



Greek Pulmonary Hypertension Registry

Insights from patients with PAH and CTEPH

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On behalf of the Hellenic Society for the Study of Pulmonary Hypertension (HSSPH)



Conflicts of interest

- Advisory Board
 - Astra Zeneca, Bayer, ELPEN, Actelion, MSD, NOVARTIS

- ✓ **Retrospective and prospective (starting from 2015) data collection**
 - PH diagnosis, classification, symptoms, comorbidities, diagnostic procedures and treatment
 - Patients who had full work up, including a 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**

Greek Pulmonary Hypertension Registry Expert Centers

9 expert centers in Greece

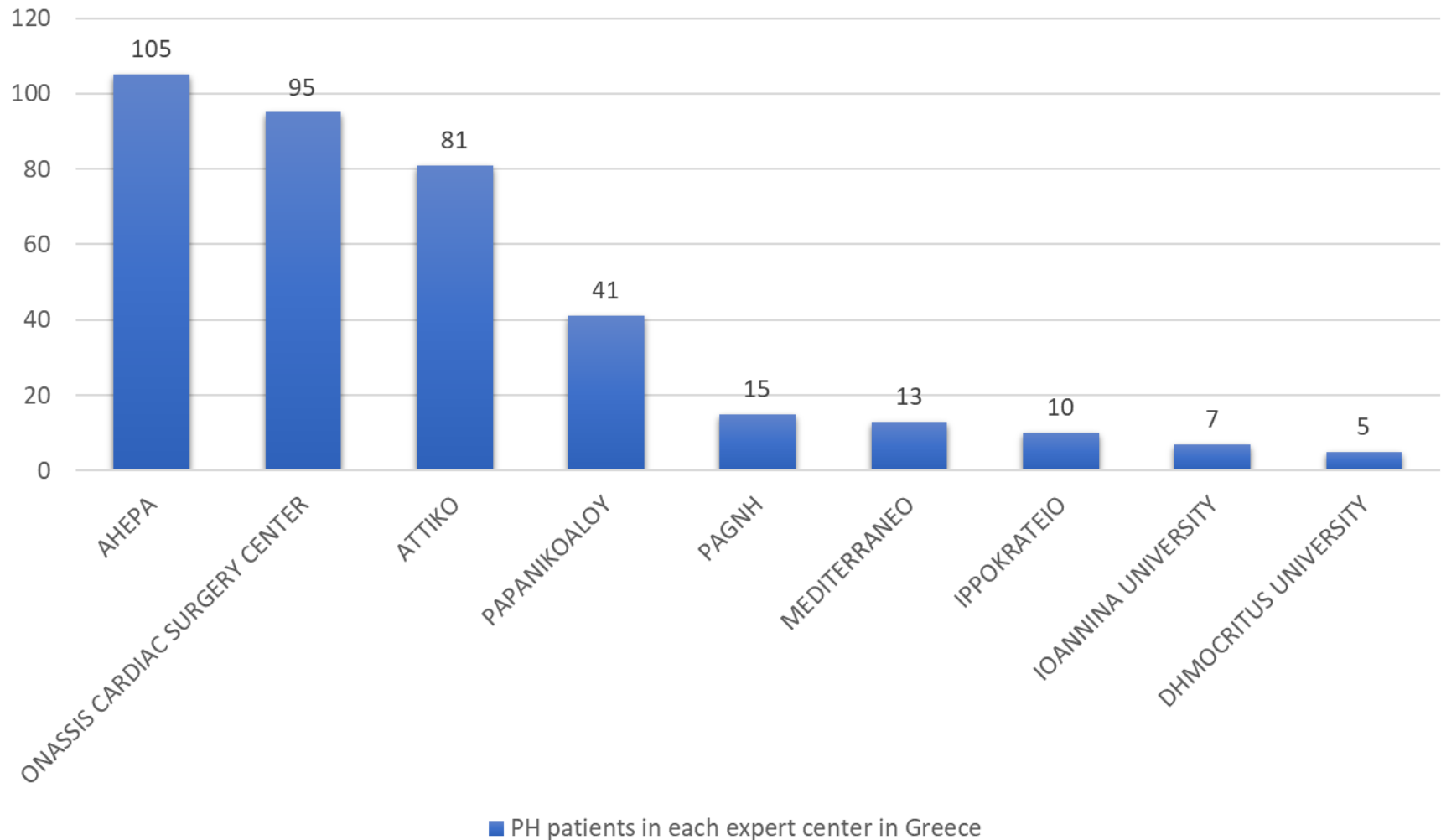
1. Cardiology Department, AHEPA University Hospital, Thessaloniki
2. Multidisciplinary Pulmonary Hypertension Service, Attiko General Hospital, Athens
3. Cardiology- Pediatric Cardiology Department, Onassis Cardiac Surgery Center, Athens
4. Cardiology Department, Mediterraneo Hospital, Glifada, Athens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



Greek Pulmonary Hypertension Registry Center participation

Greek
PAHTool
Registry

PH patients in each expert center in Greece



Greek Pulmonary Hypertension Registry

Data Management



Center number: null ▼

Theme ▼ Language ▼ Hi George Giannakoulas ▼

First visit

ID

CC

PH

CH

6T

HD

EC

OT

BT

Tx

Previous

Next

Ok

Close

Identification

Last name

First name

Gender

Male

Female

Ethnicity/Race

[Select a ethnicity] ▼

Date of last news

Date

05/06/2018

Global number

0

Center number

AM

AMKA

Clinical trials

Protocol Name ^

Inclusion date

End of study date

Add

Edit

Close

Delete

Birth

Birth Date

dd/mm/yyyy

Country

[Select a country] ▼

Zip Code

City

Address

Contacts

Other information

BioBank

Yes

No

informed consent authorization

Yes

No

Wizard

☒ Classification

☒ Previous history

☒ Current history

☒ 6 MWT

☒ Hemodynamics

☐ Echocardiogram

☐ Other tests

- ☐ ABG
- ☐ ECG
- ☐ Chest radiograph
- ☐ CPET
- ☐ Lung function tests
- ☐ Pulmonary angiography
- ☐ Chest CT Scan
- ☐ Ventilation/Perfusion lung scan
- ☐ Abdominal ultrasound
- ☐ Polysomnography
- ☐ MRI

