ORIGINAL ARTICLE

Norwood procedure for hypoplastic left heart syndrome: BT shunt or RV-PA conduit?

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Background: The Norwood procedure is the first stage palliative procedure for hypoplastic left heart syndrome (HLHS). Traditionally the pulmonary circulation has been supplied via a modified Blalock Taussig (BT) shunt but a recent modification, adopted in some UK centres, substitutes a conduit between right ventricle and pulmonary arteries (RV-PA conduit). It is argued that this will result in a more favourable balance between pulmonary and systemic circulations.

Aim: To compare the early postoperative haemodynamic profile between patients undergoing a BT shunt or an RV-PA conduit.

Methods: Retrospective review in a tertiary referral PICU of 51 children with HLHS undergoing the Norwood procedure with either a BT shunt (Group 1; n = 23) or an RV-PA conduit (Group 2; n = 28). Data items were extracted at 10 set time points in the initial 96 h, postoperatively.

Results: Diastolic BP was significantly lower in Group 1 ($p < 0.001$) with a trend towards a higher systolic BP and no difference in mean BP. No between-group differences were found in markers of pulmonary blood flow (PaO_2 , PaCO_2 , $\text{PaO}_2/\text{FiO}_2$ ratio), or in markers of systemic blood flow (blood lactate, oxygen extraction ratio), or in estimated ratio of pulmonary:systemic blood flow ($\text{Qp}:\text{Qs}$). Despite lower diastolic blood pressure in Group 1 renal and hepatic function did not differ over five post-operative days between groups.

Conclusions: With the exception of a higher diastolic blood pressure in the RV-PA conduit group, we found no difference in the early haemodynamic profile between patients undergoing an RV-PA conduit or a BT shunt.

Hypoplastic Left Heart Syndrome (HLHS) describes a group of cardiac anomalies characterised by underdevelopment of the left side of the heart with severe hypoplasia of the ascending aorta. In 1983 Norwood first described a series of infants who underwent successful surgical palliation using a staged approach.¹ Cardiac centres still quote mortality rates between 5 and 40% following the Norwood procedure.²

As the native main pulmonary artery is used in the construction of a neo-aorta during the Norwood procedure an alternative source of pulmonary blood flow becomes necessary. Traditionally, the pulmonary circulation following the Norwood procedure has been supplied by a modified Blalock Taussig (BT) shunt, with a GORE-TEX tube connecting the subclavian artery and the pulmonary artery. BT shunt flow to the lungs occurs throughout the cardiac cycle, resulting in potential 'steal' from the systemic circulation during diastole. Furthermore coronary perfusion following the Norwood procedure is usually dependent on retrograde flow down a diminutive ascending aorta and can potentially be compromised by diastolic run-off and a low diastolic pressure.

In 2003 Sano and colleagues reported a modification of the Norwood procedure by placing a larger GORE-TEX graft between the right ventricle and the pulmonary arteries.³ This right ventricle to pulmonary arteries (RV to PA) conduit has been proposed as a better alternative to a modified BT shunt and preliminary data from one or two centres has suggested a better outcome.^{4–6} It is hypothesized that the conduit will provide a more stable post-operative balance between pulmonary and systemic circulations with a lower pulmonary:systemic blood flow ratio ($\text{Qp}:\text{Qs}$). Improved coronary perfusion as a result of less diastolic run-off may improve myocardial performance. On the other hand creation of an RV-PA conduit may require a longer aortic cross-clamp time and involves an incision into the right ventricular muscle, a ventriculotomy,

both of which may adversely affect myocardial function in the post-operative period.

We undertook this study to compare early postoperative haemodynamic data and markers of pulmonary and systemic blood flow between patients undergoing a Norwood procedure with a modified BT shunt or an RV-PA conduit.

METHODS

At Birmingham Children's Hospital, the first Sano modification of the Norwood procedure was performed in March 2002. We retrospectively reviewed the case-notes of 51 consecutive cases undergoing a Norwood procedure between May 2001 and June 2003. During this interval 23 cases underwent a Norwood procedure with modified BT shunt (Group 1) and 28 underwent a Norwood procedure with RV to PA conduit (Group 2).

Informed consent was obtained from the parents of each patient about all details of the operative procedure. The study was approved by Birmingham Children's Hospital Research Department.

