Epilepsy with Myoclonic Atonic Seizures (Doose Syndrome)

What Is Epilepsy with Myoclonic Atonic Seizures?

Epilepsy with myoclonic atonic seizures is a rare type of epileptic encephalopathy. Children with the condition will have:

- Different types of seizures
- Intellectual disability and developmental delay can occur in some patients
- Abnormal findings on an EEG

Epilepsy with myoclonic atonic seizures begins between 2- and 6-years-old, and the course of the disease is variable. Some patients have seizures controlled by medication, but a large number of children will develop intractable epilepsy with some degree of intellectual disability and developmental delay.

What Kinds of Seizures Happen in Epilepsy with Myoclonic Atonic Seizures?

There are different seizure types in epilepsy with myoclonic atonic seizures. They can cause a variety of symptoms.

Generalized tonic-clonic seizures:

In this type of seizure, a child:

- Has convulsions, or rigid muscles and rhythmic body jerks
- Rolls the eyes back
- Cries out
- May pee or poop
- Can't respond during seizure
- Is confused and sleepy after the seizure

Atypical absence seizures:

In this type of seizure, a child:

- Blanks out or has staring spells that last 5–20 seconds
- May flutter their eyes or look upward
- Is unaware of what is going on during the seizure
- Returns to normal activity and won't remember having the seizure after it’s over
- Has lip-smacking, chewing movements

Myoclonic seizures:

In this type of seizure, a child:

- Has brief muscle twitches or jerks in the upper arms, shoulders, or neck
- Has movements on one or both sides of the body at the same time

usually is awake and can think clearly during and right after the seizure

Atonic seizures:

In this type of seizure, a child:

- Has a sudden fall (sometime just a head drop)

Myoclonic-Atonic seizures:

In this type of seizure, a child:

- Has a brief jerk involving neck, shoulder and limbs followed by a fall (loss of muscle tone)

What Causes Epilepsy with Myoclonic Atonic Seizures?

The cause of epilepsy with myoclonic atonic seizures has not been established yet, but it is probably caused by genetic abnormalities.

How Is Epilepsy with Myoclonic Atonic Seizures Diagnosed?

A pediatric neurologist (a doctor who treats brain, spine, and nervous system problems) can diagnose the condition by doing tests such as:

- EEG
- VEEG, or video electroencephalography (EEG with video recording)
- MRI

How Is Epilepsy with Myoclonic Atonic Seizures Treated?

Seizures in epilepsy with myoclonic atonic seizures are treated with antiseizure medications. In addition, children with epilepsy with myoclonic atonic seizures are often good responders to the ketogenic diet.

How Can Parents Help?

Caring for a child with epilepsy with myoclonic atonic seizures syndrome can be challenging. Work with your child's care team to set up medical visits, therapies, and to create 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 during a seizure. Your child may need to wear a helmet to prevent head injury during seizures.

Because it could lead to a tonic-clonic seizure, 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