The Natural and Surgically Modified History of Anomalous Pulmonary Veins From the Left Lung

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Background. Reconstruction of anomalous left pulmonary veins (ALPV) requires an anastomosis at a nonanatomic position, posing the potential risk of pulmonary vein obstruction (PVO).

Methods. The 514 patients who were diagnosed with a pulmonary vein abnormality from 1990 to 2010 were reviewed. Thirty-eight patients (7.4%; median age, 1.4 years; interquartile range, 0.1 to 5.7 years) were identified. ALPV was diagnosed as an isolated anomaly in 23 (61%) or as part of mixed total anomalous pulmonary venous drainage in 15 (39%). Patients were divided into 3 groups (group 1: partial ALPV, treated; group 2: total ALPV, treated; or group 3: partial ALPV, untreated). Freedom from poor clinical potential risk of PV obstruction (PVO). Various reconstructive techniques and approaches have been described with reasonable short-term outcomes [1, 8, 9]. Our group recently reported the application of sutureless repair for ALPV as a part of mixed total anomalous pulmonary venous drainage (TAPVD) [10]. This study described our management of ALPV in the last 20 years and attempted to identify high-risk subgroups for PVO after repair. We also sought to analyze the functional outcomes of untreated ALPV to define the natural history of this entity.

Results. Repair in 30 ALPV patients (79%) was performed with direct anastomosis to the left atrium (n = 26 [68%]) or by sutureless repair (n = 4 [11%]). Two deaths occurred in group 2 (heart failure and PVO). The 4 reoperations in group 2 were prompted by PVO and occurred within 6 months of the initial repair. There was a nonsignificant trend of lower freedom from poor outcomes in group 2 (74.6%) vs group 1 (100%) at 10 years (p = 0.105). There was no difference in the incidence of any left PVO among the groups (p = 0.381). Severe left PVO did not develop in group 3 (n = 8 [21%]).

Conclusions. Total ALPV carries a high risk of early PVO. Thus, the optimal surgical approach remains elusive. Untreated partial ALPV remained unobstructed during midterm follow-up. Therefore, surgical treatment may not be necessary in patients with partial ALPV.

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divided into two types: partial ALPV—only 1 left upper PV is anomalous, and total ALPV—both left upper and lower PVS are anomalous.

Surgical Technique

**Direct Anastomosis of the PV(s) to the Left Atrial Appendage.** This procedure was routinely performed with cardiopulmonary bypass. The vertical vein from the ALPV was divided at the junction to the innominate vein. The location of the incision on the left atrial appendage (LAA) depended on the location and length of the vertical vein. If the vertical vein was located anteriorly or was relatively short, the tip of the LAA was opened, and the trabeculae were trimmed. The vertical vein was then anastomosed to the LAA with an anterior pericardial patch augmentation. The posterior wall was anastomosed using a continuous suture technique with fine Prolene sutures (Ethicon, Somerville, NJ), and the anterior anastomosis was done with an interrupted suture technique. If the vertical vein was located posteriorly, the base of the LAA was used for the anastomosis with anterior patch augmentation.

Sutureless Repair. This technique was used in patients with ALPV as a part of mixed TAPVD [10]. A detailed surgical description has been previously reported [10, 11]. In brief, the posterior pericardium and the anterior wall of the ALPV confluence were opened and unroofed. The anterior wall of the individual PVSs were cut back and unroofed, if necessary. The LA was then opened transversely, and the incision was extended into the base of the LAA. The anastomosis between the LA and the posterior pericardium was performed with a continuous suture technique using 6-0 or 7-0 Prolene sutures.

Outcome Assessment

PVO was determined by review of the echocardiographic reports. PVO was defined as mild PVO (mean gradient of 3 to 5 mm Hg), severe PVO (mean gradient of >6 mm Hg), and any PVO (any patient with a mean gradient >3 mm Hg). Patients with ALPV were grouped to compare the natural and surgically modified history based on the anatomy and treatment strategy as follows: group 1—partial ALPV, treated; group 2—total ALPV, treated; and group 3—partial ALPV, untreated.

