International Journal of Contemporary Pediatrics
Ravikumar SG et al. Int J Contemp Pediatr. 2026 Sep;13(9):1858-1861
https://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20262982

Case Report

Primary hypokalemic periodic paralysis with sinus bradycardia: a case report and review of literature

Sugapradha G. Ravikumar*, Premkumar B.

Department of Pediatrics, Trichy SRM Medical College Hospital and Research Centre, The TN Dr. MGR Medical University, India

Received: 28 June 2026
Accepted: 04 August 2026

***Correspondence:**
Dr. Sugapradha G. Ravikumar,
E-mail: grsugapradha@gmail.com

ABSTRACT

Hypokalemic periodic paralysis (HPP) is a rare autosomal dominant disorder caused by mutations affecting skeletal muscle ion channels, most commonly CACNA1S or SCN4A. It manifests as recurrent episodes of flaccid paralysis associated with low serum potassium levels, often precipitated by rest after exertion, high-carbohydrate intake, or intercurrent illness. Although the condition typically presents during adolescence, diagnosis is often delayed due to its episodic nature and overlap with other causes of acute weakness. The estimated incidence is 1:100,000. HPP with cardiac arrhythmias have been reported but sinus bradycardia is not reported so far. Hence, we present a rare case of primary HPP type 2 with sinus bradycardia and a pathogenic genetic variant. A 16-year-old male child born to third-degree consanguineous parents, presented to the emergency department with an acute episode of fever, vomiting, headache, generalized weakness with difficulty in getting up or standing since waking up that morning. The weakness initially involved the lower limbs and then progressed to the upper limbs within few hours. Electrocardiogram showed sinus bradycardia with prominent U waves. Serum potassium was 1.2 mmol/l with sodium, magnesium and calcium within normal limits. The paralysis and ECG abnormality became normal following replacement of potassium. There was a similar history in father and paternal uncle. Genetic study detected a heterozygous missense variant in exon 12 of the SCN4A gene (chr17: g.63959279G>C; Depth: 85x) that results in the amino acid substitution of glycine for arginine at codon 669 (p.Arg669Gly; ENST00000435607.3) causative of HPP, type 2. HPP is a rare neuromuscular disease that can present with cardiac arrhythmias. We present a case of primary HPP type 2 with sinus bradycardia which resolved spontaneously with potassium supplementation.

Keywords: Hypokalemia, Periodic paralysis, Bradycardia, Genetic variant

INTRODUCTION

Hypokalemic periodic paralysis (HPP) is a genetic neuromuscular disorder characterized by episodes of focal or generalized skeletal muscle paralysis associated with hypokalemia.[1] Hypokalemic periodic paralysis type 2 is caused by heterozygous mutations in the SCN4A gene. Attacks typically present during the second decade of life, spontaneously or, most commonly, triggered by carbohydrate-rich meals, alcohol, or rest after strenuous exercise.[2] Attacks last for 4-24 hours, may present during/after sleep, attacks relieved by potassium administration, attacks usually decrease or disappear after age 40 years, progressive interictal weakness is common, myotonia is usually not seen, vacuolar myopathy may occur, tubular aggregates in muscle fibers may occur. Herein we describe a 16-year-old male patient with primary hypokalemic periodic paralysis type 2 and sinus bradycardia followed by a systematic literature review.

CASE REPORT

A 16-year-old male child born to third-degree consanguineous parents, presented to the emergency

department with an acute episode of generalized weakness with difficulty in getting up or standing since waking up that morning. The weakness initially involved the lower limbs and then progressed to the upper limbs within few hours. This was not associated with any loss of consciousness or sensory loss or any cranial nerve involvement. The child had febrile episodes for past 3 days before the episode which resolved on medications and was associated with an episode of vomiting. A family history of similar episodes of sudden onset of weakness involving all 4 limbs which resolves on its own in a single day was present in the child's father and paternal uncle. Detailed history revealed an intake of carbohydrate rich diet the previous day.

On clinical examination the child was conscious, oriented and afebrile with a heart rate of 40/min. respiratory rate, blood pressure and saturation were stable. Neurological examination showed a proximal as well as a distal muscle weakness (MRC power scale 1/5) and areflexia of all group of muscles. However, the respiratory and cardiovascular examination was unremarkable except for the bradycardia with no other neurological findings.

Electrocardiogram showed sinus bradycardia with prominent U waves (Figure 1). Serum potassium was 1.2 mmol/l with sodium, magnesium and calcium within normal limits.

MRI spine done was normal. Thyroid function tests were normal.

A probable diagnosis of hypokalemic periodic paralysis was made and the patient was treated with IV potassium and oral supplementation of potassium, resulting in complete resolution of symptoms and signs within 12 hours. As family history suggested a possibility of autosomal dominant inherited trait, genetic study had been done.

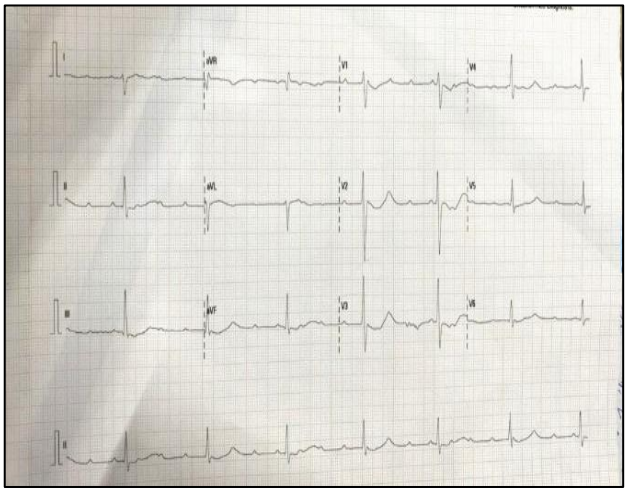

Figure 1: ECG showing sinus bradycardia with prominent U waves.

