

# 周邊神經與肌肉疾病

花蓮慈濟醫院 神經內科部

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# Overview

- Peripheral nerve lesion
- Peripheral neuropathy symptoms
- Peripheral neuropathy pattern
  - Mononeuropathy
  - Mononeuropathy multiplex
  - Polyneuropathy
    - Acquired : Guillain-Barre Syndrome
    - Hereditary: Charcot-Marie-Tooth disease

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# Peripheral nerve lesions

- Spinal cord (Myelopathy)

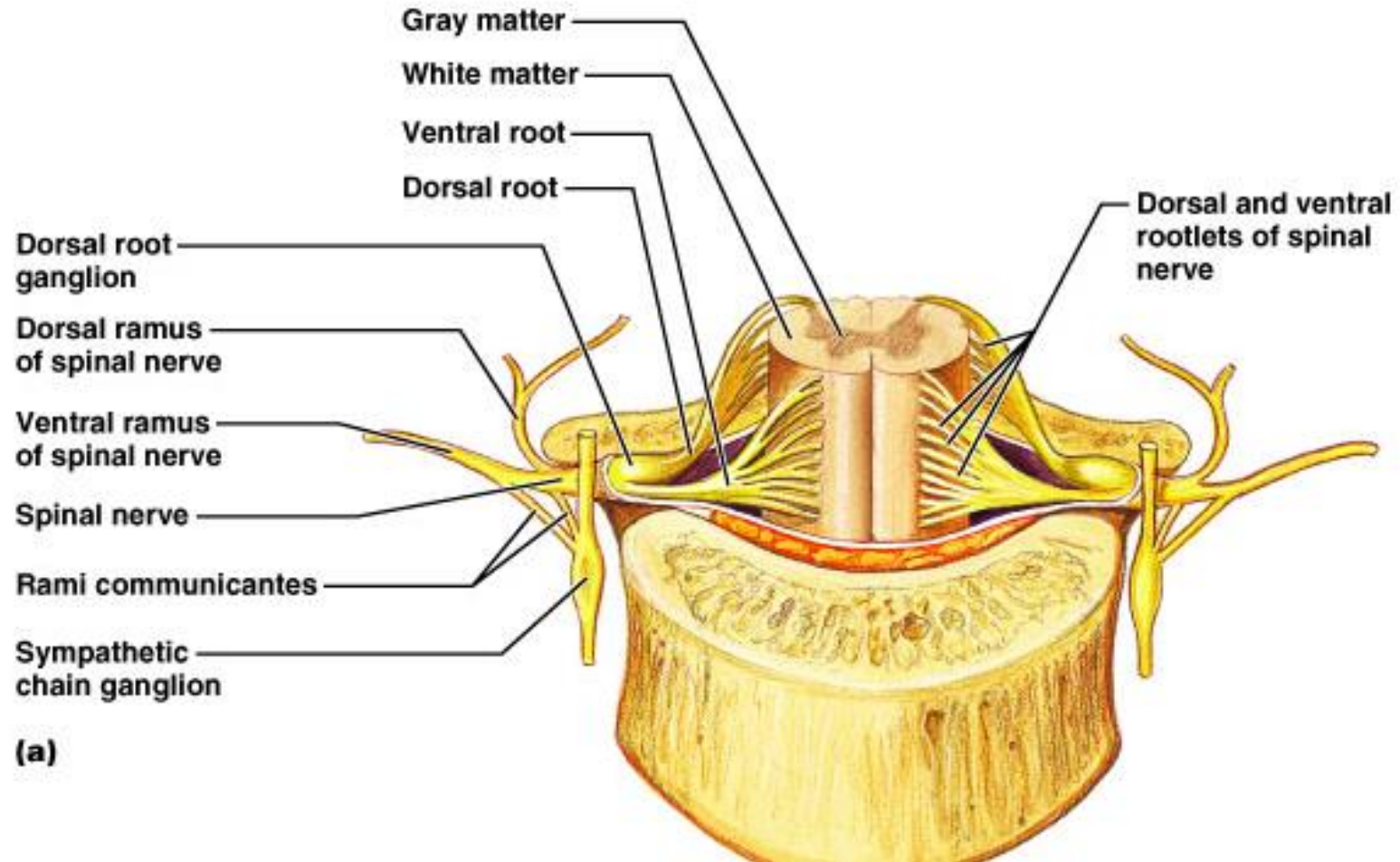
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- Root (Radiculopathy)

- Plexus (Plexopathy)

- Nerve (Neuropathy)

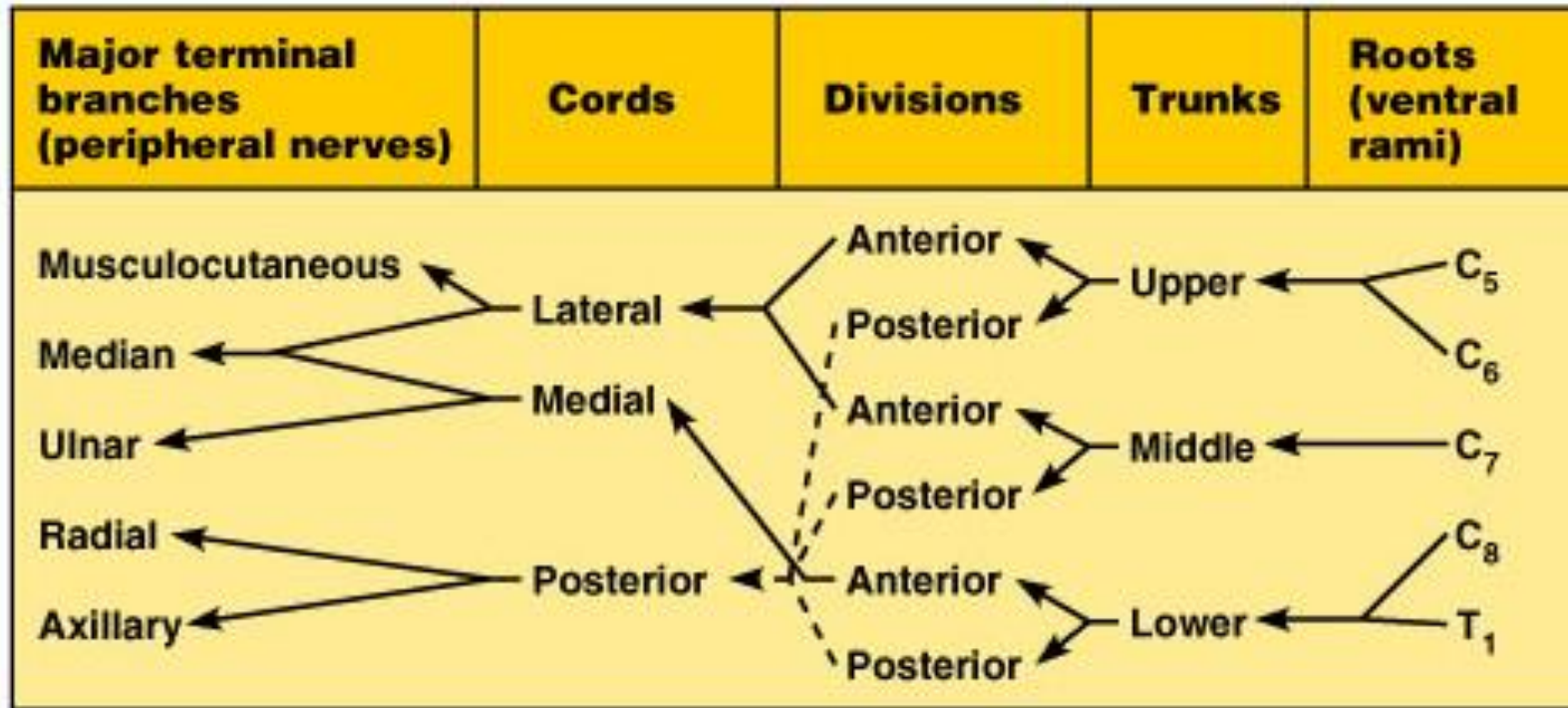
# Myelopathy, Radiculopathy



# Myeloradiculopathy



# Plexopathy (Plexus lesion)



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# Peripheral neuropathy symptoms

