TEXAS NEUROLOGICAL SOCIETY SUMMER 2016

Broca's Area

The Voice of Texas Neurology

President's Message

Deborah Carver Hodges

Dear Colleagues

I first want to say how honored I am to be your president of the Texas Neurological Society (TNS). The TNS continues to be a model for other state neurological societies to emulate. This past September, I participated in the annual meeting for all the state run neurological societies sponsored by the American Academy of Neurology. It is amazing to me that so many states are still just starting out, or have not had the success that we have taken for granted with TNS. Here are just a few of the successes that we have had over the recent years.

TNS continues to evolve and find ways to best help its members. In 2014, the TNS board decided with all the changes in the practice of neurology, including the politics and the business of medicine, the TNS also needed to change and expand. To keep members better informed of all the political changes and to have some one who would fight for us at the State Legislative level, TNS hired a lobbyist, Greg Herzog. As a lobbyist for the state legislature, he serves to advocate for the TNS and all texas neurologists. One of the current issues is the balance-billing proposal that is being heavily debated both inside and out the house of medicine.

In 2014 Kristi Berrier was also hired as our medical economic advisor. She along with Dr. Stuart Black, have been a great source of information for members on all recent changes in the business of neurology including ICD-10, ACA and PQRS. Most recently, they posted a new article on the TNS website called the Hassel Factor which has important information on what we, as physicians, and our patients can do to help with unresolved insurance claims.

The education programs have always been a main focus for TNS. Every year the TNS puts together two very high quality educational programs for members at very low costs. This past winter conference was no exception. Dr. Gary Clark, for the pediatric portion, and Dr. Reeta Achari put together a remarkable list of talks by excellent speakers. This coming summer conference headed by Dr. Mary Ellen Vanderlick looks to continue this trend and includes another pre-conference half-day section of pediatric neurology talks and post conference talks on practice management.

The Texas Neurological Society remains one of the largest state neurological societies. What originally started back in 1974 with around 30 members has grown to 700. TNS, however, would love to have more members participate. Currently, we have three committees: education, medical economics and legislative advocacy. If you have any interest in joining one or all of them, please let us know.

In the meantime, I hope to see you at the summer conference at the La Cantera Hill Country Hotel in San Antonio July 15th and 16th.

Sincerely,

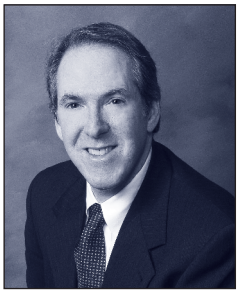
Deborah Carver MD

Mark Your Calendar

2016 SUMMER CONFERENCE

July 15-16

La Cantera
Hill Country Hotel
San Antonio, Texas



Editor's Notes

Randolph W. Evans, MD

This issue

I thank our officers and other contributors for their excellent submissions to this issue. We look forward to seeing you at the TNS 13th Annual Summer Conference in San Antonio, July 15-16. Mary Ellen Vanderlick, program director, Bob Fayle, education committee chair, and the education committee have planned an excellent program.

Many days float by as we see one routine disorder after another as well as the many medically unexplained ones. And then some days the rare and unusual cases.

CASE ONE. A Dirty Dog

This is a 53 year old female who reported that for the prior 6 months, she had been smelling a dirty dog smell typically 2-3 times a day although she could go 2-3 days without the smell with a duration of 30 seconds to 1 hour with an average of 5". She had headaches associated with the smells lasting 1 hour or days with frequent shorter episodes about 5 times before a headache immediately followed by a headache described as a bifrontal throbbing with an intensity of 6/10 associated with nausea, light and noise sensitivity but no vomiting, aura. She took ibuprofen with relief in 1 hour. During the episodes, there was no alteration of consciousness. She had no triggers. She denied depression, anxiety, or increased stress.

Prior to 6 months before, she had occasional headaches since her 20s described as a bitemporal aching with an intensity of 7/10 associated with light and noise sensitivity but no nausea or aura. She would take acetaminophen with relief in 1-2 hours. Stress was a trigger.

She saw a cardiologist about one month after onset and had a CT of the brain without contrast 1/19/15 which was normal. She saw two ENT physicians who found normal exams.
She denied depression, anxiety, or increased stress.

Past medical history of hyperlipidemia on pravastatin. Family history: sister has migraine. Neurological examination was normal.

MRI of the brain with and without contrast was normal. A complete blood count, chemistry profile, thyroid functions, erythrocyte sedimentation rate, antinuclear antibody, rheumatoid arthritis factor, and Sjogren's antibodies were negative. A routine EEG was normal during which she had no episode was normal. She declined a prolonged EEG.

During the next 10 months, she had the dirty dog smell every 2 weeks without a headache lasting about 15-20 minutes.

Question
What is the diagnosis?

Discussion
The olfactory hallucinations (phantosmias) are consistent with migraine auras. Coleman et al (Coleman ER, Grosberg BM, Robbins MS. Olfactory hallucinations in primary headache disorders: case series and literature review. Cephalalgia. 2011;31:1477-89) reported 14 adult cases seen in a New York headache clinic in a 30 month period (prevalence of .66%) and 25 cases from the literature. The typical hallucination lasted 5-60 minutes, occurred shortly before or with the onset of head pain and was of a highly specific and unpleasant odor, most commonly a burning smell. Phantosmias occurred before the onset of headaches in 64.1% and the rest during the headaches. In the majority of cases, phantosmias diminished or disappeared with initiation of migraine preventive treatment.

In a retrospective series of 11 adults from Venice, Italy (Mainardi F, Rapoport A, Zanchin G, Maggioni F. Scent of aura? Clinical features of olfactory hallucinations during a migraine attack (OHM). Cephalalgia. 2016 Mar 31 [Epub ahead of print], the duration of the olfactory hallucinations ranged from 3 minutes to 24 hours. Testing including ambulatory EEG was normal. The hallucinations occurred before the headache in all and also could start during the headaches in 3 patients. Two patients also had episodes of hallucinations without headache similar to the case presented. Migraine preventive medications were typically effective.

The patient in this case declined a trial of a migraine preventive medication.

In a pediatric headache clinic in Essex, UK over a 3 year period, olfactory hallucinations were reported exclusively during headache attacks by 2.5% all of whom had migraine (71% with other auras; in 80%, olfactory aura occurred with other aura symptoms) and occurred in 3.9% of the migraine population (Ahmed MA, Donaldson S, Akor F, Cahill D, Akilani R. Olfactory hallucination in childhood primary headaches: case series. Cephalalgia. 2015;35(3):234-9). Olfactory hallucinations shortly followed the onset of headaches and lasted from 15 to 50 minutes.

It is quite unlikely that the episodes in this case are due to partial seizures because of the long duration. Olfactory auras have a prevalence rate between 0.6% and 16% in temporal lobe epilepsy. In a series of 13 patients with olfactory epileptic auras (age of onset 7-45 years, mean 20.5), 7 described an unpleasant smell, five neutral, and 2 pleasant smells (Acharya V, Acharya J, Lüders H. Olfactory epileptic auras. Neurology. 1998;51(1):56-61). Auras lasted for 5-30 seconds. MRI of the brain showed tumors in 10 which was restricted to the temporal lobe in 9. Interictal EEG showed mesial temporal spikes in all patients. In other series, patients report mostly unpleasant smells (Chen C, Shih YH, Yen DJ, et al. Olfactory auras in patients with temporal lobe epilepsy. Epilepsia. 2003;44(2):257-60).

Unpleasant odors localize to the olfactory bulb and medial temporal (especially the uncus) and pleasant odors to the insula (Perven G, So NK. Epileptic auras: phenomenology and neurophysiology. Epileptic Disord. 2015;17(4):349-62).

CASE TWO. NEW DAILY PERSISTENT HEADACHE AND BEHÇET'S

This is a 19 year old Hispanic female who developed a severe headache with an intensity of 10/10 with light and noise sensitivity almost 2 years prior to the headache consultation which had been daily since. She described a bitemporal and the back of the head pressure, right more than left sided, with an intensity of 5-8/10 associated with nausea, vomiting occasionally, light and noise sensitivity but no aura. Light and noise increased the pain. Tramadol which she was taking twice a week did not help.

One week prior to the onset of the headache, she had acute abdominal pain with a negative evaluation. A few months after the onset of the headaches, she developed polyarthralgias, ulcers of the mouth and vagina, and decreased vision in the right eye due to uveitis (with recovery over several months). She saw a rheumatologist who diagnosed Behçet's and started Imuran about 1.5 years prior to the consult and Humira a few months later with improvement. She denied depression. Past medical history was otherwise negative. Neurological exam was normal There was bilateral greater occipital nerve tenderness.

Bilateral greater occipital nerve blocks were performed by injection of 3 cc each of 1% lidocaine. Bilateral supraorbital, supratrochlear, and auriculotemporal nerve blocks were performed by injection of .5 cc each of 1% lidocaine. She was prescribed amitriptyline which she declined to start.

MRI of the brain and MRV of the brain were negative.

The headaches resolved for one month following the blocks and then recurred about 3 days per week with an intensity of 6/10 relieved in 2-3 hours with ibuprofen. The same blocks were performed again and she was started on topiramate titrated to 100 mg daily. On follow-up 3 months later, the headaches had resolved for one month and then recurred 1-2 days per week for 1 month and then daily again for 1 month with an intensity of 7/10. The same blocks were performed, topiramate was stopped, and she was started on venlafaxine xr 37.5 mg/d to be titrated up as tolerated.

Two months later, she reported the headaches had resolved for 1.5 months and then had increased to daily for the prior week. She could not increase the venlafaxine dose due to nausea and declined switching to amitriptyline. She was placed on baclofen 10 mg ½-1 tid prn. The blocks were performed. Three months later, she reported the headaches had resolved for 1 month and then recurred twice a week with an intensity of 6/10 lasting 2-4 hours with medication. She had stopped venlafaxine and baclofen 6 weeks prior due to hives. The same blocks were performed and she was placed on methocarbamol prn.

Questions

What is the cause of the headaches? Is there any evidence for the nerve blocks being effective?

Discussion

Behçet's syndrome is names after Hulusi Behçet (1889-1948), a professor of dermatology in Istanbul, who reported 3 patients with orogenital ulcerations and eye inflammation, in 1937. Behçet's syndrome is rare in the United States with a prevalence of .38/100,000 population in Omsted County, Minnesota (Calamia KT, Wilson FC, Icen M, et al. Epidemiology and clinical characteristics of Behçet's disease in the US: a population-based study. Arthritis Rheum 2009; 61:600) but is more common from eastern Asia to the Mediterranean with the highest prevalence in Turkey of 80-370/100,000. Young adults ages 20-40 are typically affected.

This patient had typical symptoms. Most patients initially have recurrent oral apthous ulcerations and 75% or more have genital ulcerations (of the scrotum or vulva) which are similar to oral aphthae (Smith EL, Yazici Y. Clinical manifestations and diagnosis of Behçet's syndrome. UpToDate. 2016). Ocular disease occurs in 25-75% often uveitis. Fifty percent have a nondeforming arthritis of medium and large joints. Abdominal pain can be also be a manifestation due to ulcerations in some cases.

Neurological disease occurs in 5-10% of patients in most series more in men than women (Saip S, Akman-Demir G, Siva A. Neuro-Behçet syndrome. Handb Clin Neurol. 2014;121:1703-23). Parenchymal disease includes the following: multifocal (diffuse) disease (including brainstem, cerebral, or spinal cord disease), myelopathy, cerebral disease (including encephalopathy, hemiparesis, hemisensory loss, seizures, dysphagia, and mental changes such as psychosis and cognitive dysfunction), and optic neuropathy. Most cases have vascular-inflammatory CNS disease with focal or multi-focal parenchymal involvement mostly presenting with a subacute brainstem syndrome and hemiparesis.

