TEXAS NEUROLOGICAL SOCIETY SUMMER 2013

Broca's Area

The Voice of Texas Neurology

President's Message

Mark Schwartze, MD

TNS Members,

I can remember as a medical resident sitting in the office of one of the past presidents of the Texas Neurological Society admiring his many achievements and awards. He was particularly proud to have been the recent president of TNS. He knew I was interested in neurology and he encouraged me to join TNS if I returned to Texas which I did. Today, he would be pleased and very proud to see how we, as neurologists and as an organization, have progressed. The Texas Neurological Society has developed a well-deserved reputation as being one of the best and strongest state societies. This is largely due to the excellent educational programs, strong statewide membership, and outstanding state and national leaders in our specialty. It is also because TNS works to help the membership adapt and remain relevant with the continuous changes to medicine, neurology and patient care.

There is great concern about new regulations and intrusions into our practices. Moreover, we need to react to these to avoid penalties and sanctions. Also, there is an enticement, a "carrot", for reacting early enough to benefit from reimbursement and incentive programs. The Medical Economic Committee chaired by Stuart Black, MD has started a site on the TNS web page called the Medial Economics Corner (ME Corner) which addresses issues affecting us and our practices. Currently, there is an article about the incentive program, Physician Quality Reporting System (PQRS) on the website. Neurologists who do not participate in PQRS as well as other incentive programs; electronic prescribing and electronic health record, are facing multiple payment adjustments in the future. Future payment adjustments will be based on what is reported in 2013.

This year the state legislature is in session. The Texas Medical Association (TMA) sponsored "First Tuesdays". The first Tuesday of February, March, April and May were set aside for physicians and medical students to go with their county medical societies and meet with their legislators. As one of the many April "First Tuesday" participants, I was able to see firsthand how valuable this day is. We were briefed on bills by the TMA and then, collectively, we carried our message to our legislators. They listened attentively and hopefully we made an impression on the ones with whom we met. We specifically discussed the proposed Medicaid reform and expansion, the concerns we all had about the Texas Advanced Directive Act and the need to restore funding to Graduate Medical Education. The legislation to restore funding for Graduate Medical Education and the Texas Advanced Directive Act appear favorable. However, I don't think there will be expansion of Medicaid in Texas this session. In order to protect medicine in Texas we must become interactive with our state legislators. If they are not aware of what seems obvious to us, they could unknowingly vote incorrectly.

As I mentioned, much of TNS' reputation is built on its exceptional education programs. We read past program evaluations and try to shape future programs from them. This year our summer conference, under the direction of Ed Fox, MD, as program chair, will focus on multiple sclerosis. This is timely given the new oral medications that are available and the new medication that will be available soon. It is going to be a great conference--held at the Westin La Cantera in San Antonio-- and I would like to encourage you to attend and bring your family.

See you in San Antonio.

Mark Your Calendar

2013 SUMMER CONFERENCE

July 19-20, 2013

Westin La Cantera
Hill Country Resort
San Antonio, Texas

(more details see page 4)

Editor's Notes

Randolph W. Evans, MD

This issue

I thank our officers and other contributors for their excellent submissions to this issue. We are delighted to introduce the new resident section initiated by TNS Resident Representative, Brent Bluett, who has provided the inaugural case study. We much appreciate the efforts of Stuart Black for deciphering the indecipherable with his article on the Physician Quality Reporting System (PQRS) and initiating the Medical Economics Corner on the TNS website with additional timely articles.

We look forward to seeing you at the TNS Annual Summer Conference in San Antonio July 19-20. Ed Fox, program chair; Bob Fayle, education committee chair; and the education committee have planned a terrific program. Be sure to make your Westin La Cantera Hill Country Resort reservation and register in time for the early bird discounts.

The Touch Typing Divide

One of the most useful classes I ever took was a hot summer school class in 1965 after 7th grade learning how to type on an typewriter at Pershing Junior High School in Houston which still only had area fans and no air conditioning. I may have been the only boy in the class (a bonus). My friends thought I was crazy to spend my time this way. However, I could clearly see the value of learning this skill as my father is an emeritus university professor and was a prolific author typing manuscripts at home frequently. And until recently, I was only typing manuscripts and emails with great delight.

If you're about 35 or younger, you're probably a good typist. But if you're older, you may be typing challenged which results in lots of wasted time in the age of EHRs. Before using an EHR, I was spending about one hour a day dictating office consults (at a cost of about $15,000 per year). Now, like many of you, I type the note as I talk to the patient saving time and money.

Many of your do or will (if you get an EHR) dictate using Dragon Voice or a transcriptionist for your EHR note. But have you considered to learn how to touch type? Think how much time (and money if using a service) you will save for a minimal investment in training in a skill which you can practice for hours a day? There are many excellent free typing instruction programs available on the internet.

Suffering Fools Gladly

Some days it is difficult to be patient and understanding with some patients or their family and friends who vehemently disagree with whatever you say because of incorrect information from the internet or a cousin or anecdote, etc. Or they may have opinions and views that you find unusual.

Perhaps the phrase, "suffering fools gladly," may pop into your head. The phrase originally came from Tyndale's 1534 translation of the Bible where Paul was ripping into the decadent citizens of Corinth for turning away from his own authoritative teaching and falling for a bunch of second-rate false apostles. "For ye suffers fools gladly," Paul says with withering sarcasm, "seeing ye yourselves are wise." (Brooks D. Suffering fools gladly. New York Times, January 3, 2013).

The suspicion that some people have unusual or controversial beliefs is not unfounded. Public Policy Polling performed a nationwide survey in March, 2013. Here are some of the results.

Do you believe there is a link between childhood vaccines and autism, or not? Do, 20%

Do you believe the moon landing was faked, or not? Do, 7%

Do you believe that shape-shifting reptilian people control our world by taking on human form and gaining political power to manipulate our societies, or not? Do, 4%

Do you believe media or the government adds secret mind-controlling technology to television broadcast signals, or not? Do, 15%

Do you believe that the pharmaceutical industry is in league with the medical industry to "invent" new diseases in order to make money, or not? Do, 15%

Do you believe Paul McCartney actually died in a car crash in 1966 and was secretly replaced by a lookalike so The Beatles could continue, or not? Do, 5%

Red Ear Syndrome

Brent Bluett discusses "neck-tongue syndrome" in the resident section which may not be familiar to some of you. I discuss Red Ear Syndrome (RES) which is also rare and may also not be familiar.

Case. — This is a 62 year old male seen in April, 2013 with a 3 year history of episodes which can occur as often as 2-3 times per week or he can go for 8 months with no episodes. He describes a heat on the right ear which gets bright red and a pull or itching of the right posterior cervical area lasting from 2 to 15 minutes. He has no precipitants.

He has a history of migraine visual aura without headache for a few years and a 7 year history of neck pain which can be daily with numbness going to the left 4,5 fingers at times. A cervical MRI showed multi-level degenerative changes.

Past history of viral meningoencephalitis one year ago and hyperlipidemia. Neurological exam was normal.

Discussion. — Since Lance first described RES in 1994 (Lance JW. The mystery of one red ear. Clin Exp Neurol 1994;31:13–18), more than 80 cases have been reported (Ryan S, Wakerley BR, Davies P. Red ear syndrome: a

review of all published cases (1996-2010). Cephalalgia. 2013;33(3):190-201; Queiroz LP. Unusual headache syndromes. Headache. 2013;53(1):12-22; Evans RW, Lance JW. The red ear syndrome: An auriculo-autonomic cephalgia. Headache. 2004;44:835-836). The disorder is characterized by episodic burning pain, usually in one ear lobe, associated with flushing or reddening of the ear with a duration of seconds to hours (constant in 2 cases) in children and adults. The average age for idiopathic cases is 35 years with 62% females and for secondary cases 50 years with 70% females.

In individuals, one ear, alternating ears, or occasionally both ears can be involved in attacks that can occur rarely or up to 20 per day. The redness can occur without pain. The pain may radiate to the cheek, forehead, a strip behind or below the mandible, behind the ear, occipit, and the ipsilateral upper posterior neck. Attacks may be spontaneous or precipitated (in 31% of idiopathic cases and 63% of secondary cases) by touching the ear, drinking, coughing, chewing, sneezing, neck movement, exercise, stress, or exposure to heat or cold.

To understand secondary causes of RES, it is helpful to recall the sensory supply of the ear which includes C2 and C3 and cranial nerves V, VII, IX, and X. The anterosuperior ear lobe is supplied by the auriculotemporal nerve (from V3) and the inferoposterior ear lobe is supplied by the greater auricular nerve (C2 and C3).

RES can be idiopathic or occur in association with migraine (during or between headache episodes), thalamic syndrome, atypical glossopharyngeal and trigeminal neuralgia, upper cervical spine pathology (cervical arachnoiditis, cervical spondylosis, traction injury, Chiari malformation, or herpes zoster of the upper cervical roots), and dysfunction of the temporomandibular joint.

