Balloon Pulmonary Valvuloplasty in Tetralogy of Fallot: Effects on Growth of Pulmonary Annulus and Transannular Patch

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Background: *Balloon pulmonary valvuloplasty and its benefit on the growth of pulmonary annulus and pulmonary artery in tetralogy of Fallot (TOF) patients remains controversial. The purpose of the present study was to determine the growth of pulmonary valve annulus and pulmonary artery and to evaluate the need of transannular patch during total surgical correction in patients with tetralogy of Fallot (TOF) after balloon pulmonary valvuloplasty.*
Material and Method: *Fifty-one severely hypoxic TOF patients, who underwent balloon dilatation of pulmonary valve, were included in the present study. The clinical outcomes, oxygen saturation, and echocardiographic parameters before and after balloon dilatation were analyzed.*
Results: *There were 33 males and 18 females. The mean age was 3 years 5 months old (range, 1 month-15 years 8 months old). The mean oxygen saturation increased from 73.9 $\pm$ 9.1 to 84.8 $\pm$ 6.7% immediately after the procedure (p-value < 0.05). There was no serious procedural-related complication. At the mean follow-up period of 2 years and 4 months, the mean Z-score of pulmonary annulus size increased from -2.56 SD to -1.87 SD (p-value < 0.05) and the right pulmonary artery size from -0.29 SD to + 0.46 SD (p-value < 0.05). Thirty-seven patients (66.1%) underwent corrective surgery. Pulmonary transannular patch was performed in 11 of 37 patients (29.7%).*
Conclusion: *Pulmonary valve dilatation in patients with TOF is safe. It promotes the growth of the pulmonary valve annulus and pulmonary artery and may decrease the need of transannular patch at the time of surgical correction.*

Keywords: *Balloon pulmonary valvuloplasty, Tetralogy of Fallot (TOF)*

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The role of balloon pulmonary valvuloplasty as a palliative procedure in tetralogy of Fallot (TOF) remains controversial. Its effect on the growth of pulmonary valve annulus and pulmonary arteries is questionable. In TOF with small pulmonary valve annulus, transannular patch is needed in total surgical correction[1-6]. Nevertheless, potential complications due to pulmonary regurgitation after transannular patch have been reported in long term follow-up after surgery. These include right ventricular dilatation and dysfunction, ventricular tachycardia, and a higher risk of sudden death[7,8]. Balloon pulmonary valvuloplasty in TOF may promote the growth of the pulmonary annulus and pulmonary arteries and may decrease the incidence of transannular patch in total repair of TOF.

In the present study, the authors analyzed result of balloon dilatation of pulmonary valve in patients with TOF in our cardiac center. The purposes were to determine the growth of pulmonary valve annulus and pulmonary arteries after balloon dilatation and to evaluate the intermediate results including clinical outcomes after total surgical correction.

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Material and Method

Between 1996 and 2006, there were 56 TOF patients who underwent balloon dilatation of the pulmonary valve in Cardiac Center, King Chulalongkorn Memorial Hospital. The medical records and echocardiographic data were available for retrospective study in 51 patients (18 female and 33 male). The mean age at balloon valvuloplasty was 3 years and 5 months old (ranged from 1 month to 15 years 8 months old) and the mean weight was 13.4 kg (ranged from 3.2 to 44.6 kg). Two patients had Down's syndrome and one was diagnosed with Beckwith-Wiedemann syndrome. Balloon dilatation of the pulmonary valve was performed as a palliative procedure because of marked cyanosis with the mean arterial oxygen saturation of 73.9 $\pm$ 9.1% (ranged from 50% to 88%) and patient's profiles not suitable for total surgical repair. The technique of balloon pulmonary valve dilation was the same as balloon valvuloplasty in valvular pulmonary stenosis. The balloon size was about 1 to 1.5 times of the pulmonary annulus diameter measured by angiography in lateral view.

