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Cardiac Catheterization Is Necessary Before Bidirectional Glenn and Fontan Procedures in Single Ventricle Physiology

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Abstract. There are significant mortality and morbidity in bidirectional Glenn (stage II) and Fontan (stage III) procedures for congenital heart diseases with single ventricle physiology. In order to minimize the mortality and morbidity, the presence or absence of hemodynamic and anatomical abnormalities, such as poor ventricular function, coarctation of the aorta, pulmonary artery distortion, high pulmonary resistance, and abnormal collateral vessels, should be evaluated. Echocardiography and magnetic resonance imaging (MRI) can visualize ventricular size and coarctation of the aorta but may fail to visualize pulmonary artery distortion. Furthermore, cardiac catheterization is needed to measure pulmonary resistance. If the pulmonary resistance is >3 Wood units/m^2 before stage II, the risk for the bidirectional Glenn operation may be high. Abnormal aortopulmonary collateral vessels may develop after the stage II procedure and echocardiography and MRI may not visualize these vessels. Coil embolization may be required to treat these vessels. In conclusion, cardiac catheterization is required to evaluate abnormalities, which may be treated by interventional catheterization, and to reduce mortality and morbidity of stage II and III procedures.

Key words: Hypoplastic left heart syndrome — Glenn operation — Fontan operation — Cardiac catheterization

In high-risk patients with single ventricle physiology, staged procedures including bidirectional cavopulmonary anastomosis (Glenn shunt) followed by the Fontan procedure are sometimes performed to minimize the risk of Fontan operation [4, 11, 20, 21]. In patients with hypoplastic left heart syndrome, Norwood operation is initially performed in the newborn period (stage I) and then bidirectional Glenn shunt (stage II) followed by Fontan operation (stage III) are usually performed [2]. Indication for bidirectional Glenn shunt and subsequent Fontan operation in high-risk patients have been controversial. Poor ventricular function with an ejection fraction less than 0.4, more than moderate atrioventricular valve regurgitation, anomalous pulmonary venous return, a distortion of the pulmonary artery such as nonconfluent or almost nonconfluent pulmonary artery, hypoplastic pulmonary artery with a Nakata index less than 200-250 mm^2/m^2, pulmonary hypertension with a mean pulmonary arterial pressure of more than 20 mmHg, and a pulmonary vascular resistance of more than 3 Wood units.m^2 are known to be risk factors for Fontan operation [4, 6, 11, 17, 20, 21]. In patients with hypoplastic left heart syndrome, mortality and morbidity of stage II and III procedures are still high. In order to minimize mortality and morbidity, hemodynamic and anatomical abnormalities should be detected before these procedures are performed. It is controversial whether cardiac catheterization is necessary before bidirectional Glenn shunt and Fontan operation when sufficient data are available from echocardiography and magnetic resonance imaging (MRI).

Echocardiography and MRI

Left ventricular ejection fraction can be calculated from M-mode echocardiography and sometimes from two-dimensional echocardiography [14]. Echocardiographic measurement of right ventricular ejection fraction is tedious and time-consuming [8], and right ventricular function is usually estimated only qualitatively. However, it is possible to measure right and left ventricular ejection fraction noninvasively with MRI [18]. Anomalous pulmonary venous return can be diagnosed using echocardiography. The

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presence or absence of atrioventricular valve regurgitation and its severity can be estimated using echocardiography [13]. The central pulmonary artery is usually visualized and the presence or absence of a distortion of the pulmonary artery may be diagnosed using echocardiography. Assessment of the peripheral pulmonary arteries and measurement of the Nakata index are often difficult with echocardiography. It is possible to measure Nakata index and pulmonary blood flow using MRI [5, 10].

In patients who have undergone the Norwood procedure, re-coarctation of the aorta may exist [3]. The presence or absence of coarctation of the aorta can be determined by echocardiography and MRI, but in the case of mild coarctation, pressure measurement with catheter may be required to determine whether catheter intervention should be performed.

