A new diagnostic algorithm for assessment of patients with single ventricle before a Fontan operation

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Objectives: Cardiac catheterization has a low diagnostic yield before a Fontan operation, and magnetic resonance imaging and computed tomography are reliable alternatives to invasive angiography. A new diagnostic algorithm to avoid cardiac catheterization in ''low-risk'' subjects before a Fontan operation is proposed.

Methods: The proposed algorithm would identify ''high-risk'' subjects on the basis of risk factors on medical history, echocardiography, and noninvasive angiography. The efficacy of this algorithm in screening for subjects deemed to be inoperable after catheterization was evaluated retrospectively in 151 children. For this analysis, results of conventional angiography (assumed to be equivalent to noninvasive angiography) were used.

Results: According to the algorithm, 95 (63%) of 151 subjects had no risk factors (''low risk'') whereas 56 (37%) of 151 had 1 risk factor or more (''high risk''). Nine (6%) of 151 subjects were found to be inoperable after catheterization and all 9 were correctly classified as high risk by the algorithm. In the 135 of 151 subjects who underwent a Fontan operation, the algorithm predicted an adverse postoperative outcome with a sensitivity of 51% and specificity of 78%. However, this prediction was not improved by including elevated pulmonary artery pressure or ventricular filling pressure as additional risk factors.

Conclusions: The proposed algorithm effectively screened for subjects who were deemed unsuitable for a Fontan procedure. In addition, omitting preoperative invasive hemodynamic assessment did not impair prediction of adverse postoperative outcomes. Prospective evaluation of such a noninvasive diagnostic strategy before the Fontan operation is warranted.

The Fontan operation is the final surgical stage for most patients with a functionally single ventricle, resulting in a marked change in cardiovascular physiology.[1] Since inception, detailed hemodynamic assessment and angiographic evaluation of the extracardiac thoracic vasculature by cardiac catheterization before surgery has been considered to be mandatory. However, for most other types of congenital heart disease, the use of preoperative catheterization has become infrequent. In part, this is due to the advent of newer noninvasive diagnostic techniques such as echocardiography, magnetic resonance imaging (MRI), and computed tomography (CT) as reliable replacements for invasive x-ray angiography.[2-6] Cardiac catheterization continues to be used routinely for children who are referred for a Fontan operation to perform hemodynamic assessment for the possibility of elevated pulmonary artery pressure (PAP) or increased ventricular filling pressure (VFP), which can only be identified by invasive testing. However, in the modern era of timely staged palliation, it is unusual for patients with functionally single ventricle to be subjected to unrestrictive pulmonary blood flow and have significant pulmonary vascular disease with resultant pulmonary hypertension. Ventricular diastolic dysfunction is a risk factor for patients undergoing the Fontan operation, but it may be difficult to identify, especially with load-dependent parameters such as VFP.[7]

It is rare for a patient to be deemed unsuitable for a Fontan operation as a result of routine invasive hemodynamic testing in the absence of known risk factors.[8] Thus, given the low yield of catheterization, it may be possible to limit hemodynamic testing to only high-risk subjects who have been identified by clinical criteria and noninvasive techniques. However, it is not known whether using such an approach would allow the identification of all subjects who are inoperable. It is also not known whether the omission of routine invasive hemodynamic testing in low-risk patients would impair the ability to predict adverse postoperative outcomes.

As a first step toward answering these questions, in the present study, we developed a diagnostic algorithm for assessment of patients before a Fontan operation. In this

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Abbreviations and Acronyms
CPB = cardiopulmonary bypass
CT = computed tomography
ICU = intensive care unit
MRI = magnetic resonance imaging
NPV = negative predictive value
PAP = pulmonary artery pressure
PPV = positive predictive value
VFP = ventricular filling pressure

algorithm, invasive hemodynamic assessment is limited to only high-risk subjects who have been identified by screening for the presence of clinical risk factors and after assessment by noninvasive imaging. We hypothesized that (1) this new diagnostic algorithm can successfully identify subjects who are inoperable and (2) omission of invasive hemodynamic testing does not impair the ability to predict adverse postoperative outcomes in low-risk patients. We retrospectively tested the efficacy of this algorithm in a series of patients who had undergone routine cardiac catheterization to determine suitability for a Fontan operation.