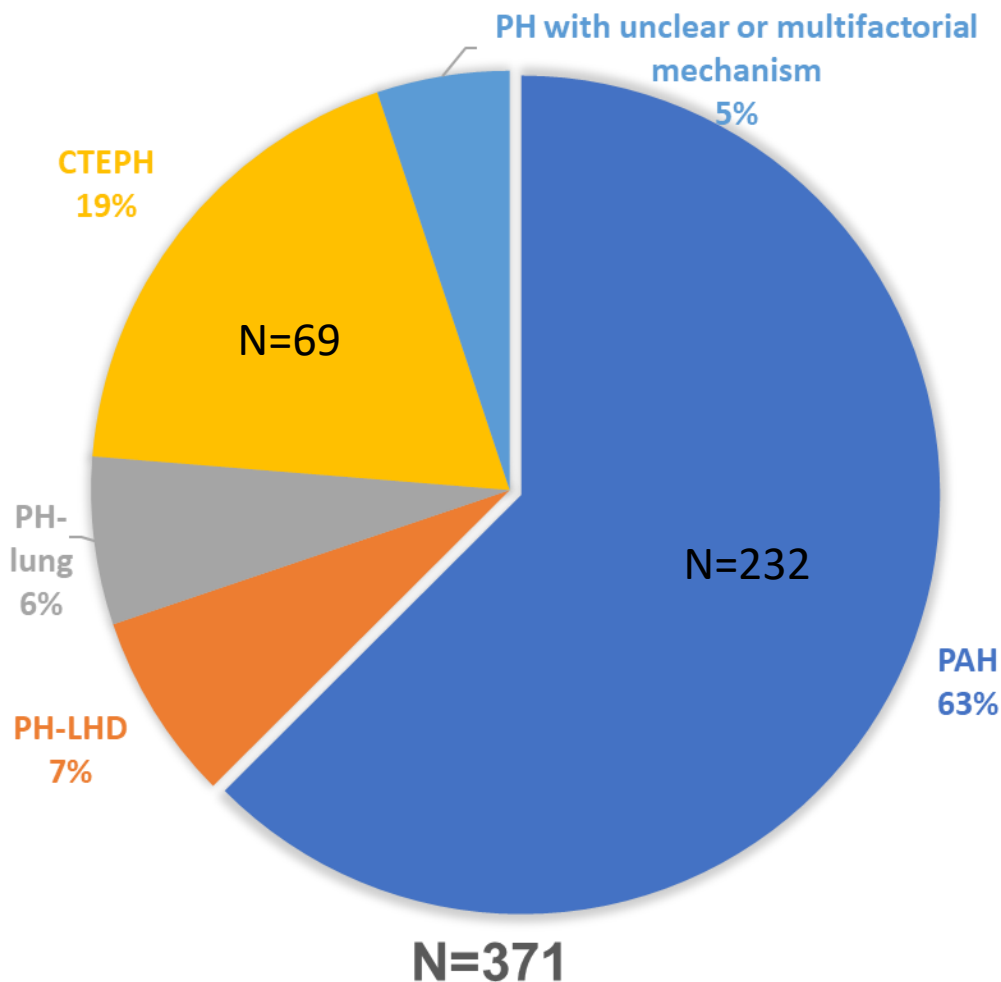
☐ Biological tests

- ☐ RBC and iron balance
- ☐ Biomarkers

Greek Pulmonary Hypertension Registry

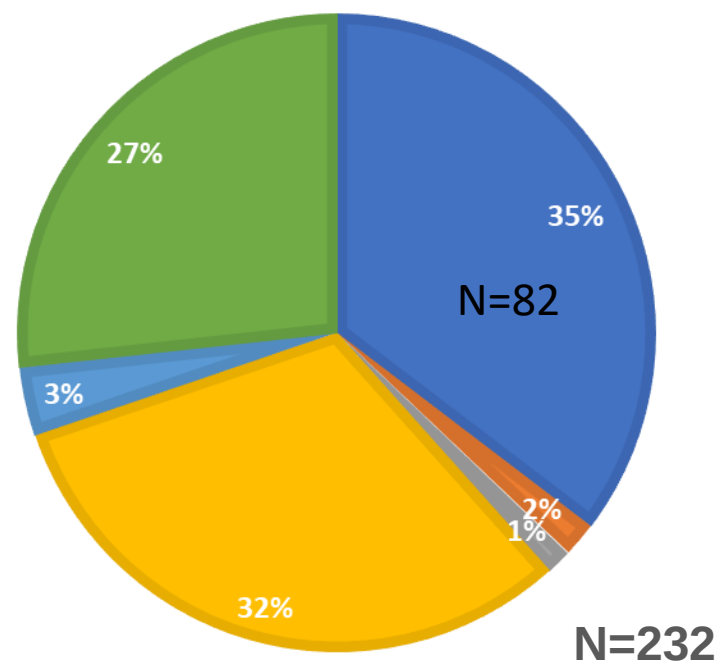
PH and PAH Classification (2015-2018)

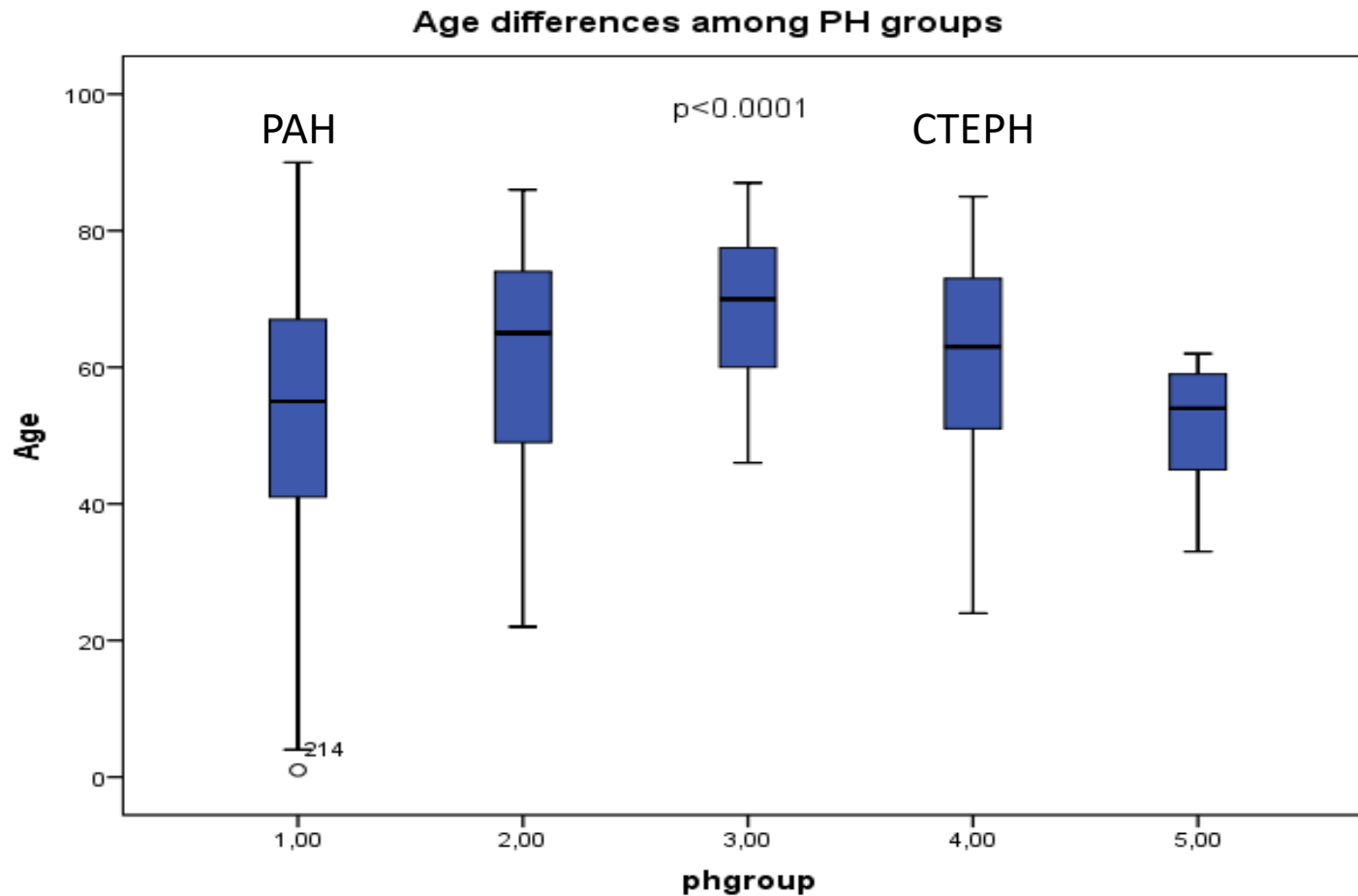
CLASSIFICATION OF PATIENTS WITH
PULMONARY HYPERTENSION IN
GREECE



CLASSIFICATION OF PATIENTS
WITH PAH IN GREECE

- Idiopathic PAH
- Heritable PAH
- Drugs and toxins induced PAH
- Connective tissue disease
- Portal hypertension
- Congenital heart disease





