GEFS+ (Generalised epilepsy with febrile seizures plus)

What is GEFS+?
GEFS+ is an epilepsy syndrome seen in families. Different family members can have different types of seizures, but seizures are usually generalised types and the epilepsy often starts after an initial period of febrile convulsions in early childhood.

What types of seizures are seen?
GEFS+ is characterised by an initial period of febrile seizures during early childhood, the febrile seizures may persist after the age where they usually disappear (6 years of age) and afebrile seizures may co-exist with febrile seizures or appear later in childhood. Afebrile seizures are generalised types (tonic clonic, absence, myoclonic).

GEFS+ is usually identified when the doctor recognises the pattern of an individual child having febrile and afebrile seizures and finds out from taking a careful family history that there are other family members with febrile and afebrile generalised seizures. Taking a good family history is very important in diagnosing this epilepsy syndrome. It is important that people such as grandparents are consulted to obtain the history of whether family members had seizures in their childhood.

If a young child has only had typical febrile convulsions, and there is a family history of febrile convulsions and afebrile generalised seizures, a diagnosis of GEFS+ cannot be established until the child progresses to having afebrile seizures. This is because febrile convulsions, GEFS+ and other epilepsies including epilepsy with myoclonic-ataonic seizures (Doose syndrome) and Dravet syndrome can start in similar ways and which diagnosis a child has, depends on how their seizures evolve over time.

Treatment is only required when afebrile seizures appear, as there is no good evidence that anti-seizure medications are useful for febrile convulsions. It is very important that certain anti-seizure medications are not used for GEFS+, carbamazepine can aggravate the condition and result in more seizures. If anti-seizure medications are required then valproate, topiramate and benzodiazepines are the best medications to use. Seizures can be triggered by sleep deprivation, by intercurrent illnesses (e.g. viruses) and...
in some (not all) individuals by light stimulation.

What effects are there on learning?

GEFS+ does not typically affect a child in any other way – children are expected to develop and learn normally.

What effects are there on behavior?

GEFS+ does not typically affect a child’s behavior.

What causes GEFS+?

There are several different genes that may cause the GEFS+ clinical pattern. Some of these genes can also cause other clinical epilepsy syndromes, including Dravet syndrome and epilepsy with myoclonic-atonic seizures (Doose syndrome).

How common is it?

GEFS+ is not uncommon, but is often not recognised due to the need for a good family history (often from matriarchs in the family who will recall whether family members had seizures in their early childhood).

What is the long-term outlook?

Seizures in GEFS+ are expected to improve with age, often stopping by puberty or in the early teens. Learning and development are expected to be unaffected.

Information reviewed by Dr. Kate Riney, Mater Hospital Brisbane.

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