

## COMPUTER BECOMES THERAPY, LINK TO WORLD FOR PATIENT

by Anne Federwisch, OTR

A lot has changed since Jim Lubin began exploring cyberspace 14 years ago. Then, the technology merely allowed him to run an electronic bulletin board with his computer, two floppy disks, and a 300-baud modem. Only computer professionals and enthusiasts were interested in communicating via computer.

The information highway has changed as more people have joined Jim in cyberspace via the Internet and the World Wide Web. And Lubin's use of the computer has also changed from an interest to a necessity.

In a matter of hours on May 17, 1989, acute transverse myelitis left Lubin a ventilator user and paralyzed below the second cervical vertebra (C2 level). Yet Lubin has not allowed quadriplegia to keep him from going on-line with his computer for up to nine hours a day. His Web site (which includes his *disAbility Resources* on the Internet) has become popular with more than 100 people "visiting" Lubin and his page each day.

Lubin remembers only the beginning of that May 17. He awoke with a sore shoulder, and his mother, Helena Lubin, suggested he take the day off from his job as a supervisor in the assembly department of Heart Technology, a manufacturer of angioplasty equipment in Redmond, Washington. But Lubin did not want to stay home from a job he loved because of what he thought was only passing discomfort.

Within the first hour of work it became apparent that the sore shoulder was a symptom of a

more serious problem. Lubin lost consciousness. His heart stopped. Paramedics from the fire station across the street from Heart Technology had to revive him several times.

When Helena Lubin arrived, the paramedics informed her that they did not think Jim would survive. "But I started calling to Jim, and he seemed to respond to my voice," she said. "Later, the doctors came right out and said the very fact that Jim lived is a miracle."

Jim spent the next 21/2 months in intensive care. Physicians initially suspected a cerebrovascular accident, Lyme disease, or an adverse reaction to chemicals used at his job. An MRI revealed acute transverse myelitis, an inflammation of the spinal cord, at the C2 level. Physicians theorize a viral infection reached Lubin's spinal cord

through a minute crack in his spinal column that may have resulted from a fall from a tree as a child.

From August 1989 through January 1990, Lubin was in rehabilitation at the University of Washington Medical Center in Seattle. "Rehabilitation was kind of boring most of the time



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because I was a C2 quad — I couldn't do much," he said.

Lubin said therapy grew more interesting once he gained access to a computer. His regular occupational therapist called in Denis Anson, MS, OTR, a computer access specialist and lecturer in the division of occupational therapy in the University of Washington's department of rehabilitation medicine, to determine the best way for Lubin to use the computer.

Anson taught Lubin how to use Morse code with a sip-and-puff interface. Lubin gently inhales and exhales — sips and puffs — into an air switch connected to a computer. An adaptive device translates each sip into a dot in Morse code and each puff into a dash. The dots and dashes are translated into commands and letters on the computer screen.

Lubin mastered Morse code and the sip-and-puff technique in less than a month. A special valve (Passy-Muir Tracheostomy Speaking Valve) lets air go into his trach but blocks it from going out, allowing him to speak and to sip and puff while on the ventilator. "That way I have to exhale through my mouth and/or nose. Also I can hold my breath this way," Lubin said.

Lubin said that only a little air is needed to activate the sip-and-puff switches. "It's more just moving my tongue to change the pressure in my mouth. I just do very fast sips and puffs. I keep exhaling through my nose while I am typing."

Lubin now uses the sip-and-puff technique at home on his computer to access the Internet. Using an adaptive device called Adap2U, made by AdapTek Interface in Mercer Island, Washington, Lubin sips and puffs into an air switch to type into the computer.

Using the technique, Lubin can now type 17 words a minute. He can keep up with almost

Traumatic spinal cord injury is the most familiar cause of quadriplegia. Other disorders can also cause spinal cord injury, such as myelitis, a condition causing inflammation of the spinal cord. Myelitis may involve any level of the spine, but when limited to a few segments, it is called transverse myelitis. Usually due to a viral infection, the cause is often unknown. Symptoms may include back pain, loss of coordination, urinary retention, and loss of bowel control. The syndrome may develop over a period of months or years or be of sudden onset with no symptoms at all. There may be some recovery of function.

everyone in on-line chat rooms, in which computer users "converse" by typing on their computer screens.

Lubin does much more than entertain himself with the computer. In exchange for his service provider account (his access to cyberspace), Lubin provides on-line support for GENie (General Electric Network for Information Exchange). Via e-mail he answers users' questions for two to three hours a day. He also occasionally programs for his former employer, Heart Technology, in exchange for computer equipment.

In early 1994, Lubin began compiling information for his personal Web site so he could link to his favorite sites more easily. Eventually, he put his Web site on-line so others could access it. His current Web home page includes links he has set up to sites relating to astronomy, computers, geography, government, magazines, movies, music, news, shopping, and television. His most popular Web page is his *disAbility Resources*, a compilation of resources of interest to people with disabilities and those who work with them. Among the resources are links to information about Social Security and the Americans with Disabilities Act.

