

# Experience With the Norwood Operation for Hypoplastic Left Heart Syndrome

Juan A. García-Hernández,<sup>a</sup> Juan D. González-Rodríguez,<sup>a</sup> Adoración I. Martínez-López,<sup>a</sup> David Canalejo-González,<sup>a</sup> Antonio Romero-Parreño,<sup>a</sup> José Santos de Soto,<sup>b</sup> Mercedes Loscertales-Abril,<sup>a</sup> Aurelio Cayuela-Domínguez,<sup>d</sup> and Mauro Gil Fournier-Carazo<sup>c</sup>

<sup>a</sup>Unidad de Gestión Clínica de Cuidados Críticos y Urgencias, Hospital Infantil Universitario Virgen del Rocío, Sevilla, Spain

<sup>b</sup>Sección de Cardiología Pediátrica, Hospital Infantil Universitario Virgen del Rocío, Sevilla, Spain

<sup>c</sup>Servicio de Cirugía Cardiovascular, Hospital Infantil Universitario Virgen del Rocío, Sevilla, Spain

<sup>d</sup>Unidad de Apoyo a la Investigación, Hospital Infantil Universitario Virgen del Rocío, Sevilla, Spain

**Introduction and objectives.** To describe our experience and to identify risk factors for in-hospital mortality.

**Methods.** Between October 1991 and June 2005, 42 children underwent the Norwood procedure. In the first 30 patients, pulmonary circulation was established using a modified Blalock-Taussig shunt (Group 1), while a right ventricle to pulmonary artery conduit was used in the remaining 12 (Group 2). Preoperative anatomic features and procedural factors were analyzed with respect to their impact on mortality. Postoperatively, data were collected on arterial blood pressure, arterial and venous oxygen saturation, arterial pH, venous pCO<sub>2</sub>, the PaO<sub>2</sub>/FiO<sub>2</sub> ratio, tissue oxygen extraction, and dead space fraction. The association between each individual variable and mortality was investigated.

**Results.** Thirty patients (71.4%) had both aortic and mitral atresia, 8 (19%) had either aortic or mitral atresia, and 4 (9.5%) had no valvular atresia. There was no statistically significant difference in postoperative mortality between the groups 1 and 2 (12/22 [54.5%] vs 7/12 [58.3%];  $P=0.56$ ). The only significant risk factor for in-hospital mortality was a longer cardiopulmonary bypass time ( $P=0.01$ ) and, for intraoperative mortality, primary rather than delayed sternal closure ( $P=0.004$ ). Venous pCO<sub>2</sub>, the mean dead space fraction, and tissue oxygen extraction all tended to be higher among infants who died, but the difference was not statistically significant.

**Conclusions.** Use of a right ventricle to pulmonary artery conduit did not improve postoperative survival. Both a long cardiopulmonary bypass time and primary sternal closure were associated with increased mortality.

**Key words:** Congenital heart disease. Cardiopulmonary bypass. Hypoplastic left heart syndrome. Norwood operation.

Correspondence: Dr. J.A. García Hernández.  
Unidad de Gestión Clínica de Cuidados Críticos y Urgencias.  
Hospital Infantil Universitario Virgen del Rocío.  
Ruiseñor, 5, portal 3, 3.º B. 41010 Sevilla. España.  
E-mail: garcier@gmail.com

Received November 17, 2006.

Accepted for publication March 29, 2007.

## Resultados de la intervención de Norwood para el síndrome del corazón izquierdo hipoplásico

**Introducción y objetivos.** Describir nuestra experiencia e identificar factores de riesgo de mortalidad hospitalaria.

**Métodos.** Entre octubre de 1991 y junio de 2005 intervenimos a 42 niños con la técnica de Norwood. Los 30 primeros recibieron una fístula de Blalock-Taussig (grupo 1) y los 12 restantes, un conducto entre el ventrículo derecho y la arteria pulmonar (grupo 2). Se analizaron los factores anatómicos y de la técnica con respecto a la mortalidad. Se recogieron variables del postoperatorio, incluidas la presión arterial, la saturación arterial y venosa de oxígeno, el pH arterial, la pCO<sub>2</sub> venosa, la relación PaO<sub>2</sub>/FiO<sub>2</sub>, la extracción tisular de oxígeno y el espacio muerto, para estudiar su asociación con la mortalidad.

