MedicalXpress

Restoring breathing capacity in Duchenne muscular dystrophy by activating the brain

September 27 2017

New research published in *The Journal of Physiology* today suggests that enhancing breathing via the brain may limit deficiencies in respiratory capacity in Duchenne muscular dystrophy (DMD) patients.

Duchenne muscular dystrophy (DMD) is a fatal genetic neuromuscular disease, which can lead to respiratory failure. Respiratory muscle dysfunction is recognized in DMD, however a thorough assessment of nervous system control of respiratory muscles is lacking.

Lead investigators David Burns and Ken O'Halloran at University College Cork, in conjunction with collaborator labs at the University of Calgary and Trinity College Dublin, performed experiments in mice lacking dystrophin, the muscle protein that malfunctions in DMD. These mice have been shown to be a useful pre-clinical model of DMD as they demonstrate many of the hallmark features of respiratory muscle dysfunction in DMD patients.

In the young dystrophin-deficient mice, the respiratory control system was impaired on multiple levels. Importantly, the researchers found that the brain was compensating for this by increasing activation of the diaphragm muscle.

This study only offers a snapshot of the deficits and compensations at one time point in the progression of the disease. It is essential the findings from the study in the dystrophin-deficient mice are confirmed in human DMD patients before its potential applications in these patients

can be considered.

David Burns, one of the lead investigators on the study, said:

'Further study of physiological neuromuscular mechanisms that compensate for the absence of dystrophin is needed. A better understanding may provide insight with potential applications to a range of neuromuscular diseases, beyond DMD.'

More information: David P. Burns et al, Sensorimotor control of breathing in the mdx mouse model of Duchenne muscular dystrophy, *The Journal of Physiology* (2017). DOI: 10.1113/JP274792

Provided by The Physiological Society