Conclusion: Our study population pts suffering of severe IPAH presented with cardiac autonomic system impairment, demonstrated mainly as abnormal TS in HRT. Interestingly a significant positive correlation was revealed between TS and physical capacity expressed as maximum oxygen consumption in CPET.

P6340
A comprehensive risk assessment at early follow-up determines prognosis better than at diagnosis in pulmonary arterial hypertension

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Background: Risk stratification and treatment strategies still continue to evolve in pulmonary arterial hypertension (PAH). The aims of the present study were to validate the PAH risk assessment tool presented in the recent guidelines, and to test the benefit of reaching a low-risk profile at early follow-up.
Methods: Between January 2008 and February 2018, a total of 97 incident cases were enrolled from 3 PAH centres (mean age 52±16, female/male 85% / 15%) and followed for a median duration of 24.03 months. Thirty (31%) patients had died. Each patient's risk group was defined by a calculated risk score at baseline and at the time of follow-up within 1 year of diagnosis, using the following variables: World Health Organization functional class, 6-min walking distance (6 MWD), NT-proBNP levels, RA area, pericardial effusion (PE) by echocardiography, and right atrial pressure, cardiac index, and mixed venous oxygen saturation by right heart catheterization. Patients were classified as "Low", "Intermediate", or "High risk".
Results: At baseline, functional class (p=0,01; HR=1,95), 6 MWD (p=0,027; HR=1,82), PE by ECHO (p=0,023; HR=1,70), and mean risk score (p=0,05; HR=1,926) were associated with survival. At follow-up, functional class (p=0.0001; HR=3,65), 6 MWD (p=0.0001; HR=5,63), NT-proBNP levels (p=0,033; HR=2,32), PE by ECHO (p=0,001; HR=2,26), RA area (p=0,008; HR=5,02), and mean risk score (p=0.0001; HR=5,22) were associated with survival. Both mean risk scores, at baseline (p=0,034; HR=2,196; %95 C.I=1,06 - 4,548) and follow-up (p=0.0001; HR=6,098; %95 C.I=2,993 - 12,421), were associated with an increased mortality in multivariable analysis adjusted for age, sex, hypertension, diabetes, and coronary artery disease (table 1). Survival differed between the risk groups at early follow-up (figure 1).

Table 1. Multivariable Cox regression analysis of mean risk scores assessed at baseline and early follow-up

	Sig.	HR		Sig.	HR
Mean risk score at baseline	0,034	2,196	Mean risk score at follow up	0.0001	6,098
Hypertension	0,697	1,21	Hypertension	0,636	1,27
Diabetes	0,619	0,758	Diabetes	0,242	0,507
Coronary artery disease	0,478	0,605	Coronary artery disease	0,549	0,663
Sex	0,783	1,178	Sex	0,555	1,445
Age	0,001	1,065	Age	0.0001	1,076

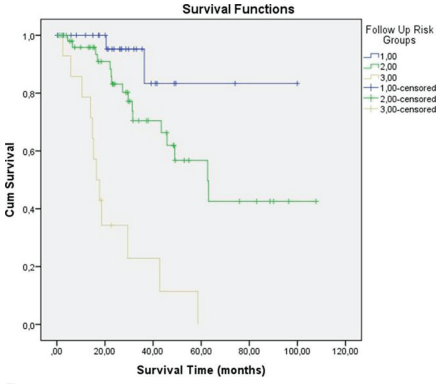

Figure 1

Conclusions: Our findings suggest that the risk assessment tool predicts survival better at early follow-up than at baseline and the aim of reaching a low-risk profile at early follow-up is valid in PAH.

P6341
Haemodynamic and exercise effects of different types of initial oral combination therapy in pulmonary arterial hypertension

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Background: Pulmonary arterial hypertension (PAH) is a severe disease for which different therapeutic strategies are available.
Objectives: The aim of the study is to compare functional and haemodynamic changes after 3–6 months of first line combination therapy or monotherapy with different endothelin receptor antagonists (ERA) and phosphodiesterase 5 inhibitors (PDE5-I) in a single centre.
Methods: Thirty consecutive naïve patients with World Health Organization (WHO) functional class II-III PAH were randomized to recieve monotherapy (Ambrisentan or Tadalafil) or first line combination therapy (Ambrisentan+Tadalafil), while we retrospectively analyzed the data of 153 patients with PAH in WHO functional class I-III who received monotherapy (ERA or PDE5-I) or first line combination therapy (Bosentan+Sildenafil or Macitentan+Sildenafil). At baseline and after 3–6 months of treatment, 6-minute walk distance (6MWD) and right-heart catheterization were performed. Statistical analysis was performed using Student's t-test for the analysis of baseline data versus 3–6 months within each group and for the comparison of the absolute changes between the groups.
Results: Results are shown in the table.