- Motor
- Sensory
- Autonomic

# Motor

- Weakness
- Muscle atrophy
- Fasciculation
- Decreased deep tendon reflex

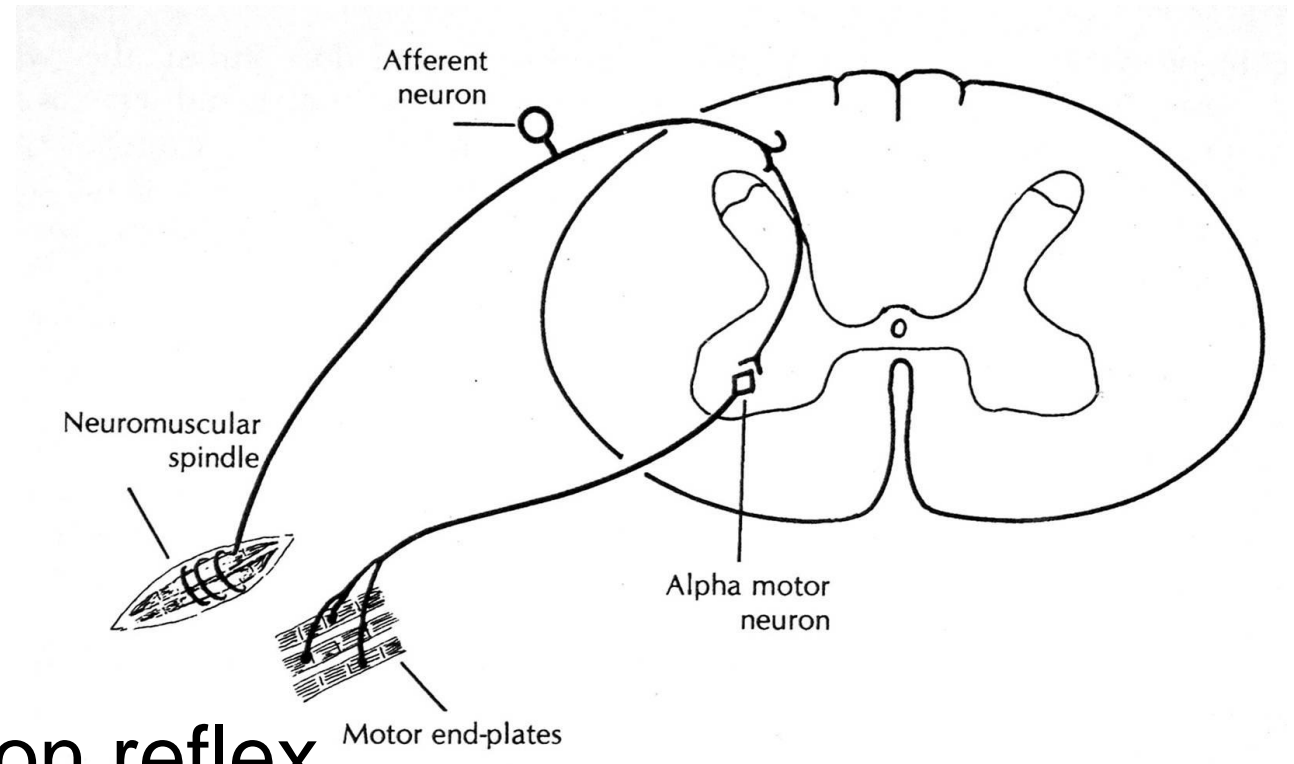
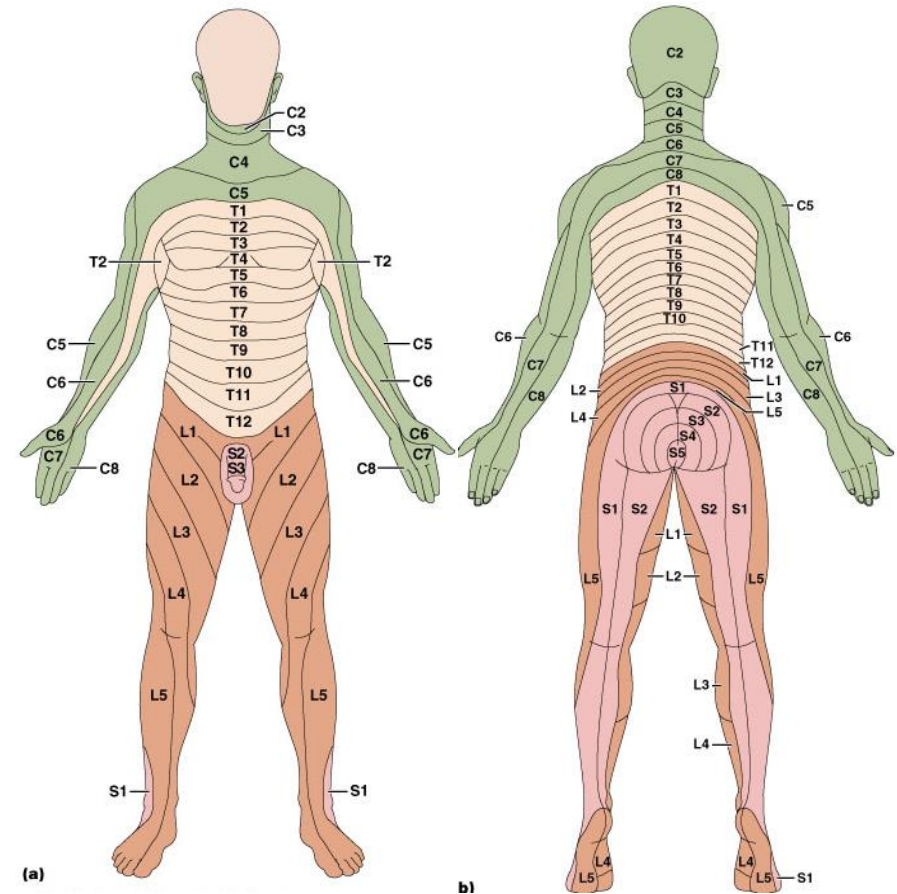


Figure 5-11. Monosynaptic stretch reflex arc.

# Sensory

- Dermatome pattern
- One peripheral nerve distribution pattern
- Asymmetrical, Symmetrical (Glove-stock pattern)



# Autonomic

## 6 AREAS AFFECTED WHEN AUTONOMIC NERVES ARE DAMAGED

### EYES

*Difficulty adjusting from  
light to dark*

### BLADDER

*Loss of bladder control  
Urinary retention  
Urinary tract infections*

### DIGESTIVE SYSTEM

*Indigestion or heartburn  
Nausea or vomiting  
Diarrhea or constipation  
Bloating  
Loss of appetite*

### HEART & BLOOD VESSELS

*Dizziness or fainting  
Difficulty breathing  
Abnormal blood pressure  
High heart rate  
Heart attack (without  
warning signs)*

### SEX ORGANS

*Erectile dysfunction  
Difficulty achieving orgasm  
during sex (women)  
Vaginal dryness*

### SWEAT GLANDS

*Lack of sweat  
Excessive sweating  
Dry skin on feet*



# 醫師國考題

- 下列何者非周邊神經病變之常見症狀？
  - A. 運動功能障礙
  - B. 肌腱反射增強
  - C. 感覺功能障礙
  - D. 自律神經病變

Ans: B

# Overview

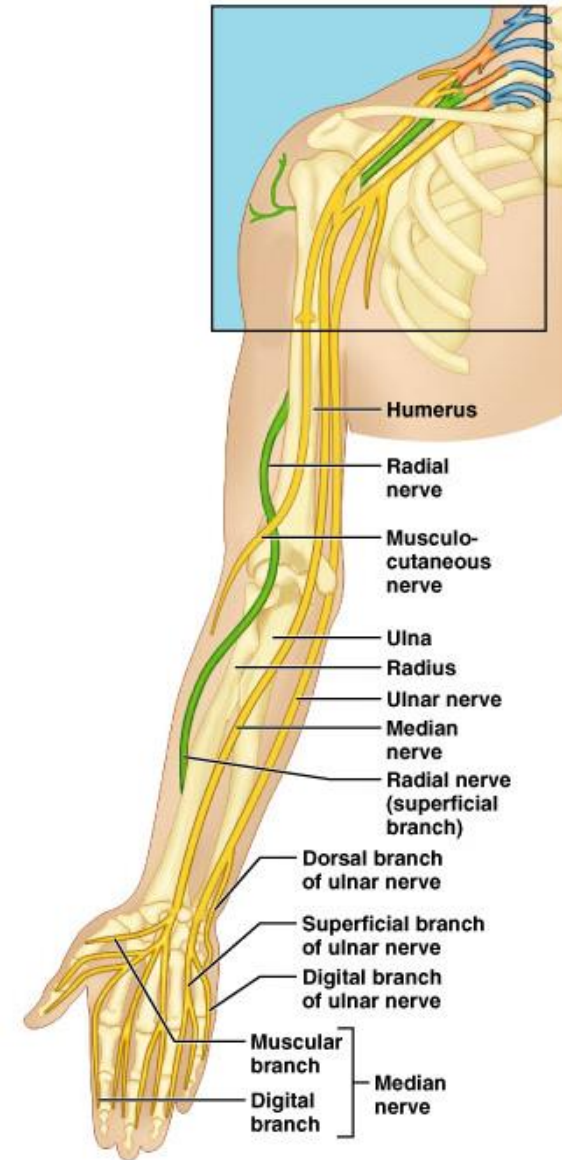
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# Mononeuropathy:

