Imperial College London

100 years of living science

Pulmonary Hypertension Registries

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What is a registry?

"an organised system that uses observational methods to collect uniform data on a population defined by a particular disease, condition, or exposure, and that is followed over time."

European Medicines Agency, 2015

Survival in Patients with Primary Pulmonary Hypertension

Results from a National Prospective Registry

Gilbert E. D'Alonzo, DO; Robyn J. Barst, MD; Stephen M. Ayres, MD; Edward H. Bergofsky, MD; Bruce H. Brundage, MD; Katherine M. Detre, MD; DrPH; Alfred P. Fishman, MD; Roberta M. Goldring, MD; Berton M. Groves, MD; Janet T. Kernis, MPH; Paul S. Levy, ScD; Giuseppe G. Pietra, MD; Lynne M. Reid, MD; John T. Reeves, MD; Stuart Rich, MD; Carol E. Vreim, PhD; George W. Williams, PhD; and Margaret Wu, PhD

■ *Objective:* To characterize mortality in persons diagnosed with primary pulmonary hypertension and to investigate factors associated with survival.

■ *Design:* Registry with prospective follow-up.

■ *Setting:* Thirty-two clinical centers in the United States participating in the Patient Registry for the Characterization of Primary Pulmonary Hypertension supported by the National Heart, Lung, and Blood Institute.

■ *Patients:* Patients (194) diagnosed at clinical centers between 1 July 1981 and 31 December 1985 and

Primary pulmonary hypertension has been considered a progressive, intractable, and often fatal disease (1-4). Further, treatments used late in its course, such as continuous prostacyclin infusion or heart-lung transplant, are expensive and of limited availability. These factors necessitate some form of priority assignment for patients. Prioritization is based primarily on prognosis and severity of disease, among other factors.

Although estimates of survival in primary pulmonary hypertension are often pessimistic (1-4), considerable variation in these estimates does occur (5-8), as does

D'Alonzo GE, *et al. Ann Intern Med* 1991; 115:343-9

How can registries help above clinical trials?

"Real world" patients in diverse settings:

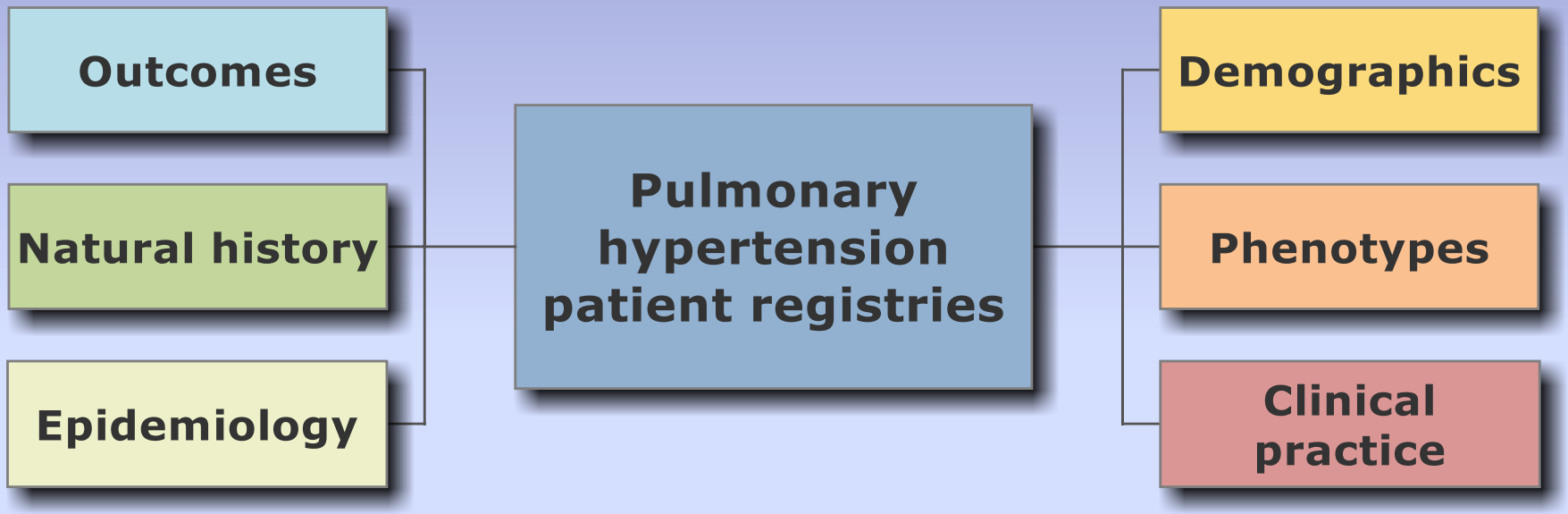
- **Elderly and children**
- **Multiple comorbidity**
- **Different countries**

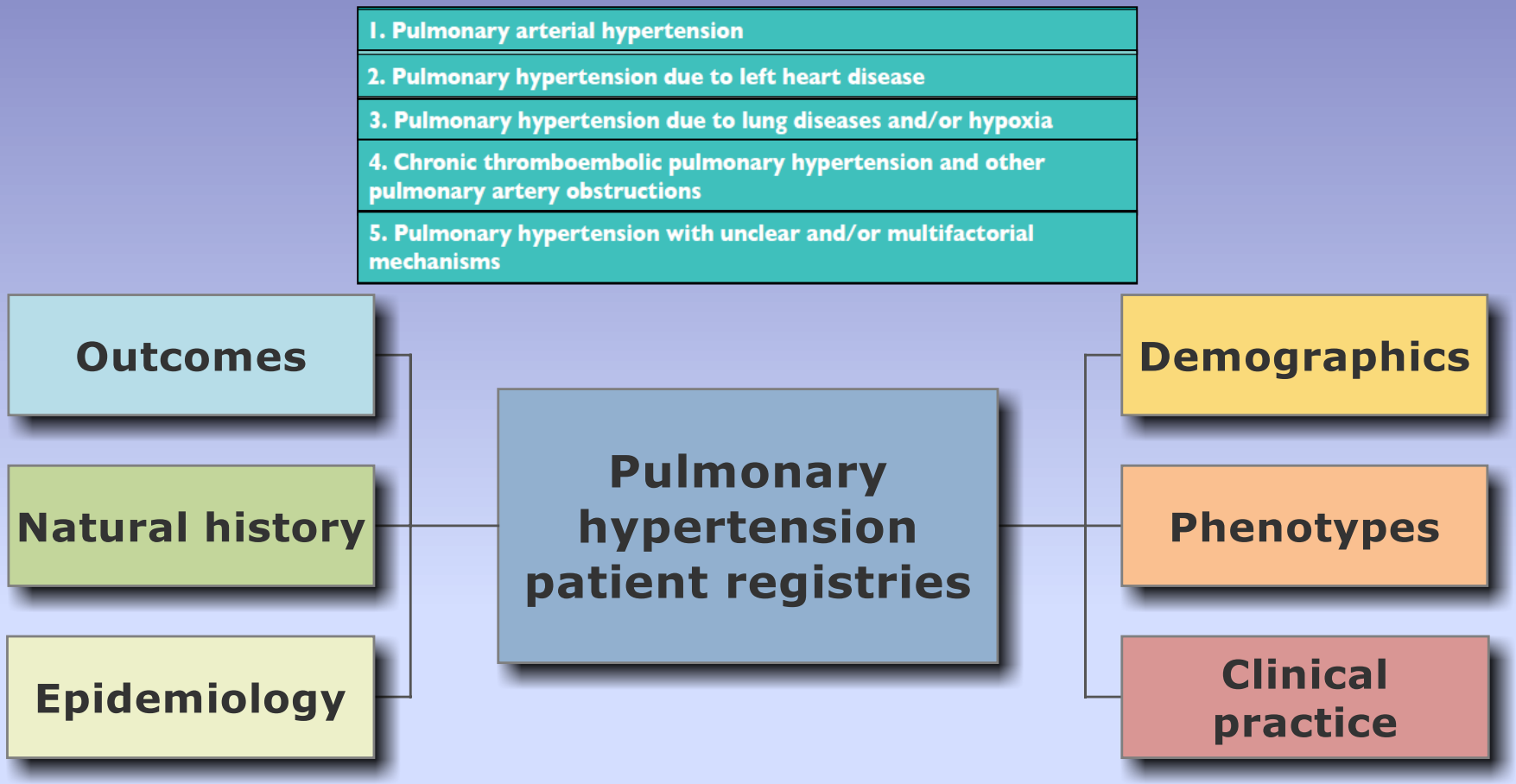
Describing clinical practice
Filling in gaps in the evidence
Understanding how evidence is applied

