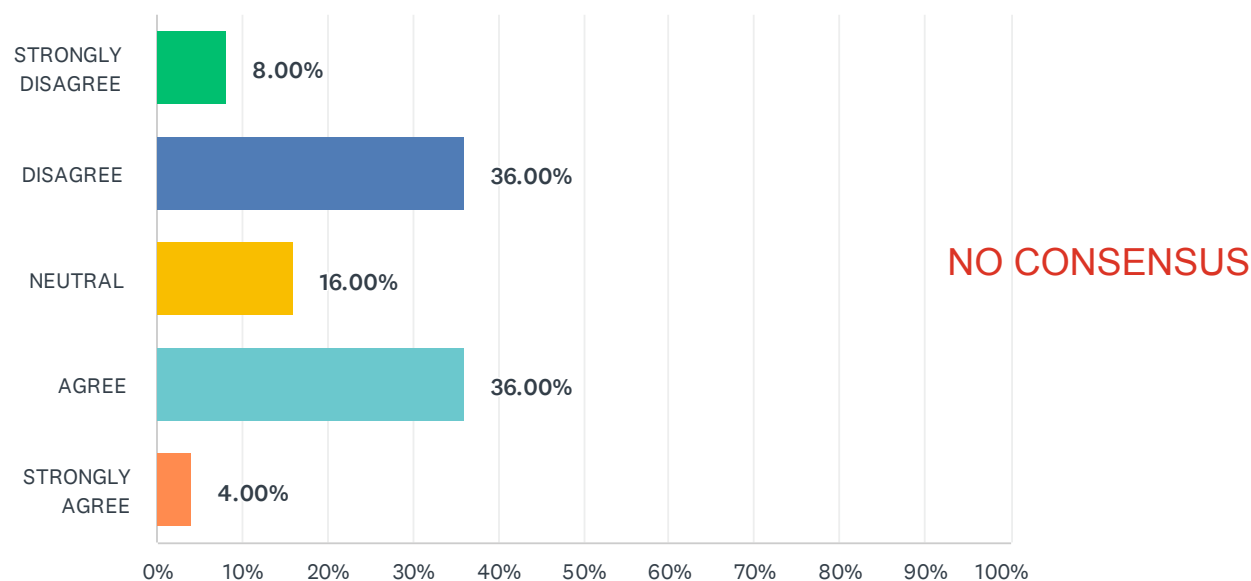
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	4.00%	1
NEUTRAL (3)	0.00%	0
AGREE (4)	44.00%	11
STRONGLY AGREE (5)	52.00%	13
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	5.00	4.44	0.70



## The specific genotype of the MELAS patient should be considered in the management of an acute SLE

Answered: 25 Skipped: 0

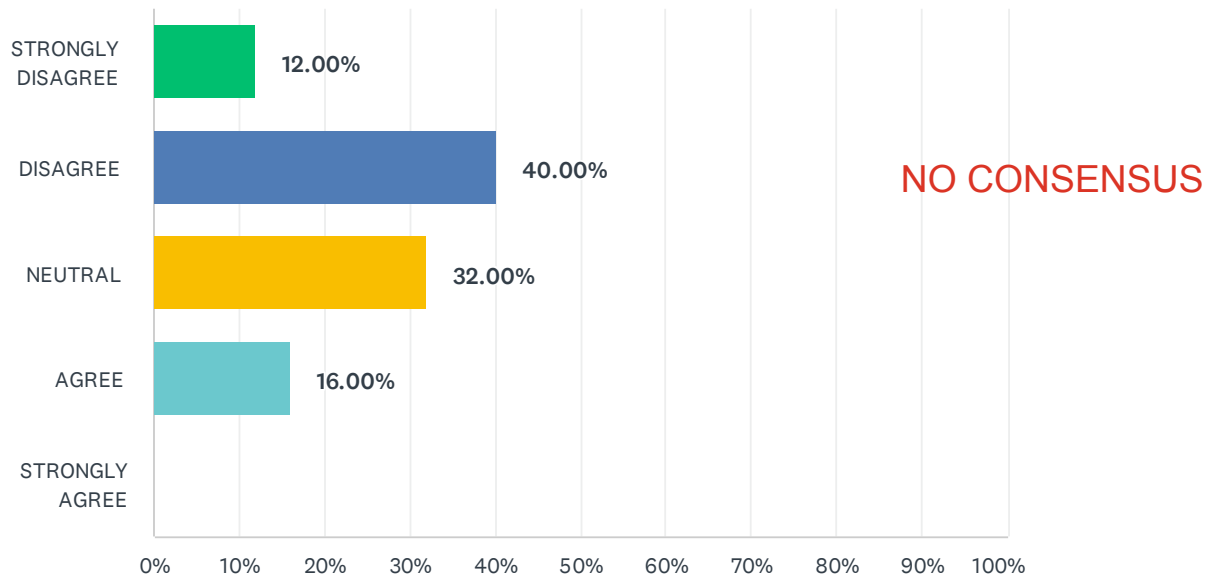


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	8.00%	2
DISAGREE (2)	36.00%	9
NEUTRAL (3)	16.00%	4
AGREE (4)	36.00%	9
STRONGLY AGREE (5)	4.00%	1
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	3.00	2.92	1.09

A “mito-cocktail” containing coenzyme Q10, vitamins, and supplements should be prescribed for the acute management of SLE.

Answered: 25 Skipped: 0

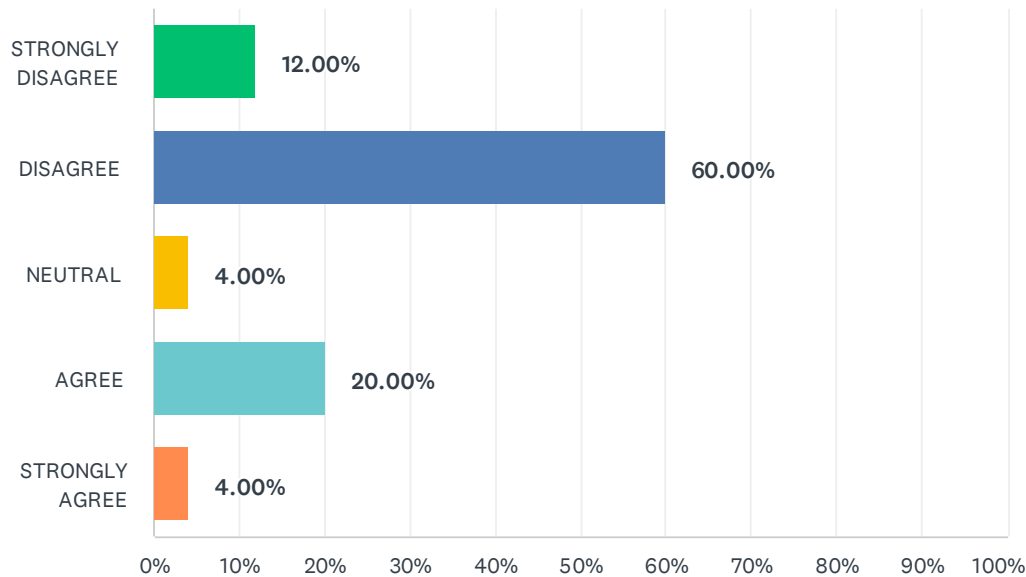


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	12.00%	3
DISAGREE (2)	40.00%	10
NEUTRAL (3)	32.00%	8
AGREE (4)	16.00%	4
STRONGLY AGREE (5)	0.00%	0
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	4.00	2.00	2.52	0.90

## Oral L-arginine should be used as long-term treatment to reduce SLE frequency

Answered: 25 Skipped: 0



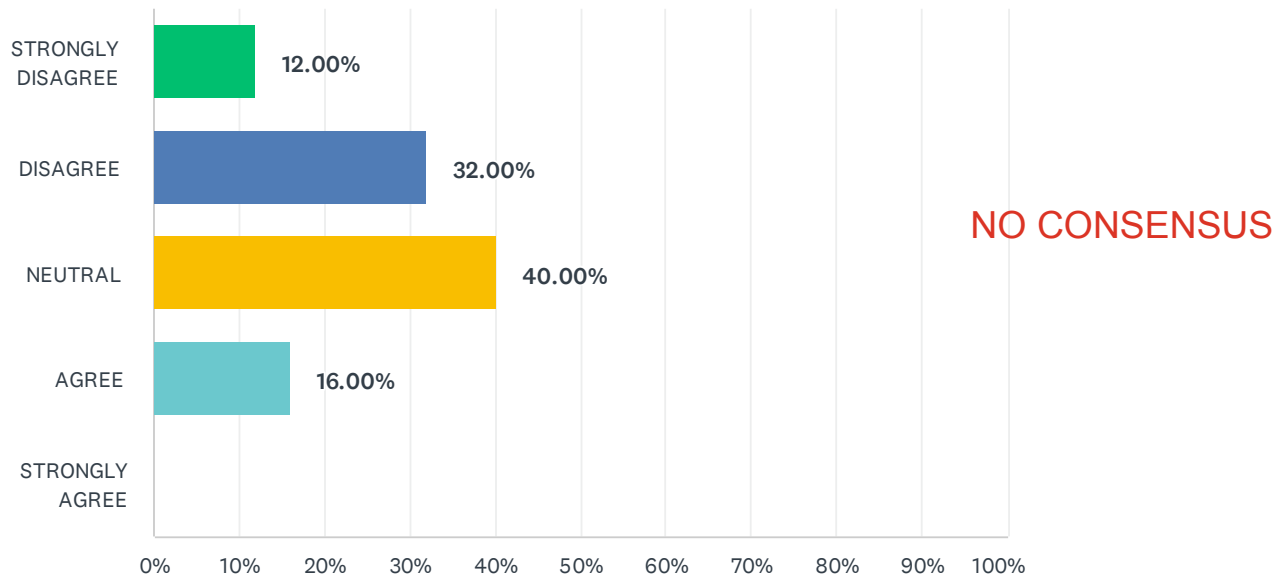
**NEGATIVE CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	12.00%	3
DISAGREE (2)	60.00%	15
NEUTRAL (3)	4.00%	1
AGREE (4)	20.00%	5
STRONGLY AGREE (5)	4.00%	1
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	2.00	2.44	1.06

## Oral citrulline should be used as long-term treatment to reduce SLE frequency

Answered: 25 Skipped: 0

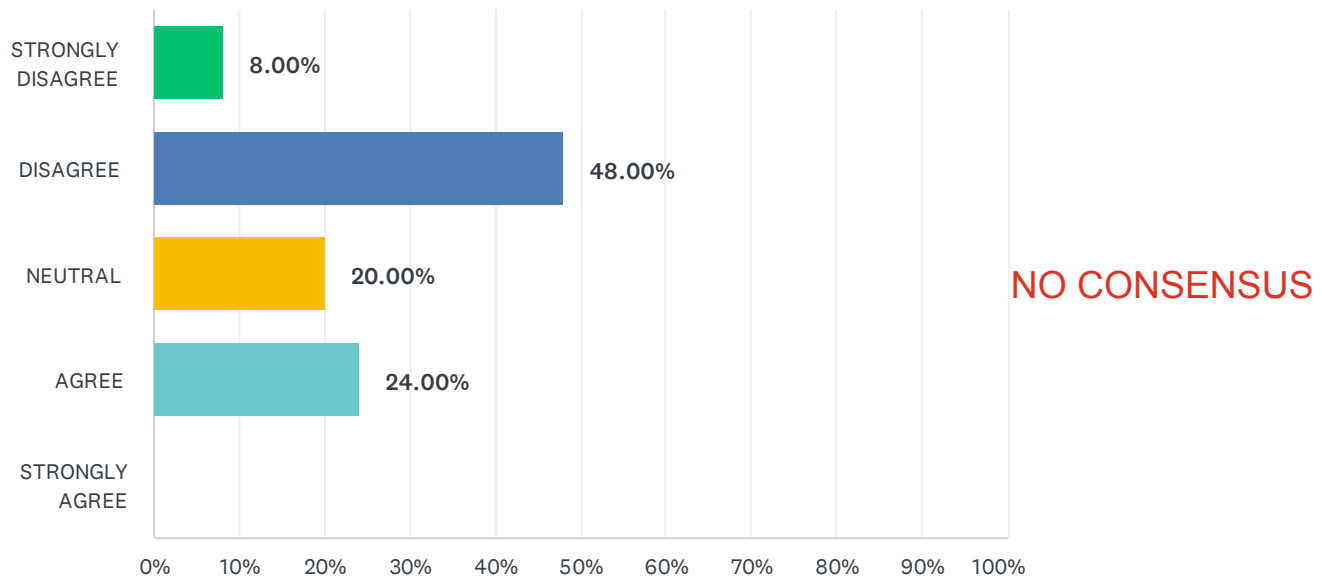


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	12.00%	3
DISAGREE (2)	32.00%	8
NEUTRAL (3)	40.00%	10
AGREE (4)	16.00%	4
STRONGLY AGREE (5)	0.00%	0
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	4.00	3.00	2.60	0.89

A “mito-cocktail” containing coenzyme Q10, vitamins and supplements should be prescribed for long-term treatment to reduce SLE frequency.

