To the Editor: In adults, palatal myoclonus occurs months after the onset of a lesion involving the triangle of Mollaret (inferior olive, red nucleus, and contralateral cerebellar nuclei); it may be caused by an infarct, demyelination, or a neoplasm. There have been few cases in children and none in which palatal myoclonus was the presenting feature of an evolving lesion, as in the following case.

C.M., a 5½-year-old boy, had been treated for acute lymphocytic leukemia with prednisone, intrathecal methotrexate, and cranial irradiation. He had been in bone marrow and CNS remission for 2 years, with no clinical or CT evidence of latent adverse effects. Except for a mild respiratory infection, he had been well; following a dose of intrathecal methotrexate, he was admitted for the onset of "twitching" eyebrows, dysphagia, and dysarthria.

Examination revealed a cushingoid boy with no fever, rash, or striae. He was alert and followed three-step commands, but did not speak and uttered only broken syllables. Cranial nerve functions were normal except for continuous, rhythmic, "blinking" contractions of the orbicularis oculi muscles. The movements were bilateral, sometimes more pronounced on the right. Similar movements of the soft palate were synchronous with the orbicularis movements, at a rate of about 1.5 per second. The eyes were not affected; pupils and fundi were normal. Facial strength was equal and full, and a gag reflex was present. Proximal limb wasting and weakness were attributed to steroid myopathy.

During the next week, right facial contractions appeared and were asynchronous with the orbicularis and palatal movements. Following this, right hand and toe jerks appeared, also out of phase with the facial movements. The palatal myoclonus became intermittent. After the fifth hospital day, the child became lethargic.

Brainstem evoked potentials and brain CT were normal. EEG revealed a left slow-wave focus that followed the facial contractions but preceded limb jerks (figure). Later, the EEG showed encephalopathic changes compatible with the child's lethargy. On the second day, the CSF fluid was acellular with normal protein and glucose, but the peripheral blood white cell count was less than 1,000. Though smears and cultures for bacteria, tuberculosis, and fungi were negative, viral cultures yielded a picornavirus. On the ninth hospital day, CSF was still normal except for myelin basic protein content of 8.4 mg/ml. On the sixteenth day, the CSF showed 15 white cells and protein of 57 mg/ml. By that time, encephalitis was manifest by obtundation, slow EEG, and diffusely increased contrast in cortical areas on CT. By the end of the second week, there were asynchronous myoclonic jerks of the eyes, right side of the face, left shoulder, and right hand and foot. Phenytoin and phenobarbital had no effect; IV diazepam, administered under EEG monitoring, abolished the movements without affecting the EEG. The myoclonus, palatal and generalized, was completely controlled by a combination of clonazepam (0.08 mg/kg/d in three divided doses) and diazepam (1 mg po tid).

A brain biopsy on the seventeenth day revealed microglial nodules suggesting an acute viral infection, but cultures for herpes and picornavirus were negative. The white matter appeared normal with no leukemic infiltrates. Over the following 6 months, the patient remained free of myoclonus, with a normal mental status but with persistent dysarthria and dysphagia as well as a right hemiparesis.

Matsuo and Ajax suggested denervation supersensitivity as the underlying mechanism for palatal myoclonus. They noted the "obligatory" lag time between onset of a lesion (hemorrhage, infarction, and so on) and the appearance of the palatal myoclonus. The onset of the movement disorder was always at least 2 months after the inciting event, with a mean interval of 10 to 11 months. Thus, in all reported cases, palatal myoclonus has been a new symptom related to an old, established lesion.

Our patient had been in remission from acute lymphocytic leukemia.
nia for over 2 years. He had received intrathecal methotrexate and cranial radiotherapy. There was no CT evidence of leuкоencephalopathy, and all the clinical and pathologic evidence pointed to an acute encephalitis.

On electrophysiologic studies, EMG spikes of facial muscles were followed sequentially by a cortical discharge on EEG and the limb contraction (figure). This sequence was consistent with brainstem or subcortical origin of the discharge, spreading sequentially to adjacent facial muscles, cortex, and finally to the limbs.\(^1\)

The onset of palatal myoclonus in this patient correlated with the acute onset of encephalitis, presumably due to a picornavirus, and was the first sign of brainstem dysfunction. CSF, brainstem evoked potentials, and CT were normal at that time. EEG confirmed the subacute onset of encephalitis, presumably due to a picomavirus, and was cranial radiotherapy. There was no CT evidence of leukoencephalopathy.

