Does the shunt type determine mid-term outcome after Norwood operation?†

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Abstract

OBJECTIVES: With improved short-term outcomes the right ventricular to pulmonary artery shunt (Sano) has become the preferred pulmonary blood source in the Norwood procedure in many centres. However, most studies analysed consecutive cohorts, with a first modified Blalock–Taussig shunt (BT) followed by the Sano cohort. Besides, neither comprehensive preoperative risk analysis nor outcome beyond 1 year of age was investigated.

METHODS: This study reviews 109 neonates undergoing the Norwood procedure in the same interval between October 2002 and December 2009. The Sano (38) or BT shunt (71) was assigned according to the surgeon’s preference. Two neonates subsequently underwent successful biventricular repair and were excluded. The Aristotle comprehensive score (ACS) was used to evaluate preoperative risk, with high-risk patients (n = 39) classified as having an ACS ≥20, and low-risk patients (n = 68) given an ACS <20.

RESULTS: Mean Aristotle score at the Norwood operation was 18.8 ± 0.4 and 18.9 ± 0.3 (P = 0.9) in Sano and BT, respectively. Mean follow-up interval was 4.1 ± 2.1 years (range: 1.7–8.9 years). Actuarial survival was similar, stabilizing from the 8th postoperative month onwards at 78.6 ± 4.9% (95% CI: 67.0–86.5%) for Sano and 78.4 ± 6.8% (95% CI: 61.4–88.6%) for BT; P = 0.95. Midterm actuarial survival was higher in low-risk patients, 88.2 ± 3.9% (95% CI: 77.8–93.9%) than in high-risk patients: 61.5 ± 7.8% (95% CI: 44.5–74.7%, P = 0.0003). No survival benefit was detected in low- or high-risk cases for either shunt type. Risk factors for midterm mortality were cardiorespiratory failure requiring ventilation (13/34, P = 0.004), and ACS ≥20 (15/39, P = 0.001), but not shunt type (8/37, P = 0.95). Increased number of shunt-related interventions before the Glenn procedure were noted with Sano (32.4 versus 6.5%, P = 0.002).

CONCLUSIONS: Preoperative risk factors, regardless of shunt type, influence midterm survival after the Norwood procedure with an excellent outcome in low-risk patients, while high-risk cases still incur a significant mortality. Sano shunt interventions occurred with increased numbers. Although, Sano shunt may be the only feasible option in some instances, given the possible negative effects of ventriculotomy on right ventricle function, the widespread use of Sano shunt should be reconsidered.

Keywords: Hypoplastic left heart syndrome • Norwood • Sano • RV–PA conduit • Aristotle score

INTRODUCTION

Two primary treatment strategies for neonates with hypoplastic left heart syndrome are currently applied in most congenital heart centres. In the ‘classic’ Norwood procedure (NP) the pulmonary blood flow is provided by a modified Blalock–Taussig shunt (BT), a polytetrafluoroethylene graft connecting the subclavian or innominate artery to the pulmonary artery. The second main procedure is called the Sano modification, in which a right ventricle to pulmonary artery shunt (Sano shunt) provides adequate pulmonary blood flow.

Several potential advantages of the Sano over the BT shunt were discussed as related to the improvement of short-term outcome, such as the absence of diastolic run-off and improved coronary perfusion. This was shown to correlate with a more stable perioperative course, improved weight gain and mortality [1]. Potential disadvantages in the long term could include a deleterious effect on right ventricular function, arrhythmias or aneurysm formation due to the ventriculotomy, greater remodelling of the ventricular myocardial extracellular matrix [2], additional volume load following regurgitation from the non-valved...
RV–PA shunt, impaired growth of the pulmonary artery, higher intervention rates on the Sano shunt and the need for an earlier stage II procedure [3, 4]. Most of these studies were not randomized, overlooking consecutive cohorts (a BT cohort followed by the Sano cohort). Therefore, it is unclear whether superior outcome could also be related to improved perioperative care or mere chance. A recently published multi-institutional randomized study has shown an improved early transplant-free survival for the Sano modification. However, there was no survival benefit beyond 12 months, due to higher numbers of unintended cardiovascular interventions and complications in the Sano subgroup during the first year [4].

This study evaluates the midterm survival of neonates with hypoplastic left heart syndromes or variants. These underwent a Norwood operation with Sano or BT shunts in the same time interval. The study took place in a single congenital heart centre with a follow-up interval of at least 18 months. The Aristotle comprehensive score was used to compare preoperative and postoperative risk between study groups.

