

Changing Natural History of Pulmonary Hypertension in the last 20 years

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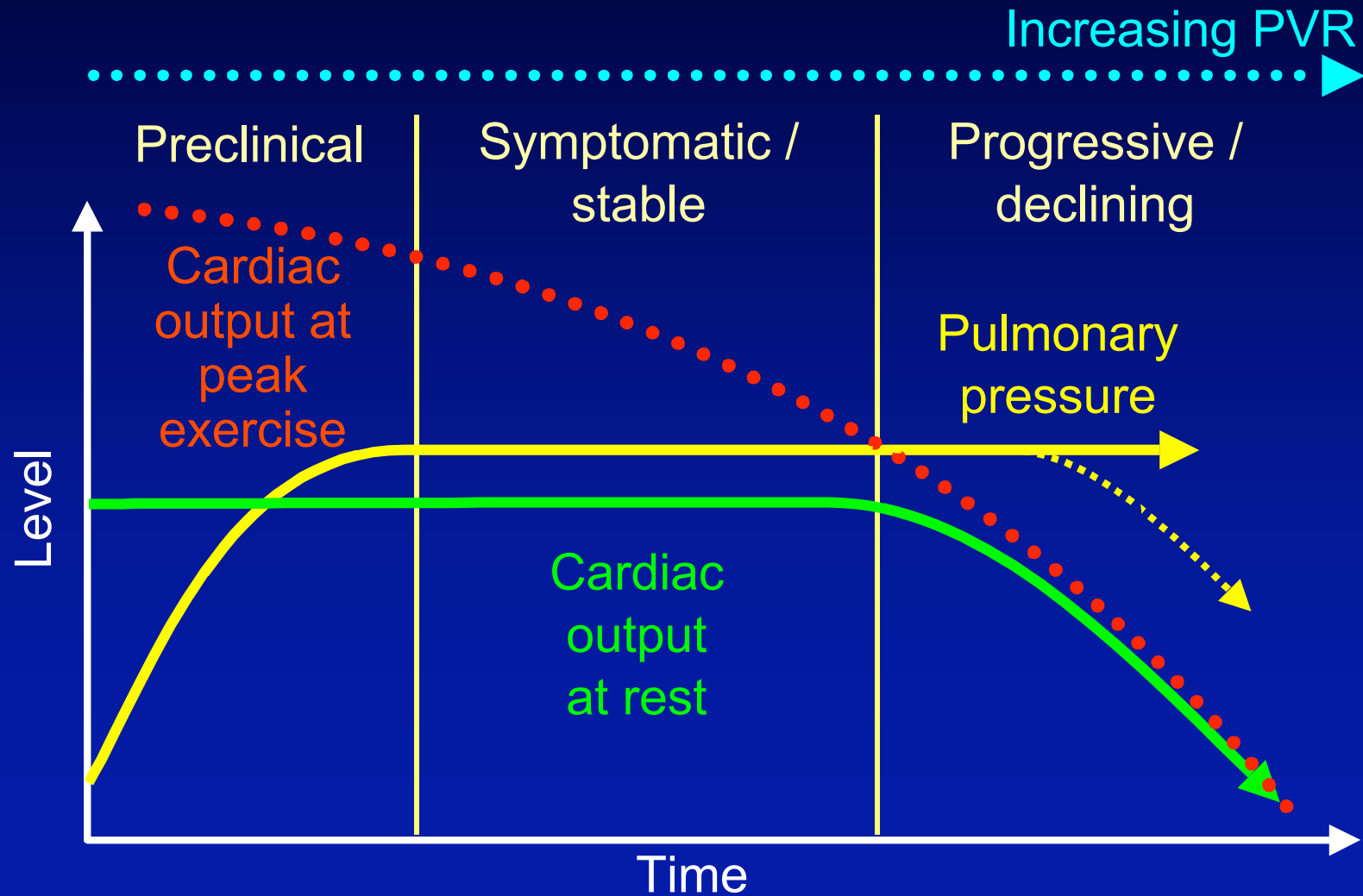
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Classical outcome in PAH



Natural History of PAH

- Although idiopathic PAH is perceived as a progressive disease, usually with a poor outcome, the natural history of the disease is heterogeneous, with some patients dying within months of diagnosis and others living for decades.
- The outcome of patients with other types of PAH has been less well described.
- Over the past decade, advances in medical therapies has altered the course of the disease and have made decisions regarding lung transplantation more complicated.

