Surgical repair of partial anomalous pulmonary venous connection in adulthood: A 4-dimensional flow magnetic resonance imaging postoperative evaluation

Martina Rizzo,[a] Lamia Ait-Ali, MD, PhD,[b] Duccio Federici, MD,[c] Pierluigi Festa, MD,[d] Valeria Piagneri, MD,[e] Federico Landra, MD,[f] Matteo Cameli, MD, PhD,[f] and Gianfranco Montesi, MD,[g] Siena and Massa, Italy

From the [a]Cardiac Surgery Unit, Department of Thoracic and Cardiovascular Disease, Santa Maria alle Scotte Hospital, University of Siena, Siena, Italy; [b]Institute of Clinical Physiology, CNR, Monasterio Foundation, Massa, Italy; [c]Paediatric Cardiac Surgery and GUCH Unit, Heart Hospital, Monasterio Foundation, Massa, Italy; [d]Paediatric Cardiology and GUCH Unit, Heart Hospital, Monasterio Foundation, Massa, Italy; [e]Diagnostic Imaging Unit, Heart Hospital, Monasterio Foundation, Massa, Italy; [f]Division of Cardiology, Department of Medical Biotechnologies, University of Siena, Siena, Italy; and [g]Cardiac Surgery Unit, Department of Thoracic and Cardiovascular Disease, Santa Maria alle Scotte Hospital, Siena, Italy.

Institutional Review Board Number and Approval: 12/2023 of July 31, 2023. Patient written informed consent for publication of study data was obtained.

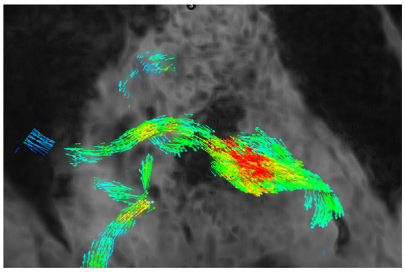
Received for publication Aug 2, 2023; revisions received Aug 23, 2023; accepted for publication Sept 16, 2023.

Address for reprints: Martina Rizzo, Cardiac Surgery Unit, Department of Thoracic and Cardiovascular Disease, Santa Maria alle Scotte Hospital, Viale Mario Bracci, 16, Siena, Italy 0577-585311 (E-mail: rizzo.mr@yahoo.com).

JTCVS Techniques 2023;■:1-4

2666-2507

https://doi.org/10.1016/j.xjtc.2023.09.017



4-D flow CMR vectors projection shows flow from the PVC to the LA.

CENTRAL MESSAGE

4-D flow CMR is a useful, noninvasive tool for hemodynamic study of complex intracardiac repair in congenital heart disease.

See Commentary on page XXX.

▶ Video clip is available online.

Partial anomalous pulmonary venous connection (PAPVC) is an uncommon congenital anomaly encountered in 0.4% to 0.7% of autopsies.[1] The anomaly usually is associated with atrial septal defect (ASD) of a sinus venous type, rarely with an intact atrial septum (IAS), or with other cardiac diseases.[1,2] We present a case of surgical correction of an adult patient affected by PAPVC with IAS and severe mitral valve prolapse (MVP). Postoperative 4-dimensional (4-D) flow cardiac magnetic resonance (CMR) was performed to assess the status of PAPVC correction.

CASE REPORT

A 66-year-old man was referred to our hospital for worsening dyspnea in New York Heart Association class III due to severe MVP. A transthoracic echocardiography confirmed the diagnosis, showing an enlarged right atrium (RA) (30 cm^2), a right ventricle with preserved function (tricuspid annular plane systolic excursion 17), and an estimated systolic pulmonary artery pressure of 50 mm Hg. The Scimitar sign was found on chest x-ray (Figure 1, *A*). Thoracic computed tomography 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 RA at the distance of 1 cm from the superior vena cava outlet, with an IAS (Figure 1, *B*, *D* and *E*). Cardiac catheterization demonstrates pulmonary artery precapillary and postcapillary hypertension, pulmonary artery pressure of 60/33, mean 43 mm Hg, pulmonary vascular resistance index of 5 Wood units, and Qp/Qs of 3, 4. The patient was referred for surgery. The procedure was performed through a median sternotomy, and the diagnosis of right PAPVC, with the venous collector draining into the RA, was confirmed. The MVP was repaired via a transeptal incision, and the surgical ASD was corrected with a heterologous pericardial patch to direct the flow from the pulmonary venous collector (PVC) to the left atrium (LA) (Figure 1, *C* and *F*). The postoperative course was uneventful, and the patient was discharged on postoperative day 6. At the 5-year follow-up, transthoracic echocardiography demonstrated a mild regression in the dimension of the RA (23 cm^2),

