

Infantile Spasms

Infantile spasms are a type of epilepsy. You may also hear infantile spasms called West syndrome after Dr. William West, who described infantile spasms in his own son in the 1840s.

What are infantile spasms?

Infantile spasms usually begin in the first year of life. Spasms are a type of seizure. When your baby has a spasm, any of the following things may happen:

- arms and legs jerk up quickly
- head drops forward
- arms fly out
- legs go up to the chest
- muscles become stiff and then relax
- eyes look to the side or up
- breathing changes

Babies experiencing infantile spasms will sometimes look as though they have severe stomach pain or colic, but they are not in pain. Spasms usually happen when the baby is waking up, but they can happen at other times. The baby may have one spasm at a time, or many spasms in a row. Many spasms in a row are called a cluster. Each spasm usually lasts less than 10 seconds. A cluster or severe spasms can last for minutes.

After a spasm, the baby may cry or laugh. The spasms do not cause pain, but the baby may cry because the quick jerking movement surprises them.

After infantile spasms start, the baby may stop developing

Once the spasms start, parents often notice that their babies stop developing. They stop doing

the things they did before, such as smiling, rolling over or sitting. They may also stop following things with their eyes (tracking) and looking at people in the eyes. They do not learn to do new things.

What causes infantile spasms?

Infantile spasms can happen because of many different problems. Sometimes they happen in babies who experience delayed development. These babies may already have a known problem with their brains. These problems can start:

- Before they are born.
 - If the brain does not form properly during fetal development
 - If there is a chromosomal abnormality that causes a disorder that affects the brain, such as Down Syndrome or Tuberous Sclerosis, and increases the risk of getting infantile spasms
 - If there is a problem with the body's metabolism, causing too much or too little of necessary body chemicals
 - If there is damage to the brain from a clot or bleeding in the brain or an infection
- Around the time they are born.
 - If they do not receive enough oxygen at birth
 - If they experience trauma during birth
- In the first few months of life.
 - Due to brain infection
 - Due to brain injury

Sometimes babies are developing normally when they start having infantile spasms and the cause is unknown.

Infantile spasms are divided into 3 groups:

- **Symptomatic:** When we know what has caused the spasms.
- **Cryptogenic:** When a baby had problems with development before the spasms started, but we cannot find a cause.
- **Idiopathic:** When a baby's development was normal before the spasms started and we cannot find a cause.

Signs and symptoms of infantile spasms

Your doctor will diagnose your baby with infantile spasms if they experience all of the symptoms below:

- Your baby has the type of seizures usually seen in infantile spasms that are described above.
- Your baby's EEG (electroencephalogram) test shows a pattern of brain waves called hypsarrhythmia.
- Your baby has stopped developing or has stopped learning to do new things since the seizures started.

Tests to diagnose infantile spasms

Your baby will need various tests before and after being diagnosed with infantile spasms.

Electroencephalogram

Your baby will have an EEG. This test looks at the patterns of electricity that the brain cells make. It does not hurt.

A technologist will put small gold discs called electrodes on your baby's scalp. Sometimes your

baby will need medicine to sleep during the EEG.

The EEG helps the doctor decide if your baby has infantile spasms. The EEG will also be used to check how well the treatment is working.

CT scan and MRI of the brain

Most babies with infantile spasms will have a CT scan or MRI of the brain.

- A CT scan is a special X-ray that uses a computer to take pictures of the brain.
- An MRI uses a magnet, radio signals and a computer to make pictures of the brain.

These pictures help the doctors see if your baby's brain looks normal or not. This may help the doctor find the cause of your baby's infantile spasms.

Other tests

The doctors may also do other tests to look for the cause of the infantile spasms:

- The doctors may test your baby's blood and urine to look for infections and chemical changes.
- If the doctors cannot find the cause of the infantile spasms using these tests, they may do a blood test to look at your baby's genes to see if there are any abnormalities.
- The doctors may look at your baby's skin using a Woods lamp. Some conditions that cause infantile spasms also cause changes in the skin. This lamp uses a special light to show these changes.

Because there are so many causes of infantile spasms, the tests will be different for each baby. The doctors and nurses will explain any other tests your child may need.

Infantile spasms are treated with medicine

Two medicines are the best at stopping infantile spasms. One is called vigabatrin (Sabril) and the other is called ACTH (Synacthen).

At SickKids the doctor will probably put your baby on vigabatrin first. Usually, after 2 to 3 weeks of treatment, your baby will have an EEG and be seen by the neurologist. If the spasms have not stopped or the hypsarrhythmia pattern on the EEG does not go away, the neurologist may decide to start your baby on ACTH or sometimes another medicine called prednisolone or another anti-seizure medicine.

Treatment will stop the infantile spasms in most children, but in some children the spasms will not stop.

Vigabatrin

Vigabatrin (Sabril) is medicine that is given by mouth. It comes as a pill that can be crushed and mixed with small amounts of liquid such as a spoonful of water, milk, or juice, or a spoonful of food such as apple sauce or baby cereal. It also comes as a powder called a sachet that can be dissolved in a liquid. Make sure that your baby takes all of the liquid or food so that he or she gets all of the medicine. For suggestions on how to give the medicine to your baby see the AboutKidsHealth Medicines Video: How to Give Your Child Liquid Medicines at Home. Keep the medicine at room temperature in a dry place.