METHODS

Proposed Diagnostic Algorithm

To consider the possibility of eliminating routine preoperative cardiac catheterization, one must develop an alternative diagnostic strategy that will provide data for assessing suitability for the operation and for predicting an adverse postoperative outcome. We developed such a diagnostic algorithm (Figure 1) for assessment before a Fontan operation on the basis of factors that have been shown to be associated with an increased risk of being inoperable or having adverse outcomes after a Fontan operation.[9-15] In this algorithm, data from history, echocardiography, and noninvasive angiography (using MRI or CT) are used (Table 1). Subjects with 1 or more risk factors are classified as ''high risk'' and undergo conventional cardiac catheterization, whereas subjects without any risk factors are classified as ''low risk'' and undergo a Fontan operation without cardiac catheterization. Angiographic evaluation of the pulmonary vasculature and systemic venous anatomy is important because abnormalities in these areas are not reliably detected by echocardiography. Hence, several angiographic criteria are included in our algorithm. Owing to the retrospective nature of this study, noninvasive angiographic data from MRI or CT were not available. Hence, data from x-ray angiography were used to identify angiographic risk factors. For the purpose of this study, it was assumed that MRI or CT imaging is equivalent to x-ray angiography, as shown previously by numerous investigators.[2-6,16]

Evaluation of Algorithm

To test its diagnostic efficacy, we applied the diagnostic algorithm retrospectively to 151 consecutive children with a functionally single ventricle who underwent cardiac catheterization after the age of 1 year at the Morgan Stanley Children's Hospital of New York Presbyterian between 2000 and 2005, presumably to assess suitability for a Fontan operation. The Institutional Review Board of Columbia University Medical Center gave permission for the analysis of existing clinical data. Data recorded for each subject included the following: (1) presence of risk factors (Table 1) on history, echocardiography, and x-ray angiography performed at the time of referral for preoperative evaluation; (2) suitability for the Fontan operation as determined by the subject's cardiologist, who had access to all available data, including results of cardiac catheterization; (3) PAP and VFP at cardiac catheterization; (4) interventional procedures performed at catheterization; and (5) for subjects who subsequently underwent a Fontan operation, details of the operation (type of Fontan, creation of a fenestration, duration of cardiopulmonary bypass), postoperative course (duration of intensive care unit [ICU] stay, and duration of pleural drainage), and data on survival to 1 year after the operation. An adverse postoperative outcome was defined as the occurrence of one or more of the following events: (1) death within 1 year of the operation; (2) prolonged pleural drainage (>15 days); (3) prolonged ICU stay (>10 days); and (4) cardiac catheterization within a 3-month period after the operation.

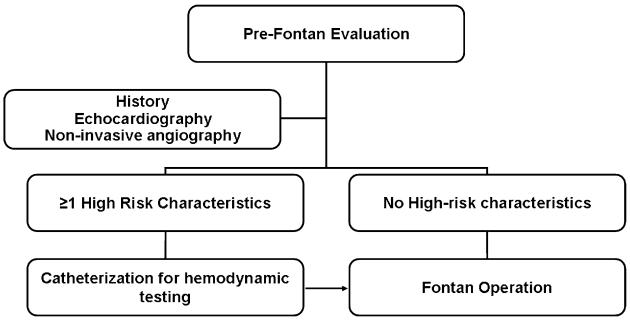

FIGURE 1. Proposed diagnostic algorithm: Characteristics on history, echocardiography, or noninvasive angiography are used to identify high-risk subjects. Low-risk subjects undergo the Fontan operation without preceding catheterization, whereas high-risk subjects undergo catheterization before the operation for hemodynamic testing.