Greek PAH registry

Demographics

Demographics of patients with PAH (N=232)

	N(%) / median (IQR)
Age, (years)	55.0 (27.0)
Female sex, n (%)	149 (64.2)
BMI, (Kg/m ²)	25.5 ± 7.7
Incident patient, n (%)	89 (38.4)

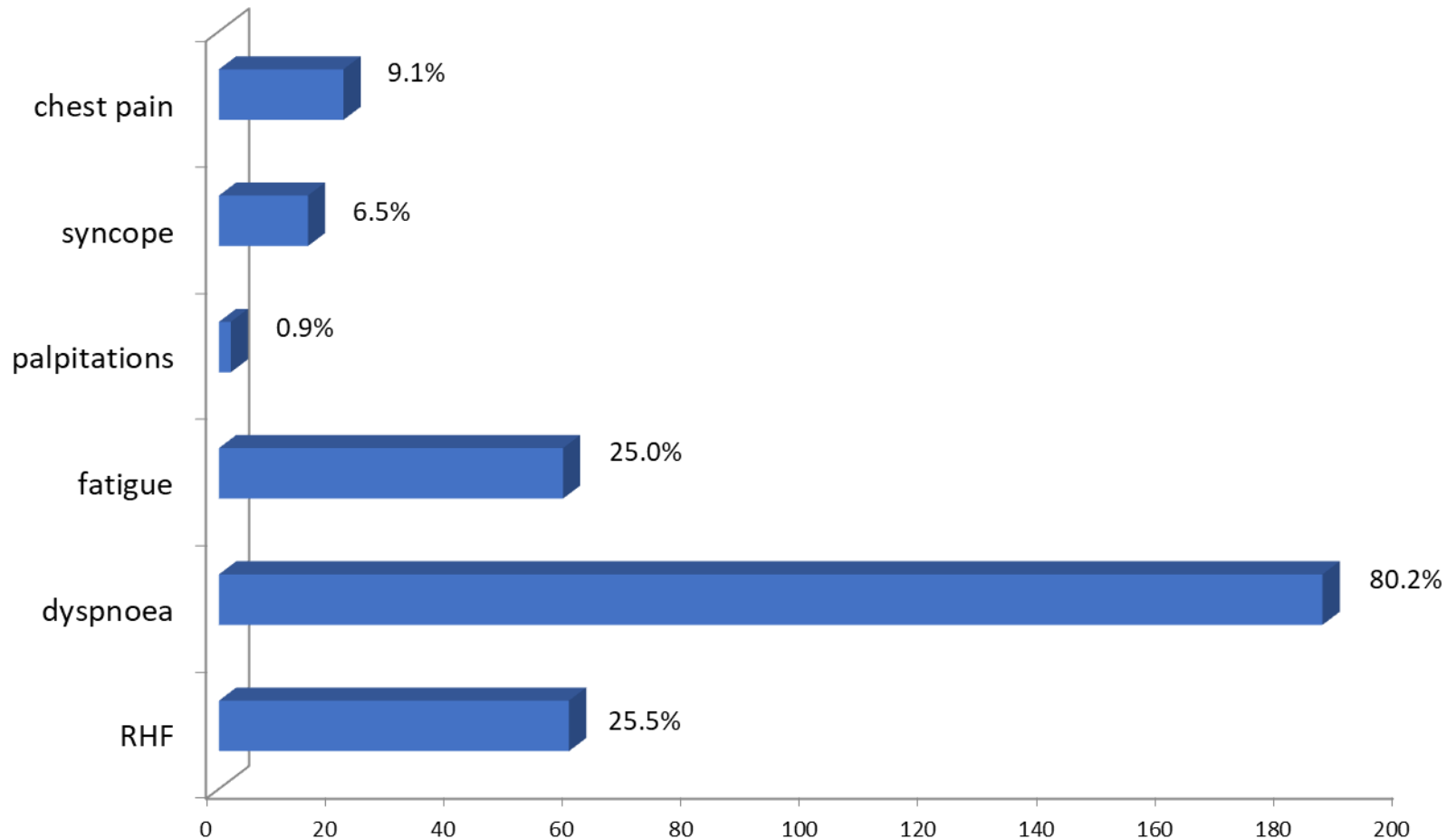
Comorbidities of patients with PAH

	N (%)
Atrial fibrillation	8(3.4)
Arterial hypertension	58(25.0)
Diabetes mellitus	29(12.5)
Dyslipidemia	27(11.6)
CAD	16(6.9)
Dialysis	1(0.4)
Liver disease	12(5.2)
Previous PE	1(0.4)
Previous DVT	1(0.4)
Splenectomy	0(0.0)
Hematological disease	10(4.3)
Thyroid disease	37(15.9)
Obesity (BMI≥30kg/m ²)	59(25.4)
COPD	20(8.6)
Smoking	9(3.9)
Depression	1(0.4)

