

Fabry disease

Theoretical usefulness of electrophysiologic evaluation

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Fiber types

LARGE FIBERS
A β & motor fibers

SMALL FIBERS
A δ C

Diameter

5-15 μ m

1-5 μ m

0,3-1,5 μ m

Myelin sheath

+++

+

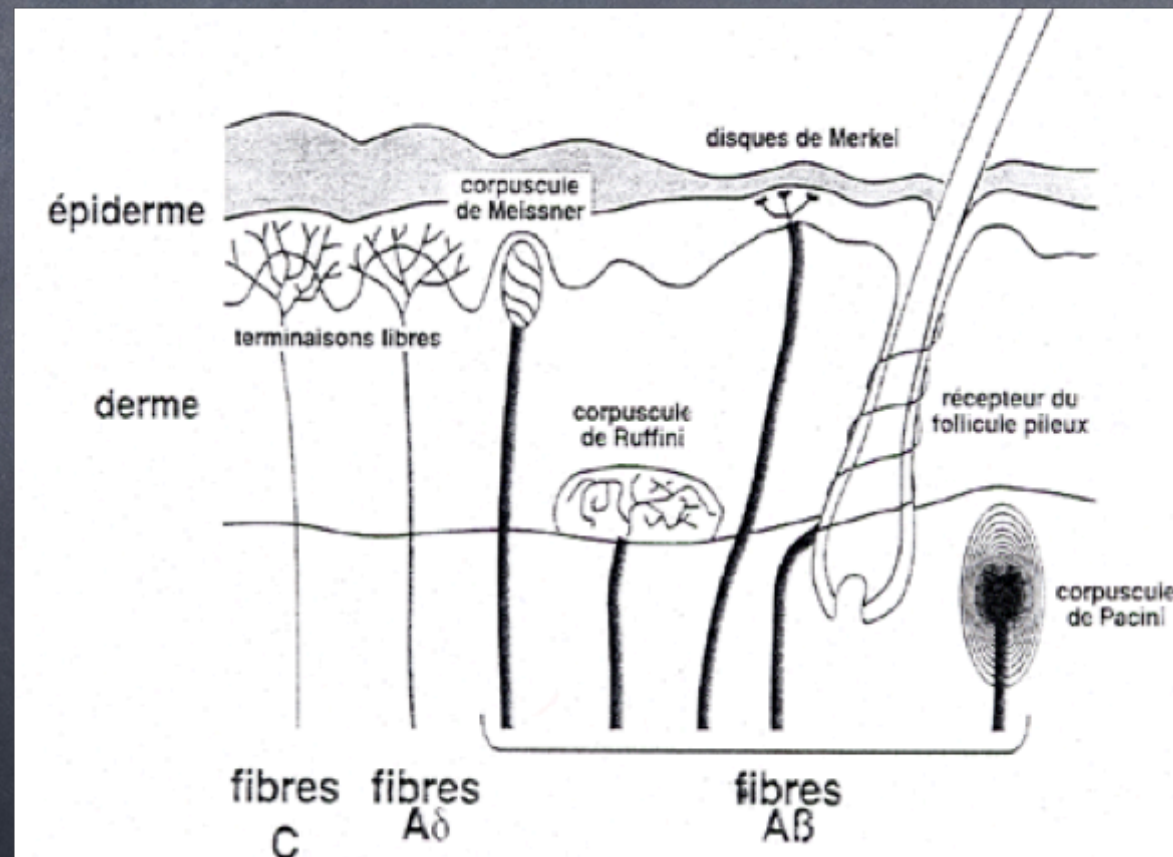
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Conduction velocity

40-100 m/s

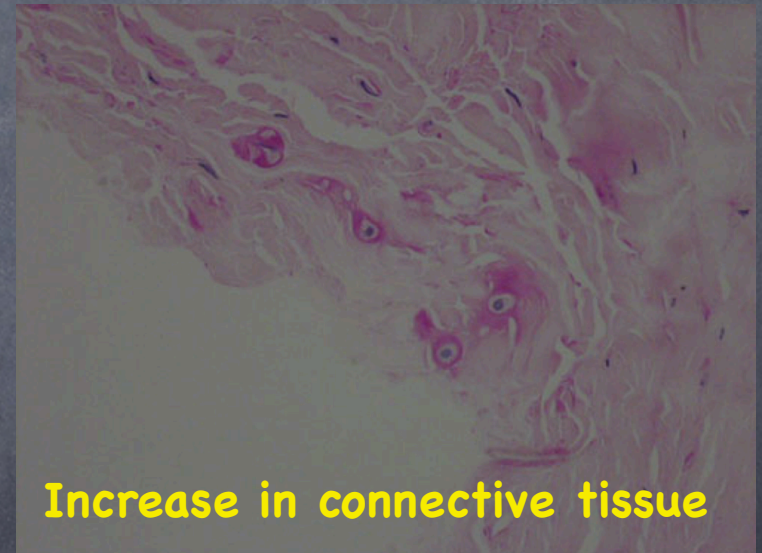
5-40 m/s

1-2 m/s



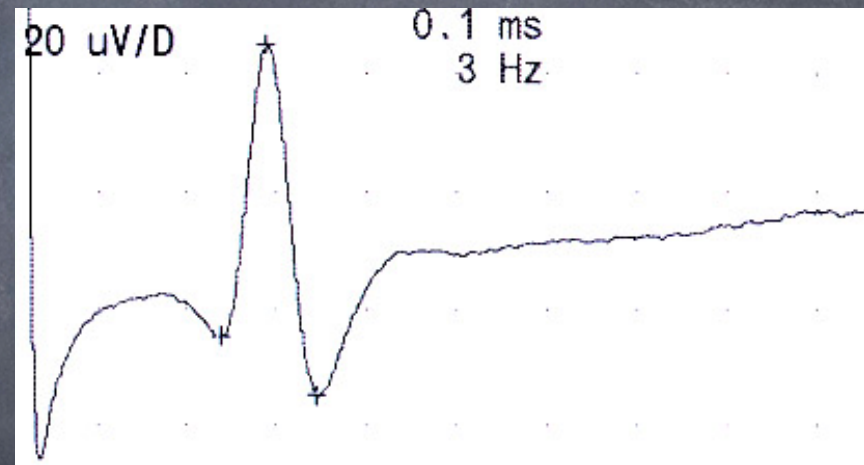
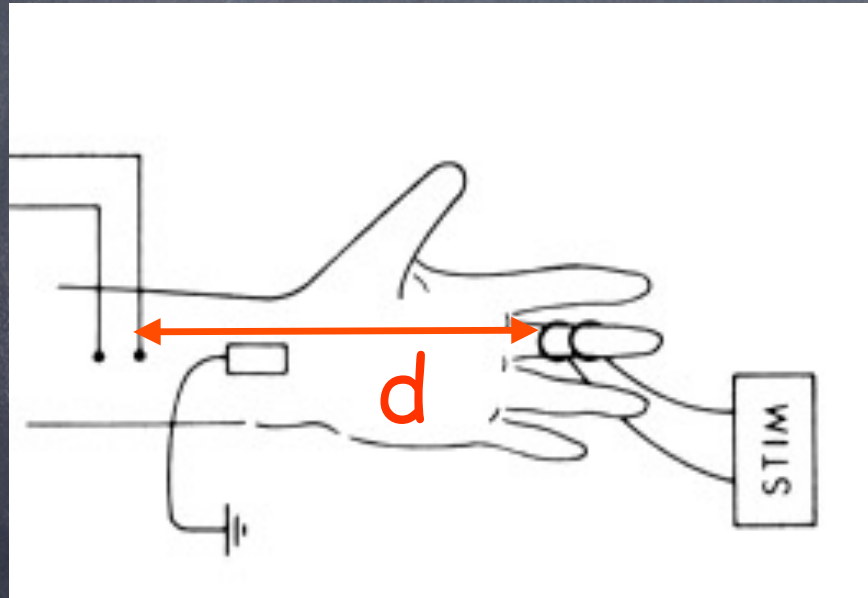
Pathologies

