

# Conventional Repair of an Intraoperatively Diagnosed Congenitally Corrected Transposition of the Great Arteries in an Adult

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## ABSTRACT

**Background:** Congenitally corrected transposition of the great arteries (ccTGA) is a rare condition that accounts for just 1% of all congenital heart disease. Diagnosis of ccTGA often is missed in adulthood, despite imaging and cardiology consultation.

**Case report:** We present the case of an intraoperatively diagnosed ccTGA with severe tricuspid valve regurgitation and secundum atrial septal defect in a 54-year-old woman, who preoperatively was diagnosed with mitral valve regurgitation in atrioventricular and ventriculoarterial concordance heart. Intraoperatively, options considered were anatomical repair with atrial-arterial double switch operation after retraining the left ventricle or a conventional repair that focused on the associated defects without addressing the discordant connections. Considering our patient's age and condition, we decided to carry on with the conventional repair to prevent further systemic right ventricle dysfunction that may lead to poor outcome and decreased survival. She was discharged one week after surgery and resumed her normal activity at 3-month follow up.

**Conclusion:** Although it rarely happens, CHD such as ccTGA in an adult must always be considered. Careful examination is essential. The treatment of ccTGA in an adult is challenging, with more limited options compared with pediatric patients. However, early management could still provide favorable outcomes.

## INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare condition, accounting for just 1% of all congenital heart disease (CHD) and is characterized by atrioventricular (AV) and ventriculoarterial (VA) discordance [Baumgartner 2010]. To eliminate systemic right ventricle

(SRV) dysfunction and tricuspid regurgitation (TR), the anatomic repair to restore AV and VA concordance has been preferred over the conventional repair that focuses on addressing the associated defects while maintaining the SRV. However, results of anatomic repair are unfavorable in adults because of the inability to recondition the left ventricle (LV) [Baumgartner 2010; Dobson 2013].

We present a case of conventional repair on an intraoperatively diagnosed ccTGA with secundum atrial septal defect (ASD) in a 54-year-old woman.

## CASE REPORT

A 54-year-old woman was referred to our center with mitral valve (MV) regurgitation. She complained of dry cough, especially when lying down, and dyspnea on exertion over the past few months. No chest pain, palpitation, or previous history of heart disease was reported. Her physical function was classified as New York Heart Association functional class II. The heart rhythm was regular with 3/6 holosystolic murmur at the apex. No S3, rales, or peripheral edema was present.

Twelve-lead electrocardiogram showed sinus rhythm, left axis deviation, and P mitrale. Q waves were present in lead V1, II, III, and aVF. Chest radiograph exhibited cardiomegaly, and coronary angiogram revealed no coronary artery disease.

Initial transthoracic echocardiography examination (TTE) and transesophageal echocardiography examination (TEE) reported levocardia, left atrial (LA) dilatation, normal LV wall motion with ejection fraction (EF) of 63.8%, and moderate-severe MV regurgitation with prolapse of the posterior leaflet of the MV. (Figure 1) The examination also showed a secundum-type ASD with a diameter of 5 mm and a left-to-right shunt. (Figure 2) Our initial diagnosis was moderate-severe MV regurgitation with secundum-type ASD, and we decided to perform ASD closure with MV replacement.

After midsternotomy, we found that the aorta arose from the morphologic RV (mRV) while the pulmonary artery (PA) arose from the morphologic LV (mLV). (Figure 3) ccTGA was suspected and confirmed using intraoperative TEE that showed AV and VA discordance. There also was severe TV regurgitation with thickened and flail posterior TV leaflet. Both ventricles showed good function.

After immediate intraoperative discussion, we decided to carry on the surgery with TV replacement and ASD closure procedure. A mechanical valve (SJM Masters) was implanted

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with pledged sutures via LA incision. Before decannulation, the patient developed first degree AV block, prompting the need for temporary pacing wire insertion. Postoperative TEE revealed satisfactory repair with no TR and paravalvular leak.

One week after surgery, she had full resolution of symptoms and stable condition. She was discharged with anticoagulant, analgetics, beta blockers, and diuretics. At 3-month follow up, she had resumed her normal activity.

