Results of Rapid Two-stage Arterial Switch Operation in Patients with Transposition of the Great Arteries : One-year Postoperation

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Abstract

The rapid two-stage arterial switch operation is an alternative therapy for patients with simple transposition of the great arteries who present beyond the neonatal period and have low left ventricular pressure. It provides normal ventricular function compared to the atrial switch operation. Between July 1994 and February 1997, there were 13 such infants who had rapid two- stage arterial switch operation performed at Siriraj Hospital. There was 1 late death (11 months after the operation). All 12 survivors (mean age 22.4±5.7 months) were clinically evaluated and had echocardiography performed at 14.8±4.9 months after the operation. All were asymptomatic. Echocardiogram revealed a residual small atrial septal defect (1 case), small ventricular septal defect (1 case), mild supravalvar neopulmonary stenosis (2 cases), bicuspid neoaortic valve without stenosis (2 cases), dilated neoaortic sinus of Valsalva (6 cases, 50%) and mild neoaortic insufficiency (11 cases, 91.7%). The left ventricular function was hyperdynamic after pulmonary artery banding and significantly decreased to normal level at the time of study (shortening fraction of 43.8±10.7 *vs* 29.2±3.8%, respectively, $p = 0.0005$). The wall thickness was significantly increased after pulmonary artery banding and decreased overtime (0.48±0.08 *vs* 0.32±0.05 cm, respectively, $p < 0.0005$). The left ventricular dimension was significantly increased both after pulmonary artery banding and at the time of study (2.06±0.42 *vs* 3.32±0.30 cm, respectively, $p < 0.0005$). The left ventricular mass was significantly increased after pulmonary artery banding and at the time of study (21.79±7.79 *vs* 33.08±7.40 g/m^2, respectively, $p = 0.0005$). The mortality and morbidity of rapid two-stage arterial switch operation are low. However, long-term follow-up should be monitored.

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The rapid two-stage arterial switch operation (ASO) is an attractive therapy for developing countries, lacking of centers offering ASO, where a number of infants with simple transposition of the great arteries (TGA) are referred beyond the first month of life. Preservation of systemic ventricular function is expected with the ASO compared with atrial switch operation[1]. There are an increasing number of reports that show good early and late results of ASO for simple TGA by either a primary or two-stage ASO[2-4]. At Siriraj Hospital, we started to perform rapid two-stage ASO in patients with simple TGA who presented late and had low left ventricular pressure in July 1994. The purposes of the present study were to evaluate the results (approximately 1 year after the operation) including 1) the parameters obtained by echocardiography: the left ventricular wall thickness, dimension, mass, function and interventricular septal thickness compared to the parameters obtained after the first stage operation (pulmonary artery banding, PAB). 2) the outcome, residual lesion and complication of the rapid two-stage ASO, such as supravalvar stenosis and neoaortic regurgitation.

PATIENTS AND METHOD

All 13 patients who had undergone the rapid two-stage ASO for simple TGA (with intact ventricular septum or insignificant left-to-right shunt of ventricular septal defect) at Siriraj Hospital between July 1994 and February 1997 were enrolled in the present study. The specifics of this patient population including anatomic variations, preoperative clinical data, operative procedure, serially echocardiographic evaluation before and after the first stage operation and the immediate results of the operation have been reported in detail elsewhere[5]. Every patient was clinically evaluated and performed echocardiographic examination at approximately 1 year after the ASO.

