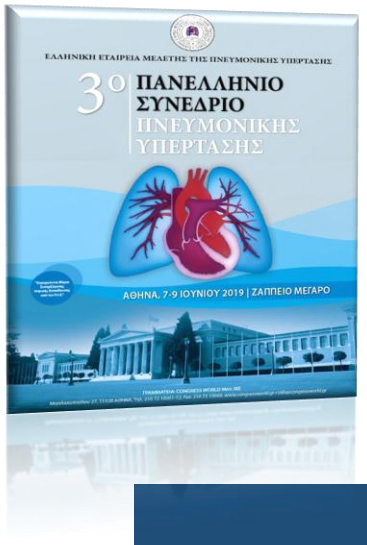


Hellenic PulmOnary HyPertension rEgistry (HOPE): Current data 2019

Pulmonary Arterial Hypertension

Arvanitaki Alexandra, MD, MSc, PhDc

1st Cardiology Department, AHEPA University Hospital

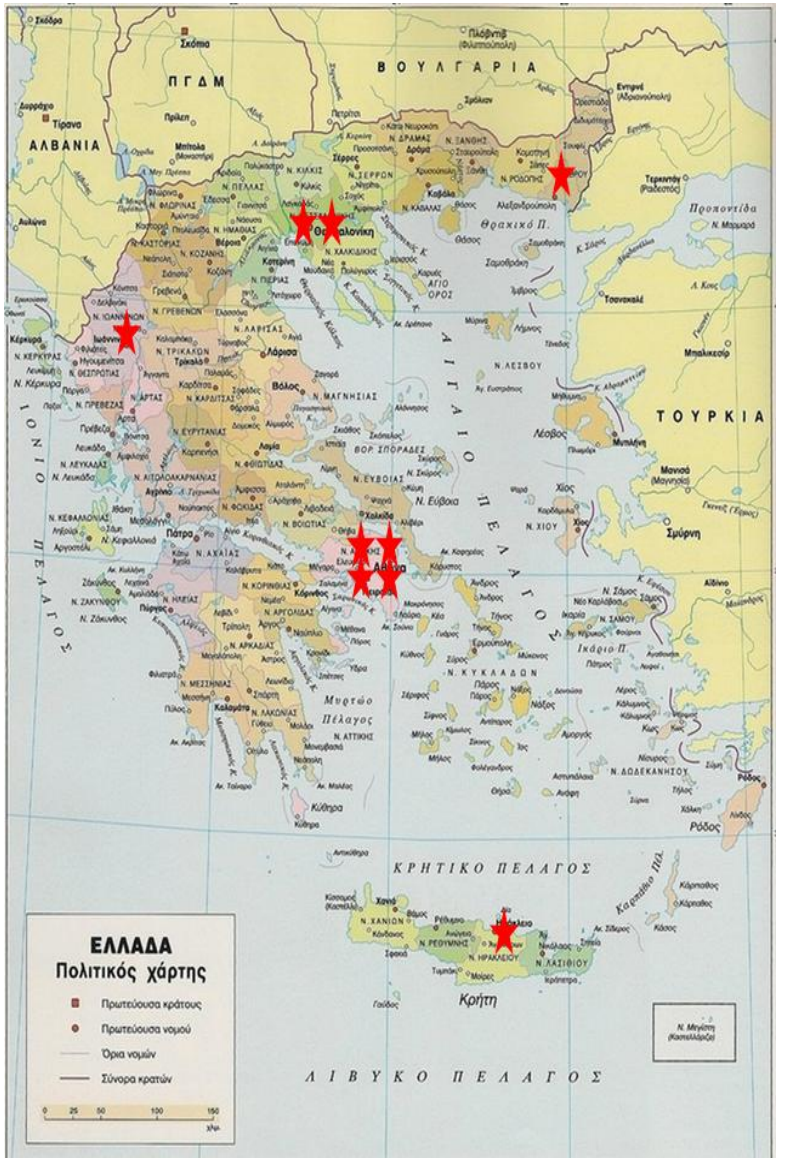
Greek PAHTool Registry

Hellenic PulmOnary Hypertension rEgistry (HOPE)
1. Expert Centers

Greek PAHTool Registry

10 expert centers in Greece

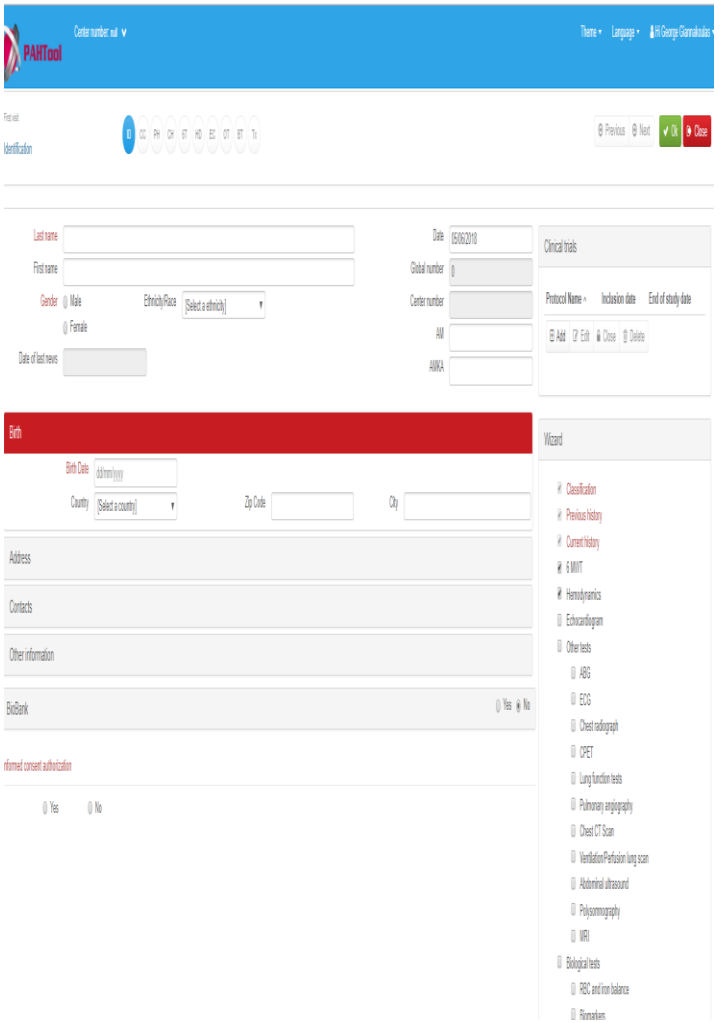
1. Cardiolody Department, AHEPA University Hospital, Thessaloniki
2. Multidisciplinary Pulmonary Hypertension Service, Attiko General Hospital, Athens
3. Cardiology- Pediatric Cardiology Department, Onassis Cardiac Surgery Center, Athens
4. Cardiology Department, Mediterraneo Hospital, Glifada, Athens
5. Respiratory Failure Unit, Papanikolaou General Hospital, Thessaloniki
6. Pulmonary Department, PAGNI, Irakleio, Crete
7. Cardiology Department, Hippokrateio General Hospital, Athens
8. **Laiko General Hospital, Athens**
9. Cardiology Department, Democritus University of Thrace, Alexandroupolis
10. 2nd Cardiology Department, Ioannina University Hospital



Hellenic PulmOnary Hypertension rEgistry (HOPE)
2. Methods and Data Management

Greek PAHTool Registry

- ✓ **PAH tool launched from 2015**
- ✓ **Retrospective and prospective data collection till April 2019**
  - PH diagnosis, classification, symptoms, comorbiditie s, diagnostic procedures and treatment
  - Patients who had full work up, including an RHC
  - First visit and Follow up visits
- ✓ **PAH tool**
  - **IRB approval**
  - **Informed consent** from all participants
  - Internet based secure data entry
- ✓ **Data extraction and statistical analysis**



Hellenic PulmOnary Hypertension rEgistry (HOPE)
3. Definition of PH

Greek PAHTool Registry

European Heart Journal
doi:10.1093/eurheartj/ehv317

ESC/ERS GUIDELINES

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

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

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

Table 3 Haemodynamic definitions of pulmonary hypertension[a]

Definition	Characteristics[a]	Clinical group(s)[b]
PH	PAPm ≥25 mmHg	All
Pre-capillary PH	PAPm ≥25 mmHg PAWP ≤15 mmHg	1. Pulmonary arterial hypertension 3. PH due to lung diseases 4. Chronic thromboembolic PH 5. PH with unclear and/or multifactorial mechanisms
Post-capillary PH	PAPm ≥25 mmHg PAWP >15 mmHg	2. PH due to left heart disease 5. PH with unclear and/or multifactorial mechanisms
Isolated post-capillary PH (Ipc-PH)	DPG <7 mmHg and/or PVR ≤3 WU[c]	
Combined post-capillary and pre-capillary PH (Cpc-PH)	DPG ≥7 mmHg and/or PVR >3 WU[c]	

CO = cardiac output; DPG = diastolic pressure gradient (diastolic PAP − mean PAWP); mPAP = mean pulmonary arterial pressure; PAWP = pulmonary arterial wedge pressure; PH = pulmonary hypertension; PVR = pulmonary vascular resistance; WU = Wood units.

[a]All values measured at rest; see also section 8.0.

[b]According to *Table 4*.

[c]Wood Units are preferred to dynes.s.cm^{-5}.

Table 4 Comprehensive clinical classification of pulmonary hypertension (updated from Simonneau et al.[5])

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 disease (Table 6)
  - 1.4.5 Schistosomiasis

1'. Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis
- 1'.1 Idiopathic
- 1'.2 Heritable
  - 1'.2.1 EIF2AK4 mutation
  - 1'.2.2 Other mutations
- 1'.3 Drugs, toxins and radiation induced
- 1'.4 Associated with:
  - 1'.4.1 Connective tissue disease
  - 1'.4.2 HIV infection

1''. Persistent pulmonary hypertension of the newborn

2. Pulmonary hypertension due to left heart disease
- 2.1 Left ventricular systolic dysfunction
- 2.2 Left ventricular diastolic dysfunction
- 2.3 Valvular disease
- 2.4 Congenital / acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies
- 2.5 Congenital /acquired pulmonary veins stenosis

3. Pulmonary hypertension due to lung diseases and/or hypoxia
- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental lung diseases (Web Table III)

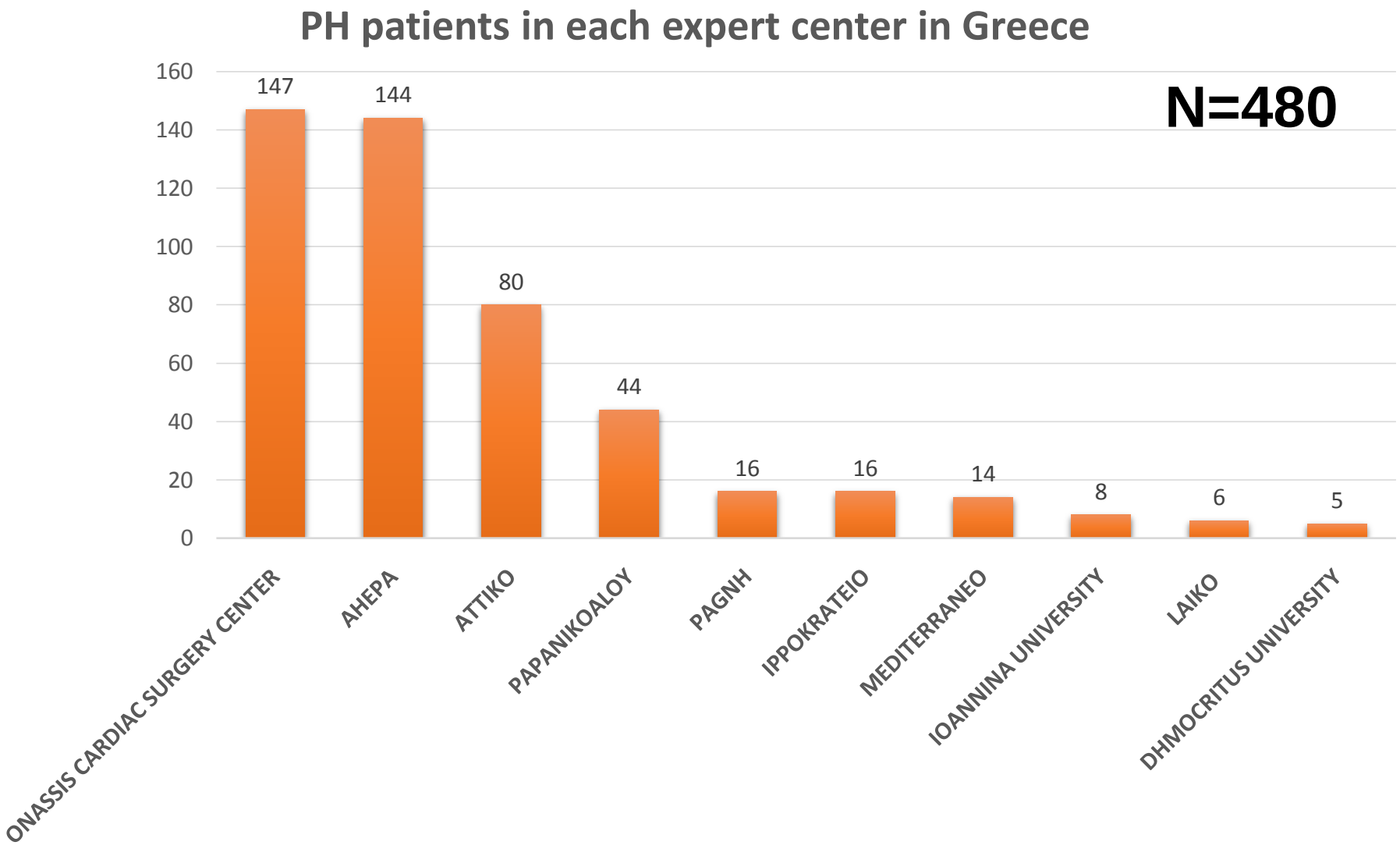
4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions
- 4.1 Chronic thromboembolic pulmonary hypertension
- 4.2 Other pulmonary artery obstructions
  - 4.2.1 Angiosarcoma
  - 4.2.2 Other intravascular tumors
  - 4.2.3 Arteritis
  - 4.2.4 Congenital pulmonary arteries stenoses
  - 4.2.5 Parasites (hydatidosis)

5. Pulmonary hypertension with unclear and/or multifactorial mechanisms
- 5.1 Haematological disorders: chronic haemolytic anaemia, myeloproliferative disorders, splenectomy
- 5.2 Systemic disorders, sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: pulmonary tumoral thrombothic microangiopathy, fibrosing mediastinitis, chronic renal failure (with/without dialysis), segmental pulmonary hypertension

BMPR2 = bone morphogenetic protein receptor, type 2; EIF2AK4 = eukaryotic translation initiation factor 2 alpha kinase 4; HIV = human immunodeficiency virus.

