



Symposium SSHP

Geneva, September 2nd 2010



**Schweizerische Gesellschaft für Pulmonale Hypertonie SGPH
Société Suisse sur l'Hypertension Pulmonaire SSHP
Società Svizzera di Ipertensione Polmonare SSIP
Swiss Society for Pulmonary Hypertension SSPH**

The Swiss pulmonary hypertension registry:

what does it learn us?



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**Why
do we need a
pulmonary
hypertension
registry?
???**

PH: diagnostic algorythm

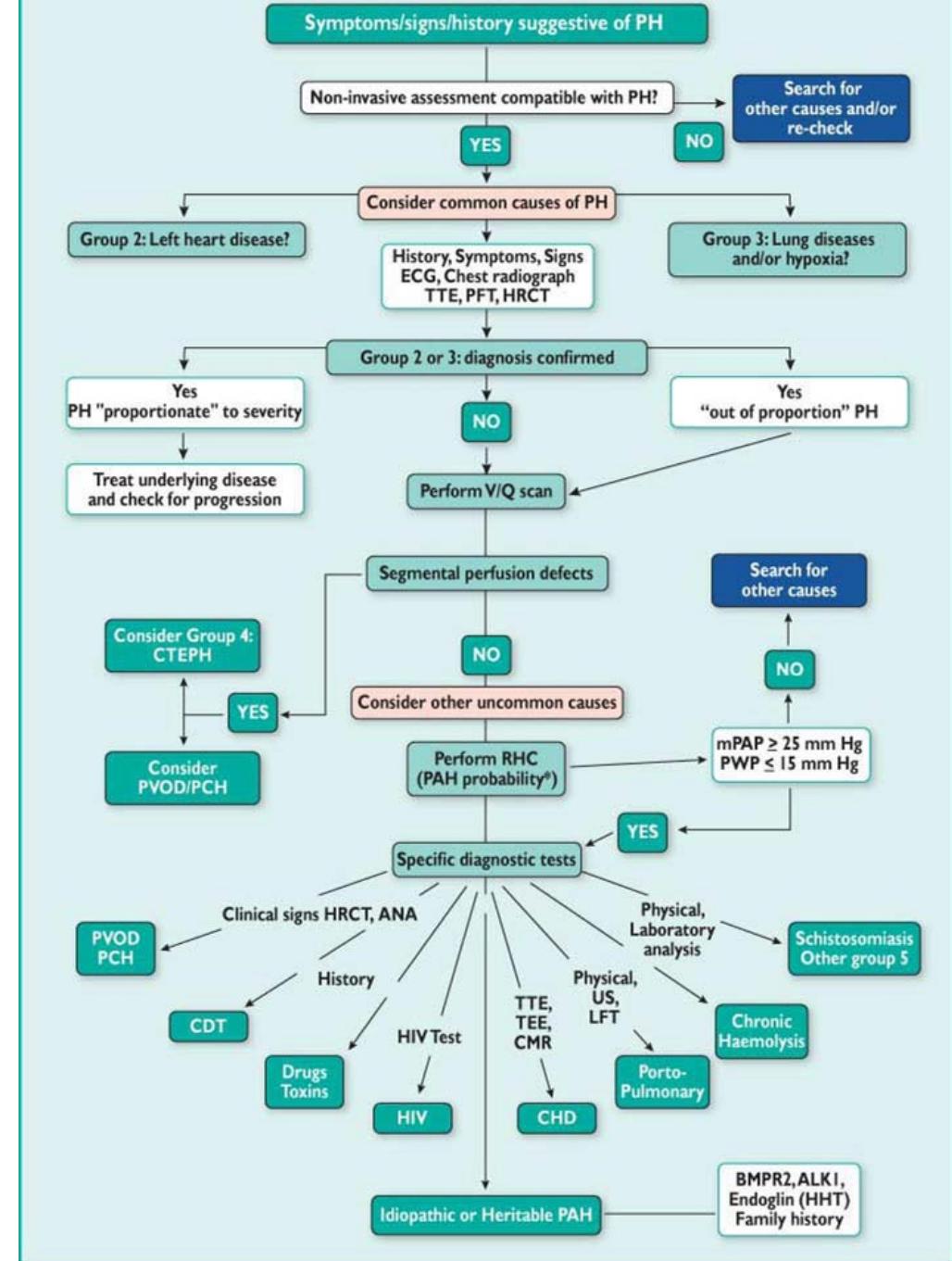


Eur Heart J 2009; 30(20):2493-537

Eur Respir J 2009; 34:1219-1263

ESC / ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

The Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)



Pulmonary arterial hypertension (PAH)



PAH is a rare disease = orphan disease

“orphan disease”: affect < 1:2000 individuals



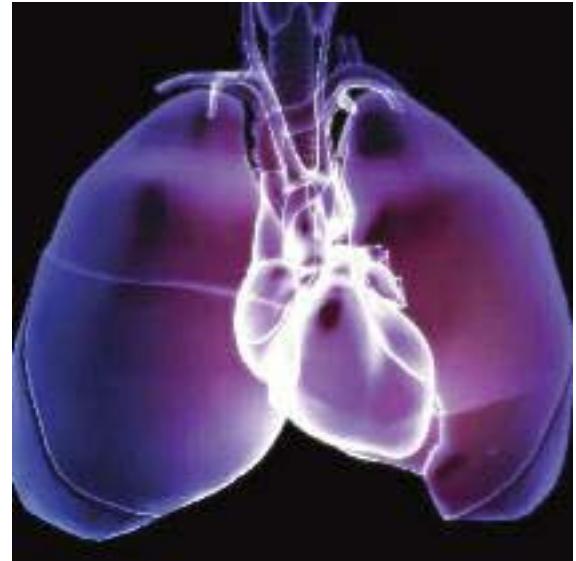
USA:	<200'000
EU:	< 5/10'000
AUS:	< 2000
J:	< 50'000
WHO:	< 6.5-10/10'000

Why a PH registry?



To yield informations on:

- diagnosis / subgroups
- prognosis
- demographics
- hemodinamics
- treatment modalities
- treatment outcome
- **survival**



The swiss PAH – registry



initiated 1999 - retrospective analysis 1990-1999
 - prospectively since 1999

collect information into:

- prevalence
- epidemiology
- characteristics
- therapy
- outcome

of PAH patients in Switzerland

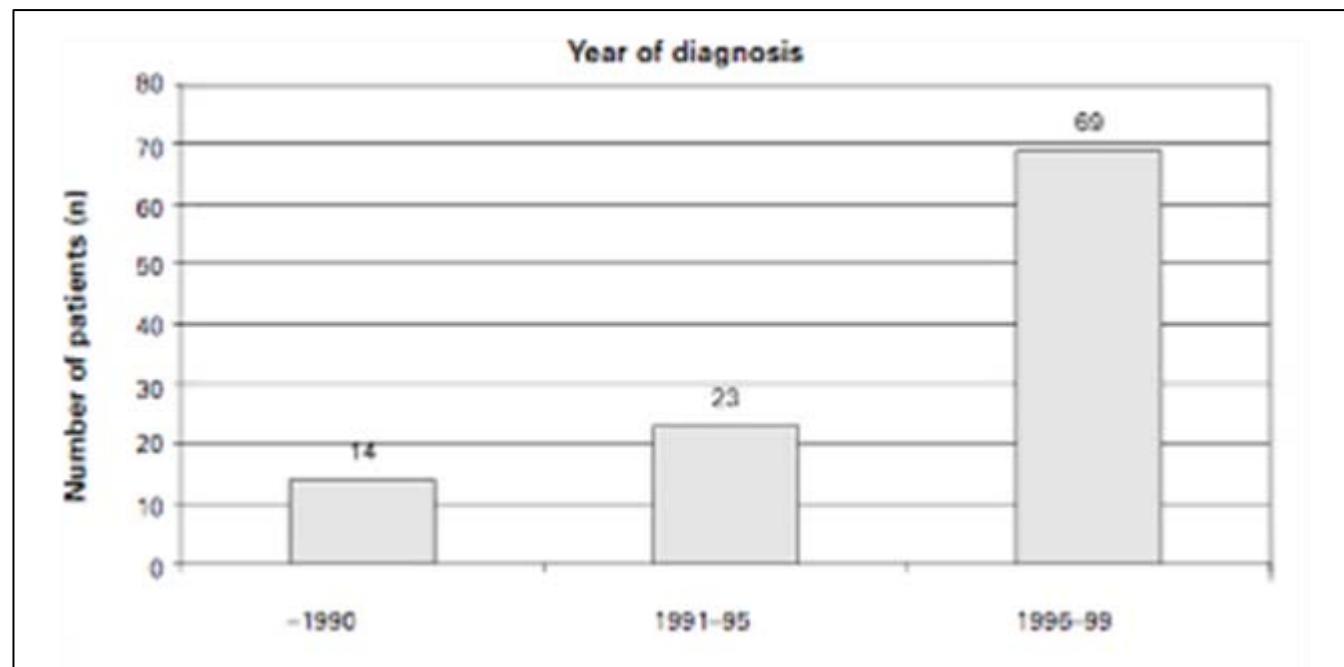
Swiss PH registry: patients characteristics 1999 (retrospective data 1991-1999)

