Treatment of Task Specific Upper Limb Dystonia using Ultrasound-guided Botulinum Toxin Injection

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Introduction and aims: Focal task specific dystonia (FTSD) is a movement disorder that has been described in medical literature since the 19th century.(1,2) It presents with "sustained muscle contractions, frequently causing twisting and repetitive movements".(3) FTSD most commonly affects the upper limb and is often initiated by voluntary movements such as writing, leading to the term graphospasm, or more commonly-used 'writer's cramp'. The precise pathophysiology of the disorder is poorly-understood but electromyographic (EMG) studies demonstrate co-contraction of antagonist muscles.(5) Botulinum toxin (BTx) is an effective treatment, it selectively blocks cholinergic transmission resulting in muscle paralysis or weakening, so-called 'chemodenervation' (figure 1). We provide ultrasound-guided injections at a joint neurology-radiology clinic, patients with dystonia are assessed clinically then examined with US. Under ultrasound (US) guidance BTx is selectively targeted to the muscle felt responsible for symptoms. BTx is reported to be effective for several months, therefore patients normally return to clinic every 4-6 months. We present the feedback provided by patients on their return and highlight potential complications.
The primary aims are to evaluate the effect of treatment in terms of patient-reported improvement in symptoms and also to evaluate patient-reported side effects. Secondary aims were to evaluate the optimum "Dysport" Botulinum toxin A doses for individual forearm muscles and to establish the average number of treatment episodes required to achieve a satisfactory treatment regime.

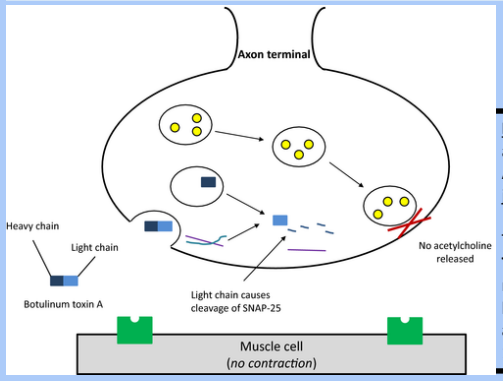

Figure 1 – The mode of action of Botulinum Toxin A at the neuromuscular junction

Taken from **Wong & Tincelli.** Management of refractory overactive bladder. The obstetrician and gynaecologist. 2016. 18 (3) 173-181

Methods: A combination of clinical evaluation and ultrasound (US) of the muscles, looking for abnormal uncoordinated movement within individual muscles, was used to determine which muscles to inject. Ultrasound was then used to guide injection of intramuscular Dysport A botulinum toxin (figure 2).
Patients return for further assessment once symptoms have returned and the next treatment is adjusted based on patient feedback using a 4 point scoring system (see results). We retrospectively analysed the assessment and treatment of all patients attending with upper limb FTSD over a 17 year period. 15 patients underwent a total of 124 treatment episodes. Feedback was available for 108 episodes. (Feedback was usually not available for the most recent episode as the patient had not yet returned).
The following information was extracted from the electronic patient records: patient demographics; muscles determined to be abnormally active; Dysport A botulinum toxin dose; patient score after each episode; complications reported; total number of treatments per patient. We assessed the number of visits required to achieve a satisfactory treatment regime, and extracted data for botulinum dosage and muscle injected when this was achieved.

Results:
15 patients (7F :8M), mean age at first treatment of 48 years (range 30-63).

Total number of treatment episodes: 124 (range 1-22 episodes per patient). Abnormal muscle movement was visible on ultrasound on 31% of patients.
On average it took 2.7 treatment episodes (range 1-4) to optimise treatment regime in terms of muscle selection and BTx dose. Optimisation did not occur for 2 patients who withdrew from treatment after 1 and 2 treatment episodes respectively.

Mean patient feedback score once optimal site and dose of injection established:
10 patients - good or very good improvement
3 patients - mild or moderate improvement
2 patients - no improvement

Patient scores following the initial adjustment phase there were 75 optimised treatment episodes: Good/very good improvement: 47 episodes (63%)
Mild/moderate improvement: 21 episodes (28%)
No effect: 7 episodes (9%).
Worse: 0 episodes (0%)
Complications: 4 episodes of initial too much weakness of muscle, lasting between 4-8 weeks before resolving. No cases of allergy or adverse drug reaction were found.

