Electrodiagnostic approach and sub-classification

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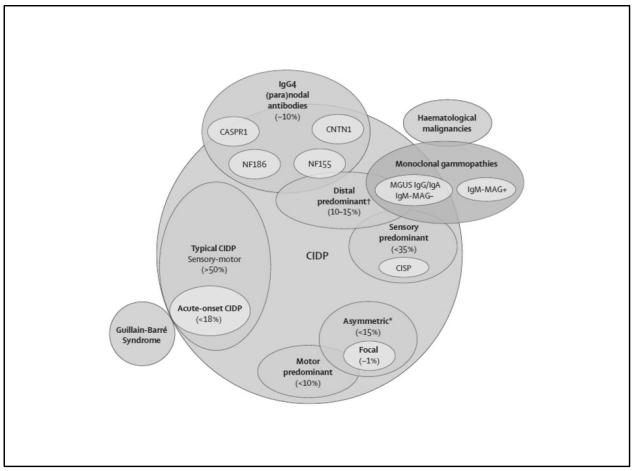


Electrophysiological classification

Acute inflammatory demyelinating polyradiculoneuropathy

(a) Ho *et al*[3]

Patients must have one of the following in two or more nerves during the first 2 weeks of illness:

- Motor conduction velocity <90% lower limit of normal (LLN) (<85% if amplitude of compound muscle action potential after distal stimulation (dCMAP) is <50% LLN)
- Distal motor latency >110% upper limit of normal (ULN) (>120% if dCMAP is <100% LLN)
- Evidence of unequivocal temporal dispersion
- F-response latency >120%

(b) Hadden *et al*[4]

At least one of the following in each of at least two nerves, or at least two of the following in one nerve if all others inexcitable and dCMAP≥10% LLN:

- Motor conduction velocity<90% LLN (<85% if dCMAP is <50% LLN)
- Distal motor latency >110 % ULN (>120% if dCMAP is <100% LLN)
- amplitude of compound muscle action potential after proximal stimulation/dCMAP ratio <0.5 and dCMAP ≥20% LLN
- F-response latency >120%

Acute motor axonal neuropathy

(c) *Ho et al*[3]

- No evidence of demyelination as defined in (a)
- dCMAP <80% LLN

(d) Hadden *et al*[4]

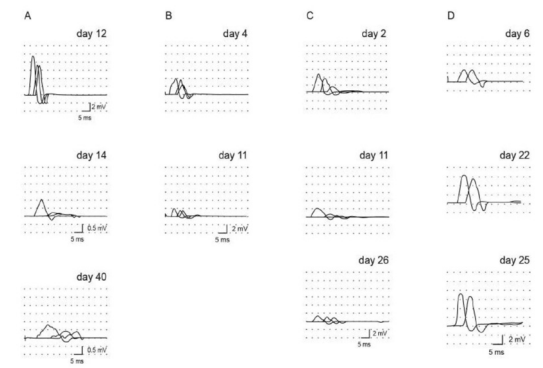
- None of the features of demyelination in any nerve as defined in (b) (except one demyelinating feature allowed in one nerve if dCMAP is <10% LLN)
- dCMAP <80% LLN in at least two nerves

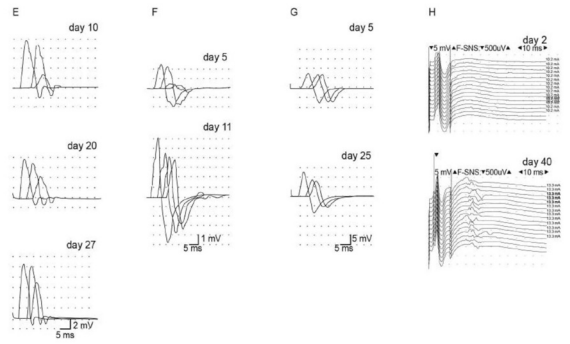
Acute motor and sensory axonal neuropathy

(e) Feasby *et al*,[6] Rees *et al*[17]

- No evidence of demyelination as defined in (a)
- dCMAP <80% LLN in at least two nerves
- Sensory nerve action potential <50% LLN in at least two nerves

Patterns of motor NCS abnormalities in GBS





Rajabally's criteria

Rajabally's criteria

1) Acute inflammatory demyelinating polyneuropathy (AIDP)

At least one of the following in at least two nerves:

- MCV <70% LLN
- DML >150% ULN
- F-response latency >120% ULN, or >150% ULN (if distal CMAP <50% of LLN)
  OR
- F-wave absence in two nerves with dCMAP ≥20% LLN, with an additional parameter, in one other nerve
  OR
- pCMAP/dCMAP amplitude ratio <0.7 (excluding the tibial nerve), in two nerves with an additional parameter, in one other nerve

2) Axonal GBS (including inexcitable forms)