	N	surviving	dead	p
6' walk test (m) 0.002	62	363 ±135	235 ±155	
SvO2(%)	66	66.5 ±9.5	57.9 ±10.1	0.006
PAPm (mm Hg) 0.06	82	53 ±12	59 ±13	
PAPs (mm Hg)	82	81 ±19	84 ±21	0.05
PVR (dyn.sec.cm-5)	67	892 ±419	959 ±455	0.11
PaO2(kPa)	72	9.1 ±1.8	9.2 ±2.3	0.9
HA class II vs III/IV(n)	94	18 vs 44	2 vs 18	0.015

Stricker SH et al, swissmedwyk 2001; 131:846-850



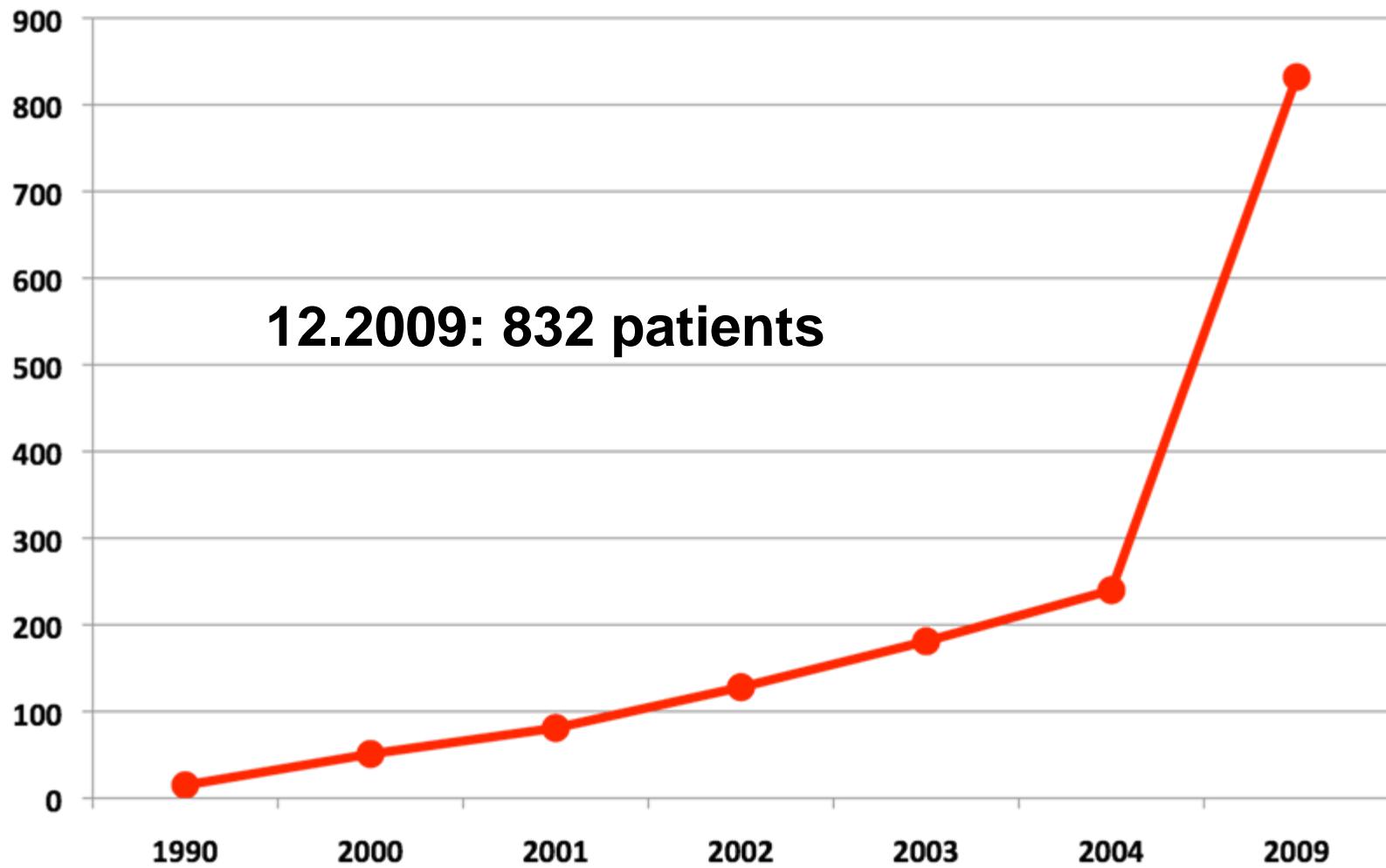
Swiss PH registry: number of patients



	1999	2000	2001	2002	2003	2004	total
Number of patients included	15	36	30	47	63	59	250
Lost of follow-up	2	4	0	4	4	10	24
Number of patients included in the registry each year							

Stricker H et al, swiss med wkly 2001; 131:346-350
Tueller T et al, swiss med wkly 2008; 138:379-384

