Now called
Ventilator-Assisted Living

IVUN News

International Ventilator Users Network
Saint Louis, Missouri USA

Spring 2003 ■ Vol. 17, No. 1

A Classic Case of Hypoventilation

Bryna Golden (brynagolden@myfastmail.com)

Although I would not be diagnosed with a neuromuscular disease until I was 21 years old, I grew up with a number of ailments and health problems. In my teens, I was diagnosed with restrictive lung disease and vital capacity of about 30% that was attributed to scoliosis. Yet, even after fully recovering from a spinal fusion to correct the scoliosis, my shortness of breath continued and walking from class to class carrying my books caused inordinate fatigue.

By my last year in high school, I was having a terrible time waking up every day. I felt hung over in the mornings, my head throbbing and feeling like a bowling ball, my stomach so upset and jittery that I was unable to eat until lunchtime. I occasionally would wake up from my sleep choking and consequently developed an aversion to going to bed.

I had a very hard time getting myself to school, and some days I did not make it. My doctors' only explanations were that there is no illness that makes a person sick in the morning. They tried to convince my parents that my symptoms were the result of my being a nervous child and suggested that I just didn't want to go to school. Fortunately for me, my parents believed me when I insisted that there was something physically wrong, and it was their empathy, support, and belief in me that helped me through an extremely difficult and painful time.

After high school, I attended a few semesters of college, but walking around the campus exhausted me so much that, despite my enthusiasm for my studies and schoolwork, I had no energy left for learning and was often too ill and tired to concentrate. I dropped out of college and found a market research job that allowed me to work afternoons and evenings (the times of day I felt best). I also began playing keyboard in a rock band.

By February 1985, I was 21, living on my own with roommates, working fulltime, playing gigs, recording with my band ... and feeling physically awful. The strangling episodes were happening several times a night, and I was terrified of going to sleep. I often had the sensation of floating above myself while watching as I strangled and choked. I would awaken shaking and feeling like the room was spinning. I actually wondered if I really was crazy, since my doctor (supposedly the best in his field) was not concerned that there was anything wrong with me.

Finally, after suffering for weeks with a cough that would not go away, having choking spells every time I fell asleep, and experiencing frequent incontinence while sleeping – a new problem I found terribly embarrassing and perplexing – I felt I could not go on. I insisted to my doctor that something was really wrong with me, but his response was to yell at me and tell me to stop complaining because there was nothing wrong with me.

One week later, I ended up in the ER in respiratory failure with bronchitis, strep throat, and mononucleosis. Later in the ICU, it was recognized that I stopped breathing in my sleep, my oxygen saturation (SaO_2) levels were dangerously low, and my carbon dioxide (CO_2) levels were dangerously high. I had been experiencing the classic signs and symptoms of hypoventilation (underventilation). *(See sidebar on page 6.)*

My new doctors suspected that I had a neuromuscular disease that caused the respiratory weakness that in turn caused the pulmonary problems. When I related all my symptoms and how long they had been going on, they told me that I was lucky to be alive. Rather than feeling depressed at the discovery of these new health problems, I felt great relief that finally someone acknowledged an actual physical cause for what I had been going through all those years. It was not just all in my head!

CONTINUED ON PAGE 6

Which Ventilator? Who Decides?

Ventilator users often have limited access to a trial of different types of ventilators and are at a disadvantage in finding the one which is best suited to their needs, the most comfortable to use, and the most affordable by their insurance carriers. Access to interface options and the opportunity to try a range of interfaces is also limited.

Recently an Italian study of European ventilators, published in *Chest,* the journal of the American College of Chest Physicians, focused on the problem. Entitled "Comparison of five bilevel pressure ventilators in patients with chronic ventilatory failure: A physiologic study," the study compared patient-ventilator action and comfort of the Helia (Saime, S.A.), O'Nyx (Puritan Bennett), Harmony (Respironics, Inc.), RespiCare CV (Dräger HomeCare), and PV102 (Breas Medical AB).

The article included two key statements:

"In our study, despite the fact that the ventilators were set at the patients' comfort level, the sensation reported during the trial was different with each ventilator studied, indicating that each patient experienced different sensations with each individual ventilator. This may be relevant in light of the fact that NPPV [noninvasive positive pressure ventilation] needs the patient's cooperation, which cannot be obtained under conditions of discomfort."

"As no significant relationship was found between comfort and pulmonary mechanics, we can argue that there is no objective method with which to determine which ventilator will be tolerated by an individual patient. The results of this study indicate the need for trials with different ventilators before prescribing NPPV in order to assess the best compliance for the individual patient."

The authors concluded "... the choice of the ventilator for home NPPV therapy should be made after a comparison of different ventilators and should be tailored to the individual patient."

"In essence, this [study] is another plea for patient-centered care and participation by the patient in relevant care decisions. If only we practiced in a system where that were even remotely possible ..."

