

# Special Protocols for Special Patients



By Mike Rubin

When a condition is easily treatable but rare, how much should EMS try to do?

Ca-ssis/iStock/Thinkstock

**D**anny was 7 days old when he had his first bleed.

“It was just a bruise,” says Darlene Shelton, his paternal grandmother, “but from that point on he never had fewer than three of them at a time. It should have been a clue that something wasn’t right.”

Danny’s doctors didn’t realize their young patient had hemophilia, a rare clotting disorder, even when his gums started to hemorrhage at 10 months. “It was during his afternoon nap,” Shelton recalls. “A very small cut caused a horror scene of blood two hours later.”

Danny’s parents decided not to wait for an ambulance and drove him to the closest hospital, 20 minutes away in Tuscaloosa, Ala., where the ER staff stopped the bleeding and sent him home. “They didn’t do any special testing, even though it was entirely too much blood for normal gums and took way too many measures to get it stopped,” Shelton says.

Danny bled again the next morning—same spot, followed by a visit to the same ER, only this time the doctor ran a series of clotting tests that led to the hemophilia diagnosis.

“Even if they’d caught it sooner, the clotting factors Danny needed weren’t any closer than Birmingham—an hour away,” says Shelton.

“Danny’s hemophilia isn’t as severe as it could be, but the longer he has to wait for treatment, the greater the risks.”

How prepared should EMS be to help patients like Danny? To what extent can we treat rare diseases pre-hospitally?

The first step is to focus on what’s treatable.

## How Many, How Sick?

Approximately 30 million people in the U.S. have at least one of roughly 7,000 illnesses considered rare, meaning they affect fewer than 200,000 people—about 0.6% of the population.<sup>1</sup> Eighty percent of those 7,000 diseases are genetic, and half of the sufferers are children, 30% of whom will not live until their fifth birthdays.<sup>1</sup>

Only 5% (350) of those illnesses account for 80% of patients with rare diseases.<sup>1</sup> The list of conditions for EMS to even consider treating is much smaller—perhaps no more than a few dozen—because 95% lack FDA-approved therapy.<sup>1</sup>

**Darlene Shelton is spearheading a campaign to rewrite protocols that would allow EMS providers to treat her grandson, Danny, who has hemophilia.**

After culling out the untreatable, EMS still faces these issues:

» *Incidence of rare disease within the region*—Are there patients in your catchment area known to suffer from any unusual-but-treatable conditions? If not, can you review a year or two of data to see if any of those illnesses show up in patients' medical histories? Preparing for acute episodes might be worthwhile even without prior cases.

» *How prepared patients and their families are to treat exacerbations*—Some families are practically self-sufficient in that they have the means and training to handle most flare-ups. Others will rely almost entirely on prompt assistance from EMS.

» *Whether treatment is time-sensitive*—How many of the diseases being considered will worsen if not treated prehospitally? How long can patients afford to wait?

» *Whether treatment is affordable and falls within prehospital providers' standards of care*—Do responders need supplementary



training, protocols, supplies or equipment to treat targeted rare illnesses? If so, at what costs?

### **Low-Incidence, High-Consequence Care**

One region that has done its homework and been proactive about treating uncommon ailments is Long Island's Suffolk County, a 900-square-mile New York City suburb that's a target-rich environment for textbook trauma and illness. Among the county's 1.5 million inhabitants, though, are some with conditions barely covered in those textbooks—the ones

most EMS providers read, at least.

"A number of years ago, we heard about a child out east with congenital adrenal hyperplasia (CAH)," says the county's EMS director, Bob Delagi, whose department oversees more than 100 distinct agencies answering over 120,000 calls annually. "We developed a protocol for administration of Solu-Cortef specifically for the squad treating that patient."

Solu-Cortef, a steroid, supplements the body's own stress hormones—often critically short in CAH victims. The disease can be

# *You're awesome*

## AND WE APPRECIATE YOU!

Thank you for dedicating your  
life to saving others.



**Happy EMS Week!**

May 21 -27, 2017

For More Information Circle 21 on Reader Service Card

fatal without prompt care, which is why some CAH sufferers have their own supplies of Solu-Cortef for acute episodes. Even those families usually welcome assistance from experienced responders. The question is, how practical is it for EMS agencies to offer such specialized care?

“We have to make sure what the patient needs doesn’t exceed the provider’s scope of practice or the agency’s resources,” Delagi says. “CAH was relatively easy, partly because it’s similar to Solu-Medrol, another steroid we carry, and partly because Solu-Cortef is inexpensive (less than \$15 per dose).”

Customizing care for small subsets of patients can be an administrative burden, though, because those responsible for medical direction and QA/QI need to know which providers can do what in the field. That gets complicated when treatment protocols vary by agency. Delagi and his staff looked for an opportunity to incorporate CAH into the county’s standard of care.

“As we became aware of more children in the area with adrenal insufficiency, we rolled out training first to the local agencies, then eventually to everyone. Now care for CAH is folded into our countywide protocol for pediatric hypoperfusion.”

CAH isn’t the only rare condition recognized by SCEMS, says Delagi: “We have about 30 residents with LVADs (left-ventricular assist devices). We were one of the first regions to put out an advisory to EMS providers about those patients. We also offer clinical training through Stony Brook Hospital, a regional LVAD center.”

Suffolk County’s EMS protocols include three pages about LVADs, including probable emergency scenarios and recommended actions for both ALS and BLS providers.

“We’re just trying to be as progressive as we can with these low-likelihood, high-consequence conditions,” Delagi says. “We can’t address them all, but we do want to give our providers the education and tools to assist patients whenever possible.”

