Infantile Epileptic Spasms Syndrome

What Is Infantile Epileptic Spasms Syndrome?

Infantile epileptic spasms syndrome is a rare but serious type of epilepsy. It's associated with a unique, very abnormal brain wave pattern that can cause serious harm to a young, developing brain. This type of epilepsy can happen during the first 2 years of life, most commonly between 5-7 months of age. Babies with the condition also might have slowed development or loss of skills (like babbling, sitting, or crawling). The epileptic spasms can be very hard to control, and many children go on to have other kinds of seizures later in life.

The term West syndrome was used when a baby has infantile spasms, a unique EEG pattern (called hypsarrhythmia), and developmental delay. In 2022 the International League Against Epilepsy updated the nomenclature of several childhood epilepsy syndromes and the term infantile epileptic spasms syndrome includes patients with West syndrome and those presenting epileptic spasms who do not fulfill all the criteria for West syndrome.

What Do Epileptic Spasms Look Like?

Epileptic spasm is a type of seizure that starts suddenly and lasts a second or two. They often come one after another in a cluster that lasts several minutes. They happen most often just after a child wakes up.

A baby having an epileptic spasm might have:

- The head bent forward with arms flung out and the knees pulled into the body (described as "jackknife")
- The head bent back with the arms and legs straightened
- Small movements in the neck or other parts of the body, such as the eyes widening and rolling up

What Causes Infantile Epileptic Spasms Syndrome?

Infantile epileptic spasms syndrome can be caused by brain malformations, infections, perinatal brain injury, etc. Epileptic spasms also can happen in babies with some types of metabolic and genetic disorders. However, sometimes the etiology is not identified.

How Is Infantile Epileptic Spasms Syndrome Diagnosed?

Infantile epileptic spasms syndrome is diagnosed by a pediatric neurologist. Testing may include:

- Blood tests and urine tests (to look for infections or illnesses)
- EEG (it usually shows hypsarrhythmia, but not every child with epileptic spasms will have this finding).
- VEEG, or video electroencephalography (EEG with video recording)
- MRI

How Is Infantile Epileptic Spasms Syndrome Treated?

Infantile epileptic spasms syndrome usually is treated with seizure medicines or steroids. If medicines don't control the spasms, a special diet, such as the ketogenic diet, might help. Sometimes, doctors may recommend epilepsy surgery.

What Problems Can Happen?

Most children with infantile epileptic spasms syndrome will have problems with their development and may need help with daily tasks throughout their lives.

How Can I Help My Child?

Caring for a child with infantile epileptic spasms syndrome can be challenging. Work with your child's care team to set up the medical visits, therapies and a treatment plan that provides your child with a good quality of life.

Make sure that you and other adults and caregivers (family members, babysitters, teachers, coaches, etc.) know what to do if one happens. Your doctor may prescribe an emergency medicine to give if your child has a long seizure or many seizures in a short amount of time. Be sure to ask your doctor about a seizure rescue plan for your child

What Else Should I Know?

If your child has epilepsy, your doctor and the care team can answer questions and offer support. They also might be able to recommend a local support group. Online organizations can help too, such as:

- Epilepsy Foundation
- CDC – Managing Epilepsy
- Infantile Spasms Action Network (ISAN)