letters to the editor

Membership in ASIM encouraged

I would like to remind the members of the AAN of an important policy decision of the Academy and to request active support for the project.

Three years ago the Executive Board established a liaison with the American Society of Internal Medicine, an association which represents internists, neurologist, and other subspecialists in the socioeconomic areas of medicine. Specifically, ASIM:

1. Supports through policy and action the repeal of federally mandated health planning, including CON restrictions.
2. Fought successfully against patient package inserts.
3. Provides practice management services for members and assists in resolving reimbursement problems with third parties.
4. Represents the interest of neurologists and their patients on local and state issues through ASIM state societies.

ASIM recently adopted the following resolution that addresses an area of great concern to neurologists, third party restrictions on reimbursement for CT scans:

> "Third party coverage for a technical procedure performed by a physician competent by training and experience in the procedure, should not be denied solely because of differences in the setting in which the procedure is performed (as long as the setting is medically appropriate) or differences in the specialty designations of the physician performing the procedure.
>
> "Reimbursement for covered technical procedures performed in the ambulatory or outpatient setting should be at a level at least equal to the level of payment in the inpatient setting."

The statement is based on an AAN resolution introduced into the 1981 ASIM House of Delegates. ASIM will use its resources to urge adoption of this policy by health insurers and will support the AAN in introducing the resolution to the AMA.

The Academy together with seven other subspecialty societies has a voting delegate status in the House of Delegates of ASIM. In addition, the Practice Committee of the AAN receives organizational help, legislative tracking information, and the benefit of ASIM's political and socioeconomic experience through the parttime services of Bob Doherty of the ASIM staff. AAN delegate status assures neurologists of a significant voice in the policy decisions of the ASIM. Neurologists, as ASIM members, may participate through the ASIM grass roots structure at local, state, and national levels.

We must strengthen the voice of neurologists in ASIM. To do so we must maintain our delegate status, which requires increasing the number of neurologists who are active ASIM members. Board eligibility or certification in neurology qualifies neurologists for membership. The ASIM House of Delegates meets October 1, 1982. I would like to request that all neurologists consider supporting the Executive Board decision by joining the ASIM. Please call ASIM—800-368-5652—for membership applications and information.

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Transient hyperammonemia in the preterm infant

To the Editor: The article by Ellison and Cowger[1] deserves a few comments. Since the original report of Ballard et al[2] at least 12 patients have been reported instead of only one.[3-9] We agree with the authors that a remarkably favorable evolution is the rule in this condition; however, one of our patients was left with spastic quadriplegia, epilepsy, and severe psychomotor retardation. In our five patients there were no girls and both cases of Ellison and Cowger were boys. Among other reported cases, there were 13 boys and 5 girls. Perhaps this is another example of "male disadvantage"[10] in perinatal morbidity and mortality. Contrary to the experience of Ellison and Cowger,[1] our patients with transient neonatal hyperammonemia were not completely unresponsive to stimuli at the time of severe ammonia elevations. At that specific moment, although they were ventilated and although they showed fixed pupils and had absent vestibulo-ocular responses, they all responded with stereotypic elicited arm movements after the thoracic wall was touched or the lower ribs were gently squeezed. The arm movements were always antigravity movements but the degree of extension and abduction followed by flexion and adduction varied from one baby to the other. On abdominal palpation they also showed jerky movements of the lower limbs like a puppet on a string. The segmental input seemed specific, because movements were not elicited by head movements or by tactile nociceptive or proprioceptive stimuli of the limbs. We have seen this reaction also in newborns with hyperammonemia due to urea cycle defects.

Finally, we do not agree with the authors that "CSF protein content higher than 140 to 150 mg per deciliter may be the clue to an underlying hyperammonemic condition" because values up to 200 mg per deciliter are normal in preterm neonates and higher values are not at all specific for hyperammonemia.[11]

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1981;31:767-70.
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To the Editor: We read with interest the article on transient hyperammonemia by Ellison and Cowger.[1] We have studied a preterm infant with the same disorder who failed to respond to exchange transfusions.