Statistical Analysis

Continuous data are presented as median (IQR). Discrete data are presented as frequency (percentage). The level of statistical significance was set at a p value of 0.05 or less. Differences between the groups were analyzed with the Mann-Whitney U test. Event frequencies were compared with the χ² or Fisher test. Freedom from poor clinical (death/reoperation) and functional outcomes (any PVO, mean pressure gradient >3 mm Hg) were analyzed with Kaplan-Meier analysis and a log-rank test. Predictors for death/reintervention or any PVO were explored with Cox regression. Statistical analysis was performed using SPSS 17.0 software (IBM Corp, Armonk, NY).

Results

Patient demographics are reported in Table 1. Various coexisting anomalies were present in 30 of 38 patients (79%). Of 23 patients who had isolated ALPV, 14 patients had partial ALPV and 9 patients had total ALPV. Preoperative echocardiography demonstrated PVO in 6 patients, which was mild in 4 and severe in 2. In patients with mixed TAPVD, 4 had partial ALPV and 11 had total ALPV. The ALPV patterns observed in this cohort are shown in Figure 1. There were 11 patterns seen in patients associated with mixed TAPVD.

Surgical repair was performed in 30 of 38 patients (79%; Fig 2). The surgical techniques are outlined in Table 2. Direct anastomosis between the PV and LA was performed in 26 patients (68%). Types of ALPV were isolated in 19 patients (partial ALPV in 10 and total ALPV in 9) and were part of mixed TAPVD in 7 (all had total ALPV). Sutureless repair was used in 4 patients who had total ALPV in the setting of mixed type TAPVD.

Eight patients with partial ALPV were left untreated. Four patients with isolated partial ALPV were not

![Image](https://via.placeholder.com/15x678)

**Table 1. Patient Demographics**

<table>
<thead>
<tr>
<th>Variables</th>
<th>No. (%) or Median (IQR)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at operation/assessment, y</td>
<td>1.4 (0.1–5.7)</td>
</tr>
<tr>
<td>Body weight, kg</td>
<td>8.9 (3.9–20.8)</td>
</tr>
<tr>
<td>Body surface area, m²</td>
<td>0.48 (0.25–0.72)</td>
</tr>
<tr>
<td>Diagnosis</td>
<td></td>
</tr>
<tr>
<td>Isolated ALPV partial ALPV</td>
<td>14/38 (36%)</td>
</tr>
<tr>
<td>Total ALPV</td>
<td>9/38 (23%)</td>
</tr>
<tr>
<td>Mixed TAPVD partial ALPV</td>
<td>4/38 (11%)</td>
</tr>
<tr>
<td>Total ALPV</td>
<td>11/38 (29%)</td>
</tr>
<tr>
<td>Coexisting cardiac anomaly</td>
<td></td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>16/38 (42%)</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>2/38 (5%)</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>6/38 (16%)</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>3/38 (8%)</td>
</tr>
<tr>
<td>Others</td>
<td>9/38 (24%)</td>
</tr>
<tr>
<td>Surgical technique</td>
<td></td>
</tr>
<tr>
<td>Direct anastomosis</td>
<td>26 (67)</td>
</tr>
<tr>
<td>Sutureless repair</td>
<td>4 (11)</td>
</tr>
<tr>
<td>Untreated</td>
<td>8 (22)</td>
</tr>
<tr>
<td>Approach</td>
<td></td>
</tr>
<tr>
<td>Median sternotomy</td>
<td>30 (100)</td>
</tr>
<tr>
<td>Left thoracotomy</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Cardiopulmonary bypass</td>
<td>30 (100)</td>
</tr>
<tr>
<td>Cardiopulmonary bypass time, min</td>
<td>82 (62–131)</td>
</tr>
<tr>
<td>Concomitant procedure</td>
<td></td>
</tr>
<tr>
<td>Atrial septal defect closure</td>
<td>16/30 (53%)</td>
</tr>
<tr>
<td>Ventricular septal defect closure</td>
<td>2/30 (7%)</td>
</tr>
<tr>
<td>Patent ductus arteriosus closure</td>
<td>6/30 (20%)</td>
</tr>
<tr>
<td>Pulmonary stenosis repair</td>
<td>3/30 (10%)</td>
</tr>
<tr>
<td>Others</td>
<td>8/30 (27%)</td>
</tr>
</tbody>
</table>

ALPV = anomalous left pulmonary vein(s); IQR = interquartile range; TAPVD = total anomalous pulmonary venous drainage.
treated because they did not have any other cardiac anomaly or right ventricular volume overload. In 4 patients with mixed TAPVD, ALPV was not repaired during TAPVD repair because the ALPV was small and technically challenging or appeared to be hemodynamically insignificant. Of these 8 who were not treated, physiologic assessment by magnetic resonance imaging (MRI) in 3 patients showed pulmonary-to-systemic flow ratios (Qp/Qs) of 1.3, 1.4, and 1.7, respectively.

