CASE REPORT

Isolated left-sided partial anomalous pulmonary venous connection in a child

Bir çocukta tek başına sol-tarafta kısmi anormal pulmoner venoz bağlantı

İsmihan Selen Onan, M.D.,[1] Onur Sen, M.D.,[1] Selman Gökalp, M.D.,[2] Burak Onan, M.D.[1]

[1]Department of Cardiovascular Surgery, İstanbul Mehmet Akif Arsoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey

[2]Department of Pediatric Cardiology, İstanbul Mehmet Akif Arsoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey

Summary– Isolated left-sided partial anomalous pulmonary venous connection with intact interatrial septum is a rare diagnosis in childhood. In these cases, a vertical vein drains the left upper pulmonary lobe into the brachiocephalic vein and finally to the right atrium. Surgical treatment is performed to prevent right ventricular failure and pulmonary artery disease in advanced age. In this report, the rare entity of isolated left-sided anomalous pulmonary venous connection in a 14-year-old girl and successful minimally invasive surgery without cardiopulmonary bypass are described.

Özet– Tek başına sol tarafta kısmi anormal pulmoner venöz bağlantı ile sağlam interatriyal septum birlikteliği çocukluk çağında nadir bir durumdur. Bu olgularda, bir vertikal ven sol üst pulmoner venöz sistemi brakiyosefalik vene ve daha sonrada sağ atriyuma boşalır. Cerrahi tedavi ileri yaşlarda sağ ventrikül yetersizliği ve pulmoner arter hastalığını engellemek amacıyla uygulanır. Bu yazıda, 14 yaşındaki kız hastada nadir bir durum olan tek başına sol taraf kısmi anormal pulmoner venöz bağlantının, kardiyopulmoner baypas olmaksızın minimal invaziv olarak yapılan başarılı cerrahi tedavisi sunuldu.

Partial pulmonary venous connection (PAPVC) is a congenital anomaly present in 0.4% to 0.7% of postmortem examinations.[1–4] The incidence of left-sided PAPVC is only between 10% and 18.2% of all reported cases of PAPVC and most patients present with an associated atrial septal defect.[1] It is an uncommon congenital abnormality in which some, but not all, of the pulmonary veins connect to the right atrium or to one of its venous tributaries. Clinically, these cases are mostly detected in adulthood, and the diagnosis of left-sided PAPVC is an unusual event in childhood. The major morbidities are right ventricular failure and pulmonary artery disease as patients grow and, therefore, surgical treatment is indicated.

Presently described is the rare entity of isolated left-sided PAPVC in a child, characterized by drainage of the left upper pulmonary vein into the left brachiocephalic vein through a vertical vein with intact atrial septum. A discussion of surgical treatment without cardiopulmonary bypass is also provided.

Abbreviation:

PAPVC Partial pulmonary venous connection

CASE REPORT

A 14-year-old female presented with dyspnea on exertion ongoing for the last year. The patient had no history of heart failure or recurrent respiratory tract infections. Her family history was not significant from a cardiac standpoint. On admission, vital signs were normal. Physical examination revealed no abnormality of cardiopulmonary system. Biochemical test results were normal. Electrocardiogram revealed the presence of right axis deviation. A chest X-ray showed a normal cardiothoracic ratio with a slight increase in pulmonary vascularity. Transthoracic echocardiogram revealed dilated right-sided cardiac chambers with

Received: December 08, 2016 *Accepted:* February 07, 2017

Correspondence: Dr. İsmihan Selen Onan. İstanbul Mehmet Akif Ersoy Göğüs Kalp ve Damar Cerrahisi, Egitim ve Araştırma Hastanesi, Kalp ve Damar Cerrahisi Kliniği, 34303 İstanbul, Turkey.
Tel: +90 212 - 692 20 00 / 1129 e-mail: selenibis@hotmail.com

good tricuspid valve coaptation and left ventricular fractional shortening of 32%. Left upper pulmonary vein was connected to the left brachiocephalic vein via a vertical vein. Interatrial septum was intact, and no other intracardiac defect was observed. Cardiac catheterization revealed single drainage of the left upper pulmonary lobe into the brachiocephalic vein through the vertical vein of about 13 mm in diameter (Figure 1; Video 1*). Angiogram also excluded dual-drainage of pulmonary veins to the left atrium simultaneously with vertical vein. The pulmonary to systemic blood flow (Qp/Qs) ratio was calculated as 1.6. Contrast-enhanced computed tomography of the chest confirmed the pathology and illustrated anatomical details, such as the proximity of the left atrium to the vertical vein. After providing informed consent, the patient underwent minimally invasive surgery for repair of PAPVC.

