European Cystic Fibrosis Society

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Cystic Fibrosis Research News

Title:

Skeletal muscle contractility and fatigability in adults with cystic fibrosis

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What was your research question?

Cystic fibrosis (CF) transmembrane conductance regulator (CFTR) is expressed in human skeletal muscle, suggesting specific limb muscles abnormalities in CF patients. We tested the theory that CF patients with mild to moderate lung disease may have an altered ability for contracting their muscles and increased muscle fatigue compared to healthy people (controls).

Why is this important?

Recent evidence suggests that limb muscle function is frequently decreased in CF patients and has important clinical implications, including reduced exercise tolerance and quality of life, which in turn may impact patient's survival. It remains however unclear whether limb muscle dysfunction is the consequence of physical inactivity only or whether some factors specific to CF may be involved. Determining whether CF patients have or haven't abnormalities in their muscle limbs should help to develop interventions aiming to improve muscle function.

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What did you do?

Using non-invasive measurements, we compared limb muscle function of 15 CF patients with mild to moderate lung disease with 15 healthy controls of similar age, gender and levels of daily physical activity. Using voluntary muscle contractions and muscle contractions evoked by magnetic stimulation, we evaluated strength, endurance, mechanisms of fatigue and recovery capacity of the thigh muscle. We also determined the volume of the thigh using magnetic resonance imaging (MRI).

What did you find?

We found that CF patients have similar muscle endurance, mechanisms of fatigue and recovery compared to healthy controls. They had only small reductions in maximal muscle strength and non-voluntary ability for contracting their muscles which disappeared when reporting the results relative to the muscle size of the thigh.

What does this mean and reasons for caution?

These results suggest that a small reduction in muscle size, rather than an actual difference in the functioning of limb muscles possibly caused by the CF gene, is responsible for the difference in muscle performance between patients with mild to moderate lung disease and healthy controls. It is still possible that the CF gene may impact muscle function, hence, the present results should be confirmed in a larger sample of patients considering different classes of CFTR mutation.

What's next?

The present data further encourage the incorporation of physical strength training programs aiming to increase skeletal muscle mass in CF patients, which may in turn have positive effects on functional capacities, quality of life and survival.

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