Acute Demyelinating Encephalomyelitis [ADEM] in children
By Dr Rachel Kneen, Consultant Paediatric Neurologist, Alder Hey Children’s Hospital, Liverpool

What is ADEM?
This 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 illnesses are often quite ordinary and easily forgotten, like a cold, sore throat or tummy upset. The immune system has a kind of delayed over-reaction which starts to inflame nerve coverings affecting their normal function. The peak age of onset is 3 – 10 years, but children younger and older can also be affected.

What is the illness like?
It usually starts quite promptly. 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. When the doctor examines the child he/she finds broad ranging signs in the nervous system which often indicate that more than one area of the nervous system is involved. Seizures (convulsions or fits) can occur in up to one third of children, but the seizures are not usually difficult to control.

How common is it?
It is fairly rare; a large UK teaching hospital will admit about 10-15 children per year with the disorder. This means there will only be perhaps 1 or 2 cases in every local district hospital per year. Children that go to a local district hospital are likely to be moved to the large teaching hospital due to the initial severity of the illness and the need for rehabilitation in the recovery period. It is more much common in children than in adults. Adult neurologists do not use the same clinical definitions as paediatric neurologists do when considering ADEM as a diagnosis so it is difficult to make comparisons between children and adults who are given this diagnosis.
What happens in hospital?
Affected children are likely to be given antibiotics and antiviral treatment straight away through a drip while investigations proceed. This is because the symptoms may mimic those of Meningitis (inflammation of the protective covering of the brain) or Viral Encephalitis (inflammation of the brain substance caused by a virus). Blood tests will be done.

A lumbar puncture (LP, sometimes called a spinal tap), if the child is well enough, is crucial for helping confirm the diagnosis. The LP allows the doctor to sample the cerebrospinal fluid (CSF) that surrounds the brain and the spinal cord. A lumbar puncture 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 (see next paragraph). However the LP still needs to be done and will be performed a couple of days after admission to hospital as it can still provide very useful information at this stage.

Computed tomography (CT) brain scans are usually unhelpful (the images look normal), but they are often done in an emergency department as they are the only type of scan available. A Magnetic Resonance Imaging (MRI) scan of the brain and sometimes spinal cord will be arranged next. Their images are very helpful in confirming the diagnosis. A CT and a MRI are tests used to scan the brain in order to show the extent of inflammation. 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. Most children are managed without intensive care, but if the site of the brain inflammation affects the breathing pattern, or makes the child unresponsive or they have problems following seizures, then a period of controlled ventilation in intensive care may be required.

Can it be treated?
It can be treated. Medication which help to dampen down the inflammatory response can speed up the recovery. Most often a three to five day course of an intravenous corticosteroid medicine called methyl prednisolone is sufficient to achieve this. Following this, a course of corticosteroids are usually given by mouth for a few weeks, with a tailing off schedule, to sustain the process. This medicine is called prednisolone. 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 corticosteroids.

There are other therapies that can be tried, but they are only given to children who do not respond to the corticosteroid medicines. They include infusions of immunoglobulins given over a few days or a process of cleaning the blood called Plasma Exchange.

**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, so do not become disheartened if your child takes some weeks and sometimes longer to make that recovery. It can be faster for some children. Your child may be in hospital for a few weeks or months for rehabilitation.

In about half of cases the MRI scan will show persisting evidence of the episode, even when there has been a full recovery. This doesn’t matter that much - what matters is what the child is able to recover functionally. For this reason, some paediatric neurologists do not arrange a repeat or follow up MRI scan.

**Can there be long term consequences?**
Reports do suggest that in those children who do not make a full recovery their areas for support are likely to be in the field of learning or behaviour. They may also have some physical problems. This makes it even more important for the child's care to include interdisciplinary rehabilitation while in hospital and afterwards in his/her own community services. So follow-up and links are important locally as well as at the original hospital. The team involved is likely to be made up of physiotherapists (physio's), speech and language therapy, psychology and the education/ teaching service – depending on the individual’s particular needs.

Sometimes parents report behavioural changes which should be thought of in the same context as the learning support needs.
Will it happen again?
Most children who get ADEM do not get a second episode. Doctors and clinical research workers are tightening up on the definition of the diagnosis so that the children and their parents can be given the most up to date information about the disorder. However sometimes there is a recurrence, which can present differently, for instance with spinal cord involvement, or with a different set of neurological symptoms from the first episode. Clinicians now call such an illness MDEM (multiphasic demyelinating encephalomyelitis)]. This usually happens within a few months of the initial ADEM illness. Some hospitals in the UK are involved in a study looking at the risk of recurrence. Information about this study is available on the website www.childdemyelination.org.uk.

Is this the same as Multiple Sclerosis (MS)?
No. 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 recoveries over time, allowing the child and family to put the illness behind them in due course, and look forward confidently once more.