Effect of stress related to the 9/11/2001 terror attack on seizures in patients with epilepsy
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Abnormal co-contraction in yips-affected but not unaffected golfers: Evidence for focal dystonia

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Up to 30% of golfers develop the yips, an inability to complete a golf stroke, most often affecting short putts, which worsens with anxiety. Yips may be organic (task-specific dystonia) or psychological (anxiety or “choking”). We previously found abnormal trains of 4 to 8 Hz, rhythmic, co-contracting bursts of EMG activity in arm muscles of three golfers suggesting a movement disorder. This led to the current investigation.

Methods. We studied 20 age- and handicap-matched male right-handed golfers, 10 with the yips and 10 without. Handicap is the golfer’s average score over par over the past 10 rounds of golf. Surface EMG electrodes were placed bilaterally on the pectoralis major, deltoid, biceps, triceps, wrist flexors, pronator teres, flexor pollicis longus, wrist extensors, abductor pollicis brevis, and abductor digiti minimi. EEG electrodes were in a standard montage. Recordings were made at a sampling rate of 1,000 Hz, bandpass 1 to 200 Hz using the Neuroscan system (Neuroscan Compumedics, El Paso, TX). Investigators were not blinded. Pre-putting conditions at rest and with arm activation maneuvers, including writing, were recorded. Somatosensory evoked potentials (SEP) were performed by median nerve stimulation at 2.2 Hz using an average ear reference.

Putting was performed on an indoor, artificial, flat putting green surface that was 12 feet long with a hole 2 feet from one end. Subjects were evaluated standing with arms relaxed with and without holding the putter. They then performed 75 putts: 30 putts of 6 feet, 10 putts of 3 feet, and 35 putts of 8 feet. The number of putts made and distance from the hole were recorded for each putt. An electronic photocell recorded the initiation of each stroke and the point of impact.

EEG-EMG polygraphy was assessed for abnormal activation of muscles including oscillating discharges, co-contractions, and high-amplitude short duration discharges. The N30 amplitude was measured from the averaged SEP at Fz and Cz. Rectification of EMG activity for all leads was performed 6 seconds before the first putter movement, through to at least 2 seconds after the ball was hit.

Results. There was no difference in age or current handicap, but yips-affected golfers had better “best” handicaps and had been golfing for longer than yips-unaffected golfers (table). Prior to putting no abnormal movements were found. During putting two yips-affected golfers reported feeling the yips, yet all yips-affected golfers appeared to have a visible jerk or twist of the wrist or forearms. There were no differences between groups in right or left forearm EMG activity being dominant, wrist flexor vs wrist extensor activity being dominant, or whether there was phasic flexor or extensor bursting. At 200 ms prior to impact of the putter with the ball there was co-contraction of wrist flexor and extensor muscles in 5/10 of the yips-affected golfers (in all putting conditions) and 0/10 of the yips-unaffected golfers (p = 0.06, exact McNemar test). The rest of the golfers had either no specific pattern or had reciprocal wrist flexor and extensor activity (6/10 yips-unaffected vs 3/10 yips-affected). SEP data revealed significant (one electrode) or trend (two electrodes) to higher amplitude N30 waves in the yips-affected group (see table).

There was a trend for yips-affected co-contracting golfers to be older, have higher handicaps, have yips for fewer years, make fewer putts, and have a greater degree of error in missing the putts than the non-co-contractors.

Discussion. Electrophysiologic analysis of age- and handicap-matched yips-affected vs yips-unaffected golfers found 5/10 yips-affected and 0/10 yips-unaffected golfers have wrist flexor/extensor co-contractions, even with just two of the yips-affected golfers having symptoms during testing. As co-contraction is a hallmark of dystonia, the yips appears to be a movement disorder, such as a task-specific dystonia, in some golfers. The SEP results may also support the yips being a focal dystonia.

Other explanations for the co-contraction might include another abnormal movement physiology or a phenomena called “double pull” in which agonists and antagonists apparently co-contract when under conditions of high arousal (although not studied with electrophysiology). As the co-contracting golfers golfed longer and had lower handicaps they were likely not anxious and 3/5 lacked a subjective feeling of the yips.

