Exercise Testing in Cystic Fibrosis

Patients with cystic fibrosis (CF) suffer progressively impaired exercise tolerance, and it is this—the inability to do things, to keep up with the physical tasks of daily life and play—that represents the biggest insult to patients’ quality of well-being. Exercise tolerance, expressed as the maximal workload achievable or as peak oxygen consumption on a progressive cycle ergometer test, has been shown to be a high and significant correlate with survival in CF. Therefore, it seems reasonable to measure exercise tolerance in these patients, and in fact, a recent consensus conference listed exercise tolerance as an important outcome variable to consider in CF intervention trials; yearly exercise testing is considered a standard of care in the United Kingdom.

Since Godfrey and Mearns described the response of patients with CF to exercise, the “maximal” exercise test on a cycle ergometer or treadmill (with collection of inspired and expired gases for analysis and measurement of minute ventilation, and with calculation of oxygen consumption and carbon dioxide production) has been considered the gold standard. However, this type of challenge does not duplicate children’s daily activity; it is also expensive, as well as tiring for the patient, and it requires a sophisticated laboratory not available in all institutions. These considerations have led investigators to consider different kinds of tests for evaluating CF patients’ exercise tolerance (Table 1). These tests have included supramaximal “sprint” tests and various submaximal tests, including walk tests and, as reported in this issue of Pediatric Pulmonology, a step test. All these tests have been shown to have validity, reliability, and definite advantages over the other tests; of course, the dark side is that they all have disadvantages as well. Balfour-Lynn and colleagues introduce into the realm of CF exercise tests the Great Ormond Street 3-minute step test, a variation on a theme in use in adult cardiac testing since the 1920s. They found a standardized test on a 15 cm step, at a cadence of 30 steps/min, to be tolerable for 91% of their patients and to be a reliable and reproducible way of increasing heart rate and breathlessness over the resting values, as well as decreasing oxyhemoglobin saturation ($S_{a,O_2}$). They found similar, but smaller, increases in breathlessness and pulse and comparable changes in $S_{a,O_2}$ with a 6 minute walk test.

This new test is a useful addition to the CF clinical testing armamentarium, in that it is a test that does not depend on patient motivation as maximal tests do, it should not vary as walk tests do with encouragement, and it is portable, allowing testing in the field. However, it has some clear limitations and is unlikely to become the exercise test for CF. With a set step height, its workload varies depending on the subject’s height (and weight). The investigators suggest that an important use of exercise testing is to follow exercise tolerance over time or with interventions, yet they have not validated this test as sensitive to changes with time or interventions. Particularly around the time of puberty, it will be very difficult to compare the outcome measures (pulse, $S_{a,O_2}$, breathlessness) of tests separated by months or years. This step test measures exercise tolerance only insofar as it assesses heart rate, $S_{a,O_2}$, and breathlessness during and after an exercise challenge (useful things to measure, without question). Since most subjects were able to complete the test, it does not measure maximal functional work capacity, nor even a set, known, reproducible submaximal workload.

Choosing the appropriate exercise test for CF depends on the specific question being asked and the aspect of exercise tolerance that is of interest:

- **Is it muscle strength?** Then a strength measure, e.g., the maximum amount of weight lifttable one time (1 RM) would be applicable.

- **Are anaerobic, supramaximal energy bursts available?** Then an anaerobic test, e.g., the Wingate test might be the most appropriate.

- **What is the maximum workload sustainable for a minute or the maximum amount of oxygen that can be brought in and processed?** The traditional progressive cycle or treadmill test will answer these questions.

- **Are ventilatory and cardiac responses to progressively increasing workloads appropriate?** If so, once again, the traditional progressive cycle or treadmill test is superb. Even a progressive test that does not push the patient to a maximal effort would suffice.

- **How much oxygen is needed to prevent exercise desaturation?** A simple double test on treadmill or cycle will do the trick: one in room air at progressively more difficult loads until desaturation occurs and then another with
supplemental oxygen at that workload. If desaturation still occurs, repeat the test with a higher $F_{I,O_2}$.

What is the ability of the patient to perform real-life work and the cost of that work in terms of oxygenation and heart rate? A 2-, 6-, 9-, 10, 12 or 12-minute walk test can give that information (in specific terms of distance walked in the given time), with a baseline for comparison with a repeat test after intervention (or the passage of time). Perhaps the “shuttle walking test,” which is similar to the walk tests in that it has subjects walk back and forth over a measured course, will be useful. In the shuttle walking test, the speed of traversing the course is specified and increases incrementally each minute.18,19 These tests can be used as maximal tests, with distance walked (times completing a 10-meter shuttle distance) as the outcome variable.

Conducting these tests takes experience and consistency. Some of the tests are highly dependent on patient effort (all the maximal tests, all the walking tests). Even the amount of encouragement investigators give the subjects must be standardized, as encouragement influences distance walked in a standard walk test as much as any intervention could be expected to do.14 It is encouraging to see more attention being paid to the development of useful exercise tests for patients with CF. Since there almost certainly will never be a single perfect exercise for all situations for a CF patients test, it probably makes sense for each CF Center to become familiar with one or two tests, and use them consistently.

—DAVID M. ORENSTEIN, MD

Department of Pediatrics
School of Medicine
Department of Health, Physical, and Recreation Education
School of Education
University of Pittsburgh
Children’s Hospital of Pittsburgh
Pittsburgh, Pennsylvania

REFERENCES

6. Cabrera M, Lough M, Doershuk C, DeRivera G. Anaerobic per-