

Arriving at Correct Diagnosis, Treatment: A Long and Frustrating Process

More than a year ago, after strenuous exertion, I became very short of breath and dizzy, and realized that I had gone into atrial fibrillation (A-Fib). In the emergency room, the cardiologist focused on my being 61 as the cause. I had a normal lipid panel and a normal stress test. I reviewed A-Fib causes online and found that sleep apnea was a possible underlying cause and that post-polio syndrome (PPS) was associated with sleep hypoventilation. (I contracted polio at age 4 in 1953.) I asked the cardiologist for a pulmonary consult. He flatly refused, telling me to accept that I was just getting older.

This first-person account was written by a retired physician who, for personal reasons, prefers to remain anonymous. The author spent a year in a rehabilitation hospital after contracting polio at age 4 and went on to become an accomplished skier until the late effects of polio curtailed active participation in outdoor sports.

This account highlights the struggle some people with neuromuscular conditions face in finding appropriate consultation and treatment, particularly if they do not live near a major medical center.

Over the next five months of multiple meds, the A-Fib became more frequent and was always very symptomatic. I was dizzy and short of breath even walking 20 feet. Episodes would last for an hour or two. At every cardiologist and electrophysiologist visit, I explained the polio/sleep hypoventilation concerns and requested a pulmonary consult. They both refused repeatedly.

After four months of paroxysmal A-Fib, I had to quit my job. I obtained my records and test results and noticed that my ultrasound indicated severe pulmonary hypertension and blood count showed high hematocrit of 46. I have never smoked, rarely drank, never used drugs, did not have COPD, lived at only 800 feet altitude and had become sedentary because of the A-Fib. So there was no reason for such a high hematocrit. High hematocrits correlate with hypoxia (oxygen deprivation), again suggesting sleep hypoxia.

Seeing the worrisome ultrasounds indicating severe pulmonary hypertension, I contacted the local medical

school pulmonary hypertension clinic nurse. She took my polio and pulmonary concerns to her pulmonologist. He understood and set up pulmonary function testing.

A full six months after the start of A-Fib, the medical college pulmonologist discovered that while sleeping I had 97 minutes below 88 percent oxygen saturation, with hypoventilation, obstructive sleep apnea. The official diagnosis is now chronic respiratory failure, neuromuscular type. He sent me to a neurologist who did EMG/NCV (tests that record the electrical activity of muscles and nerves) and confirmed the extent of my prior polio.

I have new weakness in muscles not previously affected as well as new breathing and probably swallowing concerns. She said the left leg collapsing despite reasonable muscle bulk was because I had a reduced number of motor neurons going to my muscles. When I used my muscles a lot, their neurotransmitters became depleted more rapidly and would stop working.

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From Around the Network

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

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

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To be sure you receive email updates from PHI and IVUN, set your spam filters to allow messages from info@post-polio.org and info@ventusers.org.

Product News

Vent users interested in changing their interface and/or ventilator should discuss the change with their pulmonologist and home health care provider.

GoLife for Men and GoLife for Women, new nasal pillows from Philips Respironics, conform to the different contours of the male or female face. Featuring soft facial “arms” that hug the cheeks, the pillows maintain a secure seal, even during movement. GoLife for Women includes petite, small and medium cushion sizes. GoLife for Men has small, medium and large cushions included.

www.healthcare.philips.com/us_en/homehealth/sleep/golife/index.wpd



Newport Medical Instruments, Inc., the manufacturer of the HT50® and HT70® ventilators, was acquired by Covidien (which had previously acquired Puritan Bennett) in late March. The acquisition is expected to be completed in June 2012.
www.ventilators.com; www.covidien.com

Discontinued vents reminder. PLV®-100 vents have been discontinued but units still in use will be serviced through December 31, 2014. Philips Respironics is transitioning PLV users to the Trilogy100. <http://trilogy100.respironics.com>
 Puritan Bennett™ Legendair vents, also discontinued, will be serviced until May 2015. Legendair users are being transitioned to the Puritan Bennett™ 560.
www.covidien.com/rms/pages.aspx?page=OurBrands/PuritanBennett

Cough Devices. Three new units are on the market, but none are available yet in the United States. Comfort Cough™ is manufactured by Seoil Pacific Group in South Korea (www.seoilpacific.co.kr) and anticipates 510K clearance from the FDA in May. Nippy Clearway is manufactured by B & D Electromedical in the United Kingdom (www.nippyventilator.com). The Pulsar is a Siare® Engineering International Group product from Italy (www.siare.it).