Hellenic PulmOnary Hypertension rEgistry (HOPE)
4.Patient Recruitment

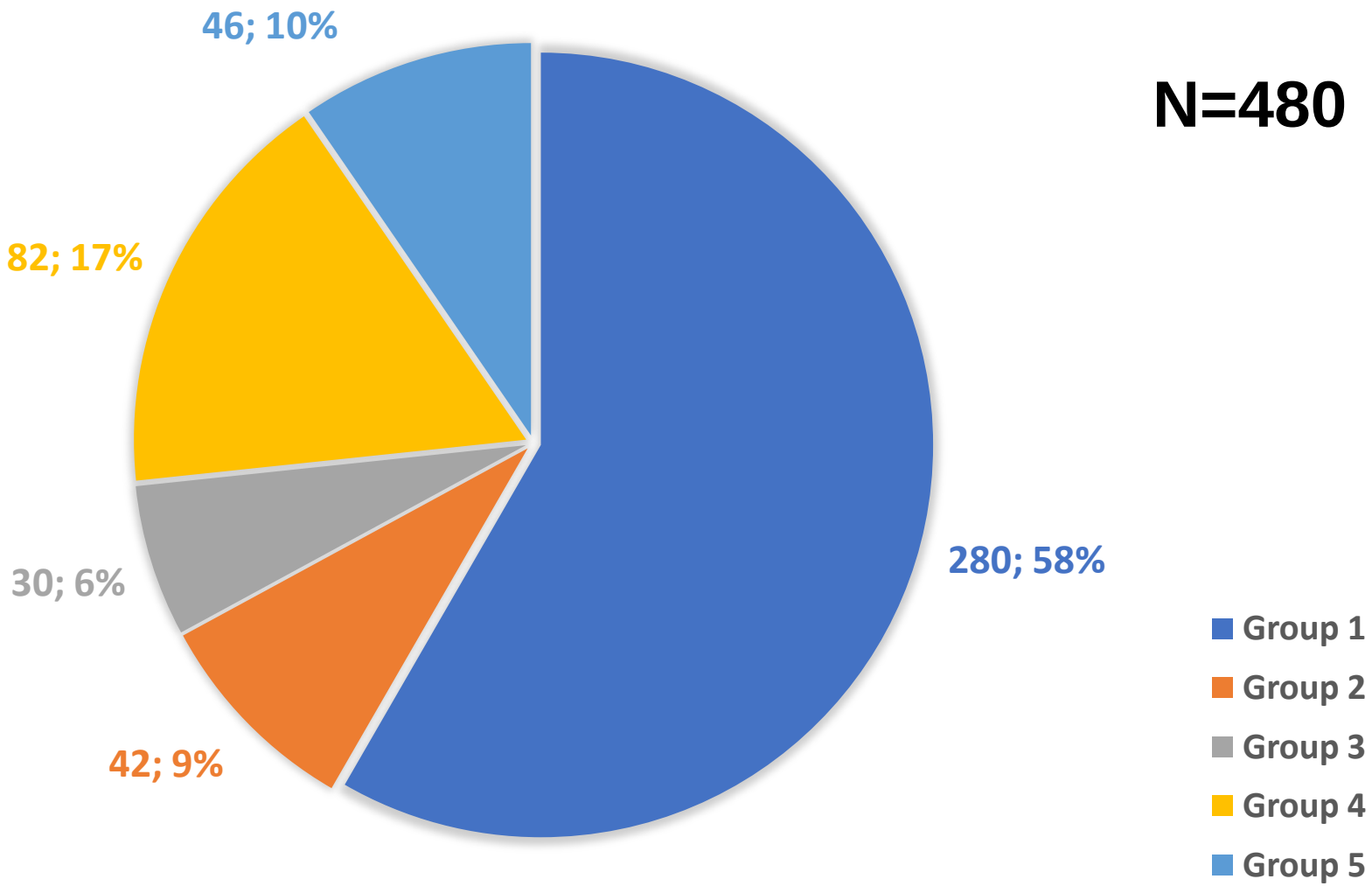
Greek PAHTool Registry



Hellenic PulmOnary Hypertension rEgistry (HOPE)
5.Classification of PH

Greek PAHTool Registry

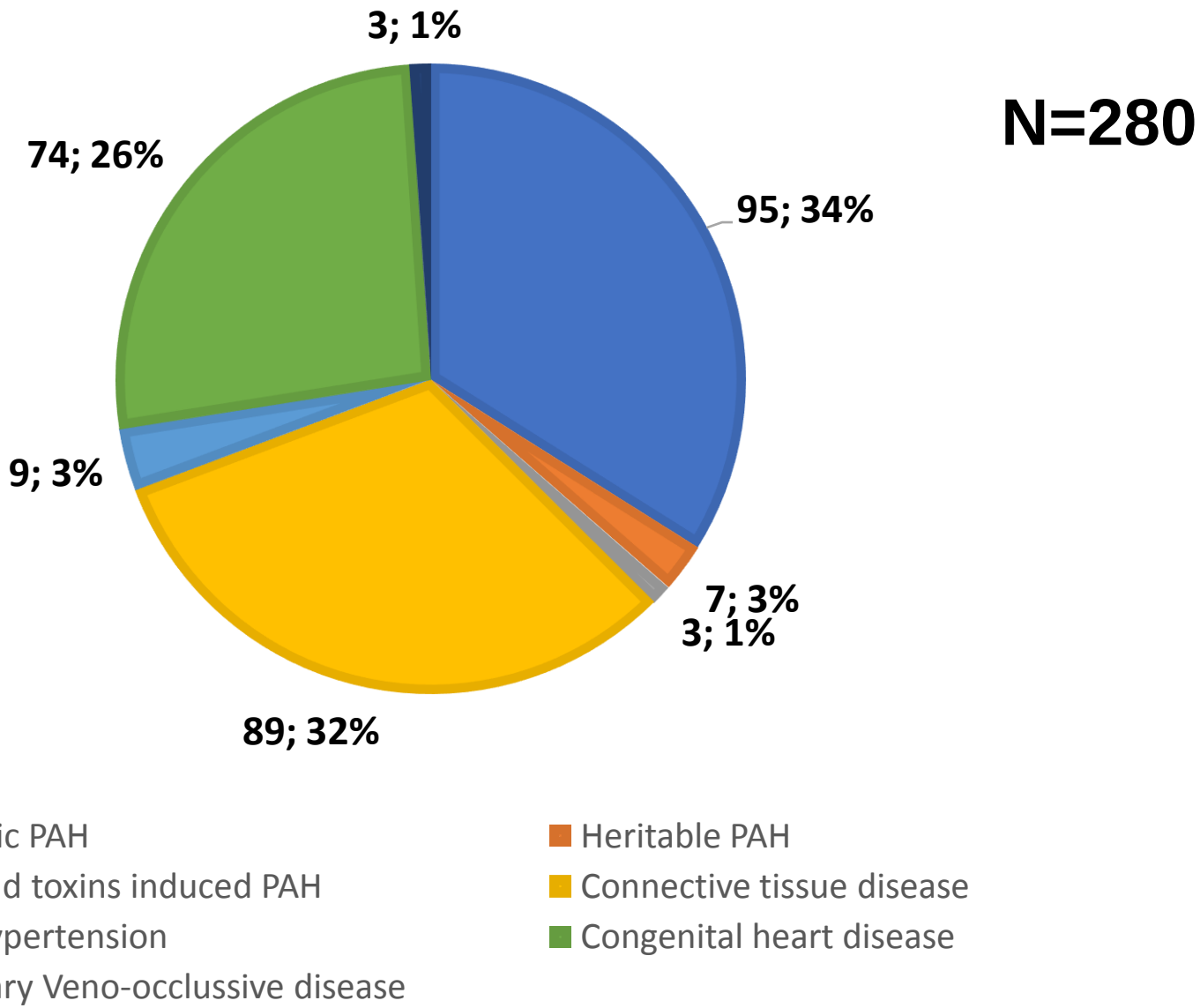
CLASSIFICATION OF PATIENTS WITH PULMONARY HYPERTENSION IN GREECE



Hellenic PulmOnary Hypertension rEgistry (HOPE)
6. Classification of PAH

Greek PAHTool Registry

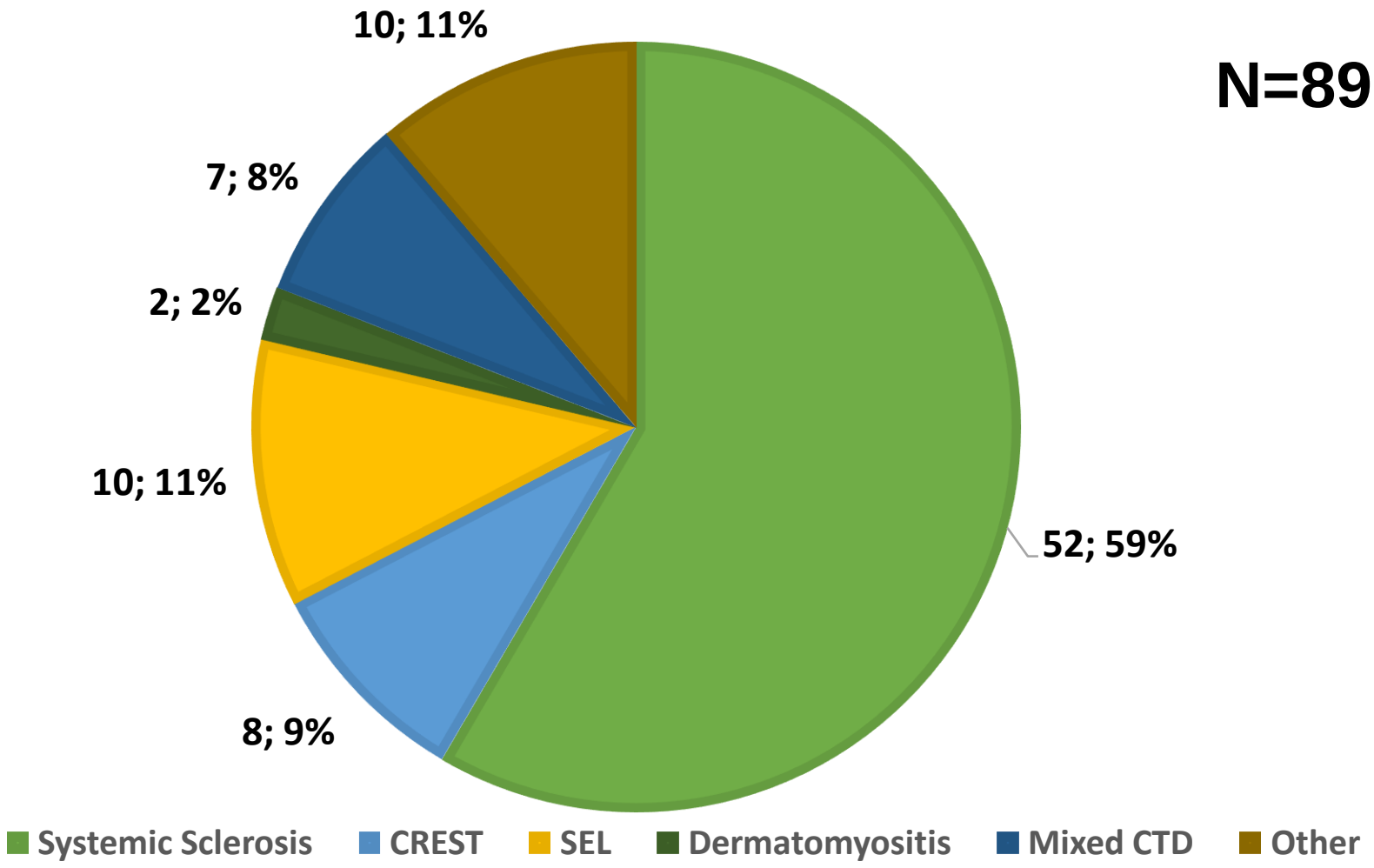
CLASSIFICATION OF PATIENTS WITH PAH IN GREECE