Echocardiographic assessment: All patients were sedated with chloral hydrate 50-75 mg/kg. The echocardiographic examination was performed with a Hewlett-Packard Sonos 1000 (Hewlett-Packard Co., Palo Alto, Calif). The examination included two-dimensional, Doppler, color-flow mapping and M-mode echocardiographic imaging in parasternal long and short axis, apical and subcostal views. The basic studies included assessment of residual anatomic defects, and valvular function. The presence and severity of neoaortic insufficiency were assessed by color Doppler echocardiography and graded on a four-point scale by measuring the size of the proximal regurgitant jet width (absent; upto 4 mm, mild; 4 to 6 mm, moderate; > 6 mm, severe)[6]. M-mode echocardiography of circular short axis left ventricle was obtained in parasternal short axis view. Measurements were obtained by averaging data from three cardiac cycles at the end of diastole included left ventricular short-axis dimension, posterior wall thickness, interventricular septal thickness. The left ventricular mass was calculated by the formula of Devereux and associations[7]. The shortening fraction was also calculated. All these parameters were compared to those obtained after the PAB and the last parameter was compared to those obtained initially before the operation.

Statistical analysis

The categorical variables were presented as count. The continuous variables were presented as mean ± SD and compared by using the Student's *t*-test for paired samples. The left ventricular mass of 3 values; initial before the operation, after pulmonary artery banding and at the time of the study, were compared by using ANOVA analysis for repeated measurements. P value of less than 0.05 was considered significant.

RESULTS

During the study period, 13 patients underwent rapid two-stage ASO. There was 1 late death at 11 months after the surgery because of persistent pulmonary hypertension and intractable right heart failure. Therefore, only 12 cases of survivors, comprising of 5 females (41.7%) and 7 males (58.3%) were evaluated. The mean age was 22.4 ± 5.7 months with the range from 16 to 35 months. The mean duration between the second stage, ASO and the time of the present study was 14.8 ± 4.9 months with the range from 11 to 25 months. All were asymptomatic. One case had improved, his hemiparesis had not fully recovered, however, he can walk and has normal intelligence. Systolic murmur, not greater than grade II, was heard over the left sternal border in 4 cases. Other physical examination was normal.

Echocardiographic evaluation: The abnormal findings of two-dimensional and Doppler echocardiography are shown in Table 1. Every case has normal atrioventricular valvar function. All

Table 1. Summary of two-dimensional and Doppler echocardiographic findings in our patients.

Findings	n	%
Small atrial septal defect (5 mm)	1	8.3
Small ventricular septal defect (2 mm)	1	8.3
Supravalvar neopulmonary stenosis*	2	16.7
Bicuspid neoaortic valve**	2	16.7
Dilated sinus of Valsalva of neoaortic valve	6	50.0
Aortic insufficiency, mild	11	91.7

* Mild pressure gradients were obtained (16 and 30 mmHg, respectively) at anastomotic site.
** no stenosis

Table 2. The comparison of echocardiographic parameters obtained after the first stage, pulmonary artery banding (PAB), and at the present study (post ASO) of our patients (n=12).

Variables	Post PAB		Post ASO		P value
Shortening fraction (%)	27.80-63.2	(43.8±10.7)	26.10-39.0	(29.2±3.8)	0.0005
LVPW (cm)	0.40-0.67	(0.48±0.08)	0.24-0.41	(0.32±0.05)	< 0.0005
LVID (cm)	1.00-2.67	(2.06±0.42)	2.98-4.07	(3.32±0.30)	< 0.0005
IVT (cm)	0.44-0.62	(0.51±0.07)	0.24-0.57	(0.44±0.10)	0.077
LV mass (g/m^2)	7.30-36.37	(21.79±7.79)	21.05-44.71	(33.08±7.40)	0.0005

LVPW, left ventricular posterior wall thickness; LVID, left ventricular internal dimension; IVT, interventricular septal thickness

cases except one have mild neoaortic (native pulmonary valve) regurgitation by color-flow mapping. Focusing on native pulmonary valve by retrospective reviews of initially echocardiographic studies (before the surgery), there was only 1 case with mild regurgitation, 1 case with asymmetric (eccentric) valvar closure and 2 cases with bicuspid valve.

