

# **Hyperkalemic Periodic Paralysis**

## **Preliminary Survey Results & Management Guidelines**

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# History of HyperPP

- 1st family identified 60 years ago
  - **Episodes:** frequent short attacks of paralysis
  - **Triggers:** rest after exercise, stress, and certain foods
  - **Genetics:** autosomal dominant, complete penetrance
  - **Reference:** Tyler et al, 1951
- 2<sup>nd</sup> family 5 years later
  - K<sup>+</sup> levels were high in some members
  - K<sup>+</sup> administration precipitated attacks
  - **Reference:** Gamstorp, 1956
- These two reports clearly **distinguished hyperPP** from the more common **hypokalemic periodic paralysis**

# Intro to HyperPP

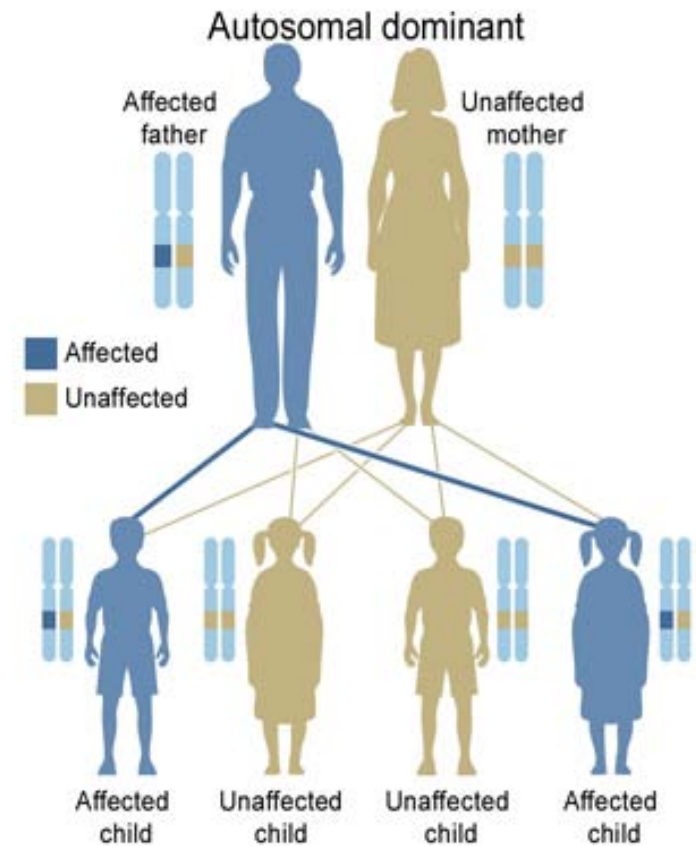
- **What:** episodes of muscle weakness
  - Induced by increased K<sup>+</sup> levels
  - +/- paramyotonia congenita (PMC)
- **Why:** autosomal dominant

