

**Supplementary materials Table S1.** Analysis of cytokine concentration in patients with CTEPH, CHD, CTD, IPAH in relation to healthy individuals.

Type of PAH		Cytokine concentration [pg/ml]				
		IL-2	IL-4	IL-6	IL-10	IFN- $\gamma$
CTEPH	Mean $\pm$ SD	8.14 $\pm$ 0.44	2.87 $\pm$ 0.19	23.86 $\pm$ 6.47	6.93 $\pm$ 0.71	5.34 $\pm$ 0.18
	Median (range)	7.89 (7.74-8.82)	2.86 (2.61-3.21)	22.16 (11.12-31.23)	7.07 (5.93-7.99)	5.35 (5.06-5.57)
CHD	Mean $\pm$ SD	29.16 $\pm$ 9.67	6.09 $\pm$ 2.81	18.96 $\pm$ 8.51	19.84 $\pm$ 5.61	4.37 $\pm$ 2.06
	Median (range)	26.66 (16.30-50.12)	5.89 (1.67-12.24)	17.90 (9.35-30.46)	14.65 (3.95-35.30)	4.26 (0.40-10.03)
CTD	Mean $\pm$ SD	5.82 $\pm$ 1.03	5.89 $\pm$ 2.56	33.10 $\pm$ 13.08	11.00 $\pm$ 1.56	4.88 $\pm$ 0.13
	Median (range)	6.09 (4.30-6.91)	4.69 (3.23-14.33)	30.97 (10.57-40.88)	10.68 (8.76-13.66)	4.93 (4.63-5.05)
IPAH	Mean $\pm$ SD	32.19 $\pm$ 8.32	2.57 $\pm$ 0.67	46.66 $\pm$ 10.47	5.72 $\pm$ 3.78	9.20 $\pm$ 3.26
	Median (range)	23.80 (9.39-50.12)	1.74 (0.64-9.24)	45.96 (12.30-60.30)	4.91 (2.07-15.71)	8.04 (5.58-23.30)
Healthy volunteers	Mean $\pm$ SD	2.76 $\pm$ 1.00	4.80 $\pm$ 0.31	4.13 $\pm$ 2.11	4.06 $\pm$ 1.02	2.38 $\pm$ 1.19
	Median (range)	2.36 (0.48-7.16)	4.67 (4.29-5.31)	3.17 (0.15-17.20)	4.08 (2.77-6.16)	2.34 (0.61-4.38)
<i>p-value</i>		0.000*	0.000*	0.000*	0.000*	0.000*
<i>p-value</i>	HV vs. CTEPH	0.000*	0.000*	0.000*	0.000*	0.072*
	CTEPH vs. CHD	0.000*	0.000*	0.031*	0.000*	0.124
	CTEPH vs. CTD	0.032*	0.000*	0.028*	0.017*	0.277
	CTEPH vs. IPAH	0.000*	0.000*	0.000*	0.000*	0.000*
	HV vs. CHD	0.000*	0.027*	0.000*	0.000*	0.000*
	CHD vs. CTD	0.000*	0.048*	0.000*	0.011*	0.312
	CHD vs. IPAH	0.043*	0.000*	0.000*	0.000*	0.000*
	HV vs. CTD	0.000*	0.973	0.000*	0.000*	0.000*
	CTD vs. IPAH	0.000*	0.000*	0.013*	0.000*	0.000*
	HV vs. IPAH	0.000*	0.000*	0.000*	0.142	0.000*

Statistically significant results are marked with \*.

**Supplementary materials Table S2.** Sperman rank correlation analysis for patients with CHD

Parameters	R	t(N-2)	p-value
Wiek & Lym [10 <sup>3</sup> /mm <sup>3</sup> ]	-0 .647	-4 .156	0 .000
Wiek & RVSP [mmHg]	-0 .644	-4 .124	0 .000
Wiek & HGB [g/dl]	-0 .604	-3 .710	0 .001
Wiek & PASP [mmHg]	-0 .572	-3 .415	0 .002
Wiek & mPAP [mmHg]	-0 .565	-3 .353	0 .003
HGB [g/dl] & PLT [10 <sup>3</sup> /mm <sup>3</sup> ]	-0 .463	-2 .561	0 .017
Wiek & PVR dyne/s/cm-5]	-0 .456	-2 .509	0 .019
6MWT [m] & CO [L/min.]	-0 .428	-2 .269	0 .033
NT-proBNP [pg/ml] & HGB [g/dl]	-0 .419	-2 .263	0 .033
BMI & 6MWT [m]	-0 .413	-2 .178	0 .040
IL-2 [pg/ml] & NK	-0 .413	-2 .224	0 .036
CI [L/min/m2] & Mean pressure in the right ventricle	-0 .412	-2 .217	0 .036

