Diagnostic Assessment Before Fontan Operation in Patients With Bidirectional Cavopulmonary Anastomosis
Are Noninvasive Methods Sufficient?

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OBJECTIVES
This study was designed to determine if a subset of patients who have undergone bidirectional cavopulmonary anastomosis could be identified in which catheterization was of little benefit before completion of the Fontan procedure.

BACKGROUND
Diagnostic evaluation before Fontan procedure has typically included cardiac catheterization. However, the overall management strategy for patients with functional single ventricle has evolved to include staging bidirectional cavopulmonary anastomosis in most, and it has become uncommon to exclude patients from Fontan based on catheterization data.

METHODS
Patients who underwent bidirectional cavopulmonary anastomosis and had complete echocardiograms and catheterizations within three months of each other between January 1992 and October 1997 were evaluated with a series of clinical and echocardiographic characteristics to identify a subset in whom catheterization was predicted to be of little added value ("no-cath" group). The predictive value and sensitivity of these criteria in excluding patients who required additional intervention, were excluded from Fontan, or died within 30 days of Fontan was determined.

RESULTS
A total of 99 patients who underwent bidirectional cavopulmonary anastomosis at 6.7 months (range 2.9 months to 14 years) were studied; 46 met criteria for the "no-cath" group. Noninvasive criteria stratified all patients who died (n/H11005 5) or did not proceed to Fontan (n/H11005 1) and 9 of 11 who required additional interventions to the "cath" group. Thus, the negative predictive value of these criteria was 93%.

CONCLUSIONS
Our data suggest that catheterization before Fontan could be avoided in a large percentage of patients without adversely affecting outcome; prospective evaluation of this strategy is warranted. (J Am Coll Cardiol 2004;44:184–7) © 2004 by the American College of Cardiology Foundation

Noninvasive techniques are commonly used as the sole means of evaluation before surgical repair of congenital heart disease. Past studies have demonstrated the accuracy of echocardiography alone for preoperative diagnosis in many lesions including atrial septal defects, complete atrioventricular (AV) canal, and tetralogy of Fallot (1–5).

Echocardiography is also commonly used as the sole diagnostic modality before the first palliative procedure in patients with single-ventricle lesions (6,7). Diagnostic evaluation before Fontan procedure has typically included noninvasive assessment with echocardiography as well as invasive hemodynamic and angiographic evaluation by cardiac catheterization (8,9). The goals of this diagnostic assessment are: 1) to identify patients in whom the Fontan operation should not be performed because of excessive risk; and 2) to identify patients in whom additional interventions (either by catheter before surgery or in the operating room concomitant with the Fontan procedure) are required.

The overall management strategy for patients with functional single-ventricle heart disease has evolved since the enunciation of the so-called Fontan laws (10). At our institution, most patients proceed to Fontan surgery after antecedent bidirectional cavopulmonary anastomosis. It has become uncommon to exclude patients from Fontan based on hemodynamic parameters obtained at catheterization (11). We hypothesized that, after a previous bidirectional cavopulmonary anastomosis, a subset of patients could be identified in whom catheterization was of little added benefit before total cavopulmonary anastomosis. Based on clinical experience we delineated a series of criteria to discriminate between patients who would benefit from catheterization before Fontan (cath) and those who would not (no-cath). We then tested the value of these criteria in predicting either poor outcome or the need for additional intervention.

METHODS
The surgical database at our institution was queried to identify all patients who underwent bidirectional superior cavopulmonary anastomosis between January 1992 and October 1997.
For inclusion in the study we required that patients have a complete echocardiogram performed within three months of cardiac catheterization in anticipation of completion of the Fontan procedure. According to our laboratory standards, complete echocardiograms in this setting include assessment of systemic and pulmonary venous return, flow at the atrial septum, AV and semilunar valve function, ventricular performance, pulmonary artery architecture, and aortic arch anatomy and flow velocity. Medical records, operative notes, and cardiac catheterization data were reviewed. Independent observers blinded to outcomes reviewed echocardiograms and angiograms.

Patients were divided into those in whom catheterization was predicted to be unnecessary (no-cath) and those who were predicted to require catheterization (cath) before proceeding to the Fontan operation. Patients were included in the cath group if they met any one of the criteria, including two clinical features (pulse oximetry <76%, hemoglobin concentration >18 g/dl) and six echocardiographic features (Table 1). The clinical course of all patients was reviewed to determine which patients were subsequently excluded from proceeding to Fontan, required additional interventions (at catheterization or surgery), or died within 30 days of Fontan. Predictive value and sensitivity of the discriminating criteria were determined. Summary data are expressed as mean (range).

