J.Neurol. 223. 191-198 (1980)

Journal of
Neurology

Involuntary Motor Phenomena in the Locked-In Syndrome

G. Bauer*, F. Gerstenbrand, and W. Hengl

Universitätsklinik für Neurologie, Anichstraße 35, A-6020 Innsbruck, Austria

Summary. Several involuntary movements were observed in patients who were totally immobile except for vertical gaze due to bilateral pyramidal transverse lesion at the pontine or midbrain level. In those conscious locked-in patients extensor spasms and flexor spasms could be elicited by nonspecific stimulation. Mimic pain reactions, pathological crying, and primordial screaming ('cat crying') were also noted. Other motor patterns were whining, moaning, groaning, sighing, and yawning. The pathophysiological implications of these observations are discussed,

Key words: Locked-in syndrome - Motor automatism - Extensor and flexor spasms.

Zusammenfassung. Mehrere unwillkürliche motorische Phänomene wurden bei vollständig gelähmten Patienten mit bilateralen Pyramidenbahnläsionen in Pons- oder Mittelhirnebene beobachtet. Durch unspezifische Stimulation wurden bei diesen bewußtseinsklaren Locked-in-Patienten Streck- und ausnahmsweise auch Beugesynergismen ausgelöst. Komplexere Verhaltensmuster waren mimische Schmerzreaktionen, Zwangsweinen und ein undifferenziertes, automatisches Schreien („cat crying"). Weitere Schablonen waren Winseln, Stöhnen, Grunzen. Seufzen und Gähnen. Die pathologischen Erklärungsmöglichkeiten dieser Phänomene werden diskutiert.

Introduction

The locked-in **syndrome** (LiS) denotes a neurological condition due to bilateral transsection of pyramidal tracts at the level of pons or cerebral peduncles leading to complete immobility except for vertical gaze. The syndrome shows several varieties in terms of the symptoms and course, the subject of a previous paper [2]. In locked-in patients a number of movements had been observed that were incompatible with the extent of voluntary paralysis. Since the LiS represents a

* To whom correspondence should be sent

0340-5354/80/0223/0191/S 01,60

unique natural transsection experiment with preserved consciousness, the investi gation of those motor phenomena promised some insight in the organization and localization of non-pyramidal motor systems. The present paper gives a description and list of these movements and outlines the possible explanations.

Case Reports

All cases had already been published by Bauer et al. [2]. The following reports condense the involuntary motions observed in these patients, and further details may be gleaned from the previous paper.

Case I. M.A., male, aged 68, was a classical LiS due to a ventral pontine infarction verified by autopsy. Except for vertical gaze the patient was virtually immobile. The sixth day after admission to the hospital the patient *muttered* some low sounds, followed by sighing and change in respiration rate. ¯Those manifestations seemed to be time-locked to verbal commands, but we learned from the patient via eye blink communication that he was neither able to make them voluntarily nor could he suppress them. Breathing was otherwise very regular and machinelike with intermingled periods of Cheyne-Stokes respiration which the patient could not change voluntarily.

Case 2. C.E., female, aged 44, was admitted with a classical LiS due to thrombotic occlusion of the basilar artery and hemorrhagic infarction of the ports. She opened her eyes on command, but no meaningful communication was possible. The EEG was normal with a reactive 9-10 cps rhythm. During this stage *decerebrate posturing* could be elicited by exogenous stimuli, especially by neck deflection. The patient's condition deteriorated rapidly to a bulbar brain syndrome. At this time the muscle tone was flaccid, and the extensor spasms had disappeared. The patient died three days after admission.

Case 3. W.R., female, aged 42, suffered from an incomplete LiS with remnants of lateral gaze and right arm motility, After three days, she was comatose with diffuse slowing in EEG due to secondary pulmonary infection and metabolic derangement. *Decerebrate posturing*¯ was observed during the locked-in condition and also in coma. At admission, the facial component of a *pain reaction* was noted, although the VII nerve muscles were otherwise completely paralyzed. Breathing was machinelike and became insufficient in coma, so artificial respiration was needed. At autopsy, thrombotic occlusion of the basilar artery and infarcted transsection at the mid-pontine level were found.

Case 4. R.D., male, aged 65, was admitted in an incomplete locked-in condition with inconspicuous movements of all extremities and voluntary downward gaze but no voluntary facial movements. Secondary to painful stimuli a marked *pain reaction* occurred, and *decorticate posturing* could also be triggered easily. Breathing was ataxic with prolonged apnoic periods. A hemorrhagic infarction of the pons was demonstrated at autopsy.

