SOMENE
SOCIEDAD MEXIQUENSE DE NEUROLOGÍA A.C

Sistema Nervioso Periférico

Neuropatías, Miopatías y Enfermedades de la Unión Neuromuscular

Dr. Perfecto Oscar González Vargas

Neurólogo

INNN – HMP MP – SOMENE - UAEM

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CEREBRUM DIVINA LUX RATIO SALUS SCIENTIAE · MEXICO ·

AMEVASC
Asociación Mexicana de Enfermedad Vascular Cerebral, A.C.

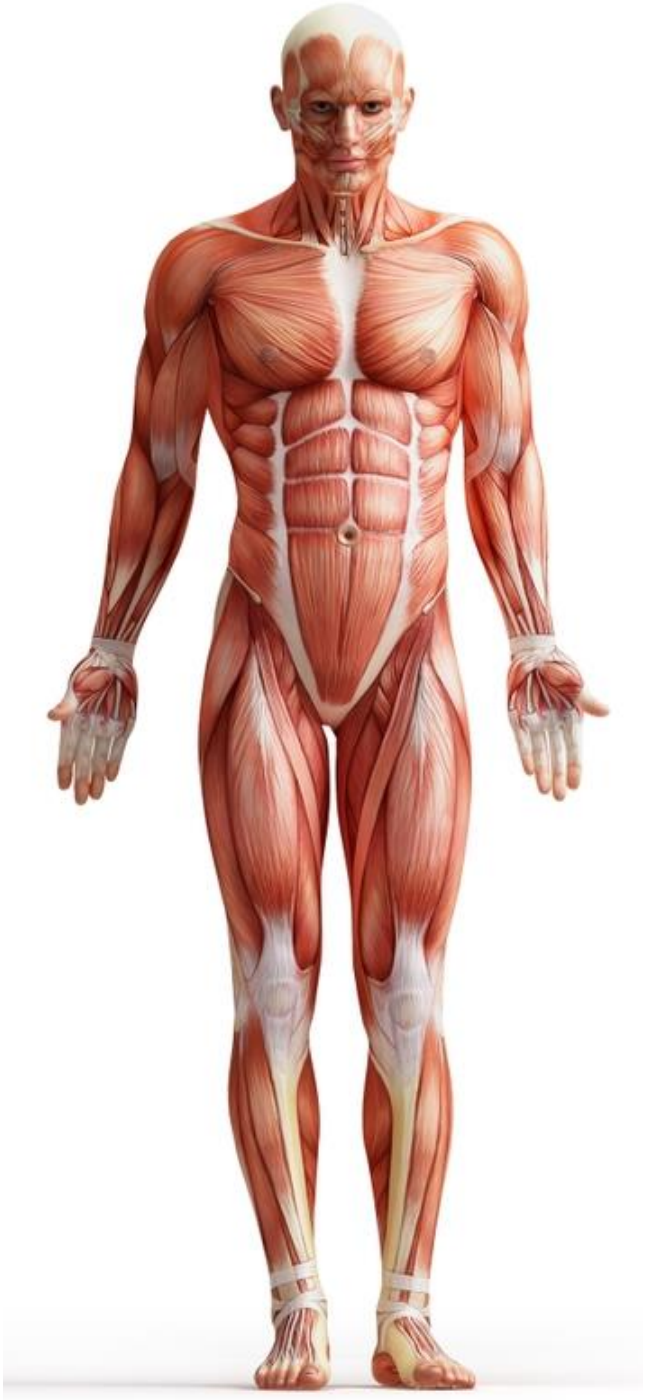
International Parkinson and Movement Disorder Society

CAMELICE

Academia Mexicana de Neurología, A.C.

Miopatías

- Son patologías del funcionamiento o de la estructura del músculo.
- Clínicamente:
    - Debilidad proximal
    - Simétrica



Cintura Escapular

Cintura Pélvica

Miopatias.

Datos Clínicos

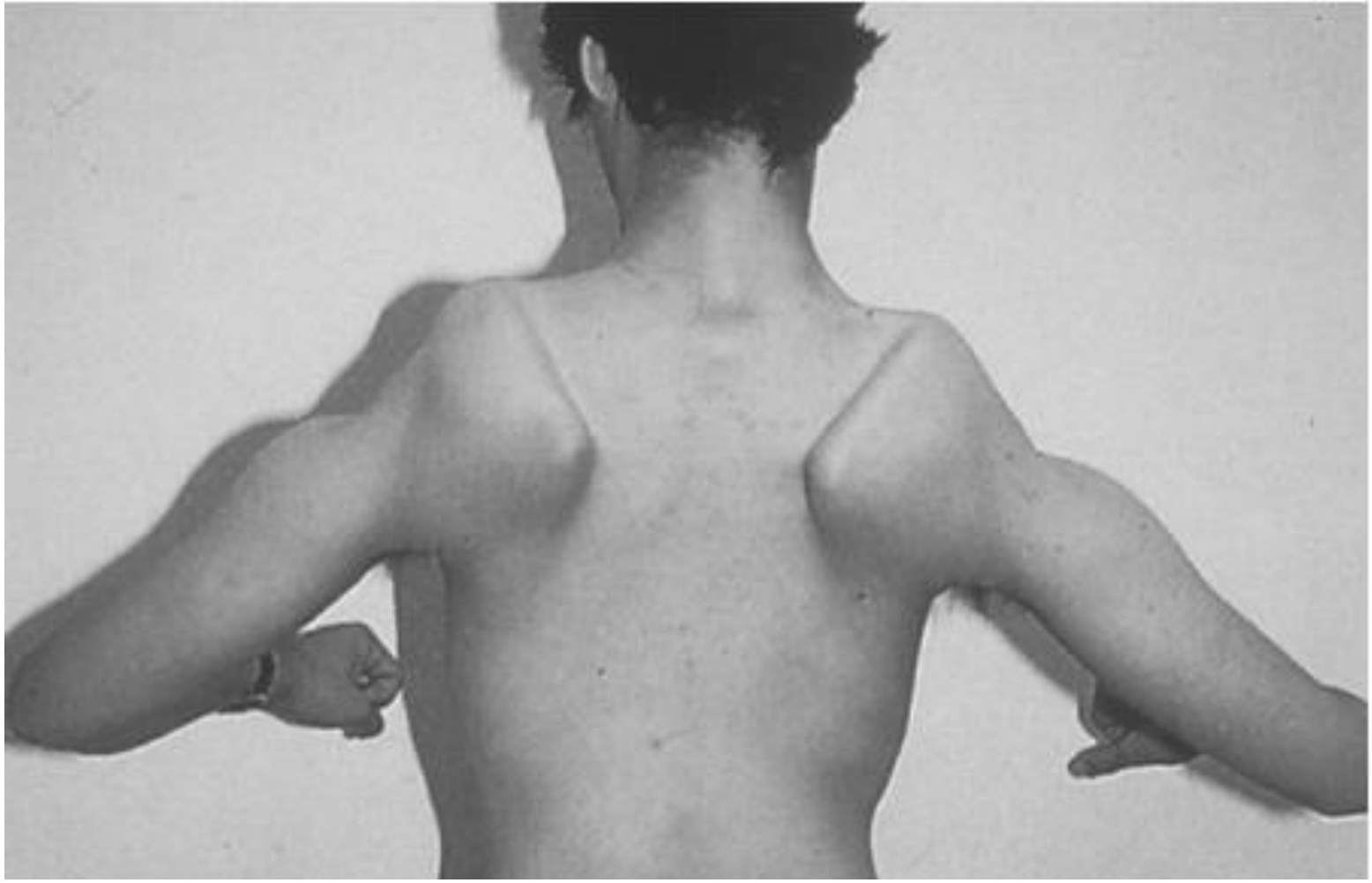
- Debilidad y fatigabilidad
- Mialgias
- Espamos
- Rigidez muscular. *Stiff man*
- Miotonía
- Masa muscular (disminuída o incrementada). Pseudohipertrofia.

Fenómeno Miotónico

Postura Lordótica

Source: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson JL, Loscalzo J: *Harrison's Principles of Internal Medicine, 18th Edition*: www.accessmedicine.com

Lordotic posture, exaggerated by standing on toes, associated with trunk and hip weakness.



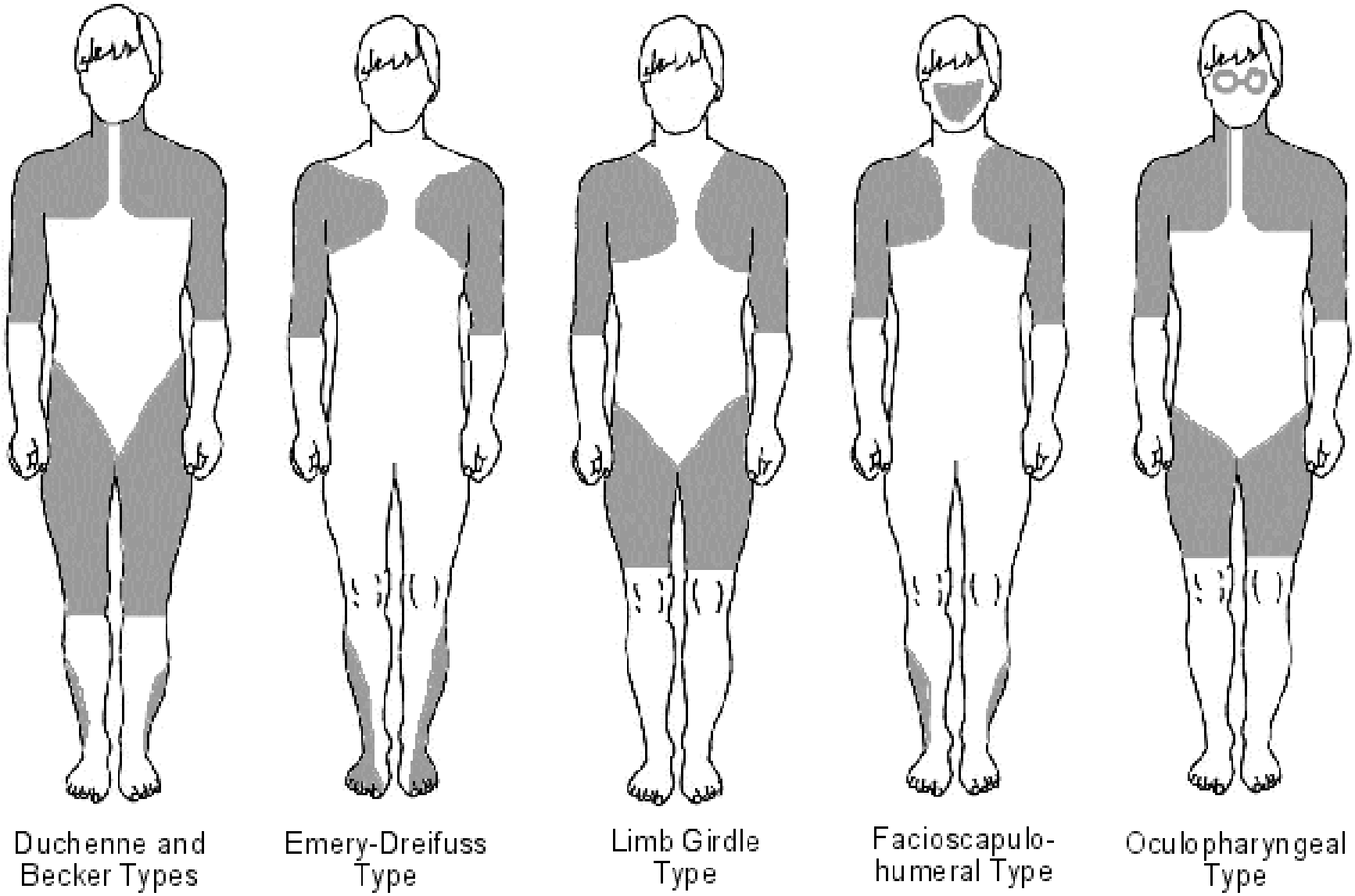
Escapula Alar

Facioscapulohumeral dystrophy with prominent scapular winging.

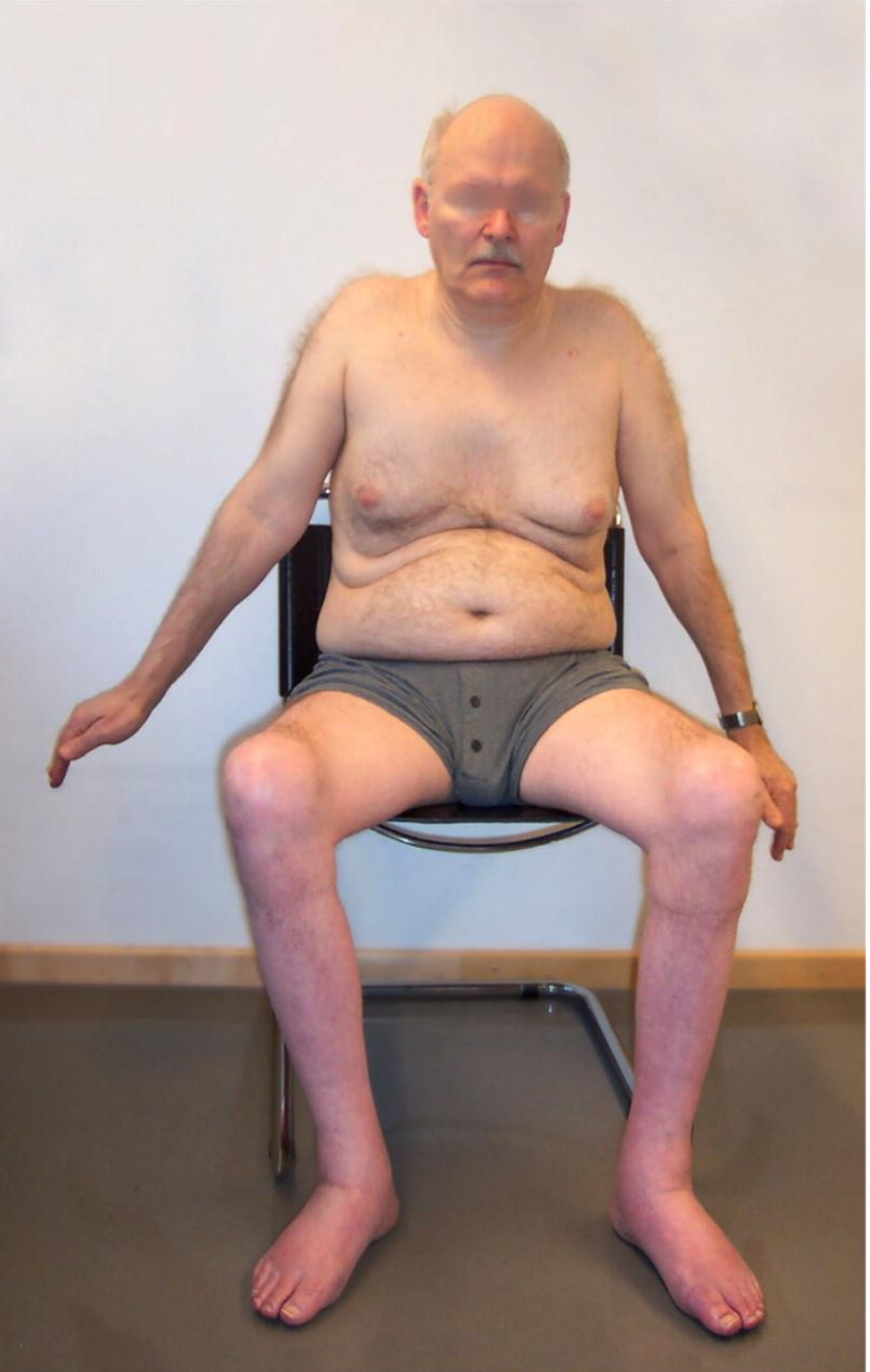
¿signo de..?