Background PAH

Prognosis of PAH is poor

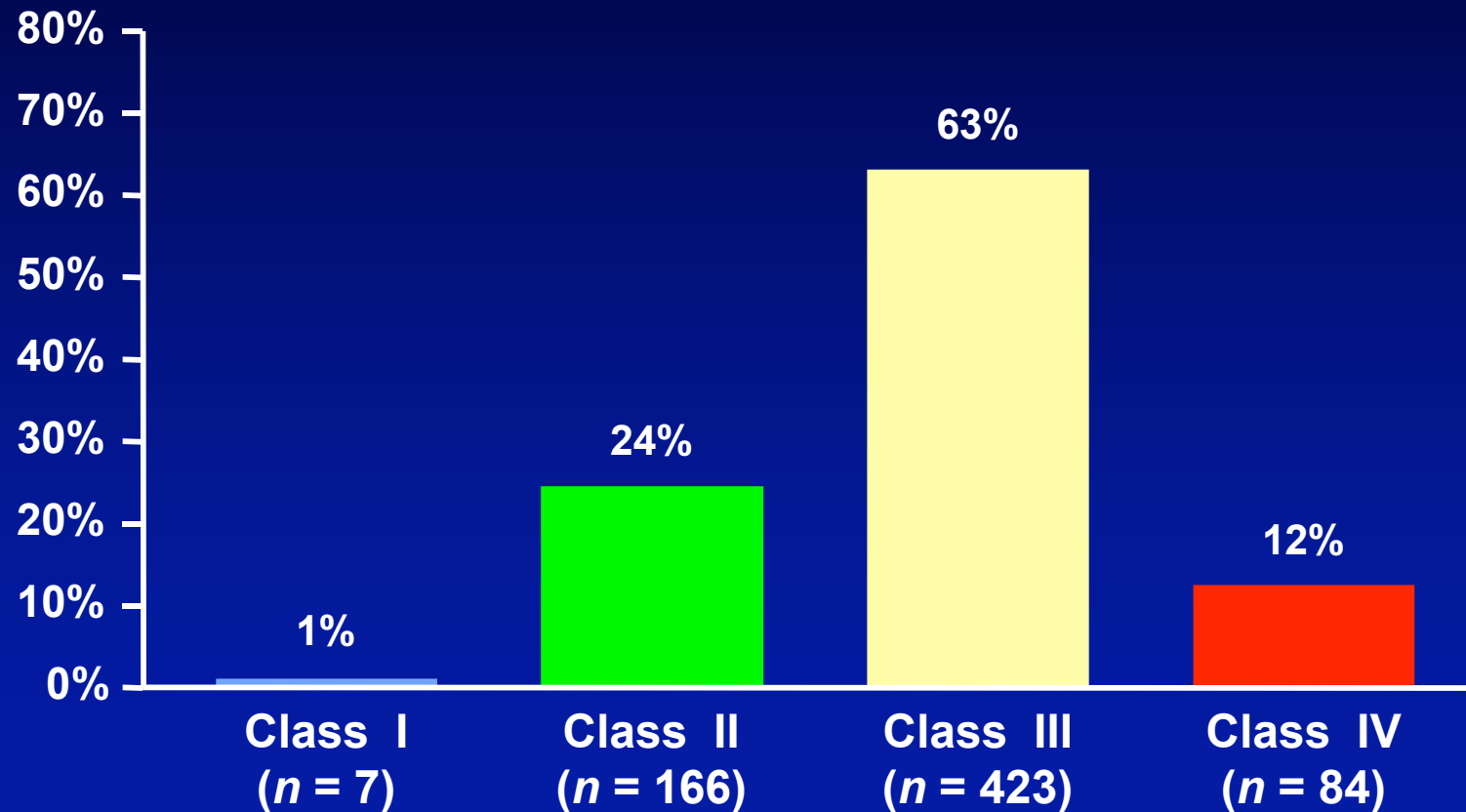
Survival in Patients with Primary Pulmonary Hypertension

