

# MDA/ALS NEWSMAGAZINE

The Latest Information About Amyotrophic Lateral Sclerosis • March/April 2010 • Vol. 15, No. 2

## ALS TDI Progress Report: Full Steam Ahead

### Inside:

When nature calls:  
Toileting with ALS

What happens at the end  
in ALS?



# Research Roundup



## Value of high-fat, high-calorie diets to be tested in ALS

An MDA-supported study comparing three tube-feeding formulas — one high-calorie, normal-fat; one high-calorie, high-fat; and one average-calorie, average-fat — in people with ALS is now being conducted at the five centers that make up the MDA/ALS Clinical Research Network.

The trial will test the hypothesis, suggested by observational studies and studies in mice, that a high-fat and/or high-calorie diet can increase survival in ALS. Participants must be tolerating tube feedings and must meet other study criteria.

The centers that make up the MDA/ALS Clinical Research Network are California Pacific Medical Center, San Francisco; Columbia University, New York; Emory University, Atlanta; Massachusetts General, Boston; and Methodist Hospital, Houston.

For more information, contact Katy Shaver at Massachusetts General Hospital at (617) 643-7434 or [kshaver@partners.org](mailto:kshaver@partners.org); or see the Clinical Trials section of the MDA Web site ([www.mda.org](http://www.mda.org)).

## Trial of SOD1 blocker now open

An MDA-supported, phase 1 clinical trial of the experimental drug ISIS-

SOD1-Rx in people with the SOD1 familial (inherited) form of ALS opened in February 2010.

This study is being performed at Massachusetts General Hospital in Boston and five other U.S. sites. Timothy Miller at Washington University has received MDA support to work with Isis Pharmaceuticals of Carlsbad, Calif., to develop ISIS-SOD1-Rx.

The trial will test the safety and tolerability of this “antisense” compound, which is designed to block production of the SOD1 protein in people who have developed ALS because of inherited mutations in the SOD1 gene. Such mutations result in ALS in approximately 1 percent to 3 percent of cases.

The investigators will infuse ISIS-SOD1-Rx into the fluid that surrounds the brain and spinal cord, a delivery method that targets the cells that produce the toxic SOD1 protein.

Details of genetic testing and other study requirements will be discussed with potential study participants. Contact Pat Andres at (617) 724-8995 or (877) 458-0631 (toll-free) or [andres1@partners.org](mailto:andres1@partners.org).

## Researcher studying altered communication in ALS, other diseases

Speech-language pathologist Carolyn Baylor at the University of Washington in Seattle is seeking help from people with ALS and other diseases that can affect speech and communication in developing a questionnaire for health care providers to use. Participants must use speech as their primary communication method, although they may use writing or augmentative devices at times.

“Our goal is that this questionnaire

will be one of several tools that clinicians can use to help people with communication disorders improve their participation in activities they want to do,” Baylor explains on her Web site at <http://staff.washington.edu/cbaylor>.

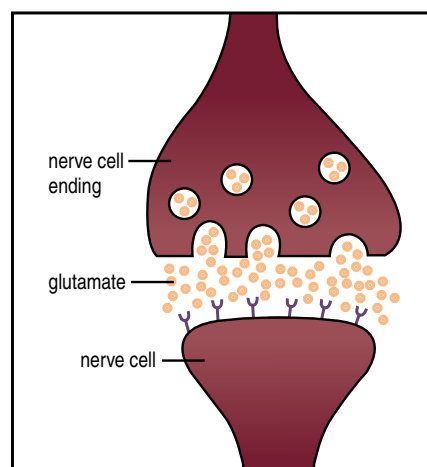
Participation involves about an hour of filling out questionnaires at home, online or on paper. Contact Carolyn Baylor at (206) 221-3563 or [commpart@u.washington.edu](mailto:commpart@u.washington.edu).

## Ceftriaxone study remains open

A 540-person trial of intravenous ceftriaxone in people with ALS remains open at locations throughout the United States and Canada.

Ceftriaxone is an antibiotic that may increase the levels of a protein that helps clear potentially toxic glutamate from the areas surrounding nerve cells. Laboratory studies suggest the drug may help protect motor neurons, the main cells damaged in ALS. In this study, ceftriaxone is being administered intravenously.

Contact Sarah Titus at (617) 726-1398 or [stitus@partners.org](mailto:stitus@partners.org). □



Glutamate is a necessary carrier of signals between nerve cells. However, it has to be cleared away from cells regularly to avoid toxic buildup. Ceftriaxone may aid in that process.

# ALS TDI: Moving Forward on Multiple Fronts

## Drug development, new partnerships, new Web features and more were covered in ALS TDI's first research update of the year

*In January 2007, MDA and the nonprofit biotech ALS Therapy Development Institute (ALS TDI) of Cambridge, Mass., launched a three-year, \$36 million drug discovery partnership. In January 2010, MDA extended that partnership with a \$2.5 million grant. The biotech's Web site is [www.als.net](http://www.als.net).*

ALS TDI's first research update webcast of 2010, hosted on Jan. 14, included a rundown on the status of the Institute's drug development pipeline and its leading therapeutic, ALS TDI 00846; recent grant and collaboration news; an innovative addition to its online message board (ALS Forum); and an introduction to the "Dashboard," the face of the Institute's information management system.

