

Off-pump hepatic to azygos connection via thoracotomy for relief of fistulas after a Kawashima procedure: Ten-year results

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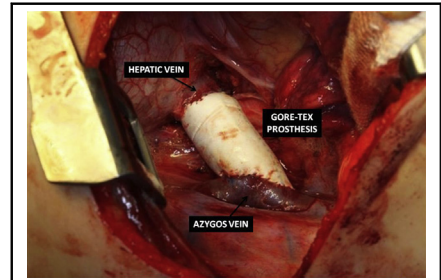
ABSTRACT

Objectives: An almost universal incidence of developing pulmonary arteriovenous fistulas after the Kawashima operation has been reported. Exclusion of the hepatic venous flow from the pulmonary circulation causes the development of these malformations. Redirection of hepatic venous flow to the pulmonary circulation mostly leads to the regression of the arteriovenous fistulas.

Methods: We analyzed 11 patients with arteriovenous fistulas that developed after the Kawashima operation. The hepatic-to-azygos shunts were performed with an off-pump technique through a lateral thoracotomy in all but one. Operative and postoperative data were retrospectively collected.

Results: No intraoperative complications occurred, and no patient died in the hospital. Up to 10-year follow-up showed a significant postoperative improvement of patients' oxygen saturation and New York Heart Association class. Apart from 2 re-thoracotomies for bleeding in 1 patient, no complications occurred and no patient died during follow-up. Two other patients underwent reoperation for an undiagnosed additional hepatic vein. The improvement of patients' oxygen saturation and New York Heart Association class persisted during the follow-up period.

Conclusions: The surgical connection can be performed safely with an off-pump technique that avoids the risks related to extracorporeal circulation and circulatory arrest. The results at 10 years follow-up confirmed the efficacy and safety of the surgical technique described. (*J Thorac Cardiovasc Surg* 2015;149:1524-30)



HAS via lateral thoracotomy.

Central Message

The HAS by interposition of a vascular prosthesis can be performed safely with an off-pump technique through a lateral thoracotomy. The redirection of hepatic venous flow to the pulmonary circulation leads to the regression of arteriovenous fistulas.

Perspective

The redirection of the hepatic blood to the pulmonary bed can be performed with different techniques and approaches. We describe the HAS by using a polytetrafluoroethylene (Gore-Tex; WL Gore & Associates Inc, Flagstaff, Ariz) vascular prosthesis performed with an off-pump technique through a lateral thoracotomy. The described technique is safe and has excellent clinical results at short- and medium-term follow-ups. This technique avoids the risks of repeat sternotomy and the possible complications related to the cardiopulmonary bypass. Moreover, the extracardiac connection provides adequate and balanced flow distribution of the hepatic blood to both lungs.

See Editorial Commentary page 1530.

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In 1984, Kawashima and colleagues¹ described a quasi-total cavopulmonary connection in patients with univentricular atrioventricular connection, interrupted inferior caval vein, and azygos or hemiazygos continuation where the superior caval vein was connected to the right pulmonary artery, leaving the hepatic veins connected to the atrium. Kawashima and colleagues¹ published their surgical technique 4 years before de Leval and colleagues² first publication on the total cavopulmonary connection. The “Kawashima operation” is currently a recognized surgical technique to achieve a quasi-complete cavopulmonary

Abbreviations and Acronyms

CT	= computed tomography
HAS	= hepatic to azygos shunt
ICU	= intensive care unit
NYHA	= New York Heart Association

connection in cases of interruption of the inferior caval vein, which usually occurs in the context of left atrial isomerism.^{3,4}

However, the medium- and long-term follow-ups after the Kawashima operation show a high incidence of gradually reducing systemic oxygen saturation due to the formation of pulmonary arteriovenous fistulas.⁵⁻⁷ It was suggested that the exclusion of hepatic venous blood flow from the pulmonary circulation caused the development of arteriovenous fistulas.⁵⁻⁸ The reversibility of these arteriovenous fistulas after redirection of hepatic venous blood flow to the pulmonary circulation supported the concept that a lack of a hitherto unidentified hepatic factor in the pulmonary blood causes the development of the fistulas.⁷⁻¹⁰ Different techniques have been described to redirect the hepatic flow to the pulmonary circulation, and we described the current technique in 2008.^{9,11-14} In our study, we describe an off-pump technique to make a connection between the hepatic vein and the azygos or hemiazygos vein using a vascular prosthesis through a lateral thoracotomy. We report on the outcomes up to 10 years of 11 patients in whom this technique was used to redirect hepatic flow to the lungs.

MATERIAL AND METHODS

Study Design: Patient Selection

The Institutional Review Boards of the University Medical Center Groningen and Rikshospitalet Oslo University Hospital agreed to waive the need for an informed consent because data were collected as part of routine medical care and patients were not individually identifiable.

