



## EMG CASE No. 67, December 2003

### Presenting Symptom(s):

Asymmetric upper extremity weakness

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**Appropriate Audience:** Residents and practicing physicians

**Learning Objectives:** After completing this educational activity, participant will be able to:  
1) differentiate between the different subtypes of motor neuron disease, 2) define conduction block, and 3) understand how a diagnosis may change over time as new diseases are discovered and described more fully.

**This case is no longer available for CME credit.**

### History

The patient is a 46-year-old male who presents to clinic with a complaint of weakness mostly involving his right arm.

The patient initially was seen in clinic 12 years ago. At that time, he had a 3 year history of "muscle spasms" throughout the right arm which progressed, causing thumb weakness. There was no history of trauma. The patient denied any pain other than that associated with the spasms. He denied any numbness. He also complained of some mild generalized weakness.

- Prior to continuing, please develop a differential diagnosis.
- Is there any additional information regarding the clinical history that might be helpful in clarifying your differential list?

### Differential diagnosis

With the very limited information we have so far, there are numerous possible etiologies including:

Brain lesion

Stroke

Tumor

Spinal cord and nerve root

Myelopathy

Tumor

Radiculopathy

Syrinx

Plexopathy



Tumor  
Brachial Plexitis or Parsonage-Turner syndrome

Motor neuron disease

Amyotrophic lateral sclerosis and its variants  
Spinal muscular atrophy

Peripheral Nerve

Mononeuropathy

Median mononeuropathy  
Ulnar mononeuropathy at the wrist  
Multiple mononeuropathies

Demyelinating motor neuropathy

Multifocal motor neuropathy  
Atypical chronic inflammatory  
demyelinating polyradiculopathy  
Paraneoplastic motor neuropathies

Polyneuropathy

Neuromuscular junction disorders

Myopathy

Muscular dystrophy  
Inflammatory myopathy  
Drug-induced myopathy

### **Commentary I**

Of this rather extended list of possibilities, a few stand out as more likely and a few as improbable.

A peripheral mononeuropathy is a common explanation of asymmetric distal weakness. The most common, entrapment neuropathy involving the upper extremity, a median mononeuropathy at the wrist, will cause weakness of thumb abduction and opposition, and, in some late cases, atrophy of the thenar compartment. Usually sensory symptoms predominate and precede actual weakness, and our patient does not complain of any classic sensory symptoms per se. Common things being common, a median mononeuropathy is high on the list of possibilities. An ulnar mononeuropathy at the wrist, particularly one that involves the distal deep palmar motor branch only, may present with hand weakness without sensory symptoms.



A cervical radiculopathy is also a very common diagnosis and may cause asymmetric weakness involving the upper extremity. Again, our patient does not complain of the pain, paresthesias or other sensory abnormalities that are usually associated with the weakness. However, given the frequency of radiculopathies in general, a C7 radiculopathy is possible in our patient.

Motor neuron disease is much less common, but certainly can present with asymmetric distal upper extremity weakness. Some patients may use the term "spasm" to describe the fasciculations that are common with this disease. This type of disorder, by definition, rarely causes any sensory complaints. The most common form of idiopathic motor neuron disease, amyotrophic lateral sclerosis (ALS), is unlikely as it typically is more rapidly progressive than has been the case for our patient. However, primary lateral sclerosis and progressive muscular atrophy are forms of idiopathic motor neuron disease with more prolonged courses. Polio is a form of infectious motor neuron disease. The weakness is usually asymmetric as in this case, but is preceded by an illness and generally involves the lower extremities before the upper extremities. It would be extremely unlikely in our patient unless he was both not immunized (possible given his age) AND had traveled to an endemic area. HIV is another infectious agent that can produce a motor neuropathy similar to polio. Spinal muscular atrophy Type III and Type IV, and other very, very rare variants, are inherited forms of motor neuron disease and are possible causes of a relatively slowly progressive disorder causing weakness.

Myopathy is another potential cause of weakness that is not associated with prominent sensory complaints. The weakness tends to be symmetric and involve proximal muscles before distal. However, there are forms of myopathy that can be asymmetric such as inclusion body myositis and facioscapulohumeral muscular dystrophy. Types of myopathy that may affect distal muscles preferentially include myotonic dystrophy, distal hereditary myopathy and inclusion body myositis. Myopathy would also be a reasonable explanation of the "mild generalized weakness" mentioned at the end of the history.

