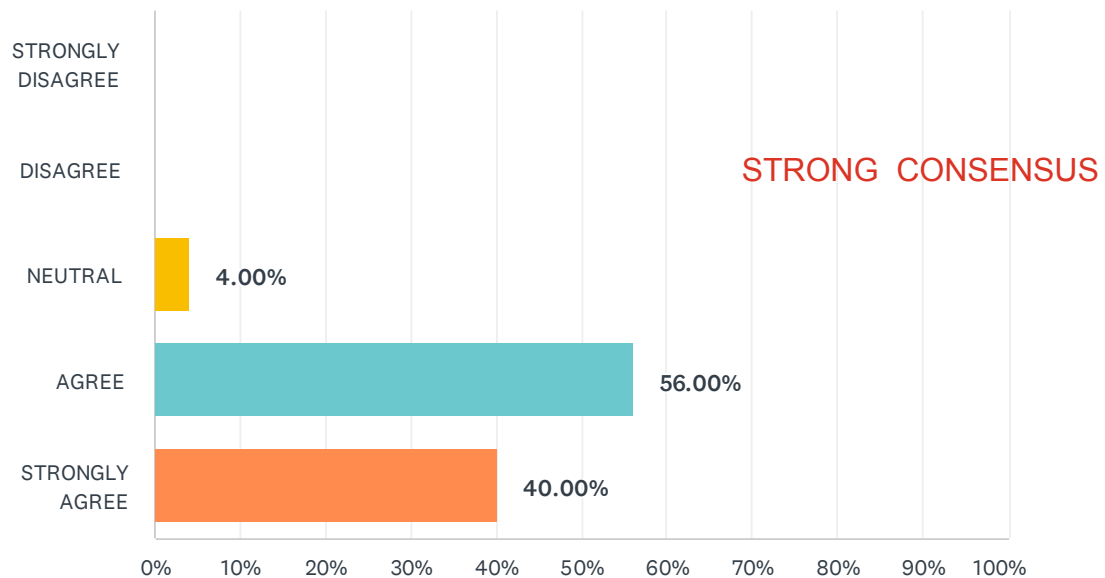


## MELAS DEFINITION

**MELAS is a primary mitochondrial syndrome characterized by one or more stroke-like episodes with associated epileptic and/or encephalopathic features, in the context of either genetic or biochemical or muscle histological evidence of oxidative phosphorylation dysfunction, caused by a pathogenic mtDNA variant.**

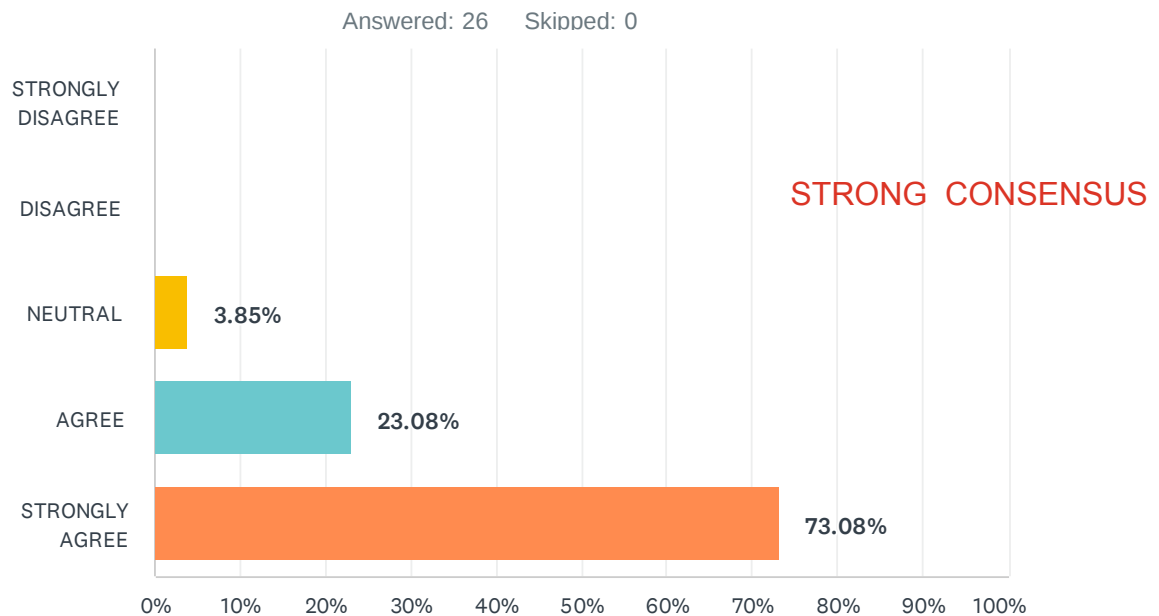
Answered: 25 Skipped: 0



ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	4.00%	1
AGREE (4)	56.00%	14
STRONGLY AGREE (5)	40.00%	10
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.00	4.36	0.56

The following modified 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 with or without EEG abnormalities.

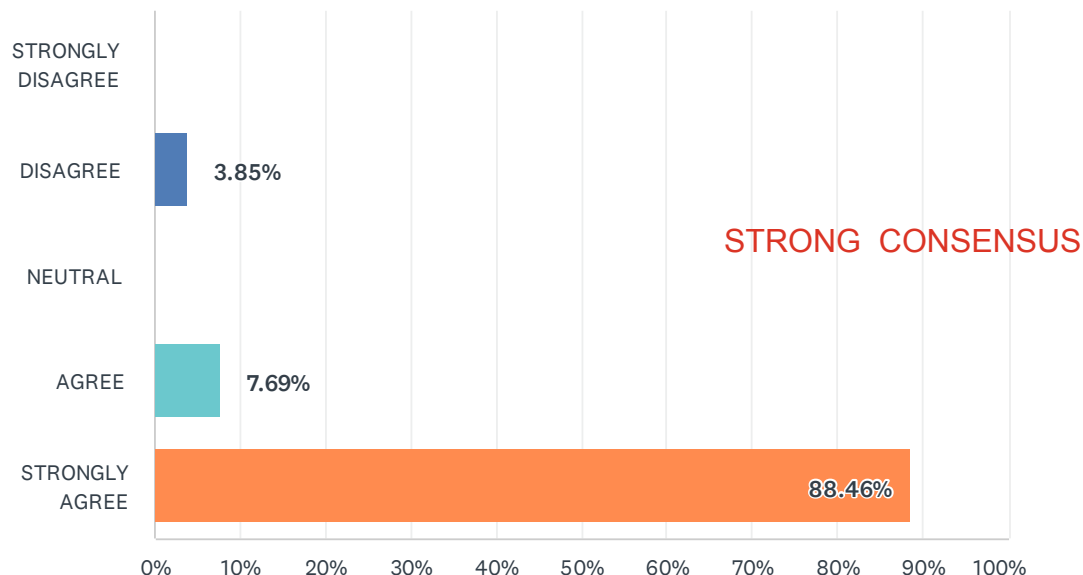


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	3.85%	1
AGREE (4)	23.08%	6
STRONGLY AGREE (5)	73.08%	19
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	5.00	4.69	0.54

SLE can be caused by mutations in either mitochondrial or nuclear DNA and is not limited to the m.3243A>G variant in the MT-TL1 gene.

Answered: 26 Skipped: 0

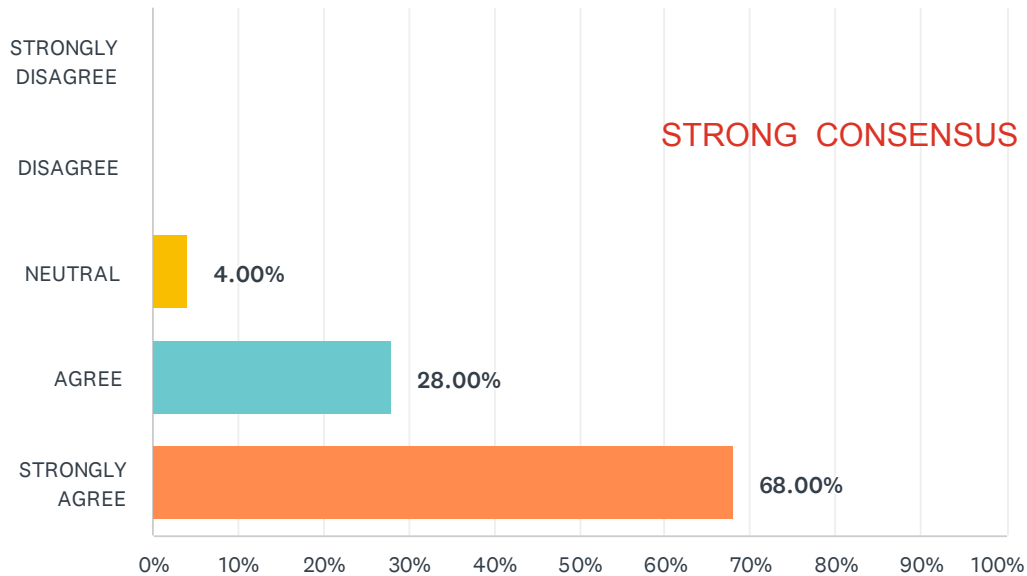


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	3.85%	1
NEUTRAL (3)	0.00%	0
AGREE (4)	7.69%	2
STRONGLY AGREE (5)	88.46%	23
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	5.00	4.81	0.62

MELAS syndrome can be caused by various pathogenic variants in mitochondrial DNA and is not limited to the m.3243A>G variant in the MT-TL1 gene.

Answered: 25 Skipped: 0

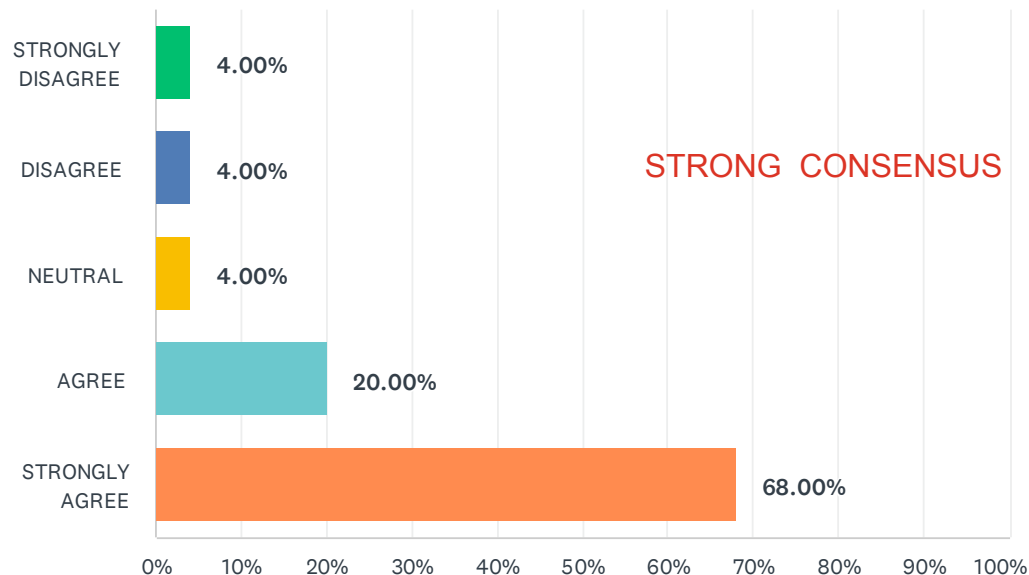


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	4.00%	1
AGREE (4)	28.00%	7
STRONGLY AGREE (5)	68.00%	17
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	5.00	4.64	0.56

The term MELAS—an acronym for Mitochondrial Encephalomyopathy, Lactic Acidosis, and Stroke-like episodes—is a traditional diagnostic label. A full phenotype may not always be present; however, for the syndrome to be defined as MELAS, a stroke-like episode must occur.