We selected from both institutional databases those patients who underwent a surgical connection of the hepatic to the azygos vein, hepatic to azygos shunt (HAS), because of the development of arteriovenous fistulas after a Kawashima operation. The operations were performed between June 2002 and August 2011; thus, the maximum follow-up is 10 years.

Inclusion criteria were a prior Kawashima operation with secondary hypoxia due to pulmonary arteriovenous fistulas. Twelve patients satisfied the inclusion criteria. One patient was excluded because of refusal until now of the surgical procedure. The presence of arteriovenous fistulas was suspected by a progressive decrease of oxygen saturation by pulse oximetry during follow-up compared with the oxygen saturation recorded immediately after the Kawashima operation. Three patients had similar oxygen saturation levels before HAS compared with the time of their Kawashima operation. However, these patients showed poor clinical condition and cyanosis at exercise. The presence of pulmonary arteriovenous fistulas demonstrated by contrast echocardiography and cardiac catheterization determined the indication to proceed with the HAS procedure.

Two-dimensional contrast echocardiography was used in all patients to confirm the presence of pulmonary arteriovenous fistulas. This was performed by injection of agitated saline solution through a peripheral vein with simultaneous echocardiographic imaging of the left atrium. Pulmonary arteriovenous fistulas were considered present when contrast could be visualized in the left atrium with a short delay (3-5 cardiac cycles or 2-4 seconds) after contrast injection. In 9 patients, in addition to contrast echocardiography, cardiac catheterization was performed to visualize the fistulas and reduced oxygenation of pulmonary venous blood established.

A computed tomography (CT) angiography examination was performed to define the anatomy with particular focus on the connection between the hepatic veins and the atrium. Contrast was injected in an antecubital vein, and after 40 seconds the scan was started so optimal hepatic vein enhancement was obtained. The CT data were reconstructed on a 3-dimensional workstation by using a multiplanar reformat and volume-rendering technique.

Data Collection and Patients' Characteristics

Preoperative, operative, and postoperative data from all 11 patients were collected retrospectively. Baseline patient characteristics are listed in [Table 1](#).

Surgical Technique

Ten HAS procedures were performed through a thoracotomy, and 1 HAS procedure was performed through a median re-sternotomy. The surgical approach was planned on the basis of the CT scan findings, where the thoracotomy was done on the same side as the major inferior systemic vein, thus, right in case of an azygos continuation and left in case of a hemiazygos continuation. The HAS procedures were performed through a left thoracotomy in 7 patients with a hemiazygos continuation and through a right thoracotomy in 3 patients with an azygos continuation. All operations were performed off pump, as previously described.⁹ Through a thoracotomy at the level of the seventh or eighth intercostal space, the hepatic vein and the azygos or hemiazygos vein could be identified easily. A polytetrafluoroethylene (Gore-Tex; WL Gore & Associates Inc, Flagstaff, Ariz) vascular prosthesis was used to make the connection. The diameter of the vascular prosthesis varied between 10 and 18 mm. The choice of the polytetrafluoroethylene (Gore-Tex) vascular prosthesis diameter depends on the diameter of the hepatic vein and the azygos or hemiazygos vein: The prosthesis diameter should be proportional to both veins.

The hepatic vein was partially closed by using a side-biting vascular clamp. An end-to-side anastomosis was performed between the polytetrafluoroethylene (Gore-Tex) vascular prosthesis and the hepatic vein. Afterward, an end-to-side anastomosis was performed between the polytetrafluoroethylene (Gore-Tex) vascular prosthesis and the azygos or hemiazygos vein by partial closure of the azygos or hemiazygos vein, again with a side-biting vascular clamp. After de-airing, the shunt was opened and started functioning ([Figure 1](#)).

The connection of the hepatic veins to the atrium was always closed by using a suture or staples. A division of the hepatic vein could be performed where there was enough space to place 2 vascular clamps or to use a combined stapler/cutting device. The division was performed in 2 patients.

All patients received postoperative acetylsalicylic acid at a dosage of 100 mg daily per os in adult patients and at a weight-based dosage in pediatric patients. In the presence of atrial arrhythmia, previous thromboembolic events, and thrombophilia, such as factor V Leiden or protein C/S deficiency, patients received vitamin K antagonist instead of acetylsalicylic acid. The international normalized ratio target was between 2 and 3. It was recommended to continue lifelong antithrombotic or anticoagulation therapy. At follow-up, all patients were still receiving antithrombotic or anticoagulation treatment.

Follow-up

The clinical follow-up was performed at regular outpatient visits. Physical examination, echocardiographic evaluation, laboratory tests, measurement of oxygen saturation by pulse oximetry, and evaluation of New York Heart Association (NYHA) class were routinely performed in all patients.

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