

## Outcome of Heart-lung or Double-lung transplantation in Pulmonary Hypertension Secondary to Congenital Heart Diseases

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### Introduction

Heart and lung transplantation or double lung transplantation with shunt closure is a curative option for patients with pulmonary arterial hypertension secondary to congenital heart diseases. Its benefit remains controversial given the slow evolution of the disease. We aim to investigate outcome of patients with CHD and PAH listed for transplantation.

### Methods

- We investigated outcomes of 90 patients with PAH-CHD listed for transplantation in a single center.

### Results

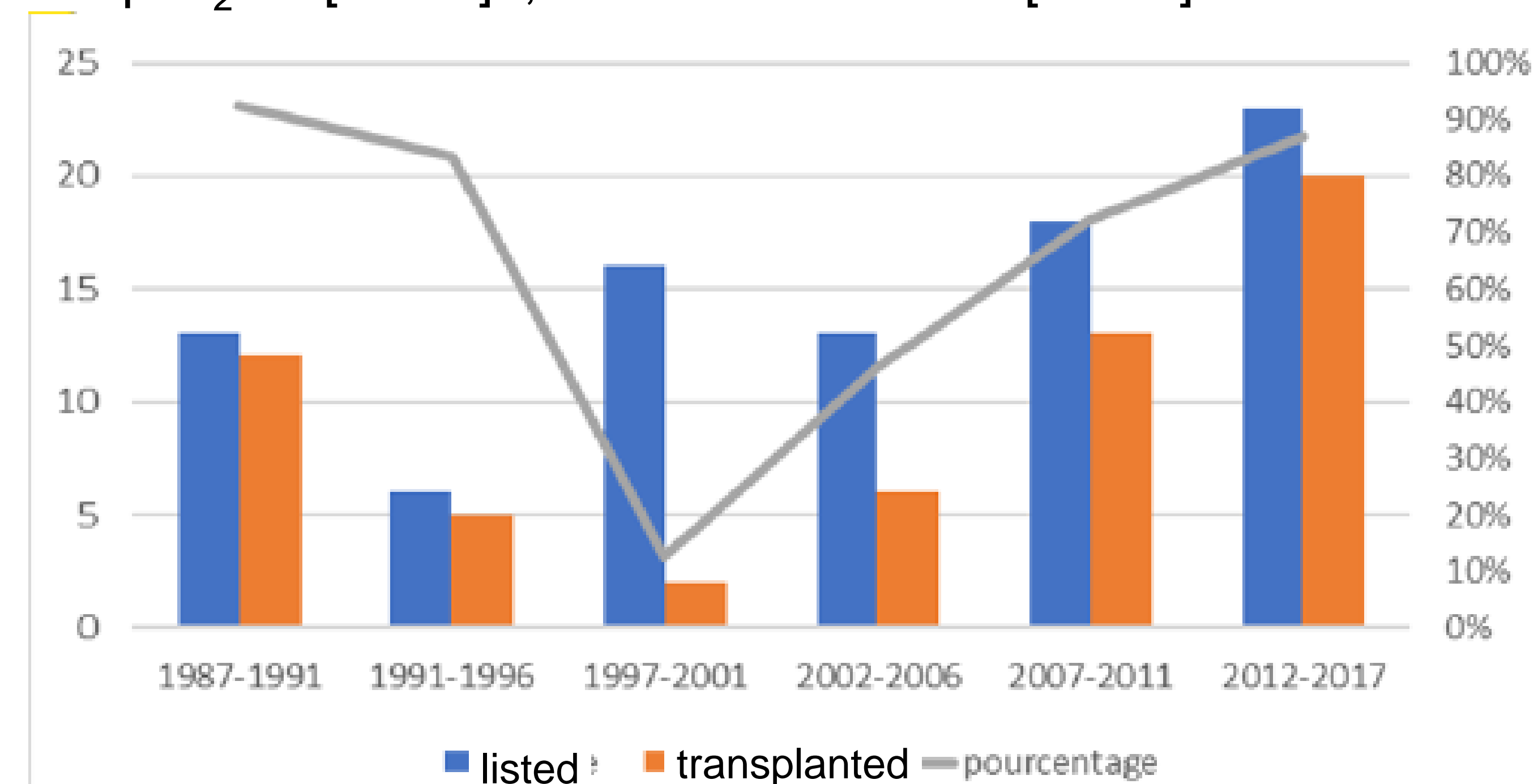
- Patients had Eisenmenger syndrome in 66 cases (75%) and post corrective surgery PAH in 22 cases (25%)

	n	nyha	Freq.	Percent
<b>Initial CHD</b>		2	2	2.33
<b>ASD</b>	29	3	35	40.70
<b>VSD</b>	24	4	49	56.98
<b>PDA</b>	9			
<b>TA</b>	4			
<b>SV</b>	10			
<b>AVSD</b>	3			
<b>Other</b>	11			
<b>Previous CHD surgery</b>	32 (35.6%)			
<b>Palliative surgery</b>	11			
<b>CHD repair</b>	21			

	position shunt	Freq.	Percent
<b>absent</b>		4	4.55
<b>mixte</b>		5	5.68
<b>post</b>		49	55.68
<b>pre</b>		30	34.09
<b>Total</b>		88	100.00

