Rev Esp Cardiol. 2014;67(2):114–119

Original article

Characteristics and Outcomes of Transposition of Great Arteries in the Neonatal Period

Anna Turon-Viñas,[a] Ana Riverola-de Veciana,[a,*] Julio Moreno-Hernando,[a] Joaquim Bartrons-Casas,[b] Fredy H. Prada-Martínez,[b] Javier Mayol-Gómez,[c] and José M. Caffarena-Calvar[c]

[a] Servicio de Neonatología, Unitat Integrada Hospital Clínic-Sant Joan de Déu, Esplugues de Llobregat, Barcelona, Spain
[b] Servicio de Cardiología Pediátrica, Unitat Integrada Hospital Clínic-Sant Joan de Déu, Esplugues de Llobregat, Barcelona, Spain
[c] Servicio de Cirugía Cardiovascular, Unitat Integrada Hospital Clínic-Sant Joan de Déu, Esplugues de Llobregat, Barcelona, Spain

Article history:
Received 27 January 2013
Accepted 27 June 2013
Available online 23 November 2013

Keywords:
Congenital heart disease
Transposition of the great arteries
Neonatal cardiac surgery
Arterial switch operation

ABSTRACT

Introduction and objectives: Transposition of the great arteries is a prevalent congenital heart defect with a high survival rate and a good long-term outcome, especially if managed with early surgical intervention during the neonatal period. In this study, our main objective was to describe patient characteristics and outcomes and to identify possible predictors of early and long-term morbidity and mortality.
Methods: Retrospective analysis through review of clinical and surgical charts of patients with transposition of the great arteries admitted to the service of neonatology during 2000-2011.
Results: The study included 136 patients; 119 of them had undergone corrective surgery during the neonatal period. Patients were divided into 3 groups: group I, 81 cases of isolated transposition; group II, 24 cases with ventricular septal defect; and group III, 31 with "complex" transposition of the great arteries. The overall postoperative survival was 96.7% (115 of 119 patients); no patients from group I died after surgery. Duration of surgery, intubation, inotropic treatment, and length of stay were higher in patients in groups II and III. The overall survival rate after an average of 6 years of follow-up was 90.4% 123 of 136 patients, with no deaths after discharge in group I. The most frequent residual defect during cardiac follow-up was supravalvular pulmonary stenosis, in 33 of 113 patients that had follow-up data.
Conclusions: In our study, the survival rate was high in patients with transposition of great arteries and especially in those undergoing arterial switch. The number of subsequent residual heart defects was low.

Características y evolución de la transposición de grandes vasos en el periodo neonatal

Palabras clave:
Cardiopatía congénita
Transposición de grandes vasos
Cirugía cardiaca neonatal
Switch arterial

RESUMEN

Introducción y objetivos: La transposición de grandes vasos es una cardiopatía congénita frecuente, con alta supervivencia y escaso número de secuelas, especialmente si se interviene precozmente, en el periodo neonatal. Nuestro objetivo es describir las características y la evolución de los pacientes afectados y determinar posibles factores pronósticos de morbimortalidad precoz y a largo plazo.
Métodos: Estudio retrospectivo mediante revisión de historias clínicas y base de datos de los pacientes con transposición de grandes vasos ingresados entre los años 2000 y 2011.
Resultados: Ingresaron 136 pacientes afectados de transposición de grandes vasos, de los que 119 se sometieron a cirugía correctora durante el periodo neonatal. Los pacientes se dividieron en tres grupos: grupo I, 81 transposición de grandes vasos simple; grupo II, 24 con comunicación interventricular concomitante, y grupo III, 31 transposiciones «complejas». La supervivencia postoperatoria fue de 96,7% (115 de 119 pacientes), aunque ningún paciente del grupo I falleció. La duración de la cirugía, la intubación y el uso posquirúrgico de inotrópicos y el tiempo de ingreso fueron mayores para los pacientes de los grupos II y III. Tras una media de 6 años de seguimiento, el 90,4% (123 de 136) de los pacientes estaban vivos. La lesión residual más frecuente en el seguimiento fue la estenosis supravalvular pulmonar en 33 de 113 pacientes seguidos.
Conclusiones: En nuestro estudio, la supervivencia en el conjunto de pacientes con transposición de grandes vasos, y especialmente los sometidos a *switch* arterial, es alta. Los pacientes con formas más complejas tienen más complicaciones hospitalarias, pero no tras el seguimiento.

* Corresponding author: Servicio de Neonatología, Hospital Sant Joan de Déu, Pg. St. Joan de Déu 2, 08950 Esplugues de Llobregat, Barcelona, Spain.
E-mail address: ariverola@hsjdbcn.org (A. Riverola-de Veciana).

