Preoperative evaluation of candidates for total cavopulmonary connection: The role of echocardiography and cardiac catheterization

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Congenital heart disease; Paediatrics; Single ventricle; Cyanosis

Summary
Aim. — To evaluate the information provided by echocardiography and cardiac catheterization in patients with partial cavopulmonary connection (PCPC) with or without additional pulmonary blood flow (PBF) in whom total cavopulmonary connection (TCPC) was planned.

Methods. — We retrospectively evaluated the results provided by echocardiography and cardiac catheterization in 110 consecutive patients with PCPC (35 with isolated PCPC, 38 with associated antegrade PBF, 37 with associated retrograde PBF) in whom TCPC was planned.

Results. — Eight patients had superior vena cava syndrome; all others suffered from cyanosis and fatigue. Pulmonary artery size could be determined in 54% of patients without additional PBF, in 47% of those with associated retrograde PBF, and in 68% of those with associated PBF (p = 0.20). Concomitant clinical signs of increased central venous pressure and echocardiographic signs of ventricular dysfunction and/or hypoplastic pulmonary arteries identified four patients in whom TCPC was not low-risk. PAP exceeding 16 mmHg was found in 9% of patients without and 16%
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Background

PCPC usually precedes conversion to TCPC in patients with
single ventricle physiology. PCPC can be the first palliation
in patients with low PAP, but it is often preceded by BT shunt
in patients with duct-dependent physiology or by pulmonary
banding in those with pulmonary overflow. PCPC can be the
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reach the age of 2 to 3 years with moderate cyanosis at
rest. This is the age at which TCPC is performed in most
centres, generally by using an extracardiac conduit [1,2].
When TCPC is planned at an older age [3], an additional
native or surgically created source of PBF can be left in
place at the time of PCPC. This strategy is often adopted
by some cardiac surgeons of the French school [4]. TCPC is
generally preceded by cardiac catheterization, intended to
measure mean PAP, to assess pulmonary artery size and to
treat possible associated anomalies.

For low-risk TCPC, 10 inclusion criteria were initially
proposed [5], but some of these have been considered
overly restrictive [6,7]. At present, the criteria for low-risk
TCPC are mean PAP not exceeding 14 to 16 mmHg, normal
pulmonary artery size [3,8] and absence of ventricular dys-
function and/or severe incompetence of AV valve(s) [3,8,9].
The most useful index to measure pulmonary artery size
is the Nakata index [10]. The accepted Nakata index for
patients scheduled for TCPC is at least 200 mm2/m2 [3,8,9].

The aim of this study was to evaluate retrospectively
patients with PCPC with or without additional PBF who
underwent echocardiography and cardiac catheterization
before TCPC over a 10-year period. We had two main objec-
tives: to determine whether clinical information associated
with echocardiographic findings could have been sufficient
to identify low-risk candidates for TCPC and to identify the

Abbreviations

AV atrioventricular
BT Blalock-Taussig
CT computed tomography
PAP pulmonary pressure
PBF pulmonary blood flow
PCPC partial cavopulmonary connection
TCPC total cavopulmonary connection

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proportion of patients in whom cardiac catheterization was necessary either to contraindicate TCPC and/or to treat associated anomalies.

**Methods**

**Study population**

From January 1998 to September 2007, 110 patients with PCPC underwent scheduled cardiac catheterization before TCPC. Consecutive paediatric patients (less than 18 years) with PCPC with or without additional PBF in whom TCPC was planned were included. Patients were excluded for severe ventricular dysfunction associated with severe incompetence of the AV valve(s) needing medical treatment, hepatic disease, interrupted inferior vena cava and chromosomal diseases apart from trisomy 21.

We identified three groups of patients:

- isolated PCPC \( (n = 35) \);
- PCPC with associated retrograde flow \( (n = 37) \);
- PCPC with associated antegrade flow \( (n = 38) \).

**Clinical, echocardiographic and haemodynamic evaluation**

Hospital records and echocardiographic, catheterization and surgical files were reviewed. Complications occurring during catheterization and/or surgery were sought and recorded. All children underwent clinical and echocardiographic assessments and cardiac catheterization at Necker—Enfants- Malades. The study was approved by the institutional review committee. Parents gave consent to catheterization and to inclusion into the study.

The following clinical findings were noted: fatigue, superior vena cava syndrome (defined as facial and neck swelling), hepatomegaly and systemic oxygen saturation at rest. The echocardiographic findings recorded were:

- systolic dysfunction of the single ventricle, defined as ejection fraction of less than 50\% [9];
- increased filling pressure, defined as inferior vena cava depression during inspiration of less than 50\%;
- moderate to severe incompetence of AV valve(s) [8,9];
- visualization and size of pulmonary arteries.

The Simpson technique was used to calculate ejection fraction, although we are aware that this method has limited value for patients with a single ventricle.

