Outcome of the Norwood procedure in the setting of transposition of the great arteries and functional single left ventricle☆

Attilio A. Lotto, Riad Hosein, Timothy J. Jones, David J. Barron *, William J. Brawn

Birmingham Children’s Hospital, NHS Foundation Trust, Birmingham, UK

Received 13 December 2007; received in revised form 6 August 2008; accepted 6 September 2008; Available online 8 November 2008

Abstract

Objective: To assess the surgical results of the Norwood procedure and subsequent clinical outcome in the setting of transposition of the great arteries (TGA) with a dominant morphologic left ventricle. Methods: Among 486 patients who underwent the Norwood procedure from 1988 to 2007 at our institution, there were 37 patients with TGA and left ventricular dependant circulation with the following associated lesions: double inlet left ventricle (DILV) (n = 24), tricuspid atresia (n = 9), ventricular septal defect (VSD) with hypoplastic right ventricle (RV) (n = 4). Outcomes for all three-staged procedure were compared with the overall Norwood group. Results: Early mortality was 21.6% (8/37) compared to 26.7% (120/449) in the overall Norwood group (p = ns). There was only one subsequent death giving a 5- and 10-year actuarial survival of 72.8 ± 7.4% compared to 55.3 ± 2.6% and 52 ± 2.9% at 5 and 10 years for the overall series (p = 0.06). Median follow-up was 4.7 (0.7–10.2) years. Eighteen patients underwent stage III completion at 3.9 ± 1.5 years from the second stage with no mortality. Preoperative mean pulmonary artery (PA) pressure and transpulmonary gradient were respectively 11.6 ± 3.4 and 5.2 ± 3.3 mmHg. All patients had good left ventricle (LV) function at time of stage III. All patients except one are currently in NYHA I. One patient (with DILV) had congenital heart block and required a pacemaker. There was no postoperative heart block. The systemic outflow was unobstructed in all patients and no patient required any additional intracardiac procedure. Conclusions: The Norwood procedure provides good palliation in this subgroup of patients and avoids the need for subsequent intracardiac operations, maintaining unobstructed systemic outflow tract and avoiding the risk of postoperative heart block.

Keywords: Norwood procedure; Transposition of the great arteries; Double inlet left ventricle; Tricuspid atresia

1. Introduction

In hearts with transposition of the great arteries (TGA) and a functional single left ventricle, the aorta is supported by a rudimentary right ventricle and a ventricular septal defect (VSD) [1]. The VSD is positioned between the muscular apical trabecular septum and the muscular outlet septum with the potential for becoming restrictive resulting in progressive subaortic obstruction [1,2]. This arrangement can be found in hearts with double inlet left ventricle (DILV), but also in hearts with absent right or left atrioventricular connection where the other atrium is connected to a dominant left ventricle [1]. Moreover, aortic coarctation, with or without arch hypoplasia, is a recognised associated anomaly in a high proportion of patients with complex cardiac lesions [3].

Surgical management of this difficult group of patients is not uniformly agreed. Patients without any evidence of subaortic obstruction can be simply managed with a pulmonary artery (PA) band with or without arch repair as required [4–8]. However, even with the arch repaired, the risk of a PA band alone in these patients is the progressive development of subaortic obstruction in the future with an unpredictable time course [9–12].

This study has excluded patients without any evidence of systemic outflow obstruction and is specifically concerned with patients who required both aortic arch repair and in whom the aorta arose from a muscularised right ventricular chamber distant from the left ventricle (LV).

Enlargement of the VSD through the aortic valve has been suggested, but this can be technically difficult and postoperative heart block due to damage of the conduction system is a significant complication [8,13,14].

We have adopted the concept of creating an unobstructed outflow by mean of a Norwood type of procedure [12,15,16] as an alternative approach. This potentially adds to the complexity of the initial procedure and may expose patients to a higher operative risk [15].

