The use of botulinum toxin in head and neck disorders

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Not only can botulinum toxin be used for cervical and laryngeal dystonia but in this article its use and complications in the management of oromandibular dystonia, Frey's syndrome, crocodile tears and drooling is described.

This short article will describe the use of botulinum toxin (BTX) in the management of oromandibular dystonia, a principal movement disorder affecting the tongue and jaw. BTX may also be used to reduce exocrine gland secretion, so conditions such as drooling, crocodile tears and Frey's syndrome may also be successfully controlled by the use of this neurotoxin.

DROOLING

Indications for the use of BTX

Drooling is largely a result of the inability of the oral musculature, including the tongue, to propel saliva from the mouth into the hypopharynx. The common groups of patients in which this occurs are those suffering from cerebral palsy, stroke, motor neurone disease and Parkinson's disease.

Saliva production can be reduced with the use of atropine-like drugs, such as hyoscine, which have become popular as a transdermal administration with a self-adhesive patch. Unfortunately, some patients may be allergic to the adhesive, requiring administration of the drug orally. Radiotherapy has also been used in the management of this condition, but there is a general reluctance to irradiate healthy glandular tissue in the young and middle aged.

BTX may be considered as a suitable initial treatment before considering surgical management by either submandibular gland duct re-routing or excision of the submandibular glands.

Method

Dysport (Ipsen, Slough, Bucks) may be administered by injecting 60 units directly into each submandibular gland. This will usually be successful in controlling drooling in the majority of patients, for anything up to 4 months. However, in some patients, drooling continues to be a problem, and therefore the parotid glands can be injected in addition to the submandibular gland. The bulk of the glandular tissue of the parotid lies between the ramus of the jaw and the mastoid process. Sixty units of Dysport may be delivered just underneath the external auditory canal – an injection administered at this point, between 1 and 1.5 cm deep, is unlikely to strike the facial nerve.

Complications

BTX may leak into the middle constrictor and cause atony of this muscle. The patient usually complains of an inability to initiate swallow, with accumulation of saliva in the back of the mouth, which may be aspirated. Many patients may already have their swallow compromised by the underlying medical condition, and so there is a distinct risk of aspiration occurring, particularly in those patients who have not had a percutaneous gastrostomy.

Long-term reduction of saliva flow predisposes to the development of dental caries and gingival disease. Therefore, care is needed in maintaining good oral hygiene.

FREY'S SYNDROME

Frey's syndrome or gustatory sweating occurs following salivary gland surgery or injury. The patient usually complains of preauricular sweating over the skin covering the parotid gland, triggered by the smell of cooking food, or when eating.

Many patients attempt to control the sweating with antiperspirants; however, these are often not particularly successful. Before considering a surgical treatment, such as tympanic neurectomy, management with BTX may be considered. This can usually control the gustatory sweating for approximately 4 months, and can be repeated as required.

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Method

It is often helpful to try and delineate the area of sweating by undertaking a starch iodine test. Starch iodine powder is dusted on to the affected area after which sweating is triggered by giving the patient some food to chew. As the sweat flows, the pale starch iodine powder turns a blue/black colour. The area of sweating is thus delineated, and a photograph may be taken, clearly showing the affected skin.

A 500-unit vial of Dysport is dissolved in 10 ml of normal saline, giving a concentration of 5 units of Dysport in 0.1 ml of solution. The affected skin is marked out in 1.5 cm squares, and subcutaneous injection of 0.15 ml of the solution (7.5 units of Dysport) is injected. This dose can be increased up to 15 units per square, but there is an increased risk of the skin sagging, as a result of paralysis of the adjacent platysma muscle.

Complications

Complications are rare. The commonest problem is inadequate treatment resulting from failure to map out the extent of the skin involved in the gustatory sweating process.

CROCODILE TEARS

Crocodile tears are a common sequelae to facial nerve injury or paralysis which has recovered. Oral medication is poorly tolerated and not desperately effective for managing this condition. Consequently, BTX has now become the mainstay in the management of this disorder.

Method

The lachrymal gland is located subcutaneously in the lateral third of the eyelid. It may extend underneath the orbital margin. The injection needs to be deep enough to penetrate the subcutaneous tissues, but obviously not so deep as to injure the underlying globe of the eye or extraocular muscles. Forty units of Dysport administered directly into the lachrymal gland will normally control this condition.

Complications

If the injection is too medially placed, it can leak into the levator palpebrae superioris muscle. This will lead to a ptosis. If the injection is placed too deeply, there is a risk of the tip of the needle penetrating the globe. The lateral rectus muscle may also be weakened or paralysed, leading to a complaint of double vision.

OROMANDIBULAR DYSTONIA

Oromandibular dystonia is a common dystonia (Duffey et al, 1998), accounting for approximately 8% of all types of dystonia. It is unusual for it to present in childhood and the peak age of onset is between the ages of 30 and 50 years. It is commonly present with other dystonias such as blepharospasm, laryngeal dystonia and torticollis.

