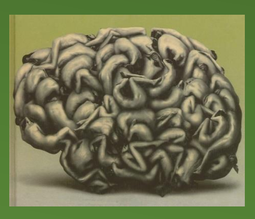
Functional magnetic resonance imaging by using laser stimoli in Charcot-Marie-Tooth syndrome: a case study



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Objective

Charcot-Marie-Tooth (CMT) disease is a genetically heterogeneous group of disorders. Pain is a less common symptom complained by CMT patients [1]. In this study we evaluated the cortical activations in a patient with congenital neuropathy to observe possible variations of pain matrix area.

Materials and Methods

We described the case of a 39-year-old male patient affect by CMT syndrome compared with one sex age matched healthy subject (HS). The nociceptive system was evaluated by Laser Evoked Potentials (LEPs). Moreover, a 3,0 T Magnetic Resonance Imaging (MRI) and fMRI (functional Magnetic Resonance Imaging) by using laser stimuli were performed (Fig. 1). Laser stimuli were applied at dorsum of feet and hands by using a Nd:YAP laser device. Stimulus intensity was set up at 2.0J, slightly shifting the stimulation point in order to avoid stimuli habituation and skin lesions. A high-field fMRI and muscular MR were performed. We constructed 10 blocks of rest and tasks respectively, each consisting of 5 volumes. Statistical analysis was performed by using the GLM. All analyzes were corrected by Bonferroni ($p < 0.05$).

Fig. 1 Devices: A) 3.0 T Magnetic Resonance Imaging; B) Stimul 1340 Nd:YAP

Results

The LEP examination showed an increase of latency and an amplitude reduction respect to HS (Fig. 2). The stimulation of upper and lower limbs produced more signifincantly activation in insula ($p<0.05$ correct) and anterior cingulate cortex ($p<0.05$ correct) respect to HS (Fig. 3). Moreover, the muscular MRI highlighted atrophy of the posterior compartment of lower limbs. The muscular echography showed an iper-echogenicity and diffuse and inhomogeneous alterations of the lower limbs muscles.