**Resultados.** En total, 30 (71,4%) pacientes tenían atresia aórtica y mitral; 8 (19%) tenían atresia aórtica o mitral y 4 (9,5%) no tenían atresia. No hubo diferencias significativas en la mortalidad postoperatoria entre los grupos 1 y 2 (12/22 [54,5%] frente a 7/12 [58,3%];  $p=0,56$ ). El único factor de riesgo de mortalidad hospitalaria fue un tiempo de circulación extracorpórea prolongado ( $p=0,01$ ), y el de mortalidad intraoperatoria, el cierre primario del esternón ( $p=0,004$ ). La pCO<sub>2</sub> venosa, el espacio muerto pulmonar y la extracción tisular de oxígeno fueron superiores en los niños fallecidos, pero las diferencias no fueron significativas.

**Conclusiones.** El uso de un conducto entre el ventrículo derecho y la arteria pulmonar no mejoró la supervivencia postoperatoria. Un tiempo de circulación extracorpórea prolongado y el cierre primario del esternón se asociaron con un aumento de la mortalidad.

**Palabras clave.** Cardiopatía congénita. Circulación extracorpórea. Síndrome del corazón izquierdo hipoplásico. Norwood.

**ABBREVIATIONS**

PICU: pediatric intensive care unit  
 $\text{PaO}_2/\text{FiO}_2$ : respiratory quotient  
 $\text{Vd}/\text{Vt}=(\text{PaCO}_2-\text{EtCO}_2/\text{PaCO}_2)$ : lung dead space  
 $\text{EtO}_2=(\text{SaO}_2-\text{SvO}_2/\text{SaO}_2)$ : tissue oxygen extraction

**INTRODUCTION**

The hypoplastic left heart syndrome is a serious congenital cardiopathy that leads to surgery in the first days of life. The treatment is palliative and it is made in several stages, of which the intervention of Norwood<sup>1</sup> is the first. It attempts to make the right ventricle act like a systemic ventricle, connecting the trunk of the pulmonary artery to the aorta. The pulmonary flow is attained through a fistula between the aorta and the pulmonary artery. Although the results obtained in the last years have improved,<sup>2,3</sup> it continues to have a high mortality. The main problem of this intervention is the hemodynamic instability; in order to try and avoid it, in year 2003 the modification of Shunji Sano,<sup>4</sup> consisting of the substitution of the fistula for a non-valvulated tube introduced between the right ventricle and the pulmonary artery. In our hospital, the first Norwood intervention was made in 1991,<sup>5</sup> and in 2003 we incorporated the Sano modification, based on the good results obtained in others centers.<sup>6,7</sup> The objectives of this study are: *a*) to communicate our experience with the classic Norwood technique and to determine if we have managed to increase the survival with the modification of Sano, and *b*) to identify prognostic factors of early hospital mortality among the different variables.

**METHODS****Patients**

The study was planned as prospective, descriptive, and observational, to be carried out in the period between October 1991 and June 2005. We included a total of 42 diagnosed children with hypoplastic left heart syndrome from the Hospital Virgen del Rocío of Seville. Among the anatomical characteristics of the cardiopathy we emphasized that the average diameter of the ascending aorta was 3.4 mm (rank, 1-8 mm); in 23 (54.8%) children it was  $\leq 3$  mm, in 12 (28.6%) it was between 3 and 5 mm, and in 7 (16.7%) it was  $\geq 5$  mm. In 30 (71.4%) patient the diagnosis was aortic atresia and mitral atresia, in 8 (19%), aortic atresia or mitral atresia and in 4 (9.5%), of aortic stenosis and mitral stenosis. 40.5% (17/42) had another associated cardiopathy and coarctation of the aorta was the most frequent.

**Surgical Intervention**

The intervention was made at an mean age of 10.1 days (range, 3-24 days) and an mean weight of 3.3 kg (range, 2.3-4.6 kg). The first 30 patients (group 1) underwent the classic<sup>1</sup> Norwood technique, receiving a modified Blalock-Taussig fistula of different diameters, and in the other 12 (group 2) the Sano<sup>4</sup> variant was used. All the interventions were made under extracorporeal circulation (ECC) under deep hypothermia, and periods of circulatory shutdown for the reconstruction of the aortic arch were employed, done in most of cases (30/42; 71%) by means of a terminoterminal suture between the pulmonary artery and the aorta; when, by technical reasons, this was not possible (12/42; 28.6%), we used an aortic graft. At the beginning of the operation dobutamine was administered at doses of 5  $\mu\text{g}/\text{kg}/\text{min}$ , low doses of adrenalin that oscillated between 0.05 and 0.1  $\mu\text{g}/\text{kg}/\text{min}$ , and a milrinone perfusion at 0.5  $\mu\text{g}/\text{kg}/\text{min}$ . During the process of retrieval from ECC, we administered to an inspiratory fraction of oxygen of 100% and inhaled nitric oxide at 20 ppm, attempting to reduce the pulmonary vascular resistance to the maximum, which were increased at that moment. The  $\text{P}_{50}$  of the ECC time, aortic clamping and circulatory shutdown was of 185.5 min (range, 173-267), of 56.5 min (range, 50-65), and 56 min (range, 49-72), respectively. In 19 (45.2%) cases the thorax was left open in the operating room, deciding on a deferred closure of the sternum in the pediatric intensive care unit (PICU) due to hemodynamic instability upon finalizing the intervention.

