

## Hypokalemia Complicating Duchenne Muscular Dystrophy

BRUCE McDONALD, M.D.,<sup>a,b</sup> AND SETH A. ROSENTHAL, B.A.<sup>b</sup>

<sup>a</sup>*Department of Pediatrics, Bridgeport Hospital, Bridgeport, Connecticut;* <sup>b</sup>*Yale University School of Medicine, New Haven, Connecticut*

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Although patients with Duchenne muscular dystrophy (DMD) have been shown to have decreased total body potassium levels, serum potassium levels have generally been thought to be within normal limits. We report two siblings with DMD noted to be hypokalemic in conjunction with a respiratory illness. Hypokalemia may have exacerbated the pre-existing pulmonary insufficiency in these patients. The literature concerning hypokalemia and DMD is reviewed, and recommendations for the closer monitoring of serum potassium levels in patients with DMD are presented.

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### INTRODUCTION

Duchenne muscular dystrophy is a genetically determined disorder which causes progressive muscular weakness in male children and adolescents. The diagnosis may be considered when there are clinical signs of clumsiness or leg weakness, or when there is the incidental finding of elevated levels of aspartate aminotransferase (AST) and alanine aminotransferase (ALT) (formerly SGOT and SGPT). The disease is progressive, leading to the need for assisted ambulation by the age of ten years and progressive pulmonary insufficiency secondary to weakness of respiratory musculature by the end of the second decade of life. The patient's demise is most often related to the respiratory insufficiency.

Patients with muscular dystrophy have previously been reported to have decreased muscle potassium content [1], decreased total body potassium [2], and serum hypokalemia [3]. This latter condition may cause additional weakness of the respiratory muscles and exacerbate the underlying chronic pulmonary insufficiency. We report two siblings with Duchenne muscular dystrophy and hypokalemia. In one patient the hypokalemia was severe and may have contributed to his death.

### CASE REPORTS

#### *Patient 1*

A ten-year-old boy with the diagnosis of Duchenne muscular dystrophy was admitted to the hospital because of respiratory difficulty. He had been well until three days prior to admission when he developed a low-grade fever, congestion, and cough. During this time he became anorectic and was able to take only clear liquids devoid of potassium. Although normally he could ambulate with a leg brace, he experienced increased muscle weakness, was unable to ambulate, and developed swallowing difficulty. There was no vomiting or diarrhea.

On physical examination he was noted to have severe and generalized muscular weakness and lethargy, with temperature, 100.2°F; pulse, 110/minute; respirations,

TABLE 1  
Laboratory Studies of Patient 1

Time	Serum K+ (mEq/L)	Urine K+ (mEq/L)
Admission	1.7	—
5 hours	1.7	14
10 hours	2.1	20
24 hours	2.9	—
48 hours	3.6	—

26/minute; blood pressure, 110/70 mm Hg. There were diminished breath sounds at the bases of both lungs and absent bowel sounds on auscultation.

The following laboratory data were reported: WBC, 21,000 cells/mm<sup>3</sup> with a predominance of neutrophils; arterial blood gas (face mask O<sub>2</sub>), pH 7.22, pCO<sub>2</sub> 60 mm Hg, PO<sub>2</sub> 144 mm Hg. His electrolytes were: Na, 140 mEq/L; K, 1.7 mEq/L; CO<sub>2</sub> content, 21 mEq/L. The chest radiograph revealed bilateral basilar infiltrates.

An electrocardiogram revealed small, flat, T waves but no evidence of an arrhythmia. He was treated with intravenous antibiotics and potassium chloride (0.2 mEq/kg/hour). Additional laboratory studies were obtained as shown in Table 1.

Despite ventilatory assistance the patient's condition deteriorated, and he expired 60 hours after admission.

#### *Patient 2*

A 12-year-old sibling of the previous patient was admitted because of similar respiratory symptoms. He had been diagnosed as having Duchenne muscular dystrophy by muscle biopsy nine years prior to this admission. His clinical course had not been unusual, and at the time of admission he was able to ambulate with leg braces and a walker. Although he had experienced an illness similar to his brother, he had been able to continue to eat a normal diet in diminished quantity and subjectively did not experience an increase in his muscular weakness.

On physical examination, he was noted to have generalized muscular wasting and weakness, but was alert and in no acute respiratory distress, with temperature 102°F, pulse 120/minute, respiration 24/minute, blood pressure 105/80 mm Hg. There were decreased breath sounds at both lung bases. Bowel sounds were normal. The chest radiograph revealed small bilateral basilar infiltrates.

