

PERIODIC PARALYSIS

A RARE CONUNDRUM

(MY APPROACH)

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- PLEASE: If you want to see me, make an appointment by phone call. If you are not already my patient (i.e.: I haven't seen you in my office), I – personally – will not answer your questions or requests. Do NOT send me any personal records about you/your condition unless my office requests it from you.

STEPHEN F. LEWIS, MD

- Education: Davidson College; University of NC, Greensboro; Wake Forest University School of Medicine; Abington Memorial Hospital (now Abington-Jefferson Health); Thomas Jefferson University Hospital.
- CURRENT: SOLO, PRIVATE PRACTICE. (that means I get to do what I want to do, when – and for how long – I want to do it)
 - I have now seen almost 7 dozen patients with complaints consistent with a Periodic Paralysis
 - My gut reaction is that only a third have “Genetically Proven” PPP
 - Occasionally, patients have come to see me THINKING they had a PP, but had some other rare disorder divined after our investigations



CONTENTS

- History
- Types
- Cause
- Diagnosis
- Treatment
- Prognosis



LLAMA



HISTORY

- Cavare, 1853.
- Potassium salts used 1901 (Buzzard) and 1902 (Mitchell)
- 1930's fluctuations in K^+ noted, including familial cases without K^+ changes
- Thyrotoxicity as triggering event also noted

TYPES

PRIMARY

HYPOKALEMIC

HYPERKALEMIC

ANDERSON-TAWIL

NORMOKALEMIC

[CHLORIDE-CHANNEL]

SOMETHING ELSE

TYPES

SECONDARY

THYROTOXIC

SOMETHING ELSE

WE DON'T KNOW

“YOU'RE JUST CRAZY”



ALPACA



CAUSE

- Genetics
 - Sodium Channel
 - Calcium Channel
 - Potassium Channel
 - Chloride Channel
 - We Don't Know

CAUSE

- Something Else



GUANACO



DIAGNOSIS

- Here's where it gets tricky.
 - Go to a PHYSICIAN (a person who went to medical school and became licensed to practice medicine)
 - Tell him your HISTORY
 - Be chronological, including any KNOWN Family Disorders
 - *Avoid Medical Terms!*
 - Don't be disparaging of health care workers
 - Provide documents as you're able
 - Don't send them ahead of time
 - Don't expect him to read them all right then and there

DIAGNOSIS

- Here's where it gets tricky.
 - Allow him to perform a thorough EXAMINATION if requested
 - Don't try to "demonstrate" your symptom unless asked
 - Don't feel like if you don't have weakness at the time of the exam that you should – or that the physician needs to see it to believe it

DIAGNOSIS

- Here's where it gets tricky.
 - DO NOT PULL THE “DR. GOOGLE CARD” (do not suggest possible diagnoses gleaned from your reading on the web)

DIAGNOSIS

- “Tests”
 - Genetics
 - Metabolic Panel (Between and, if possible, During episode)
 - Electromyography with Nerve Conduction Studies, Repetitive Nerve Stimulation
 - Urine Electrolytes
 - Thyroid Functions
 - (EKG, Echocardiogram, ?Cardiologist, ?Nephrologist, ?Psychiatrist, ?Dietician, PT/OT)
 - Occasionally: Neuroimaging, Spinal Puncture



VICUNA



TREATMENT

- Depends on the Diagnosis

TREATMENT

- HYPOKALEMIC TYPE
 - Potassium Salts
 - Carbonic Acid Anhydrase Inhibitor
 - KEVEYIS (dichlorphenamide)
 - Diamox (acetazolamide)
 - Topamax (topiramate)
 - Zonegran (zonisamide)

TREATMENT

- HYPOKALEMIC TYPE
 - Potassium-Sparing Diuretics
 - Aldactone (spironolactone)
 - Inspra (eplerenone)
 - Amiloride
 - triamptere

TREATMENT

- HYPERKALEMIC TYPE
 - Carbonic Acid Anhydrase Inhibitors (KEVEYIS, Diamox, Topamax, Zonegran)
 - “Sugars” (Coke, Caro Syrup, Liquid Chocolate, Glucose Water, etc.)
 - Albuterol inhalation
 - Potassium-Wasting Diuretics
 - HCTZ
 - Lasix (furosemide)
 - [ACE Inhibitors]

TREATMENT

- ANDERSON-TAWIL SYNDROME
 - Seems to require a combination of the above (which way do they swing?)
 - Beta Blockers, Cardiac Pacemaker

TREATMENT

- OTHER DRUGS
 - Pain relief: Muscle Relaxants, NSAIDs, gabapentin, anti-muscle spasm Meds: baclofen, tizanidine, benzodiazepines, Vimpat (lacosamide), mexilitene
 - Sleep Aides: ?, Ambien (zolpidem)
 - SSRIs (I typically avoid SNRIs)
 - Mestinon (pyridostigmine)

TREATMENT

- GENERAL PRINCIPLES
 - Rest
 - Know your limits
 - DIET (Carbs/Sugars, Salt (sodium/potassium))
 - Look for TRIGGERS
 - Foods
 - Exercise
 - Hot/Cold
 - Stress (Makes the world go 'round...but...)

PROGNOSIS

- Wide Variety of out-comes
- In general, most people lead productive lives if they can work around their triggers
- Remember: This is a part of you, like your fingers, eyes, and brain. Learning how to live with the difficulties you may encounter is imperative. Learning how to cope with the stumbling blocks of life is imperative.

VICUÑA



ALPACA



GUANACO



LLAMA



WILD

DOMESTIC

WEBSITES

- Medline Plus - <https://medlineplus.gov/>
 - Basic information on Hyperkalemic, Hypokalemic, and Thyrotoxic periodic paralysis
- Genetics Home Reference - <https://ghr.nlm.nih.gov/>
 - More detailed information emphasizing genetics with links to other authoritative sources. Written for lay people but good for clinicians, too. Links to scientific articles and clinical trials.
- GARD – Genetic and Rare Diseases Info Center - <https://rarediseases.info.nih.gov/>
 - Clinical information, links to research, and supportive organizations. Information in English and Spanish