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Alcohol improves dystonia: Opioid system involvement?

To the Editor: The finding that ethanol infusions improve dystonic scores in patients with spasmodic torticollis may provide some insight about neurotransmitter dysfunction in this disorder. Analgesic doses of nitrous oxide also ameliorate the symptoms,\(^5\) and nitrous oxide interacts with the endogenous opioid system (EOS) in animals\(^6,7\) and humans\(^8,9\) as well as in vitro receptor-binding assays.\(^10,11\) Some of the effects of ethanol may be mediated by the opioid system,\(^12\) and nitrous oxide ameliorates the alcohol withdrawal state.\(^13\) Therefore, the beneficial effects reported by Biary and Koller\(^1\) may have been produced by stimulation of the EOS.

If so, their data would support our hypothesis that there is an underactivity of the EOS in torticollis.\(^2\)

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Respiratory dyskinesia in Parkinson’s disease

To the Editor: De Keyser and Vincen~\(^1\) described a patient with Parkinson’s disease who had levodopa-induced respiratory disturbance that was suppressed by tiapride, a dopamine receptor blocker. We recently studied a similar patient with levodopa-induced respiratory dyskinesia that was managed by adjusting the dosage of levodopa, which we think is a better approach.

A 77-year-old man had Parkinson’s disease for 8 years, with good control of his symptoms by Sinemet 10/100, one-half tablet three times a day. For 6 months, he had episodes of tachypnea, dyspnea, and perpiration. The episodes occurred 30 to 40 minutes after each dose of Sinemet and lasted 15 to 45 minutes. Cardiopulmonary evaluation was normal. In the hospital, several respiratory episodes were time-related to taking Sinemet. There were also orofacial dyskinetic movements during the attacks. Three to 4 hours after Sinemet, the anti-Parkinson effects wore off, and he had hypomimia, tremor at rest in the hands and legs, moderate cogwheel rigidity, stooped posture, and shuffling gait without armwaving.

On the third hospital day, the Sinemet was discontinued, and the respiratory attacks disappeared within 24 hours. After baseline spirometry (figure 1) the patient was given one tablet of Sinemet 10/100, and within 1 hour, his respiratory rate rapidly increased from 8 breaths per minute to a rate of 30 per minute. In addition, the minute volume increased from 15 l/min to 40 l/min, but arterial blood gases did not change. The respiratory distress was associated with orofacial dyskinesia and with profuse diaphoresis. By adjusting the Sinemet dosage to one-fourth tablet of Sinemet 10/100 six times a day, the Parkinson symptoms have been well controlled, and he has had no further respiratory symptoms.

Respiratory dyskinesia is an uncommon complication of levodopa therapy.\(^2\) It may be associated with laryngeal dystonia or other dys-
induced dyskinesias "without increasing parkinsonian disability." However, tiapride was presumably selected as an antidyskinesia agent because of its preferential effect on the mesolimbic system. In contrast, the other neuroleptic drugs block the nigrostriatal pathway and thus may have a more adverse effect on the Parkinson symptoms. Despite this theoretical differentiation, tiapride may increase parkinsonian disability. Our patient illustrates that levodopa-induced dyskinesias can be controlled by merely adjusting levodopa dosage, rather than by blocking the dopamine receptors with tiapride which, despite some reports, might exacerbate parkinsonism. Finally, this dopamine-antagonist is not readily available to clinical neurologists in the United States.

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Reply from the Authors: The respiratory disturbance described by Jankovic and Nour differs in several ways from the one we reported. In their patient, it was a side effect of chronic levodopa therapy, was associated with orofacial dyskinesia, and consisted of a marked increase in respiratory rate. From figure 1, their patient was already tachypneic at baseline spirometry (respiratory rate, 36 per minute), but the reason is not clear from their letter. In our patient, the respiratory disturbance occurred after starting levodopa therapy, was not associated with orofacial or limb dyskinesia, and was characterized by a breathing pattern that was irregular in rate and depth.

Although the underlying mechanism may be completely different, in both cases dyspnea was related to intake of levodopa. We entirely agree that if the side effect can be avoided by simple adjustment of levodopa dosage, that is the most logical approach. In our patient, parkinsonian disability was severe, and doses of levodopa 200 mg three times a day were required to obtain favorable improvement. At doses of 100 mg of levodopa, she already experienced dyspnea, and a simple adjustment of levodopa dosage would have been not as effective as administration of tiapride.

