TEXAS NEUROLOGICAL SOCIETY SUMMER 2015

Broca's Area

The Voice of Texas Neurology

President's Message

Robert F. Leroy, MD

Dear Colleagues,

I wish to reflect on the activity of the Texas Neurology Society since the winter meeting. The most important activity has been the planning for the summer meeting in Fort Worth. I thank Drs. Loftus and Ready for the program. It is an expanded meeting with our traditional general neurology and partnering with the Southern Headache Society. It will prove practical and enlightening. In addition, I hope you enjoy the great City of Fort Worth. The downtown Sundance Square is great evening fun with dining and exploring. During the day, family activities range from culture to cowboys. Plan to visit the stockyards and the zoo. The Kimbell is a world class museum across the street from the Modern. During the July afternoon, explore the Botanical Gardens especially the Japanese Gardens. Motor over to Cowboy stadium, Six Flags and Hurricane Harbor in Arlington. The 2016 winter meeting in Austin as always will prove the best CME and camaraderie for the cost.

The advocacy activity of the TNS has been centered on the Texas Legislature. Bills we have monitored and opined on include protecting scope of practice from optometrists, dentists and chiropractors. There has been activity concerning High CBD and low THC marijuana for the treatment of refractory epilepsy – TNS has been supportive of further research. It will impact neurological practice because it is for the 140,000 Texans who have refractory epilepsy and requires neurological diagnosis of refractory epilepsy. In addition there has been a bill concerning the licensure of electrodiagnostic technologists. This appears to be better for a higher standard of performance but may limit availability of technologists in offices and laboratories for EEG, video EEG, IOM, sleep studies, and EMG/NCV. There might be a slippery slope for future scope of practice issues.

There have been discussions about availability of board members for issues from the membership. This has been brought up concerning current Maintenance of Certification and ICD-10. Of course, membership can always contact board members and we welcome involvement as a member or joining the leadership.

The health of Texas Neurology Society remains robust. Financially we are stable. However, our survey of members during the 2015 winter meeting suggests that we are an aging membership and are largely private practitioners. The renascence of the TNS in the late 1990's was based on intellectual leadership from the academic neurology departments which has waned. We are probably needing to reach out to young faculty, residents and fellows for the future. The challenge of changing medical economic models may require us to change TNS.

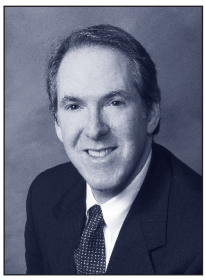
See you in Fort Worth!

Mark Your Calendar

2015 SUMMER CONFERENCE

July 31-Aug 2, 2015

Sheraton Fort Worth
Fort Worth, 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 Summer Conference in Fort Worth, July 31-August 2. Brian Loftus, program chair; Bob Fayle, education committee chair; and the education committee have planned an excellent program in coordination with Michael Ready and the Southern Headache Society for the first joint meeting.

Since headache will be the subspecialty focus for the summer conference, here are two headache cases (not that I ever need an excuse to present headache cases).

A One Month Headache in a Migraineur

This is a 35 year old female with a history of occasional migraine with visual aura as a teen and migraine without aura since her 20s occurring 1-2 times per month described as a bifrontal aching with light and noise sensitivity.

One month prior, she looked behind the seat in the car and developed a mild crick in her neck. A few hours later, she developed a constant nuchal-occipital and generalized pressure and throbbing without associated symptoms with an intensity of 9-10/10. Her pcp gave her Imitrex and Midrin which helped a little. Over a couple of weeks, the headaches were only bifrontal. After one month, the headaches were still constant, bifrontal and right temporal with an intensity of 3-10/10. Neuro exam normal
Question: What is the diagnosis?

Could this be migraine status? This is possible but the headache was different than prior migraines without associated light and noise sensitivity to fit migraine criteria. Can migraine status last a month?

Migraine status has been rarely studied. In a retrospective French study of 25 patients with status migrainosus seen in a tertiary care center (out of 8821 migraineurs over 11 years), the demographics were as follows: mean age at first episode, 39 years; male: female, 84%; duration 4.8 weeks (3-10); relapse of status migrainosus, 32%; and delay of relapse, 61.5 months (Beltramone M, Donnet A. Status migrainosus and migraine aura status in a French tertiary-care center: An 11-year retrospective analysis. Cephalalgia. 2014;34(8):633-637). Precipitating factors included the following: stress/anxiety, 69%; menses, 31%; and lack of sleep, 6.3%. Sixty percent had to be hospitalized for intravenous treatment. The great majority of patients had the same attack frequency before and after the status and most cases occurred in those with low-frequency migraine attacks.

Since migraine is a diagnosis of exclusion and the headache was different than her prior migraines, a migraine mimic or secondary cause should be excluded (Evans RW. Migraine mimics. Headache. 2015;55(2):313-22).

Neuroimaging and follow-up: MRI of the brain showed no ischemic lesions. MRA of the neck suggested a dissection of the proximal to mid left vertebral artery. The CTA was also suggestive but not diagnostic of a left vertebral dissection. A cerebral arteriogram showed a left vertebral artery aneurysm at the C6 level with pseudoaneurysm just distal to the narrowing with 50% stenosis. She was placed on heparin and warfarin for 3 months. MRA after 3 months showed recanalization. The headaches were still fairly constant 7 months later.

Questions: How often are headaches associated with cervical artery dissection and where is the headache in vertebral artery dissection? Can CAD cause new daily persistent headache? How often is the imaging indeterminate in diagnosing dissection? How often do strokes occur with dissection? Is antiplatelet or anticoagulation treatment preferred?

Headache or neck pain is the only symptom of spontaneous cervical artery dissection in 8% and can mimic migraine with and without aura and migraine status (Arnold M, Cumurciuc R, Stapf C, Favrole P, Berthet K, Bousser MG. Pain as the only symptom of cervical artery dissection. J Neurol Neurosurg Psychiatry. 2006;77(9):1021-4). The headache has a thunderclap onset in about 20% of cases. The incidence of spontaneous cervical carotid artery dissections is about 2.6/100,000/year and for cervical vertebral artery dissections about 1.5/100,000/year.

Headache occurs in about 70% of those with cervical vertebral artery dissection (VAD) with head or neck pain preceding other neurological symptoms and/or signs by a mean time of 14.5 hours (Hsu YC, Sung SF. Spontaneous vertebral artery dissection with thunderclap headache: a case report and review of the literature. Acta Neurol Taiwan. 2014;23(1):24-8). VAD is typically a severe ipsilateral occipital or posterior neck throbbing or steady and sharp pain but can be bilateral and rarely bilateral or generalized (Arnold M, Bousser MG. Clinical manifestations of vertebral artery dissection. Front Neurol Neurosci. 2005;20:77-86). The headache is rarely associated with migraine features such as nausea, vomiting, photophobia, or phonophobia, and visual aura.

The mean duration of the headache in one study was 8.3 days (range 2-35 days). As in this case, there are reports of CAD, more so ICAD, resulting in a secondary new daily persistent headache (Mokri B. Headache in cervical artery dissections. Curr Pain Headache Rep. 2002;6:209-216).

Headache is also common in cervical internal carotid artery dissection (ICAD), occurring in 60-95% and preceding other neurological symptoms and/or signs by a mean time of 4 days. The pain of ICAD, which is ipsilateral in 91% of cases, is typically localized to the frontal or temporal area, jaw, ear, and/or orbit and is more often aching than throbbing. A partial Horner syndrome occurs in about 25% of cases with ptosis and miosis. ICAD can have migraine features including nausea and vomiting.

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Seemingly forever (although the first description of spontaneous cervical artery dissection was 100 years ago [Turnball HM. Alterations in arterial structure and their relation to syphilis. Q J Med 1915;8:201-54), neurologists have debated the best medication treatment for cervical dissections (the jury is still out on the benefit of endovascular therapy). And now we may have the best answer we may have for many decades.