*hyper* = high

*kalemia* = blood potassium level

*periodic* = occurring at intervals

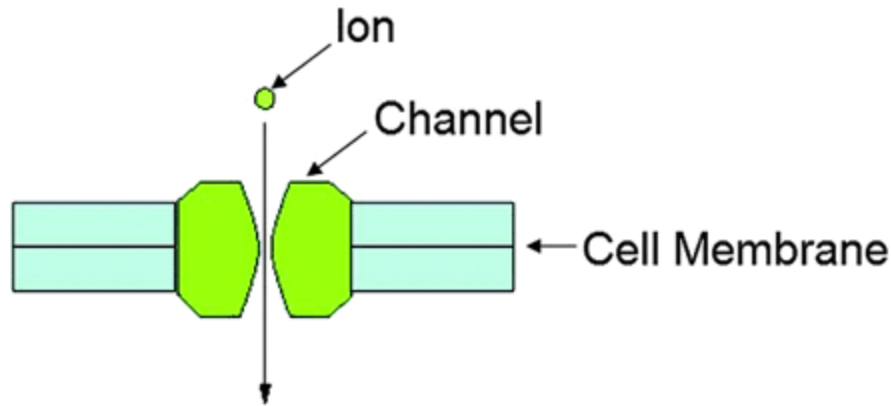
*paralysis* = loss of ability to move



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# Intro to HyperPP

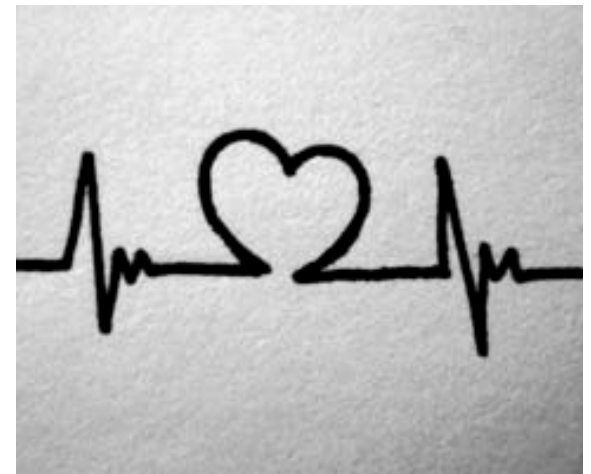
- **How:** mutated skeletal muscle sodium channel



- **Who:** onset in infancy or early childhood; prevalence 1:200,000; M = F
- **When/where:** usually triggered by cold exposure, rest after exercise, fasting, stress, ingestion of potassium, e.g. bananas or orange juice, or anesthesia

# Signs & Symptoms

- **Attacks of weakness**
  - Focal, affecting one limb, OR generalized flaccid paralysis
  - Spares consciousness, cranial muscles, respiration
  - +/- muscle pain with attacks
  - +/- arrhythmias from high  $K^+$



# Signs & Symptoms

- +/- Myotonia between attacks
  - **Myotonia**: delayed muscle relaxation after contraction (i.e., stiffness/rigidity)
- Adolescence/early adulthood:
  - Attacks most frequent
- Between ages 30 and 60:
  - Attacks decrease in frequency
  - Permanent muscle weakness

# Our Survey

- Recruitment through PPA, PPA medical advisors
- 4 main categories of questions
  - General Information
  - Diagnosis & Symptoms
  - Treatment and Management
  - Special Situations and Conclusion
- 72 responses → 43 documented mutations, from USA, Canada, Germany, India, Singapore
  - Some participants chose not to answer certain questions so not all questions had 43 responses



# Respondent Characteristics

<b>Diagnosis</b>	HyperKPP (22) HyperKPP with paramyotonia congenita (21)					
<b>Age</b>	0-20 (3)	21-30 (6)	31-40 (8)	41-50 (11)	51-60 (5)	61+ (10)
<b>Sex</b>	Male (21)	Female (20)	Blank (2)			
<b>Family History</b>	Child (25) Maternal grandparent (21) Aunt/Uncle (16) Sibling (14) Parent (9) Grandchild (9) Paternal grandparent (4)					
	<b><i>All mutation-positive respondents had family history of HyperKPP</i></b>					

# Comorbidities:

(? Due to no exercise & high carbs?)

- **High triglycerides/cholesterol (7)**
- **Thyroid problems (7)**
- **High blood pressure (4)**
- **Migraines (4)**
- **CAD (3)**
- **Arrhythmias (3)**
- **DM2 (3)**
- **Kidney problems (2)**

# Diagnosis

<b>Age at first attack</b>	0-1 yrs (13)	2-10 yrs (18)	11-20 yrs (5)	
<b>Time to diagnosis (~50% &gt; 7 yrs)</b>	< 1 yr (5)	1-5 yrs (2)	<b>7-10 yrs (5)</b>	<b>&gt;11 yrs (15)</b>
<b>Specialists</b>	<ul style="list-style-type: none"> <li>• <b>29/43 (~70%) saw 2-3 physicians before diagnosis</b></li> <li>• <b>Neurologists were felt to be most valuable (rated 4.5/5)</b></li> </ul>			
<b>Misdiagnoses (~40%)</b>	Malingering/ Faking it (8)	Conversion disorder (4)	HypoKPP (3)	Depression (1)