Answered: 25 Skipped: 0

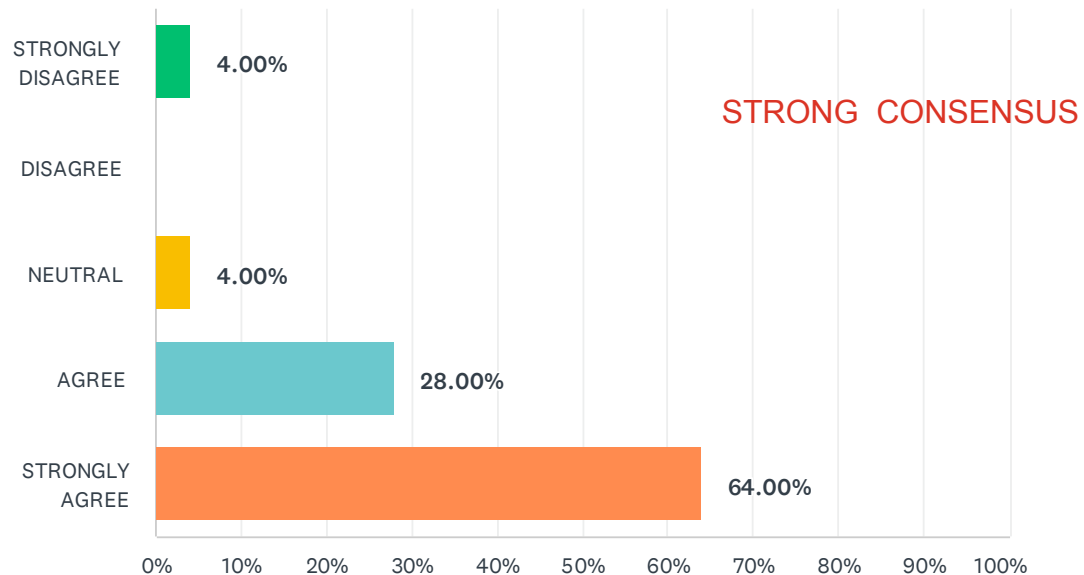


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	4.00%	1
DISAGREE (2)	4.00%	1
NEUTRAL (3)	4.00%	1
AGREE (4)	20.00%	5
STRONGLY AGREE (5)	68.00%	17
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	5.00	4.44	1.02

MELAS syndrome is a well-defined clinical entity; therefore, terms such as 'MELAS-like' or 'MELAS spectrum' should be avoided.

Answered: 25 Skipped: 0

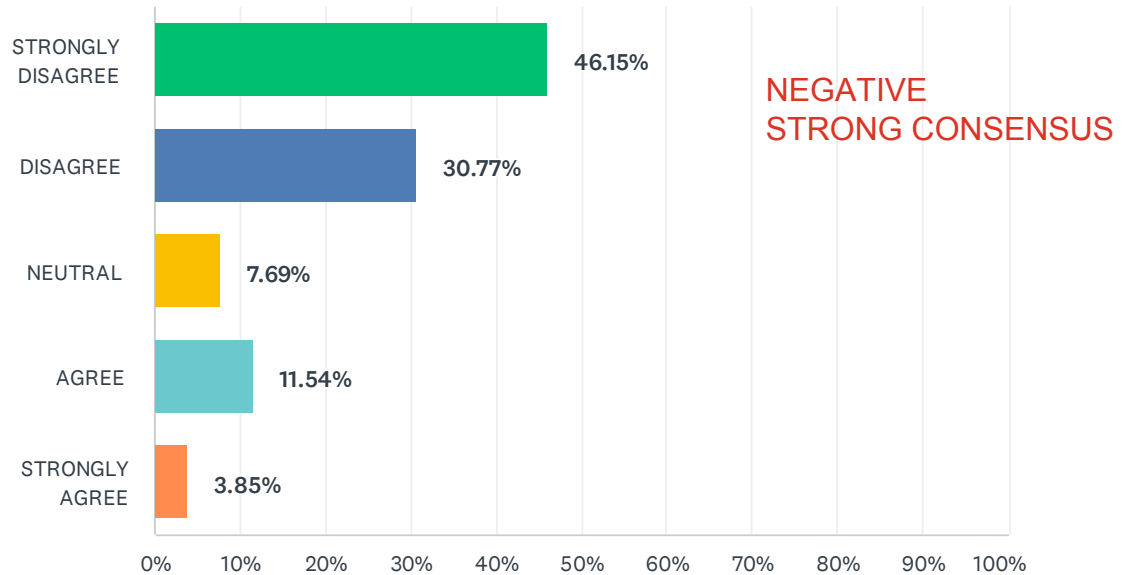


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	4.00%	1
DISAGREE (2)	0.00%	0
NEUTRAL (3)	4.00%	1
AGREE (4)	28.00%	7
STRONGLY AGREE (5)	64.00%	16
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	5.00	4.48	0.90

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

Answered: 26 Skipped: 0

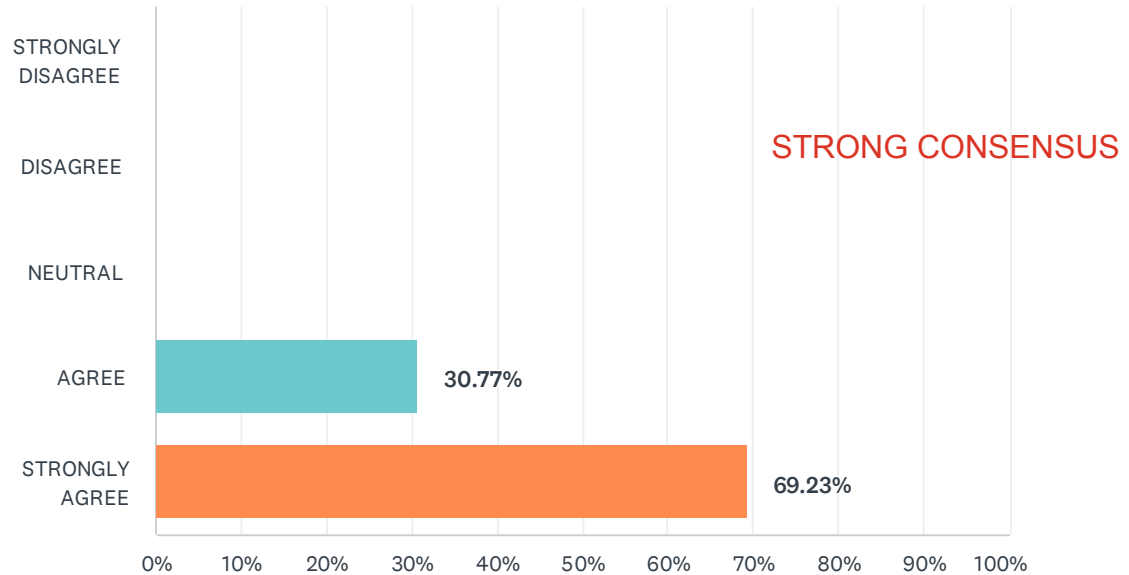


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	46.15%	12
DISAGREE (2)	30.77%	8
NEUTRAL (3)	7.69%	2
AGREE (4)	11.54%	3
STRONGLY AGREE (5)	3.85%	1
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	2.00	1.96	1.16

## The efficacy of intravenous L-arginine is unproven in the acute phase of SLE.

Answered: 26 Skipped: 0



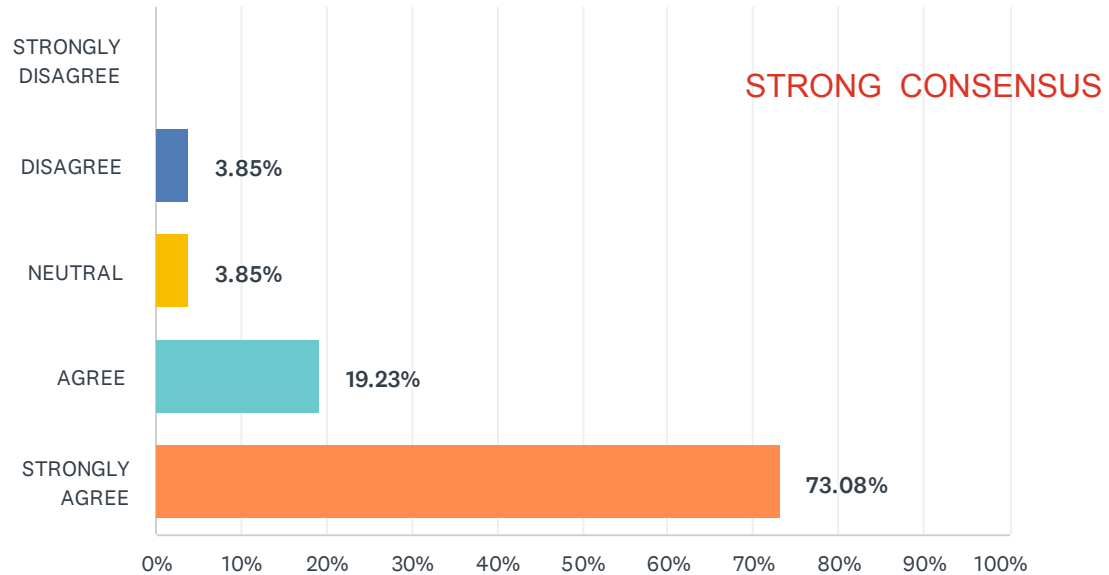
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	30.77%	8
STRONGLY AGREE (5)	69.23%	18
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
4.00	5.00	5.00	4.69	0.46



## Randomised controlled clinical trials are needed to assess the efficacy of intravenous L-Arginine in the acute phase of SLE management