Transthoracic echocardiographic study was performed to compare the pulmonary annulus and pulmonary arterial sizes between before balloon pulmonary valvuloplasty and during follow-up after the procedure. All parameters were transformed to Z-score in the number of standard deviation (SD) from the mean normal value indexed by body surface area. Echocardiographic study was also performed in the patients after total surgical correction. The data included the sizes of the pulmonary annulus and pulmonary arteries, pressure gradient across right ventricular outflow tract, degree of pulmonary valve regurgitation by color flow quantifications (grading as 1+ to 3+), right ventricular ejection fraction by Modified Simpson's rule, left ventricular ejection fraction by Modified Simpson's rule, right ventricular end diastolic volume, right ventricular dilatation index (ratio of right and left ventricular end diastolic dimension, normal value < 0.5)[9], right ventricular length, and right ventricular dimension.

Statistical analysis

Continuous variables were compared by t-test. Categorical variables were compared by Pearson's Chi-square or Fischer's exact tests as appropriate. A probability p-value of < 0.05 was considered significant.

Results

After balloon dilatation, the mean arterial oxygen saturation increased from 73.9 $\pm$ 9.1 to 84.8 $\pm$ 6.7% immediately after the procedure (p-value < 0.05). There was no serious complication related to the procedure. However, nine of 51 patients (17.6%) did not respond to balloon valvuloplasty and developed severe hypoxia while waiting for surgical total correction. These nine patients needed Blalock-Taussig shunt.

Of the available data in 51 patients, 37 patients had total surgical correction. Eight patients were waiting for corrective surgery (two with Down syndrome, one with Beckwith-Wiedemann syndrome). One patient died from underlying biliary atresia and sepsis. Five patients were lost to follow-up.

At the mean follow-up period of 2 years and 4 months post balloon dilatation, the mean diameters of pulmonary annulus, and left and right pulmonary arteries increased significantly. There was also a significant increase of the mean Z-score of the pulmonary annulus and right pulmonary artery, but not the left pulmonary artery (Table 1).

The mean age at complete correction in the 37 patients was 6 years and 8 months old (range from

Table 1. The mean diameter and mean Z-score of pulmonary valve annulus, left and right pulmonary artery before and after balloon pulmonary valvuloplasty (at the mean follow-up of 2 years 4 months)

			Mean diameter (mm)	p-value	Mean Z-score (SD)	p-value
PV	Pre	n = 37	8.33 $\pm$ 2.70	<0.05	-2.56 $\pm$ 1.35 SD	<0.05
	Post	n = 33	10.68 $\pm$ 3.30		-1.87 $\pm$ 1.47 SD	
LPA	Pre	n = 29	7.37 $\pm$ 2.97	<0.05	+0.07 $\pm$ 1.73 SD	0.66
	Post	n = 29	8.56 $\pm$ 2.79		+0.23 $\pm$ 1.50 SD	
RPA	Pre	n = 30	7.31 $\pm$ 2.55	<0.05	-0.29 $\pm$ 1.48 SD	<0.05
	Post	n = 30	8.99 $\pm$ 2.39		+0.46 $\pm$ 1.40 SD	

PV = pulmonary valve annulus; LPA = left pulmonary artery; RPA = right pulmonary artery; SD = standard diviation

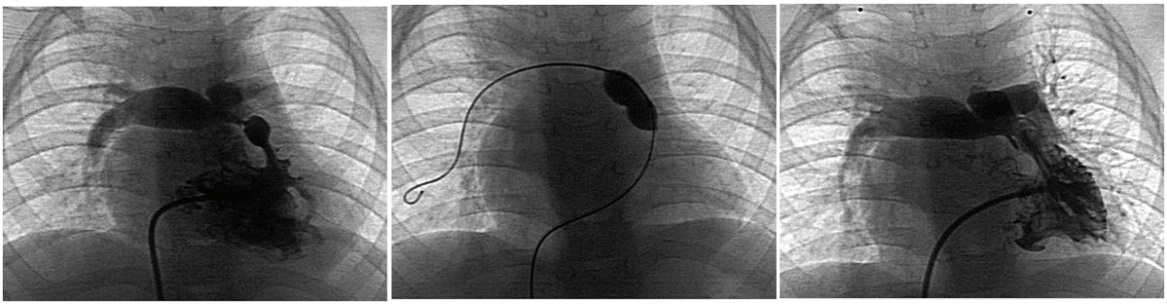

Fig. 1 Right ventriculograms in tetralogy of Fallot patients, pre and post balloon pulmonary valvuloplasty. (left, pre-balloon) there was severe pulmonary valvular stenosis with hypertrophic narrowing of the infundibulum; (middle) the balloon was inflated to dilate the pulmonary valve; (right, post-balloon) the pulmonary valve was wide open