Entrapment neuropathy  
Compressive neuropathy



(c)

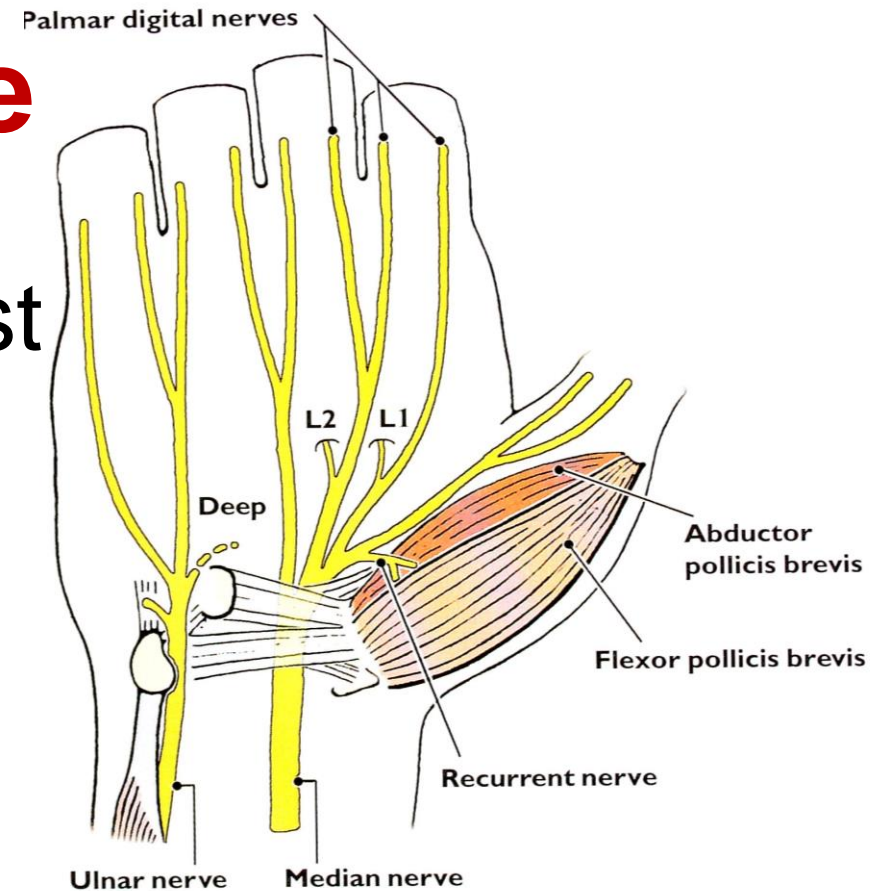


# Carpal tunnel syndrome

Median neuropathy at the wrist

Symptoms:

