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Dystonia: a surgeon's perspective

Tipu Z. Aziz*, Alexander L. Green

Department of Neurosurgery, The John Radcliffe Hospital, Oxford, UK

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SUMMARY

Surgery for dystonia has a history stretching back for centuries including myotomy and other procedures on the musculoskeletal system. In the last century lesional procedures, mainly involving the pallidum became popular. More recently, with the advent of deep brain stimulation, bilateral medial pallidal stimulation has become commonplace. This review describes the issues with patient selection, technical aspects of implantation and effects as well as complications of the technique. Some of the rarer types of dystonia that have also been treated with DBS are also described.

1. Introduction

Although dystonic conditions have been treated surgically for centuries e.g. myotomy for torticollis, and stereotactic lesional procedures have been used for decades (Fig. 1), the search for better, less hazardous therapies has continued. With experience of pallidotomy and the alleviation of dystonia that affected some parkinsonian patients [1] it became apparent that pallidal stimulation might be effective in dystonia. Since bilateral surgery for generalised dystonia was indicated [2], stimulation had the advantage of less side effects than lesions and at the time, high frequency stimulation was thought to mimic lesions [3].

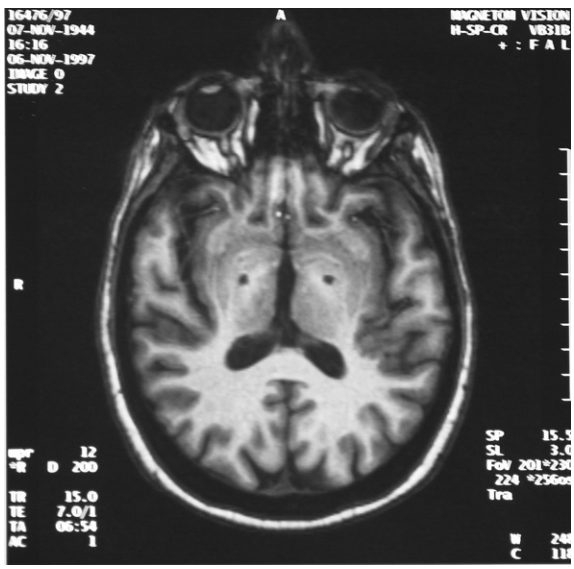

Fig. 1. Post-operative axial MRI scan showing bilateral pallidotomies (Globus Pallidus internus) in a patient with dystonia.

The earliest example of deep brain pallidal stimulation for dystonia was that reported by Mundinger in 1977 [4]. Since then, numerous case series have confirmed efficacy of the therapy and in the case of generalised dystonia, Class 1 evidence came in the form of a randomised controlled trial [5]. Currently there is no Class 1 evidence for spasmodic torticollis, but there are prospective controlled trials in the literature supporting the efficacy of this therapy in patients with failed medical management [6].

It is now known that not all patients benefit to the same degree with deep brain stimulation and degree of improvement may be multi-factorial [6]. It is by accumulating available literature of patient assessments and results that a better picture will emerge for accurate decision making. One would have to consider patient selection, surgical technique, choice of hardware implants, stimulation settings, avoidance of complications and possible avenues to manage non-responders.

2. Patient selection

One of the problems facing surgical decision making is the nebulous nature of the term categorised as *dystonia*. The condition may be primary, associated with a genetic predilection such as the DYT1 gene, secondary to a metabolic disorder such as PANK2, drug intake such as tardive dyskinesia, or secondary to brain injury. Dystonia can also be an additive deficit to another condition such as that which attends cerebral palsy or be task-specific.

In order to compare results across groups, it is essential to have standardised assessments such as the Burke, Fahn, Marsden rating scale to generalised dystonia [7], the TWSTRS scale for spasmodic torticollis [8] and the Abnormal Involuntary Movement scale for unusual disorders [9]. Task specific disorders are less amenable to rating scales but as for all patients, functional disability scales are also valuable. Given that neuropsychological abnormalities are

* Corresponding author. Tipu Z Aziz. Department of Neurosurgery, Nuffield Department of Neurosurgery, Level 3, The West Wing, The John Radcliffe Hospital, Headington, Oxford, OX3 9DU, UK.
E-mail address: tipu.aziz@nds.ox.ac.uk (T. Aziz).