Lance postulates that the cause might be an antidromic discharge of nerve impulses in the third cervical root and greater auricular nerve in response to some local pain-producing lesion in the upper neck or trigeminal areas of innervation. Al-Din et al suggest that primary and secondary cases may be due to activation of the trigeminal-autonomic reflex (Al-Din AS, Mir R, Davey R, Lily O, Ghaus N. Trigeminal cephalgias and facial pain syndromes associated with autonomic dysfunction. *Cephalalgia.* 2005;25:605-611).

A variety of treatments have been tried with variable success including gabapentin, amitriptyline, indomethacin, flunarizine, nimodipine, and ibuprofen. Local anesthestic block or section of the third cervical root might be helpful. Some cases are resistant to treatment.

In this case, RES is associated with migraine aura without headache and cervical degenerative spine disease.

The New TNS Medical Economics Update Corner

Stuart B Black MD, FAAN
TNS Medical Economics Committee Chair

The creation of the discipline "*Medical Economics*" is based on a 1963 article written by the American Economist and winner of the 1972 Nobel Memorial Prize in Economics, Kenneth Arrow PhD.

In his article, "Uncertainty and the Welfare Economics of Medical Care" Arrow stipulated that Medical Economics is entirely focused on medical services and not health care per se. Arrow's 1963 article essentially started the discussion about health care markets. As Medical Economics influence health care markets, economic issues also result in changes in how physicians practice medicine. Over the years, following numerous major health care legislative acts, measuring and reporting "quality" now ties physician reimbursements to the formula Value = Quality divided by Cost.

Many of the mandated quality measurement programs are confusing, overlapping and onerous. While Medical Economics influence health care markets, it is still not clear how patient care will be affected. As legislative mandates continue to add quality measures to existing programs, the burden includes more accountability, public reporting, the expense of purchasing and maintaining EHRs, accomplishing Meaningful Use and the increased risk of practice audits. There are also some who are concerned that the unintended consequence of incorporating the numerous legislative mandates could influence the traditional doctor/patient interaction and relationship.

Current physician monetary incentives for compliance will turn into economic penalties for noncompliance. To assist our colleagues in navigating this evolving landscape, the TNS has developed a new Medical Economics section on our website; the "ME Update Corner". The TNS encourages our members to visit the "ME Update Corner" at www.texasneurologist.org and periodically review the website to take advantage of the relevant contributions which will be added on an ongoing basis.

The TNS goal is to provide information which will hopefully be helpful in dealing with some of these emerging changes.

TNS 10th Annual Summer Conference Preview

Edward Fox, MD, PhD
TNS Summer Program Director

The 2013 Summer Conference is approaching, and will be held at the beautiful Westin La Cantera in San Antonio on July 19-20. Held on Friday afternoon and Saturday morning, it will have eight speakers covering a variety of topics. On Friday, the session is co-sponsored by the National Multiple Sclerosis Society, and topics will include discussion of the many new therapies available for MS treatment. Additional speakers will cover the definition of "treatment failure," how to evaluate cognitive problems in MS, and a very intriguing talk about the environmental factors that may trigger the disease.

On Saturday, the diverse topics will include concussions and chronic traumatic encephalopathy, the physiatrist's role in neurologic gait dysfunction, and an update on sleep disorders. The ethics hour will be a practical discussion on how to accurately document levels of disability for the Social Security Administration, with special attention on how to save time and effort in getting the right decision.

Please make time in your busy summer schedule for a get-away to the Texas Hill Country and enjoy the atmosphere as well as the educational opportunity. Further information about the meeting and registration materials are available at www.texasneurologist.org.

See you there!

Legislative Update – May 2013

Sara Austin, MD
TNS Legislative Chair

I am dating this article, because in another two months, everything could change. Let's see, where to start? The 800 pound gorilla right now seems to be Medicaid expansion, and the Medicaid 'bump' for primary care.

The AAN hosted "Neurology on the Hill" in Washington, DC at the beginning of April. It was well attended by 158 neurologists from across the country; nine from Texas. Our 'ask' was for our representatives to sign on to a bill adding neurology to those specialties who are getting paid Medicare rates to see Medicaid patients for 2013 and 2014 (courtesy of Obamacare). Those specialties are family practice and internal medicine (including the internal medicine subspecialists). We joined a coalition with the psychiatry and OB/GYN specialties in hopes of showing the importance of being included—strength in number. Though we really have no reasonable expectation that this bill will pass, it did give us the opportunity to explain what we do, and how few neurologists in the state take Medicaid, and how broken the entire system actually is.

Back in Texas and later that next week, the Texas Medical Association wrote a letter to the conference committee for the budget (a group of House and Senate members who reconcile the House budget with the Senate budget) asking that they increase **all** Medicaid rates to Medicare levels for the next biennium. Rather a 'hail Mary' pass, but shoot, you never know, and we appreciated the effort.

Knowing this was a long shot, the TNS then wrote our own letter asking the committee to consider at least raising the rate for E&M codes to Medicare levels for 2013 and 2014 if they could not increase all Medicaid rates. We hand delivered that letter to the members of the budget committee. There is a general agreement that neurology access for Medicaid patients is truly drying up here in Texas. I was feeling a little hopeful that the legislature was going to be able to find at least some extra money for Medicaid this biennium, until the Governor weighed in asking for a big tax cut. That's pretty disappointing.

There is still a lot of talk around the Texas Capital about Medicaid expansion which could conceivably add two million low income folks to the Medicaid rolls in 2014. It looks like all bills that pertain to that have died in committee. However, it appears that there is some language in the Senate budget that might allow for some expansion. So, after the session is over something may be able to be cobbled together. It is about $10 billion dollars over 10 years from the federal government. I think it will be hard to pass up that kind of money, especially for the hospitals.

Overall, this Texas legislative session has been a positive one for doctors. An agreed-to bill with the TMA and nurse practioners has almost passed both houses and will soon head to the Governor. This allows increased supervision of mid-levels, rules for supervision that make more sense, and slightly more autonomy for nurses who have practiced for a while, *all under the direction supervision of a physician (as it should be).* A bill to streamline DPS renewal with license renewal is also about to pass, and the 'silent PPO' bill that the TMA has been lobbying for more than 10 years has passed both houses and is heading out to be signed. We are working on a bill to standardize prior authorization forms that is working its way slowly thru the process. If it does

not make it this year, hopefully it will the next session. We have also successfully kept many bad bills bottled up in committee or in calendars, so those are considered wins as well.

On a federal level, I am sorry to say that it is very unlikely there will be a long term SGR fix. They have us over a barrel, and there is no real reason to not keep us there. The AAN continues to meet with CMS about the nerve conduction study code cuts but it is unlikely any changes will be made for at least another year. The AAN now have two full time lobbyists on the Hill and they have hired Dr. Bruce Sigsbee part time. He is a very effective speaker about neurology economics so hopefully he will be a help.

Please continue to respond to Action Alerts from the AAN and send letters to your Congressmen. Join TexPAC and BrainPAC as you are able and remember your participation makes a difference!

Letter to the Editor

BLIP SYNDROME

To the editor:

I appreciated your article regarding "blip syndrome." I had a patient with similar symptoms about one week prior to your article in Broca's Area arriving on my desk. Patients with these types of diagnoses are rare and to read your review of the literature was helpful to me and the patient as I had never heard of the syndrome previously. Interestingly, his symptoms virtually resolved with elimination of caffeine from his diet.

Bill Davis, MD
New Braunfels

American Academy of Neurology Update – June 2013

Practice Management Resources

Updated Sports Concussion Guideline Available
Review resources for physicians, patients and caregivers, sports coaches and athletic trainers at www.aan.com/go/practice/concussion.

Are You Prepared for and EHR Incentive Program Audit?
Eligible providers and hospitals attesting in either the Medicare or Medicaid Electronic Health Record (EHR) Incentive Program may be subject to an audit.

CMS has developed a supporting documentation for audits fact sheet to help eligible professionals prepare for an audit. This and other resources are available at www.cms.gov/Regulations-and-Guidance/Legislation/EHRIncentivePrograms/EducationalMaterials.html.

AAN Releases FAQ on Balancing Medicare Incentive Programs and Penalties
View the document online at http://www.aan.com/globals/axon/assets/10781.PDF

Practice Management Webinars - Register at www.aan.com/view/webinar.

E/M: Minimize Mistakes, Maximize Reimbursement – June 25
Public Policy Resources

US Facing a Neurologist Shortage
Americans with brain diseases such as Alzheimer's disease, Parkinson's disease or multiple sclerosis (MS) who need to see a neurologist may face longer wait times or have more difficulty finding a neurologist, according to a new study published in the online issue of Neurology®. Visit www.aan.com to learn more.

AAN Advocates Visit Capitol Hill
AAN members took part in the 11th annual Neurology on the Hill April 21-22. This is one of the Academy's signature advocacy events, and 144 neurologists representing 43 states, 130 congressional districts, and Washington, DC, met with 216 members of Congress to discuss the need for fair Medicare and Medicaid reimbursement to help prevent a shortage of neurologists available to care for people affected by neurologic disease. Visit www.aan.com/go/advocacy/hillreport or www.aan.com/noh for more information.

