cmtpediatricscale

Charcot-Marie-Tooth disease Pediatric Scale equipment and training resource kit

Updated June 2012

Based on the publication: Burns J. Ouvrier R. Estilow T. Shy R. Laurá M. Pallant J. Lek M. Muntoni F. Reilly M. Pareyson D. Acsadi G. Shy M. Finkel R. Validation of the CMT Pediatric Scale as an outcome measure of disability. *Annals of Neurology* 71(5), 642–652, 2012.

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Contents

Final version of the 11-item CMTPedS data form

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Initial Evaluation ❑ Re-Evaluation ❑ Date:

Patient Profile					
ID:		D.O.B:	Age (yrs):	Gender: Boy ❑ Girl ❑	
Height (m):	Weight (kg):	Dominant Hand: L ❑ R ❑	Dominant Foot: L ❑ R ❑	Diagnosis:	
Symptoms:	Foot pain ❑	Leg cramps ❑	Unsteady ankles ❑	Daily trips and/or falls ❑	Hand pain ❑
	Hand weakness ❑	Hand tremor ❑	Sensory symptoms (e.g. pins and needles, tingling, numbness, prickling) ❑		

Lunge test (degrees)		Left:	Right:
Foot Posture Index	Talar head palpation		
	Curves above and below lateral malleolus		
	Inversion/eversion of the calcaneus		
	Bulge in the region of the talonavicular joint		
	Congruence of the medial longitudinal arch		
	Abd/adduction of forefoot on rearfoot (too-many-toes)		
	Total (-12 to 12)		

Hand Dexterity			
1. Functional Dexterity Test (sec)		**2. Nine-hole peg test (sec)**	

Strength	**Trial 1**	**Trial 2**	**Trial 3**	**Average**
3. Hand grip (*N*)				x2:
4. Foot plantarflexion (*N*)				
5. Foot dorsiflexion (*N*)				

Sensation	**0**	**1**	**2**	**3**	**4**	**Score**
6. Pinprick	Normal	Decreased below or at ankle bones	Decreased at or below midline of calf	Decreased above calf midline up to and including knee	Decreased above knee (above top of patella)	
7. Vibration	Normal	Reduced at first metatarsal bone	Reduced at ankle	Reduced at knee (tibial tuberosity)	Absent at knee and ankle	

Balance	**Assistive device required** (e.g. AFO) Y/N. Describe device and footwear:									
8. Bruininks Oseretsky Test	Raw Score		*Conduct second trial only if examinee does not earn the maximum score on the first trial*							Point score
	Trial 1	**Trial 2**								
Standing with feet apart on a line-eyes open			**Raw**	**0.0-0.9**	**1.0-2.9**	**3.0-5.9**	**6.0-9.9**	**10**		
			Point	0	1	2	3	4		
Walking forward on a line			**Raw**	**0**	**1-2**	**3-4**	**5**	**6**		
			Point	0	1	2	3	4		
Standing on one leg on a line-eyes open			**Raw**	**0.0-0.9**	**1.0-2.9**	**3.0-5.9**	**6.0-9.9**	**10**		
			Point	0	1	2	3	4		
Standing with feet apart on a line-eyes closed			**Raw**	**0.0-0.9**	**1.0-2.9**	**3.0-5.9**	**6.0-9.9**	**10**		
			Point	0	1	2	3	4		
Walking forward heel-to-toe on a line			**Raw**	**0**	**1-2**	**3-4**	**5**	**6**		
			Point	0	1	2	3	4		
Standing on one leg on a line-eyes closed			**Raw**	**0.0-0.9**	**1.0-2.9**	**3.0-5.9**	**6.0-9.9**	**10**		
			Point	0	1	2	3	4		
Standing on one leg on a beam-eyes open			**Raw**	**0.0-0.9**	**1.0-2.9**	**3.0-5.9**	**6.0-9.9**	**10**		
			Point	0	1	2	3	4		
Standing heel-to-toe on a balance beam			**Raw**	**0.0-0.9**	**1.0-2.9**	**3.0-5.9**	**6.0-9.9**	**10**		
			Point	0	1	2	3	4		
Standing on one leg on a beam-eyes closed			**Raw**	**0.0-0.9**	**1.0-2.9**	**3.0-4.9**	**5.0-7.9**	**8.0-9.9**	**10**	
			Point	0	1	2	3	4	5	
Balance Subscale from the *Bruininks Oseretsky Test of Motor Proficiency, Second Edition (BOT-2)*. Copyright© 2005 NCS Pearson, Inc. Adapted and reproduced with permission. All rights reserved									**Total**	

Motor Function	**Assistive device required** (e.g. AFO) Y/N. Describe device and footwear:		
9. Gait	Foot drop: No❑ Some❑ Yes❑	Difficulty heel walking: No❑ Some❑ Yes❑	Difficulty toe walking: No❑ Some❑ Yes❑
10. Long jump (cm)		**11. Six-minute walk test (m)**	

Item Scores (0-4)											**Total Score (0-44)**
1.	2.	3.	4.	5.	6.	7.	8.	9.	10.	11.	

