

# Unusual association of transposition of great arteries with infradiaphragmatic pulmonary venous return

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## ABSTRACT

A 21-day-old baby with transposition of the great arteries with intact ventricular septum, infradiaphragmatic totally anomalous pulmonary venous connection, and atrial septum defect underwent combined arterial switch operation, totally anomalous venous connection repair, and atrial septum defect closure, using a right-sided approach and temporary pulmonary veins occlusion, with no postoperative and 6-months follow-up complications. Complete anatomical correction is the most conceivable treatment for this unusual pathology; right-sided approach instead lifting the heart toward the right pleural cavity to perform left atrium-to-pulmonary veins anastomosis limits heart displacement and avoids nonphysiological three-dimensional alterations; moreover, ligation and division of vertical vein allow to obtain more tissue for anastomosis; temporary occlusion of pulmonary veins while performing anastomosis is a simple procedure that allows to avoid deep hypothermic circulatory arrest or low flow systemic perfusion. Combination of these details facilitates intra- and postoperative management, especially in combined demanding cases.

**Keywords:** Arterial switch operation, congenital heart disease, great vessel anomaly, neonate

## INTRODUCTION

Transposition of the great arteries (TGA) and totally anomalous pulmonary venous connection (TAPVC) is an unusual combination, and few cases are reported in the literature.<sup>[1-7]</sup> Combined anatomical correction is the preferable treatment, but in most cases, physiological correction was reported in the past series.<sup>[2-4]</sup> In addition, TGA associated with infradiaphragmatic TAPVC was even more rare,<sup>[5,6]</sup> and surgical technique details, especially regarding optimal exposure, limitation of anatomical distortions, and cardiopulmonary bypass or myocardial protection strategies, could play a role to reduce the incidence of postoperative complications.

## CLINICAL SUMMARY

A 21-day-old female baby (weight 2800 g) was brought to our cardiac surgery center with the diagnosis of

TGA with intact ventricular septum associated with infradiaphragmatic TAPVC [Figure 1] and atrial septum defect (8 mm).

The baby underwent surgical repair in hypothermic cardiopulmonary bypass instituted through aortic and bicaval cannulation. The proximal portion of the pulmonary venous collector was isolated; interatrial groove and superior and inferior venae cavae were mobilized. The vertical vein was ligated inferiorly close to the diaphragm and divided [Figure 2a]. After aortic cross-clamp and cardioplegic arrest with the del Nido cardioplegic solution, the right and left atria were opened, and pulmonary veins were temporary occluded with removable neurosurgical clips usually used for intracranial aneurysms occlusion. Pulmonary venous collector was opened such as the

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proximal portion of the vertical vein to obtain more tissue [Figure 2b]. Left atrium-to-pulmonary veins' anastomosis was performed with two PDS II 6/0 (Ethicon®) hemi-continuous running sutures, without displacing the heart, from the right side of the operative field [Figure 3a and b], retracting the right atrium to the left side. Then, arterial switch operation (ASO) and atrial septum defect closure with pericardial patch with precalibrated interatrial communication (3.5 mm) were performed. The aorta was anterior with type A coronary arteries according to the Yacoub classification. Trap-door technique was used to perform coronary arteries-to-neo-aorta anastomosis. Aortic cross-clamp and CPB time were 177 and 322 min, respectively; del Nido cardioplegic solution was repeated after 60 min. Modified ultrafiltration was applied. A peritoneal drain was inserted intraoperatively and was removed in postoperative day (POD) 6, and nitric oxide was used until POD 5. The sternum was left open, and delayed chest closure was performed in POD 3. The baby was weaned from mechanical ventilation in POD 7. Postoperative course was uneventful, and the patient was discharged in POD 32.

At 6-month follow-up, echocardiography showed no pathological findings.

## DISCUSSION

TGA combined with TAPVC is an unusual association. Few cases with combined TAPVC and TGA were reported in the past series, in which an atrial switch procedure was prevalently performed;<sup>[2-4]</sup> in the series reported by Lopes *et al.*,<sup>[5]</sup> in only one baby, an anatomical repair was performed and another one was also reported in a 6-day-old baby with TGA and TAPVC draining into the coronary sinus.<sup>[7]</sup>

Mykychak *et al.* reported a case of a 5-h-old baby with TGA with intact ventricular septum and infradiaphragmatic TAPVC treated with ASO and left atrium-to-venous collector anastomosis performed displacing the heart in the right pleural cavity but complicated by pulmonary vein obstruction (PVO); PVO was treated after 60 days through a reoperation with intraoperative direct balloon catheter dilatation.<sup>[6]</sup> Pulmonary vein obstruction is a possible complication that could influence the outcome. Hypoplastic or stenotic pulmonary veins and absence of a common confluence are considered risk factors, while “sutureless” technique has been demonstrated to be an independent protective factor.<sup>[8,9]</sup>

