

# MEDICAL PRACTICE

## Clinical Topics

### Register of cases of subacute sclerosing panencephalitis

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From July 1975 to October 1976, 30 patients were reported for inclusion in the register of cases of subacute sclerosing panencephalitis (SSPE). Four, however, did not fulfil the criteria for diagnosis—namely, characteristic clinical picture, electroencephalographic (EEG) appearances, measles antibody titres in serum and cerebrospinal fluid (CSF), and histological findings—and were therefore excluded. Of the remaining 26 patients, 18 were regarded as definite cases of SSPE and eight as probable cases.

#### Present series

##### DIAGNOSTIC CRITERIA

Many of the 26 patients did not satisfy all of the criteria for the diagnosis of SSPE (table I). Nevertheless, all were considered beyond any reasonable doubt to have the disease, cases being recorded as definite only when there was no doubt whatsoever. All were being managed by their clinicians as cases of SSPE.

Of the definite cases, all but one (case 18; table I) had measles antibodies in the serum and CSF, and all but two (cases 10 and 18) a typical clinical picture. In case 10 the serological findings were gross and the progress of the disease was typical of SSPE. There was little written information on case 18, and we accepted the opinion of the neurologist concerned that the patient was a definite case of SSPE.

Among the probable cases the presence of measles antibodies in the serum and CSF was the main criterion used to indicate SSPE; in

TABLE I—Presence of criteria positive for diagnosis of SSPE

Case No	Sex	Characteristic clinical picture	Measles antibody titres in serum and CSF	EEG appearances	Histological findings*	Previous measles infection
<i>Definite SSPE</i>						
1	M	+	+	+		+
2	M	+	+	+		+
3	M	+	+	+		+
4	M	+	+	+		+
5	F	+	+	+		+
6	F	+	+	+	+	+
7	M	+	+	+		+
8	F	+	+	+		+
9	M	+	+	+		+
10	M	+	+	+		+
11	M	+	+	+		+
12	M	+	+	+		+
13	M	+	+	+		+
14	M	+	+	+		+
15	M	+	+	+		+
16	M	+	+	+		+
17	M	+	+	+		Vaccine
18	F	+	+	+		+
<i>Probable SSPE</i>						
19	M		+			+
20	M		+			+
21	M	+	+	+		+
22	M		+			+
23	F		+			+
24	M		+			+
25	F	+				+
26	F		+			+

\*Histological studies were done in only three cases—one on necropsy material and two on brain biopsy specimens.

case 21 the diagnosis was put in doubt only by negative brain biopsy findings.

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#### GEOGRAPHICAL DISTRIBUTION

Twenty-two of the patients lived in Britain, one of whom was of Indian origin and had her measles infection in Tanzania. The remaining four—two from the Middle East, one from Italy, and one from

Greece—were referred to Britain for investigation. In the USA SSPE appears to be more common in rural areas and small towns.<sup>1</sup> There is no evidence of this in Britain.

SEX AND AGE AT ONSET

Nineteen of the patients were male and seven female. This ratio (2.7:1) is consistent with previous findings.

The age distribution was similar to that in previous reports (table II).<sup>2,3</sup> The average age at onset of the disease (10.7 years), however, was higher than found previously (8.5 years) owing to the inclusion of two patients whose disease was diagnosed at 17 and 27 years respectively. Although SSPE usually occurs much earlier than this, cases have been reported in patients aged up to 32 years.<sup>4</sup>

TABLE II—Age at onset of SSPE

No of cases	Age in years					Total
	3-5	6-8	9-11	≥12	Unknown	
1	5	13	5	2	26	

AGE AT TIME OF MEASLES INFECTION

A history of previous measles infection was elicited in 13 of the definite cases and six of the probable cases (table I). We assumed that the remaining patients had had subclinical infections; antibody titres showed that they had been in contact with the virus. The age at which the infection occurred (table III) was similar to the earlier data and shows that children who go on to develop SSPE suffer measles infection at an unusually early age. Seven patients had measles before their second birthday. Of the seven patients with no history of measles, one had received measles vaccination.

