

ONLINE SUPPLEMENTARY MATERIAL

(Standard Operating Procedures for Test Conduct and Test Reporting)

For detailed instructions for the test set-up, conduct and reporting of cardiopulmonary exercise testing (CPET) with measures of pulmonary gas exchange and ventilation the reader should refer to the European Cystic Fibrosis Society Exercise Work Group Statement on Exercise Testing (1) and the European Respiratory Society Statement on Standardisation for CPET in Chronic Lung Diseases (49).



Exercise Working Group

TEST INSTRUCTIONS

W_{peak} Cycle Ergometer Test

The peak power (W_{peak}) cycle ergometer test is similar to cardiopulmonary exercise testing, but involves running the protocol without ‘breath-by-breath’ measures of pulmonary gas exchange and ventilation. The W_{peak} test is an incremental cycling test (ramp or minute-by-minute stages) that involves the participant maintaining a constant pre-determined cadence (pedal speed) as workload is progressively increased until exhaustion. The primary measurement from the test is W_{peak} , alongside other measures of physiology and perceptuals. The following procedures are adapted from the European Respiratory Society (ERS) (49) and European Cystic Fibrosis Society (ECFS) (1) statements.

Equipment

- Calibrated cycle ergometer
- Pulse oximeter to continuously measure oxygen saturation (SpO_2 , preferably forehead sensor) and heart rate (HR)
- Scales to assess leg effort and breathlessness (e.g., Borg Scale)
- A source of oxygen
- Telephone (in case of emergency)
- Defibrillator (compulsory, depending on the testing location and conditions)
- ECG (optional)
- Supplemental O_2 (optional)

Pre-test preparations

- Refrain from eating for at least 2 hours before testing
- Participants should not exercise vigorously within 24 hours prior to completing the test
- Comfortable clothing should be worn with appropriate footwear.
- Usual pre-exercise bronchodilator and dose should be administered at least 10 minutes before the test
- Set-up the cycle ergometer to ensure appropriate settings for the participant. For small paediatric participants a paediatric ergometer and/or paediatric pedal cranks / vertical and horizontal saddle adjustments may be required
- The participant should sit in a chair next to the cycle ergometer at the starting position for ~10 minutes prior to the start of the test. During this time, contraindications should be checked, and pre-test measures (e.g., SpO₂, HR, effort, breathlessness) completed
- Measure symptoms on a standardised scale immediately prior to test commencement
- A warm-up period before the test is not required as the protocol comprised a warm-up
- When possible, effort should be made to familiarise participants with the equipment and what it feels like to cycle at a set cadence whilst workload increases prior to the test. If this cannot be undertaken at a separate visit then rest should be undertaken before the final test then begins.

Protocol

- A resting phase of 3 minutes, seated on the cycle ergometer, before started exercise should be used to collect HR, SpO₂ and ratings of effort and breathlessness. Resting values should be record towards the end of this phase when the participant has been seated in position.
- This is followed by a 3 minute period of unloaded cycling. Most cycle ergometers do not allow 0 W, this should be ~10 W, with this work rate stated in the test report. This value can be obtained from the yearly cycle ergometer calibration report.
- In participants with more severe CF lung disease and / or severe deconditioning, this unloaded phase may be shortened to 2 minutes (49).
- The incremental test begins – aiming for a total duration of the incremental phase of 8-12 minutes. This can be either an incremental ramp test or minute-by-minute stages. Minute-by-minute stages may be preferable in clinical practice where the cycle

ergometer may not be connected to a computer that imposes the work rate and the test must be run manually.

- The choice of work rate depends on a number of factors, such as age, sex, body size, disease severity, pulmonary function, fitness level, etc.
- The increments can be adjusted dependent on fitness levels, e.g.
 - e.g. a fit 80 kg young man may have a target workload of 320 W (4 W/kg), such that a 30W/min ramp or minute-by-minute work rate increment is appropriate, whilst a sedentary 55 kg woman may be expected to have a target workload of 110 W (2 W/kg), suggesting that a 10W ramp should be chosen. For children and adolescents, an equation predicting peak workload has been published (50).
 - Further reductions should be made in advanced lung disease e.g. FEV₁ <40% predicted
- HR and SpO₂ should be monitored throughout exercise and effort and breathlessness ratings recorded at the end of each minute and at end-exercise
- For the measurement of effort and breathless during exercise the following instruction may be useful:

“During the test, we are going to ask you about the intensity of your breathlessness and leg discomfort at that point in time. As you should avoid speaking during the test, you will use a finger of your hand to point which number between 0 and 10 [amend based on scale being used] best reflects the intensity of each of these sensations”. (49)
- Upon exhaustion, defined as the point at which the participant can no longer maintain the required cadence despite strong verbal encouragement, or that exhaustion has been reached in the view of the tester, the recovery phase should begin.
- The recovery phase should last 2-3 minutes of cycling at a slow cadence (~ 30 rpm), although a longer recovery may be needed. HR, SpO₂, effort, breathless and symptoms should continue to be monitored throughout recovery.

Outcomes

- Peak power output (W_{peak}) – average of the final 30 seconds
- Heart rate (ECG if available) at peak exercise
- Continuous SpO₂ and SpO₂ at peak exercise. If SpO₂ drops below 90%, power output and HR at this point should be recorded.
- Subjective perception of breathlessness and effort

Prediction equations

There are numerous different prediction equations for CPET outcomes available (51), but only few report normal reference equations for W_{peak} (e.g. (52, 53)). Reference equations for the Godfrey protocol are presented in Hebestreit et al. (2015) (1).



