

Caring for a patient with hypokalemic periodic paralysis: A case report

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Abstract

Hypokalemic Periodic Paralysis (HypoPP) is a condition that causes episodes of extreme muscle weakness typically beginning in childhood or adolescence. The disorder is three to four times more commonly clinically expressed in men. Mutations in the CACNA1S (calcium voltage-gated channel subunit alpha 1 S) or SCN4A (sodium voltage-gated channel alpha subunit 4) gene can cause Hypokalemic Periodic Paralysis. The primary form is genetic and follows an autosomal dominant pattern of inheritance. The secondary forms may complicate a number of medical disorders. Echocardiography, electromyography, muscle biopsy, lab investigations are useful for diagnosis. The treatment of choice is avoidance of triggering factors, adherence to a diet low in sodium, carbohydrate and rich in potassium, and with the use of oral potassium supplementation. Nurses have a great role to play in the management of a patient with HypoPP. For a comprehensive understanding of the condition 'Hypokalemic Periodic Paralysis' here is an article with the details of the disease - definition, other names, epidemiology, etiology, pathophysiology, clinical manifestations, diagnostic measures, management, complications and prevention presented along with a case report focusing on nursing management.

Keywords: HypoPP - hypokalemic periodic paralysis, CACNA1S - calcium voltage gated channel subunit alpha 1 S, SCN4A - sodium voltage gated channel alpha subunit 4, ROM - range of motion

Introduction

Hypokalemic Periodic Paralysis is a condition that causes episodes of extreme muscle weakness typically beginning in childhood or adolescence. Most often, these episodes involve a temporary inability to move muscles in the arms and legs. Attacks cause severe weakness or paralysis that usually lasts from hours to days. Some people may have episodes almost every day, while others experience them weekly, monthly, or only rarely. Attacks can occur without warning or can be triggered by factors such as rest after exercise, a viral illness, or certain medications. Often, a large carbohydrate rich meal or vigorous exercise in the evening can trigger an attack upon waking the following morning. Although affected individuals usually regain their muscle strength between attacks, repeated episodes can lead to persistent muscle weakness later in life. People with Hypokalemic Periodic Paralysis have reduced levels of potassium in their blood during episodes of muscle weakness. Researchers are investigating how low potassium levels may be related to the muscle abnormalities in this condition^[1].

Definition

Hypokalemic Periodic Paralysis is a rare genetic disorder with autosomal dominant inheritance and characterized by recurrent attacks of skeletal muscle weakness with

associated hypokalemia which is precipitated by stress, cold, carbohydrate load, infection, glucose infusion, hypothermia, metabolic alkalosis, anesthesia, and steroids^[2].

Other Names

- Familial Hypokalemic Periodic Paralysis
- HOKPP
- HypoKPP
- HypoPP
- Primary Hypokalemic Periodic Paralysis
- Westphall disease¹

Epidemiology

Hypokalemic Periodic Paralysis is the most common of the periodic paralysis, but is still quite rare, with an estimated prevalence of 1 in 100,000. Hypokalemic Periodic Paralysis may be familial with autosomal dominant inheritance or may be acquired in patients with thyrotoxicosis. Men tend to experience symptoms of this condition more often than women. The disorder is three to four times more commonly clinically expressed in men. Approximately one third of cases represent new mutations^[3].

Etiology and Pathophysiology

Mutations in the CACNA1S (calcium voltage gated channel subunit alpha 1 S) or SCN4A (sodium voltage gated channel

alpha subunit 4) gene can cause Hypokalemic Periodic Paralysis. These genes provide instructions for making proteins that play an essential role in skeletal muscles. For the body to move normally, these muscles must contract and relax in a coordinated way. Muscle contractions are triggered by the flow of certain positively charged ions into muscle cells. The CACNA1S and SCN4A proteins form channels that control the flow of these ions. The channel formed by the CACNA1S protein transports calcium ions into cells, while the channel formed by the SCN4A protein transports sodium ions. Mutations in the CACNA1S or SCN4A gene alter the usual structure and function of calcium or sodium channels. The altered channels cannot properly regulate the flow of ions into muscle cells, which reduces the ability of skeletal muscles to contract. Because muscle contraction is needed for movement, a disruption in normal ion transport leads to episodes of severe muscle weakness or paralysis. A small percentage of people with the characteristic features of Hypokalemic Periodic Paralysis do not have identified mutations in the CACNA1S or SCN4A gene. In these cases, the cause of the condition is unknown [4].

