The significance of pulmonary annulus size in the surgical management of transposition of the great arteries with ventricular septal defect and pulmonary stenosis

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Objective: Aortic translocation has received growing attention in the management of complete transposition with ventricular septal defect and pulmonary stenosis, but the criteria regarding pulmonary stenosis for selecting this option have yet to be established. The aim of this study is to evaluate the significance of pulmonary annulus size with the outcome after the arterial switch operation.

Methods: Between November 1996 and September 2008, 250 patients underwent the arterial switch operation for complete transposition. Among them, 8 patients with a pressure gradient greater than 30 mm Hg, bicuspid pulmonary valve, and an aortic Z-score of the pulmonary annulus less than 0 were included in this retrospective study. The median age was 19.1 months (range, 0.5–80.0 months). The median follow-up was 39.7 months (range 9.1–139.5 months).

Results: At latest follow-up, the Z-score of the neo-aortic annulus increased from -1.50 ± 1.13 (range, -3.42 to -0.35) to 1.10 ± 1.15 (range, -0.8 to 2.10) ($P < .01$). No patient had a significant pressure gradient across the left ventricular outflow tract. There was 1 early death and there were no late deaths. Two reoperations were performed in 1 patient for neo-aortic stenosis at 81 months and 110 months after the operation. Latest echocardiogram revealed grade 0 or 1 neo-aortic insufficiency.

Conclusion: It was possible to extend the indication for the arterial switch operation with acceptable outcome to the patient with a Z-score of about -3 of the pulmonary annulus despite bicuspid pulmonary valve. Inasmuch as the arterial switch operation has benefits over the other options, a large-scale study is required for more reasonable triage in this group of patients. (J Thorac Cardiovasc Surg 2010;139:135-8)

The Rastelli operation has been the standard surgical option in the management of transposition of the great arteries (TGA) with ventricular septal defect (VSD) and pulmonary stenosis (PS) since its introduction,¹ but recently, two large series reported less optimal outcomes of the Rastelli procedure.^{2,3} In contrast, the aortic translocation that had been first described in 1984⁴ showed excellent midterm outcomes and performance of the left ventricle with better outflow alignment,^{5,6} although the aortic translocation still had a problem with the right ventricular outflow tract (RVOT).⁶ The arterial switch operation (ASO) is the established procedure, and many surgeons have experienced stable outcomes with it,^{7,8} even in patients with a bicuspid pulmonary valve or other left ventricular outflow tract obstructions (LVOTO).⁹⁻¹¹ However, the guidelines for selection of the

surgical option with regard to PS has not been established in patients with TGA with VSD and PS.

In this study we evaluated the outcomes after the ASO in patients who are possible candidates for aortic translocation, focusing on the pulmonary annulus size.

PATIENTS AND METHODS

Between November 1996 and September 2007, 8 patients underwent the ASO for TGA with VSD, bicuspid pulmonary valve, and small pulmonary annulus. One of them had undergone palliation with an aortopulmonary shunt before the ASO. All patients had a pressure gradient greater than 30 mm Hg across the left ventricular outflow tract (LVOT) (median, 42.5 mm Hg; range, 30–81 mm Hg), bicuspid pulmonary valve, and an aortic Z-score of the pulmonary annulus of less than 0 (median, -1.25 ; range, -3.42 to -0.35) (Table 1). In 3 patients, LVOTO at the subvalvular level was combined (1 tissue tag, 2 hypertrophic conal septa) (Table 1). All patients had interatrial communication and VSD. Coronary arterial pattern was usual in all patients. The median age and weight at operation were 19.1 months (range, 0.5–80 months) and 10.0 kg (range, 3.3–18.8 kg).

In the same period, 9 patients underwent the Rastelli operation for TGA with VSD and PS. Among them, an elective Rastelli operation was performed in 5 patients who had an aortic Z-score of the pulmonary annulus of less than -4 . In the other patients, an operative plan was determined for the Rastelli operation in the operating room owing to dysplastic pulmonary valve or unrelievable tunnel-form subaortic stenosis.

