

The Right and Left Ventricular Function After Surgical Correction with Pericardial Monocusp in Tetralogy of Fallot: Mid-Term Result

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Abstract

Background : Surgical repair of tetralogy of Fallot (TOF) with reconstruction of the right ventricular (RV) outflow tract invariably results in pulmonary regurgitation (PR). Chronic PR has been associated with RV dysfunction and ventricular arrhythmia. Pericardial monocusp has recently been used at Ramathibodi Hospital to preserve pulmonary valve function.

Objectives : First, to study the competency of the pericardial monocusp, one-year after correction. Second, to assess the right and left ventricular (LV) functions after surgery. Third, to assess correlation between severity of PR and the characters of electrocardiography (ECG) and chest X-ray (CXR) after correction.

Method : A cross-sectional study was conducted in patients who, had undergone total correction for TOF at least one year ago. The past medical history was retrospectively reviewed from the medical records. The patients who underwent surgical correction with and without pericardial monocusp were recruited into group I and group II, respectively. The clinical symptoms, QRS duration from ECG, and cardio-thoracic (CT) ratio from CXR were analyzed. From the echocardiographic standpoint, the LV systolic function was determined by LV fractional shortening (LVFS), whereas the RV systolic function was determined by the tricuspid annular plane systolic excursion (TAPSE). Restrictive physiology of the RV was determined by presence of antegrade flow across the pulmonary valve during diastole.

Results : Sixty four patients were enrolled in the study, 7 in group I and 57 in group II. The median follow-up time after the surgery was 6.5 years, which was 3 years in group I and 7 years in group II ($p < 0.01$). All patients in group I (100%) and 45 (80.4%) in group II had moderate or severe PR. The severity of PR, the RV and LV systolic functions were not statistically significantly different between the two groups ($p > 0.01$). The median of the LVFS was 32.4 per cent, and of the TAPSE was 10.5 mm. There was no restrictive physiology of the RV in all patients. There were no significant correlations between symptoms, CT-ratio, QRS duration and the severity of PR.

Conclusions : The pericardial monocusp could neither reduce severity of PR nor improve right and left ventricular functions after 3 years follow-up post-operatively. However, the right and left ventricular performances in mid-term period remained insignificantly changed and severity of PR could not be predicted from symptoms and simple laboratory investigations.

Key word : Tetralogy of Fallot, Total Correction, Pulmonary Regurgitation

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Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease with an incidence of 0.1/1,000 live births⁽¹⁾. Total surgical correction of TOF has been accomplished with success since the mid 1950s⁽²⁾. At present, the morbidity and mortality rates of the operation are extremely low. However, there are many reports of progressive right ventricular (RV) dilatation and dysfunction, ventricular arrhythmias, and sudden death after the conventional repair technique⁽³⁻⁵⁾.

Many factors influence the long-term outcome after total correction. Post-operative pulmonary regurgitation (PR) has been implicated as a major contributor to RV dysfunction and development of ventricular arrhythmia⁽⁶⁻⁸⁾. Several reports also correlate PR with reduced capacity for exercise⁽⁹⁻¹¹⁾. The transventricular repair has been the traditional approach to total correction of TOF⁽¹²⁾. However, extensive right ventriculotomy causes progressive RV dilatation, which may contribute to right heart failure and RV arrhythmias^(13,14). Older age at repair has also been associated with a less favorable outcome in several long-term follow-up studies⁽¹⁵⁻¹⁷⁾.

The transannular patch approach leaves the patients with free PR, which may eventually lead to RV volume overload^(18,19). An alternative to the transannular patch has been proposed, including fabricated pericardial or homograft monocusp valve⁽¹⁹⁻²¹⁾.

The pericardial monocusp technique is frequently used at Ramathibodi Hospital, as it is inexpensive and easily constructed. However, in Thailand, no report has been demonstrated for its durability and the improvement of post-operative outcome. The objectives of this study were to determine the competency of the pericardial monocusp after correction, to assess the right and left ventricular function after surgery, and to assess correlation between severity of PR and the electrocardiography (ECG) and chest X-ray (CXR) findings in the mid-term period (3-5 years follow-up) after correction.

