



# Lung transplantation for PAH

K Hoetzenecker MD PhD

Lung Transplant Program Vienna

Department of Thoracic Surgery, Medical University of Vienna

# Disclosures

Consultant for Medtronic (ECMO dual-lumen cannulas)

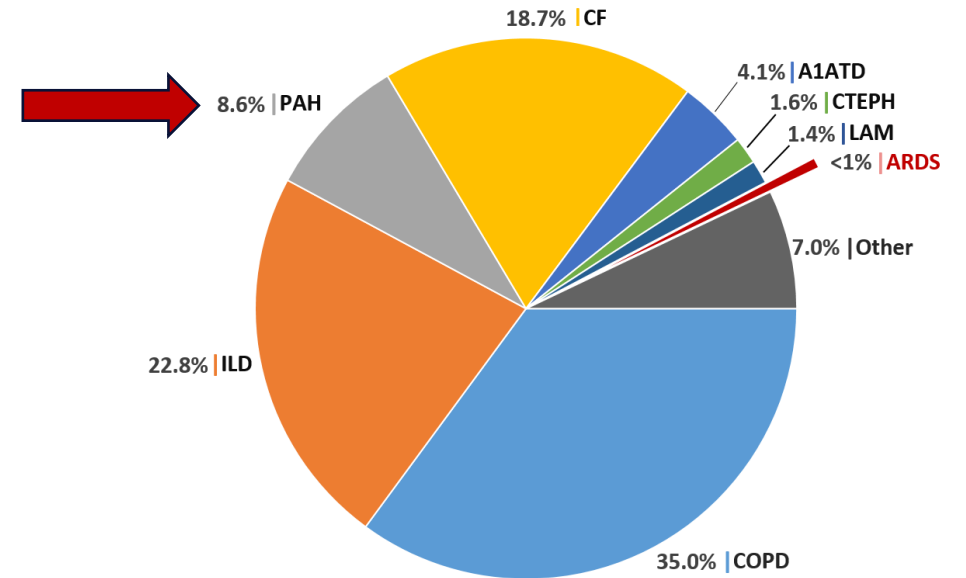
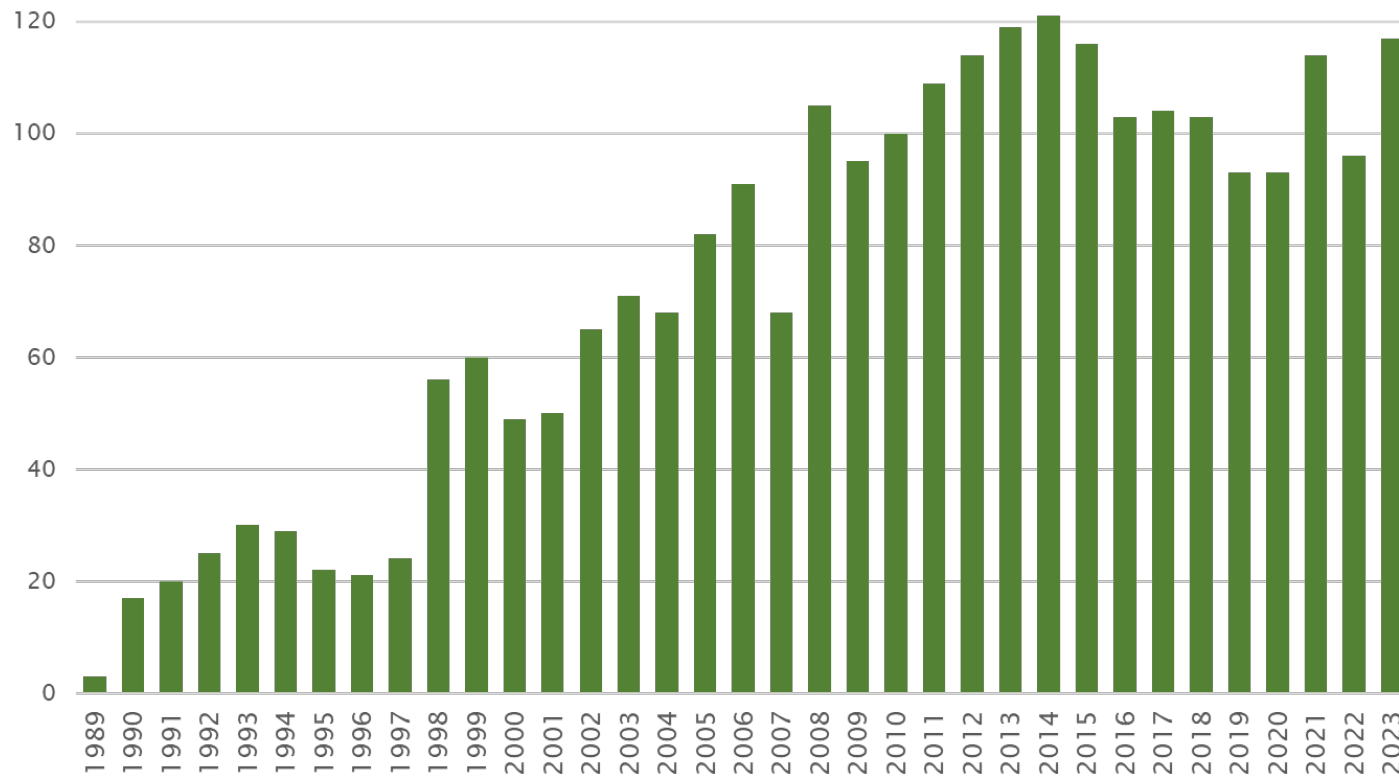
All patients have given their permission to show their photographs

# I will talk about...

- ① Indication and timing for LTx
- ② Choice of procedure (bilateral LTx, HTLx, single lung Tx, etc.)
- ③ Postoperative management/cardiac remodeling