Hellenic PulmOnary Hypertension rEgistry (HOPE)
7.Classification of PAH-CTD

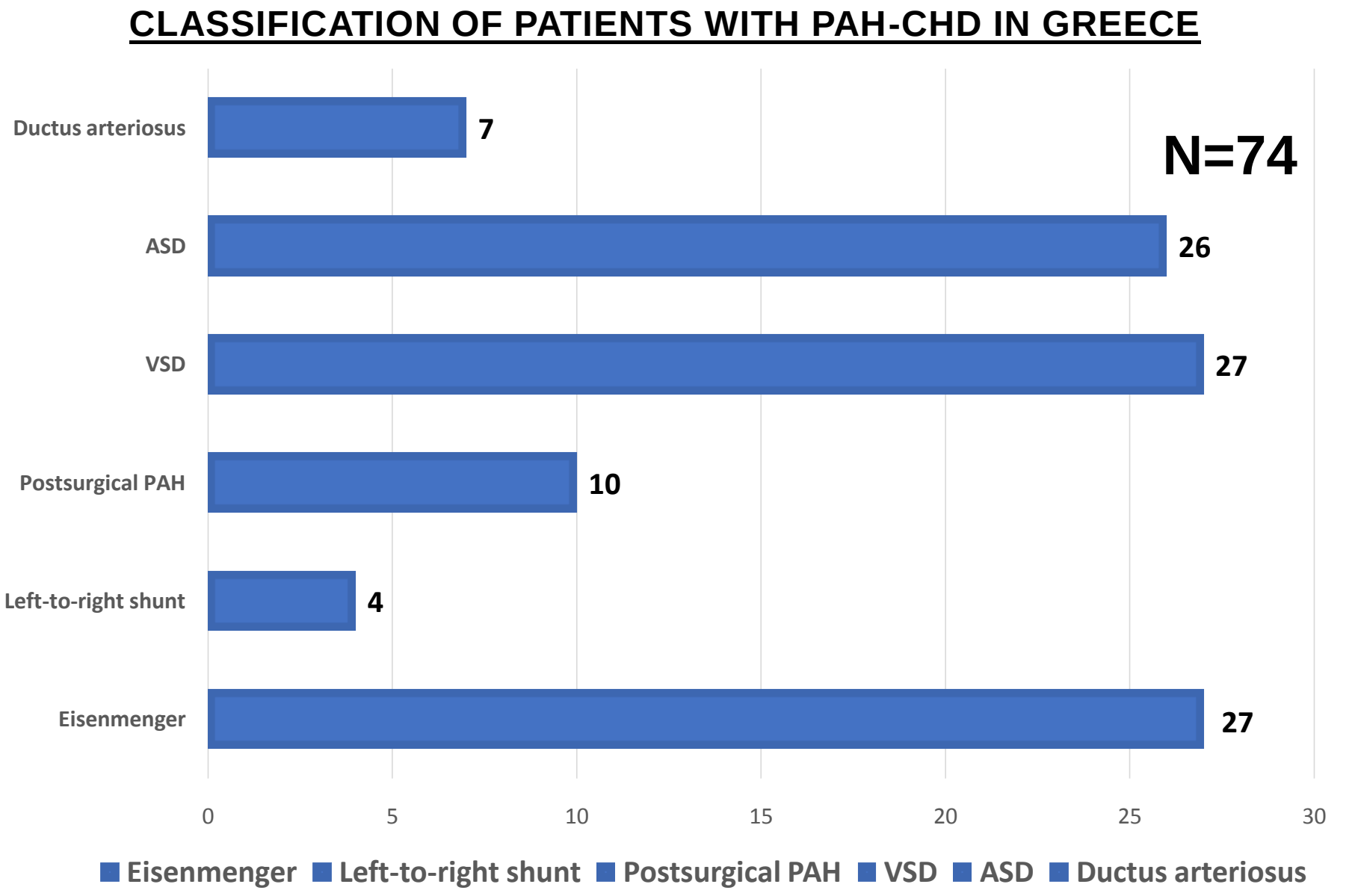
Greek PAHTool Registry

CLASSIFICATION OF PATIENTS WITH PAH-CTD IN GREECE



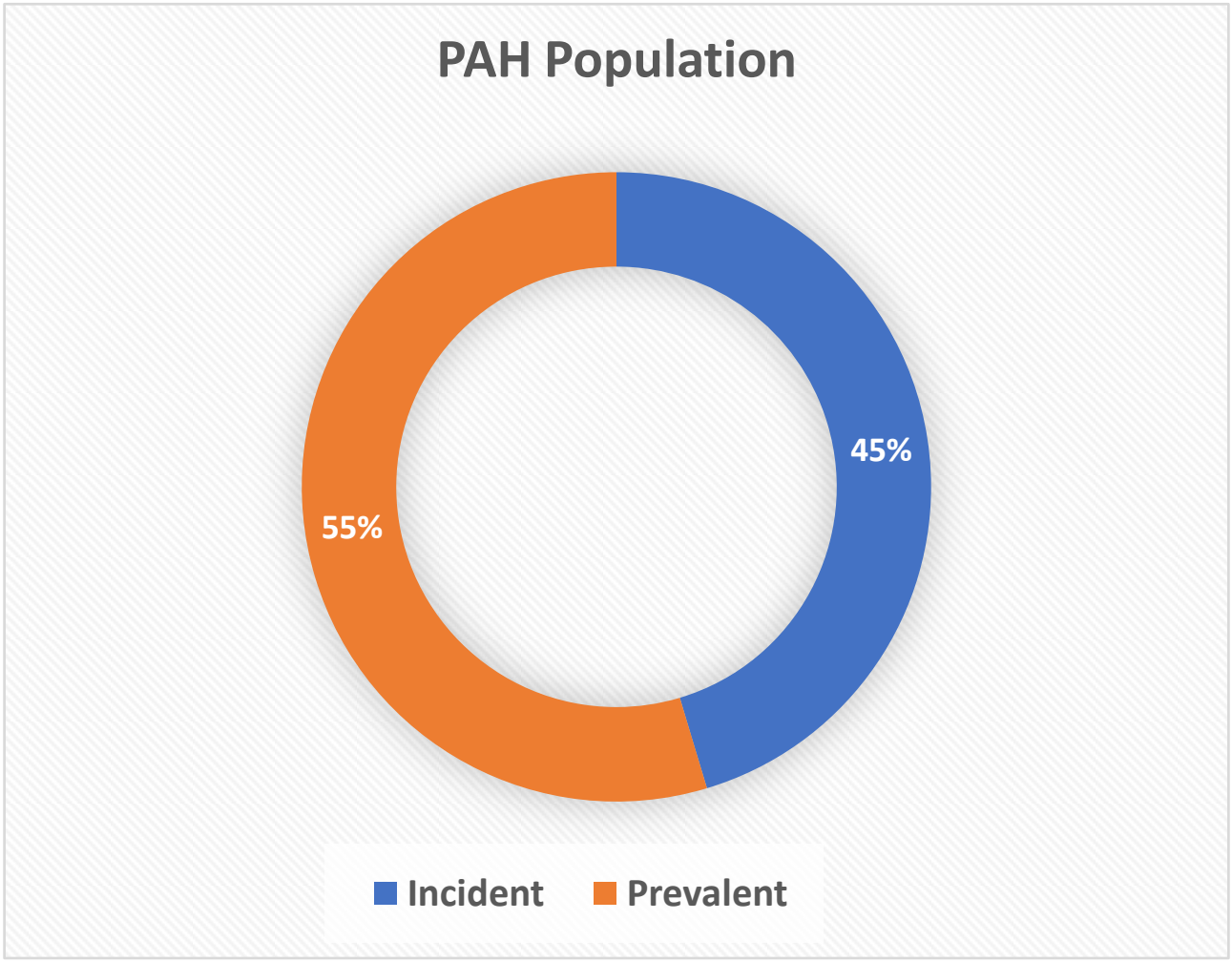
Hellenic PulmOnary Hypertension rEgistry (HOPE)
8.Classification of PAH-CHD

Greek PAHTool Registry



Hellenic PulmOnary Hypertension rEgistry (HOPE)
9. Incidence in PAH

Greek PAHTool Registry



Hellenic PulmOnary Hypertension rEgistry (HOPE)
10. Baseline PAH Characteristics

Greek PAHTool Registry

	PAH Total	IPAH/ HPAH/Drugs	PAH-CTD	PAH-CHD	Portal hypertension	P-Value
Subjects	280	105 (38)	89 (32)	74 (26)	9 (3)	
Female	186 (66.4)	67 (63.8)	71 (79.7)	44 (59.5)	3 (33.3)	**0.004**
Age, years	56.3 (25)	55.1 (23)	63.1(18)	39.6 (37)	56.2 (17)	**<0.0001**
BMI, (kg/m²)	25.8 (7.7)	27.9 (7.3)	27.4 (7.8)	25.3 (5.7)	29.4 (7.8)	**<0.0001**
Comorbidities						
Atrial Fibrillation	20 (7.1)	6 (5.7)	3 (3.4)	7 (9.4)	4 (44.4)	**0.000**
Arterial Hypertension	73 (26.1)	39 (37.1)	25 (56.2)	8 (8.9)	1 (1.1)	**0.001**
Dyslipidemia	36 (12.9)	15(14.2)	18 (20.2)	3 (4.5)	0	**0.013**
Diabetes	37 (13.2)	23 (22)	7 (7.8)	3 (4.5)	4 (44.4)	**0.000**
CAD	22 (7.9)	12(11.4)	9 (10.1)	0	0	**0.02**
Obesity	74 (26.4)	34 (32.4)	25 (28.1)	11 (14.8)	4 (44.4)	0.11
Hypothyroidism	48 (17.1)	13 (12.3)	24 (26.9)	11 (14.8)	0	0.08
Smoking	64 (22.9)	33 (31.4)	18 (20.2)	6 (8.1)	5 (55.5)	**0.002**
Symptoms						
Dyspnea	231 (82.5)	86 (81.9)	81 (91.0)	53 (71.6)	8 (88.8)	**0.013**
Fatigue	154 (55.0)	55 (52.4)	51 (57.3)	40 (54.0)	7 (7.8)	0.495
Palpitations	22 (7.9)	12 (11.4)	6 (67.4)	4 (5.4)	0	0.565
Chest pain	22 (7.9)	12 (11.4)	6 (67.4)	4 (5.4)	0	0.565
Syncope	21 (7.5)	11 (10.5)	5 (5.6)	4 (5.4)	1	0.493
PHE	64 (22.9)	21 (20.5)	18 (20.2)	12 (17.5)	1	0.20

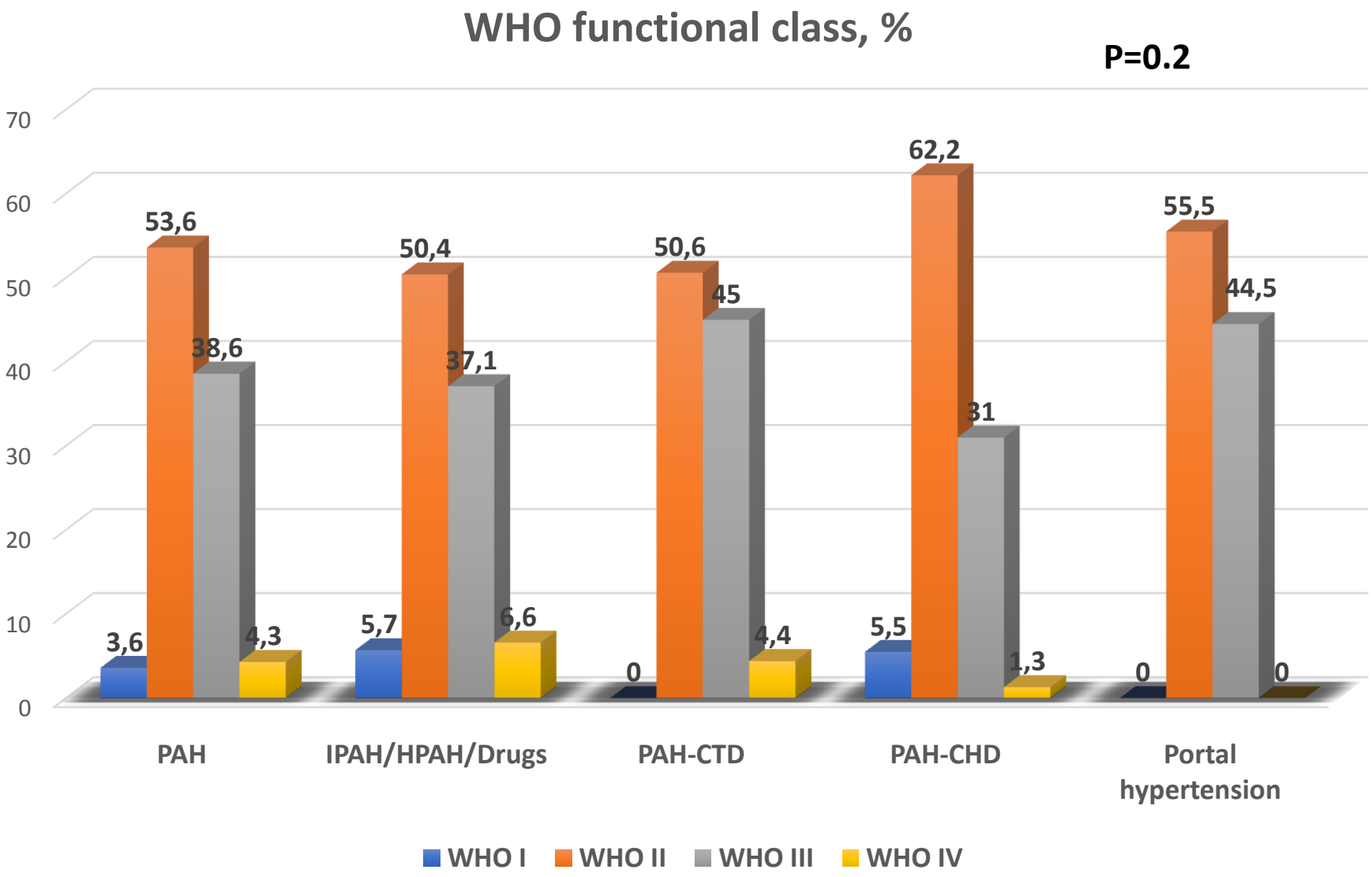
Hellenic PulmOnary Hypertension rEgistry (HOPE)
11. Baseline PAH Diagnostics

Greek PAHTool Registry

	PAH Total	IPAH/ HPAH/Drugs	PAH-CTD	PAH-CHD	Portal PH	P-Value
6-MWD, m N=202	382 (176)	410 (170)	325 (1963)	405 (168)	400 (96)	**0.013**
NT-pro BNP, pg/mL N=128	466 (1467)	483 (1046)	624 (2011)	452 (854)	400 (1386)	0.701
Echocardiography						
RA area (cm2)	22 (6)	22 (6)	21 (7.1)	25 (4)		0.8
RVEDD, mm	38 (16)	39.5 (19.3)	32 (13)	42 (13)	45 ± 13	0.08
TAPSE, mm	19 (6)	18 (6.3)	20 (4.5)	17 (9)	21.6 ± 1.5	0.09
TR Vmax, m/s	4 (1.1)	4 (0.8)	3.6 (0.9)	4.5 (1.4)	4 (1.1)	**0.008**
RVSP, mmHg	70 (33.2)	74.5 (30.5)	60.1 (24.2)	89 (56.5)	63.5 (30.2)	**0.001**
Right heart catheterization, n=261						
m RAP, mmHg	8 (6)	9 (6)	6 (4.3)	9 (5.8)	9 (7)	**0.003**
m PAP, mmHg	45 (20.5)	50 (18.0)	39 (15.0)	51 (36.0)	37 (24)	**<0.0001**
PAWP, mmHg	11 (5.3)	11 (6.0)	11 (4.8)	12 (4.0)	10 (10)	0.437
CO, L/min	4.2 (1.97)	4.2 (1.6)	4.2 (1.8)	4 (2.4)	5.8 (4.5)	**0.044**
CI, L/min/m²	2.45 (0.92)	2.3 (0.8)	2.45 (0.8)	2.63 (1.2)	2.9 (0.9)	**0.007**
PVR, WU	10.4 (7.0)	16.3 (10.7)	10.5 (8.8)	14.5 (13.2)	8.8 (7.8)	**<0.0001**
HR, bpm	78 (15.5)	77 (13)	80 (16.5)	85 (22)	75 (17)	**0.001**
SvO2, %	70 (11.1)	68 (8.6)	69.7 (9.7)	75 (11.5)	72.9	**<0.0001**

Hellenic PulmOnary Hypertension rEgistry (HOPE)
12. Functional Class at baseline

Greek PAHTool Registry



Hellenic PulmOnary Hypertension rEgistry (HOPE)
13. 1-year mortality risk stratification at baseline

Greek PAHTool Registry

Mortality in pulmonary arterial hypertension: prediction by the 2015 European pulmonary hypertension guidelines risk stratification model

Marius M. Hoeper[1,2], Tilmann Kramer[3,4], Zixuan Pan[5], Christina A. Eichstaedt[5], Jens Spiesshoefer[6], Nicola Benjamin[5], Karen M. Olsson[1,2], Katrin Meyer[1], Carmine Dario Vizza[7], Anton Vonk-Noordegraaf[8], Oliver Distler[9], Christian Opitz[10], J. Simon R. Gibbs[11], Marion Delcroix[12], H. Ardeschir Ghofrani[13], Doerte Huscher[14], David Pittrow[15], Stephan Rosenkranz[3,4] and Ekkehard Grünig[2,5]

TABLE 1 Variables and cut-off values used for risk stratification

	Low risk	Intermediate risk	High risk
WHO FC	I/II	III	IV
6-min walking distance m	>440	165–440	<165
BNP ng·L^{-1}	<50	50–300	>300
NT-proBNP ng·L^{-1}	<300	300–1400	>1400
Right atrial pressure mmHg	<8	8–14	>14
Cardiac index L·min^{-1}·m^{-2}	⩾2.5	2.0–2.4	<2.0
Sv_{O_2} %	>65	60–65	<60

WHO FC: World Health Organization functional class; BNP: brain natriuretic peptide; NT-proBNP: N-terminal fragment of pro-brain natriuretic peptide; Sv_{O_2}: mixed venous oxygen saturation.

