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# Hypokalaemic periodic paralysis in an African—Case report*

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**Summary**

The case history of a typical case of hypokalaemic periodic paralysis is described. A 21-year-old male Ugandan African presented with his second attack of complete flaccid tetraplegia. Serum potassium levels, ECG changes and response to therapy were entirely consistent with the diagnosis. Family history was negative. It is possible that this rare disorder has not previously been reported in an African. Possible mechanisms involved in the disorder are briefly discussed.

**Résumé**

Un africain en Ouganda, agé de 21 ans, a présenté pour la deuxième fois avec la paralysie complète de tous les membres à cause de la paralysie hypokaliémique périodique (maladie de Westphal). On l'a confirmé par les mésures du potassium dans le sang, par les changements electrocardiographiques, et par les effets de thérapie. Dans sa famille il resta seul avec cette maladie. Peut-étre reste-t-il seul africain avec cette maladie, quant'aux journaux medicaux jusqu'à ce moment. Il y a une petite analyse du mode du cours pathologique.

## Introduction

Hypokalaemic periodic paralysis, a rare genetically-determined disorder, has not been reported in an African in Uganda, and possibly not elsewhere in Africa.

The publication of a case, very typical in all respects save for the absence of a positive family history, is therefore of considerable epidemiological interest and may lead to further useful information concerning its prevalence (or absence) in other parts of the continent. Furthermore, possible errors of diagnosis and management may be avoided once attention has been drawn to the existence of such a disorder which is both easy to diagnose and highly rewarding to treat. Lastly, the nature of the pathological process in the paralytic phase of the disease is a most intriguing and still very imperfectly understood problem, the unravelling of which is likely to throw light on the dynamics of the muscle membrane and the excitation-contraction coupling process in normal as well as affected muscles.

## Case report

Mr G.K., a Muganda male aged 21 years, was admitted to Mulago Hospital on 9 June 1971 at 19.10 h with the complaint of paralysis of all four limbs since the morning of the day of admission. He had been well until that morning when, on waking, he had found that he could not move his arms and legs. There was no difficulty with breathing, speaking or swallowing and he had normal control of bladder and rectum. Sensation was normal and there were no paraesthesiae. Mental function, eyesight and hearing were unaffected. He denied any history of antecedent febrile illness, trauma, drugs or poisoning. He had not taken alcohol or an unduly heavy meal the night before, nor had he undergone strenuous exertion.

He remembered a similar episode of weakness of the legs four years previously, in September 1967, when he was 17 years old. He had been hospitalized in the same ward for 3 days. No definite diagnosis had been reached; hysteria was suspected. Otherwise, he had always been very fit. He had no symptoms suggestive of thyrotoxicosis or myotonia.

There was no family history of a similar disorder.

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On examination, his general condition was excellent. He appeared to be euthyroid. Blood pressure was 150/80 mmHg. There were no abnormal signs in the heart or chest. He was fully conscious, cooperative and mentally entirely healthy. The fundi and cranial nerves were normal. In particular there was no weakness of speaking or swallowing. There was a profound flaccid arreflexic weakness of all muscles of the limbs and trunk except for the muscles of respiration. Abdominal and plantar responses were absent. All modalities of sensation were normal.

The diagnosis of hypokalaemic periodic paralysis was suspected and confirmed by finding on the same day a serum potassium of 1·4 mEq/l and changes in the ECG typical of hypokalaemia, that is ST segment depression, flattened T waves and prominent U waves (Turner, 1967). Blood sugar, blood urea and cerebrospinal fluid were normal. The patient was given 1 g of potassium chloride three times a day. The following morning at 0830 he had a strong grip and sluggish reflexes. At 1300 he walked to the toilet; power had returned in full and reflexes were normally brisk. No myotonia was detectable. Serum potassium had risen to 2·7 mEq/l and the ECG was returning to normal. The patient was discharged fully fit on the fourth day.

The progressive return to normal of the ECG is shown in Figs 1–3. The changes are most striking in the precordial leads, the abnormal U waves being noted best in V2 and V3.

The patient has not been seen since.