AAN Attending TNS Summer Conference
AAN staff will be in attendance at the TNS Summer Conference to answer questions and obtain member feedback. Stop by our booth to discuss your membership and find tools and resources on important neurological issues. Contact dshowers@aan.com for more information.

Expert Opinion:
Distinguishing Essential Tremor from Parkinson's Disease

Mary Ann Thenganatt, MD
Assistant Professor of Neurology, Baylor College of Medicine
Parkinson's Disease Center and Movement Disorders Clinic, Houston

Case:

A 40 year-old woman with a family history of essential tremor (ET) develops a kinetic tremor and, a few years later, a postural head tremor. Her tremor worsens considerably such that she has severe and debilitating tremor by the age of 65. At age 70, she also develops a rest tremor with no other parkinsonian features.

Questions:

1. What are the overlapping features of ET and Parkinson's Disease (PD)?
2. How can these two disorders be distinguished, both clinically and through laboratory studies?

Expert Opinion

Essential tremor (ET) and Parkinson's Disease (PD) are two of the most common adult-onset tremor disorders. The prevalence of ET and PD increases with age; PD is estimated to be 1.8% in individuals over the age of 65, and that of ET has been estimated to be 4.6% in this same age group.[1, 2] ET and PD patients may exhibit overlapping clinical features and, to further complicate the matter, patients with ET may eventually meet criteria for an additional diagnosis of PD (i.e., "ET+PD"). Thus, the two disorders may co-exist within the same individual, and having one seems to increase the risk of developing the other.

The criteria commonly used for the diagnosis of ET are that proposed by the Movement Disorder Society.[3] For definite ET, the diagnostic criteria require the presence of persistent, bilateral postural tremor of the forearms for at least five years. Kinetic tremor may be present, but is not necessary for the diagnosis. No other abnormal neurological signs may be present, except for Froment's sign, which is a cogwheel phenomenon without rigidity. The United Kingdom Parkinson's Disease Society Brain Bank criteria require postmortem confirmation for the diagnosis of definite PD.[4] The diagnosis of probable PD requires bradykinesia and one of the following additional features: rigidity, 4-6 Hz rest tremor, or postural instability (not caused by primary visual, vestibular, cerebellar or proprioceptive dysfunction). In addition, three supportive features are required.

Clinicians are often faced with the prospect of distinguishing ET from PD, which can be a diagnostic challenge in early stages of disease when clinical signs are subtle. One study observed that one third of patients who were diagnosed as ET was misdiagnosed, with PD being the most common true diagnosis.[5] Various tremor types (rest, postural, kinetic, intention) may be seen in both ET and PD. Detailed clinical examination with attention to specific features of tremor and associated neurological findings may help distinguish these two diseases.

CLINICAL EXAMINATION

Rest tremor

Rest tremor is a cardinal feature of PD, and when it is accompanied by bradykinesia and rigidity, PD is high on the differential diagnosis. Rest tremor, however, is also seen in 20-30 % of ET patients.[2] Rest tremor in ET typically involves the arm; in contrast to PD, in which it may occur in the arm, leg, or both.[6] Studies have shown that rest tremor occurs in ET patients with longer disease duration and more severe postural and kinetic tremor that those without rest tremor.[7, 8] Thus, onset of rest tremor in a patient with relatively mild action tremor and early on in the disease course would be atypical for ET and should raise the possibility of a diagnosis of PD.

Action tremor

Action tremor is the hallmark feature of ET and can be further subdivided into postural, kinetic and intention tremor. Action tremor in ET patients is usually, though not always, bilateral, and it is typically asymmetric.[9] Small to moderate side-side differences are the rule rather than the exception. Postural tremor occurs when holding a body part (e.g., arm, head, leg) motionless against gravity. Re-emergent tremor is a particular type of postural tremor; when the patient holds

his arms extended, the tremor commences after a variable latency of one to several seconds. Kinetic tremor occurs with voluntary movement (e.g., pouring, writing). Intention tremor occurs with goal-directed movement (e.g., finger-nose-finger movement) and worsens as the body part (e.g., finger) approaches the target.

Yet just as rest tremor may occur in patients with ET, action tremor may be found in patients with PD. It is not uncommon to encounter patients with PD who have various forms of action tremor without an additional diagnosis of ET. Studies have shown that the postural tremor in PD may be similar in frequency to the rest tremor in PD.[10] A particular type of postural tremor, called re-emergent tremor is highly suggestive of PD.[11] This tremor occurs after a variable latency period when assuming an outstretched posture. In patients with PD, the frequency of the re-emergent tremor tends to be similar to that of their rest tremor. Kinetic arm tremor is another form of action tremor, classic for ET. Studies have shown that the kinetic tremor of ET may be more severe and of higher amplitude than that of an ET patient's postural tremor.[12] In contrast to the situation in ET, the amplitude of kinetic tremor in PD has been shown to be lower than that of the rest and postural tremor.[13] Intention tremor (e.g., during the finger-nose-finger maneuver) is more suggestive of ET than PD. ET patients with intention tremor are more likely to be older with longer disease duration and more severe tremor overall.[14] They are also more likely to have voice, head and truncal tremor. These patients with intention tremor may also have frank dysmetria in addition to tremor.[15]

Cranial tremor

Head tremor is more commonly observed in ET patients than those with PD. Women with ET are more likely to have head tremor than men.[16] The head tremor of ET is typically a postural tremor that resolves at rest. Head tremor has been observed in PD but is rare. Head tremor in PD is described as a rest tremor with a similar frequency (4-6Hz) as the arm rest tremor and responsive to levodopa.[17]

Jaw tremor is classically associated with PD and it typically occurs when the mouth is closed at rest. Again, jaw tremor may occur in ET but is much less common. Jaw tremor may occur in up to 18% of ET patients and has been observed to be a predominantly postural tremor (occurring during voluntary mouth opening) or kinetic tremor (occurring while speaking).[18]

Bradykinesia

Bradykinesia is a cardinal sign of PD demonstrated by decreased facial expression, slowness with rapid alternating movements, difficulty arising from a chair and reduced arm swing. Although bradykinesia is not traditionally associated with ET, studies have demonstrated slowed rapid alternating movements, finger tapping and reduced arm swing in a small proportion of ET patients.[19] However, a reduction in amplitude and cessation of movement (pauses or freezing) during rapid successive movements has not been demonstrated in ET.

Archimedes' Spirals and Handwriting

Archimedes' spiral analysis can be accomplished by qualitative visual inspection at the bedside. ET spirals tend to have a peak spiral amplitude that lines up along an axis. One study found the majority of right hand spirals to have an axis corresponding to the numbers 2 and 3 on the face of a clock; the majority of left hand spirals had an axis corresponding to 10-12 o'clock (a 90 degree angle to the right hand axis).[20] When evaluating PD spirals, the hand with greater bradykinesia may produce smaller spirals (e.g., more compact with a smaller diameter).

Having a patient write can also be informative. ET patients may have obvious tremor when forming letters. In contrast, the hallmark feature of PD is micrographia with the letters becoming smaller and smaller as they continue to write.

LABORATORY EVALUATION

Neuroimaging

The dopamine transporter (DAT) is a presynaptic protein that is used as a biomarker for dopaminergic nigrostriatal neurons.[21] Single photon emission tomography (SPECT), with cocaine derivative tracers binding to DAT, can thus be used as a measure of dopamine deficiency seen in PD. DAT-SPECT scans were FDA-approved in 2011 to help distinguish ET from parkinsonian syndromes. While this scan can be a helpful diagnostic tool, it is not without limitations and needs to be interpreted carefully. DAT-SPECT scans should not replace a detailed clinical examination and should be interpreted in the context of the entire clinical picture.

Genetics

During the past 10 years, there has been increasing interest in the search for susceptibility genes for PD and ET. A number of genetic forms of PD have been identified, including both autosomal dominant and recessive forms;[22] however, genetic testing is not used for diagnostic purposes of PD and is not used to distinguish PD from ET. The genetic causes of ET are not as well defined. Linkage studies have identified three genetic loci in ET families.[23] In 2009, a genome-wide

continued on next page

association study reported an association between a variant in the *LINGO1* gene and ET.[24] While further studies also demonstrated this association, not all studies have been confirmatory. Interestingly, in several studies, variants in the *LINGO1* gene have also been associated with PD.

Postmortem

The pathological hallmark of PD is neuronal loss in the substantia nigra pars compacta and the presence of Lewy bodies-neuronal inclusion bodies composed of the protein α-synuclein. The study of the pathological changes in ET is in its infancy relative to that of PD. Few quantitative controlled studies have been performed. Studies at the Essential Tremor Centralized Brain Repository have indicated that the majority of ET cases have postmortem changes in the cerebellum.[25] While post-mortem studies help us learn more about these two diseases they do not aid in diagnosis during a patient's lifetime.