The left ventricular function was near normal or normal in every case with the mean shortening fraction of 29.2±3.8 per cent. The wall thickness was significantly increased after the PAB[5] and significantly decreased at the time of the study ($p < 0.0005$, Table 2). The internal dimension was significantly increased both after the PAB[5] and at the present study ($p < 0.0005$), as shown in Table 2. The left ventricular mass was significantly increased after the PAB[5] and at the present study ($p = 0.0005$). The interventricular septal thickness was not significantly increased after the PAB[5] and decreased overtime ($p = 0.077$).

DISCUSSION

Arterial switch operation has recently been the treatment of choice for simple transposition of the great arteries either by a primary or two-stage approach[2,8,9]. Banding of the pulmonary artery (the first stage) to induce left ventricular hypertrophy followed by ASO (the second stage) within 1-2 weeks has been performed when a primary ASO was considered high risk because of inadequate left ventricular hypertrophy[10,11]. In the present study, the left ventricular mass, reflecting the hypertrophy, was significantly increased after pulmonary artery banding[5] and at follow-up. The banding interval should be minimized as soon as possible as previously reported to reduce risk factors for late dysfunction[11]. Ventricular hypertrophy secondary to an acute pressure stimulus may also be associated with focal areas of necrosis,[12] and limitations of coronary vascular reserve[13]. However, the immature myocardium is relatively resistant to the adverse effects of hypertrophy that

have been described in mature animals[14,15]. Ventricular function may be adversely affected in some individuals by a two-stage approach compared with a primary approach[16,17]. Higher rate of acquisition of ventricular mass after pulmonary artery banding, a higher ratio of left ventricular to right ventricular pressure before ASO and a lower ejection fraction after pulmonary artery banding were found to be associated with lower ventricular contractility at late follow-up[10]. Later age of pulmonary artery banding and longer duration of the left ventricular preparatory period were identified as factors that might adversely affect late left ventricular function[16-18]. In the present study, taken as a group, the left ventricular function was supranormal after the pulmonary artery banding[5] and significantly reduced to normal level at the time of the study, as in previous reports[10,19]. The left ventricular dimension was also significantly increased and the posterior wall thickness correlated inversely with follow-up, as in the previous report[10].

The frequency of mild *versus* no neoaortic regurgitation was significantly higher in the rapid two-stage ASO group than one-stage ASO[1,19]. The cause is uncertain, however, the possibilities are 1) intrinsic difference between the native aortic and pulmonary valves, which might lead to intolerance of the anatomic pulmonary valve to the force of the systemic circulation as shown in patients with other congenital heart diseases up to 60 per cent of cases[6]. Observations of pathologic specimens from patients with dTGA have suggested that the pulmonary valve cusps are not equal in size, leading to eccentric closure[20]. 2) the surgical procedure itself could contribute because the neoaortic root is abnormal as a result of the coronary reimplantation sites and the aortic circumferential suture line. 3) the pulmonary artery banding may directly damage the valve mechanism[1]. However, the degree of dysfunction was mild and did not progress on serial evaluation[1]. In the present study, although the majority of our patients (91.7%) had neoaortic regurgitation, not greater than mild degree by color-flow mapping echocardiography, the auscultatory findings and clinical were normal.

All of our survivors were asymptomatic after the second stage ASO. There was 1 case of late death, and 1 case of residual hemiparesis which may have been caused by clot embolization during the operation, as described elsewhere[5].

SUMMARY

Presently, the rapid two-stage arterial switch operation is an attractive therapy in developing countries for patients with simple transposition of the great arteries who present late and have low left ventricular pressure. It preserves ventricular function. The complication is low; such as supravalvar neopulmonary stenosis. Although the neoaortic regurgitation is frequently demonstrated by color-flow echocardiography, the clinical finding is insignificant. However, long-term follow-up should be monitored.