All echocardiographic evaluations were performed using a GE Vingmed Vivid 7 ultrasound system connected to a 5-MHz probe (General Electrics, Fairfield, Connecticut; USA). Catheterization was performed, according to the presence of forward flow to the lungs, via the femoral vein or the internal jugular vein. Heparin at a dose of 50 UI/kg or 100 UI/kg was administered when angiography was performed. Pulmonary angiography was performed by injecting 1 mL/kg of contrast medium. The diameters of the right and left pulmonary arteries were measured immediately proximal to their first branches and used to calculate cross-sectional areas. The combined pulmonary artery areas were expressed as the Nakata index as previously described [10].

During the study period, non-invasive imaging of pulmonary arteries was not performed systematically.

TCPC was contraindicated when both PAP was more than 16 mmHg and the Nakata index was less than 200 mm\(^2\)/m\(^2\), in the absence of additional flow; when PAP remained more than 16 mmHg after test occlusion of any additional flow; or when ventricular dysfunction with or without incompetence of AV valve(s) was accompanied by increased atrial or pulmonary wedge pressure despite medical treatment.

**Statistical analyses**

Results are expressed as numerical values and percentages for categorical variables and as mean ± standard deviation (SD) for continuous variables.

Comparisons between the three groups (isolated PCPC, PCPC with associated retrograde PBF and PCPC with associated antegrade PBF) were performed using Anova for continuous variables and the \( \chi^2 \) test or Fisher’s exact test for categorical data. Comparisons of continuous variables between patients with and without additional PBF were performed with Student \( t \)-test, whereas comparisons of categorical variables were performed with the Pearson \( \chi^2 \) test or Fisher’s exact test, as appropriate.

All statistical analyses were performed using the R software package (http://cran.r-project.org/). Statistical significance was considered as a \( p \)-value of less than 0.05. All tests were two-sided.

**Results**

Previous surgical treatment and associated sources of pulmonary flow are illustrated in Table 1.

**Clinical signs**

Clinical signs and echocardiographic findings are described in Table 2. Mean age at catheterization was \( 8.3 \pm 4.2 \) years (range 1.6 to 17.9 years). Patients with retrograde PBF were significantly older than those without additional flow \( (9.5 \pm 4.3 \) years versus \( 7 \pm 4.3 \) years, \( p = 0.014 \)). Mean systemic oxygen saturation and the presence of fatigue were similar in patients with and without additional PBF. Of the eight patients with superior vena cava syndrome, six (75\%) had additional PBF. Hepatomegaly was found in seven (9\%) patients with additional PBF and in no patient without additional PBF \( (p = 0.09) \).

**Echocardiographic findings**

Pulmonary artery size could be analysed, as expected, in only 54\% of patients without additional PBF, in 47\% of those with associated retrograde PBF and in 68\% of those with associated antegrade PBF \( (p = 0.20) \). Moderate to severe incompetence of AV valve(s) was found in nine patients with additional PBF \( (12\%) \) and in one patient without additional PBF \( (3\%) \). Ventricular dysfunction was found in 13 patients with additional PBF \( (17\%) \) and in four without additional PBF \( (11\%) \).
Table 1  Previous surgery and additional pulmonary blood flow in the three groups of patients.

<table>
<thead>
<tr>
<th>Associated source of PBF</th>
<th>Associated retrograde PBF (n = 37)</th>
<th>Associated antegrade PBF (n = 38)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>26 BT shunt, 4 Norwood, 1 PB</td>
<td>2 Norwood-Sano, 15 pulmonary banding, 3 PB and aortic coarctation repair</td>
</tr>
<tr>
<td></td>
<td>2 PA reconstruction, 6 AVF, 1 BT shunt</td>
<td>1 PA reconstruction, 1 PA reconstruction</td>
</tr>
</tbody>
</table>

Of eight patients with superior vena cava syndrome, two had hepatomegaly, seven had moderate to severe incompetence of AV valve(s) and three had ventricular dysfunction. Pulmonary artery size could be analysed in only four patients and was defined as normal in three and small (less than \(-2\) z-values) in one.

Coexistence of clinical signs of increased central venous pressure and echocardiographic signs of ventricular dysfunction and/or hypoplastic pulmonary arteries identified four patients in whom TCPC would not be low-risk.

Cardiac catheterization

Haemodynamic characteristics are shown in Table 3. Mean PAP was 12.3 ± 4.3 mmHg, mean aortic pressure was 70.3 ± 13.7 mmHg and mean pulmonary artery size (Nakata index) was 209.9 ± 42.7 mm²/m². Although PAP exceeding 16 mmHg was found in 9% of patients without and in 16% of those with associated PBF, this difference was not statistically significant (p = 0.38). Mean aortic pressure was higher in patients with associated retrograde flow, but not significantly so.