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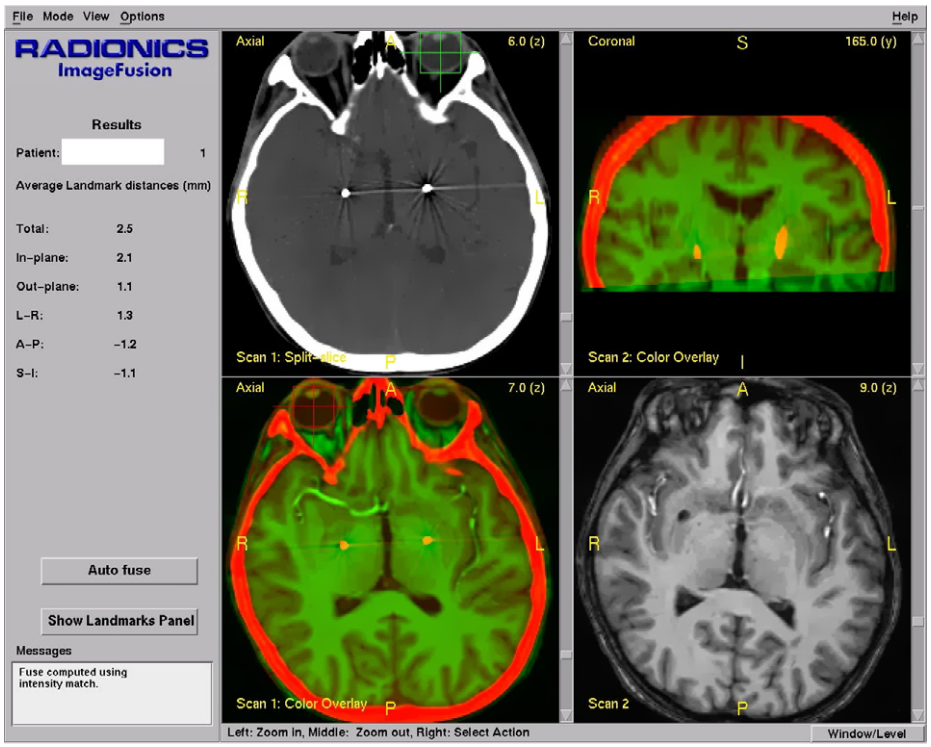

Fig. 2. Image fusion software showing post-operative CT scan fused with the pre-operative MRI scan. Electrodes are in the medial pallidum bilaterally.

common in dystonia [10] a full neuropsychological assessment is essential.

It is still not possible to select patients on firm evidence-based criteria, but different groups have shown that there are some apparently common predictors. It has been the experience of several groups that secondary dystonias have a lesser rate of response than primary dystonias [11,12]. In generalised dystonia, myoclonic jerks can respond over a very brief period of time i.e. days whereas it is quite clear that sustained postural abnormalities can take months to over a year to fully respond [13]. Thus, the question of whether a patient has failed to benefit from surgery can be difficult.

3. Surgical technique

Surgical technique for deep brain stimulation (DBS) of the medial pallidum is highly variable across surgical groups [14–20]. In dystonic patients, it has been our preference to treat patients under full anaesthesia to avoid the problems of patient discomfort and postural difficulties affecting optimum positioning. With modern MRI imaging it is possible to visualise the actual target nucleus. To avoid stereotactic inaccuracies that attend MRI scans, we use CT/MRI fusion, merging the preoperative MRI scans to an intraoperative stereotactic CT scan (Fig. 2). Electrode placement is performed anatomically and confirmed with an intra-operative CT scan. Following this, the electrodes are immediately internalised with the implantable pulse generator (IPG) or in some cases externalised for a few days to study the neurophysiology of the condition in that particular patient or to confirm efficacy.

There are numerous papers to support the use or lack of the use of microelectrode recording to acquire the target [21–31]. There are no studies directly comparing the two but it seems reasonable to use microelectrode recording if targeting entirely based upon MRI imaging due to inherent possible inaccuracies due to field distortion [32–34]. However, there is a well documented risk attached to such recording [35,36]. We do not use micro recording at Oxford.

One of the useful early predictors of outcome is the loss of pain after surgery which is apparent immediately. Pain is virtually a universal feature of dystonia and in our experience is the first symptom to respond. Following this, within the first few days, jerking movements diminish. Numerous authors have also shown that the fixed postural components take months to respond [13,37,38]. We find that during the acquisition of the pre-operative MRI scan under general anaesthesia, we can examine patients to confirm that the static or fixed postural abnormalities relax. If they do not, representing fixed flexion deformities, it is unlikely that they will respond to surgery.

4. Choice of implant

To date, the vast majority of implanted systems have been those of one manufacturer though this will change as new manufacturers have entered the market. The choice that surgeons generally face today will be common to all devices.

Dystonia is virtually always a bilateral condition requiring electrodes to be implanted in both hemispheres. Some surgeons prefer to implant separate single channel IPGs on each side. Others prefer to implant a dual channel device. Theoretically, separate devices mean that it is unusual for both pacemakers to run out of battery life simultaneously with its possible consequent rebound dystonia which can be life threatening. Also, single channel pacemakers have a lower profile that the much larger dual channel ones and this is of importance in children and very thin patients.

However, the dual channel device does inherently imply fewer surgical procedures and therefore fewer complications. Patients have to be followed up regularly to ensure the pacemaker is changed prior to the battery life expiring. It is our preference to implant the dual channel device in the vast majority of patients and replace them routinely when near end of life.

Rechargeable pacemakers are a new addition to the market place. These devices rely on the patient recharging the IPG on a regular basis via a radiofrequency coil positioned over the device. The frequency of recharging will depend on the voltage and frequency