Outcomes of patients with fetal heterotaxy syndrome in the current era

Takei K., Takeda A., Yamazawa H., Furukawa T., Izumi G., Sasaki O.
Hokkaido University Hospital, Sapporo, Japan

Background: Patients with heterotaxy syndrome (HS) have a wide range of outcomes depend on their associated anomalies. Previous reports had shown that patients with asplenia syndrome (AS) had worse outcome than polysplenia syndrome (PS) and live-born patients with AS were associated with survival rate of 21-60%. Considering progress in management and surgical procedure for these patients, we reassessed outcomes of patients with fetal HS in this study.

Methods: From October 2010 to December 2014, 22 consecutive patients were diagnosed HS in utero. We excluded a patient associated with congenital diaphragmatic hernia from this study. We obtained clinical data of these patients from clinical records and assessed.

Results: Fetal diagnoses were made at a median gestational age of 27 weeks (range 19-36). Of 21 patients, 11 (52%) were diagnosed with AS and 10 (48%) were diagnosed with PS. 2 patients with advanced atrioventricular block (AVB) were elected termination of pregnancy. All AS patients were live birth and had single-ventricle physiology. Of 11 AS patients, 8 (73%) were alive. Of 10 PS patients, 8 were live birth and 5 were considered to be candidates for biventricular repair. 4 (50%) patients were alive and 3 were waiting for biventricular repair. 4 patients (3 AS and 1 PS) were associated with total anomalous pulmonary venous connection and 2 were died soon after birth because of pulmonary venous obstruction (PVO). The other 2 were alive, but no patient had undergone Glenn or Fontan procedure. There were 4 PS patients with advanced AVB who had undergone pacemaker implantation shortly after birth. 2 were delivered prematurely at gestational ages of 28 and 35 weeks because of hydrops fetalis. Of these 4 patients, 3 were died because of pacing failure, postoperatively, and sepsis after operation for congenital biliary atresia. There was no patient who suffered from severe bacterial infection except for an above-mentioned patient.

Conclusions: PVO and AVB remained to be solved problems for HS patients. Outcomes in fetal HS patients without these associated defects were acceptable.