Statistical Analyses

Statistical analyses were performed with commercially available statistical software (STATA SE 9.0; Stata Corp, College Station, Tex). To evaluate the efficacy of the proposed diagnostic algorithm in identifying subjects who are inoperable, we calculated sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV), likelihood ratios, and post-test probability as previously described.[17-19] We also generated 95% confidence intervals (CI) for sensitivity, specificity, PPV, and NPV. To evaluate the proposed diagnostic algorithm's efficacy in predicting adverse postoperative outcomes, we similarly calculated sensitivity, specificity, PPV, NPV, likelihood ratios, and post-test probabilities. To test the hypothesis that the omission of invasive hemodynamic testing does not impair the ability to predict adverse postoperative outcomes, we repeated the above analyses after including the presence of elevated PAP (>15 mm Hg) and/or VFP (>10 mm Hg) as additional risk factors in the proposed algorithm. Furthermore, to identify predictors of an adverse postoperative outcome as previously defined, we used multivariable logistic regression. Variables included for analysis were age at operation, PAP, VFP, ventricular dysfunction, atrioventricular valve regurgitation, creation of a fenestration, presence of heterotaxy or a defined genetic syndrome, cardiopulmonary bypass (CPB) time, and the type of Fontan operation (lateral tunnel or extracardiac conduit). In addition, further analyses were performed to identify predictors of the following individual outcomes: duration of ICU stay, duration of pleural drainage, and death within 1 year after operation. Data for duration of ICU stay and duration of pleural drainage were skewed and underwent logarithmic transformation before analysis to satisfy normalcy requirements. Multivariable linear regression was used to identify the predictors of log-transformed duration of ICU stay and log-transformed duration of pleural drainage, and multivariable logistic regression was used to identify predictors of death within 1 year after operation. For all multivariable modeling using either logistic or linear regression, a ''backward elimination'' method was used for selection of variables and $P < .05$ was required for inclusion of a variable in the final model.

TABLE 1. Characteristics of high-risk subjects

History
Chronic respiratory or lung disorder (chest x-ray abnormality lasting > 3 months/need for supplemental oxygen > 3 months)
Heterotaxy
Genetic syndrome
Echocardiography
≥ Moderate atrioventricular valve regurgitation (qualitative grading)
≥ Moderate ventricular dysfunction (qualitative grading)
≥ Moderate aortic insufficiency (qualitative grading)
Pulmonary vein stenosis (mean Doppler gradient > 3 mm Hg)
Angiography
≥ Moderate pulmonary artery branch stenosis or discontinuity (qualitative grading)
Pulmonary vein stenosis (qualitative assessment)
Systemic venous anomaly (excluding left superior vena cava)

RESULTS

A total of 151 consecutive children (63% male) who underwent cardiac catheterization for assessment before a Fontan operation between 2000 and 2005 at Children's Hospital of New York Presbyterian were included in this study. Subject diagnoses are shown in Table 2. Hypoplastic left heart syndrome was the most common diagnosis, in approximately one third of the subjects. A majority (147/151, 97%) of subjects had undergone a superior cavopulmonary anastomosis with either a bidirectional Glenn (n = 117) or hemi-Fontan (n = 30) operation. According to the proposed algorithm, 95 (63%) subjects had no risk factors (low-risk group) and 56 (37%) had 1 or more risk factors (high-risk group).

Identification of Inoperable Subjects by the Proposed Algorithm (Figure 2)

A total of 9 subjects were deemed inoperable for the Fontan operation by their cardiologist after undergoing cardiac catheterization. The proposed algorithm performed well in screening for inoperability, and all of the 9 inoperable subjects were correctly classified in the high-risk group. However, 47 eligible subjects were also classified as high risk. In identifying inoperable subjects, the algorithm's sensitivity (100%, 95% CI 66–100) and NPV (100%, 95% CI 97–100) were high whereas the specificity (67%, 95% CI 59–75) and PPV (16%, 95% CI 8–28) were low. For a subject classified as high risk by the algorithm, the likelihood ratio was 3.03 and the post-test probability of being inoperable was 19% (increased from a pre-test probability of 6%). On the other hand, for a subject classified as low risk by the algorithm, the likelihood ratio was 0.015 (assuming a sensitivity of 99% instead of the actual 100% to allow calculation) and post-test probability of being inoperable was 0.1% (decreased from a pre-test probability of 6%). The characteristics of the 9 subjects who were inoperable are described in Table 3. All inoperable subjects had risk factors on history, echocardiography, or angiography and 6 of the 9 had abnormal PAP and/or VFP. Several subjects had multiple risk factors. No inoperable subject had an elevated PAP and/or VFP without an associated risk factor on history, echocardiography, or angiography.

