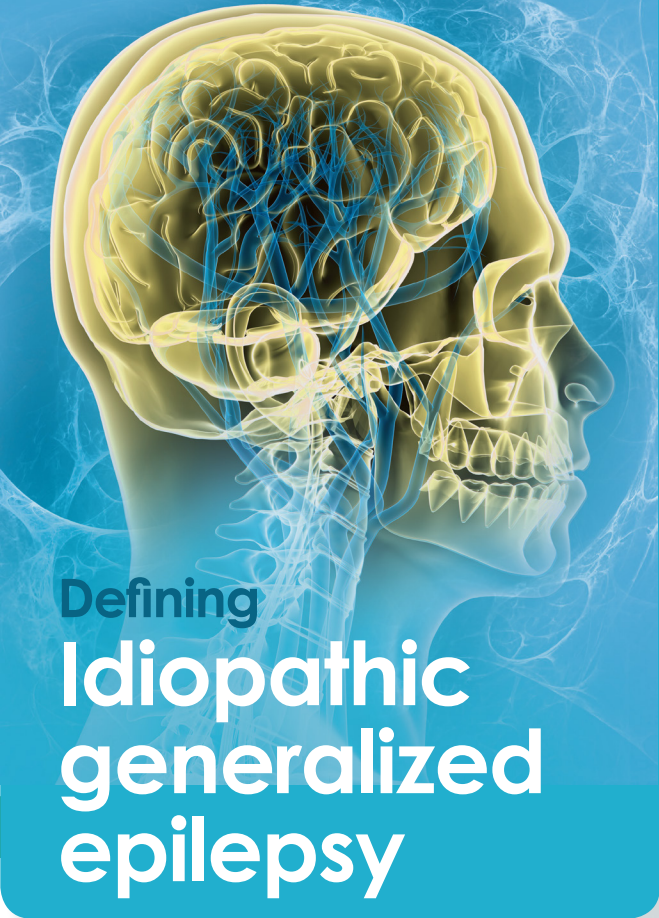


A GUIDE FOR PATIENTS & FAMILIES

WHAT IS IDIOPATHIC GENERALIZED EPILEPSY?

Idiopathic generalized epilepsy, abbreviated IGE, is a group of epilepsy that has very distinct features. It is also called “primary” generalized epilepsy. The term “idiopathic” is misleading, because it usually means “cause unknown,” which is not true here. They are genetic, and in fact were recently renamed “genetic generalized epilepsy.”



Defining Idiopathic generalized epilepsy

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CHARACTERISTICS

These epilepsies have the following characteristics:

- They are genetic and not caused by any brain physical abnormality. This means that the brain is anatomically normal.
- They basically represent a genetic low threshold (or high susceptibility) for seizures.
- There is often, but not always, a family history of epilepsy. (If everyone had 50 siblings, patients with IGE would more often have a family member affected).
- For some specific types (for example, juvenile myoclonic epilepsy) the chromosome and gene have even been identified. Others will almost certainly be identified in the near future.
- Seizures are especially sensitive to sleep deprivation
- They tend to begin during childhood or adolescence, although they may not be diagnosed until adulthood.

- People with IGE have normal intelligence, normal neurological examination, and normal MRIs.
- Although they are clearly genetic, they are not transmitted predictably like, for example, hemophilia or cystic fibrosis.

TYPE OF SEIZURES

- Patients with IGE have one or more of 3 types of (primary generalized) seizures: myoclonic, absence and generalized tonic-clonic seizures.
- One type may be the only or main type in a given patient.
- Generalized tonic-clonic (“grand-mal”) seizures are convulsions of the whole body lasting 1-2 minutes, and are the most common and most dramatic type of seizures.
- Absence seizures are brief staring spells with arrest of activity, often with eye fluttering, which last just a few seconds.
- Myoclonic seizures are very brief isolated body jerks that tend to occur in the morning.

Defining Idiopathic generalized epilepsy

HOW DO WE MAKE THE DIAGNOSIS?