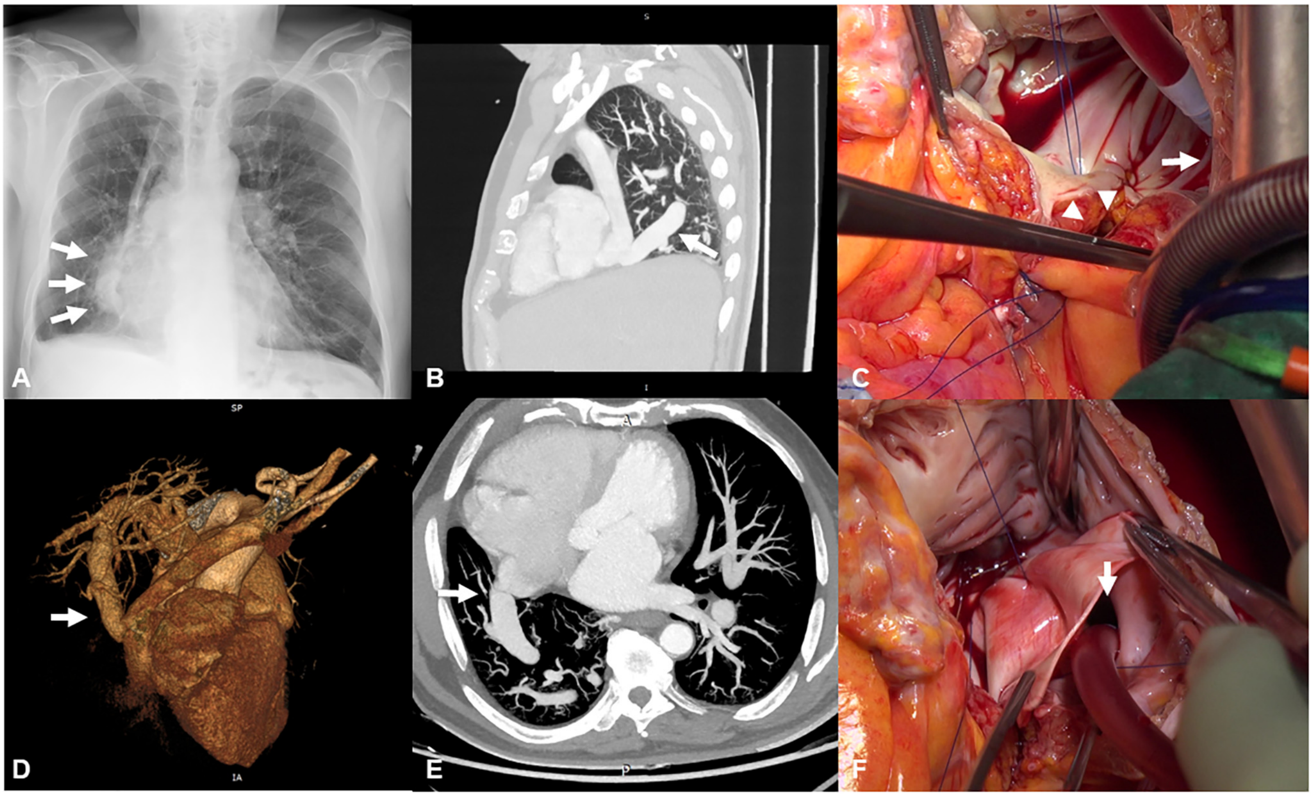


FIGURE 1. Preoperative and intraoperative imaging. A, *Arrows* show scimitar sign in chest x-ray. B-E, *Arrows* indicate PVC in thoracic computed tomography. D, *Arrow* shows PVC in 3-dimensional computed tomography rendering. C, *Arrowheads* show iatrogenic ASD in surgical view. F, *Arrow* shows PVC outlet partially sutured with pericardial patch.

