tant detectable effects on the short-term; as we have shown, there are not such detectable effects. We have provided a comprehensive description of the ventilatory and thoraco-abdominal pattern, operational volumes, and unassisted cough peak flow before and immediately after a single MI-E treatment.

We found no improvement effects on operational volumes or unassisted cough peak flow, only on breathing frequency. We do not think that our study provides misconceptions about the objective intended use of MI-E devices. It simply provides new and original insights. We have shown that, immediately after treatment, MI-E does not provide lung recruitment, does not affect thoraco-abdominal volume variations (and therefore respiratory muscle action and control), and does not improve unassisted cough peak flow and vital capacity. In contrast, the significant short-term effect is on breathing frequency, which remains significantly lower for 6.2 ± 1.8 min after MI-E treatment, which is a too short period to recommend MI-E to reverse rapid and shallow breathing patterns or to prefer MI-E over other, less expensive techniques. For this reason, we have not encouraged the use of MI-E to reduce breathing frequency and thereby improve dyspnea, and we have underlined in the discussion the need for further studies aimed to investigate the long-term effects of MI-E on breathing pattern.

In conclusion, we do not believe that our paper provides any "miscommunication," "doubtful clinical message," or improper recommendations of a preferred device. Actually, our study confirms that "there is no reason to use a MI-E device in stable DMD patients with not effective (as written by Toussaint et al) unassisted cough peak flow to specifically target lung volume recruitment." Conversely, we fully agree with Joshua Benditt, who observed that our study "supports the very important and growing notion that respiratory support for patients with neuromuscular weakness is much more than just a focus on the noninvasive ventilator and actually requires a holistic approach to ventilation, cough function, and maintenance of the mechanical properties of the lung and chest wall in a way that is as close to normal as possible."

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Assessment of Peripheral Muscle Function in Cystic Fibrosis: Why and How?

There is a growing body of evidence, in people with cystic fibrosis, to support pe-

ripheral skeletal muscle abnormalities that may contribute to increased fatigability and reduced physical activity. This may, in turn, further exacerbate exercise intolerance and reduce health-related quality of life, the latter of which is associated with increased risks of hospitalization and poorer survival in people with cystic fibrosis (see Gruet et al[1] for a review). Peripheral muscle testing in cystic fibrosis is thus relevant for both clinical and research purposes, notably for early detection and monitoring of limb muscle abnormalities, designing targeted therapies, and evaluating their effectiveness; however, at present, it is rarely used. Development and validation of suitable testing protocols for this patient group should help to increase its uptake. Ideally, a test should evaluate all components of muscle function (ie, strength, endurance, power, fatigability) that may be impaired in people with cystic fibrosis. To be suitable for use in clinical and research settings, the tests must be valid, reliable, and feasible, with normative reference data available to assist with data interpretation.

In this issue, Sheppard et al[2] evaluated the validity of 4 field tests (ie, 30-s sit-to-stand test, stair-climb power test, vertical jump height, and triple hop distance) as surrogate measures of quadriceps strength and power in subjects with cystic fibrosis. The rationale for this study centered around the fact that computerized dynamometry (eg, Cybex, CSMi, Stoughton, MA; or Biodex System 4 Pro, Biodex Medical Systems, Shirley, NY), which is the standard tool to evaluate peripheral muscle performance in humans, is costly and requires sufficient space and technical expertise. Based on their observed moderate-to-strong correlations between the performance on this battery of tests and quadriceps strength and the power assessed by computerized dynamometry, the authors recommended the use of these tests (and particularly the stair-climb power test) to assess quadriceps strength and power of people with cystic fibrosis. Although these findings underline the potential of such simple field tests to evaluate muscle performance in people with cystic fibrosis, these results should be interpreted with caution and several issues should be considered before these tests are considered fully appropriate for implementing into clinical practice.

These tests have primarily been used in older adults (eg, stair-climb power tests, 30-s sit-to-stand test) or healthy young adults (eg, vertical jump height and triple hop distance). Despite the increased survival age associated with cystic fibrosis, this is not particularly representative of the typical age of the cystic fibrosis population. Moreover, there is a lack of full validation studies, especially in people with cystic fibrosis. Some of these tests are dependent on technical aspects and may thus have a large learning effect, especially in sedentary individuals who are unaccustomed to this type of effort. Postural control, which is impaired in some people with cystic fibrosis,[3] may also confound the performance in some tests (eg, triple hop distance) and test-retest reliability may be an issue. Moreover, as acknowledged by Sheppard et al[2] their study, similar to previous work on this topic,[4,5] is limited by the small sample size and possible selection bias. The absence of normative reference data for some of these tests within the age range of the cystic fibrosis population is also an issue.

It is therefore difficult to state whether the performance on a given test for a given patient is abnormal and, subsequently, decide on the most appropriate intervention to put in place. Accordingly, multi-centric validation of these tests (ie, learning effect, inter- and intra-rater reliability, construct validity, responsiveness, and minimally clinically important difference) in people with cystic fibrosis is needed in addition to the development of normative data for children and young adults. Although we agree that the correlations reported by Sheppard et al[2] show interesting promise, for the reasons stated herein (ie, sample size, validity issues), it seems premature to use these tests as a valid surrogate of quadriceps strength in this population. Similar to the 1-min sit-to-stand test,[4,5] these tests should be viewed as additional and complementary measures of limb muscle function to be considered for people with cystic fibrosis. The tests used by Sheppard et al[2] have the advantage that they may better reflect activities of daily living compared with single-joint isometric or isotonic maneuvers and thus should display better ecological validity. However, confirmation of this is needed.