## DISCUSSION

Congenitally corrected transposition of the great arteries is a rare condition, accounting for just about 1% of all CHD and is characterized by AV and VA discordance [Baumgartner 2010]. Even without associated lesions, the survival of

ccTGA is poor, with only 50% at 40 years [Murtuza 2011]. In some cases, patients are asymptomatic and the diagnosis can go unrecognized until adulthood [Baumgartner 2010; Lampropoulos 2013]. The incidence of congestive heart failure and SRV dysfunction increases with age and by the age of 45 years, clinical heart failure is found on 67% of patients with associated abnormalities [Hornung 2010]. To our knowledge, our patient age was the oldest reported operated ccTGA in Indonesia to date.

Around 90% of ccTGA is accompanied by associated anomalies like ventricular septal defect (70%), pulmonary stenosis (40%), and systemic TV anomalies [Baumgartner 2010]. The TV is abnormal in 94% ccTGA patients and is frequently Ebstein's anomaly [Hornung 2010]. Atrial septal defect, as found in our case, is an uncommon finding in ccTGA. Another common finding in ccTGA is conduction system anomaly, due to the unusual position of AV node and bundle of His [Baumgartner 2010]. First degree AV block, as shown in our patient, is uncommon as most patients developed total AV block occurring at a rate of 2% per year [Baumgartner 2010; Oliver 2012; Talwar 2013].

Diagnosis of ccTGA often is missed in adulthood, despite

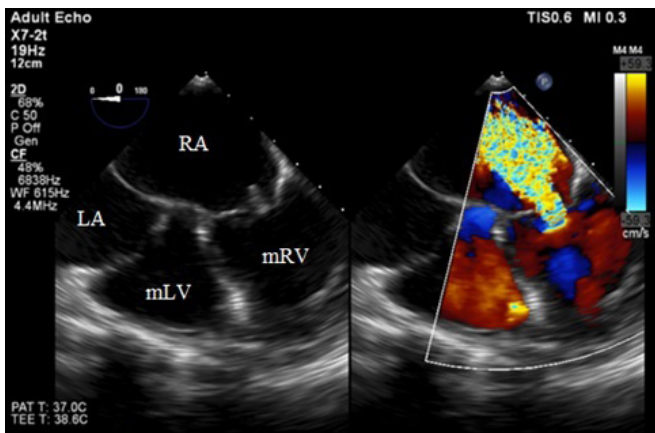


Figure 1. Four-chamber view of the TEE illustrating ccTGA with severe tricuspid valve (TV) regurgitation and flail posterior TV leaflet previously misinterpreted as a normal heart with MV regurgitation. Note the trabeculated morphologic right ventricle (mRV) on the left side of the heart. LA, left atrium; RA, right atrium; mLV, morphologic left ventricle; mRV, morphologic right ventricle

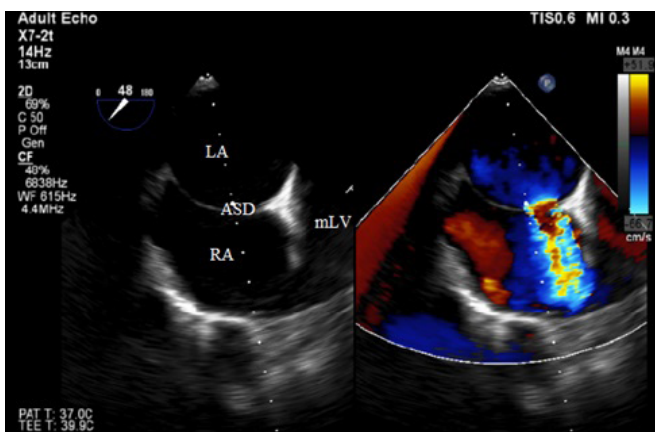


Figure 2. TEE demonstrating ASD with left-to-right shunt. LA, left atrium; RA, right atrium; mLV, morphologic left ventricle; ASD, atrial septal defect

