Brain dystrophin-glycoprotein complex: Persistent expression of beta-dystroglycan, impaired oligomerization of Dp71 and up-regulation of utrophins in animal models of muscular dystrophy

ABSTRACT

A link has been established with membranes at the vascular-glial interface in the forebrain, suggesting that -dystroglycan is associated with them.

INTRODUCTION

A major issue in the treatment of muscle dystrophy is the persistent expression of beta-dystroglycan and consequent loss of function of utrophins. The aim of the present study was to investigate whether and to what extent beta-dystroglycan gene is expressed in muscle of human patients with muscle dystrophy.

Methods:

A total of 60 patients with muscle dystrophy were selected for the study, including 52 patients with a total of 16 cases. A total of 9 patients were randomly assigned to receive either intravenous vitamin D 3 or placebo (n=9). Thirty-five patients were assigned to receive either According to the main hypotheses, deficiency in dystrophin inevitably leads to muscular dystrophy as its membrane cytoskeletal component is not present and results in impaired sarcolemmal integrity and clustering of ion channel complexes. A lack of dystrophin leads to the disintegration of sarcolemmal components, which makes muscle fibers from patients with Duchenne muscular dystrophy more susceptible to necrosis. This could explain why all dystopian-associated glycoproteins are reduced in bulk skeletal muscle and may also cause a decrease in osmotic stability and greater vulnerability to stretch-induced injury in dystropy-deficient muscle fibres. There are seven promoters that drive the different types of dystrophin protein (Dp) isoforms.

CONCLUSION

Remarkable conclusions This report indicates that -dystroglycan is not found at high concentrations in central neurons of the forebrain region, but may be located at the interface between endothelial cells and glia. These structures may represent endfeet on astrocytes at least to the blood-braIN barrier, and it is unlikely that this is due to up-regulation of utrophin isoforms. However, it was shown that Dp71 exists in contrast to its normally oligomery. Molecular mechanism has been linked to