



## **Subacute-sclerosing panencephalitis (SSPE)**

### **A Chronic Encephalitis as a result of Measles**

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#### **What is this?**

SSPE stands for subacute sclerosing panencephalitis and refers specifically to an encephalitis which can rarely follow measles virus infection. It can affect children and adults when the measles virus persists in the brain.

'Subacute' means it has a slow onset and, usually, a gradually emerging 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 onset varies from one individual to another.

Unfortunately this is a progressive form of encephalitis without a cure. Though some treatments have given temporary improvement to some individuals, the disease process is ultimately overwhelming, being fatal within one year of diagnosis in the majority of cases.

A 'Chronic Encephalitis' is one that has a slow time course such as this.

#### **How common is it?**

It is not common. It occurs in about 4 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 after acute measles infection, before symptoms are seen. Children catching measles under 2 years of age are more vulnerable to the condition. 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, the symptoms seen are physical as well. Two factors may be operating: The measles virus remains in the brain in a slightly altered form. Also 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 hard to spot as an illness. Usually it begins with a slight change in personality and ability to function at work, or, for a child to cope with school. This may be a noticeable untidiness in hand writing for instance, or difficulty in doing ordinary daily tasks, or a change in the power of expression in conversation. Occasionally brief jerks of the limbs or loss of muscle control signal fits at this stage.

It is possible in the earlier phase for the brain to become swollen giving signs and symptoms of raised pressure.

Then within about two months other movement problems emerge. These may be unwanted uncontrolled movement of limbs which come and go, or a gradual emergence of stiffness and spastic muscle tone and posture which can be one sided. As these signs emerge the intellectual deterioration also progresses. Fits can be very troublesome. Vision, or recognizing what is seen, becomes affected by this stage.

Distressingly for family and carers, dementia and physical disability become severe and the child, or adult, who is thus affected becomes totally dependent. Finally problems affecting

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feeding, swallowing and respiration contribute to the terminal phase.

### How is it diagnosed?

Brain scans may be normal in the early phase, but eventually the sclerosing nature of the disease shows up as signal change on the magnetic scanner. Before that the diagnosis is usually made from examining lumbar puncture fluid and measuring the level of specific measles antibody there and in the blood. This is very high.

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

### Is there any treatment?

Antiviral agents, such as isoprinosine by mouth; ribavirine intravenously or injected into the cerebro-spinal fluid [CSF]; Interferon alpha into the CSF, or beta injected under the skin may have a modifying effect when given as a long-term treatment. Intravenous immunoglobulins, which are in common usage for immune disorders, have also been reported to have some benefit. However the disease eventually resumes its course and no cure has been reported up to this time.

Carbamazepine has proven beneficial for many sufferers in its improvement of seizures.

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.

### 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 RNAs" have been shown to be effective in research on the measles virus and SSPE measles virus.\* [Otaki et al 2006]

### 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\* [Campbell et al].

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, which is a disease of the wild virus.

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.

### References

Otaki M, Sada K et al. Inhibition of measles virus and SSPE virus by RNA interference. *Antiviral Res* 2006;70:105-11;

Campbell H, Andrews N et al. Review of the effect of measles vaccination on the epidemiology of SSPE. *Int J Epidemiol*. 2007 Dec;36(6):1334-48

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