The Journal of THORACIC AND CARDIOVASCULAR SURGERY

SURGERY FOR CONGENITAL HEART DISEASE

SINGLE-STAGE REPAIR OF AORTIC ARCH OBSTRUCTION AND ASSOCIATED INTRACARDIAC DEFECTS WITH PULMONARY HOMOGRAFT PATCH AORTOPLASTY

Christo I. Tchervenkov, MD
Stephen A. Tahta, MD
Luc Jutras, MD
Marie J. Béland, MD

Objective: Intracardiac malformations associated with coarctation and aortic arch hypoplasia have traditionally been repaired in 2 stages, with a high mortality rate. We review our experience with single-stage biventricular repair of intracardiac defects associated with aortic arch hypoplasia by means of pulmonary homograft patch aortoplasty. **Methods:** Between October 1988 and October 1997, 39 of 40 consecutive patients underwent single-stage biventricular repair for aortic arch obstruction and associated intracardiac defects. The median age at operation was 17 days and the mean weight was 3.71 ± 1.09 kg. Nineteen patients had either dextrotransposition of the great arteries or the Taussig-Bing anomaly. Sixteen patients had multiple left-sided obstructive lesions (2 cases of critical aortic stenosis, 3 of subaortic stenosis and ventricular septal defect, and 11 of hypoplastic left heart complex). One patient had an associated complete atrioventricular septal defect. Four patients had only an associated ventricular septal defect. Through a median sternotomy, the hypoplastic aortic arch was enlarged with a pulmonary homograft patch in 36 patients. In 4 patients an extended end-to-end anastomosis was performed. **Results:** There were 2 early deaths (5%) and 2 late deaths (5%). One late death was not cardiac related.

From the Divisions of Cardiovascular Surgery and Cardiology, The Montreal Children's Hospital, McGill University, Montreal, Quebec, Canada.

Read at the Seventy-eighth Annual Meeting of The American Association for Thoracic Surgery, Boston, Mass, May 3-6, 1998.

Received for publication May 8, 1998; revisions requested June 11, 1998; revisions received Aug 5, 1998; accepted for publication Aug 10, 1998.

Address for reprints: Christo I. Tchervenkov, MD, Director, Cardiovascular Surgery, The Montreal Children's Hospital, 2300 Tupper St, Rm C-827, Montreal, Quebec, Canada H3H 1P3.

J Thorac Cardiovasc Surg 1998;116:897-904

Copyright © 1998 by Mosby, Inc.

0022-5223/98 \$5.00 + 0 12/6/93762

The mean follow-up time was 36 months (range 1 month-9 years). The recoarctation rate after pulmonary homograft patch aortoplasty was 8.3%, but after exclusion of those patients with associated left-sided obstructive lesions this decreased to 0%. No aneurysm formation in the aorta has occurred. The actuarial survival at 8 years is 89% $\pm$ 10%.

Conclusions: Single-stage biventricular repair of aortic arch obstruction and associated intracardiac defects can achieve excellent survival. We recommend pulmonary homograft patch aortoplasty because it achieves complete relief of anatomic afterload with a tension-free anastomosis and low incidence of recoarctation. (J Thorac Cardiovasc Surg 1998;116:897-904)

Aortic arch obstruction with associated intracardiac defects has always presented a formidable surgical challenge. The traditional 2-stage approach of initial coarctation repair and palliation followed by subsequent intracardiac repair has been associated with a high mortality rate.¹⁻⁴ A few centers report improved survival with a single-stage repair of the intracardiac defect and reconstruction of the hypoplastic aortic arch almost exclusively through the extended end-to-end anastomosis technique.^{1,5-7} In this report we review our results during a 9-year period with a single-stage repair of aortic arch hypoplasia and associated intracardiac defects with pulmonary homograft aortoplasty.

Patients and methods

Between October 1988 and October 1997, 39 of 40 consecutive patients with aortic arch hypoplasia and associated intracardiac defects underwent a single-stage biventricular repair at the Montreal Children's Hospital. There were 21 boys and 19 girls. The median age at operation was 17 days (range 4 days-2 years), with 27 neonates and all except 2 younger than 3 months. The mean weight was 3.70 ± 1.08 kg. A 10-day-old neonate underwent a 2-stage repair for a Taussig-Bing anomaly with aortic arch obstruction because of hemodynamic instability after induction of anesthesia. This patient initially underwent resection and extended end-to-end anastomosis and pulmonary artery banding through a left thoracotomy. He successfully underwent the arterial switch operation, ventricular septal defect (VSD) closure, and pulmonary artery debanding at 22 days old.

All patients were evaluated before the operation by 2-dimensional and Doppler echocardiography. Cardiac catheterization was performed in only a few cases.

