

European Network on Myalgic Encephalomyelitis/Chronic Fatigue Syndrome (*EUROMENE*)

COST action CA15111

Deliverable 11

**Evaluation scales for assessment of neurological symptoms associated with
ME/CFS and usable in diagnostic**

WG4 - Leader Prof Jerome Authier

Canadian consensus criteria (CCC) → clinical criteria for ME/CFS

domain **Neurological / Cognitive**

Two or more of the following difficulties should be present:

- Confusion
- Impairment of concentration and short-term memory consolidation
- Disorientation
- Difficulty with information processing, categorizing and word retrieval (Word-finding problems)
- Perceptual and sensory disturbances (for example spatial instability and disorientation and inability to focus vision)
- Ataxia, muscle weakness and fasciculations are common.
- Overload phenomena:
 - cognitive overload,
 - sensory overload (for example photophobia and hypersensitivity to noise) and/or emotional overload,
→ crash eriods and/or anxiety.

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EUROMENE paper in *Cortex*



Neuropsychological
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*Neurological
impairment or dysfunctioning?*

International consensus criteria (ICC) → clinical criteria for ME

domain **Neurological impairments**

At least one symptom from three of the following four symptom categories

1. Neurocognitive impairments

- a. Difficulty processing information
- b. Short-term memory loss

2. Pain

- a. Headaches
- b. Significant pain in muscles, muscle-tendon junctions, joints, abdomen or chest
→ generalized hyperalgesia, widespread pain (fibromyalgia), myofascial

3. Sleep disturbance

- a. Disturbed sleep patterns
- b. Unrefreshed sleep

4. Neurosensory, perceptual and motor disturbances

- a. Neurosensory and perceptual: e.g. inability to focus vision, sensitivity to light, noise, vibration, odour, taste and touch; impaired depth perception
- b. Motor: e.g. muscle weakness, twitching, poor coordination, feeling unsteady on feet, ataxia

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noise, vibration, odour, taste
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- *Screening → Epworth
sleepiness scale*

- *Search for sleep apnea
syndrome?*

- *Sleep medicine specialist?*

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Pain: Fibromyalgia

1990 American College of Rheumatology criteria
→ trigger points = decreased threshold
ACR 2010 symptom severity



RESEARCH
EDUCATION
TREATMENT
ADVOCACY



The Journal of Pain, Vol 20, No 6 (June), 2019: pp 611–628
Available online at www.jpain.org and www.sciencedirect.com

Critical Reviews

AAPT Diagnostic Criteria for Fibromyalgia



2018 AAPT diagnostic criteria → to facilitate fibromyalgia diagnosis

Dimension 1 includes core diagnostic criteria, which are three:

- (1) multisite pain defined ≥ 6 or more pain sites (from 9 possible sites);
- (2) Moderate to severe sleep problems OR fatigue;
- (3) MSP plus fatigue or sleep problems > 3 months.

Other dimensions reinforce diagnostic conviction: common features, epidemiology, psychiatric comorbidities, functional consequences and risk factors

Dimension 1: Core Diagnostic Criteria → inclusion/exclusion

1. **Multi-Site Pain** defined as 6 or more pain sites from a total of 9 possible sites (see Fig. 1)