Signs: Tinel sign/ Phalen test

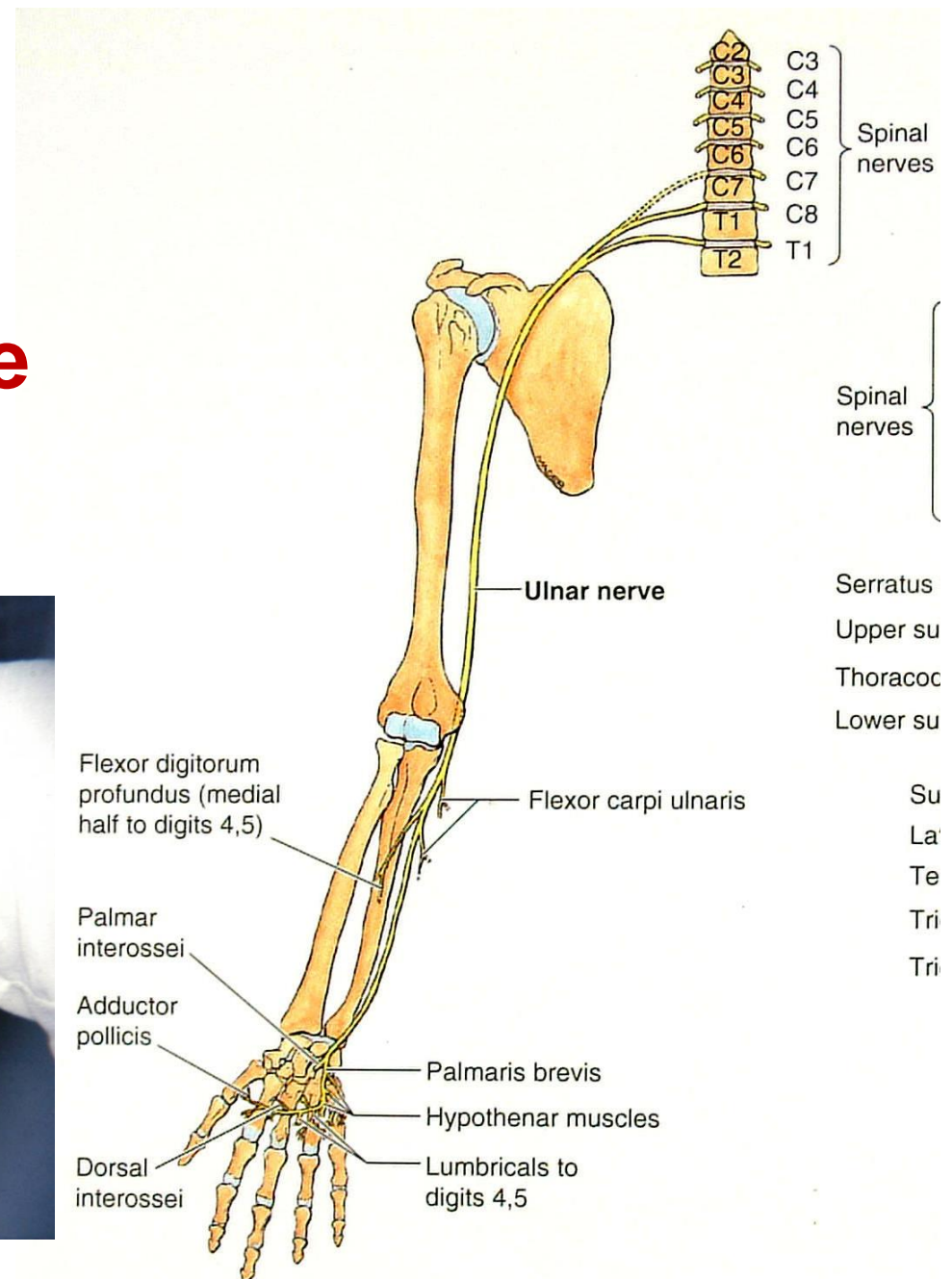


**Figure 6.38.** Essential nerves of the hand: median and ulnar nerves (L1 and L2, lumbrical branches of median nerve).

# Tardy ulnar palsy

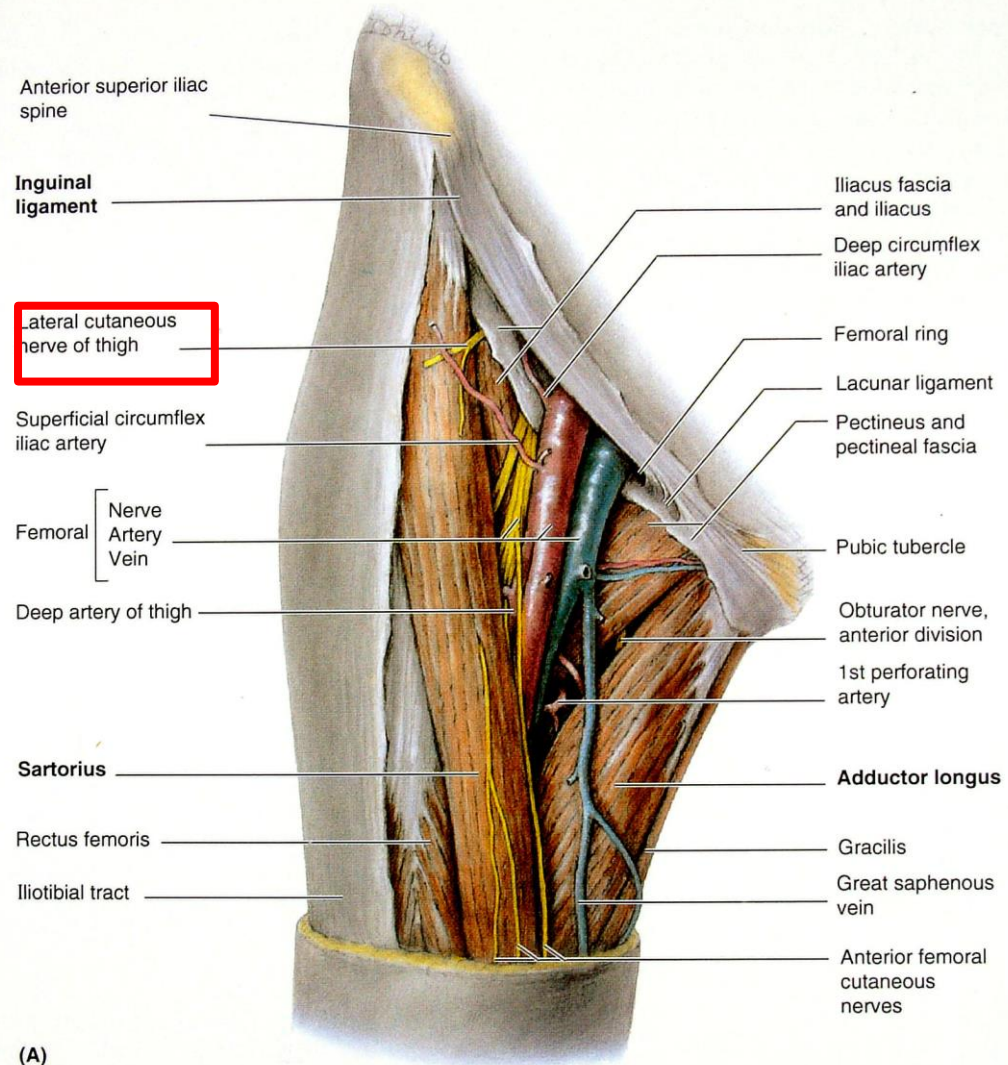
# Cubital tunnel syndrome

## Ulnar nerve



# Meralgia paresthetica

## Lateral femoral



Lateral Femoral Cutaneous Nerve

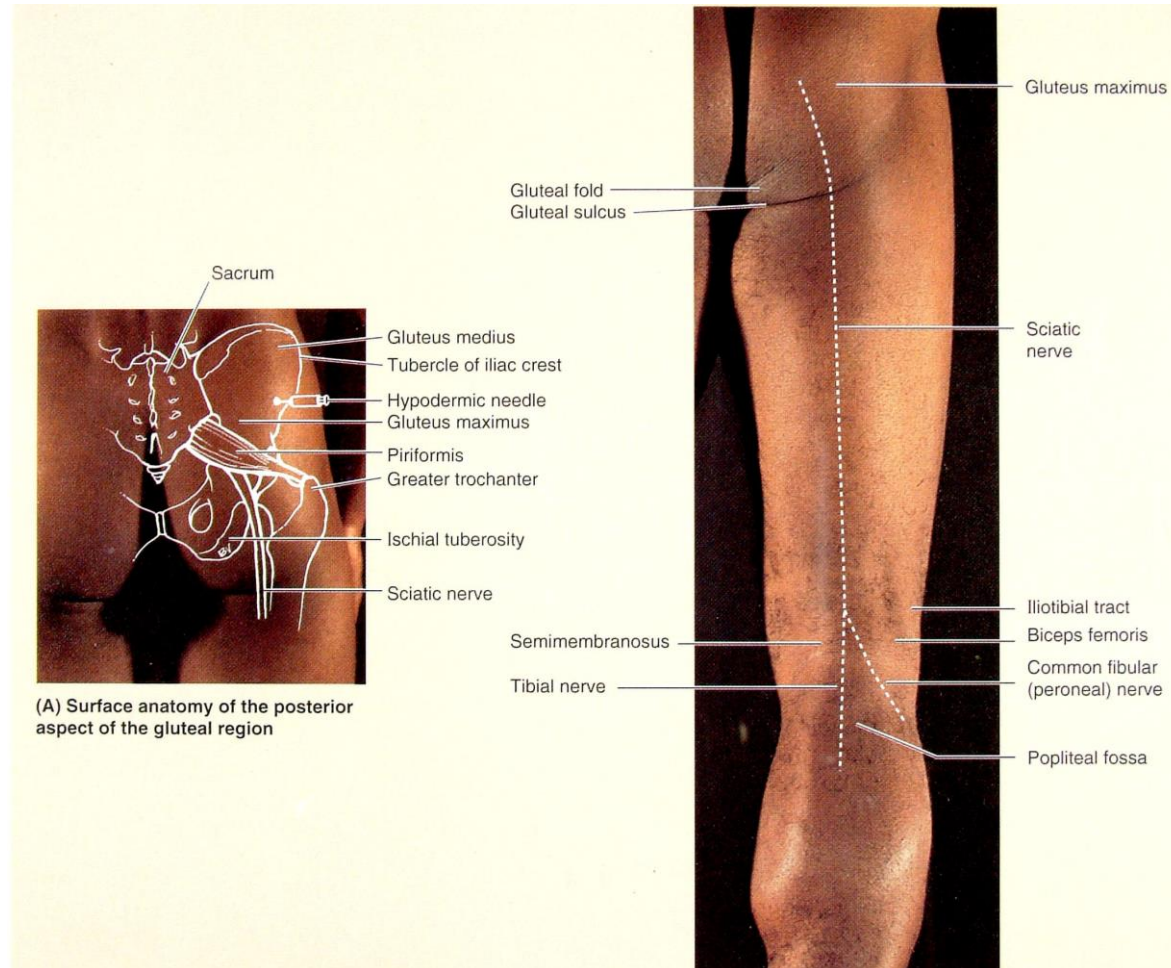
Sensory Distribution of Lateral Femoral Cutaneous Nerve



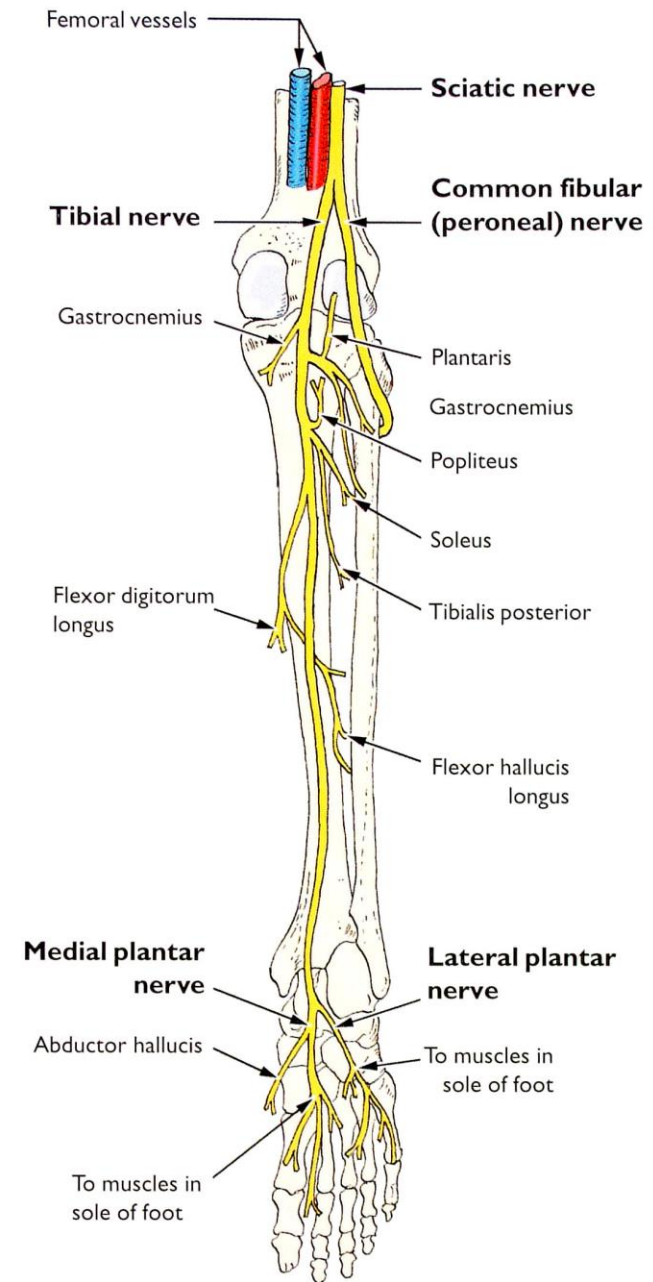
# Sciatica

Lumbar-sacral radiculopathy

Piriformissyndrome



# Common peroneal nerve palsy



**Figure 5.32.** Scheme of motor distribution of the tibial nerve in the posterior crural compartment and in the sole of the foot.