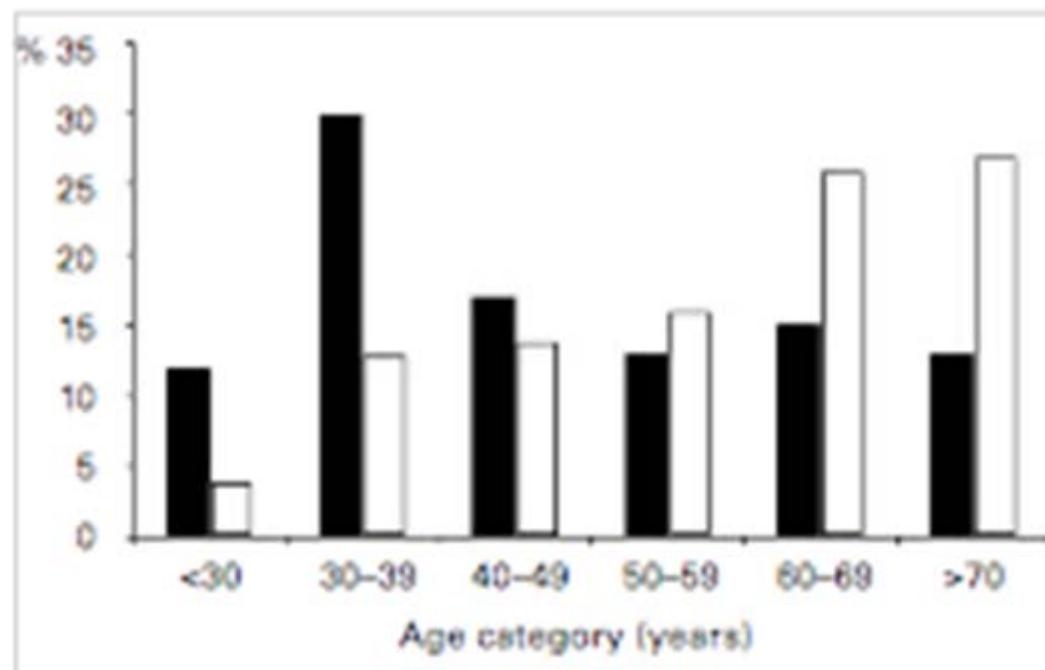
Swiss PH registry: number of patients



Source: Alabus / Swiss PH Registry



Swiss PH registry: age distribution



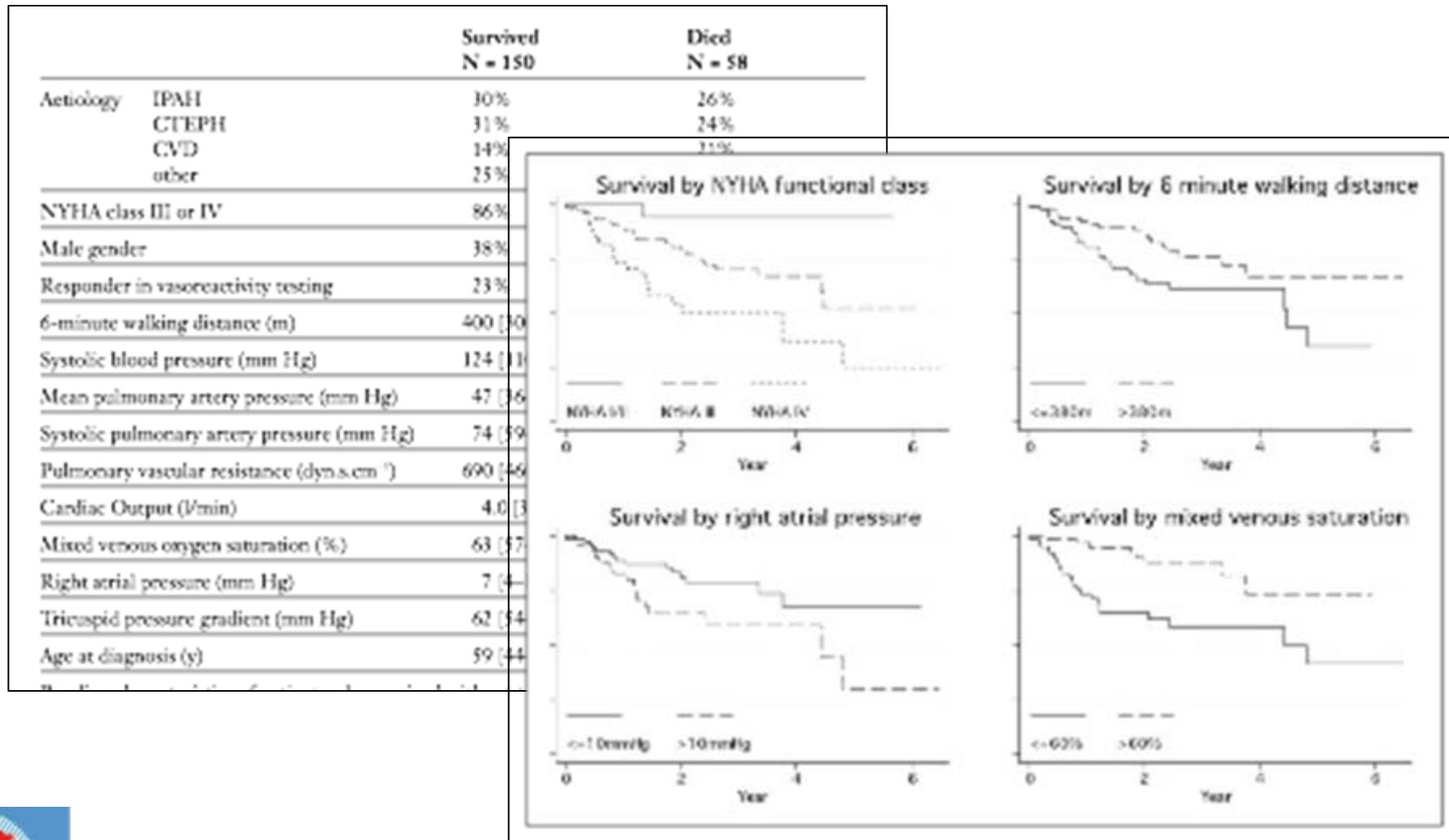
black: retrospective data 1991 - 1999
white: prospective data 1999 - 2004

Swiss PH registry: demographics 2004

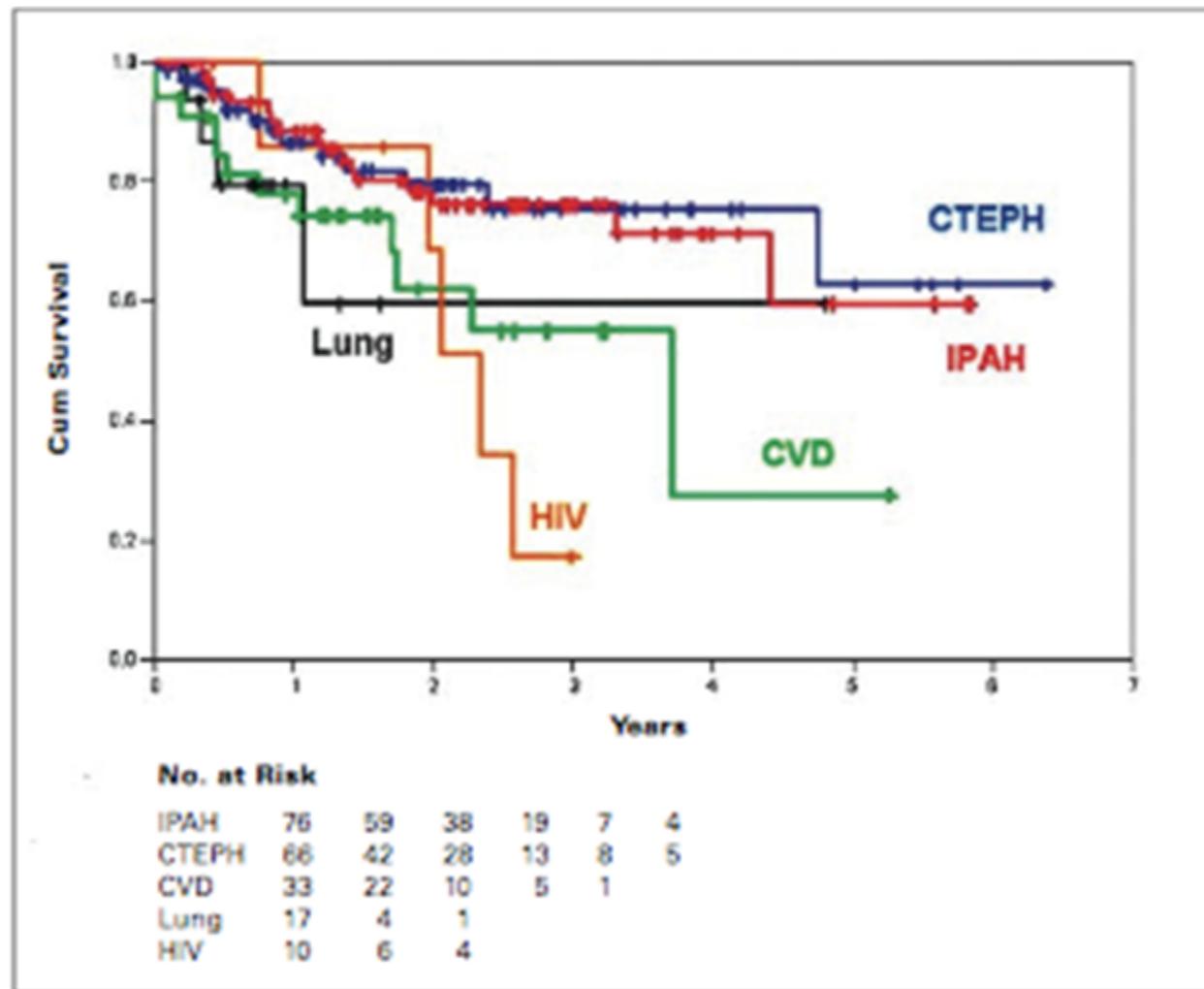
250 patients	Females 147 (59%)
median age	F 59y (46-70), M 63y (45-70)
median follow-up	18.8 months (9-31)
median time between visits	3.0 months (2-5)
most patients:	IPAH or CTEPH NYHA III-IV
Prevalence	all PAH: 55 / million adult inhabitants IPAH: 8.6 / million
Incidence	all PAH: 3.5 / million