The Cervical Artery Dissection in Stroke Study (CADISS) was a randomized trial at hospitals with specialized stroke or neurology services (39 in the UK and 7 in Australia) of 250 patients with 118 extracranial carotid or 132 extracranial vertebral dissections with onset of symptoms within the prior 7 days (CADISS trial investigators. Antiplatelet treatment compared with anticoagulation treatment for cervical artery dissection (CADISS): a randomized trial. Lancet Neurol. 2015 Apr;14(4):361-7) with a mean age of 49 years (range 18-87). Presenting symptoms in the 250 patients were as follows: ischemic stroke, 195; TIA, 29: headache, 22; neck pain, 22; Horner's syndrome, 4. Patients were randomly assigned to antiplatelet (aspirin, dipyridamole, or clopidogrel alone or in combination) or anticoagulant drugs (unfractionated heparin or low-molecular weight heparin followed by warfarin aiming of an INR of 2-3) for 3 months with the specific treatment decided by the local clinician. In the antiplatelet group, 22% received aspirin alone, 33% received clopidogrel alone, 1% received dipyridamole alone, 28% received aspirin and clopidogrel, and 16% received dipyradomole. In the anticoagulant group. 90% received heparin and warfarin and 10% received warfarin alone.

The diagnosis of dissection could not be confirmed on central imaging review in about 20% of participants despite evidence of dissection on angiographic imaging or cross-sectional imaging through the vessel wall. In some cases, imaging was of poor quality. In others, alternative diagnoses were suggested including atherosclerosis with an atretic artery, a narrowed artery or adherent thrombus without dissection.

Excluding patients where dissection could not be confirmed, stroke occurred in 3 of 101 patients in the antiplatelet group (3%) vs 1 of 96 patients (1%) in the anticoagulant group (OR 0·346, 95% CI 0·006–4·390; p=0·66). There was one subarachnoid hemorrhage in the anticoagulant group. All stroke events occurred in the first 10 days after randomization. There were no deaths in the total population. In the intention to treat population, ipsilateral TIA occurred in 1% of the antiplatelet group vs 3% of the anticoagulant group.

So there was no difference in efficacy of antiplatelet and anticoagulant drugs in preventing stroke and death. The overall risk of stroke was low.

The study might have not have enrolled a small high risk group, those with early recurrent strokes (Kasner SE. CADISS: a feasibility trial that answered its question. Lancet Neurol. 2015 Apr;14(4):342-3). Any future trials of newer oral anticoagulants and to detect small effect size may require 5000 participants and require 500 sites and 10 years to complete recruitment.

An Orthostatic Headache

This is a 41 year old male with no prior headache history who developed a back of the head throbbing with an intensity of 9-10/10 when upright and 1/10 when supine associated with nausea and vomiting. Past medical history was negative. Neurological examination was normal.

Question: What are the causes of orthostatic headaches?
The most common cause of an orthostatic headache is a cerebrospinal fluid (CSF) leak, most commonly iatrogenic after a dural puncture. Other than iatrogenic, CSF leaks can be traumatic or spontaneous.

Spontaneous intracranial hypotension (SIH) has an estimated annual incidence of 5/100,000 with a female to male ratio of 2:1. The peak incidence is around the age of 40 years. SIH is usually due to a spontaneous CSF leak at the thoracic or cervicothoracic junction. A MRI of the brain with contrast may demonstrate diffuse dural enhancement in 80% of cases. In some cases, the location of the leak cannot be located despite extensive testing. A minority of patients may have an underlying connective tissue disorder such as Marfan's or Ehlers-Danlos but the cause in most is not known.

There are other causes of orthostatic headaches (Mokri B. Spontaneous low pressure, low CSF volume headaches: spontaneous CSF leaks. Headache. 2013 Jul-Aug;53(7):1034-53). Orthostatic headache can be the most prominent feature of postural orthostatic tachycardia syndrome (POTS) (Mokri B, Low PA. Orthostatic headaches without CSF leak in postural tachycardia syndrome. Neurology. 2003;61:980-982). Occasionally following decompressive surgery for Chiari malformation, an orthostatic headache may occur with a CSF leak.

Following large decompressive craniectomies for life-threatening cerebral edema, orthostatic headaches may occur which can improve with cranioplasty (Mokri B. Orthostatic headaches in the syndrome of the trephined: Resolution following cranioplasty. Headache. 2010;50:1206-1211). Individuals with increased compliance of the dural sac (especially with large lumbar dural sacs and stigmata of connective tissue disease) may develop orthostatic headaches (Leep Hunderfund AN, Mokri B. Orthostatic headache without CSF leak. Neurology. 2008;71:1902-1906). Finally, orthostatic headaches can be present with colloid cysts of the third ventricle (Spears RC. Colloid cyst headache. Curr Pain Headache Rep. 2004;8:297-300) and a supratentorial meningioma (Smith RM, Robertson CE, Garza I. Orthostatic headache from supratentorial meningioma. Cephalalgia. 2/9/15 [online before print].

Neuroimaging: A MRI of the brain showed diffuse meningeal enhancement. MRI of the spine showed a small ventral epidural CSF collection at the C8-T1 level. Past medical history was negative.

Questions: Are patients without Ehlers-Danlos or Marfan's at risk for vascular abnormalities?

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Schievink and Deline (Schievink WI, Deline CR. Headache secondary to intracranial hypotension. Curr Pain Headache Rep. 2014;18(11):457) recommend screening for two vascular abnormalities which occur with a higher frequency in those with SIH than in the general population, intracranial saccular aneurysm and aortic root dilatation, based upon single studies.
MRA scans of the brain were performed in 93 patients (mean age 42.2 years, 70% females) with SIH finding 8.6% intracranial saccular aneurysm (mean age 51.4 years (Schievink WI, Maya MM. Frequency of intracranial aneurysms in patients with spontaneous intracranial hypotension. J Neurosurg 2011;115:113-5). None of the patients with aneurysms had a recognized systemic connective tissue disorder or family history of aneurysm. In a control population of 291 patients (mean age 54.8 years, 56% females), 1% had intracranial aneurysms. Study limitations included not obtaining MRA scans on all patients with SIH and not matching study and control patients for important risk factors.

In a study of 50 consecutive patients with SIH aged 12-67 years (Pimienta AL, Rimoin DL, Pariani M, Schievink WI, Reinstein E.Echocardiographic findings in patients with spontaneous CSF leak. J Neurol. 2014 Oct;261(10):1957), 6 had aortic root dilatation (5 without connective tissue disorders). The authors recommend that patients with SIH without a connective tissue disorder may benefit from baseline echocardiographic screening. For those with evidence of a dilated thoracic aorta, they suggest a yearly follow-up for the first 3-4 years to determine the risk of progressive and rupture. For those with an initial normal study, follow-up exams in two and five year intervals. They also recommend confirmation of their findings in larger cohorts.

National MS Society designates Dr. Ann Bass as a Partner in MS Care

The Neurology Center of San Antonio's Dr. Ann Bass has earned the designation of a Partner in MS Care as a result of her specialization in and commitment to multiple sclerosis. Partners in MS Care is a National MS Society program that recognizes and supports quality MS care, encourages strong partnerships between MS Clinics and the Society to create optimal care and support for people living with multiple sclerosis.

Dr. Bass completed her neurology residency in 1997 at the University of Texas Health Science Center in San Antonio. Dr. Bass has specialized in multiple sclerosis since 2000, and has more than 1,000 patients living with MS under her neurological care and management. She serves on the National MS Society, South Central Clinical Advisory Committee, and has been a principle investigator in many multi-center clinical trials for the treatment of multiple sclerosis. In 2001, Dr. Bass furthered her commitment to people affected by MS by establishing a comprehensive MS clinic that includes an infusion center.

Other Partners in MS Care are the following: Texas Institute for Neurological Disorders, Sherman; Baylor Institute for Rehabilitation, Dallas; University of Texas Southwestern Medical Center at Dallas; Amarillo Diagnostic Clinic; Maxine Mesinger Multiple Sclerosis Comprehensive Care Center, Houston; Pediatric MS Clinic at Texas Children's Hospital; Bellaire Neurology; University of Texas Health Science Center, San Antonio; The Multiple Sclerosis Treatment Center of Dallas; Kane Hall Barry Neurology, Bedford and Keller; MS Clinic of Central Texas/Central Texas Neurology Consultants, Round Rock; and The University of Texas Health Science Center at Houston.