In an accompanying editorial Drs. Graziano Carlon and Arthur Combs commented, "Ironically, the real issues pointed out by the current study have nothing to do with the NPPV device characteristics, per se. Rather they are as follows:

1. How many clinicians are aware of the full range of marketed devices and interfaces available or their relevant characteristics, so that they may prescribe the best possible NPPV for a given patient?
2. How many communities have medical equipment distribution and reimbursement systems that can support such [appropriate] individualization of care?
3. How many patients have the opportunity to try a spectrum of devices, interfaces, and modes, select those most comfortable, then further test their selections over time at home under varying conditions?
4. What are the drivers for innovation and product development among manufacturers? Is there a search for engineering solutions that do correlate with patient comfort?
5. Does measurement of comfort lend itself to a Euclidean quantitative relationship, of the types: if $a > b$ and $b > c$, then $a > c$? That is, if a patient says that (s)he prefers ventilator b to ventilator a, and ventilator c to ventilator b, can we safely assume that ventilator c will be preferred to ventilator a in a direct comparison? The order of comparison may be a major confounding factor, as is often the case in market testing of consumer products. This possibility would require not only that all ventilators be tested, but also that each of them be tested against each other, an even more daunting task."

Carlon and Combs conclude, "In essence, this [study] is another plea for patient-centered care and participation by the patient in relevant care decisions. If only we practiced in a system where that were even remotely possible ..."

Joseph Lewarski, BS, RRT, President, Hytech Homecare, Mentor, Ohio, and Home Care Section Chair for the American Association for Respiratory Care (Joe.Lewarski@hytree-hytech.com), agrees with the editorial. "Centers of excellence, which exist in few markets, are ideal places to test and trial patients and devices, including the ventilators and the interfaces.

"The cold reality is that this is not practical or financially possible in most cases. Most clinicians

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Home or Institution?

A man with ALS uses 24-hour tracheostomy positive pressure ventilation (since 1999) and lives at home. His wife works fulltime to support them and to retain medical insurance, which provides 16 hours of licensed visiting nurse (LVN) in-home care every day. With the current nursing shortage, the home health agency cannot always provide LVNs when they are needed, and the couple is often advised that a skilled nursing facility would be a better place for his care.

A young woman with muscular dystrophy, age 31, lives in a New York state nursing home. Her request for 16 hours of daily nursing care to enable her to move in with her sister was denied with the explanation, "... the client's health and safety cannot be maintained in the community."

Nick Dupree, a ventilator user due to Duchenne muscular dystrophy, approaches his 21st birthday knowing that he will no longer be eligible for Medicaid services that would allow him to remain in his Alabama home and in his community. The alternative: living in a nursing home in another state.

All of these individuals should be able to live independently at home within their communities with home services and attendant care, which is more cost-effective than institutional care. But why is living at home still such a problem? A law, a Supreme Court decision, and proposed legislation are supposed to make it happen.

The Americans with Disabilities Act (ADA) is a landmark civil rights statute designed to halt all practices that segregate persons with disabilities and those that treat them differently. In Olmstead v. L.C., the 1999 Supreme Court decision that stated "... unjustified institutional isolation of persons with disabilities is a form of discrimination ..." upheld the ADA's integration mandate. Olmstead requires states to end unnecessary institutionalization of individuals with chronic conditions and disabilities who can remain at home with community support services in "... the most integrated setting appropriate to their needs."

This means each state must use its Medicaid and other funds to support people with disabilities to live in the community rather than in nursing homes or other institutions. To do this effectively, a state must make certain that "the money follows the person." (www.accessiblesociety.org/topics/ada/olmsteadoverview.htm)

Nick Dupree challenged Medicaid ... and won. (www.nickscrusade.com) An intense media campaign and lobbying by AIMMM (Advancing Independence: Modernizing Medicare and Medicaid) focused attention on Dupree. The Department of Health and Human Services approved a Medicaid waiver enabling Alabama to continue support at home for people with disabilities who might otherwise lose their home care services simply because they turn 21.

Although every state has chosen to provide certain services under home- and community-based waivers, they are unevenly distributed, have long waiting lists, and reach just a small percentage of eligible individuals.

MiCASSA (Medicaid Community Attendant Services and Supports Act) is continually reintroduced into Congress, but never passes. It is long-awaited legislation to bring the Medicaid system into accord with the Olmstead decision by establishing a national program of community-based attendant services and supports for people with disabilities. This bill would allow the Medicaid dollars to follow the person, and allow eligible individuals, or their representatives, to choose where they would receive services and supports. Any individual who is entitled to nursing home or other institutional services would have the choice where and how these services are provided. (www.adapt.org) ■

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Humidification

Louie Boitano, MS, RRT, Northwest Assistive Breathing Center, Pulmonary Clinic, University of Washington, Seattle (boitano@u.washington.edu)

Our lungs are the only internal organs in our bodies that are directly exposed to the outside environment through the air we breathe. Our nasal and sinus passages condition the air we breathe before it reaches the lungs by warming and humidifying the air, keeping the lungs both warm and moist.

Mucus captures the impurities (microscopic particles, bacteria, fungal spores, etc.) in the air we breathe. The mucus and captured impurities are then removed by tiny hair-like structures called cilia that line the airways and act as a conveyor (think mucociliary escalator) to clear the lungs. Breathing dry, cool air for long periods of time can cause the dehydration of mucus that lines the airways of the lungs. Dehydrated, thickened mucus can either slow or stop mucociliary clearance. Thickened mucus then becomes a potential source for lung congestion and infection.

When you have a tracheostomy and breathe through a tracheostomy tube, your natural humidification system is bypassed. Providing adequate humidity to the air you breathe becomes important in maintaining your lung health.

There are different levels of ventilator humidification.

