Study of Strength-Duration Properties and iMAX in CMT1A patients

Maëlle TYBERGHEIN
Florence MANTO
François WANG
Antoine PANELLI

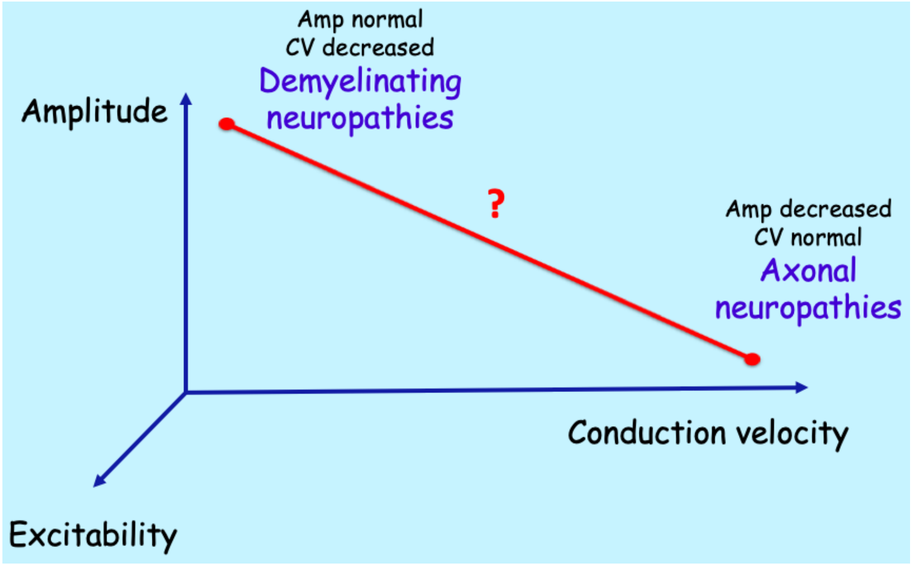
INTRODUCTION

Utility of Axonal Excitability in Electrodiagnostic (EDX)

- **Current Practice:**
  - Diagnostic are primarily based on changes in amplitude or conduction velocity.
  - Classifications will typically label neuropathies as **axonal** or **demyelinating.**

- **Limitations in Conventional EDX:**
  - Simplistic categorization does not fully capture the complexities of neuropathies.

- **Need for Improved Characterization**:
  - Excitability studies in peripheral neuropathies could help to understand the behavior of axonal membranes
  - More precise identification of pathophysiological mechanisms
  - Can lead to better patient management and treatment strategies.



F Wang

INTRODUCTION

Propriétés d'excitabilité axonale dans la maladie de Charcot-Marie-Tooth de type 1A

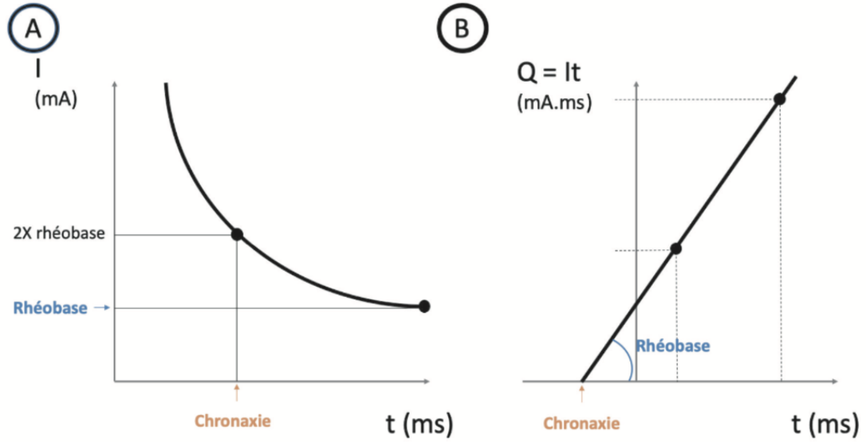
Tyberghein M, Manto F, Wang FC
Centre Hospitalier Universitaire de Liège, Belgique

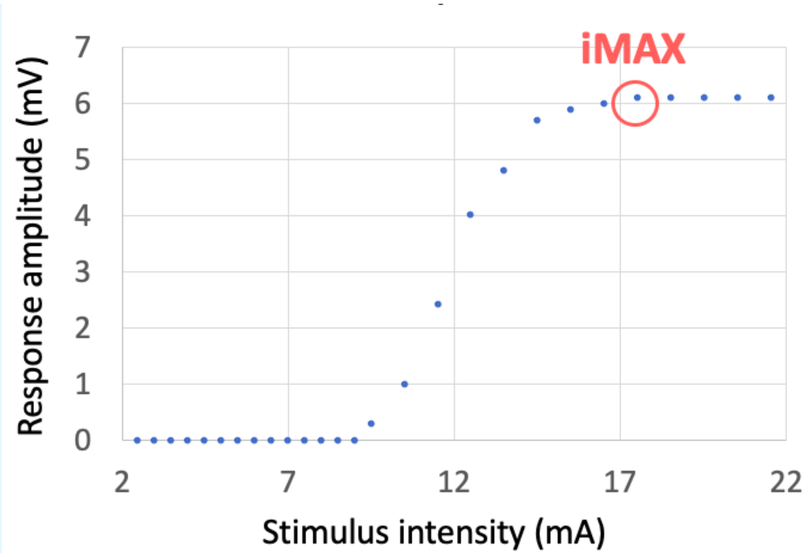
In the study:

Two key excitability parameters using conventional EDX machine in healthy individuals and CMT1A patients: **strength-duration properties** and **iMAX**.

