Peninsula Cerebra Research Unit
PenCRU Childhood Disability Research

What's the Evidence?

Brain Surgery to Reduce Seizures in Children with Sturge-Weber Syndrome

Key findings

- Sturge-Weber Syndrome is a rare condition that varies in severity from person to person.
- A range of surgical procedures can be used to reduce seizures, but this makes it difficult to compare approaches.
- Current evidence suggests that disconnective brain surgery such as hemispherectomy and posterior quadrantectomy are likely to be effective to reduce seizures.
- Future research requires more comprehensive follow-up and standardised ways to assess outcomes.

Published October 2016

What were we asked?

- A parent of a child with Sturge-Weber Syndrome and drug-resistant epilepsy asked us: 'Which of the several different types of brain surgery is most effective in reducing seizures in children with Sturge-Weber Syndrome?'

What did we do?

- In July 2016 we searched a range of academic databases including; NHS Evidence, the Cochrane Library, Pubmed and TRIP database, using the search terms: 'neurosurgery', 'seizures', 'children' and 'Sturge Weber'. We also looked at relevant charity websites, and asked an expert to check a draft of this summary.

What did we find?

- Sturge-Weber is a rare syndrome affecting connective tissues. The characteristics are varied but include learning disability, glaucoma, a congenital facial (port-wine) birth mark and epilepsy.[1]
- Epilepsy severe enough to consider surgery is not common.[2]

When is brain surgery appropriate and what are the different types of brain surgery?

- Surgical treatment of epilepsy involves the removal, modification or disconnection of an area of the brain with the aim to stop seizures occuring.[3]
- National Institute for Health and Care Excellence (NICE) guidance recommends that referral for brain surgery is

considered for children who do not respond to anti-epileptic medications, and when certain criteria are met. These criteria include a diagnosis of Sturge-Weber Syndrome [4]

- A decision about whether surgery should be considered in children with Sturge-Weber Syndrome and drug resistant epilepsy will be made considering signs that the syndrome is progressive. For example, if there is persistent seizures despite medication, an increase in the duration of seizures, the disorienting symptoms after a seizure and/or the extent of the brain lesions.[1]

- A number of tests are carried out that help to determine whether surgery is appropriate and the appropriate procedure. These include EEGs and MRI brain scans and assessments of memory, intelligence and behaviour. The tests will vary depending on individual circumstances. To find out more about the tests that may be involved, please see the *signposts to other information* section below.

- Epilepsy brain surgery will only go ahead if the tests show that the benefits are likely to be higher than the risk of complications. The risks can vary for the individual but may include:
  - Memory problems
  - More focal/less severe seizures
  - Reduced vision to one side
  - Increased one-sided paralysis
  - Behaviour problems.[5]

  Other complications from surgery include infection, bleeding, stroke, and death in a very small number of people.[3]

- The risk of complications has to be balanced against risks of uncontrolled epilepsy. These include progressive memory and behavioural difficulties and risk of sudden death.[3]

- The timing of surgery is likely to depend on specific patient characteristics. Seizures which start before 6 months old have been associated with poorer developmental outcomes.[1] As seizures may also exacerbate brain injury in Sturge-Weber Syndrome, [6] early surgical treatment (under age 1 year) may be recommended.

- However, age at the time of surgery does not appear to have an adverse effect on seizure control, with older children still benefitting from surgery[2, 10]. The long-term effects of epilepsy surgery at a younger age, or close to the start of seizures compared to surgery at an older age, or later in the course of the disease are not known.[7]

- There are different types of epilepsy brain surgery. The type of procedure will depend on a number of factors, including the nature of seizures experienced, where in the brain seizures begin and the extent of the brain abnormality. Different procedures may be combined.

- Some of the more commonly performed types of epilepsy surgery include:

  - *Resection*

  This type of surgery is used when it is clear which part of the brain the seizures start in. This small part of the brain is removed.

  - *Disconnection*

  Parts of the brain can be disconnected to interrupt seizure pathways, such as the temporal, occipital and parietal lobes, this is sometimes called posterior quandrantectomy.

- *Hemispherectomy/hemispherotomy*

In this surgery, one half of the brain is disconnected from the other because one half of their brain is abnormal.

- *Corpus callosotomy*

This type of surgery is mainly used for generalised seizures, particularly frequent 'drop attacks'. In this surgery, the two hemispheres (halves) of the brain are separated.

Which brain surgery is most effective to reduce seizures in children with Sturge Weber Syndrome?

- We found five published case series that report the seizure outcomes of different brain operations for individuals with Sturge-Weber.

