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Brief Report

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Cardiac MRI for Fontan candidates after intrapulmonary-artery septation

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Abstract

Intrapulmonary-artery septoplasty may be effective for establishing two-lung Fontan circulation in patients with unilateral pulmonary circulation. However, evaluation of the function of each lung by conventional modalities can be challenging in these patients due to differing sources of blood flow to the left and right lungs following intrapulmonary-artery septation. Herein, we report a case in which two-lung Fontan circulation was successfully achieved after using cardiac MRI along with conventional modalities to evaluate pulmonary circulation.

The Fontan procedure is frequently performed in patients with functional single-ventricle physiology.[1] Surgical outcomes for this procedure have improved with technical advancements[2]; however, contraindications for the Fontan procedure include unbalanced pulmonary artery development with unilateral pulmonary artery hypoplasia and/or pulmonary venous obstruction.[3] In previous reports, we evaluated the efficacy of intrapulmonary-artery septation in patients with unilateral pulmonary circulation.[4,5]

Traditionally, pulmonary function has been evaluated using cardiac catheterisation, echocardiography, and lung perfusion scanning to determine patient eligibility for the Fontan procedure. However, in patients with intrapulmonary-artery septation, it can be difficult to evaluate parameters such as pulmonary blood flow, pulmonary resistance, and pulmonary venous physiology when assessing the function of each lung owing to the differences in the blood flow sources to the left and right lungs. Here, we report a case of successful two-lung Fontan circulation after using cardiac MRI along with conventional modalities for evaluation of pulmonary circulation.

Case report

As anonymised patient data were used, and the need for a written informed consent for publication of patient information and images was waived for this case report.

A male neonate weighing 2344 g was diagnosed with double-inlet left ventricle, transposition of the great arteries, ventricular septal defect, and mild valvular pulmonary stenosis. He had undergone a staged palliation procedure on day 15 of hospitalisation at another hospital. Patent ductus arteriosus ligation and main pulmonary artery banding were performed. At 8 months of age, he underwent bidirectional cavopulmonary anastomosis and pulmonary artery plasty and recovered uneventfully.

However, at 4 years and 2 months of age, the Fontan procedure was planned, but cardiac catheterisation revealed decreased left pulmonary circulation with left pulmonary artery hypoplasia and left pulmonary venous obstruction, and the Fontan procedure was absolutely contraindicated.

The patient was referred to our hospital at 5 years and 1 month of age. At 5 years and 3 months (body weight 15 kg), we performed intrapulmonary-artery septation using a 4-mm expanded polytetrafluoroethylene tube graft to the left pulmonary artery and unilateral cavopulmonary anastomosis to the right pulmonary artery with intrapulmonary- septation. The septation patch (0.1-mm expanded polytetrafluoroethylene sheet [GORE®-PRECLUDE®; Gore® Medical Products, Flagstaff, AZ, USA]) was sutured obliquely between the right and left pulmonary arteries. Pulmonary artery anterior-roof reconstruction was performed via patch augmentation with a 0.4-mm expanded polytetrafluoroethylene patch (GORE-TEX; Gore® Medical Products). Left pulmonary venous obstruction release included extensive resection of the stenotic and atretic lesions and concomitant sutureless repair to improve left pulmonary circulation and ensure that the patient would be able to undergo the Fontan procedure. Six months after intrapulmonary-artery septation, cardiac MRI and catheterisation were performed to evaluate pulmonary haemodynamics (Fig 1).

Cardiac MRI was performed using Ingenia, a 1.5-T whole-body system (Philips healthcare, Amsterdam, Netherlands).

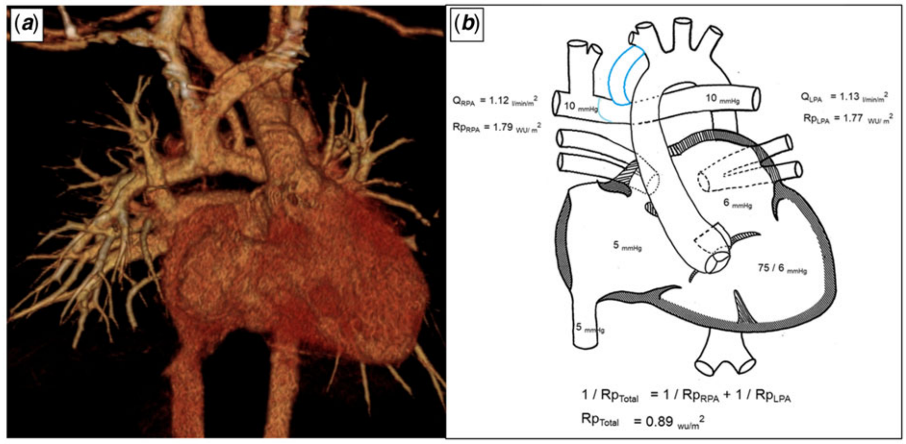

Figure 1. (***A***) Computed tomography scan; (***B***) Evaluation of pulmonary hemodynamics by CMR and catheterization after IPAS. BSA, body surface area; CMR, cardiac magnetic resonance; IPAS, intrapulmonary-artery septation; LPA, left pulmonary artery; Qp, pulmonary blood flow; Rp, pulmonary resistance; RPA, right pulmonary artery.