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REFERENCES

1. Colan SD, Boutin C, Castaneda AR, et al. Status of the left ventricle after arterial switch operation for transposition of the great arteries: Hemodynamic and echocardiographic evaluation. J Thorac Cardiovasc Surg 1995; 109: 311-21.
2. Norwood WI, Dobell AR, Freed MD, et al. Intermediate results of arterial switch repair: A 20-institute study. J Thorac Cardiovasc Surg 1988; 96: 854-63.
3. Planche C, Bruniaux J, Lacour Gayet F, et al. Switch operation for transposition of the great arteries in neonates: A study of 120 patients. J Thorac Cardiovasc Surg 1988; 96: 354-63.
4. Wernovsky G, Hougen TJ, Walsh EP, et al. Midterm results after the arterial switch operation for transposition of the great arteries with intact ventricular septum: Clinical, hemodynamic, echocardiographic and electrophysiologic data. Circulation 1988; 77: 1333-44.
5. Laohaprasitiporn D, Soongswang J, Nana A, et al. Immediate results of rapid two-stage arterial switch operation for simple transposition of great arteries. Thai Heart J 1998; 11: 1-6.
6. Jenkin KJ. Hanley FL, Colan SD, et al. Function of the anatomic pulmonary valve in the systemic circulation. Circulation 1991; 84 (Suppl): III 173-9.
7. Devereux RB, Lutas EM, Casale PN, et al. Standardization of M-mode echocardiographic left ventricular anatomic measurements. J Am Coll Cardiol 1984; 4: 1222-30.
8. Castaneda AR, Norwood WI, Jonas RA, et al. Transposition of the great arteries and intact ventricular septum: Anatomical repair in the neonate. Ann Thorac Surg 1984; 38: 438-43.
9. Idriss FS, Ilbawi MN, DeLeon SY, et al. Arterial switch in simple and complex transposition of the great arteries. J Thorac Cardiovasc Surg 1988; 95: 29-36.
10. Boutin C, Wernovsky G, Sanders SP, et al. Rapid two-stage arterial switch operation: evaluation of left ventricular systolic mechanics late after an acute pressure overload stimulus in infancy. Circulation 1994; 90: 1294-303.
11. Jonas RA, Giglia TM, Sanders SP, et al. Rapid, two-stage arterial switch for transposition of the great arteries and intact ventricular septum beyond the neonatal period. Circulation 1989; 80 (Suppl I): I 203-8.
12. Moore GW, Hutchins GM, Bulkley BH, et al. Constituents of the human ventricular myocardium: Connective tissue hyperplasia accompanying muscular hypertrophy. Am Heart J 1980; 100: 610-6.
13. Lamping KG, Dole WP. Acute hypertension selectively potentiates constrictor responses of large coronary arteries to serotonin by altering endothelial function in vivo. Circ Res 1987; 61: 904-13.
14. Flanagan MF, Fujii AM, Colan SD, et al. Myocardial angiogenesis and coronary perfusion in left ventricular pressure-overload hypertrophy in the young lamb: evidence for inhibition with chronic protamine administration. Circ Res 1991; 68: 1458-70.
15. Rakusan K, Flanagan MF, Geva T, et al. Morphometry of human coronary capillaries during normal growth and the effect of age in left ventricular pressure-overload hypertrophy. Circulation 1992; 86: 38-46.
16. Borow KM, Arensman FW, Webb C, et al. Assessment of left ventricular contractile state after anatomic correction of transposition of the great arteries. Circulation 1984; 69: 106-12.
17. Sievers HH, Lange PE, Onnasch DG, et al. Influence of the two stage anatomic correction of simple transposition of the great arteries on left ventricular function. Am J Cardiol 1985; 56: 514-9.
18. Lange PE, Sievers HH, Onnasch DG, et al. Up to 7 years of follow-up after two-stage anatomic correction of simple transposition of the great arteries. Circulation 1986; 74 (Suppl I): I 47-52.
19. Iyer KS, Sharma R, Kumar K, et al. Serial echocardiography for decision making in rapid two-stage arterial switch operation. Ann Thorac Surg 1995; 60: 658-64.
20. Kovalchin JP, Allen HD, Cassidy SC. Pulmonary valve eccentricity in d-transposition of the great arteries and implications for the arterial switch operation. Am J Cardiol 1994; 73: 186-90.

