SAPIENZA
UNIVERSITÀ DI ROMA
Dipartimento di Neurologia e Psichiatria
Unità di Malattie Neuromuscolari

Diagnostica Neurofisiologica

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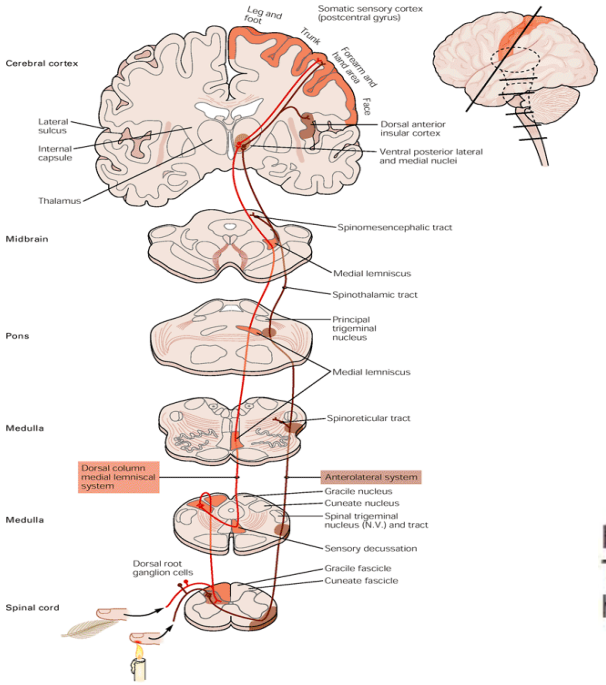
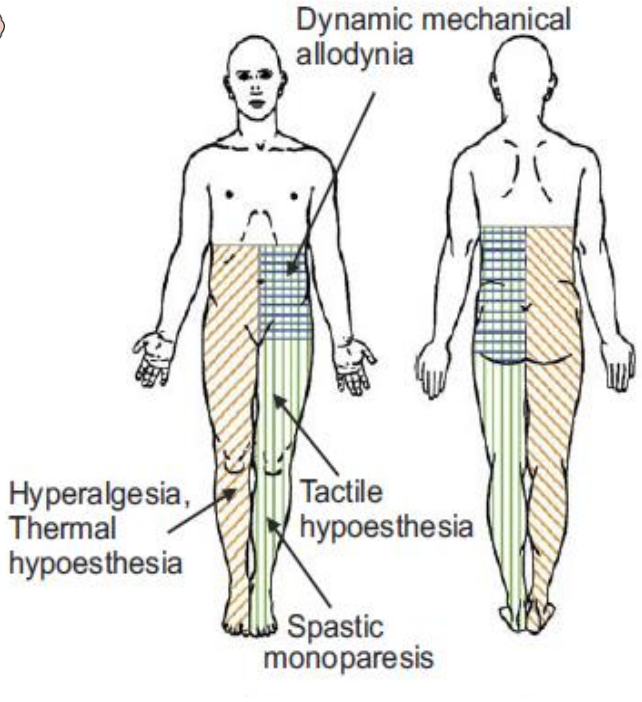
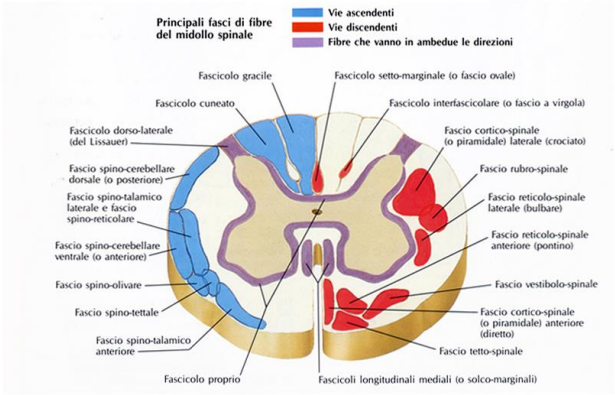
Approccio al paziente neurologico

- Anamnesi
- Distribuzione neuroanatomica
- Esame obiettivo (bedside examination)
- Diagnostica strumentale



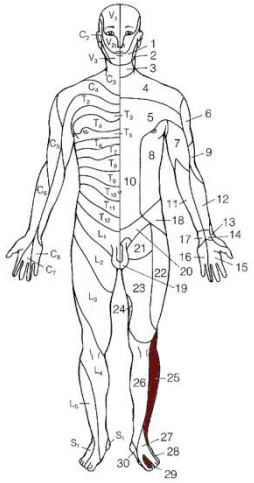
Brown Sequard Syndrome



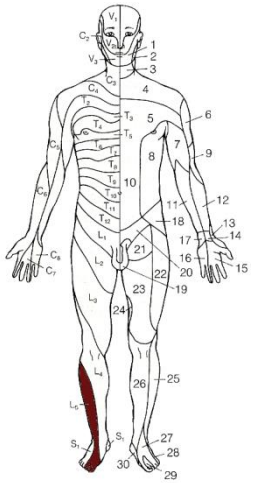


Clinical diagnosis

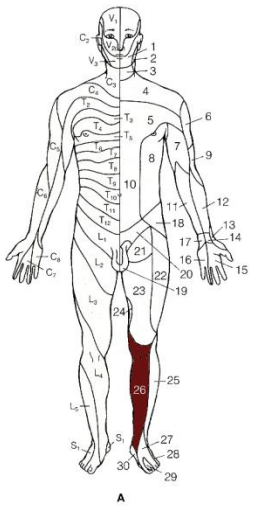
Peroneal mononeuropathy



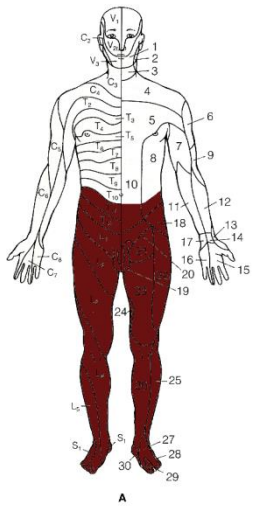
L5- radiculopathy



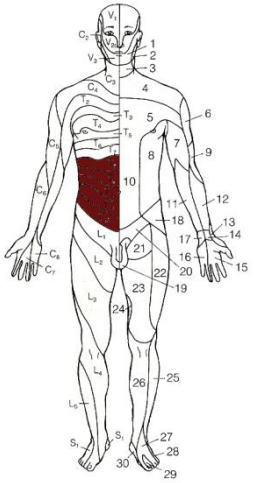
Saphenous mononeuropathy



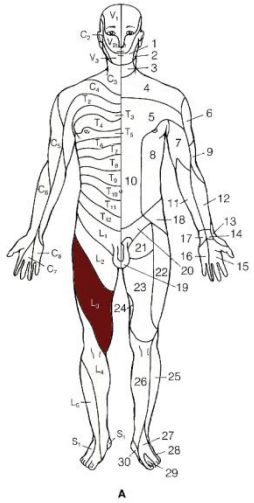
Myelopathy



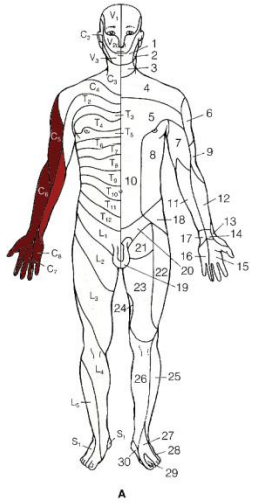
Herpes Zoster



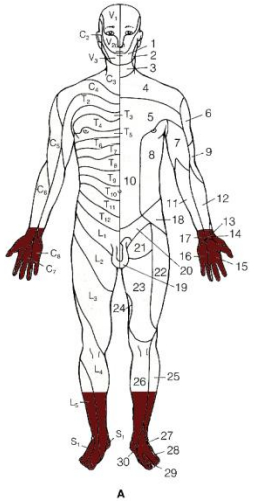
L3- radiculopathy



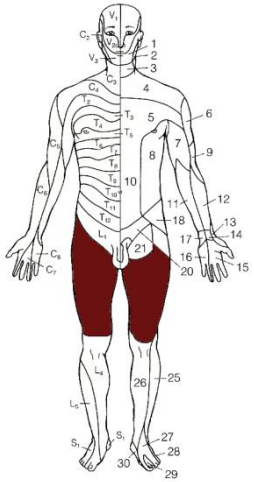
Brachial plexopathy



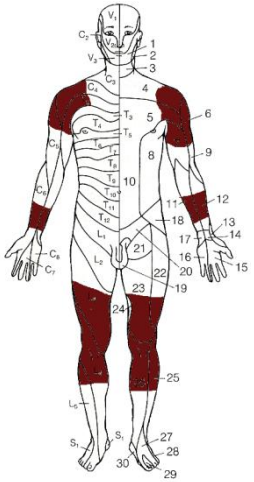
Polyneuropathy



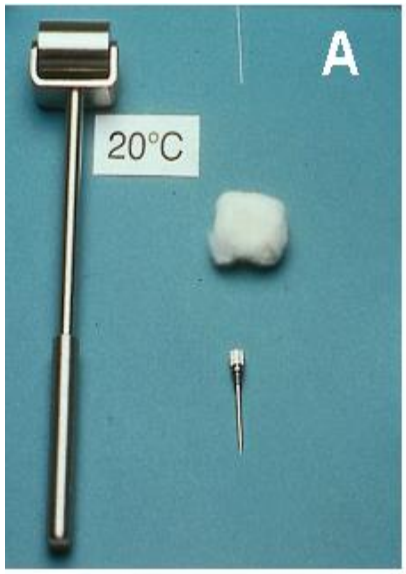
Amiotrophy

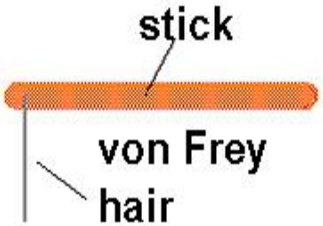


Fibromyalgia



Bedside examination
Quantitative sensory testing





B

Diagnostica delle patologie del sistema nervoso periferico

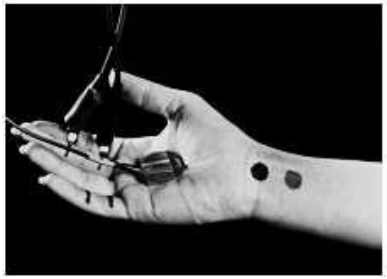
Esami diagnostici:

- ENG/EMG
- Biopsia cute
- Riflessi trigeminali
- Potenziali evocati

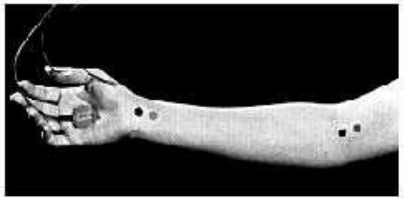
Esame elettroneurografico (ENG)

ENG sensitiva

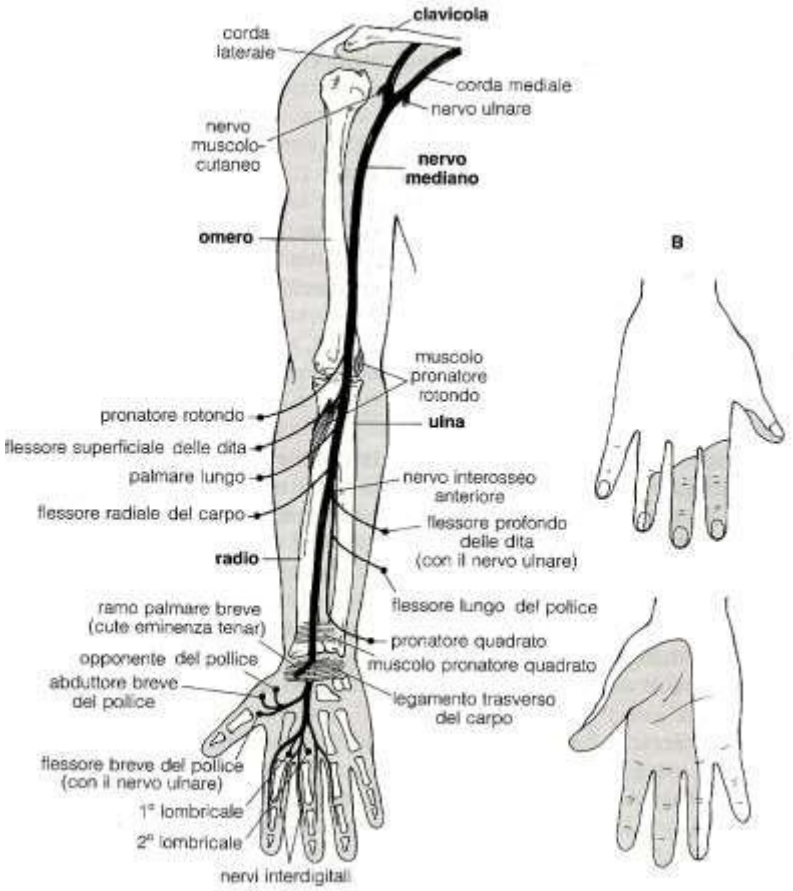
Rec distale



Rec prossimale

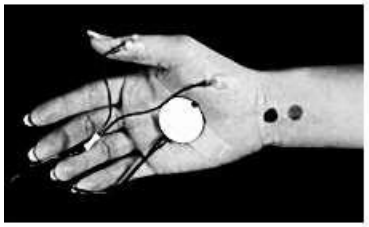


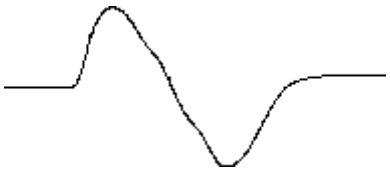
Nervo mediano



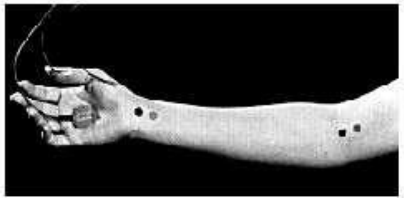
ENG motoria

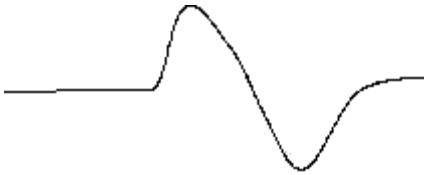
Stim distale





Stim prossimale





Parametri da valutare

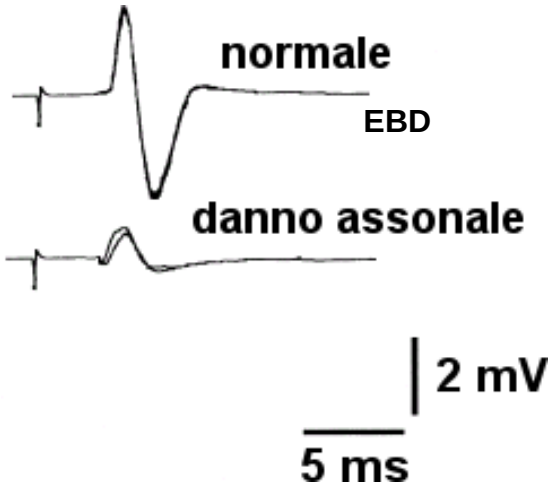
- Presenza/assenza del potenziale
- Ampiezza del potenziale (µV)
- Latenza del potenziale (msec)
- Morfologia del potenziale
- Velocità di conduzione (m/sec)

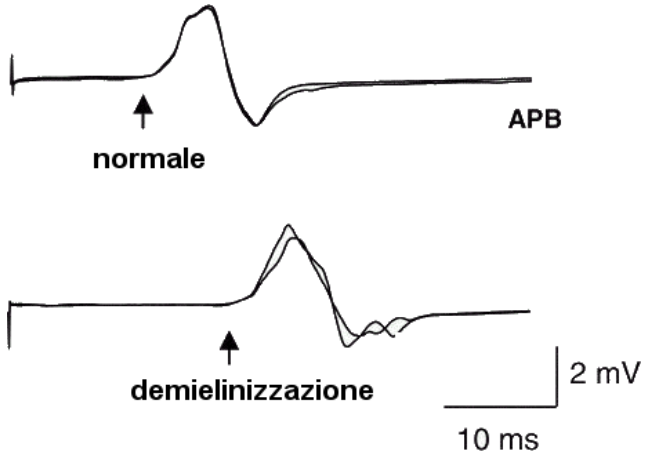
$$\frac{\text{distanza tra i due punti di stimolazione (mm)}}{\text{tempo di conduzione tra i due punti (ms)}}$$

- Distribuzione del danno

Classificazione neurofisiologica patologie del nervo periferico (neuropatie)

-**Assonali** (diabetica, alcolica, tossico-carenziale, genetica)
-**Demielinizzanti** (Neuropatie infiammatorie, genetica)





Classificazione delle neuropatie

-Sensitive

-Motorie

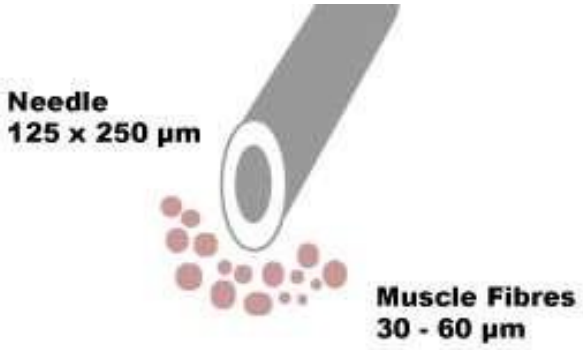
-Sensitivo-motorie

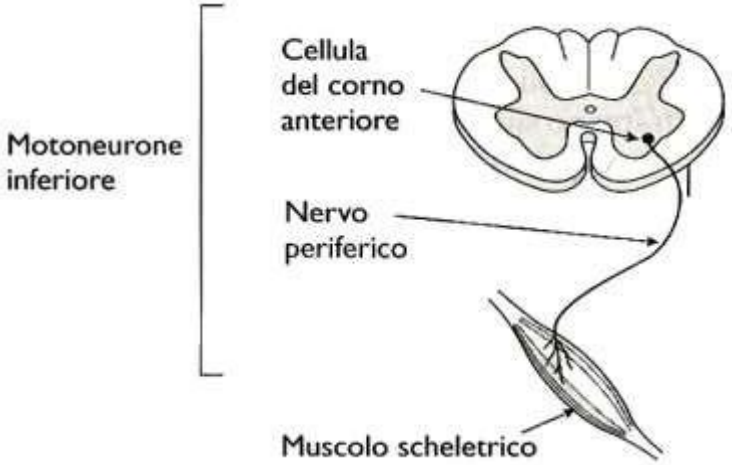
-Mononeuropatie (trauma, infiltrazione)

