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## Familial Hypokalemic Periodic Paralysis in Blacks†

Jack M. Bernstein, MD\* and Michael Kleerekoper, MD\*\*

*Hypokalemic periodic paralysis is an uncommon disease that has rarely been reported in blacks. Historical, clinical, and pathological findings in three black patients, in addition to those previously reported in the literature,*

*suggest that the spectrum of the disease is identical in both blacks and whites. Considering the disease as either different or unknown in the black population is probably not justified.*

Familial hypokalemic periodic paralysis is an uncommon syndrome characterized by flaccid paralysis of the skeletal muscles associated with hypokalemia. The paralytic episodes are usually associated with profound hypokalemia and almost always aborted by the administration of potassium. Death due to either cardiac arrhythmias or paralysis of respiratory muscles is unusual. In North America, the disease has been reported almost exclusively in Caucasians.

We have been able to find only four reports (1-4) comprising four black families in whom this disease existed. In this paper, we report an additional two black families with familial hypokalemic periodic paralysis.

### Case Reports

#### Case 1

A 22-year-old black woman was admitted to the hospital for evaluation of "low potassium." At the age of nine, she went to sleep one night on a hard floor, and when she awoke the next morning she found that she could not move, although she could talk and swallow. She was given orange juice and within several hours regained her strength. Soon after, she was seen by a physician who told her that she had "low potassium"; she received oral potassium supplementation which she has taken since. Attacks of paralysis occurred after she ate a meal, especially those rich in carbohydrates, or drank soda pop. Occasionally, attacks were precipitated by cold or sleep. Exercise followed by rest induced an attack although continuing to exercise slowed the onset of weakness. Attacks occurred more frequently in the winter than in the summer, and thus, she needed less potassium triplex in the summer. She denies that she has chronic diarrhea or diabetes, uses diuretics, or eats large amounts of licorice.

Her father (Case 2) is the only other affected member of this family.

Physical examination revealed a young black woman in no apparent distress. General physical examination was normal except for a scar over the lower back secondary to an operation for scoliosis. Neurologic examination revealed minimal proximal weakness of the lower extremities. Blood pressure was 106/70, pulse 96 and regular.

Laboratory examination revealed sodium 132, potassium 4.1, chloride 104, and  $\text{HCO}_3^-$  24 with normal glucose and  $\text{T}_4$  on admission. On the third hospital day, 75.0 gm of glucose was infused over one-and-a-half hours. Two hours after first receiving the glucose, the patient evidenced an objective decrease in strength with weakness of plantar flexion and extension. In the next hour, the patient became almost completely paralyzed; only her facial and respiratory muscles were spared, and she was areflexic. The serum potassium at this time was 2.1 mEq/L. An electromyogram at this time showed a marked loss of motor unit activity, but the remaining motor units appeared normal in configuration and duration. Frequent P waves and fibrillations were seen. On full contraction, there was rapid recruitment of motor units that appeared small and polyphasic. The patient was given

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intravenous and oral potassium and by the next morning she had recovered most of her strength. Repeat EMG showed marked improvement, but some myopathy remained. A muscle biopsy (Fig. 1) showed a vacuolar myopathy.

The patient is presently doing well. She takes potassium supplements several times a week when she feels an attack beginning, and these successfully abort the attacks.

### Case 2

A 51-year-old black man, the father of the patient described above, went to sleep one night at the age of 13 and awoke paralyzed. He remained paralyzed although he was able to speak and swallow. He was carried from place to place, and after three days, he regained his strength. Major attacks came on approximately every three months and were precipitated by eating, especially if followed by rest or too much exercise. "Keeping moving" sometimes ameliorated the attacks. By chance, he discovered that drinking orange juice or eating bananas shortened the period of paralysis.

In 1955 at the age of 26, he was evaluated at another institution where hypokalemic periodic paralysis was diagnosed and potassium supplementation after each meal was prescribed.