Case 5. B.H., a woman aged 66. was reported in detail under case number 7 by Bauer et al. [2]. Although considered totally locked-in, weak facial movements resembling a *pain reaction* could be induced by powerful noxious stimuli. Breathing was ataxic from the very beginning of the disease which turned out to be an extensive pontine hemorrhage.

Case 6. S.A., a woman aged 67, was described in detail under case number 8 in the previous paper. During the acute phase of the nearly classical LiS *groaning* was noted. It occurred spontaneously and also secondary to attempted verbal communication. Seven days after admission, *yawning* accompanied by typical generalized body stretching was noted. The observation was made while recording an EEG. The tracing was judged within normal limits, and cerebral electrical activity remained unchanged during the yawning. The patient's condition improved to an incomplete LiS. Ocular motility became free, some crude sounds could be uttered voluntarily and quadriplegia was also slightly improved, During this phase the patient exhibited compulsory *mimic reactions* by far exceeding the extent of motions already possible by

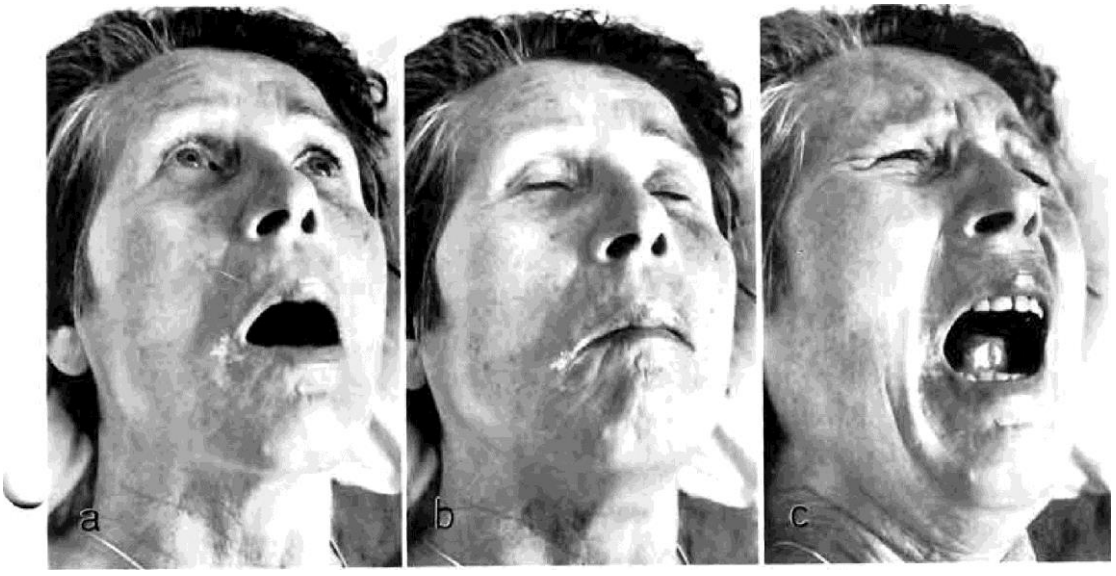

Fig. 1 a–c. *Case 6*, S.A., female, 67 years: a maximal voluntary attempt to open the mouth, b maximal voluntary attempt to close the eyes, and *c* compulsory mimic reaction. The extent of facial movements exceed the voluntary ones

Table 1. List of involuntary motor phenomena reported in the literature and in the present series

Motor phenomenon	Author	Present series
Flexor spasm (decorticate posturing)	Nordgren et al. [29]	Case 4
Extensor spasm (decerehratc posturing)	Dehaene and Martin [7] Feldman [9] Markand and Dyken [23] Nordgren et al. [29] Shafey et al. [39] Wilkus et al. [441	Case 2 and 3
Pain reaction	Markand [22J	Cases 3, 4, 5 and 7
Compulsory mimic reaction (pathologic crying)	Feldman [9] Markand and Dyken [23] Messert et al. 1251	Cases 6 and 8
`Cat crying'	Biemond [4]	Cases 6 and 7
Whining. moaning, groaning	Feldman [9] Poeck [33]	Cases 1 and 6
Yawning	Gschwend [15] Karp and Hurtig [19] Nordgren et al. [29]	Case 6
Chewing. slicking	Nordgren et al, [29]	—
Sighing	—	Case 1
Coughing	Feldman [9] Nordgren et al. [29] Wilkus et al. [44]	—