Miopatias. Debilidad Muscular

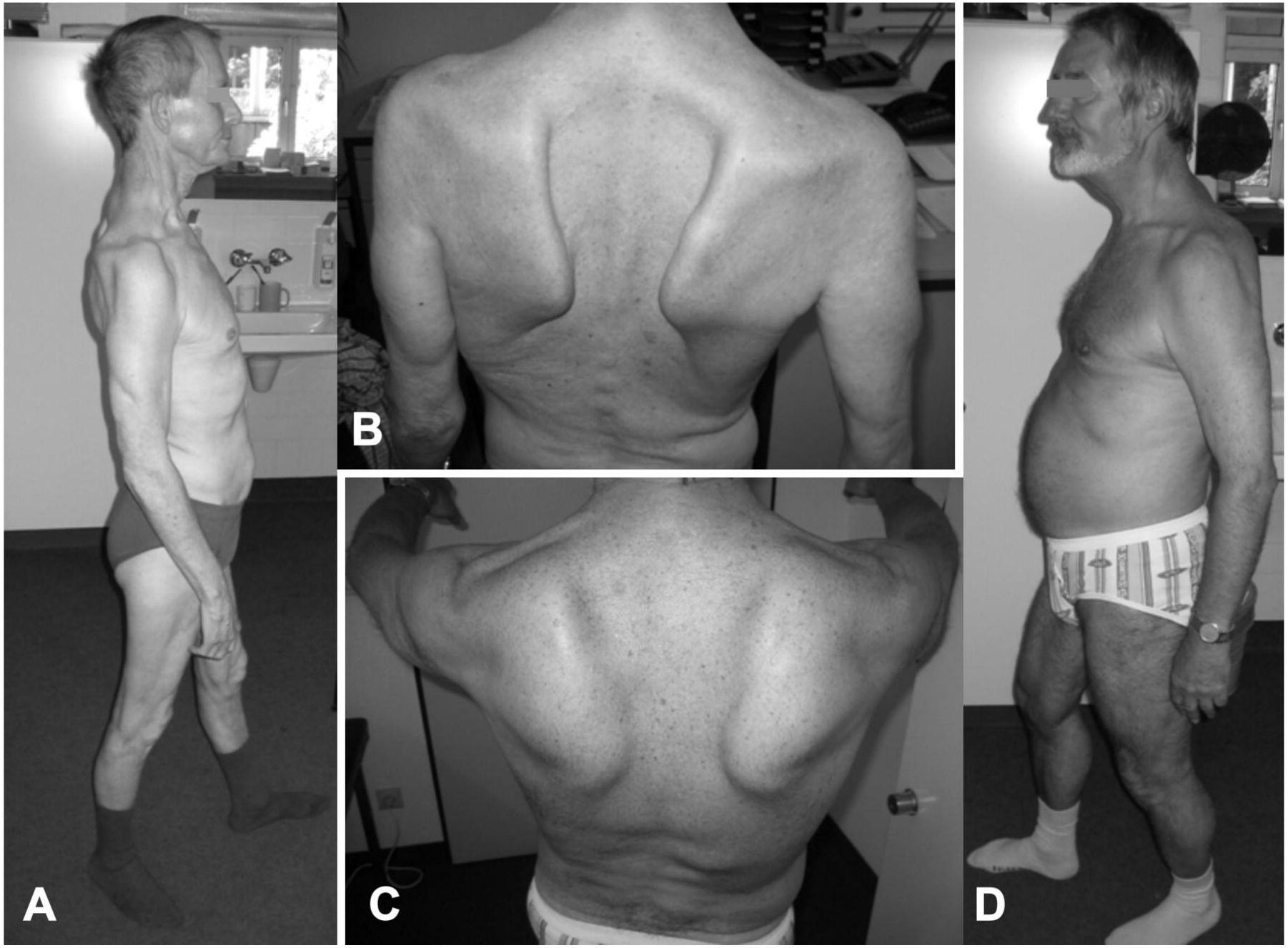
- Intermitentes:
  - Parálisis Hipokalémica, Miastenia Gravis , Mioglobinuria
- Persistes:
  - Miopatías Inflamatorias. Polimiositis
  - Distrofias Musculares. Degenerativas



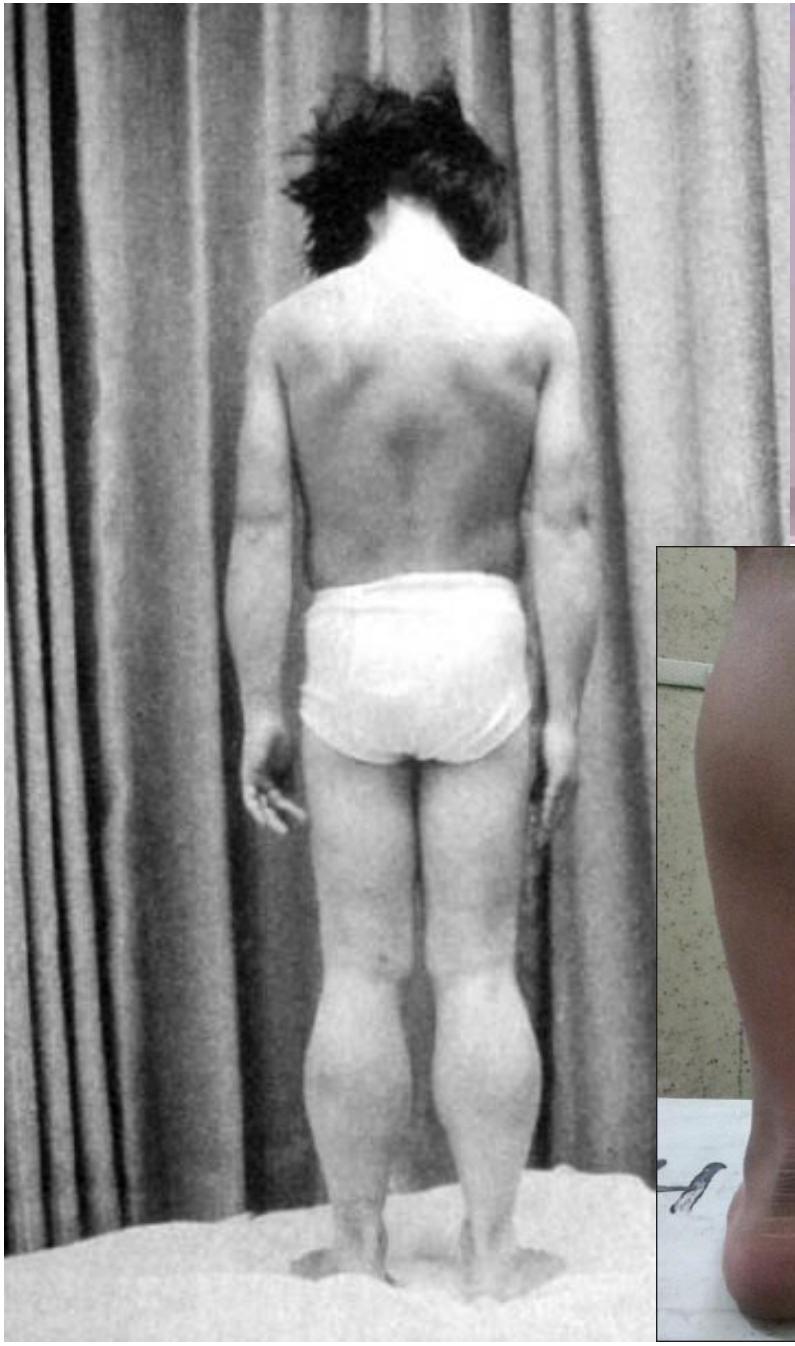
Main areas of muscle weakness in different types of dystrophy

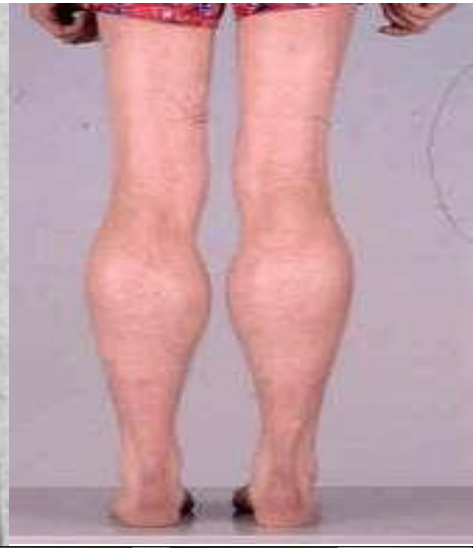


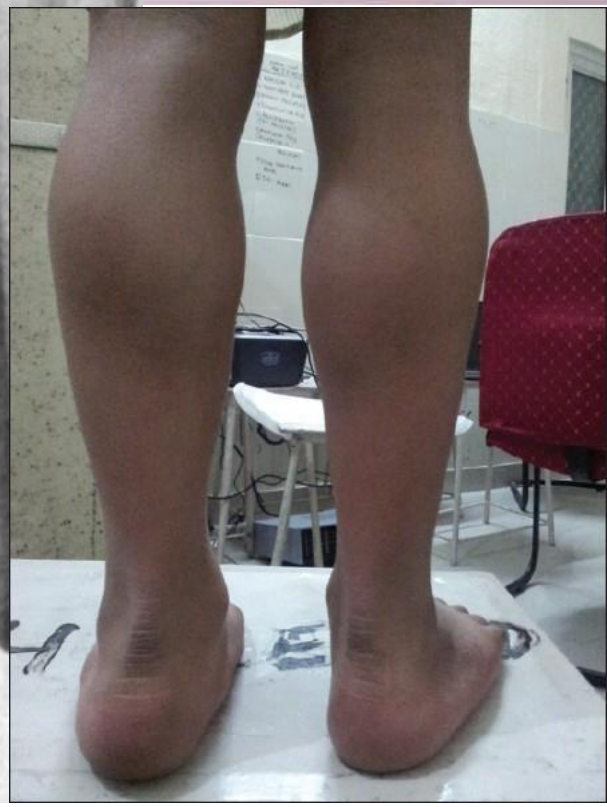
Síndrome Escapulohumeral


A
B
C
D

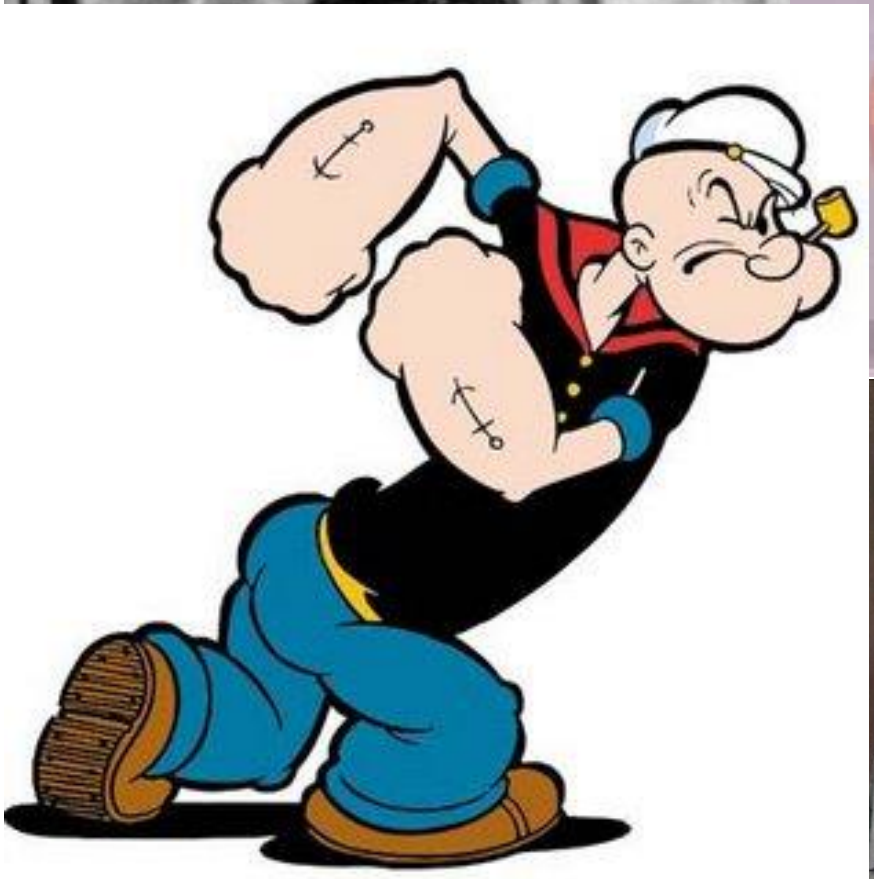
Pseudo-hipertrofia.

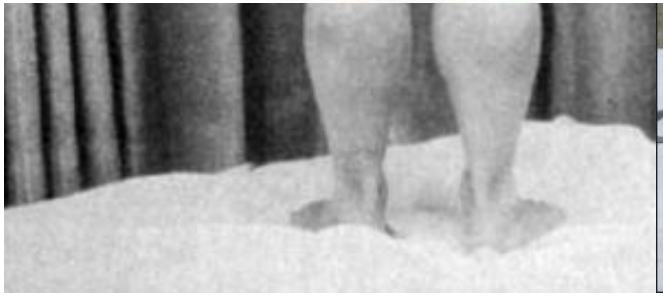






Pseudo-hipertrofia.





Miopatias.
Diagnóstico

- Laboratorio. CPK

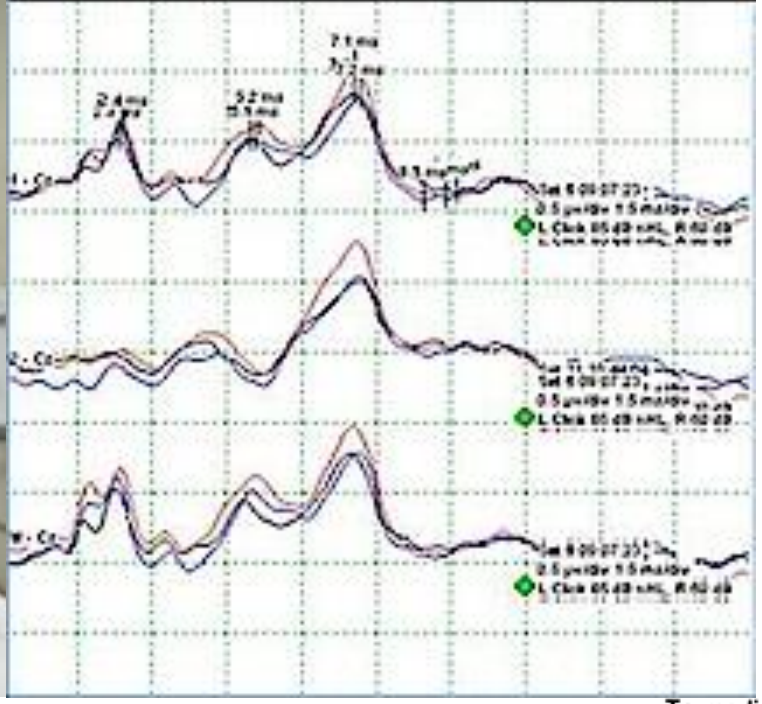
- Electrodiagnóstico:

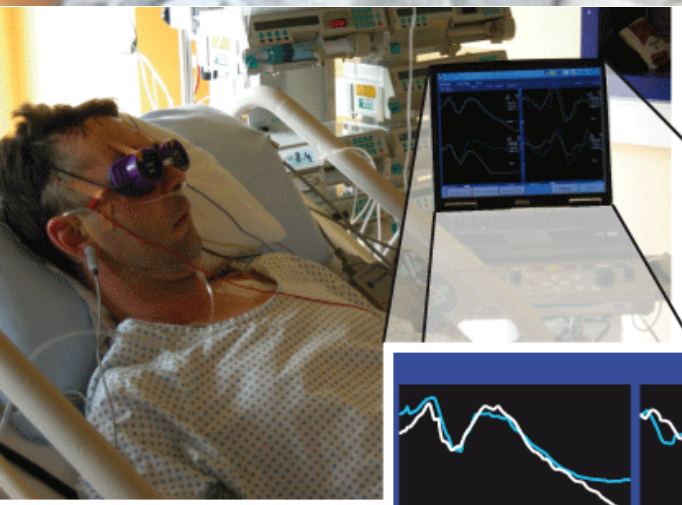
  Electromiografía. **EMG**
- Diagnóstico Molecular
- Biopsia

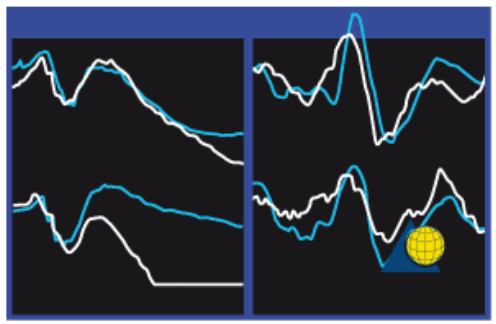
Miopatias.