A demyelinating motor neuropathy such as multifocal motor neuropathy or atypical chronic inflammatory demyelinating polyradiculopathy (CIDP) could certainly cause asymmetric distal arm weakness that would be slowly progressive. Fasciculations often occur in both of these diseases and may explain our patient's "muscle spasms." These diseases are quite rare, but certainly need to be considered in our patient. A paraneoplastic syndrome is even more rare, but possible as well.

Most of the other possibilities listed are very unlikely. A stroke may cause right hand weakness, but it wouldn't be slowly progressive. A brain tumor could also cause right hand weakness, but typically would progress more quickly. A spinal cord tumor or a myelopathy is pretty unlikely to cause isolated right hand weakness. A syrinx could cause asymmetric hand weakness, but this is usually associated with decreased sensation to pain and temperature and with a deep, relentless pain in the affected area. A plexopathy caused by tumor or a brachial plexitis is very rare, is unlikely to involve just the thumb and would almost certainly cause more prominent sensory complaints. Neuromuscular junction disorders are rare and almost always involve more proximal musculature before distal. Polyneuropathy and multiple mononeuropathies would almost certainly cause more prominent sensory complaints.



**History, continued**

The patient had not traveled outside of the United States and had no risk factors for HIV. He did not smoke. He worked as a mechanic for Dow Chemical.

Physical examination at that time demonstrated normal tone, decreased strength in the distal right upper extremity, 2+ reflexes bilaterally and no sensory deficits. Unfortunately, the notes did not elaborate further.

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

**Commentary II**

The patient has no risk factors for an infectious motor neuron disease and, as a non-smoker, has minimal risk factors for a paraneoplastic syndrome so these can be moved even further down on our list. As a mechanic, he does do a lot of work with his hands, making a mononeuropathy a bit more likely.

The physical examination as described doesn't help us a great deal. The fact that no sensory abnormalities are noted may cause us to put a mononeuropathy or a radiculopathy further down on our list, but the sensory examination is always subjective and often unreliable. Normal reflexes make radiculopathy, myopathy, motor neuron disease and demyelinating motor neuropathy less likely, but all of these certainly can present with normal reflexes initially.

**History, continued**

At that point, the treating physician decided to do an EMG which showed the following:

MOTOR NERVE CONDUCTION STUDIES							
NERVE	SIDE	STIM SITE	RECORD	Cm	AMPL	LAT	CV
Peroneal motor	R	Ankle	EDB	90	5.4	5.6	
	R	Below knee	EDB	325	5.0	12.5	47.1
	L	Ankle	EDB	90	6.1	6.3	
	L	Below knee	EDB	325	5.4	13.3	46.4
	L	Above knee	EDB	100	5.2	14.8	66.7
Tibial motor	L	Ankle	Abd Hal	80	6.3	4.6	



	L	Knee	Abd Hal	470	2.9	16.3	40.2
Ulnar motor	R	Wrist	Hypothenar	70	7.7	3.3	
	R	Below elbow	Hypothenar	265	7.1	7.4	64.6
	R	Above elbow	Hypothenar	110	7.2	9.6	50.0
	L	Wrist	Hypothenar	70	8.9	3.4	
		Below elbow	Hypothenar	250	3.6	8.0	54.3
		Above elbow	Hypothenar	110	3.6	10.2	50.0
Median Motor	L	Wrist	Thenar	70	11.3	3.8	
		elbow	Thenar	261	10.0	8.3	58.0
Peroneal F-resp	R	Ankle	EDB			58.9	
	L	Ankle	EDB			55.5	
Tibial F-resp	L	Ankle	Abd hal			66.6	
Ulnar F-resp	L	Wrist	Hypothenar			32.4	

Temporal dispersion and/or partial conduction block was noted in the left tibial and left ulnar motor responses.

