CASE REPORT

Temporary Decompression of the Right Ventricle to Assess the Right Ventricle-dependent Coronary Circulation in Pulmonary Atresia with Intact Ventricular Septum

We repaired a case of pulmonary atresia with intact ventricular septum in which the blood supply to the left anterior descending coronary artery depended on the right ventricle. At the time of a bidirectional Glenn operation, total cardiopulmonary bypass with venous drainage from the right atrium was performed in order to evaluate the safety of right ventricular decompression required for a planned Fontan operation. We confirmed the dependence of the coronary perfusion on the right ventricle by demonstrating transient depression of the ST segment in the epicardial electrocardiogram during temporary decompression of the right ventricle. To prevent ischemic myocardial damage, we then performed an extracardiac Fontan operation with a temporary venous shunt and without cardiopulmonary bypass. (Jpn J Thorac Cardiovasc Surg 2005; 53: 400-403)

Key words: extracardiac Fontan operation, pulmonary atresia with intact ventricular septum, right ventricle-dependent coronary circulation

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For a patient of pulmonary atresia with intact ventricular septum (PA-IVS), decompression of the right ventricle is usually contraindicated when coronary perfusion is strongly dependent on it. However, in certain cases where there is less dependence, the right ventricle can be decompressed safely. Therefore, determination of the extent to which the coronary circulation depends on the right ventricle is essential for deciding the appropriate surgical approach.

In the case reported here, we assessed the dependence of the coronary perfusion on the right ventricle by means of temporary decompression during a Glenn operation, which was performed as staged palliative surgery.

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Case

An infant boy whose pulmonary atresia with intact ventricular septum was diagnosed by means of echocardiography underwent cardiac catheterization when he was 16 days old. Since the right ventriculogram showed fistulas to the right and left coronary arteries, he underwent a left modified Blalock-Taussig shunt when he was 21 days old. The right ventriculogram obtained 1 month after the shunt operation showed the right ventricular end-diastolic volume was 38% normal and the diameter of the tricuspid valve annulus was 5.0 mm (37% of normal; Z-value=-3.7). Selective coronary artery angiography showed that the proximal left anterior descending coronary artery was interrupted and that the distal portion was opacified during the ventricular systolic phase (Fig. 1). The right coronary artery was intact.

At the age of 1 year, the patient underwent a bidirectional Glenn operation using a partial cardiopulmonary bypass. To avoid decompression of the right ventricle, systemic venous drainage was performed only from the superior caval vein with the heart beating. Throughout the procedure an epicardial electrocardiogram on the
Before the venous drainage

After temporary decompression of RV

After the termination of the venous drainage

Fig. 1. Angiography after Blalock-Taussig shunt. Left anterior descending coronary artery was interrupted proximally (A, arrow) and opacified distally during the ventricular systolic phase on the right ventriculogram (B).

Fig. 2. After temporary decompression of the right ventricle by venous drainage from the right atrium, the epicardial electrocardiogram on the inferior wall of the ventricle showed depression of the ST segment a few seconds later. The ST change normalized a few minutes after termination of the venous drainage from the right atrium.

Before the venous drainage

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inferior wall of the ventricle was monitored. After completion of the bidirectional Glenn anastomosis, we assessed the dependence of the coronary circulation on the right ventricle, by temporarily decompressing it. A 20-Fr. right-angled venous cannula was placed in the inferior caval vein at the junction to the right atrium. Under constant monitoring of right ventricular pressure, a slow but total cardiopulmonary bypass with venous drainage from the right atrium was performed. Although the mean systemic perfusion pressure was kept to approximately 60 mmHg, the epicardial electrocardiogram showed depression of the ST segment a few seconds after the right ventricle had been decompressed to 30 mmHg. The change in the ST normalized a few minutes after venous drainage from the right atrium had ended (Fig. 2). The perfusion pressure could be kept stable during the procedure, and weaning from the cardiopulmonary bypass was smooth.

When he was 4 years old, the patient underwent an extracardiac Fontan operation without cardiopulmonary bypass. The main pulmonary artery was clamped proximally to the Glenn anastomosis and to the left modified Blalock-Taussig shunt. The pulmonary trunk was then divided, a 16-mm ringed expanded polytetrafluoroethylene tube (W. L. Gore & Associates; Flagstaff, AZ, USA) was anastomosed to the incised main pulmonary artery in an end-to-side fashion. A 22-Fr. right-angled venous cannula was placed in the inferior caval vein at the junction to the right atrium, and a temporary venous shunt was made by connecting the cannula to another 22-Fr. right-angled venous cannula that was inserted at the right atrial appendage. The inferior caval vein was clamped and divided just above the venous cannula, with a stable venous pressure of 8 mmHg measured at