

Subacute sclerosing pan-encephalitis (SSPE): a chronic encephalitis as a result of measles

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What is SSPE?

SSPE is a very rare type of encephalitis which can follow natural (wild) measles virus infection.

- '*Subacute*' means a slow start and, usually, a gradual progression.
- '*Sclerosing*' means a reaction which damages and scars the brain.
- '*Pan-encephalitis*' means that all areas of the brain are affected.
- '*Chronic Encephalitis*' means a type of encephalitis that has a slow time course.

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 a slowing down of the disease process or a remission following use of certain drug combinations, however, most of those affected die within about five years of diagnosis.

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 natural measles infection and less vaccination coverage. It was thought to be rare in Western countries where there was an effective measles immunisation programme. However, with the recent low intake of the vaccination against measles in some Western countries, we may see more cases of SSPE in the future.

The measles vaccination will protect against SSPE as long as the child has not been exposed to the measles virus before getting the vaccine. SSPE occurs more often in children or young adults who were younger than two years old when they had the initial measles infection. Boys are affected more often than girls.

Stages of the illness

The initial symptoms of SSPE can be very subtle and there are several stages of this illness. After the initial measles infection, the virus lies hidden in brain cells.

The symptoms of SSPE usually start around 6-8 years after the measles infection and are related to destruction of the infected brain cells. At first, the problems noted are subtle and may be hard to spot as an illness. Usually, it begins with a change in personality and 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 speech during conversation. This can initially lead doctors or other specialists to think that the child or young adult is suffering from psychological or psychiatric problems. Seizures (fits or convulsions) or involuntary movements, such as jerks of the limbs (often known as myoclonus) start and it is often at this point that further investigations are initiated. The start of these abnormal movements forms the next phase of the illness.

Within a few months other movement problems start such as unwanted and uncontrolled movement of limbs which come and go, a gradual emergence of stiffness, and increased (spastic) muscle tone which can be more noticeable down one side of the body. As these signs emerge, there is progression of the brain deterioration and it appears that

the child or adult is suffering from a type of dementia. Seizures can be very troublesome and don't respond well to treatment. Vision, or recognizing what is seen, becomes affected by this stage.

The dementia and disability become severe and the person affected becomes totally dependent on the family or carers for all their needs. Finally, problems affecting feeding, swallowing and breathing contribute to the final stage of the illness. The patient may stay in this period for many months or even years. Death is usually caused by pneumonia. 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.

Diagnosis

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 changes affecting certain parts of the brain (the cerebral cortex, deep nuclei and white matter) can be detected on magnetic resonance imaging (MRI) scans.

If the diagnosis is suspected, the level of specific measles antibodies (measles 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.

In the early stages of the illness an electroencephalogram (EEG) may be performed. This looks at brain wave activity. The findings on an EEG can be suggestive of SSPE with characteristic patterns of wave forms being noted. These are not seizures in themselves, but show a disturbance of the normal electrical activity in the brain caused by the disease process.

Is there any treatment?

Antiviral agents and a medication called Interferon Alpha may alter the effects of the illness when given as a long-term treatment. Immunoglobulins (a blood product that is given through the veins) and plasmapheresis (cleaning the blood of inflammatory chemicals) have also been reported to have some benefit. Other medications have not been demonstrated to have any significant effect on the disease course and unfortunately, large scale trials cannot be undertaken due to the rarity of the disease. Despite initial improvements or a slowing in progression, the disease eventually resumes its course and no cure has been reported up to this time. Sometimes patients can die as a result of complications related to the treatments themselves.

Symptom control is very important and can improve the patient's quality of life. Anticonvulsant (medication to stop fits) drugs can be helpful for controlling seizures. Other medications can be used to relax stiff muscles. Patients will ultimately require feeding via a nasogastric tube (tube passed from the nose into the stomach) or a gastrostomy (tube directly into the stomach from the tummy wall inserted by a small operation).

The main part of management is a co-ordinated approach to care, by the health professionals, once the condition is recognised. With a multidisciplinary approach much can be done to relieve discomfort, support nutrition and daily care. The child, or young person, and their family will need emotional and practical support. It is recommended that early referral to a psychologist, social worker, palliative care team and hospice is offered.

What about research into SSPE?

There is interest in the problem of disturbed immunity and how it might be reversed but no clear treatment has yet emerged. Another approach considered is to inhibit or suppress the persisting measles virus by stopping the virus' genetic material (RNA) building proteins needed for the virus to survive.

Can SSPE be prevented?

SSPE can be prevented by good uptake of the measles vaccination. This has been a problem in recent years as the uptake has dropped and outbreaks of measles have been reported in many areas of Europe and the USA. Ultimately, some of these children may get SSPE. There is no evidence to suggest that the measles vaccine itself causes SSPE. Continuing dedication to immunisation programmes is essential to try and eliminate this devastating disease.

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Thank you!

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