Hellenic PulmOnary Hypertension rEgistry (HOPE)
14. Follow up Characteristics

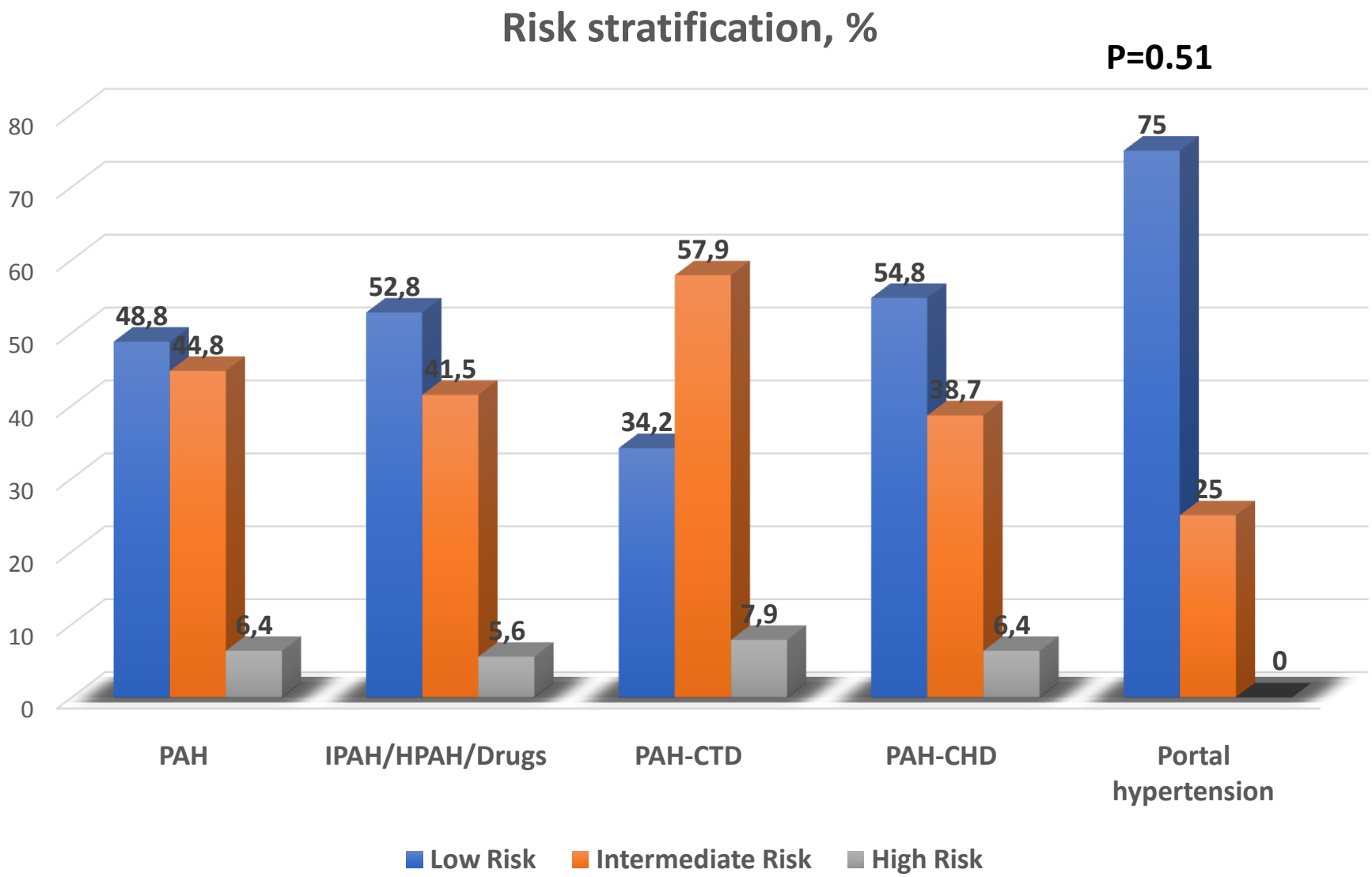
Greek PAHTool Registry

Differences in functional and hemodynamic parameters at follow up in PAH.

	Baseline n=126	Follow up n=126	
Age, years	53.7 ± 15.9	56.7 ± 13.7	**<0.0001**
NYHA I/II	74 (58.7)	77 (61.1)	**<0.0001**
NYHA III/IV	52 (41.2)	49 (38.8)	
Risk Stratification			
Low Risk	48 (38)	61 (48.4)	**0.008**
Intermediate Risk	69 (54.7)	57 (45.2)	
High Risk	9 (7.1)	8 (6.3)	
6MWD, m n=55	368.5 ± 173.9	418.7± 163.8	**0.001**
NT-proBNP, n=60	1373 ± 1933	962 ± 1294	0.09
Echocardiography, n=38			
RA area (cm2)	21.3 ± 3.9	23.1 ± 3.9	0.14
TAPSE, mm	18.5 ± 5.3	18.1 ± 4.6	0.6
TR Vmax, m/s	3.9 ± 0.6	3.8 ± 0.6	0.11
RVSP, mmHg	75.7 ± 24.3	69.3 ± 23.4	0.05
Right heart catheterization, n=50			
m RAP, mmHg	9.3 ± 4.5	7.8 ± 3.5	**0.04**
m PAP, mmHg	48.7 ± 12.8	42.6 ± 12.9	**<0.0001**
PAWP, mmHg	11 (5.3)	11 (6.0)	0.12
CO, L/min	4.6 ± 1.4	5.6 ± 1.7	**<0.0001**
CI, L/min/m^2	2.5 ± 0.7	3.1 ± 0.9	**<0.0001**
PVR, WU	16.3 ± 7.8	10.9 ± 5.5	**<0.0001**
SVO2, %	69.2 ± 8	71.3 ± 7.5	0.05

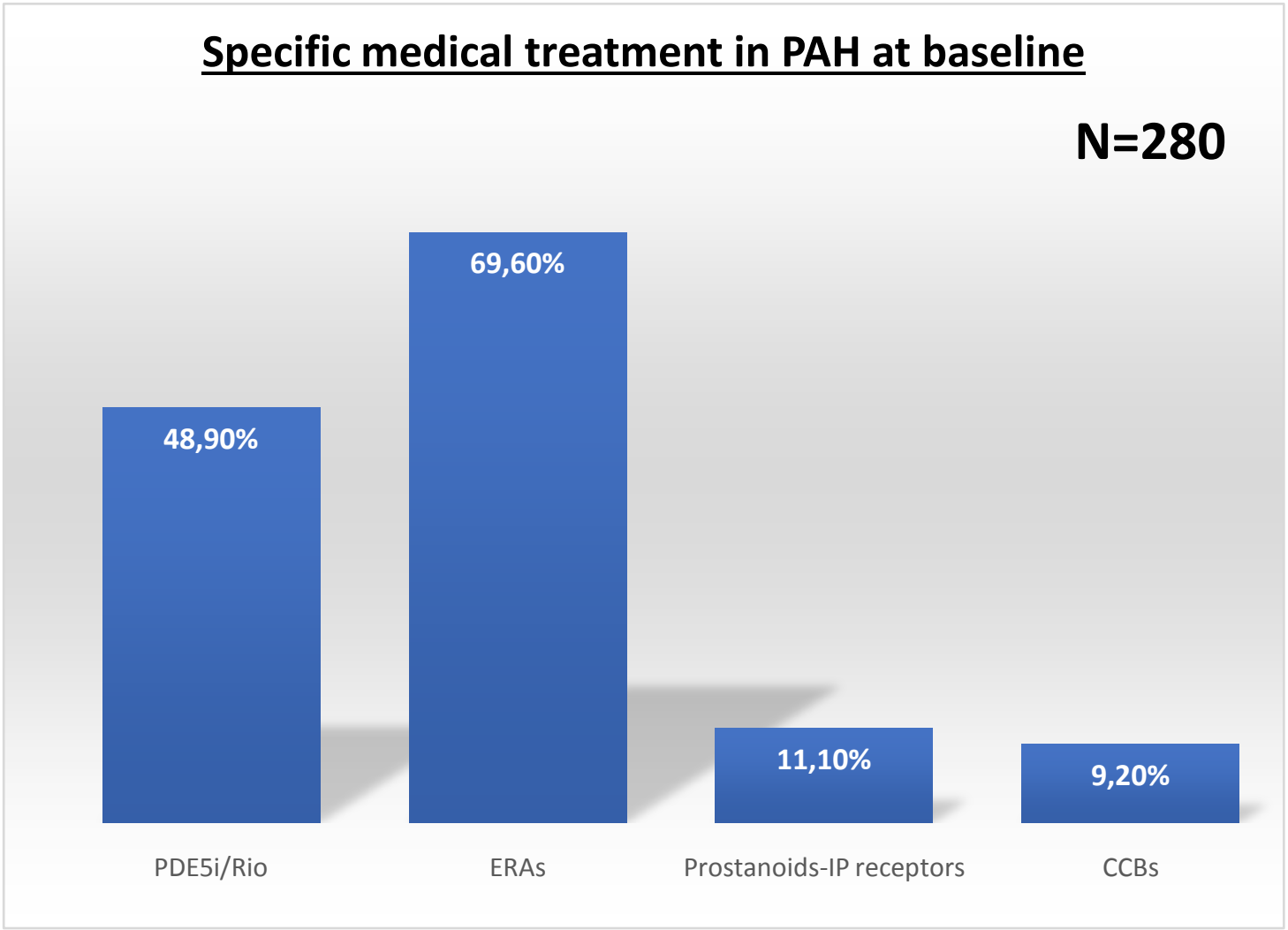
Hellenic PulmOnary Hypertension rEgistry (HOPE)
15. Follow up Risk Stratification

Greek PAHTool Registry



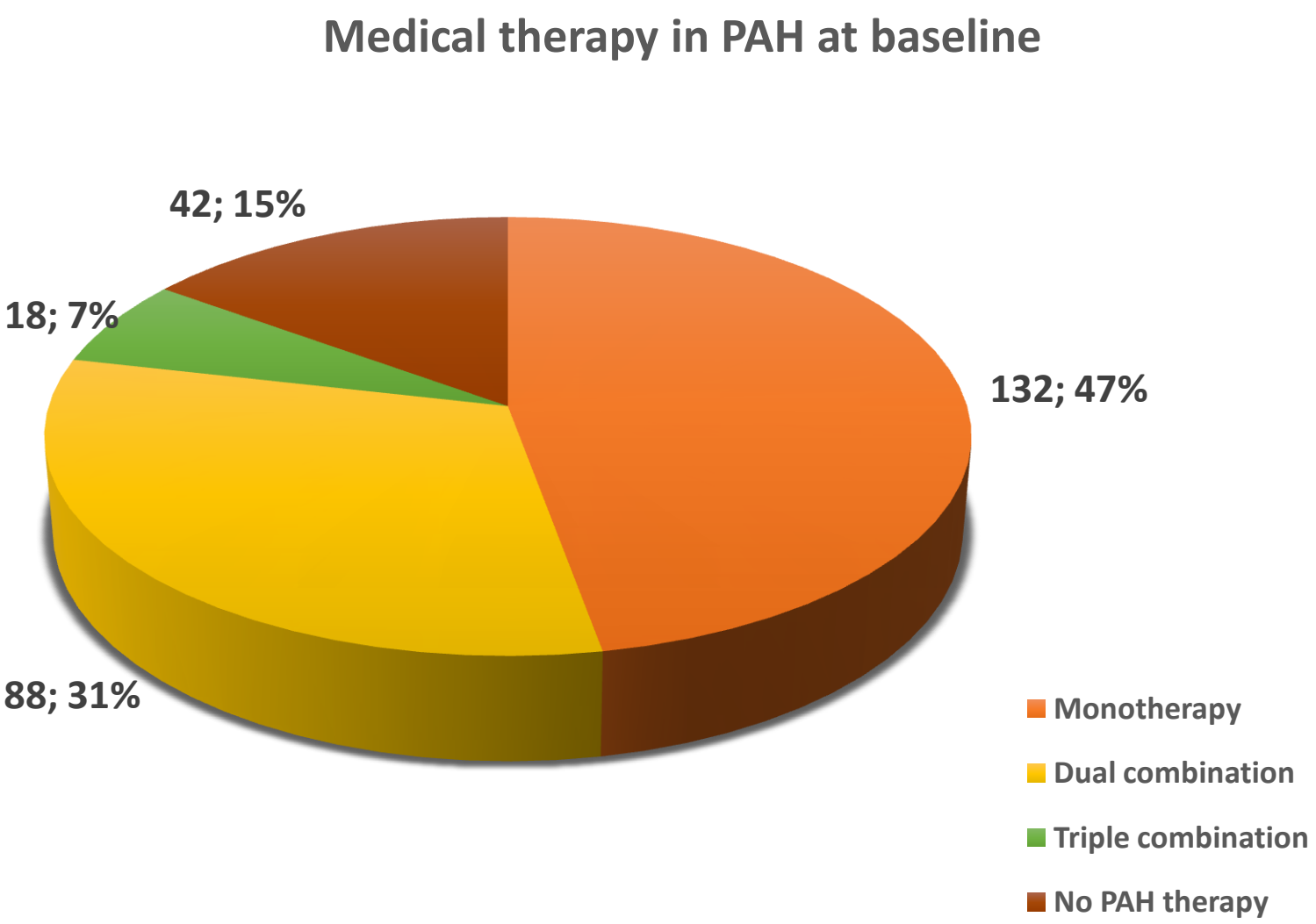
Hellenic PulmOnary Hypertension rEgistry (HOPE)
16. Pharmacotherapy at baseline

Greek PAHTool Registry



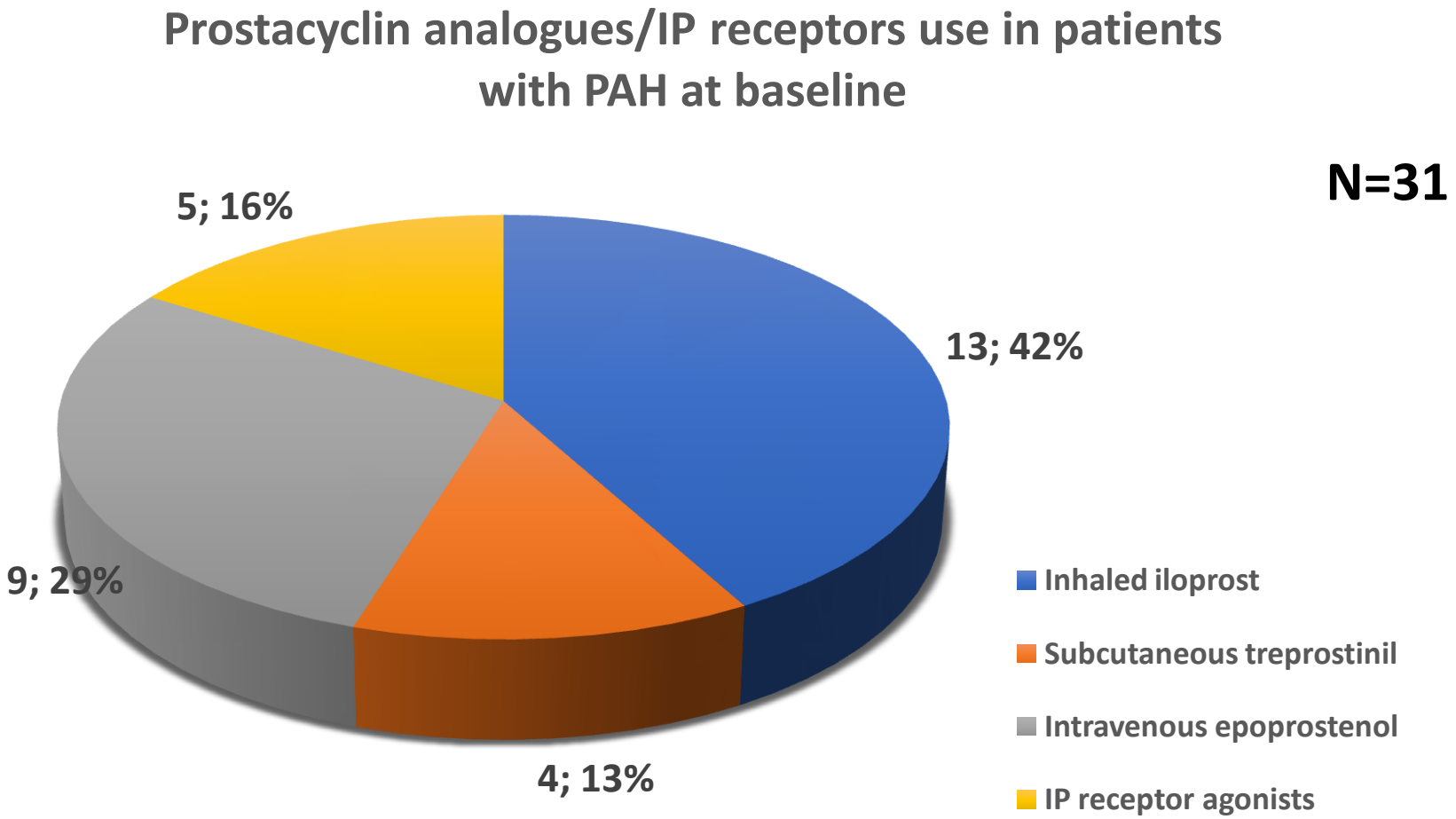
Hellenic PulmOnary Hypertension rEgistry (HOPE)
16. Pharmacotherapy at baseline