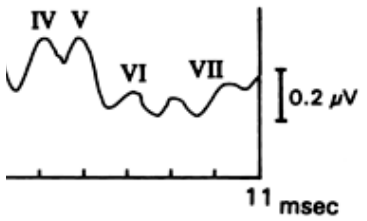
Neurofisiología

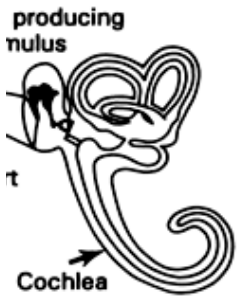
- Electromiografía. EMG.
- Velocidades de Conducción Nerviosa
- Potenciales Evocados:
  - Visuales. PEV
  - Auditivos. PEAT
  - Somatosensoriales. PESS

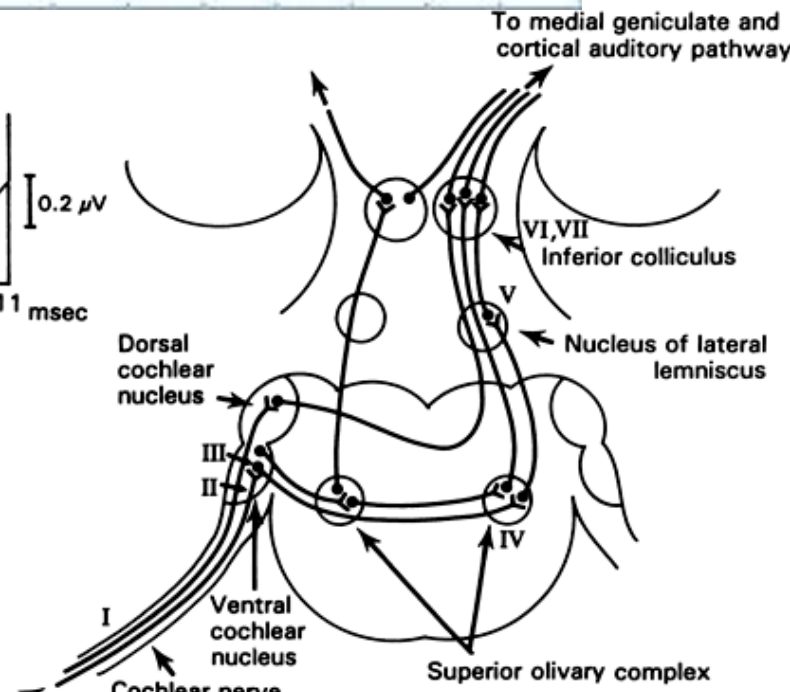






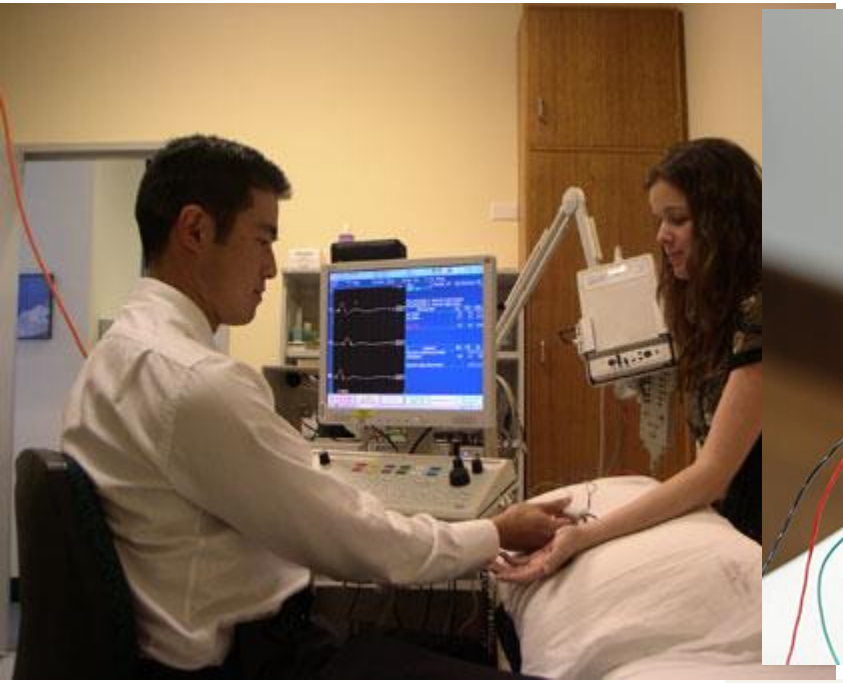

IV V
VI
VII
0.2 μV
11 msec

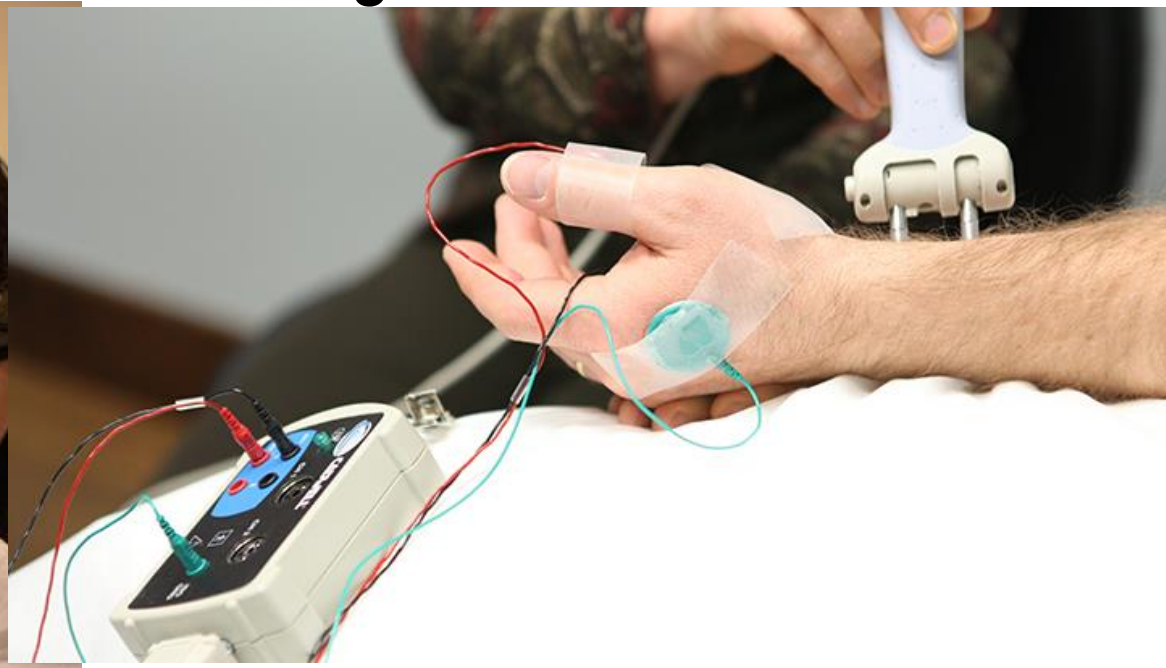

producing
nulus
t
Cochlea

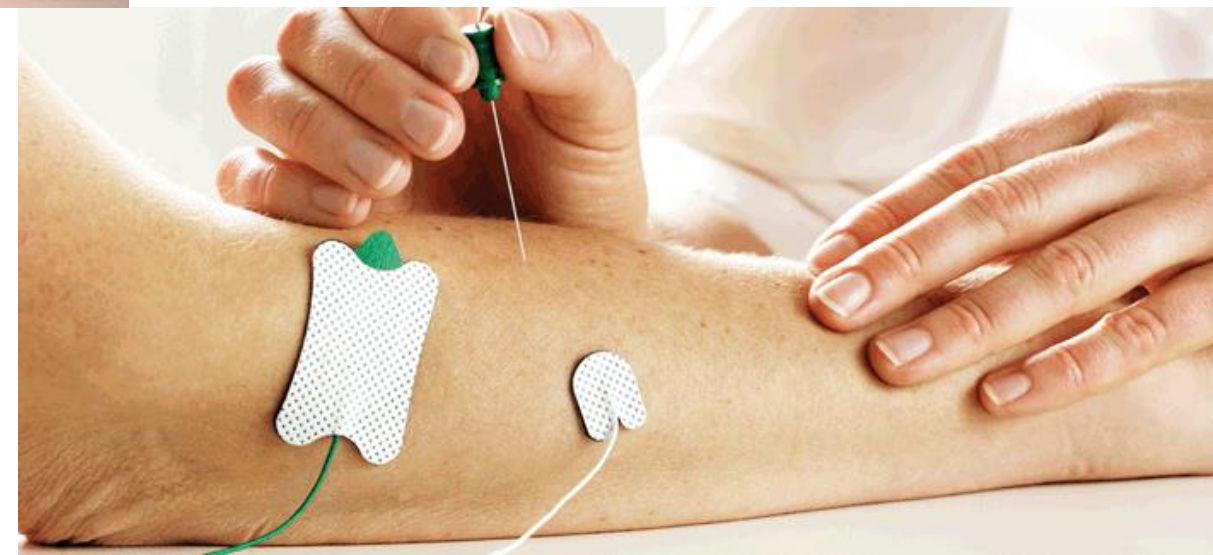

To medial geniculate and
cortical auditory pathway
VI,VII
Inferior colliculus
V
Nucleus of lateral
lemniscus
Dorsal
cochlear
nucleus
III
II
IV
I
Ventral
cochlear
nucleus
Cochlear nerve
Superior olivary complex

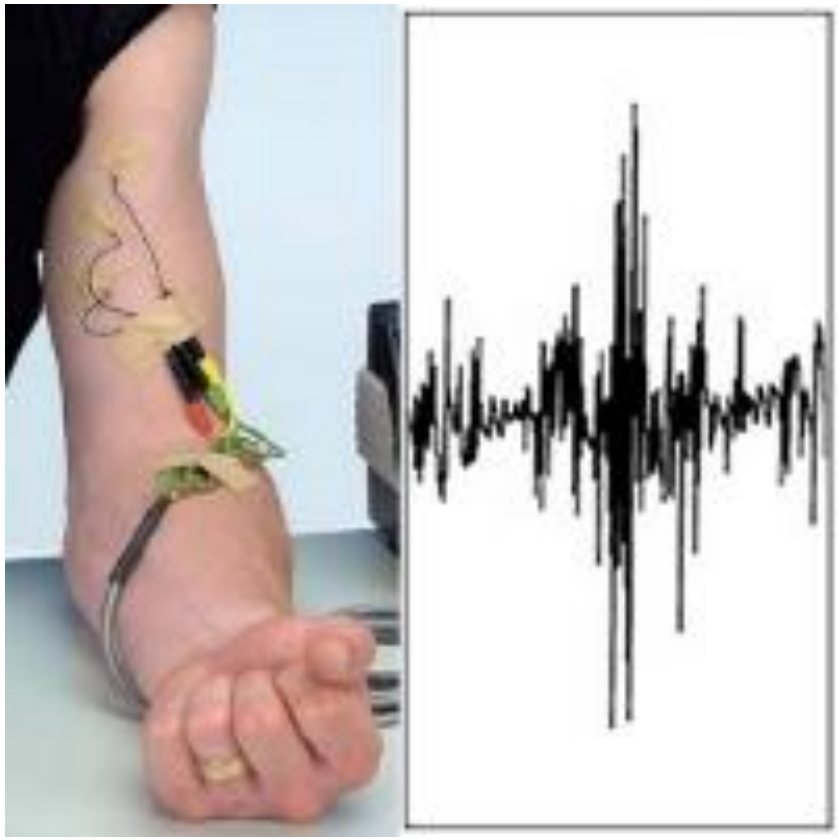
Miopatias.

Neurofisiología









Miopatias.
Tratamiento

- Certeza diagnóstica.
- Nada efectivo para las Distrofias Musculares.

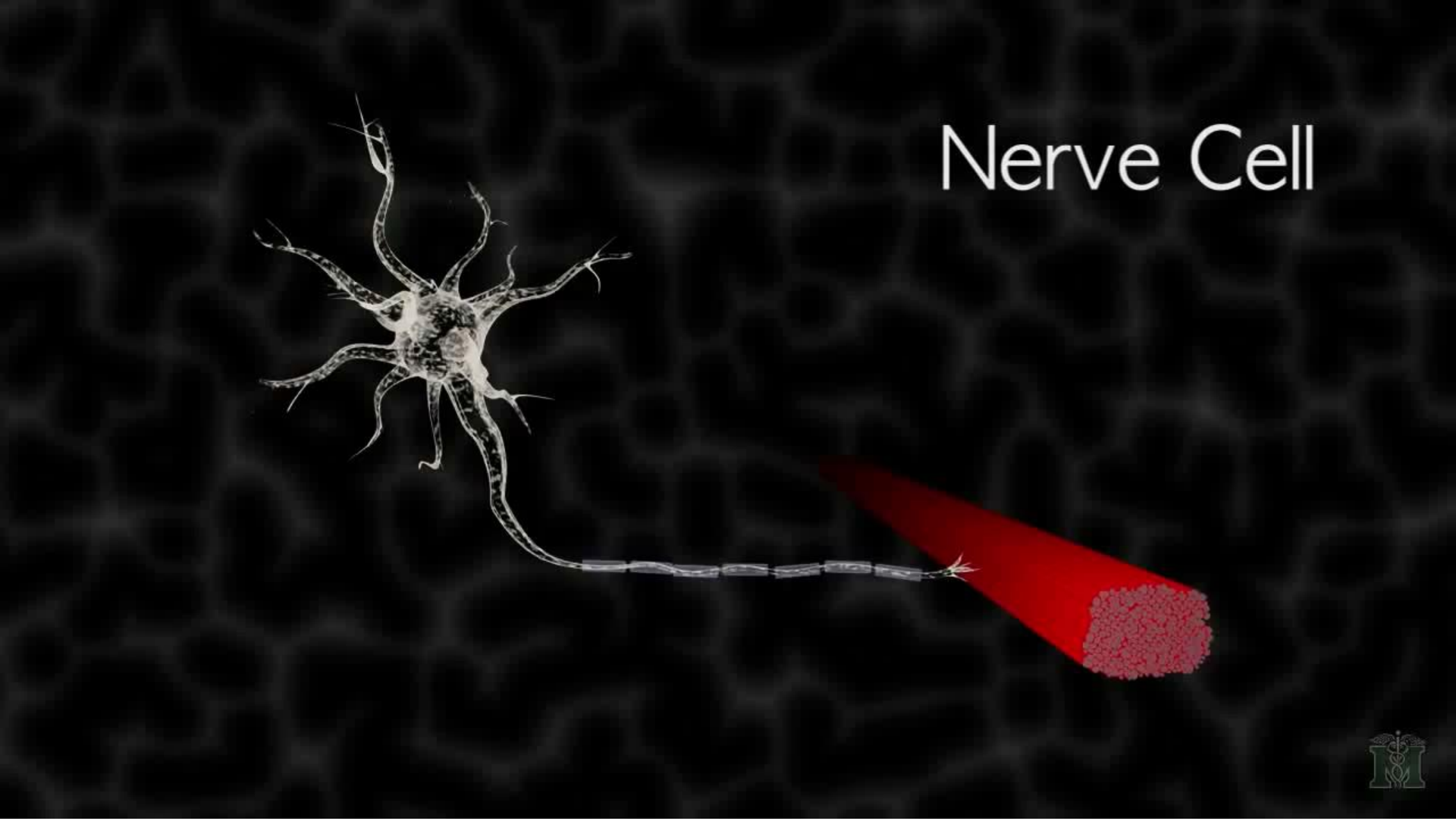
Ante un paciente con Miopatía no Inflamatoria buscar el tratamiento en 3er nivel.

