**Case Report**
Volume 14:03, 2024

Journal of Clinical Case Reports

ISSN: 2165-7920 Open Access

# Hypokalemic Periodic Paralysis in Afghanistan, Study of Case Report

**Mirwais Ramozi*, Sayed Mohammad Reza Hosseini, Abdullah Rastin and Abass Ali Ramozi**

*Department of Medical Research, Kateb University, Kabul, Afghanistan*

## Abstract

Hypokalemic Periodic Paralysis (HPP) is a form of periodic paralysis, a rare autosomal dominant channelopathy caused by the skeletal muscle ion channel mutations. This is a heterogeneous group of muscle diseases. It is characterized by episodes of flaccid and sudden muscle weakness by a fall in potassium level in the blood.

Here, a case of 15-year-old male is presented. The patient was referred to our emergency department because of sudden onset of paralysis of his both upper and lower limbs. After comprehensive evaluation, significantly lower potassium content was determined. The patient's symptoms resolved after potassium replacement and he was discharged without deficit.

The right differential diagnosis should rule out other causes of weakness and paralysis and allow timely treatment.

**Keywords:** Hypokalemic periodic paralysis • Autosomal dominant • Channelopathy • Case report • Anderson Tawil syndrome

**Abbreviations:** PP: Periodic Paralysis, HPP: Hypokalemic Periodic Paralysis, FPP: Familial Periodic Paralysis, TSH: Thyroid Stimulating Hormone, $T_3$: Triiodothyronine, $T_4$: Thyroxine, TPP: Thyrotoxic Periodic Paralysis, ATS: Anderson Tawil syndrome

## Introduction

Periodic Paralysis (PP) is a rare group of neuromuscular disorders in skeletal muscles. It occurs in different forms hereditary by channelopathy. Its most characteristic is episodic flaccid paralysis attacks. It is autosomal dominant but penetrance can be variable, men struggle more severely than women. PP according to concomitant serum potassium level is classified into two main groups (Hyperkalemic, Hypokalemic) and sub-group (Normokalemic) [1].

Hypokalemic Periodic Paralysis (HPP) is an autosomal dominant channelopathy associated with low level of serum potassium due to point mutations in sodium or calcium channels. The beginning of this form is the first or second decade of life. HPP is characterized by episodic flaccid paralysis attacks which most commonly occur at the night or early in the morning associated with low serum potassium levels [2].

Attacks are distinguished by recurrent transitory episodes of muscle weakness that vary from mild to severe complete flaccid paralysis which is potentially fatal due to the involvement of the respiratory muscles, and life threatening cardiac arrhythmias [3]. The proximal muscles are involved more severely than distal muscles which start from lower limbs and revolve to girdle muscles and progress the upper limbs. However, sensory functions are normal [4]. The episode progress over minutes to hours and last for several minutes, or persist for several days. Although some patients experience one episode in their lifetime, some experience multiple episodes in a day, a week, a month or less often [5]. The most trigger factors in HPP are high intake of carbohydrates and salt, over eating, alcohol, dehydration, sudden change in temperature, hard physical activity, and rest after exercise. Additional triggers are cold, stress, excitement, fear, prolonged immobility use of glucosteroids, and anesthetic procedure [6].

Although rare, periodic paralysis must differentiate from other causes of weakness and paralysis so that the proper treatment can be initiated quickly.

Therefore, this case report is presented with the aim of diagnosing hypokalemic periodic paralysis and timely treatment of this disease.

## Case Presentation

A 15-year-old boy with no significant past medical history present to the emergency department with sudden onset of weakness of both

***Address for Correspondence:** Mirwais Ramozi, Department of Medical Research, Kateb University, Kabul, Afghanistan, Tel: 93730221656; E-mail: mramozi2019@gmail.com*



**Received:** 17 April, 2024, Manuscript No. JCCR-24-96244; **Editor assigned:** 20 April, 2024, PreQC No. JCCR-24-96244 (PQ); **Reviewed:** 03 May, 2024, QC No. JCCR-24-96244; **Revised:** 09 May, 2024, Manuscript No. JCCR-24-96244 (R); **Published:** 17 May, 2024, DOI: 10.37421/2165-7920.2024.14.1602

upper and lower limbs. The patient had gone to bed at night after a football play and full dinner with no weakness and pain, awoke at the midnight unable to move his upper and lower extremities. The weakness was bilateral and consist both the proximal muscles of the shoulders and hips as well as distal limbs. He had no respiratory and swallowing difficulty and was able to move his neck and facial muscles. The patient denied any pain and paresthesia. Prior to this episode, he had been healthy and denied any recent diarrhea, shortness of the breath, chest pain, or weight change. He did not take any medications and denied use of alcohol or drugs. There was no history of similar episodes and no significant illnesses with none of his family members.

