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Commentary: When is a bidirectional cavopulmonary shunt a bad idea?

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CENTRAL MESSAGE

Proceed with caution when cardiac magnetic resonance imaging–derived pre–bidirectional cavopulmonary shunt superior vena cava blood flow is low. The “conventional” staging sequence of palliation for a functional single ventricle may be inappropriate in this setting.

In the early 1970s, Fontan and Baudet¹ and Kreutzer and colleagues² reported successful separation of the systemic and pulmonary circulations in tricuspid atresia. Since then, surgeons and cardiologists have sought palliative procedures designed to produce the “ideal” total cavopulmonary connection (TCPC) candidate. In 1985, Hopkins and colleagues³ reported on the efficacy of the bidirectional cavopulmonary shunt (BCPS) as a palliative procedure, thereby launching the era of staged systemic venous pathway redirection.⁴ In recent years, the timing and applicability of the BCPS have been adjusted to meet the needs of a growing population of complex functional single-ventricle patients who are now surviving early infancy.

In 2012, the Toronto group reported directly measured superior vena cava (SVC) blood flow during the BCPS procedure.⁵ Low intraoperative pre-BCPS SVC flow correlated with poor clinical outcome. The current report, presented in this issue of the *Journal*,⁶ reviews all patients selected for BCPS at The Hospital for Sick Children between January 2012 and December 2017. Every patient underwent pre-BCPS cardiac catheterization, cardiac magnetic resonance imaging (CMR), and echocardiography. The methodology used for CMR data acquisition and analysis is well established,⁷ and the use of CMR as the primary pre-BCPS evaluation has been suggested previously.⁸

Univariate analysis identified moderate to severe atrioventricular valve regurgitation, pre-BCPS absolute low SVC blood flow, and prolonged cardiopulmonary

bypass (CPB) time as risk factors for poor outcome. In multivariate analysis, only pre-BCPS indexed SVC flow was a risk factor for poor outcome.

This isolated risk factor finding is a bit surprising and somewhat counterintuitive. Is low pre-BCPS SVC blood flow an independent variable, or is it a surrogate for a complex interaction between multiple variables? The authors do not provide any information regarding the physiological status of patients during CMR. Their patient cohort is quite heterogeneous, and the previous methods of palliation are varied. Is it possible that patients palliated by the Hybrid technique, patients with hypoplastic left heart syndrome, and patients with atrioventricular (AV) valve regurgitation (ie, patients requiring longer CPB time during BCPS) develop low SVC blood flow during CMR? Cerebral blood flow may be influenced by arterial PCO₂ levels^{9,10} and other anatomic and physiological considerations.¹¹

The adverse effect of thrombosis in the upper central venous circulation on SVC blood flow can be anticipated. We have avoided the use of internal jugular lines whenever possible in infants born with a functional single ventricle. In addition, although we recommend measuring SVC pressure at the completion of BCPS, we do not recommend leaving an indwelling jugular vein catheter in place following the procedure.

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This report is an important contribution because it calls our attention to a subset of patients that are probably not good candidates for a conventionally programmed single-ventricle pathway. Low pre-BCPS SVC flow may indicate that the second-stage procedure should be designed to address the comorbid factors (eg, AV valve regurgitation, peripheral pulmonary artery stenosis) before contemplating a BCPS procedure. In essence, low pre-BCPS SVC flow may be a discriminating factor that sorts out “gray zone” candidates for BCPS. We agree with the authors’ conclusion that some single-ventricle patients may be better served by early cardiac transplantation. Finally, it is worth noting that most Fontan/Kreutzer survivors over age 30 years have not undergone a BCPS procedure, indicating that it is not absolutely necessary that every patient undergo BCPS before TCPC or that all candidates for TCPC undergo the procedure before age 4 years.

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