Answered: 25 Skipped: 0

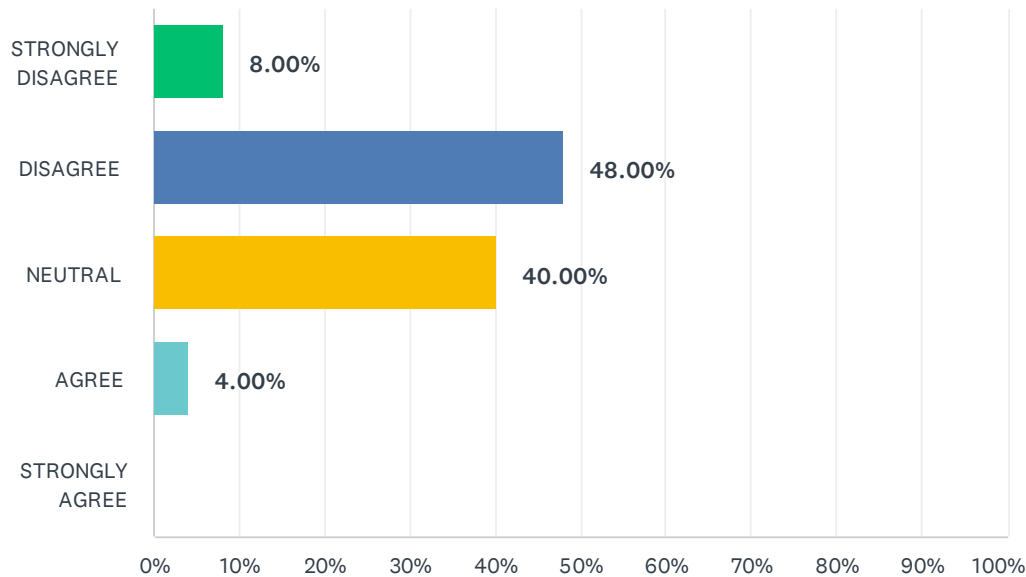


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	8.00%	2
DISAGREE (2)	48.00%	12
NEUTRAL (3)	20.00%	5
AGREE (4)	24.00%	6
STRONGLY AGREE (5)	0.00%	0
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	4.00	2.00	2.60	0.94

## Taurine should be used as a long-term medication to reduce SLE frequency

Answered: 25 Skipped: 0



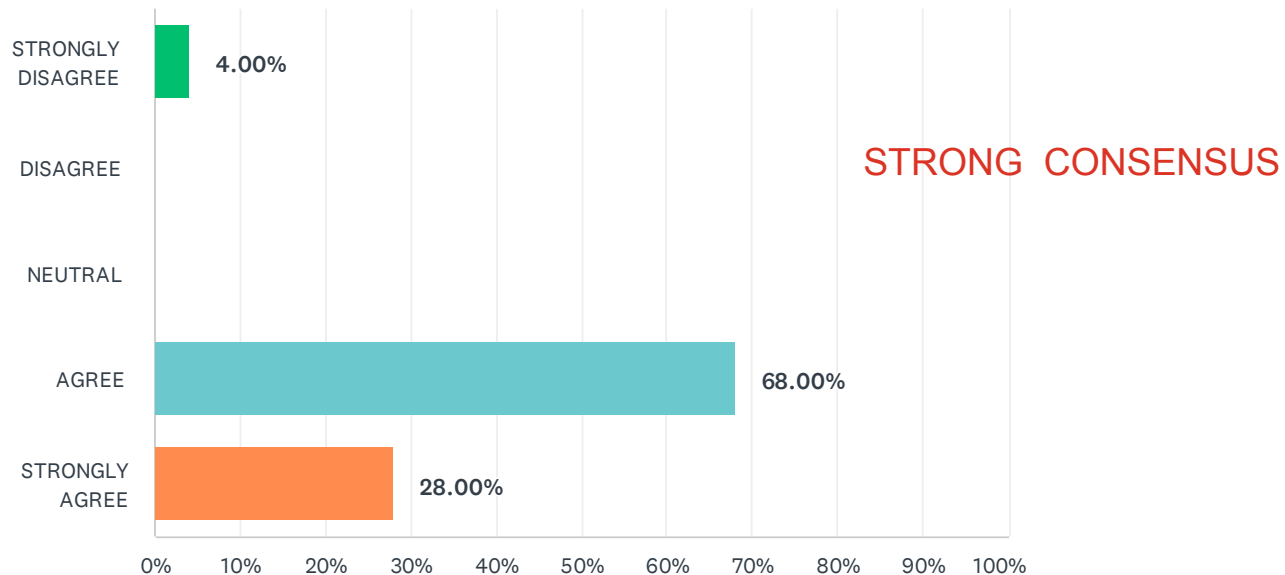
NO CONSENSUS

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	8.00%	2
DISAGREE (2)	48.00%	12
NEUTRAL (3)	40.00%	10
AGREE (4)	4.00%	1
STRONGLY AGREE (5)	0.00%	0
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	4.00	2.00	2.40	0.69

## The role of antioxidants such as CoQ10 analogues in the long-term prophylaxis of SLE requires further investigation.

Answered: 25 Skipped: 0

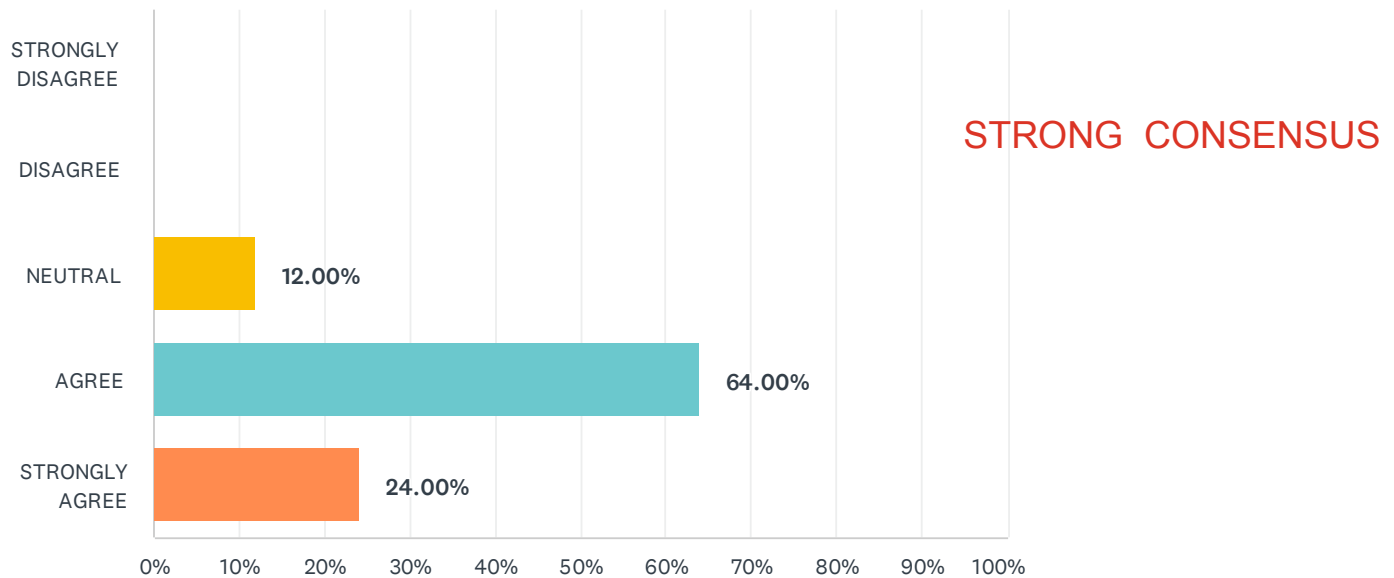


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	4.00%	1
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	68.00%	17
STRONGLY AGREE (5)	28.00%	7
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	4.00	4.16	0.78

## Anti-seizure medications might be useful to treat acute psychotic symptoms associated with stroke-like episodes or acute encephalopathic episodes

Answered: 25 Skipped: 0



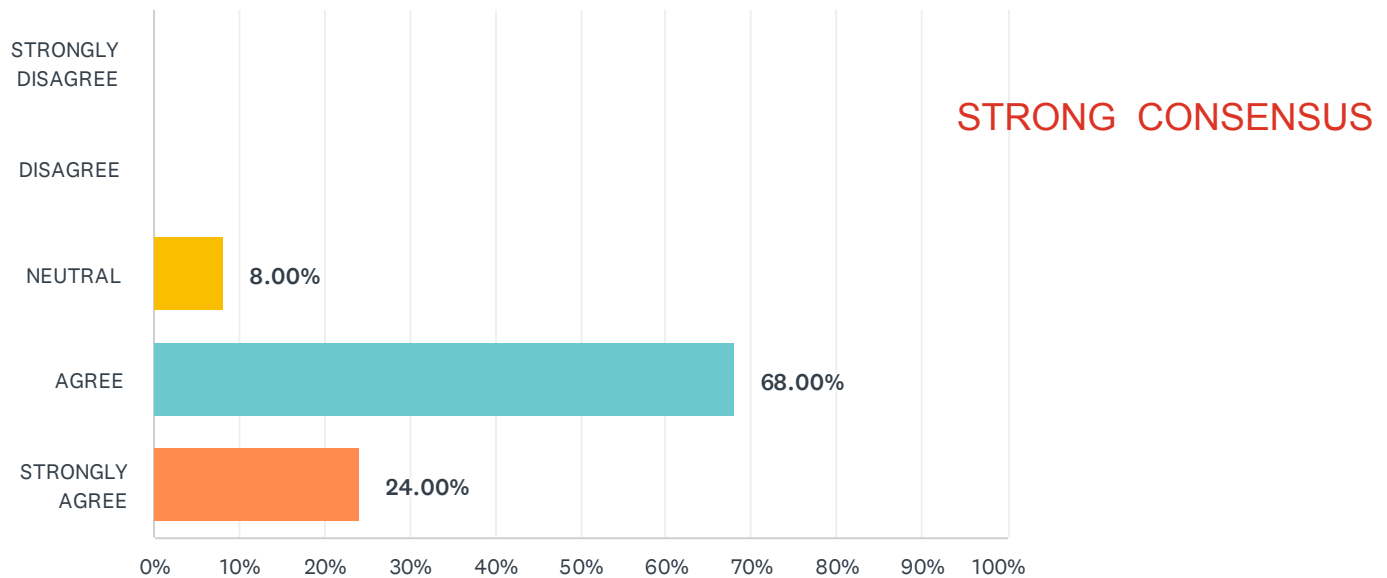
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	12.00%	3
AGREE (4)	64.00%	16
STRONGLY AGREE (5)	24.00%	6
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.00	4.12	0.59



Atypical antipsychotic drugs like risperidone or quetiapine can be useful to manage acute psychotic symptoms associated with stroke-like/ acute encephalopathic episodes.

