Deep brain stimulation

When is DBS used to treat dystonia?

Deep brain stimulation works most effectively for people who have an inherited (genetic) form of dystonia or for those where the dystonia has no identified cause (this is called idiopathic). The treatment is provided for those with inherited or idiopathic dystonia who have a severe generalised dystonia, neck (cervical) dystonia or dystonic tremor when other treatment options (such as botulinum toxin injections and oral medications) have failed to provide adequate relief.

The effectiveness of DBS is thought to reduce as the proportion of life lived with dystonia increases so, for children, ideally DBS should be offered early, preferably within a few years of the onset of symptoms, although the right timing will vary by individual.

Unfortunately, DBS does not work as well for the majority of those with acquired dystonias (which have a known but non-genetic cause such as cerebral palsy, a disorder of the metabolism or brain damage). However, it is still sometimes provided as it can have benefits for certain carefully selected patients. The DBS implanting centre team will be able to discuss this with you. If the dystonia is drug-induced (tardive) then DBS can work well.

A rigorous patient assessment, selection and de-selection process is carried out prior to the operation. In addition to the type of dystonia, a number of other factors are taken into account – for instance DBS may not be

What is Deep brain stimulation?

Deep brain stimulation (DBS) is a surgical procedure in which two thin, insulated electrodes are inserted into the brain. These electrodes are then connected by a wire under the skin to a battery usually implanted in the chest or in the abdomen. The battery operates similarly to a pacemaker delivering targeted electrical pulses that block the signals that cause the symptoms of dystonia.

The battery is implanted below the skin on the chest wall (or sometimes the lower abdominal wall) so is barely visible but an outline of its shape and of the wires connecting it to the brain may be visible.

How does it work?

The carefully controlled, minute electrical currents delivered through the electrodes on both sides of the brain can have a beneficial effect on the involuntary muscle contractions caused by dystonia. As a result, the symptoms of dystonia such as abnormal movements and postures and/or dystonic tremor can be eased. In addition, DBS can reduce the pain caused by dystonia. The electrodes are usually implanted into an area known as the Globus Pallidus Interna (GPI). Stimulation of this area is known as pallidal stimulation. Occasionally, another part of the brain, the thalamus, is targeted instead.
undergoes a series of scans of the brain which receiving a psychological assessment. The patient next
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DBS can have a significant effect in easing the symptoms of dystonia but it is important to go into the operation with realistic expectations.
For inherited/idiopathic dystonias, on average DBS reduces the severity of symptoms by slightly more than 50% but experience varies widely between individuals. Some (around 20% of patients receiving DBS) get very little benefit. At the other extreme, some can experience symptom reduction of 80% or more. Overall, patients with the DYT1 gene experience the best average results but the vast majority of those who meet the clinical criteria for receiving DBS gain substantial relief.

After DBS some patients are able to stop receiving other treatments for their dystonia, but many patients do need to continue receiving botulinum toxin injections and/or oral medication.

What does the procedure involve?
The first step is a rigorous assessment to ensure the patient is suitable – this will include a comprehensive review of medical history and a psychological assessment. The patient next undergoes a series of scans of the brain which enable the implanting team to identify the target areas of the brain for placement of the electrodes. Patients often want to know if their hair will be shaved off prior to the operation. Practice on this varies between treatment centres – some don’t shave the hair at all, others a patch around the implant area and others all the hair.

The operation to implant the electrodes and battery takes a few hours and the procedure is performed while the patient is asleep. Patients report the process as exhausting but not usually painful. Fortunately, the brain itself has no pain receptors and feels no pain.

Depending on the centre, the DBS system can be activated 1–2 days or a few weeks after surgery. The stimulator will start to reduce symptoms a few days after activation, but the improvement is usually gradual and may take several months to reach its full extent. During this time, the healthcare professional who programs the device will go through several programming sessions adjusting the settings of the device to achieve optimal results. The wait for the stimulator to be fully effective can be a time of anxiety so it is important to be aware from the outset that patience is required.

After the operation, the patient may also need additional support such as physiotherapy, psychological support and/or pain management.

Even after the initial tuning process for the stimulator has been completed, regular visits to the implant centre will be necessary. Given the variable nature of dystonia, keeping the stimulator at its optimum performance is an ongoing process requiring commitment from both patient and clinicians.

What are the risks?
All surgical procedures carry a degree of risk both from the surgery and the introduction of anaesthesia. However, when complications do occur they are usually mild and short-lived.

The risks of the procedure should be discussed with the treatment centre prior to going ahead. One leading UK implant centre reports the main risks of the procedure at their centre as follows: stroke / intracranial bleeding / hemiplegia (below 1% of cases), seizure (2%) and infection from the implanted device (2–4%).

Other risks include malfunction of the device (1.8%) or fracture of the lead (0.7%). Other problems that can arise over time include speech problems (dysarthria) but these can usually be reversed by adjusting the settings on the device.

How long does the battery last?
It is important to understand that the battery wears out so it will be necessary to have a surgical procedure to replace it every few years. Controlling dystonic symptoms requires a high power setting so non-rechargeable batteries can wear out in 18 months to 2 years. However, rechargeable batteries are available that can last for up to 9 years – but they do require the patient to commit to recharging regularly.

What if the stimulator stops working?
To minimise the risk of problems with the stimulator, care needs to be taken to avoid interference with the device. Magnetic resonance imaging (MRI) scans are sometimes restricted and diathermy (which is the use of high-frequency electromagnetic currents as a form of physical or occupational therapy) should be avoided. If in any doubt, you should always check with the implant centre before undergoing any such procedures.

It is possible (but unusual) for the stimulator to stop working suddenly – either because of a device malfunction or because the power runs down unexpectedly. If this happens, it is important to contact the treatment centre immediately as, in some rare cases, this can bring on a potentially dangerous dystonic storm (the medical term is status dystonicus) where the dystonic symptoms return with severity.

How do I get referred to DBS?
Referral to DBS can only be by a consultant (adult or paediatric) specialising in movement disorders. To get the procedure approved, they will need to confirm that you meet the clinical criteria described above and that all appropriate alternative treatment options have been ruled out. However, once this is done, the procedure, battery replacements and other follow-up support should be funded by the NHS. There are a number of specialist treatment centres across the UK that can provide DBS for dystonia.