



**Down's Syndrome
Association**

A Registered Charity No. 1061474

Health Series



Epileptic Spasms in Children



The
Down Syndrome
Medical Interest
Group

A Down's Syndrome Association publication

Our resources and Information Team are here to help

Please see our website for up-to-date information: www.downs-syndrome.org.uk

If you would like to talk about any of the issues raised in this resource, then please get in touch with our helpline by calling 0333 1212 300 or by emailing us on info@downs-syndrome.org.uk.

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The Down's Syndrome Medical Interest Group (DSMIG)

This resource has been produced in collaboration with the Down's Syndrome Medical Interest Group (DSMIG).

DSMIG was launched in 1996 and is a registered charity. It is a network of healthcare professionals – mainly doctors – from the UK and Republic of Ireland whose aim is to share and disseminate information about the medical aspects of Down's syndrome and to promote interest in the specialist management of the syndrome.

Epileptic Spasms (sometimes called Infantile Spasms, West syndrome or Salaam attacks) are uncommon epileptic seizures which can occur in infants with Down's syndrome. They also occur in infants without Down's syndrome, occurring in less than one in every 2000 infants. They are more likely to occur in children who have some difficulties with their development. Epileptic Spasms occur in around 5% of children with Down's syndrome, making them much more likely to occur than in other children.

What will I see?

Epileptic Spasms usually occur in children aged less than 12 months. They most often start between 3 and 6 months old. Families may notice that the child's head will suddenly nod forwards. Sometimes the arms or legs will also quickly move and the child may become upset. The movements are very brief (lasting less than a second), but may occur in "clusters" where several of these movements occur close together. These clusters may happen after the child wakes from sleep. Some families also notice that their child's development pauses or slows at this time, or that the child becomes less responsive and stops smiling as much.

What else could it be?

Small babies may make unusual movements for many different reasons. Some babies will have an exaggerated "startle" reflex, which might be seen if there is a loud noise and the baby "jumps". Some babies may arch their backs or feel uncomfortable if milk comes back up from their stomach. This is called "reflux", and commonly occurs in babies with Down's Syndrome.

Many children, particularly those who have some difficulties with learning, will have unusual movements from time to time. Some babies will have movements similar to those seen in Epileptic Spasms but the tests are normal. These children may be given a diagnosis of "benign infantile myoclonus". This is a condition which will not require any treatment and will get better as the child grows.

If there are movements that you are concerned about, your doctor may find it useful to see a video recording of them. If you see unusual movements try to write down a description, with a date and time, straight after you see the movements. Some parents find it helpful to think about what happened just before the episode (was the baby feeding/sleeping/playing etc.?), what happened during the episode (think about your child's face/colour/arms/legs/eyes/eyelids/body), and what happened straight after the episode (was the child upset/drowsy etc.?). It is also useful to record what the episode looked like, how long it lasted, and whether there were several episodes close together.

What should I do if I'm worried?

If you see movements like this in your child you should see your GP or family doctor, usually within the next few days. Give them a description of the movements and explain that you are concerned about Epileptic Spasms. These movements can be very subtle, and it may be quite hard for the doctor to work out if these are normal baby movements or are worrying so the more information you can give your doctor the better. If your doctor is also concerned, they will refer you to a specialist. Initially this will usually be a Paediatrician (doctor specialising in children) and then a Paediatric Neurologist (doctor specialising in children's brains and nerve diseases). You would usually be seen by the specialist the same or next day. Most children will need to stay in hospital during this time to begin tests and treatment.

What tests may be carried out?

The test to look for Epileptic Spasms is a brain wave test called an electroencephalogram (or EEG for short). This test involves putting lots of sensors on your child's head with either glue or a cap and looking at the brain wave traces. It is not painful but it can be difficult, especially if your child is very active. This test happens in hospital and usually takes around an hour. It is usually helpful if the child has a sleep or nap during the test, if they are able. Sometimes the specialists will ask to video the child during the EEG to record the movements during the brain wave tracing. In some children with Epileptic Spasms the EEG will show a jagged appearance, which is called "hypsarrhythmia" (pronounced hips-a-rith-me-a). This will confirm the diagnosis of Epileptic Spasms, and children may also be described as having "West Syndrome". Some children may have an abnormal brain wave trace which is not typical, or the test may need to be repeated, including during a period of sleep.

Epileptic Spasms can be related to a range of underlying conditions, including Down's syndrome. For infants where there has been no previous concern about their development, the Epileptic Spasms may be the first sign that there is a developmental problem, and a numbers of tests will be arranged to try and find out what the underlying problem may be. For a child with Down's syndrome, we already know what the underlying condition is, but blood tests and imaging of the brain (e.g. MRI scan) may be recommended.

What treatment is available?

We know that children appear to have better outcomes when they are given treatment for Epileptic Spasms as soon as possible (ideally within days of the diagnosis being confirmed). Sometimes two medicines are given together.

One treatment that is often started in the UK is a type of steroid. Steroids are a substance that all of our bodies naturally produce to help us respond to stress. This medicine is usually given for around 14 days only. It may be dissolved in water and given with a syringe into the child's mouth several times a day, or occasionally given by injection (usually into the side of the leg) every other day.