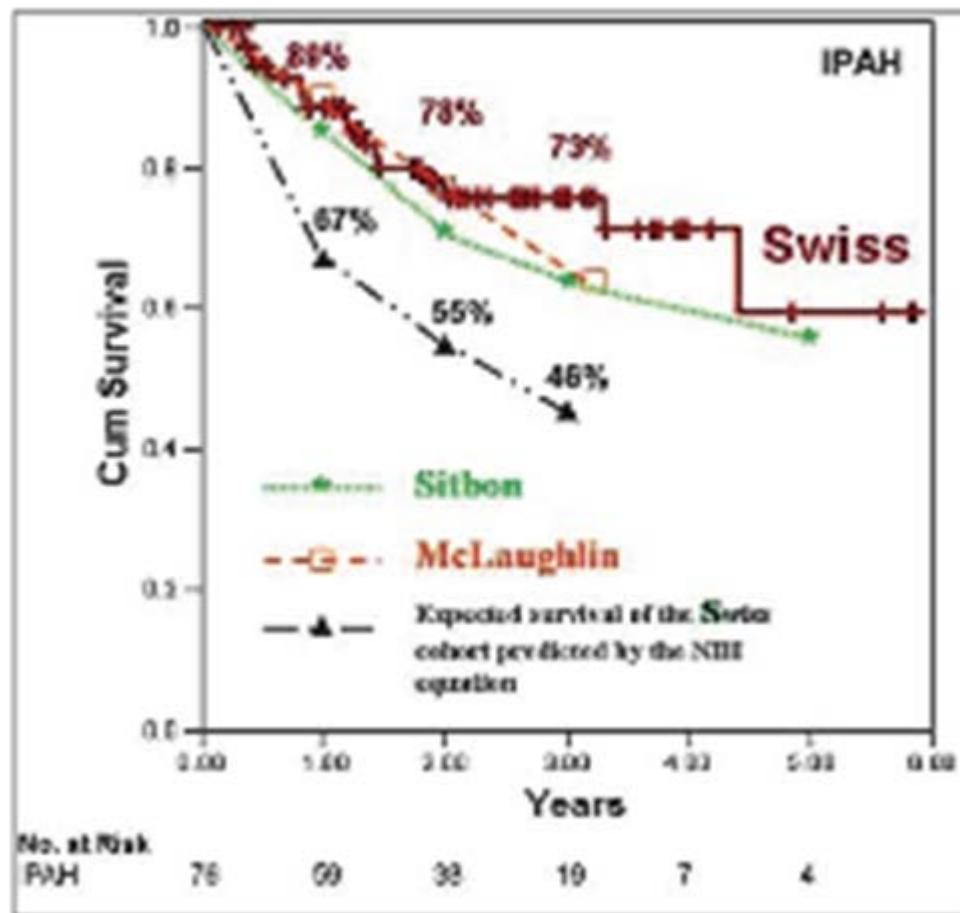
Swiss PH registry: patients characteristics 2004 (prospective data 1999-2004)



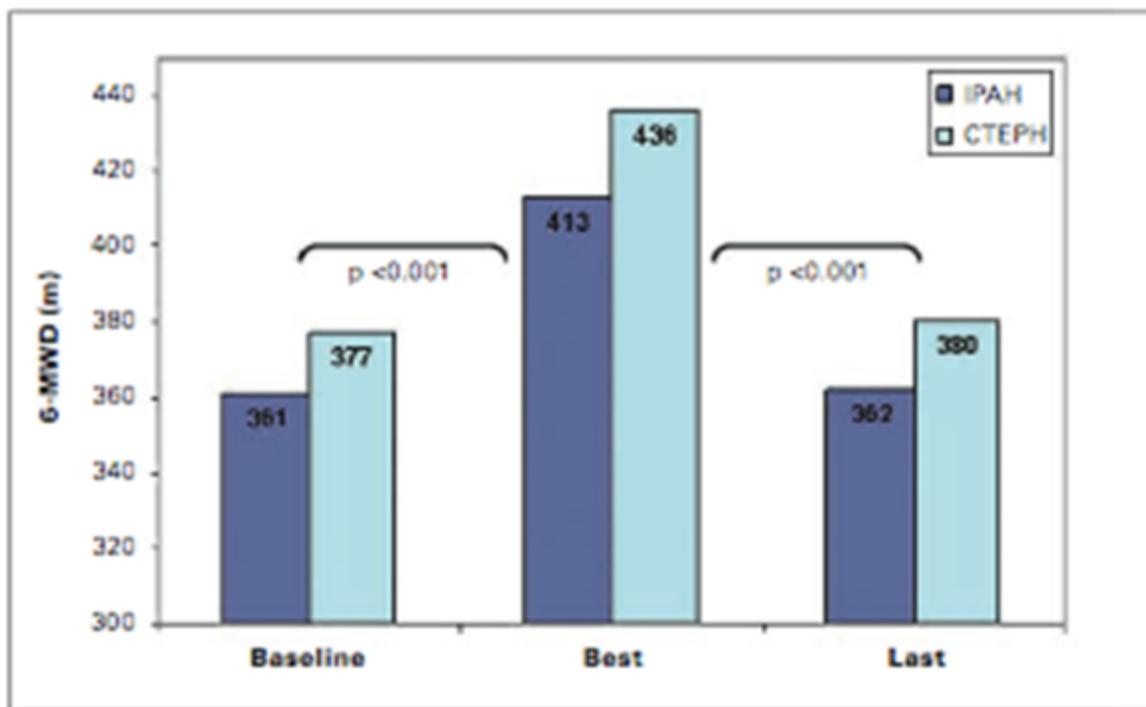
Swiss PH registry: survival 2004



Swiss PH registry: survival CH vs. NIH-formula and literature data (IPAH)



Swiss PH registry: evolution over time



	Baseline		Best		Last	
	IPAH	CTEPH	IPAH	CTEPH	IPAH	CTEPH
NYHA 1 (%)	0	0	1	6	4	4
NYHA 2 (%)	8	14	23	20	19	14
NYHA 3 (%)	69	60	48	40	37	44
NYHA 4 (%)	23	26	28	34	40	38

IPAH: Idiopathic pulmonary artery hypertension, CTEPH: Chronic thromboembolic pulmonary hypertension, NYHA: New York Heart Association

Swiss PH registry: modality of treatment

	Oxygen (%)		Diuretics (%)		OAK (%)	
	IPAH	CTEPH	IPAH	CTEPH	IPAH	CTEPH
Baseline	25	22	33	36	59	79
Best	41*	30	43	55*	84**	89
Last	42	38	49	61	84	98

IPAH: Idiopathic pulmonary artery hypertension, CTEPH: Chronic thromboembolic pulmonary hypertension, OAK = oral anticoagulation. *p <0.05; **p <0.001

	IPAH			CTEPH		
	BL	Best	Last	BL	Best	Last
Specific Therapy*	26 of 83 (31%)	63 of 75 (84%)	67 of 75 (89%)	22 of 70 (31%)	44 of 60 (73%)	46 of 57 (81%)
<u>Single drug</u>						
CCB (%)	8	2	0	9	5	4
Ilo inh (%)	50	22	16	71	51	30
Ilo iv (%)	8	6	5	4	0	2
Bos (%)	30	37	25	15	23	26
Sil (%)	9	4	3	0	5	9
<u>Multiple drugs</u>						
Bos+Sil (%)	0	2	5	4	2	7
Bos+Ilo (%)	4	19	16	0	7	7
Ilo+Sil (%)	0	2	3	0	5	13
3 and > (%)	0	6	27	0	2	2