PATIENTS AND METHOD

A cross-sectional study was conducted in those who had undergone total repair of TOF at Ramathibodi Hospital and remained alive more than 1 year after the operation. The authors excluded patients with atrioventricular septal defect, double-outlet right ventriculoarterial connection, absent pulmonary valve syndrome, and TOF in whom transannular patch, infundibular patch or pulmonary valvulotomy were not performed. Surgical history and follow-up status were obtained from the database of the Division of Pediatric Cardiology, Ramathibodi Hospital. Pre-operative status, age at operation, body weight, previous palliative procedure, concomitant cardiac anomaly, grade of pulmonary valve stenosis, diameter of pulmonary valve annulus, diameters of

right and left pulmonary arteries and McGoon ratio were collected for the pre-operative database. The operative records, post-operative status and complications were reviewed.

Patient evaluation included an interview for symptoms, physical examination, 12-lead electrocardiogram (ECG), chest radiography, transthoracic two-dimensional and Doppler echocardiography. Symptoms of palpitation or syncope, New York Heart Association (NYHA) functional class status were recorded, as well as the grade of pulmonary stenosis and pulmonary regurgitant murmur on physical examination. Twelve-lead ECG was evaluated for presence of right bundle branch block (RBBB), arrhythmia and QRS duration. The cardiothoracic ratio (CT-ratio) was calculated from CXR. Transthoracic two-dimensional, and Doppler echocardiography were performed on all patients using a multi-frequency (5-8 MHz) transducer interfaces with the Hewlett-Packard SONOS 4500 ultrasound system. Right ventricular and left ventricular end-diastolic dimensions were measured from a two-dimension directed M-mode recording in the parasternal long-axis view at the level of the mitral valve leaflet, then the right and left ventricular end-diastolic dimension ratio (RV/LV EDD ratio) was calculated. The left ventricular fractional shortening (LVFS) was derived from M-mode recording from the parasternal short-axis view, and normal LVFS was between 25-46 per cent⁽²²⁾. The right ventricular systolic function was

assessed by excursion of the tricuspid valve annulus in systole (TAPSE) recorded in the apical four-chamber view⁽²³⁾ (Fig. 1) and converted into right ventricular ejection fraction (RVEF) by the equation of $RVEF (\%) = 3.2 \times TAPSE (mm)$ ⁽²⁴⁾. The RVEF would be abnormal if less than 40 per cent⁽²⁴⁾. Systolic and diastolic Doppler pulmonary flow recordings were made with the sample placed at mid portion between the pulmonary valve leaflets and the bifurcation. The presence of diastolic antegrade pulmonary flow coincident with atrial systole on pulse Doppler recording as an indicator of RV diastolic dysfunction⁽²⁵⁾ (Fig. 2). Pulmonary stenosis (PS) was determined by the presence of the right ventricular outflow tract velocity more than 1.5 m./sec. PR was defined as the presence of diastolic regurgitant jet into the right ventricle and graded into mild, moderate and severe according to the ratio of regurgitant jet width and diameter of pulmonic valve annulus: mild PR, ratio less than 30 per cent; moderate PR, ratio 30-70 per cent; severe PR, ratio more than 70 per cent⁽²²⁾.

The patients were divided into two groups, group I repaired with pulmonary pericardial monocusp, and group II repaired without pericardial monocusp.

Statistical analysis

Data analysis was done with Epi Info (version 6) program. Group data are expressed as mean \pm SD. Student's *t*-test was used to compare normally

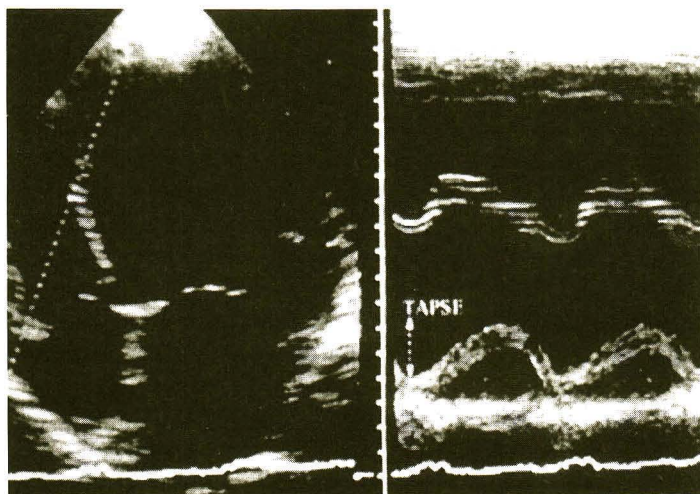


Fig. 1. Tricuspid annular plane systolic excursion (TAPSE) in apical-four-chamber view.

