Should We Address the Neopulmonic Valve? Significance of Right-Sided Obstruction After Surgery for Transposition of the Great Arteries and Coarctation

Yasutaka Hirata, MD, Jonathan M. Chen, MD, Jan M. Quaegebeur, MD, and Ralph S. Mosca, MD

Division of Cardiothoracic Surgery, Department of Surgery, Columbia University College of Physicians and Surgeons, New York, New York

Background. **The combination of transposition of the great arteries and coarctation of the aorta (TGA/CoA) presents a surgical challenge. We have adopted a concurrent aortic arch repair and arterial switch operation with excellent results. These patients tend to have a small aortic (ie, neopulmonary) annulus. This study evaluates the significance of right-sided obstruction after single-stage repair of TGA/CoA.**

Methods. **Between May 1991 and May 2006, 53 patients with TGA/CoA or Taussig-Bing/CoA (n = 20; 38%) underwent a complete single-stage repair. Surgical technique involved the arterial switch operation and ventricular septal defect closure when present in 51 patients (96%). The aortic arch was enlarged in 36 patients (75%) with patch augmentation. Two patients (4%) underwent the augmentation of the right ventricular outflow tract (RVOT) at the first operation.**

Results. **There was one hospital death (operative mortality, 1.9%) and one late death. The difference between the preoperative aortic and pulmonary annulus was significant (6.7 vs 10.4 mm, $p < 0.001$). The peak pressure gradient across the proximal RVOT at discharge was 16 ± 16 mm Hg (range, 0 to 62 mm Hg). There have been six reoperations (11%) and four catheter interventions (7.5%) for right-sided obstruction. Freedom from reintervention/reoperation for right-sided obstruction at 1, 5, and 10 years is 95%, 87%, and 80%, respectively.**

Conclusions. **Neonatal single-stage repair for TGA/CoA achieves excellent survival without transannular patch repair at the first operation. Although some of the patients have pressure gradient across the RVOT, these lesions were amenable to reintervention with minimal morbidity.**

(Ann Thorac Surg 2008;86:1293–8)

The surgical treatment of transposition of the great arteries (TGA) has made great advances in the past 20 years. Although TGA can be repaired using the arterial switch operation with a low operative risk, the combination of TGA and coarctation of the aorta (TGA/CoA) or double -outlet right ventricle with subpulmonary ventricular septal defect CoA (Taussig-Bing/CoA) presents a surgical challenge [1–4].

We have adopted a concurrent aortic arch repair and arterial switch operation during the past 15 years, with excellent results. These patients tend to have a small aortic (ie, neopulmonary) annulus, and its significance on reintervention and reoperation is considerable. Some have suggested enlarging the right ventricular outflow at the time of the arterial switch operation [5–7]. However, the necessity of this intervention in the newborn period remains unclear. This study was undertaken to delineate the significance of right-sided obstruction after the single-stage repair of TGA/CoA and Taussig-Bing/CoA.

Material and Methods

This study received approval by the Institutional Review Board of Columbia University. We retrospectively reviewed the cardiac surgery database and catheter intervention database at the Morgan Stanley Children's Hospital of New York (MS-CHONY) for those patients with the diagnosis of TGA/CoA or Taussig-Bing/CoA who

Accepted for publication March 25, 2008.

Presented at the Forty-fourth Annual Meeting of The Society of Thoracic Surgeons, Fort Lauderdale, FL, Jan 28–30, 2008.

Address correspondence to Dr Hirata, The Division of Cardiothoracic Surgery, Columbia University College of Physicians and Surgeons, 3959 Broadway, CHN-273, New York, NY 10032; e-mail: yh2240@columbia.edu.

Published by Elsevier Inc

0003-4975/08/$34.00
doi:10.1016/j.athoracsur.2008.03.059

underwent single-stage aortic arch reconstruction with arterial switch operation between May 1991 and May 2006. The cardiac diagnosis was based primarily on two-dimensional and Doppler echocardiography.

