

Editorial



Evaluation of Pulmonary Blood Flow Pulsatility in Patients Undergoing Various Fontan Palliation Techniques

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Conflict of Interest

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Since Fontan palliation was introduced in 1968, improved survival has been achieved in univentricular heart patients, but long-term adverse events remain a problem.¹⁾

Since the original description of the Fontan operation, multiple modifications of the procedure have been introduced. The three most common types of Fontan circulation are atriopulmonary connection (APC), lateral tunnel (LT), and extracardiac total cavopulmonary connection (EC).

In Fontan palliation, the caval veins are connected without a pumping ventricle to the pulmonary arteries. The Fontan circulation has unique hemodynamics characterized by nonpulsatile lung perfusion, chronic systemic venous hypertension, and low cardiac output.^{1,2)} Lack of pulsatile flow is one of the mechanisms of pulmonary vascular bed abnormalities.^{3,4)} Chronic privation of pulsatile pulmonary flow is detrimental for endothelial function, capillary recruitment, and pulmonary vascular development, all of which impact pulmonary vascular resistance, as seen in animal models.^{5,6)}

Theoretically, adding pulsation to the pulmonary artery in Fontan circulation may promote pulmonary artery development and preserve endothelial function.⁵⁻⁸⁾

However, Kalia et al.⁹⁾ reported that maintenance of pulmonary flow pulsation with some forms of forward flow at the time of Fontan palliation did not alter short-term outcomes or long-term prognosis although it tended to increase postoperative oxygen saturation.

Whether adding pulsation to the pulmonary artery in Fontan circulation leads to improved hemodynamic outcomes remains unclear.

There have been a few reports on pulsatility index (PI) evaluation of pulmonary artery flow in different types of Fontan circulation.^{10,11)} Klimes et al.¹⁰⁾ reported cardiac magnetic resonance imaging-derived pulmonary artery pulsatility in Fontan patients. The APC Fontan group had a normal pulsatility, a dilated right atrium and partial backward flow in the inferior vena cava (IVC), demonstrating suboptimal Fontan circulation. LT- and EC-type Fontan both produced unidirectional antegrade flow in the IVC but pulsatility was very low, which may increase pulmonary vascular resistance contributing to late Fontan failure.

In this issue of the *Journal of Cardiovascular Imaging*, Shabanian et al.¹²⁾ investigated PI measured by 2-dimensional echocardiography in patients with different types of Fontan palliation and normal controls. Their cohort included several modifications of EC Fontan that involved adding pulsation to pulmonary circulation. In their study, all enrolled patients had good clinical status and favorable hemodynamic parameters. Near-normal PI was demonstrated in APC Fontan patients, and lowest PI was observed in the LT and EC Fontan groups. Furthermore, they added pulsation to Fontan circulation via several modifications to the EC-type Fontan procedure in selected patients, which was confirmed by increased PI.

This study has limitations including a small sample size of patients with favorable outcomes at a single center, and thus selection bias was inevitable. Moreover, echocardiographic-based PI is technically challenging due to the poor acoustic window from chest wall deformities related to recurrent sternotomy and geometric variation in the shape and spatial relationships of actual vessels and conduits.

Despite these limitations, PI is a noninvasive and simple but reliable parameter. PI can be used to evaluate the hemodynamic status of Fontan patients. Further studies are needed to elucidate the hemodynamics associated with pulsatile pulmonary arteries in Fontan circulation.

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