COMMENTARY ON CLINICAL CASE

Does the patient in the introductory case above have ET or ET-PD?

This woman likely only has ET with long-standing, progressive kinetic and head tremor. She develops rest tremor, which can be seen in advanced ET cases and does not meet criteria for additional Parkinson's disease.

Conclusion

Distinguishing ET from PD is important in terms of selecting appropriate therapy as well as counseling patients about disease progression. As the same types of tremor may occur in both disorders, the clinician should be aware of patterns suggestive of ET versus PD and categorize accordingly. Nonetheless, this is still a best estimate; clinical features may evolve or new signs may develop and the diagnosis may need to be revised accordingly. A diagnosis of ET + PD should only be assigned when a patient fulfills the clinical criteria for each individual diagnosis. Laboratory testing is only supportive and should be interpreted in the context of the entire clinical picture.

References

1. de Rijk MC, Launer LJ, Berger K, et al. Prevalence of Parkinson's disease in Europe: A collaborative study of population-based cohorts. Neurologic Diseases in the Elderly Research Group. Neurology 2000;54:S21-23.
2. Louis ED, Ferreira JJ. How common is the most common adult movement disorder? Update on the worldwide prevalence of essential tremor. Mov Disord 2010;25:534-541.
3. Deuschl G, Bain P, Brin M. Consensus statement of the Movement Disorder Society on Tremor. Ad Hoc Scientific Committee. Mov Disord 1998;13 Suppl 3:2-23.
4. Hughes AJ, Ben-Shlomo Y, Daniel SE, Lees AJ. What features improve the accuracy of clinical diagnosis in Parkinson's disease: a clinicopathologic study. 1992. Neurology 2001;57:S34-38.
5. Jain S, Lo SE, Louis ED. Common misdiagnosis of a common neurological disorder: how are we misdiagnosing essential tremor? Arch Neurol 2006;63:1100-1104.
6. Rajput AH, Rozdilsky B, Ang L. Occurrence of resting tremor in Parkinson's disease. Neurology 1991;41:1298-1299.
7. Cohen O, Pullman S, Jurewicz E, Watner D, Louis ED. Rest tremor in patients with essential tremor: prevalence, clinical correlates, and electrophysiologic characteristics. Arch Neurol 2003;60:405-410.
8. Louis ED, Asabere N, Agnew A, et al. Rest tremor in advanced essential tremor: a post-mortem study of nine cases. J Neurol Neurosurg Psychiatry 2011;82:261-265.
9. Louis ED, Wendt KJ, Pullman SL, Ford B. Is essential tremor symmetric? Observational data from a community-based study of essential tremor. Arch Neurol 1998;55:1553-1559.
10. Henderson JM, Yiannikas C, Morris JG, Einstein R, Jackson D, Byth K. Postural tremor of Parkinson's disease. Clin Neuropharmacol 1994;17:277-285.
11. Jankovic J, Schwartz KS, Ondo W. Re-emergent tremor of Parkinson's disease. J Neurol Neurosurg Psychiatry 1999;67:646-650.
12. Brennan KC, Jurewicz EC, Ford B, Pullman SL, Louis ED. Is essential tremor predominantly a kinetic or a postural tremor? A clinical and electrophysiological study. Mov Disord 2002;17:313-316.
13. Zimmermann R, Deuschl G, Hornig A, Schulte-Monting J, Fuchs G, Lucking CH. Tremors in Parkinson's disease: symptom analysis and rating. Clin Neuropharmacol 1994;17:303-314.

14. Louis ED, Frucht SJ, Rios E. Intention tremor in essential tremor: Prevalence and association with disease duration. Mov Disord 2009;24:626-627.
15. Deuschl G, Wenzelburger R, Loffler K, Raethjen J, Stolze H. Essential tremor and cerebellar dysfunction clinical and kinematic analysis of intention tremor. Brain 2000;123 (Pt 8):1568-1580.
16. Hardesty DE, Maraganore DM, Matsumoto JY, Louis ED. Increased risk of head tremor in women with essential tremor: longitudinal data from the Rochester Epidemiology Project. Mov Disord 2004;19:529-533.
17. Roze E, Coelho-Braga MC, Gayraud D, et al. Head tremor in Parkinson's disease. Mov Disord 2006;21:1245-1248.
18. Louis ED, Rios E, Applegate LM, Hernandez NC, Andrews HF. Jaw tremor: prevalence and clinical correlates in three essential tremor case samples. Mov Disord 2006;21:1872-1878.
19. Jimenez-Jimenez FJ, Rubio L, Alonso-Navarro H, et al. Impairment of rapid repetitive finger movements and visual reaction time in patients with essential tremor. Eur J Neurol 2010;17:152-159.
20. Louis ED, Yu Q, Floyd AG, Moskowitz C, Pullman SL. Axis is a feature of handwritten spirals in essential tremor. Mov Disord 2006;21:1294-1295.
21. Vlaar AM, van Kroonenburgh MJ, Kessels AG, Weber WE. Meta-analysis of the literature on diagnostic accuracy of SPECT in parkinsonian syndromes. BMC Neurol 2007;7:27.
22. Kumar KR, Djarmati-Westenberger A, Grunewald A. Genetics of Parkinson's disease. Semin Neurol 2011;31:433-440.
23. Zimprich A. Genetics of Parkinson's disease and essential tremor. Curr Opin Neurol 2011;24:318-323.
24. Stefansson H, Steinberg S, Petursson H, et al. Variant in the sequence of the LINGO1 gene confers risk of essential tremor. Nat Genet 2009;41:277-279.
25. Louis ED, Faust PL, Vonsattel JP, et al. Neuropathological changes in essential tremor: 33 cases compared with 21 controls. Brain 2007;130:3297-3307.

Resident Section

Neck -Tongue Syndrome

Brent Bluett, DO
TNS Resident Repersentative
University of Texas Southwestern, Austin

I present two patients with neck-tongue syndrome (NTS), review the clinical features, pathophsysiology, and treatment.

Case 1:

A 16 year old female softball pitcher, who develops recurrent transient right sided neck pain, followed by numbness of her tongue ipsilaterally. Her episodes occur mainly while pitching. The patient is without significant past medical, social, or family history. Neurologic examination shows no focal abnormalities. An MRI of the brain was performed and was negative for acute or chronic intracranial abnormalities. Most fast pitch softball pitchers use what is best described as a ¾ to full windmill delivery where the arm stops abruptly at the end of the down stroke. The pitcher also tends to step forward with their non dominant leg at delivery. This causes a good amount of torque on the right side of the body (if the pitcher is right handed) with contraction of the cervical muscles on that side; this in turn can cause compression of the cervical nerve roots.

Case 2:

A 40 year old female sports physical therapist with a past medical history significant for osteoarthritis, and a severe neck injury that occurred in high school, requiring a cervical collar for a few months. She did not break anything but was unable to flex her neck against resistance for more than 2 years. Since this injury, she has had a restricted range of motion when looking to the left, only being able to rotate her head approximately 45 degrees to the left. Any further rotation results in sudden pain that starts on the left side of her neck and radiates into the left side of her face, with associated numbness of her tongue and loss of vision in her left eye. The pain is severe enough that it would make her drop to the ground, and lasted approximately 30 seconds to one minute. She believes she has had 4-5 incidents over 20 years after her injury in high school, but has always felt like something has "not been quite right with the top of the left side of her neck" since her injury in high school. Unfortunately, in 2012, she was involved in a motor vehicle accident where she was 'T-boned'. Since then, she has been having these episodes more frequently, approximately once every two weeks. She has seen a chiropractor in the past, but never had manipulation of her neck because of muscle spasms. Neurological exam showed no focal deficits. An MRI of the brain and cervical spine performed in 2012 were both normal.

Both patients' constellations of symptoms represent the uncommon, yet well described entity, of NTS.

Discussion:

The literature reports two forms of NTS – complicated and uncomplicated, based upon the presence or absence of an underlying disease process (inflammatory or degenerative).

The International Headache Society lists this entity as a "Cranial Neuralgia and Central Cause of Facial Pain". The diagnostic criteria include:

A: Pain lasting seconds or minutes, with or without simultaneous dysaesthesia, in the area of distribution of the lingual nerve and second cervical root and fulfilling criteria B and C.

B: Pain has acute onset.

C: Pain is commonly precipitated by sudden turning of the head.

NTS was first described in 1980 by Anthony and Lance.[1] It is an uncommon disorder characterized by acute unilateral occipital pain and subsequent numbness of the ipsilateral tongue lasting seconds to approximately 5 minutes. This is generally precipitated by sudden movement, usually rotation, of the head. One study has estimated the prevalence in the general population at approximately 0.22%,[2] with approximately 50 cases described in literature. A benign, familial form of neck-tongue syndrome, likely autosomal dominant, has also been reported. This type occurs without anatomic abnormality, and resolves spontaneously during adolescence.[3] As the patient in Case 1 has no underlying past medical history, and meets all three criteria for NTS, the diagnosis based on the IHS classification is Uncomplicated NTS. The patient in Case 2, however, does have an underlying degenerative condition, osteoarthritis, which meets the criteria for Complicated NTS.