Bridging Apps (<http://bridgingapps.org>) is an innovative volunteer web community of parents, therapists, doctors and teachers who seek to share information on ways to use educational/therapy apps on the latest technology devices – iPad, iPhone, iPod, Android and others – to support developmental learning goals for people with disabilities. Components of this forum include app lists, practical how-to information, success stories, community feedback and a review system that identifies skills embedded in particular apps. ▲

Flying While Using a Vent

Joan L. Headley, Executive Director, International Ventilator Users Network, St. Louis, Missouri, info@ventusers.org

What's the problem?

Stories of users of home mechanical ventilation denied access to flights is not an everyday occurrence, but when they are told, the sequence of events makes no sense based on facts and is extremely upsetting to the traveler.

IVUN has provided travel information for many years. (See www.ventusers.org/edu/valnews/topic2.html) But the reaction to 9/11 has resulted in more complications for everyone flying into and out of the United States, particularly vent users.

Earlier this year, IVUN launched a project to gather information about flying with a vent: from users of home mechanical ventilation, from manufacturers and from airlines.

For traveling vent users, we posted a survey on our website at www.ventusers.org/adv/issues.html. It is available in English, Dutch, French and Spanish, and we will be accepting input until June 1. Users of HMV, both 24/7 and nighttime only, are encouraged to complete it. We have recorded data from 35 users (22 English, 6 Dutch, 3 Spanish, 4 French).

The second group we surveyed was the manufacturers of portable ventilators. To date, IVUN received responses from CareFusion, Covidien, GE/Breas, Impact Instrumentation, Newport Medical Instruments, Philips Respironics, Siare Engineering International Group, ResMed and Weinmann. Manufacturers of portable devices who are not listed should contact IVUN at info@ventusers.org so that their companies' information is included in the final report.

We also contacted numerous airlines regarding their policies on

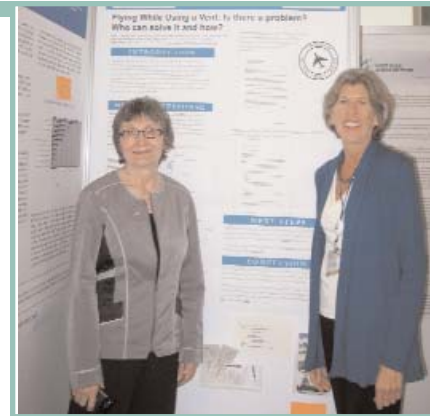
using a ventilator during flight. To date, cooperation in providing information has been minimal at best.

Is there a solution?

Maryze Schoneveld van der Linde, a ventilator user from the Netherlands, is assisting IVUN with this international advocacy effort. She is actively advocating with the European Parliament to improve the rights of air passengers with disabilities.

At the recent international conference on HMV in Barcelona, Bernard Gaudon spoke about his experiences in traveling with a ventilator. He will post his entertaining presentation on <http://bgaudon.tetraconcept.com>, a site on which he tells his story as a polio survivor and ventilator user.

Also in Barcelona, Dr. João Carlos Winck, São João Hospital, Portugal, gave an informative talk entitled "Air travel and respiratory failure." He asked if it is safe for patients with respiratory disorders to fly, discussing concerns for people with COPD, spinal cord injury, obesity hypoventilation syndrome and neuromuscular conditions. He discussed the reasons for oxy-



IVUN Executive Director Joan L. Headley, left, and IVUN Information Specialist Judith R. Fischer with the IVUN poster on "Flying While Using a Ventilator" at the International Conference on Home Mechanical Ventilation held recently in Barcelona, Spain.

gen use and ventilator support during flight.