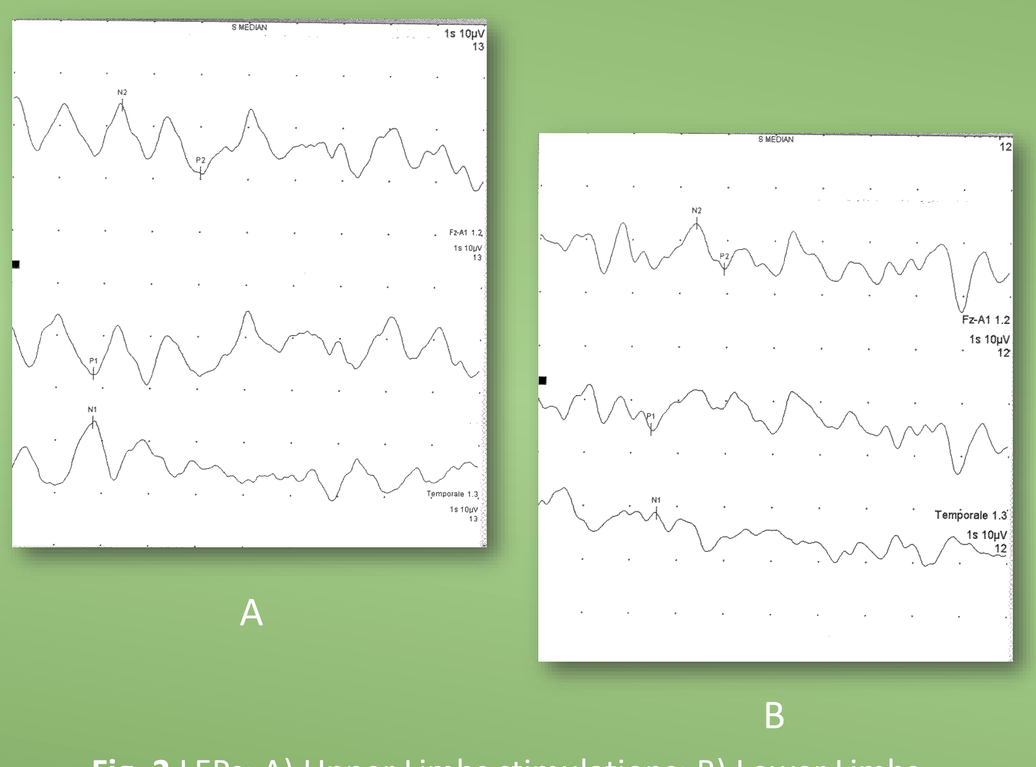

Fig. 2 LEPs: A) Upper Limbs stimulations; B) Lower Limbs stimulations

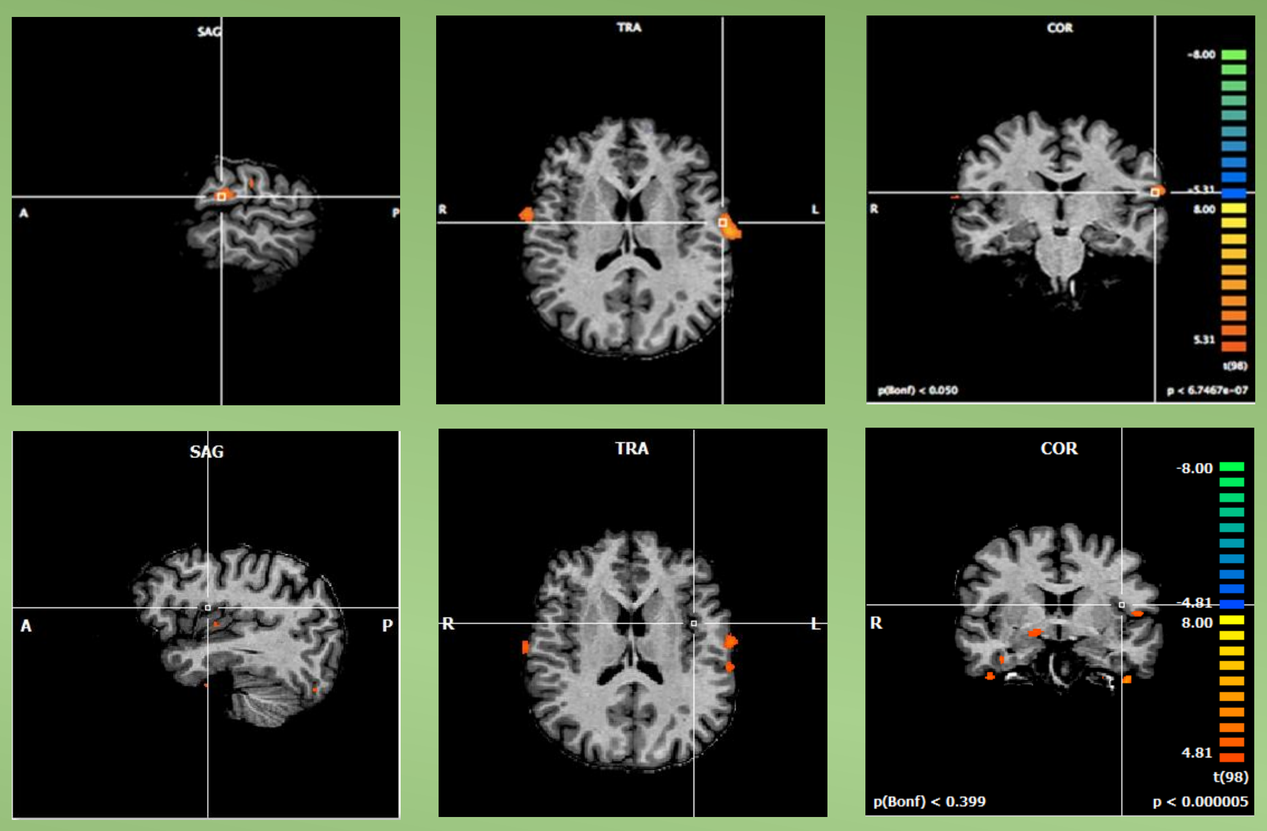

Fig. 3 fMRI: Cortical activations during upper and lower limbs stimulations

Discussion/Conclusion

The insula is a region known to play a role in the representation of bodily states, and the region comprising the posterior insula and anterior cingulate constitutes a functional area containing networks devoted to pain processing [2]. Several activations could be due to a major sensibility of nociceptive system in CMT patient. In our case, the amplitude N2/P2 amplitude to foot stimulation was reduce than that in HS, suggesting an Aδ-fiber impairment in this neuropathy.

In our patient, a reduced N2/P2 LEP amplitude suggests an Aδ -fiber decrease, mainly involving the longer axons of the lower limbs. Moreover, our results showed that the Aδ -axonal loss represents a risk factor for developing neuropathic pain. On the other hand, it is known that neuropathic pain can be sustained by a peripheral sensitization with mechanism by which nervous fibers develop spontaneous activity, lowered activation threshold, ectopic neural firing along the nerve. These further mechanisms cannot be excluded in our patient.

References

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2. Garcia-Larrea L, Perchet C, Creac'h C, Convers P, Peyron R, Laurent B, MauguiÃ¨re F, Magnin M. Operculo-insular pain (parasylvian pain): a distinct central pain syndrome. Brain 2010;133:2528-2539.

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