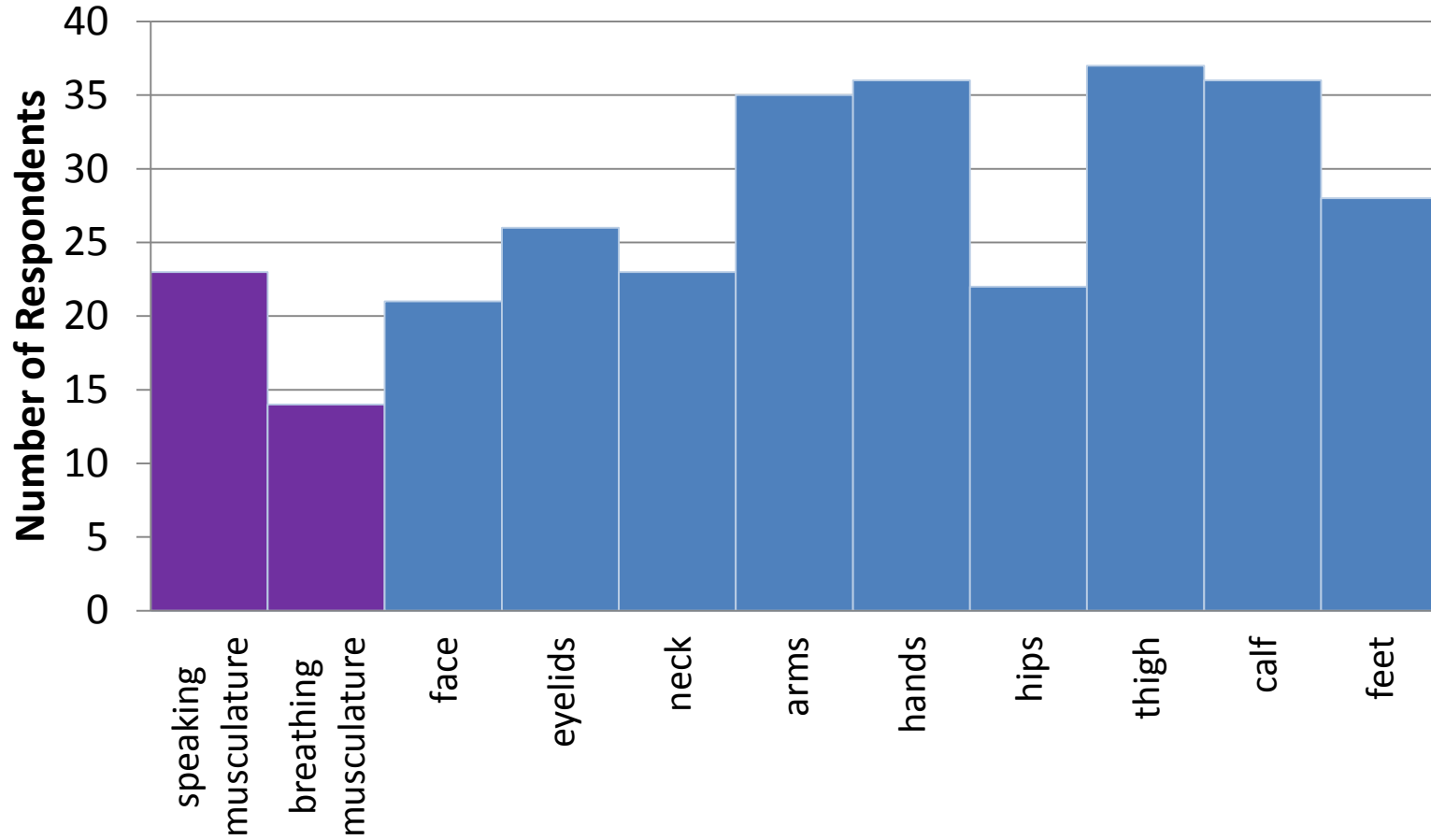
# Attack Characteristics

<b>Warning period</b>	<60 min (30) 1-5h (3)
<b>Type of attacks</b>	Weakness + stiffness (27) Primarily weakness (26) Primarily stiffness (17)
<b>Body parts</b>	Partial body (19) Total body (11) Only muscles used prior to attack (9)
<b>Severity of attacks/ function</b>	Mild (15) Moderate (21) Severe (5)

