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CASE REPORT
Other

Abnormal wire's trajectory during edge-to-edge mitral valve repair—a rare case report of inferior vena cava anomaly

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Background Inferior vena cava (IVC) anomalies are rare and diagnosed incidentally as most patients are asymptomatic. We present a case where an abnormal course of the wire during percutaneous mitral valve repair revealed abnormal IVC anatomy leading to procedure termination. We summarized all IVC anomalies relevant to cardiovascular physicians and designed a simplified tool to illustrate their course for differential diagnosis.

Case summary A 78-year-old female presented with severe and symptomatic mitral regurgitation. The heart team decided to proceed with a percutaneous option, considering the patient's high surgical risk. While ascending from the femoral vein, the wire took an abnormal course to the left side of the vertebrae and continued beyond the cardiac silhouette downwards the right atrium (RA). We decided to abort the procedure due to the high risk for vascular complications assuming the need to cross it with the device's delivery system. Retrospective computed tomography analysis revealed an interrupted IVC at the level of the renal vasculature and azygos continuation toward the RA via a dilated superior vena cava. The patient was referred to surgery and had successful mitral and tricuspid valve repair and was discharged home in good health.

Discussion The increased number of minimally invasive percutaneous procedures, especially for valvular heart disease, mandates a profound understanding of the arterial, and venous system anatomy. Inferior vena cava anomalies represent a group of anomalies with different paths and variations and have a tremendous impact on all aspects of the procedure.

Keywords Inferior vena cava • Mitral valve repair • Anomaly • Femoral vein • Superior vena cava • Mitral regurgitation • Case report

ESC Curriculum 4.3 Mitral regurgitation • 2.1 Imaging modalities • 4.9 Multivalvular disease • 6.1 Symptoms and signs of heart failure

Learning points

- Inferior vena cava anomalies are rare and easily misdiagnosed as most patients are asymptomatic.
- Deep vein thrombosis at early age and signs of vena cava dilatation on chest imaging may be the only clues.
- Cardiovascular operators should be aware of different vascular anomalies, plan the procedure accordantly, and choose alternative access when applicable.

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Introduction

The inferior vena cava (IVC) develops primarily between the 6th and 8th gestational weeks in a sequential growth, regression, and anastomosis process.[1] The complexity of the embryogenesis process may lead to numerous anatomical variations of IVC formation. We present a rare case of azygos continuation of interrupted IVC identified during percutaneous mitral valve repair.

Timeline

Office visit (November 2018)	The patient complained of exertional dyspnoea. Transthoracic echocardiography showed biatrial enlargement, left ventricular dilatation, moderate to severe mitral regurgitation, with peak pulmonary artery pressure of 43 mmHg.
Progression of symptoms (October–December 2021)	Dyspnoea and fatigue become daily, with leg oedema appear more frequently. Transoesophageal echocardiography revealed severe mitral regurgitation, causing an increase in right-sided volume and pressure with mild to moderate tricuspid regurgitation.
Evaluation for transcatheter mitral valve replacement (TMVR) (February 2021)	TMVR computed tomography (CT) analysis indicated valve dimensions (an annulus area of 2140 mm^2 and a diameter of 54.3 mm x 47.1 mm) too large for TMVR. Abdominal CT analysis with arterial angiogram reported no major finding.
Percutaneous mitral valve repair (February 2021)	An attempt of mitral valve repair using the Abbott MitraClip system was aborted due to the abnormal trajectory of the inferior vena cava.
Surgical mitral valve repair (June 2021)	The patient underwent a successful combined mitral and tricuspid valves repair and surgical treatment for atrial fibrillation (Maze procedure) along with left atrial appendage ligation.
Follow-up (August 2021)	The patient reports significant improvement in daily life activity, denies any exertional dyspnoea, and engages in cardiac rehabilitation.

Case presentation

A 78-year-old female with hypertension, hyperlipidaemia, coronary artery disease, and atrial fibrillation, presented with shortness of breath over the last several months. She is unaware of any significant diseases in her family, and she had no major surgeries except for vaginal hysterectomy, which was uneventful.