Axonal GBS

None of the above features of demyelination in any nerve (except one demyelinating feature allowed in one nerve if dCMAP <10% LLN), and at least one of the following:

- dCMAP <80% LLN in two nerves
- F-wave absence in two nerves with distal CMAP ≥20% LLN, in absence of any demyelinating feature in any nerve
- pCMAP/dCMAP amplitude ratio <0.7, in two nerves (excluding the tibial nerve
- F-wave absence in one nerve with distal CMAP ≥20% LLN
  OR pCMAP/d CMAP amplitude ratio <0.7 (excluding the tibial nerve), in one nerve; with **IN ADDITION**, dCMAP <80% LLN in one other nerve

Inexcitable

- If dCMAP absent in all nerves (or present in only one nerve with dCMAP <10% LLN)

3) Equivocal

- Abnormal range findings however not fitting criteria for any other group

Uncini's criteria

1) Acute inflammatory demyelinating polyneuropathy (AIDP)

At first or second study at least one of the following in at least two nerves:

- MCV <70% LLN
- DML >130 % ULN
- dCMAP duration >120% ULN
- pCMAP/dCMAP duration >130%
- F-response latency >120% ULN
  OR one of the above in one nerve PLUS:
- Absent F waves in two nerves with dCMAP >20% LLN
- Abnormal ulnar SNAP amplitude and normal sural SNAP amplitude

2) Axonal GBS

Acute motor axonal neuropathy (AMAN)

At first and second study none of the above AIDP features in any nerve (demyelinating features allowed in one nerve if dCMAP <20% LLN)

At first study at least one of the following in each of two nerves:

- dCMAP < 80% LLN
- pCMAP/dCMAP amplitude ratio < 0.7 (excluding tibial nerve)
- isolated F wave absence (or < 20% persistence)

At second study:

at least one of the followings in two nerves is evidence of axonal degeneration:

- persistent or further reduction of dCMAP amplitude
- pCMAP/dCMAP amplitude ratio < 0.7 at first test which recovers because of decrease of dCMAP without increased temporal dispersion (dCMAP duration ≤ 120% ULN and pCMAP/dCMAP duration ratio ≤ 130%)

at least one of the followings in two nerves is evidence of reversible conduction failure:

- >150% increase dCMAP amplitude without increased dCMAP duration (≤120% ULN)
- pCMAP/dCMAP amplitude ratio <0.7 at first test which improves more than 0.2 because of increased pCMAP without temporal dispersion (pCMAP/d CMAP duration ratio ≤ 130%)
- isolated F wave absence (or <20% persistence) that recovers without increased minimal latency (≤120% of ULN)

Uncini's criteria

Acute motor and sensory axonal neuropathy (AMSAN)

At first study

- the same criteria of AMAN in motor nerves
  PLUS
- SNAP amplitudes <50%LLN in at least two nerves

At second study:

- evidence for axonal degeneration and reversible conduction failure in motor nerves as in AMAN
- **there is evidence of axonal degeneration in sensory nerves** if SNAP amplitude in two nerves it is stable or decreased
- **there is evidence of reversible conduction failure in sensory nerves** if SNAP amplitude in two nerves it is increased (>50% in median and ulnar nerves and >60% in sural)

3) Inexcitable

At first or second study

- Distal CMAP absent in all nerves (or present in only one with distal CMAP <10% LLN)

4) Equivocal

At first or second study

- Abnormal findings not fulfilling any of the above criteria

Guillain–Barré syndrome subtypes: A clinical electrophysiological study of 100 patients

A.-M. Grapperon [a,], M. Berro [a], E. Salort-Campana [a], A. Verschueren [a], E. Delmont [a,b], S. Attarian [a,c]*

Table 3 – Electrophysiological classifications of Guillain–Barré syndrome (GBS) patients.

	Hadden criteria	Rajabally criteria
All patients (*n* = 100)		
Axonal	9	41
Demyelinating	52	41
Normal	8	8
Equivocal	31	10
Classic GBS (*n* = 69)		
Axonal	6	29
Demyelinating	38	33
Normal	1	0
Equivocal	24	7
Miller–Fisher syndrome (MFS) (*n* = 8)		
Axonal	0	1
Demyelinating	2	1
Normal	6	6
Equivocal	0	0
Pharyngeal–cervical–brachial (PCB) variant (*n* = 2)		
Axonal	1	1
Demyelinating	0	0
Normal	0	0
Equivocal	1	1
Paraparetic Guillain–Barré syndrome (P-GBS) (*n* = 12)		
Axonal	4	8
Demyelinating	6	2
Normal	0	0
Equivocal	2	2
Unclassified (*n* = 9)		
Axonal	1	3
Demyelinating	6	1
Normal	1	1
Equivocal	1	4

The electrodiagnosis of Guillain-Barré syndrome subtypes: Where do we stand?

