The first cohort of prospective Fontan surgical planning patients with follow-up data: How accurate is surgical planning?

Phillip M. Trusty, MS,^a Zhenglun Alan Wei, PhD,^a Timothy C. Slesnick, MD,^b Kirk R. Kanter, MD,^c Thomas L. Spray, MD,^d Mark A. Fogel, MD,^e and Ajit P. Yoganathan, PhD^a

ABSTRACT

Objective: Fontan surgical planning is an image-based, collaborative effort, which is hypothesized to result in improved patient outcomes. A common motivation for Fontan surgical planning is the progression (or concern for progression) of pulmonary arteriovenous malformations. The purpose of this study was to evaluate the accuracy of surgical planning predictions, specifically hepatic flow distribution (HFD), a known factor in pulmonary arteriovenous malformation progression, and identify methodological improvements needed to increase prediction accuracy.

Methods: Twelve single-ventricle patients who were enrolled in a surgical planning protocol for Fontan surgery with pre- and postoperative cardiac imaging were included in this study. Computational fluid dynamics were used to compare HFD in the surgical planning prediction and actual postoperative conditions.

Results: Overall, HFD prediction error was $17 \pm 13\%$. This error was similar between surgery types ($15 \pm 18\%$ and $18 \pm 10\%$ for revisions vs Fontan completions respectively; $P = .73$), but was significantly lower ($6 \pm 7\%$; $P = .05$) for hepatic to azygous shunts. Y-grafts and extracardiac conduits showed a strong correlation between prediction error and discrepancies in graft insertion points ($r = 0.99$; $P < .001$). Improving postoperative anatomy prediction significantly reduced overall HFD prediction error to $9 \pm 6\%$ ($P = .03$).

Conclusions: Although Fontan surgical planning can offer accurate HFD predictions for specific graft types, methodological improvements are needed to increase overall accuracy. Specifically, improving postoperative anatomy prediction was shown to be an important target for future work. Future efforts and refinements to the surgical planning process will benefit from an improved understanding of the current state and will rely heavily on increased follow-up data. (J Thorac Cardiovasc Surg 2019;157:1146-55)

Comparison of predicted and postoperative hepatic flow distribution (indicated by percentage) for a representative patient. The overlay shows predicted (yellow) and postoperative (purple) anatomies.

Central Message

Fontan surgical planning accurately predicts hepatic flow distribution for specific graft types and will benefit from improved anatomical predictions.

Perspective

Fontan surgical planning can provide accurate predictions of hepatic flow distribution for specific graft types. Anatomical prediction was shown to be a key methodological shortcoming in the surgical planning process. With continued improvements, surgical planning might be a useful tool to avoid pulmonary arteriovenous malformations in Fontan patients.

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From the ^aWallace H. Coulter Department of Biomedical Engineering, Georgia Institute of Technology and Emory University, Atlanta, Ga; ^bDivision of Cardiology, Department of Pediatrics, ^cDivision of Cardiothoracic Surgery, Children's Healthcare of Atlanta, Emory University School of Medicine, Atlanta, Ga; and ^dDivision of Pediatric Cardiothoracic Surgery, and ^eDivision of Cardiology, Children's Hospital of Philadelphia, Philadelphia, Pa.

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Address for reprints: Ajit P. Yoganathan, PhD, 387 Technology Circle NW, Suite 200, Atlanta, GA 30313 (E-mail: ajit.yoganathan@bme.gatech.edu).

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Fontan surgical planning is an image-based, collaborative effort, which is hypothesized to result in improved patient outcomes. The general process has been detailed in multiple publications and has been implemented for select cases over the past decade.¹⁻⁴ Despite its use in dozens of cases, few follow-up data have been available to evaluate the accuracy of surgical planning predictions.

Scanning this QR code will take you to the article title page to access supplementary information.

