Hypokalemic Periodic Paralysis

Experience from a secondary level Hospital

Dr. Philip Finny
Duncan Hospital, Raxaul
Bihar, India
India still has 70% rural population
1440 inpatients per month
Learning Outcomes

- Identify the clinical context in which to consider Hypokalemic Periodic Paralysis (HPP)
- Planning the appropriate lab evaluation for HPP
- Review the comprehensive management of HPP
- Recognize the serious complications of HPP if improperly treated
- Understand the pathway from research to practical application in the hospital setting
25 year old gentleman presented with acute onset of weakness of all 4 limbs on waking up in the morning

Clinically, he has grade 3 weakness in the upper limbs and grade 2 weakness in the lower limbs, Deep Tendon Reflexes hyporeflexic, flexor plantar response

No history of trauma, recent viral infections, diarrhea

No past history of similar problem
What could be the possible differential diagnosis?

- This is a syndrome of acute areflexic quadriplegia
- Possible causes include:
  - Guillain Barre Syndrome
  - Hypokalemic Periodic Paralysis (HPP)
  - Acute Myasthenic Crisis
  - Acute Polymyositis
  - Acute Intermittent Porphyria
Clinical and Laboratory Evaluation

- **GBS**
  - Ascending paralysis
  - Preceding history of viral infection, diarrhea
  - EMG/Nerve Conduction Studies are useful to confirm the diagnosis

- **HPP**
  - Serum electrolytes
  - Any other indirect way?
A case from 1994

- The Duncan Hospital was without an electrolyte analyzer
- What is the test?
- EKG
  - reveals U waves
Clinical and Laboratory Evaluation

- **Myasthenic Crisis**
  - More common among young women
  - Past history of diplopia, dysphagia, and other symptoms suggestive of myasthenia
  - EMG and repetitive nerve stimulation
  - Ach antibodies
  - Associated thyroid dysfunction is common

- **Polymyositis**
  - Muscle pain
  - Proximal myopathy
  - Elevated CPK
Acute intermittent porphyria
- History of intermittent abdominal pain
- Polyneuropathy
- History of urine changing color on exposure to sunlight
- Urinary porphobilinogen and 24 hour urinary delta ALA elevation
The junior doctor had admitted two young men on one night with acute areflexic quadriparesis

He wanted to refer them to a higher center with a provisional diagnosis of GBS

It was the first day of the new physician who had just joined the Duncan Hospital
As the ECG was suggestive of hypokalemia, he treated with KCl and both the patients were able to walk home within 24 hours.

A referral was averted.

There were many other similar patients seen over the coming year at this hospital and this prompted the physician to study this problem systematically.
Preponderance of Hypokalaemia as a Cause of Acute Onset Quadriplegia in Northern India/Southern Nepal

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Abstract

Of 68 patients who were admitted with acute quadriplegia to a hospital in northern India, over 70% were found to be hypokalaemic. The most common cause of hypokalaemia was that associated with gastroenteritis (54%). These patients had all received intravenous fluids previously. It is likely that their hypokalaemia was caused by gastrointestinal loss compounded by parenteral fluid replacement. The next most common group of hypokalaemia-associated quadriplegia had no obvious cause for hypokalaemia (38%).

Hypokalaemia-induced quadriplegia is a potentially life-threatening illness which can be readily treated with potassium supplements. The physician should consider hypokalaemia in patients who present with acute onset quadriplegia, and even if diagnostic tests for hypokalaemia are not available, should consider a judicious trial of potassium supplementation empirically, provided that there are no contraindications.
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Background

- **Definition**
  - The acute weakness of all four limbs associated with an initial $K < 3.5$ mmol/L
  - HPP is a potentially life-threatening medical emergency
  - It is the most common cause of acute areflexic quadriplegia in North Bihar
  - Many of these patients are sub optimally treated and many have recurrent episodes
Aetiological, clinical and metabolic profile of hypokalaemic periodic paralysis in adults: A single-centre experience

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ABSTRACT

Background. Hypokalaemic periodic paralysis constitutes a heterogeneous group of disorders that present with acute muscular weakness. In this analysis, we discuss the aetiological factors that appear to be more common in the Indian population.

Methods. From 1995 to 2001, 31 patients presented with periodic paralysis (mean age 34.5 years, range 11–68 years). Of the 31 patients, 19 were men. The clinical and laboratory data of these patients were analysed. Patients were investigated for possible secondary causes of hypokalaemia.

Results. There were 13 patients (42%) with renal tubular acidosis, 13 with primary hyperaldosteronism (42%), 2 each with thyrotoxic periodic paralysis and sporadic periodic paralysis, and 1 with Gitelman syndrome.

