Results of modified Norwood’s operation for hypoplastic left heart syndrome

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Abstract
Objective: The aim of the study was to analyze intermediate results of treatment of the hypoplastic left heart syndrome based on selective indication criteria. Methods: Between February 1997 and May 1999 38 patients with hypoplastic left heart syndrome (n = 35), or with functional variant of hypoplastic left heart syndrome (n = 3) were admitted to our department. Contraindications for surgery were birth weight <2500 g, diameter of ascending aorta <2 mm, severe tricuspid regurgitation persisting after initial stabilization, pulmonary regurgitation more than mild, dysfunction of the systemic right ventricle and failure to effectively resuscitate circulation before surgery. Results: Based on these criteria surgery was not indicated in 17 patients. Twenty-one infants were operated on by modified Norwood’s procedure using only autologous great vessel tissue for reconstruction of systemic outflow. Overall hospital mortality was 14% (three patients). Eighteen survivors (86%) were discharged with well-balanced circulation. There was one late death (5%). Thirteen patients had already undergone the second stage (bi-directional Glenn) with no death. The mean follow-up was 13.2 ± 9.1 months (range 4–32 months). Considering both early and late events the probability of survival for the whole group (n = 21) from the time of surgery was 86% at 1 month, 80% at 12 months, and it remained unchanged at 18 and 24 months of follow-up. Conclusions: Only a limited number of European countries offer surgical treatment of hypoplastic left heart syndrome. Promising intermediate results (80% survival rate after stage I and II) achieved at our department do not only reflect overcoming the learning curve but also a selective approach to indication for surgery as well. In a country with limited resources selective approach to the patients with hypoplastic left heart syndrome is justified. © 2000 Elsevier Science B.V. All rights reserved.

Keywords: Hypoplastic left heart syndrome; Risk factors of the first-stage palliation; Modified Norwood procedure; Selective indication criteria

1. Introduction

The hypoplastic left heart syndrome (HLHS) is characterized by various hypoplasia of the left-sided heart structures, where mitral stenosis (MS) or mitral atresia (MA) is combined with aortic stenosis (AS) or aortic atresia (AA). An integral part of HLHS is a hypoplasia of the left ventricle (LV), ascending aorta, aortic arch, and a critical coarctation of aorta [1]. In the Slovak Republic about 15 patients with HLHS are born annually. Without surgical intervention HLHS is universally a lethal condition, accounting for 25% of all cardiac deaths within the first week of life.

Most neonates with HLHS are otherwise well-developed babies without major associated malformations.

Principally, there are three options in the management of children with this type of congenital heart defects. Firstly, children with HLHS are left untreated; secondly, they undergo heart transplantation, and thirdly, a staged-approach leading to a modified Fontan procedure is employed. The first-stage palliation is associated with the highest risk. Its aim is to eliminate the pressure overload of a single-ventricle circulation by creating an unobstructed outflow tract and aortic arch, and to provide a well-defined shunt-dependent pulmonary circulation, hence, to minimize the volume overload of a single-ventricle circulation and to ensure the growth of pulmonary arteries (PA) [2–4].

The objective of this study is to define selective operative criteria on the prospective basis and to analyze intermediate results of the treatment of HLHS in the Slovak Republic since 1997.
2. Materials and methods

2.1. Patients

Between February 1997 and May 1999, 38 patients with the diagnosis of HLHS were admitted to Department of Cardiovascular Surgery, Children’s University Hospital in Bratislava. In our center contraindications for surgery include immaturity, body weight below 2500 g, diameter of aortic root below 2 mm, severe tricuspid regurgitation persisting after initial stabilization, more than a mild pulmonary regurgitation, the right ventricular dysfunction and inability to stabilize baby before the surgery due to multiple reasons: severe hypoxia associated with intact interatrial septum and failure to perform effective atroioseptostomy; late referral with unresponsive shock and severe metabolic acidosis. The degree of preoperative tricuspid or pulmonary regurgitation was graded as none, mild, moderate, or severe by color Doppler mapping of the regurgitant jet in the right atrium and right ventricle, respectively. Based on these criteria surgery was not considered in 17 patients (Table 1). A total of 21 patients underwent the first-stage palliation. With respect to anatomic subtypes 14 patients had a combination of MS and AS, three patients had MS and AA, one patient had MA in combination with AA, and three patients had variant of HLHS (Table 2). In the same period, 13 patients have undergone subsequently the second-stage procedure (bi-directional Glenn).

