

RCP MATHEC FORM: INFLAMMATORY DEMYELINATING POLYNEUROPATHY (CIDP)

This form, as well as those pertaining other autoimmune diseases, can be downloaded at the following link: <https://www.mathec.com/soignants/rcp-mathec/>

For each patient with CIDP: Please complete this form for each patient you wish to submit to the RCP MATHEC meeting and send it between 8 and 3 days before the desired date of assessment to the following address: valentina.unfer@aphp.fr or Fax to: **+33 01.42.49.94.78**

Referring Physician

Last Name : _____ **First Name :** _____
Email : _____
Name and address of hospital : _____
Telephone: _____ **Fax :** _____

Referring Neurologist (if different)

Last Name : _____ **First Name :** _____
Email : _____
Name and address of hospital : _____
Telephone : _____ **Fax :** _____

Patient presented

Last Name (maiden name) : _____ **Last name used:** _____
First Name : _____ **Sex :** _____
Date of birth : _____
Address : _____
Telephone : _____

Diagnosis and initial level of disease burden

Date of first symptoms : _____

Date of Diagnosis : _____

EFNS/PNS classification criteria (Annex 1) :

Description :
 Clinical Criteria :

EMG criteria :

Supportive criteria (see table 5 page 6) :

Attach the last electromyogram

Progression of CIDP since diagnosis

List different lines of therapy used, treatment response, and complications encountered

Clinical profile at last follow-up Date :

Weight :

Height :

a. Clinical examination

• **Motor**

➤ **MRC** (*Kleyweg RP, Muscle Nerve 1991*)

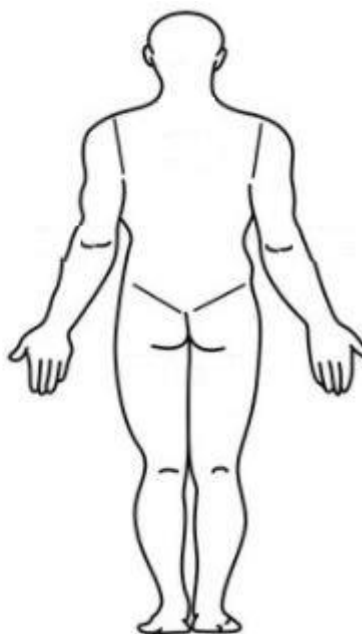
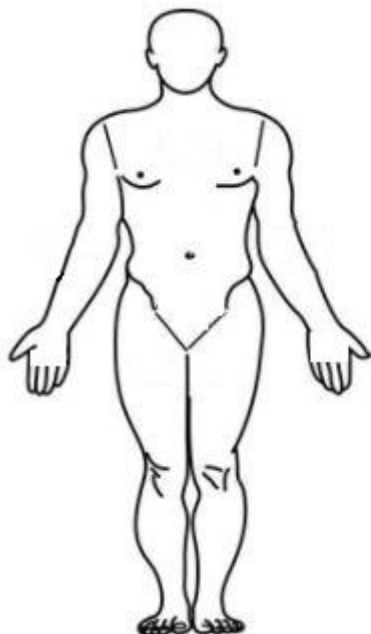
	R	L
Deltoid		
Biceps		
Extensor digitorum muscle		
Abductor digiti minimi muscle of hand		

	R	L
Psoas		
Hamstring		
Tibialis anterior muscle		
Quadriceps		

➤ **ROT :**

• **Sensory**

- Sensory symptoms and topography :



b. Scores

• **Overall Neuropathy Limitations Scale** (*Graham RC, J Neurol Neurosurg Psychiatry, 2006*)

ONLS MS : ONLS MI :

• **Six Minute Walk Test (SMWT)**

Distance :m (Normal value : m)

- **INCAT Sensory sum score** (*Merkies IS, Neurology 2000*)

Sensory score	Specify Side	Pinprick	Vibration	Total
Arm		/4	/4	/8
Leg		/4	/4	/8
Score		/8	/8	/16