Limitations in this study include its small size, laboratory and not golf course location, and unblinded data collection and analysis. Despite these limitations, the results suggest some cases of

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Table Comparison of yips-affected and unaffected golfers (paired t test)

<table>
<thead>
<tr>
<th>Variable</th>
<th>No.</th>
<th>Unaffected</th>
<th>Affected</th>
<th>Difference</th>
<th>p</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>10</td>
<td>49.30 (17.85)</td>
<td>50.30 (14.91)</td>
<td>1.00 (3.77)</td>
<td>0.42</td>
<td>−1.70 to 3.70</td>
</tr>
<tr>
<td>Years golfing</td>
<td>10</td>
<td>25.90 (16.99)</td>
<td>37.60 (12.37)</td>
<td>11.70 (14.58)</td>
<td>0.03</td>
<td>1.27 to 22.13</td>
</tr>
<tr>
<td>Handicap</td>
<td>10</td>
<td>7.80 (6.44)</td>
<td>6.60 (6.28)</td>
<td>−1.20 (3.58)</td>
<td>0.32</td>
<td>−3.76 to 1.36</td>
</tr>
<tr>
<td>Best handicap</td>
<td>10</td>
<td>6.30 (5.54)</td>
<td>3.50 (4.86)</td>
<td>−2.80 (2.62)</td>
<td>0.01</td>
<td>−4.67 to −0.93</td>
</tr>
<tr>
<td>N20L</td>
<td>8</td>
<td>2.07 (0.38)</td>
<td>2.44 (1.02)</td>
<td>0.37 (1.08)</td>
<td>0.36</td>
<td>−0.53 to 1.27</td>
</tr>
<tr>
<td>N20R</td>
<td>8</td>
<td>2.11 (0.70)</td>
<td>2.47 (0.64)</td>
<td>0.36 (1.04)</td>
<td>0.36</td>
<td>−0.51 to 1.24</td>
</tr>
<tr>
<td>P25L</td>
<td>8</td>
<td>1.84 (1.30)</td>
<td>2.60 (1.11)</td>
<td>0.76 (1.05)</td>
<td>0.08</td>
<td>−0.12 to 1.64</td>
</tr>
<tr>
<td>P25R</td>
<td>8</td>
<td>2.19 (1.41)</td>
<td>2.80 (1.06)</td>
<td>0.61 (1.77)</td>
<td>0.36</td>
<td>−0.87 to 2.09</td>
</tr>
<tr>
<td>FzL</td>
<td>8</td>
<td>1.72 (0.50)</td>
<td>2.82 (1.34)</td>
<td>1.11 (1.39)</td>
<td>0.06</td>
<td>−0.06 to 2.27</td>
</tr>
<tr>
<td>FzR</td>
<td>8</td>
<td>1.70 (0.83)</td>
<td>2.29 (0.91)</td>
<td>0.58 (1.41)</td>
<td>0.28</td>
<td>−0.59 to 1.76</td>
</tr>
<tr>
<td>CzL</td>
<td>8</td>
<td>1.40 (0.41)</td>
<td>2.35 (1.00)</td>
<td>0.95 (0.82)</td>
<td>0.01</td>
<td>0.26 to 1.63</td>
</tr>
<tr>
<td>CzR</td>
<td>8</td>
<td>1.44 (0.90)</td>
<td>1.89 (0.85)</td>
<td>0.45 (1.21)</td>
<td>0.33</td>
<td>−0.56 to 1.47</td>
</tr>
</tbody>
</table>

Values are mean (SD).

SEP = somatosensory evoked potentials. L and R refer to side of median nerve stimulation.
the yips are organic and further study on a golf course or under anxiety-provoking conditions is warranted. A better understanding of the yips may lead to advances for other task-specific disorders.