voluntary innervation (Fig. 1). Those compulsory mimic reactions were frequently, but not invariably, accompanied by a penetratingly loud *crying.* Urging the patient to stop was never successful, and it never became clear whether the crying was secondary to external or internal distress. The next month crying was frequently repeated each day, The repetition seemed to follow an endogenous pattern linked with the well-developed sleep cycles. There were hours of regular crying at a rate of once or twice a minute. In clinical jargon we coined the term *'cat crying'* for this rather troublesome symptom. When paralysis of cranial nerve muscles further improved, cat crying and eventually compulsory mimic reactions disappeared.

Case 7. B.M., a woman aged 27, was admitted after sudden loss of responsiveness due to an incomplete LiS with preserved vertical eye movements and convergence. Mimic *pain reactions* and cat-crying-like phonations could be released by noxious stimuli and with apparent emotional distress, i.e., seeing her relatives. Those involuntary movements were demonstrable in the acute and subacute phase but they disappeared with improvement of cranial nerve functions.

Case 8. P.R., a woman aged 61, suffered from an incomplete LiS due to a pontine vascular accident demonstrated by CAT. With improvement of the caudal cranial nerve functions, *compulsory mimic reactions* occurred. The involuntary motions were much more forceful than were the responses to command. The patient wore a tracheal tube so phonation phenomena could not be reliably judged.Compulsory mimic reactions were clearly secondary to emotional stimuli. The patient also signaled by eye blinks that the mimic pattern was accompanied by overwhelmingly negative feelings like being incurably crippled or separated from her family. An automatic repetition rate such as the one in case 6 was never observed.

Table I gives a list of the involuntary phenomena in the cases reported, completed by corresponding observations of other authors.

Discussion

Normal motor functions are the result of an interaction of sensory input, of processing by integrative cortical areas, and of programing by the primary motor cortex. This program is carried out with the support of the extrapyramidal system, of the cerebellum, or primordial motor patterns built-in at different levels of the brain stem, and of reflexes mediated by the spinal cord. Under pathological conditions abnormal involuntary motor phenomena appear or normal movements occur in an uncontrolled automatic fashion. The studies of those motor phenomena have brought remarkable insights in the organization of the motor system, occasionally marred by somewhat whimsical speculations about the biological goal of these patterns. However, motor phenomena might be divided into normal reactions, complex automatic behavior, and simple reflexes.

Normal reactions are variable and represent appropriate responses to the changing demands of the environment. Automatic motor patterns such as *motorische Schablonen* [201 or *motorische Primitivschablonen* [13] appear in a stereotyped fashion and are considered primordial behavior organized somewhere in the subcortical structures and released under pathological conditions by the lacking influence of cortical inhibiting systems [13, 20, 34]. Therefore, automatic motor patterns have been observed in coma [5, 14, 32], in chronic vegetative state or apallic syndrome [13, 17], in cases of bilateral supranuclear palsy [34], in cerebral malformations [6, 12, 26], and in normal inborns [30]. The examination of motor phenomena in high pyramidal transsections like the US can be of great value to disclose the localization of the so-called centers of such motor patterns.

In LiS, it seems to be easy to differentiate involuntary motor phenomena from remnants of pyramidal movements. The conscious patients use the later ones for communication. However, most of the pathological patterns are stimulus-sensitive, and it might be hard to decide in a particular case, whether the movements are secondary to command or to nonspecific stimulation. Repeated examination and the consideration of the unvariable type of the pathological motor pattern may lead to the proper diagnosis.

Extensor spasms (decerebrate posturing) were frequently *seen* with the LiS (Table 1) and were invariably stimulus-sensitive. Neck deflection was especially effective (case 2). Extensor spasms are identical with Jackson's cerebellar fits and are a constitutional sign of midbrain affection due to transtentorial herniation [5, 10, 14. 24. 32, 37, 43] or functional dissolution down to the midbrain level due to metabolic or anoxic encephalopathies 13, 36]. Gamper [12] showed both extensor and flexor spasms in the ancephalic midbrain. Since extensor spasms were also seen in localized brain stem lesions with intact cortical functions [8, 11, 16, 38, 42], the term decerebrate posturing is intrinsically fallacious [9]. This statement is strongly supported by the observations in LiS. Extensor spasms disappear with dissolution of bulbar brain functions [14. 32]. These clinical observations suggest that extensor spasms are stimulus-sensitive-composed reflexes built-in within the bulbar brain and released by interruption of inhibiting pyramidal influences somewhere above the midbrain level. These limits were already established by the experimental work of Sherrington [40,41]. The closer localization lies within the multisynaptic system of the brain stem intimately related to the ascending reticular activating system [18]. A connection of the stretch reflex with a diffuse activating system in the brain stem was already suspected as early as 1926 by Gamper [12].

