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Although all preoperative imaging can be considered surgical planning (SP), it is defined in this article as the act of using preoperative data to simulate the surgical procedure or the result of the procedure. It is the combination, all or in part, of 3-dimensional (3D) medical imaging, applied computer vision, computer-aided design, and computational fluid dynamic (CFD) modeling to mimic and provide visual guidance for surgical procedures. This simulation is generally performed with multiple anatomic and physiological states to determine the robustness of the procedure. There are multiple advantages to this approach such as assessing standard interventions and creation of new surgical strategies without risking the patient’s health; this can potentially have both clinical and economic benefits.

Since its first mention ≈30 years ago, SP is now a routine part of interventions in fields such as neurosurgery and orthopedics. Translating this paradigm to cardiovascular interventions provides not only enhanced 3D visualization but also the potential to interface with physics-driven computational solvers (eg, CFD) to predict the hemodynamic outcomes. Considering the complexity of fluid-solid interactions and the highly time-varying component of the cardiovascular system, these efforts are largely lagging behind those in the neurological and orthopedic communities, but recent advances appear promising.

The Fontan operation for single ventricle (SV) patients, in which a conduit (the total cavopulmonary connection [TCPC]) is placed to channel systemic venous return passively into the pulmonary arteries (PAs), is the paradigm for this approach. This category of lesions is the leading cause of morbidity and mortality in congenital heart disease, and although it is generally associated with acceptable short-term outcomes, Fontan failure remains problematic. Progressive ventricular dysfunction, protein-losing enteropathy, poor exercise tolerance, pulmonary arteriovenous malformations (PA VMs), and liver dysfunction are some of the most commonly cited complications. These morbidities are multifactorial, and the underlying causes in many cases are unknown; however, mounting evidence suggests that TCPC hemodynamics play an important role in their development. For example, exercise intolerance may be related to nonlinear increases in TCPC power loss (PL) with increasing exercise level, which contributes substantially to ventricular preload limitations. The SP approach can yield clues to these complications and potentially avoid them by simulating the multiple TCPC geometries and hemodynamics in a patient-specific fashion to determine the optimal design.

There are 2 main goals to date of SP in the SV patient. One is to minimize the PL in the Fontan baffle (systemic venous pathway). As mentioned, this allows passive flow of blood from the systemic venous return to enter the lungs without the use of a pumping ventricle. Because of this, minimizing the PL at this level is important to allow easier transit of blood into the lungs. A second goal is to distribute an equal amount of hepatic blood flow to both lungs. It is known that some form of an unidentified hepatic factor (HF) inhibits the formation of PA VM (ie, lack of this factor produces PA VMs); in lungs with PA VMs, introduction of hepatic blood flow (and with it the HF) causes the PA VM to regress. Certain types of SV patients are a setup for a lack of HF (bidirectional Glenn and heterotaxy patients), so it is clear that a benefit would accrue if SP could design baffles to maintain HF levels to both lungs (see below). These 2 goals are amenable to SP because the geometry of the Fontan baffle can be altered to minimize energy loss and direct blood appropriately.

The SP Approach

The overall SP procedure as it relates to the Fontan operation and the TCPC is summarized in Figure 1 (top) and contains 4 basic steps with 2 assessment stages. Preoperative imaging is obtained using cardiac magnetic resonance (CMR) for anatomy and flows followed by detailed image processing to determine the current hemodynamics and physiology. Virtual surgery is performed on a workstation in conjunction with bioengineers, cardiologists, and surgeons to determine various options, with CFD subsequently performed to obtain the
physiology and hemodynamics of each option. Finally, the team meets to determine which option is optimal, and then surgery is performed.

The SP approach that we developed requires patient-specific anatomy. Although static steady-state free-precession CMR is the method presented, the reconstruction tools can be applied to different types of CMR (eg, angiography) and imaging modalities (eg, computed tomography), as long as they provide enough anatomic details for segmentation. The following is more detail on each phase of the process.