Antonino Uncini [a,*], Satoshi Kuwabara [b]

Main differences between the serial studies approach (SSA) and the one study approach (OSA) in electrodiagnosis of Guillain-Barré syndrome (GBS) subtypes.

	No. of studies	Method to assess changes at serial studies	Results of serial studies	Cut-off for RCF in intermediate segments	Association of RCF with GBS subtype and anti-gangliosides Ab
SSA	Serial	Initial diagnosis made with old criteria has been compared with reference diagnosis obtained considering the patient's whole elctrophysiological history and results of anti-gangliosides Ab	22–38% of patients changed subtype	<0.7 p/d CMAP amplitude at first study which improves, at follow-up, more than 0.2	RCF mainly associated with axonal subtypes and antigangliosides Ab
OSA	One	The same criteria sets have been employed at two consecutive studies	Frequencies of subtypes did not change. Subtype diagnosis was dependent on the specific criteria set. 31-35% of individual patients changed subtype	Resolution of CB by at least 30% increase of p/d CMAP amplitude	RCF not associated with GBS subtypes and anti-gangliosides Ab

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CIDP

EFNS diagnostic criteria

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

Joint Task Force of the EFNS and the PNS. Journal of Peripheral Nervous System 15 (2010) 1-9

Table 1. Electrodiagnostic criteria.

(1) Definite: at least one of the following
  (a) Motor distal latency prolongation ≥50% above ULN in two nerves (excluding median neuropathy at the wrist from carpal tunnel syndrome), or
  (b) Reduction of motor conduction velocity ≥30% below LLN in two nerves, or
  (c) Prolongation of F-wave latency ≥30% above ULN in two nerves (≥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 ≥20% of LLN + ≥1 other demyelinating parameter[a] in ≥1 other nerve, or
  (e) Partial motor conduction block: ≥50% amplitude reduction of the proximal negative peak CMAP relative to distal, if distal negative peak CMAP ≥ 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
  ≥30% amplitude reduction of the proximal negative peak CMAP relative to distal, excluding the posterior tibial nerve, if distal negative peak CMAP ≥ 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.

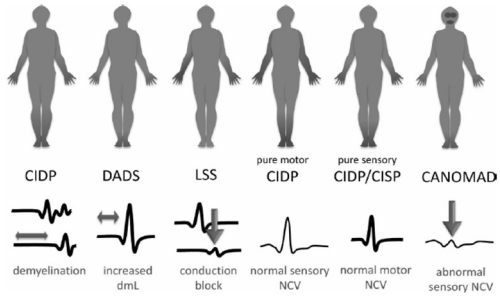
Table 5. Supportive criteria.

1. Elevated CSF protein with leukocyte count <10/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 Points)

Table 6. Diagnostic categories.

Definite CIDP
  Clinical criteria 1 (a or b) and 2 with electrodiagnostic criterion 1; or
  Probable CIDP + at least one supportive criterion; or
  Possible CIDP + at least two supportive criteria
Probable CIDP
  Clinical criteria 1 (a or b) and 2 with electrodiagnostic criterion 2; or
  Possible CIDP + at least one supportive criterion
Possible CIDP
  Clinical criteria 1 (a or b) and 2 with electrodiagnostic criterion 3
CIDP (definite, probable, possible) associated with concomitant diseases.

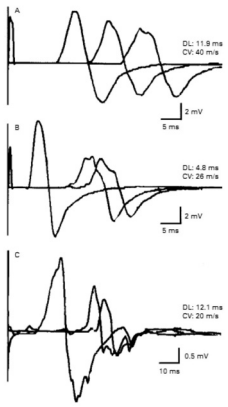
Electrophysiological hallmarks of CIDP subtypes



RESEARCH PAPER

Different electrophysiological profiles and treatment response in 'typical' and 'atypical' chronic inflammatory demyelinating polyneuropathy

Satoshi Kuwabara, Sagiri Isose, Masahiro Mori, Satsuki Mitsuma, Setsu Sawai, Minako Beppu, Yukari Sekiguchi, Sonoko Misawa