Dr. Winck also referenced the timely and instructive "Managing passengers with stable respiratory disease planning air travel: British Thoracic Society recommendations," developed by the Society's Air Travel Working Group. The revised version was published September 2011 (Volume 66 Supplement 1 of *Thorax*.) The guidelines may be downloaded at www.brit-thoracic.org.uk/guidelines/air-travel-guideline.aspx.

Final report

IVUN will continue to gather information, and we encourage all members to visit the website and complete the survey by June 1. We will prepare a final report and fact sheets that will be posted on its website – www.ventusers.org in the fall of 2012.

Recognizing the fact that flying isn't the only way to travel, IVUN will create an online searchable database for all ventilator users to report any kind of travel problem or success. We will notify IVUN Members when it is available for input and searching. ▲

Notes from 13th International Conference on Home Mechanical Ventilation (JIVD) and 4th European Respiratory Care Association (ERCA) Conference

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

More than 1,300 respiratory care physicians and respiratory therapists gathered for a joint conference in Barcelona in March. Program planners Joan Escarrabill Sanglas, MD, Patrick Léger, MD, and Dominique Robert, MD, presented a full schedule of panels and practical sessions led by the top experts in home mechanical ventilation (HMV) for both children and adults.

A recurring theme was the increasing demand for HMV (due in part to better vents and masks for children). However, fewer financial resources and increasing health costs in depressed economies worldwide mean that even countries with universal health care are cutting back on services for vent users in the home.

People with obesity hypoventilation syndrome are a growing population of vent users, not only in the United States, but around the world, and several panels discussed how to diagnose sleep and breathing problems, how to ventilate them during surgeries, such as gastric bypass, and which is the best vent and best interface.

Transcutaneous monitoring of carbon dioxide levels (or $tcPCO_2$) is favored by many European physicians to monitor high CO_2 and underventilation in people with chronic respiratory failure. Arterial blood gases (ABGs) and overnight oximetry are useful tools to a point, but ABGs only measure one point in time and do not reflect high CO_2 overnight.

Physicians from Chile, Slovenia, Serbia and Poland described setting up home ventilation centers in their countries. The most recent program is at Benghazi Medical Center in Libya.

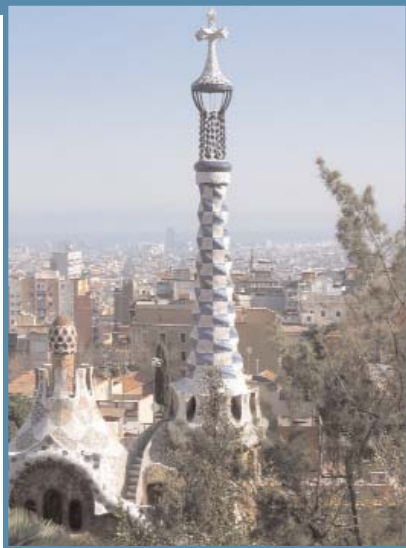
The session on interfaces provided a key finding: Skin problems develop from movement of the mask, not from pressure or tightness of the mask.

Several panels focused on the different disorders, development and physiology of infants and children and their challenges as vent users for physicians. *Guidelines for the respiratory management of children with neuromuscular weakness* is due in spring 2012 from the British Thoracic Society.

Discussions and topics for more than 100 posters included how, when and where to initiate noninvasive ventilation (NIV) in ALS, COPD, etc.; caregiver training and programs to keep vent users at home; use of pressure-targeted or volume-targeted ventilators; recent guidelines on HMV from Australia, Canada and Germany; tracheostomy ventilation; diaphragmatic pacing; and use of NIV in the ICU and palliative and end-of-life care. (See story on IVUN's poster "Flying While Using a Vent" on p. 3.)

Ventilator equipment manufacturers were represented by their European distributors, and several new assisted coughing units were on display. (See From Around the Network on p. 2.)

Special thanks and acknowledgment are owed JIVD's incomparable Brigitte Hautier for ensuring that the conference ran smoothly. www.jivd-france.com ▲



Barcelona is a showplace for Gaudi architecture, such as Park Guell.



JIVD's Brigitte Hautier kept the conference running smoothly.

Conference DVD with sessions, audio and slides, (#1201J) are available for €105 (\$138 USD) from OneScience, 47, rue Marcel Dassault, F-92100 Boulogne Billancourt, France; +33 1 69 30 79 52 fax; www.e-onescience.org.

