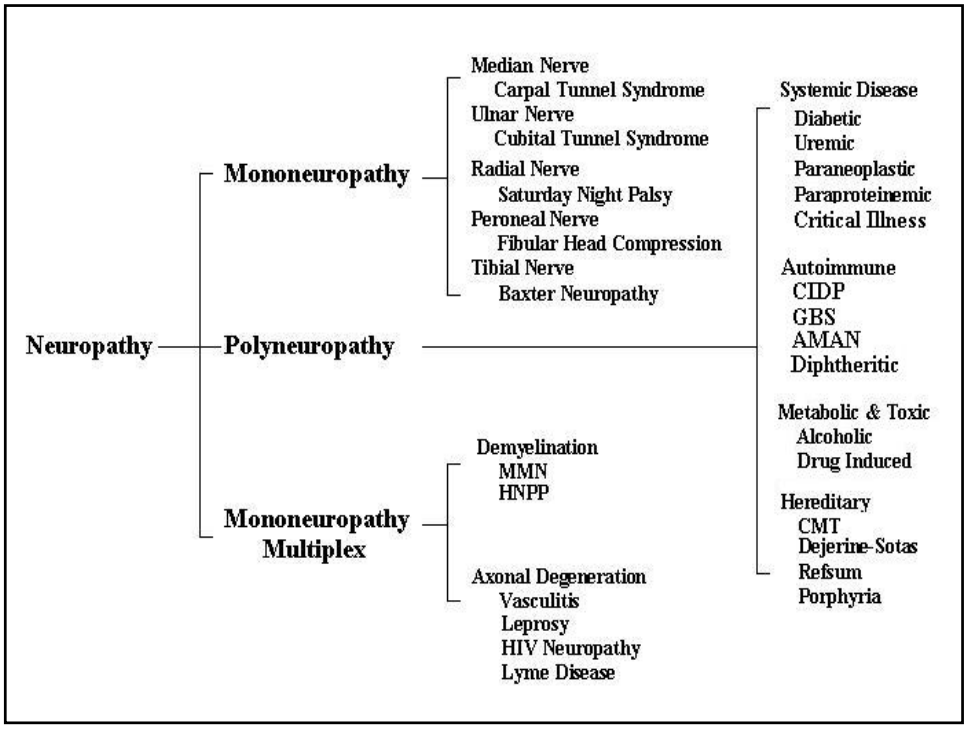



## Different Dx of Neuropathy

Neuronopathy
- Anterior horn cell
- Dorsal ganglion cell
- Autonomic ganglion

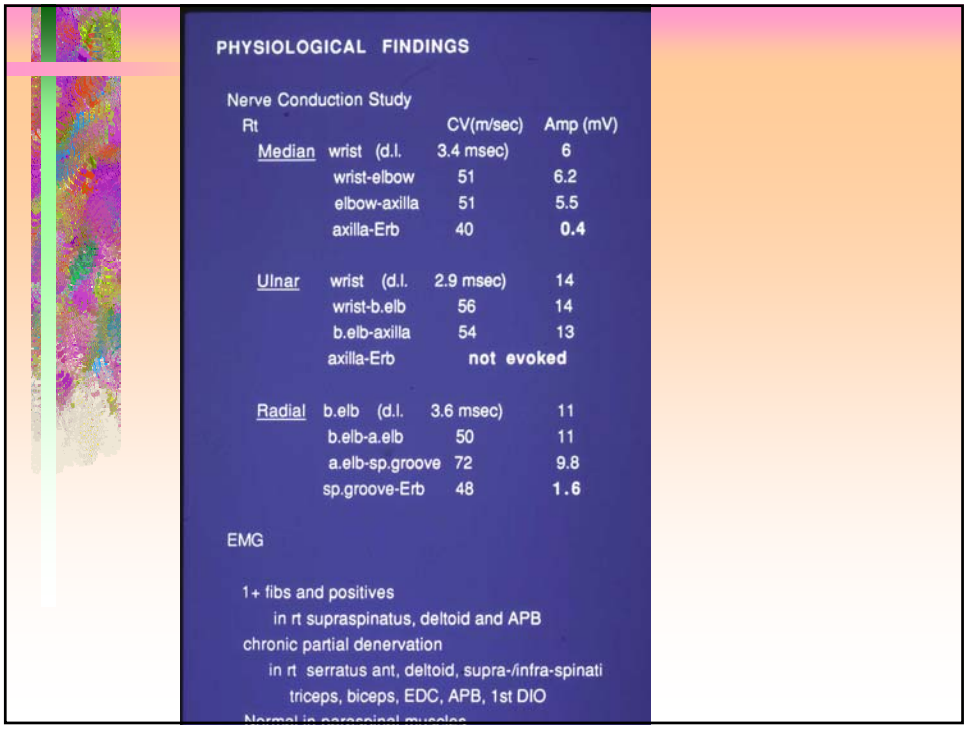
Axonopathy
- dying back axonopathy (fast transport)
- axonal atrophy (slow transport)
- giantaxonal neuropathy
- central distal axonopathy