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TNS 18th Annual Winter Conference Poster Winners

1st Place-	*Matthew Freeman, MD*
2nd Place-	*Ahmed Yassin, MD*
Tie 3rd Place-	*Divyanshu Dubey, MD*
	and Jason Thonhoff, MD

Still Alice: A Wonderful Movie with Superb Acting That Sends the Wrong Clinical Message

By Ronald Devere MD FAAN
Director Alzheimer's Disease and Memory Disorders Center – Austin, Texas

Shortly before the Academy Awards I had the opportunity to see the movie, Still Alice, at a private showing with about one hundred care givers of Alzheimer's patients, as well as patients with mild cognitive impairment and early stage Alzheimer's disease in attendance. This event was sponsored by the Capital of Texas Chapter of the Alzheimer's Association in Austin, Texas. The story line was heart felt and the acting superb, portraying all the love and care that occurs in reality. Julianne Moore, as the Alzheimer's patient, Alice, certainly deserves her Academy Award for Best Actress. However, based on many audience questions after the movie, I believe the clinical message portrayed was incorrect, and only added more fear of the disease to an already fearful public.

As presented, Alice is a PhD linguistic professor at Columbia University who has just celebrated her 50th birthday. In the early part of the movie she begins to have memory problems and word finding difficulty, and also begins getting lost on her regularly frequented jogging trail nearby the University. Then she decides on her own to seek medical help and visits a physician (not stated, but presumably a neurologist) for a medical opinion. During the office cognitive evaluation she was asked six or seven very basic questions that suggested cognitive impairment. Nothing is mentioned about further tests being ordered and/or reviewed at the follow-up visit. At the follow-up visit she is told that the MRI of her brain and her blood work were normal. Two additional tests that had been ordered were reviewed. One was a genetic panel of blood tests for familial Alzheimer's disease, which was positive. This is usually to detect dominant genes such as Presenilin 1 and 2 on chromosomes 14 and 1. Her test and that of the one daughter who was tested were positive for one of the genes. She also had an amyloid PET scan that was positive for high levels of amyloid in the brain and along with the positive genetic test compatible with familial Alzheimer's disease. My concerns are twofold: the methods by which the diagnosis was obtained, and how Alice came to seek out her physician. Both require clarification and send the wrong message.

1. Alice began having memory problems early on and that was followed by her getting lost on her usual jogging trail. Ostensibly, her family members were unaware of any cognitive changes until she told her husband that she had Alzheimer's disease. This is very unusual for patients with mild cognitive impairment or Alzheimer's because family members are usually the first to recognize this problem. More than ninety percent of patients who seek medical attention for memory changes go to see a doctor because the care givers are worried and recognize the problem before the patient.

2. In the movie the cognitive testing consisted of only five to six questions, which is well below the standard for in-office cognitive testing. I understand that the movie is not going to show an entire cognitive evaluation, but they could easily have mentioned that more detailed testing was done, such as neuropsychological testing.

3. Alice's family history for Alzheimer's disease suggested that only her father possibly had dementia. Clinically, it would not be the standard to order detailed genetic testing in a fifty-year-old patient with one family member with Alzheimer's disease, and without some very basic information from the treating physician about how very rare these genetic mutations are in the population. The familial form of Alzheimer's disease makes up less than 0.5 percent of all people with the disorder.

4. Amyloid PET scans are a relatively new test and are very helpful in the diagnosis of dementia. However, they are not approved by any insurance plan, including Medicare, and they cost over $5000 out of pocket. Currently they are done mostly at research centers for those who are willing to pay for them, but that is very infrequent. Medicare covered testing for the degenerative dementia usually includes FDG PET scans and spinal fluid bio-markers, and it is also usually covered by private insurance carriers.

After the movie I was asked numerous questions from the audience, as well as in my practice about ordering genetic testing for Alzheimer's regardless of the age and family history. They also wanted to know when and where they could have the amyloid PET scan done. I realize the movie was based on the 2007 novel by Lisa Genova, but the script writers and producers should have made every attempt to check the facts about a disease that is so prevalent and is causing such an epidemic of fear. Even putting a disclaimer on the screen stating the content as related in the movie may not have been entirely accurate or had been dramatized would at least help to reduce some of the fear of getting Alzheimer's disease.

Advocacy Update – May 2015

By Sara Austin, TNS Legislative Chair and Greg Herzog, TNS Lobbyist

Yippee! Whooo hoo!, and Wowza! THE SGR IS FIXED! Yes, hard to believe but very true, Congress actually worked together to pass the bill that took the SGR off the table completely and the bill was signed by the President on 4/15. Dr. Michael Burgess (R- TX Denton) wrote the bill and gets most of the credit for getting it through. I'm not sure exactly what went right this time except that the policy seemed to be something they could support (and maybe they ran out of credibility blaming each other). Senator Cornyn did his share of heavy lifting to make sure it passed so we want to give him credit as well. There is a small update now to the Medicare fee schedule every year for the next 4 years (.5%) and then starting in 2020 there will be bigger updates for those of us using alternative payment methods. It's not clear exactly what those are yet, so stay tuned. And yes, that makes us nervous, but at least we won't be facing a 25% reduction every year.

This has been a busy year for the house of medicine in the Texas Legislature. I know that some of you have been to First Tuesdays, thank you. Contacting your legislator's office about certain bills is also very helpful. The TNS has a great advantage now that we have our own lobbyist. Greg Herzog has been so helpful following issues and alerting us to bills that are coming up that might affect us. In addition, he has relationships with many of the legislators and can arrange meetings when needed. We have not had any of that in the past and believe me, it has really helped.

The TNS weighed in on CBD (cannabis oil) for epilepsy in a hearing. Our position that more research should be allowed was well received. Both the AAN and TMA are asking that marijuana be reclassified as a Schedule II drug on the federal level so that it is easier for studies to be conducted.

Greg and Dr. Kim Monday have been working on a bill to license neurodiagnostic technologists. As filed, the legislation was lacking. TNS has worked with the bill author, State Representative Greg Bonnen, MD to make several important improvements. The NDT licensure body would be housed at the Texas Medical Board. NDTs would not have direct access to patients or independent use of needle EMGs. TNS will remain very vigilant as the bill moves through the process to ensure that the agreed to language stays on the bill.

I have been working on the Advanced Directives work group for the TMA's Council on Legislation. We have a work group that has been talking frequently and working through bills that were filed. Probably at least one bill will get thru this year pertaining to artificial nutrition and hydration, but hopefully no more than that. It is important that you educate your legislators about end of life issues because there are some people out there with some very weird ideas – I'm thinking that a little education from physicians who see it every day would help.

Even in the land of plenty, which is the Texas budget this year, Medicaid remains severely underfunded. The House budget added $460M to extend the primary care bump to Medicare parity, the Senate is not even considering it as best we know. The specifics will be worked out in conference and we are talking to the conference members reminding them how difficult it is for Medicaid patients to access adult and pediatric neurologists. Our TNS lobbyist, Greg Herzog, was able to ask Dr. Greg Bonnen to push a rider to the budget that at least would let them consider adding Neurology to the bump. Cross your fingers.

GME looks to be pretty well funded this session which we are happy with. Scope is always an issue and there are some bills that are winding their way through, but things are moving slowly and so the chances of bills we don't like getting thru are getting smaller with each passing day.

As always, we encourage you to make a connection with your legislator (and to let us know if you are able to do that). Stay involved, your opinion is respected and is needed.

It's That Time Again....

Make time on your schedule for the Texas Neurological Society Summer meeting! The meeting will be in Fort Worth on Friday, July, 31 2015 through Sunday, August 2, 2015 and has been expanded to three days to include general neurology presentations as well as a joint meeting of the Southern Headache Society. Headache presentations will be open to TNS members with appropriate CME hours, as well. Brian Loftus and Michael Ready are the program directors, and have done an excellent job in organizing this joint meeting – giving us the opportunity to take advantage of the headache talks.

The Sheraton Fort Worth is an excellent hotel with wonderful accommodations. Start planning now for another good summer program.

Register today at www.texasneurologist.org

New Committee Formed to Help TNS Physicians

By Kristi Berrier, TNS Medical Economics Consultant

At its July 2014 meeting, the TNS Board of Directors had a lengthy discussion regarding the possibility of forming a specialty IPA to help members navigate the complex landscape of managing a neurology practice in Texas. Subsequently, the IPA Subcommittee of the Medical Economics Committee was convened in September to explore the possibility of forming a specialty Independent Physician Association (IPA). After extensive discussions regarding the costs and benefits of creating an IPA, the subcommittee decided to table the IPA concept for possible re-exploration at a later date.

