Blood flow distribution in a large series of patients having the Fontan operation: A cardiac magnetic resonance velocity mapping study

Kevin K. Whitehead, MD, PhD,[a] Kartik S. Sundareswaran, MS,[b] W. James Parks, MD,[c]
Matthew A. Harris, MD,[a] Ajit P. Yoganathan, PhD,[b] and Mark A. Fogel, MD, FACC[a]

Objectives: Our goal was to determine flow distribution in the cavopulmonary connections of patients with and without bilateral superior venae cavae who had the Fontan procedure. No large series exists that establishes the flow distributions in Fontan patients, which would be an important resource for everyday clinical use and may affect future surgical reconstruction.

Methods: We studied 105 Fontan patients (aged 2–24 years) with through-plane phase contrast velocity mapping to determine flow rates in the inferior and superior venae cavae and left and right pulmonary arteries. Superior caval anastomosis type included 40 bidirectional Glenn shunts (of which 15 were bilateral) and 53 hemi-Fontan anastomoses; Fontan type included 69 intra-atrial baffles, 28 extracardiac conduits, and 4 atriopulmonary connections.

Results: Total caval flow was 2.9 ± 1.0 L · min^{-1} · m^{-2}, with an inferior vena cava contribution of 59% ± 15%. Total pulmonary flow was 2.5 ± 0.8 L · min^{-1} · m^{-2}, statistically less than caval flow and not explained by fenestration presence. The right pulmonary artery contribution (55% ± 13%) was statistically greater than the left. In patients with bilateral superior cavae, the right cava accounted for 52% ± 14% of the flow, with no difference in pulmonary flow splits (50% ± 16% to the right). Age and body surface area correlated with percent inferior caval contribution ($r = 0.60$ and 0.74, respectively). Superior vena cava anastomosis and Fontan type did not significantly affect pulmonary flow splits.

Conclusions: Total Fontan cardiac index was 2.9 L · min^{-1} · m^{-2}, with normal pulmonary flow splits (55% to the right lung). Inferior vena caval contribution to total flow increases with body surface area and age, consistent with data from healthy children.

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In the early 1970s, Fontan,[1] Kreutzer,[2] and their associates independently developed strategies for palliation in patients with tricuspid atresia that involved baffling the caval veins directly to the pulmonary arteries. This strategy has since been modified and adopted for the treatment of all patients with a single usable ventricle.[3-7] Staged reconstruction of the modified Fontan operation has become the standard by which single ventricle is palliated. Despite success at many institutions, managing these patients remains one of the most controversial and challenging aspects of pediatric cardiology.

During routine management, it may be useful to measure flow rates in various parts of the systemic venous pathway to make management decisions. This allows measurement of cardiac output and the determination of flow splits to the pulmonary arteries. Measurement of these flows may also prove to be beneficial in planning future surgical reconstructions. Cardiac magnetic resonance imaging (CMR) provides an accurate, noninvasive means of measuring flow rate in each limb of the cavopulmonary connection.[8,9] However, no large series exists that has used CMR to investigates the flow rates and distributions in patients having the Fontan operation. The goal of the study was to determine blood flow distribution and flow rates in these patients using CMR in the inferior and superior venae cavae (IVC and SVC) and the left and right pulmonary arteries (LPA and RPA). These data will serve as a guide for the clinical management of these patients. In addition, the effect of patient age and size, presence or absence of left SVC, and Fontan type on both the IVC contribution to total Fontan flow and LPA/RPA flow splits are determined.

METHODS

Patients

We studied 105 subjects with CMR phase-contrast velocity mapping. They were enrolled at either Children's Hospital of Philadelphia (CHOP) or Children's Healthcare of Atlanta (CHOA) over a 6-year period from March 2001 through June 2007. All protocols were approved by the

From the Division of Cardiology,[a] Children's Hospital of Philadelphia, Philadelphia, Pa; the Department of Biomedical Engineering,[b] Georgia Institute of Technology and Emory University; and the Sibley Heart Center,[c] Children's Health Care of Atlanta, Emory University School of Medicine, Atlanta, Ga.

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Address for reprints: Kevin K. Whitehead, MD, PhD, Cardiology, 2nd Floor Main Bldg, Children's Hospital of Philadelphia, Philadelphia, PA 19104 (E-mail: whiteheadk@email.chop.edu).

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Abbreviations and Acronyms

BSA	= body surface area
CHOA	= Children's Healthcare of Atlanta
CHOP	= Children's Hospital of Philadelphia
CMR	= cardiac magnetic resonance imaging
IVC	= inferior vena cava
LPA	= left pulmonary artery
MRI	= magnetic resonance imaging
RPA	= right pulmonary artery
SVC	= superior vena cava

institutional review boards of both institutions and informed consent was obtained from all participants or their legal guardians.