Greek PAH registry

Symptoms

Most frequent symptoms in patients with PAH

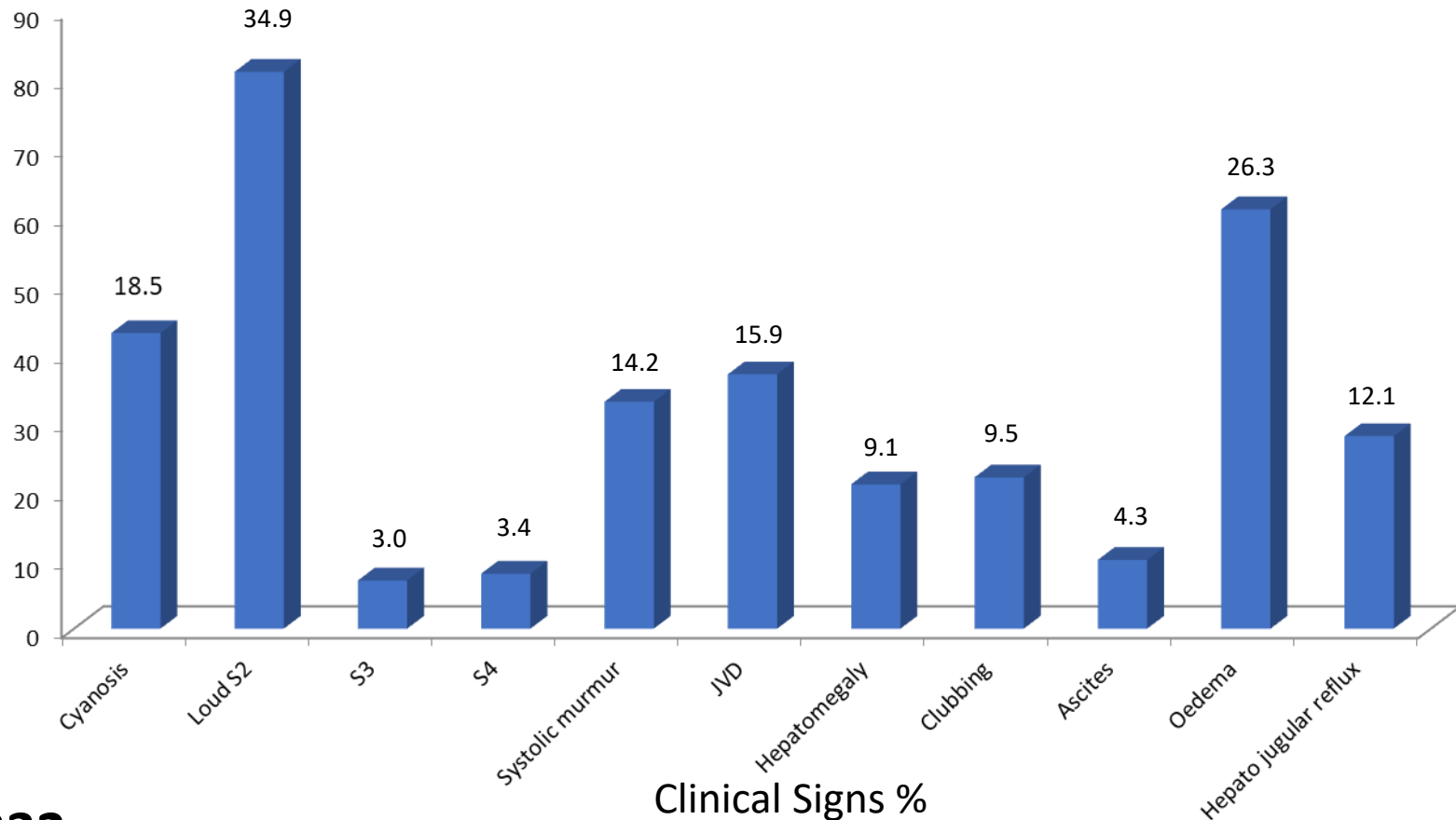


N=232

Greek PAH registry

Clinical Signs

Clinical Signs in patients with PAH

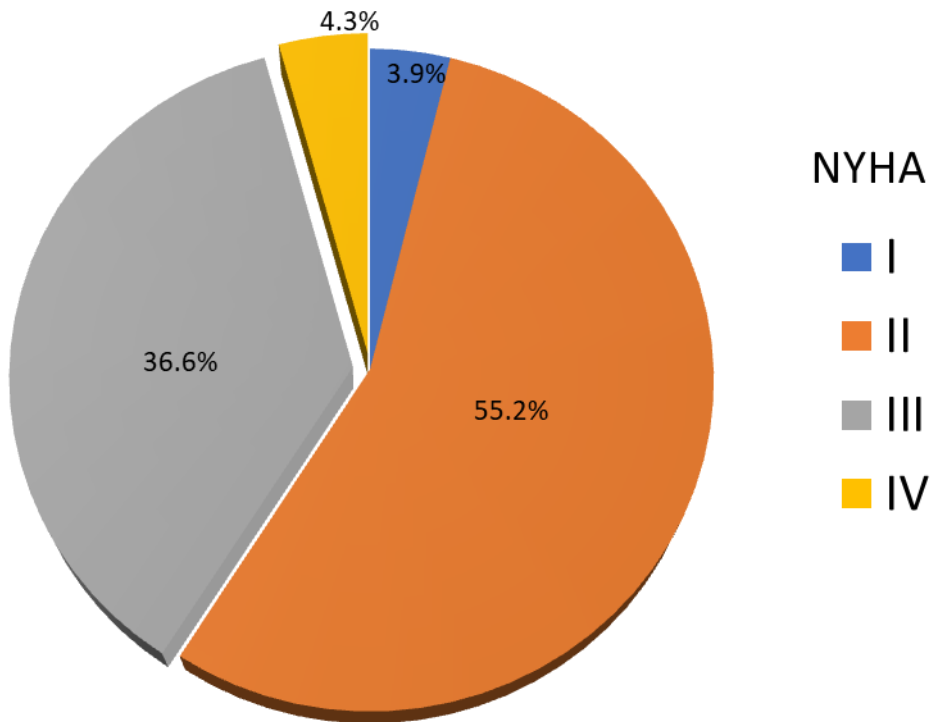


N=232

Greek PAH registry

Functional and neurohormonal characteristics

WHO FUNCTIONAL CLASS



Functional and neurohormonal indices

6-MWD, (m) (n=17)	335.0 (216.5)
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NT-proBNP, (pg/mL) (n=36)	174.0 (376.0)
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N=232

Greek PAH registry

Diagnostic parameters (I)

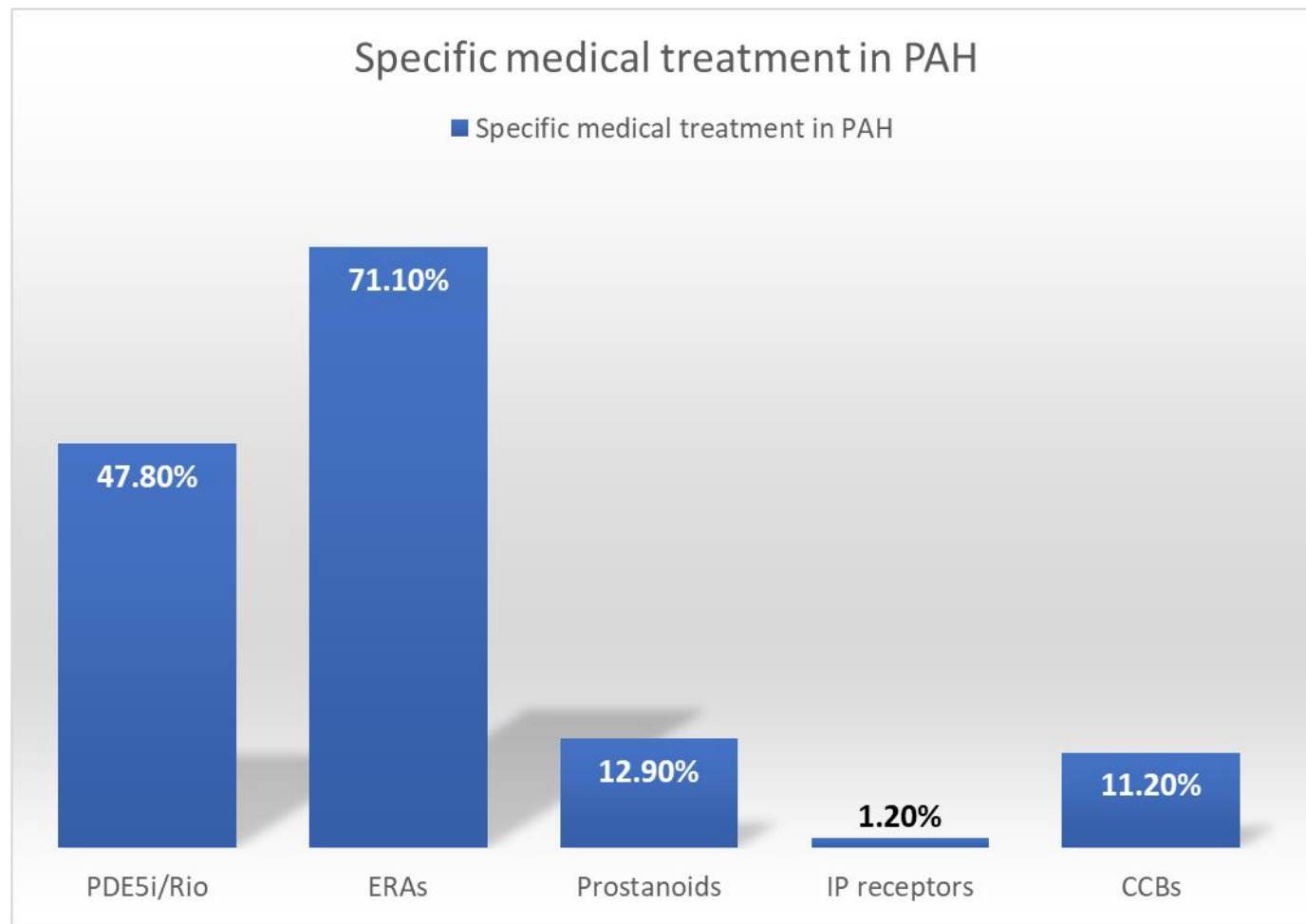
Echocardiography	N(%) / Median (IQR)
LVEF %	61.0(13.0)
RV hypertrophy,	49.0(21.1)
RV enlargement	97.0(41.8)
RVEDD, mm	32.0(12.0)
Abnormal RV function (severe)	25.0(10.8)
TAPSE, mm	18.0(6.0)
TR Vmax, m/s,	3.9(0.9)
RVSP, mmHg	60.8(28.4)

