
THE ERYTHROMELALGIA ASSOCIATION 2008 MEMBER STUDY

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Objectives and Methods

Objectives

- To increase the understanding of Erythromelalgia
- To engage membership in advancing the research of the disease
- To share treatment results and ideas among current and future members, physicians and researchers

Study Methods

- Survey questionnaire developed jointly with TEA members.
- Survey was deployed online and on paper. Invitations were emailed to 646 members, with two subsequent reminders. Another 81 paper surveys were mailed to those without email.
- Emails were corrected and resent. An additional 85 surveys were mailed to those who might not have received the emails.
- Survey responses (paper & Internet) were received from 6/04/08 through 10/13/08. A total of 427 responses were received (357 via Internet, 70 paper). The response rate was 66%.

Summary

- Respondents to this survey were:
 - ☐ 96% Caucasian
 - ☐ 80% female
 - ☐ 77% within the age range of 41-80
- EM is very distinctive
 - ☐ 99% of sufferers experience flares, primarily in their feet (95%), but also in their hands (54%) and/or face (31%)
 - ☐ Of the 95% with foot flares:
 - 40% flare only in their feet
 - 29% flare only in feet and hands
 - 25% flare in all three: feet, hands and face
 - 6% flare only in feet and face
 - ☐ 93% experience burning and redness in affected areas
 - ☐ 96% experience regular flares if their environment is not controlled
 - ☐ 85% experience pain at the skin level and 56% also feel pain deep into their body
 - ☐ 97% feel their condition is exacerbated by weather conditions, primarily heat
 - ☐ 97% have been diagnosed with EM by a doctor; 58% of these credit themselves with helping the doctor determine the diagnosis
 - ☐ 87% feel certain things trigger flares [of those answering yes: 82% identified exercise, 49% taking a shower, 45% elevated emotions, 44% alcohol, 36% certain foods]
- Yet the disease appears to follow few patterns
 - ☐ Any age person can be affected
 - ☐ Symptoms start across all ages about evenly
 - ☐ Only 28% report symptoms starting with a defining event
 - ☐ 57% report symptoms steadily worsening, but 17% report symptoms improving over time
 - ☐ 64% report no period of even a few days in which they have been symptom-free
 - ☐ 60% indicate at least some ability to control their symptoms, mostly by altering the temperature or airflow of their environment
 - ☐ 53% have a separate condition which accompanies their EM, 34% do not
 - ☐ Although no single other condition stands out, Raynaud's is the most frequently reported, appearing in 42% of those who named another condition (or 20% of all survey respondents), followed by peripheral neuropathy at 29% (13% of all respondents) and thyroid disease at 25% (11% of all respondents)
 - ☐ Only 46% report their perspiring as normal even when not flaring; another 38% can't sweat at all, and that number swells to 88% during flares
 - ☐ No relationship was found between symptoms and age, gender, whether another condition is present and the ability to control flares

- There does seem to be a genetic component to the disease
 - 5% have a relative diagnosed with EM, and another 20% suspect that a relative has EM symptoms
 - These percentages are enormous considering how rare the disease is in the general population
- No treatment works consistently for all diagnosed with EM; although 79% aggressively seek treatment
 - Anticonvulsants¹ have the greatest effect among prescription medicines, helping 43%, followed by SNRI's² at 23% and NSAID's³ at 21.7%
 - Supplements/regimens and invasive therapies were credited as helpful by fewer respondents, but some provided comparable relief among those who did use them, particularly the Mayo Clinic's compounded topical cream (containing 1% Amitriptyline/5% Ketamine in a Plo Gel), magnesium supplements and acupuncture
- Only 26% of respondents feel confident that their doctor knows enough to diagnose and treat EM for others; 49% believe their doctor is not educated enough to do so

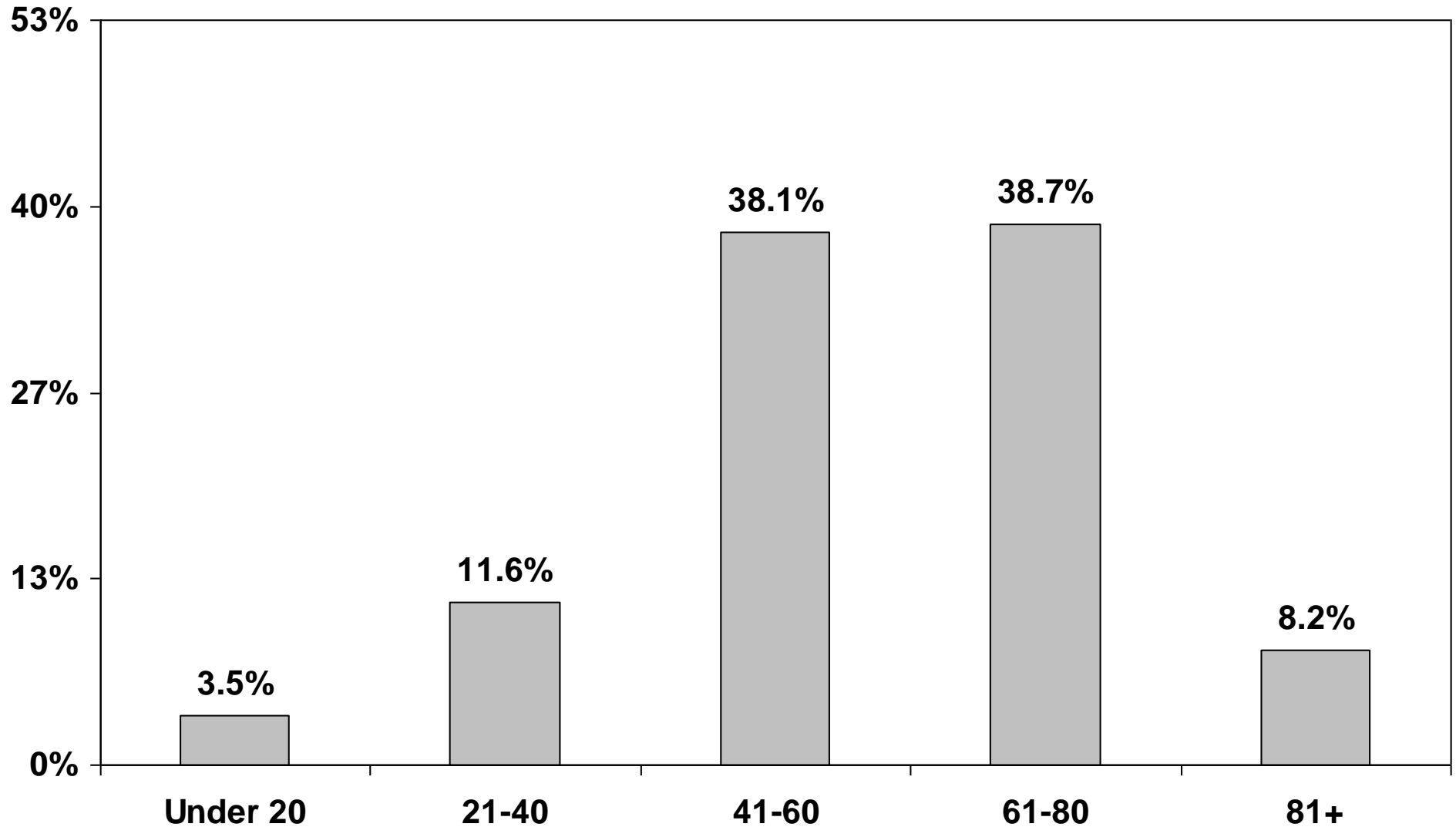
NOTE: For all subsequent slides " N= " refers to the number of responses used