Answering questions not amenable to clinical trials

Assessment of novel interventions

Limits to conceptual understanding of real-world data


Outcomes
Natural history
Epidemiology
Pulmonary hypertension patient registries
Demographics
Phenotypes
Clinical practice


1. Pulmonary arterial hypertension
2. Pulmonary hypertension due to left heart disease
3. Pulmonary hypertension due to lung diseases and/or hypoxia
4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions
5. Pulmonary hypertension with unclear and/or multifactorial mechanisms
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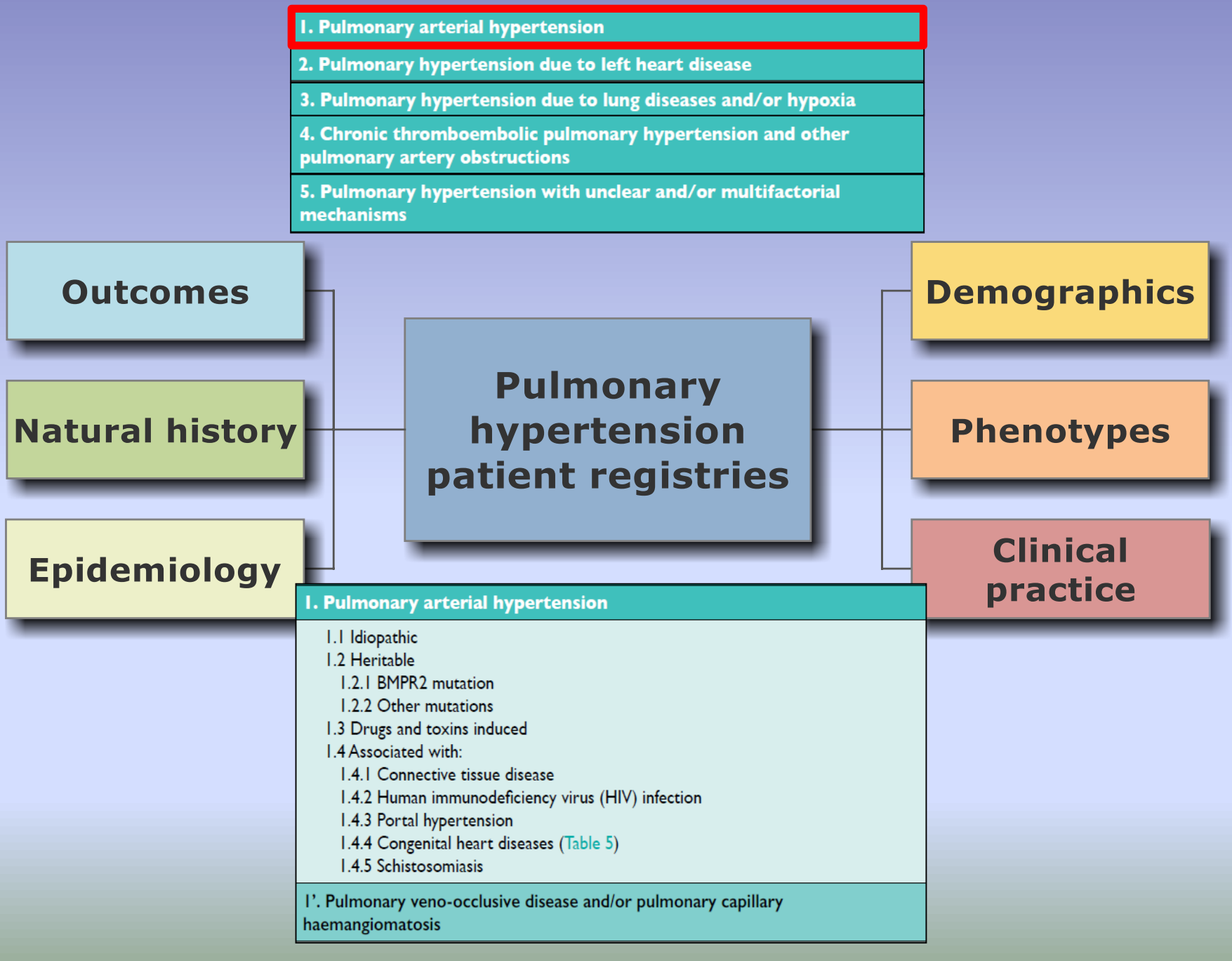
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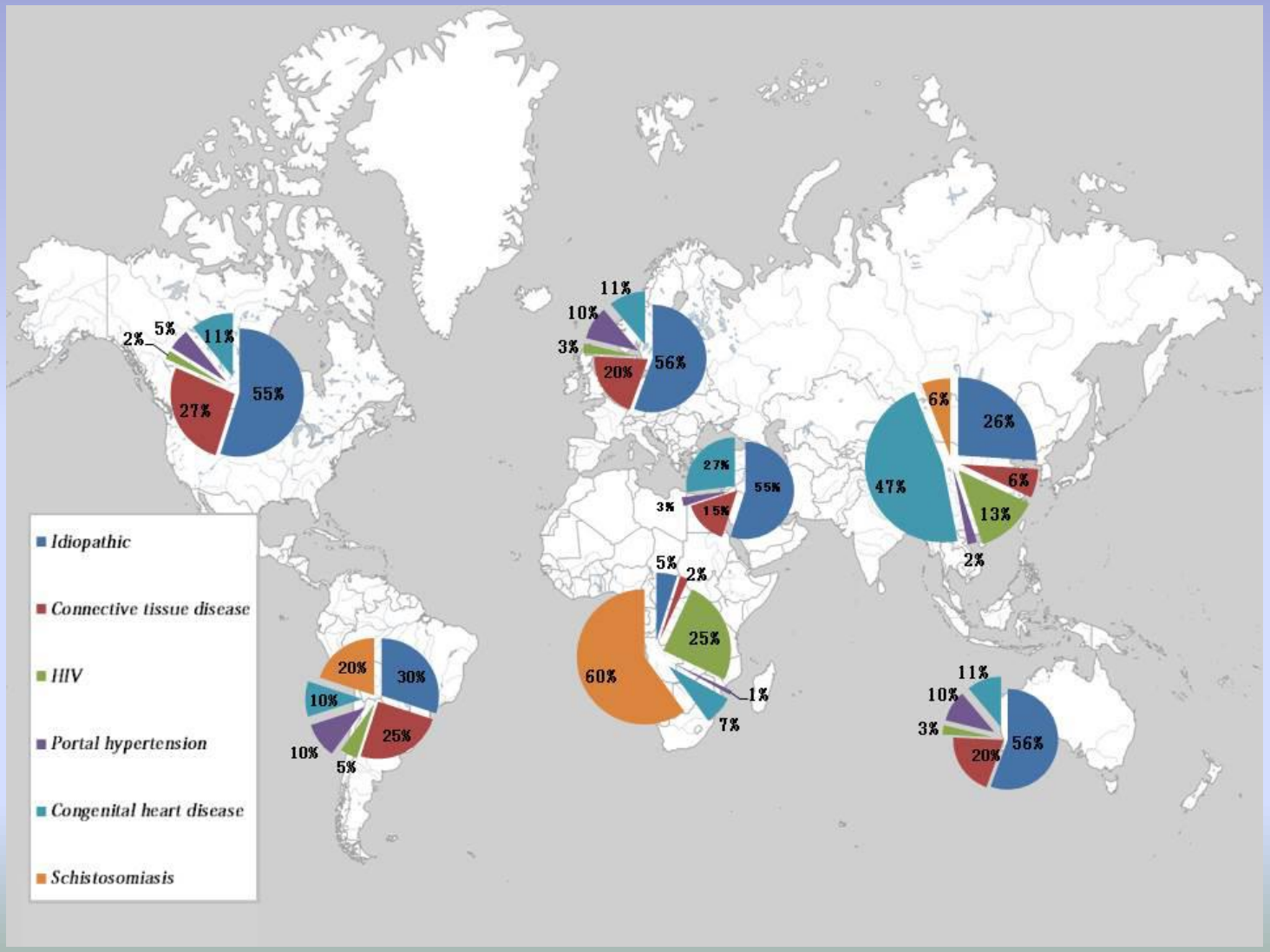
1. Pulmonary arterial hypertension
1.1 Idiopathic
1.2 Heritable
1.2.1 BMPR2 mutation
1.2.2 Other mutations
1.3 Drugs and toxins induced
1.4 Associated with:
1.4.1 Connective tissue disease
1.4.2 Human immunodeficiency virus (HIV) infection
1.4.3 Portal hypertension
1.4.4 Congenital heart diseases (Table 5)
1.4.5 Schistosomiasis
1'. Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis

