

Department of Neurology | Fall Issue 2021

MESSAGE FROM THE CHAIR

Dear Friends and Colleagues:

The COVID roller coaster has continued into this fall locally although hopefully we are coming off of our final surge! Unfortunately, we felt compelled to cancel our planned 50th Anniversary of the Department/ Alumni Reunion but we will reschedule for 2022. Be on the lookout for a save the date announcement.

Our department continues to expand. We added three new faculty this summer in neurocritical care, pediatric neurology and general neurology and established a new community practice in Aiken, South Carolina. On the residency front, we are excited to welcome our six new interns. Perhaps our most diverse group ever, our new residents come to us from Ecuador, China, Somalia, University of Washington, Wake Forest, and Morehouse medical school.

In this issue, we highlight our ALS center, the first in Georgia when it was established in 2007. The center offers a multidisciplinary clinic approach, access to numerous clinical trials and translational research studies and is the byproduct of Dr Rivner's devotion to his patients.

Please keep in touch and hope to see you soon.

Best Wishes,



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REMINDER!

Virtual Neurology Grand Rounds are streamed live weekly:

- Thursday mornings at 8 am
- Contact Deana Brown at (dbrown3@augusta.edu) with questions about how to join.

NEW FACULTY MEMBERS

Nilufer Yalcin, MD Assistant Professor

Dr. Nilufer Yalcin grew up in Turkey and moved to Boston, MA in 2012 shortly after she graduated from medical school. She worked as a post-doctoral research fellow for three years at Massachusetts General Hospital and Spaulding Rehabilitation Hospital with focus on cortical spreading depolarization and transcranial magnetic stimulation on stroke recovery. She completed her neurology residency training at Tufts Medical School in Boston, MA and her fellowship training in neurocritical care at Rush University Medical Center in Chicago, IL. Her main research and clinical interest is the role of cortical spreading depolarization in SAH, TBI and acute stroke patients. Outside of work, she loves spending time with her husband and her daughter, enjoys hiking, camping and wine tasting.





Tyler Vines, MD Assistant Professor

Dr. Tyler Vines a native South Carolinian, was born and raised in North Augusta, SC. He received his undergraduate degree from the College of Charleston and his medical degree from the Medical University of South Carolina. He married a beautiful woman in Charleston and then returned to the CSRA, where he completed his neurology residency at the Medical College of Georgia. In his spare time, he enjoys relaxing with his family, listening to an eclectic range of music, gardening, reading, playing games, and biking on the Greenway. He is a member of First Baptist Church of North Augusta.



Marleny Aquino-Cabrera, MD Assistant Professor

Dr. Marleny Aquino has joined the Augusta University faculty. She earned her medical degree at the Universidad Central Del Este in the Dominican Republic, where she graduated summa cum laude. She completed her pediatric residency at the Pediatric Hospital of the University of Puerto Rico. Dr. Aquino completed a child neurology residency at Boston Medical Center, and her pediatric epilepsy fellowship at The University of Texas Southwestern Medical Center, Dallas. Dr. Aquino has an interest in the care of neurologic complex children and adolescents. She is passionate about caring for children with epilepsy and believes in providing excellent patient care. Dr. Aquino's interests outside of medicine include traveling, spending time with her family, and volunteering in the local and international community.



FEATURE ARTICLE

Augusta University ALS Center

Michael H. Rivner, MD

ALS also known as "Lou Gehrig's disease" is a progressive degenerative disease of Motor Neurons. It usually starts in one region of the body and moves to other regions as the disease progresses. Death usually occurs within 5 years of the initial symptom. Besides causing paralysis of the limbs it also interferes with talking and eating. Ultimately it will cause respiratory failure which is usually the cause of death. Patients with ALS have both lower motor neuron disease symptoms as well as upper motor neuron disease symptoms. Lower motor neuron symptoms are due to loss of motor neurons in the spinal cord and brainstem that directly innervate muscles. These symptoms are muscle weakness, wasting, fasciculations and abnormal neuropathic changes on EMG. Upper motor neuron symptoms are due to degeneration of neurons in the brain that innervate the lower motor neurons. These symptoms are increased muscle tone, spasticity, cramps and increased reflexes. Finding both in a single region almost always

Goals of AU ALS Center

- Provide comprehensive and compassionate ALS care
- Provide access to ALS research
- Provide rapid and thorough diagnoses of patients suspected of having ALS
- Multi-displinary clinic
 - Physician, PT, OT, SLP, Dietician, RT, Social work
 - Equipment vendors
 - ALSA

Table 1: Goals of AU ALS Center

indicates ALS. At this time, the cause of ALS is unknown, and the treatment options are limited. It is the goal (Table 1) of the Augusta University ALS clinic to help patients with this disease by offering them the best treatment options currently available, state of the art supportive care, emotional care, and access to ALS clinical trials. Another goal is to try to develop novel therapies for this disease and do research to find its cause.