## A Missouri Initiative

Meanwhile, back in Tuscaloosa, Danny was needing emergent care for bleeding two or three times a year. The Sheltons had started carrying Danny’s clotting factors in their car in case he had an acute episode away from home.

In 2014, after 4-year-old Danny and his family had moved closer to Darlene in southeast Missouri, she discovered state law prohibited paramedics and even ED docs from administering Danny’s own IV drugs to him.

“It’s scary to live with a rare disease and carry your own lifesaving medication, knowing you’re the only one who can give it,” says Shelton.

“Danny’s meds cost about \$50,000 a month—thank God for insurance—but we try not to stress the cost. If there’s an emergency, you want paramedics to feel at ease about giving those drugs. It’s not hard: You pop a cap off a glass vial and mix with saline, then draw it into a syringe. There’s no reason to be afraid of messing it up; Danny can do it himself when he’s not sick. But when I speak with veteran paramedics, they’re still reluctant to administer any medications not in their possession because their protocols don’t cover that.”

So what can a proactive, protective grandmother do to change that mind-set? She can rewrite the protocols, which is what Dar-



## EMERGENCY MEDICAL SCIENCE

### Earn Your Degree Online!

#### Paramedic Certification (Hybrid Program)



- Fully CAAHEP Accredited
- Hybrid Paramedic program offered through LCC Continuing Education Department. Only four on-site visits required for skills training and evaluations. All coursework is done online. Tuition is \$360.
- Clinicals can be completed in your area. Contact LCC regarding available areas.
- Different course options that allow currently credentialed EMTs, AEMTs, or individuals with no certification to train for their Paramedic certification.
- Graduates are eligible for the NCOEMS Paramedic exam and the National Registry Paramedic exam.

#### Associate Degree in Emergency Medical Science — Bridging

- All degree classes offered 100% online.
- Currently credentialed state and national Paramedics earn up to 45 credits toward their degree just for being certified!
- Designed for demanding EMS work schedules. Complete the degree at your own pace!

Ranked Third in the Nation by [bestcolleges.com](http://bestcolleges.com)!



231 Hwy. 58 South,  
Kinston, NC  
[www.lenoircc.edu](http://www.lenoircc.edu)



(252) 527-6223, ext. 115

✉ [jgtilghman38@lenoircc.edu](mailto:jgtilghman38@lenoircc.edu)

For More Information Circle 22 on Reader Service Card

lene is doing with help from Missouri's State Advisory Council on EMS.

"They really stepped up and told me, 'We're going to help you fix the problem,'" Shelton says. "We have a legal team and medical advisors. We're getting there."

That legal team originated a bill to address protocol changes and cover liability across the state. "EMS is regulated service by service in Missouri," Darlene explains, "so you need a law to provide regional protocols statewide."

"Usually the next question is, 'How is the training going to be paid for?' Fortunately we have lots of home health agencies and drug companies on board with certified nurse educators. They cover things like cardiac and endocrine disorders, in addition to hemophilia."

"A big part of the training is just raising awareness about people with rare diseases carrying their own medications and the importance of treating them as soon as possible. That way, when they get to the hospital, the priority can be evaluation rather than emergent care."

In case you're wondering, neither Darlene nor anyone in her family has a medical background. "My experience is in accounting," she says. "My husband's a general contractor, and Danny's parents are engineers. We're just a very active family trying to help not only Danny but others with rare illnesses."

"We're not political people. When we embarked on this mission, it was a huge learning curve. We decided to start with Missouri, but I've been having conversations with groups in California and Texas who've been monitoring our progress."

"I'm hoping we can change protocols nationwide within five years."

For more information on treating rare conditions, visit Shelton's website, [www.dannysdose.com](http://www.dannysdose.com).

## REFERENCE

1. Global Genes. *Rare Diseases: Facts and Statistics*, [www.globalgenes.org](http://www.globalgenes.org).

## ABOUT THE AUTHOR



**Mike Rubin** is a paramedic in Nashville and a member of EMS World's editorial advisory board. Contact him at [mgr22@prodigy.net](mailto:mgr22@prodigy.net).

**MACS is Making Ambulance Crews Safer with the Mac's Bariatric Ambulance Lift.** With a 1,300 lbs. weight capacity the lift is prepared for any job you can roll on it. The platform is universal and will accommodate whatever equipment you choose to use. The Bumper Stow Technology makes the lift available to the ambulance crew at all times and functions as a bumper and step.

**Don't Strain Your Back...Go See Mac!**

Ambulance Built  
by Excellence Inc.



[sales@macsliftgate.com](mailto:sales@macsliftgate.com)  
[www.macsliftgate.com](http://www.macsliftgate.com)

**800-795-6227**

For More Information Circle 23 on Reader Service Card

**ON-LINE OR ON-CAMPUS:**  
Earn your bachelor's or  
master's degree through WCU!  
[emc.wcu.edu](http://emc.wcu.edu)



**ESTABLISHED IN 1977, WCU'S EMERGENCY MEDICAL CARE PROGRAM WAS THE FIRST PARAMEDIC PROGRAM LEADING TO THE BACHELOR OF SCIENCE DEGREE IN THE NATION.**

- Undergraduate concentrations are available in health services management or in science (pre-med)
- Accredited by Committee on Accreditation of Educational Programs for EMS Professions (CoAEMSP)
- EMS management and EMS education concentrations available in the Master of Health Sciences program

For more information:

828-227-3516 / [mhubble@wcu.edu](mailto:mhubble@wcu.edu)

**Western  
Carolina**  
UNIVERSITY

For More Information Circle 24 on Reader Service Card