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CASE REPORT

BEGINNER

CLINICAL CASE SERIES

4-Dimensional Flow by Cardiac Magnetic Resonance Informs Surgical Planning in Partial Anomalous Pulmonary Venous Return

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ABSTRACT

Four-dimensional flow cardiac magnetic resonance enhances the visualization of blood flow in a 3-dimensional volume throughout the cardiac cycle, thus dramatically improving visualization of pulmonary venous anatomy by cardiac magnetic resonance. We demonstrate the impact of 4-dimensional flow on diagnosis and surgical planning for partial anomalous pulmonary venous return. **(Level of Difficulty: Beginner.)** (J Am Coll Cardiol Case Rep 2020;2:672–7)

CASE 1

A 7-year-old child was referred for surgical repair of a superior sinus venosus defect (SVD). Transthoracic echocardiogram (TTE) demonstrated right-sided heart enlargement, a left superior vena cava (LSVC) draining into the coronary sinus, and concern for partial anomalous pulmonary venous return (PAPVR).

LEARNING OBJECTIVES

- Diagnostic accuracy of PAPVR is important for surgical planning.
- 4-dimensional flow CMR offers improved diagnostic sensitivity with the addition of the temporal dimension.

SVDs are invariably associated with PAPVR (1). The classic surgical approach is placement of a patch-baffle that closes the defect and redirects anomalous pulmonary veins into the left atrium, with a risk for venous obstruction (2). Superior insertion of pulmonary veins extends the length of the baffle with increased risk for obstruction. The Warden procedure is an alternative in which the proximal right superior vena cava (RSVC) is divided and anastomosed to the right atrial appendage, thereby leaving the lower RSVC as a conduit for anomalous veins to be directed into the left atrium (3,4).

Although there are no strict guidelines, it has become our institutional practice to perform cardiac

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magnetic resonance (CMR) on all patients with SVD, given the high incidence of PAPVR and the limitations of TTE. Steady-state free precession (SFFP) and 2-dimensional (2D) phase contrast imaging is performed to assess ventricular volumes and shunt fractions. Contrast-enhanced magnetic resonance angiography (CE-MRA) is used to capture a 3-dimensional (3D) volume and define the vascular anatomy; however, angiography is limited by the ability to opacify all structures of interest with a single contrast medium administration.

This patient underwent CMR on a 1.5-T Siemens Aera device (Siemens Healthcare, Erlangen, Germany). Imaging demonstrated a severely enlarged right ventricular diastolic volume of 153 ml/m^2 and estimated a pulmonary-to-systemic flow ratio of 2.6:1. CE-MRA showed anomalous right upper pulmonary venous (RUPV) drainage to the RSVC; however, administration of contrast material timed to opacify the pulmonary veins limited visualization of the relevant systemic venous anatomy (**Figures 1A and 1B**).

The 4-dimensional (4D) flow imaging was performed following contrast administration by using Siemens WIP 785A with the following: field of view (FOV), 280 $\times$ 196 mm^2; matrix, 160 $\times$ 72; slice thickness, 2 mm; echo time (TE), 2.3 ms; repetition time (TR), 38.7 ms; velocity encoding (VENC), 200 cm/s; flip angle, 15°; and parallel acquisition technique (PAT), 2. Total acquisition time was slightly <9 min. Analysis was completed with Arterys version 19.06.2 (Arterys Inc., San Francisco, California). Visualization of the entire vasculature with dynamic flow imaging clearly demonstrated 2 RUPVs draining into the RSVC; 1 vein inserted posteriorly, just cranial to the azygous vein, whereas a second vein inserted on the right aspect of the RSVC at the level of the right pulmonary artery (**Figures 1C and 1D**). The right middle pulmonary vein (RMPV) was also seen draining to the right atrium at the RSVC-right atrial junction (Video 1). Additionally, 4D flow allowed visualization of the LSVC draining into the coronary sinus with an absent bridging vein. In this case, the temporal dimension of 4D flow allowed for improved visualization of the venous anatomy by separating systolic from diastolic flow patterns, which is quite challenging with standard CE-MRA. Alternative time-resolved 3D MRA sequences can provide temporal information, albeit at the cost of spatial resolution, and these images are limited by contrast distribution.