Myelinopathy
- demyelinating
- hypomyelinating

症例　　４９歳　男性　　設計技師

主訴：右上肢脱力

２年前より次第に右上肢の挙上困難が出現、
次第に設計のためPenを持つのも困難になって来た。
同時に、右上肢の筋萎縮も出現、
某大学病院でALSの診断を受ける。



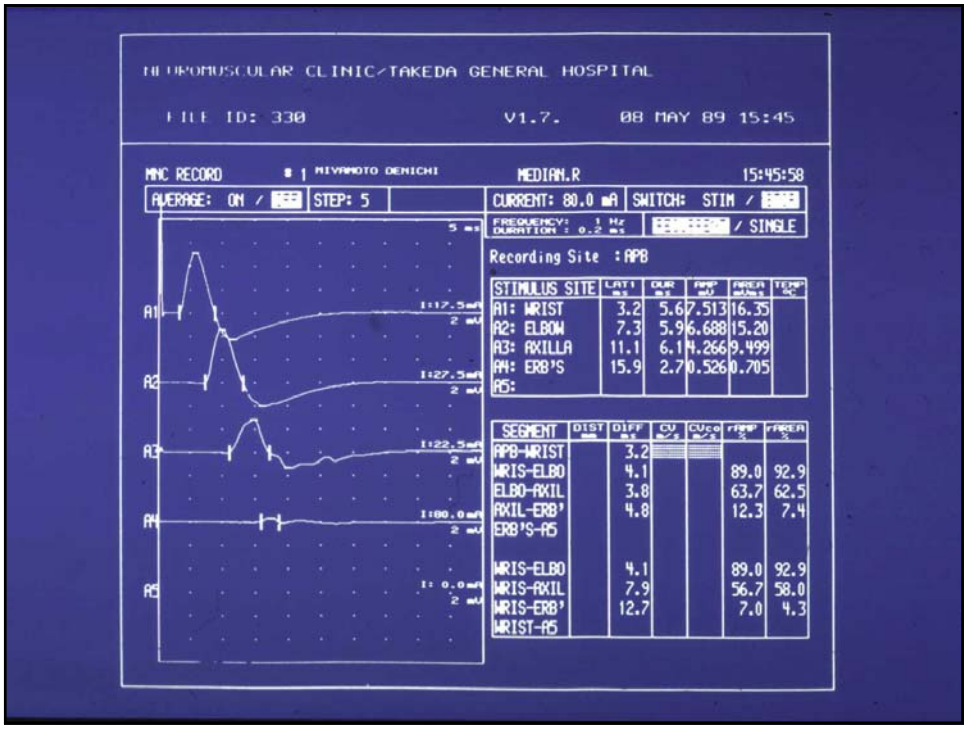
PHYSIOLOGICAL FINDINGS

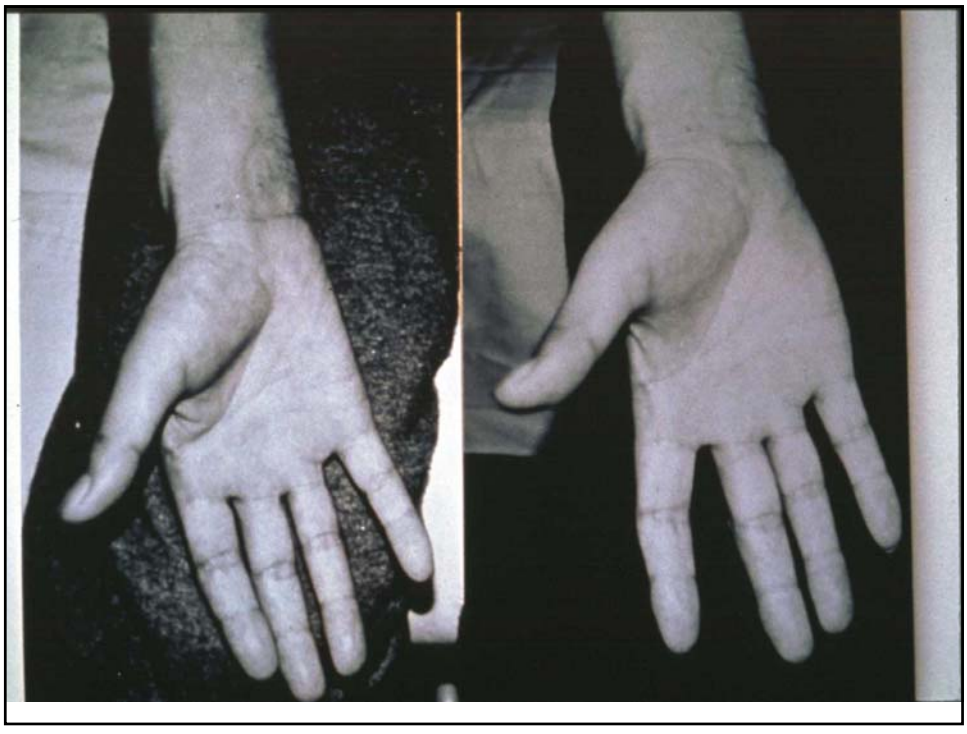
Nerve Conduction Study

| Rt | | CV(m/sec) | Amp (mV) |
|---|---|---|---|
| Median | wrist (d.l. | 3.4 msec) | 6 |
| | wrist-elbow | 51 | 6.2 |
| | elbow-axilla | 51 | 5.5 |
| | axilla-Erb | 40 | 0.4 |
| Ulnar | wrist (d.l. | 2.9 msec) | 14 |
| | wrist-b.elb | 56 | 14 |
| | b.elb-axilla | 54 | 13 |
| | axilla-Erb | not evoked | |
| Radial | b.elb (d.l. | 3.6 msec) | 11 |
| | b.elb-a.elb | 50 | 11 |
| | a.elb-sp.groove | 72 | 9.8 |
| | sp.groove-Erb | 48 | 1.6 |

EMG

1+ fibs and positives
in rt supraspinatus, deltoid and APB
chronic partial denervation
in rt serratus ant, deltoid, supra-/infra-spinati
triceps, biceps, EDC, APB, 1st DIO
Normal in paraspinal muscles


NEUROMUSCULAR CLINIC/TAKEDA GENERAL HOSPITAL
FILE ID: 330
V1.7.
08 MAY 89 15:45
MNC RECORD
MIYAMOTO DENICHI
MEDIAN.R
15:45:58
AVERAGE: ON / OFF
STEP: 5
CURRENT: 80.0 mA
SWITCH: STIM / REC
FREQUENCY: 1 Hz
DURATION : 0.2 ms
/ SINGLE
Recording Site : APB
STIMULUS SITE
A1: WRIST 3.2 5.6 7.513 16.35
A2: ELBOW 7.3 5.9 6.688 15.20
A3: AXILLA 11.1 6.1 4.266 9.499
A4: ERB'S 15.9 2.7 0.526 0.705
A5:
SEGMENT
APB-WRIST 3.2
WRIS-ELBO 4.1 89.0 92.9
ELBO-AXIL 3.8 63.7 62.5
AXIL-ERB' 4.8 12.3 7.4
ERB'S-A5
WRIS-ELBO 4.1 89.0 92.9
WRIS-AXIL 7.9 56.7 58.0
WRIS-ERB' 12.7 7.0 4.3
WRIST-A5