The hospital records of all patients were reviewed for details of preoperative assessment, operative management, and hospital course. The preoperative echocardiographic recordings at presentation were reviewed to determine the location and morphology of the ventricular septal defect (VSD), and diameter of the aortic and pulmonary annuli.

The observed left- and right-sided lesions were retrospectively analyzed to validate the hypothesis that in this anomaly, the great artery anatomy is mainly associated with a size discrepancy between the pulmonary annulus and the aortic annulus, with the latter (ie, neopulmonary annulus) being much smaller than the neoaortic root.

Patients

Between May 1991 and May 2006, 53 patients with the diagnosis of TGA/CoA or Taussig-Bing/CoA underwent single-stage aortic arch repair and arterial switch at MS-CHONY. Mean birth weight was 3.2 ± 0.7 kg, and mean age at the operation was 8.7 ± 9.4 days. All patients were taken to the operating room with the intent to perform a single-stage complete repair. Most patients had an associated VSD, often with concurrent conal septal malalignment. Only 2 patients had intact ventricular septum. Relative hypoplasia of the ascending aorta was often present when compared with the pulmonary trunk. The relationship between the great vessels varied widely: 15 patients (28%) had side-by-side great arteries, and 21 (40%) had Taussig-Bing/CoA. Demographic and preoperative characteristics are listed in Table 1.

Table 1. Preoperative Characteristics of the 55 Patients

Characteristic	Value
Sex, No.	
Male	31
Female	22
Operative data, mean ± SD (range)	
Age, days	8.8 ± 9.3 (2–46)
Weight, kg	3.21 ± 0.68 (1.3–4.7)
Aortic annulus, mm	6.7 ± 1.3
Pulmonary annulus, mm	10.4 ± 1.4 mm
Coronary anatomy	
1 LCx 2 R	33 (62%)
1 L 2 RCx	5 (9%)
1 LR 2 Cx	2 (4%)
1 R 2 LCx	2 (4%)
Single coronary ostium	4 (8%)
Other	7 (13%)
VSD, No. (%)	
Subpulmonary	32 (60)
Perimembranous	6 (11)
Muscular	13 (25)
No VSD, No. (%)	2 (4)
Arch obstruction, No. (%)	
Arch hypoplasia	38 (72)
Discrete coarctation	15 (28)
Great vessel relationships, No. (%)	
Anteroposterior	38 (72)
Side-by-side	15 (28)
Taussig-Bing anomaly	21 (40)

L = left anterior descending artery; R = right coronary artery; Cx = circumflex artery; RCx = right circumflex; SD = standard deviation; VSD = ventricular septal defect.

Surgical Technique

All patients underwent median sternotomy alone. Cardiopulmonary bypass was instituted, patients were cooled to 18°C, and the aortic arch was reconstructed under deep hypothermic circulatory arrest. After resection of the coarctation shelf and ductal tissue, arch reconstruction was performed by either end-to-end anastomosis in 13 patients (25%) or patch augmentation in 40 (75%) patients, of whom 31 had homograft patch and 9 had a glutaraldehyde-treated autologous pericardial patch.

After completion of the aortic arch repair, systemic arterial perfusion was resumed, and the arterial switch operation was performed. The closure of the VSD was achieved through the original pulmonary artery (PA) in 35 patients (66%) and through the right atrium in 12 (23%). Right ventriculotomy was used for only 1 patient. Either fresh or glutaraldehyde-treated pericardium was used to reconstruct the defect in the neopulmonary root after detachment of the coronary ostia. One pantaloon shaped patch or two small patches for each ostium was used at the surgeon's discretion.

Five patients did not undergo the LeCompte maneuver to prevent compression of the left coronary artery button. The subaortic area was always inspected at the time of operation, and subaortic muscle resection was performed for 8 patients (16%). The aortic valve (ie, neopulmonic valve) was preserved as much as possible. Eventually, 1 patient underwent transannular patch repair, and 1 patient had a right ventricle-pulmonary artery conduit at the time of initial operation. Intraoperative and perioperative patient data are summarized in Table 2.

