



## May 1997 EMG Case-of-the-Month

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### HISTORY

A 29-year-old right-handed man reports a three-day history of upper and lower limb weakness, moreso in the upper limbs, worse on the left side. He denies pain and numbness. There is no history of trauma, and no bowel or bladder incontinence.

- **Prior to continuing, please develop a differential and list each diagnosis in order of likelihood.**
- **Is there any additional information from the clinical history that might be helpful in clarifying your differential list or changing its order of priority?**

### HISTORY CONTINUED

He denies any other medical problems and takes no regular medications. He did have a viral upper respiratory tract infection starting about 3 weeks ago. He works as a carpenter and denies any occupational exposures.

- **If necessary, please revise your differential diagnosis based on the additional clinical history.**
- **On what details of the physical examination do you think you should focus at this point?**

### PHYSICAL EXAMINATION

Strength examination in the upper limbs (Left/Right using 5 point MRC scale) is: shoulder abduction 2/3, elbow flexion 4-/4, elbow extension 4/4, wrist flexion 4-/4, wrist extension 4-/4, thumb abduction 4/4, dorsal interossei 4-/4. Lower limb strength is 4/5 in all muscles tested at the hip and knee joints; distally ankle dorsiflexion is 3+/4, ankle plantarflexion 4/4 (not tested by standing), ankle eversion 3+/3, ankle inversion 4/4.

Sensation is intact to pinprick and 2 point discrimination throughout the upper and lower limbs. Muscle stretch reflexes are 1+ and symmetric at the biceps and triceps, but absent at the brachioradialis. Lower limb reflexes are 1+ and symmetric at the knees and absent at the ankles. Plantar responses are flexor.

Mental status is intact. Cranial nerve examination is remarkable only for mild facial muscle weakness bilaterally, affecting both upper and lower areas of the face.

- **At this point, review your differential diagnosis and revise as appropriate.**
- **Determine a final working differential diagnosis from which to design your electrodiagnostic study.**
- **Formulate your approach to the electrodiagnostic study.**



**ELECTROPHYSIOLOGIC DATA**

<b>ELECTROMYOGRAPHY</b>										
N = normal incr = increased decr = decreased 0 = absent 1+ = minimal 4+ = maximal crd = complex repetitive discharge fasc = fasciculation potential myk = myokymic discharge myt = myotonic discharge nmt = neuromyotonic discharge p wave = positive sharp waves fibrillation = fibrillation potentials recr = recruitment amp = amplitude dur = duration poly = polyphasic potentials										
R/L	MUSCLE	INSERTION		SPONTAN		VOLUNTARY				
		activ	p wave	fib	other	recr	amp	dur	poly	effort
R	biceps brachii	N	0	0	0	decr	N	N	N	decr
R	1 <sup>st</sup> dorsal interosseous	N	0	0	0	decr	N	N	N	decr
R	tibialis anterior	N	0	0	0	decr	N	N	N	decr
R	medial gastroc	N	0	0	0	decr	N	N	N	decr

<b>SENSORY NERVE CONDUCTION</b>									
nr = no response									
NERVE	LATENCY (ms)			AMPLITUDE (µV)			CONDUC VEL (m/s)		
	R	L	Norm	R	L	Norm	R	L	Norm
ulnar (14cm)	3.0	-	-	59	-	-	-	-	-
median (14cm)	3.2	-	-	33	-	-	-	-	-
sural (14cm)	3.8	-	-	40	-	-	-	-	-

<b>MOTOR NERVE CONDUCTION</b>								
nr = no response								
NERVE	LATENCY (ms)		AMPLITUDE (mV)		CONDUC VEL (m/s)		TEMPORAL DISPERSION	
	R	L	R	L	R	L	R	L
ulnar	-	-	-	-	-	-	-	-
wrist	3.1	-	7.7	-	-	-	-	-
below	7.4	-	5.3	-	47.0	-	-	-



elbow								
above elbow	12.2	-	3.1	-	31.0	-	-	-
axilla	13.4	-	2.6	-	58.0	-	-	-
median	-	-	-	-	-	-	-	-
wrist	4.6	-	7.7	-	-	-	-	-
elbow	8.4	-	6.1	-	53.0	-	-	-
axilla	11.7	-	4.3	-	58.0	-	Y	-
peroneal	-	-	-	-	-	-	-	-
ankle	5.1	-	8.8	-	-	-	-	-
fib. head	10.9	-	4.8	-	49.0	-	-	-
popl. fossa	13.3	-	2.6	-	29.0	-	Y	-

F-WAVE								
# = number of stimuli P = persistence CD = chronodispersion F:M = ratio of average F-wave amplitude to M-wave amplitude								
R/L	NERVE	#	LATENCY (ms)			CD (ms)	P (%)	F:M (%)
			min	mean	max			
R	ulnar to ADM	10	absent	-	-	-	-	-
R	median to APB	10	absent	-	-	-	-	-
R	peroneal to EDB	10	absent	-	-	-	-	-

- **On the basis of the clinical and electrodiagnostic evaluation, formulate your final impression by determining the most likely diagnosis. List other possibilities that are not excluded by the data. Eliminate those diagnoses not supported by the data.**

**DIAGNOSTIC IMPRESSION**

Acquired acute demyelinating peripheral polyneuropathy. Differential diagnosis includes AIDP (acute inflammatory demyelinating polyradiculoneuropathy, or Guillain-Barré syndrome), HIV-associated neuropathy, paraneoplastic syndromes, multiple myeloma or other gammopathies, and some toxins (e.g. arsenic in the acute phase).