SENSORY NERVE CONDUCTION STUDIES							
NERVE	SIDE	STIM SITE	RECORD	Cm	AMPL	Peak LAT	CV
Sural	L	Calf	Ankle	140	7.8	4.2	42.4
Ulnar	L	Wrist	5 <sup>th</sup>	140	16.6	3.6	53.8
Median	L	Wrist	Index	140	25.4	3.4	53.8



NEEDLE ELECTROMYOGRAPHY									
INSERTional activity: N, sust, unsust									
FIB: 0, 1+, 2+, 3+, 4+									
OTHER: 0 or fascic, myotonia, myokymia									
EFFort: N, decr									
RECRuitment: N, inc or dec 1+, 2+, 3+, 4+									
AMPliitude: N, inc or dec 1+, 2+, 3+, 4+									
DURation: N, inc or dec 1+, 2+, 3+, 4+									
POLYphasia: N, inc or dec 1+, 2+, 3+, 4+									
R/L	MUSCLE	INSER	FIB	OTH	EFF	REC	AMP	DUR	POL
R	Biceps brachii	N	+/-	0	N	Dec 2+	Inc 1+	Inc 1+	Inc 1+
R	Pronator teres	sust	2+	0	N	Dec 3+	Inc 2+	Inc 2+	Inc 2+
R	FDI (hand)	N	+/-	0	N	Dec 2+	Inc 2+	Inc 2+	Inc 1+
R	Ext Digitorum communis	N	0	0	N	Dec 3+	Inc 3+	Inc 2+	Inc 2+
R	Anterior tibialis	sust	2+	0	N	Dec 2+	Inc 4+	Inc 3+	Inc 2+
L	Anterior tibialis	Sust	2+	0	N	Dec 3+	Inc 3+	Inc 3+	Inc 2+
L	Vastus medialis	N	0	0	N	Dec 2+	Inc 2+	Inc 2+	Inc 1+
L	Medial gastrocnemius	N	1+	0	N	Dec 1+	Inc 1+	Inc 1+	N
L	Ext digitorum communis	sust	1+	0	N	Dec 3+	Inc 2+	Inc 2+	Inc 3+
L	Biceps brachii	sust	1+	0	N	N	Inc +/-	N	N
L	Abd pollicis brevis	unsust	+/-	0	N	N	N	N	N
L	Parasp – mid lumbar	N	0	0					

The interpretation of this study was the following: There is electrodiagnostic evidence for a very slowly progressive chronic neurogenic process causing diffuse motor denervation and sparing sensory responses.



## Commentary III

Obviously, our differential diagnosis now must be revised quite a bit. A mononeuropathy or even multiple compressive mononeuropathies could not cause such a diffuse picture. Mononeuritis multiplex could be as diffuse as this, but it usually would cause more prominent sensory symptoms and would cause nerve conduction abnormalities in the sensory as well as in the motor nerves. A radiculopathy, again, is too focal. Multiple radiculopathies might just possibly cause a diffuse picture such as this in a much older patient, but not in our healthy 36 year old. Again, the lack of prominent sensory complaints or findings also makes this very unlikely.

Motor neuron disease is a very likely possibility given the EMG findings of widespread denervation and reinnervation. Unilateral upper extremity wasting and weakness are the first symptoms (as opposed to lower extremity or bulbar symptoms) in 40-60% of patients. However, again, the rate of progression of symptoms in our patient is likely too slow for ALS - progression to death averages 3 years and our patient has had his symptoms for that long. But other forms of motor neuron disease exist.

Progressive bulbar palsy involves the preferential degeneration of bulbar nuclei not associated with either anterior horn cell dysfunction or upper motor neuron signs. It is very rare in its pure form. Face, tongue and neck weakness predominate and lead to dysphagia and dysarthria. This disease tends to be more rapidly progressive than typical ALS and our patient clearly doesn't have this.

Primary lateral sclerosis is a very rare idiopathic disorder marked by progressive upper motor neuron involvement with sparing of the lower motor nerves. Symptoms include spasticity, weakness, increased reflexes, Babinski signs and pseudobulbar speech. Atrophy and fasciculations are less common. The course of this disease tends to be more prolonged than for ALS. This is not very likely in our patient, though, because tone and reflexes are normal.

About 9-15% of patients with idiopathic motor neuron disease present with a pure lower motor neuron syndrome called progressive muscular atrophy (PMA). These patients have distal limb atrophy, fasciculations and cramps with no sensory complaints. Symptoms are commonly asymmetric. Reflexes may be present but usually are reduced in the effected limbs. The clinical course is prolonged with slow progression to the proximal limb muscles. This seems like a possible diagnosis for our patient.