The aim of this study was to assess the mortality of the Norwood type approach in patients with TGA and single left ventricle. Furthermore we examined the surgical and clinical
outcome of the subsequent stages leading to the Fontan circulation with particular attention on the natural history of the systemic outflow tract.

2. Materials and methods

2.1. Patient population

Between January 1988 and January 2007, 486 patients underwent a stage I Norwood type of procedure at the Birmingham Children’s Hospital NHS Trust, UK. Among them we identified 37 patients (7.6%) with TGA and a single functional ventricle of left morphology and a systemic outflow obstruction (group I).

As outlined above, patients with unrestricted systemic outflow and normal aortic arch dimensions were excluded from this study.

We compared the preoperative, intraoperative and postoperative findings of this group of patients with the results of the remaining 449 patients (group II) who underwent the Norwood procedure during the same time period.

The study was a retrospective review and was registered with the Birmingham Children’s Hospital R&D department; ethical approval was applied for through the COREC (Central Office for Research Ethics Committees) but was waived after ethical approval was applied for through the CRREC (Central Research Ethics Committee) of the Birmingham Children’s Hospital NHS Trust, UK. Among them we recruited 37 patients (7.6%) with TGA and a single functional ventricle of left morphology and a systemic outflow obstruction (group I).

As outlined above, patients with unrestricted systemic outflow and normal aortic arch dimensions were excluded from this study.

We compared the preoperative, intraoperative and postoperative findings of this group of patients with the results of the remaining 449 patients (group II) who underwent the Norwood procedure during the same time period.

The study was a retrospective review and was registered with the Birmingham Children’s Hospital R&D department; ethical approval was applied for through the COREC (Central Office for Research Ethics Committees) but was waived after consultation in respect of being retrospective with anonymous data. No patients were refused surgery during the period of the study.

2.2. Morphology

The anatomical morphology of group I is shown in Table 1. All patients had obstruction either at the level of the systemic outflow and/or at level of the aorta. Anomalies related to the aorta were: interrupted aortic arch in 2 patients and aortic coarctation in 4 patients; hypoplasia of the ascending aorta, defined by a diameter less than the patient’s weight in kg plus 1, was present in 14 patients; 5 patients had a hypoplastic aortic arch, 4 of whom with associated aortic coarctation; 6 patients had a long tubular narrow aortic arch with borderline dimensions.

Obstructions to the systemic outflow were: one patient had restrictive VSD detected at birth, five patients developed a restricted VSD after the first few weeks of life, and significant subaortic stenosis was present in six patients.

Thirty-one patients presented at neonatal age and 22 patients had a duct dependent circulation on prostaglandin E₂ at time of operation.

These and other associated anomalies are summarised in Table 2.

<table>
<thead>
<tr>
<th>Table 1: Anatomical characteristic of 37 patients with functional left ventricle and transposition of the great arteries.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anatomical diagnosis</td>
</tr>
<tr>
<td>Double inlet left ventricle</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
</tr>
<tr>
<td>Hypoplastic right ventricle and VSD</td>
</tr>
</tbody>
</table>

VSD: ventricular septal defect.

<table>
<thead>
<tr>
<th>Table 2: Associated anomalies.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Associated anomalies</td>
</tr>
<tr>
<td>Interrupted aortic arch</td>
</tr>
<tr>
<td>Aortic coarctation</td>
</tr>
<tr>
<td>Hypoplastic ascending aorta</td>
</tr>
<tr>
<td>Hypoplastic-narrow aortic arch</td>
</tr>
<tr>
<td>Restrictive VSD</td>
</tr>
<tr>
<td>Subaortic stenosis</td>
</tr>
<tr>
<td>Congenital heart block</td>
</tr>
<tr>
<td>Patients on PGE₂ at time of surgery</td>
</tr>
</tbody>
</table>

VSD: ventricular septal defect; PGE₂: prostaglandin E₂. Hypoplastic ascending aorta and arch defined by a diameter less than the patient’s body weight in kg plus 1.