All patients had aortic arch obstruction by virtue of hypoplasia with or without aortic coarctation. Patients with interrupted aortic arch were excluded from this study, as were those with a single-ventricle physiology who could not be considered for a biventricular repair. Aortic arch hypoplasia was defined as the transverse aortic arch diameter less than the weight of the baby in kilograms + 1 mm.⁵ Only 5 patients

did not have a discrete coarctation. The mean diameter of the ascending aorta for this series was 6.6 ± 1.5 mm. The proximal aortic arch (between the innominate and left carotid arteries) and the distal arch (between the left carotid and the left subclavian arteries) had mean diameters of 4.0 ± 0.8 mm and 3.1 ± 0.7 mm, respectively. The aortic isthmus (between the left subclavian artery and the ductus arteriosus) had a mean diameter of 2.2 ± 0.7 mm.

The heterogeneous set of associated intracardiac defects is presented in Table I. The patients were divided into 2 groups according to the presence or absence of additional left-sided obstructive lesions. Group I consisted of 24 patients without left-sided obstructive lesions, including 8 patients with subaortic stenosis and transposition of the great arteries (TGA) and VSD or the Taussig-Bing anomaly, because that region is excluded from the systemic circulation after the repair. Group II consisted of 16 patients with associated left-sided obstructive lesions, all class II or III according to Kirklin's classification.⁸ Eleven of those patients had the diagnosis of hypoplastic left heart complex, consisting of multiple hypoplastic structures in the left heart-aorta complex, including the mitral valve anulus, left ventricular cavity, left ventricular outflow tract (LVOT), aortic valve anulus, ascending aorta, and aortic arch. Inclusion criteria included presence of antegrade aortic flow and absence of intrinsic aortic valve or mitral valve stenosis, with the obstruction caused by hypoplasia only. All patients with hypoplastic left heart complex had LVOT and aortic valve diameters at least 4 SD below the normal mean. They also had a mitral valve diameter and left ventricular dimensions at least 2 SD below the normal mean.

Preoperative status. Ductal dependency and administration of prostaglandin E₁ treatment were necessary in 25 patients (63%). Thirty-three patients (83%) were treated for congestive heart failure. Seventeen patients (43%) were critically ill, requiring ventilatory support and in some cases inotropic support.

Surgical technique. All patients except 1 (previously described) underwent single-stage repair through a median sternotomy with the use of deep hypothermic circulatory arrest. The often diminutive ascending aorta was carefully

Table I. Associated intracardiac defects in 40 patients with aortic arch hypoplasia (with or without coarctation)

Additional anatomic diagnosis	No.
Group I	
TGA + intact ventricular septum	3
TGA + VSD	4
TGA + VSD + SAS	4
Taussig-Bing anomaly	4
Taussig-Bing anomaly + SAS	4
VSD	4
Complete atrioventricular canal	1
Group II	
VSD + SAS	3
Critical aortic stenosis	2
Hypoplastic left heart complex*	11
+ Parachute mitral valve	4
+ VSD	4
+ Partial atrioventricular canal	1

VSD, Ventricular septal defect; SAS, subaortic stenosis.

*See text for definition.

cannulated on the rightward side near the base of the innominate artery. Single venous cannulation of the right atrium was used for patients in group II and before 1994 in the patients in group I. Since 1994, double venous cannulation was used in these latter patients to perform the intracardiac repair with cardiopulmonary bypass, reserving circulatory arrest only for the aortic arch repair. Myocardial protection was accomplished in all patients with single-dose crystalloid cardioplegic solution and ice-cold slush solution in the pericardial cavity.

The hypoplastic aortic arch repair was carried out with a pulmonary homograft patch in 36 patients, according to our previously published technique.⁹ This consisted of opening its undersurface from approximately 1.5 cm distal to the insertion of the ductus in the upper descending aorta, across the entire aortic arch and into the ascending aorta. The entire opened aorta was then enlarged with an appropriately fashioned pulmonary homograft patch. In the transposition complexes the enlargement was carried down to the transected aorta, thus correcting any size discrepancy present. In the patients with hypoplastic left heart complex, the entire ascending aorta was enlarged down to just distal to the aortic valve commissural posts. In the other patients varying lengths of the ascending aorta were enlarged according to the length and degree of hypoplasia. The resection and extended end-to-end anastomosis was used in only 4 patients, according to the technique described by the Marie-Lannelongue group.⁶

The intracardiac repairs were performed by previously described techniques for each specific malformation. Subaortic stenosis was present in 8 group I patients. In 7 patients it was resected through the ventriculotomy incision, which was then enlarged with an outflow pericardial patch. In 1 patient the severely stenotic area could not be enlarged with a transannular patch because of an abnormal coronary artery. The patient underwent a modified Damus-Kaye-Stansel/

