



# The first successful double switch operation for congenitally corrected transposition of the great arteries in Slovenia

Prva popolnoma uspešna poprava pri prirojeni popravljeni transpoziciji velikih arterij v Sloveniji

Miha Weiss,<sup>1</sup> Anže Djordjević,<sup>2</sup> Roman Gebauer,<sup>3</sup> Miroslav Elek,<sup>4</sup> Mirko Topalović,<sup>4</sup> Teja Senekovič,<sup>4</sup> Andraž Zupan,<sup>5</sup> Matija Jelenc,<sup>1</sup> Mirta Koželj<sup>4,6</sup>

## Abstract

Congenitally corrected transposition of the great arteries is a rare congenital heart defect characterized by atrioventricular and ventriculoarterial discordance and can be potentially associated with several other concomitant anomalies, such as ventricular septal defect or congenital complete atrioventricular block. Different surgical options are used for treating the condition. Anatomic repair yields the best long-term outcomes; however, it is technically the most challenging. We present a case of a 3-year old female patient who was electively admitted for complete repair after pulmonary artery banding and pacemaker implantation soon after birth. Subsequently, an anatomic repair (double switch) was performed with an uneventful postoperative course and excellent mid-term outcomes.

## Izveček

Prirojeno popravljena transpozicija velikih arterij je redka prirojena srčna napaka, pri kateri gre za neskladje med odnosi preddvorov in prekatov ter prekatov in izstopajočih velikih arterij (aorta, pljučna arterija). Pridružene so lahko tudi druge napake, kot sta okvara medprekatnega pretina ali prirojeni popolni preddvorno-prekatni blok. Obstaja več možnosti

<sup>1</sup> Department of Cardiovascular Surgery, Division of Surgery, University Medical Centre Ljubljana, Ljubljana, Slovenia

<sup>2</sup> Department of Cardiac Surgery, University Medical Centre Maribor, Maribor, Slovenia

<sup>3</sup> Division of Surgery, Children's Heart Center, 2nd Faculty of Medicine, Charles University in Prague and Motol University, Prague, Czech Republic

<sup>4</sup> Department of Paediatric Cardiology, University Medical Centre Children's Hospital Ljubljana, Ljubljana, Slovenia

<sup>5</sup> Institute of Radiology, University Medical Centre Ljubljana, Ljubljana, Slovenia

<sup>6</sup> Faculty of Medicine, University of Ljubljana, Ljubljana, Slovenia

**Correspondence / Korespondenca:** Anže Djordjević, e: [anze.djordjevic@ukc-mb.si](mailto:anze.djordjevic@ukc-mb.si)

**Key words:** congenitally corrected transposition of the great arteries; ventricular septal defect; congenital complete atrioventricular block; double switch procedure; congenital heart surgery

**Ključne besede:** prirojeno popravljena transpozicija velikih arterij; okvara medprekatnega pretina; prirojeni popolni preddvorno-prekatni blok; popolno kirurško popravilo; otroška srčna kirurgija

**Received / Prispelo:** 16. 10. 2020 | **Accepted / Sprejeto:** 13. 11. 2020

**Cite as / Citirajte kot:** Weiss M, Djordjević A, Gebauer R, Elek M, Topalović M, Senekovič T, et al.. The first successful double switch operation for congenitally corrected transposition of the great arteries in Slovenia. *Zdrav Vestn.* 2021;90(11–12):624–7. **DOI:** <https://doi.org/10.6016/ZdravVestn.3180>



