

# Successful total correction of Transposition of Great Artery in Surabaya's rural area experience: serial cases



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## ABSTRACT

**Background:** Transposition of the great arteries (TGA) is a cyanotic heart disorder characterized by the aorta being positioned to the pulmonary artery's right-anterior. Arterial switch operation (ASO) is the main surgical procedure for TGA correction that has favorable outcomes. This case report aims to explain the findings of TGA cases that were performed successfully by ASO surgery at Dr. Soetomo Hospital in Surabaya.

**Case Presentation:** We reported two cases of dextro-TGA, which were confirmed from anamnesis, physical examination and echocardiographic. Both cases have been successfully carried out with an arterial switch operation (ASO) procedure with a post-operative length of 14-15 days with stable hemodynamics.

**Conclusion:** TGA is a congenital heart defect with a rapid worsening of clinical signs if not detected early. The early correction will improve post-correction outcomes in patients.

**Keywords:** arterial switch operation, congenital heart defect, transposition of great artery, TGA total correction

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## INTRODUCTION

Transposition of the great arteries (TGA) is a heart defect condition when the connections between the large arteries in the heart are exchanged. In contrast to normal conditions, the aorta exits the right ventricle. It runs anterior to the pulmonary artery, while the pulmonary artery exits the left ventricle and runs posterior to the aorta. TGA is one of the most common cyanotic congenital heart defects, with a 5% prevalence of all congenital heart defects or approximately 30/100.000 live births.<sup>1,2</sup>

Diagnosis can be confirmed by asking the patient's history, physical examination and supporting procedures such as echocardiography and cardiac catheterization. Survival depends on the degree of mixing between the two circulations and the type of TGA. Patients with intact ventricular septum can survive because of aortopulmonary blood flow through the patent ductus arteriosus or are maintained by giving prostaglandin E1 and left-to-right bypass through the open foramen ovale due to high pressure of the

left atrium accompanied by an incompetent foramen ovale valve. Unfortunately, this mixing of blood can only meet a small amount of tissue oxygen demand, which does not respond to high concentrations of oxygen supplementation.<sup>3</sup> The primary treatment for TGA correction is arterial switch operation (ASO) that has favorable outcomes. A study by Anderson et al. suggested that an ASO procedure done on the 3rd day after birth could prevent left ventricular function deterioration, lowering post-operative complications and mortality. Hence, reduce hospital stay and costs effectively.<sup>4</sup> In this case report, we reported two cases of TGA surgery in newborns that were successfully performed at Dr. Soetomo Hospital in Surabaya.

## CASE PRESENTATION

### Case 1

A 3-month-old boy presents with a history of shortness of breath and a bluish discoloration of the skin that worsens with crying. The patient has a heart defect and is being treated with oral medications.

We found SpO<sub>2</sub> 63-78%, lips and limbs cyanosis, grade II-III murmur and Gallop on the 2<sup>nd</sup> ICS of the left parasternal line on physical examination. Other parameters are within normal limits. Laboratory tests are within normal limits. The patient was diagnosed with TGA-VSD and Moderate Pulmonary Hypertension, confirmed by echocardiography and cardiac catheterization as in [Figure 1](#).

We did ASO, closed VSD, and made PFO. The estimated duration of Box and CPB were 262 minutes and 239 minutes, respectively. We use histidine-tryptophan-ketoglutarate (Custodiol®) as cardioplegia. Post-operative results were evaluated through physical examination, chest X-ray and echocardiography. The patient was discharged with stable hemodynamic after 14 days.

### Case 2

A 1.5-month-old boy was treated with cyanosis since birth and was unable to cry after birth. Physical examination revealed SpO<sub>2</sub> 70-80%, cyanosis of the lips and limbs, a grade II-III holosystolic murmur was heard on ICS-2 left parasternal line.

Other parameters are within normal limits. Laboratory tests are still within normal limits. The patient diagnosed with TGA-IVS with small ASD without pulmonary hypertension, confirmed by echocardiography as in [Figure 2](#).