The mean subject age was 11.5 years, ranging from 2 to 28 years. The mean time from the Fontan operation for the 82 patients for whom the data were available was 8.2 years. The SVC anastomosis and Fontan type were recorded from the medical record for each patient, when available. There were 40 patients with bidirectional Glenn shunts, 53 with hemi-Fontans, and 12 who either had older atriopulmonary-type connections or else had insufficient information to determine the type of SVC connection. The Fontan type included 69 intra-atrial baffles, 28 extracardiac conduits, and 4 classic atriopulmonary connections. Four patients had their operations at outside institutions and their Fontan types were not completely defined. There were 15 patients with bilateral SVCs. The presence of an open fenestration was recorded by the surgical note determining whether there was a surgical fenestration and by the most recent echocardiogram documenting whether the fenestration was still patent.

All patients were required to undergo a 1-hour magnetic resonance imaging (MRI) scan. Older patients performed breath-holds for image acquisition, whereas younger patients or patients unable to cooperate were sedated per institution protocol and averaging was used. No patient had arrhythmias that precluded imaging in the scanner. Patients were excluded if artifact precluded obtaining velocity maps in all 4 or 5 (for bilateral SVCs) limbs of the Fontan pathway.

CMR

All CMR scans were performed at either CHOP or CHOA using either a 1.5-T Siemens Magnetom Avanto (Siemens Medical Systems, Iselin, NJ) or a 1.5-T GE Signa (GE Medical Systems, Milwaukee, Wis). Velocity maps of each venous vessel (SVC, IVC, left SVC) and pulmonary artery (LPA, RPA) were obtained. In addition, aortic outflow was available in 78 patients. Five patients were excluded from the study for artifact in one or both pulmonary arteries.

Phase-Encoded Velocity Mapping

Retrospectively gated, through-plane phase-encoded velocity maps were obtained in the SVC, IVC, RPA, and LPA and in most cases of the proximal ascending aorta. Care was taken to be perpendicular to flow and to obtain slice positions and orientations that were (1) distal to the azygos insertion (when still present) into the SVC, (2) proximal to the RPA upper lobe branching, and (3) distal to the aorta–pulmonary anastomosis (if present). Multiplanar reconstruction was used to set the position and angle of the imaging plane for phase-encoded velocity mapping. A set of sample parameters is given in Table 1.

Data Analysis

An in-house program in Matlab (The Mathworks, Natick, Mass) was used to read and process acquired images. A gradient-based active contour algorithm was implemented for the semiautomatic segmentation of the vessel of interest in all the cardiac phases.[10] A contour was manually outlined around the vessel of interest. This contour would automatically evolve on the basis of gradient-based forces until it identified the vessel boundary for all the cardiac phases. The segmentation was visually inspected for accuracy, and incorrect contours were adjusted manually. The segmented pixel values were converted into velocity values, which were integrated over the entire vessel area for each cardiac phase to extract flow for that phase. Mean flow rates were computed by averaging the flows through all the cardiac phases.

These flows were indexed to body surface area (BSA) for comparison. Fractional contributions of IVC and RPA to total blood flow were calculated and correlated with BSA and age. All population statistics are reported as a mean ± the standard deviation. Differences between statistics are reported as mean difference ± the standard error. Total pulmonary blood flow, total caval blood flow, and aortic flow were compared using a paired Student *t* test. The data were disaggregated by Fontan type, SVC anastomosis type, and the presence or absence of a left SVC. These groups were compared by Student *t* test for independent samples to determine whether flow splits vary by group.

RESULTS

The results of the Fontan flow analysis are summarized in Table 2. The contribution of IVC flow to total systemic venous return was 59% ± 15%. The contribution of RPA flow to total pulmonary blood flow was 55% ± 13%, which was significantly greater than half ($P = .003$). Total pulmonary flow was measured at 2.4 ± 0.7 $L \cdot min^{-1} \cdot m^{-2}$, compared with 2.8 $L \cdot min^{-1} \cdot m^{-2}$ for the measured total caval flow, a difference of 14% ($P < .001$).

In patients with bilateral SVCs, the right SVC accounted for 52% ± 14% of the flow. No difference was noted in the pulmonary flow splits for patients with bilateral SVCs, with 48% ± 19% to the right lung.

A potential source of difference between the systemic venous and pulmonary flows is the presence of a fenestration in the Fontan repair, which would allow a right-to-left shunt before blood reaches the branch pulmonary arteries. To investigate this, the data were disaggregated by the presence or absence of a fenestration at the time of the MRI. Although the ratio of pulmonary to caval blood flow was slightly higher in patients without an open fenestration (0.86 ± 0.24 vs 0.84 ± 0.20), the difference was small and not statistically significant.

Age and BSA were correlated with the IVC fraction of total systemic venous return ($r = 0.60$ and 0.74, respectively; $P < .05$). The IVC fraction appeared to increase in a logarithmic pattern with BSA (Figure 1). Conversely, there was no correlation between RPA fraction of pulmonary blood flow and age or BSA.

Effect of Left SVC and Fontan Type

Table 2 summarizes the comparison of patients with bilateral SVCs with those without, and it compares those with intracardiac versus extracardiac Fontans. There was no significant effect of SVC anastomosis type, Fontan type, or presence of a left SVC on the pulmonary flow splits or the fractional contribution of the IVC to caval flow.

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