

Successful and Safe Use of Botulinum Toxin A in a Patient with Subclinical Myasthenia Gravis and Lingual/Brachial-Manual Dystonia

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Myasthenia gravis (MyGr) receptor antibody titers (AChR Ab) may be elevated in patients with cranial manual dystonia (Duane & Stuart, *Mov. Disord* 1999). This white female had late childhood onset right brachial-manual dystonia and at age 36 years developed lingual dystonic symptoms with fluctuating hyperkinetic dysarthria. Rest improved speech transiently. Two relatives have similar symptoms. AChR Ab titers were elevated and single fiber EMG revealed jitter in tongue and extremity muscles. Steroid and later plasmapheresis therapy at another center had no effect on patient symptoms. After separate trials of oral anticholinergics, baclofen, gabapentin, L-Dopa and lithium were unsuccessful, frontalis muscle injections of botulinum toxin A were applied. 10 units (Botox®) produced minimal effects but 20 units four weeks later produced isolated unilateral frontalis weakness. Subsequently two tongue sites at 10 units each produced minor improvement without excessive weakness or dysphagia. Six weeks later 4 tongue sites @ 12.5 units each (50 IU total) were well tolerated and produced brief modest improvement in motor speech. This form of subclinical My.Gr. may not be an absolute contraindication to botulinum toxin therapy for dystonia. However, symptom relief was suboptimal and generalization can not be made to typical My.Gr.

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