- One paper reviewed the results of brain surgery at one hospital in France for 27 children. This study found that all 8 children who had a hemispherectomy, and 11/19 children who had a resection, were seizure-free after the surgery.[8]

- A second paper reviewed the results of brain surgery carried out at two hospitals (France & Canada) for 20 patients (mostly children). This review found that 5 children who had hemispherectomy were all seizure free after surgery; 8/14 who had resection became seizure-free and 1 who had corpus callosotomy, needed resective surgery before a reduction in seizures was observed.[2]

- The third paper reviewed the results at one hospital in America, of 8 children with Sturge-Weber; 4 children had hemispherectomy and 3 of these were seizure-free or almost seizure-free following surgery. 4 other children had resection and 1 of these was seizure-free/almost seizure-free afterwards.[9]

- The fourth paper specifically reported the results of 10 children with Sturge-Weber Syndrome who had a type of disconnection surgery called posterior quadrantectomy in Japan. This study reported 8/10 were seizure-free following surgery.[10]

- In the preceding cases series when resection did not lead to seizure freedom, the surgery was often incomplete. This means that the target area of the brain could not be completely eliminated due to its closeness to other crucial functioning parts of the brain.

- The fifth paper reported the results of hemispherectomy surgery in a Canadian hospital for 24 children, including six who had Sturge-Weber. All six were seizure-free after hemispherectomy, but three children required treatment for glaucoma after surgery.[11]

- A questionnaire was completed by 32 parents of children with Sturge-Weber who had received a hemispherectomy at one of eighteen different hospital across the world. The results reported that 26/32 children were seizure free afterwards.[12]

- While the questionnaire suggested that almost all parents were happy the surgery had taken place, 38 parents did not respond. So it is possible, the results may be biased to those who had positive outcomes.

What do we think?

- The evidence suggests disconnective brain surgeries (hemispherectomy/ posterior quadrantectomy) which interrupt the epileptic pathway, rather than surgery to remove small areas of the brain, are likely to be effective to reduce seizures for children with Sturge-Weber Syndrome.

- However, comparing the effectiveness of different surgeries is difficult because there are different surgical procedures and Sturge-Weber is a rare syndrome, with wide varying symptoms.
- The evidence largely consists of studies that have looked back at cases from one or two hospitals, and report whether the procedures delivered were effective in reducing or eliminating seizures.
- The processes used to select candidates for surgery, surgical techniques and measures and definitions of freedom of seizures vary widely. Therefore it is not possible to use the available evidence to give general advice for children with Sturge-Weber Syndrome.
- The chances of reduced seizures after surgery will vary from person to person. The doctors involved in each individual case are best placed to advise.
- Hospitals collecting data about procedures and outcomes prospectively for all children, and reporting their results regularly, would help provide further guidance on which factors predict effective surgery.
- Future research requires a more comprehensive, standardised outcome assessment. As well as looking at the timing of surgery and the long-term seizure outcomes, this assessment could include agreed visual, motor, behavioural, cognitive and quality of life outcome measures.
- Any child in the UK for whom surgery is considered should be reviewed and evaluated at one of the four Children's Epilepsy Surgery Service (CESS) in England or the similar service in Scotland.

Signposts to other information

For more information on Sturge-Weber Syndrome:

- www.sturgeweber.org.uk/
- www.sturge-weber.org/
- www.gosh.nhs.uk/medical-information-0/search-medical-conditions/sturge-weber-syndrome

Epilepsy Action provide information on the Children's Epilepsy Surgery Service in the UK and the tests to determine the appropriate surgery:

- www.epilepsy.org.uk/info/treatment/epilepsy-surgery/children
- www.epilepsy.org.uk/info/treatment/epilepsy-surgery#tests

Epilepsy Action also provides links to epilepsy organisations in other countries which can provide further information:

- www.epilepsy.org.uk/about/international-epilepsy-organisations

The Hemispherectomy Foundation have a list of questions to ask before surgery:

- http://hemifoundation.homestead.com/sws_hemi_questions.html

Youth Health Talk has videos of young people describing their experience of epilepsy brain surgery:

- www.healthtalk.org/young-peoples-experiences/epilepsy/brain-surgery-and-vns

We would like to hear your feedback on this summary – please email us at pencru@exeter.ac.uk if you have any comments or questions.

Published October 2016

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Note: the views expressed here are those of the Peninsula Cerebra Research Unit (PenCRU) at the University of Exeter Medical School and do not represent the views of the Cerebra charity, or any other parties mentioned. We strongly recommend seeking medical advice before undertaking any treatments/therapies not prescribed within the NHS.