

Table S1. Clinical, echocardiographic, right heart catheterisation features, and outcome of the idiopathic pulmonary hypertension patients who have been studied

| Patient | Gender | Age at first diagnosis | NYHA functional class at diagnosis | Age at last follow-up | Saturation on room air | Therapy | Echo TAPSE | Echo RVSP | RHC mPAP | RHC mRAP | RHC CI | RHC PVRi | RHC SVO2 | Outcome |
|---------|--------|------------------------|------------------------------------|-----------------------|------------------------|--|------------|-----------|----------|----------|--------|----------|----------|--|
| 1 | F | 1 | II | 1 | 87 | Sildenafil, Bosentan | 9 | 90 | 62 | 10 | 2 | 22 | 60 | On transplant list |
| 2 | M | 6 | II | 16 | 93 | Sildenafil, Bosentan, Treprostinil | 12 | 85 | 107 | 17 | 3.6 | 18.8 | 69 | On transplant list |
| 3 | F | 4 | II | 13 | 80 | Sildenafil, Ambrisentan, Treprostinil | 13 | 130 | 80 | 13 | 5.3 | 24 | 66 | On transplant list |
| 4 | F | 11 | IV | 16 | 88 | Sildenafil, Bosentan, Epoprostenol (upfront therapy) | 12 | 90 | 82 | 9 | 2 | 23 | 41 | Much improved with therapy (currently NYHA I). Epoprostenol switched to Treprostinil |
| 5 | F | 5 | II | 9 | 93 | Sildenafil, Bosentan, Epoprostenol | 9 | 100 | 41 | 8 | 5.2 | 8.2 | 84 | Lung transplant |
| 6 | M | 17 | III | 17 | 92 | No therapy. Addressed straight to lungs transplant | 16 | 125 | 57 | 16 | 2.8 | 18.8 | 52 | Death while waiting for lung transplant |
| 7 | M | 11 | III | 13 | 90 | Sildenafil, Bosentan, Iloprost | 11 | 110 | 51 | 10 | 3.7 | 9.3 | 76 | Death while waiting for lung transplant |
| 8 | M | 0.1 | III | 15 | 97 | Sildenafil, Macitentan | 22 | 75 | 48 | 9 | 2.6 | 10.6 | 67 | Small PDA + atrial septostomy + atrial stenting |
| 9 | F | 0.1 | IV | 16 | 93 | Sildenafil, Bosentan, Epoprostenol | 17 | 120 | 50 | 7 | N/A | 20 | 56 | PDA stenting (reverse Potts shunt) |
| 10 | F | 2 | III | 8 | 95 | Sildenafil, Bosentan, Epoprostenol | N/A | 85 | 40 | 8 | N/A | 8.7 | 65 | Transplant assessment |

Legends: M: male; F: female; NYHA: New York Heart Association; TAPSE: tricuspid annular plane systolic excursion; N/A: not acquired; RVSP: right ventricular systolic pressure; RHC: right heart catheterisation; mPAP: mean pulmonary arterial pressure; mRAP: mean right atrial pressure; CI: cardiac index; PVRI: pulmonary vascular resistance; SVO₂: mixed venous oxygen saturation; PDA: patent ductus arteriosus.