Online supplement table 1: Clinical features and pulmonary hemodynamics at baseline (T0) and after 5 months (T1) under PAH-drug therapies in 37 patients\*

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| --- | --- | --- | --- |
| Variables | T0 | T1 | *p* value# |
| Clinical features  NYHA class (n=)  I or II  III  IV  6MWD  Resting hemodynamics | 7  28  2  392 ± 112 | 22  13  2  420 ± 123 | -  0,05 |
| HR (beats/min) | 75 ± 14 | 70 ± 13 | 0.009 |
| mPAP (mm Hg) | 45 ± 9 | 40 ± 10 | <0.001 |
| CI (L/min/m²) | 2.5 ± 0.6 | 2.8 ± 0.7 | 0.001 |
| SVI (ml/m2) | 33.7 ± 8.9 | 40.7 ± 9.1 | <0.001 |
| PVR (Wood Units) | 9.4 ± 4.1 | 7.0 ± 3.1 | <0.001 |
| SvO2 (%) | 63 ± 8 | 65 ± 6 | 0.110 |
| Exercise hemodynamics |  |  |  |
| Workload (W) | 20 ± 10 | 21 ± 11 | 0.068 |
| HR (beats/min) | 108 ± 17 | 101 ± 17 | <0.001 |
| mPAP (mm Hg) | 64 ± 12 | 60 ± 12 | 0.002 |
| CI (L/min/m²) | 3.5 ± 1.0 | 4.2 ± 1.1 | <0.001 |
| SVI (ml/m2) | 32.8 ± 9.9 | 41.8 ± 10.3 | <0.001 |
| PVR (Wood Units) | 9.5 ± 4.1 | 7.2 ± 2.8 | <0.001 |
| Exercise minus rest values |  |  |  |
| ΔHR (beats/min) | 33 ± 13 | 32 ± 13 | 0.388 |
| ΔsPAP (mm Hg) | 27 ± 13 | 29 ± 11 | 0.474 |
| ΔCI (L/min/m²) | 1.0 ± 0.7 | 1.4 ± 0.8 | <0.001 |
| ΔSVI (ml/m2) | -0.9 ± 5.2 | 1.1 ± 5.3 | 0.012 |
| Pressure-Flow slope (mm Hg/l/min) | 14.1 ± 8.6 | 11.0 ± 6.6 | 0.020 |
| **Isoflow mPAP (mm Hg)** | **62 ± 13** | **49 ± 15** | **<0.001** |

Abbreviations: 6MWD= 6-minute walk distance; HR= heart rate; mPAP= mean pulmonary artery pressure; CI= cardiac index; SVI= indexed stroke volume; PVR= pulmonary vascular resistance; SvO2=mixed venous oxygen saturation, Δ= exercise minus rest values; sPAP= systolic pulmonary artery pressure; Pressure-Flow=pressure-flow relationships

\*Idiopathic PAH (n=14), heritable PAH (n=3), PAH associated with the scleroderma spectrum (n=8), PAH associated with portal hypertension (n=5), PAH associated with systemic lupus erythematosus (n=3), PAH associated with human immunodeficiency virus (n=2), PAH associated with congenital heart disease (n=2)

#*p* values of the paired *t* tests