The beneficial hemodynamic effects of selective patent vertical vein following repair of obstructed total anomalous pulmonary venous drainage in infants

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Abstract

Objectives: Postoperative low cardiac output may persist after repair of total anomalous pulmonary venous drainage (TAPVD) because of a relatively small and non-compliant left atrium and left ventricle. We examined the effects of selective vertical vein patency on postoperative hemodynamics.

Methods: Thirty-four patients less than 3 months of age with TAPVD were operated from July 1993 to June 2000. The mean age at operation was 21 ± 8 days (range, 3–62 days) and the mean weight was 3 ± 0.2 kg (range, 2–4.1 kg). Supracardiac type drainage was found in 12 (35%), cardiac in three (9%), mixed in one (3%), and infracardiac in 18 (53%) patients. Twenty-two patients (65%) had obstructed venous drainage. All operations were performed with deep hypothermic circulatory arrest. Supracardiac, mixed and infracardiac types were repaired through a posterior approach, whereas, in the cardiac type, the coronary sinus was unroofed and the atrial septal defect was patched. The decision whether to keep the vertical vein open was made at the end of the operation and was based on the hemodynamic state of the patient.

Results: There were no operative deaths. The suture on the vertical vein was released in 22 patients who had obstructed pulmonary venous drainage (infracardiac type, n = 18; supracardiac type, n = 3; and mixed type, n = 1), resulting in a significant drop in the left atrial pressure from 19 ± 2 to 12 ± 2 mmHg (P < 0.05), and in the mean pulmonary artery pressure from 42 ± 6 to 35 ± 3 mmHg (P < 0.05), associated with an immediate increase in the mean arterial blood pressure from a mean of 46 ± 3 to 60 ± 4 mmHg (P < 0.05). During a mean follow-up of 38 ± 6 months (range, 8–71 months), there were no late deaths. Follow-up, two-dimensional echocardiography with Doppler studies demonstrated good left ventricular function and trivial or no left to right shunt through the vertical vein in those patients in whom the snare was released. Conclusions: Maintaining the vertical vein patent in a selective group of patients with infracardiac total anomalous venous drainage contributes to a favorable outcome following surgery.

Keywords: Vertical vein; Obstructed pulmonary venous drainage; Infants

1. Introduction

The results of repair of total anomalous pulmonary venous drainage (TAPVD) have improved over recent years, and the surgical mortality currently reported in patients less than 3 months of age ranges from 0 to 5% [1,2]. However, postoperative low cardiac output may persist because of a small and non-compliant left atrium and ventricle or recurrent episodes of pulmonary hypertension.

In order to allow the left heart to adapt and maintain adequate cardiac output, various surgical techniques have been employed in order to construct an unrestricted connection between the pulmonary venous confluence and the left atrium [3,4]. Ligation or interruption of the vertical vein and incorporation of the vertical vein as part of the new anastomosis have been described [5]. Some authors [17] have been concerned about the occurrence of acute liver necrosis after surgery with ligation of the vertical vein. The technical modifications have been directed at reducing the recurrence of pulmonary vein stenosis. However, they do not address the problems of a relatively small, non-compliant left atrium and ventricle, which may lead to the development of low cardiac output immediately after surgery.

The purpose of this study is to present our experience with selective patency of the vertical vein with particular emphasis on the hemodynamic effects following repair of TAPVD.
2. Materials and methods

Between July 1993 and June 2000, 34 consecutive patients with isolated TAPVD were operated at the Soroka Medical Center in Beer Sheba, Israel, and Children’s Hospital in New Orleans, LA, by one surgeon (J.C.). The hospital records, preoperative and postoperative echocardiograms, and cardiac catheterization data were retrospectively reviewed. Patients with associated cardiac lesions were excluded from this study. The surgical techniques and postoperative management protocols were identical in both institutions. Age at the time of the repair ranged from 3 to 62 days (mean, 21 ± 8 days) and weight ranged from 2 to 4.1 kg (mean, 3 ± 0.2 kg). Twenty-six patients required intubations and mechanical ventilation prior to surgery for respiratory distress and 21 patients required inotropic support for hemodynamic instability. Cardiac catheterization was performed in eight patients in whom accurate anatomical pattern of the pulmonary venous return could not be established by echocardiography. Supracardiac drainage was found in 12 (35%), cardiac drainage into the coronary sinus in three (9%), and infracardiac drainage in 18 (53%) patients. A combination of supra- and infracardiac drainage was found in one (3%) patient. Twenty-two (65%) patients had obstructed pulmonary venous drainage (Table 1). The ductus arteriosus was patent in 29 patients. The pulmonary artery pressure was measured in all patients prior to the institution of cardiopulmonary bypass. Pulmonary hypertension was defined as a right ventricular/left ventricular pressure ratio of > 0.6.