The ASO was conducted with moderate hypothermic cardiopulmonary bypass, the Lecompte maneuver, and trapdoor coronary transfer. To relieve the LVOTO, we performed commissurotomy of the pulmonary (neo-aortic) valve

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Abbreviations and Acronyms

ASO	= arterial switch operation
LVOT	= left ventricular outflow tract
LVOTO	= left ventricular outflow tract obstruction
PS	= pulmonary stenosis
RVOT	= right ventricular outflow tract
TGA	= transposition of the great arteries
VSD	= ventricular septal defect

in 4 patients, tissue tag resection in 1, and resection of hypertrophic conal septum in 2 through the pulmonary (neoaortic) valve. All VSDs were closed through the tricuspid valve, and all neopulmonary arteries were reconstructed with single pantaloony-shaped fresh autologous pericardium.

The median follow-up was 39.7 months (range, 9.1–139.5 months) and all patients were included to be followed up to the end date of this study (June 2008). LVOT and cardiac performance were assessed by echocardiography during follow-up.

Our institutional review board/ethical committee approved this retrospective study and waived the informed consent. Data are presented as median with range using Microsoft Excel 2007.

RESULTS

The median cardiopulmonary bypass time and myocardial ischemic time were 213 minutes (range, 140–274 minutes) and 128 minutes (range, 96–157 minutes), respectively. Bypass was weaned smoothly and the sternum was closed in all patients. The median mechanical ventilation time, stay in the intensive care unit, and hospital time were 78 hours (range, 22–255 hours), 6 days (range, 2–15 days), and 9.5 days (range, 7–24 days), respectively.

There was 1 early death. A 35.4-month-old boy underwent standard ASO and a concomitant LVOTO-relieving procedure including commissurotomy and resection of the hypertrophic conal septum (Table 1). Echocardiography in the immediate postoperative period showed both good ventricular contractility and no residual LVOTO with a low dosage of dopamine ($5 \mu\text{g} \cdot \text{kg}^{-1} \cdot \text{min}^{-1}$). He died of tension pneumothorax and subsequent pulmonary edema on the third postoperative day.

Two reoperations were required in 1 patient who had undergone ASO with commissurotomy and resection of a tissue tag at 30.4 months of age. At 81 months after the operation, recommissurotomy was performed for neoaortic valve stenosis with an LVOT pressure gradient of 77 mm Hg; then a 21-mm mechanical prosthesis was implanted without annular enlargement 29 months later (at 11.7 years of age). At 2 years of follow-up, echocardiography revealed no significant pressure gradient across the mechanical valve, and there was no thromboembolic or hemorrhagic complication.

The Z-score of the pulmonary (neoaortic) annulus has caught up with somatic growth at latest follow-up (Figure 1), and in a patient who required reoperation, valve replacement was performed without an additional annular enlargement procedure. The pressure gradient across the LVOT decreased and was not significant in all but 1 reoperative case (Figure 2).

In all patients, the latest echocardiogram showed aortic regurgitation of grade 0 or 1 and no significant stenosis or regurgitation of the RVOT. All patients were in New York Heart Association functional class I and did not require any cardiac medication at latest follow-up.

TABLE 1. Patients' characteristics

				LVOTO			Preop				Last follow-up				
	Age (mo)	Bwt (kg)		Bicuspid	Subvalvular	PG (mm Hg)	PV size (mm)	PV Z-score‡			SaO ₂ (cath)	Previous palliation	LVOT procedure	Interval (mo)	PG (mm Hg)
1*	30.4	12.9		Yes	Tissue tag	49	11	−0.6	73%	—	Commissurotomy. Tissue tag resection	140	—	1.3	—
2†	35.4	14.3		Yes	Hypertrophic conal septum	64	11	−1.8	78%	BAS	Commissurotomy. Conal septum resection	—	—	—	—
3	79.6	18.8		Yes	—	81	12.4	−1	83%	—	—	111	9	2.0	0
4	39.8	14.8		Yes	—	71	11	−1.1	77%	BAS, RMBT	Commissurotomy	58	23	1.4	0
5	2.3	4.9		Yes	—	36	8	−0.4	67%	—	—	40	12	1.9	0
6	7.8	7		Yes	—	36	6	−3.4	—	—	Commissurotomy	29	14	−0.8	0
7	0.6	3.3		Yes	Hypertrophic conal septum	30	6	−1.4	75%	BAS	Commissurotomy. Conal septum resection	11	29	2.1	1
8	0.5	3.28		Yes	—	36	5	−2.7	80%	BAS	—	9	19	−0.2	0