Non-parenchymal disease includes the following: cerebral venous thrombosis (CVT), intracranial hypertension syndrome (pseudotumor cerebri), acute meningeal syndrome, and uncommonly stroke due to arterial thrombosis, dissection, or aneurysm. Isolated CVT and intracranial hypertension account for about 10-20% of neurological cases.

Headache is the most common neurological symptom in Behçet's occurring in 56-83% of cases. Headache can be due to parenchymal disease, vascular disease (cerebral venous thrombosis), increased intracranial pressure without sinus venous thrombosis, meningitis, associated with ocular inflammation, and isolated headache not accompanied by neuro- Behçet's or uveitis (Evans RW, Akman-Demir G. Behçet syndrome and headache. Headache. 2004;44(1):102-4; Kale N, Agaoglu J, Icen M, Yazici I, Tanik O. The presentation of headache in neuro-Behet's disease: a case-series. Headache. 2009 Mar;49(3 Behçet's):467-70; Vishwanath V, Wong E, Crystal SC, et al. Headache in Behçet's syndrome: review of literature and NYU Behçet's syndrome center experience. Curr Pain Headache Rep. 2014;18(9):445).

Continued on page 5

Unusual Presentation of Chrionic Sensory Neuropathy


Kavitha Tirumalasetti, MD

Kavitha Tirumalasetti, MD
University of Texas at Austin Dell Medical School

Sara Austin, MD
Seton Brain and Spine Institute

With special thanks to
Bridgett Wallace, PT, DPT,
360 Balance and Hearing, Austin, TX

Case Presentation

A 71 year old female presented with a 15 year history of lower extremity numbness. Her symptoms began in 2000 with numbness in her feet, and she was found to have a positive Romberg sign. Her EMG was reportedly abnormal, and she was diagnosed with a sensory peripheral neuropathy. She began having poor balance and frequent falls without vertigo. She was reevaluated with lower extremity EMG in 2008 and found to have absent SNAPs, borderline low CMAPs, and normal EMG. Screening labs, genetics screening with a complete ataxia panel and CT head/neck imaging were unremarkable. She developed intermittent diplopia in 2010. She had vestibular testing in 2012 and was diagnosed with Meniere's disease. She revealed her older brother had a peripheral neuropathy and similar milder symptoms.

Physical examination remarkable for multi-directional nystagmus; full strength of all extremities; decreased pinprick and vibratory sensation; diminished upper extremity reflexes and absent lower extremity reflexes; lower extremity dysmetria; truncal ataxia; wide-based, ataxic gait with unsteady station, and she ambulated with a walker.

Her previous workup included normal laboratory studies such as TSH, RPR, vitamin B12, folate, vitamin E, ferritin, ESR, HbA1C, rheumatoid factor, copper, ceruloplasmin, and SPEP. MRI brain in 2013 showed incidental 1 cm vertex meningioma, otherwise unremarkable. Recent upper extremity EMG in 2015 again showed absent SNAPs, borderline low CMAPs, and she had mild distal chronic denervation changes. She was referred again for vestibular evaluation, which confirmed complete loss of vestibulo-ocular reflex on the left with significant weakness on the right, and corrective saccades and nystagmus on smooth pursuit. These constellation of symptoms and characteristic vestibulo-ocular findings supports the diagnosis of this distinct syndrome.

Introduction

The association between cerebellar ataxia and bilateral vestibulopathy (CABV) was first described in the 1990s (1). In 2004, the visually-enhanced vestibulo-ocular reflex (VVOR) was recognized as a characteristic sign associated with this syndrome (1,2). Impaired VVOR reflects a failure of 3 compensatory eye movements: vestibulo-ocular reflex, smooth pursuit, and the optokinetic reflex. Sensory peripheral neuropathy was identified in these patients as well, but not thought to be a feature until 2011, and the syndrome was renamed as cerebellar ataxia neuropathy vestibular ataxia syndrome (CANVAS) (3,4).

- Once thought to be sporadic, affected sibling pairs have been identified, suggesting an autosomal recessive inheritance pattern (late-onset) (4).
- Neuropathy is predominately length-dependent, with small and large fiber sensory involvement; can also see mild motor axonal loss (3).
- Downbeating nystagmus is a common finding (6).
- Some degree of cerebellar atrophy may be present on brain MRI (though there are a few patients described with normal MRIs). Additionally, MRI may be normal in earlier stages of the disease (3,6).
- An otopathologic case report revealed the patient had severe atrophy of bilateral vestibular nerves, with significant loss of Scarpa's ganglion cells (84%) when compared to the normal cell count for age (5).
- In a recent study by T.Y. Wu et al, 83% of patients had evidence of autonomic dysfunction; most common symptoms are hypohidrosis and orthostatic hypotension (6,7).
- Spinocerebellar ataxias (particularly SCA 3), Friedreich's ataxia, multiple system atrophy of cerebellar type, and Wernicke's encephalopathy share some similar features and are important conditions to differentiate from (3,6).
- Management is symptomatic and preventative including risk assessment for falls, vestibular rehabilitation, and neuropathic pain management (pregabalin most effective) (7).
- CANVAS is a progressive condition, though with variability in mobility decline; most patients described require assistance with a gait aid or wheelchair (3).

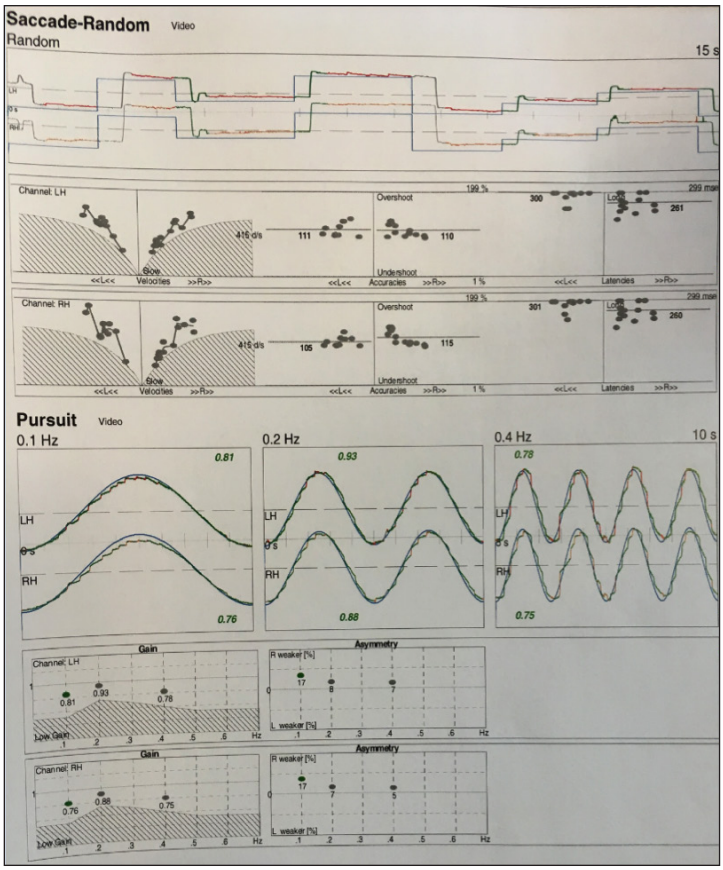
Discussion

We present a patient with a longstanding history of sensory neuropathy, which was the only finding for several years. In retrospect, her early symptoms of poor balance and a positive Romberg sign suggests there was impairment of the vestibular and/or proprioceptive functions during initial presentation. She developed nystagmus and diplopia years later, and interestingly it wasn't until her second vestibular evaluation that impairment in VVOR was found.

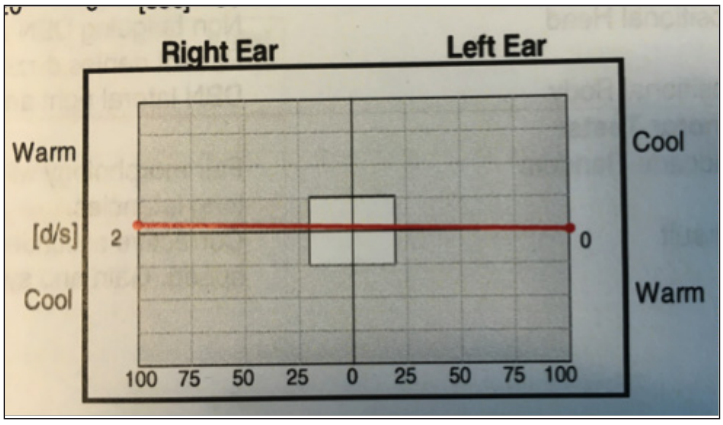
Screening for similar syndromes was unremarkable, including genetic testing for SCAs and FRDA. Her clinical features and neuroimaging did not fit the pattern for

multiple system atrophy, Wernicke's encephalopathy, or another neurodegenerative process. Of note, her MRI brain did not show cerebellar atrophy, which has been described in a few CANVAS patients in the literature.

Vestibular evaluation in our patient



Oculomotor testing. (Top) increased latencies and corrective saccades strongly suggest cerebellar involvement (saccades yield a high level of specificity). (Bottom) corrective saccades that worsens with increased speed, suggestive of central involvement (pursuit yields a high level of sensitivity).



Oculocephalics testing. Images show bilateral weakness in calorics with warm air stimulation – with zero response on the left.

CONCLUSIONS

CANVAS should be in the differential for a sensory ataxic neuropathy. The vestibular features may develop years after the neuropathy becomes apparent. In patients with 'sensory ataxia', particularly in whom the ataxia is marked, one might consider getting vestibular testing.

REFERENCES:

1. Bronstein, Adolfo M., Mossman, Stuart., Luxon, Linda M. The neck-eye reflex in patients with reduced vestibular and optokinetic function. Brain 1991; 1-11.
2. Migliaccio, Americo A., Halmagyi, G. M., McGarvie, Leigh A., Cremer, Phillip D. Cerebellar ataxia with bilateral vestibulopathy: description of a syndrome and its characteristic clinical sign. Brain 2004; 127: 280-293.
3. Szmulewicz, David J. et al. Sensory neuropathy as part of the cerebellar ataxia neuropathy vestibular areflexia syndrome. Neurology 2011; 76: 1903-1910.
4. Szmulewicz, David J. et al. Cerebellar ataxia, neuropathy, vestibular areflexia syndrome (CANVAS): a review of the clinical features and video-oculographic diagnosis. Annals of the New York Academy of Sciences 2011.
5. Szmulewicz, David J., Merchant, Saumil N., Halmagyi, G Michael. Cerebellar ataxia with neuropathy and bilateral vestibular areflexia syndrome (CANVAS): a histopathologic case report. Otol Neurotol 2011; October, 32(8): e63-e65.
6. Wu, Teddy Y., Taylor, Jennifer M., Kilfoyle, Dean H., et al. Autonomic dysfunction is a major feature of cerebellar ataxia, neuropathy, vestibular areflexia 'CANVAS' syndrome. Brain 2014; 1-8.
7. Szmulewicz, David J., McLean, Catriona A., MacDougall, Hamish G. et al. CANVAS an update: Clinical presentation, investigation and management. Journal of Vestibular Research 2014; 24: 465-474.
8. Peterson, Jens Alexander, Wichmann, Werner W., Weber, Konrad Peter. The pivitol sign of CANVAS. Neurology 2013; 81: 1642-1643.