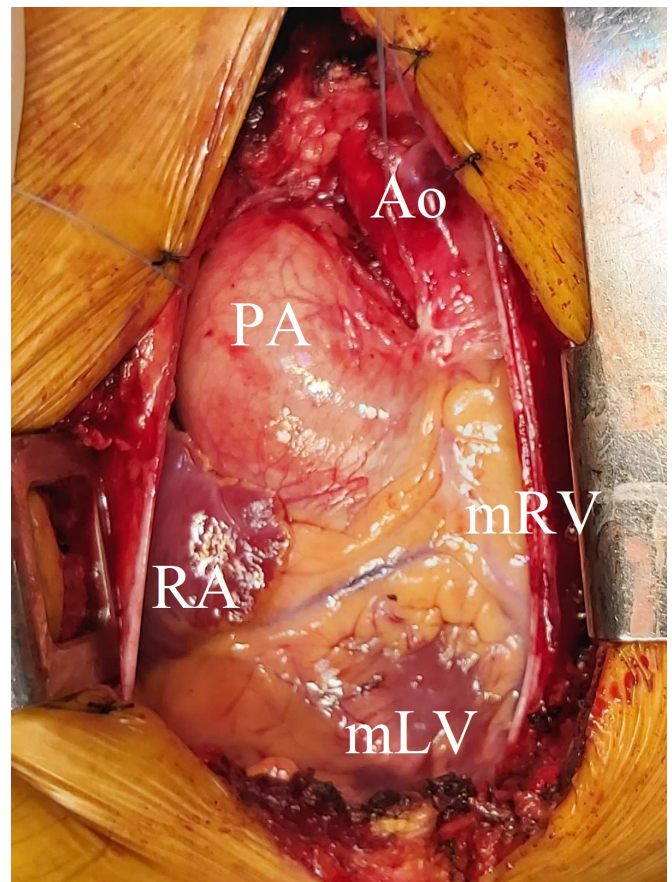


Figure 3. Intraoperative findings suggestive of ccTGA. Note that the aorta arose from the mRV while the PA arose from the morphological LV. PA, pulmonary artery; Ao, aorta; RA, right atrium; mLV, morphologic left ventricle; RV, right ventricle

imaging and cardiology consultation [Beauchesne 2002]. As it occurred in our case, we misinterpreted systemic TR in ccTGA as MV regurgitation. Our failure to diagnose this patient preoperatively led to intraoperative diagnosis and prompt on-site treatment modification.

Surgical treatment for ccTGA varies widely, according to individual conditions. Our patient presented with a SRV dysfunction and severe TR. Tricuspid regurgitation is a major contributor to the development of SRV dysfunction and surgery for severe regurgitation should be performed early before right ventricular ejection fraction <45% [Baumgartner 2010; Filippov 2016]. Intraoperatively, considered options were (a) anatomical repair with atrial-arterial double switch operation after retraining the LV, and (b) conventional repair that focused on the associated defects without addressing the discordant connections. Anatomic repair, which restores AV and VA concordance and repositions LV and MV into the systemic position, could treat or prevent the SRV failure. However, previous studies showed that age >12 years correlates with higher unsuccessful LV retraining and mortality [Baumgartner 2010; Filippov 2016; Poirier 2004]. Moreover, there is a risk of new systemic LV dysfunction after atrial-arterial double switch operation [Dobson 2013].

Considering our patient's age and condition, we decided to carry on with the conventional repair to prevent further SRV dysfunction, which may lead to poor outcome and decreased survival [Talwar 2013; Filippov 2016]. We proceeded with TV replacement due to tricuspid annular dilatation and abnormal TV leaflets not amenable to repair.

Various complications after repair have been reported, including progressive TR, SRV failure, and dysrhythmias, while death commonly is caused by SRV failure [Hornung 2010; Hsu 2016]. van Son reported that the 5-year and 10-year survival rate of patients with preoperative EF  $\geq$ 44% as in our case was 100%, and Hsu stated overall 16-year survival rate was 80% [Hornung 2010; Hsu 2016]. Nevertheless, we planned close monitoring for signs of SRV and systemic TR in addition to pharmacological support in this patient.

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