trated electrophysiologically that cor- 
neomandibular and palomental reflexes 
are stimulus-strength-dependent.

To our knowledge, the arm-mouth reflex has never been described previously, the most 
similar reflex being the hand-mouth reflex 
of Babkin. 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. 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. 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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References


Letters

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

Sir: Lennox originally described pro- 
longed stupor associated with bilaterally 
synchronous spike-wave discharges in the 
electroencephalogram (EEG). Although 
many authors described this condition as 
"petit mal status", Niedermeyer and 
Khalifeh 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-
elleptic patients. We report a case 
observed in portal-systemic encephalo-
pathy.

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 dis-
oriented 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 hus-
band's name, but not calculate "10 - 7". 
There was amnesia about that morning's 
events. She appeared to be in a slightly 
sporadic condition. Neurological examination revealed hyperreflexia of extremities and 
flapping tremor. On admission, EEG 
examination was performed immediately 
and showed continuous bilaterally syn-
chronous 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 µg/dl (normal; 30-86 µg/dl). During 
her admission, lactulose and kanamycin 
were orally administered. By the next day, 
she was stuporous and not responding to 
stimulation. The ammonia level was 
still high (149 µg/dl). These high 
levels of ammonia remained high during 
admission. Her EEG recording in a non-
soporadic state showed slow alpha 
activity mixed with sporadic theta waves.
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* described continuous bilaterally synchronous 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 But* 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.* Our case was clinically stuporous, but not comatose, with hyperammonaemia. 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. Other metabolic factors besides hyperammonaemia are considered to act together on the diencephalon, mesencephalon and ascending reticular formation of the brain stem, 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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Fig. EEG in spike-wave stupor

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