1, 2, 3 - See Appendix 1 for descriptions

Respondent Make-up

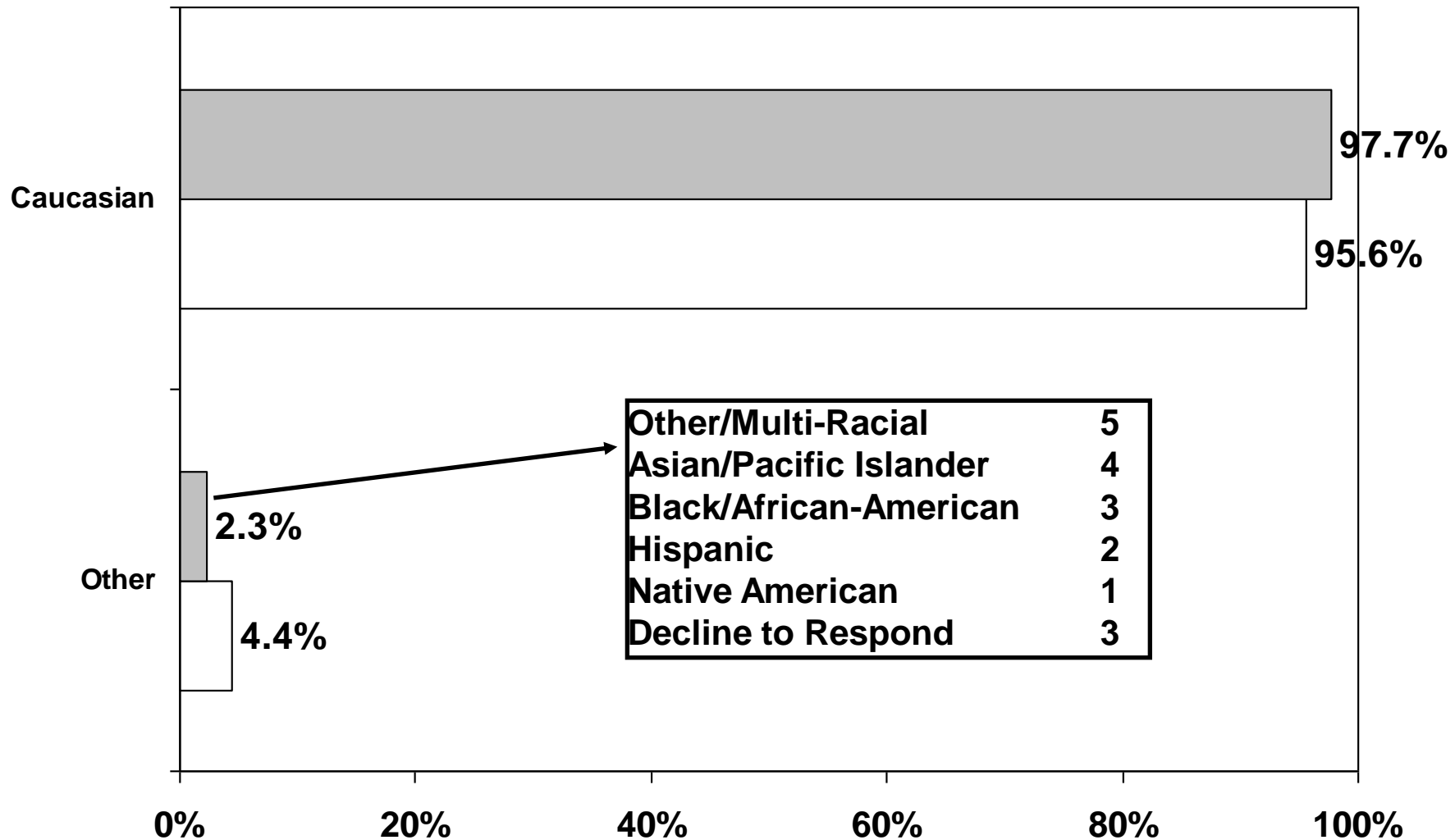
Respondent Age

N=318

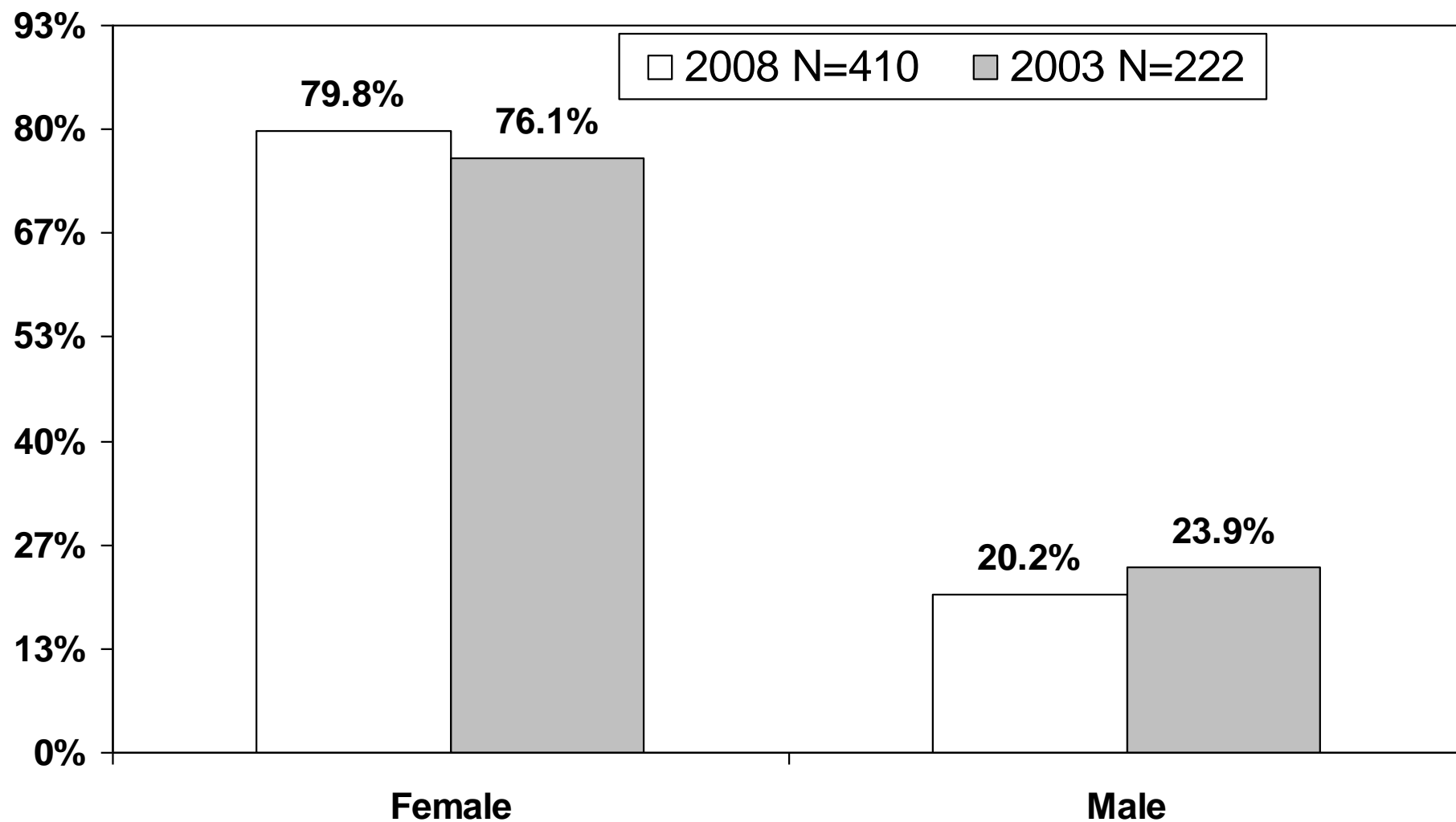


Ethnic Background

□ 2008 N=405 ■ 2003 N=222

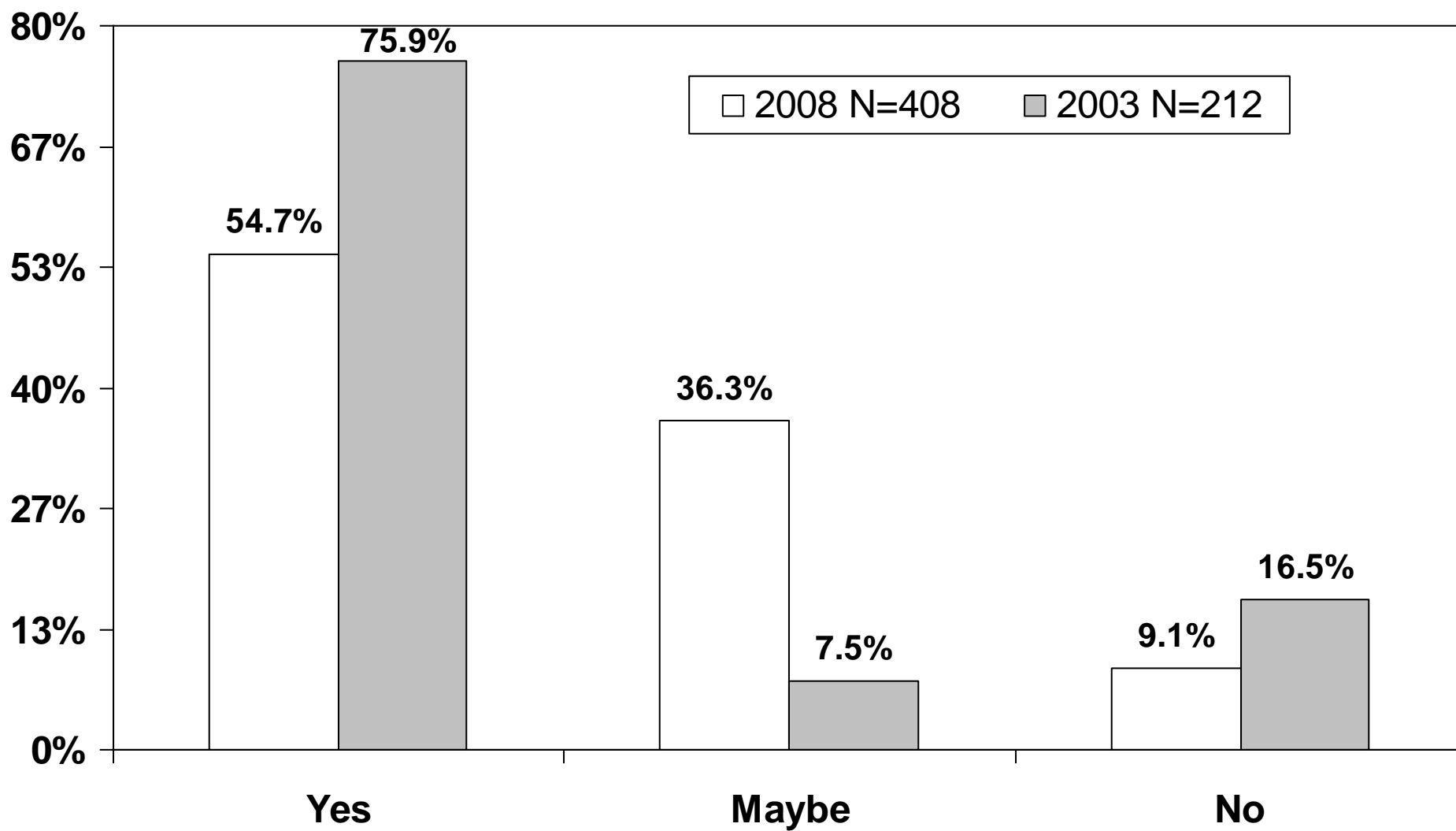


Gender



Medical Research Participant

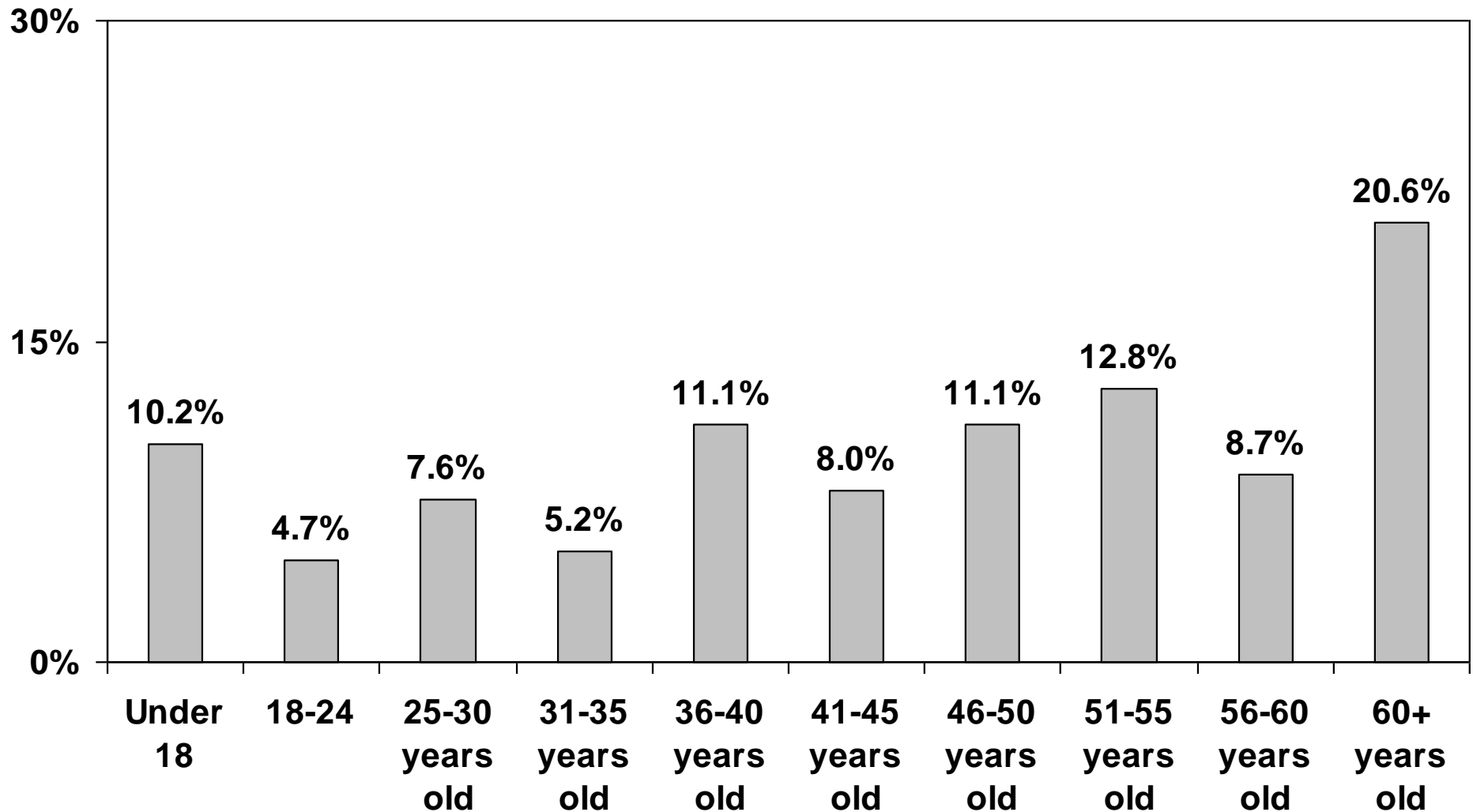
Would you be willing to participate in medical research concerning EM if the opportunity presented itself?



Disease Onset and Diagnosis

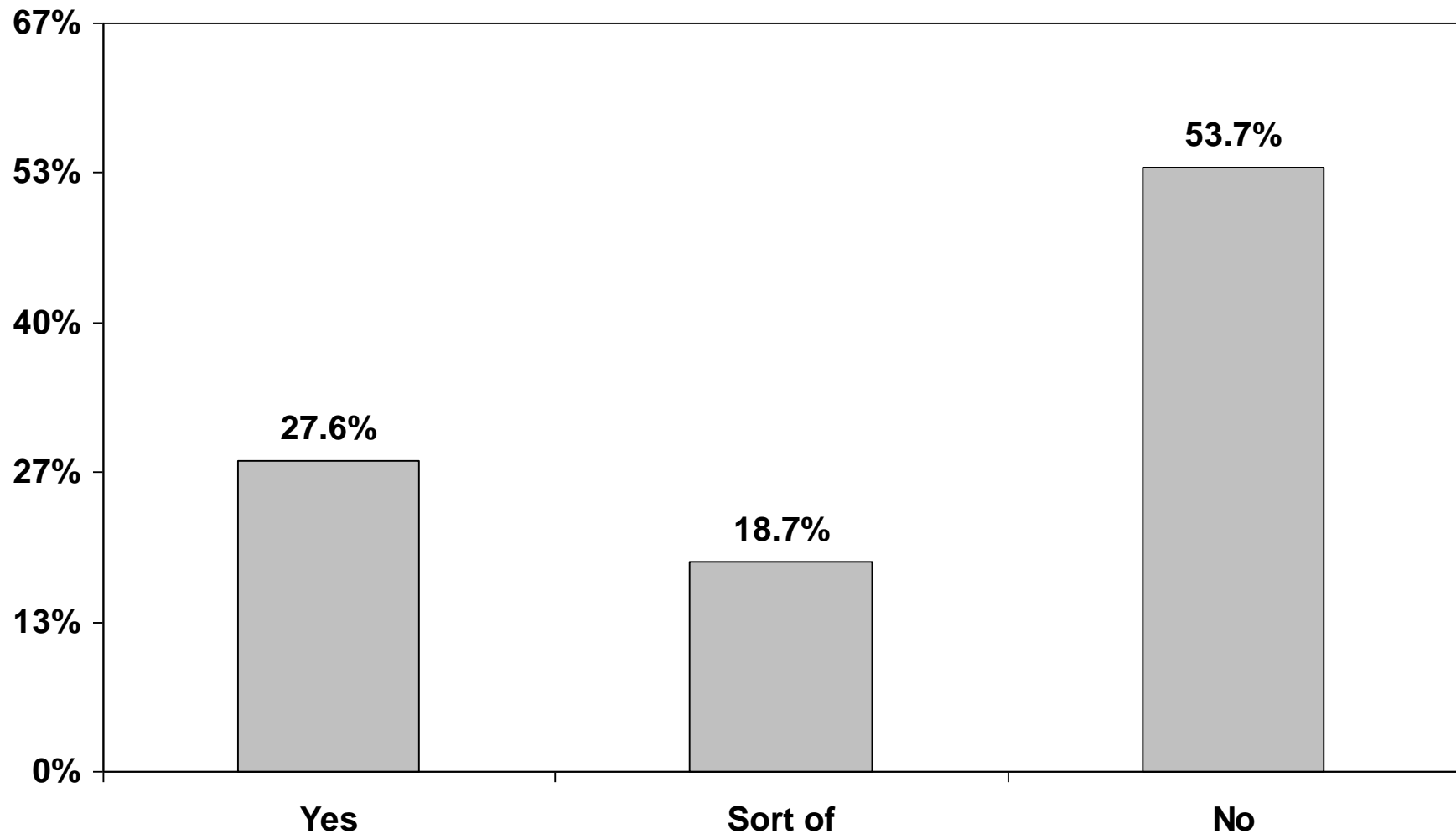
Age When Symptoms Emerged

At what age did you first become aware of your Erythromelalgia (EM) symptoms? N=423



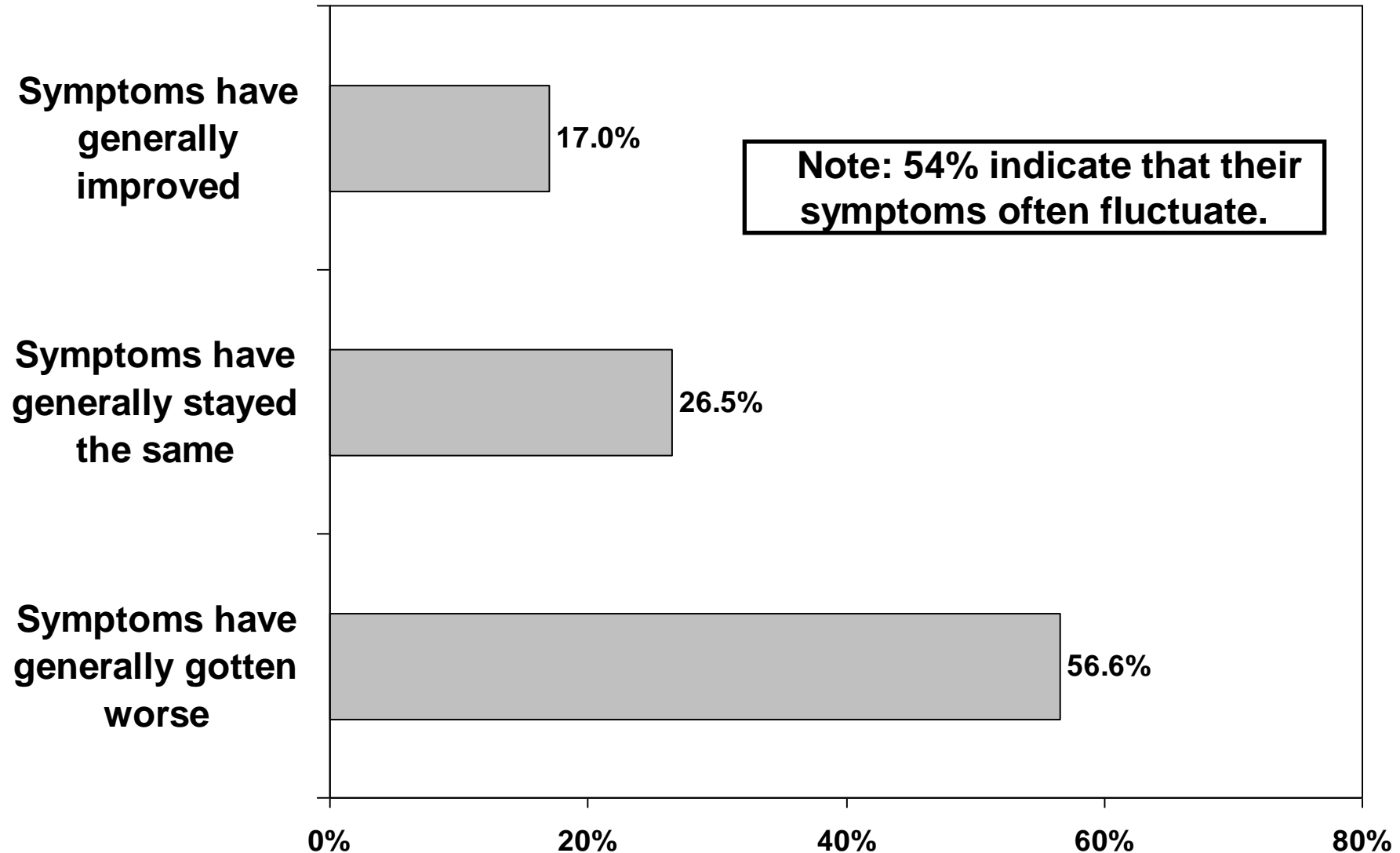
Defining Event Starting Symptoms

Do you feel your symptoms started after some sort of defining event like trauma or change in your life? N=423



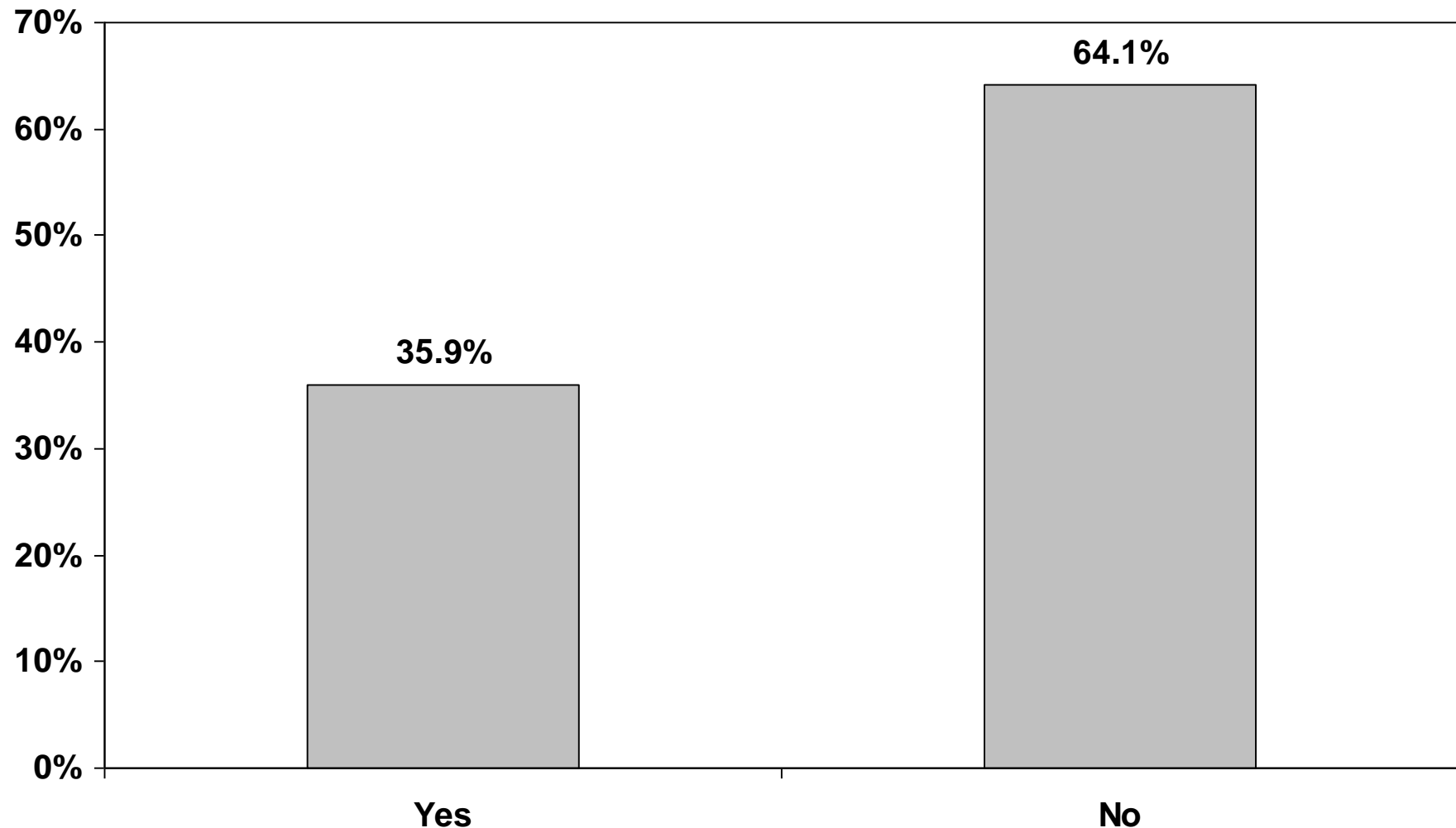
Progression of Symptoms

How have your symptoms changed since you first became aware of them? N=412



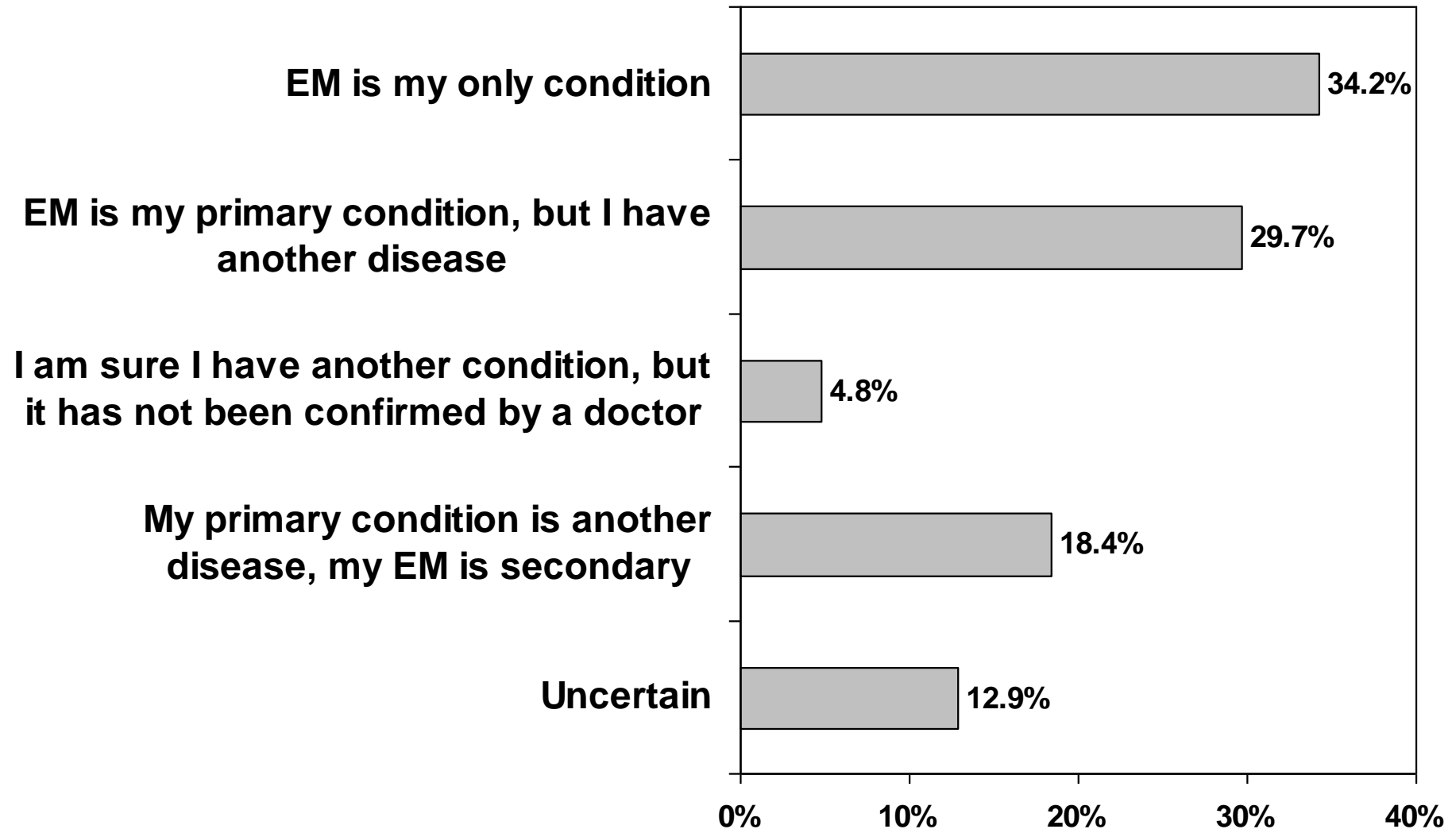
Symptom-free Days

Since your EM symptoms started have you had periods of more than a few days where you were symptom-free? N=401