2.2. Operation technique and perfusion protocol

The operation was performed in a deep hypothermia (at rectal temperature of 18°C) and circulatory arrest (Table 2) [5,6]. All patients were operated on by a modified technique using only autologous great vessel tissue for reconstruction of systemic outflow and 3.5 mm BT shunt (Fig. 1) [7,8]. Mediastinum was approached through the midline sternotomy and the thymus gland was completely removed. Arch vessels were dissected free and mobilized before opening the pericardium. Following opening of the pericardium, both pulmonary arteries were encircled. Then, cardiopulmonary bypass was instituted by cannulation of arterial duct (PDA) and the right atrial appendage. The mobilization of aortic arch and isthmic aorta was completed during cool-

Table 1
Reasons for contraindication of surgical treatment (February 1997–May 1999, n = 17)

<table>
<thead>
<tr>
<th>Reason</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic annulus &lt;2 mm</td>
<td>4</td>
</tr>
<tr>
<td>Severe tricuspid regurgitation</td>
<td>3</td>
</tr>
<tr>
<td>More than a mild pulmonary regurgitation</td>
<td>1</td>
</tr>
<tr>
<td>Failure to effectively resuscitate circulation</td>
<td>4</td>
</tr>
<tr>
<td>Major associated defects</td>
<td>2</td>
</tr>
<tr>
<td>Weight &lt;2500 g</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>17</td>
</tr>
</tbody>
</table>

ing of the patient. PA was transected just beneath the bifurcation, and the latter was enlarged with a glutaraldehyde-treated pericardial patch. Following at least 20 min of cooling, the circulation was arrested at a temperature of 18°C, arch vessels were temporarily occluded by tourniquets, and an antegrade blood cardioplegia was administered from a side port of the arterial cannula after clamping of the descending aorta. All cannulas were removed from the heart. Then, the right atrium was opened and the interatrial septum (IAS) was excised. The mobilization of the descending aorta was completed after PDA transsection. The entire isthmic part of the aorta was removed with adjacent ductal tissue. Then, aortic arch was incised in its concavity, and the part of ascending aorta was opened. The posterior wall of the aortic arch and descending aorta were partially sutured by end-to-end anastomosis. Subsequently, aortic arch and portions of descending and ascending aorta were enlarged with the pulmonary trunk creating thus an unobstructed aortic arch. A modified Blalock–Taussig shunt using a 3.5-mm Gore-Tex tube was constructed between the take-off of the right subclavian artery and the right pulmonary artery after re-instituting the circulation and warming the patient. Epicardial pacing wires were placed on the right atrium and ventricle. Monitoring catheters were placed into the right atrium and to the superior vena cava (SVC) to measure the filling pressure of the systemic ventricle and mixed venous oxygen saturation. A modified ultrafiltration was routinely used after weaning from the cardiopulmonary bypass.

2.3. Postoperative management

If myocardial distention or chest wall edema developed, the sternum was left primarily open. At the completion of the operation a latex membrane was sewn to the skin edges and Ioban dressing was applied. The sternum was closed when tissue edema subsided, usually on postoperative days 3 to 5.

Dopamine at low (5 µg/kg per min) or moderate (10 µg/kg per min) doses was used as a standard inotropic agent. Amrinone was used in patients with persisting low cardiac output.

