TEXAS NEUROLOGICAL SOCIETY WINTER 2019

Broca's Area

The Voice of Texas Neurology

A message from the President

TNS is proud to be part of the AAN's efforts to protest the new CMS proposal to collapse the E/M codes levels 2-5 which would have taken away about $43 million annually in Medicare payment for E/M services from neurologists. CMS agreed to delay the implementation until 2021. This delay demonstrated the importance of advocacy by professional organizations like the AAN and TNS. TNS signed on to the AAN's petitions letter. Beside national efforts, we are working at the state level to tackle important legislative issues that concern neurological care and practice in Texas.

Escalating drug prices have become a national and state issue alike. The AAN has taken great efforts to establish a pilot program to develop an international pricing index to align pricing with that in peer nations. This is a program that TNS supports.

Regulatory overburden is an important cause of neurologist burnout. It is partly related to preauthorization and electronic medical records requirements and similar issues that distract neurologists from what they do the best: patients care. We are inviting experts to our summer and winter conferences to talk about these issues and we are working locally and nationally to improve our working conditions.

Our conferences continue to be very well attended and attract national experts as speakers. Your evaluation remarks have helped us to improve these programs so please continue to send us your feedback.

The TNS membership has increased by 220 new members mostly in response to our discounted group rate to the academic programs. That is about a 25% increase of the active membership. Almost all university neurology programs took advantage of the offer which enriches and diversifies our membership. This will also be reflected in the quality of the programs, advocacy and leadership provided by and to TNS.

In the last board meeting, board members voted to offer patients support groups the opportunity to apply for a TNS grant. This grant would help in their efforts to advance patients awareness of neurological diseases. The beginning of a new relationship between the TNS and patient support groups will increase awareness of TNS' advocacy on behalf of our patients. $5000 has been allocated in the 2019 budget for this grant.

After an intense recruitment campaign, the board hired a new lobbyist, Tom Holloway. He will represent TNS on behalf of its membership (as directed by the board) during the next state legislative session. Look to the legislative update found later in this addition of Broca's for more details.

Another issue the board addressed was opening up board member-at-large nominations to the membership beginning later in 2019 for the 2020 board. The nomination committee will screen the nominations and present its selection to the board for voting. This democratic process will give every member the right to nominate him/herself or others to ensure wide representation.

Aziz Shaibani, MD

Nominations for the 2020 TNS Lifetime Achievement award was circulated to the membership earlier this fall. The board will be making its final decision on the recipient in February. The 2019 awardee, Dr. Gary Clark, is an original contributor to neurology literature and patients care and to the TNS. He is the head of a world renowned pediatric neurology program at Texas Children's hospital and has lead excellent pediatric neurology TNS sessions over the years.

We continue to prosper and be the society that other neurology societies in the nation want to emulate. That brings a lot of pride to our work and activism. This could only happen with the support and efforts of the TNS members, board, committees, advocacy group and our executive director, Ky Camero. I would like to profusely thank them all for their vision, time and efforts.

I am concluding my year with satisfaction and gratitude.

CONTENTS

We've gone digital!

OPEN THIS NEWSLETTER IN FIREFOX OR GOOGLE CHROME FOR THE BEST INTERACTIVE EXPERIENCE.

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 22nd Annual TNS Winter Conference at the Hyatt Regency Austin. Yessar Hussein, adult program director, Gary Clark, pediatric program director, Bob Fayle, education committee chair, and the education committee have planned an excellent program.

THE FIRST FOOTBALL CONCUSSION CRISIS.

The first football concussion crisis occurred over 100 years ago when a significant number of concussions and deaths resulted in recommendations to outlaw the game or at least make it safer (Harrison EA. The first concussion crisis: head injury and evidence in early American football. Am J Public Health. 2014;104(5):822-833). After at least 45 football players died from injuries sustained in football form 1900 to October, 2005 (which the "Chicago Tribune" called college football's "death harvest"), a number of schools including Columbia, Duke, Northwestern, Stanford, Cal, and Army cancelled football.

President Roosevelt saved football when he had a private meeting with the coaches of the three biggest college programs (Harvard, Yale, and Princeton) urging them to save the sport by reducing the violence (Miller JJ. The big scrum: how Teddy Roosevelt saved football. Harper Collins, 2011) The Intercollegiate Athletic Association passed 19 new rules in 1906 to try and make football safer including creating a neutral zone, stopping the game when a player fell on the ball, banning the flying wedge, requiring 10 yards rather than 5 for a first down, and establishing the forward pass.

After he was kicked and hit in the head in the Army-Navy game of 1893 and a Navy doctor told him this could result in instant insanity or death, Reeves (later to be an admiral and "the father of carrier aviation") invented the first football helmet (a moleskin hat with earflaps) (Stamp J. Leatherhead to radio-head: the evolution of the football helmet. October 1, 2012. Smithsonian.com. available at https://www.smithsonianmag.com/arts-culture/leatherhead-to-radio-head-the-evolution-of-the-football-helmet-56585562/). The NCAA made helmets mandatory in 1939 and the NFL required helmets in 1943.

Nichols and Smith, surgeons who were in charge of the Harvard football squad, performed the first epidemiological study of concussion in football during the 1905 season (Nichols EH, Smith, HB. The physical aspect of American football. Boston Medical and Surgical Journal. 1906;154:1-8). "Cases of concussion were frequent, both during practice and games. In fact, but two games were played during the entire season in which a concussion of the brain did not occur. …Players who had concussion were at once carefully examined to exclude the possibility of middle meningeal hemorrhage. …the injured men were in every case compelled to go to the infirmary, where they remained over night. … Concussion was treated by the players as a trivial injury and rather regarded as a joke. The real seriousness of the injury is not certain. Our own experience with the after-effects of the cases if not sufficient for us to draw any definite conclusions, but from conversation with various neurologists, we have obtained very various opinions in regard to the possibility of serious after-effects."

A SURVEY OF NEUROLOGISTS ON POSTCONCUSSION SYNDROME.

Many thanks to those of you who participated in our survey on postconcussion syndrome (Evans RW, Ghosh K. A Survey of Neurologists on Postconcussion Syndrome. Headache. 2018 Jun;58(6):836-844). The abstract is the following:

BACKGROUND AND OBJECTIVES:

Postconcussion syndrome (PCS) has been controversial for more than 150 years. As there have not been any surveys of PCS among neurologists in the United States since 1992, another was performed using most of the prior items to assess current opinions and practices and whether there have been any changes since 1992.

METHODS:

Two hundred and eighty-nine neurologists attending the Texas Neurological Society 20th Annual Winter Conference continuing medical education meeting in 2017 were supplied the survey instrument with registration materials. The 25 item instrument (including 7 new items) contained items on demographics, definitions, causation, prognosis, medicolegal aspects, testing, and treatment. Forty percent of attendees completed the survey.

RESULTS:

The majority of respondents agree with the following: PCS is a clearly defined syndrome with a solid basis for determining prognosis with an organic basis; accept the authenticity of patients' reports of symptoms; effective treatment is available for headaches lasting 3 months or more; headaches persist in over 20% 1 year after injury; and cognitive rehabilitation is effective. The majority of the respondents do not agree with the following: symptoms improve in a relatively short period of time and quickly resolve once litigation is settled; effective treatment is available for PCS; and return to play guidelines are strongly evidence based. 68.4% disagree with the following: I would support my son or grandson (or if you do not have one,

← BACK TO PAGE 1
NEXT PAGE →

1905 Harvard-Yale foot ball game (from https://www.worthpoint.com/worthopedia/1905-harvard-yale-football-game-153116757)

relative's or friend's) playing football. CONCLUSIONS: There has been growing acceptance of the organicity of PCS among neurologists in the last 25 years. There is significant concern over the long-term sequelae of concussion as most respondents would not recommend that their son or grandson play football.

THE RETIRED NFL PLAYERS CONCUSSION SETTLEMENT.

The NFL settlement with former players (20,517 registered) who accused he League of failing to warn and protect players against long-term neurological risk of repetitive head trauma became effective on January 7, 2017. The benefits include "monetary awards for diagnoses of Death with CTE before April 22, 2015 (the Final Approval Date), ALS, Parkinson's Disease, Alzheimer's Disease, Level 2 Neurocognitive Impairment (i.e., moderate Dementia) and Level 1.5 Neurocognitive Impairment (i.e., early Dementia) (see Injury Definitions). All valid claims under the Settlement, without limitation, will be paid in full throughout the 65-year life of the Settlement." (NFL Concussion Settlement. In RE: National Football League Players' Concussion Litigation. No. 2:12-md-02323 (E.D. Pa). Available at https://www.nflconcussionsettlement.com/Un-Secure/FAQDetails.aspx?q=2#2. Accessed on November 24, 2018.

Fifteen neurologists in Texas are qualified to perform Monetary Award Fund (MAF) examinations.

There has been controversy over application of the criteria for cognitive impairment (Hurley D. For Your Patients-Concussion: Why Some Neurologists Are Calling 'Foul' Over Criteria for NFL Concussion Settlement. Neurology Today. 18(19):1,38-39, October 4, 2018; Letters to the Editor: Criteria for NFL Concussion Assessment. Neurology Today. 18(22):6, November 15, 2018).

To determine level 1.5 ("early dementia) or 2.0 ("moderate dementia") neurocognitive impairment, the neurologist uses the history, neurological exam, and neuropsychological testing. In addition, the retired player must exhibit functional impairment general consistent with the criteria in the Clinical Dementia Rating Scale (CDR) in the areas of community affairs, home & hobbies, and personal care which results from cognitive loss and not other causes (such as a physical handicap or injury, chronic pain, or sleep apnea). A sworn statement of a third party is used to corroborate the retired players' functional impairment when there are no other documents available to corroborate. The diagnostic criteria, testing and documentation must be generally consistent. The neurologist is to use "his or her best judgment" in assigning CDR scores and sound medical judgement and best practices in assigning impairment ratings.

← BACK TO PAGE 1
NEXT PAGE →

Editor's Notes *(cont.)*

This process is much different from the one we use to determine cognitive impairment in clinical practice where we obtain a history from the patient and sometimes family members and friends, perform a neurological exam including the MMSE or MoCA, and obtain testing which may include blood tests, a scan of the brain, and neuropsychological testing. Blood tests, a scan of the brain, and polysomnograpy to exclude other causes of cognitive impairment are not done as part of the retired player's evaluation.

The settlement only used the last 3 parts of the CDR (not the first 3 parts). Although the CDR has been validated for use in Alzheimer's disease (Rikkert MG, Tona KD, Janssen L, Burns A, Lobo A, Robert P, Sartorius N, Stoppe G, Waldemar G. Validity, reliability, and feasibility of clinical staging scales in dementia: a systematic review. Am J Alzheimers Dis Other Demen. 2011;26(5):357-65), the use of the last 3 parts of the CDR have not been validated in this population of retired players. There is significant potential for under and overreporting of functional impairment with the required use of the CDR and a clinical affidavit.

The program has audited 57% of claims. The appeals claim that persons with level 2 do not drive and do not or cannot work. However, this is not accurate since about 1/3 of people with Alzheimer's disease drive and some people with mild Azheimer's disease still work. MMSE scores above 24 have been alleged to be inconsistent with cognitive impairment which is clearly inaccurate as the test may be influenced by age and education and is not sensitive for mild dementia. In a study of 304 consecutive referrals to a university based outpatient memory clinic, there were 70 subjects with MMSE scores of 30/30 or 29/30. 43% were found to have moderate to severe memory impairment (Lacy M, Kaemmerer T, Czipri S. Standardized Mini-Mental State Examination Scores and Verbal Memory Performance at a Memory Center Implications for Cognitive Screening. Am J Alzheimer's Disease & Other Dementias 2015;30:145-152).