## Discussion

The clinical presentation of this case does not differ in any important respect from the classical picture originally described by Shakhnovitch (1882) and by Westphal (1885) (see Pratt, 1967), and reviewed by McArdle (1963). The patient was a male, with onset in the second decade of life, experience of initial weakness on waking, profound flaccid paralysis at peak, sparing of bulbar and respiratory muscles, together with marked hypokalaemia and typical ECG changes, and complete recovery on oral potassium within 24 h. He showed no evidence of myotonia or thyrotoxicosis. The only unusual feature was the negative family history, although sporadic cases do occur, much more commonly in males. Detailed information about the historical background, clinical picture and genetic basis of the disease is given by Pratt (1967).

The uniquely interesting feature in this case lies not in his clinical presentation (which was typical) but in his being the first recorded African with the disorder. Its occurrence in Africans was not known to Trowell (1960) who had reviewed very thoroughly the literature up to that date. Nor is it mentioned by Osuntokun (1971) in his massive review of almost 10,000 neurological cases seen at Ibadan between 1957 and 1969. Girard, Dumas & Jacquin-Cotton (1970) in a report of a single case of acquired hypokalaemic paralysis due to an acute diarrhoeal episode in an African Senegalese woman with chronic renal disease, mention that they have seen no cases of the hereditary periodic type over a period of 10 years. Other reports of the pattern of neurological diseases in the African also make no mention of the occurrence of periodic paralysis (Hutton, 1956; Gelfand, 1957; Reef, Lipschitz & Block, 1958; Kaushik, 1961; Cosnett, 1964; Haddock, 1965; Ojiambo, 1966; Spillane, 1969; Dada *et al.*, 1969; Billinghurst, 1970).

Even if its occurrence in Africans is not so rare as it might seem, the disease is so unlikely to occur that its differential diagnosis is worth a brief mention. The spontaneous onset of flaccid tetraplegia without preceding symptoms, is rather too sudden for acute infective polyneuritis (Guillain-Barré syndrome), porphyria or poliomyelitis. The absence of sensory disturbance and sphincter involvement tends to exclude spinal cord disorders such as compression, subluxation of the odontoid peg and acute necrotic myelopathy (transverse myelitis). The age incidence is wrong for basilar artery insufficiency and the duration of paralysis too long for cataplexy. Hyperkalaemic period paralysis (*adynamia episodica hereditaria*), an even rarer disorder than the hypokalaemic disease, is characterized by younger age at onset and more frequent and transient attacks of paralysis related to exercise and meals. A still rarer disorder, normokalaemic periodic paralysis, resembles the hyperkalamic type.

It is possible that the most frequent error is to diagnose hysteria or psychoneurosis, as happened during the first attack in this case.

The exact nature of the pathological process in hypokalaemic periodic paralysis is still far from clear. The clinical picture differs considerably from that seen in hypokalaemia due to deficient intake of potassium or excessive loss of potassium from the gastro-intestinal tract of urine. The site of the disorder lies only in the skeletal muscles, not in the

