

# **Deep Brain Stimulation for Dystonia Frequently Asked Q & A's**

Deep brain stimulation (DBS) is also known as Activa® Therapy, branded by Medtronic® the world's largest medical device company.

## **What is DBS?**

A patient undergoes surgery to implant a medical device, usually called a Kinetra™ . This is similar to a cardiac pace-maker. It is connected to electrodes which are precisely positioned in both sides of the brain. Continuous stimulation through these electrodes, blocks the signals which cause the physical symptoms of the disease. The aim of the treatment is for patients to regain control over their movement.

All the individual components of the DBS system are implanted underneath the skin so they are barely visible; the Kinetra is implanted underneath the skin usually on the chest wall or occasionally in the lower abdominal wall.

## **How does it work?**

The electrodes are implanted into an area of the brain known as the Globus Pallidus Interna (GPi) The minute electrical currents which are carefully controlled, have a beneficial effect on the major disabling symptoms of dystonia such as the involuntary muscle contractions. These contractions cause abnormal movements and postures which can be painful. It is not known exactly how it all works. The GPi is an area of the brain which is extensively involved in controlling and co-ordinating movement. Stimulation of this area is also known as pallidal stimulation.

## **What does the operation involve?**

Before a patient is accepted for surgery, a detailed medical history is taken and they will have to undergo a series of assessments, often including neuropsychological tests. This is

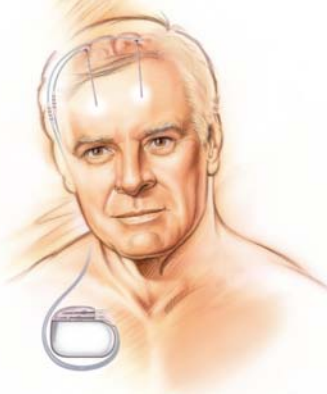
done to ensure that DBS is the best option for the patient. A video of the patient can also be made so that a patient's movement can be compared after surgery. Once the procedure and process has been fully discussed with the patient, and all the implications understood, the patient is offered the treatment.

Next the patient undergoes a series of brain scans, usually both MRI and CT, which allows the surgeon to decide exactly where to place the electrodes in the GPi. Before the scans, a lightweight frame will be fixed to the patient's skull to prevent movement within the scanner. They are usually performed under a general anaesthetic. These scans help the team to plan the route and the final position the wires will take, avoiding other important areas of the brain next to where the wires need to be.

The wires are then implanted under a general anaesthetic. This is unlike the procedure for patients with Parkinson's disease or Essential tremor, who may have the surgery awake under sedation.

Two small burr holes are made on either side of the skull. The electrodes are passed through these, down to the planned target area in the GPi. There are four in-line electrodes at the end of a DBS lead. The aim is to introduce as many as possible of them within the depth and volume of the target area. The exact placement for the electrodes is then decided from the scans and other tests carried out in the operating theatre. When the electrodes are in the right place, the holes are closed and the other end of the DBS™ leads (not containing the electrodes) are connected individually to two extension cables. The two extension cables are tunnelled underneath the skin behind the ear, on the same side of the neck to the chest wall. They are both connected to the Kinetra stimulator which is then put into a small 'pocket' made under the skin. All the wounds are closed and dressed.

The patient usually then has another scan to check that the electrodes are in exactly the right place. They are transferred to recovery for a brief period of observation, before being transferred onto a neurosurgical or neurological ward.



Diagrammatic drawing of the placement of the DBS stimulator components

N.B. The Kinetra may be placed on the lower abdominal wall in children and in female patients for cosmetic reasons

What happens next depends on the centre where the surgery takes place. The stimulator may be programmed with an initial set of electrical parameters, either straight away or within a few days. A physician programmer uses radio waves to 'talk' to the implanted Kinetra. This is not painful or invasive. The physician can then adjust many variables of the stimulation, including which electrodes are selected, to ensure that the best effect is achieved. It is usual for the patient to need many adjustments over the next few months.

### **How long will I be in hospital?**

The amount of time that you have to remain in hospital depends on the centre implanting the device. Patients are usually admitted for further tests, one to three days before surgery. The length of the operation also depends on the technique used by each centre, but it often lasts between 3-6 hours from start to finish.

As long as the electrodes are accurately placed, without

complications, the recovery period usually lasts from between 3 to 5 days. The patient is then discharged with clear instructions about the amount of activity that is safe, and with a series of outpatient appointments.

### **How long will it take to work?**

Patients who have DBS for Parkinson's disease and Essential tremor often see improvements immediately or within a few hours. It often takes several months however, for people with dystonia to see benefits from GPi stimulation. Patience is essential. Everyone is different and so every person responds differently to stimulation. Many patients do notice soon after surgery, that they no longer have the chronic pain which comes from the muscle spasms and contorted postures. This is normally an indication for doctors that patients will also see improvements with the involuntary muscle contractions and abnormal postures. The tremor associated with dystonia may also improve quickly.