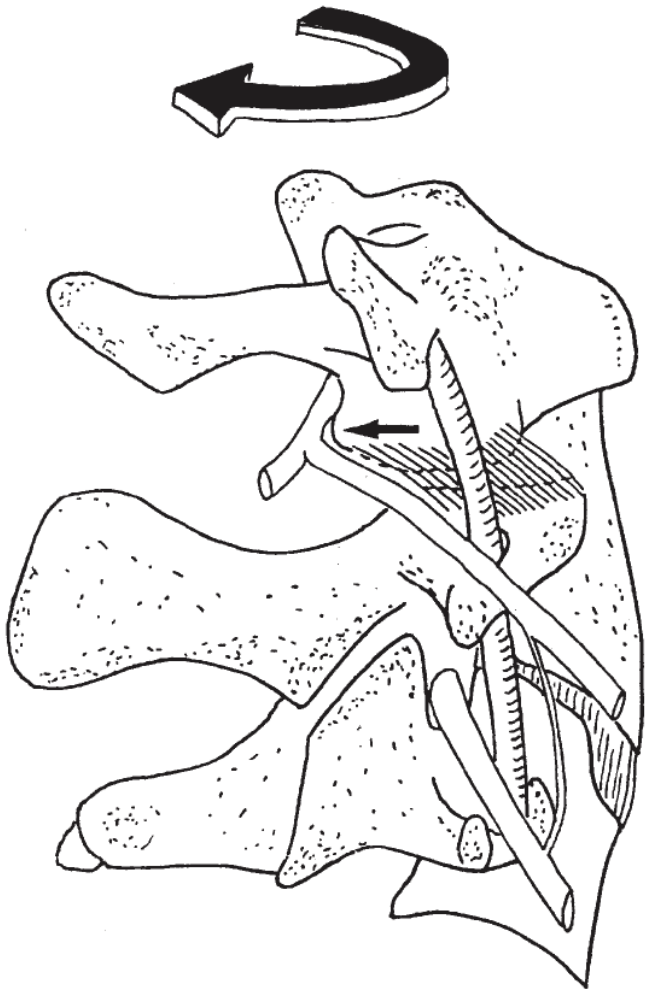

Fig. 1

Lateral view of the right atlantoaxial joint; the atlas has rotated to the right. The small arrow shows the inferior articular process impinging the C2 spinal nerve and ventral ramus. (Evans RW, Lance JW. Transient headache with numbness of half of the tongue. Headache 2000;40: 692–3; with permission.)

Acknowledgement: special thanks to Dr. Sara Austin for her numerous contributions to this article.

The symptoms of NTS are the result of transient subluxation of the atlantoaxial joint that stretches the joint capsule and the C2 ventral ramus, which contains proprioceptive sensory fibers from the tongue originating from the lingual nerve to the hypoglossal nerve to the C2 root (Fig. 1)[4]. Primary or "uncomplicated" NTS can occur without obvious abnormalities. Secondary or "complicated" NTS includes co-existing conditions such as congenital anomalies of the cervical spine. This includes Chiari-1 malformation, ankylosing spondylitis, degenerative spondylosis, rheumatoid arthritis, tuberculous atlantoaxial osteoarthritis, cervical acute transverse myelopathy, and also following head and neck trauma.[5] When evaluating patients with possible NTS, upper cervical spine and particularly atlantoaxial instability should be considered, along with muscle spasm.

There is no evidence regarding the efficacy of any particular therapeutic approach to NTS. Initially conservative treatment is recommended, which includes avoidance of neck trauma and the use of non-steroidal anti-inflammatory drugs (NSAIDs) and drugs to alleviate neuropathic pain.[6] Treatment commonly used includes cervical collars, chiropractic manipulation, analgesics/ NSAIDs, and muscle relaxants. Neuropathic agents including carbamazepine, gabapentin, and amitriptyline can be employed. Steroids, injections of local anesthetic, nerve resection, and cervical fusion are generally considered as a last resort.[7]

References

1. Lance, J., Anthony, M. "Neck Tongue Syndrome on Sudden Turning of the Head". Journal of Neurology, Neurosurgery, and Psychiatry, 1980, 43, 97-101.
2. Sjaastad O, Bakketeig LS. Neck-tongue syndrome and related (?) conditions. *Cephalalgia.* 2006;26: 233-240.
3. Lewis DW, Frank LM, Toor S. Familial neck tongue syndrome. *Headache.* 2003;43:132-134.
4. Evans, R., Lance, J. Transient Headache with Numbness of Half of the Tongue. *Headache.* 2000; 40: 692-693
5. Queiroz, L. Unusual Headache Syndromes. *Headache* 2013;53:12-22
6. Orrell RW, Marsden CD. The neck-tongue syndrome. *J Neurol Neurosurg Psychiatry.* 1994;57: 348-352.
1. Elisevich K, Stratford J, Bray G, Finlayson M. Neck tongue syndrome: Operative management. *J Neurol Neurosurg*

PQRS for Neurologists

Stuart B Black MD, FAAN
Chief of Neurology
Co-Director: Neuroscience Center
Baylor University Medical Center at Dallas

Please note this article can be found on the TNS website under the ME Update Corner

The Physician Quality Reporting System (PQRS) is designed to provide incentive payment to eligible professionals who meet the guidelines on reporting data on quality measures for covered professional services furnished on Medicare patients. There are over 300 measures in the 2013 program. PQRS has proven to be daunting to many Neurologists. Trying to understand the requirements and how to implement the measures is often confusing. It is also important to recognize that while PQRS may provide a reimbursement to Neurologists, those physicians who elect not to participate or are determined to be unsuccessful in reporting during the 2013 program year will receive a payment penalty starting in 2015. In addition CMS plans to publish the names of those practitioners who successfully participated in PQRS. There is also indication that CMS may also highlight the names of practitioners who do not participate in PQRS.

As is common with other CMS programs, PQRS has a defined vocabulary referred to as the "Glossary of Terms". The definitions of the terms can be found in the CMS 2013 Physician Quality Reporting System Implementation Guide. In Appendix A: Glossary of Terms, there are 29 items defined. The following are some of the more important terms to help understand PQRS.

Glossary of Terms:

Eligible Professional (EP): Refers to the list of professionals eligible to participate in PQRS. Since this update is written for Neurologists, the text below will reference either Neurologists or Eligible Professional

Encounter: Encounters with patients during the reporting period which include: CPT Category 1 E/M service codes, CPT Category 1 procedure codes, or HCPCS codes specific for PQRS.

CPT Category 11 Codes: A set of supplemental CPT codes intended to be used for performance measurement. For PQRS, CPT Category 11 codes are used to report quality measures on a claim for measurement calculation PQRS is reported using Category 11 CPT Codes

CPT Category 11 Codes are generally 4 numbers followed by "F" or 4 numbers preceded by "G"

e.g. 1200F for Seizure frequency and G8851 for adherence to positive airway pressure therapy

G-Codes for PQRS: Are a set of CMS defined temporary HCPCS codes used to report quality measures on a claim. G-Codes are maintained by CMS

Measure: Performance Measure is a quantitative tool (e.g., rate, ratio, index, percentage) that provides an indication of performance in relation to a specified process of outcome.

Measure Tags: Measure Tags are reporting frequencies or timeframe requirements. For example, "report each visit", "once during the reporting period", "report each episode".

The measure restrictions limit the frequency of reporting that may be necessary in certain circumstances. An example would be patients with a chronic illness for whom a particular process of care is provided only periodically

Measure Tags are found in the instructions for each measure specification

Eligible Cases: Eligible Cases are defined as a patient population that receive a particular process of care or achieve a particular outcome. The Eligible Cases are defined by demographic information, certain International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) Diagnosis, Current Procedural Terminology (CPT) and Healthcare Common Procedure Coding System (HCPCS) codes specified in the measures that are submitted by *Individual Eligible Professionals* as part of a claim for *covered services* under the *Physician Fee Schedule*.

Denominator Codes (Eligible Cases) and Numerator Quality- Data Codes: Quality measures consist of a numerator and a denominator that permit the calculation of the percentage of a defined patient population that receive a particular process of care or achieve a particular outcome

Denominator: The denominator is associated with a given population that may be counted as Eligible Cases to meet a measure's inclusion requirements and defines the Eligible Cases for a measure. For Neurologists reporting PQRS, most denominator codes will usually be ICD-9 codes

Numerator: Describes the clinical action required by the measure for reporting and performance. The

clinical action to be counted must meet a measure's requirements (i.e., patients who received the particular service or obtained outcome that is being measured). PQRS measure numerators are CPT Category 11 codes and G-codes.