Table II. Summary of results by group

	Group I (n = 24)	Group II (n = 16)
Median age (d)	29	9
Early deaths*	0	2 (13%)
Late deaths*	1 (4%)	1 (6%)
Recurrent aortic arch obstruction*	0	4 (25%)
Patients undergoing reoperation (no.)	2	7
Reoperations (no.)	2	10
For aortic arch obstruction	0	5
For LVOT obstruction	0	5
For other	2	2
Actuarial survival (%)	91% ± 8%	73% ± 16%
Freedom from reoperation	89% ± 8%†	27% ± 8%‡

*Number of patients with percentage in parentheses.

†At 8 years.

‡At 3 years.

Rastelli operation. The VSD was closed into the main pulmonary artery and the systemic circulation was brought over the transected proximal main pulmonary artery after enlargement of the aortic arch with a pulmonary homograft patch. Right ventricle to distal pulmonary artery continuity was established with a valved pulmonary homograft. In group II, 3 patients with VSD also had subaortic stenosis. One patient underwent resection with VSD closure. In the second patient the posterior malalignment VSD was closed through the pulmonary valve with an undersized patch anchored distally on the left ventricular aspect of the conal septum, as described by Luciani and associates.¹⁰ The third patient had severe unresectable stenosis and underwent the previously described modified Damus-Kaye-Stansel/Rastelli operation. All patients with hypoplastic left heart complex had tunnel subaortic stenosis, which was treated conservatively.

The mean cardiopulmonary bypass time was 202 ± 70 minutes and the mean circulatory arrest time was 52 ± 22 minutes. Delayed sternal closure was necessary in 16 cases.

Follow-up. All follow-up data were collected from the clinic visits to the divisions of cardiovascular surgery and cardiology. The patients have been followed up annually with physical examination, chest radiography, and echocardiography. The patients with suspected abnormalities were further assessed with cardiac catheterization or magnetic resonance imaging. Patients followed up elsewhere had data collected from parents and referring cardiologists. Mean follow-up time for all hospital survivors was 36 ± 32 months (range 1 month–9 years).

Results

There were 2 in-hospital deaths among the 40 patients (5%), both in group II. Both early deaths were in patients with hypoplastic left heart complex. One patient was unable to come off cardiopulmonary bypass because the hypoplastic left side of the heart was

unable to support the systemic circulation. Despite revision to a Norwood operation, the patient died during the operation. The other patient acquired persistent junctional ectopic tachycardia during the postoperative period, which necessitated systemic cooling. This patient died during a cardiac catheterization 14 days after operation that was performed to assess an extremely small LVOT.

Among the 38 early survivors, the median ventilatory support time was 8 days (range 8 hours–5 months). The median hospital stay was 24 days (range 6 days–8 months).

There were 2 late deaths, 1 in each group, for an overall total mortality rate of 10%. The total mortality rate in group I was 4.2%; it was 18.8% in group II. In group I the only death occurred from a hepatoblastoma at 39 months after cardiac repair (parents refused liver transplantation). In group II a late death occurred at 39 months after the original repair, after reoperation for severe tunnel subaortic stenosis. Severe pulmonary hypertension and a single right coronary artery were likely causes of the poor outcome. The actuarial survival for all patients in this series at 8 years is $89\% \pm 10\%$.

There have been 4 known cases of recurrent aortic arch obstruction; all were managed surgically. All 4 cases occurred in group II. One patient had undergone initial repair by resection and extended end-to-end anastomosis. The other 3 recurrences were after pulmonary homograft patch aortoplasty (3/36 patients, 8.3%). The overall rate of recurrent aortic arch obstruction was 10% (0% in group I and 25% in group II).

A total of 12 reoperations have been performed in 9 of the 38 early survivors. There were 2 reoperations in 2 patients in group I. One patient underwent reoperation for recurrent pulmonary stenosis and right pulmonary artery stenosis 21 months after repair of TGA, VSD, and aortic arch obstruction. The second patient was reoperated on for severe mitral regurgitation 2 months after repair of complete atrioventricular canal and aortic arch obstruction. Because of a single papillary muscle, the mitral valve cleft was not closed at the initial operation. The mitral valve was repaired by papillary muscle splitting and closure of the mitral valve cleft. Seven patients in group II required 10 reoperations. There were 5 reoperations in 4 patients for recurrent aortic arch obstruction, with no deaths. These 4 patients underwent resection and end-to-end anastomosis. One patient with a second recurrence also underwent another resection and patch aortoplasty. Four patients underwent 5 reoperations for LVOT obstruction from 12 to 39 months after the first operation. One

other patient was reoperated on at 14 months for recurrent aortic stenosis, both valvular and supra-valvular, and aortic arch obstruction. She underwent a revision of aortic valvotomy, ascending aortoplasty with a polytetrafluoroethylene patch, and resection of recurrent aortic arch obstruction with end-to-end anastomosis.

