ground of the elaborate and atheotid phenomenology alone.

WIM VERHAGEN
MWIM HORSTINK
Dept Neurology
SLH NOTERMANS
Dept Clinical Neurophysiology,
St Radboud Hospital,
University of Nijmegen,
The Netherlands

References
1 Spillane JD, Nathan PW, Kelly RE, Marsden CD. Painful legs and moving toes. Brain 1971;94:541–56.
19 Eccles RM, Lundberg A. Synaptic actions in motorneurons by afferents which may evoke the flexion reflex. Arch Ital Biol 1959;97:199-221.

Accepted 1 September 1984

The arm-mouth reflex in a patient with amyotrophic lateral sclerosis

Sir: Numerous pathological or primitive reflexes may appear in a variety of diseases of the central nervous system including amyotrophic lateral sclerosis. We have observed reflex opening of the mouth upon passive flexion of the forearm in a patient with an advanced state of amyotrophic lateral sclerosis. To our knowledge, this reflex, which we have called arm-mouth reflex, has never been previously described.

A 57-year-old Japanese man noticed wasting and weakness of both hands in April 1981. During the next nine months progressive weakness of lower limbs and difficulty in swallowing and speaking developed. The patient's history was otherwise normal. He was admitted to hospital on 11 January 1982. Findings on general physical examination were normal as was his mental status. Bilateral weakness was present mildly in facial muscles and moderately in bulbar muscles with dysphagia and dysphonia. Atrophy and fasciculations in the tongue were observed. Weakness was also present mildly in neck flexors, moderately in all limb girdle and proximal limb muscles and severely in bilateral hand muscles. Deep tendon reflexes were symmetrically hyperactive in association with Babinski's reflex. Jaw jerk was exaggerated and superficial abdominal reflexes were absent. Sensation and coordination were normal. Cerebrospinal fluid examination and radiographs of the skull and spine were normal. Electromyographic examination of the right biceps brachii, first dorsal interossei, and rectus femoris showed fibrillation potentials, positive sharp waves, an increase in potential amplitudes and polyphasic potentials with a decrease in the maximum interference pattern. Pulmonary function tests showed that his vital capacity was 49%. A diagnosis of amyotrophic lateral sclerosis was made. The patient became almost aphagic and apnoeic by June 1982 and required the use of a feeding tube and a respirator. Muscle weakness also progressed rapidly. Neurological examination in April 1983 showed that the patient was alert and oriented but was totally aphagic, aphoniac, dysphagic and quadriplegic in extension. Supranuclear ophthalmoplegia and facial diplegia were noted. Forced crying was frequently seen. Deep tendon reflexes were symmetrically depressed but the sustained jaw clonus and head-retraction reflex were elicited. Tactile stimulation of the tongue resulted in reflex masticatory movement. At this clinical state, the arm-mouth reflex was observed, and consisted of a wide opening of the mouth upon passive flexion of the forearm and could only be elicited by flexion of either forearm. The response movements were restricted to the perioral musculature and was constantly elicited by repetitive stimulation. The appearance, latency and duration of the response were absolutely dependent upon the strength of the stimulus (fig): faster and greater flexion of the forearm resulted in more prolonged and wider opening of the mouth at a shorter latency, whereas slower flexion of the forearm elicited the response less often. Flexions and extensions of other joints and sensory stimuli including pain and tendon tap of the triceps brachii did not elicit opening of the mouth.

The reflex opening of the mouth by the passive flexion of the forearm described here can be classified as a primitive brainstem reflex for the following reasons. Firstly, passive flexion of the forearm was the only stimulus that resulted in a stereotyped response of the perioral musculature at distinct latent periods following the stimulus. Secondly, the appearance, latency and duration of the response were dependent upon the strength of the stimulus. These physiological properties are in accordance with those of primitive brainstem reflexes.1-3 It has been demons-
trated electrophysiologically that cor- 
neomandibular and palmomental 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 con-
ists 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 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 indi-
cated that they are clinically distinguish-
able, 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 popu-
lation.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 clini-
cally valuable sign of severe destruction of 
the corticobulbar tract.

Y KURODA
K ODA
H SHIBASAKI
Division of Neurology, 
Department of Internal Medicine, 
Saga Medical School, 
Nabeshima, Saga 840-01, 
Japan

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.

References

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

Sir: Lennox1 originally described prolonged stupor associated with bilateral 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 behavior. That morning she had been disoriented and mentally dull. Prior to this episode, a similar episode which lasted few hours occurred in January 1983. During the 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 µg/dl (normal; 30–86 µg/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 µg/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.