Continued from page 3

Our patient had headaches consistent with the isolated type fitting new daily persistent headache (NDPH) with migraine features. When isolated headache occurs, different studies have variably found predominant migraine or tension type. In one prospective study of 44 patients with Behçet's, headache was reported in the past year by 38 (86%), 24 with episodic headache and 14 with chronic daily with 3 of them meeting criteria for NDPH (Vishwanath V, Wong E, Crystal SC, et al. Headache in Behçet's syndrome: review of literature and NYU Behçet's syndrome center experience. Curr Pain Headache Rep. 2014;18(9):445).

There are no prospective placebo controlled trials of preventive treatment for NDPH (Evans RW. New daily persistent headache. Headache. 2012;52 Suppl 1:40-44)). The treatment for NDPH with migraine features and chronic migraine are the same although NDPH is less responsive to treatment.

Topiramate was not effective. She developed hives while on venlafaxine and baclofen.

I empirically gave her bilateral greater occipital nerve and trigeminal nerve blocks which have been very effective although the evidence for efficacy is largely anecdotal or open label (Blumenfeld A, Ashkenazi A, Evans RW. Occipital and trigeminal nerve blocks for migraine. Headache. 2015;55(5):682-9). The blocks are often effective without corticosteroids (supraorbital and supratrochlear blocks should not be performed with corticosteroids because of the risk of lipodystrophy).

Tics and Tourette Syndrome

Joseph Jankovic, MD

Professor of Neurology, Distinguished Chair in Movement Disorders, Director, Parkinson's Disease Center and Movement Disorders Clinic, Director, Tourette Association of America Center of Excellence, Department of Neurology, Baylor College of Medicine, Houston, Texas

Joseph Jankovic, MD

Tourette syndrome (TS) is a childhood-onset neurological disorder characterized by multiple motor and phonic tics and a wide range of behavioral problems, including attention deficit with hyperactivity (ADHD) and obsessive-compulsive disorder (OCD) (Thenganatt and Jankovic, 2016). The Diagnostic and Statistical Manual of Mental Disorders Fifth Edition defines TS by the presence of multiple motor and phonic tics with onset prior to age 18 and lasting at least 1 year. Epidemiological studies have shown that 20-30% of children exhibit tics sometime during their childhood and 2-3% of children develop TS. TS symptoms are typically at their worst just prior to puberty and spontaneously improve by the age of 20 years, but in about a third of the patients the symptoms persist through adulthood.

There is no diagnostic test for TS and, therefore, recognition of the full spectrum of phenomenology of tics and various behavioral co-morbidities is critical to the diagnosis. Tics, the clinical hallmark of TS, are relatively brief and intermittent movements (motor tics) or sounds (vocal or phonic tics). This division into motor and vocal/phonic tics, however, is artificial, because vocal/phonic tics are actually motor tics that involve respiratory, laryngeal, pharyngeal, oral, and nasal musculature. Motor tics typically consist of sudden, abrupt, transient, often repetitive and coordinated (stereotypical) movements that may resemble gestures and mimic fragments of normal behavior, vary in intensity, and are repeated at irregular intervals. Tics are usually intermittent and may be repetitive and stereotypic. They may occur as brief bouts or bursts and they tend to wax and wane with unpredictable periodicity. In addition to variable frequency and intensity, tics often change anatomic location. Typically, tics can be volitionally suppressed, although this might require intense mental effort and they may also increase when suppression is no longer needed, such as when returning home from school. In contrast to other hyperkinetic movement disorders that are usually suppressed during sleep, motor and phonic tics tend to persist during all stages of sleep.

Tics may be simple or complex. Simple motor tics involve only one group of muscles, causing a brief, jerk-like movement. They are usually rapid and brief (clonic tics), but they may be slower, causing a briefly sustained abnormal posture (dystonic tics) or an isometric contraction (tonic tics). Examples of simple clonic motor tics include blinking, nose twitching, and head jerking. Motor and phonic tics are preceded by premonitory sensations in over 80% of patients. This premonitory phenomenon consists of sensations or discomforts localized to the anatomic region of the tic, such as a burning feeling in the eye before an eye blink, tension or a crick in the neck that is relieved by stretching of the neck or jerking of the head, a feeling of tightness or constriction that is relieved by arm or leg extension, nasal stuffiness before a sniff, a dry or sore throat before throat clearing or grunting, and itching before a rotatory movement of the scapula. Besides these local or regional sensations, premonitory phenomenon may be more generalized, poorly described feeling, such as an urge, anxiety, anger, and other psychic sensations. The pathophysiology of premonitory sensations is not well understood but they represent an example of increasingly recognized sensory aspects of movement disorders (Patel et al., 2014). Simple dystonic tics include blepharospasm, oculogyric movements, bruxism, sustained mouth opening, torticollis, and shoulder rotation. Tensing of chest, abdominal or limb muscles is an example of a tonic tic. Blocking tics are due to either prolonged tonic or dystonic tics that interrupt ongoing motor activity such as speech (intrusions) or a sudden inhibition of ongoing motor activity (negative tic). Complex motor tics consist of coordinated, sequenced movements resembling normal motor acts or gestures that are inappropriately intense and timed. Additional examples of complex motor tics include gesturing "the finger" and grabbing or exposing one's genitalia (copropraxia) or imitating gestures (echopraxia). Some complex motor tics may be difficult to differentiate from compulsions, which frequently accompany tics, particularly in TS. Simple phonic tics typically consist of sniffing, throat clearing, grunting, squeaking, screaming, coughing, blowing, and sucking sounds. Complex phonic tics include linguistically meaningful utterances and verbalizations, such as shouting of obscenities or profanities (coprolalia), repetition of someone else's words or phrases (echolalia), and repetition of one's own utterances, particularly the last syllable, word or phrase in a sentence (palilalia).

Several different scales are used to assess various aspects of TS. The most widely used instrument to "measure" tics is the Yale Global Tic Severity Scale (YGTSS) which consists of two broad domains: Total Tic Severity (with 2 sub-domains: Motor and Phonic Tics) and Impairment. Within each category there are five dimensions, score 0 - 5: number of tics, frequency, intensity, complexity, and interference. The total tic score ranges from 0 to 50; the usual ranges in most studies is 15 to 30. A health-related quality of life (HR-QOL) has been developed and validated for internal consistency, test-retest reliability and and against other clinical scales.

Pathogenesis of TS is still not well understood but various clinical, genetic, physiological, and imaging studies suggest that TS represents a developmental genetic disorder resulting in abnormal function in the corticostriatal-thalamic-cortical circuit which then leads to motor and behavioral disinhibition (Black et al, 2014). Despite numerous attempts, the gene or genes responsible for TS have not yet been identified. One of the reasons for this is that, as we demonstrated several years ago, TS has bileneal transmission, meaning that both parents are usually affected to some degree.

Many patients with TS do not require any form of therapy beyond education and counseling about the disease. Although behavioral therapy, such as Comprehensive Behavioral Intervention for Tic Disorders (CBIT) and habit reversal therapy (HRT), is increasingly advocated in the treatment of TS, there is marked paucity of

well-trained therapists who are certified in these behavioral techniques. Thus, behavioral therapy is often difficult to access, maintain, and afford.

By the time children or adults with TS are referred to our Movement Disorders Clinic their symptoms are usually quite troublesome, requiring some form of pharmacological intervention (Gilbert and Jankovic, 2014). If the patient has only focal tics, such as frequent and persistent blinking which may lead to sustained eye closure (blepharospasm), forceful neck movements ("whiplash tics"), or loud vocalizations including shouting of obscenities (coprolalia), an injection of botulinnum toxin (BoNT) into the affected muscles may be the best therapeutic option, providing effective relief of the tics for at least 3-4 months. We and others showed that BoNT not only improves the motor component of the tic but also the premonitory urge.

Numerous controlled and open label trials have found that of the various pharmacologic agents used for tic suppression, the dopamine receptor–blocking drugs (neuroleptics) and dopamine depleting drugs (e.g. tetrabenazine) are clearly most effective (Jankovic, 2015; Thenganatt and Jankovic, 2016). Currently, pimozide, haloperidol, and aripiprazole, are the only drugs approved by the Food and Drug Administration (FDA) for the treatment of TS, but we almost never use these drugs in our Movement Disorders Clinic because of a variety of potential side effects, including tardive dyskinesia. Until recently we have used fluphenazine as the first-line anti-tic pharmacotherapy, since it is quite effective in controlling tics and appears to have a relatively low risk of side effects effects (Wijemanne et al., 2014). Since I first obtained my Notice of Claimed Investigational Exemption for a New Drug (IND) for tetrabenazine in 1979, I have used this drug in thousands of patients with various hyperkinetic movement disorders including TS. Tetrabenazine (Xenazine) was approved by the FDA in 2008 for the treatment of chorea associated with Huntington disease, but it is also a powerful anti-tic drug that has the advantage over typical and atypical neuroleptics in that it does not cause tardive dyskinesia (Jankovic J, Clarence-Smith, 2011). As a vesicular monoamine transporter type 2 (VMAT2) inhibitor, tetrabenazine depletes dopamine presynaptically. While generally effective in reducing abnormal involuntary movements including tics, tetrabenazine is associated with some adverse effects, such as somnolence, nausea, depression, insomnia, akathisia, and parkinsonism, that may limit its utility. Furthermore, the active metabolites of tetrabenazine, alpha-dihydrotetrabenazine (α HTBZ) and beta dihydrotetrabenazine (β HTBZ), have short half lives thus requiring tetrabenazine to be administerd at least 3 times per day. To address some of the limitations of tetrabenazine, Auspex, a wholly owned subsidiary of Teva Pharmaceutical Industries (Petach Tikva, Israel) has developed a deuterated form of tetrabenazine (previously referred to as SD-809 or TEV-50717). The deuterium placement forms a stronger bond with carbon that requires more energy for cleavage, thus attenuating metabolism, resulting in longer half-lives and other potential advantages. In a pilot study involving 23 patients with moderately severe TS, deutetrabenazine we showed that the drug was associated with a 37.6% reduction in tic severity ($p<0.0001$); other measures also showed improvement without significant adverse effects (Jankovic et al., 2016). Another VMAT2 inhibitor, NBI-98854 (also known as Valbenazine, developed by Neurocrine Biosciences), administered only once/day is also being investigated in tardive dyskinesia and in TS. A novel D1 dopamine receptor antagonist, Ecopipam (Psyadon Pharmaceuticals), has been reported to be effective in the treatment of TS-related tics in an open label trial and is currently being investigated in children with TS. All three drugs, deutetrabenazine, Valbenazine, and Ecopipam are currently being investigated in the treatment of TS by the movement disorders team at Baylor College of Medicine. Finally, if medications and BoNT fail to obtain satisfactory relief of their symptoms DBS targeting thalamus, globus pallidus or other brain regions may need to be considered (Viswanathan et al., 2012).

In 2015 the Parkinson's Disease Center and Movement Disorders Clinic (PDCMDC) was designated by the Tourette Association of America (TAA) as its first Center of Excellence (COE). We are honored by this prestigious designation which is an affirmation of our committement to excellence in clinical care, research and education related to TS. In addition to the multidisciplinary care (which includes behavioral, medical and surgical therapies), the PDCMDC is currently conducting dozens of clinical trials in various movement disorders, including four investigating novel treatments in TS. For further information about the TAA COE and the PDCMDC's experimental therapeutics research program, training, and other clinical and educational activities visit www.jankovic.org.