Nakata index was significantly higher in patients with associated antegrade flow than in those without associated flow (p = 0.004) or in those with associated retrograde flow (p = 0.002, Fig. 1). Of eight patients with normal Nakata index and PAP exceeding 16 mmHg, one had isolated PCPC, one had associated retrograde flow and six had associated antegrade flow.

There were 16 associated procedures: one BT embolization, 10 patent ductus arteriosus embolizations, one arteriovenous fistula embolization, two dilatations of recoarctation, one pulmonary artery dilatation and one dilatation and stenting of an occluded innominate vein. Interventional catheterization was more often performed in patients with retrograde PBF (27%) than in those with antegrade PBF (8%).

Table 2  Clinical and echocardiographic characteristic of patients.

<table>
<thead>
<tr>
<th></th>
<th>Isolated PCPC (n = 35)</th>
<th>Associated retrograde PBF (n = 37)</th>
<th>Associated antegrade PBF (n = 38)</th>
<th>Group 1,2,3 (p)</th>
<th>Isolated PCPC vs additional PBF (p)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex (F/M)</td>
<td>14/21</td>
<td>17/20</td>
<td>21/17</td>
<td>0.41</td>
<td>0.29</td>
</tr>
<tr>
<td>Age, years</td>
<td>7 ± 4.3</td>
<td>9.5 ± 4.3</td>
<td>8.3 ± 3.9</td>
<td>0.048</td>
<td>0.03</td>
</tr>
<tr>
<td>Systemic oxygen</td>
<td>81 ± 5.9</td>
<td>81 ± 6.8</td>
<td>82 ± 6.7</td>
<td>0.88</td>
<td>0.97</td>
</tr>
<tr>
<td>saturation (mean ± SD)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fatigue, n (%)</td>
<td>19 (54)</td>
<td>16 (43)</td>
<td>19 (50)</td>
<td>0.63</td>
<td>0.45</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>2 (6)</td>
<td>4 (11)</td>
<td>2 (5)</td>
<td>0.65</td>
<td>1</td>
</tr>
<tr>
<td>syndrome, n (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hepatomegaly, n (%)</td>
<td>0</td>
<td>3 (8)</td>
<td>4 (11)</td>
<td>0.17</td>
<td>0.09</td>
</tr>
<tr>
<td>Visualization of PA,</td>
<td>19 (54)</td>
<td>18 (47)</td>
<td>26 (68)</td>
<td>0.20</td>
<td>0.67</td>
</tr>
<tr>
<td>n (%)</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Incompetence of AV</td>
<td>1 (3)</td>
<td>5 (14)</td>
<td>4 (11)</td>
<td>0.31</td>
<td>0.16</td>
</tr>
<tr>
<td>valve(s), n (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Ventricular</td>
<td>4 (11)</td>
<td>6 (16)</td>
<td>7 (18)</td>
<td>0.70</td>
<td>0.42</td>
</tr>
<tr>
<td>dysfunction, n (%)</td>
<td></td>
<td></td>
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</table>

AV: arteriovenous; PA: pulmonary arteries; PBF: pulmonary blood flow; PCPC: partial cavopulmonary connection.
or isolated PCPC (9%) \((p=0.03)\). There were no immediate or late complications of cardiac catheterization.

In accordance with the results of haemodynamic evaluation, TCPC was contraindicated in 11 patients (two with isolated PCPC and nine with additional PBF, \(p=0.50\)). Reasons for contraindication were: coexistence of \(PAP\) exceeding 16 mmHg and Nakata index less than 200 in four patients, ventricular dysfunction in four and lack of normalization of \(PAP\) after test occlusion of pulmonary banding in one. Therefore, haemodynamic investigation confirmed that clinical and echocardiographic evaluation identified the four patients in whom TCPC was not considered low-risk because of ventricular dysfunction, but was necessary to identify the remaining seven patients.

**Follow-up**

Eighty-four patients had TCPC at a median of 62 days after cardiac catheterization (2 days to 1.7 years); in 73, a fenestration was created. Thirty-eight patients had complications (23 had effusions, four arrhythmias, four cerebral embolism, four sepsis, two ventricular dysfunction, two bleeding). There were two perioperative deaths due to haemorrhage \((n=1)\) and ventricular dysfunction \((n=1)\). One of the eight patients with superior vena cava syndrome had TCPC, complicated by pleural effusion. Neither preoperative \(PAP\) nor Nakata index were predictors of postoperative complications and/or death.

At the time of writing, 15 patients (one with superior vena cava syndrome) are on a waiting list for TCPC. Of four patients in whom TCPC was contraindicated because of ventricular dysfunction, two underwent cardiac transplantation. There were three deaths, 2 to 4.6 years after catheterization, but these were not causally related to catheterization. Six patients are living with PCPC and additional PBF, without major systemic desaturation or severe ventricular dysfunction.