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Presynaptic dopaminergic pathology in Chediak–Higashi syndrome with parkinsonian syndrome

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Chediak–Higashi syndrome (CHS) is a rare autosomal-recessive lysosomal storage disorder caused by mutations of the lysosomal trafficking regulator gene (LYST) on chromosome 1q42. The disease is characterized by partial oculocutaneous albinism, immune deficiency, peroxidase-positive granules in leukocytes and giant granules in other tissues (e.g., neurons, astrocytes), and neurologic disturbances. There are two distinct clinical patterns. In the childhood form, the hematologic system is affected with immune deficiency, frequent bacterial infections, and an accelerated lymphoproliferative phase followed by early death. Neurologic signs and symptoms are uncommon. In the adult form of CHS, which occurs between late childhood and early adulthood, hematologic dysfunction is milder and neurologic signs and symptoms are more prevalent: dementia, pyramidal and spinocerebellar signs and symptoms, peripheral polyneuropathy, and a parkinsonian syndrome. The pathophysiology of the neurologic manifestations is still unknown.

Case report

A 22-year-old man was admitted to the Neurology Department because of progressive gait disturbance for 2 years. From childhood he had had partial albinism of the eyes and a convergent strabismus. During childhood and adolescence, infections often developed and a coagulation disturbance led to frequent bleeding. The general examination revealed oculocutaneous albinism and pes cavus. He had a masklike face plus rigidity of the neck and all extremities; there was no tremor. Saccadic pursuit was present. The patient also presented with paraparesis, which was pronounced distally and on the left side. Deep tendon reflexes were diminished in the arms and absent in the legs. Babinski sign was positive on both sides. Except for reduced vibratory sensation at the ankles, findings from sensory examinations were normal. A stumbling gait caused frequent falls. Neuropsychological tests of memory (immediate and later recall), visuomotor speed, verbal fluency, and constructive abilities, and others were all clearly pathologic. His Mini-Mental State Examination score was 25/30 points.

The blood smear showed peroxidase-positive granules within lymphocytes and neutrophils. The hemogram depicted neutropenia, the bleeding time was delayed to 300 seconds (normal 94 to 193). Spontaneous migration and chemotactic activity of neutrophils and fluorescence-activating cell scanning (FACS) analysis of leukocytes were normal. Tibial nerve somatosensory evoked potentials to the left leg were delayed (P 40 after 51.2 msec). Sensory and motor nerve conduction studies were normal beside both sural nerves where no action potential could be elicited.

Brain MRI and EEG were normal. Disturbed presynaptic dopaminergic circuitry was demonstrated by markedly reduced binding in [123I]N-omega-fluoropropyl-2beta-carbomethoxy-3beta-(4-iodophenyl)nortropane (FP-CIT) SPECT on both sides (figure 1a) on visual analysis. On iodobenzamide (IBZM) SPECT, the quotient of the region of interest was calculated. It was marginally lowered (left side: 1.47; right side: 1.45; normal reference: 1.54 ± 0.05). After an L-dopa test (250 mg), the patient improved from 24 to 8 points in selected areas (handwriting, speech, facial expression, rigidity, postural stability, body bradykinesia) of the Unified Parkinson’s Disease Rating Scale. We initiated continuous L-dopa therapy, and the patient’s condition stabilized. After 1 year of treatment, he stopped taking the medication and became rap-
Effect of stress related to the 9/11/2001 terror attack on seizures in patients with epilepsy

Pavel Klein, MB, BChir; and Leonie van Passel, MD

Stress facilitates seizures in patients with epilepsy. The frequency with which this occurs is uncertain. Estimates range widely, from 14 to 67% of patients affected, mainly because there is no uniform definition of stress. One way of bypassing the problem of the definition of stress is to examine the effects of seizures on seizures of an event that is generally recognized as stressful. The 9/11/2001 terrorist attack was such an event in the United States. In the current study, we examined the effect of 9/11 attack-related emotional stress on seizures among patients with epilepsy in an area directly involved in one of the attacks, Washington, DC, by comparing seizure frequency before and after the attack. In addition, because we had studied the association of subjectively perceived stress with seizures in the same patients before 9/11, we compared the change in seizure frequency following 9/11 with patients’ own assessment of stress as a seizure precipitant.

