

**Deep brain stimulation for medically refractory tremor in Wilson's disease: a single case and review of the literature**

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A 41-years-old woman was referred to our outpatient clinic for a five year slowly progressive onset of speech impairment and tremor involving the arms and head. She had previously received a genetically confirmed diagnosis of Wilson's disease (WD) at the age of three years. At the admission neurological examination showed: a) action and postural tremor of the upper limbs (left more than right); its amplitude increased with a longer duration of posture (i.e wing-beating tremor); b) horizontal head tremor (i.e. "no-no"); c) voice tremor (video 1). Brain T2-MRI showed the typical "face of the giant panda sign" ( i.e. normal intensity of red nuclei and lateral portions of substantia nigra pars reticulata with high signal intensity of tegmentum and hypointensity of the superior colliculus) with no other evidence of basal ganglia structural changes.

Because of first line (primidone and propranolol) and second line (topiramate, pregabalin, clonazepam) therapy agents for tremor were unsuccessful, a surgical approach was proposed; the patient underwent to Deep Brain Stimulation (DBS) of ventral intermediate (Vim) thalamic nucleus. High stimulation output led to a significant reduction of her tremor amplitude of both arms and head; speech impairment remained stable and no potential DBS-related side effects were reported.

Vim has emerged as the most effective and established target for medically refractory tremor in patients with essential tremor (ET) [1]. Even though the pathophysiology of tremor is different between WD and ET our experience and published evidence support the potential role of Vim DBS as an effective and safe approach in carefully selected WD patients, although the presence of structural changes in the basal ganglia may limit the therapeutic success of the surgical procedure [2].

**References:**

[1] Krack P, Volkmann J, Tinkhauser G, Deuschl G. Deep Brain Stimulation in Movement Disorders: From Experimental Surgery to Evidence-Based Therapy. *Mov Disord.* 2019.

[2] Hedera P. Treatment of Wilson's disease motor complications with deep brain stimulation. *Ann N Y Acad Sci.* 2014.