2. Moderate to severe **sleep** problems OR **fatigue**

3. MSP plus fatigue or sleep problems must have been present for **at least 3 months**

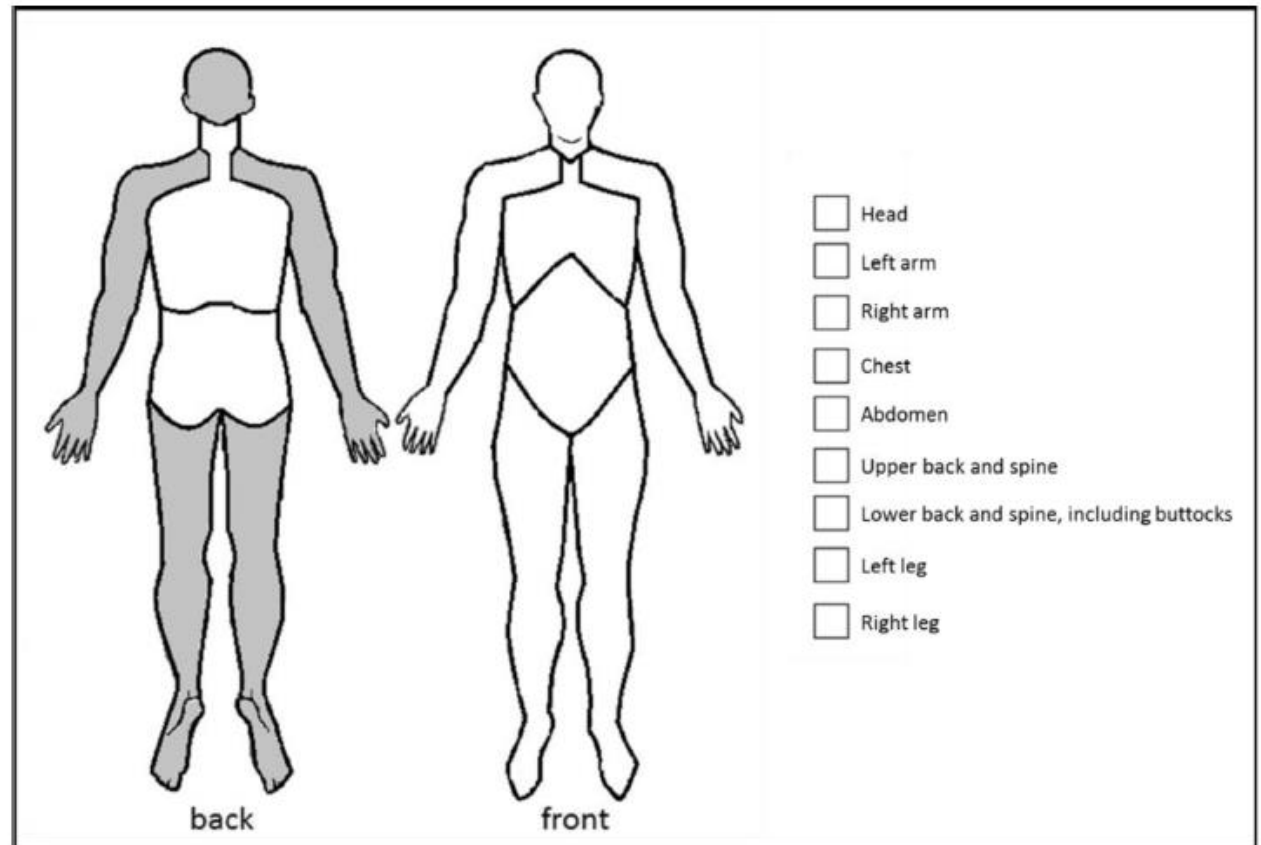


Figure 1. Number of painful bodysites.

Patients are asked to check the areas in which they experience pain on the 2-view manikins (ignoring the pre-shaded areas). Alternatively, patients may use the checklist of body sites. The number of separate sites are summed from a maximum of 9 body sites.

Pain: Neuropathic pain

DN4 – QUESTIONNAIRE

To estimate the probability of neuropathic pain, please answer yes or no for each item of the following four questions.

INTERVIEW OF THE PATIENT

QUESTION 1:

Does the pain have one or more of the following characteristics?

YES

NO

Burning

☐

☐

Painful cold

☐

☐

Electric shocks

☐

☐

QUESTION 2:

Is the pain associated with one or more of the following symptoms in the same area?

YES

NO

Tingling

☐

☐

Pins and needles

☐

☐

Numbness

☐

☐

Itching

☐

☐

EXAMINATION OF THE PATIENT

QUESTION 3:

Is the pain located in an area where the physical examination may reveal one or more of the following characteristics?

YES

NO

Hypoesthesia to touch

☐

☐

Hypoesthesia to pinprick

☐

☐

QUESTION 4:

In the painful area, can the pain be caused or increased by:

YES

NO

Brushing?

☐

☐

YES = 1 point

NO = 0 points

Patient's Score:

/10

Neuropathic pain if score ≥ 4
sensitivity: 83%; specificity: 90%

Bouhassira D, Attal N, Alchaar H, et al. "Comparison of pain syndromes associated with nervous or somatic lesions and development of a new neuropathic pain diagnostic questionnaire (DN4)." Pain 114.1-2 (2005): 29-36.