When Quality Measures are calculated in terms of a numerator and denominator, the results are a percentage of a defined patient population that receives a particular process of care or achieves a particular outcome

Example of Numerator and Denominator:

Denominator: All patient visits with a diagnosis of Epilepsy

Numerator: Report the CPT Category 11, Seizure Type(s) and Current Seizure Frequency(ies) in development designated for this numerator 1200F

PQRS reports are issued to an individual National Provider Identifier (NPI) and payment is under the group Tax Identification Number (TIN)

More detailed information on these two major components of PQRS are described in the *2013 CMS Physician Quality Reporting System (Physician Quality Reporting) Implementation Guide* found on the CMS website

How can I avoid the payment penalty in 2015?

The easiest way for a Neurologist to avoid a 1.5% payment penalty in 2015 is to successfully report on at least one individual measure at each encounter for >50% of all eligible patients

For example: If using the documentation of seizure type and frequency, the Neurologist needs to report this measure for at least 50% of their epilepsy patients.

*To satisfy CMS rules and **receive** the incentive*

Individual Measures and Measures Groups:

If reporting via registry:

>80% on 3 or more individual measures

20 or more unique patients (>50% must be Medicare) for 1 or more measures group

If reporting via claims:

>50% on 3 or more individual measures for the 12 month reporting period

1 or more measures group of the 12 month reporting period for 20 or more unique patients

Group Practice Reporting Option (GPRO):

Report at least 3 Measures and report each measure for at least 80% of the group Practice's Medicare Part B FFS patients seen during the reporting period to which the measure applies

If the Neurologist chooses to report on a diagnosis with no neurologic specific measures, (e.g. migraine), he/she could choose 3 general measures. This may include such measures as medication reconciliation or tobacco use. However, it is important to understand that when choosing more general measures, those measures may not be specific to a listed diagnosis. In that case, every Medicare patient may be eligible to be considered for that measure, (e.g. medication reconciliation measure). Reporting would then apply to everyone. If a measure is not defined by a particular diagnosis code the eligible patient population would be all Medicare patients.

PQRS reporting can be submitted in the following ways:

To CMS on the Medicare Part B claims form (Part B 1500 billing form)

To a qualified Physician Quality Reporting registry

To CMS via a qualified EHR

To a qualified Physician Quality Reporting data vendor

The 3 different reporting options available for PQRS:

Reporting Individual Measures: Easiest to understand. Most Neurologists will report Individual Measures

Reporting Group Measures Options: Clinically related measures focused on chronic and high cost conditions. Most measures are not as applicable to a Neurology practice

Group Practice Reporting Option (GPRO): Overall more applicable to Internal Medicine and large multispecialty groups

Explaining the 3 different reporting options available for PQRS:

Individual Measures:

Least complicated for Neurology PQRS reporting and will probably be used by most Neurologists

Individual Measures are reported using claims, a PQRS qualified registry, or a PQRS qualified EHR

There are over 300 PQRS measures available

There are some clinical topics for individual PQRS

continued on next page

measures specific to neurology, such as stroke and epilepsy

Neurologists may also choose general individual measures such as fall screening, pain management, medication reconciliation, smoking cessation and the use of the electronic health record. When using general measures, reporting may need to be on every eligible Medicare patient

When using individual measures, to avoid the penalty, the Neurologist must report on at least one individual measure (see above)

Measures Group reporting:

4 or more measures grouped together

It is anticipated, when using measures group, most Neurologists will primarily use the clinical topics of individual group measures that exist for the following neurological diseases:

Parkinson's disease, Dementia, Sleep, Back pain

If choosing to report on a measures group, all measures in the group must be reported for all applicable patients. Each patient within the eligible professional's patient sample must be reported a minimum of once during the reporting period

For example, choosing the Parkinson's disease measures group means reporting all six measures in the group for all Parkinson's patients

Report via claims or PQRS qualified registry.

Not available for EHR reporting

To receive the bonus, the Neurologist must report on 20 patients who qualify for the measures group

Under the 2013 program, greater than 50% of those patients must be Medicare patients

Failure to reach 20 patients does not meet the requirement

To avoid the 2015 penalty when reporting measures group, the Neurologist must report on 1 measures group. But failure to reach 20 patients does not meet requirement. Thus, if 20 patients are not reached, to avoid the penalty, choose another measures group or another reporting option.

The Group Practice Reporting Option (GPRO):

For at least two or more providers in a group

Different criteria of reporting depending on the group size

Group size: 2+ eligible professionals

Group size: 25-99 eligible professionals

Group size: 100+ eligible professionals

Single – specialty Neurology practices will be less likely to choose group reporting because these measures are more geared toward the primary care provider

Details about the Neurology Specific PQRS Measures

Disease	# of Measures	Reporting Mechanism
Epilepsy	Three	Claims, Registry
Parkinson's disease	Six	Registry only Measures
Dementia	Nine	Claims, Registry
Sleep Apnea	Four	Registry

Stroke has seven measures but stroke is currently hospital level reporting and not individual EP reporting. Stroke can be reported as follows: four by claims and 6 by registry

The top 5 PQRS measures used by Neurologists are as follows:

(Cohen AB, Sanders AE, et.al..Quality measures for neurologists: Clinical Practice 2013l; vol 3;44-50)

Adoption and use of electronic health record (PQRS # 124)

Inquiry regarding tobacco use (#114)

Documentation and verification of current medications (#130)

Advising smokers to quit smoking (#115)

DVT prophylaxis for Ischemic stroke or Intracranial hemorrhage (31)

Incentives For Reporting:

2012	2013	2014	2015	2016
+0.5%	+0.5%	+0.5%	0.0%	0.0%

Penalties For Not Reporting:

2012	2013	2014	2015	2016
0.0%	0.0%	0.0%	-1.5%	-2.0%

As illustrated, there are still incentive payments for PQRS through 2014

The Patent Protection and Affordable Care Act (PPACA) requires penalty starting in 2015 (based on 2013 reporting), for providers who do not satisfactorily report PQRS

For a Neurologist to get started in PQRS:

Determine which reporting option is best for your practice

Select 3 measures group to submit (to receive incentive)

Select 1 measures group to submit (to avoid penalty)

Perform a personal review of the measure specifications chosen

PQRS is reported using Category 11 CPT Codes (described above)

There are CPT Modifiers which explain the reason for not performing a quality procedure:

1PModifier: Procedure not performed due to medical reasons

2P Modifier: Procedure not performed due to patient reasons

3P Modifier: Procedure not performed due to system reasons

8P Modifier: Procedure not performed due to reasons otherwise not specified

General Examples:

Example #1: Choosing to report Epilepsy:

There are 3 Measures for reporting Epilepsy:

1. "Percentage of patient visits with a diagnosis of epilepsy who had the type(s) of seizure(s) and current seizure frequency(ies) for each seizure type documented in the medical record"
2. "All visits for patients with a diagnosis of epilepsy who had their etiology of epilepsy or with epilepsy syndrome(s) reviewed and documented if known, or documented as unknown or crytogenic"
3. "All female patients of childbearing potential (12-44 years old) diagnosed with epilepsy who were counseled about epilepsy and how its treatment may affect contraception and pregnancy at least once a year"

To illustrate, when reporting the first measure, the CPT Category 11, "Seizure Type(s) and Current Seizure Frequency(ies)", is designated 1200F in the numerator

The Neurologist must document the reason for not performing a measure by appending the modifier to the CPT 11 Code. Thus, for "Seizure Type(s) and Current Seizure Frequency(ies)"

Medical Reason 1200F-1P

Patient Reason 1200F-2P

Another example of documenting the reason for not performing a measure: *For Counseling for Women of Childbearing Potential with Epilepsy* (Measure 3 above)

To document the medical reason for not performing that measure, append the modifier to the CPT 11 code 4340F as follows: 4340F–1P

The Neurologist could choose the 3 epilepsy measures and apply exclusion to the male patients. Reporting must still be on 50% of eligible patients if reporting via claims and for 80% of eligible patients if through a qualified registry.

If the Neurologist were reporting on epilepsy, *just using one measure*, it is worth re-emphasizing that the *Neurologist must still report this measure on at least 50% of his/her epilepsy patients to avoid the penalty*.

Example #2: A Neurologist chooses to report on measure 126: Diabetic foot and Ankle Care, Peripheral Neuropathy-Neurological Evaluation, *primarily to avoid the PQRS penalty.*

He/she can bill as little as one individual measure to avoid the penalty. However, the physician still must report on at least 50% of eligible patients (those meeting the numerator and denominator criteria outlined in the measure specifications).