- Mean age was 32.5 ± 10.5 yo (10 patients < 18 yo)
- 6'WT 336 ± 108 m
- Right heart failure n=37 (53.6%), ascitis n=9,
- inotropic support (n=10) ECMO n=4 (TGA-ASD x2 – APVR)
- Orotracheal intubation n=5
- CI Creat < 30 ml/min n=3
- mPAP 69 ± 19 mmHg, RAP 10 ± 9 mmHg
- Qs 2.9 ± 1.1 l.min<sup>-1</sup>.m<sup>-2</sup>, PVR 22 ± 13 WU
- SpaO<sub>2</sub> 87 [77-91], SVC saturation 60 [52-70]



- Double Lung transplantation n=8 + 3 percut. ASD closure
- Heart and lung transplantation n=46 / ECC 223 ± 55 minutes
- Post transplant. ECMO n=10 (18.5%) / Dialysis n=18 (39.1%)
- Bleeding n=15 (27.8%)
- Ventilation duration 5 days [2-15]
- ICU duration 15 days [5-26]
- Hospital stay 33 days [23-60]

Listed n=90 (emergency list n=19)

3.5 months [11j-14m]

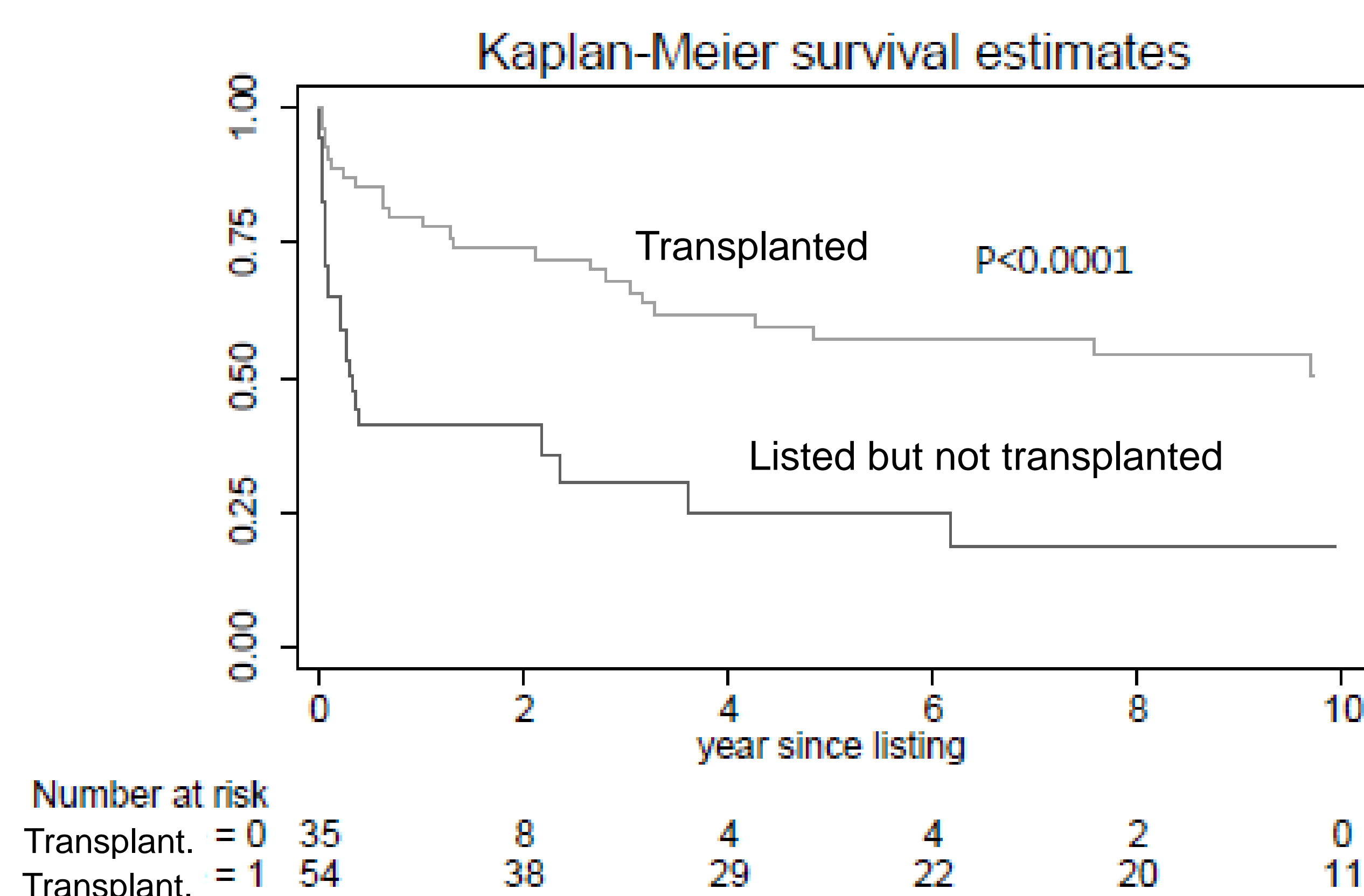
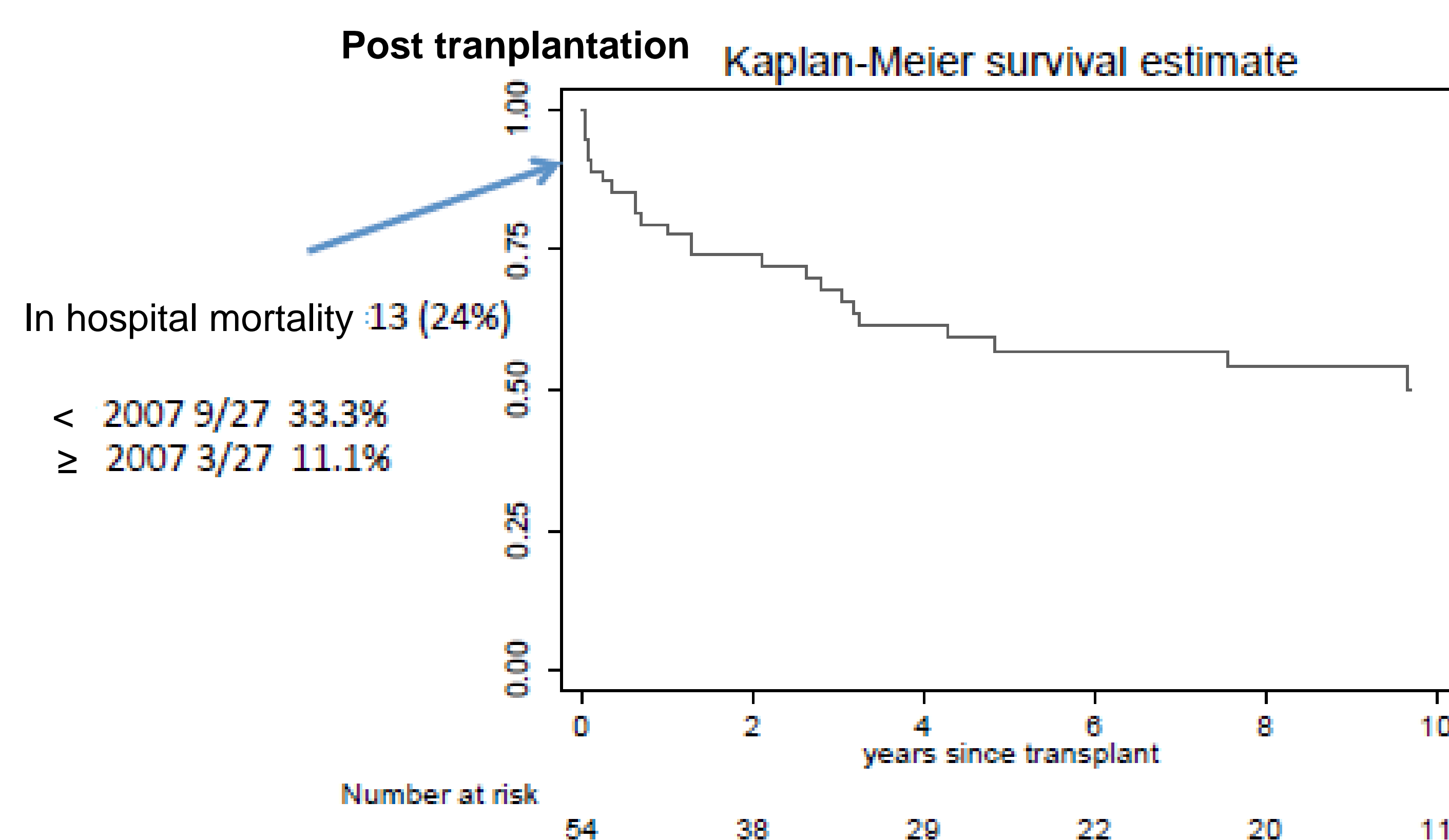
Transplantation n=54

no transplantation n=36

Deaths n=24

Emergency list available in France since 2006 (HLT) and 2007 (Lung transplant)

- Transplantation rate :**  
< 2007 : 27/48 (56.2%) ≥ 2007 27/42 (64.3%) p=NS
- Transplantation delay:**  
< 2007 : 3.1months [0-12.4] ≥ 2007 3.9 [0.7-18.7] p=NS
- Mortality rate on list**  
< 2007 : 19/48 (39.5%) ≥ 2007 5/42 (11.9%) p=0.004



### Conclusion

- High mortality rate on listing (>25%)** / selected at-risk end-stage PAH-CHD population
- High mortality rate post transplantation** but decreasing over time with increased experience. Transplantation in patients with PAH-CHD remains at high risk..
- Long term survival > 50% at 10 y.** post transplantation
- Transplanted patients outcome is better compared to patients listed but not transplanted**