Abbreviations and Acronyms

CMR	= cardiac magnetic resonance
HFD	= hepatic flow distribution
IVC	= inferior vena cava
PAVM	= pulmonary arteriovenous malformation
TCPC	= total cavopulmonary connection

A common motivation for Fontan surgical planning is the progression (or concern for progression) of pulmonary arteriovenous malformations (PAVMs). PAVMs are extremely rare in the average population (2/100,000), but are much more common in Fontan patients in whom a poorly designed total cavopulmonary connection (TCPC) might lead to unbalanced hepatic flow distribution (HFD), a known factor in PAVM formation/progression.⁵⁻¹¹ This type of surgical planning can involve Fontan revision cases (a previous Fontan surgery resulted in PAVMs and therefore must be revised) as well as Fontan completion cases (stage 2-3, often performed in patients with more complex anatomies and hence the need for added insight to determine the best surgical strategy). In either case, the purpose of Fontan surgical planning is to determine which TCPC design will result in balanced HFD. The goal to optimize HFD has led to a variety of Fontan connections.

Although limited to case studies and relatively short-term follow-up data, several previous studies have provided a preliminary understanding of surgical planning prediction accuracy.² Sundareswaran and colleagues published the first use of surgical planning to correct PAVMs (determined by an increase in oxygen saturation) in 2009.¹² Unfortunately, this single-patient case report included no postoperative imaging data to compare the predicted and postoperative HFD. Haggerty and colleagues provided the most thorough study to date, which included only 4 patients with short follow-up times (2 patients less than 1 month).¹³ In their study HFD and graft resistance were compared and showed “sufficient agreement” between the predicted and postoperative states. However, the use of steady flow conditions and fixed outlet flow splits, as well as the small sample size, limit the generalizability of these findings.

The purpose of this study was to evaluate the accuracy of Fontan surgical planning predictions and identify which methodological improvements are needed to improve prediction accuracy. Specifically, in this study we focused on the prediction of HFD. Pre- and postoperative imaging data from patients enrolled in the surgical planning process were used to address these questions. This study offers, to our knowledge, the first assessment of prediction accuracy and methodological shortcomings for Fontan revisions and Fontan completion cases using longer-term follow-up data not previously available.

METHODS

Patient Selection

A total of 12 single-ventricle patients were included in this study. All patient data were received from the Children’s Hospital of Philadelphia and Children’s Healthcare of Atlanta under institutional review board approval (H17434 and H09279, respectively) with a waiver of consent. Inclusion criteria were that the patient: (1) was enrolled in the surgical planning process before surgery; (2) had pre- and postoperative cardiac magnetic resonance (CMR) and phase contrast CMR imaging; and (3) imaging quality was sufficient for accurate anatomical segmentation as well as flow segmentation at every TCPC inlet/outlet. This resulted in Fontan revision cases (n = 5) and Fontan completion cases (n = 7). All patients in this study were enrolled in surgical planning because of PAVM development or the concern for PAVM development due to atypical vasculature or clinical history. Clinical data including age, sex, body surface area, imaging and surgery dates, and diagnosis were obtained for each patient.

Magnetic Resonance Imaging Acquisition

All CMR imaging was performed with a Siemens 1.5 T magnetic resonance imaging system (Siemens Medical Solutions, Malvern, Pa). Patients were scanned supine, head first in the scanner with electrocardiogram leads placed. After localizers were obtained, a stack of contiguous, static, diastolic steady state free precession images were obtained from the diaphragm to thoracic inlet to assess anatomy and provide inputs for computational fluid dynamics modeling. Slice thickness was generally 3–4 mm and in plane resolution was 1 × 1 mm.

Through plane, retrospectively gated, phase contrast magnetic resonance imaging was used to assess flow in the cavae, branch pulmonary arteries, and across the aortic valve. Inferior vena cava (IVC) flow was measured supra-hepatically. Velocity encoding was generally 150 cm/s for the aorta and 60 cm/s for the other vessels (superior vena cava, Fontan baffle, right pulmonary artery [RPA], and left pulmonary artery [LPA]). Slice thickness was generally 3 mm with in-plane resolution of 1 × 1 mm. The number of phases was a function of the heart rate and ranged from 20 to 30. Postoperative CMR images were acquired at the follow-up times specified in Table 1.

Anatomic Reconstruction and Blood Flow Segmentation

Patient-specific anatomies were reconstructed from axial CMR images using methods previously developed.^{14,15} Geomagic Studio (Geomagic Inc, Research Triangle Park, NC) was used to fit a surface around the reconstructed point-cloud and export the surface for mesh generation. Patient-specific blood flow waveforms were segmented from phase contrast magnetic resonance images for all vessels of interest using previously validated methods.^{16,17} Pulmonary flow distribution was calculated as:

$$PFD = \frac{Q_{LPA}}{Q_{LPA} + Q_{RPA}}$$

where Q is flow rate. Changes in flows from pre- to postoperative were calculated as:

Flow Change = |Postoperative Q – Preoperative Q| to show absolute magnitude changes in flow rates.