Miastenia Gravis

Miastenia Gravis
Características Clínicas Principales

- Enfermedad de la Unión Muscular
- Debilidad y Fatigabilidad Muscular FLUCTUANTE
- Enfermedad Autoinmune
- Timoma
- Anticuerpos contra el Receptor de Acetilcolina
- Dx: Prueba del Edrofonio (Tensilón) y Anticuerpos Séricos Circulantes

Nerve Cell



Clasificación de Osserman

Tabla I. Clasificación de Osserman.

Estadio	0	Sin datos clínicos
Estadio	I	Debilidad muscular de músculos del ojo
Estadio	II	Debilidad que afecta otro grupo muscular que no es el ocular
	IIA	Afecta extremidades, músculos axiales o ambos
	IIB	Afecta músculos orofaríngeos o respiratorios
Estadio	III	Debilidad moderada afectando otro grupo muscular que no sea el ocular
	IIIA	Afecta extremidades, músculos axiales o ambos
	IIIB	Afecta músculos orofaríngeos o respiratorios
Estadio	IV	Debilidad severa afectando otro grupo muscular que no sea el ocular
	IVA	Afecta extremidades, músculos axiales o ambos
	IVB	Afecta músculos orofaríngeos o respiratorios
Estadio	V	Paciente intubado con apoyo o no de ventilación mecánica, que no incluye al paciente en el manejo postoperatorio en un paciente con MG

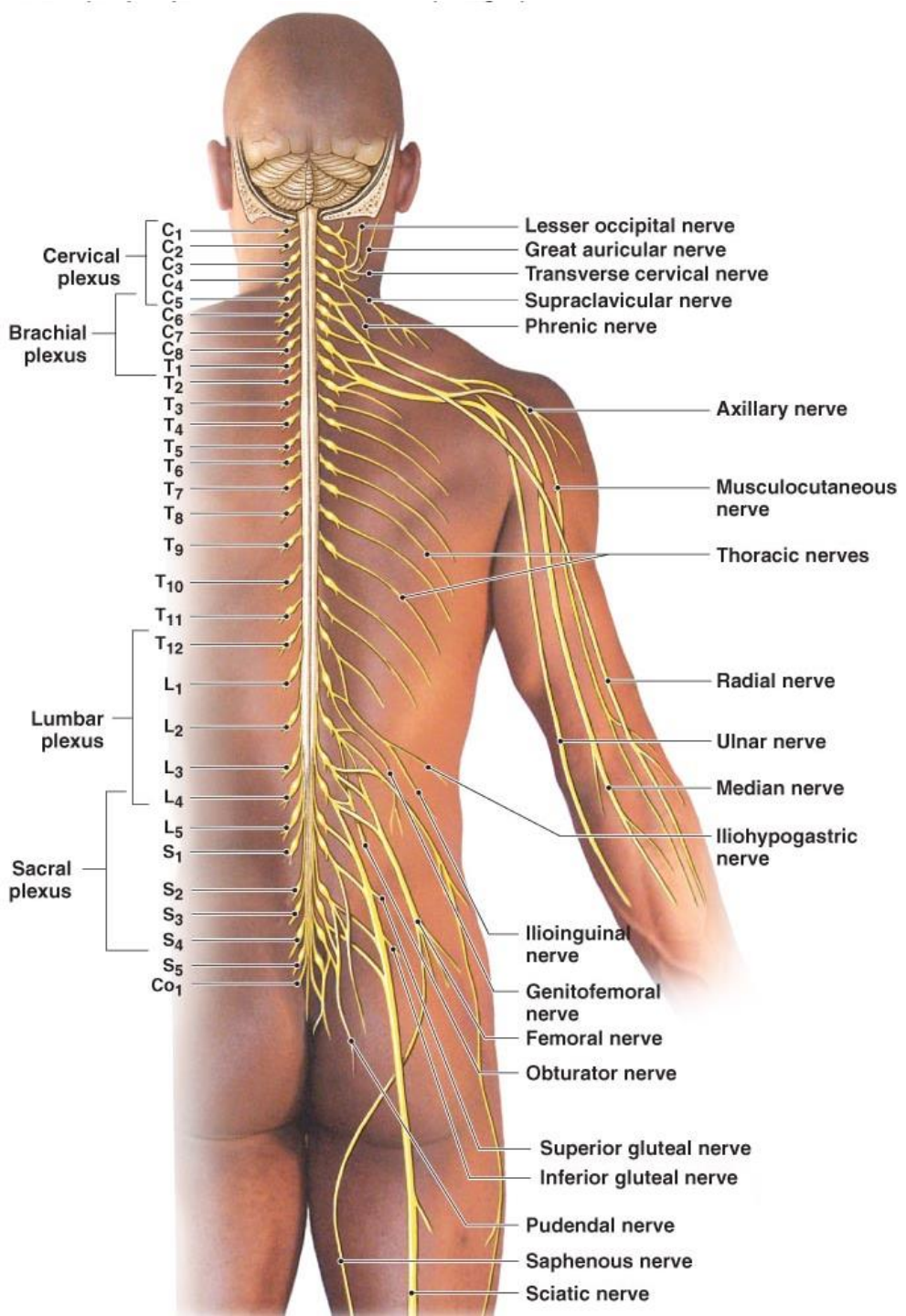
Tomado de: Osserman KE, Genkins G. Studies on myasthenia gravis; a reference for Health Care Professionals. In: Myasthenia Gravis Foundation of America. HYPERLINK http://www.myasthenia.org November, 2003.

Miastenia Gravis

Tratamiento

- Colinérgicos.
- Esteroides
- Inmunosupresores. Azatioprina, Ciclosporina, Micofenolato
- Plasmaféresis. Gammaglobulina
- Timectomía
- Hay fármacos que empeoran la MG.

Polineuropatías

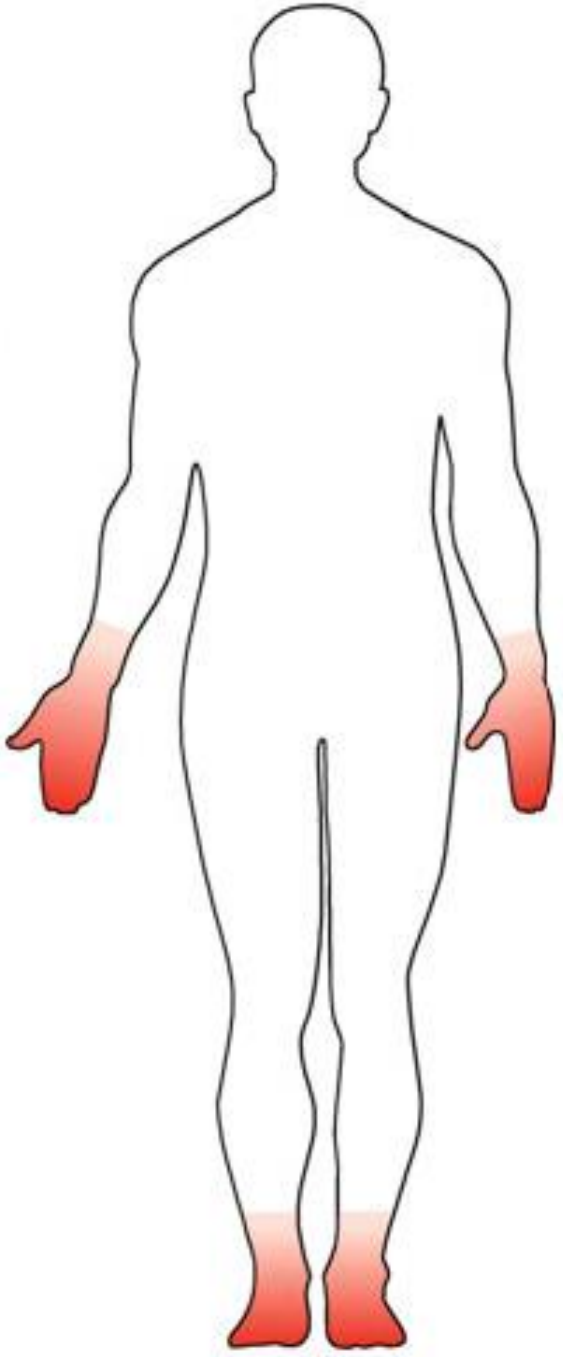


Nervio Periférico

A. Sensitivo

B. Motor

C. Autonómico



Nervio Periférico
Sintomatogía

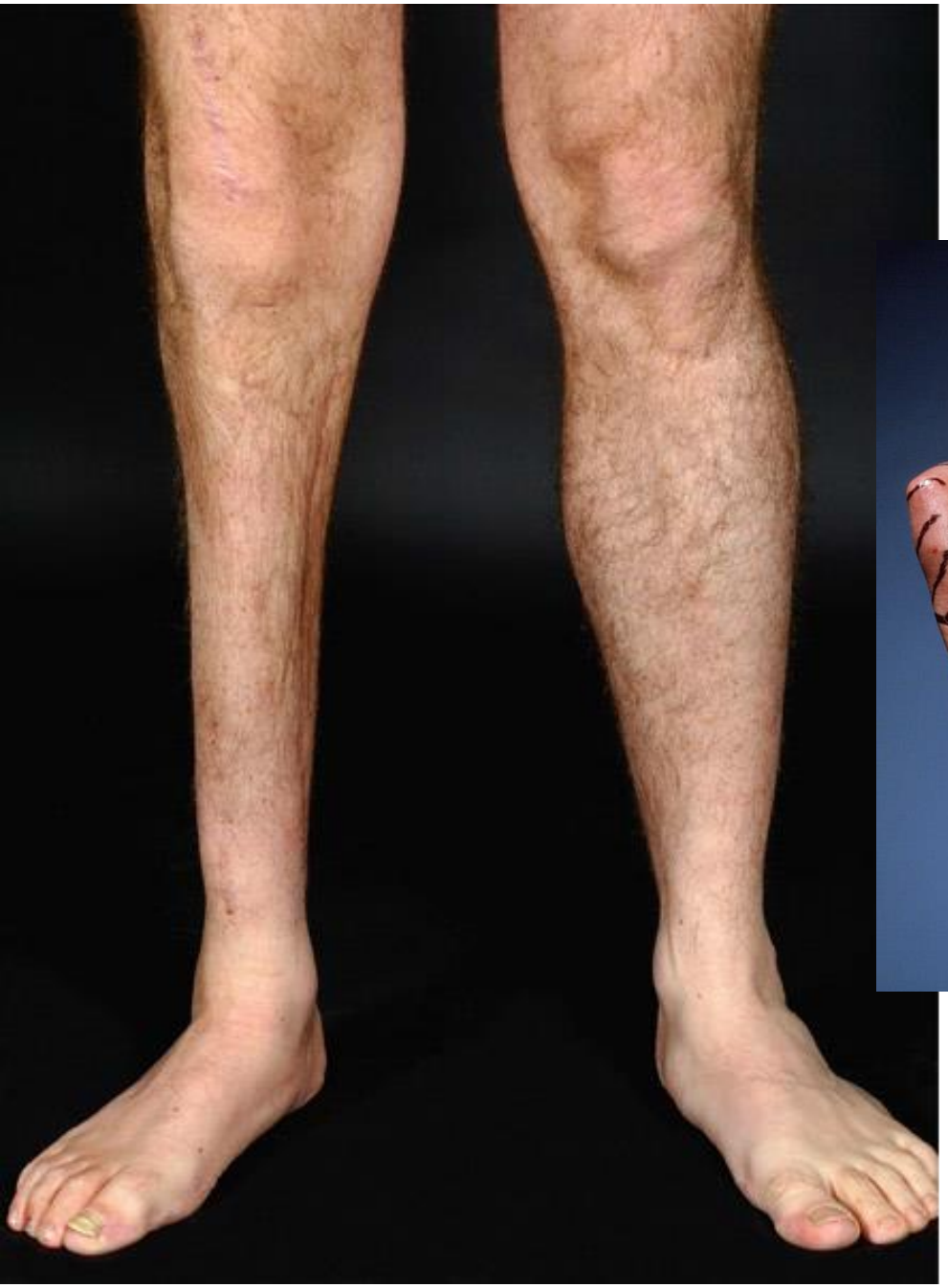
A. **Sensitivo**. Disestesias, parestesis. Habitualmente en “guante y calcetín”

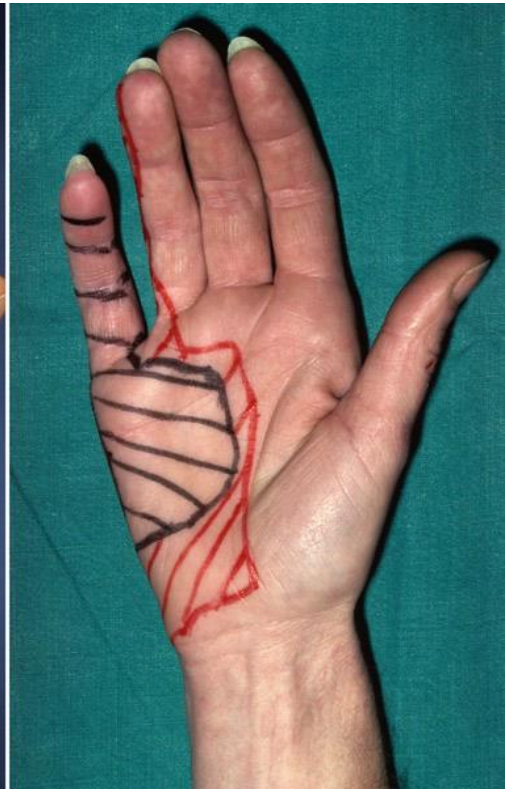
B. **Motor.** Debilidad motora DISTAL

C. **Autonómico.** Problemas en control del sudor, temperatura, hipotensión, erección etc

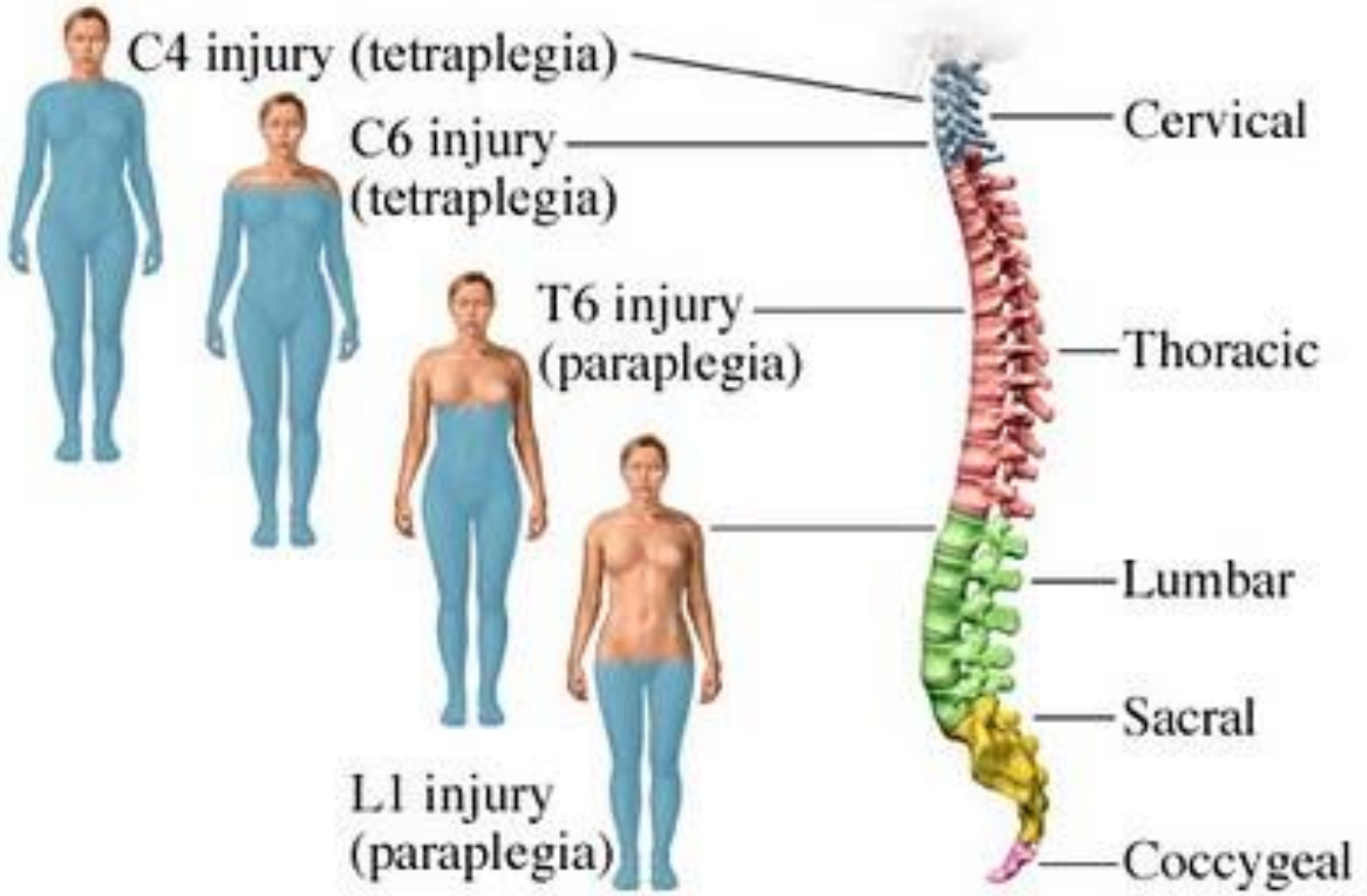
Nervio Periférico = **Síndrome de Neurona Motora Inferior**

1. **Debilidad**
2. **Arreflexia o Hiporeflexia**
3. **Atrofia**
4. **Fasciculaciones**





¿Clínica de un Nervio Periférico?



