# Cardiovascular Magnetic Resonance in Unsuspected Interrupted Inferior Vena Cava

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Interrupted inferior vena cava (IVC) is a rare anomaly. Anomalies of IVC are clinically important for cardiologists and radiologists who plan to intervene in the right heart. We describe three cases of IVC interruption diagnosed by cardiac magnetic resonance imaging study.

## Introduction

Anomalies of inferior vena cava (IVC) have been known to anatomists since 1793[1], but IVC interruption with azygous and hemiazygous substitution is a rare anomaly which has been documented recently[2].

Usually patients are asymptomatic; however, this finding can be associated with cardiac and visceral anomalies[1] such as the heterotaxy syndrome (HS) with asplenia or polysplenia. Situs ambiguous, or heterotaxy, refers to visceral malposition and dimorphism associated with indeterminate atrial arrangement[3]. It differs from situs inversus because the latter represents a mirror image of the normal anatomy, i.e: the systemic atrium is on the left with a left-sided trilobed lung. Also, the liver, gallbladder, and inferior vena cava are present on the left side. The pulmonary atrium is on the right side with a right-sided bilobed lung. The stomach, spleen, and aorta are also present on the right side. In the heterotaxy syndrome, the arrangement of body organs is not orderly as seen in situs solitus or situs inversus. Interrupted IVC is the most consistent finding in heterotaxy with polysplenia[4]. These anomalies of IVC are clinically important, particularly for cardiologists and radiologists who plan to intervene on the right heart or to assess hemodynamics using IVC as the venous channel to enter the right side of the heart.

In this study, we describe three cases of IVC interruption with or without HS incidentally diagnosed by cardiac magnetic resonance (CMR) imaging study (1.5T Excite GE, Milwaukee, WI) with clinical implications for patient management. Non-contrast breath hold spin echo and steady state free precession (SSFP) cine images were performed in all cases. Gadolinium was neither used nor required in any of the three cases. A composite of axial and coronal imaging was performed in all cases.

## Keywords

Vena Cava, Inferior/abnormalities; Cardiovascular Abnormalities; Magnetic Resonance Spetroscopy/diagnostic use.

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## Case Report

### Case 1

A 69-year-old Caucasian female underwent cardiac catheterization for the evaluation of chest pain and mild pulmonary hypertension. The left heart catheterization was uneventful and she did not have significant coronary artery disease. While performing the right heart catheterization through the right femoral vein access, the Swan Ganz catheter was seen to course along the left side of the spine as it ascended into the thoracic cavity. The operator was able to advance it without difficulty up to the level of the aortic arch. Beyond the arch, it could not be further advanced into the right atrium. To delineate the catheter course, iodinated contrast was hand injected into the illiac vein. The dye injection revealed a venous structure crossing over to the left of the spine approximately at the level of the second lumbar vertebra (L2). This vein continued to course on the left side up to the level of the aortic arch and then connected to the right heart (Figure 1). The anomalous venous connection was not fully characterized and the operator ordered a CMR to assess it better.

The CMR clearly revealed that the IVC was interrupted just beyond the renal veins. The hemiazygous vein originated from the left renal vein and received the terminal portion of the IVC. The hemiazygous vein then ascended into the left hemithorax and ultimately drained into a persistent left superior vena cava (PLSVC). The PLSVC in turn drained into a markedly dilated coronary sinus (Figure 2). The CMR also clarified the subdiaphragmatic visceral drainage on the right side in the absence of the usual course of the IVC. The liver was seen to directly drain into the right atrium via hepatic veins. The patient was found to have the usual position of the heart i.e. levocardia with normal chamber size and systolic function along with situs solitus. A left-side aortic arch with hyper arterial bronchi system was visualized. The liver was bridging most of the abdomen with polysplenia featuring HS. The patient otherwise had no associated congenital heart disease (CHD).

### Case 2

A 46-year-old female underwent CMR to evaluate a dilated coronary sinus seen on the transthoracic echocardiogram. A suspicion for CHD, including a PLSVC,

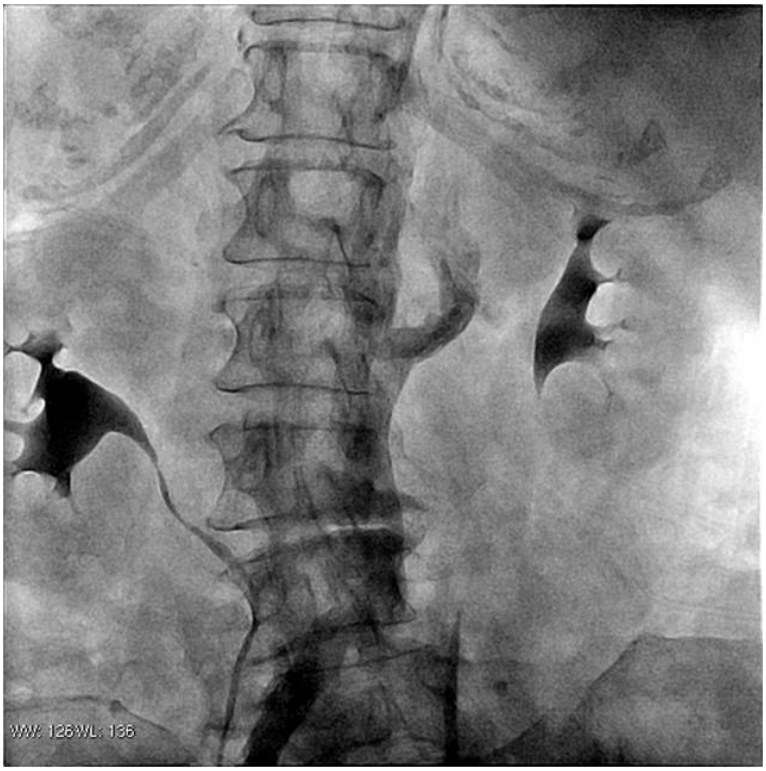


**Figure 1 -** *Abnormal abdominal venous drainage. The catheter was not able to progress upper the femoral vein. Dye was injected and revealed venous continuation arching up to aortic and then connecting to right side heart.*