Greek PAHTool Registry



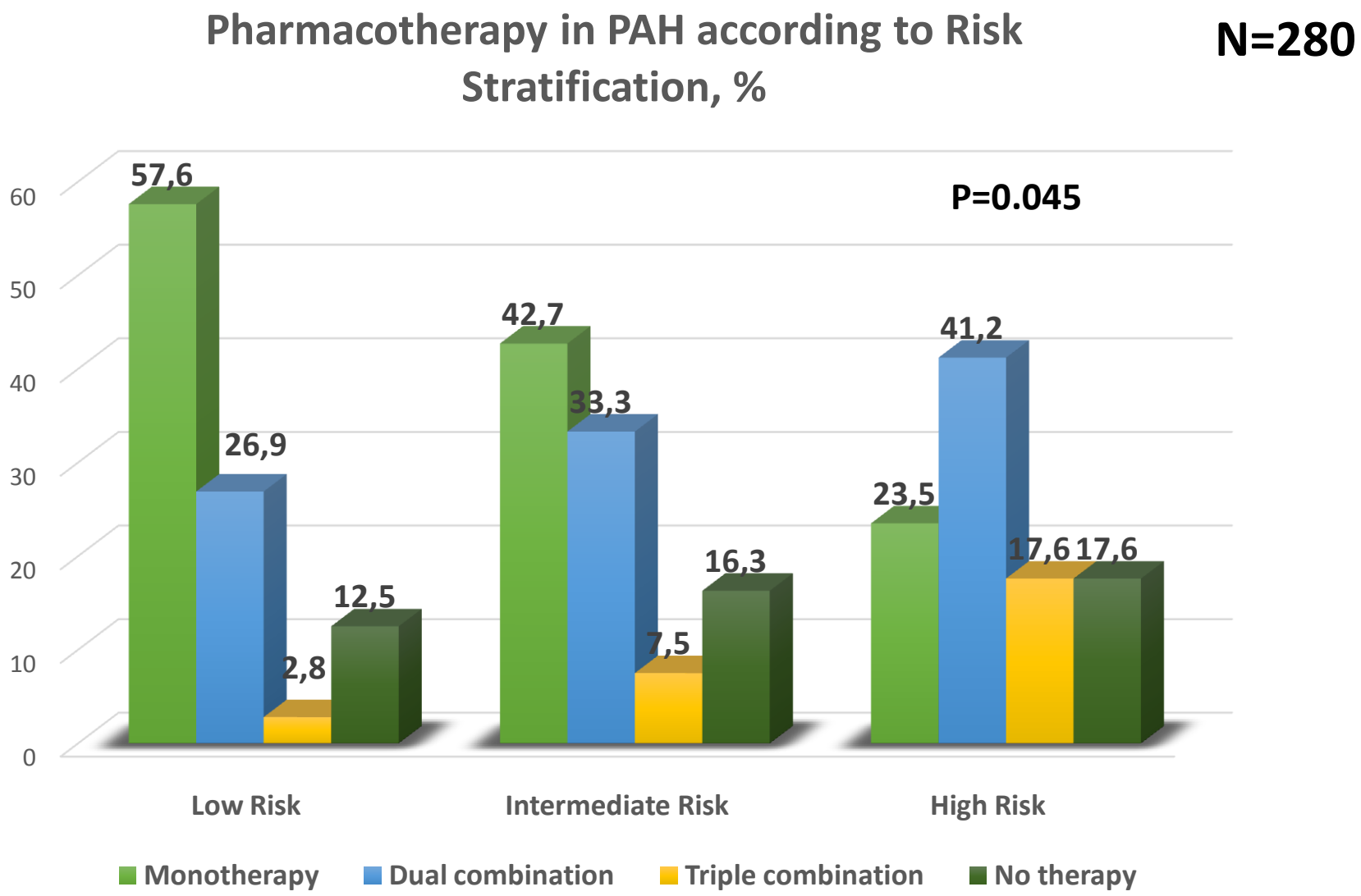
Hellenic PulmOnary Hypertension rEgistry (HOPE)
16. Pharmacotherapy at baseline

Greek PAHTool Registry



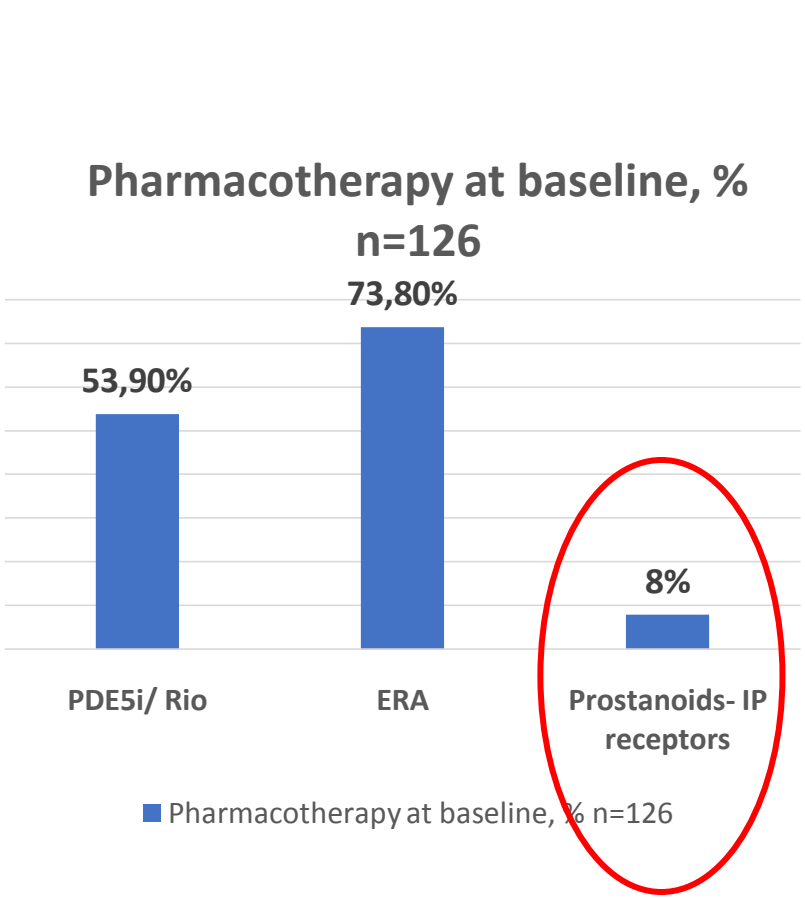
Hellenic PulmOnary Hypertension rEgistry (HOPE)
16. Pharmacotherapy at baseline

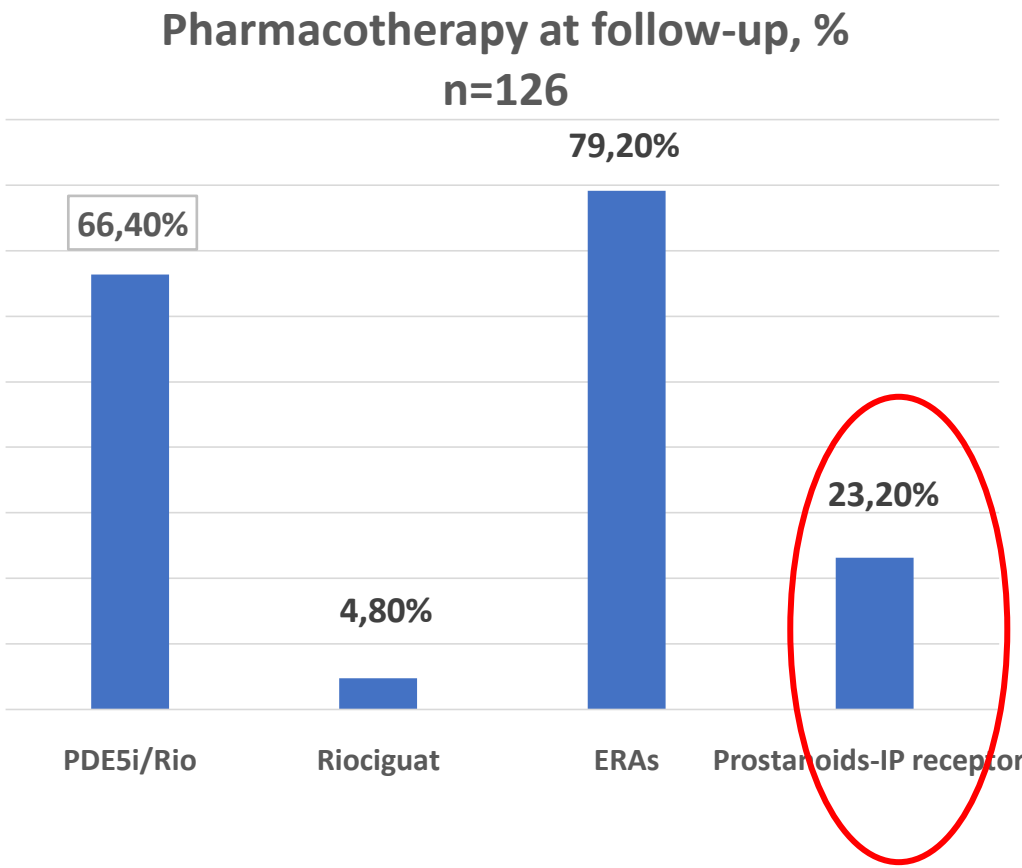
Greek PAHTool Registry



Hellenic PulmOnary Hypertension rEgistry (HOPE)
17. Pharmacotherapy at Follow up

Greek PAHTool Registry





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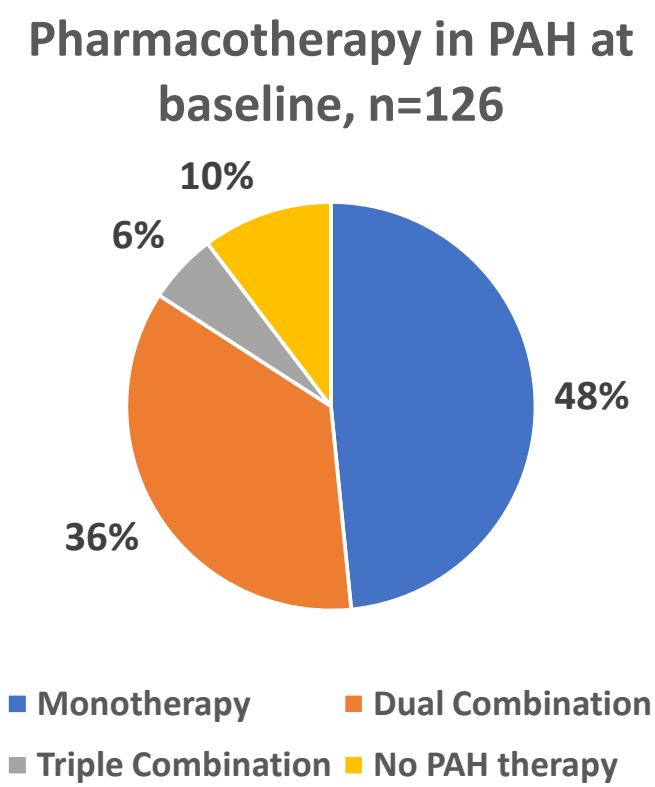
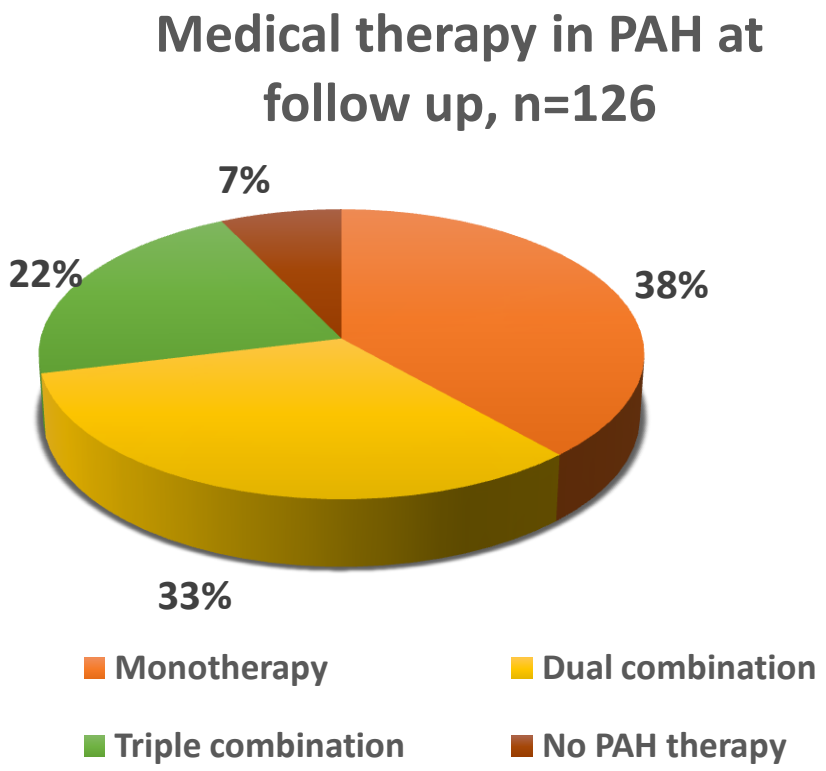
Hellenic PulmOnary Hypertension rEgistry (HOPE)
17. Pharmacotherapy at Follow up

