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Hepatic to azygos connection by autologous pericardial roll for Fontan completion

Tao Shi, Zhan Gao and Zhongdong Hua

Pediatric Cardiac Surgery Center, Fuwai Hospital, CAMS and PUMC, National Center for Cardiovascular Diseases, Beijing, China

Brief Report

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Author for correspondence:
Zhongdong Hua, Ward 3 of Pediatric Cardiac Surgery Center, Fuwai Hospital, No. 167 North Lishi Road, Xicheng District, Beijing 100037, China. Tel: +86(10)88396555.
E-mail: clusterheadache@126.com

Abstract

Techniques for performing the Fontan completion procedure after Kawashima procedure have developed in recent years. This paper presents a case in which we performed an off-pump hepatic-to-azygos connection by autologous pericardial roll via thoracotomy as a modified Fontan completion. The operation wound was limited, and the short-term follow-up was satisfactory. Notably, it was found that the autologous pericardium might require a shorter period of anticoagulation treatment than what is needed for artificial prosthetic materials.

Fontan completion, resulting in the drainage of the hepatic venous blood flow in the pulmonary circulation, induces the regression of the pulmonary arteriovenous malformations after Kawashima procedure.[1] Apart from the widely used extracardiac conduit technique, the hepatic-to-azygos connection is one of the optional techniques for Fontan completion after Kawashima procedure. This paper presents a case in which we performed an off-pump hepatic-to-azygos connection by autologous pericardial roll via thoracotomy as a modified Fontan completion.

Case presentation

A 6-year-old boy presented for his third heart operation. At birth, he displayed severe cyanosis and intermittent hypoxic spells, and he was diagnosed with dextrocardia, a single ventricle, pulmonary atresia, and an interrupted inferior caval vein. He had received a modified Blalock–Taussig shunt and a bidirectional Glenn procedure (also called Kawashima procedure) at 9 months and 3 years of age, respectively. His oxygen saturation had increased to 88% after the second operation, and he became almost asymptomatic afterward. A pre-operative CT scan showed the absence of the hepatic segment of the inferior caval vein with azygos continuation, and the hepatic vein drained directly into the atrium (Fig 1). The McGoon ratio was 2.1, and the Nakata index was 227.4 mm^2/m^2. The mean pulmonary artery pressure was 9 mmHg, and no pulmonary arteriovenous malformations were noticed by catheter angiography. For his third operation, we planned to perform a Fontan completion procedure. A right posterolateral thoracotomy at the level of the seventh intercostal space was completed under general anaesthesia. The hepatic vein and the enlarged azygos vein (diameter ~12 mm) were identified after releasing the inferior pulmonary ligament. The azygos vein ran along the spine, and the hepatic vein drained directly into the atrium. We planned to make a pedicled pericardial tunnel, but the available pericardium was quite limited. The autologous pericardium of a size 20 × 50 mm was carefully dissected from the atrium, and its adhesions were carefully removed to maintain a smooth inside surface. The piece of the pericardium was then formed into a roll (diameter ~16 mm) for the hepatic-to-azygos connection. The hepatic vein's pressure rose to 22 mmHg when its proximal section was clamped with forceps, which might exclude the possibility of undiagnosed branches. End-to-side anastomosis was then performed between the pericardial roll and the hepatic vein with a continuous suture using 6-0 Prolene, and in turn the pericardial roll and the azygos vein. After de-airing, the shunt was opened, and it began functioning. The connection of the hepatic vein and the atrium was then cut and sutured. No intra-operative and post-operative complications occurred. A post-operative CT scan showed the hepatic-to-azygos connection (Fig 2). The oxygen saturation had increased to 100% at discharge. This patient was advised to take warfarin daily for at least 6 months, and the target of the international normalised ratio was set at 1.8–2.5. The velocity of the azygos vein was 27 cm/s on discharge and 33 cm/s at the 3-month follow-up.

