# Tetralogy of Fallot Associated with a Rare Type Partial Anomalous Pulmonary Venous Return

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Tetralogy of Fallot (TOF) is rarely associated with partial anomalous pulmonary venous return (PAPVR). Unidentified PAPVR, however, might increase the risk of pulmonary valve replacement in repaired TOF patients by right ventricular (RV) dilatation and RV dysfunction. Here, we present a case of a 19-year-old male who received a correction of TOF 18 years ago and a rare type of PAPVR was identified during the follow up period. The anomalous pulmonary veins were connected to the left hepatic vein, left superior vena cava, and the right superior vena cava. Performing a pulmonary valve replacement, PAPVR was also corrected by an intra-atrial baffle with a new approach using the venous plexus between the left hepatic vein and the right hepatic vein.

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**Abstract**

Tetralogy of Fallot (TOF) is rarely associated with partial anomalous pulmonary venous return (PAPVR). Unidentified PAPVR, however, might increase the risk of pulmonary valve replacement in repaired TOF patients by right ventricular (RV) dilatation and RV dysfunction. Here, we present a case of a 19-year-old male who received a correction of TOF 18 years ago and a rare type of PAPVR was identified during the follow up period. The anomalous pulmonary veins were connected to the left hepatic vein, left superior

vena cava, and the right superior vena cava. Performing a pulmonary valve replacement, PAPVR was also corrected by an intra-atrial baffle with a new approach using the venous plexus between the left hepatic vein and the right hepatic vein.

**Keywords**

Tetralogy of Fallot, Partial anomalous pulmonary venous return, Intra-atrial baffle

**Introduction**

A rare but clinically important association is that of anomalous pulmonary venous connection with tetralogy of Fallot (TOF). In repaired TOF patients, right ventricle (RV) dilatation and dysfunction are common sequelae which pulmonary valve regurgitation (PR), residual ventricular septal defects, and pulmonary artery stenosis are well known contributing factors. The presence of partial anomalous pulmonary venous return (PAPVR), however, is a rare risk factor. Herein, we report a surgical case of unique pattern of PAPVR in a 19-year-old male who had previously received TOF correction. Appropriate consent forms were obtained from the patient.

**Case report**

At the age of 11 months, the patient was diagnosed as TOF and received a TOF repair with a transannular patch for right ventricular outflow tract reconstruction and discharged without major complications. Routine cardiac echocardiogram was reviewed once a year, however, moderate PR and RV dilatation developed 10 years after the operation which was earlier than expected. Cardiac magnetic resonance image (MRI) studies and cardiac computed tomography (CT), which were decided for further evaluation, showed a rare type of PAPVR (Fig. 1). The right upper-middle pulmonary veins (RUMPVs) were connected to the right superior vena cava (SVC) and the left upper pulmonary vein had a connection with the left SVC which drained to the left atrium (Fig. 2A, 2B). There was a connecting vein between the bilateral SVC (Fig 1). The left lower pulmonary vein (LLPV) was draining to the left hepatic vein (LHV) (Fig. 2C), and LHV had communications between the middle hepatic vein (MHV) as a venous plexus (Fig. 2D). Kinking at the origin of the left pulmonary artery (LPA) was also revealed. Deciding to repair the anomalous pulmonary drainage when the pulmonary valve replacement (PVR) is necessary, cardiac MRI studies were reviewed every three years. The last MRI finding, which was performed 1 year before admission, showed increasing RV end-diastolic volume index (RV EDVI, 174ml/m2) and RV end-systolic volume index (RV ESVI, 71ml/m2) with 38 percent of PR fraction. The pulmonary to systemic flow ratio (Qp/Qs) was 2.86 in cardiac catheterization studies, and planned an elective operation.

The patient received an elective open heart surgery through a redo median sternotomy. Cardiopulmonary bypass (CPB) was supported by cannulating the ascending aorta, right SVC, and the right femoral vein. After cross clamping the aorta, an incision was made from the lateral wall of right SVC to right atrium, and followed by a partial atrial septectomy. RUMPVs were rerouted by baffling to the atrial septal defect using Gore-tex vein graft (GTVG, Gore-Tex vascular graft, W.L. Gore assoc. Inc, Elkton, MD). The diaphragmatic surface of the pericardium was opened to expose the LLPV and the LHV. Internal closure between the LLPV and the LHV was performed using GTVG to separate the hepatic flow with the LLPV. Rerouting the LLPV was performed by baffling from the opening of the LHV to the baffle for the RUMPVs, within the lumen of the inferior vena cava (IVC) (Fig. 3). The incision of right SVC and right atrium was closed with using an additional GTVG. Aortic cross clamp was released and root vent was started. PVR was performed using 25mm St. Jude valve (St. Jude Medical, Minneapolis, MN) with LPA angioplasty. After an uneventful weaning from CPB support, the left SVC was ligated using a surgical clip to prevent the systemic venous blood mixing with the LUPV drainage. Extubating after 12hr of ICU care, the patient moved to the general ward and discharged after 8 days without significant complication. Postoperative CT showed decreased RV EDVI (58 ml/m2), RV ESVI (21 ml/m2) and demonstrated unobstructed venous return from the lungs (Fig. 4).

