Pediatric Cardiothoracic Surgery in Patients With Unilateral Pulmonary Agenesis or Aplasia

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Background. Unilateral pulmonary agenesis or aplasia (UPA), a rare developmental defect of the lung, is sometimes associated with congenital heart and tracheal diseases. The purpose of this study was to assess our experience of pediatric cardiothoracic surgery in UPA patients.

Methods. Cardiothoracic surgery for congenital heart defect or tracheal stenosis performed between 1981 and 2010 in 8 UPA patients (ageneis in 5 and aplasia in 3) was reviewed retrospectively. Associated cardiac anomalies included ventricular septal defect, double outlet right ventricle with pulmonary atresia, total anomalous pulmonary venous connection, and interrupted aortic arch complex.

Results. For 7 patients with right UPA and 1 patient with left UPA, 12 cardiothoracic operations were performed, including 8 cardiac procedures in 4 patients and 4 tracheal procedures in 4 patients. Cardiac palliative repairs included Blalock-Park anastomosis, systemic-to-pulmonary artery shunt, and pulmonary artery banding. Cardiac definitive repairs included ventricular septal defect closure, subaortic membrane resection, modified Konno procedure, total anomalous pulmonary venous connection repair, and Rastelli-type operation. Tracheal repairs were costal cartilage tracheoplasty and slide tracheoplasty. The median age at surgery was 8 months and median body weight was 6.2 kg; the median operative time was 6.5 hours. There were 3 hospital deaths and 1 late death, with the 1-year mortality rate of 25%. Cardiopulmonary bypass-induced acute lung injury has occurred in 3 cases, 2 of which required extracorporeal membrane oxygenation support. Younger age of less than 1 month and prolonged cardiopulmonary bypass time of more than 200 minutes were related to operative risk factors for hospital mortality and morbidity.

Conclusions. Most of the pediatric cardiothoracic operations in UPA patients were successfully performed through an optimal surgical approach and procedure, but they still presented surgical risks of high mortality and morbidity. Perioperative management of UPA patients should be provided with a precise understanding of anatomic configuration and a careful consideration of underlying risk factors.

Developmental lung defect is a rare congenital anomaly, which is thought to result from bronchopulmonary foregut abnormalities in the fourth gestational week. The exact incidence of this anomaly remains unknown, but it was found in approximately one in 10,000 to 20,000 autopsies. The lesion occurs either unilaterally or bilaterally, with an equal distribution between the right and left lungs. Although its etiology is not fully understood, genetic, teratogenic, and mechanical factors, such as vitamin A or folic acid deficiency or the use of salicylates, may be responsible [1, 2]. Developmental lung defects have generally been classified into three categories according to the degree of pulmonary involvement: (1) agenesis, complete defect of parenchyma, bronchus, and artery; (2) aplasia, defect of parenchyma, and artery with rudimentary bronchus; and (3) hypoplasia, hypoplastic parenchyma and artery with rudimentary bronchus. Unilateral lesion of the anomaly presents as ipsilateral shift and rotation of the heart and mediastinum to the empty hemithorax, which results in a great displacement, distortion, and compression of the great vessels and airway [3, 4]. Although bilateral lesion of the anomaly is mostly incompatible with life, patients with unilateral lesion may be asymptomatic and diagnosed on a routine chest roentgenogram, or may present with respiratory distress and recurrent respiratory tract infection. Associated congenital anomalies, including
Cardiovascular, musculoskeletal, gastrointestinal, and urogenital systems are present in 50% to 75% of patients [4].

Unilateral pulmonary agenesis or aplasia (UPA) is characterized by total absence of the lung and vascular tissues, which is differentiated from pulmonary hypoplasia. Because UPA patients have no lung volume in hemithorax, the shift of the heart and mediastinum with consequent distortion of the vessels and airway can be greater in these patients than in patients with pulmonary hypoplasia. In general, UPA itself has an increased risk of mortality and morbidity, and the prognosis of right UPA is much worse than that of left UPA. Associated congenital anomalies are related to high mortality and morbidity [5]. Although successful cases of cardiothoracic surgery in UPA children have previously been reported [6–12], actual surgical outcomes are still unknown. The aims of the present study were to review our 30-year experience of pediatric cardiothoracic surgery in UPA patients, and to assess the surgical outcomes.