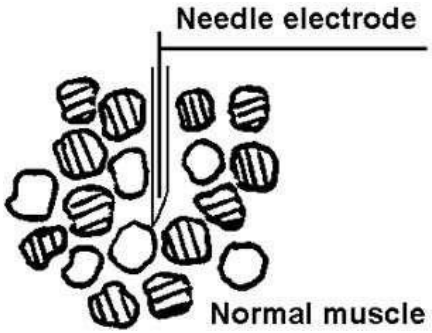
-Multineuropatie (ischemia)

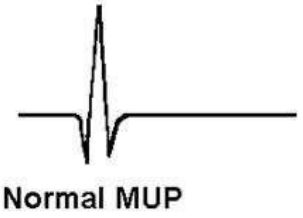
-Polineuropatie (tossiche, dismetaboliche)

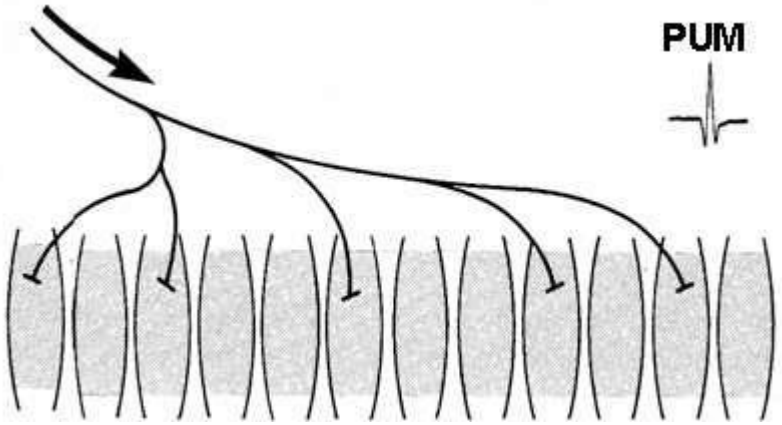
Elettromiografia





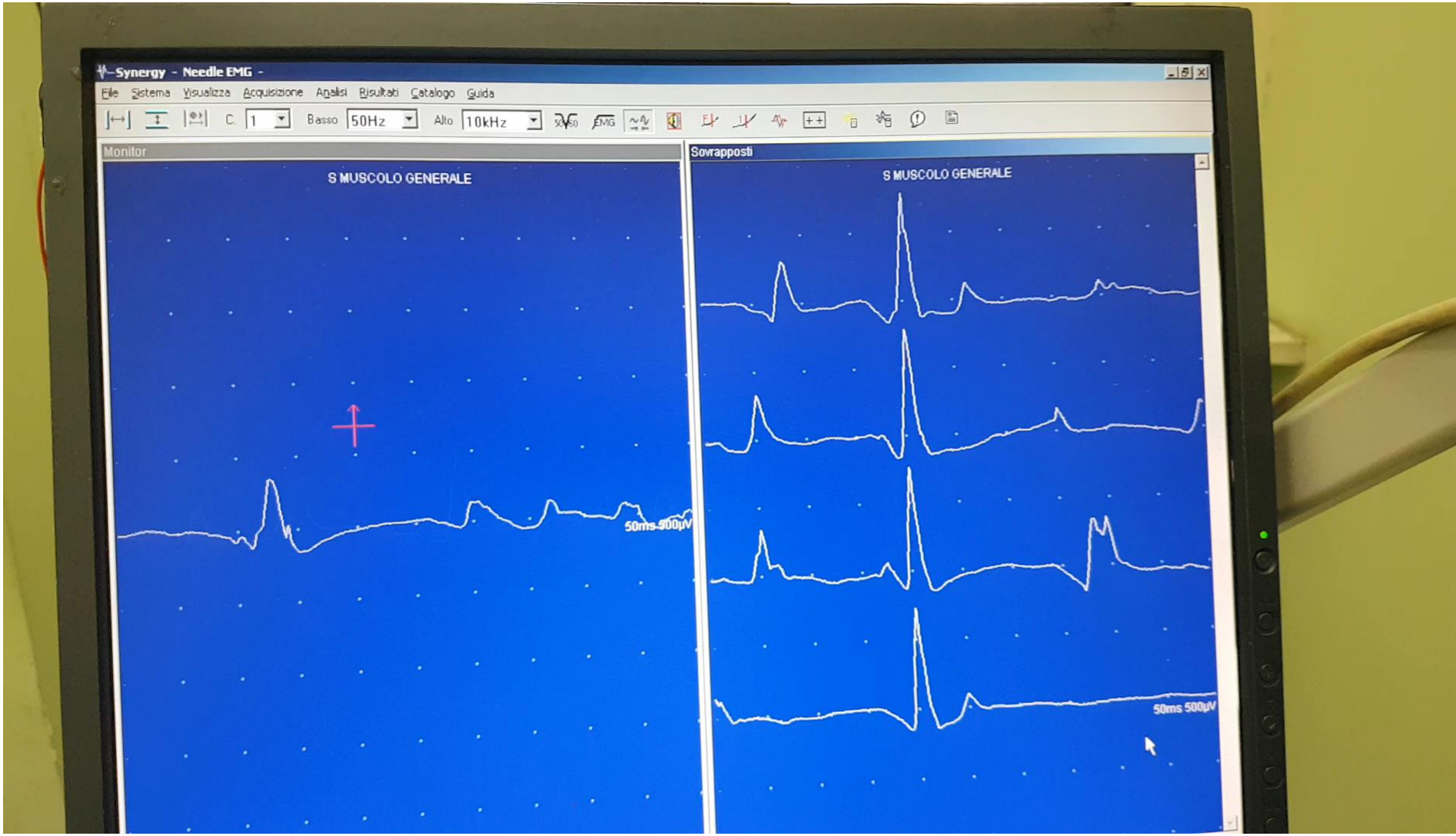




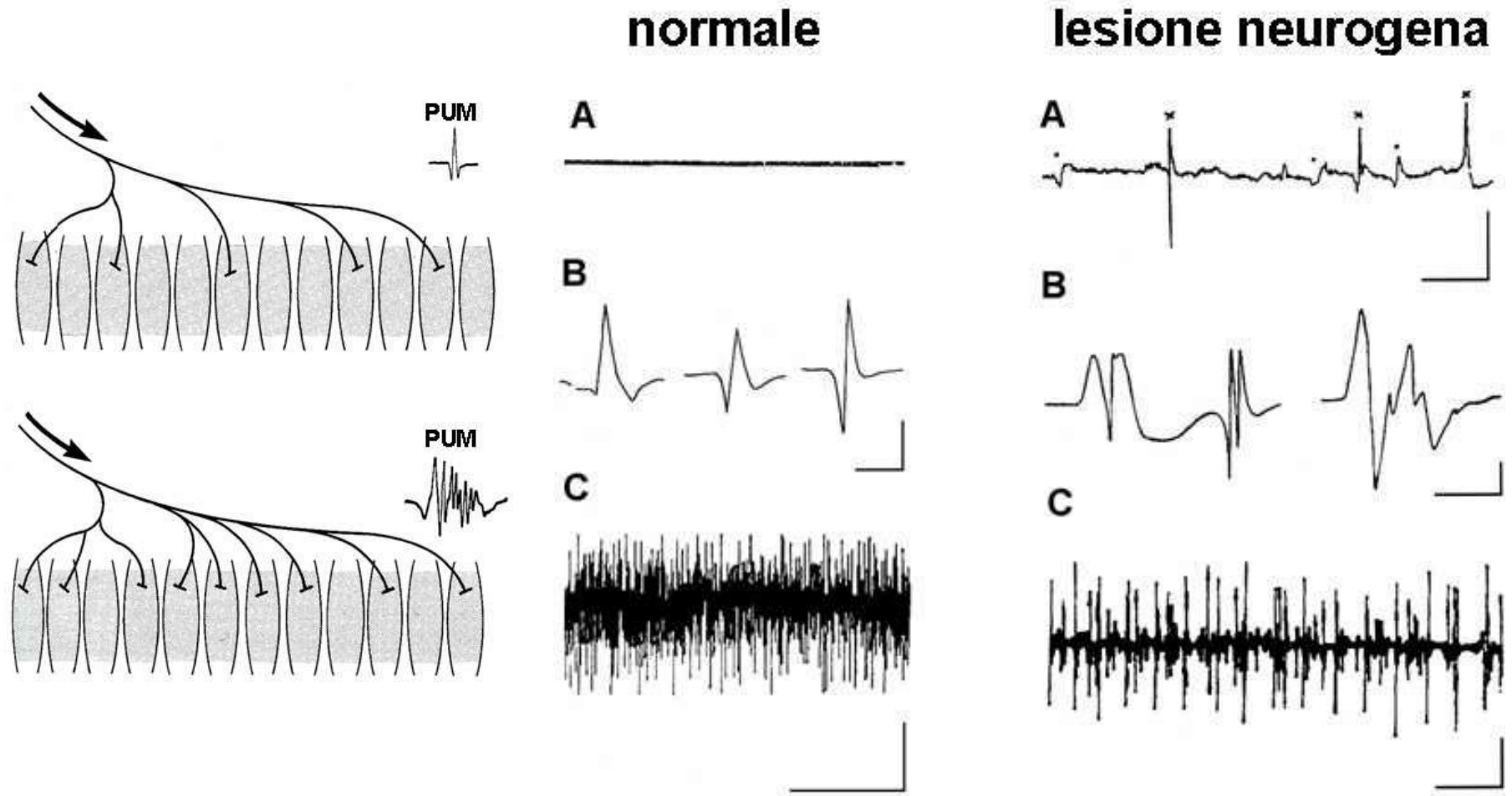


Parametri da valutare

- Presenza/Assenza attività a riposo (denervazione)
- Ampiezza dei PUM (mV)
- Morfologia dei PUM
- Durata dei PUM (msec)
- Reclutamento


Synergy - Needle EMG -
File Sistema Visualizza Acquisizione Analisi Risultati Catalogo Guida
C. 1 Basso 50Hz Alto 10kHz
Monitor
S MUSCOLO GENERALE
50ms 500µV
Sovrapposti
S MUSCOLO GENERALE
50ms 500µV

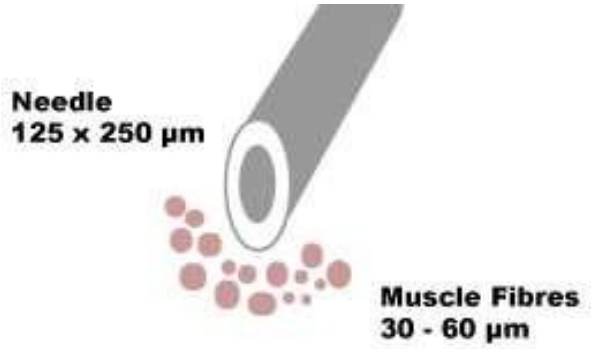
EMG nelle lesioni nervose

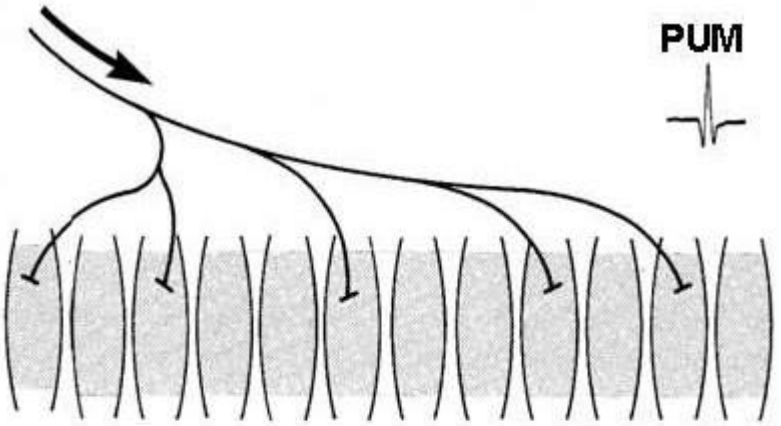


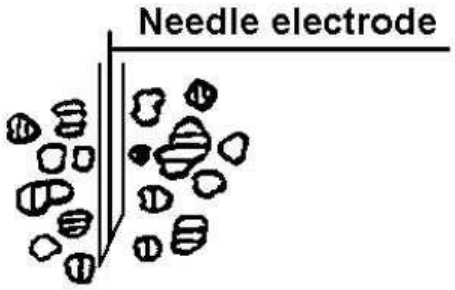
I segni di danno neurogeno compaiono nelle lesioni del contingente motorio periferico:

- malattie del nervo (radice spinale, nervo periferico).
- malattie del II neurone di moto.

EMG nelle miopatie

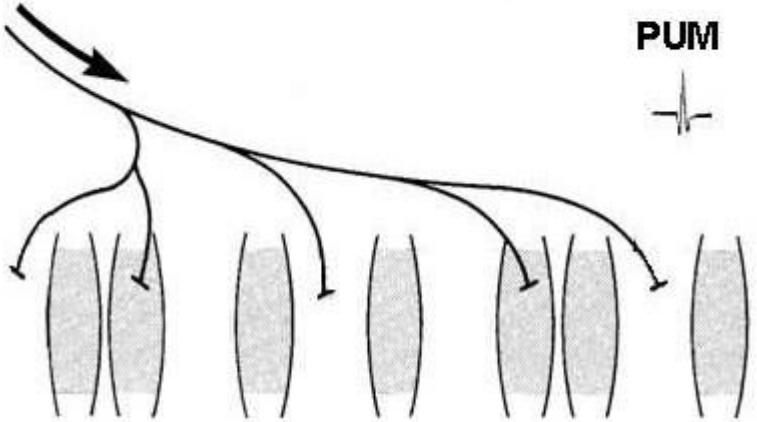




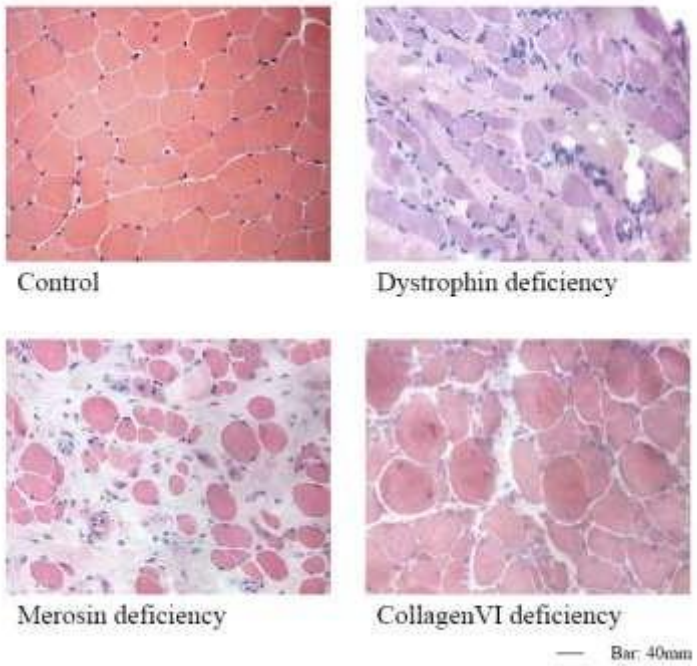


Myopathic changes

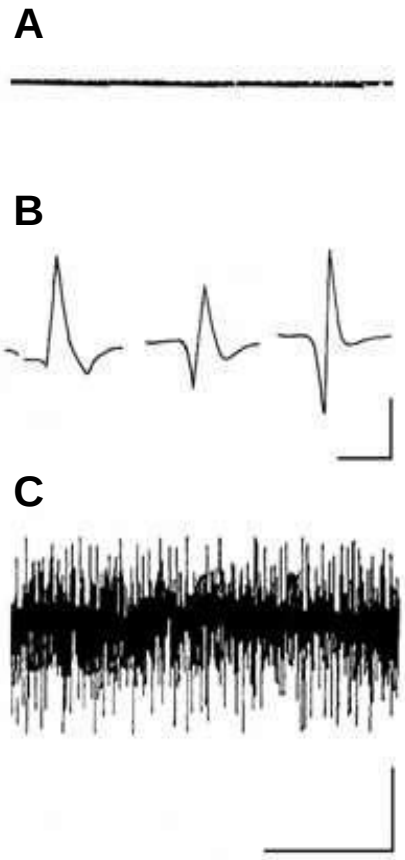
Small MUP



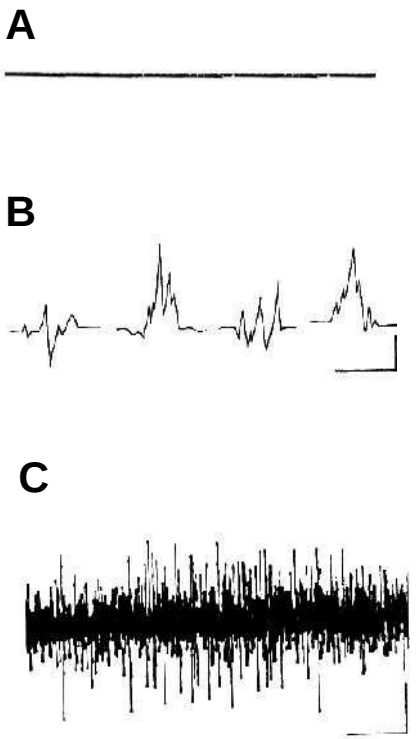
Muscle Dystrophies -Muscle biopsy



Normale



Miopatia



ENG: utile nelle **neuropatie**. Permette di differenziare le neuropatie assonali dalla demielinizzanti, con importanti implicazioni terapeutiche e prognostiche.