Direct surgical inspection through a midline sternotomy confirmed the pulmonary venous anatomy, as seen with 4D flow. The patient underwent a successful Warden procedure and was discharged home 3 days following surgery. Post-operative TTE demonstrated unobstructed caval and pulmonary venous drainage.

CASE 2

A full-term newborn with the prenatal diagnosis of D-transposition of the great arteries (D-TGA) presented with cyanosis and underwent balloon atrial septostomy. TTE confirmed D-TGA and a patent ductus arteriosus and raised concern for PAPVR of the left upper pulmonary vein (LUPV).

PAPVR is a reported but rare association with D-TGA (5). The LUPV can be redirected during the arterial switch operation, although with prolonged bypass time. CMR was planned to confirm the venous anatomy and estimate the flow burden through the LUPV. CE-MRA demonstrated anomalous drainage of the LUPV into the innominate vein (**Figure 2A**). A 2D phase contrast image estimated the flow through the anomalous vein; however, this was limited by the presence of multiple shunt lesions complicating the acquisition and analysis. Multiple 2D phase contrast planes had to be adjusted and acquisition repeated to optimize accuracy, given the limited spatial resolution.

4D flow imaging performed after contrast administration used the following: FOV, 250 $\times$ 175 mm^2; matrix, 160 $\times$ 72; slice thickness, 1.5 mm; TE, 2.3 ms; TR, 38.7 ms; VENC, 250 cm/s; flip angle, 15°; and PAT, 2. Total acquisition time was 6.5 min. Post-processing with flow assessment found that the LUPV received 20% of pulmonary venous return. These calculations matched estimates provided by 2D phase contrast imaging; however, the single 4D acquisition was accomplished in one-half the time of repeated 2D phase contrast imaging. Additionally, the 4D flow calculations could be internally validated by the ability to confirm both

ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

D-TGA = D-transposition of the great arteries

FOV = field of view

LUPV = left upper pulmonary vein

MRA = magnetic resonance angiography

PAPVR = partial anomalous pulmonary venous return

PAT = parallel acquisition technique

RMPV = right middle pulmonary vein

RSVC = right superior vena cava

RUPV = right upper pulmonary vein

SVD = sinus venosus defect

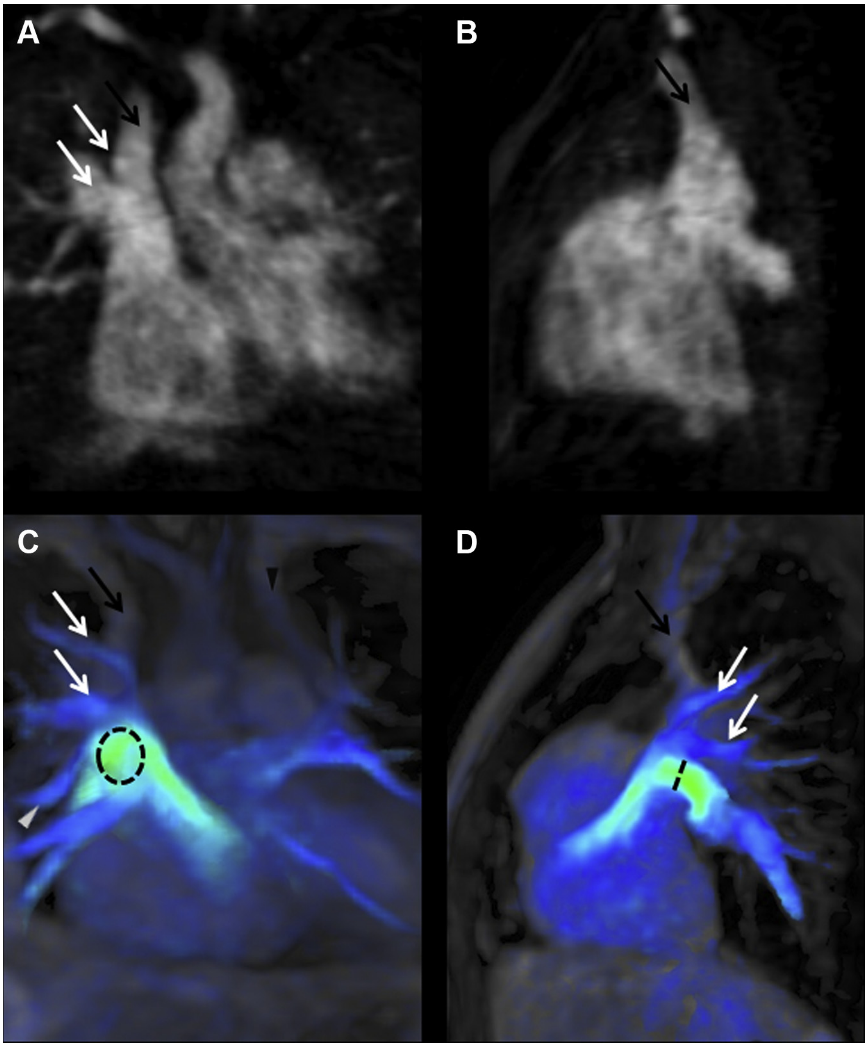
TE = echo time

TR = repetition time

TTE = transthoracic echocardiogram

VENC = velocity encoding

FIGURE 1 Case 1: Right PAPVR and Sinus Venous Defect



Contrast-enhanced magnetic resonance angiography in the **(A)** coronal and **(B)** sagittal projections suggestive of anomalous, right upper pulmonary veins **(white arrows)** draining to the right superior vena cava **(black arrow)**. The 4-dimensional flow images in the **(C)** coronal and **(D)** sagittal projections demonstrate the right superior vena cava **(black arrows)**, left superior vena cava **(black arrowhead)**, anomalous, right upper pulmonary veins **(white arrows)**, right middle pulmonary vein **(white arrowhead)**, and superior sinus venosus defect **(black dotted lines)**. PAPVR = partial anomalous pulmonary venous return.

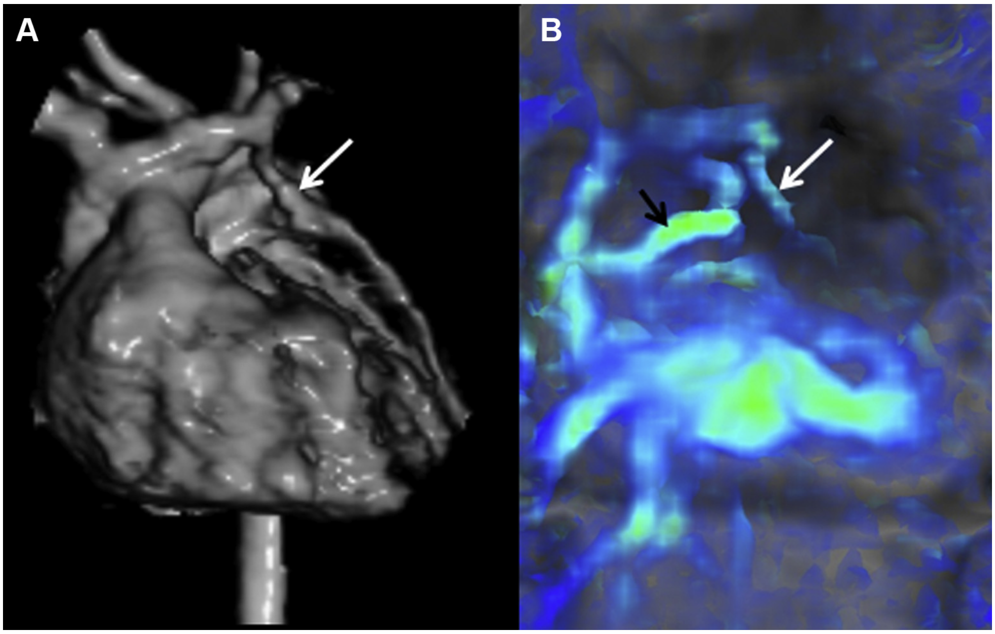
systemic and pulmonary inflows and outflows from the same data set. Dynamic flow imaging further depicted the anatomic relationship of the LUPV relative to the anatomy of unrepaired D-TGA (Figure 2B).