Case report. A 2.5 kg boy was delivered after a normal pregnancy of 35 weeks duration. At 12 hours of age he had generalized seizure activity followed at 24 hours by coma. At 36 hours, the plasma ammonia level was 2575 μg per deciliter. A diagnosis of transient hyperammonemia of prematurity was made by determination of blood citrulline, arginine, argininosuccinate, glycine, and volatile fatty acids, and urine levels of orotic acid. Despite four three-volume exchange transfusions, plasma ammonia remained above 1000 μg per deciliter and showed no response to a 24 hour infusion of arginine (4 mM per kilogram per day). At 62 hours, three hours of hemodialysis through an "umbilical shunt" lowered plasma ammonia from 2184 to 275 μg per deciliter concomitant with complete reversal of the neurologic depression. Plasma ammonia levels continued to fall spontaneously thereafter, and remained less than 100 μg per deciliter, even after a normal protein diet had been instituted. At the time of discharge he appeared neurologically intact, though some chronic pulmonary changes were evident, probably as a result of earlier ventilator therapy.

Survivors of transient hyperammonemia of prematurity have been shown to have normal neurologic outcomes.[1,2] It appears that in most instances adequate therapy can be provided solely by exchange transfusions. However, failure of a patient to respond to exchange transfusions should prompt aggressive management of hyperammonemia since this does not preclude a normal outcome.

Our decision to utilize hemodialysis rather than peritoneal dialysis was based on an earlier experience in treating hyperammonemia secondary to ornithine transcarbamylase deficiency[3] and the expertise with this technique within our institution. In this case, we calculated ammonia clearance to be 40 ml per minute (uncorrected for surface area) at a blood flow through the dialyzer of 60 ml per minute. This is considerably higher than we would have expected using peritoneal dialysis[4] (10 ml/min/M^2) and certainly took much less time to accomplish. The rapidity with which our patient's neurologic depression was reversed prompts us to recommend hemodialysis as an effective and relatively safe therapy in centers equipped for the procedure.

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Reply from the Authors: We appreciate the comments of Jaeken et al. In particular, the stereotypic movement they describe on touching the thoracic wall or squeezing the lower ribs is of interest. We have not used that maneuver in our neurologic examination, but we shall.

We did not suggest that CSF protein of greater than 140 to 150 mg per deciliter was specific for hyperammonemia but only indicated that this could be a clue to the diagnosis. In our neonatal intensive care unit, CSF protein values are generally less than this in preterm infants unless there has been a cerebral disorder such as intracranial hemorrhage. Either the stereotypic movement as described, the elevated CSF protein, or the comatose infant may be sufficient to alert the physician to checking the serum ammonia and making the diagnosis.

We read with interest the additional case of hyperammonemia of the preterm infant described by Dr. Donn et al. Whether or not their patient failed to respond to exchange transfusions is perhaps open to question. Four exchange transfusions were performed with decrease in plasma ammonia from 2575 μg per deciliter to a level above 1000 μg per deciliter. Two of the patients in our article required more than four transfusions—one had 10 and another 5.[1] Hemodialysis may well be more efficacious than peritoneal dialysis in the removal of ammonia. But that may not be the important issue. We

have no experience in hemodialysis in our neonatal intensive care unit at The Medical College of Wisconsin but continue to perform occasional peritoneal dialyses, more generally for neonates with kidney failure. It would seem more appropriate to utilize those procedures which are efficacious and safe, which is most true of peritoneal dialysis.

As noted in our article, the most important aspect is proper diagnosis so that appropriate therapy can be initiated, yielding an infant who is neurologically intact.

