

Juvenile myoclonic epilepsy is a fairly common epilepsy syndrome that usually begins when a child is between 12 and 18 years old. The child will have one or several myoclonic jerks, usually shortly after waking up. About three years after the myoclonic jerks begin, most children with juvenile myoclonic epilepsy start having generalized tonic-clonic seizures as well.

## What are other terms for juvenile myoclonic epilepsy?

Other terms for juvenile myoclonic epilepsy that you may come across include:

- adolescent myoclonic epilepsy
- Janz syndrome
- JME
- juvenile myoclonic epilepsy of Janz

## What causes juvenile myoclonic epilepsy?

Juvenile myoclonic epilepsy is an idiopathic generalized epilepsy syndrome with a strong genetic component. One-third to one-half of affected children have a family history of seizures or epilepsy. In some cases, children had febrile seizures or childhood absence epilepsy before they developed juvenile myoclonic epilepsy.

The genetic basis of juvenile myoclonic epilepsy is not completely clear. More than one gene appears to be involved, and the disorder has been linked to different genes in different families.

Citovenneté et

Immigration Canada

In rare cases, children with juvenile myoclonic epilepsy also have structural brain abnormalities, but these do not seem to affect the response to treatment or the outlook. One MRI study found that people with juvenile myoclonic epilepsy and other idiopathic generalized epilepsies had subtle differences in grey matter volume, grey matter distribution, and brain metabolism compared to people without idiopathic generalized epilepsy.

## What are the features of juvenile myoclonic epilepsy?

Children with juvenile myoclonic epilepsy usually begin having seizures when they are between eight and 24 years old, with most children starting to have seizures between the ages of 12 and 18.

The child has myoclonic jerks with sudden, brief muscle contractions on one or both sides of the body. The jerks mainly affect the shoulders and arms; if the child is holding an object, it may be thrown across the room. Occasionally the jerks may affect the legs or even the entire body, in which case the child may fall down. The child remains conscious during the seizures.

The jerks usually happen soon after the child wakes up in the morning or after a nap. They can interfere with normal activities like eating breakfast, brushing teeth, or putting on makeup. The child and her parents may assume that the jerks are normal, or that they are caused by nervousness or clumsiness. For this reason, it can be easy to overlook the seizures.







The seizures can also be triggered by early awakening, lack of sleep, alcohol, drugs, fasting, menstruation, or (in about one-third of cases) flashing or flickering lights. They may also happen when the child is tired or relaxed. One study found that complex activities such as writing, calculating, or drawing triggered seizures in 76% of people with juvenile myoclonic epilepsy.

In some cases, the seizures do not cause visible movements. The child only feels an electric shock-like feeling inside her body.

Once a child has had juvenile myoclonic epilepsy for several years, she will usually start to have generalized tonic-clonic seizures. These seizures usually begin with a series of myoclonic jerks, which become more and more severe and spread through the body. The child then has a tonic-clonic seizure. The child may be able to tell from the myoclonic jerks that she is about to have a tonic-clonic seizure, and can lie down in a safe place to avoid injury.

Between 15% and 30% of children with juvenile myoclonic epilepsy also have absence seizures, which usually begin when the child is about 11 ½ years old. Children with childhood absence epilepsy may develop juvenile myoclonic epilepsy later on.

The child's intelligence usually remains normal. However, some researchers have reported that children with juvenile myoclonic epilepsy may have an immature personality, poor social adjustment, or difficulty with tests that need mental flexibility and abstract reasoning.

Children with this syndrome may also fail to follow their course of treatment properly.

#### How many other children have juvenile myoclonic epilepsy?

Because children and their parents may not realize that myoclonic jerks in the morning are abnormal and may not tell their doctor about them, it is not certain how many children have juvenile myoclonic epilepsy. Doctors are becoming more aware of this syndrome. It is estimated that about 10% of cases of epilepsy, and perhaps more, are juvenile myoclonic epilepsy.

### How do you know that a child has juvenile myoclonic epilepsy?

A description of the seizures is very important for your child's doctor to make a diagnosis of juvenile myoclonic epilepsy. Your child's doctor will need to know what the jerks look like and if your child drops or throws things in the morning after she wakes up. He will also ask if any family members have or had seizures.

The progressive myoclonic epilepsies (PMEs), such as Unverricht-Lundborg disease and Lafora's disease, also cause myoclonic jerks, and your child's doctor will consider them as a possible cause. These conditions also cause progressive neurological deterioration with movement problems and dementia. By contrast, if a child has juvenile myoclonic epilepsy, her physical and neurological examinations are usually normal.

The child will usually have a specific EEG pattern between seizures. The doctor may want to obtain an EEG after the child has been sleep-





deprived for some time to see any changes in the EEG while the child is sleeping or tired.

If the diagnosis of juvenile myoclonic epilepsy is clear, the doctor will not usually need to obtain an MRI or CT scan.

### How is juvenile myoclonic epilepsy treated?

Juvenile myoclonic epilepsy is treated with antiepileptic drugs. For some children or adolescents, lifestyle changes may help to control seizures as well.

The anti-epileptic drug valproate is an effective treatment in 86% to 90% of people with juvenile myoclonic epilepsy. If it is not effective, or if it causes side effects, lamotrigine may be tried. Newer anti-epileptic drugs may also be useful.

Because factors such as tiredness, irregular sleep patterns, alcohol, and missing doses of medication can all trigger seizures, it is important for teens with juvenile myoclonic epilepsy to adopt regular lifestyle habits and follow their course of treatment carefully. Teens should sleep for eight to 10 hours per night and avoid staying up late or waking up early. They should avoid alcohol and large amounts of caffeine, found in coffee, tea, and cola drinks. If the child or teen is expecting a change in her sleep cycle, for instance if she is flying to a different time zone, she should talk with her doctor about techniques to prevent sleep deprivation.

Citoyenneté et Immigration Canada Juvenile myoclonic epilepsy often starts at an age when the child or teen is starting to stay up late to study, go to parties, or drink alcohol. She may feel that the syndrome and the lifestyle changes that are needed can interfere with her social life. Juvenile myoclonic epilepsy is sometimes linked with lack of compliance with treatment. For all these reasons, some doctors recommend counselling to help children and teens manage the condition.

Because juvenile myoclonic epilepsy can affect learning and social development, your child's doctor may recommend a neuropsychological evaluation

#### Nore information

Drug Therapies for Epilepsy

# What is the outlook for a child with juvenile myoclonic epilepsy?

It is rare for seizures in juvenile myoclonic epilepsy to go away. Anti-epileptic drugs control seizures for most people with this syndrome, but the relapse rate if the medication is stopped is up to 90%. For this reason, withdrawing medication is not recommended even if the child has been free of seizures for a long time. The child may need to take medication for the rest of her life.

It is possible that early treatment of the seizures may lead to better seizure control. Children with mostly myoclonic seizures and only a few tonicclonic seizures have the best chance of complete seizure control.