Estimated global distribution of the most prevalent forms of PAH



Hoeper MM et al 2016

Epidemiology and outcomes of PAH reported from registries from various countries

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Columbia University registry[9]	1994–2002	84	81%	42 (14)	..	..	87%	75%
Scottish morbidity records*[10]	1986–2001	374	70%	52 (12)	7·6	26·0	..	..
The International Primary Pulmonary Hypertension study, Belgium†[11]	1992–94	24	..	..	1·7	..	..	..
French registry‡[5,12,13]	2002 and 2003	674	65%	50 (15)	2·4	15·0	89%	55%
REVEAL§[14–17]	2006 and 2007	2967	80%	50 (14)	2·0	10·6	91%	75%
Spanish registry¶[18]	1998–2008	886	71%	45 (17)	3·7	16·0	89%	77%
UK and Ireland registry‖[19]	2001–09	482	70%	50 (17)	1·1	6·6	93%	73%
Danish registry[20]	2000–12	134	58%	50 (21)	..	..	86%	73%
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UK National Audit 2014[22]	2004–14	2940	65%	Female: 60 (··), male: 58 (··)	..	..	86%‖	63%‖
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Data are mean (SD). *Inclusion age was restricted to 16–65 years. †Inclusion age was restricted to 18–70 year. ‡Inclusion age was 18 years or older. §Inclusion age older than 3 months, only 16% of the patients were incident. ¶Inclusion age in inclusion criteria not stated. ‖Only idiopathic, heritable, and drug-associated pulmonary arterial hypertension.