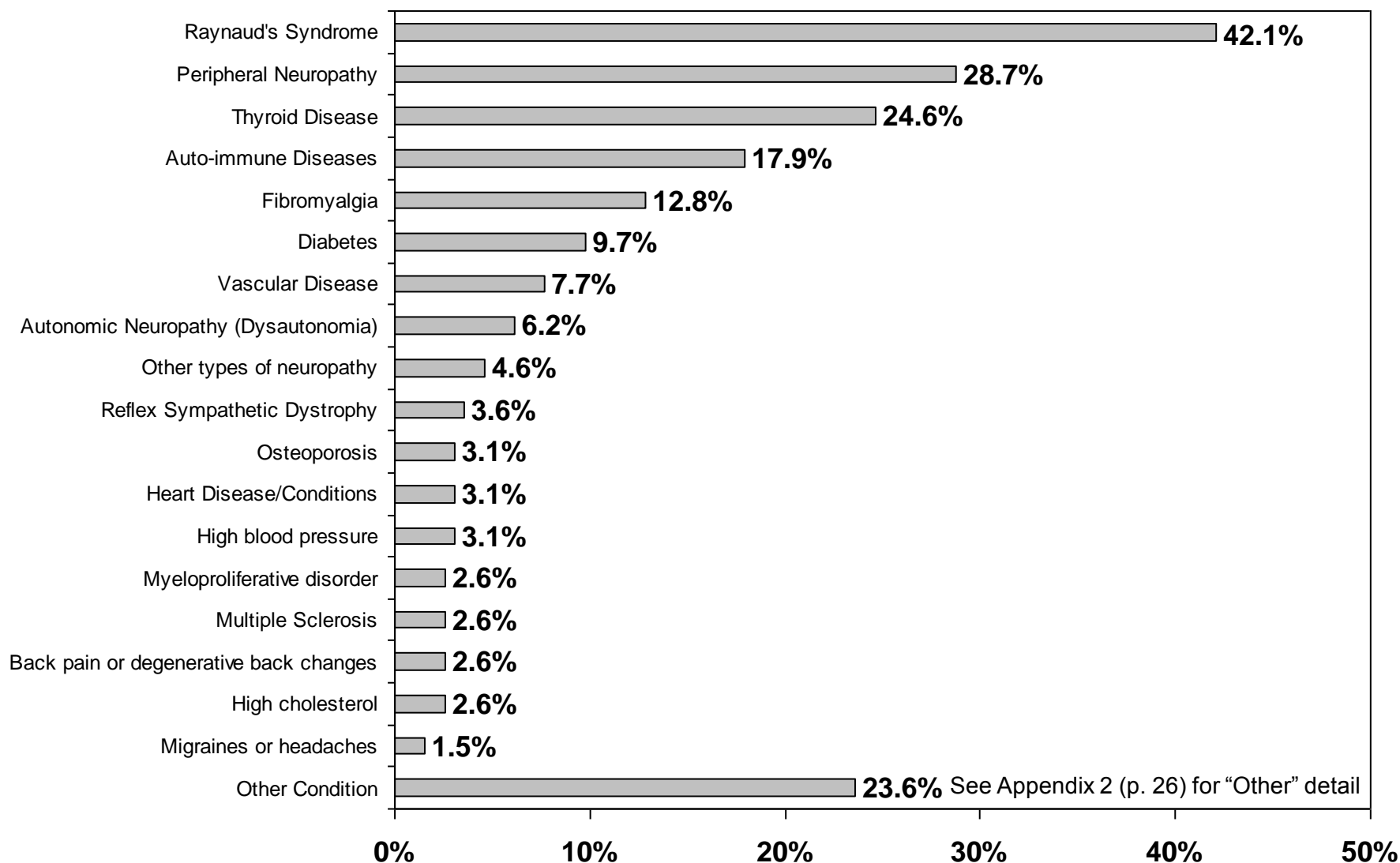
Accompanying Conditions

Other diseases can accompany EM, either in a primary role or a secondary role. Which of the following applies to you? N=418



Diseases or Conditions With EM

What other diseases or conditions do you have that accompany your EM?
(Select all that apply) N=195



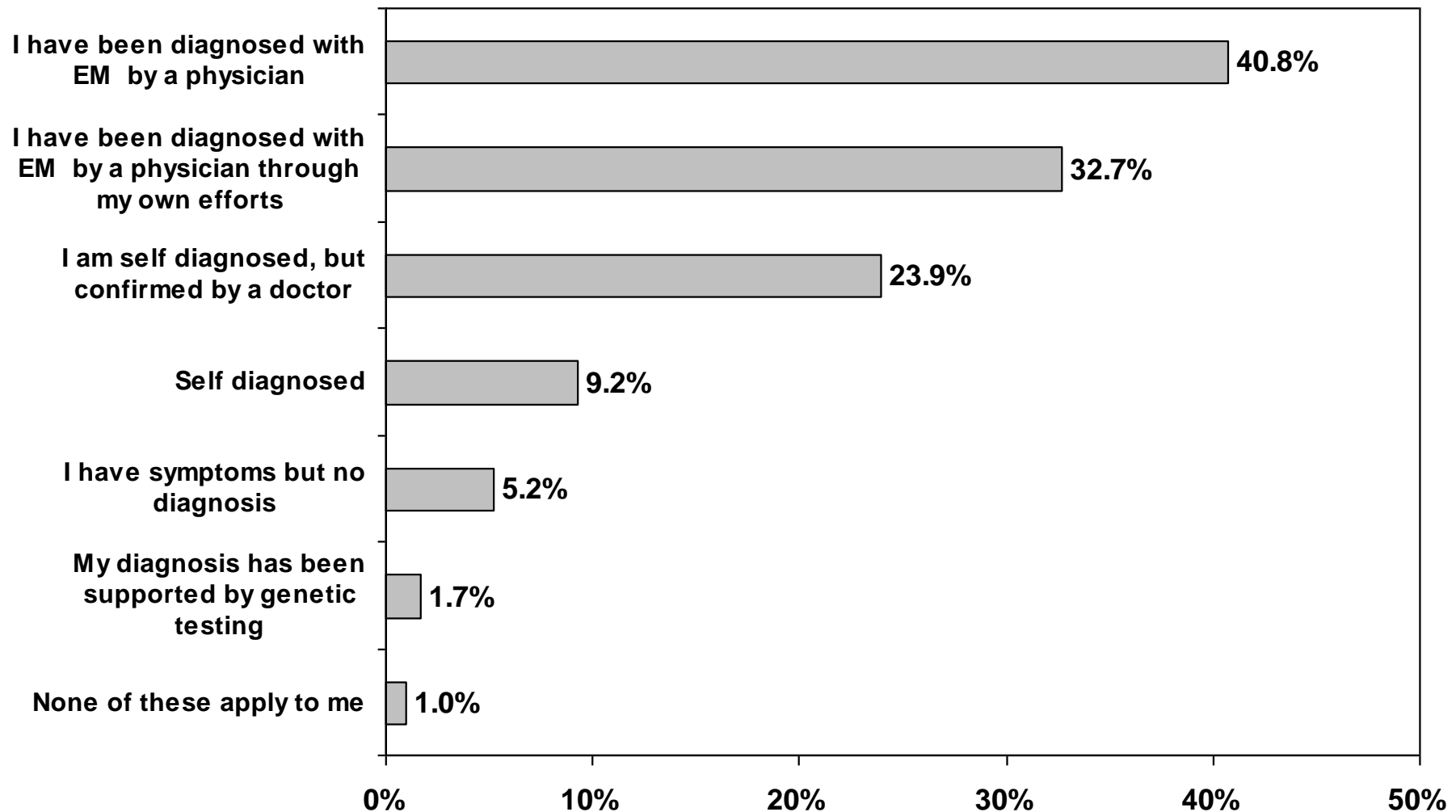
Diseases or Conditions with EM: “Other” Responses

What other diseases or conditions do you have that accompany your EM?

- Antiphospholipid Syndrome (2)
- Anxiety, ADHD
- Asthma
- Benign brain tumor
- Bowen's Disease
- Breast Cancer
- Bronchiectasis Disease
- Burning mouth
- Chronic Fatigue Syndrome
- Chronic renal failure / on dialysis
- Colitis
- Depression
- Eczema
- Ehlers Danlos – (hyper mobility)
- Fibrosing Alveolitis
- Food allergies
- Hepatitis C
- Interstitial Cystitis (2)
- Irritable Bowel Syndrome (2)
- Lichen planus
- Macrocytic anemia
- Menieres Disease
- Metatarsalgia
- Monoclonal gammopathy
- MTFHR - blood clot disorder
- Multiple vasculitis
- Muscle spasms of feet & calves
- Non-malignant tumor on my pituitary gland
- Obstructive Sleep Apnea
- Panic Disorder
- Parkinson's (MSA) - Multisystem Atrophy
- Periodic paralysis
- Perniosis - also known as chilblains
- Polymyalgia rheumatica
- Postural Orthostatic Tachycardia Syndrome (POTS)
- Premature ovarian failure (early menopause)
- Problems with blood clots
- Prostate surgery
- Pseudohypoparathyroidism
- Psychiatric disorder
- Reactive hypoglycemia
- Restless Leg Syndrome
- Rosacea
- Scoliosis
- Sleep disorder
- Temporal arthritis
- Thalassemia
- von Willebrand Disease
- Waldenstrom's Macroglobulinemia

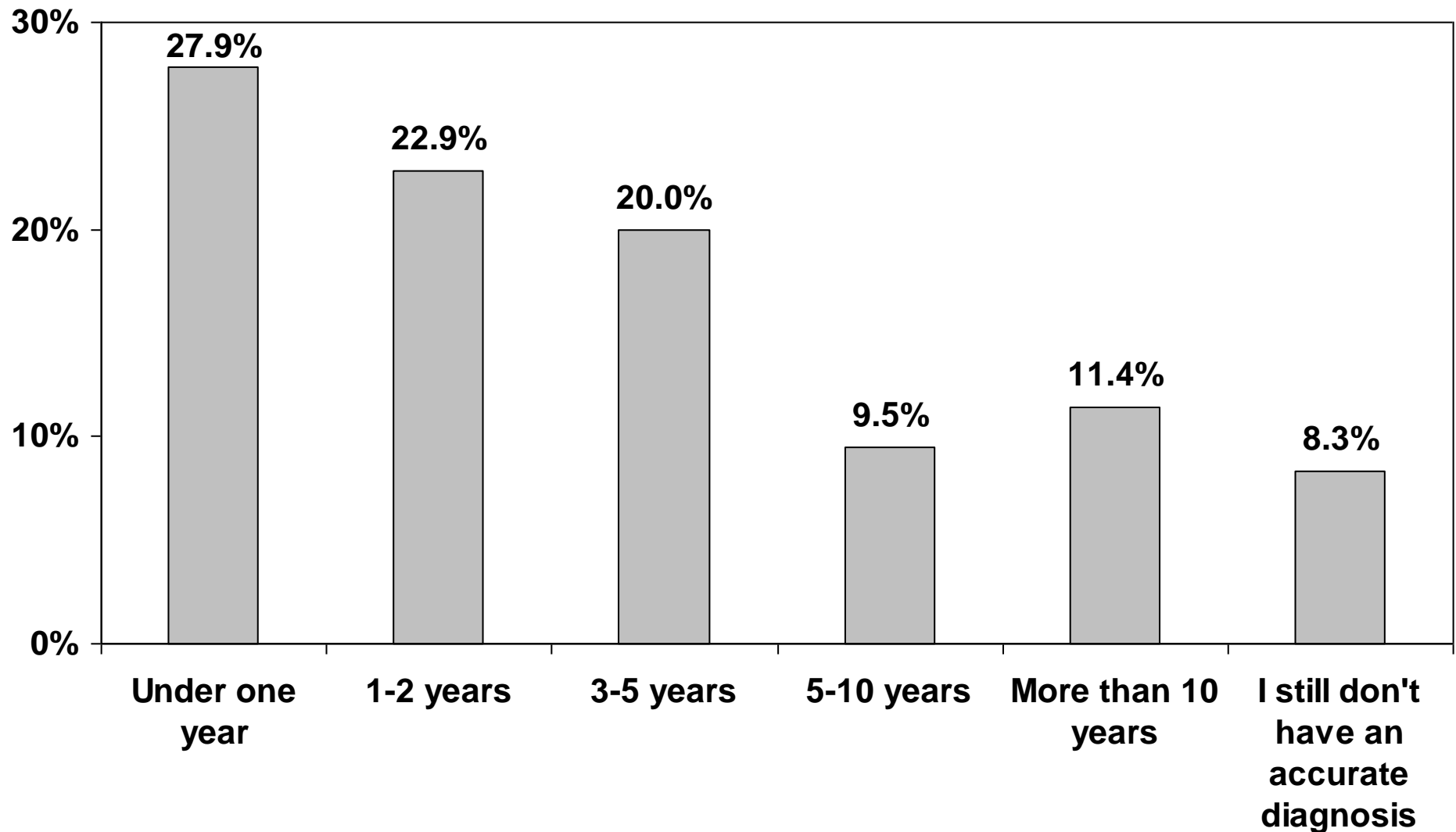
EM Diagnosis

Please indicate which of the following statements applies to your EM diagnosis.
(Select all that apply) N=422



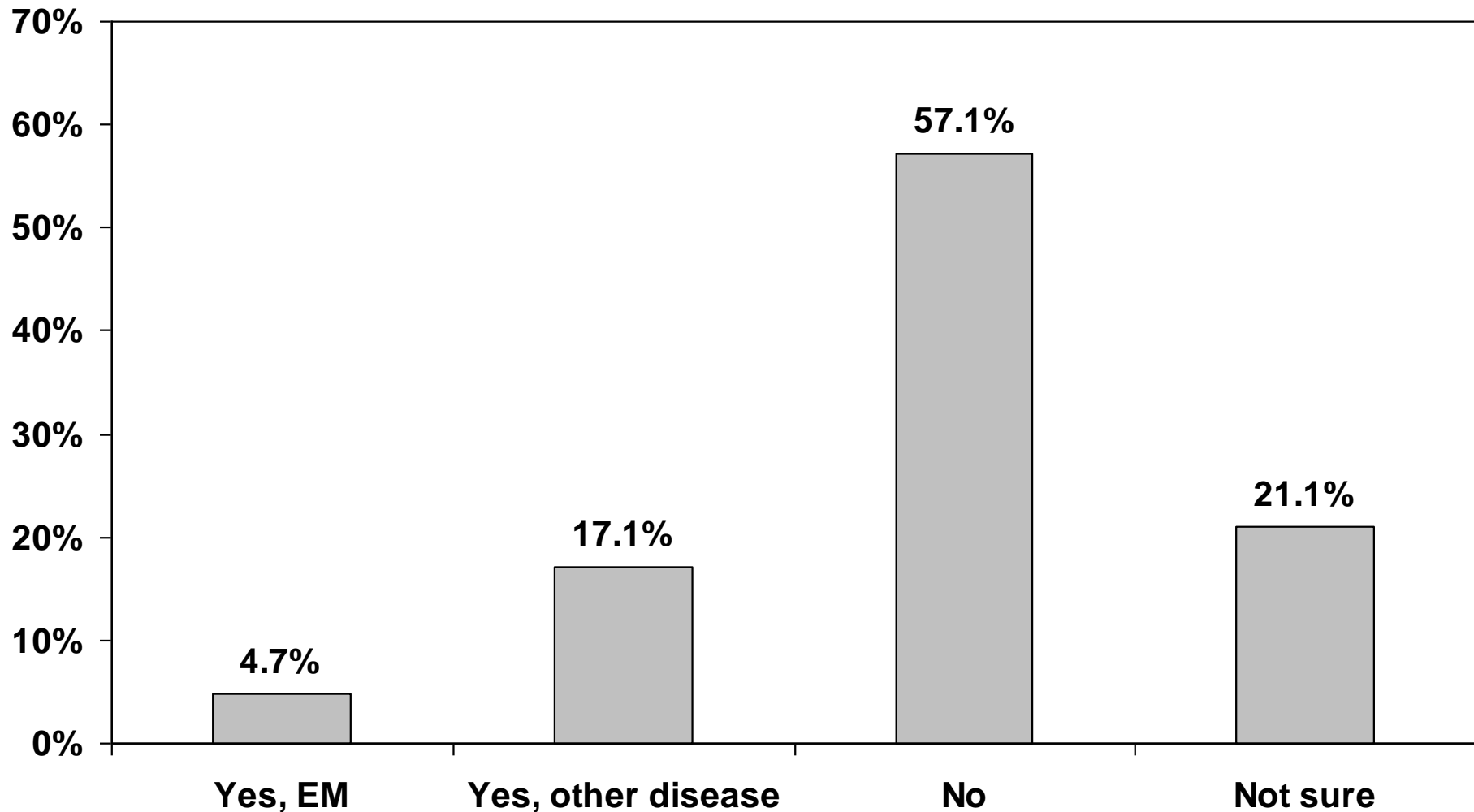
Time From First Symptoms to Diagnosis

Approximately how long did it take you or the medical community from the time you first had EM symptoms until you had an accurate EM diagnosis? N=420