**Postoperative Treatment**

The PICU treatment initiated in the operating room was continued, with the objective to optimize cardiac output and to reduce to the systemic and pulmonary vascular resistance to the maximum. The inspiratory fraction of oxygen and nitric oxide were being reduced progressively, once a stable  $\text{SaO}_2$  in the optimal accepted range was obtained, which we considered between 75% and 80%. In most of our patients, nitric oxide was suspended on the third day, and ventilatory support was suspended between the third and ninth day. Sedation and analgesia were obtained with midazolam and fentanyl, and we only resorted to the neuromuscular paralysis with vecuronium in children whose sternal closure was deferred. The process of weaning off the respirator began once the child was hemodynamically stable and when the thorax had been closed, in children with deferred closure. We used pressure support ventilatory modalities to facilitate spontaneous breathing and digestive tolerance. We initiated enteral nutrition through a transpyloric tube on the second postsurgical day, preferentially with hydrolyzed maternal milk or proteins. In the analytical control we measured arterial and venous blood gases, oxygen saturation ( $\text{SO}_2$ ) by

noninvasive, and continuous pulse-oxymetry and the carbon dioxide (CO<sub>2</sub>) through capnography. We made an immediate postsurgical heart ultrasound to evaluate ventricular function, the size of the auricular defect, the competence of the tricuspid valve, and the flow through the fistula, or the ventriculopulmonary tube.

### Data Recovery and Analyzed Variables

Hospital mortality, constituted by intrasurgical and postsurgical mortality was analyzed before discharge. In addition, groups 1 and 2 were compared in relation to mortality, intrasurgical, and postsurgical. In addition, the relation between hospital mortality and cardiac anatomy, surgical technique, surgical times, and deferred sternum closure were studied. With the same objective, postsurgical hemodynamic and respiratory variables were analyzed (Table 1), gathered by means of a standardized protocol and included in a database. The analyzed variables were systolic, diastolic, and mean arterial pressure, the arterial and venous saturation of oxygen, arterial pH, venous pCO<sub>2</sub>, PaO<sub>2</sub>/FiO<sub>2</sub> relation, tissue oxygen extraction (SaO<sub>2</sub>-SvO<sub>2</sub>/SaO<sub>2</sub>), and the pulmonary dead space (CO<sub>2art-et</sub>CO<sub>2</sub>/CO<sub>2art</sub>). Tissue oxygen extraction (EtO<sub>2</sub>) measures the oxygen consumption by waves, so that when tissue perfusion is jeopardized, it slows blood flow and increases its extraction, lowering the S<sub>v</sub>O<sub>2</sub> and increasing the venous PCO<sub>2</sub>. The tissue oxygen extraction and venous pCO<sub>2</sub> allow the determination of cardiac output. The reason to have chosen, in addition, pulmonary dead space (Vd/Vt) was based on the fact that, in the intervention of Norwood, the pulmonary flow is jeopardized and zones of the lung have a predominance of ventilation over perfusion. This fact produces an increase of the dead space, which is translated in a difficulty to exhale carbon dioxide. Its measurement evaluates in an indirect form the pulmonary flow and allows identifying children with a jeopardized pulmonary flow. The fraction of the pulmonary dead space is considered normal if between 0.2 and 0.4. The successive determinations of the variables previously mentioned were done in the following sequence: first, in operating room when coming out of ECC; second, after the immediate admission into the PICU; third, at 24 h of the first day; fourth, at 8 h of the second day; fifth, at 18 h of the second day; sixth, at 8 h of the third day; and the seventh and last one, at 18 h of the third day.