BrownGreer, the MAF settlement administrator, states, "You should bill the Player or his insurance for the services performed as you would for any other patient." (NFL concussion settlement program: qualified MAF physician manual. As of 3/1/18). However, billing the claimant's insurance is highly problematic. Insurance may not cover the medico-legal encounter. Even if covered, the time involved in learning about the rules for the determination, the encounter (which may include reviewing extensive prior medical records, obtaining and scoring the Clinical Dementia Rating and third party affidavit, and current neuropsychological testing), determination of a compensable diagnosis, if any, and submission of materials is much more than insurance compensation would cover.

In addition, neurologists should be wary of establishing a physician-patient relationship with a claimant they will probably see once. Many of the claimants have major depression and suicidal ideation, significant cognitive impairment or unrelated medical co-morbidities. Neurologists could potentially be sued by the retired player or their families for failure to diagnose or treat. Examples might be suicide, vascular complications of untreated hypertension, cognitive impairment due to another undiagnosed condition, or poor decision making due to cognitive impairment. The neurologists and the claimants may be better served by informing the claimants at the time of their visit that they are there for an independent medical examination and there is no physician-patient relationship.

TNS RECOMMENDS A RETROSPECTIVE STUDY OF THE CLAIMANTS IN THE RETIRED NFL PLAYERS' CONCUSSION SETTLEMENT.

Although the NFL is funding concussion research (such as a safer helmet), there are no plans to analyze the results of the neurological and neuropsychological evaluations of claimants. TNS recommends the following:

> Texas Neurological Society press release
> June 20, 2018
>
> Traumatic brain injury in sports is a major public health issue in the United States as there are up to 3.8 million sports related mild traumatic brain injuries per year. The long term consequences are poorly understand.
>
> There is growing concern over the risk of chronic traumatic encephalopathy (CTE) in contact sports which has been reported in football players as young as 17. There are approximately 3 million children ages 6-18 years playing football (including 1 million high school players), 70,000 collegiate players, and 1,696 in the NFL.
>
> There are 20, 504 retired football players who have registered for the NFL Concussion Settlement. The NFL has announced that they will pay for concussion research. We strongly encourage the NFL, with permission from the retired players, to pay for an independent retrospective study to be performed by independent neurology experts to assess how many have cognitive impairment, Parkinson's disease, amyotrophic lateral sclerosis, and other neurological disorders and what the risk factors may be.

If you would like to submit an article for *Brocas*, deadline for the Spring edition is May, and for the Winter edition is December. All membership is invited to participate.

← BACK TO PAGE 1
NEXT PAGE →

TNS Advocacy Report

Fall 2018

Sara Austin, MD, Legislative Chair

Tom Holloway, TNS Lobbyist

CHANGING OF THE GUARD

First and foremost, we're sad to announce that our longtime lobbyist, Greg Herzog, has accepted a position as the Director of Government Affairs for a major biopharmaceutical company and will no longer be representing the Texas Neurological Society at the Texas Legislature. Those of you who had the opportunity to work with Greg over the years know what a great person he is and how hard he worked to advance the interests of neurology at the Capitol. Though we're sad to see him go, we wish him all the best in his new endeavor.

Consequently, we are pleased to announce that Tom Holloway will be taking over as the Texas Neurological Society's new government affairs consultant as we prepare for the 86th Legislative Session. Tom is a veteran legislative staffer who has spent the last 12 years working in the Texas House and Senate. For the past seven years, he has served as the chief of staff to Senator Charles Schwertner, MD – a practicing orthopedic surgeon and Chairman of the Senate Committee on Health and Human Services. We feel very fortunate to have such an experienced advocate working on our behalf this session, and I know Tom is eager to meet each of you at the upcoming Winter Conference and at First Tuesdays during the legislative session.

THE MIDTERM ELECTION AND ITS IMPACT ON TEXAS

As many of you know, November's midterm elections shattered all expectations for voter participation in Texas and sent shockwaves through state's political landscape. In the end, we saw more than 8.3 million Texans turn out to cast their ballots – a staggering number that nearly doubled the record-setting turnout from the prior midterm election in 2014 (4.9 million) and fell just short of capturing the overall turnout record (8.9 million) set during the 2016 presidential race.

This result was largely attributed to growing voter dissatisfaction with President Trump, as well as a well-funded and energetic campaign engineered by longshot Democratic hopeful Beto O'Rourke against Texas' junior U.S. Senator, Republican Ted Cruz. As the final votes were tallied, O'Rourke exceeded all expectations to come within 225,000 votes of toppling Cruz and ending a streak of statewide Republican dominance that has endured for more than 20 years. Even in defeat, O'Rourke's coattails lifted Texas Democrats to unexpected victories across the state…most notably in the halls of the Texas Legislature, where Democrats succeeded in flipping two seats in the 31-member Texas Senate, and an astonishing 12 seats in the Texas House of Representatives.

SHIFTING PRIORITIES AND A CHANGE IN TONE

As we head into the upcoming legislative session in beginning in January, the Texas House now stands at a significantly diminished 83-67 Republican majority – still solidly Republican to be sure, but a far cry from the 100+ supermajority the party enjoyed only a few years ago. Perhaps even more significant, the Senate Democrats now find themselves just one vote shy of the margin needed to block consideration of legislation on the floor of the Texas Senate – significantly upsetting the balance of power in the state's conservative upper chamber. These new developments mean that Republican leaders in both the House and Senate will likely have to pursue a greater degree of compromise with members across the aisle if they hope to advance an agenda through Texas Legislature in 2019.

Yet, the most interesting result from Election Night was perhaps a less obvious one: the stark and somewhat surprising difference between the electoral performance of more socially-conservative Tea Party Republicans and their more moderate, business-minded Republican counterparts.

While popular center-right Republicans like Governor Greg Abbott, Comptroller Glenn Hager, and Land Commissioner George P. Bush all garnered relatively strong support from the voters (55.8%, 53.2%, and 53.7%, respectively), statewide incumbents known more for their rigid, ideologically-conservative views like Lt. Governor Dan Patrick (51.3%), Attorney General Ken Paxton (50.6%), and Agriculture Commissioner Sid Miller (51.3%) faired significantly worse – with some managing to eek out the narrowest of victories against largely unknown and underfunded Democratic challengers.

These closer-than-expected statewide results, taken with the somewhat dramatic partisan political shift in the House and Senate, seem to indicate a significant rebuke from the voters over hot-button social issues like the so-called "bathroom bill" from last session, and may signify a coming realignment of Republican political priorities toward more practical, middle-of-the-road issues like fixing the Texas' ailing school finance system, addressing the state's troublingly high uninsured population, and finding sensible solutions to reducing local property taxes.

← BACK TO PAGE 1

NEXT PAGE →

TNS Advocacy Report (*cont.*)

A NEW SPEAKER OF THE TEXAS HOUSE (WITH NEUROLOGY CONNECTIONS!)

When current House Speaker Joe Straus (R-San Antonio) announced last October that he planned to step down at the end of 2018, it sent tongues wagging across Austin about who would hold the Speaker's gavel when in the 2019 Legislative Session. One by one, more than half dozen candidates entered the race to succeed Straus, though none appeared to enjoy the broad support necessary to claim an outright victory.

The crowded field seemed to open the door for a group of 25-30 center-right Republicans to unite behind an effort to draft veteran lawmaker Dennis Bonnen (R-Angleton) into the race to serve as the next leader of the Texas House. A few short weeks later, Bonnen announced he had the secured the support of 109 of the chamber's 150 members, thereby capturing the Speaker's gavel and demonstrating a clear mandate to lead the Texas House of Representatives in 2019.

In addition to being the first new face to lead the Texas House in a decade, the selection of Dennis Bonnen as the next Speaker of the Texas House is significant for another reason: his brother is Dr. Greg Bonnen, MD (R-League City), the only practicing neurosurgeon serving in the Texas Legislature. As we enter the next legislative session, the Texas Neurological Society should find itself uniquely positioned to use our relationships with both Bonnen brothers to move our agenda forward.

PHYSICIAN FEE SCHEDULE

This year, the Texas Neurological Society will be keeping a close eye on a number of proposals at the state and federal level that have the potential to impact our practices and affect the all-important relationship between physicians and their patients.

First and foremost, we're pleased to report that CMS has agreed to postpone a controversial proposal to collapse the physician fee schedule ($135 for all new patients, $95 for follow-up appointments, regardless of complexity) until at least 2021. Many groups, including the Texas Medical Association and the American Academy of Neurology, lobbied aggressively against this proposed change to the reimbursement schedule and will continue to do so in the future.

In announcing the delay, CMS indicated that postponing implementation of the rule would allow for additional research on the proposal and give regulators a chance to offer further changes – perhaps allowing payment for a level 5 new patient visit, with levels 99202-99204 collapsed into one code.

We now know that the E/M coding and payment structure will remain unchanged in 2019 and 2020. Needless to say, this has been a worrisome conversation for neurologists and we are following it carefully. Neurologists should continue to use either the 1995 or 1997 documentation guidelines to document E/M office visits billed to Medicare until the new rule comes into place in 2021 (if it does actually get approved at that time).

TEXAS MEDICAL BOARD SUNSET LEGISLATION

As we prepare for the 86th Legislative Session in January, the Texas Neurological Society is monitoring a number of legislative developments on the state level – most immediately, ensuring the legislative reauthorization of the Texas Medical Board to continue operations for another 12 years.

By law, every state agency in Texas is required to undergo a thorough, periodic review by the Texas Sunset Commission at least once every 12 years to maintain their statutory authority. This process allows legislators the opportunity to conduct a thoughtful evaluation of each agency, offering wholesale reforms to the agency's management and operations if necessary. If the Sunset bill for a particular agency should fail to pass, the agency loses its statutory authority and is "sunsetted" out of existence.

Last session, the legislature failed to pass the Texas Medical Board Sunset Bill, consequently placing the Texas Medical Board and the medical license of every Texas physician in jeopardy. In a last minute attempt to avert disaster for every licensed physician in Texas, the legislature effectively "kicked the can down the road" by opting to delay the effective Sunset date of the Texas Medical Board until 2019.

This session, the Texas Neurological Society will be working with TMA and other medical specialty groups to pass the Texas Medical Board Sunset Bill and reauthorize the board for another 12 years.

DO-NOT-RESUSCITATE ORDERS & DIAGNOSING BRAIN DEATH

Last session, the Texas Legislature passed legislation to place a number of new conditions on the issuance of a valid do-not-resuscitate order, including establishing specific notification protocols, creating a process to revoke a valid DNR order issued by a physician, and outlining circumstances under which a physician could be held civilly or even criminally liable.

This session, the Texas Neurological Society be working to oppose any legislation that seeks to create further legal liability or disruption to the process of issuing a do-not-resuscitate order in circumstances where continuation of futile medical treatment would violate a physician's sense of medical ethics and prolong the pain and suffering of their patient. TNS will also be working to oppose any legislation that interferes with a physician's ability to exercise their independent medical judgment with regard to establishing a diagnosis for their patients – particularly as it relates to the diagnosis of "brain death."

OPIOID ABUSE & THE TEXAS PRESCRIPTION MONITORING PROGRAM

As most of you are aware, the Texas Prescription Monitoring Program (PMP) is an electronic database maintained by the Texas State Board of Pharmacy to monitor prescription data for controlled substances. Under a law passed in 2015, physicians will soon be required to clear all prescriptions for Schedule II, III, IV, and V drugs through the Texas Prescription Monitoring Program. As currently constituted, this mandate threatens to

← BACK TO PAGE 1
NEXT PAGE →

TNS 2019
Summer Conference

HOUSTON MARRIOTT MARQUIS

JULY 26-27

There will be nationally recognized speakers addressing the microbiome and the gut-brain axis, advances in genetic testing and headache secondary to intracranial hypotension.