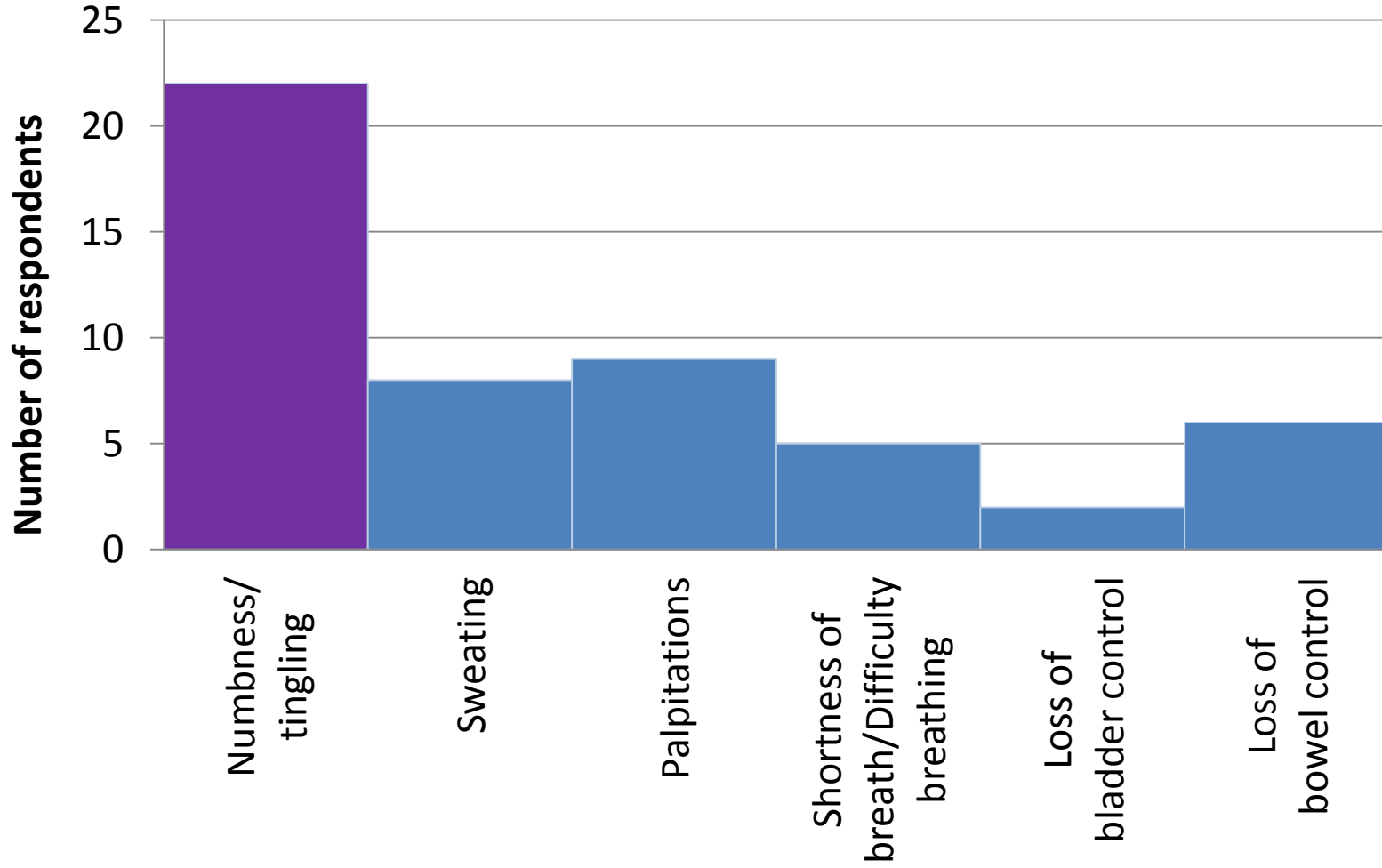
# Attack Characteristics, continued

<b>Frequency</b>	<1-3 X/month (15) 1-6 X/week (13) 1-4 X/day (9)		
<b>Duration</b>	<1 hour (12) 1-4 hours (7) 20+ hours (8)		
<b>Time of day</b>	Morning (20) Upon waking (14) During sleep (18)	Afternoon (9)	Evening (6) Night (10)