right ventricle preserved systolic function (tricuspid annular plane systolic excursion 22), and significantly reduced pulmonary artery pressure of 30 mm Hg. A 4-D flow CMR sequence was performed in a 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 × 1.5 × 1.5 mm, and flip angle 7°. 4-D flow CMR data were processed using Arterys Cardio AIMR (Arterys Inc). The good surgical result was confirmed with a patent pulmonary venous baffle (Figure 2, *A* and *C*) and no residual shunt (Qp/Qs = 1). Flow parameters were evaluated with 4-D flow CMR in the PVC, intra-atrial baffle, and left pulmonary vein (Table E1). Flow peak speed was slightly higher at the level of the intra-atrial baffle than in the proximal PVC and left pulmonary vein, and an organized flow profile was visualized with vectors and streamlines projection (Figure 2, *B*, *C*, *E* and *F*; Video 1).

DISCUSSION

PAPVC correction with the 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; thus, modification in blood rheology and circulation through the baffle and 4-D flow CMR allow a noninvasive flow quantitative and qualitative evaluation.[3-5] We report a case of a venous return of the right pulmonary vein in the RA near the superior vena cava. Five years after surgical repair, 4-D flow CMR allows the anatomic 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 flow peak speed, whereas in the qualitative analysis with streamline projection of the flow from the PVC through the baffle in the RA to the LA, there was no evidence of formation of whirling motion (Figure 2, *C* and *F*). Furthermore, at vector projection, the deviated venous flow appeared with an organized flow profile in the trajectory from the PVC to the left ventricle with mild acceleration (Figure 2, *B* and *E*).

CONCLUSIONS

4-D flow CMR is emerging as a promising, noninvasive technology in presurgical and postsurgical repair of congenital heart disease, as PAPVC with ASD of sinus venous

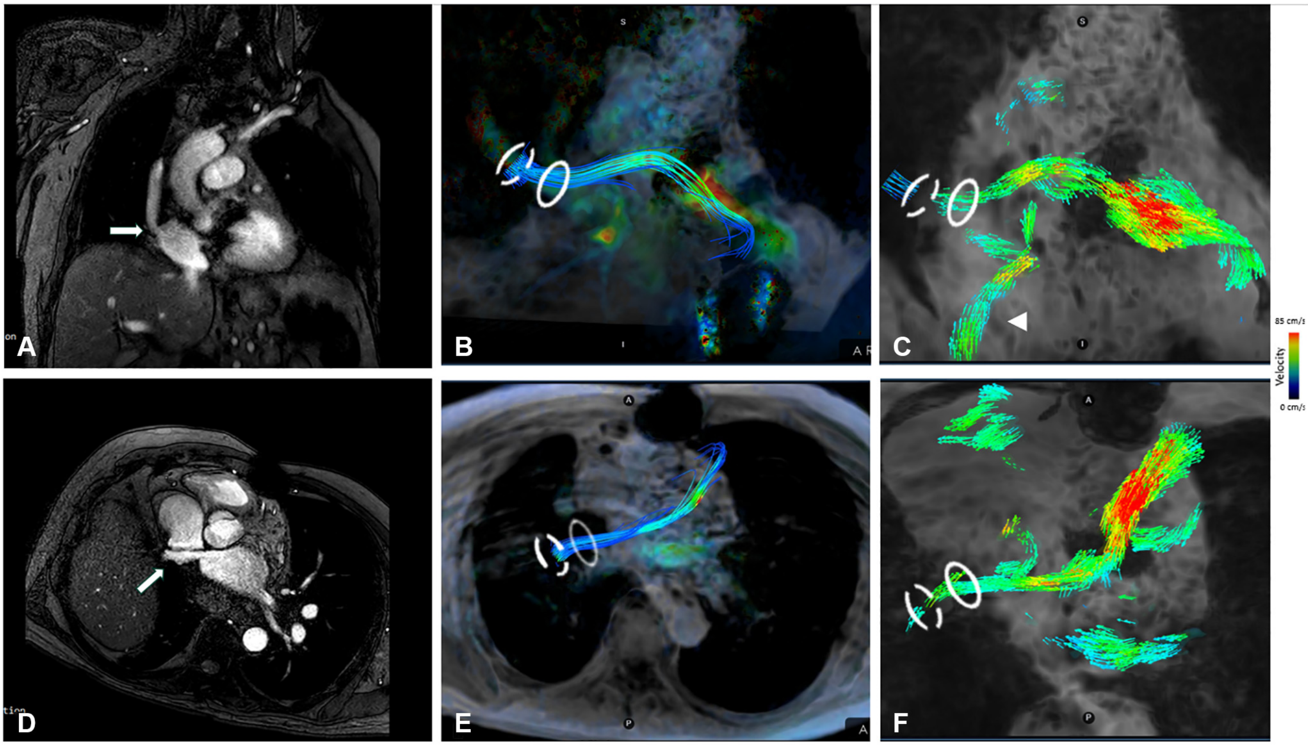
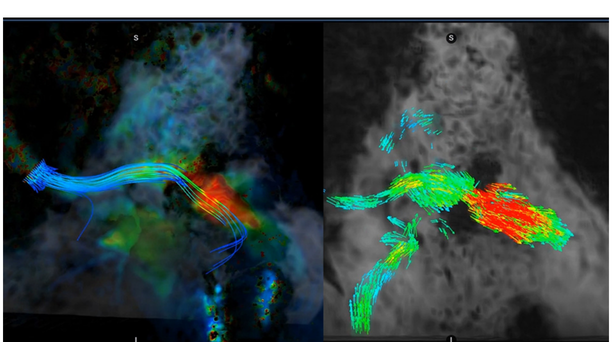