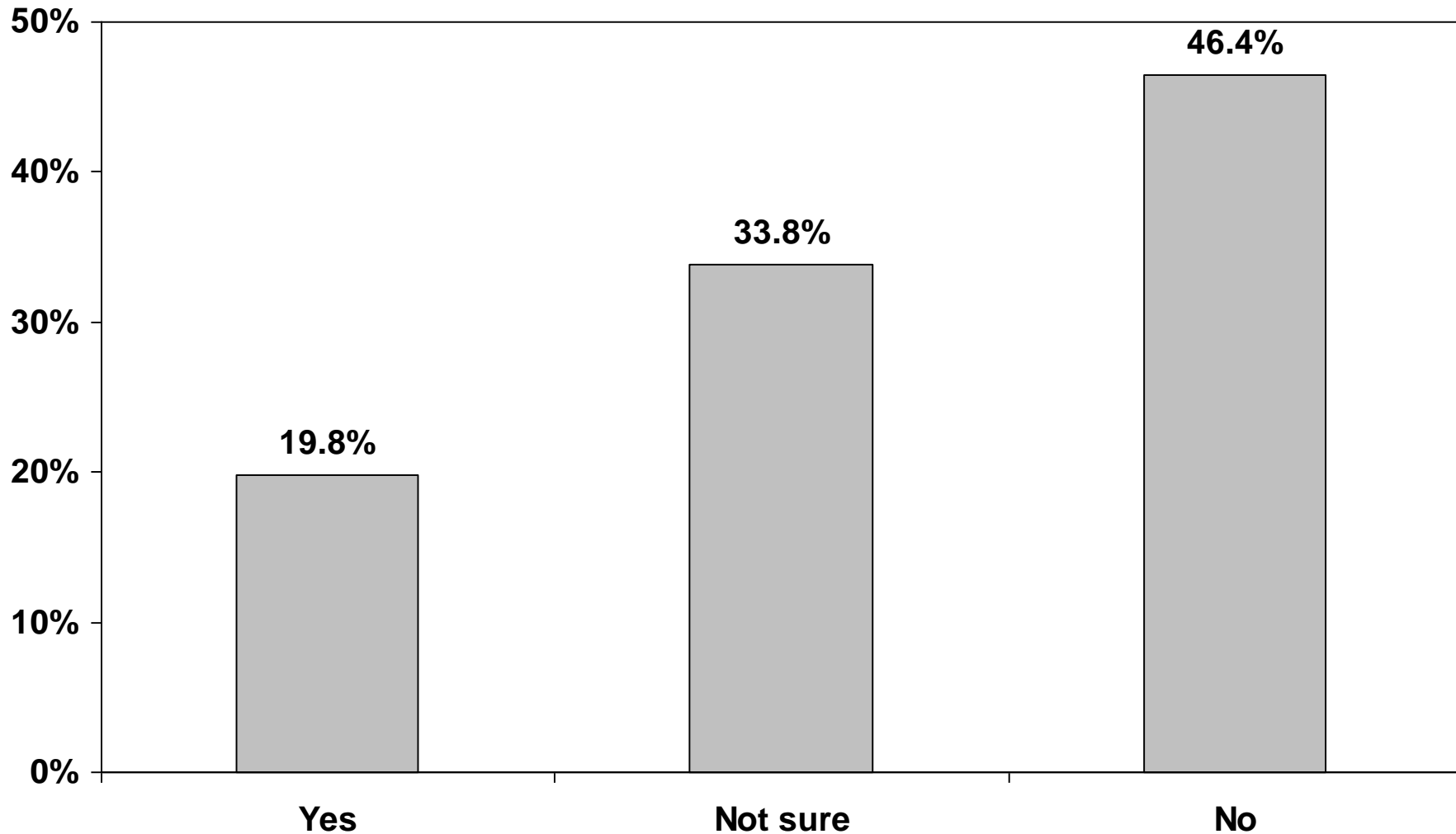
Blood Relatives with EM - Diagnosed

Do you have any blood relatives that have been diagnosed with EM (or any of the other diseases mentioned on page 8)? N=422



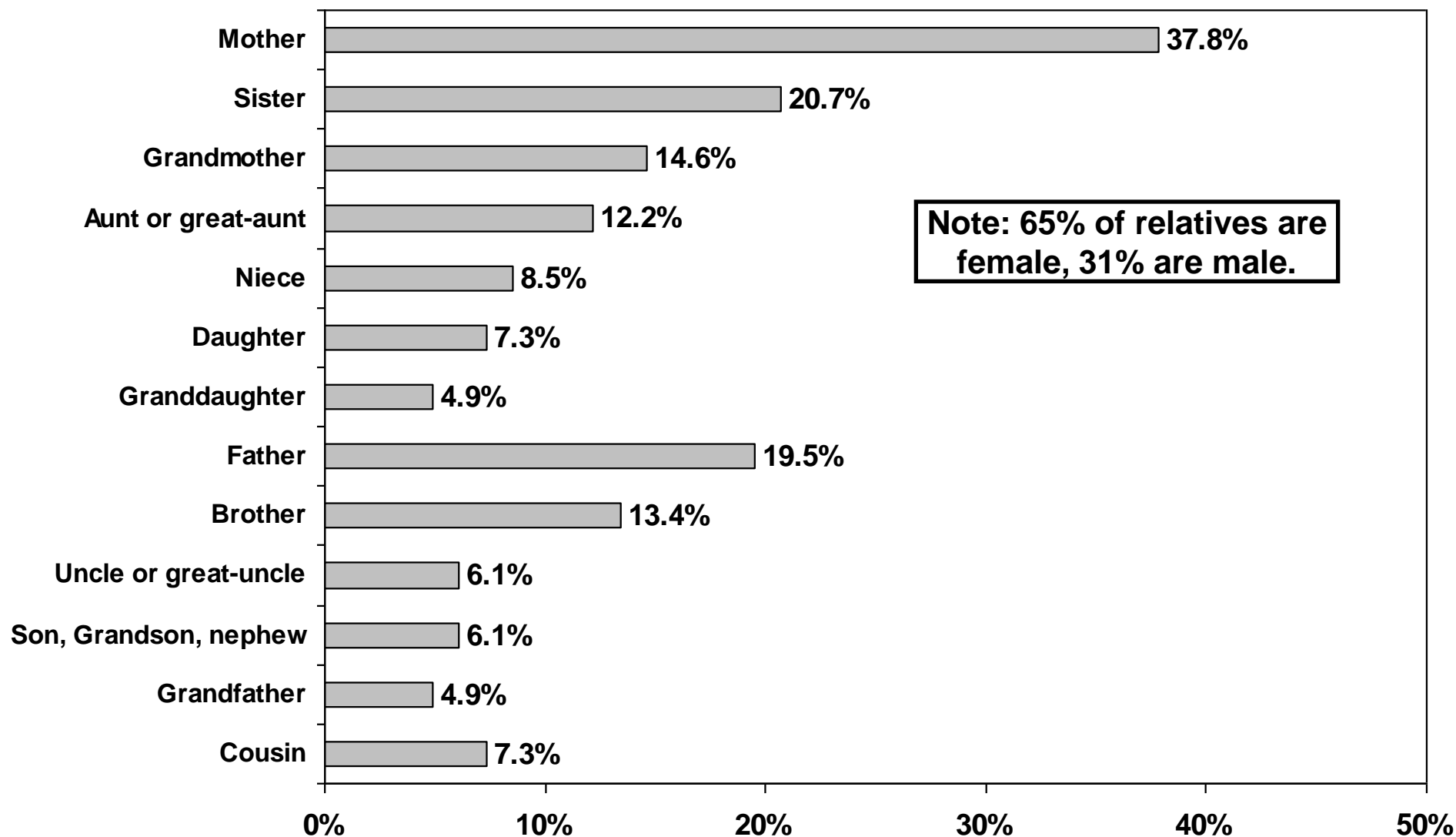
Blood Relatives with EM - Undiagnosed

Do you think any of your blood relatives have EM symptoms but have not actually been diagnosed with EM? N=422



Blood Relatives with EM

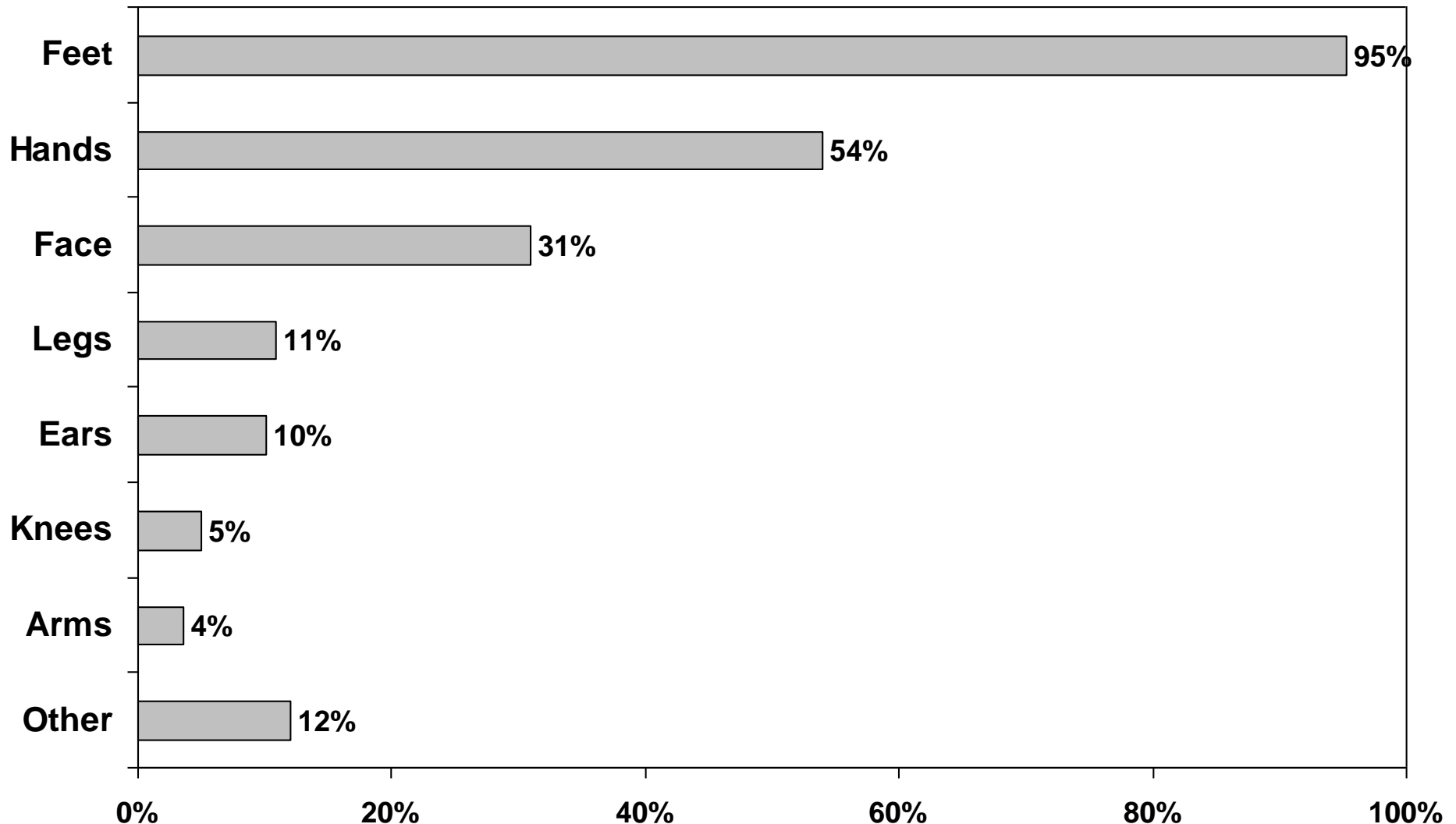
List your relations that have been diagnosed with EM or who you think might have EM symptoms. **N=82**



Disease Symptoms

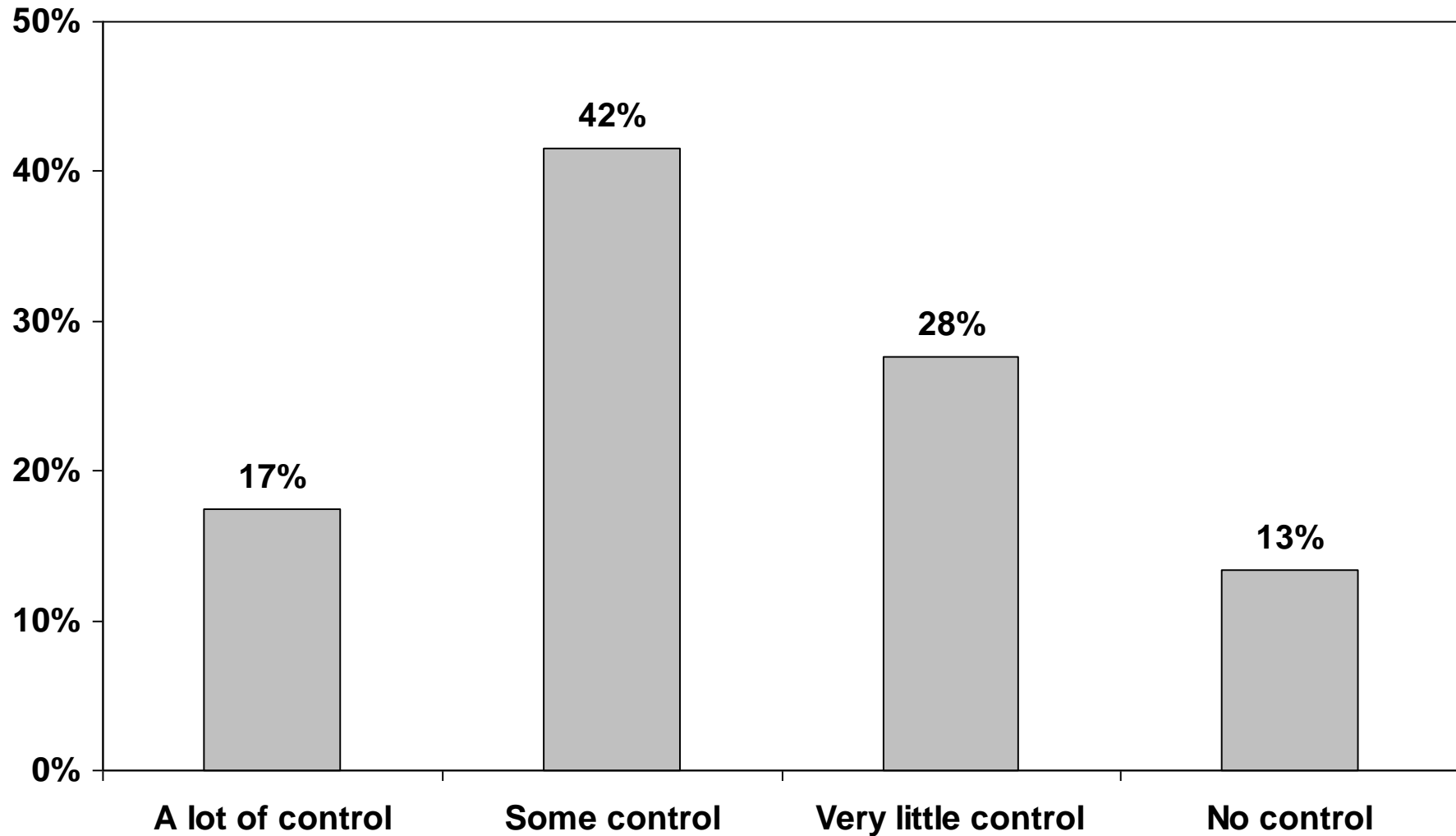
Location of Flares

Please indicate in which of the following locations you experience painful flares which you attribute to your EM. (Select all that apply) N=423



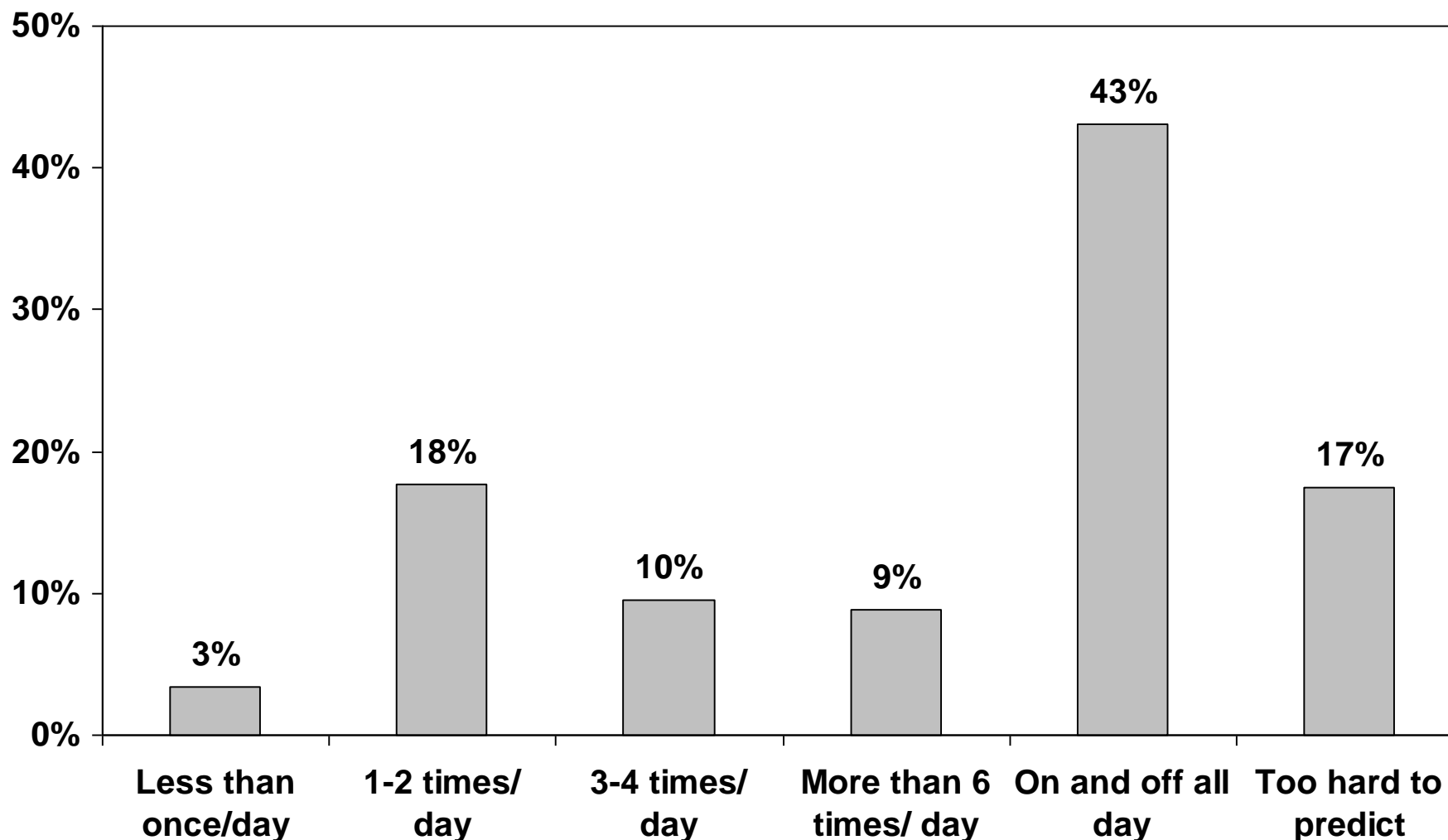
Control Over the Frequency of Flares

Based on actions you take to control your environment, how much control do you have over the frequency of you EM flares? N=412