1. CMR: The minimum imaging required as input for the current process consists of the following:
   - high-resolution stack of cardiac-gated, static axial steady-state free-precession images to reconstruct the bidirectional Glenn or TCPC and surrounding structures; this can be done with or without T2 preparation (eg, whole-heart imaging). Angiography is not optimal because it is not cardiac gated, and the borders of the cardiovascular structures tend to be blurred.
   - Through-plane, retrospectively gated, phase-encoded velocity maps across all vessels of interest, including all cavae, the TCPC immediately inferior to the branch PAs (to account for fenestration flow), hepatic veins if needed, and across the branch PAs at a minimum. In addition, phase-encoded velocity maps are performed across the aortic root, the main PA (if present), the pulmonary veins, and the descending aorta at the level of the diaphragm.
   - Optional imaging includes 4-dimensional flow stack in the coronal plane across the TCPC to determine the in vivo flows and to compare with the calculated CFD flows, which will subsequently be generated.

2. Anatomic reconstruction: Our group has previously developed and extensively used techniques5,6 (Figure 1, bottom) to accurately reconstruct patient anatomy from CMR images that include (1) interpolation, which ensures isotropic sizing for each volume element of the stack, (2) segmentation, which can handle incomplete vessel edge definitions, and (3) 3D reconstruction, which defines a smooth and coherent 3D surface from the segmented stack. Analysis of phase-encoded velocity maps data (velocity segmentation): This methodology uses parametric active contours with gradient vector flow,7 in which the user specifies an initial contour that evolves under the influence of internal and external energy fields to sit precisely on the vessel border. Flow artifacts and noise are eliminated with an automatic, adaptive median filtering approach. Validation against manual segmentation demonstrated an excellent agreement between the 2 methods with <1% difference in resulting flow rates.8
3. CFD: By numerically solving the basic fluid mechanics (Navier-Stokes equations) and mass conservation equations, CFD methods provide flow and pressure fields in a physical domain for given boundary conditions. CFD solver software especially tailored to address the complexities and challenges of modeling the cardiovascular system is used to simulate transient, internal flows in arbitrarily complex shapes such as TCPC anatomies. Through numerous experimental test cases, the time-averaged velocity fields and flow profiles have had excellent agreement.
Figure 3. Surgical planning in a Fontan patient with heterotaxy. A, All 3-dimensional images are viewed from posterior. Original anatomy is at the top, and the 3 virtual surgical options are below. Y graft, azygous (Azyg) connection to the hepatic baffle and hepatic baffle connection to the azygous are shown from left to right. B, Flow structures (top), pressure drop (middle), and hepatic flow (bottom) for the original anatomy (left column) and the 3 surgical options are shown. Arrow points to the vortex formation in the original anatomy. IV indicates innominate vein; IVC, inferior vena cava; LPA, left pulmonary artery; RPA, right pulmonary artery; and SVC, superior vena cava. Reprinted from Sundareswaran et al17 with permission from the publisher. Copyright © 2009, Elsevier Inc. Authorization for this adaptation has been obtained both from the owner of the copyright in the original work and from the owner of copyright in the translation or adaptation.
with both experimental and in vivo imaging data. Control volume PL and hepatic flow distribution are the primary end points from these simulations and are used to evaluate and discriminate between surgical options.

4. Virtual surgery: After the anatomy and phase-encoded velocity maps data are analyzed, CFD is used to determine the current physiology and hemodynamics. Detailed discussions among bioengineers, cardiologists, and surgeons occur to determine the best surgical options to test.

A major milestone for the systematic investigation of alternate Fontan surgical procedures is the ability to modify the preoperative anatomy to model different postoperative configurations. To this end, our group has developed a robust and practical interface that reproduces surgical motions (SURGEM) via 2 free-form haptic devices, which is used to perform this procedure.

Finally, an assessment of all options is done by bioengineers, cardiologists, and surgeons, and the best option is implemented.

5. Lumped parameter and compliance modeling: Lumped parameter models are a widely used tool for simulating the cardiovascular system. By exploiting analogies between fluid flow and electric circuits, systemic responses can be assessed at a global level combining simplified models for each relevant section of the cardiovascular system without the need to model anatomic detail. Recently, such reduced order models have been combined with 3D CFD solvers as a means to improve the accuracy and realism of the prescribed boundary conditions. This coupling strategy can be a particularly valuable tool to naturally approximate the postoperative flow conditions needed to properly evaluate the various surgical options, given the complex hemodynamic changes that occur after Fontan surgery. This is a useful aid.