9 months to 16 years and 7 months old), and the mean duration after balloon pulmonary valvuloplasty was 3 years and 8 months (range from 3 years to 4 years and 5 months). The patients were divided into two groups. Group A, composed of 26 patients (70.3%), did not require transannular patch during total correction due to good size of pulmonary valve annulus. The other 11 patients (29.7%) in group B had small annulus and required transannular patch. The mean Z-score of the pulmonary annulus measured by angiography after balloon pulmonary valvuloplasty in group A was -1.36 SD, compared with the mean of -2.24 SD in group B (p-value of 0.08).

Echocardiographic study after surgical correction was available in 25 patients (67.6%). There were 18 patients in group A and seven patients in group B. The mean duration after surgical repair was 4 years and 6 months. The mean age was 11 years and 2 months old (range from 2 years and 1 month to 22 years and 11 months old). The comparison between transannular patch and non-transannular patch patients showed no significant difference between all echocardiographic parameters except for the Z-score of the right pulmonary artery diameter, which was greater in group B (p-value of 0.05). None of the patients had moderate or severe aortic and mitral regurgitation. Color flow Doppler revealed the degree of pulmonary regurgitation as followed; group A: 1+, n = 5; 2+, n = 9; 3+, n = 4, and group B: 2+, n = 2; 3+, n = 5.

Discussion

The palliative treatment in severe hypoxic TOF patients whose anatomy was not suitable for total correction is systemic to pulmonary artery shunt. However, there were literatures[1-6] about good results of balloon pulmonary valvuloplasty as an alternate palliative method in such patients. Before the year 2005, there was a long waiting time for cardiac surgery for cardiac children in Thailand. Khongphattanayothin A[10] reported that the median waiting time for TOF patients was 452 days (1.24 year) and more than 10% of these patients waited for more than 4 years for total correction. The delay in systemic to pulmonary artery shunt depended on facility of each medical center. This resulted in mortality of about 20% while the patient was waiting for Blalock-Taussig shunt[10]. After shunting, hypoplastic pulmonary annulus remained relatively small[11] and the need for transannular patching was not always reduced at the time of complete repair[12]. The advantage of balloon pulmonary valvuloplasty is not only to increase blood flow to the lungs with relief of cyanosis but may also promote the growth of the pulmonary annulus in order to reduce the need for transannular patching at the time for complete correction[5]. Although this technique was associated with some complications[6], the previous studies had shown that this technique was safe without significant complications and could reduce the severity of cyanosis[1-4]. Qureshi SA reported a series of 15 TOF patients treated by balloon dilatation. The authors noticed an increase in systemic oxygen saturation and a decrease in the systolic gradient across the right ventricular outflow tract. In 70% of patients, corrective operation could be delayed or no further palliation was required in the follow-up period[1]. Sreeram N reported on a wider series of patients. Most of the patients in the present study had significant immediate improvement in systemic arterial oxygen saturation, a significant increase in pulmonary annulus diameter at follow-up angiography and the complete correction could be delayed[2]. Sluysman T reported the reduced incidence of

transannular patching in TOF patients who underwent balloon dilation of pulmonary valve[3]. The presented results are similar to many previous reports in improvement of pulmonary annulus diameter, reduction of cyanosis and no significant complication. Seventy percent of the presented patients who underwent total surgical correction did not need transannular patch.