Methods. Cognitively normal adult patients with epilepsy seen consecutively in the Georgetown University Hospital (GUH) Epilepsy Clinic between 9/12 and 12/20/01 were evaluated. GUH is 3 miles from the Pentagon, the Washington target of the 9/11/2001 attack. Greater Washington area residents present in the area on the day of the attack who had kept seizure diaries for ≥3 months before and 1 month after the attack and had had one or more seizures during the last 2 years were included in the study. Patients with nonepileptic seizures were excluded. Patients interviewed before 10/10/2001 were reinterviewed later in the study. Seizure frequency was assessed using seizure diaries. Average monthly seizure frequency during 3 months before 9/11 was compared with seizure frequency during the month after 9/11. Seizure exacerbation was defined as a >50% increase in seizure frequency. Patients completed a questionnaire rating the severity of emotional stress (absent, mild, moderate, or severe) experienced during and after the attack. Additional questions addressed sleep deprivation, anti-epileptic drug change, noncompliance, drug use, and menstruation.

Table 9/11, stress, and seizures

<table>
<thead>
<tr>
<th>Feature</th>
<th>No. of patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total no. of patients</td>
<td>66</td>
</tr>
<tr>
<td>Stressed/directly affected by 9/11</td>
<td>28 (42)</td>
</tr>
<tr>
<td>Moderately/severely stressed</td>
<td>15 (23)</td>
</tr>
<tr>
<td>Directly affected</td>
<td>6 (10)</td>
</tr>
<tr>
<td>With “stress-related seizures”*</td>
<td>45 (68)</td>
</tr>
<tr>
<td>Seizure exacerbation after 9/11</td>
<td>8 (12)</td>
</tr>
<tr>
<td>In patients stressed by 9/11</td>
<td>8/28 (29)</td>
</tr>
<tr>
<td>In patients not stressed by 9/11</td>
<td>0/38 (0)</td>
</tr>
<tr>
<td>In patients directly affected by 9/11</td>
<td>4/8 (50)</td>
</tr>
<tr>
<td>In patients with “stress-related seizures”*</td>
<td>7/45 (16)</td>
</tr>
<tr>
<td>In patients without “stress-related seizures”*</td>
<td>1/21 (5)</td>
</tr>
</tbody>
</table>

* “Stress-related seizures”: Patients used a questionnaire to evaluate the percentage of their seizures that occurred in association with stress. Fifty-six of 66 (85%) patients filled out the questionnaire before 9/11/01 and 10 after 9/11. Patients were defined as having “stress-related seizures” when they estimated that >10% of all their seizures occur after a perceived change in stress level.
family member died during the attack or the patient was in a building threatened during the attack).

Eight (12%) patients experienced seizure worsening during the month after 9/11 compared with the average monthly seizure frequency during the 3 months before 9/11. The average seizure frequency of patients stressed by 9/11 for the 3 months before 9/11 was 1.44 seizures/patient/month compared with 2.08 seizures/patient during the month after 9/11 (p = 0.18, Wilcoxon two-sample test). Clinical features of patients with seizure exacerbation are presented in table E-1 on the www.neurology.org.

Eight (29%) of the 28 patients stressed by the attack had seizure exacerbation compared with 0 of 38 patients not stressed by the attack (Fisher exact test, p = 0.0005).

Four of eight (50%) patients with seizure exacerbation were directly affected by the attack vs 2 of 58 patients without seizure exacerbation (p = 0.001). Leaving out patients directly affected by 9/11, 4 (14%) of 29 patients stressed by the attack had seizure exacerbation compared with 0 of 38 patients not stressed by the attack (p = 0.028).

Forty-five of 66 (68%) patients perceived their seizures to be stress related ("stress-related seizures") in >10% of all seizures and 35% of patients in >50% of all seizures. Seven of 45 patients with stress-related seizures experienced seizure exacerbation after 9/11 compared with 1 of 21 patients with non-stress-related seizures (p = 0.286).

Discussion. Our study indicates that a stressful event is associated with seizure exacerbation only in a small proportion (12%) of exposed patients with epilepsy, but in a higher proportion (29%) of patients stressed by the event and in 50% of patients directly affected by the event. Our finding is similar to those from two previous studies of seizures during a uniformly recognized stressful event. Eight percent of patients exposed to missiles attacks in Israel during the 1991 Gulf War experienced increased seizure frequency. Twenty-six percent of patients experienced seizure exacerbation during a flood in Holland. By contrast, 33 to 65% patients attribute seizures to stress in reports that use subjective patient evaluation without definition of stress.

We conclude that stress can exacerbate seizures in a minority of patients with epilepsy.