The Beginnings of SP in Fontan Patients

One of the first attempts at using the SP approach in Fontan patients was fairly elementary: performing virtual angioplasty on a stenotic left PA (LPA). Pekkan et al. noted that in a large cohort of SV patients, 35% had significant narrowing of the LPA attributable to the large patulous aortic reconstructions and hypothesized that relief of this obstruction would

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**Figure 4.** Prediction of pulmonary arteriovenous malformations (PAVMs) in a heterotaxy patient. Time-averaged flow structures in the original cardiac magnetic resonance (top left) and at different phases of the cardiac cycle (top right) with P being the phase and the following number being the phase. Bottom right. The graph of percentage of hepatic blood vs phase, noting that little flow goes to the right pulmonary artery (RPA). Bottom left. The global flow structure at the second cardiac magnetic resonance with hepatic vein flow (HepV) in blue, right superior vena cava (SVC) flow in red, left superior vena cava flow (LSVC) in gold, and azygous (Az) flow in green. Note different color maps for the top left.
significantly improve hemodynamics and PL. By using CFD and modifying the geometry with specialized software, they found not only that left lung perfusion increased (which is no surprise) but also that PL decreased with decreasing severity of the stenosis; this effect was more pronounced with diffuse stenosis than discrete stenosis. There were high PLs (30 mW) with an 85% stenosis, which were cut in half with virtual angioplasty to no narrowing. Adding a fenestration decreased the TCPC pressure but also decreased both left and right lung perfusion and increased hemodynamic PL.

This was followed by another study in which the Fontan baffle was actually moved in a case with both bilateral superior vena cava connected to the ipsilateral PA (Figure 2A and 2B).

In that report, the inferior vena cava (IVC) baffle was connected to the right superior vena cava (RSVC)–right PA (RPA) anastomosis; although there were smooth and steady flow fields in other portions of the systemic venous pathway, flow disturbances and collisions were found at the RSVC-RPA connection with baffle flow. Left superior vena cava flow preferentially streamed to the LPA with the central PAs being perfused by flow from the IVC and RSVC. When simulations were run with <50% of the cardiac output streaming to the LPA, little flow was noted in the central PAs, which would be a setup for thrombus formation (by flow stasis) and left PAVMs because of lack of HF (see below). By modifying the anatomy and shifting the baffle to the central PA, not only was PL decreased by 7%, but also no flow stasis was observed in the central PA, and a substantial amount of IVC blood (and HF) coursed to both lungs. Although not mentioned in this study, CFD modeling would be able to determine the optimal degree of angulation of the Fontan baffle if it was not anatomically possible to place it directly in the central PA or to determine the offset to one branch PA in situations when there was unequal distribution of blood flow.

**SP and PAVMs: Tackling a Difficult Problem**

By definition, PAVMs are a complex tangled web of abnormal arteries and veins connected by >1 fistula in the lung, bypassing the alveoli (no capillary bed of its own). One obvious problem this creates is cyanosis because bypassing the alveoli prevents gas exchange. In addition, the fistulas are a low resistance circuit, drawing blood away from the normal regions of the lung where gas exchange can occur to regions of the lungs where it cannot, creating another dimension to the problem. As mentioned above, an unidentified HF can prevent these PAVMs from occurring, although VEGF, nitric oxide, and other vasoactive substances are also thought to play a role.

It is estimated that 3% of SV patients after bidirectional Glenn have PAVMs that can develop rapidly, although they can resolve just as rapidly after completion of the Fontan procedure, presumably because of delivery of HF to the lungs. Lending credence to the notion of HF is a report by AboulHosn et al in which PAVMs were found in lungs of Fontan patients with discontinuous branch PAs whose blood flow was solely from the SVC; after reconnection, the PAVMs regressed.

SV patients with heterotaxy, specifically those with interrupted IVC with azygous continuation to an SVC, are especially susceptible to PAVM development. In these individuals, because the hepatic veins enter the right atrium directly, a Kawashima operation (ie, a bidirectional Glenn procedure in this setting) directs all blood from the lower and upper body to the PAs except hepatic blood. Therefore, to baffle this blood into the lungs and to complete the Fontan operation, a conduit or baffle must be created. Because hepatic venous flow is only 20% to 25% of the cardiac output, the challenge becomes creating a baffle in such a fashion to distribute hepatic flow equally to both lungs. A baffle too close to one branch PA may stream hepatic blood to that lung, causing the contralateral lung to develop PAVMs. This
becomes an even more complex decision when there are bilateral SVCs, each connected to the ipsilateral branch PA.