Test Report – W_{peak} Cycle Ergometer Test

Date of test: _____ (dd/mm/yyyy)

Date of birth: _____ (dd/mm/yyyy)

First name: _____

Last name: _____

Indication of test: _____

Medications: _____

Bronchodilator/time since last dose: _____

Oxygen supplementation: Yes _____ (L/min) No

Unloaded work rate (W): _____

Work rate increments used (W/min): _____

Test Results

Did the participant experience an adverse event? Yes No

Yes, please specify:

W_{peak} (W): _____ Reason for test termination: _____

Incremental phase duration (min): _____

Pre-test

Post-test

Resting heart rate: _____ bpm

Peak heart rate: _____ bpm

Resting SpO₂: _____ %

SpO₂ at peak exercise: _____ %

Comment on SpO₂ signal quality: _____

*Nadir SpO₂: _____ %

**Based on continuous SpO₂ measurement*

Resting breathlessness: _____

Peak breathlessness: _____ [state scale used]

Resting leg effort (RPE): _____

Peak leg effort: _____ [state scale used]

Test

Time (mins)	SpO ₂ (%)	HR (bpm)	Dyspnoea (scale used)	Leg effort (scale used)
Rest				
Unloaded 1				
Unloaded 2				
Unloaded 3				
1				
2				
3				
4				
5				
6				
7				
8				
9				
10				
11				
12				
13				
14				
15				

Recovery 1				
Recovery 2				
Recovery 3				
Recovery 4				
Recovery 5				

Comments



Exercise Working Group

TEST INSTRUCTIONS

6-Minute Walk Test

The 6-minute walk test (6MWT) is a functional exercise capacity test and measures the distance that the participant can walk on a flat surface in 6-minutes. The following procedures are adapted from the European Respiratory Society (ERS) and American Thoracic Society (ATS) technical standards (54).

Equipment

- Stopwatch/smartphone
- Mechanical lap counter
- Pulse oximeter to continuously measure oxygen saturation (SpO₂) and heart rate (HR)
- Two small cones to mark the turnaround points
- Scales to assess leg effort and breathlessness (e.g., Borg Scale)
- Chair(s) along the walking course for the patient to rest if required
- A source of oxygen (for recovery / emergency; O₂ tank optional during the test)
- Telephone (in case of emergency)
- Defibrillator (compulsory, depending on the testing location and conditions)

Pre-test preparations

- There is a substantial learning effect when the test is repeated. At least one familiarisation test should be performed (9, 55).
- If two tests are performed on the same day, at least 30 minutes rest should be ensured between tests. Following European Respiratory Society (ERS) and American Thoracic

Society (ATS) technical standards (9), two tests should be performed and the test with the largest walking distance should be recorded.

- Comfortable clothing should be worn with appropriate footwear.
- Usual pre-exercise bronchodilator and dose should be administered at least 10 minutes before the test.
- Participants should not exercise vigorously within 2 hours of completing the test.
- Refrain from eating for at least 1 hour before the test.
- A warm-up period before the test is not required.
- The participant should sit in a chair located at the starting position for ~10 minutes prior to the start of the test. During this time, contraindications should be checked, and pre-test measures (e.g., SpO₂, heart rate) completed.
- Measure symptoms on a standardised scale immediately prior to test commencement.

Test instructions

- The participant is provided with the following instructions before the start of the test (56):

“The object of this test is to walk as far as possible in 6 minutes. You will walk along this hallway between the markers, as many times as you can in 6 minutes. I will let you know as each minute goes past, and then at 6 minutes I will ask you to stop where you are. Six minutes is a long time to walk, so you will be exerting yourself. You may get out of breath or become tired. You are permitted to slow down or stop and rest as necessary. If you rest, please resume walking as soon as you are able. You will walk back and forth around the cones. You should pivot briskly around the cones and continue back without stopping. Remember that the object of the test is to walk as far as possible in 6 minutes, but don’t run or jog. Try not to talk during the test unless you have a problem. Do you have any questions? When you start walking, I will start the timer.”
- During the test, the participant is allowed to have a rest when needed.
- Standardised encouragement should be provided:
 - After the first minute, tell the participant: *“You are doing well. You have 5 minutes to go.”*
 - After the second minute, tell the participant: *“Keep up the good work. You have 4 minutes to go.”*

- After the third minute, tell the participant: *“You are doing well. You have 3 minutes to go.”*
- After the fourth minute, tell the participant: *“Keep up the good work. You have 2 minutes to go.”*
- After the fifth minute, tell the participant: *“You are doing well. You have 1 minutes to go.”*
- With 15 seconds to go, tell the participant: *“In a moment, I am going to tell you to stop. When I do, just stop right where you are.”*
- If the participant stops during the test, ask the participant to resume walking whenever they feel able.
- SpO₂ should be recorded continuously throughout the 6MWT (9).
- If the participant stops to rest during the test, note the time (do not stop the timer). Instruct the participant to continue walking when they feel able.
- The assessor should not pace the participant throughout the 6MWT.
- At the end of the test, record HR, SpO₂, and subjective perception of breathlessness and leg effort, symptoms and the final distance covered. Ask the participant about their limiting symptom(s).

Outcomes

- Distance (i.e., the larger distance of two tests is recorded)
- Heart rate
- SpO₂ at peak exercise and nadir if continuous measurement available
- Subjective perception of breathlessness and effort

Prediction equations

There is wide variation in the reference values used for 6MWD (9, 32).



Test Report – 6-Minute Walk Test

Date of test: _____ (dd/mm/yyyy)

Date of birth: _____ (dd/mm/yyyy)

First name: _____

Last name: _____

Indication of test: _____

Medications: _____

Bronchodilator/time since last dose: _____

Oxygen supplementation: Yes _____ (L/min) No

Corridor length: _____

Test Results

Did the participant take a break(s) during the test? Yes _____ sec No

Yes _____ sec

Yes _____ sec

Did the participant experience an adverse event? Yes No

Yes, please specify:

Number of laps: _____

Reason for test termination: _____

Total distance: _____

Pre-test

Post-test

Resting heart rate: _____ bpm

Peak heart rate: _____ bpm

Resting SpO₂: _____ %

SpO₂ at peak exercise: _____ %

Comment on SpO₂ signal quality: _____

*Nadir SpO₂: _____ %

**Based on continuous SpO₂ measurement*

Resting breathlessness: _____

Peak breathlessness: _____ [state scale used]

Resting leg effort (RPE): _____

Peak leg effort: _____ [state scale used]

