



Infantile Spasms

Facts

Infantile Spasms (IS) is a devastating epilepsy syndrome that usually strikes infants between three to six months old.¹ Children with IS can experience hundreds of spasms per day.¹ Of children who develop IS, 85 percent do so before the age of 12 months² and as many as 20 percent of infants affected die before age five.² The spasms that occur in IS are often initially mistaken for colic because the babies double up during a spasm and typically cry afterwards.² While spasms usually cease by age five, more than half of children with IS develop other types of seizures.¹

General Facts

- ◆ IS, also known as West syndrome, is a condition that typically includes the triad of infantile spasms; a characteristic abnormal, chaotic brain wave pattern on EEG; and mental retardation.³
- ◆ In the United States, IS constitutes about two percent of childhood epilepsies, but 25 percent of epilepsies with onset in the first year of life.³
- ◆ There are an estimated 8,500 infants in the United States diagnosed with IS.⁴
- ◆ Spasms frequently occur just before sleep or upon awakening.³
- ◆ A typical spasm, called a flexor spasm, causes the infant to bend forward and stiffen the body, arms and legs.³ This may resemble a self-hugging motion and is often followed by a cry.³ Extensor spasms, which cause the infant to arch the torso, can also be associated with symptomatic IS.³ The most common type of spasm incorporates both flexor and extensor spasms.³
- ◆ Infants suffer spasms that typically last for one to five seconds and occur in clusters of up to 100 spasms at a time.¹
- ◆ IS has a devastating effect on neurological development of the child. Children with uncontrolled spasms suffer impaired nervous system development and may never attain critical developmental milestones and frequently regress from previously established milestones.⁵

Epidemiology

- ◆ There are three classifications for IS: Symptomatic IS, cryptogenic IS and idiopathic IS with symptomatic IS being the most common and having a worse prognosis than cryptogenic or idiopathic IS.³
- ◆ The most common causes of symptomatic IS are Tuberous Sclerosis Complex (TSC), a genetic condition characterized by seizures and lesions of the skin and nervous system, and perinatal asphyxia (lack of oxygen to the brain) usually caused by a drop in maternal blood pressure or interference during delivery with blood flow to the infant's brain.³ Ten to 30 percent of IS cases are caused by TSC.³
- ◆ Approximately one-fifth of children with IS develop Lennox-Gastaut syndrome (LGS)⁵, a severe, catastrophic childhood epilepsy characterized by multiple types of seizures, behavioral disturbances and developmental delay.⁶

Diagnosis and Treatment Goal

- ◆ The electroencephalogram (EEG) of a child with IS will typically reveal a characteristic abnormal brain wave pattern called hypsarrhythmia. This chaotic, random, high-voltage pattern that arises from multiple areas of the brain and spreads is often useful in confirming the diagnosis.⁵
- ◆ The primary goal of any treatment for IS is spasm cessation to allow neurological development to resume.³

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Sources:

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