Discussion

In 2008, Lopez et al[2] performed the first off-pump hepatic-to-azygos connection via thoracotomy to resolve pulmonary arteriovenous malformations by using a polytetrafluoroethylene vascular prosthesis for the connection. It significantly reduced the complications associated with

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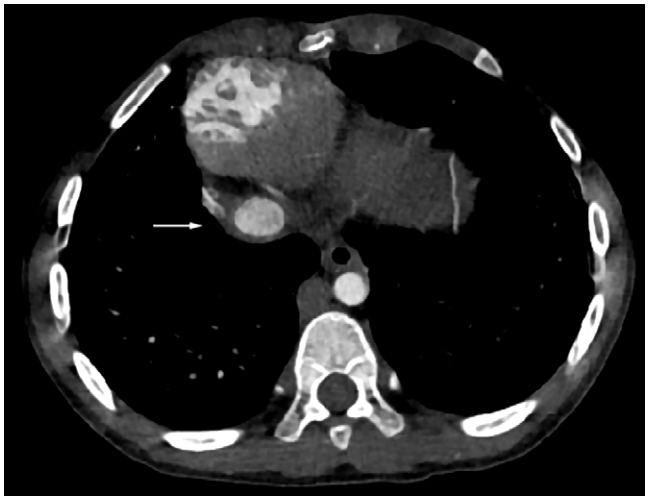

Figure 1. Pre-operative CT scan showing the hepatic vein (arrow) drained directly into the atrium, the enlarged azygos vein parallel to the aorta.

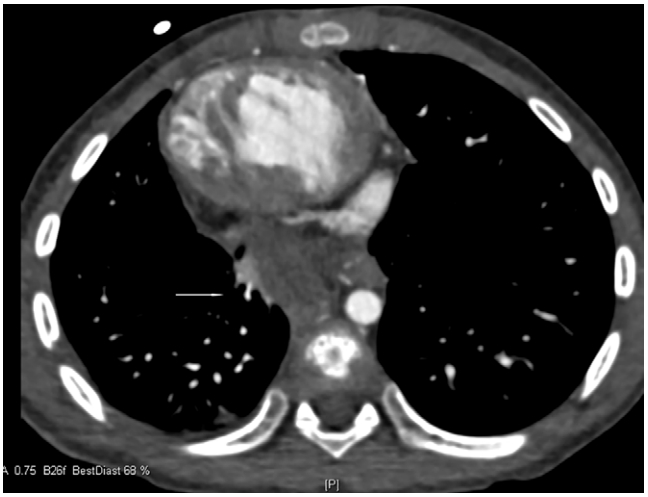

Figure 2. Post-operative CT scan showing the hepatic-to-azygos connection (arrow).

re-sternotomy and cardiopulmonary bypass compared with the traditional extracardiac conduit technique. McElhinney et al[3] argued that a direct hepatic-to-azygos connection might provide more reliable mixing and a bilateral distribution of hepatic venous blood, while avoiding the formation of unilateral pulmonary arteriovenous malformations. A 10-year follow-up study confirmed the approach's efficacy and safety.[4]

Prosthetic material is commonly used for making the hepatic-to-azygos connection, such as polytetrafluoroethylene, with consequent lifelong antithrombotic treatment required. Reports have surfaced that favour the pedicled autologous pericardial roll as an ideal option for an extracardiac conduit.[5,6] In vivo, the conduit has the potential to grow in a parallel manner with somatic development.[5] Immunohistologic examination showed the presence of endothelium and the absence of calcification and fibrosis 9 years after a pedicled autologous pericardium implantation.[6] We believe that an isolated autologous pericardium might also be an ideal material. Although the isolated autologous pericardium lacked the growth potential, it might require a shorter period of anticoagulation treatment than what is needed for artificial prosthetic materials.

The timing of the staged palliation remains under dispute. If the hepatic-to-azygos connection was performed simultaneously with the Kawashima procedure, it would be analogous to performing a one-stage Fontan completion.[7] To the contrary, to prolong the uncomplicated stage as long as possible, Arrigoni et al[3] suggested making the hepatic-to-azygos connection after initial signs of development of pulmonary arteriovenous malformations. However, there have been cases reporting that not all pulmonary arteriovenous malformations can be reversed.[8] Performing the hepatic-to-azygos connection for the Fontan completion prior to when pulmonary arteriovenous malformations might form seemed to be a reasonable solution.

It was essential to have an operation plan to evaluate the pattern of hepatic vein drainage due to its variability, including isolated drainage through bilateral independent branches and other potential routes. For the branches of the hepatic vein, the surgeons used to make an atrial cuff as a conduit for the hepatic-to-azygos connection. Creating an intra-atrial tunnel was mentioned as well as connecting the branches outside the atrium by prosthetic conduits. Ligating directly has also been reported, especially in cases of reoperation due to the undiagnosed branches.[4]

We tentatively used the autologous pericardial roll for the hepatic-to-azygos connection because it might require a short period of anticoagulation treatment. Although the short-term evaluation was satisfactory, a long-term follow-up would confirm the prognosis.

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Conflicts of Interest. None.

Ethical Standards. Fuwai Hospital Committee on Clinical Investigation approved the review of patient medical records.

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