Test 1

Time (mins)	SpO ₂ (%)	HR (bpm)	Dyspnoea (scale used)	Leg effort (scale used)
Rest				
1				
2				
3				
4				
5				
6				
Recovery 1				
Recovery 2				
Recovery 3				

Test 2

Time (mins)	SpO ₂ (%)	HR (bpm)	Dyspnoea (scale used)	Leg effort (scale used)
Rest				
1				
2				
3				
4				
5				
6				
Recovery 1				
Recovery 2				
Recovery 3				

Comments <hr/> <hr/> <hr/> <hr/>
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TEST INSTRUCTIONS

10 m Modified Shuttle Test

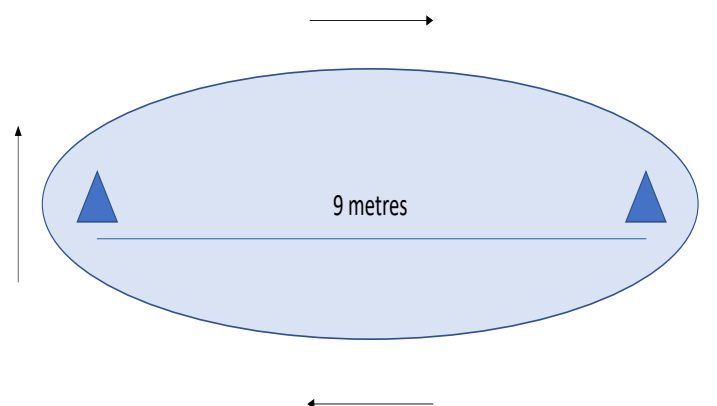
The 10 m Modified Shuttle Test (10 m MST) is an incremental exercise capacity test and measures the distance that the participant can walk or run on a flat surface. The 10 m MST is available in 15- and 25-level versions where the speed is controlled by a series of pre-recorded signals. Speed increases until the participant can no longer continue. The following procedures are adapted from the European Respiratory Society (ERS) and American Thoracic Society (ATS) technical standards.

Equipment

- Tape measure
- 10 m markers
- Test scoring sheet
- Stopwatch/smartphone
- Pulse oximeter to measure oxygen saturation (SpO₂) and heart rate
- Scales to assess leg effort and dyspnoea (e.g., Borg Scale, Dalhousie Scale)
- Telephone (in case of emergency)
- Defibrillator (compulsory, depending on the testing location and conditions)



- The MST takes approximately 30 minutes to complete (including preparation and participant recovery time). In centres where this test is not established and/or where the participant is naïve to the test, a practice test is likely to be necessary, and thus the time required for testing, at least initially, may be considerably longer.
- Only standardised instructions from the CD should be used. In contrast to the six-minute walking test, no encouragement should be given throughout the MST.
- A comfortable ambient temperature and humidity should be maintained for all tests.
- The testing track must be the same for all tests for a participant:
 - Cones are placed nine metres apart.
 - The distance covered around the cones is 10 m.



Pre-test preparations

- Instruct the participant to dress comfortably and to wear appropriate footwear.
- Any prescribed inhaled bronchodilator medication should be taken within one hour of testing or when the participant arrives for testing.
- The participant should rest for at least 15 minutes before beginning the MST.
- Record:
 - Blood pressure
 - Heart rate
 - SpO₂Dyspnoea score*
** Note: Show the patient the dyspnoea scale and give standardised instructions on how to obtain a score*

During the test

The participant is given the following instructions

- *The object of the progressive shuttle test is to walk or run as necessary, for as long as possible, there and back along the 10 m course, keeping to the speed indicated by the beeps on the audio recording.*
- *You will hear these beeps at regular intervals.*
- *You should walk or run at a steady pace, aiming to turn around the cone at one end of the course when you hear the first beep, and at the other end when you hear the next.*
- *The speed for the first minute is very slow. You have 20 seconds to complete each 10 m shuttle, so don't go too fast.*
- *At first you will be able to walk, but as the beeps get closer together you will need to speed up at the end of each minute.*
- *Your aim should be to follow the set rhythm for as long as you can.*
- *Each single beep signals the end of a shuttle and each triple beep signals an increase in speed.*
- *You should stop the test only when you become too breathless to maintain the required speed or can no longer keep up with the set pace, e.g. you can't run any faster.*
- *The test is progressive. In other words, it is easier at the start and harder at the end.*
- *The test will start in 15 seconds, so get ready at the start now. Level one starts with a triple beep after the 4 second countdown.*

Each minute, record heart rate, SpO₂ and dyspnoea, as well as number of shuttle that is completed as on the recording sheet.

Monitor the participant for untoward signs and symptoms.

Ending the test

The test ends if any one of the following occur:

- The participant is **more than** 0.5 m away from the cone when the beep sounds (allow one lap to catch up).
- The participant reports that they are too breathless to continue.
- The participant exhibits any of the following signs and symptoms:
 - Chest pain that is suspicious of / for angina.
 - Evolving mental confusion or lack of coordination.
 - Evolving light-headedness.
 - Intolerable dyspnoea.
 - Leg cramps or extreme leg muscle effort
 - Any other clinically warranted reason.

At the end of the test

- Seat the participant or, if the participant prefers, allow to them to stand.
- Immediately record oxygen saturation (SpO₂), heart rate, dyspnoea and leg effort ratings.
- Two minutes later, record SpO₂ and heart rate to assess the recovery rate.
- Record the total number of shuttles.
- Record the reason for terminating the test. The participant can be asked: “*What do you think stopped you from keeping up with the beeps?*”
- The participant should remain in a clinical area for at least 15 minutes following an uncomplicated test.

Learning effects

There is a small but statistically significant difference between two incremental shuttle tests performed (mean differences of 20 m and 25 m for tests performed on the same day, and 23.5 m for tests performed on different days) (57, 58). This learning effect is considered to be clinically important when evaluating change over time. However, conducting 2 shuttle tests to maximal volitional capacity would be challenging for the participant and any repeat tests are usually done to evaluate the benefits of an intervention such as a course of treatment or the effectiveness of a training program.