FIGURE 2. 4-D flow CMR streamline and vector projection. A and D, CMR coronal and transversal plane. *White arrows* show PVC in the RA. B and C, Coronal plane with blood flow trajectory from the PVC through the patent baffle to the LA. E and F, Transversal plane with flow trajectory from the PVC to the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

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.



VIDEO 1. 4-D flow CMR vectors and streamlines projection: Vectors (right) and streamlines (left) projection show the deviated venous blood flow from the PVC to the LA through the intra-atrial baffle. Video available at: https://www.jtcvs.org/article/S2666-2507(23)00363-2/fulltext.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

1. Graham G, Dearani JA, Mathew J, Miranda WR, King KS, Schaff HV, et al. Partial anomalous pulmonary venous connection with intact atrial septum: early and midterm outcomes. *Ann Thorac Surg*. 2023;115:1479-84. https://doi.org/10.1016/j.athoracsur.2022.04.031
2. Majdalany DS, Phillips SD, Dearani JA, Connolly HM, Warnes CA. Isolated partial anomalous pulmonary venous connections in adults: twenty-year experience. *Congenit Heart Dis*. 2010;5:537-45. https://doi.org/10.1111/j.1747-0803.2010.00458.x
3. Ebel S, Kühn A, Aggarwal A, Köhler B, Behrendt B, Gohmann R, et al. Quantitative normal values of helical flow, flow jets and wall shear stress of healthy volunteers in the ascending aorta. *Eur Radiol*. 2022;32:8597-607. https://doi.org/10.1007/s00330-022-08866-5
4. Shadden SC, Arzani A. Lagrangian postprocessing of computational hemodynamics. *Ann Biomed Eng*. 2015;43:41-58. https://doi.org/10.1007/s10439-014-1070-0
5. Dyverfeldt P, Bissell M, Barker AJ, Bolger AF, Carlhäll CJ, Ebbers T, et al. 4D flow cardiovascular magnetic resonance consensus statement. *J Cardiovasc Magn Reson*. 2015;17:72. https://doi.org/10.1186/s12968-015-0174-5

TABLE E1. Four-dimensional cardiac magnetic resonance hemodynamic flow parameters

Parameter	PVC	I-AB	LPV
F mL/sec	2500	2500	2800
FPS cm/sec	65.2	70.2	47.1

PVC, Pulmonary venous collector; *I-AB*, intra-atrial baffle; *LPV*, left pulmonary vein; *F*, flow; *FPS*, flow peak speed.