Distal pattern

Intermediate pattern

Diffuse pattern

Table 2 Electrophysiology in typical CIDP and MADSAM

	Typical CIDP (n=60)	MADSAM (n=34)	p Value
(A) Motor nerve conduction study: mean (SD)			
Median			
Distal latency (ms)	8.0 (3.1)	4.7 (2.5)	<0.001
Terminal latency index	0.31 (0.13)	0.48 (0.19)	<0.001
CMAP amplitude (mV)	5.0 (3.7)	7.8 (3.4)	<0.001
Proximal/distal ratio	0.74 (0.21)	0.56 (0.30)	0.005
Conduction velocity (m/s)	34.7 (11.5)	37.1 (11.7)	NS
F wave latency* (ms)	50.8 (12.2)	38.2 (18.1)	0.002
Ulnar			
Distal latency (ms)	5.4 (2.0)	4.1 (2.3)	0.006
Terminal latency index	0.38 (0.015)	0.47 (0.017)	0.01
CMAP amplitude (mV)	4.6 (2.5)	6.4 (4.0)	0.02
Proximal/distal ratio	0.71 (0.24)	0.62 (0.26)	NS
Conduction velocity (m/s)	35.9 (13.0)	39.9 (14.9)	NS
F wave latency* (ms)	46.7 (15.2)	43.8 (17.2)	NS
(B) Distribution of demyelination (the EFNS/PNS electrodiagnostic criteria)			
Distal alone	20%	3%	0.02
Intermediate alone	8%	65%	<0.001
Distal and intermediate	43%	11%	<0.001
Unclassified	29%	21%	
(C) Sensory nerve conduction study			
Abnormal median-normal sural response	53%	11%	<0.001

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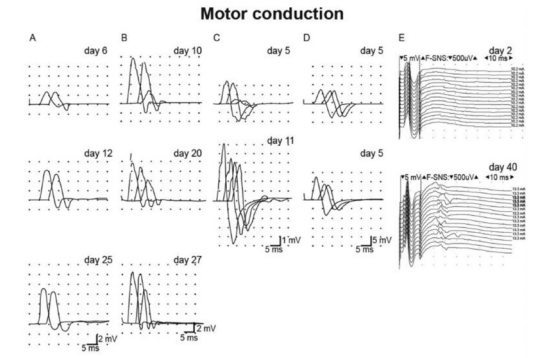
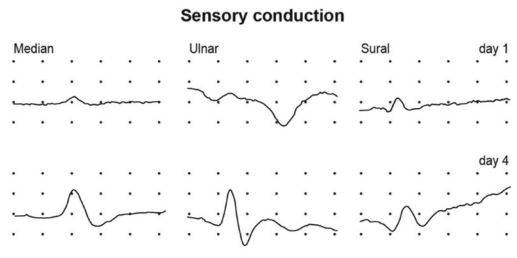
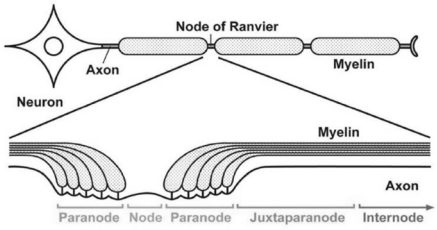
Nodo-paranodopathies

RCF



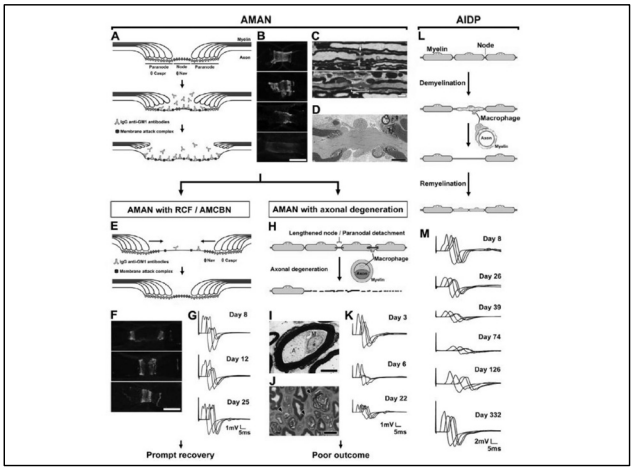


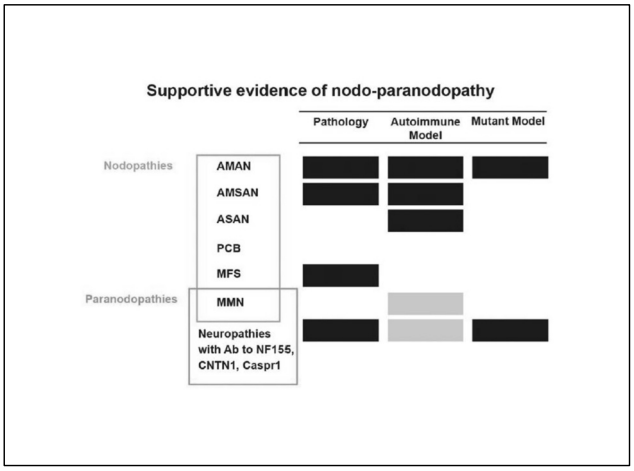
Nodo-paranodopathy



Nodo-paranodopathy: Beyond the demyelinating and axonal classification in anti-ganglioside antibody-mediated neuropathies

Antonino Uncini [a,*], Keiichiro Susuki [b], Nobuhiro Yuki [c]





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