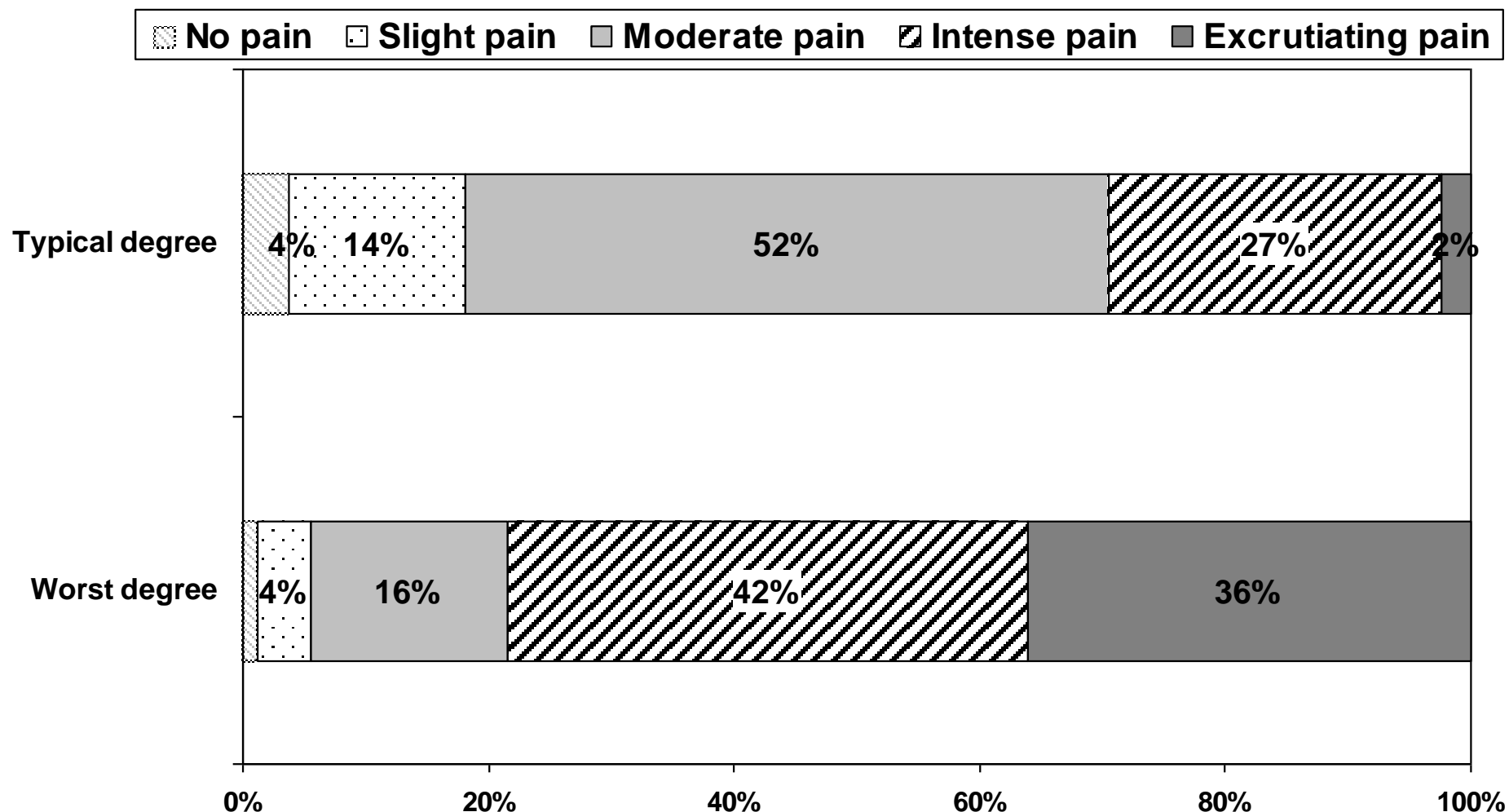
Frequency of Flares w/No Intervention

If you did NOTHING to control your EM flares, how often would you experience a flare-up?
N=418



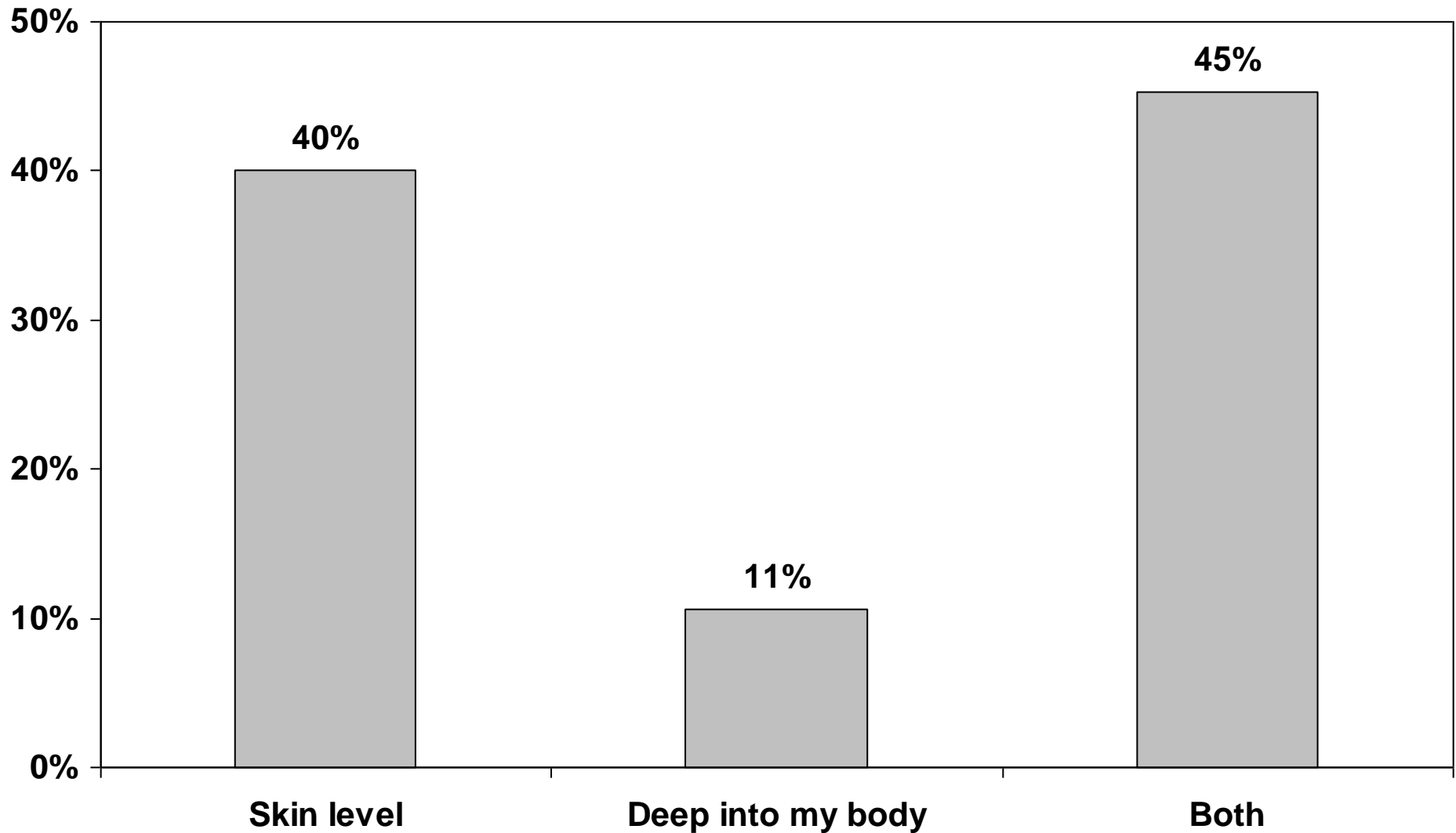
Degree of Pain During Flare Up

Please indicate the worst degree of pain you might feel during a flare-up and the typical degree of pain you feel during a flare up. N=423



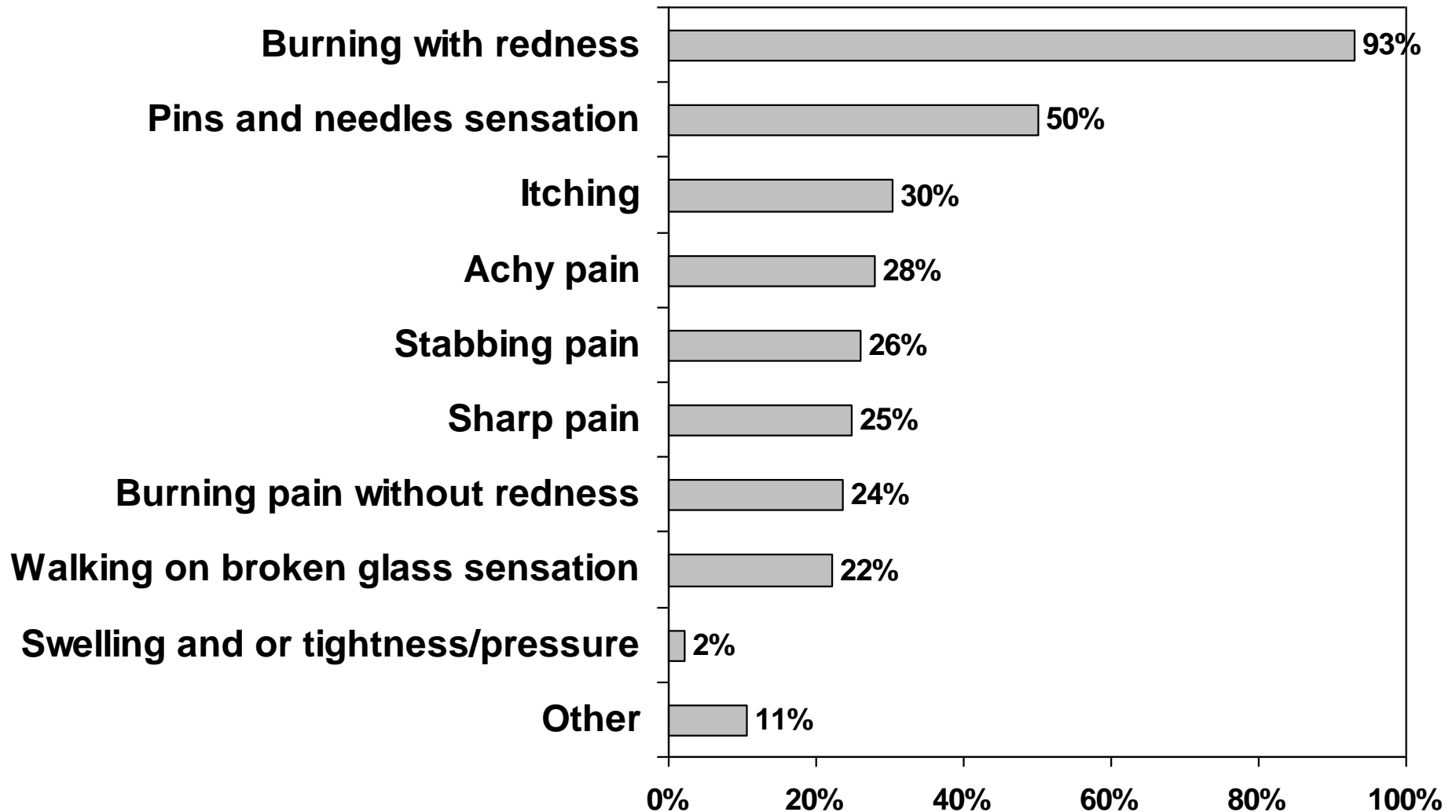
Level of EM Pain

At what level would you say you experience your EM pain? N=400



Type of Pain with Flares

Please indicate the type of pain you experience with your EM flares.
(Select all that apply) N=420



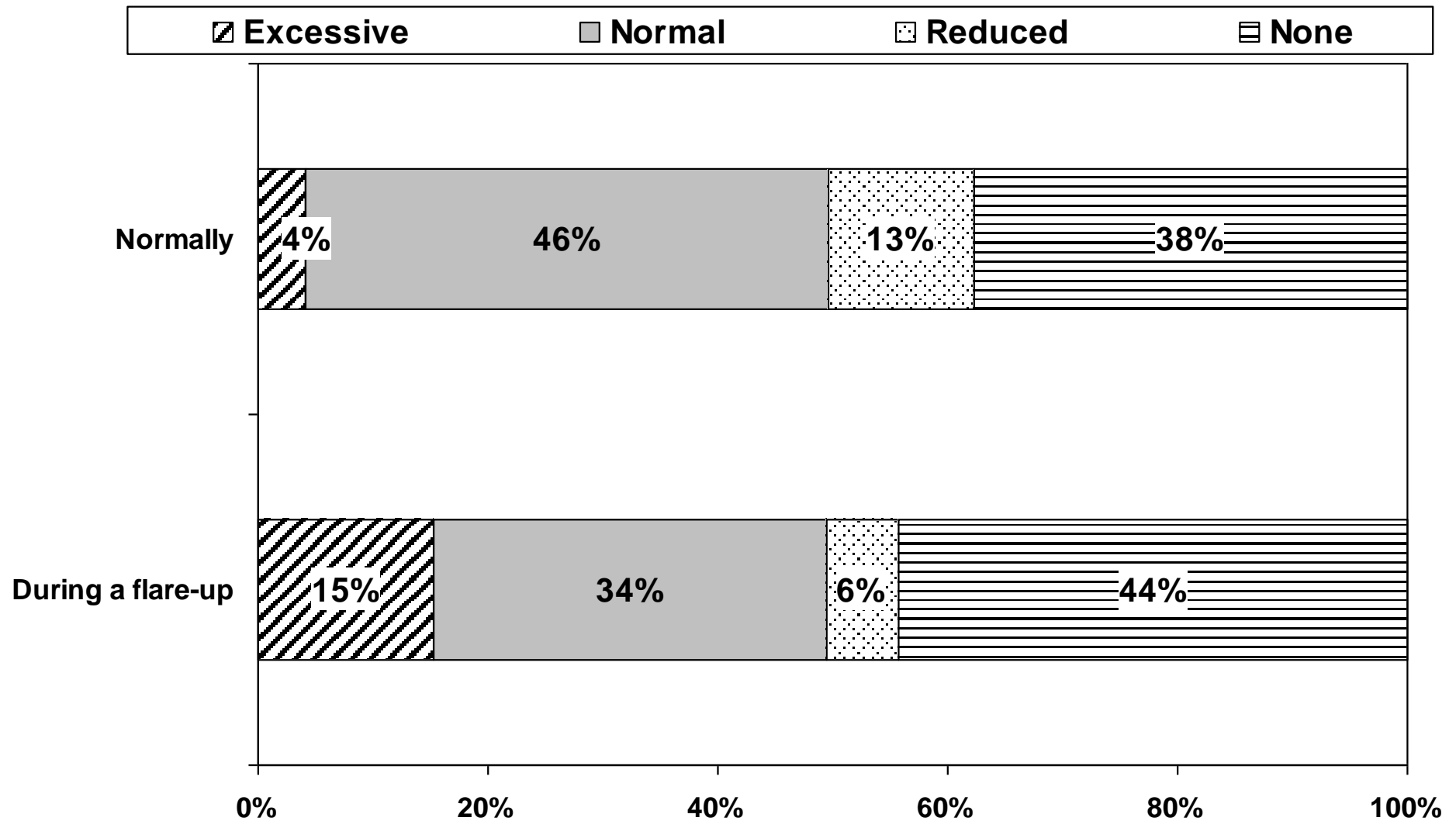
Type of Pain: 'Other' Responses

Please indicate the type of pain you experience with your EM flares.

