**Supplement**

**Materials and Methods**

***Setting***

Shizuoka Children's Hospital is a pediatric facility that was established in 1977. Our cardiovascular center performs approximately 280 cardiac surgeries annually—200 on-pump and 80 off-pump cardiopulmonary bypass surgeries. We accept consultations not only from our regional referral area but from all over Japan. A cardiac ICU (CICU) specializing in pediatric heart diseases is available and staffed 24 h a day by three pediatric cardiac intensivists and three dedicated fellows, working in collaboration with a cardiovascular surgeon. The CICU has 12 beds, and every patient who undergoes cardiovascular surgery is admitted to the CICU for postoperative management.

***Routine management***

*Surgical procedure*

At Shizuoka Children's Hospital, 15–20 cases of Fontan surgery are performed annually. When planning Fontan procedures, we determine the route of the conduit from each anatomical feature. Intracardiac or intra-extracardiac root is selected depending on the anatomical difficulty of installing an extracardiac conduit and the position of reflux of the hepatic vein. Fenestrations are constructed in patients at high risk of abnormally high venous return pressures. We did not set a clear threshold, such as central venous pressure. We made a comprehensive decision based on various data on whether to construct fenestrations. A median sternotomy was performed, and cannulas were placed in the ascending aorta, superior vena cava, and inferior vena cava to establish total cardiopulmonary bypass. Most of the operations involve a total cavopulmonary connection using a 16-mm or 18-mm extracardiac conduit. Intracardiac or intra-extracardiac total cavopulmonary connection was selected according to the orientation and morphology of the cardiovascular system. Ventricular fibrillation was induced during the creation of fenestrations, and cardioplegic arrest was instituted during simultaneous valvuloplasty, endocardial repair, intracardiac total cavopulmonary connection, and intra-extra cardiac total cavopulmonary connection. After being weaned from cardiopulmonary bypass, all patients underwent modified ultrafiltration.

*Anesthetic management*

Midazolam was administered transrectally in anxious children. Induction of anesthesia was performed by rapid intravenous infusion of propofol. Nasotracheal intubation was performed, and propofol and remifentanil were continuously administered during the procedure to maintain anesthesia. Similarly, vecuronium and fentanyl were intermittently administered. All patients received methylprednisolone 30 mg/kg and tranexamic acid preoperatively. We administered sufficient heparin before establishing cardiopulmonary bypass, and this was reversed with protamine postoperatively. With the aim of early extubation at 2–3 h postoperatively, the dose of fentanyl was kept below 30 μg/kg. Intercostal nerve blocks were not performed.

*CICU management*

At our hospital, all cardiovascular surgery patients are admitted to the CICU while intubated with the intent to extubate them as early as possible. We initially use a Siemens Maquet Servo-i ventilator (SOMA TECH INTL, Bloomfield, CT) on pressure-regulated volume control mode. Once spontaneous breathing resumes, we switched to pressure support ventilation. Extubation is performed once the following criteria are met: stable circulation, systolic blood pressure that is above the fifth percentile for the child’s age; normal lactate level (<2 mmol/L); adequate hemostasis; maintenance of respiratory function [spontaneous breathing test (fraction of inspired oxygen ≤0.5; positive end-expiratory pressure ≤5 cmH20, peak inspiratory pressure ≤10 cmH2O)]; and awaking while maintaining a sufficient airflow through the airway. Acetaminophen and low-dose intravenous continuous infusion of fentanyl and non-steroidal anti-inflammatory drugs were used for analgesia as needed. To maintain adequate circulation, dopamine, dobutamine, and, if necessary, continuous small doses of epinephrine were infused. In addition to the maintenance dose, blood transfusion and albumin preparations were used whenever they were appropriate.

**Result**

Respiratory failure was the most common cause of prolonged ventilation (n = 11, 52%), followed by circulatory instability (n = 6, 29%), hemorrhage and cardiac tamponade (n = 2, 9%), postoperative cerebral infarction (n = 1, 5%), and delayed postoperative waking (n = 1, 5%). Respiratory failure was caused by pulmonary edema and parenchymal damage in eight patients (73%), upper airway obstruction in two (18%) patients, and phrenic nerve palsy in one patient (9%).

 Although there is no clear definition of respiratory failure, it was assumed that the ventilator weaning does not proceed, and the cause was evaluated to be the respiratory system, including the airways and lungs. The definition of circulatory instability was mentioned in the CICU management section. Hemorrhage was treated as a case of bleeding that required a hemostasis procedure. Delayed postoperative waking was defined as a prolonged decline in consciousness that was evaluated as inability to maintain the airway after extubation.

**Supplementary Table S1.**

Postoperative profile

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|  | Early extubation group, n = 102 | Prolonged ventilation group, n = 21 | p value |
| SAP – admission (mmHg, IQR) | 88 (82–95) | 85 (75–95) | <0.01 |
| 12 h | 92 (80–101) | 86 (80–96) | 0.07 |
| 24 h | 93 (82–101) | 82 (77–96) | <0.01 |
| MAP – admission (mmHg, IQR) | 64 (59–70) | 63 (58–71) | 0.82 |
| 12 h | 68 (62–77) | 66 (60–70) | 0.16 |
| 24 h | 67 (61–74) | 65 (57–72) | 0.06 |
| CVP – admission (mmHg, IQR) | 14 (13–16) | 14 (13–16) | 0.81 |
| 12 h | 12 (10–13) | 14 (12–16) | 0.04 |
| 24 h | 12 (10–14) | 14 (12–17) | 0.19 |
| Lactate - admission (mmol/L, IQR) | 2.36 (1.73–3.06) | 3.20 (2.42–4.36) | 0.04 |
| 12 h | 1.35 (1.13–1.59) | 1.68 (1.21–2.05) | 0.27 |
| 24 h | 1.36 (1.13–1.66) | 1.43 (1.17–1.92) | 0.78 |
| SaO2 – admission (%, IQR) | 98.9 (97.9–99.3) | 98.3 (95.7–99.3) | 0.12 |
| 12 h | 98.1 (96.3–98.8) | 97.5 (93.3–97.9) | 0.06 |
| 24 h | 97.4 (95.1–98.7) | 96.1 (90.9–98.3) | 0.18 |
| Inotropic score – admission (score, IQR) | 8 (8–9) | 8 (8–11) | 0.03 |
| 12 h | 4 (3–6) | 6 (4–9) | 0.02 |
| 24 h | 0 (0–4) | 6 (4–9) | 0.02 |
| Extra volume 0–12 h (mL, IQR) | 491 (336–697) | 732 (592–1058) | <0.01 |
| extra volume 12–24 h | 159 (76–303) | 386 (159–665) | <0.01 |
| iNO (n, %) | 49 (48%) | 17 (81%) | <0.01 |
| Ventilation time (hours, IQR) | 7.0 (4.8–12.9) | 70.0 (42.6–115.4) | <0.01 |
| Noninvasive ventilation (n, %) | 10 (10%) | 3 (14%) | 0.56 |

CVP: central venous pressure, iNO: inhaled nitric oxide, IQR: interquartile range, MAP: mean arterial pressure, SAP: systolic arterial pressure, SaO2: oxygen saturation

**Supplementary Figure S1**

Distribution of ventilation times