TABLE 2. Subject characteristics

Diagnosis	N
Hypoplastic left heart syndrome	53 (35%)
Unbalanced atrioventricular canal defect	26 (17%)
Double-inlet left ventricle	18 (12%)
Tricuspid atresia	17 (11%)
Pulmonary atresia with intact ventricular septum	12 (8%)
Others	25 (17%)

Subjects Who Underwent a Fontan Operation

At the time of data analysis, 135 of the 142 operable subjects had undergone the Fontan operation at a median age of 3.2 years (interquartile range 2.5–3.9 years).

Preoperative characteristics. Of the 135 subjects who underwent a Fontan operation, 95 were in the low-risk group and 40 were in the high-risk group. PAP and VFP were similar in the low-risk (11 ± 3 and 9 ± 2 mm Hg) and high-risk groups (12 ± 3 and 9 ± 3 mm Hg; $P > .5$ for both). In the low-risk group, 24 (27%) children had an elevated PAP (>15 mm Hg, n = 7) and/or VFP (>10 mm Hg, n = 23), whereas in the high-risk group, 22 (55%; $P = .0009$ using a 2-sided Student t test) had an elevated PAP (n = 5) and/or VFP (n = 22). However, no patient undergoing a Fontan operation had a PAP greater than 23 mm Hg or a VFP greater than 14 mm Hg.

Operative details and early postoperative outcomes. The Fontan operation was performed with a lateral tunnel in 99 (74%) and an extracardiac conduit in 35 (26%) subjects. A fenestration was performed in 81 (60%) subjects. Within the first month after the operation, there were 2 deaths and 1 patient required heart transplantation. Median duration of stay in the ICU was 6 days (interquartile range 4–8 days). Median duration of pleural drainage was 9 days (interquartile range 7–12 days). An adverse postoperative outcome as previously defined occurred in 26% (95% CI 19–34%) of subjects.

Prediction of postoperative outcomes using the algorithm with and without additional hemodynamic data. As seen in Table 4, the proposed algorithm had modest success in predicting adverse postoperative outcomes. Compared with the entire study population, presence in the high-risk group increased the probability of an adverse outcome whereas presence in the low-risk group decreased the probability of an adverse outcome. The occurrence of an adverse postoperative outcome was significantly associated with a high-risk status ($\chi^2 = 10.8$; $P = .001$; odds ratio 3.75, 95% CI 1.53–9.16). However, prediction of an adverse postoperative outcome was not improved by including an elevated PAP and/or VFP

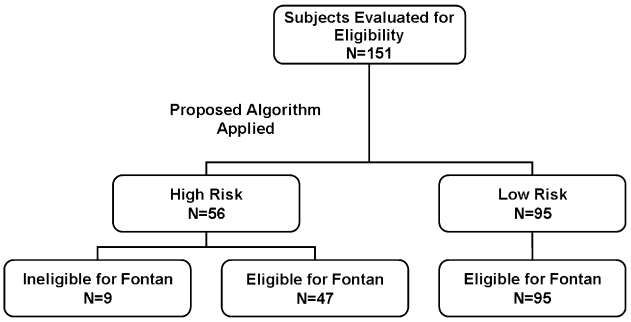

FIGURE 2. Predicting operative eligibility: Results of retrospective testing of the algorithm on 151 children presenting for evaluation before a Fontan operation. All 9 ineligible (inoperable) subjects were correctly classified as high risk. All low-risk subjects were deemed eligible.

as an additional risk factor, and with this modified algorithm, an adverse outcome was not significantly associated with high-risk or low-risk status ($\chi^2 = 3.3$; $P = .07$).