Answered: 26 Skipped: 0

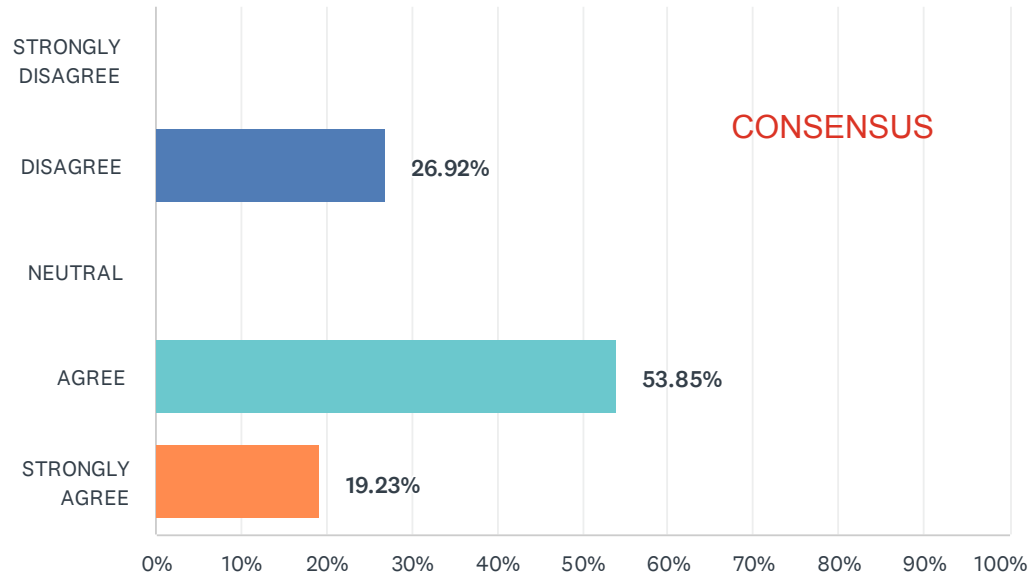


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	3.85%	1
NEUTRAL (3)	3.85%	1
AGREE (4)	19.23%	5
STRONGLY AGREE (5)	73.08%	19
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	5.00	4.62	0.74

## The use of intravenous L-arginine in the acute phase of SLE is considered safe on a case by case basis

Answered: 26 Skipped: 0

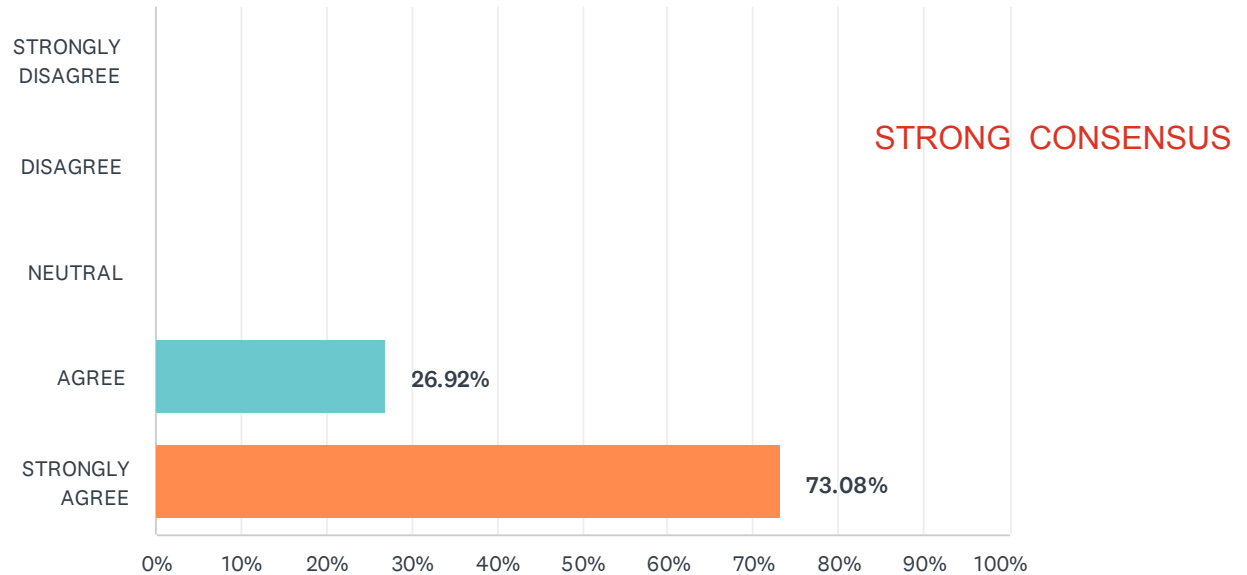


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	26.92%	7
NEUTRAL (3)	0.00%	0
AGREE (4)	53.85%	14
STRONGLY AGREE (5)	19.23%	5
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	3.65	1.07

## The efficacy of oral L-arginine as long-term treatment to reduce SLE frequency in unproven

Answered: 26 Skipped: 0

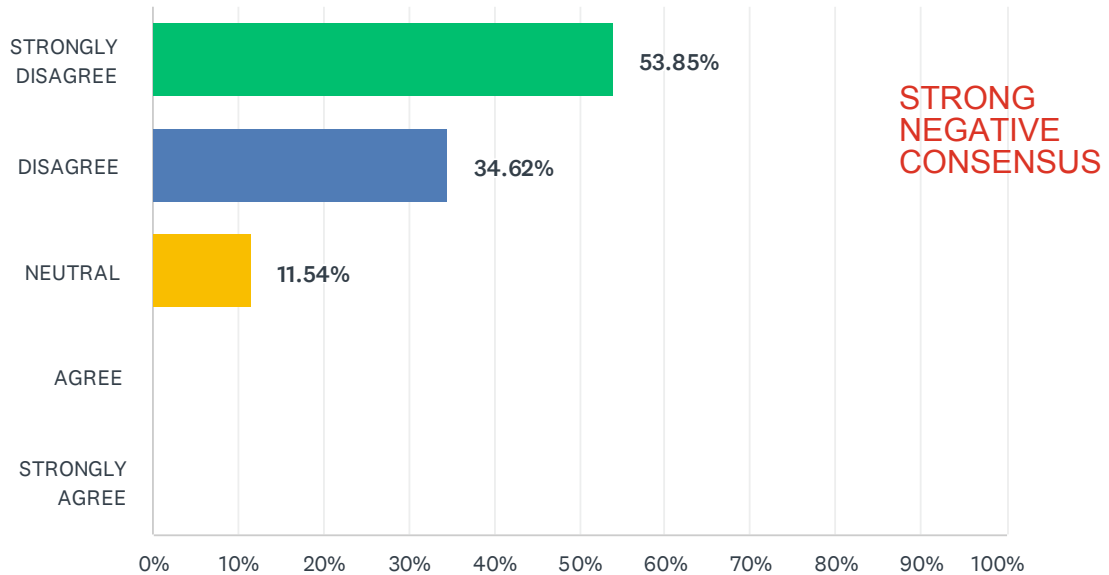


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	26.92%	7
STRONGLY AGREE (5)	73.08%	19
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
4.00	5.00	5.00	4.73	0.44

## Oral Taurine should be used as a long-term medication to reduce SLE frequency in MT-TL1 gene related MELAS

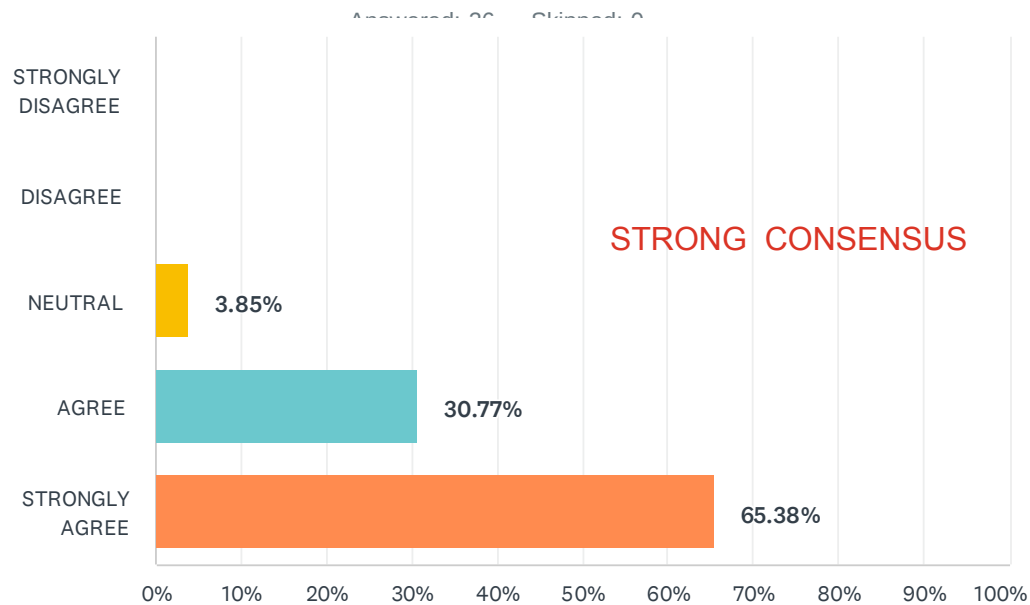
Answered: 26 Skipped: 0



ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	53.85%	14
DISAGREE (2)	34.62%	9
NEUTRAL (3)	11.54%	3
AGREE (4)	0.00%	0
STRONGLY AGREE (5)	0.00%	0
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	3.00	1.00	1.58	0.69

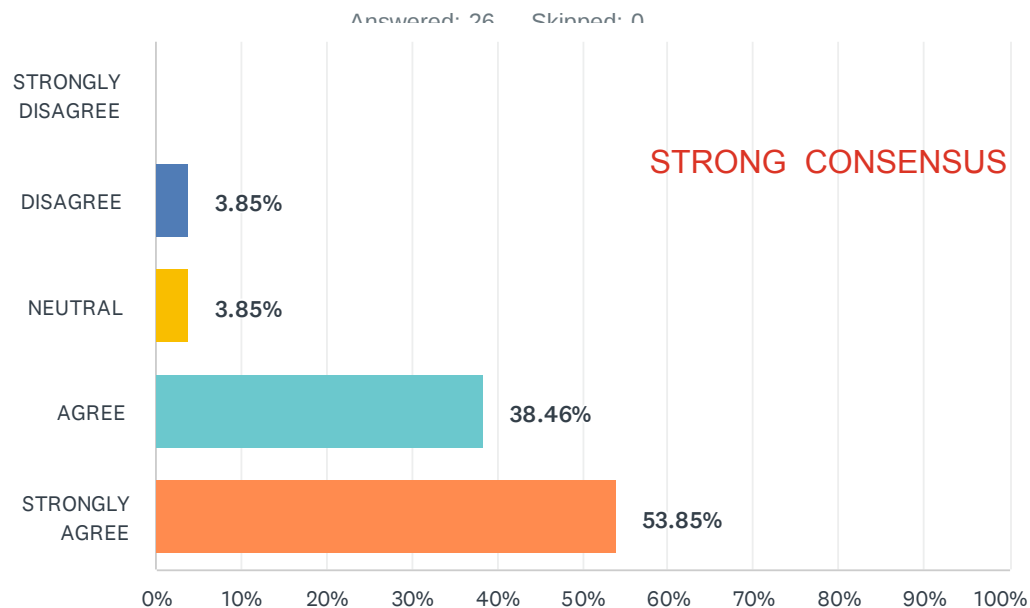
The efficacy of oral taurine as long-term treatment to reduce SLE frequency in MT-TL1 gene related MELAS is unproven



ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	3.85%	1
AGREE (4)	30.77%	8
STRONGLY AGREE (5)	65.38%	17
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	5.00	4.62	0.56

## More data are needed to investigate efficacy and safety of long-term oral taurine supplementation in tRNA LEU UUR related MELAS

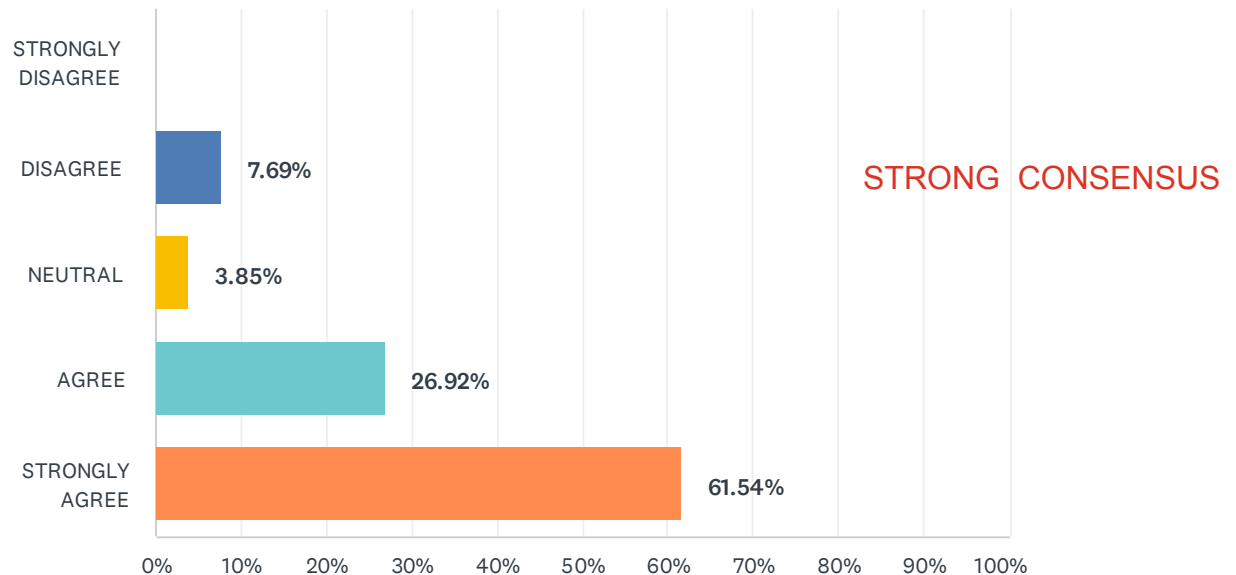


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	3.85%	1
NEUTRAL (3)	3.85%	1
AGREE (4)	38.46%	10
STRONGLY AGREE (5)	53.85%	14
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	5.00	4.42	0.74

## ASMs should be used in SLE if there is a strong clinical suspicion of seizure activity, irrespective of EEG findings.

Answered: 26 Skipped: 0

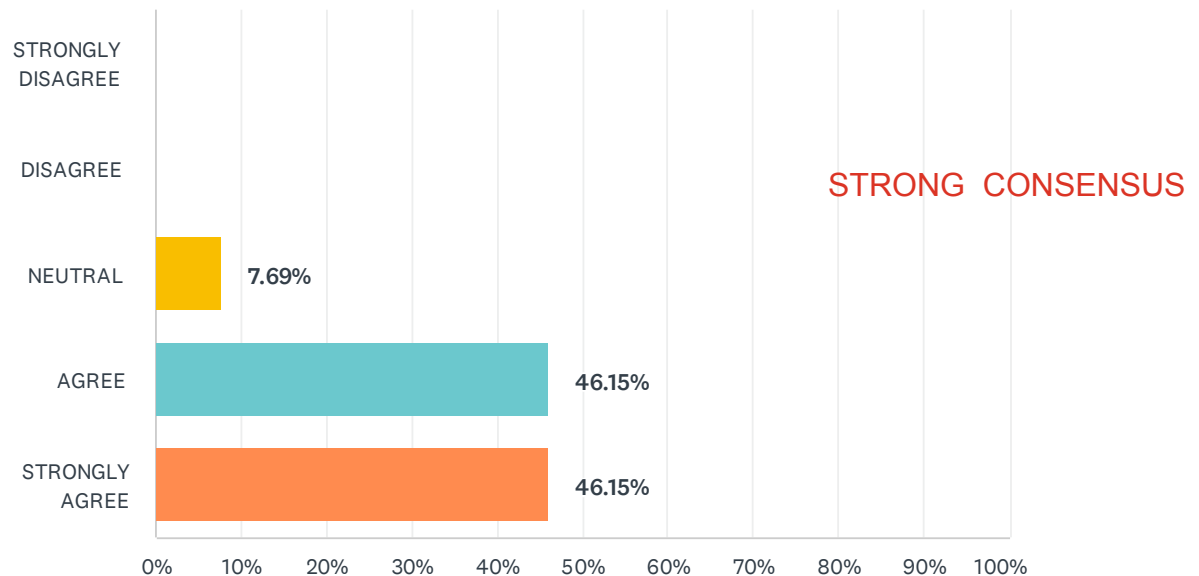


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	7.69%	2
NEUTRAL (3)	3.85%	1
AGREE (4)	26.92%	7
STRONGLY AGREE (5)	61.54%	16
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	5.00	4.42	0.88

## Anti-seizure medications are important in treating stroke-like episodes or acute encephalopathic episodes, including its possible psychotic manifestations.

Answered: 26 Skipped: 0



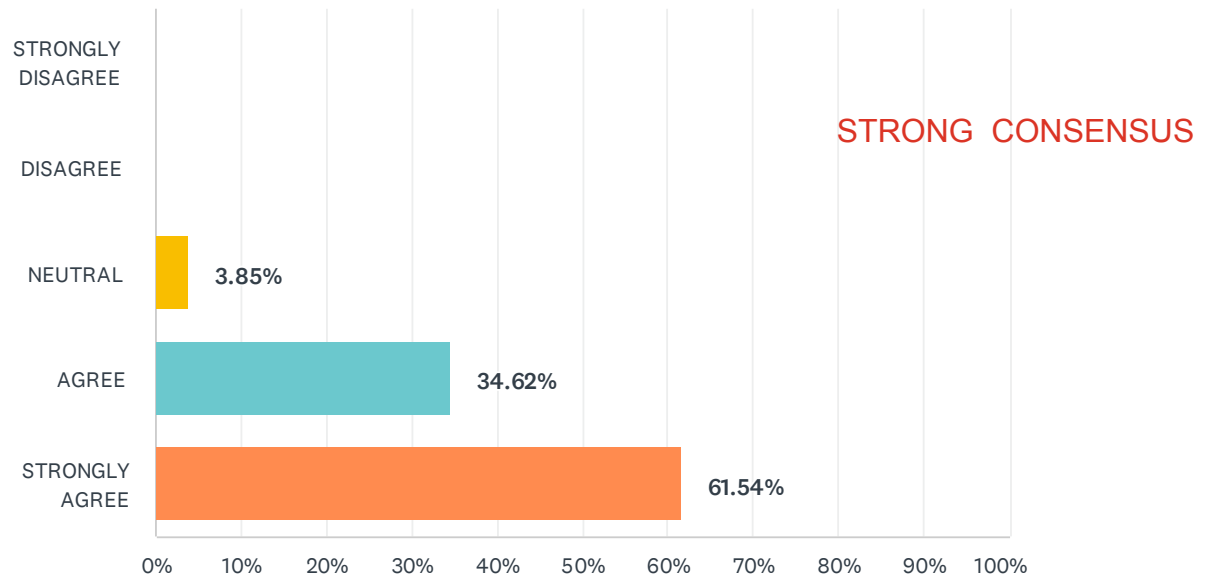
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	7.69%	2
AGREE (4)	46.15%	12
STRONGLY AGREE (5)	46.15%	12
TOTAL		26

BASIC STATISTICS				
Minimum 3.00	Maximum 5.00	Median 4.00	Mean 4.38	Standard Deviation 0.62



## Antipsychotic drugs can be used to manage acute psychotic symptoms associated with stroke-like episodes.

Answered: 26 Skipped: 0

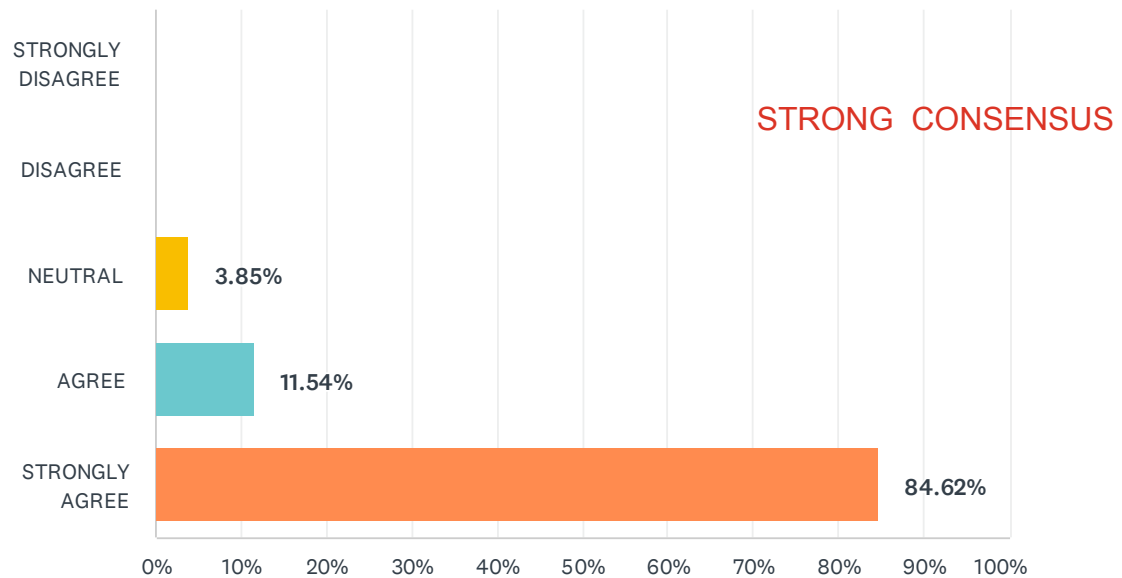


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	3.85%	1
AGREE (4)	34.62%	9
STRONGLY AGREE (5)	61.54%	16
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	5.00	4.58	0.57

There is no rationale nor evidence that dietary supplements are useful to manage acute psychotic symptoms associated with stroke-like/acute encephalopathic episodes.