**Patients and Methods**

This study was approved by our Institutional Review Board, and the need for individual consent was waived. Clinical medical records and databases were retrospectively reviewed. Between August 1981 and August 2010, 8 UPA patients underwent pediatric cardiothoracic surgery at Kobe Children’s Hospital. The patient characteristics are summarized in Table 1. Right UPA was present in 7 patients, and left UPA was present in only 1 patient. UPA was detected in all patients using chest roentgenography, and subsequent chest computed tomography (CT) and bronchoscopy were necessary for definitive diagnosis (Fig 1). Preoperatively, mechanical ventilator support and balloon tracheoplasty were needed in patient 5, and tracheostomy was needed in patient 6.

**Cardiothoracic Surgery**

Twelve operations for a variety of cardiothoracic defects were performed in the setting of UPA: 8 cardiac procedures in 4 patients, and 4 tracheal procedures in 4 patients (Table 2). There were no simultaneous cardiac and tracheal operations.

**Cardiac Palliative Repair**

**Blalock-Park Anastomosis.** An anastomosis between the left subclavian artery and the descending aorta was performed through a left posterolateral thoracotomy [13]. Pulmonary artery banding was performed concomitantly.

**Systemic-to-Pulmonary Artery Shunt.** Through a left posterolateral thoracotomy, a 4-mm expanded polytetrafluoroethylene graft was anastomosed from the aortic arch to the main pulmonary artery [10].

**Pulmonary Artery Banding.** Through a median sternotomy, the main pulmonary artery was banded with a 2.5-mm-wide expanded polytetrafluoroethylene strip after patent ductus arteriosus ligation.

**Cardiac Definitive Repair**

Total cardiopulmonary bypass (CPB) with a moderate systemic hypothermia was established between the ascending aorta and both venae cavae, except case 5 in which venous cannulas were inserted into the innominate vein and the inferior vena cava.

### Table 1. Patient Characteristics

<table>
<thead>
<tr>
<th>Pt. No.</th>
<th>Sex</th>
<th>Lung Defect</th>
<th>Cardiac Position</th>
<th>Associated Cardiothoracic Anomalies</th>
<th>Underlying Genetic Disorder</th>
<th>Initial Clinical Symptom</th>
<th>Onset of Symptom</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Female</td>
<td>Right agenesis</td>
<td>Dextrocardia</td>
<td>Tracheal stenosis</td>
<td>–</td>
<td>Respiratory distress</td>
<td>10 months</td>
</tr>
<tr>
<td>2</td>
<td>Male</td>
<td>Right agenesis</td>
<td>Dextrocardia</td>
<td>IAA, VSD, ASD, SAS</td>
<td>–</td>
<td>Stridor</td>
<td>After birth</td>
</tr>
<tr>
<td>3</td>
<td>Male</td>
<td>Right agenesis</td>
<td>Dextrocardia</td>
<td>TAPVC, ASD, PDA</td>
<td>Goldenhar syndrome</td>
<td>Cyanosis</td>
<td>After birth</td>
</tr>
<tr>
<td>4</td>
<td>Male</td>
<td>Left agenesis</td>
<td>Levocardia</td>
<td>DORV, PA, VSD</td>
<td>–</td>
<td>Cyanosis</td>
<td>After birth</td>
</tr>
<tr>
<td>5</td>
<td>Female</td>
<td>Right aplasia</td>
<td>Dextrocardia</td>
<td>VSD, ASD, PDA, tracheal stenosis</td>
<td>Goldenhar syndrome</td>
<td>Respiratory distress</td>
<td>After birth</td>
</tr>
<tr>
<td>6</td>
<td>Female</td>
<td>Right agenesis</td>
<td>Dextrocardia</td>
<td>Tracheal stenosis</td>
<td>–</td>
<td>Respiratory tract infection</td>
<td>10 months</td>
</tr>
<tr>
<td>7</td>
<td>Female</td>
<td>Right aplasia</td>
<td>Dextrocardia</td>
<td>Tracheal stenosis, VSD</td>
<td>VATER association</td>
<td>Respiratory distress</td>
<td>After birth</td>
</tr>
<tr>
<td>8</td>
<td>Female</td>
<td>Right aplasia</td>
<td>Dextrocardia</td>
<td>Tracheal stenosis, ASD</td>
<td>VATER association</td>
<td>Respiratory distress</td>
<td>4 months</td>
</tr>
</tbody>
</table>

