Case Report / Olgu Sunumu
doi: 10.18678/dtfd.1580769

Duzce Med J, 2025;27(1):102-104
Düzce Tıp Fak Derg, 2025;27(1):102-104

Tetralogy of Fallot, Absent Vena Cava Superior, Persistent Left Vena Cava Superior, and Interrupted Vena Cava Inferior: A Case of Struggle in Open Heart Surgery

Fallot Tetralojisi, Vena Cava Süperior Yokluğu, Persistan Sol Vena Cava Süperior ve Kesintili Vena Cava İnferior: Açık Kalp Cerrahisindeki Güçlük Üzerine Bir Olgu

Hande İŞTAR
0000-0002-7150-0171
Buğra HARMANDAR
0000-0002-7487-1779

Department of Cardiovascular Surgery, Muğla Sıtkı Koçman University Faculty of Medicine, Muğla, Türkiye

ABSTRACT

Interruption of vena cava inferior (VCI) is extremely rare, in occurrence in isolation or association with asplenia or polysplenia syndromes. In this abnormality, the infrahepatic segment of the VCI is absent, by representing the inadequacy of fusion of subcardinal embryological parts of the VCI. It is compensated by an azygos or hemiazygos vein that continues on the posterior wall of the thoracic cavity. In this case report, a case of incidentally diagnosed interrupted VCI in a patient diagnosed with tetralogy of Fallot (TOF), major aortopulmonary collateral artery (MAPCA), right arcus aorta, absence of right vena cava superior (VCS), and persistent left vena cava superior (PLVCS), and its successful surgical treatment was presented. If the hepatic vein confluence is of adequate size, the cannulation for cardiopulmonary bypass circuit through the hepatic vein confluence is safe in case of interrupted VCI.
Keywords: Superior vena cava; inferior vena cava; persistent left vena cava superior; tetralogy of Fallot.

ÖZ

Kesintili vena kava inferior (vena cava inferior, VCI), izole olarak ya da aspleni veya polispleni sendromlarıyla birlikte meydana gelmesiyle, oldukça nadir olarak görülür. Bu anomalide, vitellin ve subkardinal embriyolojik VCI'nın füzyonundaki yetersizlik nedeniyle, VCI'nın infrahepatik segmenti gelişmemiştir. Bu durum, torasik boşluğun posterior duvarında devam eden azygos ya da hemiazygos veni tarafından kompanze edilir. Bu olgu sunumunda, Fallot tetralojisi (tetralogy of Fallot, TOF), majör aortikpulmoner kollateral arter (major aortopulmonary collateral artery, MAPCA), sağ arkus aorta, sağ vena kava süperior (vena cava superior, VCS) yokluğu ve persistan sol vena kava süperior (persistent left vena cava superior, PLVCS) tanısı olan bir hastada tesadüfen teşhis edilen kesintili VCI vakası ve başarılı cerrahi tedavisi sunulmuştur. Hepatik ven konflüensi yeterli büyüklükteyse, kesintili VCI durumunda hepatik ven konflüensinden açık kalp cerrahisi dolaşımı için kanülasyon güvenlidir.
Anahtar kelimeler: Superior vena kava; inferior vena kava; persistan sol vena kava superior; Fallot tetralojisi.

Corresponding Author
Sorumlu Yazar
Hande İŞTAR
hande13@msn.com

Received / Geliş Tarihi : 06.11.2024
Accepted / Kabul Tarihi : 13.02.2025
Available Online / Çevrimiçi Yayın Tarihi : 12.03.2025

