

Extra-anatomic correction of persistent left superior vena cava draining into the left atrium associated with atrial septal defect.

Maria Grandinetti¹, Arianna Di Molfetta¹, Piero Farina², Gianluigi Perri³, Francesca Graziani¹, Angelica Bibiana Delogu¹, Rosa Lillo¹, Natalia Pavone¹, Piergiorgio Bruno¹, Antonio Amodeo⁴, and Massimo Massetti¹

¹Policlinico A Gemelli

²Catholic University

³Bambino Gesù Pediatric Hospital

⁴Bambino Gesù Children Hospital

July 31, 2020

Abstract

Persistent left superior vena cava draining into the left atrium is a rare condition. We herein report the case of a 34-year-old male admitted at our institution for paroxysmal atrial fibrillation. Investigations revealed a large ostium secundum atrial septal defect, with a persistent left superior vena cava draining into the left atrium, while the coronary veins drained separately into the left atrium. Surgical correction was performed, closing the defect with a patch and connecting the left superior vena cava to the right atrial appendage with an extra-cardiac conduit.

History of presentation

A 34-year-old male was admitted to our hospital following the sudden onset of palpitation and shortness of breath. His past medical history, otherwise unremarkable, included hypertension and overweight (BMI 28.2 Kg/m²). Physical examination revealed clubbing and low SpO₂ (90%).

Investigations

Electrocardiogram revealed atrial fibrillation, which reverted spontaneously to sinus rhythm. Transthoracic echocardiogram revealed a large *ostium secundum* atrial septal defect (ASD), 41x42mm in size, with left-to-right shunt, pulmonary hypertension (60mmHg) and dilatation of the right ventricle. Computed tomography (CT) scan showed: (1) a persistent left superior vena cava (LSVC) draining into the left atrium (LA) in close proximity to the left atrial appendage, (2) the absence of a left brachiocephalic vein, (3) the abnormal independent drainage of the coronary veins in the LA. The great cardiac vein drained into the postero-lateral wall, the marginal vein into the lower wall and two middle cardiac veins into the medial-lower wall of the LA (Fig. 1; Fig 2). Cardiac catheterisation confirmed pulmonary hypertension (pulmonary arterial pressure 70/44mmHg; mean 45mmHg, wedge pressure 14mmHg), arterial SpO₂ 91%, systemic venous saturation 70% and pulmonary artery saturation 90%, Qp/Qs 2.8. Nitric oxide caused a significant reduction of pulmonary artery and wedge pressures, with an increment of the Qp/Qs to 11.5. The patient was referred and accepted for elective surgery.

Management

Surgery was performed through full median sternotomy. The absence of the left brachiocephalic vein was confirmed. The LSVC entered the LA cephalad and medial to the left superior pulmonary vein (Fig 3 and

Fig 4). Cardiopulmonary bypass was established, with bicaval cannulation for venous drainage. The aorta was crossclamped and the heart was arrested with a single shot of cold histidine-tryptophan-ketoglutarate (HTK) solution delivered in antegrade fashion. The right atrium was opened and the ostium of the LSVC was examined through the large ASD. The close proximity of the ostium of the LSVC to the ostium of the left superior pulmonary vein precluded the placement of an interatrial baffle, therefore the ASD was closed with an autologous pericardial patch. The LSVC was divided, its distal end ligated and its proximal end anastomosed to the right atrium with the interposition of a 16mm Gore-Tex vascular graft (Gore, Tempe, AZ)(Fig 3). The patient had an uneventful postoperative course and made a full recovery. Four years following surgery, he is in excellent general condition and the echocardiogram shows no residual shunt, with a normal right ventricle and normal pulmonary pressures.

Discussion

Persistence of the left superior vena cava (LSVC) is the most common anomaly of the systemic venous connection, and in the absence of associated defects may be considered a variant of normal. Its presence can be relevant when a central venous catheter or a transvenous pacemaker is needed. Also, a LSVC can interfere with optimal venous drainage and exposure during some cardiac surgical procedures [1-6]. A persistent LSVC usually drains into the coronary sinus, but sometimes can drain into the left atrium leading to right-to-left shunt. When needed, surgical connection is possible with either intra- or extra-cardiac procedures [1-6]. The few cases reported in literature have been treated with: *a*) division and re-implantation of the LSVC to right atrium [1,4]; *b*) an intra-atrial baffle or graft to divert flow from the LSVC to the right atrium and to close the ASD [1, 2, 4, 7]; *c*) anastomosis of the LSVC to the left pulmonary artery [6,8,9] or *d*) simple ligation of the LSVC [1, 2, 3, 4-6, 8].