Important to know: There are two different types of vender reporting

Direct Vender Reporting through a Direct Qualified EHR Vendor

Data Submission Vendors (DSV): outside vendors used by the EHR

Neurologists using an EHR that is not qualified for EHR direct reporting must submit quality measures through a Data Submission Vendor if they want to use an EHR-based reporting method

Risk of data integrity issues: When converting data from one system to another, there is always the risk of losing information. Reporting through the registry-based or DSV that uses registry submission options could risk data integrity. Be sure to consider this when choosing a PQRS reporting method

Some PQRS reporting methods are more complex than others. Claims-based and group practice reporting options appear to be the most complex

continued on page 19

Treatment of Seizures in Patients with Significant Drug Interactions and Co-Morbidities

Lola Morgan, MD and Jose E. Cavazos, MD, PhD
South Texas Comprehensive Epilepsy Center
University of Texas Health Sciences Center San Antonio

CASE HISTORY: A 66 year old right handed male with history of coronary artery disease and hyperlipidemia managed with aspirin and simvastatin who presents with recurrent episodes of blank stare, garbled speech with loss of consciousness for the past 6 weeks. Neurologic examination is unremarkable. MRI brain reveals moderate deep white matter disease and an EEG reports epileptiform discharges emanating from the left fronto-temporal region.

QUESTION: What factors will influence choice of treatment for seizures in a patient with a history of cardiovascular disease?

EXPERT OPINION: Many patients who have cardiovascular disease (CAD) are managed medically based on clinical recommendations for lipid lowering and use of antiplatelet agents. Because development of epilepsy is common in the elderly, it is useful to be aware of potential interactions for medications utilized for the highly prevalent condition of coronary artery disease.

Review of basic pharmacology provides the framework to understand drug-drug interactions. Drugs undergo biotransformation primarily in the liver along with other tissues such as the intestines, skin, lungs and kidneys, typically aiming to change compounds into more hydrophilic molecules that can be more easily excreted by the kidneys. The biotransformation processes include phase I (oxidation, reduction and hydrolysis) occurring in the subcellular structure of the microsomes and are mediated by the cytochrome P450 family of enzymes. Environmental and genetic factors can significantly influence the activity and result in clinically relevant variations in a patient's metabolism of a drug[1].

Many first generation and several second generation antiepileptic drugs (AEDs) share a common feature of enzyme induction (see Table 1) which can result in an increase in the metabolism of different substrates targeted by the particular cytochrome p450 enzyme along with a decrease in the action of the inducer. Additionally, coadministered drugs can be affected by the process with an acceleration of their own metabolism, a process known as autoinduction[2].

Knowledge of these basic tenets of biotransformation assists in the selection of anticonvulsant therapy in the setting of multiple potential drug-drug interaction. Statins have become a mainstay of treatment for hyperlipidemia. The goal of management of hyperlipidemia is to utilize a statin to lower the low-density lipoprotein cholesterol level to 70 to 100 mg per dL in patients with coronary artery disease[3]. Statins are metabolized by the cytochromic p450 system, in particular the 3A4 family, and would be expected to have reduced serum levels in the presence of an 3A4 enzyme inducing antiepileptic medication. This

Table 1: properties of antiepileptic medications

Enzyme inducing antiepileptic drugs	Non enzyme inducing antiepileptic drugs	Carbonic anhydrase inhibitor activity
phenobarbital		
Phenytoin	levetiracetam	
Carbamazepine	Gabapentin	
Oxcarbazepine Eslicarbazepine	Tiabadine	
Primidone	Pregabalin	
topiramate (weak, usually at doses of 200 mg or greater per day)		topiramate
vigabatrin (?)	Zonisamide	zonisamide
lamotrigine (very weak induction, not clinically significant)	acetazolamide	acetazolomide
rufinamide (weak inducer of 3A4 enzymes)	Lacosamide	
Felbamate	valproic acid ethosuximide Clonazepam Clobazam	

[adapted from Johannessen and Landmark: Antiepileptic Drug Interactions. Current Neuropharmacology, September 2010;8(3): 254-267]

Table 2:

TABLE 65-7 Antiepileptic Drugs Elimination Pathways and Major Effects on Hepatic Enzymes

Antiepileptic Drugs	Major Hepatic Enzymes	Renal Elimination (%)	Induced	Inhibited
Carbamazepine	CYP3A4; CYP1A2; CYP2C8	<1	CYP1A2; CYP2C; CYP3A; GT	None
Ethosuximide	CYP3A4	12–20	None	None
Felbamate	CYP3A4; CYP2E1; other	50	CYP3A4	CYP2C19; β-oxidation
Gabapentin	None	Almost completely	None	None
Lacosamide	CYP2C19	70%	None	None
Lamotrigine	GT	10	GT	None
Levetiracetam	None (undergoes non-hepatic hydrolysis)	66	None	None
Oxcarbazepine (MHD is active oxcarbazepine metabolite)	Cytosolic system	1 (27 as MHD)	CYP3A4; CYP3A5; GT	CYP2C19
Phenobarbital	CYP2C9; other	25	CYP3A; CYP2C; GT	None
Phenytoin	CYP2C9; CYP2C19	5	CYP3A; CYP2C; GT	
Pregabalin	None	100	None	None
Rufinamide	Hydrolysis	2%	CYP3A4 (weak)	CYP2E1 (weak)
Tiagabine	CYP3A4	2	None	None
Topiramate	Not known	70	CYP3A (dose dependent)	CYP2C19
Valproate	GT; β-oxidation	2	None	CYP2C9; GT epoxide hydrolase
Vigabatrin	None	Almost completely	CYP2C9	None
Zonisamide	CYP3A4	35	None	None

CYP, cytochrome P450 isoenzyme system; GT, glucuronyltransferase.
Data from Faught,[30] Leppik,[31] Patsalos et al.,[32] Halford and Lapointe,[33] Cada et al.,[34] and Sabril [package insert].[35]

Table 65-7 from Rogers S and Cavazos JE. Chapter 65: Epilepsy. In Dipiro J, Talbert RL, Yee G, Matzke G, Wells B, Posey LM (eds.): Pharmacotherapy: A Pathophysiologic Approach, 8th Edition, pp 979-1006. McGraw-Hill, 2010.

interaction has been demonstrated between atorvastatin and phenytoin where bioavailability of atorvastatin was reduced by phenytoin coadministration. It was observed that dose adjustment may be required to maintain adequate atorvastatin exposure when coadministered with phenytoin. It has also been proven in the combination of simvastatin and carbamazepine. Atorvastatin and Simvastatin are metabolized by the 3A4 iso-enzyme family of the cytochromic p450 system. Carbamazepine and phenytoin are inducers of 3A4 (see table 2). In contrast, lamotrigine is not an inducer or inhibitor of the 3A4 iso-enzyme family. No interaction between atorvastatin and lamotrigine was observed when both medications were co-administered[4]. A less effective statin, pravastatin is unaffected by 3A4 induction. Because of the importance of statin use in patients with coronary artery disease and stroke, knowledge of the properties of the antiepileptic medications is essential for appropriate selection or alternatively, dosing of the statin to counteract the interaction.

Antiplatelet therapy is an important component of CAD management because platelet aggregation at atherothrombotic plaque sites can produce clinically significant thrombosis and resultant MI. The most common antiplatelet agents used in the United States are aspirin and clopidogrel[5]. Aspirin has several important interactions with antiepileptic medications with which a clinician should be aware. Of note, salicylates may enhance the adverse effect of zonisamide, topiramate and acetazolamide resulting in an increase in the metabolic acidosis which can be observed during use of these medications. All three anticonvulsants share carbonic anhydrase inhibition. Clinical symptoms of metabolic acidosis include drowsiness, hyperventilation, vomiting, confusion and lethargy. The onset may take days to weeks to manifest.[6,7] The mechanism of this interaction is unclear. Salicylates appear to reduce carbonic anhydrase inhibitor protein binding and decrease carbonic anhydrase inhibitor excretion by the kidneys. In addition, carbonic anhydrase inhibitor-induced decreases in plasma pH might result in a higher concentration of nonionized salicylate, which can more readily enter the central nervous system resulting in clinical symptoms[8,9]. While the effect appears to be dose dependent so that cardiac patients taking low dose aspirin are at lesser risk, it is still advisable that the combination of aspirin and an antiepileptic medication containing the carbonic anhydrase inhibitory mechanism be avoided. If this is not possible, close monitoring of metabolic acidosis is warranted.

Aspiring additionally can have effects on other anticonvulsant medications. In particular, the serum level of valproic acid can be increased resulting in clinical symptoms of toxicity[10].

It has been noted that aspirin may increase the serum level of phenytoin; however, little change in the free fraction of phenytoin is observed. Therefore, no symptoms of toxicity are observed. It is advisable to monitor both a free and total fraction when checking levels of phenytoin[11].

It is notable that clopidogrel does not have any clinically significant interactions with any of the anticonvulsant

medications. This absence of interactions is noted for both enzyme inducing and non enzyme inducing antiepileptic medication.

CASE HISTORY: A 45 year old right handed male presents to the emergency room after having a witnessed seizure described by her spouse as eye deviation to the left, head version to the left followed by a fall with a generalized seizure. The patient underwent brain MRI which revealed a mass in the right frontal lobe described as a hyperintense lesion on T2 surrounded by vasogenic edema. A brain biopsy is performed and histology reveals glioblastoma multiforme. He is seen by a neuro-oncologist who plans for resection followed by radiochemotherapy.

QUESTION: What is the best selection of antiepileptic medication for the patient while he is receives definitive treatment?