# Areas Affected by Attacks



# Associated Symptoms

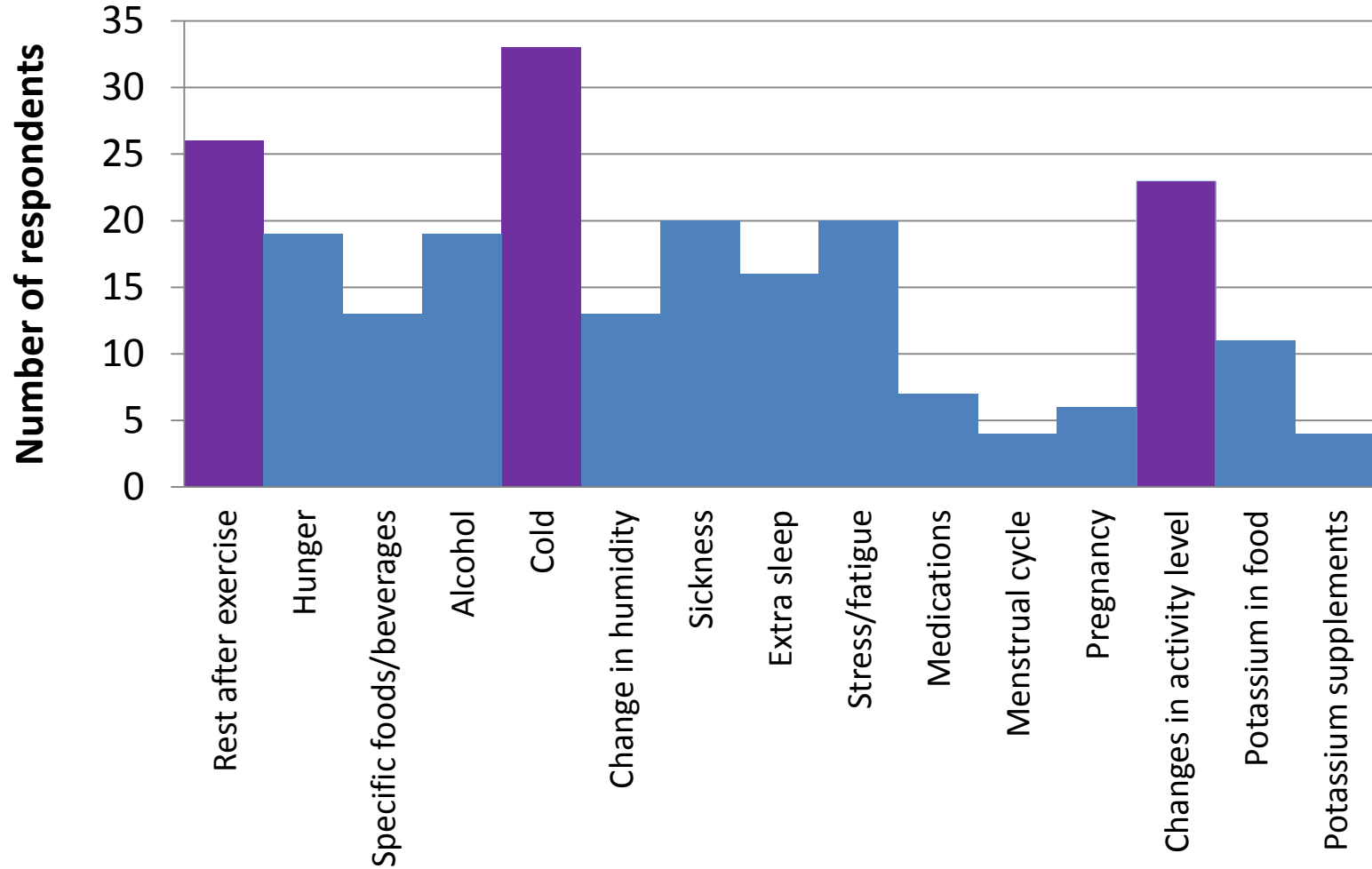


# Symptoms Surrounding Attacks

- Symptoms After Attacks
  - Muscle Pain (56%) (N=24)
  - Clumsiness (35%) (N=15)
  - Extreme fatigue (30%) (N=13)
  - Irritability (30%) (N=13)
  - Mental dullness (14%) (N=6)
- Mood Around Attacks (~30-40%)
  - Irritable (17)
  - Lethargic (15)
  - Depressed (12)



# Triggers for Attacks



# Disease Effects

<b>Weight</b>	<b>Weight gain (16) (37%)</b>	<b>No effect (25)</b>	
<b>Progressive myopathy</b>	<b>Yes (13) (30%)</b>	<b>No (17)</b>	<b>Unsure (12)</b>
<b>Hypokalemic weakness</b>	<b>No (20)</b>	<b>Yes (7) (16%)</b>	
<b>Effects on aspects of life</b>	<b>Mental health (14) Physical health (21)</b>	<b>Family life (20) Relationships (11)</b>	<b>Work (27) School (19)</b>

# Medication Management

<b>Medications for acute attacks</b>	None (5) Diuretics* (5) Magnesium (4)	Beta-2 agonists* (4) Antiarrhythmics* (3)	Anticonvulsants (2) Potassium (2) Glucose tablets (2)
<b>Ability to abort attacks</b>	All of the time (0) Most of the time (12)	Some of the time (12) Occasionally (5)	<b>Never (9)</b>
<b>Chronic medications</b>	<b>Diamox (12)</b> <b>HCTZ (10)</b> Salbutamol (6)	Mexitil (5) Daranide (2) Glucose tablets (1)	Lasix (1) IV Ca gluconate (0)
<b>Level of control</b>	<b>Needs improvement (15)</b>	Mostly controlled (15)	Optimal (2)
<b>Time to regimen</b>	<b>&lt;2 years (3)</b>	<b>2-4 years (2)</b>	<b>9-20 years (4)</b>
<b>Rx to avoid</b>	<i>(Open-Ended)</i> : <b>Potassium, corticosteroids</b> , antibiotics, statins, diuretics		