- large fiber neuropathy : increased incidence of carpal tunnel syndrome (25% of patients ?)
 - sensory neurography
 - motor neurography
 - EMG
- small fiber neuropathy
 - sympathetic skin response
 - R-R interval variation
 - laser evoked potentials



Increase in connective tissue

Sensory neurography



LSD

Sensory neurography


droit Médian
Medianus
0µV/D 20µV/D EMG1
5ms/D

Paume-Poignet
oy.:0 Rejets:0

Dig I - Poignet
Dig II - Poignet
Dig III - Poignet
Dig IV - Poignet
Poignet - Thénar

Droit Sensitif Medianus

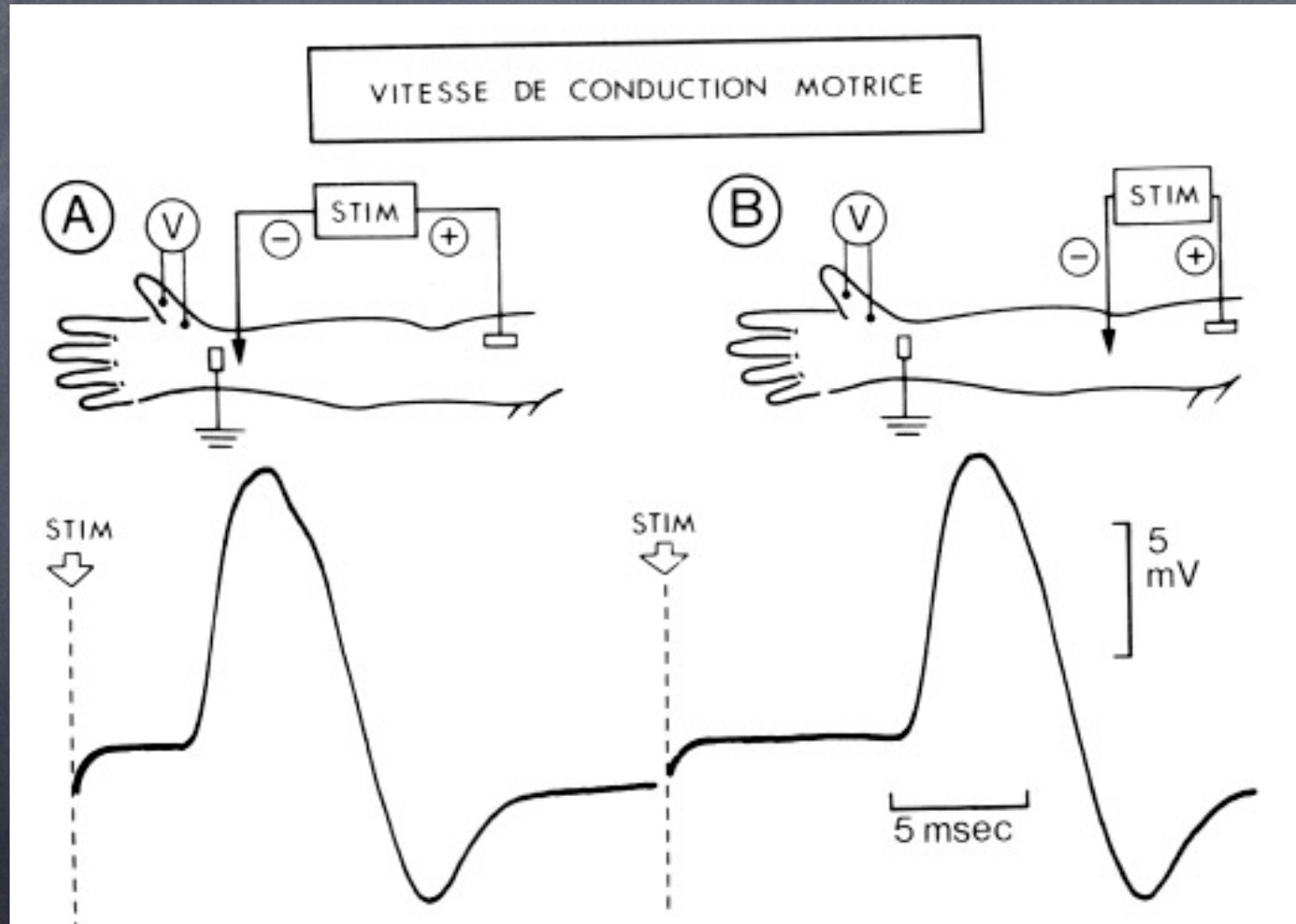
Sites	Dist [mm]	CV [m/s]	Amp [µV]	Lat [ms]	pLat [ms]	Area [ms*µV]	Stim [mA]	Temp [°C]