Results from a National Prospective Registry

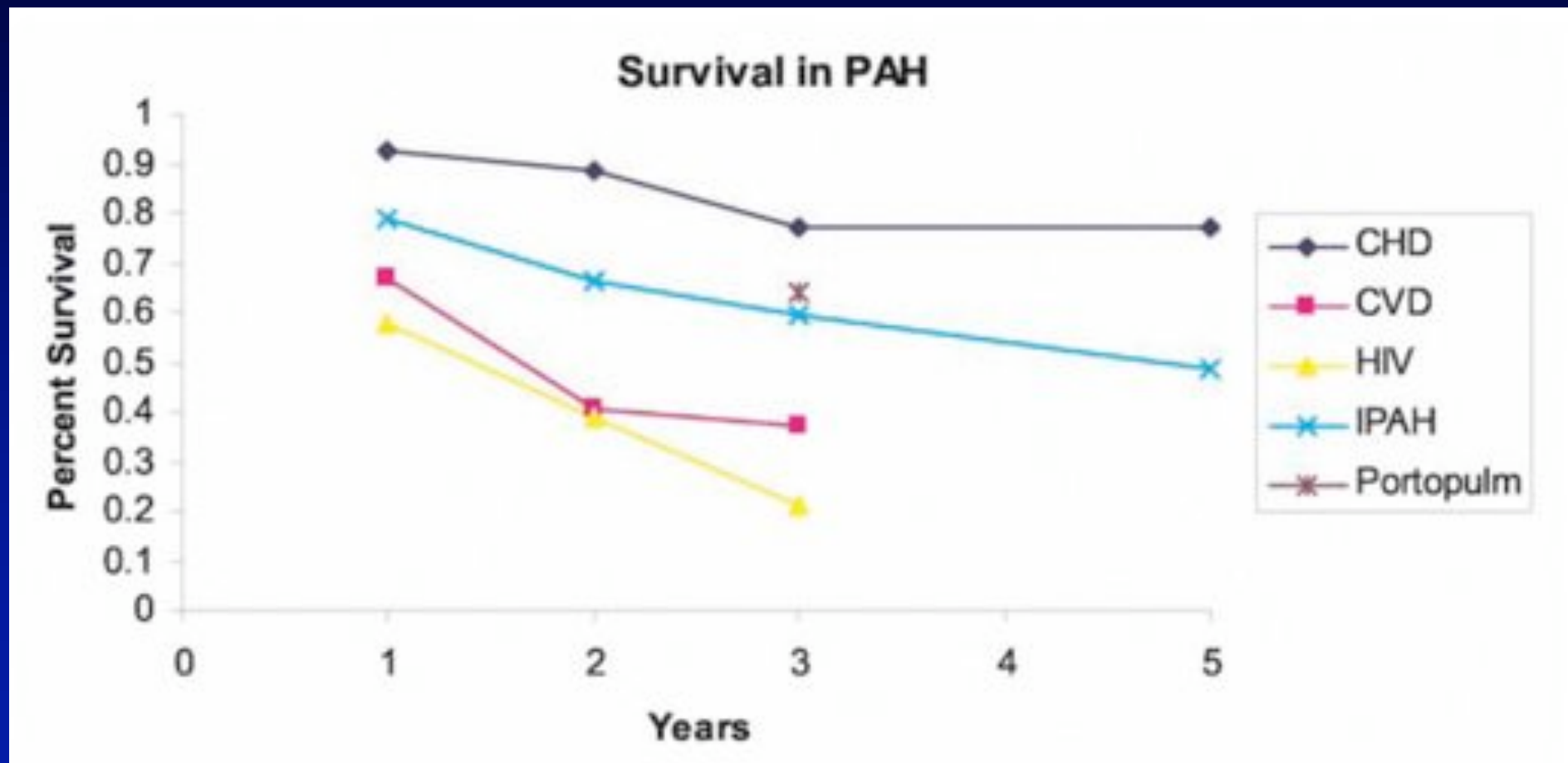
	Median survival time (months)
<i>NYHA class I and II</i>	58.6
<i>NYHA class III</i>	31.5
<i>NYHA class IV</i>	6.0