Artifical nose. Use of an artificial nose also called a heat-moisture exchanger, works by holding some of the moisture that is normally lost on exhalation. This consists of a paper wick network enclosed in a plastic housing with open ends that can be connected to a home ventilator breathing circuit between the tracheostomy tube and the exhalation valve. The paper wick captures moisture on exhalation and provides moisture with inhalation. Hudson RCI (www.hudsonrci.com), Portex (www.portexusa.com), and Boston Medical (www.bosmed.com) are among several companies that manufacture artificial noses.

Pass-over humidification. A water chamber, also called a pass-over humidifier, is in the circuit without heat. This provides a subtle amount of moisture. An example is the LX Pass-over Humidifier (Respironics Inc., www.respironics.com). Pass-over humidifiers are primarily used for non-invasive mask ventilation or CPAP therapy (the ConchaTherm 2000 from Hudson RCI), but do not provide enough moisture for ventilation through a tracheostomy.

Heated water chamber with temperature adjustable up to 39°C. Water droplets are likely to appear in the ventilator circuit but are not dangerous if the circuit contains a water trap and the circuit is monitored on a regular basis. Examples are the HumidAire™ (ResMed, www.resmed.com) and the MR410 humidification system (Fisher & Paykel Healthcare Corporation, www.fphcare.com).

Heated water chamber with heated wire in hose at the same temperature. The heated wire breathing circuit supports both uniform heat and humidity (beneficial for cooler climates). Water droplet buildup in the hose is less likely to be a problem. An example is the HC500

Fisher & Paykel's HC500

Treating Bronchitis with H_2O

Jerry Daniel (JerryPD2000@aol.com)

In 2000 I marked 45 years of successful positive pressure ventilation by tracheostomy, but there was a dark cloud hanging over my head. My chronic bronchitis was getting worse, and prednisone and antibiotic Cipro treatments were becoming more frequent but less effective. My frogbreathing-powered cough was not working, and my bronchial tree seemed tough and dry. I had frequent hospitalizations and even considered moving from the state of Washington back to Southern California for a more favorable climate.

My pulmonologist and respiratory therapists in the hospital tried to make me comfortable with the Cascade humidifier, but I resisted it. Then they brought in the Fisher & Paykel HC500 heated hose humidifier. I was more receptive because it looked compatible with my LP10 ventilator. I have now used the HC500 all night and intermittently during the day for three years. The humidifier is always inline with the LP10. The secretions are under control; I switch the heating elements off and on as needed. Tracheal suctioning did not seem to work before, but now it works well. I have not needed prednisone treatments, antibiotics, or to be hospitalized, and the humidifier is easy to maintain at home.

humidification System (Fisher & Paykel). Not all insurance carriers will reimburse for this type of humidification system.

A heated humidifier in the ventilator breathing circuit warms the air and provides enough moisture to prevent mucus dehydration.

A common problem encountered with heated humidification is condensation or "rain out" – when warmed humidified air cools as it travels through the breathing circuit surrounded by cooler air.

This condensed moisture must be removed before it either restricts the flow of air through the breathing circuit or causes choking by entering the tracheostomy tube. A water trap can be placed at the lowest point in the breathing circuit to collect the condensation.

The heated humidifier must be set at a temperature high enough (30-34°C) to provide adequate humidity after a portion of the water vapor is lost to condensation in the breathing circuit. The circuit can be insulated to decrease the amount of water lost to condensation, but this can be bulky and impractical.

Heated wire breathing circuits generally require significantly more support because they are non-disposable and must be cleaned regularly. The more common disposable breathing circuits with water traps are more convenient to replace, but require regular monitoring for water accumulation and emptying of the water trap in the breathing circuit. ■

Calendar 2003

APRIL 30-MAY 1	**The ALS Association's Annual Leadership Development/ALS Clinical Conference.** Renaissance Hotel, Washington, DC. Contact ALSA, 818-880-9007, alsaconference@@alsa-national.org.
JUNE 20-22	**Families of SMA Conference.** Hyatt Regency-Capitol Hill, Washington, DC. Contact Families of SMA, 800-886-1762, www.fsma.org.
JUNE 26-28	**CCHS Family Network Conference.** Caribe Royale Resort Suites, Orlando, Florida. Contact Mary Vanderlaan, 607-432-8872, www.cchsnetwork.org.
JULY 10-13	**Parent Project Muscular Dystrophy Conference.** Omni Netherland Hilton, Cincinnati, Ohio. Contact Parent Project, 800-714-KIDS, www.parentprojectmd.com.

In Memoriam

IVUN expresses appreciation for contributions to its operating fund in memory of ...

Wayne Abney
Bob Horn, PhD

IVUN expresses appreciation for contributions to The Research Fund in memory of ...

Doug Martin, PhD
Don Willfong

Contributions (tax-deductible) to either fund in honor or in memory of friends, loved ones, and colleagues may be mailed to IVUN, 4207 Lindell Boulevard, #110, Saint Louis, Missouri 63108 USA, and are gratefully received to continue the vital work of the International Ventilator Users Network.

The Research Fund

Thanks to ventilator user Morton Freilicher and a $10,000 gift from The Edouard Foundation, the corpus of The Research Fund is $285,000. Grant awards are provided using the income earned from the principal.

The Fund supports researchers through either The Thomas Wallace Rogers Memorial Respiratory Research Grant: to study the cause(s) and treatment of neuromuscular respiratory insufficiency and the effects of long-term home mechanical ventilation, OR The Post-Poliomyelitis Research Grant: to study the cause(s), treatment, and management of the late effects of polio.