Data included operative variables, haemodynamic parameters, inotropic and vasodilator support (table 1). We examined markers of pulmonary blood flow, gas exchange and markers of systemic blood flow (table 1). $\text{Qp}:\text{Qs}$ at each timepoint was estimated using the formula $(\text{SaO}_2 - \text{SvO}_2)/(\text{SpvO}_2 - \text{SpaO}_2)$. A value of 96% was assumed for SpvO_2 . SvO_2 was measured from a central venous line in the superior vena

Abbreviations: BT, Blalock Taussig; CPB, cardiopulmonary bypass; CVP, central venous pressure; DHCA, deep hypothermic circulatory arrest; ETCO_2 , end-tidal carbon dioxide; HLHS, hypoplastic left heart syndrome; OE, oxygen extraction; PaO_2 , arterial partial pressure of oxygen; PaCO_2 , arterial partial pressure of carbon dioxide; PRVC, Pressure Regulated Volume Control; $\text{Qp}:\text{Qs}$, pulmonary:systemic blood flow; RV-PA, right ventricle to pulmonary artery; SaO_2 , arterial oxygen saturation; SvO_2 , venous oxygen saturation

Table 1 Summary of the data variables collected for the study

Patient demographics	Gestational age, chronological age, sex, birth weight, antenatal diagnosis
Operative variables	Duration of CPB, aortic cross clamp and DHCA, minimum core temperature
Haemodynamic parameters and inotropic/vasodilator support	Heart rate, systolic BP, diastolic BP, mean BP, CVP
Pulmonary blood flow and gas exchange	SaO ₂ , PaO ₂ , FiO ₂ , PaCO ₂ , ((PaCO ₂ – ETCO ₂)/PaCO ₂)
Systemic blood flow and oxygen delivery	Blood lactate, SvO ₂ , OE ratio, renal and liver function
Pulmonary/systemic blood flow ratio	Qp:Qs

BP, blood pressure; CPB, cardiopulmonary bypass; CVP, central venous pressure; DHCA, deep hypothermic circulatory arrest; ETCO₂, end-tidal carbon dioxide; OE ratio, oxygen extraction ratio; PaO₂, arterial partial pressure of oxygen; PaCO₂, arterial partial pressure of carbon dioxide; Qp:Qs, pulmonary to systemic blood flow ratio; SaO₂, arterial oxygen saturation; SvO₂, venous oxygen saturation.

cava or inferior vena cava. Following the Norwood procedure SpaO₂ is identical to SaO₂. Parameters were recorded at 10 set time points after return to the intensive care unit (0, 4, 8, 12, 24, 36, 48, 60, 72, 96 h).

Diagnosis of HLHS was limited to patients with mitral stenosis or atresia, aortic stenosis or atresia, with diminutive left ventricle and systemic right ventricular circulation. Operations were performed using deep hypothermic cardiopulmonary bypass with periods of circulatory arrest for arch reconstruction, which was performed using a pulmonary homograft patch in both groups. In Group 1, the pulmonary anastomosis of a modified Blalock-Taussig shunt was completed with a polytetrafluoroethylene tube (GORE-TEX), to the upper border of the right pulmonary artery. The main PA was then divided at the level of the bifurcation and the defect in the PAs repaired with a pulmonary homograft patch. A 3.5 mm shunt was used in patients ≥ 2.5 kg ($n = 21$) and a 3 mm shunt was used if < 2.5 kg ($n = 2$), proximally anastomosed to the innominate artery. In Group 2 the main PA was divided as above and the defect patched with pulmonary homograft. An opening was then cut into the patch to receive the GORE-TEX® tube. The proximal end of the conduit was anastomosed to a ventriculotomy in the distal infundibulum of the right ventricle. A 5 mm shunt was used in all patients ≥ 2.0 kg ($n = 26$) and a 4 mm shunt if < 2.0 kg ($n = 2$).

Antegrade cerebral perfusion was introduced during the period of this study and used when the head-and neck vessels were of suitable size to accommodate the arterial cannula during arch reconstruction. The introduction of antegrade cerebral perfusion was made independently of the conversion to a modified Norwood procedure with a right ventricle-pulmonary artery conduit.