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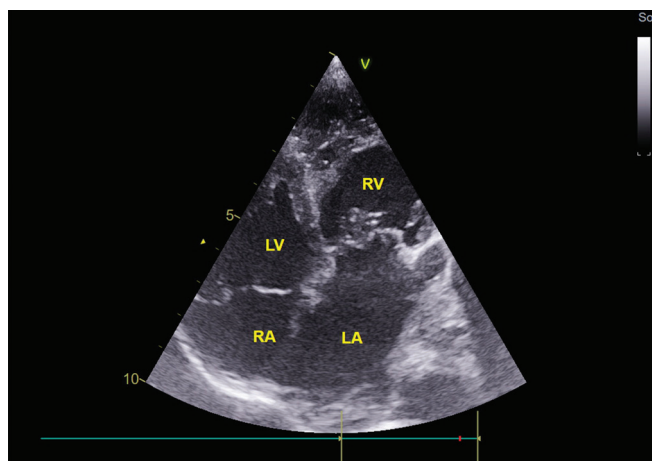
kirurškega zdravljenja. Popolno anatomsko popravilo prinaša najboljše dolgoročne rezultate, vendar je tehnično najzahtevnejše. Prikazujemo primer triletne deklice, ki je imela prvo operacijo kmalu po rojstvu (bandažiranje pljučne arterije, vstavitev srčnega spodbujevalnika), zdaj pa je bilo uspešno opravljeno popolno popravilo z odličnim srednjeročnim izidom zdravljenja.

## 1 Introduction

Congenitally corrected transposition of the great arteries (ccTGA) is a combination of discordant atrioventricular (AV) and ventriculoarterial connections (1). With an incidence of 1/33.000 live births ccTGA accounts for 0.05% of congenital cardiac malformations (2). Since both ventricles and great arteries (aorta and pulmonary artery (PA)) are transposed, normal physiology of blood flow is observed (3). Deoxygenated blood returns to the right atrium (RA) and through the mitral valve (MV) into the left ventricle (LV), pumping it through the pulmonary valve into the PA. Oxygenated blood returns to the left atrium (LA) and through the tricuspid valve (TV) into the right ventricle (RV), which in turn pumps it against systemic pressure through the aortic valve into the aorta. RV failure and/or TV regurgitation are the most common indications for surgical repair, followed by concomitant anomalies, such as ventricular septal defect (VSD), congenital complete AV block or LV outflow tract (LVOT) obstruction (4). Presenting with AV and ventriculoarterial discordance, it is plausible that these patients live without surgical corrections into adulthood due to almost physiological blood flow. Individuals with ccTGA present a broad spectrum of clinical presentations, from asymptomatic to heart failure or even sudden cardiac deaths (5). Transthoracic echocardiography with computed tomography or magnetic resonance angiography adding valuable information regarding coronary artery anatomy remains the golden diagnostic standard. VSDs are common in patients with ccTGA with reported incidence of 60% of all affected individuals (6). A dysplastic TV is found in 75% of cases with VSDs (3). The congenital AV block is the consequence of atrial and ventricular septal malalignment and extensive membranous septum filling the gap, thus preventing the AV node to lie appropriately in the triangle of Koch (3). Here, we present the first Slovenian successfully treated case of ccTGA managed at the University Medical Centre Ljubljana, Slovenia.

## 2 Case presentation

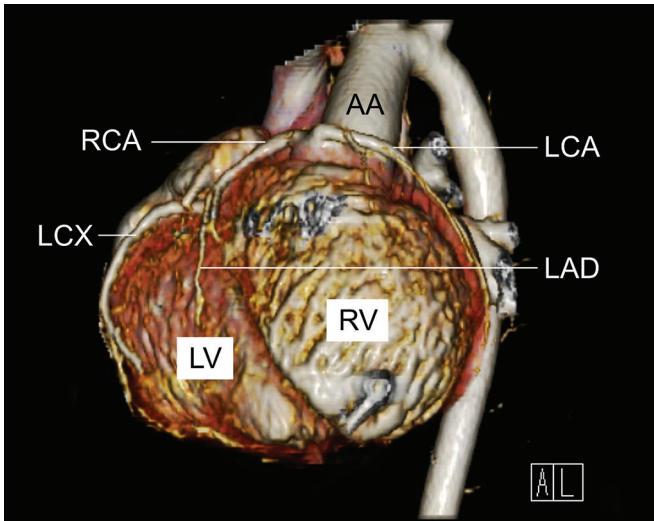
A 3-year old Caucasian female child with diagnosed ccTGA after PA banding (performed 2 days after birth) intended for LV pre-conditioning and a single chamber VVI pacemaker (PM) implantation (in order to spare



**Figure 1:** Preoperative transthoracic echocardiogram image in four-chamber view depicting atrioventricular and ventriculoarterial discordance.