**DIFFERENTIAL DIAGNOSIS**

The clinical differential diagnosis of a young person with acute onset of weakness after a viral infection is limited. Given the distal areflexia and absence of long tract signs, the lesion probably lies within the peripheral nervous system rather than centrally. Since there was an acute onset, inherited neuropathies are unlikely. Thus, the differential diagnosis includes



AIDP (either demyelinating or axonal form), infection (HIV, possibly diphtheria), Lyme disease, acute poisoning (arsenic, medications, or other nerve toxins), or a paraneoplastic process.

## FORMULATION OF ELECTROPHYSIOLOGIC STUDIES

While the clinical picture is quite consistent with an acute onset polyneuropathy, clinical information alone cannot distinguish between axonal and demyelinating processes and cannot always pick up subtle sensory lesions. The electromyographic examination is constructed to answer several questions:

1. Is the neuropathy axonal or demyelinating?
2. What is the degree of sensory vs. motor involvement?
3. Is this in fact an acquired rather than inherited neuropathy?
4. What is the prognosis (though it is likely too early to answer this question)?

Nerve conduction studies, examining several segments in each of several nerves, are most useful in demonstrating a demyelinating polyneuropathy. Five features are commonly seen in acquired segmental demyelinating neuropathies:

1. Prolonged distal latencies
2. Slowed conduction velocities, usually patchy
3. Temporal dispersion
4. Conduction block
5. Prolonged or absent F-wave latencies

Inherited neuropathies are somewhat similar but have relatively uniform (as opposed to segmental or patchy) slowing of conduction velocities, and they do not usually have temporal dispersion or conduction block.

Thus, in this patient, the ulnar, median, and peroneal nerves are studied over multiple segments in order to maximize the chance of detecting patchy slowing or conduction block. F-waves are assessed to look for proximal slowing or conduction block. Early after onset of AIDP, F-wave abnormalities may be the only electrodiagnostic findings.

Needle EMG is not expected to be helpful this soon after onset of symptoms, assuming onset of the neuropathy was really three days prior to examination. It is too early for signs of axon loss (fibrillations, positive sharp waves) to be seen. When present, neither the finding of fibrillations, nor their density is reliable at predicting outcome; distal CMAP amplitudes are better. Reduced recruitment, with fewer numbers of rapidly firing motor unit action potentials, is an expected early result of demyelination and conduction block.

Limited needle EMG screen is performed to look for evidence of unexpected prior axon loss.

Prognosis is probably better established at about 3 weeks post onset of symptoms, or at least a week after the nadir of the clinical course, allowing time for axonal degeneration to occur. There are some studies suggesting that distal CMAP amplitudes are correlated with outcome. Specifically, a CMAP amplitude greater than 10% of the lower limit of normal is associated with a good outcome; smaller amplitudes suggest the possibility of a poor outcome.



## DISCUSSION OF ELECTRODIAGNOSTIC FINDINGS AND FORMULATION OF AN IMPRESSION

Several abnormalities stand out in the nerve conduction study results. These include:

1. Prolonged distal latencies in the median and peroneal nerves.
2. Patchy slowing of conduction velocities, including ulnar across the elbow and peroneal nerve from popliteal fossa to fibular head.
3. Temporal dispersion seen in the median and peroneal nerves with proximal stimulation.
4. Conduction block is present in the ulnar nerve (forearm and across elbow), the median nerve (forearm and arm), and peroneal nerve (both segments studied). While the definition of conduction block is controversial, one should be suspicious when drop in amplitude >20% is seen over a 25cm or less segment.
5. All F-waves are absent.

Distal compound muscle action potential amplitudes are normal, however, it is too early to base a good prognostic statement upon these findings.

Sensory nerve studies are normal. In most patients with AIDP, the sural potential is normal while median and ulnar sensory responses are more commonly, but not always, abnormal. Thus the electrodiagnostic medical consultant should not rule out a demyelinating polyneuropathy based upon a normal sural or other sensory potential.

These findings are all consistent with an acute acquired demyelinating neuropathy. While AIDP is at the top of the list, the electrodiagnostic medical consultant cannot conclude, based only upon electrophysiologic testing, that this is AIDP. Thus, if specific diagnoses are mentioned, a listing of other acquired demyelinating neuropathies should be included in the impression as well. The reader is referred to Donofrio and Albers for an excellent summary of the differential diagnoses for motor > sensory acquired demyelinating polyneuropathies as well as other polyneuropathies.

## BIBLIOGRAPHY

1. Asbury AK, Cornblath DR: Assessment of current diagnostic criteria for Guillain-Barré syndrome. *Ann Neurol* 1990;27: (Suppl)S21-S24.
2. Donofrio PD, Albers JW: AAEM Minimonograph #34: Polyneuropathy: Classification by nerve conduction studies and electromyography. *Muscle Nerve* 1990;13:889-903.
3. Miller RG, Peterson GW, Daube JR, et al: Prognostic value of electrodiagnosis in Guillain-Barré. *Muscle Nerve* 1988;11:769-774.