Spinal muscular atrophy Type III, or Kugelberg-Welander disease, is typically a slowly progressive, autosomal recessively inherited disorder. This disease typically presents with symmetric, proximal weakness in patients 3-30 years old. Insidious weakness about the hip girdle progresses to involve the shoulder girdle. Ambulation is maintained for 10-30 years after onset of the disease and the later the onset, the longer the period of ambulation expected. As this disease is typically symmetric and proximal, it is less likely in our patient.

SMA Type IV also usually displays autosomal recessive inheritance. Age of onset is between 30-60 years, with the mean age of initial presentation 35-45 years. This form of SMA typically presents with asymmetric atrophy of the intrinsic hand muscles. Fasciculations are common. During the early phases of the disease, reflexes may be normal, but later they are



decreased or absent. The disease is slowly progressive. Weakness spreads from the hand to the foot intrinsic and distal leg. The shoulder girdle, proximal arm muscles and pelvic girdle are involved late in the course of the disease. This also seems like a possible diagnosis for our patient.

Comparing SMA type IV and PMA, which present with very similar symptoms, patients with SMA type IV generally present younger than those with PMA (mean age of onset in the third and fourth decade as opposed to the fifth). The male to female ratio for SMA Type IV is somewhat higher than for PMA (6-7:1 as opposed to 5.6:1). The mean survival is similar - 7-10 years for SMA Type IV and 7 years for PMA. The most obvious difference between the two disorders is that SMA Type IV shows an inheritance pattern and PMA does not.

There are other very rare forms of inherited motor neuron disease. Monomelic amyotrophy is an autosomal recessively inherited disorder that typically causes a slowly progressive loss of muscle bulk, usually affecting the hand intrinsic muscles of one limb, with actual strength being relatively preserved. The disease occurs almost exclusively in men between the ages of 15 and 30. EMG findings are often bilateral. Distal spinal muscular atrophy is usually an autosomal dominantly inherited disorder that presents in the first 2 decades of life. It has two major forms of presentation, one of which does typically cause bilateral, symmetric or asymmetric hand intrinsic weakness and wasting. Chronic asymmetric spinal muscular atrophy presents as weakness and wasting in either an upper or lower extremity, beginning either distally or proximally. The mean age of onset is 32 years and there is no inheritance pattern. The rate of progression is slow, and typically as the first limb progresses, another limb starts and progresses in the same pattern as the first. Hexosaminidase deficiency is usually inherited as an autosomal recessive disorder and causes lower extremity weakness and cramping. Patients typically present in their second or third decades. Upper extremity strength is normal, but fasciculations and EMG findings are noted in both the upper and lower extremities. Although all of these diseases are possibilities in our patient, they are all very rare, and should go further down on our list of possibilities.

Both idiopathic motor neuron disease and spinal muscular atrophy were suggested as possible diagnoses that could cause the electrodiagnostic picture recorded above, although the latter was thought to be more likely, given the lack of upper motor neuron signs on examination and the chronic nature of the changes on needle examination.

Careful examination of this study reveals that temporal dispersion and/or partial conduction block was noted in the left tibial and left ulnar motor responses. Also the F responses were prolonged. It is unusual for motor conduction velocities and F responses to be abnormal in MND, particularly when they are recorded in muscles that are not clinically involved and atrophic (these findings were on the left side in our patient with right thumb weakness). It was mentioned in the report that these findings might be consistent with the diagnosis of multifocal motor neuropathy. This disease was just beginning to be described back in 1990, which may be why it wasn't further considered at that time.

### **History, continued**

The patient was then lost to follow up for approximately 12 years. During the intervening time, the patient developed increasing weakness in his right arm and new onset of weakness in his left arm. He also had developed mild bilateral foot drop. He complained of cramping involving all of his extremities. He continued to deny bowel or bladder complaints,



double vision, voice change, swallowing difficulty, shortness of breath, neck weakness or sensory changes. Interval past medical history was unremarkable.

**Physical Examination**

Neurologic examination revealed strength to be 5 throughout except the following (R/L): hand intrinsics 3+/4, abductor pollicis brevis 2/5, and anterior tibialis 4+/5-. Reflexes were not obtainable in the right upper extremity and were decreased to the same degree but present everywhere else. Sensory examination was intact to light touch, pinprick, proprioception and vibration throughout. Dupuytren's contractures were noted in the right fourth finger and the left third finger. Fasciculations were not noted. Physical examination was otherwise completely normal.

- At this point, review your differential diagnosis and revise as appropriate.