ผลการผ่าตัดด้วยวิธี rapid two-stage arterial switch ในผู้ป่วย transposition of the great arteries: ภายหลังการผ่าตัด 1 ปี

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ได้ศึกษาผู้ป่วย transposition of the great arteries ที่อายุมากกว่า 1 เดือน ร่วมกับมีความดันเลือดในเวนตริเคิลซ้ายต่ำ และได้รับการทำผ่าตัดรักษาด้วยวิธี rapid two-stage arterial switch ที่ โรงพยาบาลศิริราช ตั้งแต่เดือนกรกฎาคม 2537 ถึง กุมภาพันธ์ 2540 จำนวน 13 ราย มีผู้ป่วย 1 ราย เสียชีวิตภายหลังการทำผ่าตัด 11 เดือน ดังนั้นผู้ป่วย 12 รายที่เหลือ (อายุเฉลี่ย 22.4±5.7 เดือน) ได้รับ การตรวจร่างกายทั่วๆ ไป และตรวจหัวใจด้วยเครื่องมือคลื่นเสียงสะท้อนความถี่สูง ภายหลังการทำผ่าตัด 14.8±4.9 เดือน ทุกรายไม่มีอาการผิดปกติ ผลการตรวจหัวใจพบว่ามีรูรั่วขนาดเล็กที่ผนังหัวใจห้องบนและห้องล่างอย่างละ 1 ราย มีการตีบแคบเล็กน้อยของหลอดเลือดแดงของปอดบริเวณที่มีการตัดต่อ (เหนือลิ้นหัวใจ) 2 ราย ลิ้นหัวใจเอออร์ติกใหม่มีลักษณะ bicudpid แต่ไม่มีการตีบแคบ 2 ราย มีการขยายใหญ่ของ sinus of Valsalva ของลิ้นหัวใจเอออร์ติกใหม่ 6 ราย (ร้อยละ 50) และมีการรั่วเล็กน้อยของลิ้นหัวใจเอออร์ติก ใหม่ 11 ราย (ร้อยละ 91.7) การทำงานของเวนตริเคิลซ้ายพบว่า มากกว่าปกติภายหลังการทำผ่าตัดขั้นที่ 1 (pulmonary artery banding) และลดลงเป็นปกติอย่างมีนัยสำคัญในการศึกษาครั้งนี้ (shortening fraction ร้อยละ 43.8±10.7 และ 29.2±3.8 ตามลำดับ, p = 0.0005) ความหนาของผนังเวนตริเคิลซ้ายเพิ่มขึ้นอย่าง มีนัยสำคัญภายหลัง pulmonary artery banding และลดลงตามกาลเวลา (0.48±0.08 และ 0.32±0.05 ซม. ตามลำดับ, p < 0.0005) ส่วนขนาดของเวนตริเคิลซ้ายเพิ่มขึ้นอย่างมีนัยสำคัญทั้งภายหลังการทำ pulmonary artery banding และในการศึกษาครั้งนี้ (2.06±0.42 และ 3.32±0.30 ซม. ตามลำดับ, p < 0.0005) มวลของเวนตริเคิลซ้ายเพิ่มขึ้นอย่างมีนัยสำคัญเช่นกันทั้งหลังการผ่าตัด pulmonary artery banding และในการศึกษานี้ (21.79±7.79 และ 33.08±7.40 กรัม/ม^2 ตามลำดับ p = 0.0005) อัตราเสี่ยง และภาวะแทรกซ้อนภายหลังการทำผ่าตัดพบน้อย แต่ควรติดตามผู้ป่วยในระยะยาวต่อไป

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