2.3. Surgical management

2.3.1. Stage 1

The surgical technique has been described elsewhere [15,17] and is summarised below. Deep hypothermia to 18 °C with periods of circulatory arrest was employed for all repairs. The surgical technique used to reconstruct the systemic outflow tract depended upon the anatomy as described below:

- In the setting of a normal sized ascending aorta and arch, a direct DKS anastomosis was made between the main PA and the underside of the proximal arch and ascending aorta without augmentation of the aortic arch.

- In patients with coarctation or arch hypoplasia the coarctation was resected followed by an end-to-end anastomosis with patch augmentation of the arch and ascending aorta using pulmonary artery homograft. A separate incision was then cut into this patch to receive the main PA [17]. In our practice this is the technique used for the Norwood procedure.

- If the vessels were in complete antero-posterior relationship then both the ascending aorta and main PA were transected at the same level and the facing edges sutured together to create a ‘double-barreled’ outflow to the heart. The arch was then augmented with a homograft patch and anastomosed to this double-barreled outflow.

- In all patients, regardless of the technique used for outflow tract reconstruction, a 3–3.5 mm Gore-Tex (Gore-Tex, WL Gore & Associates UK Ltd., Livingston, Scotland) shunt was placed between the proximal brachiocephalic artery and the proximal right PA according to patient weight (3 mm for patients <2.5 kg and 3.5 mm for patients ≥2.5 kg). An atrial septectomy was performed in all patients and selective cerebral perfusion was used during arch reconstruction since 2002.

2.3.2. Stage 2

The surgical technique for the second stage consisted of construction of bidirectional superior cavopulmonary anastomosis (Bidirectional Glenn shunt) performed during a period of deep hypothermic circulatory arrest at 18 °C. Any narrowing in the central pulmonary arteries was addressed at the time with a separate patch of pulmonary homograft.

2.3.3. Stage 3

During the study period, the surgical technique employed for completion of the Fontan circulation evolved as
previously described [18]. From 1988 to 1995, the Fontan procedure was performed using an atrio-pulmonary connection. In 1995 the unit adopted the total cavopulmonary connection (TCPC) technique. This initially involved a lateral atrial tunnel. However, since 1998 the Fontan procedure has been performed using an extracardiac conduit TCPC with the interposition of a Gore-Tex tube (W.L. Gore & Associates (UK) Ltd., Livingston, Scotland) between the IVC and the right PA. Fenestration of the circuit was initially according to surgeons’ preference but has become routine practice since 2002.

2.4. Follow-up

Patients’ records were reviewed retrospectively. Clinical outcomes were examined by reviewing the most recent outpatient follow-up or by contacting the local cardiologist to provide current clinical status. Need for further intervention, in particular need for intracardiac reoperations and relief of outflow obstruction, were recorded.

2.5. Statistical analysis

Data were examined by means of analysis of variance with a commercial statistical software package (SPSS for Windows, version 12; SPSS Inc, Chicago, Ill). Continuous variables are expressed as mean ± standard deviation (SD) and comparative univariable analyses between group I and group II have been made using the t-test, Mann–Whitney U-test or Wilcoxon signed rank test. Binomial or ordinal data are expressed as percentage and comparative univariable analyses between group I and group II have been made using the χ² test, two-sided Fisher’s exact test or binomial logistic regression. Cumulative stage I mortality was considered as in-hospital plus interstage mortality; comparison between the two groups was performed in three different periods as follows: from January 1992 to December 1997; from January 1998 to December 2001; from January 2002 to January 2007. Actuarial survival of group I and group II was estimated by using the Kaplan–Meier product limit method. A probability value of p < 0.05 was taken to represent a statistically significant difference between groups.

3. Results

The in-hospital mortality for the Norwood stage I in group I was 16.2% (6/37 patients). When compared to the 26.7% (120/449 patients) mortality in group II, there was not a significant difference (p = 0.17).