**Commentary IV**

Both SMA Type IV and PMA are still possible diagnoses, given the clinical progression. However, looking at his old EMG 12 years later, the diagnosis of multifocal motor neuropathy had to be further considered. A repeat EMG was ordered.

**Electrophysiologic Data**

MOTOR NERVE CONDUCTION STUDIES							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL	LAT	CV
Peroneal motor	R	Ankle	EDB	90	3.5	5.2	
	R	Below knee	EDB	310	2.4	13.4	37.8
	L	Ankle	EDB	90	2.2	5.7	
	L	Below knee	EDB	300	1.8	13.6	38.0
Tibial motor	R	Ankle	Abd hal	80	3.2	4.6	
	L	Ankle	Abd hal	80	3.0	4.5	
Median motor	R	Wrist	Thenar	70	3.3	5.6	
	R	Elbow	Thenar	260	0.5	13.5	32.9
	L	Wrist	Thenar	70	7.0	3.6	
	L	Elbow	Thenar	250	5.0	8.4	52.1
Ulnar motor	R	Wrist	Hypothenar	70	8.0	3.7	
	R	Below elbow	Hypothenar	220	2.6	14.3	20.8
	L	Wrist	Hypothenar	70	6.2	3.4	



	L	Below elbow	Hypothenar	220	1.2	9.5	36.1
Tibial F-response	R					NR	
Median F-response	R					62.3	

SENSORY NERVE CONDUCTION STUDIES							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL	LAT	CV
Sural	L	Calf	Ankle	140	15.7	3.3	42.4
Median	R	Wrist	Index	140	23.4	2.9	63.6
Ulnar	R	Wrist	5th	140	15.0	3.3	53.8

NEEDLE ELECTROMYOGRAPHY									
INSERtional activity: N, sust, unsust									
FIB: 0, 1+, 2+, 3+, 4+									
OTHER: 0 or fascic, myotonia, myokymia									
EFFort: N, decr									
RECRuitment: N, inc or dec 1+, 2+, 3+, 4+									
AMPLitude: N, inc or dec 1+, 2+, 3+, 4+									
DURation: N, inc or dec 1+, 2+, 3+, 4+									
POLYphasia: N, inc or dec 1+, 2+, 3+, 4+									
R/L	MUSCLE	INSER	FIB	OTH	EFF	REC	AMP	DUR	POL
R	Anterior tibialis	Sust	2+	0	N	Dec 1+	Inc 1+	Inc 1+	Inc 2+
R	Medial gastrocnemius	N	0	0	N	N	N	N	N
R	Vastus lateralis	N	0	0	N	Dec 1+	Inc 2+	Inc 2+	Inc 1+
R	FDI (hand)	Sust	3+	crd	N	Dec 3+	Inc 1+	Inc 1+	Inc 1+
R	Abd pollicis brevis	Sust	3+	0	N	Dec 4+	0	0	0
R	Biceps brachii	N	0	0	N	N	N	N	N



- On the basis of both the clinical and electrophysiologic evaluations, formulate your diagnostic impression. List the most likely diagnosis first and follow in order with the other possibilities that are not excluded by the data. Eliminate those diagnoses not supported by the data.
- Are there additional electrophysiologic data that you feel would further delineate the diagnosis? (Remember, collecting data that are not needed for the diagnosis is costly and uncomfortable for the patient.)

### **Diagnostic Impression**

The widespread denervation/reinnervation combined with the definite conduction block noted in the right median, right peroneal, and both ulnar motor nerves supports the diagnosis of a multifocal demyelinating neuropathy. The absent tibial F response and prolonged median F response also support the diagnosis of a demyelinating neuropathy. The normal sensory conduction studies make the diagnosis of multifocal motor neuropathy more likely than CIDP.

### **Commentary V**

In 1982, Lewis et al. first reported five patients with a chronic, asymmetric, motor and sensory neuropathy more pronounced in the upper limbs than lower limbs. Electrophysiologically, it was characterized by persistent multifocal conduction blocks in motor but not sensory nerves. The authors considered it an asymmetric form of CIDP. During 1985-86, four patients were reported with a chronic, asymmetric pure motor neuropathy without, or with little, sensory impairment. Again this was associated with persistent multifocal partial motor conduction block. Since then, more than 300 patients with what is now called multifocal motor neuropathy (MMN) have been reported.