Predictors of an adverse outcome. On multivariable modeling, an adverse postoperative outcome, as previously defined, was independently associated with a defined genetic syndrome ($P = .02$, odds ratio 17.7, 95% CI 1.5–208), heterotaxy syndrome ($P = .02$, odds ratio 4.7, 95% CI 1.3–17.1), a longer duration of CPB ($P = .008$, odds ratio 1.01, 95% CI 1.003–1.02 for 1-minute increase in duration of CPB), and with the absence of a fenestration ($P = .01$, odds ratio 0.29, 95% CI 0.11–0.74). On further analysis to identify predictors of individual adverse outcomes, a longer ICU stay (after logarithmic transformation) was associated with a longer duration of CPB ($P < .0001$, coefficient = 0.006, 95% CI 0.003–0.009 for 1-minute increase in duration of CPB), heterotaxy syndrome ($P = .01$, coefficient = 0.5, 95% CI 0.11–0.88), and a defined genetic syndrome ($P = .002$, coefficient 0.74, 95% CI 0.08–1.4). Longer duration of pleural drainage (after logarithmic transformation) was independently associated with a longer duration of CPB ($P = .002$, coefficient = 0.003, 95% CI 0.001–0.006 for 1-minute increase in duration of CPB), heterotaxy syndrome ($P = .04$, coefficient = 0.36, 95% CI 0.01–0.7), and the absence of a fenestration ($P = .008$, coefficient = –0.29, 95% CI –0.49 to –0.08). Notably, there were no significant associations between adverse postoperative outcomes and preoperative hemodynamic measurements (PAP or VFP).

Survival to 1 year. Five subjects died within 1 year after the operation, all of whom were classified in the high-risk group according to the algorithm. The characteristics of these subjects and the clinical events leading to death in each case are shown in Table 5. Four of the 5 had an unbalanced atrioventricular canal defect. All 5 subjects had prominent and often multiple noninvasive risk factors. Three of the 5 subjects had heterotaxy, and 3 had severe atrioventricular valve regurgitation. Only 1 of the 5 had abnormal preoperative hemodynamics. The preoperative PAP was within normal limits in all 5 children and the preoperative VFP was elevated in 1 of the 5 children. On multivariable modeling, the presence of greater than moderate atrioventricular valve regurgitation ($P = .003$, odds ratio 46.3, 95% CI 3.6–593) and heterotaxy syndrome ($P = .049$, odds ratio 17.3, 95% CI 1.01–172) were independently associated with risk of mortality within 1 year after the Fontan operation. PAP and VFP were not associated with risk of mortality within 1 year.

Interventional Procedures Performed

Eighty-five transcatheter interventions were performed in 73 (48%) of 135 subjects who subsequently underwent the

TABLE 3. Subjects deemed inoperable for a Fontan operation

No	Diagnosis	PAP (mm Hg)	VFP (mm Hg)	Echo risk factors	Angiographic risk factors	Clinical risk factors
1	Unbalanced AVCD	20	12	Severe ventricular dysfunction, prosthetic AV valve stenosis	—	—
2	HLHS	19	18	—	—	Chronic lung disease, Noonan syndrome
3	Unbalanced AVCD	25	18	Severe ventricular dysfunction, moderate AVVR	Left pulmonary vein stenosis	Chronic lung disease, tracheostomy
4	Mitral atresia, DORV	27	21	Severe AVVR, decreased ventricular function	Hypoplastic RPA	Chronic lung disease, tracheostomy
5	Mitral atresia, DORV, scimitar syndrome, TAPVC	24	13	—	Scimitar vein	Right lung hypoplasia
6	HLHS	10	11	—	Discontinuous LPA	—
7	HLHS	14	8	—	LPA occluded by thrombus	—
8	HLHS	9	8	—	Occluded SVC	Chronic lung disease
9	HLHS	10	9	—	Severe stenosis of head and neck vessels	—

AV, Atrioventricular; *AVCD,* atrioventricular canal defect; *AVVR,* atrioventricular valve regurgitation; *DORV,* double-outlet right ventricle; *Echo,* echocardiographic; *HLHS,* hypoplastic left heart syndrome; *LPA,* left pulmonary artery; *PAP,* pulmonary artery pressure; *RPA,* right pulmonary artery; *SVC,* superior vena cava; *TAPVC,* totally anomalous pulmonary venous connection; *VFP,* ventricular filling pressure.