**Discussion**

This study shows that cardiac catheterization was necessary to identify patients with normal pulmonary artery size and elevated \(PAP\). The results showed that:

- clinical signs identified only a minority of patients in whom TCPC was contraindicated;
- echocardiography allowed measurement of pulmonary artery size in a minority of patients, although it identified all patients in whom TCPC was contraindicated because of ventricular dysfunction;
- \(PAP\) was elevated in a subgroup of patients with normal pulmonary artery size and associated antegrade or retrograde PBF;
- interventional catheterization was often needed in patients with associated retrograde flow, mostly to occlude associated sources of PBF.

Patients with cyanotic heart disease invariably have reduced exercise tolerance [11] and are at risk of cerebral embolism, due to increased blood viscosity and clot formation [12]. Thus, completion of cavopulmonary connection is advocated by most authors, whenever possible. Patients with PCPC are a heterogeneous population because of the possible presence of additional flow to the lungs.
Although many authors do not agree with this policy [13], some surgeons of the French school often prefer to add or preserve additional PBF, in order to maintain systemic saturation around 80% and perform TCPC at an older age. Thus, in our series, TCPC was performed in relatively old patients, mostly when there was at least one source of retrograde PBF.

As previously shown, we confirmed that additional PBF prevents the decrease of pulmonary artery size observed after PCPC [2,14]. Maintaining pulsatile pulmonary flow and adequate total pulmonary perfusion might promote pulmonary artery growth [15]. It has been suggested that, in the absence of additional flow, TCPC should be performed early after PCPC to prevent excessive decrease of the Nakata index [14]. However, several reports describe potential disadvantages of additional sources of PBF. BT shunt promotes distortion and/or stenosis of pulmonary arteries and requires reconstruction of the vessels at the time of PCPC or before TCPC [16]. In our series, three out of 38 patients with BT shunt underwent pulmonary artery repair. Antegrade pulsatile flow promotes pulmonary artery growth [15]; however, it could prompt the development of pulmonary vascular obstructive disease and preclude TCPC [16].

We confirmed that patients with additional antegrade flow have larger pulmonary arteries and found increased PAP in a subgroup of patients with antegrade PBF and normal Nakata index. As previously reported, we also observed pulmonary artery distortion in patients with pulmonary banding (two out of 18 in our series) or interposition of a conduit between the single ventricle and the pulmonary arteries [17].

High-quality imaging of pulmonary arteries in PCPC patients is mandatory before TCPC. Non-invasive imaging of pulmonary arteries using multislice CT or magnetic resonance imaging allows a precise definition of pulmonary artery size and of associated anomalies [18,19]. However, at the time of this study, we could measure pulmonary artery size only using echocardiography and angiography. We confirmed that echocardiography is not appropriate to visualize pulmonary artery size in patients with PCPC [19].

Although we found normal PAP in all but one patient with normal Nakata index and isolated PCPC, we found elevated PAP in a subgroup of patients with associated PBF and normal pulmonary artery size. Thus, correct imaging of pulmonary arteries could be insufficient to authorize TCPC in a subgroup of patients.

We believe that high-quality non-invasive imaging of pulmonary arteries associated with clinical evaluation could avoid cardiac catheterization in patients with isolated PCPC but not in those with additional sources of PBF. A prospective study in patients scheduled for TCPC is ongoing in our institution to evaluate the information provided by cardiac catheterization and CT scan. At present, measurement of PAP with non-invasive methods remains experimental [20].

Apart from information concerning pulmonary artery anatomy and PAP, a relevant percentage of our patients with PCPC needed interventional catheterization to treat residual aortic coarctation, dilate pulmonary arteries and suppress additional sources of PBF. Preoperative closure of BT shunt or patent ductus arteriosus, although potentially accompanied by a decrease of systemic saturation, avoids uncontrolled bleeding during TCPC.

Preoperative PAP is not a predictor of complicated postoperative outcome [17]. We confirmed these results and suppose that the lack of correlation is due to the fact that patients with elevated PAP were not considered candidates for TCPC. Similarly, we confirmed that pulmonary artery size, depressed ejection fraction and AV valve regurgitation, well-known risk factors influencing early and late mortality after TCPC [9], were not predictors of postoperative complications in our series, probably due to patient selection.

Study limitations

Although the study population was relatively large, the majority of our patients had associated sources of antegrade or retrograde flow to the lungs. Thus, the difference between groups rarely reached statistical significance. During the study period, non-invasive imaging of pulmonary arteries was not performed systematically.

Conclusion

Cardiac catheterization is still recommended before TCPC, mostly in patients with additional PBF, to identify patients with high PAP and those needing interventional catheterization. The comparison between the information provided by cardiac catheterization and CT scan will be the object of a future study.

References