INTRODUCTION

Congenital heart surgery is challenging by nature. Different intra and extra cardiac abnormalities may be in close relation in the same patient, and it may be difficult to choose which abnormality is of the greatest importance and the priority for surgical treatment. For example, cyanotic and abnormal systemic venous return can present together. In isolation, interrupted vena cava inferior (VCI) is very rare and a compensatory azygos or hemiazygos vein continuity is present. Interrupted VCI and

azygos or hemiazygos continuity are present in occurrence with 0.6-2% of people with congenital heart diseases, <0.3% in isolation in normal people (1). It is associated in the literature with tetralogy of Fallot (TOF), truncus arteriosus, absence of the pulmonary artery, atrioventricular septal defect, pulmonary valvular stenosis, pulmonary atresia, interrupted aortic arch, atrial septal defect, coarctation of aorta, pulmonary venous stenosis, common atrium, common atrioventricular valve, and double outlet right ventricle (2-4). Here we share our experience of the surgical correction of TOF, major aortopulmonary collateral artery (MAPCA) in association with interrupted VCI, absence of vena cava superior (VCS), right arcus aorta, and persistent left vena cava superior (PLVCS).

CASE REPORT

A 2-year-old female, 10 kg in weight, patient was admitted to our center due to cyanosis with a room air saturation of 78%. She was diagnosed on echocardiography with features of TOF and a MAPCA arising from the ascending aorta. A PLVCS was enlarged; however, the right VCS was not well determined. The right aortic arch and an interrupted VCI were present. On catheterization, the exact anatomy was determined and an interrupted VCI was seen (Figures 1A, and 1B). The McGoon ratio was measured as 2.18 due to well-developed right and left pulmonary arteries. We decided total correction for the TOF and MAPCA pathologies in light of the adequate McGoon ratio and low room air saturation. Informed consent was taken from the patient's relatives.

Following median sternotomy, we observed that the hypoplastic main pulmonary artery was situated to the left side of the aorta. The right aortic arch and a well-developed PLVCS were present. Right-sided VCS was not observed and the innominate vein course was through the coronary sinus. Instead of an appropriate entrance of the VCI, we observed an enlarged confluence made by multiple hepatic veins with a central course and a central entrance to the right atrium (Figure 1C). After systemic heparinization, aortic and PLVCS cannulations were performed as usual. MAPCA division was performed. Following cardiopulmonary bypass (CPB) establishment, cannulation of the hepatic vein confluence was performed. Systemic hypothermia (32° C) and Custodiol cardioplegia delivered via the antegrade route were used. Through the right atriotomy and atrial septectomy, a sump cannula was inserted in the left atrium. Another sump cannula was used for the aspiration of blood coming from the innominate vein through the coronary sinus. Venous return to the reservoir of the CBP machine was safe and adequate in spite of the unusual venous source due to the challenging anatomy. Ventricular septal defect closure was performed using a Gore-Tex graft. The muscle resection was performed to resolve the right ventricular outflow tract. The main pulmonary artery was opened longitudinally. We observed a bicuspid pulmonary valve. Glutaraldehyde-treated autologous pericardium was fashioned to the main pulmonary artery in an extended manner, cross annularly to obtain an enlargement for the right ventricular outflow. The atrial septum was repaired and weaning from CPB was performed as usual. Following decannulation of the hepatic vein confluence, we did not observe any stenosis. The postoperative course was uneventful. The patient was discharged on the 10th day postoperatively. Throughout the hospitalization, we did not observe any abnormal liver test results or hepatomegaly. No rhythm disturbance occurred postoperatively.

DISCUSSION

Interrupted VCI and azygos or hemiazygos venous continuity occurs in 0.6 to 2% of people with congenital heart diseases. It occurs <0.3% in isolation in normal people (1). Interrupted VCI and azygos or hemiazygos continuity at the level of the liver frequently presents in patients in addition to heterotaxy syndrome, who exhibit left isomerism of the atrial appendage, i.e. polysplenia syndrome. Chen et al. (4) performed a study of imaging of interrupted VCI and concluded that 96.4% of the cohort had left atrial isomerism. Splenic abnormalities were common and 11.8% had a normal cardiac structure; however, 30 of the 34 patients had a simple to severe degree of congenital heart disease. The most frequently associated congenital heart diseases were pulmonary stenosis or atresia and double outlet right ventricle, in ratio of 70.6% and 61.8% of patients, respectively (4). 44.1% of cohort studies showed that sinus bradycardia can occur in