Given the small contribution of the LUPV, the patient underwent a successful arterial switch operation without addressing PAPVR. The patient recovered and was discharged home on postoperative day 8. His serial cardiology follow-up will include monitoring for evidence of right-sided heart dilation.

CASE 3

A 9-year-old child with a complex chromosomal abnormality, developmental delay, and PAPVR of the left pulmonary veins was referred for CMR to confirm the pulmonary venous anatomy and evaluate right heart size.

FIGURE 2 **Case 2: Left PAPVR in Unrepaired D-TGA**



(A) Volume rendering of a contrast-enhanced magnetic resonance angiography demonstrates partial anomalous pulmonary venous return (PAPVR) of the left upper pulmonary vein **(white arrow)** to the innominate vein. **(B)** The 4-dimensional flow images confirm the left upper pulmonary vein anatomy **(white arrow)**, as well as shunt lesions including a patent ductus arteriosus **(black arrow)**.

Cine imaging demonstrated a right ventricular diastolic volume of 205 ml/m^2. The pulmonary-to-systemic flow ratio was 2.8. CE-MRA demonstrated a large, left-sided pulmonary venous confluence draining vertically to the innominate vein, as well as anomalous RUPV drainage to the RSVC (**Figures 3A and 3B**). The 4D flow imaging was performed with the following: FOV, 351 $\times$ 262 mm^2; matrix, 256 $\times$ 144; slice thickness, 2 mm; TE, 2.6 ms; TR, 45.4 ms; VENC, 150 cm/s; flip angle, 15°; and PAT, 3. Further review demonstrated an anomalous RMPV crossing the midline that had not been appreciated on the CE-MRA (Video 2). Dynamic flow imaging helped confirm the RMPV joining the left venous confluence (**Figures 3C and 3D**).

This anatomy posed an interesting surgical challenge. Typically, in left-sided PAPVR the vertical vein is transected and anastomosed to the left atrial appendage (6). However, the anomalous right-sided veins require further surgical redirection with use of a baffle.

Intraoperative inspection confirmed the foregoing anatomy, and the patient underwent a modified Warden procedure, including division of the left-sided vertical vein and anastomosis to the right atrial appendage because of concerns about kinking of the vertical vein if anastomosed to the left atrial appendage. The cardiac portion of the divided RSVC was baffled along with the drainage from the left vertical vein through the atrial septum into the left atrium, and the cranial portion of the RSVC was anastomosed to the roof of the right atrium. Post-operative TTE demonstrated unobstructed venous drainage.

DISCUSSION

PAPVR comprises a spectrum of unique anatomy and is often concurrent with congenital heart disease. A complete understanding of pre-operative anatomy and physiology is paramount to successful outcomes. Use of 4D flow CMR has shown tremendous promise in providing reliable, noninvasive shunt fractions, which are typically challenging with 2D phase contrast (7-10).