## Pipeline progress

ALS TDI Chief Scientific Officer Steve Perrin provided an update on ALS TDI's leading therapeutic candidate, a protein biologic known as ALS TDI 00846.

The compound is an antibody (immune system protein) designed to modulate the immune response through a pathway called CD40L. ALS TDI scientists have determined that in mice with an ALS-like disease, CD40L pathway activity is increased ("up-regulated") in the spinal cord, skeletal muscle and sciatic nerve.

ALS TDI 00846 hits the CD40L target, a particular cell-surface recep-

tor that is present on a number of different types of cells associated with the immune system, Perrin reported.

Multiple ALS TDI mouse studies of the compound have shown a small but significant increase in survival, muscle function and survival of motor neurons (which normally die in ALS); and a decrease in the harmful activation of normally supportive cells called astrocytes.

Perrin noted that once scientists were confident the drug was working, they then began trying to figure out how it worked. Pinpointing that mechanism may help in the identification of related drugs with better safety profiles, greater specificity, or the ability to hit similar pathways that produce an even greater effect.

"It's a very important question," said Perrin. "Once we find a drug that has an effect similar to the size of the effect we're seeing with this drug, we could treat them as a combo therapy and try to amplify the effect with a combination of the two drugs at various doses."

Study results in mice have shown that the drug knocks down almost every one of the approximately 20 genes in the CD40L pathway. But it remains to be seen whether the CD40L pathway data from mice with ALS is relevant to humans with the disease.

Over the last three years, Perrin noted, ALS TDI has partnered with several MDA clinics nationwide that have been collecting samples of blood, muscle, skin and fat from people with ALS. The collection currently contains approximately 400 blood samples, which scientists at the Institute have profiled, along with an equal number of non-ALS samples, in order to measure patterns of gene activity (gene



"expression"). They found a subset of patients — approximately half — have highly upregulated genes in the CD40L pathway.

Perrin affirmed that a great deal of work has been done on 00846, including testing in hundreds of mice. He said the Institute hopes to move the compound into clinical trials by the end of 2010, and that it's working to find a pharmaceuticals partner to help move the compound through testing and into the clinic as quickly as possible.

In addition to 00846, Perrin noted, ALS TDI has many drugs in its pipeline and ALS TDI researchers are working on 30-plus different potential therapeutics.

## New funding, partnerships

ALS TDI Communications Director Rob Goldstein reported on a number of developments on the Institute's funding front, including a renewal of "critical funding" from MDA.

Goldstein thanked MDA for providing "a renewal of their partnership and investment in ALS research," with the new \$2.5 million grant aimed at continuing the search for therapeutic

tic targets in ALS. The funds were awarded through MDA's Augie's Quest, a fast-track ALS research initiative spearheaded by ALS Co-Chairs Augie and Lynne Nieto. Four times a year, representatives from MDA and ALS TDI, along with a number of expert ALS researchers, will review the goals and progress associated with the milestone-driven grant.

Another key funding partnership, announced Jan. 12, has come in the form of a multiyear grant awarded through the 2010 Defense Appropriations Act, signed by President Obama on Dec. 22, 2009, and provided to ALS TDI through the Department of Defense. The money will fund development and advancement of new therapeutics.

Goldstein also reported on the Applied Proteomics partnership announced Dec. 12, 2009, that will use animal and human samples to identify and validate protein markers for ALS. Protein markers — changes in protein levels that correlate with disease progression — are useful for diagnosis and analyzing response to treatment.

The work “will help us to further understand and prioritize what's going on,” both in the ALS model and with human samples collected from clinics, “to identify key points that we can attack with new therapeutics,” Goldstein explained. This in turn may help speed clinical trials by allowing phase 1 trials to be bypassed.

Goldstein said the proof-of-concept work is complete and data will be presented at the next ALS TDI research update webcast in April.

## New Web site feature

The ALS Forum — ALS TDI's lively and comprehensive message board for people with ALS and their caregivers — has welcomed the newly formed ALS

Forum Response Committee. The committee meets once a week to discuss some of the topics raised on the Forum, and posts responses designed to inform participants and help steer discussions.

“These posts are not meant to be an ‘answer’ or to replace any specific advice you are given by your personal medical team,” said Perrin in announcing the new feature.

The hope, said Goldstein, is that the experts' responses will contain “both information from the preclinical, and experience from the clinical side of research,” and that these responses “will enable readers to become better informed and therefore better armed to make treatment decisions in consultation with their personal medical team.”

Forum moderators at ALS TDI will identify topics from among those that come up on the message boards and submit them to the committee for review and response. There are no plans to allow direct questions from readers as, Goldstein said, “the idea is to add value to the conversation taking place between ALS patients, caregivers and others on the ALS Forum — not replace that system altogether.”

Response Committee members include three ALS experts with close ties with MDA: Merit Cudkowicz, Jonathan Katz and Stanley H. Appel. Other members are ALS TDI pre-clinical experts John Lincecum and Fernando Vieira, and medical/science writer Carey Goldberg.

MDA research grantee Cudkowicz at Massachusetts General Hospital in Boston also is director of the MDA/ALS center at that institution and serves as an adviser to MDA's translational research program. Jonathan Katz serves as co-director of the MDA/ALS Center at California Pacific Medical Center in San Francisco.