Surgical Planning Prediction

For complex Fontan cases, surgical planning has been used to compare fundamentally different surgical options (extracardiac conduit vs Y-graft vs hepatic to azygous, etc). In addition, this process is used to determine which location on the pulmonary arteries the graft should be anastomosed. SURGEM III, a solid modeling software designed specifically for Fontan surgical planning was used to generate the surgical planning anatomy

TABLE 1. Demographic and surgical data

Patient	Sex	Diagnosis	Surgery type	PAVMs*	Surgical option	Age at surgery, years	Age at follow-up, years	Follow-up, months	HFD prediction, % LPA	Postoperative HFD, % LPA	HFD error
1	F	H, PA	Revision	Left	Hep to AZ	4.7	12.8	97.8	60	56	4
2	F	H	Revision	Left	Y-graft	19.0	19.0	0.3	38	45	7
3	M	H, D	Revision	Right	Hep to AZ	11.6	15.0	40.4	100	100	0
4	F	U	Revision	Right	Y-graft	12.7	12.8	1.6	53	32	21
5	M	H, PA	Revision	Left	ECC	17.5	18.3	9.0	26	71	45
6	M	HLHS	Fontan completion	–	ECC	1.3	4.3	35.9	17	51	34
7	M	H, HLHS	Fontan completion	–	Y-graft	2.6	2.6	0.2	27	48	21
8	F	PA, TH	Fontan completion	–	Y-graft	3.0	8.3	63.7	60	77	17
9	M	PA, DILV	Fontan completion	–	ECC	2.2	2.2	0.3	73	80	7
10	F	H	Fontan completion	–	Hep to AZ	1.1	1.1	0.3	51	56	5
11	F	H, DORV	Fontan completion	–	Hep to AZ	3.2	4.2	11.7	71	87	16
12	F	H, HLHS	Fontan completion	–	Hep to Inn	1.4	1.5	2.1	48	25	23

PAVM, Pulmonary arteriovenous malformation; HFD, hepatic flow distribution; LPA, left pulmonary artery; F, female; H, heterotaxy; PA, pulmonary atresia; Hep to AZ, hepatic to azygous shunt; M, male; D, dextrocardia; U, unbalanced canal; ECC, extracardiac conduit; HLHS, hypoplastic left heart syndrome; TH, tricuspid hypoplasia; DILV, double-inlet left ventricle; DORV, double-outlet right ventricle; Hep to Inn, hepatic to innominate vein. *Data indicate the lung that contained the malformations.

prediction.¹⁸ First, the preoperative TCPC (for Fontan revisions) or bidirectional Glenn (for Fontan completion) and hepatics were imported. With input from the respective clinician, the desired surgical option was then created and exported as a surface mesh. Preoperative flow waveforms reconstructed from phase contrast–magnetic resonance images were directly used as the “predicted” flow waveforms. This technique makes the simplifying assumption that postoperative flows will be identical to preoperative flows. It is important to mention that during the surgical planning process, multiple surgical options are created. However, for the sake of comparing surgical planning predictions with postoperative data, only the prediction of the actual implemented surgical option is considered.

Computational Fluid Dynamics

The 3-D anatomy (from either the preoperative scan, surgical planning prediction, or postoperative scan) was imported into ANSYS Workbench (ANSYS Inc, Canonsburg, Pa), where vessel extensions of length $10 \times$ (vessel diameter) were added to overcome entrance effects and establish an appropriate velocity profile. A polyhedral mesh of approximately $D_{IVC}/20$ -mm elements was used to achieve mesh-independent results, where D_{IVC} is the diameter of the IVC.¹⁹ All simulations were performed using ANSYS Fluent (Release 17.1; ANSYS Inc), which is a finite volume pressure-based Navier–Stokes solver. Blood was modeled as a single-phase Newtonian fluid ($\mu = 0.04$ g/cm·s; $\rho = 1.06$ g/cm³). The appropriate patient-specific blood flow waveforms extracted from phase contrast–magnetic resonance imaging were used as boundary conditions for each TCPC inlet and outlet.^{19,20} Twenty cardiac cycles were simulated to overcome transition effects and achieve period stability, using the final cycle for data analysis.