Transannular patch is generally not preferred in complete correction of TOF because it may increase the frequency of re-operation due to significant patch-induced pulmonary regurgitation. Commonly, the transannular patch was needed when Z-score of the pulmonary valve annulus was -3 SD or less and could be avoided when the Z value was greater than -2SD[6]. In the present study, the mean Z-score of the pulmonary annulus after balloon dilation in group A (-1.36 SD) was greater than that in group B (-2.24 SD), but there was no statistical difference between means of both groups (p-value = 0.08). The incidence of transannular patch during total repair of TOF was reported to be from 36-100%[6]. Only 30% of our cases needed transannular patch. Thus, the balloon dilatation of pulmonary valve may be beneficial in decreasing the incidence of transannular patch.

Ho KW reported the late complications after TOF repair (mean postoperative follow-up duration of 23.5 $\pm$ 12.1 years)[13]. The most common complications were pulmonary regurgitation, residual RVOT obstruction, VSD patch leakage, and arrhythmias. Pulmonary regurgitation can produce progressive right ventricular dilatation, reduction in exercise capacity, arrhythmia, and sudden death. With the mean post-operative follow-up duration of 4.6 $\pm$ 2.7 years in the presented cases, the color Doppler echo-cardiography demonstrated evidence of pulmonary regurgitation in all of our patients. The presented data could not clearly demonstrate the difference in the degree of severity of pulmonary regurgitation after corrective surgery between the groups with and without transannular patch. In addition, there was no significant difference in echocardiographic parameters between both groups. Finally, the outcome of complete TOF repair, after balloon pulmonary valvuloplasty, in the presented cases was quite good. None of the patients underwent any re-operations.

Conclusion

Pulmonary valve dilatation in patients with TOF is safe. It promotes the growth of the pulmonary valve annulus and pulmonary artery and may decrease the incidence of transannular patch use at the time of total correction.

References

1. Qureshi SA, Kirk CR, Lamb RK, Arnold R, Wilkinson JL. Balloon dilatation of the pulmonary valve in the first year of life in patients with tetralogy of Fallot: a preliminary study. Br Heart J 1988; 60: 232-5.
2. Sreeram N, Saleem M, Jackson M, Peart I, McKay R, Arnold R, et al. Results of balloon pulmonary valvuloplasty as a palliative procedure in tetralogy of Fallot. J Am Coll Cardiol 1991; 18: 159-65.
3. Sluysmans T, Neven B, Rubay J, Lintermans J, Ovaert C, Mucumbitsi J, et al. Early balloon dilatation of the pulmonary valve in infants with tetralogy of Fallot. Risks and benefits. Circulation 1995; 91: 1506-11.
4. Hwang B, Lu JH, Lee BC, Hsieng JH, Meng CC. Palliative treatment for tetralogy of Fallot with percutaneous balloon dilatation of right ventricular outflow tract. Jpn Heart J 1995; 36: 751-61.
5. Kreutzer J, Perry SB, Jonas RA, Mayer JE, Castaneda AR, Lock JE. Tetralogy of Fallot with diminutive pulmonary arteries: preoperative pulmonary valve dilation and transcatheter rehabilitation of pulmonary arteries. J Am Coll Cardiol 1996; 27: 1741-7.
6. Godart F, Rey C, Prat A, Muilwijk C, Francart C, Vaksmann G, et al. Early and late results and the effects on pulmonary arteries of balloon dilatation of the right ventricular outflow tract in tetralogy of Fallot. Eur Heart J 1998; 19: 595-600.
7. Gatzoulis MA, Balaji S, Webber SA, Siu SC, Hokanson JS, Poile C, et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. Lancet 2000; 356: 975-81.
8. Ghai A, Silversides C, Harris L, Webb GD, Siu SC, Therrien J. Left ventricular dysfunction is a risk factor for sudden cardiac death in adults late after repair of tetralogy of Fallot. J Am Coll Cardiol 2002; 40: 1675-80.
9. Borowski A, Ghodsizad A, Litmathe J, Lawrenz W, Schmidt KG, Gams E. Severe pulmonary regurgitation late after total repair of tetralogy of Fallot: surgical considerations. Pediatr Cardiol 2004; 25: 466-71.
10. Khongphatthanayothin A, Layangool T, Sittiwangkul R, Pongprot Y, Lertsapcharoen P, Mokarapong P. Pediatric heart surgery waiting