NYHA FC at diagnosis

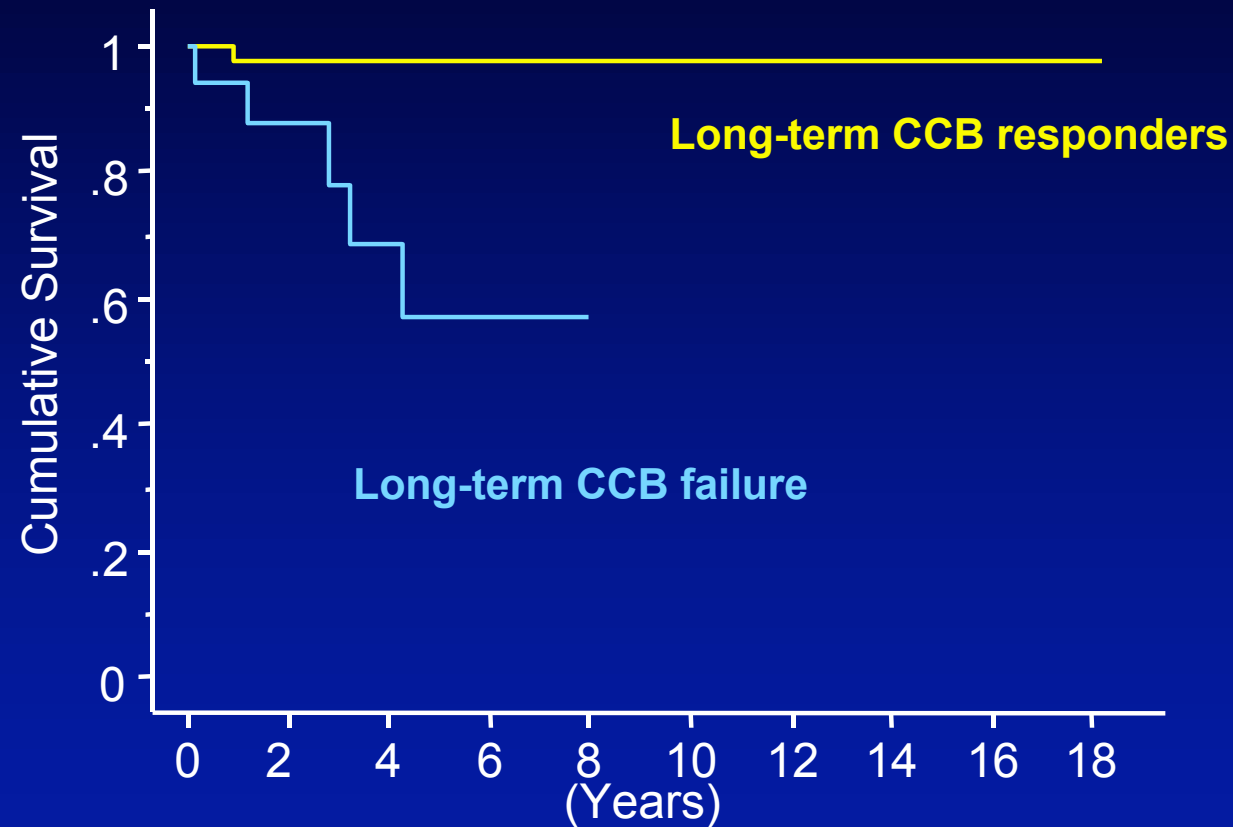
French National Registry



Survival in different forms of PAH