Tiapride may indeed increase parkinsonian symptoms, but much less than classic neuroleptic drugs and other starting levodopa therapy, such as metoclopramide or sulpiride. However, it does not significantly increase parkinsonian disability if the levodopa dosage is adjusted concurrently. Careful adjustment of tiapride/levodopa therapy can suppress dyskinesias and allow maximal antiparkinsonian benefit from levodopa.

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An additional pilomotor seizure

To the Editor: In view of the recent interest in pilomotor seizures, we describe one more case.

A 56-year-old man was admitted because of diffuse headaches for 1 month. For 2 weeks he noted an unusual smell, which was followed by the feeling that his hair was standing on end. This sensation began in the legs and spread to the arms, trunk, and head; it was stronger on the left. He felt anxious, sensed flushing in the face, and was "in a cold sweat" during the 1-minute episodes. Afterwards he felt tired. He sometimes had nausea but no epigastric symptoms. There were no attacks for 1 week, but then they recurred. Examination showed only symmetric horripilation. The vital signs, skin color and temperature, consciousness, speech, and behavior remained normal. Contrast-enhanced CT was normal, but none was obtained during an attack. The diagnosis was uncinate seizures with autonomic features, and he was given phenytoin, 300 mg daily. The attacks did not recur.

Three months later he was readmitted after a complex partial seizure that involved the right arm. He had aphasia, left papilledema, right homonymous field cut, and right hemiparesis. CT showed a large right temporoparietal, white-matter, ring-enhancing lesion with marked mass effect. Left parietal craniotomy disclosed a glioblastoma multiforme. He died 2 weeks later.

This is the sixth report of pilomotor seizures. In four cases, a seizure disorder began with this form of seizure; in two, other types of seizure preceded the onset of pilomotor seizures. Piloerection was asymmetric in four cases; ipsilaterally predominant in ours, starting ipsilaterally to the lesion and generalizing in two. Anticonvulsant controlled symptoms in three patients. They had no effect in one, although craniotomy had a transiently beneficial effect. Seizures appeared after temporal lobectomy in one patient and continued after that procedure in another.

Glioblastoma multiforme was responsible for four of six cases. A possible antecedent infection and prior epilepsy with temporal resection were noted in the others. The infiltrative nature of astroglial tumors with preservation of normal structures accounts for the selectivity of manifestations. The high frequency of astrocystomas means they will be the cause of most cases of pilomotor seizures. Pilomotor seizures are more common than is apparent; they may be transient or obscured by more severe symptoms. The symptoms may also not be mentioned by patients unless specifically sought.

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Cysticercosis acquired in the United States, without compatible travel history

To the Editor: There have been few reports of acquired cysticercosis in the United States since 1979 and apparently no previous reports of cerebral cysticercosis in an urban-dwelling American who never traveled and whose family members never traveled outside this country.

This 6-year-old girl, well until the evening of admission, suddenly had difficulty speaking and fell to the right. Minutes later, her eyes deviated to the right, and there were right-sided tonic-clonic move-

ments that became generalized. In the emergency room, the seizure stopped. On examination, she was afebrile, with an extensor plantar response on the right. Thereafter, the hospital course was unremarkable. She was treated with phenytoin. CT showed an increased density in the periphery of the left parietal lobe, with no enhancement (figure). Skull films were negative. CSF, routine blood studies, and EEG were negative. The serum titer of antibodies for cysticercosis was 1:125; 6 weeks later, it was 1:64 (normal, less than 1:32). No antibodies were found in CSF.

The diagnosis was suspected because of the calcified lesion on CT, the afebrile seizure, and the history of frequent pork ingestion. In one series of children with cerebral cysticercosis, lesions were seen by CT in all cases, but plain skull films were abnormal in only 10%. When there were completely calcified lesions, they were usually found in the peripheral areas of the cerebral hemisphere, as in our case. Our patient had seizures, as in the vast majority of children with cysticercosis.

This case raises several public health concerns. Some pork in the United States may be infected with the larval form, *Cysticercus cellulosae*, and is not being prepared properly for human consumption. Also, egg-harboring feces of infected individuals may contaminate the food or drink of individuals who have never traveled to endemic regions, which better explains the etiology in our patient since the family denied eating any undercooked meat.