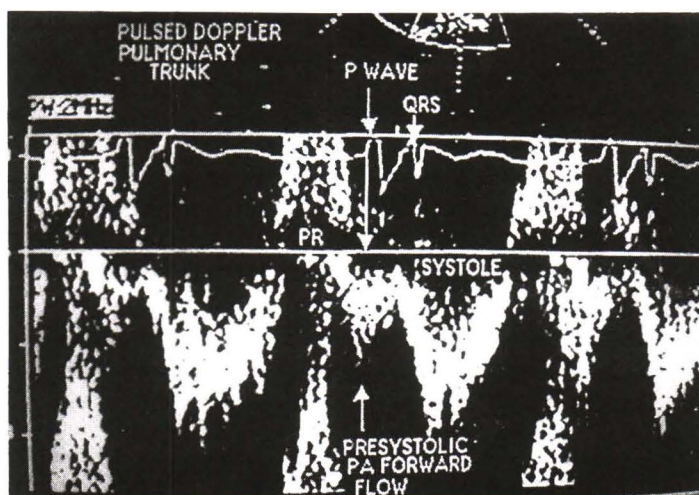


Fig 2. Restrictive right ventricular (RV) physiology⁽³⁰⁾. The pulse wave Doppler is in the pulmonary artery (PA). With atrial contraction, indicated by the P wave on the ECG, there is abrupt termination of the pulmonary regurgitation with presystolic forward flow into the PA, reflecting decrease RV compliance.

distributed variable, otherwise, a Mann-Whitney *U* test was performed. Statistical significant difference was inferred at a *p*-value of <0.01.

RESULTS

Patients

Sixty-four patients (35 males and 29 females) with repaired TOF between 1988 and 1998 fulfilled the inclusion criteria and were enrolled in the study. All of them were in NYHA class I. Their median age was 13 years (range 4-26 years). The median age at total repair was 6 years (range 2-16 years). The median follow-up time after total repair was 6.5 years (range 1-14 years). Their mean pulmonary valve annulus index was 1.20 ± 0.18 mm. The mean McGoon ratio was 1.98 ± 0.37 . The mean cardiac bypass time was 123.8 ± 37.2 min. Twenty-seven patients had previous palliative surgery: 20 patients with modified Blalock-Taussig (BT) shunt, and 7 patients with classical BT shunt.

Table 1 shows the characteristics of the patients in each group of the study. There were 7 patients in group I and 57 in group II. The median age at total repair and median follow-up time after repair in group I was significantly less than group II ($p < 0.01$). The cardiac bypass time and aortic cross clamp time in group I was significantly greater than that in group II ($p < 0.01$). Sex, percentage of previous

palliative shunts, pulmonary valve annulus diameter index and McGoon ratio were not statistically significantly different between the two groups.

Echocardiographic data (Table 2)

None had significant residual ventricular septal defect (VSD) or residual right ventricular out-flow gradient. One patient had a VSD patch aneurysm, and 1 had mild subaortic obstruction. There was no difference of RV/LV EDD ratio between the 2 groups, (0.83 ± 0.32 versus 0.67 ± 0.21 , $p = 0.16$). The means of LVFS, TAPSE and RVEF were within the normal ranges without significant difference between the 2 groups. All of the patients in group I and 44 patients (80.4%) in group II had moderate or severe PR. The existence of right ventricular restrictive physiology was investigated in all patients, and none of them appeared to have a forward flow in the pulmonary artery during atrial systole, indicating non-restrictive RV.