Intermittent bedside calculation of Qp/Qs based on known systemic oxygen saturation, mixed venous oxygen saturation from SVC and assumed pulmonary venous
saturation (95%) was performed very frequently in operating room and during postoperative period. $Q_p/Q_s$ was calculated using a Fick equation, assuming that the dissolved oxygen was zero [9]. All therapeutic interventions (adjustment of ventilator regimen, inotropic support, afterload reduction) were based on continuous assessment of clinical condition in conjunction with frequently repeated calculation of $Q_p/Q_s$. Sedation and muscle paralysis continued for the next 12–24 h postoperatively according to hemodynamic condition except in patients with sternum primarily

Fig. 1. (A): The main pulmonary artery is transected distally. Isthmic part of aorta is excised. The aortic arch is opened into the ascending aorta. (B): The descending aorta is anastomosed to the posterior wall of the aortic arch. Proximal main pulmonary artery is anastomosed to the descending aorta, aortic arch and distal ascending aorta. MPA, mean pulmonary artery; MBT shunt, modified Blalock–Taussig shunt.

Fig. 2. (A): Selective injection of reconstructed systemic outflow demonstrating unobstructed aortic arch and descending aorta. (B): Selective injection of BT shunt demonstrating pulmonary artery architecture.
left open when it continued for 12–24 h after successful sternal closure. The median length of ventilation was 6 days (range 2–19 days) and the median length of stay in intensive care unit (ICU) was 8 days (range 6–44 days) postoperatively. For comparison, the median ICU stay was 3 days and the length of ventilation was 1 day in all other remaining patients operated on in our department.

2.4. Late management

Elective cardiac catheterization was performed 2–3 weeks before the second-stage of palliation (Fig. 2). The second-stage bi-directional Glenn procedure was performed 5–10 months after the first-stage operation, whenever electively possible at the age of 6 months. Operation was performed through median sternotomy with cardiopulmonary bypass (CPB) on beating heart. Anastomosis between SVC and PA was placed centrally to augment the origin of left pulmonary artery (LPA). Aortic arch augmentation with the patch, if necessary, was performed using hypothermic circulatory arrest.

2.5. Statistical analysis

Data were analyzed using a statistical program (JMP Statistical Analysis, Cary, NC). The primary outcome variable was survival after operation. Early failure was defined as death within 30 days of operation and late failure was defined as death beyond 30 days after operation. Multiple clinical parameters were analyzed for their possible impact on survival using univariate analysis with Pearson’s chi-square for categorical variables and Student’s t-test for continuous variables (P < 0.05). The Kaplan–Meier method was used for actuarial survival analysis. All results are expressed as mean ± SEM.

3. Results

3.1. Early mortality and morbidity

Three of 21 operated patients died in hospital (14%). One patient died from interstitial pneumonia on postoperative day 11 and two patients died due to failure to control balance between Qp and Qs on postoperative days 1 and 4, respectively.

Early postoperative complications are listed in Table 3. Univariate analysis of certain variables (age, weight, sex, anatomic subtype, diameter of ascending aorta, length of cardiopulmonary bypass) did not find any risk factor for death.

3.2. Late mortality

There was one late death (5%) 7 months after the first-stage palliation probably due to pneumonia. Autopsy was not obtained.

3.3. Long-term survival

Of 17 survivors (81%), one patient was re-operated at 3 months after the first procedure due to a gradual development of interatrial restriction. Temporary compression of left bronchus was noted in one patient (Table 3). At the mean age of 6.9 ± 1.9 months, thirteen of 17 survivors were converted to bi-directional Glenn (the second-stage procedure). Preoperative angiography revealed mild stenosis of the origin of the left pulmonary artery in six patients (35%). Aortic arch obstruction was noted in one patient (5.8%) with catheter-measured gradient 30 mmHg. Concomitant procedures at the stage II palliation were as follows: one reconstruction of the pulmonary artery by patch augmentation, and repair of the neoaortic arch obstruction. There was no early or late death. Three patients had deep sternal wound infection (DSWI). DSWI was defined in accordance with the guidelines of the Centers for Disease Control and Prevention [10]. Our protocol includes re-exploration with thorough debridement of all necrotic tissues, primary closure of the sternum with closed irrigation system using betadine (povidone-iodine) solution. Ten days antibiotic course of vancomycin and amikacin is administered until no specific culture is available.