**METHODS**

**Patients**

The 109 neonates included in the study underwent the NP for hypoplastic left heart syndrome (n = 83) or variants of hypoplastic left heart syndrome (n = 26) from October 2002 to December 2009. Two patients (Sano and BT shunt types) had successful biventricular repair and were removed from further analysis. No patient was denied surgery in the period because of preoperative poor condition. Heart transplantation was not a treatment option. Males predominated (n = 69; 79.5%). The age at the time of operation ranged from 1 to 27 days (mean 7.8 ± 0.4 days); the weight was 3.2 ± 0.05 kg (range: 1.7–4.4 kg).

**Management**

A multidisciplinary protocol was followed as described previously [5]. Unstable patients and those referred in shock were ventilated preoperatively and stabilized using inotropic after load reduction therapy with dopamine 3–6 µg/kg/min, milrinone 0.5 µg/kg/min and phentolamine 2–8 µg/kg/min. If pulmonary overcirculation could not be controlled medically, NP was undertaken. Treatment of patients with obstructive pulmonary venous return was conducted as described before [6]. Surgery included aortic arch augmentation using pulmonary homograft material, atrial septectomy and placement of a polytetrafluoroethylene shunt from either the innominate artery (BT, n = 70, diameter 3.5–5 mm) or the right ventricle (Sano, n = 37, diameter 5–6 mm) to the pulmonary artery, the Sano shunt being placed on the right side of the augmented ascending aorta. The aortic arch was reconstructed during continuous antegrade cerebral perfusion. Modified ultrafiltration was applied in every case. Monitoring of systemic venous oxygen saturation (SvO₂) was performed to estimate Qp/Qs in all patients. Inotropic management usually included dopamine 3–6 µg/kg/min and milrinone 0.5–0.9 µg/kg/min. Norepinephrine or epinephrine was added if supplementary inotropic support became necessary. Infusion of milrinone or phentolamine (2–8 µg/kg/min), was commenced in cases of low SvO₂, acidosis, rising lactate levels or low urinary output. The sternum was generally left open for 1 day (median duration) to achieve haemodynamic stabilization. Rapid-response extracorporeal membrane oxygenation (ECMO), used selectively since 2003, was installed for 11 neonates with low cardiac output. Survivors were treated with aspirin 2 mg/kg daily and usually with digoxin, carvedilol (0.1–1 mg/kg/day) or captopril (treatment of pulmonary overcirculation) until the second-stage palliation. Patient’s parents were trained for daily monitoring of weight and peripheral oxygen saturations at discharge [7]. The results were checked and confirmed by weekly telephone or E-mail contact. Haemoglobin levels were monitored every other week and maintained at the level of 14 g/dl. After a period of 3–6 months or when the patient’s bodyweight reached 5 kg, cardiac catheterization was performed followed by bidirectional cavopulmonary (Glenn) anastomosis. Shunt-banding intervention was indicated in case of pulmonary circulation associated with a low systemic output not responsive to medical therapy. Interventional dilation and/or stenting was realised to manage shunt stenosis or kinking associated with oxygen saturation <70% at rest. Dilation with or without stenting of aortic isthmus was indicated when the systolic blood pressure gradient went over 30 mmHg or if the calibre dropped >30% from arch to descending aorta. The Fontan circulation by means of extracardiac total cavopulmonary connection was completed at 1–3 years of age, or when patients reached a body weight of 10–15 kg. Before this Fontan, a cardiac catheterization was routinely performed, and haemodynamic data and total lower lobe index were collected [8] to assess cardiac function and development of pulmonary vascular bed. Follow-up echocardiograms were carried out by an experienced cardiologist and ventricular function was evaluated as normal or mildly depressed, moderately depressed and severely depressed. Tricuspid regurgitation was graded as mildly or more than mildly incompetent if the proximal tricuspid regurgitant jet width was <2.5 mm. It was graded moderate to severe if the jet was at least 2.5 mm [9]. The obstructed pulmonary venous return was defined as a persistent foramen ovale diameter of <2.5 mm on the Doppler colour flow mapping, or as a continuous non-phasic flow with a velocity ≥1.2 m/s on the pulse Doppler echocardiography sampling [10].

