Potential therapy for congenital muscular dystrophy

December 30 2008

Current research suggests laminin, a protein that helps cells stick together, may lead to enhanced muscle repair in muscular dystrophy. The related report by Rooney et al, "Laminin-111 restores regenerative capacity in a mouse model for alpha 7 integrin congenital myopathy," appears in the January 2009 issue of *The American Journal of Pathology*.

Muscular dystrophy is a group of inherited genetic diseases that cause progressive muscle weakness. In one type of muscular dystrophy, patients with mutations in the adhesion molecule alpha 7 integrin experience delayed developmental milestones and impaired mobility. There is currently no treatment or cure for alpha 7 integrin congenital myopathy.

Interactions of alpha 7 integrin with laminin, an extracellular protein found surrounding muscle fibers, promote muscle cell health and survival. Alpha 7 integrin has also been implicated in muscle repair. To determine if alpha 7 integrin is critical for muscle repair, researchers led by Dr. Dean Burkin at The University of Nevada School of Medicine examined the response to muscle damage in alpha 7 integrin-deficient mice. They found that alpha 7 integrin-deficient muscle exhibited defective muscular regeneration. Injection of laminin-111, however, restored muscle repair and regeneration.

Paper: Rooney JE, Gurpur PB, Yablonka-Reuveni Z, and Burkin DJ: Laminin-111 restores regenerative capacity in a mouse model for alpha 7 integrin congenital myopathy. Am J Pathol 2009 174: 256-264

Source: American Journal of Pathology