Flexor spasm (decorticate posturing) was rarely observed in LiS (Table I). Case 4 was an incomplete LiS with motor remnants of all four extremities. From the observations in transtentorial herniations [14, 32] one has to draw the conclusion that flexor spasms are integrated above the midbrain-diencephalic border. This might be the reason why it is exceptionally observed in LiS and only in the incomplete form.

Pain reactions, compulsory *mimic reactions (Zwangsweincn)* and compulsory screaming *(cat crying)* have been separately listed in Table 1, although these patterns cannot be distinguished by the mere description of the motor phenomenon. The pain reaction represents a normal phenomenon secondary to specific stimulation, abolished with certain depressed cerebral states and exaggerated with others [18]. Compulsory mimic reactions *(Zwangsweinen)* occur with nonspecific stimulation and are frequently repeated [34]. The differentiation from an exaggerated pain reaction may be impossible and meaningless in a practical as well as in a theoretical sense. Cat crying occurs spontaneously in an automatic repetition rate with an extremely pronounced vocal component.

In LiS, pain reactions were seen in four cases of the presented series during the acute phase but were reported in literature only once [221. Its facial and vocal components are integrated in the periaqueductal gray [18]. Its occurrence in LiS suggests that the pathways to the corresponding cranial nerve nuclei do not use

the pyramidal tracts interrupted in LiS but instead a primordial motor system within the dorsal part of the brain stern.

There are several observations of compulsory mimic reactions *(Zwangsweinen)* in chronified LiS (Table 1). It occurs spontaneously and in a rhythmic pattern like other cases of severe supranuclear palsy [35]. Pathological crying is organized in the brain stem and usually appears in conjunction with bilateral pyramidal or extrapyramidal motor disorders [34]. Laughing was never observed in LiS. Therefore, it has to be integrated in the rostral brain stein above the lesions leading to a LiS. This is in line with the findings in malformations [12, 26].

Cat crying may be separated from the vocal component of the pain or compulsory mimic reactions. It is crude and shrill like the screaming of prematures. Neumann [27, 28] described spontaneous and automatic screaming in children with tumors in the rostral brain stem. Sherrington [40] and Catel and Krauspe [6] suppose a primordial system for vocalization in the oral brain stem whose irritation or disinhibition might also he responsible for cat crying in LiS.

Yawning was also seen in LiS (Table 1). Observations in the apallic syndrome [13], in cerebral malformations [6. 12], in inborns and prematures [30], and in the early phases of transtentorial herniations [32] make it likely that it is integrated in the lower brain stem, an area undisturbed by ventral pontine syndromes. Yawning also contains generalized body stretching resembling extensor spasms [12, 32], and the whole sequence of behavior is finished by swallowing [30]. Connecctions with the sleep-inducing system [15], the arousal system [12], or the respiration 'center' [30] were conjectured.

The REM sleep and to a lesser degree, the NREM sleep is altered in LiS [9, 21. 22. 23]. The changed sleep pattern is not due to immobility such as that in high cervical lesions [1] but to the direct involvement of the sleep-inducing system within the brain stem. REM sleep reduction was only seen with paralyzed horizontal gaze, whereas incomplete LiS with preserved lateral eye movements had comparatively undisturbed sleep cycles [23]. These findings prompted the assumption that the paramedian pontine reticular formation known to be responsible for conjugate gaze movements at the pontine level might also be related to induction or priming the REM sleep. However, the relation of the sleep system to yawning remains unclarified.

Breathing is certainly affected in LiS. The pattern may be very regular (cases 1 and 3) resembling the central neurogenic hyperventilation [31] or even ataxic (cases 4 and 5) [29]. Peiper [30] considered the nervous structures for yawning to be part of the respiratory center, suppressed under normal circumstances, but activated or released with disturbed breathing. This conjecture cannot be confirmed or rejected by the findings in LiS.