[mmHg]			
Wiek & 6MWT [m]	-0.397	-2.072	0.050
PVR dyne/s/cm-5] & HGB [g/dl]	0.400	2.140	0.043
6MWT [m] & mPAP [mmHg]	0.414	2.179	0.040
CD45 & CD4	0.454	2.499	0.020
CD4 & CD3	0.454	2.499	0.020
6MWT [m] & HGB [g/dl]	0.457	2.464	0.022
PLT [ $10^3/\text{mm}^3$ ] & IFN- $\gamma$ [pg/ml]	0.462	2.551	0.018
PVR dyne/s/cm-5] & Lym [ $10^3/\text{mm}^3$ ]	0.469	2.600	0.016
6MWT [m] & PASP [mmHg]	0.469	2.546	0.018
CO [L/min.] & IL-10 [pg/ml]	0.471	2.615	0.015
PLT [ $10^3/\text{mm}^3$ ] & Wiek	0.492	2.768	0.011
PVR dyne/s/cm-5] & 6MWT [m]	0.497	2.745	0.012
CD19 & CD4/CD8	0.516	2.954	0.007
HGB [g/dl] & IL-2 [pg/ml]	0.518	2.964	0.007
6MWT [m] & RVSP [mmHg]	0.533	3.025	0.006
NT-proBNP [pg/ml] & IL-4 [pg/ml]	0.535	3.103	0.005
NT-proBNP [pg/ml] & IL-10 [pg/ml]	0.540	3.142	0.004
PASP [mmHg] & Lym [ $10^3/\text{mm}^3$ ]	0.612	3.789	0.001
PVR dyne/s/cm-5] & RVSP [mmHg]	0.642	4.107	0.000
RVSP [mmHg] & Lym [ $10^3/\text{mm}^3$ ]	0.643	4.112	0.000
Lym [ $10^3/\text{mm}^3$ ] & HGB [g/dl]	0.654	4.230	0.000
PVR dyne/s/cm-5] & PASP [mmHg]	0.660	4.307	0.000
PASP [mmHg] & HGB [g/dl]	0.670	4.425	0.000
mPAP [mmHg] & HGB [g/dl]	0.672	4.448	0.000
RVSP [mmHg] & HGB [g/dl]	0.719	5.068	0.000
CD4 & CD4/CD8	0.740	5.393	0.000
mPAP [mmHg] & RVSP [mmHg]	0.744	5.454	0.000
mPAP [mmHg] & PASP [mmHg]	0.763	5.780	0.000
mPAP [mmHg] & Lym [ $10^3/\text{mm}^3$ ]	0.790	6.318	0.000
PVR dyne/s/cm-5] & mPAP [mmHg]	0.793	6.369	0.000
CD45 & CD8	0.826	7.188	0.000
CD8 & CD3	0.826	7.188	0.000
CI [L/min/m <sup>2</sup> ] & CO [L/min.]	0.890	9.586	0.000
PASP [mmHg] & RVSP [mmHg]	0.961	16.962	0.000

Supplementary materials Table S3 Sperman rank correlation analysis for patients with CTD

Parameters	R	t(N-2)	p-value
Wiek & PLT [ $10^3/\text{mm}^3$ ]	-0.916	-6.040	0.001
IL-2 [pg/ml] & IL-4 [pg/ml]	-0.833	-3.989	0.005
IL-4 [pg/ml] & IFN- $\gamma$ [pg/ml]	-0.833	-3.989	0.005
IL-2 [pg/ml] & IL-4 [pg/ml]	-0.833	-3.989	0.005
IL-4 [pg/ml] & IFN- $\gamma$ [pg/ml]	-0.833	-3.989	0.005
CD19 & NK	-0.733	-2.854	0.025
Wiek & mPAP [mmHg]	-0.713	-2.687	0.031