TABLE 4. Prediction of adverse postoperative outcomes using the algorithm with and without hemodynamic data

	Proposed algorithm	Algorithm including PAP, VFP
Sensitivity (95% CI)	51% (34%–69%)	62% (45%–79%)
Specificity (95% CI)	78% (68%–86%)	55% (45%–65%)
PPV (95% CI)	45% (29%–62%)	29% (19%–40%)
NPV (95% CI)	82% (73%–89%)	81% (70%–89%)
High-risk status		
Likelihood ratio	2.32	1.37
Pre-test probability	26%	26%
Post-test probability	45%	25.4%
Low-risk status		
Likelihood ratio	0.63	0.69
Pre-test probability	26%	26%
Post-test probability	18%	20%

CI, Confidence interval; *NPV,* negative predictive value; *PAP,* pulmonary artery pressure; *PPV,* positive predictive value; *VFP,* ventricular filling pressure.

Fontan operation. A majority of these interventions included embolization of aortopulmonary collateral arteries (n = 64, 75%). Other procedures performed included embolization of venovenous collaterals (n = 15), stenting of pulmonary artery stenosis (n = 3), angioplasty for recurrent coarctation of the aorta (n = 2), and embolization of a right ventricular sinusoid (n = 1). Interventions were performed in 23 (58%) of 40 subjects in the high-risk group and in 50 (56%) of 90 subjects in the low-risk group. No relationship was seen between the performance of an intervention and an adverse postoperative outcome.

DISCUSSION

In this study, we proposed and evaluated a diagnostic algorithm that does not use routine cardiac catheterization for low-risk subjects before the Fontan operation. In a retrospective evaluation, the proposed algorithm was successful as a screening tool to identify subjects who are inoperable. In addition, adverse postoperative outcomes, including death within 1 year after the operation, were not associated with preoperative hemodynamic parameters; hence if such an algorithm were used to screen patients, the omission of routine invasive hemodynamic measurements would not impair the ability to predict adverse early postoperative outcomes.

Identifying Inoperable Subjects

In identifying subjects who are inoperable, the proposed algorithm is highly sensitive (100%) but has a low specificity (67%). To maintain a high level of sensitivity, we chose to use a low threshold for classifying a subject as high risk ($\geq$1 risk factor), and this likely resulted in a low specificity. However, this low specificity is acceptable because this algorithm is being proposed only as a screening tool to identify high-risk patients who would then undergo invasive testing. Notably, the inoperable subjects had relatively prominent and often multiple risk factors and all were classified as high risk according to the proposed algorithm. Furthermore, inoperable status was rare (6%), likely resulting from a consistent policy of staged palliation and the rarity of pulmonary vascular disease. If the proposed algorithm were used to screen subjects who require cardiac catheterization, all the inoperable subjects would be correctly classified as high risk and would undergo cardiac catheterization. It should be noted that these results are based on the assumption that MRI or CT imaging is equivalent to conventional x-ray angiography.

Predicting Adverse Postoperative Outcomes

In addition to assessing eligibility, prediction of adverse postoperative outcomes is another goal of preoperative testing. In this study, we found that the proposed diagnostic algorithm allowed modest prediction of adverse postoperative

TABLE 5. Mortality within 1 year

				Preoperative hemodynamics (mm Hg)			
No.	Age at Fontan (y)	Diagnosis	Nonhemodynamic risk factors	PAP	VFP	Timing of death (days after Fontan)	Clinical events related to mortality
1	1.7	Unbalanced AVCD	Trisomy 21, severe AVVR	12	10	89	Prolonged pleural effusion, thrombus in Fontan pathway, sepsis
2	3.8	Unbalanced AVCD	Heterotaxy	12	9	90	Prolonged pleural and pericardial effusion, thrombus in Fontan pathway
3	7	Unbalanced AVCD	Heterotaxy	13	9	25	Low cardiac output, renal failure, JET
4	2	Unbalanced AVCD	Heterotaxy, severe AVVR	10	7	22	Low cardiac output, respiratory failure.
5	2	HLHS	Severe AVVR, occluded LPA	9	5	80	Prolonged pleural effusion

AVCD, Atrioventricular canal defect; *AVVR,* atrioventricular valve regurgitation; *HLHS,* hypoplastic left heart syndrome; *JET,* junctional ectopic tachycardia; *LPA,* left pulmonary artery; *PAP,* pulmonary artery pressure; *VFP,* ventricular filling pressure.