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List of Equipment

Item	Approximate Cost (USD$)
1. Functional Dexterity Test:	
Functional Dexterity Test (FDT): North Coast Medical: www.ncmedical.com	$90
Digital stopwatch (included with 9-Hole Peg Test Kit)	
2. Nine Hole Peg Test:	
Rolyan® 9-Hole Peg Test Kit and Digital stopwatch	$80
Sammons Preston, Bolingbrook, IL, USA: www.pattersonmedical.com	
3. Grip strength:	
Citec hand-hand dynamometer with grip strength applicator, spare batteries (6V)	$1900
C.I.T. Technics, Haren, The Netherlands: www.citec.nu	
*CMTPedS Users receive a 10% discount until 31.12.12. Contact: info@citec.nu	
4. Plantarflexion strength:	
Citec hand-hand dynamometer incl. Grip Strength Applicator, spare batteries (6V)	as above
5. Dorsiflexion strength:	
Citec hand-hand dynamometer incl. Grip Strength Applicator, spare batteries (6V)	as above
6. Pinprick: Neurotips™ (100): Owen Mumford Ltd, Oxford, UK: www.owenmumford.com	$20
7. Vibration:	
Rydel Seiffer tuning fork, C 64 Hz / c 128 Hz detachable clamps	
Arno Barthelmes & Co. GmbH - Tuttlingen – Germany: www.barthelmes.info	$150
8. Balance	
Bruininks-Oseretsky Test of Motor Proficiency, 2nd Ed (BOT-2): Balance Beam with Bag	$197
NCS Pearson, Upper Saddle River, NJ, USA: www.pearsonassessments.com + stopwatch	
9. Gait (not applicable)	
10. Long Jump: Tape measure (150 cm)	$10
11. Six-minute walk test: Two small markers, lap counter, stopwatch, tape measure (30 m)	$60
Other equipment for patient profiling:	
Foot Posture Index: Score sheet and manual: www.leeds.ac.uk/medicine/FASTER/fpi.htm	Free
Lunge test: Baseline Digital Inclinometer: http://www.bpp2.com/	$112

Item Instructions

1. Functional Dexterity Test

Background/Purpose: The Functional Dexterity Test (FDT) is a measure of hand dexterity that provides information regarding the use of the fingers and hand for daily tasks requiring 3-jaw chuck prehensions e.g. buttoning, tying shoe laces, screwing a nut and bolt. The FDT has demonstrated good validity and reliability in healthy controls[1,2] and adults with CMT.[3] It has been correlated to light touch-pressure sensation as tested via the Semmes Weinstein monofilament.[4]

Test Position: The child is asked to sit on a chair with the FDT pegboard on a table in front of him/her. The proximal edge of the pegboard is placed 10 cm from the edge of the table.

Testing Procedures: The clinical evaluator explains to each child that the purpose of the test is to turn over the pegs as fast as possible with their dominant hand. The child is instructed to start at the top at the opposite side of the board to the hand and continue in a zigzag manner (left to right and right to left, or vice versa).

The evaluator says,

> "I don't want you to turn your hand towards the ceiling (supinate), touch the board for help in turning the peg, touch the peg with your other (free) hand, allow the peg to touch your chest, or drop a peg. Here, I want you to watch me and then you can practice."

The clinical evaluator demonstrates by turning over 4 pegs. The child is given a practice trial of turning over 4 pegs with the dominant hand first.

After the practice trial the clinical evaluator says:

> "I want you to turn over the pegs as fast as you can. Are you ready? Go."

The evaluator starts the stopwatch at the word 'go' and stops the stopwatch when the child releases the last peg. The time is recorded in seconds.

For those unable to perform the FDT due to CMT severity or are very slow, give a score of **150 seconds** which is greater than the maximum childhood CMT value.

Note: Adult FDT test scoring assigns the following standardized penalties: 5 additional seconds for supinating or touching the board with a peg and 10 additional seconds for dropping a peg. Previous studies suggest that assignment of 'adult-grade' penalties to children is inappropriate because of the young child's shorter attention span and distractibility. It has been decided to base our calculations on the data without adult penalties (time-only). Instead, if a penalty occurs (supinating, board touch, dropping) just stop the stopwatch and replace the peg. This repeated effort would capture a time penalty within itself.[5]

2. Nine Hole Peg Test

Background/Purpose: The nine-hole peg test is used to examine fine motor ability, dexterity and hand/eye coordination as a measure of hand function. The nine-hole peg test has been shown to be highly repeatable and valid in healthy children and adults with CMT, [6-9] and norms are available.[7]

Test Position: The dominant hand is tested at a desk and chair with the pegboard centered in front of the participant and the container side on the same side as the hand being tested.

Testing Procedures: The evaluator demonstrates with 4 pegs. The child is given a practice trial with 4 pegs using their dominant hand.

The evaluator says,

> "This is a timed test to see how fast you can put the pegs in and then take them back out. Pick up each peg one at a time. I want you to do the best that you can. Are you ready? Go."

The stopwatch is started when the evaluator says 'go' and ends when the last peg is returned to the container. If a peg is dropped, stop the stopwatch and replace the peg to its initial position. The time it takes, in seconds, to insert all nine pegs and remove them again is recorded.

For those unable to perform the nine-hole peg test due to CMT severity or are very slow, give a score of **150 seconds** which is greater than the maximum childhood CMT value.

3. Grip strength

Background/Purpose: Grip strength is quantified using the Citec hand-held dynamometer. Quantitative muscle testing of hand strength using standardized procedures has been shown to be highly reliable and valid in children and adults with a variety of neuromuscular conditions.[6,8,10-12]

Test Position: The child is asked to sit in a chair with their feet supported. The testing arm is positioned with the shoulder adducted and in neutral rotation, elbow flexed to 90 degrees, forearm in neutral with the wrist between 0 and 30 degrees of extension and 0 to 15 degrees of ulnar deviation.

Testing Procedure: Three *valid* trials of 3 to 5 seconds for each muscle group of the dominant hand is recorded and averaged. The child is asked to grasp the hand-held dynamometer with the fingers wrapped around the handle. The child is instructed to squeeze the handle as hard as they can.