Some clinical studies have shown that the maximum benefit from stimulation may occur approximately 6 to 12 months after the stimulator is implanted and that no further benefit can be expected after that period.

For Patients to see improvements, a number of things must occur. Firstly both sets of electrodes must be accurately placed in the GPi on both sides of the brain. Secondly the physician programmer must also find the optimum stimulation settings. Finally any other medical considerations have to be taken into account. The best combination is not achieved the first time the Kinetra is activated and programmed after surgery. Patients should expect that they will have to undergo frequent programming sessions to get the best clinical improvement for the symptoms being targeted. Sometimes not all of the symptoms can be equally improved, and patients may still need to have other therapies such as Botulinum toxin injections for Blepharospasm.

## **What are the risks?**

There are three main types of risks: those that are associated with any brain surgery; risks related to the device and having a foreign body inside you and those connected with the stimulation itself.

All surgical procedures have the same degree of risk associated with having the surgery itself and the anaesthetic. There are a number of risks which relate to having the electrodes inserted.

1. The risk of stroke which is reported to be between 1 - 3% in the latest clinical literature
2. intracranial hemorrhage
3. hemiplegia
4. headaches and seizures.

However, the advances in MRI technology have helped the neurosurgical teams to implant the electrodes into a precise position via the best routes, avoiding major blood vessels.

Having the stimulator hardware implanted may carry a small risk of infection. Skin can breakdown over the Kinetra unit or where the leads come out of the burr holes. There may be a small risk of parts of the stimulator failing, for example a DBS lead may break which would mean that the therapy would have to be stopped until the problem was resolved. If the stimulation is stopped, this may cause the symptoms to temporarily get worse again. This is known as the 'rebound effect' which lasts until the part is replaced.

The GPi is near parts of the brain which do not coordinate movement. Stimulating one of these areas by mistake can cause visual phenomena, speech disturbances, dyskinetic movements and sensory disturbances. Because the stimulator can be adjusted an infinite number of times, side effects caused by the stimulation usually do not last and can be reduced or eliminated whilst still working on the areas effected by dystonia.

## **What if it does not work?**

There are a number of factors which significantly increase the chance of surgery being a success:

1. Detailed patient assessment is carried out before surgery and from the assessment, medical staff have a clear selection and deselection process
2. The electrodes are precisely placed in the brain
3. Accuracy in targeting means that electrodes are placed in exactly the right place
4. The team are experts in managing patient care after the stimulator has been implanted.

Before the stimulator is implanted, the physician, patient and their family discuss what improvements can be expected for the symptoms which are being targeted and from this, realistic expectations and goals are set. Every patient has an individual response to stimulation but published data shows an improvement in mean dystonia movement scores of 51%, with many patients showing more than a 75% improvement.

One of the benefits of DBS is that the electrodes do not destroy brain tissue so if it is decided that a satisfactory clinical outcome has not been achieved, the DBS system can simply be turned off or removed (explanted). You would have to have an operation under a general anaesthetic to have the stimulator removed.

## **How do I go about being considered for DBS?**

To find out whether you are suitable for DBS, you need to be referred by your GP to a specialist neurologist or to the specialist implanting neurosurgeon, who has an interest or is an expert in this area. It is often difficult to find a specialist neurologist and you may have to look outside of the immediate area where you live. Ideally the neurologist should be part of a team which includes a specialist functional neurosurgeon, neuropsychologist, anaesthetist, and specialist nurses.

There are a number of UK neurosurgical centres which are

able to accept referrals as well as a number of specialist neurologists who assess patients and where appropriate, refer them to the centres. See p12 for a list of centres, neurosurgeons and specialist neurologists who offer the surgery.

After a patient has been referred, they have to undergo a detailed selection and de-selection process. If it is decided that a patient is suitable, then the centre offering the surgery then has to get funding and authorization from the patients GP and local PCT. This process can take a long time and is still not a guarantee that surgery will take place. If approved, the patient is put on a DBS neurosurgical waiting list.

### **Where can I get the operation?**

There are many neurosurgical centres in the UK that accept referrals for patients who are considering DBS. The Dystonia Society has a complete list and there is also a list of centres, neurologists and neurosurgeons on page 12 of this booklet. It may be necessary for a patient to travel to a neighbouring county for surgery as there is not a centre in every county. The patient may want to take the list of centres offering the surgery with them when they go to see the GP for a referral.

### **How long does the recuperation take?**

As long as there are no complications as a result of the surgery, patients usually stay in hospital for 3 to 5 days after the device is implanted. This may vary however, according to the different practices of each centre. Some centres are involved in clinical research programmes. These centres may ask patients whether they are willing to take part in their studies. This may involve a slightly longer stay in hospital.

