

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, dopa-responsive 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, tetrabenazine, 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 generalised dystonic spasms. Genetic analyses revealed that she was negative for the DYT1 gene. Medical therapy including L-dopa, benzhexol, clonazepam and botulinum toxin had not provided any long-lasting benefit. Her Fahn and Marsden dystonia rating scale scores were 109/120 for movement and 29/30 for disability. No changes were observed until stimulation was initiated one month after implantation

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Professor Aziz studied physiology at University College London graduating in 1978. During this time he developed his keen interest in the role of the basal ganglia in movement disorders. He studied medicine at King's College London (1978-1983) and obtained his surgical fellowship in 1987 following which he has pursued a career in neurosurgery. He is currently a consultant neurosurgeon at the Radcliffe Infirmary, Oxford and Charing Cross Hospital, London. He is an expert in functional neurosurgery and has a special interest in the surgical treatment of movement disorders including dystonia.



Mr. John Yianni trained at University College London, qualifying in 1996. He completed his basic surgical training in Oxford, obtaining his surgical membership in 1999. Subsequently he joined the Oxford Movement Disorder Group based at The Radcliffe Infirmary Oxford, where he is currently clinical research fellow working towards an MD. His field of interest includes stereotactic functional neurosurgery for movement disorders, in particular dystonia.

of bilateral electrodes into the posteroventral internal globus pallidus (GPi). She subsequently experienced gradual improvement in most aspects of dystonia. At 3 months her Fahn and Marsden rating scores had improved to 47/120 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 neurectomy 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.

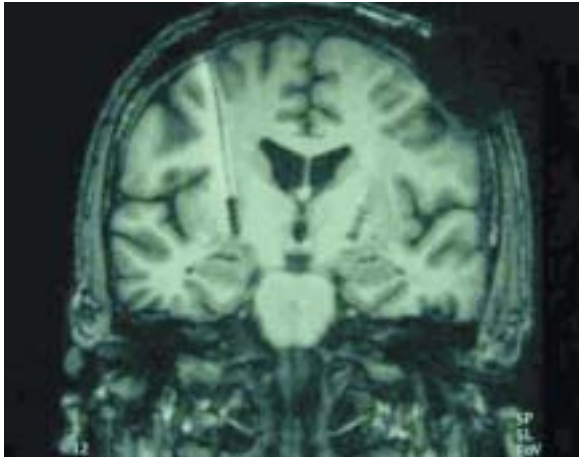
The introduction of stereotactic surgery allowed Bertrand (1978) to combine thalamotomy and peripheral denervation with improved outcomes. Further development of stereotactic techniques coupled with satisfactory results encouraged its use by functional neurosurgeons who have attempted to treat dystonia by lesioning a variety of different deep brain



Pre-op: Severe generalised dystonia with bulbar features. At the time of surgery she was bed-bound, doubly incontinent, anarthric and dysphagic.



Post-op: Some dystonic posturing of left foot and right hand remains. Speech improving and swallow normal. Mobilising independently and attending school.



Post-operative MRI of a patient following implantation of bilateral DBS electrodes.

structures including the internal capsule, cerebral peduncles, dentate nucleus, various basal ganglia and thalamic targets. As in other hyperkinesias, medial pallidotomy was the first stereotactic operation to be performed, initially for spasmodic torticollis in 1953 by Riechert followed by its use for generalised dystonia in 1957 by Cooper. Unfortunately in comparison with thalamic surgery only a few small series of pallidotomy for dystonia were published at that time. Hence by the 1960's, thalamotomy was emerging as the stereotactic procedure of choice. In 1976 Cooper published the results of thalamotomies that he had performed on over two hundred patients, reporting good or moderate improvement in 70% of his patients series. There was also some evidence that lesioning this target benefited patients with secondary dystonia, hemidystonia and tardive dystonia. However, in contrast to pallidotomy, the high incidence of postoperative dysarthria and dysphagia usually prevented surgeons from performing simultaneous bilateral thalamic surgery. Also, compared to Cooper's original series, subsequent studies from other centres have produced more variable and generally less impressive results. Consequently the ideal subcortical target for lesional surgery has remained the subject of much discussion although more recent evidence favours the medial pallidum.

The success of Deep Brain Stimulation (DBS), within the last few years, as treatment for a number of different movement disorders could soon see it as the first-line treatment for dystonia refractory to medical intervention. It has the advantages over lesional surgery of being reversible, adaptable and avoids concern about the effects of lesioning the developing brain in the case of children. DBS also allows bilateral surgery to be undertaken because of the reduced level of morbidity involved. As dystonic posturing may be very severe, DBS is usually performed under general anaesthesia for dystonia, unlike functional surgery for tremulous disorders, which usually occurs with the patient fully awake.

A new method for successfully treating spasmodic torticollis by implantation of stimulators into the thalamus was described by Mundinger in 1977. Since then, it has been demonstrated that targeting 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

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