N=232

Hemodynamics, Median (IQR)

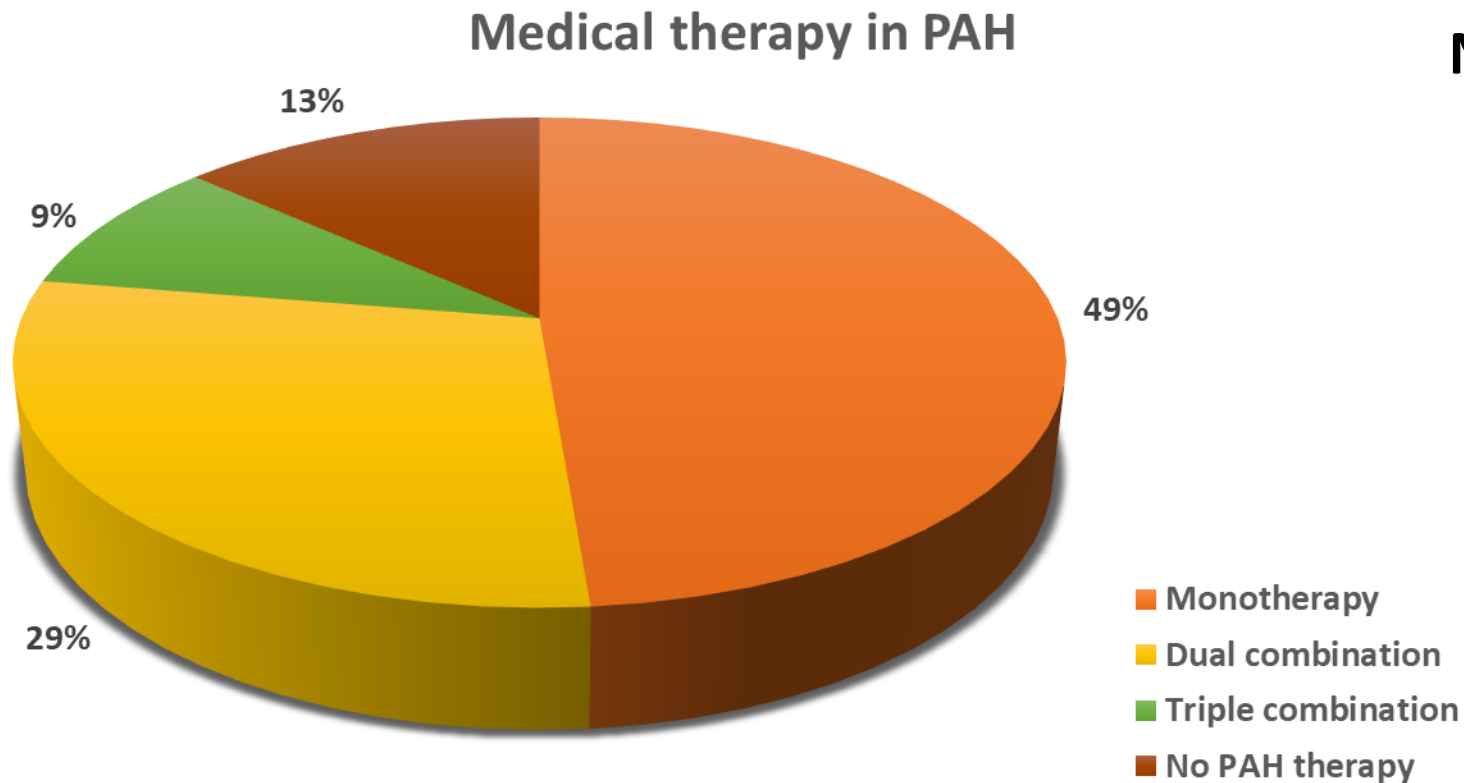
RAP, (mmHg)	6.0(9.5)
sPAP, (mmHg)	75.0(20.0)
dPAP, (mmHg)	28.0(15.0)
mPAP, (mmHg)	60.0(6.0)
PAWP, (mmHg)	9.0(5.5)
CO, (L/min)	4.5(2.15)
CI, (L/min/m ²)	2.6(1.0)
PVR, (WU)	8.4(5.2)
PVRi, (WU*m2)	15.7(9.3)
SVO ₂ , (%)	68.0(19.0)
HR, (bpm)	77.0(16.0)

N=232



Greek Pulmonary Hypertension Registry

Medical therapy in PAH patients (II)



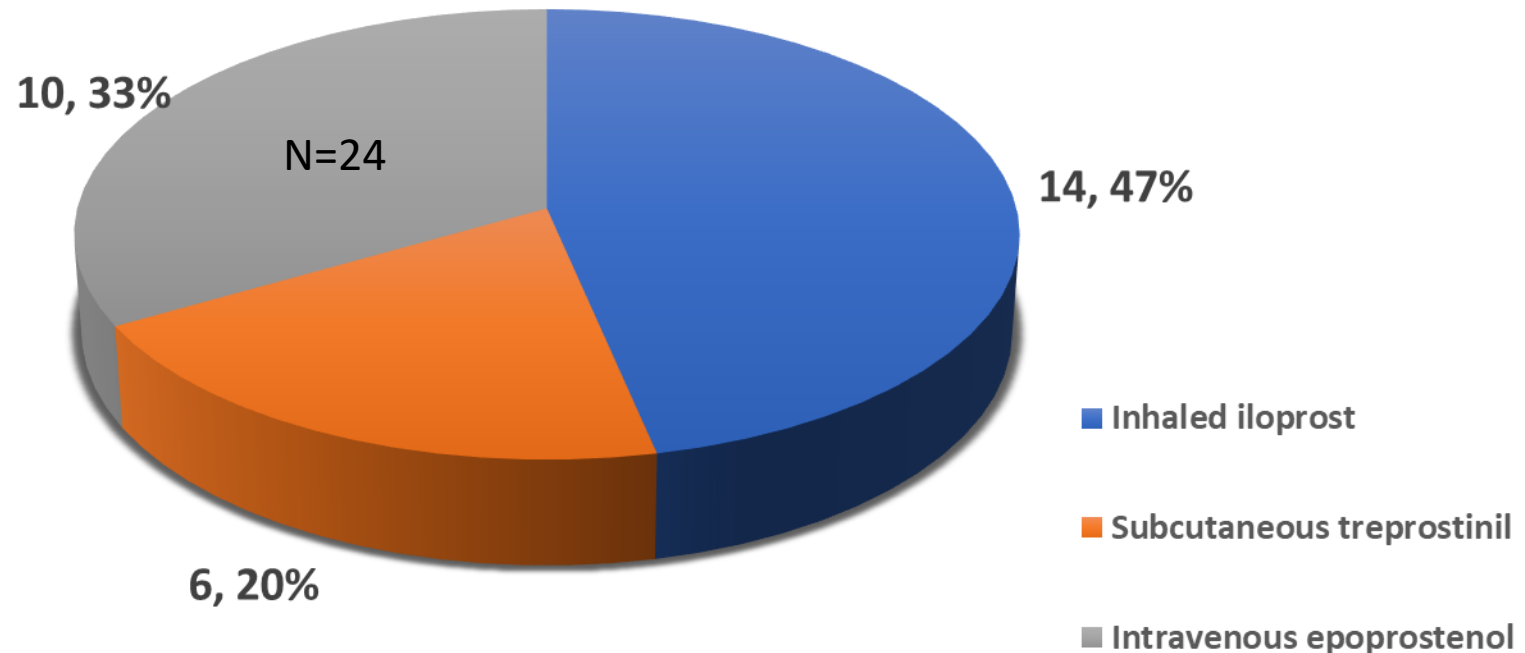
Dual combination	PAH (N=68), n (%)
PDE5i/Rio + ERA	57 (84)
PDE5i/Rio + Prostacyclin analogue	8 (11.7)
ERA+ Prostacyclin analogue	3 (3.7)