Reference values

Reference equations to predict distance achieved on the MST-15 are available – see below. There are, as yet, no reference equation data for predicting performance on the MST-25.

$$\text{Predicted distance on MST} = 1449.701 - (11.735 \times \text{age}) + (241.897 \times \text{gender}) - (5.686 \times \text{BMI})$$

Where male gender = 1, female gender = 0.

(59-61)

The table below provides information on levels and distances covered within MST-25 and MST-25 protocols:

Metrics of the 15-level and 25-level Modified Shuttle Tests:

Level	Shuttle time (s)	Total distance covered (m)
1	20.00	30
2	15.00	70
3	12.00	120
4	10.00	180
5	8.57	250
6	7.50	330
7	6.67	420
8	6.00	520
9	5.46	630
10	5.00	750
11	4.62	880
12	4.29	1,020
13	4.00	1,170
14	3.75	1,330
15	3.53	1,500
16	3.33	1,680
17	3.16	1,870
18	3.00	2,070
19	2.86	2,280
20	2.73	2,500
21	2.61	2,730
22	2.50	2,970
23	2.40	3,200
24	2.31	3,480
25	2.22	3,750

Test Report – 10 m Modified Shuttle Test



Date of test: _____ (dd/mm/yyyy) Date of birth: _____
(dd/mm/yyyy)

First name: _____ Last name: _____

Indication of test: _____

Oxygen supplementation: Yes _____ (L) No

Test Results

Did the participant take a break during the test? Yes _____ sec No

Did the participant experience an adverse event? Yes No

Yes, please specify:

Number of completed shuttles: _____ Reason for test termination: _____

Pre-test

Post-test

Resting heart rate: _____ bpm

Peak heart rate: _____ bpm

Resting SpO₂: _____ %

SpO₂ at peak exercise: _____ %

Comment on SpO₂ signal quality: _____

*Nadir SpO₂: _____ %

**Based on continuous SpO₂ measurement*

Resting breathlessness: _____

Peak breathlessness: _____ [state scale used]

Resting leg effort (RPE): _____

Peak leg effort: _____ [state scale used]

Comments

Level	Shuttle time	Shuttle number																				Distance
19	2.86	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	2280
		21																				
20	2.73	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	2500
		21	22																			
21	2.61	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	2730
		21	22	23																		
22	2.50	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	2970
		21	22	23	24																	
23	2.40	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	3200
		21	22	23	24	25																
24	2.31	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	3480
		21	22	23	24	25	26															
25	2.22	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	3750
		21	22	23	24	25	26	27														

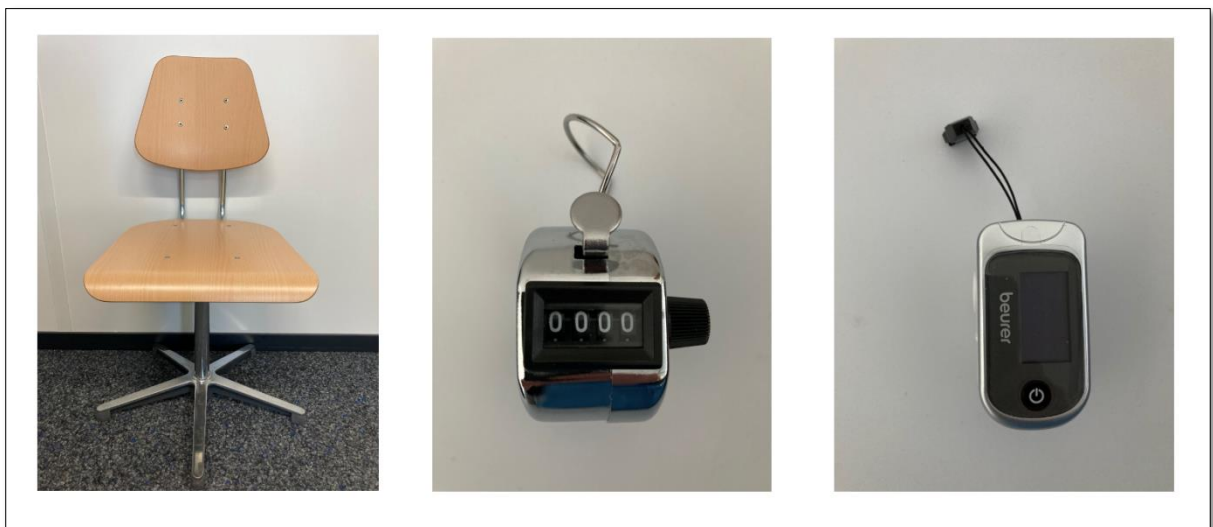
TEST INSTRUCTIONS

1-minute Sit-to-Stand Test

The 1-minute sit-to-stand test (1-min STS) is a functional exercise capacity test and measures the maximum number of sit-to-stand repetitions that a person can perform for one minute.

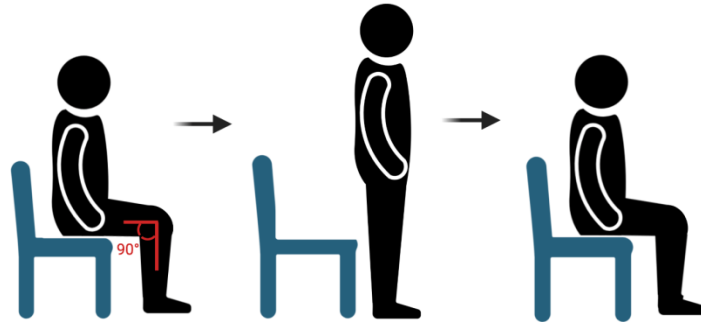
Equipment

- Height adjustable chair
- Tape measure
- Stopwatch/Smartphone
- Counter (compulsory)
- Pulse oximeter to measure oxygen saturation (SpO₂) and heart rate
- Scales to assess effort and breathlessness (e.g., Borg Scale, Dalhousie Scale)
- Telephone (in case of emergency)
- Defibrillator (compulsory, depending on the testing location and conditions)



Pre-test preparations

- Place the chair against the wall for safety reasons.
- Adjust the seat height to have approximately 90° knee angle flexion (see picture). Do not test young children with a standard chair (46-48 cm). Where possible, an adjustable chair that allows for 90° knee angle flexion should be used.