The surgical repair strategy is carried out in two stages; the first step is to perform

cardiac catheterization and balloon atrial septostomy (BAS) to improve intracardiac blood mixing. The next steps are ASO, PDA ligation and closure of ASD using primary sutures. The estimated duration of Box and CPB were 291 minutes and 214 minutes, respectively. We use histidine-tryptophan-ketoglutarate (Custodiol®) as

cardioplegia. Post-operative results were evaluated through physical examination, chest X-ray and echocardiography. The patient was discharged with stable hemodynamic after 15 days.

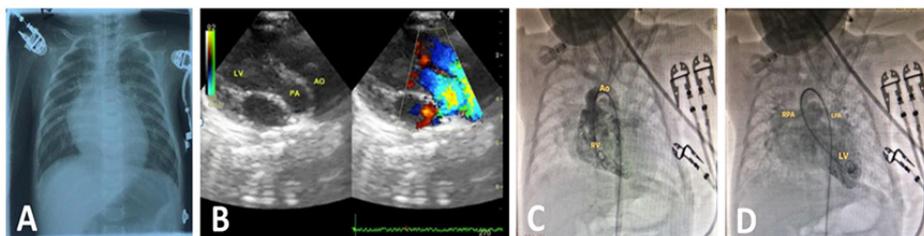
## OUTCOMES

No post-operative complications were reported in the first case. Six months after surgery, the patient showed clinical improvement with normal growth and development, as in [Figure 3](#). The second patient, seven days post-operatively showed signs of pneumonia and was treated with antibiotics based on sputum culture. Pneumonia resolved within five days, as shown in [Figure 4](#). Both patients fully recovered and continued at the outpatient clinic.

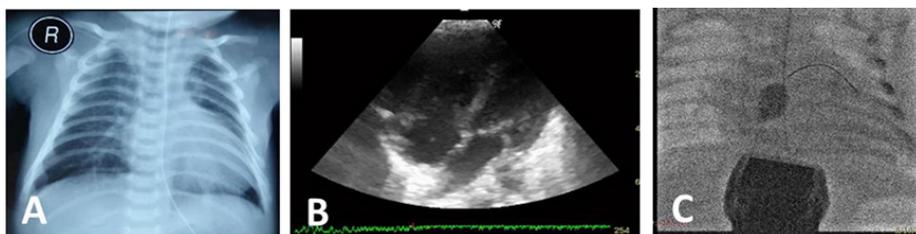
## DISCUSSION

We reported 2 cases of TGA handled by the Cardiac Surgery team at RSUD Dr. Soetomo Surabaya as in [Table 1](#). Both patients were male with an age range of 1.5-3 months—both cases presented with complaints of blue lips and fingertips that worsens with crying. TGA is a congenital heart defect with a high incidence compared to other types of heart defects, which is about 9.9% of all types of congenital heart disease. Epidemiologically, TGA is more common in boys or with a ratio of 3.2: 1.<sup>2</sup> We report two male infants with clinical findings that suggest congenital heart disease.

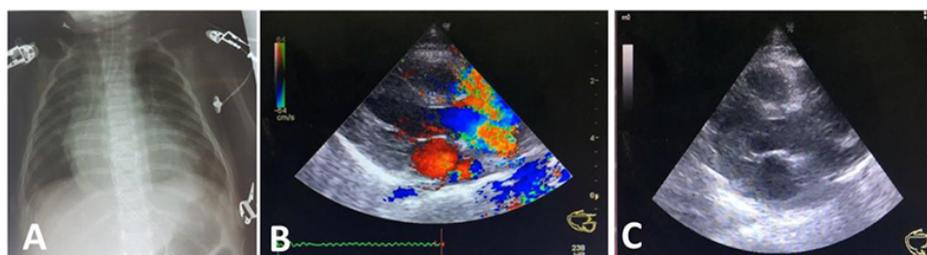
Diagnosis in both cases is based on a thorough physical examination and several investigations. From physical examination in both cases, abnormalities were found on the physical examination of the heart. The clinical appearance of patients with congenital heart disease depends on the type of defect. In TGA, patients will have central cyanosis. Clinical onset and severity vary widely depending on anatomical and physiological variations that affect the degree of mixing between the two circulatory circuits in the heart. Symptoms of congestive heart failure, tachypnea, tachycardia, diaphoresis, poor weight gain, gallop rhythm and hepatomegaly. In both cases, signs of central cyanosis were obvious. On auscultation of the heart, murmurs and gallops are heard,