0 = Normal, 1 = Distal interphalangeal abnormality 2 = Ankle / wrist abnormality, 3 = elbow / knee abnormality, 4 = Shoulder / hip abnormality

Normal values of Rydell-Seiffer tuning fork: for patients over 41 years of age ≥ 6.0 , and patients under 40 years of age ≥ 6.5

- **Modified Rankin Score** (*Banks JL, Stroke 2007*)

0 No symptoms at all

1 No significant disability despite symptoms; able to carry out all usual duties and activities

2 Slight disability; unable to carry out all previous activities, but able to look after own affairs w/o assistance

3 Moderate disability; requiring some help, but able to walk without assistance

4 Moderately severe disability; unable to walk and attend to bodily needs without assistance

5 Severe disability; bedridden, incontinent and requiring constant nursing care and attention

Cardiovascular assessment

SBP :mmHg

DBP :mmHg

Heart rate :beats/min

ECG :

Echocardiogram: Date : .../.../...

LVEF :% EF :% sPAP :mmHg TAPSE :m/s

Dilated cardiomyopathy :

Valvular insufficiency :

Pericardial effusion : PH :

Pulmonary assessment

smoking : never currently previous Packs per year: Quitting Date / .../...

Thoracic CT scan: no yes Date...../...../20.....

Result :

Pulmonary function tests

Date/...../20.....

Air flow/volume	% Mean predicted value
FVC	
FEV	
FEV/VC max	
Spirometry	
VC max	
TLC	
RV	
Diffusion	
Corrected DLCO	
Corrected DLCO/VA	

Conclusion :

Laboratory parameters Date...../...../20.....

Biochemistry : Hb :.....g/dl WBC :...../mm³ Lymphocytes :..... /mm³
Platelets :...../mm³ Creatinine:.....µmol/l Albumin :.....g/l

Assessment of inflammation: CRP :.....mg/l. fibrinogen :.....g/l

Infectious disease assessment :

	Serology		PCR	
	Results	Date	Results	Date
HIV 1/2 Ag+Ac				
HTLV1/2				
CMV				
EBV				
HBV	Ac HBs			
	Ag HBs			
	Ac HBc			
	Ac HBe			
HCV				
HHV8				
HSV1/2				
Toxoplasmosis				
TPHA / VDRL				

Hepatitis B vaccination : no yes
Pneumococcal vaccination: non yes
Flu vaccine: no yes

CT sinus scan + Consultation in otorhinolaryngology: Date/...../20.....

Panoramic radiograph + Consultation in oral medicine : Date/...../20.....

Gynecologic consultation + mammogram: Date/...../20.....

Current treatments

Annexe 1

Clinical, electrodiagnostic, and supportive diagnostic criteria (Van den Bergh PY, *European Journal of Neurology* 2010)

Table 4 Clinical diagnostic criteria

(1) Inclusion criteria

(a) Typical CIDP

Chronically progressive, stepwise, or recurrent symmetric proximal and distal weakness and sensory dysfunction of all extremities, developing over at least 2 months; cranial nerves may be affected; and

Absent or reduced tendon reflexes in all extremities

(b) Atypical CIDP (still considered CIDP but with different features)

One of the following, but otherwise as in (a) (tendon reflexes may be normal in unaffected limbs):

Predominantly distal (distal acquired demyelinating symmetric, DADS) or

Asymmetric [multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), Lewis-Sumner syndrome] or

Focal (e.g., involvement of the brachial or lumbosacral plexus or of one or more peripheral nerves in one upper or lower limb)

Pure motor or

Pure sensory (including chronic immune sensory polyradiculopathy affecting the central process of the primary sensory neuron)

(2) Exclusion criteria

Borrelia burgdorferi infection (Lyme disease), diphtheria, drug or toxin exposure probably to have caused the neuropathy

Hereditary demyelinating neuropathy

Prominent sphincter disturbance

Diagnosis of multifocal motor neuropathy

IgM monoclonal gammopathy with high titre antibodies to myelin-associated glycoprotein