- Throbbing (5)
- Electrical Sensation in Feet (4)
- Crushing Pain (3)
- Feels like a layer of skin has been ripped off (3)
- Feels like a layer of skin has been ripped off & then salt rubbed into the open wounds
- Carpet feels like sharp stones
- After blow torch is touched to gasoline drenched feet
- Bee-stings
- Boiling acid poured over feet
- Bottom of feet incredibly sore
- Buzzing pain in toes and bottom of feet
- Cramping
- Crawling sensations
- Deep bone pain
- Extreme skin sensitivity
- Facial bone, teeth, and jaw pain
- Feeling of being on fire
- Feeling of large painful blisters on bottom of feet
- Feeling of raw flesh, burnt, stinging nettle rash, itching, burning
- Feels like I'm walking on metal nuts/bolts, skin feels like it's 3/4 sizes too small
- Feet and ankles feel like they will burst
- Heaviness and aching in arms and/or legs
- It's like a bad toothache in my limbs sometimes
- Like an asphalt burn on the bottoms of my feet
- Sinus pain from EM on face
- Pain in ball of my right foot where it all started.
- Pulsing
- Writhing and crying
- Skin feels ready to split
- Twitches
- Uncomfortable hot skin
- Walking ankle-deep in burning hot sand
- Walking on blisters
- Walking on fire with burned off skin
- Walking on rocks sensation

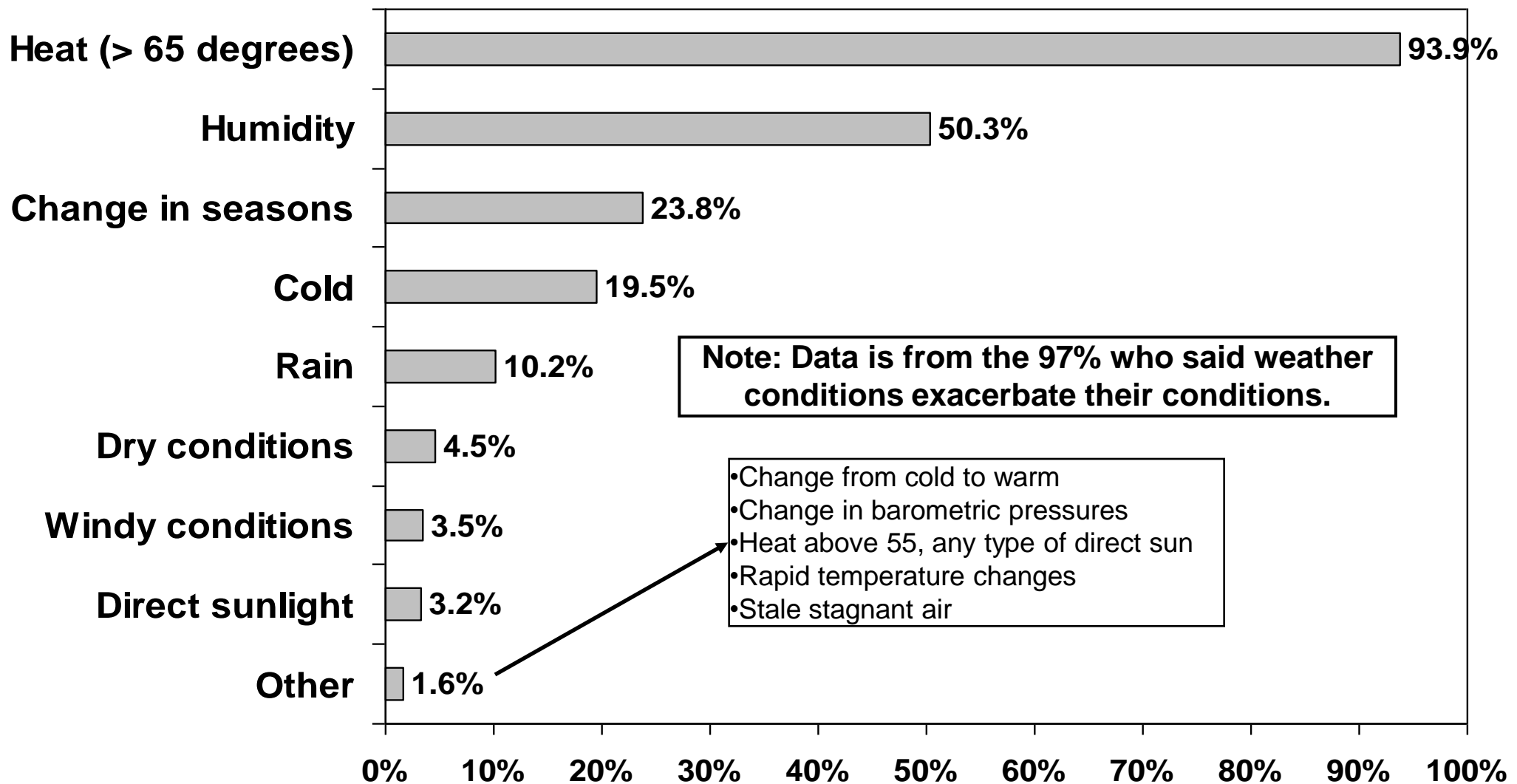
Perspiration

Which of these choices most closely describes your perspiration (sweating)? N=419



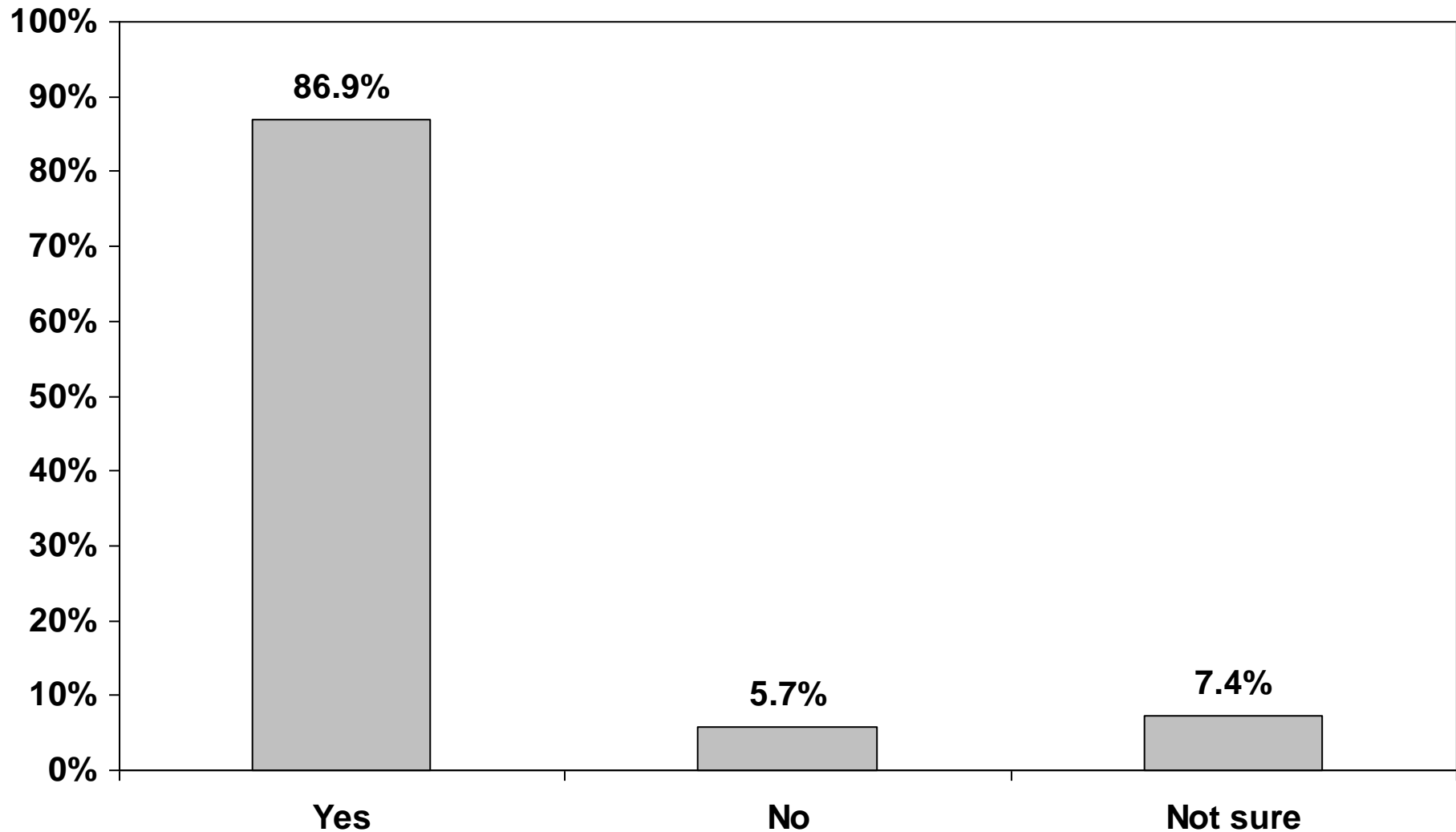
Weather Conditions and EM

Which of the following weather conditions, if any, cause or exacerbate your EM flares?
(Select all that apply) N=376



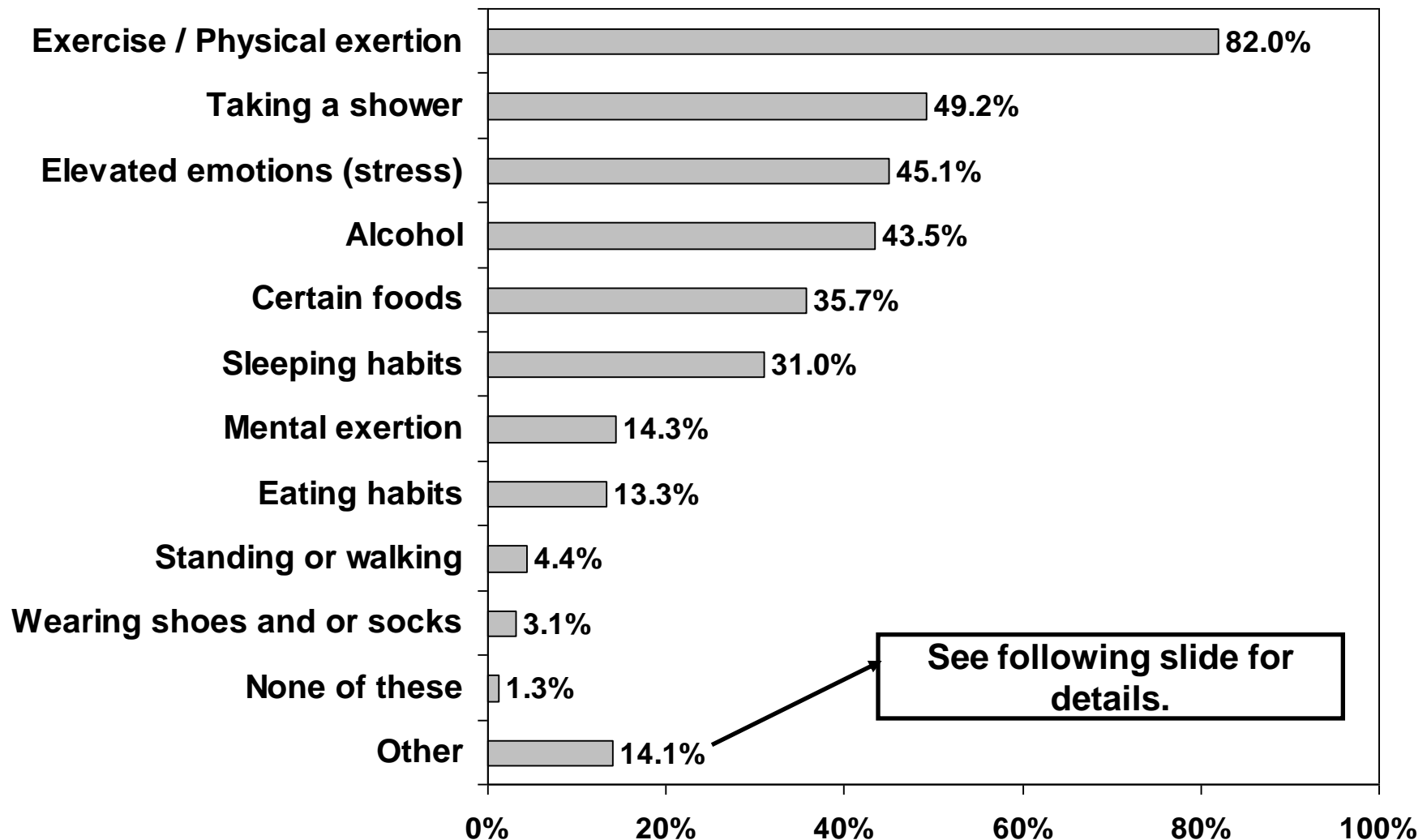
EM Triggers

Have you established that certain things you do can trigger your EM symptoms or make them worse? N=420



EM Triggers (cont'd)

Which of the following do you feel might trigger your EM symptoms to occur?
(Select all that apply) N=391



Triggers: “Other” Responses

Which of the following do you feel might trigger your EM symptoms to occur?

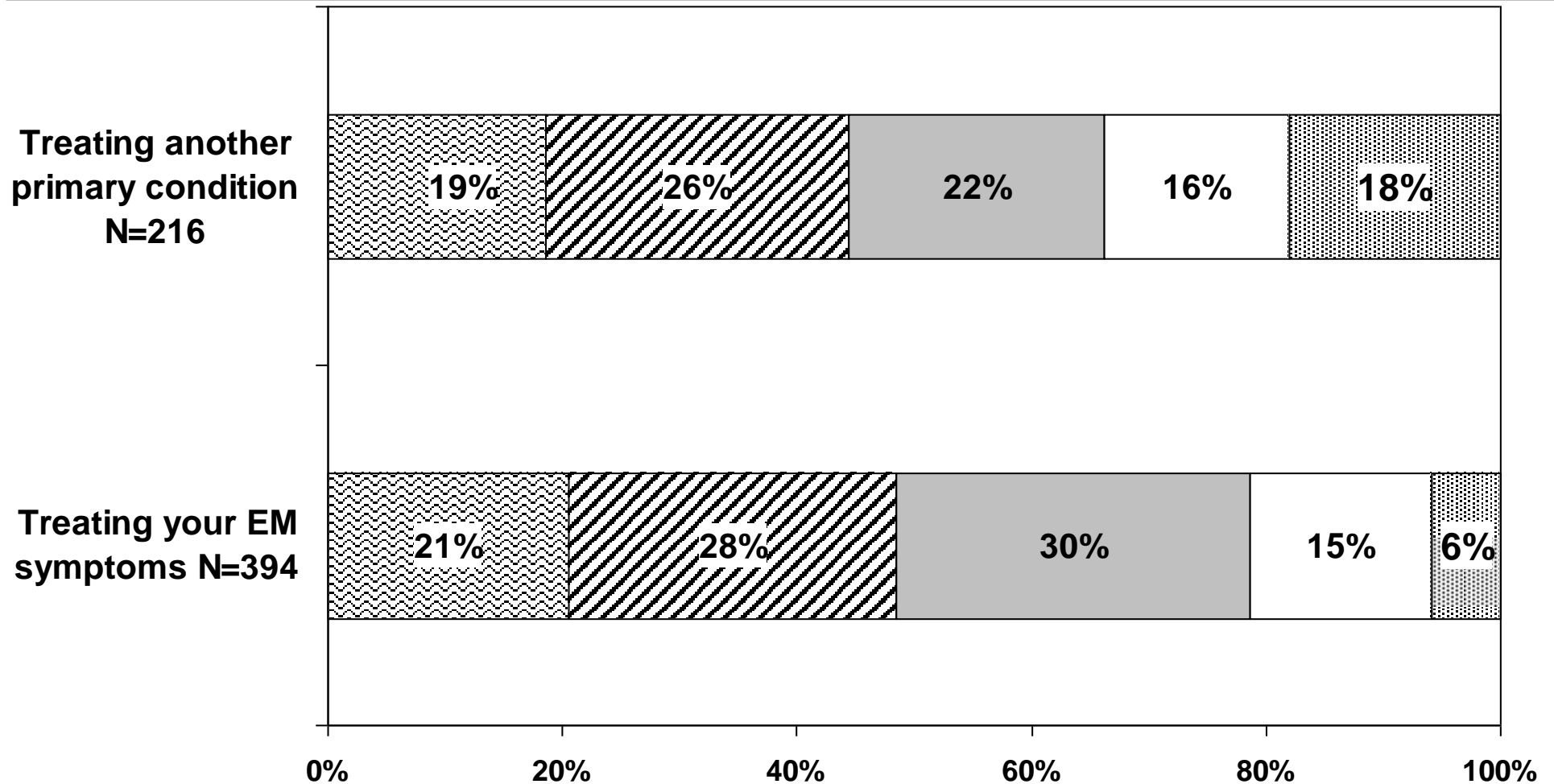
- Legs down/not elevated (7)
- Exposure to direct sunlight (6)
- Hot tub/spa/hot bath (6)
- Being in a warm place (5)
- Type of footwear (4)
- Elevation of limbs (2)
- Allowing my hands or feet to get really cold - EM flares as they warm
- Also toes get warm/hot in the evening time
- Ambient temperature
- Being warmly dressed or under bed covers in cold weather
- Bending over, standing for more than a few minutes
- Cold/ice compresses
- Contact with running water (hand washing); when sitting on softer surfaces
- Covering feet (including use of rich creams)
- Drinking hot beverages, sitting for long periods of time
- Elevated body heat
- Feet on warm surface or covered
- Feet up with toes point up
- Holding a warm object like a laptop or plate, when stressed like when giving a presentation
- Hot flashes
- Infection
- My 12 days of progesterone
- Positional (e.g.: legs dependent, or lying on my side will always trigger EM flare)
- Raynaud's in my toes causes blanching, then I get EM if I warm up too quickly
- Resting; any change in heart rate
- Room temperature above 69 degrees, sitting with feet down, lying in bed
- Smoking
- Too many blankets in bed
- Vasodilator drug or body cream

Treatment

EM Treatment

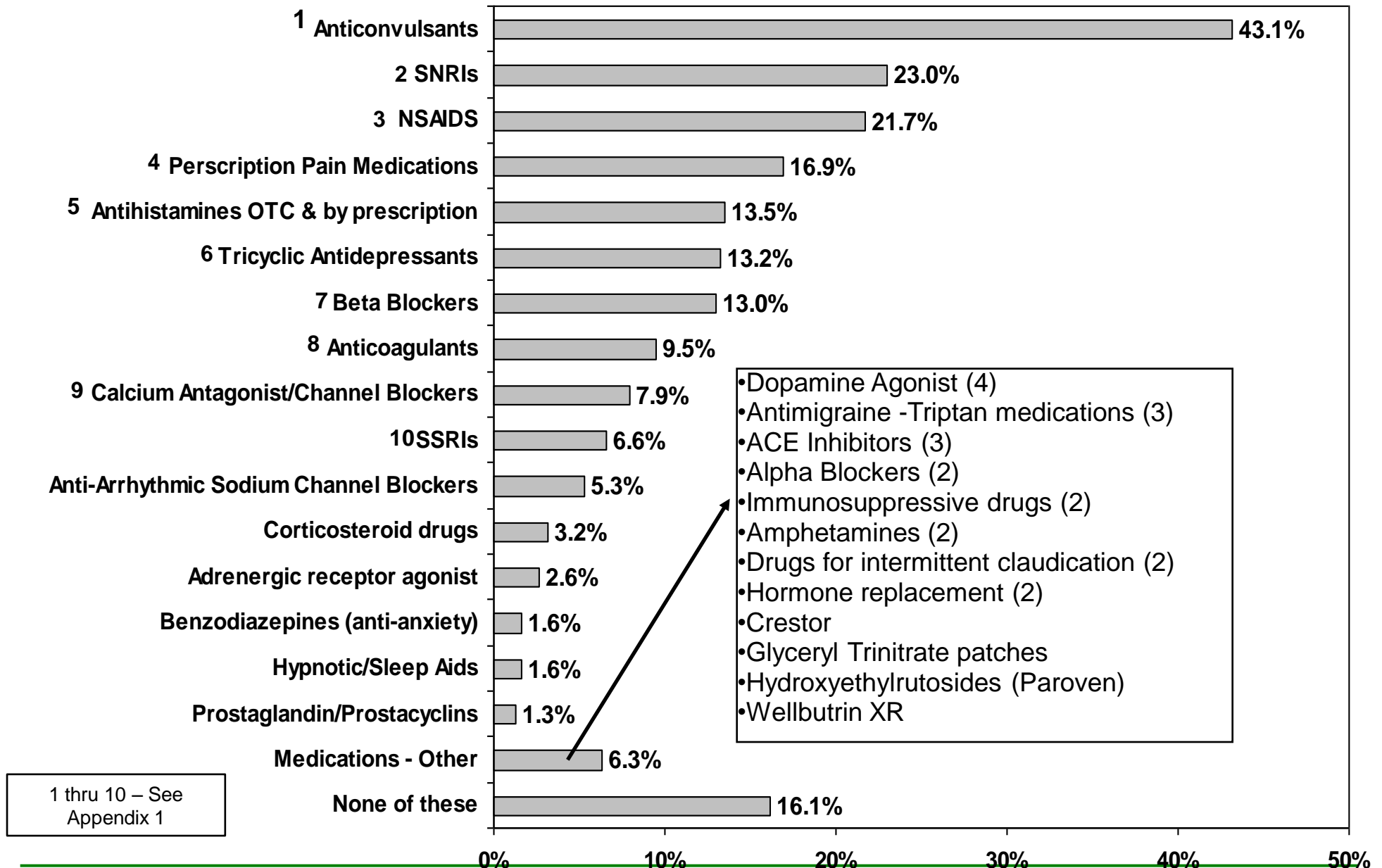
Please rate how aggressively you treat your EM condition or another primary condition that affects your EM.