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Reference

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Lateralization of brainstem lesions

To the Editor: We have recently presented preliminary evidence to support the hypothesis that lateralization of brainstem dysfunction can be demonstrated by the use of brainstem auditory evoked potential (BAEP).[1] However, in my opinion, the cases of brainstem disease reported by Oh et al[2] do not support the contention that BAEP can accomplish this goal for the following reasons:

1. Two of the nine cases result from vascular occlusion of the circulation of the pons (one left anterior inferior cerebellar artery). In both cases, no potential was elicited following stimulation of the ear ipsilateral to the side of the lesion. Although this may appear to represent lateralization, no mention of the integrity of the peripheral auditory receptor was made and indeed this structure could have been lesioned by ischemia since the internal auditory artery supplying the cochlea is a branch of the anterior inferior cerebellar artery in 39% of the general population. Similarly, the third case made no mention of the histopathology of the cochlea and a "low amplitude of BAEP with the absence of wave V" may be attributed to a lesion of the peripheral receptor organ.

2. Another case presented was a "left midbrain-pons metastatic tumor" which showed "prolongation of latency to waves II-V" following left ear stimulation and normal BAEP in the right ear (figure 3). However, on analysis of their data, the left BAEP appears to be within normal variation except possibly for hearing loss. Indeed it is the right ear stimulation which appears to have a characteristic brainstem dysfunction, suggested by low amplitude wave V relative to the preceding waves but was reported to be normal.

3. The two cases of right and left midbrain infarction showed prolonged latency of wave V following stimulation of the cochlea ipsilateral to the lesion. This result could be interpreted as either a peripheral hearing loss or a central conduction defect (figure 4). Unfortunately, special audiometric evaluation and latency-intensity function were not performed to clarify this problem in differential diagnosis.[3,4]

In conclusion, I feel that this report has not convincingly demonstrated lateralization of lesions in the brainstem using BAEP. Although I certainly would not discourage the authors from pursuing their goal, I cannot agree that the data provide convincing evidence to support this hypothesis.

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Reply from the Authors: Cases 1 and 2 had sudden hearing loss in the left ear at the time of infarction, which was obvious on clinical examination. Unilateral hearing loss in the lateral pontine syndrome is attributed to infarction of either the cochlear nuclei or the cochlear organ itself. We did not discuss this point because it was not relevant to the central issue of our paper. Case 1 did not have any potential in the left BAEP (figure 1) at 75 dB stimulation. However, at 95 dB stimulation, left BAEP had wave I with latency of 1.96 msec, but no subsequent waves. This shows that the hearing loss in case 1 was predominantly due to infarction of the cochlear nuclei. Case 2 did not have any potential in the left BAEP at 75, 85, or 95 dB stimulation, indicating cochlear infarction as the main cause of hearing loss.

Case 3 did not have any hearing impairment on clinical examination. Clinically, this patient had a classical "locked-in syndrome" without any lateralizing signs. On the basis of BAEPs we predicted the lateralization of the lesion, later proved by autopsy. Latency of wave I in the left BAEP was 2.13 msec, but the interwave latencies to waves II-IV were normal. We did not study the histopathology of the cochlea.

Case 5 showed normal latency (1.92 msec) of wave I and prolonged latencies to waves II-V in the left BAEP. Identification of wave I was helped by Cz-Ac recording, varying dB stimulation, and changing of click polarity. Right BAEP was normal by all standards. In cases 6 and 8, wave I latency in the involved side was normal.

Thus, prolongation of wave V latency should be interpreted to be due to central conduction defect. In fact, the left BAEP in case 6 (figure 4) had prolonged latency of wave I but normal interwave latencies.

Unlike animal experiments, the study of human clinical data is often marred by unwanted variables. We admit that some of our cases did not have the ideal "single level lesion" or pathologic proof. However, we believe that our cases represent the best clinical examples available to address this issue. More detailed clinical, BAEP, and pathologic correlations are needed to resolve the complex issue of human "generators" of BAEP waves and their laterality. Since our paper was presented, this issue has been further explored.[1-3] We are pleased to have stimulated this line of investigation.