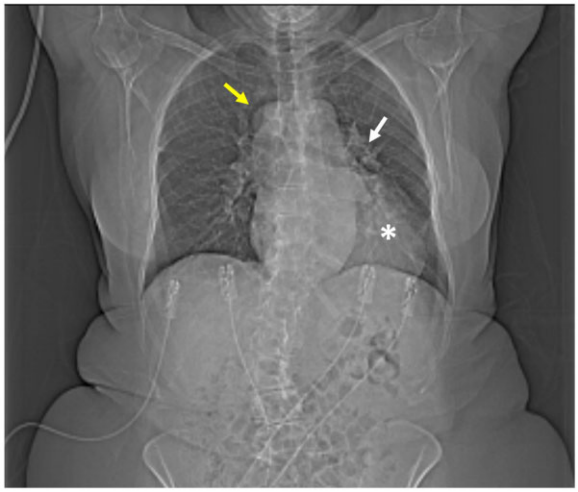
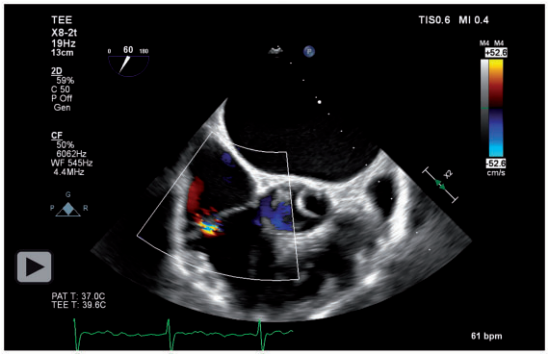


Figure 1 A plain chest X-ray on admission indicating the enlargement of the left ventricle silhouette (asterisk) and a perihilar haze (white arrow). Note the focal enlargement of the paratracheal strip indicating the silhouette of dilated azygos vein arch on the right superior mediastinum (in retrospective analysis, white arrow).



Video 1 Pre-procedure transoesophageal echocardiography in different views.

Physical examination on admission indicated signs of mild heart failure with a continuous holosystolic murmur at the cardiac apex and rales at the base of the lung. Chest X-ray shows mild pulmonary oedema and a significant dilated left heart silhouette (*Figure 1*). Transoesophageal echocardiography (TOE) indicated a left ventricle (LV) ejection fraction of 50%, severely dilated left atrium (LA) and right atrium (RA), severe mitral regurgitation with malcoaptation, and centrally directed flow jets, and mild to moderate tricuspid regurgitation (*Video 1*). The proximal isovelocity surface area derived effective regurgitant orifice area measured 0.43 cm^2 with LV end-systolic diameter of 42 mm.

A cardiac heart team discussed all treatment options, including valve replacement surgery, percutaneous valve repair (PMVR) using a

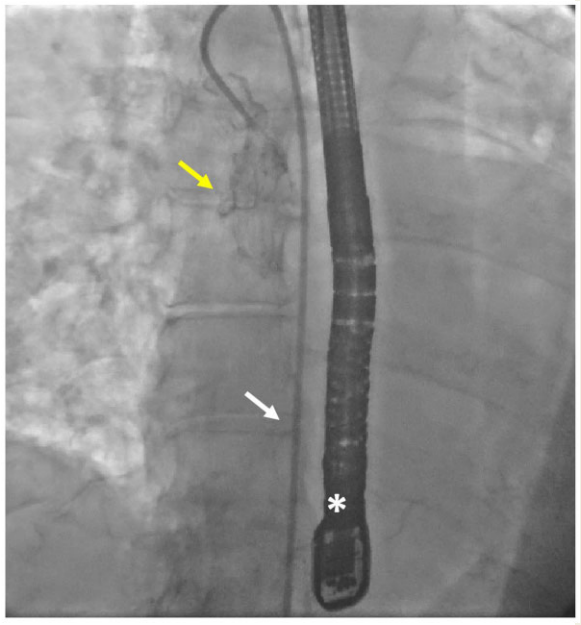


Figure 2 Fluoroscopy during edge-to-edge mitral repair with transoesophageal echocardiography guided (asterisk) indicating an abnormal position of the wire ascending from the femoral vein at the left side of the vertebrae (white arrow) and contrast flow jet from multipurpose A catheter-directed downward and to the left side indicating right-sided chamber (yellow arrow). TOE, transoesophageal echocardiography; MPA, multipurpose A.