5= Very aggressively 4=Aggressively 3=Somewhat aggressively
2=Passively 1=Not at all



Most Successful Medications

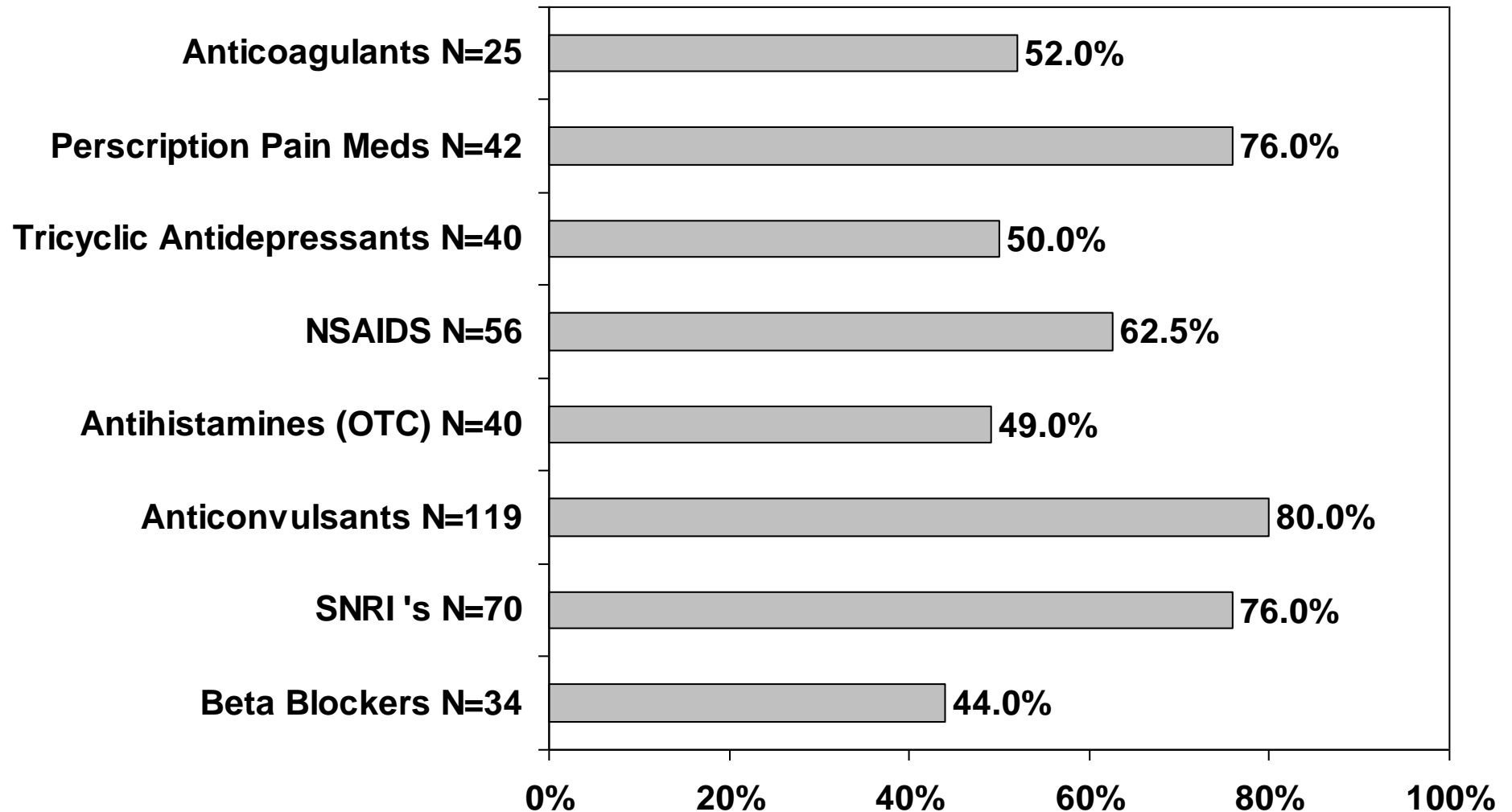
With which of the following **medications and/or treatments** have you had the MOST SUCCESS in treating your EM or primary condition? (Select NO MORE THAN 5) N=378 ~ ~ ~ Percent selecting each item.



Medication Relief

Please RATE each medication or treatment's effectiveness for treating your EM symptoms.

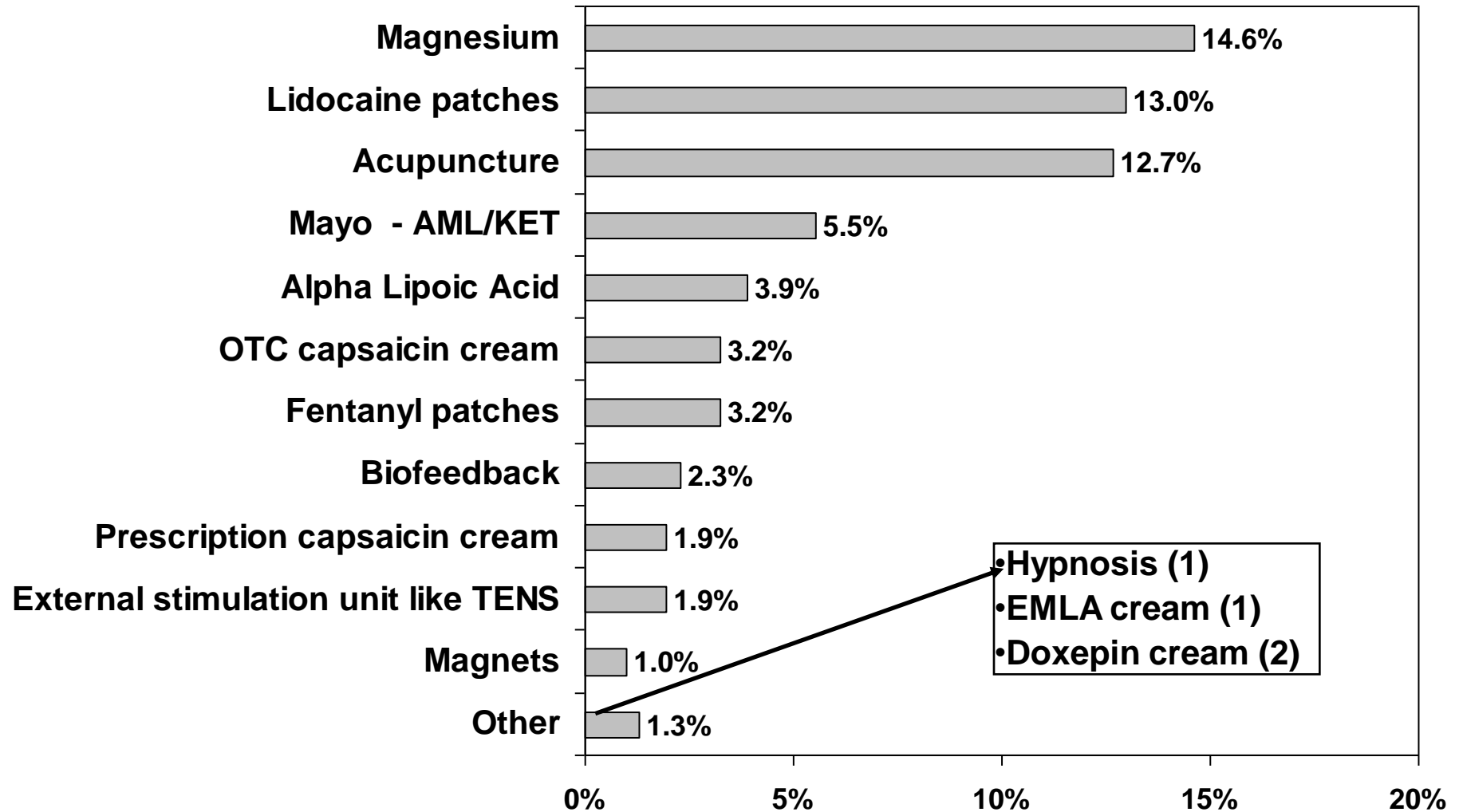
Percent expressing "Good" or "Moderate" relief shown in this chart



Most Successful Supplement/Regimen

With which of the following topical creams, supplements and/or regimens have you had the MOST success in treating your EM symptoms or your primary condition? (Select NO MORE THAN 4) N=308

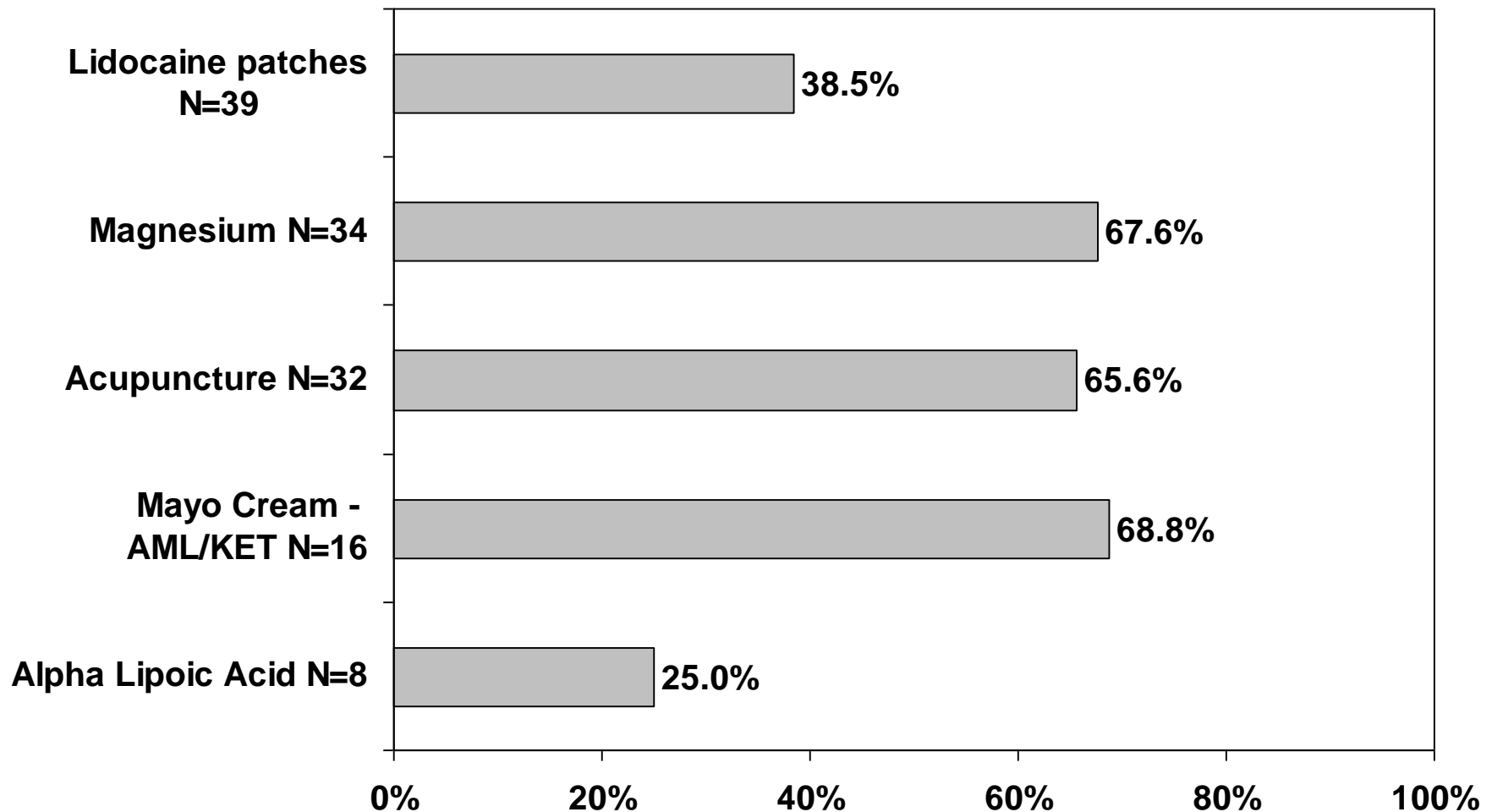
This graph shows the percentage of respondents selecting the listed item.



Supplement/Regimen Relief

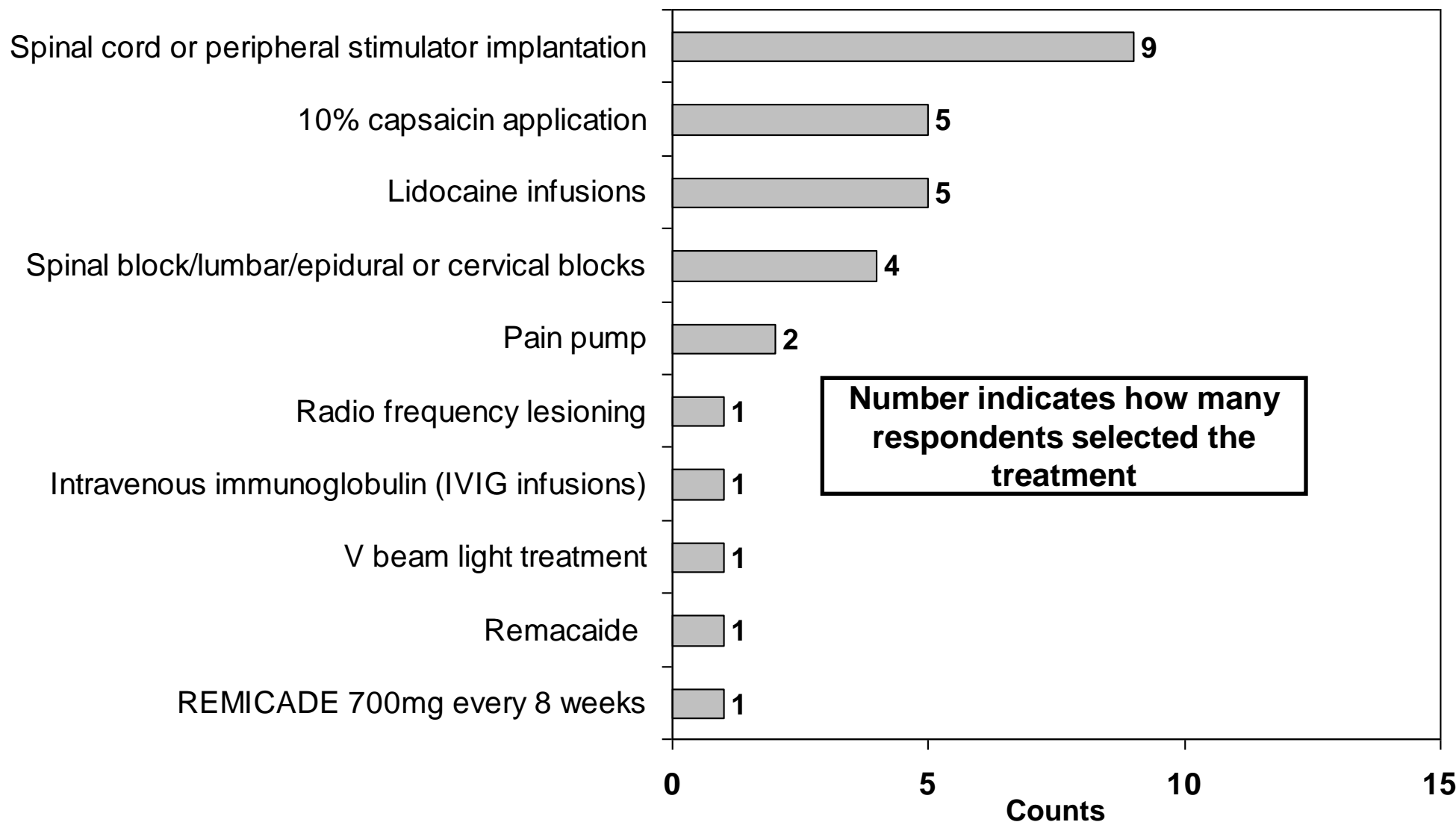
Please RATE each topical cream, dietary supplement or regimen's effectiveness for treating your EM symptoms.

