

Partial endocardial cushion defect with Raghیب's syndrome: a rare case report

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Raghیب's syndrome is characterized by the combination of abnormal drainage from the left superior vena cava into the left atrium, the presence of an atrial septal defect, and the absence of a coronary sinus. Although surgical repair of Raghیب's syndrome is often indicated, its correction in combination with partial endocardial cushion defect is extremely rare. In this article, we present a 24-year-old female patient with partial endocardial cushion defect accompanied by Raghیب's syndrome who was corrected successfully with surgery.

Keywords: Coronary sinus; endocardial cushion defect; superior; vena cava.

Absence of coronary sinus is a rare eventuality in persistent left superior vena cava (PLSVC), which is usually associated with interatrial defect.^[1] In general, PLSVC drains into the right atrium through the coronary sinus (92%) or directly into the left atrium (8%, unroofed coronary sinus).^[2] The latter may lead to right-to-left shunting, cyanosis, paradoxical embolism, and a misdiagnosis of Eisenmenger's syndrome.

In this article, we report a 24-year-old female case with partial endocardial cushion defect and coronary sinus atresia, unroofed coronary sinus, and PLSVC draining directly into the left atrium, which was diagnosed intraoperatively and corrected successfully.

CASE REPORT

A 24-year-old female patient was admitted to our clinic with symptoms of dyspnea and fatigue. She had no history of rheumatologic disease or rheumatic fever or Down syndrome. The results of the physical examination were within normal limits without clubbing or cyanosis. On cardiac examination, the patient had a pansystolic murmur in the fifth intercostal space on the left side at the midclavicular line. Her vital signs were normal with an oxygen saturation of 90%. An electrocardiogram showed sinus rhythm and left axial deviation. Echocardiography demonstrated a large ostium primum defect and third-degree mitral regurgitation (Figure 1). No other associated cardiac malformations were demonstrated.

After written informed consent, median sternotomy and aortic-bicaval cannulation were

performed under cardiopulmonary bypass. In the inspection of the venous anatomy, the innominate vein was absent. After aortic cross-clamping and antegrade blood cardioplegia administration, right atriotomy was performed. The coronary sinus was not visualized. Multiple Thebesian vein ostiums in the right atrium were observed. Persistent left superior vena cava was draining to the left atrial roof. A vent sucker was placed in the PLSVC and snared for good operative exposure and prevention of brain-related complications. There were normal pulmonary venous connections. There was also a cleft in the mitral valve anterior leaflet.

Surgical correction was performed initially. The cleft in the mitral valve was repaired using multiple interrupted 6-0 polypropylene sutures and the flow of the PLSVC was diverted to the right atrium including Thebesian veins with an 8 mm polytetrafluoroethylene (PTFE) prosthetic tube graft, which was cut vertically (Figure 2). The atrial septum was reconstructed with an autologous pericardial patch which was used for closing primum ASD (Figure 2).

Postoperatively, oxygen saturation improved to 99%. Following an uneventful recovery, the patient was discharged in the fifth postoperative day. Postoperative anticoagulant therapy was administered for eight weeks. At sixth months, she was free of symptoms and in sinus rhythm. Repeated echocardiography also

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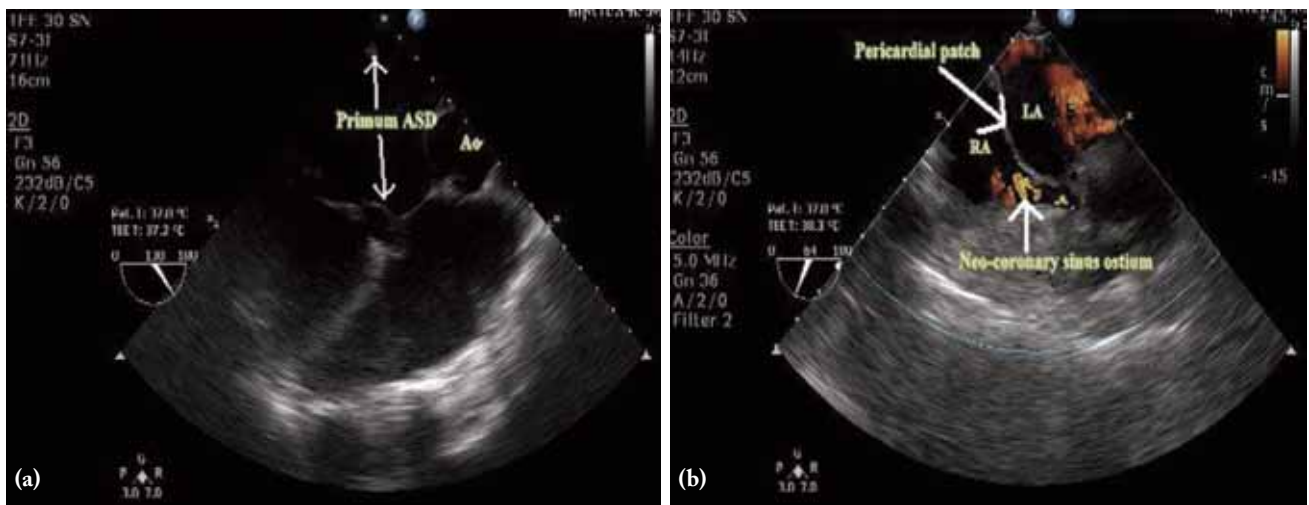