To investigate the potential accuracy of surgical planning if methodological improvements allowed for more accurate anatomy and flow predictions, 2 additional simulations were run representing either “improved” anatomy or flow predictions. This resulted in 4 simulations for each patient:

1. Predicted (simulation uses predicted anatomy and predicted flows);
2. Actual postoperative (simulation uses postoperative anatomy and postoperative flows);
3. “Improved” anatomy prediction (simulation uses actual postoperative anatomy and predicted flows). The postoperative anatomy represents a perfect anatomical prediction from the surgical planning process; and

4. “Improved” flow prediction (simulation uses predicted anatomy and actual postoperative flows). The postoperative flows represent a perfect flow prediction from the surgical planning process.

In addition, preoperative simulations (preoperative anatomy and preoperative flows) were also run for Fontan revision cases. This allowed an investigation of the relationship between preoperative HFD and PAVM progression.

HFD

HFD was quantified by seeding massless particles at the IVC and calculating the total flux of particles leaving the left and right pulmonary arteries. HFD was defined as:

$$HFD = \frac{\theta_{LPA}}{\theta_{LPA} + \theta_{RPA}}$$

where θ is the total flux of particles throughout a cardiac cycle. The error in HFD prediction is defined as:

$$\text{Error in HFD prediction} = |HFD_{\text{post-op}} - HFD_{\text{predicted}}|.$$

Anatomy Comparison

To compare the predicted and postoperative anatomies, the TCPCs were first registered to account for differences in imaging coordinate systems. A mesh comparison software (CloudCompare, version 2.10) was then used to quantify average and maximum deviations between the surfaces of the 2 TCPCs. This was done for the full TCPC and the graft alone. Graft insertion offset was calculated by measuring the distance between the predicted and postoperative anastomosis locations (distance between center points of each anastomosis). For Y-grafts, the largest insertion offset of the 2 branches was used.

Statistical Analysis

SPSS version 25 (IBM Corp, Armonk, NY) was used for statistical analyses. The Shapiro–Wilk test was used to determine normality for each parameter. Pearson and Spearman correlations were used to investigate bivariate correlations between HFD prediction error and clinical, hemodynamic, and anatomic parameters for linear and monotonic

TABLE 2. Fontan revision and Fontan completion comparison

Variable	Revision	Fontan completion	P value
Age at surgery, y	13.1 ± 5.7 (12.7-10.1)	2.1 ± 0.9 (2.2-1.7)	<.001
Age at follow-up, y	15.6 ± 2.9 (15.0-5.8)	3.5 ± 2.5 (2.6-2.8)	<.001
Follow-up time, mo	30 ± 41 (9-68)	16 ± 25 (2-64)	.492
HFD prediction error	15 ± 18 (7-31)	18 ± 10 (17-16)	.795
IVC flow change, L/min	0.22 ± 0.16 (0.15-0.32)	0.25 ± 0.19 (0.17-0.33)	.782
SVC flow change, L/min	0.35 ± 0.13 (0.4-0.24)	0.44 ± 0.32 (0.34-0.56)	.573
AZ flow change, L/min	0.61 ± 0.45 (0.63-0.82)	0.20 ± 0.17 (0.2-0.32)	.104
LPA flow change, L/min	0.57 ± 0.19 (0.57-0.32)	0.48 ± 0.37 (0.35-0.67)	.638
RPA flow change, L/min	0.80 ± 0.60 (0.91-1.13)	0.25 ± 0.22 (0.14-0.39)	.052
IVC flow change, %	42 ± 42 (35-62)	41 ± 26 (32-35)	.955
SVC flow change, %	36 ± 21 (35-40)	48 ± 29 (52-44)	.462
AZ flow change, %	49 ± 30 (51-58)	40 ± 28 (48-52)	.656
LPA flow change, %	38 ± 21 (30-41)	59 ± 35 (64-38)	.274
RPA flow change, %	62 ± 40 (55-58)	41 ± 53 (19-32)	.471
PFD change, %	9 ± 9 (4-14)	14 ± 6 (13-12)	.261
TCPC deviation, mm	3.3 ± 0.8 (3-1.4)	1.6 ± 0.5 (1.5-0.9)	.001
TCPC max deviation, mm	17.6 ± 2.0 (19-3.5)	9.2 ± 3.7 (9.4-4.1)	.004
Graft deviation, mm	5.6 ± 2.8 (6.3-5.1)	2.8 ± 1.9 (2.7-3.0)	.062
Graft max deviation, mm	15.3 ± 4.4 (15.5-8.3)	9.3 ± 4.5 (9.4-6.2)	.046
Graft insertion offset, mm	13.5 ± 8.3 (15.0-16.1)	5.8 ± 4.5 (6.8-7.5)	.063