Answered: 26 Skipped: 0

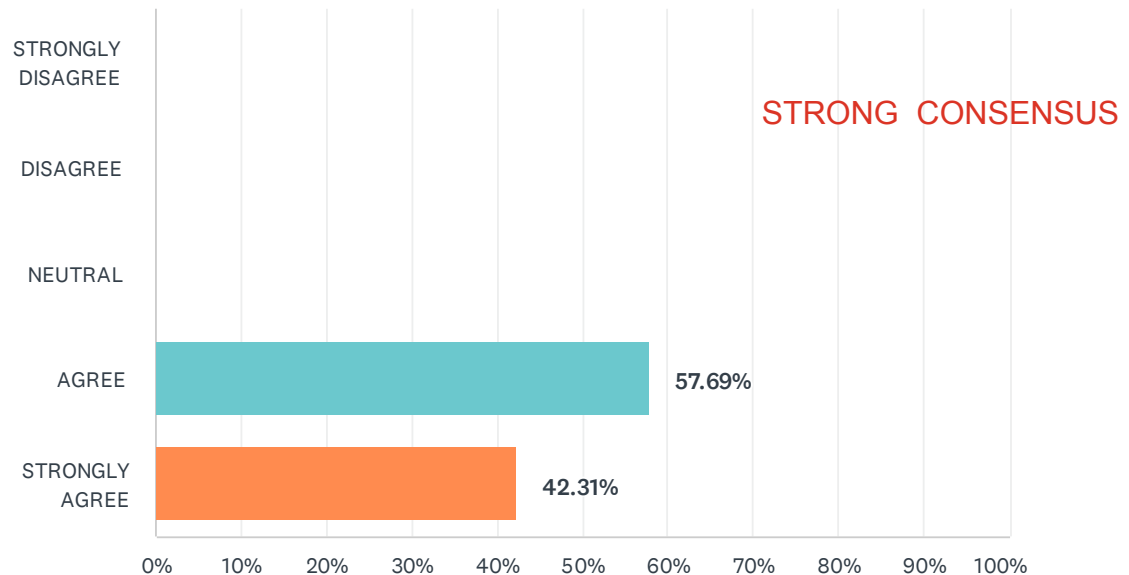


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	3.85%	1
AGREE (4)	11.54%	3
STRONGLY AGREE (5)	84.62%	22
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	5.00	4.81	0.48

## Cognitive rehabilitation should be considered to treat cognitive symptoms in MELAS

Answered: 26 Skipped: 0

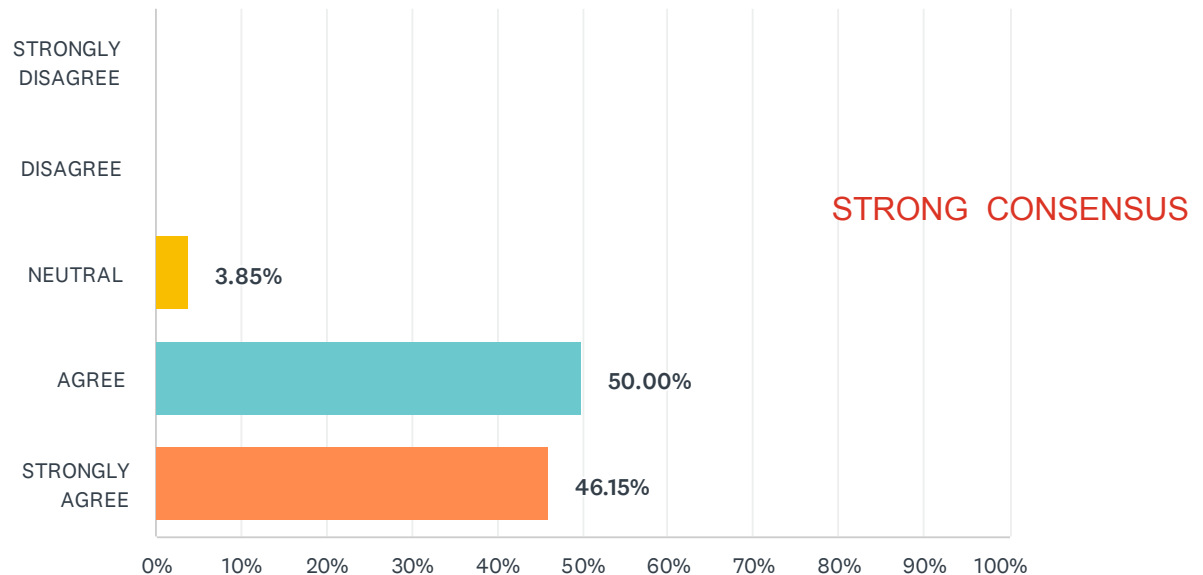


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	57.69%	15
STRONGLY AGREE (5)	42.31%	11
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
4.00	5.00	4.00	4.42	0.49

## SSRI and SNRI should be used in patients with MELAS according to best psychiatric practices to treat depression and anxiety

Answered: 26 Skipped: 0

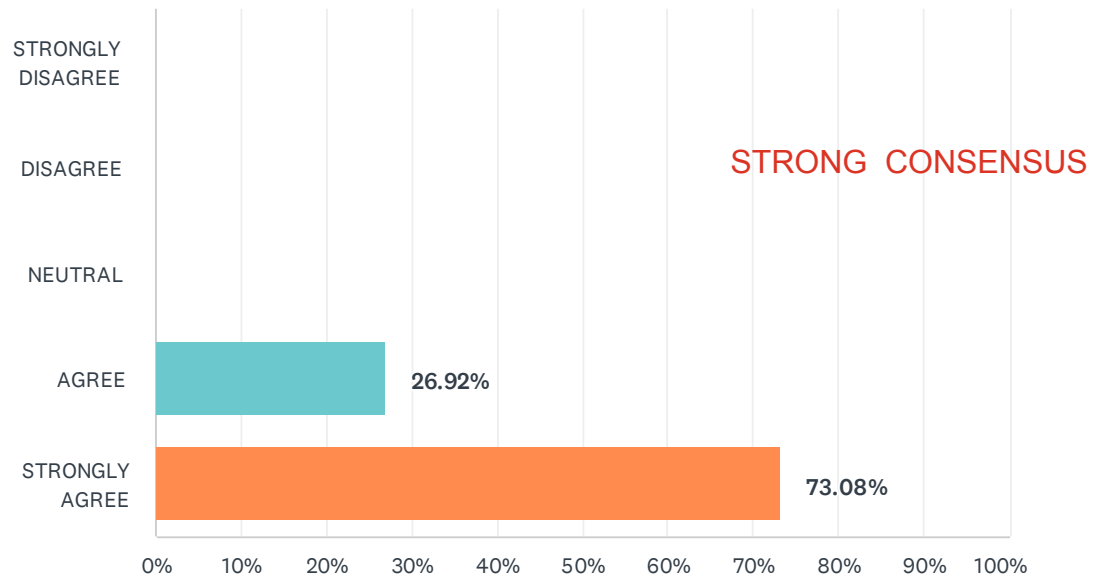


ANSWER CHOICES		RESPONSES	
STRONGLY DISAGREE (1)		0.00%	0
DISAGREE (2)		0.00%	0
NEUTRAL (3)		3.85%	1
AGREE (4)		50.00%	13
STRONGLY AGREE (5)		46.15%	12
TOTAL			26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.00	4.42	0.57

## Cognitive deficits are common in patients with MELAS. Multi-domain cognitive assessments are recommended.

Answered: 26 Skipped: 0

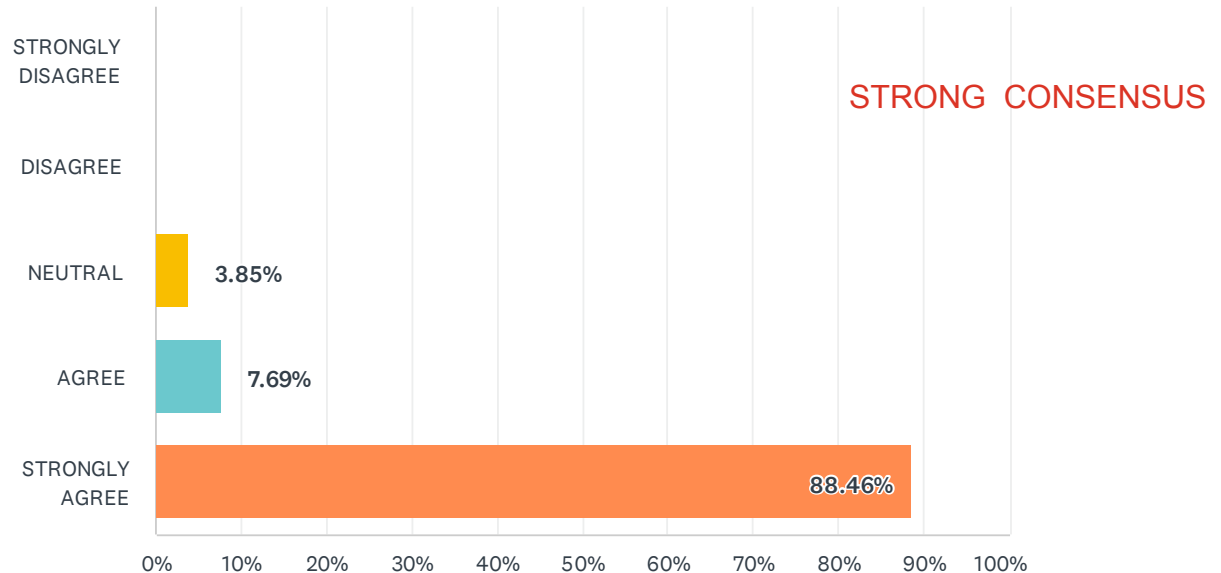


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	26.92%	7
STRONGLY AGREE (5)	73.08%	19
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
4.00	5.00	5.00	4.73	0.44

## NEW There is currently no evidence demonstrating that oral citrulline is effective in the acute phase of SLE.

Answered: 26 Skipped: 0

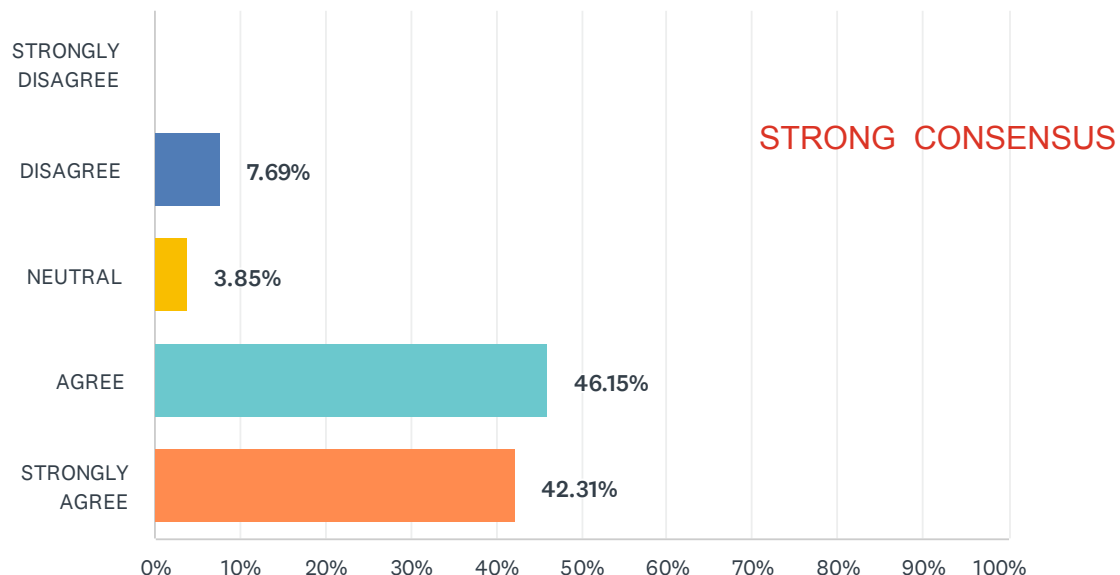


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	3.85%	1
AGREE (4)	7.69%	2
STRONGLY AGREE (5)	88.46%	23
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	5.00	4.85	0.46

