**Table S3** Clinical Characteristics of the Recruited Subjects

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| --- | --- | --- | --- |
|  | PH (n=6) | controls (n=6) | *p* value |
| Age (years) | 4.62(0.31-14.37) | 2.20(0.91-11.60) | 0.631 |
| Male/female | 5/1 | 4/2 | / |
| **Echocardiography** | | | |
| LVDd (mm) | 3.35(2-4.37) | 2.85(2.35-4.10) | 0.831 |
| LVDs (mm) | 2.15(1.25-2.72) | 1.75(1.55-2.55) | 0.915 |
| LVEF (%) | 66(55.50-69.25) | 67(61.50-69.50) | 0.745 |
| LVFS (%) | 35(28.75-38) | 36(32.25-37.50) | 0.827 |
| MPA (mm) | 0.70(0.65-1.15) | 1.20(1.05-1.20) | 0.133 |
| TRPG (mmHg) | 21(19.75-22.75) | 52.50(39-90) | 0.004 |
| TRV (m/s) | 2.29(2.22-2.38) | 3.60(3.12-4.73) | 0.004 |
| **Blood Tandem Mass Spectrometry** | | | |
| Arginine (μM) | 25.56(12.45-34.03) | 13.31(4.59-21.84) | 0.200 |
| Glutamine (mM) | 560.90(458.45-608.55) | 400.31(381.06-454.05) | 0.016 |
| Caproyl carnitine (μM) | 0.08(0.06-0.08) | 0.03(0.02-0.05) | 0.005 |
| Octenyl carnitine (mM) | 0.24(0.15-0.42) | 0.185(0.14-0.28) | 0.423 |
| Palmityl carnitine (mM) | 1.34(0.73-1.74) | 0.98(0.72-1.36) | 0.423 |

Data are presented as median (interquartile ranges, IQRs). Differences were assessed by non-parametric statistical method. *p* value <0.05 was considered statistically significant. LVDd, left ventricular end diastolic dimension; LVDs, left ventricular end systolic dimension; LVEF% left ventricular ejection fraction; LVFS%, left ventricular fractional shortening; MPA, main pulmonary artery; TRPG, tricuspid regurgitation pressure gradient; TRV, tricuspid regurgitation velocity.