TABLE III—Age at time of measles infection

No of cases	Age in years						Total
	<1	1-	2-	3-	4-	≥5	
2	5	4	2	2	4	19	

DELAY BETWEEN MEASLES AND SSPE

The average gap between the measles infection and the onset of SSPE (6.6 years) agrees with previous findings in Britain and the

USA (table IV). The patient who received measles vaccine developed SSPE 4.2 years later. The American data show that SSPE develops sooner after measles vaccination (average 3.2 years) than after the natural infection (average 7.0 years).<sup>4</sup>

ONE HUNDRED YEARS AGO Gunshot-wounds and diseases of the hip-joint, for which the operation of excision has been performed, present to the surgeon difficulties in the after-treatment which are seldom met with in other surgical operations. The patient is utterly helpless; he is very likely prostrated with disease or by the wound, or the operation itself may have produced great exhaustion; he may be bound by splints or appliances considered necessary for promoting extension or counterextension; and the fact of his lying in this helpless condition is apt to bring about a series of complications, owing to the formation of bedsores, which in many instances are stated to have been the cause of patients succumbing; besides these, the surgeon has to contend with the copious discharge from the seat of wound, or that made by the operation, saturating the dressings, which, if not promptly removed, will become offensive, and may give rise to the formation of extensive abscesses or other complications. There is also the necessity for rearranging the sheeting and bedding, and for removing excreta by suitable utensils; and, at the same time, it is absolutely necessary to maintain the limb at perfect rest.

In investigating the experience of others with regard to how these arrangements have been carried out, we find that but little has been recorded; at the same time, nearly all recognise the importance of strict attention to the after-treatment, but lay greater stress on the

TABLE IV—Delay between measles infection and onset of SSPE

No of cases	Delay in years							Total
	<2	2-	4-	6-	8-	10-12	Unknown	
1	1	6	4	5	2	7	26	

OUTCOME

Seven patients were known to have died. The date of death of one of them, who died at home, is unknown. The other six survived 42, 6, 30, 13, 5, and 1 month respectively, the average survival time being 16 months. The longest surviving patient was a 30-year-old man whose symptoms of SSPE first appeared in 1962.

Conclusion

A scheme for the international collection of data has been proposed by the Virus Diseases Unit of the World Health Organisation. GD will be the co-ordinator for the UK, and the virology laboratory at the Institute of Child Health has been designated as the centre for serological analysis.

The incidence of SSPE appears to be falling along with the decline in measles notifications. The proportion of cases with a history of measles vaccination may be expected to rise, though work in the USA suggests that the risk of developing SSPE after vaccination is less than after natural measles infection. Of the 30 patients notified to the register since the last report in July 1975, only five had their onset in the current period. It is most important that all cases should be reported as they occur so that an accurate surveillance of the disease may be maintained.

References

- <sup>1</sup> Brody, J A, and Detels, R, *Lancet*, 1970, 2, 500.
- <sup>2</sup> Dick, G, *British Medical Journal*, 1973, 3, 359.
- <sup>3</sup> Dick, G, *British Medical Journal*, 1975, 3, 238.
- <sup>4</sup> Modlin, J F, Jabbour, J T, and Witte, J J. To be published.

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formation and application of splints for keeping the limb in a proper position. Professor Volkmann of Halle observes that, if a mattress or something could be invented by which the cleansing of the wound in these cases, the defaecation of the patient, and, at the same time, his utter immobility, could be maintained, it would be a great boon. The same authority remarks: "With regard to the great mortality after the operation of excision of the hip-joint, much has depended upon the want of care in the after-treatment. Three times I resorted to excision; twice after the battles of Beaumont and Sedan, once in Dijon; all these patients died; two, as I believe, principally in consequence of bedsores." And, as a reason for the fatal issue of most of the cases which had submitted to excision of the hip-joint during the late war, he notices the impossibility of bestowing upon the sufferers that care which is necessary after such an operation. "As long," he says, "as we are not able to put these patients on good beds, in order to place the limb in proper position, we dare not expect success." "No excision," he continues, "in fact, scarcely an operation exists which requires such care and unwearied patience in the after-treatment as excision of the hip-joint. The excised joint must be kept at rest, and the patient must be so laid that the wound at the hip is not pressed and the surgeon can have free access to it; that the dressing and cleansing of the same may be accomplished without moving the patient." (*British Medical Journal*, 1877.)