Fig. 1 describes the results of the surgical stages for patients in group I.

Comparison of the preoperative characteristic between the two groups revealed that patients in group I were older at time of operation (Table 3). This reflected the fact that, unlike HLHS, the patients in group I were not all duct dependent lesions and so did not all present as early. Because of the different anatomical substrate between the two groups, the mean ascending aorta diameter was also significantly bigger in group I (Table 3).

The cardiopulmonary bypass, aortic cross-clamping and circulatory arrest times were comparable between the two groups (Table 3). Cerebral perfusion times during arch repair (instituted in 2002) were included in the total circulatory arrest times.

All patients in group I underwent a classic Norwood type of procedure with a shunt between the brachiocephalic artery and the right PA, as did 60% of the patients in group II. However, following the development of the ‘Sano’ modification [22] with a right ventricle to PA Gore-Tex shunt, this has become the routine procedure for hypoplastic left heart syndrome in this unit since 2002, accounting for 40% of the patients in group II (p < 0.0001).

Two patients (6.4%) of group I, both with TGA and TA, died in the interstage period while waiting for the stage II procedure. When compared to the 10.9% (36/329) of group II, the difference was not statistically significant (p = 0.4).

The cumulative mortality for the stage I Norwood procedure in the group I patients (in-hospital and interstage period) has decreased since 2002. In the period from 1992 to 1997 it was 25% (4/16 patients), from 1998 to 2002 it was 33% (3/10 patients) and from 2002 to the end of the study period it was 9% (1/11 patients). Nonetheless, there were no significant differences between the two groups when mortality was compared across the three eras (Table 4).

Fig. 1. Flow chart of the surgical pathway leading to the Fontan circulation in group I. DILV: double inlet left ventricle. TA: tricuspid atresia. Hypo pl. RV: hypoplastic right ventricle.
Of the initial 37 patients, all 29 survivors underwent the second stage procedure at a mean age of 7.1 ± 3.6 months and with an interstage time of 6.3 ± 3.0 months. Five patients needed PA augmentation (three patients of the left PA, one patient of the central PA and one patient of the right PA) at time of the procedure with a pulmonary homograft patch.

In-hospital mortality for group I was 3.4%, represented by one patient who developed postoperative sepsis, ARDS and died of multiorgan failure. Of the 293 survivors in group II, 20 are still waiting to undergo the Norwood stage II procedure. Of the remaining 273, 12 patients (4.0%) died in hospital following the Norwood stage II. The difference in mortality between the two groups was not significant (p = 0.9). Currently, 18 patients in group I have undergone the third stage with completion of the Fontan circulation. The mean interstage time was 3.8 ± 1.4 years from stage II and the mean patients’ age was 3.9 ± 1.5 years. All patients scheduled for surgery underwent a preoperative cardiac catheterisation that demonstrated a mean PA pressure of 11.6 ± 3.4 mmHg and a transpulmonary gradient of 5.2 ± 3.3 mmHg. One hundred and seven patients in group I had a successful Fontan circulation: of these patients underwent an unobstructed systemic outflow from early life, avoiding the deleterious effects of gradual outflow obstruction and subsequent myocardial hypertrophy. Subsequent progression to a Fontan circulation is associated with a low postoperative mortality and good long-term clinical outcome.

All patients in group I had a successful Fontan circulation: 2 patients had an atriovenous connection, 4 patients had a lateral tunnel and 12 patients had a total cavopulmonary connection (TCPC) with a Gore-Tex tube conduit (W.L. Gore & Associates (UK) Ltd., Livingston, Scotland). In 6 patients the Fontan circulation was not fenestrated and in 12 patients a fenestration was constructed. Mean postoperative intensive care stay was 1.07 ± 0.27 days, mean intercostal chest drains duration was for 10.1 ± 5.8 days. Total postoperative hospital stay was 14.7 ± 6.2 days.