The results are summarized in Table II.

Discussion

The surgical treatment of intracardiac defects with associated aortic arch obstruction is a challenging undertaking. Until recently these patients were approached in 2 stages, with the coarctation repaired first through a left thoracotomy, often with concomitant pulmonary artery banding. The intracardiac repair was deferred to a second procedure. In the last decade a few centers have been repairing these defects in a single operation, with improved survival rates.^{1,5-7,9} In this report we present our results since 1988 with a consistent single-stage repair of intracardiac defects and aortic arch obstruction with pulmonary homograft patch aortoplasty.

Traditionally patients with coarctation of the aorta have been divided into 3 groups: isolated coarctation, coarctation with VSD, and coarctation with complex intracardiac defects. The largest multiinstitutional study, involving 27 institutions, was performed by the Congenital Heart Surgeons Society.² Among the 171 patients with isolated coarctation the total mortality rate was 13%; it was 17% for the 155 patients with coarctation and VSD. The mortality rate increased to 45%, however, for the 106 patients with coarctation and complex intracardiac defects. Of particular interest is the stratification of the coarctation with or without VSD groups with respect to the presence and number of associated additional left-sided obstructive lesions. In the absence of additional left-sided obstructive lesions, the total mortality was only 8% among 280 patients. One additional left-sided obstructive lesion (Kirklin class II) was associated with a mortality rate of 40% and the presence of 2 or more additional left-sided obstructive lesions (Kirklin class III) further increased the mortality rate to a prohibitive 88%. These dismal results are probably the reason for the recommendation that patients with class III hypoplastic left heart syndrome be subjected to a univentricular approach.⁸ It is also clear that patients with additional left-sided obstructions should be considered in the complex coarctation group and that the traditional approach of coarctation repair through left thoracotomy in these patients leads to poor results.

The single-stage repair was first reported by Pigott

and associates¹¹ for 5 patients with TGA and aortic arch obstruction. There were 4 survivors. The superiority of the single-stage approach was first demonstrated by Planché and colleagues¹ in 1993, also for patients with TGA, VSD, and aortic arch obstruction. This experience was further updated by Conte and associates⁶ for a total of 71 patients. The total mortality rate for the single-stage approach in 27 patients was only 19%, as opposed to 45% for the 2-stage approach used in the other 44 patients. Other authors have also reported improved results with the single-stage approach, with total mortality rates of around 20%.^{5,7} In 1997 we reported no early deaths and only 1 late noncardiac death among 12 patients with transposition complexes with aortic arch obstruction treated by the single-stage approach.⁹

In the largest series from a single institution on the surgical management of neonatal coarctation, consisting of 307 patients from Marie-Lannelongue in Paris, the total mortality rate for isolated coarctation was an outstanding 2%, and for coarctation with VSD it was only 6%.⁶ On the other hand, the mortality rate for the complex coarctation group (110 patients) was much higher at 40%. However, the 31 patients treated by a single-stage operation had a significantly lower mortality rate (23%) than did the 79 patients subjected to a 2-stage approach (47%).

The reason for the high mortality rate associated with the 2-stage approach may be multifactorial. The overwhelming association of neonatal coarctation with tubular hypoplasia of the aortic arch has been unrecognized until recently. Aortic arch hypoplasia was present in 81% of the patients in Conte and associate's series,⁶ with the association being highest in the group with complex intracardiac lesions (93%). Conservative repair of the coarctation in these patients would not address the aortic arch hypoplasia, leaving anatomic obstruction to be faced by the unrepaired heart. For this reason several groups, including the Marie-Lannelongue group, advocate an extended end-to-end technique of repair to a point proximal to the left common carotid artery.⁶ Although there is evidence that the tubular hypoplasia of the aorta grows after simple coarctation repair,^{12,13} growth probably does not occur fast enough to allow consistent survival of the unrepaired heart. After subsequent repair, the heart freshly operated on still has to face the residual anatomic afterload of the hypoplastic arch, decreasing survival. Furthermore, the placement of a pulmonary artery band in these patients may lead to a doubly obstructed heart. Finally, in some patients the complexity of the intracardiac malformation may be overwhelming, itself pre-

cluding survival unless repaired, despite an adequate coarctation repair. The 2-stage approach psychologically elevates the coarctation to the status of primary malformation, relegating the heart defect to a secondary status. We believe that it should be the other way around for most patients with associated intracardiac defects. The cardiac malformation should take precedence, with the coarctation being repaired simultaneously.