**Data collection and statistical analysis**

Preoperative and perioperative data were collected retrospectively to estimate the Aristotle comprehensive score. Preoperative risk was evaluated by means of the Aristotle complexity score [11]. For further analysis of the clinical course, patients with an Aristotle comprehensive score of 20 and more were classified as high risk and those with a score under 20 as low risk [12]. Inquiry for follow-up and late survival took place at the end of August 2011. Follow-up data collection were complete. Analysis for mortality was limited to factors affecting at least four patients. Kaplan–Meier curves for actuarial survival were calculated using the GraphPad Prism (San Diego, CA, USA). The log-rank test assessed the statistical differences between two groups. All other (univariate) analyses were performed using the statistical software package SPSS 18.0 (SPSS, Inc., Chicago, IL, USA). Data were summarized as mean ± SEM. Independent Student’s t-test was used to compare means of parametric variable for significant differences between groups. Levene’s test was employed to test for equality of variances. The χ² test or Fisher’s exact test was used.
for non-parametric data analysis as appropriate. A difference was considered significant at a P-value of <0.05.

**RESULTS**

**Preoperative risk factors**

As displayed in Table 1, preoperative and operative data as well as hospital stay after Norwood, Glenn and Fontan operations did not reveal significant differences between BT and Sano groups of patients, except for the age at the time of the Norwood operation: 6.6 ± 0.5 days versus 8.5 ± 0.6 days (P = 0.01), respectively. In each group, 26 patients reached the stage of the Fontan operation at a comparable mean age (2.9 ± 0.2 and 3.4 ± 0.3 years) with statistically equivalent mean weights (13.1 ± 0.5 and 13.5 ± 0.3 kg).

Before the NP, factors pertaining to the calculation of the Aristotle comprehensive score (Table 2) as well as the mean Aristotle comprehensive score and the number of patients reaching a complexity score of 20 or more points were not significantly different between the BT and Sano groups (P range = 0.20–0.9).

**Outcome**

Hospital mortality following NP (10.5 versus 12.5%, P = ns), inter-stage mortality (5.9 versus 8.1%, P = ns) and hospital mortality following Glenn operation (6.5 versus 1.8%, P = ns) were similar for Sano and BT, respectively (Fig. 1). The mean follow-up interval was significantly longer in the Sano group (4.9 ± 0.3 years) than in the BT group (3.8 ± 0.3 years, P = 0.01). Actuarial survival was similar as of the 8th postoperative (NP) month: 78.6 ± 4.9% (95% CI: 67.0–86.5%) in the Sano group and 78.4 ± 6.8% (95% CI: 61.4–88.6%, P = 0.95) in the BT group (Fig. 2). As illustrated in Fig. 3, survival was significantly superior (P = 0.0003) for patients having an Aristotle score ≤20 points: 88.2 ± 3.9% (95% CI: 77.8–93.9%) when compared with those having a score of 20 points or more: 61.5 ± 7.8% (95% CI: 44.5–74.7%).

**Rapid-response extracorporeal membrane oxygenation support.** Eleven neonates required ECMO support for 3 days after the NP (median, range 0–186 days) because of a low systemic output (n = 10) and a respiratory failure following respiratory syncytial virus infection (n = 1). After a median support time of 4 days (range: 12 h–14 days), seven patients could be weaned and all survived. Incidence of ECMO support was not significantly different between Sano and BT shunt groups. A tendency towards higher incidence of ECMO was observed in patients with Aristotle comprehensive scores of 20 or more (19.3 versus 8.1%, P = 0.1).

**Risk factor analysis.** Operation-dependent and operation-independent risk factors related to the Aristotle comprehensive score analysis were evaluated for Sano and BT shunt groups with respect to hospital and overall mortality.

**Hospital mortality after the Norwood procedure.** Mechanical ventilation to treat cardiorespiratory failure within 48 h before first-stage palliation was a significant risk factor for hospital mortality in the Sano group (4/13 = 30.8%, P = 0.01) and BT group (7/21 = 30%, P = 0.002). Similarly, Aristotle score of 20 or more was a mortality risk factor for both shunt types: Sano, 4/11 = 36.4%, P = 0.005; and BT, 8/28 = 28.6%, P = 0.002. Shock resolved at the time of surgery was not a risk factor for the Sano group with 1/6 = 16.7%, P = ns, but was related to increased mortality in the BT group (5/17 = 29.4%, P = 0.01). Hospital mortality was not related with obstructed pulmonary venous return in the Sano group (0/5 = 0%, P = ns), as opposed to the corresponding results in the BT group (6/18 = 33%, P = 0.007).