**Figure 1.** Preoperative case 1. (A) Chest X-ray shows cardiomegaly and egg on string appearance; (B) Transesophageal echocardiography shows VA discordance; (C, D) Cardiac catheterization shows contrast filling on the RV that runs to the Aorta and fills the LV via a VSD



**Figure 2.** Preoperative case 2. (A) Chest X-ray shows cardiomegaly and egg on string appearance; (B) Transthoracic echocardiography showing VA discordance; (C) The patient has had a Balloon Atrial septostomy (BAS).



**Figure 3.** Post-Operative Evaluation Case 1. (A); (B); (C)



**Figure 4.** Post-operative evaluation case 2. (A) Chest X-ray evaluation seven days post-operatively showing a picture of pneumonia; (B) Chest X-ray evaluation 12 days post-operatively showing improvement of pneumonia after five days of antibiotic therapy; (C)

**Table 1. Summary of patients' clinical characteristics**

Cases :	Case 1	Cases 2
Sex/Age	K/ Male / 3 months	E/ Male/ 1.5 months
Chief Complaint	Lips and fingertips are blue when crying.	Lips and fingertips are blue when crying.
History of disease	None	None
Family history	None	None
Physical Exam.	SpO <sub>2</sub> 63-78%, Auscultation: murmur (+) and Gallop	SpO <sub>2</sub> 70-80%, Auscultation: murmur (+) holosystolic ICS II PSL (S) gr II/VI
Laboratorium	Hb 17.3 d/dL, HCT 55,9%	Hb 18.1 g/dL, HCT 55.8%
Radiology (Chest x-rays)	Cardiomegaly (CTR 60%), egg on a string appearance	Cardiomegaly (CTR 60%), egg on a string appearance
Echocardiography	d-TGA + Moderate perimembranous VSD + Mild MR	D-TGA with ASD Secundum L to R Shunt + moderate PDA post-BAS (46% pre-BAS saturation, 87% post-BAS saturation).
Intraoperative	- Normal size heart, looks bluish, sufficient contractility - Aorta from RV, MPA from LV - 15 mm perimembranous VSD	- Bluish heart, normal size, adequate contractility, anterior aorta MPA, MPA from LV, aorta from RV, a.coronaria 2 pieces right & left - 4 mm PDA, confluent RPA and LPA - ASD size 1 cm
Procedure	- Arterial switch - VSD closure	- Double ligation PDA - ASD closure with primary suture - Arterial switch - Insertion of the peritoneal dialysis tube
Follow-up	Intensive care for five days	Intensive care for 14 days

which leads to a suspicion of congenital heart disease. The finding of a murmur is generally associated with obstruction of the left ventricular outlet, PDA presence or due to septal defect.<sup>2</sup>

Clinical findings that lead to congenital heart disease suspicion must be determined according to the type and appropriate therapy. Plain photographs (x-rays) in some cases with typical characteristic imaging can help in the early stages. In TGA, the superior mediastinal space is narrowed, giving the appearance of egg-on-string or egg-on-its-tied. Cardiomegaly with increased pulmonary vascular patterns can be seen when accompanied by VSD. Electrocardiography is not very specific. The common ECG finding is a deviation of the QRS complex axis to the right associated with right ventricular hypertrophy. Biventricular hypertrophy can be found when the left ventricle has been overloaded.<sup>2</sup> Plain chest radiographs were performed on both patients. The plain radiograph supports the TGA findings in the presence of egg-on-string features and cardiomegaly. Plain photo examination was also carried out to eliminate the possibility of lung pathology. In both cases, the lungs

were within normal limits, and there was no increase in bronchovascular patterns. The electrocardiographic examination was performed in patient 2 with findings of sinus rhythm and deviation of the right QRS axis.