¿Clínica de un Nervio Periférico?

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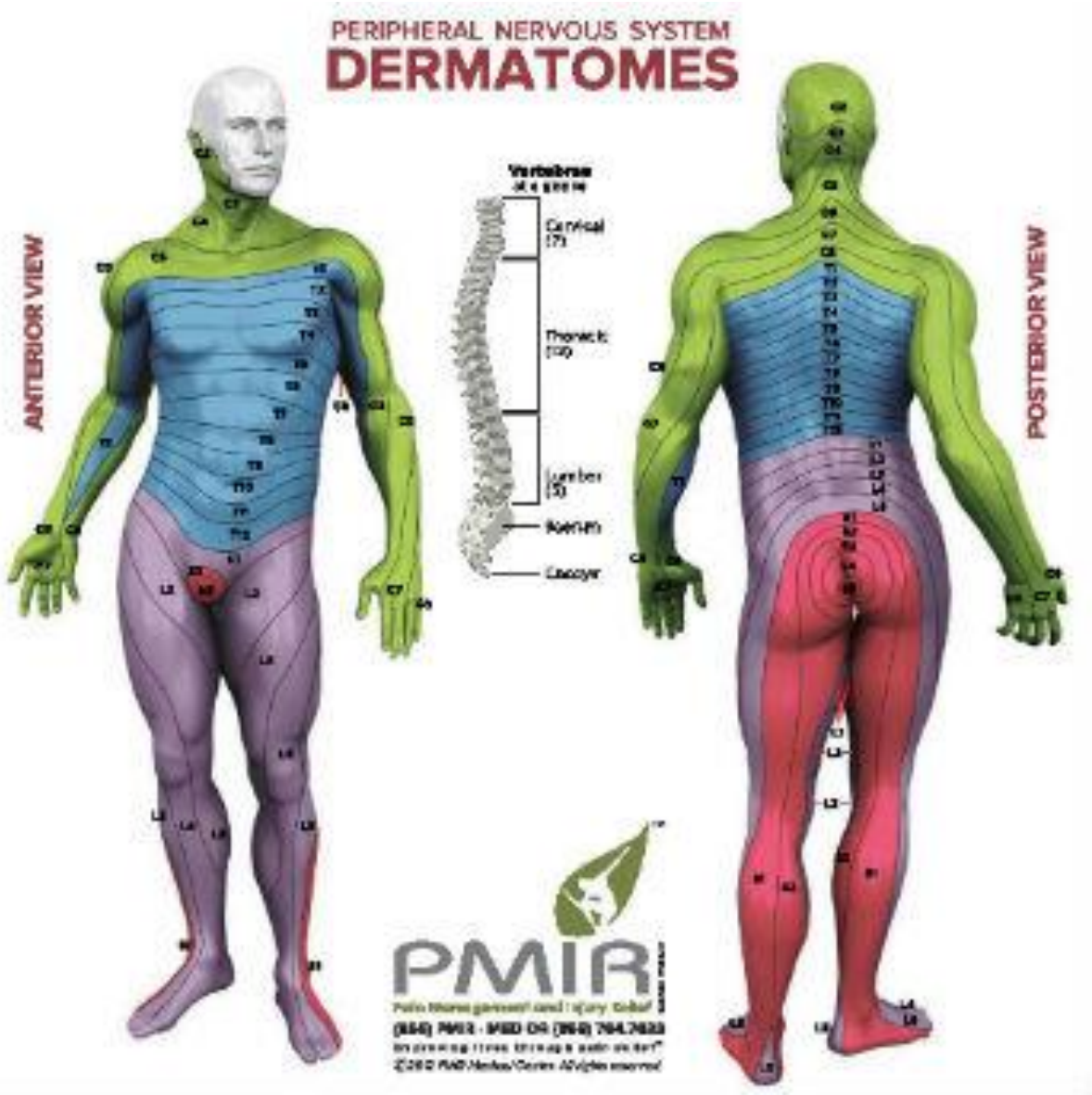
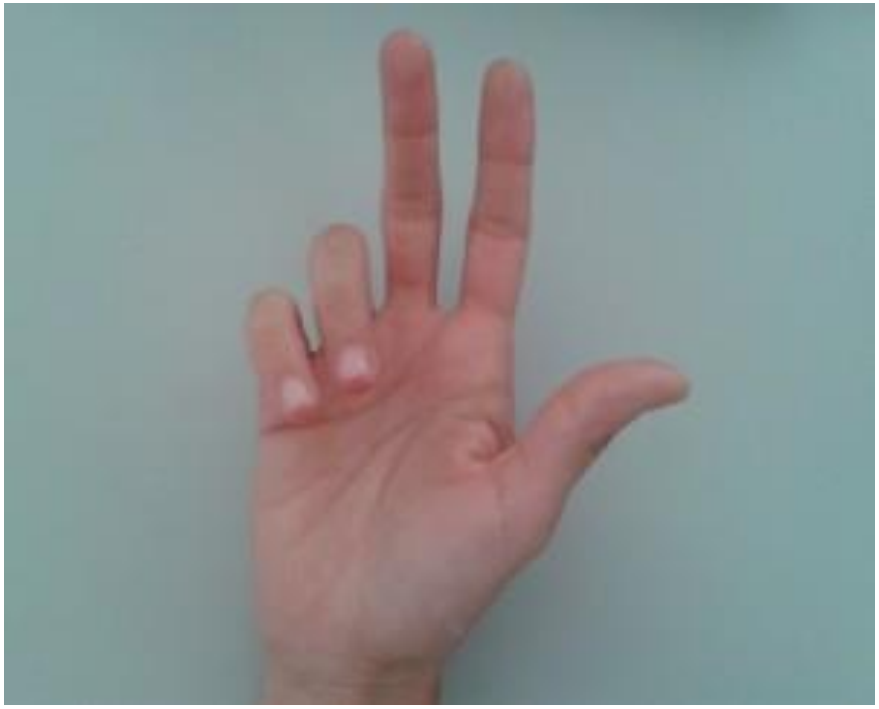
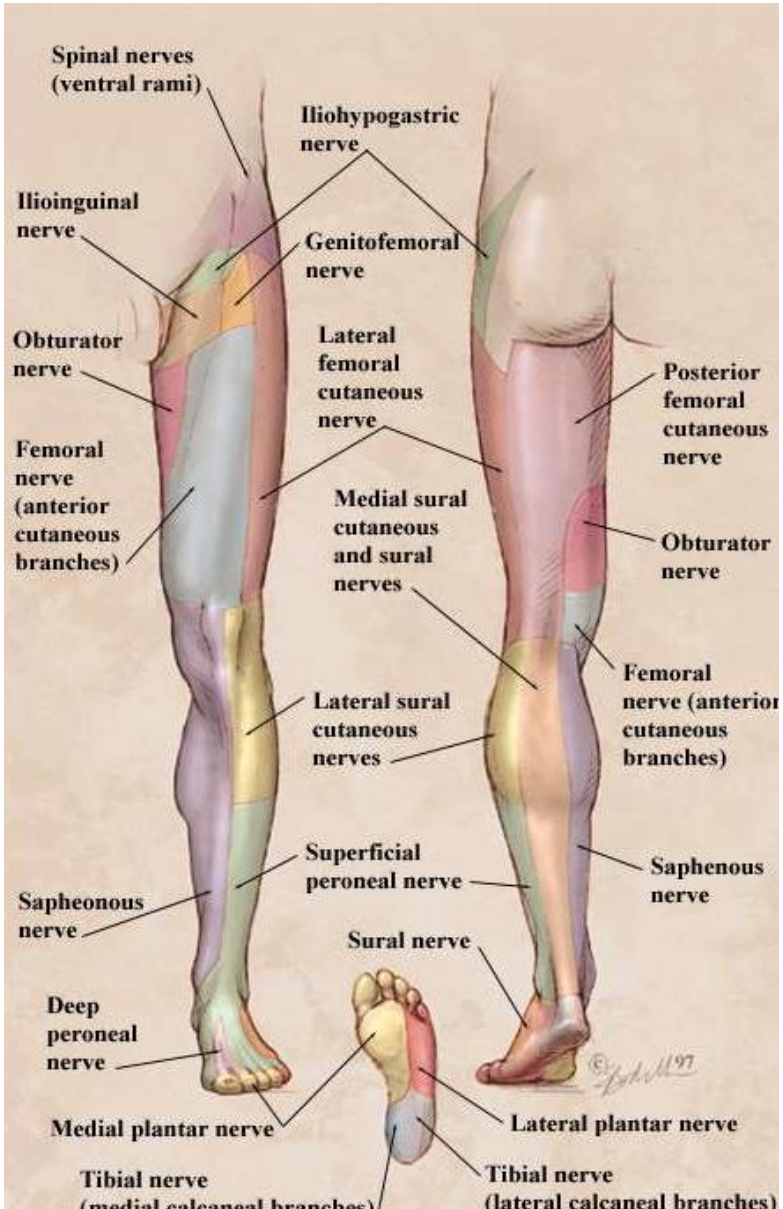


¿Clínica de un Nervio Periférico?





PNP

Clasificación

A. Por Evolución Temporal. Aguda, subaguda, crónica.
B. Por Etiología
C. Por Distribución. Distal, simétrica.
D. Por Patología. Desmielinizantes o axonales.

Clasificación por **Evolución Temporal**

A. **Agudas**. Días-4 semanas

B. **Subagudas**. 4-8 semanas

C. **Crónicas**. > 8 semanas

Evolución Temporal. Patologías comunes

A. **Agudas**. Días-4 semanas. Sx Guillain-Barré, tóxicas

B. **Subagudas**. 4-8 semanas PNP Diabética

C. **Crónicas**. > 8 semanas Genéticas

Evolución Temporal. Patologías comunes

A. **Agudas**. Días-4 semanas. Sx Guillain-Barré, tóxicas

B. **Subagudas**. 4-8 semanas PNP Diabética

C. **Crónicas**. > 8 semanas Genéticas

D. **Recaídas**.

Polineuropatía Desmielinizante Inflamatoria Crónica **CIDP**, Porfiria.

Clasificación por el

Número de Nervios Afectados

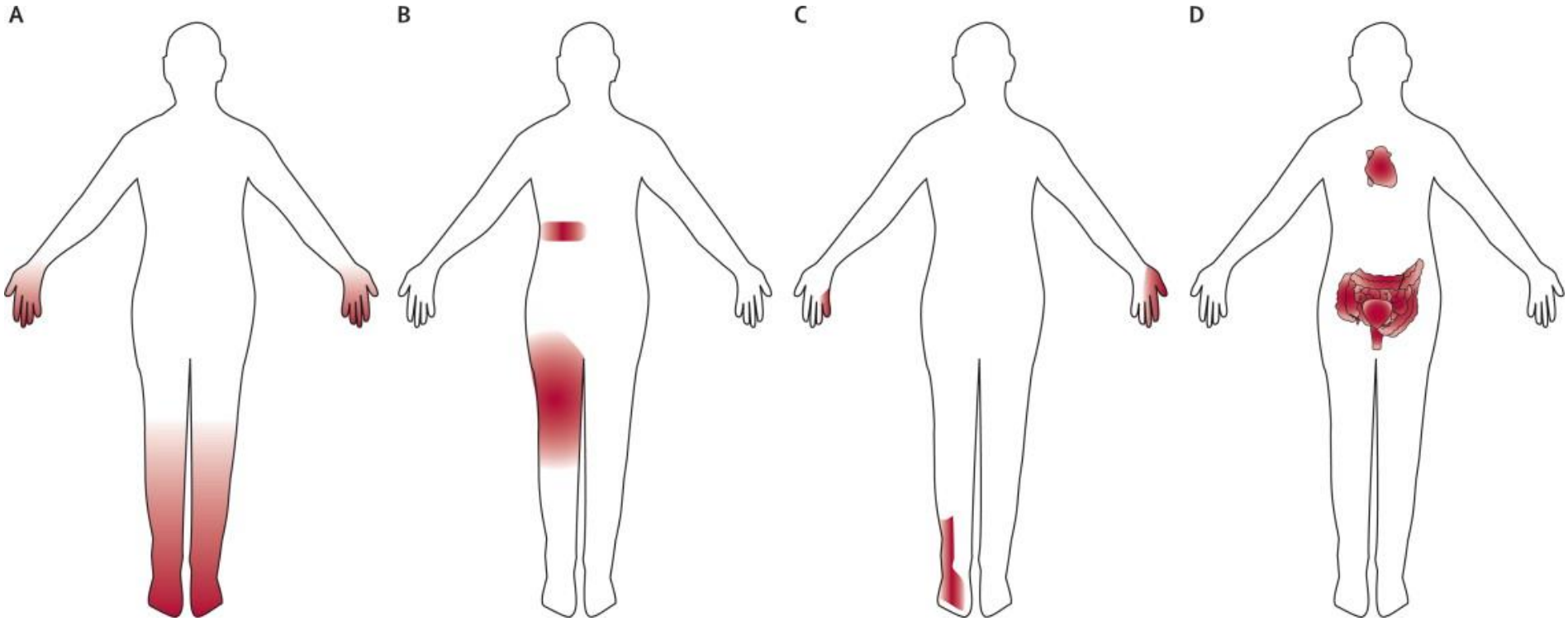
Mononeuropatía	**Polineuropatía**	**Mononeuritis Múltiplex**
Atrapamiento	Metabólicas	Vasculitis
Sx Tunel Carpo	Diabética	AR
Ciática	Sx GB	

Considerar si es:

Proximal-Distal,

Simétrica-Asimétrica,

Motora-Sensitiva-Autonómica.



