extra-junctional acetylcholine receptors be secondary to regeneration of new motor end effective for three to four months after injection release of Ach for several days means the onset of unit. There is some evidence suggesting BoNT may junction, decreasing contraction of the motor unit. There is some evidence suggesting BoNT may also modify sensory and proprioceptive input via an effect on central motor neurons. Continued release of ACh for several days means the onset of effect may be two to four days. From a therapeutic point of view, treatment generally remains effective for three to four months, after injection and restoration of muscle activity is thought to be secondary to regeneration of new motor end plates, reduced levels of acetylcholinesterase and extra-junctional acetylcholine receptors. There are currently four neurotoxin formulations available in New Zealand: Botox (Allergan), AbobotulinumtoxinA (Dysport, Ipsen), IncobotulinumtoxinA (Baxilum, Merz) and RimabotulinumtoxinB (Myobloc, Solstice). Although BoNT is a foreign protein, secondary and response with production of antibodies against botulinum toxin A is extremely rare. There is very little evidence of tachyphylaxis. Botulinum toxin units are defined by mouse toxicity trials and while comparative efficacy is fairly similar in regards to potency, speed of onset, therapeutic duration and immunogenicity, there is no true “equivalent” dosing paradigm between different BoNT preparations. The lethal dose of Botox is estimated to be 2500-3000 units for a 70kg person. Typical perioral treatments are in the order of 20-50 units.

Facial dystonias Benign essential blepharospasm and hemifacial spasm are two of the most common disorders affecting the face. These facial dystonias can render patients functionally blind and adversely affect quality of life, causing depression, anxiety, ocular pain and difficulties with driving.

Benign essential blepharospasm (BEP) is a bilateral pattern of focal dystonia involving the muscles surrounding the eyes that manifests clinically as sustained spasm of the eyelid protractors resulting in excessive blinking, photophobia and persistent eye closure in the absence of any adnexal cause. BEP affects 32/100,000 people, typically in the fifth to sixth decade of life; affecting women more frequently than men in a 3:1 ratio. The aetiology is unknown.

A subset of blepharospasm patients display forceful contraction of jaw and tongue, as well as chin thrusting, consistent with focal cervical dystonia, a combination commonly described as cranial dystonia or “Meige syndrome”.

Hemifacial spasm (HFS) is characterised by involuntary intermittent sustained contractions of the muscles of facial expression supplied by the facial nerve. Hemifacial spasm commonly is of insidious or subacute onset, peaking in middle age, although it can occur anywhere from the third to seventh decade of life but is less common than BEP. Women are affected more frequently than men, 2:1. It is usually chronic in nature and may cause patients significant social disability.

The most commonly identified aetiology is vascular compression of the fifth cranial nerve around the Meckel’s cave, which may cause compression and ischemia of the cranial nerve, and may cause patients significant social disability. The most commonly identified aetiology is vascular compression of the fifth cranial nerve around the Meckel’s cave, which may cause compression and ischemia of the cranial nerve.

Neurotherapeutical treatments are available but patients may elect for a less invasive treatment modality.

High-level evidence exists for the treatment of facial dystonias with BoNT and clinical response rates are reported to be greater than 90% and clinically correlate with improved quality of life. Studies show persisting efficacy over a 15-year period in patients treated for BEB, HFS and Meige syndrome, with very low incidence of adverse effects. The large duration of effect and dose required for treatment remains unchanged in long-term follow-up studies, once an effective dose has been established by titration.

Subcutaneous injections into orbicularis oculi of upper and lower lids and the corrugator supercilii muscles of the upper eyelid are performed. Injections with blepharospasm are typically injected in three to four sessions while those with hemifacial spasm every four to six months.

Efficacy can be evaluated two to four weeks after initial injection and the sites and doses titrated to respond for subsequent injections. The clinical safety of BoNT injections has been well established in large series studies and, thankfully, most adverse events are minor and self-limiting, occurring in around 5-10% of cases. The titration period carries a risk of both under and overtreatment, but this reduces as the effective treatment regimen is identified. Bruising, swelling and haematoma formation are the most common adverse events. Injections into perioral area can cause blepharochalasis, diplopia, lower-lid malpositions including entropion and ecrropion. Excessive weakening of orbicularis can lead to lagophthalmos and corneal epitheliopathy, making artificial lubricants essential in the early post-injection period. An early review after initial treatment is usually prudent. The injection technique is extremely important in minimising side effects and maximising therapeutic effect.

Summary BoNT has revolutionised treatment of perioral facial movement disorders, particularly the extensive surgery that used to be patients’ only treatment option. The increasing use of BoNT for cosmetic reasons risks being underused and it should be considered as a possible cause in all patients seen with ocular and facial dystonias. Fortunately, such side effects are almost always transient, mild and respond well to conservative management.

References

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