Diagnosis in both cases was confirmed after echocardiography examination found an atrioventricular concordance and a ventriculoarterial discordance. Echocardiography is still the definitive diagnostic tool in cases of heart defects. In the 4-chamber imaging, we can assess the atria-ventricle relationship. The TGA morphology can be best evaluated by a 5-chamber view, long-axis parasternal or subcostal view. The pulmonary artery is a branched artery (bifurcation) and posteriorly located that exits the left ventricle. At the same time, the aorta is characterized by a coronary head and neck in the right ventricle. The proximal parts of the two arteries run in parallel, which under normal conditions will cross so that it is quite easy to find from this examination. From the short-axis appearance, the pulmonary artery is in the middle, with the aorta located anteriorly and slightly to the right. Doppler studies

need to be carried out to confirm other defects. Catheterization was done when the operator still needed data about anatomy and hemodynamic aspects that could not be obtained from the echocardiographic examination.<sup>2,3</sup> Both of the cases we reported were complex TGA or TGA accompanied by other cardiac defects.

The type of lesion determines surgery in a TGA. The ASO (atrial switch operation) procedure is the procedure of choice used to achieve complete physiological and anatomical improvement. This procedure's advantages were associated with long-term outcomes characterized by good left ventricular function, sinus rhythm, and low mortality rates with a survival rate of 88% at 10 and 15 years. Rare post-operative complications are primarily associated with prolonged peri-operative ischemia, aortic regurgitation and coronary artery obstruction, leading to myocardial ischemia or even infarction. Low reoperation rates have also been reported, with pulmonary stenosis at the reconstruction site being the most common cause.<sup>2,5</sup>

In some studies, transposition with an

intact septum, newborn babies should be operated on in the first two weeks of life. Because at that time, the left ventricle had not experienced significant involution, and its contractility could still support systemic circulation. In older neonates and young infants, the left ventricle may need to be retrained before ASO is attempted, with a pulmonary artery band. Recent reports have suggested extending primary switch arterial operation limits in patients with TGA and intact ventricular septum from 2 weeks after birth to 2-3 months of age. This reduces the need for pulmonary artery banding and systemic-pulmonary artery bypass before primary correction, associated with pulmonary valve dilatation (neoortic valve) and neoartical insufficiency when the arterial switch is performed later. This strategy's implication is the need for longer post-arterial switch support because of the untrained left ventricle.<sup>5</sup>

Until the mid-twentieth century, the treatment of transposition was limited to a few palliative steps. The natural history of the disease with its poor prognosis was an undeniable fact. At that time, the mean life expectancy for the transposed newborn was 0.65 years, and the one-year mortality rate was 89.3%. With the emergence of newer and better surgical techniques and post-operative intensive care, the scenario has changed, and a very long-term survival rate of nearly 90% has been reported. A current corrective surgery modality potential shows a low 10-year re-intervention rate (6%) and 88% symptom-free survival.<sup>6</sup> Nonetheless, several recent studies have shown decreased exercise performance, impaired cognitive function, and unsatisfied life quality. Therefore, prenatal diagnosis is still an important key to reduce the complications of managing patients with congenital heart disease, especially TGA.<sup>7</sup>

## CONCLUSION

The very low mortality rate has replaced the neonates with a previous poor natural history with D-TGA. Most of the treated patients could live to adulthood, with a 20-year survival rate of nearly 90%. The outcome of a surgical procedure is determined by preoperative, intraoperative, and post-operative care. Dr. Soetomo General Hospital has successfully carried out 2 cases of TGA correction. Careful time management and selection of cases will further increase the success rate of TGA corrections, as shown in our case series, even though our facilities are limited. The treatment period for both patients was under three weeks, and they had fully recovered without further post-operative complications.

## CONFLICT OF INTEREST

The authors declare that there is no competing interest regarding the manuscript.

## ETHICAL CONSIDERATION

This research was conducted based on the ethical conduct of research from the Ethics Committee of the Airlangga University, Dr. Soetomo-Surabaya General Hospital.

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## AUTHOR CONTRIBUTION

All of the authors equally contributed to the study from the conceptual framework, data gathering, and data analysis until interpreting the study results on publication.

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