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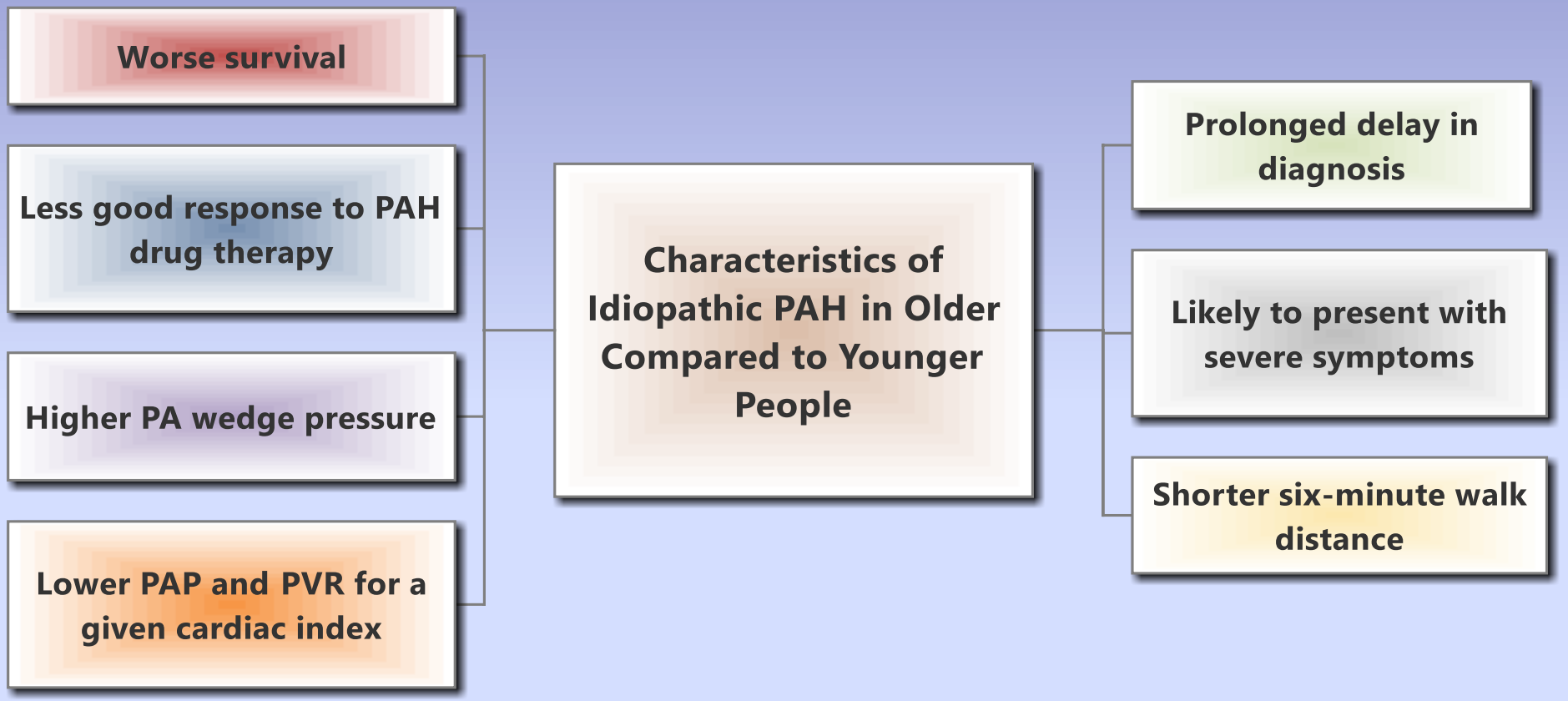

Hoeper M et al 2012; Ling Y et al 2012; Charalampopoulos A et al 2013

Haemodynamics at presentation in the Global TOPP Registry (tracking outcomes and practice in **paediatric** pulmonary hypertension) in 19 countries

Mean age 7 years, range 0 – 17 years
57% iPAH, 38% congenital heart disease, 10% chronic lung disease

	All patients (N = 480)	Class I (N = 62)	Class II (N = 227)	Class III (N = 158)	Class IV (N = 33)
No. patients with HC (n)	472	62	226	155	29
RA pressure (mm Hg)	7.1 (3.77)	6.5 (2.94)	7.0 (3.90)	7.1 (3.43)	8.9 (5.43)
	0–25	0–14	0–25	1–17	0–21
Mean pulmonary arterial pressure (mm Hg)	57.8 (18.80)	49.1 (15.74)	57.8 (19.66)	60.7 (17.36)	61.3 (20.49)
	25–143	25–83	25–143	26–126	27–103
Systemic arterial pressure (mm Hg)	68.0 (14.41)	63.9 (13.86)	68.2 (14.59)	68.9 (14.17)	70.4 (14.71)
	37–113	38–103	38–112	37–112	44–113
Cardiac index (L/min/m^2)	3.7 (3.74)	3.7 (1.54)	3.6 (1.61)	4.0 (6.14)	3.4 (1.48)
	1–75	1–12	1–11	1–75	1–8
PAP/SAP	0.9 (0.27)	0.8 (0.21)	0.9 (0.28)	0.9 (0.27)	0.9 (0.30)
	0.3–2.4	0.4–1.3	0.4–1.9	0.3–2.4	0.4–1.6
PVRI (wood units $*$ m^2)	16.6 (11.62)	13.1 (11.61)	16.9 (12.48)	17.1 (9.85)	19.0 (12.54)
	3–96	3–89	3–96	3–49	4–56
SVRI (wood units $*$ m^2)	20.4 (10.82)	18.5 (11.61)	20.2 (10.55)	21.2 (10.51)	22.5 (12.45)
	1–89	5–89	5–85	1–59	6–59
PVRI/SVRI	0.9 (0.60)	0.7 (0.26)	0.9 (0.58)	0.9 (0.75)	0.9 (0.32)
	0.2–8.3	0.2–1.3	0.2–4.3	0.2–8.3	0.3–1.7

Data are mean (SD) and range unless stated otherwise.
RA: right atrial, PAP: mean pulmonary arterial pressure, SAP: mean systemic arterial pressure, PVRI: indexed pulmonary vascular resistance.
SVRI: indexed systemic vascular resistance.

Beghetti M et al 2016

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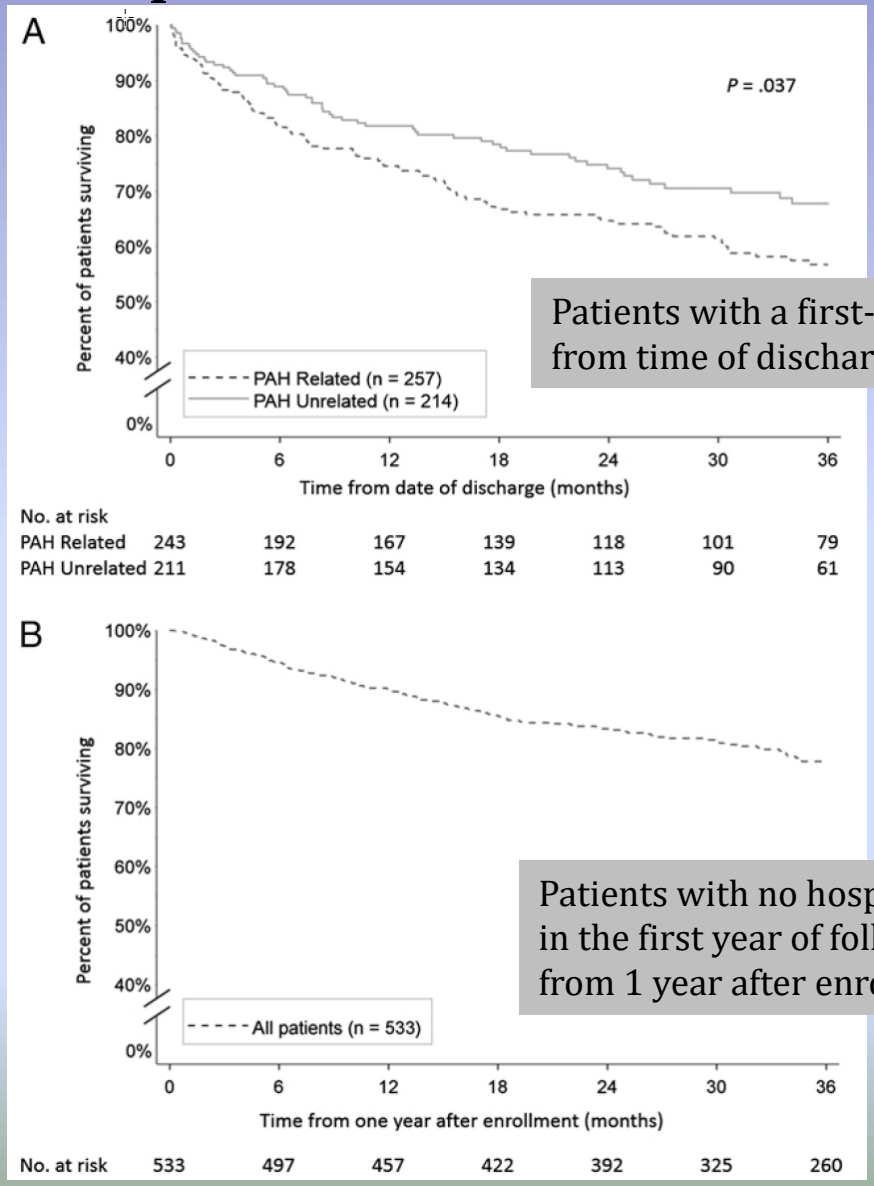
PAP/SAP	0.9 (0.27)	0.8 (0.21)	0.9 (0.28)	0.9 (0.27)	0.9 (0.30)
	0.3–2.4	0.4–1.3	0.4–1.9	0.3–2.4	0.4–1.6
PVRI (wood units $*$ m^2)	16.6 (11.62)	13.1 (11.61)	16.9 (12.48)	17.1 (9.85)	19.0 (12.54)
	3–96	3–89	3–96	3–49	4–56
SVRI (wood units $*$ m^2)	20.4 (10.82)	18.5 (11.61)	20.2 (10.55)	21.2 (10.51)	22.5 (12.45)
	1–89	5–89	5–85	1–59	6–59
PVRI/SVRI	0.9 (0.60)	0.7 (0.26)	0.9 (0.58)	0.9 (0.75)	0.9 (0.32)
	0.2–8.3	0.2–1.3	0.2–4.3	0.2–8.3	0.3–1.7