Data are presented as mean ± standard deviation (interquartile range). Change represents absolute difference between preoperative and postoperative flows. HFD, Hepatic flow distribution; IVC, inferior vena cava; SVC, superior vena cava; AZ, azygous vein; LPA, left pulmonary artery; RPA, right pulmonary artery; PFD, pulmonary flow distribution; TCPC, total cavopulmonary connection.

relationships, respectively. Depending on normality, either a Wilcoxon rank sum test or a 2-sample *t* test was used to test for equal medians between the revision and Fontan completion groups, as well as between various surgical connection types. A paired sample *t* test was used to test for differences between surgical planning methodologies (current vs improved anatomy/flow). Statistical significance was determined using *P* < .05. Values are shown as average ± standard deviation (median, interquartile range).

RESULTS

Clinical Data

Clinical and surgical data are given in Table 1. The cohort consisted of 5 Fontan revisions and 7 Fontan completion cases. Implemented surgical options included 4 hepatic to azygous shunts, 4 Y-grafts, 3 traditional extracardiac conduits, and 1 hepatic to innominate vein connection. Average follow-up time was 22 ± 32 [6, 39] months.

Revisions Versus Fontan Completions

Age at surgery and age at follow-up were significantly different between the revision (13.1 ± 5.7 [12.7, 10.1] and 15.6 ± 2.9 [15.0, 5.8] years) and Fontan completion (2.1 ± 0.9 [2.2, 1.7] and 3.5 ± 2.5 [2.6, 2.8] years) cases respectively (*P* < .001 for both; Table 2). Follow-up time was not significantly different between the revision and Fontan completion cases (30 ± 41 [9, 68] and 16 ± 25

[2, 64] months respectively; *P* = .49, Table 2). No significant differences in pre- to postoperative changes in flow rates were seen between the revision and Fontan completion cases (Table 2). Additionally, no flow rates (grouped according to surgery type or vessel) showed consistent directionality in flow rate changes. Significant differences in geometric variations (between the predicted and actual postoperative anatomy) were seen between the revision and Fontan completion cases in terms of the TCPC as a whole and the graft alone (Table 2).

HFD Prediction Error

The predicted and postoperative HFD can be found in Table 1. Overall, the HFD prediction error was 17 ± 13 [17, 17]%, and was not significantly different between revisions (15 ± 18 [7, 31]%) and Fontan completion (18 ± 10 [17, 16]%) cases (*P* = .73; Table 2). Fontan completion predictions underestimated HFD in 6 of 7 cases, whereas revisions were evenly split between overestimations and underestimations. Computational fluid dynamics results comparing the predicted and postoperative streamlines for all Fontan revisions and Fontan completion cases are shown in Figures 1 and 2, respectively. In addition, Figure 1 shows the preoperative HFD for all Fontan revisions, confirming a lack of hepatic flow to the lung with PAVMs. Overall, no

FIGURE 1. Streamline comparison for the preoperative (*Pre-Op*), predicted, and postoperative (*Post-Op*) states for all Fontan revision cases. Patients 1, 2, and 5 were diagnosed with left-sided pulmonary arteriovenous malformations (PAVMs), and patients 3 and 4 with right-sided PAVMs. PAVMs regressed in each case when the revision resulted in increased hepatic flow to the affected lung. Hepatic flow distribution is noted as the percent of hepatic flow distribution to the left pulmonary artery (*LPA*). *SVC*, Superior vena cava; *IN*, innominate vein; *RPA*, right pulmonary artery; *AZ*, azygous vein; *FC*, Fontan conduit; *LSVC*, left superior vena cava.

