



COST is supported by the  
EU Framework Programme  
Horizon 2020

## **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*



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# 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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- a. Neurosensory and perceptual: e.g. noise, vibration, odour, taste
- b. Motor: e.g. muscle weakness, twitching, feet, ataxia

- *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](http://www.jpain.org) and [www.sciencedirect.com](http://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  $\geq 6$  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.

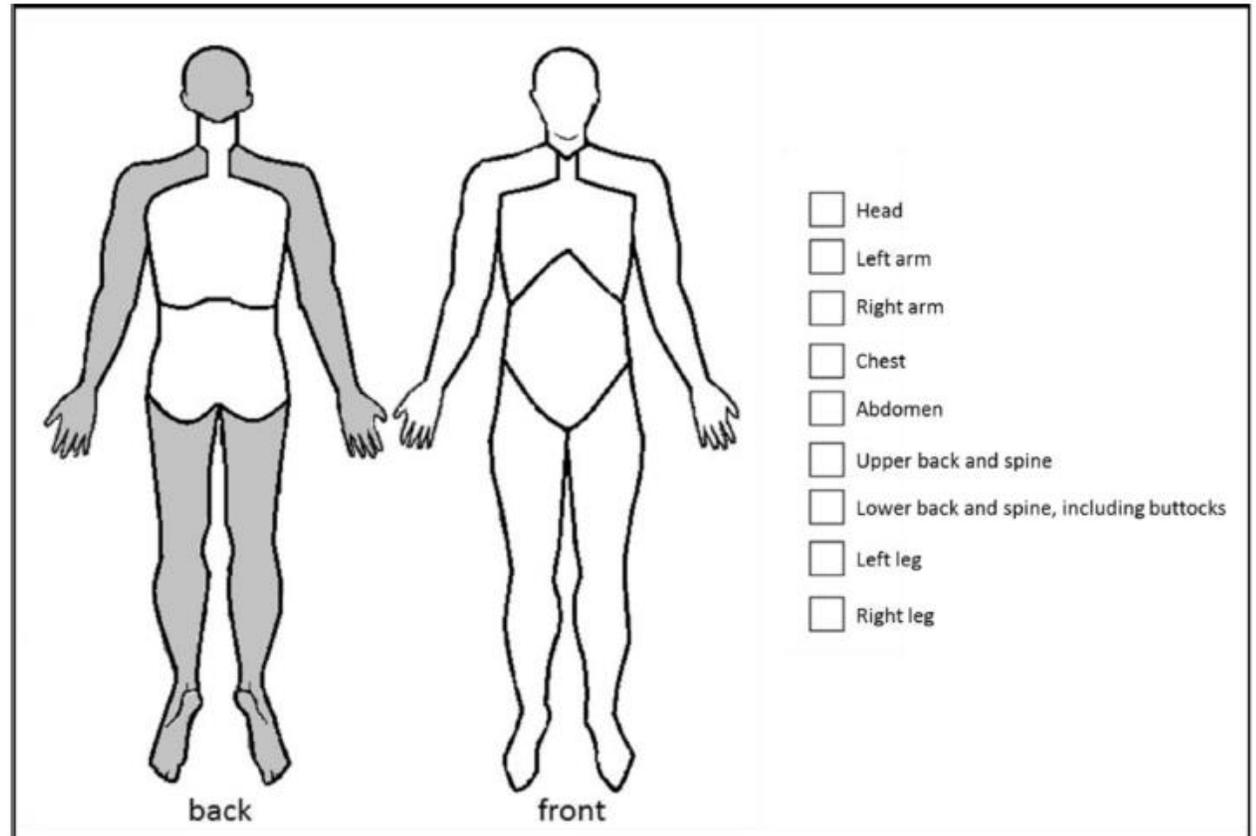
Other dimensions reinforce diagnostic conviction: common features, epidemiology, psychiatric comorbidities, functional consequences and risk factors

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.

Neuropathic pain if score  $\geq 4$   
sensitivity: 83%; specificity: 90%

INTERVIEW OF THE PATIENT		
<b>QUESTION 1:</b>		
Does the pain have one or more of the following characteristics?	YES	NO
Burning .....	<input type="checkbox"/>	<input type="checkbox"/>
Painful cold .....	<input type="checkbox"/>	<input type="checkbox"/>
Electric shocks .....	<input type="checkbox"/>	<input type="checkbox"/>
<b>QUESTION 2:</b>		
Is the pain associated with one or more of the following symptoms in the same area?	YES	NO
Tingling .....	<input type="checkbox"/>	<input type="checkbox"/>
Pins and needles .....	<input type="checkbox"/>	<input type="checkbox"/>
Numbness .....	<input type="checkbox"/>	<input type="checkbox"/>
Itching .....	<input type="checkbox"/>	<input type="checkbox"/>

EXAMINATION OF THE PATIENT		
<b>QUESTION 3:</b>		
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 .....	<input type="checkbox"/>	<input type="checkbox"/>
Hypoesthesia to pinprick .....	<input type="checkbox"/>	<input type="checkbox"/>
<b>QUESTION 4:</b>		
In the painful area, can the pain be caused or increased by:	YES	NO
Brushing? .....	<input type="checkbox"/>	<input type="checkbox"/>

YES = 1 point  
NO = 0 points

Patient's Score:	/10
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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

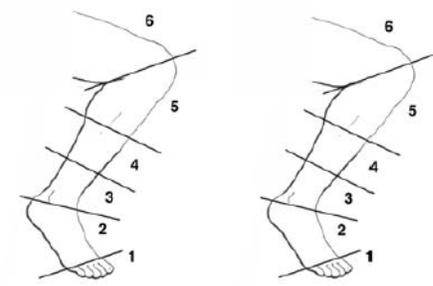
Patient Name
Study Number
Visit

## 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

Left Leg      Right Leg



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 time	<input type="text"/>	<input type="text"/>
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)**

# 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