The amount of medicine you give is increased each day over a few days to a very high dose. If the vigabatrin stops the spasms, your baby will stay on it for at least 6 months. Your doctor will

tell you when your baby is ready to stop taking the medicine.

Here is what to do if your baby throws up the medicine or if you forget a dose:

- If your baby throws up the medicine within 30 minutes after taking it, give the dose again.
- If your baby throws up more than 30 minutes after taking the medicine, do not give any more until the next time it is supposed to be given.
- If the medicine is not given on time, give it as soon as you remember or can, unless the next dose is scheduled within 4 hours.
- If the next dose of the medicine needs to be given in less than 4 hours, do not give the dose you forgot. Give the next dose a little earlier and then return to giving the medicine at the usual times after that. Do not give 2 doses at the same time.

Side effects of vigabatrin

Your baby may have some of the following side effects:

- changes in how well the baby can see, in particular loss of peripheral or side vision, which is sometimes called tunnel vision
- wanting to sleep more than usual, especially in the first few days to 3 weeks of treatment
- upset stomach
- constipation
- hyperactivity, restlessness, always wanting to move
- being awake a lot in the middle of the night
- irritability, fussiness, hard to make happy
- weight gain
- arms and legs become floppy, also called decreased muscle tone or hypotonia

Not all babies get these side effects. Your baby may get some of these side effects and not others.

A note on tunnel vision

Doctors at The Hospital for Sick Children have monitored many children taking vigabatrin to see how vigabatrin affects their eyesight. They have found that:

- Changes in eyesight because of vigabatrin happen in fewer than 10% of children who are younger than 2 years old.
- It is very rare to see eye changes in children who have been taking vigabatrin for less than 6 months.
- Serious changes to the eyes are more likely to happen after 1 to 2 years of treatment, but they are still uncommon.
- Most children will not have their eyesight changed by vigabatrin.

How to deal with side effects of vigabatrin

- **Changes in eyesight:** Your baby will have an eye test called an electroretinogram (ERG) within a short time after starting on vigabatrin. Vigabatrin can sometimes cause narrowing of the peripheral vision, which means that a child cannot see as well to the side. The ERG will help the doctor know if this is a problem for your child. An ERG will be done every 3 to 6 months while your child is taking vigabatrin to see if there are any changes. One more ERG will be done after your child stops taking vigabatrin.
- **Sleepiness:** Your baby will become less sleepy over time as he or she gets used to the medicine. Make sure your baby gets enough to eat and drink. Wake your baby up if it is

time to feed him or her. If your baby is very sleepy and will not eat or drink, call the doctor who gave you the prescription for vigabatrin.

ACTH

ACTH (Synacthen) is given by needle into the leg. We will arrange for a nurse to come to your home to give this medicine to your baby.

ACTH is started at a high dose. At SickKids the dose is lowered slowly over 6 weeks and then the treatment is stopped.

The side effects are usually worse at first when the dose is higher. The side effects will go away after the ACTH is stopped.

The nurse must check your baby's blood pressure before giving the medicine. Your baby's blood needs to be tested every 1 to 2 weeks and the urine tested 2 times a week.

Sometime doctors will suggest using prednisolone instead of ACTH. It is a similar type of drug as ACTH and causes the same side effects, but the dose is not as high and it is given by mouth.

Side effects of ACTH

Your baby may have some of the following side effects:

- increased chance of getting a serious infection
- not sleeping well, although some babies are more sleepy and lethargic
- irritability: your baby may be very fussy and hard to calm down
- hunger or wanting to eat more
- weight gain
- puffy-looking face

- upset stomach, such as vomiting (throwing up) or stomach pain
- high blood pressure
- changes in the chemicals of the body, such as high sugar or high salt
- acne: red, raised bumps on the face
- tremor (mild shaking)
- rarely, thinning of the bones that makes them easy to break
- very rarely, thickening of the heart muscle

Not all babies get these side effects. Your baby may get some of these side effects and not others. These side effects will go away after the ACTH or prednisolone is stopped.