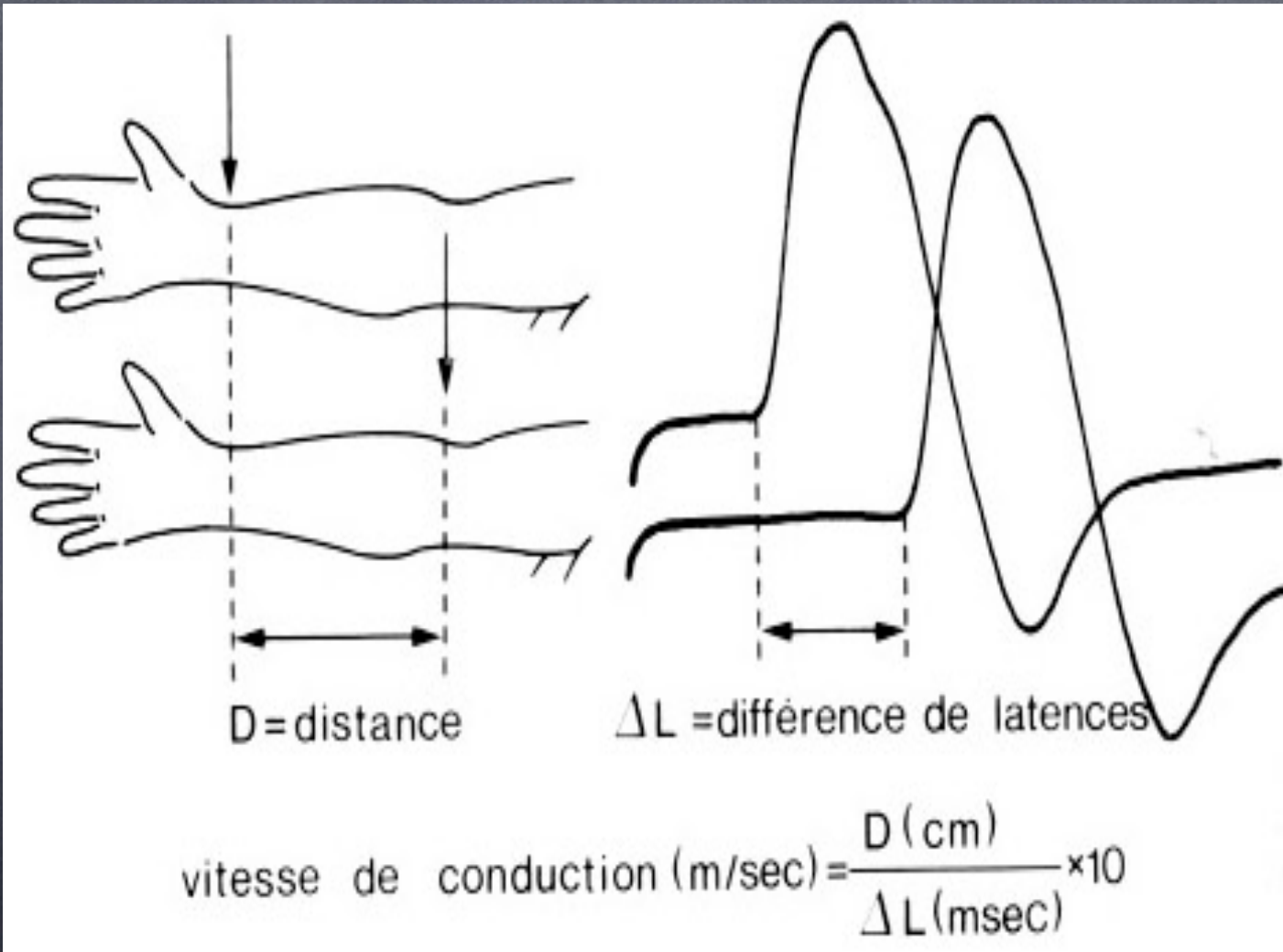
Sens. d'entrée Sensibilité du moy. Données CN Déplacer vers l'historique Superposer

Nouveau Site Filtrer Autre côté Gauche Nouveau nerf Paramètres

Motor neurography



Motor neurography



Motor neurography

Matrices Courts segments F8 Sensitives F9 Réponses F F10 Réponses F facilité F11 Réponses F doublé F12 Silence temporal F7 Nerve proche F8

Droit Médian-Ulnaire 0.0mA 0.2ms 1.0Hz

Medianus
Enreg.: APB
5mV/D EMG1
5ms/D

Poignet

Droit Moteur Medianus | APB


Sites	Lat [ms]	Amp [mV]	Dur [ms]	Area [ms*mV]	Stim [mA]

Segments Dist [mm] CV [m/s] Amp [%] Dur [%] Area [%]

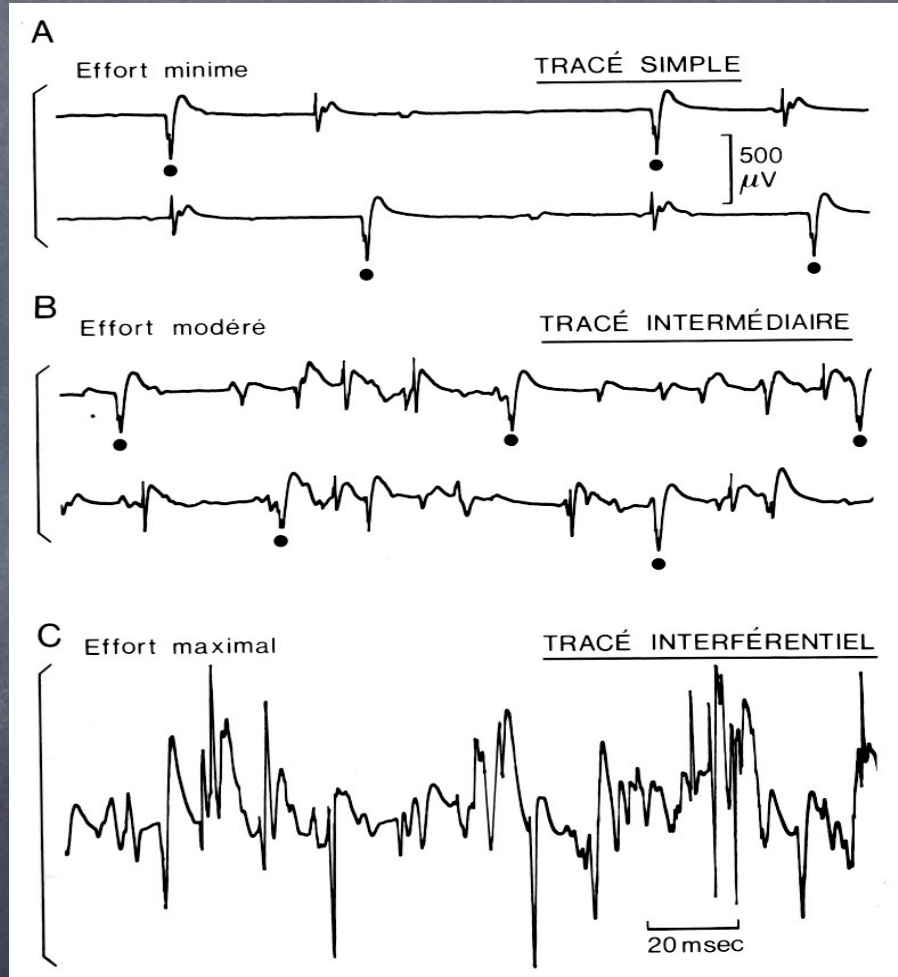
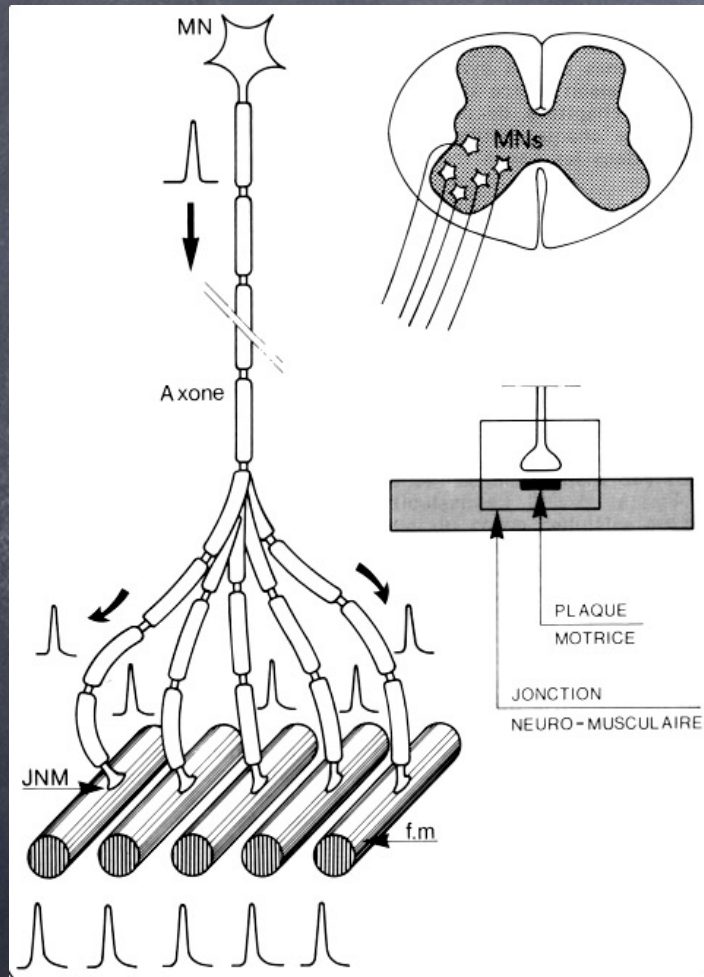
Droit Moteur Medianus | Lombrical

