Dr. Feelgood

Using sartorial style and a patient-centric approach, **Richard Bedlack** offers optimism in his quest to cure and treat those with ALS.

BY DANNY HOOLEY

PHOTOGRAPHY BY ALEX BOERNER + SHAWN ROCCO

In the examining room at the Duke Neurological Disorders Clinic, Richard Bedlack looks more like the eccentric rock star he wanted to be growing up than the physician he became.

At a youthful fifty-one, he wears his salt-and-pepper hair in an upswept typhoon wave. On this September day, he's wearing a midnight purple suit and black pointy-toed boots, his long, lanky frame evoking a mid-'60s Carnaby Street style à la British mod-rock hero Paul Weller (or singer Elvis Costello, if your eyes fixate on his big black-rimmed glasses).

But there's a weathered black leather doctor's bag on the floor next to him. It was a gift from his parents when Bedlack graduated from the University of Connecticut School of Medicine in 1995. Despite the finery he's wearing, it's the old-fashioned accessory that reveals the most about him.

> "I save my best outfits for Tuesday, when I see my ALS patients."

At the moment, sixty-eight-year-old Martha "Marty" Stephens has his full attention. She was diagnosed with amyotrophic lateral sclerosis (or ALS) in March 2016.

She's still verbal, and she gets around mostly on a three-wheeled Pride Go-Go scooter. Stephens does a lot less walking with her braces now. She gets winded when doing simple tasks, like getting dressed.

Bedlack performs a quick but thorough physical exam, tapping her elbows and knees. Her right knee doesn't show much reflex. Stephens tells Bedlack her left hand is weaker than it was at the last visit. She's right-handed.

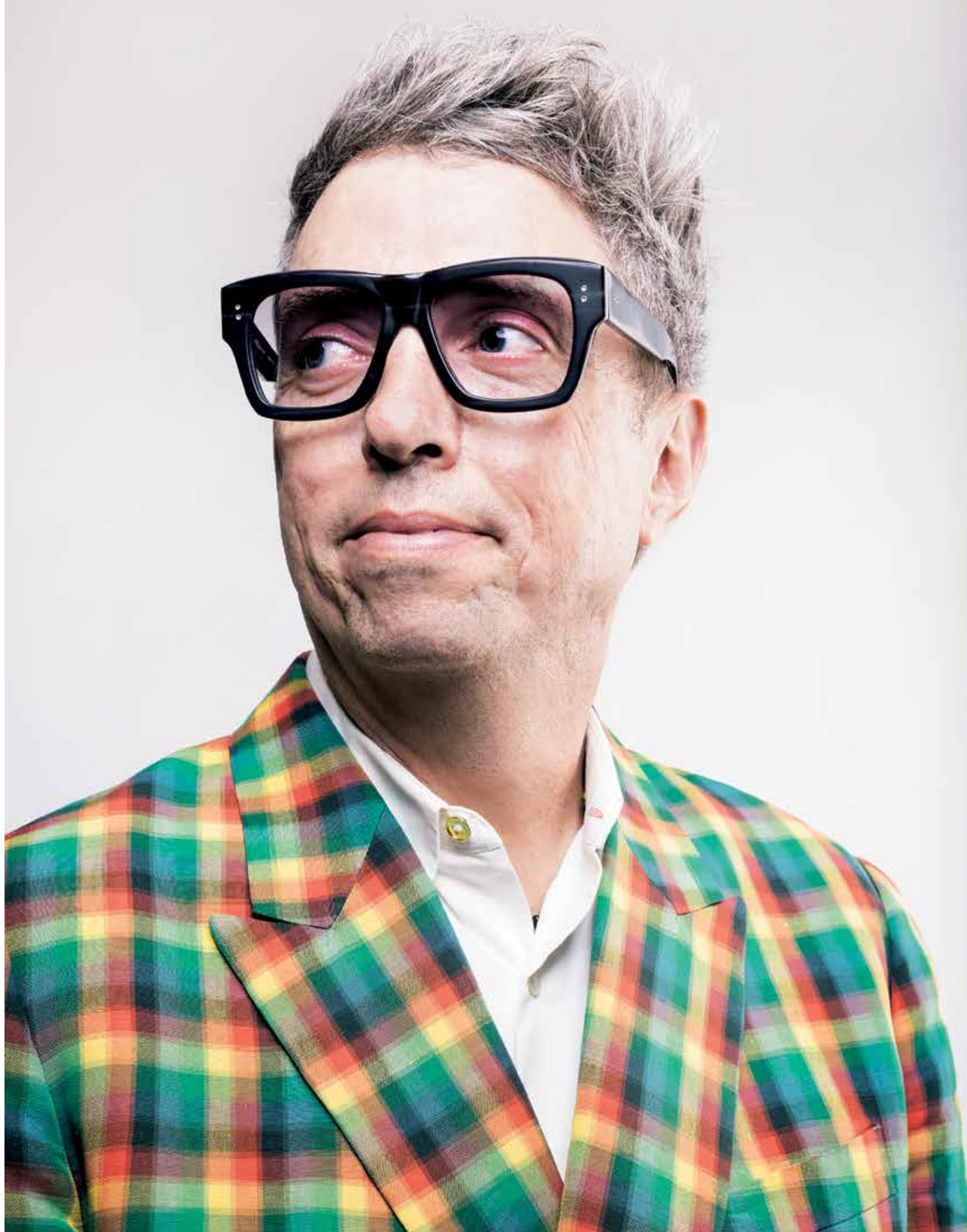
"Are you still cutting your food okay?" Bedlack asks.