2.1. Operative technique

The operation was performed under profound hypothermic circulatory arrest at a rectal temperature of 18°C. The cooling time to 18°C ranged from 16 to 24 min (mean, 18 ± 1.5 min) and the circulatory arrest time ranged from 32 to 47 min (mean, 38 ± 4 min). Multi-dose blood cardioplegia was used for myocardial protection and started with the initiation of circulatory arrest. For cardiac drainage, the approach was through the right atrium as described by Malm [6]. The coronary sinus was unroofed and the opening was performed in eight patients in whom accurate anatomical pattern of the pulmonary venous return could not be established by echocardiography. Supracardiac drainage was found in 12 (35%), cardiac drainage into the coronary sinus in three (9%), and infracardiac drainage in 18 (53%) patients. A combination of supra- and infracardiac drainage was found in one (3%) patient. Twenty-two (65%) patients had obstructed pulmonary venous drainage (Table 1).

Table 1

<table>
<thead>
<tr>
<th>Type of Pulmonary Vein Drainage</th>
<th>Obstructed (%)</th>
<th>Non-obstructed (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supracardiac (n = 12)</td>
<td>3 (25)</td>
<td>9 (75)</td>
</tr>
<tr>
<td>Cardiac (n = 3)</td>
<td></td>
<td>3 (100)</td>
</tr>
<tr>
<td>Infracardiac (n = 18)</td>
<td>18 (100)</td>
<td></td>
</tr>
<tr>
<td>Mixed (n = 1)</td>
<td>1 (100)</td>
<td></td>
</tr>
<tr>
<td>Total number (n = 34)</td>
<td>22 (65)</td>
<td>12 (35)</td>
</tr>
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</table>

For the infracardiac type, the apex of the heart was dislocated to the right side and superiorly, thus exposing the pulmonary venous confluence. The repair was carried out by creating a wide anastomosis between the common pulmonary vein, with the incision being extended to the upper right pulmonary vein and the left atrium after it was opened longitudinally. The atrial septal defect was closed via a right atriotomy without leaving a fenestration. In the mixed drainage type, repair was accomplished by combination of transatrial and apical approaches. After completion of the repair, a 5/0 Prolene suture was placed around the draining vein at the junction with the innominate vein for the supracardiac drainage or below the lower pulmonary veins and just above the diaphragm for the infracardiac drainage. The suture was passed through a snare of an 8 French polyethylene tube. The suture was fixed in position with a series of hemoclips placed on the suture and the polyethylene tube.

Left atrial and pulmonary artery pressures were monitored continuously after the operation. All patients were sedated and paralyzed during the first 24 h following surgery. Pulmonary hypertension was treated with hyperventilation, sedation and inhaled nitric oxide as required.

2.2. Statistical analysis

All values are reported as means and standard error of the mean. Numerical and categorical data were analyzed by the Student’s t-test and the Chi-square test, respectively. For all analyses, a P value of less than 0.05 was considered significant.

3. Results

There were no operative deaths (<30 days). Table 2 shows the hemodynamic data in patients with and without obstructed venous drainage immediately after surgery. In patients with unobstructed venous drainage (supracardiac, n = 9; cardiac, n = 3), there was no hemodynamic instability and the vertical vein was permanently ligated without untoward effects. However, each of the patients with preoperative obstructed drainage had varying degrees of hemodynamic deterioration following repair. Loosening of the
Table 2
Comparison of the hemodynamic data after surgery between patients with obstructed vs. unobstructed venous drainage*