ASD = atrial septal defect; DORV = double outlet right ventricle; IAA = interrupted aortic arch; No. = number; PA = pulmonary atresia; PDA = patent ductus arteriosus; Pt. = patient; SAS = subaortic stenosis; TAPVC = total anomalous pulmonary venous connection; VSD = ventricular septal defect.
VSD CLOSURE. Through a right posterolateral thoracotomy, the perimembranous VSD was closed with a Dacron (C.R. Bard, Haverhill, PA) patch through a right atrial approach [10].

SUBAORTIC MEMBRANE RESECTION. A right posterolateral thoracotomy was performed in resecting the subaortic fibromuscular membrane to relieve subaortic stenosis through a J-shaped oblique aortotomy.

MODIFIED KONNO PROCEDURE. Through median sternotomy, a modified Konno procedure [14] to relieve subaortic stenosis was performed. The septal incision, longitudinal aortotomy, and right ventricular infundibulotomy were closed using equine pericardial patches.

TAPVC REPAIR. Through a right posterolateral thoracotomy, a side-to-side anastomosis of the pulmonary venous confluence and the left atrium was performed through a superior approach [10].

RASTELLI-TYPE OPERATION. Through median sternotomy, an intraventricular tunnel was constructed from the VSD to the aorta with the Dacron patch through a right ventricular approach. The right ventricular outflow tract was reconstructed using a tailored pericardial roll with a polytetrafluoroethylene monocuspid valve.

**Tracheal Repair**

Standard median sternotomy with a transverse neck incision was used for the surgical approach.

COSTAL CARTILAGE TRACHEOPLASTY. The tracheal stenotic segment was enlarged by a rib cartilage graft [12, 15].

SLIDE TRACHEOPLASTY. Normothermic cardiopulmonary bypass was established by cannulating the ascending aorta and the right atrium (RA). After dividing the tracheal stenotic segment at the midpoint, sliding oblique anastomosis was performed [12, 16].

**Statistical Analysis**

All values were expressed as median with range for nonnormally distributed variables. Univariate analysis was performed to identify risk factors for hospital mortality and morbidity, such as acute lung injury (ALI). Variables included age, body weight, sex, UPA (agenesis or aplasia, right or left), and CPB time. Data were analyzed using StatView J-5.0 software (SAS Institute, Cary, NC). Differences were considered statistically significant at \( p < 0.05 \).

**Results**

Operative data are summarized in Table 2. The median age at surgery was 8 months and median body weight was 6.2 kg, and the median operative time was 6.5 hours. In patients undergoing surgery with CPB, median CPB time was 183 minutes.

**Cardiac Palliative Repair**

Blalock-Park anastomosis (case 2) and pulmonary artery banding (case 9) in patients with right UPA were performed without difficulty through a standard approach. In case 7 (left UPA), surgical access to the aortic arch and the main
pulmonary artery was easy and adequate for the systemic-to-pulmonary artery shunt through left thoracotomy.

Cardiac Definitive Repair

Median sternotomy in cases 5 and 8 was carefully performed with the lung deflated because the unilateral lung extended into the contralateral hemithorax of the absent lung and covered the heart. After the lung was mobilized out of the contralateral hemithorax and off the pericardium, the Rastelli operation in case 8 (left UPA) was successfully performed with easy aortic and venous cannulation of CPB and acceptable exposure of the intracardiac anatomy. In case 5 (right UPA), however, venous cannulation of CPB was performed in the innominate vein because of difficult access in the superior vena cava and the RA (Figs 2A, 3A). The subaortic area was also difficult to access for the modified Konno procedure. Occasionally, we stood at the left side of the operating table to obtain satisfactory exposure of the surgical field and easy handling in this case. Right thoracotomy in cases 3, 4, and 6 (right UPA) allowed easy cannulation of CPB (Fig 2B) and adequate exposure of surgical sites for VSD closure and TAPVC repair. However, the access to the subaortic area was not perfect for performing subaortic membrane resection.