### Follow-Up

We analyzed, additionally, late mortality and the patients who underwent a second and third procedures, that is the Glenn and Fontan procedures, were classified. The degree of function of survivors was determined during their last check-up in the outpatient clinic of cardiology.

### Statistical Analysis

Descriptive statistic of the qualitative variables was done, using absolute and relative frequencies. Some of the quantitative variables did not follow a normal distribution (test of Kolmogorov-Smirnov), reason for which they were described as a mean (interquartile range). The comparison of variables according to groups was made with the test of  $\chi^2$  for the qualitative variables, and with the Mann-Whitney U for the quantitative variables. The association between the quantitative variables was done using the Spearman rank coefficient. All the analyses were made with SPSS software, version 14.0. A *P* value less than .05 was considered significant.

### RESULTADOS

Global hospital mortality was 64.3% (27/42), 66.6% (20/30) in group 1, and 58.3% (7/12) in group 2 (Table 2). The age and the weight of the children at the moment of the intervention did not have a significant statistical relationship with mortality (Table 3). In relation to the cardiac anatomy, hospital mortality was 73.3% (22/30) in the group of children with mitral and aortic atresia, 50% (4/8) in the group with mitral or aortic atresia, and of 25% (1/4) in the group with stenosis (Table 4). The children who had an ascending aorta diameter of  $\leq 3$  mm presented a hospital mortality of 69.6% (16/23); those that had a diameter between 3 and 5 mm, of 66.7% (8/12), and those that had a diameter  $\geq 5$  mm, of 42.9% (3/7) (Table 5). These differences, which relate the cardiac anatomy to hospital mortality, although spectacular, did not have statistical meaning. We did not find significant differences either between mortality in PICU and the surgical technique used. The one obtained in group 1 was 54.5% (12/22), after excluding from the analysis 8 children who died in the operating room, and for group

**TABLE 1. Analyzed Variables**

---

Age and weight at the time of the intervention
Anatomy of the cardiopathy
Diameter of the ascending aorta
Associated congenital cardiopathies
Surgical techniques (Norwood vs Sano)
Time on extracorporeal circulation
Time of cardiac anoxia
Time of cardiac arrest
Deferred sternum closure
Arterial oxygen saturation (S <sub>a</sub> O <sub>2</sub> )
Venous oxygen saturation (S <sub>v</sub> O <sub>2</sub> )
Arterial pH (pHa)
Systolic arterial pressure
Dyastolic arterial pressure
Mean arterial pressure
Respiratory coefficient (PaO <sub>2</sub> /FiO <sub>2</sub> )
Tissue oxygen extraction (S <sub>a</sub> O <sub>2</sub> -SvO <sub>2</sub> /SaO <sub>2</sub> )
Pulmonary dead space (CO <sub>2art-et</sub> CO <sub>2</sub> /CO <sub>2art</sub> )

---

**TABLE 2. Relationship Between the Surgical Technique With Operating Room Mortality, Pediatric Intensive Care Unit, and Total Hospital Mortality\***

	Deaths Operating Room	Deaths PICU	Hospital Mortality	Survivors
Group 1 (n=30)	8/30 (26.6%)	12/22 (54.5%)	20/30 (66.6%)	10/22 (45.4%)
Group 2 (n=12)	0	7/12 (58.3%)	7/12 (58.3%)	5/12 (41.6%)
Total (n=42)	8/42 (19%)	19/34 (55.9%)	27/42 (64.3%)	15/34 (44.1%)

\*PICU indicates pediatric intensive care unit.

**TABLE 3. Differences in the Following Variables Among the Groups of Surviving Children and the Ones That Died in the Pediatric Intensive Care Unit\***

Variables	Dead	Survivors	P
Age, mean (SD), days	9.6 (4.7)	11.1 (5.7)	.351
Weight, mean (SD), kg	3.3 (0.4)	3.3 (0.6)	.588
Time ECC, min	245 (177.5-300)	180 (157.5-184.5)	.010†
Time AC, min	58 (50-71)	56 (49-58)	.161
Time CA, min	56 (50.5-77.5)	55.5 (48-60)	.355
Deferred sternum closure	10/19 (52.6%)	9/19 (47.4%)	.152

\*AC indicates aortic clamping; ECC, extracorporeal circulation; CA, cardiac arrest.

†Statistical significance.

Variables of age and weight are expressed as a typical mean and standard deviation. The rest of the variables are expressed as percentiles: P<sub>50</sub> (P<sub>25</sub>-P<sub>75</sub>).