Other topics include clinically relevant updates in Psychiatry and Narcolepsy along with practice management recommendations addressing maximizing revenue opportunities and technology to improve your practice.

Thank You to our 2018 TNS Summer Conference Supporters

GOLD

CSL Behring
Mitsubishi Tanabe Pharma America
Sunovion Pharmaceuticals
UCB, Inc.

SILVER

Acadia Pharmaceuticals
Eisai, Inc.
Sanofi Genzyme
US WorldMeds

BRONZE

Amgen
Medtronic
PHRK Intervention Inc., dba Southside Device, LLC
Supernus Pharmaceuticals, Inc
Validus Pharmaceutical LLC

dramatically slow the ability of physicians to treat patients and write prescriptions in a timely manner.

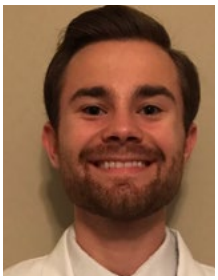
While the Texas Neurological Society is supportive of the overarching goals and purpose of the Texas Prescription Drug Monitoring Program, we have practical concerns about the impact its implementation will have on physician practices. TNS supports the requirement to report all Schedule II drugs to the PMP beginning in 2019, as scheduled, but will seek to delay reporting of lower classification Schedule III, IV, and V drugs for an additional two years in order to allow the legislature to monitor implementation and make any necessary adjustments to the program or software during the 2021 Legislative Session.

MEDICAL MARIJUANA & COMPASSIONATE CANNABINOID USE

In 2015, the Texas Legislature passed the Texas Compassionate Use Act – the state's first law permitting the limited application of medical cannabinoid oil to treat patients diagnosed with severe, intractable epilepsy. This law permits the highly regulated sale of oils with low levels of tetrahydrocannabinol (THC), the psychoactive element in marijuana, and high levels of cannabidiol (CBD), a non-euphoric component used to treat epilepsy and other chronic medical conditions.

Considering the relative success of the 2015 Compassionate Use Act law and the rapidly changing public opinion regarding medical marijuana, we anticipate a host of bills to be filed this session that will seek to expand the use of medical marijuana and other cannabinoid derivatives to treat additional medical conditions. The Texas Neurological Society intends to closely monitor legislation related to medical cannabis, including efforts to expand the Texas Compassionate Use Act to include additional medical conditions not currently covered under the relatively narrow limitations of the current law. TNS is particularly interested in the potential application of CBD oil and other cannabinoid derivatives to treat a host of conditions involving involuntary muscular spasticity, such as epilepsy, multiple sclerosis, and ALS.

← BACK TO PAGE 1
NEXT PAGE →



An Atypical Presentation of Spontaneous Intracranial Hypotension

Ryan P. Coburn
Medical student, University of Texas McGovern School of Medicine, Houston, Texas

INTRODUCTION

Spontaneous intracranial hypotension (SIH) is due to leakage of cerebrospinal fluid (CSF) from a leak in the dura mater, which is almost always spinal, mostly thoracic or at the cervicothoracic junction. An underlying connective tissue disorder can lead to dural weakness and leaks. About 25% of cases are due to ventral dural tears associated with calcified thoracic disc herniations or osteophytes. A CSF-venous fistula (between the CSF and paraspinal veins) is a recently identified cause of SIH. Minor trauma or an inciting event, such as sex or sneezing, can cause leaks though rupture of spinal epidural cysts, perineural cysts, or cause a tear in the dural nerve sheath.

The annual incidence is 5 per 100,000, with a female to male ratio of 2:1. The most common presenting symptom is orthostatic headache. The headache is variable in character, ranging from dull and focal pain, to generalized and incapacitating. Other headache patterns include the following: a non-orthostatic chronic daily headache; a paradoxical headache worse when supine and better upright; thunderclap headache; intermittent CSF leaks which may cause intermittent headaches with headache free intervals; and can mimic primary cough or exertional headache.

The most common associated symptoms, seen in approximately half of patients, are neck pain, nausea, and vomiting. Other associated symptoms include tinnitus, vertigo, dizziness, visual changes, photophobia, and unsteady gait. Rare complications include parkinsonism, quadriparesis, and a decreased level of consciousness, even coma.

MRI of the brain is abnormal in about 80% of cases and may show the following: smooth and diffuse pachymeningeal (dura and arachnoid) enhancement (the most common abnormality), sagging of the brain with cerebellar tonsillar herniation, subdural fluid collection, enlarged venous sinuses, and pituitary enlargement. Spinal MRI may show dilated epidural veins, and extradural or subdural collection of fluid. Others studies to confirm or find a spinal CSF leak are radioisotope cisternography, CT myelography, and MR myelography.

For patients that present acutely, conservative measures may help. These include bed rest, oral hydration, and caffeine (200-300 mg bid or tid). Those that are unresponsive to conservative measures may respond to epidural blood patches (50% require more than one and some may require up to 4-6). The patch may be given at the thoracolumbar junction or lower lumbar area, or at the cervical or thoracic spine if the site of the leak is found. An infusion of 10-20 cc of autologous blood may be tried for the first, and 20-100 cc for subsequent patches if the first does not work, with subsequent blood patches at least 5 days apart. Epidural patching with fibrin glue can be done if the site of the CSF leak can be identified, which may be effective for the approximately 1/3 of patients who failed epidural blood patches. Neurosurgical intervention can be considered for those with a clearly identified source of the CSF leak which is refractory to multiple epidural blood patches.

A frequent complication of CSF leak closure with epidural blood patching (in over half of cases) or surgical closure is rebound intracranial hypertension, where CSF pressure increases above normal levels and leads to a new type of headache. This headache can be mild to severe in intensity and may become worse when recumbent or can be non-positional. Nausea and blurry vision may be present, however papilledema is often absent. The symptoms may be brief but can also persist for weeks to months. Treatment is with acetazolamide or topiramate.

CASE REPORT

A 39 year old right handed male with an unremarkable past medical history presented with new onset headaches for three weeks, described as a generalized pressure with an intensity of 5-6/10, lasting only a few seconds and triggered by head movement. For about 4 weeks, he experienced intermittent "whooshing" sounds in his left ear that were independent of movement. He also experienced orthostatic dizziness. The headache and tinnitus decreased in frequency with supine positioning. He had not experienced any relief with over the counter ibuprofen. He denied any antecedent head or neck trauma, recent illness, history of headaches, or previously experiencing similar symptoms. There were no accompanying symptoms such as neck pain, nausea, or vomiting.

Two weeks prior he saw an ENT and had a normal exam and audiogram. MRI with and without contrast revealed diffuse dural enhancement on T1 with contrast, consistent with SIH. A complex arachnoid cyst left of the falx cerebelli and a developmental venous anomaly of the right cerebellar hemisphere were also noted on the MRI report. MRI scan of the cervical, thoracic, and lumbar spine is pending.

Neurologic exam was normal.

DISCUSSION

The presence of diffuse dural enhancement on brain MRI strongly suggested a diagnosis of SIH. However, the headaches seen in this patient were atypical for SIH, given the necessity of head movement while in the upright position to elicit pain. Common associated symptoms of SIH were absent, such as neck pain, nausea, or vomiting.

← BACK TO PAGE 1
NEXT PAGE →

He also had orthostatic dizziness. Dizziness, changed and decreased hearing, deafness, tinnitus, and orthostatic tinnitus may be due to stretching of the eighth nerve or pressure changes in the perilymphatic fluid of the inner ear.

With the variability in the headaches and associated symptoms, SIH is often under-recognized, with a mean time of 13 months before correct diagnosis is reached. Thus it is important that SIH be considered as a differential diagnosis in patients with headache, even with atypical presentation. Early recognition can prevent unnecessary suffering, as full recovery often occurs with conservative treatment or epidural blood patches.

The author has no conflict of interest.

REFERENCES

Berroir S, B. Loisel B, A. Ducros A, e al, M. Boukobza, C. Tzourio, D. Valade, M-G. Bousser. Early epidural blood patch in spontaneous intracranial hypotension Neurology Nov 2004; , 63 (10): 1950 1951.

Karm M-H, Choi J-H, Kim D, Park JY, Yun HJ, Suh JH. Predictors of the Treatment Response of Spontaneous Intracranial Hypotension to an Epidural Blood Patch. Hanaoka. K, ed. Medicine. 2016;95(18):e3578..

Kranz PG, Gray L, Amrhein TJ. Spontaneous Intracranial Hypotension: 10 Myths and Misperceptions. Headache. 2018;58(7):948-959.

Lin J, Zhang S, He F, Liu M, Ma X. The status of diagnosis and treatment to intracranial hypotension, including SIH. The Journal of Pain. 2017;18(1):4.

Mokri B, Maher CO, Sencakova D. Spontaneous CSF leaks: Underlying disorder of connective tissue. Neurology Mar 2002, 58 (5) 814-816

Schievink WI. Misdiagnosis of Spontaneous Intracranial Hypotension. Arch Neurol. 2003;60(12):1713–1718.

Schievink WI. Spontaneous Spinal Cerebrospinal Fluid Leaks and Intracranial Hypotension. JAMA.2006;295(19):2286–2296.

Primary Dural MALT Lymphoma Mimicking Multiple Meningiomas

Divya Mella MD
University of Texas Southwestern Medical Center, Neuro-Oncology

CASE SUMMARY

A 46-year-old woman developed a tender, subcutaneous mass in the center of her forehead over 9 months. Brain MRI revealed two extra-axial enhancing masses (figure 1) consistent with meningiomas. She underwent a bifrontal craniotomy and removal the left frontal lobe extra-axial tumor. The immunohistochemical profile was consistent with MALT (mucosa associated lymphoid tissue) lymphoma. Staging did not reveal involvement of any other site. Primary dural lymphomas are rare and are usually MALT type B cell lymphomas. Radiologically they are often mistaken for meningiomas. They are characterized by indolent behavior and favorable response to treatment. [1, 2]

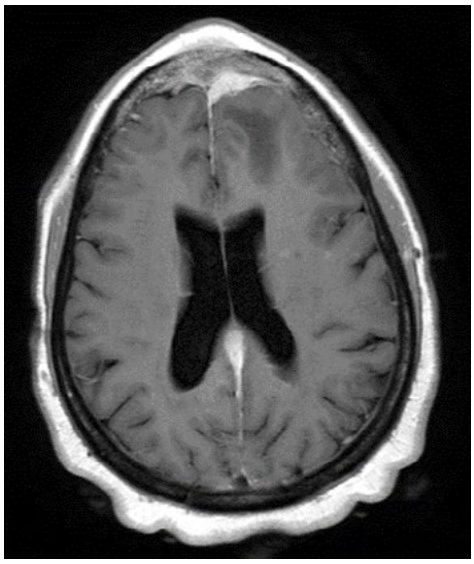


Figure 1: Gadolinium enhanced axial T1-weighted MRI shows 2 extra-axial enhancing masses: one along the left frontal convexity with intraosseous involvement of frontal calvarium crossing the midline, the second along the fronto-parietal falx cerebri with dural tail.

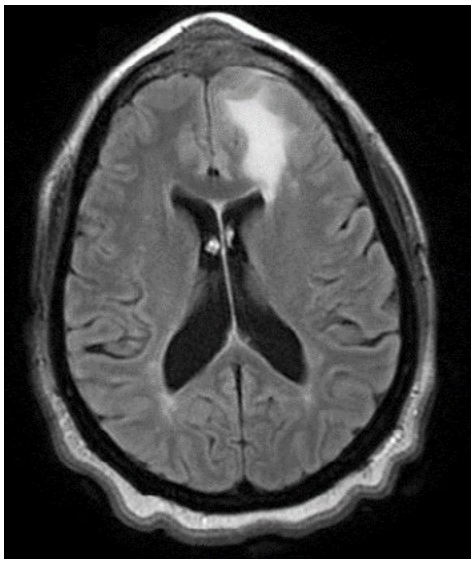


Figure 2: T2-weighted FLAIR image shows mass effect on the left frontal lobe.