Answered: 25 Skipped: 0

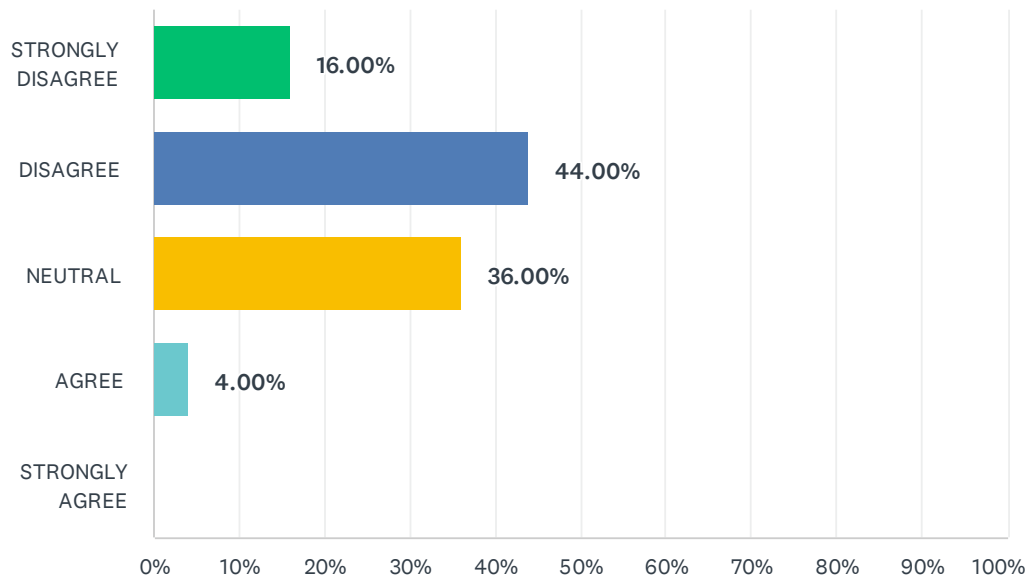


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	8.00%	2
AGREE (4)	68.00%	17
STRONGLY AGREE (5)	24.00%	6
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.00	4.16	0.54

Nutritional supplements like creatine monohydrate or coenzyme Q10 might be useful to manage acute psychotic symptoms associated with stroke-like/ acute encephalopathic episodes.

Answered: 25 Skipped: 0



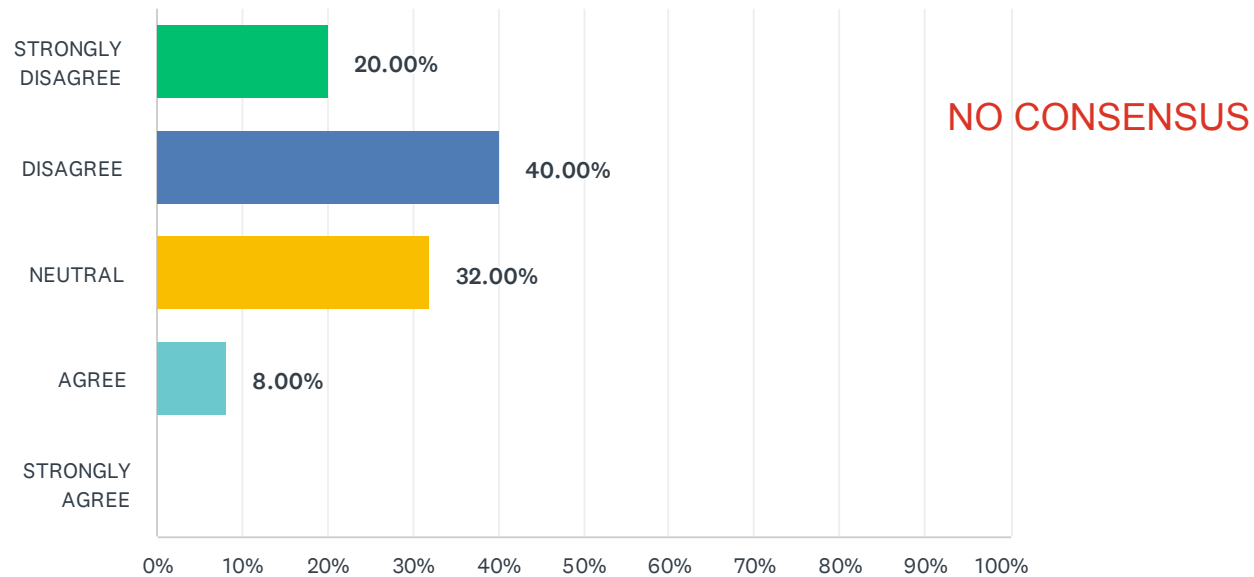
NO CONSENSUS

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	16.00%	4
DISAGREE (2)	44.00%	11
NEUTRAL (3)	36.00%	9
AGREE (4)	4.00%	1
STRONGLY AGREE (5)	0.00%	0
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	4.00	2.00	2.28	0.78

## L-Arginine might be useful to treat acute psychotic symptoms associated with stroke-like episodes or acute encephalopathic episodes

Answered: 25 Skipped: 0

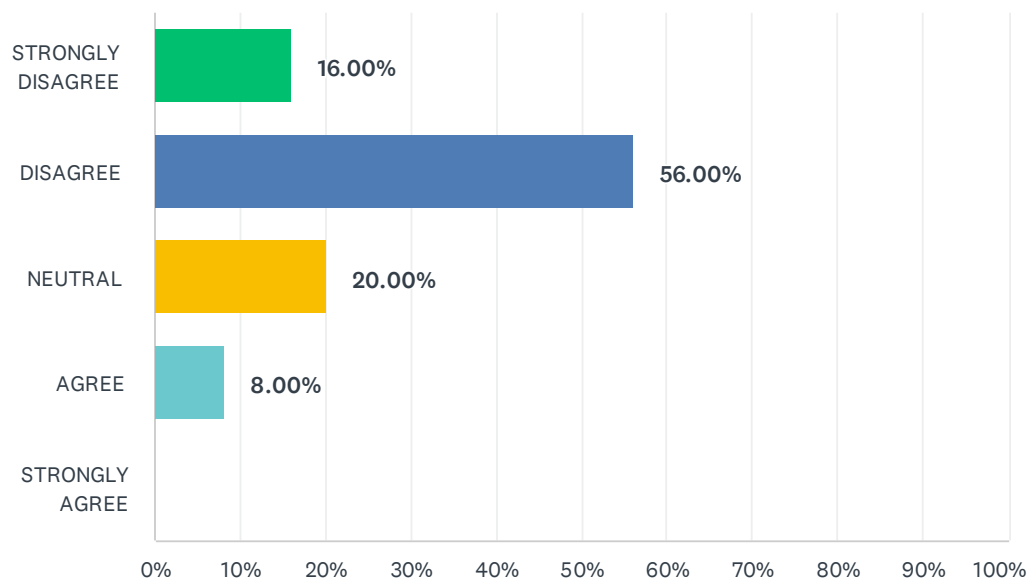


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	20.00%	5
DISAGREE (2)	40.00%	10
NEUTRAL (3)	32.00%	8
AGREE (4)	8.00%	2
STRONGLY AGREE (5)	0.00%	0
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	4.00	2.00	2.28	0.87

## Oral L-arginine and coenzyme Q might be useful to prevent psychotic episodes in PMD patients.

Answered: 25 Skipped: 0



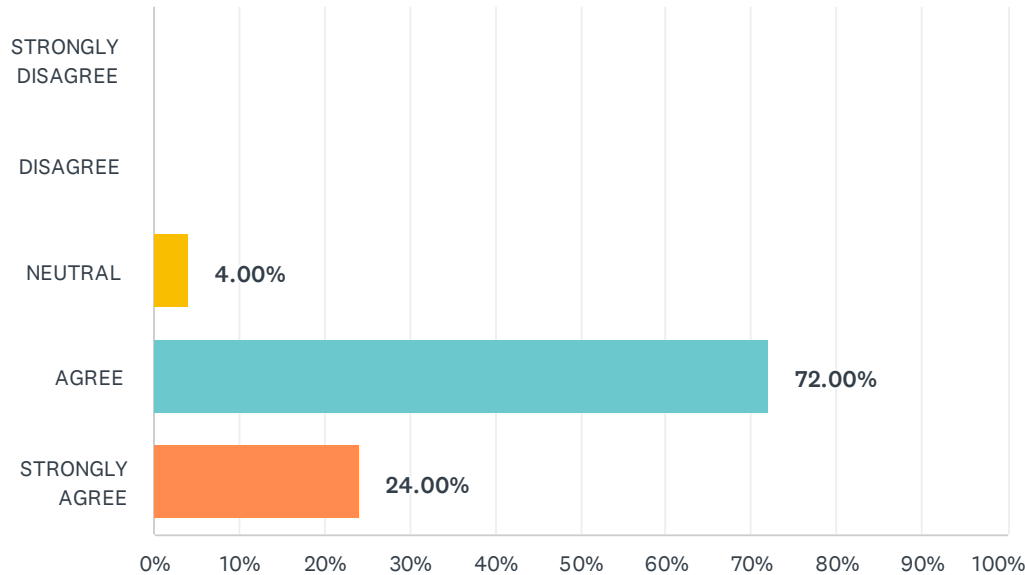
NEGATIVE CONSENSUS

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	16.00%	4
DISAGREE (2)	56.00%	14
NEUTRAL (3)	20.00%	5
AGREE (4)	8.00%	2
STRONGLY AGREE (5)	0.00%	0
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	4.00	2.00	2.20	0.80

## Cognitive rehabilitation might be useful to treat long-term psychiatric and cognitive symptoms

Answered: 25 Skipped: 0



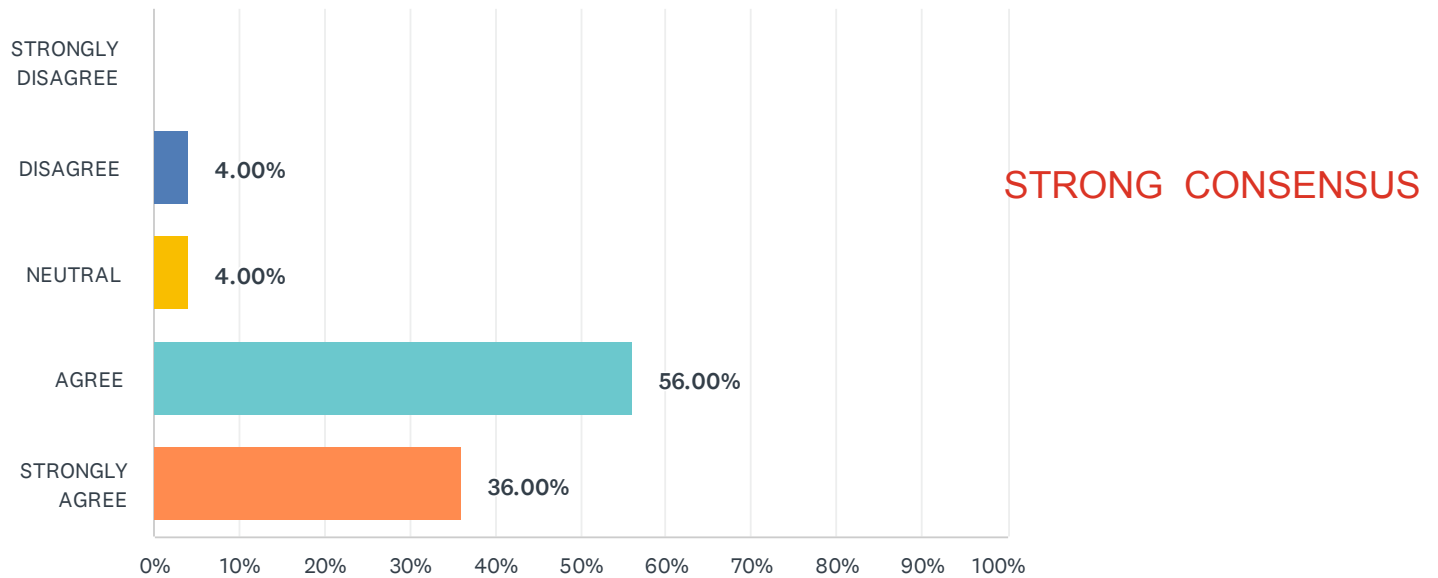
**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	4.00%	1
AGREE (4)	72.00%	18
STRONGLY AGREE (5)	24.00%	6
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.00	4.20	0.49

