Restrictive left atrial outflow adversely affects outcome after the modified Norwood procedure

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Abstract

Objective: Moderate restrictive foramen ovale in neonates with hypoplastic left heart syndrome (HLHS) is considered to be favourable, reducing pulmonary overcirculation, before modified Norwood operation. However, some newborns with severe restriction of interatrial communication will have pulmonary vascular disease at birth, which correlates with increased perioperative mortality. This article studies the post-Norwood hemodynamic patterns and outcome for the particular group of HLHS newborns with restrictive left atrial outflow compared to other patients. Methods: Restrictive left atrial outflow is defined as mitral and/or aortic atresia with intact ventricular septum, and restrictive foramen ovale, with 3 mm diameter or less with mean interatrial pressure gradient more than 5 mmHg at preoperative echo-Doppler. Four neonates fulfilled these criteria among 18 consecutive patients, who underwent Norwood procedure from October 2002 to December 2003. Mean arterial pressure, heart rate, mean common atrial pressure, urinary output, central venous and arterial oximetry data, serum lactate levels, and dosages of milrinone, phentolamine and norepinephrine were collected at 0, 6, 12, 18 and 24 h after operation. Data were summarized as mean ± SEM. For univariate comparison of different variables, Student’s t-test was used. Results: The postoperative hemodynamic pattern of patients with restrictive left atrial outflow was characterized by hypoxemia and low cardiac output. Arterial (66 ± 3.0% vs 76 ± 1.0%, P = 0.01) and central venous (37 ± 1.2 vs 52 ± 1.1%, P = 0.001) oxygen saturations were much lower than in patients without restriction. Arterio-venous oxygen saturation difference was wider (29 ± 2.4% vs 23 ± 0.9%, P = 0.02) and serum lactate levels were higher (10.8 ± 3.0 vs 2.8 ± 0.2 mmol/l, P = 0.03). Common atrial pressures were more elevated (12 ± 0.8 vs 8 ± 0.3 mmHg, P < 0.001) and higher norepinephrine doses were needed (0.44 ± 0.15 vs 0.06 ± 0.01 μg/kg/min, P = 0.03). The difference for the mean arterial pressures did not reach the significance level (48 ± 2.0 vs 51 ± 2.0 mmHg, P = 0.2). Operative mortality was higher 75% (3/4) compared to 14.3% (2/14, P = 0.04) for the other patients. Conclusions: Restrictive left atrial outflow adversely affects outcome after modified Norwood procedure. Abnormal pulmonary vasculature leading to insufficient pulmonary perfusion is incriminated. To improve outcome, implantation of larger size modified Blalock-Taussig or right ventricle-to-pulmonary artery shunts and routine use of postoperative mechanical assist device should be considered.

Keywords: CHD; Norwood; Hypoplastic left heart syndrome; Pulmonary vascular resistance; Hemodynamics; Outcomes

1. Introduction

Since Norwood reported in 1983 about the first patient with hypoplastic left heart syndrome (HLHS) who successfully underwent the first stage palliation and subsequently the Fontan operation [1], there have been numerous surgical and management improvements, leading to increased survival for neonates with HLHS. The perioperative mortality of the Norwood procedure, which is now applied to a variety of complex cardiac defects with functional single ventricle and systemic outflow tract obstruction, is reported to be around 10-20% in specialized centers [2,6]. On the other hand, the operative risk remains higher for patients with additional extracardiac and cardiac anomalies, in particular with severe restriction of interatrial communication, not only after reconstructive surgery but also while awaiting orthotopic cardiac transplantation, because of pulmonary venous hypertension [2-4].

In October 2002 we introduced continuous monitoring of systemic venous oxygen saturation in our management protocol of the Norwood procedure. This resulted in overall improvement of outcome. However, it brought little change in the postoperative evolution for those neonates with preoperative restrictive left atrial outflow. This article studies the post-Norwood hemodynamic patterns and outcome for this particular group of HLHS patients with limited exit from left atrium compared to patients without outlet obstruction out of left atrium.

