

Twin Vent Users

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My sister Sheila and I are identical twins who have been challenged with limb girdle muscular dystrophy (LGMD) since the age of eight. Being diagnosed with muscular dystrophy was no easy task for the doctors.

At age two, I was hospitalized in isolation for two weeks with polio, which only left me with a limp. At age eight, Sheila started limping exactly



Sheila (left) and Nancy

like me. Because we were such closely-knit twins, people thought that the limp was out of sympathy for each other. Our mother became quite concerned and took us to our pediatrician who told her she was just a young mother worrying about nothing.

After observing us climbing aboard the school bus differently than the other children, she decided to take us to Children's Hospital in Boston. It was there that a muscle biopsy confirmed a diagnosis of muscular dystrophy. The prognosis was terrible: we would not walk after our teens nor live out our 20s. But we proved them wrong, walking until we were 25 and living well beyond our 20s. We are now 63.

When we turned 34, Sheila started having trouble breathing but because she had a very stressful job, our mother and I kept telling her it was

nerves. We almost killed her. She suddenly lost a lot of weight, and her voice became very soft. Thus began our love affair with Nicholas Hill, MD, our pulmonary doctor. When Sheila went to see Dr. Hill for the first time, a blood gas test confirmed she was indeed having trouble breathing. After a few visits, Dr Hill told her she needed to start using a ventilator.

My whole family was shocked because we associated a ventilator with death, not life. Oh, how wrong we were! We have lived full and productive, happy lives for 30 years, despite needing to use our ventilators all the time. Now we use a bilevel unit with nasal pillows during the day and a nasal mask at night. When we go out, we use a mouthpiece with the HT50® ventilator that hangs on the back of our power wheelchairs.

We have traveled extensively, mostly when we only needed to use the bilevel at night. Dr. Hill likes to tell the story of Sheila using the pneumo-wrap, a breathing machine that was like a vacuum cleaner. It took two personal care attendants to get her comfortable in it. I started using the bilevel, then new on the market, and decided to take a trip to Disneyworld,

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International Ventilator Users Network's mission is to enhance the lives and independence of home mechanical ventilator users and polio survivors through education, advocacy, research and networking.

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Special thanks ...

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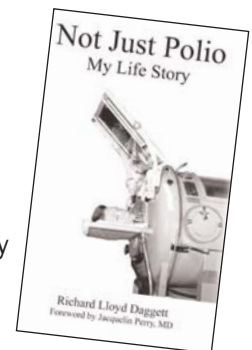
Dr. Nicholas Hill Honored



Nancy Nolan, Nicholas Hill, MD, Sheila Nolan in 2009. Dr. Hill, Chief, Pulmonary, Critical Care & Sleep Medicine, Tufts Medical Center, Boston, received the Margaret Pfrommer Honorary Lecture and Award from the American College of Chest Physicians in 2009. His lecture focused on his relationship with the Nolans (See page 1.) as an example of the partnership between patients and doctors in long-term home care.

Book by Polio Survivor Vent User

Not Just Polio, (www.polioassociation.org/Not_Just_Polio_-_Downey_Patriot.pdf) Richard Daggett's autobiography, is now available through Amazon and Barnes & Noble. Daggett includes many compelling details about his wonderful family and life in Southern California, as well as his ties with Rancho Los Amigos, one of the respiratory centers funded by the March of Dimes in the 1950s for respiratory polio survivors. But as Daggett makes clear, polio is only one area of his life. Proceeds from the sale of the book will go to the Polio Survivors Association, of which Daggett is president, and the Amigos Fund of Rancho Los Amigos National Rehabilitation Center. \$14.95, 148-page paperbound.