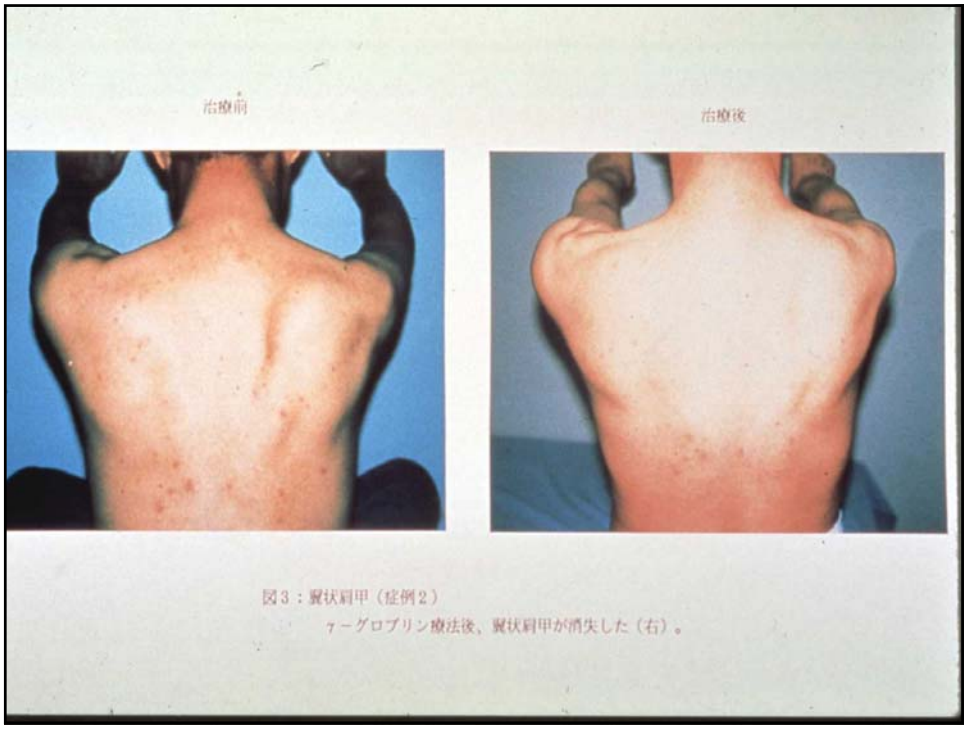
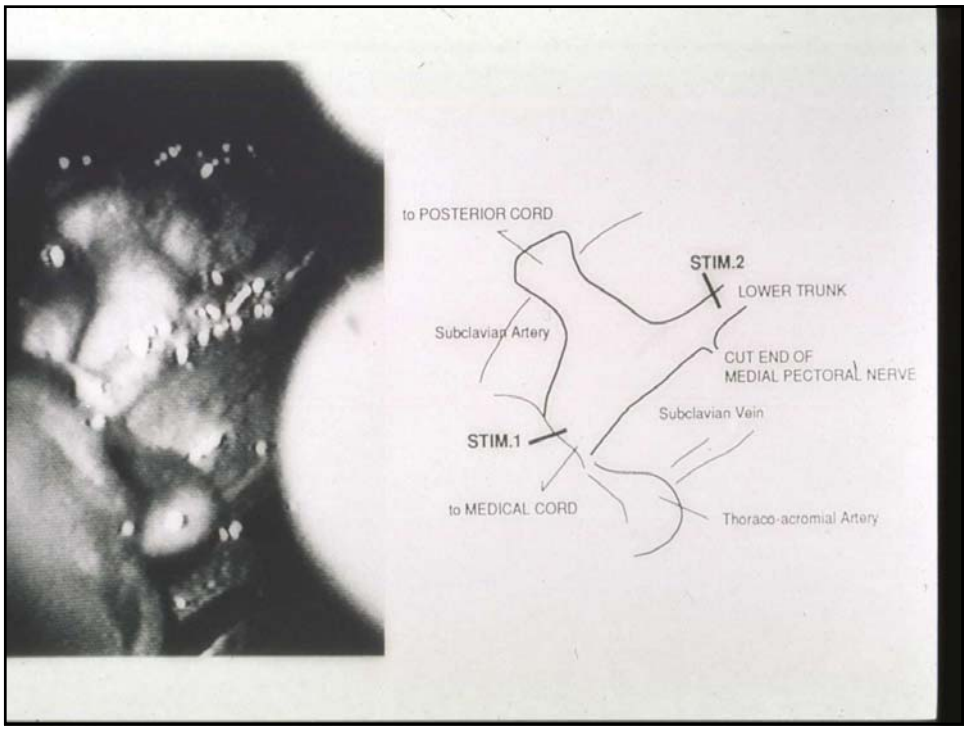
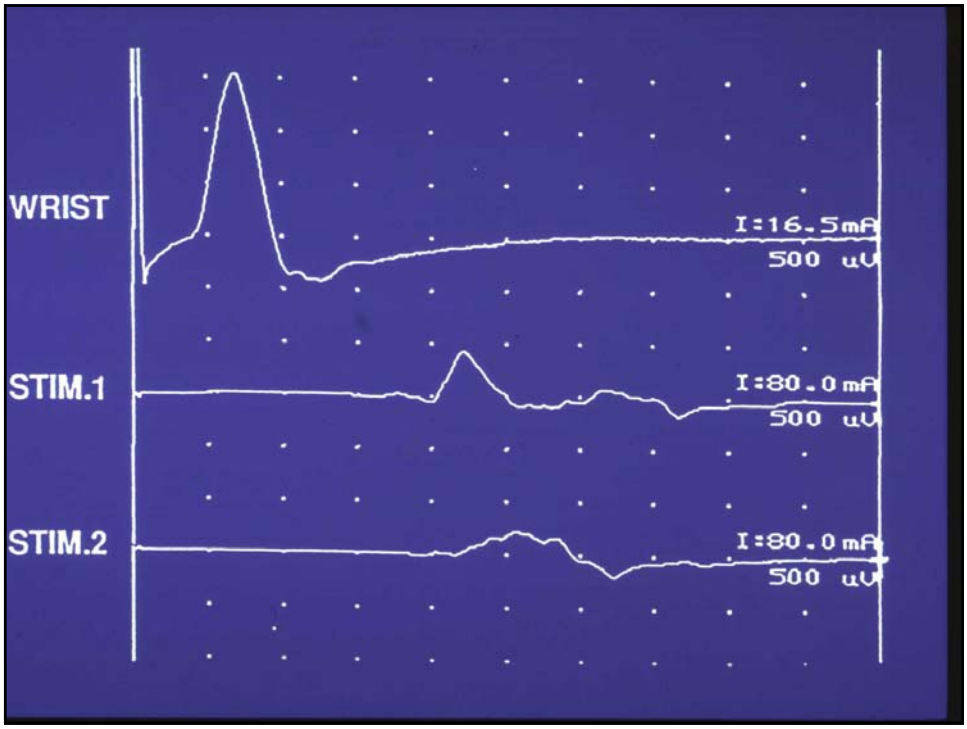
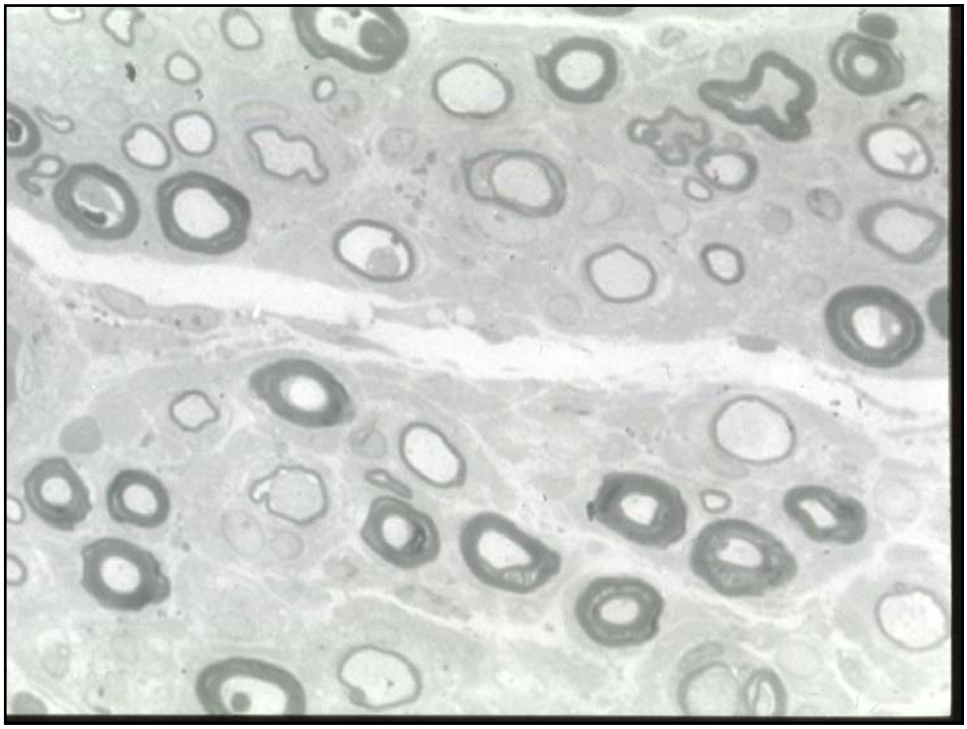