# LTx for PAH is rare...

## Total # of lung transplantations - Vienna n = 2560



# LTx for PAH is rare ... ISHLT registry data (1992-2018)

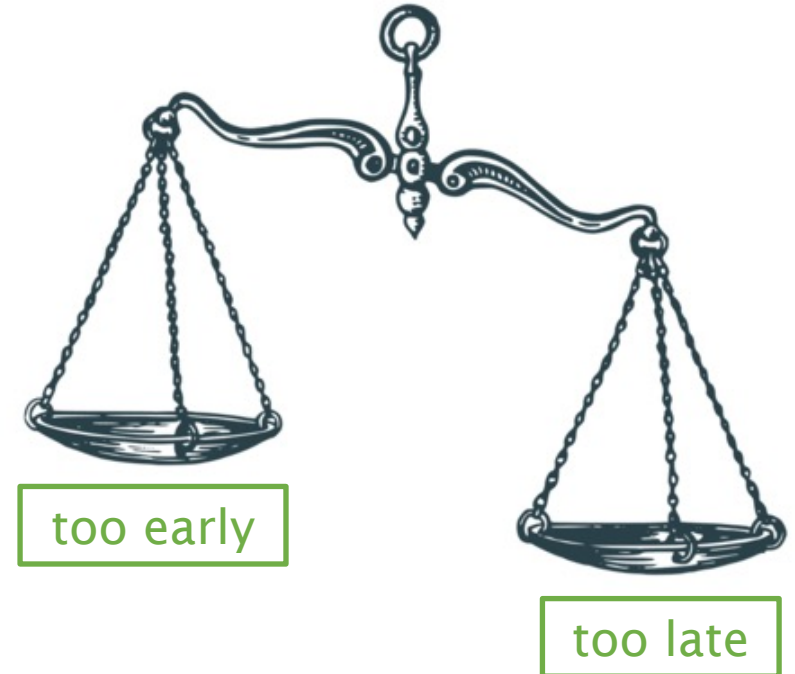


	Jan 1992-Dec 2000 (N = 11,796)	Jan 2001-Dec 2009 (N = 21,806)	Jan 2010-Jun 2018 (N = 33,891)	P-value
<b>Diagnosis</b>				<b>&lt;0.0001</b>
- COPD	4162 (37.3%)	7102 (33.1%)	8917 (26.5%)	
- A1ATD	1169 (10.5%)	1131 (5.3%)	1054 (3.1%)	
- CF	1717 (15.4%)	3470 (16.2%)	4771 (14.2%)	
- IPAH	578 (5.2%)	563 (2.6%)	945 (2.8%)	
- IPF	1677 (15.0%)	4899 (22.8%)	9755 (29.0%)	
- Retransplant	394 (3.5%)	921 (4.3%)	1364 (4.1%)	
- Other	1471 (13.2%)	3392 (15.8%)	6822 (20.3%)	

→ Evidence mostly limited to single-center retrospective analysis

# Indications for Lung Transplantation

- Chronic end-stage lung disease
- Failed medical management
- For most patients the ultimate treatment
  - Trading one medical challenge for another
  - BUT: excellent quality of life possible

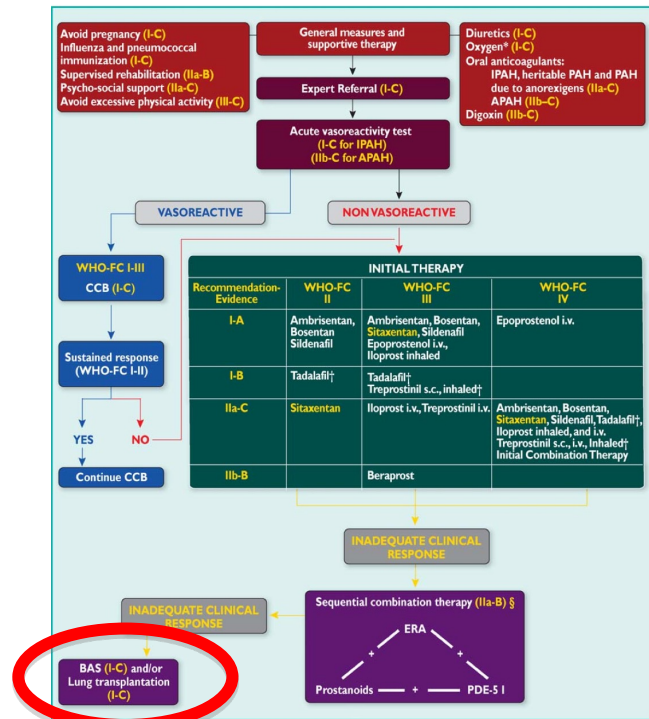


*J Heart Lung Transplant.* 2021 Nov;40(11):1349-1379.