Data are mean (SD) and range unless stated otherwise.
RA: right atrial, PAP: mean pulmonary arterial pressure, SAP: mean systemic arterial pressure, PVRI: indexed pulmonary vascular resistance.
SVRI: indexed systemic vascular resistance.

Beghetti M et al 2016

Natural history: hospitalization in PAH in REVEAL predicts worse survival

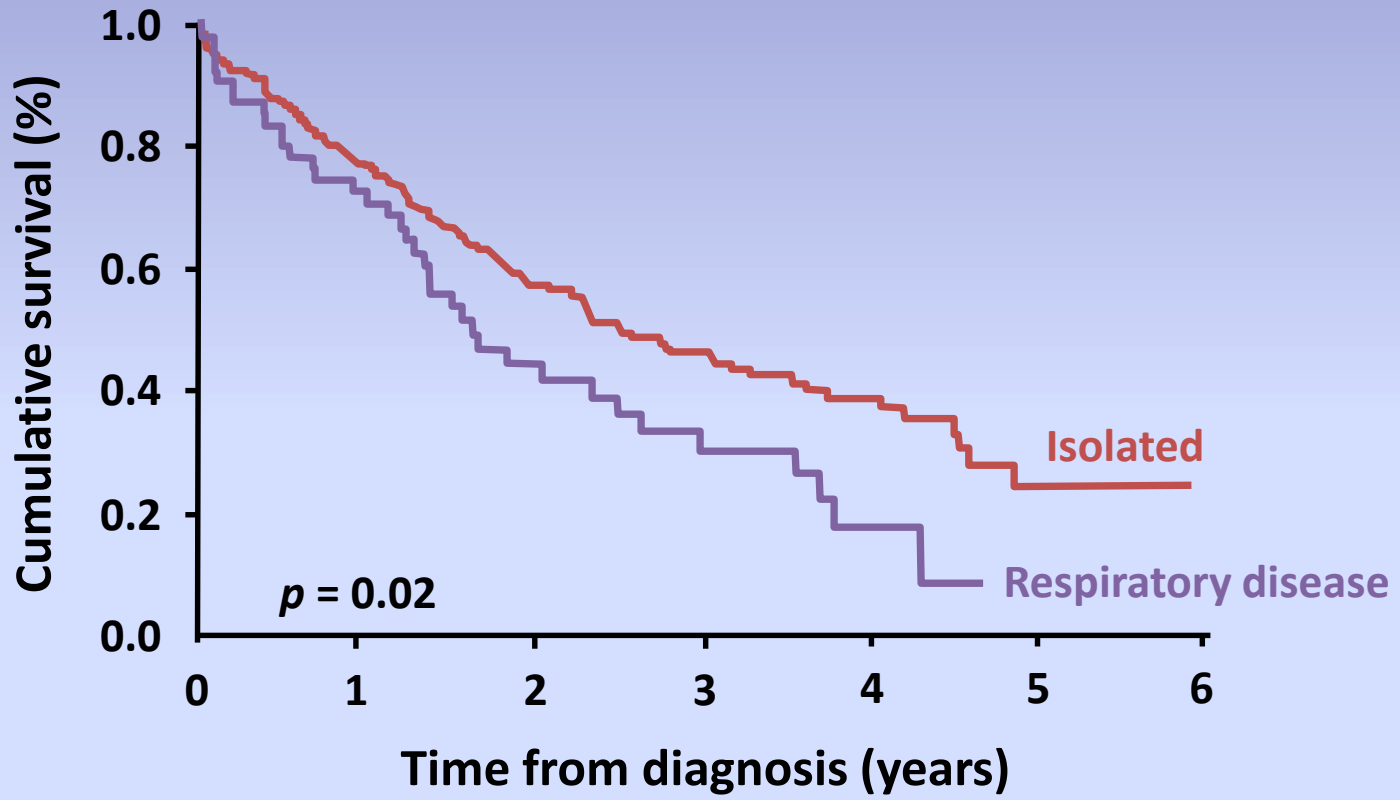


Patients with a first-time hospitalization from time of discharge

Patients with no hospitalization in the first year of follow-up from 1 year after enrolment