- It may be difficult, and at times impossible, to make a precise diagnosis of epilepsy type (beyond just “epilepsy” or “seizure disorder”).
- Age of onset is usually early in life.
- The types of seizures seen in IGE are described above.
- Intellectual functions, neurological exam, and imaging (MRI) are normal in IGE.
- The electroencephalogram (EEG) is the only definitive test to confirm the diagnosis of IGE. Unfortunately, a single EEG is often normal in patients with epilepsy, in which case it does not help. When abnormal, the EEG in IGE is very characteristic, with various combinations of generalized spike-wave complexes, spikes, or polyspikes, sometimes triggered by flashing lights (photosensitivity).

SUBTYPES OF IGE

Depending on age at onset, predominant seizure type(s), and EEG findings, several subtypes of IGE exist:

- Childhood absence epilepsy: typically begins between 4 and 10 years of age. The main seizure type is typical absences, which are brief (about 10 seconds), but occur frequently (usually >10 daily). About 50% of patients also have generalized tonic clonic seizures, and very few also have myoclonic seizures. EEG shows a very characteristic pattern of spike-wave complexes.
- Juvenile myoclonic epilepsy (JME) typically begins in adolescence (12-18 years), and is characterized by myoclonic jerks, particularly in the morning. EEG shows a very characteristic pattern called generalized polyspikes, often with generalized 3-5 Hz spike-wave complexes.
- Epilepsy with grand-mal seizures consists of generalized tonic clonic seizures mainly, which may be more frequent in the morning. EEG can show either of the above patterns or generalized spikes.

WHY IS IT IMPORTANT TO KNOW EXACTLY WHICH TYPE OF EPILEPSY I HAVE?

- Because treatment options are not exactly the same for IGE and partial (focal) epilepsies. Some drugs work better than others for this type of epilepsy (see below).
- Because surgery is never an option for this type of epilepsy.
- Because prognosis is generally different from (better than) focal epilepsy.

TREATMENT

- The IGEs are usually easy to treat, i.e., they respond to medications in about 80-90% of patients.
- Many IGEs are outgrown in young adulthood, with the exception of JME.
- Many patients with IGE are wrongly diagnosed with “complex partial seizures” (partial epilepsy), which results in a poor choice of medications such as, for example, phenytoin (Dilantin), carbamazepine (Tegretol), or gabapentin (Neurontin).
- Many drugs are available to treat epilepsy, but they are not all appropriate for this type of epilepsy.
- Phenytoin (Dilantin), carbamazepine (Tegretol), gabapentin (Neurontin), tiagabine (Gabitril), oxcarbazepine (Trileptal) are good drugs for partial (focal) seizures, but do not work well in IGE. In fact they can make some seizures worse. This is unfortunate because phenytoin (Dilantin) and carbamazepine (Tegretol) are the most often prescribed drugs for seizures in the U.S.
- The classic drug of choice for IGE is valproic acid (Depakote).
- For patients (usually children) with absence seizures only, ethosuximide (Zarontin) is also an option.
- Other medications can be used in IGE. Phenobarbital and primidone (Mysoline) are old drugs that tend to cause sedation. Several newer drugs are also options for IGE, including lamotrigine (Lamictal), topiramate (Topamax), levetiracetam (Keppra), and zonisamide (Zonegran).
- Some newer medications have not yet been tested for IGE so it is not clear whether they should be used.

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Defining

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HOW LONG DOES TREATMENT LAST?

- With the exception of juvenile myoclonic epilepsy (JME), the epilepsies are often outgrown (roughly 50% of the time) in young adulthood, so weaning may be tried.
- JME is the only type that is not outgrown, so attempts to stop drugs in JME usually fail.

OTHER TREATMENTS

Non-drug treatments like the ketogenic diet and vagus nerve stimulation (VNS) are rarely considered in IGE, because response to medications is usually excellent, but they can be used.

As mentioned above, epilepsy surgery is never an option for IGE.

ADDITIONAL READING

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