Ensure hand-held dynamometer visual display is away from the patient.

In accordance with the manufacturer's instructions, the displayed grip value must be **multiplied by 2** (grip applicator passes on measured strength in a 1:2 ratio).

For those unable to perform grip strength due to CMT severity or are very weak, score **0 Newtons**.

4. Plantarflexion strength
5. Dorsiflexion strength

Background/Purpose: Plantarflexion and dorsiflexion strength is quantified using the Citec hand-held dynamometer. Using standardized procedures the reliability and validity of using hand-held dynamometry to measure foot strength has been shown to be acceptable in children[13] and adults.[14]

Test Position: The child is seated with hips flexed and knees comfortably extended (long sitting). The evaluator stabilizes the lower limb proximal to the ankle joint.

The dynamometer is positioned according to the muscle group being tested.

Plantarflexion: The hand-held dynamometer is positioned against the plantar surface of the foot just proximal to the metatarsal heads (Figure 1). In older children normal plantarflexors are too strong to test, so we are measuring weakness. For plantarflexion, if the patient overpowers the clinical evaluator, score **250 Newtons.**

Dorsiflexion: The hand-held dynamometer is positioned against the dorsal surface of the foot just proximal to the metatarsal heads (Figure 2).

Testing Procedures: Each child is assessed using the "make" test, whereby the assessor holds the hand-held dynamometer stationary while the child exerts a maximal force against it. The maximum effort should be recorded at the available mid-range position of the foot.

Three *valid* trials of 3 to 5 seconds for each muscle group of the dominant foot is recorded and averaged.

For those unable to perform plantarflexion or dorsiflexion strength due to CMT severity or are very weak, score **0 Newtons**.

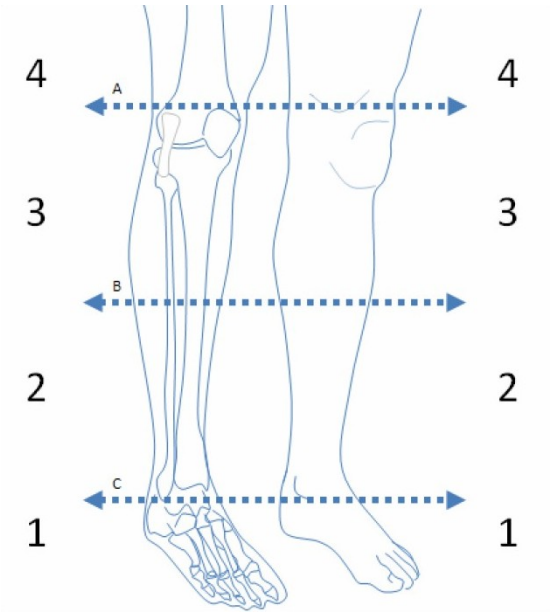
6. Pinprick

Background/Purpose: The sensory exam requires the use of instruments that are strange to most children, and for that reason the clinical evaluator should carefully explain to each child what sensations to expect from each instrument before starting the exam. To help familiarize, let the child practice on him or herself.

Test Position: Dominant lower limb only is tested. All children should have their eyes closed during the test, in order to help them keep focus throughout the exam.

Testing Procedures: The first important part of this exam is to determine if the child is able to discriminate between the dull and sharp sides of the Neurotip™. This test should be first performed in a region with "expected" normal sensation (distal thigh, above top of the patella). If the child does not feel pain, score 4 and go to the next test. If he/she does feel pain, the test will be performed in the lower limbs (distal-to-proximal direction) and graded according to the 4 levels shown below from the *CMT Neuropathy Score* (CMTNS-2nd, 2010):

0 Normal
1 Decreased below or at ankle bones (Note: Below the line passing at ankle malleoli)
2 Decreased at or below the midline of the calf
3 Decreased above the calf midline, up to and including knee (Knee = Top of the patella.)
4 Decreased above the knee (above top of the patella)



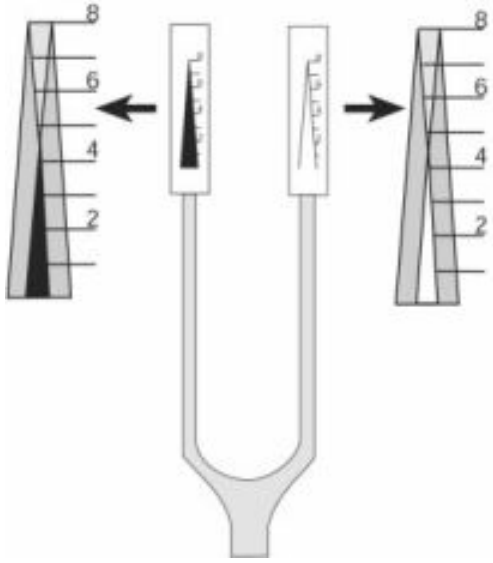
7. Vibration

Background/Purpose: As above, the sensory exam requires the use of instruments that are strange to most children, and for that reason the clinical evaluator should carefully explain to each child what sensations to expect from each instrument before starting the exam. To help familiarize, let the child practice on him or herself.

Test Position: Dominant lower limb only is tested. All children should have their eyes closed during the test, in order to help them keep focus throughout the exam.

