

# Rastelli procedure in transposition of the great arteries associated with ventricular septal defect and pulmonary stenosis: A report of 3 consecutive cases in choray hospital

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## ABSTRACT

Transposition of the great arteries (TGA) is a complex cyanotic congenital heart disease, and only 10% of children with TGA can be alive through infancy. Rastelli operation is a standard surgical method for patient with TGA associated with ventricular septal defect (VSD) and pulmonary stenosis (PS). The aim of this report was to show short-term outcome of Rastelli operation in TGA; 3 patients (aged from 3 months to 4 years) were diagnosed TGA, VSD, atrial septal defect (ASD) and PS (2 severe pulmonary stenosis, 1 pulmonary atresia) with cyanosis, failure to thrive, baseline oxygen saturation values of 62% to 75%. All of 3 was performed Rastelli procedure and hospital discharged after 14 – 17 days postoperatively with saturation > 95%, no early complication, reoperation or death. Rastelli procedure in TGA associated with VSD, ASD and PS is initially well performed at the department of Pediatric Cardiac Surgery, Cho Ray hospital with satisfactory short-time outcome.

**Keywords:** Transposition of the great arteries, Rastelli procedure, pulmonary stenosis, pulmonary atresia.

## 1. INTRODUCTION

Transposition of the great arteries (TGA) is a complex cyanotic congenital heart disease

(CHD) that accounts for about 10% among CHD and patients with TGA and VSD accounts for 25% in all cases of TGA [1]. TGA is described as ventriculoarterial discordance that the aorta derives from left ventricle and the pulmonary artery (PA) derives from right one. This malposition separates circulation into 2 isolated parts: systemic and pulmonary circulations that oxygen-poor blood is pumped to systemic while oxygenated blood is pumped to lungs. Patients with TGA need another defect such as ventricular septal defect (VSD), atrial septal defect (ASD) to mix the blood between 2 separated circulations to improve systemic blood oxygenation. Without operations, only 10% of children with TGA can be alive through infancy.

Arterial switch operation now has been a gold standard of surgical technique for TGA in neonates that has excellent surgical result in neonatal period [2, 3]. Rastelli procedure was firstly performed in 1969 and now it has been become a standard surgical method to treat patient with TGA, VSD and pulmonary stenosis [4, 5]. In Rastelli procedure, VSD was closed and

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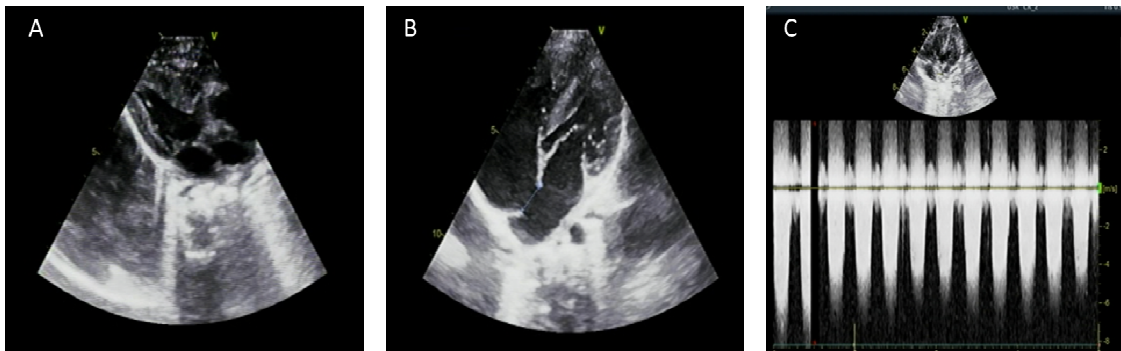
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using a right ventricle - pulmonary artery conduit with tissue valve inside to relieve pulmonary obstruction. Rastelli procedure may be a good choice for patients with TGA associated with VSD and pulmonary stenosis, who were not operated in neonatal period.

