



PH Resource Network

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This newsletter, compiled by the Education Committee of PH Resource Network, is intended to provide a professional forum to share, learn, network and enhance our practice and understanding of PH.

The information provided in the newsletter is not an endorsement of any particular method or form of treatment but a "sharing" of information that may help you in your PH plan of care and broaden your understanding of clinical situations.

LUNG TRANSPLANTATION FOR PULMONARY HYPERTENSION

by Monica Horn, R.N., CCTC

Lung transplant candidates are people with end-stage lung disease for which there is no other medical therapy available or the medical therapy has been ineffective for them. PH patients have more medical therapies available currently than ever before. Some physicians believe, however, that lung transplantation should be discussed with every newly diagnosed PH patient, as back-up intervention in the event of medical treatment failure. Predicting the proper time for transplant referral of a PH patient can be difficult.

Lung transplantation has been performed for more than 40 years with most of the successful cases having been performed since the 1980s. Advances in surgical techniques and development of improved immunosuppression have improved survival rates. Although survival rates vary slightly from center to center and also with other variables such as the age of the patient, lung transplant recipients have a survival rate of 83-92% in the first year.¹ Survival may decrease by a small percentage—3%-5% per year—due to infection or rejection. There may also be complications of side effects of immu-

nosuppression such as post transplant lymphoproliferative disorder (PTLD), a lymphoid proliferation seen in 1-20% of solid organ transplant recipients that usually responds to reduction of immunosuppression if detected early.²

Prior to acceptance for listing, transplant candidates must be evaluated for any preexisting condition that would decrease chances of a successful outcome. These conditions might include other organ failure, current infection, current or recent cancer, current or recent substance abuse/dependence, psychiatric conditions, or history of non-adherence to medical instructions.

The type of lung transplant recommended for a PH patient may be determined by consideration of the patient's PH diagnosis. If the patient has congenital heart disease or a failing heart, a heart-lung transplant may be recommended. Due to relatively low survival rates of heart-lung transplantation, many transplant centers recommend lung transplantation while the heart is still functional. That is, lung transplant may be considered for



that time frame of disease progression when cardiac function is fairly well preserved. That may be a window of opportunity that should be carefully followed and assessed.

Depending on the condition and age of the lung transplant candidate, either a single lung transplant or a bilateral lung transplant may be preferred. Generally, pediatric patients are given bilateral lung grafts to promote normal growth and activity.

Donors may be deceased/brain-dead donors or living lobar lung donors. There has been some experience with non-heart beating donors more recently. Deceased lung donors' lungs are transplanted separately in the anatomically correct side of the thorax. Lungs or lung lobes need to be healthy, the right size, and of a compatible blood type. Lung lobe donors are adult volunteers that are usually family members or close personal friends. In the living donor lung transplantation procedure, two healthy adults donate the lobes with one right lower lung lobe and one left lower lung lobe being utilized.

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UNOS, the United Network for Organ Sharing, is a private organization under contract by the federal government in the United States to oversee all solid organ allocation in this country. All approved transplant centers and OPOs (organ procurement organizations) must adhere to the strict standards mandated by this organization. Centers and OPOs enter data for the organization's database from which standards of practice, survival, and other quality indicators are tracked. Patients or the general public may access data on the website www.ustransplant.org.

The LAS (Lung Allocation Score) system, a UNOS tool utilized since 2005, is used to determine if candidates 12 years of age and older meet criteria for more urgent donation. Parameters such as age, size, diagnosis, oxygen requirements, six-minute walk distance, mechanical ventilation, pulmonary artery pressures, pulmonary function values and functional class are entered and the system calculator determines a score. The higher the score, the higher the priority the patient has for donor offers.

Medical centers who evaluate PH lung transplant candidates, are challenged with advising referred candidates about successful timing for lung transplant listing. Prediction of donor availability for patients with advanced disease whose medical therapy is failing is difficult. The International Society for Heart and Lung Transplantation (ISHLT) guidelines for pulmonary arterial hypertension patients' referral and transplantation include consideration of functional class of III or IV with rapidly progressive disease and declining stability as measured by decreasing 6-minute walk test, decreasing cardiac index, increasing

right atrial pressure and functional class not improving/declining on maximal therapy.³ Although candidates are individually considered, many transplant physicians state that early referral is key given the challenges of donor availability.