## SSRI and SNRI might be useful to treat depression and anxiety.

Answered: 25 Skipped: 0

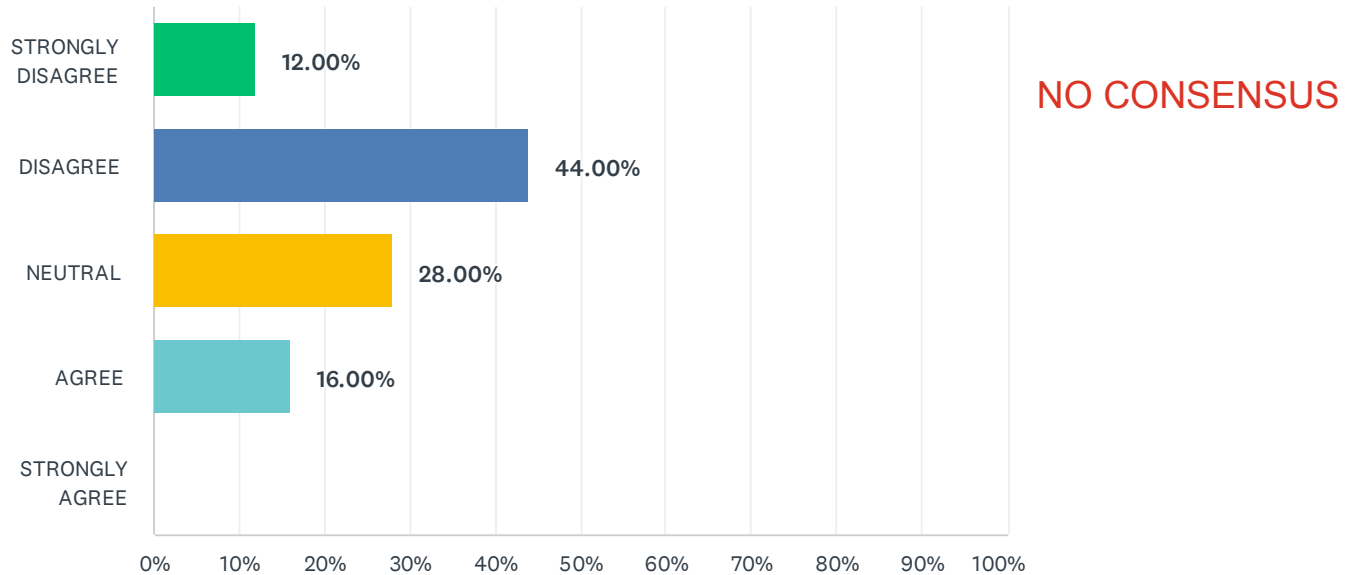


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	4.00%	1
NEUTRAL (3)	4.00%	1
AGREE (4)	56.00%	14
STRONGLY AGREE (5)	36.00%	9
TOTAL		25

BASIC STATISTICS				
Minimum 2.00	Maximum 5.00	Median 4.00	Mean 4.24	Standard Deviation 0.71

## Oral L-arginine and coenzyme Q might be useful to improve or preserve cognition

Answered: 25 Skipped: 0

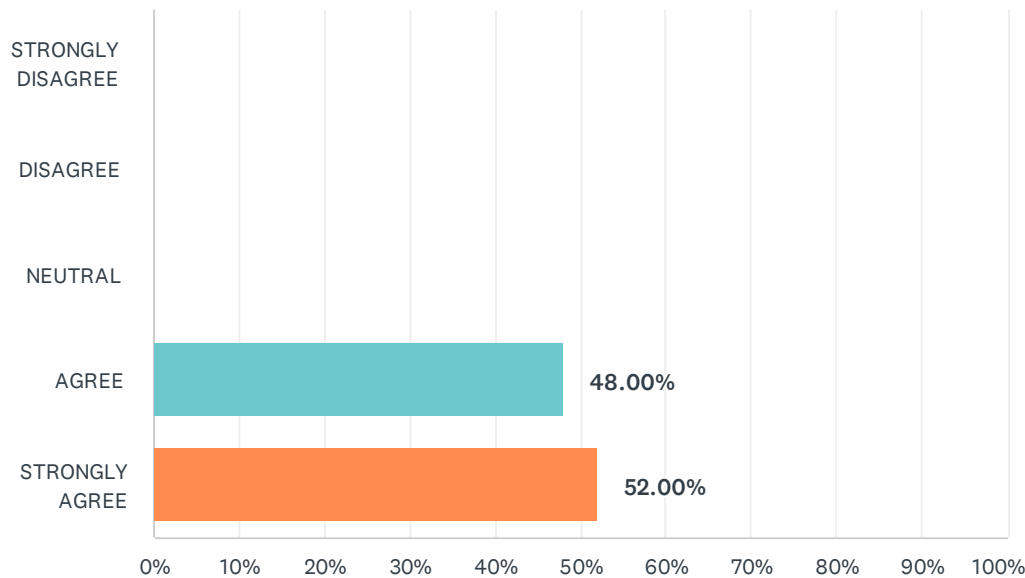


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	12.00%	3
DISAGREE (2)	44.00%	11
NEUTRAL (3)	28.00%	7
AGREE (4)	16.00%	4
STRONGLY AGREE (5)	0.00%	0
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	4.00	2.00	2.48	0.90

In patients with MELAS syndrome, headache should be systematically assessed to enable timely intervention and optimize quality of life.

Answered: 25 Skipped: 0



**STRONG CONSENSUS**

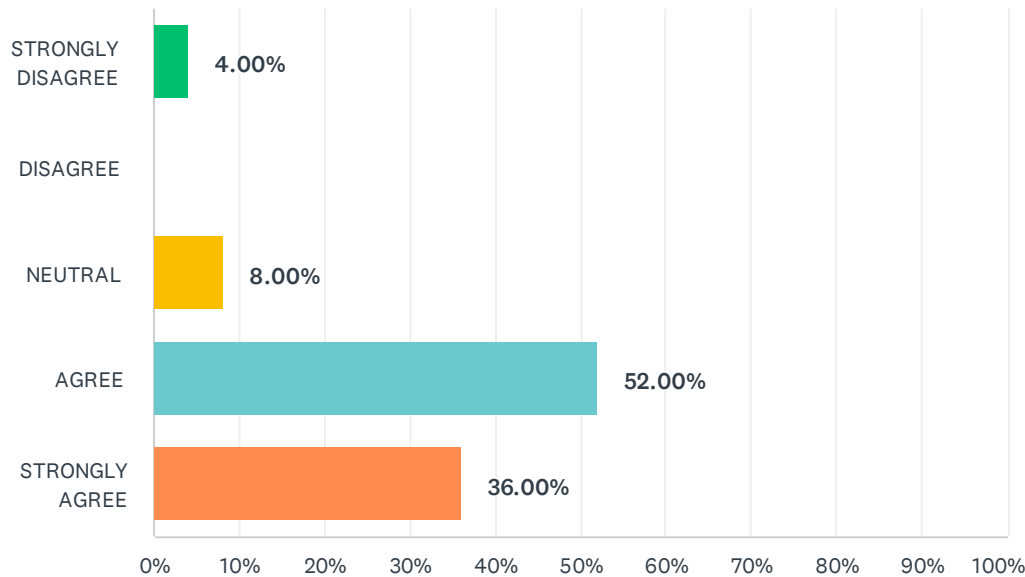
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	48.00%	12
STRONGLY AGREE (5)	52.00%	13
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
4.00	5.00	5.00	4.52	0.50



## Pharmacological and non-pharmacological headache management in MELAS syndrome can be applied regardless of genotype and age.

Answered: 25 Skipped: 0



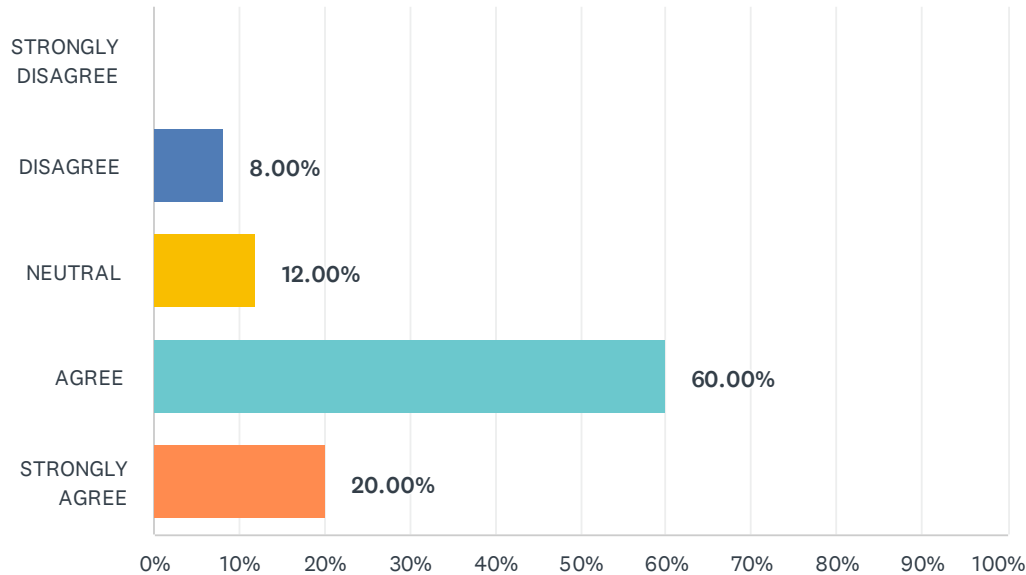
**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	4.00%	1
DISAGREE (2)	0.00%	0
NEUTRAL (3)	8.00%	2
AGREE (4)	52.00%	13
STRONGLY AGREE (5)	36.00%	9
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	4.00	4.16	0.88

## Persistent headache unresponsive to treatment should prompt neuroimaging to exclude a stroke-like episode.

Answered: 25 Skipped: 0

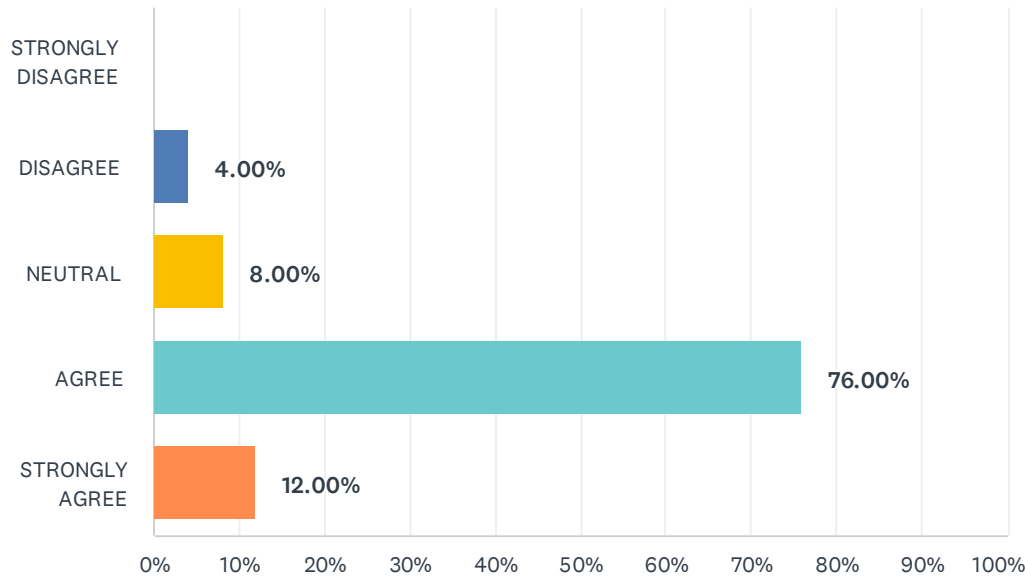


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	8.00%	2
NEUTRAL (3)	12.00%	3
AGREE (4)	60.00%	15
STRONGLY AGREE (5)	20.00%	5
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	3.92	0.80

There are no specific contraindications to the use of standard pharmacological classes for acute headache treatment (NSAIDs, triptans, paracetamol, combination analgesics, opioids, ditans, and gepants).

