Peninsula Cerebra Research Unit
PenCRU Childhood Disability Research

What's the Evidence?

Seizures: emergency medication and outcomes in Sturge-Weber Syndrome

Key findings

- NICE guidance is that emergency medication should be administered after five minutes of prolonged seizure activity to try and prevent children entering status epilepticus.
- Children who have frequent episodes of seizures may have individually tailored guidelines which should be followed when they have a seizure.
- The long-term effect of prolonged seizures on learning disability is not clear.

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Published September 2013

What were we asked?

A parent asked whether there is any evidence for:

a) how long after seizure onset (i.e. 5, 10, 30 minutes) it is best to use emergency medication (buccal midazolam or rectal diazepam) when treating a child with Sturge-Weber syndrome who is having a seizure, to reduce the likelihood of entering prolonged or repeated seizures or status epilepticus?

b) whether delaying treatment with emergency medication after seizure onset in children, or being unable to stop seizure activity, increases the risk of learning disability?

What did we do?

We searched the NHS Evidence website, NICE guidelines, and PubMed.

What did we find?

- Sturge-Weber syndrome is a neurological condition characterised by a port-wine stain birthmark on the face, seizures, hemiplegia, glaucoma, and learning disabilities. Most children with Sturge-Weber Sydrome (SWS) develop seizures by the age of 2 years.[1]

- There is more information about Sturge-Weber syndrome and epilepsy on these websites:

www.ninds.nih.gov/disorders/sturge_weber/sturge_weber.htm

www.sturgeweber.org.uk

www.nhs.uk/Conditions/Epilepsy.

- Status epilepticus is a period of prolonged convulsive seizure activity lasting more than 30 minutes, or two or more discrete

seizures without a return to normal neurological function in between.

Is there any evidence for how long after seizure onset (i.e. 5, 10, 30 minutes) it is best to use emergency medication (buccal midazolam or rectal diazepam) when treating a child with Sturge-Weber Syndrome who is having a seizure, to reduce the likelihood of entering prolonged or repeated seizures or status epilepticus?

- Sturge-Weber Syndrome (SWS) is a rare neurological syndrome so there is limited evidence from research. The majority of studies are retrospective series, and there is information from expert opinion.

- Estimates suggest that between 40-83% of children with SWS do not respond to seizure treatment.[1-3] This increases the likelihood of entering status epilepticus.

- NICE has published guidance on treating seizures in children and young people.

- The NICE guidance for treating prolonged convulsive seizures states that emergency medication should be given after five minutes, to try to stop the seizure and prevent the likelihood of entering status epilepticus.[4]

- The NICE guidance also states that children who have frequent episodes of seizures may have individually tailored treatment regimens which should be followed when they have a seizure. It may have been found that they respond better to some medications than others.

- There is no rationale for suggesting that children with SWS should receive treatment for acute seizures at a longer duration than other children with epilepsy. Available evidence supports the early treatment of seizures as an important aspect of management.[5]

Is there any evidence that delaying treatment with emergency medication after seizure onset in children, or being unable to stop seizure activity, increases the risk of learning disability?

We found no experimental studies specifically evaluating the 'timing' of emergency medication for seizures for children with Sturge-Weber Syndrome.

The effects of status epilepticus (a seizure lasting longer than 5 minutes) depend on the cause of the seizure, the age at which the child first experienced seizures, and how long the seizure lasts. [6]

It is difficult to say whether learning disability in later life is related to underlying conditions, or a direct effect of status epileptics.[6, 7]

There is evidence that early onset, hard to control seizures in children with Sturge-Weber Syndrome may predict more severe neurological impairments.[8, 9]
Some research suggests that prolonged or frequent seizures may contribute to poor cognitive outcomes, and for that reason it is important that parents and carers of children with SWS should be aware of how to recognise and promptly treat seizures.[8]

What do we think?

We found no evidence specifically regarding the most effective time at which to administer emergency seizure medication specifically in children with Sturge-Weber Syndrome.

However NICE guidance states that emergency medication should be administered after five minutes of seizure activity, and that children who have frequent episodes of seizures may have individually tailored guidelines. This guidance is relevant to children with SWS.

The relationship between effects of duration of seizure and outcomes such as intellectual ability is unclear. There is evidence to suggest that prolonged or frequent seizures may be associated with poorer neurological outcomes.

Please email your feedback on this summary to pencru@exeter.ac.uk

References

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