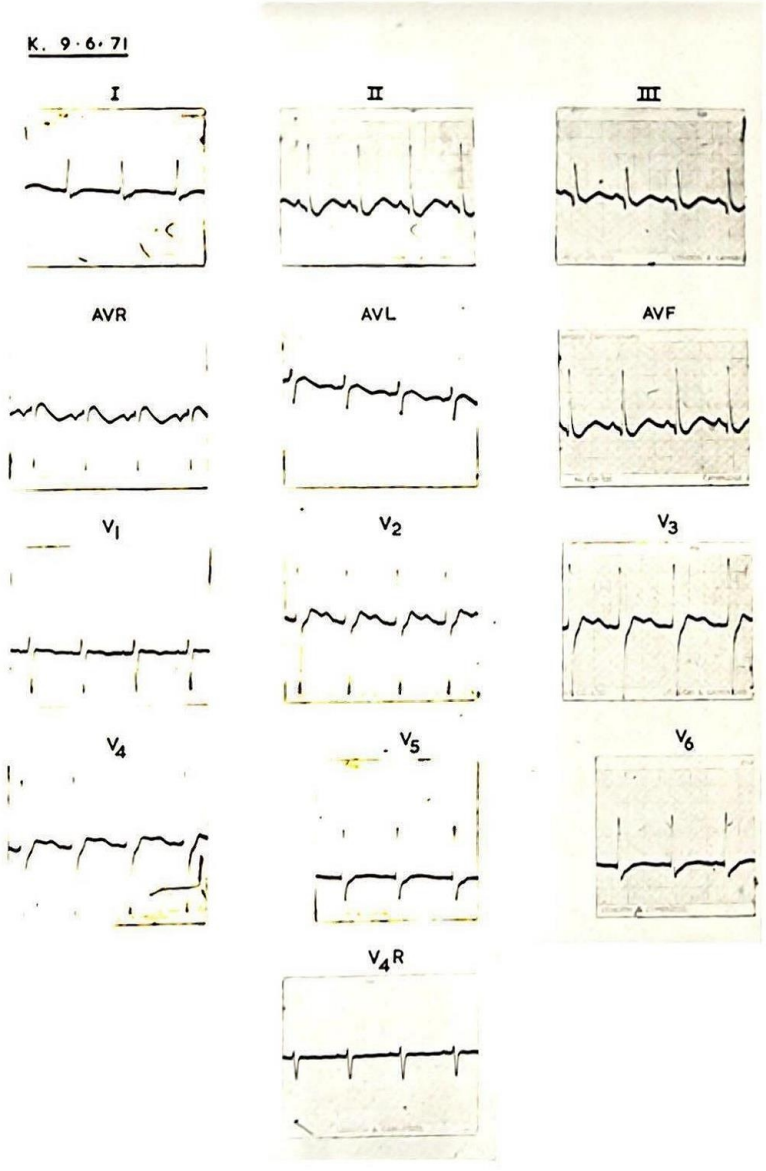


FIG. 1.

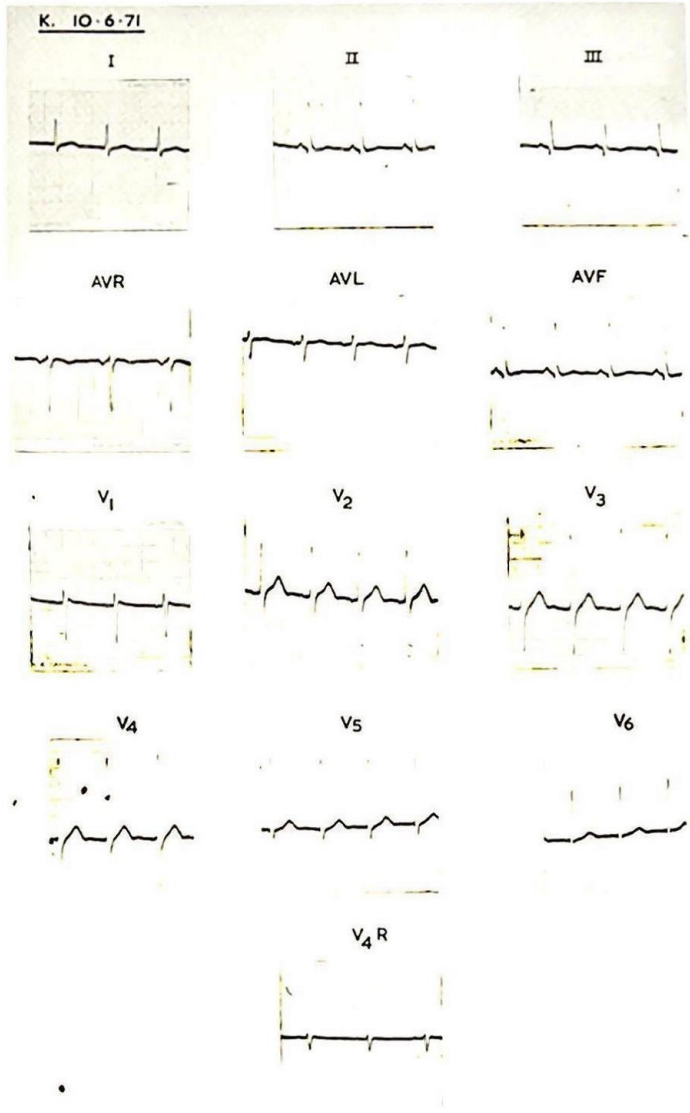


FIG. 2.