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2. Methods

2.1. Definition

Restrictive left atrial outflow is defined as mitral and/or aortic atresia, with intact ventricular septum, and restrictive interatrial communication (Fig. 1). This is judged restrictive at echo-Doppler echocardiography when the maximal width of the color flow jet through the atrial septal defect is 3 mm or less from subcostal, long- and short-axis views and the mean interatrial Doppler gradient is more than 5 mmHg, measured during the entire cardiac cycle, over three consecutive beats, from the subcostal views without angle correction.

2.2. Patients

Between October 2002 and December 2003, 18 consecutive patients at German Paediatric Heart Institute, Sankt Augustin, Germany underwent modified Norwood operation for HLHS or for complex forms of single ventricle with systemic outflow obstruction. Four patients (group I) fulfilled the criteria of restrictive left atrial outflow with a mean interatrial communication of $1.8 \pm 0.5$ mm, range 1-3 mm and interatrial gradient $16 \pm 4.6$, range 8-24 mmHg. 14 patients did not have restrictive left atrial outflow (group II). Atrioseptostomy was not performed. Age, weight, diagnosis and preoperative clinical condition were not statistically different between the two groups (Table 1). All patients of group I were neonates and had classic HLHS anatomy [5].

![Fig. 1. Definition ‘Restrictive left atrial outflow’: Patients with HLHS and ‘restrictive left atrial outflow’ have a restrictive PFO, mitral atresia and/or aortic atresia with intact ventricular septum. The only exit for pulmonary venous blood is through the PFO. Restriction of PFO therefore, directly translates into restriction of the pulmonary venous return. Ao, aorta, LA, left atrium, RA, right atrium, RV, right ventricle, PA, pulmonary artery, PDA, persistent ductus arteriosus, PFO, persistent foramen ovale, rPFO, restrictive persistent foramen ovale. Arrows indicate pulmonary venous blood return, VS: ventricular septum.](image)

Table 1

<table>
<thead>
<tr>
<th>Variable</th>
<th>Group I (n=4)</th>
<th>Group II (n=14)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median age (days)</td>
<td>11 (8-14)</td>
<td>8 (4.275)</td>
<td>0.55</td>
</tr>
<tr>
<td>Weight (g)</td>
<td>3545±100</td>
<td>3375±165</td>
<td>0.42</td>
</tr>
<tr>
<td>Diagnosis</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>HLHS (n pts.)</td>
<td>4</td>
<td>8</td>
<td>0.25</td>
</tr>
<tr>
<td>DILV + TGA, complex IAA, TA (n pts.)</td>
<td>0</td>
<td>6</td>
<td>0.25</td>
</tr>
<tr>
<td>Aortic atresia (n pts.)</td>
<td>3</td>
<td>4</td>
<td>0.27</td>
</tr>
<tr>
<td>Mitral atresia (n pts.)</td>
<td>2</td>
<td>2</td>
<td>0.19</td>
</tr>
<tr>
<td>Aortic annulus diameter (mm)</td>
<td>4.5±1.2</td>
<td>5.5±0.5</td>
<td>0.39</td>
</tr>
<tr>
<td>Tricuspid incompetence mild</td>
<td>3</td>
<td>7</td>
<td>0.60</td>
</tr>
<tr>
<td>Preoperative condition</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SaO2 (%)</td>
<td>81±2</td>
<td>88±2</td>
<td>0.06</td>
</tr>
<tr>
<td>Rate of respiration (breaths/min)</td>
<td>58±10</td>
<td>69±5</td>
<td>0.32</td>
</tr>
<tr>
<td>Mechanical ventilation (n pts)</td>
<td>3</td>
<td>3</td>
<td>0.08</td>
</tr>
<tr>
<td>Inotrope therapy (n pts)</td>
<td>1</td>
<td>2</td>
<td>1.0</td>
</tr>
</tbody>
</table>

Data are displayed as mean±SEM. Group I, pts. with restricted left atrial outflow; Group II, pts. without interatrial restriction; DILV, Double inlet left ventricle; HLHS, hypoplastic left heart syndrome; IAA, interrupted aortic arch; SaO2, systemic arterial oxygen saturation; TA, Tricuspid atresia; TGA, transposition of the great arteries.