Another drug, sometimes used together or instead of the steroid medicine, is called Vigabatrin. This is a medicine also used for other types of Epilepsy. It usually comes in a sachet which can be mixed with a drink and taken twice a day. This medicine is sometimes taken for up to several months.

These medicines are usually given for a short period of time, and the EEG may be repeated when your child is on treatment. Sometimes families will stop seeing the abnormal movements when a child is on treatment but the EEG may still be abnormal. Sometimes these medicines do not work and other treatments may have to be tried. The medical team will need to monitor your child closely to look for any possible side effects of any medicine used.

Will my child get better?

Some children with Epileptic Spasms will recover fully, having no further seizures and no additional problems with their development. Some children will continue to have seizures as they grow older. The seizures seen in these children may look different to the spasms that they had when they were babies.

Some children with Epileptic Spasms may go into "remission" when the seizures stop, but sometimes the seizures will then restart. Any of these scenarios may occur in a child with Down's syndrome, but in many cases the response to treatment is good, the spasms disappear quickly with treatment, and the seizures do not recur.

Children who do not have Down's syndrome who develop Epileptic Spasms often go on to have problems with their development and long-term learning difficulties. Children who have Down's syndrome will already be expected to have some difficulties with their learning. Doctors will need to monitor your child's development as they get older as some children who have a second diagnosis of Epileptic Spasms will go on to need more support and help with their learning than is typical for children with Down's syndrome. In some cases, a co-diagnosis of Autism Spectrum Disorder may subsequently be made.

What about other types of epilepsy and Down's syndrome?

People with Down's syndrome are more likely than those without Down's syndrome to develop epilepsy in their lifetime. Epilepsy is a medical term to describe when the cells in a person's brain have a tendency to give off abnormal electrical activity. As our brains control our bodies, this abnormal electrical activity may result in the person having unusual movements or sensations (sometimes called a "seizure"). There are lots of different types of epilepsy (so some people prefer the term "the Epilepsies") because there are lots of different ways in which the brain cell electrical activity can impact on our bodies.

It is estimated that around 1 in every 10 people with Down's syndrome will develop epilepsy at some point in their lives, compared to around 1 in every 100 people who do not have Down's syndrome. Just under half of the people with Down's syndrome who have epilepsy develop it when they are less than a year old, mainly with Epileptic Spasms, but other types of seizure can occur, most often in teens or early adult years, though they can also occur in later life. Epilepsy occurring at any age needs medical assessment, and medicines may be offered to try and control it.

GP's learning disability register

People with learning disabilities experience poorer health compared to the rest of the population, but some ill health is preventable. Over one million people in the UK have a learning disability but only 200,000 are on their GP's learning disability register.

We know that people with a learning disability often have difficulties accessing health services and face inequalities in the service they receive. The Government is asking parents and supporters to speak to their GP and ensure their sons/daughters or the people whom they support are registered. It is hoped that this drive will ensure better and more person centered health care for people with learning disabilities.

The Learning Disability Register is a record of people with a learning disability who are registered with each GP practice. The Register is sometimes referred to as the Quality Outcomes Framework (QOF) Register.

If you are not sure you are on the Register, you can ask the receptionist at your GP Practice to check for you.

The doctor may have made a note on the record that a person has Down's syndrome but this does not automatically mean they have been put on the Register. When you speak to the GP about being registered, the needs and support of the person in health settings can be discussed. This information can be entered on the person's Summary Care Record (SCR) so that all health professionals at the practice know about their needs and how best to support them.

If the person is over 16 years of age or older, they must give their consent (see section in this resource about the Mental Capacity Act 2005):

- for information about their support needs to be added to their SCR
- to which information can be shared and with whom

It's never too early (or late) to join your GP's Learning Disability Register; you can join at any age. It's a good idea for children with a learning disability to join the learning disability register at an early age. This means adjustments and support can be put in place before they reach adult

Reasonable adjustments in health care

You may have heard of the term 'reasonable adjustments' and wondered what it meant. Since the Disability Discrimination Act (1995) and the Equality Act (2010) (this does not apply to Northern Ireland) public services are required by law to make reasonable adjustments to help remove barriers faced by people with disabilities when trying to use a service. The duty under the Equality Act to make reasonable adjustments applies if you

are placed at a substantial disadvantage because of your disability compared to people without a disability or who don't have the same disability as you.

So for people with physical disabilities reasonable adjustments may include changes to the environment like ramps for the ease of wheelchair users. For people with learning disabilities 'reasonable adjustments' may include easy read information, longer appointments, clearer signs at the practice, help to make decisions, changes to policies, procedures and staff training.

If a patient with Down's syndrome is NOT on their GP's Learning Disability Register, then reasonable adjustments to care for that person cannot be anticipated and made.

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The Down's Syndrome Association provides information and support on all aspects of living with Down's syndrome.

We also work to champion the rights of people with Down's syndrome, by campaigning for change and challenging discrimination.

A wide range of Down's Syndrome Association publications can be downloaded free of charge from our website.

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