"At first I just included sites I've found helpful," Lubin said. "Then people began writing to me and saying they had an interesting Web site, so I added those. They started saying they really appreciated my site, so I figured I should update it more often. The easier it is for people to find resources, the better it is."

The computer is Lubin's primary means of connecting with the world. Getting out of the house can be difficult. Lubin must make reservations two days in advance to use a public van that can accommodate his wheelchair. "I don't go out of the house much," he said. "But I'm very active on the computer. I can make good friends over the computer. I'm always helping people."

Lubin is one of an estimated 200,000 people with spinal cord injuries in the United States. And of the 16,024 people listed in the National Spinal Cord Injury Database, only 1.1 percent have impairment at the C2 level. Many people with quadriplegia use computerized controls to interact with their environments, performing tasks including turning on lights. Lubin takes that further by using his computer to interact with the world.

Helena Lubin is proud of her son and the way he helps people despite his paralysis. "Jim is a positive person, and he's always been that way," she said.

She said that physicians predicted a significant change in his condition in the seventh year after

onset of the acute transverse myelitis. "From this year on, we should see changes," she said. "Jim is experiencing pain on his right side, but we don't know for sure if his condition will improve."

Jim Lubin is pleased to see others recognize the therapeutic potential of on-line computing for people with disabilities. A few years ago he advised the publishers of a spinal cord injury newsletter to put the publication on-line. "I had stopped reading it because it was a hassle," he said. "Someone (had to be) here that could turn it (the page); I just didn't want to be a bother." Now that the publishers have followed Lubin's advice and put the newsletter on the Internet, Lubin can browse it independently.

Lubin's quadriplegia may severely limit his physical abilities, but he has no limitations in cyberspace. "(On the computer) people don't know I'm disabled unless I tell them," he said.

**EDITOR'S NOTE:** Jim uses trach positive pressure with PLV-100 ventilators, one on his wheelchair and one at bedside, both of which were purchased by his insurance company. His mother and brother are his primary caregivers, plus Jim receives 80 hours of nursing care per week. He has recovered feeling/sensation throughout his body except for his right hand and arm. He can now sip-and-puff up to 19 words per minute.

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### **International Ventilator Users Network (I.V.U.N.)**

*links ventilator users with each other and with health care professionals interested in home mechanical ventilation and home care.*

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## **ADAPTING TO MASKS AND BiPAP®**

*by David Ronfeldt*

As a respiratory polio survivor, I spent many months during 1949-1950 using an iron lung. After that, I was able to breathe quite well on my own, despite limited vital capacity, until 1978, when I was hospitalized twice with respiratory failure. I began using a chest shell at night. This was adequate until 1994, when the chest shell ceased to fit well.

Thus began my efforts to switch to mask ventilation and the BiPAP® system. I rushed into the transition, trying to force my way during the December holiday break with time off from work. It turned into a major ordeal, and after a couple of weeks of uncertain ventilation, mounting sleep deprivation, and erratic progress, despite encouraging efforts by many sides, I retreated to an old, modified chest shell for several months.

I renewed my effort, this time using a gradual approach in which I started a night's sleep with the mask and BiPAP®, then switching to the chest shell after a few hours to complete the night. I tried afternoon catnaps to gain additional experience adapting. Eventually, over a six-week period, the time spent with the mask lengthened.

I have spent over two years using only the mask and BiPAP® for respiratory support during the night and for day-time naps. Because of this experience, I believe there are three steps to adaptation: getting accustomed to the mask, learning to fall asleep, and staying asleep.

### **Getting Accustomed to the Mask**

The process begins by wearing the mask in order for the face to become accustomed to it. This may take several weeks. It is important to get the right mask in the right size. I tried several masks and nasal pillows, but none felt right until the Sullivan "bubble" mask which I have used for two years. (Fortunately, my doctor recommended that the vendor provide several different masks for me to try. This is not standard practice, but it should be. Perhaps a "beginner's package" of several different masks?)

One weird experience with the mask was a sensation that the air inside it, after exhalation, was not fresh. It seemed a bit stifling. I was reassured that the sensation was common for first-timers. I relieved it by opening one of the tiny ports I found on the mask and that provided a sensation of fresh air. After I became accustomed to the mask, I found no reason to continue keeping the port open. I was informed that I shouldn't have

done this because it has the effect of reducing the air inflow pressure. I had not felt much difference in the pressure, and a bit of relief here and there made it easier to cope with the mask experience.

I encountered a tendency in some therapists to fit me with mask sizes which were a bit small; I think a bit large may be preferable. I was told that I would have to shave off my mustache for the mask to fit well, but my mustache never proved a source of leaks. I deduced that if the mustache hairs are fairly thick, but not bushed out, and long — down to the top of the upper lip, the mask may press down on the hairs lengthwise and not lose the seal. If the hairs are trimmed short, then the mask may ride on the stubby tips of the hairs and lose the seal.