ECG findings

The mean QRS duration was 151 ± 34 m.sec. Sixty-two patients had right bundle branch block and none had ventricular tachycardia. One patient had a premature ventricular contraction without symptoms. No atrial arrhythmia was observed.

Table 1. Baseline data in the two studied groups.

Variables	Group I (n = 7)	Group II (n = 57)	P-value
Sex (M/F)	3/4	32/25	0.58
Palliative shunt (%)	42.8	42.1	0.63
Age (year) at total correction [median (max, min)]	5 (2, 5)	6.5 (2, 16)	<0.01
Time (year) after surgery [median (max, min)]	3 (1, 4)	7 (1, 14)	<0.01
Pulmonic valve annulus diameter index (mm/m ²) (Mean \pm SD)	0.85 \pm 0.19	0.88 \pm 0.17	0.96
McGoon ratio (Mean \pm SD)	2.02 \pm 0.19	1.98 \pm 0.39	0.255
Cardiac bypass time (min) (Mean \pm SD)	154.0 \pm 30.9	119.3 \pm 36.3	<0.01
Aortic cross clamp time (min) (Mean \pm SD)	106.8 \pm 16.9	67.3 \pm 25.6	<0.01

Table 2. Comparison of echocardiographic data between the two studied groups.

Variables	Group I (n = 7)	Group II (n = 57)	P-value
LVFS (%) (Mean \pm SD)	31.58 \pm 5.71	30.44 \pm 8.60	0.58
RV/LV EDD Ration (Mean \pm SD)	0.67 \pm 0.21	0.83 \pm 0.32	0.16
TAPSE (mm) (Mean \pm SD)	14.14 \pm 4.81	13.69 \pm 3.81	0.31
RVEF (%) (mean \pm SD)	45.3 \pm 13.4	43.8 \pm 12.4	0.28
Patients with moderate or severe PR (%)	100	75.4	0.15

LVFS : left ventricular fractional shortening,

RV/LV EDD ratio : right ventricular-left ventricular end-diastolic dimension ratio,

TAPSE : tricuspid annular plane systolic excursion, PR : pulmonary regurgitation

Correlation between severity of PR and ECG and CXR findings

Fifty three patients (79.7%) had moderate or severe PR, and 13 (20.3%) had mild PR. There was no significant difference of cardio-thoracic (CT) ratio measured from CXR, QRS duration from ECG, RV/LV EDD ratio, LVFS, TAPSE and RVEF between patients with moderate or severe PR and those with mild PR. However, 4 patients (8%) with moderate or severe PR had QRS duration over 180 m.sec and 1 had premature ventricular contraction.

Right ventricular outflow tract (RVOT) aneurysm

In the echocardiographic study, 5 patients had RVOT aneurysm and all of them had severe PR. Their chest X-ray revealed an enlarged and calcified pulmonary trunk. Their clinical and laboratory findings are demonstrated in Table 3.

DISCUSSION

Transannular patching of the right ventricular outflow tract in the surgical repair of TOF gives an excellent result in relieving the obstruction. However, this procedure invariably leaves PR. Long-term

PR may cause dilation of RV and has impacts on the RV systolic and diastolic functions and also may cause ventricular arrhythmia in the late post-operative period. Monocusp constructed from autologous pericardium has been utilized to minimize PR. The present study demonstrated that after 3 years follow-up, all of the patients who underwent pericardial monocusp for repair of TOF had developed a significant degree of PR, similar to previous reports of excellent early but late limited valve function due to tissue degeneration⁽¹⁹⁻²¹⁾. This tissue valve intends to improve early post-operative hemodynamic status from reduction of the RV volume load, especially in those who have pre-operative RV dysfunction. Recently, Roughneen PT, et al. used a monocusp constructed with polytetrafluorethylene (PTFE) to circumvent tissue degeneration. The result of a 17-month-follow-up of 7 patients with PTFE monocusp was good, and most of the patients had only mild to moderate PR⁽²⁶⁾.

The long-term right ventricular adaptation and systolic function after repair of TOF was determined by multiple factors such as early cyanosis, myocardial protection, extent of right ventriculotomy

Table 3. Clinical and laboratory data in post-repair tetralogy of Fallot patients who developed right ventricular outflow tract aneurysm.