Tracheal Repair

To relieve direct aortic compression of the trachea (Fig 3B), slide tracheoplasty was conducted with translocation of the trachea anterior to the aortic arch in cases 10 and 12. This modification provided us with a good surgical view of the trachea to reconstruct. We performed tracheopexy in case 10 and aortopexy in case 12 concomitantly.

Postoperative Outcomes

The median follow-up period was 4.1 years (range, 50 days to 22 years). There were 3 hospital deaths (cases 5, 6, and 11) and 1 late death (case 9). The 30-day and 1-year mortality rates were 0% and 25.0%, respectively. In 3 cases, ALI occurred immediately after CPB. Of these cases, extracorporeal membrane oxygenation support was required for 3 days after surgery in case 4, and for 4 weeks in case 5. Patient 2 of the cases died of multiple complications.
organ failure and infection at 50 days after modified Konno procedure. Patient 3 required prolonged ventilator support with inhaled nitric oxide for ALI treatment, and died of severe renal failure on postoperative day 100. Patient 7 died of septic shock due to pseudomonal infection on postoperative day 108. Patient 5 underwent tracheostomy on postoperative day 27 because of difficulty in tracheal extubation, and died of unanticipated endotracheal tube obstruction at home 3 years later. The postoperative course was uneventful in the other cases. Univariate analysis showed that the younger age of less than 1 month and prolonged CPB time of more than 200 minutes were related to operative risk factors for hospital mortality and morbidity in UPA patients ($p = 0.0285$ and $p = 0.0304$, respectively).

Comment

This retrospective review involved clinical assessment of our 30-year experience of pediatric cardiothoracic surgery in UPA patients. We present 12 cardiothoracic operations in 8 patients in this study. Patients with UPA have unique anatomic features such as an ipsilateral shift and rotation of the heart and mediastinum to the empty hemithorax \[3, 4\]. Surgeons are required to identify the anatomic displacement and to make certain modifications to normal surgical techniques when operating on such patients.

Congenital heart defect in combination with UPA occurs very rarely, and previously published articles are mostly limited to case reports. To the best of our knowledge, only 12 surgical cases of congenital heart defect with UPA, except for our cases, have been reported in the literature (Table 3). The anatomic displacement of the heart in UPA patients can affect technical aspects of cardiac surgery, including the establishment of CPB and the intracardiac repair. In left UPA cases, Palma and colleagues \[17\], Guo and colleagues \[9\], and Akishima and associates \[18\] reported that VSD or atrial septal defect (ASD) closure were performed through right atrial approach on standard CPB with cardiac arrest through median sternotomy.

### Table 3. Reported Cases of Pediatric Cardiac Surgery in Patients With Unilateral Pulmonary Agenesis or Aplasia

<table>
<thead>
<tr>
<th>First Author [Ref]</th>
<th>UPA</th>
<th>CHD</th>
<th>Age</th>
<th>Sex</th>
<th>Operative Procedure</th>
<th>Surgical Approach</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boxer [7], 1978</td>
<td>Agenesis, L</td>
<td>TAPVC</td>
<td>4 mon</td>
<td>F</td>
<td>TAPVC repair</td>
<td>–</td>
<td>Alive</td>
</tr>
<tr>
<td>Akishima [18], 1994</td>
<td>Agenesis, L</td>
<td>VSD</td>
<td>2 mon</td>
<td>M</td>
<td>VSD closure</td>
<td>Median sternotomy</td>
<td>Alive</td>
</tr>
<tr>
<td>Finci [22], 1999</td>
<td>Agenesis, R</td>
<td>TAPVC</td>
<td>–</td>
<td>–</td>
<td>TAPVC repair</td>
<td>–</td>
<td>Died</td>
</tr>
<tr>
<td>Ishikawa [23], 1999</td>
<td>Agenesis, R</td>
<td>VSD, ASD</td>
<td>20 mon</td>
<td>M</td>
<td>VSD/ASD closure</td>
<td>Right thoracotomy</td>
<td>Alive</td>
</tr>
<tr>
<td>Pietrzykowski [11], 2006</td>
<td>Agenesis, R</td>
<td>ASD</td>
<td>6 years</td>
<td>M</td>
<td>ASD closure</td>
<td>Median sternotomy</td>
<td>Alive</td>
</tr>
<tr>
<td>Chou [24], 2007</td>
<td>Agenesis, R</td>
<td>VSD, ASD</td>
<td>–</td>
<td>M</td>
<td>VSD/ASD closure</td>
<td>–</td>
<td>Alive</td>
</tr>
<tr>
<td>Agniesis, R</td>
<td>ASD</td>
<td>–</td>
<td>F</td>
<td>ASD closure</td>
<td>–</td>
<td>Alive</td>
<td></td>
</tr>
<tr>
<td>Chiang [8], 2009</td>
<td>Agenesis, L</td>
<td>TA, VSD, ASD</td>
<td>5 mon</td>
<td>M</td>
<td>BDG, ASD enlargement</td>
<td>–</td>
<td>Alive</td>
</tr>
<tr>
<td>Palma [17], 2010</td>
<td>Agenesis, L</td>
<td>VSD, ASD</td>
<td>2 years</td>
<td>M</td>
<td>VSD/ASD closure</td>
<td>Median sternotomy</td>
<td>Alive</td>
</tr>
<tr>
<td>Guo [9], 2011</td>
<td>Agenesis, L</td>
<td>VSD</td>
<td>5 years</td>
<td>F</td>
<td>VSD closure</td>
<td>Median sternotomy</td>
<td>Alive</td>
</tr>
</tbody>
</table>