Data regarding lung candidate waiting time is published regularly by the Organ Procurement and Transplantation Network (OPTN). All transplant centers in the United States enter information in this database. Median waiting times for lung transplant candidates listed in 2003-2004 ranged from 445-680 days.⁴ There were 2,029 lung candidates waiting for organs in the United States as of 12/22/2008.⁵ There were 1,042 lung donors in 2008.⁶ These numbers illustrate the scarcity of lung donors in comparison to lung transplant candidates that are waiting.

Some of the interventions that have been developed to address these challenges include utilizing an appeal process with the UNOS LAS system.⁷ This allows transplant centers to appeal to a UNOS review board comprised of transplant medical professionals, to essentially plead the case of their patient with valid clinical information to support a higher score representing a greater need for more rapid transplantation.

Living donor organs can also be used. Since two healthy adult donors must donate lobes, it may not always be realistic to have volunteers available who are able to qualify for the procedure. However, transplant centers that perform living donor lobar lung transplants have lobar lung transplant patient survival rates comparable to, or slightly better than, the deceased donor transplants with few reported

donor complications.⁸ This slight difference may be due to the lobes being donated by healthy individuals without the inflammatory mechanisms of brain death physiology and trauma associated with the cause of death of a deceased donor.⁹ Of course, early aggressive medical treatment following current guidelines and continuation of maximal medical support is vital in contributing to stability during the waitlist period.

The listed transplant candidate should have the transplant team and PH therapy team, if at separate institutions, continue close communication during the waitlisted period. The transplant team should be aware of the PH patient's current measures of therapy effectiveness such as changes in functional class, decreasing 6-minute walk, and changes in right ventricular function. If these parameters are changing, the transplant team may be able to advocate for an appeal to the waitlist score.

When lung transplantation is successful, patients describe an improved quality of life.⁹ As with other therapies, there are medication side effects and complications making it imperative that there be careful medical follow-up. Lung transplantation, however, remains a valuable option for selected PH patients who have maximized their drug therapy, yet begin or continue to deteriorate. Early referral is essential to allow for preserved or recoverable right ventricular function as well as a possible prolonged wait for donor organs. ☪

*-by Monica Horn, RN, CCTC
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Meet the Author



Monica V. Horn, RN, CCTC

Monica has been a registered nurse for 36 years, having worked with pediatric patients at Children's Hospital Los Angeles (CHLA) for the last 27 years. After working with medical-surgical adults, an obstetrics office, and a large pediatric hospital's tracheostomy unit/ICU in the Midwest early in her nursing career, she relocated with her husband to California in 1981. A PICU/CTICU nurse at CHLA since 1981, she took the position of Cardiothoracic Transplant Coordinator in January of 1994, and began working with children who needed/received heart, heart-lung, or lung transplants. When her transplant team started seeing IV epoprostenol patients who were on the lung transplant list, shortly after taking the transplant coordinator position, she also became the pulmonary hypertension coordinator. She has authored or co-authored more than 21 published journal articles and 48 abstract presentations since 1996.

Over 850 cardiothoracic surgery cases are performed yearly at Children's Hospital Los Angeles, an international leader in pediatrics and teaching hospital affiliated with the Keck School of Medicine of the University of Southern California. Drs. Vaughn Starnes and Cynthia Herrington head the Cardiothoracic Transplant surgical team. Drs. Jacqueline Szmuszkovicz, JD Mentee, Marlyn Woo, and Eithne McLaughlin provide medical support for pre-transplant and/or post-transplant Pulmonary Hypertension patient management.

Name Our Newsletter!

The Education Committee would like to give an innovative name to this newsletter, representing the dedication and spirit of members of the PH Resource Network. A few suggested titles are listed below. Pick one, and send your vote to PHRN@PHAssociation.org by **March 1st**. The next issue of the newsletter will come out in Spring 2009, sporting a fresh new name!