events but, more importantly, the addition of invasive hemodynamic parameters to this algorithm did not improve this prediction. Hence, if the proposed algorithm was used to screen subjects who require cardiac catheterization, the omission of routine invasive hemodynamic testing in the low-risk subjects would not impair the ability to predict adverse postoperative outcomes. Previous studies have demonstrated that invasive hemodynamic parameters can be used to predict postoperative outcomes.[9-12] However, in our study, we did not find a significant relationship between preoperative invasive hemodynamic parameters and postoperative outcomes. In fact, we found that adverse early postoperative outcomes were associated with nonhemodynamic factors such as the presence of heterotaxy or genetic syndromes and the duration of CPB. Hemodynamic parameters also were not predictive of mortality within 1 year. The only predictors of mortality were the presence of greater than moderate atrioventricular valve regurgitation and heterotaxy syndrome. The lack of association between preoperative hemodynamics and adverse postoperative outcomes may be related to a consistent policy of timely staged palliation in the current surgical era and the resultant rarity of significantly elevated PAP or VFP.

Interventional Procedures

In addition to assessment of eligibility and prediction of postoperative outcomes, cardiac catheterization also provides an opportunity for interventional procedures before the Fontan operation. In a significant proportion of our study population (48%), an intervention was performed during cardiac catheterization, which most frequently (75%) consisted of embolization of aortopulmonary collateral vessels. The clinical utility of routine embolization of small aortopulmonary collateral vessels is not known. Data both supporting and criticizing this practice have been presented by various investigators; hence, this practice is of unproven benefit.[20-25] With the proposed algorithm, subjects with significant aortopulmonary collaterals would be identifiable using noninvasive angiography and subsequent coil embolization could be performed if deemed necessary. Further investigation is needed to determine whether embolization of small aortopulmonary collateral vessels improves clinical outcomes.

Study Limitations

This study had several important limitations. First, the study is limited by its retrospective design. Hence, although the algorithm would omit cardiac catheterization for low-risk subjects, all the patients in this study underwent cardiac catheterization. Second, although data obtained with noninvasive angiographic techniques such as MRI or CT are a component of the proposed diagnostic algorithm, the accuracy of MRI or CT was not directly compared with x-ray angiography, although this has been demonstrated previously by several investigators in children with complex congenital heart disease, including those with a functionally single ventricle.[2-6,16] A recently published randomized trial found that cardiac MRI is a safe, effective, and less costly alternative to routine catheterization in the evaluation of selected patients before a bidirectional Glenn operation.[16] Third, follow-up beyond 1 year after the operation was not available for this study and hence long-term outcomes were not evaluated. Finally, the effect of interventional procedures such as the coiling of aortopulmonary collateral arteries in low-risk patients, while controversial, was not assessed in this study. Although the long-term benefit of these interventions in low-risk subjects has not been established, it is not known whether eliminating cardiac catheterization and interventional procedures would affect outcome.

CONCLUSION

In summary, we have shown that a noninvasive diagnostic algorithm such as the one proposed here could potentially be used as a screening tool to identify subjects who are inoperable for a Fontan procedure. Second, our results indicate that preoperative hemodynamic parameters are not associated with early postoperative outcomes. Adverse postoperative outcomes were associated with risk factors identified on medical history, such as heterotaxy and genetic syndromes. Hence, the omission of routine cardiac catheterization in low-risk subjects would not impair identification of inoperable subjects or those at risk for adverse early postoperative outcomes. The use of the proposed algorithm could help avoid the risk of cardiac catheterization and reduce the cost of evaluation in low-risk subjects while allowing the opportunity for invasive hemodynamic testing and/or interventional procedures in high-risk subjects. Owing to the retrospective nature of this study and because MRI or CT imaging was not performed in our subjects, the results should be considered preliminary until further prospective validation of such a strategy is performed. It should be noted that high-quality MRI or CT imaging is necessary if such a strategy is used. Finally, further investigation is warranted to identify other predictors of adverse outcomes after the Fontan operation and to evaluate whether routine embolization of small aortopulmonary collateral arteries improves outcomes.

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