As an alternative to developing an IPA, it was decided to form a subcommittee to the Medical Economics Committee called the "Medical Economics Resources Committee" (MERC). The committee will serve as a networking resource to help TNS members find solutions to medical economics issues they may be facing. These solutions might come from fellow TNS members, TNS staff, AAN staff or the TMA. The AAN has expressed an interest in using this committee and its work as a possible template for improving communications with members in other state Neurological Societies.

A gap analysis conducted by the AAN examined areas of concern for neurologists across the country. Those issues included: ACOs; episodes of care/bundled payments; value based payment modifiers; PQRS; Coding; MOC and adding Advanced Practice Providers (Nurse Practitioners and Physician Assistants). These are some of the issues the committee will be focused on with intent of assisting our TNS members.

It is apparent from a recent announcement by CMS that these issues are highly relevant for all physicians. On January 26, 2015, CMS announced goals for the continued implementation of a value-based payment model to replace the existing fee for service model. In the announcement, CMS identified four categories of health care payment:

- Category 1 – fee-for-services (FFS) with no link of payment to quality
- Category 2 – fee-for-service with a link to quality (e.g. Physician Value-Based Modifier)
- Category 3* – alternative payment models build on fee – for-service architecture (e.g. ACOs, Bundled payments)
- Category 4* – population-based payment (e.g. Pioneer and New Generation ACOs)

* - Considered alternative payment models by CMS

CMS has set a goal to have 85 percent of all Medicare reimbursements in categories 2-4 by 2016, with an increase to 90 percent in 2018 with at least 50 percent of them being alternative payment models (categories 3-4). It is important for physicians to understand that CMS is not implementing the shift away from traditional fee-for-service and toward value-based payment in a vacuum. It is working with other stakeholders, including private payers and large employers to accelerate the transition, not just for Medicare and Medicaid patients, but for those insured by commercial payers as well. As explained in the announcement:

> *"When providers encounter new payment strategies for one payer, but not others, the incentives to fundamentally change are weak. In fact, a provider that alters its system to prevent admissions and succeed in an alternative payment environment may lose revenue from payers that continue fee-for-service payments."*
>
> *~ January 26, 2015 CMS Media Release*

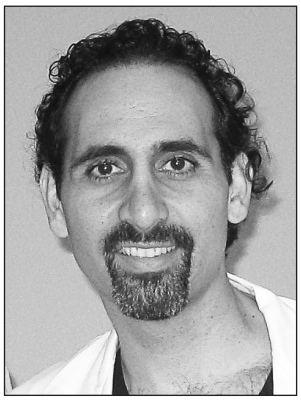
Some TNS members have not participated in the EHR Incentive Program/Meaningful Use or PQRS, reasoning that they have a relatively small or even no Medicare patient population. These members believe that, by avoiding Medicare they may be avoiding the transition from fee-for-service to value-based payment systems. However, based upon the literature and CMS reporting, in the near future there will be a transition when Medicare is not the only payer requiring physicians to provide quality data and implement cost control measures in order to receive full reimbursement. The transition won't happen overnight - it takes time to test and evaluate reimbursement and reporting models while identifying the accepted methods for benchmarking and risk adjustment. Nevertheless, private payers will be following in the footsteps of CMS; sooner rather than later.

The Medical Economics Resource Committee is committed to helping Texas neurologists face the multitude of impending changes with confidence; while focusing on our mission of providing excellent neurological care for our patients. The committee has created answers to frequently asked questions on medical economics issues that can be found on the TNS website. TNS members can submit additional questions to the committee via an email link provided on the TNS website. The TNS Medical Economics Resource Committee members include: Stuart Black (Chair), Robert Leroy, Kim Monday, Sarah Austin, William Gilmer, Randy Evans, Gary Tunnell, Deborah Carver, Erin Furr and Tommy Yee.

"When providers encounter new payment strategies for one payer, but not others, the incentives to fundamentally change are weak."

Chiasmal Syndromes

By Rod Foroozan, MD
Neuro-Ophthalmologist
Associate Professor of Ophthalmology and Neurology
Baylor College of Medicine



Case Presentation

A 35 year-old woman developed the sudden onset of headache and blurred vision. She saw an eye doctor and was noted to have 20/20 visual acuity in each eye and full visual fields to confrontation . Extraocular motiltiy and ocular alignment were normal. Funduscopy was normal in each eye, with no evidence of optic disc edema.

Over the next 48 hours the headache and blurred vision worsened. She went to an emergency room and computed tomgrahy (CT) with axial views and without contrast of the head was said to be normal. Optic neuritis was considered and it was suggested that she see a neuro-opthalmologist.

Two days later visual acuity was 20/20 in each eye and color vision was normal in each eye. The pupils were brisk with no relative afferent pupillary defect. Automated perimetry showed temporal defects in each eye (Figure 1). An MRI of brain with contrast was done urgently and showed a sellar/suprasellar mass with multiple signal intensities suggestive of hemorrhage into a pituitary adenoma and consistent with pituitary apoplexy (Figure 2). She was sent to the emergency room for urgent neurologic and neurosurgical consultation.

Discussion

The optic chiasm is an important neuro-anatomic landmark. Pathology that affects the chiasm helps emphasize key principles of neuroanatomy. The most common visual consequence of pituitary tumors is chiasmal compression, and visual findings remain important indicators of central nervous system pathology in many patients with sellar/suprasellar lesions.

Anatomy

The intracranial optic nerves exit the optic canals and travel at a 30- 45 degree angle to make up the optic chiasm. The optic chiasm is 12-18 mm in diameter and contains the decussation of nasal nerve fibers from each of the optic nerves. 1 Inferior retinal fibers representing the superior visual field cross first, while macular fibers cross more posteriorly. The chiasm lies 10 mm above the sella turcica, which contains the pituitary gland.

The space inferior to the chiasm and above the sella is the basal cistern. Anteriorly, the chiasm is exposed to cerebrospinal fluid (CSF); posteriorly it abuts the hypothalamus and the anterior portion of the third ventricle. The individual variation in relative position contributes to clinical variation in the presentation of chiasmal syndromes.

Presenting symptoms and signs

A thorough eye examination (including visual acuity, external examination, pupils, intraocular pressure, ocular motility, and examination of the anterior and posterior segments) should be considered in all patients suspected of having a chiasmal disorder. Color vision should also be tested to detect subtle defects of central visual function. Examination of the visual field, preferably with automated or kinetic perimetry, can provide critical clinical information, and may provide the initial indication of pathology affecting the optic chiasm.

Visual loss

Visual acuity and color vision may be abnormal in patients with suspected chiasmal disease and a visual disturbance. Macular neurons make up 30% of the chiasm; therefore, many chiasmal disorders result in central visual field defects or disorders of central vision, such as color perception abnormalities.2 However in more subtle and incomplete chiasmal syndromes central visual function is initially spared.

Visual field testing is often the most important clinical tool in localizing pathology to the chiasm.2 Patterns of visual field defects include:

- Bitemporal hemianopia/quadrantanopia
- Unilateral central defect with contralateral hemianopia or temporal visual field loss
- Homonymous hemianopia

Bitemporal hemianopia is the classic finding in a patient with a compressive chiasmal lesion; however, the field defects can be subtle because the deficits need not be complete. Given individual variation in the position of the chiasm as well as the growth pattern of the lesion, it is important to remember that a patient with a lesion compressing the chiasm may not have the classic bitemporal hemianopic field defect.3,4 Depending on variations of anatomy there may be more involvement of the optic nerve, causing monocular visual field loss characteristic of other optic neuropathies. Involvement of the more posterior portion of the chiasm may cause temporal defects which are smaller, around fixation, and with extension to involve the optic tract, may lead to a homonymous hemianopia.

Optic nerve involvement in chiasmal disease can result in an ipsilateral central defect with contralateral temporal defect, a "junctional scotoma." A unilateral temporal visual field defect may also be the first sign of a chiasmal syndrome, so that the finding of preferential loss of the temporal visual field suggests that pathology involves the optic chiasm until proven otherwise.

Diplopia

Diplopia is caused by two mechanisms in a patient with a chiasmal lesion. First, diplopia can result from involvement of the cavernous sinus and cranial nerves caused by lateral expansion of a pituitary tumor (such as with pituitary apoplexy) or other lesions such as carotid aneurysms. A second mechanism causing variable vertical or horizontal diplopia due to disruption of latent heterophorias is known as hemifield slide. Hemifield slide occurs in patients with

dense bitemporal defects as images formed on corresponding portions of each retina fall into the affected nasal retina, and therefore are not seen. 2 In some patients treatment with optical measures such as prism can be helpful to reduce the frequency of diplopia.5

Optic disc pallor and cupping

Classically, optic disc pallor from a compressive chiasmal lesion is in a band pattern that spares the inferior and superior portions of the disc. It should be noted that optic disc pallor indicates a more chronic process and may be subtle.1 The absence of optic disc pallor should not preclude an evaluation for a chiasmal disorder in a patient with suggestive clinical findings.