SELECTED REFERENCES

1 Black KJ, Jankovic J, Hershey T, McNaught KS, Mink JW, Walkup J. Progress in research on Tourette syndrome. J Obsessive Compuls Relat Disord. 2014;3(4):359-362.

2 Gilbert DL, Jankovic J. Pharmacological treatment of Tourette syndrome. J Obsessive-Compulsive and Related Disorders, 2014;3:407–414.

3 Jankovic J, Clarence-Smith K. Tetrabenazine for the treatment of chorea and other hyperkinetic movement disorders. Expert Review of Neurotherapeutics 2011;11:1509-23.

4 Jankovic J. Therapeutic developments for tics and myoclonus. Mov Disord. 2015;30:1566-73.

5 Jankovic J, Jimenez-Shahed J, Budman C, Coffey B, Murphy T, Shprecher D, Stamler D. A pilot study of deutetrabenazine in tics associated with Tourette syndrome. Presented at the 68th Annual Meeting of the AAN, Vancouver, BC, Canada, 4/16/16.

6 Thenganatt MA, Jankovic J. Recent advances in understanding and managing Tourette syndrome. F1000Res. 2016 Feb 9;5.

7 Patel N, Jankovic J, Hallett M. Sensory aspects of movement disorders. Lancet Neurol. 2014 Jan;13(1):100-12.

8 Viswanathan A, Jimenez-Shahed J, Baizabal Carvallo JF, Jankovic J. Deep brain stimulation for Tourette syndrome: target selection. Stereotact Funct Neurosurg. 2012;90:213-24.

9 Wijemanne S, Wu LJ, Jankovic J. Long-term efficacy and safety of fluphenazine in patients with Tourette syndrome. Mov Disord. 2014;29:126-30.

DR. AZIZ SHAIBANI
(HOUSTON)
FOR THE BOOK ROYALTY DONATION TO THE SOCIETY.

Frequent Falls In The Elderly: Think NPH!

Gustavo C. Román, MD, DrHC
Methodist Neurological Institute
Houston Methodist Hospital
Houston, TX
Professor of Neurology
Weill Cornell Medical College, Cornell University
New York, NY


Gustavo C. Román MD DrHC

Introduction

Frequent falls in the elderly are a leading cause of injury resulting in $35 billion in direct medical costs; by 2030 the number of fatal falls among older adults is projected to reach 100,000 per year with an associated cost of $100 billion[1] as a result of fractures, head injury and other traumatic lesions. Many elderly patients with history of frequent falls fail to be diagnosed with idiopathic Normal-Pressure Hydroce alus (iNph), a potentially treatable condition.[2] Although Nph was described 50 years ago[3,4] the condition known as "chronic senile hydrocephalus" [5] was first reported by Morgagni in 1727, indicating the significant increase in iNph prevalence after 70 years of age.[6-8]

Although Alzheimer's disease (AD) and other neurodegenerative pathologies do occur in elderly patients with confirmed iNph,[9] AD should not be the default diagnosis in an older patient presenting with the typical symptomatic iNph triad of cognitive decline, bladder incontinence, and abnormal gait. In fact, the progression of neurodegeneration in AD does not explain the occurrence of abnormal gait. The classic neuropathological studies of Braak and Braak,[10-13] demonstrated that tau deposits, neurofibrillary tangles, neuronal disconnection and cortical atrophy, typically follow a trans-synaptic progression from early lesions in the nucleus basalis of Meynert,[13] to the hippocampus-parahippocampal cortex, finally affecting prefrontal-frontal regions.[10-12] Even at Braak & Braak stages V-VI of advanced AD (Figure 1) there is minimal involvement of supplementary motor cortical areas that could produce alterations of gait or bladder control.

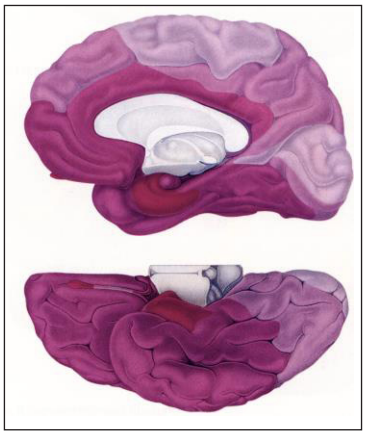


Figure 1. Braak and Braak Stages V-VI of advanced Alzheimer's disease. Notice sparing of cortical supplementary motor areas for control of lower limbs and bladder (Diagram by Leroy Baudouin).

The 4th element for the diagnosis of Nph is the demonstration of ventriculomegaly. This is sometimes the most prominent feature of the clinical syndrome. The radiologists reading the brain images (CT or MRI) consistently note the increase in ventricular size but the explanation provided usually judges that the ventricular enlargement is "consistent with age" or "proportional to the degree of cortical atrophy." This is akin to determining the degree of anemia without the values of hemoglobin or hematocrit. A report telling the clinician that there is "moderate-to-severe anemia" without numerical values cannot be used to determine the need for blood transfusion. Obtaining a numerical value for the ventricular size helps to solve this problem. The ventricular measurement most frequently used is the Evans index,[14] a venerable 74-year-old heirloom from the days of pneumoencephalography that measures the ratio of the widest diameter of the frontal horns to the widest diameter of the brain on the same axial slice (Figure 2). The normal values of the Evans index range from 0.25 to 0.30. With the availability of computer programs that accurately quantify ventricular-CSF volume, cortical and white matter volumes, as well as lobar and total brain volumes, these problems may be relegated soon to the history of medicine. In the interim, neurologists are advised to measure the Evans index in brain MRI/CT from all their patients.

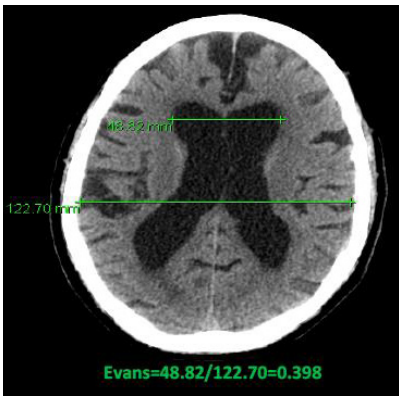


Figure 2. The Evans index measures the ratio of the widest diameter of the frontal horns to the widest diameter of the brain on the same axial slice (normal ≤0.30).

NPH symptoms: The Hakim triad

In 1965, the original description of Nph in The New England Journal of Medicine by Adams, Fisher, Hakim, et al.[3] emphasized the "progressive dementia characterized by forgetfulness, psychomotor retardation and unsteady gait." The same year, a second paper by Hakim and Adams,[4] reviewed the importance of intraventricular hydrostatic pressure on the physiopathology of the syndrome and recorded the so-called Hakim's triad consisting of abnormal gait, incontinence and cognitive decline. (In medical students' parlance, the 3Ws: wobbly, wet and wacky). Although the initial report emphasized the presence of treatable dementia, patients with ventriculomegaly may not have the complete triad. Progressive gait difficulty may be the only manifestation, unaccompanied by concurrent decline in mentation; gait

plus bladder problems without cognitive decline is another form of presentation.[15-17]

Cognitive decline: The cognitive problems observed in Nph include deficits of memory and executive dysfunction predominantly frontal in nature. Hakim and Adams[4] called it "psychomotor retardation" and described "lack of impulsivity, expressed as apathy, disinterest, and lack of spontaneity." Successful ventriculoperitoneal shunting (VPS) results in significant neuropsychological improvement.[18] The Rey Auditory Verbal Learning Test (RAVLT) appears to be highly sensitive to cognitive improvement in Nph; post-shunt improvement is also observed in the Mini-Mental State Examination (MMSE), phonemic verbal fluency tests and the Trail Making test A, independent of age, gender or follow-up interval.[18]

Sphincter incontinence: The incontinence of sphincters also appears to be frontal in origin. According to Hakim and Adams[4] "… the patient being unaware of the contents of bladder and bowel and incapable of making any arrangement for the somewhat precipitate action of these organs." Urinary incontinence is usually discounted as being the result of often-present prostate problems in aged men or an exacerbation of chronic bladder stress incontinence in elderly women. Frontal-type stool incontinence is less common but it also occurs.

Gait: In 1982, C. Miller Fisher[19] provided a careful description of the gait disturbance in 30 elderly patients with Nph. Imbalance was the most frequent complaint, described by patients as "poor balance, off balance, unsteady, wobbly, staggering, and drunken." Gait is uneven and energy consuming and patients have a tendency to fall and difficulty on stairs and curbs. Symptoms fluctuate from day to day and from one week to the next. Patients often have difficulty initiating gait movements and the feet are "glued" to the floor or "magnetized." This may be due to the presence of plantar grasp reflex and tonic feet pressor response. Very slow walking speed, short steps, and ataxia in the vertical direction have been demonstrated by computer-assisted gait analyses.[20] Feet are placed on the ground with variable force. With eyes closed the ataxia, postural instability and unsteadiness of gait increase markedly. As the disorder progresses the steps become shorter, shuffling and scuffing occur; turning becomes precarious and is usually done by pivoting in one leg; i.e., the so-called "compass sign." At this point, patients usually require a cane or a walker to ambulate and are at very high risk of hip fractures.

Patients with occult Nph complain of weakness and tiredness after walking short distances. In advanced stages, the patient is unable to regain balance and becomes wheelchair-bound and totally incapacitated; standing, sitting, and finally turning over in bed become impossible.[19] According to Hakim and Adams[4] "With the latter the plantar reflexes were usually extensor and the tendon reflexes brisk, particularly in the legs. An element of ataxia seemed to have been present but was always difficult to evaluate, and clear-cut intention tremor and dysmetria were not demonstrable." Thus, plantar reflexes can be either flexor or extensor bilaterally, or extensor on one side and flexor on the other.[19] Sensory complaints are common. Patients indicate that the legs feel funny, numb, weak, stiff, hot or cold; and the complaints increase with exercise.

Frequent falls: Recurrent falls are the end result of the above gait alterations with loss of righting reflex. Falls are unprovoked, unaccompanied by dizziness, faintness or loss of consciousness. Patients often blame the fall on minimal irregularities of the floor surface or difficulty negotiating stairs. The incapacity to break the fall is compared by patients to having the feet shackled. Patients may give a history of falling forward, backwards or to the sides. It is not uncommon to discover the presence of hydrocephalus when the patients undergo brain imaging in the course of an emergency room evaluation for head injury resulting from a severe fall. In our experience, subdural hematomas are often present alongside ventriculomegaly. For similar reasons, ventriculomegaly may also be found in elderly patients undergoing surgical treatment for hip fracture.

Differential Diagnosis.

The main conditions included in the differential diagnosis of Nph include Parkinson's disease, Binswanger's disease,[21,22] subcortical ischemic white matter lesions and lacunar dementia,[23-25] progressive supranuclear palsy,[26] and multisystem degeneration. C. Miller Fisher observed that in severely impaired patients unable to walk the legs function well when the patient is on the back; this led him to suggest that the gait disorder is a frontal gait apraxia or hydrocephalic astasia-abasia.[19] In rare patients, attempts to walk produce great anguish (French: astasia-abasia trépidante or vibrating astasia-abasia). The abnormal gait can be parkinsonian in nature, in particular "lower-body parkinsonism" or arteriosclerotic parkinsonism.[27] Nph may be particularly difficult to diagnose in elderly patients with severe rheumatoid arthritis or osteoarthritis.[28]

The alterations of gait in iNph are essentially identical to those observed in patients with extensive ischemic lesions of the periventricular white matter in Binswanger disease,[22] as well as those noted in patients with multiple lacunar strokes (i.e., "lacunar dementia").[25] The gait of the latter patients was called by Dejerine "marche à petits pas" or short-step gait. The simultaneous occurrence of Nph and these vascular conditions is relatively common and the potential beneficial effect of VPS should not be denied to vascular patients a priori without the large-volume spinal tap test.