Greek PAHTool Registry





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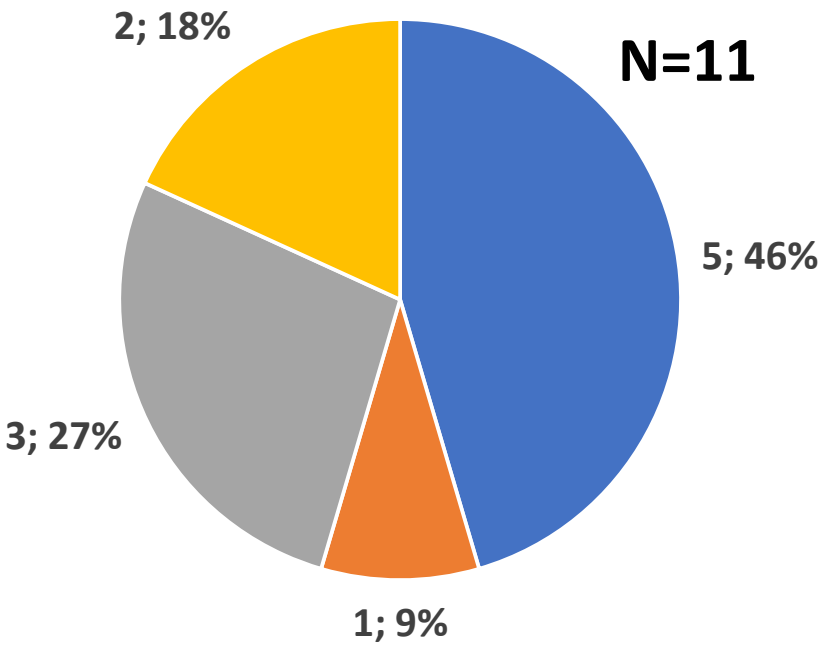
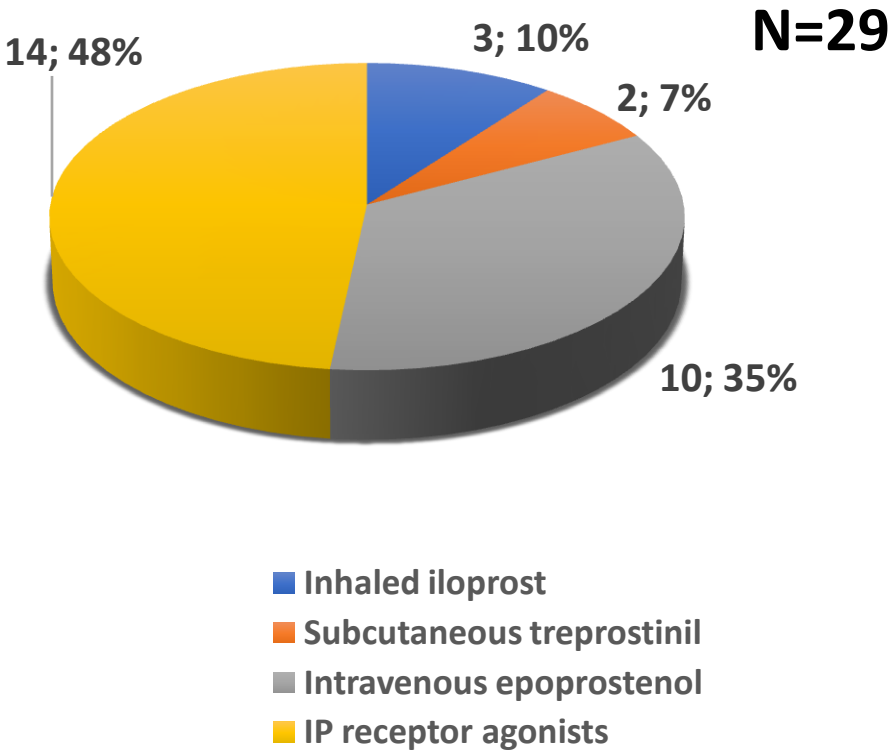
Hellenic PulmOnary Hypertension rEgistry (HOPE)
17. Pharmacotherapy at Follow up

Greek PAHTool Registry

Prostacyclin analogues/IP receptor agonists at baseline, n=126



Prostacyclin analogues/IP receptor agonists in patients with PAH at follow up, n=126



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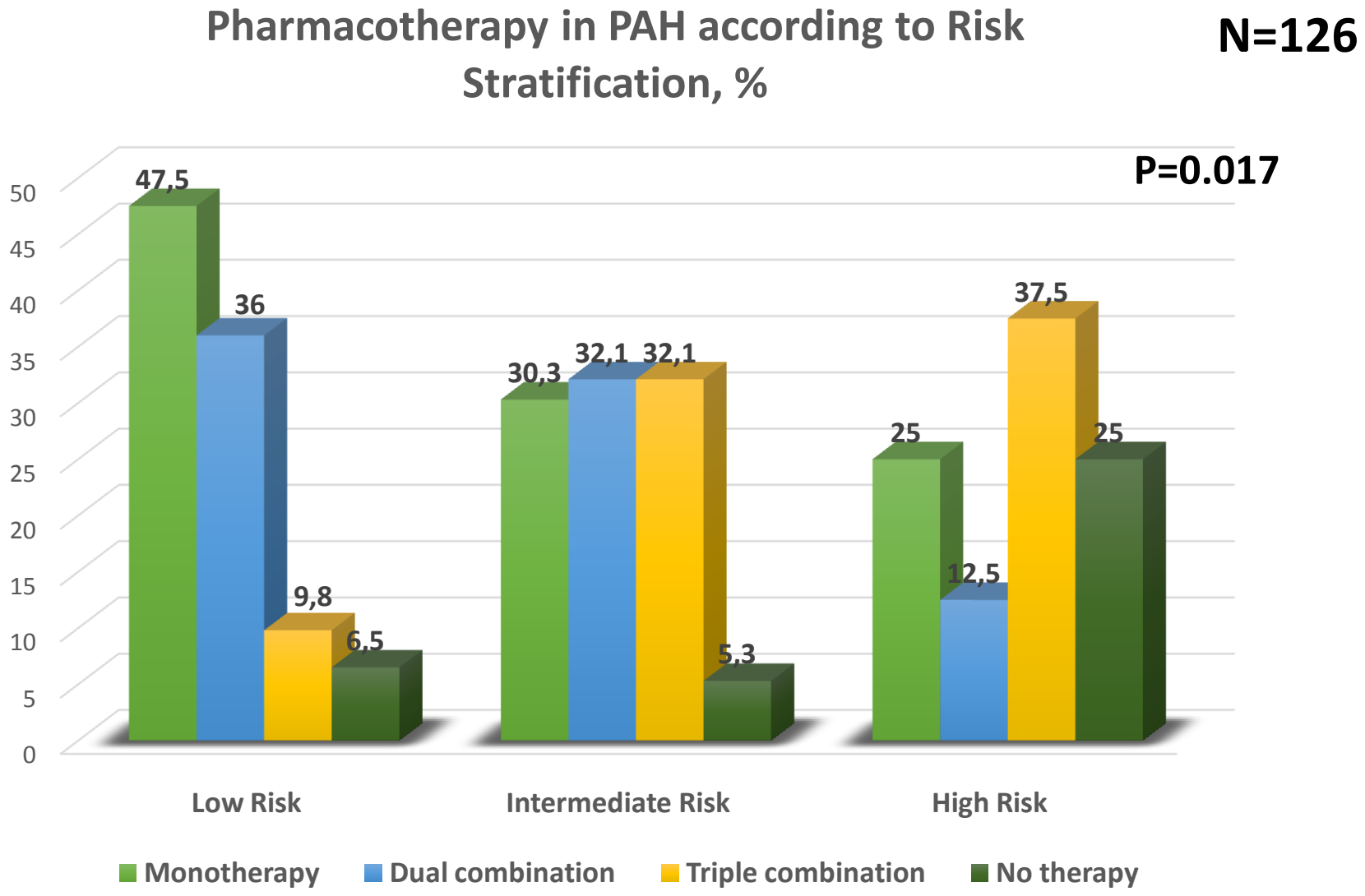
Hellenic PulmOnary Hypertension rEgistry (HOPE)
17. Pharmacotherapy at Follow up

Greek PAHTool Registry



Eur Respir Rev 2012; 21: 123, 8–18
DOI: 10.1183/09059180.00008211

EUROPEAN RESPIRATORY UPDATE

REVEAL: a contemporary US pulmonary arterial hypertension registry

M.D. McGoon* and D.P. Miller#

TABLE 2 Pulmonary arterial hypertension-specific medications among patients meeting traditional haemodynamic criteria in the REVEAL registry at enrolment

	ERA#	PDE-5 inhibitor¶	Prostacyclin analogue		
			Intravenous epoprostenol	Inhaled iloprost	Treprostinil+
Overall use§	1147 (47.0)	1194 (49.0)	480 (19.7)	237 (9.7)	307 (12.6)
Monotherapy	452 (18.5)	417 (17.1)	188 (7.7)	23 (0.9)	84 (3.4)
Combination with one oral therapyƒ	291 (11.9)	290 (11.9)	243 (10.0)	138 (5.7)	148 (6.1)
Combination with one prostacyclin analogue	224 (9.2)	305 (12.5)	2 (0.1)	3 (0.1)	5 (0.2)
Combination with >1 other therapy	180 (7.4)	182 (7.5)	47 (1.9)	73 (3.0)	70 (2.9)
NYHA/WHO functional class I/II					
Overall use	468 (47.1)	474 (47.7)	187 (18.8)	71 (7.1)	111 (11.2)
Monotherapy	216 (21.7)	187 (18.8)	83 (8.4)	4 (0.4)	26 (2.6)
Combination with one oral therapyƒ	110 (11.1)	109 (11.0)	85 (8.6)	41 (4.1)	60 (6.0)
Combination with one prostacyclin analogue	76 (7.6)	110 (11.1)			
Combination with >1 other therapy	66 (6.6)	686 (6.8)	19 (1.9)	26 (2.6)	25 (2.5)
NYHA/WHO functional class III					
Overall use	525 (47.7)	567 (51.5)	218 (19.8)	130 (11.8)	161 (14.6)
Monotherapy	181 (16.4)	177 (16.1)	80 (7.3)	16 (1.5)	43 (3.9)
Combination with one oral therapyƒ	147 (13.4)	147 (13.4)	117 (10.6)	78 (7.1)	79 (7.2)
Combination with one prostacyclin analogue	114 (10.4)	160 (14.5)	2 (0.2)	2 (0.2)	4 (0.4)
Combination with >1 other therapy	83 (7.5)	83 (7.5)	19 (1.7)	34 (3.1)	35 (3.2)
NYHA/WHO functional class IV					
Overall use	55 (44.4)	61 (49.2)	44 (35.5)	14 (11.3)	15 (12.1)
Monotherapy	5 (4.0)	14 (11.3)	17 (13.7)	2 (16)	5 (4.0)
Combination with one oral therapyƒ	16 (12.9)	16 (12.9)	21 (16.9)	8 (6.5)	4 (3.2)
Combination with one prostacyclin analogue	18 (14.5)	15 (12.1)			
Combination with >1 other therapy	16 (12.9)	16 (12.9)	6 (4.8)	4 (3.2)	6 (4.8)

Data are presented as n (%). Combinations with one oral therapy, with one prostacyclin analogue, and with more than one oral therapy are mutually exclusive and exclude calcium channel blockers. Blinded clinical trial patients are excluded from this presentation (n=87). ERA: endothelin receptor antagonist; PDE-5: phosphodiesterase type-5; NYHA/WHO: New York Heart Association/World Health Organization. #: 953 on bosentan, 106 on sitaxsentan and 89 on ambrisentan; ¶: 1,147 on sildenafil and 47 on tadalafil; +: treprostinil use includes 159 on intravenous, 112 on subcutaneous, 28 on inhaled and nine on oral treprostinil; §: n=2,435; ƒ: oral therapy is defined as bosentan, sildenafil, ambrisentan, sitaxsentan and tadalafil. Reproduced from [15] with permission from the publisher.

42%

37%

58%

Hoeper MM, Kramer T, Pan Z, et al. Mortality in pulmonary arterial hypertension: prediction by the 2015 European pulmonary hypertension guidelines risk stratification model. Eur Respir J 2017; 50: 1700740

Compera Registry

TABLE 2 Characteristics of the patients included in the baseline risk stratification group

	N	Low risk	Intermediate risk	High risk	All
Subjects n		196	1116	276	1588
Age years		53±18	66±14	65±17	64±16
Female		66	63	66	64
BMI kg·m^{-2}		26±5	28±6	28±7	28±6
PAH aetiology					
I/D/H-PAH		101 (52)	760 (68)	199 (72)	1060 (67)
CTD-PAH		52 (27)	234 (21)	61 (22)	347 (22)
HIV-PAH		4 (2)	16 (1)	2 (1)	22 (1)
PoPH		20 (10)	60 (5)	9 (3)	89 (6)
CHD-PAH		19 (10)	46 (4)	5 (2)	70 (4)
WHO FC class I/II/III/IV	1530	2/47/43/0 (unknown n=8)	0/8/78/11 (unknown n=3)	0/1/53/43 (unknown n=3)	0/11/70/15 (unknown n=4)
6MWD m	1262	442±100	299±109	186±103	298±126
NT-proBNP ng·L^{-1}	1003	151 (86–351)	1404 (597–3042)	4006 (2519–6743)	1573 (526–3498)
BNP ng·L^{-1}	249	44 (25–96)	180 (93–377)	549 (418–784)	236 (101–523)
Haemodynamics					
Right atrial pressure mmHg	1506	5±3	8±4	13±5	8±5
mPAP mmHg	1588	43±13	44 ±12	50±12	45±13
PAWP mmHg	1588	8±3	9±4	10±3	9±3
Cardiac index L·min^{-1}·m^{-2}	1497	3.1±0.7	2.3±0.7	1.6±0.4	2.3±0.8
PVR dyn·s·cm^{-5}	1588	547±273	743±372	1149±522	784±431
Sv_{O_2} %	1420	72±5	64±8	53±8	63±9
Initial therapy (within 3 months after diagnosis)					
CCB		9%	4%	1%	4%
ERA		43%	36%	45%	39%
PDE-5i/sGC		63%	72%	78%	72%
PCA		1%	2%	7%	3%
Monotherapy		81%	86%	73%	83%
Combination therapy		19%	14%	27%	17%
Anticoagulation		31%	44%	48%	43%

Hoeper MM, Kramer T, Pan Z, et al. Mortality in pulmonary arterial hypertension: prediction by the 2015 European pulmonary hypertension guidelines risk stratification model. Eur Respir J 2017; 50: 1700740

Compera Registry

TABLE 3 Variables obtained between 3 months and 2 years after treatment initiation of patients included in the follow-up risk stratification group

	N	Low risk	Intermediate risk	High risk	All
Subjects n		261	650	183	1094
Age years		52±17	66±14	73±11	64±16
Female		68	65	62	65
BMI kg·m^{-2}		26±6	29±7	29±6	28±7
PAH aetiology					
I/D/H-PAH		158 (61)	460 (71)	138 (75)	756 (69)
CTD-PAH		55 (21)	122 (19)	36 (20)	213 (20)
HIV-PAH		6 (2)	9 (1)	1 (1)	16 (2)
PoPH		25 (10)	28 (4)	2 (1)	55 (5)
CHD-PAH		17 (7)	31 (5)	6 (3)	54 (5)
WHO FC class I/II/III/IV	964	11/72/17/0	1/20/76/4	0/0/81/19	3/28/64/6
6MWD m	713	472±100	324±99	140±83	349±133
NT-proBNP ng·L^{-1}	626	145 (78–279)	900 (366–1851)	3788 (2072–5465)	887 (263–2339)
BNP ng·L^{-1}	146	27 (20–76)	136 (88–250)	457 (361–685)	190 (100–371)
Haemodynamics					
Right atrial pressure mmHg	374	5±2	8±5	15±5	8±5
mPAP mmHg	384	40±13	44±12	50±9	43±12
PAWP mmHg	378	9±3	10±4	12±5	10±4
Cardiac index L·min^{-1}·m^{-2}	360	3.2±0.8	2.3±0.7	1.7±0.4	2.6±0.8
PVR dyn·s·cm^{-5}	376	449±211	681±315	967±334	632±326
Sv_{O_2} %	349	72±5	63±7	55±8	65±8
Initial therapy (within 3 months after diagnosis)					
CCB		9%	3%	1%	4%
ERA		59%	55%	48%	55%
PDE-5i/sGC		72%	75%	79%	75%
PCA		4%	6%	7%	5%
Monotherapy		51%	59%	61%	57%
Combination therapy		47%	40%	36%	41%
Anticoagulation		36%	51%	62%	49%