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References

Ictal cardiorespiratory arrest in Panayiotopoulos syndrome

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Panayiotopoulos syndrome (PS) is a syndrome of childhood susceptibility to benign autonomic seizures and affects approximately 6% of children with nonepileptic seizures. The clinical features are infrequent, often single, focal seizures comprising autonomic symptoms; behavioral changes; and other ictal clinical manifestations. Half of the seizures progress to become generalized; two thirds occur during sleep. The seizures can last 5 to 15 minutes, but some are prolonged, lasting hours, constituting autonomic status epilepticus (ASE). Even after severe seizures, the patient recovers within a few hours. Prognosis is excellent. Remission usually occurs within 1 to 2 years from onset. EEG is characterized by multifocal spikes that predominate in the posterior regions. Ictal EEG shows frontal or posterior onset. Ictus emeticus (nausea, retching, and vomiting) is the most prominent autonomic symptom. Other mild autonomic manifestations include pallor, flushing or cyanosis, mydriasis or miosis, cardiorespiratory and thermoregulatory alterations, bladder or bowel incontinence, hypersalivation, and abnormal intestinal motility. We present a patient with severe ASE and cardiorespiratory arrest to illustrate that, although PS seizures are usually benign with eventual remission, affected children may have severe and potentially fatal seizures. 

Case report. The patient is a 6-year-old first child of nonconsanguineous parents. He was born at term after an uneventful pregnancy, and his psychomotor development was normal. Personality and family history is unremarkable.

The seizure occurred while traveling in a car with his parents. He fell asleep but woke up a half hour later complaining of feeling sick and having frontal headache. He then vomited and had visual hallucinations. Subsequently, his eyes deviated to the right and he became pale, unresponsive, and flaccid with incontinence of urine and feces. He vomited again. On arrival to a hospital emergency department after 50 minutes from onset, he was apneic, with no pulse and dilated unresponsive pupils.

He was resuscitated, intubated, mechanically ventilated, and then transferred to the intensive care unit with a diagnosis of ASE. Two hours later, he became responsive and gradually recovered without any neurologic deficit.

Neurologic examination, brain MRI, EKG, and echocardiogram were normal. The EEG showed high-amplitude, right-sided occipital spikes (figure). Two follow-up EEGs, 3 days and 9 months later, showed multifocal high-amplitude sharp-wave complexes mainly in the bioccipital regions. The spikes were accentuated by sleep.

On follow-up 9 months after the seizure, he is well and unmedicated and has no seizures or any other abnormality.

Discussion. This child had a single ASE seizure typical of PS in all aspects but with severe cardiorespiratory symptoms. Breathing and cardiac irregularities have recently been reported in PS, but these may be more common and severe than currently described. Brief apnea or heavy and irregular breathing occurs in 7% and tachycardia is a consistent finding on ictal recordings. One of 48 patients with PS had similarities to our case: a boy aged 3 years was dozing in his mother's car when he had a severe autonomic seizure with cardiorespiratory arrest later witnessed by a physician who resuscitated him.

The pathophysiology of autonomic seizures in PS is unclear. Autonomic seizures and ASE with the symptomatology and sequence as described by Panayiotopoulos are specific to childhood, although 10 to 20% may be due to lesional brain pathology. These seizures do not occur in adults. An explanation for this is that children are more vulnerable to autonomic and autonomic disturbances. It is likely that epileptic discharges from various cortical locations trigger a low threshold for child emetic centers and the hypothalamus, as illustrated with ictal EEG documenting a frontal or posterior origin at a stage of only autonomic manifestations.
Sudden deafness from stroke

V. Leussink, MD; P. Andermann, MD; K. Reiners, MD; W. Shehata-Dieler, MD; T. Günthner-Lengsfeld, MD; and M. Naumann, MD

Acute bilateral deafness often suggests a peripheral otologic disorder or psychogenic cause. Rarely, it is a rare symptom of bilateral temporal lobe infarcts or vertebrobasilar system ischemia. A 74-year-old right-handed woman had sudden onset of complete bilateral deafness, which was noticed by relatives but not the patient herself. Past medical history revealed arterial hypertension and a previous stroke with mild transient hemiparesis on the left side 5 years earlier with no long-term sequelae. The patient's relatives reported a long-standing mild hypoacusis pronounced on the left ear.