A longtime MDA adviser and research grantee, Appel is director of the MDA/ALS Clinical Research Center at the Methodist Neurological Institute in Houston. An MDA clinic director since the 1970s, Appel is a member of MDA's Board of Directors, has served as chairman of the MDA Scientific Advisory Committee and currently serves as an adviser on matters concerning MDA's translational research efforts.

Forum readers may access the experts' responses at this direct link: [www.als.net/aboutus/committee.aspx](http://www.als.net/aboutus/committee.aspx).

## 'Dashboard' points the way

Perrin also briefed viewers on “The Dashboard,” the face of the ALS TDI information management system that is an effort to bring together all information on ALS, including thoughts and comments on journal papers and related literature, lab processes and any relevant “information in the scientist's brain.”

The Dashboard electronically captures data and puts it into a database, and tracks scientist input. It's hoped it will inspire ideas that lead to projects and then serve as a project management tool by helping researchers gauge where the project stands, what's holding it up and how to proceed.

The idea, Perrin said, is to “capture live and archive forever.”

## Next up

The next quarterly ALS TDI research update will take place April 22 at 6 p.m. EST. To participate in the webcast, go to [www.als.net](http://www.als.net). Registration (free) is required.

The first-quarter 2010 webcast has been archived on the Institute's Web site. (Click on “Get Involved” tab at the top of the page, then click on the “Research Update” banner.) □

# Share Your Story, Make a Difference

ALS doesn't play favorites. Athletes and couch potatoes, parents and grandparents, even teenagers — anyone can get it. The letters "ALS" truly could stand for "Anyone's Life Story."

For the past three years, in order to put a human face on this little-known but devastating disease, MDA has posted the online series "ALS: Anyone's Life Story," during May's national ALS Awareness Month. Each day in May, a different person with ALS is profiled online, telling about the individual's hopes, fears, challenges and most of all, determination.

Local media are alerted when a member of their community is featured in Anyone's Life Story, sometimes leading to additional newspaper and TV coverage about the disease and its affect on individuals and families.

Right now, MDA is selecting 31 individuals from around the country for this year's series. Might you or someone you know be a good candidate? If so, contact your local MDA health care service coordinator and let her or him know of your interest by **March 15**. (You can be connected to your local office by calling 800-572-1717.)

To view the profiles that have run in past years, go to the MDA/ALS Division Web site ([www.als-mda.org](http://www.als-mda.org)) and click on Resources at the top, then Anyone's Life Story. □



## Letter to the Editor:

### Gulf War vets with ALS not forgotten

In your January-February issue, I found your article about Gulf War ALS ("Research Roundup: Soil bacteria implicated in ALS-Gulf War connection") very interesting. My husband Randy is one of those 40 from the Gulf who have ALS. Randy has outlived most of the veterans who were diagnosed with Gulf War ALS. Randy is also one of the Marine officers who testified before Congress about his exposure to toxins, which led to the ALS/Gulf investigation. It is nice to know these veterans have not been forgotten and studies are still being done to prove what we have suspected for years.

Randy has done extremely well with ALS, his positive attitude and motivation are encouraging. In your articles, let people know life does not have to stop just because you have ALS.

Kim Hebert  
Emerald Isle, N.C.

# The Ups and Downs of Hygiene Slings

by Bill Norman

When nature calls, a hygiene sling may be just the right answer for many with ALS.

Like standard slings that are used to transfer, transport or reposition patients, a hygiene (toileting) sling is suspended from a patient lift that can be maneuvered to the toilet on wheels or a ceiling track.

But unlike standard slings, hygiene slings allow lift passengers to stay in the sling while using the toilet, making the whole process quicker and easier.

## Commode models differ

Although some slings are characterized as commode slings and serve the same purpose, their design is considerably different than that of hygiene slings.

Commode slings typically have a fairly small hole cut in the bottom of the sling fabric, which usually fully encompasses the buttocks. Hygiene slings are open from the middle or upper back all the way to the back of the thighs just above the knees.

Because of their open design, hygiene slings are easier to use without making a mess. They also facilitate personal cleanup after toileting.



Victor hygiene sling in combination with a portable lift.

## Design is important

When selecting a sling of any type, several factors need to be considered, especially since costs can range from less than \$100 to \$1000 and more:

**Weight and size of the user.** The load-bearing capacity of slings can vary from several hundred pounds to more than half a ton. The same is true of the lifts that support slings. Both need to be capable of accommodating the patient's size and weight.

**Special physiological conditions.** Lift users who require head support should use a high-back sling with a

safety belt, advises the Lift Doctor, a panel of clinical and bioengineering lift specialists at Liko ([www.liko.com/na/](http://www.liko.com/na/), 888-545-6671). Regardless of the user's physical abilities, all slings should have a safety belt option, "in order to avoid any possibility of your patient slipping while in the sling."

**Function.** Hygiene slings are specifically designed for toileting and bathing use.

**Sling material.** Most slings are fabricated of nylon and available with either padded or unpadded leg flaps. Don Krebs, owner of Access



*Victor hygiene sling with head support function.*

to Recreation ([www.accesstr.com](http://www.accesstr.com), 800-634-4351), warns that unpadded slings can bunch up and pinch the skin. He also recommends sling material that can be washed easily.