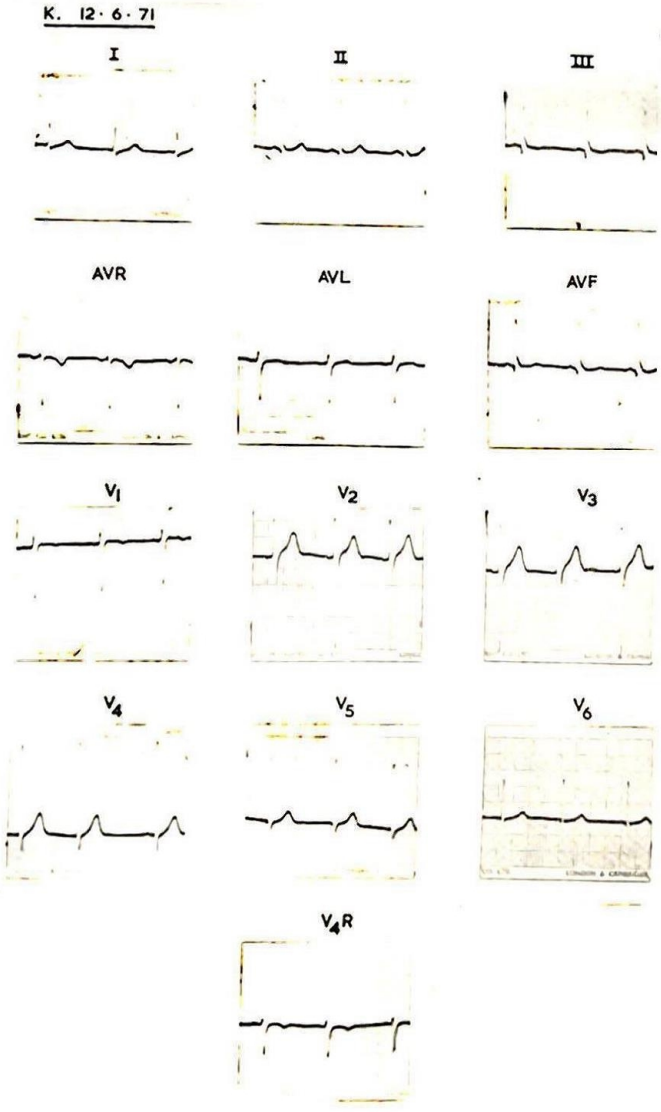


FIG. 3.

nerves or smooth muscles of the gut. It is likely that the hypokalaemic is the result and not the cause of the pathological process which is associated with the paralysis. Recent work (Hofmann & Smith, 1970) has shown that the muscle membrane potential is lower than normal, suggesting that the sodium pump mechanism may be marginally defective, a state of affairs liable to be aggravated by cold or exertion (both of which may precipitate paralysis). While the sodium pump is not working normally, there would be a drift into the muscle cell of sodium ions and water (as has been observed). Sudden reactivation of such a marginally defective sodium pump would result in the sudden extrusion of sodium ions and the sudden inflow of potassium ions into the muscle cells, the latter causing a sudden fall in the extracellular (i.e. serum) concentration of potassium. Some of the intracellular potassium might be rendered ionically inactive by being caught up with the deposition of glycogen, granules of which can be seen in the muscles. Such a mechanism could elegantly explain the fall in serum potassium. But it would not explain the paralysis itself. There now seems to be evidence that there is also a structural disturbance in the sarcoplasmic reticulum of the muscles and a flaw in the excitation-contraction coupling process, and that the fundamental defect here may be inadequate release of calcium ions from the sarcoplasmic reticulum (Weller & McArdle, (1971). How the inadequate release of calcium ions from the sarcoplasmic reticulum is related to a defective sodium pump at the membrane awaits elucidation. Possible mechanisms are discussed by Bradley (1969) who draws attention to a number of similarities present in all three forms of periodic paralysis.

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