**Fig 1.** Anomalous left pulmonary vein (ALPV) patterns of patients in this cohort. (CS = coronary sinus; INNV = innominate vein; IVC = inferior vena cava; LA = left atrium; LLPV = left lower pulmonary vein; LLUP = left upper pulmonary vein; RA = right atrium; RLPV = right lower pulmonary vein; RMPV = right middle pulmonary vein; RUPV = right upper pulmonary vein; SVC = superior vena cava; TAPVD = total anomalous pulmonary venous drainage; VV = vertical vein.)

**Fig 2.** Anatomic variation and treatment is shown for patients with isolated anomalous left pulmonary veins (ALPV) and total anomalous pulmonary venous drainage (TAPVD).
Clinical Outcome

One hospital death occurred in a patient who had double-outlet right ventricle with partial ALPV. The patient died of pulmonary hypertension and right ventricular failure, which were unrelated to the partial ALPV. During a median follow-up of 4.3 years (IQR, 0.7 to 12.6 years), 4 patients required reoperation (Table 3):

- Patient 1, with isolated total ALPV, underwent a direct anastomosis, and stenosis developed at the vertical vein and at the anastomotic site. Reaugmentation of the anastomotic site and the vertical vein was performed, without success.
- Patient 2, with isolated total ALPV, underwent a direct anastomosis between the vertical vein and the LAA. Severe stenosis developed at the site of the anastomosis at the vertical vein, which had a hairpin turn (Fig 3). Conventional reoperation with reaugmentation of the anastomotic site was unsuccessful. She subsequently underwent modified sutureless repair with the PV confluent that was located posterior to the pericardium and the LA. The connection has remained patent 2 years after repair.
- Patient 3, with mixed TAPVD, underwent a direct anastomosis between the left PVs and LA, and stenosis developed at the site of anastomosis. The anastomosis was revised at reoperation, and the PVs have remained unobstructed for 20 years.
- Patient 4, with mixed TAPVD, presented with left PVs connected to the coronary sinus and right PVs connected to the inferior vena cava (Scimitar veins). The initial operation was unroofing the coronary sinus and creating a baffle from the right PVs to the LA. The right PVs and left lower PV had severe stenosis 2 months after the initial operation, and the patient required extracorporeal membrane oxygenation because of pulmonary hypertension. The patient underwent repeat repair for post-repair PVO but died 2 days later of a cerebral infarction.

After reoperation, all PVs, except the left upper PV, remained severely stenosed distal to the anastomosis. Patients 1, 2, and 3 had only mild left PVO, but reoperation was performed because discrete stenosis was found on MRI. Successful relief of PVO at reoperation was achieved in 2 of the 4 patients (50%).

Freedom from poor clinical outcomes (death or reoperation) was 85.5% at 5, 10, and 15 years after repair. There was no difference in poor clinical outcomes between those who were and were not surgically treated (85.2% vs 87.5% at 15 years, p = 0.957). There was no difference in freedom from poor outcomes between isolated ALPV (87.0%) and mixed TAPVD (81.8%) groups (p = 0.977). Freedom from poor outcome was comparable among the groups at 15 years after repair (group 1, 100%; group 2, 76.2%; group 3, 87.5%; p = 0.292; Fig 4). Group 3 did not require reoperation during follow-up. No risk factors for death or reoperation were identified.