Test instructions

- Start position: Participants sitting on a chair.
- The participant is provided with the following instructions: *"The objective of this tests is to stand up and sit down on a chair as often as possible in one minute. Put your legs hip-width apart and your feet flat on the floor under your knees so that there is an angle of about 90 degrees. Place your hands on your hips. Now try to stand up and sit down as many times as possible within one minute. Do not use your hands or arms to assist movement. When sitting down you must touch the chair with your buttocks and when standing up your knees must be fully straight. You may get out of breath or become tired. You are permitted to slow down or stop and rest as necessary. After "Ready, set, go" you can start. I will tell you when you have 15 seconds left. If I speak to you and correct you, don't get upset, just continue the test. Optional: I will also ask you before and after the test about your perceived breathlessness and effort."*
- Standardised instructions should be provided:
 - After 45 seconds, tell the participant: *"15 seconds left until the test is over."*
- Do not verbally encourage the participant during the test

Outcomes

- Number of repetitions
- Heart rate
- Oxygen saturation
- Subjective perception of breathlessness and effort
- STS Power Index (optional, see (42))

References values

Age group	Female					Male				
	Percentiles					Percentiles				
Years	p2.5%	p25%	p50%	p75%	p97.5%	p2.5%	p25%	p50%	p75%	p97.5%
5-7	39	52	56	63	72	42	52	60	68	79
8-10	41	58	64	68	77	41	55	61	66	77
11-13	44	53	61	67	80	43	57	61	66	75
14-16	35	42	53	60	76	38	49	55	60	72
---	---	---	---	---	---	---	---	---	---	---
20-24	31	39	47	55	70	27	41	50	57	72
25-29	30	40	47	54	68	29	40	48	56	74
30-34	27	37	45	51	68	28	40	47	56	72
35-39	25	37	42	50	63	27	38	47	58	72
40-44	26	35	41	48	65	25	37	45	53	69
45-49	25	35	41	50	63	25	35	44	52	70
50-54	23	33	39	47	60	24	35	42	53	67
55-59	21	30	36	43	61	22	33	41	48	63
60-64	20	28	34	40	55	20	31	37	46	63
65-69	19	27	33	40	53	20	29	35	44	60
70-74	17	25	30	36	51	19	27	32	40	59
75-79	13	22	27	30	43	16	25	30	37	56

Note: Reference values for adults ($n = 6,929$, 20-79 years of age) are based on the publication by (62)). Reference values for children and adolescents ($n = 547$, 5-16 years of age) are based on the publication by (63)).

Learning effects

There is a substantial learning effect when the test is repeated (24, 41, 63, 64). It is important that the person to be tested performs at least on practice test to have an accurate assessment of

functional exercise capacity. If two tests are performed on the same day, consider a rest of about 30 minutes between the first and second test to allow for sufficient recovery.

Minimal important difference

The minimal important difference, i.e., the smallest difference in 1-min STS that is perceived as beneficial by adults with CF is 5 repetitions. These data are based on a small study with 16 individuals with CF and should be confirmed by larger studies in the era of new modulator therapy. Data for children are not yet available.

Practical tips

- Typically, the lowest SpO₂ value that the participant drops to (i.e. nadir value) is observed within the first minute of recovery (42, 65). We therefore suggest not to use SpO₂ recordings immediately post-exercise, i.e., wait until a stable signal is achieved and record the lowest SpO₂.

Test Report – 1-Minute Sit-to-Stand Test



Date of test: _____ (dd/mm/yyyy) Date of birth: _____ (dd/mm/yyyy)
First name: _____ Last name: _____
Indication of test: _____
Oxygen supplementation: Yes _____ (L) No
Chair height: _____ cm Leg length (optional): _____ cm

Test results

Did the participant take a break during the test? Yes _____ sec No
Did the participant experience an adverse event? Yes No
Yes, please specify:

Number of STS repetitions: _____
Reason for test termination: _____

Pre-test

Post-test

Resting heart rate: _____ bpm	Peak heart rate: _____ bpm
Resting SpO ₂ : _____ %	SpO ₂ at peak exercise: _____ %
Comment on SpO ₂ signal quality: _____	
	*Nadir SpO ₂ : _____ %
	<i>*Based on continuous SpO₂ measurement</i>
Resting breathlessness: _____	Peak breathlessness: _____ [state scale used]
Resting leg effort (RPE): _____	Peak leg effort: _____ [state scale used]

Comments:



TEST INSTRUCTIONS

Quadriceps muscle strength testing

Quadriceps muscle strength testing involves measuring the maximal voluntary force produced by the knee extensors under isometric modality.

Equipment

- Medical chair with back rest allowing variable settings for inclination
- Seat belt fixed to the chair
- Strain gauge dynamometer (measurement range from 0 to, at least, 1,000 N)
- Inextensible ankle strap
- Amplifier and dedicated software for biomedical signal processing
- Computer (for visual feedback and storing the data)
- Tape measure for measuring the lever arm

Pre-test preparations

- Install the person on the chair and adjust the settings to have both knee and hip of the dominant leg at 90° angle flexion.
- Attach the leg to the force sensor ~1-2 cm above the malleoli of the ankle joint.
- Measure the distance between the knee joint center of rotation and the point of force application (lever arm).
- Secure the person to the chair with the seat belt placed across the waist.

Test instructions

- The aim of this test is to perform maximal voluntary contractions (MVC) of the knee extensors and determine the peak force.