The controversy also extends to patients with coarctation and VSD, where the difference between the single-stage and the 2-stage approach is not as obvious. In the Congenital Heart Surgeons Society multi-institutional study,² initial repair of coarctation with concomitant pulmonary artery banding carried a mortality rate of 9%, compared with mortality rates of 21% for coarctation repair alone and 22% for single-stage repair of the coarctation and VSD. The difference in mortality rate, however did not reach statistical significance. Furthermore, although the lowest mortality rate was seen among the 34 patients who underwent coarctation repair and pulmonary artery banding, 18 of those patients had not yet undergone VSD closure by the time results were analyzed for the study.

Since 1988, we at the Montreal Children's hospital have consistently repaired cardiac defects and associated aortic arch obstruction with a single operation through a sternotomy approach. This was possible in 39 of 40 consecutive patients. The early mortality rate was 5% (2 deaths), with no deaths in group I and a 12.5% mortality rate in group II. The total mortality rate was 10% (2 late deaths, 1 in each group), with mortality rates of 4.2% in group I and 18.8% in group II. These mortality rates are much lower than those reported in the literature with a 2-stage approach for similar patients.^{1,2,4,8,13-15}

Only 4 patients in our series (10%) had a VSD and aortic arch obstruction without additional left-sided obstructive lesions. These defects were repaired simultaneously without any deaths. The single-stage approach for these patients merits further assessment, perhaps in a prospective randomized study.

Hypoplastic left heart complex is a term that we introduced at the Thirty-fourth Annual Meeting of The Society of Thoracic Surgeons in New Orleans in 1998 to denote a subset of patients with class III hypoplastic left heart syndrome who should be considered for biventricular repair.¹⁶ We believe that the biventricular approach will probably result in a better long-term prognosis than will a univentricular approach, which is based on the morphologically right ventricle as a systemic pump. Our criteria for biventricular repair are as follows: (1) antegrade flow in the ascending aorta, (2)

no intrinsic aortic or mitral valve stenosis (obstruction by hypoplasia only), and (3) adequate left ventricular function. The surgical objectives were complete relief of the extracardiac anatomical afterload, elimination of all intracardiac shunts to fully preload the left side of the heart, and a conservative approach to the hypoplastic inflow and outflow tracts of the left ventricle. This was accomplished with pulmonary homograft patch aortoplasty of the aortic arch and ascending aorta and closure of any interventricular and interatrial communications. We have observed rapid growth of the hypoplastic left heart structures, with the exception of the subaortic region.

Several techniques have been proposed through the years for repair of coarctation and aortic arch hypoplasia. Synthetic patch aortoplasty has fallen out of favor because of the risk of aneurysmal growth and sudden rupture.^{17,18} It was first proposed in 1957 to replace the circumferential suture line in the resection with end-to-end anastomosis technique for coarctation repair.¹⁹ The incidence of aneurysm formation with the synthetic patch aortoplasty, now well studied, has ranged from 1.3% to 26%.^{17,20-22}

Complete conduit reconstruction of the aorta in early life has been avoided as a result of the lack of growth potential. The subclavian flap angioplasty technique gained in popularity because of its ease of use and because it does not require extensive dissection. However, it does not address the aortic arch hypoplasia proximal to the left subclavian artery and is not a technique that can be used with the single-stage approach. Currently the most widely used technique for repair of coarctation with aortic arch hypoplasia is resection with extended end-to-end anastomosis.⁴⁻⁷ The main advantages of this technique are the resection of the coarctation shelf and abnormal periductal tissue and the augmentation of the tubular hypoplasia of the aortic arch with no foreign material. Disadvantages are the greater tension at the anastomosis and the potential for leaving relative obstruction at the proximal end. The incidence of recoarctation has varied between 4% and 20%, depending on the series and the classification of the defects.^{2,4-7}

Although a pulmonary homograft patch has been used widely in the Norwood procedure for hypoplastic left heart syndrome, its use for repair of coarctation and aortic arch hypoplasia has remained largely unreported. The only report of the use of homograft aortoplasty was by Pigott and associates.¹¹ Our almost exclusive use of pulmonary homograft patch aortoplasty in the single-stage repair was based on the following principles: (1) the aortic arch must be of adequate size after the repair,

regardless of the original diameter, (2) we must have the ability to enlarge the ascending aorta, particularly important in the group II patients with multiple left-sided obstructions, (3) there must be no tension at the anastomosis, particularly in patients undergoing the arterial switch operation with circumferential neoaortic anastomosis, and (4) it is important to correct the marked discrepancy between the proximal neoaorta and the distal aorta commonly present in patients with transposition complexes. We have used this technique in 90% of our patients since 1988. Our overall reintervention rate for recoarctation after pulmonary homograft patch aortoplasty is 8.3%, comparable to the incidence observed for the extended end-to-end technique^{2,4-7} as well as incidences of recoarctation after the Norwood procedure.²³⁻²⁵ There were no recoarctations in group I; all reinterventions occurred in the group II patients with additional left-sided obstructions. We can only speculate that the residual intracardiac obstructions may have led to a borderline cardiac output with inadequate volume and pressure loading of the aortic arch. Aneurysm formation has not been observed in our patients; however, it is too early to make definitive statements regarding this problem.