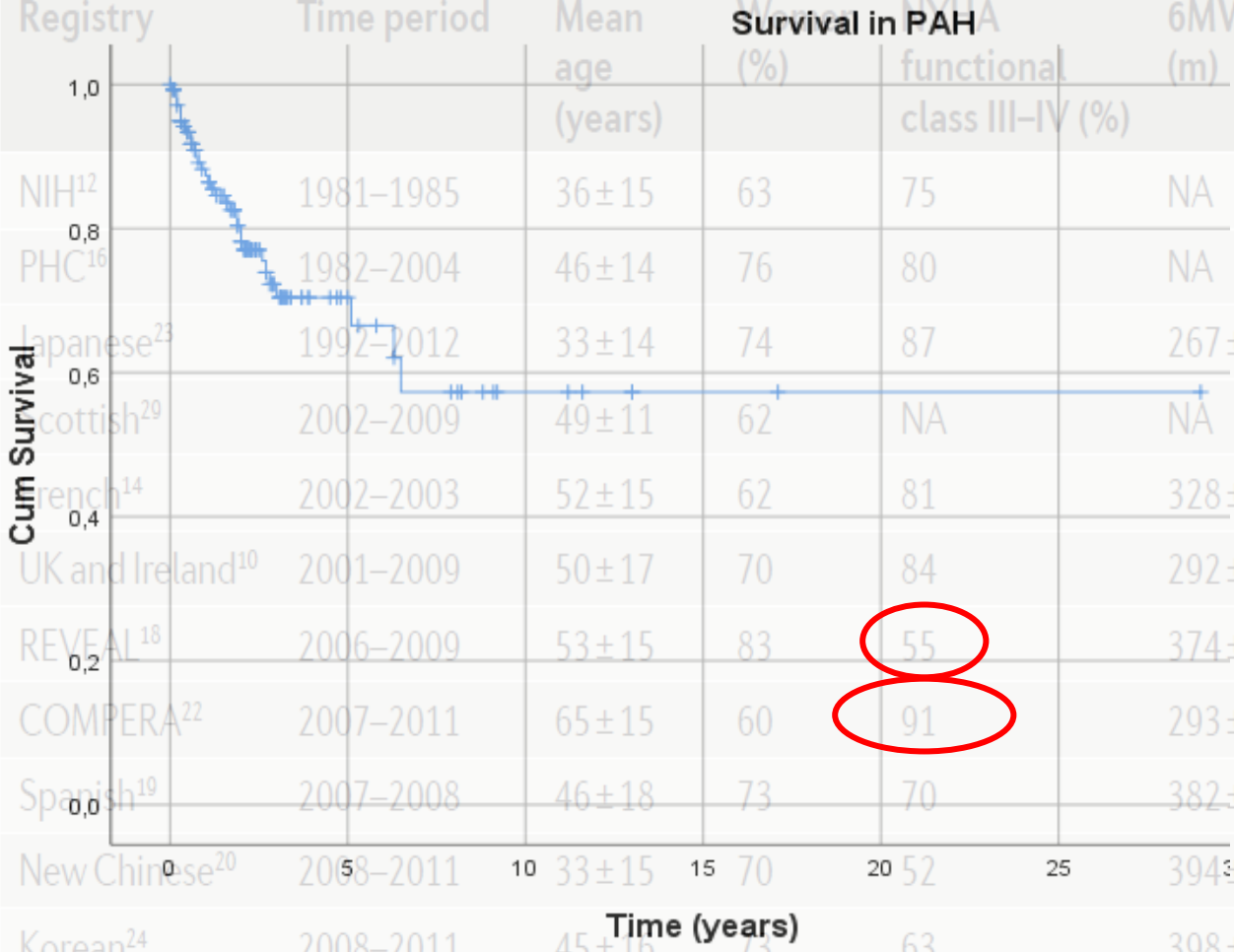
Data are presented as mean±SD, n (%) or median (interquartile range), unless otherwise stated. BMI: body mass index; PAH: pulmonary arterial hypertension; I/D/H: idiopathic, drug-associated or hereditary; CTD: connective tissue disease; PoPH: portopulmonary hypertension; CHD: congenital heart disease; WHO FC: World Health Organization functional class; 6MWD: 6-min walking distance; NT-proBNP: N-terminal fragment of pro-brain natriuretic peptide; BNP: brain natriuretic peptide; mPAP: mean pulmonary arterial pressure; PAWP: pulmonary arterial wedge pressure; PVR: pulmonary vascular resistance; Sv_{O_2}: mixed venous oxygen saturation; CCB: calcium channel blocker; ERA: endothelin receptor antagonist; PDE-5i: phosphodiesterase-5 inhibitor; sGCs: stimulator of soluble guanylate cyclase; PCA: prostacyclin analogue.

Hellenic PulmOnary Hypertension rEgistry (HOPE)
18. Survival Analysis

Greek PAHTool Registry

33 deaths / 143 patients in 394 patient-years

Median FU = 2 years (2.45)

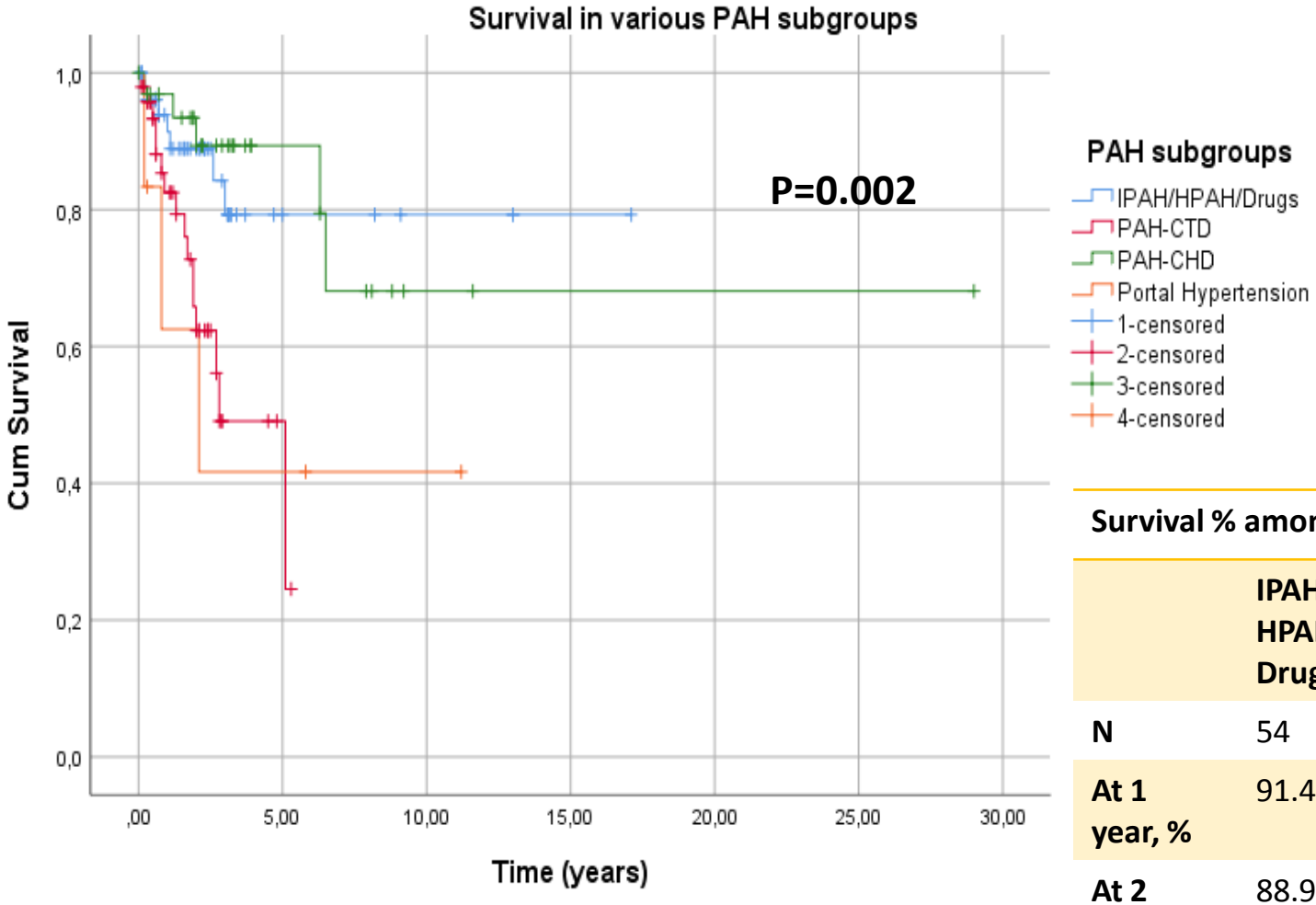


Survival in PAH, %	
At 1 year, %	87.3
At 2 year, %	78.2
At 3 year, %	70.4

Mortality rate: 8,3 deaths / 100 patients-years

Hellenic PulmOnary Hypertension rEgistry (HOPE)
18. Survival Analysis

Greek PAHTool Registry



Survival % among PAH subgroups

	IPAH/ HPAH/ Drugs	PAH-CTD	PAH-CHD	PoPAH
N	54	48	53	6
At 1 year, %	91.4	82.5	96.9	62.5
At 2 years, %	88.9	62.3	89.4	41.7
At 3 years, %	79.3	49.1	**89.4**	**41.7**

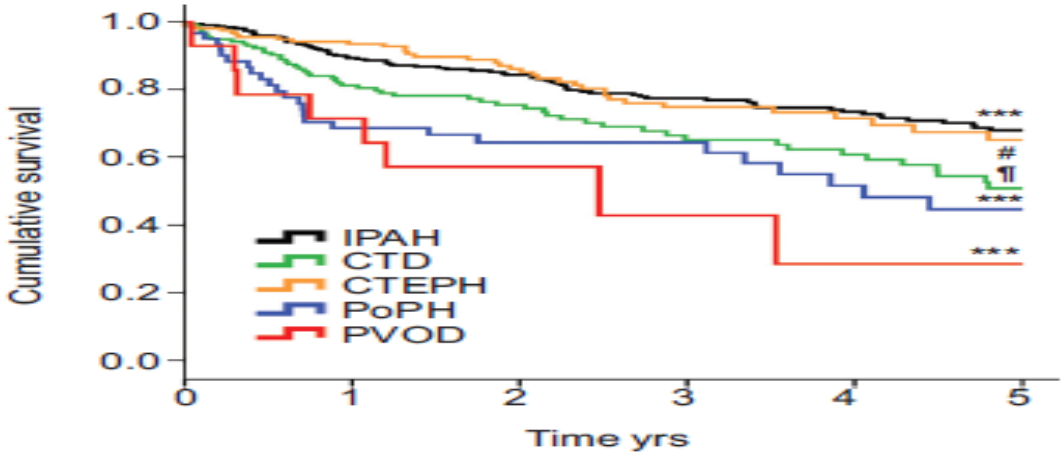
Median FU = 2 years (2.45)

Hellenic PulmOnary Hypertension rEgistry (HOPE)
18. Survival Analysis

Greek PAHTool Registry

Survival in pulmonary hypertension in Spain: insights from the Spanish registry

Pilar Escribano-Subias*, Isabel Blanco#, Manuel López-Meseguer¶, Carmen Jimenez Lopez-Guarch*, Antonio Roman¶, Pilar Morales+, María Jesús Castillo-Palma§, Javier Segoviaƒ, Miguel A. Gómez-Sanchez* and Joan Albert Barberà# on behalf of the REHAP investigators**

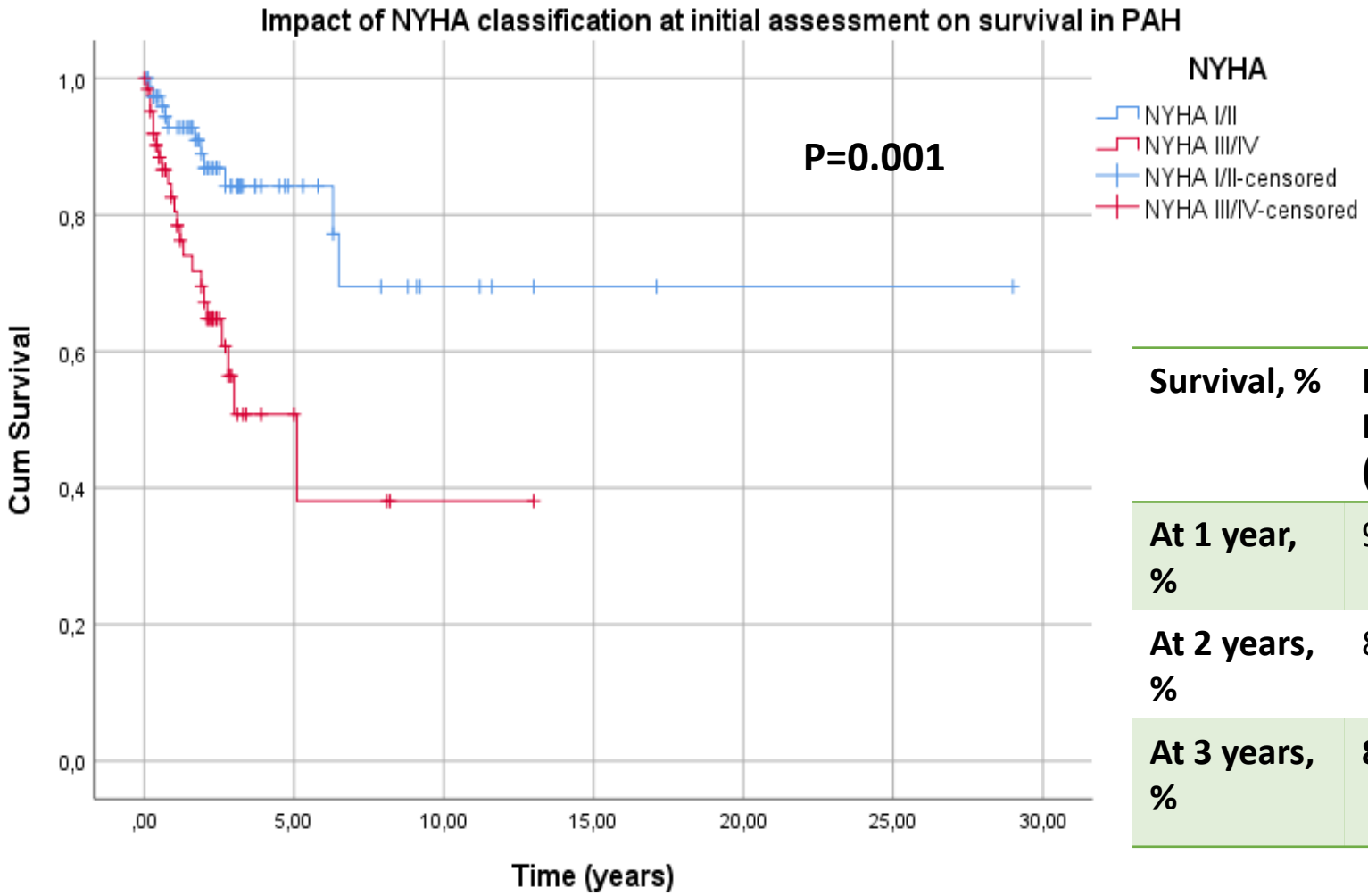


At risk n							p-value#
IPAH	314	304	236	179	136	108	
CTD	157	149	98	67	46	37	0.023
CTEPH	162	153	117	82	51	34	0.951
PoPH	61	58	34	26	21	14	<0.001
PVOD	15	14	8	4	3	2	<0.001

FIGURE 1. Kaplan–Meier estimates of 5-yr survival from time of diagnosis in different pulmonary hypertension subtypes. IPAH: idiopathic pulmonary arterial hypertension; CTD: connective tissue disease; CTEPH: chronic thromboembolic pulmonary hypertension; PoPH: portopulmonary hypertension; PVOD: pulmonary veno-occlusive disease. The p-value for the overall comparison is <0.001. #: compared with IPAH.