2.3. Surgical management

Perioperative management and operative conduct followed a multidisciplinary protocol. This included appropriate preoperative stabilization, operation with aortic arch augmentation using pulmonary homograft material, atrial septectomy, and placement of a polytetrafluoroethylene (Goretex®; W.L. Gore & Associates, Inc., Flagstaff, AZ) shunt either from the innominate artery ($n=14$, diameter 3.5-5 mm) or from the right ventricle ($n=4$, diameter 5 or 6 mm) to the pulmonary artery according to the surgeons’ preference. All patients were cooled down to 18° nasopharyngeal temperature on cardiopulmonary bypass (CPB), 3 mg/kg phentolamine being administered to facilitate cooling and rewarming. The aortic arch was reconstructed with continuous antegrade cerebral perfusion via the MBTS anastomosed to the innominate artery. In case of extended reconstruction of the aortic arch to the proximal ascending aorta the heart was selectively perfused via a cardiopulmonary bypass placed in the ascending aorta just above the aortic annulus. This allowed performing the Norwood procedure on a beating heart in 4 patients, with only a short period of complete circulatory arrest for atrioseptectomy. After completion of operation, the heart was assisted with partial CPB as long as necessary to achieve serum lactate levels below 4 mmol/l and normal sinus rhythm. Rarely, sequential atrioventricular pacemaker stimulation was instituted before weaning the patient from CPB. Operative techniques and CPB times were not statistically different for the two groups of patients (Table 2). Modified ultrafiltration was always applied.

Oximetric catheters (4F Edwards Life Sciences, Irvine, CA, USA) were placed through the right atrium into the superior vena cava to allow continuous monitoring of systemic venous oxygen saturation ($SvO_2$) [6]. An additional line was placed in the common atrium for pressures monitoring and infusion of inotropic drugs. Sternal was routinely left open usually for two days until hemodynamic stable conditions were achieved.
Table 2
Operative patients characteristics

<table>
<thead>
<tr>
<th>Variable</th>
<th>Group I (n=4)</th>
<th>Group II (n=14)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Operation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Norwood operation (n pts)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>with MBTS</td>
<td>3</td>
<td>11</td>
<td>0.59</td>
</tr>
<tr>
<td>with RV/PA conduit</td>
<td>1</td>
<td>3</td>
<td>1.0</td>
</tr>
<tr>
<td>Selective coronary perfusion</td>
<td>0</td>
<td>4</td>
<td>0.52</td>
</tr>
<tr>
<td>Complete circulatory arrest time</td>
<td>8.8±1.9</td>
<td>5.6±0.9</td>
<td>0.12</td>
</tr>
<tr>
<td>(min)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cardiopulmonary bypass time (min)</td>
<td>352±37.3</td>
<td>268.1±21</td>
<td>0.11</td>
</tr>
<tr>
<td>Normalized shunt size (mm²/kg)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Diameter/ body weight (mm²/kg)</td>
<td>1.1±0.04</td>
<td>1.1±0.02</td>
<td>0.95</td>
</tr>
<tr>
<td>Cross-sectional area/body weight</td>
<td>3.0±0.1</td>
<td>3.3±0.2</td>
<td>0.32</td>
</tr>
</tbody>
</table>

Data are displayed as mean±SEM. Group I, pts. with restricted left atrial outflow; Group II, pts. without interatrial restriction; MBTS, modified Blalock Taussig shunt; RV/PA conduit, right ventricular to pulmonary artery conduit.