Patients in group II had a mean postoperative intensive care stay of 2.2 ± 4 days, and the mean duration of intercostal tube drainage was 16.0 ± 4 days. Total postoperative hospital stay was 21.5 ± 15.7 days.

Mortality for this stage was zero for group I and 0.9% (1/107 patients) in group II and this difference was not statistically significant.

There are 10 patients in group I and 134 in group II awaiting the third stage procedure.

### 3.1. Follow-up

The follow-up of group I is 100% complete at a median time of 4.7 years (range 0.7—10.2 years). All patients except one are at present in NYHA class I with a mean functional class of 1.1 ± 0.2. Serial echocardiography performed in the postoperative period has demonstrated an unobstructed systemic outflow in all patients. None of the patients who survived the stage I Norwood needed intracardiac reintervention. One patient with congenital heart block needed a permanent pacemaker system.

The actuarial survival curves (Fig. 2) showed a trend towards a better survival in group I compared with group II at 5 years (group I 72.8 ± 7.4% vs group II 55.3 ± 2.6% p = ns) and at 10 years (group I 72.8 ± 7.4% vs group II 52 ± 2.9%, p = 0.06).

### 4. Discussion

The major finding of this study is that a Norwood procedure, when applied in patients with TGA and functional single left ventricle, carries an acceptable mortality comparable to other patients undergoing this procedure. Moreover, it offers an unobstructed systemic outflow from early life, avoiding the deleterious effects of gradual outflow obstruction and consequent myocardial hypertrophy. Subsequent progression to a Fontan circulation is associated with a low postoperative mortality and good long-term clinical outcome.

Importantly, none of the patients suffered postoperative heart block or needed reoperations for intracardiac obstruc-

---

### Table 3
Preoperative characteristics and operative results in group I and group II.

<table>
<thead>
<tr>
<th>Operative procedure</th>
<th>Group I TGA—single LV</th>
<th>Group II Norwood group</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient characteristics</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patient number</td>
<td>37 (70.2%)</td>
<td>449</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>26 (70.2%)</td>
<td>275 (60.1%)</td>
<td>ns</td>
</tr>
<tr>
<td>Mean age (days)</td>
<td>21.1 ± 35.5</td>
<td>8.3 ± 3.3</td>
<td>0.003</td>
</tr>
<tr>
<td>Mean weight (kg)</td>
<td>3.4 ± 0.6</td>
<td>3.1 ± 0.8</td>
<td>0.006</td>
</tr>
<tr>
<td>Mean asc ao (mm)</td>
<td>4.6 ± 1.1</td>
<td>3.3 ± 0.1</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Operative data</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean CPB time (min)</td>
<td>88.8 ± 39.2</td>
<td>89.7 ± 31.3</td>
<td>ns</td>
</tr>
<tr>
<td>Mean cross-clamp (min)</td>
<td>53.8 ± 22.9</td>
<td>55.2 ± 9.3</td>
<td>ns</td>
</tr>
<tr>
<td>Mean circ arrest (min)</td>
<td>42.9 ± 22.9</td>
<td>41.1 ± 22.7</td>
<td>ns</td>
</tr>
</tbody>
</table>

### Table 4
Mortality of group I vs group II in three different periods.

<table>
<thead>
<tr>
<th>Period</th>
<th>Group I</th>
<th>Group II</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>January 1992—January 1997</td>
<td>25.0% (4/16)</td>
<td>37.8% (42/111)</td>
<td>ns</td>
</tr>
<tr>
<td>January 1997—January 2002</td>
<td>33.3% (3/10)</td>
<td>40.8% (56/137)</td>
<td>ns</td>
</tr>
<tr>
<td>January 2002—January 2007</td>
<td>9.1% (1/11)</td>
<td>28.0% (51/182)</td>
<td>ns</td>
</tr>
</tbody>
</table>

* p: ns for intergroup and intragroups comparisons.
tions which have previously been highlighted as potential risks in this group of patients [2,9,10].