Sites	Lat [ms]	Amp [mV]	Dur [ms]	Area [ms*mV]	Stim [mA]

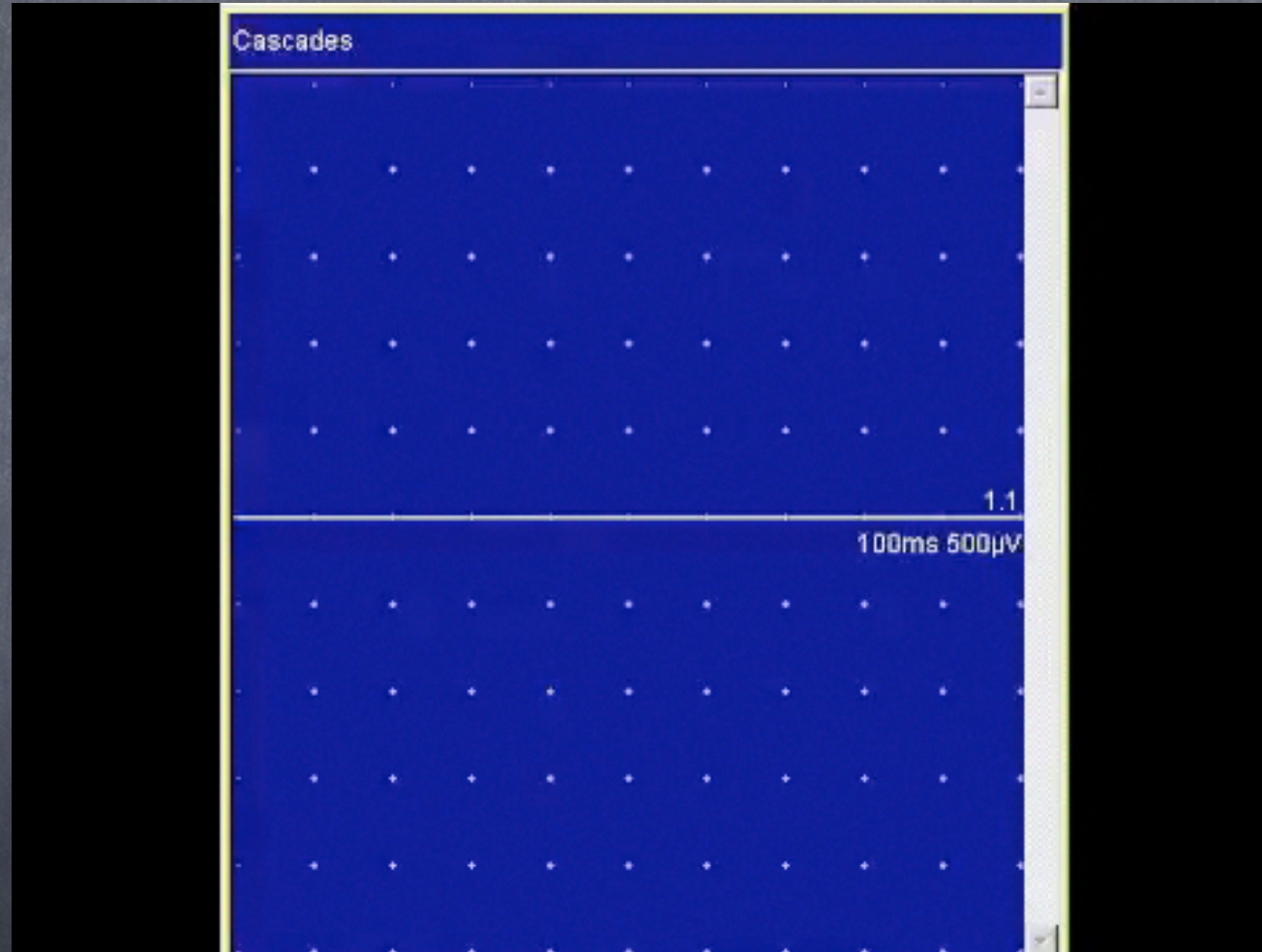
Segments Dist [mm] CV [m/s] Amp [%] Dur [%] Area [%]