Nelle sezione del nervo: assenza dei potenziali potenziali sensitivi e motori.
Nella neuropatia da intrappolamento: rallentamento della velocità di conduzione attraverso la zona compressa.
Nelle polineuropatie assonali: riduzione di ampiezza dei potenziali sens e mot
Nelle neuropatie demielinizzanti: rallentamento della velocità di conduzione.

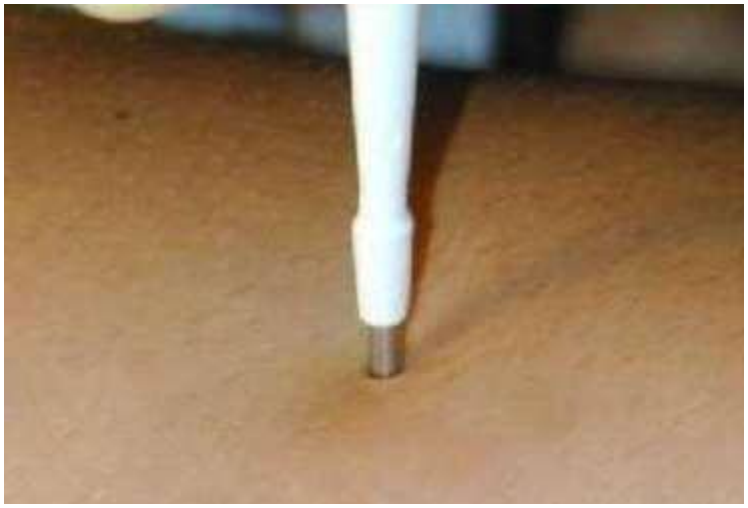
Non indicata nei **danni** del **II neurone di moto** o nei danni **radicolari**.

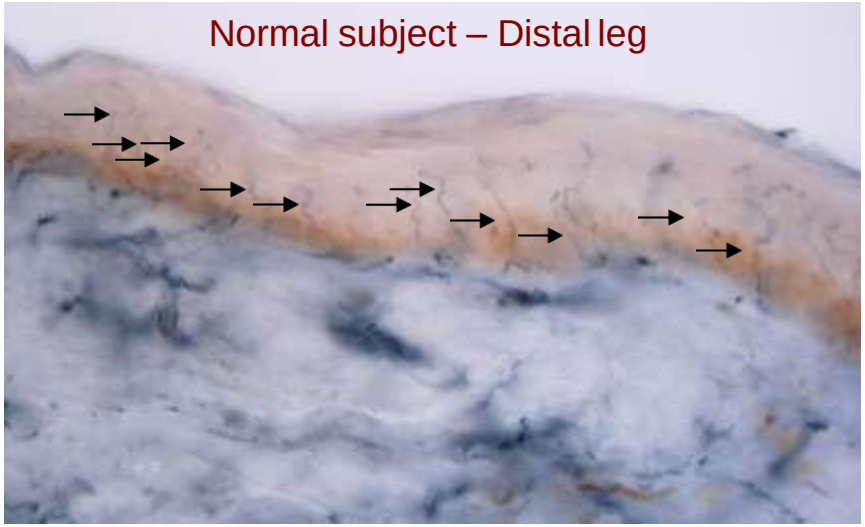
EMG: esame invasivo. Necessita della collaborazione del paziente. Utile nella valutazione dei **danni** a carico del **contingente motorio periferico**, **dal II neurone di moto al nervo periferico**. Esame essenziale nella valutazione delle **miopatie.**

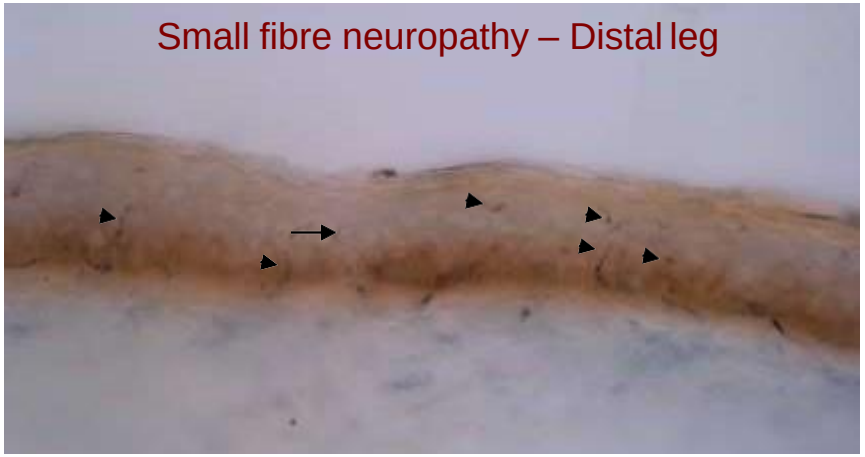
Non indicata nelle **polineuropatie.**

*Sia l'ENG, sia l'EMG **non** sono indicati nei disturbi sensitivi e motori causati da lesione del sistema nervoso centrale.*

Skin biopsy



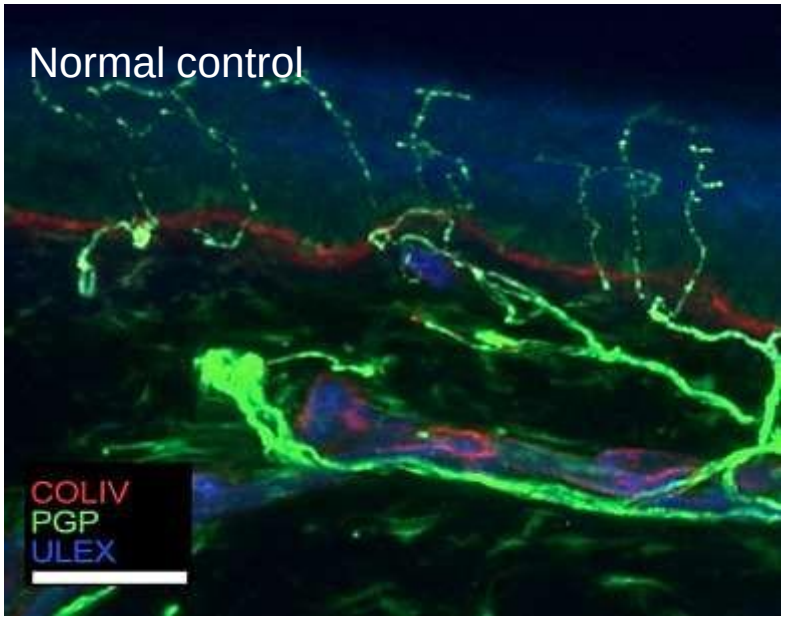


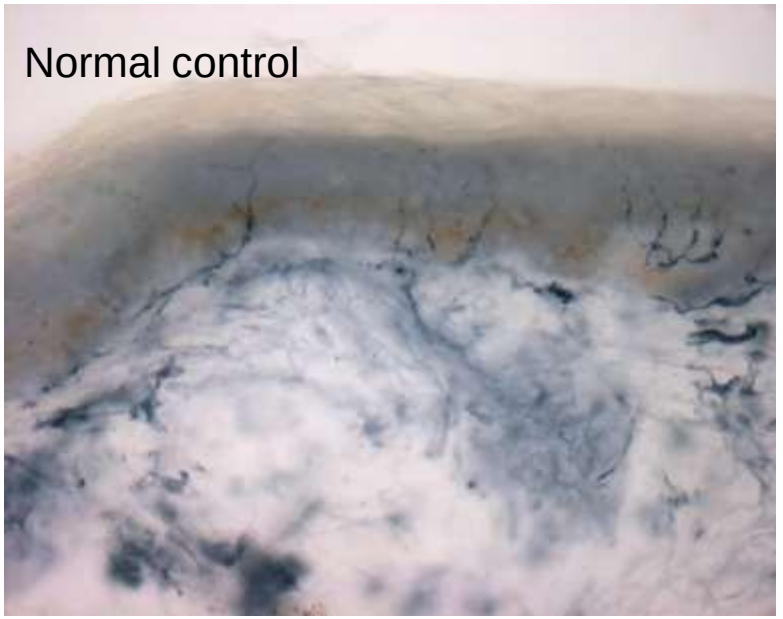


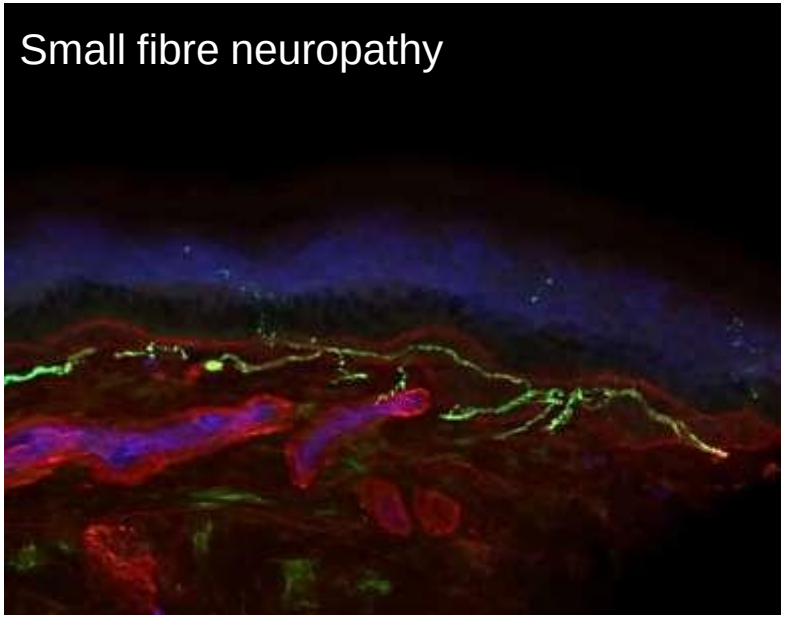
→ Normal nerve fibres

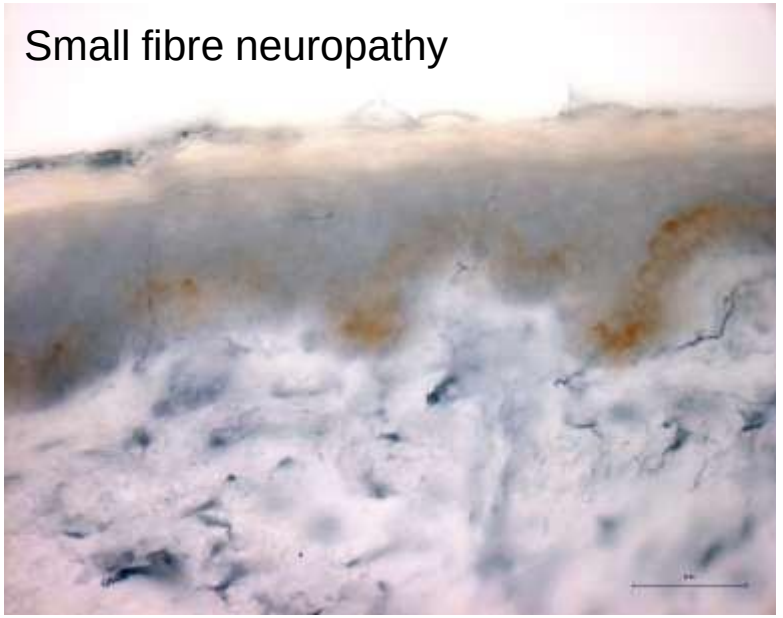
▸ Axonal abnormalities

Comparative morphometry

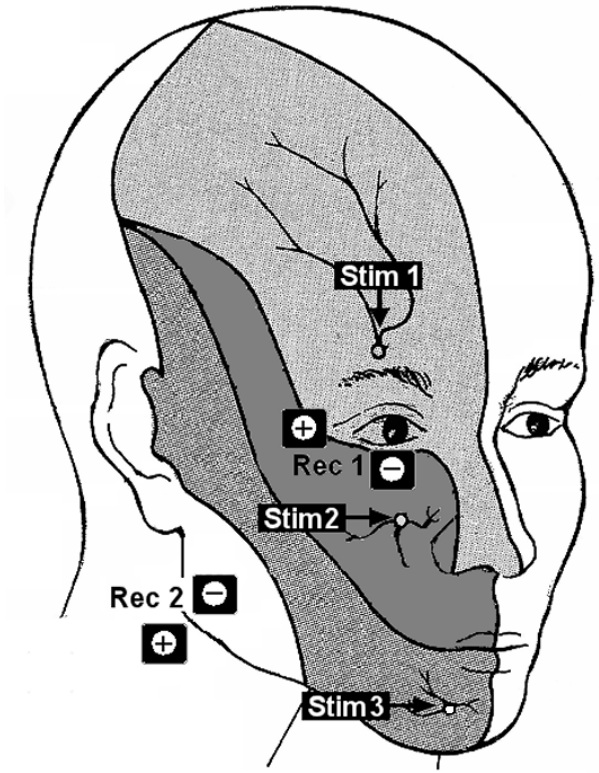


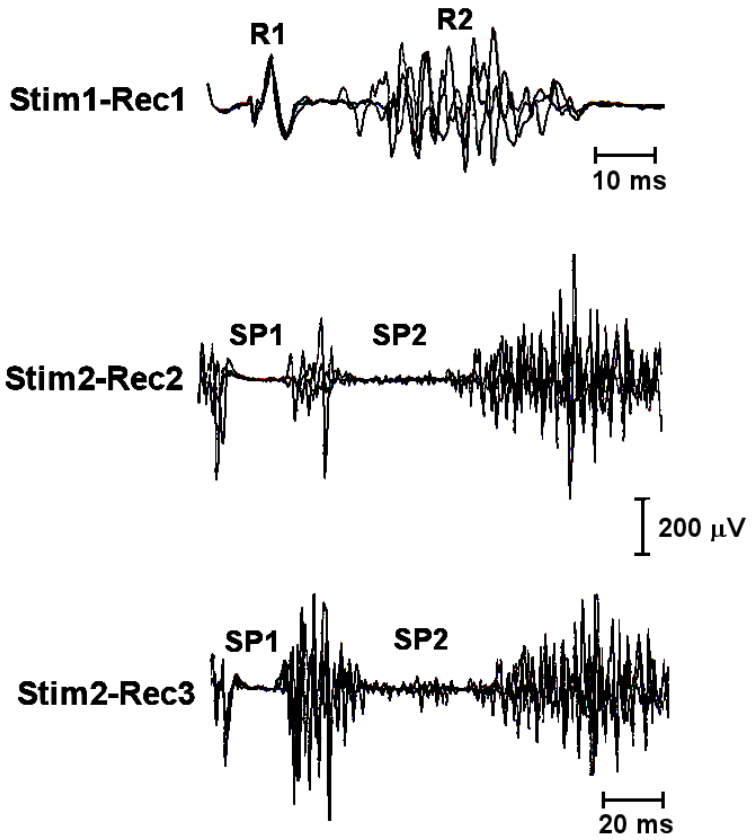






Trigeminal reflexes





Blink reflex: anatomy

R1: short **oligosynaptic** circuit, **homotopycal**, Strictly **segmental**

The whole circuit lies in the **pons**

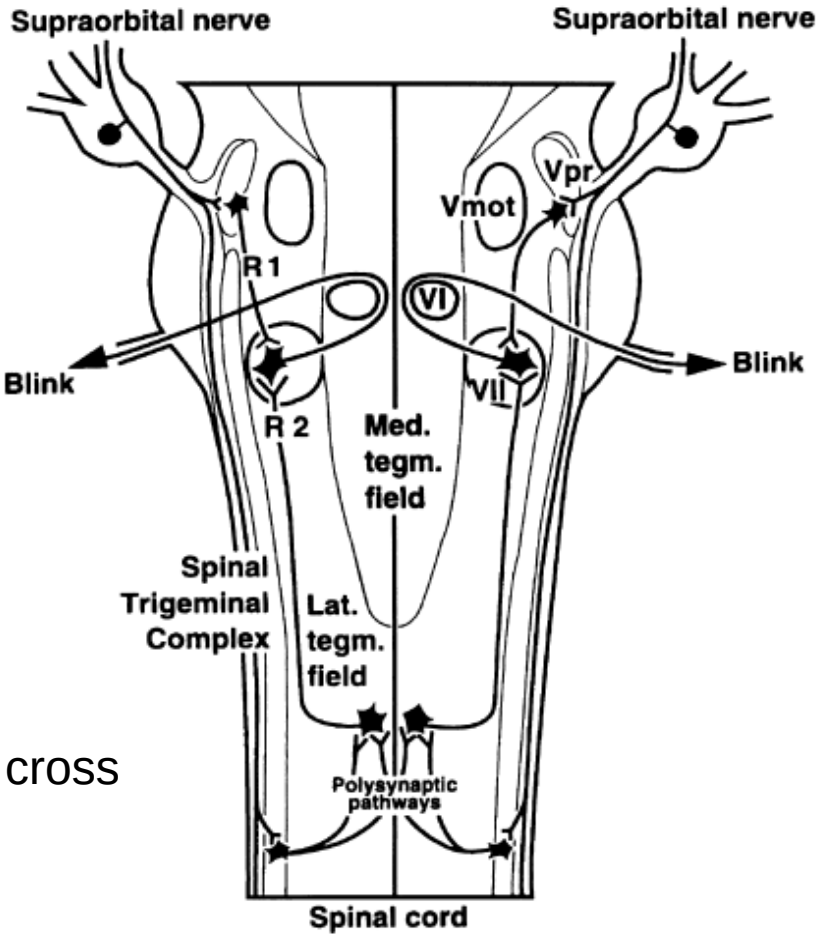
Trigeminal afferent impulses conducted by Aβ fibers