The first grant awarded $20,000 in 2001 to the study "Ventilator Users' Perspective on the Important Elements of Health-Related Quality of Life," available online at www.post-polio.org/QofLFINALREPORT-Sept2002.pdf.

The second grant awarded $25,000 in 2003 to a team of researchers from the University of Michigan for a study comparing the differences of aging between women and men with a neuromuscular disease (polio).

A "Call for Proposals" for a third grant to awarded for 2005 will be issued in early 2004. Check IVUN's website (www.post-polio.org/grf-call.html) or future issues of *IVUN News*. To receive an announcement, send your information to IVUN, 4207 Lindell Boulevard, #110, Saint Louis, Missouri 63108 USA or e-mail ventinfo@post-polio.org. ■

After a week in the hospital, and with the bronchitis almost gone, I was discharged with an O_2 compressor and told to use it whenever I slept. I felt like I was in a haze the first couple of days at home, and by the third day I was almost in a coma. Rushed back to the ER, I was admitted in respiratory failure again. This time the doctors considered a tracheotomy, but then decided to transfer me to the Pulmonary ICU at the University of California, Irvine Medical Center. I remained there for a month while many tests were conducted, including an inconclusive muscle biopsy. I was finally discharged with a diagnosis of unidentified neuromuscular disease, sleep apnea, restrictive lung disease, and chronic respiratory failure.

One month later, after trying several negative pressure devices, I began using an Emerson rocking bed and O_2 through a nasal cannula whenever I slept. I used the rocking bed from 1985 to 1989 and while it helped to a certain degree, I felt I should have been breathing and sleeping better. I was still having strangling dreams, headaches, and confusion. Also, the large cumbersome nature of the rocking bed meant I was unable to spend a night away from home. This situation made it impossible for me to travel with the band for out-of-town performances.

My pulmonary doctor at UCI attempted to switch me over to a portable ventilator, but was unable to find a mask that I was able to tolerate. I have a very thin face and prominent nose, so the masks would only make a good seal if they were so unbearably tight that they left painful pressure sores on the bridge of my nose. In 1989, he made arrangements for me to see Ahmet Baydur, MD, at Rancho Los Amigos Medical Center and to spend a night there trying different types of noninvasive interfaces. The stay was very successful, and soon after I had my own PLV®-100 volume ventilator with an ADAM circuit and nasal pillows. (I now use nasal pillows with Breeze™ SleepGear™ for more comfort.)

The ventilator changed my life. Once I became accustomed to using it, it was a huge relief to go to sleep and let the machine

What is Hypoventilation?

Robert A. Lebby, MD (rlebby@barlow2000.org)

The work of inspiring air, or breathing in, is performed primarily by the diaphragm although other muscles play a smaller role. With many neuromuscular diseases, these muscles become weak and less able to expand lungs and draw air in. Other factors making it harder to fully expand the lungs in some people, such as Bryna, include lung and chest wall stiffness/inelasticity and scoliosis (spinal curvature).

Smaller breaths can result in less overall airflow or hypoventilation. If adequate airflow does not occur through the lungs, then carbon dioxide (CO_2) is not effectively eliminated and may build up in the blood. Blood oxygen levels (O_2) may also decrease due to the hypoventilation, high CO_2 levels, or other reasons.

The symptoms of hypoventilation may be subtle at first, especially in people with neuromuscular disease whose problems evolve very slowly allowing the body to adapt to the gradual changes. The earliest problems usually occur during sleep because the control of breathing changes with different sleep stages and because sleeping in the supine position may worsen breathing.

The signs and symptoms of nocturnal (sleep-related) hypoventilation include many that Bryna describes in her article:

- sleep initiation and sleep maintenance insomnia
- anxiety about going to sleep
- restless/fragmented sleep with frequent awakenings
- shallow breathing or pauses in breathing
- awakening from sleep with choking sensation
- nightmares, night sweats, bedwetting, or need to urinate frequently
- night-time or morning headaches
- excessive daytime sleepiness and need to nap during the day
- worsening mental status, impaired memory, concentration, cognition
- inability to lie flat during sleep/need to sleep sitting up (othopnea).

Other symptoms of hypoventilation include:

- shortness of breath, breathlessness with minimal activity
- fatigue or exhaustion from normal activities
- claustrophobia and/or feeling that the air in the room is somehow bad
- anxiety
- difficulty in speaking for more than a short time
- quiet speech with fewer words per breath
- inability to lie flat (even while awake) due to shortness of breath.

Anyone experiencing a combination of these symptoms deserves a respiratory evaluation, preferably by a pulmonary physician experienced in neuromuscular diseases, sleep, and hypoventilation. Together, the physician and the individual can formulate a treatment plan for assisted ventilation.

breathe for me. I was able to wake up feeling rested and without headaches, and I was able to travel with the psychedelic rock band I formed, Babylonian Tiles (www.babylonian-tiles.com). I am the lead singer, keyboardist, and songwriter. Our band recorded three internationally released CDs, and we have had five US tours, on the road for more than a month at a time. Touring would not be possible for me without the ventilator.

An extensive surgical cervical fusion in 1995 stabilized my very weak neck. I remained intubated about six hours after surgery but once fully awake, I was extubated and my pulmonologist switched me over to my PLV®-100, which I used until I could sit up and breathe on my own.