Genetic variant interpretation

A heterozygous missense variant in exon 12 of the SCN4A gene (chr17:g.63959279G>C; Depth: 85x) that results in amino acid substitution of glycine for arginine at codon 669 (p.Arg669Gly; ENST00000435607.3) was detected (Table 1). This variant located in the "Ion transport protein" domain of SCN4A protein (PF00520).

Table 1: Genetic variant interpretation.

Gene (Transcript)	Location	Variant	Zygosity	Disease (OMIM)	Inheritance	Classification
SCN4A (-) (ENST00000435607.3) NM_000334.4	Exon 12	c.2005C>G (p. Arg669Gly)	Hetero-zygous	Hypokalemic periodic paralysis, type 2 (OMIM#613345)	Autosomal dominant	Likely Pathogenic (PP1_Sup, PM1_S, PM2_M, PP3_Sup)

*_VS - Very Strong, _S-Strong, _M-Moderate, _Sup-Supporting

Patient was being followed up regularly and there was no recurrence of symptoms.

DISCUSSION

HPP is a rare genetic neuromuscular disorder with an estimated incidence of 1:100,000 characterized by episodes of focal or generalized skeletal muscle paralysis associated with hypokalemia.[1] Attacks typically present during the second decade of life, spontaneously or, most commonly, triggered by carbohydrate-rich meals, alcohol, or rest after strenuous exercise.[1,2]

In HPP the potassium level is most often between 0.9 mmol/l and 3 mmol/l.[3] In our patient the serum potassium was 1.2 mmol/l. The ECG abnormality is a significant clinical finding in hypokalaemia. The classical signs of hypokalaemia were ST-segment depression, T-wave depression or inversion, increased U-wave amplitude, AV block, and QT prolongation.[4,5] Our patient electrocardiogram showed sinus bradycardia with prominent U waves (Figure 1).

Diagnosis is based on genetic testing showing heterozygous pathogenic mutations in the CACNA1S or SCN4A genes, present in 70-80% and in 10% of cases,

respectively.[6] Hypokalemic periodic paralysis, type 1 is characterized by a mutation in the dihydropyridine-sensitive skeletal muscle calcium channel gene, CACNA1S. Hypokalemic periodic paralysis, type 2 is associated with mutations in the voltage-sensitive skeletal muscle sodium channel gene, SCN4A.[6] In addition, disease-causing mutations in the genes KCNJ2 and KCNJ18, which code for the inward rectifier potassium (Kir) channel, have also been identified as contributors to Hypokalemic periodic paralysis and can lead to a condition known as Andersen-Tawil syndrome. In cases with negative genetic testing, HPP is diagnosed based on clinical presentation, serum potassium levels during paralytic attacks, and reduction in compound muscle action potential amplitude during or after exercise.[6,7]

In our patient genetic study revealed a heterozygous missense variant in exon 12 of the SCN4A gene (chr17: g.63959279G>C; Depth: 85x) that resulted in the amino acid substitution of glycine for arginine at codon 669 (p. Arg669Gly; ENST00000435607.3) (Table 1). This variant was located in the "Ion transport protein" domain of the SCN4A protein (PF00520).

HPP is caused by a skeletal muscle channelopathy. Normally, acetylcholine is released and binds to specific receptors at the neuromuscular junction.[6] Once a threshold is reached, voltage-gated sodium channels, predominantly Nav1.4, are activated.[3] Propagation of action potentials induces a conformational change of the dihydropyridine receptor (DHPR), leading to opening of the type 1 ryanodine receptor (RyR1) anchored in the sarcoplasmic membrane, resulting in increased intracellular calcium, which triggers muscle contraction.[6] CACNA1S, located in the chromosome 1q32.1, encodes the Cav1.1 protein, pore-forming a1-subunit of the L-type calcium channel DHPR, while SCN4A encodes the pore-forming a-subunit of the skeletal muscle voltage-gated sodium channel Nav1.4.[6] Mutations in these genes alter the conduction for monovalent cations, leading to an alternative conduction pathway known as “current leak,” which mainly manifests at resting membrane potentials.[6] Although this would be expected to cause hyperpolarization of the membrane, in the presence of hypokalemia there are decreased outward potassium currents through inward rectifier channels, with a net potassium inward current, leading to paradoxical depolarization and subsequent paralysis.[6]

Periodic paralysis hypokalaemia may occur in several clinical settings. The diagnosis should require an extensive search for the underlying aetiology. The treatment may vary according to the cause. In this patient, we have excluded the possibility of thyrotoxic periodic paralysis (TPP). TPP is related to hyperthyroidism. It is the most common form of HPP and is seen primarily in young Asian males.[7]

Treatment of periodic paralysis hypokalaemia consists of slow and careful correction of low potassium level. The dose of correction may not exceed of 10 mmol/hour. The very rapid correction exposes the risk of hyperkalaemia.[8] Close monitoring by neurological examination and repeat examination potassium level is essential.[9,10] Therapeutic education that focused on avoiding triggers is very important. It prevents the occurrence of episodes in the future.[11]

HPP is a rare genetic neuromuscular disorder, hypokalemia must be corrected slowly, genetic study is essential to identify the variants, thyroid function tests must be done and periodical follow up is needed.

CONCLUSION

HPP is a rare neuromuscular disease that can present with cardiac arrhythmias. We present a case of primary hypokalemic periodic paralysis type 2 with sinus bradycardia which resolved spontaneously with potassium supplementation.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Ravikumar SG, Premkumar B. Primary hypokalemic periodic paralysis with sinus bradycardia: a case report and review of literature. Int J Contemp Pediatr 2026;13:1858-61.