His laboratory data were reported as follows: WBC, 18,900 cells/mm<sup>3</sup> with a predominance of neutrophils; Hb, 11.7 gm/dl; Hct, 34 volume percent. Serum electrolytes were as follows: Na, 136 mEq/L; K, 2.4 mEq/L; CO<sub>2</sub> content, 26 mEq/L. Urine K+ was 14 mEq/L.

He was treated with intravenous antibiotics and potassium chloride. After 48 hours, his serum potassium was 4.3 mEq/L. The respiratory symptoms slowly subsided, and he was discharged on the eighteenth hospital day, at which time his serum potassium was 4.4 mEq/L.

#### DISCUSSION

These two patients developed symptoms of respiratory decompensation in conjunction with a presumed viral respiratory illness. There was evidence of bilateral infiltrates, which is not unexpected in patients with muscular dystrophy who are afflicted with mild respiratory illnesses. Pneumonia may develop because of general-

ized muscular weakness and a diminished respiratory excursion or because of difficulty in handling normal respiratory tract secretions. Both patients also presented with severe hypokalemia, which is known to result in poor muscular contraction and which may have resulted in a severe and irreversible exacerbation of the pulmonary insufficiency in one of the patients.

Although there are a number of reports documenting diminished total body potassium [4,5], the level of serum potassium is generally normal [6,7], and there is only one previous case report of hypokalemia [3] which was considered to be of clinical significance. There has been speculation that the hypokalemia may be genetically determined and may be the primary defect in muscular dystrophy, leading to diminished contractile ability of the muscles [8]. Other investigators have shown that the diminished total body potassium is related to a decrease in intracellular potassium concentrations in the dystrophic muscle tissue. Fat and collagenous fibers which replace muscle bundles in dystrophic muscle have workedly diminished water and potassium content [2,9]. It is this replacement of potassium-rich normal muscle fibers with potassium-poor fibers which leads to the diminished total body potassium. There was no evidence in these two patients or in other studies [10] of renal potassium wasting.

During the course of the respiratory illnesses, both patients became anorectic, which precipitated their hospital admissions. The first patient noted progression of his normal degree of muscular weakness, resulting in difficulty eating and swallowing solid foods. He was able to ingest only clear liquids which were deficient in potassium. Patient 2 continued to ingest his usual diet but in a diminished quantity. It is our conviction that, because of the abnormal potassium reservoir, diminished intake of potassium resulted in a low serum potassium unrelated to renal wasting. Although a cardiac arrhythmia was not evident, there was evidence of depressed T waves and ST segments on the electrocardiogram. This hypokalemia and exaggerated muscular weakness complicated the treatment of the respiratory illness and may have caused the irreversible cardiorespiratory failure in patient 1.

Patients with advanced muscular dystrophy should be prospectively monitored for hypokalemia as part of their standard care. In addition, it may be advantageous to monitor serum potassium levels in patients with muscular dystrophy and to encourage oral supplementation during illnesses which may limit ingestion of a normal diet. High potassium-containing foods or additional supplementation should be encouraged in those patients known to have borderline-low, or low, serum potassium.

## REFERENCES

1. Cumings JN: The potassium content of muscle in disease. *Brain* 39:153-156, 1939
2. Bland WH, Lederer M, Cassen B: The significance of decreased body potassium concentrations in patients with muscular dystrophy and nondystrophic relatives. *N Engl J Med* 276:1349-1352, 1967
3. Soloway SS, Mudge GH: Acute hypokalemia as a possible cause of death in a patient with advanced muscular dystrophy. *Johns Hopkins Med J* 144:166-167, 1979
4. Bland WH, Cassen BL, Lederer M: Body potassium content in patients with muscular dystrophy. *Ann NY Acad Sci* 110:282-290, 1963
5. Bland WH, Cassen B, Lederer M: Decreased body potassium in nondystrophic relatives of patients with muscular dystrophy: A biochemical trait. *N Engl J Med* 270:197-198, 1964
6. Danowski TS, Wirth PM, Leinburger MH, Randall LA, Peters JH: Muscular dystrophy III. Serum and blood solutes and other laboratory indices. *AJDC* 91:346-355, 1956
7. Tyler FH, Perkhoff GT: Studies in disorders of muscle VI. Is progressive muscular dystrophy an endocrine or metabolic disorder. *Arch Int Med* 88:175-190, 1951

8. Howland JL: Abnormal potassium conductance associated with genetic muscular dystrophy. *Nature* 251:724–725, 1974
9. Shy GM, Cumming DJ, Berg L, Horvath B: Muscular dystrophy potassium exchange in residual muscle. *J Appl Phys* 8:33–36, 1955
10. Garst JB, Vignos PJ, Hadaday M, Matthews D: Urinary sodium, potassium and aldosterone in Duchenne muscular dystrophy. *J Clin Endo Metab* 44:185–188, 1977