1885-5857/$ – see front matter
http://dx.doi.org/10.1016/j.rec.2013.06.017

Abbreviations

AS: arterial switch
DORV: double outlet right ventricle
ECC: extracorporeal circulation
TGA: transposition of the great arteries
VSD: ventricular septal defect

INTRODUCTION

Congenital heart defects are the most common major congenital malformations. They are an important cause of morbidity and mortality in the neonatal period, occurring in 4-10/1000 live births.[1]

In the last three decades, there has been increased survival in these patients, mainly due to advances in diagnostic fetal and neonatal echocardiography, the extensive use of prostaglandin E1, and technological progress in cardiac surgery units.[2]

Unrepaired critical heart defects can result in progressive and irreversible secondary damage to various organs. Reconstructive surgery in this period can reduce mortality and prevent secondary damage.[3]

Transposition of the great arteries (TGA) accounts for 5% to 7% of congenital heart disease. In recent decades, TGA has been studied with more precision. Van Praagh et al.[4] have clarified the underlying anatomy and include TGA in conotruncal heart defects that include malposition of the great arteries. Double outlet right ventricle (DORV), double outlet left ventricle, and anatomically corrected transposition are also included in this group.[4]

About half of the patients with TGA have no other cardiac malformations, and are designated as isolated TGA.[4–6] The remaining cases are designated as complex TGA, that is, TGA accompanied by other malformations.

The main treatment for isolated TGA is surgical correction during the neonatal period.[7] Several surgical techniques are available, but the technique of choice is the arterial switch (AS) operation, described by Jatene et al. in 1975.[8] High survival rates have been reported, with few medium- or long-term sequelae.[9–11] The aims of this study were:

- To determine the outcome of neonates with TGA who underwent AS during the study period.
- To describe differences in the outcome of neonates with isolated TGA, transposition with ventricular septal defect (VSD), or complex transposition accompanied by other malformations who underwent AS.
- To determine the prognostic factors of morbidity and mortality in neonates who underwent a simple AS procedure or AS combined with another surgical procedure.
- To determine the long-term cardiac outcome of patients with TGA.

METHODS

Study Population

This retrospective study consecutively included all patients admitted to the service of neonatology with a diagnosis of TGA. The study period was from April 2000 to December 2011. During this period, 7935 patients were admitted to the service of neonatology. Of these patients, 781 (10.3%) were diagnosed with congenital heart disease, including 136 (17.4%) with isolated or complex TGA; 59 (43.6%) of these were intrahospital transfers (from the maternity unit) and 77 (56.6%) were referred from other centers.

Mean birth weight was 3227 (506) g and mean gestational age was 39 (1.7) weeks; 102 (75%) were boys and 34 (25%) were girls.

Patients and Study Groups

The patients were divided into 3 groups according to echocardiographic findings: group I, 81 neonates with isolated TGA (59%); group II, 24 neonates with TGA and VSD (18%), and group III, 31 neonates with "complex" TGA treatable by AS. The latter group consisted of 12 patients with DORV and transposed arteries, 11 patients with aortic coarctation and/or hypoplastic aortic arch, and 8 patients with pulmonary stenosis and VSD (23%).

Echocardiographic Study

In all cases, congenital heart defects were identified using 2-dimensional color Doppler echocardiography. The heart defects were classified according to the recommendations of the International Nomenclature and Database Conference for Pediatric Cardiac Surgery.[12] In patients with more than one heart defect, the defect with the greatest hemodynamic effect was chosen.

Initial Management of Patients

Following hospital protocol, pregnant women with a diagnosis of fetal congenital heart disease were referred to prenatal monitoring and early neonatal management. After birth, central venous cannulation was immediately performed for the administration of prostaglandin E1. Within the first 24 h of life, a Rashkind balloon atrial septostomy procedure was performed in patients with isolated TGA and restrictive foramen ovale. Corrective surgery was scheduled in the first weeks of life.

Surgical Technique

Corrective surgery was performed with extracorporeal circulation (ECC) and hypothermia according to the AS technique described by Jatene et al.[8] If present, VSD was corrected using a heterologous pericardium patch during the same procedure. Patients with DORV and TGA were treated by AS and VSD closure whenever it was possible to maintain continuity between the ventricles and the great arteries. Aortic coarctation was corrected by coarctectomy with end-to-end anastomosis extending to the inferior aspect of the aortic arch.

An atrial switch (Senning procedure) was performed when needed, rerouting deoxygenated vena caval blood to the pulmonary artery and oxygenated blood to the aorta. The coronary artery pattern at the time of surgery was described according to the Leiden classification.[13]

Early Postoperative Treatment

Following surgery, the patients were readmitted to the service of neonatology, where they were treated according to standard guidelines. After the AS procedure, inotropic infusion was begun with milrinone and dopamine, adjusting doses according to clinical response. All patients received analgesia with fentanyl and mechanical ventilation. Extubation was scheduled within 24 to 48 h, providing there were no respiratory complications (pulmonary edema, atelectasis, pneumonia).