EMG

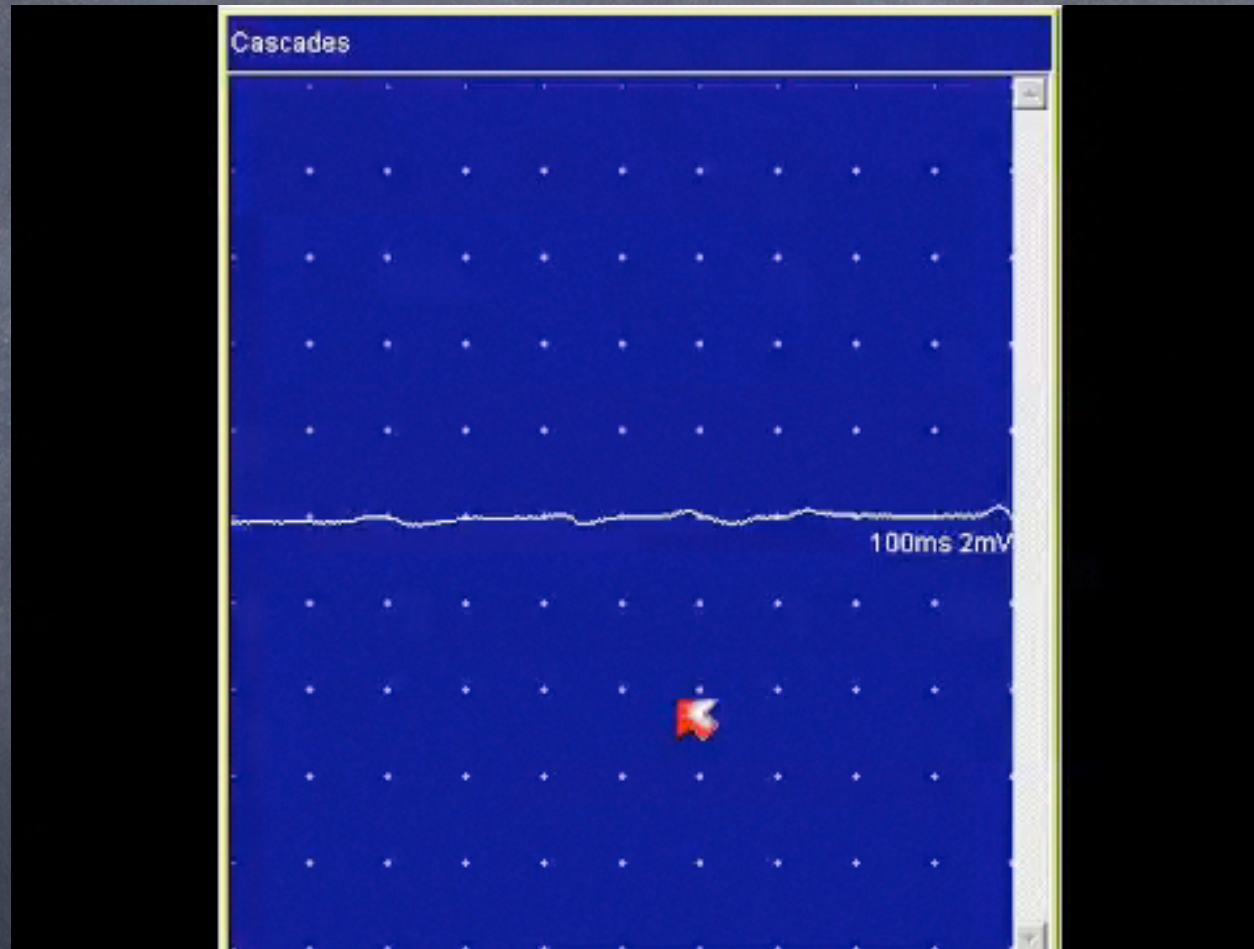


EMG



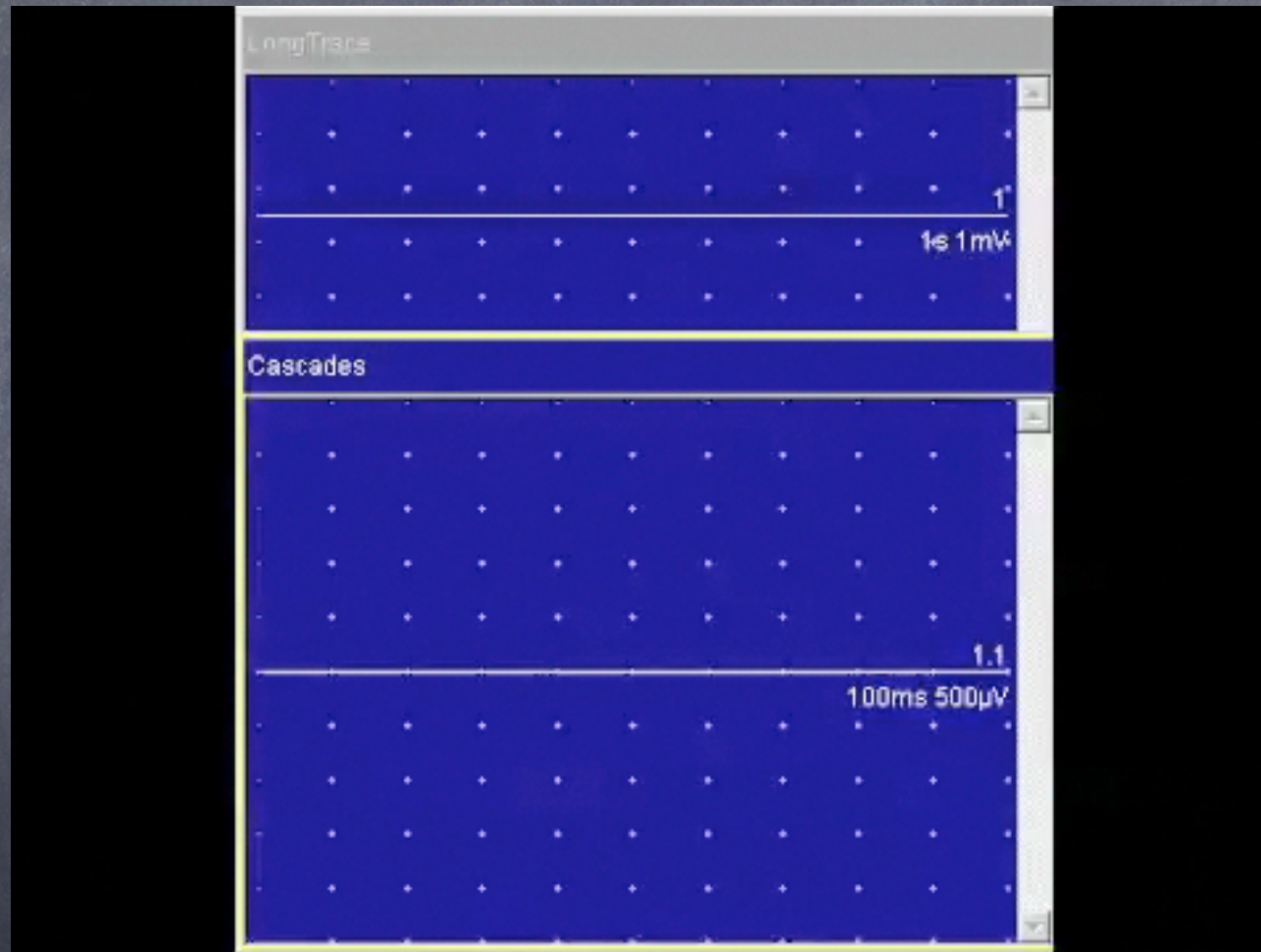
Normal

EMG



Neuro

EMG



Myo

Carpal tunnel syndrome

Gauche Médian
Medianus
5µV/D 20µV/D EMG1
0.8ms/D

14.5mA 0.1ms 3Hz

Gauche Sensitif Medianus

Sites	Dist [mm]	CV [m/s]	Amp [µV]	Lat [ms]	pLat [ms]	Area [ms*µV]	Stim [mA]	Temp [°C]
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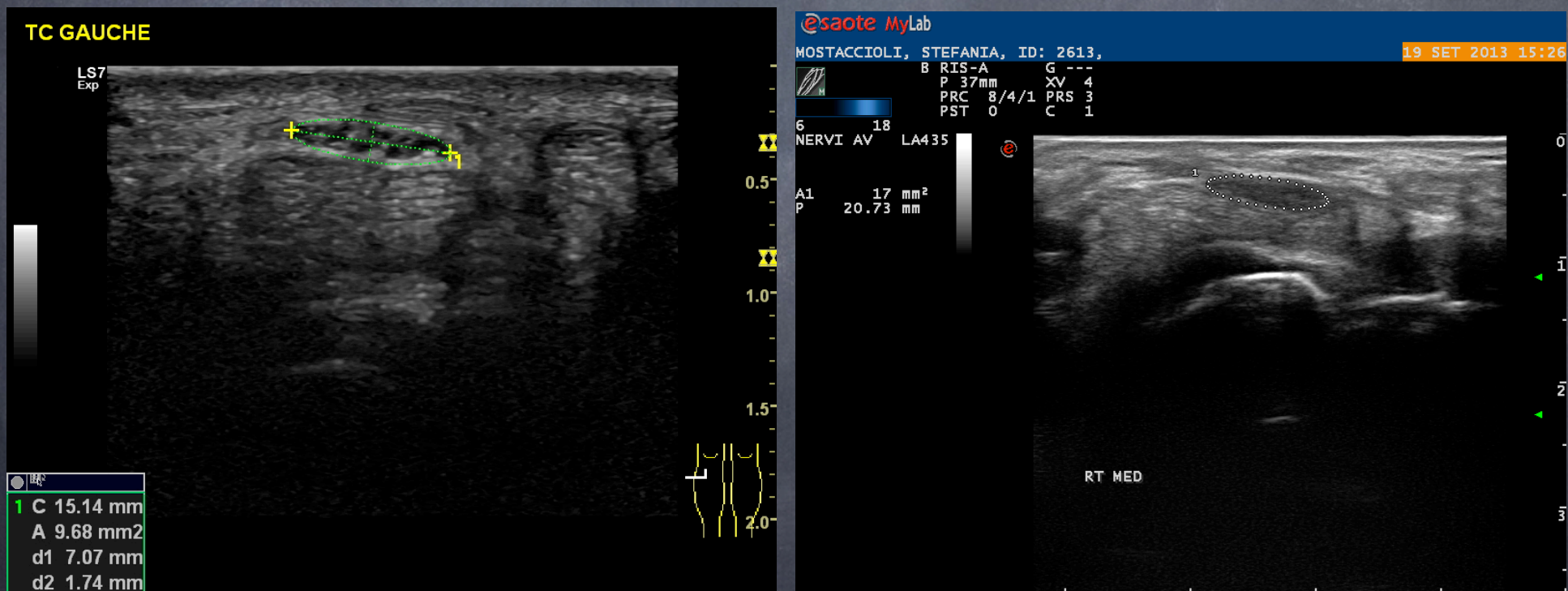
Paume- Poignet
Moy.:0 Rejets:0