図3：翼状肩甲（症例2）
γ－グロブリン療法後、翼状肩甲が消失した（右）。




WRIST
I:16.5mA
500 uV
STIM.1
I:80.0 mA
500 uV
STIM.2
I:80.0 mA
500 uV



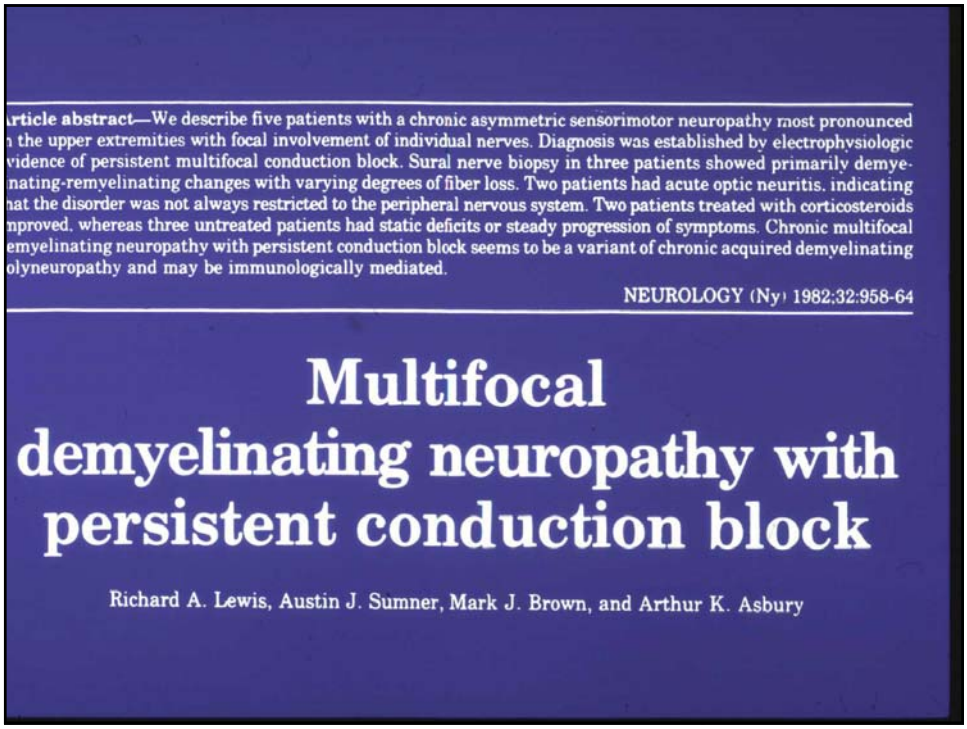
PHYSIOLOGICAL CONSEQUENCES OF DEMYELINATION

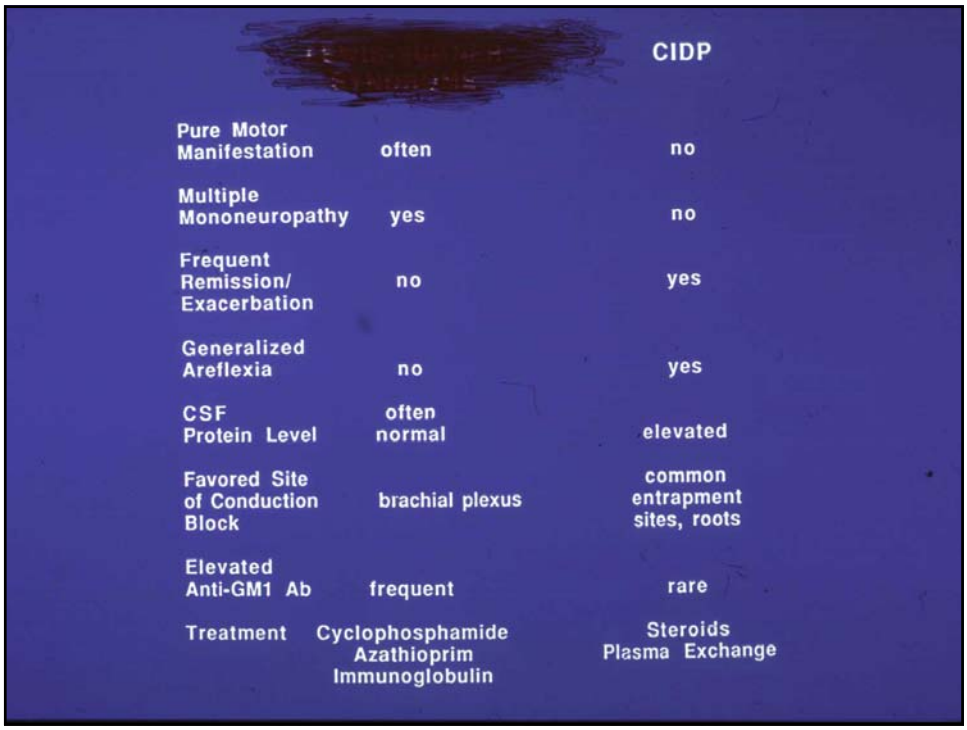
1. COMPLETE CONDUCTION BLOCK

2. SLOWED CONDUCTION

3. FAILURE TO TRANSMIT HIGH-FREQUENCY IMPULSES (RATE-DEPENDENT BLOCK)
4. ECTOPIC IMPULSE GENERATION

5. EPHAPTIC TRANSMISSION


rticle abstract—We describe five patients with a chronic asymmetric sensorimotor neuropathy most pronounced
the upper extremities with focal involvement of individual nerves. Diagnosis was established by electrophysiologic
vidence of persistent multifocal conduction block. Sural nerve biopsy in three patients showed primarily demye-
nating-remyelinating changes with varying degrees of fiber loss. Two patients had acute optic neuritis, indicating
hat the disorder was not always restricted to the peripheral nervous system. Two patients treated with corticosteroids
nproved, whereas three untreated patients had static deficits or steady progression of symptoms. Chronic multifocal
emyelinating neuropathy with persistent conduction block seems to be a variant of chronic acquired demyelinating
olyneuropathy and may be immunologically mediated.
NEUROLOGY (Ny) 1982;32:958-64
Multifocal demyelinating neuropathy with persistent conduction block
Richard A. Lewis, Austin J. Sumner, Mark J. Brown, and Arthur K. Asbury


CIDP
Pure Motor Manifestation
often
no
Multiple Mononeuropathy
yes
no
Frequent Remission/ Exacerbation
no
yes
Generalized Areflexia
no
yes
CSF Protein Level
often normal
elevated
Favored Site of Conduction Block
brachial plexus
common entrapment sites, roots
Elevated Anti-GM1 Ab
frequent
rare
Treatment
Cyclophosphamide Azathioprim Immunoglobulin
Steroids Plasma Exchange

## HISTORY

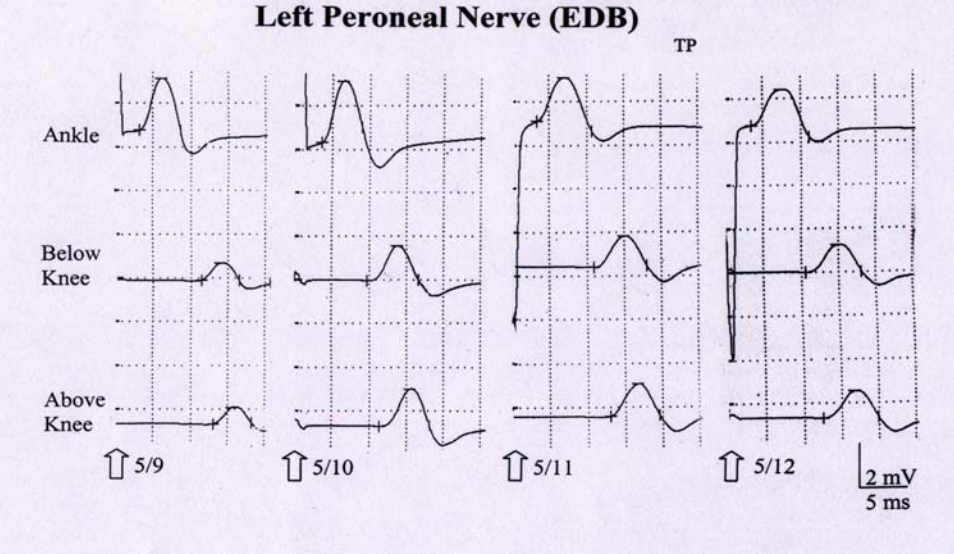
A 21 year old white male, known asthmatic, developed purpura on palms and soles 4 days PTA followed, 2 days later, by lower limb weakness and loss of balance. Hx of respiratory infection and diarrhea.