ASD = atrial septal defect; BDG = bidirectional Glenn shunt; CHD = congenital heart disease; CoA = coarctation; F = female; L = left; LPA = left pulmonary artery; M = male; mon = month; R = right; TA = tricuspid atresia; TAPVC = total anomalous pulmonary venous connection; UPA = unilateral pulmonary agenesis or aplasia; VSD = ventricular septal defect.
associates [11], who reported ASD closure through a left atrial approach using total circulatory arrest and deep hypothermia through median sternotomy in a patient with right UPA, commented that it would be difficult to insert the venous cannula into the RA, which was positioned more posteriorly than usual. Previously, Ishikawa and coworkers [18] reported VSD/ASD closure on standard CPB with cardiac arrest through right atrial approach with right thoracotomy in a patient with right UPA.

Before surgery, we currently obtain anatomic information to determine the surgical approach and procedure by three-dimensional multidetector-row computed tomography angiography (3D-MDCTA). For the right UPA case, the 3D-MDCTA image of anterior view showed that it would be difficult to visualize the superior vena cava and RA because of the rightward displacement of the heart (Fig 3A). Based on this preoperative CT assessment, we inserted the venous cannula of CPB into the innominate vein. In the right UPA cases, it was easy to access the superior vena cava and RA for the cannulation of CPB and the intracardiac repair through right thoracotomy. However, the exposure of the subaortic area through either right thoracotomy or median sternotomy may not be deemed adequate to perform the release of subaortic stenosis in a patient with right UPA (cases 4 and 5). Further prospective planning against surgical handling problems, such as poor exposure and access, is needed.

Congenital tracheal stenosis with UPA is also a rare occurrence, and recently published papers describing tracheoplasty for congenital tracheal stenosis with UPA are summarized in Table 4. As far as we have found, a total of 29 UPA patients, except for our cases, have previously undergone tracheal surgery. Right UPA was present in 21 patients, and left UPA in 8. Backer and colleagues [6] described 6 UPA patients with tracheal stenosis and associated cardiac anomalies requiring simultaneous repair: pulmonary artery sling in 4 patients, and VSD in 2. Of these patients, a 4-month-old infant underwent VSD closure through the left atrium and translocation of the trachea anterior to the aortic arch to facilitate the autograft tracheoplasty.

Costal cartilage tracheoplasty in our case 1 was the first successful surgical treatment of long segment congenital tracheal stenosis [15], and slide tracheoplasty has been performed in our institution since 1997 [12]. In the present study, we experienced 3 cases of slide tracheoplasty in patients with right UPA. Displacement and rotation of the heart to the right hemithorax has the potential to cause an aortic compression of the trachea (Fig 3B). Based on the preoperative CT assessment, we reconstructed the trachea with a translocation of the trachea anterior to the aortic arch in cases 10 and 12. To date, postoperative courses after the modified slide tracheoplasty have been uneventful without any airway distortion and obstruction. Aortopexy or tracheopexy were also useful to prevent postoperative tracheomalacia [19].