PH: therapy

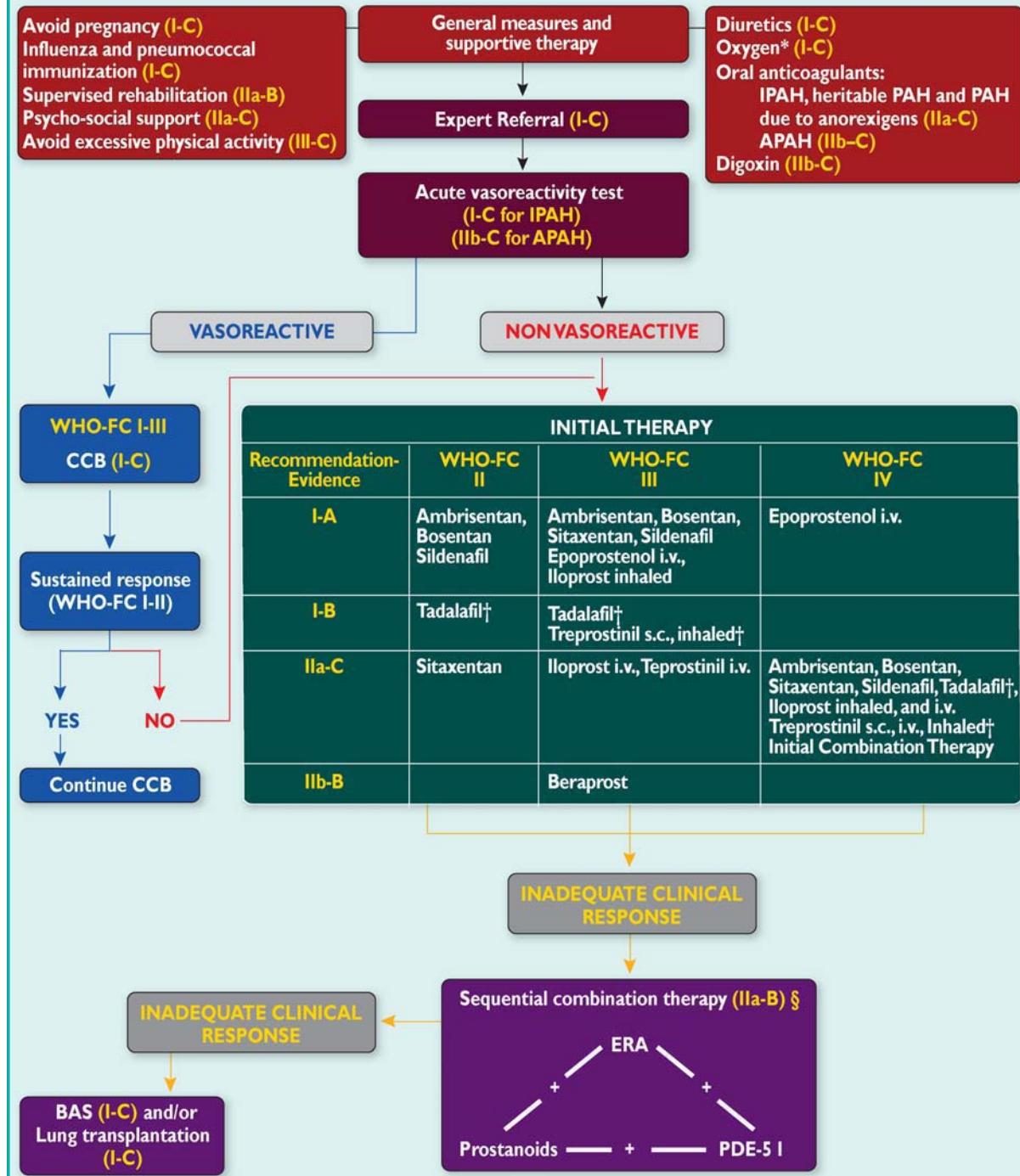


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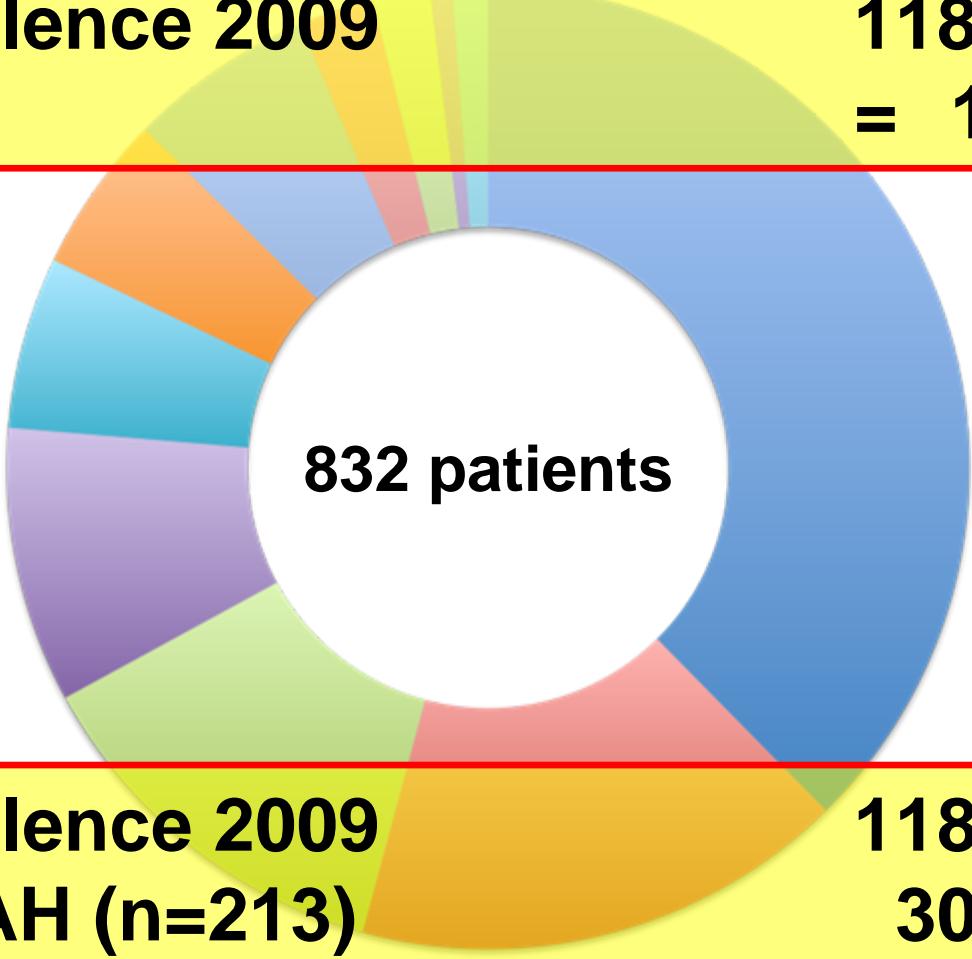


Swiss PH Registry: number of patients (12.2009)

Prevalence 2009

118 / million inhab.
= 1:8475

ZH-LUSZ



Prevalence 2009
IPAH (n=213)

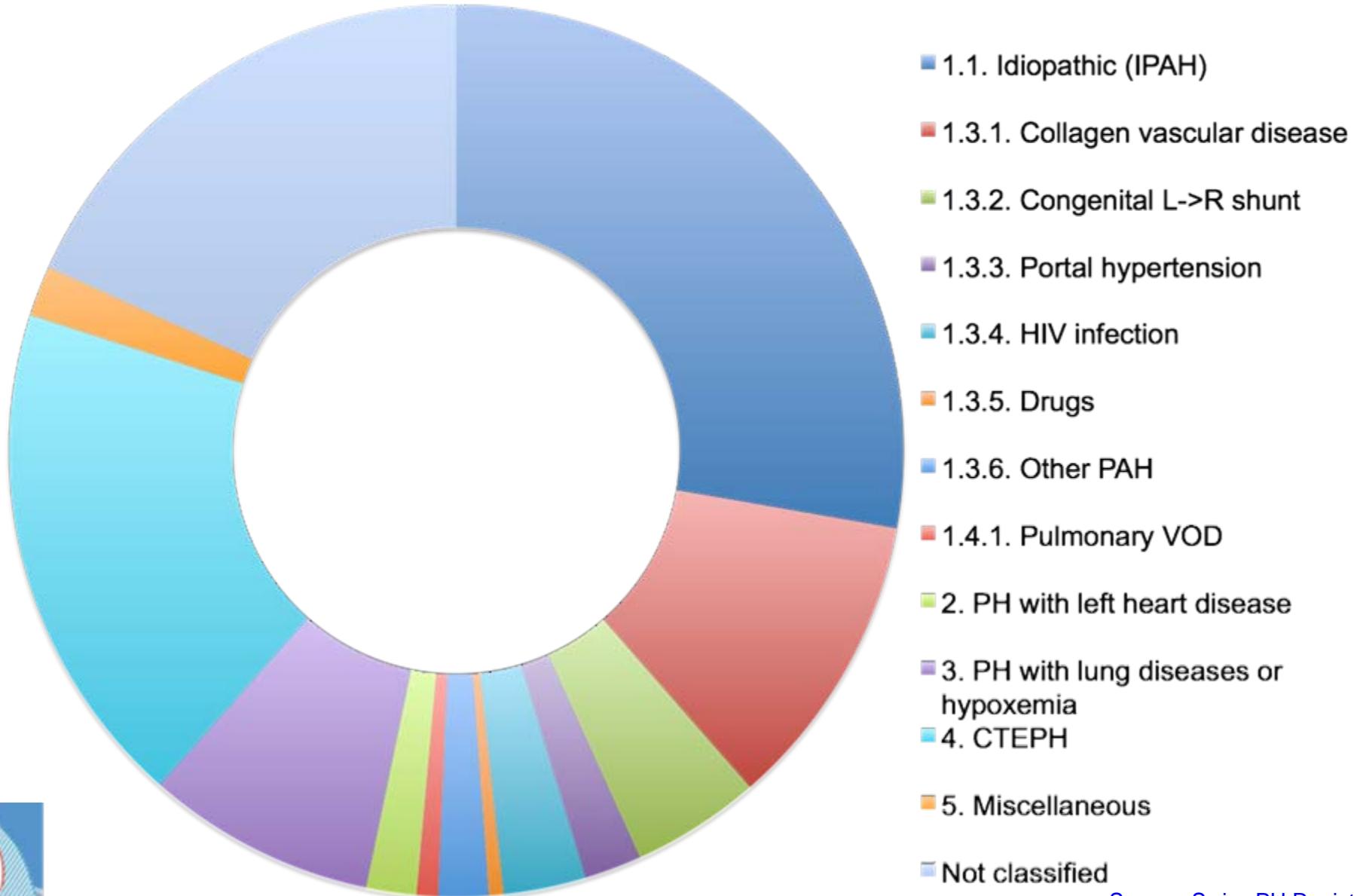
Incidence (mean 2004-2009)