Ask The Experts

QUESTION: Could your experts please clarify who establishes the settings on a ventilator for a person with a neuromuscular condition?

ANSWER: Diana Guth, BA, RRT, Home Respiratory Care, Los Angeles, California, Diana@hrcsleep.com

In the United States, the physician is supposed to write the order for the settings, but there is the option of writing an open-ended order with setting ranges. This is for companies like ours who have knowledgeable RTs who work with the patients to achieve both comfort and effectiveness. We inform the prescribing physician what the settings are after we set them. Some companies will ONLY accept orders with specific settings. This is very problematic for the proper care of the patients. The physicians are only making educated guesses. The patients' conditions change so the settings need to be adjusted, especially in patients with progressive neuromuscular disorders such as ALS. ▲

Are you a ventilator user or health professional with a question about home mechanical ventilation?

Send it to info@ventusers.org, and IVUN will find experts to answer it.

CONFERENCE CALENDAR THROUGH DECEMBER 2012

MAY 10-12. Spring FOCUS on Respiratory Care, Sleep Medicine and Critical Care Nursing Conference.
Gaylord Opryland Hotel, Nashville, Tennessee. www.foocus.com

MAY 18-23. American Thoracic Society International Conference. San Francisco, California.
www.thoracic.org

JUNE 21-24. FSMA Family and Professionals Annual Conference. Bloomington, Minnesota.
www.fsma.org

JUNE 28–JULY 1. Parent Project Muscular Dystrophy 2012 Connect Conference.
Harbor Beach Marriott, Ft. Lauderdale, Florida. www.parentprojectmd.org

SEPTEMBER 1-5. European Respiratory Society Annual Congress. Vienna, Austria.
www.ersnet.org

OCTOBER 3-5. Fall FOCUS on Respiratory Care, Sleep Medicine, and Critical Care Nursing Conference.
Bally's Hotel & Casino, Las Vegas, Nevada. www.foocus.com

OCTOBER 15-18. Medtrade: Connecting the HME Industry. Georgia World Congress Center, Atlanta, Georgia. www.medtrade.com

OCTOBER 20-25. CHEST, American College of Chest Physicians Annual Conference.
Atlanta, Georgia. www.chestnet.org

NOVEMBER 10-13. AARC International Respiratory Congress. New Orleans, Louisiana.
www.aarc.org

DECEMBER 1-7. International Symposium on ALS/MND. Sheraton Chicago, Chicago, Illinois.
International Alliance of ALS/MND Associations. www.alsmndalliance.org.
Hosted by Les Turner ALS Foundation, www.lesturnerals.org ▲

A Long and Frustrating Process

continued from page 1

She told me to take time to sit down and rest during the day to replenish my neurotransmitters. She is right. Even a five- or 10-minute rest at a store or mall gives me much added strength.

The pulmonologist sent me to an electrophysiologist who did an excellent cardiac ablation (a procedure that cauterizes an area of heart muscle) that has kept me A-Fib free so far. Post-ablation, I started on food supplements and a fitness program which doubled my strength in three months. I can now do ordinary tasks easily again, like getting up from chairs, going up stairs and closing the hatch on my SUV.

After six months I went off warfarin (an anti-coagulant), and after seven months, I went off dronedarone (cardiac arrhythmia medication).

A cardiac catheterization done at the same time as the ablation showed that the pulmonary hypertension was either nonexistent or mild. The high-risk anesthesia team pulled me smoothly through that eight-hour procedure. Post-op, my X-ray showed obvious atelectasis (lung collapse), and I said it's time to start CPAP. So I stayed on the hospital CPAP 24/7 until discharge.

The evening of discharge, they brought CPAP to my home. It was a ResMed AutoSet with an H5i heated humidifier, and a ResMed Mirage Quattro full face mask, size medium. As I became used to this CPAP, I noticed that one to three times every night I would awaken from a deep sleep with the CPAP putting out its top pressure of 20, and the mask slapping my face hard with pressure leaks into my eyes.