Of the 13 patients with renal tubular acidosis, 10 had proximal and 3 distal renal tubular acidosis. Three of these patients with renal tubular acidosis had Sjogren syndrome. The patients diagnosed to have renal tubular acidosis had significantly lower serum bicarbonate (18.7 [4.6] v. 29.6 [5.0] mEq/L; p < 0.05) and higher levels of chloride (107.5 [6.0] v. 99.5 [3.4] mEq/L; p < 0.05) compared with those who had primary hyperaldosteronism, although the potassium values were similar.

METHODS

Thirty-one patients with HPP were identified over a 6-year period (1995–2001). Their mean age was 34.5 years (range 11–68 years; 19 men). The mean duration of illness before presentation was 23 months (range 1–92 months) and they had had a mean of 4 episodes (median 3, range 1–15 episodes). The data of all patients who presented with HPP were analysed retrospectively (Table 1). HPP was defined as acute loss of muscle power with inability to walk and documented plasma potassium levels <3.5 mEq/L during the episode. The clinical data collected included age, sex, ethnic
Scenario at the Duncan Hospital in 2008

- HPP was prevalent in the this region of Bihar with nearly 20–25 patients admitted annually in our hospital
- Quite a few of them were recurrent admissions
- They were being treated with IV and oral potassium supplements and discharged within 24–48 hours
- However, a work up for secondary causes was not being done
- This prompted a thorough investigation into this problem
Aims and Objectives of the Study

- To identify underlying causes for HPP in patients presenting to a secondary care centre in rural North India
- To develop a simple algorithm for evaluation and treatment of HPP at a primary/secondary care hospital in India
Methodology

- We analyzed data collected from 84 patients over 3 years (Feb 2009 – Jan 2012)
Results

Age Range

Patient age: 14-70 yrs
Mean age: 36.7 yrs

Distribution by Gender

- Male: 46%
- Female: 54%
Secondary Causes of HPP

Renal Tubular Acidosis: 58
Idiopathic HPP: 9
Conn's Syndrome: 2
Acute Gastroenteritis: 4
Gitelman's/Bartter's: 8
Thyrotoxicosis: 8
Familial HPP: 8
Unclear etiology: 0

N=84
Seasonal Changes in Occurrence of HPP

![Graph showing seasonal changes in total cases and weather variables](image-url)
Seasonal Changes in Occurrence of HPP

Seasonal Variation

- Hot Season (April - October): 13%
- Cold Season (Nov - March): 87%
Key Findings

- 67 (79.8%) patients with HPP had a definite underlying secondary cause
- 80% of patients had a severe hypokalemia (Potassium ≤ 2 mmol/L)
- 32 (38%) patients were aged 30 – 39 years
- 73 (86.9%) of our patients presented in summer months
Key Findings

- 2 patients presented with respiratory paralysis that required ventilation
- 30 (35.7%) patients had recurrent episodes of HPP with the number of recurring episodes ranging up to 10
Hypokalaemic periodic paralysis in rural northern India – most have secondary causes

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Abstract

Hypokalaemic periodic paralysis (HPP) is a life-threatening condition. Our aim was to study the clinical profile and laboratory parameters of HPP patients and to develop an algorithm to determine the causes of HPP. 84 patients presented with HPP over a 3 year period. 58 (69.0%) were found to have renal tubular acidosis (RTA). The other causes were idiopathic HPP (8 (9.5%)), acute gastroenteritis (4 (4.8%)), suspected primary hyperaldosteronism and familial HPP (2 each (2.4%)) and suspected Gitelman/Bartter Syndrome and thyrotoxic periodic paralysis (1 each (1.2%)). The number of cases peaks in the hot season. Over a third of the patients (35.7%) had recurrent episodes. 80% had secondary HPP and therefore a biochemical evaluation is mandatory. A simple algorithm was developed. Both health professionals and patients need further education regarding this problem in order to improve diagnosis and treatment and to improve compliance.
A simple algorithm for determining secondary cause of HPP

1. Acute quadriparesis, paraparesis, or impending respiratory paralysis and serum $K^+ < 3.5$ mmol/L
2. Rule out acute gastroenteritis and drug use (e.g., diuretics)
3. HPP
4. ABG
   - Metabolic acidosis
     - RTA (High Cl, Low $\text{HCO}_3$)
   - Normal
     - 1) Thyrotoxicosis
     - 2) Idiopathic
     - 3) Familial
   - Metabolic alkalosis
     - With Hypertension
       - Primary hyperaldosteronism
     - Without Hypertension
       - Bartter or Gitelman Syndrome
Take Home Points

- HPP is a life threatening condition and a GBS mimic
- 70 percent of acute flaccid quadriplegiasis were due to HPP in this region
- 80% had secondary causes in our series of 84 India and Nepali pts
- 35% were recurrent presentations
- Peak incidence in summer months
- Exclude secondary treatable causes such as RTA, Conn’s and Thyrotoxicosis
- Familial HPP was of low incidence in our series
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Questions?