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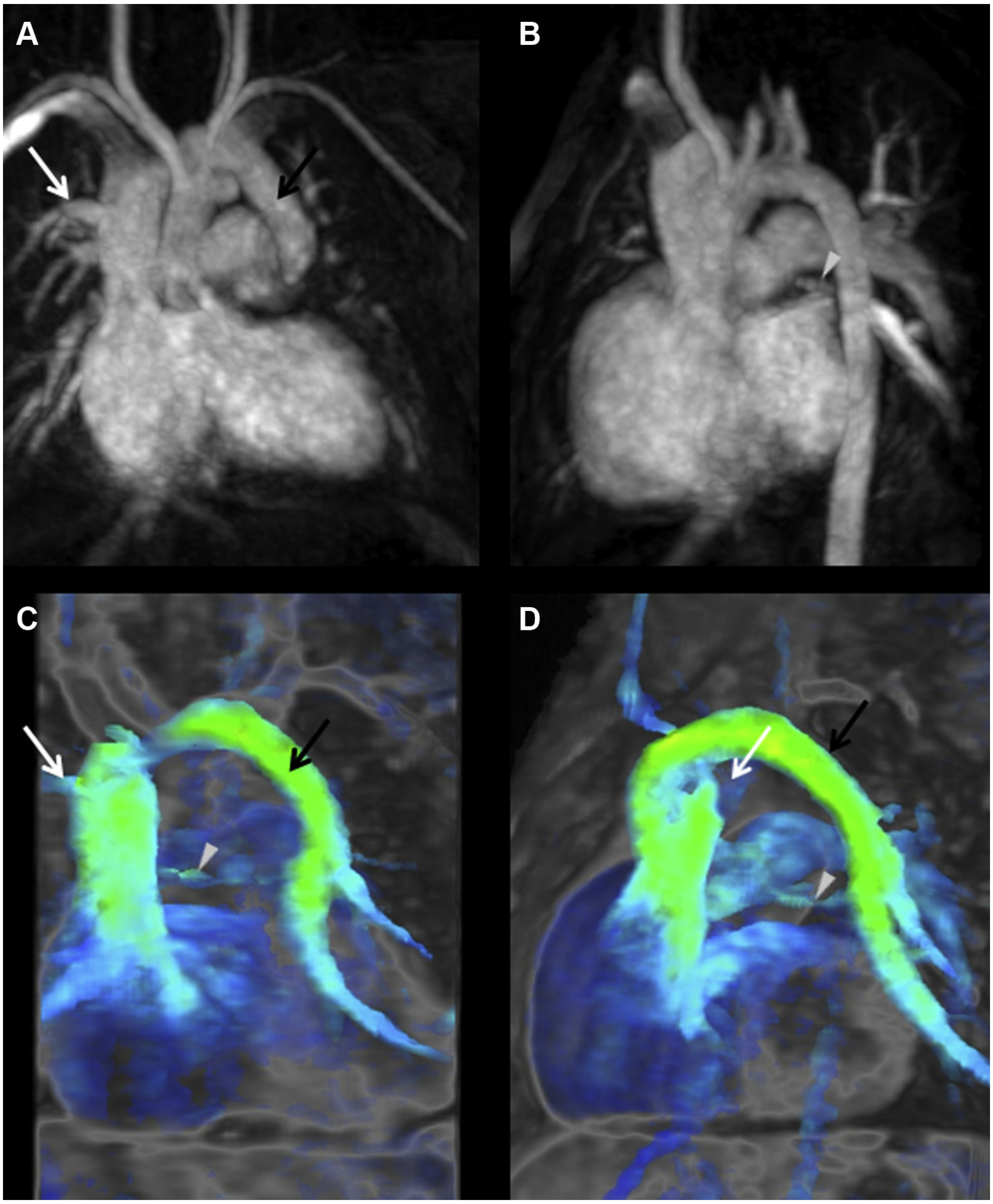
FIGURE 3 **Case 3: Complex Right and Left PAPVR**



Contrast-enhanced magnetic resonance angiography in the **(A)** coronal and **(B)** sagittal projections and 4-dimensional flow images in the **(C)** coronal and **(D)** oblique sagittal projections demonstrate the anomalous left pulmonary venous drainage **(black arrow)**, the anomalous right upper pulmonary vein **(white arrow)**, and the anomalous right middle pulmonary vein draining to the left-sided pulmonary venous confluence **(white arrowhead)**. 4D flow images allow improved visualization of subtle venous structures by focusing on diastolic flow patterns. PAPVR = partial anomalous pulmonary venous return.

CONCLUSIONS

This case series demonstrates the routine use of 4D flow as an important diagnostic tool that enhances diagnostic sensitivity and has the potential to improve surgical planning and patient outcomes in PAPVR.

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KEY WORDS cardiac magnetic resonance, congenital heart defect, imaging, pediatric surgery

APPENDIX For supplemental videos, please see the online version of this paper.