The Cardiothoracic Surgeon

CASE REPORT Open Access

An adult with left isomerism having an interrupted inferior vena cava and persistent left superior vena cava associated with partial atrioventricular canal defect and complete heart block — a case report

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Abstract

Background: Persistent left superior vena cava is a well-recognized thoracic venous abnormality, most of which do not present with any symptoms. Its opening into the left atrium is uncommonly encountered and, if present, is noted to be associated with various other complex intracardiac malformations, which further complicates its management. Herein, we highlight a case of an adult with persistent left superior vena cava with a spectrum of other cardiac anomalies rarely encountered with it, which was effectively managed by tailored surgical intervention.

Case presentation: A 26-year-old female on meticulous evaluation was found to have left isomerism with an interrupted inferior vena cava and hemiazygos continuing as a persistent left superior vena cava. It was also associated with common atrium, partial atrioventricular canal defect, and complete heart block. This complex array of anomalies was managed surgically. Common atrium was closed with atrial neo-septation using autologous pericardial patch tailored in a way, such that the persistent left superior vena cava drained into right atrium with left atrial appendage being on right side of neo-septum. Cleft in the left atrioventricular valve was also repaired. Despite preoperative complete heart block, the patient had an uneventful postoperative recovery and was in atrial fibrillation with controlled ventricular rate, thus precluding the need for permanent pacemaker implantation.

Conclusions: A rare spectrum of adult congenital cyanotic heart disease was successfully managed with thorough preoperative evaluation and judicious intraoperative management. Surgical management should be tailored based on specific anatomy for reduction of morbidity and optimum results.

Keywords: Heterotaxy syndrome, Persistent left superior vena cava, Partial atrioventricular canal defect, Common atrium

Background

Persistent left superior vena cava (PLSVC) is a well-recognized thoracic venous abnormality [1], most of which do not present with any symptoms. It is an ancillary finding, with prevalence of 0.2 to 3% in general population and 1.3 to 11% in patients with congenital heart defects. It most frequently drains into the right atrium through coronary sinus. Its drainage and opening into the left atrium (LA)

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are uncommon, but if present, it is associated with various complex intracardiac malformations such as heterotaxy syndrome, interrupted inferior vena cava (IVC), atrioventricular canal defect (AVCD), and single-ventricle physiology [1–5], which further complicates its management.

We hereby report successful management of a rare case of an adult presentation of heterotaxy syndrome with left isomerism, interrupted IVC which continues as hemiazygos, draining into PLSVC, which in turn opens into left side of common atrium, and also associated with cleft left atrioventricular (AV) valve, partial AVCD, and complete heart block.

Case presentation

A 26-year-old female presented with cyanosis since childhood and dyspnea functional class II (New York Heart Association) since the past 2 years. She had repeated episodes of loss of consciousness, for which she was investigated by computed tomography of the brain. It revealed no significant abnormality.

On physical examination, the patient had central cyanosis with oxygen saturation of 64% at room air. Cardiovascular system examination revealed a grade 3/6 pansystolic murmur in mitral area. Per-abdomen examination showed the absence of fundal tympanic note in the left hypochondrium, suggesting heterotaxy syndrome clinically.

On further evaluation, chest X-ray revealed cardiomegaly, right ventricular type of apex and bi-atrial enlargement, and prominent pulmonary arteries with plethoric lung fields. Electrocardiography showed complete heart block with heart rate of 50/min (Fig. 1A). Transthoracic echocardiography showed common atrium, normal pulmonary venous drainage, and partial AVCD with two distinct AV valves. There was mild right AV valve regurgitation and moderate left AV valve regurgitation due to cleft with no pulmonary stenosis. Right ventricular systolic pressure (RVSP) was 34 mmHg with normal biventricular function (Fig. 1B).

A cardiac contrast-enhanced computed tomography was performed, which showed interrupted IVC continuing as hemiazygos, draining into PLSVC which in turn opened into left side of common atrium. Hepatic veins and right superior vena cava (RSVC) opened into right side of the common atrium. Dilated pulmonary artery, transverse liver, right-sided gastrium, and polysplenia with bilateral morphologic left bronchi (Fig. 2) were other significant findings noted. Cardiac catheterization was performed, which revealed left to right shunt at atrial level with additional anatomical delineation (Fig. 1 C and D). PVR was not measured during cardiac catheterization; however, we considered proceeding with surgery based on the normal RVSP in the echocardiography.