Single-stage repair of aortic arch obstruction and associated intracardiac defects has achieved excellent survival rates. Results are superior to those published for the 2-stage approach, and single-stage repair should therefore be considered the surgical approach of choice. Furthermore, the extensive enlargement of the aortic arch and ascending aorta has achieved much improved survival for the patients with additional left-sided obstructions. We recommend pulmonary homograft patch aortoplasty because it achieves complete relief of the anatomic afterload with a tension-free anastomosis and a low incidence of recoarctation.

REFERENCES

1. Planché C, Serraf A, Comas JV, Lacour-Gayet F, Bruniaux J, Touchot A. Anatomic repair of transposition of the great arteries with ventricular septal defect and aortic arch obstruction. *J Thorac Cardiovasc Surg* 1993;105:925-33.
2. Quaegebeur JM, Jonas RA, Weinberg AD, Blackstone EH, Kirklin JW. Outcomes in seriously ill neonates with coarctation of the aorta: a multiinstitutional study. *J Thorac Cardiovasc Surg* 1994;108:841-54.
3. Vogel M, Freedom RM, Smallhorn JF, William WG, Trusler GA, Rowe RD. Complete transposition of the great arteries and coarctation of the aorta. *Am J Cardiol* 1984;53:1627-32.
4. Vouhé PR, Trinquet F, Lecompte Y, Vernant F, Roux PM, Touati G, et al. Aortic coarctation with hypoplastic aortic arch. *J Thorac Cardiovasc Surg* 1988;96:557-63.
5. Karl TR, Sano S, Brawn W, Mee RB. Repair of hypoplastic or

- interrupted aortic arch via sternotomy. *J Thorac Cardiovasc Surg* 1992;104:688-95.
6. Conte S, Lacour-Gayet F, Serraf A, Sousa-Uva M, Bruniaux J, Touchot A, et al. Surgical management of neonatal coarctation. *J Thorac Cardiovasc Surg* 1995;109:663-75.
7. Sandhu SK, Beekman RH, Mosca RS, Bove EL. Single-stage repair of aortic arch obstruction and associated intracardiac defects in the neonate. *Am J Cardiol* 1995;75:370-3.
8. Kirklin JW, Barratt-Boyes BG. Coarctation of the aorta and interrupted aortic arch. In: *Cardiac surgery*. 2nd ed. New York: Churchill Livingstone; 1993. p. 1269-70.
9. Tchervenkov CI, Tahta S, Cecere R, Beland MJ. Single-stage arterial switch with aortic arch enlargement for transposition complexes with aortic arch obstruction. *Ann Thorac Surg* 1997;64:1776-81.
10. Luciani GB, Ackerman RJ, Chang AC, Wells WJ, Starnes VA. One-stage repair of interrupted aortic arch, ventricular septal defect and subaortic obstruction in the neonate: a novel approach. *J Thorac Cardiovasc Surg* 1996;111:348-58.
11. Pigott JD, Chin AJ, Weinberg PM, Wagner HR, Norwood WI. Transposition of the great arteries with aortic arch obstruction. *J Thorac Cardiovasc Surg* 1987;94:82-6.
12. Brouwer MH, Cromme-Dijkhuis AH, Ebels T, Eijgelaar A. Growth of the hypoplastic aortic arch after simple coarctation resection and end-to-end anastomosis. *J Thorac Cardiovasc Surg* 1992;104:426-33.
13. Siewers RD, Eteddgui J, Pahl E, Tallman T, del Nido PJ. Coarctation and hypoplasia of the aortic arch: will the arch grow? *Ann Thorac Surg* 1991;52:608-14.
14. DeLeon SY, Downey FX, Baumgartner NE, Ow EP, Quinones JA, Torres L, et al. Transsternal repair of coarctation and associated cardiac defects. *Ann Thorac Surg* 1994;58:179-84.
15. Ziemer G, Jonas RA, Perry SB, Freed MD, Castaneda AR. Surgery for coarctation of the aorta in the neonate. *Circulation* 1986;74(3 Pt 2):125-31.
16. Tchervenkov CI, Tahta SA, Jutras L, Béland MJ. Biventricular repair in neonates with hypoplastic left heart complex. *Ann Thorac Surg*. In press, 1998.
17. Bromberg BI, Beekman RH, Rocchini AP, Snider AR, Bank ER, Heidelberg K, et al. Aortic aneurysm after patch aortoplasty repair of coarctation: a prospective analysis of prevalence, screening tests and risks. *J Am Coll Cardiol* 1989;14:734-41.
18. Kumar A, Miller R, Finley JP, Roy DL, Gillis DA, Nanton MA. Follow-up after patch aortoplasty for coarctation of aorta. *Can J Cardiol* 1993;9:751-3.
19. Vosschulte K. Isthmusplastik zur behandlung der aorten isthmusstenose. *Thoraxchirurgie* 1957;4:443-50.
20. Venturini A, Papalia U, Chiarotti F, Caretta Q. Primary repair of coarctation of the thoracic aorta by patch graft aortoplasty: a three-decade experience and follow-up in 60 patients. *Eur J Cardiothorac Surg* 1996;10:890-6.
21. Mendelsohn AM, Crowley DC, Lindauer A, Beekman RH. Rapid progression of aortic aneurysms after patch aortoplasty repair of coarctation of the aorta. *J Am Coll Cardiol* 1992;20:381-5.
22. Del Nido P, Williams W, Wilson G, et al. Synthetic patch angioplasty for repair of coarctation of the aorta: experience with aneurysm formation. *Circulation* 1986;74(suppl):I32-6.
23. Jonas RA. Intermediate procedures after first-stage Norwood operation facilitate subsequent repair. *Ann Thorac Surg* 1991;52:696-700.