# When is the ideal timepoint to consider Tx in PAH patients?

In case of inadequate clinical response to primary therapy

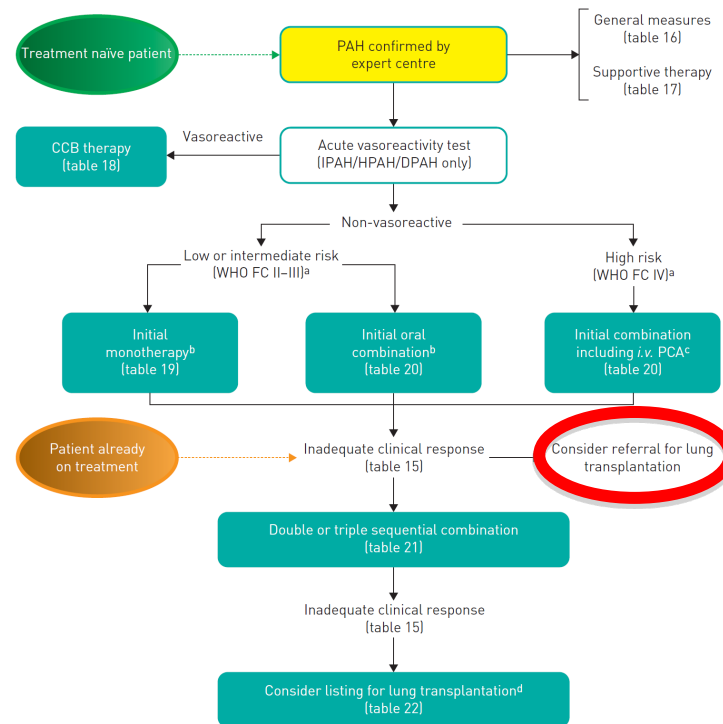
2009



ESC/ERS/ISHLT Guidelines

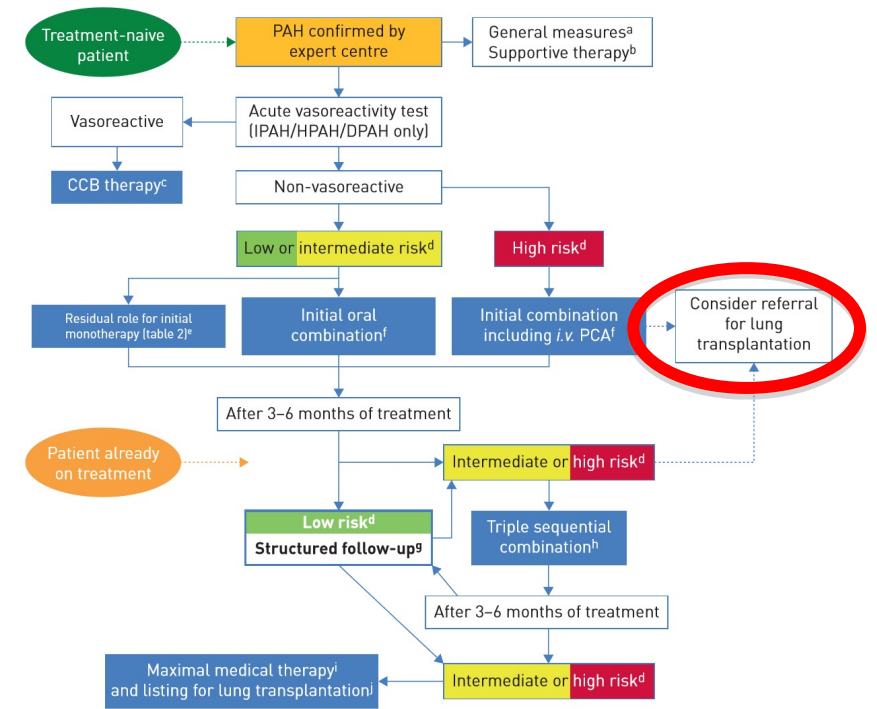
Eur Respir J. 2009 Dec;34(6):1219-63.

2015



Eur Respir J. 2015 Dec;46(6):1855-6.

2019



Eur Respir J. 2019 Jan 24;53(1).

# Problems arising when referring a patient too late



VS.



# Let's transplant - BUT which procedure?



# In the early days of Tx for PAH the heart was considered too damaged → Heart/Lung Tx



The NEW ENGLAND  
JOURNAL of MEDICINE

ORIGINAL ARTICLE

Volume 306:557-564 March 11, 1982 Number 10

## Heart-lung transplantation: successful therapy for patients with pulmonary vascular disease

*BA Reitz, JL Wallwork, SA Hunt, JL Pennock, ME Billingham, PE Oyer, EB Stinson, and NE Shumway*

We report our initial experience with three patients who received heart-lung transplants. The primary immunosuppressive agent used was cyclosporin A, although conventional drugs were also administered. In the first patient, a 45-year-old woman with primary pulmonary hypertension, acute rejection of the transplant was diagnosed 10 and 25 days after surgery but was treated successfully; this patient still had normal exercise tolerance 10 months late. The second patient, a 30-year-old man, underwent transplantation for Eisenmenger's syndrome due to atrial and ventricular septal defects. His graft was not rejected, and his condition was markedly improved eight months after surgery. The third patient, a 29-year-old woman with transposition of the great vessels and associated defects, died four days postoperatively of renal, hepatic, and pulmonary complications. We attribute our success to experience with heart-lung transplantation in primates, to the use of cyclosporin A, and to the anatomic and physiologic advantages of combined heart-lung replacement. We hope that such transplants may ultimately provide an improved outlook for selected terminally ill patients with pulmonary vascular disease and certain other intractable cardiopulmonary disorders.

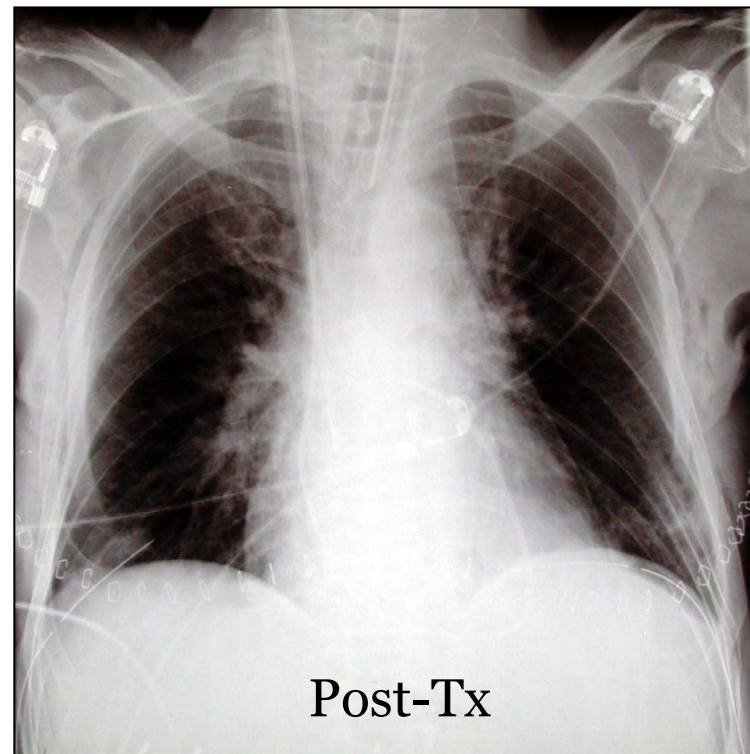
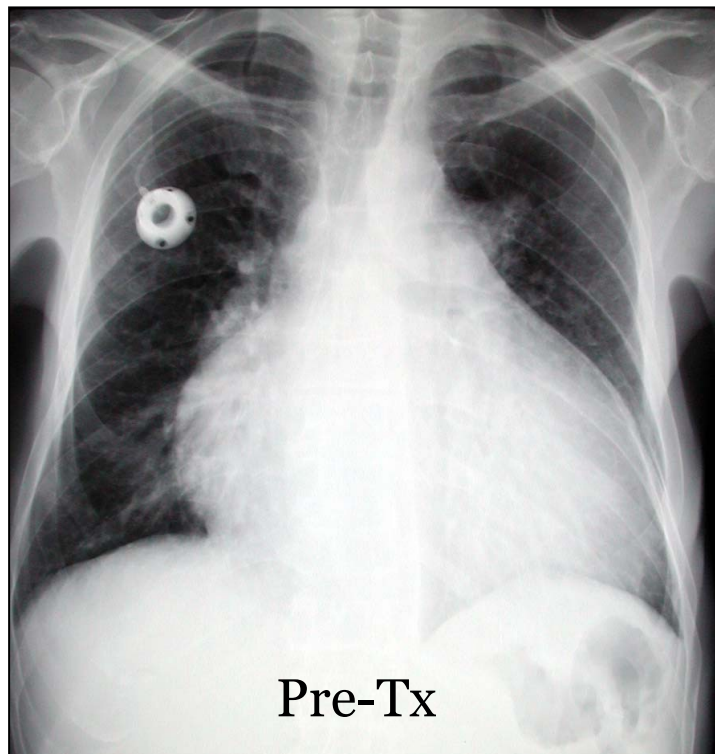