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Migraine and CSF pleocytosis

To the Editor: I found it personally satisfying to review the report of Bartleson et al on conditions simulating migraine and attended by cerebrospinal fluid pleocytosis.[1] I have seen a 38-year-old woman who noted onset and worsening of slurred speech, right arm numbness, right hand clumsiness, and incoordinated gait over 72 hours. This was not associated with headache, visual disturbance, or dizziness. Examination performed after some improvement showed only minimal impairment of fine, distal movements of the right hand. History disclosed periodic headaches confined to both temples and the frontalnasal region. A maternal aunt had suffered from migraine. Eight years earlier bilateral upper extremity numbness prompted cessation of oral contraceptives. Radionuclide scan and CT were normal. An EEG showed mild slowing over the left midtemporal region. In the following several weeks she exhibited fluctuating episodes, lasting a day or two, of similar speech and right sided body dysfunction, but she also noted more frequent and variable headaches of moderate severity which lacked temporal relationship to other neurologic complaints. Despite renewed complaint of a dead-like feeling in the entire right arm, no objective sensory or motor disturbance was identified, with the exception of an asymmetric plantar response on the right. A three-vessel cerebral angiogram excluded AVM or extracranial carotid disease. Three days later lumbar puncture showed normal CSF pressure with clear and colorless fluid. CSF protein was 37 mg%, glucose 67 mg%, and there were 17 white cells, 100% lymphocytes. CSF immunoglobulin G level was 16.6 mg per deciliter (normal, 0 to 8.6). She was afebrile and there was no peripheral leukocytosis. The ESR was 25 mm per hour. Collagen-vascular workup and CSF microbiologic studies were negative. No serologic tests for viral infection were performed. Cold agglutinin study was not elevated.

The unilateral symptoms and signs, as well as the history of headache and later acknowledgment of more severe headaches, led us to consider complicated migraine, though the temporal profile of headache offered weak correlation with other complaints. Further, there was no family history of hemiplegic migraine. Even the sterile CSF pleocytosis was accepted as possibly related to migraine proper, though it might have been a response to angiography performed 72 hours earlier. The elevated CSF IgG, though not pathognomonic, pointed strongly to demyelinating disease.

Others have commented about severe headaches associated with the cerebral hemiplegic form of multiple sclerosis.[2] It may well be that this woman had two common diseases: migraine and multiple sclerosis. On the other hand, a 38-year-old woman developing apparent classic signs of migraine for the first time in a setting of CSF pleocytosis and elevated quantitative IgG raises the issue of a migrainous syndrome precipitated by an acute expression of demyelinating disease. A course of time may prove helpful to further understanding this patient in question. The observation of Dr. Bartleson et al should serve to arouse suspicion of certain migrainous syndromes, but I'm not entirely in agreement with their final conclusion suggesting that such a constellation of symptoms and CSF findings constitutes a benign syndrome.

The patient was seen after the above was written. She has new complaints of paresthesias of hands and feet, and as often as 40 times a day had abrupt loss of tone on the right side of her body with slurred speech lasting only seconds. These paroxysmal features might suggest demyelinating disease, but she has also enjoyed reduction of headaches on propanolol therapy.

Acknowledgment

I would like to thank Dr. Gerhard Witte for the kind referral of the above described case.

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with cerebrospinal fluid pleocytosis. Neurology (Ny) 1981;31:1257-62.
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To the Editor: We are interested in the report by Dr. Bartleson[1] and colleagues of six cases of a migrainous syndrome with pleocytosis. Our experiences with a similar patient lead us to doubt, however, the etiology of this syndrome.