REFERENCES

1. Kumar, S., et al., Primary low-grade B-cell lymphoma of the dura: a mucosa associated lymphoid tissue-type lymphoma. Am J Surg Pathol, 1997. 21(1): p. 81-7.

2. Kambham, N., Y. Chang, and A.Y. Matsushima, Primary low-grade B-cell lymphoma of mucosa-associated lymphoid tissue (MALT) arising in dura. Clin Neuropathol, 1998. 17(6): p. 311-7.

← BACK TO PAGE 1
NEXT PAGE →

Medical Management of Multiple Cavernoma Syndrome

Rance A. Boren[1], MD, Hannah C. Justice[2], Christopher S. Eddleman, MD[3]

[1]Brownwood Regional Medical Center, Brownwood, TX
[2]Howard Payne University, Brownwood, TX
[3]Hendrick Health System, Abilene, TX

The authors declare no conflicts of interest and received no funding for this project. The patient gave informed consent for publication of the case report, which was drafted according to the CARE guidelines and contains no identifying information.

INTRODUCTION:

Familial cerebral cavernous malformation (CCM) syndrome is an autosomal dominant disorder with variable penetrance that accounts for approximately 20% of all cavernoma cases. Although these lesions were historically considered hamartomas that enlarged through sequential hemorrhage, the formation of cavernomas is now understood as a dynamic process of disordered angiogenesis which in some patients manifests as a lifelong tendency to develop new lesions.1 It is felt that a two-hit genetic mechanism, involving both the inherited germ-line mutation and a second somatic mutation or inactivation, accounts for much of the variability in penetrance of this disorder, but it also appears that various biochemical triggers may lead to cavernoma growth and rupture in genetically susceptible individuals. 2

When necessary, the mainstay of treatment is surgical resection; stereotactic radiotherapy is not recommended in the familial syndrome due to the risk of triggering growth of additional lesions.3,4 However, a number of promising avenues for medical therapy are being pursued...therapeutic targets under investigation include angiogenic growth factors, cell adhesion molecules, second-messenger systems, and mediators of autophagy, inflammation, and oxidative stress.2,5 The most-studied pathway involves inhibition of Rho-Kinase A, but its most potent inhibitor, Fasudil, is not available in the United States; data regarding the benefit of statins in this pathway has been contradictory.5,6,7 Other potential medications showing promise include propranolol, Vitamin D, sulindac, rapalogs, and VEGF inhibitors, among others.2,8 Unfortunately, there are no controlled human studies of these medications.3

Numerous loss-of-function mutations in three cerebral cavernous malformation (CCM) genes (CCM1/KRIT1, CCM2/Malcaverin, and CCM3/PDCD10) have been identified. These genes encode products that are intimately involved in maintenance of normal vascular endothelium, each probably acting through multiple pathways and in association with the others.1,9 The substantial overlap of these CCM pathways suggests that medicines with downstream activity may be effective regardless of genotype, but it is possible that specific agents may be more effective in one mutation than in others. Gene editing may hold promise as well.

The high degree of variability in clinical presentation makes prediction of long term outcomes difficult, but attempts have been made to characterize both clinical and laboratory markers of disease aggressiveness.10 Early age of onset, two or more symptomatic hemorrhages, and high lesion burden have been identified as indicators of chronic disease severity. Acute disease aggressiveness is characterized by symptomatic hemorrhage, documented lesion growth, and de- novo lesion formation. A combination of low vitamin D and non-HDL cholesterol levels (less than 118.8) seems to have predictive value for chronic disease activity according to the equations 0.74 x VitD + 0.75 x Non-HDL-C. Below, we report a patient demonstrating both acute and chronic disease aggressiveness and an unfavorable serum biomarker profile and discuss management options.

CASE REPORT:

A 19 year-old male presented to the emergency department with a three-week history of right frontal headache. His computerized tomography (CT) scan showed a 2 x 0.9 x 2.4 cm expansile, osteolytic lesion eroding through the inner table of the frontal bone. Postsurgical changes were noted in the right temporal lobe and cerebellum. A small air-fluid level was seen in the right maxillary sinus. Eight months previously he had vomiting and headache associated with a 3 x 4 x 3 cm non-hemorrhagic cerebellar cavernoma; in retrospect his scan then showed a smaller (1.7 x 1.8 x 0.5 cm) lytic lesion with thinning of the inner table. A scan four years earlier was normal in both areas. (Figure1).

His history was remarkable for four prior surgeries for symptomatic cerebral cavernous malformations, beginning at age nine. Family members in four generations were affected. He was otherwise healthy, though he had been smoking since age 12 at an average of two packs per day (four per day on presentation). His skull was tender to palpation over the lesion. He also had a painful 2 x 3 cm soft tissue mass lateral to the right knee that he said had been present for a decade. He reported that it had enlarged intermittently with apparently spontaneous bruising until about age 15, with persistent pain thereafter. His vitamin D level was 34.1 ng/ml, and his fasting non-HDL cholesterol was 89 mg/dl, for a combined value of 114.9. Genetic testing revealed a deletion in the CCM2/Malcaverin gene.

Magnetic resonance imaging (MRI) of the head confirmed an enhancing mass consistent with an intraosseous cavernous hemangioma, with slight local mass effect on the underlying brain (Figure 2). There were three additional abnormalities on gradient imaging consistent with cerebral cavernomas and a small enhancing area in the cerebellum (susceptibility-weighted imaging, which is more sensitive for cavernoma detection11, was not available at our institution.)

← BACK TO PAGE 1
NEXT PAGE →

Technecium-99 scintigraphy revealed another lesion in the right orbital roof and one in the distal left tibia (Figure 3). When questioned about the left ankle he reported that it had been intermittently painful since childhood, but he was told it was "growing pains." An x-ray of the ankle at age thirteen was normal, but one at age sixteen showed a 2 x 3 cm sclerotic "bone cyst" in the distal tibia.

His tumor was removed via a right frontal craniectomy. Gross examination showed punctuate erosion of the outer table by vascular channels and complete destruction of the inner table. (Fig 4) Pathology revealed dilated venous sinusoids consistent with a cavernous malformation. Subsequently an MRI of the knee confirmed a 3 x 1 x 4 cm superficial vascular lesion in the region of the iliotibial band and a deeper lesion along the popliteus tendon (Figure 5). The presumptive diagnosis of cavernoma was confirmed at surgery. EKG abnormalities during his preoperative evaluation led to a chest CT…incidental note was made of a 1 cm vascular lesion in the right lower lung which was not present on a CT of the abdomen a year earlier.

DISCUSSION:

In the familial cavernoma syndrome extra-neural cavernomas are well-known to occur in the vertebral bodies, retina, liver, and skin.12,13,14 Skull, long bone, pulmonary, and joint involvement, as seen in our patient, are apparently novel manifestations of the familial syndrome though sporadic cavernomas have been reported in these locations, alone or in combination with other lesions, and one patient without genetic or pathologic confirmation has been reported with multiple cerebral and pulmonary hemangiomas.15,16

The differential diagnosis of cavernous malformations is broad and varies with the site of involvement and age of the patient. Our patient's youth and known predisposition to cavernoma formation simplified the process. The bone scan was performed on the basis of a case report suggesting that this was more sensitive than MRI for detection of additional intraosseous lesions.15 In this instance two unsuspected lesions were discovered.

The reported cases of sporadic cavernomas of the skull suggest a slight female predominance, with lesions typically presenting in middle age as a slowly growing bony mass. In classic cases expansion is outward, with sparing of the inner table; the frontal bone is most commonly affected.17 Lesions are typically solitary and often asymptomatic or merely painful, but multiple lesions and inward growth with hematomas and hemiparesis have been reported.18,19 Metastasis, osteosarcoma, myeloma, and intraosseous meningioma are neoplastic possibilities, while Langerhans histiocytosis, Paget's disease, dermoid/epidermoid cysts, and fibrous dysplasia are other contenders.

In the appendicular skeleton the long bones are most commonly affected and in approximately 75% of those cases the diaphysis is involved.20 The differential diagnosis is similar to that of skull lesions. Definitive treatment consists of en bloc resection with grafting and instrumentation as needed; the location of our patient's lesion in the ankle makes surgical treatment problematic.

Synovial hemangiomas are rare and typically occur in childhood.21 These lesions may affect various joints but appear more common in the knee, where they can be intra-articular or arise from adjacent tendons. Other peri-articular connective tissues may be similarly involved. As in our patient, presentation often occurs due to atraumatic bloody effusions; diagnosis is frequently delayed. In addition to other vascular malformations the differential includes pigmented villonodular synovitis, various inflammatory arthropathies, trauma, and bleeding diatheses.

Solitary pulmonary cavernous hemangiomas are rarer still, with presenting symptoms ranging from incidental detection to pulmonary hypertension, polycythemia, or fatal hemoptysis. When they present in adulthood they are likely to be mistaken for lung cancer.22 Treatment options include watchful waiting, surgical resection, and endovascular treatment.

Postoperatively, our patient was placed on propranolol, a lipophilic non-specific beta-adrenergic antagonist that exerts several known effects on angiogensis. This inexpensive, well-tolerated agent has been found to be an effective treatment for infantile hemangioma at a dose of 3mg/kg divided BID.23 There are also encouraging preliminary results in angiosarcoma, hereditary telangectasia, and retinopathy of prematurity.24,25,26 The anti-angiogenic effects of propranolol presumably underlie the benefits seen in studies involving solid tumors.27

Several anecdotal reports support the use of propranol in cavernoma patients. Muschovi first reported the use of propranolol in a pediatric patient with a giant cavernoma in 2010. A ten day course was associated with "significant tumor shrinkage."28 Another other pediatric patient with multiple lesions has been reported to be treated with good results.28 Two adult patients (one of them a familial case) with an apparent response to this medication have also been reported.29 Both patients were hesitant to undergo surgery and had regression of their lesions while on propranolol. Another possible cavernoma patient (CCM mutation negative) with multiple hemorrhages associated with von-Willibrands's disease had abrupt cessation of hemorrhages with propranolol therapy after failure of hemostatic treatment, thalidomide, and simvistatin.30

Propranolol down-regulates the production of vascular endothelial growth factor (VEGF), which has been shown to correlate with cavernoma growth patterns.31,32,33 Propranolol has also been shown to induce apoptosis, inhibit DNA replication through the cyclin pathway, and inhibit tubule formation by endothelial precursor cells due to effects on chemotaxis and cellular migration via reduced expression of matrix metaloproteinases.34,35 In addition,

← BACK TO PAGE 1
NEXT PAGE →

Medical Management of Multiple Cavernoma Syndrome (*cont.*)

propranolol mitigates oxidative stress through metabolites. This is presumed to account for some of its cardioprotective activity.36 Lastly, since hemodynamic factors are felt to influence the growth of cavernomas, there is potential for more mundane effects through peripheral vasoconstriction or other anti-hypertensive properties.37

A high-throughput screening study of 2,100 existing drugs and other molecules identified cholecalciferol (vitamin D3) as having activity on the CCM pathways and, in a mouse model, demonstrated a reduction in cavernoma burden by MRI.5 Vitamin D has been shown to act in the oxidative stress pathways, regulate gene expression, and inhibit epithelial-to-mesenchymal transition.38,39 Furthermore vitamin D levels have been found to correlate inversely with disease severity in cavernoma patients.10 Since our patient's vitamin D level was in the "insufficient" range, supplementation was recommended following the recommendations for multiple sclerosis patients.