After a patient is discharged from hospital, they will be given clear and precise instructions about how much activity they may do. They will also usually be given a structured outpatient appointment schedule which should be closely followed. Outpatients' appointments are arranged to check that the

wounds are healing and to assess how well the stimulation is working. Adjustments to the stimulator settings may be made at this time. After the patient is discharged from hospital, they should be able to resume normal life.

### **What types of dystonia can be treated?**

The types of dystonia which respond well to DBS and which are considered to be a primary indication that surgery may be helpful are:

- Primary generalised dystonia
- DYT1 positive or negative dystonia
- Hemidystonia
- Regional dystonias like torticollis, anterocollis or retrocollis

In addition there are case reports in clinical literature for less frequently reported dystonias like craniofacial dystonia, choreo-athetoid dystonia and dystonic movements in cerebral palsy.

There is no upper age limit for patients being offered surgery, as long as there is no other illness which may complicate the surgery. This would be discussed during the patient assessment.

Unless there are exceptional circumstances, children are not considered for DBS implant until they are 7 years old although assessments may be made earlier in preparation for the child's seventh birthday.

### **What follow-up treatment is needed?**

Adhering to the follow-up schedule provided by the implanting team is crucial to maximising the clinical efficacy of the stimulation and any adjustments of medications that maybe required. In addition the patient maybe considered for additional therapies such as Botulinum toxin injections for added clinical benefits.



**It is important that during the first weeks, patients should watch the incision site carefully. The implanting team must be told if there is any sign of redness, soreness, pain over the incision site, (other than what would be expected after surgery) or anything else that is unusual.**

During the first months after the stimulator is implanted, it will be necessary to attend an outpatients' clinic to assess the clinical benefits of the stimulator and make any necessary adjustments to the stimulator settings. The adjustments are made by a physician programmer which uses radio waves to 'talk' to the stimulator. This is a painless procedure where a device is held over the stimulator. The number of outpatient appointments varies from one patient to another but eventually, the stimulator parameters plateau to provide maximum clinical effect on the symptoms being targeted. Once this stage is reached, the stimulator only needs very small infrequent changes.

The stimulator is powered by a battery. The level which the stimulator is programmed at, effects how long the battery lasts. The battery life span can be checked by a physician programmer or a patient Therapy Access Controller™ (TAC). This is a small patient programmer which is given to some patients when the implanting team agrees. The TAC allows a patient to monitor the battery life and switch the device 'on' or 'off' if necessary. When the implanting team agree, this device also allows the patient to make minor adjustments to the stimulation intensity.

Once the battery is reaching its 'end-of-life', patients need to have a small operation. The battery is removed and then replaced with a new stimulator unit. The extension cables and DBS leads are not replaced at this time. This surgery usually lasts for about one hour.

After this surgery, the new stimulator which contains the fresh battery is programmed using the same settings as the previous stimulator. The patient is then discharged from hospital.

## **How do I get an assessment for DBS?**

DBS is a complex procedure and so it is only offered to a patient who has had a detailed assessment from a multidisciplinary team, and who has not responded to, or is not suitable for, less invasive procedures.

The patient has to get a referral to a specialist team which has experience in movement disorders and DBS surgery. The process starts with a letter from the patient's GP. This is sent to either the specialist neurologist or the specialist neurosurgeon, asking them to discuss DBS and whether or not this procedure is suitable for that patient. Where the person is considered to be a potential candidate, the assessment process is started. There is a list of the specialist neurologists and neurosurgeons who offer the surgery, on page 12. Patients often have to be referred outside of their local area because there are not many specialists in this field.

The implanting team then consider, discuss, and agree, the following issues with patients:

1. All aspects of a patient's medical history are then looked at.
2. The risks and potential benefits of DBS are considered.
3. All that is involved in the surgery is discussed.
4. Realistic expectations are agreed between the implanting team, the patient and their carer.
5. The Team and the patient agree which symptoms will be targeted and what success they should expect.
6. They also discuss how long it is likely to take before the patient begins to see improvements.

If, after all these things have been discussed, the patient then decides that they want to go ahead with surgery, they are put on a waiting list. During this time the implanting

centre has to gain funding for the surgery from the patient's Primary Care Trust (PCT).