Although my unlabeled neuromuscular disease has progressed to the point that I am now only able to walk a few steps with assistance and must use a wheelchair whenever I leave the house, I still only use the ventilator when sleeping or lying down.

Currently, I am experiencing problems that I think are related to my night-time ventilation: daytime headaches and fatigue (on an almost daily basis), and frequent confusion upon awakening. My respiratory therapist suggested I might benefit from a ventilator such as the Newport HT50®, which provides both volume and pressure support, but it may be difficult to obtain. My current physician, Robert Lebby, MD, Barlow Respiratory Hospital, is ready to tackle and resolve these latest problems so I can again awake from a good night's sleep feeling refreshed and ready to seize the day. ■

ALS Sanctuary. Developed as a prototype for the inpatient care of people with ALS, the MDA/ALS Center of Hope Sanctuary at Hahnemann University Hospital in Philadelphia serves as a model for other medical centers seeking to provide compassionate and high tech care for people with ALS.

For people with ALS admitted to Hahnemann, the sanctuary is a suite-like hospital room with all the comforts of home, space for family members to stay with the individual, and state-of-the-art assistive devices.

The sanctuary is a joint project of The Center of Hope Foundation in collaboration with the Muscular Dystrophy Association (MDA), Hahnemann University Hospital/Tenet, and the MDA/ALS Center of Hope at Drexel University. For more information, contact Megan Sandora, ALS Hope Foundation (www.alshopefoundation.org).

When a Loved One Has ALS: A Caregiver's Guide. 2nd edition. 2002. The 60-page guide includes caring for the person with ALS, caring for the caregiver, end-of-life issues, and getting help, plus an extensive listing of resources and information about the programs in MDA's ALS Division (www.mdausa.org/publications/alscare). One print copy is free to each individual with ALS registered with MDA. For others, copies can be ordered online for $15 plus shipping and handling (www.mdausa.org/publications/puborder.html).

Kaleidoscope: Exploring the Experience of Disability Through Literature and the Fine Arts, a semiannual publication, seeks submissions for its next issue. The theme: "Perspectives on Aging: I Am Still Learning." Deadline is August 1, 2003 for publication January 15, 2004. To request guidelines and/or send submissions, contact Gail Willmott, United Disability Services (330-762-9755, www.udsakron.org).

Non-Invasive Respiratory Support: A Practical Guide.
2nd edition. 2001. Edited by Anita K. Simonds, MD, FRCP, Consultant in Respiratory Medicine, Royal Brompton and Harefield NHS Trust, London. 320 pages, references, illustrations. (0-340-76259-4, paperbound, £29.99 plus shipping, Hodder Arnold Publishers: www.arnoldpublishers.com).

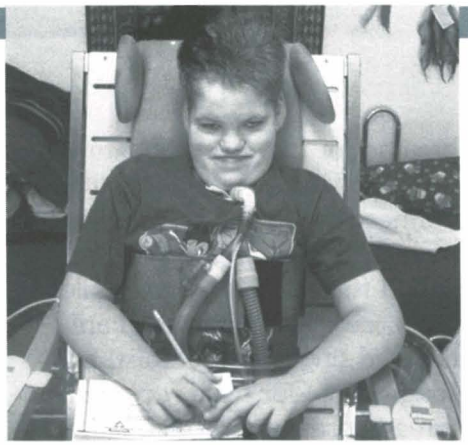
The second edition of this highly practical and informative handbook describes the indications and outcome for noninvasive ventilation (NIV) in acute and chronic ventilatory failure in a wide range of respiratory disorders. It includes how to select patients, choose equipment, and initiate therapy.

Available in the US through Oxford University Press (www.oup-usa.org/catalogs/general/subject/Medicine.html) for $49.50 plus shipping.

DMD Pioneers. Jeff McAllister (Jeff@DMDpioneers.org), who uses noninvasive ventilation due to Duchenne muscular dystrophy, started an email list especially for adults (16 years and older) with DMD. Go to http://groups.yahoo.com/group/dmdpioneers. Jeff also edits an online noninvasive ventilation newsletter (www.JeffSpace.net/NIVnews). ■

Luke's Progress

Mary Garrett (mary.rick@verizon.net)



Our son Luke has Pompe's disease, a glycogen storage disease, in which the acid maltase enzyme necessary to break down glycogen and convert it to fuel is lacking. The glycogen is stored in the lysosomes of the cells, causing muscle damage and death on the cellular level. Luke's respiratory and skeletal muscles are affected. His profile appeared in IVUN News, *Fall, 1996.*

With Luke's discharge from the PICU at Albany Medical Center in Albany, New York, in May 1995, my husband Rick and I believed that there would be a viable treatment for our son's disease within six months to a year. We thought we might be able to care for him at home for as long as two years.

Now, almost eight years later, we are still surviving, learning, struggling, and glad that we decided to bring Luke home. We have had many ups and downs, yet are happy that Luke has managed to avoid hospitalization since his initial discharge. We were told that Luke would probably die before age 2; he will be 10 years old in July.