Norman Shumway  
Stanford 1981



Bruce A. Reitz  
Stanford 1981

# In the 1990-2000s - Tx centers started to experiment with Double-Lung Tx for PAH

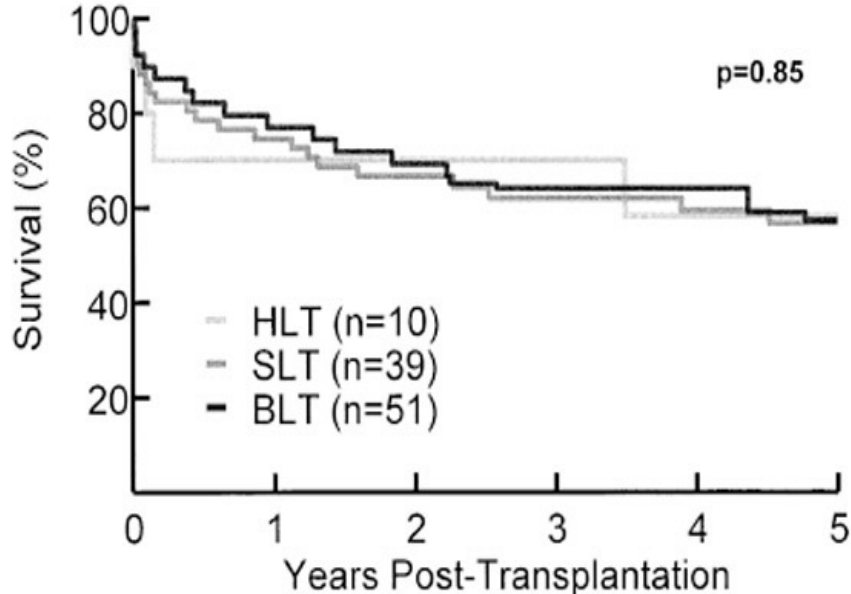


## Reverse cardiac remodelling

→ With the right expertise in postOp care, PAH patients can receive a LuTx (instead of a HLuTx)

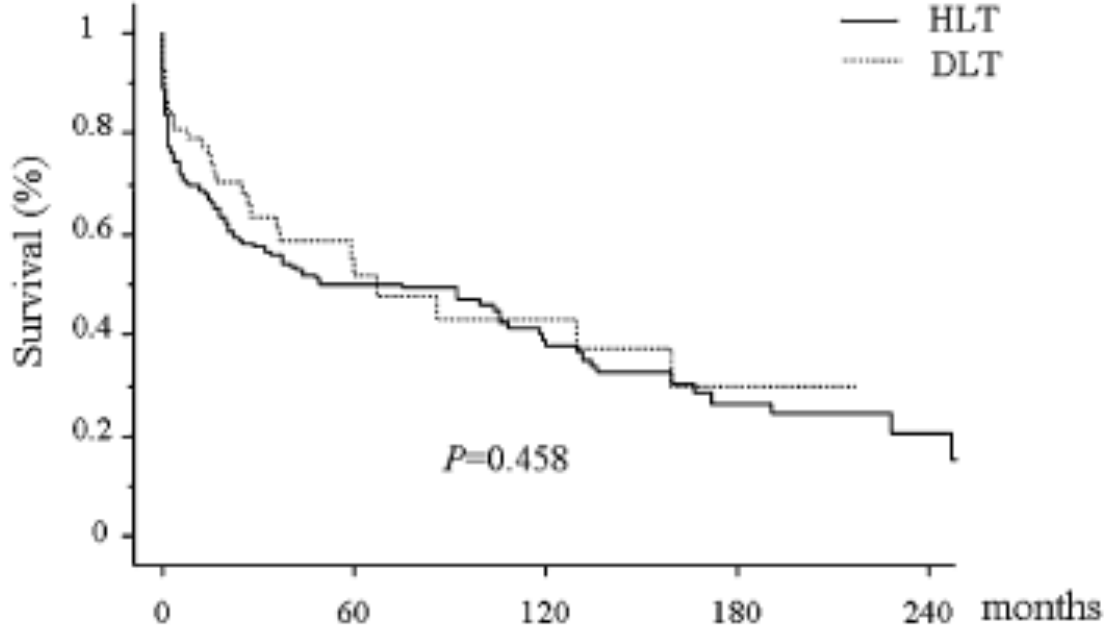
# Survival after Tx for PAH: lung Tx vs heart/lung Tx

St. Louis



Mendeloff EN et al. Ann Thorac Surg 2002;73: 209-19.

Paris

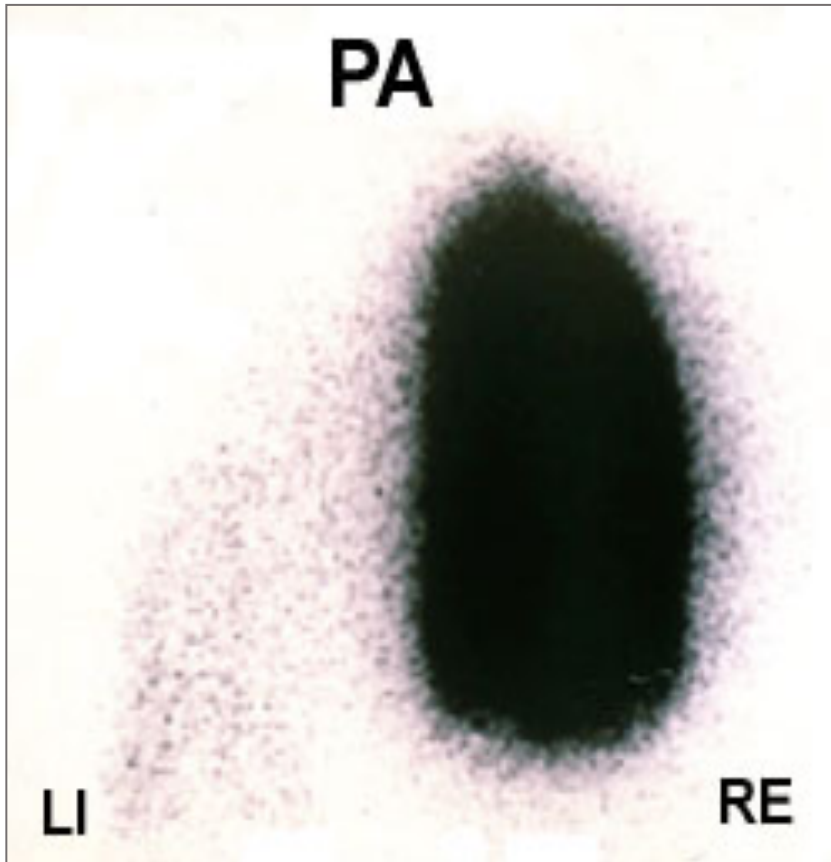


Fadel E. et al. Eur J Cardiothorac Surg. 2010; 38:277-84.