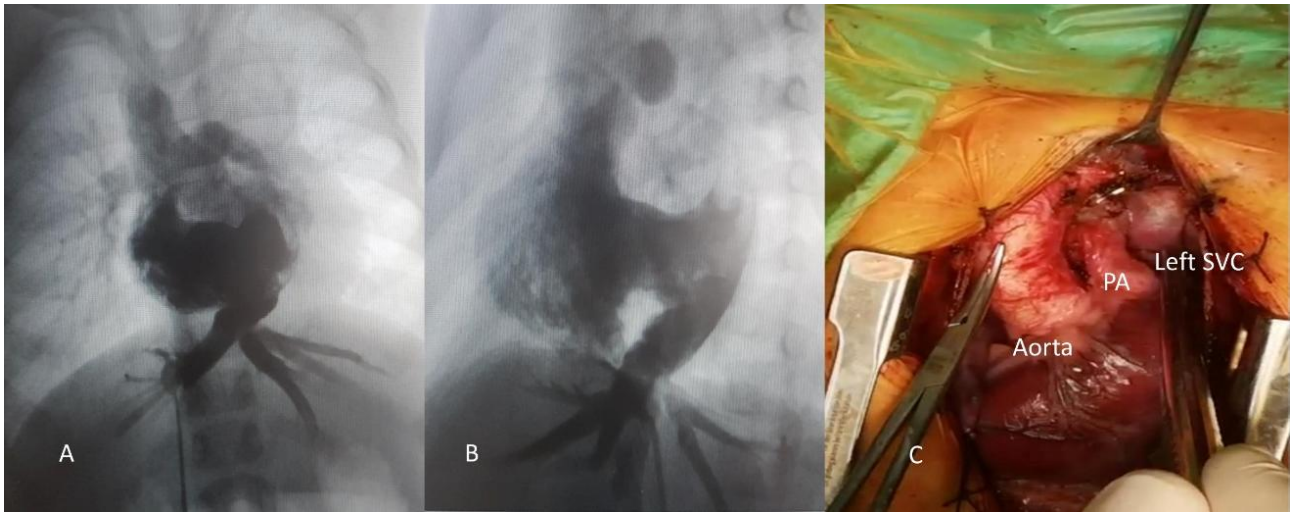

Figure 1. A, B) Angiographic view of hepatic vein confluence, **C)** intraoperative view of persistent left vena cava superior (PLVCS)

the long-term outcome. In the literature, five types of interrupted VCI have been described (5), and the most common type is interruption of the hepatic segment (4). As a result, to maintain the antegrade flow in intrauterine life, some venous anastomosis between right supracardinal and right subcardinal veins persist. They result in azygos and hemiazygous continuations (4). Due to their strong associations, an interrupted VCI should warn us of the presence of additional congenital heart diseases.

Before planning a cardiac intervention via the femoral vein or cardiac surgery in a patient with an interrupted VCI, preoperative awareness is important. Particularly in open heart surgery, while inserting the venous cannula into the VCS and VCI for CPB, two adequately sized veins are required. In the case of interrupted VCI, hepatic vein confluences have to be cannulated instead. Snaring hepatic veins after cannulation and inadequate drainage may therefore result in postoperative elevated hepatic enzymes and hepatomegaly. Moreover, in cases of intracardiac surgical repair, it is not adequate to only snare the VCS. To obtain a good exposure, it should be performed by direct sucking of the hepatic veins besides hypothermia with low flows, moreover the total circulatory arrest (6).

To complete the single ventricle type repair, it is necessary to redirect the VCI drainage into the right pulmonary artery. However, if an interrupted VCI is present and azygos or hemiazygos continuity drain the flow to the brachiocephalic vein, the Kawashima operation (consisting of end-to-side anastomosis of the VCS and the azygos or hemiazygos continuation and the confluent pulmonary artery, and division or ligation of the pulmonary artery trunk) is required. Except for hepatocardiac venous and coronary sinus flow, the total venous return drains directly into the pulmonary artery, bypassing the right-sided heart, i.e., right ventricle and right atrium (7).