If the sling will be used to transfer the user in and out of the shower or bathtub, mesh fabrics that dry quickly are an option. Vancare ([www.vancare.com](http://www.vancare.com), 800-694-4525) offers slings and



*Slings from Waverley Glen are color-coded by size and use Velcro-fastened belts for security.*

belts made of disposable material to help with infection control. The company also sells sling fabric with a nonabsorbent coating on one side to aid spot cleaning.

Waverley Glen ([www.waverleyglen.com](http://www.waverleyglen.com), 800-265-0677) makes sling seat belts with hook-and-loop (Velcro) fasteners for easily securing the user.

**Lift/compatibility.** Some companies that sell both lifts and slings maintain that their slings are not compatible with other

manufacturers' lifts. "This is no doubt explained as a liability issue," says Diane Huberty, 61, of Fort Wayne, Ind. Huberty, who received an ALS diagnosis in 1986, also is a neuro-certified registered nurse. "But the truth is that nearly all slings will fit nearly all lifts."

## Adaptive clothing works

Huberty writes a regular column ([www.living-with-als.org/Diane](http://www.living-with-als.org/Diane)) called "From Both Sides — Caring for an ALS Patient." From long-time firsthand experience with hygiene slings, she has acquired knowledge that's pertinent not only to the slings themselves, but also to clothing that's compatible with their use.

"Nobody mentions getting your pants down!" she complains on her site of sling makers' otherwise glowing descriptions of their products. "No one tells you that in order to use a sling lift for toileting, you have to give up wearing slacks and underwear! Well, I refuse to sit around bare-assed

under a drafty skirt or lap blanket waiting for a call of nature!"

Huberty and her caregiver tried making the sling work while she wore regular slacks, but getting the pants up and down proved a struggle. "That led me to try adaptive clothing," she recounts.

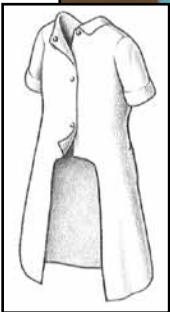
She tried slacks with zippers on both sides, but they were difficult to zip back up after re-seating in her wheelchair. Ultimately, she settled on slacks with an open back, sold by Silvert's Adaptive Clothing & Footwear ([www.silverts.com](http://www.silverts.com), 800-387-7088). "As weird as they sound, they look like ordinary slacks when you're seated," she writes on her Web site. "Of course, you can't wear underwear



*SureHands hygiene sling suspended from a ceiling-track lift.*

with them. I just put a hand towel in the wheelchair seat instead.

"When you transfer to the toilet, you don't have to do anything with them. No removing, pulling, unzipping, unsnapping needed. Just transfer to the toilet and go! There is plenty of open space underneath. I have never gotten my slacks wet or soiled. A pit



*Buck & Buck cut-away muu-muus and dusters are adaptive but not revealing. Inset: Rear view of Buck & Buck's cut-away duster.*

stop with the help of a caregiver who's familiar with the process takes less than 10 minutes, even with me on a ventilator.”

Buck and Buck ([www.buckandbuck.com](http://www.buckandbuck.com), 800-458-0600) also sells adaptive clothing, offering open-back and overlapping-back muu muus and dusters for women, and pants for men with two overlapping back panels held together by easily detached hook-and-loop tabs.

### Choose carefully

Huberty recommends doing an Internet search to find the brand and model of hygiene sling that's best for your particular needs, and then searching

further for the best price. Among the Web sites she searched were [www.livewellmedical.com](http://www.livewellmedical.com) (877-748-3935) and sites mentioned earlier in this article. She also notes that slings don't work without lifts, and that the latter should be selected as carefully as the former.

Several manufacturers suggest that slings should, in most cases, be taken out of service and replaced after two years of use — or sooner if necessary. Sling condition can be affected by frequency of use, disinfectants, detergents, frequency of washing, temperature of the wash/rinse cycles and weight of the user. □



*Silvert's open-back wheelchair pants (above) are fashionable and sling-functional.*

# To Cath or Not to Cath?

People with ALS, caregivers and professionals ponder the value of internal urinary catheters

by Margaret Wahl

When ALS weakened Jane Cheng's mother, her caregivers found it took a great deal of strength to help her transfer on and off the toilet.

"I twisted my ankle a few times when attempting to turn the pivot disk with my foot while supporting most of her weight," says Cheng, who cares for her mother in central Pennsylvania.

The family eventually obtained a patient lift, which helped with transfers. However, says Cheng, "because of the time it took to complete a transfer, my mother sometimes felt like she didn't have to urinate anymore by the time we got her on the toilet successfully."

The eventual solution for Cheng's mother, and for at least a few others with ALS, was to get a Foley catheter, which drains urine through a tube in the urethra that's anchored in the bladder by a small, inflatable balloon (see illustration, page 10). It's also called an internal or "indwelling" catheter, or a "urethral" catheter. Urine drains continuously into an attached bag, which can be hooked to a chair, wheelchair, bed rail or leg (under clothes).

"The Foley helped reduce the number of times she had to get in the lift," says Cheng, "and I think that being able to rest without worrying about getting up to use the restroom was a relief. Personally, I was relieved when the Foley was put in, because



it decreased the risk for injury during transfers; helped my Mom conserve energy for other more important matters, such as spending time with friends and family; and helped caregivers conserve energy for more important matters, such as providing care for my mom."