Other known risk factors, such as aortic atresia, interrupted aortic arch, aberrant right subclavian artery, atrioventricular valve regurgitation, renal dysfunction, bodyweight under 2.5 kg, prematurity, chromosomal or extracardiac anomaly, combination of mitral stenosis and aortic atresia, were considered and not found

**Table 1:** Patients’ characteristics and operative data

<table>
<thead>
<tr>
<th></th>
<th>Sano</th>
<th>BT shunt</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Norwood (n)</td>
<td>37</td>
<td>70</td>
<td></td>
</tr>
<tr>
<td>Age (day)</td>
<td>6.6 ± 0.5</td>
<td>8.5 ± 0.6</td>
<td>0.01</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>3.2 ± 0.1</td>
<td>3.2 ± 0.1</td>
<td>ns</td>
</tr>
<tr>
<td>Aristotle comprehensive score</td>
<td>18.8 ± 0.4</td>
<td>18.9 ± 0.3</td>
<td>ns</td>
</tr>
<tr>
<td>HLHS (number of patients)</td>
<td>33</td>
<td>33</td>
<td>ns</td>
</tr>
<tr>
<td>ACC (min)</td>
<td>38.2 ± 3.7</td>
<td>37.2 ± 2.9</td>
<td>ns</td>
</tr>
<tr>
<td>ECC (min)</td>
<td>162.8 ± 13.7</td>
<td>181.7 ± 10.5</td>
<td>ns</td>
</tr>
<tr>
<td>Length of hospital stay (days)</td>
<td>27.7 ± 3.2</td>
<td>30.4 ± 2.7</td>
<td>ns</td>
</tr>
<tr>
<td>Bidirectional Glenn (n)</td>
<td>31</td>
<td>56</td>
<td></td>
</tr>
<tr>
<td>Age (month)</td>
<td>5.4 ± 0.3</td>
<td>5.5 ± 0.2</td>
<td>ns</td>
</tr>
<tr>
<td>ECC (min)</td>
<td>65.5 ± 4.7</td>
<td>75.1 ± 5.4</td>
<td>ns</td>
</tr>
<tr>
<td>Length of hospital stay (days)</td>
<td>19.5 ± 2.6</td>
<td>16.3 ± 1.2</td>
<td>ns</td>
</tr>
<tr>
<td>Extracardiac Fontan (n)</td>
<td>26</td>
<td>26</td>
<td></td>
</tr>
<tr>
<td>Age (year)</td>
<td>2.9 ± 0.2</td>
<td>3.4 ± 0.3</td>
<td>ns</td>
</tr>
<tr>
<td>ECC (min)</td>
<td>74.0 ± 6.9</td>
<td>88.8 ± 4.9</td>
<td>ns</td>
</tr>
<tr>
<td>Length of hospital stay (days)</td>
<td>20.5 ± 1.3</td>
<td>18.9 ± 1.3</td>
<td>ns</td>
</tr>
</tbody>
</table>

Values display total number of patients or mean values ± SEM within the shunt group.


**Table 2:** Preoperative risk factors for the Norwood procedure (n = 107)

<table>
<thead>
<tr>
<th>Procedure-dependent factors</th>
<th>Sano, n = 37</th>
<th>BT, n = 70</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic atresia</td>
<td>24</td>
<td>41</td>
<td>ns</td>
</tr>
<tr>
<td>Obstructed pulmonary venous return</td>
<td>5</td>
<td>18</td>
<td>ns</td>
</tr>
<tr>
<td>Aberrant right subclavian artery (arteria lusoria)</td>
<td>4</td>
<td>4</td>
<td>ns</td>
</tr>
<tr>
<td>Procedure independent factors</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mechanical ventilation to treat cardiorespiratory failure</td>
<td>13</td>
<td>21</td>
<td>ns</td>
</tr>
<tr>
<td>Shock resolved at time of surgery</td>
<td>6</td>
<td>17</td>
<td>ns</td>
</tr>
<tr>
<td>Renal dysfunction</td>
<td>12</td>
<td>13</td>
<td>ns</td>
</tr>
<tr>
<td>Aristotle comprehensive score of at least 20 points</td>
<td>10</td>
<td>29</td>
<td>ns</td>
</tr>
</tbody>
</table>

Values display total number in each shunt group with specific risk factor.
to be risk factors for hospital or overall mortality in shunt groups.