Testing Procedures: Determine if the child is able to feel the tuning fork vibrating on a bony region with "expected" normal sensation (e.g. collar bone). Set the tuning fork into motion by compressing the prongs (tines) in a finger snapping motion. As the prongs start to oscillate, the illusion of two triangles is visible on each damper. The child is asked to indicate the moment when they can no longer perceive the decreasing vibratory stimulus. Read off the black triangle. If the child does not feel any vibration at the "expected" normal region, score 4 and go to the next test. If the child does feel vibration, the test will be performed in the lower limbs (distal-to-proximal direction) and graded according to the 4 levels described below from the *CMT Neuropathy Score* (CMTNS-2nd, 2010):

0 Normal ($\geq$5)[15]
1 Reduced at great toe (first metatarsal bone)
2 Reduced at ankle
3 Reduced at knee (tibial tuberosity)
4 Absent at knee and ankle



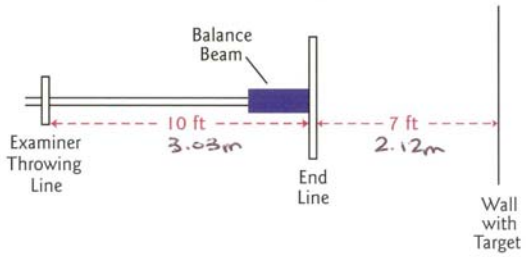
8. Balance

Bruininks-Oseretsky Test of Motor Proficiency, Second Edition (BOT-2) (NCS Pearson, Upper Saddle River, NJ, USA): Balance subtest.

Background/Purpose: The BOT-2 is an individually administered test that uses engaging, goal-directed activities to measure a wide array of motor skills. We are using the BOT-2 subtest of balance.

Tests of standing and walking balance include: standing with feet apart on a line (eyes open/closed); walking forward on a line and walking forward heel-to-toe on a line; standing on one leg on a line (eyes open/closed); standing on one leg on a balance beam (eyes open/closed); and standing heel-to-toe on a balance beam.[16] The BOT-2 incorporates age-equivalent normative data and is regarded as a reliable and valid measure of motor ability.[17]

Course set-up



General Directions

- Perform using the portion of the running course from the 'examiner throwing line' to the 'end line' and the target on the wall (see diagram above).
- Place the target on the wall at the appropriate level for the child's height, with the bottom of the target at the examiner's eye level.
- For items using the balance beam, place the beam at the 'end line'.
- For each item, conduct a second trial *only if* the child does not earn the maximum score on their first trial.
- Before administering each item, teach the task to the child using verbal and non-verbal directions as necessary to ensure the child's understanding of the task.

Tests should be performed barefoot and without an assistive device. If assistive devices are required (e.g. AFOs) they should be clearly documented with footwear details, and repeated at follow-up.

For those unable to perform Balance due to CMT severity, score **0 Points**.

See over the page for BOT-2 Balance Subtest Instructions:

BOT-2 Item 1. Standing with feet apart on a line-eyes open

Procedure:

- Child stands with feet together, dominant foot on and parallel to the line
- Child places hands on hips
- Child takes one natural step forward, placing non-dominant foot on and parallel to line, and looks at the target

Scoring:

- Record number of seconds, to nearest tenth of a second, that the child maintains proper form, up to 10 seconds
- Stop trial after 10 seconds or if the child steps off line or fails to keep hands on hips

Administration:

- Teach the task to the child. Then, say, "Stand on the line with your feet apart until I tell you to stop. Ready? Begin."
- Begin timing *when the child attains proper form*. After 10 seconds or when the child breaks proper form, say, "Stop."
- If the child does not earn the maximum score of 10 seconds, conduct the second trial. If necessary, re-teach the task after you say," Let's try it again."

BOT-2 Item 2. Walking forward on a line

Procedure:

- Child stands with feet together, dominant foot on and parallel to the line
- Child places hands on hips
- Child walks forward in a natural walking stride, placing feet on and parallel to line with each step

Scoring:

- Record number of correct steps, up to 6.
- A step is incorrect if the child steps off the line, fails to keep hands on hips, stumbles or falls. Stop trial, remind the child of proper form, and conduct second trial.

Administration:

- Teach the task to the child. Then, say, "Walk on the line until I tell you to stop. Ready? Begin."
- After 6 correct steps or an incorrect step, say, "Stop."
- If the child does not earn the maximum score of 6 steps, conduct the second trial. If necessary, re-teach the task after you say, "Let's try it again."

BOT-2 Item 3. Standing on one leg on a line - eyes open

Procedure:

- Child stands with feet together, dominant foot on and parallel to the line
- Child places hands on hips
- Child raises non-dominant leg behind him- or herself, with knee bent 90^0 and shin parallel to floor, and looks at the target

Scoring:

- Record number of seconds, to nearest tenth of a second, that the child maintains proper form, up to 10 seconds
- Stop trial after 10 seconds or if the child fails to keep raised leg lifted to at least 45^0, fails to keep hands on hips, or steps or falls off the line

Administration:

- Teach the task to the child. Then, say, "Stand on one leg on the line until I tell you to stop. Ready? Begin."
- Begin timing *when the child attains proper form*. After 10 seconds or when the child breaks proper form, say, "Stop."
- If the child does not earn the maximum score of 10 seconds, conduct the second trial. If necessary, re-teach the task after you say, "Let's try it again."

BOT-2 Item 4. Standing with feet apart on a line - eyes closed

Procedure:

- Child stands with feet together, dominant foot on and parallel to the line
- Child places hands on hips
- Child takes one natural step forward, placing non-dominant foot on and parallel to line, and closes his or her eyes

Scoring:

- Record number of seconds, to nearest tenth of a second, that the child maintains proper form, up to 10 seconds
- Stop trial after 10 seconds or if the child steps off line, fails to keep hands on hips, or opens eyes

Administration:

- Teach the task to the child. Then, say, "Stand on the line with your feet apart and your eyes closed until I tell you to stop. Ready? Begin."
- Begin timing *when the child attains proper form*. After 10 seconds or when the child breaks proper form, say, "Stop."
- If the child does not earn the maximum score of 10 seconds, conduct the second trial. If necessary, re-teach the task after you say, "Let's try it again."