118 / million inhab.
30 / million inhab.
23 / million inhab./ year

SOURCE: Swiss PH Registry

Swiss PH registry: PAH by aetiology

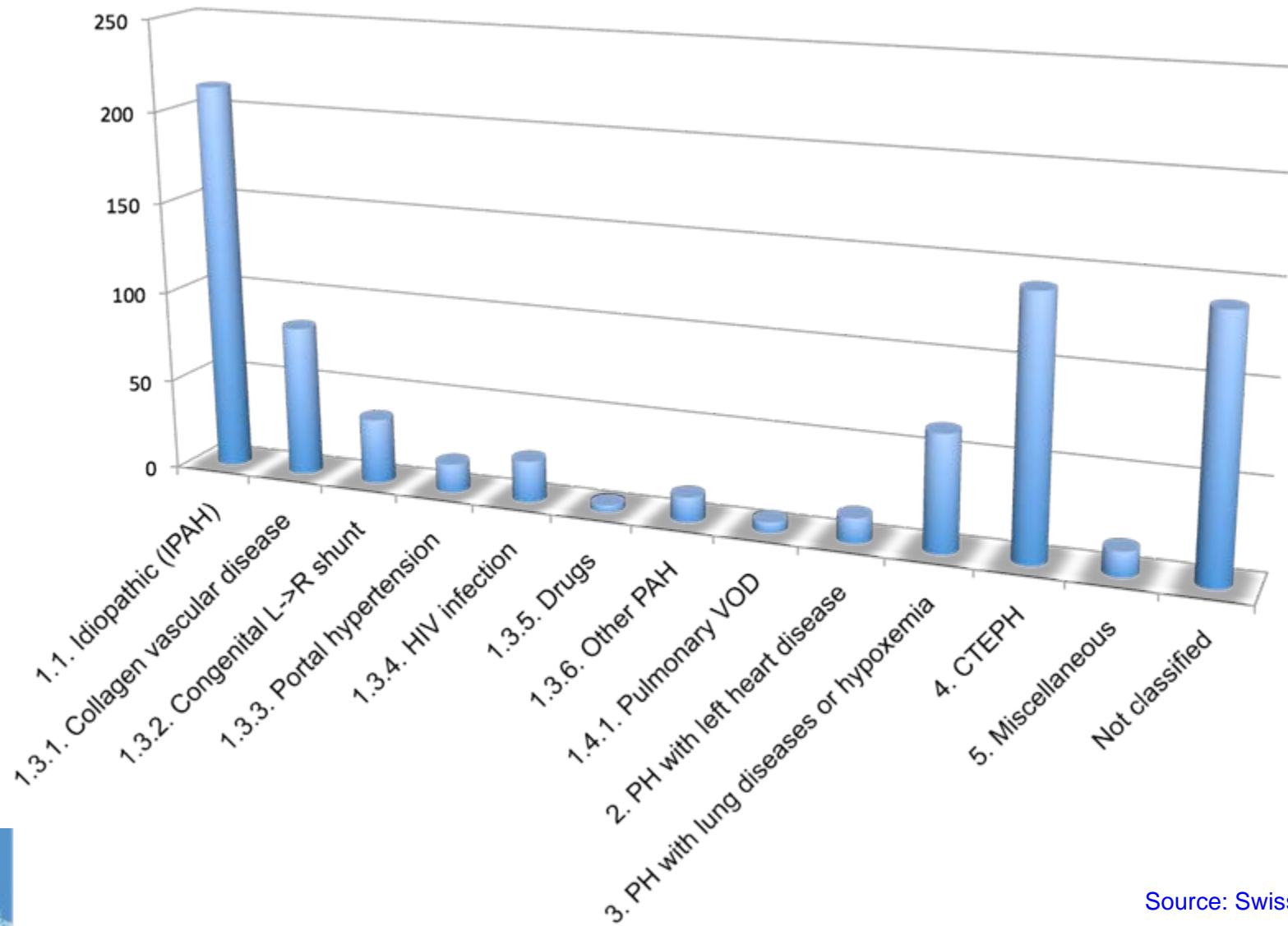
(according to WHO classification, status 12.2009)



Source: Swiss PH Registry

Swiss PH registry: PAH by aetiology

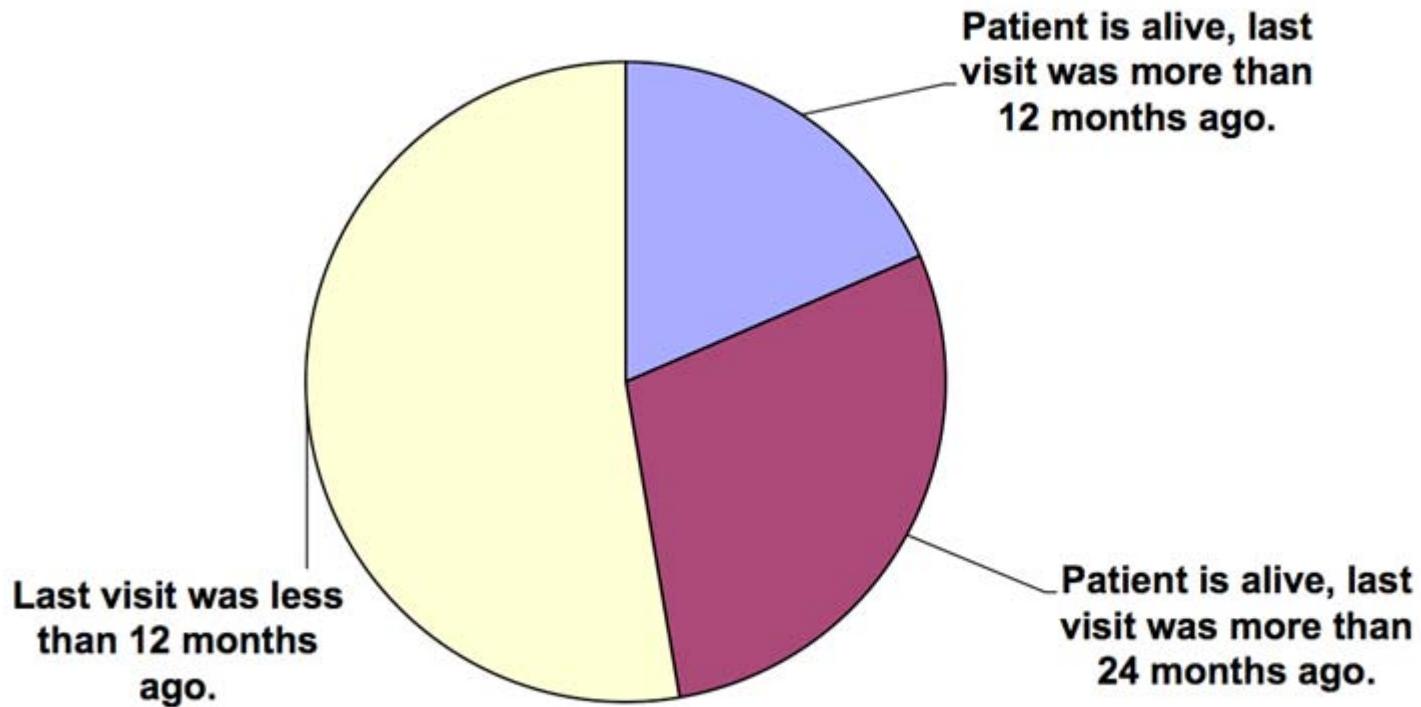
(according to WHO classification, status 12.2009)