Luke left the hospital with an LP10 and a CPAP unit to give him continuous flow. After meeting with John Bach, MD, in 1997, we changed his ventilator settings and eliminated the CPAP unit, enabling Luke to be more "portable." Two years ago, Luke obtained the LTV™ 950 ventilator (volume control) and that made him yet more "portable." With a recent growth spurt, Luke needed his ventilator settings adjusted. Luke is very helpful in managing his own care and works the controls on a manual CoughAssist™.

Luke has a busy life. He is homeschooled for 2½ hours each weekday morning and is hooked up via computer to his regular fourth-grade classroom. (He is an A student.) He uses a Passy-Muir speaking valve and vocalizes very well.

It takes two people to get Luke into a rigid body jacket to help maintain his posture and minimize his scoliosis. He is strapped onto a standing frame for 1-1½ hours each day and receives nebulizer treatments, chest percussion, and nutrition through gastrostomy tube feeds every four hours. Daily physical and speech therapies round out Luke's schedule, but I think that it is precisely this schedule that has kept him well for so long.

Luke's siblings, now 16, 14, and 12, are loving, patient, and tolerant companions and mentors and bring the world to him.

So, we live from day to day, fight our battles when we have to, enjoy our family, and hang onto our hope that Luke will soon receive enzyme replacement therapy that shows promise in halting the progression and maybe even reversing his disease. ■

2003 Camps for Ventilator-Assisted Children

March 29-April 4. VACC Camp. Contact Bela Florentin, VACC Camp, Miami Chldren's Hospital (305-662-8222, www.vacccamp.com).

June 1-6. Trail's Edge Camp. Mayville, Michigan. Contact Mary Dekeon, RRT, C.S. Mott Children's Hospital (734-763-2420, mdekeon@med.umich.edu, www.umich.edu/~tecamp).

June 1-7. Camp Pelican. Lions Camp, Leesville, Louisiana. Contact Cathy Allain (985-764-0343, cathyallain@cox.net).

June 8-13. Fresh Air Camp. Hiram House Camp, Moreland Hills, Ohio. Contact Kathy Whitford, CNP, Cleveland Clinic Foundation (216-721-7159, whitfok@ccf.org, www.freshaircamp.org).

June 21-27. CHAMP Camp and CHAMP Camp Adolescent Retreat. Recreation Unlimited, Ashley, Ohio. Contact Nancy McCurdy (317-415-5530, nmccurdy@champcamp.org, www.champcamp.org).

June 21-28. Pennsylvania Vent Camp. Camp Victory, Millville, Pennsylvania. Contact Debra Randler (717-531-5337, drandler@psu.edu, www.collmed.psu.edu/pedsvent).

June 29–July 4. Camp Inspiration. Rocky Mountain Village, Empire, Colorado. Contact Monte Leidholm, RRT, The Children's Hospital (303-837-2502, leidholm.monte@tchden.org).

August 28-31. SKIP Camp. Seashore Methodist Assembly, Biloxi, Mississippi. Contact Judy Abney, SKIP of Louisiana (985-649-0882).

Reimbursement for Ventilatory Equipment: How It Works

The Centers for Medicare and Medicaid (CMS), formerly known as the Health Care Financing Administration (HCFA), contracts with four regional insurance carriers to process claims submitted under Medicare Part B. These claims are for durable medical prostheses, orthoses, and supplies (DMEPOS). Each of the carriers, known as a Durable Medical Equipment Regional Carrier or DMERC, is headed by a medical director.

CMS assigns the DMERCs the task of developing Local Medical Review Policies (LMRP) for the purpose of processing and reviewing Medicare claims for DMEPOS.

The first step is to obtain a HCPCS code (HCFA Common Procedure Coding System) for any DMEPOS item. Obtaining a code is important because it allows health care providers to bill Medicare. Once Medicare approves a code for reimbursement, Medicaid and third party payers usually adopt the same reimbursement policy.

However, some codes are not approved for Medicare reimbursement, such as HCPCS code E0241 for a bathtub wall rail. Medicare does not reimburse for this item, but Medicaid or other third party payers may reimburse for it.

Next, the DMERCs develop a draft LMRP that details the terms and conditions for reimbursement of the equipment. The draft LMRP is required to be submitted for public comments before the final policy is released. The DMERCs notify interested parties through announcements in the *Federal Register* (www.access.gpo.gov/su_docs/aces/aces140.html) and on the CMS website (http://cms.hhs.gov/providerupdate).

Two recent examples of the process:

The **CoughAssist**™ (J.H. Emerson Co.) received HCPCS code E0482 in January 2002. The DMERC directors released a draft LMRP and asked for public comments to be submitted by late October 2002. IVUN activated its network to solicit letters in support of the CoughAssist from pulmonologists, respiratory therapists, nurses, and ventilator users who need the device to mobilize and remove secretions and prevent pneumonias. No final reimbursement policy has been issued.

The **LTV® 950** (Pulmonetic Systems, Inc.) received HCPCS code E0454 for in January 2003. No draft LMRP has been released for public comment.

The process is bureaucratically long, and with proposed reforms in Medicare and Medicaid, there is no predicting the future for HCPCS codes or LMRPs for ventilatory equipment and accessories. IVUN will continue to stay abreast of reimbursement status of ventilatory equipment and alert ventilator users and respiratory health professionals when their support is needed. ■

DMERC Websites

Region A (Northeast)
www.umd.nycpic.com and
www.tricenturion.com

Region B
(Mid-Atlantic and Midwest)
www.adminastar.com

Region C (South)
www.palmettogba.com

Region D (West)
www.cignamedicare.com

Phrenic Nerve Pacing

Christopher Reeve, a ventilator user due to a horseback riding accident in 1995, recently underwent experimental surgery by a new procedure (*IVUN News*, Summer 2001, Vol. 15, No. 2), developed at Case Western Reserve University and MetroHealth Medical Center. The procedure involves implanting phrenic nerve pacers using laparascopic (minimally invasive) surgery, instead of a thoracotomy, a major chest operation that is riskier and more costly.