A 33-year-old man was admitted with an acute onset of Broca aphasia with right hemianopsia and right hemiparesis. He had a severe diffuse headache and his temperature was 101° F. EEG showed a left temporal theta focus. CT scan and arteriography were normal. The spinal fluid showed 96 lymphocytes and 4 polys per cubic millimeter. CSF protein was 81 mg% and glucose 64 mg%. All other CSF studies including viral titers, electrophoresis, cultures, and serology were negative. All neurologic deficits resolved in twelve hours, and over 2 weeks the cell count and protein returned to normal.

Subsequently, he has been readmitted with two identical episodes marked by aphasia and right hemiparesis with CSF pleocytosis. Headache was not a prominent feature. All other studies have remained normal.

This case resembled those described by Bartleson et al,[1] in being a young man with stereotyped recurrent focal neurologic signs, lasting less than 1 day, accompanied by CSF lymphocytosis and elevated protein. Diffuse headache was a prominent feature of the first episode but not of subsequent attacks.

We agree with the assessment that the diagnosis of migraine in these cases is uncertain. As the authors point out, CSF abnormalities are rare in migraine of any kind, including hemiplegic varieties. Headache is very common, however, in almost any infectious or inflammatory process involving the CNS or meninges.

The etiology of the lymphocytic meningitis and headache in our case and those described by Bartleson et al is not known, but it probably belongs in the group of chronic recurrent meningitis. Bruyn[2] described a very similar case as being due to Mollaret meningitis, pointing out that transient focal neurologic findings do not exclude this diagnosis. The clinical picture can be in every way identical, no infectious agents can be isolated, and the CSF "endothelial cells" characteristic of Mollaret's can only infrequently be detected. Headache with nausea and visual symptoms are well described in Mollaret meningitis.[3] We feel this is a more probable explanation of this syndrome. We certainly do not feel, as Dr. Bartleson implies, that aggressive workup is unnecessary in patients with this presentation.

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To the Editor: The article by Bartleson et al[1] prompts us to report the following case.

A 21-year-old right-handed man with a personal and family history of common migraine suddenly experienced numbness and tingling of the right hand. Within 5 minutes, paresthesias involved the entire right half of the body. Ten minutes later, although comprehension was normal, he could only say "yes" or "no;" simultaneously he felt sleepy and "as if his visual field was narrowed on both sides," as he told us later. All symptoms regressed within 3 hours of onset and, except for motor aphasia during this spell, neurologic and general examination consistently gave normal results; there was no fever. His usual moderate bifrontal headache with nausea preceded other symptoms by 2 hours and lasted for 24 hours. Blood erythrocyte, leukocyte, and platelet counts; erythrocyte-sedimentation rate, VDRL, serum electrolytes, glucose, and BUN; and enhanced brain CT were normal or negative. EEG showed bilateral slowing that cleared in a few days. On day 2, CSF contained 193 lymphocytes and 32 histiocytes and polymorphonuclear neutrophils per cubic millimeter; protein content was 116 mg per deciliter and glucose level 79 mg per deciliter; cultures for bacteria, the VDRL, and gamma globulin were negative or normal. On day 22, CSF contained 80 lymphocytes per cubic millimeter and protein was 70 mg per deciliter. On day 70, protein content was 44 mg per deciliter and lymphocytes 8 per cubic millimeter. Six months later, the patient had remained free of neurologic symptoms but had had one further episode of common migraine.

Migraine is merely a clinical diagnosis and we agree with Bartleson et al that the distinction between asceptic meningitis with secondary migraine and migraine with secondary meningeal reaction may be hazardous. However, in our patient, the following favors the latter hypothesis: personal and familial history of migraine; absence of fever and other features of infection; absence of increased gamma globulin content in CSF and therefore "non-meningitic," "transudative" CSF profile;[2] the absence of such symptoms in proved cases of viral meningitis or meningoencephalitis might be an other argument for a primary migrainous mechanism. The benign course of the syndrome must be emphasized and possibly harmful investigations or treatment should be avoided.

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Reply from the Authors: We are gratified by the interest our publication has elicited and thank the various authors for their observations and comments.

