Journal of the American College of Cardiology
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Published by Elsevier Inc.

Vol. 46, No. 7, 2005
ISSN 0735-1097/05/$30.00
doi:10.1016/j.jacc.2005.06.063

Long-Term Results After Primary One-Stage Repair of Transposition of the Great Arteries and Aortic Arch Obstruction

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OBJECTIVES The study was designed to evaluate perioperative and late results after primary, single-stage arterial switch operation (ASO) associated with aortic arch obstruction repair. Outcome of patients with more than five years of follow-up were analyzed.

BACKGROUND The treatment of patients with transposition of the great arteries, or other forms of ventriculoarterial discordance suitable for an ASO, with coexisting arch obstruction is a difficult task. Single-stage repair has become the treatment of choice at many institutions, but large series with long-term results are seldom reported.

METHODS Between 1990 and 1998, a primary operation including aortic arch repair through a midline sternotomy was performed in 38 patients. The relief of arch obstruction was accomplished during a period of hypothermic circulatory arrest, employing a wide pericardial patch to enlarge the inner curvature of the entire arch in most patients.

RESULTS There were nine (24%) hospital deaths. None could be directly related to aortic arch repair, but additional risk factors for an ASO were common (right ventricular hypoplasia, complex coronary anatomy, uncommon relationship between the great vessels or severe pulmonary hypertension). There were no late deaths. Four patients required cardiac reoperation, whereas three underwent successful treatment of recurrent coarctation with balloon angioplasty.

CONCLUSIONS Infants with ventriculoarterial discordance and aortic arch obstruction represent a high-risk subgroup of candidates for an ASO. Despite a non-negligible operative mortality, single-stage primary repair represents the treatment of choice, and follow-up of operative survivors is favorable. Pericardial patch enlargement is a reliable technique for arch obstruction repair. (J Am Coll Cardiol 2005;46:1331–8) © 2005 by the American College of Cardiology Foundation

The association between transposition of the great arteries (TGA) and aortic arch obstruction is relatively infrequent, especially in the absence of a ventricular septal defect (VSD). When this combination occurs, the natural history is very poor and the treatment constitutes a difficult task (1–5). Hypoplasia of the aortic arch is more commonly encountered in specific forms of ventriculoarterial discordance, being reported in up to 50% of cases in some subgroups of double outlet right ventricle (RV), especially in the presence of a subpulmonary VSD (Taussig-Bing anomaly) (6). Historically, the approach to such complex malformations favored a two-stage treatment, including primary coarctation repair with pulmonary artery banding, followed by intracardiac repair at a later date (5–7). During the past two decades, a one-stage procedure has gradually become the treatment of choice to avoid the detrimental effects of pulmonary artery banding on the left ventricle and neoaortic root and valve (8,9).

Our group has adopted this approach since 1990. This retrospective study includes all infants who underwent single-stage anatomic repair of TGA complexes with aortic arch obstruction at the Hôpital Laënnec in Paris between July 1990 and June 1998, and describes perioperative and medium-to-long-term results.

METHODS

Patients. Between July 1990 and June 1998, 38 consecutive patients underwent an arterial switch operation (ASO) with concomitant repair of aortic arch obstruction. Most patients (31 of 38, 82%) underwent operation during the first month of life. Intravenous prostaglandin infusion to maintain patency of the ductus arteriosus and percutaneous atrial septostomy (Rashkind maneuver) were necessary in most neonates to prevent arterial desaturation. Except for two patients with type B interrupted aortic arch, diffuse arch hypoplasia was invariably present, with the area of most severe narrowing located immediately after the origin of the left subclavian artery. A localized coarctation "ridge" at the aortic isthmus was found in about three-quarters of the cases. Patients were divided into three subgroups, according to coronary anatomy: a first group included infants with Yacoub's type A anatomy (normal pattern) (10), a second group comprised type D and E coronary arteries (circumflex artery originating from the right coronary ostium, and right coronary artery originating from the left ostium, respectively), and a third group included all other more complex patterns. The presence of specific high-risk situations,

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Manuscript received December 9, 2004; revised manuscript received June 10, 2005, accepted June 21, 2005.

