

CASE REPORT

An Orthodontic Appliance Used in Management of Lingual Focal Dystonia

HEATHER M. ILLING, BDS(Hons), MSC, FDS, MOrth RCS
STEPHEN N. ROBINSON, BDS, MSC, FDS RCPS, DOrth, MOrth RCS

A 52-year-old lawyer was referred by the speech therapy department in October 1997 due to his increasing difficulty in controlling extraneous movements of his tongue. The patient had been affected in 1994 by a focal dystonia of the right wrist and hand, with the latter showing repetitive muscular contractions. The tongue became involved in October 1996, causing a significant effect on the patient's speech and a socially embarrassing protrusion of the tongue. He was referred for speech therapy, but despite intensive instruction, further speech deterioration occurred.

The patient sought alternative forms of therapy, and in July 1997, hypnosis was induced, using progressive relaxation

techniques and post-hypnotic suggestions involving reduction of tongue movement and accurate targeting of articulation points. No immediate effects were observed.

The patient was then referred to a neurologist, and various drug regimens were attempted. Benzhexol, an antimuscarinic drug, produced the side effects of dry mouth, blurred vision, and mental confusion, which required discontinuation of treatment. Baclofen, a skeletal muscle relaxant, was then prescribed to inhibit transmission at the spinal level and depress the central nervous system. The patient suffered no side effects and felt that this drug "targeted" the tongue more effectively. The option of localized botulinum toxin

injections was discussed, but the risk of dysphagia was not acceptable to the patient.

The patient also tried several methods of controlling his protrusive tongue movements, one of the most successful of which was to hold chewing gum on the lingual aspect of the mandibular incisors. Therefore, the patient was referred to the orthodontic department for fabrication of a more robust training device for the tongue.

Orthodontic Appliance

An .028" removable wire "gate" was designed for placement lingual to the mandibular incisors (Fig. 1). The appliance was retained with Adams clasps on the first molars and ball

Dr. Illing is Senior Registrar in Orthodontics and Dr. Robinson is a Consultant Orthodontist, Maxillofacial Unit, Queen Alexandra Hospital, Cosham, Portsmouth PO6 3LY, England. Dr. Robinson is also a Consultant Orthodontist at St. Mary's Hospital, Isle of Wight, and in the private practice of orthodontics in Hampshire, England.



Dr. Illing



Dr. Robinson



Fig. 1 Removable mandibular appliance, with lingual “gate” embedded in acrylic to control tongue protrusion.

clasps in the anterior segment. Cold-cured acrylic was added incrementally over the “gate” until the patient felt his tongue movement was controlled. The appliance was then constructed with heat-cured acrylic.

The patient was seen two weeks after fitting of the appliance and again three months later. During this period, he wore the training plate nearly full-time, and the clarity and articulation of his speech improved noticeably. After another three months, his speech improved even more. Therefore, the patient’s dosage of Baclofen was reduced.

The patient currently uses the training device two to three days a week and in periods of tiredness, when he usually has more difficulty in controlling extraneous tongue movements.

Eventually, we expect to make a permanent appliance of cobalt chromium.

Discussion

Dystonias are a group of neurological disorders affecting about 20,000 people in Great Britain.¹ Although they were once thought to be due to psychological distress, the various syndromes of dystonia are now established as a result of abnormal brain function, usually in the basal ganglia. A cause for these movement disorders can be established in approximately 25% of cases, with inheritance playing a role in the remainder. In many patients with idiopathic dystonia, particularly with an onset in childhood and adolescence, the illness is inherited. Most cases of adult onset occur

sporadically.

At one time, the mode of inheritance was thought to be an autosomal recessive trait common among Ashkenazi Jews. Recent studies in the United States,² Israel,³ and Great Britain⁴ have disproved this theory. A survey of 100 British families showed 85% of dystonic disorders to be due to an autosomal dominant gene with approximately 40% penetration; there was no evidence of an autosomal recessive gene.⁴ The number of dystonic Jewish patients was greater than expected, but was considered to be due to a founder effect in eastern Europe. The patient shown in this article is of Jewish origin.

Dystonia is a syndrome of sustained muscle contractions, which frequently cause twisting and repetitive movements or ab-

normal postures. The effects are worsened by attempts to move. Dystonia can affect any part of the body, but typically starts in one area as a focal dystonia. The disease progresses to generalized dystonia, where virtually all of the body is affected, in 60% of children, 35% of adolescents, and 3% of adults.¹ Dystonia can be recognized clinically by observation of the movement disorder, and later confirmed by blood and urine tests; biopsy of bone marrow, muscle, or skin; and/or MRI of the brain.

Lingual dystonia is a common feature of oromandibular dystonia (OMD) that is manifested by lateral or upward deviation and protrusion of the tongue. The protrusive movements can be particularly disruptive, causing not only severe dysarthria, but also embarrassing problems with chewing as the tongue pushes food out of the mouth. The result can be loss of weight and poor nutrition. If the pharyngeal musculature is involved, dysarthria, dysphagia, dysphonia, respiratory difficulties, and involuntary vocalizations such as humming, grunting, belching, or gasping can occur.

OMD can be alleviated with various sensory inputs, including touching the lips or chin, chewing gum, and applying

pressure in the submental area.⁵ Symptoms of lingual dystonia are alleviated in about one-third of the cases with medications such as anticholinergic drugs,⁶ Baclofen, benzodiazepines, and tetrabenazine. Although botulinum toxin injections can benefit patients with focal dystonias, especially in later onset cases, the risk of dysphagia limits the usefulness of this therapy.

Conclusion

In the case shown here, a simple removable appliance was used in the management of lingual dystonia, in conjunction with more conventional forms of therapy. Our experience emphasizes the importance of multidisciplinary care of patients with this debilitating condition.

REFERENCES

1. Marsden, C.D. and Quinn, N.P.: The dystonias, *Br. Med. J.* 300:139-144, 1990.
2. Bressman, S.B.; de Leon, D.; Brin, M.F.; Risch, N.; Shale, H.; Burke, R.E.; Greene, P.E.; and Fahn, S.: Inheritance of idiopathic torsion dystonia among Ashkenazi Jews, *Adv. Neurol.* 50:45-56, 1988.
3. Zilber, N.; Corkzyn, A.D.; Kahana, E.; Fried, K.; and Alter, M.: Inheritance of idiopathic torsion dystonia amongst Jews, *J. Med. Genet.* 21:13-20, 1984.
4. Fletcher, N.A.; Harding, A.E.; and Marsden, C.D.: A genetic study of idiopathic torsion dystonia in the UK, *Brain* 113:379-395, 1990.
5. Cardoso, F. and Jankovic, J.: Oromandibular dystonia, in *Handbook of Dystonia*, ed. J.K.C. Tsui and D.B. Calne, Marcel Dekker Inc., New York, 1993.
6. Vogels, O.J.; Maassen, B.; Rotteveel, J.J.; and Merx, J.L.: Focal dystonia and speech impairment responding to anticholinergic therapy, *Pediat. Neurol.* 11:346-348, 1994.

CORRECTION

In the JCO Roundtable on Ethics in Orthodontic Practice, Part 6 (August 1999), Dr. David Kornbluth's response on p. 449 was misquoted. To the question, "Is it ethical to criticize the work of another orthodontist when you believe such criticism may be warranted?" his reply should have read, "I am not sure it is ethical without first contacting the other orthodontist and finding out the details of the case, but I do think you must refrain from criticizing the work of a fellow orthodontist to the patient or the parents."