There are several differences between the seven patients we reported and Doctor Novom's case and we are not convinced that the case he describes is an example of "a migrainous syndrome with CSF pleocytosis." Our patients had no history of migraine-like headaches or other neurologic symptoms before or after their 1- to 12-week illness. Unlike Doctor Novom's patient, the duration of our patient's neurologic symptoms and the clear connection between those symptoms and headache were consistent with the classic form of migraine. All of our patients had at least one episode affecting each cerebral hemisphere, eliminating the possibility of a single, focal lesion. Our patients' CSF abnormalities included elevated opening pressures, an average peak protein level of 103 mg percent, and an average CSF pleocytosis of 121 leukocytes per cubic millimeter, whereas Doctor Novom's patient had normal CSF opening pressure and total protein and only 17 lymphocytes per cubic millimeter. Doctor Novom's patient had an elevated CSF IgG level which raised his suspicion about possible multiple sclerosis. While the increased total protein levels in our patients make such ratios unreliable, the CSF IgG level expressed as a percentage of total protein was less than 20 percent in each of our five patients in whom it was determined. Also, the CSF cell counts and protein levels in our patients are excessive for multiple sclerosis.[1] A diagnosis of multiple sclerosis seems quite unlikely in our patients.

Dr. Rolak et al suggest that their patient's three episodes of focal neurologic deficit, only one of which was accompanied by headache, may be due to Mollaret meningitis, another syndrome of unknown etiology. We learned by telephone communication that their patient's three episodes occurred during a period of 4 weeks. We do not believe that the seven cases we described had Mollaret's type of recurrent meningitis. Rather, our patients had multiple migrainous attacks during a single, self-limited illness which lasted just 1 to 12 weeks and they had no recurrence in follow-up averaging more than 2.3 years. Patients with Mollaret meningitis have recurrent, separate episodes of meningitis over a period of 1 year or more with symptom-free intervals lasting weeks, months, and sometimes years.[2] In addition, patients with Mollaret meningitis have prominent meningeal signs with positive Brudzinski and Kernig maneuvers but the patients with a migrainous syndrome with CSF pleocytosis did not. While transient neurologic phenomena such as generalized seizures, hallucinations, coma, diplopia, facial weakness, anisocoria, and Babinski signs may be seen in Mollaret meningitis,[2] distinctly migrainous sequences of neurologic symptoms followed by headache are rarely, if ever, seen. The presence of characteristic, large, fragile, "endothelial cells" in the CSF serves as a further differentiation between Mollaret meningitis and other causes of CSF pleocytosis. Only additional follow-up will reveal whether the case described by Dr. Rolak experiences recurrent attacks as in Mollaret meningitis or if he has now recovered from his "migrainous syndrome with CSF pleocytosis" as we suspect. In either event, an eventual, complete recovery seems assured.

The case described by Dr. Lhermitte et al had a single, transient episode of focal neurologic deficit associated with headache and accompanied by CSF abnormalities that persisted for some 10 weeks. Because their patient had a long history of common migraine headaches, Dr. Lhermitte et al understandably emphasize the possibility that the CSF findings were secondary to migraine. Our patients had no personal history of migraine and hence we favor the possibility that their multiple migrainous episodes were symptomatic of an underlying, inflammatory disorder of the central nervous system. We agree with Dr. Lhermitte et al that patients with the syndrome we described enjoy a benign outcome and that possibly harmful, invasive procedures, such as cerebral angiography, can generally be avoided.

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Corrections

"Fisher syndrome in childhood" (letter) by I. Derakhshan, July 1982, p. 787. The final sentence should end after the word "ganglion."

"Long-term follow-up after cerebral hemispherectomy: Neurophysiologic, radiologic, and psychological findings" by C. M. Verity, E. H. Strauss, P. D. Moyes, J. A. Wada, H. G. Dunn, and J. S. Lapointe; June 1982, p. 636. Figure 3 is upside down.