**Supplementary Tables**

Supplementary Table S1

|  |  |  |  |
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| Table S1. Risk factor analysis for onset of failing Fontan | | | |
| Variables |  | Univariate |  |
|  | p-value | HR | 95% CI |
| Sex | 0.177 | 0.706 | 0.426-1.170 |
| HLHS | **0.013** | 1.885 | 1.140-3.117 |
| UVH | 0.810 | 0.936 | 0.544-1.608 |
| TA | 0.069 | 0.486 | 0.223-1.059 |
| DILV | 0.338 | 0.720 | 0.368-1.409 |
| PAIVS | 0.363 | 0.521 | 0.128-2.122 |
| UAVSD | 0.299 | 1.707 | 0.623-4.682 |
| ccTGA | 0.550 | 0.703 | 0.221-2.232 |
| Others | 0.228 | 1.545 | 0.762-3.134 |
| Dominant RV | **<0.01** | 2.597 | 1.587-4.253 |
| TGA | 0.281 | 0.770 | 0.479-1.238 |
| DORV | 0.539 | 1.222 | 0.645-2.318 |
| CoA | 0.277 | 0.666 | 0.320-1.387 |
| Dextrocardia | 0.227 | 1.538 | 0.765-3.092 |
| Heterotaxy | **0.003** | 2.543 | 1.369-4.724 |
| Azygos | 0.102 | 2.132 | 0.860-5.288 |
| T (P)APVC | **0.002** | 2.713 | 1.428-5.151 |
| ASVD | 0.096 | 1.724 | 0.908-3.271 |
| Norwood/DKS | 0.024 | 1.708 | 1.703-2.721 |
| APS | 0.516 | 0.897 | 0.646-1.245 |
| PAB | 0.106 | 1.547 | 0.911-2.629 |
| Previsou BCPS | 0.085 | 2.022 | 0.908-4.503 |
| Age BCPS (months) | 0.718 | 0.998 | 0.990-1.007 |
| Weight BCPS (kg) | 0.406 | 0.975 | 0.918-1.035 |
| Pre TCPC catheter values |  |  |  |
| PAP | **<0.001** | 1.119 | 1.052-1.191 |
| LAP | 0.142 | 1.070 | 0.977-1.172 |
| TPG | **0.028** | 1.118 | 1.012-1.235 |
| SVP | 0.170 | 1.011 | 0.996-1.026 |
| EDP | 0.100 | 1.070 | 0.987-1.159 |
| MAP | 0.146 | 1.013 | 0.996-1.030 |
| SO2 | 0.450 | 0.986 | 0.949-1.023 |
| Age at TCPC | 0.633 | 1.011 | 0.968-1.055 |
| TCPC weight | 0.389 | 1.008 | 0.990-1.025 |
| TCPC height | 0.385 | 1.004 | 0.995-1.013 |
| CPB time | 0.483 | 1.002 | 0.997-1.007 |
| Need AXC | 0.693 | 1.103 | 0.678-1.794 |
| Fenestration | 0.586 | 0.811 | 0.382-1.723 |
| Concomitant procedure | 0.075 | 1.519 | 0.959-2.406 |
| Con DKS | 0.079 | 2.127 | 0.916-4.939 |
| Con AVV procedure | 0.396 | 1.307 | 0.704-2.427 |
| Con PA reconstruction | 0.160 | 1.511 | 0.849-2.691 |
| Con ASE | 0.462 | 1.406 | 0.567-3.485 |
| Con SAS Resection | 0.521 | 0.049 | 0.000-498.637 |
| Con Pacemaker | 0.320 | 1.799 | 0.566-5.720 |
| ICU stay | **<0.01** | 1.034 | 1.019-1.048 |
| Hospital stay | **<0.01** | 1.022 | 1.009-1.035 |
| Chest tube dration | **0.001** | 1.029 | 1.021-1.048 |
| Pleural effusion (< 7 days) | **0.005** | 1.957 | 1.231-3.113 |
| Chylothorax | 0.138 | 1.498 | 0.878-2.556 |
| Ascites | **0.035** | 1.706 | 1.038-2.805 |
| ICU re-admission | 0.181 | 1.425 | 0.848-2.394 |

Supplementary Table S2

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| Table S2. Cardiac catheterizazion data before TCPC | | | |  |  |
| Variables: N(%) or median (IQR) | | Total cases | Non-failing Fontan | Failing Fontan | p-value |
| Number of patients | | 634 | 558 (88.0) | 76 (12.0) |  |
|  | PAP | 9 (8-12) | 9 (8-11) | 10 (9-13) | <0.001 |
|  | LAP | 5 (4-7) | 5 (4-7) | 6 (4-7) | 0.372 |
|  | TPG | 4 (3-5) | 4 (3-5) | 4 (3-6) | 0.003 |
|  | SVP | 83 (76-92) | 83 (76-91) | 83 (77-95) | 0.169 |
|  | EDP | 8 (6-10) | 8 (6-9) | 8 (5-10) | 0.745 |
|  | MAP | 58 (51-65) | 59 (51-64) | 60 (52-68) | 0.088 |
|  | SO2 | 83 (80-86) | 83 (80-86) | 82 (79-87) | 0.929 |

Supplementary Table S3

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| Table S3. Risk factor analysis for onset of failing Fontan | | | |  |  |  |  |
| Variables |  | Univariate |  |  |  | Multivariate |  |
|  | p-value | HR | 95% CI |  | p-value | HR | 95% CI |
| HLHS | **0.013** | 1.885 | 1.140-3.117 |  |  |  |  |
| Dominant RV | **<0.001** | 2.597 | 1.587-4.253 |  | 0.010 | 2.062 | 1.192-3.568 |
| Heterotaxy | **0.003** | 2.543 | 1.369-4.724 |  |  |  |  |
| T(P)APVC | **0.002** | 2.713 | 1.428-5.151 |  |  |  |  |
| Norwood/DKS | **0.024** | 1.708 | 1.703-2.721 |  |  |  |  |
| PAP pre TCPC | **<0.001** | 1.119 | 1.052-1.191 |  | 0.004 | 1.107 | 1.032-3.568 |
| TPG pre TCPC | **0.028** | 1.118 | 1.012-1.235 |  |  |  |  |
| ICU stay (per day) | **<0.001** | 1.034 | 1.019-1.048 |  |  |  |  |
| Hospital stay (per day) | **<0.001** | 1.022 | 1.009-1.035 |  |  |  |  |
| Chest tube dration (per day) | **0.001** | 1.029 | 1.021-1.048 |  |  |  |  |
| Pleural effusion (< 7 days) | **0.005** | 1.957 | 1.231-3.113 |  |  |  |  |
| Ascites | **0.035** | 1.706 | 1.038-2.805 |  |  |  |  |

Supplementary Table S4

|  |  |  |
| --- | --- | --- |
| Table S4. Interventions for failing Fontan | |  |
| Variables | | N (%) |
| **Surgical interventions** | | 20 (26) |
|  | Pacemaker implantation | 8 (11) |
|  | Atrioventricular valve (AVV) procedure | 8 (11) |
|  | Secondary fenestration | 3 (4) |
|  | Resection of subaortic stenosis | 1 (1) |
| **Catheter interventions** | | 52 (68) |
|  | Balloon dilatation of TCPC pathway | 25 (33) |
|  | Stent implantation in TCPC pathway | 24 (32) |
|  | Closure of aortopulmonary collaterals (APCs) | 12 (16) |
|  | Closure of veno-venous collaterals (VVCs) | 12 (16) |
|  | Secondary fenestration | 4 (5) |