Patient	Age (yr)	Time after surgery (yr)	CT-ratio	QRS duration (sec)	LVFS (%)	TAPSE (mm)
1	18	8	0.58	0.14	24	13.4
2	14	8	0.61	0.14	35	14.3
3	13	9	0.64	0.16	23	13.8
4	12	7	0.63	0.14	17	14.1
5	17	9	0.65	0.18	28	16.7
Mean	14.8	8.2	0.62	0.15	25.5	15.8
SD	3.4	0.8	0.07	0.01	5.3	0.97

CT-ratio : cardio-thoracic ratio, LVFS : left ventricular fractional shortening,

TAPSE: tricuspid annular plane systolic : excursion

during initial repair, RV outflow tract incompetent, and the presence or not of the RV restrictive physiology(26-28). The present study reports normal right and left ventricular systolic function without right ventricular restrictive physiology during 10 years follow-up regardless of the severity of PR. However, 10 per cent of patients with moderate or severe PR developed RVOT aneurysm. These patients had greater CT-ratio, lower LVFS, as well as shorter TAPSE compared to patients without aneurysm. In the authors' opinion, those who had RVOT aneurysm should be closely observed for ventricular dysfunction and ventricular arrhythmia.

QRS lengthening seen after repair of TOF is the effect of surgical injury on myocardium and right bundle branch(28). QRS duration of 180 m.sec or more was predictive of ventricular tachycardia and sudden death(28). In the present study, only 1 patient with severe PR developed QRS duration more than 180 m.sec 9 years after surgery, but he had no history of syncope and was in NYHA class I without evidence of arrhythmia in 12-lead ECG. This showed

that cardiac arrhythmia was not the major problem in the mid-term period after repair of TOF as in previous studies(28,29).

There were no correlations between the severity of PR and the characters of ECG and CXR after repair of TOF. So, it would not be appropriate if only these two investigations in the follow-up of patients with total correction of TOF were relied on.

In conclusion, the pericardial monocusp could neither reduce the severity of pulmonary insufficiency nor improve right and left ventricular functions after 3 years follow-up post-operatively. Nonetheless, the right and left ventricular performances in mid-term period remained insignificantly changed and the severity of PR could not be predicted from symptoms and simple laboratory investigations.

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การทำงานของเวนทรีเคิล ในผู้ป่วย Tetralogy of Fallot หลังได้รับการผ่าตัดโดยใช้ pericardial monocusp

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ปัญหาการรั่วของลิ้นหัวใจพัลโมนารี (pulmonary regurgitation, PR) เป็นปัจจัยสำคัญที่ทำให้เวนทรีเคิลขวาทำงานผิดปกติ หรือมีเวนทรีเคิลเต้นผิดจังหวะในผู้ป่วย tetralogy of Fallot (TOF) หลังได้รับการผ่าตัดในช่วง 5 ปีที่ผ่านมา โรงพยาบาลรามคำแหงใช้การผ่าตัดด้วยการใช้เยื่อหุ้มหัวใจของผู้ป่วย ประดิษฐ์ขึ้นเป็นลิ้นหัวใจเทียม (pericardial monocusp) เพื่อช่วยลดการรั่วของลิ้นหัวใจนี้

วัตถุประสงค์ : เพื่อศึกษาความสามารถในการทำงานของ pericardial monocusp ความสามารถในการบีบและคลายตัว (systolic and diastolic function) ของเวนทรีเคิลทั้งสองด้านในผู้ป่วย TOF หลังผ่าตัดแก้ไข ด้วยคลื่นเสียงสะท้อนหัวใจ (echocardiogram) และศึกษาความสัมพันธ์ระหว่างการรั่วของลิ้นหัวใจพัลโมนารีกับอาการแสดง, ลักษณะคลื่นไฟฟ้าหัวใจ (electrocardio-gram) และภาพถ่ายรังสีปอด