Burger CD et al 2014

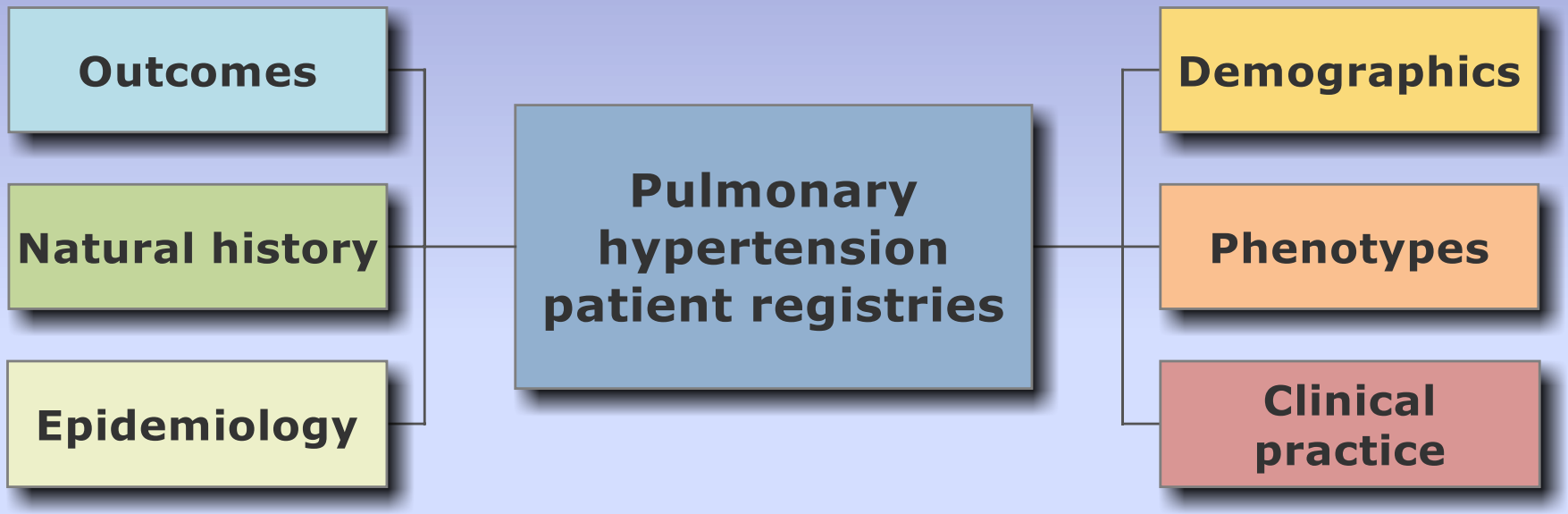
Survival in PAH associated with scleroderma with and without lung disease in the UK



Isolated	259	179	94	53	27	6	
Respiratory	56	38	18	10	3		

Condliffe R, et al. *Am J Respir Crit Care Med* 2009; 179:151–7

1. Pulmonary arterial hypertension
2. Pulmonary hypertension due to left heart disease
3. Pulmonary hypertension due to lung diseases and/or hypoxia
4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions
5. Pulmonary hypertension with unclear and/or multifactorial mechanisms



CTEPH Registries

Registry name (Ref.)	Country	Period	Number of Patients	% CTEPH	Prevalence	Incidence
ASPIRE registry (6)	UK	2001–2010	1,344	18	—	0.3–3.7*
National Audit 2012 (7)	UK	—	7,000	18.7	12.9–27.3	4.3–4.9
National Audit 2013 (8)	UK	—	7,757	19.2	10.8–38.4	4.4–4.6
Swiss registry (12)	Switzerland	1998–2012	996	25	—	—
Spanish registry (9)	Spain	1998–2008	1,028†	15.8	3.2	0.9
Portuguese registry (10)	Portugal	2008–2010	79‡	41.8	—	1.1
Condliffe et al (11)	UK	2001–2006	469§	100	—	1.4
CTEPH European registry (2)	Europe and Canada	2008–2010	679§	100	—	Up to 5.7‖

Definition of abbreviations: ASPIRE = *A*ssessing the *S*pectrum of *P*ulmonary Hypertension *I*dentified at a *Re*ferral Centre; CTEPH = chronic thromboembolic pulmonary hypertension.

*Evolution over the years.

†Registry including only PAH and CTEPH cases. Prevalence and incidence are expressed as cases per million inhabitants and per million inhabitants per year.

‡Registry including only incident PAH and CTEPH cases. Prevalence and incidence are expressed as cases per million inhabitants and per million inhabitants per year.

§Registry including only incident patients with CTEPH. Prevalence and incidence are expressed per million inhabitants per year.

‖Country with highest incidence (unpublished data from the European CTEPH registry).

Hurdman J 2012; Health & Social Care Information Centre 2012 and 2013; Escribano-Subias P 2012; Baptista R 2013; Condliffe R 2008; Mueller-Mottet S 2015; Pepke-Zaba J 2011

Patient characteristics in the International CTEPH Registry

	All Patients (n=679)	Operable Patients* (n=427)	Nonoperable Patients* (n=247)	*P* (Exploratory)
Gender, % male	50.1	53.4	44.5	0.0308
Ethnicity, % white	95.9	95.3	96.7	0.4277
Age, y, median [Q1;Q3]	63 [51; 72]	61 [48; 70]	67 [57; 74]	<0.0001
Weight, kg, median [Q1;Q3]	75 [65; 87]	76 [66; 88]	73 [63; 82]	0.0161
NYHA class, % I/II/III/IV	0.7/17.8/68.6/12.8	0.5/19.2/67.7/12.6	1.2/15.8/70.4/12.6	0.4922
6MWD, m, median [Q1; Q3] (n)	329 [245; 427] (589)	340 [250; 435] (373)	315 [223; 400] (214)	0.0219
Blood group non-0, % (n)	76.0 (366)	79.5 (249)	68.4 (117)	0.0255

Values are expressed as medians with first and third quartiles (Q1; Q3) or percentages; (n): patients with assessment; *P* values from Wilcoxon rank-sum test or Fisher exact test. NYHA indicates new York Heart Association; 6MWD, 6-minute walking distance.

*Five patients had no data on operability.