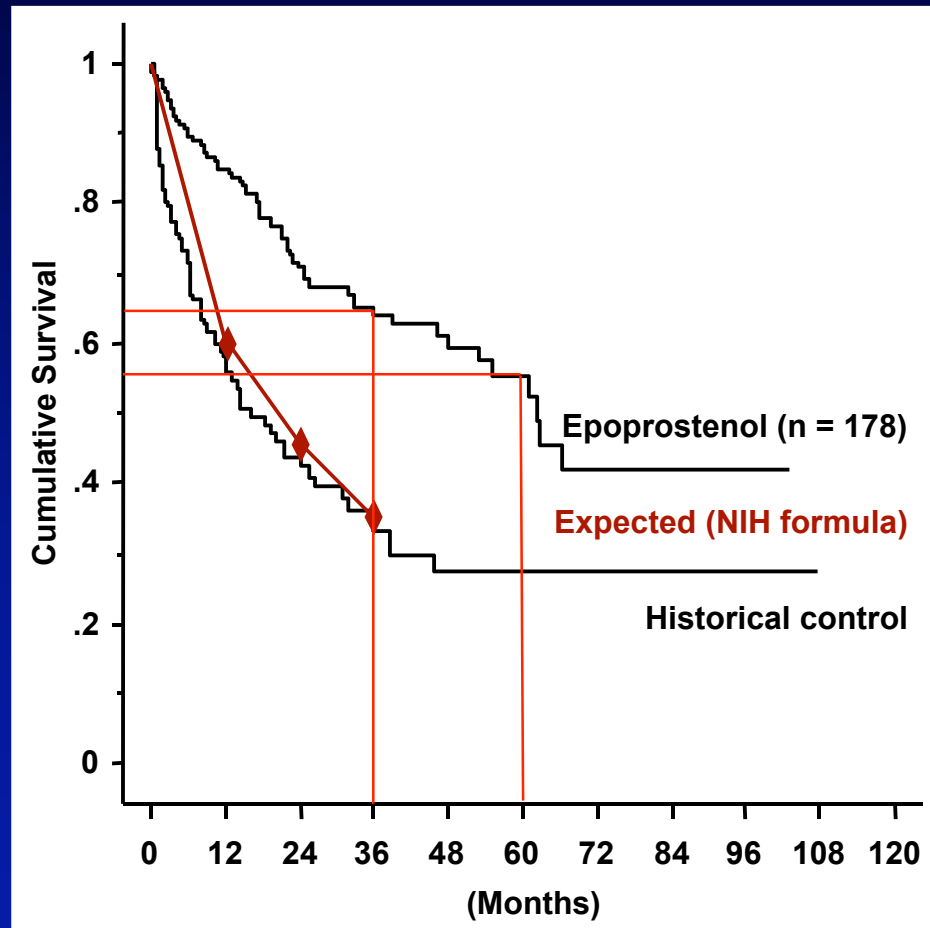
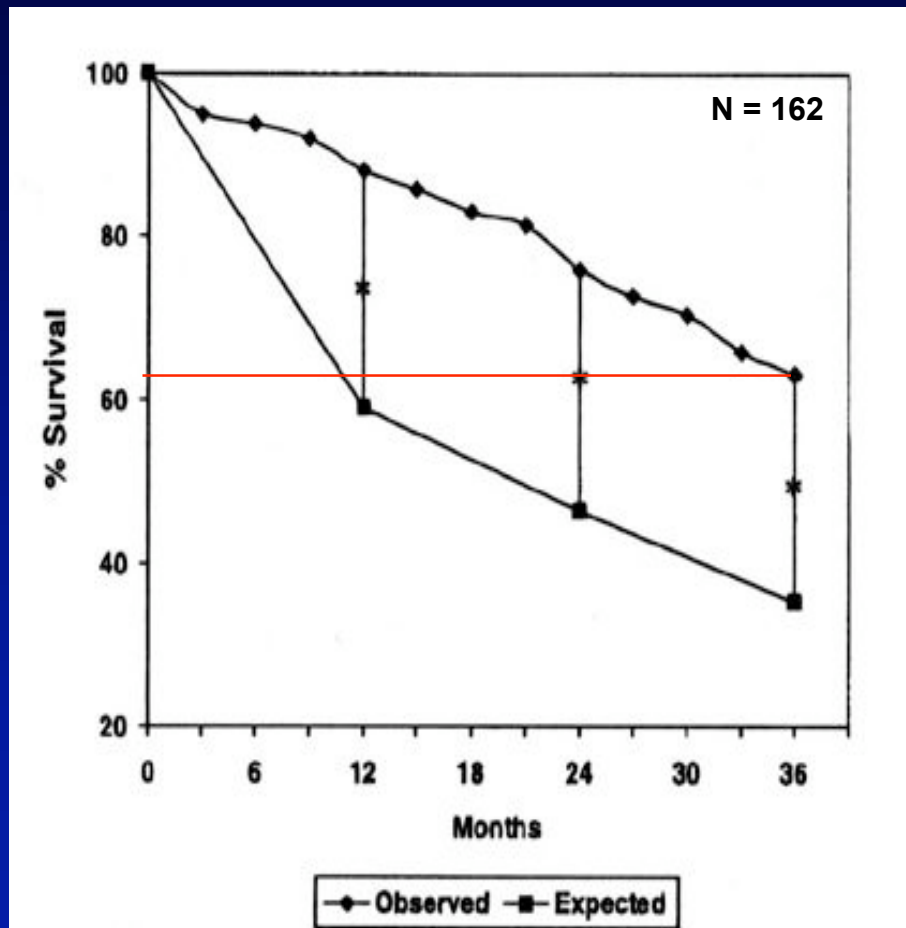
Survival in IPAH long-term CCB responders