2.4. Postoperative management

All patients received dopamine 3-6 µg/kg/min, milrinone 0.5 µg/kg/min and phentolamine 2-8 µg/kg/min. Norepinephrine was added if supplementary inotropic support became necessary. Postoperative management aimed to achieve a mean systemic arterial blood pressure of about 50 mmHg, a hematocrit between 45 and 55%, a urinary output greater than 1 ml/kg/h, SvO₂ greater than 50%, systemic arterial oxygen saturation (SaO₂) between 75 and 80%. This would correlate with an arteriovenous oxygen saturation difference of about 25%, and a pulmonary to systemic blood flow ratio (Qp/Qs) of about 1. Patients had severe ventricular dysfunction or multi-organ failure. As the patient needed dopamine (6 µg/kg/min) before Norwood procedure, three had mild tricuspid regurgitation and none had severe ventricular dysfunction or multi-organ failure.

2.5. Hemodynamic data collection and statistical analysis

Preoperative and perioperative data were collected retrospectively. Mean systemic arterial pressure, heart rate, mean common atrial pressure, urinary output, blood gas analysis, SaO₂ and SvO₂, standard base excess, serum lactate levels, and dosages of milrinone, phentolamine and norepinephrine were collected to be studied as hemodynamic data at 0, 6, 12, 18 and 24 h after operation or until the patient expired or initiation of extracorporeal membrane oxygenation (ECMO). Zero hour corresponds to the time of patient’s arrival in the intensive care unit. Data were summarized as mean±SEM. Preoperative and operative characteristics of group I and group II patients as well as variables for survivors and non survivors were compared by independent Student’s t-test for parametric data analysis. Levene’s test was used to test for equality of variances. For non-parametric data the Mann-Whitney U test, Fisher’s exact test, or χ² test was used, as appropriate. Analyses were performed using the statistical software package SPSS 11.0 (SPSS Inc., Chicago, IL). Differences were considered statistically significant at a P-value of ≤0.05.

3. Results

3.1. Availability of hemodynamic data

Table 3 displays the number of hemodynamic data available at each time. A total of 10 samples in group I and 69 in group II could be collected for each hemodynamic parameter and were analyzed. The unavailability of data was due to either early death (n=3) or the use of ECMO (n=1).

3.2. Preoperative condition

Patients in group I had a tendency towards lower systemic arterial saturation (81±2 vs. 88±2, P=0.06). They also were more often ventilated before operation (75 vs 21%, P=0.08, Table 1). However, these changes did not reach statistically significance in between the two groups. One patient needed dopamine (6 µg/kg/min) before Norwood procedure, three had mild tricuspid regurgitation and none had severe ventricular dysfunction or multi-organ failure.

3.3. Hemodynamic data analysis

Patients in group I displayed lower arterial oxygen saturations (P<0.001, Table 4), a lower central venous oxygen saturation (P=0.001), and subsequently a wider arterio-venous oxygen saturation difference (P=0.02) with a smaller oxygen excess factor (P<0.001), lower Qs (P=0.001) and a higher serum lactate level (P=0.03), indicating a lower systemic oxygen delivery. As the fraction of inspired oxygen (FiO₂) was similar (P=0.2), and the calculated Qp was lower (P=0.001) these findings corresponded to a lower pulmonary perfusion. Common atrial pressures (filling ventricular pressures) were significantly elevated (P<0.001) and as well as norepinephrine doses (P=0.03). The difference for the systemic mean arterial pressures did not reach significance level (P=0.2).