New from IVUN

“Breathing and Sleep Problems in Polio Survivors” and **“Tests for Breathing Problems If You Have a Neuromuscular Condition”** are available at www.ventusers.org/ThisWk.html#twJun3. Members without Internet access may call 314-534-0475 for print copies. The first describes problems that survivors may experience, pointing out that they can be more than obstructive sleep apnea (OSA). The second article describes a routine evaluation for respiratory function which is good for the first evaluation and for later periodic checkups. ▲

Frogbreathing: A Forgotten Breathing Aid

Tom Mecke, San Antonio, Texas, tom@mecke.info, <http://toms.page.us>

Tom has Duchenne muscular dystrophy (DMD) and turned 50 years old in January 2010. He has avoided a respiratory crisis, which can often lead to a tracheostomy, by careful monitoring of his respiratory condition and seeing a doctor willing to try non-invasive alternatives. Tom worked for Southwestern Bell until 1997 when he left to start his own company, Mecke & Associates, which designs websites, upgrades PCs and builds custom computer systems (www.meckeai.com). Tom is a DMD Pioneer, a group of individuals with DMD who have surpassed their expected life span (late 20s) by many years, even decades. (<http://dmdpioneers.org>)

I had made it to the age of 22 and was in my senior year at Texas A&M University working on an electrical engineering degree before I had any major respiratory infections. College life was busy and quite tiring, but I had noticed a decrease in my respiratory function. Around Thanksgiving, I became congested and went to the doctor to get some antibiotics, which usually cleared up any infection. However, this one kept hanging on, and finally I was so exhausted that my doctor put me in the hospital over the Christmas holidays.

Intravenous antibiotics and IPPB treatments began. My doctor tried to avoid tracheostomies if he could, and he put me into an iron lung that the hospital (Santa Rosa Hospital in San Antonio) still maintained, even in the early '80s, to give me respiratory support. After the pneumonia cleared up, I went home. However, I was soon back in the hospital, and after that infection cleared, my doctor said I would need at least respiratory support at night, and I should try to find a used iron lung. Miraculously, I found one at the Air Force surplus dealer locally. I modified and used this machine for 13 years to sleep in at night while breathing on my own using frogbreathing for extra support during the day. I was able to complete my engineering degree and to begin my career.

On one visit to the MDA clinic where I was being monitored and which was located at the medical school, several residents and medical students accompanied the doctor as he checked me out. One of the students noticed how I was taking in air so that I could talk

loud enough to be heard and asked, "Who taught you how to frogbreathe?" I asked what he was talking about and was told, "That's what you're doing." Apparently I had taught myself how to do it unconsciously.

So what is this technique with the funny name? (The medical term is glossopharyngeal breathing or GPB.) Basically, it involves using your tongue and pharynx as a sort of air pump. The glottis or vocal cords act as a check valve as you use your tongue to gulp air into your lungs. You repeat the gulping of air until you have a full breath. I used this technique at first to stack enough air to speak loudly, but eventually I needed the technique to speak audibly. (GPB usually doesn't work with a tracheostomy because the air will just leak out of the trach.)

After 13 years using the iron lung at night and GPB by day, my respiratory weakness had progressed to the point that I needed continuous support. I was referred to Joseph Viroslav, MD,

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"Some of the techniques from the polio days which have been basically forgotten can work for DMD now."

—Tom Mecke

From Vent to Diaphragmatic Pacing System: Part II

Bill Miller, Leesburg, Florida, MaxNWM@aol.com

In Part I, I described all the factors I considered before deciding to become a Diaphragm Pacing Stimulation (DPS) System user. www.ventusers.org/edu/valnews/VAL_24-2Apr10p5.pdf



Bill Miller

Blog at
<http://powerwheelchairusers.blogspot.com>

After deciding that I want to become a DPS user, I took the next steps of contacting and meeting the surgeon to discuss my candidacy, obtaining necessary medical evaluations, and gaining insurance approval.

I met with Lawrence Lottenberg, MD, FACS, who performs surgery at Shands Hospital at the University of Florida in Gainesville. He thinks I'm a good candidate since I'm relatively young, healthy and quite motivated. Because I'm on a blood thinner, Dr. Lottenberg said I'll need to be hospitalized for at least two nights: one night preceding surgery to thicken my blood, and one night after surgery to thin my blood back to a "therapeutic" level to prevent blood clots.

I needed to have my heart pacemaker and my phrenic nerve evaluated. My heart pacemaker has a dead battery and will be replaced with a completely new pacemaker. It's a demand pacemaker that only kicks in if my heart rate falls below 60 beats per minute, which almost never happens. Consequently, there was some debate about whether I need the pacemaker, but since there's no interaction between the DPS and the pacemaker, why not have it as backup?