### Learning to Fall Asleep

The next step is to fall asleep wearing the mask. I could not fall asleep for more than a few seconds in my first effort. Part of this step is just sticking with the process and I was more successful later. It also is important to get the proper settings on the BiPAP® S/T system.

I found the T setting worked best at first, partly because on the S/T setting every swallow or other movement would trigger an untimely, disorienting blast of air. I did not know about the %IPAP knob which plays a crucial role in the T setting by determining the length of time air is pushed into the mask during the inhalation cycle. It took a while to find the proper setting for me. I found that I needed to increase the IPAP setting from about 10, which was fine while I was awake, to a little more than 12, to compensate for shallower respiration during sleep.

Gradually, as I became used to the mask and made small refinements in the settings, I started falling asleep for brief periods with less and less delay, but I still was not staying asleep for long.

### Staying Asleep

The third step is staying asleep for several hours. By now, I had a good mask and the right IPAP settings. I had been using an EPAP setting of 4, but one night I dropped it to 3, and immediately had my first good night's sleep. Another change that helped was to shift from the T to the S/T setting. I had adapted enough that I felt the T setting was not quite matching what my lungs wanted to do. I now had enough experience to know when to swallow so as not to trigger the inhalation cycle in the S/T setting.

I know that doctors and respiratory therapists prefer to determine the proper settings, but when an individual is having trouble adapting, he or she should take an active role in experimenting

with the settings — a point with which my doctor agrees.

### Retrospect

The whole process took many months during 1995, but I consider myself adjusted and thankful for the mask and BiPAP® system. However, this does not mean I am a completely happy user. I continue to have a difficult time with extreme dry mouth — in which my lips, gums, tongue, roof of my mouth, and front of my throat are dried out and stuck together. Saliva flow has ceased. It seems to start right after I fall asleep and continue through the night. Therapists and other users insist I must be leaking air through my mouth, but I am convinced this is not the case. A humidifier can help with nose and throat dryness, but I am not bothered by that, and the humidification I have tried has had no effect on my dry mouth. I know what may make it worse, e.g., some foods and drinks, medications, higher settings, but I cannot find a way to resolve the problem. I have had hopes for a chinstrap, lipseal, hose positions to ease the mask's pressure on the upper lip, herbal syrups, certain foods, etc., but to no avail.

Thanks to the BiPAP® system and mask, I know I am breathing better than I did with the chest shell. The new technology is a lot easier to travel with than the old and so much more reliable and sturdy that I do not need to carry a backup machine — decisive pluses!

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## LIVING WITH ARTHROGRYPOSIS

*by Janet Prebul*

I was born with arthrogryposis 25 years ago, a condition that affects the nerve supply to the muscles. I have limited movement in my arms and legs. Because I do not bend very well at the waist, I usually stand or lie down. I rarely ever sit, usually only when I ride in a car. I can walk, but use a specially-designed, one-of-a-kind electric wheelchair when I go out that allows me to remain standing.

I have used tracheostomy positive pressure ventilation (with a Bear 2 ventilator and LP6 as backup) since I was 12 years old, due to a bout with pneumonia. I have about seven hours of ventilator-free time during the day (five hours free, then two hours with the ventilator, before the last two hours free). I use oxygen during my free time. My

trach is plugged with a trach button; I do not have a Passy-Muir valve. The trach helps clear excess secretions, because I have a difficult time



coughing them up when I become heavily congested. I have been hospitalized with pneumonia more times than I can remember, and frankly more times than I care to remember. Some of those times were very tough, but I have always managed to bounce back.

Currently, I receive eight hours of nursing care per week, divided into two sessions. The nurse assists with my breathing treatments, and helps me to get out of bed and to dress in the morning. My parents are my primary caregivers.

Having respiratory problems and using a ventilator has slowed me down, but it certainly hasn't stopped me. I have completed one year of college and would eventually like to obtain a degree in psychology in order to work with chronically ill or disabled children.

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## CCHS REGISTRY

Over the last several years, Drs. Debra Weese-Mayer and Jean Silvestri at Rush Children's Hospital in Chicago have evaluated nearly 50 children with idiopathic congenital central hypoventilation syndrome (CCHS) and others with alveolar hypoventilation of other etiologies. Through thorough questioning of the referring physician and parents, coupled with careful review of the medical records, they have been able to diagnose children with CCHS as distinct from other etiologies of alveolar hypoventilation, and to make a number of observations and offer insights about children with CCHS.

Based upon these years of experience with diagnosis and clinical management, coupled with their search for patterns of inheritance and investigation into the molecular genetics of CCHS, they have established an International CCHS Registry. The purpose is to standardize the evaluation of children with a presumptive diagnosis of CCHS, maintain an ongoing database to be accessed for further research, and expand a blood bank for research into genetic testing of children with CCHS, their parents, and siblings.

Families interested in participating in the genetic research studies should contact Angela Huffman, RN, The International CCHS Registry, 622 Murdock, Rush Children's Hospital, 1653 West Congress Parkway, Chicago, Illinois 60612 (312/942-2723).