Patrones Comunes y Patologías “Características”

A. **Distal, Simétrica, <u>Motora</u>**. Sx de Guillain-Barré

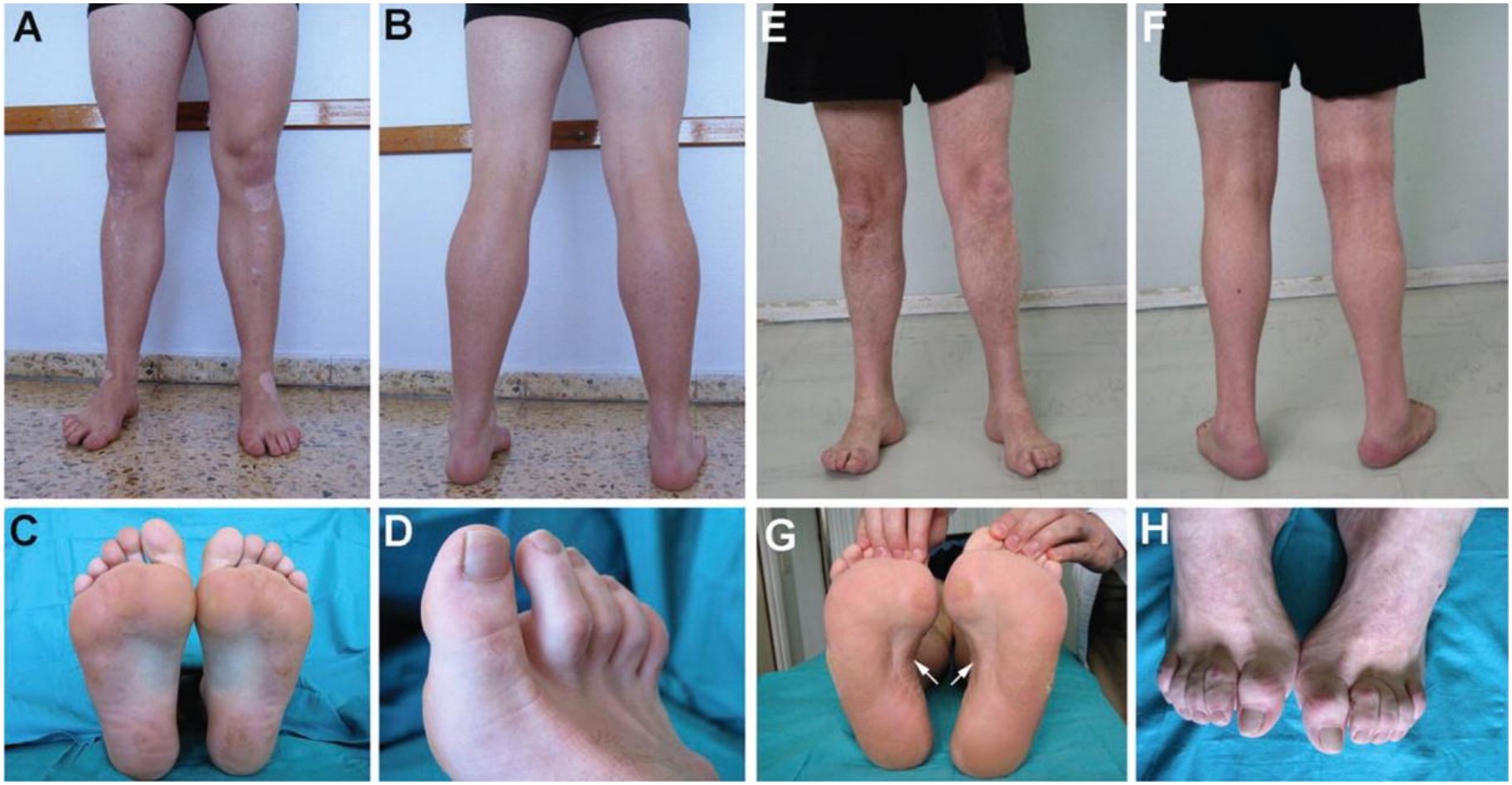
B. **Distal, Simétrica, <u>Sensitiva</u>**. Metabólica, Tóxica

C. **Distal, Simétrica, <u>Sensitiva-Motora</u>.** Metabólica, Fármacos

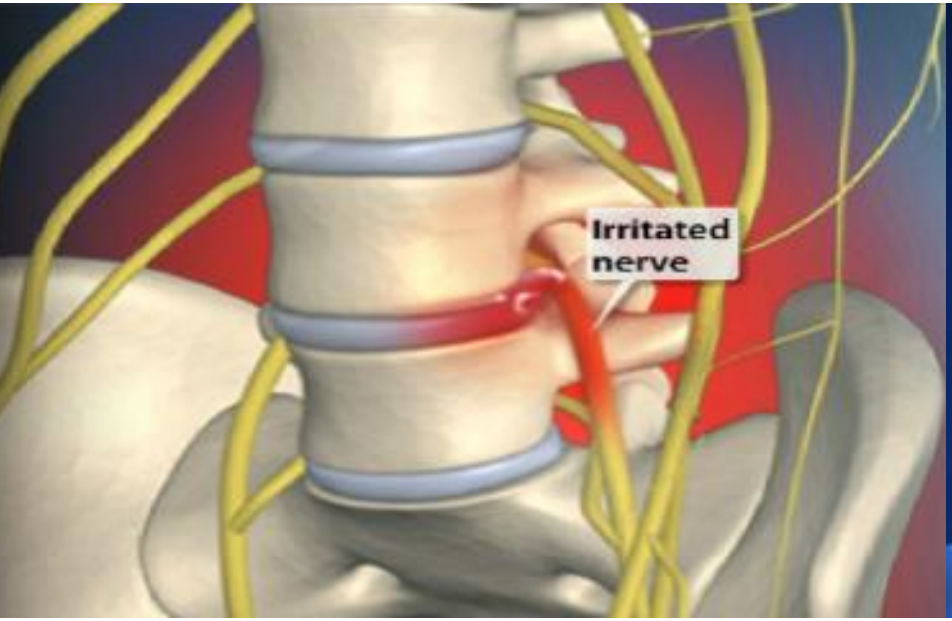
D. **PNP con Síntomas Autonómicos.** DM, Hereditaria, Porfiria

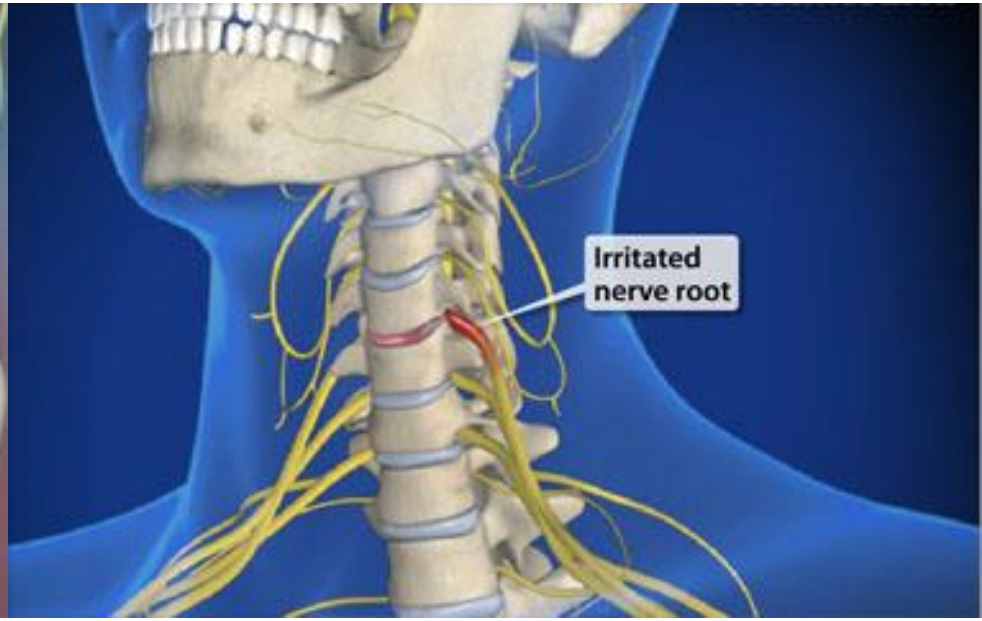
¿Hereditaria?

Charcot-Marie-Tooth

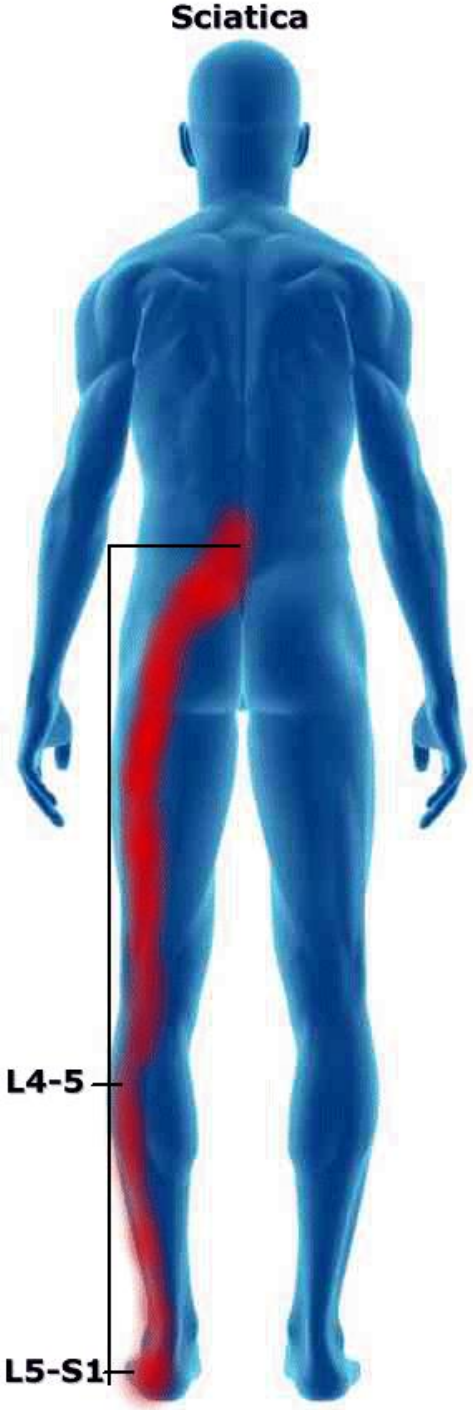


Radiculopatías



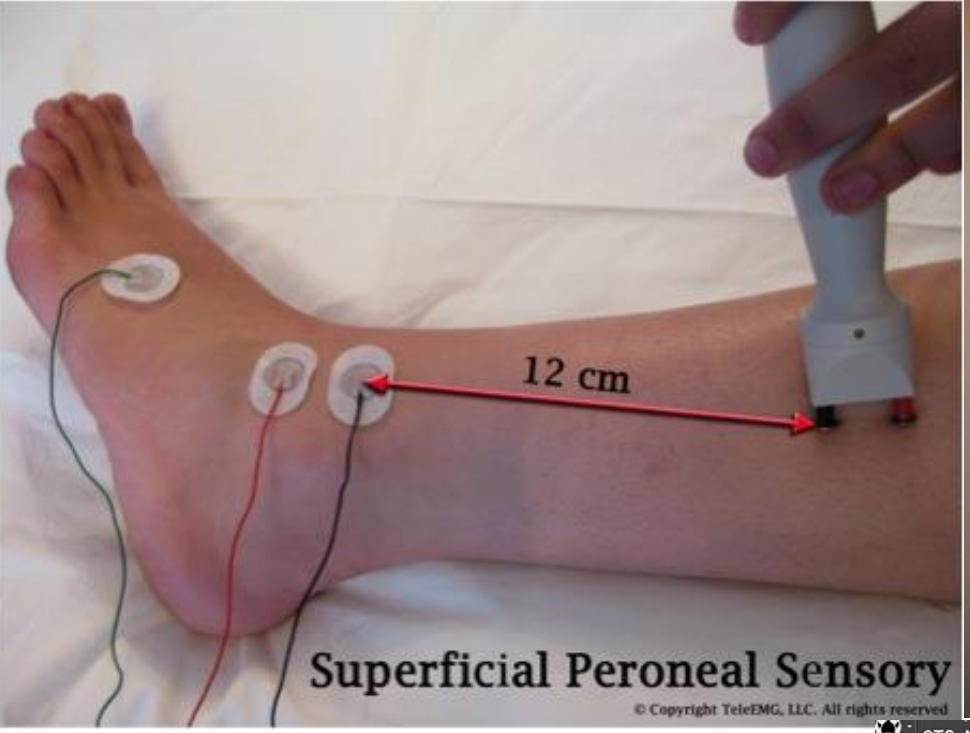


Sciatica



Diagnóstico de las PNP

A. Clínico
B. Electrodiagnóstico. Velocidad de conduc nerviosa
C. LCR. Guillain-Barré
D. Patológico. Biopsia de nervio periférico



CTS: Median and Ulnar... | Right Ulnar | Record | 08:34:32

Switch: STOP | Rate: 2 Hz | Level: 0.0 mA | Dur: 0.1 ms | Single
Stim: 1

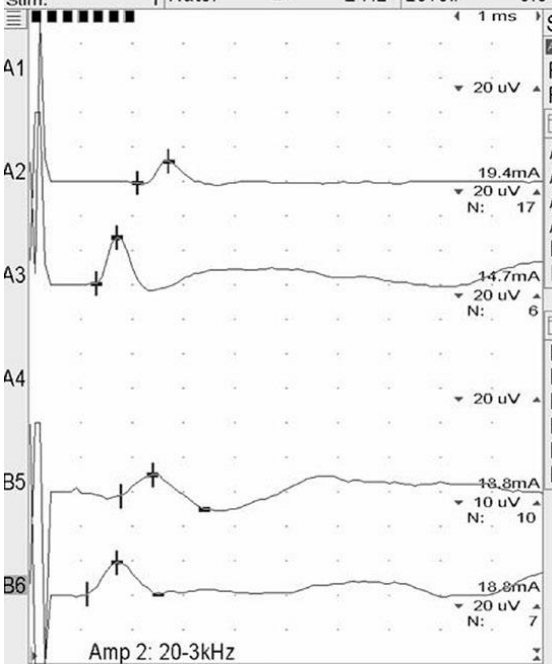
Step: 6 | Average: On | Sig. Enhancer: On

A1 A2 A3 A4 B5 B6

Recording Site A: Wrist
Recording Site B: Wrist

Stimulus Site	Lat1 ms	Lat2 ms	Amp uV
A1: Digit II (index finger)			
A2: Digit III (long finger)	2.1	2.7	14
A3: Mid palm	1.3	1.7	27
A4: Digit IV (ring finger)			
B5: Digit V (little finger)	1.8	2.4	10
B6: Mid palm	1.1	1.7	18

Segment	Dist mm	CV m/s
Digit II (index finger)-Wrist		
Digit III (long finger)-Wrist	131	62
Mid palm-Wrist	80	62
Digit IV (ring finger)-Wrist		
Digit V (little finger)-Wrist	110	62
Mid palm-Wrist	80	71



Tratamiento

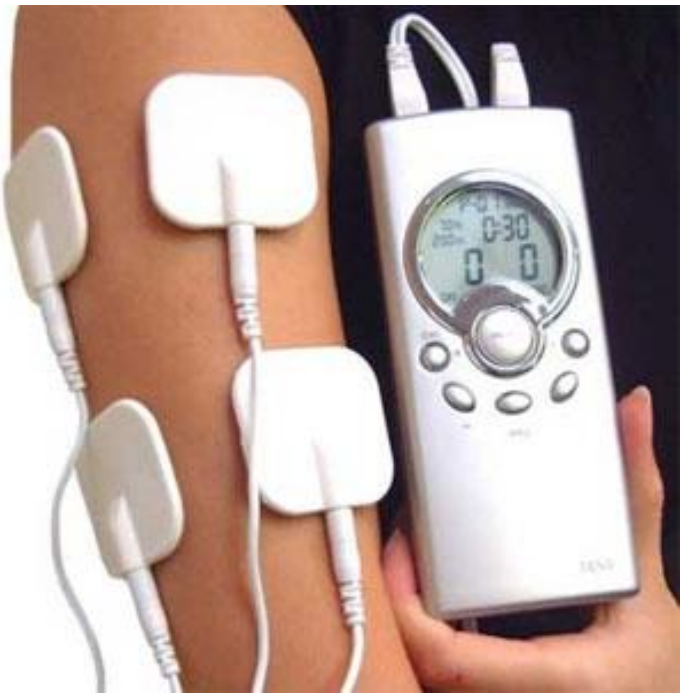
A. Debe ser específico para la etiología.
B. El diagnóstico certero en patologías poco comunes (gammopatías, autonómicas, hereditarias, etc) es difícil.
C. Habitualmente el tratamiento es sintomático