Cheng even speculates that "maybe my mom's ALS wouldn't have progressed as rapidly if she had not spent all her time and energy getting up and down from the toilet." And, she says, "It's sad to even try to think about the things we could have done together during that time, instead of struggling in the bathroom."

## The 'pee problem'

Almost everyone with ALS faces difficulties with handling urination at some point. The problems are rarely with urination itself, but with handling the process in a dignified, quick and convenient manner, without jeopardizing health.

Medical professionals often balk at the insertion of any type of internal

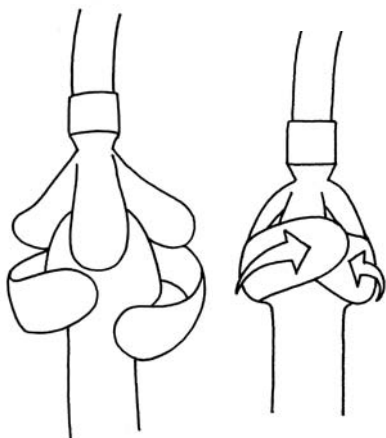
urinary catheter because of the fear of infection. But some people want them anyway and are willing to incur some risk for the other advantages.

In the initial phases of ALS, a urinal often is a good solution to the "pee problem." These are easier for men than women to use, but there are some new styles that work for some women as well (see Resources, page 11). Urinals can be tucked into a large bag when away from home, as it's generally easier to use one in a public restroom than to transfer on and off a toilet.

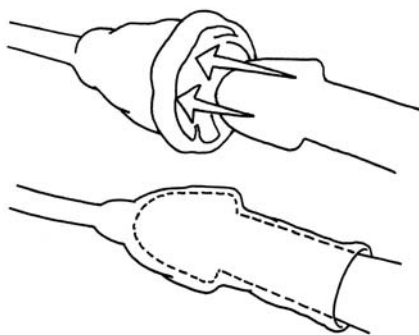
If urinals become unworkable as ALS progresses, the next step could be a mechanical patient lift to help facilitate toilet transfers, or an external urinary drainage device — again easier for men than women, although not out of the question for both sexes (see Resources, page 11). Other than skin irritation, leaks and detachments, there's virtually no risk with an external catheter or urine collection device.

For women in particular, though, the external devices are tricky and may not provide the necessary coverage, comfort or convenience.

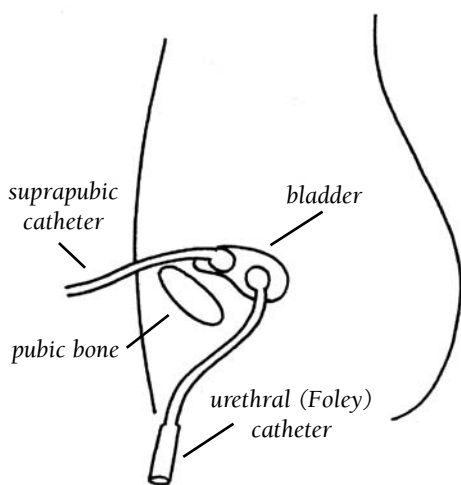
## External and Internal Catheters



BioDerm's Liberty external catheter seals to the tip of the penis.



Condom-style catheters attach to the penis like a condom.



Foley catheters are inserted through the urethra in males and females. Suprapubic catheters are surgically inserted through the abdomen into the bladder in either sex.

## Foley catheters: The downside

Medical professionals generally frown on Foley catheters and other internal catheters for ALS patients. ALS, they say, usually doesn't affect urinary functions specifically, even though in general, muscle weakness can make it very difficult to get to a bathroom, use a toilet or sometimes even use a urinal or bedpan. (Some people with ALS also experience urinary urgency.)

Their main objection to an indwelling catheter is the danger of infection this type of device poses.

"It's easy for a Foley catheter to become contaminated by bacteria," says nurse Lora Clawson, director of ALS Clinical Services at the MDA/ALS Center at Johns Hopkins University in Baltimore. "The anatomical structure of where the urethra is in relation to the rectum leads to cross-contamination, even with normal female hygiene, let alone a catheter."

Antibiotics can be used to successfully treat urinary tract infections, but, Clawson cautions, they're not without their own risks. "Anytime you're exposed to an antibiotic, you can develop antibiotic resistance," she says. "After a time, there are constant resistant bacteria present. Antibiotics also can cause nausea, vomiting, diarrhea, skin rashes and yeast infections."

She prefers that ALS patients try other solutions before opting for an indwelling catheter. "There are some new designs of female urinals that I encourage patients to try," Clawson says. "The mouth of the urinal has to be form-fitting to a woman's body."

Some women, she says, have come up with "creative solutions," such as wearing a dress without underwear, sliding down to the edge of the wheelchair and then peeing in a cup or other container.

For men, she says, anatomy

makes the urinary challenges much easier. Many men with ALS can use a traditional urinal without much trouble. They also have the option of an external catheter, which fits over the penis and connects to a tube (see illustration, left). It doesn't carry any of the infection-related complications of the indwelling catheter, although it can cause skin irritation.

Gabriela Harrington-Moroney, a nurse at the Eleanor and Lou Gehrig MDA/ALS Center at Columbia University Medical Center in New York, is also unenthusiastic about indwelling catheters because of concerns about infection and the difficulty of getting patients the nursing care that these devices require.