"I get…a lot of help," she answers, as her husband, Jim Stephens, watches nearby.

In his chair, Bedlack leans in to listen as she talks. He tells her that the weakness in her arm and both legs has not progressed as much as he expected. "You were progressing pretty fast for a while there," he says. "I don't know why you've leveled out, but I'll take it. Have you been taking any supplements that might account for that?" She replies that she hasn't.

Before he leaves the room, Bedlack offers to help her get in touch with a technician to adjust the motorized wheelchair at home that's been vexing her. Then he asks her to look over some upcoming ALS studies when she gets back and perhaps choose one to participate in—preferably one that doesn't involve too many doctor visits, for her own comfort and convenience.

Her husband drives her from their home in Fuquay-Varina about every ninety days for an appointment here. As always, the Stephenses have a long day ahead of them. They already had visited with occupational and physical therapists that day, before they even got to Bedlack. "It's a multidisciplinary thing," says Jim Stephens. "We see somewhere between six and eight people every time we come out."



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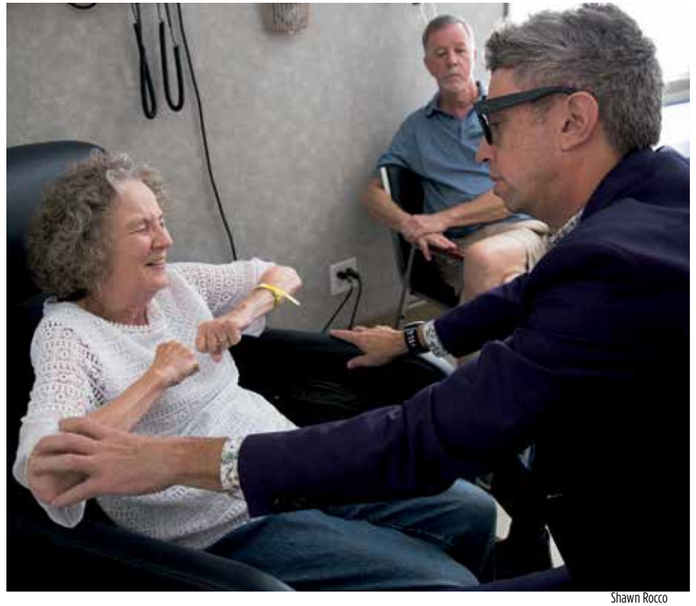


Shawn Rocco

Rick Bedlack, as he likes to be called, is a renowned expert on ALS—sometimes known as Lou Gehrig's disease—an often-baffling disease that attacks nerve cells in the brain and spinal cord in diverse ways, and progresses at different rates with different patients. ALS can't be diagnosed with one definitive test. Approved treatments can slow the progression but not reverse the damage of ALS on the body.

