

Lung Transplantation in Patients with Pulmonary Hypertension

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- **Information for Friends and Family** (www.phassociation.org/Patients/LungTransplant/InfoFamilyFriends)
- **Before Lung Transplant** (www.phassociation.org/Patients/LungTransplant/Before)
- **After Lung Transplant** (www.PHAssociation.org/Patients/LungTransplant/After)
- **Join PHA's Transplant Email Group** (www.PHAssociation.org/TransplantGroup)

Lung transplantation is a treatment option for selected patients with pulmonary hypertension (PH) when medical therapy is no longer effective. Lung transplantation can prolong survival and improve quality of life, but some patients have significant complications and may not realize its potential benefits. Although lung transplantation offers a potential cure for PH coupled with improved quality of life, it may lead to shorter survival, on average, compared to other solid organ transplants. Potential complications include organ rejection, infections, side effects of drugs that suppress the immune system and lung transplant recipients need to be followed closely by a transplant center that may not be close to home.

Some lung transplant patients survive ten years or more with a good quality of life; however, some patients survive only a few months or less because of serious complications. Even at the best transplant centers, the survival statistics for lung transplants are lower overall than for kidney, liver or heart transplants. The current lung transplant survival statistics from the International Society for Heart and Lung Transplantation Registry show an overall one-year survival of 74 percent and a five-year survival of 45 percent. Thus, survival is less than five years for more than half of the patients who receive a lung transplant. The major long-term problem is chronic rejection, which results in persistent worsening function of the transplanted lung or lungs. Chronic lung rejection occurs earlier and more frequently in lung transplant recipients as compared to recipients of other transplanted organs.

Unfortunately, the timing of lung transplantation cannot be controlled, even though it is a critical issue for many patients. The majority of donor lungs are obtained from donors who die from accidental injuries, making the availability of donor organs unpredictable. The number of patients who need lung transplants considerably exceeds the number of available donor lungs, and lungs are transplanted according to the recipient's indication for transplant (e.g. pulmonary hypertension, chronic obstructive pulmonary disease, pulmonary fibrosis). A number of other factors are used to calculate the lung allocation score (the current process by which patients are listed to obtain a lung transplant), which reflects disease severity and urgency of transplantation as well as the likelihood that a patient will survive to at least one year following transplantation. Emergency lung transplantation is sometimes possible, but it cannot be done unless the patient has already been evaluated by a transplant center.

The distribution of donated organs in the United States is regulated by the United Network for Organ Sharing, and additional information is available on their website (www.unos.org). Outside of the United States, the process of getting listed for lung transplantation is, in general, much more difficult and the availability of organs is somewhat limited but varies from country to country. Additional information about lung transplantation worldwide is available at the website of the International Society for Heart and Lung Transplantation (www.isHLT.org).

Deciding on the optimal time to be listed for transplantation is often difficult. This is especially true in patients receiving epoprostenol (Flolan) or other intensive PH treatment, although three separate retrospective analyses that included nearly 400 patients with idiopathic PH receiving epoprostenol found that failure to improve clinically or hemodynamically one year after starting therapy was associated with a significantly worse outcome. In general, PH in its early stages does not immediately threaten the life of the patient, so transplantation is not recommended at that time. On the other hand, the possibility of transplantation should be discussed before patients are too ill. When transplantation is considered late in the course of the disease, there is a substantial risk that the patient will not survive until donor lungs are available. The approach followed by many centers is to add a patient (if eligible; see below) to the transplant waiting list at the time he or she initiates epoprostenol or other aggressive therapy. If the patient subsequently improves with treatment, he or she may choose to become inactive on the waiting list. Once listed for transplantation, patients do have the right to transfer their care to another center. It is also possible to be placed on more than one list in different geographic regions.

Transplantation of a single lung works very well for many lung diseases, and many patients with PH have had a single lung transplant with good long-term results. However, nearly all transplant centers currently prefer to transplant both lungs (double-lung transplant) for patients who have PH, in part because there are generally fewer postoperative complications. Transplantation of the heart and both lungs (heart-lung transplant) may be needed in some patients and, in general, waiting time is longer when more than one organ is needed. Living donor lung transplantation (taking a lobe of one lung from a living person) is possible but requires two other healthy individuals, much larger in stature than the recipient, to each donate one lobe of a lung. Thus, living donor transplantation is mainly performed in pediatric patients who receive lung tissue from adult donors. Few living donor transplantations have been performed, and this option is available at only a few centers. At least one donor parent has died due to complications of the donation surgery.

Candidate selection criteria for lung transplant and post-transplant treatment regimens vary from center to center. The majority of centers transplant patients with idiopathic PH and congenital heart disease-related PH. Patients with PH related to diseases that affect more than just the lungs, such as those with scleroderma, are generally evaluated on a case-by-case basis and are not considered for transplantation at some centers. The age cutoff for listing PH patients may be somewhat lower than for patients with other types of end-stage lung disease requiring only single-lung transplantation. Generally, to be considered for double-lung transplant patients must be less than 60 years old and for heart-lung transplant they must be less than 50 years old. Systemic illnesses such as poorly-controlled diabetes with related complications in other organs, significant coronary artery disease or cancer (other than localized skin cancers) most often exclude patients from consideration for lung transplantation at the majority of centers. In addition, a detailed psycho-social evaluation is performed at all centers to determine whether patients are fully committed to lung transplantation and whether there will be adequate social support after transplantation. Support from family or friends is essential for a successful outcome because of the complicated post-transplant care. Patients should therefore inquire about the specific acceptance and rejection criteria at the center performing their evaluation.

Lung transplantation remains the only curative treatment for PH. However, while it may cure PH, it presents a wide range of new challenges for patients. Lung transplantation is most effective for patients who have a positive attitude and are not troubled by the need to take multiple medications daily or to have frequent medical testing.

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This information is for general information only. These guidelines may not apply to your individual situation. You should rely on the information and instructions given specifically to you by your PH specialist and/or the nurses at your PH Center. This information is general in nature and may not apply to your specific situation. It is not intended as legal, medical or other professional advice, and should not be relied upon as a substitute for consultations with qualified professionals who are familiar with your individual needs.