All patients are doing well with adequate growth. Mild tricuspid regurgitation was noted in three patients (17.6%). Neurologic examination revealed only mild tone alteration (hypotonia) in five patients (29%) at one year of age, the rest of patients showed normal neurologic development. Four patients are awaiting the second-stage repair (Fig. 3).

Patients were followed on average for 13.2 ± 9.1 months (range 4–32 months). Considering both early and late events the probability of survival for the whole group (n = 21) from the time of surgery was 86% (CL 79–93%) at 1 month, 80% (CL 71–89%) at 12 months and it remained unchanged at 18 and 24 months of follow-up (Fig. 4).

4. Discussion

The treatment of HLHS is one of the most demanding cardio surgical procedures not only due to technical reasons, but also due to inherent hemodynamic instability in postoperative period. The first-stage palliation represents a typi-
ical therapeutical paradox, when a complex reconstruction of the outflow tract of the systemic ventricle and formation of a shunt-dependent pulmonary circulation does not significantly improve the hemodynamics immediately after surgery. Before and after the procedure, the patient has only one effective systemic pump (right ventricle), and the distribution of pulmonary and systemic blood flow is related to the ratio of pulmonary and systemic resistance. This might be the reason for extremely high initial mortality associated with the first-stage procedure. The initial mortality of the first-stage procedure has not been reduced below 20% even in the best centers [11–16]. Multiple preoperative risk factors related to stage I mortality are considered significant [13,14]. Pre-stage I anatomy (ascending aorta <2 mm, anatomic subtype MS/AA), low birth weight (weight <3000 g), inadequacy of resuscitative efforts (preoperative pH) strongly influenced both operative and intermediate mortality among stage I survivors according to Boston group reports [13]. Preoperative moderate or severe tricuspid regurgitation was defined as a significant risk factor in long-term survival after palliative surgery for HLHS in accordance with Philadelphia’s experience [15].

Based on this data selective operative criteria were introduced in our institution on prospective bases before the program for HLHS was launched.

Modified technique using only autologous great vessel tissue for reconstruction of systemic outflow reported by Bu’Lock [7] and Fraser [8] in 1995 was used in all patients. The one month postoperative survival in our report was 86% among 21 patients after the first stage palliation. We did not find any significant risk predictors for death, but this might have been influenced by our selection criteria. There was only one late death, 7 months after initial operation among 18 survival.

In a period between the first and subsequent stages, candidates for a Fontan-type of repair must be followed systematically in order to identify early possible surgical residues. Residual aortic arch obstructions, obstruction of interatrial communication, distortion of pulmonary arteries are encountered most commonly [13]. Modified technique increases the risk of compression of the left main bronchus and left pulmonary artery. In addition, the central pulmonary artery stenosis is more likely to occur, because the main pulmonary artery is divided more distally. The incidence of LPA origin stenosis is high (35%) in our group of patients, but only one patient needed patch augmentation of central PA during the second stage. The rest of patients were fixed by centrally located Glenn anastomosis. There was only one patient with temporary compression of left main bronchus which resolved spontaneously with the patient growth. Aortic arch obstruction necessitating surgical intervention was noted only in one patient. There was no death in patients undergoing the bi-directional Glenn shunt.

Eighty percent survival rate after stage I and II of palliation of HLHS sing a modified Norwood’s procedure is promising, however, the number of patients treated and the length of follow-up do not permit any integrated conclusions. A selective approach to indication eliminates, in part, patient groups at the highest risk. This approach is justified especially in countries with limited resources.

5. Conclusion

The hypoplastic left heart syndrome represents a complex medical and ethic problem. Only few European countries offer the surgical treatment of HLHS. The reason is a persistently high mortality (25–40%) of the first-stage procedure even in the best centers. Promising intermediate results (80% survival rate after stage I and II) achieved at our department do not only reflect overcoming the learning curve but also a selective approach to indication for surgery as well.
References