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Surgical Repair of Partial Anomalous Pulmonary Venous Connection In Adulthood: A 4-D Flow MRI Postoperative Evaluation

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Glossary Of Abbreviation

4-D Flow CMR: Four-Dimensional Flow Cardiac Magnetic Resonance
PAPVC: Partial Anomalous Pulmonary Venous Connection
IAS: Intact Atrial Septum
ASD: Atrial Septal Defect
MVP: Mitral Valve Prolapse
NYHA: New York Heart Association
TTE: Trans Thoracic Echocardiography
PAPs: Pulmonary Arterial Pressure Systolic
Thoracic – CT: Thoracic – Computed Tomography
SVC: Superior Vena Cava
PVRi: Pulmonary Vascular Resistance Index
RA: Right Atrium
LA: Left Atrium
PVC: Pulmonary Venous Collector
I-AB: Intra- Atrial Baffle
LPV: Left Pulmonary Vein
FPS: Flow Peak Speed
CHD: Congenital Heart Defect
Central Message

Four-Dimensional Flow CMR is a useful, non-invasive tool for hemodynamic study of complex intracardiac repair in congenital heart disease.

Central Picture:

4-D flow CMR vectors projection shows flow from pulmonary venous collector to left atrium.

INTRODUCTION

Partial anomalous pulmonary venous connection (PAPVC) is an uncommon congenital anomaly encountered in 0.4% to 0.7% autopsies. The anomaly is usually associated with atrial septal defect (ASD) of sinus venous type, rarely with an intact atrial septum (IAS), or in association with other cardiac diseases. We present a case of surgical correction of an adult patient affected by PAPVC with IAS, and severe mitral valve prolapse (MVP). Postoperative Four-Dimensional Flow Cardiac Magnetic Resonance (4-D Flow CMR) was performed to assess the status of PAPVC correction.

CASE REPORT

A 66-year-old male was referred to our hospital for worsening dyspnea in NYHA class III due to severe MVP. A transthoracic echocardiography (TTE) confirmed the diagnosis, showing an enlarged right atrium (30 cm²), a right ventricle with preserved function (TAPSE 17), and an estimated PAPs of 50 mmHg. The Scimitar sign was found at chest X-ray. Thoracic-CT showed a right-sided heart, a hypoplastic right lung, and a PAPVC with a venous collector of the diameter of 19 mm draining almost all the right lung to the right atrium at the distance of 1 cm from the superior vena cava (SVC) outlet, with an IAS. Cardiac catheterization demonstrates pulmonary artery precapillary and postcapillary hypertension, PAP of 60/33 mean 43 mmHg, a pulmonary vascular resistance index (PVRi) of 5 WU, and a Qp/Qs of 3.4. The patient was referred
for surgery. The procedure was performed through median sternotomy, and the diagnosis of right PAPVC, with the venous collector draining into the right atrium (RA), was confirmed. The MVP was repaired via a transeptal incision, then the surgical ASD was corrected with a heterologous pericardial patch to direct the flow from the pulmonary venous collector to the left atrium (LA) [figure 1 C, F]. The postoperative course was uneventful, and the patient was discharged on the sixth postoperative day. At the five-year follow-up, TTE demonstrated a mild regression in the dimension of the right atrium (23 cm²), right ventricle preserved systolic function (TAPSE 22), and significantly reduced PAP of 30 mmHg. A 4-D Flow CMR sequence was performed in 3 Tesla scanner (Ingenia, Philips Healthcare) in an axial orientation with the following parameters: velocity encoding of 130 cm/s, reconstructed spatial resolution 1.5 x 1.5 x 1.5 mm, flip angle 7°. 4D flow CMR data were processed using Arterys Cardio AIMR (Arterys Inc., San Francisco, CA). The good surgical result was confirmed with a patent pulmonary venous baffle [figure 2 A, C] and no residual shunt (Qp/Qs=1). Flow parameters were evaluated with 4-D Flow CMR in the pulmonary venous collector (PVC), in the intra-atrial baffle (I-AB), and in the left pulmonary vein (LPV) [Supplemental Table 1]. Flow Peak Speed (FPS) was slightly higher at the level of the I-AB than in proximal PVC and LPV, and an organized flow profile was visualized with vectors and streamlines projection [figure 2 B, E, C, F, VIDEO 1].

DISCUSSION

PAPVC correction with single-patch technique allows the deviation of the blood flow from the PVC to the LA. The surgical correction can lead to flow hemodynamic changes due to the altered anatomy, hence modification in blood rheology and circulation through the baffle, and 4-D flow CMR allows a non-invasive flow quantitative and qualitative evaluation [3,4,5]. We report a case of a venous return of the right pulmonary vein in the right atrium near the SVC. Five years after surgical repair, 4-D flow CMR allows the anatomical and blood flow evaluation. In our patient, we confirmed the surgical result with no residual shunt (Qp/Qs=1) and baffle obstruction. In our study,
we made a quantitative evaluation at the site of surgical deviation of the flow, resulting in a mild increment of FPS, while at the qualitative analysis with streamline projection of the flow from the PVC through the baffle in the RA to the LA, there is no evidence of formation of whirling motion [figure 2 C, F]. Furthermore, at vector projection, the deviated venous flow appears with an organized flow profile in the trajectory from the PVC to the left ventricle with mild acceleration [figure 2 B, E]. 4-D Flow CMR is emerging as a very promising non-invasive technology in pre and post-surgical repair of CHD, as PAPVC with ASD of sinus venous type, and with IAS, or in association with other cardiac diseases. However, further studies are needed to validate the application of measurement to assess advanced hemodynamic parameters and detect overall clinical outcomes in congenital heart disease. (IRB number approval: 12/2023 of 31 July 2023), patient written informed consent for publication of study data was obtained.

**Figure Legend:**

**Figure 1:** Preoperative and intraoperative imaging; A: Arrows show scimitar sign in Chest X-Ray; B-E: arrows indicate PVC in Thoracic-CT; D: arrow shows PVC in 3D-CT rendering; C: Arrowheads show iatrogenic ASD in surgical view; F: arrow shows PVC outlet partially sutured with pericardial patch.

**Figure 2:** 4-D Flow CMR streamline and vector projection; A, D: CMR coronal and transversal plane, white arrows show PVC in the right atrium; B, C: Coronal plane with blood flow trajectory from the PVC through the patent baffle to the LA; E, F: Transversal plane with flow trajectory from the PVC to the LA. Arrowhead: Inferior vena cava flow in the right atrium; Dashed circle: PVC flow to the right atrium; Solid circle: intra-atrial baffle flow deviation.

**Video Legend:**
Video 1: 4-D Flow CMR vectors and streamlines projection: Vectors (right) and streamlines (left) projection shows the deviated venous blood flow from the pulmonary venous collector to the left atrium through the intra-atrial baffle.

References:


**Supplemental Table 1: 4–D Flow CMR haemodynamic flow parameters:**

<table>
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<tr>
<th></th>
<th>PVC</th>
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<td>47,1</td>
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**Supplemental Table 1:** Flow (F), Flow Peak Speed (FPS), in the pulmonary venous collector (PVC), in the intra-atrial baffle (I-AB), and in the left pulmonary vein (LPV).