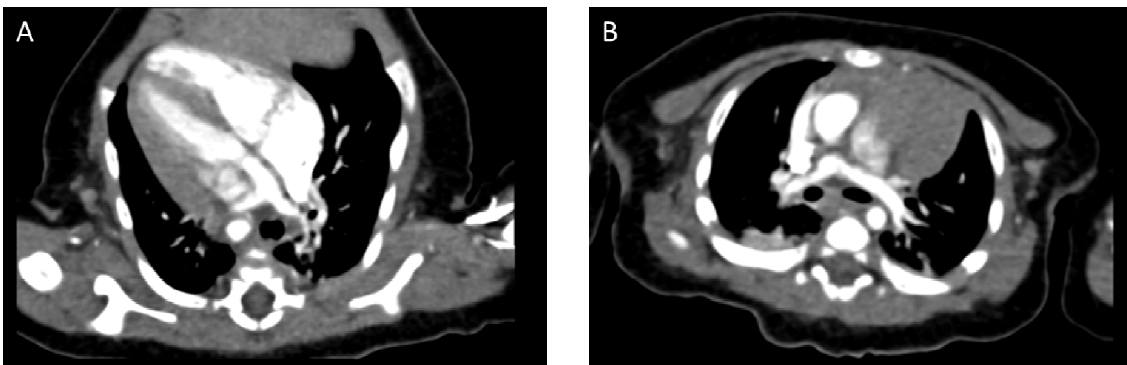
Rastelli procedure has been reported as the safe and effective operation with low mortality [6, 7]. However, Rastelli procedure is a complex cardiac surgery and it is only performed at some professional Heart center in Vietnam. The aim of this report was to show short-term outcome of Rastelli operation in 3 consecutive cases of TGA associated with VSD, ASD and PS, who were operated at the department of Pediatric Cardiac Surgery, Cardiovascular center, Cho Ray hospital.

## 2. PATIENTS AND METHODS

Since November 2021, 3 patients (aged from 3 months to 4 years) have undergone Rastelli procedure for the treatment of TGA, associated with VSD, ASD and pulmonary stenosis at our hospital. Echocardiography and computed tomography scan (CT-Scan) were performed for diagnosis. Echocardiography were used to evaluate the diameters of VSD and transpulmonary pressure gradient (PG) for determine whether pulmonary stenosis or not. Obstruction of right ventricular outflow tract (RVOTO) and left ventricular outflow tract (LVOTO) were identified with pressure gradient higher than 30 mmHg on echocardiography. Sizes of pulmonary artery and branches were firmly evaluated by contrast CT-scan.



**Figure 1:** Echocardiography in TGA diagnosis. (A) large VSD with two parallel great arteries. (B) large ASD. (C) transpulmonary pressure gradient for determine whether pulmonary stenosis



**Figure 2:** CT-scan in TGA diagnosis. (A) the pulmonary artery connects to the left ventricle. (B) pulmonary stenosis.

All patients were conducted to the open heart surgery with full sternotomy, cardiopulmonary bypass (CPB) supporting, antegrade cardioplegia with hypothermia at about 28 - 30°C intraoperatively. The VSD were closed with biologic patch, performed intraventricular tunnel as left ventricular outflow tract. The pulmonary artery were resected and oversaw nearly the annulus and were then reconstructed by valved conduit which connected from right ventricular infundibulum to pulmonary artery so that the valve locates near pulmonary bifurcation.

### 3. RESULTS

All patients presented with cyanosis, failure to thrive and systolic murmur grade 3/6 at lower left sternal border. The patent ductus arteriosus (PDA) defect help to maintain pulmonary blood flow in patients with pulmonary stenosis or atresia. 2 patients received Rashkind procedure that helps increase blood mixing in atrial floor had higher in baseline saturation than others. After surgery, the oxygen saturation (SpO<sub>2</sub>) of all patients were improved over 95%. Echocardiography before hospital discharge revealed no residual VSD, unobstructed right ventricular outflow tract. Patients discharged hospital from 8 to 17 days after operation with no complication.