In phrenic nerve pacing, electrodes implanted near the phrenic nerve connect to a control box worn outside the body to electrically stimulate the diaphragm, the primary muscle needed for breathing, and control breathing.

The main advantage of phrenic nerve pacing is the elimination of the need for mechanical ventilation. Other advantages include improved vocalization and sense of smell, although a tracheostomy is usually still necessary because of possible upper airway collapse during sleep and sudden operational failure.

The system works best in people with high spinal cord injuries who have intact phrenic nerves and normal chest walls. It is not effective for people with neuromuscular disease such as post-polio syndrome because of chest wall abnormalities and because tidal volumes cannot be routinely modified or as precisely controlled. ■

For more information, contact Anthony DiMarco, MD, MetroHealth Medical Center, (afd3@po.crwu.edu).

Advocacy/Policy Websites

www.disabilityresources.org
General guide to disability resources on the Internet.

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www.ncd.gov
National Council on Disability.

◆◆◆

www.adapt.org
Americans Disabled for Attendant Programs Today (ADAPT). For the latest on MiCASSA (Medicaid Community Attendant Services and Supports Act).

◆◆◆

www.dredf.org
Disability Rights Education and Defense Fund, Inc.

◆◆◆

www.jfa.com
Justice For All e-mail network. Alerts on congressional actions for disability issues.

◆◆◆

www.aimmm.org
Advancing Independence: Modernizing Medicare and Medicaid (AIMMM).

◆◆◆

www.accessiblesociety.org
The Center for an Accessible Society.

◆◆◆

www.familiesusa.org
Families USA's Health Action Network.

◆◆◆

http://cms.hhs.gov
For press releases and policy information on Medicare and Medicaid from the US Department of Health and Human Services.

◆◆◆

www.protectionandadvocacy.com
National Association of Protection and Advocacy Systems (NAPAS), an association of federally mandated programs that protect the rights of persons with disabilities.

Ninth International Conference on Noninvasive Ventilation: "From the ICU to Home"

October 23-25, 2003
Caribe Royale Resort Suites, Orlando, Florida

Sponsored by the American College of Chest Physicians (ACCP)
In conjunction with the National Jewish Medical and Research Center, International Ventilator Users Network (IVUN), and Journées Internationales de Ventilation á Domicile

Ventilator Users Sessions

Thursday, October 23, 1:00–4:30 pm
"Challenges Facing Ventilator Users"
"Empowerment: Organizing and Advocating"

Friday, October 24, 8:30–11:30 am
"Transitions from Teen to Adult, from Middle Age to Elderly"
"Intimacy and Sexuality"

More Topics

Noninvasive ventilation applications in COPD, ALS, and congestive heart failure; surgical and anesthetic considerations for ventilator users; nasal and oronasal masks; assisted cough; patient-provider teams; cough/stress incontinence; ethical case discussions in chronic care; reimbursement issues; caregivers in the home; new ventilation technologies for home care; pediatric ventilation; tracheostomies; and more.

Speakers

Ventilator Users: Larry Becker, PhD; Linda Bieniek, CEAP; Laura Hershey; David Jayne; Audrey King; Bill Miller; and Barbara Rogers.

Faculty: Nicholas Hill, MD; Barry Make, MD; E.A. Oppenheimer, MD; Allen Goldberg, MD; Nicolino Ambrosino, MD; Patrick Léger, MD; Susan Sortor Léger, RRT; Josh Benditt, MD; Gerald Teague, MD; Angela King, RRT; Bart Celli, MD; Mark Elliott, MD; Anita Simonds, MD; Ole Norregaard, MD; Norma Braun, MD; Nancy Collop, MD; Peter Gay, MD; Dominique Robert, MD; and more.

Post-Graduate Courses

October 26, 2003
"Noninvasive Ventilation in Acute Care"
"Noninvasive Ventilation in the Long-term Setting"

Hotel reservations.

The Caribe Royale Resort Suites (800-323-8300, 407-238-8000, www.cariberoyale.com). The conference rate for a queen-sized double room or standard king is $185 per night plus applicable taxes. To receive this rate, ask for "ACCP Noninvasive Ventilation" when making reservations.

Registration fees for ventilator users

Noninvasive ventilation conference *(includes continental breakfast)*:
Before August 29, 2003: $100 After August 29, 2003: $150.
Postgraduate course *(includes lunch)*:
Before August 29, 2003: $200 After August 29, 2003: $250.

For a registration form, contact the ACCP (847-498-1400, www.chestnet.org).

that I work with make recommendations for home ventilators and accessories based on product and brand exposure and knowledge. Since there is almost no scientific evidence regarding the performance characteristics of NPPV interfaces and accessories, marketing leads the way.