Functional Outcome

Complete echocardiographic follow-up was available in 26 patients (68%). During the follow-up period, mild left PVO developed in 8 patients and severe left PVO developed in 1 patient (Table 3). In group 3, mild left PVO developed at the vertical vein from the left upper PV draining into the inferior vena cava in 1 patient at 14 years of age, but without clinical symptoms. There was no late severe left PVO in any of the three groups. Overall freedom from any (mild or more) left PVO was 72.2%, 60.2%, 32.1%, respectively, at 5, 10, and 15 years (Fig 5A). There was no difference in any left PVO between surgical and nonsurgical groups at 15 years after repair (32.6% vs 50.0%, p = 0.183; Fig 5B). No difference in left PVO was seen between isolated ALPV (36.3%) and mixed TAPVD (36.4%) groups at 15 years (p = 0.701). There was no difference in freedom from any left PVO among the groups at 10 years after repair (group 1, 53.3%; group 2, 57.8%; and group 3, 100.0%; p = 0.381; Fig 5C). No severe left PVO developed in group 3. No risk factors for left PVO were identified.

Comment

ALPV is a rare cardiac anomaly that includes a challenging diversity of anatomic arrangements. The major technical challenge is to create a nonobstructive venous connection in a nonanatomic fashion. Furthermore, the surgical indications for asymptomatic patients with left upper lobe vein-only ALPV remain debatable.

The main purpose of this study was to outline the anatomic patterns and evaluate the natural and surgically modified history of ALPV. Survival is excellent after repair for isolated ALPV or ALPV as a part of mixed TAPVD. However, early PVO and the subsequent risk of reoperation highlight the need for improvements in existing surgical methods. Reduction in the early risk of PVO is important because there is no significant risk of late PVO regardless of ALPV pattern. Patients with untreated ALPV rarely developed late PVO.

Surgical Technique and Approach

In the setting of ALPV, the vertical vein from the ALPV generally crosses the left pulmonary artery and runs

Table 2. Type of Operation and Pulmonary Veins

<table>
<thead>
<tr>
<th>Variable*</th>
<th>Isolated ALPV</th>
<th>Mixed TAPVD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, y</td>
<td>0.5 (0.3–0.7)</td>
<td>0.1 (0.1–0.2)</td>
</tr>
<tr>
<td>Group 1: Treated, partial ALPV</td>
<td>10</td>
<td>0</td>
</tr>
<tr>
<td>Direct anastomosis</td>
<td>10</td>
<td>0</td>
</tr>
<tr>
<td>Sutureless repair</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Group 2: Treated, total ALPV</td>
<td>9</td>
<td>11</td>
</tr>
<tr>
<td>Direct anastomosis</td>
<td>9</td>
<td>7</td>
</tr>
<tr>
<td>Sutureless repair</td>
<td>0</td>
<td>4</td>
</tr>
<tr>
<td>Group 3: Untreated, partial ALPV</td>
<td>4</td>
<td>4</td>
</tr>
</tbody>
</table>

* Continuous data are shown as median (interquartile range) and discrete data as frequency.

ALPV = anomalous left pulmonary vein(s); TAPVD = total anomalous pulmonary venous drainage.
vertically towards the innominate vein. The divided edge of the vertical vein acquires a “hairpin turn” to reach the LAA, especially in the anatomic arrangement of total ALPV. Because of this unique anatomic relationship, there is a risk of kinking the vertical vein or creating stenosis at the anastomotic site, or both. ElBardissi and associates [8] reported the use of an end-to-end anastomosis between the vertical vein and LAA with interrupted sutures in the anterior circumference of the anastomosis through a left thoracotomy in the adult population. Use of a pericardial patch on the anterior aspect after creating continuity between the vertical vein and LAA through median sternotomy was reported by Ban and colleagues [12]. Ports and associates [13] described the technique that uses the entire back wall of the LA instead of the LAA, using cardiopulmonary bypass, which may result in a more anatomic connection. A side-to-side connection between the vertical vein and LAA without dividing the vertical vein may be an option to avoid kinking of the vertical vein, although we did not apply this technique in this series.

The sutureless repair may be a good surgical option if the PV confluence is located just behind and maintains fibrous continuity with the pericardium. This procedure can be done in a very similar fashion to the way we perform primary sutureless repair for TAPVD [14] as long as the tissue between the posterior pericardium and ALPV have a relatively strong tissue continuity. We recently performed a modified sutureless repair in a patient with isolated total ALPV that resulted in an unobstructed flow pathway of the total left PV. More experience with follow-up by MRI is required to determine the efficacy of primary sutureless repair in patients with total ALPV. The sutureless repair is our preferred strategy for the management of total ALPV as a part of mixed TAPVD [10].