2 it was 58.3% (7/12) ( $P=.561$ ) (Table 2), and in this group no child died (Table 5). This difference of operating room mortality between groups 1 and 2 turned out to be significant ( $P=.05$ ) (Table 2). In relation to the surgical times, only the time of ECC was associated significantly with mortality, so that it was greater in those that died in the operating room (Table 6) and in PICU (Table 3). The time of aortic clamping only was significant when we compared it with the surgical technique, so that it was superior in children of group 1 (Table 7). The children to whom the thorax was closed in the operating room (19 cases) had a greater intrasurgical mortality than the children with closure deferred to the PICU ( $P=.004$ ) (Table 6). Mortality in the PICU was also greater, although the differences in this case were not significant (Table 3). The deferred closure was most frequently made in children in-group 2 in relation group 1 ( $P=.014$ ) (Table 7). As far as the postsurgical monitorization in the PICU, there were no significant differences between the groups of survivors and deceased in the following variables: S<sub>a</sub>O<sub>2</sub>, arterial S<sub>v</sub>O<sub>2</sub>, pH, PaO<sub>2</sub>/FiO<sub>2</sub>, and systolic, diastolic, and mean arterial pressure. Nevertheless, although there were no differences in the P<sub>v</sub>CO<sub>2</sub>, the EtO<sub>2</sub> and the pulmonary dead space (Vd/Vt), the deceased displayed superior numbers.

Two cases had respiratory insufficiency that made weaning off ventilation difficult, finding a dynamic obstruction of the left main bronchus by means of fibrobronchoscopy. The study of the airways by means

**TABLE 4. Anatomical Variants of the Cardiopathy and Their Relation to Mortality**

Anatomy of the Cardiopathy	Patients	Deaths
Mitral and aortic atresia	30	22 (73.3%)
Mitral or aortic atresia	8	4 (50%)
Mitral atresia	6	2 (33.3%)
Aortic atresia	2	2 (100%)
No atresia	4	1 (25%)
Total	42	27

**TABLE 5. Diameter of the Ascending Aorta and its Relationship With Mortality**

Diameter Ascending Aorta	Patients	Deaths
≤3 mm	23/42 (54.8%)	16/23 (69.6%)
>3 to <5 mm	12/42 (28.6%)	8/12 (66.7%)
≥5 mm	7/42 (16.7%)	3/7 (42.9%)
No.	42	27

**TABLE 6. Differences in the Following Variables Between the Groups of Children That Survived the Intervention and Were Admitted to the Pediatric Intensive Care Unit and the Ones That Died in the Operating Room\***

Variables	Deaths in the Operating Room	Survivors	P
Time ECC, min	281.5 (245-300)	182 (170-245)	.024†
Time AC, min	60 (50-71)	56 (50-64)	.879
Time CA, min	75 (51-87)	56 (48.5-66.5)	.161
Deferred sternum closure	0/19 (0%)	19/19 (100%)	.004†

\*AC indicates aortic clamping; ECC, extracorporeal circulation; CA, cardiac arrest.

†Statistical significance.

Variables are expressed as percentiles: P<sub>50</sub> (P<sub>25</sub>-P<sub>75</sub>).

of computerized tomography (CT) with three-dimensional reconstruction and magnetic resonance (MR) demonstrated that it was produced by an extrinsic compression of the neo-aorta. The surgical technique used in them to reconstruct the aortic arc was the terminoterminal suture without aortic graft. Both patients

**TABLE 7. Differences Among the Variables of Groups 1 and 2\***

Variables	Group 1	Group 2	P
Operating room deaths	8/30; 26.7%	0/12; 0%	.050†
Deaths in PICU	12/22; 54.5%	7/12; 58.3%	.561
Time ECC, min	216.5 (175-291)	176.5 (141-191.5)	.103
Time CA, min	59 (55-71)	50 (43-55)	.013†
Time Ca, min	57.5 (51-77.5)	50 (44-56)	.111
Deferred sternum closure	10/30 (33.3%)	9/12 (75%)	.014†
Hour ventilation	216 (120-480)	76.5 (8-389.5)	.167
Hours of stay in the PICU	480 (384-624)	76.5 (8-492)	.056†

\*AC indicates aortic clamping; ECC, extracorporeal circulation; CA, cardiac arrest; PICU, pediatric intensive care unit.

†Statistical significance.

Group 1: classic Norwood.

Group 2: Sano variant.

Numeric variables expressed in percentiles: P<sub>50</sub> (P<sub>25</sub>-P<sub>75</sub>).

died due to respiratory insufficiency before a tensile stent could be implanted in the bronchial light.