In patients with TGA and a single left ventricle, the VSD is at risk of becoming restrictive. This may happen due to a natural tendency for this particular type of muscular VSD to become smaller, as supported by Rao [9]. Progression to a restrictive VSD may also be accelerated by surgical palliation: pulmonary artery banding may lead to myocardial hypertrophy and hence VSD restriction with subaortic obstruction [2,10]. Freedom et al. reported in patients with single ventricle physiology and TGA initially palliated with pulmonary artery band a high incidence (84.4%) of developing subaortic obstruction [10].

In a series of 26 patients with DILV or TA and TGA, Jensen et al. reported their results with initial PA band with or without aortic arch reconstruction [6]. A significant subaortic obstruction, with a gradient equal or more than 10 mmHg between the ascending aorta and the left ventricle, developed in more than half of the patients [6]. Webber et al. reported their results in a similar group of patients, initially palliated with PAB and aortic arch repair [7]. Fifteen of the 16 survivors needed relief of subaortic obstruction after the initial palliation [7].

Aortic arch hypoplasia is an associated anomaly found in a high proportion of patients with complex cardiac lesions [3]. A left thoracotomy approach has been described, undertaking repair of aortic arch hypoplasia and simultaneous placement of a PA band. However, concerns have been raised about the capability to achieve an unobstructed aortic arch repair through a left thoracotomy, particularly in case of diffuse tubular arch hypoplasia [12,14]. Odim et al. reported 15 patients with single ventricle heart and arch hypoplasia operated on with this strategy [5]. In the follow-up period, 3 of the 13 survivors developed a significant recurrent aortic arch gradient after the initial palliation [5].

Clarke et al. reported a 30% recoarctation rate requiring reoperation in a group of 25 patients who underwent PAB and coarctation repair through a left thoracotomy for single ventricle and TGA [4].

In the series by Jahangiri et al. 11 (46%) of the original 24 patients with a similar diagnosis to group I in this paper had an associated aortic coarctation, which was repaired in 9 patients at the time of the PA band [19]. In their analysis, they found a significant reduced survival in patients with obstructed aortic arch [19].

In our series all patients had obstruction either at the level of the systemic outflow and/or at level of the aorta. In particular most of the patients had obstruction at aortic level, which was dealt at time of surgery. The addition of a PAB in patients with residual aortic gradient will cause double obstruction to ventricular outflow, which in turn may promote accelerated ventricular hypertrophy.

Aortic obstruction and myocardial hypertrophy with impairment in diastolic function are two adverse risk factors for the long-term success of the Fontan circulation [11,18]. PAB may also promote pulmonary artery distortion with secondary dysfunction of the pulmonary valve, which will become the systemic valve at the time of a DKS procedure [4]. In the report by Odim et al. 3 out of the 13 survivors developed pulmonary valve regurgitation [5].

Despite these concerns, some authors have preferred initial palliation of these patients by means of PAB with or without aortic arch or coarctation repair. This strategy requires close observation of the subaortic and aortic area and an early DKS procedure should obstruction develop [4—8]. Using a vigorous surveillance to detect subaortic obstruction, Odim et al. were able to recognise early development of subaortic obstruction in 8 of 15 patients following pulmonary artery band [5]. These patients underwent a DKS procedure at a median age of 3.6 months while the remaining 47% did not develop subaortic obstruction and progressed to a bidirectional Glenn shunt at a median age of 9.75 months [5].

In a recent study by the same group, Lan et al. suggested that systemic obstruction associated with a gradient but without ventricular hypertrophy, does not influence long-term outcome [8].