Patterns of ALPV and Surgical Technique
Two patterns of ALPV were found in this study: (1) isolated ALPV in which left PV(s) have anomalous PV drainage but normal right PVs connect to the LA and (2) ALPV associated with mixed TAPVD in which the left PV returns separately to the right-sided circulation in the setting of TAPVD [10, 15]. Isolated ALPV can be repaired by direct anastomosis between the PV and the LA, with or without cardiopulmonary bypass. In contrast, the unique anatomy of an asymmetric or a separate PV confluence in mixed TAPVD makes direct anastomosis between the PV and the LA difficult [15, 16]. The sutureless technique was originally developed for patients with anatomic stenosis occurring after TAPVD repair [11, 17]. Our group reported the extended use of this technique to mixed TAPVD, which has so far eliminated the need for PVO-related reoperation in this anatomic subset of patients [10]. Surgical repair was done using this technique in approximately 30% of our patients and resulted in no recurrent PVO during follow-up. However, the ALPV confluence is not always located in the anterior hilum in continuity with the posterior pericardium, which is necessary to make a sutureless connection. This represents

<table>
<thead>
<tr>
<th>Pt</th>
<th>PV Anatomy</th>
<th>Extent of ALPV</th>
<th>Operation</th>
<th>PVO Severity</th>
<th>Extent of ALPV</th>
<th>Operation to PVO</th>
<th>Operation to PVO</th>
<th>Reoperation</th>
<th>Technique</th>
<th>Patch augmentation</th>
<th>Time From Operation to Reoperation</th>
<th>Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Isolated ALPV</td>
<td>Partial</td>
<td>Direct anastomosis</td>
<td>Mild</td>
<td>Left upper PV</td>
<td>1.5 y</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>2</td>
<td>Isolated ALPV</td>
<td>Partial</td>
<td>Direct anastomosis</td>
<td>Mild</td>
<td>Anastomosis</td>
<td>12 y</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>3</td>
<td>Isolated ALPV</td>
<td>Partial</td>
<td>Direct anastomosis</td>
<td>Mild</td>
<td>Left upper PV</td>
<td>7 y</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>4</td>
<td>Isolated ALPV</td>
<td>Partial</td>
<td>Direct anastomosis</td>
<td>Mild</td>
<td>Anastomosis</td>
<td>14 y</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>5</td>
<td>Mixed TAPVD</td>
<td>Total</td>
<td>Not repaired</td>
<td>Severe</td>
<td>Left upper PV</td>
<td>4 d</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>6</td>
<td>Isolated ALPV</td>
<td>Total</td>
<td>Direct anastomosis</td>
<td>Mild</td>
<td>Anastomosis</td>
<td>4 d</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>7</td>
<td>Isolated ALPV</td>
<td>Total</td>
<td>Direct anastomosis</td>
<td>Mild</td>
<td>Left lower PV</td>
<td>3 mo</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>8</td>
<td>Mixed TAPVD</td>
<td>Total</td>
<td>Direct anastomosis</td>
<td>Mild</td>
<td>Anastomosis</td>
<td>2 mo</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>9</td>
<td>Mixed TAPVD</td>
<td>Total</td>
<td>Direct anastomosis</td>
<td>Mild</td>
<td>Left lower PV</td>
<td>2 mo</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
</tbody>
</table>

ALPV = anomalous left pulmonary vein; PV = pulmonary vein; PVO = pulmonary vein obstruction; TAPVD = total anomalous pulmonary venous drainage.
an important limitation of sutureless repair for ALPV. Therefore, the anatomy should be carefully assessed by imaging, such as computed tomography or MRI, to determine the most appropriate type of surgical repair.