I kept tightening the mask but developed a worrisome pressure sore on my nose. I bought a ResMed Gecko gel nasal pad, size medium, which

helped considerably. But I developed unilateral diplopia (seeing double) in both eyes and wondered if it was from excessive mask pressure and the large gel pad. No one knew. I bought a smaller mask, and I also bought a second Gecko nose pad, size small. The diplopia became milder but is still present.

Even with the smaller mask strapped on tightly, on CPAP every night I would be awakened by the pressure at 20 burping out of the mask which kept slapping my face. With the mask super tight, I would awaken at the top of a breath, and it felt like I was trying to breathe out into an already full balloon pushing back at me.

Neither the pulmonary nurse nor the CPAP nurse understood the problem. So it was back to online searching. Several websites suggested that people with PPS needed BiPAP. I contacted IVUN describing my mask pressure problem, and the information IVUN provided seemed to indicate that I needed BiPAP.

The pulmonary hypertension pulmonologist understood immediately. He referred me to the sleep specialist pulmonologist who handles neuromuscular ventilation needs. She did some more testing that qualified me for BiPAP. My insurance gave me a Philips Respironics BiPAP AVAPS. She set this on variable IPAP 4 to 25, EPAP 4, backup breathing at 8, tidal volume at 450 ml. I tried different rise times and decided the 1 setting felt best.

After almost a year, I use the BiPAP every night and do not really notice it any more. It feels easier to breathe with it on. I still use a tightly fitting ResMed Mirage Quattro full face mask, size small, with the ResMed Gecko gel nose pad.

Recently, I developed a gray spot in my central vision and an arc of flash-



ing lights at the outer edge. The ophthalmologist found a posterior vitreous detachment, but he did not know of any association to the mask pressure on my eyes. He did feel that the previous CPAP mask high-pressure air leaks caused dry eyes that were probably the cause of the unilateral diplopia and suggested over-the-counter artificial tears four times a day.

Now that I have become so aware of my breathing, I realize that when relaxed during the day, I do not breathe well; I go into a fog and then fall asleep. In retrospect, this has been happening for about two years. I have discovered that during the fog, I can pull myself out of this. With 10 to 15 deep breaths, I become totally alert again. Also, actively rocking in a rocker while watching TV completely stops this from happening. I would be interested to hear if anyone else in the neuromuscular respiratory failure group has had experience with this and has any suggestions. If so, please contact info@ventusers.org. ▲

NEW TO IVUN?

This is our 26th year of publishing a newsletter.

Past issues may be found at: International Ventilator Users Network (IVUN), www.polioplace.org/history/collection/international-ventilator-users-network-ivun

Join IVUN!

...online at shop.post-polio.org and receive *Ventilator-Assisted Living*.

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

\$30 Subscriber – Bi-monthly *Ventilator-Assisted Living* and *IVUN Membership Memo* (both delivered electronically).

Yes, I want post-polio news, too.

\$55 Subscriber Plus – *Ventilator-Assisted Living* (bi-monthly; electronic) AND *Post-Polio Health* (quarterly; print)

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- Person of your choice (include name and address) or
- Person who has expressed financial need to IVUN.



[CLICK HERE TO LEARN WHY](#)

Smartphones? iPads? Tablets? Do you use any of this technology in connection with your vent therapy? If you could somehow 'connect' all of your caregiver communications, prescription information, progress reports, sleep logs – anything connected with your vent therapy – through one of these devices, would you use it? Please take a minute to complete a short survey. The results will be shared in a subsequent distribution of this newsletter. Thanks.

[Please click here to begin.](#)

RESEARCH REMINDER

The Research Fund of IVUN is accepting proposals for a \$25,000 grant to be distributed in 2013.

The deadline for Phase I application is **May 4, 2012.**

The criteria are posted at Research Fund Call for Proposals for 2012. Funds may be used for a small exploratory study or to complete an ongoing project or they may be combined with other grant funding to complete a larger project.

If you have any questions, please email info@ventusers.org or call 314-534-0475.

Yes, I want to support IVUN's mission of education, research, advocacy and networking and its comprehensive website, www.ventusers.org.

Membership at the following levels includes ALL benefits PLUS special recognition in IVUN publications:

- \$250 Bronze Level Sustainer**
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