- Provide instructions to the participant as follows:

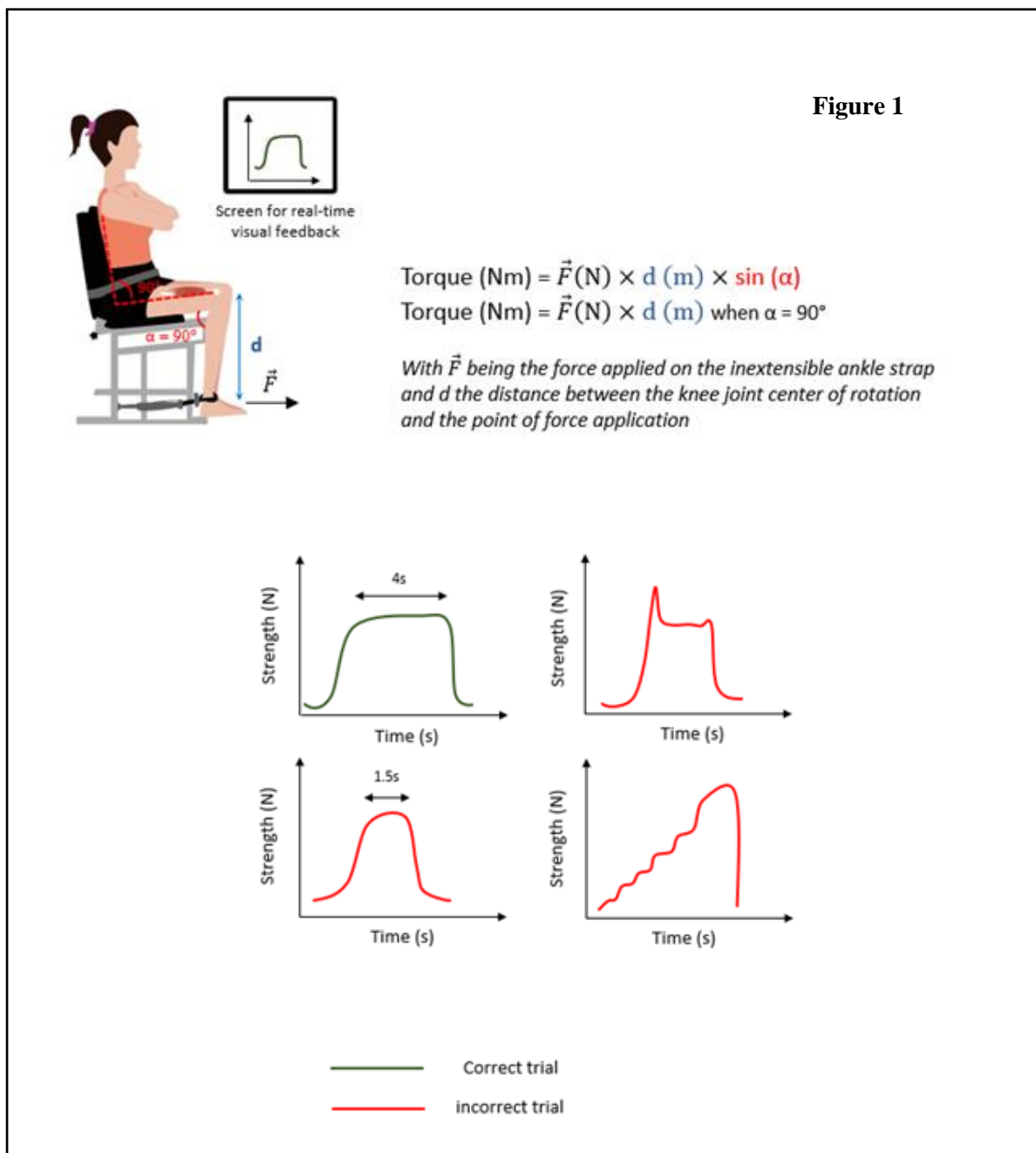
“The aim of this test is to measure the strength of your thigh. The movement is a leg extension against a fixed resistance. It is important to push as hard and fast as possible and to maintain your maximal effort during the whole procedure. Using a countdown, I will tell you the moment at which you have to push. I will encourage you during the entire duration of the contraction and then indicate when you can relax. You will repeat this procedure several times. During each trial, it is important that you keep your arm crossed on your abdomen. Remember that each trial requires a maximal effort. Do you have any questions? [...] We will begin with a short warm-up”.
- Specific warm-up should include submaximal intermittent isometric contractions (e.g. 5 second contraction, 5 second relaxation) at increasing force levels (i.e. without exceeding ~ 40% of maximal perceived effort to avoid muscle fatigue) for ~ 3 minutes. This warm-up also allows the participant to be familiarised with the visual feedback they will receive during the test.
- Start with two initial MVC (not considered for analysis) permitting to familiarise the participant with maximal manoeuvres and check the stability of the measurement chain.
- Perform at least three attempts with the aim to obtain three reproducible MVC varying less than 5% (which is usually obtained in less than 6 trials) and keep the highest peak value among these three attempts for analysis.
- It is important for each MVC to obtain a force plateau of at least 3 seconds (see Figure 1).
- The total contraction time should range between 4-6 seconds.
- A 60 second recovery period should be observed between each trial.
- Vigorous encouragements are necessary, using the same frequency and intensity during each trial, using a repetition of single words like “go, go, go...” or “push, push, push...”

Outcomes

- Maximal voluntary contraction, expressed in Newton (N) and Newton meters (Nm)

Practical tips

- It is important to ensure that the appropriate posture is maintained by the participant throughout each attempt.
- Classic compensatory movements include movements of the arms and/or progressive rise of the buttocks from the chair. This usually results in increasing force level throughout the 4-6 second duration of the MVC (see Figure 1), so the participant should be informed that they must stay on the chair at all times.



Test Report – Quadriceps muscle strength testing



Date of test: _____ (dd/mm/yyyy) Date of birth: _____ (dd/mm/yyyy)

First name: _____ Last name: _____

Indication of test: _____

Lever arm: _____ cm

Test Results

Did the participant experience an adverse event? Yes No

Yes, please specify:

Peak Force (N): _____ Peak Force (Nm): _____

Comments

ONLINE SUPPLEMENTARY MATERIAL (ECFS survey of Exercise Practitioners)

Survey summary:

Methods: A group from the European CF Society Exercise Working Group (ECFS-EWG), including members from the Physiotherapy Specialist International Interest Group (PhySIG), aimed to develop guidance and standard operating procedures for an agreed selection of exercise tests, that allow for the comprehensive functional evaluation of people with CF. To facilitate test selection (considerations e.g. most-used, best-validated), an international panel ($n=64$; 81.3% Europe), comprising physicians (5%), exercise scientists (14%), physiotherapists (78%), physiotherapy assistants and fitness instructors (3%), were consulted to obtain a snapshot of current practice versus the evidence base. Statements summarising our recommendations developed within this article were then presented to members of the wider ECFS-EWG at the January 2023 bi-annual meeting with agreement/disagreement obtained. Further stakeholder feedback on the test instructions and reporting templates was provided by physiotherapy experts from CFPhysio.com. A bias towards physiotherapists working with pwCF is noted.

