

Nicholas P LeFevre DO₁
UPMC-Lititz

Introduction

Hypokalemic periodic paralysis (HkPP) is a rare neuromuscular disorder causing transient skeletal muscle weakness. It belongs to a group of disorders consisting of ion channel defects known as channelopathies.

Case Presentation

- 73 year old female with a past medical history of systemic lupus erythematosus, Sjogren's syndrome, and hypokalemic periodic paralysis, presents to the emergency department
- Reports quadriparesis 2 hours ago upon waking and persistent dehydration
- On exam, muscle strength 2/5 in upper extremities and 0/5 in lower extremities
- The previous day the patient underwent a colonoscopy
- After emergence from anesthesia, she was nonverbal for 10 minutes, and unable to move extremities for 20 minutes; no labs were drawn at that time
- Attacks occur almost daily since she was 20 years old, typically lasting 30 minutes, and most often after waking
- A quick review of the patient's chart reveals a history of hypokalemia
- Started IV fluids and ordered a BMP

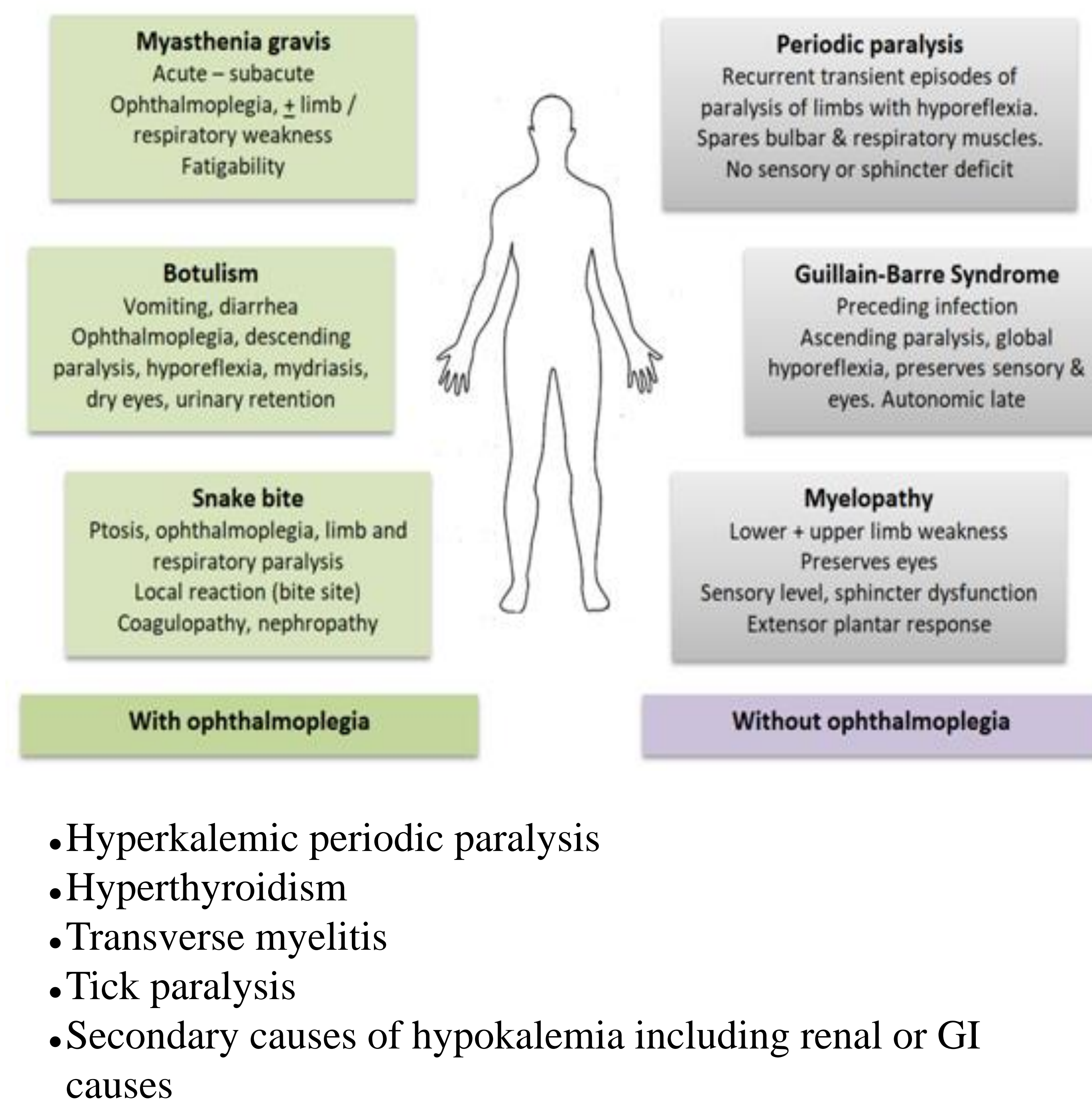
Labs

Na	138
K	3.6
Cl	110
Mg	2.2
Ca	9.1

Discussion

- HkPP follows an autosomal dominant inheritance pattern and has a prevalence of 1 in 100,000 people
- Onset up to the late 20's but usually occurs by teenage years
- 70% of cases are caused by a mutation of the CACNA1S gene that affects dihydropyridine sensitive voltage-gated calcium channels in skeletal muscle
- A stressor increases potassium uptake into the myocyte via Na/K channels; neurons become paradoxically depolarized and the altered channel is refractory to signaling
- Episodes occur suddenly, may last hours to days, and are triggered by exercise, stress, immobility, or high-carbohydrate meals
- Patients may complain of paresthesia, dry mouth, extreme fatigue, mental dullness, or dyspnea during an episode
- Potassium levels are typically normal between episodes and may be normal during an episode; mean potassium level during an episode is 2.6
- Consciousness is preserved and respiration is minimally affected
- Muscle weakness usually affects proximal > distal muscles and legs > arms
- Acquired cases have been seen in a hyperthyroid state known as thyrotoxic periodic paralysis
- Acute treatment should include 60 to 120 mEq of potassium and IV fluids
- Long term treatment includes acetazolamide and spironolactone

Differential Diagnosis



Conclusion

Although rare, hypokalemic periodic paralysis should be considered in a patient presenting with a history of intermittent muscle weakness coupled with any degree of hypokalemia. Pre-operative optimization may have prevented the need for additional medical intervention.

References

- Abbas H, Kothari N, Bogra J. Hypokalemic periodic paralysis. *Natl J Maxillofac Surg.* 2012;3(2):220–221. doi:10.4103/0975-5950.111391
- Guttman L, Conway R. Hypokalemic Periodic Paralysis. In: Wilterdink J, ed. *UpToDate.* Providence, Rhode Island: UpToDate, 2019. <https://www.uptodate.com/contents/hypokalemic-periodic-paralysis>. Accessed March 30, 2020.
- HA D, Padmaperuma P. *Clinical Syndromes Of Acute Flaccid Paralysis.* 2018. [image] Available at: <<https://medcraveonline.com/EMIJ/images/EMIJ-06-00189-g002.png>> [Accessed 1 April 2020].
- Matthews E, Portaro S, Ke Q, et al. Acetazolamide efficacy in hypokalemic periodic paralysis and the predictive role of genotype. *Neurology.* 2011;77(22):1960–1964. doi:10.1212/WNL.0b013e31823a0cb6