All patients were ventilated in Pressure Regulated Volume Control (PRVC) mode (Servo 300), and ventilation was adjusted to achieve a target PaCO₂ of 5–6 kPa and arterial oxygen saturation 75–85%. No patient received supplemental inspired CO₂ or a hypoxic gas mixture. The chest was left open electively in all cases, using a fenestrated GORE-TEX patch to the skin, and was closed when the clinical condition was felt to be stable.

STATISTICAL ANALYSIS

Data are represented as mean (standard deviation) or median (range). Patient characteristics, operative variables and outcome variables were compared between the groups using a Student *t* test, Mann Whitney U test or Fisher's exact test, as appropriate. A Repeated Measures General Linear Model analysis was used to evaluate the post-operative variables for within-group effects over time and between-group effects (SPSS 12.0 for Windows). Differences were considered significant at a *p* value < 0.05 .

RESULTS

Group characteristics

The two groups did not differ significantly with respect to age at procedure (Group 1: 5 days (1–18) vs Group 2: 5 days (2–15); $p = 0.57$), gender (Group 1: 30% female vs Group 2: 39% female; $p = 0.56$) or antenatal diagnosis (Group 1: 65% antenatal vs Group 2: 57% antenatal; $p = 0.77$). Birth weight was lower in Group 2 (Group 1: 3.3 (0.5) kg vs Group 2: 2.9 (0.7) kg; $p = 0.04$).

Operative variables

Significant differences were found in the duration of circulatory arrest, aortic cross clamp and cardiopulmonary bypass between the groups. In Group 1 the circulatory arrest time was longer (46 mins (11–70) vs 23 mins (6–65) $p = 0.003$), but aortic cross clamp (49 mins (38–96) vs 61 mins (12–76); $p = 0.001$) and cardiopulmonary bypass times (69 mins (45–322) vs 106 mins (45–138); $p = 0.001$) were shorter than Group 2.

Haemodynamics

Over the first 96 hours post-operatively the heart rate (fig 1) and CVP were not significantly different between groups. Diastolic BP was significantly lower in Group 1 ($p < 0.001$) with a trend towards a higher systolic BP ($p = 0.06$) but no difference in mean BP ($p = 0.12$) (fig 2). The calculated pulse pressure was significantly higher in Group 1 ($p < 0.001$). Significant within group changes with time were evident in both groups for diastolic BP ($p = 0.003$), systolic BP ($p < 0.001$), heart rate ($p < 0.001$), and CVP ($p = 0.03$).

Inotropic/vasodilator therapy

No difference was found in mean adrenaline or dobutamine dose between the groups. Sodium nitroprusside was used less often in Group 2 (62% vs 95% of cases; $p = 0.01$) with a trend towards a greater use of milrinone (33% vs 14% of cases; $p = 0.18$).

Figure 1 Heart rate profile over the first 96 h in intensive care for the Blalock Taussig (BT) and Sano RV-PA groups.

Figure 2 (A) Systolic blood pressure (BP) and (B) diastolic BP profile over the first 96 h in intensive care for the Blalock Taussig (BT) and Sano RV-PA groups.

Gas exchange and markers of pulmonary blood flow

No between-group differences were found in PaO_2 , PaCO_2 , SaO_2 , FiO_2 or $\text{PaO}_2/\text{FiO}_2$ ratio. There were no differences in $(\text{PaCO}_2 - \text{ETCO}_2)/\text{PaCO}_2$, a marker of dead-space ventilation that can reflect alterations in pulmonary blood flow. Within-group changes with time were evident for FiO_2 and $\text{PaO}_2/\text{FiO}_2$ ratio ($p < 0.001$) (fig 3), but not for PaO_2 .

Markers of systemic blood flow

No between-group differences were found in lactate profile over 96 hours ($p = 0.81$), though a marked reduction with time was evident within each group ($p < 0.001$) (fig 4).

Missing data for SvO_2 prevented us from performing a repeated measures analysis. Instead we compared available data points between the groups at four post-operative time points (4 ($N = 25$), 8 ($N = 22$), 12 ($N = 23$) and 24 hrs ($N = 22$)) using a Mann Whitney U test. No significant between-group difference was found at any of these time points. A similar analysis was undertaken for oxygen extraction ratio and estimated Qp:Qs , with no significant between-group differences detected.