Answered: 25 Skipped: 0

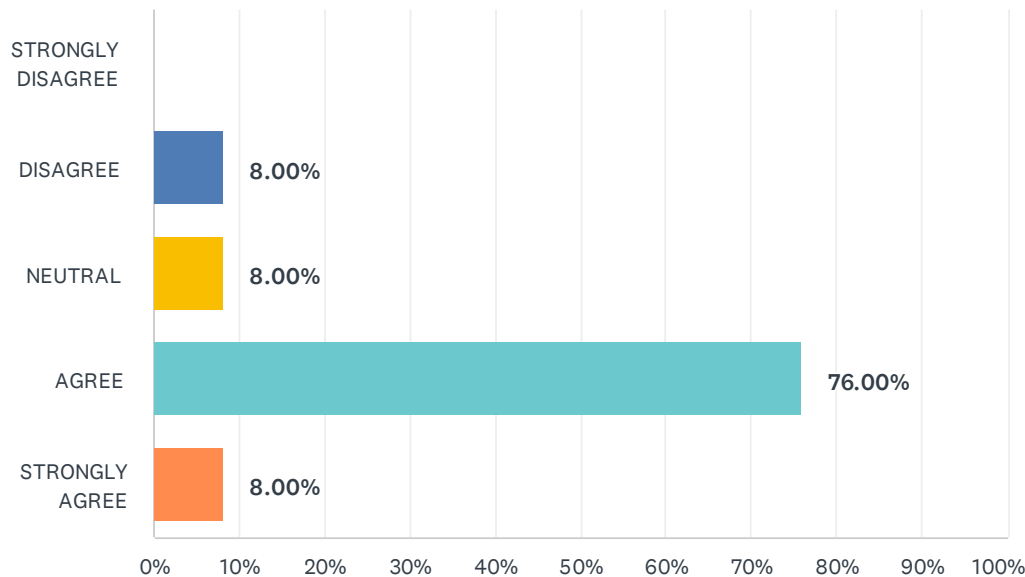


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	4.00%	1
NEUTRAL (3)	8.00%	2
AGREE (4)	76.00%	19
STRONGLY AGREE (5)	12.00%	3
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	3.96	0.60

There are no specific contraindications to the use of standard pharmacological classes for migraine prevention (antidepressants, beta blockers, topiramate, botulin toxin, calcium channel blockers and other blood pressure-lowering agents).

Answered: 25 Skipped: 0



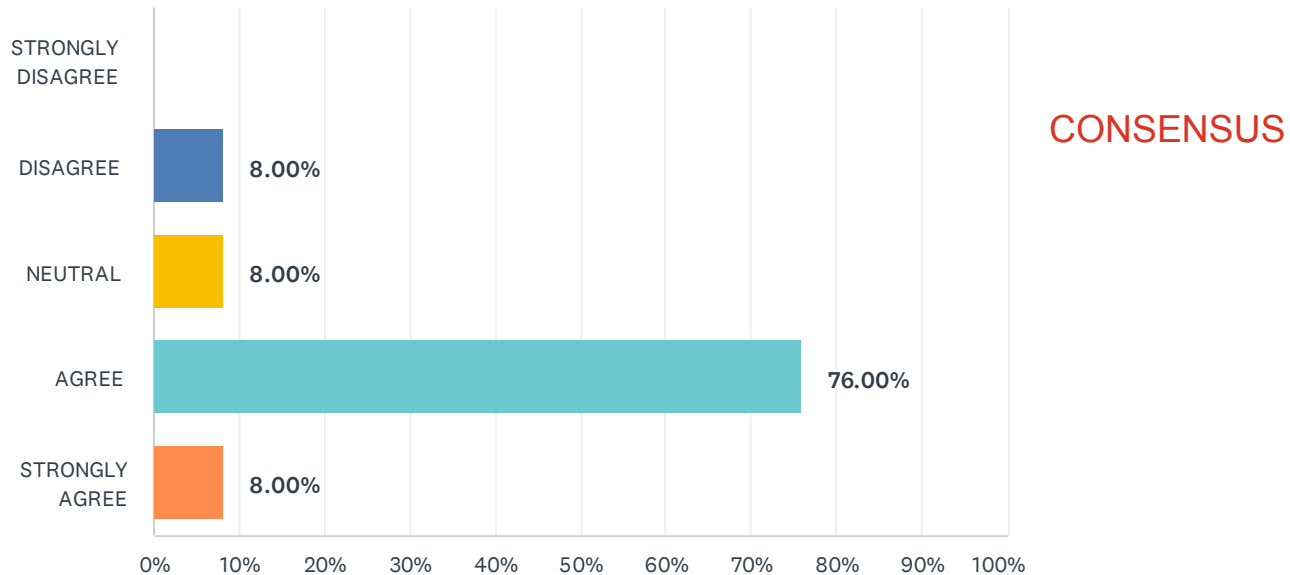
CONSENSUS

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	8.00%	2
NEUTRAL (3)	8.00%	2
AGREE (4)	76.00%	19
STRONGLY AGREE (5)	8.00%	2
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	3.84	0.67

There are no specific contraindications to the use of standard pharmacological classes for migraine prevention (antidepressants, beta blockers, topiramate, botulin toxin, calcium channel blockers and other blood pressure-lowering agents).

Answered: 25 Skipped: 0

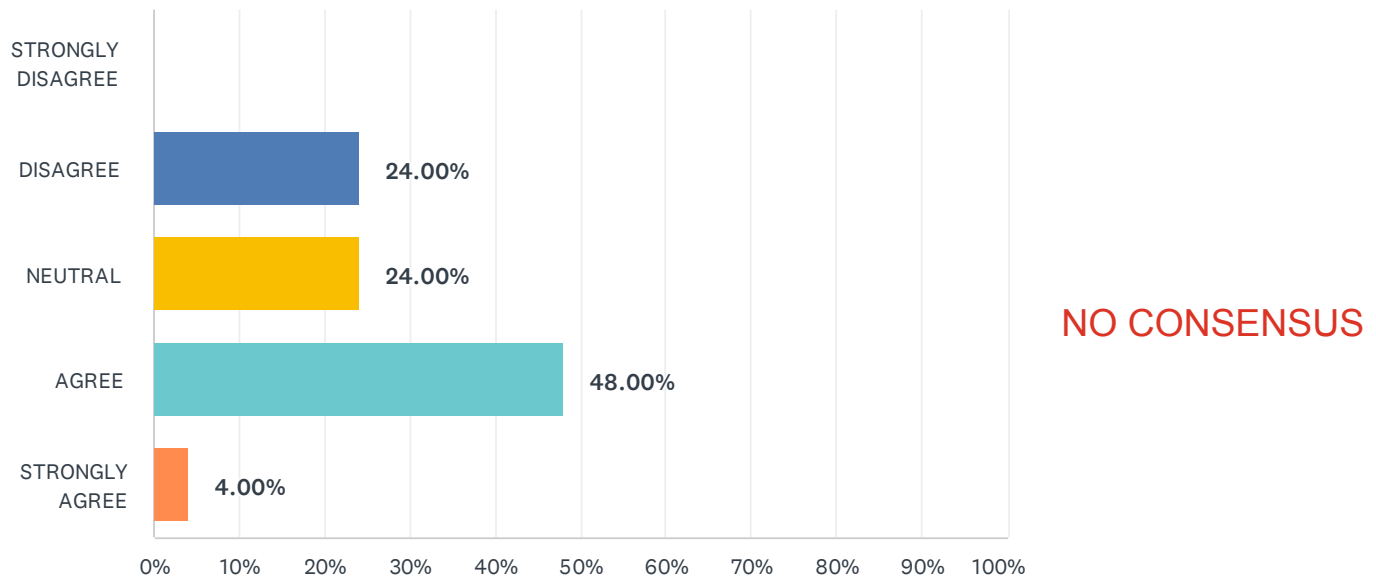


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	8.00%	2
NEUTRAL (3)	8.00%	2
AGREE (4)	76.00%	19
STRONGLY AGREE (5)	8.00%	2
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	3.84	0.67

Valproate may be considered for migraine prevention in patients not harbouring POLG variants and without liver impairment (or at risk of liver impairment), with careful monitoring.

Answered: 25 Skipped: 0

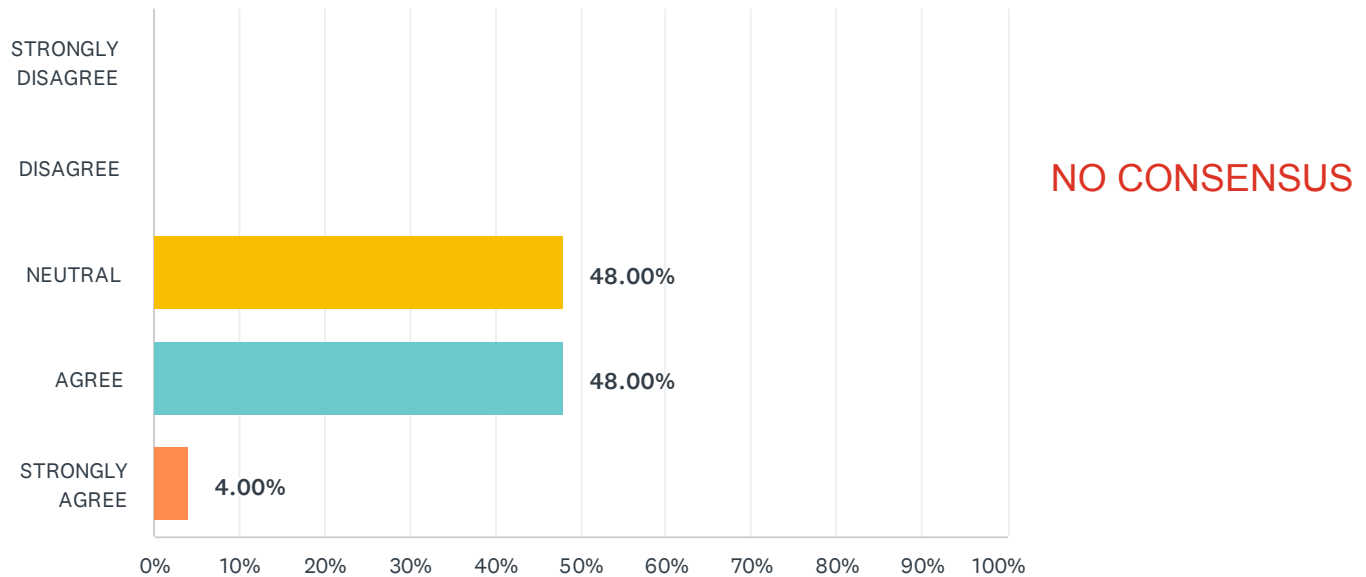


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	24.00%	6
NEUTRAL (3)	24.00%	6
AGREE (4)	48.00%	12
STRONGLY AGREE (5)	4.00%	1
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	3.32	0.88

## Monoclonal antibodies targeting the CGRP pathway are effective and well-tolerated for migraine prevention.

Answered: 25 Skipped: 0

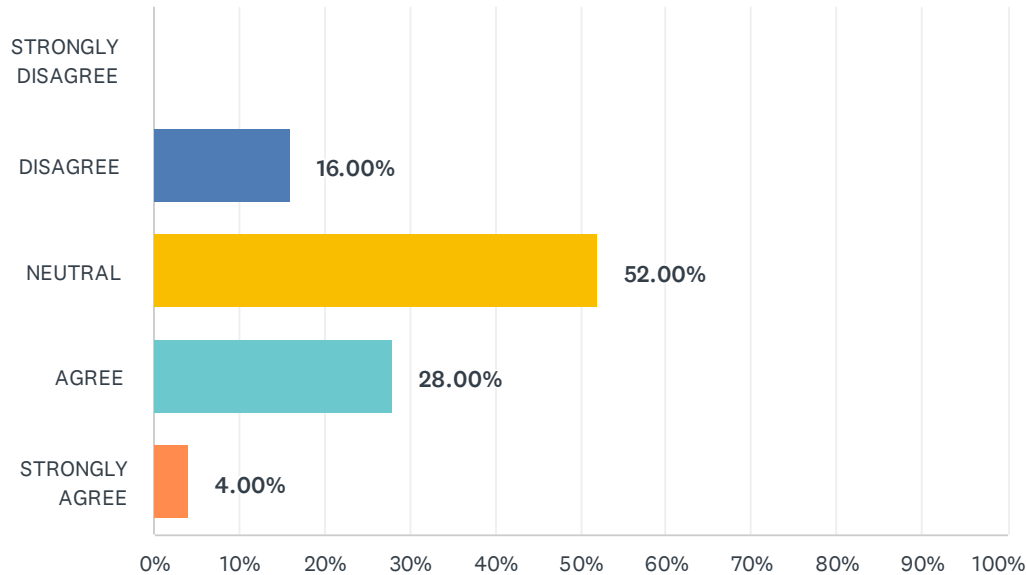


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	48.00%	12
AGREE (4)	48.00%	12
STRONGLY AGREE (5)	4.00%	1
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.00	3.56	0.57