Some patients with compressive lesions involving the anterior visual pathways may develop non-glaucomatous optic disc cupping. Optic disc cupping in a band pattern has been noted in chiasmal compression.6

Diagnostic studies

Neuroimaging

Neuroimaging should be considered in any patient with a newly diagnosed bitemporal hemianopia. MRI is typically the most useful ancillary study when evaluating chiasmal disease. It both localizes and characterizes chiasmal pathology in order to determine the diagnosis.7 A CT scan with contrast may be of some value, especially in emergent settings or in patients that have contraindications to MRI. Imaging with CT may also reveal acute blood, bony changes of the sella, and calcium which are poorly seen with MRI. In most patients, MRI is better suited to assess the soft tissue of the intracranial optic nerves, the chiasm (including contrast enhancement and thickening), and the area around the sella. Diffusion tensor imaging may be helpful to assess the visual pathways when standard sequences are not revealing.

It is important to provide the neuroradiologist with clinical details and concerns for chiasmal pathology. Depending on the duration and type of the lesion, in some patients the neuroimaging findings may not completely predict the pattern and severity of visual loss.

Optical coherence tomography

Optical coherence tomography (OCT) has become a valuable tool to evaluate retinal nerve fiber layer (RNFL) thickness in glaucoma and in patients with other types of optic neuropathy.9,10 Because it is not dependent on responses from the patient, OCT still may be completed in patients who are not able to perform perimetry.

There is evidence that OCT may be a useful predictive tool to assess potential visual field recovery in patients undergoing pituitary tumor resection.11 One study noted that in patients with visual field defects but relatively normal RNFL on OCT, visual recovery following decompressive surgery was better than in those patients with attenuation of the RNFL on OCT.10 Correlations with structure and function in chiasmal dysfunction have continued to improve.12

Etiologies and clinical approach to chiasmal disease

Most diseases that affect the chiasm can be broadly placed into two categories, those that are intrinsic to the chiasm and those that are extrinsic, summarized in the Table. These include potentially life threatening (including apoplexy, aneursym, and infection) and blinding conditions. While neuroimaging will typically distinguish the most likely causes, other ancillary testing, such as lumbar puncture may be helpful in some patients. In other patients tissue biopsy may be necessary to secure a diagnosis.

Table: Etiologies of Chiasmal Disorders, modified and reprinted with permission from Foroozan, R. Chiasmal syndromes. *Curr Opin Ophthalmol.* Dec 2003, Vol. 14, 6, pp. 325-31.

Congenital	Traumatic	Iatogenic	Intrinsic Lesions	Extrinsic lesions
Albinism	Motor vehicle accident	Radiation	Glioma	Pituitary adenoma
Achiasmia	Skull fracture	Surgical injury	Demyelination	Craniopharyngioma
		Fat packing	Chiasmal inflammation/ infiltration	Meningioma
		Empty sella	Ganglioma	Aneurysm
		Dopamine agonists	Cavernoma	Mucocele
			Histiocytosis	Lymphocytic hypophysitis
			Ischemia	Hydrocephalus
			Infection	Arachnoid cyst
				Epithelial cyst
				Dysgerminoma
				Metastasis
				Abscess/Mycotic aneurysm

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Figure 1: Automated perimetry showed temporal defects in each eye.

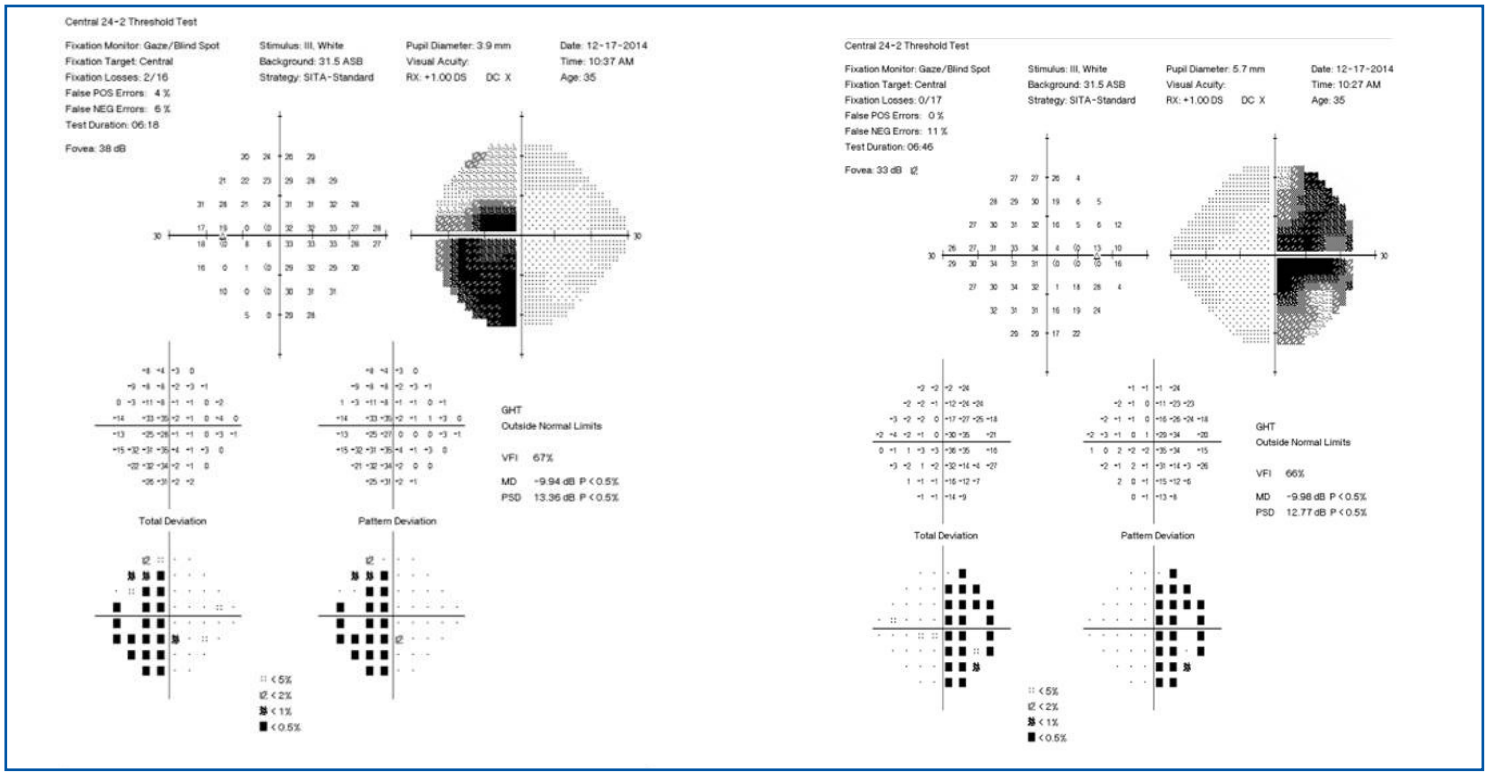
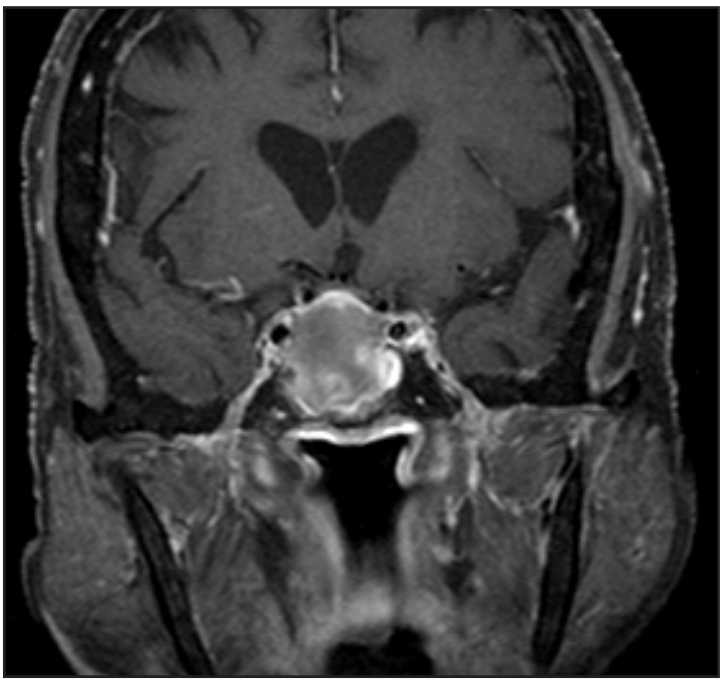


Figure 2: An MRI of brain with contrast showed a sellar/suprasellar mass with heterogeneous signal intensities suggestive of hemorrhage into a pituitary adenoma and consistent with pituitary apoplexy.