End organ function

There was no difference in renal or hepatic end-organ function over 5 post-operative days between the groups (table 2). No

Figure 3 Changes in $\text{PaO}_2/\text{FiO}_2$ ratio (kPa) over the first 96 hours in intensive care for the Blalock Taussig (BT) and Sano RV-PA groups.

difference was found in the proportion of cases managed with peritoneal dialysis (Group 1: 25% vs Group 2: 30%; $p = 1.0$).

Outcomes

No difference was found in survival to hospital discharge (Group 1: 78% vs Group 2: 82%; $p = 0.9$) or in the proportion of cases requiring urgent chest re-exploration (Group 1: 23% vs Group 2: 19%; $p = 1.0$), requiring cardiopulmonary resuscitation (Group 1: 18% vs Group 2: 21%; $p = 0.74$), time to chest closure (Group 1: 2 days (1–7) vs Group 2: 3 days (2–26); $p = 0.13$), PICU length of stay (Group 1: 7 days (4–69) vs Group 2: 9 days (5–28); $p = 0.09$), or hospital length of stay (Group 1: 18 days (9–81) vs Group 2: 18 days (11–38); $p = 0.67$).

DISCUSSION

Over the past 20 years there have been dramatic advances in the surgical treatment of HLHS, which is the commonest cardiac cause of neonatal death.^{7–9} Studies have looked at modifying surgical technique and manipulating post-operative cardiovascular physiology in order to improve cardiac output and reduce mortality.^{10–15}

In 2003, Sano *et al* published the results of 19 consecutive cases of infants undergoing his modification of the Norwood procedure using an RV to PA conduit.³ It has been hypothesised that the RV-PA conduit will result in greater postoperative stability, improved myocardial performance, a lower pulmonary to systemic blood flow ratio and better systemic perfusion.^{3 4 17} To date, comparative studies have emanated from a very limited number of centres, and have largely suggested benefits of the RV-PA conduit.^{4–6 17} A number of studies have concentrated on late haemodynamic profile, at the time of cardiac catheterisation before undertaking the second stage procedure, and have reported a lower Qp:Qs and better pulmonary artery growth in the RV-PA conduit group.^{17–19}

This study compares in greater detail than previous studies the early postoperative haemodynamic profile. It confirms the findings of a lower diastolic blood pressure in the BT shunt group, which has been a consistent finding in previous studies,^{4–6 17} but no difference in mean blood pressure as a result of a higher systolic pressure. Although concern has been expressed that a lower diastolic BP together with reduced diastolic flow in the descending aorta may significantly compromise flow to the kidneys, liver and gut,⁶ no difference in clinical sequelae has previously been reported. We too found no evidence to support an important difference in renal and hepatic blood flow as a consequence of the lower diastolic blood pressure.

In this study we were not able to detect differences in markers of pulmonary or systemic blood flow between the groups. This contrasts with Pizarro *et al*,⁶ who reported a higher PaO_2 in the early post-operative period in the BT shunt group, suggesting a higher pulmonary blood flow, as well as a requirement for more frequent ventilator manipulation to balance Qp:Qs . These differences are likely to be explained by the use of a larger BT shunt (4 mm) by Pizarro and colleagues compared to a 3.5 mm shunt in this study. In contrast we found no difference in ventilator management or in gas exchange between the groups.

Aortic cross clamp time was significantly longer in our patients undergoing a Norwood repair with RV-PA conduit, which, together with a ventriculotomy, could adversely affect myocardial function. Mair and colleagues¹⁷ found significantly higher Troponin levels in the RV-PA group, supporting greater myocardial injury.

Interpretation of the shorter circulatory arrest time and longer CPB time in the RV-PA group is confounded by the introduction of antegrade cerebral perfusion during this study

Figure 4 Blood lactate over the first 96 hours in intensive care for the Blalock Taussig (BT) and Sano RV-PA groups.

and the greater use of RV-PA repair after this time-point. Antegrade cerebral perfusion was not specifically targeted at either group but this technique was used in a greater proportion of Group 2 patients.

