IS DBS BETTER THAN BOTULINUM TOXIN IN PRIMARY DYSTONIA – NO!!

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Primary dystonias are chronic and often disabling conditions with a widespread spectrum of presentation mainly in young people.Primary dystonias are classified as pure dystonia, dystonia plus or paroxysmal dystonia syndromes.

Botulinum toxin has been used in treatment of primary dystonia for a longtime. In 1996, in a comparative prospective, double-blind trial (Brans JW, Lindeboom R et al) Botulinum toxin type A was reported to be significantly more effective than trihexyphenidyl in the treatment of cervical dystonia. Longterm followup of 100 patients (Haussermann P, Marczoch S, et al 2003) on botulinum toxin therapy was reported to be effective in approximately 60% over more than 10 years. Only 3% of patients were secondary nonresponders.The safety and efficacy of botulinum toxin for the treatment of focal hand and cranial dystonias are well-established. Studies of these adult-onset focal dystonias reveal both shared features, such as the dystonic phenotype of muscle hyperactivity and overflow muscle contraction and divergent features, such as task specificity in focal hand dystonia which is not a common feature of cranial dystonia. The physiologic effects of botulinum toxin in these 2 disorders also show both similarities and differences.

Key studies of the physiology of dystonia have focused on focal hand dystonia as it offers several advantages over other forms of dystonia. First, hand cortical sensory and motor representations can be readily identified on brain imaging and selectively targeted for such techniques as transcranial magnetic stimulation (TMS). The task specificity of focal hand dystonias permits separation of dystonic movements, hand movements that do not elicit dystonia and resting conditions. Finally, the frequently unilateral nature of focal hand dystonia provides an unaffected hand for comparison. Multiple research methodologies have demonstrated a loss of various types of inhibition in focal hand dystonia, including impaired reciprocal, intracortical, surround and interhemispheric inhibition.

Botulinum toxin blocks cholinergic nerve endings in muscle spindles as well as at the neuromuscular junction, demonstrated by the fact that botulinum toxin can lessen the tonic vibration response (TVR) in patients with limb dystonia. Modulation of spindle afferent input to the central nervous system likely contributes to botulinum toxin's efficacy in dystonia and it has been suggested the central effects of the toxin are largely explainable by the toxin's modulation of peripheral afferent input to the central nervous system. Motor cortex plasticity can be assessed by paired associative stimuli (PAS) in which peripheral nerve and transcranial magnetic stimulation of the contralateral motor cortex combine to alter the excitability of the motor cortex. Using TMS motor evoked potentials of the hand as a measure of motor cortical excitability, patients with cervical dystonia in combination with hand dystonia or dystonic tremor were found to have increased PAS-induced excitability. One month after botulinum toxin injections to the neck muscles only, PAS no longer facilitated the motor evoked potential response in hand muscles. Abnormal excitability returned as the clinical effects of the toxin wore off 3 months later. Botulinum toxin was found to reverse white matter abnormalities detected by diffusion tensor imaging in cervical dystonia.

As regards DBS, patients with dystonia are usually implanted at a younger age compared with parkinsonian and essential tremor patients, underlining the importance of long-term outcomes and possible interaction with disease progression. In addition, the long-term management of dystonia patients with DBS can be complicated by a number of problems mostly related to the implanted hardware and stimulation settings. Symptom exacerbation after initial benefit has been reported in a few cases. It is not known whether this is related to potential tolerance or habituation to stimulation or to progression of the underlying disease. Moreover, DBS has been shown not to alter the abnormal physiology in dystonia.