Bwt, Body weight; LVOTO, left ventricular outflow tract obstruction; PG, pressure gradient; PV, pulmonary valve; Sao_2 , arterial oxygen saturation; LVOT, left ventricular outflow tract; NeoAoV, neoaortic valve; NeoAR, neoaortic regurgitation; BAS, balloon atrial septostomy; RMBT, right modified Blalock-Taussig shunt. *Reoperation; †early mortality; $\ddagger$ Z-score was calculated using Cincinnati Children's Hospital formula.¹²

FIGURE 1. Changes in Z-score of the pulmonary (neoaortic) annulus. F/U, Follow-up.

DISCUSSION

TGA with VSD and PS is uncommon; the prevalence was less than 1% of all congenital heart diseases⁵ and about 20% in patients with TGA.⁹ Since 1989, PS or LVOTO was combined in 16 (6.4%) patients among 250 patients who underwent the ASO for TGA or variants in our center.

Among surgical options for TGA with VSD and PS, the Rastelli operation has been used as the standard procedure for many years, but its long-term outcome was less optimal in terms of the LVOT, RVOT, and even survival.^{2,3} It was time for attention to be turned toward aortic translocation.

Morell,⁵ Yeh,⁶ and their associates reported excellent results with aortic translocation, with better LVOT performance than that provided by other surgical options; however, the authors noted a still high reoperation rate for RVOT: 64% reintervention-free rate at 15 years.⁶ Hu and colleagues¹³ described the modification of the aortic translocation procedure, called double root translocation. In their report, the indication for aortic translocation was only a pressure gradient across the LVOT of more than 50 mm Hg. They translocated the pulmonary roots to preserve the competence and growth potential of the pulmonary valve, but during short-term follow-up (range, 2–26 months), 9 of 11 patients had pulmonary insufficiency—mild in 3 and mild to medium in 6. Overall, however, although aortic translocation is better than the Rastelli operation in terms of the LVOT, the outcome is similar when compared with repaired tetralogy of Fallot in terms of the RVOT.

In 1990, Wernovsky and colleagues⁹ suggested that mild pulmonary valve abnormalities and surgically reparable subpulmonary stenosis did not preclude an ASO. Uemura and colleagues¹⁰ reported that the ASO can be performed in patients with a bicuspid pulmonary valve. In this report, all patients had a pulmonary annulus larger than 100% of normal reference value. A recent anatomic

FIGURE 2. Changes in pressure gradient across the LVOT. F/U, Follow-up.

study by Hazekamp, Portela, and Bartelings¹⁴ indicated that a hypoplastic pulmonary annulus precluded an ASO. But how small is small? Sohn and colleagues¹¹ noted that the ASO could be performed in patients with combined LVOT abnormalities, including an aortic Z-score of the pulmonary annulus ranging from -3.16 to 3.79 , regardless of valve morphology.

Regarding the growth potential of the semilunar valve, McElhinney and coworkers¹⁵ reported that the aortic annulus diameter in patients with critical aortic stenosis who survived the early period after balloon aortic valvuloplasty increased to the normal range within months, and over time, the Z-score of the aortic annulus was generally normal. From this result, it can be inferred that in some of the patients who underwent aortic translocation or its modifications, simple ASO and concomitant relief of pulmonary (neoaortic) stenosis can be performed with future catch-up growth of the neo-aortic annulus. In our study, all patients had a bicuspid pulmonary valve and a Z-score of the pulmonary annulus less than 0 (lowest, -3.42) preoperatively. We could observe that the diameter of the neo-aortic annulus caught up with somatic growth and was normalized with time (Figure 1).

This study is limited by the inclusion of a small number of highly selected patients who had a variety of pulmonary annulus sizes and by its retrospective methodology. Despite these limitations, we found that our data could be useful for decision-making in this group of patients.

In conclusion, the ASO can be performed safely in patients with TGA and VSD with a bicuspid pulmonary valve and a Z-score of the pulmonary annulus less than 0 (lowest, -3.42). The diameter of the neo-aortic annulus caught up with somatic growth, and the function of the neo-aortic valve was preserved during follow-up. A large-scale study may be required to extend the safe indication of the ASO in this group of patients.

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