Intraoperatively, all radiological findings were confirmed (Fig. 3A). RSVC was found to be draining normally into right side of the common atrium with no bridging innominate vein. Left atrial isomerism was also noted.

Cardiopulmonary bypass was established with cannulation of RSVC, PLSVC, and cardiac part of IVC on the right side. After satisfactory cardioplegic arrest, right side of common atrium was opened, and the anatomy was inspected. Although the right AV valve was competent, the left AV valve was moderately regurgitant with cleft. Hence, it was repaired with interrupted polypropylene sutures (Fig. 3B).

The opening of left atrial appendage (LAA) was narrow and closely related to the opening of PLSVC. An attempt to leave the LAA opening into LA would cause kinking of the baffle, which may further lead to obstruction of PLSVC drainage. Hence, the atrial neo-septation with pericardial patch was tailored, such that the PLSVC drained into right atrium, with LAA being on the right side of neo-septum (Fig. 3C and Fig. 4).

The patient came off cardiopulmonary bypass in junctional rhythm with regular RR interval, narrow QRS complexes, and heart rate of 80/min. However, two atrial and two ventricular epicardial pacing wires were put in view of preoperative complete heart block. Intraoperative transesophageal echocardiography was performed, which showed intact neo-septal pericardial patch with no residual shunt and unobstructed normal systemic and pulmonary venous drainage. LA was adequately sized with trivial left AV valve regurgitation having gradient of 7/4 mmHg and trivial right AV valve regurgitation with RVSP of 28 mmHg. Normal biventricular function was noted (Fig. 3D).

The patient had an uneventful postoperative recovery. Patient remained with stable heart rate and did not require epicardial pacing in the entire postoperative period. Despite preoperative complete heart block, the patient regained junctional rhythm with good ventricular rates in immediate postoperative period, which settled to atrial fibrillation with controlled ventricular rate at the time of discharge, thus precluding the need for permanent pacemaker implantation. At discharge, transthoracic echocardiography findings were consistent with intraoperative transesophageal echocardiography findings. Oral anticoagulation was considered to prevent thromboembolic complications due to atrial fibrillation, and long-term follow-up was recommended for monitoring the rhythm.

She was asymptomatic at 1 year follow-up and continues to be in atrial fibrillation with controlled ventricular rate on oral anticoagulants.