From 1 to 3 interneurons to the **facial motorneurons**

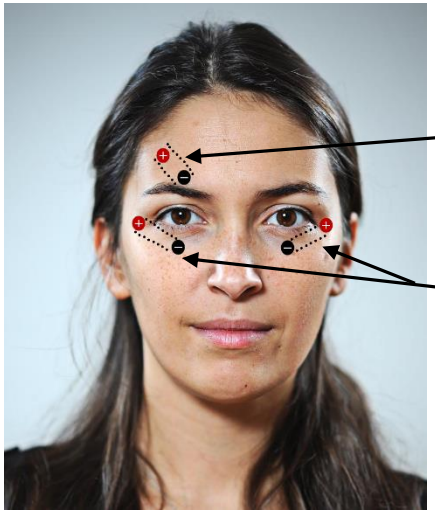
R2: **polysynaptic** circuit, ascending **ipsilateral** and **contralateral**

The circuit reach the **medulla oblongata** and cross the midline

To the **facial motorneurons**



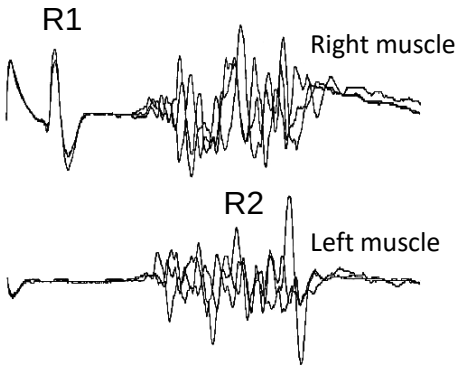
Blink reflex



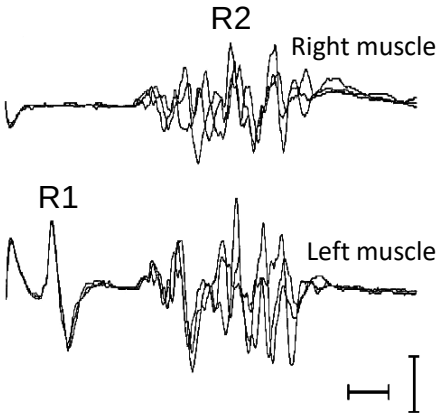
Supraorbital nerve stimulation
(0.1 ms, 15-35 mA)

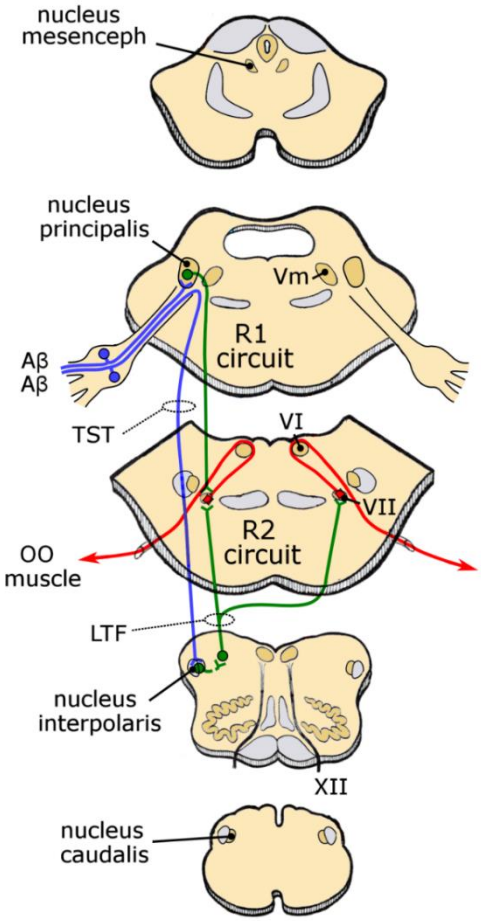
Orbicularis oculi recording
(4 non-averaged trials)

Right simulation

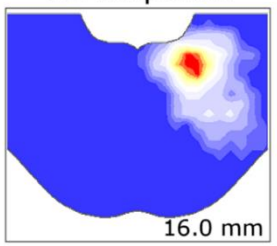


Left simulation

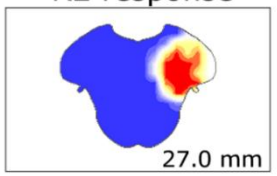




R1 response



R2 response



Blink reflex

R1 **stable response**, **resistant** to all suprasegmental influences, including supratentorial lesions, disorders of consciousness, and cognitive factors.

→ **Reliable** in disclosing **peripheral lesions**

R2 is relatively **unstable**, **habituates** rapidly to repetitive rhythmic stimulations, and is strongly **modulated by suprasegmental influences**, cortical and basal ganglia dysfunction, disorders of consciousness, and cognitive factors

→ Allows **differentiation between** damage to the **afferent** (trigeminal) and **efferent** (facial) arcs of the reflex

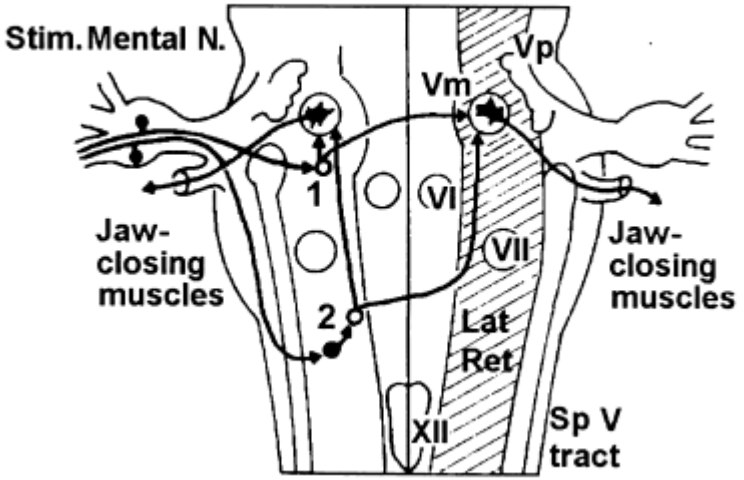
Masseter inhibitory reflex: anatomy

SP1: **short-latency** response, mediated by 1 **inhibitory interneuron**

The whole circuit lies in the **mid**-**pons**

Trigeminal afferent impulses conducted by Aβ fibers

Projections onto the **trigeminal jaw closing motoneurons bilaterally**

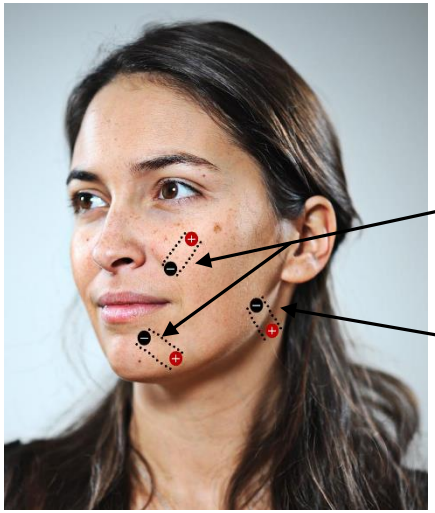


SP2: **polysynaptic** circuit, ascending **ipsilateral** and **contralateral**

The circuit is a chain of excitatory interneurons in the medullary lateral reticular formation, tha last inhibitory

To the **trigeminal motoneurons**

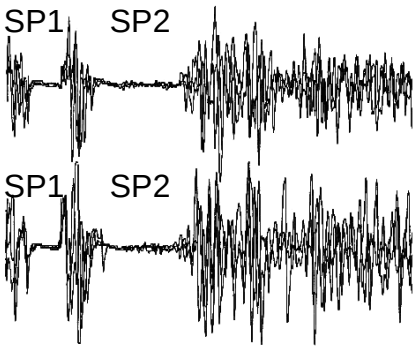
Masseter inhibitory reflex



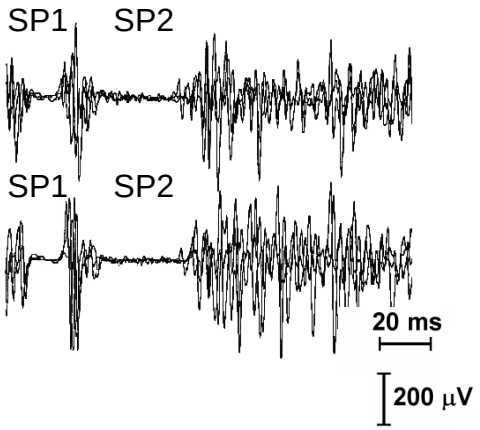
Infraorbital OR mental nerve stimulation
(0.1 ms, 15-45 mA during clenching)

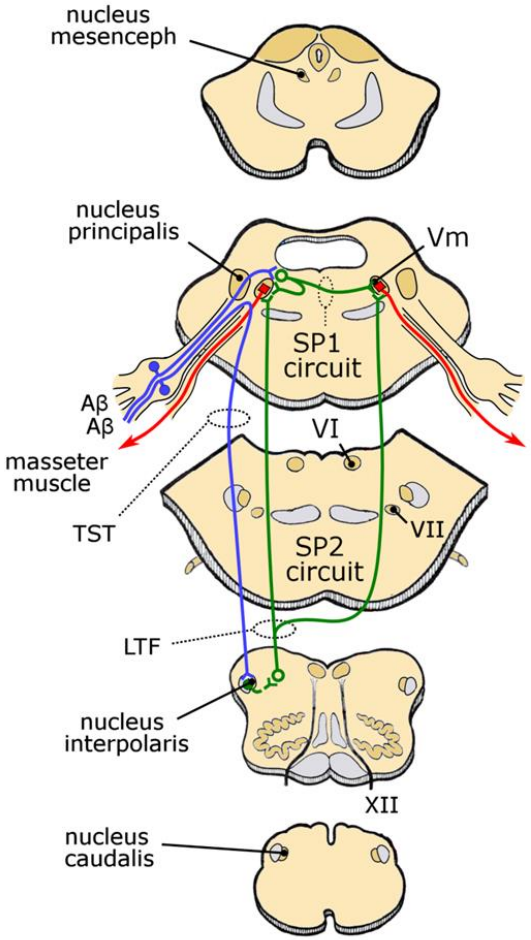
Masseter muscle recording,
7 non averaged trials

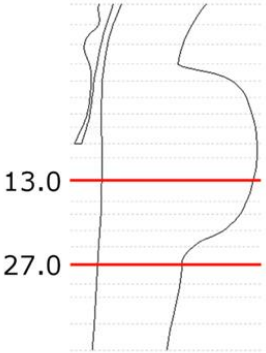
Right simulation



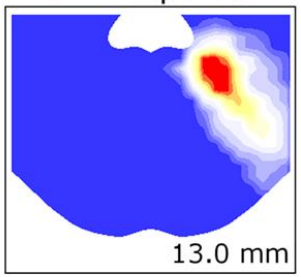
Left simulation



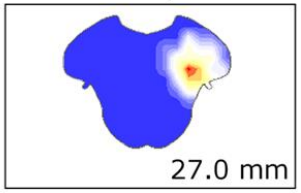




SP1 response



SP2 response



Masseter inhibitory reflex

As in blink reflex studies, **the pattern of abnormality** of the masseter inhibitory reflex provides **important information on the site of the lesion**

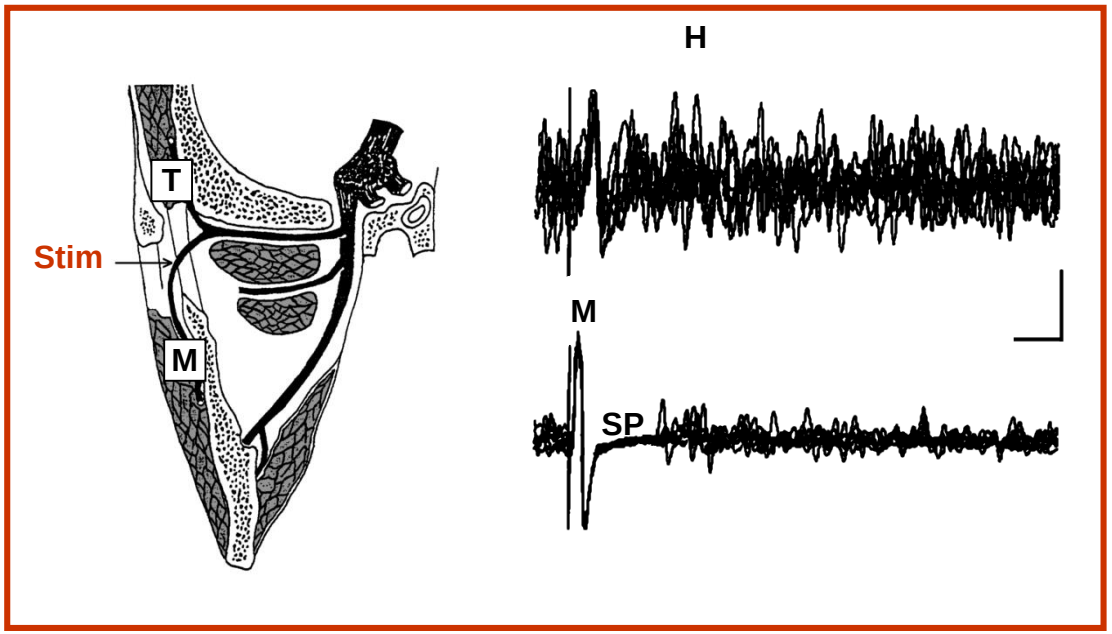
Typical abnormalities of brainstem reflexes and their diagnostic indications

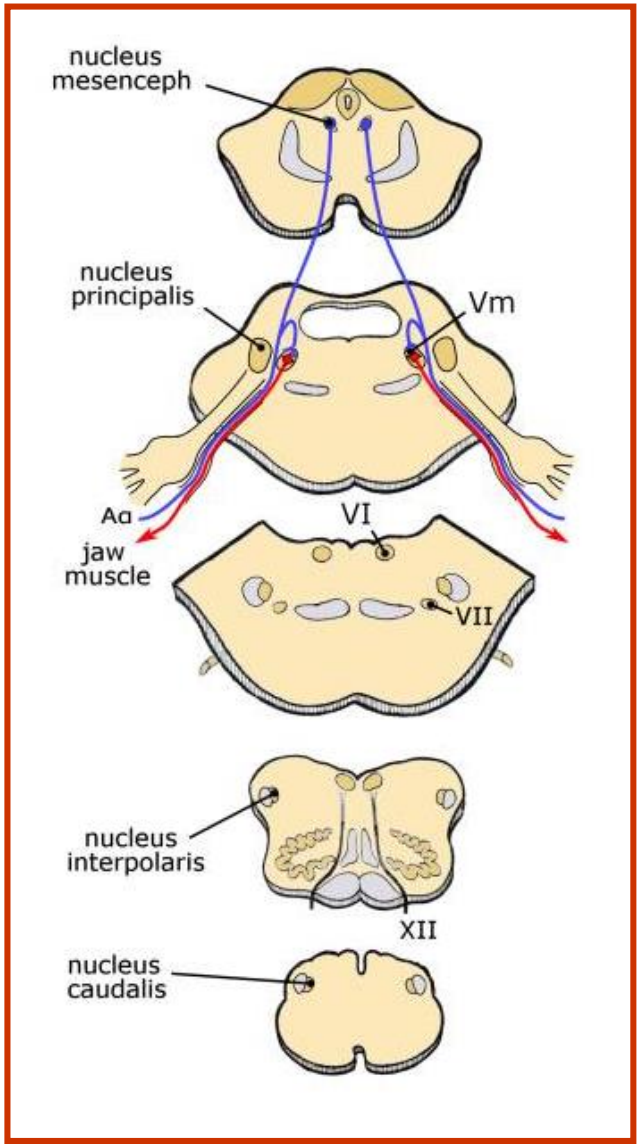
Lesion/disease	Abnormal responses to ipsilateral stimulation[a]	Abnormal responses to contralateral stimulation[a]
1. Facial neuropathy	R1 and d-R2	c-R2
2. Focal trigeminal neuropathy (distal)	Either R1 or SP1 or JJ (R2 or SP2)	Normal
3. Focal trigeminal neuropathy (retrogasserian and root entry zone)	R1 and SP1 and JJ (R2 and SP2)	Normal
4. Sensory polyneuropathy	R1 and SP1 and JJ (R2 and SP2)	R1 and SP1 and JJ (R2 and SP2)
5. Sensory-motor polyneuropathy	All abnormal	All abnormal
6. Ganglionopathy	R1 and SP1 (R2 and SP2)	R1 and SP1 (R2 and SP2)
7. Dorsal midbrain lesion	JJ	Normal
8. Dorsal pontine lesion	All can be abnormal according to the precise site of lesion	Normal
9. Midline medullary lesion	c-R2 and c-SP2	c-R2 and c-SP2
10. Lateral medullary lesion	d- and c-R2; d- and c-SP2	Normal
11. Suprasegmental (pyramidal)	Normal	d- and c-R2 (d- and c-SP2)
12. Suprasegmental (extrapyramidal)	Enhanced excitability of R2 and SP2	Enhanced excitability of R2 and SP2

[a] R1, early blink reflex; d- and c-R2, direct and crossed late blink reflex; SP1 and SP2, early and late masseter inhibitory reflex; JJ, jaw jerk. Responses in parentheses are less frequently affected.