**Interstage mortality.** In both Sano and BT groups, the risk factors assessed with the Aristotle comprehensive score were not significantly related with increase in mortality.

**Overall mortality.** There was a tendency towards an increase in mortality in relation to mechanical ventilation applied to treat cardiorespiratory failure for the Sano group 5/13 = 38.5%, $P = 0.1$. A significantly higher mortality was observed in the BT group (8/21 = 38.1%, $P = 0.01$) in the same circumstance. An Aristotle score of 20 or more was a mortality risk factor for both shunt types: 5/11 = 45.5%, $P = 0.04$ (Sano) and 10/28 = 35.7%, $P = 0.04$ (BT). Shock resolved at the time of surgery was not a risk factor for overall mortality with the Sano group with 1/6 = 16.7% ($P = ns$), but was related to mortality in the BT group (7/17, 41.1%, $P = 0.03$). Likewise, hospital mortality was not related with obstructed pulmonary venous return for Sano (0/5 = 0%, $P = ns$), whereas there was a relation for the BT group (hospital mortality of 7/18 = 38.9%, $P = 0.04$).

**Follow-up data**

**Interventions between Norwood and Glenn operations.** Interventions between stages 1 and 2 were common. They consisted of dilation and/or stenting of the aortic arch (Sano: 10/34 = 29.4%; BT: 16/62 = 25.8%, $P = 0.8$). A greater number of shunt-related interventions were recognized in the Sano group 11/34 = 32.4 versus BT 4/62 = 6.5%, $P = 0.002$, mainly dilation or stenting, i.e. Sano in 9/34 = 26.5% and BT in 2/62 = 3.2%, $P = 0.0012$. The other interventions were shunt bandings: Sano in 2/34 = 5.8%, BT in 2/62 = 3.2%, $P = ns$.

**Echocardiography data.** Incidence of more than mildly impaired right ventricular function was similar at the last follow-up echocardiogram, as well as occurrence of more than mild tricuspid regurgitation.
Catheterization data before Fontan completion. End-diastolic pressure was not significantly different at cardiac catheterization before completion of the Fontan circulation. Pulmonary vascular bed evaluated by total lower lobe index was equally developed (Table 3). Only one patient in the Sano group suffered from protein-losing enteropathy. None has required heart transplantation to date.

Table 3: Follow-up data

<table>
<thead>
<tr>
<th></th>
<th>Sano</th>
<th>BT shunt</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients on follow-up</td>
<td>30</td>
<td>55</td>
<td></td>
</tr>
<tr>
<td>Follow-up interval (years)</td>
<td>4.9 ± 0.3</td>
<td>3.8 ± 0.3</td>
<td>0.01</td>
</tr>
<tr>
<td>Echocardiographic findings</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderately impaired ventricular function</td>
<td>20</td>
<td>23</td>
<td>ns</td>
</tr>
<tr>
<td>Number of patients &gt; mild tricuspid regurgitation</td>
<td>5</td>
<td>16</td>
<td>ns</td>
</tr>
<tr>
<td>Catheterization data</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Number of patients having catheterization before the Fontan operation</td>
<td>26</td>
<td>26</td>
<td></td>
</tr>
<tr>
<td>EDP (mmHg)</td>
<td>9.1 ± 0.6</td>
<td>8.8 ± 0.4</td>
<td>ns</td>
</tr>
<tr>
<td>Total lower lobe index (mm²/m²)</td>
<td>144 ± 8.6</td>
<td>154.7 ± 7.7</td>
<td>ns</td>
</tr>
</tbody>
</table>

Values display total number of patients or mean values ± SEM in each shunt group. EDP: end-diastolic pressure measured during cardiac catheterization before Fontan.

Catheterization data before Fontan completion. End-diastolic pressure was not significantly different at cardiac catheterization before completion of the Fontan circulation. Pulmonary vascular bed evaluated by total lower lobe index was equally developed (Table 3). Only one patient in the Sano group suffered from protein-losing enteropathy. None has required heart transplantation to date.

COMMENT

Since Dr Sano reintroduced the right ventricular to pulmonary shunt using a non-valved Goretex shunt, many others have embraced his idea and were able to demonstrate more stable haemodynamics and improved early outcome [1, 9]. Also, the Sano shunt may be the only possible option for inadequately developed innominate artery or an aberrant subclavian artery. Besides, the Sano shunt may be beneficial in hypoplastic left heart syndrome complicated by coronary fistulae or neoaortic valve insufficiency.