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## CCHS UPDATE: MATT STONE

Matt Stone, now 11 years old, appeared in *I.V.U.N. News*, Spring 1995. Nancy Stone, Matt's mother, relates the following in the March 1997 issue of the *CCHS Family Newsletter*:

"Even though Matt has had a relatively uncomplicated case of CCHS, I still remember the feelings of being sick and of being cooped up after various respiratory illnesses, feelings of isolation — both social and physical. I don't think I realized at the time how many illnesses Matt had, as they were rather mild (he was never hospitalized for illness and he usually recovered fairly quickly), but it still meant staying home, extra ventilator time, missing school, etc. Matt has been using nasal ventilation since 1994, and has had only a couple of minor colds. At his last doctor's check-up, he had gained 6 pounds and grown two inches (in three months). We have been very pleased with his progress.

"If anyone would like to discuss his switch to nasal intermittent positive pressure ventilation and the PB335, please call us at 317/299-0801 or e-mail: [bstone@indy.vax.iupui.edu](mailto:bstone@indy.vax.iupui.edu)."

### MOVING?

Please send both your old and new addresses to:  
I.V.U.N.

4207 Lindell Boulevard, #110  
Saint Louis, Missouri 63108-2915 U.S.A.

## FORUM ON HOME MECHANICAL VENTILATION

MAY 30, 1997

### ■ THE DANISH EXPERIENCE

*Grethe Nyholm, RNP, Home Ventilation Department, University Hospital, Copenhagen*

Ventilator care in Denmark is divided between two centers. Center West in Arhus maintains 124 ventilator users out of a population of 2.7 million; Center East in Copenhagen maintains 142 ventilator users from a population of 2.5 million. At each center, there is uniform criteria for someone to begin home ventilation.

At my center in the east, there is an outpatient clinic and a sleep disorder laboratory. I am head nurse of a ward with ten beds for ventilator users. The ratio of nurses to patients is usually 2:1. Respiratory polio survivors have a sleep study every year, beginning when they started ventilator use and then every three months to determine if ventilator use is benefiting them. Our center has a coordinating nurse who cares for the outpatients. He educates patients and their families on ventilator use, registers and checks all the equipment, and coordinates home oxygen therapy.

With Denmark's social system, every person with a severe disability receives paid attendant care 24 hours per day. Ventilators are provided free of charge, as is service. If someone is a 24-hour ventilator user, he or she receives two ventilators — one for bedside and one for a wheelchair. Specially adapted flats for people with disabilities, as well as cars, can be obtained on favorable terms without the high Danish tax. Also paid attendant care is available for holidays.

In 1952, Denmark experienced its largest polio epidemic. Many polio survivors had to be venti-



lated manually or "bagged." A small Danish ventilator, the Pulsula, was invented. It had many advantages, one being a built-in humidifier, and it is still being used.

Today, my center follows 24 patients with post-polio syndrome. The complaints at admittance were fatigue, headache, sleeping disorders, and respiratory symptoms. Ten patients have tracheostomies (seven after a crisis, three after a trial of noninvasive ventilation failed). Mean vital capacity was 800 ml. Fourteen patients with a larger vital capacity of 1700 ml use the BiPAP® system during the night. Polysomnography on this group before ventilation revealed severe nocturnal hypoxia and an increase in PCO<sub>2</sub>. BiPAP® effectively reversed the hypoxia and reduced PCO<sub>2</sub>. In the case of pulmonary hypertension, invasive ventilatory treatment must be recommended.

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### ■ THE JAPANESE EXPERIENCE

*Yoichi Sakakihara, MD, PhD, Department of Pediatrics, University of Tokyo*

The incidence of polio has been drastically reduced in recent years. In the near future, the number of people with mechanical ventilation due to post-polio disability will decrease, and the spectrum for people with disabilities who need mechanical ventilation will also change.

In a national survey I conducted in Japan in 1993, questionnaires were mailed to 2,425 hospitals with departments of pediatrics. From those, 282 hospitals were taking care of 434 longterm (more than 3 months) ventilator-assisted children (under 20 years of age) — either in a hospital or at home. Only 61 out of 434 children needing longterm ventilation were at home. Average duration of home ventilation was 3.9 years, but 11 children had been ventilator-assisted for 10 years.

The basic disorders causing respiratory insufficiency included muscular dystrophy and other



myopathies, hypoxic encephalopathy, Werdnig-Hoffmann disease (spinal muscular atrophy Type I), chronic lung disorders of prematurity, Ondine's curse (CCHS), drowning, and congenital heart disease.

Of the many issues related to ventilator-assisted children, the major question is why only 10-15% are living at home. One problem might be associated

with the cost of ventilator care, but it is easier now that ventilators are provided for free. All the patients have some reimbursement or some money from the government (about \$400 per month for maintenance and supplies), so this does not explain why in-home ventilator care is unpopular in Japan. Another problem is that paid attendant care is not provided by insurance. In-home nursing service, only about 1.2 hours per day, is not as readily available as in the United States, where the average is 14.8 hours per day.