So, should we give up on strength measurements if computerized dynamometry is not available? Quite simply, no. Chairs with a fixed strain gauge offer an attractive (often underused) alternative for measuring limb muscle strength. Not only are commercially available or custom-built chairs with strain gauges (which allow isometric strength measurements) much cheaper than computerized dynamometry (usually ~$5,000–$7,000), but strain gauge measures of quadriceps strength also show excellent validity (ie, vs computerized dynamometry) and are highly reliable (as much as are Biodex values[6]) in both healthy individuals[7] and those individuals with chronic respiratory disorders.[6] Moreover, normative data are available when using standardized procedures.[8] Such chairs are also easily transportable and can be used to assess muscle function in other chronic respiratory conditions.[9,10]

Also, such isometric set-ups also enable assessments of limb muscle endurance and fatigability, which are often impaired in people with respiratory disease.[10-12] However, fixed strain gauges have not yet been adapted to evaluate muscle power in cystic fibrosis. In the absence of computerized dynamometry, an interesting alternative could be the use of peak power achieved during short-duration cycling tests, such as the Wingate test. The Wingate test has often been used in cystic fibrosis as an indicator of anaerobic capacity[13] and is well tolerated in this population. Such testing also provides a well-established measure of muscle power, which demonstrates strong agreement with other tests of muscle power determined when using computerized dynamometry (eg, repeated isokinetic contractions).[14]

In conclusion, we thank Sheppard et al[2] for providing further support for the argument that the future of exercise testing in the cystic fibrosis clinic should, perhaps, include appropriate assessments of peripheral muscle function rather than focusing only on aerobic exercise function. Specifically, their findings support recent evidence,[4,5] to substantiate the need to develop and implement simple functional tests that allow rapid evaluation of limb muscle function for people with cystic fibrosis. Moreover, their preliminary findings clearly call for further validation of such tests in the cystic fibrosis population and the establishment of appropriate normative reference values. In the meantime,

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we strongly encourage the use of chairs with strain gauge measurements, which have been shown to be clinically feasible, to evaluate strength, endurance, and fatigability of limb muscles in people with cystic fibrosis.[9] Ultimately, the use of common methodology across cystic fibrosis centers to assess limb muscle function will promote the development of large prospective and retrospective multi-center studies, which are needed to (1) help us truly understand the prevalence, incidence, and development of limb muscle abnormalities in modern day cystic fibrosis; and (2) develop personalized therapeutic strategies targeting these issues.

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Clinical Usefulness of Functional Tests of Leg Muscle Strength and Power in Adults With Cystic Fibrosis

We thank Drs Gruet and Saynor for their insightful comments on our article, "Functional tests of leg muscle strength and power in adults with cystic fibrosis."[1] We believe that the comments they raised have stimulated an interesting discussion on the clinical implications of our study. First, Gruet and Saynor correctly stated that there are no reference values from age-matched healthy controls for the 4 tests selected for our study (stair-climb power test, 30-s sit-to-stand test, vertical jump height, and triple hop distance). We agree that, to establish a "diagnosis" or to determine the degree of functional impairment, as a percentage of predicted, we certainly need some normative values for comparison. Nevertheless, in clinical practice, many times, it is the change over time that is important to evaluate progression of disease. In certain circumstances, a comparison of an individual's own baseline may be more meaningful than the percentage of predicted compared with normative data from healthy controls. This is especially so with chronic diseases, for example, cystic fibrosis. For example, sometimes the change of lung function, that is FEV_1, is used as one of the criteria to define cystic fibrosis pulmonary exacerbation or to determine the effectiveness of clinical interventions during a pulmonary exacerbation. Therefore, we think that, even without appropriate age-matched normative reference values for the tests at this point, these 4 tests still have their usefulness clinically for assessment of the change over time in leg muscle strength and power in adults with cystic fibrosis. Besides, it is often debatable about the most appropriate "control" for people with cystic fibrosis. When healthy people are used as controls in cystic fibrosis studies, it always raises the question whether the differences in the outcomes are due to a respiratory disease or are specifically due to the condition of cystic fibrosis. To overcome this issue, Wells et al[2] included subjects with primary ciliary dyskinesia as a "respiratory comparison group" to control for the effects of impaired respiratory function on their outcome of interest, skeletal muscle metabolism. Further discussion is needed to decide on the best controls for people with cystic fibrosis.

Second, we agree with Gruet and Saynor that the vertical jump test and triple hop distance tests require motor skills, coordination, and good balance to perform well. For this reason, in our study, we ensured that the tester first demonstrated the task and that the subjects were allowed to have a trial run before the measurements were made. The subjects repeated each test 3 times, and the average score was used for analysis to account for variability in perfor-

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