# Mononeuropathy

Infection, Inflammatory

Facial nerve palsy



# Mononeuropathy or Radiculopathy

Herpes zoster: Ganglionopathy

Sensory ganglion

Geniculate ganglion, Trigeminal  
ganglion, Dorsal root ganglion



# 醫師國考題

- 有關疱疹病毒神經病變 ( neuropathy of herpes zoster ) 之敘述，下列何者最不恰當？
  - A. 可能引起角膜 ( cornea ) 損傷
  - B. 疱疹病毒主要侵犯運動神經元
  - C. 好發於肋間神經
  - D. 好發於三叉神經第一分支

A n s : B



# Ramsay-Hunt syndrome

Geniculate ganglion



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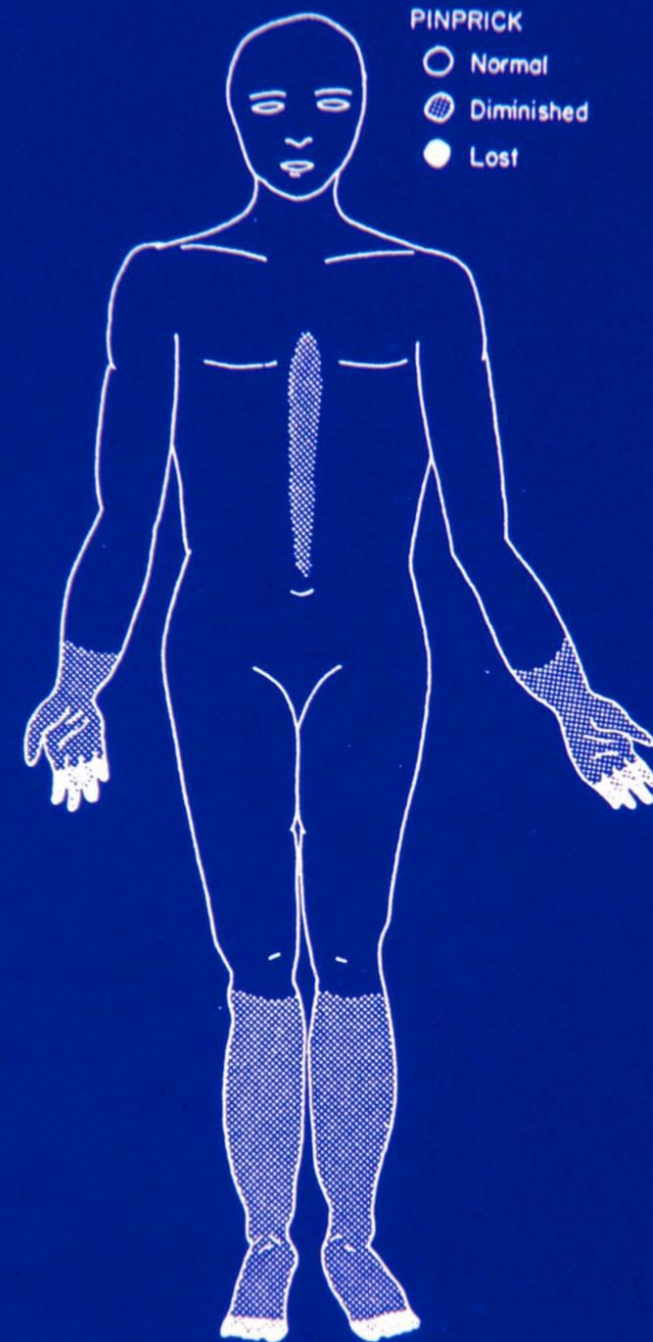
# Mononeuropathy multiplex

- Autoimmune disease
- Rheumatic arthritis
- Systemic lupus erythematosus (SLE)
- Sjogren syndrome (Sicca complex)
- Vasculitis (Polyarteritis nodosa)
- Malignancy
- DM

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# Polyneuropathy: Symmetrical Stocking-glove pattern



**FIGURE 6.** Stocking-glove pattern of sensory loss in an advanced stage of distal axonopathy. The area of diminished sensation over midthorax ("cui-rass distribution") reflects involvement of distal ends of intercostal nerves.

# **Polyneuropathy: Acquired, Hereditary**

- **Diabetes**
- **Alcohol**
- **Nutrition**
- **Guillain-Barre syndrome**
- **Trauma (Radiation injury, Burn injury, Electric shock)**
- **Hereditary**
- **Environment (Toxin, Drugs)**
- **Rheumatic (Autoimmune disease)**
- **Amyloid**
- **Paraneoplastic (Malignancy)**
- **Infection**
- **Systemic disease (Metabolic, hepatic, Uremic, Thyroid)**
- **Tumor**

# Clinical symptoms and signs

- **Motor deficits**
- **Decreases in tendon reflexes**
- **Fasciculation and cramp**
- **Hypoesthesia and hyperesthesia**
- **Paresthesia and dysesthesia**
- **Sensory ataxia and tremor**
- **Deformity and trophic changes**
- **Autonomic disorders**

# **Polyneuropathy: Acquired, Hereditary**

## **Acquired**

**Guillain-Barre Syndrome (GBS)**

**Acute Inflammatory Demyelinating  
Polyneuropathy (AIDP)**

**Chronic Inflammatory Demyelinating  
Polyneuropathy (CIDP)**

## **Hereditary**

**Hereditary Sensory Motor Polyneuropathy**

**Charcot-Marie-Tooth disease**

**Hereditary Neuropathy with Liability to  
Pressure Palsy (HNPP)**



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# 新聞報導

## 接種賽諾菲出現2例神經病變 與流感疫苗關聯待查

2020年10月31日 中央社



(中央社記者張茗暄台北31日電) 繼台中一名男子接種流感疫苗後出現神經病變症狀，有名基隆女子也現疑似症狀。疫情指揮中心發言人莊人祥表示，2個案均接種賽諾菲疫苗，但批號不同，是否與疫苗有關仍待釐清。

基隆一名50多歲李姓女子接種流感疫苗9天後四肢突然發麻且有味覺喪失症狀，向中央通報疫苗不良反應事件，後續出現吞嚥困難、無法進食等症狀，目前插管住進加護病房，院方初步懷疑為急性多發性神經炎，即所謂的「格林巴利症候群」(GBS)。

中央流行疫情指揮中心發言人莊人祥表示，多數研究都顯示，流感疫苗和此疾病沒有相關，僅少數研究顯示有微小相關，且個案出現味覺喪失症狀，有醫師研判和格林巴利症候群相關性可能不大，另因病毒感染都可能造成味覺喪失，如無相關危險因子，不會特別採檢。