The second issue is an ethical issue. There is disagreement among pediatricians in Japan on whether to initiate ventilator care for children with Werdnig-Hoffmann's disease. I conducted another survey of about 30 doctors currently taking care of children with Werdnig-Hoffmann's. I asked the doctors if they were satisfied with the quality of life for their patients. Unfortunately, most of the doctors answered that they were unsatisfied with the quality of life, but most of the children were living in the hospital. Physicians have no guidelines for these ethically difficult cases. There is no ethical consensus for the prolongation of life and much debate is going on about that now.

In conclusion, the major issues concerning home mechanical ventilation in Japan are insufficient nursing for home care, no guidelines for ventilator cases, no ethical consensus for prolonging life, and inadequate quality of life for ventilator users.

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## ■ THE JAPANESE EXPERIENCE

*Kimiyo Satoh*

I was born in 1962 and I am now 34 years old. The name of the disease which caused my disability is Kugelberg Welander disease (spinal muscular atrophy Type III). I use trach positive pressure ventilator 24 hours a day. I move around on a gurney.

Twenty-two years ago, when I started to use a ventilator, there was no portable ventilator in Japan yet; what we had was a ventilator about the size of a refrigerator. Therefore I had no choice but to stay in bed all day. At that time, I was told by a doctor that I could only survive in a ward of the hospital all my life. I had no hope in my future.

While living in a hospital bound by rules just like a prison, I always had a yearning for freedom. I never abandoned my dream to live in the outside world. One

day I was handed a brochure and learned about a portable ventilator for the first time. In the brochure, I saw a picture of a disabled person moving around with the ventilator mounted in the back of his wheelchair; that picture gave me a big courage.

When I was 27, I got out of the hospital and started to live on my own. Having no institutionalized assistance from society, it wasn't as easy as I had imagined to live on my own, but the freedom that I gained was very precious and irreplaceable by anything else. The road to actualizing my dream was very harsh. There was no information at all around me about people living at home using a ventilator, and the medical specialists kept telling me that it was dangerous to take ventilators out of the hospital. It took a long time to convince them otherwise.

After I started to live on my own, I established Japan Ventilator Users Network (J.V.U.N.). I had no friends who used ventilators then; I needed a network where ventilator users could share their



feelings, problems, information, and other experiences. Regrettably there is no governmental financial support for J.V.U.N., but 800 people support our organization. About 100 out of the 800 are ventilator users. J.V.U.N. publishes a newsletter called *Another Voice*.

Currently, it is estimated that there are about 8,000 ventilator users in Japan, but those living at home constitute less than one percent of this figure. Most ventilator users are still confined to hospitals. The primary reason is that most of the medical specialists think of a ventilator as life support equipment.

People with disabilities have been told that having a severe disability and living with a ventilator is worthless. But for us, the parties concerned, a ventilator is just like a wheelchair or a pair of crutches, a tracheal tube like pierced jewelry. We have been tenaciously insisting that ventilators are merely tools to support our independence.

In 1992, the movement of ventilator users and their families bore fruit; national medical insurance started to cover home ventilation therapy. Ventilators were able to be rented free of charge in 1994. (The government pays the hospitals the cost of the ventilator equipment.) Until then a ventilator user had to purchase a ventilator which costs as much as 3 million yen (approximately \$25,000 USD).

However, we can only rent one ventilator, and if we need two ventilators in case one fails, we must purchase the backup ventilator. We receive an allowance of about \$400 per month from the central and local government to pay for maintenance and some supplies. Attendant care assistance for ventilator users is not guaranteed by the government, and the reality is that in-home care is supported by the devoted efforts of family members.

Tracheal suction care cannot be legally performed except by medical doctors and nurses; home helpers are not allowed to perform any home medical care, even if it is applying skin lotion. Visiting nurses come about one to three times a week for two hours per visit, which hardly guarantees medical care needed 24 hours a day. School teachers also cannot suction. Consequently, the radius of action of many ventilator users is very limited.

I think suction care can be performed by anyone who has received basic training. I know it very well because I perform suction on myself. Suction care is everyday care, just like eating and

changing the body position, and attendants and family members can be easily trained to do it.

Even with these problems, home ventilator users are increasing. We are especially proud of the children with Werdnig-Hoffmann's disease who use a ventilator and attend regular elementary school, studying together with nondisabled children. We ventilator users are messengers who can tell the world about the brightness and wonderfulness of human lives.