**Discussion**

In recent studies, planning a PVR for repaired TOF patient has focused at the timing, considering irreversible dysfunction of the RV and repeated intervention.[1] These studies aimed at several factors including RV volume status, exercise function, arrhythmia, and tricuspid regurgitation etc., however, standard indication is still not established.[1,2] In addition, a range of surgical suggestions with a broad spectrum of patient status including other cardiac lesions which causes significant hemodynamics are making surgeons decision more complicated. Our case was an extreme rare case that a repaired TOF patient was developing PR accelerated by a mixed type PAPVR.

TOF with anomalous pulmonary venous connection is extremely rare (0.6% of TOF patients) and there are only few reports in the English medical literature.[3] This case, moreover, is unique according to a mixed type PAPVR pattern which RUMPVs, LUPV, and LLPV were connected to right SVC, left SVC, and LHV. Redington et al. reported four (0.3%) of PAPVR patients among 1183 TOF patients which only corrected the TOF lesion without rerouting the anomalous pulmonary vein (one or two lobar vessels) during the 17 years of follow up period.[3] In this case, however, rerouting was performed because the four out of five lobar pulmonary veins were connected to systemic veins and significant volume overload was identified. In LLPV rerouting, baffling and internal closure technique was used instead of direct implantation to the left atrium since the LLPV had an intra-pulmonary course toward the subdiaphragmatic area and the venous plexus between the LHV and MHV had a sufficient flow. Internal closure technique was used to separate the hepatic venous flow and a baffle was placed within the lumen of IVC which is a similar fashion with correcting in 'Scimitar syndrome' with a long baffle.[6] Hepatic collaterals had developed well assuming the pressure gradient between the LHV and MHV, which the precise mechanism is still unclear.[7,8]

Innominate vein is a well-known site for anomalous connection of left pulmonary veins, however, there have been rare reports which left pulmonary veins connected to IVC.[4,5] To our knowledge, this might be the first reported instance of a unique pattern of PAPVR which anomalous pulmonary drains were connected to the right SVC, left SVC, and LHV.

## Conclusion

Tetralogy of Fallot is rarely associated with anomalous pulmonary venous return. Careful assessment of the pulmonary venous connection is required in all patients with TOF since PAPVR can accelerate RV dilatation, RV dysfunction, and PR. As rerouting the anomalous pulmonary drainage to the hepatic vein, using the venous plexus between the hepatic veins should be considered.

## Conflict of interest

The authors declare that there is no conflict of interest

## Author contributions

All authors participated in the research; J.A.S performed the literature search, and wrote the manuscript. N.R.K. performed the literature search, designed the figures. W.Y.L. revised the manuscript.

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**Figure legends**

**Figure 1** . A schema showing a rare type of partial anomalous pulmonary venous return. LHV, left hepatic vein; LLPV, left lower pulmonary vein; LSVC, left superior vena cava; LUPV, left upper pulmonary vein; RMPV, right middle pulmonary vein; RUPV, right upper pulmonary vein.

**Figure 2** . A, right upper pulmonary vein (RUPV) and right middle pulmonary vein (RMPV) drain to the right superior vena cava. B, left upper pulmonary vein (LUPV) drains to the left superior vena cava (LSVC). C, left lower pulmonary vein (LLPV) drains to the left hepatic vein (LHV) and to the inferior vena cava (IVC). D, venous plexus (circle) between the LHV and the middle hepatic vein (MHV).

**Figure 3** . Operative view showing rerouting right upper-middle pulmonary veins and left lower pulmonary vein by intra-atrial baffling. LLPV, left lower pulmonary vein; RUMPV, right upper-middle pulmonary vein.

**Figure 4** . Post-operative computed tomography shows unobstructed pulmonary venous drainage to the left atrium. A, RUMPVs and LLPV are drained to left atrium via the atrial septal defect (asterix). B, LUPV is drained to the left atrium. LLPV, left lower pulmonary vein; LUPV, left upper pulmonary vein; RLPV, right lower pulmonary vein; RUMPVs, right upper-middle pulmonary veins.