FIGURE 2. Streamline comparison between the predicted and postoperative (*Post-Op*) states for all Fontan completion patients. Hepatic flow distribution is noted as the percent of hepatic flow distribution to the left pulmonary artery (*LPA*). The overlay column shows a comparison of the predicted (*yellow*) and *Post-Op* (*purple*) anatomies. *SVC*, Superior vena cava; *RPA*, right pulmonary artery; *FC*, Fontan conduit; *AZ*, azygous vein; *LSVC*, left superior vena cava; *IN*, innominate vein.

FIGURE 3. Effect of connection type on hepatic flow distribution (HFD) prediction error. A, High variability in prediction error was observed within and across graft types. B, Hepatic to azygous (*Hep to AZ*) shunts showed significantly lower HFD prediction errors than other connection types. ECC, Extracardiac conduit.

significant correlations were found between HFD prediction error and age at surgery, age at follow-up, or follow-up time. Moderate correlations were seen between the percent change in IVC flow rate ($r = 0.60$; $P = .04$) and the change in pulmonary flow distribution ($r = 0.60$; $P = .04$) from pre- to postoperative with HFD prediction error.

Connection Types

HFD prediction error was associated with surgical connection type. A comparison of HFD prediction error between graft types is shown in Figure 3. Hepatic to azygous shunts had significantly lower prediction errors than other connection types (6 ± 7 [5, 12]% vs 22 ± 13 [21, 22]% respectively; $P = .05$; Figure 3, B). In addition, a strong, positive correlation was seen between HFD prediction error

and graft insertion offset within the Y-graft and extracardiac conduit groups ($r = 0.99$; $P < .001$; Figure 4, D). Example cases of low, moderate, and high graft insertion offsets are shown in Figure 4, A-C.

Potential Methodological Improvements

The HFD prediction error associated with the “improved” postoperative anatomy and flow predictions and the current methodology is shown in Table 3. Although a reduced or identical prediction error was seen for most patients using either methodological improvement (8 of 12 and 7 of 12 for the improved anatomy and improved flow scenarios, respectively), a more substantial reduction in error was seen when the improved anatomy was used. The current HFD prediction error (17 ± 13 [17, 17]%) was significantly reduced by improving anatomy prediction

FIGURE 4. Relationship between hepatic flow distribution (HFD) prediction error and graft insertion offset for extracardiac conduit and Y-graft Fontan connections. Graft insertion offset describes the distance between the predicted and implemented graft insertion locations. Representative cases are shown for (A) low, (B) moderate, and (C) high graft insertion offsets. A strong correlation (D) was seen between prediction error and offset for these connection types. The overlay figures (panels A-C) show a comparison of the predicted (blue) and postoperative (red) total cavopulmonary connections on the left, and show a color map of the offset between the predicted and postoperative grafts on the right for each representative case. All cases use the same color scale.

TABLE 3. Comparison of HFD prediction errors between the current surgical planning process and potential methodological improvements

Patient	Surgery type	Vessels present	Connection type	HFD prediction error		
				Current	Improved anatomy prediction	Improved flow prediction
1	Revision	AZ, INN	Hep to AZ	4	6	11
2	Revision	AZ	Y-graft	7	13	1
3	Revision	AZ, LSVC	Hep to AZ	0	0	0
4	Revision	–	Y-graft	21	10	29
5	Revision	AZ	ECC	45	16	58
6	Fontan completion	–	ECC	34	18	18
7	Fontan completion	AZ	Y-graft	21	7	38
8	Fontan completion	AZ	Y-graft	17	6	14
9	Fontan completion	LSVC	ECC	7	13	5
10	Fontan completion	AZ	Hep to AZ	5	2	3
11	Fontan completion	AZ, LSVC	Hep to AZ	16	3	23
12	Fontan completion	AZ, INN	Hep to Inn	23	18	13
Average	–	–	–	17 ± 13 (17-17)	9 ± 6 (9-12)*	18 ± 17 (14-24)

The presence of any nonstandard vessels (in addition to the inferior/superior vena cava and left/right pulmonary artery) is indicated in the vessels present column. *HFD*, Hepatic flow distribution; *AZ*, azygous shunt; *INN*, innominate vein; *Hep to AZ*, hepatic to azygous shunt; *LSVC*, left superior vena cava; *ECC*, extracardiac conduit; *Hep to Inn*, hepatic to innominate connection. *Statistically significant difference (paired sample *t* test) from the current method.