<table>
<thead>
<tr>
<th></th>
<th>Unobstructed</th>
<th>Obstructed</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>MAP (mmHg)</td>
<td>66 ± 4</td>
<td>46 ± 3</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>LAP (mmHg)</td>
<td>12 ± 2</td>
<td>19 ± 2</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>MPAP (mmHg)</td>
<td>26 ± 3</td>
<td>41 ± 3</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Arterial PO2 (mmHg)</td>
<td>211 ± 20</td>
<td>78 ± 15</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Arterial blood pH</td>
<td>7.38 ± 0.03</td>
<td>7.28 ± 0.02</td>
<td>&lt; 0.05</td>
</tr>
</tbody>
</table>

* MAP, mean arterial blood pressure; LAP, left atrial pressure; MPAP, mean pulmonary artery pressure; PO2, partial pressure of oxygen.

snare resulted in a significant decrease (P < 0.05) of the left atrial pressure in 22 patients; infracardiac (n = 18), supracardiac (n = 3) and mixed (n = 1). The decrease of left atrial pressure was associated with a significant increase (P < 0.05) in the mean arterial blood pressure and correction of metabolic acidosis (Table 3).

Pulmonary hypertension after surgery was more common in patients with preoperative obstructed pulmonary venous drainage: 22 patients compared with two patients without preoperative obstructed veins (P < 0.01). A pulmonary artery to systemic blood pressure ratio of > 0.6 remained in nine patients with the infracardiac type, and two with the mixed type, 12 h postoperatively and decreased significantly (P < 0.05) to a ratio of < 0.6 during a mean time of 28 ± 9 h (range, 6–54 h). Forty-eight hours after surgery, the vertical vein remained open in all patients with the infracardiac type (n = 18) following repeated attempts to snare it. The draining vein into the superior vena cava was tightened in the intensive care unit in four patients (supracardiac type, n = 3; and mixed type, n = 1) 6, 10, 12 and 18 h after surgery with no subsequent deterioration in their hemodynamic state. Inotropic support with dopamine (>3 g/kg per min) was required in 26 patients for a mean duration of 4.5 ± 1 days (range, 2–7 days) after surgery. The mean duration of mechanical ventilation was 5.2 ± 2 days (range, 2–16 days). Nitric oxide inhalation therapy was used in 15 patients for a mean duration of 32 ± 11 h (range, 12–58 h). Twelve patients (35%) underwent delayed sternal closure at a mean time of 31 ± 7 h (range, 18–52 h).

Follow-up was complete and ranged from 8 to 71 months (mean, 38 ± 6 months). There were no late deaths. Pulmonary vein stenosis occurred in one patient with mixed drainage type at 3 months after the first operation necessitating re-operation with patch augmentation of the left pulmonary veins–left atrial anastomosis. This patient is currently free of symptoms. All patients are in New York Heart Association functional class 1 without any medications. Serial two-dimensional echocardiography with Doppler color flow studies demonstrated a gradual decrease of the blood flow through the vertical vein and no pressure gradient across the pulmonary venous confluence–left atrial anastomosis. None of the patients required late reoperation as a result of left to right shunt through the vertical vein. No neurological abnormalities have occurred in this series either immediately after the operation or during the follow-up period.

4. Comment

The operative outcome after repair of TAPVD in infants has improved significantly in recent years with a reported mortality of less than 5% [1,2,4]. Increased mortality has been previously correlated with the preoperative clinical and hemodynamic parameters, such as the need for endotracheal intubation, type of anatomic connection, gender, age at operation, the size of the intraatrial communication, pulmonary venous obstruction, pulmonary hypertension, elevated pulmonary vascular resistance and left heart volumes [3,4]. Preoperative resuscitation, better perioperative care, improved surgical techniques, and early diagnosis by echocardiography and Doppler color flow studies without the need for cardiac catheterization have contributed to better results [14]. Data from this and other recent studies [8–11] have shown that the type of pulmonary venous connection, younger age (<1 month), or the need for preoperative mechanical ventilation have not resulted in increased surgical death.

Various studies have related the improved results to more precise surgical techniques aimed at achieving structural alignment and augmentation of the left atrial cavity, including the use of the vertical vein as a patch [4,5]. Other authors [3] advocated the use of absorbable suture material and interrupted fine sutures in order to minimize the possibility of early anastomotic narrowing and to allow future growth. A continuous suture technique has been our standard method of performing the anastomosis between the pulmonary venous confluence and the left atrium and we have not seen an increased incidence of early or late anastomotic constriction.