PVR dyne/s/cm-5] & RVSP [mmHg]	0 .672	2 .404	0 .047
mPAP [mmHg] & PLT [10 <sup>3</sup> /mm <sup>3</sup> ]	0 .678	2 .442	0 .045
CD8 & CD19	0 .683	2 .476	0 .042
mPAP [mmHg] & RVSP [mmHg]	0 .694	2 .553	0 .038
CD45 & CD19	0 .700	2 .593	0 .036
CD19 & CD3	0 .700	2 .593	0 .036
PVR dyne/s/cm-5] & IL-6 [pg/ml]	0 .717	2 .719	0 .030
IL-4 [pg/ml] & CD8	0 .750	3 .000	0 .020
CD4 & CD4/CD8	0 .750	3 .000	0 .020
CD4 & CD19	0 .750	3 .000	0 .020
mPAP [mmHg] & IL-6 [pg/ml]	0 .804	3 .573	0 .009
IL-4 [pg/ml] & IL-10 [pg/ml]	0 .833	3 .989	0 .005
IL-4 [pg/ml] & CD19	0 .850	4 .269	0 .004
CD45 & CD4	0 .850	4 .269	0 .004
CD4 & CD3	0 .850	4 .269	0 .004
PASP [mmHg] & IL-4 [pg/ml]	0 .863	4 .511	0 .003
PVR dyne/s/cm-5] & mPAP [mmHg]	0 .975	11 .533	0 .000
CI [L/min/m <sup>2</sup> ] & CO [L/min.]	0 .983	14 .248	0 .000

Supplementary materials Table S4 Sperman rank correlation analysis for patients with CTEPH

Parameters	R	t(N-2)	p-value
CD8 & CD4/CD8	-0 .964	-10 .200	0 .000
CD4 & CD8	-0 .903	-5 .946	0 .000
mPAP [mmHg] & Lym [10 <sup>3</sup> /mm <sup>3</sup> ]	-0 .874	-4 .758	0 .002
IL-10 [pg/ml] & Lym [10 <sup>3</sup> /mm <sup>3</sup> ]	-0 .729	-3 .017	0 .017
Lym [10 <sup>3</sup> /mm <sup>3</sup> ] & PASP [mmHg]	-0 .728	-2 .810	0 .026
IL-2 [pg/ml] & mPAP [mmHg]	-0 .669	-2 .384	0 .049
IL-4 [pg/ml] & mPAP [mmHg]	-0 .669	-2 .384	0 .049
IL-10 [pg/ml] & Mean pressure in the right ventricle [mmHg]	-0 .669	-2 .384	0 .049
CD45 & NK	0 .636	2 .333	0 .048
NK & CD3	0 .636	2 .333	0 .048
IL-6 [pg/ml] & NK	0 .661	2 .489	0 .038
CD45 & CD8	0 .661	2 .489	0 .038
CD8 & CD3	0 .661	2 .489	0 .038
NK & IL-6 [pg/ml]	0 .661	2 .489	0 .038
IL-6 [pg/ml] & PVR dyne/s/cm-5]	0 .667	2 .366	0 .050
IL-2 [pg/ml] & Mean pressure in the right ventricle [mmHg]	0 .669	2 .384	0 .049
IL-4 [pg/ml] & Mean pressure in the right ventricle [mmHg]	0 .669	2 .384	0 .049
IL-10 [pg/ml] & mPAP [mmHg]	0 .669	2 .384	0 .049
BMI & CO [L/min.]	0 .683	2 .476	0 .042

IL-6 [pg/ml] & CD45	0 .697	2 .749	0 .025
CD3 & IL-6 [pg/ml]	0 .697	2 .749	0 .025
IL-2 [pg/ml] & Lym [10 <sup>3</sup> /mm <sup>3</sup> ]	0 .729	3 .017	0 .017
IL-4 [pg/ml] & Lym [10 <sup>3</sup> /mm <sup>3</sup> ]	0 .729	3 .017	0 .017
IFN- $\gamma$ [pg/ml] & BMI	0 .791	3 .663	0 .006
CI [L/min/m <sup>2</sup> ] & CO [L/min.]	0 .867	4 .596	0 .002
mPAP [mmHg] & RVSP [mmHg]	0 .870	4 .675	0 .002
CD4 & CD4/CD8	0 .903	5 .946	0 .000
mPAP [mmHg] & PASP [mmHg]	0 .929	6 .635	0 .000
PASP [mmHg] & RVSP [mmHg]	0 .967	9 .989	0 .000