Hellenic PulmOnary Hypertension rEgistry (HOPE)
18. Survival Analysis

Greek PAHTool Registry

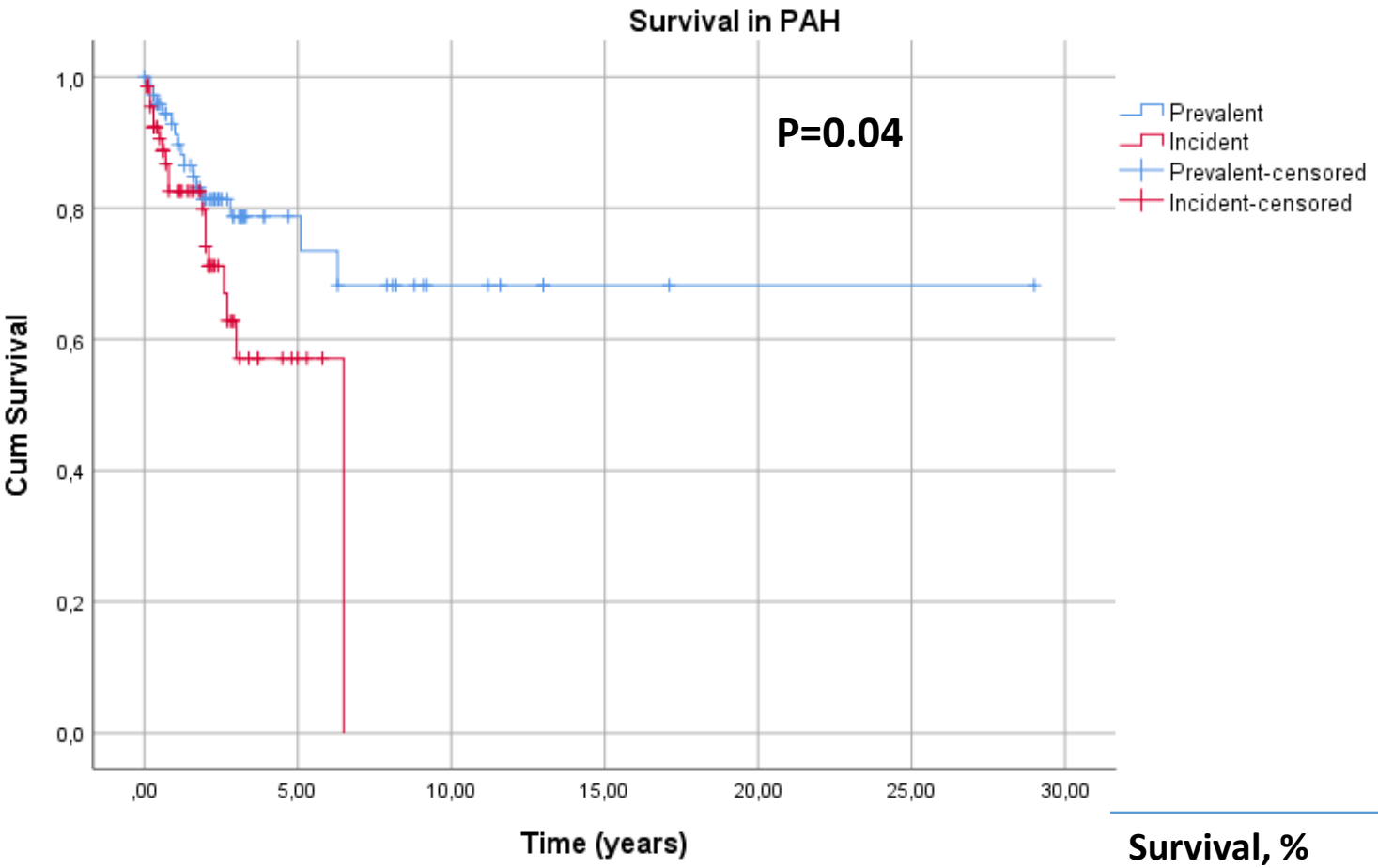


Survival, %	NYHA I/II (N=79)	NYHA III/IV (N=64)
At 1 year, %	92.8	80.5
At 2 years, %	86.9	67.2
At 3 years, %	**84.2**	**50.8**

Median FU = 2 years (2.45)

Hellenic PulmOnary Hypertension rEgistry (HOPE)
18. Survival Analysis

Greek PAHTool Registry

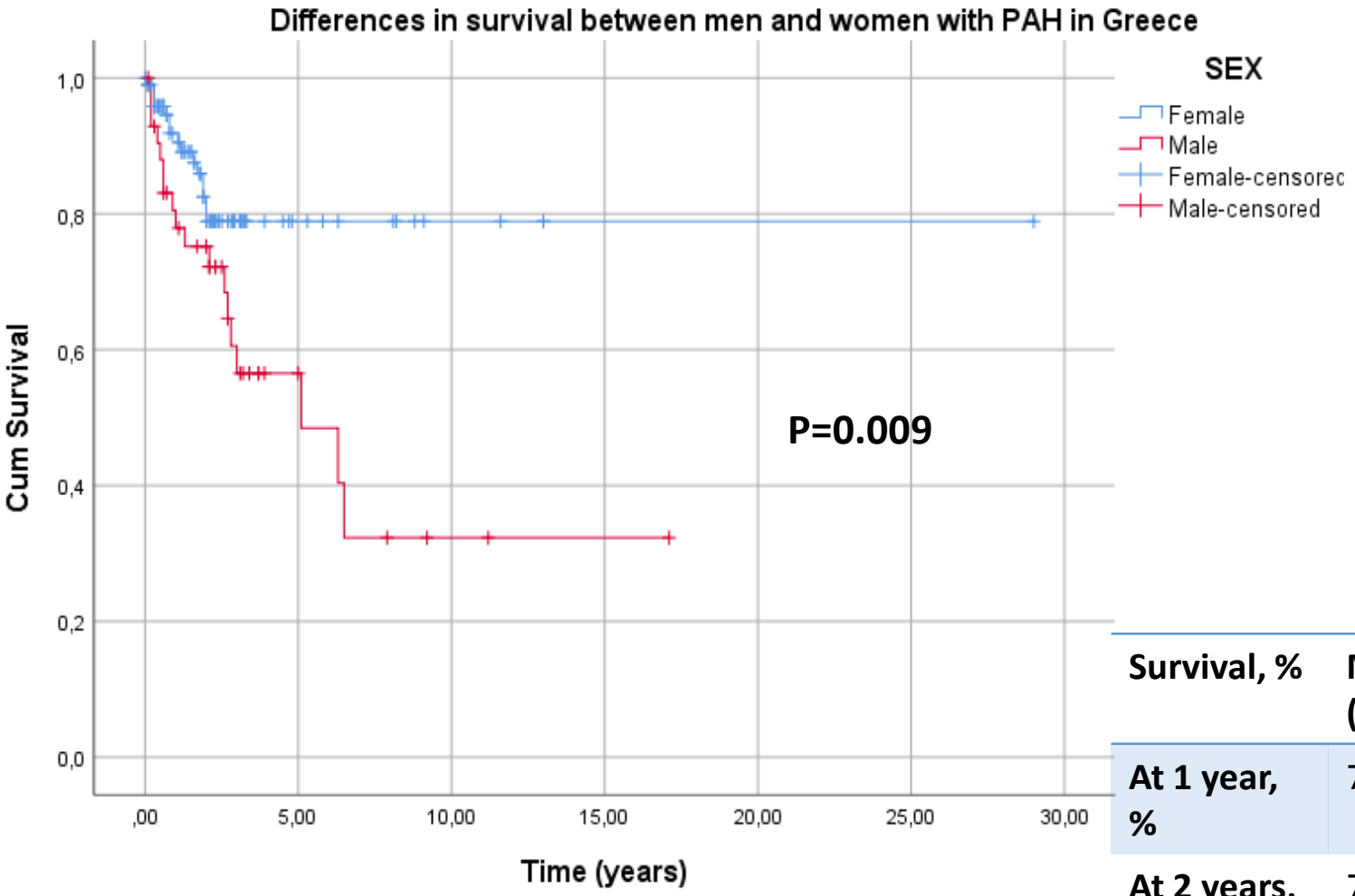


Median FU = 2 years (2.45)

Survival, %	Incident (N=69)	Prevalent (N=74)
At 1 year, %	82.6	91.3
At 2 years, %	74.1	81.4
At 3 years, %	**57.1**	**78**

Hellenic PulmOnary Hypertension rEgistry (HOPE)
18. Survival Analysis

Greek PAHTool Registry

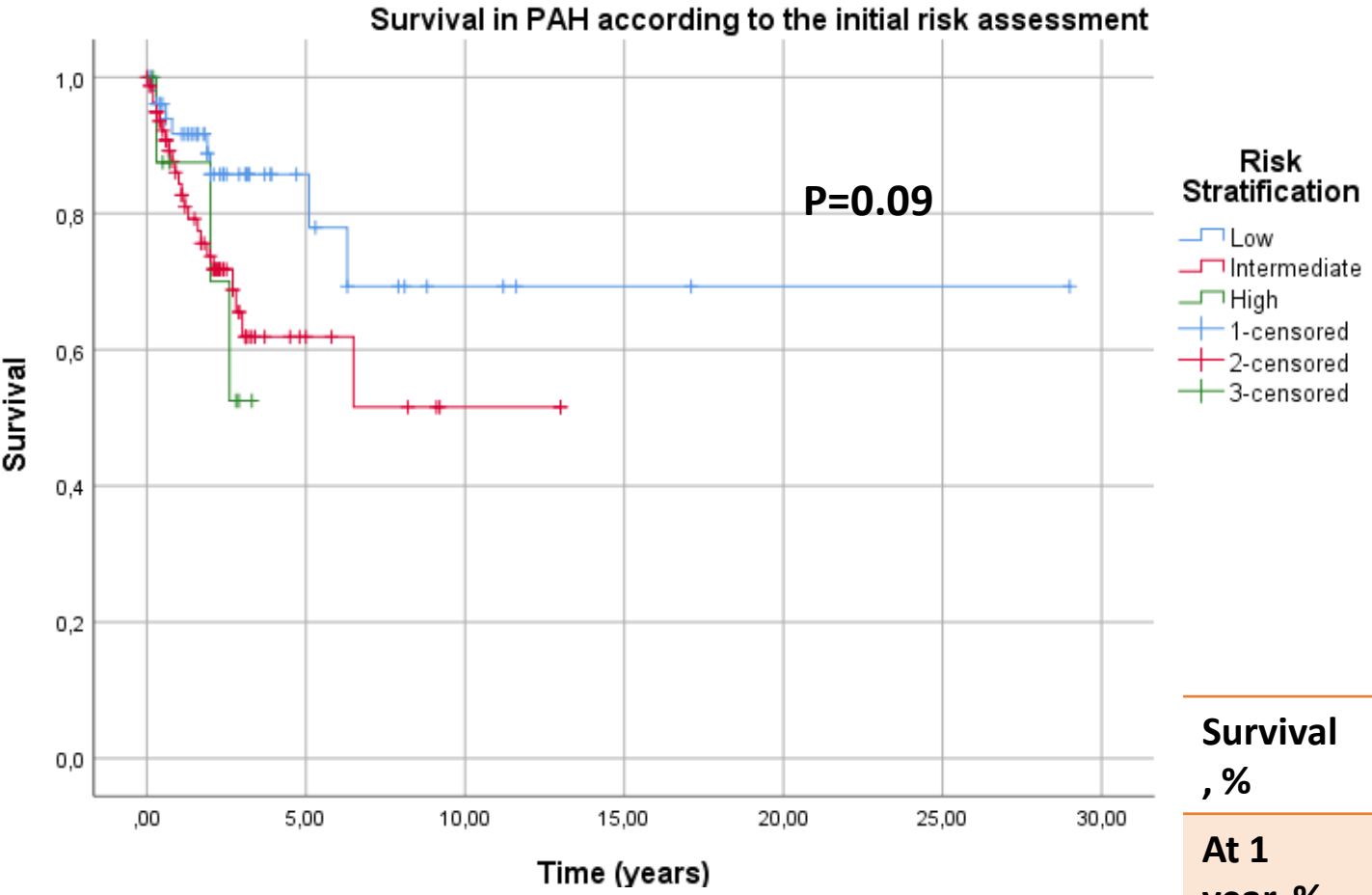


Survival, %	M (N=43)	F (N=100)
At 1 year, %	77.9	91.9
At 2 years, %	75.2	82.5
At 3 years, %	**56.5**	**78.9**

Median FU = 2 years (2.45)

Hellenic PulmOnary Hypertension rEgistry (HOPE)
18. Survival Analysis

Greek PAHTool Registry



Median FU = 2 years (2.45)

Survival, %	Low N=53	Median N=81	High N=9
At 1 year, %	91.7	84.4	87.5
At 2 years, %	85.7	73.7	70
At 3 years, %	**85.7**	**61.9**	**52.5**

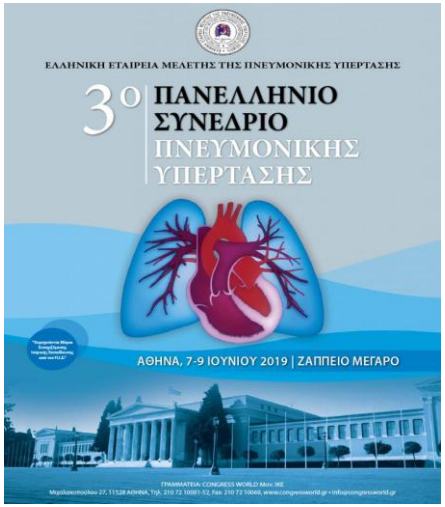
Hellenic PulmOnary Hypertension rEgistry (HOPE)
19. Take home messages

Greek PAHTool Registry

- 280 patients with PAH were recorded.
- Nearly half patients (45%) were newly diagnosed.
- Female sex was the dominant sex among all subgroups except from PoPAH.
- The majority of patients (57.1%) had no or mild symptoms and were at intermediate 1-year mortality risk (56.8%).
- The most common comorbidities were hypertension and obesity.
- Over time there was a significant improvement in hemodynamics and 6MWD.
- There was an increased use of combination therapy and prostanoids especially IP receptor agonists over time.
- 1- and 3-year survival in PAH was 87.3% and 70% respectively.
- The lowest survival rate was detected in PoPAH and PAH-CTD subgroups, in men, incident patients and in severely symptomatic patients (NYHA III/IV).

Greek PAHTool Registry

Ευχαριστώ



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