Types

There are two forms: Primary and Secondary

The primary form is genetic and follows an autosomal dominant pattern of inheritance. The primary form usually manifests in the first 2 decades of life and is more common in men than women.

The secondary forms may complicate a number of medical disorders including, but not limited to, primary and acquired distal renal tubular acidosis, primary and acquired Fanconi syndrome, Thyrotoxicosis, Hyperaldosteronism, Bartter syndrome, Gitelman syndrome, and medication reactions (theophylline, amphotericin, diuretics, laxatives, chemotherapy agents such as cisplatin). Late onset Hypokalemic Periodic Paralysis is usually secondary and is characterized by hypokalemia, which may be persistent in contrast to primary where hypokalemia is periodic [3].

Clinical Manifestations

Symptoms include attacks of muscle weakness or paralysis that come and go. There is normal muscle strength between attacks. Attacks usually begin in the teen years, but they can occur before age 10. How often the attacks occur varies. Some people have attacks every day. Others have them once a year. During attacks the person remains alert. The weakness or paralysis most commonly occurs at the shoulders and hips and also affect the arms, legs, muscles of the eyes, and muscles that help with breathing and swallowing. The weakness or paralysis occurs on and off, most commonly occurs on awakening or after sleep or rest. It is rare during exercise, but may be triggered by resting after exercise and also by high carbohydrate, high salt meals, stress, pregnancy, heavy exercise, and cold. An attack usually lasts for several hours up to a day. Another symptom may include eyelid myotonia [3].

Diag Nostic Measures

History collection

HypoPP suspicion is based on a family history of the disorder. Other clues to the disorder are muscle weakness

symptoms that come and go with normal or low results of a potassium test.

Physical examination

- Between attacks, a physical examination shows nothing abnormal.
- Before an attack, there may be leg stiffness or heaviness in the legs.
- During an attack, muscle reflexes are decreased or absent.
- Muscles go limp rather than staying stiff.
- Muscle groups near the body, such as the shoulders and hips, are involved more often than the arms and legs.

Laboratory findings

During an attack of muscle weakness, blood potassium level is low. This confirms the diagnosis. There is no decrease in total body potassium. Blood potassium level is normal between attacks.

Electrocardiogram (ECG): may be abnormal during attacks.

Electromyography (EMG): usually normal between attacks and abnormal during attacks [4].

Muscle biopsy: often shows the presence of single or multiple centrally placed vacuoles.

Provocative test: patients whose attacks are too infrequent, for study of a spontaneous attack to be feasible require provocative testing with glucose and insulin administration. These tests are potentially hazardous and require careful monitoring [5].

Treatment

The goals of treatment are relief of symptoms and prevention of further attacks.

Treatment varies depending on the intensity and duration of the paralytic attack. Minor attacks may resolve spontaneously. Moderate attacks may be self-treated in a nonmedical setting by ingestion of oral potassium salts.⁶ Oral potassium chloride (0.2 to 0.4 mmol/kg) should be given to patients with severe weakness and repeated at 15 to 30 minutes intervals depending on the response of ECG, serum potassium level and muscle strength. When patients are unable to swallow or are vomiting, intravenous therapy may be necessary. Small repeated boluses of potassium chloride (0.1mmol/kg) may be administered over 5 to 10 minutes with careful monitoring of the ECG and serum potassium level. If potassium is administered as a dilute solution (20 to 40 mmol/L) in 5% glucose or in physiologic saline solution, the serum potassium may decline, and weakness may worsen. Mannitol is preferred vehicle for administered intravenous potassium in such situations, since it facilitates rapid return of the serum potassium level to normal but does not cause the lowering of the serum potassium level that may be caused by glucose or saline solutions [5]. There is no known curative treatment for hypoPP related myopathy; physiotherapy may help to maintain strength and motor skills [6].