Neurosensory, perceptual and motor disturbances

- Refer to neurologist ? The analysis of subtle changes requires experience
- **Motor function**
 - examination of muscles : muscle bulk, search for abnormal activity (fasciculations, rippling, etc..)
 - muscle strength
 - manual testing (MRC)
 - dynamometer
 - fatigability
 - dynamometer
 - 6-mn walk test (6MWT)

6 Minute Walk Test → sub-maximal exercise test used to assess aerobic capacity and endurance

→ distance covered over a time of 6 minutes

- easy to do; very reproducible for one individual → follow-up
- but reference values depend on gender, age, weight etc...

6 MWT : distance (m) = $218 + [5,14 \times \text{height in cm}] - [5,32 \times \text{age}] - [1,8 \times \text{weight in kg}] + [51,31 \times \text{gender}]$ (0 for female, 1 for male)

Neurosensory, perceptual and motor disturbances

Sensory function → light touch, pinprick, temperature, vibration, proprioception

- Several scales for scoring neuropathic symptoms: Neuropathy Impairment Score of the lower limb (NIS-LL), Michigan Diabetic Neuropathy Score (MDNS), modified Toronto Clinical Neuropathy Score (mTCNS), Total Neuropathy Score-clinical (TNS-C), Neuropathy Disability Score (NDS);
- **Early Neuropathy Score (ENS)** → assess key abnormalities in early neuropathy
 - sensory loss → monofilament testing on the hallux
 - vibration testing → Rydel-Seiffer tuning fork on the interphalangeal joint of the hallux,
 - pin perception on the hallux using a nickel-plated steel, size #2 safety pins,
 - cold perception using metal thermal disks on the dorsum of the foot,
 - ankle reflexes

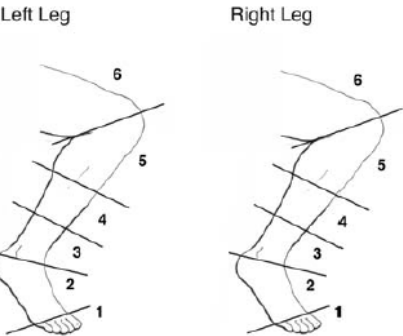
Bilateral testing → 0 = normal result; 1 = reduced result; 2 = absent result

Patient Name
Study Number
Visit

The Utah Early Neuropathy Scale

Motor Examination	Left	Right
0 normal		
2 weak		
Great Toe Extension	<input type="text"/>	<input type="text"/>
Total both sides (out of 4)	<input type="text"/>	

Segments for pin sensation reporting



Pin Sensation:	L	R
0 normal		
1 for each segment with reduced sensation	<input type="text"/>	<input type="text"/>
2 for each segment with absent sensation	<input type="text"/>	<input type="text"/>
Total both sides (out of 24)	<input type="text"/>	

Allodynia/Hyperesthesia	L	R
0 normal		
1 if present in toes or foot	<input type="text"/>	<input type="text"/>
Total both sides (out of 2)	<input type="text"/>	

Large Fiber Sensation	L	R
0 normal		
1 diminished		
2 absent		
Great toe vibration	<input type="text"/>	<input type="text"/>
time	s	s
Great toe joint position	<input type="text"/>	<input type="text"/>
Total both sides (out of 8)	<input type="text"/>	

Deep Tendon Reflexes	L	R
0 normal		
1 diminished		
2 absent		
Ankle	<input type="text"/>	<input type="text"/>
Total both sides (out of 4)	<input type="text"/>	

Total Score (out of 42)	<input type="text"/>
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Assessment of neurological symptoms

- Interview, symptoms description
- Clinical neurological examination
- Pain → AAPT scale, DN4 score
- Motor/fatigability → handgrip muscle strength
- Sensory function → ENS scale

***Refer to neurologist
if CNS/PNS involvement
is suspected***

Lab investigations (if necessary)

- EMG, small nerve fiber investigation (Laser EP, Sudoscan, QST)
- Muscle biopsy, skin biopsy for intra-epidermic NF density quantification
- CNS EP, visual, sensory, auditory, motor
- Brain MRI
- Polysomnography