Recently, an association between infection and cavernoma formation has been suggested.41 Lipopolysaccharide was implicated as a stimulant of the toll-like receptor 4 (TLR4) system and the authors demonstrated that blockade of this pathway could prevent cavernoma formation in a mouse model. This supports the growing impression that local biochemical "third-hits" may play a role in cavernoma formation. Our patient had radiographic evidence of chronic rhinosinusitis with an active infection around the time of his presentation. Review of his earlier scans showed substantial sinus disease fluctuating in location and extent as far back as 2009. The significance of this is unclear given the frequent appearance of sinusitis as an incidental finding, but the idea that activation of inflammatory cytokines and other signaling molecules could cross over from one disease to the other must be considered.

Lastly, our patient was advised to eliminate nicotine. This compound has well-documented pro-angiogenic properties, including stimulation of endothelial cell proliferation, migration, and tubule formation through numerous pathways.42 A study of CCM1 patients with the Common Hispanic Mutation failed to show an effect of smoking on disease severity as measured by lesion count, though there was a slight association between pack-years and the chance of hemorrhage.43 However, review of the data indicates that smokers were over-represented, suggesting some effect of tobacco on study entry.44

Our patient demonstrated an aggressive clinical course, with seven symptomatic lesions before age of 20, and developed extracerebral cavernomas in unusual locations for a patient with the familial syndrome. He was well-informed about his disease and its natural history…in addition to his own problems he had witnessed symptoms in numerous relatives. He understood that there was only indirect or anecdotal evidence for an effect of propranolol and vitamin D on cavernomas. When given the choice between a low-risk therapy with uncertain benefit and an apparently high chance of future problems, he chose the former. This approach, while speculative, appears justified given the safety profile of propranolol, the growing body of clinical evidence to support its off-label use in disorders of angiogenesis, and the morbidity associated with hemorrhage. Lastly, this case highlights the potential for cavernomas to form at unusual sites, and patients with this syndrome should be made aware of this possibility.

REFERENCES

Haasdijk R, Cheng C, Maat-Kievit A, Duckers H: Cerebral cavernous malformations: From Molecular pathogenesis to genetic counseling and clinical management. Eur J Hum Genet 2012;20:134-140.

Retta S, Glading A: Oxidative stress and inflammation in cerebral cavernous malformation disease pathogenesis: Two sides of the same coin. Int J Biochem Cell Biol 2016;81:254-270.

Akers A, Al-Shahi Salman R, Issam A, et al: Guideline for the clinical management of cerebral cavernous malformations: Consensus recommendations based on systematic literature review by the Angioma Alliance scientific advisory board clinical experts panel. Neurosurgery 2017;80:665-680.

Golden M, Saeidi S, Liem B, Marchand E, Morrison L, Hart B: Sensitivity of patients with familial cerebral cavernous malformations to therapeutic radiation . J Med Imaging Radiat Oncol 2015;59:134-136.\

Gibson C, Zhu W, Davis C, et al: Strategy for identifying repurposed drugs for the treatment of cerebral cavernous malformation. Circulation 2015;131:289-299.

Yin L, Morishige K, Takahashi T, et al: Fasudil inhibits vascular endothelial growth factor-induced angiogenesis in vitro and in vivo. Mol Cancer Ther 2007;6:1517-1525.

Shenkar R, Shi C, Austin C, et al: RhoA kinase inhibition with fasudil versus simvastatin in murine models of cerebral cavernous malformations. Stroke 2017;48:187-194.

Bravi L, Rudini N, Cuttano R, et al: Sulindac metabolites decrease cerebrovascular malformations in CCM3 knockout mice. PNAS 2015;112: 8421-8426.

Gore A, Lampugnana M, Dye L, Dejana E, Weinstein B: Combinatorial interaction between CCM pathway genes precipitates hemorrhagic stroke. Dis Model Mech 2008;1:275-281.

Girard R, Khanna O, Shenkar R, et al: Peripheral plasma vitamin D and non-HDL cholesterol reflect the severity of cerebral cavernous malformation disease. Biomark Med 2017;10:255-264.

de Sauza, J, Dominguez R, Cruz L, Domingues F, Iasbecj T, Gasparetto E: Susceptibility-weighted imaging for the evaluation of patients with familial cerebral cavernous malformations: A comparison with T-2 weighted fast spin-echo and gradient-echo sequences. AJNR 2008; 3: 154-158.

Toldo I, Drigo P, Mammi I, Marini V, Carollo C: Vertebral and spinal cavernous angiomas associated with familial cerebral cavernous malformation. Surg Neurol 2009;71:167-171.

Drigo P, Mammi I, Battistella P, Riochieri G, Carollo C: Familial cerebral hepatic, and retinal cavernous angiomas: A new syndrome. Child's Nerv Syst 1994;10:209-209.

Sirvante J, Enojolas O, Wassef M, Tournier-Lasserve E, Labauge P: Frequency and phenotypes of cutaneous malformation in a consecutive series of 417 patients with familial cerebral cavernous malformations. JEADV 2009;23:1066-1072.

Sasagawa Y, Akai T, Yamamoto K, et al: Multiple cavernous hemangiomas of the skull associated with hepatic lesions. Neurol Med Chir (Tokyo) 2009;49:162-166.

Giannopoulos S, Pelidou S, Kryitis A. Multiple organ cavernous hemangiomas involving brain, skin, and lung. Cornea 2008;1:128.

Heckkl, S, Aschoff A, Kunze S: Cavernomas of the skull: review of the literature 1975-2000. Neurosurg Rev 2002;25:56-62.

Gottfreid O, Gluf W, Schmidt M: Cavernous hemangioma of the skull presenting with subdural hematoma. Neursurg Focus 2004;17:Clinical Pearl 1, 2004.

Nasi D, Somma L, Iacoangeli M, et al: Calvarial bone cavernous hemangioma with intradural invasion: An unusual aggressive course-case report and literature review. Int J Surg Case Rep 2016;22:79-82.

Kaleem Z, Kyriakos M, Totty W: Solitary skeletal hemangioma of the extremities. Skeletal Radiol 2000;29:502-513.

Wang M, Chen C, Yeh L, et al: Synovial hemangioma of the knee joint: A case report. Chin J Radiol 2008;34:239-243.

Chen Q, Li H, Hu B, Jin M, Guo J. Multiple Cavernous Hemangiomas of the lung. Ann Thorac Surg 2014;98:1835-1837.

Leaute-Labreze C, Hoeger P, Mazereeuw-Hatier J, et al: A randomized, controlled trial of oral propranolol in infantile hemangioma. N Engl J Med 2015;372:735-746.

Pasquier E, Andre N, Street J, et al: Effective management of advanced angiosarcoma by the synergistic combination of propranolol and vinblastine-based metronomic chemotherapy: A bench to bedside study. EBioMed 2016;6:87-95.

Mei-Zahev M, Blau H, Bruckheimer E, Zur E, Goldschmidt N: Topical propranolol improves epitasaxis in patients with hereditary hemorrhagic telangectasia-a preliminary report. J Otolaryngol Head Neck Surg 2017;46:58.

Buhrer C, Bassler D: Oral Propranolol: A new treatment for retinopathy of prematurity? Neonatology 2015;108:49-52.

← BACK TO PAGE 1

Pasquier E, Ciccoline J, Carre M, Giacometti S, Fanciullino R, Pouchy C: Propranolol potentiates the anti-angiogenic effects and anti-tumor efficacy of chemotherapy agents: Implications in breast cancer treatment. Oncotarget 2011;2:797-809.

Muschovi M, Alexiou G, Stefanik K, Tourkantoni T, Prodromou N: Propranolol treatment for a giant infantile brain cavernoma. J Child Neurol 2010;25:653-655.

Berti I, Marchetti F, Skabar A, Zennaro, Zanon D, Ventura A: Propranolol for cerebral cavernous angiomatosis: A magic bullet. Clin Pediatr 2014;53:189-190.

Zabramski J, Kalani M, Filippidis A, Spetzler R: Propranolol treatment of cavernous malformations with symptomatic hemorrhage. World Neurosurg 2016;88:631-639.

Reinhard M, Schuchardt F, Meckel S, et al: Propranolol stops progressive multiple cavernoma in an adult patient. J Neurol Sci 2016;367:15-17.

Jung K, Chu K, Jeong S, Park H, Bae H, Yoon B: Cerebral cavernous malformations with dynamic and progressive course-correlation study with vascular endothelial growth factor. Arch Neurol 2003;60:1613-1618.

Maiuri F, Cappbianca P, Gangemi M, et al: Clincal progression and familial occurrence of cerebral cavernous angiomas: the role of angiogenic and growth factors. Neurosurg Focus 2006;21(1):E3.

Wu S, Wang B, Chen L, et al: Clinical efficacy of propranolol in the treatment of hemangioma and changes in serum VEGF, bFGF and MMP-9. Exp Ther Med 2015;10:1079-1083.

Kum, J, Khan Z: Mechanisms of propranolol action in infantile hemangioma. Dermatoendocrinol 2014;6:e979699. Available at: https://www.tandfonline.com/doi/full/10.4161/19381980.2014.979699 Accessed Dec 18, 2017.

Lamy S, Lachambre M, Lord-Dufour S, Beliveau R: Propranolol suppression angiogenesis in vitro: Inhibition of proliferation, migration, and differentiation of endothelial cells. Vascul Pharmacol 2010;53:200-208.

Mak I, Weglicki W: Potent antioxidant properties of 4 hydroxyl-propranolol. JPET 2004;308:85-90.

Little J, Issam A, Jones S, Ebrahim Z: Vascular pressures and cortical blood flow in cavernous angioma of the brain. J Neurosurg 1990;73:553-559.

Urberti F, Lattuada D, Morsanuto V, Nava U, Bolis G, Vacca G: Vitamin D protects human endothelial cells from oxidative stress through the autophagic and survival pathways. J Clin Endocrinol Metab 2013;99: 1367-1374.

Larriba M, Garcia de Herreros A, Munoz A: Vitamin D and the epithelial to mesenchymal transition. Stem Cell Int 2016: DOI: 10.1155/2016/6213872 Available at: https://www.hindawi.com/journals/sci/2016/6213872/ [Accessed Dec 4, 2017.]

Tang A, Choi J, Kotzin J, et al: Endothelial TLR4 and the microbiome drive cerebral cavernous malformations. Nature 2017;545:305-310.

Lee J, Cooke J: Nicotine and pathological angiogenesis. Life Sci 2012;91:1058-1064.

Choquet H, Nelson J, Pawlikowska L, et al: Association of cardiovascular risk factors with disease severity in cerebral cavernous malformation type 1 subjects with the common hispanic mutation. Cerebrovasc Dis 2014;37:57-63.

Centers for Disease Control and Prevention: State tobacco activities tracking and evaluation. https://nccd.cdc.gov/STATESystem/rdPage.aspx?rdReport=OSH_STATE.Highlights&rdRequestForwarding=Form [Accessed May 2018.]