Table S1. Participant demographics.

Variable	n (%) = 64 (100.0)	
Expert group	Physician ($n = 3$ [4.7]) Exercise scientist / clinical exercise physiology ($n = 9$ [14.1])	Physiotherapist ($n = 50$ [78.1]) Physiotherapy technician / assistant / fitness instructor ($n = 2$ [3.1])
Geographical location	Europe ($n = 52$ [81.3])	Outside of Europe ($n = 12$ [18.8])
Experience working with people with CF (years)	< 1 ($n = 1$ [1.6]) 3-5 ($n = 7$ [10.9])	1-3 ($n = 3$ [4.7]) > 5 ($n = 53$ [82.2])

Note: CF, cystic fibrosis

Results: In the last two years, 90.6% of respondents reported using exercise tests, the most common of which were the 6-minute walk test (6MWT; 79.4%), cycle ergometer cardiopulmonary exercise testing with gas analysis (CPET, 52.4%), the 1-minute sit-to-stand test (1minSTS; 39.7%), cycle ergometer tests without gas analysis (30.2%) and the 15-level 10-m modified shuttle test (28.6%). When asked to rank exercise tests (excluding the accepted gold standard CPET) based on perceptions of those most rigorously validated, the top 5 were: 1) cycle ergometer tests without gas analysis, 2) treadmill tests without gas analysis, 3) 6MWT, 4) 1minSTS and 5) the A-step test.

Table S2. Perceived usefulness of exercise tests available for use with people with cystic fibrosis.

Exercise test	Used in clinical practice past 2 years (n [%])	Ranked usefulness of test					
		Not at all useful	Slightly useful	Somewhat useful	Very useful	Extremely useful	Not able to judge (no experience with test)
CPET (with gas analysis) – cycle ergometer	33 (52.4)	0	1 (1.6)	3 (4.7)	10 (15.6)	34 (53.1)	16 (25.0)
CPET (with gas analysis) – treadmill	10 (15.9)	0	0	2 (3.1)	9 (14.1)	20 (31.3)	33 (51.6)
Cycle ergometer test without gas analysis	19 (30.2)	0	2 (3.1)	10 (15.6)	21 (32.8)	2 (3.1)	29 (45.3)
Treadmill test without gas analysis	14 (22.2)	0	1 (1.6)	13 (20.3)	14 (21.9)	1 (1.6)	35 (54.7)
1-minute sit-to-stand test	25 (39.7)	3 (4.7)	6 (9.4)	19 (29.7)	12 (18.8)	3 (4.7)	21 (32.8)
Modified shuttle test 15-level (10 m)	18 (28.6)	2 (3.1)	2 (3.1)	24 (37.5)	6 (9.4)	3 (4.7)	27 (42.2)
Modified shuttle test 25-level (10 m)	12 (19.0)	2 (3.1)	2 (3.1)	13 (20.3)	10 (15.6)	3 (4.7)	34 (53.1)
20 m shuttle test	1 (1.6)	2 (3.1)	1 (1.6)	9 (14.1)	4 (6.3)	1 (1.6)	47 (73.4)
6-minute walk test	50 (79.4)	2 (3.1)	7 (10.9)	22 (34.4)	19 (29.7)	7 (10.9)	7 (10.9)
12-minute walk test	0 (0.0)	3 (4.7)	1 (1.6)	4 (6.3)	5 (7.8)	0	51 (79.7)
3-minute step test	11 (17.5)	4 (6.3)	4 (6.3)	10 (15.6)	9 (14.1)	2 (3.1)	35 (54.7)
Alfred step test (A-step)	5 (7.9)	3 (4.7)	1 (1.6)	3 (4.7)	4 (6.3)	4 (6.3)	49 (76.6)
Handgrip strength test	17 (27.0)	1 (1.6)	7 (10.9)	12 (18.8)	7 (10.9)	2 (3.1)	35 (54.7)
Quadriceps strength test	6 (9.5)	1 (1.6)	1 (1.6)	8 (12.5)	8 (12.5)	3 (4.7)	43 (67.2)
Other	11 (17.5)	-	-	-	-	-	-

Note: CPET, cardiopulmonary exercise testing. Data are presented as n (%).

Table S3. Groups for whom exercise testing can be used in clinical practice.