Due to the heterogeneity of the anatomic substrate of these patients, it is difficult to predict which patients will develop subaortic obstruction. Nonetheless, there is a group of patients who may achieve good long-term results with a more classic approach of PAB, particularly in the absence of aortic hypoplasia. Strict adherence to a vigilant follow-up is therefore essential for early detection of any development of aortic gradient before ventricular hypertrophy occurs.

A different surgical strategy of relieving the subaortic obstruction by enlargement of the VSD was firstly reported by Cheung et al. [13]. They have reported their long-term results in a group of 24 patients operated on with this strategy [19]. Their patients were older with a median age of 3.2 years and there were significant associated postoperative complications including 8% heart block, 12.5% aortic insufficiency, (thought to be due to damage to the aortic leaflet), and 8% recurrent obstruction. The difficulty in enlarging the VSD at young age is also reflected by an early mortality of nearly 60% in patients less than 1-year old [19]. Some authors, including ourselves, believe that exposure of the subaortic area can be very difficult in neonates and an additional patch enlargement of the rudimentary right ventricle might be necessary [14,15]. Furthermore, in cases of obstruction due to abnormal attachments of the atrioventricular valve, VSD enlargement may not be effective [14]. Despite the constant course of the conduction axis in hearts with usual atrial arrangements and a dominant left ventricle [1], postoperative heart block occurs in 8—34% patients post procedure [8,19]. Lan et al., in a large series of patients with diagnosis of TGA and DILV or TA, reported their long-term results with different surgical strategies [8]. Among 140 patients, 44 underwent VSD enlargement and 34% needed a postoperative pacemaker because of direct injury to the conduction system. Furthermore, their multivariate analysis showed that pacemaker requirement was an independent risk factor for mortality [8].

Other authors have suggested performing a palliative arterial switch operation in the neonatal period for these patients [20,21]. The main advantage of this approach is to produce an unobstructed systemic outflow tract. However, the unpredictable progressive VSD restriction may significantly limit pulmonary blood flow over necessitating a systemic to pulmonary shunt. Given the other options that are available, this may add unnecessary complexity to a
functional univentricular circulation. For these reasons the experience with this approach is limited to a few cases where the early mortality was between 14.5% and 33% [13,20,21].

We believe that a Norwood procedure is the most effective procedure for this group of patients in the neonatal period. The main drawback of this approach is the potential increase in surgical risk and the complexity of the operation. Our results show that when a Norwood procedure is performed in this group of patients, mortality is comparable to that obtained in patients with other diagnoses, mainly hypoplastic left heart syndrome. In this series there is a trend for a lower mortality in the TGA single ventricle group, but it has not reached statistical significance. Moreover, the survival curves show that, after an initial attrition, there are no late deaths between 5 and 10 years with a survival rate of 72.8%. Furthermore, when we analysed our data according to the era of the operation, only 1 patient of the 11 patients operated since 2002 has died, with a mortality of 9%. We believe that this strategy provides unobstructed outflow thereby reducing early hypertrophy and avoiding the need for subsequent intracardiac reoperations. Preserved ventricular function is an important factor contributing to a successful Fontan circulation [18]. Secondly, by avoiding the potential injury to the conduction system, the Norwood procedure avoids the morbidity and mortality associated with pacemaker induced cardiomyopathy [8].

Having a single morphologic left ventricle as the functional pumping ventricle may be responsible for the good long-term results and it would be logical to predict that this group will have better outcomes than a morphologic right ventricle. However, in a recent publication reporting our mid-term results of the Fontan circulation, left or right ventricular morphology did not influence the long-term results, reflecting that other factors play a more important role in outcome [18].

Tchervenkov et al. reported in 2001 the results of a comparative study between PAB and Norwood operation in patients with the anatomical substrate of single ventricle and systemic obstruction [12]. They showed a 43% mortality in the group treated with PAB with or without aortic arch repair and the four remaining patients in this group experienced a high number of reinterventions due to new onset or recurrent subaortic or aortic obstruction [12]. The group of patients treated with a Norwood procedure had a lower mortality rate of 27%, which is comparable to the mortality of group I (22%) in the present study. Furthermore, patients in this group have required significantly less reinterventions [12].