Greek Pulmonary Hypertension Registry

Medical therapy in PAH patients (III)

Prostacyclin analogues use in patients with PAH

N=30



Greek CTEPH registry

Demographics

N=69

Demographics of patients with CTEPH

	N=69
Age, (years)	62.8 ± 16.2
Female sex, n (%)	39 (56.5)
BMI, (Kg/m ²)	28.3 ± 5.7
Incident patient, n (%)	35 (50.7)

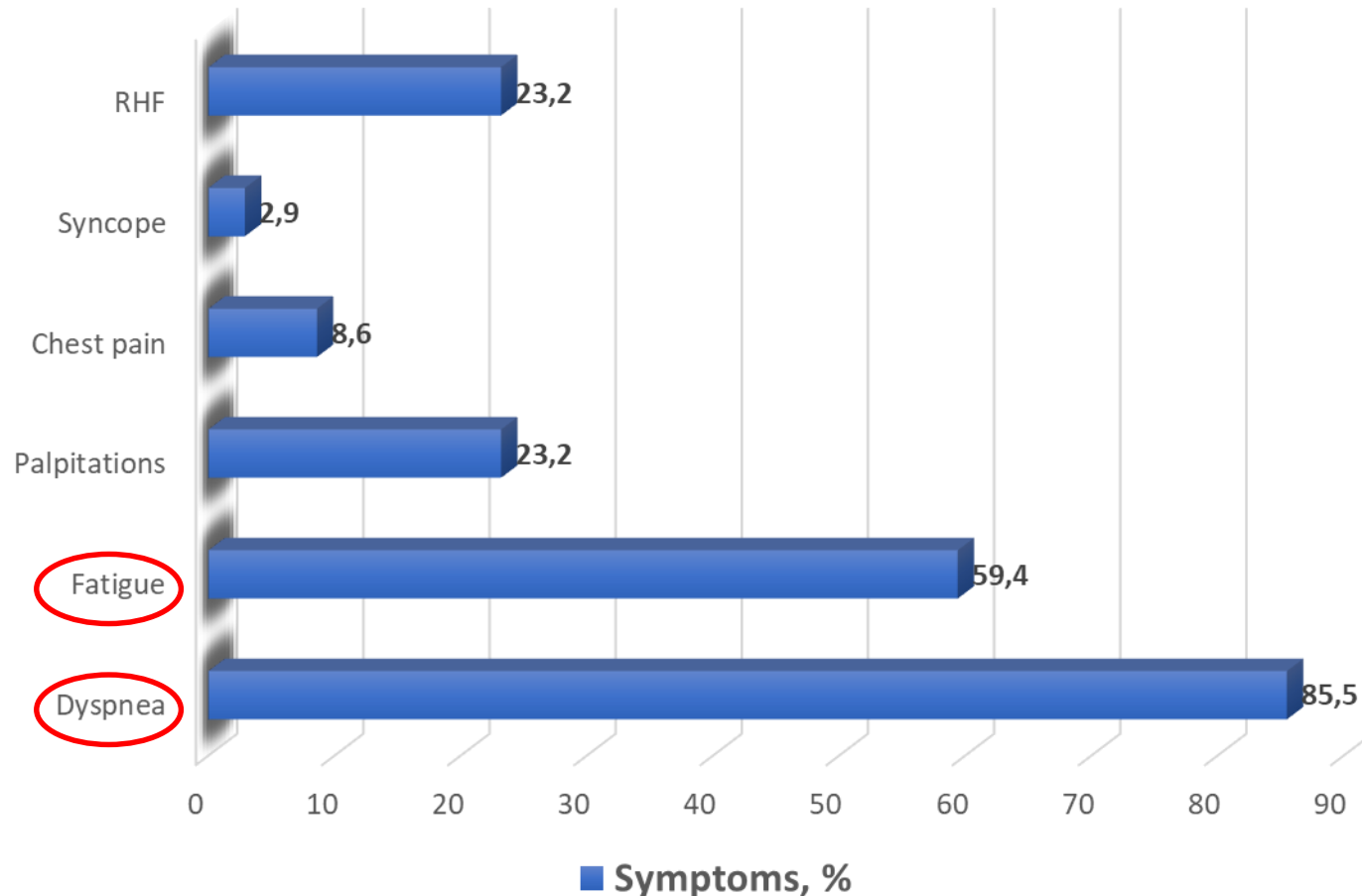
Comorbidities of patients with CTEPH

	N (%)
Atrial fibrillation	1 (1.4)
Arterial hypertension	27 (37.1)
Diabetes mellitus	6 (8.6)
Dyslipidemia	7 (10.1)
CAD	0
Dialysis	1 (1.4)
Liver disease	0
Previous PE	38 (55)
Previous DVT	4 (5.8)
Splenectomy	7 (10.1)
Hematological disease	16 (23.2)
Thyroid disease	8 (11.6)
Obesity (BMI≥30kg/m ²)	17 (24.6)
COPD	9 (13)
Smoking	19 (27.5)
Depression	5 (7.2)

Greek CTEPH registry

Symptoms

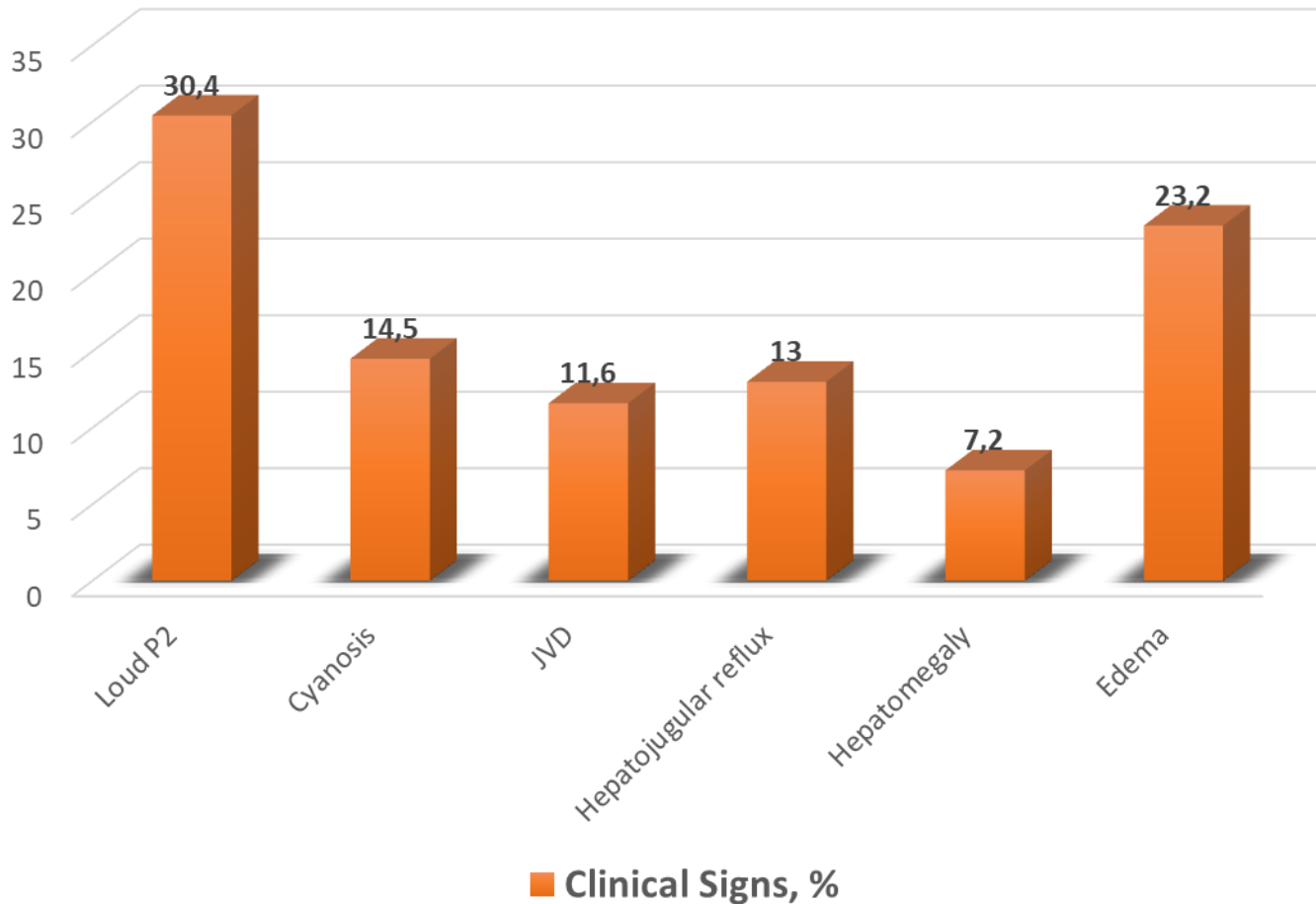
Most frequent symptoms in patients with CTEPH



