Recent advances in the surgical treatment of dystonia

Dystonia is an interesting neurological disorder that continues to cause the clinician difficulties in formulating appropriate management strategies. Therapy is linked closely to the classification of dystonia and so the characterisation into aetiological sub-type and distribution of the condition should be established prior to devising any treatment plan. In a small minority of patients (eg. Wilson’s disease, doparesponsive dystonia (DRD)), specific treatment can be instituted but in the majority of cases therapy is symptomatic, directed at decreasing the intensity of the dystonic contractions. However a lack of knowledge relating to the underlying pathophysiology has hindered the discovery of effective pharmacological treatments for most forms of dystonia. Nevertheless because of the reversibility and responsiveness of DRD to L-dopa therapy, all patients with childhood onset dystonia should therefore be given an adequate trial of this drug. Unfortunately, treatment of dystonia with oral agents is otherwise generally unsatisfactory. For those with symptoms and signs unresponsive to levodopa, other oral medications, including anticholinergics, tetraabenazine, baclofen and benzodiazepines, may provide mild to moderate relief. More effective treatment exists for the focal dystonia in particular the use of botulinum toxin, although injections of toxin into the affected muscle groups tends only to produce transient relief and generally need to be repeated every 3-6 months. For patients with more widespread dystonia, or those with disease refractory to medical therapy or botulinum toxin injection, there appears now an increasing role for functional neurosurgical intervention.

Case Report: Idiopathic Torsion Dystonia

This 7 year old girl first began to exhibit features of dystonia at the age of 3 years. Her condition was progressive in nature to the point where at presentation, she was anarthric, fully dependent on her parents for care and in constant pain due to the dystonic contractions. At the time of surgery rating scale scores were 109/120 for movement and 14/29 for disability. She continued to improve and was able to communicate, attend school, walk unaided and remain continent.

The first recorded case of surgery for dystonia dates back to 1641 when the German Physician Minnius treated torticollis by sectioning the sternocleidomastoid muscle. The Russian surgeon Buyalsky (1850) appears to have performed the first spinal accessory nerve section for spasmodic torticollis followed by Morgan in 1867 and Collier in 1890. Spinal cord root section to treat spasmodic torticollis, involving unilateral section of the first three anterior cervical roots, was first proposed over a century ago by Keen (1891). This procedure of cervical rhizotomy was refined over the years by surgeons including Dandy in 1928 who combined intradural section of the cervical sensory and motor roots with accessory nerve section. By 1979 variations of this procedure were still considered the operation of choice for cervical dystonia refractory to medical therapy. However, long-term follow up has disputed the effectiveness of these techniques. The issue of long term efficacy, together with the high incidence of denervation related complications, has now led to the virtual abandonment of these procedures. Extensive muscle resections, microvascular decompression of the accessory nerve, peripheral facial neurctomy and cervical cord stimulation are further examples of procedures that have been used to treat dystonia but that have also fallen out of favour. Apart from intrathecal baclofen infusions, practically all the surgical methods for treating generalised dystonia, preceding the stereotactic era, have either been ineffective or of poor comparable benefit. This has consequently given rise to the replacement of these operations by functional stereotactic procedures for patients with dystonia.

The introduction of stereotactic surgery allowed Bertrand (1978) to combine thalamotomy and peripheral denervation with improved outcomes. Further development of stereotactic techniques coupled with satisfactory results encouraged its use by functional neurosurgeons who have attempted to treat dystonia by lesioning a variety of different deep brain...
Thalamic nuclei can produce favourable results in a number of different forms of dystonia. For example, Vercueil (2001) employed this technique in twelve patients with generalised dystonia resulting in a satisfactory outcome in five of the patients.

Because of the success of thalamotomy and neurophysiological evidence implicating the thalamus in the pathogenesis of dystonia, the pallidum was not initially the favoured target for DBS. There are only a few reports of the effects of pallidal stimulation in dystonia and these are mainly case reports or small case series. Although to date there do not appear to be any formal comparative studies of thalamic versus pallidal stimulation, there are several instances where patients with stimulators in both deep brain structures appear to have benefited more from pallidal rather than thalamic stimulation.

Present evidence favours the view that Gpi is superior to thalamic stimulation for primary and secondary dystonia and it would appear that DBS is one of the most effective means of alleviating dystonia. Generalised dystonia, particularly in those patients who are positive for the DYT1 gene, is the best indication followed by spasmodic torticollis, where respectively mean 70% and 40% improvements have been reported. Post-traumatic dystonias with visible brain lesions on imaging do not appear to respond well to DBS. Furthermore, it is also important to note that a feature of these dystonic conditions is that the response is gradual, manifesting as a progressive improvement in the condition over months to years. Experience gained from the patients treated by our group suggests that maximal or near maximal improvement occurs at about one year in patients with generalised dystonia. Those with spasmodic torticollis improved at a slower rate, gaining most benefit approximately two years post-surgery. Longer-term follow-up will be needed to confirm that these benefits are maintained and also to help ascertain what the optimal parameter settings are.

Further Reading

Correspondence Address
Mr. John Yianni1 and Prof. Tipu Aziz1,2
1The Oxford Movement Disorder Group, Department of Neurological Surgery, The Radcliffe Infirmary, Oxford
2University Department of Physiology, Oxford University, Oxford
E-mail: tipu.aziz@physiol.ox.ac.uk, Fax:01865224786

We would like to thank Medtronic for their sponsorship of this article.

Contact details are -
Mr. Clive Woodard, UK Manager - Activa Therapy, Medtronic (UK) Ltd, Suite One, Sherbourne House, Croxley Business Centre, Watford, Herts WD1 8YE
Tel: 01923 212213
Email: clive.woodard@medtronic.com