At neurologic examination, the patient was alert but did not respond to any verbal commands or environmental sounds, nor did she react to loud noises. She did not realize why her relatives had taken her to hospital. Spontaneous speech was fluent with a few phonematic paraphasias but poorly modulated. Except for brisker deep tendon reflexes on the left, the result of the previous stroke, no discoordination or motor or sensory deficits were detectable.

A complete otologic microscopic examination showed normal findings on the right side and evidence of a previous chronic middle ear inflammatory process on the left side. Pure tone and speech audiometry revealed no responses up to the upper limit of the audiometer output for pure tones at frequencies 125 to 8,000 Hz as well as for speech signals. Auditory inmanitance measurements showed normal tympanograms on both ears. The ipsilateral stapedius reflex was present in all tested frequencies at intensities ranging between 90 and 105 db hearing level (HL) on the right side. On the left side, the ipsilateral stapedius reflex was absent most likely be explained by the chronic middle ear process.

Auditory brainstem evoked response (ABR) elicited with rectangular clicks, 100 μs/phase, showed normal waveform morphology and wave latencies at 80 db HL bilaterally. The ABR thresholds were at 60 db HL on both ears.

Bilaterally, amplitudes, latencies, and interpeak latencies were within normal limits for peaks I to V, presenting no abnormalities of the peripheral acoustic nerve or its pathways across the brainstem.

Diffusion-weighted cranial MRI showed an acute infarct of the left middle temporal lobe, and T2-weighted MRI displayed a contralateral residual defect most likely resulting from the previous infarct of the right middle cerebral artery (figure, A and B). Considerable hypometabolism in [18F]fluorodeoxyglucose (FDG) PET was visible in the left middle temporal, inferior frontal, and occipitotemporal gyri and in the right perfusion area of the middle cerebral artery (see figure, C). A group of 10 healthy subjects served as control for these PET scan findings.
in the differential diagnosis of acute hearing loss. The underlying lesions in temporal lobes or brainstem may be small but are detectable by modern imaging techniques.

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References

Spontaneous CSF leak treated with percutaneous CT-guided fibrin glue
Jonathan P. Gladstone, MD; Kent Nelson, MD; Naresh Patel, MD; and David W. Dodich, MD

Spontaneous intracranial hypotension (SIH) is characterized by CSF hypovolemia usually occurring secondary to a spontaneous brech in the spinal dura mater. While patients with SIH often recover spontaneously or with autologous epidural blood patch (EBP), up to 1/3 of patients fail to respond to repeated EBP. While surgical therapy can be curative in these cases if the leak can be localized, the cost, morbidity, and difficulty identifying the site of the CSF leak during surgery makes this option less than optimal.

We report a patient with a persistent symptomatic CSF leak who recovered following a percutaneous CT-guided fibrin glue injection at the site of a CSF leak.

Case report. A 44-year-old nurse was evaluated for a 3-month history of chronic daily headache. Three months prior to presentation, the patient developed five discrete episodes of exertional headache over a 10-day period. Each episode resolved completely after a night’s sleep. In between episodes, she was headache-free.
The increased risk of ALS in migrants to and from Guam suggests co-occurrence in patients and families of a parkinsonism-dementia complex (PDC) was observed in the Chamorro population of Guam. Between 1940 and 1965, an unusually high incidence of ALS with orthostatic features, followed by headaches that appeared in the afternoon (second half of the day headache) that gradually evolved into a persistent chronic daily headache. Delayed CT myelography demonstrated a CSF leak at the T3 level. After failure to respond to repeated EBP, CT-guided percutaneous injection with fibrin glue provided prompt and sustained relief of headache and associated symptoms.

Fibrin glue (also known as fibrin sealant) is a preparation that mimics the final stages of blood coagulation by forming a stable physiologic fibrin clot that can assist both hemostasis and wound healing. CT-guided injection of fibrin glue at the site of a spinal CSF leak may obviate the need for surgery. Complete and long-lasting benefit has now been reported in five of seven treated cases, including this case. With more experience and appropriate patient selection, this technique may be an important alternative to surgical therapy for patients with intractable SIH secondary to persistent CSF leaks.