**Risk of PVO in Isolated ALPV: Partial vs Total ALPV**

We had speculated that partial ALPV might have a higher risk of PVO because of the smaller size of the vertical vein. In this series, however, significant PVO occurred only in patients with total ALPV. This might be related to its particular anatomy and physiology; that is, the combination of the surgically induced hairpin turn of the left PV resulting from surgical connection to the LA and the larger volume of redirected PV blood flow in total ALPV. There was no reoperation or severe PVO in 10 patients with isolated partial ALPV, implying the risk of PVO is not directly linked to the size of the vertical vein. Rather, surgically induced kinking or compression seen in total ALPV evidently has a more important effect on PVO.

**Reoperation for PVO in Patients With ALPV**

We previously reported the efficacy of sutureless repair for postrepair PVO [18]. If PVO occurs at the level of anastomosis to the LA, sutureless repair might be effective in relieving PVO, although 1 patient in this series had significant residual PVO after redo sutureless repair that led to the patient's death. Reoperation for isolated total ALPV is more technically challenging. Augmentation of the vertical vein or anastomotic site at the LAA may not effectively address the problem of kinking and compression of the left PV. One patient was saved by "rescue" sutureless repair, which was feasible because the patient had an anteriorly located PV confluence. A side-to-side anastomosis between the vertical vein and LAA as a rescue procedure might be an option for patients with selected anatomic configurations.

**Should All ALPV Be Surgically Treated?**

Surgical indications for asymptomatic patients with isolated partial or total ALPV are controversial.
Surgical repair is generally considered beneficial in asymptomatic patients when a significant left-to-right shunt (Qp/Qs of ≥2.0), PVO, and right-sided heart failure are present [6–8]. Nonetheless, unlike an atrial septal defect where Qp/Qs increases as right ventricular compliance increases [19], the amount of left-to-right shunt through the ALPV is usually fixed, which may cause less hemodynamic sequelae. Given that there is a considerable risk of early PVO after surgical repair, surgical indication and timing should be determined carefully. Serial MRI with Qp/Qs measurements are helpful for decision-making in this entity.

There is, however, little debate that surgical repair is indicated for mixed TAPVD. We propose from this experience that surgical intervention is also warranted for total ALPV as part of mixed TAPVD. Our experience with 4 such patients showed reasonable PV outcome by means of primary sutureless repair because all 4 patients are free from any PVO up to 5 years after repair. There is some controversy over partial ALPV as part of mixed TAPVD. Our strategy is to leave it untreated as long as the other 3 PVs are connected to the LA without obstruction. Repair for mixed TAPVD is typically performed in the neonatal period; therefore, a partial ALPV can be very small, posing a significant risk of PVO. This study showed that there are no significant adverse effects of untreated partial ALPV. This supports nonsurgical management of these patients.

Study Limitations
The main limitation of this study is its retrospective, nonrandomized nature. Patient numbers are relatively small. Echocardiography at follow-up was available only in 68% of patients, and thus, the data may not be completely representative of the entire cohort.

Conclusions
Reconstruction of ALPV can be done with a reasonably low occurrence of PVO and reoperation. The total form of
ALPV, in which the entire left PV drainage connects anomalously, appears to carry a higher risk of early PVO and subsequent reoperation. A reliable surgical technique to minimize PVO in this anatomic subgroup remains an area of investigation. The sutureless technique (ie, atrial-to-pericardial anastomosis) appears to be a reliable option for total ALPV as part of mixed TAPVD. Untreated partial ALPV remained unobstructed and did not cause any hemodynamic sequelae, suggesting that a nonsurgical approach is a reasonable option in this anatomic subset.

We thank Nobuko Yamamoto for her illustrations.

References


DISCUSSION

DR NAHIDH W. HASANIYA (Loma Linda, CA): Again, thank you for a very good series, though small but has lots of information. For the partial type, I understand you did direct implantation into the atrium or the other pulmonary vein?

DR KOTANI: We basically created a confluence between the left atrium and the pulmonary vein or vertical vein.

DR HASANIYA: Right. Of course these cases will need pump. We had a case last week. In fact, we did the case off-pump and anastomosed the left upper lobe to the atrial appendage. Do you have any experience with that, and is this something you recommend?

DR KOTANI: I think that previous papers have suggested a left thoracotomy approach in older children or in adult patients. In our patient group, the median age is 1 year old, which is younger relative to previous reports. In the case of small children, we routinely use cardiopulmonary bypass to make a wide anastomosis.