## NEW Future clinical trial is required to evaluate the efficacy of oral citrulline as long-term treatment to reduce SLE frequency

Answered: 26 Skipped: 0

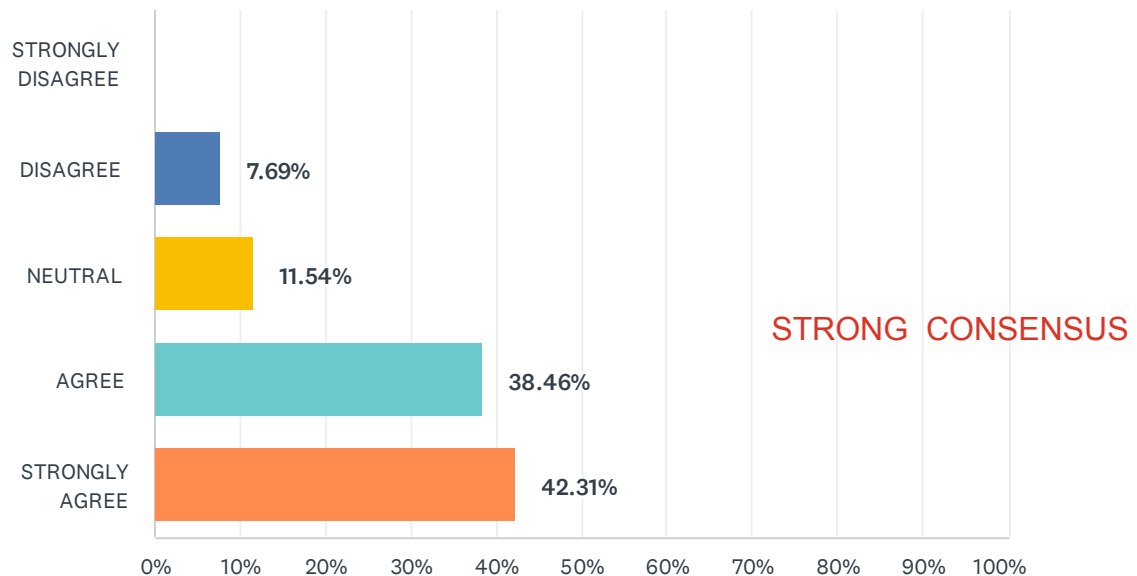


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	7.69%	2
NEUTRAL (3)	3.85%	1
AGREE (4)	46.15%	12
STRONGLY AGREE (5)	42.31%	11
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	4.23	0.85

Based on the available experience, the use of i.v. steroids (either methylprednisolone or dexamethasone) may have a beneficial effect in the acute phase of SLE

Answered: 26 Skipped: 0



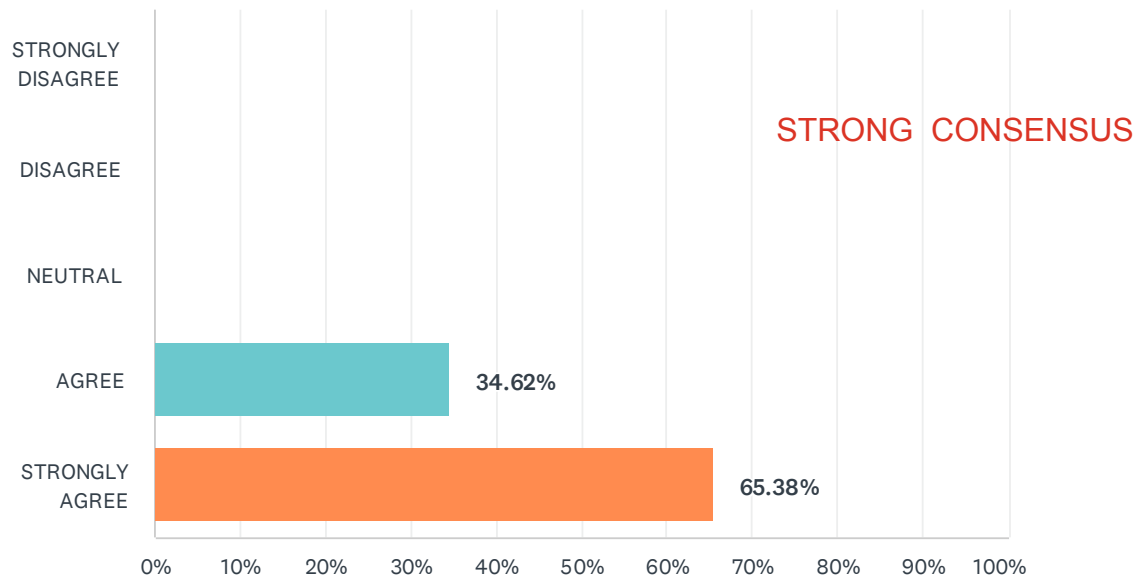
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	7.69%	2
NEUTRAL (3)	11.54%	3
AGREE (4)	38.46%	10
STRONGLY AGREE (5)	42.31%	11
TOTAL		26

BASIC STATISTICS				
Minimum 2.00	Maximum 5.00	Median 4.00	Mean 4.15	Standard Deviation 0.91



**NEW: There is no evidence that coenzyme Q10 formulation or its analogues, vitamins or other dietary supplements have a beneficial effect in the acute management of SLE.**

Answered: 26 Skipped: 0

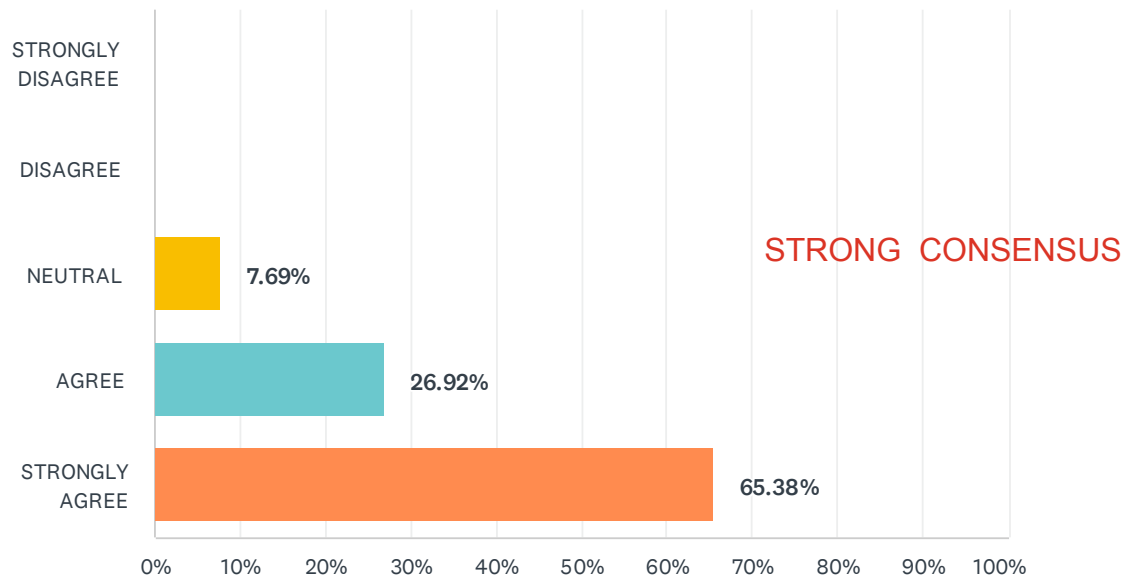


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	34.62%	9
STRONGLY AGREE (5)	65.38%	17
TOTAL		26

BASIC STATISTICS				
Minimum 4.00	Maximum 5.00	Median 5.00	Mean 4.65	Standard Deviation 0.48

There is no evidence that coenzyme Q10 formulation or its analogues, vitamins or other dietary supplements have a beneficial effect in long-term treatment to prevent SLE

Answered: 26 Skipped: 0

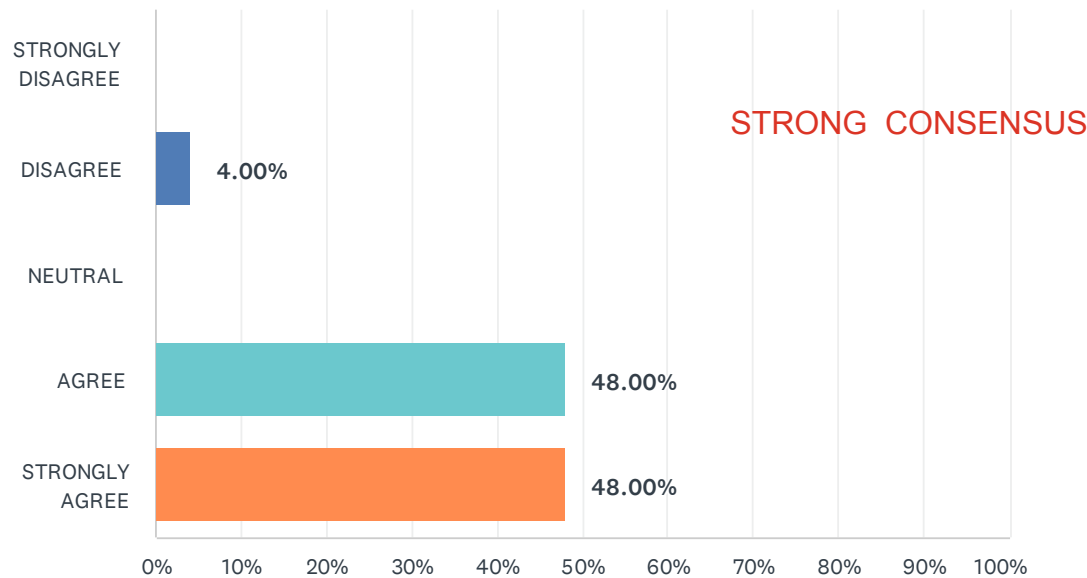


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	7.69%	2
AGREE (4)	26.92%	7
STRONGLY AGREE (5)	65.38%	17
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	5.00	4.58	0.63

There is no known contraindication to the use of monoclonal antibodies for the prevention of migraine in patients with MELAS, but their use should be carefully monitored until more evidence is accumulated.