Pepke-Zaba J et al 2011

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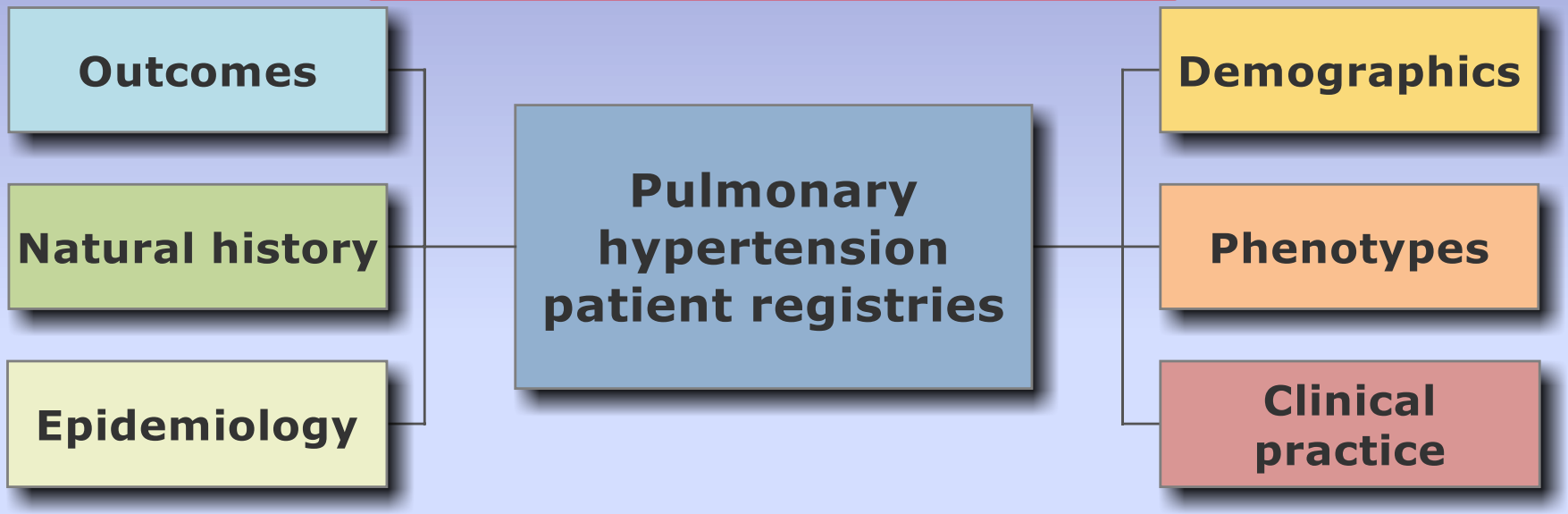
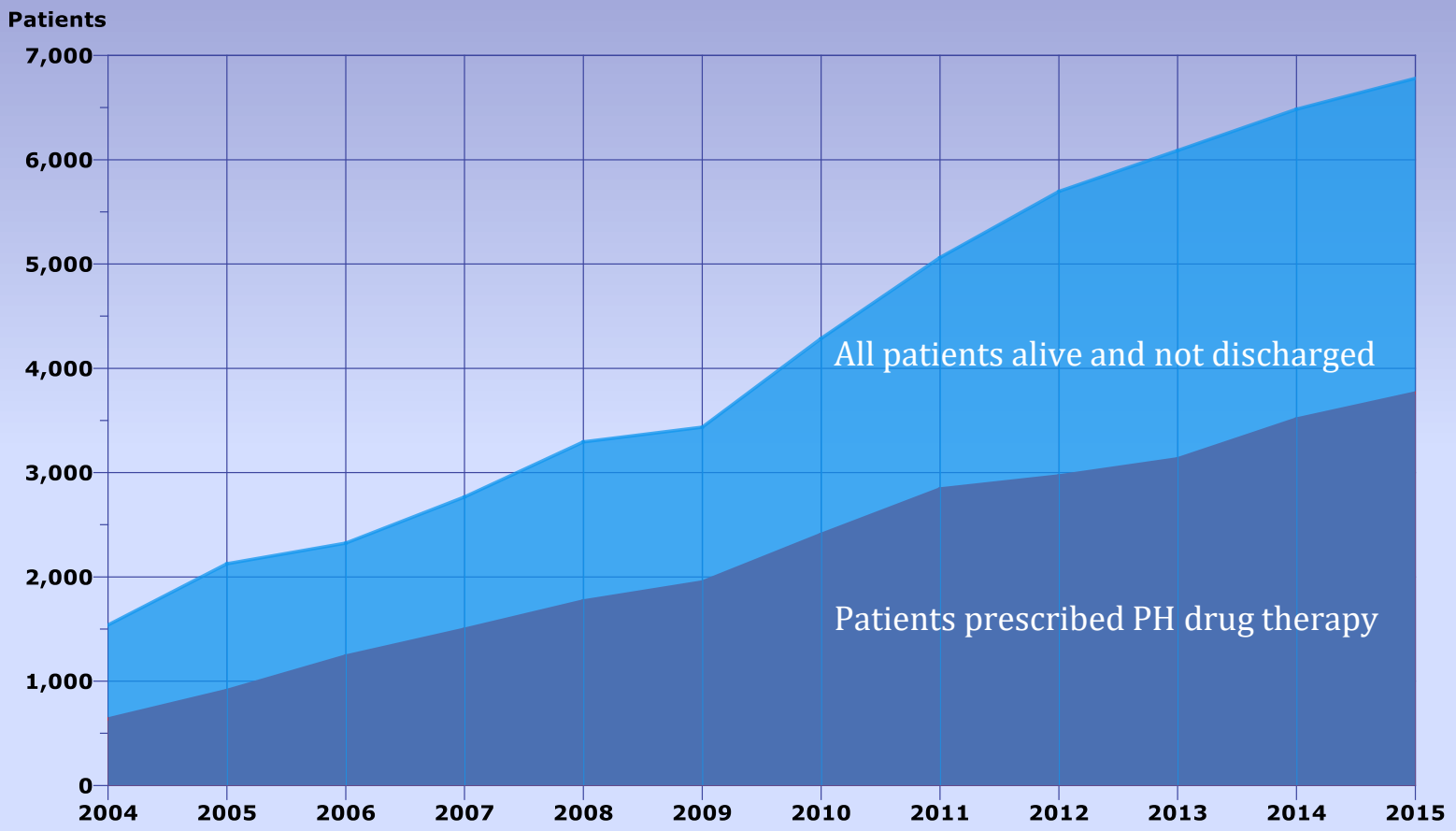
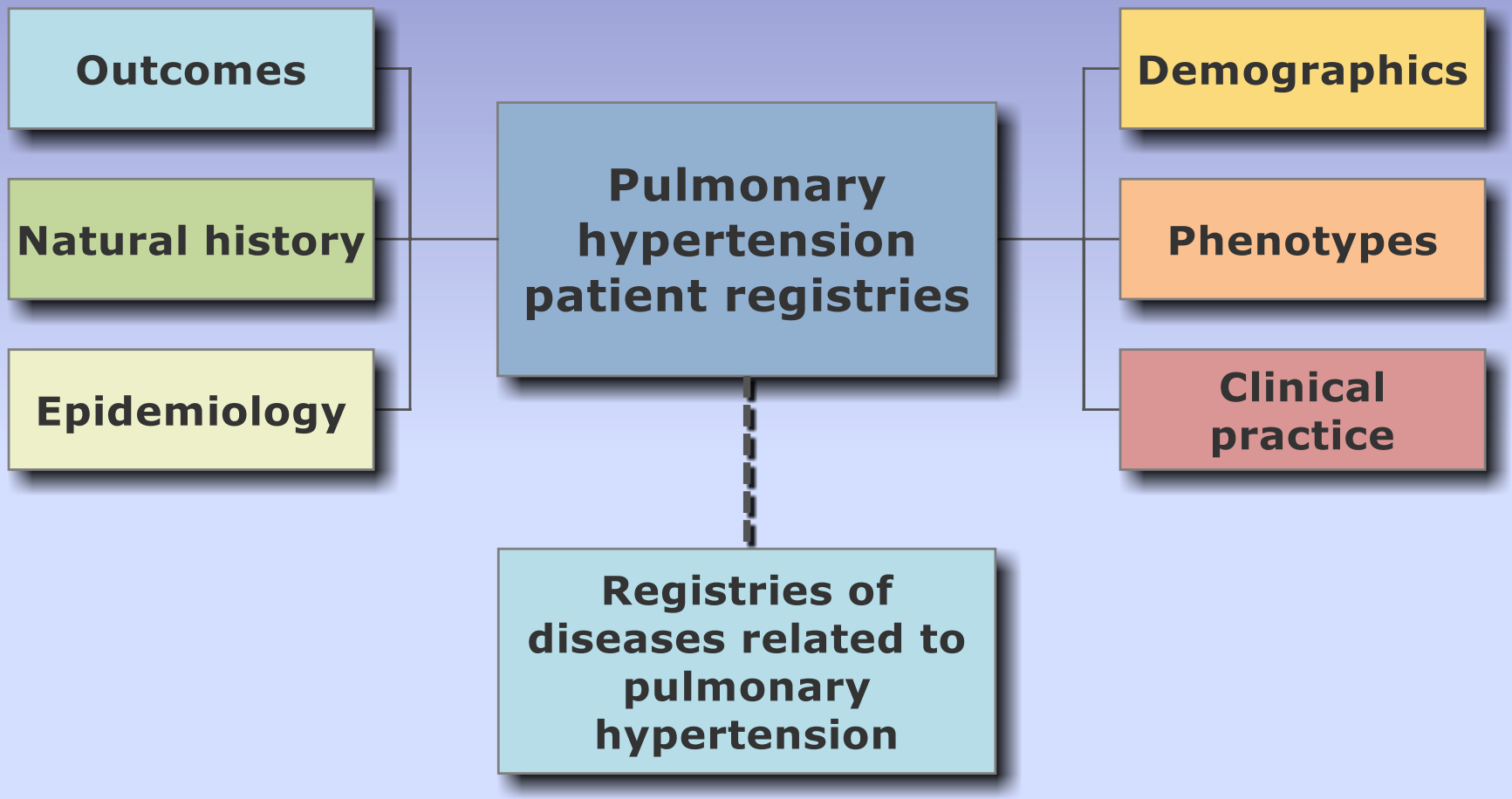