Dig I - Poignet
Dig II - Poignet
Dig III - Poignet
Dig IV - Poignet
Poignet - Thénar

Arrêter moyenneur
Sens. d'entrée
Sensibilité du moy.
Données CN
Déplacer vers l'historiq
Superposer
Nouveau Site
Effacer
Autre côté
Droit
Nouveau nerf
Paramètres

Carpal tunnel syndrome

Ultrasonography



Sympathetic skin response

- Efferent part of the SSR reflex : **myelinated sympathetic fibers** from the spinal cord that terminate in paravertebral sympathetic ganglia. Postganglionic fibers are **non myelinated (type C)** and innervate the eccrine sweat glands.
- The central part of the reflex arch is not fully understood yet. It is presumably polysynaptic with a connection to the structures of hypothalamus, ventrolateral part of the brainstem, medial and basal parts of the frontal lobe and medial part of the temporal lobe

Sympathetic skin response

- SSR can be evoked by different types of stimuli
 - electrical stimulation of peripheral nerve in the extremity
 - noise
 - brisk inspiration
 - cough effort
 - lie

Sympathetic skin response

RCS bruit RCS stim FS

Droit Mains et pieds 0.0mA 0.2ms 1.0Hz

Enreg.: Main
0.5mV/D EMG1
1s/D

Bruit

Bruit - Pied
Bruit - Main droite
Bruit - Pied droit
Bruit - Main gauche
Bruit - Pied gauche

Droit Moteur - Main droite

Sites	Lat [ms]	Amp [mV]
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Droit Moteur - Pied gauche

Sites	Lat [ms]	Amp [mV]
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Droit Moteur - Pied droit

Sites	Lat [ms]	Amp [mV]
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Droit Moteur - Main

Sites	Lat [ms]	Amp [mV]
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Droit Moteur - Pied

Sites	Lat [ms]	Amp [mV]
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Historique

- Moyenne
- Afficher tout
- Afficher les plus récents 5
- Déplacement auto. vers historique

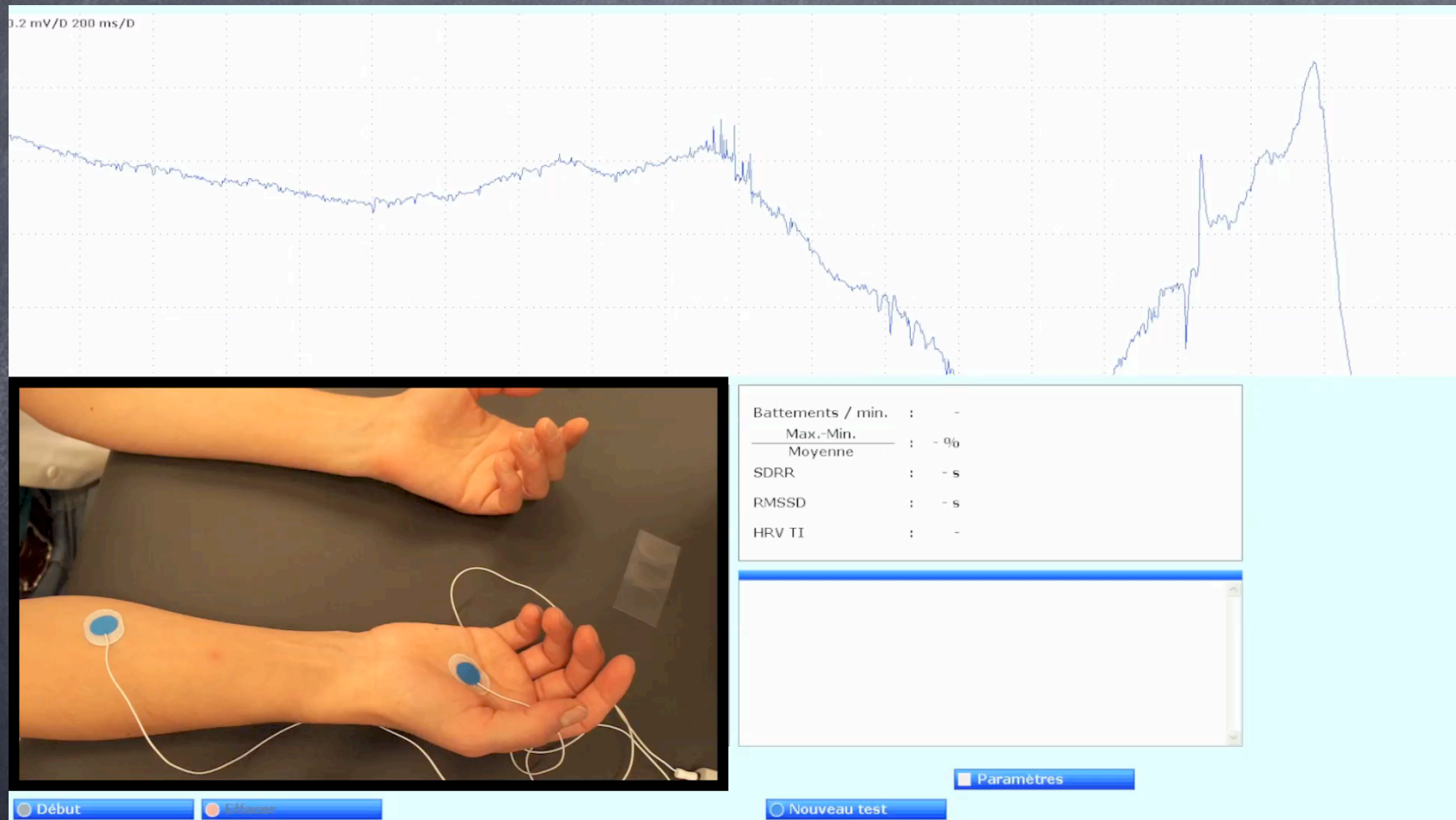
Données CN Déplacer vers Historique Superposer