24. Farrell PE, Chang AC, Murdison KA, Baffa JM, Norwood WI, Murphy JD. Outcome and assessment after the modified Fontan procedure for hypoplastic left heart syndrome. *Circulation* 1992;85:116-22.
25. Chang AC, Farrell PE, Murdison KA, Baffa JM, Barber G, Norwood WI, et al. Hypoplastic left heart syndrome: hemodynamic and angiographic assessment after initial reconstructive surgery and relevance to modified Fontan procedure. *J Am Coll Cardiol* 1991;17:1143-9.

Discussion

Dr Claude Planché (*Paris, France*). I first congratulate you on an excellent series with remarkable results in surgical management of complex cardiac anomalies associated with aortic arch obstruction. Aortic arch obstruction includes a wide range of anatomic anomalies of various degrees of severity that should dictate the method of repair. Extended resection and end-to-end repair require extensive mobilization of the descending aorta and can be challenging, as you pointed out, when the proximal aortic arch is involved. Aortic arch reconstruction with pulmonary homograft patch is a relatively simple standardized and reproducible technique that is applicable in all forms, even those involving the ascending aorta. In some cases, however, a new aorta will consist almost entirely of prosthetic material, particularly at the level of the aortic isthmus.

It has been our experience that even pulmonary homograft can calcify. Can you comment on the technical aspect of surgical management of recoarctation in such a case? Is it possible to use balloon dilatation? In your series 5 patients did not have a discrete coarctation but underwent an aortic arch repair. Can you comment on this particular aspect?

The study confirms our experience that excellent survival can be achieved with single-stage biventricular repair of complete cardiac anomaly. However, we consider that the 2-stage management of this anomaly may be preferable in some situations, such as multiple VSD, straddling atrioventricular valves, or preoperative brain damage. Can you give your precise selection criteria for 1- versus 2-stage repair?

With respect to the group of patients with hypoplastic left ventricle, in our experience with patients with intact ventricular septum and only 1 atrial septal defect, reported on last year at the American Association for Thoracic Surgery meeting, we observed that isolated coarctation repair was generally enough to initiate the left ventricular growth; concomitant closure of the atrial septal defect led to severe left ventricular diastolic failure. In addition, when analyzing your results, we observed that the recoarctation rate was nearly 30% among the survivors of this particular group, despite an adequate repair and the growth of the left ventricle. Could you comment on these particular points?

I totally agree that use of pulmonary homograft patch is helpful in repair of the aortic arch obstruction, particularly in forms involving the ascending aorta and the proximal part of the aortic arch.

Dr Tchervenkov. We have been inspired by you in many cases, reassured that the techniques we use in Montreal are, at least we believe, the right ones.

With respect to recoarctation, the patients who required reoperation for recoarctation also required reoperation for other reasons. There were patients, usually in the group with hypoplastic left heart complex, who required reoperation at the same time for recurrent residual LVOT obstruction. I think that if we deal exclusively with an isolated recoarctation at the distal end with a nicely enlarged aortic arch, we would agree that the appropriate management would be balloon dilatation.