→ Double lung transplantation has become the standard of care for PAH

# What about single lung Tx for PAH?

**NOT RECOMMENDED!!!**



## *Postoperative period:*

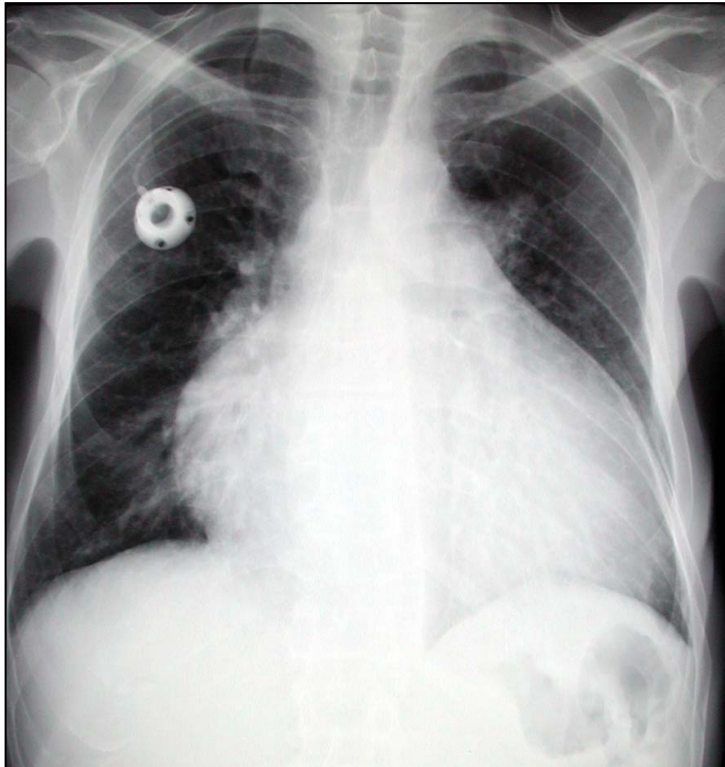
- 1. Volume overflow of single-lung graft*
- 2. Hemodynamic instability*
- 3. Primary graft dysfunction*