(9 ± 6 [9, 12]%; *P* = .03, paired sample *t* test), but remained nearly the same when only an improved flow prediction was used (18 ± 17 [14, 24]%; *P* = .73, paired sample *t* test). When the 2 potential methodological improvements were compared, improved postoperative anatomy prediction resulted in equivalent or more accurate HFD predictions for 9 of 12 patients compared with improved flow prediction.

DISCUSSION

Previously limited by a lack of postoperative data, the current study offers, to our knowledge, the first assessment of prospective Fontan surgical planning accuracy for Fontan revisions and Fontan completions using medium-term postoperative data. This study incorporates a unique data set resulting from more than a decade of surgical planning experience and provides a methodological assessment necessary for the improvement of surgical planning accuracy.

Although an exact “cutoff” for HFD to prevent PAVMs is currently unknown, and might vary between patients, the Fontan revision results (Table 1, Figure 1) confirm a lack of hepatic flow to the lung with PAVMs in each case. Furthermore, PAVMs regressed in each case when the revision resulted in increased hepatic flow to the affected lung. In combination with previous studies, these results emphasize the importance of achieving a balanced HFD through appropriate TCPC design.

In this study, HFD prediction error averaged 17 ± 13 [17, 17]% across all patients. Prediction error was similar between revision and Fontan completion cases, but differed across connection types. Intuitively, hepatic to azygous connections are more robust to variations in surgical implementation because all hepatic flow will join the azygous flow and then travel through the entire azygous vein before

interacting with other flows regardless of exact placement of the shunt. No colliding flows from multiple vessels are present locally in hepatic to azygous connections, in contrast with Y-graft and extracardiac conduit connections where slight offsets might substantially change the interactions between various inlets and therefore stray from predicted results (Figure 4, D).²¹ Therefore, hepatic to azygous predictions were quite accurate, whereas predictions for other connection types were more varied.

Methodologic Improvements

Capitalizing on the available postoperative data, various simulations were run using the postoperative anatomy or flows as a surrogate for an “improved” prediction. Although it is unlikely that anatomy or flow prediction techniques will ever produce exact matches to postoperative outcomes, this analysis is instructive by showing the full potential of surgical planning accuracy if methodological improvements in either of these areas offered extremely accurate predictions. To reiterate, improvements in anatomy prediction led to a significant (*P* = .03) reduction in HFD prediction error. Interestingly, improvements in flow prediction did not result in similar error reduction (Table 3). These findings stress 2 critical points: (1) postoperative anatomy prediction is a primary factor in HFD prediction; and (2) anatomy prediction methods must improve for Fontan surgical planning to provide more accurate HFD predictions.

Again, improved flow prediction alone did not result in more accurate HFD predictions (*P* = .73; Table 3). In general, HFD is primarily driven by graft placement.²² In a complex connection such as the TCPC, relatively small offsets in graft placement and angulation can largely alter the collisions and interactions among flows from various

VIDEO 1. Discussion of surgical planning predictions, accuracy, and implications by Dr Timothy Slesnick. Video available at: [https://www.jtcvs.org/article/S0022-5223\(18\)33252-5/fulltext](https://www.jtcvs.org/article/S0022-5223(18)33252-5/fulltext).

vessels.²² These variations can affect the preferential streaming of inlet flows including hepatic flow, which in turn will determine HFD. Naturally, severe changes in individual flow rates can affect HFD prediction; however, this was not observed in this cohort.

Though it is common to use indexed flow rates (normalized for body surface area) in pediatric studies involving changes over time, raw flow rates are shown in this study (Table 2) to emphasize the changes in actual input to the surgical planning process. Although an indexed flow rate might remain constant over a several-year follow-up, the actual flow rate (and therefore the flow rate that needs to be predicted) does not. These raw data better represent how boundary conditions for the surgical planning process change over time.