clip, and transcatheter mitral valve replacement (TMVR) study trial. The patient underwent an abdominal and chest computed tomography (CT) angiogram to assess the feasibility of TMVR, which indicated large MV dimensions excluding her from the procedure. No other major abnormal findings were reported on CT except for cholelithiasis and suspected renal cysts.

Considering the patient's high mortality risk for surgery (3.96% by the Society of Thoracic Surgeons score), unfavourable anatomy for TMVR, and patient preference, we decided to proceed with a PMVR using an edge-to-edge technique.

Under ultrasound guidance, we introduced a 6 Fr sheath into the right femoral vein using a micropuncture catheter and confirmed its location by angiography.

While ascending, the wire took an abnormal path on the left side of the chest and beyond the cardiac silhouette and then downwards towards the RA (*Figure 2*). A multipurpose catheter showed RA trace and a flow-jet directed to the left side supporting the location right-sided chambers (*Video 2*). Transoesophageal echocardiography confirmed that the trajectory was from the superior vena cava (SVC) to the RA without passing through the IVC-RA junction (Supplementary material online, *Figure S1*). Given this, we decided to stop the procedure as it cannot safely be performed.

Retrospective analysis of the CT revealed an interruption of the IVC at the level of the hepatic segment. The IVC was drained directly into dilated azygos veins along with the hemiazygos and the accessory veins and entered the RA via the SVC. The massive enlargement of the LA led to significant compression of the azygos vein before draining to the SVC (*Figure 3* and corresponding *Figure 4D*, *Video 3*, Supplementary material online, *Video S1*).

Follow-up

The patient was referred to surgery, where the mitral and the tricuspid valves were repaired successfully and discharged home in good health. Two months following the surgery, the patient reported a significant improvement in daily life activity and denied exertional dyspnoea. She engaged in cardiac rehabilitation and felt much better than before the surgery.

Discussion

The volume of percutaneous cardiovascular procedures is increasing worldwide.[2] New techniques and devices are continually developing to treat valvular diseases, heart defects, pulmonary embolism, and arrhythmias and require using femoral veins as preferred access.[3,4]

Anomalies of the IVC are extremely rare and therefore not widely known. Considering that in most cases, patients are asymptomatic makes the diagnosis even more challenging and probably underestimated. Clinical clues for IVC anomaly could be one or more of the following: a known congenital heart anomaly, asplenia or polysplenia, deep vein thrombosis at an early age, varicose vein, and haematochezia.[5–14]

Definite diagnosis using CT venogram and US imaging could aid for proper procedural planning and prevent unnecessary complications. Once the IVC anomaly is confirmed, operators should also consider the minimum luminal diameter across the vascular path to fit with the device's dimension and the alignment of the delivery system when entering the RA (*Table 1*).[15] Choosing an alternative access route as the brachial, subclavian, axillary, jugular, and hepatic veins or use a retrograde approach via the arterial system may be reasonable.

We review anomalies with a clinical significance for cardiovascular interventionalists.

Double or duplicated inferior vena cava

Double or duplicated inferior vena cava (DIVC) is one of the most common variants in IVC anomalies seen in 0.2–3% of the population. The most common variant of DIVC is the separate drainage of the left common iliac artery to the left IVC, forming two parallel veins on both sides of the aorta. The left and right IVCs usually join to form a dilated azygos vein, which drains to the RA via the SVC (*Figure 4B*). The dilated veins can be easily mistaken with a cyst, tumour, and lymphadenopathy on abdominal CT or US and could be potentially dangerous when a biopsy is attempted.[7–9]