Follow-Up

Reviews of clinical records and contacts with the referring cardiologists served for collection of hospital and follow-up data. Echocardiographic reports were available for all patients. Four patients were lost to follow-up (follow-up rate, 92%), and the mean follow-up was 94 ± 64 months.

Statistical Analysis

Continuous variables were expressed as the mean ± standard deviation. The univariate association between risk factors and postoperative outcome was assessed by using either the χ^2 test for dichotomous risk factors or a t test for continuous risk factors. Multivariate analysis was performed by logistic regression model. The Kaplan-Meier method was used to estimate the probability of survival and

Table 2. Surgical Data of the Patients

Characteristic	Value No. (%) or Mean ± SD
Aortic arch repair	
Patch enlargement	40 (75)
Autologous pericardium	9 (17)
Pulmonary homograft	15 (28)
Aortic homograft	1 (2)
Cryopreserved pericardium	15 (28)
End-to-end anastomosis	13 (25)
VSD repair	
Through pulmonary valve	38 (72)
Through right atrium	12 (23)
Through right ventricle	1 (2)
Neo-PA sinus reconstruction	
2 patches	44 (83)
1 patch	9 (17)
RVOT procedure	
Transannular patch repair	1 (2)
RV-PA conduit	1 (2)
Subaortic muscle resection	8 (16)
CPB, total time min	211 ± 39
Aortic cross-clamp time, min	130 ± 31
DHCA time, min	40 ± 17

CPB = cardiopulmonary bypass; DHCA = deep hypothermic circulatory arrest; PA = pulmonary artery; RV = right ventricle; RVOT = right ventricular outflow tract; SD = standard deviation.

freedom from adverse outcome event. Analyses were carried out with JMP software (SAS Institute, Cary, NC).

Results

Survival

One hospital death (operative mortality, 1.9%) due to sudden AV block occurred in a 1.3-kg girl with Taussig-Bing/CoA with severe intrauterine growth restriction. She had multiple dysmorphic features including omphalocele, ear tags, and vertebral abnormalities. She was doing well postoperatively, but sudden AV block developed on postoperative day 2. Although the chest was opened and a pacing wire was placed, she did not respond to pacing. One late death occurred in a patient who underwent full repair for Taussig-Bing/CoA and was discharged 1 month later. He had been doing well, but came in 5 months postoperatively with respiratory syncytial virus infection. His respiratory condition deteriorated, and he died 1 month later.

Reintervention

There were eight reoperations and six catheter interventions for 11 patients (20%) during this period. Actuarial freedom from overall reintervention was 92% (95% confidence interval [CI], 82% to 97%) at 1 year, 81% (95% CI, 69% to 90%) at 5 years, and 74% (95% CI, 59% to 85%) at 10 years (Fig 1). There was no death from reoperation or catheter-based interventions.

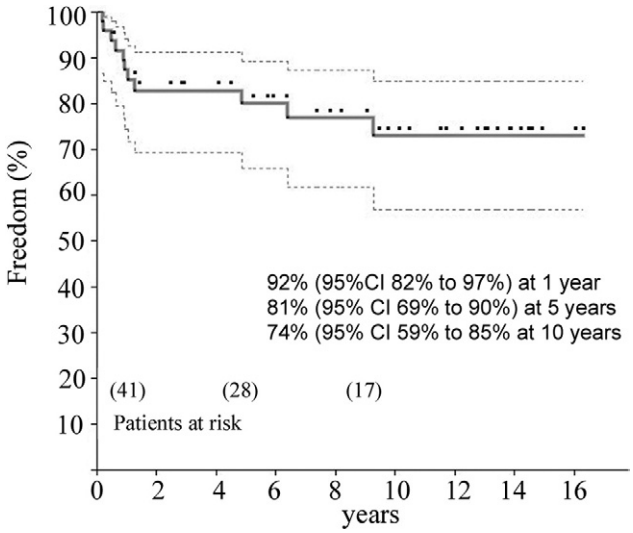

Fig 1. Freedom from all reintervention. Dashed lines represent 95% confidence limits.

