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# Inferior vena cava return in left atrium: a rare presentation with severe mitral regurgitation

*Sol atriyumda inferior vena kava dönüşü: şiddetli mitral yetersizlik ile nadir bir tablo*

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***ABSTRACT***

Herein, we present a rare case of abnormal connection of the inferior vena cava to the left atrium with an interatrial communication associated with severe mitral regurgitation which altered clinical presentation. Successful correction was achieved after mitral valve repair and re-routing of the inferior cava flow to the right atrium.

***Keywords:*** Cardiac septal defect; mitral valve insufficiency; sinus venosus atrial septal defect; systemic venous return.

***ÖZ***

Bu yazıda, klinik tabloyu değiştiren şiddetli mitral yetersizlik ile ilişkili interatriyal iletişimli sol atriyumda inferior vena kavanın anormal bağlantısı olan nadir bir olgu sunuldu. Mitral kapak tamiri ve inferior cava akımının yeniden sağ atriyuma yönlendirilmesinin ardından başarılı bir düzeltme sağlandı.

***Anahtar sözcükler:*** Kardiyak septal defekt; mitral kapak yetersizliği; sinüs venozus atriyal septal defekt; sistemik venöz dönüş.

Inferior vena cava (IVC) draining into the left atrium is a rare anomaly of systemic venous return.[1-3] In the literature, several cases with various anatomical associations have been published.[1-6]

Herein, we report a female case with a Marfanoid morphotype, presenting with an abnormal return of the IVC into the left atrium associated with an interatrial communication and severe mitral regurgitation.

## CASE REPORT

A 16-year-old schoolgirl was referred for surgical treatment of severe mitral regurgitation associated with an interatrial communication. Physical examination revealed a good overall condition with dyspnea New York Heart Association (NYHA) Class II without cyanosis. Blood pressure was 107/65 mmHg, oxygen saturation 96%, weight 42 kg and height 166 cm. The patient suffered from several phenotypic features mimicking Marfan syndrome; however, she did not fulfill the Ghent's criteria for the definite diagnosis of Marfan syndrome. She presented with skeletal malformations including moderate pectus excavatum, arachnodactyly, joint hypermobility, an arm span to height ratio greater than 1.05; and a decrease in visual acuity. The cardiac examination showed a 3/6 systolic ejection murmur at the apex, split and fixed S2. There was no sign of the right heart failure and all findings were non-specific. Electrocardiography showed a regular sinus rhythm at 8/bpm and biatrial hypertrophy. Chest radiography revealed a cardiothoracic ratio of 0.5 with a prominent pulmonary artery segment and increased pulmonary vascular markings.

Echocardiography demonstrated a moderately dilated left ventricle with normal systolic functions and dilated right cavities; an aortic root diameter of 20 mm without regurgitation, Grade 3 mitral regurgitation by A2 and P2 prolapse and trivial tricuspid regurgitation. A low-situated interatrial communication was also diagnosed with significant left to right shunting (QP/QS>2). Computed tomography scan demonstrated a large IVC connecting directly to the floor of left atrium (Figure 1).

The patient was operated on by median sternotomy under cardiopulmonary bypass and aortic

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cross-clamping. The IVC was cannulated directly in its intrapericardial portion. After right atriotomy, an inferior sinus venosus defect was found and there was a complete rim of the septal tissue surrounding it with approximately 0.5 cm of atrial tissue at the inferior margin. The orifice of the IVC was well-visualized and was located within the left atrial cavity. The inferior right pulmonary vein was low-situated and connected to the left atrium, but unrelated to the IVC (Figure 2). The mitral valve was explored trough a transseptal approach after enlarging the defect trough the oval fossa and repaired using conventional techniques. The abnormal return was corrected by a Dacron patch sewn down on the floor of the left atrium and back on the top edge of the interatrial communication to direct the IVC flow into the right atrium. Postoperative course was uneventful. Repeated echocardiography demonstrated a competent mitral valve with no residual shunt. The patient was discharged in the 10th postoperative day under medical treatment with a diuretic and vasodilators.