Other causes for a demyelinating neuropathy including POEMS syndrome, osteosclerotic myeloma, diabetic and non-diabetic lumbosacral radiculoplexus neuropathy. PNS lymphoma and amyloidosis may occasionally have demyelinating features

Table 1 Electrodiagnostic criteria

(1) Definite: at least one of the following

(a) Motor distal latency prolongation $\geq 50\%$ above ULN in two nerves (excluding median neuropathy at the wrist from carpal tunnel syndrome), or

(b) Reduction of motor conduction velocity $\geq 30\%$ below LLN in two nerves, or

(c) Prolongation of F-wave latency $\geq 30\%$ above ULN in two nerves ($\geq 50\%$ if amplitude of distal negative peak CMAP $< 80\%$ of LLN values), or

(d) Absence of F-waves in two nerves if these nerves have distal negative peak CMAP amplitudes $\geq 20\%$ of LLN + ≥ 1 other demyelinating parameter^a in ≥ 1 other nerve, or

(e) Partial motor conduction block: $\geq 50\%$ amplitude reduction of the proximal negative peak CMAP relative to distal, if distal negative peak CMAP $\geq 20\%$ of LLN, in two nerves, or in one nerve + ≥ 1 other demyelinating parameter^a in ≥ 1 other nerve, or

(f) Abnormal temporal dispersion ($> 30\%$ duration increase between the proximal and distal negative peak CMAP) in ≥ 2 nerves, or

(g) Distal CMAP duration (interval between onset of the first negative peak and return to baseline of the last negative peak) increase in ≥ 1 nerve (median ≥ 6.6 ms, ulnar ≥ 6.7 ms, peroneal ≥ 7.6 ms, tibial ≥ 8.8 ms)^b + ≥ 1 other demyelinating parameter^a in ≥ 1 other nerve

(2) Probable

$\geq 30\%$ amplitude reduction of the proximal negative peak CMAP relative to distal, excluding the posterior tibial nerve, if distal negative peak CMAP $\geq 20\%$ of LLN, in two nerves, or in one nerve + ≥ 1 other demyelinating parameter^a in ≥ 1 other nerve

(3) Possible

As in (1) but in only one nerve

To apply these criteria, the median, ulnar (stimulated below the elbow), peroneal (stimulated below the fibular head), and tibial nerves on one side are tested. If criteria are not fulfilled, the same nerves are tested at the other side, and/or the ulnar and median nerves are stimulated bilaterally at the axilla and at Erb's point. Motor conduction block is not considered in the ulnar nerve across the elbow and at least 50% amplitude reduction between Erb's point and the wrist is required for probable conduction block. Temperatures should be maintained to at least 33°C at the palm and 30°C at the external malleolus (good practice points).

CMAP, compound muscle action potential; ULN, upper limit of normal values; LLN, lower limit of normal values.

^aAny nerve meeting any of the criteria (a-g).

^bIsoe S. *et al.*, in press [16].

Table 5 Supportive criteria

1. Elevated CSF protein with leukocyte count $< 10/\text{mm}^3$ (level A recommendation)
2. MRI showing gadolinium enhancement and/or hypertrophy of the cauda equina, lumbosacral or cervical nerve roots, or the brachial or lumbosacral plexuses (level C recommendation)
3. Abnormal sensory electrophysiology in at least one nerve (good practice points):
 - a. Normal sural with abnormal median (excluding median neuropathy at the wrist from carpal tunnel syndrome) or radial sensory nerve action potential (SNAP) amplitudes; or
 - b. Conduction velocity $< 80\%$ of lower limit of normal ($< 70\%$ if SNAP amplitude $< 80\%$ of lower limit of normal); or
 - c. Delayed somatosensory evoked potentials without central nervous system disease
4. Objective clinical improvement following immunomodulatory treatment (level A recommendation)
5. Nerve biopsy showing unequivocal evidence of demyelination and/or remyelination by electron microscopy or teased fibre analysis (good practice point)