Exercise test	Group						
	Children	Adults	Mild lung disease	Moderate lung disease	Severe lung disease	All groups listed	Not able to judge (no experience with test)
CPET (with gas analysis) – cycle ergometer	20 (31.3)	23 (35.9)	20 (31.3)	19 (29.7)	10 (15.6)	29 (45.3)	15 (23.4)
CPET (with gas analysis) – treadmill	12 (18.8)	17 (26.6)	12 (18.8)	15 (23.4)	4 (6.3)	13 (20.3)	33 (51.6)
Cycle ergometer test without gas analysis	11 (17.2)	13 (20.3)	10 (15.6)	11 (17.2)	4 (6.3)	24 (37.5)	26 (40.6)
Treadmill test without gas analysis	12 (18.8)	15 (23.4)	11 (17.2)	12 (18.8)	1 (1.6)	17 (26.6)	30 (46.9)
1-minute sit-to-stand test	14 (21.9)	26 (40.6)	9 (14.1)	18 (28.1)	27 (42.2)	13 (20.3)	22 (34.4)
Modified shuttle test 15-level (10 m)	23 (35.9)	25 (39.1)	15 (23.4)	23 (35.9)	13 (20.3)	11 (17.2)	27 (42.2)
Modified shuttle test 25-level (10 m)	22 (34.4)	23 (35.9)	19 (29.7)	17 (26.6)	5 (7.8)	8 (12.5)	31 (48.4)
20 m shuttle test	10 (15.6)	13 (20.3)	10 (15.6)	9 (14.1)	1 (1.6)	7 (10.9)	44 (68.8)
6-minute walk test	18 (28.1)	30 (46.9)	6 (9.4)	16 (25.0)	35 (54.7)	14 (21.9)	8 (12.5)
12-minute walk test	6 (9.4)	10 (15.6)	7 (10.9)	9 (14.1)	4 (6.3)	1 (1.6)	48 (75.0)
3-minute step test	12 (18.8)	19 (29.7)	11 (17.2)	17 (26.6)	11 (17.2)	5 (7.8)	33 (51.6)
Alfred step test (A-step)	5 (7.8)	8 (12.5)	6 (9.4)	6 (9.4)	4 (6.3)	5 (7.8)	49 (76.6)
Handgrip strength test	7 (10.9)	15 (23.4)	14 (21.9)	13 (20.3)	15 (23.4)	14 (21.9)	34 (53.1)
Quadriiceps strength test	7 (10.9)	10 (15.6)	9 (14.1)	9 (14.1)	10 (15.6)	11 (17.2)	42 (65.6)

Note: CPET, cardiopulmonary exercise testing. Data are presented as *n* (%).

Table S4. Perception of clinimetric properties of exercise tests available for use with people with cystic fibrosis.

Exercise test	Perception					
	Very poor	Poor	Acceptable	Good	Very good	Not able to judge (<i>no experience with test</i>)
CPET (with gas analysis) – cycle ergometer	0	0	0	5 (7.8)	45 (70.3)	14 (21.9)
CPET (with gas analysis) – treadmill	0	0	0	7 (10.9)	28 (43.8)	29 (45.3)
Cycle ergometer test without gas analysis	0	2 (3.1)	9 (14.1)	16 (25.0)	11 (17.2)	26 (40.6)
Treadmill test without gas analysis	0	2 (3.1)	10 (15.6)	17 (26.6)	6 (9.4)	29 (45.3)
1-minute sit-to-stand test	0	2 (3.1)	23 (35.9)	14 (21.9)	2 (3.1)	23 (35.9)
Modified shuttle test 15-level (10 m)	0	1 (1.6)	16 (25.0)	16 (25.0)	4 (6.3)	27 (42.2)
Modified shuttle test 25-level (10 m)	0	2 (3.1)	15 (23.4)	10 (15.6)	5 (7.8)	32 (50.0)
20 m shuttle test	0	3 (4.7)	8 (12.5)	4 (6.3)	2 (3.1)	47 (73.4)
6-minute walk test	1 (1.6)	4 (6.3)	25 (39.1)	15 (23.4)	10 (15.6)	9 (14.1)
12-minute walk test	1 (1.6)	3 (4.7)	5 (7.8)	6 (9.4)	2 (3.1)	47 (73.4)
3-minute step test	2 (3.1)	5 (7.8)	10 (15.6)	9 (14.1)	5 (7.8)	33 (51.6)
Alfred step test (A-step)	1 (1.6)	4 (6.3)	1 (1.6)	8 (12.5)	2 (3.1)	48 (75.0)
Handgrip strength test	0	2 (3.1)	11 (17.2)	10 (15.6)	6 (9.4)	35 (54.7)
Quadriceps strength test	0	2 (3.1)	9 (14.1)	5 (7.8)	4 (6.3)	44 (68.8)

Note: CPET, cardiopulmonary exercise testing. Data are presented as *n* (%).

Table S5. Perceived rankings of the validation of exercise tests available for use in people with cystic fibrosis.

Exercise test	Rank				
	1	2	3	4	5
Cycle ergometer test without gas analysis	33 (62.3)	9 (17)	3 (5.7)	3 (5.7)	5 (9.4)
Treadmill test without gas analysis	10 (25)	16 (40)	5 (12.5)	4 (10)	5 (12.5)
1-minute sit-to-stand test	2 (5.3)	6 (15.8)	11 (28.9)	15 (39.5)	4 (10.5)
Modified shuttle test 15-level (10 m)	3 (7.5)	12 (30)	9 (22.5)	10 (25)	6 (15)
Modified shuttle test 25-level (10 m)	7 (18.9)	10 (27)	9 (24.3)	4 (10.8)	7 (18.9)
20 m shuttle test	1 (7.1)	2 (14.3)	8 (57.1)	2 (14.3)	1 (7.1)
6-minute walk test	9 (16.1)	14 (25)	16 (28.6)	9 (16.1)	8 (14.3)
12-minute walk test	1 (7.1)	2 (14.3)	4 (28.6)	5 (35.7)	2 (14.3)
3-minute step test	0	3 (12.5)	11 (45.8)	8 (33.3)	2 (8.3)
Alfred step test (A-step)	1 (4)	3 (12)	9 (36)	3 (12)	9 (36)
Handgrip strength test	0	5 (20)	7 (28)	7 (28)	6 (24)
Quadriceps strength test	2 (10)	4 (20)	7 (35)	5 (25)	2 (10)

Note: CPET, cardiopulmonary exercise testing. Data are presented as *n* (%).

Other tests currently being used in clinical practice included:

- Canadian Agility and Movement Scale Assessment
- Submaximal tests, Wingate testing
- ½ Bruce treadmill test
- Inpatient exercise testing (1-15-minute walking test) for adults with CF at a comfortable pace for the individual, for as long as they can as comfortably continue, possibly depending on how unwell they are during an acute exacerbation following admission to hospital. The aim of this informal home-grown test is to determine whether they desaturate during walking and require oxygen therapy.
- Biodex isokinetic strength testing

Consensus agreement was sought from members of the ECFS Exercise Working Group at the January meeting. Members were asked to comment if they did not agree with the summary statements presented in this paper or to rate their agreement on a Likert scale from 1-5. The mean response to all statements presented was 4/5.

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