Legend: LA – left atrium, RA – right atrium, LV – morphologically left ventricle, RV – morphologically right ventricle. Image is from authors' own archive.

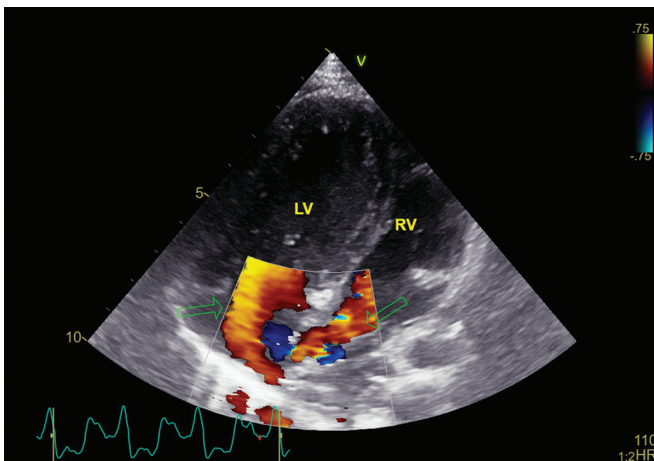
the atrial tissue until final correction) due to a congenital complete AV block was admitted to the Department of Paediatric Cardiology for definite treatment. Preoperative transthoracic echocardiogram showed an 8-mm large non-restrictive perimembranous VSD with a left-to-right shunt, a good functioning LV without obstruction in the LVOT, a hypertrophic RV with an ejection fraction of 65% and without obstruction in the RVOT, a dysplastic TV with moderate to severe regurgitation (indexed vena contracta 6 mm) and without stenosis (Figure 1). Abnormal coronary artery anatomy with a common origin of both coronaries from the left coronary sinus was confirmed on a computed tomography scan (Figure 2). There was no need to perform invasive coronary angiography (7). At surgery, a standardized double switch operation was performed (8). Standard median re-sternotomy with distal ascending aorta and bicaval cannulation was performed. Under mild hypothermia, the aorta was cross-clamped and the heart was arrested using antegrade cardioplegia. With the right transatrial approach, the perimembranous VSD was closed using a Dacron (a condensation polymer of ethylene glycol and terephthalic acid) patch. The MV was functioning well and did not require any surgical repair. Then, the aorta and PA were separated and a single coronary button was excised. A pericardial patch was used to reconstruct the neo-PA. The PA band was removed and



**Figure 2:** Computed tomography angiography image depicting a single origin of both coronary arteries with left anterior descending (LAD) arising from the right coronary artery.

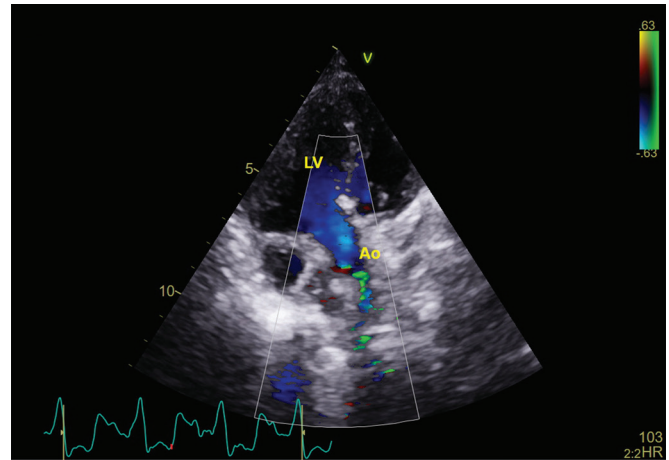
Legend: LCX – left circumflex artery, RCA – right coronary artery, LCA – left coronary artery, AA – ascending aorta. Image is from authors' own archive.

the coronary button was reimplemented into the neo-aorta. Both the neo-PA and neo-aorta were switched (without the Lecompte manoeuvre) and anastomosed to their respective counterparts. Finally, the Senning procedure or the atrial switch was performed. With the Senning surgical repair, a baffle is created within the atria, which reroutes deoxygenated blood from both caval veins to the



**Figure 3:** Postoperative transthoracic echocardiography image in four-chamber view with colour flow Doppler depicting pulmonary (left arrow) and systemic venous baffle (right arrow).