Further motor phenomena like whining, moaning, groaning, chewing. and coughing were incidentally observed in LiS (Table 1). With those phenomena. the differentiation to residues of voluntary motions is particularly difficult except for coughing which appeared clearly reflex-like to tracheal suction [29. 44]. Chewing or other oral automatisms were never seen in the cases reported and only once in the literature [29]. In contrast, oral motor patterns reappear first in apallic patients [13] and were fully developed in cerebral malformations [12. 26]. This

contrast indicates an important difference between primary or prolonged and acute decerebrate or cerebrobulbar disconnection syndromes.

References

1. Adev, W. R., Bors, E., Porter, R. W.: EEG sleep patterns after high cervical lesions in man, Arch. Neurol. **19**, 377-383 (1968)
2. Bauer. G., Gerstenbrand, F., Rumpl, E.: Varieties of the locked-in syndrome. J. Neurol. **221**. 77-91(1979)
3. Batter., G., Niedermeyer, E.: Acute convulsions. Clin. Electroencephal. 10. 127-144 (1979)
4. Biemond, A.: Thrombosis of the basilar artery and the vascularisation of the brain stem. Brain **74**, 300-317 (1951)
5. Buthenuth, J., Fuchs. E. C., Schilfter, R., Wolf, P.: Klinische Kriterien zur Bestimmung der Komaticre, akt. neural, 2. 81-102 (1975)
6. Catel, W., Krauspe, C. A.: Über die nervöse Leistung und den anatomischen Bau einer menschlichen Hirnmißbildung (Meroencephalie mit Merokranie), Jb. Kinderheilk. **129.** 1-54 (1930)
7. Dehaene, J., Martin, J. J.: "Locked-in" syndrome. .A clinicopathological study of two cases. Europ. Neurol. **14**, 81-89 (1976)
8. Dorndorf, W., Gänshirt, H.: Die Klinik der arteriellen Gefäßverschlüsse. In: Der Hirnkreislauf. H. Gänshirt (ed.), pp. 512-629**.** Stuttgart: Thieme 1972
9. Feldmann, M. H.: Physiological observations in a chronic case of "locked-in" syndrome. Neurology (Minneap.) 21. 459-478 (1971)
10. Fischgold, H., Mathis, P.: Obnubilations, comas et stupeurs. Etudes electroencephalographiques. EEG Clin. Neurophysiol. 1959, Suppl. 11, 124 p.
11. Fog, M., Hein-Sorensen. O.: Mesencephalic syndromes. In: Handbook of clinical neurology, P.J. Vinken, G. W. Bruyn (eds.). Vol. II. pp. 272-285. Amsterdam: North Holland 1969
12. Gamper, E.: Bau and. Leistungen eines menschlichen Mittelhirnwesens (Arhinencephalie mit Encephalocele), zugleich ein Beitrag zur Teratologie und Fasersystematik, II. Klinischer Teil. Z. ges. Neurol. Psychiatry 104. 49-120 (1926)
13. Gerstenbrand F.: Das traumatische apallische Syndrom. Wien, New York: Springer 1967
14. Gerstenbrand, F., Lücking, C. H.: Die akuten traumatischen Hirnstammschaden. Arch. Psychiatr. Nervenkr. **213**. 264-281 (1970)
15. Gschwend, J.: Gähnen als einziges Verhaltensmuster der Gesichts- und Kiefermuskulatur bei einem Patienten mit einem Ponsgliom. Fortschr. Neurol. Psychiatr. **45**. 652-655 (1977)
16. Halsey, J. H., Downie, A. W.: Decerebrate rigidity with preservation of consciousness. J. Neurol. Neurosurg, Psychiatry **29**. 350-354 (1966)
17. Ingvar, D. H., Brun, A.: Das komplette apallische Syndrom. Arch. Psychiatr. Nervenkr. **215**, 219-239 (1972)
18. Jouvet, M.: Coma and other disorders of consciousness, In: Handbook of clinical neurology, P.J. Vinken. G. W. Bruyn (eds.), Vol. III. pp. 62-79. Amsterdam, New York. Oxford: North Holland 1969
19. Karp, J.S., Hurtig, H.I.: "Locked-in" state with bilateral midbrain infarcts. Arch. Neurol. **30**. 176-178 (1974)
20. Kretschmer. E.: Der Begriff der motorischen Schablonen und ihre Rolle in normalen und pathologischen Lebensvorgängen. Arch. Psychiatr. Neurol. **100**. 1-3 (1953)
21. Markand, 0.N.: Sleep abnormalities in patients with brain stem lesions. Neurology (Minneap.) 25, 385 (1975)
22. Markand, 0.N.: Electroencephalogram in "Locked-in" syndrome. EEG Clin. Neurophysiol. **40.** 529-534 (1976)
23. Markand. 0.N., Dyken, M. L.: Sleep abnormalities in patients with brain stem lesions. Neurology (Minneap.) **26**. 769-776 (1979)
24. McNealy. D.E., Plum, F.: Brainstem dysfunction with supratentorial mass lesions. Arch. Neurol. **7**. 10-32 (1962)
25. Messert. B., Orrison, W.W., Hawkins, M. J., Quaglieri, Ch. E.: Central pontine myelinolysis. Neurology (Minneap.) **29**. 147-160 (1979)