TOF can present with other venous return abnormalities. Shah et al. (8) presented a case report that both the VCS were absent and only the VCI and ambiguous innominate vein were draining the systemic venous flow into the cardiac cavity. In our case, the surprising intraoperative discovery of the absence of both VCS made the TOF total repair difficult while cannulation before CPB. However, after cannulation of the VCI and the ambiguous innominate vein, also with total circulatory arrest, TOF total correction became possible (8).

Even though isolated interrupted VCIs are asymptomatic in the general population, there have been some case reports of lower limb oedema, pelvic vein or pulmonary thrombosis, and deep venous thrombosis as severe symptoms (9,10).

CONCLUSION

Our case was challenging due to an absent right VCS, interrupted VCI, PLVCS, and well-developed hepatic vein confluence when we performed venous cannulation before repairing the TOF and MAPCA. The hepatic vein drainage was fortunately adequate in the CPB period. We did not observe a serious hepatic dysfunction postoperatively. We believe this association is the first to be published in the literature. Further investigations are crucial.

Informed Consent: Written informed consent was obtained from the patient for publication and accompanying images.

Conflict of Interest: None declared by the authors.

Financial Disclosure: None declared by the authors.

Acknowledgments: None declared by the authors.

Author Contributions: Idea/Concept: Hİ; Design: Hİ; Data Collection/Processing: Hİ; Analysis/Interpretation: BH; Literature Review: Hİ, BH; Drafting/Writing: Hİ; Critical Review: BH.

REFERENCES

1. Zafar SI, Halim A, Khalid W, Shafique M, Nasir H. Two cases of interrupted inferior vena cava with azygos / hemiazygos continuation. J Coll Physicians Surg Pak. 2022;32(8):S101-3.
2. Bista B, Ferris J, Na N, Krishnam M, Urgun D. Unilateral absence of pulmonary artery and azygous continuation of interrupted inferior vena cava. Radiol Case Rep. 2020;15(6):688-90.
3. Bansal N, Ghosh R, Sankhyan LK, Chatterjee S, Chatterjee S, Bose S. Left isomerism with bilateral superior vena cava, interrupted inferior vena cava and tetralogy of Fallot. Ann Pediatr Cardiol. 2020;13(4):364-7.
4. Chen SJ, Wu MH, Wang JK. Clinical implications of congenital interruption of inferior vena cava. J Formos Med Assoc. 2022;121(10):1938-44.
5. Abu-Hilal LH, Barghouthi DI, AbuKeshek T, Tamimi H, Khatib H, Dayeh AH. Interrupted inferior vena cava syndrome discovered incidentally after minimally invasive mitral valve repair in a 31-year-old female patient: A case report. Int J Surg Case Rep. 2023;109:108621.
6. Pillai JB, Kpodonu J, Yu C, Borger MA. Heterotaxy syndrome with azygous continuation-causing pseudo Budd-Chiari syndrome after cardiopulmonary bypass. Ann Thorac Surg. 2006;81(5):1890-2.
7. Kawashima Y, Kitamura S, Matsuda H, Shimazaki Y, Nakano S, Hirose H. Total cavopulmonary shunt operation in complex cardiac anomalies: A new operation. J Thorac Cardiovasc Surg. 1984;87(1):74-81.
8. Shah TR, Hiremath CS, Diwakar A, Soman Rema KM. Absent superior vena cava in tetralogy of Fallot. Ann Card Anaesth. 2018;21(2):205-7.
9. Cheung CKM, Law MF, Wong KT, Tam MTK, Chow KM. Massive pulmonary embolism in a patient with polysplenia syndrome and interrupted inferior vena cava with azygous continuation. Arch Med Sci. 2018;14(1):251-3.
10. Koc Z, Oguzkurt L. Interruption or congenital stenosis of the inferior vena cava: prevalence, imaging, and clinical findings. Eur J Radiol. 2007;62(2):257-66.