With the help of the ALS association of Georgia, the clinic was opened in September 2003 as a half day monthly multi-disciplinary clinic. Over the years our clinic has grown to a twice a month full day clinic serving around 150-200 patients with ALS and related diseases. On November 2007, we became the 31th ALSA certified clinic in the US and the first in Georgia. Our center has always been actively involved in ALS research even before the clinic was started participating in the CNTF ALS trial in 1993. In 2008 we joined the NEALS ALS consortium to become more involved in clinical research. To meet the needs of ALS patients in central and southwestern Georgia we started a satellite clinic in Macon Georgia on March 2010. This clinic also continued to grow and now serves 100 ALS patients. It has since become an independent clinic but has close ties to the Augusta Clinic as Dr Michael Rivner, the AU ALS clinic director also serves as the clinic director in Macon.

The diagnosis of ALS is often clinical. On the average, it takes 1 year for a patient to be diagnosed. The main reason for this delay is that there is no single test that is diagnostic of this condition. It requires a physician knowledgeable about ALS to evaluate all the patient's clinical information to make the correct diagnosis. Often the diagnosis is not possible until the patient has enough clinical symptoms making early diagnosis difficult. AU currently has two physicians who specialize in ALS (Figure 1), Dr Michael Rivner and Dr Ben Barnes. Dr Michael Rivner did his neurology training and neuromuscular fellowship here at MCG and has been taking care of ALS patients for around 40 years. Dr Ben Barnes did his Neurology training at the University of Kentucky and his neuromuscular fellowship at Duke University. It is their goal to try to see all patients suspected of having ALS within a month and do a thorough and prompt evaluation so the period of uncertainty about a potential ALS diagnosis is limited.



Figure 1: Clinic Director--Dr Michael H. Rivner and Clinic Co-Director Dr Ben Barnes—ALS Clinic Physicians

At the diagnostic appointment, Drs Rivner and Barnes will thoroughly go over the patient's history and exam. They will review the patient's records from elsewhere including any MRIs the patient might have. Often a diagnostic EMG test is required as part of the evaluation. The appointment is setup so that if an EMG is required it can be done on the same day. This way the patient often can be told the diagnosis on the day they visit the clinic. If they are diagnosed with ALS, patients will be started on appropriate treatments and given a return appointment to our multi-disciplinary ALS clinic. If another condition is discovered, Drs Rivner and Barnes are trained to treat all neuromuscular and related conditions.



Figure 2: Occupational Therapist—Stephanie Johnson and Physical Therapist—Tracey Rogers examining a patient.

The multi-disciplinary ALS clinic is the central feature of our ALS Center. This allows our patients to receive all needed ALS care with one visit rather than having to make many appointments. The ALS clinic team consist of clinic coordinator Ashley Creswell who makes sure patients are scheduled correctly and checks to make sure that their treatment needs are setup correctly. Our nerve conduction technologists Rebecca Fulmer and Loreen Ingram-Moore play an integral role in the running of the clinic. It is their job to do the nerve conduction studies on patients seen in our clinic which is often essential to make the correct diagnosis. In addition, they facilitate clinic flow, do the ALS rating scales and generally make sure that the clinic operates smoothly and efficiently in addition to making sure that our physician orders are carried out. Our physical therapist (Figure 2)--Tracey Rogers, occupational therapists—Stephanie Johnson and Tracey Boggs, Speech pathologists—Sarah Smith and

Allie Harrill, and Dietician—Tayler Crumpton evaluate our patient's continuing needs. It is essential that all these areas are assessed as limb mobility, speech and swallowing difficulties are part of this disease. Early intervention made possible by being examined by these therapists improves clinical outcomes. Our patients also have pulmonary function tests done by our respiratory therapists but due to COVID currently these are done by our nerve conduction technologists. It is important to monitor these functions since early respiratory intervention improves survival. We have vendors in our clinic to facilitate getting and maintaining adaptive equipment. These are Marivic Beebe, our augmentative communication specialist, Brian Byler, our DME specialist and Julie Sailors, our respiratory equipment specialist. We have representatives from both the Georgia and South Carolina ALS associations to help our patients with resources available to them from the ALS association. Finally, we have an excellent nursing staff lead by our Nurse manager Shaun Glass.