Prevention of primary manifestations

The goal of preventive treatment is to reduce the frequency and intensity of paralytic attacks. This may be achieved by:

- Avoidance of triggering factors
- Adherence to a diet low in sodium and carbohydrate, rich in potassium, and with the use of oral potassium supplementation.
- If dietary intervention and oral potassium supplementation are not effective in preventing attacks, treatment with a carbonic anhydrase inhibitor (acetazolamide or dichlorphenamide) may be necessary.
- Acetazolamide (125 to 1000 mg/d in divided doses) or dichlorphenamide (50 to 200 mg/d) abolishes attacks in most cases ^[5].
- If carbonic anhydrase inhibitors are not tolerated or not effective after prolonged use, alternatives include potassium sparing diuretics such as triamterene (25 to 100 mg/d), spironolactone (25 to 100 mg/d), or eplerenone ^[5, 6].

Prevention of secondary complications

Creating a safe environment, getting help in case of paralytic attack, and preventing falls and accidents are critical. An affected person experiencing a paralytic attack must have access to potassium as well as physical assistance and companions must be informed of the risk in order to enable rapid treatment. Anaesthetic complications should be prevented by strict control of serum potassium concentration, avoidance of large glucose and salt load, maintenance of body temperature and acid-base balance, and careful use of neuromuscular blocking agents with continuous monitoring of neuromuscular function. It is unknown whether prevention of paralytic attacks also prevents the development of myopathy. Individuals with known pathogenic variants in one of the genes associated with hypoPP who developed myopathy without having experienced episodes of weakness have been reported.

Surveillance

The frequency of consultations is adapted to the individuals signs/symptoms and response to preventive treatment. Periodic neurologic examination with attention to muscle strength in the legs should be performed to detect long lasting weakness associated with myopathy. For those taking acetazolamide, the following are indicated every three months: complete blood count, electrolytes, glucose, uric acid, and liver enzyme levels. Renal ultrasound should be performed annually.

Agents/circumstances to avoid

1. Factors that trigger paralytic attacks (e.g., unusually strenuous effort, carbohydrate rich meals or sweets, cold, stress/excitement/fear, high salt intake, prolonged immobility, oral or intravenous glucosteroids, certain anaesthetic procedures, alcohol) should be avoided when possible ^[6].

Complications

- Kidney stones (side effect of acetazolamide)
- Irregular heartbeat during attacks
- Difficulty in breathing, speaking, or swallowing during attacks (rare)

- Muscle weakness that worsens over time ^[4]

Prevention

HypoPP cannot be prevented. Because, it can be inherited. Genetic counselling may be advised for couples at risk of the disorder. Treatment prevents attacks of weakness. Before an attack, there may be leg stiffness or heaviness in the legs. Doing mild exercise when these symptoms start may help prevent a full blown attack ^[7].

The following case report highlights the symptoms, treatment and preventive measures for Hypokalemic Periodic Paralysis.

Case Report

Mrs. X, a 55 year old female was brought to the hospital (Emergency Room) with recurrent seizures and unresponsiveness. She also had a history of vomiting, decreased sleep, muscle weakness, drooping of eyes, headache and pain all over the body since two weeks. She was taken to a local hospital at her home town and received analgesics and antiemetics. On arrival to emergency room, Temperature: 100°F, Heart rate: 100 beats per minute, Respiration: 20 breaths per minute and Blood pressure: 180/100 mm of Hg. Patient was connected to four litres of oxygen via face mask. Computed Tomography of the brain (plain) was done and was found to be normal. Blood investigations was done and it showed a marked decrease in serum potassium (2.9 mEq/l) level. She was diagnosed as Hypokalemic Periodic Paralysis. Condition of the patient was explained to the relatives and she was admitted to Medical Intensive Care Unit. Potassium correction was done (40 mEq). Within one hour she regained her consciousness. She is a known case of Bipolar Affective Disorder and was on regular treatment. She also exhibited the following symptoms like restlessness, agitation, disorientation, anxious mood, increased talking, crying. She was on restraints. Her past history revealed that she suffers from rheumatoid arthritis and not much information was available about its treatment. She was treated with potassium supplements, antiepileptics, proton pump inhibitors, antibiotics, steroids and analgesics. She was hemodynamically stable. After two days, she was shifted to medical ward. She got discharged in a healthy state after three days and was advised to come for follow up visits.

