[CASE REPORT]

Kawashima Procedure After Staged Unifocalizations in Asplenia With Major Aortopulmonary Collateral Arteries

Short title; UF AND KAWASHIMA IN ASPLENIA

Yukiko Ban, MD,* Mio Noma, MD, PhD,* Hitoshi Horigome, MD, PhD,†
Hideyuki Kato, MD,* Chiho Tokunaga, MD, PhD,* Yuzuru Sakakibara, MD, PhD,*
and Yuji Hiramatsu, MD, PhD*

*Departments of *Cardiovascular Surgery and †Pediatric Cardiology,*
University of Tsukuba, Tsukuba, Japan,

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Address for correspondence:
Yuji Hiramatsu, MD, PhD
Department of Cardiovascular Surgery, University of Tsukuba
1-1-1 Tennodai, Tsukuba 305-8575, Japan
Tel; +81 29 853 3210
Fax; +81 29 853 3097
e-mail: yuji3@md.tsukuba.ac.jp

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ABSTRACT

We report a Kawashima procedure (total cavopulmonary shunt) successfully carried out for asplenia syndrome, pulmonary atresia and major aortopulmonary collateral arteries. At the age of eight the patient underwent staged bilateral unifocalizations utilizing confluent central pulmonary arteries concomitant with bilateral modified Blalock-Taussig shunts. As the result of an interrupted inferior vena cava with azygous continuation, the patient required a Kawashima procedure with augmentation of the central pulmonary arteries for definitive palliation one year later. Cyanosis, respiratory distress and ventricular function improved.

(81 words)

The presence of major aortopulmonary collateral arteries (MAPCAs) in patients with single ventricle physiology is an extremely rare but serious condition. In this patient group, it is still unclear if pulmonary vascular resistance can be maintained at low enough levels to sustain a stable long term Fontan-type circulation (1). We report a successful case of staged unifocalizations followed by Kawashima procedure (total cavopulmonary shunt) in asplenia with MAPCAs.

A baby girl was diagnosed with asplenia syndrome, single ventricle morphology, common atrioventricular canal, pulmonary atresia, right aortic arch, single left superior vena cava, interrupted inferior vena cava with azygous continuation, and MAPCAs. Because of the size of her central pulmonary arteries surgery was not an option until age six when angiography revealed diminutive but confluent central pulmonary arteries and at least four MAPCAs arising from the descending thoracic aorta [Figure 1]. At that time, due to pulmonary over-circulation, the patient's arterial oxygen saturation (SaO_2) was about 90% with marked cardiomegaly and it was decided that her best option was surgery.

The patient underwent staged bilateral unifocalizations. The first procedure was a left sided uniforcalization with a modified Blalock-Taussig shunt (5mm) performed through a left lateral thoracotomy. This procedure was followed four months later by a right sided unifocalization and a modified Blalock-Taussig shunt (4mm) through a right lateral thoracotomy.

Catheterization 10 months after the second unifocalization showed low pulmonary vascular resistance (1.4 Wood units), relatively low pulmonary arterial pressure (mean right and left pulmonary arterial pressures of 12.5 and 14 mmHg, respectively) and reasonable pulmonary vascular beds (pulmonary arterial index 140 mm^3/m^2).

At 9 years of age, she underwent Kawashima procedure, augmentation of the central pulmonary arteries with an autologous pericardial patch, and takedown of bilateral modified Blalock-Taussig shunts. Catheterization after the Kawashima procedure showed a well reconstructed pulmonary blood pathway with no remarkable obstruction and acceptable pulmonary arterial pressure (mean 15 mmHg) [Figure 2]. Two years after this final procedure, the patient is in good condition with SaO_2 of 85%.

Comment

The surgical strategy for pulmonary atresia with ventricular septal defect (PA/VSD) and MAPCAs has evolved substantially and general principles have been well established. The standard protocol includes early midline single-stage unifocalization of all sources of pulmonary flow and intracardiac repair. This strategy has provided a high rate of early complete repair with favorable right ventricular pressure (2). It has been reported that over 90% of patients with MAPCAs have PA/VSD as their intracardiac anatomy with very few having single ventricle physiology (1). In a recent study of patients with MAPCAs who underwent unifocalizations, only 4.1% (14 patients) had single ventricles including six with asplenia (1). Among the 6 with asplenia, only 1 patient successfully underwent the Fontan procedure.

The prevalence and the clinical implications of MAPCAs in patients with asplenia remain unclear. The natural long-term fate of unifocalized MAPCAs often follows progressive stenosis, occlusion or growth failure which results in the loss of bronchopulmonary segments (3). As a result the presence of MAPCAs may lead to less favorable pulmonary vascular resistance before or even after completion of cavopulmonary connections and preclude the maintenance of long term Fontan

circulation in these patients.

A survey of the literature revealed only 4 reported patients with asplenia and MAPCAs who had achieved successful staged repair. One patient underwent biventricular repair (4), and 3 other patients achieved Fontan-type completion (1, 5, 6). Our report should be an important addition to the literature as the first case of Kawashima procedure carried out successfully in a patient with asplenia and MAPCAs.

Kawashima procedure incorporates most (about 85%) of the systemic venous return into the pulmonary circulation, excluding coronary sinus and hepatic venous flow (7). Although this particular near-total Fontan circulation usually brings SaO_2 into the range of 85 to 90%, the inability to use a two-stage approach to achieve Fontan-type permanent palliation is associated with higher surgical risk. Recent reports recommend various fenestration techniques in high-risk Kawashima procedures (8). However, since fenestrated Kawashima may result in unsatisfactory oxygen saturation levels, it should not be considered a definitive procedure (7). Therefore in staged unifocalizations in patients with MAPCAs and IVC interruption, recruitment of as many pulmonary segments as possible to the central pulmonary artery is a prerequisite for creating a greater chance of Kawashima completion without fenestration. In this case we carefully looked for the chances to recruit MAPCAs to the diminutive but confluent central pulmonary arteries, and successfully reconstructed a reasonable pulmonary vasculature by staged unifocalizations.

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Figure Legends

Fig 1. *(A) Angiogram before unifocalizations showing multiple major aortopulmonary collateral arteries (white arrows) arising from the descending aorta. (B)Diminutive but confluent central pulmonary arteries (*) are connecting to a right upper lobe collateral.*

Fig 2. *Angiogram after Kawashima operation showing well reconstructed pulmonary vasculature. The catheter is passed antegrade into the left superior vena cava through the azygos vein.*