Autoimmune encephalitis

This information sheet from Great Ormond Street Hospital (GOSH) explains the causes, symptoms and treatment of autoimmune encephalitis and where to get help.

What is autoimmune encephalitis?

Autoimmune encephalitis is a group of rare neurological condition causing inflammation of the brain. It can follow on from a minor infection such as a cold, and is the result of the immune system becoming mis-programmed. There are several types of autoimmune encephalopathy – the most common of which is acute disseminated encephalomyelitis (ADEM) – see our separate information sheet for further information. The next most common in children is anti-N-methyl-D-aspartate (anti-NMDA) receptor encephalitis. Other types not covered in this information sheet include limbic encephalitis, Hashimoto encephalopathy and Rasmussen encephalitis.

What causes anti-N-methyl-D-aspartate (anti-NMDA) receptor encephalitis?

It is an autoimmune disorder although further research is needed to prove the actual mechanism of how the condition develops and what makes this more likely to happen in some people rather than others. Autoimmune disorders occur when the body mistakenly attacks itself rather than a foreign invader such as a bacterium or virus.

What are the signs and symptoms of anti-NMDAR receptor encephalitis?

The presentation of symptoms in children will be acute, meaning there will be an obvious and sudden change in the child's health. These changes will have usually happened over a short period of time. Sometime before the onset of symptoms, children may have a flu-like illness with a headache and high temperature. Once they have seemed to recover from this illness, other symptoms appear. These include altered mental state, behavioural changes and agitation. They can have seizures or fits, abnormal movements, hallucinations, sleep disturbance and decreased consciousness, which may require an admission to the children's intensive care unit. Speech changes may also develop, such as losing previously-fluent speech or lack of speech entirely.

How is anti-NMDAR autoimmune receptor encephalitis diagnosed?

Anti-NMDAR autoimmune receptor encephalitis can be difficult to diagnose and often needs specialist tests. Frequently, doctors consider infection as the cause of symptoms but to exclude these, bloods tests and a lumbar puncture may need to be carried out. Samples of blood and cerebrospinal fluid will often show markers of inflammation and specific antibodies. An MRI scan of the brain (and sometimes the spine) will be helpful in making the diagnosis and is usually normal. Electroencephalogram (EEG) tests will also be carried out to view any abnormalities in brain wave pattern.

How is anti-NMDAR autoimmune receptor encephalitis treated?

The aim of treatment is to dampen down the immune system to stop abnormal signalling so improving the symptoms and reducing lasting damage.

Steroid treatment can reduce some symptoms and stop new symptoms from developing. Once the diagnosis is made, they are usually given into a vein once a day for the next three to five days. Intravenous immunoglobulin (IVIG) is often prescribed alongside steroid treatment, also given into a vein. Plasma exchange may also be suggested when the condition is very severe. Rituximab is another medicine prescribed to reduce inflammation. Rituximab removes some of the white blood cells in the body called B cells. Removing these cells reduces the production of antibodies that may play a role in your child's illness.

What happens next?

Almost all children and young people make a good neurological recovery. This can take a number of months. The symptoms that tend to improve first are the problems with movement, reduced level of consciousness and seizures. Behavioural problems tend to continue or come back later. Children may have continued difficulties with memory and attention, but these can often be improved greatly with support from a variety of therapists. Support in school may be helpful for some children. Recovery may take up to two years.

Autoimmune encephalopathy can recur – around one-fifth of children have further episodes. These can be prevented to an extent by long term immunosuppressive medicines such as azathioprine and mycophenolate mofetil (MMF) and regular follow up monitoring.

Further information and support

The Encephalitis Society offer information and support to anyone affected by autoimmune encephalopathy. Call them on 01653 699599 or visit their website at www.encephalitis.info

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