Nouveau Site Effacer Autre côté Gauche Nouveau nerf Paramètres

R-R interval variation

- R-R interval variation is based on the variability of heart rate in a relaxed state and following hyperventilation.
- Heart rate variability is the result of the instantaneous relationship between excitatory, i.e., **sympathetic**, and inhibitory, i.e., **parasympathetic**, neural influences on the sino-atrial node automatism

R-R interval variation



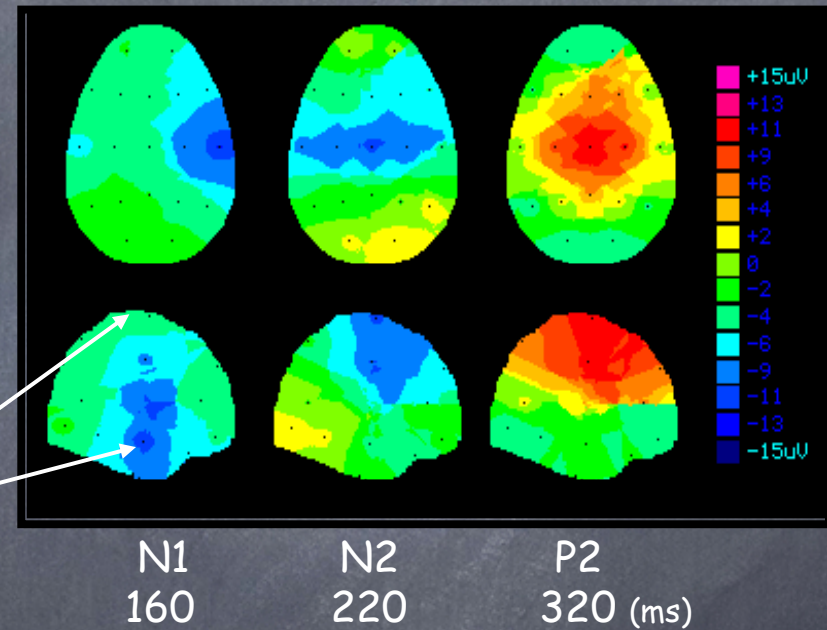
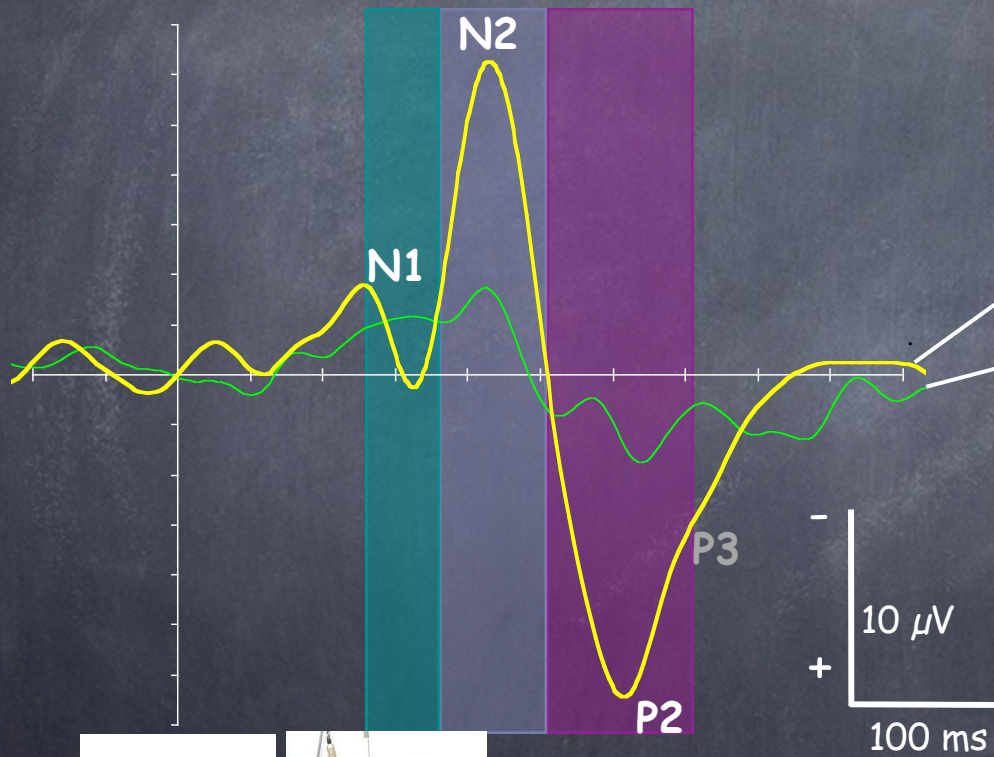
Laser evoked potentials

Characteristics of nociceptif stimuli	Electrical	Thermal			Mechanical	Chemical
		conduct.	radiation			
		<i>thermode</i>	<i>convent.</i>	<i>laser</i>		
Specificity	---	---	+++	+++	---	+++
Noninvasive	+++	+++	+++	+++	+++	---
Reproducibility	+++	±	±	+++	±	±
Time locking	+++	---	---	+++	±	---

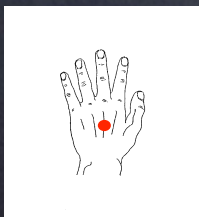
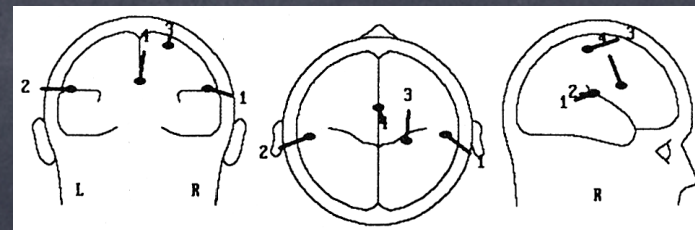
Laser evoked potentials