NPH evaluation

There are clinical and imaging criteria for the diagnosis of Nph from a European-USA meeting of experts,[15] and from a similar Japanese group,[16] but no universally accepted method predicts accurately which patients would respond to surgical treatment with VPS. Most recently, the American Academy of Neurology published guidelines regarding response to shunting and predictors of response.[2] The single test with the highest predictive value is CSF drainage, either by large-volume lumbar puncture (LVLP), or by constant drainage of CSF via lumbar drain.

At Houston Methodist Hospital the evaluation of patients suspected of iNph is performed, using the protocol summarized

Continued on page 10

Continued from page 9

in Table 1, by a team that includes specialists in neurology, neuropsychology, physical therapy, neuroradiology and neurosurgery.

NEUROPSYCHOLOGY

Prior to the LVLP, a certified clinical neuropsychologist evaluates the following cognitive domains: global cognition (MMSE), memory, orientation, language, executive function, and praxis. Twenty-fours after the LVLP the same person performs the second evaluation, modified to avoid learning and practice effects.

PHYSICAL THERAPY

On the day of admission for the LVLP, a pre-trained certified Physical Therapist tests and scores the patients' gait and balance within a few hours prior to and after the LVLP using Tinetti's test[29] and Berg's Balance (BB) test.[30]

SPHINCTER CONTROL

For a period of in-hospital observation (24 hours), pre- and post-LVLP, the patients' accompanying relative is instructed to notify the nurse if the subject requests to void or to evacuate, or if incontinence occurred.

LARGE-VOLUME LUMBAR PUNCTURE

A routine lumbar puncture is performed under fluoroscopy; ideally, a total of 50 mL of CSF is collected; opening and closing pressures are obtained.

Surgical treatment with implantation of a ventriculoparitoneal shunt (VPS) is usually recommended only in patients that present clear improvement in gait post LVLP, with or without improvement in bladder control. Only a minimal number of patients show improvement in cognitive evaluation post LVLP. For patients considered to be non-surgical candidates the use of acetazolamide (Diamox®) is recommended at relatively low doses (125-500 mg/day).[31]

SUMMARY

The treatment of Nph can be quite rewarding, both from the viewpoint of prevention of head injury, trauma and hip fractures in the elderly, as well as for the improvement of quality of life with disappearance of incontinence, freedom of ambulation and in some selected cases, cognitive improvement. As predicted 50 years ago, Nph is still one of the few curable causes of dementia in the elderly.

- Pre-Lumbar Puncture
  - Cognitive evaluation by neuropsychology
  - Physical therapy evaluation: gait & balance
  - Sphincter continence
- Large-volume LP: 50 mL under fluoroscopy
- Post-LP: Repeat pre-LP protocol within 24 hours.
  - Caregiver global impression of change

Table 1. NPH Evaluation at Houston Methodist Neurological Institute
(According to GC Román)

REFERENCES:

1. Houry D, Florence C, Baldwin G, Stevens J, McClure R. The CDC Injury Center's response to the growing public health problem of falls among older adults. Am J Lifestyle Med 2016 Jan-Feb; 10 (1). doi:10.1177/1559827615600137.
2. Halperin JJ, Kurlan R, Schwalb JM, et al. Practice Guideline: Idiopathic normal pressure hydrocephalus: Response to shunting and predictors of response. Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology. Neurology 2015; 85: 2063–2071.
3. Adams RD, Fisher CM, Hakim S, Ojemann RG, Sweet WH. Symptomatic occult hydrocephalus with "normal" cerebrospinal-fluid pressure. A treatable syndrome. N Engl J Med 1965; 273: 117–126.
4. Hakim S, Adams RD. The special clinical problem of symptomatic hydrocephalus with normal cerebrospinal fluid pressure. Observations on cerebrospinal fluid hydrodynamics. J Neurol Sci 1965; 2: 307–327.
5. Missori P, Paolini S, Currà A. From congenital to idiopathic adult hydrocephalus: A historical research. Brain 2010;133:1836–1849.
6. Brean A, Eide PK. Prevalence of probable idiopathic normal pressure hydrocephalus in a Norwegian population. Acta Neurol Scand 2008;118:48–53.
7. Iseki C, Kawanami T, Nagasawa H, et al. Asymptomatic ventriculomegaly with features of idiopathic normal pressure hydrocephalus on MRI (AVIM) in the elderly: A prospective study in a Japanese population. J Neurol Sci 2009;277:54–57.
8. Jaraj D, Rabiei K, Marlow T, Jensen C, Skoog I, Wikkelsø C. Prevalence of idiopathic normal-pressure hydrocephalus. Neurology 2014;82:1449–1454.
9. Hamilton R, Patel S, Lee EB, et al. Lack of shunt response in suspected Idiopathic Normal-Pressure Hydrocephalus with Alzheimer disease pathology. Ann Neurol 2010; 68:535–540.
10. Braak H, Braak E. On areas of transition between entorhinal allocortex and temporal isocortex in the human brain. Normal morphology and lamina-specific pathology in Alzheimer's disease. Acta Neuropathol (Berl) 1985;68(4): 325-332.
11. Braak H, Braak E. Neuropathological stageing of Alzheimer-related changes. Acta Neuropathol (Berl) 1991; 82(4): 239-259.
12. Braak H, Braak E. Diagnostic criteria for neuropathologic assessment of Alzheimer's disease. Neurobiol Aging 1997; 18 (4 Suppl): S85-88.
13. Sassin I, Schultz C, Thal DR, Rub U, Arai K, Braak E, Braak H. Evolution of Alzheimer's disease-related cytoskeletal changes in the basal nucleus of Meynert. Acta Neuropathol (Berl) 2000; 100 (3): 259-269.
14. Evans WA Jr. An encephalographic ratio for estimating ventricular enlargement and cerebral atrophy. Arch Neurol Psychiatry 1942; 47: 931-937.
15. Relkin N, Marmarou A, Klinge P, Bergsneider M, Black PM. Diagnosing idiopathic normal-pressure hydrocephalus. Neurosurgery 2005;57:S4–S16.
16. Mori E, Ishikawa M, Kato T, et al. Guidelines for management of idiopathic normal pressure hydrocephalus: Second edition. Neurol Med Chir (Tokyo) 2012;52:775–809.
17. Williams MA, Malm J. Diagnosis and treatment of idiopathic normal pressure hydrocephalus. Continuum 2016; 22 (2, Dementia): 579-599.
18. Peterson KA, Savulich G, Jackson D, Killikelly C, Pickard JD, Sahakian BJ. The effect of shunt surgery on neuropsychological performance in normal pressure hydrocephalus: a systematic review and meta-analysis. J Neurol 2016; doi:10.1007/s00415-016-8097-0
19. Fisher CM. Hydrocephalus as a cause of disturbances of gait in the elderly. Neurology 1982; 32: 1358-1363.
20. Sørensen PS, Jansen EC, Gjerris F. Motor disturbances in Normal-Pressure Hydrocephalus. Special reference to stance and gait. Arch Neurol 1986;43:34-38.
21. Román GC. Senile dementia of the Binswanger Type: A vascular form of dementia in the elderly. JAMA 1987;258:1782-1788.
22. Román GC. White matter lesions and normal-pressure hydrocephalus: Binswanger disease or Hakim syndrome? AJNR 1991;12: 40-41.
23. Masdeu JC, Wolfson L, Lantos G, et al. Brain white-matter changes in the elderly prone to falling. Arch Neurol 1989;46:1292–1296.
24. George AE. Chronic communicating hydrocephalus and periventricular white matter disease: A debate with regard to cause and effect. AJNR 1991;12:42–44.
25. Román GC. Lacunar dementia. In, Senile Dementia of the Alzheimer type. Hutton JT and Kenny AD (Editors), Alan R. Liss, Inc., New York, 1985; 131–151.
26. Dubinsky RM, Jankovic J. Progressive supranuclear palsy and a multi-infarct state. Neurology 1987; 37: 570–576.
27. Nutt JG, Marsden CD, Thompson PD. Human walking and higher-level gait disorders, particularly in the elderly. Neurology 1993;43:268-279.
28. Rasker JJ, Jansen ENH, Haan J, Oostrom J. Normal-pressure hydrocephalus in rheumatic patients. A diagnostic pitfall. N Engl J Med 1985;312:1239–1241.
29. Tinetti ME. Performance-oriented assessment of mobility problems in elderly patients. J Am Geriatr Soc 1986;34:119–126.
30. Berg KO, Wood-Dauphinee SL, Williams JI, Maki B. Measuring balance in the elderly: Validation of an instrument. Can J Public Health 1992;83 (suppl 2): S7–S11.
31. Alperin N, Oliu CJ, Bagci AM, et al. Low-dose acetazolamide reverses periventricular white matter hyperintensities in iNph. Neurology 2014; 82:1347–1351..

Neurology on the Hill 2016

Mike Amery, AAN Senior Legislative Counsel, Federal Affairs

The largest Neurology on the Hill ever took place on March 1 in Washington DC with 184 AAN members, 35 stroke advocates and 253 congressional offices visited. This was the 13th year that the American Academy of Neurology brought its passionate members to the nation's capital to advocate on behalf of their patients and profession.

Your colleagues were more unified than just their green bow ties and scarves. They were there to carry three messages to their lawmakers. First, Congress must pass the Furthering Access to Stroke Telemedicine Act (FASTact) that will allow Medicare to pay for stroke telemedicine consultations in urban areas. Second, Congress must support full funding for the BRAIN Initiative at NIH. Finally, "Meaningful Use" is anything but, and we seek the elimination of meaningless regulations that take precious time away from neurologists and patients.

These office meetings on the Hill are important not only to make our case, but to see how the legislators line up on the issues. Our participating neurologists are sharing this information with staff and this will help us with our follow-up tactics. We're seeing members of Congress steadily signing on as cosponsors of the FAST Act and in support of the BRAIN Initiative as a result of AAN member advocacy.

This year, the AAN broaden its advocacy to reach and include members of the public for the first time. We had great allies in the stroke participants and the American Heart Association/American Stroke Association. It was a very beneficial collaboration as these individuals put a human face to the issues facing neurologists and neurology as a profession. . Their personal stories were compelling and gave a sense of urgency to passing the FAST Act as soon as possible.

With another Neurology on the Hill in the books, we look forward to next year! The application for 2017 Neurology on the Hill will open this fall so keep an eye out for an email. In the meantime, stay up to date on AAN advocacy in Washington DC by reading the Capitol Hill Report at AAN.com.

2016 Texas Delegation

- Sara Austin, MD, FAAN – Austin, TX
- David Evans, MBA – Dallas, TX
- Rajani Caesar, MD – Longview, TX
- Kent Ellington, MD – Austin, TX
- Shamin Masrour, MD – Austin, TX
- Timea Hodics, MD – Dallas, TX
- Eddie Patton, MD, MS – Sugar Land, TX
- Carlayne Jackson, MD, FAAN – San Antonio, TX
- Shilpa Chitnis, MD, PhD, FAAN, FANA – Dallas, TX

Sports Concussion Assessment and Treatment: A Case-Based Application of the Literature

Bert B. Vargas, MD, FAAN
Associate Professor of Neurology and Neurotherapeutics
Director, Sports Neurology and Concussion Program
University of Texas Southwestern Medical Center
Dallas, Texas

Case Presentation

A 16-year-old male junior varsity football player with no reported previous medical history and no history of concussion presented to the emergency department after sustaining a suspected concussion during a game with a rival high school that evening. He was witnessed to sustain an on-field helmet-to-helmet collision when attempting to dive into the end zone for a touchdown. Witnesses reported a brief loss of consciousness lasting "a few seconds", after which he was noticeably confused and perseverating, repeatedly asking the score.