Greek CTEPH registry

Clinical Signs

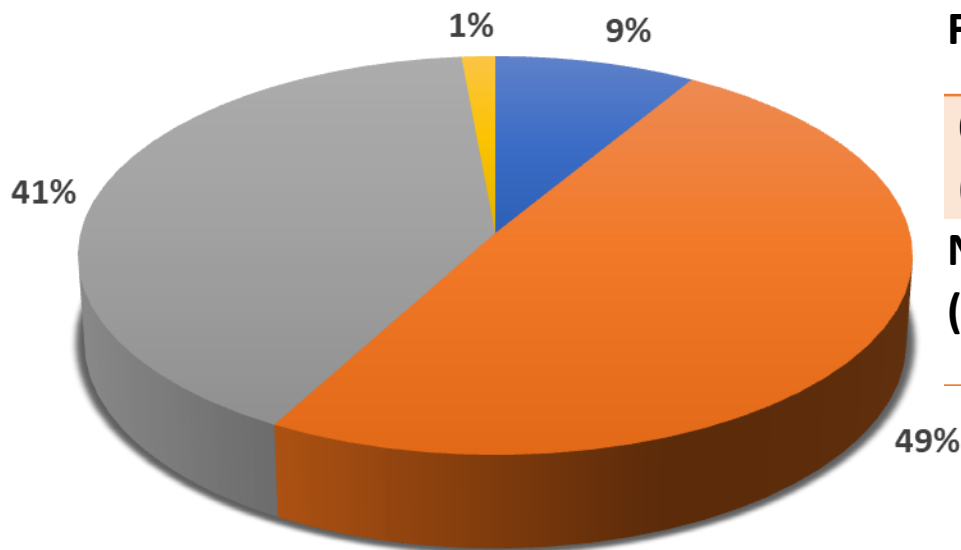
Clinical Signs in patients with CTEPH



Greek CTEPH registry

Functional and neurohormonal characteristics

WHO functional class



■ NYHA I ■ NYHA II ■ NYHA III ■ NYHA IV

Functional and neurohormonal indices

6-MWD, (m) (n=58)	333.3 ± 165.5
NT-proBNP, (pg/mL) (n=36)	356 (10 - 3220)

N=69

Greek CTEPH registry

Diagnostic parameters (I)

Lung function test, median (IQR)

FEV1, (% pred)	83.6(70.0-97.0)
FVC, (% pred)	82.1(74.8-98.5)
FEV1/FVC, (%pred)	76.99(71.5-82.8)
TLC, (%pred)	82.0(73.0-97.0)
DLCO/SB, (%pred)	69.0(57.2-79.0)

Other diagnostic procedures

V/Q scintigraphy	44 (63.8)
Abnormal V/Q scan	31 (70.5)
Chest CT	35 (50.7)
Central thrombi	11 (31.4)
Peripheral thrombi	16 (45.7)
Mural thrombi	6 (17.1)
Pulmonary Angiography	15 (21.7)
Proximal lesions	10 (66.7)

Greek CTEPH registry

Diagnostic parameters (II)

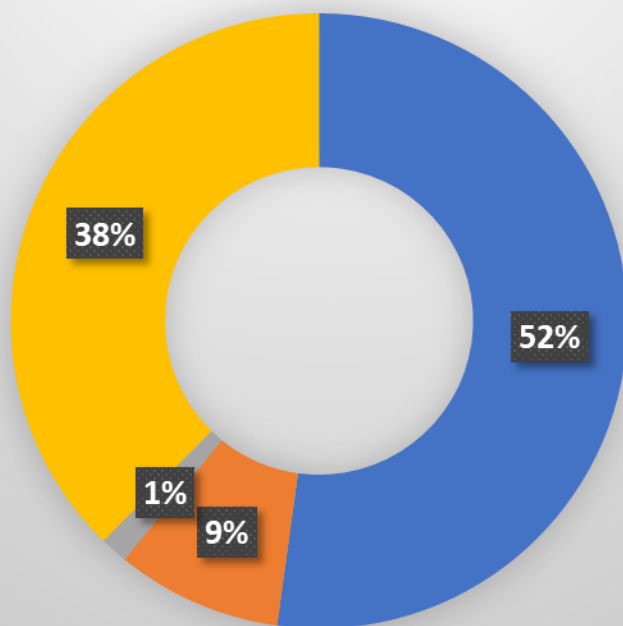
Echocardiography

LVEF, %	60 (50-68)
RV hypertrophy, n (%) (63.7%)	15 (34)
RV enlargement, n(%) (72.5%)	38 (76)
RVEDD, mm (31.9%)	32.9 ± 12.6
Abnormal RV function, n (%)	43 (62.3)
TAPSE, mm (n=39)	20 (9 – 28)
TR Vmax, m/s, (56.5%)	3.7 ± 0.7
RVSP, mmHg (65.2%)	71.7 ± 22.4

Hemodynamics, 94.2%

RAP, (mmHg)	6 (1-21)
sPAP, (mmHg)	74.1 ± 19.6
dPAP, (mmHg)	26.5 ± 7.4
mPAP, (mmHg)	43.4 ± 10.8
PAWP, (mmHg)	10.4 ± 3.4
CO, (L/min)	4.6 ± 1.4
CI, (L/min/m ²)	2.5 ± 0.7
PVR, (WU)	6.6 (2.6-18.2)
PVRi, (WU*m2)	12.6 (4.3-36)
SVO ₂ , (%)	66.2 ± 9.6
HR, (bpm)	78.4 ± 10.2

PAH targeted therapy in CTEPH patients N=69



- Monotherapy
- Dual combination
- Triple combination
- No medical therapy

Circulation



Chronic Thromboembolic Pulmonary Hypertension (CTEPH): Results From an International Prospective Registry

Joanna Pepke-Zaba, Marion Delcroix, Irene Lang, Eckhard Mayer, Pavel Jansa, David Ambroz, Carmen Treacy, Andrea M. D'Armini, Marco Morsolini, Repke Snijder, Paul Bresser, Adam Torbicki, Bent Kristensen, Jerzy Lewczuk, Iveta Simkova, Joan A. Barberà, Marc de Perrot, Marius M. Hoeper, Sean Gaine, Rudolf Speich, Miguel A. Gomez-Sanchez, Gabor Kovacs, Abdul Monem Hamid, Xavier Jaïs and G rald Simonneau

