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| **Supplementary Table S1** Univariable analysis for predictors of heart transplantation and mortality in patients during the late post-Fontan period | | |
|  | Univariable |  |
|  | HR (95% CI) | *p*-value |
| Basic demographics | | |
| Sex, male | 4.25 (0.89–20.2) | 0.07 |
| Hypoplastic left heart syndrome | 3.97 (1.12–14.1) | **0.03** |
| Right ventricular physiology | 13.1 (1.66–103.4) | **0.02** |
| Heterotaxy | 0.97 (0.12–7.70) | 0.98 |
| Fontan type,  Atrio–pulmonary connection  Lateral tunnnel | 0.33\*10-7 (0-Inf)  0.85 (0.18–4.03) | 1.0  0.8 |
| Fenestration at Fontan | 6.38 (0.79–51.3) | 0.08 |
| Age at Fontan, y | 0.50 (0.19–1.31) | 0.16 |
| Hospital stay after Fontan, days | 1.04 (1.01–1.08) | **<0.01** |
| Glenn hemodynamics | | |
| Pulmonary arterial pressure, mmHg | 0.91 (0.60–1.37) | 0.64 |
| Transpulmonary pressure gradient, mmHg | 0.80 (0.48–1.33) | 0.38 |
| Atrial pressure, mmHg | 1.20 (0.78–1.87) | 0.42 |
| SaO2, % | 0.84 (0.65–1.09) | 0.20 |
| PVRI, WU·m2 | 1.46 (0.48–4.38) | 0.50 |
| PVRI/SVRI\*a | 27.1 (0.70–1048) | 0.08 |
| Clinical variables at evaluation | | |
| Age at evaluation, y | 0.95 (0.84–1.08) | 0.47 |
| Duration after Fontan, y | 0.98 (0.87–1.10) | 0.69 |
| Indication for Cath, surveillance evaluation | 0.60 (0.15–2.33) | 0.46 |
| Body surface area, m2 | 1.17 (0.21–6.49) | 0.86 |
| Body mass index | 0.99 (0.90–1.08) | 0.79 |
| Fenestration, patent | 3.93 (1.11–14.0) | **0.03** |
| Medication  Diuretics  ACEI/ARB  Beta-blocker  Sildenafil | 2.93 (0.82–10.5)  1.56 (0.44–5.52)  2.36 (0.58–9.64)  2.26 (0.45–11.3) | 0.10  0.49  0.23  0.32 |
| NYHA Ⅲ/Ⅳ | 32.8 (4.05–267) | **<0.01** |
| PLE/PB | 2.94 (0.75–11.6) | 0.12 |
| Use of pacemaker | 1.37 (0.29–6.47) | 0.69 |
| Echocardiography  Ventricular dysfunction\*b  AVVR  PG at fenestration, mmHg  CoA, SAS\*c | 5.31 (1.24–22.7)  1.03 (0.22–4.89)  5.41 (0.45–64.9)  0.000000014 (0–Inf) | **0.03**  0.97  0.18  1.0 |
| Hemoglobin, g/dL | 1.09 (0.77–1.54) | 0.63 |
| Blood urea nitrogen, mg/dL | 1.08 (0.98–1.20) | 0.11 |
| Creatinine, mg/dL | 0.78 (0.025–23.8) | 0.88 |
| Mechanical ventilation | 0.47 (0.14–1.65) | 0.24 |
| Hemodynamic variable at evaluation | | |
| Fontan pressure, mmHg | 1.24 (1.05–1.46) | **0.01** |
| Fontan pressure, ≥14 mmHg | 3.68 (0.78–17.5) | 0.1 |
| Fontan pressure, ≥17 mmHg | 5.73 (1.60–20.6) | **<0.01** |
| Cardiac index, L/min/m2 | 1.38 (0.91–2.09) | 0.13 |
| Cardiac index ≥3.0 L/min/m2 | 1.38 (0.91–2.09) | 0.13 |
| Mean SP, mmHg | 0.94 (0.88–1.01) | 0.07 |
| PVRI, WU·m2 | 1.25 (0.57–2.75) | 0.57 |
| PVRI, ≥4.0 WU·m2 | 0.00000030 (0–Inf) | 1.0 |
| SVRI, WU·m2 | 0.89 (0.78–1.01) | 0.08 |
| SVRI, <13 WU·m2 | 4.21 (1.11–16.0) | **0.04** |
| PVRI/SVRI\*b | 2.01 (1.10–3.66) | **0.02** |
| PCWP, mmHg | 1.22 (1.03–1.44) | **0.02** |
| Transpulmonary pressure gradient, mmHg | 1.03 (0.67–1.59) | 0.9 |
| Perfusion pressure, mmHg | 0.91 (0.85–0.98) | **0.01** |
| Perfusion pressure, <53 mmHg | 10.3 (1.29–81.7) | **0.03** |
| Qp/Qs | 0.55 (0.028–10.9) | 0.7 |
| Qp/Qs <0.65 | 6.25 (1.70–22.9) | **<0.01** |
| SaO2, % | 0.87 (0.81–0.93) | **<0.01** |
| SaO2, <90% | 9.31 (2.28–38.1) | **<0.01** |
| SvO2, % | 0.89 (0.84–0.96) | **<0.01** |
| Hemodynamic category | | |
| Category A (CI≥3, SVRI≥13) | 2.82 (0.59–13.5) | 0.19 |
| Category B (CI<3, SVRI≥13) | 0.83 (0.23–2.95) | 0.77 |
| Category C (CI≥3, SVRI<13) | 5.71 (1.51–21.6) | **0.01** |
| Category D (CI<3, SVRI<13) | 3.85 × 10-8 (0–Inf) | 0.99 |
| *ACEI* angiotensin converting enzyme inhibitor, *ARB* angiotensin receptor blocker, *AVVR* atrioventricular valve regurgitation*, Cath* catheterization*, 95% CI* 95% confidence interval*, CoA* coarctation of aorta*, HR* hazard ratio*, NYHA* New York Heart Association functional classification*, PB* plastic bronchitis, *PCWP* pulmonary capillary wedge pressure*, PG* pressure gradient*, PLE* protein losing enteropathy*, PVRI* pulmonary vascular resistance index*, Qp/Qs* pulmonary flow/systemic flow*, SaO2* systemic arterial oxygen saturation*, SP* systemic pressure*, SvO2* systemic venous oxygen saturation*, SVRI* systemic vascular resistance index*.*  \*a Per 0.1  \*b more than moderate  \*c > 30 mmHg | | |