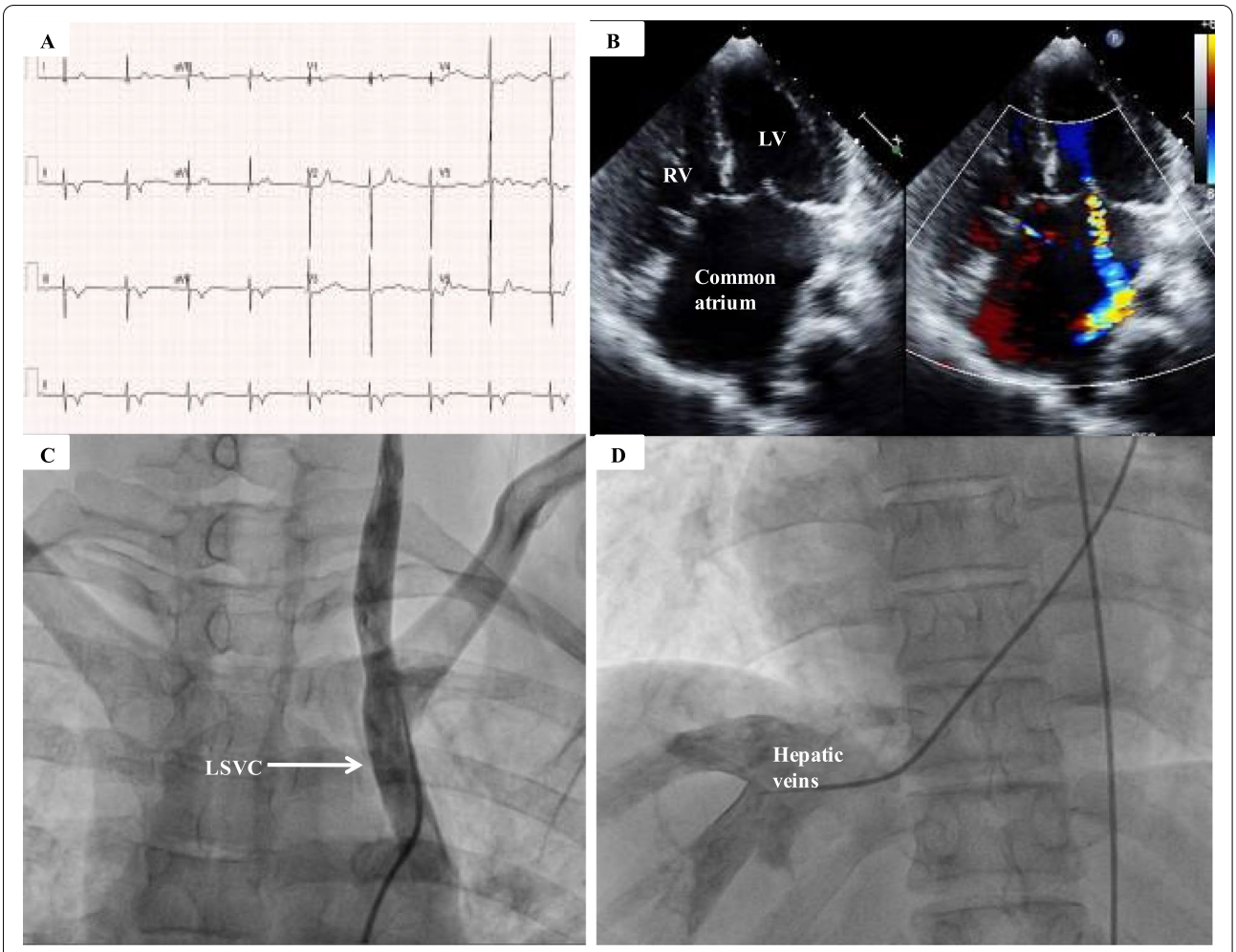

Fig. 1 **A** Preoperative electrocardiography showing complete heart block. **B** Apical 4-chamber transthoracic echocardiography showing common atrium with moderate left AV valve regurgitation. **C** Cardiac catheterization showing IVC continuing as hemiazygos — PLSVC and draining to left part of common atrium. **D** Cardiac catheterization showing catheter course: IVC — hemiazygos vein, PLSVC — common atrium — hepatic veins

Discussion

PLSVC is the most prevalent thoracic venous abnormality [1]. PLSVC opening into the LA is associated with various complex intracardiac malformations [1, 3]. Bilateral SVC is also a component of the right and left isomerism syndromes, which are further associated with complex cardiac malformations such as common atrium, interrupted IVC, AVCD, and single-ventricle physiology [1, 4, 5].

The PLSVC descends lateral to the aortic arch and anterior to hilum of the left lung. It enters the pericardium in the posterior atrioventricular groove into the coronary sinus. It can drain into coronary sinus or LA. When it drains into LA, it may form a part of unroofed coronary sinus syndrome [4]. The presence of PLSVC opening into LA with intracardiac malformations such as common atrium, interrupted IVC, and AVCD [1, 4, 5] further complicates the surgical management.

Common atrium is defined as the condition characterized by complete or near-complete absence of interatrial septum, with or without AVCD [6]. It has been reported in patients with heterotaxy syndrome, Ellis-van Creveld syndrome, and AVCD with or without associated Down syndrome [7].

Traditionally, such cases have been managed with the creation of a neo-interatrial septum in such a way that the baffle redirects the opening of PLSVC into right atrium and pulmonary veins into LA. The opening of PLSVC is relatively constant, anteriorly to the opening of left superior pulmonary vein, lying posterior and inferior to the opening of LAA [1, 3]. Yaku et al. in their study pointed out that adequate partitioning of the common chamber is the key to avoiding systemic or pulmonary venous