Source: Swiss PH Registry



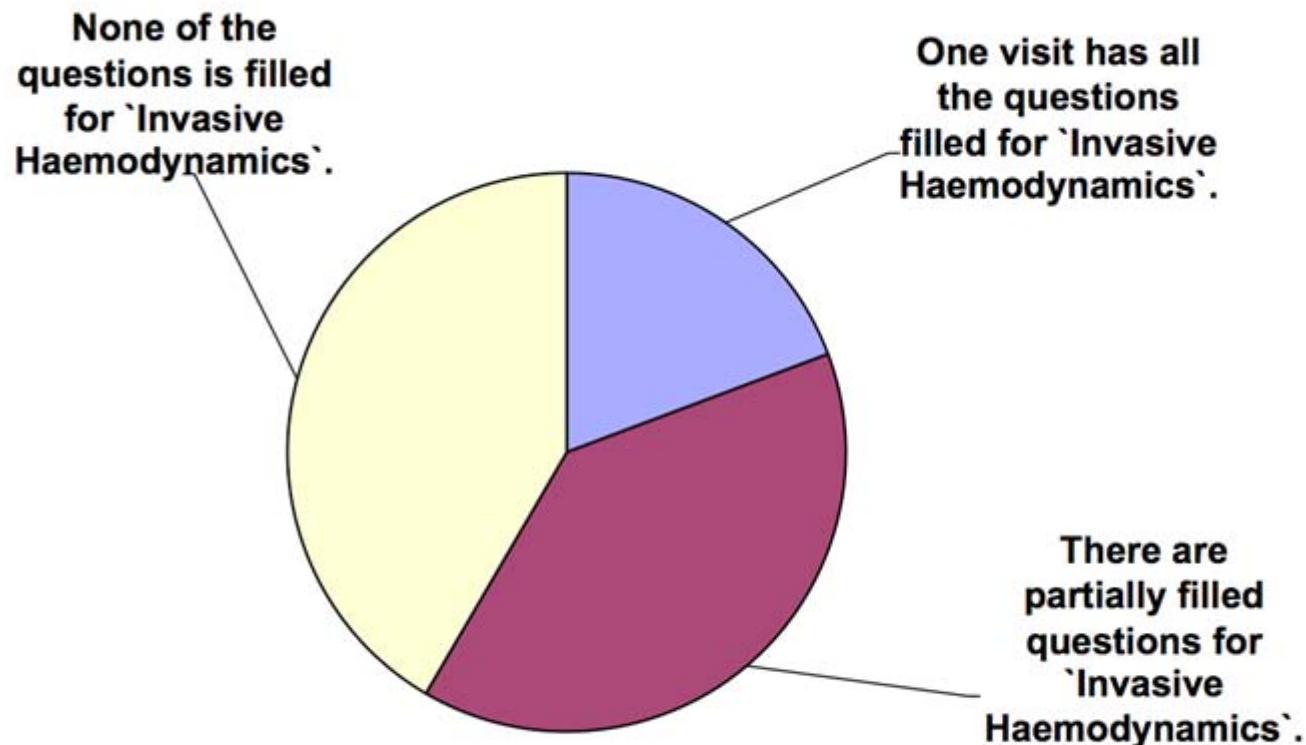
Swiss PH registry: demographic status (12.2009)



Source: Alabus / Swiss PH Registry

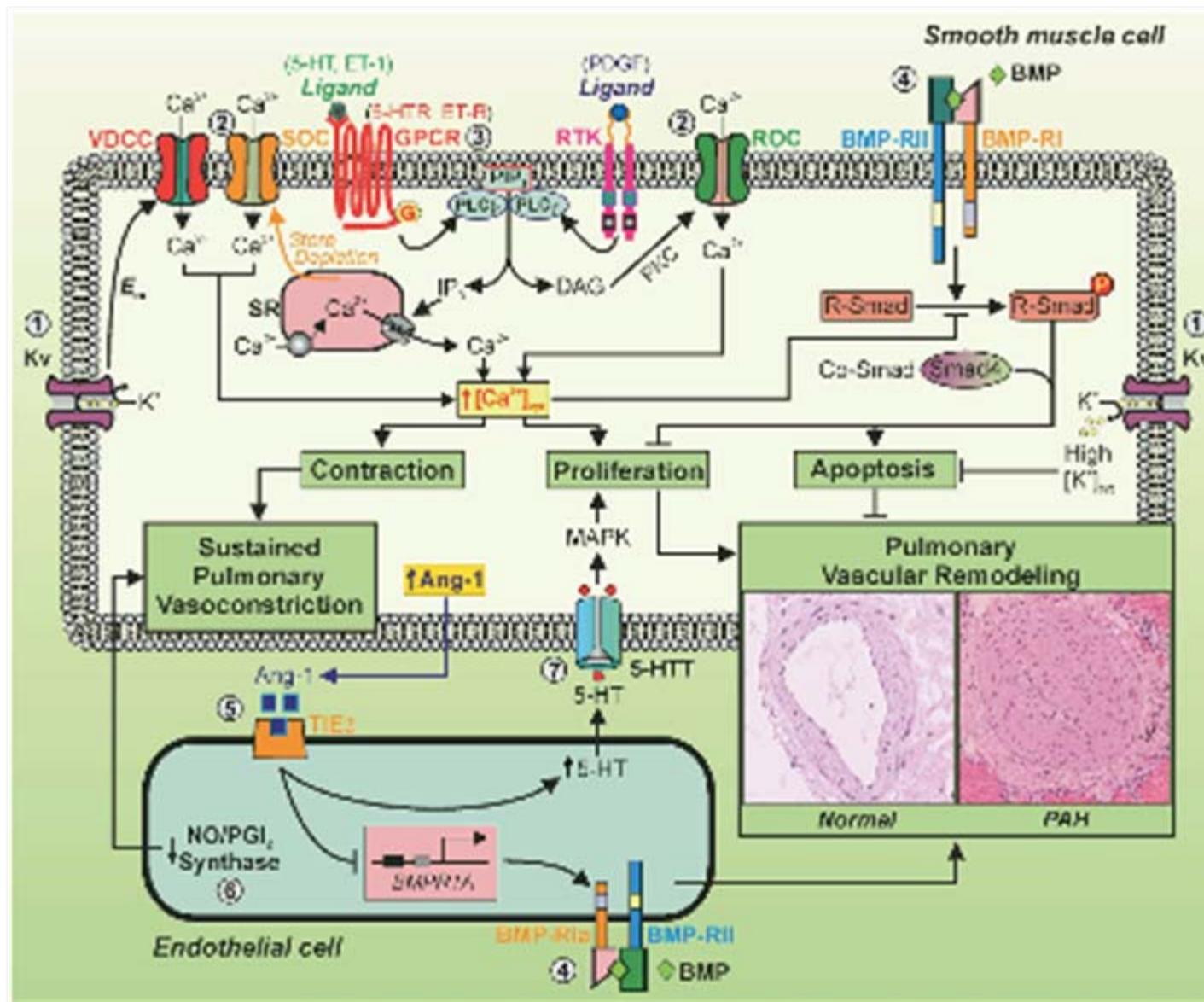


Swiss PH registry: baseline right heart catheter (12.2009)





PAH pathogenesis: relevant pathways



Conclusion / 1



- pulmonary arterial hypertension is a rare disease
- a national PAH register is crucial for a better knowledge of the disease
- the PAH-register provide us information on:
 - epidemiology and prevalence of PAH (different subgroups) in Switzerland
 - clinical course of the disease
 - treatment outcome
 - (time to clinical worsening)
 - **survival**

Conclusion / 2



Data from PAH-registers have limitation and need correlation to RCT

Is a good quality control tool:

- comparaison with data from other national registries or from major international centers
- comparaison between swiss centers

Knowledge on improvements during time,
specially in the era of specific PH therapies

Conclusion / 3



All PAH patients should be included in a national registry.