Right-Sided Obstruction

The difference between the preoperative aortic and pulmonary annuli was significant (6.7 vs 10. 4 mm, $p < 0.001$). The postoperative peak pressure gradient across the right ventricular outflow tract (RVOT) at discharge was 16 ± 16 mm Hg (range, 0 to 62 mm Hg). There were six reoperations and four catheter interventions for right-sided obstruction. Actuarial freedom from reoperation or reintervention for the right-sided obstruction was 95% (95% CI, 87% to 98%) at 1 year, 87% (95% CI 76% to 94%) at 5 years, and 80% (95% CI 66% to 90%) at 10 years (Fig 2).

Four patients underwent operation for recurrent RVOT obstruction. Three patients underwent a transannular patch and 1 patient underwent infundibular muscle resection without a transannular patch. Actuarial freedom from reoperation for RVOT obstruction was 97% (95% CI,

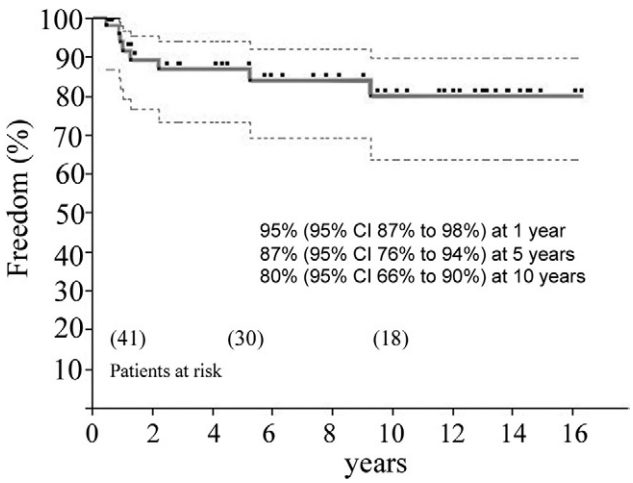

Fig 2. Freedom from reintervention for right-sided obstruction. Dashed lines represent 95% confidence limits.

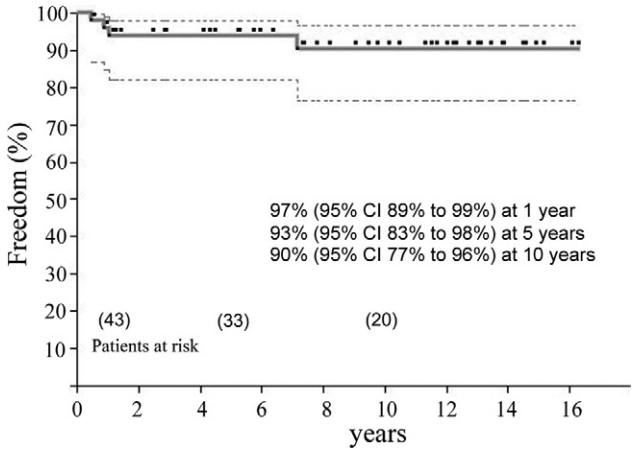

Fig 3. Freedom from reintervention for the proximal right-sided obstruction. Dashed lines represent 95% confidence limits.

89% to 99%) at 1 year, 93% (95% CI, 83% to 98%) at 5 years, and 90% (95% CI, 77% to 96%) at 10 years (Fig 3).

Univariate analysis revealed aortic valve *z* score, 1-patch PA reconstruction technique and Taussig-Bing anomaly as risk factors for the RVOT obstruction. On multivariate analysis, the preoperative aortic valve *z* score was shown to be a risk factor for RVOT obstruction. In our study, no specific risk factor was found to significantly predict the intervention for distal PA obstruction (Table 3).

Small Aortic Valve z Score

The *z* scores were less than −2 in 13 patients (25%). Two patients underwent RVOT repair at the time of the first operation (transannular patch repair and RV-PA conduit placement). Of those patients who did not undergo RVOT repair at the time of the operation, 2 patients (*z* scores, −3.17, −2.23) eventually underwent RVOT repair 6 months and 12 months later. Eight patients with a *z* score of less than −2 were free from RVOT reoperation (Table 4).