There are limitations to a study of this sort that compares a new procedure with an earlier control group. The published comparative studies to date have all involved comparison of a cohort of patients undergoing an RV to PA conduit with a historical cohort undergoing a BT shunt. Inevitably there is an evolution in surgical technique associated with any new procedure or practice,²⁰ such that the findings of this study might have been different if a later cohort of RV to PA conduit patients had been studied. In addition selection of cases may result in an uneven balance of complexity and bias the results in one direction. Changes in intensive care management may also impact on the results. In this study we found evidence of a move away from sodium nitroprusside towards greater use of milrinone, reflecting greater use of milrinone following publication of the PRIMACORP study.²¹

Current surgical practice across the UK is mixed with some centres adopting the RV-PA conduit whilst others continue with the BT shunt. A prospective randomised controlled trial comparing the two techniques is needed to determine whether important clinical outcomes are improved with the RV-PA conduit modification of the Norwood procedure. We must await with interest the results of an ongoing randomised trial in North America.

CONCLUSIONS

With the exception of a higher diastolic blood pressure in the RV-PA conduit group, we found no difference in the early haemodynamic profile, markers of pulmonary blood flow,

What is already known on this topic

- Hypoplastic Left Heart Syndrome is the commonest cardiac cause of neonatal death.
- Surgical treatment of HLHS includes staged Norwood palliation traditionally with a modified BT shunt.
- Postoperative pulmonary to systemic blood flow balance is important in maintaining adequate cardiac output and systemic perfusion.

What this study adds

- With the exception of a higher diastolic blood pressure in the RV-PA conduit group, no differences in early haemodynamic profile, markers of pulmonary blood flow, markers of systemic oxygen delivery or end organ function were found.
- A prospective randomised trial is needed to determine whether important clinical outcomes are improved with an RV-PA conduit modification.

markers of systemic oxygen delivery or end-organ function in patients undergoing an RV-PA conduit rather than a BT shunt.

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Competing interests: None declared.

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Table 2 Markers of renal and liver function before and after surgery for the Blalock Taussig (BT) and Sano RV-PA groups

		Pre-op	D1	D2	D3	D4	D5
Creatinine ($\mu\text{mol/l}$)	BT shunt	86.9 (31.9)	80.5 (18.5)	93.1 (22.1)	107.6 (35.8)	111.3 (55.6)	104.6 (72.4)
	RV-PA	76.8 (20.1)	80.0 (18.7)	95.3 (28.1)	106.5 (32.1)	102.8 (35.4)	87.8 (31.8)
Urine output (ml/kg/h)	BT shunt	—	2.8 (1.5)	3.0 (1.8)	3.0 (1.6)	2.9 (1.2)	3.7 (1.2)
	RV-PA	—	2.5 (1.7)	3.2 (1.7)	3.6 (1.7)	3.7 (1.7)	4.0 (1.4)
Fruzemide (mg/kg/day)	BT shunt	—	3.0 (0-24)	2.0 (0-24)	2.6 (0-24)	2.2 (0-17)	2.0 (0-3)
	RV-PA	—	2.7 (0-20)	3.0 (0-30)	2.0 0-24()	1.2 (0-8)	1.3 (0-3)
ALT (units)	BT shunt	51 (15-448)	27 (18-124)	35 (15-142)	54 (16-109)	66 (21-141)	62 (25-108)
	RV-PA	26 (16-126)	23 (7-96)	24 (11-259)	25 (13-261)	32 (10-293)	33 (16-294)
Bilirubin (units)	BT shunt	112 (40-284)	105 (40-1555)	117 (35-248)	131 (31-268)	84 (17-269)	57 (15-251)
	RV-PA	114 (38-248)	99 (23-219)	106 (23-245)	91 (24-278)	60 (13-282)	36 (16-255)

Renal function is described by changes in serum creatinine, urine output and requirement for diuretic over the first five post-operative days. Liver function is described by changes in serum bilirubin and alanine transaminase (ALT). Data are mean (SD) or median (range).

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IMAGES IN NEONATAL MEDICINE.....

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An unusual complication of phototherapy

Figure 1 Mosquito net enhanced phototherapy.

Having started work in warmer climes recently, I had prepared myself to face the unexpected. While doing a ward round, I came up to a baby with jaundice who was meant to be under phototherapy overnight.

“So how long was he under the lights?” I questioned, somewhat perplexed by his increasing level of jaundice.

“Well, he was only really under for about an hour” was the ashamed response.

“What happened, I thought I said...”

“He was getting bitten by the insects and was screaming all night!”

It turned out that the insects were being attracted to the blue lights and found our helpless victim lying underneath. And so began the new “mosquito net enhanced phototherapy”.

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