Our patient presented with a combination of persistent LSVC draining into the LA, a large *ostium secundum* ASD, coronary veins draining into the LA and pulmonary hypertension responding to pulmonary vasodilators. The multiple anomalous connections between the systemic and pulmonary circulation did not translate into a particularly high Qp/Qs, likely due to concomitant presence of left-to-right and right-to-left shunts. However, as fast fibrillation entered the picture the patient decompensated. An interatrial baffle would have been our treatment of choice, but the ostium of the left superior pulmonary vein was at risk of obstruction due to its proximity to the ostium of the LSVC. Therefore, an extracardiac technique was used.

As there is no standardised procedure for the correction of a persistent LSVC draining into the LA, whenever surgery is considered, the choice of the technique should take into account several variables such as the patient's age, the intra-cardiac anatomy, the size of both the right superior vena cava and the pulmonary artery, the pulmonary vascular resistance and the site of drainage in the left atrium.

Conclusion

LSVC into LA can be corrected with different intra- and extra-cardiac approaches. Strategy should be tailored to each patient, keeping into account anatomical and physiological factors.

Learning objectives

- 1) Recognizing an unusual combination of ostium secundum, coronary veins and LSVC draining into the left atrium, with indication for surgical correction, in a patient who presented with atrial fibrillation and desaturation.
- 2) Knowing how to correct an anomalously connected LSVC.

References

- [1] De Leval MR, Ritter DG, McGoon DC, Danielson GK. (1975) Anomalous systemic venous connection surgical considerations. *Mayo Clin Proc* 50:599–610.
- [2] Rastelli GC, Ongley PA, Kirklin JW. (1965) Surgical correction of common atrium with anomalously connected persistent left superior vena cava: report of case. *Mayo Clin Proc* 40: 528–32.

- [3] Zimand S, Benjamin P, Frand M, Mishaly D, Smolinsky AK, Hegesh J. (1999) Left Superior Vena Cava to the Left Atrium: Do We Have to Change the Traditional Approach? *Ann Thorac Surg* 68:1869±72
- [4] Shumacker H, King H, Waldhausen J. (1967) The persistent left superior vena cava. Surgical implications, with special reference to caval drainage into the left atrium. *Ann Surg* 165:797– 805.
- [5] Kabbani SS, Feldman M, Angelini P, Leachman RD, Cooley DA. (1973) Single left superior vena cava draining into the left atrium: surgical repair. *Ann Thorac Surg* 16:518–25.
- [6] Komai H, Naito Y, Fujiwara K. (1996) Operative technique for persistent left superior vena cava draining into the left atrium. *Ann Thorac Surg* 62:1188–90.
- [7] Sugimoto K, Matsuo K, Ohba M. (2012) Intra-Atrial Rerouting and Maze Procedure for an Adult Patient in Cor Triatriatum, Persistent Left Superior Vena Cava, and Atrial Fibrillation *Ann Thorac Surg* 93:2056–8
- [8] Reddy VM, McElhinney DB, Hanley FL. (1997) Correction of left superior vena cava draining to the left atrium using extra- cardiac techniques. *Ann Thorac Surg* 63:1800–2.
- [9] Takach TJ, Cortelli M, Lonquist JL, Cooley DA. (1997) Correction of anomalous systemic venous drainage: transposition of left SVC to left PA. *Ann Thorac Surg* 63:228–30.

Figure Legend

Fig.1: 3D CT scan reconstruction showing the persistent left superior vena cava (LSVC) draining into the left atrium in close proximity to the left atrial appendage.

Fig. 2: 3D CT scan reconstruction showing the separate drainage of the coronary veins into the left atrium.

Fig. 3: Scheme of anatomy before (left) and after (right) surgical correction. §: persistent LSVC, *: ostium of the persistent LSVC; Blue arrowhead: *ostium secundum* atrial septal defect (ASD) with left-to-right shunt; Black arrowhead: obliterated ostium of the LSVC; Blue arrow: ASD closed with a patch of pericardium; #: 16-mm prosthetic graft interposed between the LSVC and the right atrium.