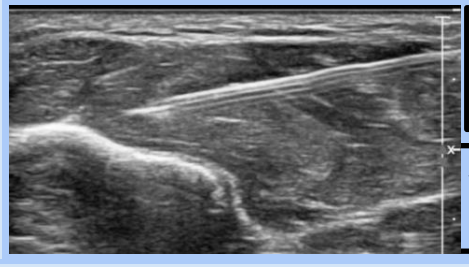

Figure 2 – Ultrasound demonstrating injection needle within extensor digitorum longus muscle

Indication	Muscle (* abnormal muscle activity seen on US)	Dose Botulinum toxin A (units)
Complex forearm movement/tremor	Supinator*	75-100
Graphospasm	FPL	10
Graphospasm	Adductor pollicus	10
Graphospasm	FCU	20-35
Wrist/ digit flexion/ subluxation	FDS* FCU*	20-30 40-50
Graphospasm	FCU FCR	15 15
Graphospasm	FPL*	20-35
Dystonic tremor	Pronator teres	50-60
Graphospasm	FCU*	150
Graphospasm	EPL	30
Graphospasm	FDS FDP	20 20
Graphospasm	FDP	7.5-10
Graphospasm	EDL	25

Table 1 – Indication, muscles targeted (+/-prescence of abnormal muscle activity *) and dose of toxin required for optimal effect in 13 patients

Discussion: Clinical guidance of injections uses anatomical landmarks and examination while moving a joint. While this technique is considered adequate in large muscles, it is more challenging in peripheral muscles.(6) EMG is shown to be superior to clinical guidance of BTx injection in all but the largest (gastrocneumius) muscle when leg and arm muscles were targeted in children with cerebral palsy.(7) EMG however, requires a degree of patient compliance and can be painful due to the size of electrodes and needles used. Alternative reported methods include injection of botulinum mixed with contrast medium and imaging with either fluoroscopy or cross-sectional imaging thereafter.(6)
Ultrasound provides a painless, radiation-free modality to localise muscles. Significant differences in injection location were found when comparing clinical examination using anatomical landmarks to ultrasound guidance when treating spasticity of small muscles of the forearm.(8) Injection needles can be directed accurately under ultrasound guidance and real-time imaging allows localisation of adjacent structures including nerves and blood vessels. Visualisation of the injectate entering the muscle is also beneficial given that the Botulinum staying within the muscle fascia is important to the duration of effectiveness.(8) Ultrasound has been shown to be superior to clinical examination in small studies assessing injection in the forearm or leg in post-stroke spasticity. (9, 10)
Our study further supports ultrasound as a method of muscle localisation in FTSD with 63% of injections reported as providing very good or good improvement and fewer than 10% providing no effect. The complication rate is likely to be reduced by ultrasound guidance providing additional anatomical information regarding surrounding structures. The complication rate in our study is low at 4%, this related to too high a dose being administered which could be rectified by adjustments at subsequent injections. Indeed, it has been suggested that effective targeting of BTx with ultrasound in FTSD and appreciating the injectate being administered specifically within the target muscle may help reduce the dose required.(6) We feel the fact that the majority of patients continue to attend for treatment for up to 22 episodes is an indication of patient satisfaction, with large numbers of good responses being reported. The 2 patients who decided to withdraw from treatment, both did so before reaching the mean number of visits shown to be required for optimising treatment. Given the condition is notoriously difficult to treat, with no satisfactory alternative to BTx injection, these results indicate a successful treatment method.
This is a retrospective study of a single treatment of focal dystonia. The scoring system used has not been validated. It was adopted as a pragmatic solution because dystonia is often a complex and subtle condition with individual effects on each patient. Patients usually have difficulty putting a description to their problem. The scoring used relates directly to the patient's perception of the treatment on their individual clinical problem. Altering this is the primary aim of treatment.

References: **1. Karp & Alter K.** Toxins. 2017 Dec 29;10(1):20. **2. Pearce JMS.** Journal of Neurology, Neurosurgery & Psychiatry. 2005 Apr 1;76(4):513–513. **3. Albanese et al.** Mov Disord. 2013 Jun 15;28(7):863–73. **4. Schuele et al.** Neurology. 2005 Jan 25;64(2):341–3. **5. Lin et al.** Journal of Hand Therapy. 2009 Apr;22(2):109–14. **6. Walter & Dressler.** Expert Review of Neurotherapeutics. 2014 Aug;14(8):923–36. **7. Chin et al.** J Pediatr Orthop. 2005 Jun;25(3):286–91. **8. Henzel et al.** PM&R. 2010 Jul;2(7):642–6. **9. Picelli et al** Clin Rehabil 2014;28(3):232-42 **10. Picelli et al.** Am J Phys Med Rehabil 2012;91(11): 957-64

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