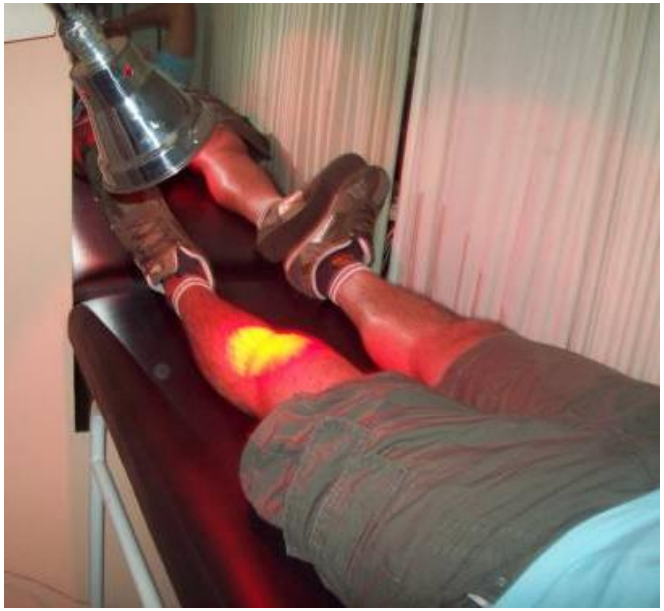
Tratamiento Sintomático

ADT. Amitriptilina

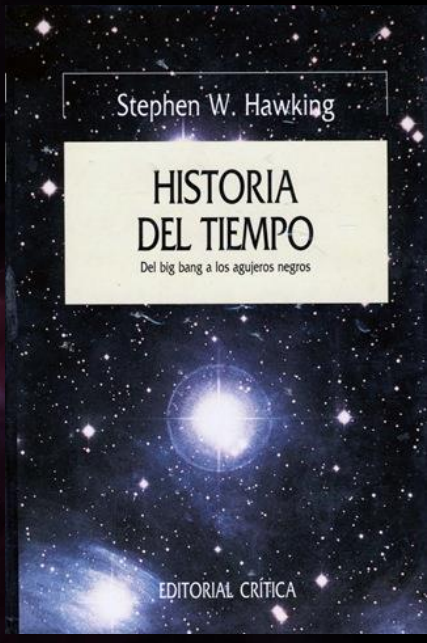
FAE. Carbamacepina, topiramato
gabapentina, pregabalina,
duloxetina

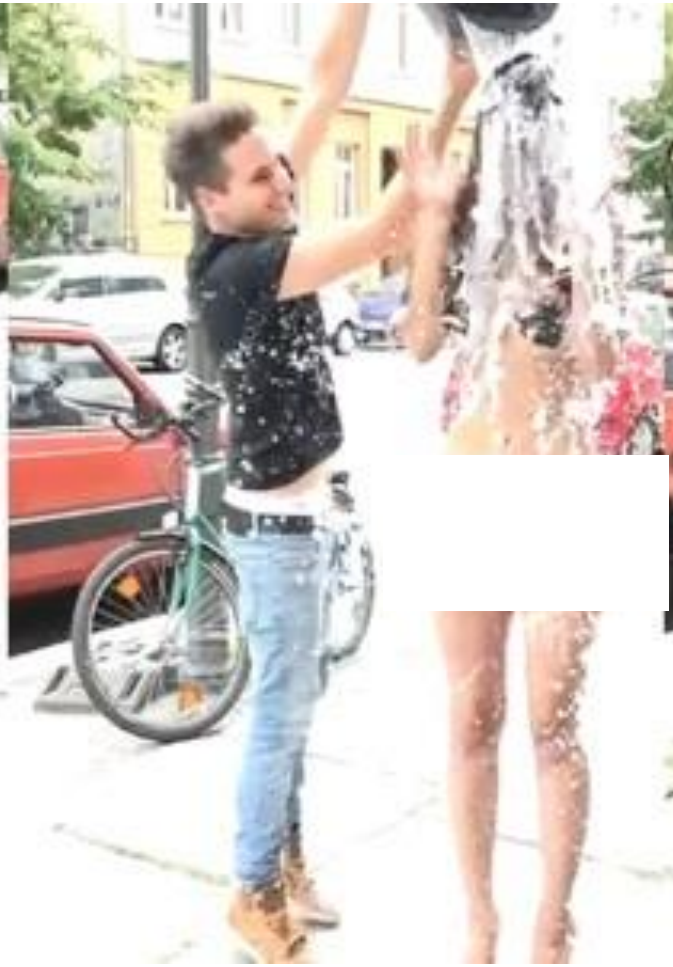
Dosis bajas.





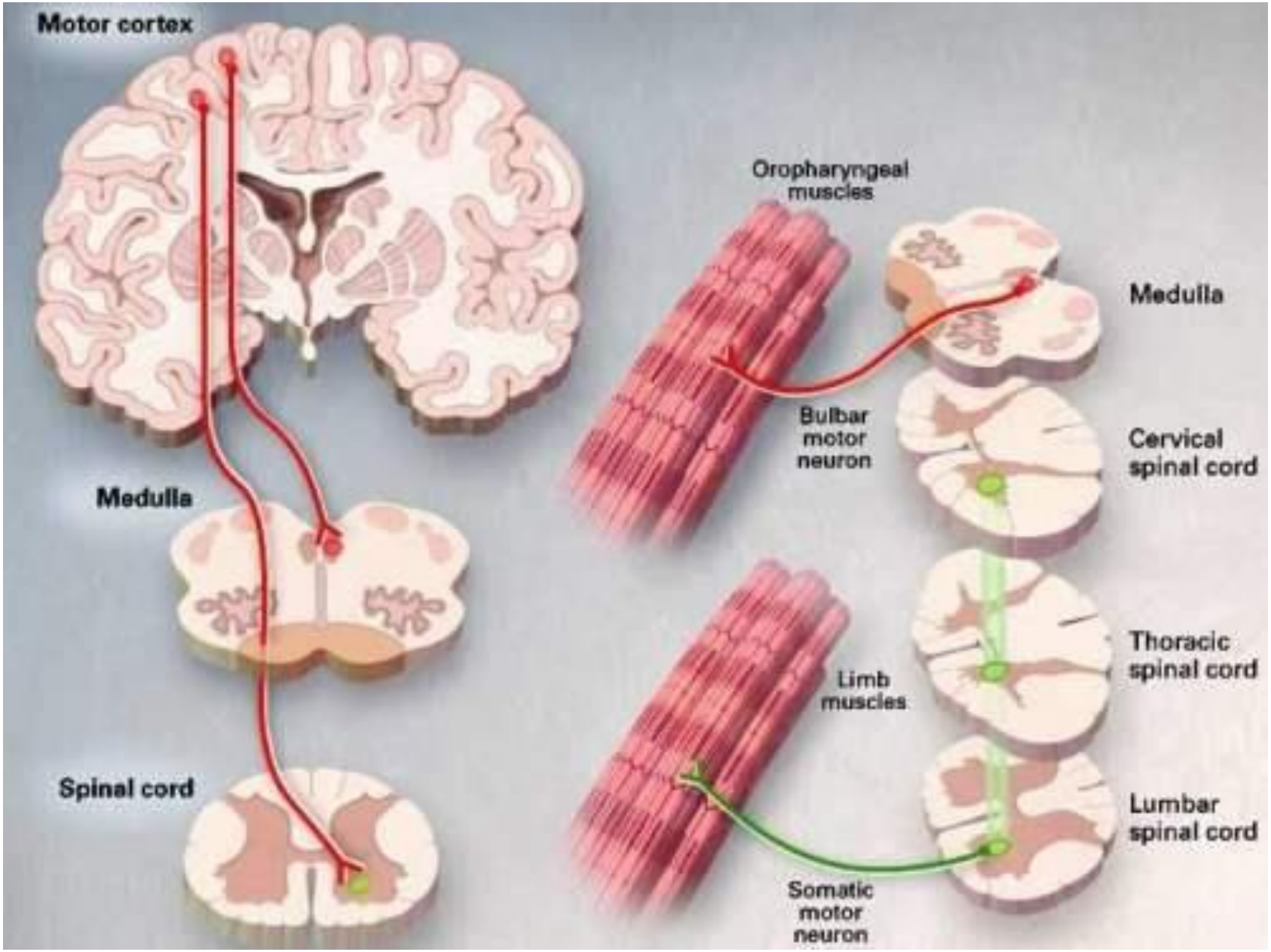
Otra Patología


Stephen W. Hawking
HISTORIA DEL TIEMPO
Del big bang a los agujeros negros
EDITORIAL CRÍTICA


Lou Gehrig

Stephen Hawking


Motor cortex
Medulla
Spinal cord
Oropharyngeal muscles
Bulbar motor neuron
Medulla
Cervical spinal cord
Limb muscles
Thoracic spinal cord
Lumbar spinal cord
Somatic motor neuron

Esclerosis Lateral Amiotrófica
ELA (ALS)

TABLE 3-4 Amyotrophic Lateral Sclerosis Features

Men more than women
Clusters
Peak at 50–70 years
Asymmetrical
Upper and lower motor neuron findings
Cramps
Fasciculations
Lack of pain early
Respiratory insufficiency begins nocturnally
Emotional lability
Lack of bowel and bladder involvement
Paucity of sensory findings

Criterios Diagnósticos

El Escorial

Tratamiento

Todo y Nada

Síndrome de Guillain-Barre

GBS

- Eponym that **acute immune-mediated polyneuropathies**
- Myelin is target
- Starts at level of nerve root=conduction blocks & muscle weakness

Octubre 1916
2 casos

Georges Guillain
(1880-1967)

André Strohl
(1887-1977)

Jean Alexandre Barré
(1880-1967)

The New England
Journal of Medicine

Volume 255 JULY 12, 1956 Number 2

AN UNUSUAL VARIANT OF ACUTE IDIOPATHIC POLYNEURITIS (SYNDROME OF OPHTHALMOPLEGIA, ATAXIA AND AREFLEXIA)*

MILLER FISHER, M.D.†

BOSTON

THE purpose of this communication is to report 3 cases of an acute neurologic illness characterized among other features by total external ophthalmoplegia, severe ataxia and loss of the tendon reflexes. The clinical picture in all 3 cases was so similar as to constitute an easily recognizable syndrome. The presenting symptoms and signs were most alarming for the attending physician on each occasion — unnecessarily so, since the course of the illness appears to be benign. The cause of the syndrome was obscure until, in the most recent case, a great rise in the protein of the cerebrospinal fluid in the late stages of the illness indicated a close relation to the Guillain–Barré type of polyneuropathy. Although it is therefore likely that the illness under discussion is but another variant of so-called infective polyneuritis in which limb involvement is minor, the diagnosis is hardly to be suspected at the onset unless one realizes that such an atypical form exists. Moreover, several features of the neurologic disturbance are worthy of emphasis — for example, the striking symmetry of the ocular palsies and the severity of the ataxia. For these reasons it is considered worth while to call attention to this interesting though rare condition.

Several different terms have been used for the type of polyneuropathy referred to herein, such as Guillain–Barré syndrome, acute infective polyneuritis, for 3 days. On the 1st day of illness, a slight transient dizziness had been noted — not a rotational feeling, but an unsteady sensation in the head. Later in the day there was a feeling of malaise, and the chest felt as if it were tightening up "like an ordinary cold." About 4 p.m., on trying to climb a stepladder, he noted that the unsteady feeling in the head had become worse. When he awoke the next morning his head "felt like a balloon," and on going to the bathroom, he walked "like a drunken sailor." The feet tended to lift up very lightly like feathers and could not be placed properly on the floor again but instead plopped down. The dizziness in the head seemed distinct from this disturbance of gait. At some time during the 2d day horizontal diplopia appeared on near vision, but not for distant objects. Vision was not as distinct as usual. In the evening the fingers of both hands became numb, and the arms felt sore and lame. The lower limbs were not affected. The tightness in the chest, which had become worse, was situated deep and centrally, directly behind the lower sternum. A mild, deep cough had appeared, and coughing produced a sharp, splitting pain in the mid-frontal region. The patient did not use alcoholic beverages and had been taking a well balanced diet up to the onset of his illness.

At the 1st hospital, examination by Dr. Robert K. Davis, on the 3d day of illness, showed paralysis of abduction of the right eye and questionable impairment of the 4th nerve. The pupils were large and reacted only slightly to light. The deep tendon reflexes were hypoactive (the patient thought they had always been so), and there was marked ataxia on the finger-nose and heel-knee tests. Flexion of the right arm was weak. The temperature was 99°F., and the blood pressure 120/76. Examination of the blood and urine was not remarkable. The blood Wassermann test was negative. The cerebrospinal fluid examined on admission was clear, and contained 1 lymphocyte per cubic millimeter. The sugar was 72 mg., and the protein 36 mg.

Charles Miller Fisher (1913-2012)

1956 3 Casos

GBS

- 1.2- 2.3/100,000 hab
- H:M igual, pero en México es mayor en H
- Cualquier edad

Pathophysiology

- Usually postinfectious
- Ab production against specific gangliosides/glycolipids
- Conduction block and flaccid paralysis

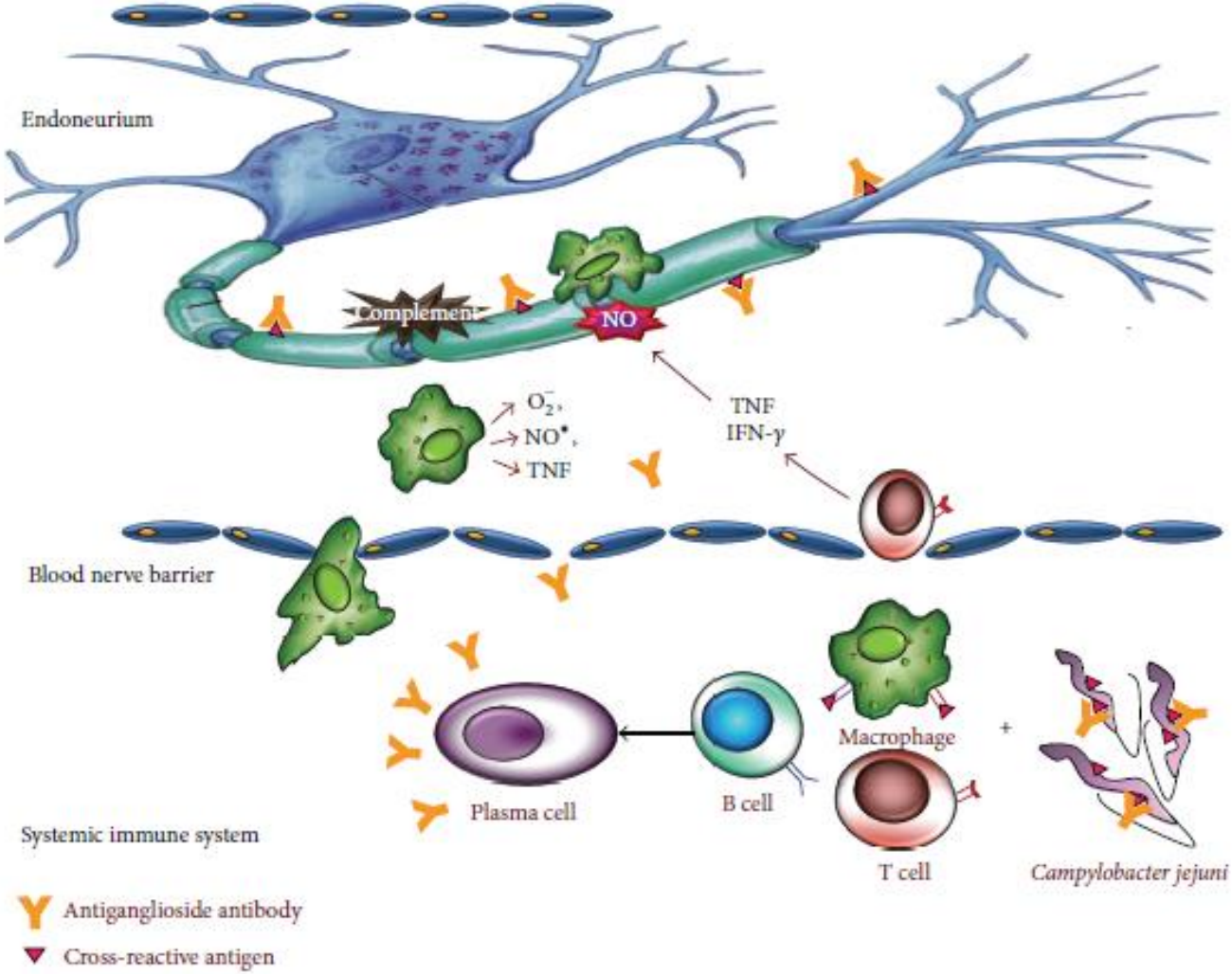


FIGURE 3: Origin and contribution of antiganglioside antibodies and *C. jejuni* infection to Guillain-Barré syndrome pathogenesis. A bacterial cross-reactive antigen recognized by macrophages and T cells that help B cells to produce antiganglioside antibodies, which penetrate blood-nerve barrier and activate complement. These antibodies bind with specific nerve gangliosides and *C. jejuni* antigen as well. Activated endoneurial macrophages release cytokine and free radicals (nitric oxide), invade compact myelin, periaxonal space, and sometimes block nerve conduction or cause axonal degeneration. Activated T cells release proinflammatory cytokines, fix complement, damage Schwann cell, and ultimately produce dissolution of myelin.