With respect to the 5 patients subjected to enlargement of the hypoplastic aortic arch in the absence of coarctation, this really goes along with our overall philosophy for these patients. I believe that if you leave tubular hypoplasia behind in a patient with isolated coarctation after repair with a normal heart, the normal heart will overcome the anatomic afterload and that arch will probably grow. If you have tubular hypoplasia of the aortic arch after coarctation repair in addition to intracardiac defects, however, that relative hypoplasia or that relative anatomic afterload will have to be faced by the unrepaired heart. Furthermore, leaving the tubular hypoplasia behind in a repaired heart that has to recover from the ischemic insults may also lead to problems. We have therefore taken the philosophic approach that all these patients with tubular hypoplasia with complex intracardiac malformations should undergo enlargement.

This approach was actually confirmed in 1 patient in this series whom I classified as belonging to the single-stage group. Actually this patient had a borderline aortic arch of 5 mm according to Roger Mee's criteria, the weight of the baby + 1 mm, and I thought that this arch would be adequate. After the switch operation and the VSD closure, approximately half an hour after coming off cardiopulmonary bypass, the patient had a rapid progressive deterioration to the point of near arrest. On 2 occasions resuscitation on bypass did not prevent the further episode. We decided to take the whole switch down, enlarge the aortic arch, and redo the switch. After 10 hours in the operating room and close to 4 hours of cross-clamp time, the patient came off bypass with ease and gave us no trouble afterward. This really confirmed for me that, in some patients without coarctation but with hypoplasia of the aortic arch and complex intracardiac defects, this hypoplasia has to be addressed.

With respect to the indications for the single-stage approach, I agree with you that there will be some, in my opinion rare, exceptions. We have learned today from other presentations that there are ways to handle the patients with multiple VSDs, and I think that every patient has to be treated on an individual basis. I think that some of these patients, even if they undergo palliation, are not going to do well, so we have taken the approach that we may as well go with a single-stage operation. Perhaps some patients with a difficult multiple VSD swiss-cheese septum may be excluded in our institution as well.

With respect to the question of straddling of the tricuspid valve, you have published techniques for dealing with that

straddling. I think that in many of those situations straddling, unless it is the type C severe straddling, can perhaps be handled at the same time.

With respect to the recoarctation rate among the patients with hypoplastic left heart complex, it is puzzling to us on reviewing the data that all these recoarctations happened in the group of patients with multiple left-sided obstructions. I cannot explain that. I can only advance a hypothesis that perhaps in those patients the residual hypoplasia of the left ventricle and the inflow and outflow tracts perhaps led to inadequate volume and pressure loading of the repaired aortic arch and perhaps allowed the pulmonary homograft patch to regress somewhat and not fully expand. This is something that we obviously have to observe, with increased follow-up.

Dr Frank L. Hanley (*San Francisco, Calif*). Getting back to the specifics of the arch, I think that you are stretching to try to make a physiologic argument for the recurrent arch obstruction in certain patients. One would think that if there is not enough flow to stimulate arch growth, there would be some other global manifestation of low output as well. There is an awful lot of experience around the world with this type of arch reconstruction with nonviable homograft material crossing ductal tissue, and I think that one can reliably state that no matter how well you do this operation initially there is going to be a defined incidence of recoarctation because you have nonviable tissue crossing ductal tissue. It is not unreasonable to expect that recurrence in a large series of these 2-ventricle arch repairs that would be similar to that seen after Norwood operations, for example.

There are alternative ways to repair these arches, particularly in patients with transposition and a large mismatch between the aorta and the main pulmonary artery, in hearts with Taussig-Bing syndrome and interrupted arches or coarctations, and in cases of transposition with coarctation, where you have a diffusely small ascending aorta from right above the valve all the way around the arch. We have had favorable results with transecting the aorta just above the aortic valve and then swinging the ascending aorta transversely and connecting it end to end to the descending aorta, so that the hypoplastic ascending aorta becomes a transverse aorta. After the Lecompte maneuver, the proximal main pulmonary artery is connected into that newly directed transverse arch without artificial material.

Dr Tchervenkov. I agree that there are several ways to approach this situation, and I am certainly aware of your technique for the patients with interrupted aortic arch whom you described. What we are discussing here is our technique that has been consistently applied through a 9-year period, without changing every month or every year because of an unfavorable outcome, and we are simply presenting the results of this approach during this period. It is possible that in some situations we may evolve. For example, a way to avoid leaving the coarctation shelf behind and the abnormal tissue is to simply resect that area, do a partial anastomosis, and then enlarge the undersurface with a homograft patch.