## Nerve stimulation devices should be avoided in the management of headache

Answered: 25 Skipped: 0



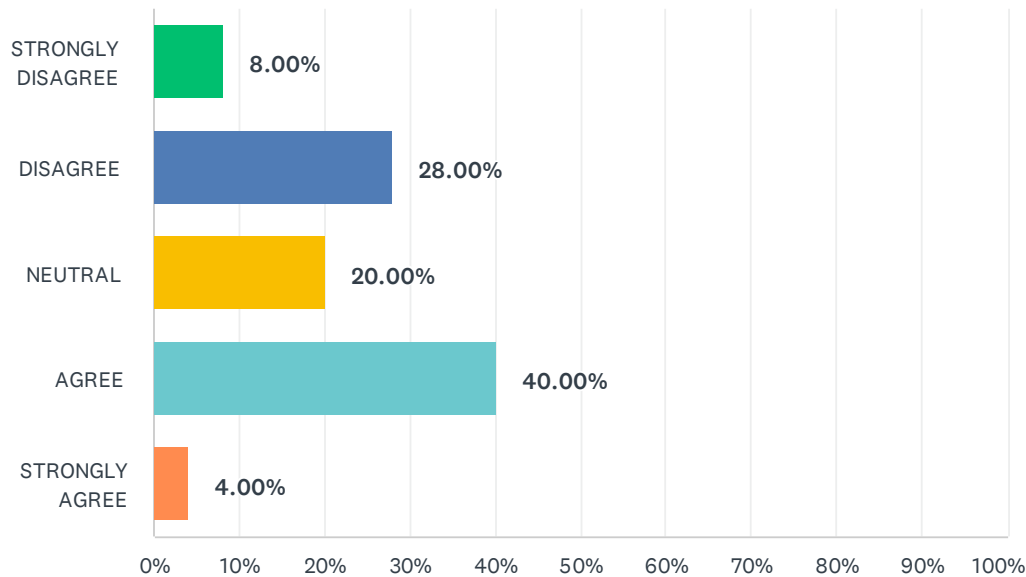
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	16.00%	4
NEUTRAL (3)	52.00%	13
AGREE (4)	28.00%	7
STRONGLY AGREE (5)	4.00%	1
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	3.00	3.20	0.75



## A “mito-cocktail” containing coenzyme Q10 and riboflavin should be considered for migraine prevention.

Answered: 25 Skipped: 0



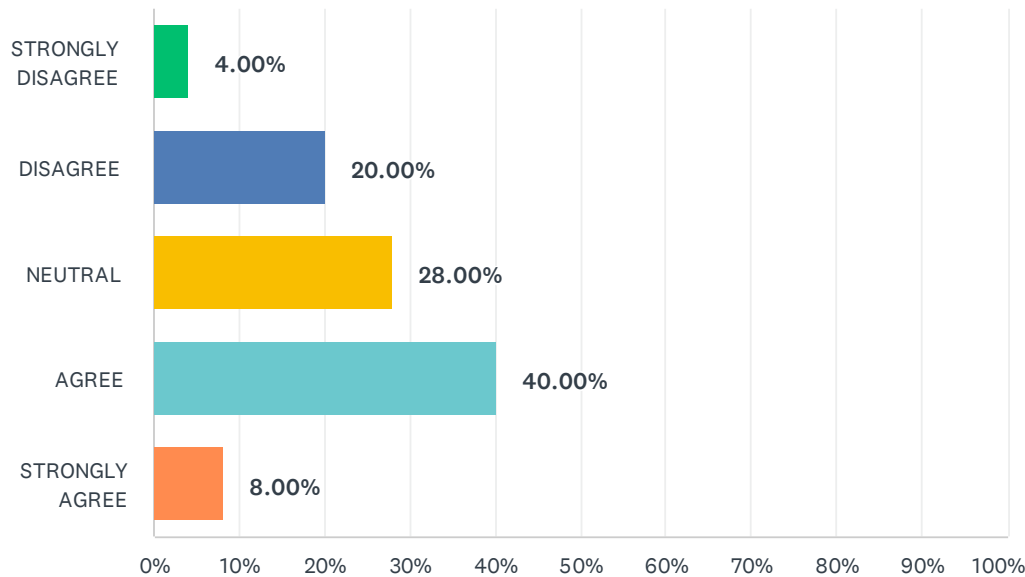
NO CONSENSUS

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	8.00%	2
DISAGREE (2)	28.00%	7
NEUTRAL (3)	20.00%	5
AGREE (4)	40.00%	10
STRONGLY AGREE (5)	4.00%	1
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	3.00	3.04	1.08

## Acute and preventive headache treatments should be considered to reduce the frequency and severity of stroke-like episodes.

Answered: 25 Skipped: 0

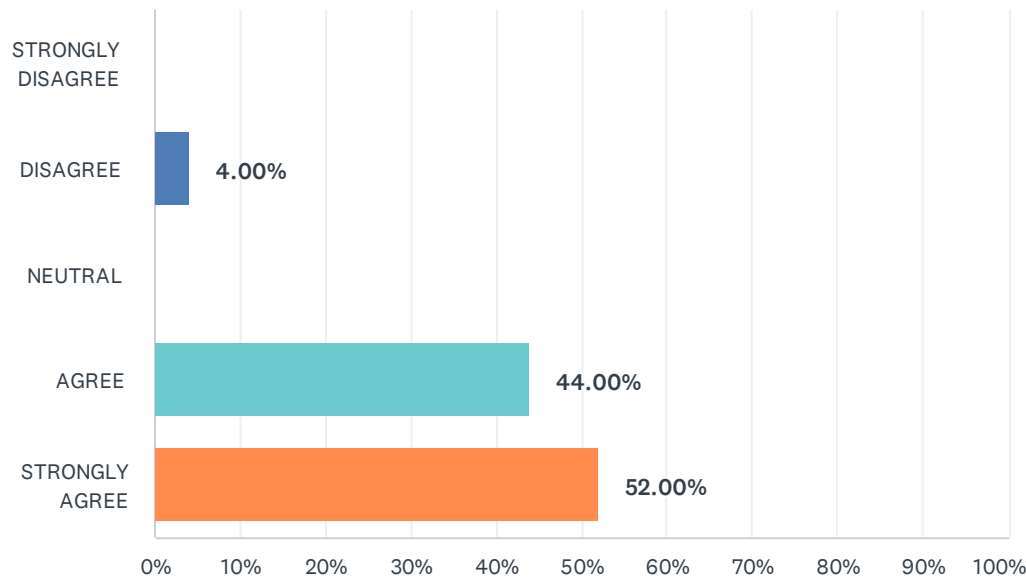


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	4.00%	1
DISAGREE (2)	20.00%	5
NEUTRAL (3)	28.00%	7
AGREE (4)	40.00%	10
STRONGLY AGREE (5)	8.00%	2
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	3.00	3.28	1.00

## The European consensus study from the InterERNS Mitochondrial Working Group (PMID: 38576261) regarding the safety and toxicity of anti-seizure medications is appropriate and should be endorsed.

Answered: 25 Skipped: 0



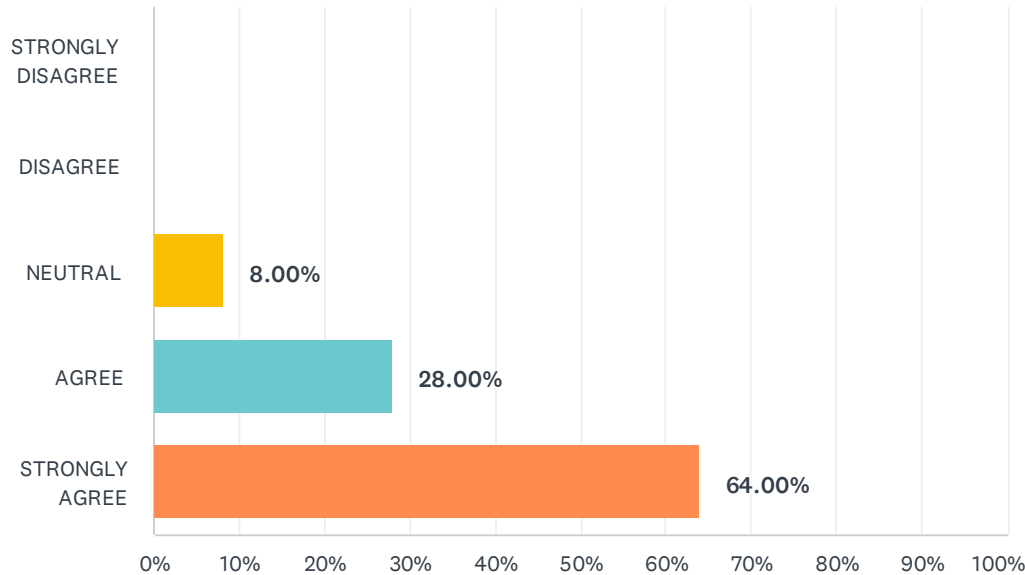
**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	4.00%	1
NEUTRAL (3)	0.00%	0
AGREE (4)	44.00%	11
STRONGLY AGREE (5)	52.00%	13
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	5.00	4.44	0.70

## EEG should be considered even in the absence of a prior history of epilepsy

Answered: 25 Skipped: 0



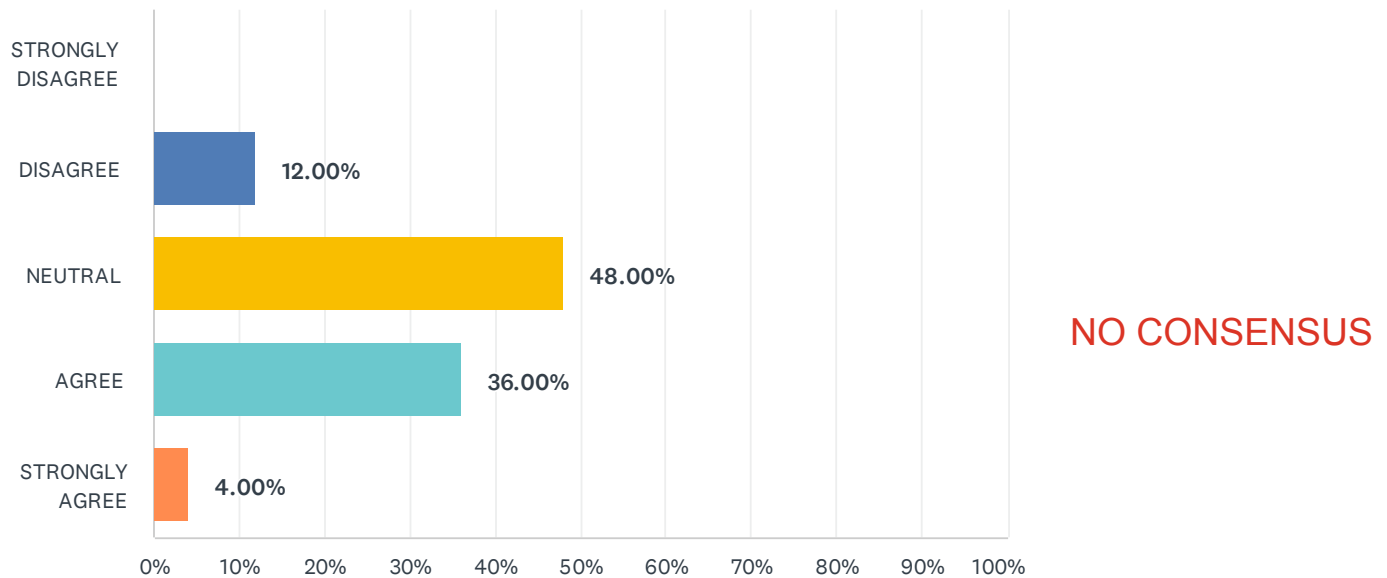
STRONG CONSENSUS

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	8.00%	2
AGREE (4)	28.00%	7
STRONGLY AGREE (5)	64.00%	16
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	5.00	4.56	0.64

## Early anti-seizure medication in patients with stroke-like episodes without seizures may influence the risk of epilepsy and recurrence of stroke-like episodes