On physical examination, consciousness was preserved and orientated with heart rate 76 beats/ min, blood pressure 110/70 mm of Hg, respiratory rate 16, no jugular venous distension, goiter and lymphadenopathy. Cardiac exam revealed no abnormalities. Examination of the lungs and abdomen were unremarkable. There were no deformities or edema of limbs and distal pulses were present and equal bilaterally. Neurologic examination revealed flaccid paralysis of all extremities which involved the proximal and distal muscles and included the hips and shoulders with diminished reflexes but all sensory and cranial nerve functions were intact. Routine biochemistry investigation revealed normal liver enzymes, complete blood count, blood sugar and creatinine except for potassium level 2.6 (3.5-5 mmol/L) with urine spot potassium 11 mmol/L (20 mmol/L). Other serum electrolytes found to be normal. ECG revealed ST-segment depression and T-wave inversions, while the PR interval was prolonged along with an increase in the amplitude of the P wave. Abdominal sonography findings were normal, CT scan of abdomen and CT scan of brain showed normal findings.

After initiation of intravenous potassium replacement, the patient's neurologic symptoms resolved completely. Follow up studies were accomplished to determine the etiology of the patient's hypokalemia. Thyroid Stimulation Hormone (TSH); Free Triiodothyronine (FT3) and Free Thyroxine (FT4) levels found normal. 24 hours' urinary sodium and potassium, serum aldosterone and renin levels were also normal. The patient was diagnosed as hypokalemic periodic paralysis.

## Results and Discussion

Muscle weakness is a common, albeit nonspecific, symptom in both emergency and outpatient settings. Although the differential diagnosis with complaints of weakness is extensive (Table 1), the focus is greatly narrowed when the patient's muscle weakness is detected on physical examination. Strokes and tumors that cause nerve compression are life-threatening. It is possible and should be ruled out first. Other relatively common neurological problems include postictal paralysis or various motor neuron disorders. Diagnosis of these disorders requires a complete medical history, with particular attention to the timing, duration, and distribution of symptoms. Periodic paralysis is often overlooked on the first workup.

| Neurologic | Inflammatory | Infectious |
|---|---|---|
| Stroke | Polymyositis | Polio |
| Post-seizure paralysis | Dermatomyositis | Diphtheria |
| Myasthenia cataplexy | | Botulism |
| Multiple sclerosis | | |

**Table 1.** Causes of acute weakness.

There are different types of periodic paralysis associated with metabolic and electrolyte disorders. Of these, Hypokalemic Periodic Paralysis (HPP) is the most common, with a prevalence of 1 in 100,000 live births [7,8].

The clinical features of the syndrome vary somewhat depending on the underlying etiology, but the most obvious feature is the sudden onset of weakness, which ranges in severity from mild, transient weakness to severe disability, leading to life threatening respiratory failure. Attacks can be triggered by viral illnesses, stress such as fatigue, or certain medications such as beta agonists, insulin, or steroids.

Disruption of sodium and calcium ion channels leads to low potassium levels and muscle dysfunction [9].

Essentially there is a problem with muscle contraction rather than nerve conduction, the tendon reflexes may be reduced or absent but sensation is usually intact. Though the serum potassium level is often alarmingly low, other electrolytes are generally normal. As total body potassium is actually normal and change in serum levels reflect potassium shifts into cells [10].

ECG changes are common, but unlike patients with true potassium deficiency (Table 2), changes do not correlate well with measured serum levels [11].

Diagnosis during paralytic episodes is difficult because patients may have normal muscle strength and normal potassium levels. Electromyograms are abnormal in some patients but are often normal, especially between episodes in which there is no clinically evident weakness.

| Potassium depletion-Renal | Potassium depletion-Extra renal | Potassium shift into cells |
|---|---|---|
| Increased aldosterone | Decreased intake | Increased insulin |
| Diuretics | Vomiting/diarrhea | Alkalosis |

| | | |
|---|---|---|
| Hypomagnesemia | Zolinger-Ellison syndrome | Thyrotoxic periodic paralysis |
| Renal tubular acidosis (Type I and II) | Fistulas | Familial hypokalemic periodic paralysis |
| Metabolic alkalosis | | |
| Lidddle's syndrome | | |