Legend: LV – morphologically left ventricle, RV – morphologically right ventricle. Image is from authors' own archive.



**Figure 4:** Postoperative transthoracic echocardiography image in four-chamber view with colour flow Doppler depicting ventriculoarterial concordance.

Legend: LV – morphologically left ventricle, Ao – aorta. Image is from authors' own archive.

MV and onwards to the pulmonary circulation. The baffle was constructed using RA and interatrial septal tissue as well as bovine pericardium. The right atriotomy was closed using *in situ* pericardium in order to prevent RA reduction. After de-airing, the cross-clamp was removed and the heart spontaneously reanimated. New atrial and ventricular epicardial PM leads were implanted. After weaning from cardiopulmonary bypass, haemostasis was achieved and the sternal wound closed in layers. The patient was extubated on the second postoperative day (POD) and discharged from the hospital on POD 12. Postoperative transthoracic echocardiography showed no obstruction in the atrial reconstruction, mild TV regurgitation, no other valvular pathology, appropriate biventricular function, no residual VSD, and no pericardial effusion (Figures 3 and 4). The anatomic restoration of AV and ventriculoarterial connections was achieved. Due to coronary artery reinsertion into the neo-aorta, acetylsalicylic acid at a dose of 1mg/kg was added to her therapy for 6 months.

### 3 Discussion

ccTGA is a rare clinical entity with unique characteristics. Adequate preoperative diagnostics (i.e. transthoracic echocardiography, computed tomography) is crucial for successful timing of the surgery and good long-term outcomes. During surgery, either biventricular

or univentricular surgical correction is indicated. Biventricular correction can be either physiological, meaning that only associated anomalies are corrected (VSD repair, TV replacement), and anatomical, which aims to restore anatomic relations, focusing especially on removing the RV from pumping into the systemic circulation (1). If an anatomic repair is not technically feasible, the Fontan palliative procedure is advocated with comparable mid-term results. Factors, precluding anatomic repair, are aberrant coronary artery anatomy, AV valves' straddling, dextrocardia or a very large area of septation required for VSD closure (9). We opted for an anatomic repair despite the coronary anomaly (single origin of both coronary arteries), partially because of favourable position regarding the neo-aorta (origin from the left coronary sinus). Anatomic repair or double switch is a challenging surgical procedure, which requires an experienced team of surgeons, paediatricians, anaesthesiologists and intensivists. Due to small numbers of children with congenital heart defects in Slovenia (approximately 120 per year); double switch was not yet performed at our centre. These children were offered either physiologic or palliative repair or were transferred to a high-volume centre. With this case, we offer a new therapeutic option for patients diagnosed with ccTGA in Slovenia. With encouraging mid-term results, we are aiming to achieve similar long-term results comparable to high-volume centres, where the survival rate is 80% 15 years after the double switch procedure (10,11).

## 4 Conclusion

To conclude, we have presented a case of a 3 years old female child with ccTGA and several associated defects (VSD, coronary artery anomaly, congenital complete AV block and dysplastic TV with significant regurgitation), which required a multidisciplinary approach and two surgical interventions before a complete correction of the anomaly. PA banding was performed and a PM implanted in the neonatal period. Limitation in physical activity, a VSD and TV regurgitation were indications for surgery. After accurate non-invasive imaging diagnostics only, a standardized double switch operation was successfully performed. Anatomic repair in ccTGA is the preferred surgical method; however, it requires an experienced surgical team with good support from other modalities (paediatric cardiology, anaesthesiology, intensive care unit, radiology). This is the first successful case of a double switch surgical repair for complex ccTGA with several associated heart anomalies in Slovenia with excellent mid-term outcomes.

## Conflict of interest

None declared.

## Informed consent of the patient

The parent of a case patient signed the informed consent form for the publication.

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