Answered: 25 Skipped: 0

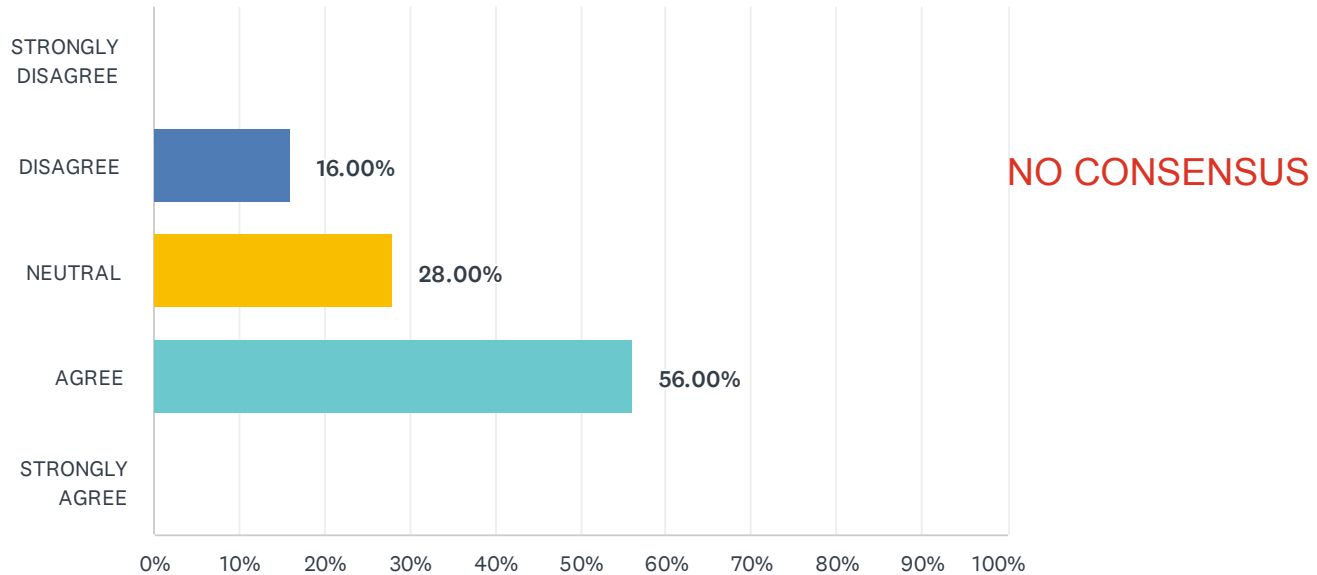


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	12.00%	3
NEUTRAL (3)	48.00%	12
AGREE (4)	36.00%	9
STRONGLY AGREE (5)	4.00%	1
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	3.00	3.32	0.73

## Alternative approaches for epilepsy management (i.e. anti-inflammatory drugs, cofactor supplementation) should be considered.

Answered: 25 Skipped: 0

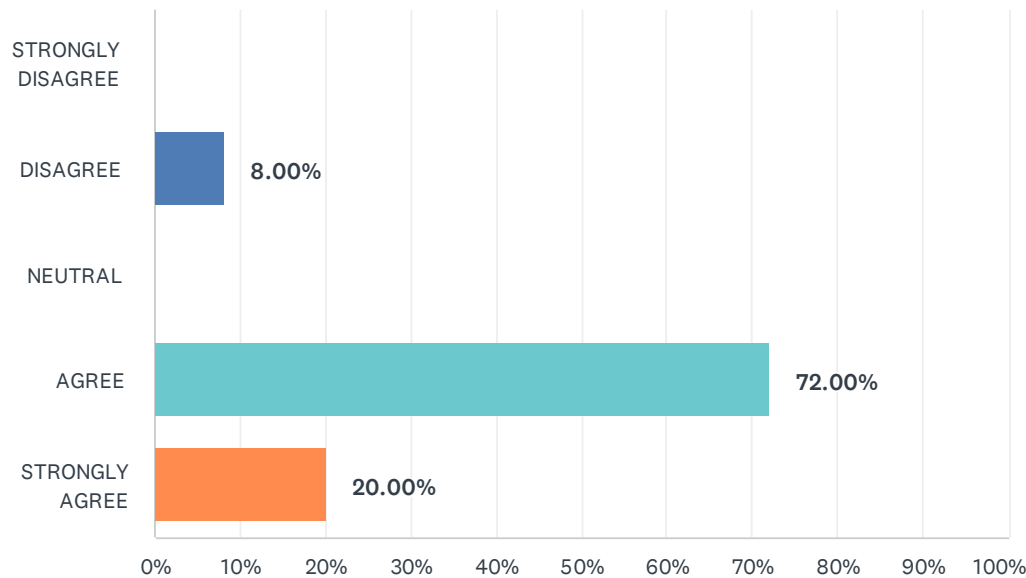


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	16.00%	4
NEUTRAL (3)	28.00%	7
AGREE (4)	56.00%	14
STRONGLY AGREE (5)	0.00%	0
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	4.00	4.00	3.40	0.75

Gastrointestinal Dysmotility is frequently seen in SLE. Conservative management with aggressive hydration and nutrition started early and continued are typically sufficient to prevent surgical intervention.

Answered: 25 Skipped: 0



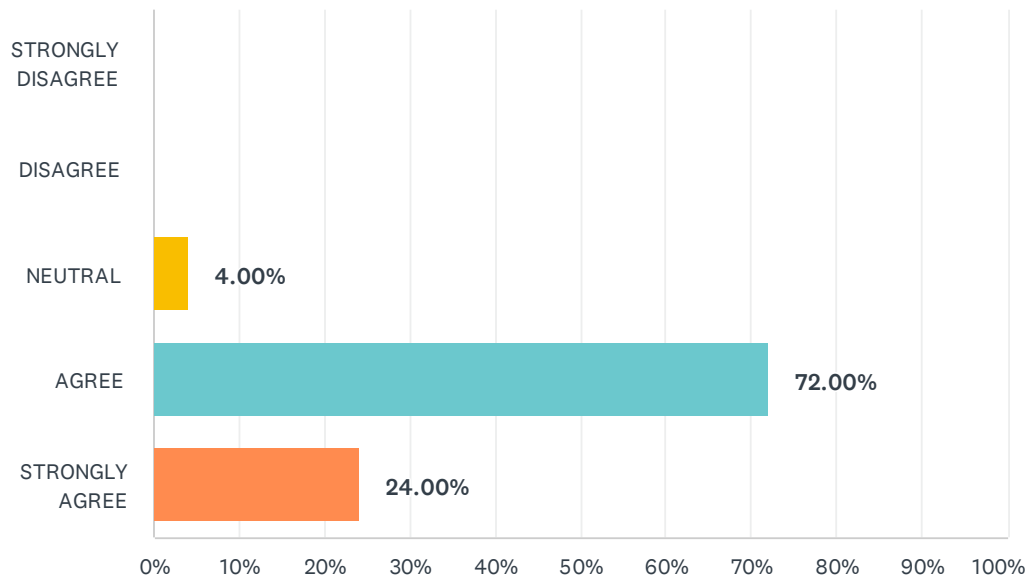
STRONG CONSENSUS

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	8.00%	2
NEUTRAL (3)	0.00%	0
AGREE (4)	72.00%	18
STRONGLY AGREE (5)	20.00%	5
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	4.04	0.72

Prokinetics should be considered if conservative measures such as laxatives fail to achieve full resolution of gastrointestinal symptoms, and to prevent IPO.

Answered: 25 Skipped: 0



**STRONG CONSENSUS**

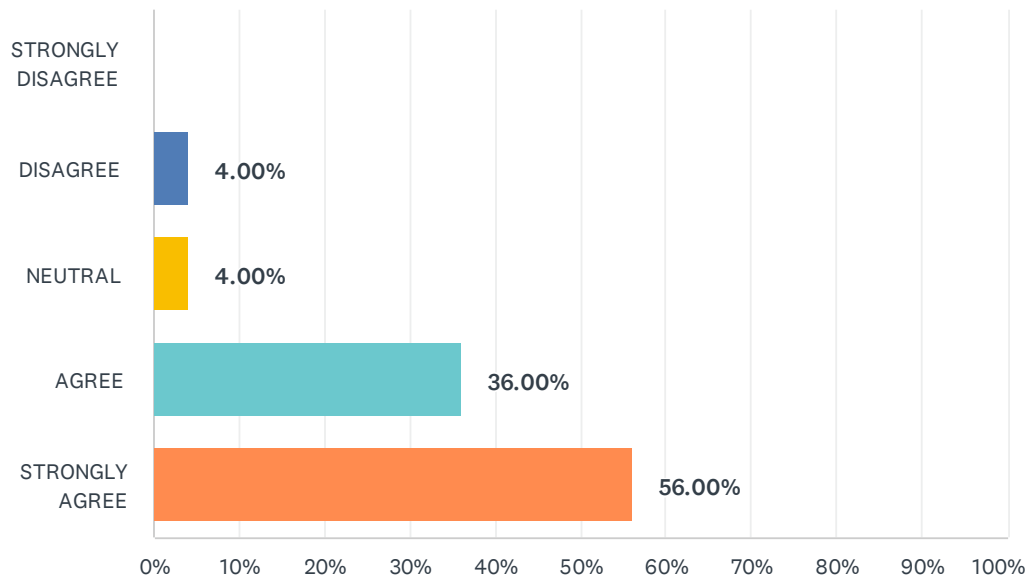
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	4.00%	1
AGREE (4)	72.00%	18
STRONGLY AGREE (5)	24.00%	6
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.00	4.20	0.49



Sometimes infections precede or complicate SLEs. Index of suspicion should be high, appropriate antibiotics should be tailored to the underlying infection.

Answered: 25 Skipped: 0



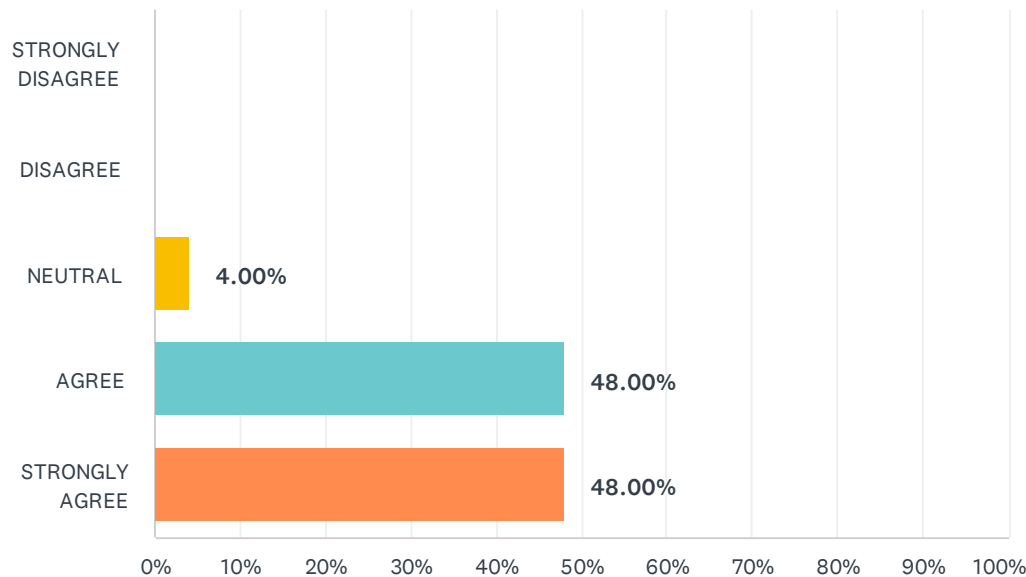
STRONG CONSENSUS

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	4.00%	1
NEUTRAL (3)	4.00%	1
AGREE (4)	36.00%	9
STRONGLY AGREE (5)	56.00%	14
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	5.00	4.44	0.75

Cardiac symptoms including cardiomyopathy, cardiac arrhythmia, acute cardiac failure, and cardiogenic shock can occur in the setting of SLE and MELAS. Patients should be monitored with continuous cardiac monitoring and echocardiogram performed if indicated

