

Acute Demyelinating Encephalomyelitis (ADEM) in children

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What is ADEM?

ADEM is a type of encephalitis caused by an inflammatory reaction in the brain and, sometimes, the spinal cord, which mainly affects the nerves in the 'white matter' of the brain. Often the child will have a history of an infection of some sort about 2-4 weeks before they become ill with ADEM. Such infections may be ordinary and easily forgotten such as a cold, sore throat or tummy upset. The immune system overreacts to this infection trigger and causes inflammation of the nerve coverings affecting how they work.

Recent research has shown that in some cases of ADEM, a molecule called Myelin Oligodendrocyte Glycoprotein antibody (MOG-Ab) can be found in the blood. MOG-Abs cause the white matter coating (myelin sheaths) to fall off nerves, giving rise to demyelination. This means messages cannot pass along nerves effectively, slowing them down or stopping them completely causing the brain's symptoms.

Symptoms of ADEM

ADEM usually starts suddenly. The symptoms can be severe and worrying. Headache, vomiting, drowsiness and neck stiffness are all common. Loss of balance and inability to walk or stand may also quickly appear. Seizures (convulsions or fits) can occur in up to one third of children, but they are not usually difficult to control.

How common is it?

ADEM is fairly rare. Typically, a large UK children's hospital will admit about 10-15 children per year with ADEM. This means that there will only be 1 or 2 cases per year in every local district general

hospital. Children that go to a local district general hospital may be moved to the regional children's hospital due to the initial severity of the illness and the need for rehabilitation in the recovery period. The peak age of onset is between 3 and 10 years, but younger and older children can also be affected. It is more common in children than in adults. Neurologists dealing with adults use different clinical definitions for ADEM than paediatric neurologists so it is difficult to make comparisons between children and adults who are given this diagnosis.

What happens in hospital?

Initially, treatment usually consists of antibiotics and antiviral drugs given through a drip while investigations proceed. This is because the symptoms of ADEM can look similar to those of meningitis (inflammation of the protective covering of the brain) or viral encephalitis (inflammation of the brain caused by a virus).

Most children are managed without intensive care, but if the site of the brain inflammation affects the breathing pattern, makes the child unresponsive or they have problems following seizures, then a period of controlled ventilation (help with their breathing using a machine) in intensive care may be required.

Investigations

- Blood tests including specialist blood tests to look for MOG-Abs.
- Lumbar puncture (LP) (sometimes called a spinal tap), if the child is well enough, is crucial for helping confirm the diagnosis. An LP may be postponed in a drowsy or unwell child because the management can go ahead anyway after a scan makes the diagnosis of ADEM likely. However, the LP still needs to be done at a later stage as it can provide very useful information.
- CT (computed tomography) brain scans are usually unhelpful in ADEM (the images can look normal), but they are often done in an emergency department as they are often the only type of scan available and can rule out other possible causes of the symptoms.
- An MRI (magnetic resonance imaging) scan of the brain and, sometimes, spinal cord is helpful in confirming the diagnosis. A child must keep still for an MRI scan so, if this isn't possible, they will require supervised sedation or a brief general anaesthetic. Each hospital has their own protocol for how they will arrange the scan. This is a very safe procedure. Often the lumbar puncture is arranged at the same time as the sedation or general anaesthetic.

Treatment

Treatments, which help to dampen down the inflammatory response (anti-inflammatory), can speed up the recovery. Most often a three to five day course of an intravenous (through a drip) steroid medicine called methyl prednisolone is sufficient to achieve this. Following this, a course of steroid (prednisolone) is usually given by mouth for a few weeks, initially at a high dose and then reducing slowly over a number of weeks to maintain the anti-inflammatory response.

There are some important side effects of these medicines. Please make sure you discuss these with the doctor who is treating your child. If your child is given a course of prednisolone, it is important to follow the instructions of how to withdraw and stop the medicine carefully. If the medicine is stopped too quickly, the child may become unwell. Make sure you tell all health care professionals you meet that your child is taking a course of steroids. Usually, children should not have vaccinations for three months after stopping steroids for ADEM. However, you should discuss the vaccination schedule with your child's doctor. There are other therapies that can be tried, but they are usually only given to children who do not respond to the steroid medicines. They include infusions of immunoglobulins (a blood product) given over a few days or a process of cleaning the blood called plasma exchange (PE).

Will my child get better?

Up to 75% of cases have a complete recovery. However the time scale for recovery varies in individual cases. The nervous system is often slow to repair itself, and it may take weeks, or sometimes months, to make a recovery. Children may be in hospital for a few weeks or months for rehabilitation.

In about half of cases, the MRI scan will show evidence of the episode, even when there has been a full recovery. For this reason, some paediatric neurologists do not arrange a repeat or follow up MRI scan. What matters is how well the child is able to recover functionally.

Can there be long term consequences?

Research suggests that children who do not make a full recovery may have learning, behavioural or physical difficulties. It is therefore very important for the child's care to include interdisciplinary rehabilitation both while in hospital and afterwards with their own community services. The team involved is likely to be made up of physical therapists (physio's), speech and language therapists,

psychologists and the education/ teaching service, depending on the individual's particular needs. Ongoing follow-up and links to the local services are important to maintain progress and continuity. Sometimes parents report behavioural changes which should be thought of in the same context as the learning support needs.

Will it happen again?

Many children who get ADEM do not get a second episode, however sometimes there is a recurrence which can present differently from the first episode, for instance with spinal cord involvement, visual disturbance or with a different set of symptoms related to the nervous system. Clinicians call such an illness Multiphasic Demyelinating Encephalomyelitis (MDEM). This usually happens within a few months of the initial ADEM illness. In recent research studies, mostly with smaller groups of patients, the risk of relapse is thought to be increased if MOG-Abs are found in the blood at the time of the first attack. Further research is actively ongoing into MOG-Abs and the role they play in predicting relapses and disease severity in ADEM patients.

Is MDEM the same as Multiple Sclerosis (MS)?

MDEM is not the same as MS. MS is very rare in children. However, a small number of recurring cases, especially where the features do not strictly fit the ADEM criteria, have been shown to be at a greater risk of developing MS in later life.

The majority of children make very good recovery over time, allowing the child and family to put the illness behind them in due course, and look forward confidently to the future.

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