Left-Sided Lesion

There were two reoperations and two catheter interventions for left sided obstruction. Actuarial freedom from reoperation or reintervention was 94% (95% CI, 84% to 98%) at 1 year, 88% (95% CI, 74% to 95%) at 5 years, and 83% (95% CI, 65% to 93%) at 10 years. There was no significant difference for the rate of reintervention for the arch obstruction based on the various methods of reconstruction.

There was no reoperation for neoaortic insufficiency or neoaortic stenosis during the study period.

Comment

Despite the improvements in surgical techniques and perioperative intensive care management, the operation for TGA/CoA or Taussig-Bing/CoA remains a surgical challenge [1, 2, 8]. It has been postulated that the malalignment of the RVOT can cause aortic arch anomalies by diverting flow from the aorta to the PA [9], resulting in more complex anatomy in these patients and an increased operative mortality [10–12]. Planché and colleagues [11] reported that RVOT obstruction occurs at a lower rate in single-stage repair than in two-stage repair, possibly because the previous PA banding can cause infundibular hypertrophy and significant postoperative RVOT obstruction.

Tchervenkov and colleagues [6] reported 13 consecutive cases of single-stage repair for TGA/CoA in which 3 patients (23%) underwent a transannular patch repair of the RVOT and 1 patient (7%) underwent a modified Damus-Kaye-Stansel operation with aortic arch augmentation. Pigott and colleagues [7] described five cases of single-stage repair for TGA/CoA, with all patients undergoing transannular patch or RV-PA conduit. In a series from Wetter and colleagues [12], RVOT obstruction was reported to often

Table 3. Analyses of the Risk Factor for Proximal and Distal Right-Sided Obstruction

(A) Univariate Analyses

Variables	RVOT No. (%)	*p* Value	Distal PA No. (%)	*p* Value
Overall right-sided operation/intervention	6 (11.3)		6 (11.3)	
Aortic *z* score		0.0404[a]		0.235
Weight		0.867		0.424
LCA from sinus 2	1/14 (7.1)	0.663	1/14 (7.1)	1.000
1-patch PA reconstruction	4/13 (30.8)	0.0331[a]	3/13 (23.1)	0.293
Taussig-Bing	5/21 (23.8)	0.0201[a]	3/21 (14.3)	0.671

(B) Multivariate Analyses

Variable	χ^2	OR	*p* Value
Aortic valve *z* score	4.20	18.150	0.0404[a]

[a] Signifies a statistically significant value ($p < 0.05$).

LCA = left coronary artery; PA = pulmonary artery; OR = odds ratio; RVOT = right ventricular outflow tract.

Table 4. First Operation and Reoperation for Patients Whose Preoperative Aortic Valve z Scores Were Less Than −2

Patient No.	Weight (kg)	Aortic Annulus (mm)	*z* Score	RVOTR at ASO	Taussig-Bing	PA Reconstruction	RVOT-PG at Discharge (mmHg)	Reoperation (Death)
1	3.0	4.5	−3.24	Transannular patch	Yes	1 patch	0	
2	3.8	6	−2.37	RV-PA conduit	Yes	1 patch	30	
3	4.0	5	−3.17	No	Yes	2 patches	10	Transannular patch (6 months)
4	3.1	5.5	−2.52	(Subaortic stenosis resection)	No	1 patch	10	
5	2.6	5	−2.51	(Subaortic stenosis resection)	Yes	1 patch	12	
6	3.6	6	−2.24	No	No	2 patches	0	
7	4.0	6	−2.24	(Subaortic stenosis resection)	Yes	1 patch	30	
8	3.0	5.5	−2.23	(Subaortic stenosis resection)	Yes	2 patches	50	Transannular patch (12 months)
9	3.0	5.4	−2.17	No	No	2 patches	20	
10	3.0	5.8	−2.13	No	No	2 patches	0	
11	3.3	6	−2.1	(Subaortic stenosis resection)	Yes	2 patches	0	
12	3.2	6	−2.1	No	No	2 patches	0	
13	3.4	6	−2.1	No	No	1 patch	45	Late death (RSV infection, 6 months)