Percent expressing "Good" or "Moderate" relief shown in this graph



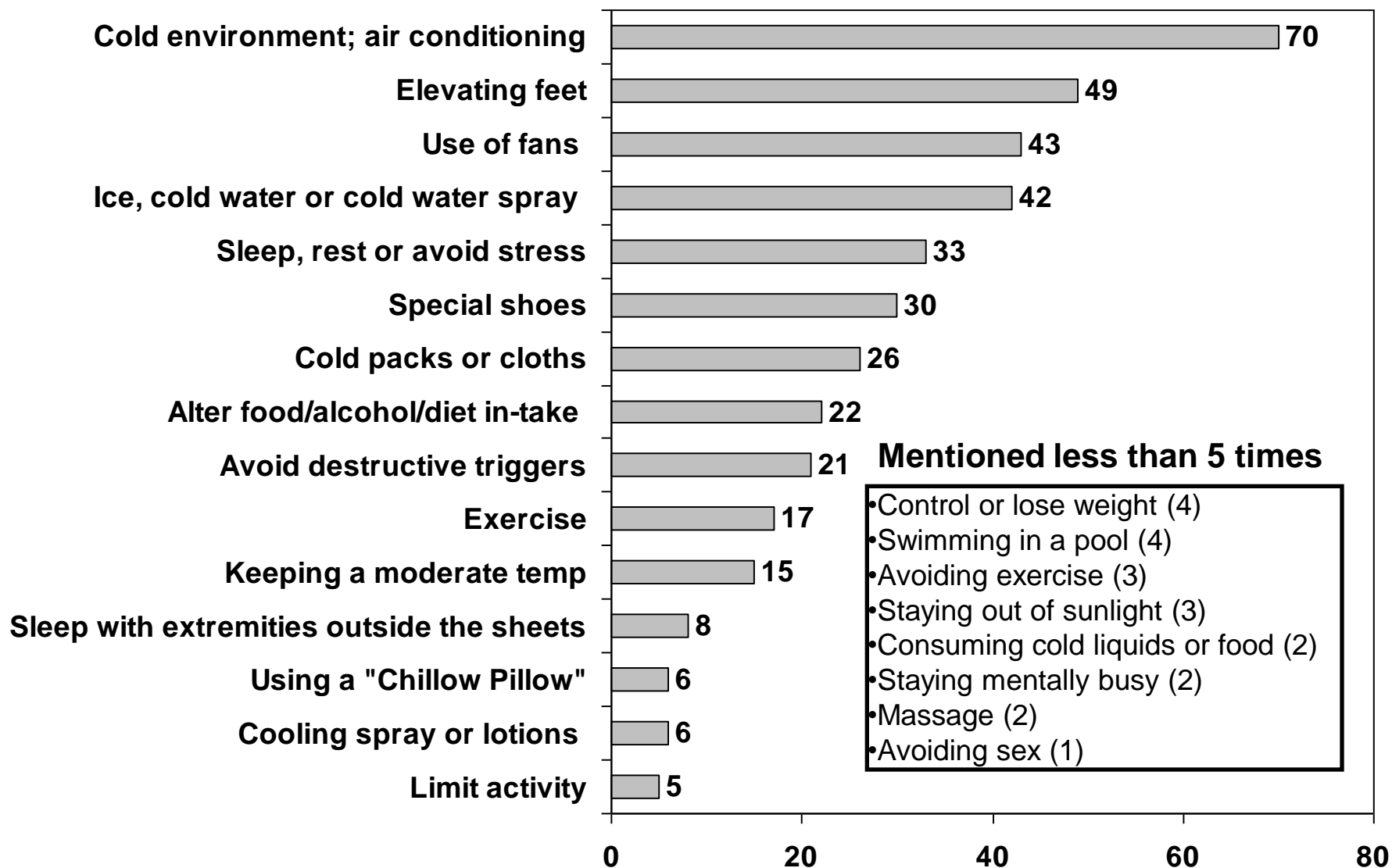
Most Successful Invasive Treatments

With which of the following invasive treatments have you had the MOST success in treating your EM or primary condition symptoms? (Select NO MORE THAN 3) N=257



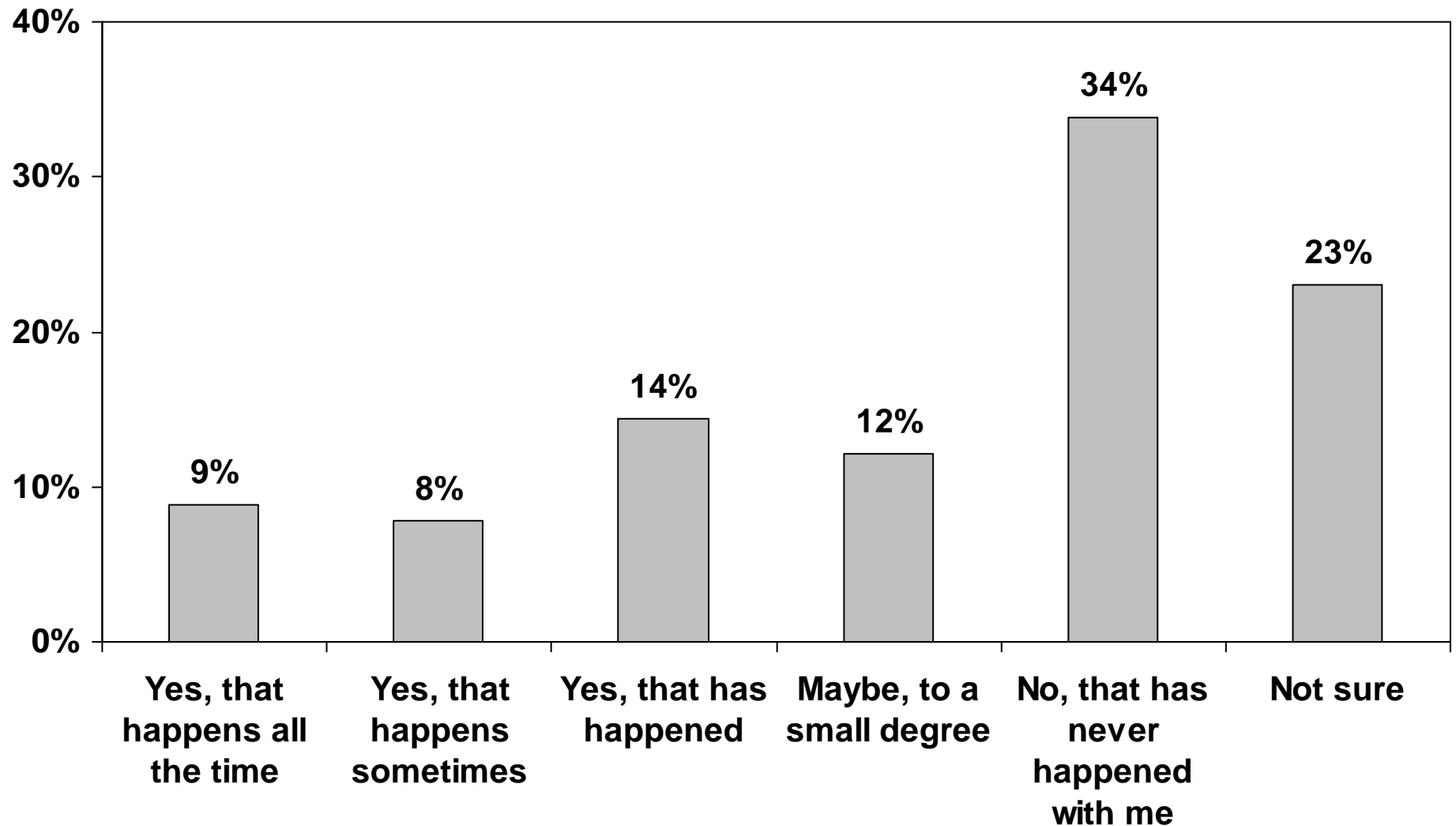
Other Treatments Used

Is there ANYTHING else other than things already covered that you have found that seems to reduce the frequency or severity of your EM symptoms? N=269



Treatments Over Time

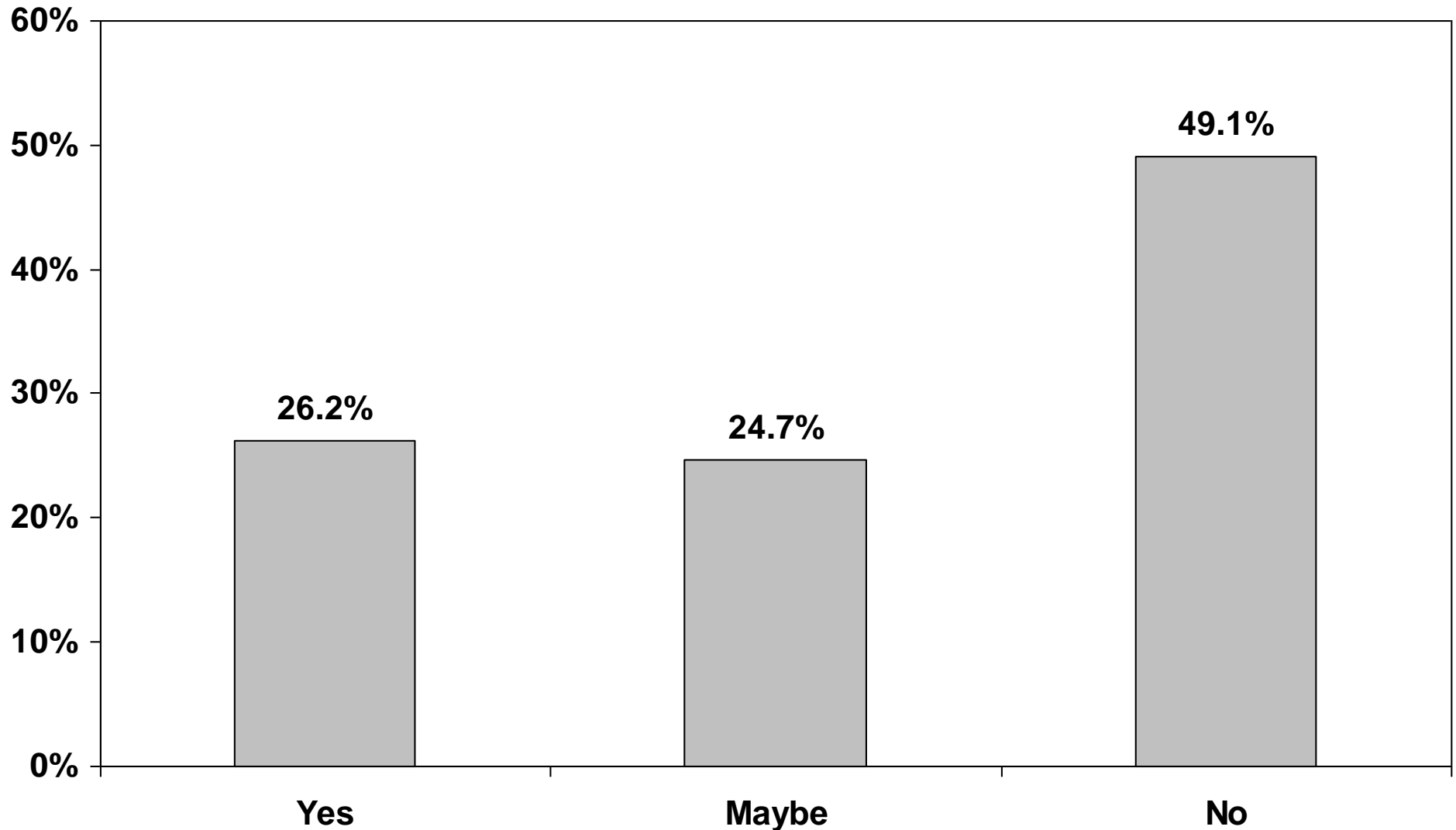
Have you found that some treatments are effective at reducing symptoms for a period of time, only to lose their effectiveness over time? N=396



Closing

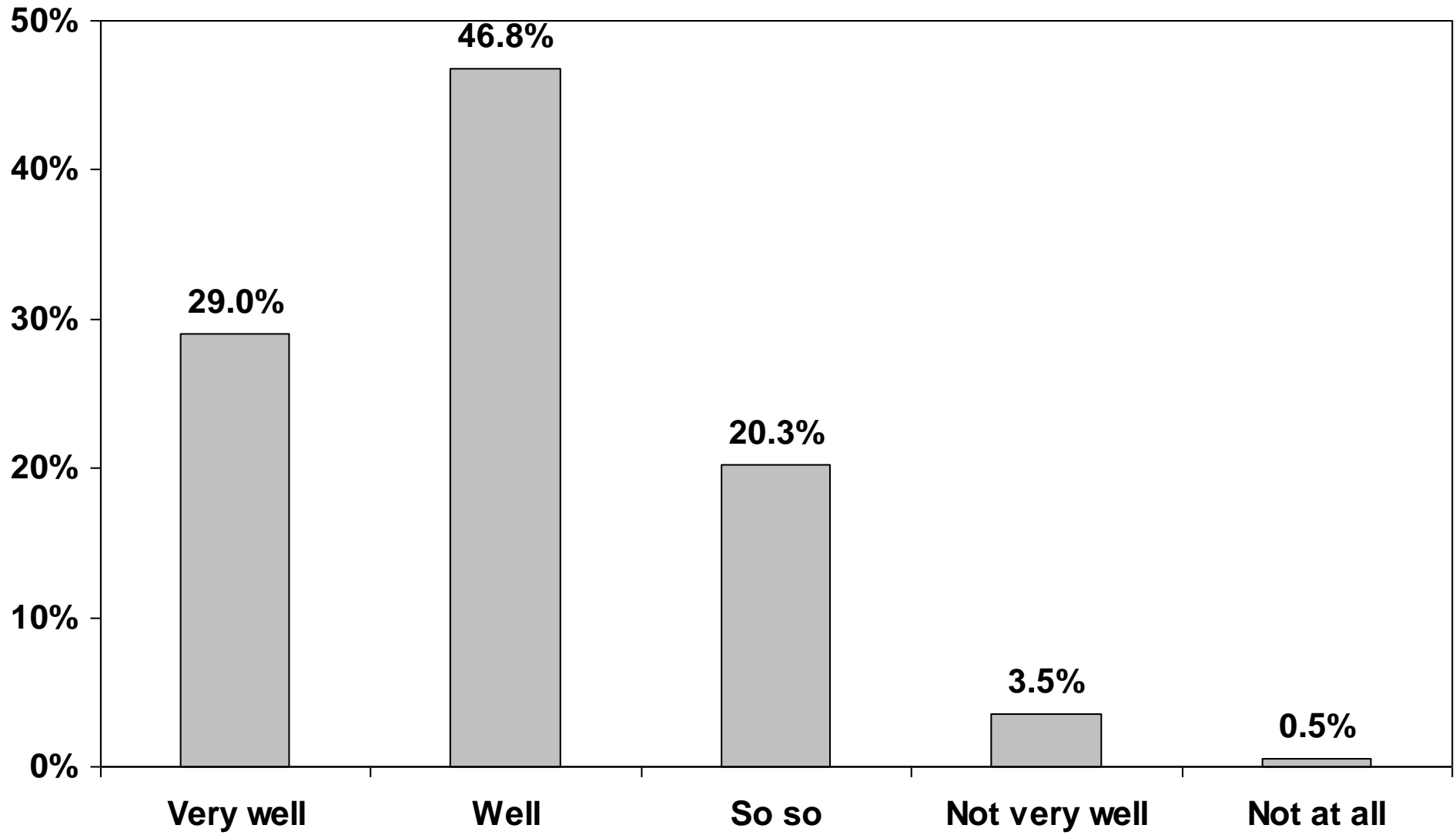
Doctor Able to Diagnose and Treat EM

Do you think your doctor is educated enough to diagnose and treat others that may have EM? N=409



Survey Relevance for Respondent

Please rate how well you feel this questionnaire provided you the opportunity to characterize your EM experience? N=400



To view the raw survey results, visit the web link below.

Of particular interest, all the write-in responses can be viewed at this site.

Note, however, that some of the bar charts will not match those in this summary. Some respondents relied on the fill-in boxes to respond to questions or otherwise elaborate on their conditions or treatments. As a result, the “Other” categories for those questions were artificially inflated, and some of the response options were undercounted.

This summary reflects adjustments based on what people wrote when given the opportunity to complete open-ended “Other” lines; the web link shows only the data as originally entered.



<http://app.sgizmo.com/reports/2747/40761/YS1IX69R3LJ3IB6AYC8HHRB7H090YB/>

Please see following pages for Appendices noted on earlier slides

Appendix 1

Drug categories – footnotes from p. 41

¹**Anticonvulsants** – drugs that aid in preventing seizures that are often prescribed for epilepsy as well as nerve pain. How some of these drugs work is not fully understood. They are thought to bind to certain areas in the brain that reduce seizures, nerve pain and anxiety. Drug names include **Klonopin (clonazepam)**, **Lyrica (pregablin)**, **Neurontin (gabapentin)**, **Tegretol (carbamazepine)**.

²**SNRIs (serotonin-norepinephrine reuptake inhibitors)** – anti-depressant medications that enhance use of both norepinephrine and serotonin in the central nervous system. Drug names are **Effexor (venlafaxine)**, **Cymbalta (duloxetine)**.

³**NSAIDS** – (Non-steroidal anti-inflammatory drugs) – drugs that relieve aches and pains and reduce fever. Drugs: **aspirin**; **Advil and Motrin (ibuprofen)**.

⁴**Prescription pain medications** – drugs with sedative or narcotic effects similar to those containing opium or its derivatives. Drugs: **Duragesic (fentanyl)**, **Utram (tramadol)**.

⁵**Tricyclic antidepressants** – older class of antidepressant medications that are less targeted in their impact on brain substances than are newer antidepressants like the SNRIs. **Elavil (amitriptyline)**, **Pamelor (nortriptyline)**, **Tofranil (imipramine)**.

⁶**Antihistamines** – drugs used to counteract the effects of immune system substances released during allergic reactions. These substances are powerful stimulants of allergy symptoms such as gastric secretions, bronchial smooth tissue constriction and vasodilation. **Benadryl (acrivastine, diphenhydramine)**, **Periactin (cyproheptadine)**, **Claritin (loratadine)**

⁷**Beta-blockers** – drugs often used for management of hypertension and cardiac arrhythmias. They block the action of substances within part of the sympathetic nervous system. **Inderal (propranolol)**, **Tenormin (atenolol)**.

⁸**Anticoagulants** – substances that delay or prevent the clotting of blood. Drugs include **aspirin**, **Coumadin (warfarin)**, **Refludan (heparin)**, **Plavix (clopidogrel)**.

⁹**Calcium channel blockers/calcium antagonists** – drugs often given to lower blood pressure. They interfere with calcium's role in maintaining vasoconstriction in blood vessels. Magnesium is sometimes called "nature's natural calcium channel blocker" because it is believed to produce a similar result. **Cardizem, Tiazac (diltiazem)**; **Norvasc (amlodipine)**.

¹⁰**SSRI (Selective serotonin reuptake inhibitors)** – anti-depressant drugs that enable serotonin to be used more efficiently. **Prozac (fluoxetine)**, **Zoloft (sertraline)**, **Paxil (paroxetine)**, **Celexa (citalopram)**.