Nursing Care

Nursing care of the patient involves assessment of patient, identification of symptoms and complications, provision of appropriate therapeutic interventions and health education.

Nursing Diagnosis: Impaired physical mobility related to skeletal muscle weakness as evidenced by muscle weakness and body ache.

Expected outcome: Client maintains normal physical mobility as evidenced by absence of muscle weakness, body ache.

Interventions

- Continually assessed the motor function by requesting client to perform certain actions.
- Assisted with full Range of motion (ROM) exercises on

all extremities and joints, using slow smooth movements.

- Encouraged the use of relaxation techniques like listening to music.
- Potassium correction was done (40mEq). Her serum potassium level came to normal level (4mEq/l).
- Inspected skin daily. There was no evidence of skin peel.
- Observed pressure areas and provided meticulous skin care every two hourly.

Evaluation: Patient's muscle weakness and body ache reduced with ROM exercises. ROM exercises and physiotherapy were given daily and muscle strength improved.

Nursing Diagnosis: Risk for electrolyte imbalance related to vomiting as evidenced by serum potassium level 2.9 mEq/l.

Expected outcome: Client maintains normal electrolyte level as evidenced by normal potassium level (4mEq/l) and absence of vomiting.

Interventions

- Assessed the level of consciousness and neuromuscular function.
- Monitored heart rate and rhythm. There was no variations in the ECG.
- Monitored respiratory rate, depth, effort. Rate was found to be 24 bpm.
- Auscultated the bowel sounds. Bowel sounds were present, 7/minute.
- Maintained accurate record of intake and output.
- Monitored the rate of IV potassium administration using infusion pump.
- Encouraged intake of foods and fluids high in potassium like bananas, oranges, dried fruits, red meat, salmon, turkey, leafy vegetables, peas, baked potatoes, tomatoes, coffee, tea.
- Diluted liquid and effervescent potassium supplements with four ounce of water and gave it after the meals.

Evaluation: Patient's vomiting reduced and hypokalemia corrected. Oral as well as IV potassium, proton pump inhibitors, foods rich in potassium like tender coconut water and bananas were continued.

Nursing Diagnosis: Activity intolerance related to body ache, muscle weakness, imposed activity restriction.

Expected outcome: Client maintains normal activity within her limit as evidenced by absence of muscle weakness, body ache and free from restraints.

Interventions

- Assessed the physical activity level. She had muscle weakness and body ache. She also was on restraints.
- Determined the patient's daily routine.
- Assisted the patient with ROM exercises on all extremities and joints using slow smooth movements.
- Provided emotional support.

- Advised patient to perform the activity more slowly in a longer time with more rest or pauses or with assistance necessary, when she was free from restraints.

Evaluation: Patient's tolerance in activity increased with exercises and assistance. She was able to resume normal activity.

Nursing Diagnosis: Anxiety related to prognosis and hospitalization.

Expected outcome: Client gets relief from anxiety as evidenced by verbalization and pleasant facial expression.

Interventions

- Assessed the anxiety level of the patient. She was in anxious mood and crying.
- Supported the patient in adhering to treatment and gave positive feedback.
- Maintained good interpersonal relationship.
- Provided diversional therapy like paper crafts when she free from restraints.
- Encouraged to express her feelings.

Evaluation: Patient's anxiety level reduced. She verbalized her feelings.

Conclusion

HypoPP responds well to treatment. Treatment may prevent, and even reverse, progressive muscle weakness. Although muscle strength starts out normal between attacks, repeated attacks may eventually cause worsening and permanent muscle weakness between attacks. Prompt therapy and nursing care goes hand in hand to achieve best outcome. Nursing care rendered need to be comprehensive and complete. The nurse needs to ensure that adequate education is provided to caregivers for ongoing care even at home.

Declaration of interest

There were no conflicts of interest reported.

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