

Deep brain stimulation for unusual movement disorders

The development of deep brain stimulation in the treatment of Parkinson's disease as an effective and safe therapy has led clinicians to explore the treatment of disorders for which there has been no effective treatment. Such therapy does not require the ablation of tissue and as a result does not preclude future newer treatments should they become available. Furthermore, because such electrodes can be externalised for extended periods the clinician and patient can both assess any benefits that may accrue. Given that no permanent lesions are made it is also possible to implant several electrodes in different target sites to select the best target for long term effects.

We present cases where deep brain stimulation has proven to be effective in some unusual movement disorders.

Camptocormia

A 38 year old gentleman was referred with a 10 year history of severe repetitive spasms of the abdominal muscle such that sitting up or standing were impossible. He had a brief course of anti-psychotics for severe depression several years previously, which may have been relevant to the causation. On the understanding that pallidal stimulation in Parkinson's disease can improve dystonic aspects of the condition, the patient was offered bilateral pallidal deep brain stimulation.

The electrodes were then externalised for a week's observation period. During this period stimulation allowed him to sit unaided and he perceived the benefit to be significant enough to go onto full implantation of the Kinetra pulse generator. Within six months he was able to walk with one stick.

After 18 months his symptoms recurred. It was found that one of the leads had fractured and after surgical re-implantation, his symptoms rapidly improved. This confirmed that the benefits seen were due to deep brain stimulation and not psychological.

Anterocollis

A 79 year old gentleman was referred with severe anterocollis of several years duration. Botulinum injections were not feasible given the muscle groups involved. He developed severe swallowing problems with repeated episodes of aspiration pneumonia requiring several admissions to the local intensive care unit. Spasmodic torticollis we knew responds to bilateral pallidal stimulation and on this basis he consented to surgery. Under general anaesthesia bilateral pallidal electrodes were implanted and

externalised. He was then observed over a week and it was noted that his neck posture was improving. He therefore underwent a full implantation of the pulse generator.

Six months after surgery he is now able to eat and drink normally and his neck has assumed an erect posture. Interestingly, EMG studies of his neck muscles before stimulation showed that the sternocleidomastoid muscle on the right was active but the contralateral was not, nor were the trapezius muscles bilaterally. With chronic stimulation normal activity has returned to all muscle groups.

Lesch-Nyhan Syndrome

Although not as obvious as the above two as a movement disorder, Lesch-Nyhan is a genetic condition characterised by crippling self-mutilation associated with dystonia. Taira *et al* have described bilateral pallidal stimulation in a 19 year old man with the intention to alleviate the dystonia. Interestingly, his self-mutilating behaviour also improved.

Senile Chorea

A 67 year old right handed lady developed choreic movements over 6 years affecting the right limbs, trunk, face, tongue and to a lesser extent the left side of her body. Investigations, including MRI and genetic testing, had failed to identify a cause for the symptoms. Activities of daily living were difficult, and she was unable to mobilise outside, due to poor mobility and embarrassment. Left pallidal and Vop thalamic electrodes were implanted in view of the predominantly right-sided symptoms. Pallidal stimulation almost completely abolished the right limb movements, and temporarily improved the orofacial chorea.

However, although after six months chorea in the right limbs remained suppressed, left-sided movements had become more prominent. This was addressed by initiating thalamic stimulation after turning off the pallidal stimulator. This eliminated almost all choreoform dyskinesia and has continued to do so for two years.

Choreo-acanthosis:

Burbaud *et al* have reported a case of a 43 year old man with choreo-acanthocytosis and severe oro-mandibular and truncal spasms. He was treated with bilateral stimulation of the Vop nucleus of the thalamus with successful alleviation of spasms such that he was able to eat and walk again.



Professor Tipu Z 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 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.



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.



Camptocormia. Pre-op (left) - Involuntary truncal flexure making normal gait impossible and causing lumbar pain. Post-op (right) - Able to stand and walk upright for short distances without support.



Senile Chorea. Pre-op (left) - Continuous choreic movements of right arm, face and tongue. Beginning to affect left hand and foot. Post-op (right) - Right limb chorea abolished with temporary improvement in other areas.

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Head tremor

Head tremor in isolation has not generally been considered an indication for surgery yet there is a report of two cases of head tremor that have been successfully treated by bilateral thalamic stimulation.

What is emerging is that quite a few unusual movement disorders may well have dystonia as the underlying aetiology. If this is so, bilateral pallidal or thalamic stimulation may well be of benefit. Given the inherent risks of surgery this should be offered only when all medical therapies have been tried to no effect. At the same time it should not be withheld until too late for benefits to accrue i.e. after postural abnormalities have become fixed.

The nature of such cases renders it unrealistic to expect randomised controlled studies of treatment efficacy, hence it is important that carefully studied case reports are published. Nevertheless, with time it is fairly clear that the number of established conditions responsive to deep brain stimulation can only expand.

Future possibilities

Levo-dopa resistant akinetic disorders are conditions for which medical treatment is palliative only. Such conditions as progressive supranuclear palsy and multi-system atrophy in common with Parkinson's disease show degeneration of a brain stem nucleus called the pedunculopontine nucleus (PPN). The PPN receives an inhibitory input from the medial pallidum which is increased in parkinsonian conditions. It has been well recognised that in rodents stimulation both chemically and electrically induces movement. We have studied the effects of stimu-

lating the PPN in the parkinsonian primate model and have shown that microinjections of bicuculline (a GABA antagonist) has a very profound effect on reversing akinesia. Given this, it is likely that deep brain stimulation of this site may be of use in alleviating the “on” medication freezing in late Parkinson's disease and hopefully in L-Dopa resistant parkinsonian disorders.

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