How to deal with the side effects of ACTH and prednisolone

- **Chance of a serious infection:** Wash your hands well with soap before touching or holding your baby. You may want to buy soap that has 'anti-bacterial' written on the label. Ask everyone who comes into your home to wash his or her hands. Anyone with an infection such as a cold or flu should not touch or hold your baby. We suggest that your baby not be in a day care while taking ACTH. It is very important that you take your baby to see your paediatrician or family doctor right away if he or she shows any signs of an infection. Some signs of infection are fever, runny nose, cough, diarrhea (watery bowel movements), bad smelling urine, and change in alertness. For more information, please see "Immunosuppression: Protecting Your Child from Infection."
- **Severe irritability or fussiness:** If it is really hard for your baby to sleep, the doctor may suggest you give your baby a medicine to help him or her sleep at night. You will be given a prescription for this. Ask for help from family or friends to take turns caring for your baby so that you can get some rest. Hold, rock, swing, give warm baths, play music, or do anything that seems to make your baby feel better. Sometimes, no matter what you do, your baby will not stop crying or being fussy.
- **Increased appetite:** Feed your baby the type and amount of food that is usually fed to a baby your child's age. Ask your baby's nurse for the information on 'What to Feed Your Baby in the First Year of Life' or speak to your paediatrician for information about the type and amount of food that is best for your baby. We can also arrange for you to speak with a dietitian.
- **Upset stomach:** The doctor will give you a prescription for a medicine that will help settle your baby's stomach.
- **High blood pressure:** The nurse will check your baby's blood pressure before giving ACTH. If your baby is taking prednisolone then their blood pressure should be checked at least once a week by your paediatrician. Blood pressure is a measurement of how strongly and how often the heart pushes blood around the body. If the blood pressure is above a certain level, the doctor may have you give a medicine to your baby that lowers the blood pressure.
- **Changes in chemicals of the body:** Your baby's blood will be tested every 1 to 2 weeks. Your baby's urine will be tested 2 times a week. The blood can be tested through your paediatrician or family doctor.

The nurse who comes into your home will test the urine. The doctor will give you a prescription for medicine to treat any problems.

It is very important to treat your child to stop infantile spasms

All children stop having infantile spasms over time, usually by 2 to 3 years of age. But if a child is not treated, he or she will stop developing. The child will not learn any new things. This will cause problems with development that last forever. Treatment that stops the infantile spasms will give your child the chance to learn and develop. This is why it is very important to give treatment to stop the infantile spasms and make the brain wave (EEG) pattern as normal as possible.

Whether your child will get better depends on the cause of the infantile spasms

Whether a child gets better depends on:

- the cause of the infantile spasms
- how well the treatment works

If a child has a symptomatic (known) cause for infantile spasms or the spasms do not stop with treatment, they are more likely to have learning problems and development problems that continue for the rest of their life. If the cause is cryptogenic or idiopathic (not known), the child has a greater chance of normal or almost normal development.

Your child may have other types of seizures later in life

Many children who have had infantile spasms will get other types of seizures. Most children

with symptomatic infantile spasms will develop other types of seizures later on.

You may need to delay your baby's immunizations

Talk to the neurologist about when your baby can have immunizations. After your baby starts taking medicine for infantile spasms, you may need to delay the regular schedule of immunizations:

- Sometimes doctors do not want a baby to have an immunization until they know whether the medicine has stopped the spasms or not.
- Vaccines will not work while a child is taking ACTH, and sometimes for many months after they have stopped taking the medicine.

You can help your baby with infantile spasms

Here are some things parents can do to help their baby with infantile spasms:

- Write down the number of spasms your baby is having and what they look like. This will help the doctor decide how well the medicine is working.
- Give your baby his or her medicine at the time suggested by the doctor.
- Play with and talk to your baby to help him or her learn new things and develop.
- Take your baby to all medical appointments.
- Make sure that doctors and pharmacists know your baby is taking vigabatrin or ACTH before starting on any other medicines.
- Look after yourself. Eat well, ask others for help and get enough sleep. If you stay healthy, you are better able to meet your child's needs.

When should I call a doctor or nurse?

Call your baby's neurologist or Neurology Clinic nurse if:

- your child has side effects to the medicine that are hard to deal with
- your child is not eating or drinking enough
- your child is sleeping most of the day and is hard to wake up
- your child shows any signs of an allergic reaction, such as hives, rash or wheezing
- your child has more spasms than usual

Call your baby's paediatrician or family doctor if your baby has any signs of infection or you have any other worries about your baby's health.

Resources to help you cope

There are many people who can help you.

- Social workers can support you and your family to help you adjust to your child's condition. They can also help with getting community supports and finding money to help pay for your child's medicine.
- Occupational therapists help with the development of your baby. They will check how your baby is developing, give treatment as needed and teach you how to help your baby learn new things. We often call this infant stimulation.
- Family, friends and your religious community may help support you in different ways.

- Support groups such as the West Syndrome Support Group in the United Kingdom (<http://www.wssg.org.uk/wssg>) or a parent mentor program (<http://www.infantilespasmsinfo.org>) can be found on the Internet. Being in touch with other parents who also have a child with infantile spasms may help.

Key Points

- Infantile spasms are a type of epilepsy that usually begins in the first year of life.
- Infantile spasms do not cause your baby pain, but they must be treated as soon as possible, or your baby will stop developing and learning new things
- After being diagnosed with infantile spasms, your baby may need tests to help find the cause of the infantile spasms. Your baby may need to have an MRI of the brain or have blood and urine tested for chemical changes or changes in the genes.
- Your baby's infantile spasms will be treated with medicine. Your baby will be treated with vigabatrin (Sabril) first. If vigabatrin does not work then your baby will be treated with ACTH (Synacthen) or prednisolone or another anti-seizure medicine.
- Many babies who have infantile spasms will develop other seizures later in life.