\*Diuretics: HCTZ, Diamox, Daranide, torsemide, Lasix

\*Beta-2 agonists: salbutamol, fenoterol

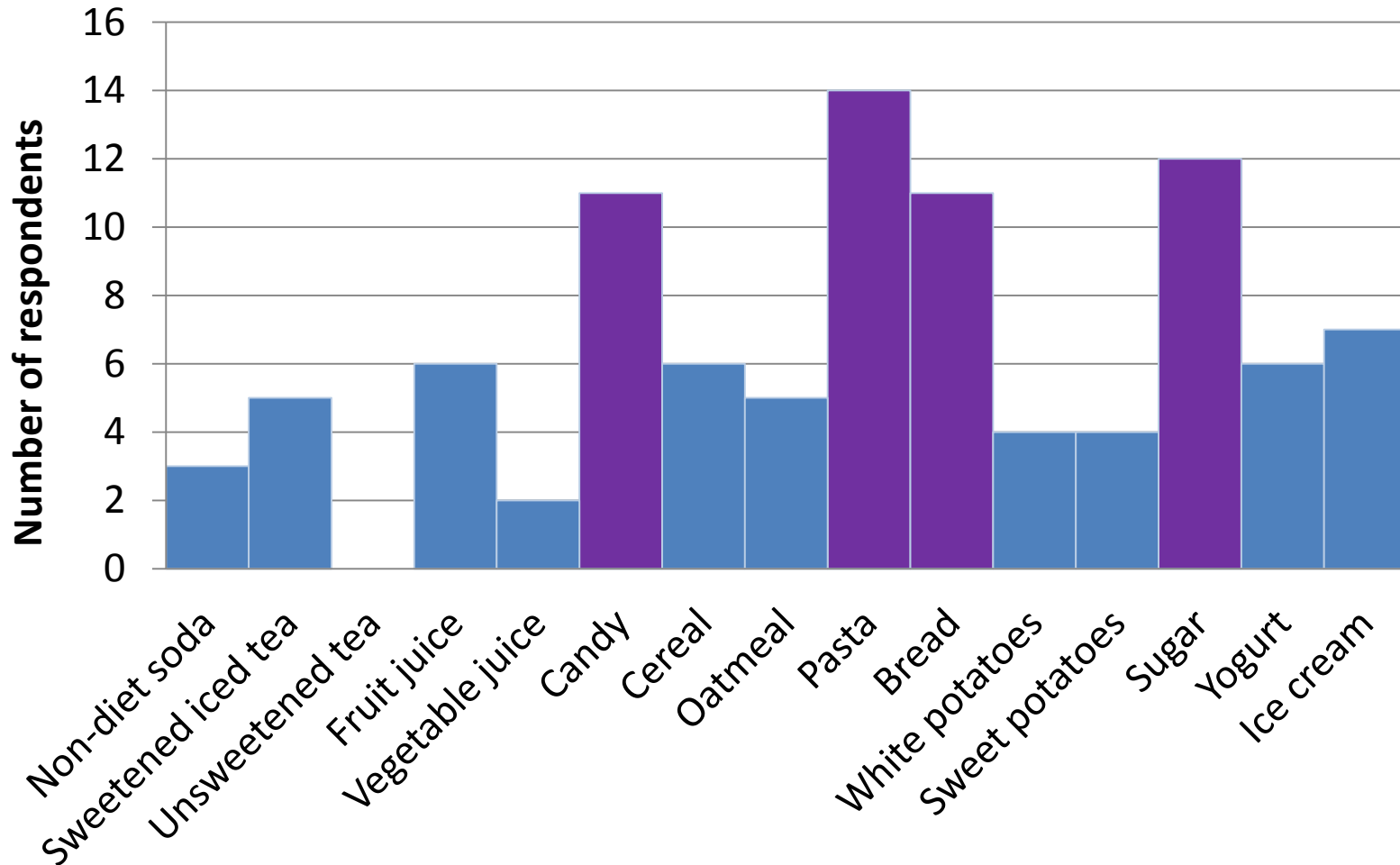
\*Antiarrhythmics: propafenone, flecainide, Mexitil

\*Anticonvulsants: Lamictal, Topamax, Trileptil, carbamazepine

# Dietary Management

Meal frequency	1 meal per day (0) 2 meals per day (2) 3 meals per day (9)	4 meals per day (6) 5 meals per day (2) 6 meals per day (1)
Frequency of carbohydrate-rich snacks	Never (1) Every 2 hours (1) Every 2.5-3 hours (4)	
Food quantity	Small (4) <b>Medium (18)</b> Large (4)	
Faster response with liquid vs. solid sweets	<b>Liquid sweets (9)</b> <b>Solid sweets (3)</b> <b>No difference/ have not noticed (17)</b>	
Foods to avoid	Bananas (15) Alcohol (10) Diet soda (6)	Dried figs (6) Tomato/vegetable juices (5) Apricots (5)

# Helpful Foods



# Other Management

<b>Acute management</b>	<b>Keeping warm (17)</b> <b>Sugary food (12)</b> <b>Gentle exercise (3)</b>
<b>Home accommodations</b>	None (19) <b>Items kept within reach near bed (11)</b> Exercise equipment (5) Rug placement (4) Bathroom railings (4) Emergency access button or necklace (1) Wheelchair accessibility (1)
<b>Exercise</b>	<b>Walking (17)</b> Yoga (3) Swimming (5) Biking (8) Jogging/Running (3) Aerobic exercise (3)