Classification of oromandibular dystonia

Jaw-opening dystonia: In jaw-opening dystonia, symmetrical involuntary pulling of the jaw open is the commonest presentation. Asymmetric opening does occur, but is rare. The principal muscles involved are the lateral pterygoids, but in some patients spasm within the anterior belly of the digastric muscle may also be palpated. The patient presents with an inability to keep the mouth shut, and consequently speech is affected. The patient is much more distressed by the fact that they are unable to keep food in their mouth. As a result, patients will often refuse to eat with other members of the family, and are very reluctant to consider visiting friends and relatives, or going to restaurants.

Jaw-closing dystonia: The patient finds it difficult to open the mouth, or to keep the mouth open, as a result of spasm within the masseter muscles, or the vertical fibres of temporalis. It is usually a symmetrical dystonia and may be accompanied by bruxism and wear on the molar teeth. Chewing is often a slow process and as a consequence, although the patient may be happy to eat with relatives and friends, it takes much longer to complete a meal.

'Bunched tongue': In this form of tongue dystonia the tongue appears to be bunched up in the back of the mouth. It is often best seen on a lateral view during video fluoroscopy. The patient has a marked difficulty in protruding the tongue and may not even be able to get the tongue as far as the incisor teeth; consequently, speech is significantly affected. Overall, movements of the tongue are sluggish, and rapid alternating movements, such as moving the tongue backwards and forwards, or from side to side, are greatly reduced in both amplitude and speed. The voice may sound as if the person has a gobstopper or large sweet in their mouth.

Tongue-protruding dystonia: In tongue-protruding dystonia, the patient has great difficulties in keeping the tongue in the mouth. If the patient is asked to open their mouth and let their tongue do what it wishes, the tongue will gradually protrude from the mouth, and may protrude so far that the tip of the tongue reaches the tip of the chin. In everyday life, the patient has grave difficulties in keeping food in the mouth. Speech is often severely affected, as a

result of the tongue protruding between the front teeth, and making closure of the lips impossible.

Lip-closing dystonia: In lip-closing dystonia the orbicularis oris muscle goes into spasm, causing the lips to pucker up, as if to make a kiss. The patient often has difficulty in smiling and is unable to broaden the mouth.

Teeth-baring dystonia: In this variation, the patient has grave difficulties in covering the teeth with the lips. The angles of the mouth are often pulled back superiorly, so that the front upper incisor and canine teeth are all seen. In severe cases, the lower incisor teeth may also be permanently on view. In this type of grinning appearance there is considerable pressure on the teeth, particularly on the lower teeth. Over many years, this may lead to absorption of the gums and loss of the lower incisor teeth. Less commonly, there may be loss of the upper teeth as well, as a result of muscular pressure on the upper dentition.

Platysma: Prominent platysmal bands are often found in patients with oromandibular dystonia. They frequently stand out during speech and can easily be seen and palpated.

Assessment

When a new patient with oromandibular dystonia is referred, it is important that a full assessment is undertaken before treatment. In addition to taking a history, which would include the social difficulties caused by the dystonia and their effects on the patient's life, it is essential that video and auditory recording are undertaken. The author recommends videoing the patient face-on, using the following regimen:

- The patient is asked to sit quietly and look at the camera
- The patient is then asked to open their mouth and to hold the mouth open for as long as they can
- They are advised not to try and control the tongue movement, but to let the tongue do what it wishes
- Twenty seconds of videoing of the tongue at rest, with the mouth open is undertaken
- The patient is then asked to open and close the mouth three times
- The patient is then asked to move the jaw from side to side with the mouth open three times, and once again to move the jaw from side to side with the mouth closed three times
- Then with the mouth open, the patient is asked to protrude the tongue as far as they can
- They are asked to attempt to touch the tip of their nose with the tongue, and to touch the tip of their chin with the tongue
- Side-to-side movement of the tongue is then recorded
- The patient is then asked to say 't t t t t'; 'putaker putaker putaker'; 'k k k k k'
- When this exercise is completed the patient is recorded eating a biscuit
- Finally, in those patients who complain of difficulty swallowing, and particularly in the oral or pharyngeal stages of swallowing, a video fluoroscopy should be undertaken.

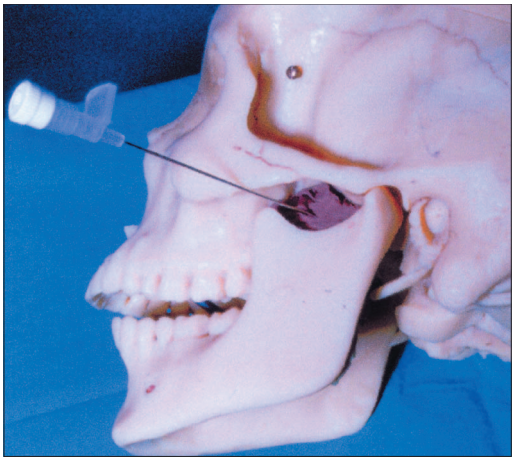
After completion of assessment most patients can usually be classified into a subgrouping depending on jaw movement, tongue movement, and lip movement. Some combinations are more common than others and jaw opening with lip retraction often go together. Jaw closing with the puckering of the lips is also a common combination.