Recently, we reported that CPB-induced ALI is one of the most serious complications of cardiac surgery in children with UPA [20]. Ischemia-reperfusion injury, fluid overload, atelectasis-reexpansion, and ventilator-induced injury of the lung during surgery can be associated with CPB-induced ALI in UPA patients. We suggest that careful perioperative management to prevent CPB-induced ALI is necessary for UPA patients. A specific neutrophil elastase inhibitor, sivelestat sodium hydrate (0.2 mg · kg$^{-1}$ · h$^{-1}$) [21], is currently administered during and after pediatric cardiac surgery in UPA patients. Extracorporeal membrane oxygenation support or ventilator support with inhaled nitric oxide is selected as necessary.

The literature review, including the present study, showed that cumulative total number of pediatric cardiothoracic operations in UPA patients is 53. Of these, 4 patients (7.7%) died within 30 days after surgery. The 1-year mortality rate after surgery was 23.1%. Congenital heart defect and tracheal stenosis in UPA patients still require great efforts to improve outcomes. The present study demonstrated the younger age (less than 1 month) and prolonged CPB time (longer than 200 minutes) as risk factors.

<table>
<thead>
<tr>
<th>First Author [Reference]</th>
<th>UPA (n)</th>
<th>R:L</th>
<th>Age</th>
<th>Sex (M:F)</th>
<th>Operative Procedure</th>
<th>Survival (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weber [25], 1991</td>
<td>5</td>
<td>3:2</td>
<td>2−6 mon</td>
<td>3:2</td>
<td>Cartilage tracheoplasty, Tracheal resection</td>
<td>2</td>
</tr>
<tr>
<td>Antonio-Pacheco [26], 2003</td>
<td>2</td>
<td>2:0</td>
<td>2 mon, 6 mon</td>
<td>1:1</td>
<td>Cartilage tracheoplasty, Tracheal resection</td>
<td>1</td>
</tr>
<tr>
<td>Kocyildirim [27], 2004</td>
<td>2</td>
<td>2:0</td>
<td>–</td>
<td>–</td>
<td>Pericardial tracheoplasty, Slide tracheoplasty</td>
<td>–</td>
</tr>
<tr>
<td>Airway Reconstruction Team [28], 2005</td>
<td>1</td>
<td>0:1</td>
<td>48 days</td>
<td>0:1</td>
<td>Slide tracheoplasty</td>
<td>1</td>
</tr>
<tr>
<td>Chou [24], 2007</td>
<td>1</td>
<td>1:0</td>
<td>–</td>
<td>0:1</td>
<td>Slide tracheoplasty</td>
<td>0</td>
</tr>
<tr>
<td>Backer [6], 2009</td>
<td>11</td>
<td>9:2</td>
<td>13 days–3.5 years</td>
<td>3:8</td>
<td>Pericardial tracheoplasty, Cartilage tracheoplasty, Tracheal resection, Slide tracheoplasty</td>
<td>9</td>
</tr>
<tr>
<td>Speggiorin [29], 2011</td>
<td>7</td>
<td>4:3</td>
<td>1–12 mon</td>
<td>2:7</td>
<td>Slide tracheoplasty</td>
<td>6</td>
</tr>
</tbody>
</table>

F = female; L = left; M = male; mon = month; R = right; UPA = unilateral pulmonary agenesis or aplasia.
factors for hospital mortality and morbidity after cardiothoracic surgery in UPA patients. Although there is a statistical limitation on the small sample size and univariate analysis, we suggest that careful management of patients with these high-risk factors is needed during and after surgery to avoid postoperative complications and improve their prognosis.

In conclusion, we reviewed our 30-year experience of pediatric cardiothoracic surgery in UPA patients. Most of the cardiothoracic operations in our institution have been successfully performed through an optimal surgical approach and procedure, but some of the UPA patients were still at surgical high risk of mortality and morbidity. Perioperative management of these patients with UPA should be provided with a precise understanding of anatomic configuration and a careful consideration of underlying risk factors.

References