A persistent left-sided or displaced inferior vena cava

Left-sided or displaced inferior vena cava (LIVC) is very similar to the normal variant of IVC (*Figure 4A*) except that its origin is not from the right side. The iliac veins drain to the LIVC on the left side of the vertebrae and then cross it anteriorly at the level of the renal arteries to form the hepatic segment of the IVC while entering the RA (*Figure 4C*). In only a minority of cases, the LIVC merges with the azygos vein and enters the RA via the SVC.[10,11]

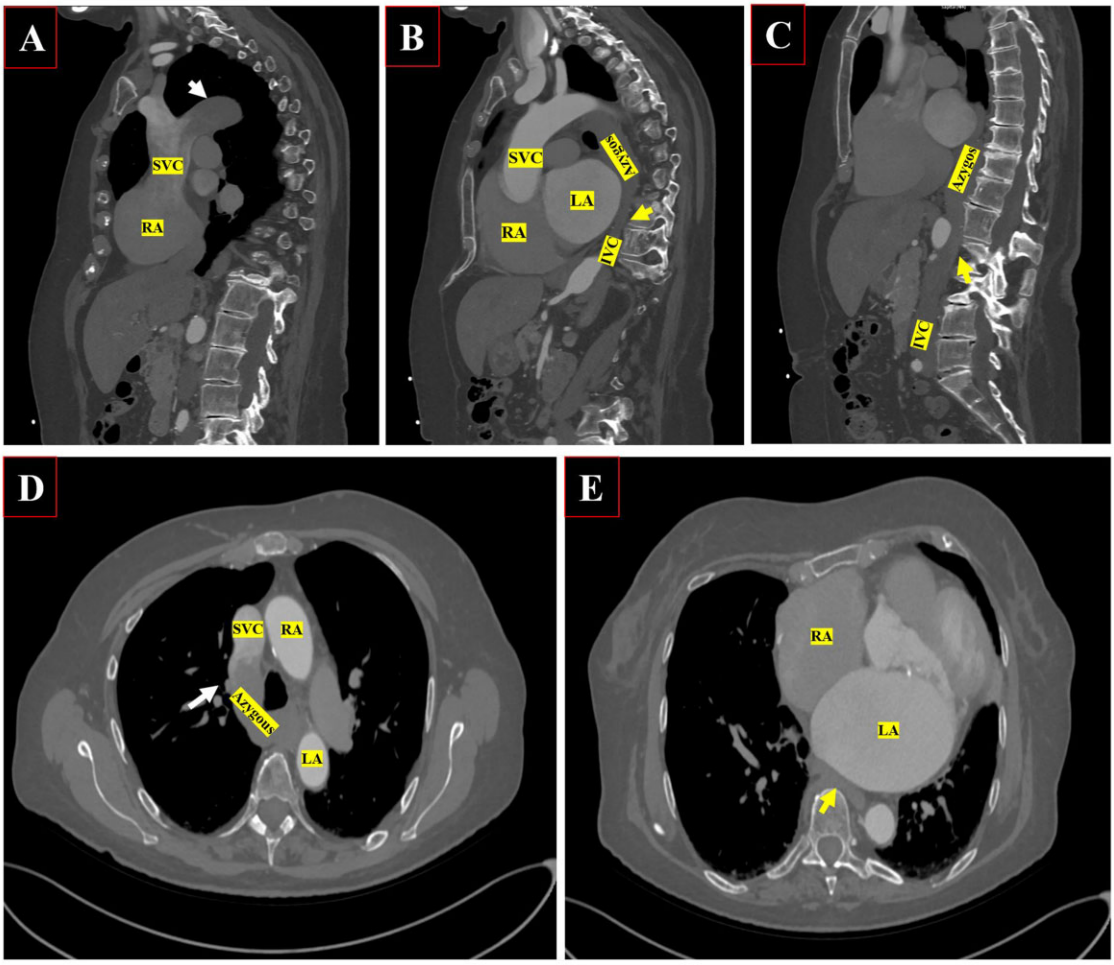
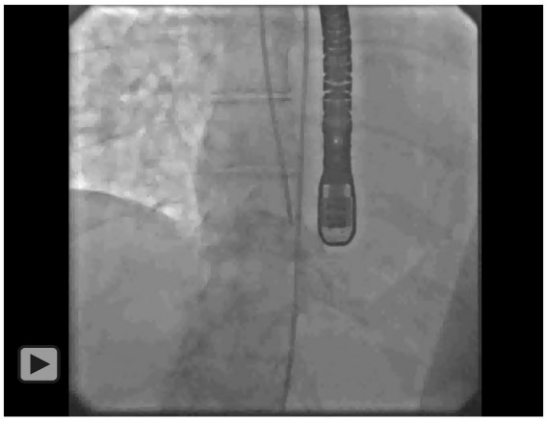
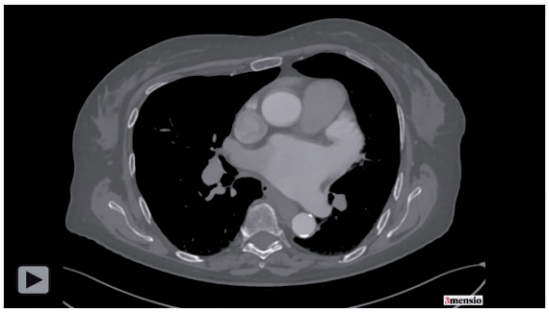