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## ■ THE AMERICAN EXPERIENCE

*E.A. Oppenheimer, MD, Department of Pulmonary and Critical Care Medicine, Southern California Permanente Group*

Since 1985, I have coordinated a regional home mechanical ventilation (HMV) program which provides support for a population of about five million people in southern California. This is approximately the same size population base as the two ventilation centers in Denmark. We had about 160 people with severe disabilities using longterm mechanical ventilation 20-24 hours per day who were trapped in our hospitals. At that time, 1985, no one thought they could be at home. Our program evaluated whether safe and successful home care could be provided. We found that if people/families had equipment, training, an adequate number of caregivers, and a support team of experienced healthcare professionals, they could be happy, successful, and safe at home. These people felt that the quality of care was better at home, in part because they usually had one-on-one care. A major problem was, and



still is, obtaining resources to pay for in-home caregivers when needed to assist the family.

In 1996, the American College of Chest Physicians (ACCP) — who published guidelines for home mechanical ventilation in *Chest* (1986) — established a new Home Care Section. ACCP has an active Web site which can help physicians and the public understand home ventilation issues better, particularly new noninvasive methods. ACCP plans to publish updated HMV guidelines later this year.

In the United States, we estimate that there are approximately 10,000 to 15,000 people using mechanical ventilation at home, with a growing number in longterm care institutions. The number of people at home has been increasing, probably because of the success of intensive care and the great advances in noninvasive mechanical ventilation, particularly nasal positive pressure ventilation. We have excellent equipment available in the United States; we have excellent home care services. There are many excellent centers experienced in HMV. But these centers are not linked or coordinated with a system to provide support to all our communities in the United States. There are still great gaps in knowledge and understanding related to how to evaluate people, as well as planning and supporting home ventilation. Thus, there is tremendous need for a clinical care network for HMV; one doesn't exist. Consequently, one of the most frequent complaints I hear from patients is that the physicians and allied health professionals are not knowledgeable.

Of the people who are using home mechanical ventilation, in our program, about 25% have COPD, about 25% have motor neuron diseases like ALS, and the remaining 50% have a variety of neuromuscular and musculoskeletal conditions. One situation frequently encountered by people with neuromuscular disease occurs when they are found to have a low arterial oxygen level. They are often given oxygen, rather than the physician realizing that this is caused by an elevated CO<sub>2</sub> level due to hypoventilation, weak respiratory muscles, and that they really need assisted ventilation, not oxygen. Another major problem is that there is not enough experience in helping them with assisted cough techniques, which help people with weak respiratory muscles to clear secretions and to avoid aspiration and pneumonia.

Tracheostomy ventilation is still probably overused in the United States. Most pulmonary physicians focus on intensive care; they are very familiar and comfortable with endotracheal tubes and

tracheostomies, but not with longterm and home ventilation. This is improving, although there is a long way to go. Many people with neuromuscular disease are advised to have a sleep study when respiratory symptoms occur; and then they are often advised that they need CPAP (used for sleep apnea).

However, CPAP puts an even greater burden on their respiratory muscles; they usually need assisted ventilation, not CPAP. Another problem is that symptoms of developing respiratory failure are not identified early enough. The progressive nature of both obstructive (such as COPD) and restrictive neuromuscular and musculoskeletal diseases is often not followed so as to anticipate respiratory failure, and plan desired options. Thus, advanced planning is neglected. Very frequently, people have a respiratory crisis and end up in an emergency room, where they have little chance to participate in decision-making, and later on undergo a tracheotomy.

There is a very individualistic social philosophy in the United States. An informed person's right to refuse or withdraw care is strongly supported legally and ethically. In Japan, in contrast, once mechanical ventilation is started, it is not accepted that if you want to stop using it, that that is an appropriate thing to do. This is the same in many parts of Europe. On the other hand, the United States has not supported universal access to health-care; other countries put a higher value on equity and social justice. The greatest obstacle to long-term home mechanical ventilation here is the lack of social and economic support for paid caregivers or attendant services. This leaves a tremendous burden on family caregivers as well as the ventilator user. Thus, the major problems I see in the United States are the lack of widespread appreciation by healthcare professionals of the issues involved in longterm HMV (particularly noninvasive ventilation), as well as the great gaps in social resources. I look forward over the coming years for increased understanding of these issues, progress in solving them, and a closer working together of I.V.U.N. and the ACCP.

**ADDRESS:** E.A. Oppenheimer, MD, Southern California Permanente Medical Group, 4950 Sunset Boulevard, Los Angeles, California 90027-5822 (213/783-7106 or e-mail: eaopp@ucla.edu)

**Frog Breathing with Gary McPherson**, a nine-minute videotape, demonstrates the technique of glossopharyngeal breathing or, as it is commonly known, frog breathing. Polio survivor McPherson uses the technique to breathe on his own without the use of a ventilator for parts of the day. The video is available for \$50 (Canadian) from the University of Alberta, Health Sciences Media Services, Carole Sweeney, 0J1 Mackenzie Centre, Edmonton, Alberta T6G 2R7 Canada (403/492-6560 or 403/492-7973 FAX).

**FES Information Center** offers a wealth of information about the application of functional electrical stimulation on people with neuromuscular disorders to provide restoration of movement or function, as in cough. Much of the information is free or at a nominal cost. Contact: FES Information Center, 11000 Cedar Avenue, Suite 230, Cleveland, Ohio 44106-3052 (800/666-2353 or 216/231-3258 FAX; e-mail: fes\_info@po.cwru.edu).

