American Association of Neuromuscular & Electrodiagnostic Medicine Annual Meeting

September 21-24, 2022

Neurologists, physical medicine and rehabilitation physicians, other allied health professionals, and researchers came together for the annual meeting of the American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM) to learn from one another through plenary sessions and workshops that focused on the management and treatment of neuromuscular diseases, including amyotrophic lateral sclerosis (ALS), muscular dystrophy, Duchenne muscular dystrophy, myasthenia gravis (MG), and spinal muscular atrophy.

The four-day meeting took place at the Gaylord Opryland Resort in Nashville, Tennessee, from Sept. 21 to 24, 2022. Below, we summarize some of the presentations that caught our attention at the conference. We will follow this up with a postconference wrap-up report soon.

Select sessions/highlights at the 2022 AANEM Annual Meeting

Lessons learned from the Duke MG Clinic Registry

The Duke MG Clinic Registry is composed of physician-derived data from patients seen at the Duke MG Clinic since 1980.

“This is one of the largest, if not the largest, such registry,” said Donald Sanders, M.D., professor of neurology at Duke University School of Medicine in Durham, North Carolina. He reported new findings from the dataset at the meeting.

“Myasthenia gravis is challenging to diagnose,” said Sanders. “Many people have symptoms resembling myasthenia gravis, but without proof, it is hard to make the diagnosis.”

Enter single-fiber electromyography.

Fully 80 percent of MG patients will have anti-acetylcholine receptor (AChR) and anti-muscle-specific tyrosine kinase antibodies. “Single-fiber electromyography can

confirm the diagnosis of myasthenia gravis in patients who don't have the antibodies but do, in fact, have jitter."

Jitter is a phenomenon that represents the abnormality at the nerve-muscle junction, Sanders said. "Single-fiber electromyography is the most sensitive in vivo test of nerve transmission. You can't have myasthenia gravis without abnormal jitter; it's a hallmark of the disease," he said.

Sanders and colleagues reported results on the use of single-fiber electromyography for the diagnosis of jitter in patients with MG initially seen in the Duke MG Clinic between 1980 and 2017. Jitter was measured with single-fiber electromyography during voluntary activation using conventional techniques, and the extensor digitorum or frontalis muscles were usually examined first.

Jitter was increased in 92 percent of initial single-fiber electromyography studies of 1,081 patients with MG and an additional 2 percent in follow-up studies. Facial muscles were more often abnormal than limb muscles, but no single muscle was more abnormal or more often abnormal in all patients, Sanders said.

Of the initial 1,081 patients, both extensor digitorum and a facial muscle were tested in 393 patients. Both were abnormal in 60 percent of patients. The facial muscle alone was abnormal in 26 percent, and the extensor digitorum alone was abnormal in 5 percent of patients. When the frontalis and orbicularis oculi were both tested in 73 patients, they were both abnormal in 20 percent, orbicularis oculi alone was abnormal in 42 percent, and frontalis alone was abnormal in 11 percent of MG patients.

Of 90 patients whose results were negative in initial studies, 26 percent were abnormal in follow-up single-fiber electromyography studies, said Sanders.

Most labs only measure jitter in one muscle, he added. "The facial muscles are most sensitive, and they measure one facial muscle and use the results to say whether this patient has the hallmark abnormality of myasthenia gravis," he said. But he suggested doctors instead select muscles based on the distribution of disease in specific patients.

Patients with ocular myasthenia may only express weakness or jitter in eye muscles, but patients with generalized MG may present differently. The message

from this registry is clear: “Don’t stop with just one muscle if a muscle other than a facial muscle is weak,” Sanders said.

Always be sure the muscle being tested is involved in the disease, Sanders added. “If you see a patient who doesn’t have weakness in the eyes, start with a muscle somewhere else,” he said.

Conventional MG treatments work well

Sanders and colleagues also reported treatment outcomes from 462 patients with MG initially seen at the Duke MG Clinic between 2000 and 2018 who were followed for at least one year. The treatment goal was defined as achieving a postintervention scale of minimal manifestations (MM) or better. A time-to-event analysis showed whether and when the patient reached the treatment goal starting at their first visit.

Overall, 68 percent of 462 patients achieved treatment goals within one year, 73 percent of 386 achieved treatment goals within two years, and 77 percent of 322 achieved treatment goals within three years after the initial visit. Moreover, 77 percent of those who were followed for at least three years achieved a postintervention scale of MM or better.

“A number of new treatments for myasthenia gravis have come out in the last few years and more are coming out,” Sanders said. “These new drugs are expensive, and we wanted to see how well patients did using conventional therapy.”