ASO = arterial switch operation; RVOTR = right ventricular outflow tract repair; PA = pulmonary artery; PG = pressure gradient; RSV = respiratory syncytial virus.

complicate the postoperative course of Taussig-Bing hearts and to be the main indication for reintervention. These authors have stressed the importance of subaortic stenosis by the conal malalignment and have been more aggressive than us in this respect. However, unlike tetralogy of Fallot, in which RV hypertrophy complicates the morphology of infundibulum, patients with TGA/CoA tend to have a less hypertrophied RV, especially when single-stage repair is applied, and more conservative management remains a possibility. Moreover, recent reports demonstrated good long-term survival after the hospital discharge [3, 4]. Thus, it may safely be said that the key for the good long-term survival is to keep the mortality as low as possible at the time of the single-stage operation.

We adopted single-stage repair with excellent survival with minimal use of transannular patch or RV-PA conduit at the time of the first operation whenever possible. Transannular patch repair or placement of the RV-PA conduit was done only once each at the time of the operation in our series. Although 4 patients underwent reoperation, freedom from reintervention was comparable to those patients with simple TGA and TGA with VSD [5]. Our limited application of concurrent procedures on the RVOT reduces cardiopulmonary bypass and cross-clamp time of the already complex operation. Moreover, we can avoid an unnecessary transannular patch with free pulmonary insufficiency by this limited approach.

In conclusion, our results are notable for three salient findings: First, outcomes after a single-stage repair for TGA/CoA or Taussig-Bing/CoA are excellent, with a low rate of reoperation or reintervention.

Second, our univariate analysis revealed Taussig-Bing anomaly, PA reconstruction with one patch, and smaller aortic valve *z* score are risk factors for RVOT obstruction. However, only a smaller aortic valve *z* score was demonstrated to be predictive in the multivariate domain, suggesting that the need for one pantaloon patch for reconstruction and the diagnosis of Taussig-Bing were surrogates of the small aortic annulus. Although a small aortic valve *z* score is a significant risk factor for the RVOT obstruction, when and at what threshold we should address RVOT is still unclear.

Finally, our strategy of one-stage repair with limited application of concurrent procedures on the RVOT tract offers excellent outcomes and has a low risk of reintervention for right-sided obstruction.

References

1. Daebritz SH, Nollert G, Sachweh JS, Engelhardt W, von Bernuth G, Messmer BJ. Anatomical risk factors for mortality and cardiac morbidity after arterial switch operation. Ann Thorac Surg 2000;69:1880–6.
2. Williams WG, McCrindle BW, Ashburn DA, Jonas RA, Mavroudis C, Blackstone EH; Congenital Heart Surgeon's Society. Outcomes of 829 neonates with complete transposition of the great arteries 12–17 years after repair. Eur J Cardiothorac Surg 2003;24:1–9.
3. Pocar M, Villa E, Degandt A, Mauriat P, Pouard P, Vouhé PR. Long-term results after primary one-stage repair of transposition of the great arteries and aortic arch obstruction. J Am Coll Cardiol 2005;46:1331–8.
4. Mohammadi S, Serraf A, Belli E, et al. Left-sided lesions after anatomic repair of transposition of the great arteries, ven-