Pulmonary Resistance and Glenn Shunt

In a series from a single institution, Takeda et al. [21] showed that mortality, including late death, of Glenn shunt was 11% in 52 high-risk Fontan candidates, and Masuda et al. [11] showed that mortality of Fontan operation after Glenn shunt was 0%. In their series, 3 of 10 patients (30%) with pulmonary resistance >3 Wood units/m^2 died and 2 of 5 patients (40%) with pulmonary artery pressure >20 mmHg died after bidirectional Glenn operation. These studies suggest that in the staged procedures, if the patients survive the bidirectional Glenn shunt, the chances of surviving the Fontan operation are high and hemodynamic evaluation before the bidirectional Glenn shunt is important. Alejos et al. [1] showed that in 129 patients who underwent bidirectional Glenn shunt, mortality was 4% when the mean pulmonary pressure was <18 mmHg and 20% when the pulmonary was >18 mmHg. Pridijan et al. [20] also showed that pulmonary resistance >3 Wood units/m^2 predicted high mortality of the bidirectional Glenn shunt. These data suggest that hemodynamic evaluation is important before bidirectional Glenn shunt is performed. However, it is not possible to measure pulmonary artery pressure and resistance accurately using echocardiography or MRI.

Although most patients had undergone Fontan operation without prior bidirectional Glenn shunt at our institution, mortality of the Fontan operation with pulmonary resistance <3 Wood units/m^2 was approximately 5%, that with pulmonary resistance between 3 and 4 Wood units/m^2 was 20%, and that with pulmonary resistance >4 Wood units/m^2 was 80% [19]. Similarly, in our institution, mortality of the Fontan operation with mean pulmonary pressure <20 mmHg was approximately 7-9%, and that with mean pulmonary pressure >20 mmHg was 26% [19].

From these data, I conclude that cardiac catheterization is necessary to measure pulmonary artery pressure and pulmonary resistance before bidirectional Glenn shunt. It is not clear if the pulmonary resistance >3 Wood units/m^2 after Glenn shunt is a risk factor for Fontan completion. However, it is usually possible to perform the stage III procedure if the pulmonary resistance is less than 3 Wood units/m^2 before the stage II procedure, although the pulmonary resistance may be more than 3 Wood units/m^2 after the stage II procedure. Therefore, it is important to evaluate the pulmonary resistance before the stage II procedure.

Accurate assessment of the pulmonary arterial pressure is critical in high-risk candidates for bidirectional Glenn and Fontan procedures. In patients with pulmonary atresia or severe stenosis with or without aortopulmonary shunts, pulmonary blood flow and pressure may be interfered with by the catheter advanced into the pulmonary artery. We showed that a 3-Fr catheter is useful to accurately measure pulmonary arterial pressure in such patients [16]. It is also possible to estimate pulmonary arterial pressure from the pulmonary venous wedge pressure [7, 15, 22].

Pulmonary Artery Distortion and Occlusion

Echocardiography frequently fails to visualize the peripheral pulmonary arteries. MRI usually visualizes the presence of the peripheral pulmonary artery stenosis, but sometimes fails to do so. One side of the pulmonary artery may be occluded by the thrombus despite an MRI suggesting the presence of both sides of the pulmonary arteries. The stage III procedure can be difficult, if not impossible, with only one side of the pulmonary artery. In a series of 158 Norwood operations, Bove and Lloyd [3] reported that 3 cases required pulmonary artery thrombectomy and/or pulmonary artery angioplasty and 12 cases of pulmonary stenosis required balloon angioplasty and stent placement. From these data, I conclude that cardiac catheterization is important to evaluate the presence or absence of peripheral pulmonary artery stenosis before stage II and III procedures.

Collateral Vessels

Abnormal aortopulmonary collateral vessels may develop after the stage II procedure, and echocardiography and MRI may not visualize these vessels. Triedman et al. [23] showed that patients who underwent the bidirectional Glenn procedure were more likely to have collateral vessels than patients who underwent the Fontan procedure (65 vs 30%).

Coil embolization may be required to treat these vessels [9]. In a series of 158 Norwood operations, Bove and Lloyd [3] reported that there were 16 cases requiring coil embolization. Furthermore, collateral vessels between the superior vena cava and left atrium may develop after the stage II procedure, and echocardiography and MRI may not visualize these vessels. These vessels may also need to be treated before the stage III procedure [12].

Conclusion

Cardiac catheterization is required to evaluate abnormalities, which may be treated by interventional catheterization, and to reduce mortality and morbidity of stage II and III procedures.

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