## *Later problems:*

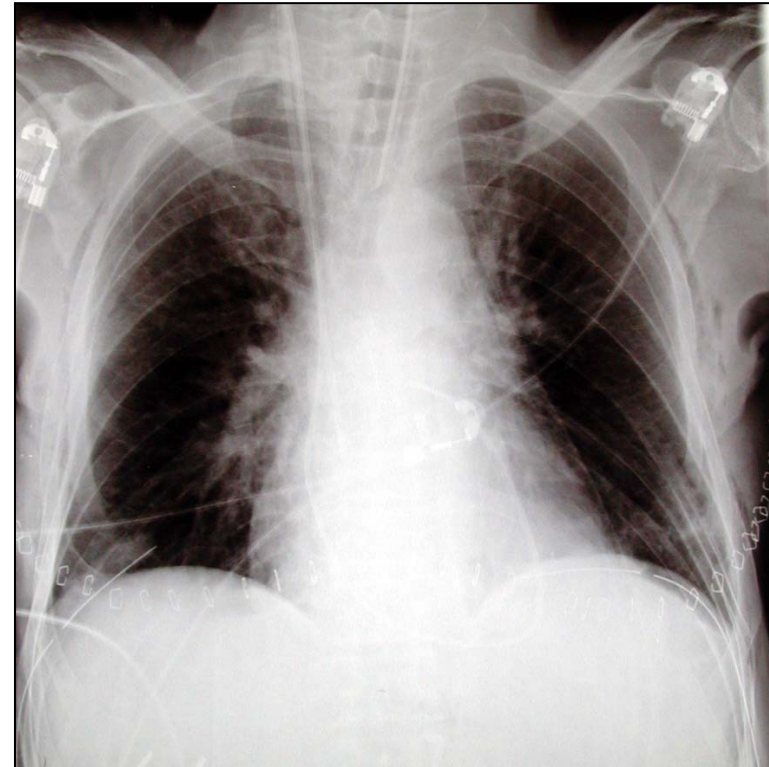
*V/Q mismatch with early CLAD*

# The early postoperative phase after LTx for PAH can be challenging 'cardiac remodeling'

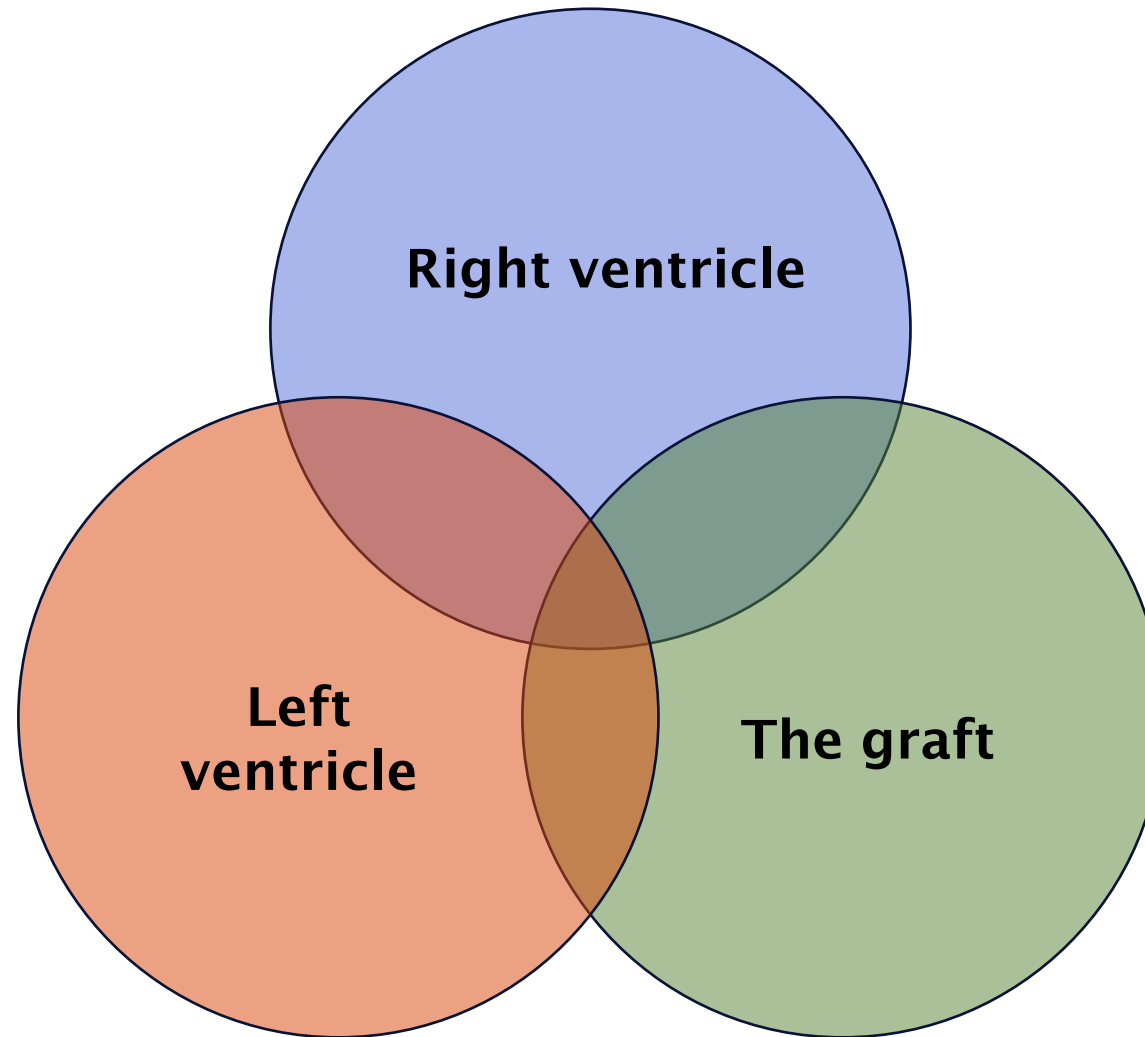
Pre-LTx



Post-LTx



# The three dimensions of early post-LTx challenges of PAH

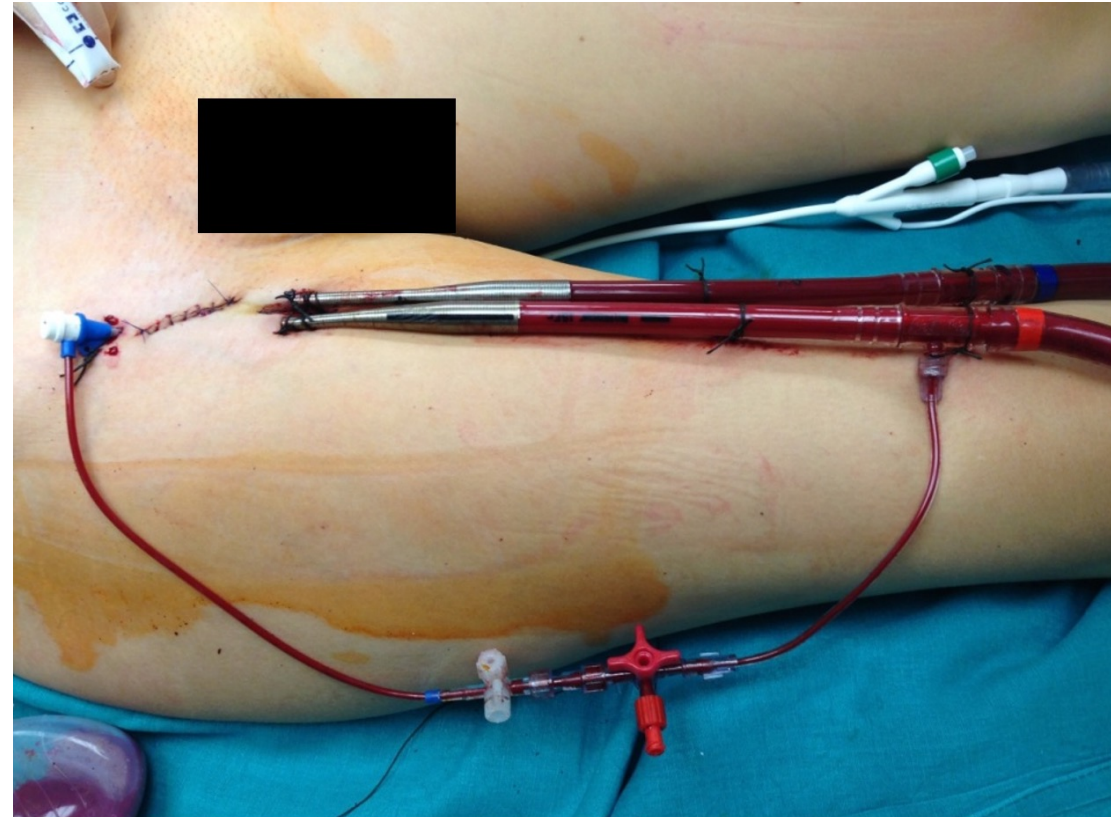




# Prophylactic postoperative ECMO

femoro-femoral  
veno-arterial

femoral artery: 15-17 Fr  
femoral vein: 17-19 Fr  
leg cannula: 8 Fr  
LMWH (anti-Xa guided)



→ postoperative VA ECMO in PAH patients - mean of 2.4 days

# ... excellent outcome with this strategy

European Journal of Cardio-Thoracic Surgery 53 (2018) 178-185

**Lung transplantation for idiopathic pulmonary arterial hypertension on intraoperative and postoperatively prolonged extracorporeal membrane oxygenation provides optimally controlled reperfusion and excellent outcome**