Answered: 25 Skipped: 0



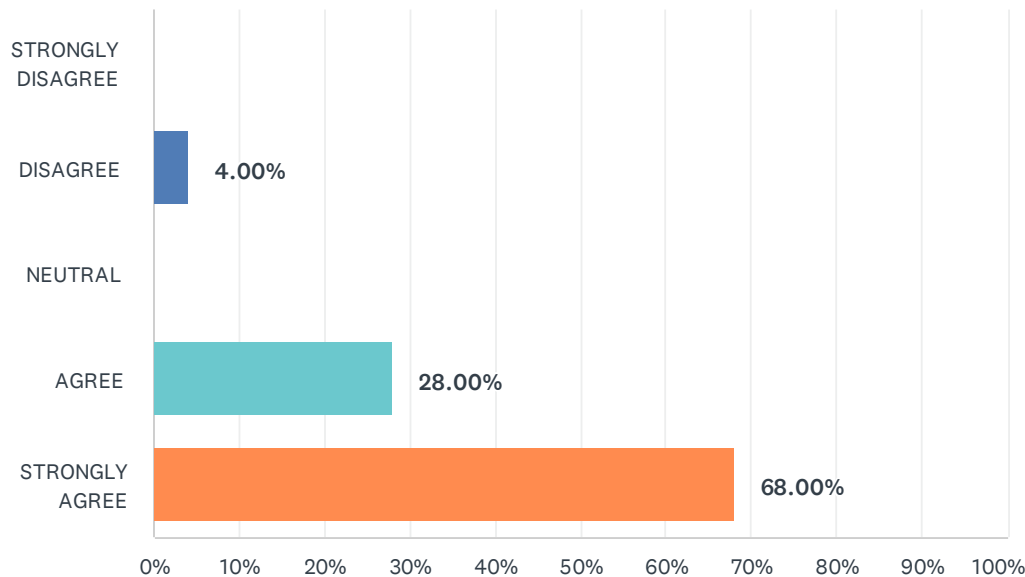
**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	4.00%	1
AGREE (4)	48.00%	12
STRONGLY AGREE (5)	48.00%	12
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.00	4.44	0.57

The onset of stroke-like episodes occur in MELAS may trigger/exacerbate multiple organ failure if accompanied by infection, metabolic crises, and increased metabolic demand that is not properly managed. Proper hydration, continued adequate caloric intake and close monitoring for complications are necessary and should start as soon as patient is admitted

Answered: 25 Skipped: 0



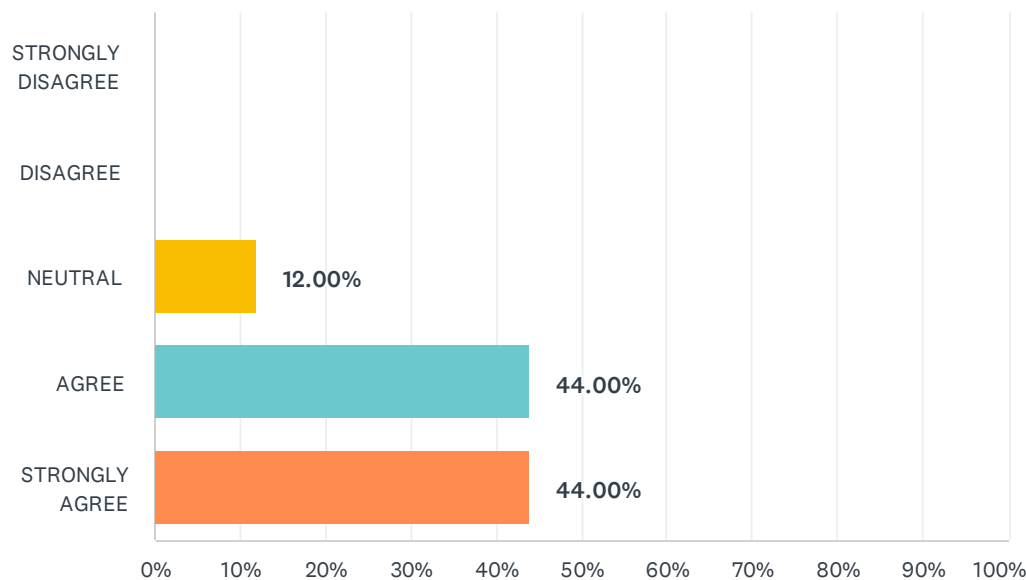
**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	4.00%	1
NEUTRAL (3)	0.00%	0
AGREE (4)	28.00%	7
STRONGLY AGREE (5)	68.00%	17
TOTAL		25

BASIC STATISTICS				
Minimum 2.00	Maximum 5.00	Median 5.00	Mean 4.60	Standard Deviation 0.69

In the acute management of metabolic acidosis in MELAS, clinicians should consider that elevated lactic acid and anion gap may be baseline findings and not all cases require treatment. Priority should be given to monitoring and interventions like fluid resuscitation, bicarbonate therapy, and metabolic substrate supplementation, aiming to stabilize severely fluctuating baseline pH while minimizing complications.

Answered: 25 Skipped: 0



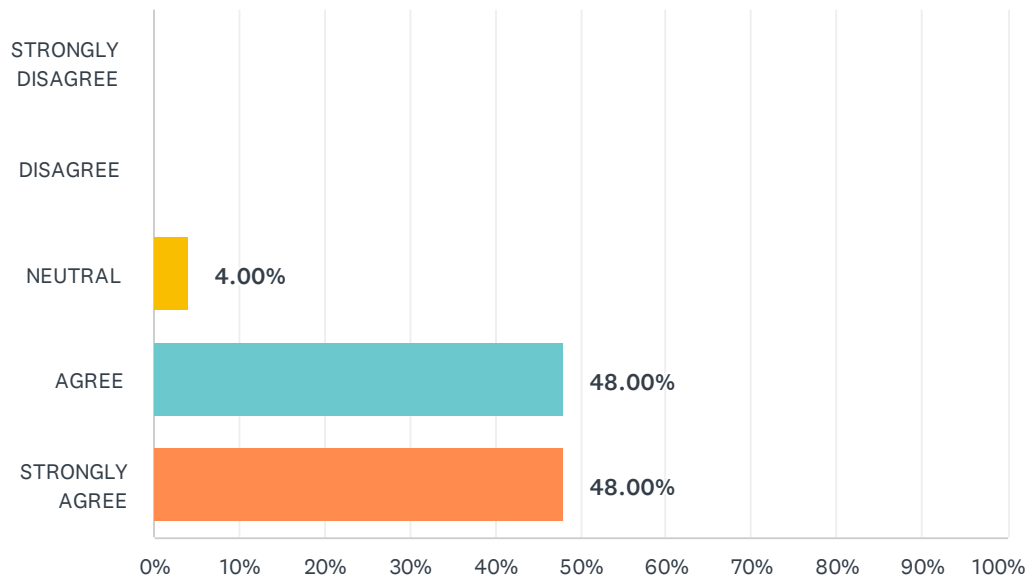
**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	12.00%	3
AGREE (4)	44.00%	11
STRONGLY AGREE (5)	44.00%	11
TOTAL		25

BASIC STATISTICS				
Minimum 3.00	Maximum 5.00	Median 4.00	Mean 4.32	Standard Deviation 0.68

Acute kidney injury can be a concomitant event in SLE and must be treated promptly. Sodium and calcium balance and the use of diuretics are the first approach. Dialysis should be initiated if necessary.

Answered: 25 Skipped: 0



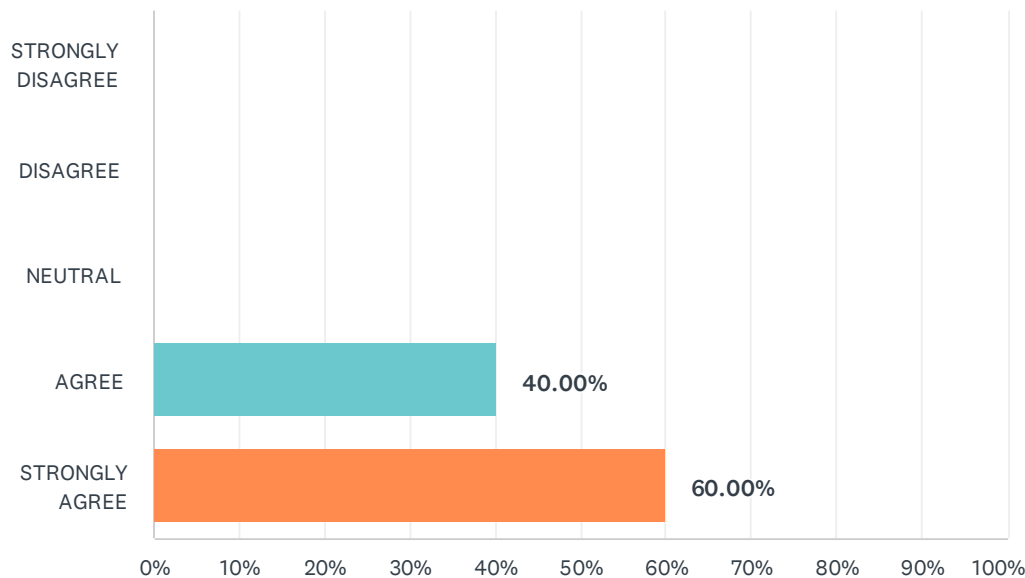
**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	4.00%	1
AGREE (4)	48.00%	12
STRONGLY AGREE (5)	48.00%	12
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.00	4.44	0.57

Rhabdomyolysis is a life-threatening complication that rarely occurs in mitochondrial disorders and is mainly caused by periods of stress, illness, or strenuous activity; it should be carefully managed with intravenous fluids and electrolyte correction to prevent further complications like kidney damage

Answered: 25 Skipped: 0



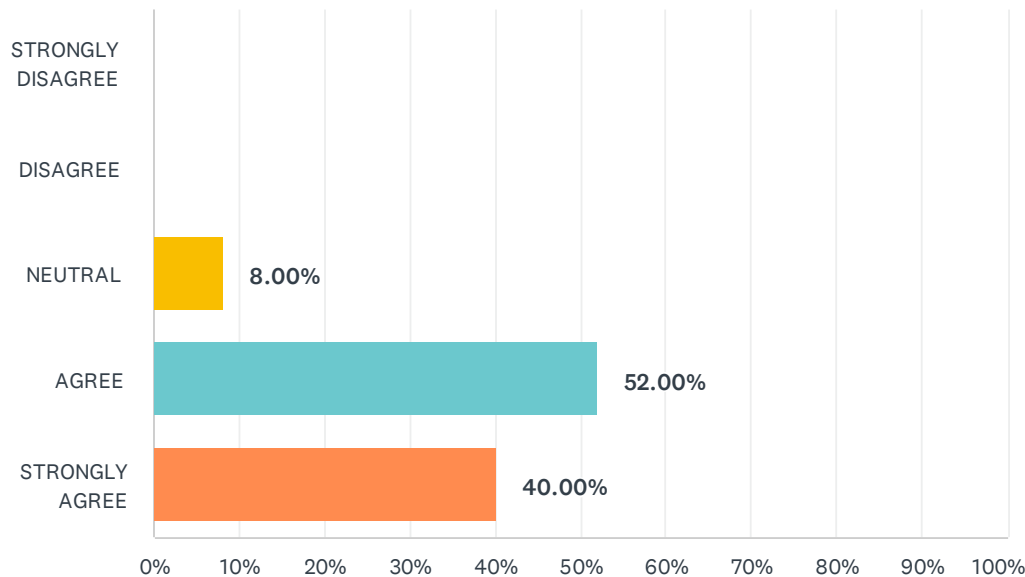
**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	40.00%	10
STRONGLY AGREE (5)	60.00%	15
TOTAL		25

BASIC STATISTICS				
Minimum 4.00	Maximum 5.00	Median 5.00	Mean 4.60	Standard Deviation 0.49

Propofol infusion syndrome is a rare but often fatal condition leading to multiorgan failure. The management of this condition includes the immediate discontinuation of the drug (propofol), hemodialysis and hemodynamic support. The use of propofol for the management of SE in SLEs in MELAS patients should be therefore avoided if possible or limited to short infusions when necessary

Answered: 25 Skipped: 0



**STRONG CONSENSUS**

ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	8.00%	2
AGREE (4)	52.00%	13
STRONGLY AGREE (5)	40.00%	10
TOTAL		25

BASIC STATISTICS				
Minimum 3.00	Maximum 5.00	Median 4.00	Mean 4.32	Standard Deviation 0.61