BOT-2 Item 5. Walking forward heel-to-toe on a line

Procedure:

- Child stands with feet together, dominant foot on and parallel to the line
- Child places hands on hips
- Child walks forward heel-to-toe, placing feet on and parallel to the line and touching heel to toe with each step

Scoring:

- Record number of correct steps, up to 6.
- A step is incorrect if the child fails to step heel-to-toe, steps off the line, fails to keep hands on hips, stumbles or falls. Stop trial, remind the child of proper form, and conduct second trial.

Administration:

- Teach the task to the child. Then, say, "Walk heel-to-toe on the line until I tell you to stop. Ready? Begin."
- After 6 correct steps or an incorrect step, say, "Stop."
- If the child does not earn the maximum score of 6 steps, conduct the second trial. If necessary, re-teach the task after you say, "Let's try it again."

BOT-2 Item 6. Standing on one leg on a line - eyes closed

Procedure:

- Child stands with feet together, dominant foot on and parallel to the line
- Child places hands on hips
- Child raises non-dominant leg behind him- or herself, with knee bent to 90^0 and shin parallel to floor, and closes his or her eyes

Scoring:

- Record number of seconds, to nearest tenth of a second, that the child maintains proper form, up to 10 seconds
- Stop trial after 10 seconds or if the child fails to keep raised leg lifted to at least 45^0, fails to keep hands on hips, steps or falls off the line, or opens eyes

Administration:

- Teach the task to the child. Then, say, "Stand on one leg on the line with your eyes closed until I tell you to stop. Ready? Begin."
- Begin timing *when the child attains proper form*. After 10 seconds or when the child breaks proper form, say, "Stop."
- If the child does not earn the maximum score of 10 seconds, conduct the second trial. If necessary, re-teach the task after you say, "Let's try it again."

BOT-2 Item 7. Standing on one leg on a beam - eyes open

Procedure:

- Child stands with dominant foot on the balance beam and non-dominant foot on the floor
- Child places hands on hips
- Child raises non-dominant leg behind him- or herself, with knee bent to 90^0 and shin parallel to floor, and looks at the target

Scoring:

- Record number of seconds, to nearest tenth of a second, that the child maintains proper form, up to 10 seconds
- Stop trial after 10 seconds or if the child fails to keep raised leg lifted to at least 45^0 , fails to keep hands on hips, or steps or falls off the beam

Administration:

- Teach the task to the child. Then, say, "Stand on one leg on the beam until I tell you to stop. Ready? Begin."
- Begin timing *when the child attains proper form*. After 10 seconds or when the child breaks proper form, say, "Stop."
- If the child does not earn the maximum score of 10 seconds, conduct the second trial. If necessary, re-teach the task after you say, "Let's try it again."

BOT-2 Item 8. Standing heel-to-toe on a balance beam

Procedure:

- Child stands with dominant foot on the balance beam and non-dominant foot on the floor
- Child places hands on hips
- Child takes one step forward, placing non-dominant foot on the balance beam and touching the heel of the front foot to toe of back foot, and looks at the target

Scoring:

- Record number of seconds, to nearest tenth of a second, that the child maintains proper form, up to 10 seconds
- Stop trial after 10 seconds or if the child fails to keep feet heel-to-toe, fails to keep hands on hips, or steps or falls off the beam

Administration:

- Teach the task to the child. Then, say, "Stand heel-to-toe on the beam until I tell you to stop. Ready? Begin."
- Begin timing *when the child attains proper form*. After 10 seconds or when the child breaks proper form, say, "Stop."
- If the child does not earn the maximum score of 10 seconds, conduct the second trial. If necessary, re-teach the task after you say, "Let's try it again."

BOT-2 Item 9. Standing on one leg on a beam - eyes closed

Procedure:

- Child stands with dominant foot on the balance beam and non-dominant foot on the floor
- Child places hands on hips
- Child raises non-dominant leg behind him- or herself, with knee bent to 90^0 and shin parallel to floor, and closes his or her eyes

Scoring:

- Record number of seconds, to nearest tenth of a second, that the child maintains proper form, up to 10 seconds
- Stop trial after 10 seconds or if the child fails to keep raised leg lifted to at least 45^0, fails to keep hands on hips, steps or falls off the beam, or opens eyes.

Administration:

- Teach the task to the child. Then, say, "Stand on one leg on the beam with your eyes closed until I tell you to stop. Ready? Begin."
- Begin timing *when the child attains proper form*. After 10 seconds or when the child breaks proper form, say, "Stop."
- If the child does not earn the maximum score of 10 seconds, conduct the second trial. If necessary, re-teach the task after you say, "Let's try it again."

For those unable to perform any of the Balance items due to CMT severity, score **0 Points**.

9. Gait

Background/Purpose: Difficulty toe walking is a gross indicator of plantarflexion weakness, difficulty heel walking is a gross indicator of dorsiflexion weakness (and Achilles tendon shortening), and the presence of foot drop is a sign of dorsiflexion weakness during gait.[17]

Testing procedure:

i. The child is asked to walk 10 steps and the clinical evaluator assesses the presence of foot drop:

If the child demonstrates NO forefoot strikes on all steps a score of 1 is recorded.
If child demonstrates SOME forefoot strikes during gait or lands flatfooted, a score of 2 is recorded.
If the child demonstrates forefoot strikes on ALL steps, a score of 3 is recorded.