"It's so **uplifting** to go there. Even with no promised cure in sight. These people are doing their best to try to make your life better."

GENTLE MAN: Richard Bedlack examines Martha Stephens, as her husband, Jim, watches.

Bedlack is an active advocate for research and an innovator in his approach to clinical trials. He's been seen on CNN, ESPN, and Fox News. He's been covered by *The Wall Street Journal*. He's in three ice-bucket challenge videos on YouTube. He travels the world (recently, to Reykjavik, Iceland) about sixty days a year to speak at ALS conferences and participate in golf and poker events to raise money for research.

He even walked the runway for 2015 Men's Fashion Week in Paris with British fashion designer Sir Paul Smith, who invited Bedlack and his wife, Shelly Miller, as guests, after he heard from a store manager that Bedlack regularly wore his suits.

Bedlack's colorful style, patient-centered philosophy, and open mind have made him the face of optimism against a terrible disease—not just in the media, or behind a speaker's podium, but in the examination room, where optimism really counts.

The exquisitely tailored dark-blue suit he wears one Wednesday, as he sits at a table in a conference room at the clinic, is rather demure by his famously flashy standards.

"I save my best outfits for Tuesday, when I see my ALS patients," he says. "You can imagine—it's a scary place for those folks to come. Because a lot of measurements are made. Unfortunately, sometimes those measurements are worse than they were the time before."

Progression of the disease necessitates a discussion of options that can be hard for a patient to hear. *Do you need to stop driving? Do you need to start using a breathing machine?* "I try to break the ice by dressing in a fun way," Bedlack says. He also strives to empower his patients during a period that can feel lonely and hopeless. Bedlack says he'll never forget the words of one speaker with ALS, at a previous event in Iceland.

"He said: 'All the doctors and scientists out there, I just want you to remember something. This disease is about people. It's not about test tubes, and mice, and things like that. And nothing about us should ever be done without us.'"

Bedlack pauses a moment to let that sink in. "And he's right."

Duke's ALS Clinic runs from 8 a.m. until around 7 p.m. every Tuesday. When he created the clinic in 2001, Bedlack was only given half a day, once each month.

It's grown from a skeleton crew to a staff of three speech therapists, two occupational therapists, two physical therapists, two respiratory therapists, a pulmonologist, a social worker, a clinic coordinator, an ALS Association representative, an ALS research nurse, and Bedlack.

"We have, probably, one of the largest, most comprehensive ALS clinics in the world," Bedlack says. "We have more than 400 patients that we follow." He adds that more than a hundred are on the waiting list. That infuriates him. "We're booking people for March 2018," he fumes. "That's not acceptable for a disease that rapidly progressive, [that] massively disabling, where we know they may not be getting options they should be getting, wherever they are."

Bedlack says it's gotten to the point where he needs at least one more full day to devote to ALS. So far, he hasn't gotten his department to agree. "We have so many folks that are waiting for appointments, and so many research opportunities, that we need another day. And that's one of the battles that I'm fighting right now."

As of early October, he's getting much-needed help from Ashley Whyte-Rayson, a Neuromuscular Fellow and one of his former residents. Now, she's his partner—and maybe someday, the inheritor of his legacy.

Bedlack came to Duke in 1995, a decision he made years after passing it up for his undergraduate studies in favor of The College of William & Mary, whose smaller size he found more relatable to his working-class upbringing in Cromwell, Connecticut. That sensibility lingers. Even though he lives right by the private Hope Valley golf course in south Durham, he'd rather tee off on the public Hillandale course with his friends on the weekend.