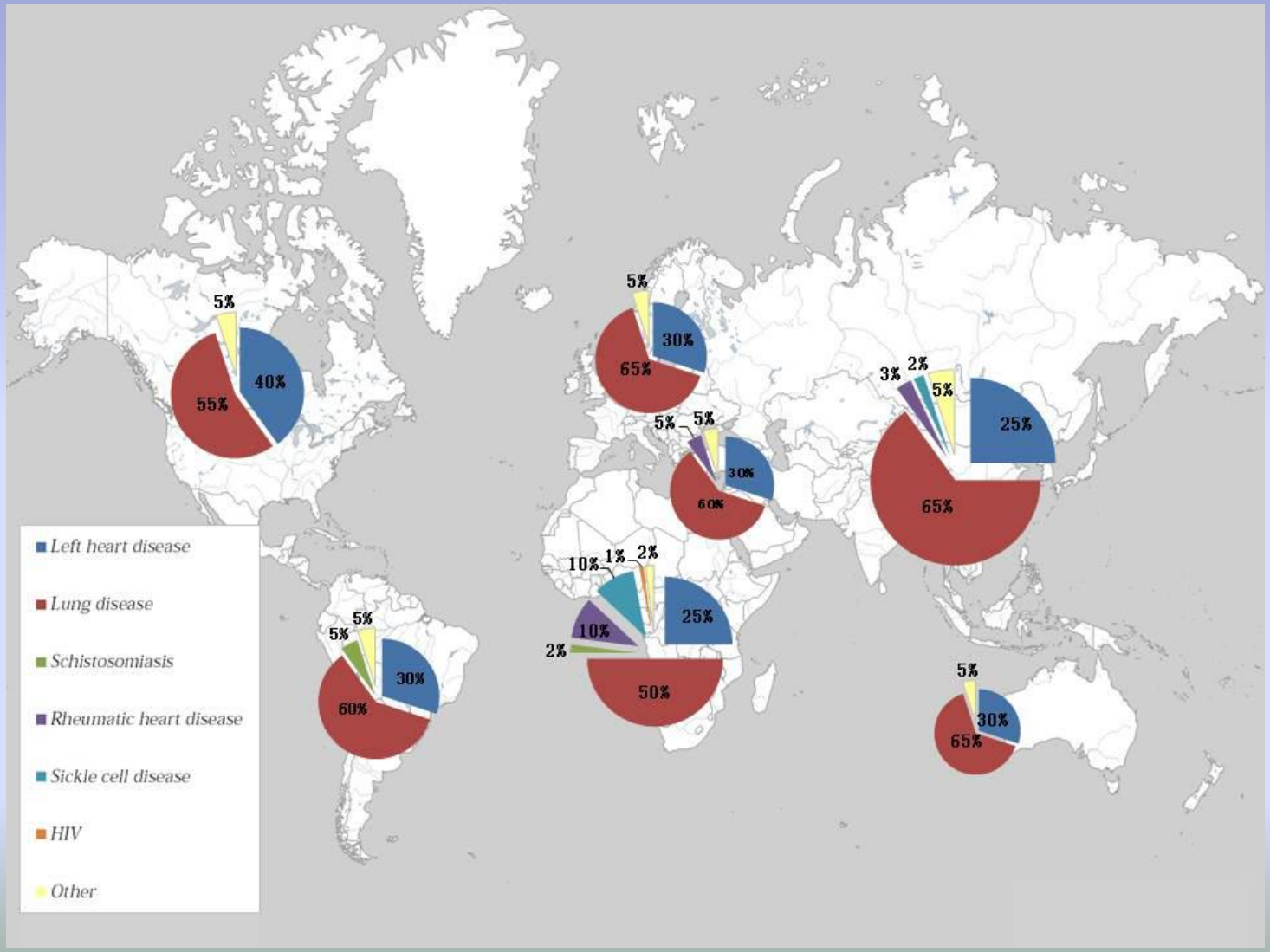
Growth of the UK Pulmonary Hypertension Service on 31st March for 12 years



Health & Social Care Information Centre 2016


Outcomes
Natural history
Epidemiology
Pulmonary hypertension patient registries
Demographics
Phenotypes
Clinical practice
Registries of diseases related to pulmonary hypertension

Estimated global distribution of the most prevalent forms of pulmonary hypertension



Hoeper MM et al 2016

Pulmonary hypertension registries into the future

- Registry of registries
- Strengthening observations and generalisability
- Linking registries to biobanks
- Individualizing patient management through big data