Cerebral cysticercosis may be diagnosed serologically in more patients with afebrile seizures and appropriate CT abnormalities, even when there is no compatible travel history.

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Dystonia and calcification of the basal ganglia: another case

To the Editor: Larsen described a family with autosomal dominant dystonia, calcification of the basal ganglia, and no abnormality of calcium metabolism. We studied a child with a similar disorder. He was born in 1973 from healthy parents; there was no familial history or consanguinity of similar disorder. After normal psychomotor development, at age 6 years, he began to show impaired speech; at age 8, pigmentary degeneration of retina was recognized, ERG was extinguished, and brain CT was normal; at age 9, there was symmetric progressive axial and segmental dystonia. At age 10, symmetric calcification of basal ganglia was evident on CT (figure).

Now, at age 12 years, his speech is almost incomprehensible because of dysarthria; the limbs are fixed in abnormal dystonic postures. Dementia is not overt, but it is difficult to evaluate IQ. Plasma and urine amino acids, lipoprotein electrophoresis, oligosacchariduria, leukocyte aroylsulfatase A, θ-beta-hexosaminidase and θ-galactosidase, CSF proteins, EEG, ECG, and ultrastructural skin biopsy examination were normal. Studies of calcium, iron, and copper metabolism revealed no abnormality.

The association of pigmentary degeneration of retina and progressive extrapyramidal symptoms may suggest Hallervorden-Spatz disease, but no diagnostic laboratory test is available, and that diagnosis could be confirmed only by postmortem examination. CT changes reported in Hallervorden-Spatz disease differ from those of our patient, and resemble the patterns of Huntington’s disease.

We do not believe that calcification of the basal ganglia in this case is a coincidence, as it may be in the elderly.

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Palatal myoclonus and ataxia: another case

To the Editor: Sperling and Herrmann’s report of palatal myoclonus and progressive ataxia reminded me of a similar patient. A 48-year-old school administrator was seen in 1983 after 4 years of progressive difficulty with speech, which had become indistinct and diminished in volume. He also had noted difficulty keeping his balance, but only in strenuous athletic activities. He had no other symptoms except for a perceived increase in salivation. Shortly after onset, CT, EEG, skull x-ray, and repetitive nerve stimulation were normal. He had abused alcohol for several years, but had never had liver failure, GI hemorrhage, jaundice, coma, or seizures. There was no family history of neurologic disease.

On examination, he was alert and mentally intact. There was palatal-pharyngeal myoclonus at approximately 130 beats per minute, perceptible through the skin of the neck. I could discern no involvement of the eyelid, respiratory muscles, larynx, tongue, or ocular muscles. His speech was dysarthric; rapid repetitive movements of the tongue were slow, with neither wasting nor fasciculations of the tongue. The cranial nerve functions were normal. He had clumsy rapid repetitive movements of both arms, but no weakness or change in tone. The remainder of the neurologic examination was normal.

Routine laboratory tests were normal, as was CSF, including gamma globulin and oligoclonal bands. Brainstem auditory evoked potentials revealed wave I with stimulation of either ear; however, the subsequent responses were poorly defined and poorly reproducible between trials (Dr. Michael J. Aminoff). CT revealed a punctate area of calcification in the region of the fourth ventricle, probably calcification of the choroid plexus. Magnetic resonance imaging (Dr. David Norman) revealed only high signal intensity in the periventricular white matter. The brainstem appeared normal, save for a somewhat enlarged fourth ventricle.

The patient was unaware of the palatal myoclonus. As it was asymptomatic, I did not try to suppress it pharmacologically.

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Walter E. Dandy’s bookplate

To the Editor: May I offer two corrections and an additional fact to Dr. Nicholas J. Lenn’s article on “Walter E. Dandy’s Bookplate” in the July issue of Neurology.

Dandy’s illustrator, who prepared the bookplate, was Doras Hager Padget. Her maiden and married names are misspelled in the article. The light shining up toward the brain in the bookplate is not an x-ray tube (as Lenn states), but the headlight that Dandy wore when he operated. Part of the bulb was blackened to avoid diffusion of light.

An interesting feature of the bookplate, which Dr. Lenn overlooked, was the pair of rongeurs whose handles flank the headlight. As far as I know, Mrs. Padget was entirely responsible for creation of the bookplate.

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