time in Thailand and its effect on mortality: a cooperative study from Chulalongkorn, Children and Chiang Mai University hospitals. J Med Assoc Thai 2005; 88 (Suppl 4): S23-9.
11. Cotrufo M, Arciprete P, Caianiello G, Fittipaldi O, de Leva F, Violini R, et al. Right pulmonary artery development after modified Blalock-Taussig shunt (MBTS) in infants with pulmonary atresia, VSD and confluent pulmonary arteries. Eur J Cardiothorac Surg 1989; 3: 12-5.
12. Pacifico AD, Kirklin JW, Blackstone EH. Surgical management of pulmonary stenosis in tetralogy of Fallot. J Thorac Cardiovasc Surg 1977; 74: 382-95.
13. Ho KW, Tan RS, Wong KY, Tan TH, Shankar S, Tan JL. Late complications following tetralogy of Fallot repair: the need for long-term follow-up. Ann Acad Med Singapore 2007; 36: 947-53.

การขยายลิ้นหัวใจพูลโมนารีด้วยบอลลูนในผู้ป่วย tetralogy of Fallot: ผลต่อการเติบโตของลิ้นพูลโมนารี และการทำ transannular patch

อิงคนิจ ชลไกรสุวัฒน์, พรเทพ เลิศทรัพย์เจริญ, อภิชัย คงพัฒนะโยธิน, วิทวัส ลออคุณ, กัญญลักษณ์ วิเทศสนธิ, วิชัย เบญจชลมาศ, จุล นำชัยศิริ

วัตถุประสงค์: *เพื่อทำการศึกษาผลของการขยายลิ้นหัวใจพูลโมนารีด้วยบอลลูนในผู้ป่วย tetralogy of Fallot ที่มีต่อการเติบโตของลิ้นหัวใจและหลอดเลือดพูลโมนารี และความจำเป็นของการทำ transannular patch ในการทำผ่าตัดหัวใจเพื่อแก้ไขความผิดปกติ*

วัสดุและวิธีการ: *ได้ทำการศึกษาในผู้ป่วย tetralogy of Fallot จำนวน 51 ราย โดยได้นำระดับความอิ่มตัวของออกซิเจนในเลือด และผลการตรวจคลื่นเสียงสะท้อนหัวใจเพื่อวัดขนาดของลิ้นหัวใจและหลอดเลือดพูลโมนารีมาเปรียบเทียบ เพื่อศึกษาความแตกต่างระหว่างก่อนและหลังการขยายลิ้นหัวใจพูลโมนารีด้วยบอลลูน*

ผลการศึกษา: *มีผู้ป่วยเพศชาย 33 ราย และเพศหญิง 18 ราย อายุเฉลี่ย 3 ปี 5 เดือน ภายหลังการขยายลิ้นหัวใจทันที พบว่าค่าเฉลี่ยของระดับความอิ่มตัวของออกซิเจนในเลือดเพิ่มขึ้นจาก 73.9 เป็น 84.8% จากการศึกษาที่ระยะเวลาหลังการขยายลิ้นหัวใจเฉลี่ย 2 ปี 4 เดือน พบว่าค่า Z-score ของขนาดลิ้นหัวใจและหลอดเลือดพูลโมนารีเพิ่มขึ้นอย่างมีนัยสำคัญ ผู้ป่วยจำนวน 37 ราย ได้รับการทำผ่าตัดหัวใจเพื่อแก้ไขความผิดปกติ โดยมีผู้ป่วยจำนวน 11 ราย จำเป็นต้องทำ transannular patch*

สรุป: *การขยายลิ้นหัวใจพูลโมนารีด้วยบอลลูนในผู้ป่วย tetralogy of Fallot มีความปลอดภัยสามารถส่งเสริมการเติบโตของลิ้นหัวใจ และหลอดเลือดพูลโมนารี และอาจลดความจำเป็นในการทำ transannular patch ในการทำผ่าตัดหัวใจเพื่อแก้ไขความผิดปกติ*