26. Monnier, M., Willi, H.: Die integrative Tätigkeit des Nervensystems beim meso-rhombo spinalen Anencephalus (Mittelhirnwesen), Monatsschr, Psychiatr. Neurol, **126**, 239-258 (1953)
27. Neumann, J.: Über die spontane automatische Lautgebung. Wien. Z. Nervenheilkd. Deren Grenzgeb. 25. 20-30 (1967)
28. Neumann, J.: Über den gleichzeitigen Verlust der spontanen automatischen Lautgebung und der Sprache. Wien. Z. Nervenheikd. Deren Grenzgeb. **25**. 31-36 (1967)
29. Nordgren, R. E., Markesbery. W. R., Fukuda, K., Reeves. A. G.: Seven cases of cerebro-medullospinal disconnection: The "locked-in" syndrome. Neurology (Minneap.) **21**, 1140-1148 (1971)
30. Peiper, A.: Die Eigenart der kindlichen Hirntätigkeit. Leipzig: Thieme 1961
31. Plum. F.: Organic disturbances of consciousness. In: Scientific foundations of neurology, M. Critchley, J. L. O'Leary. B. Jennet [eds.), pp. 193-201. Philadelphia: Davis 1972
32. Plum. F., Posner, J. B.: The diagnosis of stupor and coma. Philadelphia: Davis 1966
33. Poeck, K.: Bemerkenswerte Svmptomkombination bei Basilaristhrombose. Nervenarzt **29,** 317 (1958)
34. Poeck, K.: Pathophysiology of emotional disorders associated with brain damage. In: Handbook of clinical neurology. P. J. Vinken, Ci. W. Bruyn (eds.). Vol. III. pp. 343-367. Amsterdam. New York. Oxford: North Holland 1969
35. Poeck, K., Pilleri, G.: Pathologisches Lachen und Weinen. Schweiz. Arch, Neurol. Neurochir. Psychiatr. **92**, 323-370 (1963)
36. Prior, P. F.: The EEG in acute cerebral anoxia. Amsterdam: Excerpta Medica 1973
37. Schiffter, R., Schliack, H.: Bewußtseinsverlust. akt. neural. **2**. 69-72 (1975)
38. Scott, M., Lennon,. H. C.: Decerebrate tonic extensor convulsions as a sign of occlusion of the basilar artery. Arch. Neurol. Psychiatry **44**, 1102-1108 (1940)
39. Shafey, S. Scheinblum, A. F., Scheinberg, P.: The ventral pontine syndrome. Trans. Am. Neurol. Assoc. **93**. 21 (1969)
40. Sherrington, C. S.: Experiments on the value of vascular and visceral factors for the genesis of emotion. Proc. Roy. Soc., London **1900**. 390
41, Sherrington, C. S.: The integrative action of the nervous system. New Haven: Yale University Press 1920
42, Silverstein, A.: Acute infarctions of the brain stem in the distribution of the basilar artery. Confin. Neurol. **24**, 37-61 (1964)
43, Walker. A. E.: Brain herniations. In: Handbook of clinical neurology. P. J. Vinken, G. W, Bruyn (eds.), Vol. I, pp. 550-573. Amsterdam: North Holland 1969
44, Wilkus, R. J., Harvey, F., Moretti, L., Ojeman, Lettich, E.: Electroencephalogram and sensory evoked potentials. Findings in an unresponsive patient with pontine infarct. Arch. Neurol. (Chicago) **24**. 535-544 (1971)

Received February 14. NM