G. Cruccu, G. Deuschl / Clinical Neurophysiology 111 (2000) 371±387

Temporalis H Reflex

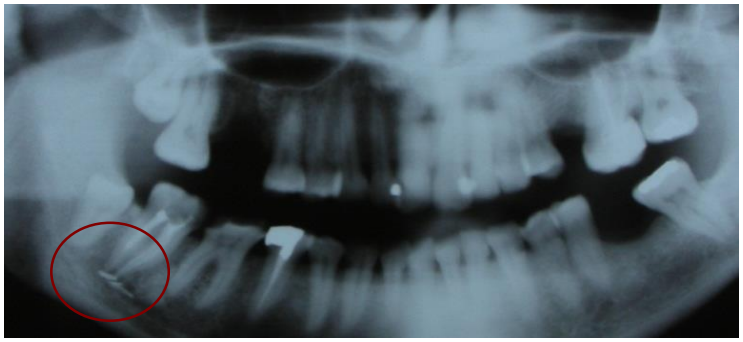


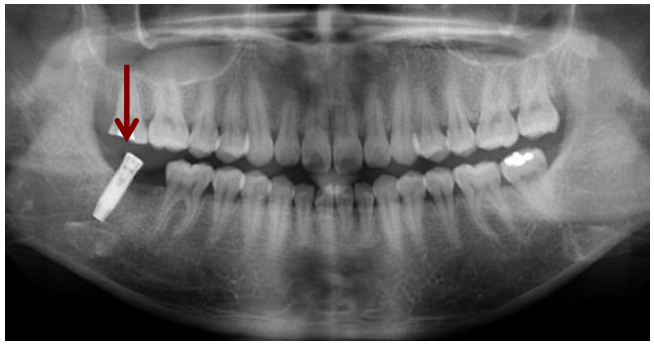


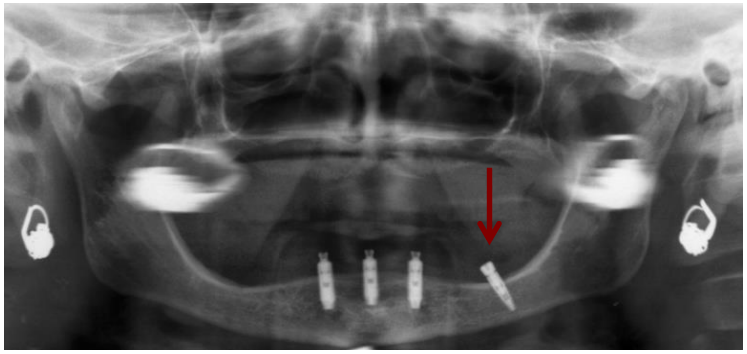
Stimulation:
masseteric nerve in the infratemporal fossa
(monopolar needle electrode; 0.1 ms, 3-5 mA)

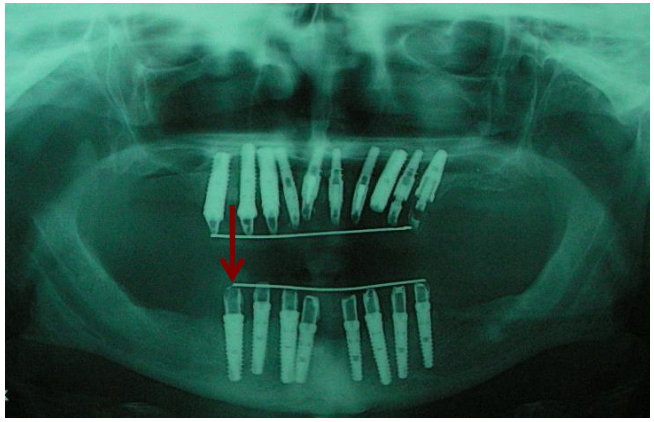
Recording:
8 non-rectified trials,
during clenching.
Latency 4.5 ms

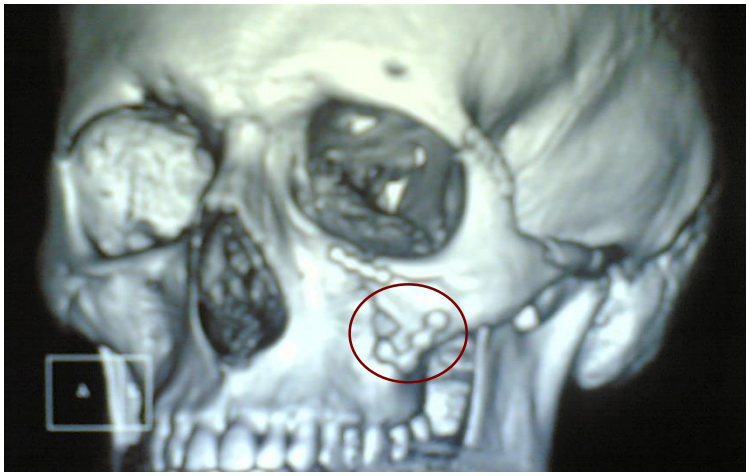
Focal trigeminal neuropathy



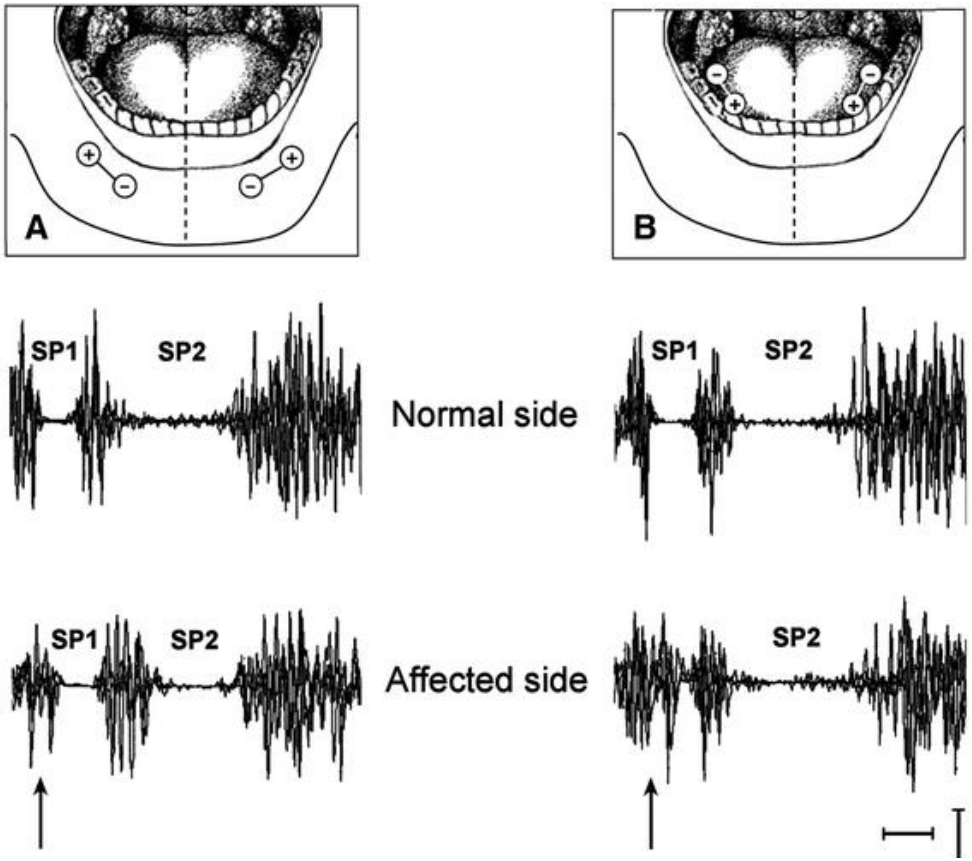








Diagnostic accuracy in iatrogenic damage



Concerning the overall diagnostic accuracy, the MIR was abnormal in 90 nerves and normal in 86 on the affected side whereas on the contralateral side it was abnormal in 2 and normal in 174, which resulted in a strong association ($P < 0.0001$; Fisher's exact test), with 51% sensitivity (CI: 0.435–0.587) and 99% specificity (CI: 0.959–0.999).

Table 1

Latency of the masseter inhibitory reflex (mean ± SD) in 160 patients with iatrogenic damage to the mandibular nerves

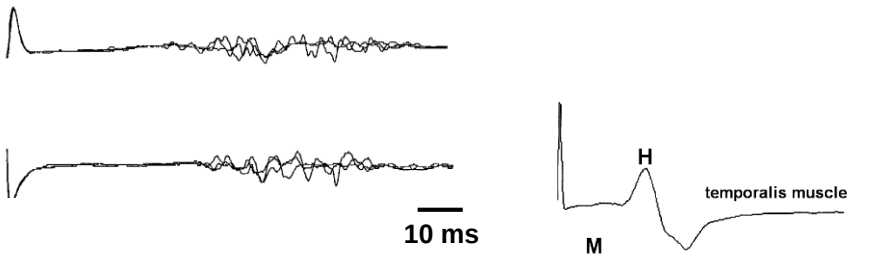
Stimulation site	Affected	Contralateral	*P**
Inferior alveolar nerve (n = 125)	12.0 ± 1.3	11.3 ± 0.8	<0.0001
Lingual nerve (n = 51)	14.3 ± 2.6	12.9 ± 1.7	<0.0001

*P** *t* test with Welch's correction

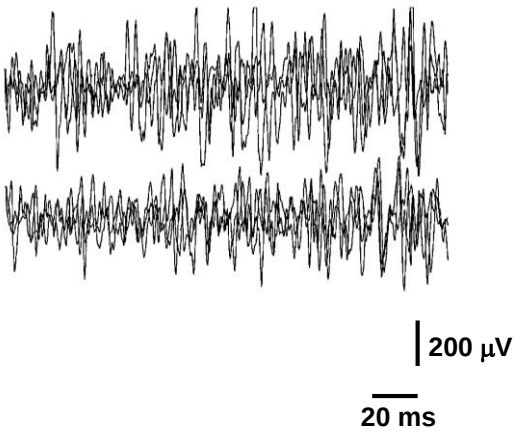
FOSMN Syndrome

Patient

Blink reflex

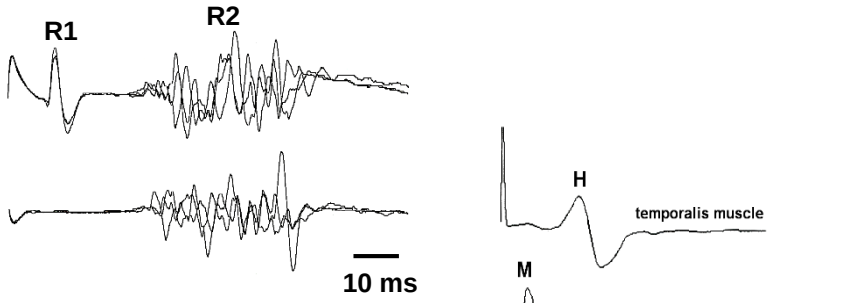


Masseter inhibitory reflex

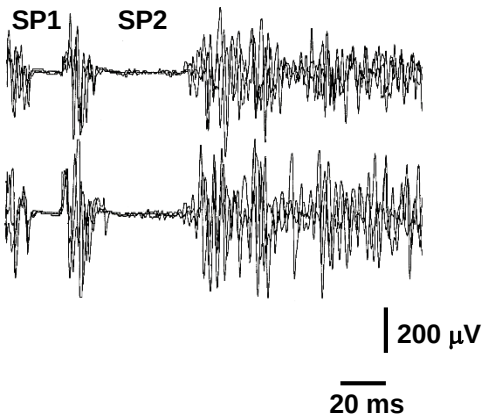


Normal responses

Blink reflex

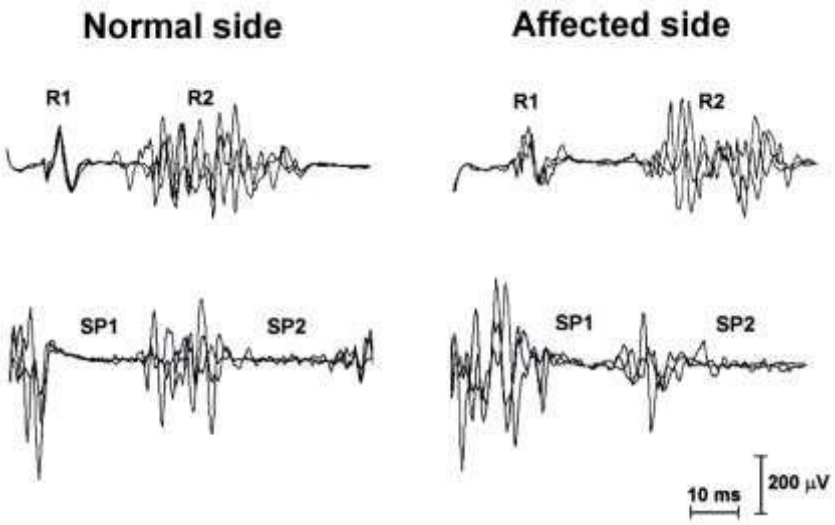


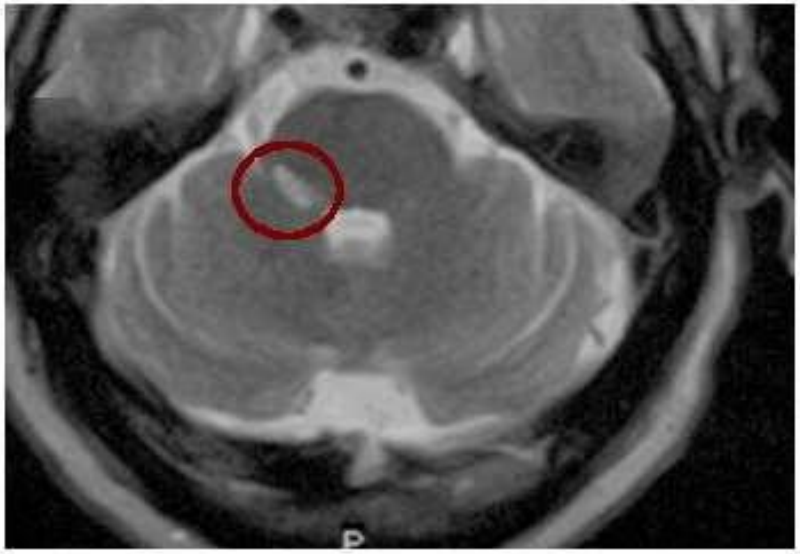
Masseter inhibitory reflex



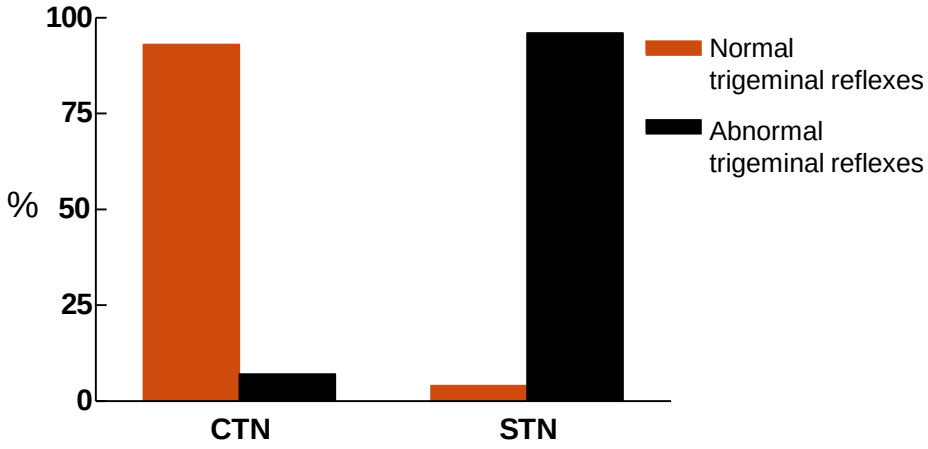
Trigeminal reflexes in trigeminal neuralgia

