

Fig. EMG recordings of arm-mouth reflex. Surface electrodes were placed on the chin and the right forearm (FA). Faster flexions (A and B) of the FA elicited reflex opening of the mouth (indicated by solid lines), whereas very slow flexion (C) of the FA did not result the response.

trated electrophysiologically that corneomandibular and palmontal reflexes are stimulus-strength-dependent.^{1,2} To our knowledge, the arm-mouth reflex has never been described previously, the most similar reflex being the hand-mouth reflex of Babkin.^{4,5} The hand-mouth reflex consists of opening of the mouth, flexion of the neck, and sometimes closing of the eyes and flexion of the forearm produced by pressure on the palm of the hand.^{4,5} In the present case, pressure on bilateral palms resulted in slight opening of the mouth but no other associated movements. The difference in the mode of stimulation and response between the two reflexes indicated that they are clinically distinguishable, although the physiological mechanisms of the two reflexes seem to be shared. The findings of prolonged opening of the mouth produced by sustained flexion of the forearm suggests that the afferent impulse of the arm-mouth reflex might originate from one of the slowly adapting mechanoreceptors. It has been reported that primitive reflexes have little clinical value because of their presence in a significant proportion of the healthy population.^{6,7} To date, however, we have not found the arm-mouth reflex in any normal

adult. The appearance of the arm-mouth reflex in a patient with an advanced state of amyotrophic lateral sclerosis suggests that the reflex was related to the disease process and resulted from severe destruction of the upper motor neuron system innervating the perioral musculature. Accordingly, we consider that the arm-mouth reflex is a clinically valuable sign of severe destruction of the corticobulbar tract.

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Triphasic spike-wave stupor in portal-systemic encephalopathy: a case report.

Sir: Lennox¹ originally described prolonged stupor associated with bilaterally synchronous spike-wave discharges in the electroencephalogram (EEG). Though many authors described this condition as "petit mal status", Niedermeyer and Khalifeh² proposed the term "ictal stupor", or "spike-wave stupor". Most of the cases of spike-wave stupor have been observed in epilepsy. A few cases have been reported as spike-wave stupor in non-epileptic patients.³ We report a case observed in portal-systemic encephalopathy.

A 59-year-old woman was admitted to Kagawa Central Hospital on 13 April 1983, because of bizarre abnormal behaviour. That morning she had been disoriented and mentally dull. Prior to this episode, a similar episode which lasted 3 hours occurred in January 1983. During examination she was cooperative, but her attention span was short. She could give her name, the time and the place, her husband's name, but not calculate "100 - 7". There was amnesia about that morning's events. She appeared to be in a slightly stuporous state. Neurological examination revealed hyperreflexia of extremities and flapping tremor. On admission, EEG examination was performed immediately and showed continuous bilaterally synchronous spike-wave discharges (fig) which took the wave form of blunt spike-wave complexes. This was not altered by photic stimulation, nor by opening and closing the eyes. Her plasma ammonia level was 188 $\mu\text{g}/\text{dl}$ (normal; 30-86 $\mu\text{g}/\text{dl}$). During her admission, lactulose and kanamycin were orally administered. By the next day, her spike-wave stupor and flapping tremor had disappeared, although the ammonia level was still high (149 $\mu\text{g}/\text{dl}$). These high levels of ammonia remained high during admission. Her EEG recording in a non-stuporous state showed slow alpha activities mixed with sporadic theta waves.

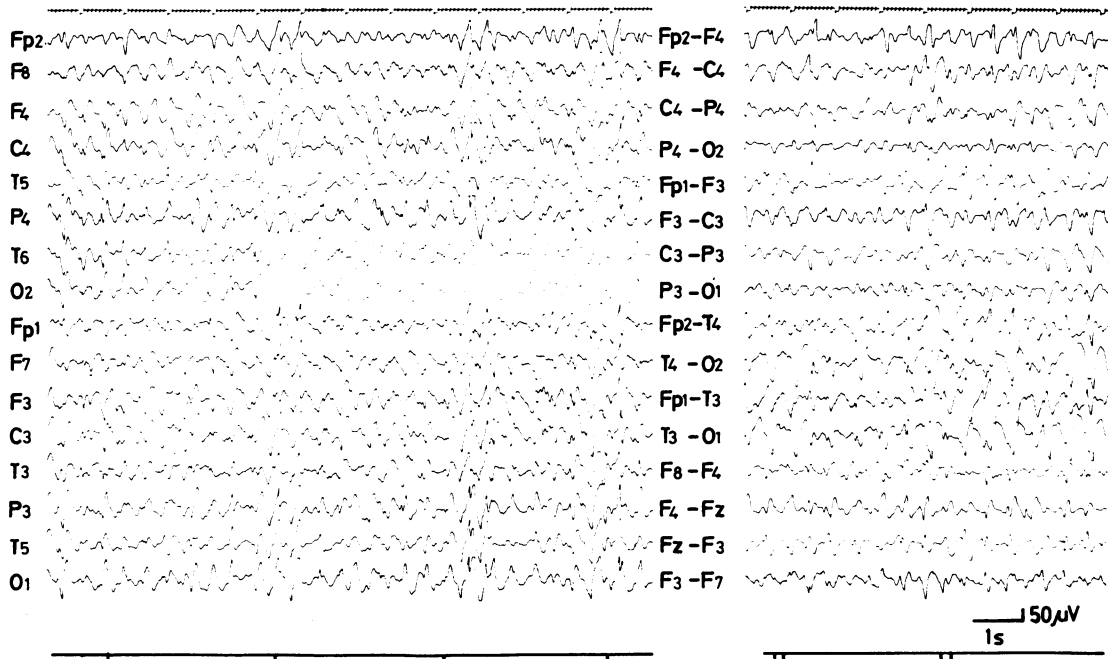


Fig. EEG in spike-wave stupor

Laboratory and radiological examinations revealed liver cirrhosis. Thus she was diagnosed as being in a spike-wave stupor due to portal-systemic encephalopathy. Six months after the episode, her EEG showed almost a normal pattern.

"Spike-wave stupor" have been used to describe an extraordinary state in epilepsy. However a similar state described as a combination of spike-wave continuity and prolonged stuporous symptom is, rarely, recognised in non-epileptic patients. Lion *et al*⁴ reported a case after trauma. Hosokawa *et al*⁵ described also the same state in two patients with cerebrovascular and Vogt-Koyanagi's disease. Hosokawa *et al*⁵ reported a case almost identical to ours, with portal-systemic encephalopathy. Poser⁶ described continuous bilaterally triphasic waves in hepatic coma. This EEG was similar to a spike-wave complex in ordinary epileptics. His patient had several grand mal convulsions during the course of illness. Bickford and Butt⁷ recognised that triphasic waves usually represent semicoma or complete unresponsiveness. In hepatic encephalopathy, the state of consciousness is not usually associated with the level of ammonia.⁸ Our case was clinically stuporous, but not comatose, with hyperam-

monaemia. Her initial EEG showed continuous bilaterally synchronous atypical spike-wave discharges of an epileptic nature. On the next day, her spike-wave stupor subsided, although she had partial amnesia about the previous day, and hyperammonaemia was still observed. Many metabolites seem to play important roles in hepatic coma.⁹ Other metabolic factors besides hyperammonaemia are considered to act together on the diencephalon, mesencephalon and ascending reticular formation of the brain stem,⁵⁻⁷ and might have produced a spike-wave stupor. It would help in the understanding of the mechanism of spike-wave stupor to investigate non-epileptic patients and it is therefore important to remember that a spike-wave stupor can be observed in non-epileptic disorders.

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