However, most of the available studies were single-centre case series. They displayed higher Sano shunt hospital survival than historical BT controls. But there was no real difference in survival rate according to shunt type for studies with comparable controls [13, 14].

This study involves the Sano and BT shunting methods in a single centre with a follow-up of at least 18 months. A homogeneous group of neonates was analysed; multi-factorial risk evaluation did not reveal significant differences between the two groups. Sano and BT shunt patients were not strictly randomized, but shunt type was selected according to the surgeon’s preference on call.

We could not reveal any survival benefit of one shunt type over the other. Moreover, there was no improved survival in high- or low-risk patients before the Norwood operation with either shunt type. With low preoperative risk, not only hospital but overall midterm survival was remarkably high with survival rates stabilizing at 90% after 8 months for both shunt types. However, high-risk neonates (with an Aristotle comprehensive score of 20 or more) remain a challenge even for an experienced team, as reflected by midterm survival rates of only 60% for Sano as well as BT shunt types. Therefore, a high Aristotle comprehensive score was the most important risk factor for mortality following the NP and overall mortality. Of course, little can be done to reduce the impact of operative-dependent factors, such as total anomalous pulmonary venous connection. These patients presenting with a single ventricle anatomy generally suffer a poor outcome [6, 10].

Hospital mortality of HLHS with obstructive pulmonary venous appears to be an important risk factor for early mortality in the BT shunt group. Whether the Sano shunt bears advantages in this entity remains to be determined. Since the incidence of obstructed pulmonary venous return in the Sano group is small, data should be evaluated with caution. As one could...
argue, with diastolic blood pressure improved by the Sano shunt, better pulmonary blood flow could be provided at the onset of an increase in pulmonary resistance (commonly seen after repair of obstructive pulmonary venous return).

The incidence of obstructive pulmonary venous return was relatively high (21.5%, 23/107), which reflects an unusual referral pattern of our centre, known to have the largest experience nation-wide with neonates presenting with this condition. Thus such patients were sent from all over Germany and even from neighbouring countries.

Shock resolved at time of surgery was not a risk factor for mortality in the Sano group, but was related to increased mortality in the BT shunt group. However, the incidence of this risk factor within the Sano group was minor, thus the survival benefit for the Sano shunt in this condition remains unclear. Other well-known risk factors for early Norwood mortality such as typical HLHS compared with its variants or the combination of mitral stenosis and aortic atresia were not associated with the increase in mortality in our cohort.

With evolving techniques and experience, prenatal diagnosis of HLHS will allow easy referral or even delivery in a specialized paediatric heart centre. With this strategy neonates are unlikely to deteriorate preoperatively and require ventilation because of cardiorespiratory failure. This preoperative ventilation is indeed the second most important risk factor (P < 0.001) for both groups of patients (Sano, P = 0.01; BT, P = 0.002). It continues to influence even midterm survival, confirming the importance of prenatal diagnosis. Similarly, Tworetzky et al. found improved early survival with prenatal diagnosis of HLHS. Others did not find a survival benefit, but after all they identified fewer adverse perioperative neurologic events.

An alternative approach for high-risk patients is the newly introduced hybrid operation, combining bilateral pulmonary banding and ductus arteriosus stenting. Early results are promising for selected patients, but interstage mortality remains high. Furthermore survival benefits could not be demonstrated in high-risk patients.

This study confirms that by means of the Aristotle comprehensive score, not only can case complexity be evaluated, but also midterm survival can be predicted. Outcome appears to be consistently related with status and concomitant cardiac and non-cardiac malformations at the time of the NP. Midterm survival is excellent for both groups with survival rates stabilizing beyond 1 year of age at ~80%, for the whole cohort, including low- and high-risk patients. Our Norwood hospital mortality compared favourably with the hospital mortality reported in a recently published multi-institutional series which was as high as 23.7% (95% CI: 22.0–25.4%). Ideally one should compare results according to case complexity such as the Aristotle complexity score. This scoring method moreover addresses the issue of patients' selection.

As for other complex congenital heart lesions, survival tends to stabilize 1 year after surgery. Late survival rates are heavily influenced by early mortality. The 1-year survival rate of 79.4% ± 3.9% in this series represents a significant improvement in comparison with the 60–70% survival benefit in the multi-institutional study reported by a multicentric study.