# Special Situations

<b>Pregnancy</b>	More severe (4)	Less severe (6)	Same severity (7)
	More frequent (7)	Less frequent (4)	Same frequency (6)
<b>Local anesthesia</b>	Did not trigger attack (27)	<b>Triggered attack (4)</b>	
<b>General anesthesia</b>	Did not trigger attack (18)	<b>Triggered attack (9)</b>	



# Prevention: Diet

- **Avoid foods rich in potassium**
  - Fruits, fruit juices
- **Avoid medications that elevate potassium levels**
  - Spironolactone, enalapril, valsartan
- **Avoid fasting**
- **Some find that high salt intake reduces attack frequency & severity**



# Prevention: Activity

- **Exercise**
  - Unclear how to advise
  - Some individuals find that continuing mild exercise after a period of more vigorous exercise can lessen an attack
- **Avoid exposure to cold**

# Prevention: Chronic Therapy

- Helpful medications:
  - **Thiazide** diuretics
  - **Acetazolamide**
  - **Albuterol inhaler** (1 to 2 puffs of 0.1 mg)
  - Your physician should **monitor your electrolyte** levels to ensure they remain in a safe range

# Management of Acute Attacks

- **MILD** - abort or prevent attacks at onset with:
  - sugar
  - mild exercise
- **MODERATE/SEVERE** – abort with:
  - thiazide diuretics (hydrochlorothiazide)
  - inhaled beta adrenergic agonists (salbutamol)
  - intravenously injected glucocorticoids?
  - intravenous calcium gluconate [for heart]

# Hypotheses

- **Treadmill at home:** exercise but help will be near
- **Pace workout:** use music for slow rhythm to limit overexertion
- **Meditation CD:** prevent a stress-induced attack when feeling overwhelmed or tense
- **Tiger Balm or Advil:** for post-attack muscle stiffness and soreness

# Special Situations: Surgery

- Make anesthesia staff aware of diagnosis
- Opioids or depolarizing agents used in anesthesia → myotonia  
→ interfere with intubation / ventilation
- Breathing problems / weakness when recovering from general anesthesia
  - Prevent carbohydrate depletion – D5W is good!
  - Avoid muscle relaxants
  - Maintain a normal body temperature
  - Keep serum potassium levels from being elevated
- Asymptomatic family members should be tested for the family's mutation before surgery so preventive measures can be instituted

# Special Situations

- When you change routine, such as sleeping late on holidays or weekends, attacks can occur more frequently
  - Wake up at your usual hour and have your regular breakfast meal
- Albuterol is safe in pregnancy

# Tips from Fellow HyperPPers

- *Know what works for you*
- *Get all the support possible*
- *Don't be afraid to talk about it*
- *Stand up for your rights*
- *Learn all you can*
- *Diagnosis is a long road, but...*
- *Get diagnosed!*

# Tips from Fellow HyperPPers

- ***Know what works for you.** My son & I both have pp but have different severity and longevity.*
- *Get regular exercise, eat frequent small meals, reduce stress.*
- *Do not sit for longer period. Keep moving. Never skip food. Do not do heavy exercise.*
- *Expect that most doctors you see are not going to believe you. But it is real, there are many people who have it and it can be managed.*
- *I spent many years being dismissed by various doctors for my symptoms. It takes a long time to find someone who has heard of these conditions, and then figuring out how to manage them is another **long road**.*
- ***Get diagnosed***



# Tips from Fellow HyperPPers

- *Don't wait to find out as much as you can. I was misdiagnosed for over 40 years with hypo and it has now affected my health*
- ***Get all the support possible, and don't be afraid to talk about it; stand up for your rights and don't put yourself in a situation you know you will suffer in.***
- *Don't lie about it, explain to others what it is (which in my childhood/teen years was impossible because no one knew what it was). Join the PP list to share experiences with others and learn from them.*
- ***Learn all you can!*** *Talk with others, learn how to manage your diet. You can take control of many things, even though mid-attack you have little to no control. When you're feeling strong - go out and do things! Don't put things off, because you never know when the next big attack might come. We already must suffer being paralyzed physically, so let's not be paralyzed by fear, as well.*

# Questions?

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