"... Currently, I am working on a comprehensive infant case with an HMO that pays about 40% below the Medicare allowable for durable medical equipment. That is about $589 per month for the ventilator. This infant requires pressure support and PEEP. The home ventilators capable of these modes cost 1½ to 3 times the cost of traditional home ventilators. If this case turns out to be short-term and the child can be weaned, I would be left owning a very expensive ventilator. So I choose to rent. The lowest wholesale rental of this category of ventilator in my market is about $550 per month. How can we even think about offering multiple ventilators and interfaces for trial when we may not even be able to provide basic services for a case?

"What we really need Euclid for is a mathematical formula that an HMO can understand that explains the cost-benefit of providing complex care in the home and paying appropriately." ■

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Carlon, C., & Combs, A. (2002). Would Euclid approve of how we select mechanical ventilators? *Chest,* 122, 1881-1883.

JOURNAL ARTICLES

Recent medical journal articles on aspects of mechanical ventilation. If you are not a member of the professional organization that publishes the journals, you can usually obtain abstracts of the articles free online; the article itself can be purchased separately.

Bach, J.R. (2002). Amyotrophic lateral sclerosis: Prolongation of life by noninvasive respiratory aids. *Chest,* 122, 92-98. (www.chestnet.org)

Campbell, R.S., Johannigman, J.A., Branson, R.D., Austin, P.N., Matacia, G., & Banks, G.R. (2002). Battery duration of portable ventilators: Effects of control variable, positive end-expiratory pressure, and inspired oxygen concentration. *Respiratory Care,* 47(10), 1173-1183. (www.aarc.org)

Chatwin, M., Ross, E., Hart, N., Nickol, A.H., Polkey, M.I., & Simonds, A.K. (2003). Cough augmentation with mechanical insufflation/exsufflation in patients with neuromuscular weakness. *European Respiratory Journal,* 21(3), 502-508. (www.ersnet.org)

DiMarco, A.F., Onders, R.P., Kowalski, K.E., Miller, M.E., Ferek, S., & Mortimer, J.T. (2002). Phrenic nerve pacing in a tetraplegic patient via intramuscular diaphragm electrodes. *American Journal of Respiratory Critical Care Medicine,* 166, 1604-1606. (www.thoracic.org)

Eagle, M., Baudouin, S.V., Chandler, C., Giddings, D.R., Bullock, R., & Bushby, K. (2002). Survival in Duchenne muscular dystrophy: Improvements in life expectancy since 1967 and the impact of home nocturnal ventilation. *Neuromuscular Disorders,* 12(10), 926-929.

Hart, N., Hunt, A., Polkey, M.I., Fauroux, B., Lofaso, F., & Simonds, A.K. (2002). Comparison of proportional assist ventilation and pressure support ventilation in chronic respiratory failure due to neuromuscular and chest wall deformity. *Thorax,* 57, 979-981. (www.thorax.bmjjournals.com)

Highcock, M.P., Morrish, E., Jamieson, S., Shneerson, J.M., & Smith, I.E. (2002). An overnight comparison of two ventilators used in the treatment of chronic respiratory failure. *European Respiratory Journal,* 20(4), 942-945. (www.ersnet.org)

Janssens, J., Derivaz, S., Breitenstein, E., de Muralt, B., Fitting, J., Chevrolet, J., & Rochat, T. (2003). Changing patterns in long-term noninvasive ventilation. *Chest,* 123, 67-79. (www.chestnet.org)

Nùrregaard, O. (2002). Noninvasive ventilation in children. *European Respiratory Journal,* 20(5), 1332-1342. (www.ersnet.org)

Stone, A.C., Nolan, S., Abu-Hijelhia, M., McCool, D., & Hill, N.S. (2003). A novel form of manually assisted ventilation. *Chest,* 123, 949-952. (www.chestnet.org)

Stuart, M., & Weinrich, M. (2001). Protecting the most vulnerable: Home mechanical ventilation as a case study in disability and medical care: Report from an NIH conference. *Neurorehabilitation and Neural Repair,* 15(3), 159-166.

IVUN News
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Inside This Issue ...

IVUN News Now 12 Pages

The Board of Directors of Gazette International Networking Institute (GINI), the governing organization of IVUN, decided to discontinue publication of its bi-annual *Rehabilitation Gazette* in 2003 and to expand the quarterly *IVUN News* to 12 pages, and to rename the publication *Ventilator-Assisted Living.* Subscribers to the *Rehabilitation Gazette* will receive this expanded newsletter.

The inaugural issue of the biannual *IVUN News* in 1987 introduced the world to the International Ventilator Users Network (IVUN). IVUN is a worldwide network of ventilator users, health care professionals, and ventilator equipment manufacturers and dealers experienced in home mechanical ventilation. Polio survivors form the nucleus of IVUN because of their use of mechanical ventilation, such as the iron lung, during the polio epidemics. Their experiences with home ventilation are invaluable for persons with high level spinal cord injuries, muscular dystrophy, ALS, and other neuromuscular diseases, and for the growing number of ventilator-assisted infants and children. Holding fast to the network's mission, *IVUN News* expanded to quarterly issues in 1998.

Articles from *IVUN News* are online (www.post-polio.org/ivun/ivun-news.html) to provide an immediate and reliable source of information for the home mechanical ventilation community. The comprehensive *IVUN Resource Directory* is also online (www.post-polio.org/ivun.d.html) and is updated continually.

Watch for the first issue of *Ventilator-Assisted Living* ... coming in the summer of 2003. ■

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