DR CHARLES B. HUDDESTON (St. Louis, MO): So I noticed that one of the patients in the partial veins group died early in the experience, and I am a little surprised because it is relatively benign. Was he struck by a car as he was discharged from the hospital or something?

DR KOTANI: Yes, that is a very good point. Thank you very much for that question. That patient had a rare anomaly. The patient had double-outlet right ventricle/ventricular septal defect, and only the left upper pulmonary vein was anomalous. The other pulmonary veins were normal but had severe pulmonary vein stenosis. This patient tolerated operation but developed recurrent pulmonary vein obstruction and subsequently died.

DR HUDDESTON: And my second question is that these patients will have a fixed shunt in the absence of other lesions. So you say that the indication for operation is from his right ventricular volume overload. Exactly how do you determine that? Is it with magnetic resonance imaging (MRI) or with an
echocardiogram? And if it is with an MRI, are there specific volume measurements on the MRI that you use to say this is enough that this requires operation? Thanks.

DR KOTANI: Thank you very much for your question. We had 8 patients who were untreated. Of these 8 patients, we had 3 patients who were assessed by MRI. The pulmonary-to-systemic flow (Qp/Qs) measurements for those patients ranged from 1.3 to 1.7. Usually, patients are assessed by echocardiography, but these were special cases for which we used MRI. As you know, it is difficult to determine right ventricular (RV) dimensions using a quantitative approach. As such, I don’t think that we have any specific cutoff for RV volume. Rather, it should be followed as it is for patients with atrial septal defects.

DR CARL LEWIS BACKER (Chicago, IL): Can you please back your slides up to that beautiful anatomic one where you showed all the different variations?

We had a recent patient who had what I would call “dual drainage” of the pulmonary vein. The patient had a coarctation of the aorta and also had partial drainage of the left upper pulmonary vein to the innominate vein. MRI showed there was also a dual connection down to the left atrium. I simply ligated the left vertical vein, and the echo post-op looked totally normal. Did you have any patients like this in your series where there was dual drainage of the anomalous pulmonary venous connection?

DR KOTANI: We didn’t have that particular case in our series. I think in small patients who have only one anomalous left pulmonary vein, it may be best to leave it alone because there is a high risk for late stenosis even if it is repaired. Outcomes may be similar if you leave it untreated.

DR S. ADIL HUSAIN (San Antonio, TX): I am going to follow up on Dr Huddleston’s question. Are there any patients in the series who received operations that initially were thought to be ones you were just going to follow long-term and then they ended up going into the surgical arm for X reason? And if there are, what was X reason?

DR KOTANI: Right. In our series, of the 8 patients who were untreated, 1 patient developed very minor pulmonary vein obstruction at 15 years after the initial diagnosis. It may be important to note that the patient had a left upper pulmonary vein going into the inferior vena cava, so it is a kind of intra-cardiac type. That particular pulmonary vein anatomy might be related to a risk of late pulmonary vein obstruction, but I am not sure.

DR BACKER: I want to poll the audience. This is a somewhat common problem that we are presented with. That is the patient who has an isolated left upper pulmonary vein that is unobstructed and draining to the innominate vein. I agree with you about looking for right ventricular volume overload. Certainly some of these patients have a Qp/Qs that is more than 1.5 to 1. If you have a patient who has isolated left upper pulmonary vein drainage and it is from half of the left lung, how many people would electively recommend repairing that?

(Audience responds.)

DR BACKER: All right. And how many people would leave it alone?

(Audience responds.)

DR BACKER: It looks like we are split about 50/50. So, Chris Caldarone, come up to the microphone and tell us what the answer is! I mean, as a Society, we should have some sort of unified answer to this basic question.

DR CALDARONE: All right. Lock the doors. Nobody leaves until we decide. Perhaps we could argue one point and the other. I’ll argue for nonoperation, and someone else can argue for operation.

If you have a left upper pulmonary vein draining anomalously to the innominate vein, it is unobstructed. There is no other residual shunt, no other residual intracardiac shunt, that patient may have a left-to-right shunt of 1.3, 1.4 probably at most, and it is never going to change with time. That is quite different than an atrial septal defect that will increase its shunt with time. So I think the shunt is controlled. It is never going to change. We have evidence out for more than a decade that it is unlikely to become stenotic. I would say just follow the patient, and if it develops stenosis, then decompress it.