Previous studies [12,13] have demonstrated that in patients with TAPVD, the left atrial volume is smaller and its compliance is diminished compared with normal hearts. In the presence of pulmonary venous obstruction, the left atrial chamber size is even smaller than in non-obstructed cases [15]. A controversy still exists as to whether the small size of the left atrium is a determinant of operative outcome and if it is necessary to augment the anastomosis and close the atrial septal defect with a patch [3]. We believe that the use of a redundant pericardial patch to close the atrial

Table 3
Postoperative hemodynamic data with the vertical vein closed and patent*

<table>
<thead>
<tr>
<th></th>
<th>Closed</th>
<th>Patent</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>MAP (mmHg)</td>
<td>46 ± 3</td>
<td>60 ± 4</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>LAP (mmHg)</td>
<td>19 ± 2</td>
<td>12 ± 2</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>MPAP (mmHg)</td>
<td>42 ± 3</td>
<td>35 ± 5</td>
<td>0.06</td>
</tr>
<tr>
<td>Arterial blood pH</td>
<td>7.28 ± 0.02</td>
<td>7.4 ± 0.04</td>
<td>&lt; 0.05</td>
</tr>
</tbody>
</table>

* MAP, mean arterial blood pressure; LAP, mean left atrial pressure; MPAP, mean pulmonary artery blood pressure.
septal defect is an important addition to the operation, which serves to augment the left atrial chamber size. Despite all these modified techniques aimed at improving the dimension of the left atrial cavity, low cardiac output has still remained a significant problem leading to hemodynamic instability and morbidity after surgery [11]. The presence of a non-compliant left atrium and ventricle reflected as high left atrial pressure has been documented in this study and may explain the development of low cardiac output after surgery. Although ligation of the vertical vein is controversial, we concur with other studies [16–18] that have shown improved surgical results when the vertical vein was left open. Based on our initial experience [19], leaving the vertical vein patent has been shown to reverse the hemodynamic deterioration and to help maintain a more stable postoperative course. In this study, the vertical vein was left open in all patients with the infracardiac drainage type with no further attempts to ligate it after the immediate postoperative period. Leaving a calibrated fenestration of the atrial septal defect patch may also help to decompress the small left atrium after repair, however, we have no experience with this approach. In accordance with other reports [17,20], this study does not show a higher operative mortality in regard to the type of anomalous drainage, particularly the infracardiac type. However, it is apparent that the incidence of low cardiac output and pulmonary hypertension after surgery is higher in this group and is most likely to be related to the presence of obstructed pulmonary veins preoperatively. The site of the venous obstruction can occur at the diaphragm level or at any level along the tortuous pathway of the blood flow through the portal system because of the high resistance of the hepatic capillary bed. It is conceivable that there would be a preferential blood flow from the pulmonary veins to the left atrium in the presence of a wide anastomosis between these structures. The small residual shunt through the vertical vein would help as a temporary vent of the small and non-compliant left heart during the early postoperative phase and allow accommodation to the increased forward blood flow. Evidently, as a result of the obstructed intra-abdominal venous pathway, there is a significant reduction in the magnitude, and eventually, complete obliteration of the left to right shunt through the vertical vein. Serial postoperative echocardiographic studies have shown, in this particular group of patients, a significant reduction of residual blood flow across the diaphragm within a short time span after the operation. Similarly, the notion that the left atrial and venricular chambers grow and become more compliant with time after successful repair has been supported by other studies [3].

Postoperative pulmonary hypertension is a major determinant of early postoperative mortality. It is noteworthy that this may occur in the absence of anatomic venous obstruction [4]. The mechanism of this condition remains unclear, however, in addition to intimal fibrous proliferation, the media of the small pulmonary arteries and veins of patients with TAPVD are unusually thick. In our series, postoperative pulmonary hypertension was more common in patients with infracardiac type drainage and was probably related to a higher prevalence of preoperative pulmonary venous obstruction.

Further study would be required to compare two groups of patients with and without patent vertical vein in regard to the duration of pulmonary hypertension, duration of ventilator and inotropic support after surgery.

In spite of the improved hemodynamics associated with selective vertical vein patency, it is clear that the postoperative course of patients with obstructed TAPVD can be complex with other appreciable complications.

We believe that the employment of this modification in the surgical management of patients with preoperative obstructed TAPVD has favorable effects on the morbidity and outcome after surgery.

References

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