The school's athletic trainer initially assessed the athlete for cervical spine injury and cleared him to participate in a concussion assessment in the locker room. A Sport Concussion Assessment Tool 3 (SCAT 3) and King-Devick (K-D) test were used for evaluation. The athlete reported symptoms of headache, photophobia, and "dizziness" but did not report any cognitive complaints. Deficits were noted on delayed recall, attention, balance, and he was unable to complete the K-D without errors. He was transported by his parents to the emergency department where his exam was described as "non-focal" and non-contrast CT of the head was read as normal. He was discharged home with a diagnosis of concussion after a brief period of observation.

In clinic the following day his parents reported that he seemed to be "more like himself" cognitively; however, he did report numerous cognitive symptoms including feeling foggy, and difficulty with concentration. He could not recall any details of his injury, could not recall details of the game for about 10 minutes before his injury, and states he "woke up" to find himself in the emergency department. He continued to complain of headache with photophobia and phonophobia which responded to 400mg ibuprofen. A focused history revealed a prior history of milder headaches starting at age 12 also with associated mild photophobia and phonophobia, occurring roughly twice per year, and also responsive to ibuprofen. A family history of migraine in both his mother and paternal aunt was also identified.

On examination, his vital signs, general, and musculoskeletal exams were normal. On neurological exam, although no gross deficits were noted, his K-D was 3 seconds delayed (with no errors), and he had maximal errors on Modified Balance Error Scoring System (mBESS) for single-leg stance, and tandem stance.

Clinical Question

- What are some of the current opinions regarding this athletes evaluation, rehabilitation, and treatment with specific attention to:
  a. Interpretation of his examination findings
  b. Need for cognitive and physical rest
  c. Eventual return to learn and return to play
  d. Treatment of his headache and prognosis

The increased public awareness surrounding concussion has led to the inevitable development of a number of tools that claim to accurately identify abnormalities consistent with concussion. Despite the ever-expanding marketplace in concussion, there remains a relative lack of evidence supporting the use of many of these tools (many of which are expensive, time-intensive, or cumbersome) and the gold standard for concussion diagnosis continues to be the clinical exam.

The SCAT 3 is frequently used as a sideline assessment tool and is a gross measure of cognitive and balance function after suspected injury. Frequently, it is also used in the clinic to evaluate and track concussion; however, it is somewhat limited by a high degree of inter-rater variability and modest sensitivity and specificity. The symptom checklist has a sensitivity of 65-89% and a specificity of 91-100% for concussion [1] and results may vary widely if the athlete is asked to complete the questionnaire on his or her own versus completing it under the supervision of the provider. Input from parents and other family members may also influence the reliability of this self-reported data.

The Standardized Assessment of Concussion (SAC) portion of the SCAT 3 is a gross estimate of cognitive function including orientation, immediate memory, concentration, and delayed recall. The SAC has a sensitivity of 80-94% and specificity of 76-91%, [1] is subject to practice effects and can be difficult to interpret without a baseline or well-established normative data.

The mBESS is the balance portion of the SCAT 3 and also has a high degree of inter-rater variability in that the scoring of errors is highly subjective. The exam includes having the

athlete maintain a stable posture with feet together and eyes closed for 20 seconds, then standing on the non-dominant leg for 20 seconds, then with the feet in tandem stance with the dominant foot forward for 20 seconds. It has a sensitivity of 34-64% and a specificity of 91%. [1] Contributing to the overall poor reliability of the mBESS is the fact that the double leg stance is under-sensitive as it is frequently normal in concussion and the single leg stance is over-sensitive as it is overly difficult (even for non-concussed individuals) with many elite athletes achieving maximal error scores even on their normal baseline examination. [2]

The K-D test is a timed rapid number naming test that requires the athlete to read a series of numbered cards as fast as possible without making any errors. Cognitive and oculomotor abnormalities that frequently occur in concussion are frequently identified on K-D when compared to a pre-injury baseline. A decrement in performance by 5 or more seconds is seen in mixed martial arts fighters after sustaining head trauma; [3] however, the manufacturer recommends that any decrement should warrant remove from play and more in-depth evaluation for concussion. The K-D is abnormal in the setting of concussion 79% of the time, but one small study determined that any identified abnormality when combining the K-D, SAC and mBESS agrees with the clinical diagnosis of concussion nearly 100% of the time. [3]

The pitfalls of "strict rest"

Despite earlier recommendations that cognitive and physical rest were the mainstays of concussion treatment, [4, 5, 6] a number of studies have recently suggested that complete rest may be ineffective and, in some cases, counterproductive. One highly cited study addressed this very question by randomizing 99 concussed youth athletes into either a 5-day "strict rest" group or a "usual care" group with 1-2 days rest followed by a graduated return-to-learn and return-to-play. [7] Key findings in this study were that at 3 days and 10 days post-injury, no between-group differences were noted in assessments of cognition or balance. Furthermore, the "strict rest" group was noted to display slower symptom resolution and a statistically significant higher symptom severity scores at the 10-day end-point.

Although this study supports current practice and beliefs that the social isolation and inactivity of "strict rest" likely leads to deconditioning, higher rates of anxiety and depression, and a more protracted recovery, these data should be interpreted with caution as there appear to be a number of flaws with the methodology used to reaching these conclusions.

It is questionable whether there was a significant enough difference between the control and treatment groups as the differences in physical exertion were not much different between groups despite the treatment group being counseled to observe "strict rest" and the strict rest group did not appear to properly observe the protocol as the mean school and after-school attendance on days 2-5 was 3.8h (when it should have been zero if the protocol had been properly observed). Arguably, the relative homogeneity between the groups may have led to an unsurprising finding that balance and neurocognitive performance were not statistically different.

Regarding the differences observed in the symptom severities, there were no baseline severity assessments to adequately gauge true symptom resolution keeping in mind that 68% of male and 76% of female athletes will have one or more symptoms at baseline – even in the absence of concussion. [8] Furthermore, at the time of initial assessment in the ED, the mean scores of the two groups differed by 2 points and, at the end of 10 days, the mean scores of the two groups only differed by a total of 5 — for a total difference of only 3 points over 10 days in the control group. It is unclear how statistically significant this difference is (as it is not reported), but the clinical significance is questionable in the context of a scale with scores that can range between 0-132.

The "sweet spot" for the frequency and intensity of cognitive and physical activity appears to exist; however, there have been no studies to adequately quantify how much is too much and how much is too little. Although most experts would agree that the extremes of "strict rest" and "full activity" are both counterproductive to the recovery process, there is also wide agreement that supportive evidence is lacking and the degree to which a concussed athlete should rest is need of further research. [9]

Current concepts regarding rest and reintegration of cognitive and physical activity

Overall, prognosis after concussion is very good with most athletes recovering to their pre-injury baseline within days to weeks. Although 90% college football players return to their baseline symptom severities, and performance on cognitive and balance testing within 7 days [10] caution should be exercised in applying this timeline to youth athletes in whom cognitive, somatic, and emotional symptoms can persist for up to 5 weeks. [11, 12] Consensus opinion is that a step-wise, graduated return to learn and return to play should be implemented with gradual increases to the athlete's physical and cognitive load as long as they remain asymptomatic at each interval. [4]

Coordinating return to learn protocols can be more difficult than return to play protocols since "cognitive rest" and recovery are difficult to define or quantify the way "physical rest" can. For lack of evidence-based guidance, some consider 1-2 days of cognitive rest appropriate in the acute phase of injury with return to the classroom (in some capacity) during the sub-acute phase. [13] During this time, school accommodations may be necessary including the allowance of extra time to complete assignments and tests.

Although the concept of "physical rest" is also somewhat nebulous, it is arguably easier to define and implement than cognitive rest. Although there is some evidence that forced exercise may be detrimental to recovery in animal models, there is a growing body of evidence supporting early implementation of symptom-limited exercise. [14] There is wide agreement that any athlete suspected of having concussion

Continued on page 14

Continued from page 13

should immediately be removed from competition and should never return on the same day; however, the specific process through which an athlete is reintegrated into physical activity remains unclear. [4] Current guidelines recommend a 6-step graduated protocol which implements higher levels of physical activity with a progression to the next level if the athlete remains asymptomatic. Unfortunately, the specific goals and levels of exertion at each step are somewhat open to interpretation.

In the setting of acute injury, low level exercise appears to be safe and potentially assists in recovery. [14] For athletes who remain symptomatic at 4-6 week after injury, the Buffalo Concussion Treadmill Test (BCTT) has been studied as a specific exercise protocol that also appears to be safe and effective for symptom recovery. The BCTT establishes the symptom exacerbation threshold after which the athlete is exercised until reaching 80-90% of the threshold heart rate for 20 minutes or terminated early if symptoms occur. This is repeated for 5-6 days per week for two weeks at which time a new symptom threshold can be established. Once the athlete is able to exercise at 85-90% of their age-predicted maximum heart rate over the course of several days, physiologic recovery is felt to have occurred and the remaining steps of the traditional return to play protocol can then be initiated. [14] Clearly, the BCTT cannot truly determine if the structural and metabolic derangements associated with concussion are fully resolved and more research is needed to help determine the role of exercise in recovery and rehabilitation.

Managing headache post-concussion

Post-traumatic headache (PTH) is the most common symptom after concussion and has been reported in up to 94% of athletes after injury. [15] The predominant phenotype of PTH is migraine or probable migraine and comprise greater than 60% of all headaches reported. [16] Unfortunately, there are no treatment guidelines specific to PTH and the current guidance is to manage according to accepted treatment algorithms based on the phenotype they most represent. [17]

PTH is frequently multifactorial and it is important to consider cervicogenic contributors (suggesting a role for trigger point injections or regional nerve blockade) as well as other issues including dehydration, and side-effects to medications which may have been prescribed to treat post-concussion symptoms. [18] In the absence of any of these contributors, a number of uncontrolled, open-label studies have demonstrated efficacy of a variety of medications commonly used for migraine in the setting of PTH. For abortive treatment, non-steroidal anti-inflammatories and triptans are commonly used and tricyclic antidepressants, β-blockers, and anticonvulsants such as valproate or topiramate have demonstrated some degree of efficacy for prophylaxis. Caution should be exercised, however, as many of these prophylactic medications can cause sedation or cognitive side effects which may be misinterpreted as residual effects of concussion potentially prolonging treatment and recovery.

Additional factors which may complicate PTH treatment include the presence of pre-existing or pre-disposition to primary headache disorders as identified on a detailed headache history and/or family history. Although it is generally felt that an athlete may only return to play after symptomatic recovery and discontinuation of any medications which may mask symptoms, identification of untreated and frequently occurring headache may have warranted prophylactic treatment even prior to concussion. These decisions are nuance and may require the guidance of both a headache and a concussion specialist. The prolonged and frequent use of abortive treatments should be also monitored closely as one recent study identified over 70% of children with headache at 3 months post-concussion as meeting criteria for medication overuse headache the majority of whom improved once the overused medication was discontinued. [19]

Although the prognosis of PTH is generally favorable, mostly resolving with other symptoms over a matter of weeks, headache may persist at 3 months in 47-78% of individuals. [18] This, coupled with the fact that athletes with PTH tend to have a prolonged recovery course, [20, 21] suggests that thoughtful and aggressive treatment of headache should be considered.