With that decided, I will have the pacemaker replaced by a cardiologist when I have the DPS surgery. The cardiologists said that it may take three days following surgery to get my blood back to therapeutic, and I'll probably need to stay in the hospital.

A successful phrenic nerve test helps secure insurance approval for the DPS surgery. The phrenic nerve is the body's natural way of stimulating the diaphragm for breathing. But

Dr. Lottenberg confirmed that the diaphragm's ability to be stimulated cannot truly be assessed until the surgeon gets inside, and that the phrenic nerve test often yields false negatives. It can yield a positive, which indicates that my diaphragm is capable of being stimulated, but a negative test doesn't eliminate the possibility for success with the DPS.

I had an appointment with a neurologist to test my phrenic nerve, but when he found out I had a heart pacemaker, he was not comfortable doing the test. Even though my pacemaker's battery is dead, he said, I still have leads going to my heart and he was concerned about stimulating my phrenic nerve under those circumstances.

After reporting that to Dr. Lottenberg and Dr. Raymond Onders, who pioneered the DPS, Dr. Onders said the neurologist could try stimulating my phrenic nerve only on my right side to avoid possible interaction with the pacemaker. But that became a moot point when I learned that my insurance company "has now passed protocol for the DPS, thus no prior authorization is needed." That statement was told to Dr. Lottenberg's assistant by the hospital's financial group, and I'm waiting to get written clarification. My understanding is that the surgery will cost more than \$20,000, and I don't want the hospital coming after me if my insurance doesn't pay for some reason.

Another insurance issue was trying to get my insurance company to state in writing that my level of nursing coverage would not be decreased when I become a DPS user. Currently I receive 44 hours per week of nursing coverage and my family covers the remaining 124 hours. For them to cover more would be quite difficult.

I wrote to my insurance company to explain that even as a full-time DPS user, I would still be dependent on a mechanical device to help me breathe, and the rest of my care would essentially remain unchanged. However, the insurance company replied that a “clinical review” would be necessary to determine the level of care I need after I get the DPS.

I wasn't pleased with that reply until I spoke with a Florida friend who has

the same insurance company and has been a DPS user for over five years. He told me that he receives 16 hours per day, 7 days per week, of nursing coverage, for a total of 112 hours. He also knows another DPS user with the same level of care and same insurance. Now I'm not concerned about a clinical review and might even be able to increase my nursing coverage. (For anyone who might have the same concern, Dr. Onders said that no DPS user has had his/her level of insurance coverage decreased, and it shouldn't be an issue for anyone.)

I don't have a firm surgery date yet, but I am hopeful it will be late June or early July. Stay tuned for Part III about the surgery. ▲

For more information about the DPS System, go to www.synapsebiomedical.com

Twin Vent Users

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leaving Sheila home. I had been trying to convince her to try the bilevel but she always found change difficult. After I went to Florida without her, she finally tried and succeeded with the bilevel. Since then we have both gone to Disneyworld together.

You would think living with a ventilator would be difficult, but it just becomes a way of life. You can choose to think of it as a burden or you can just enjoy each day as it comes. We are not saying that living on life support systems has not been challenging, but we have wonderful caregivers who like to have as much fun as we do.

Our summers are our time for fun! Last summer we traveled to Maine, Rhode Island, and many places in the

Boston area in our van. When traveling in the van, we wear our neck braces and chest straps. Our necks are very weak and floppy, so finding these neck braces (See Headmaster Cervical Collar™ details on p. 6.) was a great blessing and changed our lives.

We had a sine-wave inverter installed in our van so we could use our CoughAssist® when traveling as well as plugging in the bilevels anytime.