# Guillain-Barre Syndrome

## Risk Factors:

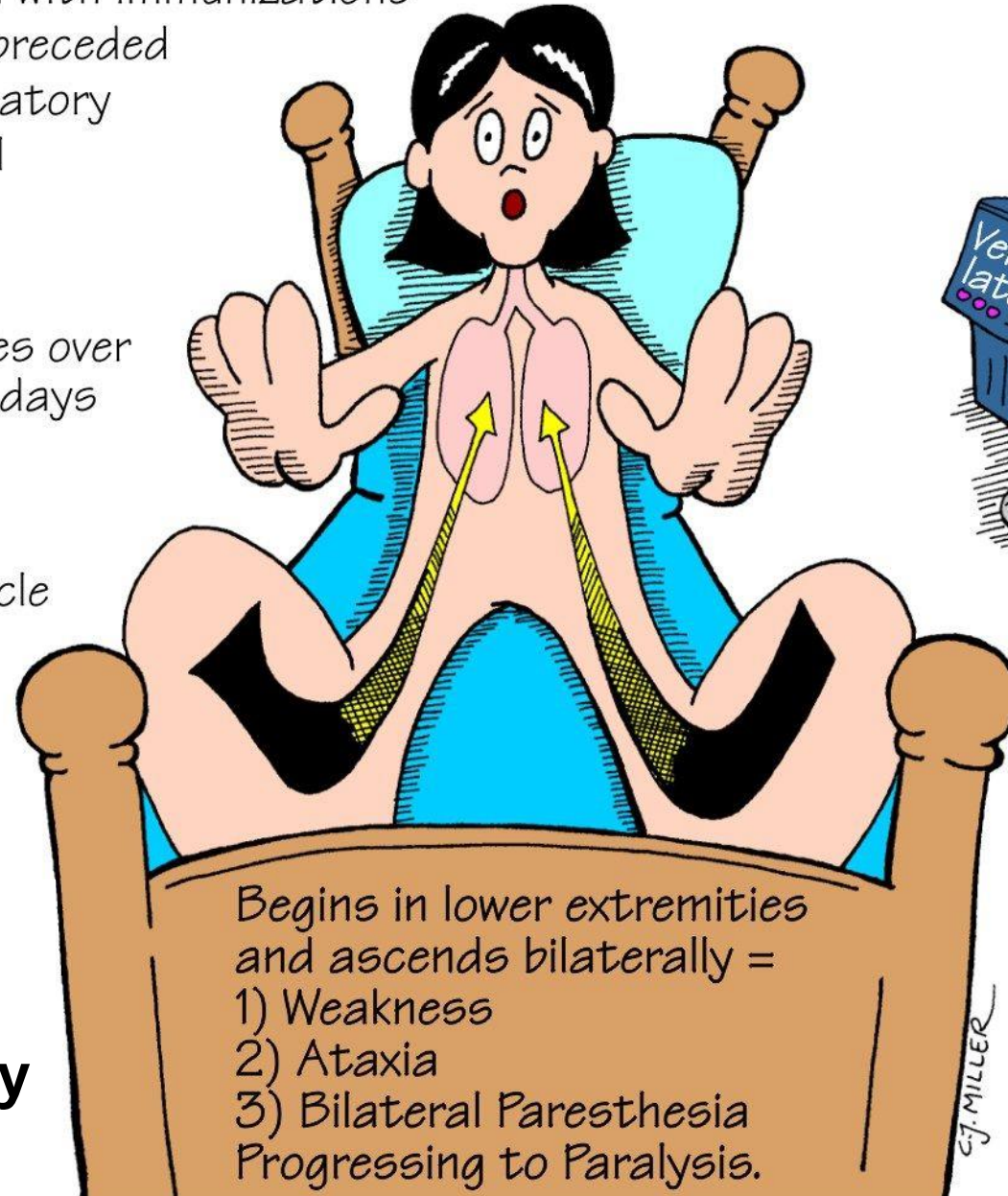
- Possibly Autoimmune
- Association with Immunizations
- Frequently preceded by mild respiratory or intestinal infection

- Progresses over hours to days

- Minimal Muscle Atrophy

NSG  
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## Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP)



Symmetrical Paralysis

Causes Problems With:

- Respiration
- Talking
- Swallowing
- Bowel & Bladder Function

CJ-MILLER

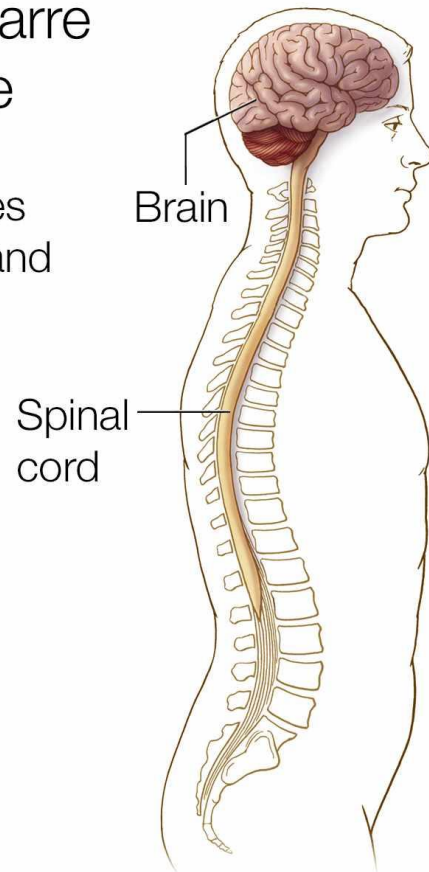
# Pathology

→ **Cell-mediated immunologic reaction**

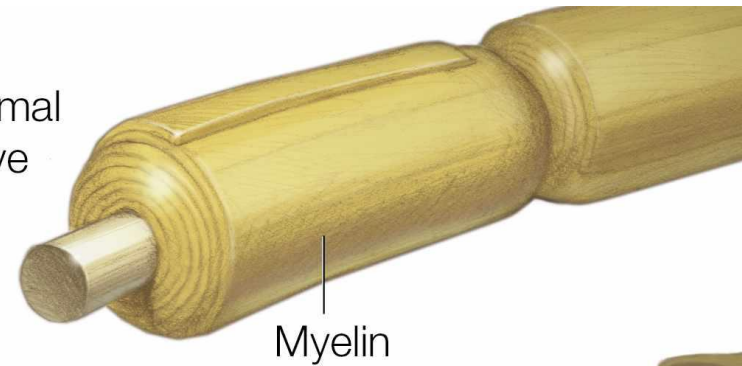
→ **Segmental demyelination**

## Guillain-Barre Syndrome

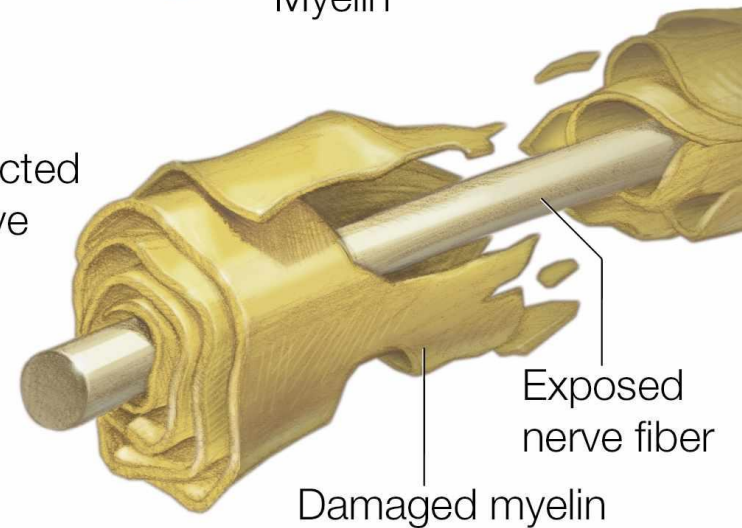
Affects nerves in the brain and spinal cord



Normal nerve



Affected nerve



# Spectrum of Guillain-Barre Syndrome

- Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP)
- Axonal GBS
- Acute Motor Axonal Neuropathy (AMAN)
- Variant
  - Miller-Fisher Syndrome (Ophthalmoplegia)
  - Sensory GBS
  - Subacute Pandysautonomia
  - Acute Vestibulopharyngitis

# Treatment of GBS

- Plasmapheresis
  - 250-300 cc/kg in divided doses
- IV Immunoglobulin (IVIg)
  - 0.4 gm/kg/day for 5 days