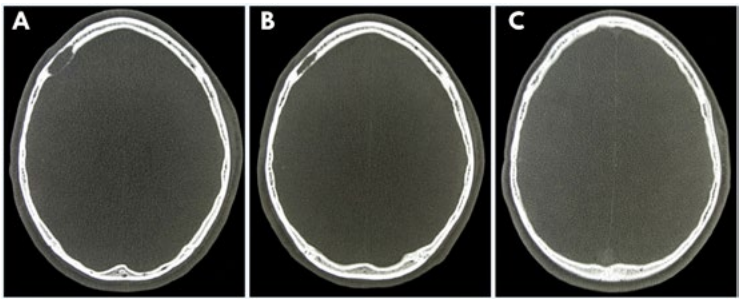


Figure 1: Unenhanced axial CT head bone windows at presentation, (B) eight months prior, (C) four years prior

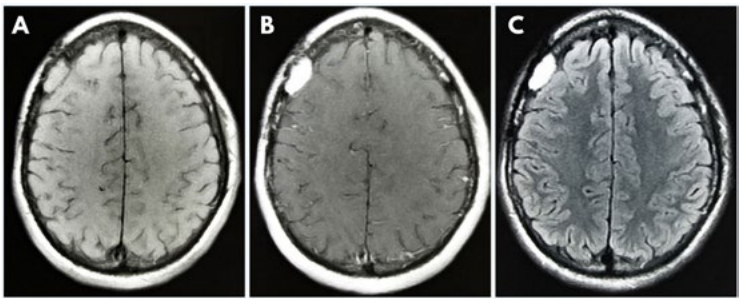


Figure 2: Axial MRI of the head T1 without contrast, (B) T1 with contrast, (C) T2 FLAIR

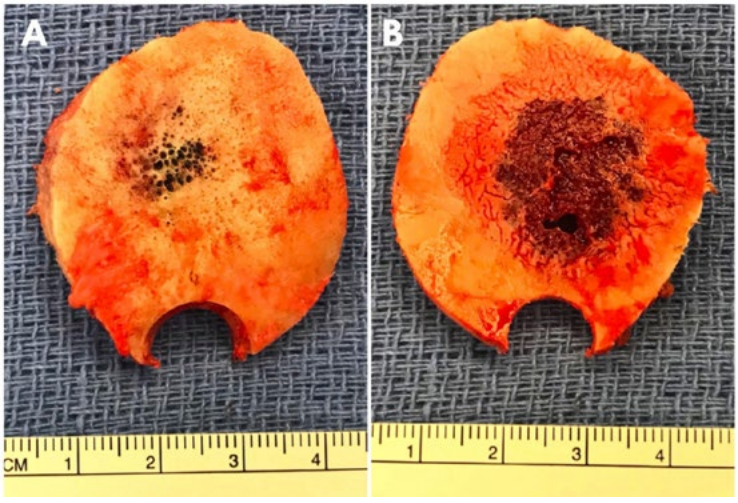


Figure 3: Craniectomy Specimen outer table, (B) inner table

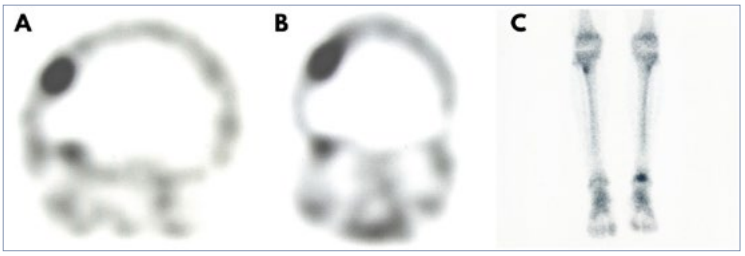


Figure 4: Technicium-99 Bone Scan (A,C) Anterior-Posterior, (B) Lateral

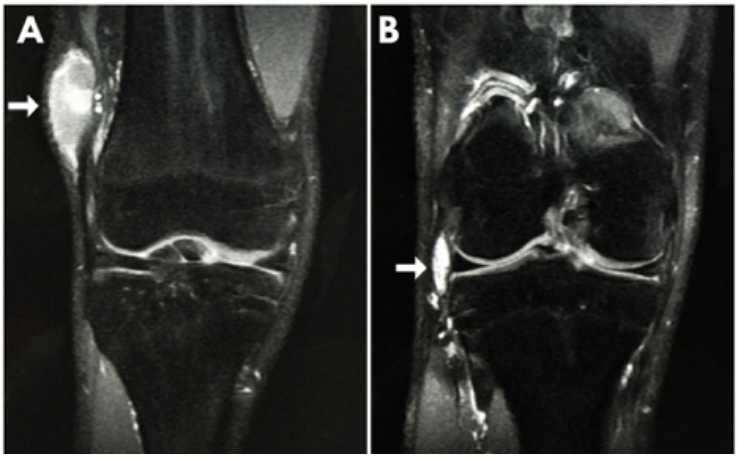
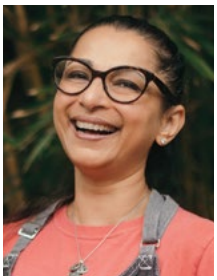


Figure 5: Coronal MRI of the right knee (A,B) T2 fat-saturated post-contrast; arrows denote cavernomas

BACK TO PAGE 1



Nutritional Neurology—What's Old is New Again

(4 Clinical Cases and a Brief Discussion)

By Madhureeta Achari, MD
Tasting-Health.com

INTRODUCTION

Nutritional illnesses are the oldest treatable medical maladies known. The phrase 'let food be thy medicine', attributed to Hippocrates, reminds us that good nutrition is essential for good health. However, food is not medicine. Food is fuel to keep the body running optimally. Food provides the essential micronutrients, vitamins and minerals, required for every metabolic process in the body. Organic compounds from foods, vitamins are coenzymes in cellular processes. Their deficiencies or insufficiencies have caused significant illnesses throughout medical history.

The role of nutrition in neurological disorders is well known. Vitamin B12 deficiency is a treatable cause of memory disorders. Vitamin A deficiency remains the main cause of night blindness worldwide and hypervitaminosis A causes pseudotumor cerebri through its influence of CSF metabolism. Vitamin A has a role in memory function. Wernicke-Kirsakoff syndrome from Vitamin B1 deficiency is well known, as is Beri-Beri with both cognitive effects and peripheral neuropathy. Vitamin B1 deficiency is making a resurgence in the era of bariatric surgery, gluten-free and ketogenic diets. Vitamin C deficiency not only causes scurvy, but has been implicated in spinal and other pain syndromes. Vitamin D deficiency is associated with white matter diseases and is a cause of positional vertigo. Multiple other disorders related to the other vitamins are well known.

Nutritional deficiencies and their sequelae are typically thought of in the setting of alcoholism, substance abuse, food deprivation, poor socioeconomic status, poor access to fresh food, critical and serious illness, chemotherapy and anorexia. However, in the current era of ever-expanding restrictive diets (gluten-free, paleo, ketogenic, vegan, vegetarian, blood type, etc), meal replacements (bars, shakes, powders, etc), and the demise of the old fashioned well-balanced meal, our patients are developing multiple nutritional deficiencies without even knowing they are at risk for serious illness. Our current nutritional illnesses are not related to the special circumstances described above. Rather, they are occurring across the board touching all facets of our population. In my experience, it is now also higher socioeconomic segments of the population, along with those with higher levels of education who fall prey to fad diets and supplement hawkers with negative consequences to their health. The sad irony is that they are getting sick in an effort to be healthier.

Another, more complex issue leading to nutritional illness is the poor quality of the food available in most grocery stores. Whether conventional or organic, most produce is cold stored for significant lengths of time rendering them nutritionally bankrupt. The apples in the grocery store are 6 to 12 months old, dipped in the right chemicals and held at the perfect temperatures to ensure sweetness and crunch, but many of the nutrients, especially vitamin C are lost. Processed foods and food replacements in the form of shakes, bars and powders offer very little real nutrition and are usually full of thickeners and sugars. Furthermore, commercial farming methods are also causes of nutritional depletion, adding to the complexity of the problem.

To combat the multitude of health/nutrition claims made by snake oil salespeople with get-rich-quick schemes, physicians need to approach nutritional illness with scientific tools. Our first task is to think about nutrition in the clinical setting and take a dietary history focused on restrictive diets and processed foods. One of the easiest methods to check for nutritional deficiencies is to obtain blood levels for vitamins and minerals. These are readily available at commercial laboratories and mostly covered by insurance plans. It has been reported that multiple vitamin deficiencies occur together.

In my practice, as micronutrients play a key role in every aspect of metabolism, it makes sense to keep levels in the high normal ranges. Using prescription supplements whenever possible helps with the precision and accuracy of treatment. When using over the counter supplements, sticking to national brands that undergo testing and national certification is preferred. As over the counter products are not regulated and few prescription forms exist anymore, keeping an eye on the levels is critical. If a level drops and the patient is taking the supplements regularly, it may be that the concentration in the supplement has decreased. I also take time to teach patients how to prepare simple meals using seasonal produce and locally farmed meats. If food is the basis of good health, then we all need to know how to shop for it, prepare it and feed ourselves.

THE HISTORY OF RECOMMENDED DAILY ALLOWANCES (RDA)/DIETARY REFERENCE INTAKES (DRI)

In 1940, Franklin D. Roosevelt, established the National Defense Research Committee "to coordinate, supervise, and conduct scientific research on the problems underlying the development, production, and use of mechanisms and devices of warfare." As a result of the war, there was a growing urgency for the government to care for populations left malnourished or starving. In 1941, the NDRC asked the National Research Council and the Institute of Medicine's Food and Nutrition Board to come up with Recommended Dietary Allowances for protein, energy, and eight vitamins and minerals they determined to be most important. This first attempt to determine human requirements for the various dietary nutrients would be the foundation for food relief efforts and would provide the government with guidelines on

← BACK TO PAGE 1
NEXT PAGE →

how to feed American troops in the field. They employed 50 of America's leading nutrition experts and reviewed all the available published evidence. The first report of Recommended Dietary Allowances was published in 1941, and after the war many countries used the United States RDAs to set their own dietary standards. These guidelines were reviewed in 2017 and revised criteria for RDA/DRI will soon be available. However, the most recent data found in NHANES (National Health and Nutrition Examination Survey) suggests that 31% of the US population remains nutritionally deficient despite supplementation of foods based on RDA criteria.

CLINICAL CASES

The following are a few of the nutritional cases that I have treated over the past 10 years. The rate of new cases is astounding with over 800 since 2016. The average financial cost before nutritional labs are performed is approximately $50,000, including multiple specialist visits, labs, scans, procedures including lumbar punctures, chiropractic and nutritionist visits, multiple prescription medications, supplements and unnecessary 'cures'.

CASE # 1: PARESTHSIA/RLS

29yo previously healthy female law student presents with a 6 month history of increasing pain and sensitivity in the feet. She also notes soreness in the calves at times. She has developed some tingling in the feet and her feet feel 'tight' at times. The pain and discomfort are present throughout the day, but become unbearable at night. She gets better with putting feet on the ground and walking around. Her MRIs of the brain, cervical and lumbar spines were normal. EMG/NCV of the lower extremities were normal. Nutritional labs showed Vitamin C deficient at 0.2 ng/dL , vitamin D deficient at 19 ng/mL, vitamin B12 deficient at 220 pg/mL, and vitamin B2 deficient at 5.2 nmol/L. The patient was given over the counter vitamin C 1000mg, vitamin B12 1000mcg, vitamin D3 5000Iu daily and prescribed ergocalciferol 50,000IU weekly. Her symptoms improved rapidly over 3 weeks, and resolved in 4 months. Repeat nutritional levels were all in high normal ranges with vitamin C 1.5, vitamin D 57, vitamin B12 817 and vitamin B2 32. The diagnoses were restless leg syndrome and paresthesias of the lower extremities due to nutritional deficiencies. As a law student and also working full time, the patient was eating mostly processed snacks and fast foods. With the addition of supplements and changing her diet over the next 6 months, she recovered completely. Over the past 2 years, she has continued to do well with improved diet alone and is no longer taking supplements.