### **What questions should I ask my specialist?**

The patient may wish to include the following questions during any discussions with the expert multidisciplinary team

- ▶ Am I a candidate for this therapy? Why? Why not?
- ▶ What are the potential risks and benefits of the therapy?
- ▶ What are the potential risks and benefits of the surgery?
- ▶ What are the side effects of the therapy? Can the side effects be controlled?
- ▶ What activities may I be able to resume as a result of the therapy? How likely is it that I will be able to walk, feed myself, write, work, drive and sleep through the night?
- ▶ How should I prepare for surgery?
- ▶ What kinds of tests will be conducted before the surgery?
- ▶ What can I expect the day of surgery?
- ▶ How long does the surgery last? Is it painful?
- ▶ How long will I need to be hospitalised?
- ▶ Will my condition improve immediately after surgery, or will it take more time?  
What precautions will I need to take after surgery?
- ▶ How often will I need to return for follow-up visits?  
How many programming sessions to adjust the stimulation can I expect?
- ▶ Will I still need to take medication after having the Activa System implanted?

## UK DBS Centres that can accept referrals for Dystonia

<b>Hospital</b>	<b>Ad- dress</b>	<b>Neurosur- geon</b>	<b>Neurologist</b>	<b>Tele- phone</b>
Ninewells	Dundee	Mr. S. M. El-jamel	Dr. R. Swingler	01382 660111 x35712
Western General Infirmary*	Edin- burgh	Prof. I. Whittle	Dr. R. Davenport	0131 537 2104
Newcastle Gen- eral*	Newcas- tle	Mr. A. Jenkins	Dr. D. Burn	0191 273 8811 x22449
Walton Centre*	Liverpool	Mr. T. R. K. Varma	Dr. M. Steiger & Dr. N. Fletcher	0151 529 5681
Queen Elizabeth Medical Centre	Birming- ham	Mrs. R. Mitchell	Dr. H. Pall & Prof. A. Williams	0121 697 8223
Hope	Salford	Mr. P. Richard- son	Dr. J. Dick & Dr. M. Kellett	0161 787 2368
Radcliffe Infir- mary*	Oxford	Prof. T. Aziz	Dr. R. Gregory	01865 224605
Frenchay*	Bristol	Mr. S. Gill	Dr. P. Heywood	0117 970 2461
Royal London	London	Mr. H. Ella- mushi	Dr. J. McAuley	020 7377 7000 x7211
Charing Cross	London	Prof. T. Aziz	Dr. P. Bain	020 8846 1182
Kings College	London	Mr. R. Selway	Dr. M. Samuel & Dr. C. Clough	020 7737 4000
National Hospital for Neurology & Neurosurgery*	London	Prof. M. Hariz	Dr. P. Dowsey- Limousin & Prof. N. Quinn	020 7837 3611
Royal Hallam- shire*	Sheffield	Mr. M. Radatz & Mr. J. Rowe	Dr. R. Grue- newald	0114 271 3302
Southern Gen- eral	Glasgow	Mr. L. Dunn	Dr. D. Grossett	0141 201 2020
University Hospi- tal Wales	Cardiff	Mr. R. Nannapanneni	Prof. A. Rosser	02920 742708
Addenbrookes	Cam- bridge	Mr. Colin Watts	Dr. Roger Barker	01223 245151

\* accepts Paediatric referrals

## **Want to Know More or Help?**

**Further information and support is available through the contacts listed on the back page.**

**Additionally if you would like to talk with someone who has had DBS about their experience you can call The Dystonia Society Helpline in the UK on 08450 956575 (local call rates apply)**

**If you have access to the internet you can download video of the DBS procedure and its impact at**

**[www.parkinsonsappeal.com](http://www.parkinsonsappeal.com)**

**We hope you have found this document useful and if you have any comments or suggestions please email [mwfeedback@btinternet.com](mailto:mwfeedback@btinternet.com)**

## Notes

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## **Want to Know More or Help?**

**The Dystonia Society** Reg Charity number: 1062595,  
46/47 Britton Street, London EC1M 5UJ  
Phone : +44 20 7490 5671  
Fax: +44 20 7490 5672  
E-mail: [info@dystonia.org.uk](mailto:info@dystonia.org.uk)  
Website: <http://www.dystonia.org.uk>  
Helpline: 08450 956575 (local call rates apply)

**The Parkinson's Appeal for Deep Brain Stimulation** Reg Charity No:263064  
15 Southampton Place,  
London WC1A 2AJ  
Tel / Fax: 020 7233 6034  
E-mail: [parkinsonsappeal@yahoo.com](mailto:parkinsonsappeal@yahoo.com)  
Website: <http://www.parkinsonsappeal.com>

**Dystonia Medical Research Foundation**  
One East Wacker Drive, Suite 2430  
Chicago, Illinois 60601-1905. America  
Phone: 001-312-755-0198  
Fax: 001-312-803-0138  
E-mail: [dystonia@dystonia-foundation.org](mailto:dystonia@dystonia-foundation.org)  
Website: <http://www.dystonia-foundation.org>

**Medtronic** <http://www.medtronic.co.uk>

**DBSforDystonia discussion group**  
<http://health.groups.yahoo.com/group/DBSforDystonia>

**ADDER**  
Website: <http://www.dystonia.co.uk>