Multiple Sclerosis Society of Canada

Acute Disseminated Encephalomyelitis (ADEM)

What is ADEM?

Acute disseminated encephalomyelitis or ADEM, is a rare neurological disorder that predominantly affects children, but can affect individuals of any age. It is characterized by a short-lived but widespread attack of inflammation in the central nervous system (brain, including optic nerve, and spinal cord) causing damage to the myelin, the protective covering of nerve fibers.

What causes ADEM?

In about 50-70 per cent of cases, ADEM follows a viral or bacterial infection, often after an upper respiratory tract infection. Although many virus and bacteria strains have been implicated with ADEM, no one virus or bacteria has been identified as the cause. ADEM appears to be an immune reaction to the infection and typically begins 7 to 14 days after the infection. This reaction causes inflammation within the CNS resulting in damage to the myelin.

Although very rare, ADEM can also occur following a vaccination, generally after the measles, mumps and rubella (MMR) vaccine.

What are the symptoms of ADEM?

Symptoms often appear quickly and usually include: fever, headache, fatigue, nausea and vomiting and in the most severe cases, seizures and coma. Neurological symptoms of ADEM will depend on the area affected in the central nervous system and may include: visual loss (inflammation of the optic nerve), confusion, weakness in arms and legs, and difficulty with balance and coordination. Symptoms tend to get worse over several days.

How is ADEM diagnosed?

ADEM should be considered when multiple neurological symptoms, accompanied by fever, headache and altered mental state closely follow an infection or vaccination.

Magnetic Resonance Imaging (MRI) is an important part of diagnosing ADEM to help rule out infections in the central nervous system and other conditions with similar symptoms. Typically, widespread multiple lesions deep in the white matter of the brain are seen on MRI in cases of ADEM.

Lumbar puncture (spinal tap) may also be used to rule out other infections. In ADEM, the cerebrospinal fluid drawn will show an increase in white blood cells.

How is ADEM treated?

Typically, initial therapy for ADEM is **corticosteroids**. Intravenous steroids are usually given over a course of 5 to 7 days, followed by a tapered course of oral steroids. Steroids are given to reduce inflammation in the central nervous system and to speed recovery of most ADEM symptoms. Following steroid treatment, most people start to recover over several days and within six months of an ADEM attack, the majority of

people will have complete, or near complete recoveries. There are a number of individuals who may experience mild to moderate long-term impairment including cognitive difficulty, weakness, loss of vision or numbness.

Some people may not respond to steroid treatment and **intravenous immuoglobulin (IVIG)** may be used. Immunoglobulin is an antibody protein found in white blood cells and has been shown to reduce immune activity in several autoimmune conditions such as ADEM.

Plasmapheresis, also called **plasma exchange** is another treatment that can be used to reduce immune activity. Plasmapheresis is a process that removes circulating antibodies thought to cause the active immune response.

Chemotherapy drugs may be used in only very severe cases of ADEM, where other treatments have not been effective.

It is possible that ADEM can reoccur within a few months of the initial diagnosis. It is treated by restarting steroids. A very small number of people diagnosed with ADEM may later develop MS but there are currently no known risk factors to predict who those individuals may be. The long-term prognosis of ADEM for the vast majority of those affected is generally very good.

Sources: National Institute of Neurological Disorders and Stroke and the Cleveland Clinic. 2014

How to reach us

Call toll-free in Canada: **1.800.268.7582**
MS Society of Canada Web: **mssociety.ca**
Email: **info@mssociety.ca**

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