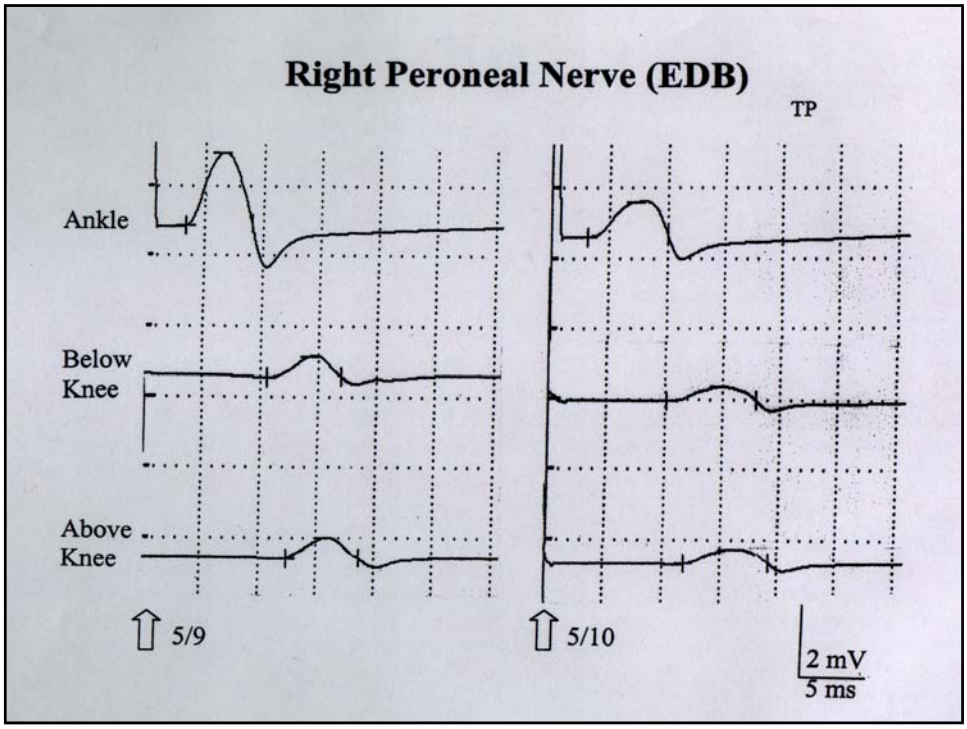
## Physical Exam Day 1

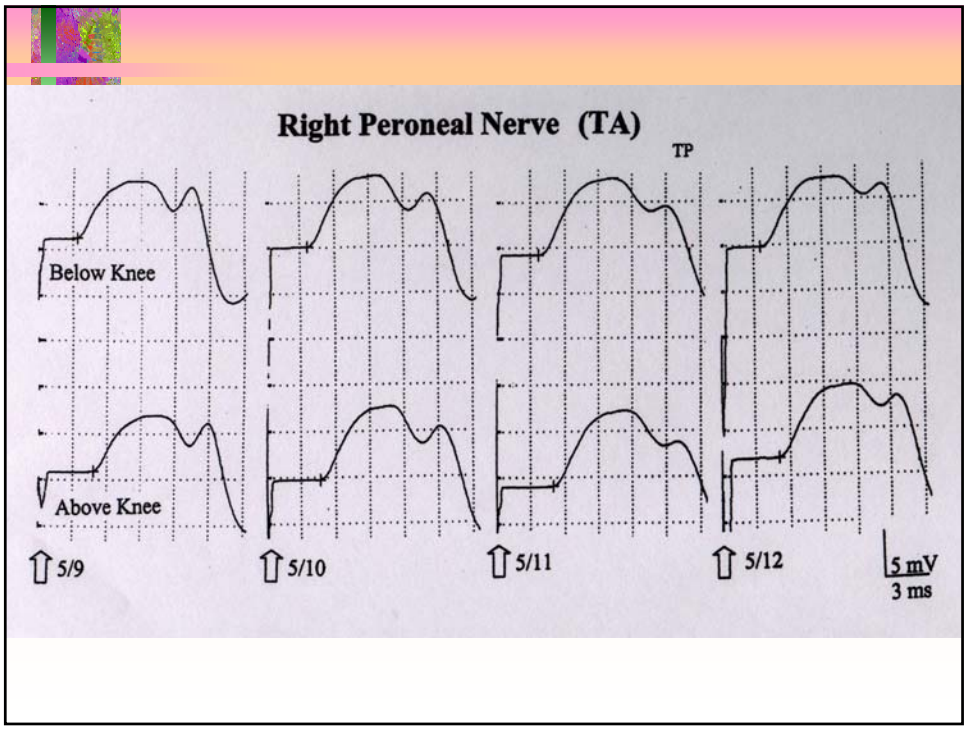
- Purpura/petechiae palms and dorsal aspects of feet
- Grips right 3/5, left 3-4/5
- Plantar Flexion right 4/5, left 5/5
- Reflex - knee 0/2, ankle 0/2
- Unable to toe walk

# Physical Exam Day 2

- Unable to stand unassisted
- Unable to lean forward or backward
- Unable to adduct right eye
- Loss of strength trunk and deltoid
- Loss of reflex left upper limb