Having an internal catheter of any type, she notes, "requires nursing intervention. A nurse has to go out every four weeks or so to change the catheter, so you have to have an ongoing visiting nurse service in place, and sometimes insurance doesn't want to cover it." (She recommends that any catheter be changed every four to six weeks and that a nurse regularly look for signs of infection or other problems.)

On the other hand, she concedes that there are problems for people who need assistance to get to the toilet or use a urinal as well. "We've had some patients with urinary tract infections because their home health care worker may go home at 8 p.m., and they don't urinate all night because they can't get up on their own."

And then, there are those quality-of-life issues, which go beyond the medical issues and can be hard to assess.

"We talk a lot as nurses and physicians about quality of life for patients," says Harrington-Moroney. "Yes, maybe there are downsides and pitfalls to catheters, but with patients with ALS, what quality of life do they want to have? Some of them won't go

to dinner because they can't get to a bathroom fast enough and don't want to wear a diaper. Maybe we should look at that more closely."

## Loves her Foley

Karin Kallweit of Toronto, a former nurse, was only 31 when she first noticed weakness and muscle wasting in her right hand that would later prove to be ALS. Now, as she approaches her 42nd birthday, she has almost no movement from her shoulders down but manages to live in her own apartment with only part-time assistance.

When she was still able to use a walker, she found she couldn't unbutton her pants in time in the bathroom. Now, she can no longer transfer to the toilet at all.

"I used pads for a little bit," she says, "but since I'd rather not have anybody during the day, one of the problems was that you need to get them changed regularly. I used to be in nursing, so I knew people who had catheters. I thought, you can always take it out if you don't like it." (Note: This may not be possible in all cases, because of bladder changes over time caused by the continuous drainage of urine.)

Her request was not initially greeted with enthusiasm. "I went to the ALS clinic and asked the nurse about it, and she acted like it was the most horrible thing," Kallweit recalls. "She said, 'You're going to get bladder infections.' She made me go to the urologist at the hospital, probably thinking he would talk me out of it. But I talked to him, and he said, 'Sure,' and put one in right in the office."

That was eight years ago. Kallweit has had two bladder infections (each after a bowel accident) and some difficulties with clogging of the catheter, but she's never regretted her decision.

"I don't need anybody during the day to change diapers," she says. "I go out whenever I want, because I don't worry about smelling like pee or sitting in a wet diaper. I'm up in my chair all day. I'm in downtown Toronto, so there's always something going on. If I go out with my girlfriends and I want to have three or four cups of coffee, I don't worry about it."

Kallweit admits the Foley isn't for everyone and does have potential complications. "If you don't drink enough, or if the catheter has a little defect, the little hole at the end of the part that is in the bladder can become plugged up. It happens every four to five months."

To help prevent infections and clogging, Kallweit drinks plenty of fluids and takes cranberry pills. She also takes a drug called Ditropan, to counteract bladder spasms.

"If you're someone who doesn't like drinking, then the catheter is probably out for you, because you'll get more infections if you don't drink enough," she says.

For herself, she says, "I wouldn't live without it."

## Suprapubic catheters

Foley catheters aren't the only game in town when it comes to internal urinary drainage.

A suprapubic catheter, instead of being inserted through the urethra, is inserted into the bladder through the abdomen, in a surgical procedure requiring anesthesia (see illustration, page 10). As with Foley catheters, suprapubic catheters can cause infections. But they have their fans too.

## Freedom and comfort

"Walk a mile in my shoes and you'd understand," says Sandra Scott about

## Resources

### At Home Medical

Suwanee, Ga.  
(770) 476-0490  
(800) 526-5895  
www.athomemedical.com  
External urine collection systems for men, external collection system for women (Convatec Female Urinary Pouch).

### BioDerm

Largo, Fla.  
(727) 507-7655  
(800) 373-7006  
www.bioderm.us  
Unique Liberty external collection system for men (video on Web site).

### West Marine

Watsonville, Calif.  
(800) 262-8464  
www.westmarine.com  
Little John portable urinal for men with Lady Jane adapter for women.

her choice of a suprapubic (above the pubic area) catheter. Scott, 60, has ALS and lives in Portage La Prairie in Manitoba, Canada.

Like Kallweit, Scott values her independence and doesn't like the idea of having to transfer to the toilet every few hours (for which she needs the help of a patient lift) or having to wear diapers. In short, she says, she didn't like having to have her life "planned around urine."

Scott at one time considered getting a Foley catheter, but she rejected the idea.

"I had experienced them before and didn't appreciate the feeling of a tube 'there' and how it felt being inserted and taken out," she says. "Also, I'm a married woman with a normal, albeit changed, sex life."

She asked her pulmonary specialist for a referral to a urologist and did some Internet research on catheters "not only for medical facts but for people with them. I could still speak and had no problem being my own advocate," Scott says.

Her suprapubic catheter was inserted above the pubic bone during outpatient surgery. The area around the insertion site ("stoma") is cleaned twice a day, and the tube is changed every four weeks by a home health care nurse. The bag is changed weekly. "It's simple to care for," Scott says.