Figure 1. (a) Preoperative and (b) postoperative transesophageal echocardiography.

showed a mild mitral insufficiency and patent flow of the PLSVC (Figure 1).

DISCUSSION

Persistent left superior vena cava is a congenital anomaly which affects 0.3% of healthy population and 1.3-10% of patients with cardiovascular disease.^[3] An association between PLSVC and absence of coronary sinus is an extremely unusual condition, usually associated with an inter-atrial defect.^[3,4] More unusual is the PLSVC, absence of coronary sinus, and partial atrioventricular septal defect.

Some authors hypothesized that this unusual anatomy is due to the failed process of embryonic venous

system lateralization at the level of the left horn and left anterior cardinal vein.^[5] The left anterior cardinal vein follows the same development of the right, while the last part of the left horn remains high on the left and behind the left atrium instead of becoming the coronary sinus. Because the left horn does not mitigate, the ostium of coronary sinus will not form.^[5]

Furthermore, clinical complications of this anomaly are cyanosis and reduced strain tolerance.^[2] Therefore, PLSVC carries more importance in the event of cardiovascular surgery (absolute contraindication of retrograde cardioplegia), central venous, and permanent transvenous pacing lead placement.^[6,7] This anomaly may also cause a misdiagnosis such as right-to-left shunting or Eisenmenger's syndrome.

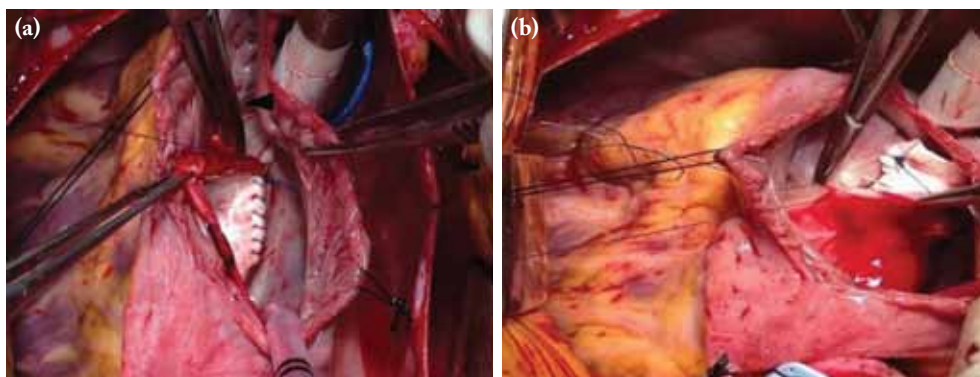


Figure 2. (a) The orifice of polytetrafluoroethylene baffle, which was used to divert the blood from the orifices of the persistent left superior vena cava into the right atrium seen under the autologous pericardial patch. ► The clamp was inside the venous baffle. (b) The primum atrial septal defect was closed with an autologous pericardial patch over the polytetrafluoroethylene baffle and the completed venous baffle directed persistent left superior vena cava flow to the right atrium.

Several surgical procedures in the correction surgery have been reported including ligation of the left SVC, intra-atrial redirection of flow from the left SVC to the right atrium, and re-implantation of the left SVC into the right atrium, pulmonary artery or SVC.^[8] Ligation of the vein obliterates the intracardiac shunt, however, this procedure is risky, unless there are large collateral links in the head which allow non-obstructed head and neck venous return into the heart. Re-implantation of the persistent left SVC is preferable, when there is a possibility that an intra-atrial baffle may obstruct systemic or pulmonary venous return due to the location of the veins orifices, in particular.^[8-10]

In conclusion, our case had partial endocardial cushion defect with an absent coronary sinus and PLSVC without any communication between SVCs. In our case, thebesian veins were opening on way of the baffle and there was the absence of intercaval communication and adequate left atrium volume. Therefore, we used a tube graft PTFE material, as we thought that the selected material could be more resistant to compression from the left atrium pressure.

Declaration of conflicting interests

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