CASE # 2: KETOGENIC DIET AND ENCEPHALOPATHY

26yo male notes trouble with speech and memory for 3 months. He has trouble with word finding and short-term memory. For the last 2 months his thinking seems 'cloudy' and he cannot understand instructions easily. He has difficulty sustaining attention and cognitive focus. He does accounting for his mother's company and has been making errors. His grades in college have also declined; he was previously an A student. He got confused with directions while driving to college. He started a ketogenic diet 4 months ago to lose weight, which he has. He continued on the diet as he was told by a nutritionist it was good for him. Prior to this he ate a wide variety of healthy foods. For his ketogenic diet, he eats bacon and eggs, beef and broccoli, chicken and shrimp and spinach salad with nuts. He eats no grains. His past history is significant for asthma and ALL, which was treated, and he is followed for this. His MRI of the brain is normal. EEG is normal. Nutritional labs revealed vitamin B1 deficient at 71 nmol/L, vitamin B2 deficient at 5.2 nmol/L, vitamin C deficient at 0.3 mg/dL, and vitamin D deficient at 30 ng/mL. He was supplemented with OTC vitamins and advised to eat a healthy variety of foods. He improved within 2 weeks and has been well since. His nutritional levels were all normal in 12 weeks and he continues with a well-balanced, varied healthy diet.

CASE # 3: NUTRITIONAL NEUROPATHY

42yo male with idiopathic painful sensory-motor demyelinating peripheral neuropathy diagnosed 8 years ago. He was placed on gabapentin and then on pregabalin with some improvement. His outside labs from 8 years ago revealed a normal HgbA1C at 5.4 and low normal B12 of 340, which was not supplemented. His examination was significant for absent DTRs in the lower extremities and stocking distribution numbness. A repeat EMG/NCV confirmed the findings. Nutritional labs showed vitamin B1 deficient at 71 nmol/L, vitamin D deficient 19 ng/mL, vitamin B12 deficient at 152 pg/mL. He was supplemented with OTC B1, D3 and B12 and given B12 injections and ergocalciferol weekly. Over the next 4 years, his neurological exam reverted to normal and his EMG/NCV was normal in 6 years. He has been doing well for 10 years and continues to take supplements with levels maintained in high normal ranges. He is no longer taking pregabalin.

CASE # 4: SCURVY

19yo female with JME diagnosed at age 17. She has stopped taking her medications due to side effects and has recurrent seizures. She had been on levetiracetam and lamotrigine which were ineffective. She has been on zonisamide and lacosimide for the past 8 months with excellent results. However, in the last 2 months, since she has been living on her own, she has developed periodic abdominal pain and cramping, nausea, joint pain and severe fatigue. She also has neck pain, headache and dizziness. She has diffuse body aches, which is worse in her hands and fingers. No changes were made to her AEDs and she has been receiving the same formulations from same manufacturers. She was evaluated by a gastroenterologist without any etiology of her symptoms identified. Examination revealed a red rash on her cheeks and tenderness at tendinous insertions in arm and legs. She had mild diffuse abdominal tenderness. Nutritional labs revealed vitamin C deficient at 0.1 mg/dL, vitamin B2 at deficient 5.2 nmol/L, vitamin B12

← BACK TO PAGE 1
NEXT PAGE →

Nutritional Neurology—What's Old is New Again (*cont.*)

deficient at 200 pg/mL, and vitamin D deficient at 28 ng/mL. Although the majority of her symptoms were due to scurvy, the other nutritional deficiencies were clearly contributing factors. Her diet consisted solely of canned chicken soups for the 2 months prior to symptom onset. With replacing all deficiencies, the entirety of her symptoms resolved over 8 weeks. She remains well and seizure free on medications at 4 years. Due to her inability to sustain a proper diet, she continues on OTC supplements with yearly levels.

SUMMARY

Although the scope of this article doesn't allow a detailed discussion of the pathophysiology of nutritional illness, the information exists in the scientific literature and continues to expand. Each of the micronutrients have specific neurological roles and thus manifestations of neurological illness. In future articles, each vitamin and its neurological pathophysiology can be discussed.

Nutritional illnesses are far more common than we expect and caused by a multitude of factors, old and new. They are easy to diagnose and treat. But first, we must consider them in the differential diagnosis. Our patients are looking for ways to improve their health and we have the knowledge, expertise and ability to help them. If we, as physicians, don't embrace the science of nutritional neurology, our patients will continue to turn to the gallery of non-medical "practitioners" who may cause more harm than good. In an ever more complicated and advancing world, it's time to get back to basics.

SELECTED REFERENCES

Bird, JK, et al Nutrients 2017, (9) 7, 655. The Risk of Deficiencies in Multiple Concurrent Micronutrients in Children and Adults in the United States.

Bruins, MD, et al Nutrients 2018, (10) 1, 47. Consideration for Secondary Prevention in High Risk Groups in High Income Countries.

Hamel, J, et al Muscle Nerve 2018 Jan: 57 (1):33-39. Acute Nutritional Axonal Neuropathy

Grim, J, et al Nutrients 2017 May 25; 9(6), E535 Selected Risk Nutritional Factors for Chemotherapy-Induced Polyneuropathy

Miesel, K, et al Neurology In Clinical Practice July 23, 2015. Scurvy in the 21st Century

Publications of the National Academies Of Science. How the RDA was Established.

2019 TNS Annual Winter Conference

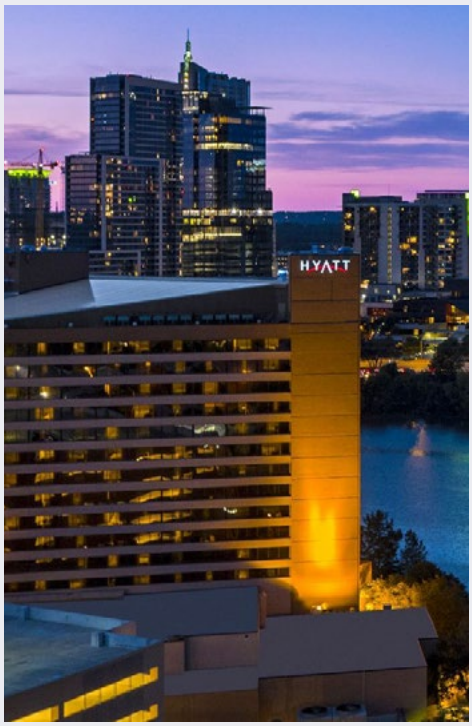
It's that time of year again when Texan neurologists flee to the capital to further our knowledge, catch up with old friends, get to know new TNS members, and share our experience with both the clinical and business aspects of our practices.

This has been an exceptional year for the Texas Neurological Society. Our membership is at a record high, and practices from both private and academic institutions have joined us, making our society stronger than ever. We've also seen some changes and advances in neurology in 2018, starting with the FDA's approval of new treatments for genetic disorders, with a few others coming down the pipeline. For the first time, the FDA has also recommended the approval of a marijuana-based drug for medical use. CMS spared us no excitement, either, having suggested changes that will impact neurology practices nationwide.

This year, we've invited well-known speakers from such universities as Baylor, Washington University, UCLA, Columbia, UTSW, and UT Austin. We'll hear from them on a broad range of topics: neuromuscular disorders, multiple sclerosis, movement disorder, epilepsy, and stroke, to name just a few. We made sure to add new topics of interest, covering issues that neurologists currently face, such as the narcotics epidemic, an update on CMS regulation, nutritional neurology, genetic ethics, and research in the medical use of marijuana.

Your attendance will strengthen TNS and make our voice more powerful. See y'all here in beautiful Austin this coming February.

Yessar Hussain, MD
TNS Winter Program Director



Feb. 1-3
Hyatt Regency
Austin

← BACK TO PAGE 1

NEXT PAGE →

Glossopharyngeal Neuralgia Associated with Cardiac Syncope

Shamin Masrour, DO
Assistant Professor, UT Southwestern Medical Center

INTRODUCTION

Glossopharyngeal neuralgia (GN) is characterized by paroxysmal pain in the throat, pharynx, tongue, and ears. Rarely, the condition can cause loss of consciousness due to bradycardia or asystole. This condition should be recognized and diagnosed appropriately as the potential cardiac consequences may be life-threatening. Below is a case presentation of a man with multiple episodes of syncope that was ultimately diagnosed with glossopharyngeal neuralgia.

CASE PRESENTATION

A 72-year-old man with no significant past medical history was initially taken to the hospital for syncope. He was awakened from sleep by episodes of severe pain in his left ear and lost consciousness after getting out of bed. EMS was called and a heart rate of 35 bpm and hypotension were noted. While hospitalized, he experienced multiple additional episodes of severe pain in his left ear and throat with bradycardia and syncope. Cardiology was consulted and a pacemaker was eventually placed. An otolaryngologist and neurologist also evaluated him during the hospitalization. Work up, including MRI brain and IAC with and without contrast and CTA head and neck, was unremarkable. There was no history of trauma. He was discharged from the hospital with a diagnosis of vasovagal syncope and possible trigeminal neuralgia.

He presented to our clinic several months after his hospitalization for evaluation of facial pain. The pain was described as a baseline continuous pain in the left ear with superimposed severe attacks, described as "stinging", in the left ear, maxillary sinus and occasionally the back of his throat lasting about a minute. Talking, yawning, and swallowing could trigger an attack. Since the pacemaker placement, he no longer experienced episodes of bradycardia or syncope.

His neurological examination, including cranial nerve exam, was normal.

The patient was diagnosed with glossopharyngeal neuralgia. He declined pharmacologic treatment as well as evaluation for surgical treatment options. Topical anesthesia did provide intermittent pain relief.

DISCUSSION

Glossopharyngeal neuralgia is a rare condition characterized by paroxysms of severe pain in the sensory distribution of the auricular and pharyngeal branches of glossopharyngeal and vagus nerves.1,2 Patients generally complain of a unilateral deep and stabbing pain in the throat, base of tongue, tonsillar fossa, larynx, jaw, and ear. The episodes last a few seconds up to 2 minutes.3 Some patients may have a lingering, deep, burning interictal pain. Triggers can include swallowing, chewing, yawning, coughing, and talking. Most cases of GN are primary or classical-attributed to neurovascular compression. About 10% of the patients are misdiagnosed as the more common trigeminal neuralgia,4 as was the case with this patient. The condition can also be easily confused with nervus intermedius neuralgia. Secondary causes of GN can include structural lesions including tumors (such as cerebellopontine angle tumors and oropharyngeal tumors), infection, arteriovenous malformation, trauma, carotid aneurysm, Eagle's syndrome, or demyelinating lesions.1,4,5, 8

In 2% of the cases,6 the condition is associated with bradycardia and cardiac syncope, when the parasympathetic vagal branches are more involved.2 The etiology is unclear but is thought to be based on the close anatomical relationship of the glossopharyngeal and vagus nerves in the medulla oblongata and the possibility of the formation of vagoglossopharyngeal reflex arc. It is believed that the afferent nerve impulses travelling within the glossopharyngeal nerve may reach the tractus solitarius in the midbrain and reach the dorsal nucleus of the vagus nerve, through collaterals.7 The sensory fibers of the glossopharyngeal nerve may also connect with fibers of the carotid sinus nerve (nerve of Hering), causing stimulation and activation of the baroreceptors of the carotid sinus.4 Other vagal symptoms can include voice abnormalities such as hoarseness and cough.

GPN treatment is similar to trigeminal neuralgia. Anticonvulsants are initially used, including carbamazepine, oxcarbazepine, phenytoin, baclofen, and gabapentin. Patients who are refractory to medical treatment can be evaluated for surgical interventions. If there is evidence of vascular compression of the nerve, microvascular decompression can also be effective.6,7

In summary, glossopharyngeal neuralgia is a rare syndrome presenting with pain in the distribution of the glossopharyngeal nerve. Infrequently, it can present with cardiac syncope and arrhythmia. Clinicians should consider glossopharyngeal neuralgia in patients presenting with paroxysmal pain in the throat or ear and be aware of the potential cardiovascular consequences.