**Nellcor Puritan Bennett** was acquired by Mallinckrodt in August 1997. Nellcor Puritan Bennett had acquired Aequitron Medical, Inc. last December. Mallinckrodt is a Saint Louis-based medical company whose products include X-ray contrast media, radiopharmaceuticals, and devices for imaging. Nellcor Puritan Bennett will be combined with Mallinckrodt's critical care unit; it will retain the same name.

**Phantom™ Nasal Mask** is a new product from SleepNet. The gel cushion conforms to the face, minimizing leaks and discomfort. It is latex-free and hypoallergenic. The dual-strap headgear has integrated swivel tubing. For more information about the mask, contact Venise Traficante, SleepNet, 1050 Perimeter Road, Manchester, New Hampshire 03103 (800/ABC-DPAP or 603/641-9440 FAX or Web site: www.dpap.com).

**Alliance for Technology Access (ATA)** is a national network of 42 community-based technology resource centers, plus technology designers, developers, and vendors. The non-profit ATA centers are the products of collaboration among people with disabilities, family members, and professionals in related fields. Registration in the program is available at different rates, online or hard copy. *Computer Resources for People with Disabilities*, 2nd edition, is available for \$17.95 (paper) or \$22.95 (disk) plus \$3 shipping and sales tax 7.25%. Contact: ATA, 2175 East Francisco Boulevard, Suite 1, San Rafael, California 94901 (415/455-4575, 415/455-0491 TTY, e-mail: atainfo@ATAccess.org, or Web site: www.ATAccess.org).

**Attention: Pneumobelt Users** Recent inquiries to the I.V.U.N. office suggest that some pneumobelt users are having difficulty in maintaining their old Bantam ventilators which power the pneumobelt, and in adjusting to a more modern ventilator, such as the PLV-100. They are also having a harder time finding replacement corset/bladders. (Ventilator users in this situation should be evaluated for signs of underventilation and perhaps move to a more effective/efficient mode of ventilation.)

In any case, I.V.U.N. would like to hear from pneumobelt users who are having similar difficulties and from anyone who has suggestions/solutions.

Write: I.V.U.N., 4207 Lindell Boulevard, #110, Saint Louis, Missouri 63108-2915 U.S.A. (314/534-0475; 314/534-5070 FAX; or e-mail: gini\_intl@msn.com).

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## MDA PUBLISHES ALS GUIDE

*When a Loved One Has ALS: A Caregiver's Guide* is a new publication from The Muscular Dystrophy Association. The 94-page manual is a good beginner's guide for those newly diagnosed with ALS and their families. The guide covers the physical aspects of the disease, including respiratory and communication problems, as well as the emotional side of caring for a person with ALS. Practical matters such as home healthcare, hospice care, living wills, and financial support are included. An extensive list of references will lead caregivers to books, organizations (although not The ALS Association), Web sites, the addresses of local MDA offices, plus a list of the medical directors at the MDA clinics.

The guide is free to any primary caregiver who is registered with MDA. The cost is \$10 to all others. Call MDA at 800/572-1717, to locate the office nearest you or e-mail: mda@mdausa.org. MDA's Web site is www.mdausa.org.

People with ALS and their caregivers looking for broader coverage of living day-to-day with ALS should consider the six newly revised manuals published by The ALS Association. These are free to people with ALS, except for shipping costs of \$3 per manual. Contact ALSA, 800/782-4747, or e-mail: eajc27b@prodigy.com. ALSA's Web site is www.alsa.org.

# MONITORING RESPIRATORY FUNCTION

by John R. Bach, MD,

Co-Director, Jerry Lewis Muscular Dystrophy Association Clinic of UMDNJ-NJMS

Dr. Bach has developed the following regimen for the management of people with muscular dystrophy and has applied it to respiratory polio survivors as well.

## OPTIMAL SCREENING

The individual is routinely screened every three to six months for symptoms of chronic alveolar hypoventilation. At this screening, the following are measured:

- Vital capacity (VC), sitting and supine
- Maximum insufflation capacity (maximum volume of air-stacked breaths) — Collins Survey Spirometer, Collins, Inc., Braintree, Massachusetts
- Assisted and unassisted peak cough flow (PCF) — Access Peak Flow Meter, HealthScan, Inc., Cedar Grove, New Jersey
- SaO<sub>2</sub> — Ohmeda Model #3760, Louisville, Colorado
- End-tidal carbon dioxide (ETCO<sub>2</sub>) monitoring — Microspan 8090 capnograph, Biochem International, Waukesha, Wisconsin

An oral-nasal interface or lipseal (Nellcor Puritan Bennett, Minneapolis, Minnesota) is used for spirometry when lip muscles are too weak to grab a mouthpiece. For assisted PCF measurements, the individual is insufflated to the maximum insufflation capacity, then the expiratory muscles are assisted by coordinating an abdominal thrust to the glottic opening. Other instruments for volume, flow, and noninvasive blood gas measurements are available. Those used by Dr. Bach are noted for the reader's convenience.