Case conclusion and outcome

This athlete was able to return to school the following day without a need for accommodations and was prescribed a graduated sub-threshold exercise plan under the guidance of physical therapy. His headaches appeared to be of similar phenotype to his his pre-existing headaches which were determined to meet criteria for migraine. Additional history revealed that ibuprofen only partially resolved his headaches and he was therefore prescribed rizatriptan which he reported as more effective at achieving total pain freedom without headache recurrence. He recovered to his baseline-level of symptoms within 10 days and was then placed on a relatively conservative graduated return to play protocol for an additional 10 days. Near the end of his return to play protocol, he was re-examined and his neurologic, K-D, SCAT3, and computerized cognitive exams were back to their pre-injury baseline. Thoughtful interpretation of the exam findings and history yielded a final return to play determination. He and his parents were counseled regarding signs and symptoms of concussion and the need to be immediately from play should they recur. His pre-existing history of migraine, history of concussion, particular sport, and position were also discussed as risk-factors for future concussion. His headache frequency returned to its pre-injury baseline and he was counseled to continue using rizatriptan for abortive migraine treatment.

Although this case was relatively uncomplicated and in many respects represents a best-case-scenario, it highlights a number of issues that the clinician frequently must address when evaluating concussed athletes. Current recommendations are mostly based on expert opinion and more well-controlled studies are clearly warranted. At this time, a more conservative approach is warranted – especially with youth athletes and any new or unusual symptom occurring in the context of suspected concussion should warrant immediate removal from play and thorough evaluation by an expert. Although a common mantra regarding suspected concussion is "when in doubt, sit them out," many believe that "when in doubt, sit them out and check them out" [22] may represent a better and more sensible approach.

REFERENCES:

1. Guskiewicz KM, Register-Mihalik J, McCrory P, et al. Evidence-based approach to revising the SCAT2: introducing the SCAT3. British journal of sports medicine 2013 Apr 1;47(5):289-93.
2. Starling AJ, Leong DF, Bogle JM, Vargas BB. Variability of the modified Balance Error Scoring System at baseline using objective and subjective balance measures. Concussion 2015(0):1-8.
3. Galetta KM, Liu M, Leong DF, et al. The King-Devick test of rapid number naming for concussion detection: meta-analysis and systematic review of the literature. Concussion 2015(0).
4. McCrory P, Meeuwisse WH, Aubry M, et al. Consensus statement on concussion in sport: the 4th International Conference on Concussion in Sport held in Zurich, November 2012. British journal of sports medicine 2013;47:250-8.
5. Silverberg ND, Iverson GL. Is rest after concussion "the best medicine?": recommendations for activity resumption following concussion in athletes, civilians, and military service members. The Journal of head trauma rehabilitation 2013;28:250-9.
6. Moser RS, Glatts C, Schatz P. Efficacy of immediate and delayed cognitive and physical rest for treatment of sports-related concussion. The Journal of pediatrics 2012;161:922-6.
7. Thomas DG, Apps JN, Hoffmann RG, et al. Benefits of strict rest after acute concussion: a randomized controlled trial. Pediatrics 2015;135:213-23.
8. Covassin T, Swanik CB, Sachs M, et al. Sex differences in baseline neuropsychological function and concussion symptoms of collegiate athletes. Br J Sports Med 2006;40:923-927.
9. Schneider KJ, Iverson GL, Emery CA, et al. The effects of rest and treatment following sport-related concussion: a systematic review of the literature. British journal of sports medicine 2013;47:304-7.
10. McCrea M, Guskiewicz KM, Marshall SW, et al. Acute effects and recovery time following concussion in collegiate football players: the NCAA Concussion Study. JAMA 2003;290:2556-63.
11. Covassin T, Elbin RJ, Nakayama Y. Tracking neurocognitive performance following concussion in high school athletes. The Physician and sportsmedicine 2010;38:87-93.
12. Sroufe NS, Fuller DS, West BT, et al. Postconcussive symptoms and neurocognitive function after mild traumatic brain injury in children. Pediatrics 2010;125:e1331-9.
13. Eastman A, Chang DG. Return to Learn: A review of cognitive rest versus rehabilitation after sports concussion. NeuroRehabilitation 2015;37:235-44.
14. Leddy JJ, Baker JG, Willer B. Active Rehabilitation of Concussion and Post-concussion Syndrome. Physical Medicine and Rehabilitation Clinics of North America 2016;27:437-54.
15. Marar M, McIlvain NM, Fields SK, Comstock RD. Epidemiology of concussions among United States high school athletes in 20 sports. The American journal of sports medicine 2012;40:747-55.
16. Lucas S, Hoffman JM, Bell KR, et al. Characterization of headache after traumatic brain injury. Cephalalgia 2012;32:600-6.
17. Gladstone J. From Psychoneurosis to ICHD-2: An Overview of the State of the Art in Post-Traumatic Headache. Headache: The Journal of Head and Face Pain 2009;49:1097-111.
18. Seifert TD. Sports Concussion and Associated Post-Traumatic Headache. Headache: The Journal of Head and Face Pain 2013 May 1;53(5):726-36.
19. Heyer GL, Idris SA. Does analgesic overuse contribute to chronic post-traumatic headaches in adolescent concussion patients?. Pediatric neurology 2014;50:464-8.
20. Kontos AP, Elbin RJ, Lau B, et al. Posttraumatic migraine as a predictor of recovery and cognitive impairment after sport-related concussion. The American journal of sports medicine 2013 May 22:0363546513488751.
21. Mihalik JP, Stump JE, Collins MW, Lovell MR, Field M, Maroon JC. Posttraumatic migraine characteristics in athletes following sports-related concussion. Journal of neurosurgery 2005;102:850-5.
22. McCrory, Paul. "Head Injuries in Sports." Sports Injuries: Prevention, Diagnosis, Treatment and Rehabilitation 2015: 2935-2951.

Make Your Plans Now!

Robert W. Fayle MD - Education Committee Chair

The TNS Summer Conference will held Friday, July 15, 2016 and Saturday, July 16, 2016 at La Cantera Resort in San Antonio, TX. La Cantera has a definite family appeal, so bring the family and enjoy the weekend. The Program Directors, Dr. Clark, pediatric neurology, and Dr. Vanderlick, adult neurology, have arranged an excellent meeting. There are both applied basic neuroscience and clinical subjects, which will be of interest to the clinical neurologist. They have considered the many requests from the members of the society to provide talks on subjects of interest, and they have done an excellent job in the preparation of the meeting.

The Pediatric session begins at 8:00 AM on Friday morning with talks on Infectious Diseases, Movement Disorders and the Ethics of Quality of Life in Pediatric Neurology.

The Adult Session begins Friday afternoon and covers whole genome sequencing, Autoimmune Encephalopathies, Neurosarcoid, new treatments for Migraine, Neuromuscular Emergencies, Myopathy and Pseudo tumor. There is a timely and important Ethics hour on The TMB rules on Pain Management.

Please plan to stay for the final program on Saturday afternoon on Practice Management, developed by the Medical Economic Committee. The TNS board and the education committee is planning to add this program in addition to the clinical meeting to provide information about the ever changing and crucially important business changes facing the neurologic practioner.

As always the TNS meeting provides a great opportunity to get together with friends and colleagues from that we do not have the chance to see regularly. Look for the registration information in this issue of Broca's Area.

Congratulations!
Stuart Black, MD

TNS 2016 Lifetime Achievement Award Recipient

Legislative Update

Sara Austin, MD, Legislative Committee Chair and Greg Herzog, TNS Lobbyist

With almost every breath of oxygen being sucked out of the political world by the race to become the President of the United States, now is a good time to draw our attention issues closer to our practices and patients. TNS continues to monitor and engage in several important topics at the state level.

Out of Network Balanced Billing

For many legislative sessions, the health plans have argued that a physician's ability to bill a patient for services provided out of network should be prohibited or severely curtailed. Furthermore, the health plans have argued to the legislature that those physicians should accept whatever payment it offers to those physicians, even though no contract exists between the two.

Organized Medicine has argued that the health plans have put together inadequate networks of physicians and offer such low-ball fees that it makes entering into a contract with a particular health plan a losing proposition. Furthermore, physicians, as independent providers of a service in a free-market system, believe that they should be able to set a price for a service and expect reasonable compensation for services provided.

The issue is complicated, especially for patients who receive a 'surprise medical bill'. The issue has garnered extreme attention in state legislatures including New York, California, and Florida. http://www.modernhealthcare.com/article/20160408/NEWS/160409899

Here in Texas, organized medicine expects a major push by the health plan lobby and the patients rights groups to pass a similar measure like the one recently adopted in Florida. While the issue of Out-of-Network Balanced Billing might today impact other specialties more directly than Neurology, the TNS Board remains worried what changes might result from physicians ability to bill out of network. Given that physicians have limited leverage with the health plans currently, how might a ban on out of network billing further erode that leverage?

TMA and its specialty societies, including TNS have been discussing acceptable legislative solutions to this issue. TMA's House of Delegates approve to actively fight the balanced billing legislation during TexMed. More to come!

Sunset Review of the Texas Medical Board

Every 12 years, each state agency in Texas is required to go through a review process by the Texas Sunset Commission. The Commission reviews the agency to make determine if it is meetings its core functions and to make recommendations to the legislature for improvements. Every agency in Texas has a sunset date, meaning that the agency is set for termination, unless the legislature proactively passes legislation reauthorizing that agency.

This cycle, the Texas Medical Board is scheduled for Sunset. Currently, the Sunset Commission is reviewing the core functions of the Board with a keen eye towards the licensure and disciplinary processes of the TMB. The Commission will also be reviewing the TMBs response to emerging issues like the use of telemedicine in Texas and the prescribing of opioids by physicians.

In order to reauthorize an agency, the legislature must pass a piece of legislation. This legislation is a bill like any other bill and is subject to amendment at any step of the process. Since the entire practice act is open for amendment, organized medicine must be very cognizant and alert to how potential changes might impact our practices and scope of practices. The Sunset Commission will issue its report this summer.

Texas Dental Board and sleep related rules

The TMA/TNS lawsuit against the Texas Dental Board continues to slowly grind forward. As you may recall, the TDB adopted rules that will allow dentists to independently diagnosis, test, and treat the medical condition of sleep apnea.

While the TDB has recently proposed rules that were a step forward, TMA and TNS continue to have significant concerns with the power granted to dentists with regard to the treatment of sleep conditions.

Our organizations remain steadfastly against this intrusion and thank you for your continued support of our position.

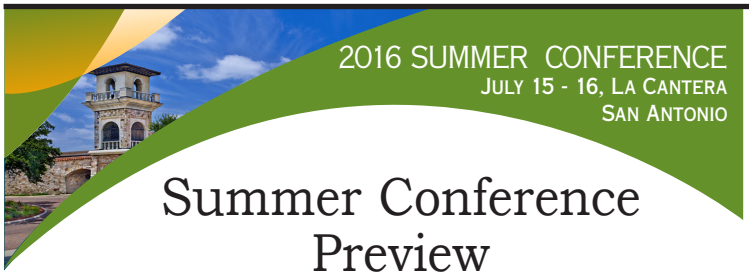
Emerging Issues

Physician prescribing practices remain under close scrutiny. The opioid addiction crisis has garnered attention from candidates and office holders from all political persuasions. Some states have even begun to consider limitations on certain types of prescriptions. The health plans are also engaged in our prescribing practices. Greater use of step therapy or fail-first protocols seem to be turning our prescription pads into "suggestion" pads. We expect these issues will be points of discussion in the months leading up to next session.