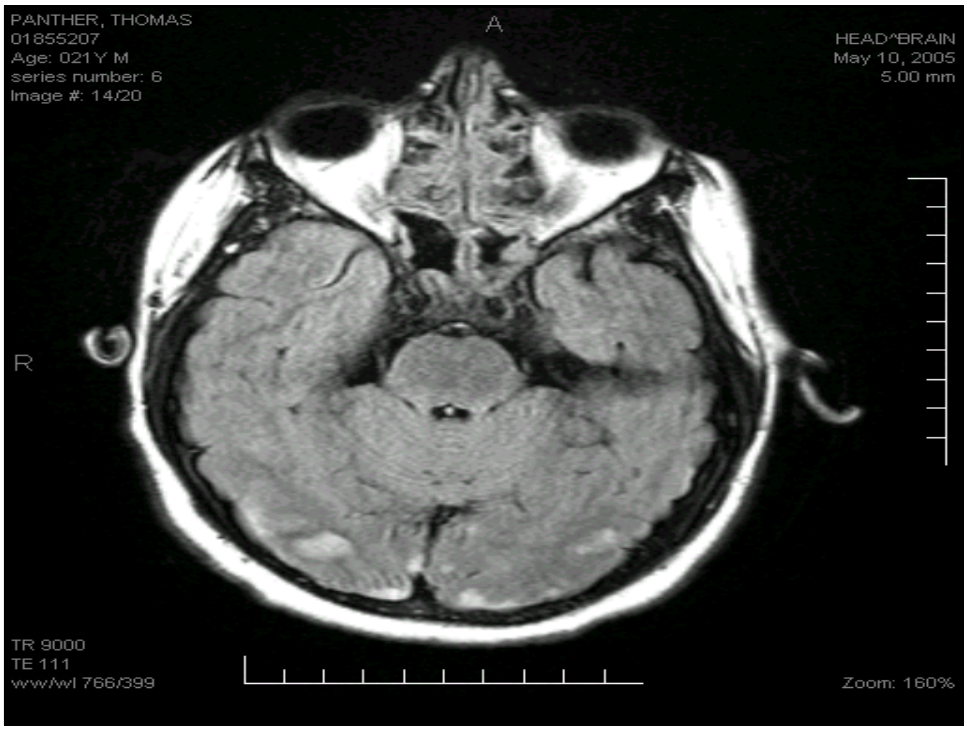



Right Peroneal Nerve (EDB)
TP
Ankle
Below Knee
Above Knee
5/9
5/10
2 mV
5 ms


Right Peroneal Nerve (TA)
TP
Below Knee
Above Knee
5/9
5/10
5/11
5/12
5 mV
3 ms

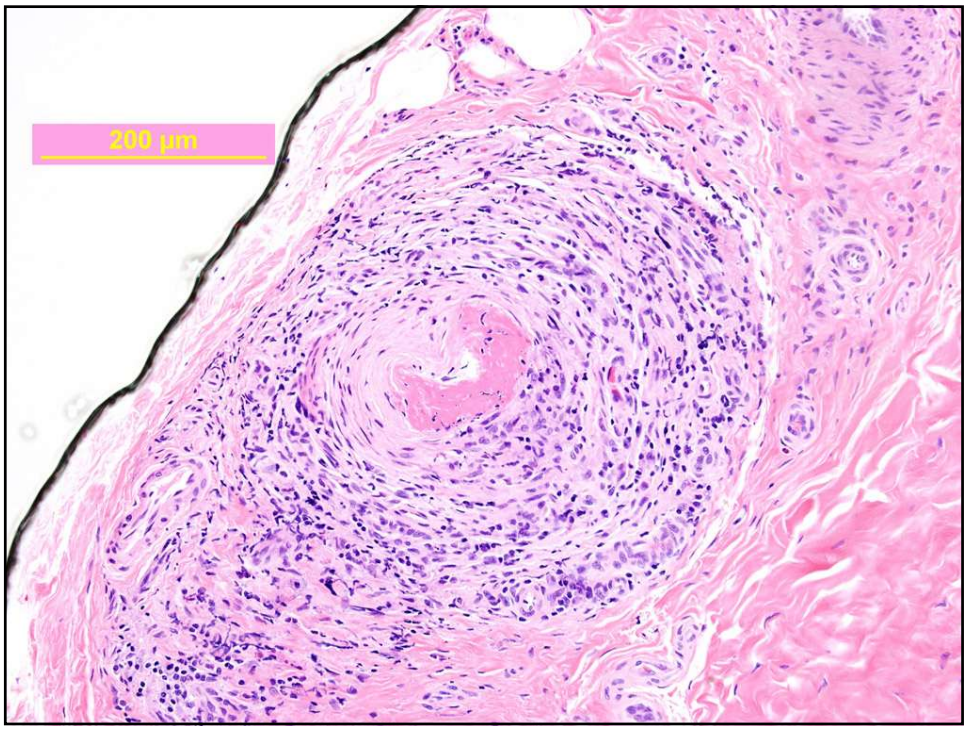
## MRI

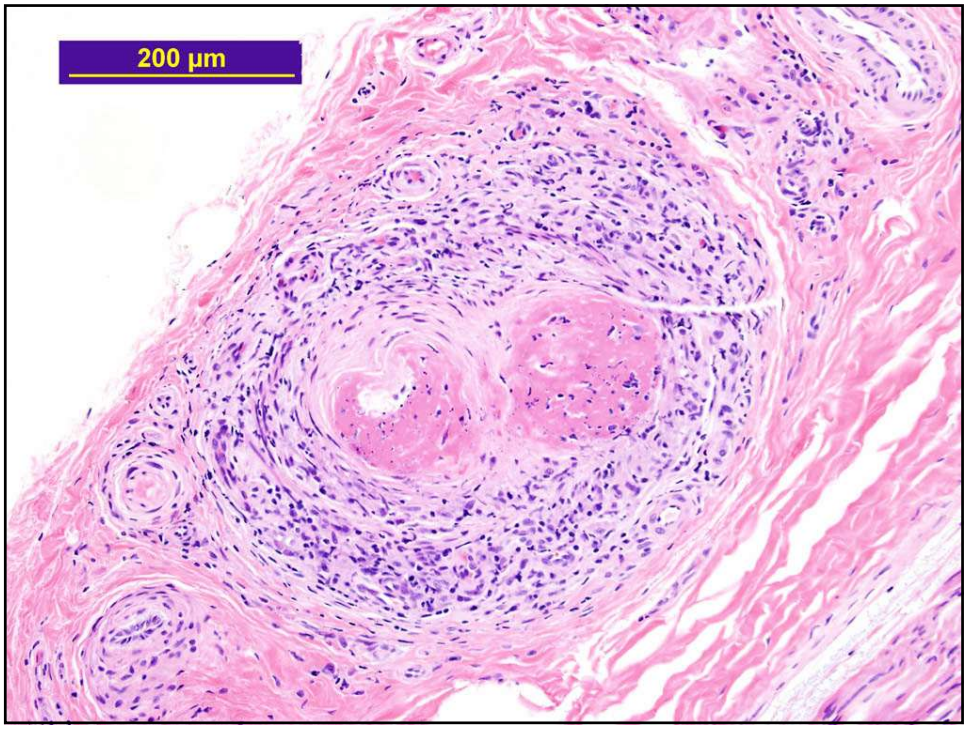
**Multiple variable sized nonenhancing areas of T2 and FLAIR hyperintensity scattered in both cerebral and cerebellar hemispheres and pons, consistent with acute infarcts.**