**A link between ALS and short residence on Guam**

Danielle Majoor-Krakauer, MD; Paul G.H. Mulder, PhD; Lewis P. Rowland, MD; and Ruth Ottman, PhD

Between 1940 and 1965, an unusually high incidence of ALS with co-occurrence in patients and families of a parkinsonism-dementia complex (PDC) was observed in the Chamorro population of Guam. So far, no genetic causes for ALS/PDC have been found. The increased risk of ALS in migrants to and from Guam suggests a combined effect of an environmental exposure and a genetic susceptibility. The declining incidence of ALS on Guam has been attributed to changes in the consuming patterns of food containing the seed of the locally growing cycad palms. These seeds were either processed traditionally in cycad flour or ingested indirectly by the consumption of indigenous flying foxes. These foxes were foraging on the cycad seeds and are supposed to contain high concentrations of the neurotoxic components of the seeds.

We reviewed information on residence on Guam in a previously conducted case-control study of ALS and investigated the possibility that residence in Guam contributed to the risk for ALS in citizens of the United States.
Methods. The study population consisted of 140 pairs of ALS patients and neurologic control subjects, matched for age (±5 years), gender (83 pairs of men and 57 pairs of women), and insurance status. Participants were patients newly diagnosed with ALS between 1989 and 1991 at the Neurological Institute, Columbia University Medical Center, in New York. The family histories of ALS, dementia, and parkinsonism and occupational and medical histories were ascertained in a semistructured interview. Information on residence outside the continental United States for a period of at least 1 month (place, date, and duration of the stay) was recorded, without alluding to specific places or continents. Information on military service was obtained, including the date of conscription, a job description, the whereabouts, and serious health problems during military service. To investigate the association between Guam and ALS, a matched-pairs exact conditional logistic regression analysis was performed, each case-control pair forming a cluster. Multivariable analysis was performed to control for confounding.

Results and discussion. The results show a significant association between short residence on Guam and ALS (odds ratio [OR] 8.0, 95% CI 1.07 to 355) (table). This association remained in multivariable analysis with age, gender, education, family history of ALS, dementia, or parkinsonism, enrollment and duration of military service, exposure to pesticides, and trauma (OR 9.40, 95% CI 1.08 to 81.91, p = 0.04). The mean age at diagnosis of patients who were on Guam was not different (66 years, range 47 to 81 years) from that of patients who had not been on Guam (60 years, range 21 to 86 years) (p = 0.6). ALS was not associated with residence in Central or South America, the Philippines, New Guinea, or other areas in the South Pacific. Neither was ALS associated with military service. There was no difference in year of enrollment (OR 1.0, 95% CI 0.87 to 1.27) or duration of military service (OR 1.0, 95% CI 0.93 to 1.23). None of the ALS patients was hospitalized or seriously injured or had a blood transfusion on Guam. Of the nine patients, six were on Guam during their military service. One ALS patient lived 1 month, seven lived 2 months, and one lived 15 months on Guam. The mean time between the end of the stay on Guam and the diagnosis of ALS was 43 years (range 27 to 57 years).

Before we can accept the association of short residence on Guam with ALS, methodologic issues have to be addressed. The goal of the original study did not specify Guam as a putative risk factor. Therefore, selection bias related to residence on Guam seems unlikely. For the same reason, we did not collect further information, for example, on eating habits during the stay on Guam. It would have been intriguing to know if patients had traditional cuisine containing cycad seeds. Because the Neurological Institute was not known for an interest in the association between ALS and Guam, it is unlikely that preferential referral of patients who stayed on Guam biased the outcome of this study. However, given our study design, we cannot fully rule out selection or information bias.

Despite the small sample size of this study, we detected an association, whereas two larger studies did not. The time from exposure to ascertainment in the two earlier studies was shorter (16 to 25 and 15 to 27 years) than in the current study (43 years, range 27 to 57 years). This could indicate that one of the key characteristics of the exposure occurring during the period of endemic ALS on Guam is the long delay in clinical expression. In conclusion, our epidemiologic data support the vision that exposure to slow-acting toxic agents is important in the pathogenesis of ALS, most likely in combination with a genetic predisposition.

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Effect of stress related to the 9/11/2001 terror attack on seizures in patients with epilepsy

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