EXPERT OPINION: Glioblastoma multiforme accounts for 50-60% of all primary brain tumors in adults and carries a median life expectancy of 15 months[12]. Seizures occur in 30-50% of patients with glioblastoma multiforme and they remain at increased risk of recurrent seizures[13]. Seizure control is an important issue in care in neuro-oncology and influences quality of life. Careful selection of an antiepileptic medication regimen can optimize clinical outcomes.

Current standard of care for glioblastoma multiforme consists of surgical resection (if possible) and radiation with adjuvant and concomitant treatment with temozolamide[14,15,16]. A retrospective study of glioblastoma patients (of which 35% were treated with temozolamide) noted that patients receiving non-enzyme inducing medications (primarily valproic acid) demonstrated both improved survival and greater hematologic toxicity as compared to those patients receiving enzyme-inducing antiepileptic drugs[17]. The authors proposed that this difference could result from the lack of enzyme induction in the primarily valproic acid group or enzyme inhibition by valproate (i.e., increased chemotherapeutic agent concentrations and effects) or some combination of these effects. According to temozolomide prescribing information, temozolomide oral clearance is an average of 5% lower with concurrent valproic acid[18].

A subsequent analysis was performed to assess whether antiepileptic drugs modulate the effectiveness of temozolomide and resulting survival. Patients receiving valproic acid had more thrombocytopenia and leukopenia than patients without an antiepileptic drug or patients taking an enzyme inducing antiepileptic drug only. The overall survival of patients who were receiving an antiepileptic drug at baseline versus not receiving any antiepileptic drug were similar. Patients receiving valproic acid alone appeared to derive more survival benefit from temozolomide and radiotherapy than patients receiving an enzyme inducing antiepileptic drug or patients not receiving any antiepileptic drug. The findings suggest that valproic acid may be preferred over an EIAED in patients with glioblastoma who require an antiepileptic drug during temozolomide-based chemoradiotherapy[19]. Future studies are needed to determine whether valproic acid increases temozolomide bioavailability or acts as a sensitizer for radiochemotherapy. The results conclude that selection of antiepileptic drug in patients with glioblastoma should be carefully considered because it may affect survival. The findings also favor the use of non-enzyme inducing antiepileptic medications to allow use of modern chemotherapy that often show increased hepatic metabolism if patients are given an antiepileptic drug which is an enzyme inducer. Common medications used in this setting include levetiracetam because its availability in oral and intravenous formulation, and comparatively a lack of drug interactions. Valproate is an alternate choice.

References

1. Brodie,M et al. Enzyme Induction in Antiepileptic Drugs: A Cause for Concern? Epilepsy, 54(1):11-27, 2013.
2. Katsung, B. Basic and Clinical Pharmacology. Chapter 4. McGraw-Hill Medical;11 edition July 31, 2009.
3. Fuster V, Badimon L, Badimon JJ, Chesebro JH. The pathogenesis of coronary artery disease and the acute coronary syndromes (1). *N Engl J Med.* 1992;326(4):242–250.
4. Bullman J, et al. Effects of lamotrigine and phenytoin on the pharmacokinetics of atorvastatin in healthy volunteers. Epilepsyia 2011 jul;52(7): 1351-8.
5. Fraker TD Jr, Fihn SD, Gibbons RJ, et al. 2007 chronic angina focused update of the ACC/AHA 2002 Guidelines for the management of patients with chronic stable angina: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines Writing Group to develop the focused update of the 2002 Guidelines for the management of patients with chronic stable angina [published correction appears in *Circulation.* 2007;116(23):e558]. *Circulation.* 2007;116(23):2762–2772.
6. Cowan RA, Harnell GG, Lowdell CP, et al, "Metabolic Acidosis Induced by Carbonic Anhydrase Inhibitors and Salicylates in Patients With Normal Renal Function," *Br Med J,* 1984, 289(6441):347-8.
7. Rousseau P and Fuentevilla-Clifton A, "Acetazolamide and Salicylate Interaction in the Elderly: A Case Report," *J Am Geriatr Soc,* 1993, 41(8):868-9.
8. Anderson CJ, Kaufman PL, and Sturm RJ, "Toxicity of Combined Therapy With Carbonic Anhydrase Inhibitors and Aspirin," *Am J Ophthalmol,* 1978, 86(4):516-9.
9. Sweeney KR, Chapron DJ, Brandt JL, et al, "Toxic Interaction Between Acetazolamide and Salicylate: Case Reports and a Pharmacokinetic Explanation," *Clin Pharmacol Ther,* 1986, 40(5):518-24.
10. Goulden KJ, Dooley JM, Camfield PR, et al, "Clinical Valproate Toxicity Induced by Acetylsalicylic Acid," *Neurology,* 1987, 37(8):1392-4.
11. Prescribing information. Dilantin (phenytoin). New York, NY: Parke-Davis, October 2011.

12. Hess KR, Broglio KR, Bondy ML. Adult glioma incidence trends in the United States, 1977-2000. *Cancer.* Nov 15 2004;101(10):2293-9.

13. Hildebrand J, Lecaille C, Perennes J et al (2005) Epileptic seizures during follow-up of patients treated for primary brain tumors. *Neurology* 65:212–215.

14. Stupp R et al. Radiotherapy plus Concomitant and Adjuvant Temozolomide for Glioblastoma. NEJM 2005; 352:987-996.

15. Stupp R, et al. Effects of radiotherapy with concomitant and adjuvant temozolomide versus radiotherapy alone on survival in glioblastoma in a randomised phase III study: 5-year analysis of the EORTC-NCIC trial.Lancet Oncol. 2009;10(5):459.

16. Athanassiou H, et al. Randomized phase II study of temozolomide and radiotherapy compared with radiotherapy alone in newly diagnosed glioblastoma multiforme. J Clin Oncol. 2005; 23(10):2372.

17. Oberndorfer S, Piribauer M, Marosi C, et al, "P450 Enzyme Inducing and Non-Enzyme Inducing Antiepileptics in Glioblastoma Patients Treated with Standard Chemotherapy," *J Neurooncol*, 2005, 72:255-60.

18. Prescribing information. Temodar (temozolomide). Kenilworth, NJ: Schering Corp., May 2007.

19. Weller M, et al. Prolonged survival with Valproic Acid Use in the EORTC/NCIC temozolomide study. Neurology 77 (12); 1156-1164 Epub 2011, Aug 11.

20. Johannessen and Landmark. Antiepileptic drug interactions. Current neuro-pharmacology 2010 Sept; 8(3): 254-267.

Treatment of Seizures in Patients

continued from page 17

There may be costs involved in registry-based reporting and data submission vendor reporting (DSV). EHR direct reporting usually do not involve additional cost

A list of 2013 Qualified EHR Direct Vendors and Qualified Registries is available on the CMS website. In addition, the AAN has been very proactive in trying to simplify the entire PQRS process. The AAN has partnered with CECity, as one registry reporting option to help meet PQRS requirements. This spring CECity is offering a discount to AAN members. CECity also has the ability to report on all the neurology related measures. More information can be found on the AAN website. In addition, the AAN toolkit for PQRS might be helpful and can be found at www.aan.com/go/practice/pay/pqrsguide.

Author's Note: The PQRS program has proven to be daunting to many colleagues who try to understand its requirements and implement the measures according to the guidelines. It is hoped that this condensed but comprehensive review of PQRS will be of benefit to our TNS membership. 17% of Neurologists participated in the PQRS program in 2010. In 2010 PQRS incentive payments for all eligible providers totaled $391,635,495 which was paid to 169,843 practitioners. The average incentive was $2,157 per practitioner. More detailed instructions on PQRS are published on the CMS and Neurology Web sites:

http://www.cms.gov/pqrs

http://www.aan.com/go/practice/pay/resources.

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

PRESORTED
FIRST-CLASS MAIL
U.S. POSTAGE PAID
AUSTIN, TX
PERMIT NO. 1787

Texas Neurological Society's 2013-2014 Officers

President
G. Mark Schwartze, MD, Waco
president@texasneurologist.org

President-Elect
Kim E. Monday, MD, Houston

Vice President
Robert F. Leroy, MD, Dallas

Secretary-Treasurer
Deborah Carver Hodges, MD,
San Antonio

Immediate Past President
Sara G. Austin, MD, Austin

Members-at-Large
Waleed El-Feky, MD, Dallas
Erin E. Furr-Stimming, MD, Houston
Ed Fox, MD, PhD, Austin
Aziz Shaibani, MD, Houston

Education Committee Chair
Robert W. Fayle, MD, Livingston

Program Director
Summer Conference 2013
Ed Fox, MD, PhD, Austin

Winter Conference 2013
Aziz Shaibani, MD, Houston

Delegate to TMA
William S. Gilmer, MD

Alternate Delegate to TMA
Ed Fox, MD, PhD, Austin

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

Executive Director
Ky Camero
401 W. 15th St.
Austin, TX 78701
exec@texasneurologist.org