tricular septal defect, and coarctation: surgical factors. J Thorac Cardiovasc Surg 2004;128:44–52.
5. Williams WG, Quaegebeur JM, Kirklin JW, Blackstone EH. Outflow obstruction after the arterial switch operation: a multiinstitutional study. Congenital Heart Surgeons Society. J Thorac Cardiovasc Surg 1997;114:975–87.
6. Tchervenkov CI, Tahta SA, Cecere R, Béland MJ. Single-stage arterial switch with aortic arch enlargement for transposition complexes with aortic arch obstruction. Ann Thorac Surg 1997;64:1776–81.
7. Pigott JD, Chin AJ, Weinberg PM, Wagner HR, Norwood WI. Transposition of the great arteries with aortic arch obstruction. Anatomical review and report of surgical management. J Thorac Cardiovasc Surg 1987;94:82–6.
8. Takeuchi K, McGowan FX Jr, Moran AM, et al. Surgical outcome of double-outlet right ventricle with subpulmonary VSD. Ann Thorac Surg 2001;71:49–52.
9. Rudolph AM, Heymann MA, Spitznas U. Hemodynamic considerations in the development of narrowing of the aorta. Am J Cardiol 1972;30:514–25.
10. Moene RJ, Ottenkamp J, Oppenheimer-Dekker A, Bartelings MM. Transposition of the great arteries and narrowing of the aortic arch. Emphasis on right ventricular characteristics. Br Heart J 1985;53:58–63.
11. Planché C, Serraf A, Comas JV, Lacour-Gayet F, Bruniaux J, Touchot A. Anatomic repair of transposition of great arteries with ventricular septal defect and aortic arch obstruction. One-stage versus two-stage procedure. J Thorac Cardiovasc Surg 1993;105:925–33.
12. Wetter J, Sinzobahamvya N, Blaschczok HC, Cho MY, Brecher AM, Grävinghoff LM, Urban AE. Results of arterial switch operation for primary total correction of the Taussig-Bing anomaly. Ann Thorac Surg 2004;77:41–6; discussion 47.

DISCUSSION

DR RODOLFO NEIROTTI (Geneva, Switzerland): I wonder if in those patients that have an associated coarctation, you inspect the right ventricular outflow tract at the time of the arterial switch operation through the aortic valve after removing the coronary arteries. And then, if there is some excess muscle, do you do something about it? We and others have found that the right ventricular outflow tract is can be abnormal in this particular group. It is not the neopulmonic valve.

DR HIRATA: Yes, we routinely inspect the right ventricular outflow tract in these patients. After division of the great vessels and removal of the coronary arteries, the surgeon is afforded a very good view of the subaortic region. This area is visually inspected and often measured with a Hagar dilator. Any significant fibromuscular tissue is resected.

DR FRANÇOIS G. LACOUR-GAYET (Denver, CO): I would like to ask you three questions:

One, I assume that 72% of the VSD were close to the pulmonary valve. In our experience, this has been a concern, because in fixing the VSD through the pulmonary valve, it can then tear on the pulmonary leaflets and further create aortic regurgitation. That is my first question.

Two, do you have any interrupted aortic arches in your series? Because those are really the ones who show a great provider of late RVOT obstruction.

Three, the transannular patch is the best solution to offer to RVOT obstruction seen late after the repair. In fact, as you know, very often there is what we call ourselves a double loop with a right coronary running in front of the pulmonary annulus. And it's very difficult to do a transannular patch and the only solution left is to do a RV to PA conduit.

DR HIRATA: Thank you Dr Lacour-Gayet. We have read with great interest your manuscripts concerning patients with TGA, VSD, and aortic coarctation. In this series, the vast majority of the VSDs were closed through the pulmonary valve. The patch is carefully trimmed to avoid over-sizing and is sewn to the muscle immediately subjacent to the valve leaflets. The function of the neoaortic valve was evaluated over time. Currently, 4 patients have mild and 1 patient moderate aortic insufficiency. Recently, 1 patient required aortic valve replacement 15 years following his initial surgery. This operation was performed last month and is not included in our data analysis. Interestingly, we feel closure of the VSD through the pulmonary valve may lead to less neopulmonary obstruction due to the absence of the patch in the right ventricular outflow tract following the procedure.

There were 2 patients with interrupted aortic arch. Both had significant tricuspid stenosis and were excluded from this study.

We did not encounter a patient who needed a transannular patch in whom a coronary anomaly would preclude its placement. However, this is an important consideration. Thank you for your comments.