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Cystic Fibrosis Research News

Title:

Peripheral muscle abnormalities in cystic fibrosis: etiology, clinical implications and response to therapeutic interventions

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What was your research question?

We summarized limb muscle abnormalities experienced by people with cystic fibrosis (CF). The focus of this overview was to describe the causes and consequences of limb muscle abnormalities in CF and how limb muscles respond to therapy (e.g. drugs or rehabilitation).

Why is this important?

CF is characterized by several systemic manifestations, including limb muscle abnormalities. Alterations in the limb muscles including reduced force, endurance and increased fatigability may have important clinical implications. These may include poor exercise capacity and reduced quality of life. It is thus important to better understand the mechanisms underlying limb muscle dysfunction in order to identify the best possible treatments targeting this problem.

What did you do?

We intended to give an overview of limb muscle abnormalities in people with CF. We first described the structural and functional alterations of limb muscles in CF. We then summarized the clinical implications of limb muscle dysfunction and discussed the potential impact of underlying factors. Finally, we described how the limb muscles of people with CF may respond to various therapeutic interventions and suggested some future research.

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What did you find?

Most people with CF have reduced muscle strength, which can be partly related to lower muscle mass. This muscle weakness leads to reduced exercise capacity. Lack of physical activity is probably a leading contributor to the observed muscle abnormalities. However, other disease related factors such as inflammation, poor nutritional status and exacerbations (flare-ups) may be important. This is particularly true in patients with severe disease. Altered CFTR protein (the damaged protein in CF) in limb muscles may also have metabolic consequences and contribute to increased susceptibility to muscle fatigue.

What does this mean and reasons for caution?

The mechanisms contributing to muscle abnormalities in CF are caused by many factors. This means that several and complementary strategies can be adopted to improve limb muscle function in CF. Exercise training is probably the most effective strategy. Other interventions may be necessary for patients who may not obtain full benefits from exercise (e.g. severe hospitalised patients). The effectiveness of pharmacological or nutritional therapies to improve muscle function in CF requires further studies. While this review focused on peripheral muscles (limb muscles), it did not cover respiratory muscles which may also be affected in people with CF and deserve specific attention.

What's next?

Larger studies are needed to further investigate the importance of limb muscle dysfunction and how abnormalities develop over time in CF. Future trials should test the efficacy of combining exercise training with other personalized therapeutic strategies targeting a specific mechanism of limb muscle dysfunction. These will likely include the stimulation of physical activity, proper and individualized nutritional support and the administration of drugs that stimulate muscle growth.

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