Trigeminal reflexes in Multiple sclerosis





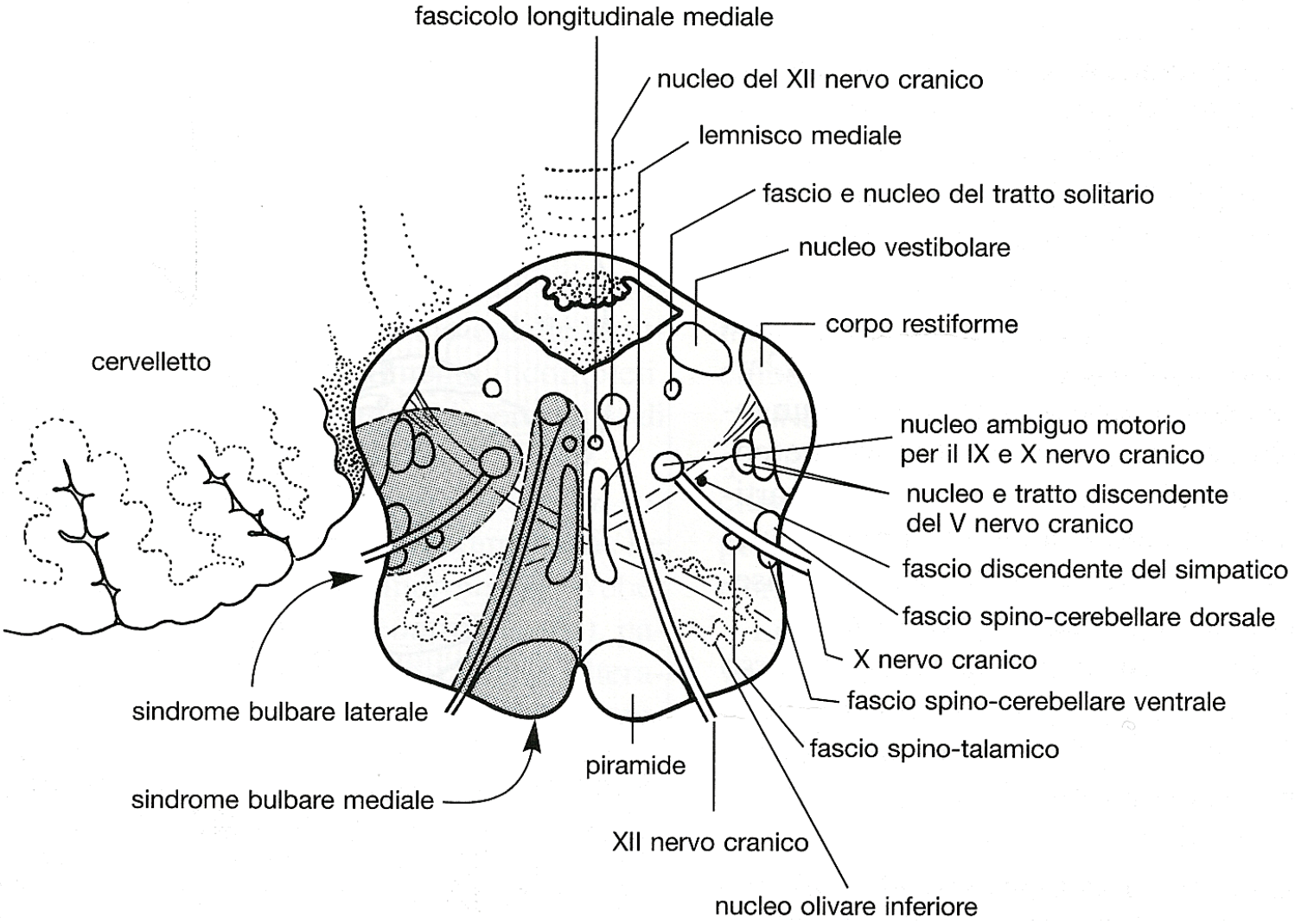
Findings in 628 patients



EFNS guidelines on trigeminal neuralgia management

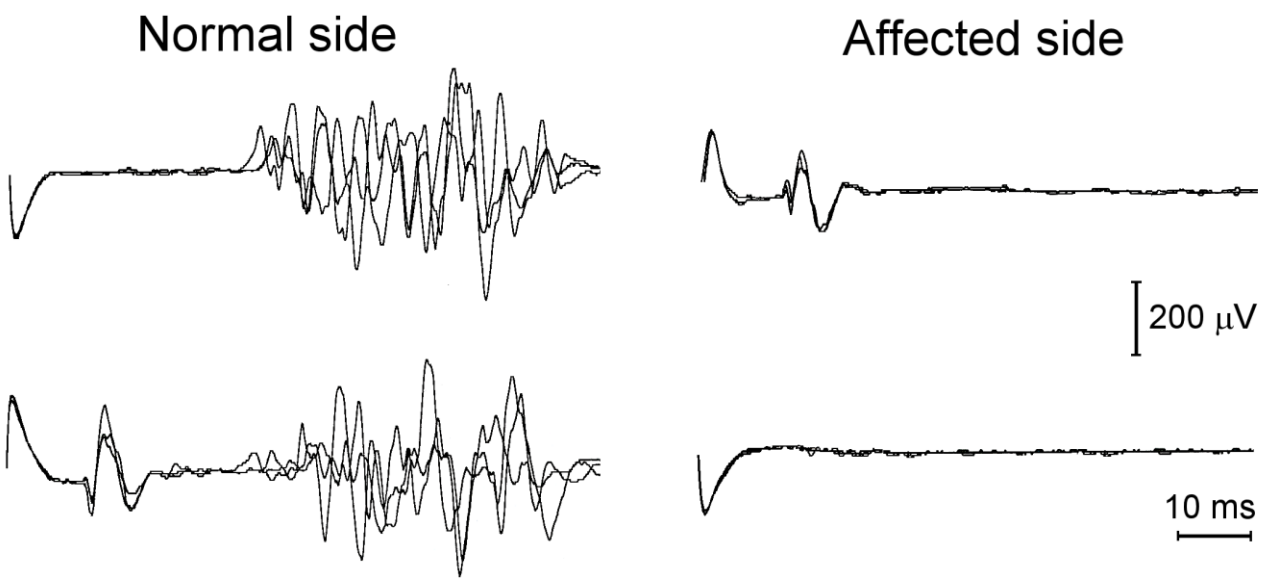
Because of a high specificity (94%) and sensitivity (87%) trigeminal reflexes are reccommended in distinguishing STN from CTN.

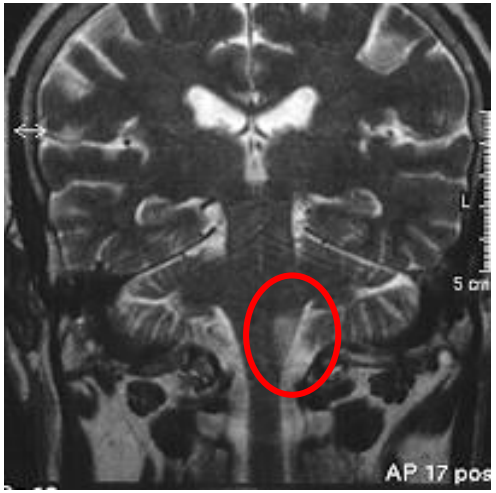
Vascolarizzazione bulbo

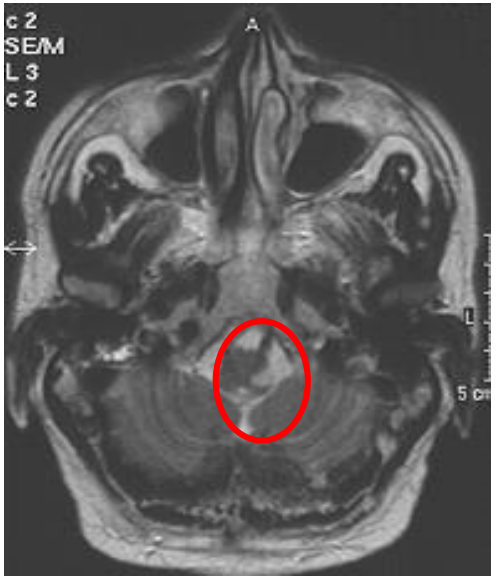


•**Omolateralmente** alla lesione: sindrome di Bernard-Horner, atassia, ipoestesia facciale termodolorifica, paralisi dell'emivelo, dell'emifaringe e dei muscoli laringei.
•**Controlateralmente** alla lesione: ipoestesia termodolorifica emisoma.

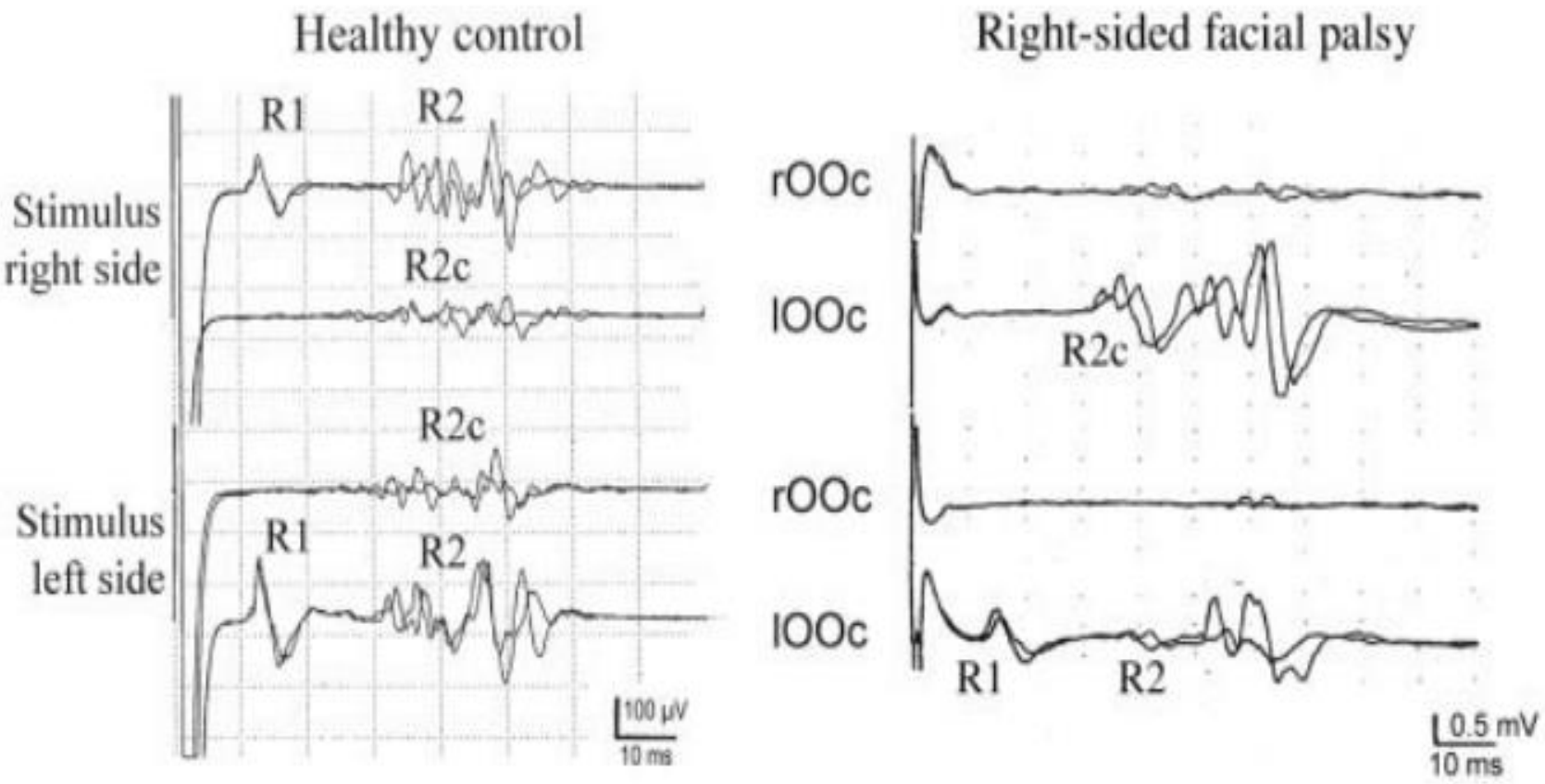
Wallenberg syndrome







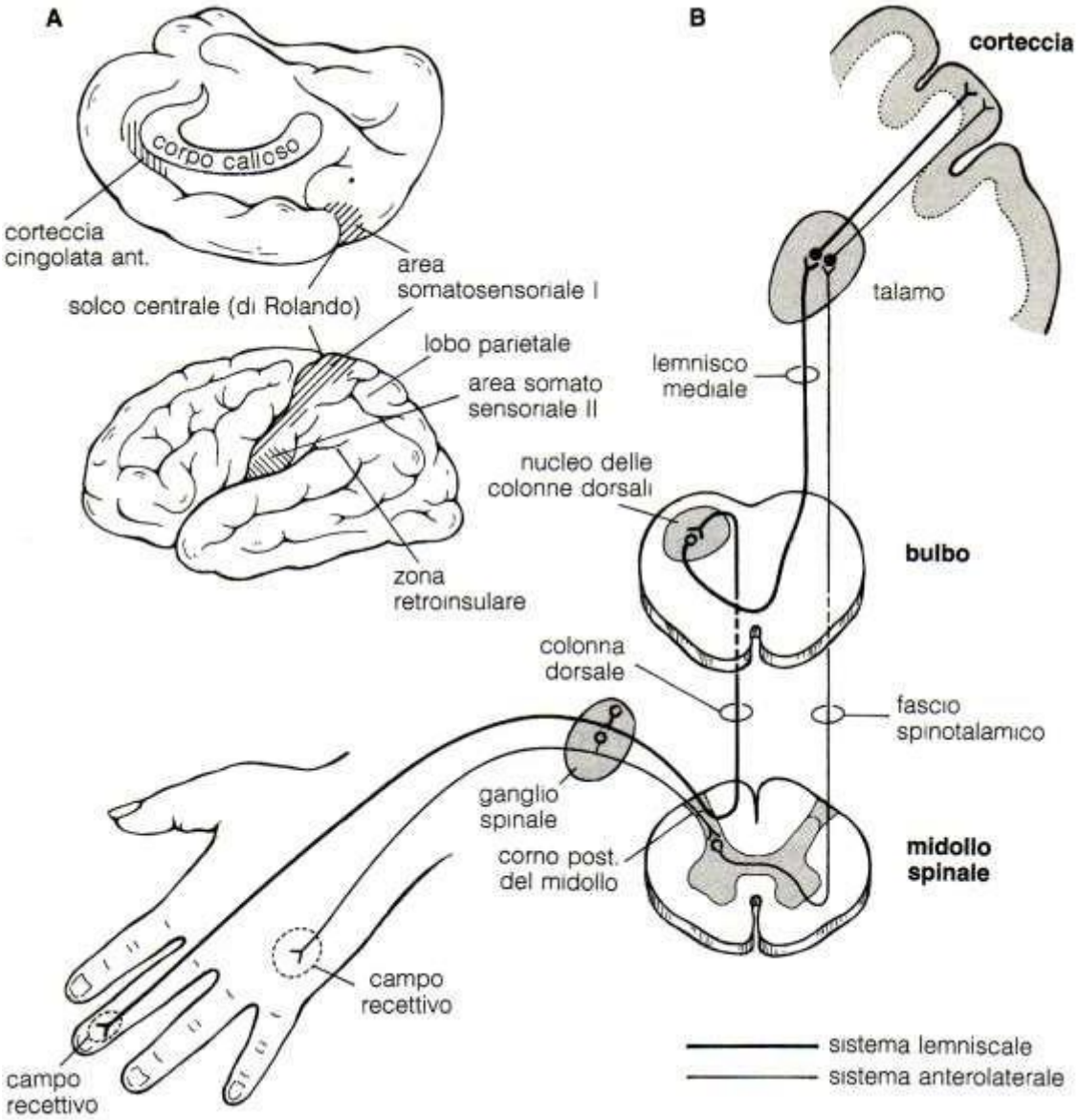
Idiopathic facial palsy



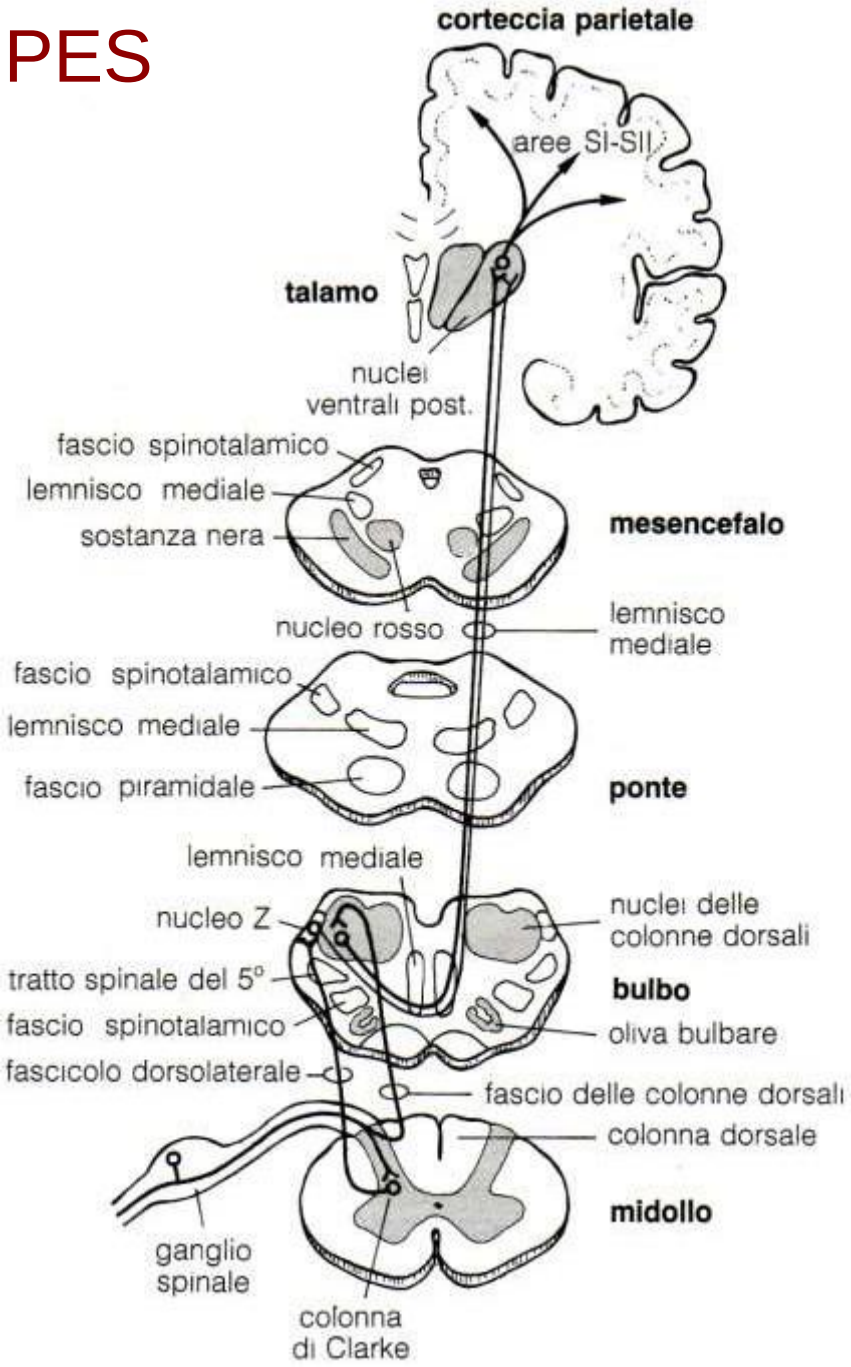
Virtually all patients show abnormalities of the R1 and R2 components in the ipsilateral orbicularis oculi muscle, regardless of the side of the stimulation