In our case, an anatomical combined correction has been performed without postoperative and mid-term complications, using a right-sided approach to limit heart displacement and two hemi-continuous running sutures to avoid purse-string effect. Temporary occlusion of pulmonary veins was applied to obtain a bloodless

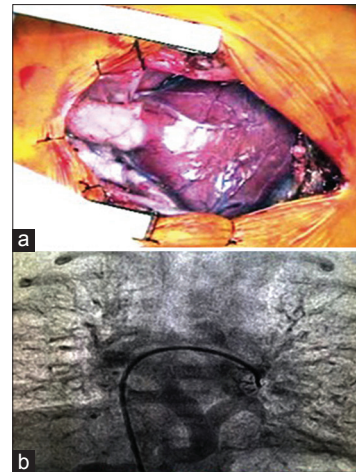


Figure 1: Transposition of the great arteries confirmed after pericardium opening (a), preoperative angiogram evidencing the total anomalous pulmonary vein connection (b)

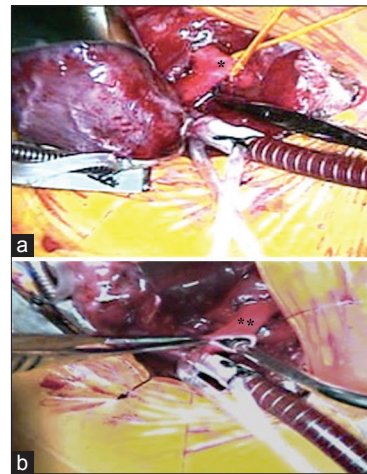


Figure 2: Vertical vein of the venous collector (\*) encircled (a) and proximal portion of the venous collector (\*\*) opened (b)

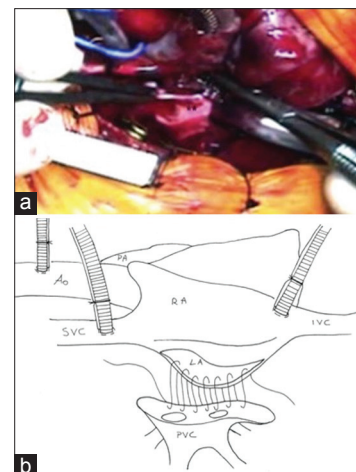


Figure 3: Left atrium-to-pulmonary vein anastomosis (\*\*) performed without displacing the heart, from the right side of the operative field (a and b). Ao: Aorta, IVC: Inferior vena cava, LA: Left atrium, PA: Pulmonary artery, PVC: Pulmonary venous collector, RA: Right atrium, SVC: Superior vena cava

operative field during left atrium-to-pulmonary vein anastomosis and to avoid deep hypothermic circulatory arrest (DHCA) or low-flow systemic perfusion.

Del Nido cardioplegic solution was administered three times to obtain a better myocardial protection in this demanding combined procedure.

Left atrium-to-pulmonary vein anastomosis could be performed according to the surgeon's preference equally from a right-sided approach as in our case, from a left-sided approach lifting the apex of the heart to the right to expose venous pulmonary collector and left atrium, or also through a transatrial approach by incising interatrial septum. The first approach limits the heart displacement and reduces the anatomical disarrangement, while anatomical distortion or exposure problems could arise with other approaches.

Chen and Xu reported enhanced surgical exposure and adequate patent anastomosis rate with a right-sided approach in 11 patients with infradiaphragmatic TAPVC, but they did not divide the vertical vein;<sup>[10]</sup> division and ligation of the vertical vein allow better mobilization of the venous collector and also provide enough tissue to perform anastomosis, but vertical vein could not be used as a pop-off; therefore, we created a 3.5 mm defect on the interatrial patch.

Right-sided approach for left atrium-to-pulmonary vein anastomosis [Figure 3b] associated with temporary pulmonary veins occlusion is a simple and safe technique that enhances exposure and reduces three-dimensional geometric alterations also in infradiaphragmatic TAPVC and allows us to avoid DHCA and its consequences, especially in combined longer operations.

#### Authors' note

The patient's parents consented to publication of the case report.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients' parents have given their consent for the patient images and other clinical information to be reported in the journal. The patients' parents understand that the patient name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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#### Conflicts of interest

There are no conflicts of interest.

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