Scott says she has a better quality of life with her suprapubic catheter than she had without it, because it's clean and odor-free, she doesn't have to wear diapers or worry about being away from her lift, and she no longer has rashes or a "sore, burning bottom" from sitting on pads in a chair.

She and her husband sleep better without "having to wake up at least twice and usually more to go through the rigmarole of putting me on and off the bedpan."

Scott says she can "wear proper clothes and underwear" and drink as much as she wants without worrying about additional trips to the bathroom.

"Increased liquid will decrease the possibility of bladder and lung infections and skin breakdown and aid bowel function," she says. "Generally, it's a lot easier to manage than 'regular' voiding."

Scott says there have been "no embarrassing accidents" and "more freedom and comfort overall" with the suprapubic approach.

## Making the decision

Consideration of an internal catheter requires a discussion with your doctor and almost certainly a referral to a urologist.

The urologist will likely assess:

- your general state of health;
- your ability to care for the catheter, including having a professional change it regularly;

- your willingness to drink plenty of fluids (or take fluids via a feeding tube) and take other recommended infection-preventing measures;
- the accessibility of medical care if complications should arise; and
- your ability to withstand anesthesia (suprapubic catheter only).

Unlike other medical interventions in ALS, such as a tracheostomy or feeding tube, there are no clear medical benefits to internal catheters and in fact, there may be medical downsides, such as the risk of infection. The main benefit, say those with internal catheters, is improved quality of life for themselves and their caregivers. These folks often take a very pragmatic view.

As one man with ALS said of his suprapubic catheter, "What's another hole in this body, anyway?" □

## MDA/ALS NEWSMAGAZINE

*Jerry Lewis*  
National Chairman

*Gerald C. Weinberg*  
President & CEO

*R. Rodney Howell, M.D.*  
Chairman of the Board

*Timmi Masters*  
Secretary

*Suzanne Lowden*  
Treasurer

*Olin R. Morris*  
Chairman, Executive Committee

*Stanley H. Appel, M.D.*  
Chairman, Medical Advisory Committee

*Louis M. Kunkel, Ph.D.*  
Chairman, Scientific Advisory Committee

*Augie & Lynne Nieto*  
Co-Chairs, MDA ALS Division

*Nancy O'Dell*  
National ALS Ambassador

*Christina Medvescek*  
Editor

The MDA/ALS Newsmagazine is published bimonthly.  
(800) 572-1717 • [www.als-mda.org](http://www.als-mda.org) • [publications@mdausa.org](mailto:publications@mdausa.org)  
©2010, Muscular Dystrophy Association. All rights reserved.





## A Peaceful Passing

### Medication, care coordination and advance planning leave little to fear at the end

by Amy Labbe

It likely crosses the mind of every individual who receives a diagnosis of ALS, if not immediately then soon thereafter — but it's the question no one wants to bring up.

*What happens at the end?*

Despite their reluctance to ask, people want to know: When the end comes is it going to hurt? Will I choke or feel like I'm suffocating? Will it be scary?

The answers to these questions, like the experience itself, can't be identical for everyone, but overall the course of ALS does tend to follow a fairly predictable path. If you're more comfortable *not* knowing about the end of your journey with ALS, perhaps this isn't the read for you. But if you're one of those who wants, who *needs* to know, then read on for some words of wisdom — and reassurance — from a number of ALS experts.

### What will happen?

The vast majority of deaths in ALS are the result of respiratory failure.

Other far-less-common causes of death include malnutrition as a result

of swallowing problems, pulmonary embolism (a blockage in one of the arteries of the lungs), abnormalities in the heart's electrical pacing system called cardiac arrhythmias, and pneumonia as the result of aspiration (when food or fluid gets into the lungs).

Mark Bromberg, director of the MDA/ALS Center at the University of Utah in Salt Lake City describes respiratory failure as a process that progresses slowly over months.

Shortness of breath develops, first with activity such as dressing, then when lying flat or sitting quietly.

At this point, people may experience a lack of energy or find they have difficulty sleeping.



Finally, says David Lacomis, director of the MDA/ALS Center at the University of Pittsburgh, “the individual’s carbon dioxide levels build up, and they slip into painless coma.”

A common phrase heard from family members whose loved ones have passed is, “It was very peaceful at the end.” Rather than struggling and gasping for air, their loved ones “went to sleep and just slipped away.”

## Will I choke or feel like I can’t breathe?

Choking, and the panic that can be induced by shortness of breath, are often the first and worst fears after a diagnosis of ALS.

“Patients fear choking,” says Lacomis, “but that would be an exceedingly uncommon cause of respiratory arrest.”

And Julie Rowin, director of the new MDA/ALS Center at the University of Illinois in Chicago, notes that although it’s common for people to worry about choking to death, she has never seen it actually happen.

As respiratory failure progresses, some may feel a shortness of breath that’s exacerbated by anxiety or fear, but in most cases medication is ad-

ministered to provide relief.

Most commonly, explains Merit Cudkowicz, director of the MDA/ALS Center at Massachusetts General Hospital in Boston, morphine (a narcotic painkiller that also acts on the central nervous system as a mood enhancer) is used to prevent any discomfort caused by shortness of breath. Depressants called benzodiazepines, such as Ativan, reduce anxiety.

At MDA clinics, physicians work closely with palliative care teams to coordinate treatment with in-home hospice care providers, assisted living facilities or inpatient hospice settings. Such cooperation helps ensure the person with ALS has the most peaceful and painless experience possible.