## DISCUSSION

As rare entities, abnormalities of systemic venous return are isolated or associated with other cardiac malformations.[1-3] They are often diagnosed incidentally, leading to mild or no hemodynamic disturbance, when they drain to the right atrium; however, they beget a right to left shunt, when they drain to the left atrium and clinically cause cyanosis.[1] Abnormal return of the IVC to the left atrium is the less common of such anomalies.[4-6] It has been reported with an intact atrial septum[4] and in association with an interatrial communication; this can be an ostium secundum defect[5] or a sinus venosus defect.[6] In the latter case, total[7] or partial[8] anomalous drainage of

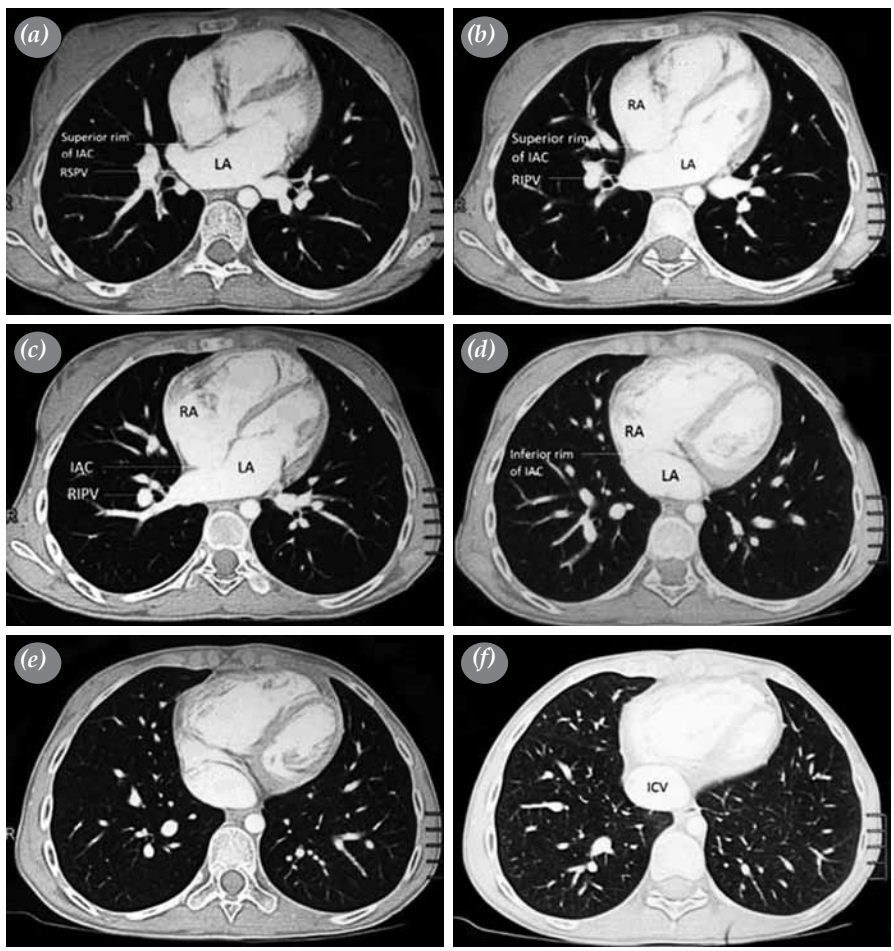


**Figure 1.** Serial axial computed tomography scans of cranial **(a)** to caudal **(f)** showing a large inferior vena cava connecting directly to the floor of the left atrium.

RA: Right atrium; IAC: Interatrial communication; RSPV: Right superior pulmonary vein; RIPV: Right inferior pulmonary vein; LA: Left atrium; IVC: Inferior vena cava.