Of the 15 given children discharged from PICU, 5 died before the second surgery and in 8 of the 10 survivors a Glenn procedure was made, with a survival of 100%. Six of the patients belonged to group 1 and 2 to group 2. The mean age and the weight of them in the last review was 19 months (range, 8-31) and 9 kg (range, 6.8-11.3 kg), and all were in Class I of the New York Heart Association classification.

## DISCUSSION

Surgical treatment of the hypoplastic left heart syndrome has been a reason for worry among surgical teams due to its elevated mortality. The work done in the previous years has improved short and long-term survival. This fact is related in part with the introduction of a modification in the classical Norwood technique, consisting in the substitution of the fistula for a tube connecting the right ventricle with the pulmonary artery. This procedure improves the growth of the pulmonary arteries with a more uniform distribution.<sup>8</sup> It has been shown, additionally, that the children present a more hemodynamically stable postoperative phase,<sup>4</sup> therefore achieving a reduction in the interstage mortality.<sup>9,10</sup> For these motives, in 2003 we decided to introduce the Sano modifications at our hospital.

The hypoplastic left heart syndrome is a cardiopathy with many anatomical variants that are frequently associated with other malformations or genetical alterations; its proper evaluation and presurgical diagnosis allows us to establish a prognosis, which is independent of the surgical technique and the postsurgical care. In this sense some authors have found a larger mortality among children with associated genetical alterations or extracardiac malformations in the univariate analysis, such as a lower gestational age or unbalanced atrioventricular canal diagnosis, and low birth weight <2.5 kg in the univariate analysis. In contrast, in the

multivariate analysis, only low birth weight and extracardiac malformations were considered as risk factors for mortality.<sup>11</sup> Others found other variables that conditioned an early mortality, such as the body surface area at the moment of intervention, the size of the ascending aorta, the function of the left ventricle, and the origin of the pulmonary flow (fistula vs ventriculopulmonary tube). Among the variables related to the surgical procedure, only the duration of ECC was an independent risk factor.<sup>12</sup> There is even a evaluation of preoperative risk scale published, in which a series of variables are scored such as the ventricular function, the presence of tricuspid regurgitation, the diameter of the ascending aorta, the characteristics of flow through an atrial septal defect and the age at the moment of intervention. A score of  $\geq 7$  points to a low probability of death, while a score  $< 7$ , indicates the opposite.<sup>13</sup>

In our series, though mortality was higher in the group with mitral and aortic atresia (Table 4), as well as in children with an aortic diameter of  $< 5$  mm (Table 5), these anatomical variations, though noticeable, were not statistically associated to mortality. For this motive, and coinciding with the findings of previous studies,<sup>11-13</sup> they have not been considered as prognostic factors for early mortality.

We did not find significant differences between the mortality in the PICU and the surgical technique employed (groups 1 and 2) (Table 7), probably due to the low number of patients currently included in group 2. Other authors, nonetheless, have found a difference<sup>4,12</sup> and promote the use of the tube from the left ventricle to the pulmonary artery because of the good results so far obtained. In our series of cases, of the 20 children that died in the first group, 8 did so in the operating room because it was impossible to take them off ECC (Table 7). Because of this, some authors support the systematic use of ventricular assist devices in the operating room and during the patients stay in the PICU, arguing that it improves cardiac output and therefore the survival, of these patients.<sup>14</sup> Since its introduction into our hospital in 2003, 12 children have

been intervened and none of them has died in the operating room (Table 2), indicating that they tolerate better the process of weaning from the ECC.

In a similar manner to other authors,<sup>12</sup> we have also proven that time on ECC is a risk factor for early mortality in the PICU, in the sense that the longer the time on ECC, the most likely the patient will die. On the contrary, time of anoxia and cardiac arrest were not significant (Table 3). Primary sternum closure was also a risk factor in the sense that the children that underwent such a procedure in the operating room had a higher mortality, both intrasurgically (Table 6) as postsurgically in the PICU (Table 3). Based on this finding, we propose the systematical deferred closure of the sternum in all of the children intervened for this cardiopathy, avoiding in this manner cardiac compression and low output that is the result of closing the thorax.