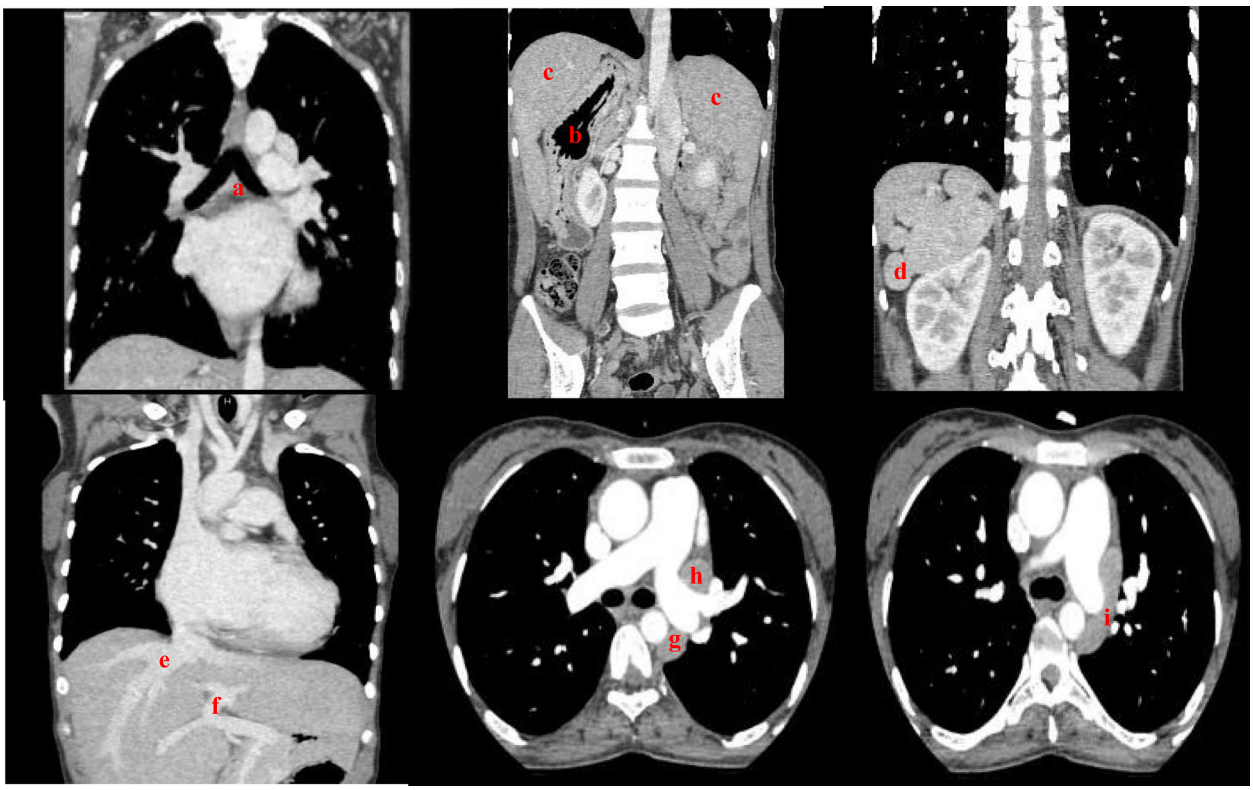

Fig. 2 Cardiac CECT demonstrating the following: (**a**) bilateral hyparterial bronchi, **b** right-sided gastrium, (**c**) transverse liver, (**d**) multiple spleniculi, (**e**) hepatic vein draining into right side of common atrium, (**f**) infra-hepatic interrupted IVC, (**g**) hemizygos continuation of IVC, (**h**) PLSVC, (**i**) confluence of hemiazygos into PLSVC

obstruction [8]. Postoperative systemic venous obstruction due to kinking of the baffle is a known complication of the procedure, requiring reoperation [8]. Hence, it is vital to place the neo-atrial septum without obstruction to systemic and pulmonary venous drainage. In our patient, the LSVC opening and the LAA orifice were in close proximity to each other, making it difficult to create a baffle without obstructing the systemic venous drainage. We considered it safe to redirect the opening of the LAA into the right atrium while creating the neo-interatrial septum, so as to ensure an unobstructed systemic venous drainage. To our knowledge, this technique of neo-septation with LAA on the right side has not been described in published literature with equivalent outcomes as compared to traditional neo-septation (Fig. 4). Thereby, we would like to highlight the positive outcome of this technique of neo-septation in our case.

Conclusions

A rare spectrum of adult congenital cyanotic heart disease comprising of hemizygos continuation to a persistent left superior vena cava with an interrupted inferior vena cava and left isomerism associated with partial atrioventricular canal defect and complete heart block was successfully managed with thorough preoperative evaluation and judicious intraoperative management. Surgical management should be tailored based on specific anatomy of systemic and pulmonary venous drainage for reduction of morbidity and optimum results.