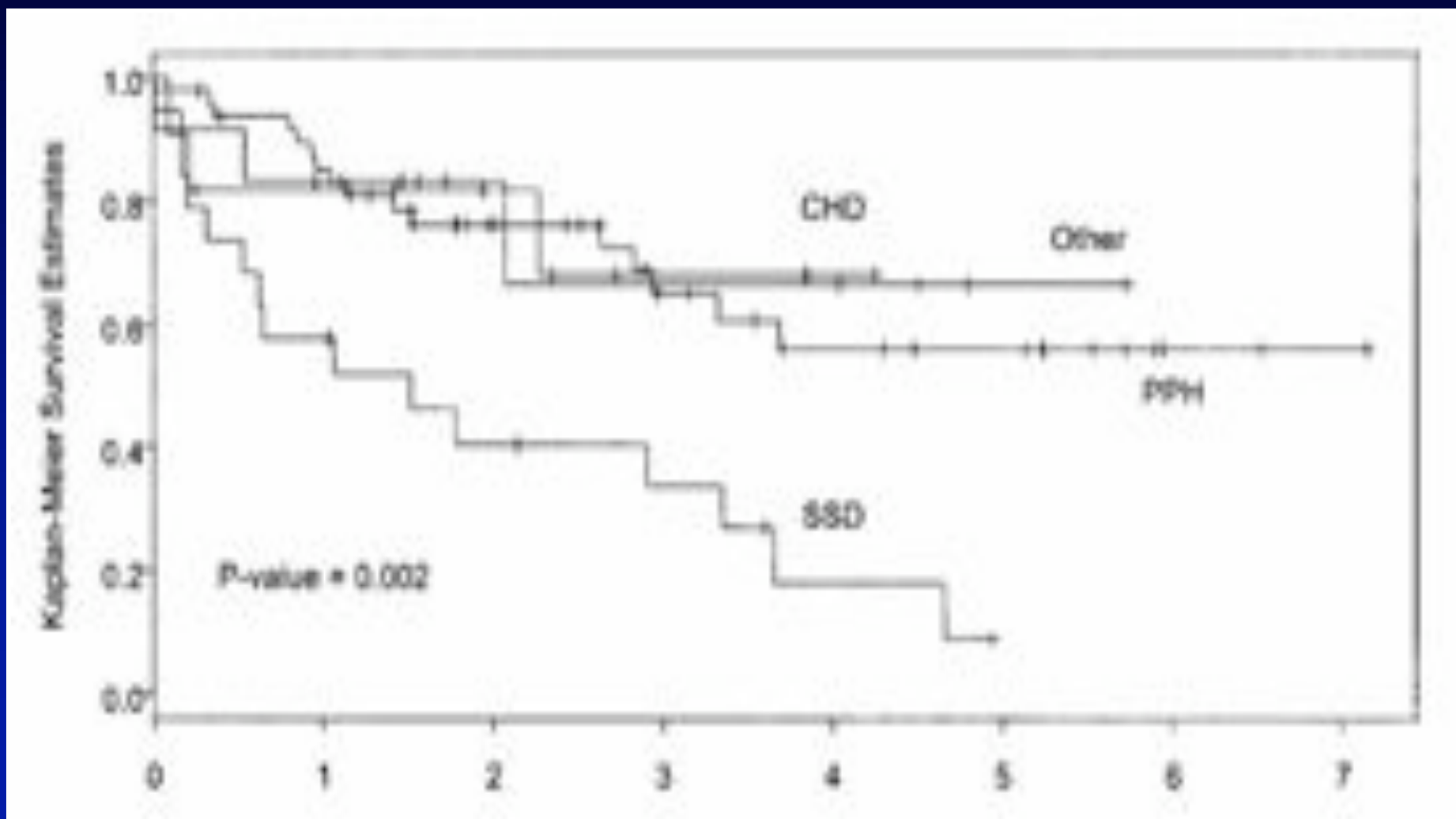
	0	2	4	6	8	10	12	14	16	18	
Subjects at risk, n	38	33	30	22	13	8	3	3	2	1	Long-term CCB responders
	19	12	7	4	0						Long-term CCB failure

Long-term epoprostenol in IPAH

Survival



Outcome in 91 Consecutive Patients with Pulmonary Arterial Hypertension Receiving Epoprostenol



Survival in IPAH

Epoprostenol or lung transplantation ?

