# *Subacute Sclerosing Panencephalitis*

## Background

Subacute sclerosing panencephalitis (SSPE) is a rare, late complication of measles and is invariably fatal. Its incidence in Australia is not known. Despite the availablility of vaccination, a large nationwide epidemic of measles occurred between mid-1992 and mid-1995, with over 4,500 cases of measles being notified in 1993. Because there is a considerable delay between measles infection and onset of SSPE , some children affected in this epidemic may still not present for several years.

Measles vaccination first became available in Australia in 1968. A two-dose measles-mumps-rubella (MMR) vaccination strategy for children aged twelve months and in early adolescence was widely implemented by 1994. In 1998, all primary school children received a second dose of MMR vaccine. From 1998, both MMR doses will be given prior to primary school entry.

## Objectives

- to estimate the incidence of SSPE in Australia
- to determine the proportion of children with SSPE who have a prior history of measles and to establish at what age measles occurred
- to determine the proportion of children with SSPE who have a prior history of measles immunisation and to establish at what age immunisation was given
- to determine whether there is an over-representation of any ethnic group or children born outside Australia in children with SSPE

## Case definition

*Any child less than 16 years of age with a typical history of SSPE; ie. insidious onset of mental deterioration followed by development of progressive motor dysfunction, dementia and decerebration and ultimately death*

***and either***

*a) Raised measles antibody titres in the CSF* ***or***

*b) EEG changes showing "periodic complexes" typical of SSPE* ***or***

*c) Typical histopathological features of the brain at autopsy* ***or***

*Any combination of a), b) or c).*

## Results and discussion

In 1998 two notifications were received, one of which was a duplicate report. The child notified was a 14 year old Melanesian boy from Vanuatu who was not an Australian citizen. He had a history of possible measles infection (diagnosed clinically) at uncertain age and his vaccination status was unknown.

Between 1995 and 1998 inclusive, there were eleven notifications of SSPE. All questionnaires were returned and indicated that four notifications were duplicates and one was an error. For one case the questionnaire provided mimimal further information. Five children with SSPE had known onset of symptoms before 16 years of age (Table 12). None had a definite history of measles vaccination.

***Table 12 Subacute sclerosing panencephalitis, 1995-1997***

| Cases | Year of Diagnosis | Vaccinated | Measles Infection | Onset of symptoms | Age at diagnosis | Place of Birth | Australian citizen |
|---|---|---|---|---|---|---|---|
| Case 1 | 1995 | Possible | 10m | 14y | 17y 6m | Australia | Yes |
| Case 2 | 1995 | Don't know | Don't know | 15y 6m | 16y 6m | Australia | Yes |
| Case 3 | 1995 | Possible | Possible | 13y 2m | 13y 4m | Australia | Yes |
| Case 4 | 1996 | Possible | 9m | 15y 6m | 15y 11m | Philippines | Yes |
| Case 5 | 1997 | N/A* | N/A* | N/A* | N/A* | Malaysia | No |
| Case 6 | 1998 | Don't know | Possible | 14y 6m | 14y 7m | Vanuatu | No |

*Not Applicable

Two children had a definite history of measles infection; both in infancy. In these children the delay between measles infection and onset of symptoms of SSPE was 13 years and two months in one child and 14 years and nine months in the other.

The delay between onset of symptoms and diagnosis ranged from one month to three and a half years.
All children had consistent EEG changes and elevated CSF measles antibody titres. One child also had features of 'subacute encephalitis' on brain biopsy.

On the basis of these four cases in Australian children, this study suggests an incidence of 0.02/100,000 per annum in children diagnosed under 16 years of age for the period 1995-8.

## Conclusion

Subacute sclerosing panencephalitis is extremely rare in the paediatric age group and is likely to become less common with the introduction of the two dose MMR by primary school entry. Due to the long delay between measles infection or immunisation and onset of symptoms of SSPE, it is likely that additional cases are being diagnosed beyond 16 years of age by adult physicians.

## Investigators

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