BioMed Research International
Volume 2013, Article ID 852195, 13 pages

Clinical Features:

- Progressive, fairly symmetric muscle weakness

  -typically starts in proximal legs

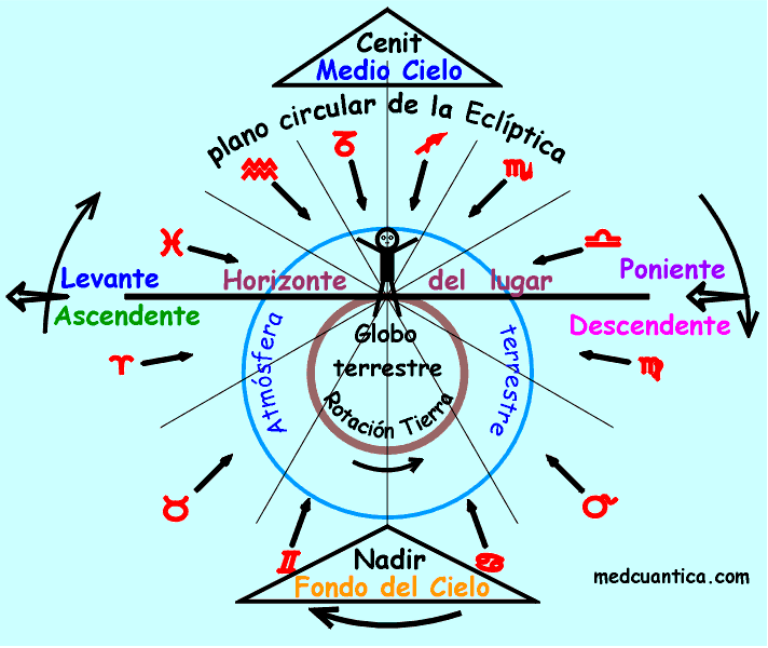
  -10% will 1st develop weakness in **face** or arms

  -severe resp muscle weakness in **10-30%** pts

  -oropharyngeal weakness in ~ 50%

Clinical Features:

- Aprox. 50% pts tienen el nadir 2 semanas
- 90% lo alcanza en 4 semanas



Course

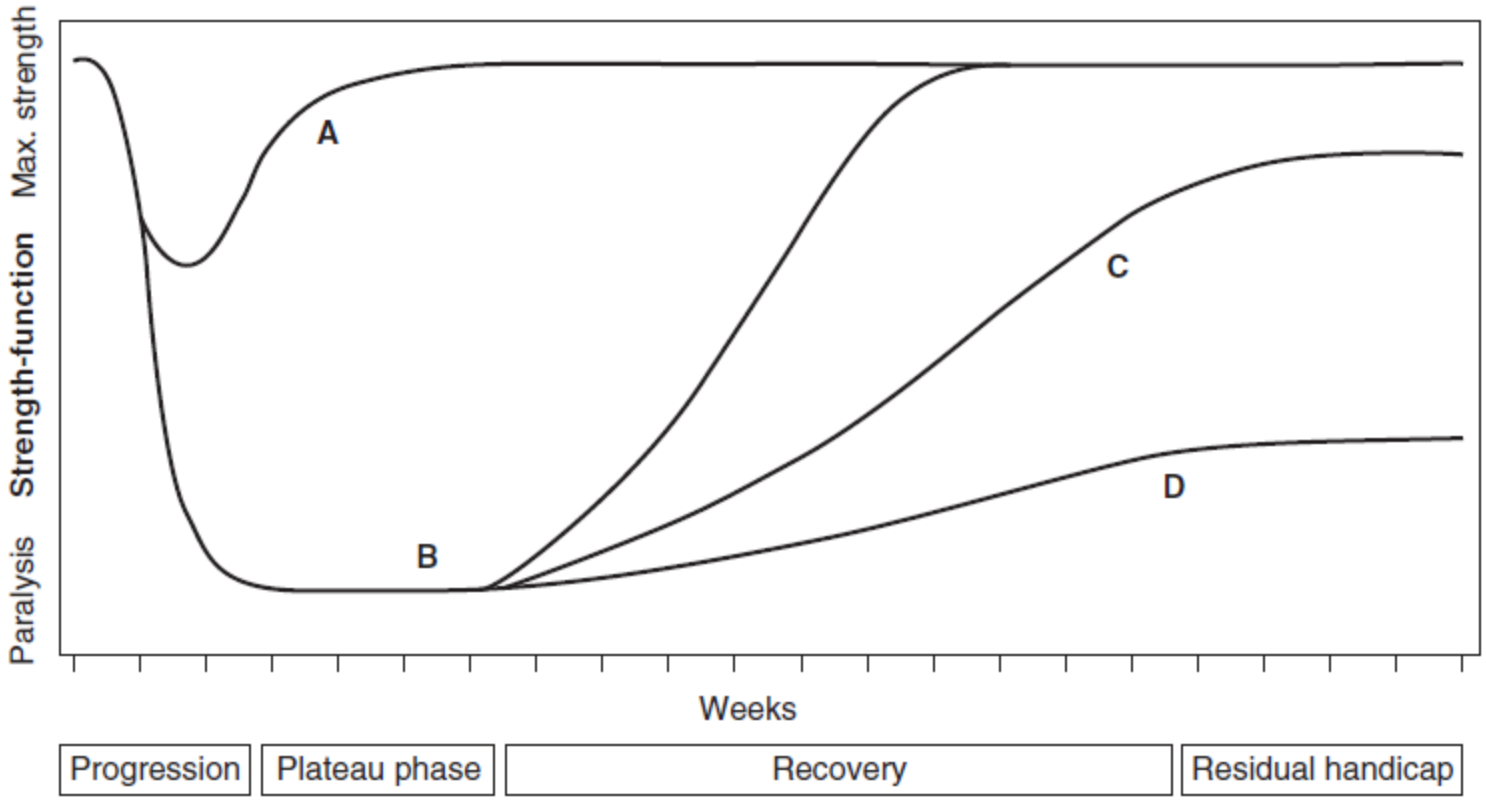


Figure 1. Heterogeneous disease course in GBS. Variation in course of disease in GBS. Indicated are the courses of disease mildly (A) and severely affected GBS patients (B – D). Some GBS patients have a severe course and recover completely (B) wh other patients have a severe course and remain mildly to severely disabled for a long period of time (C, D).

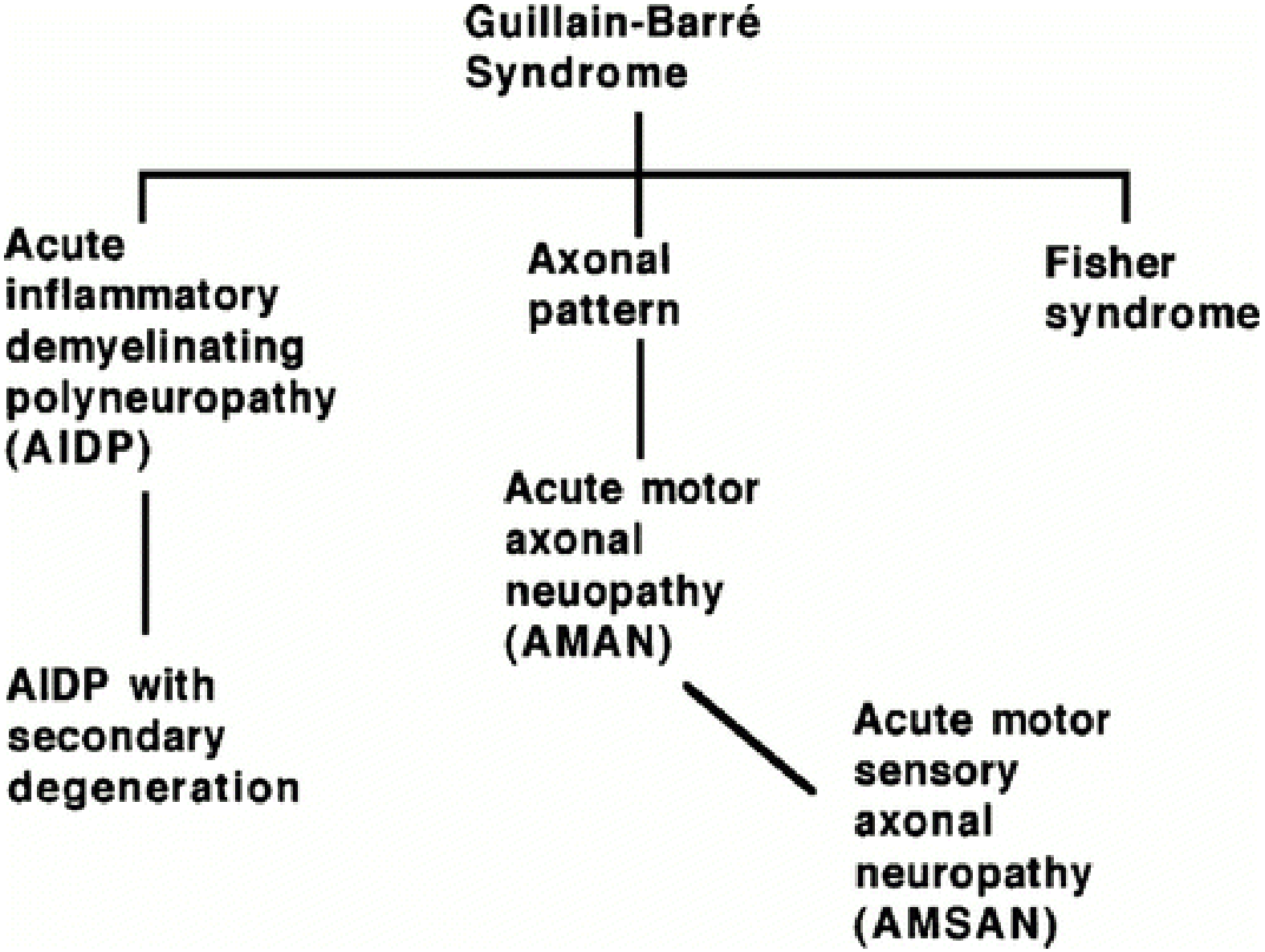
Diagnosis:

Criterios de Asbury

- **I. Requeridos** para el dx:
  - A. Debilidad Progresiva de > 1 extremidad
  - B. Areflexia
- **II. Apoyan** el dx CLÍNICO:
  - A. Progresión
  - B. Simetría (relativa)
  - C. Síntomas sensitivos leves
  - D. Involucro de Nervios Craneales. VII bilateral
  - E. Disfunción autonómica
  - F. Ausencia de fiebre

GBS=heterogenous syndrome w/ variant forms

- Think of **AIDP** as the traditional form as described previously, accts for 85-90%
- **Miller Fisher Syndrome**: opthalmoplegia, ataxia, and areflexia (5%). GQ1b antibody. Only 1/4th w/ extremity weakness
- **AMAN**: selective involv of motor nerves, DTRs are preserved, more common in Japan/China, almost all preceded by Campylobacter infxn
- **AMSAN**: more severe form of AMAN +sensory


Guillain-Barré Syndrome
Acute inflammatory demyelinating polyneuropathy (AIDP)
Axonal pattern
Fisher syndrome
AIDP with secondary degeneration
Acute motor axonal neuopathy (AMAN)
Acute motor sensory axonal neuropathy (AMSAN)

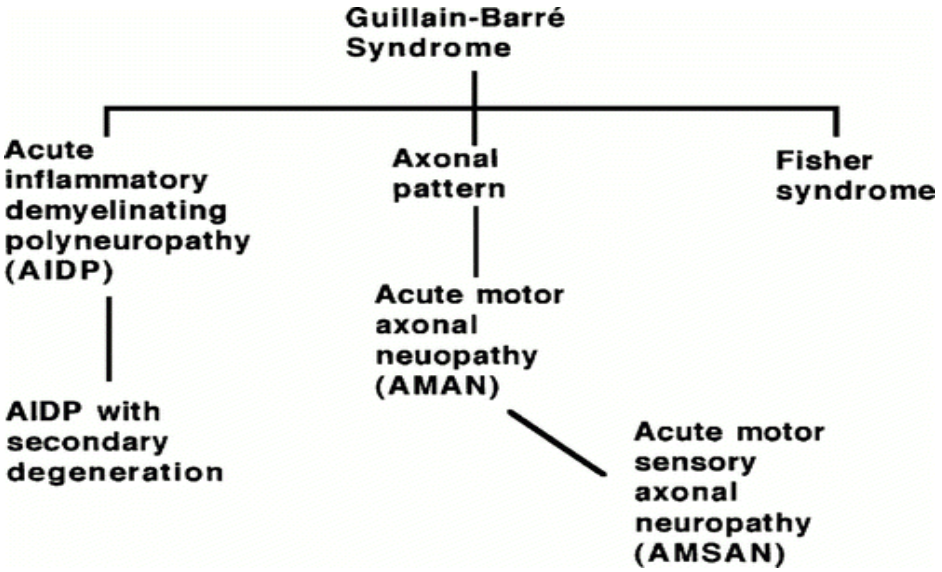
Aceptado: 24 de octubre de 2006

Subtipos electrofisiológicos del síndrome de Guillain-Barré en adultos mexicanos

Edgar Alfredo Zúñiga-González,[1] Antonio Rodríguez-de la Cruz,[2] Jesús Millán-Padilla[2]

[1]Neurólogo
[2]Neurofisiólogo

Results: The study included fifty-one patients with median age of 45.5 years (range 16-79); 37 men and 14 women, with a ratio man-woman 2.6:1; 37% were observed in winter (p = 0.56). The electrophysiological subtypes were acute motor axonal neuropathy (AMAN) 39 %; acute inflammatory demyelinating polyneuropathy (AIDP) 23.5 %; mixed neuropathy (MN) 20%; and acute motor sensory axonal neuropathy (AMSAN) 17.5 %.
Conclusions: the most frequent electrophysiological subtype in Mexican patients was AMAN. Other frequent subtypes were MN and AMSAN. Axonal variants were the predominant types.



Rev Med Inst Mex Seguro Soc 2007; 45 (5): 463-468

DDx of Polyneuropathy:

- Arsenic poisoning
- N-Hexane (glue sniffing)
- Vasculitis
- Lyme Disease
- Tick paralysis
- Sarcoidosis
- Leptomeningeal Dz
- Paraneoplastic Dz
- Critical Illness

Escala de Discapacidad (Hughes)

0. Sano
1. Signos y síntomas leves, que no impiden actividad normal
2. Camina más de 10 metros sin ayuda
3. Camina más de 10 metros con ayuda o apoyo
4. Confinado a cama o silla de ruedas
5. Con ventilación mecánica

Disease Modifying Treatment

- **IV IG** : typically given for 5 d at 0.4 gram/kg/d (may need to extend course depending on response)

- **Plasmapheresis**: usually 4-6 treatments over 8-10 days

Glucocorticoids have NO ROLE!!

Outcomes:

- 65% can walk independently @ 6 mos
- Overall, **80%** usually recover completely
- Approx **3-5% die** despite ICU care
- 2% will develop chronic relapsing Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP)

Future:

- Vaccines
- Acs Monoclonales. Eculizimab
- APT070. complement inhibitor C3/C5
- rEV576 is a recombinant form of saliva protein of a soft tick (Ornithodoros moubata)

Día Mundial del EVC
29 octubre