All of our patients underwent hyperfiltration during their time on ECC, without any complications, and without the necessity of suspending it owing to hemodynamic instability. Its use has demonstrated in other studies an improvement in hemodynamics, reflected as a reduction in the heart rate and an incremented in the systolic and diastolic pressure.<sup>15</sup>

In relation to the surgical technique, up until some years ago some authors<sup>16</sup> reconstructed the aortic arch without a homograft; in this way, it was thought, long term growth of the new aorta was benefited without any residual stenosis. In our hospital the reconstruction without a homograft was performed in as a first option in 30 patients (71%) and with it in (29%), who were those in which the first technique could not be done. Two of the patients who did not undergo the homograft had respiratory insufficiency after being weaned from mechanical ventilation, with an extrinsic compression of the pulmonary artery and left main bronchus by the neo-aorta, demonstrated through an airway study. This experience has made us consider the possibility of using a homograft in a systematic manner, as is being done by other authors with good results,<sup>12</sup> to elevate the position of the aortic arch and reducing the possibility of compressing adjacent structures.

Of the 34 children admitted to the PICU after the intervention, 19 died, which constitutes early hospital mortality in the PICU of 55.9% (Table 2). This mortality can be considered high if we compare it with that obtained by other authors.<sup>7,10</sup> As mentioned previously, the Norwood procedure was started in our hospital in 1991 and the first 3 cases were published in 1994.<sup>5</sup> Having operated 42 patients in 15 years has not allowed us to acquire sufficient experience yet. In a 12-year period, the Birmingham Children Hospital (England) intervened 333 patients, with a current mortality of 16%.<sup>12</sup> The concentration of patients in a reference center of our autonomous community will allow the intervention of more patients, which will let us gain more experience and, in consequence, increase survival. Another possible

cause that could justify mortality is the elevated proportion of children in our series with unfavorable anatomy, such as aortic and/or mitral atresia, and a reduced aortic valve diameter (Tables 4 and 5).

In relation to the analysis of the collected variables during postsurgery, children who died had higher Vd/Vt values than survivors, as a consequence or a more compromised pulmonary flow. Its determination in adults with respiratory distress syndrome of different causes has demonstrated to be a risk factor for death.<sup>17</sup> The P<sub>v</sub>CO<sub>2</sub> and EtO<sub>2</sub> values were also higher in the children that died, as a consequence of a low cardiac output and the slowing of tissue perfusion. Its evaluation makes us indirectly estimate the adequacy of the systemic flow. Differences found in P<sub>v</sub>CO<sub>2</sub>, EtO<sub>2</sub>, and Vd/Vt, between both groups, did not have any statistical value, probably due to the small sample size. Their determination has the advantage that it can be carried out easily with the common monitorization of PICU. The study of these variables is undergoing with the objective of carrying out a new analysis when the size of the sample is higher.

Medical treatment was based on a strategy of pharmacological reduction of the pulmonary and systemic vascular resistance. This way we minimized the fluctuations in the systemic and pulmonary resistance and maintained a stable circulatory and respiratory state.<sup>18</sup> The increase in systemic resistance in these children increases the myocardial workload and oxygen consumption, which explains sudden and unexpected death that is present in these patients.<sup>19</sup> For that we employed, as previously described in the methods section, nitric oxide and milrinone, the former as an exclusively pulmonary vasodilator and the latter as a pulmonary and systemic vasodilator.

Due to the advancements in medical and surgical treatments, survival of these children in the past years has increased, but they must be intervened 3 times throughout their life, with the risk inherent to this. The future, though still a ways off, will probably be the undertaking of a single procedure in the catheterism lab.<sup>20</sup>

## CONCLUSIONS

We have not demonstrated an increase in survival of children in the PICU who underwent surgery using the Sano-modified of the classic Norwood technique.

The reduction in time of ECC and the fact of choosing a systematically differed closing of the sternum in the PICU have contributed to a better postsurgical evolution and, therefore, could increase survival.

## ACKNOWLEDGMENT

We wish to express our thanks to all of the professionals of our hospital who are related to cardiology, as well as to the Asociación de Padres Corazón y Vida for their enthusiasm and the continued work developed in the caring of these children.