His father, whom he affectionately likens to Homer Simpson, worked at a nuclear power plant. His mom slowly worked her way up at Farmers and Mechanics Bank—from a sixteen-year-old teller to vice president—after putting herself through night college when he and his younger brother were little.

By the time he was thirty and fresh out of UConn, he was no longer even slightly intimidated by the size of Duke. "I came to Duke because I had this idea that I would specialize in some rare neurological disease," Bedlack says, "and build a clinic around it that was different—that offered some research."

He found a hero in Duke neuroscience professor James McNamara.

"He's built his life around epilepsy, with the idea that it's fixable," Bedlack says. "He's got one foot in clinic, trying to treat people with epilepsy with the drugs that we have. But he's got his other foot in the lab, trying to unlock the molecular mechanisms of different kinds of epilepsy in animals."

Bedlack found "his" disease that year when he was making his rounds as a resident with an attending physician. Then, as now, the resident goes in and talks to the patient, reports to the attending, and they go back in together to discuss findings with the patient.

One day, Bedlack examined a patient with ALS for the first time. "I thought it was the most amazing disease I'd ever seen," he recalls. "I thought the exam was the most stunning physical exam—I mean, it's an incredible collection of neurological abnormalities. And I was very excited to go out and see how we were going to take this terrified, tearful person and their family and empower them to have some control over what's happening."

The next conversation was a letdown for Bedlack—and a crushing blow to the patient.

"We went back in, and the attending basically said, 'This is what it is, and there's nothing anybody can do for you. You should go home and get your affairs in order.' And there were more tears, and we just walked out."

Bedlack recalls what he said to himself that day as he was driving home. "This is Duke. There's gotta be more things we can do."

He couldn't jump into lab research like McNamara, because "it's become a more complicated life for physicians" since McNamara started, he says. "We have a lot more administrative duties in and out of the hospital than physicians ever had before."

But he could do clinical research. After he finished his residency, he did a neuromuscular fellowship. He went through the Duke Clinical Research Institute Master's in Clinical Research program. Then he decided to open an ALS clinic.

Shawn Rocco

FEARLESS: Marjorie Lynne Bryan texts sentences on her cell phone to communicate.

Bedlack says that doubters were hung up on the importance of relative value units (RVUs), a measure of value for physician services in the U.S. that originated with Medicare reimbursement. "That's one way that the bean counters determine how useful you are," he remarks, clearly irritated at the thought. To this day, Bedlack is obligated to participate in similar budgetary discussions, and he's not a fan. "They always tell me how much money I'm losing for the university. It's never been about RVUs for me."

His main champion in 2001 was the head of Duke Neurology at the time, Warren Strittmatter, now a professor emeritus. Bedlack recalls that Strittmatter liked his obvious passion for fighting ALS and decided to give the proposed clinic a year and "see what happened." At the time, says Bedlack, there were two ALS clinics in the state. Even supporters of Bedlack's idea had to question whether North Carolina needed another clinic for such a rare disease. How would Duke's ALS clinic compete for pharmaceutical sponsors?

Bedlack started showing up at ALS conferences, and he thinks now that his "crazy appearance" may have helped him get noticed. He sat down with experts. He asked questions. He offered his clinic as a site for clinical-trial pharmaceutical tests on volunteers.

Within two years, Duke's ALS clinic got its first trial, for creatine, a supplement used by athletes to increase

muscle strength. "It didn't work" for ALS, says Bedlack, "but that wasn't the point." The point was to establish that his ALS clinic could quickly enroll patients for clinical trials, while maintaining compliance with protocols and avoiding dropouts.

It helped that by this time, Bedlack had a lot of patients he could recruit. Word-of-mouth about a clinic where patients were made to feel like they were involved in the process of their treatment brought in a lot of them, quickly.