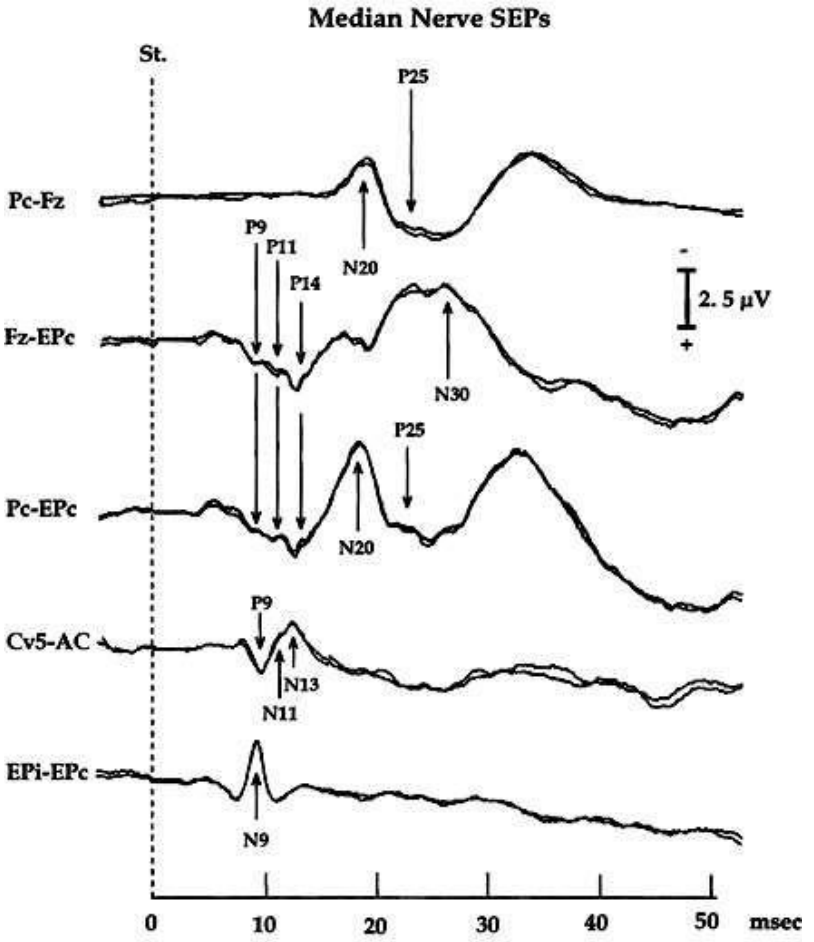
Efferent Abnormality

Sistema nervoso centrale. Studio dei potenziali evocati



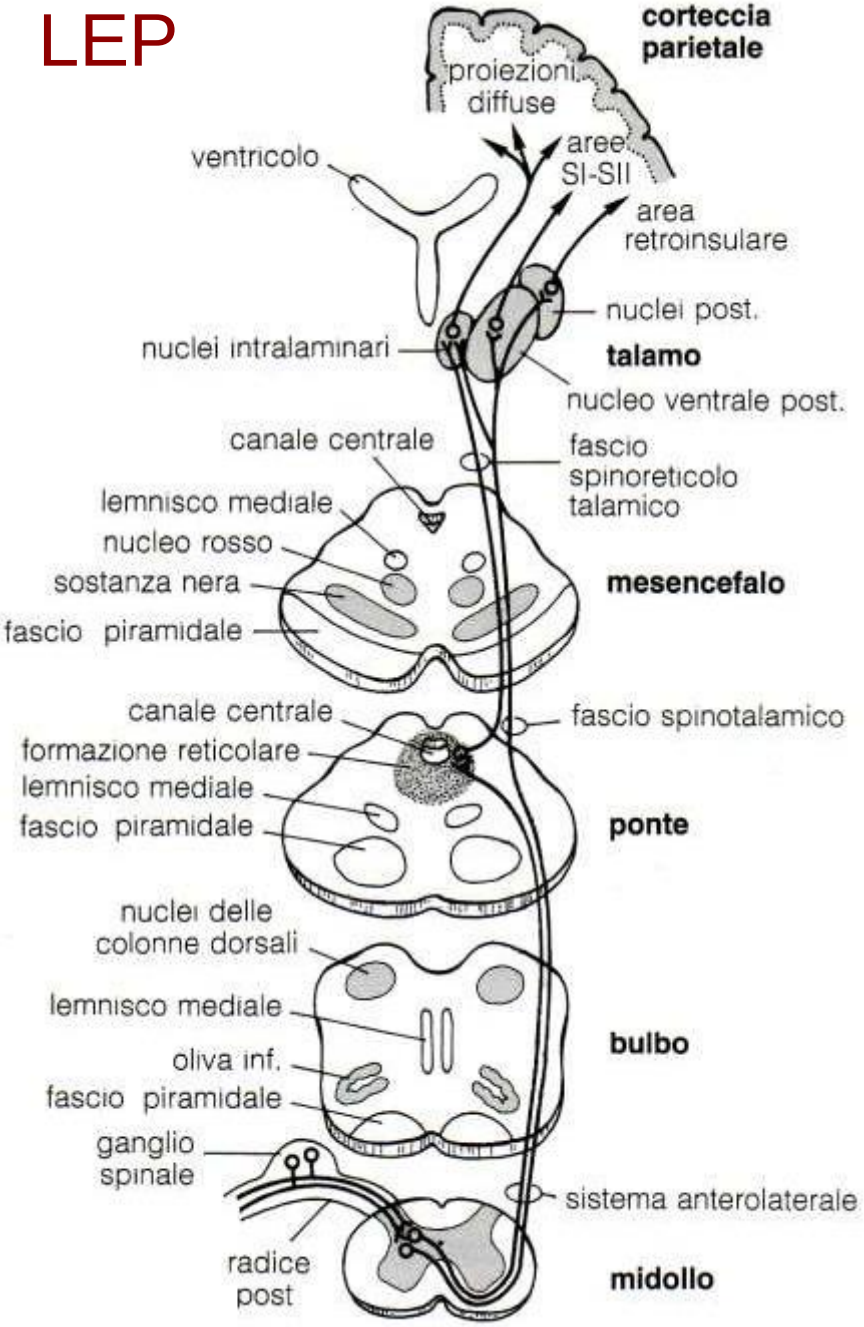
PES

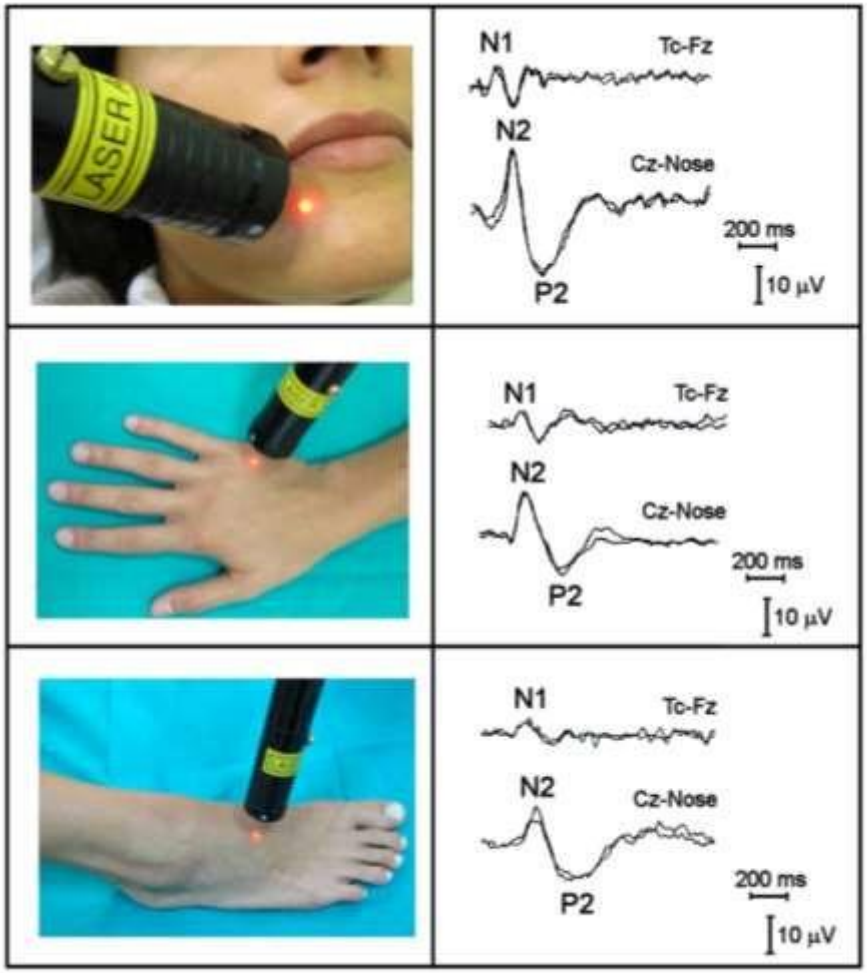




I PES (potenziali evocati somatosensoriali) sono alterati in ogni patologia in cui ci sia un danno del sistema cordoni posteriori-lemnisco mediale.

LEP

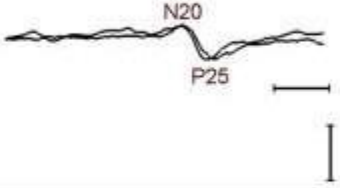

corteccia parietale
proiezioni diffuse
ventricolo
aree SI-SII
area retroinsulare
nuclei post.
nuclei intralaminari
talamo
nucleo ventrale post.
canale centrale
fascio spinoreticolo talamico
lemnisco mediale
nucleo rosso
sostanza nera
fascio piramidale
mesencefalo
canale centrale
fascio spinotalamico
formazione reticolare
lemnisco mediale
fascio piramidale
ponte
nuclei delle colonne dorsali
lemnisco mediale
oliva inf.
fascio piramidale
bulbo
ganglio spinale
sistema anterolaterale
radice post
midollo

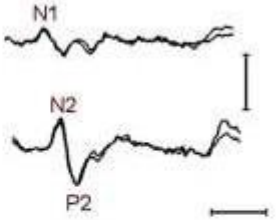

LASER
N1
Tc-Fz
N2
Cz-Nose
200 ms
P2
10 µV
N1
Tc-Fz
N2
Cz-Nose
200 ms
P2
10 µV
N1
Tc-Fz
N2
Cz-Nose
200 ms
P2
10 µV

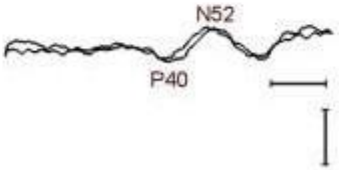
Spinal MRI

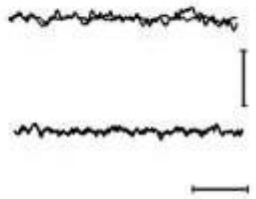
Somatosensory evoked potentials

Laser evoked potentials





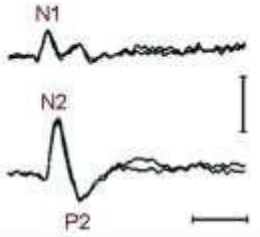


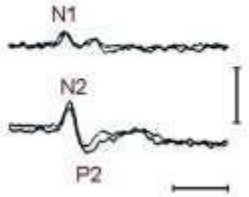


Spinal MRI

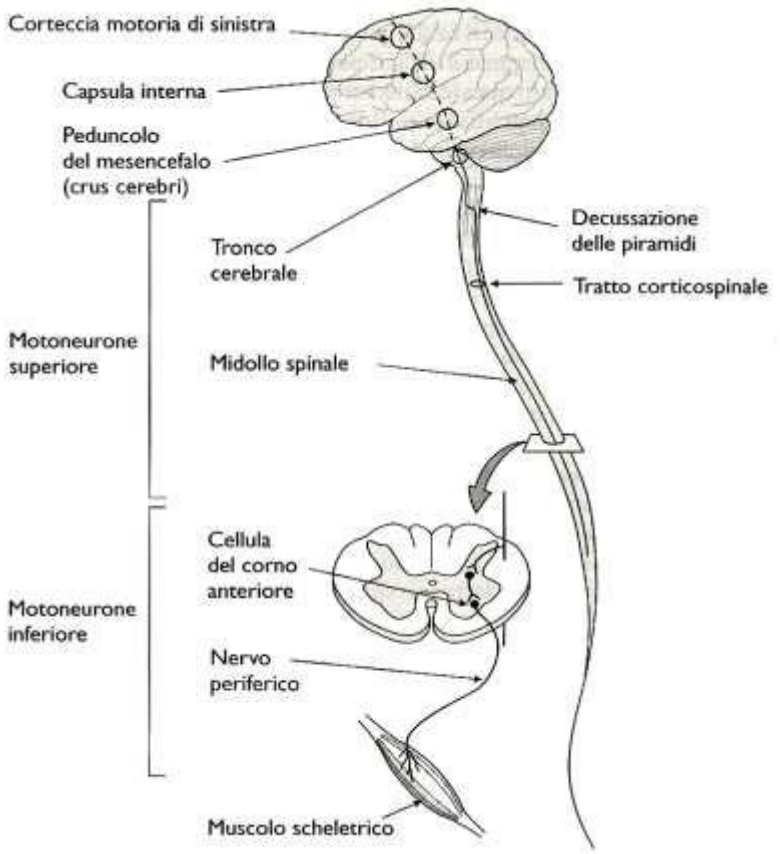
Somatosensory evoked potentials

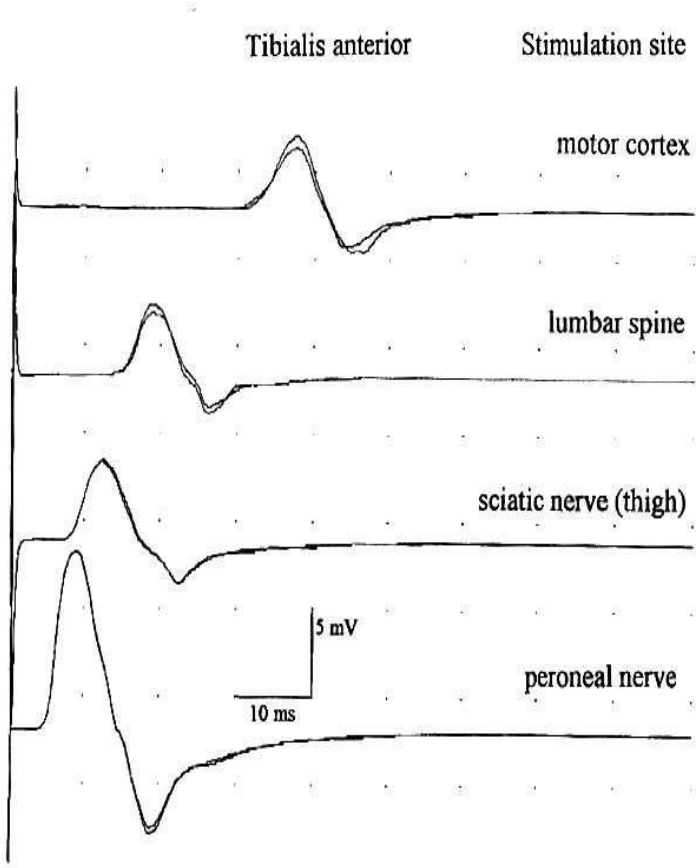
Laser evoked potentials





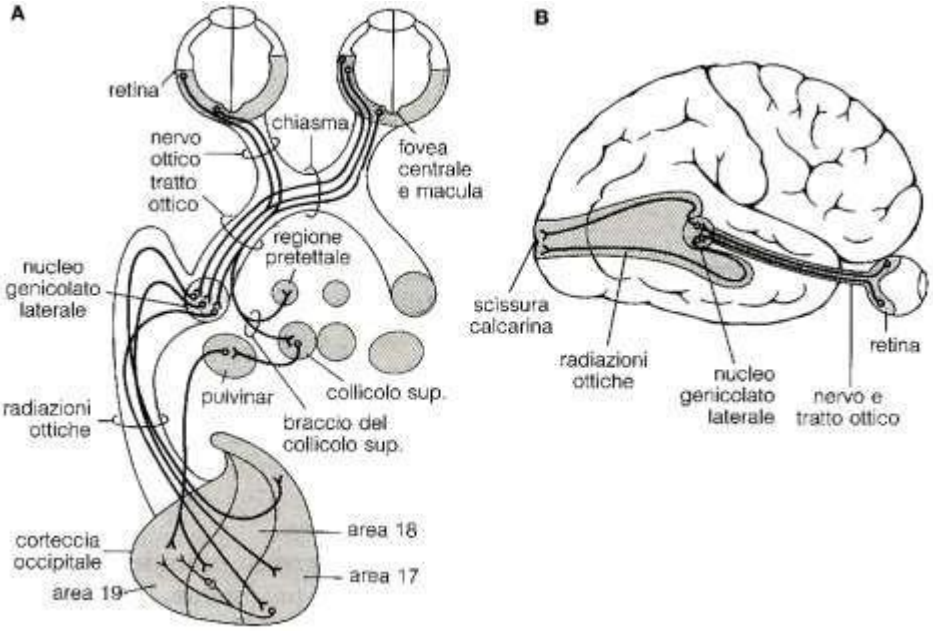
MEP

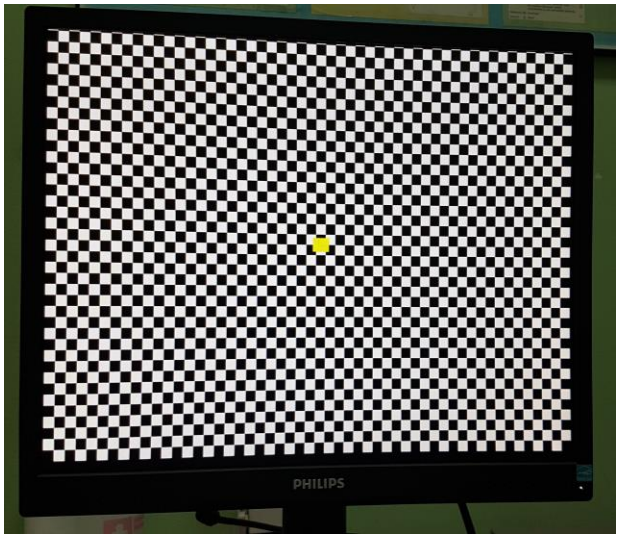


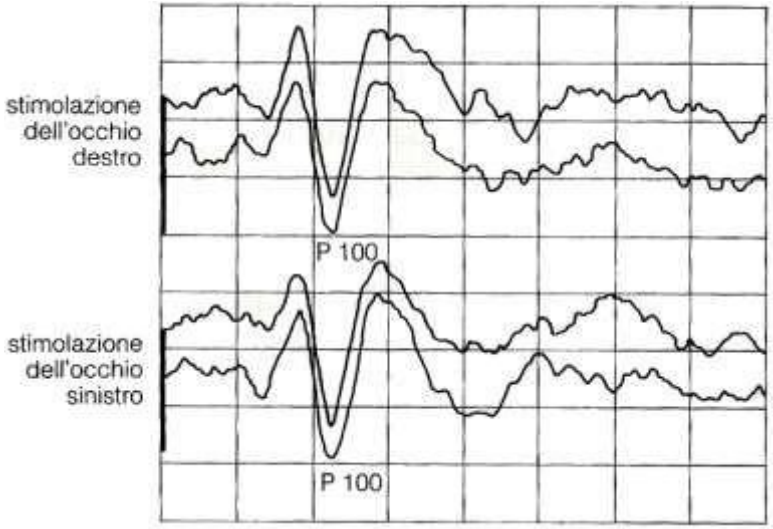


Lo stimolo magnetico attiva la corteccia motoria ed evoca potenziali evocati motori che vengono registrati nella muscolatura degli arti.