#### 3.1. The first patient

Male patient (4 years-old, 13kg, 115cm) was diagnosed TGA after delivery and urgently then performed PDA stenting. At one month old, the patient was underwent Rashkind procedure by percutaneous intervention due to augmented cyanosis.

On before operated examination, he had the

fingers clubbing with cyanosis of the lips, SpO<sub>2</sub>#75%. Echography showed the dextro-TGA, membranous VSD d#17-18mm with bidirectional shunt, pulmonary stenosis with PG#90mmHg, ASD (because of Rashkind procedure) d#14mm with bidirectional shunt, mild enlarged right ventricular (Zscore#+3.0), normal left ventricular function (EF 75%), tricuspid annular plane systolic excursion (TAPSE) was 19mm. Chest CT-scan show the stenting PDA (d#7mm) and the size of pulmonary arteries (main PA#22mm, left PA#13mm, right PA#10mm). Cardiac intervention showed the normal PA pressure (PAPs#25mmHg), pulmonary vascular resistance index was 1.67 Wood unit.

The patient was underwent Rastelli procedure with No 15 conduit (Z-score#-0.26) with tissue valve inside, ASD closed and PDA disconnected. Time for CBP was 205 minutes, aortic cross clamp time was 150 minutes.

In intensive care unit (ICU), the mechanical ventilation time was 32 hours, Milronone (0.5µg/kg/min). Patient was discharged on 15<sup>th</sup> day after operation.

#### 3.2. The second patient

Male patient (3 months-old, 5kg, 59cm) was diagnosed TGA after delivery due to severe cyanosis and urgently then performed Rashkind procedure by percutaneous intervention.

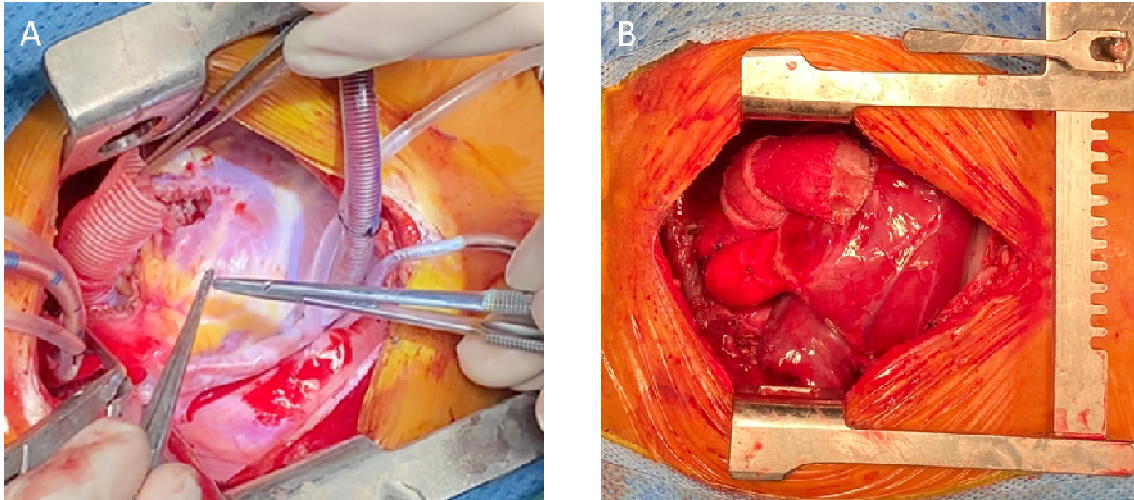
On before operated examination, SpO<sub>2</sub>#72%, echography showed the dextro-TGA, subarterial VSD d#9-10mm with bidirectional shunt, pulmonary stenosis with PG#140mmHg, ASD (because of Rashkind procedure) d#5mm with bidirectional shunt, PDA (d#3mm), normal left ventricular function (EF 73%), TAPSE was

10mm, left PA#5mm (Z-score#-0.98), right PA#5.2mm (Z-score#-0.14). Chest CT-scan show the size of PA 4mm (Zscore#-5.2).