Marjorie Lynne Bryan, a sixty-six-year-old retired middle-school art teacher from Flowery Branch, Georgia, who goes by the name Lynne, was ready to hear tough news when she first came to see Bedlack back in January of 2017. She started experiencing slurred speech nearly four years ago. "She had difficulties, not just getting misdiagnoses, but not getting the care that she wanted in Atlanta," says her husband, Mike Hammett.

> "I thought the exam was the most stunning physical exam—I mean, it's an incredible collection of neurological abnormalities. And I was very excited to go out and see how we were going to take this terrified, tearful person and their family and **empower** them to have some control over what's happening."

Bryan's initial diagnosis was for myasthenia gravis, a neuromuscular disease that disrupts normal communication between the brain and muscles. About a year's worth of treatments weren't showing any progress, and Bryan noticed that the symptoms weren't really matching the diagnosis, according to her own online research.

She and Hammett went to another neurologist, who concluded she had primary lateral sclerosis (PLS), a milder, slower-progressing and non-fatal form of ALS. That diagnosis seemed less likely when Bryan lost speech abilities and had difficulty swallowing and eating. Her right hand continued to lose function.

It took about six months, from the time Bryan found Bedlack on the Internet to their first visit to Durham in January of this year. Bedlack diagnosed her correctly in February, after exhaustive tests.

Bryan can no longer speak at all. She communicates by texting sentences that appear in large type on her cellphone display. Every so often, she'll comically raise her hand to shush Mike when she wants to say something.

On this visit, the couple, married for six years, tell Bedlack they plan to head out the next day to the beach at Hilton Head, South Carolina, maybe for the final time.

Bryan is adamant that she doesn't want a tracheostomy tube as a life-saving nor life-prolonging measure, in case the worst happens at the beach or anywhere else. She asks Bedlack to sign a Do Not Resuscitate order for her. He does, and he advises her to keep it with her at all times while she's at Hilton Head.

"She's brought that up, almost every visit," Hammett says afterward. At one point during her time with Bedlack, she types, "I'm not afraid to die," and shows it to the doctor.

Hammett praises Bedlack for the team he's assembled. The couple usually stay in Durham for two or three days, to be available to the staff for any follow-up. "It's so uplifting to go there," Hammett says, "even with no promised cure in sight. These people are doing their best to try to make your life better."

The ALS clinic is one of four main programs Bedlack created. He cofounded the ALS Clinical Research Learning Institute in 2011. The annual two-day event in Clearwater Beach, Florida, is dedicated to training selected ALS patients to learn more about studies and new therapies and become research ambassadors (advocates for clinical research). "It gives them the tools that they need to be thought leaders," Bedlack says.

He also created the ALS Untangled website, a forum for patients, clinicians, and scientists to review "alternative and off-label ALS treatments."

Soon after Bedlack started his clinic, he discovered that patients were going online in search of "something else out there" to cure ALS. Some were self-experimenting with supplements and other alternative therapies. Not surprisingly, they were reluctant to discuss it with a doctor. "They're afraid they're going to encounter some serious paternalism," Bedlack says.

In other words, they anticipate hearing *well, that's just silly* and *if that really worked, don't you think everybody would already be doing it*? ALS Untangled opens a dialogue that is respectful, empowering, and informative. "I've come to believe that most people aren't snake-oil salesmen," Bedlack says. "Even the proponents of these alternative therapies. I think that most of them have a good heart. Most of them are true believers." They just haven't pursued the science, he adds.

Another of his websites, ALS Reversals, opens its first page with: SOMETIMES PEOPLE WITH ALS GET BETTER.

That's rare, of course. Bedlack only knows of thirty-four verified reversals, which he defines as "dramatic improvement in at least one objective outcome measure"—in layperson's terms, a significant gain in at least one motor function. And that conclusion is only reached after the possibility of a condition that may be mimicking ALS's symptoms is thoroughly ruled out.