Choose one:

- “Connections”
- “esource”
- “PH On The Horizon”
- “phsource”
- “The Network Beat”
- “The Network Pulse”
- “The Network Source”
- “The PH Pulse”
- “The View”

Question Corner

Several of my PH patients seem depressed. What resources are available to these patients?



Connect your patients
to the PH community!

PH Toll Free Helpline

1-800-748-7274

Local Support Groups
and Online Message
Boards

[www.PHAssociation.org/
connect](http://www.PHAssociation.org/connect)

Depression due to chronic illness is very common. Pulmonary hypertension is a progressive chronic illness without a cure. Many patients have been faced with misdiagnosis or an extended diagnostic evaluation prior to being diagnosed with pulmonary arterial hypertension. Because initial PH symptoms are vague, many patients are told that they are deconditioned or that their symptoms are part of the normal aging process. Once diagnosed with PH, patients have often researched the internet for more information while awaiting an evaluation at a PH center or awaiting a right heart catheterization. Prognosis from community caregivers and the internet is often discouraging, as it is usually based on outdated information that may not take into account advancements in PH therapy.

Patients are faced with concerns of managing an active lifestyle with symptoms that limit their ability to complete tasks of daily living. Forced to conserve energy, ask for help, and space out activities, it is necessary for

patients to find coping strategies to adapt to their illness. Emotional impacts of evaluating self worth, caring for self and family, and dealing with insurance and financial issues related to chronic illness further contribute to the possibility of developing depression or anxiety disorders.



Counseling and evaluations for medical management of depression or anxiety disorders should be offered to patients with PH. Simple screening tools can be utilized to help identify patients who are at higher risk. Discussions with patients and family caregivers during clinic appointments should include evaluating loss of interest in meaningful aspects of life, difficulty sleeping, changes in eating patterns and emotional distress. Communication with primary care providers to moni-

tor for emotional disorders during ongoing general health visits can be helpful.

Encouraging patient involvement in PH support groups is another resource allowing patients to share coping strategies. Allowing patients to share concerns about adapting to chronic illness and share creative ways to adapt can help those with a newer diagnosis or inadequate social support. As PH providers and nurse clinicians, offering support and encouragement to those who are concerned about prognosis and treatment options is a critical part of ongoing communication. Allowing time for questions and exploring individualized treatment options provides patients control in their health decisions. Actively encouraging evaluation for emotional disorders such as depression or anxiety disorders can help patients with PH have improved quality of life. ∞

*by Traci Stewart, RN, MSN
University of Iowa
Iowa City, IA*

Have a question that you would like an expert opinion on?

Chances are another PH provider may have the very same question. This section is designed to obtain answers for you from the experts in our field. Each issue will feature answers to one or two questions that have been submitted. Submit your question to PHRN@PHAssociation.org.

Speakers Bureau: Lend Us Your Expertise

The establishment of a Speakers Bureau was an idea that sprang from PHA in response to inquiries from various organizations for persons to contact who could speak about pulmonary hypertension. The Education Committee took on this project last year, and is now ready to put it into service.

The purpose of the Speakers Bureau is to provide a list of PH professionals available to speak at various engagements such as support groups or other organizations interested in pulmonary hypertension. The Speakers Bureau list will be accessible from the PHA website. This is a voluntary program; there is no monetary compensation for presenters.

Speakers are needed in all disciplines within the Pulmonary Hypertension Resource Network. The Speakers Bureau Program would like to encourage all experienced PH nurses, respiratory therapists, pharmacists, and nutritionists to participate. It is assumed that most speakers will be asked to present within their local community.

The Speakers Bureau Program is structured on two basic components. The first establishes basic qualifications for a person to be accepted on the speaker list (described at right). The second component establishes a means to monitor, maintain and update the speaker list.

Monitoring and maintenance of the Speakers Bureau program is the responsibility of the PH Resource Network Education Committee. The Committee: (1) reviews the qualifications of a speaker prior to addition to the list; (2) updates the list on a biannual basis; and (3) reviews and maintains speaker evaluation forms. A group or

organization that uses a presenter from the speakers list will be asked to complete a simple evaluation form to be returned to PHA.