Figure 3 Sagittal and cross-sectional view contrast-computed tomography of the chest and the abdomen before the procedure. Note the enlargement of the azygos vein before draining to the superior vena cava (white arrow *A* and *D*) compressed by an enlarged left atrium (yellow arrow *B* and *E*) and the interruption of the inferior vena cava at the hepatic segment (yellow arrow *C*). CT, computed tomography; IVC, inferior vena cava; LA, left atrium; RA, right atrium; SVC, superior vena cava.



Video 2 Fluoroscopy view of contrast injection from a multipurpose catheter in right atrium.



Video 3 Sagittal view of the thorax using contrast computed tomography angiogram (arterial phase).

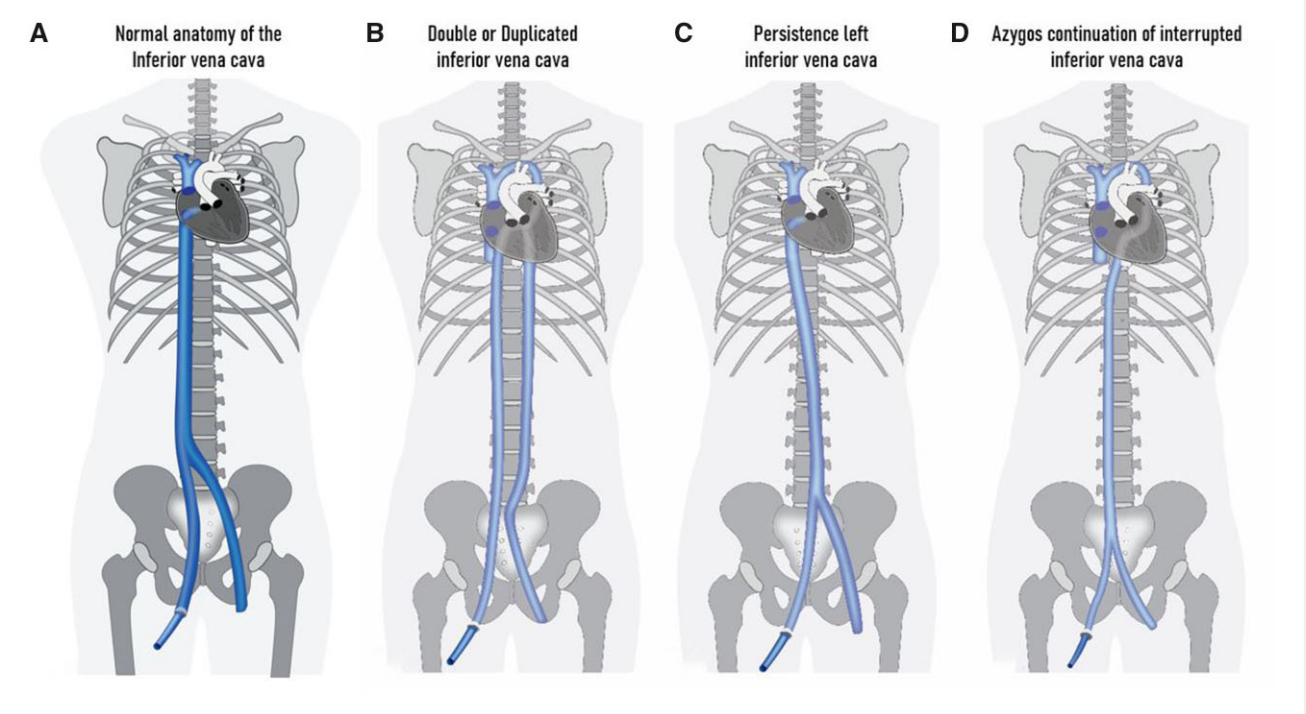


Figure 4 Illustration of the four most common inferior vena cava anomaly relevant to vascular interventionists.

Table 1 The most common inferior vena cava anomalies and related cardiovascular procedures are expected to be affected by the abnormal path, device over-dimension, and mal-alignment

Inferior vena cava anomalies	Cardiovascular procedures expected to be affected by IVC anomaly
• A double or duplicated inferior vena cava (DIVC) • Agenesis or the absence of infrarenal IVC (AIVC) • A persistent left-sided or displaced IVC (LIVC) • Azygos continuation of the inferior vena cava or the interruption of IVC (I-IVC) • Double IVC with retroaortic right renal vein and hemiazygos continuation of the IVC • Double IVC with retroaortic left renal vein and azygos continuation of the IVC	• IVC filter • Pulmonary artery thrombectomy/thrombolysis • Temporary pacemaker implantation • ASD/PFO closure • LAA closure • Edge-to-Edge mitral valve repair • Percutaneous transcatheter mitral valve replacement • Edge-to-edge tricuspid valve repair • Temporarily pacing during TAVR procedure • PDA closure • Electrophysiology study and catheter ablations • Right heart study and catheterization • VSD closure • Percutaneous balloon mitral valvuloplasty • Pulmonic valve repair and replacement