He wants to find more reversals, if they're out there, in hopes of someday replicating them. The website asks patients who may have experienced a reversal to report it and to supply medical records to verify that, indeed, a reversal has occurred.

The project has yielded potentially useful information. Dan Harrison, a medical student Bedlack worked with last year, compared demographics of those patients who showed reversals, noting the kinds of supplements they were taking. That information was compared to larger databases of people with more typical ALS.

The reversal patients were found to be younger and less likely to be white. They showed quicker initial progression. Their symptoms were more likely to start in the legs.

He also found that three out of thirty-four were taking a curcumin supplement that he plans to study next. (Curcumin is a chemical derived from the spice turmeric.) That's not to suggest he favors alternative theories over conventional medicine.

He's working on a Phase III clinical trial of the drug Tirasemtiv, a fast-skeletal-muscle troponin activator (FSMT) developed by the San Francisco company Cytokinetics. Very few drugs even make it past Phase II of the four-phase process, so Bedlack is excited by the possibilities. "We'll have results by the end of the year," he says. "And we'll see if this becomes mainstream therapy—something that helps people hold on to their strength and breathing muscles."

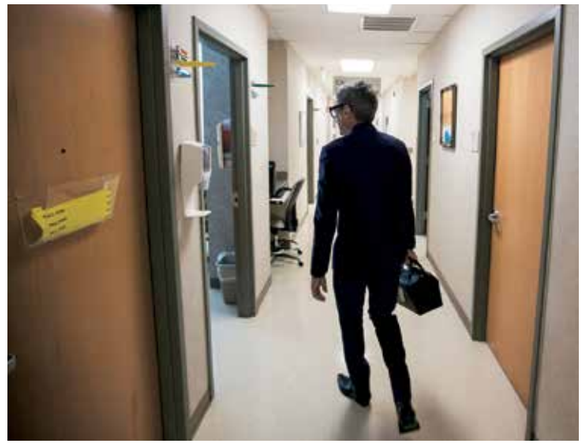
He's also optimistic about a trial for the anti-epileptic drug Retigabine. "This comes from a brand-new model of ALS," he explains. "Traditionally, our models have been based on abnormalities inserted into animals. That might not be a great model for someone who doesn't have a genetic abnormality." That includes most of his patients, whose ALS is sporadic (which means that the patient is the only member of their family to have ALS). The cause of that is still a mystery.

Experiments using skin scrapings from ALS patients have shown that cells were abnormally excitable. "That might be part of the reason why they die," Bedlack says, adding that Retigabine works well in restoring cells to normal excitability and resistance to toxins.

Bedlack says he's also happy about a yearlong trial for lunasin that's just wrapping up. Based on suggestions from his research ambassadors, he designed it so that patients at any stage of ALS could participate, and that patients could take their own measurements at home, which cuts out extra doctor visits. Plus, there was no placebo. ALS patients tend to frown on placebos.

In the absence of placebos, Bedlack used "historical controls" for comparison. For each patient enrolled in his study, he noted their progression rate prior to taking lunasin, then found, on a database of ALS patients, three documented cases not taking lunasin who had matching progression rates.

Like the creatine study that helped get him started, the lunasin trial doesn't appear to show dramatic results. But again, there's a positive to consider. The success of the trial's design is what excites him.



Shawn Rocco

TREATMENT: The ALS clinic is one of four programs Bedlack created to research and treat the disease.

"It was the fastest-enrolling trial in the history of ALS," he reports, proudly. He says he plans to use this model for future studies.

He's also seeking funds from outside the university to add more days to the ALS clinic. An annual $30,000 Tele-ALS Grant from the North Carolina chapter of the ALS Association allows him to spend Thursday afternoons connecting with patients over secure video conferencing. For one thing, he says, it allows patients at home to still see him and laugh at his crazy outfits.

"As I get funding for things, I can buy my way into doing more ALS," he says.

"I would do ALS every day if I could." ■

Hooley is a Durham-based writer, musician, and adjunct lecturer at North Carolina Central University.