**Supplementary Table S2 Comparison of Hemodynamic indices based on PAH associated with congenital heart disease**

|  |  |  |  |
| --- | --- | --- | --- |
| **Variables** | **PAH associated with congenital heart disease** **(n=35)** | **PAH not associated with congenital heart disease****(n=11)** | **p-value** |
| **Directly measured Variables** |
| VO2 (measured/ LaFarge) | 150 [148–160] | 150 [149–156] | 0.97 |
| Heart rate | 120 [102–134] | 110 [105–122] | 0.46 |
| Mean RAP (mmHg) | 7 [6–10] | 8 [6–10] | 0.87 |
| PASP (mmHg) | 50 [42–70] | 60 [39–66] | 0.67 |
| PADP (mmHg) | 23 [18–36] | 24 [18–36] | 0.80 |
| Mean PAP (mmHg) | 36 [29–51] | 42 [28–48] | 0.62 |
| Mean PCWP (mmHg) | 11 [9–11] | 10 [9–10] | 0.32 |
| Mean SAP | 56 [52–67] | 62 [57–67] | 0.33 |
| **Calculated variables** |
| TPG (mmHg) | 26 [19–40] | 33 [19–39] | 0.87 |
| PVRi (Wood units/m2) | 4.6 [3.4–9.3] | 8.3 [6.8–13.3] | 0.07 |
| Diastolic pulmonary gradient | 13 [8–26] | 14 [8–27] | 0.66 |
| PA elastance indexed (mmHg/ml/m2) | 1.2 [1.0–1.7] | 2.1 [1.5–2.6] | 0.03\* |
| Qpi (L/min/m2) | 4.7 [3.7–6.3] | 3.0 [2.4–3.5] | 0.004\* |
| Qsi (L/min/m2) | 3.9 [3.1–4.6] | 3.0 [2.6–3.5] | 0.09 |
| PA pulse pressure (mmHg) | 30 [26–36] | 27 [18–34] | 0.30 |
| PSV indexed (ml/m2) | 42 [29–52] | 27 [23–31] | 0.005\* |
| PA compliance indexed (ml/mmHg) | 1.5 [1.1–1.9] | 1.0 [0.8–1.4] | 0.05\* |
| RVSWI (g/m2 or mmHg×mL/m2) | 1042 [807–2304] | 888 [577–932] | 0.08 |
| PAPi (no units) | 4.0 [3.0–4.6] | 4.1 [2.2–5.0] | 0.88 |

mPAP - mean pulmonary arterial pressure, PA - pulmonary artery, PASP - pulmonary arterial systolic pressure, PADP - pulmonary arterial diastolic pressure, PAPi - Pulmonary artery pulsatility index, PCWP – pulmonary capillary wedge pressure, PSVi - pulmonary stroke volume index, Qpi - pulmonary flow index, Qsi - systemic flow index, RAP – right atrial pressure, RVSWI - Right ventricular stroke work index, SAP – systemic arterial pressure, \*p < 0.05 is significant