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Victory! Congress Permanently Repeals SGR

By Mike Amery, AAN Senior Legislative Counsel, Federal Affairs

The US Senate took the final step in permanently repealing the flawed Sustainable Growth Rate formula on a 92-8 vote Tuesday.

Republicans and Democrats each offered three amendments, but all failed as the Senate passed language identical to the bill passed as H.R. 2 by the US House last month. It ended a series of 17 temporary patches since 2002.

The Medicare Access and CHIP Reauthorization Act of 2015 begins to fundamentally change the way Medicare will operate in the future. It is important to understand that the bill is a starting point for changing the system in a way that improves physician participation in Medicare to ensure patient access to care.

The AAN, along with the entire physician community, supported this legislation because it's an improvement compared to the status quo. The bill is not perfect, but it does contain modest payment updates that are a stark contrast to a 21-percent cut or the prospect of Congress repeatedly using the physician community as the offset for future patches, like what occurred in 2014. The bill consolidates several burdensome programs (PQRS, MU, and VBPM) into one streamlined program that will set more realistic benchmarks for neurology practices and funds bonuses for those who perform well. The bill also preserves the fee-for-service option, which has been the desire of many solo and small group neurology practices around the country.

The AAN worked with members of Congress and their staff throughout the process of developing this bill. We now look forward to fully participating in the implementation of H.R. 2 to protect the interests of America's neurologists and the patients they serve. Just remember, this is not the end, but rather the beginning. Many of the details remain to be written, but a starting outline follows:

The Medicare Access and CHIP Reauthorization Act of 2015 (H.R. 2)

H.R. 2 is the result of a multi-year, bipartisan, bicameral process representing the most significant change to US health care policy since the Affordable Care Act. It repeals the SGR formula and enacts widespread changes to the way Medicare pays physicians by transitioning from a volume-based to a value-based system over the next decade. Below are highlights on H.R. 2:

SGR repeal: Effective immediately, the sustainable growth rate (SGR) formula has been permanently repealed. This ends the current and all future payment cuts called for by this faulty formula.

Fee updates: This legislation provides 4.5 years of 0.5 percent annual updates, through 2019. Then, from 2020 through 2025, updates are frozen at 2019 rates. For 2026 and beyond, two conversion factors will take effect:

- 0.75 percent for participants in qualifying alternative payment models
- 0.25 percent for everyone else

Merit-based Incentive Payment System (MIPS): Starting in 2019, Medicare's key quality initiatives—PQRS, Meaningful Use, and the Value-based Payment Modifier—would be consolidated into a new program, MIPS. Conversion to a single program means physicians will have more options to meet requirements and a streamlined way to track performance for all Medicare quality initiatives.

- Payment adjustments: Under MIPS, physicians will have the opportunity to earn bonuses in excess of those available under current law and would face lower penalties.
- Improved ways to demonstrate quality: Physicians will have more flexibility to select criteria they wish to report on and receive credit for improving as well as hitting performance targets.
- Technical assistance for small and rural practices: From 2016 to 2020, $100 million will be available to help practices of up to 15 professionals and rural practices to participate in MIPS or transition to an alternative payment model.

Alternative payment models (APMs): Participants in a qualifying APM are exempt from MIPS and are eligible for a 5-percent annual bonus from 2019 to 2024. The bill provides support for the development of new APMs, including those for specialty physicians.

Chronic care management: Through at least one payment code, Medicare will be required to pay for care management of patients with chronic conditions. Payment of this code will not require an annual wellness visit or an initial preventive physician exam.

Electronic health records (EHR): The bill sets a goal of interoperability of EHR systems by December 31, 2018. Should this not happen, the Health and Human Services Secretary is given discretion to adjust the penalties for Meaningful Use and/or decertify EHRs.

Quality measure development: From 2015 to 2019, $15 million per year will be available to stakeholders such as the AAN to support quality measure development.

Children's Health Insurance Program (CHIP): Funding is extended through fiscal year 2017.

Global periods for surgical services: This bill reverses the decision by the Centers for Medicare & Medicaid Services to phase out the 10- and 90-day global period

Recent Developments in Headache Medicine

By Deborah I. Friedman, MD, MPH
Professor, Neurology & Neurotherapeutics and Ophthalmology

New therapeutic targets and neuromodulation treatments for migraine and cluster headache, and completion of a clinical trial investigating the treatment of idiopathic intracranial hypertension highlight the major recent advances in the field of Headache Medicine.

Anti-calcitonin gene related peptide (CGRP) antibodies

CGRP is an attractive candidate for migraine therapy. Stimulation of the trigeminal ganglion in humans leads to release of GCRP from perivascular sensory axons to mediate vasodilation of extracerebral intracranial arteries and arteriovenous anastomoses. CGRP is also involved in pain transmission within the trigeminal system in the CNS. CGRP levels are elevated in jugular venous blood samples during migraine, and sumatriptan reduces levels of CGRP in experimental animal models of migraine. If ultimately proven safe, effective and marketed, these treatments would herald an era of migraine preventive therapy using medications specifically designed for this specific purpose. The results of two phase 2, randomized trials of CGRP antibodies for episodic migraine were presented at the 2014 American Academy of Neurology annual meeting:

- ALD403 (Alder Pharmaceuticals), administered as a single intravenous infusion, was compared to placebo infusion in 163 patients {Dodick, 2014 #1830}. At 24 weeks, those receiving active treatment had 5.6 fewer migraine days per month (a 66% decrease) vs. 4.6 fewer days for those receiving placebo. Sixteen percent of participants receiving ALD403 had no migraine days at 12 weeks (no one in the placebo group became headache free). The treatment was well tolerated with comparable side effects between the two groups.

- LY2951742 (Arteus Therapeutics) was compared to placebo in 217 patients who received biweekly subcutaneous injections for 12 weeks {Dodick, 2014 #1831}. Those receiving active treatment had 4.2 fewer migraine days per month at 12 weeks (a 64% decrease) vs. 3 fewer days in the placebo group (a 42% decrease). There were more side effects with active treatment, such as injection site pain, upper respiratory infections and abdominal pain) but overall, the medication was safe and well tolerated.

Clinical trials of a CGRP receptor antibody are underway for acute and chronic for cluster headache.

Neuromodulation

Transcutaneous supraorbital nerve stimulation

Transcutaneous supraorbital nerve stimulation (Cefaly®) was approved by the Food and Drug Administration (FDA) for prevention of migraine with and without aura. A randomized, double-blinded, sham-controlled study (PREMICE) of 67 patients was reported in 2013 [Schoenen]. Subjects underwent daily sham or verum neurostimulation for 20 minutes. The primary outcome was change in monthly migraine days and the 50% responder rate. The therapeutic gain of the device was 26%, separating from placebo stimulation after the first month. By month 3, the mean number of migraine days decreased by 6.94 to 4.88 days, compared to 6.54 vs. 6.22 in the sham group). Monthly migraine attacks, headache days and acute antimigraine drug intake were also statistically significantly decreased in the active group but not the sham group. There was no effect in pain severity or associated migraine symptoms. 70% of participants were "very or moderately satisfied" and the device was well tolerated.