Table 3
Availability of each postoperative hemodynamic parameter total amount

<table>
<thead>
<tr>
<th>Time after arrival on ICU (hour)</th>
<th>0</th>
<th>6</th>
<th>12</th>
<th>18</th>
<th>24</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group I (n=4)</td>
<td>3</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Group II (n=14)</td>
<td>14</td>
<td>14</td>
<td>14</td>
<td>13</td>
<td>69</td>
</tr>
</tbody>
</table>

The unavailability of data was due to either early death (n=3) or to the use of ECMO (n=1).
3.4. Outcome

There were five operative deaths: mortality of 27.8% (5/18); 95% confidence limit: 9.7–53.4%. The mortality was higher ($P=0.04$) in group I: 75% (3/4) compared to 14.3% (2/14) for group II. Preoperative, operative and postoperative risk factors analysed for hospital mortality are displayed in Table 5. Significant risks for death by univariate analysis were: elevated mean common atrial pressure, low systemic mean arterial pressure, low systemic venous and arterial oxygen saturation, higher lactate levels, and longer CPB times. Details about the fatal cases in the two groups of patients are displayed in Table 6, indicating earlier time of death in hypoxic shock for Group I.

4. Comment

Preoperative stable hemodynamics and preserved end-organ function are key factors for successful stage I palliation. Moderate restriction foramen ovale in neonates with hypoplastic left heart syndrome can reduce excess pulmonary blood flow and help to balance circulations and to improve systemic oxygen delivery before the Norwood operation. However, absent interatrial communication or a severe restriction (seen in about 6–10% of all HLHS cases) results in pulmonary venous hypertension, pulmonary oedema, and severe hypoxia. Urgent balloon or blade atrioseptostomy is required for survival accompanied by poor results as previously reported [3,9]. Indeed pulmonary vascular disease can develop very early, already during fetal life, causing intractable vascular resistance. Maldevelopment of pulmonary vasculature, increased arterial medial thickness with muscular extension to the smaller bronchioles, intralobular pulmonary artery tortuosity, pulmonary vein elastic hyperplasia and dilated lymphatics have been described before in several studies [3,4,10–12]. Long-term effects of these histopathologic abnormalities for the few cases surviving Norwood first stage are unknown. Rychik et al. [3] reported that only 3 out of 12 patients with HLHS and intact atrial septal defect survived the second stage palliation. In our study the patients with restrictive left atrial outflow, defined as restrictive foramen ovale and mitral and/or aortic atresia, were hemodynamically stable and could undergo reconstructive surgery without balloon atrioseptostomy, even if preoperatively pulmonary function was impaired, mirrored by lower arterial oxygen saturations and higher rate of ventilation (Table 1). This HLHS subgroup represented 22% (4/18) of our Norwood cases in the reported period, which is a relative high incidence. No reference was found in the literature matching the definition of ‘restrictive left atrial outflow’ adopted in this report.

After Norwood procedure the hemodynamic pattern of this specific subset of patients after the Norwood procedure was characterized by hypoxemia and low cardiac output, in particular with low central venous oxygen saturation (below 40%) and high serum lactate level (above 7 mmol/l, Tab 4). These complications were seen in all patients, even in the only survivor of this group. He required prolonged ventilation (9 days), high dosages of inotropes to support cardiac output, and peritoneal dialysis for renal failure. Data analysis allows to presume that pulmonary flow was reduced in all cases of this group. Increased vascular resistance secondary to preoperative pulmonary vasculature changes...
must therefore, be assumed. Qp/Qs was not different in between groups, reflecting our aim to achieve a Qp/Qs of around 1, to maximize systemic oxygen delivery, perioperatively.

In view of these findings, surgical techniques need to be discussed in order to provide more blood flow to an abnormal pulmonary vascular bed. Should larger shunts be used? A computational model stimulation of the post Norwood circulation demonstrated that larger shunts divert an increased proportion of the cardiac output to the lungs, and away from the systemic circulation [13]. Subsequently, a rather small sized shunt should be preferred to decrease pulmonary blood flow in patients with presumably normal or near normal pulmonary vascular bed. On the other hand, in case of increased pulmonary resistance, an adequately sized modified Blalock-Taussig shunt (MBTS) (eg. 3.5 mm for a 3 kg neonate) may be too small to achieve sufficient oxygenation.