EUROPEAN JOURNAL OF  
**CARDIO-THORACIC SURGERY**



Moser et al. Eur J Cardiothorac Surg. 2018

## Retrospective analysis: January 2000 and December 2014

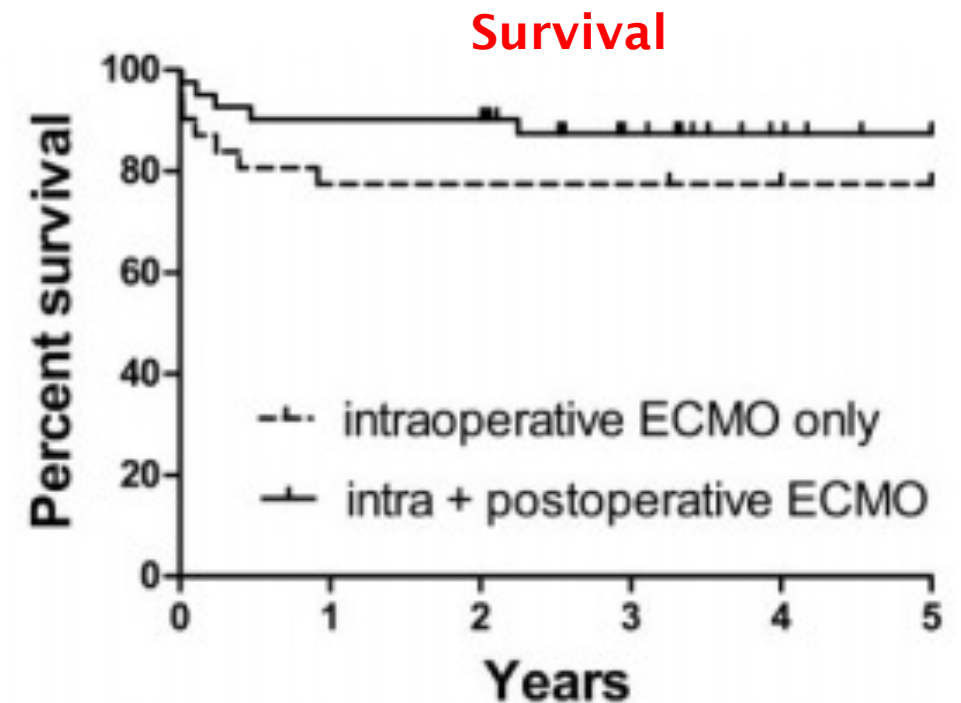
72 patients: **Group A: 41 intra + postop ECMO**

vs

**Group B: 31 intraop ECMO only**

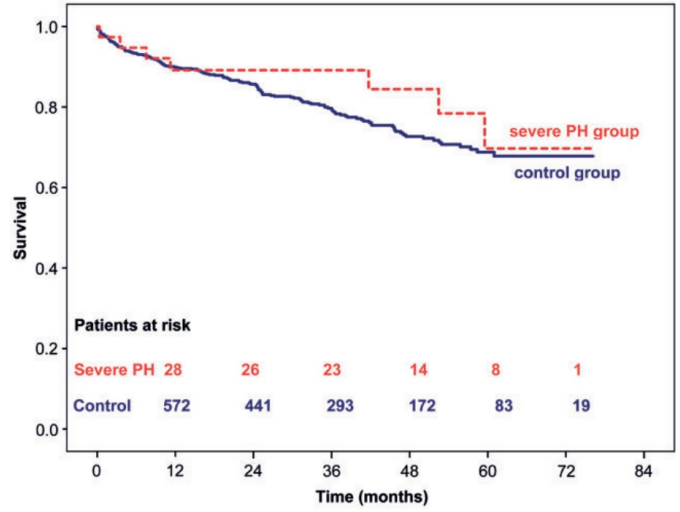
### Survival rates (p = 0.189)

	intraOP+prolonged	intraOP only
90-day	92.7 %	83.9 %
1-year	90.2 %	77.4 %
3-year	87.4 %	77.4 %
5-year	87.4 %	77.4 %