DR BACKER: All right. There’s your answer.

DR CALDARONE: Someone’s got to argue it the other way.

DR BACKER: This should be a good debate for next year. We’ll have to put it on the agenda.

DR PHILIP C. SMITH (Akron, OH): What if the technique for reimplanting the vein had a stenosis rate that was 3% instead of 60%? I would advocate for an operative approach that leaves the vein in its native position with minimal dissection, performing wide a side-to-side anastomosis with the atrial appendage. I have two of those patients, and they have been followed out for 3 and 5 years, and they haven’t had a recurrence of any stenosis. So is it a shortcoming of the technique that you are using for doing the reimplantation?

DR CALDARONE: Even if the risk of restenosis was zero, you are still doing a prophylactic operation to prevent a problem in the future. So the burden of proof is on the person who wants to do a prophylactic procedure to show that the treatment is better than the disease that would have occurred otherwise. I don’t think we have any evidence at all to say that an isolated left upper pulmonary vein draining without obstruction ever causes any morbidity or certainly it doesn’t shorten someone’s longevity. So even if there were a zero risk of pulmonary vein stenosis after your repair, what rationale would you have to do an operation solely for that?

DR BACKER: Chris, let me ask a theoretical question. As the left ventricular compliance goes up as the patient ages, can there be venovenous collaterals that develop from the lower left vein to the left upper vein? The final end point for the venous drainage of that left upper vein is the lower resistance circuit with the lower right ventricular compliance: could that be a potential cause of increased intrapulmonary venovenous shunting over time, or do you believe that does not happen, or do we not know? Chuck Huddleston is going to answer this one.

DR HUDDLESTON: Well, I was just imagining how you came up with that scenario. I have actually operated on 2 patients who developed symptoms from this particular anomaly. Both of them were these very enthusiastic joggers, and actually both of them were physicians, oddly. And this was the only thing that was found to explain why they couldn’t seem to continue their training to keep up with their peers. Both of them improved their jogging times after it was repaired. So that is my experience with that.
DR CALDARONE: Were they partial or complete?

DR HUDDLESTON: Yes, they were. That’s it. All they had was the left upper lobe, 1.2 to 1 shunt in both cases cath.

DR S. HUSAIN: I just had one tiny experience to add to what was being discussed. I think that it is possible for there to be an increase in the prevalence of flow through the anomalous vein over time. It is probably because of all the things that Chris hypothesized. And I say that because there was 1 patient that we followed over an interval of 2 years and measured an increased right ventricular end diastolic volume index in that patient. So we felt there was some evidence of progression of dilatation of the right ventricle. With no other sources of left-to-right shunting, we blame that on the vein and did go ahead with a repair of that solitary vein. So I think it is possible for it to change over time, and MRI-derived right ventricular dilation would be a way to follow that.

DR CALDARONE: I was just going to say, Carl, congratulations for stirring up a great discussion, but we should give Dr Kotani the last word.

DR JOSEPH J. AMATO (Chicago, IL): As living evidence of person with a left upper lobe vein anomaly, I was just diagnosed at the age of 75, about a month ago, of having a left upper pulmonary vein going through my innominate vein to the right superior vena cava and finally into my right atrium. A cardiac catheterization was performed because of my severe mitral valve stenosis. The anomalous pulmonary vein was visualized and the shunt measured at 1.4 Qp:Qs blood flow. The literature states that surgical correction of this anomaly alone should be considered if the blood flow ratio exceeded 1.5. So with concomitant disease like mitral valve stenosis, the question is, what do you do? And I don’t want an answer. I just wanted to relate this personal condition for a possible addition to your impending publication of these anomalies. Thank you.

DR BACKER: Dr Kotani, give us the final word, give us some words of advice to take home.

DR KOTANI: Thank you very much for all your comments. From our discussion today, it is evident that the data do not point definitively one way or the other in the management of these patients. We will need to wait for further reports and more long-term data. As the evidence accumulates, we may be able to reach a more definitive conclusion. Thank you. I am very happy to have had such a nice discussion about this topic.