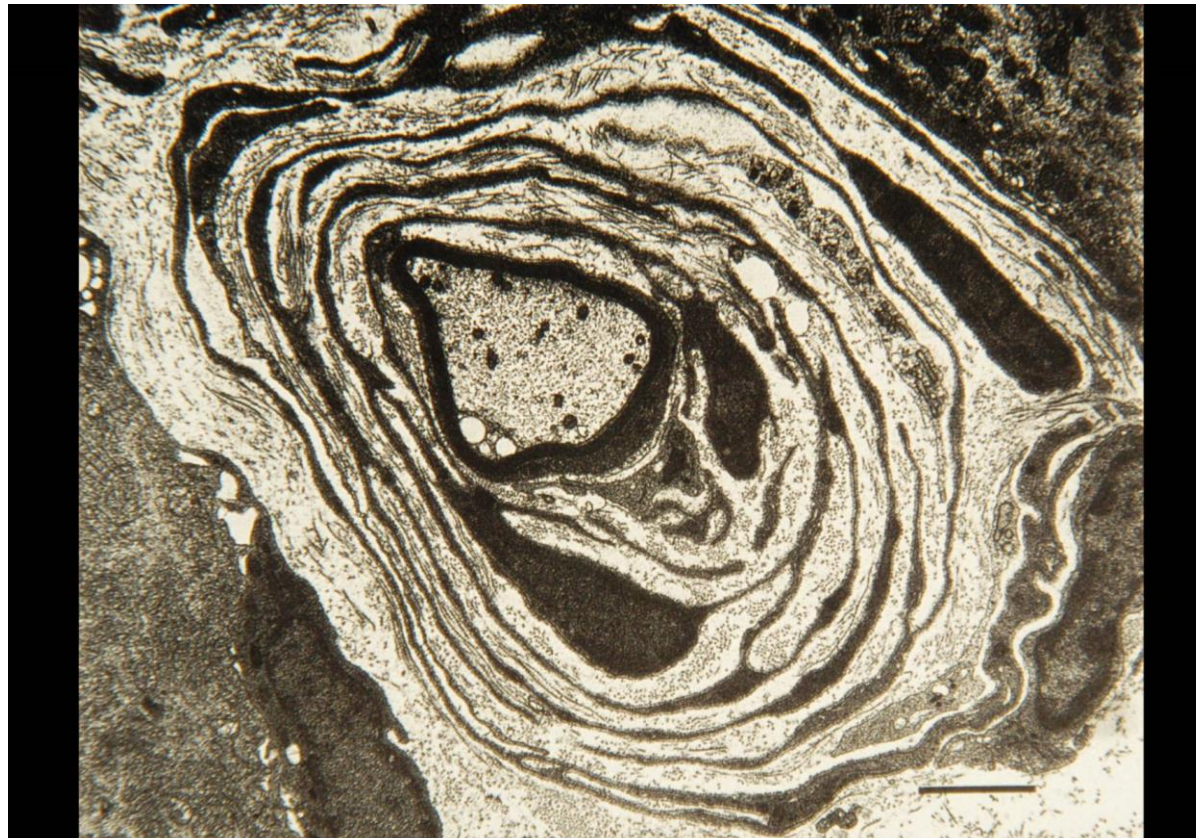
# 醫師國考題

- 42歲男性，沒有糖尿病及慢性器官衰竭病史；15天前曾有2天腹瀉，緊接著雙腳腳底有麻刺感，逐漸轉為麻木，沿著腳底一路麻到膝蓋，也發現兩側小腿的肌肉鬆垮及力量減弱，雙手使用筷子及扭瓶蓋的動作也變困難。下列有關病患的診斷臆測，何者最為正確？
- A.右側中大腦動脈的大面積中風
  - B.遺傳性運動及感覺神經病變
  - C.陳舊性的脊髓白質病變後遺症
  - D.急性的周邊神經病變

Ans:D

# Chronic Inflammatory Demyelinating polyradiculoneuropathy (CIDP)

## Demyelination-Remyelination



**Fig. 3.11.** Onion bulb. A remyelinated axon between the dorsal root ganglion cells is surrounded by layers of Schwann cell and fibroblast processes. Bar = 2  $\mu$ m. (From Adam et al. (1989), with permission.)



# Treatment of CIDP

- Steroid
  - 1 mg/Kg
  - Do not taper too far
- Azathioprine
  - 3 mg/kg
- Cyclosporine
  - 4-8 mg/kg/day
- Plasmapheresis
  - 250–300 cc/kg in divided doses
- IV Immunoglobulin (IVIG)
  - 0.4 gm/kg/day for 5 days
- Cyclophosphamide
  - 500-750 mg/m<sup>2</sup> every month

# DM (Diabetes Mellitus)

DM mononeuropathy

DM Mononeuropathy multiplex

DM polyneuropathy

DM amyotrophy (Muscle atrophy)

## Table 64–1. CLASSIFICATION OF DIABETIC NEUROPATHY

### Symmetric Polyneuropathies

Sensory or sensorimotor polyneuropathy

Autonomic neuropathy

Symmetric proximal lower limb motor neuropathy

### Focal and Multifocal Neuropathies

Cranial neuropathy

Trunk and limb mononeuropathy

Asymmetric lower limb motor neuropathy

### Mixed Forms

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# Charcot-Marie-Tooth disease

## Hereditary motor and sensory neuropathy (HMSN)

Late childhood onset

Symmetric distal motor weakness

Muscle atrophy

Most in peroneal nerves

Pes cavus

Invert champagne bottle sign

Sensory involvement

ANS not involved

# Charcot-Marie-Tooth disease



# Overview

- Myopathic disorder
- Myopathic symptoms
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  - Muscular dystrophies
    - Duchenne dystrophy
    - Becker dystrophy
  - Myotonic dystrophy

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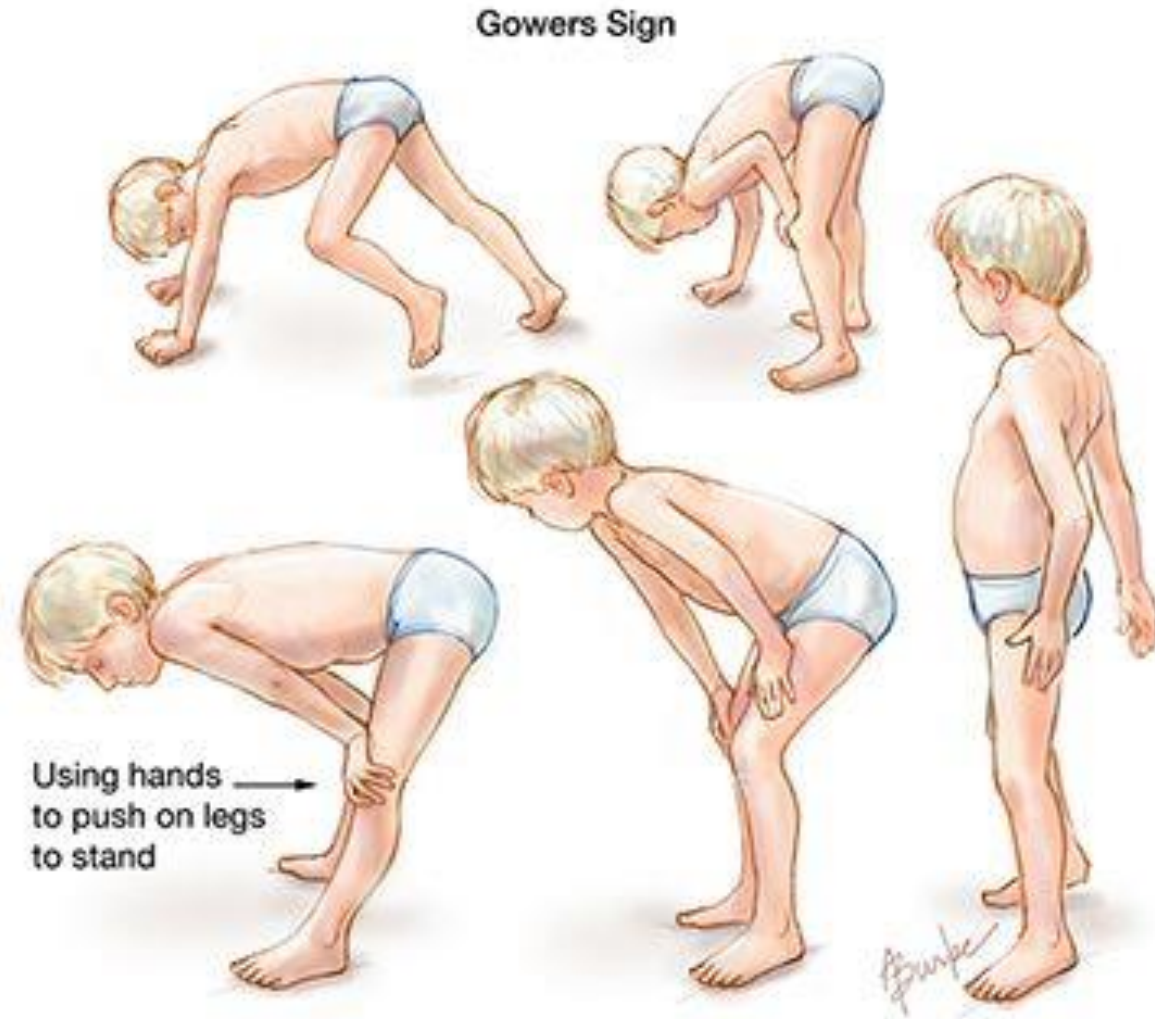
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# Myopathic disorder symptoms