Implanted occipital nerve stimulation

A randomized, double blind, multicenter study of 157 patients was performed to assess implanted occipital nerve stimulation (ONS) [Dodick DW, Silberstein SD, Reed KL, et al. Safety and efficacy of peripheral nerve stimulation of the occipital nerves for the management of chronic migraine: Long-term results from a randomized, multicenter, double-blinded, controlled study. Cephalalgia 2014 pii: 0333102414543331]. 125 patients had chronic migraine. Subjects were randomized to active or control groups for 12 weeks, then received open label active stimulation for 40 weeks. Outcomes were assessed at 52 weeks. Headache days were reduced by 6.7 + 8.4 days overall and 7.7 + 8.4 days in the chronic migraine group. More than half of patients were satisfied with treatment. However, there were 183 device or procedure-related adverse events; 8.6% required hospitalization, 40.7% required surgery and 70% of subjects had some type of adverse event.

External vagal nerve stimulation (VNS)

The results from two studies were presented at the European Headache and Migraine Trust International Congress in 2014:

- The Prevention of Chronic Migraine (EVENT) Study (Electrocore) randomized subjects to receive either VNS or sham treatment (low-voltage device generating an electric field and audible feedback) administered 3 times daily at specified time intervals. Each treatment was two 90-second stimulations administered 5-10 minutes apart [Schoenen J, Gaul C, Silberstein S. Presented at the 4th European Headache and Migraine Trust International Congress, Copehagen, September 20, 2014]. The primary endpoint was safety and tolerability with secondary clinical efficacy endpoints. The average number of headache days per 28 days was 21.1 in the active group and 22.3 in the sham group. At month 2, VNS reduced headache days by a mean of 2 per 28 days with no change in the sham group. At month 8, there was an average reduction of 9 days in the active group compared to 6 in the sham group, although the number of participants in the trial decreased by half. Longer duration of treatment was associated with fewer migraine days per month. A US study is in progress.

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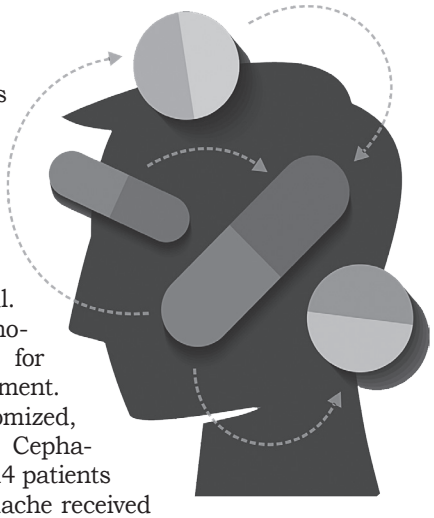

- Non-invasive VNS was studied in the PREVA (Prevention and Acute Treatment of Chronic Cluster Headache) study (Electrocore)] Schoenen J, Jensen RH, Lantéri-Minet M, et al. Stimulation of the sphenopapatine ganglion (SPG) for cluster headache treatment. Pathway CH-1: A randomized, sham-controlled study. Cephalalgia 2013;33:816-30.] 114 patients with chronic cluster headache received standard of care treatment for 2 weeks, and were randomized to standard of care (SOC) or SOC plus VNS for 4 weeks. In the third phase, all participants received SOC plus open-label VNS. The primary end point was reduction of CH attacks weekly in the last 2 weeks of the randomized phase. The mean number of CH attacks in the 4 weeks prior to enrollment was 67.3 in the SOC + VNS group and 73.9 in the SOC group. About 70% of subjects used oxygen for an acute attack. There was a statistically significant reduction in CH attacks in the VNS group compared to SOC alone (-7.6 vs. -2.0, p=0.002). After the transition to open label, the group initially assigned to receive SOC alone had a reduction of 3.3 CH attacks per week (p=0.0001) and the VNS group had an addition improvement of 1.9 attacks per week (p=.0032). 37.8% of subjects randomized to VNS had at least a 50% response rate vs. 8.3% randomized to SOC. As in the migraine study, longer duration of treatment during the study was associated with a continued reduction in CH attacks. There were no serious adverse events associated with treatment. A US study was recently completed and results are pending.

Sphenopalatine Ganglion Stimulation

The SPG is located in the pterygopalatine fossa and contains sympathetic and parasympathetic fibers. Low frequency SPG stimulation can induce cluster attacks which are aborted with high frequency stimulation. An implanted SPG stimulator with a wireless, rechargeable, remote control system is in clinical trials by Autonomic Technologies Inc. The device is inserted via trans-gingival approach by an oromaxillofacial surgeon.

A European study (CH-1) of refractory chronic CH was performed in 32 subjects with full stimulation, sub-stimulation or sham stimulation [Schoenen J, Jensen RH, Lantéri-Minet M, et al. Stimulation of the sphenopapatine ganglion (SPG) for cluster headache treatment. Pathway CH-1: A randomized, sham-controlled study. Cephalalgia 2013;33:816-30]. 28 participants completed the study. The primary outcome was pain relief in 15 minutes or a serious adverse event. 67.1% of patients receiving full stimulation had pain relief vs. 7% of those receiving the other stimulation paradigms. 68% had clinically significant improvement, 25% had pain relief in at least 50% of attacks and 35% had at least a 50% reduction in attack frequency. Recruitment for a similar study is underway in the U.S. and a larger trial for migraine is being conducted in Europe [Nagy AJ, Rapoport AM. Update on future headache treatments. Neurol Sci 2013;34(suppl 1): S101-108].

New Sumatriptan Delivery Systems

- A breath-actuated delivery of powdered sumatriptan using the OptiNose™ device (AVP-825, Avanir) was tested in a phase 3 placebo-controlled, parallel group study of 212 participants. Active treatment was 15 mg of intranasal sumatriptan. Pain relief in 2 hours was achieved in 68% of patients receiving active treatment vs. 45% of controls (p<0.01), with pain relief in 30 minutes in 42% of participants taking sumatriptan vs. 27% of controls (p<0.05).

- Iontophoretic transdermal sumatriptan, studied in a randomized, parallel-group, placebo-controlled phase 3 trial of 530 subjects (6.5 mg/4 hours), showed 2 hour pain free response in 18% with active treatment vs. 9% of controls which was sustained for 12 hours [Pierce M, O'Neill C, Felker E, Sebree T. Sumatriptan iontophoretic transdermal system: History, study results, and use in clinical practice. Headache 2013;53(Suppl S2):34-42]. Two hour pain relief occurred in 52.9% with sumatriptan vs. 28.6% of controls and was also sustained for 12 hours. There was no statistical difference in the 1 hour nausea free rate between the two groups. Most adverse events were related to application site reactions, occurring in 56.8%. The product, marketed as Zecuity™ (Teva Pharmaceuticals), was FDA-approved in 2013 with recent availability.

Idiopathic Intracranial Hypertension Treatment Trial

Despite longstanding and common usage, the effectiveness of acetazolamide for IIH treatment had never been previously demonstrated in a clinical trial. The IIHTT (National Eye Institute) studied acetazolamide vs. placebo with a supervised medical weight loss program in 165 subjects with mild visual loss from IIH [Friedman DI, McDermott MP, Kieburtz K, Kupersmith M, Stoutenburg A, Keltner J, Feldon SE, Corbett JJ, Schron E, for the NORDIC IIHTT Study Group. The Idiopathic Intracranial Hypertension Treatment Trial: Design considerations and methods. J Neuro-Ophthalmol 2014;34:107-117]. The primary outcome was change in perimetric mean deviation on Humphrey automated perimetry. The central 24 degrees of visual field were assessed, as is standard with this method of visual field testing in clinical practice. The perimetric mean deviation (PMD) is a summary variable that represents the average of light intensity (measured in decibels, dB) perceived at predetermined test locations spaced 6 degrees apart. Patients with PMD from -2 to -7 dB were included (normal is greater than -2 dB and most normal individuals have PMD between -2 and zero).

IIH was diagnosed using standard criteria, including the presence of papilledema, an elevated lumbar puncture opening pressure, no apparent secondary cause and a normal brain MRI. Potential subjects could not have received previous treatment for IIH, other than a short course of acetazolamide which was washed out prior to the baseline evaluation. Eligible participants were randomly assigned to receive either acetazolamide 500 mg BID or matching placebo tablets. The dose was gradually escalated by 250 mg weekly as tolerated to a maximum of 4,000 mg daily of acetazolamide or the equivalent of placebo tablets; the minimum allowed dose was 125 mg daily. All participants had access to a supervised weight loss program administered via telephone by weight loss coaches at the New York Obesity Nutrition and Research Center. The primary outcome was PMD in the most affected eye at 6 months.