The first article describing SP to redirect hepatic venous flow to allow PAVMs to regress was published in 2009 by Sundareswaran et al.17 That case report described an SV patient with heterotaxy, dextrocardia, and interrupted IVC with azygous continuation to the RSVC who, after a Kawashima operation and hepatic extracardiac conduit to the RPA, was noted to have a systemic saturation of 72%. During catheterization, severe left PAVMs were noted, and the patient underwent CMR and SP. The native anatomy and flow structure (Figures 3A and 3B) demonstrated vortex formation in a pouch formed by the SVC-RPA connection, blocking hepatic flow from the baffle to enter the LPA and causing streaming of this blood into the RPA (Figure 3B, left column, arrow). The LPA received 70% of the cardiac output; however, the calculated hepatic flow to the LPA was low (≈5%).

SP strategies included either increasing hepatic flow kinetic energy (unlikely), avoiding head-on collision of hepatic flow with the vortex or disrupting the vortex entirely. After long and lengthy discussions, options considered included a Y graft from hepatic veins, which straddled the pouch, connecting the azygous to the hepatic conduit and connecting the hepatic conduit to the azygous (Figure 3A and 3B). The final option noted, connection of the hepatic conduit to the azygous, demonstrated the best performance with a 66%/34% LPA/RPA split of hepatic flow, close to the global flow distribution of 70%/30% (Figure 3B, right column). This option was performed, and at the 5-month follow-up, the systemic saturation was 94% with presumed resolution of the PAVMs.

De Zélicourt et al2 described our experience using SP with 5 SV patients with interrupted IVC and severe PAVMs in an attempt to channel hepatic blood to both lungs equally. By using CMR for flows and anatomy, global and hepatic flow distributions along with PL were calculated in the current physiological setup, and virtual surgeries were performed to determine the optimal configuration of the hepatic baffle. Because of the low flow rate of hepatic blood, the conduit is sensitive to the offset with the SVC where nearly the entire cardiac output returns to the PAs; even a small offset can lead to a highly asymmetrical hepatic flow distribution. In the 3 patients with a single SVC, because of this fact, the best approach seemed to be a Y graft straddling the SVC-PA connection or a hepatic conduit to azygous connection. In patients with bilateral SVCs, the best approach seemed to be connecting the baffle to the central PA in between the 2 SVCs.

Figure 6. Surgical planning (SP) in a standard Fontan patient; SP is from the patient from Figure 5. A, Eight virtual surgeries (a–e) with varying anatomic geometries. Note that a has a separate baffle for the left hepatic vein and that lower virtual surgeries are all Y grafts; g courses long the left side of the heart. B, Flow structures corresponding to the anatomies in b. Red arrows signify regions of turbulence, whereas the yellow arrow demonstrates that the flow is too slow in the left hepatic baffle to sustain patency. Surgery (b) was chosen as optimal. C, Charts of hepatic flow distribution (top) and power loss (bottom) for all the virtual surgeries at 3 flow splits to the left pulmonary artery (LPA); surgery b seems to have the best profile (green box). mW indicates milliwatts; and PBF, pulmonary blood flow.
Proof of concept is further enhanced by the prediction of development of PA VMs. Figures 4 demonstrates data from a 6-year-old boy with SV, bilateral SVCs, interrupted IVC with azygous continuation to an left superior vena cava flow after a Kawashima operation and hepatic baffle connection to the RSVC-RPA junction. There was significant central PA stenosis and no evidence for PA VMs. The patient was referred for CMR and CFD modeling to determine the amount of hepatic flow to each lung, and results demonstrated that <10% was coursing to the left lung, implying that this lung was at risk for PAVM development. Two years later, the patient’s systemic oxygen saturation had dropped from the mid-90s to the low 80s, and cardiac catheterization revealed significant left lung PAVMs. A repeat CMR and CFD study performed at that time demonstrated <1% hepatic flow coursing to the left lung.