Like Ohye et al., we noticed more unintended interventions for the Sano shunt. This may be caused by the path of the Sano shunt across the anterior surface of the right ventricle directly posterior to the sternum. With the enlargement of the ventricle due to growth or dilation, the shunt stretches and is prone to kinking and stenosis, especially at proximal and distal anastomosis. Immediate intervention with ballooning and stenting re-established adequate pulmonary flow. This may explain similar pulmonary artery growth for both study groups before the Fontan operation. Other workers found improved pulmonary artery growth with the Sano shunt, arguing wider blood pressure amplitudes. However, these findings do not seem to be sustainable beyond the second stage until the Fontan operation.

Postoperative low cardiac output refractory to medical treatment is common after the NP. Seven out of 11 patients (63.6%) who benefited from ECMO therapy could be saved and are now midterm survivors. Rapid-response ECMO is also known to improve neurological outcome.

**STUDY LIMITATIONS**

There are several limitations to this study. First, this study is retrospective and covers a long period of time. Second, some risk factors affected small numbers of patients, hampering statistical analysis. Although all patients were treated with the same standardized protocol, patient assignment to the Sano shunt group after we experienced a high incidence of shunt-related complications and interventions in 2008. As a result, 2009 only one of 26 patients was elected to have a Sano shunt. This explains a significantly longer follow-up in Sano patients. Echocardiographic evaluation of tricuspid regurgitation according to proximal jet width is straightforward. However, with lack of objective echocardiographic parameters, estimation of right ventricular function is prone to inter-observer variability. Hence, these data should be weighed with caution.

**CONCLUSION**

For either shunt type, no higher survival rate (early- or midterm) was demonstrated, nor was any improved ventricular function observed. Incidence of shunt-related interventions was significantly increased with the Sano shunt, requiring an experienced team of interventional cardiologists and reliable home monitoring to avoid morbidity and mortality.

Midterm survival was excellent for patients with a stable preoperative condition and low Aristotle comprehensive score, reaching almost 90%. Otherwise, it drops to ~60%. As a result, prenatal diagnosis and early referral (at birth) could be of the utmost importance. More time and effort should be devoted to improve the outcome of newborns with complex forms of hypoplastic left heart syndrome.

**Conflict of interest:** none declared.

**REFERENCES**


[3] Lim DS, Peel B, Matherne GP, Kramer CM. Cardiovascular magnetic resonance of pulmonary artery growth and ventricular function after


APPENDIX. CONFERENCE DISCUSSION

Dr W. Gaynor (Philadelphia, PA, USA): We would agree with you that patient factors, particularly such things as prematurity, low birth weight, extracardiac anomalies, and especially pulmonary venous obstruction, probably have a greater effect on mortality than does the actual shunt type, and we still need to develop ways to try to overcome these. One way to overcome them is to try to individualise how we care for the patients and not say we have a strategy of always using one type of shunt but to try to pick patients who may do better.

Unfortunately, while the results are excellent, there are problems with this cohort. It is a relatively small series, particularly the number of patients with RPV-PA conduits. They were not performed in contemporaneous fashion, so there could be other changes that were not measured, in ICU management or preoperative management, despite the fact that you think you had a standardised rate of care. Obviously, there is some potential for selection bias because it is not a randomised trial. So I think we have to take some of the conclusions about the shunt types with a grain of salt.

Our data before our SVR trial began suggested that, again in non-randomised, historical controls, the RV-PA conduit might be associated with lower interstage mortality, lower early mortality, but there were significant concerns over the long-term ventricular function. Similar data has been presented for the Medical University of South Carolina. In the SVR trial what we could see with a prospective randomised trial over 15 institutions, as you said, with over 500 patients, is that there does appear to be an early survival benefit for the RV-PA conduit with concerning findings over the year, as you pointed out, that the curves begin to come together, and there may be an incidence of late ventricular dysfunction. So hopefully we are continuing the analysis of that cohort, and hopefully we can begin to find and tease out some subgroups, as you suggested in your data, in where, maybe one shunt is better than others in certain situations.

And when I responded to Viktor earlier, in my practice now I try to pick the kids who I think are at high risk for interstage mortality or an early event, particularly those children with pulmonary venous obstruction, intact atrial septum, or a very, very tiny aorta. But we know they may have instability and are at risk for interstage mortality, and we do the RV-PA conduit with those, thinking if we can get them through that period, then at least they are alive to deal with the later problems of ventricular function. And then use a B-T shunt for the other patients who we think - we do not know, you have to make a clinical decision sometimes based on not a lot of data - are at lower risk.