TABLE 1: IHS Diagnostic criteria for classical glossopharyngeal neuralgia3

A. Recurring paroxysmal attacks of unilateral facial pain in the distribution of the glossopharyngeal nerve and fulfilling criteria B
B. Pain has all of the following characteristics:
   1. Lasting from a few seconds to 2 minutes
   2. Severe intensity
   3. Electric shock-like, shooting, stabbing or sharp in quality
   4. Precipitated by swallowing, coughing, talking or yawning
   5. Not better accounted for by another ICHD-3 diagnosis

REFERENCES:

1. Blumenfeld A, Nikolskaya G. Glossopharyngeal Neuralgia. Current Pain and Headache Reports. 2013;17(7).

2. Reddy GD, Viswanathan A. Trigeminal and Glossopharyngeal Neuralgia. Neurologic Clinics. 2014;32(2):539-552.

3. Headache Classification Subcommittee of the International Headache Society. The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018;38(1):1-211.

4. Krasoudakis A, Anyfantakis D, Hadjipetrou A, Kastanakis M, Symvoulakis EK, Marathianos S. Glossopharyngeal neuralgia associated with cardiac syncope: Two case reports and literature review. International Journal of Surgery Case Reports. 2015;12:4-6.

5. Tepper SJ. Cranial Neuralgias. CONTINUUM: Lifelong Learning in Neurology. 2018;24(4):1157-1178.

6. Rozen TD. Trigeminal neuralgia and glossopharyngeal neuralgia. Neurologic Clinics. 2004;22(1):185-206.

7. Burfield L, Ahmad F, Adams J. Glossopharyngeal neuralgia associated with cardiac syncope. BMJ Case Reports. 2016.

8. Kazemi B, Akbarzadeh F. Syncopal storm caused by glossopharyngeal neuralgia. The American Journal of Emergency Medicine. 2012;30(9).

← BACK TO PAGE 1
NEXT PAGE →

A Neurologist Goes To Business School

Eddie Patton Jr., MD, MBA, MS

WHY BUINESS SCHOOL? WHY NOW?

In the spring of 2015, I went back and forth about a decision which would have a huge effect on my career and life. I was happy practicing neurology. I was active in my county medical society, state medical society, state neurological society, and national neurological society. I felt that I had a sharp grasp of advocacy issues that were affecting not only neurology but medicine as a whole. Yet with all that said, I still felt that something was missing. Medicine was going through tremendous change, and at the root of this change was how healthcare would be managed in this country. There were new terms that I was unfamiliar with, such as physician burnout. Administrative burden and elctronic health records became catch-phrases known by all. I was doing well in my practice. We were growing and increasing service lines. I began to engage more actively with my hospital administration team about things that they wanted to accomplish such as increasing productivity and clinic access. Making the connection between what was needed from a business standpoint and from a clinical standpoint became less clear. I realize then that I was doing well in the profession of medicine but didn't know enough about the business of healthcare. In medical school and residency training, we received little training on how to manage a practice and the business aspects of providing healthcare to the patients we serve. As I think back to residency, we were never taught how to bill or things that went into reimbursements for seeing patients. It was never emphasized that in an academic practice seeing three to four patients per half day is okay but if you plan to go into private practice you would have to increase your efficiency and productivity to survive. It certainly was not emphasized how things like patient satisfaction scores would impact one's payment or the increasing amount of data reporting that you were responsible for. To maximize my efforts as an advocate for patients, neurology, and medicine as a whole, I had to learn more about business. I truly felt that in order to thrive in this ever-changing healthcare landscape, a good grasp of business administration was necessary. I didn't have years of owning my own practice as a teaching guide so I decided to go back to school.

Healthcare is consuming more and more of the discussions we are having when we talk about medicine. I remember sitting as a delegate to the Texas Medical Associations annual meeting when the presiding president tof the organization Tom Garcia, MD delivered his address. He made a comment that "we are not 'providers' and our patients are not 'consumers'. We are doctors and we treat patients." What resonated with me is that medicine should not be so transactional. It should not be treated as a mere service industry. It is more about relationships between a patient and his/her physician. The way that the business of healthcare was impacting the patient-physcian relationship fascinated me and influenced me towards going to business school.

I applied to the Executive MBA program at the Jones School of Buisness campassed at Rice University. Rice University is known for its exceptional educational programs and the Jones School of Business is ranked as one of the best business schools in the country. After talking to recent graduates and faculty, I really felt that this would be a great step towards accomplishing my goals. The networking opportunities were tremendous. The geographical location in the medical center in Houston Texas, the second largest medical center in the nation, made it an ideal place to learn more about how business relates to healthcare. It was the right time for me to enroll. My practice was growing and evolving. I was taking on more and more leadership roles in the organizations I were a part of. I worried that the lack of business experience would hurt my chances of getting into business school and, if accepted, succeeding in such a demanding program. Actually, fact that I had been out of fellowship for about six years practicing medicine before going to business school was an advantage. Practicing medicine and particpating in faculty leadership provided me with more business knowledge than I had anticipated. My involvement in organized medicine had already familiarized me with some of the most pressing issues that the house of medicine was facing. I knew that business school, in some way, would help me to put it all together and I was not disappointed.

BUSINESS AND ADVOCACY

One of the biggest benefits of having a Masters of Business Administration and how it relates to medicine is my ability to better advocate for patients and physicians. Take something like prescription drug costs. Neurological drugs are among some of the most expensive drugs on the market with treatments for multiple sclerosis and dementia leading the way. But how are these prices established? How does drug pricing compare to product pricing in other industries? There is a lot that goes into product pricing. You have to thoroughly examine the landscape and competitive products. The cost of production and aquiring raw materials must be incorporated into the cost to consumers. Supply and demand curves tell you what the market will bear. Before going to business school I thought the pharmaceutical industry seemed to diverge away from some of these basic principles of price determination. It seemed that they just plucked prices out of the air. But after examining cases such as the case of the EpiPen, I learned that these executives were following some of the very same pricing principles that were used in other industries. The CEO of Mylan pharmaceutical company, which produced the EpiPen, made a business decision to increase revenue by increasing the price of a two vial box of the EpiPen from $100 to over $600 in 2016. As we move forward in this debate and figuring out ways to reduce the cost of medications for patients, it's important to understand the business model as to how these prices are established. There have been recent high-profile cases which highlight this point. The price hike trend is hurting our ability to provide price conscientious care when pharmaceutical companies increase the price of an already

← BACK TO PAGE 1
NEXT PAGE →

lower-priced drug or entering the market with a high priced pharmaceutical product. Understanding the science behind how pricing models work in other industries will allow us to better evaluate regulatory and policy efforts to curve prescrption drug costs.

CHANGES IN PRACTICE MODELS

Medicine has gone through a number of fundamental changes in recent years. Historically physicians were entrepreneurs. They finished training, secured finances to open their practices, worked out the logistics of the clinc, hired someone to collect on what he or she was billing, and after paying the bills, received compensation for their hard work. With new regulatory rules in place it has become much harder to run a private practice. The administrative burden has become overwhelming for the practicing physician. Coupling that with decreased reimbursements makes a dire situation even worse. I was at a recent presentation of the Harris County Medical Society when a graph was shown that illustrated that physician reimbursement has not kept pace with inflation. When adjusted for inflation, physicians are getting paid less than they were twenty years ago (FIGURE 1). But we are asked to do more and more. Keeping up with EHR, reporting requirements for medicare, improving patient satisfaction scores among other things has lead to frustration and burnout. For this reason more physicians are seeking alternative practice models such as becoming an employed physician. Based on a recent survey, more than 50 % of physicians are now employed physicians. There can be benefits to this. But with it comes a loss of autonomy and control of certain processes. What you give up in accepting the administrative help is the ability to make your own financial decisions without more extensive due process.

Buisness school has equiped me with the skills to do capital bugeting more effectively. Private practitioners have been doing capital bugeting for years as a part or running their practice. If you want a piece of equipment you look at the

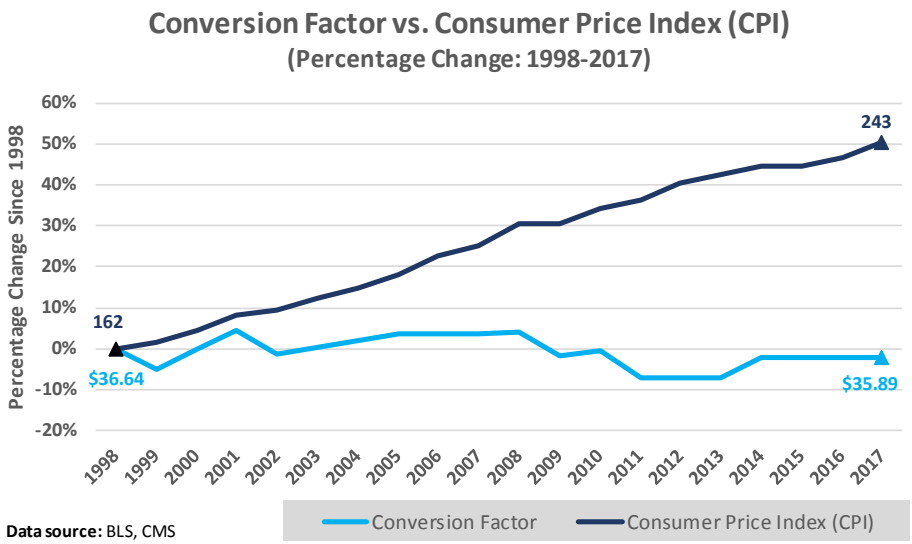


Figure 1 shows a graph of the Consumer Price Index which is one way to evaluate inflation. The CPI is graphed along with Medicare Conversion Factor (CF) which is a simplified measure of how much a physican will be reimburesed per RVU billed. The CF has remained static as CPI has increased from 1998 to 2017. Sources Bureau of Labor and Statistics and Center for Medicare and Medicaid Services.

reimbursement for that test and figure out how long it will take to pay it off. This risk to benefit analysis is simple when there is only one or two practicioners involved. When you consider a larger system, with budgets and resources that have to be dispursed among a larger pool of people, justifying a request beomes more difficult.

NETWORKING OPPORTUNITIES

One of the most surprising outcomes of my MBA is the vast and diverse network that was created by going through this program. Initially, I thought that being one of only five physicians in the class with only seven of us total in the healthcare field in a class of just over 50 would be a disadvantage. The class was heavy in the oil and gas industry and engineers. Other classmates worked in human resources, information technology or were enterpreneurers. It turned out to be an advantage. I had a different outlook and experience from other industries. This diversity turned out to be one of the hallmarks of my experience. I learned a lot about healthcare by looking at other industries. For instance the two top industries in the world right now with "big data" are the oil and gas industry and healthcare. Both of these industries have done wonders in the past few years in innovating to collect increasing amounts of data. But both industries are also struggling on what to do with this data. How do we process the data and refine it in a way that it can be useful? This is one example of many. Some of my classmates were Black Belts in Six Sigma giving them a vast knowledge about process improvement. When it comes to improving efficiency in a hospital or clinic, this knowledge is vital. By working with these individuals with years of experience and expertise, it helped me to develop and groom my own ideas about addressing clinic ineffiicencies. The Rice Business alumni network has opened doors that I havent had the chance to explore at this time, but being included in this group will help my career in the long run.

Completing an MBA program has given me a deeper understanding of our healthcare system. I am excited about the potential opportunities that come with it. It was a large sacrifice and a steep learing curve for me in the beginning. The things that I have learned will undoughtedly help me in further developing my neurology practice. This degree will have a positive impact on my advocacy efforts as well. By understanding business and how it intertwines with heathcare, I feel more prepared to have a positive impact on neurology.

← BACK TO PAGE 1
NEXT PAGE →