There are several theoretical advantages in using a rather large or less restrictive MBTS. First, most patients preoperatively have signs of pulmonary overcirculation, with SaO2 in around 90% and tachypnoea, but they can remain hemodynamically stable even with unrestricted flow to the lungs. Secondly, pulmonary vascular disease is common in HLHS even without pulmonary venous obstruction [12]. Third, pulmonary overcirculation accompanied by a large shunt can be easier be handled by decreasing systemic afterload to balance circulations rather than using a restrictive shunt and having to increase systemic afterload. Reduction of left-to-right shunt can be achieved by increasing haemoglobin concentration [14], or by decreasing systemic afterload with α-adrenoceptor antagonists, angiotensin converting enzyme inhibitors and β-adrenoceptor antagonists [15,16]. Continuous monitoring of patient’s central venous oxygen saturation helps to early detect circulations imbalance and to adjust afterload reduction. In fact, narrower arterio-venous oxygen saturation difference and higher SvO2 were seen in patients who received a larger shunt within the postoperative hours 4-24 [15]. Similarly, Photiadis et al [17] observed better postoperative hemodynamics and less inotropic support within the first 48 h in patients who received shunts, with a normalized cross-sectional MBTS area larger than 3.3 mm²/kg. Fourth, the period of pulmonary overcirculation with a large shunt is limited, since the conversion of the source of pulmonary blood flow from a systemic to pulmonary shunt to a cavopulmonary anastomosis is now routinely scheduled at the age of 4-6 months, when pulmonary vascular resistance has fallen to normal values [18]. The danger of pulmonary vascular disease, which would prohibit further palliation, may therefore, be considered to be rather small.

About 10% of all patients undergoing Norwood procedure experience sudden cardiovascular collapse accompanied by low cardiac output [19]. Since prediction of this shock is difficult, the routine use of postoperative mechanical assist device for all Norwood cases has been favoured by Ungerleider and associates and survival has been reported to be around 90% [20]. Since the postoperative course of all patients with restrictive left atrial outflow in our study was complicated by low cardiac output syndrome, there should be a low threshold for intiation of ECMO or ventilator assist device.

We conclude that neonates with hypoplastic left heart syndrome associated with restricted left atrial outflow presented with a uniform pattern of hemodynamic response after the modified Norwood operation: with hypoxemia and low cardiac output, detected by continuous monitoring of the central venous oxygen saturation. Abnormal pulmonary vasculature leading to insufficient pulmonary perfusion is incriminated. The patients represent a HLHS subgroup, with dismal postoperative course, despite aggressive ventilator and inotrope manipulations. We therefore, consider the implantation of larger shunts and the liberal use of post-operative mechanical assist device to improve the outcome of this severely ill subgroup of HLHS patients.

References


[21] Ungerleider RM, Shen I, Yeh T, Schultz J, Butler R, Silberbach M, Dr. Photiadis: I really don’t know the answer to this question, to be honest, because we didn’t do any preoperative balloontioptery, because it was not felt to be indicated for hemodynamic or oxygenation reasons.

Appendix A. Conference discussion

Dr. G. Ziemer (Tuebingen, Germany): Would you consider to do anything before surgery, like interventional enlargement of the restrictive interatrial connection and then let the patient stabilize for a week, if possible? Dr. Photiadis: Well, that could be considered. Jack Rychik, of the Philadelphia group published a series of 12 patients with intact atrial septum in two they tried in two balloon manoeuvres to better this, but of both patients died.

Dr. Ziemer: But there is a difference between intact interatrial septum and restrictive interatrial septum. In addition, there is a total difference in fetal circulation, also.

Dr. Photiadis: I really don’t know the answer to this question, to be honest, because we didn’t do any preoperative balloontioptery, because it was not felt to be indicated for hemodynamic or oxygenation reasons.

There was one patient in our institution, with intact atrial septum, in whom an atrial septectomy was performed surgically and inflow occlusion immediately after birth, but unfortunately this patient didn’t survive.