Hospice care focuses on providing comfort and quality of life in the final months or days, by supporting the physical, emotional and spiritual needs of the individual with ALS and their family members. (For more information on the ins and outs of hospice, see “Not Gloom and Doom: Demystifying Hospice,” January/February MDA/ALS Newsmagazine.)

## Is it different on a vent?

The decision to use assisted ventilation when breathing becomes diffi-

## Asking the Question No One Wants to Ask

In August 2009, Barbara Durkacz of Newcastle upon Tyne, England, wrote in a post on the ALS TDI Forums ([als.net/forum](http://als.net/forum)) an “unpleasant kind of question,” for which she hoped to find answers. She noted that what she was wondering about had been “lurking at the back of my mind for a long time.”

Durkacz says she knew from the beginning, when she received her diagnosis in 2005, what the disease would do to her, and she began wondering right away what would happen at the end. She knew someone at the time who was dying of emphysema and for whom, she observed, “every breath was a struggle for survival.”

“The first thing I said to my neurologist was, ‘I don’t want to slowly suffocate to death,’” she wrote in her forum post. “This end-stage scenario still haunts me.”

Durkacz still worries about not being able to communicate her fear and anxiety. “My fear of struggling to breathe remains very real,” she says, but notes that it’s comforting to know that medications can help “in coping with the worst of the symptoms of breathing failure.”

To those who are still asking the question, *What happens at the end?*, Durkacz recommends reading the available medical literature and talking to your physician and others with the disease. She also finds the advice and support on online forums from others with ALS “immensely valuable.”

cult may — or may not — determine how the end transpires.

In those who choose to forgo assisted ventilation, by far the most common cause of death is respiratory failure.

Noninvasive ventilation aids in alleviating shortness of breath, Rowin says, but eventually as respiratory muscles weaken, it becomes insufficient to prevent respiratory failure.

Those who use invasive ventilation (delivered through a tracheostomy or opening in the neck) may delay respiratory failure for some time. Occasionally, complications occur with equipment, such as disconnected tubing or mechanical problems, and sometimes the individual may still have a respiratory failure, such as when the lungs become unable to expand as a result of pneumonia or lung collapse.

In many cases, invasive ventilation

provides users with more time and a better quality of life. When the end does come, the experience of those on a vent and those who aren't "should be very similar," says Bromberg.

---

A common phrase heard from family members whose loved ones have passed is, "It was very peaceful at the end."

---

### What can I do to prepare?

Advance planning can alleviate some of the stress involved in facing end-of-life issues.

Shafeeq Ladha, director of the MDA/ALS Center at St. Joseph's Hospital and Medical Center in Phoenix, notes the importance of planning and discussion early in the course of the disease to prevent fear, mistakes and

discomfort in times of crisis.

Ladha advises people with ALS to create a living will with advance directives and to discuss their wishes with whomever will be making medical decisions for them in the event they're unable to do it themselves. "Too often," Ladha says, "physicians and family members are afraid or too uncomfortable to breach this topic, and so when the time for end-of-life decisions comes, no one is prepared."

Be sure to discuss palliative care options with your physician, and contact your local MDA office for assistance with finding hospice care and other support and resources.

"I think the most important message," says Rowin, "is that if the patient has an experienced neurologist and hospice care in place, they do not have to be fearful of terminal physical pain and suffering, because they will be well taken care of." □

Introducing the 2010

## Black Hawk

E D I T I O N  
P o w e r C h a i r

### Elaine's Testimonial



Elaine Ward tested the new Levo C3 and Permobil C-500 VS.

She chose the Redman Black Hawk for its excellent performance and infinitely versatile range of motion.

Test the Redman Black Hawk and you will become a believer, too.

## Power Yoga



Pictured above is a partial sampling of the Black Hawk Power Chair's range of motion. The Chair is driveable in the tilted and standing position with superior stability using a **Patented System** that *floats*.

The Redman Power Chair is highly engineered to perform with precision, comfort, and reliability, standing above all others in the industry. Learn more about the new Black Hawk Power Chair and experience a heightened sense of mobility.

**REDMAN**  **POWER CHAIR**  
Call us or visit our web site today **800-727-6684** [www.RedmanPowerChair.com](http://www.RedmanPowerChair.com)  
DRIVE | TILT | STAND THE NEW CHIEF 107 ZR<sub>3</sub> REDEFINING MOBILITY

## March/April 2010

### Check it out:

Seeking candidates for  
"Anyone's Life Story"

See page 5

**Freedom. Finally!**

Accessible Vans at a price  
**you CAN Afford**

- ★ Dedication to Customer Satisfaction
- ★ Personalized Service and Delivery
- ★ Nationwide Financing Available
- ★ Crash Test Certified
- ★ Best Conversion Warranties
- ★ 24/7 Emergency Assistance
- ★ Generous Referral Program
- ★ Post-Purchase Resale Program
- ★ YES! We Take Trade-ins.

For an additional  
**\$510 OFF**  
just tell us you saw us in MDA/ALS  
**(800) 775-8267**  
[www.amsvans.com](http://www.amsvans.com)

 **ams vans** INC  
*Your Dream of Going Places.  
We Dream of Getting You There.*