RESULTS

Patient population. A total of 305 patients underwent bidirectional cavopulmonary anastomosis between January 1992 and October 1997. Of the total, 101 patients had an echocardiogram and cardiac catheterizations within three months of each other after bidirectional cavopulmonary anastomosis with data available for review. In two cases, echocardiography studies were inadequate as defined (see the Methods section). Thus, the study population consisted of 99 patients. Of these, 94 patients had undergone hemi-Fontan and 5 had undergone bidirectional Glenn procedures. The median age at hemi-Fontan or bidirectional Glenn was 6.7 months (range 2.9 months to 14 years). The median age at echocardiogram was 19.1 months (4 months to 14 years), and the median age at cardiac catheterization was 19.1 months (4 months to 14 years), with the median time between echocardiogram and cardiac catheterization being one day (range 0 to 82 days). The median age at Fontan procedure was 21 months (range 11 months to 14 years). A total of 46 patients met criteria for the no-cath group, and 53 were stratified to cath. Among the latter group, more than one criterion was present in 12 patients (Table 2).

Because a large number of patients were excluded from this study, excluded patients were compared with those in the study population. The median age at hemi-Fontan or bidirectional Glenn among the 206 patients excluded from this study was 6.9 months (p = NS compared with study group). Mortality among the excluded patients was 15% (n = 31), higher than the study population. However, this apparent difference is explained by the fact that most of these deaths occurred early after hemi-Fontan, before the age where patients would have been eligible for entrance into this study. Of the 31 deaths, 23 occurred before 19 months of age (the average age at catheterization in this study). This leaves eight patients who died more than 19 months after hemi-Fontan, a mortality rate of 4%, not significantly different from that in the study group.

Outcomes. In 17 of the 99 patients, the subsequent course was complicated. Five died (two before Fontan and three after Fontan), and one did not proceed to Fontan. Other interventions in addition to Fontan surgery were performed in 11 patients; these are outlined in Table 3.

Table 1. Characteristics of Low-Risk Patients

<table>
<thead>
<tr>
<th>Clinical data</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Room air pulse oximetry ≥76%</td>
<td>31</td>
</tr>
<tr>
<td>Hemoglobin ≤18 g/dl</td>
<td>31</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Echocardiographic data</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left pulmonary artery visualized without stenosis</td>
<td>32</td>
</tr>
<tr>
<td>No significant atrioventricular valve insufficiency (≥1+)</td>
<td>10</td>
</tr>
<tr>
<td>No significant ventricular dysfunction (qualitative)</td>
<td>6</td>
</tr>
<tr>
<td>No aortic coarctation</td>
<td>1</td>
</tr>
<tr>
<td>An unrestricted atrial communication</td>
<td>1</td>
</tr>
<tr>
<td>No evidence of a decompressing vessel</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 2. Characteristics Predicted to Discriminate Need for Catheterization

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Number of Patients Meeting Criteria for Cath (n = 53)*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical data</td>
<td></td>
</tr>
<tr>
<td>Pulse oximetry &lt;76%</td>
<td>8</td>
</tr>
<tr>
<td>Hemoglobin &gt;18 g/dl</td>
<td>9</td>
</tr>
<tr>
<td>Echocardiographic data</td>
<td></td>
</tr>
<tr>
<td>Left pulmonary artery not seen or stenotic (19 not visualized)</td>
<td>32</td>
</tr>
<tr>
<td>(13 stenotic)</td>
<td></td>
</tr>
<tr>
<td>Significant atrioventricular valve regurgitation (&gt;1+)</td>
<td>10</td>
</tr>
<tr>
<td>Significantly diminished ventricular contractility</td>
<td>6</td>
</tr>
<tr>
<td>Aortic coarctation</td>
<td>1</td>
</tr>
<tr>
<td>Restrictive atrial septal defect</td>
<td>1</td>
</tr>
<tr>
<td>Evidence of a decompressing vessel</td>
<td>1</td>
</tr>
</tbody>
</table>

*Nine patients met two criteria and three patients met three criteria.

Abbreviations and Acronyms

<table>
<thead>
<tr>
<th>AV = atrioventricular</th>
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<td>PPV = positive predictive value</td>
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Additional Surgical or Transcatheter Interventions in Patients Undergoing Bidirectional Superior Cavopulmonary Anastomosis

<table>
<thead>
<tr>
<th>Intervention</th>
<th>&quot;No Cath&quot; Group</th>
<th>&quot;Cath&quot; Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coil embolization of decompressing vessel</td>
<td>2</td>
<td>2*</td>
</tr>
<tr>
<td>Atrioventricular valvuloplasty</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Balloon dilation of left pulmonary artery</td>
<td>0</td>
<td>3*</td>
</tr>
<tr>
<td>Balloon dilation of coarctation</td>
<td>0</td>
<td>1*</td>
</tr>
<tr>
<td>Atrial septectomy</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

*One patient had three interventions.

surgical valvuloplasty of the AV valve at the time of Fontan. This patient had mild AV valve regurgitation by echocardiogram and moderate by angiography. Of the 53 patients meeting criteria for cath, 5 died and 8 underwent additional interventions. The ability of these criteria to appropriately stratify patients to cath (negative predictive value [NPV]) was 93% (95% confidence interval 86% to 100%) with 81% sensitivity. Specificity, however, was only 52%, and the positive predictive value (PPV) was only 25% with many patients stratified to catheterization in whom no pertinent findings emerged.