Dr. D. Barron (Birmingham, United Kingdom): I was interested to know what the saturations of the patients were preoperatively.

Dr. Photiadis: They were no significantly different in between groups. They were all around 85 sometimes up to the 90s. So at that stage, it was not obvious, that they may have a problem postoperatively. That’s why we went into it, handled them as usual hypoplasts, but their outcome was worse.

Dr. Barron: I’m just concerned, because sometimes if they have saturations in the 90s, they must actually have a very high Qp.

Dr. Photiadis: You’re right.

Dr. Barron: So how restrictive really is that atrial septum compared to the patients you see who almost present like an obstructed TAPVD where they’re desaturated and very sick? These, to me, seem to be the ones that really do much worse.

Dr. Photiadis: Well, regarding preoperative saturations there was no significant difference in our subsets of patients.

Dr. B. Meyns (Leuven, Belgium): We had a similar experience in a baby who died, and then on the autopsy we found significant changes in the venous pulmonary circulation on the microcirculatory level. We thought that there might be a connection between the two. Do you have any autopsies and specifically the pulmonary autopsies on the babies that died?

Dr. Photiadis: Yes, we did have, out of the 3, we had 2, and those had pulmonary vascular changes.

Dr. B. Asfour (Sankt Augustin, Germany): As I have operated on most of the patients presented in this study, I would like to give just two short comments. Regarding the question mentioned before, to maybe do something before the operation to enlarge the intra-atrial septum—you know that the septum especially in these patients is so thick and so muscular that I think that interventional cardiologists have little chance to tear the inter-atrial septum. As this study shows, I very much prefer using the right sided modified Blalock-Taussig shunt in the Norwood procedure; however, I think that in these selected patients with abnormalities of the pulmonary vasculature maybe the RV to PA conduit may have certain advantages.

Dr. E. Bone (Ann Arbor, Michigan, USA): I thought the question that was asked earlier was a pertinent one, namely, how restrictive really are these atrial septal defects? In our experience, we’ve not identified a difference in outcomes unless the patient presents with a real picture of obstructed total veins, namely, hypoxemia and progressive pulmonary edema. There have been a number of our patients who have had significant gradients across the ASD or PFO by Doppler, sometimes as high as 10 or even 15 mmHg. Many of these patients will have pop-off veins as well. But if they present with a picture of high systemic saturation and no congestion on their chest x-ray, we’ve not seen a difference and have not made an effort to enlarge the ASD prior to the Norwood procedure.

You did ask the question about what to do preoperatively on those patients who really have a severely restrictive ASD. Even in patients with an intact atrial septum, you can actually make an ASD in the cath lab by using a biopsy probe which can make a hole in the septum, get across it, and then either get a balloon or a stent across the defect to enlarge it.

In those conditions we have seen better early survival when we have been able to satisfactorily relieve the obstruction very early in life, sometimes within hours of birth. However, late survival has not been good and most of these patients have not been particularly suitable for continued staged reconstruction. Similar results to ours in Ann Arbor were reported from the Philadelphia group.

Dr. K. Januszewska (Cracow, Poland): In our center, we routinely perform intra-atrial septectomy during the Norwood procedure and we have no problems with restrictive intra-atrial communication after that Norwood operation. What do you think of this strategy?

Dr. Photiadis: There was no question about doing the atrial septectomy during the Norwood procedure. That’s routinely done.

What I want to emphasize, that since 1983, when Norwood first successfully performed a Fontan operation after initial of Norwood operation, mortality has significantly dropped to about 20%. If we want to approach the patients, we still have problems with, e.g. those with interrupted aortic arch and anomalous pulmonary veins, we need to have a proper monitoring. Continuous monitoring of systemic venous oxygen saturations, that has been introduced by Tweddell and colleagues is to be recognized as the state of the art monitoring for this patient group, today.


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