Abnormal vascular connections between systemic arteries and the pulmonary vascular bed—systemic-to-pulmonary collateral (SPC) vessels—manifest in patients with a variety of congenital and acquired heart disease. These vessels, which originate from branches off the brachiocephalic arteries, chest wall arteries, and the descending aorta, vary markedly in size, number, course, and distribution. Although the precise mechanisms that lead to development of SPCs are incompletely understood, hypoxemia, diminished global or regional pulmonary blood flow, and nonpulsatile flow in the pulmonary arteries are some of the commonly cited contributors. Indeed, SPCs are frequently encountered in patients with cardiac anomalies that include ≥1 of these abnormalities such as severe forms of tetralogy of Fallot and functional single ventricle (FSV). In the latter group, the clinical importance of SPCs and their optimal management have been topics of intense debate for >2 decades.

The physiology of SPC flow involves both beneficial and deleterious effects. SPCs can augment blood flow to hyperperfused lung segments, improve gas exchange, and potentially inhibit the development of pulmonary arteriovenous malformations in patients in whom hepatic venous flow does not reach the pulmonary circulation. In contrast, like other shunts, SPC flow is inherently inefficient because relatively highly oxygenated blood returns to the pulmonary circulation and potentially competes with the more efficient antegrade flow to the lungs through the native pulmonary arteries. Consequently, SPC flow results in volume load on the FSV, which can lead to ventricular dilatation and dysfunction, atrioventricular valve regurgitation, and heart failure symptoms. Other deleterious effects of excessive SPCs include flow energy loss, exuberant blood flow returning through the pulmonary veins to the surgical field during cardiopulmonary bypass, respiratory distress, pulmonary hypertension, and hemoptysis. In patients with FSV, the hemodynamic burden from SPCs has been the topic of particular concern because the FSV is thought to be vulnerable to ventricular dysfunction and elevated pulmonary vascular resistance can lead to complications such as low cardiac output, central venous hypertension, pleural effusions, ascites, and protein-losing enteropathy.

The optimal management strategy of SPCs in patients with FSV continues to be the subject of debate. On 1 end of the spectrum are those who occlude SPCs only in highly selected circumstances, usually when large collaterals with exuberant flow are considered the cause of clinical compromise. Advocates of this selective approach cite absence of evidence that routine occlusion of smaller SPCs results in improved clinical outcomes such as shorter duration of postoperative pericardial and pleural effusions, shorter hospital stay, lower incidence of ventricular dysfunction, or lower mortality. On the other end of the spectrum are those who routinely target most SPCs. The rationale cited by those who favor this strategy is based on the potential for unfavorable hemodynamics due to excessive SPC flow, which may adversely affect early and late clinical outcomes. This disparate practice was recently documented by Banka et al, who found that the incidence of SPC coil occlusion among the 7 centers that participated in the Pediatric Heart Network Fontan Cross-Sectional study varied from 0% to 30%. Adding to the controversy on the use of routine coil occlusion of SPCs, multivariable analysis in their nonrandomized cohort found no significant differences in hospital length of stay, postoperative complications, or patient outcomes at cross-sectional evaluation 8.6±3.4 years after Fontan between those who received coil occlusion and those who did not.

Until recently, the debate regarding the clinical impact of SPC flow and the efficacy of transcatheter occlusion strategies has been marred by lack of a reliable in vivo method to measure the amount of blood flow carried by these collaterals. In the absence of an optimal quantitative, noninvasive technique to measure SPC flow, investigators were unable to reliably determine the hemodynamic burden caused by these collaterals and to objectively evaluate the efficacy of coil occlusion during cardiac catheterization. The recent development of a cardiac MR technique to measure SPC flow noninvasively has overcome the drawbacks of previous methods to quantify collateral flow in patients with FSV. First described in a case report by Grosse-Wartmann et al in 2007, this method was subsequently refined and systematically evaluated in small cohorts by the same group and by Whitehead et al. Using phase contrast cine cardiac MR flow measurements in the aorta, superior and inferior venae...
cavae, branch pulmonary arteries, and pulmonary veins, SPC flow can be calculated in 2 different ways: (1) SPC flow = sum of pulmonary vein flow - sum of branch pulmonary artery flow; and (2) SPC flow = ascending aorta flow - sum of caval flow (Figure). The results of both methods are then compared with each other for consistency as a measure of internal control. In addition to total SPC flow, this technique also provides information on the amount of collateral flow in each lung.