was considered. The CMR demonstrated interruption of the IVC with azygous continuation which drained into the PLSVC that drained into the coronary sinus. The coronary sinus was markedly dilated measuring 30.1 X 23.5mm. The hepatic vein directly drained into the right atrium. There was no evidence of HS and all pulmonary veins drained normally into the morphologic left atrium without further evidence of CHD.

### Case 3

A 40-year-old female underwent CMR study for the evaluation of arrhythmogenic right ventricular dysplasia (ARVD). The CMR was negative for ARVD. Again, an incidental note was made about an interrupted IVC with an extremely dilated azygous continuation, measuring 20.3mm at the level of the aortic arch where it normally drained into the right SVC. All pulmonary veins drained into the left atrium. The situs was solitus, and although the patient had polysplenia, there was no evidence of any other congenital heart disease.

## Discussion

Interrupted IVC is a common finding in HS. Incidence of congenital heart disease, such as double outlet right ventricle, atrioventricular canal and transposition of great arteries in HS is high and is around 50 – 100%[4]. Nevertheless, it can be an occasional finding in patients without HS and cardiac anomalies. However, in our case series there was no patient with cardiac congenital disease and there was only one patient who had HS.

The patient with HS in our case series did not have any congenital cardiac anomalies.

The prevalence of interrupted IVC is low. Some authors[5,6] a prevalence of 0.001% in routine computer tomography. However, the true incidence may be considerably higher, as early estimations are largely based on patients who had clinical evidence of CHD. Despite the apparent low incidence, we describe three cases where interrupted IVC was diagnosed incidentally on cardiac MRI. In all cases of interrupted IVC, the coronary sinus was dilated, which typically prompted further evaluation via CMR. In this sense, CMR high spacial resolution allows to perform a detailed venous system depiction, where even a small system as coronary venous sinus is well depicted. It is important to highlight that interrupted IVC coexists with numerous congenital cardiac and vascular anomalies such as HS and PLSVC, indicating that one needs to maintain a high level of suspicion for their surveillance in patients with dilated coronary sinus. Prognostically, this information may not be useful. However, anomalous abdomen venous drainage can be a roadblock in such patients who require right heart

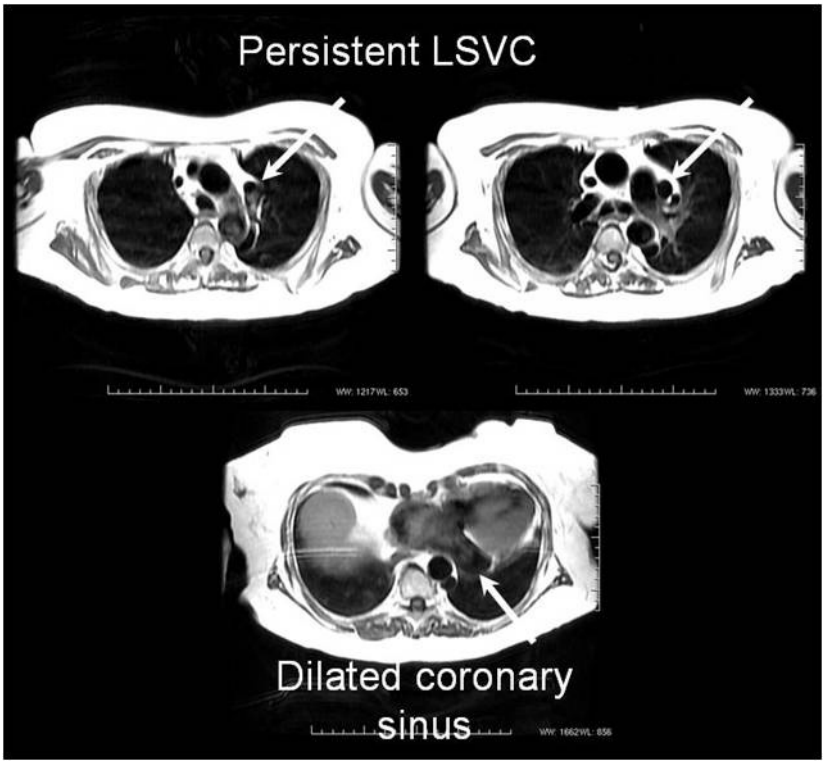


Figure 2 - *Spin echo images highlighting a PLSVC draining to a dilated coronary sinus.*

access and hemodynamics via femoral vein approach as described in our first case.

In conclusion, non-invasive CMR is an ideal radiation-free imaging technique with an inherent ability to define complex anatomy such as interrupted IVC or HS typically not diagnosed via other non-invasive or invasive strategies such as transthoracic or transesophageal echocardiography, cardiac catheterization or nuclear imaging. Performance of CMR is indispensable for cardiologists or radiologists faced with ambiguous venous anatomy in the course of routine imaging or intervention. Therefore, it can bring further information about venous system for cardiac surgeries and implantation of devices.

**Potential Conflict of Interest**

No potential conflict of interest relevant to this article was reported.

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**Study Association**

This study is not associated with any post-graduation program.

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