When we arrive at our destination, we turn on the HT50® ventilators on the backs of our wheelchairs and use them as we sightsee. People have told us we are like the Energizer bunny; we just keep on going! ▲

From Around the Network

Judith R. Fischer, IVUN Information Specialist, info@ventusers.org



iVent™ 101 Expert

New Equipment and Masks

iVent™ 101 Expert from GE Healthcare offers pressure-regulated volume control (PRVC) as well as standard ventilation modes. It is small and weighs 13.4 lbs., with an internal battery that lasts up to four hours and a color touch screen. Two additional home care models include the Signature for pressure support & SIMV (VC & PC) and the Performance for control ventilation (VC & PC). Available for use by children or adults worldwide. www.gehealthcare.com

Verone and **Verone+** nasal masks from Air Liquide Medical Systems, Inc., can be used with leak ventilation. **Verone Blue** and **Verone Blue+** can be used with valve ventilation. Verone+ and Verone Blue+ offer optimal adjustment for the forehead support pad. User's field of vision is unhindered. Available outside the USA. www.airliquidemedicalsystems.com

NP15 nasal pillow mask from Weinmann has two ball-and-socket joints that enable more freedom of movement without affecting the fit. Exhaled air is directed away from the face. Made of silicone, the pillows come in three sizes. Available outside the USA. www.weinmann.de



NP15 nasal pillow mask

Neck Support

Headmaster Cervical Collar™ from Symmetric Designs of Canada can be easily formed by hand to fit the chin and has a washable fabric cover. Worldwide distributors. www.symmetric-designs.com. See photo on p. 1 of the collars, as worn by two satisfied consumers.

Muscular Dystrophy

The diagnosis and management of Duchenne muscular dystrophy is an international consensus document about best practice in care for Duchenne muscular dystrophy (DMD). Published in *Lancet Neurology* in January and February 2010, it is the product of an extensive review process by 84 international experts (including pulmonologist David Birnkrant, MD) in DMD diagnosis and care.

Part 1: Diagnosis, and pharmacological and psychosocial management
<http://linkinghub.elsevier.com/retrieve/pii/S1474442209702716>
Lancet Neurol. 2010 Jan; 9(1):77-93

Part 2: Implementation of multidisciplinary care
<http://linkinghub.elsevier.com/retrieve/pii/S1474442209702728>
Lancet Neurol. 2010 Feb; 9(2):177-189.

Grants and Scholarships Available

CMMS Deshae Lott Ministries, Inc., is offering scholarships to individuals with severe disability who want to pursue post-graduate education. The minimum amount is \$500, but may be as high as \$1,000. Quality of life grants to individuals who are disabled and need financial assistance with in-home care are also available. Lott is a ventilator user and member of the Christian Metaphysicians and Mystics Society. Deadline for the scholarship application for each academic year is July 1. More information and applications are online: visit www.deshae.net. ▲

Frogbreathing: A Forgotten Breathing Aid

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at the Dallas Rehabilitation Institute for evaluation. He switched me to noninvasive ventilation using a PLV®-100 volume ventilator with a custom nasal mask while in bed and a mouthpiece positioned near my mouth when I was in my wheelchair. I have been using this arrangement for 15 years with good success.

GPB still comes in handy if I lose access to my mouthpiece or a hose comes off of my vent, while showering, and for use with an Ambu bag. On a few occasions, GPB may have saved my life when a ventilator tube popped off during the night, and I was able to stack up enough air to call for help.

I have had a somewhat different experience than most people with DMD. Some of the techniques from the polio days that have been essentially forgotten can work for DMD now. ▲

Reminder: All PLV® ventilators were discontinued on 12/31/2009. "All reasonable efforts" will be made by Philips Respironics to continue renting, servicing, supplying replacement parts, and technical support through 12/31/2014. www.respironics.com

MORE on www.ventusers.org

Video presentation links ...

Frogbreathing (glossopharyngeal breathing)

www.garymcperson.com/?#FrogBreathing

Lung Volume Recruitment (LVG) and glossopharyngeal breathing (GPB)

www.irrd.ca/education/slide.asp?RefName=e2r4&slideid=60

Join IVUN and receive Ventilator-Assisted Living, IVUN's bi-monthly newsletter

(February, April, June, August, October, December). The eight-page newsletter will be sent electronically. (IVUN Members without email access may request print copies by contacting IVUN). Members will also receive an electronic *IVUN Membership Memo* in alternate months. To become a Member, complete this form. *Memberships are 100 percent tax-deductible.*

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BiPAP AVAPS

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