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 1 per 50,000 cases of natural measles. It is more common in developing countries because there are 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 provided 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 handwriting, 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 magnetic resonance imaging (MRI) scans may detect changes affecting certain parts of the brain (the cerebral cortex, deep nuclei and white matter).

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?

There is no consensus for treatment approach for SSPE, although among the anti-viral and immunomodulatory treatments used either individually or in combination, the highest rate of stabilization or improvement was observed with intraventricular interferon-α (anti-viral) and oral inosiplex (mixed antiviral and immunomodulatory properties)

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 because of complications related to the treatments themselves.

The most effective way to manage SSPE is symptom control. 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.

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