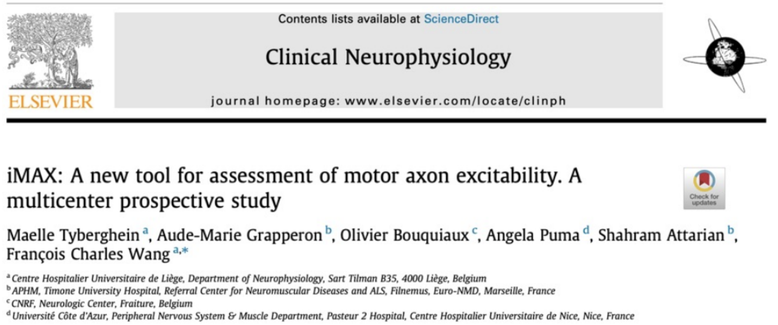
Strength-duration relationship: allows for the determination of two main parameters: first, the **rheobase**, the minimum current required to generate a response (with a preset amplitude) using a stimulus of "infinite" duration (1 ms); and second, the **chronaxie**, the time needed to evoke the target response with a stimulus whose intensity is twice the rheobase.

iMAX: represents the minimal intensity needed to produce a maximal compound muscle action potential (CMAP).





MATERIAL AND METHODS



Contents lists available at ScienceDirect

Clinical Neurophysiology

journal homepage: www.elsevier.com/locate/clinph

iMAX: A new tool for assessment of motor axon excitability. A multicenter prospective study

Maelle Tyberghein [a], Aude-Marie Grapperon [b], Olivier Bouquiaux [c], Angela Puma [d], Shahram Attarian [b], François Charles Wang [a,*]

[a] Centre Hospitalier Universitaire de Liège, Department of Neurophysiology, Sart Tilman B35, 4000 Liège, Belgium
[b] APHM, Timone University Hospital, Referral Center for Neuromuscular Diseases and ALS, Filnemus, Euro-NMD, Marseille, France
[c] CNRF, Neurologic Center, Fraiture, Belgium
[d] Université Côte d'Azur, Peripheral Nervous System & Muscle Department, Pasteur 2 Hospital, Centre Hospitalier Universitaire de Nice, Nice, France

- 50 healthy volunteers
- 14 patients diagnosed with CMT1A.

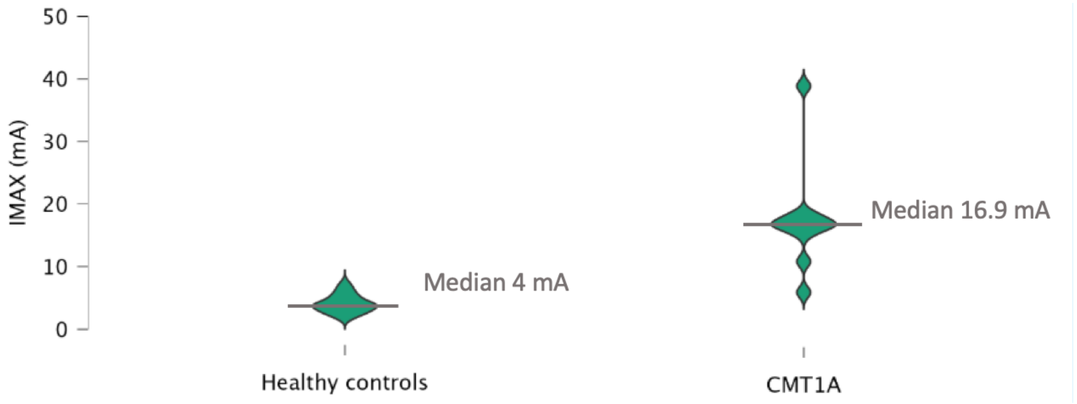
- Using EDX conventional machine

- iMAX measurements by determining **the minimal current required to evoke a maximal CMAP.**
- Study of the strength-duration properties by calculating the **rheobase** and **chronaxie**

- **Median motor nerve.**

RESULTS

iMAX and **rheobase** were significantly increased in CMT1A compared to healthy volunteers ($p<0.0001$).
Chronaxie is significantly reduced at threshold and shows a non-significant trend toward reduction at i40, compared to healthy volunteers.



	Controls (n=50)	*CMT1A (n=14)*	*p-value*
Rheobase threshold (mA)	2.20	4.87	<0,01
Rheobase i40 (mA)	2.83	11.12	<0,01
Chronaxie threshold (ms)	0.45	0.31	<0,01
Chronaxie i40 (ms)	0.47	0.42	0.46

Median CMT1A: 16.9 mA
Median Healty volunteers: 4mA

DISCUSSION - CONCLUSION

Charcot-Marie-Tooth Disease Type 1A (CMT1A)

- **Overview**: Most common inherited peripheral neuropathy
  - **Cause**: Duplication of PMP22 gene → demyelination of peripheral nerves
  - **Manifestations:** slower nerve conduction, muscle weakness, sensory deficits

- **Key Findings**:

- **Higher iMAX & Rheobase** in CMT1A: confirms and objectively supports the clinical impression that more current is needed to evoke motor responses. These results are attributed to changes in the passive properties of axonal membranes due to demyelination, resulting in current leakage.

- **Significant reduction of chronaxie at threshold and non-statistically significant trend toward reduced chronaxie at i40:** could be related to the lower density of permanent nodal sodium channels due to demyelination by increasing the nodal surface area.

Thanks for your attention!