ASD, atrial septal defect; IVC, inferior vena cava; LAA, left atrial appendage; PDA, patent ductus arteriosus; PFO, patent foramen ovale; TAVR, transcutaneous aortic valve replacement; VSD, ventricular septal defect.

Agenesis or the absence of infrarenal inferior vena cava and the azygos continuation of interrupted inferior vena cava

Agenesis or the absence of infrarenal inferior vena cava (AIVC) and interrupted inferior vena cava (I-IVC) are the rarest variants of all IVC anomalies, and the exact embryogenesis is unclear. It is believed that it may be the result of embryological failure or perinatal thrombosis resulting in IVC mal-development and the absence of a normal IVC.[12] AIVC and I-IVC share common paths as the iliac veins drain to a dilated collateral lumbar venous system, which can easily be misdiagnosed as paraspinal masses. Then, it ascends to join the hemiazygos vein and enter the RA via the SVC (*Figure 4D*) while the hepatic vein directly connects to the RA. Consequently, the azygos vein becomes dilated and forms an arch to compensate for the high volume and pressure of the IVC.[13,14]

Conclusion

Our case study aims to raise awareness of the phenomena, highlight the importance of reviewing the venous system before any percutaneous procedure and provide a simplified tool for immediate diagnosis and alternative therapeutic options.

Lead author biography

Dr Koren Ofir is an interventional cardiologist in Emek medical center in Israel and a clinical lecturer at the Faculty of Medicine of Technion-institute of Technology. Since 2021, he participates in an interventional fellowship program at Cedars-Sinai (Los-Angeles, USA) under Dr Raj R. Makkar. His main research interest is in the field of valvular heart diseases.

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Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Author disclosures

Dr. Makkar received grant support from Edwards Lifesciences Corporation; he is a consultant for Abbott Vascular, Cordis, and Medtronic and holds equity in Entourage Medical. Other authors have no conflicts of interest to disclose.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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