Multiple methods exist to predict postoperative Fontan anatomies and flows, ranging from simple (basic computer-aided design software and using preoperative flows as the “predicted” postoperative flows) to more sophisticated (designated surgical planning software and lumped parameter modeling) methods.^{18,23} As the methods have progressed in complexity, anatomy prediction methods have become faster (software is designed specifically for Fontan surgical planning) and flow prediction methods have become slower (more complex calculations and “full body” modeling). Meeting the clinical timeline for most surgical planning cases requires accelerated analysis.¹ Conveniently, the present results indicate that anatomy prediction is a primary shortcoming, which can hopefully be improved without lengthening the surgical planning process.

Accurate anatomy prediction involves predicting a viable surgical option and accurately implementing that option. Modeling a viable surgical option is heavily dependent on high-quality imaging data, clinician involvement, and inclusion of relevant anatomical landmarks. Current methods include the heart, aorta, and pulmonary circulation, but future efforts could potentially use additional organs and the process of chest closure. When a surgeon has selected the surgical option to implement,

closely replicating that option in vivo might be challenging. Little intraoperative guidance is currently offered as part of the surgical planning process. Some efforts have explored 3-D printing and augmented/virtual reality as planning/guidance tools, but further refinements are needed.²⁴⁻²⁷ In addition, growth is another difficult factor to model that might be necessary to improve surgical planning results.^{28,29} It is possible that “variations” between a predicted and postoperative anatomy are because of growth rather than “imperfect” surgical implementation.

Finally, patient 3 is a unique case that deserves attention. As shown in Figure 1, this patient had 100% HFD to the left lung in the preoperative state. This led to right-sided PAVMs and the need for a Fontan revision. Also shown in Figure 1, the surgical option implemented during the Fontan revision similarly resulted in 100% HFD to the left lung. This was predicted during the surgical planning process but implemented nonetheless because of other concerns. Importantly, not all surgical options for patient 3 resulted in 100% HFD to the left lung. This case illustrates the importance of being able to predict which options will perform poorly in addition to which ones perform well. This case also shows that surgical planning predictions are not always the main determinant for decision-making in current clinical practice. Assessing how surgical planning affects clinical decision-making and patient outcomes is an important next step when surgical planning predictions are known to offer accurate results.

If Fontan surgical planning is to be used in clinical practice, the importance and necessity of follow-up data and validation studies such as this cannot be overstated. Understanding the current accuracy and methodological shortcomings is imperative to correctly use the results and progress the surgical planning process. Future efforts and refinements to the surgical planning process will greatly benefit from an improved understanding of the current state of the art (Video 1).

Limitations

Although important conclusions can be drawn from this study, a more complete understanding of surgical planning accuracy and methodological needs with increased statistical power will require a substantial amount of data most likely through a multicenter study. Additionally, the current study includes a broad range of follow-up times and offers only a “snapshot” of the patients’ postoperative hemodynamics. If available, the inclusion of serial data in a similar study could offer a better understanding of hemodynamic changes over time. The predicted results in this study are representative of the specific surgical planning process used. Results might vary with other prediction techniques. However, this study used one of the most advanced anatomy prediction methods and we conclude that postoperative anatomy prediction is a

limiting factor, which is unlikely to change on the basis of prediction technique. Finally, 2 experienced surgeons were involved in these surgical planning cases. Although we saw no differences in prediction accuracy between the 2 surgeons, it is possible that results might vary on the basis of the surgeon involved. Finally, the results from this study do not indicate how the surgical strategy changed because of the surgical planning process. A surgical strategy was not developed before the surgical planning process for each case to determine how surgical planning influenced the original plan. Therefore, these results report prediction accuracy and do not quantify the effect on clinical decision-making.

CONCLUSIONS

Overall, HFD prediction error was $17 \pm 13\%$. This error was similar between Fontan revisions and Fontan completions, but varied across surgical connection types. Although Fontan surgical planning can offer accurate HFD predictions for specific graft types, methodological improvements are needed to increase overall accuracy. Specifically, improving postoperative anatomy prediction was shown to be an important target for future efforts that would substantially improve flow field modeling, and therefore increase HFD prediction accuracy. Future efforts and refinements to the surgical planning process will greatly benefit from an improved understanding of the current state of the art and will rely heavily on increased follow-up data.

Conflict of Interest Statement

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Key Words: Fontan, congenital heart defect, surgical planning, hepatic flow distribution, preprocedural planning, pulmonary arteriovenous malformations