## REFERENCES

- Norwood WI, Kirklin JK, Sanders SP. Hypoplastic left heart syndrome: experience with palliative surgery. *Am J Cardiol.* 1980;45:87-91.
- Williams DL, Gelijns AC, Moskowitz AJ, Weinberg AD, Ng JH, Crawford E, et al. Hypoplastic left heart syndrome: valuing the survival. *J Thorac Cardiovasc Surg.* 2000;119:720-31.
- Mahle W, Spray TL, Wernovsky G, Gaynor JW, Clark III BJ. Survival after reconstructive surgery for hypoplastic left heart syndrome. A 15-year experience from a single institution. *Circulation.* 2000;102 Suppl III:136-41.
- Sano S, Ishino K, Kawada M, Arai S, Kasahara S, Asai T, et al. Right ventricle-pulmonary artery shunt in first-stage palliation of hypoplastic left heart syndrome. *J Thorac Cardiovasc Surg.* 2003;126:504-10.
- León Leal JA, García Hernández JA, Romero Parreño A, Santos de Soto J, Álvarez Madrid A, Grueso Montero J, et al. Síndrome del corazón izquierdo hipoplásico. Iniciación de un programa terapéutico. *Rev Esp Cardiol.* 1994;47:565-7.
- Maher KO, Pizarro C, Gidding SS, Januszewska K, Malec E, Norwood W, et al. Hemodynamic profile after the Norwood procedure with right ventricle to pulmonary artery conduit. *Circulation.* 2003;108:782-4.
- McGuirk SP, Griselli M, Stumper OF, Rumball EM, Miller P, Dhillon R, et al. Staged surgical management of hypoplastic left heart syndrome: a single institution 12 year experience. *Heart.* 2006;92:364-70.
- Rumball EM, McGuirk SP, Stumper O, Laker SJ, de Giovanni JV, Wright JG, et al. The RV-PA conduit stimulates better growth of the pulmonary arteries in hypoplastic left heart syndrome. *Eur J Cardiothorac Surg.* 2005;27:801-6.
- Cua CL, Thiagarajan RR, Taeed R, Hoffman TM, Lai L, Hayes J, et al. Improved interstage mortality with the modified Norwood procedure: a meta-analysis. *Ann Thorac Surg.* 2005;80:44-9.
- Pizarro C, Mroczek T, Malec E, Norwood WI. Right ventricle to pulmonary artery conduit reduces interim mortality after stage I Norwood for hypoplastic left heart syndrome. *Ann Thorac Surg.* 2004;78:1959-63.
- Stasik CN, Goldberg CS, Bove EL, Devaney EJ, Ohve RG. Current outcomes and risk factors for the Norwood procedure. *J Thorac Cardiovasc Surg.* 2006;131:412-7.
- McGuirk SP, Stickley J, Griselli M, Stumper OF, Laker SJ, Barron DJ, et al. Risk assessment and early outcomes following the Norwood procedure for hypoplastic left heart syndrome. *Eur J Cardiothorac Surg.* 2006;29:675-81.
- Checchia PA, McGuire JK, Morrow S, Daher N, Huddleston C, Levy F. A risk assessment scoring system predicts survival following the Norwood procedure. *Pediatr Cardiol.* 2006;27:62-6.
- Underleider RM, Shen I, Yeh T, Schultz J, Butler R, Silberbach M, et al. Routine mechanical ventricular assist following the Norwood procedure—improved neurologic outcome and excellent hospital survival. *Ann Thorac Surg.* 2004;77:18-22.
- Gaynor JW, Kuypers M, van Rossem M, Wernovsky G, Marino BS, Tabbutt S, et al. Haemodynamic changes during modified ultrafiltration immediately following the first stage of the Norwood reconstruction. *Cardiol Young.* 2005;15:4-7.
- Ishino K, Stumper O, de Giovanni JV, Silove ED, Wright JG, Sethia B, et al. The modified Norwood procedure for hypoplastic left heart syndrome: early to intermediate results of 120 patients with particular reference to aortic arch repair. *J Thorac Cardiovasc Surg.* 1999;117:920-30.
- Nuckton TJ, Alonso JA, Kallet RH, Daniel BM, Pittet JF, Eisner MD, et al. Pulmonary dead-space fraction as a risk factor for death in the acute respiratory distress syndrome. *N Engl J Med.* 2002;346:1281-6.
- Nakano T, Kado H, Shiokawa Y, Fukae K, Nishimura Y, Miyamoto K, et al. The low resistance strategy for the perioperative management of the Norwood procedure. *Ann Thorac Surg.* 2004;77:908-12.
- Wright GE, Crowley DC, Charpie JR, Ohve RG, Bove EL, Kulik TJ. High systemic vascular resistance and sudden cardiovascular collapse in recovering Norwood patients. *Ann Thorac Surg.* 2004;77:48-52.
- Maher KO, Gidding SS, Baffa JM, Pizarro C, Norwood WI Jr. New developments in the treatment of hypoplastic left heart syndrome. *Minerva Pediatr.* 2004;56:41-9.