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Patient with Progressive Proximal Muscle Weakness

By Shamin Masrour, TNS Board Resident Rep.
University of Texas Southwestern Medical School Residency Programs at Austin

58 year old Burmese male presented with 6 month history of progressive bilateral upper and lower extremity weakness. His symptoms started when he was working as a dishwasher in a cafeteria. He finally quit his job, as he was no longer able to do his work. He continued to have progressive worsening of his symptoms and was admitted to the hospital for further evaluation.

The patient endorsed generalized fatigue and unintentional weight loss of 30 pounds over 6 months. He also endorsed dysphagia to solids. He denied any fevers, rash, or myalgias. His weakness consisted of difficulty lifting his arms above his head, combing his hair, and taking the stairs both up and down. He had also noticed thinning of his limbs.

Patient's past medical history included chronic back pain, GERD, and Hepatitis C, which was untreated. Patient had moved from Burma to the United States 5 years ago. He endorsed prior history of alcohol and tobacco abuse as well as history of IV Heroin use. Family history was notable for stroke in father, otherwise no history of myopathy or neuromuscular disease was noted in the family.

On general examination, patient was thinly built and moderately nourished. He had a normal cranial nerve and sensory exam. He was noted to have significant muscle atrophy around the shoulder girdle, intercostal areas, deltoid, and upper thighs. On motor examination, he had weakness in shoulder abduction, adduction, hip flexion and extension. He had diminished biceps and triceps reflexes.

Differential diagnosis for his progressive proximal muscle weakness was broad including inflammatory myopathy, paraneoplastic syndrome, toxic or infectious myopathy, and neuromuscular junction disease. Laboratory tests revealed an elevated Creatine Kinase of 8,865 and transaminitis. A full hepatitis panel was sent, which was positive for hepatitis B, in addition to his known hepatitis C infection. PPD and Quantiferon-TB Gold test were also positive and patient was found to have latent tuberculosis infection. CEA was also checked, which was highly positive and given history of dysphagia, EGD and colonoscopy were performed and were unremarkable. Other laboratory work up revealed negative antinuclear antibodies, ESR, C Reactive protein, HIV, cryoglobulin, and myasthenia gravis and myositis panel.

To proceed further with his work up, a muscle biopsy was scheduled. The patient had an MRI of the humerus to determine the best site of muscle biopsy. The MRI revealed hyperintense T2-weighted signal involving multiple muscles, including the triceps, pectoralis, biceps, anterior and posterior deltoid, subscapularis, supraspinatus, infraspinatus, teres, and serratus anterior musculature. He had a CT of the chest, abdomen, and pelvis that showed no evidence of malignancy. The patient underwent a muscle biopsy of the deltoid, which revealed inflammatory myopathy most consistent with polymyositis.

Based on clinical findings, MRI imaging, and muscle biopsy results, the patient was diagnosed with polymyositis. While awaiting pathology results, patient was started on IV methylprednisolone and IVIG. He made significant improvement after starting treatment. The plan was to ultimately start patient on Rituximab but there was concern for re-activation of the patient's hepatitis B and latent tuberculosis infection with immunosuppresants/immunomodulators. He was also started on Rifampin for treatment of latent tuberculosis infection. He was discharged from the hospital with gastroenterology, rheumatology, and tuberculosis clinic follow up.

Discussion

Polymyositis is an inflammatory disease that is an uncommon but frequently misdiagnosed disorder (1). It is mainly a disease of adults and more common after the second decade of life (2). It has a subacute onset of weeks to months and characterized by proximal and often symmetric muscle weakness (1). Distal weakness can occur later in the disease and is not as severe as proximal weakness (4). Polymyositis may be associated with other connective tissue diseases and pure polymyositis is rare (4). In our patient, a full autoimmune work up for connective tissue diseases was unremarkable.

Occasionally, the pharyngeal and neck-extensor muscles can be involved, as in this case, causing premature fatigue and difficulty holding the head up (1), and dysphagia is seen in one-third of patients (2). Creatine kinase, which is increased up to 50 times in active disease, is the most sensitive enzyme assay (1). Creatine kinase is almost always elevated in polymyositis, whereas it can often be normal in other inflammatory myopathies. There is an increased rate of malignancies with polymyositis and dermatomyositis. Patients should be carefully screened for the presence of an occult malignancy at the time of presentation and reassessed annually thereafter. In Asian patients, among whom nasopharyngeal cancer is more common, assessment of ears, nose, and throat is suggested (1).

Muscle biopsy, serum muscle enzyme concentrations, and electromyography can confirm the diagnosis. Needle electromyography shows increased insertional and spontaneous activity, with small-amplitude low-frequency fibrillation potentials and positive sharp waves (2). Pathology shows multifocal lymphocytic infiltrates surrounding and invading healthy muscle fibers. Perivascular, perimysial, or endomysial inflammation can be present (1). There is presence of CD8+ T cells and macrophages in the myofibers (5).

Steroids are usually the first-line drug for treatment of polymyositis and the majority of patients respond well to treatment (1,4). Immunosuppressants can also be used at the same time as steroids to eventually allow for a lower maintenance dose of steroids (4) or when resistance to steroids develops. IVIG can be used with disease resistance to steroids and immunosuppressants. Typically, immunosuppressive agents, such as

azathioprine and methotrexate, or IVIG are added if there is no improvement after 3-6 months of steroid use (2).

Patients with polymyositis typically have a good prognosis, however, poor prognostic factors include older age, male gender, non-Caucasian ethnicity, longer symptom duration, interstitial lung disease, cardiac involvement, dysphagia, cancer, and serum myositis-specific antibodies (2).

In this patient, there were multiple underlying problems that could have similar manifestations. This made it more difficult to determine the real underlying pathology. There have been neurologic complications noted with hepatitis virus infections and heroin use. There are reported cases of HCV and HBV infections and inflammatory myopathy (3,7). Chronic myopathy has also been noted in chronic heroin use (6). For this reason, there was a broader differential diagnosis initially for this patient's myopathy. A biopsy in this case was valuable in making the diagnosis.

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There were 161 women and 4 men in the study [Wall M, McDermott MP, Kieburtz KD, Corbett JJ, Feldon SE, Friedman DI, Katz DM, Keltner JL, Scron EB, Kupersmith MJ. Effect of acetazolamide on visual function in patients with idiopathic intracranial hypertension and mild visual loss. JAMA Neurol 2014;13(16):1641-51]. The improvement in PMD was greater in participants receiving acetazolamide than placebo (1.43 dB vs. 0.71 dB) with a therapeutic gain of 0.71 dB (p=0.05). While the overall improvement did not meet predetermined endpoints for clinical significance, greater gains were seen in patients who had more severe visual loss at baseline. Acetazolamide was also associated with greater improvements in the less affected eye, in papilledema grade in both eyes, vision-related quality of life, general quality of life, reduction in weight (-7.50 kg vs -3.45 kg) and CSF pressure. 16 subjects withdrew in each treatment group, including 1 treatment failure in the acetazolamide group vs. 6 in the placebo group. Headache disability (HIT-6) improved slightly in both groups with no difference between groups at 6 months. Acetazolamide was well tolerated and safe.

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Summer TNS meeting to feature several firsts

Brian Loftus, MD and Michael Ready, MD, Co-program directors

This summer's TNS meeting will feature several firsts. The format has been expanded to 16 hours to allow for 12 hours on headache medicine. The meeting will be help jointly with the Southern Headache Society. The speaker lists includes the highest ranking official ever to address our group – General Pete Chiarelli, US Army (retired)

The 4 hours not devoted to Headache Medicine includes hours covering the Orexin Story in the area of Sleep Medicine (Todd Swick) and the NMO Spectrum disorders (Brian Weinshenker) – a group of patients who had been previously misdiagnosed as Multiple Sclerosis. Amit Verma will cover the rational selection of AEDs. Finally, General Chiarelli will talk about Open Science, the Key of Accelerating Cures.

In the area of headache medicine, we will cover treatments that you can be prescribing and procedures that many of you can be performing today in your office setting to improve patient care. We will explore recent trends in neuromodulation as well as controversies in decompression surgery. We will cover the neuroplasticity of pain, behavioral interventions for migraine and CAM. We will discuss new therapies that will be available in just a few years. We will highlight the expanded role of Headache Centers.

We look forward to seeing you in Fort Worth July 31 to August 2, 2015. Links for sign up are available at the TNS website.

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

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