DISCUSSION

Many patients were excluded from the Fontan procedure in the early decades of its application based on selection criteria that could be accurately obtained only by cardiac catheterization (9,10). Since then, patient population, surgical approach, and diagnostic tools have dramatically changed. In our practice the vast majority of patients considered for Fontan have undergone prior bidirectional superior cavopulmonary anastomosis (12). The goal of diagnostic evaluation before completion of the Fontan procedure is to identify those few patients in whom the Fontan operation should not be performed, as well as those who require additional intervention before or at the time of Fontan. Despite these changes, current practice continues to include cardiac catheterization before Fontan. Although catheterization can be performed safely, it is not without risk. Recent studies report complications in 5% to 10% and death in 0.1% of diagnostic catheterizations (13,14). Catheterization is costly and is an additional procedure for children who are already subjected to many such procedures. Thus, we sought to determine whether noninvasive criteria could be used to define a subset of patients who could forgo preoperative catheterization without an adverse effect on outcome. Eight criteria were defined, retrospectively applied, and evaluated for predictive value. Forty-six percent of patients met criteria to avoid catheterization. Overall, 17% of patients died, required additional intervention, or did not proceed to Fontan. The selection criteria appropriately stratified all 5 patients who died before or after Fontan procedure and 8 of the 11 patients who underwent additional intervention.

Three patients who were stratified to the no-cath group received additional interventions: small decompressing veins were embolized in two patients, and one patient underwent valvuloplasty at the time of Fontan. It is not possible to definitively assess the importance of these interventions. However, it is unlikely that failure to embolize the decompressing veins would have resulted in excess cyanosis after Fontan; in both instances the vessels originated from the superior cavopulmonary circuit, and neither patient had an unusually low arterial saturation before Fontan.

Overall, the ability of our criteria to discriminate patients who could forgo catheterization (NPV) was acceptable (93%). However, the positive predictive value of this strategy was rather low; the majority of patients stratified to undergo catheterization required no additional intervention and had unremarkable courses at Fontan. The limited ability of echocardiography to adequately assess branch pulmonary artery architecture contributed to this poor predictive value (19% of cases). Application of magnetic resonance imaging to these cases would substantially improve PPV of these criteria.

Other important limitations of this work should be noted. This study is retrospective and subject to all shortcomings inherent in retrospective analyses. We chose as outcomes the need for any additional intervention and deaths. We did not attempt to determine to what extent catheterization predicted these outcomes. This approach was taken because we reasoned that attempts at such determinations would be too subject to bias in a retrospective analysis. Clearly a prospective evaluation of this management strategy must follow before its generalized application. Finally, it should be noted that at our institution coil embolization of small systemic to pulmonary arteries (commonly found in this patient group) is not routinely performed before Fontan. Some have suggested that the presence of collaterals correlates with worse outcome after Fontan and, therefore, they routinely embolize these vessels (15,16). These findings have not been reproduced in other studies (17). We reserve this intervention for instances with abundant collateral flow and hemodynamic evidence of their significance: high pulmonary artery pressure or elevated ventricular filling pressure. It seems unlikely that noninvasive criteria would correctly identify patients with more extensive collaterals. In spite of these limitations, our data suggest that catheterization before Fontan could be avoided in a large percentage of patients. We suggest that prospective evaluation of this strategy is warranted.

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REFERENCES

1. Freed MD, Nadas AS, Norwood WI, Castaneda AR. Is routine preoperative cardiac catheterization necessary before repair of secun-

Table 3. Additional Surgical or Transcatheter Interventions in Patients Undergoing Bidirectional Superior Cavopulmonary Anastomosis

Intervention                                      | "No Cath" Group | "Cath" Group |
--------------------------------------------------|-----------------|--------------|
| Coil embolization of decompressing vessel        | 2               | 2*           |
| Atrioventricular valvuloplasty                   | 1               | 3            |
| Balloon dilation of left pulmonary artery        | 0               | 3*           |
| Balloon dilation of coarctation                  | 0               | 1*           |
| Atrial septectomy                                | 0               | 1            |

*One patient had three interventions.