Answered: 25    Skipped: 0

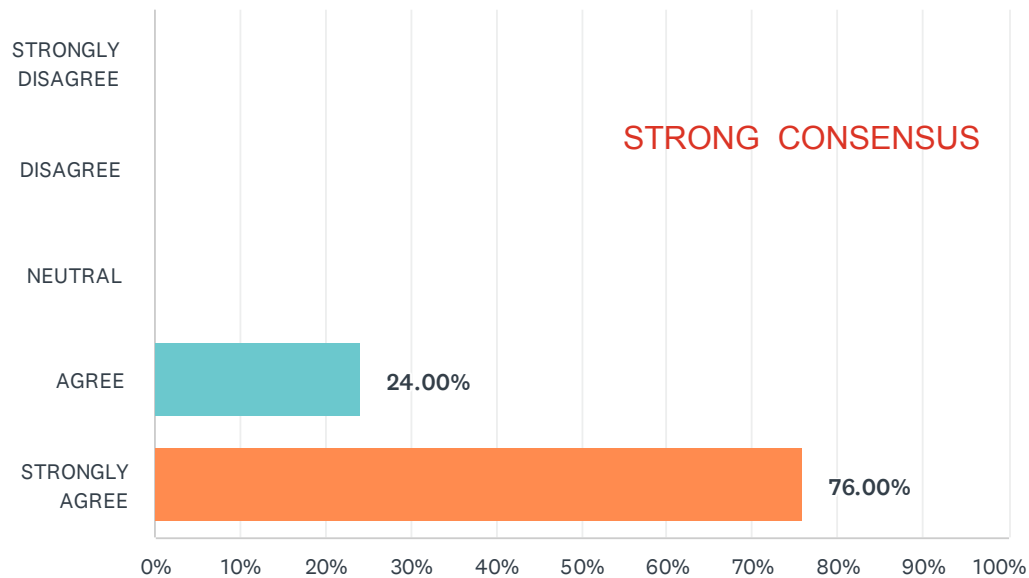


ANSWER CHOICES		RESPONSES	
STRONGLY DISAGREE (1)		0.00%	0
DISAGREE (2)		4.00%	1
NEUTRAL (3)		0.00%	0
AGREE (4)		48.00%	12
STRONGLY AGREE (5)		48.00%	12
TOTAL			25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	4.40	0.69

Stroke like episodes in MELAS may be accompanied by multi-organ failure. Cardiac, renal, and gastrointestinal function should be assessed on admission and during the course of any inpatient stay.

Answered: 25 Skipped: 0

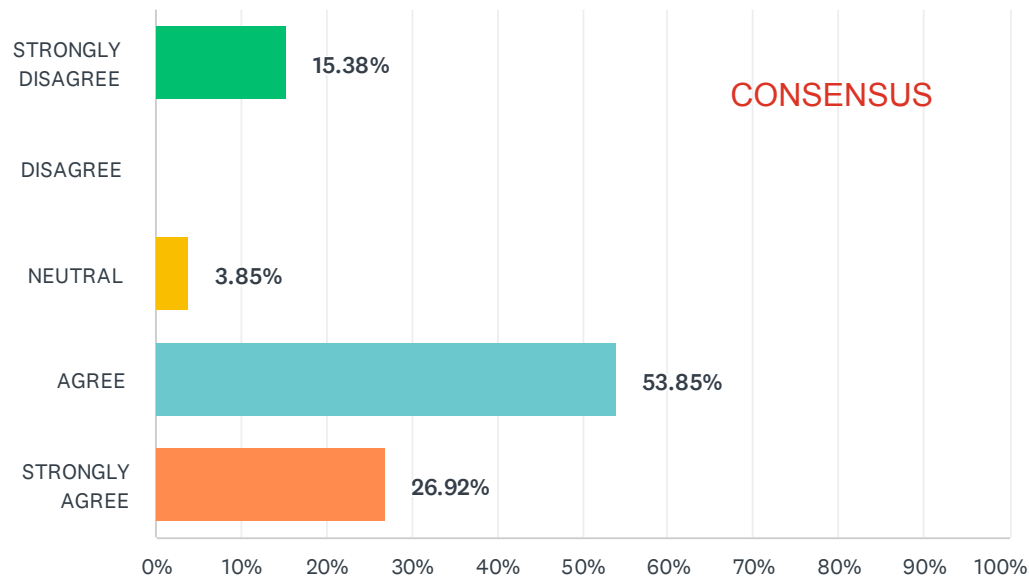


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	24.00%	6
STRONGLY AGREE (5)	76.00%	19
TOTAL		25

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
4.00	5.00	5.00	4.76	0.43

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

Answered: 26 Skipped: 0

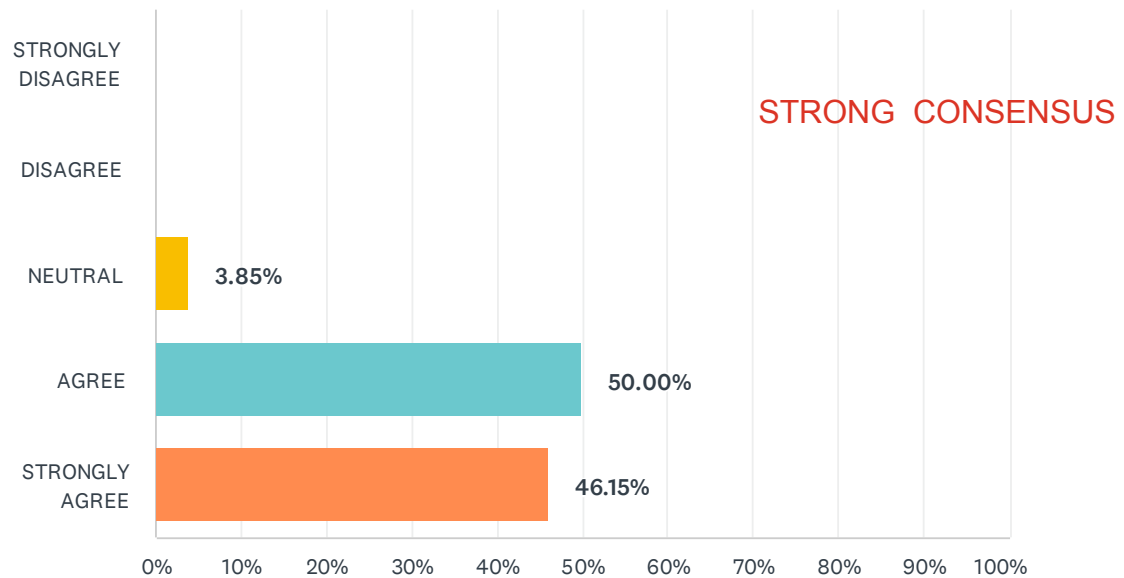


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	15.38%	4
DISAGREE (2)	0.00%	0
NEUTRAL (3)	3.85%	1
AGREE (4)	53.85%	14
STRONGLY AGREE (5)	26.92%	7
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
1.00	5.00	4.00	3.77	1.28

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

Answered: 26 Skipped: 0

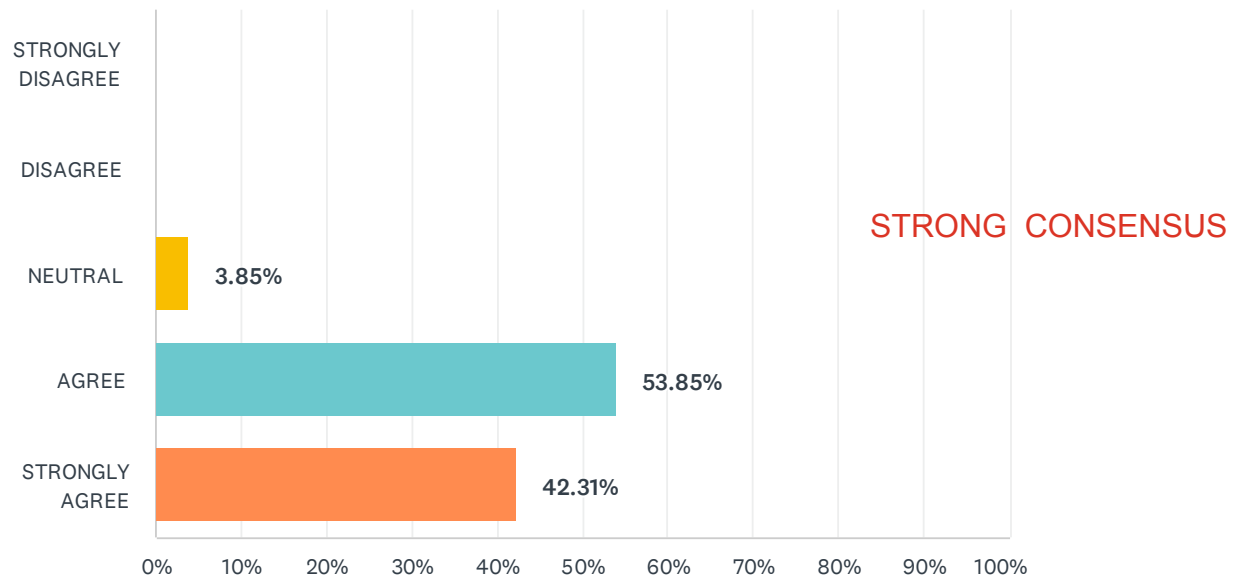


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	3.85%	1
AGREE (4)	50.00%	13
STRONGLY AGREE (5)	46.15%	12
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.00	4.42	0.57

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

Answered: 26 Skipped: 0

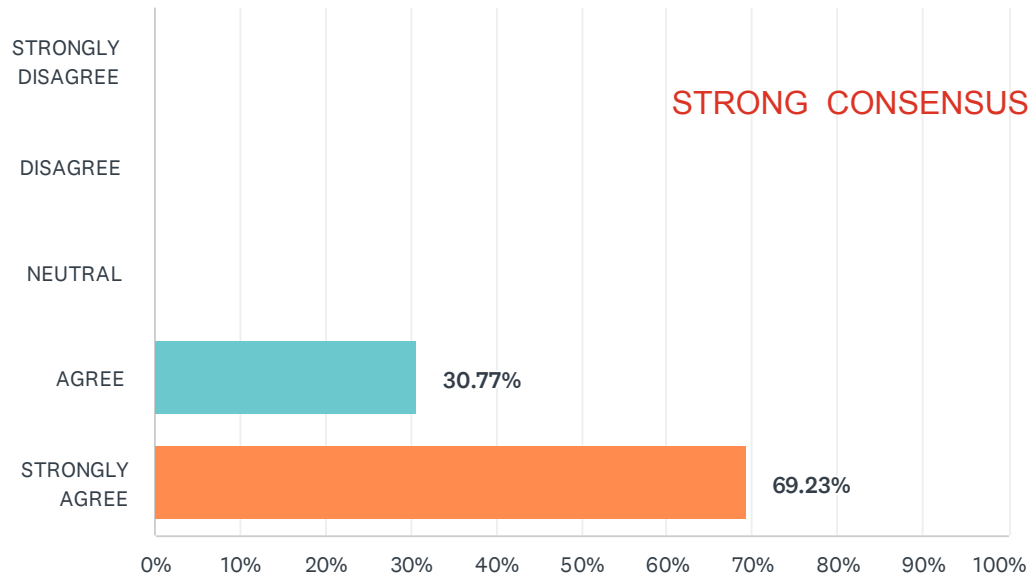


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	3.85%	1
AGREE (4)	53.85%	14
STRONGLY AGREE (5)	42.31%	11
TOTAL		26