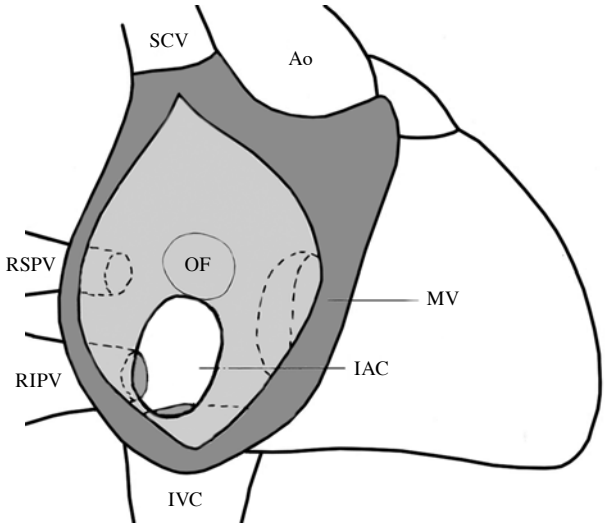


**Figure 2.** An illustration of the anatomy of the presented case.
SCV: Superior caval vein; Ao: aorta; RSPV: Right superior pulmonary vein; RIPV: Right inferior pulmonary vein; OF: Oval fossa; MV: Mitral valve; IAC: Interatrial communication; IVC: Inferior vena cava.

pulmonary veins to the IVC can be associated. Also, in this case, there is frequently overriding of the intact muscular border of the oval fossa by the mouth of the IVC.[9]

Another mechanism for the connection of the IVC to the left atrium is the presence of a large Eustachian valve, which can, then, deflect the vein flow through a defect in the oval fossa to the left atrium.[10] Occasionally, this can be mistaken for the lower margin of the defect and closed, diverting blood flow from the IVC to the left atrium.[11]

In our case the orifice of the IVC was totally situated in the left atrium and the right inferior pulmonary veins were within the left atrium and away from the orifice of the IVC. A similar case was reported by Kim et al.,[6] in which the authors described a low-lying secundum defect with the orifice of the IVC located in the left atrium and unrelated to the defect.

The association with a mitral regurgitation is uncommon and has not been reported in the literature to date. The etiology is probably genetic as in our case with several Ghent criteria[12] of Marfan syndrome (cardiovascular, musculo-skeletal, and ophthalmic), and mitral valve prolapse is one of the minor cardiovascular criteria.

Clinically the symptomatology is that of a secundum atrial septal defect with cyanosis and bidirectional shunting. It is the right-to-left due to the draining of the IVC in the left atrium and left-to-right, when left atrial pressure exceeds the right atrial pressure. However, in our case, the presence of severe mitral regurgitation suggested a more significant left-to-right shunt due to an increased left atrial pressure during systole. This also explains the absence of cyanosis and dilatation of the right cavities.

Although echocardiographic diagnosis is helpful, it does not always yield the definite diagnosis, as in our case. The diagnosis can be made more definitely with contrast echocardiography by injecting serum in the lower limb.[13] In addition, three-dimensional echocardiography has been also used successfully for the diagnosis of this anomaly.[14] Of note, cardiac catheterization remains the gold standard;[3] however, computed tomography scan also allows a diagnosis of certainty, as in our case.

Surgical treatment intends to redirect the flow of the IVC in the right atrium. Kim et al.[6] carried out the surgical correction by suturing the upper edge of the interatrial communication directly to the floor of left atrium by several separate points. In our case, we used a Dacron patch sewn at the upper edge of the IVC and the upper edge of the interatrial communication to avoid distorting the mitral annulus, which, indeed, was very close to the junction where the IVC connects to the left atrium.

In conclusion, this is a rare case of abnormal connection of the inferior vena cava to the left atrium associated with severe mitral regurgitation which altered the clinical presentation. Surgical correction was achieved successfully by redirecting the inferior vena cava flow to the right atrium and mitral valve repair.

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