The SSHP-centres have access to the swiss PAH-registry.

All swiss PAH patients should be referred to a SSPH-centre:

- for the initial work-up if PAH is suspected
- to establish indication for treatment
- for follow-up visits and register-update
(at least once a year)

Centres HTAP

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Société Suisse sur l'Hypertension Pulmonaire SSHP
Società Svizzera di Ipertensione Polmonare SSIP
Swiss Society for Pulmonary Hypertension SSPH



Centres

Bâle

Berne

Genève

Lausanne

Locarno

Zurich

Partenaires

Aarau

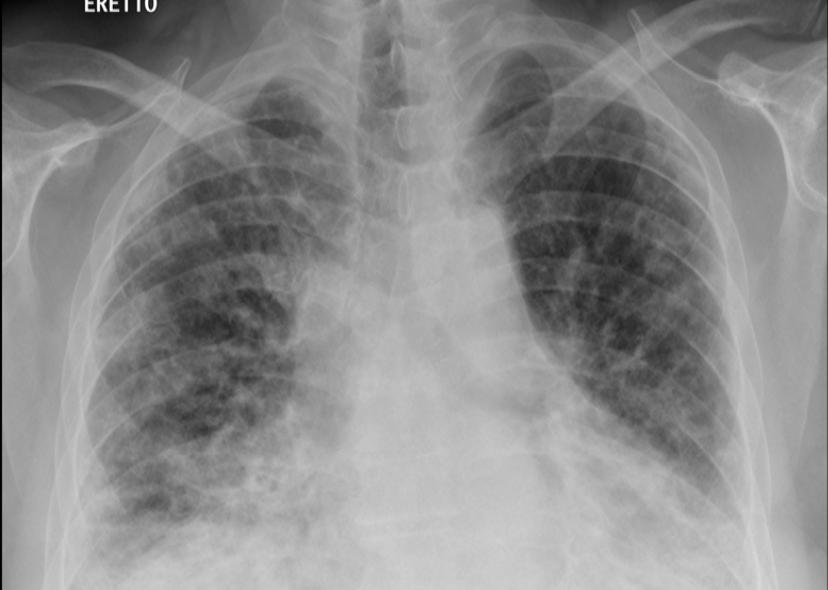
EOC

Bellinzona-Lugano-Mendrisio

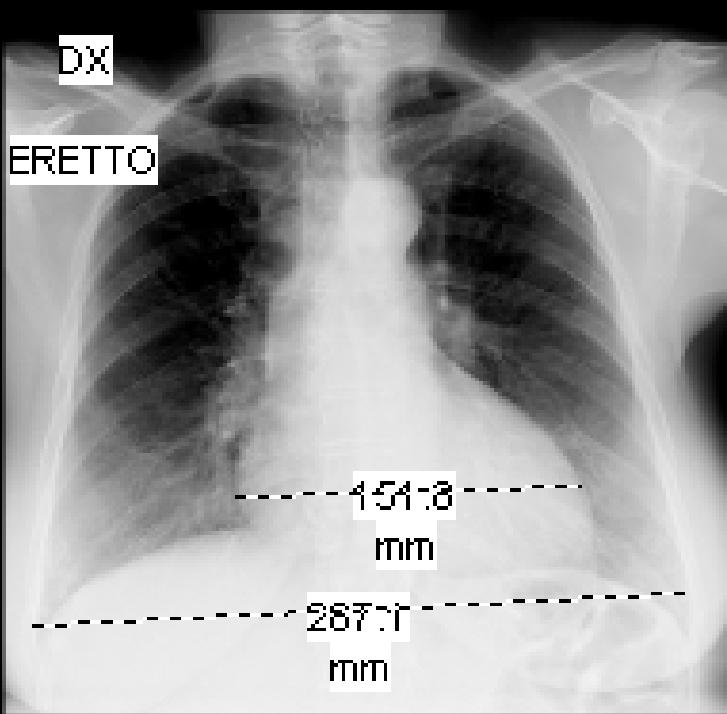
Luzern

Neuchâtel

St. Gall



32



MALATTIA REUMATOLOGICA

AR, scleroderma,

lupus, SS, ...

Interstiziopatia?

Ipertensione polmonare?

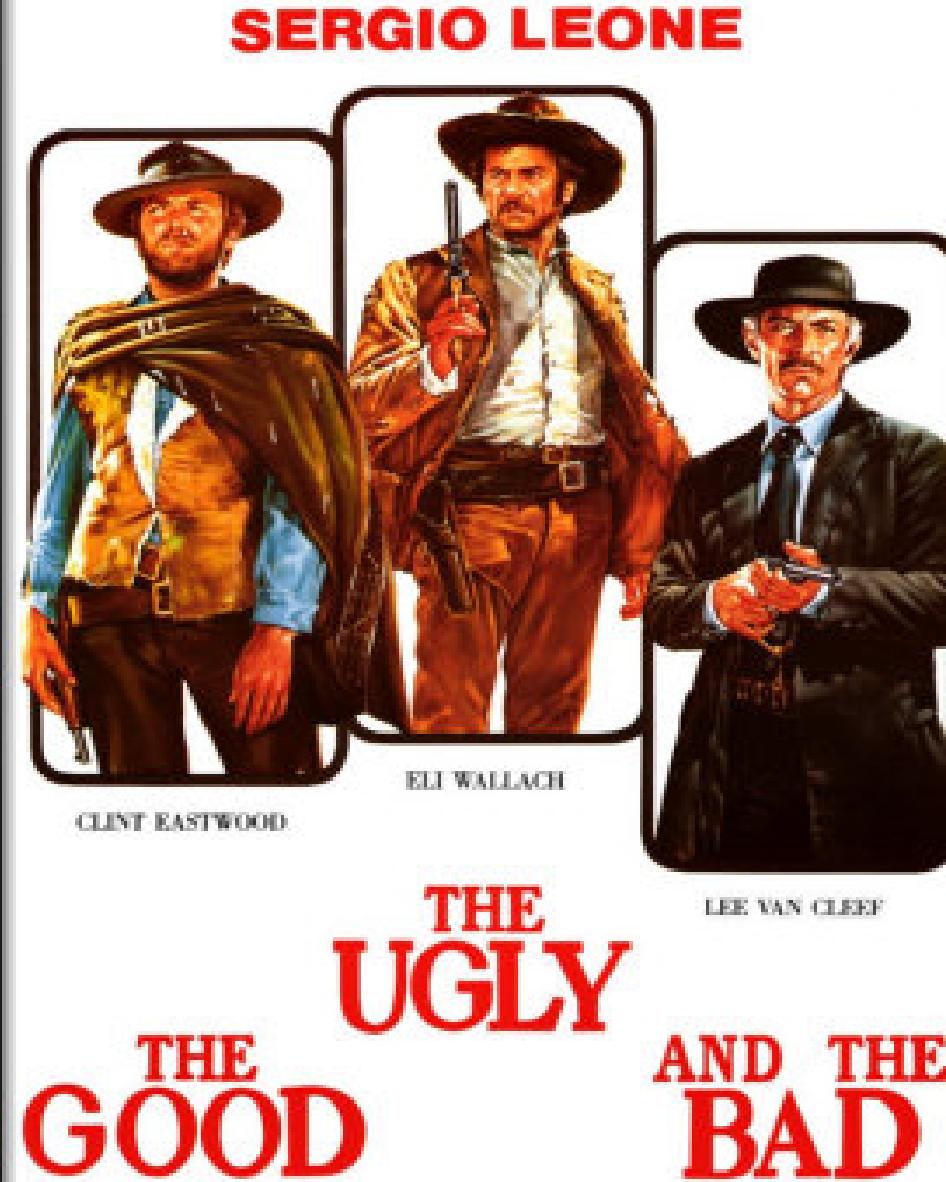
BILANCIO SPECIALISTICO!

SINTOMI RESPIRATORI

dispnea allo sforzo

tosse

Updated Classification of PH



Simonneau G et al,
J Am Coll Cardiol 2009; 53:



Grazie per l'attenzione!

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