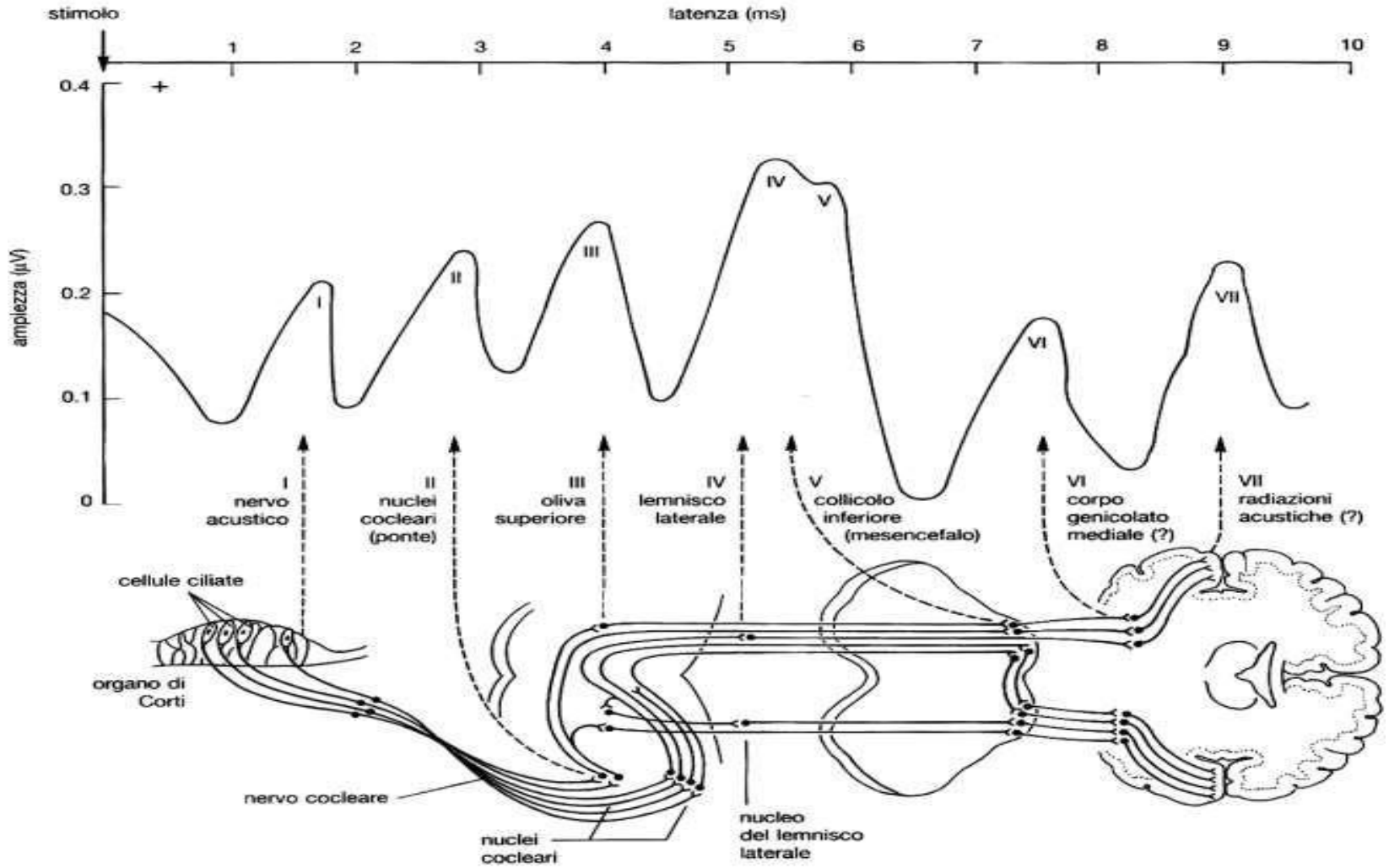
PEV


A
retina
chiasma
nervo ottico
tratto ottico
fovea centrale e macula
regione pretettale
nucleo genicolato laterale
pulvinar
collicolo sup.
braccio del collicolo sup.
radiazioni ottiche
corteccia occipitale
area 18
area 17
area 19
B
scissura calcarina
radiazioni ottiche
nucleo genicolato laterale
nervo e tratto ottico
retina


PHILIPS

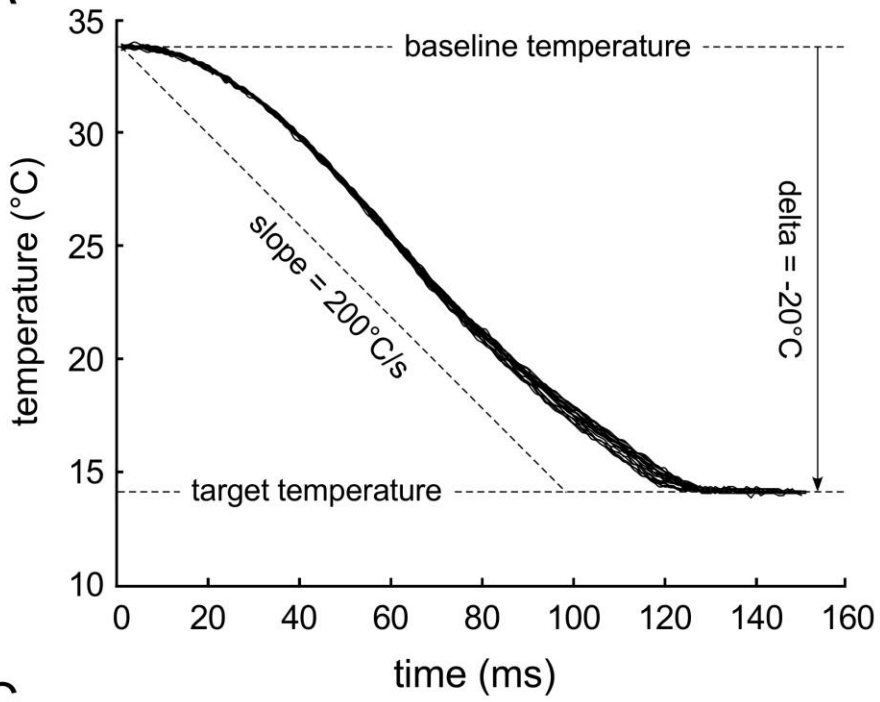

stimolazione dell'occhio destro
P 100
stimolazione dell'occhio sinistro
P 100

BAEPs

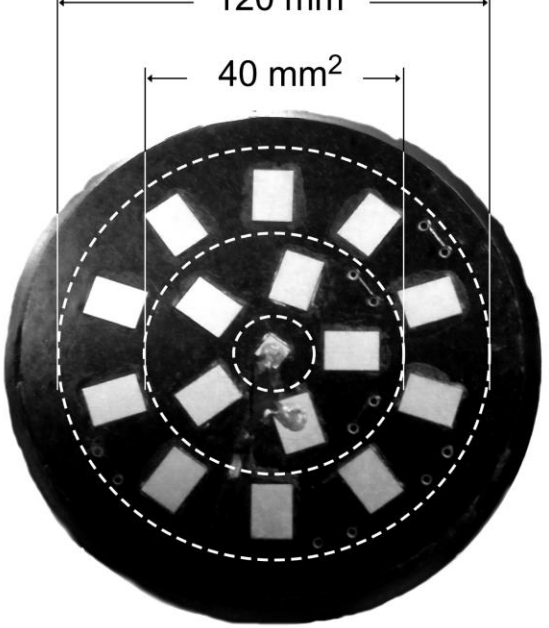

stimolo
latenza (ms)
1
2
3
4
5
6
7
8
9
10
0.4
+
0.3
0.2
0.1
0
ampiezza (μV)
I
II
III
IV
V
VI
VII
I
nervo
acustico
II
nuclei
cocleari
(ponte)
III
oliva
superiore
IV
lemnisco
laterale
V
collicolo
inferiore
(mesencefalo)
VI
corpo
genicolato
mediale (?)
VII
radiazioni
acustiche (?)
cellule ciliate
organo di
Corti
nervo cocleare
nuclei
cocleari
nucleo
del lemnisco
laterale

Cold evoked potentials

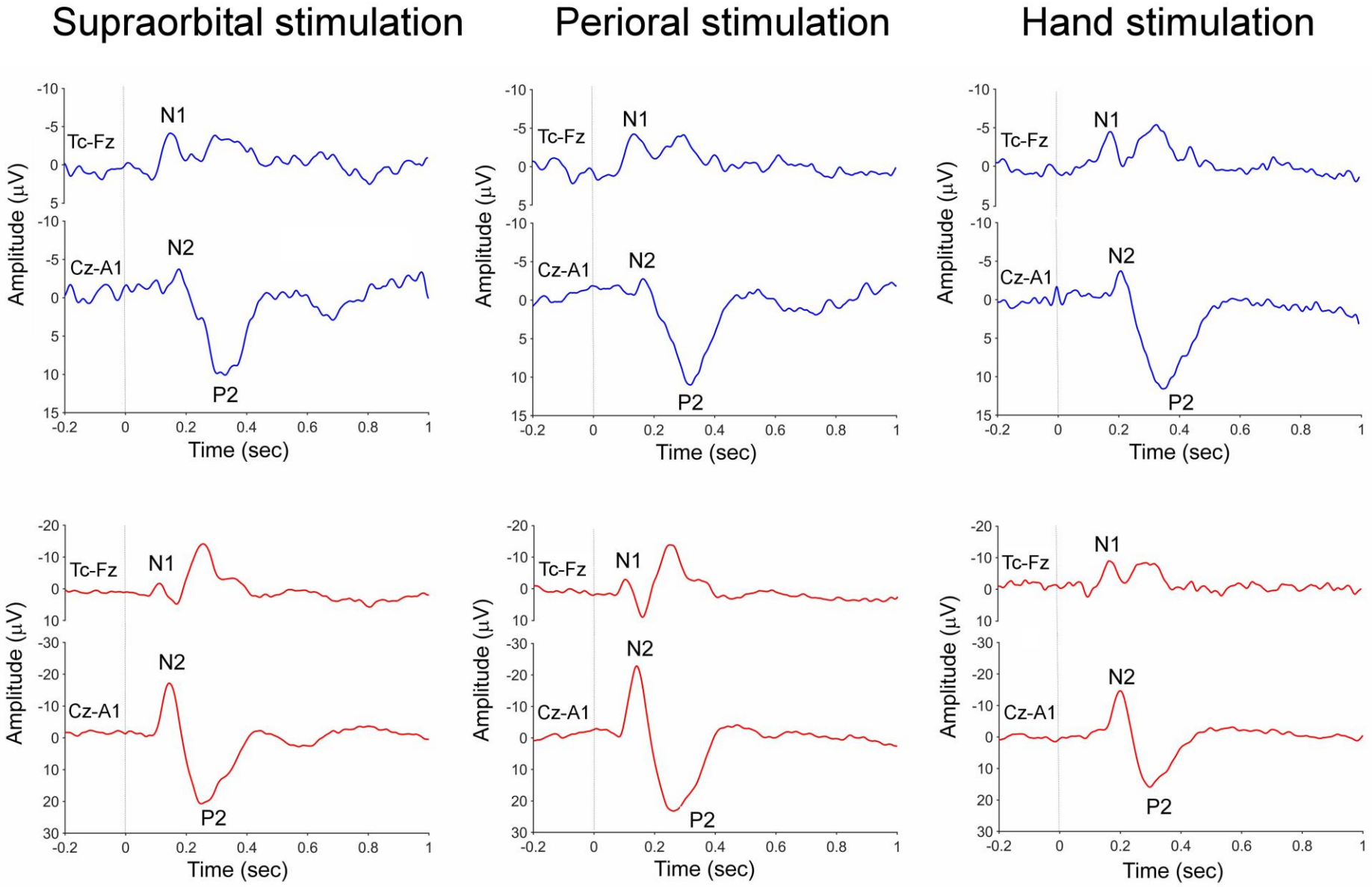
A



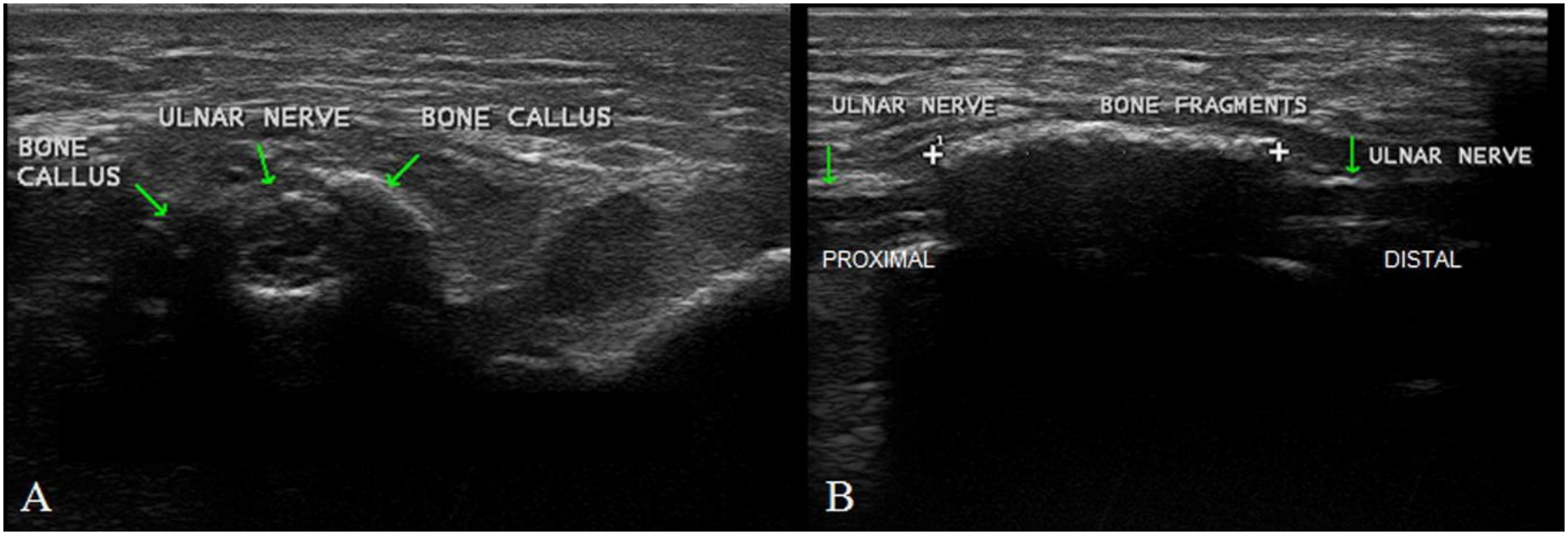
B



C


Supraorbital stimulation
Perioral stimulation
Hand stimulation
Tc-Fz
Cz-A1
N1
N2
P2
Amplitude (μV)
Time (sec)

Ecografia di nervo



ENG: Ulnar nerve sensory responses were absent. Ulnar motor nerve conduction studies, recorded from first dorsal interosseous and abductor digiti minimi, showed reduced motor nerve action potential amplitude and neuroapraxic block associated with severe slowing of conduction velocity (6 m s–1) in the above–below elbow trAt US evaluation, the ulnar nerve was normal at the elbow but could not be seen distally. After careful evaluation, we realised that the nerve entered a hyperechoic tunnel likely to be the callus osseous; longitudinally, the hyperechoic structure had a maximum length of 17 mm. Distal to this structure, the ulnar nerve was visible again, appearing hypoechoic and with increased volume (CSA: 16 mm2).act (stimulating the nerve 4 cm above and below elbow). Motor conduction velocity was normal in the arm and the forearm.

Grazie per l’attenzione!