## NIGHT-TIME TESTING

If the individual has symptoms of alveolar hypoventilation, VC below 600 ml, elevated ETCO<sub>2</sub>, or periods of daytime SaO<sub>2</sub> below 95% — continuous monitoring of nocturnal SaO<sub>2</sub> is carried out for one or two nights.

If the individual has symptoms of alveolar hypoventilation and/or the nocturnal SaO<sub>2</sub> mean/hr is below 94% during any hour of sleep, he/she is given a trial of nocturnal nasal intermittent positive pressure ventilation (NIPPV) using a portable volume ventilator.

## ASSISTED VENTILATION

The individual is encouraged to use nocturnal NIPPV nightly if she/he experiences symptomatic relief and/or if the nocturnal mean SaO<sub>2</sub> has been demonstrated to have increased. With time, more than nocturnal use becomes necessary. Seven to 16 hours per day is considered part-time use and greater than 16 hours per day is considered full-time use.

## INEFFECTIVE COUGH

The individual is at risk for respiratory failure associated with upper respiratory tract infection (URTI) when the maximum assisted PCF is below 270 L/m (4.5 L/sec). The following measures are taken before a respiratory infection occurs: the person is trained to air-stack with manual resuscitator-delivered volumes to maximum insufflation capacity, trained in manually assisted coughing, and trained in mechanical insufflation-exsufflation (mechanically assisted coughing). She/he is prescribed an oximeter.

## MANAGING AN URTI

If the person is not already using NIPPV and develops URTI, she/he is provided with rapid access (less than two hours) to a portable volume ventilator, to various mouthpieces and nasal interfaces, and to a mechanical insufflator-exsufflator (In-exsufflator, J.H. Emerson Co., Cambridge, Massachusetts).

The individual and caregivers are instructed in monitoring SaO<sub>2</sub> during URTIs. They are trained that a fall in SaO<sub>2</sub> below 95% indicates either hypoventilation and/or bronchial mucus plugging. They are trained to correct the fall in SaO<sub>2</sub> by using NIPPV and manual or mechanically assisted coughing.

The person and his/her caregivers are taught to seek prompt medical attention under any of the following circumstances: baseline SaO<sub>2</sub> decreases below 92%, dyspnea (shortness of breath) persists despite continuous ventilator use and vigorous assisted coughing, clinical dehydration is suspected, a high fever, or lethargy occurs.

**ADDRESS:** John R. Bach, MD, University Hospital, B-239, Department of Physical Medicine and Rehabilitation, 150 Bergen Street, Newark, New Jersey 07103 (973/972-4393, 973/982-5725 FAX, or e-mail: bachjr@umdnj.edu).

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## Calendar

### — 1997 —

**September 12.** *"Ventilator Dependency: Up Close and Personal,"* Cuyahoga Community College, Metropolitan Campus, Cleveland, Ohio. Contact: National Association for Ventilator-Dependent Individuals (NAVDI), P.O. Box 341, Medina, Ohio 44258. (216/778-3612 or 216/791-6362, e-mail: mail@knowledgelinc.com). Proceedings available later on the Internet at <http://www.knowledgelinc.com>.

### — 1998 —

**April 3-9.** *VACC Camp.* Contact: Cathy Klein, Ventilator Assisted Children's Center (VACC), 3200 SW 60th Court, Suite 203, Miami, Florida 33155-4076 (305/662-VACC). Deadline for applications is December 31, 1997.

**June 25-28.** *CCHS Family Conference.* Holiday Inn Select (Vanderbilt), Nashville, Tennessee. Contact: Mary Vanderlaan, 71 Maple Street, Oneonta, New York 13820 (607/432-8872 or e-mail: VanderlaanM@hartwick.edu).

### DEADLINE ...

for submission of articles, stories,  
information, etc. for the Spring 1998 issue  
of *I.V.U.N. News* is  
March 1, 1998.

Please mail, FAX, or e-mail:

*I.V.U.N. News*  
4207 Lindell Boulevard, #110  
Saint Louis, Missouri 63108-2915 U.S.A.  
314/534-5070 FAX  
e-mail: [gini\\_intl@msn.com](mailto:gini_intl@msn.com)

**THE I.V.U.N. RESOURCE DIRECTORY 1997** will be published in a separate format beginning this fall. In addition to the usual listings of health professionals and ventilator users expert in and committed to home mechanical ventilation, the *Directory* will also include ventilator equipment manufacturers, mask manufacturers, and non-profit organizations with a ventilator link.

Current *I.V.U.N. News* readers should watch for their copy later this fall. The *Directory* will also be available for \$4 postpaid. Please remit to G.I.N.I., 4207 Lindell Boulevard, #110, Saint Louis, Missouri 63108-2915.