The outlook for the State's fiscal picture does not appear to be very positive. The downturn in the Texas oil and gas economy has weakened the state's overall revenue. Texas is facing many uncertain revenue and appropriations challenges. Those of us that provide services in the Medicaid program know that physicians have not received a significant bump in reimbursements in a generation. These uncertain financial times do not bode well for new Medicaid rates; in fact, the conventional wisdom in Austin is we may need to be on guard for cuts.

https://www.texastribune.org/2016/03/31/lawsuits-oil-prices-could-spell-budget-doomsday/

Your TNS Board always welcomes your feedback and input on these and any other issues you believe are important to our specialty and our patients. Please feel free to contact me directly or the TNS Headquarters with any comments.



Summer Conference Preview

Mary Ellen Vanderlick MD, Program Director

Those challenging cases where neurosarcoidosis is in the differential. The atypical dementias, especially in young patients when an autoimmune cause is possible. What to do with your challenging pseudotumor cerebri patients.

These are the hot topics for the TNS summer conference in San Antonio Texas at the La Cantera resort on July 15 and 16, 2016.

We have Dr. Jeffrey Gelfand from UCSF to discuss autoimmune encephalopathies and his special interest, neurosarcoidosis. Dr. Christine Eng from Baylor College of Medicine will give practical implications for the human genome in testing for neurological disease. Dr. Robert Bredt from the TMB will explain our responsibilities in dispensing controlled substances to our patients.

We have experts in their fields to discuss neuromuscular emergencies and neuromuscular case presentations. And finally, a discussion on the new delivery systems for multiple generic drugs in the field of migraine. Are they worth it?

Join us for these fascinating topics this summer in San Antonio!

TNS Past Presidents Council

Bill Gilmer MD, PPC Chair

The Past Presidents Council met for the first time during the February 2016 Winter Conference with 17 past presidents attending. At the request of the board of directors, the PPC will provide a historic knowledge center for TNS and an internal focus group for the future. Your Past Presidents hold collective institutional knowledge which can help inform decision making and future direction.

Each new generation of leadership adds to that knowledge and connection base. PPC provides a deep field of contacts to continue our excellent CME programs. We offer a ready source of mentors for residents and fellows, helping make connections for those coming out into practice, or looking for a new opportunity.

We want make good use of our collective institutional memory as a valuable resource for future growth. Your leadership is passionate about TNS, committed to making it stronger and more valuable to all its members.

TNS was founded in 1975 by a handful of forward thinking Texas Neurologists. It has grown into the largest and most successful neurology organization in the country, second only to the American Academy of Neurology.

In Remembrance...

Martin Steiner, MD

On January 18, 2016, the city of Houston lost one its long time leaders in neurology, when Martin Steiner,M.D. died prematurely at the age of 74 from an aortic dissection.

Marty grew up in New York. He graduated from Florida State University in 1963. He attended the University of Florida College of Medicine, obtaining his M.D. degree in 1967. In 1967, Marty performed an internship at Ben Taub General Hospital in Houston Texas. He then returned to the east coast for a residency in neurology at Mount Sinai Hospital. It was there that he initially developed his clinical skills in neurology under the mentorship of Morris Bender, M.D., who not only was an internationally famous neurologist, but also Marty's father-in-law. Suffice it to say, Marty dealt with pressure very well.

In 1971, Marty returned to Texas as a Major in the Air Force at Wilford Hall Medical Center in San Antonio. Following his military service, Marty and his family settled in Houston to begin his private practice in clinical neurology.

I first met Marty in 1975. We became good friends and partners for more than 40 years. In 1982, we joined Ray Martin and Ron Devere to form Houston Neurology Associates, a relationship that lasted for more than 25 years.

Marty was an active participant in the profession of neurology. He served in various leadership rolls. He was a past president of the Houston Neurological Society, as well as a past president of the Texas Neurological Society.

Marty was a very generous, principled and compassionate person. He was all always offering to help others.

He had many interests and was curious about the world. He enjoyed cooking, boating, diving, skiing and recreational traveling. He loved visiting all of his children, and especially, his grandchildren.

Marty Steiner is survived by his beloved wife Marilee, with whom he shared many wonderful adventures in the last 8 years, and his three daughters and sons in law, as well as grandchildren. His first wife, Barbara (Bender) Steiner, died in 2006.

Marty Steiner will be greatly missed by those of us who were fortunate enough to enjoy his friendship.

Written by: Len Hershowitz, MD
Houston Neurology Associates

Medical Economics and Resources Committee Update

Stuart Black, MD, Medical Economics Committee Chair and Kristi Berrier, TNS Medical Economics Advisor

At the February 2016 Winter Conference, the Medical Economics and Resource Committee sponsored a Practice Management Symposium entitled "Changes in CMS Reimbursement: CMS Medicare Access and CHIP Reauthorization Act of 2015 (MACRA)". Attendees learned about the essential aspects of MACRA, the impact it may have on reimbursement and how existing CMS programs might be modified to accommodate MACRA implementation. The committee would like to thank the speakers and panelists who participated:

- Edward Patton, MD, MS - AAN Government Relations Committee member, Chair of the Federal Advocacy Workgroup
- Shannon Vogel, FHIMSS – Director of the Health Information Technology Department, Texas Medical Association
- Daniel Spirn, JD, MA – AAN Regulatory Counsel
- Stuart Black, MD – Chair TNS Medical Economics Resource Committee, Member AAN Medical Economics Committee and Alternative Payments Team

On April 27, 2016, the Department of Health and Human Services released its proposed final rule on MACRA. The 962 page document describes how the agency anticipates implementing the physician payment reforms passed by the House and Senate in a rare landslide vote last April. The proposed rule gives insight into how Medicare will shift from traditional fee for service reimbursement to value based reimbursement for physicians and providers. Between now and June 27, the AAN and AMA, among other entities, will be evaluating and responding to the proposed rule.

In order to help keep members informed about MACRA and other practice management issues, the TNS Board of Directors voted at its February meeting to add a Practice Management Symposium to the 2016 summer meeting in July. The symposium, free to meeting attendees, will be held Saturday, July 16 from 12:45 to 2:45. Dr. Stuart Black, a member of the AAN Payment Alternatives Team, will be presenting on MACRA. In his talk, Dr. Black will provide information on how physician reimbursement under MACRA will be calculated, the latest news on the development of Alternative Payment Models (APM) and Physician Focused Payment Models (PFPM - a new model which the AAN is involved in developing that will allow Neurologists to participate in and APM), and how the AXON Registry will help Neurologist satisfy quality reporting standards. Katie Shepherd, AAN Associate Director of Medical Economics, will also be speaking. Physicians are welcome to invite their office staff to what promises to be a very informative Practice Management Symposium.

For news and up to date information about Texas Neurological Society visit:
www.texasneurologist.org

19th Annual Winter Conference Supporters

Sunovion
Bronze Sponsor,
Attendee Lanyards
and Envelopes Sponsor

Cynapsus Therapeutics Inc.
Bronze Sponsor

Abbvie
Bronze Sponsor

Houston Methodist
Internet Passkey Sponsor

Avanir
Meeting Sponsor

Lundbeck
Meeting Sponsor

Medtronic
Meeting Sponsor

Poster Winners Winter Conference

First Place	**Proleta Datta, MD**
Second Place	**Parunyou Julayanont, MD**
Third Place	**Divyanshu Dubey, MD**

Welcome New Members!

Alexandra E. Armitage, APRN
Darlene Bedwell, PA-C
Mary Concepcion, NP
Melinda Ann Gottschalk, PA-C
Shivaram K. Gowdagere, MD
Clay Johnston, MD
Rachel Kilian
Batool Kirmani, MD
Rosa Luna, NP
Bhuvanaprasad Lakshmi Mandalapu, MD
Cornelius Matwijecky, MD
Raja Mehanna
Claudia Regina Padilla, MD
Sudama Reddi
Alpa Shah, MD
Kiran Shah, MD
britt stone
Madhavi Thomas, MD
Gabrielle Tucker, NP
Laura Wu
Terry William Zuehsow, PA-C

Texas Neurological Society

2016 Summer Conference Registration

Save by registering early! Your registration fee includes lectures, breakfast, and breaks.

Register online at www.texasneurologist.org

Please Print or Type:

Name: _____

Address: _____

City: _____ State: _____ Zip: _____

Email: _____

Phone: _____ Fax: _____

Contact person & phone number in case of on-site emergency: _____

Registration (please check appropriate box and note amount due below)

	By July 1, 2016	After July 1, 2016
❑ TNS Member	$175	$225
❑ TNS Resident Member	$0	$50
❑ Non Member	$275	$300

(Registration for the TNS conference is intended for healthcare providers including physicians, nurses, and physician assistants. It is not intended for pharmaceutical or industry representatives.)

Amount Due for Registration: $_____

Please mail or fax registration to:
Texas Neurological Society
401 W. 15th St., Suite 100
Austin, TX 78701
Fax (512) 370-1626

Call (512) 370-1532 or email Krista.cottingim@texmed.org with any questions

TNS Dues:

❑ TNS membership 2016 - $75

Support Your Future Partner!

❑ Yes, I would like to donate $25 to help a resident member attend the TNS conference

Syllabus: Do you want a printed syllabus: ❑ Yes ($60) ❑ No, I will view the presentations online

Social Activies: Check if you plan to attend: ❑ Friday Welcome Reception: + _____ guests

❑ Practice Management Symposium

Total Amount Due: $ _____

Payment Information

Enclosed is a check for $_____ Make checks payable to Texas Neurological Society.

Please charge my credit card: ❑ Visa ❑ MasterCard ❑ AMX

Card #: _____ Exp. Date: _____

Cardholder name: _____ Security Code: _____

Signature: _____

Refunds: Written notice of cancellation must be received by July 1, 2016 in order to receive refund minus $25 processing fee.

In accordance with the American with Disabilities Act, please let us know in writing, of any special accommodations you may need.

Registration Deadline: July 1, 2016 ★ Hotel Deadline: June 23, 2016

Texas Neurological Society
401 West 15th Street, Suite 100
Austin, Texas 78701-1680

PRSRT STD
U.S. POSTAGE PAID
PERMIT NO. 1787
AUSTIN, TX

Texas Neurological Society's 2016-2017 Officers

President
Deborah Carver Hodges, MD
San Antonio

President-Elect
Erin E. Furr-Stimming, MD, Houston

Vice President
Aziz Shaibani, MD, Houston

Immediate Past President
Robert F. Leroy, MD, Dallas

Members-at-Large
Waleed El-Feky, MD, Dallas
Ed Fox, MD, PhD, Austin
Doug Lewis, MD, Amarillo
Gary Clark, MD, Houston
Brian Loftus, MD, Houston

Education Committee Chair
Robert W. Fayle, MD, Houston

Program Director
Mary Ellen Vanderlick, Houston
Summer Conference 2016

Eddie Patton, Houston
Winter Conference 2017

Delegate to TMA
Ed Fox, MD, PhD, Austin

Alternate Delegate to TMA
Sara Westgate, MD, Austin

Newsletter Editor
Randolph W. Evans, MD
1200 Binz St, Ste 1370
Houston, TX 77004-6952

Executive Director
Ky Camero
401 W. 15th St.
Austin, TX 78701
512.370.1532

Save the Date!

TNS Winter Meeting
2017
February 24-26
Hyatt Regency
Austin, TX