JACC Vol. 46, No. 7, 2005
October 4, 2005:1331–8

Abbreviations and Acronyms

ASO = arterial switch operation
CI = confidence interval
HCA = hypothermic circulatory arrest
PHT = pulmonary hypertension
RV = right ventricle/ventricular
RVOTO = right ventricular outflow tract obstruction
TGA = transposition of the great arteries
VSD = ventricular septal defect

namely, a single or paracommissural coronary ostium, an intramural coronary, or a course between the great vessels of either coronary artery, was also analyzed separately. The two latter conditions were distinguished because the proximal segment of two of four coronary branches coursing between the great vessels was tangential to the aortic wall but not truly intramural (4,11,12). The majority of patients had an associated VSD that was often related to a variable degree of conal septal malalignment. Consequently, some degree of right ventricular outflow tract obstruction (RVOTO) was present in about a quarter of the cases, whereas a perimembranous VSD was more common in TGA with near-anteroposterior great vessels. Relative hypoplasia of the ascending aorta was always present when compared with the pulmonary trunk and determined an important mismatch between the two great vessels, the pulmonary artery diameter being 25% to 30% (or more) greater than the aorta. Also, the relationship between the great vessels varied widely, with almost one-half of the patients showing a non-anteroposterior anatomy. An unbalanced anatomy with a smaller than normal RV is more common in TGA with arch obstruction (2,4,13). In such instances, RV hypoplasia was retrospectively defined when the tricuspid valve annulus diameter measured two standard deviations below normal values. Demographic and preoperative characteristics are listed in Table 1.

Table 1. Preoperative Characteristics of the Study Population (n = 38)

Characteristic	Number of Patients	%
Girl/boy	10/28	
Age at operation (days)	29 ± 50 (3–251)	
Weight at operation (kg)	3.26 ± 0.54 (2.10–4.86)	
Prostaglandin infusion	28	74
Atrial balloon septostomy	22	58
Active infection at operation	6	16
Aortic arch anatomy		
Diffuse aortic arch hypoplasia	36	95
Localized aortic coarctation	27	71
Interrupted aortic arch	2	5
Coronary anatomy		
Type A	23	60
Type D or E	8	21
Other	7	18
Single coronary ostium	3	8
Paracommissural coronary ostium	2	5
Coronary between the great vessels	4	10
Intramural coronary artery	2	5
VSD	30	79
Malalignment	15	5
Perimembranous	11	3
Other	4	1
RVOTO	10	26
Valvular	6	6
Subvalvular	3	3
Both	1	1
Great vessels' relationship		
Antero-posterior	21	55
Oblique	5	13
Side-by-side	11	29
Leftward anterior aorta	1	3
Hypoplastic RV	9	24
Bicuspid pulmonary valve	2	5
Dysplastic tricuspid valve	1	3
Situs viscerum inversus	2	5

RV = right ventricle; RVOTO = right ventricular outflow tract obstruction; VSD = ventricular septal defect.

Surgery. All patients underwent surgery through a median sternotomy. After the institution of cardiopulmonary bypass, always using a single arterial cannula in the ascending aorta, the aorta was cross-clamped at 28°C core temperature during systemic cooling and the myocardium protected according to a modified Buckberg protocol (antegrade warm induction, cold infusion every 20 min, and controlled warm aortic root reperfusion). The ascending aorta and pulmonary trunk were then transected and the Lecompte maneuver performed in all cases.

After reaching 18°C, extracorporeal perfusion was stopped and the aortic arch reconstructed under hypothermic circulatory arrest (HCA). Since November 1996, all patients but one underwent selective antegrade cerebral perfusion through the innominate artery during HCA. An extended resection and end-to-end anastomosis was performed in nine patients, mainly in the early period. In five of nine instances, however, associated patch enlargement of the ascending aorta and proximal arch to overcome the mismatch between the arterial trunks was unavoidable. As a consequence, patch enlargement became the technique of choice for aortic arch repair, extending from below the insertion of the ductus arteriosus to the transection line in the ascending aorta. This also applied to infants with interrupted aortic arch, who were the only patients to undergo resection and end-to-end anastomosis after 1995. Twenty-nine patients underwent patch repair alone, employing glutaraldehyde-treated autologous pericardium in most cases.

After completion of the aortic arch repair, systemic arterial perfusion was resumed, and the ASO performed (14). The VSD patches were sutured in place either through the right atrium and tricuspid valve or through the great arteries, or both, always avoiding a right ventriculotomy. The conal septum was excised in three infants with double outlet RV and RVOTO (subaortic stenosis). The Rashkind atrial septal defect was only loosely closed with one or two separate stitches, with the aim to leave a restrictive interatrial communication in patients likely to develop perioperative pulmonary hypertension (PHT). A patch of fresh