The multi-disciplinary approach ensures that our patients receive the best care possible from a dedicated team of specialists. Our allied health specialists receive special training to deal with ALS patients and their problems. Part of our goal to pick up problems early so that we can provide prompt care of all medical problems. Following clinic, our ALS team has a post clinic meeting where our patients are discussed, and we make the best plan possible for their care. The entire team is involved in creating our patient's personalized integrated care plan. We offer our patients the latest treatment options for ALS. Much of the treatment used currently is supportive care. After taking care of ALS patients for decades, we have learned much about the role of supportive care in ALS. While these treatments are not curative, they do improve life by allowing the patient to do more and delay the decline seen in this disease.

A large part of our Center's efforts revolves around ALS Research coordinated by our Research Operations Coordinator, Brandy Quarles and her assistant Kristy Bouchard. As a member of the NEALS consortium our center is involved in many ALS treatment trials. We recently enrolled the last patient in the Anelixis (AT-1501) trial. This is a preliminary trial of a monoclonal antibody against CD40. It is hoped that reducing the inflammatory response will reduce the progression of symptoms in this disease. Currently we have two open ALS trials that are actively recruiting patients, the Medicinova and Apellis trial. Medicinova (Ibudilast) is a macrophage inhibitory factor which redues the inflammatory response. Apellis (Pegcetacoplan) is a complement inhibitor acting at C3. We have several future ALS trials lined up including the Pheonix and the Healey platform trials. The Pheonix trial is a phase III study of Sodium Phenylbutyrate and Tauroursodeoxycholic acid which had promising results in an earlier phase II trial (Centaur trial). The Healey platform trial represents a novel approach to clinical trial methodology. This trial uses a shared placebo group to allow multiple drugs to be tested simultaneously. This methodology allows fewer patiens to be assigned to the placebo group. Having multiple trials running at our ALS centers allows our patients the opportunity to participate in the development of new therapies that might lead to a cure for ALS.

We are excited to have recruited Dr Eric Vitriol and his team to AU. He working on important basic research that will hopefully allow us to understand the cause of ALS. As mentioned above, the cause of ALS is unknown. In the last 40 years many genes have been associated with hereditary ALS but their relationship to sporadic ALS is unknown. Theories about the posssible etiology for ALS have not changed much in this time frame (Table 2). It is only by understanding the cause of ALS that we will be able to design effective therapies for this disease. Dr Vitriol hopes to develop an In-Vitro model of the nerve and muscle which will allow him to study the pathology of ALS. He is interested in the Actin Cytoskeleton of the motor neuron and looking at defects in it that can lead to cell death. We are also actively developing an ALS database that will increase our understanding of the clinical manifestations of ALS. Perhaps this will allow us to classify our patients better according to clincal phenotypes. Perhaps what we are calling ALS is actually

Etiology of ALS: Current Theories

- Excitotoxicity—Anti-glutamate agents, Ca influx
- Oxidative Stress—Antioxidants
- Impaired Trophic Factors
- Inflammation—Microglia, Macrophages
- Apoptosis—Programed cell death
- Glia Cells and their role
- Role of oligodendrocytes, Astrocytes, microglia
- Inclusions, ubiquitinated proteins, TDP43, Protein aggregates
- Trauma—Concussions, Military
- Prions and other infectious agents
- Breakdown in Nuclei to Cell communications—abnormal nuclear pores

 Table 2: Etiology of ALS: Current Theories

many different diseases. We are also evaluating the financial impact of this disease. Keelie Denson, a fourth year medical student, is spearheading this effort.

In summary, the AU ALS center is working hard to provide the best possible care for our ALS patients, provide opportunities for clinical research and studying the biology behind the disease.

- To schedule an apportment in the clinic contact us at 706-721-2681.
- To learn more about the research being done at our clinic go to <u>2020 Augusta University ALS Conference</u> or https://conf2020.alsauh.com

Previous funding to our clinic provided by our annual walks have helped us develop the program we have today. Unfortunately due to COVID, we had not been able to hold these walks recently.

We would appreciate donations that will allow our clinic to operate and grow. Please go to https://giving.alsauh.com/ or https://www.augusta.edu/giving/gift.php?fund=210780 to reach our donation page. Donations to the AU ALS Center are tax deductable.