The patient was underwent Rastelli procedure with No 12 conduit (Z-score#+1.7) with tissue valve inside, ASD closed. Time for

CBP was 205 minutes, aortic cross clamp time was 150 minutes.

In intensive care unit (ICU), the mechanical ventilation time was 96 hours, Dobutamine (0.5µg/kg/min). Patient was discharged on 17<sup>th</sup> day after operation.



**Figure 3:** Connect right ventricle - pulmonary artery conduit. (A) connect the valve conduit to right ventricle. (B) the valve locates near pulmonary bifurcation.

### 3.3. The third patient

Male patient (4 months-old, 5.6kg, 62cm), was hospitalized because of cyanosis (SpO<sub>2</sub>#72%). Echography showed the dextro-TGA with pulmonary atresia subarterial VSD d#8-9mm with bidirectional shunt, ASD d#10-11mm with bidirectional shunt, PDA (d#2mm), normal left ventricular function (EF 79%). Chest CT-scan show the size of PA 2.4mm (Zscore#-8.86), narrow pulmonary valve annulus (PVA) diameter (d# 2.4mm).

The patient was underwent Rastelli procedure with No 13 conduit (Z-score#+1.9) with tissue valve inside, ASD closed. Time for CBP was 240 minutes, aortic cross clamp time was 205 minutes.

In intensive care unit (ICU), the mechanical ventilation time was 72 hours, Dopamine (8µg/kg/min). Patient was discharged on 14<sup>th</sup> day after operation.

### 4. DISCUSSION

Rastelli operation is one of surgical methods for patients with TGA, large VSD, PA stenosis or whom TGA and large VSD that misdiagnosis or missed operation in neonates. Rastelli procedure has been reported as good surgical result in both short-term and long-term and low mortality. However, residual RVOTO, and LVOTO are still main indicators of reoperation and pulmonary artery hypertension should be considered.

Low early mortality has been reported in

many cardiac surgery centers, about 6% in the first 5-year, decreasing along with the increasing of experience in both surgical technique and intensive care [7].

Residual RVOTO is one of the common indications of reoperation after Rastelli operation. Huang and colleagues reported about 15% of patients have to replace conduit 5 years after RO [7]. Conduit sizing based on the body surface area is very important that relates reoperation. In our report, conduit size ranges from 12 to 15 (Z-score ranges from -0.26 to +1.9) and on the echocardiography before discharge showed no RVOTO. All patients should follow up frequently after operation to evaluate when the conduit becomes inadequate.

LVOT is created between VSD and aortic annulus by using biologic patch. A restrictive VSD can limit blood flow of LVOT. In some situations, with small VSD, surgeons may enlarge VSD by incising upper-anteriorly [7, 8]. In this report, VSD diameter ranges from 9-18mm so it is large enough and does not need to incise.

In neonatal period, high pulmonary vascular resistance helps left ventricle be trained under high pressure so that arterial switch operation can be done. After 2 months old, when pulmonary vascular resistance changes, TGA patients without protective factor (PA banding, stenosis or atresia) will develop pulmonary artery hypertension rapidly because of VSD [9]. In our report, all patients were diagnosed with PA stenosis or atresia and have PDA. All of them should be followed up to diagnose and treat pulmonary vascular disease as a consequence of overcirculation.

## CONCLUSION

Rastelli operation for patients with TGA, VSD and pulmonary stenosis is initially well performed at the department of Pediatric Cardiac Surgery, Cho Ray hospital with no early mortality, complications, or reoperation.

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