Experience With the Norwood Operation for Hypoplastic Left Heart Syndrome

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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_2, the PaO_2/FiO_2 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*=.56). The only significant risk factor for in-hospital mortality was a longer cardiopulmonary bypass time (*P*=.01) and, for intraoperative mortality, primary rather than delayed sternal closure (*P*=.004). Venous pCO_2, 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.*

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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 intervinimos 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_2 venosa, la relación PaO_2/FiO_2, 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_2 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
PaO_2/FiO_2: respiratory quotient
$Vd/Vt=(PaCO_2–EtCO_2/PaCO_2)$: lung dead space
$EtO_2=(SaO_2–SvO_2/SaO_2)$: tissue oxygen extraction

INTRODUCTION

The hypoplasic 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[1] 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,[2,3] 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,[4] 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,[5] and in 2003 we incorporated the Sano modification, based on the good results obtained in others centers.[6,7] 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 hypoplasic left heart syndrome from the Hospital Vírgen 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 ≤3 mm, in 12 (28.6%) it was between 3 and 5 mm, and in 7 (16.7%) it was ≥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[1] Norwood technique, receiving a modified Blalock-Taussig fistula of different diameters, and in the other 12 (group 2) the Sano[4] 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 µg/kg/min, low doses of adrenalin that oscillated between 0.05 and 0.1 µg/kg/min, and a milrinone perfusion at 0.5 µg/kg/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 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 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 hemodynamicaly 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 (SO_2) by