Through the late 1950s, the attacks came further and further apart, and he was able to decrease his potassium supplements until he took them only when his weakness started. In this way, complete paralysis was prevented. The periodic weakness was replaced by a generalized permanent weakness, and his lower extremities were more involved than his upper extremities. At present, he only rarely takes potassium.

General physical examination was negative except for evidence of chronic lung disease, keratosis pilaris, and generalized muscle wasting. Reflexes were physiologic and symmetrical. Muscular strength was reduced; the most severe weakness involved the flexors of the hip and knee. The quadriceps femoris exhibited only minimal weakness. Both the deltoids and the triceps were weakened, but the biceps brachialis was normal. Accompanying the weakness was marked muscle wasting in these muscle groups. Laboratory examination revealed sodium 140, potassium 3.9, chloride 100,  $\text{HCO}_3$  37. Complete blood count, urinalysis, and VDRL were all normal or negative.

Presently the patient is doing well and takes no medications. His weakness does not significantly impair his activity, except for some difficulty in climbing stairs.

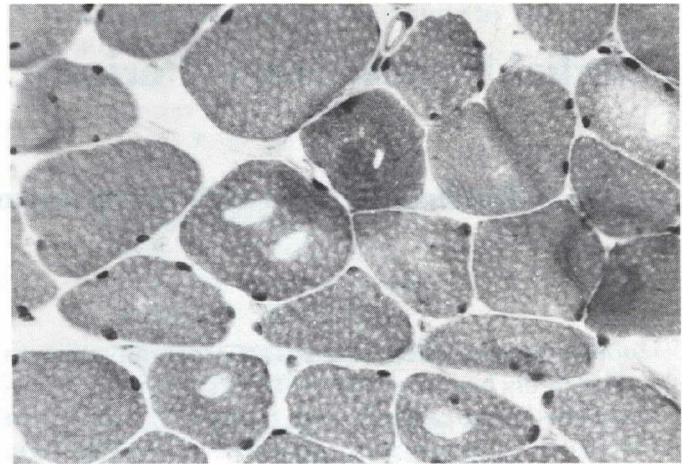


Fig. 1  
Muscle biopsy.

Vacuoles are present in several of the muscle fibers (hematoxylin and eosin). Original magnification 25 X, enlarged to approximately X 750.

### Case 3

A 29-year-old black man was admitted to the hospital with quadriplegia. He had his first attack of weakness at the age of 13, when he awoke one morning paralyzed. He was admitted to another hospital, but the cause of the weakness was not found, and he was discharged after several days, after he recovered spontaneously. During the next year, when he had several further episodes of paralysis, a diagnosis of hypokalemic paralysis was established. Over the next several years, the patient was hospitalized for paralysis many times. Attacks were precipitated by emotional upset, sleep, alcohol intake, heavy meals, or excessive exercise. In 1971, his therapy was changed to spironolactone (50 mg. p.o. t.i.d.), and he was placed on a low carbohydrate, low sodium diet. In spite of this, he had a severe attack of paralysis with respiratory decompensation that required intubation. His potassium at that time was 1.6 mEq/L. Since then, the patient has done well with only occasional bouts of weakness. A brother died at the age of 21 during an attack of paralysis, although no other family members, including parents and three siblings, are affected.

Physical examination revealed a young black man with quadriplegia who was able to move his head and neck. He was areflexic. The remainder of the examination was normal. Blood pressure was 130/80, and pulse 80 and regular.