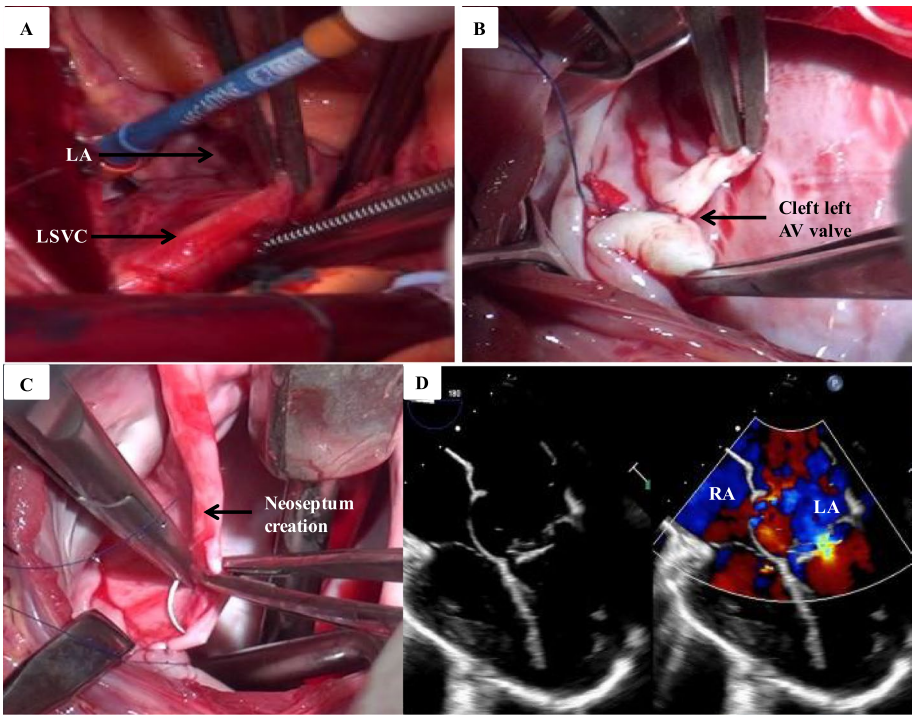

Fig. 3 Intraoperative images demonstrating the following: **A** LSVC draining to LA, **B** partially closed cleft of left AV valve, **C** creation of neo-septum using autologous untreated pericardium, **D** post repair transesophageal echocardiography demonstrating intact neo-septum with trivial left AV valve regurgitation

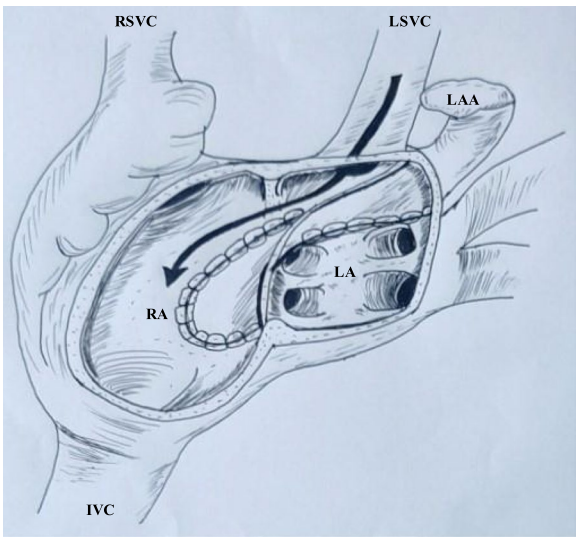

Fig. 4 Surgical diagram depicting atrial neo-septation with pericardial patch tailored such that the PLSVC drains to RA with LAA on right side of neo-septum. AV, atrioventricular; IVC, inferior vena cava; LA, left atrium; LAA, left atrial appendage; LSVC, left superior vena cava; LV, left ventricle; RA, right atrium; RSVC, right superior vena cava; RV, right ventricle

Abbreviations
AV: Atrioventricular; AVCD: Atrioventricular canal defect; IVC: Inferior vena cava; LA: Left atrium; LAA: Left atrial appendage; PLSVC: Persistent left superior vena cava; RSVC: Right superior vena cava; RVSP: Right ventricular systolic pressure.

Acknowledgements
Not applicable

Authors' contributions
All authors contributed equally in the case report as follows: concept/design, data collection, and analysis/interpretation, MWA, SMH, RI, and ND. Critical revision and approval of the article: ND. The authors read and approved the final manuscript.

Funding
This study has not been funded by any party whatsoever.

Availability of data and materials
The data that supports the findings of this study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
Consent for publication was obtained.

Competing interests
The authors declare that they have no competing interests.

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Received: 5 April 2022 Accepted: 27 May 2022
Published online: 04 June 2022

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