วิธีการศึกษา : เป็นการศึกษาแบบ cross-section ในผู้ป่วย TOF ที่ได้รับการผ่าตัดแก้ไขด้วยวิธี transannular patch จากโรงพยาบาลรามคำแหง แล้วเป็นเวลาอย่างน้อย 1 ปี การศึกษาแบ่งผู้ป่วยเป็น 2 กลุ่ม คือ ผู้ป่วยกลุ่มที่ 1 และกลุ่มที่ 2 เป็นผู้ป่วยที่ได้รับการผ่าตัดโดยใช้และไม่ใช้ pericardial monocusp ตามลำดับ ผู้ป่วยแต่ละรายจะได้รับการซักประวัติและตรวจร่างกาย ร่วมกับการตรวจคลื่นไฟฟ้าหัวใจ ภาพรังสีปอด และคลื่นเสียงสะท้อนหัวใจโดยวัด right and left ventricular end-diastolic dimension ratio (RV/LV EDD ratio) และ fractional shortening ของเวนทรีเคิลซ้าย (LVFS) เพื่อประเมินความสามารถในการบีบตัวของเวนทรีเคิลซ้าย ตรวจ tricuspid annular plane systolic excursion (TAPSE) เพื่อประเมินความสามารถในการบีบตัวของเวนทรีเคิลขวาและตรวจหา diastolic antegrade flow across pulmonary artery ซึ่งแสดงถึงความผิดปกติในการคลายตัวของเวนทรีเคิลขวา ความรุนแรงของ PR จะแบ่งเป็นระดับน้อย ปานกลาง และรุนแรง โดยการตรวจ color Doppler

ผลการศึกษา : มีผู้ป่วยในการศึกษา 64 ราย เป็นผู้ป่วยในกลุ่มที่ 1 จำนวน 7 ราย และในกลุ่มที่ 2 จำนวน 57 ราย ค่ามัธยฐานของเวลาที่ได้รับการตรวจหลังการผ่าตัดเท่ากับ 3 และ 7 ปีในผู้ป่วยกลุ่มที่ 1 และ 2 ตามลำดับ จากการตรวจด้วยคลื่นเสียงสะท้อนหัวใจ พบว่า ผู้ป่วยกลุ่มที่ 1 มี RV/LV EDD ratio เฉลี่ยต่ำกว่าในกลุ่มที่ 2 (0.67 และ 0.83) แต่ไม่พบว่ามีนัยสำคัญทางสถิติ ($p > 0.01$) และพบว่าค่าเฉลี่ยของ LVFS, TAPSE และ RVEF ระหว่างทั้งสองกลุ่มไม่มีความ

แตกต่างกันอย่างมีนัยสำคัญทางสถิติเช่นกัน ($p > 0.01$) ผู้ป่วยทุกรายในกลุ่มที่ 1 และร้อยละ 80.4 ในกลุ่มที่ 2 มี PR ระดับปานกลางหรือรุนแรง การศึกษานี้ไม่พบว่ามี diastolic antegrade flow ในหลอดเลือดพัลโมนารี ในผู้ป่วยรายใดเลย มีผู้ป่วยที่ไม่มีลิ้นหัวใจพัลโมนารีรั่วหรือรั่วเพียงเล็กน้อยทั้งสิ้น 13 ราย (ร้อยละ 20.3) และมีลิ้นหัวใจรั่วระดับปานกลางถึงรุนแรงจำนวน 51 ราย (ร้อยละ 79.7) พบว่า CT-ratio และ QRS duration ของทั้ง 2 กลุ่มไม่ต่างกัน

บทสรุป : จากการติดตามผลในระยะเวลา 3 ปีในผู้ป่วย TOF หลังผ่าตัดแก้ไข การใช้ pericardial monocusp ไม่สามารถช่วยลดความรุนแรงของ PR หลังผ่าตัดไปแล้วได้ โดยที่การทำงานของเวนทริเคิลทั้งซ้ายและขวายังอยู่ในเกณฑ์ปกติ และการใช้ภาพรังสีปอดหรือการตรวจคลื่นหัวใจ ไม่สามารถคัดกรองผู้ป่วยที่มีลิ้นหัวใจพัลโมนารีรั่วระดับปานกลางหรือรุนแรงได้

คำสำคัญ : tetralogy of Fallot, การผ่าตัดแก้ไข, การรั่วของลิ้นหัวใจพัลโมนารี

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