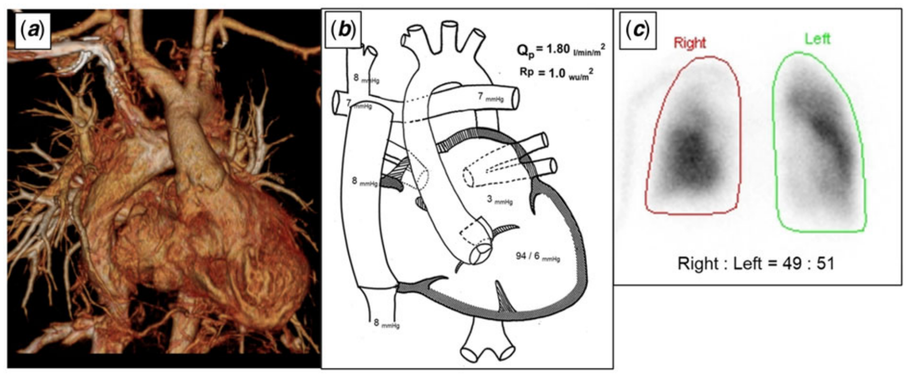

Figure 2. (***A***) Computed tomography scan; (***B***) Evaluation of pulmonary hemodynamics by catheterization and (***C***) lung perfusion scanning after the Fontan procedure. Qp, pulmonary blood flow; Rp, pulmonary resistance.

Phase-contrast cardiac MRI data from the right and left pulmonary arteries in front of the branch were obtained. cvi42 software (Circle Cardiovascular Imaging, Calgary, Canada) was used to quantify pulmonary blood flow, which was defined as right or left anterograde pulmonary blood flow rather than pulmonary venous blood flow (systemic-to-pulmonary collateral flow was excluded). Subsequently, we calculated the pulmonary blood flow of the right and left pulmonary artery, and these values were normalised to the body surface area. Pulmonary artery pressure and left atrial pressure were measured using cardiac catheterisation. Pulmonary resistance was calculated by dividing the trans-pulmonary gradient (mean pulmonary pressure – left atrial pressure) by indexed pulmonary blood flow. Thus, we calculated the pulmonary resistance in each lung and estimated the total Rp (1/pulmonary resistance $_{\text{total}}$ = 1/pulmonary resistance $_{\text{RPA}}$ + 1/pulmonary resistance $_{\text{LPA}}$) to be obtained following the Fontan procedure. The body surface area was 0.67 m^2, and the mean pulmonary artery pressure was 10 mmHg for both the left and right pulmonary arteries. Pulmonary blood flow $_{\text{RPA}}$ and pulmonary blood flow $_{\text{LPA}}$ measured by cardiac MRI were 1.12 l/min and 1.13 l/min, respectively. Pulmonary resistance $_{\text{RPA}}$ was 2.39 Woods Units/m^2 (WU/m^2), pulmonary resistance $_{\text{LPA}}$ was 2.38 WU/m^2, and pulmonary resistance $_{\text{total}}$ was 1.2 WU/m^2. These results indicated that two-lung Fontan circulation could be achieved, and the patient underwent the Fontan procedure at 6 years and 1 month of age. The postoperative course was uneventful, and he was discharged on the 12th postoperative day.

Six months postoperatively, the patient underwent evaluation through catheterisation and lung perfusion scanning (Fig 2). The mean pulmonary arterial pressure was 7 mmHg for both the left and right pulmonary arteries. The pulmonary blood flow and pulmonary resistance were 1.80 l/min/m^2 and 1.0 WU/m^2 by the oximetric (Fick) method, respectively. Lung perfusion scanning showed nearly even left and right pulmonary artery blood flow (right lung: left lung = 49:51). Five years passed following the Fontan procedure without postoperative complications, and outpatient follow-up is ongoing.

Comment

This case showed that two-lung Fontan circulation could be successfully achieved after using cardiac MRI along with conventional modalities to evaluate pulmonary circulation.

In CHD, cardiac MRI using phase-contrast through-plane sequences plays an important role in quantifying pulmonary blood flow.[6] Previous studies have demonstrated a significant correlation between several cardiac MRI-derived parameters and catheterisation-quantified pulmonary resistance. For example, Bell et al. reported that cardiac MRI and cardiac catheterisation combine noninvasive measurement of pulmonary blood flow using phase-contrast imaging with invasive pressure measurement to accurately determine pulmonary resistance.[7] Therefore, it became common practice in our hospital to combine cardiac MRI and cardiac catheterisation to evaluate patients after intrapulmonary-artery septation. By calculating pulmonary blood flow and pulmonary resistance for both lungs and estimating the total pulmonary resistance, we determined whether two-lung Fontan circulation could be established in patients who had undergone intrapulmonary-artery septation. In this case, the pulmonary resistance

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between the unaffected and affected (Blalock–Taussig shunt side) lung was approximately 1:1, and thus a successful two-lung Fontan circulation could be established.

Our approach to this case had some limitations. First, standardised methods for performing and analysing cardiac MRI have not yet been defined. Therefore, operator experience and inter- and intraoperator variability may affect the reproducibility of these results. Phase-contrast measurements may be affected by turbulent flow, which may in turn result in measurement inaccuracies. Second, it is difficult to define a clear cut-off value for pulmonary resistance in each lung obtained by our method, which could be used to definitively determine whether two-lung Fontan circulation can be achieved. Therefore, more cardiac MRI data should be collected to standardise cardiac MRI and its analysis and to clarify the indications for the Fontan procedure following intrapulmonary-artery septation.

Conflicts of interest. None.

Ethical standards. This article does not contain any studies with human participants performed by any of the authors.

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