*Detecting foot drop can be subtle, so ask the child to walk additional steps or confirm during the 6-min walk test.

ii. The child is then asked to heel walk 10 steps:

If the child demonstrates NO difficulty heel walking, a score of 1 is recorded.
If the child demonstrates SOME difficulty heel walking, a score of 2 is recorded.
If the child demonstrates difficulty heel walking on ALL steps, a score of 3 is recorded.

iii. The child is then asked to tip-toe walk for 10 steps:

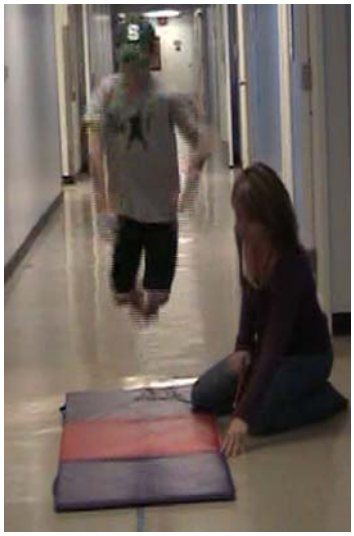
If the child demonstrates NO difficulty tip-toe walking, a score of 1 is recorded.
If the child demonstrates SOME difficulty tip-toe walking, a score of 2 is recorded.
If the child demonstrates difficulty tip-toe walking on ALL steps, a score of 3 is recorded.

10. Long Jump

Background/Purpose: Long Jump is a reliable measure of power and coordination in children.[18]

Test Position: Long jump should be performed barefoot and without an assistive device. If assistive devices are required (e.g. AFOs) they should be clearly documented with footwear details, and repeated at follow-up.

Testing Procedures: The child starts standing with the feet even and shoulder width apart behind a line on a carpeted floor or firm cushioned mat. The child is instructed to jump as far forward as they can, taking off and landing with both feet. The distance from the start line to the point where the **heel of the foot nearest** to the start line touched the floor is measured using a tape measure.[19]



Teach the task to the child and allow practice. Perform one time only, unless invalid.

Caution: Use carpet or padded mat for comfort if required.

For those unable to perform long jump due to CMT severity, score **0 centimeters**.

11. Six-minute walk test

Background/Purpose: The 6-minute walk test (6MWT) is a clinical measure of submaximal functional endurance capacity.[20,21] High test-retest reliability has been reported in children aged 4-18 years, and recently in boys aged 4-12 y with Duchenne muscular dystrophy (ICC=0.91) with no obvious learning effect.[22] The 6MWT is administered according to the American Thoracic Society guidelines with modifications to keep the child focused.[22,23] Lack of motivation and understanding can affect the 6MWT performance in children so clear instructions and abundant encouragement are required to ensure test accuracy, feasibility, and safety.

Set-up: Walking course is set in a quiet corridor and marked with small marker cones at *exactly* 0 and 25 meters based on the uniform standard protocol for pediatric neuromuscular disorders recently published.[22] Ensure the same floor surface at each visit. The 6MWT is performed by one child at a time to preclude any influence of competition between children.

Testing Procedures: Six-minute walk test should be performed barefoot and without an assistive device. If assistive devices are required (e.g. AFOs) they should be clearly documented with footwear details, and repeated at follow-up. Before the start of the 6MWT, the child sits and rests for 3 minutes. Children are instructed to walk as many laps as possible in 6 minutes without running. Briefly demonstrate the process of walking around the cones. A practice trial is not given for this lengthy test because of concerns about the impact of associated fatigue, and in light of evidence that a familiarization trial does not enhance reliability.[24]

The evaluator asks the child to stand and place his or her toes at the starting line. When the child is ready, the clinical evaluator says, ''Ready, set, go,'' and starts the stopwatch. Children are given standardized instructions to try to cover as much distance as possible in 6 minutes without running at approx 15 seconds intervals or as required (e.g. 'great work' and 'keep it up').[22]

At each minute-split, the timer announces the number of minutes completed and the number remaining. Each time the child rounds a cone, record the lap on the tally (lap) counter. Parent should not accompany child. The cumulative distance to the nearest lap marker is recorded and any remaining distance is measured to the nearest centimeter.

Caution: Bandage any loose callosities, blisters, wounds to avoid further trauma. If a fall occurs ensure the child is unharmed, and assist them back to a standing position and resume.

For those unable to perform the 6MWT due to CMT severity or are very weak, score **0 meters**.

6MWT course at Great Ormond Street Hospital, London

Patient Profile Instructions

Complete *ID, DOB, Age, Gender, Height, Weight, Dominant Hand/Foot, Diagnosis*

Symptoms

Common patient/parent complaints in children with CMT include foot pain (27%), hand pain (24%), leg cramps (36%), hand weakness (48%), unsteady ankles during walking (72%), daily trips and/or falls (47%), hand tremor (25%), sensory symptoms (e.g. pins and needles, tingling, numbness, prickling) (13%).[6,17]

Each child/parent is asked about the presence or absence of each symptom and marked as present or absent.

Note: Look for tremor throughout assessment and ask about symptom history if observed.