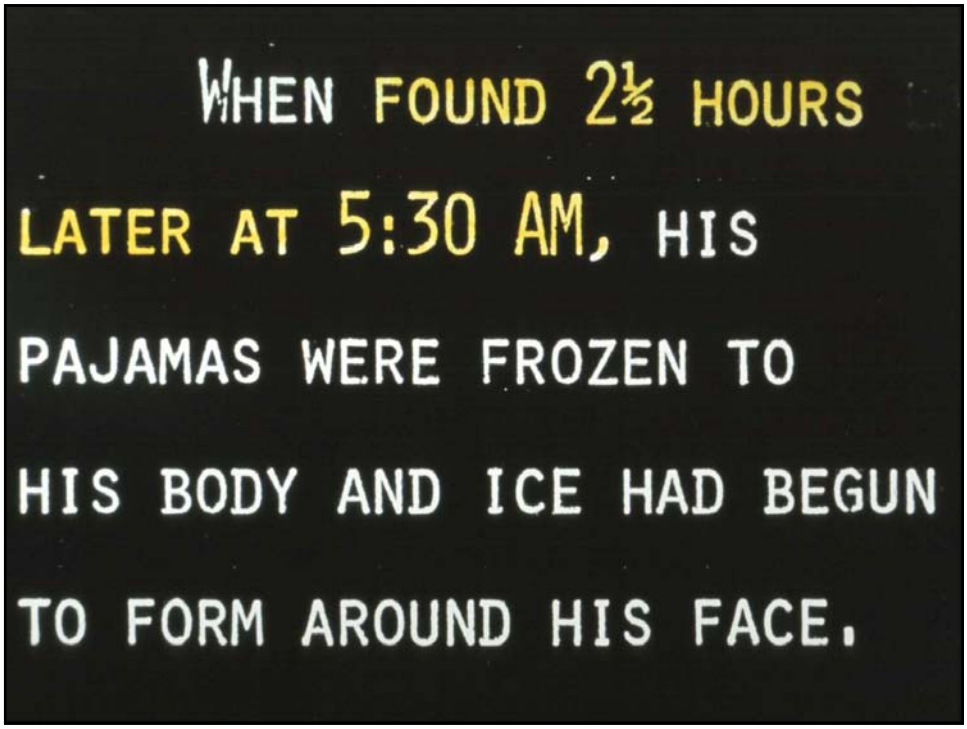
## Differential Diagnosis

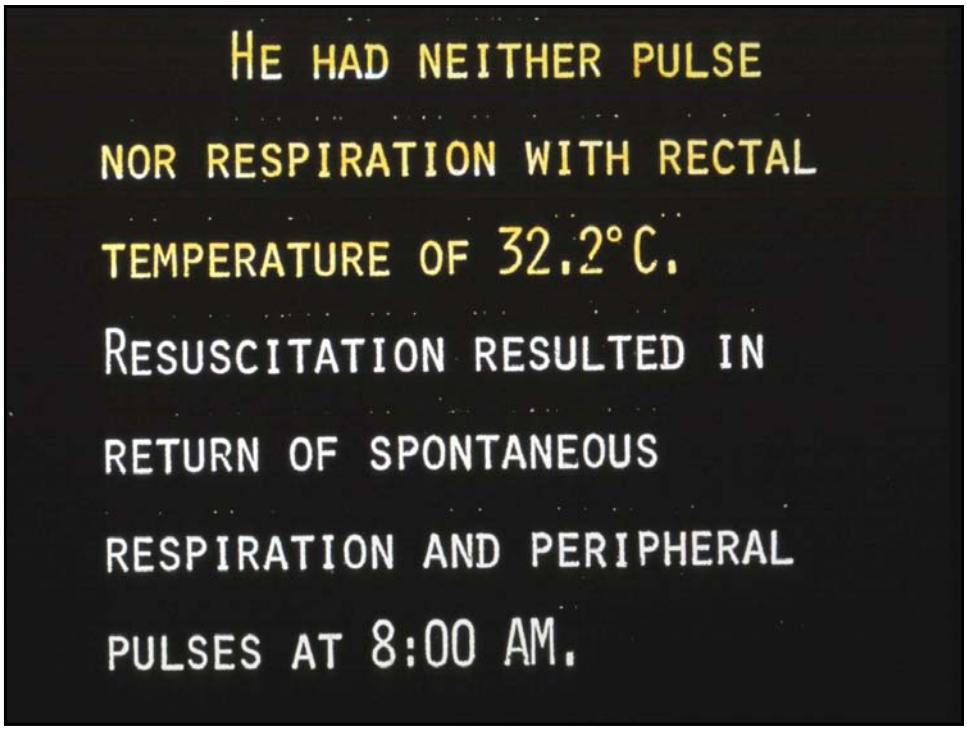
- Guillain Barré Syndrome
- Mononeuritis multiplex
- Churg Strauss vasculitis
- Multiple ischemic infarcts





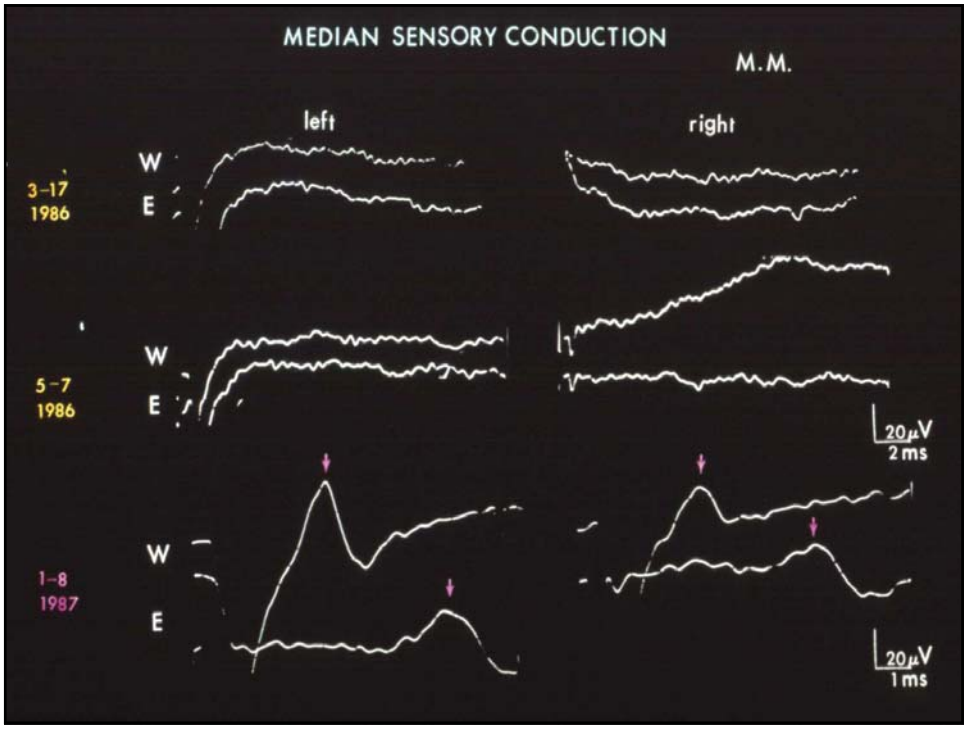

A 2½ YEAR OLD BOY WANDERED OUT OF THE MOBILE HOME INTO A CORN FIELD ON A FRIGID WINTER NIGHT IN IOWA.


WHEN FOUND 2½ HOURS
LATER AT 5:30 AM, HIS
PAJAMAS WERE FROZEN TO
HIS BODY AND ICE HAD BEGUN
TO FORM AROUND HIS FACE.

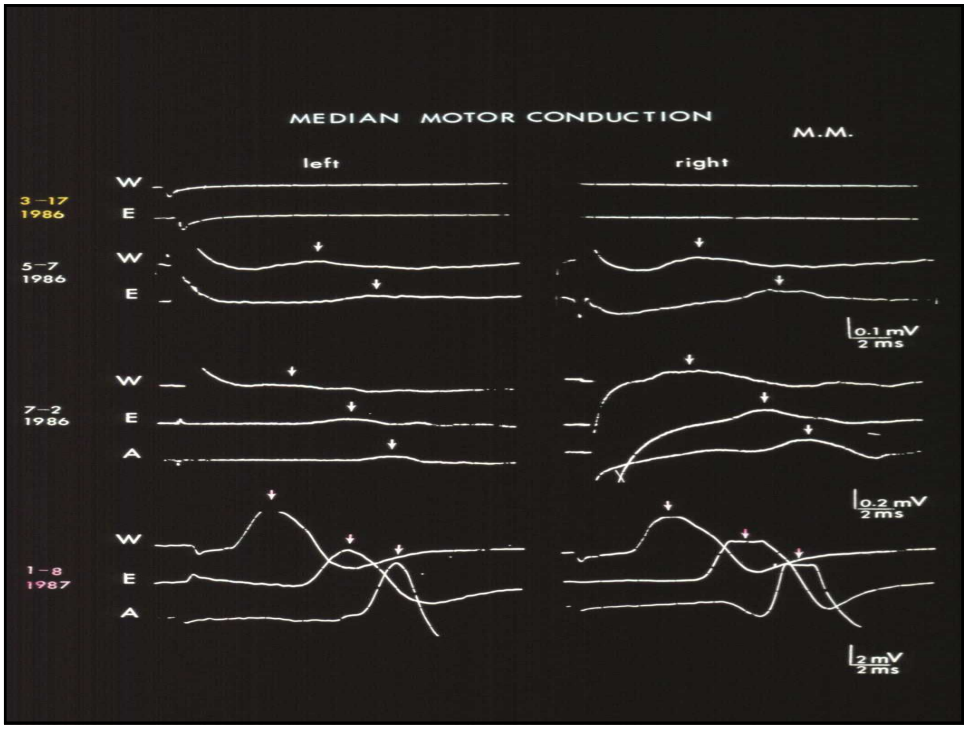

HE HAD NEITHER PULSE
NOR RESPIRATION WITH RECTAL
TEMPERATURE OF 32.2°C.
RESUSCITATION RESULTED IN
RETURN OF SPONTANEOUS
RESPIRATION AND PERIPHERAL
PULSES AT 8:00 AM.

