Renal dysfunction is rare in Fukuyama congenital muscular dystrophy.

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Abstract

BACKGROUND: The leading cause of death in patients with Fukuyama congenital muscular dystrophy (FCMD) is congestive heart failure or respiratory dysfunction, which is same as that in Duchenne muscular dystrophy (DMD). Recent studies reported that renal dysfunction is a common complication and an increasing cause of death in advanced DMD. It can be attributable to circulatory instability or inappropriate use of drugs for treating cardiac dysfunction.

METHODS: We retrospectively evaluated renal function in 38 genetically diagnosed patients with FCMD (range, 1.3-32.9 years; mean age, 13.7 ± 6.9 years) using cystatin C. We examined possible relationships of cystatin C with blood natriuretic peptide and creatinine levels along with cardiac echocardiography findings.

RESULTS: Twenty-five patients were treated for cardiac dysfunction. Elevated cystatin C level was detected only in two, who also showed proteinuria, glycosuria, hematuria, and extremely high β2-microglobulin levels on urine tests, and were thus diagnosed with renal tubular cell damage. Because both patients were treated for intractable epilepsy with various antiepileptic drugs, including valproic acid (VPA), and had low serum carnitine levels, renal tubular cell damage was considered as an adverse effect of VPA. Unlike patients with DMD, no patient with FCMD had renal dysfunction. Such a rare occurrence of renal dysfunction can be attributable to mild cardiac dysfunction, short disease duration, and careful and early fluid management.

CONCLUSION: Renal dysfunction is rare in patients with FCMD; however, renal tubular cell damage should be ascertained, particularly in those undergoing VPA treatment for epilepsy.

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