Absolute change (±SD)	Monotherapy Ambrisentan or Tadalafil n= 11	First line combination Ambrisentan + Tadalafil n= 19	p-value		Monotherapy Ambrisentan n= 6	Monotherapy Tadalafil n= 5	p-value
RAP (mmHg)	0 ± 4	-3 ± 5	0.113	RAP (mmHg)	-1 ± 4	+1 ± 3	0.426
mPAP (mmHg)	-11 ± 10	-18 ± 9	0.042	mPAP (mmHg)	-14 ± 11	-7 ± 9	0.314
CI (l/min/m²)	+0.6 ± 0.5	+1.3 ± 0.7	0.004	CI (l/min/m²)	+0.6 ± 0.6	+0.6 ± 0.1	0.962
PVR (WU)	-4 ± 1	-7 ± 3	<0.001	PVR (WU)	-4 ± 1	-4 ± 2	0.899
MVO_2 (%)	+4 ± 6	+10 ± 6	0.005	MVO_2 (%)	+1 ± 5	+7 ± 4	0.051
6MWD (m)	+33 ± 78	+89 ± 52	0.026	6MWD (m)	+18 ± 95	+51 ± 56	0.515
	Monotherapy Bosentan or Sildenafil n= 44	First line combination Bosentan + Sildenafil n= 20	p-value		Monotherapy Bosentan n= 23	Monotherapy Sildenafil n= 21	p-value
RAP (mmHg)	-1 ± 6	-6 ± 6	0.002	RAP (mmHg)	0 ± 7	-1 ± 5	0.420
mPAP (mmHg)	-5 ± 9	-13 ± 11	0.002	mPAP (mmHg)	-3 ± 7	-7 ± 9	0.080
CI (l/min/m²)	+0.4 ± 0.5	+1.3 ± 0.9	<0.001	CI (l/min/m²)	+0.2 ± 0.4	+0.5 ± 0.6	0.055
PVR (WU)	-3 ± 4	-11 ± 9	<0.001	PVR (WU)	-2 ± 4	-4 ± 3	0.181
MVO_2 (%)	+5 ± 8	+14 ± 12	0.004	MVO_2 (%)	+2 ± 8	+8 ± 8	0.020
6MWD (m)	+35 ± 77	+113 ± 145	0.029	6MWD (m)	+15 ± 80	+55 ± 70	0.087
	Monotherapy ERA or PDE5-I n= 56	First line combination Macitentan + Sildenafil n= 28	p-value		Monotherapy ERA n= 28	Monotherapy PDE5-I n= 28	p-value
RAP (mmHg)	-1 ± 5	-2 ± 5	0.087	RAP (mmHg)	-1 ± 5	-1 ± 5	0.935
mPAP (mmHg)	-5 ± 8	-14 ± 11	<0.001	mPAP (mmHg)	-5 ± 7	-6 ± 8	0.656
CI (l/min/m²)	+0.4 ± 0.6	+1.0 ± 0.6	<0.001	CI (l/min/m²)	+0.5 ± 0.6	+0.4 ± 0.6	0.565
PVR (WU)	-3 ± 4	-7 ± 5	<0.001	PVR (WU)	-3 ± 4	-4 ± 4	0.498
MVO_2 (%)	+6 ± 7	+8 ± 7	0.155	MVO_2 (%)	+3 ± 7	+8 ± 8	0.041
6MWD (m)	+41 ± 97	+45 ± 45	0.831	6MWD (m)	+46 ± 88	+37 ± 106	0.738

Legend: CI = cardiac index, mPAP = mean pulmonary arterial pressure, MVO_2 = mixed venous oxygen saturation, PVR = pulmonary vascular resistance, RAP = right atrial pressure, 6MWD = 6 minute walking distance.

Conclusions: Monotherapy and first line combination therapy both improve hemodynamic profile and exercise capacity in patients with PAH; however, first line combination therapy is associated with larger improvements as compared with monotherapy. Monotherapy with ERAs and PDE5-Is showed similar results. Also combination therapy with Bosentan+Sildenafil, Ambrisentan+Tadalafil and Macitentan+Sildenafil had similar efficacy.

P6342
Risk assessment according to the 2015 ESC guidelines risk prediction model of patients with chronic thromboembolic pulmonary hypertension (CTEPH)

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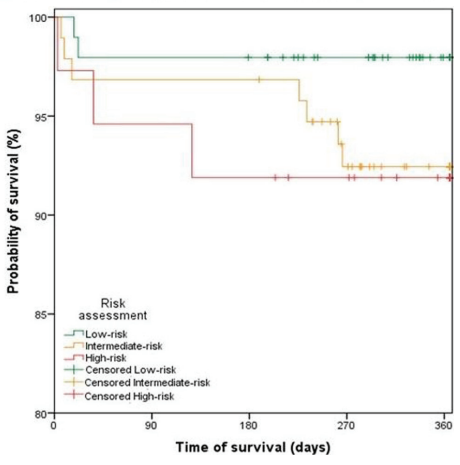
Introduction: Pulmonary endarterectomy (PEA) is the treatment of choice for patients with chronic thromboembolic pulmonary hypertension (CTEPH). However, data on the long-term course after PEA are limited and strategies for risk assessment have not been investigated in CTEPH patients thus far.
Purpose: The aim of the present study was to investigate whether the ESC 2015 guidelines risk prediction model developed for PAH patients allows risk stratification of CTEPH patients after PEA.
Methods: CTEPH patients treated with PEA in an experienced German centre between January 2014 and December 2015 were included in the present study. The ESC 2015 guidelines risk prediction model was used to classify patients into

Figure 1. Probability of 1-year survival after PEA according risk classes



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