

### Total cavopulmonary connection in patients with the Down syndrome

*Furukawa T. (1), Park I. (2), Yoshikawa T. (2), Takahashi Y. (3)  
Department of Pediatrics, Juntendo University Urayasu hospital, Chiba, Japan (1); Department of Pediatrics, Sakakibara Heart Institute, Tokyo, Japan (2); Department of Pediatric Cardiovascular Surgery, Sakakibara Heart Institute, Tokyo, Japan (3)*

**Objectives:**

Total cavopulmonary connection (TCPC) operation is rarely performed for a functional single ventricle in a child with Down syndrome. Therefore, the postsurgical outcomes are not well known. Patients with Down syndrome are at a risk of developing persistent pulmonary hypertension and airway obstruction, which may affect the outcome of univentricular repair. We evaluated mortality and related factors after TCPC in Down syndrome.

**Methods:**

Between January 2004 and March 2010, we identified 8 patients with Down syndrome among 235 patients who had undergone TCPC. The course before TCPC, preoperative data, and postoperative course were evaluated. In addition, clinical parameters and the course after TCPC were compared between Down syndrome (N = 8, 6 boys and 2 girls) and Non-Down syndrome (N = 227) groups.

**Results:**

The median age at the time of TCPC was 4.1 years (3.4–5.5 years). The preoperative mean pulmonary artery pressure was  $13.9 \pm 1.81$  mmHg. We observed respiratory complications in 2 patients, surgical site infection in 3 patients, and chylothorax in 2 patients. No significant difference was observed between the Down and Non-Down syndrome groups in preoperative data and mortality rate: Down syndrome group, 1 of 8 patients (12.5%); Non-Down syndrome group, 5 of 227 patients (2.2%). However, when the clinical course after TCPC was examined, the duration of mechanical ventilation ( $P = 0.039$ ), length of ICU stay ( $P = 0.009$ ), duration of pleural drainage ( $P = 0.027$ ), and length of hospital stay ( $P = 0.007$ ) were found to be significantly prolonged in the Down syndrome group.

**Conclusions:**

Patients with Down syndrome tended to show prolonged recovery after TCPC, and suspected etiologies include respiratory tract disease, prolonged chylothorax, and surgical site infection.