Conventional MG treatments include anticholinesterase agents, chronic immunomodulating treatments such as steroids and other immunosuppressive drugs, plasmapheresis, intravenous immunoglobulin, and/or thymectomy. These treatments are used in varying degrees depending on the characteristics of the individual MG patient.

There are several new MG drugs on the market, including ravulizumab for adults with generalized MG (gMG) who are anti-AChR antibody-positive, efgartigimod for gMG in adults who test positive for the anti-AChR antibody, and eculizumab for adults with gMG who are anti-AChR antibody-positive.

None of these newer agents were used in the patients in this analysis. “The treatment goal is to get patients to the point of no symptoms or disability or mild weakness in muscles that doesn’t affect activities of daily living,” Sanders said.

“The majority of myasthenia gravis patients achieved the treatment goal and they do so within a median time of less than a year,” he said.

There is no difference between men and women or those who do or do not have antibodies to the anti-AChR, Sanders added.

The message is clear: “Start with the old stuff first and only use the new molecules when you don't get a satisfactory response from the conventional drugs,” Sanders said.

Being proactive when treating Duchenne muscular dystrophy

It is time to be more proactive when treating patients with Duchenne muscular dystrophy, said Aloysia Schwabe, M.D., an associate professor of physical medicine and rehabilitation at Baylor College of Medicine in Houston, in a talk on the current thinking on the management of inherited muscle diseases.

“We have new medications and gene therapies so our patients will live even longer,” Schwabe said.

Proactivity starts with creating a multidisciplinary care team led by a motor neuron specialist with a focus on early intervention. “The goal is to be proactive, employ diagnostics available, and intervene early,” Schwabe said.

In the early stages of Duchenne muscular dystrophy when patients are aged 3 to 6 years, they may show weakness in neck flexors, difficulty with stairs, and proximal hip weakness. “Our very early patients don’t have a lot of physical limitations, but may be starting to develop silent cardiac abnormalities,” Schwabe said. This suggests a role for early screening and referring female carriers to cardiologists at diagnosis.

When kids with Duchenne muscular dystrophy reach 6 to 12 years old, they may show progressive weakness, Trendelenburg gait from a defective hip abductor mechanism, and contractures.

In terms of pulmonary complications, being proactive entails recommending exercises to promote chest wall expansion. “Cough assist is now a standard of care in kids with Duchenne and Becker muscular dystrophy,” said Schwabe. Ensure immunization is complete, including flu shots, as these kids are at a much higher risk for flu-related respiratory failure, advised Schwabe.

It is also important to monitor gait and assess function strength and range of motion every six months starting at the early ambulatory stage.

Today’s orthoses can be created by 3D printers. “They are lightweight, durable, and well-received by patients both young and old,” Schwabe said.

After age 12 years, patients enter the early nonambulatory stage and are likely to develop scoliosis and contractures, cardiomyopathy, restrictive lung disease, and upper-extremity weakness.

To get a head start on osteoporosis risk, doctors now look at lateral spine films at routine appointments to determine if patients have silent vertebral fractures and start intravenous bisphosphonates if they do, Schwabe said.

“With the advent of steroids, the incidence of scoliosis in Duchenne muscular dystrophy has decreased, but if you don’t receive steroids, there is a high incidence of scoliosis. Discuss steroids with patients and their families at diagnosis, and start treatment during the early ambulatory state,” Schwabe advised.

As adults, these patients can become nonambulatory, dependent on a ventilator, thin, and at risk for cardiac decompensation and contractures.

“As the disease progresses and patients become more and more physically disabled, we start to transition from pediatric care to adult care,” Schwabe said.

“We need to be better at supporting them during this transition and [provide] needed anticipatory guidance on how to handle scenarios that present themselves, including emergencies,” said Schwabe.

And the winner is…

Richard Bedlack, M.D., Ph.D., received the AANEM 2022 Public Recognition Award at the meeting. This award honors folks who have made extraordinary contributions toward increasing public awareness of muscle and nerve disorders.

Bedlack is a professor of neurology at Duke and director of the Duke ALS Clinic. He is involved in the international *ALSUntangled* program, which uses social networking to investigate alternative and off-label treatment options for patients with ALS. He also started the ALS Reversals Program, which seeks to better understand cases of recovery from ALS and to make these happen more often.

“I am very grateful that the AANEM has recognized my work,” said Bedlack. “I’ve had so much fun and met so many amazing people with ALS who have taught me so many important lessons.”

Bedlack has had some of these lessons embroidered on his suit so he never forgets them. “Never take the little thing in life like motor neurons for granted, and it is possible to heal patients of their symptoms even when we can’t cure the underlying disease,” Bedlack said. “ALS patients may seem fragile, but never underestimate their resilience, and hope is the most powerful thing we have.”