Mosca et al. also reported excellent results in a population of 38 patients with TA or DILV and TGA treated with a Norwood like procedure [16]. In their report, the actuarial survival at 5 years was 71%, and 60% of their patients had undergone a successful Fontan [16].

Although we have adopted the RV-PA conduit modification of the Norwood procedure for HLHS since 2002 [22], in the case of TGA and single left ventricle we continue to employ and recommend a modified Blalock—Taussig shunt. The reason for this surgical approach is mainly anatomical. As the right ventricle inflow is guarded by a potentially restricted VSD, the shunt has to originate from the left ventricle. Since the left ventricle in these hearts is posteriorly located, the course of the shunt would be long and tortuous posing a higher risk for compression and kinking. Furthermore, a left ventriculotomy could potentially risk damaging the left atrioventricular valve apparatus.

Our late results show good performance status in the group of patients who underwent completion of the Fontan circulation. None have had to undergo reoperations for repair of intracardiac obstructions or recoarctation. Other groups before us have reported similar results, showing very good functional results at completion of the Fontan circulation [16].

We can conclude that the Norwood operation, when employed in patients with TGA and single left ventricle carries a low mortality. It provides an unobstructed systemic outflow tract enabling progression to a Fontan circulation with low mortality and morbidity while achieving a good performance status in the long term.

4.1 Limitations of the study

One of the limitations of this study is that we considered patients operated over a long period of time, during which surgical technique has evolved. Likewise, preoperative and postoperative care has changed and improved over time.

A second limitation is the absence of a control group with comparable anatomical background operated with a different strategy.

References


Appendix A. Conference discussion

Dr R. Lange (Munich, Germany): I just noticed that you seemed to wait a little bit longer in the left ventricle group before you performed the operation. Is there any reason for that?

Dr Lotto: I think that probably reflects the period that these patients had been operated, ranging from the early 90s to nowadays. Nowadays we are operating these patients for the stage II earlier. That probably reflects the long period of time considered.

Dr A. Corno (Liverpool, United Kingdom): I see that you have never used the Sano modification in the single ventricle group. The reason is that you wanted to avoid a ventriculotomy in the functional systemic ventricle or to avoid a kinking between the proximal anastomosis of the shunt and the distal end of the shunt? And if the answer is that you wanted to avoid a ventriculotomy in the systemic ventricle, did you study the difference between the effects of a ventriculotomy on a systemic ventricle or in the systemic morphologically right ventricle for any other Norwood?

Dr Lotto: There are a few things to say about it. First of all, the so-called right ventricle-to-PA conduit is not possible in this group because they have got a diminutive right ventricle, so the conduit will still be underneath the restricted VSD, or potentially restricted VSD. Secondly, if you wanted to do your ventriculotomy on the left, that will create a very long and tortuous conduit with potentials for kinking. And, thirdly, the ventriculotomy might be just where the supravalvular apparatus of the left AV valve is, creating problems with the inflow of the conduit.

Dr S. Sano (Okayama, Japan): It seems to me that some of your patients are categorised as single ventricle with systemic ventricular outflow tract obstruction. Is there any option for Damus-Kaye-Stansel operation rather than Norwood type operation? To me, some of your patients would be better to have a DKS operation rather than Norwood operation. We will present similar data tomorrow morning.

Dr Lotto: First of all, they represent a long experience in Birmingham, so the operation technique has evolved through the years. The other thing to consider is that 14 out of 37 patients had a hypoplastic ascending aorta where enlargement, Norwood type, of the ascending aorta has been done, so probably they reflect that heterogeneity of technique.

Dr V. Tsang (London, United Kingdom): Do you use native tissue anastomosis for your arch construction?

Dr Lotto: We use homograft.