Lunge test (degrees)

Flexibility of ankle joint dorsiflexion is measured weight bearing using the lunge test. This technique has been shown to be reliable and valid in children.[25]

Test Position: The child is asked to place their foot perpendicular to a wall and lunge their knee toward the wall.[26] The foot is progressively moved away from the wall until the maximum angle of ankle joint dorsiflexion is obtained without the heel lifting. Pronation and supination of the subtalar and midtarsal joints is restricted by ensuring that the foot is positioned perpendicular to the wall, and the child lunges directly over the midline of the foot (second toe). Children are instructed to hold onto the wall for balance and the contralateral leg is placed in a comfortable position. While the child is positioned in maximum dorsiflexion range, a digital inclinometer is aligned with the midline of the Achilles tendon. The number of degrees is recorded.

Alternate Position: If weight bearing lunge is not possible, a non-weight bearing measurement in prone is conducted. Position the child in prone with the knee flexed to 90 degrees. Passively dorsiflex the ankle to the maximum range of motion and measure the maximum dorsiflexion range of motion using the inclinometer. If this approached used, mark on the CMTPedS.

Foot Posture Index

Foot structure of each foot of all children is assessed using the Foot Posture Index (FPI), a diagnostic tool that evaluates the multisegmental and multiplanar aspects of the foot using six criteria that together enable the foot to be scored along a continuum of cavus (supinated) to planus (pronated) features.[27]

The FPI allocates a score between -2 and +2 to each of the six criteria:

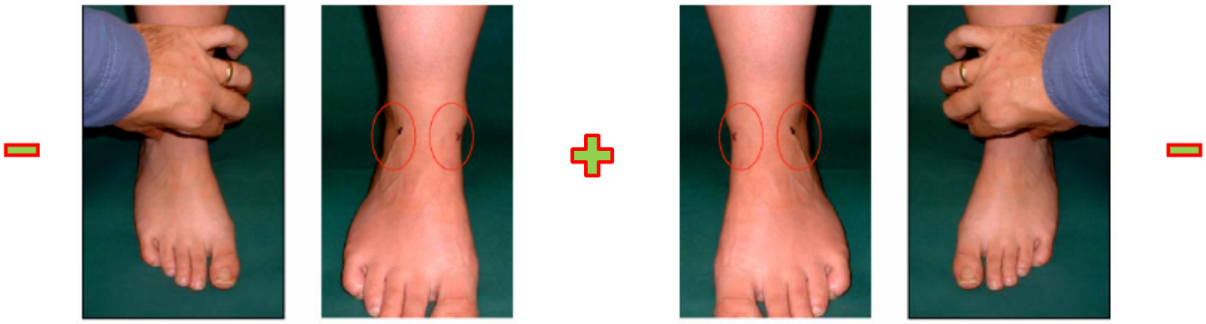
1. Talar head palpation
2. Curves above and below the lateral malleolus
3. Inversion/eversion of the calcaneus
4. Prominence in the region of the talonavicular joint
5. Congruence of the medial longitudinal arch
6. Abduction/adduction of the forefoot on the rearfoot)

Scores are allocated for each criterion with a score of 0 denoting a neutral position, -2 for clear signs of supination, and +2 for clear signs of pronation. The aggregated score ranges from -12 (extremely supinated/pes cavus) to +12 (extremely pronated/pes planus).

Test Position: All observations of the FPI are made with the participant in a relaxed stance with double limb support. They are asked to take several steps on the spot and then to relax and stand still, with their arms by their side and looking straight ahead. Ensure the child does not swivel to see what is happening as this will change the foot posture. The FPI should take 2 minutes.

Scoring information and instruction is detailed below. For more information and full training manual: http://www.leeds.ac.uk/medicine/FASTER/fpi.htm

1.Talar head palpation



Right Foot

Left Foot

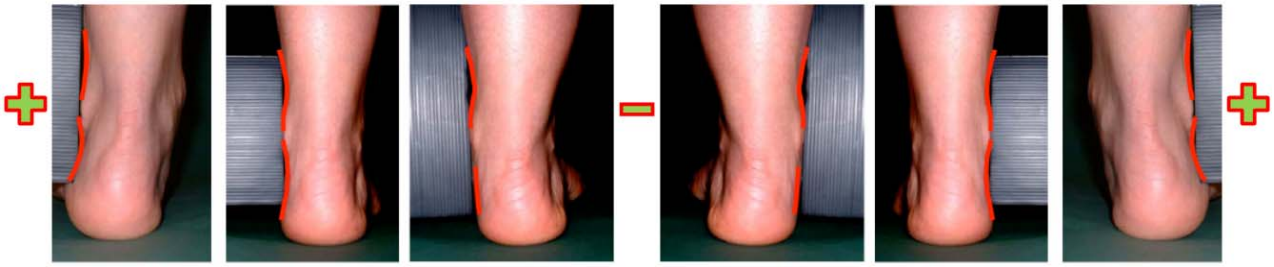
-2	-1	0	+1	+2
Talar head palpable on **lateral** side/but not on medial side	Talar head palpable on **lateral** side/ slightly palpable on medial side	Talar head **equally** palpable on lateral and medial side	Talar head palpable on **medial** side/slightly palpable on lateral side	Talar head palpable on **medial** side/but not on lateral side

+2	+1	0	-1	-2
Talar head palpable on **medial** side/but not on lateral side	Talar head palpable on **medial** side/slightly palpable on lateral side	Talar head **equally** palpable on lateral and medial side	Talar head palpable on **lateral** side/ slightly palpable on medial side	Talar head palpable on **lateral** side/but not on medial side

