Is hypokalaemic periodic paralysis more prevalent in Maori?

The primary catchment area for Christchurch Hospital has a population of just over 367,000 of whom just over 12,000 (3.3%) are Maori. The presentation of two unrelated Maori men with hypokalaemic periodic paralysis to the department of Neurology, Christchurch Hospital in 1996 prompted review of previous cases diagnosed locally. A case history is presented plus a summary of the recorded Christchurch experience since 1983.

A 48 year-old Maori man was unable to get out of bed on waking and complained of weakness of all limbs. There was no family history of episodic weakness and he was on no medication. Examination revealed a well-looking man 3/5 to 3+/5 weakness in all muscles of the upper limbs bilaterally (worse proximally), 2/5 weakness at the hip and knee bilaterally and 3/5 weakness at the ankles and toes. There was no wasting nor fasciculation and tone and reflexes were normal. The remainder of his examination was unremarkable. Blood tests revealed hypokalaemia with a potassium of 2.0 mmol/L, sodium 144 mmol/L, glucose 8.5 mmol/L, CK 573 U/L (normal range: 25-175). An ECG was normal. Potassium supplementation was not given. The following morning the neurological examination was normal and potassium 3.7 mmol/L. He was discharged on no treatment but presented the following day having woken again with weakness; at this review the left ankle jerk was absent, serum potassium 2.7mmol/L. Full strength, normal reflexes and normokalaemia again returned after 24 hours. A 24-hour urine collection showed no renal potassium wasting and an arterial blood gas was normal, as were thyroid function tests. Investigations for secondary causes of hypokalaemia were negative. Exercise to exhaustion and oral glucose loading (50g/h for 15h) did not provoke hypokalaemia or weakness. EMG and repetitive nerve stimulation were not performed. On review of his past history and medical notes it became clear that he had suffered a similar episode in 1984 and one or two previous episodes in his early twenties. They all occurred on waking and lasted less than 24 hours. During hospital admission in 1984 his potassium was measured at 2.3mmol/L while weak, returning to 5.3 the following day.

The records of all previously recorded cases of hypokalaemic periodic paralysis seen by a neurologist at Christchurch Hospital since 1983 were examined. Cases of hypokalaemic periodic paralysis associated with thyrotoxicosis were excluded.

There were three cases diagnosed with hypokalaemic periodic paralysis during this period in addition to the case presented. Two of these were Maori, all were male. One of the Maori cases reviewed was a 38-year-old man who presented with a 20 year history of recurrent episodes of weakness which occurred on waking or after rest following exercise. Severe episodes resulted in inability to walk for approximately one hour. Full strength did not return for 24-48 hours. There was no family history of similar illness, including 15 siblings. Intravenous glucose infusion resulted in hypokalaemia (nadir 2.3 mmol/L) which was associated with clinical limb weakness and reduced CMAP amplitude on EMG, both returning to normal with normokalaemia. The other Maori case presented with recurrent episodes of mild weakness over at least 18 months and was admitted to hospital during a more severe episode of weakness. Weakness was described as mild in the upper limbs and moderate in the lower limbs and trunk. He fell walking on several occasions and was unable to rise from a chair without using his arms. Serum potassium was 2.3 mmol/L, CK 277. The potassium level returned to normal without supplementation and full power return spontaneously within a few days. Tests for secondary causes of hypokalaemia were negative and EMG and muscle biopsy findings were non-specific. No provocation testing was performed.

Two cases labelled possible periodic paralysis were also found. One was a Maori man who presented with a single episode of weakness with hypokalaemia provoked by a carbohydrate-rich meal. The weakness and hypokalaemia resolved spontaneously but he has been lost to follow-up.

Three of four cases diagnosed with hypokalaemic periodic paralysis in Christchurch since 1983 are of Maori descent. Two additional cases of possible periodic paralysis exist, one of whom is Maori. The low (3.3%) Maori population in Canterbury makes the probability of our findings being due to chance alone low ($p<<0.001$ by binomial probability distribution) regardless of the 'possible' Maori case being considered. A chance finding remains unlikely even if only two of the Maori cases are accepted as definite hypokalaemic periodic paralysis ($p<0.01$). The very small number of cases makes a definite conclusion difficult. The low number of cases reflects the low prevalence of the condition but also may be due to underdiagnosis or diagnosis by other agencies in the region without referral to a neurologist or entry in

hospital records. There is nothing to suggest to us that these possible confounding factors would create an ethnic bias in favour of Maori receiving the diagnosis. It has been suggested, rather, that Maori people are less likely than non-Maori in New Zealand to come to medical attention in the event of illness.[1] A prevalence of three cases in a population of 12,000 Maori (2.5/10000) is high compared with the prevalence reported in some other populations, for example 0.4/100 000 in Finland and 1.25/10000 in Denmark.[2] Underdiagnosis within the Maori population remains likely as two of our cases presented at least 20 years after the first onset of symptoms and the estimated prevalence in other populations has risen markedly with increased knowledge of and interest in the condition.[2] The high incidence of permanent muscle weakness in other series is a cause for concern but was not evident in these patients.[3,4] None of our cases reported a family history of periodic paralysis and they may represent sporadic incidence of the condition, but the possibility of undetected related cases remains. Two of the Maori patients were not able to give an accurate family history due to prolonged isolation from other family members who live in the North Island.

We conclude that the prevalence of hypokalaemic periodic paralysis is likely to be substantially higher among the Maori than the non-Maori in New Zealand. This association has not been previously reported, to our knowledge. Further study of the genetics of hypokalaemic periodic paralysis in the Maori is required. We are currently investigating the prevalence of this disorder in the Auckland region, which has a much greater Maori population. ■

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