Supplementary materials Table S5 Sperman rank correlation analysis for patients with IPAH

Parameters	R	t(N-2)	p-value
PVR dyne/s/cm-5] & CO [L/min.]	-0 .649	-4 .096	0 .000
PVR dyne/s/cm-5] & CI [L/min/m <sup>2</sup> ]	-0 .592	-3 .522	0 .002
Wiek & 6MWT [m]	-0 .584	-3 .452	0 .002
CI [L/min/m <sup>2</sup> ] & IL-10 [pg/ml]	-0 .489	-2 .685	0 .013
IL-4 [pg/ml] & IFN- $\gamma$ [pg/ml]	-0 .485	-2 .657	0 .014
IL-4 [pg/ml] & IFN- $\gamma$ [pg/ml]	-0 .485	-2 .657	0 .014
Wiek & IL-2 [pg/ml]	-0 .469	-2 .549	0 .018
IL-6 [pg/ml] & CD19	-0 .466	-2 .527	0 .019
Mean pressure in the right ventricle [mmHg] & IL-2 [pg/ml]	-0 .462	-2 .498	0 .020
CO [L/min.] & IL-2 [pg/ml]	-0 .432	-2 .295	0 .031
CO [L/min.] & IL-10 [pg/ml]	-0 .429	-2 .280	0 .032
Wiek & mPAP [mmHg]	-0 .426	-2 .260	0 .034
Wiek & PASP [mmHg]	-0 .416	-2 .193	0 .039
NT-proBNP [pg/ml] & IL-10 [pg/ml]	-0 .402	-2 .106	0 .046
PVR dyne/s/cm-5] & IL-6 [pg/ml]	0 .406	2 .131	0 .044
CD45 & CD4/CD8	0 .435	2 .319	0 .030
CD3 & CD4/CD8	0 .435	2 .319	0 .030
CO [L/min.] & IFN- $\gamma$ [pg/ml]	0 .438	2 .338	0 .028
PVR dyne/s/cm-5] & IL-2 [pg/ml]	0 .444	2 .375	0 .026
IL-10 [pg/ml] & CD4/CD8	0 .444	2 .375	0 .026
CD8 & CD19	0 .465	2 .516	0 .019
Mean pressure in the right ventricle [mmHg] & mPAP [mmHg]	0 .470	2 .555	0 .018
IL-10 [pg/ml] & CD4	0 .475	2 .591	0 .016
CD4 & CD8	0 .477	2 .602	0 .016
6MWT [m] & RVSP [mmHg]	0 .518	2 .901	0 .008
mPAP [mmHg] & IL-6 [pg/ml]	0 .539	3 .066	0 .005
6MWT [m] & PASP [mmHg]	0 .568	3 .309	0 .003
CI [L/min/m <sup>2</sup> ] & IFN- $\gamma$ [pg/ml]	0 .581	3 .423	0 .002
CD45 & CD19	0 .613	3 .722	0 .001
CD19 & CD3	0 .613	3 .722	0 .001
CD4/CD8 & CD19	0 .693	4 .611	0 .000

CD4 & CD19	0 .703	4 .742	0 .000
PVR dyne/s/cm-5] & mPAP [mmHg]	0 .724	5 .036	0 .000
CD45 & CD4	0 .812	6 .680	0 .000
CD3 & CD4	0 .812	6 .680	0 .000
CD4 & CD4/CD8	0 .816	6 .774	0 .000
CD45 & CD8	0 .838	7 .379	0 .000
CD3 & CD8	0 .838	7 .379	0 .000
CI [L/min/m2] & CO [L/min.]	0 .869	8 .432	0 .000
PASP [mmHg] & RVSP [mmHg]	0 .982	25 .057	0 .000