Some individuals may feel comfortable talking about a wide range of PH subjects; others may have a specific area of interest or expertise. On the speakers list, an individual may specify what they would be available to talk about. For example: I have a presentation on exercise that I have given to our local support group that I feel would be of benefit to others. I could put my name on the list, specifying that I would be available to present on that specific topic only.

This is the first announcement regarding this new program. The Speakers

Bureau PHA website cannot become active without you! Consider your many talents and come forward to share your experience with patients, families and medical providers.

I would like to extend a special note of thanks and recognition to Wendy Hill, N.P., University of Southern California, for her efforts in taking the lead on this project. We are hopeful that this will be a successful program with potential speakers in all regions of the United States who offer knowledgeable, interesting, and informative presentations. ☺

*by Marilyn Schmidt, RN
BryanLGH Medical Center
Lincoln, NE*

PH Resource Network Speaker Qualifications

- An individual must have one or more of the following degrees: R.T, R.N., C.N.S., N.P., PharmD, or P.A.
- An individual must have a minimum of three years experience as a clinical or research provider, nurse or coordinator caring for and interacting with patients with pulmonary hypertension
- An individual must have knowledge of pulmonary hypertension, diagnosis, management and treatment
- An individual must be a member of PH Resource Network

All information is self-reported by the individual seeking to be included on the speaker list and credentials will not be verified. For this reason, the PHA and the PH Resource Network do not provide any speaker endorsement.

Questions about the Speakers Bureau?

Contact **Marilyn Schmidt**, Education Committee Chair
marilyn@schmidt006.com

To obtain an application:

Contact **Roice Fulton**, PHA Medical Services Program Associate
Roice@PHAssociation.org



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www.PHAssociation.org

The mission of the PH Resource Network is to improve the lives of pulmonary hypertension patients and family members through providing support and education for healthcare professionals by:

- Developing strategies to improve patient care and establishing guidelines for healthcare professionals
- Supporting research opportunities
- Providing opportunities for professional development

The PH Resource Network Education Committee would like to encourage your participation in making this a useful network tool for members. **Suggest an idea** for a topic, **volunteer** to write a feature article, **ask us** a question, **pass along** a great article you have come across or just **give us your feedback** about the newsletter. Submit questions, ideas, comments to Roice Fulton, at Roice@PHAssociation.org. Past issues of the newsletter are available online at www.phassociation.org/PHRN/newsPHRN.asp.

The Education Committee members are: Marilyn Schmidt, RN (chair); Cathy Anderson-Severson, RN, BSN; Shiela Bostelman, MSN, CPNP, CCTC; Aimee Doran, MS, RN, CPNP; Cynthia Fink, RN, BSN; Kimmarie Hammond, ANP-C; Lisa Hartin, RN, BSN; Wendy Hill, NP; Bradford Jones, BSN; Natalie Kitterman, RN; Pamela Rotella, RN; Traci Stewart, RN, MSN.



Save the Date: September 24-26, 2009



The **Pulmonary Hypertension Association**
invites you to attend the **2009 PH Resource Network Symposium**

Leading Progress, Creating Partnerships: Empowering the Interdisciplinary PH Team

September 24 – 26, 2009

Hyatt Regency Crystal City in Arlington, Va.

Registration for this unique event opens March 2009

New for 2009

Dual-Track Skill Level Programming
Research Poster Abstract Session*
Additional CNE, CEU & CPE credits

*Abstract submission guidelines available at
www.PHAssociation.org/PHRN/Symposium/Abstracts.asp

*Thanks to an unrestricted educational grant from Actelion Pharmaceuticals US Inc., the first 300 PH-treating allied healthcare professionals register for **FREE**. PLEASE NOTE: A \$50 deposit is required to reserve a space. This fee will be reimbursed after attendance; however, no-shows are ineligible for reimbursement.*

"It was the best organized, most informative and most useful of any conference I have ever attended in my 22 years of nursing. Thank you for a great experience and helping make my journey of nursing more successful."

~ a 2007 PHRN Symposium attendee

For the latest updates on registration, speakers and topics, visit www.PHAssociation.org/PHRN/Symposium or call PHA at 301-565-3004 x761.