**Table 2.** Causes of hypokalemia.

HPP occurs in several settings with different underlying etiologies, the diagnosis may need extensive investigations since the treatment varies according to the cause. HPP may occur occasionally in the form of Familial Hypokalemic Paralysis (FHP); it is a poorly understood disorder that may occur spontaneously or as a result of autosomal dominant inheritance [8]. This form of periodic paralysis is felt to be the result of impaired cellular potassium regulation, possibly due to sodium or calcium channel abnormalities [9-12]. Mutations in the CACNA1S and SCN4A genes that cause sodium channel defects leading to abnormal potassium ion flux have been identified. As a result, the muscle cannot contract efficiently. The condition is hypokalemic because a low extracellular potassium ion concentration causes the muscle to repolarize to the resting potential more quickly [13]. The first attack usually occurs between ages 5 and 35 years, but the frequency of attacks is highest between ages 15 and 35 years as our case is 15 years, and subsequently decreases with age [14]. In hypokalemic familial periodic paralysis, the most trigger factors are high intake of carbohydrates and salt, over-eating, alcohol, dehydration, surgery, pregnancy, sudden change in temperature, strenuous exercise or hard physical activity, and rest after exercise. Similar episode was observed in our case as the patient had gone to bed after a football play and full dinner. A simple exercise challenge, which is relatively safe, is somewhat helpful when serum potassium is high or low. In particular, ECG, TSH, free $T_3$ and free $T_4$ are indicated laboratory investigations, with renal and adrenal function also recommended which rule out secondary periodic paralysis, Thyrotoxic Periodic Paralysis (TPP), and other forms of PP, All the above examinations were performed in our case [15]. In Acute management of HPP treatment options include oral or Intravenous (IV) potassium administration. For prophylaxis the patient should be advised to avoid triggers such as high-carbohydrate and/or high-salt meals, alcohol, strenuous exercise and stress. Dichlorphenamide and acetazolamide is approved for HPP [16]. Which has also been useful for our patient?

Thyrotoxic Periodic Paralysis (TPP) occurs by hyperthyroidism condition. it is the most common form of HPP, seen mainly in Asian men, occurring in 1.9% of Japanese hyperthyroid patients overall and up to 8% of Japanese hyperthyroid men [17,18]. Clinical features are similar to other forms of HPP but include symptoms of thyrotoxicosis such as weight loss, tachycardia, and anxiety. However, in patients who develop HPP, symptoms of hyperthyroidism are often quite mild and can be overlooked.

Rarely, HPP can result from significant gastrointestinal or renal potassium losses. In these cases, the body's total potassium is depleted and requires aggressive replacement. Endocrine abnormalities such as hyperinsulinemia and primary hyperaldosteronism are associated with HPP [19]. Surgical removal of the aldosterone producing tumor is preferred, although symptoms are often manageable with spironolactone.

Hyperkalemic periodic paralysis and paramitonia congenita are rare forms of periodic paralysis associated with SCN4A mutations that cause gain of function abnormalities in the sodium channel that lead to prolonged excitability of muscle cells [20]. As a result, these conditions are exacerbated by repeated activity and, in some cases, exposure to cold. Patients often have facial paralysis, which affects the lower extremities to a lesser extent. Most patients do not require treatment, but are instructed to avoid debilitating situations. Mexiletine, which reduces the sensitivity of muscle tissue to nerve impulses, may have several benefits.

Anderson Tawil Syndrome (ATS) is a rare multisystem, autosomal dominant disorder which is characterized by three main aspects: Periodic paralysis, ventricular arrhythmia, and skeletal developmental anomalies. It is caused by mutation of the KCNJ2 gene in 60% of cases. A diagnosis of ATS can be made when a person has two of these three main characteristics. Sixty percent of affected individuals exhibit the full triad, while 80 percent express two of the three main features.

Mutations in this gene alter the structure and function of potassium channels, inhibiting the flow of potassium ions to muscle cells and causing periodic paralysis and long QT syndrome. Acetazolamide can prevent paralytic episodes, and antiarrhythmic or beta-blockers can prevent ventricular ectopic, but limited data are available.

## Conclusion

Hypokalemic familial periodic paralysis is an autosomal dominant disorder which runs through families with occasional episodes of muscle weakness. It is a disorder caused by impaired cellular potassium regulation, due to mutation in sodium or calcium ion channels. The episodes of attacks begin during first or second decade of life with highest frequency of attacks between ages 15 and 35 and decreases with age. Differential diagnosis of HPP with other forms of PP and secondary HPP is made by history, clinical and intensive laboratory manifestations.

The patient presented with sudden onset of limb paralysis and markedly low level of potassium. By conducting physical, laboratory examinations and history, the patient was diagnosed with hypokalemic familial periodic paralysis. Treatment options include oral and intravenous potassium supplementation and dichlorphenamide/acetazolamide for prophylaxis. The paralysis resolved completely following potassium replacement and he began a course of acetazolamide prior to being discharged from the hospital. At the time of discharge, he had no neurologic findings and has not suffered any further episodes of paralysis.

## Conflicts of Interest

The authors declare that there are no competing interests.

## Author's Contributions

Conception and designing of the study; Mirwais Ramozi, Sayed Mohammad Reza Hosseini writing this paper; Mirwais Ramozi, Sayed Mohammad Reza Hosseini.