2.Curves above and below the lateral malleolus

Left Foot

Right Foot

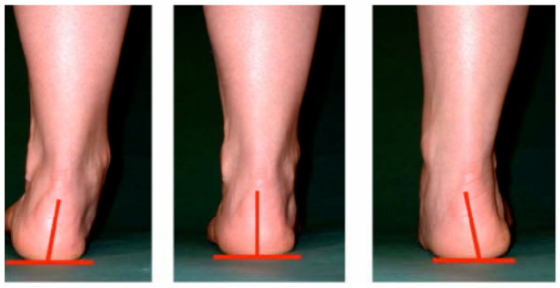


+2	+1	0	-1	-2
Curve below malleolus *markedly* more concave than curve above malleolus	**Curve below malleolus more concave** than curve above malleolus	**Both infra and supra malleolar curves roughly equal**	**Curve below the malleolus concave but flatter/** more shallow than the curve above malleolus	**Curve below the malleolus either straight or convex**

-2	-1	0	+1	+2
Curve below the malleolus either straight or convex	**Curve below the malleolus concave but flatter/** more shallow than the curve above malleolus	**Both infra and supra malleolar curves roughly equal**	**Curve below malleolus more concave** than curve above malleolus	**Curve below malleolus *markedly* more concave** than curve above malleolus

3.Inversion/eversion of the calcaneus

Left Foot



Right Foot

+2	+1	0	-1	-2
More than an estimated **5° everted** (valgus)	**Between vertical and** an estimated **5° everted** (valgus)	Vertical	**Between vertical and** an estimated **5° inverted** (varus)	**More than** an estimated **5° inverted** (varus)

-2	-1	0	+1	+2
More than an estimated **5° inverted** (varus)	**Between vertical and** an estimated **5° inverted** (varus)	Vertical	**Between vertical and** an estimated **5° everted** (valgus)	**More than** an estimated **5° everted** (valgus)

4.Bulge in the region of the talonavicular joint

Left Foot

Right Foot

+2	+1	0	-1	-2
Area of TNJ **bulging markedly**	Area of TNJ **bulging slightly**	Area of TNJ **flat**	Area of TNJ **slightly,** but definitely **concave**	Area of TNJ **markedly concave**

-2	-1	0	+1	+2
Area of TNJ **markedly concave**	Area of TNJ **slightly,** but definitely **concave**	Area of TNJ **flat**	Area of TNJ **bulging slightly**	Area of TNJ **bulging markedly**

5.Congruence of the medial longitudinal arch

Supinated foot (-2)

Pronated foot (+2)

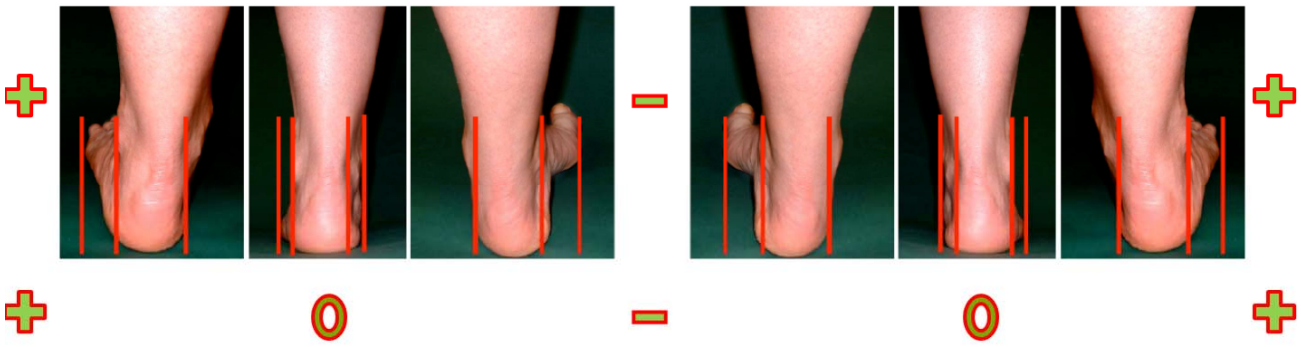
-2	-1	0	+1	+2
Arch highly **acutely angled** towards the posterior end of the medial arch	Arch **moderately** high and slightly acute posteriorly	Arch height **normal** and concentric ally curved	Arch **lowered** with some flattening in the central portion	Arch very low with **severe flattening** in the central portion – arch making **ground contact**

6.Abduction/adduction of forefoot and rearfoot

(too-many-toes)

Left Foot

Right Foot



+2	+1	0	-1	-2
No medial toes visible . Lateral toes clearly visible	**Lateral** toes clearly **more** visible than medial	Medial and lateral toes **equally** visible	**Medial** toes clearly more visible than lateral	**No lateral** toes visible . Medial toes clearly visible

-2	-1	0	+1	+2
No lateral toes visible . Medial toes clearly visible	**Medial** toes clearly more visible than lateral	Medial and lateral toes **equally** visible	**Lateral** toes clearly more visible than medial	**No medial** toes visible . Lateral toes clearly visible

Calibration Tasks

Citec hand-held dynamometer

Every 3-months check force readings with a known weight which has been verified by a biomedical department or calibrated scales.

Using the technique shown whereby a 5 kilogram known weight (49 Newtons) is applied to the Citec hand-held dynamometer.

For alternate weight, use Force Convertor website: www.unitconversion.org/unit_converter/force.html

Frequently Asked Questions

1. Why does the Citec hand-held dynamometer turn off during testing?

This can occur when the battery casing is loose and requires tightening which can happen because we frequently change applicators. Otherwise the batteries may need changing.

2. Why does the Citec hand-held dynamometer need frequent adjustments to calibrate to zero?

The new Citec has an auto-zero function. With the earlier Citec device, keeping it +/- 2 N is acceptable and should not require too much adjustment. If beyond +/- 2 N, either adjust blue wheel on side or just add or subtract the starting value from the force produced.

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