Grand average Cz and T4 (n = 10)
Left hand

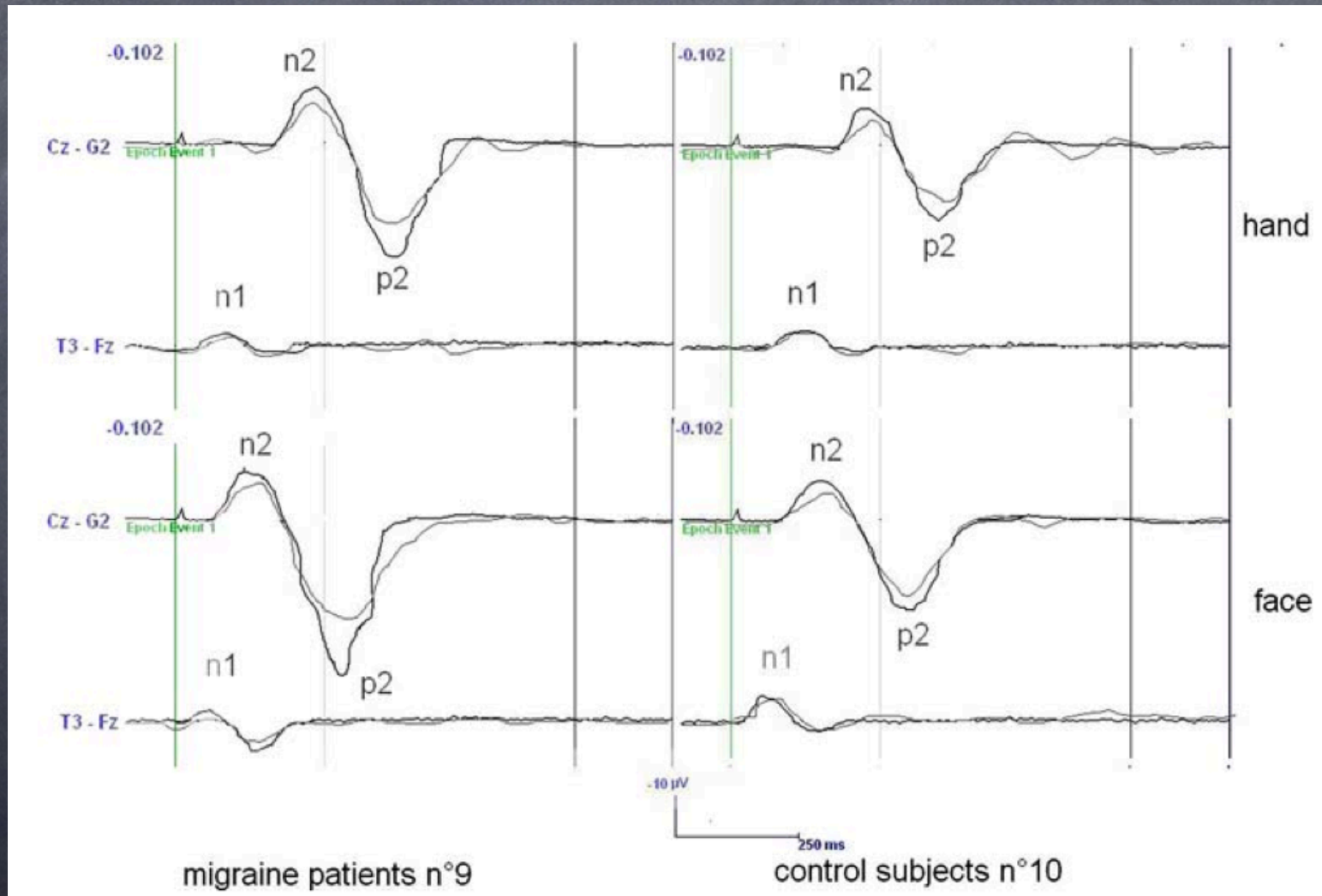
Topographical map
left hand stimulation



Dipolar modeling of late LEPs

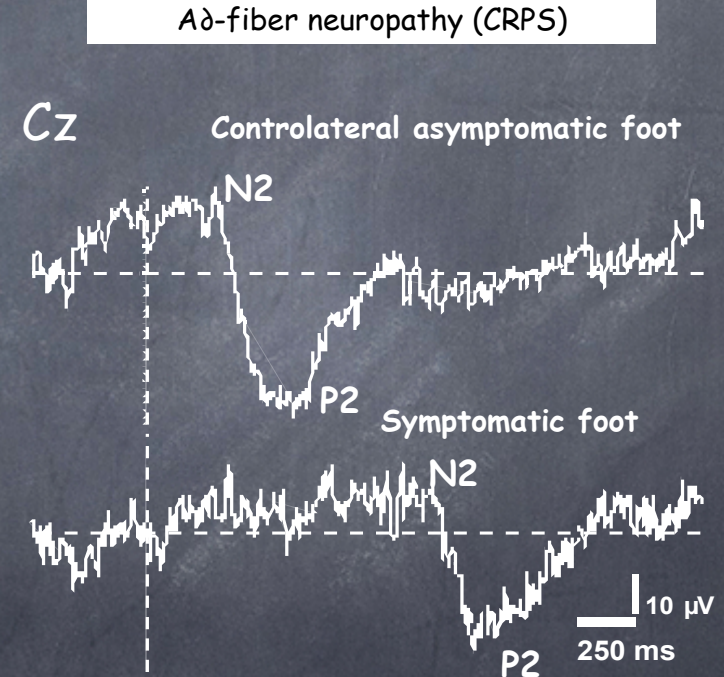
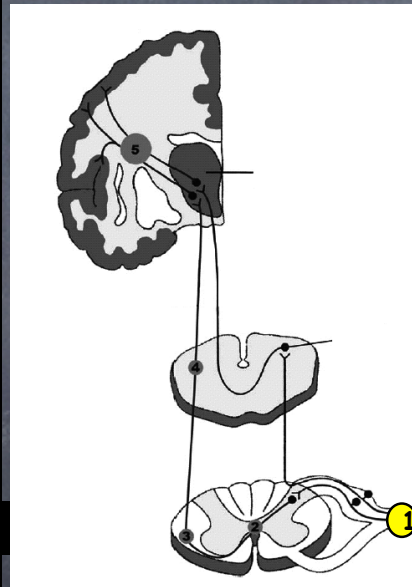
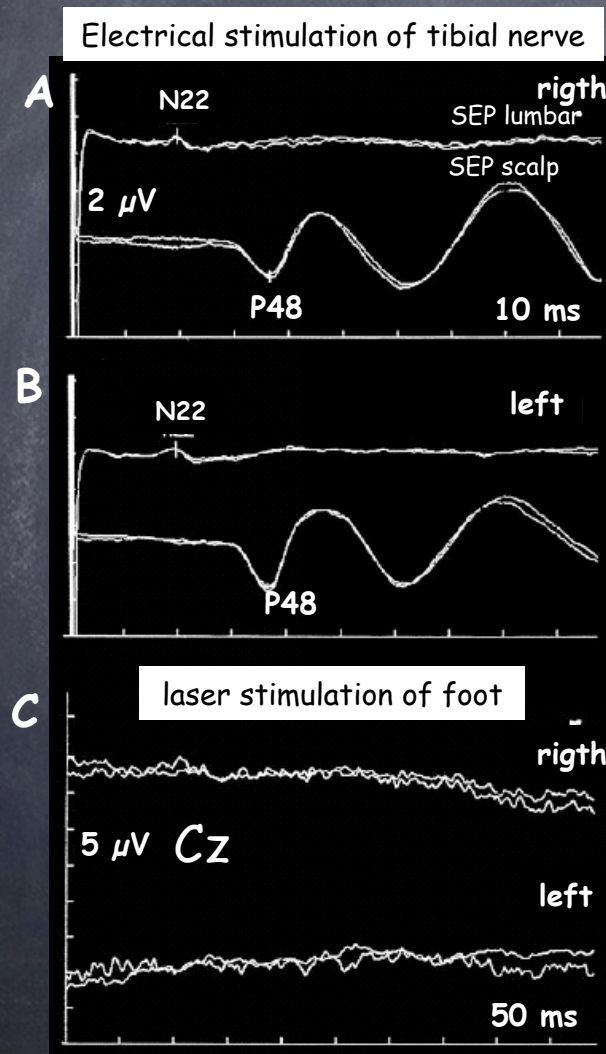


Laser evoked potentials



Laser evoked potentials

1. Small fiber neuropathies



A sural nerve biopsy at the symptomatic foot showed a reduction in A δ -fiber density of 64%