**Beyond PAVMs**

SP has other applications in the SV patient such as in the routine Fontan. Figure 5 contains CMR images from a 3-year-old SV patient with heterotaxy, dextrocardia, infra-diaphragmatic total anomalous pulmonary venous connections, midline liver after bilateral bidirectional Glenn, and pulmonary vein repair. As can be seen, the left hepatic veins entered separately (on the left side of the atrial septum) from the IVC and right hepatic veins (right side of the atrial septum), so connecting these presented a challenge. In addition, the pulmonary veins and pulmonary venous confluence seem to be in the path of a number of possible Fontan conduits. Figure 6A and 6B has 8 possible scenarios explored anatomically and with flow structures, including a conduit coursing up the left side of the cardiac mass. The main conduit for the left hepatic veins and the IVC-right hepatic veins, was thought impractical because of low flow in the left conduit and the risk of thrombosis (orange arrow). All other geometries except b had flow disturbances thought to be less than optimal (red arrows). Figure 6C is the quantitative data for hepatic flow distribution and PL demonstrating that b was the best option as well.

SP can also be used to evaluate new and novel conduit designs. For example, a study published in 2007 proposed the Optiflow design for the systemic venous pathway. In that proposal, the SVC and Fontan conduit connections to the branch PA were suggested to be a diamond-shaped configuration (Figure 7); this allowed lower PL (with the exception of a low flow 2 L/min in the systemic venous pathway relative to a standard Fontan at all flow splits to the RPA. Because this may not be practical in some cases, a Y graft was also proposed and evaluated (Figure 7); this was found to also have better PL and flow structures than the standard Fontan. This was followed by a study by Marsden et al, who also proposed 2 Y-graft variants—off-the-shelf and area-preserving designs—that were evaluated at rest and at exercise; they found that the area-preserving graft had greater efficiency than the other in both conditions, decreased caval pressures at exercise, and better distributed IVC flow. In 2012, that same group demonstrated that HF was better distributed by the Y design than either the classic T junction or an offset of the baffle from the SVC.

**Smoke and Mirrors?**

The images are appealing and SP seems useful clinically, but do the flow structures and hemodynamics have any basis in reality, or is this just smoke and mirrors? We answered that question in 3 different ways, all of which demonstrate that CFD and SP faithfully describe what occurs in vivo:

1. A qualitative comparison of the flow structure between angiography and what CFD predicts (Figure 8A)
2. Comparison between the flow structure in vivo as directly measured by CMR 4-dimensional flow imaging and what CFD predicts (Figure 8B, left).
3. Quantitative comparison between CFD flow structure in SP-designed Fontan conduits and the superimposition of the preoperative boundary conditions (flows at the inlets and outlets) on the postoperative anatomy the surgeon
actually created; the resultant flows were close to each other (Figure 8B, right).

**Implications and Conclusion**

The use of SP in designing, redesigning, and building a better Fontan uses CMR for anatomy and flows along with CFD and advanced bioengineering and computing environments. It has been used successfully and most extensively in SV patients with PAVMs to direct hepatic blood flow evenly to both lungs and to predict PAVMs; however, it is currently being extended to the routine Fontan patient along with testing out and designing more optimized Fontan geometries. More work is definitely needed to refine the process to make it more robust, quicker, and easier; at the moment, few centers are capable of this technology. In the future, SP will most likely be extended to other surgical lesions. SP holds the promise of truly individualized medicine and surgery, optimizing surgical outcome virtually and therefore minimizing the risk to the patient.

**Sources of Funding**

This work was partially funded by 2 National Institutes of Health grants: the Bioengineering Research Partnership R01 HL67622-01 and R01 HL098252-01.

**References**


**Disclosures**

Dr Fogel has a grant from Edwards Life Sciences and Siemens Medical Solutions and is a medical monitor for an imaging agent from Kereos. The other authors report no conflicts.


**KEY WORDS:** Fontan procedure ■ magnetic resonance imaging
Imaging for Preintervention Planning: Pre- and Post-Fontan Procedures
Mark A. Fogel, Reza H. Khiabani and Ajit Yoganathan

doi: 10.1161/CIRCIMAGING.113.000335
Circulation: Cardiovascular Imaging is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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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:
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