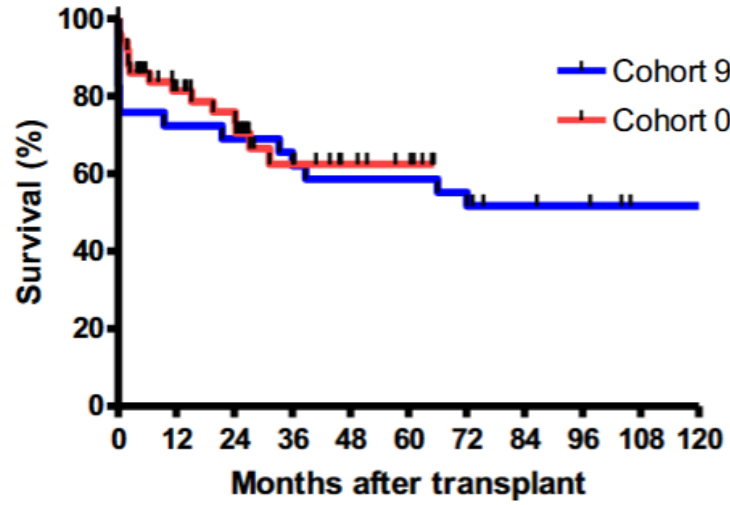


# Recent survival outcome of double-lung transplantation for PAH

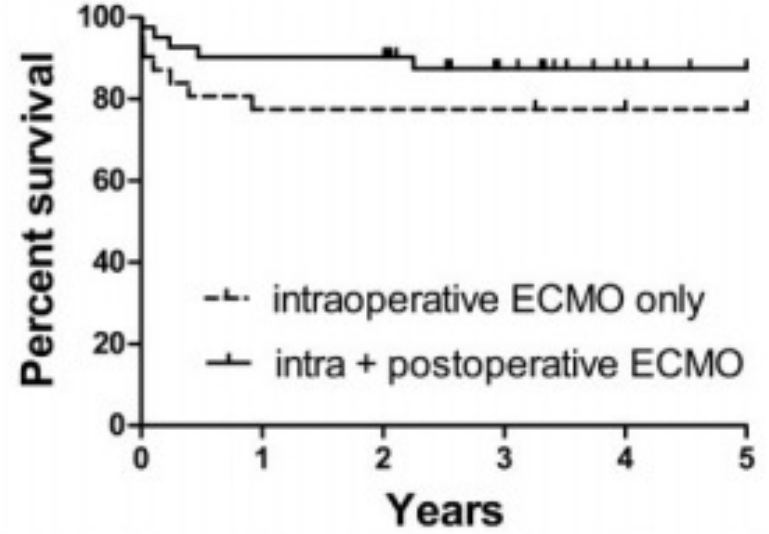
Hannover



Toronto



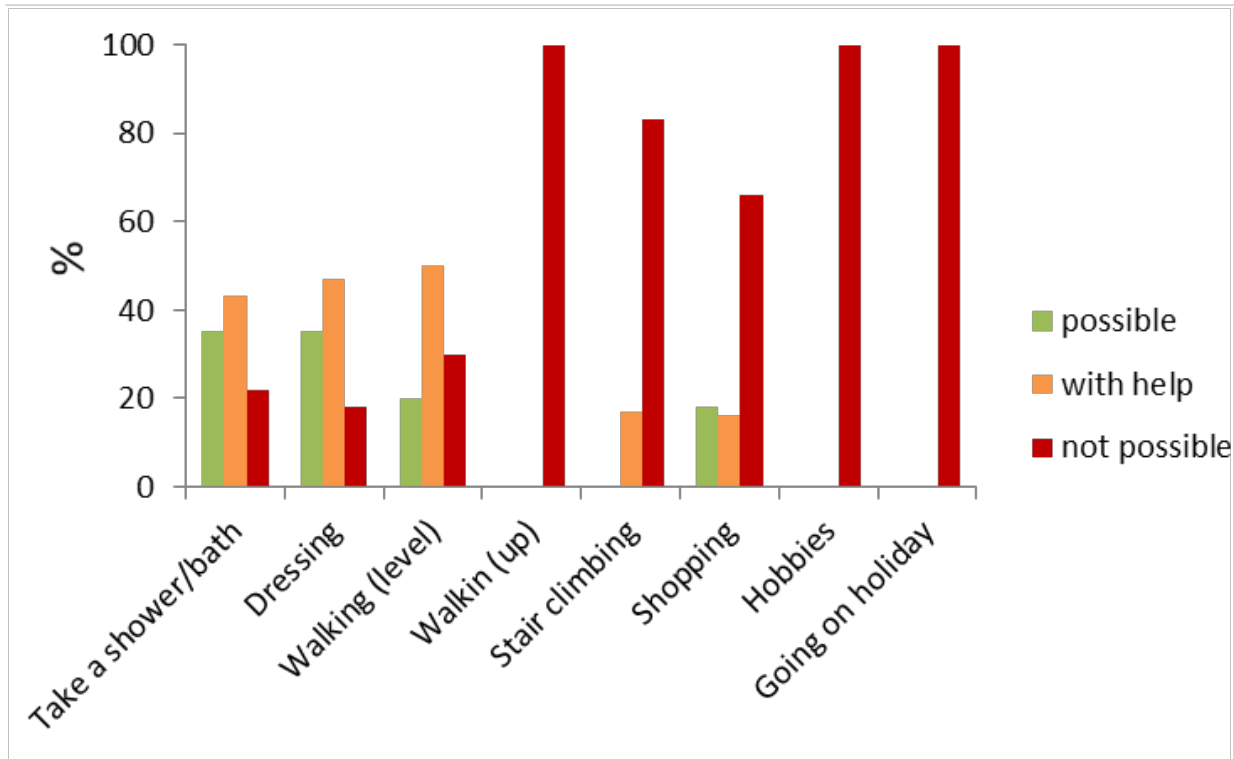
Vienna



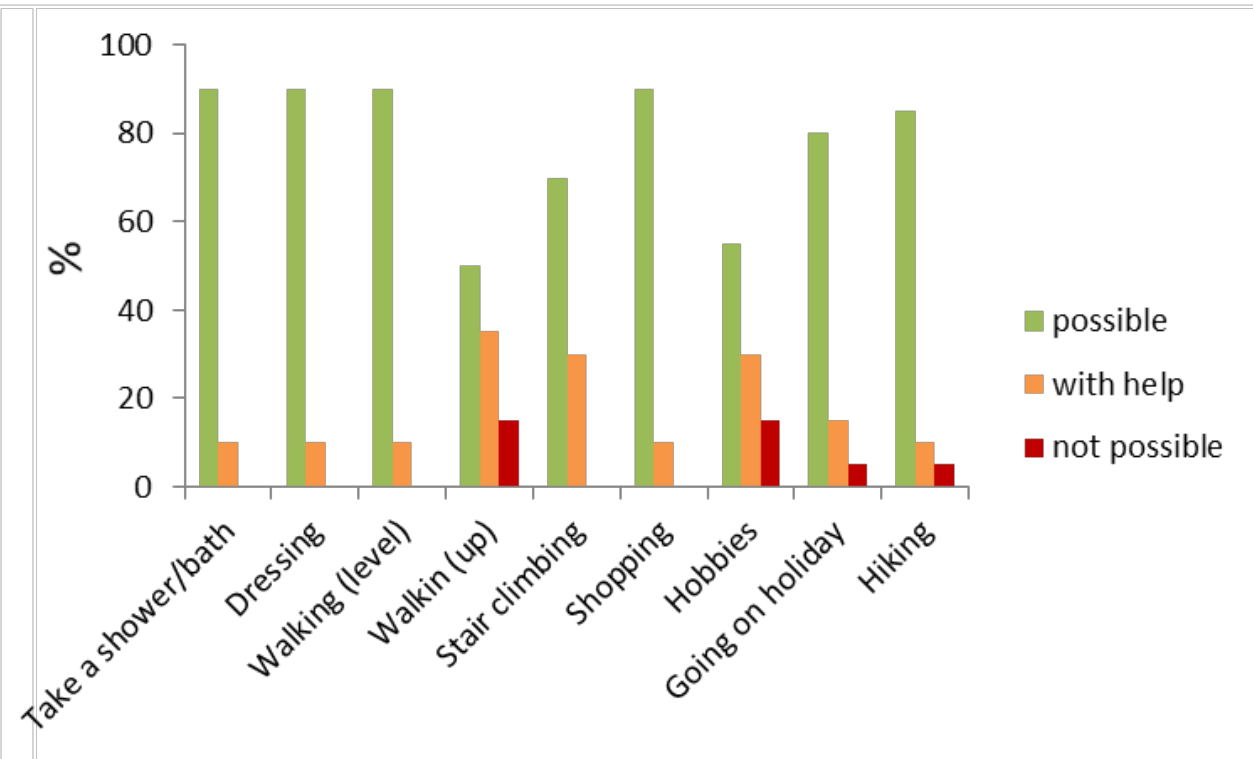
5-year: 65-75%

# Is mere survival a good outcome measure?

## Activities before Tx



## Activities one year after Tx



# Going to extremes...

## THE TIMES

### Lung transplant patients conquer Kilimanjaro



The group of 24 patients reach the summit

Simon Ashton was sat in his office in Merseyside when he received an unusual phone call from the Medical University of Vienna.

Could he organise an expedition to Mount Kilimanjaro within three months? Of course he could. Is a group of 24 people, plus 140 staff, possible? Yes, OK. Would it be a problem if the climbers were lung transplant patients, complete with their surgeons, doctors, psychologists and medical research equipment in tow? “We sat down and thought – can we really do this,” he recalled. “Then we said, ‘Yes we can’.”





## Summary: LTx for PAH

- Provides excellent outcome in experienced centers
- 1-year survival > 90 %, 5-year survival > 75 %
- Early selection of possible candidates
- Referral to LTx after failure of initial treatment