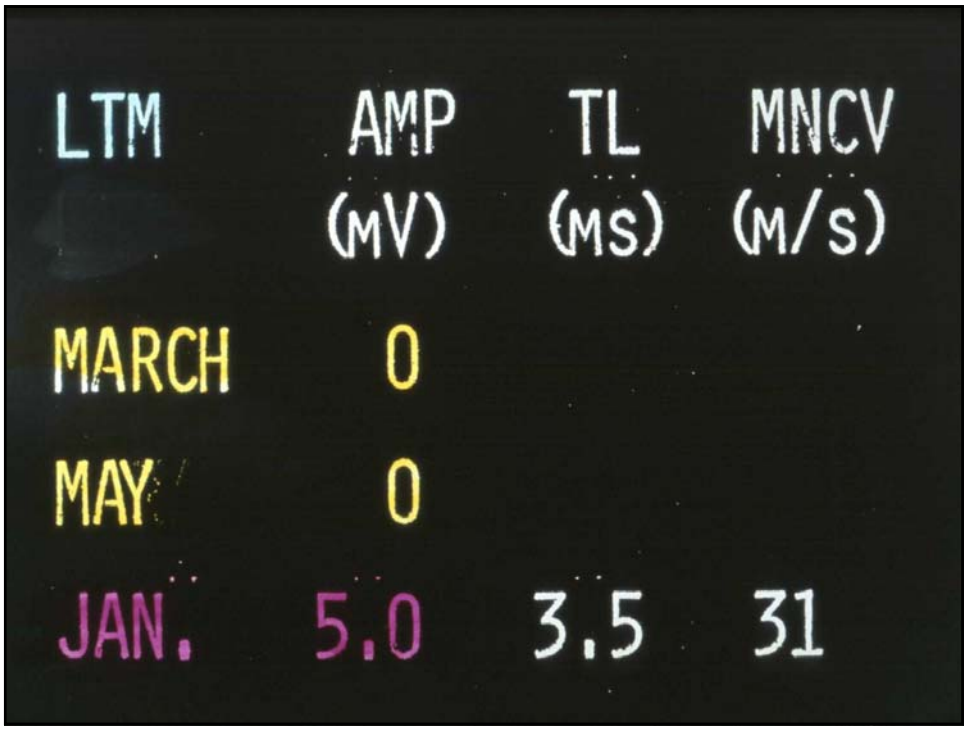
DESPITE GENERALIZED SEIZURES, HIS GENERAL CONDITION IMPROVED OVER THE NEXT FEW DAYS. HOWEVER, HE COULD NOT STAND, RAISE HIS ARM OR HOLD OBJECTS IN HIS HANDS. HE HAD HYPOTONIA AND HYPOREFLEXIA.

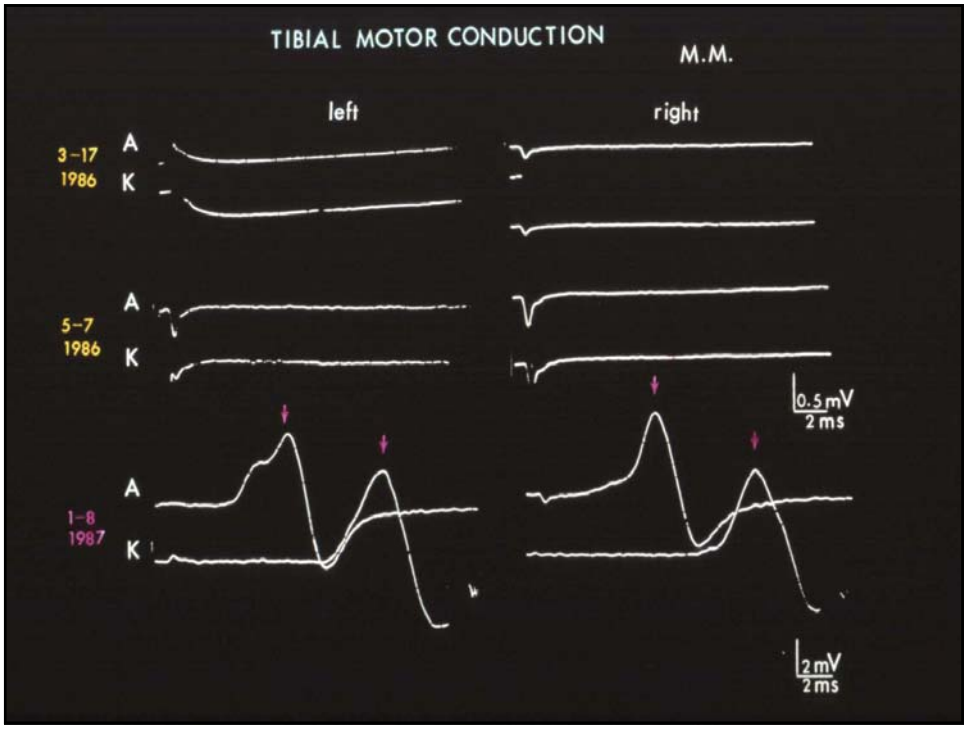
| LMS | AMP (uV) | TL (ms) | SNCV (m/s) |
|---|---|---|---|
| MARCH | 0 | | |
| MAY | 0 | | |
| JULY | 0 | | |
| JAN. | 40 | 2.5 | 40 |

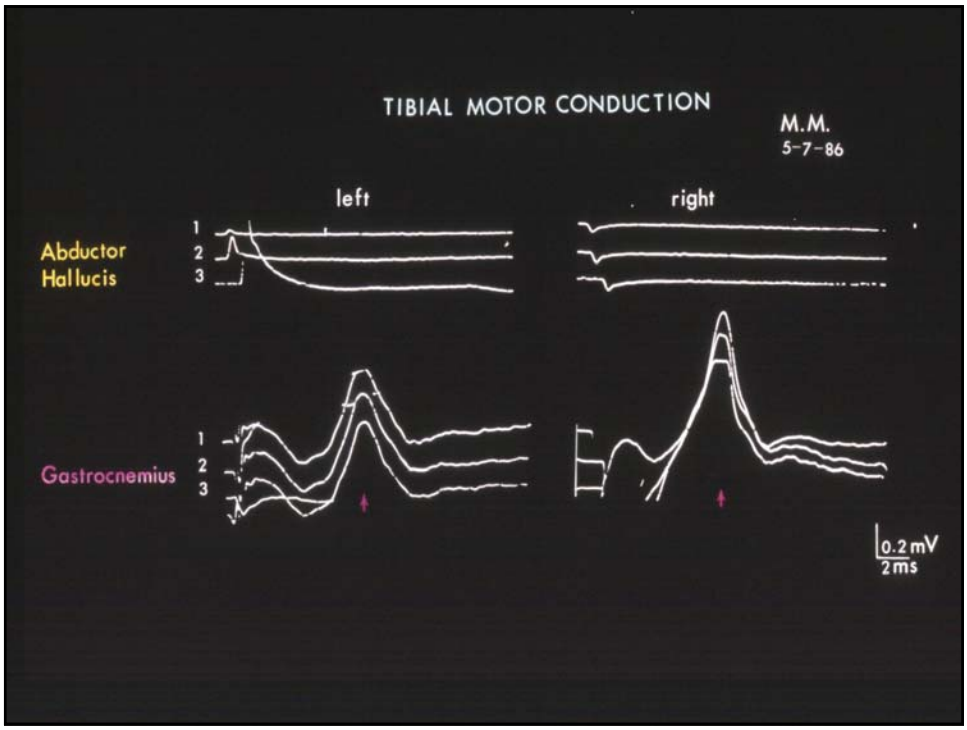



| LMM | AMP (mV) | TL (ms) | MNCV (m/s) |
|---|---|---|---|
| MARCH | 0 | | |
| MAY | 0.1 | 4.6 | 30 |
| JULY | 0.1 | 3.9 | 31 |
| JAN. | 4.0 | 3.3 | 34 |


MEDIAN MOTOR CONDUCTION
M.M.
left
right
3–17
1986
5–7
1986
7–2
1986
1–8
1987
W
E
A
0.1 mV
2 ms
0.2 mV
2 ms
2 mV
2 ms


LTM
AMP
(mV)
TL
(ms)
MNCV
(m/s)
MARCH
0
MAY
0
JAN.
5.0
3.5
31


TIBIAL MOTOR CONDUCTION
M.M.
left
right
3-17
1986
A
K
5-7
1986
A
K
0.5 mV
2 ms
1-8
1987
A
K
2 mV
2 ms


TIBIAL MOTOR CONDUCTION
M.M.
5-7-86
left
right
Abductor
Hallucis
1
2
3
Gastrocnemius
1
2
3
0.2 mV
2 ms