Circulation. 2011;124:1973-1981; originally published online October 3, 2011;
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

Table 6. PAH-Targeted Therapy Initiated at Diagnosis

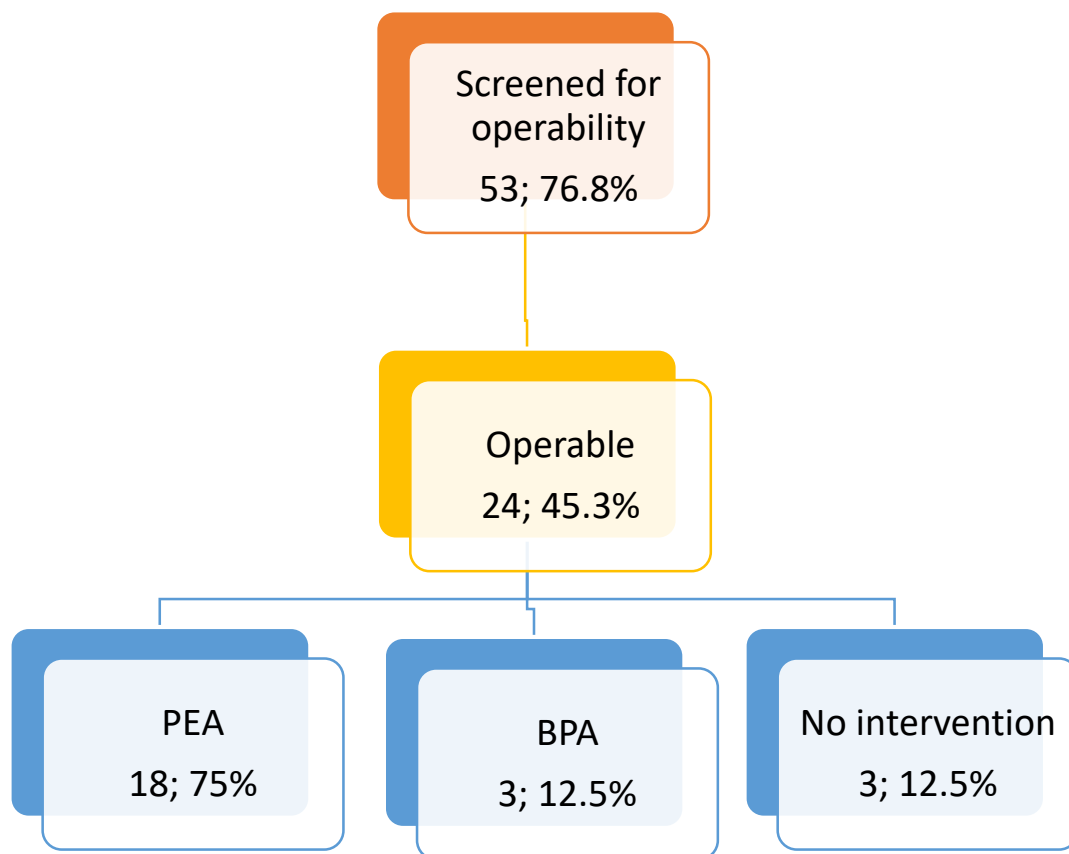
	All Patients (n=679)	Operable Patients* (n=427)	Nonoperable Patients* (n=247)	P (Exploratory)
PAH-targeted therapy, % (n)	37.9 (676)	28.3 (427)	53.8 (247)	<0.0001
Phosphodiesterase type V inhibitor, %	17.5	16.2	19.4	0.2923
Endothelin receptor antagonist, %	21.7	12.2	37.7	<0.0001
Prostacyclin analogue, %	2.7	1.6	4.5	0.0443
Combination therapies, %	4.0	1.6	7.7	0.0002

P values from Fisher exact test. (n): patients with assessment. PAH indicates pulmonary arterial hypertension.

*Five patients had no data on operability.

Greek Pulmonary Hypertension Registry

Interventional therapies in CTEPH



European Heart Journal
doi:10.1093/eurheartj/ehv317

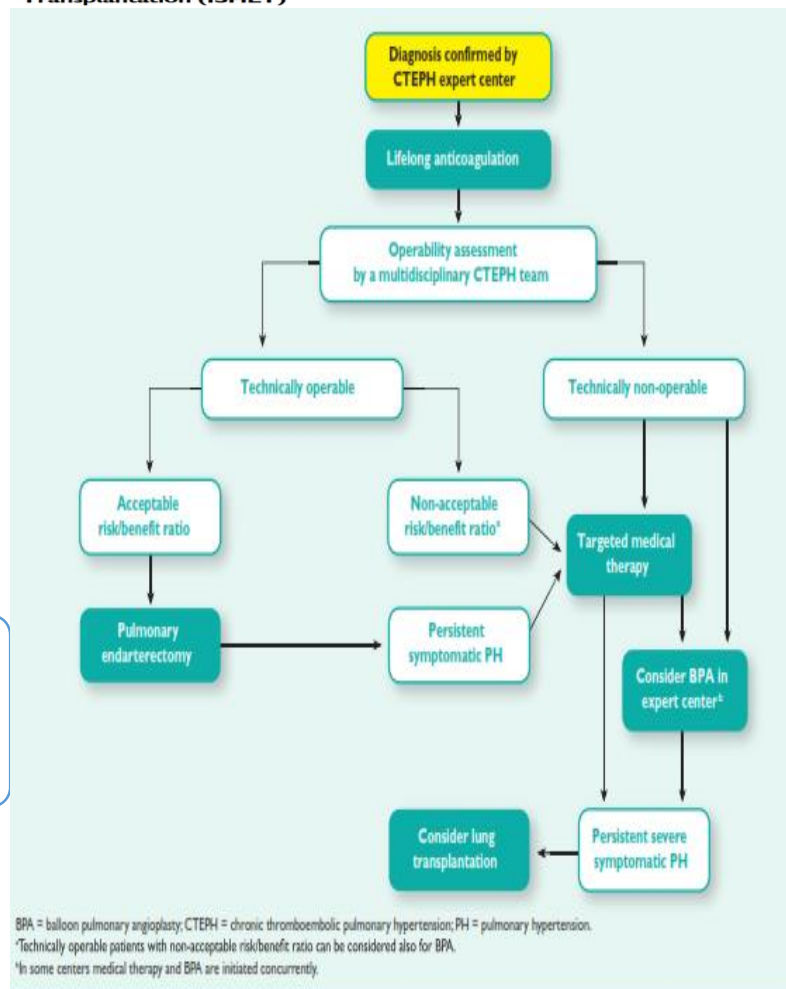
ESC/ERS GUIDELINES



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)



Greek CTEPH Registry

Key messages

- ✓ 69 consecutive patients were included
- ✓ Dyspnea and fatigue were the most common presenting symptoms
- ✓ History of PE was present in 55% of patients
- ✓ Half of patients were on WHO class II, while 40% were in WHO class III
- ✓ Nearly all patients underwent RHC to confirm diagnosis
- ✓ V/Q scintigraphy was performed in 2/3 with 70.5% having abnormal lesions
- ✓ CTPA was performed in 1/5 of patients
- ✓ About 80% of patients received an oral anticoagulant
- ✓ 45.3% of patients screened, were considered operable, of which 75% PEA, while 3 patients underwent BPA
- ✓ Inoperable patients and those that did not undergo PEA received medical treatment
- ✓ We emphasize the need for establishing a designated PEA center in Greece
- ✓ In highly selective CTEPH patients PBA emerges as an effective alternative treatment option.