So, my major question for you. You have shown that you have basically gone away from the RV-PA conduit. If you have a full-term, 3 kg baby who presents with basically intact atrial septum, pulmonary venous congestion, who requires a stent or an emergency septectomy, has a 1-1/2 mm aorta, I know what I would choose in that situation, but what would you choose at that point for the patient who requires a stent or an emergency septectomy, has a 1-1/2 mm aorta, I know what I would choose in that situation, but what would you choose at your institution based on your data and the other data in the literature? Dr Photiadis: You are absolutely right, the major limitation of this study is its retrospective nature. But as we could prove, randomisation can also be faulty in terms of detecting high-risk patients, as you said. And so we think that using the comprehensive Aristotle score gives you a good picture of how high the preoperative risk of a patient is, and thus is also good tool when consulting with the patient’s parents of what the outcome may be, as I said, with low risk having an excellent outcome.

With the question you asked regarding patients with highly restrictive pulmonary venous return, such as an intact, it is difficult. I think, as you said, the number of our Sano patients with this special entity is small. One may argue that with the improved diastolic pressure that has been proven for the RV-PA conduit, there is better blood flow in situations where the pulmonary vascular resistance suddenly increases as we commonly see after repair of obstructive pulmonary venous return, and thus may have survival benefit. However, our data cannot prove this, and a larger series is needed to look at this issue. We are undertaking an MRI study which may give us more information on the status of the right ventricular function three or more years after a Norwood operation.

Dr Gaynor: But back to the question, your data does suggest that there is a survival advantage with the RV-PA conduit and obstructed pulmonary venous return, and you have to make a clinical decision.
Re: Does the shunt type determine mid-term outcome after Norwood operation?

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There is always a debate as to which shunt is better in a Norwood procedure [1-4]. Some say the modified Blalock-Taussig (MBT) shunt is better than the right ventricle-pulmonary artery (RV-PA) shunt, while others say the RV-PA shunt is better than the MBT shunt. Photiadis et al. [1] compared their experience of MBT shunts with that of RV-PA shunts in 109 patients. They concluded that there was no significant difference in the survival rates between the two shunt types. However, the incidence of shunt-related interventions was significantly increased with the RV-PA (Sano) shunt group. Ohye et al. [2] reported the results of the first multicentre, randomized trial of the Norwood procedure with comparison of an RV-PA shunt and MBT shunt. They summarized that the 12-month transplantation-free survival was higher with the use of an RV-PA shunt than with the use of an MBT shunt. However, the RV-PA shunt was associated with a higher rate of unintended cardiovascular interventions and complications during the first 12 months after randomization. Their conclusion was that there was no significant difference between the two groups with respect to transplantation-free survival beyond 12 months.

The Norwood procedure using an MBT shunt creates pulmonary atresia from aortic atresia, while the idea of an RV-PA shunt is to create tetralogy of Fallot from aortic atresia. Postoperative management after the implantation of an MBT shunt in patients with pulmonary atresia was not easy because it is important to keep a balance between systemic and pulmonary circuits. Babies with an MBT shunt sometimes collapse suddenly in the ward or at home. However, almost no babies experience a sudden collapse in tetralogy of Fallot. While many surgeons and many centres report almost no or minimal mortality in patients with MBT shunt, and that it is easy to keep a balance between systemic and pulmonary circuits, the discharge mortalities in neonates after MBT shunts were 7.2 and 10.6%, in the Society for Thoracic Surgeons database [5] and in the European Society for Cardio-Thoracic Surgery database (http://www.eactscongenitaldb.org/index.php?LANG=en&level=1&struct=14), respectively. These data clearly show that the MBT shunt has a high mortality, and this is the reality of the situation.

Creating a tetralogy of Fallot instead of pulmonary atresia was the simple idea behind the RV-PA shunt when I started using the RV-PA shunt to treat the hypoplastic left heart syndrome. This was because postoperative management after the implantation of a MBT shunt was not easy.

We all worry about the adverse effects on RV function after a ventriculotomy, however, there are many papers that have demonstrated no adverse effects in the use of a RV-PA shunt [3, 6]. The site and size of the ventriculotomy as well as the size of the shunt are important. Initially, we created an RV hole by...