Laboratory examination revealed sodium 140, potassium 1.7, chloride 112,  $\text{HCO}_3$  25.  $\text{T}_4$  was normal. Intravenous supplementation with potassium resulted in almost complete restoration of strength within 24 hours. The

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**TABLE I**  
**Characteristics of Periodic Paralysis in Blacks**

Reference	Family	Sex	Age of Onset	Precipitating Factors	Other Family Members Affected
1	1	Male	34	Meals, exercise, sleep	Son
2	2	Male Male	23 24	Exercise, sleep Exercise, high carbohydrate meal, sleep	Total of 27 members in 4 generations, 15 men and 12 women; 3/27 died during an attack.
3	3	Male	27	High carbohydrate meals, exercise, sleep	
Present report (Case 1)	4	Female	9	Cold, sleep, exercise, heavy meals	
Present report (Case 2)		Male	13	Heavy meals, sleep, exercise	Daughter
Present report (Case 3)	5	Male	13	Emotional upset, sleep, alcohol, heavy meals, exercise	Brother (died during attack)

patient was discharged, and potassium supplementation was prescribed. He has done well since.

### Discussion

Hypokalemic periodic paralysis classically has its onset during the first to third decade of life. Within each family, the age of onset is relatively constant though it may vary from family to family. It is thought to be inherited as an autosomal dominant with incomplete penetrance and affects men more than women (5).

Clinically, several factors may predispose to an attack: exposure to cold temperatures, vigorous exercise followed by rest, eating a meal rich in carbohydrates, or emotional upset (5,6). Eating refined carbohydrates or a large carbohydrate meal is the most common, precipitating cause of paralysis. Most attacks occur as paralysis when the patient awakens. Attacks may last several hours to several days. They occur less and less frequently with the passage of time, sometimes completely abating in later life. Permanent myopathic weakness may occur as the sequela of years of recurrent attacks (6).

Histologically, affected muscles show a vacuolar myopathy that is characteristic of hypokalemic paralysis but is also seen in other chronic hypokalemic states, systemic lupus erythematosus, chloroquine myopathy, steroid myopathy, and chronic polymyositis (7).

The pathology of the attacks is not completely under-

stood. Between attacks of paralysis, the potassium content of muscle increases dramatically, suggesting an intracellular shift of the potassium (8). In vitro studies by Hofman and Smith (8) have shown that all diseased muscle specimens are significantly depolarized in vitro, irrespective of the patient's clinical condition. Insulin, at normal serum potassium concentrations, further depolarizes the fibers. These findings may account for the patient's sensitivity to carbohydrate ingestion and to insulin.

The disease has been treated with potassium supplementation or spironolactone. Recently, acetazolamide has been found to be effective in the prophylaxis of hypokalemic, as well as hyperkalemic, paralysis (9).

On the basis of the five black families on whom we have clinical data, it would seem that the disease behaves identically to that in whites (6) (Table I). In blacks, the disease begins in the second or third decade of life, but occasionally occurs late in the first or early in the fourth decades. Of 34 documented or clinically suspect cases (not including six cases [4] on whom there is no published clinical data), 12 women and 22 men were affected, indicating male predominance in spite of probably dominant inheritance (2).

Factors which precipitated attacks (Table I) were identical to those noted in the literature (5,6). We were particularly impressed with the onset of weakness in two of

our patients (Cases 1 and 3) after they drank sweetened soda pop and ate high carbohydrate meals. Four of the 34 patients died during attacks, a prevalence similar to that which had previously been reported (5).

Progression to permanent myopathic weakness as the patient grows older and attacks occur less often also seems to be the pattern in blacks. Our second patient, who is 51 years old at the present time, has obvious diffuse muscle wasting and generalized weakness although he no longer has acute exacerbations. His daughter (Case 1), who still suffers from hypokalemic paralysis, also has some early, permanent myopathic changes.

Histologically, both our first patient and that of Martinez-Catinchi, et al (3) had vacuolar myopathies with minimal atrophic changes. These findings are identical to those which have been previously reported (5).

## Conclusion

We have seen three cases of hypokalemic periodic paralysis in two black families. The total number of blacks reported in the literature now totals 40 in six separate, apparently unrelated families. Clinical features including age of onset, inheritance, precipitating factors, mortality, course and prognosis appear to be identical to those seen in the Caucasian population. To consider the disease in blacks as "almost unheard of" (5) or different from that seen in whites is probably not justified.

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