BASIC STATISTICS				
Minimum 3.00	Maximum 5.00	Median 4.00	Mean 4.38	Standard Deviation 0.56

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

Answered: 26 Skipped: 0



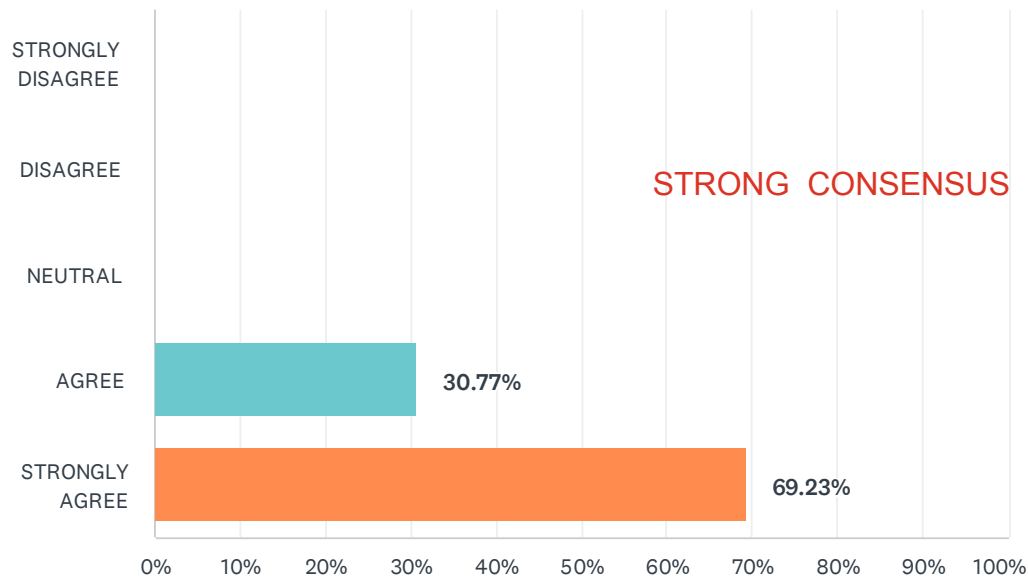
ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	30.77%	8
STRONGLY AGREE (5)	69.23%	18
TOTAL		26

BASIC STATISTICS				
Minimum 4.00	Maximum 5.00	Median 5.00	Mean 4.69	Standard Deviation 0.46



Cardiac symptoms including cardiomyopathy, cardiac arrhythmia, acute cardiac failure, and cardiogenic shock can occur in the setting of SLE and MELAS. Patients should be closely monitored for these complications

Answered: 26 Skipped: 0

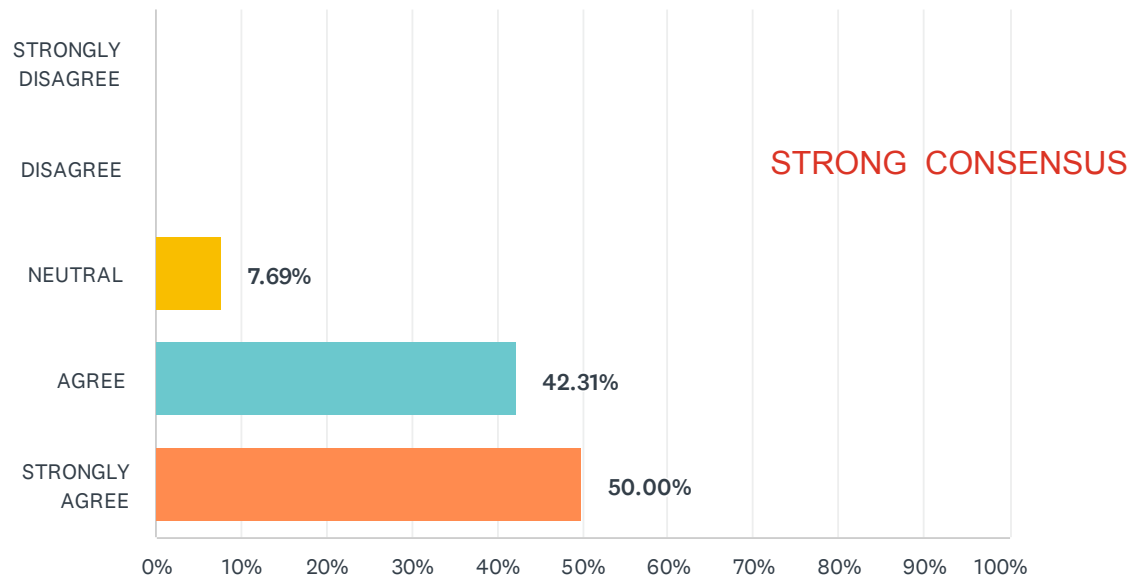


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	30.77%	8
STRONGLY AGREE (5)	69.23%	18
TOTAL		26

BASIC STATISTICS				
Minimum 4.00	Maximum 5.00	Median 5.00	Mean 4.69	Standard Deviation 0.46

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. In case of severe lactic acidosis, we recommend appropriate hydrations with fluids, avoiding high dextrose concentration and using bicarbonate in case of severe base deficit.

Answered: 26 Skipped: 0

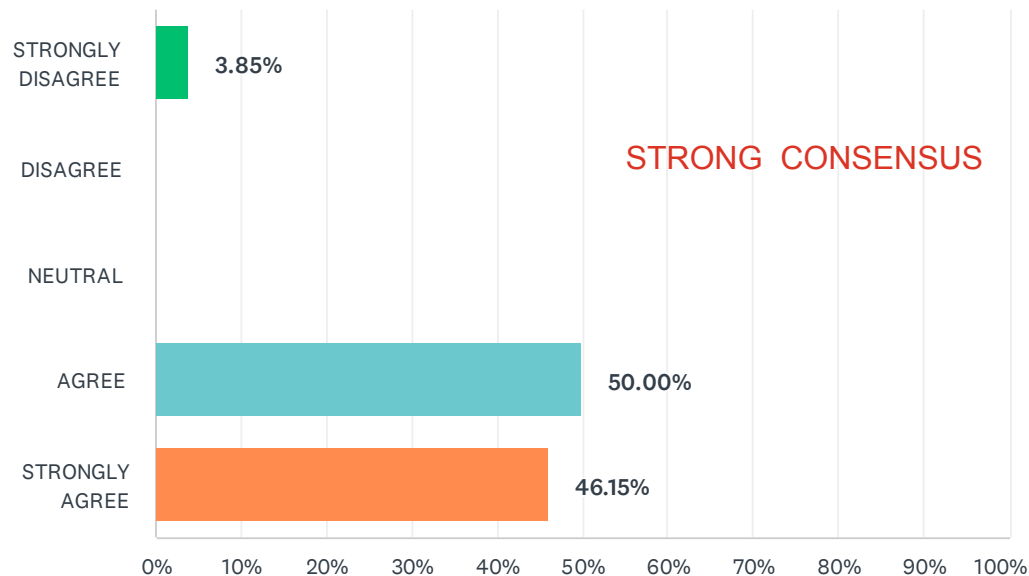


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	0.00%	0
NEUTRAL (3)	7.69%	2
AGREE (4)	42.31%	11
STRONGLY AGREE (5)	50.00%	13
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
3.00	5.00	4.50	4.42	0.63

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

Answered: 26 Skipped: 0

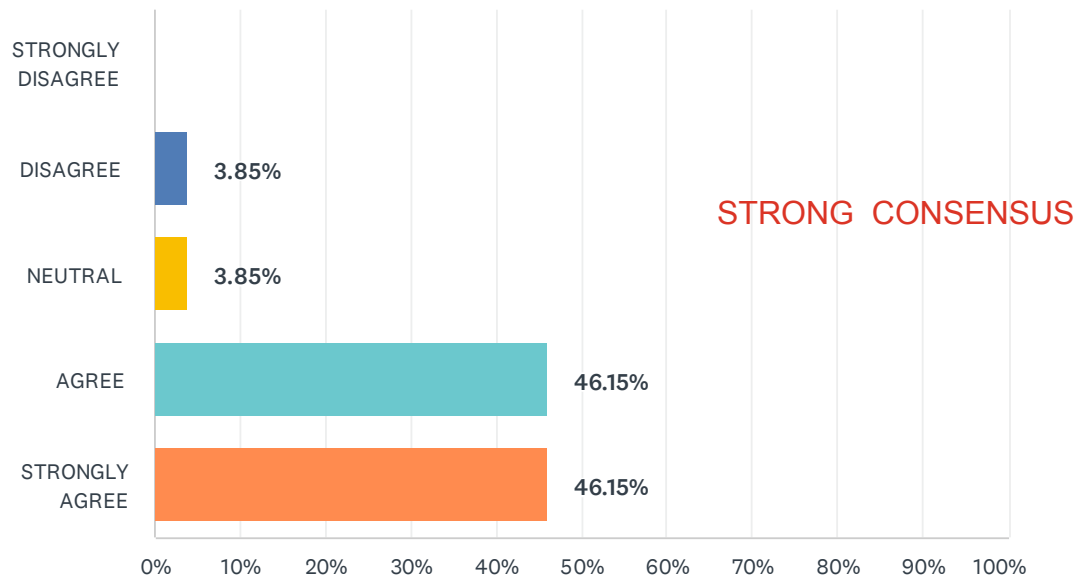


ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	3.85%	1
DISAGREE (2)	0.00%	0
NEUTRAL (3)	0.00%	0
AGREE (4)	50.00%	13
STRONGLY AGREE (5)	46.15%	12
TOTAL		26

BASIC STATISTICS				
Minimum 1.00	Maximum 5.00	Median 4.00	Mean 4.35	Standard Deviation 0.83

In MELAS Rhabdomyolysis is an extremely rare life-threatening complication; it should be carefully managed with intravenous fluids and electrolyte correction to prevent further complications like kidney damage

Answered: 26 Skipped: 0



ANSWER CHOICES	RESPONSES	
STRONGLY DISAGREE (1)	0.00%	0
DISAGREE (2)	3.85%	1
NEUTRAL (3)	3.85%	1
AGREE (4)	46.15%	12
STRONGLY AGREE (5)	46.15%	12
TOTAL		26

BASIC STATISTICS				
Minimum	Maximum	Median	Mean	Standard Deviation
2.00	5.00	4.00	4.35	0.73