Treatment

Jaw-opening dystonia: The lateral pterygoid muscle is best approached through the coronoid notch (*Figure 1*) (Hawthorne, 2002). A 2 cm, 23 gauge needle is used to inject 60 units of Dysport, and the needle is advanced up to the hilt in most patients. This injection can be painful, particularly if the tip of the needle comes into contact with the periosteum. In those patients where the anterior belly of the digastric muscle is palpable, a further 30 units of Dysport can be injected into this muscle. However, most patients are usually well controlled by limiting the injection to the lateral pterygoids.

Jaw-closing dystonia: In most patients, both temporalis and masseter muscles are involved in jaw-closing dystonia, although palpation of these muscles is recommended to confirm that this is the case. Occasionally it may be found that only one of these muscles is involved. In a typical adult, 80 units of Dysport are usually

Figure 1. Injection approach for the lateral pterygoid.



required to control the spasms of the masseter muscles, while 60 units are usually sufficient for the temporalis muscles.

Bunched tongue: In those adult patients with the obvious bunched tongue presentation a transcutaneous injection into the bulk of the tongue in the mid-line is recommended. On the first occasion, injection of 15 units of Dysport is recommended in adults. This can gradually be increased up to 50 units, but above 50 units side effects start to become more common. A 2 cm, 23 gauge needle is usually sufficient to carry out this injection.

Tongue-protruding dystonia: The author prefers a double injection technique in those suffering with tongue protrusion. This involves 10–15 units being delivered into the bulk of the muscle, in a transcutaneous injection in the mid-line, and a further 10–15 units being delivered into the anterior one-third of the tongue. This can be done as an oral injection in the mid-line, although the anterior third of the tongue can be approached from below, through the skin. Once again, the total dose into the tongue can be gradually increased to 60 units, although above 60 units it is common for complications and/or side effects to occur.

Lip-closing dystonia: Where the orbicularis oris is in spasm, injections in the 12 o'clock, 6 o'clock, 10 o'clock and 2 o'clock positions, of 10 units of Dysport, are usually effective in controlling the problem.

Teeth-baring dystonia: Careful assessment of the video is required in this condition. Often the levator anguli superiores and the depressor anguli superiores are involved. In addition, where there is considerable spasm of the orbicularis oris as well, pressing against the teeth, injections into this muscle may also be required. Each patient usually has to be assessed on an individual basis. However, a total dose of 40–60 units is often all that is required to manage the worst of the problem.

Platysma: When platysmal bands are present, injection of 20–40 units of Dysport into each band will often give relief and improve the overall cosmetic appearance of the patient.

COMPLICATIONS

Injections into the lateral pterygoid can occasionally cause problems with a velar incompetence. The patient becomes aware of a hypernasal speech, and may find that liquids come out of the nose while drinking. This is usually a dose-related complication and is unusual when the dose of Dysport is restricted to between 40 and 60 units injected into each lateral pterygoid.

Should the tongue become flaccid, the patient may find increasing difficulties with drooling, as they are unable to propel the saliva from their mouth into the hypopharynx. The patient may develop a different speech problem instead, having poor speech as a result of poor tongue control and a flaccid tongue. Gross overdosage should not occur, but if an excessive amount of toxin is inadvertently injected the tongue will loll into a position dependent on gravity. Consequently, it may prolapse into the hypopharynx, causing breathing difficulties on lying down, or may tend to fall out of the mouth on leaning forward with the mouth open. In these circumstances, tongue control may be a problem, although the author has never seen, and is unaware of, a total loss of airway as a result of excessive use of BTX within the tongue.

Injections into the facial muscles controlling lip movement often lead to an asymmetry of the lips and the patient may find a degree of flaccidity in the upper lip irritating. It may also affect plosive speech sounds. When the medial pterygoid is injected it is usually only required on one side because of asymmetric pulling of the jaw to one side. This rarely causes problems but hypotonia of the oropharynx may occur, leading to a difficulty in propelling a bolus of food into the oesophagus. This occurs more commonly when bilateral injections have been given. HM

Conflict of interest: none.

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KEY POINTS

- Botulinum toxin is an effective treatment, with few side effects, in the management of oromandibular dystonia, drooling, Frey's syndrome and crocodile tears.
- Complications are usually dose related, and are often a result of the toxin weakening the actions of adjacent muscles to the site of injection. As a rule, they are short-lived, as the toxin wears off over a period of weeks.
- Owing to a lack of systemic effects, or sedation, it is well tolerated by those patients with oromandibular dystonia.
- Many find it a more effective and well-tolerated treatment than oral medication with drugs such as tetrabenezine, benzhexol, gabapentin and clonazepam.