Fact Sheet

The Encephalitis Society
Support, Awareness & Research for Inflammation of the Brain

SSPE: a Chronic Encephalitis as a Result of Measles

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This factsheet aims to provide the people affected by Encephalitis, their families, friends and carers with a better understanding of how the encephalitis is diagnosed and what are the challenges in establishing this diagnosis.

What is SSPE?

SSPE stands for subacute sclerosing panencephalitis and refers specifically to a type of Encephalitis which can follow natural (wild) measles virus infection. After the initial measles infection, the virus lies passive in brain cells. It does not cause SSPE for several years (average 6 years) when eventually an inflammatory response is initiated against the infected cells. It is more common in children younger than 2 years who have had primary measles infection, although the condition (SSPE) manifests itself much later- older children and adults.

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Unfortunately SSPE is a progressive form of Encephalitis without a cure. Despite multiple attempts, no satisfactory treatment has been developed. In a few cases there has been remission following use of a certain drug or drug combination. However most of those affected die within about 5 years of diagnosis.

Explanation of terms:

'Subacute' means a quite slow start (typically around 9 months) and, usually, a gradually progression.
'Sclerosing' means that a reaction sets in which damages and scars the brain.
'Panencephalitis' means that all areas of the brain can be affected, though the outbreak varies from one individual to another.
'Chronic Encephalitis' means a type of Encephalitis that has a slow time course such as SSPE.

How common is it?

SSPE is a rare condition. It occurs in about 2 per 100,000 cases of natural measles. It is more common in developing countries because there is a higher rate of measles infection in such countries. It is rare in Western countries where there is an effective measles immunisation programme.

When does it happen?

There is a delay of several years (5-10 years) after an acute measles infection, before symptoms are seen. The initial symptoms can be very subtle. Children catching measles under 2 years of age are more vulnerable to the condition because of immune system immaturity. Boys are more likely to be affected than girls. Very rarely SSPE comes on more quickly and progresses more rapidly, particularly if measles is

caught by the infant around the time of birth. SSPE can also be rapid if it appears in a mother during her pregnancy.

What happens?

The brain is affected but as the brain controls the body, there are also physical symptoms. Two factors may be operating:
1. The measles virus remains in the brain in an altered (mutated) form,
2. The individual's immune response to the virus is abnormal and ineffective in getting rid of it.
This sets up a kind of inflammatory reaction, particularly around small blood vessels in the brain which can be seen under the microscope. Nerve cells [neurones] in such areas of the brain are damaged and lost progressively.

How is the child or adult affected?

There are several stages.
At first the problems are subtle and may be hard to spot as an illness. Usually it begins with a change in personality. It looks like the child affected is misbehaving. The ability to function at work or for a child to cope with school is altered. It may be a noticeable untidiness in hand writing, a difficulty in doing ordinary daily tasks, or a change in the power of expression in conversation. This can lead doctors or other specialists think that the child or young adult is suffering from psychological or psychiatric problems.
Often seizures (fits) or involuntary movements, such as jerks (often known as myoclonus) start and further investigations are then initiated. The start of these abnormal movements forms the next phase of the illness.

Then within about two months other movement problems start such as unwanted and uncontrolled movement of limbs which come and go, or

a gradual emergence of stiffness and increased (spastic) muscle tone and posture which can be one sided. As these signs emerge the intellectual deterioration also progresses. Seizures can be very troublesome and don't respond well to anticonvulsant medication. Vision, or recognizing what is seen, becomes affected by this stage.

Distressingly for family and carers, dementia and physical disability become severe and the person affected becomes totally dependent. Finally problems affecting feeding, swallowing and respiration contribute to the terminal phase. Death is usually caused by pneumonia.

How is it diagnosed?

Doctors should consider SSPE in a child or young adult who presents with a dementia-like illness, especially if they develop seizures or their vision is affected. Brain scans may be normal in the early phase, but eventually some non specific changes can be detected on MRI scans affecting certain parts of the brain (cortex, deep nuclei and white matter).

If the diagnosis is suspected, the level of specific measles antibodies (measles specific IgG) can be measured in the cerebrospinal fluid (CSF) and blood. A lumbar puncture is needed to obtain the spinal fluid. Parts of the measles virus (RNA) can also be detected in the CSF. The high levels of the measles IgG in the spinal fluid help to confirm the diagnosis.

The electroencephalogram (EEG) can also be suggestive at an early stage, with characteristic brief complexes of wave forms appearing periodically every few seconds. These are not seizures in themselves, but show a disturbance of the normal electrical pattern caused by the disease process.

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Is there any treatment?

Antiviral agents and Interferon alpha may have a modifying effect when given as a long-term treatment. Intravenous immunoglobulins and plasmapheresis which are treatments often used in other neurological immune disorders, have also been reported to have some benefit. Other medications which have been tried include amantidine, steroids and cimetidine. Large scale trials have not been undertaken due to the rarity of the disease. Sadly, despite initial improvements or a slowing in the natural progression, the disease eventually resumes its course and no cure has been reported up to this time.

Symptom control is also important. Sodium valproate or benzodiazepines have proven beneficial for many sufferers in its improvement of seizures. Other medication such as baclofen may be helpful for stiff muscles.

It is essential that co-ordinated medical and community care are instituted at the earliest opportunity once the condition is recognised. Much can be done to relieve discomfort and support nutrition and daily care. The child or young person and their family will need emotional and practical support. Referral to a psychologist, social worker, palliative care team and hospice is usually offered.

What about research?

There is interest in the problem of disturbed immunity and how it might be helped, though no clear line of treatment has yet emerged. Another approach considered is to inhibit or suppress the persisting measles virus by down-regulating the virus's RNA [Ribo Nucleic Acid] – its genetic messaging system that builds its own proteins. "Interfering

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RNAs" have been shown to be effective in research on the measles virus and SSPE measles virus.

Can SSPE be prevented?

This looks very promising through a successful measles vaccination programme with a good uptake in a population.

There has been a comprehensive review published in 2007.
The reviewers conclude that thorough vaccination programmes do protect the population from SSPE and indeed have the potential to eliminate SSPE by eliminating measles.

There is no evidence to suggest that the measles vaccine causes SSPE.

Unfortunately such an aim of elimination has not yet been achieved and the condition can and does still appear in adults and children. Continuing dedication to immunisation is essential.

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We try to ensure that the information is accurate and up-to-date as possible. None of the authors of the above document has declared any conflict of interest which may arise from being named as an author of this document.

The authors have used evidence, academic and professional experience in writing this factsheet. If you would like more information on the source material and references the author used to write this page please contact the Encephalitis Society.

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