- Dysfunction of muscle fibers
- Progressive proximal muscle weakness (general rule)
- Muscle atrophy
- Inherited or acquired



# Myopathic disorder

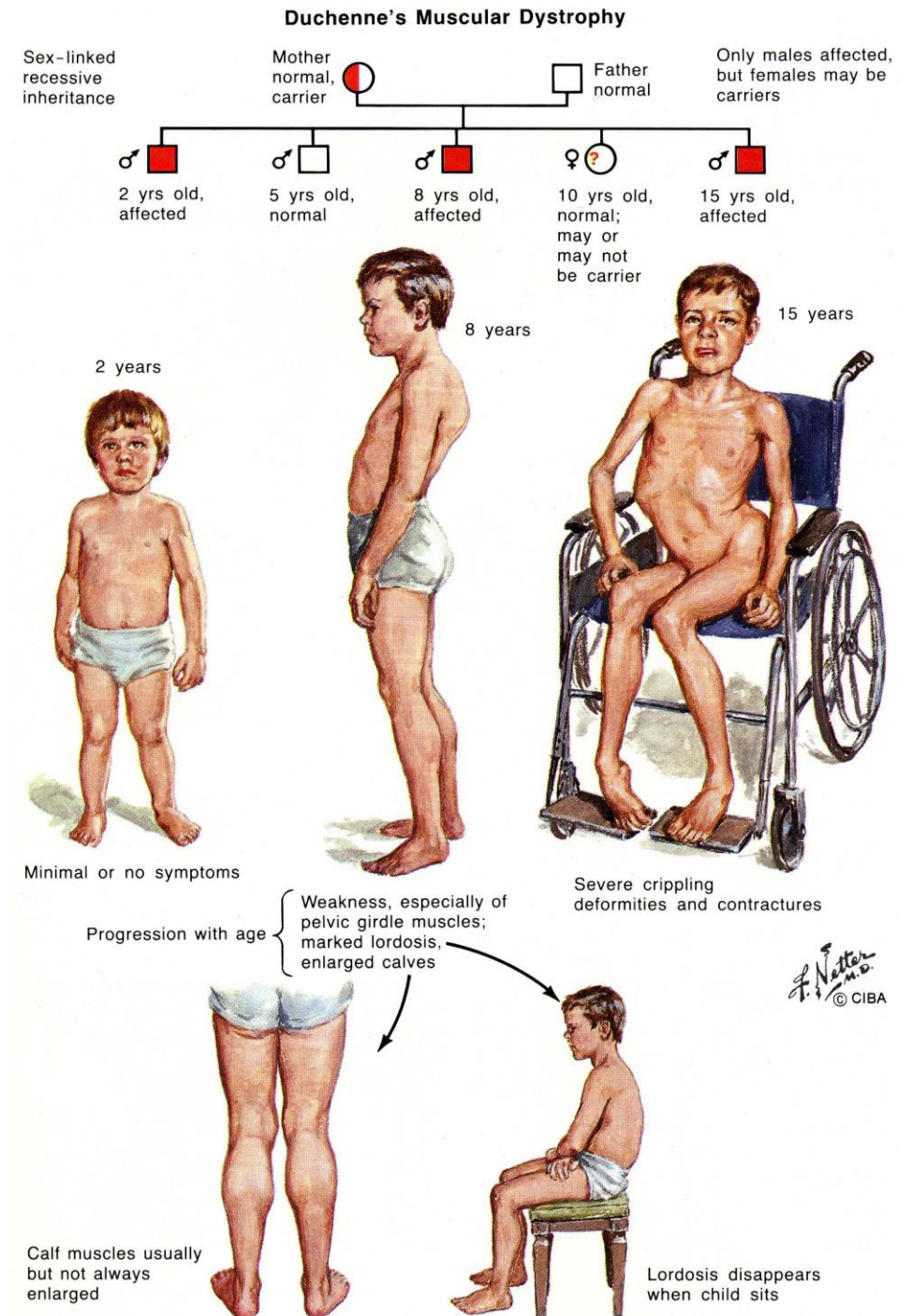


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# Muscular dystrophies

- Dystrophin protein
- Sex-linked inheritance

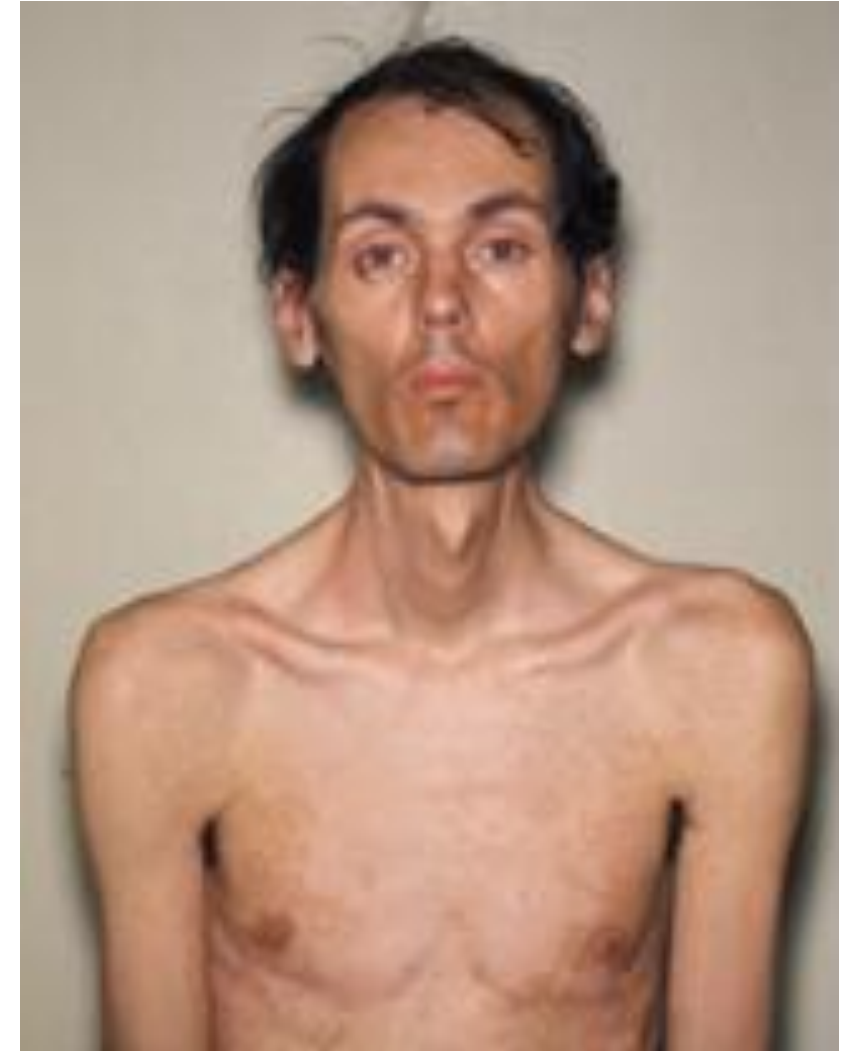


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# Myotonic dystrophy

- Myotonia
- Delayed muscle relaxation
- Dominant inheritance
- Type I myotonic dystrophy → distal muscle weakness, with DM, cataract, arrhythmia, bald
- Type II myotonic dystrophy → proximal muscle weakness, milder symptoms



# 醫師國考題

- 關於肌強直營養不良（ myotonic dystrophy ） ， 下列敘述何者錯誤？
  - A. 自體顯性遺傳
  - B. 以近端無力為主，常見複視
  - C. 經常合併多系統疾病，例如白內障、糖尿病、心律不整等
  - D. 血液中的肌肉酵素一般是正常或輕微上升

A n s : B

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# 參考資料

- Clinical neurology 7<sup>th</sup> edition
- Merritt's neurology 13<sup>th</sup> edition
- 109/110年醫師(二)國考題