

## Vagus Nerve Stimulation

For most children and adults with CDKL5 their epilepsy does not respond to medication. This is known as refractory or intractable epilepsy. It is becoming more common for these patients with CDKL5 to have a Vagus Nerve Stimulator. The youngest patient our support groups have known to have the procedure is 9 months of age. The average age of children that are currently undergoing the surgery is between 3-5.

### What is Vagus Nerve Stimulation (VNS)?

Vagus Nerve Stimulator therapy uses a pulse generator to send mild electrical stimulations to the vagus nerve with the aim of reducing the number, length and severity of seizures. NICE guidelines state that "VNS is indicated for use as an adjunctive therapy to reduce the frequency of seizures in patients whose epileptic disorder is dominated by partial or generalised seizures that are refractory to anti-epileptic medication."

### What does VNS surgery involve?

VNS surgery is carried out, in full consultation with parents and the child's Neurology team, by a neurosurgeon. The operation normally takes around one hour and is usually day surgery, recovery will depend on whether the child will be released the same day or the next. The neurosurgeon makes two small cuts, one in a natural crease on the left of the neck, and one in the left-hand side of the chest, below the collarbone. The generator is placed under the skin in the chest. A thin, flexible wire connects the generator to the left vagus nerve in the neck. The small scars from the two cuts fade over time.

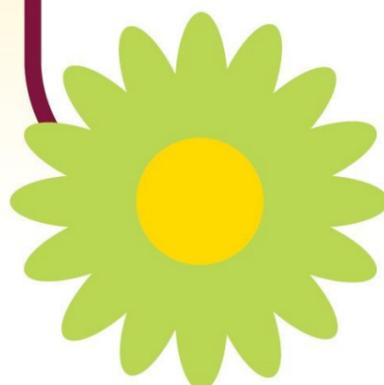
The generator is checked during the surgery but is usually left switched off for two weeks after surgery. This is to help the body heal. After that, it is usually switched on by a epilepsy nurse specialist in a clinic. They will gradually increase the settings (adjustments) over a number of weeks, the frequency of these adjustments will be determined by local policy and also how well the patient seizure control is with each adjustment. For example if a patient is having good seizure control at a certain level, then they may not need regular adjustments.

### What benefits of having VNS surgery can we expect?

It varies from individual to individual to see the positive effects of VNS. For some there are some immediate benefits as soon as stimulation reaches a therapeutic level however, it can take up to two years before full benefits are seen. For many parents they report that the magnet is able to either stop or reduce the length and severity of a seizure from the beginning. Other benefits of having the VNS which parents have noticed is the decrease in seizure activity and severity and increase in the quality of life of the patient. This in turn have a positive impact on the carers.

### Are there any risks of having VNS surgery?

With any operation that involves a general anaesthetic there is risk of a reaction to the anaesthetic. Because the surgery is invasive there is also a small risk of bleeding and infection. The Neurosurgeon will take all the necessary precautions to prevent this and you will be given dressings to ensure that the wound site is kept clean, you will be given a follow-up appointment to ensure that the site is healing well. There can be other rare complications and your surgeon will discuss this before the operation takes place and prior to you signing the medical consent form. There might be some pain for a while from the area of the implant after VNS surgery and this can be managed with over the counter pain meds. For more intense pain then seek advice from the GP.



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### What is the magnet and how should the magnet be used?

The magnet is used to add a burst of stimulation for 1 minute in the event of a seizure. The generator is normally set so that when the magnet activated the generator the amplitude of stimulation is at a high rate. The magnet can be used anytime during a seizure, but it is most likely to work when it is used towards the beginning of a seizure. You should use the magnet if...

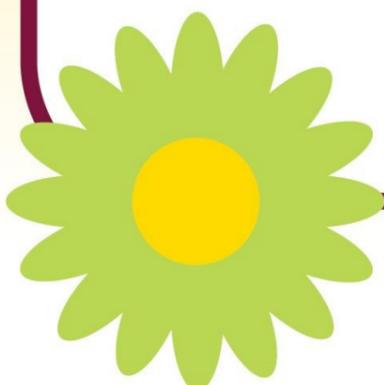
- You see seizure activity beginning to start.
- At the start of a seizure.
- Or when if you notice the patient is having a seizure.

Generally the epilepsy nurse, or community nurse will be able to provide training on the use of the magnet to education and carers for example. The magnet can be used to also switch off the generator if a MRI is needed for example.

Because the magnet is strong there are some safety tips that you should be aware of.

- Keep your magnet in a different pocket or on your belt buckle, away from your wallet as it can affect credit/debits cards for example.
- If you use a computer frequently, don't wear the magnet on your wrist – it may get too close to the computer. Wear it on your belt buckle or put it on the floor when you are working at the computer.
- Don't put magnets on top of a television, stereo, or other electronic device.
- Don't enter rooms or places that have strong electronic or magnetic fields or that have warnings for people with pacemakers or other implanted devices.
- Ask your doctor or nurse to write a letter to use while traveling that explains the VNS device and magnets. Wands used during security checks could affect the VNS device or the magnet could trigger a security alarm.

If in any doubt about the use of the magnet then should can consult the VNS Therapy Patient Manual which should have been supplied at the point of surgery, or alternatively it can be downloaded from Cyberonics or alternatively your epilepsy specialist nurse.



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### What are the Side-effects of VNS therapy?

VNS therapy is not a drug, so doesn't have the same sort of side-effects as epilepsy medicines, and it doesn't interact with other medicines. However, for some patients that have CDKL5, they may have a "honeymoon" as they do with drugs. The most common side-effects reported from having the VNS implant.

- Temporary hoarseness/change in voice tone
- Sore/tickling throat
- Shortness of breath
- Coughing

Generally these side-effects happen during stimulation, and usually get less over time. However, If you continue to have side-effects, talk to your epilepsy nurse, to see if they can help.

### Will they continue with Epilepsy medicines after VNS surgery?

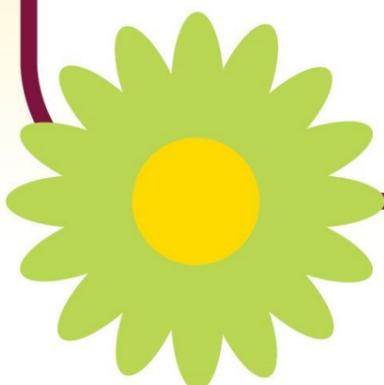
VNS is not used to replace medications. However, if there is a positive improvement with the VNS then it is possible that the Neurologist will look to reduce medications over time.

### When will the generator battery need replacing?

The generator does not last for ever and therefore at some point, it will need replacing. It usually lasts for around three to six years, depending on the settings used. The higher the frequency and output of the settings, the faster the battery will run down. The epilepsy nurse will ensure that the VNS is checked every 3 months or so and they are able to see when the battery is running low. If seizures increase and you know that the battery is nearing the end of its life it is important that you seek support from the epilepsy nurse so that the generator can be replaced at its earlier convenience so that seizure control is not impacted too greatly. The surgery for the replacement is similar to the first, however, the wires are kept in situ and are checked to ensure that they are still in tact.

### Where can I get further information?

Further information is available from Cyberonics, who make the VNS devices. To see a video of the switching on a VNS please visit the website [www.curecdkl5.org](http://www.curecdkl5.org)



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