The disease causes asymmetric weakness in the extremities, with the arms usually being affected earlier and more severely than the legs. The weakness is often related to the distribution of individual nerves and fasciculations and cramping are common. Atrophy of the involved muscles occurs as the disease progresses. Reflexes are often reduced in a patchy distribution which usually coincides with the areas of weakness. Cranial nerve involvement and phrenic nerve palsy are extremely rare. Although most patients have impaired ADLs because of decreased dexterity or fatigue, only 2 deaths related to the disease have been reported. Steadily progressive, step wise or spontaneously remitting course have all been described. The actual prevalence of MMN is unknown, but 1-2 cases per 100,000 have been estimated. The mean age of onset is 40 years, and 80% present with first symptoms between 20 and 50 years. The male:female ratio is 2.6:1.

Electrodiagnostically, it is characterized by the presence of persistent multifocal, partial conduction block (CB) in motor nerves outside the usual sites of nerve compression. The CB in MMN is most frequent in the ulnar, median and peroneal nerves and can be very proximal, requiring nerve root stimulation. The conduction velocity is often normal, or only mildly reduced, even at the level of the CB, which is evidence of how focal the demyelination is. Mildly prolonged F-responses are fairly common but the sensory responses are usually normal.



Conduction block (CB) has been defined as a reduction in the CMAP amplitude or area obtained by proximal versus distal stimulation of motor nerves, in absence of, or with only focal, abnormal temporal dispersion. The degree of reduction that is necessary to be considered CB varies from 20-50% in the literature, but the AAEM has reached the following consensus:

Definite CB requires a minimum of 50% reduction in CMAP amplitude in the nerves of the upper extremity, and 60% in the nerves of the lower extremity, in the absence of abnormal temporal dispersion.

Probable CB requires a similar reduction of amplitude with increased temporal dispersion, or 10% less reduction without increased temporal dispersion.

These criteria have been imposed to separate real CB from the apparent CB that can be observed in chronic demyelination or axonal loss, where a markedly increased range of conduction velocities, or increased polyphasia, with reduced number of motor unit potentials may result in overlap and cancellation of the positive and negative components of different motor unit action potentials. This may lead to reduced proximal CMAP amplitude that can mimic a true CB and is called phase cancellation. However, these stringent criteria may lead to a delay in the diagnosis of a treatable disease, so some authors have suggested that CB not be mandatory for the diagnosis of MMN in patients with a typical presentation.

Other laboratory findings include a serum creatinine kinase that is slightly to moderately elevated in up to 2/3 of patients. Protein in the CSF may be slightly increased (up to 80 mg/dl). Serum protein electrophoresis may reveal a monoclonal gammopathy, mostly of the IgM type. The presence of serum antibodies, mostly IgM, to the ganglioside GM1, has been found in 30-60% of patients and helps to support the diagnosis of MMN. GM1 is found throughout the central and peripheral nervous system. It is found at the level of the nodes of Ranvier, outer myelin, and motor end plate. It is more highly concentrated in motor than in sensory nerve myelin, which may help explain why the motor nerves are primarily affected in MMN. The positive predictive value of serum IgM anti-GM1 antibodies was 92% when the test was used to help distinguish between patients with MND and those with MMN (as we've seen, one of the main diagnostic dilemmas with MMN). Our patient did not have anti-GM1 antibodies in his serum.

There have been 4 randomized, double-blinded, placebo controlled, cross over studies that have confirmed the effectiveness of intravenous immunoglobulin (IVIg) in the treatment of MMN. Almost 80% of the 170 reported patients treated with IVIg improved with this therapy (some studies used improvement in motor strength, while other studies used improvement in disability score as their outcome measure). In most responding patients, improvement occurs within a week of therapy, but it only lasts a few weeks and has to be maintained with periodic IVIg infusions. Improvement is greatest in the most recently involved areas, with minor to no effect on stabilized deficits. The reduction or resolution of CB has been shown in some, but not all, the nerves in patients treated with IVIg. However, there is minimal to no effect on anti-ganglioside antibodies if they are even present. Some patients appear to become resistant to IVIg, but often will respond again if the dosage is increased or if the time interval between infusions is decreased. Our patient has been



treated with IVIG. The progression of his disease has been halted and he has regained some of the strength in his lower extremities. His right hand remains weak and atrophied.

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