In this issue of Circulation: Cardiovascular Imaging, Glatz et al\textsuperscript{16} report that higher SPC flow measured by cardiac MR in 44 patients before Fontan surgery was associated with longer postoperative duration of chest tube drainage and longer hospital length of stay after adjusting for presence of baffle fenestration and Fontan type. Keeping in mind the small sample size and the selective cohort (only 44 of 162 examples included in the calculation of SPC flow. In addition, flow measurement in the inferior vena cava can be challenging due to the short distance between the entrances of the hepatic veins and the atrial floor as well as the swirling, inhomogeneous low-velocity flow profile. Another unresolved question is whether flow measurement in the descending aorta just below the diaphragm can reliably substitute for flow measurement in the inferior vena cava. An advantage of substituting descending aorta flow for inferior vena cava flow is the more organized flow profile, which potentially results in a more accurate measurement. The pitfall of this approach is that SPCs originating below the diaphragm and decompressing veins in pre-Fontan patients are worth noting. In the absence of an in vivo reference standard for flow measurements, it is difficult to confirm the accuracy of SPC flow measurements in patients with FSV. On the other hand, the cardiac MR approach allows for internal assessment of consistency by comparing 2 distinct sets of flow measurements as well as comparison between ventricular stroke volumes measured by cine MR and aortic stroke volume measured by the phase contrast technique. Another challenge is the reproducibility of the flow measurements in patients with FSV. The interobserver reproducibility reported by Glatz et al (intraclass correlation coefficient = 0.8) and by Grosse-Wartmann et al (intraclass correlation coefficient = 0.73) is modest, likely due to compounding measurement variations from multiple samples included in the calculation of SPC flow. Finally, a logistic standpoint, the acquisition time of 8 to 9 cine phase contrast MRI sequences (Figure) usually exceeds 20 minutes, depending on heart rate and imaging parameters that determine spatial and temporal resolutions. The clinical use and accuracy of faster acquisition strategies, including breath-hold and real-time techniques,\textsuperscript{20} are yet to be tested in this patient group.

The study of Glatz et al also highlights new opportunities to gain insights into the clinical consequences of SPC flow in patients with FSV and the efficacy of transcatheter therapy. Given that the available literature indicates that equipoise exists with regard to highly selective versus routine ap-
proaches to catheter-based occlusion of SPCs in patients with FSV, the time is now ripe for a prospective randomized clinical trial to determine which approach provides superior early and midterm clinical benefits. The newly developed cardiac MR technique for SPC flow measurement can be instrumental for patient selection and baseline characterization as well as for assessment of treatment efficacy. In parallel to this effort, a prospective observational multicenter cohort using standardized data collection can inform us about the natural history of SPCs in patients with FSV and how this cardiac MR technique can be used for risk stratification. Studies with this quantitative tool, if properly validated and applied, will provide better data to standardize practice and improve outcomes for patients with complex congenital heart disease.

Disclosures

None.

References


Key Words: Editorials cardiovascular magnetic resonance imaging coil embolization congenital heart disease flow measurement Fontan procedure
Quantification of Systemic-to-Pulmonary Artery Collateral Flow: Challenges and Opportunities
Tal Geva

Circ Cardiovasc Imaging. 2012;5:175-177
doi: 10.1161/CIRCIMAGING.111.972182
Circulation: Cardiovascular Imaging is published by the American Heart Association, 7272 Greenville Avenue,
Dallas, TX 75231
Copyright © 2012 American Heart Association, Inc. All rights reserved.
Print ISSN: 1941-9651. Online ISSN: 1942-0080

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circimaging.ahajournals.org/content/5/2/175

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation: Cardiovascular Imaging can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation: Cardiovascular Imaging is online at:
http://circimaging.ahajournals.org//subscriptions/