Critical revision of the paper; Abdullah Rastin, Abass Ali Ramozi All the authors read the final draft and approved submission.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

## References


1. Fontaine, Bertrand. "Periodic paralysis." *Adv Genet* 63 (2008): 3-23.
2. Matthews E, R Labrum, MG Sweeney, and R Sud, et al. "Voltage sensor charge loss accounts for most cases of hypokalemic periodic paralysis." *Neurology* 72 (2009): 1544-1547.
3. Alkaabi, Juma M, Ahmed Mushtaq, Fatma N Al-Maskari, and Nagi A Moussa, et al. "Hypokalemic periodic paralysis: A case series, review of the literature and update of management." *Eur J Emerg Med* 17 (2010): 45-47.
4. Kung, Annie WC. "Thyrotoxic periodic paralysis: A diagnostic challenge." *J Clin Endocrinol Metab* 91 (2006): 2490-2495.
5. Welland, Natasha Lervaag, Helge Haestad, Hanne Ludt Fossmo, and Kaja Giltvedt, et al. "The role of nutrition and physical activity as trigger factors of paralytic attacks in primary periodic paralysis." *J Neuromuscul Dis* 8 (2021): 457-468.
6. Soule, Benjamin R, and Nicole L Simone. "Hypokalemic Periodic Paralysis: A case report and review of the literature." *Cases J* 1 (2008): 1-5.
7. Fontaine B, J Vale-Santos, K Jurkat-Rott, and J Reboul, et al. "Mapping of the hypokalaemic periodic paralysis (HypoPP) locus to chromosome 1q31–32 in three European families." *Nat Genet* 6 (1994): 267-272.
8. Jurkat-Rott, Karin, Holger Lerche, Yvonne Weber, and Frank Lehmann-Horn, et al. "Hereditary channelopathies in neurology." *Adv Exp Med Biol* 686 (2010): 305-334.
9. Lin, Shih-Hua, Yuh-Feng Lin, Dung-Tsa Chen, and Pauling Chu, et al. "Laboratory tests to determine the cause of hypokalemia and paralysis." *Arch Intern Med* 164 (2004): 1561-1566.
10. Kelley, David E, Hossein Gharib, Frank P Kennedy, and Ralph J Duda, et al. "Thyrotoxic periodic paralysis: Report of 10 cases and review of electromyographic findings." *Arch Intern Med* 149 (1989): 2597-2600.
11. Wang, Weiqing, Lei Jiang, Lei Ye, and Na Zhu, et al. "Mutation screening in Chinese hypokalemic periodic paralysis patients." *Mol Genet Metab* 87 (2006): 359-363.
12. Cannon, Stephen C. "Channelopathies of skeletal muscle excitability." *Compr Physiol* 5 (2015): 761.
13. Baig, Maarij, Prateek Juneja, Nicole Baig, and Kruti Ray, et al. "a rare case of a hypokalemic periodic paralysis while undergoing treatment for addison's disease." *Chest* 160 (2021): A964.
14. Statland, Jeffrey M, Bertrand Fontaine, Michael G Hanna, and Nicholas E Johnson, et al. "Review of the diagnosis and treatment of periodic paralysis." *Muscle Nerve* 57 (2018): 522-530.
15. Okinaka, Shigeo, Kazuo Shizume, Shiro Uno, and Akira Watanabe, et al. "The association of periodic paralysis and hyperthyroidism in Japan." *J Clin Endocrinol Metab* 17 (1957): 1454-1459.
16. Shizume, Kazuo, Yoshimasa Shishiba, Kanji Kuma, and Shiro Noguchi, et al. "Comparison of the incidence of association of periodic paralysis and hyperthyroidism in Japan in 1957 and 1991." *Endocrinol Jpn* 39 (1992):315-318.
17. Ogawa, Touru, and Keita Kamikubo. "Hypokalemic periodic paralysis associated with hypophosphatemia in a patient with hyperinsulinemia." *Am J Med Sci* 318 (1999): 69-72.
18. Huang, Shan, Wei Zhang, Xueli Chang, and Junhong Guo, et al. "Overlap of periodic paralysis and paramyotonia congenita caused by SCN4A gene mutations two family reports and literature review." *Channels* 13 (2019): 110-119.
19. Daroff, Robert B, and Michael J Aminoff. Encyclopedia of the neurological sciences. Oxford: Academic Press; p. 743-9, (2014).
20. Issa, Ziad F, John Michael Miller, and Douglas P Zipes. Clinical arrhythmology and electrophysiology: A companion to Braunwald's heart disease. Philadelphia: Elsevier; p. 976-1041, (2019).


**How to cite this article:** Ramozi, Mirwais, Sayed Mohammad Reza Hosseini, Abdullah Rastin, and Abass Ali Ramozi. "Hypokalemic Periodic Paralysis in Afghanistan, Study of Case Report." *J Clin Case Rep* 14 (2024): 1602.