The left chest was entered through a small posterolateral thoracotomy via the left fourth intercostal space. The left vertical vein was dissected and mobilized from the hilum to the innominate vein (Figure 2, upper view). The pericardium was opened posterior to the left phrenic nerve, and a large window was created. After 5000 units of heparin were administered, a side clamp was placed below the insertion of the vertical vein into the innominate vein, and a small Yaşargil clamp was placed across the proximal portion. The vertical vein was transected and the incision was repaired using 5/0 Prolene suture (Ethicon, Inc., Somerville, NJ, USA). A side clamp was placed across the base of the left atrial appendage. Left atrial appendage was amputated and its trabeculae were trimmed. Anastomosis between the vertical vein and the left atrial appendage was performed using 6/0 polydioxanone suture in a running fashion (Figure 2, lower view). The clamp on the vertical vein was removed first for de-airing of the vein. The left lung was then expanded. The pericardium was partially closed. One chest tube was placed in the left hemithorax and the patient was extubated 1 hour after the operation.

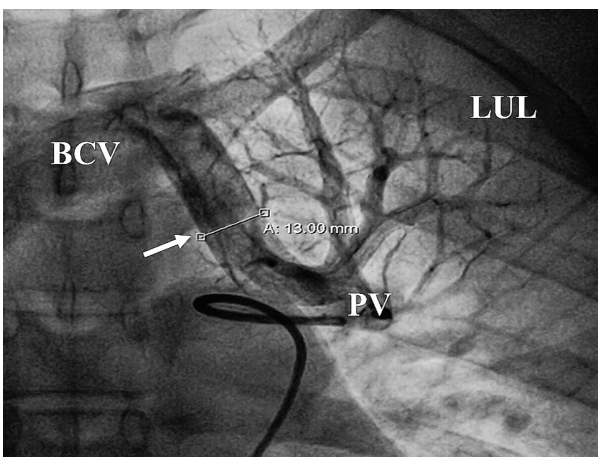

Figure 1. Cardiac catheterization shows the drainage of the pulmonary vein (PV) of the left upper lobe (LUL) into the brachiocephalic vein (BCV) through a vertical vein (arrow).

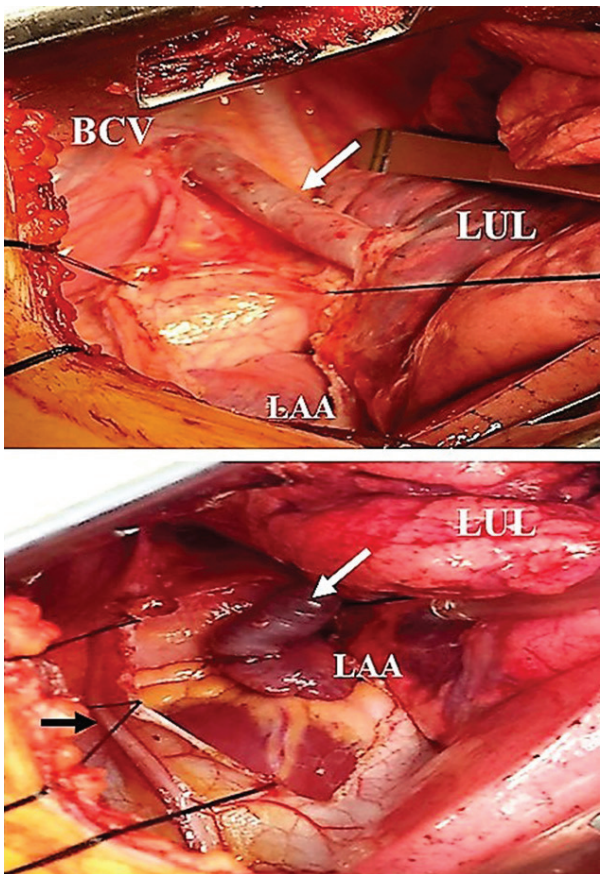

Figure 2. Upper view: Operative view shows the vertical vein (white arrow), which is draining the left upper lobe (LUL) into the brachiocephalic vein (BCV). Lower view: Vertical vein is transected from its connection to the brachiocephalic vein and anastomosed to the left atrial appendage (LAA). It is placed posteriorly to the phrenic nerve (black arrow) through a pericardial window.

Postoperative course was uneventful. The patient was discharged from the hospital on the fourth day after the operation. Transthoracic echocardiography examination identified no anastomotic pulmonary vein stenosis postoperatively. The patient was clinically in good condition on follow-up at 6 months after operation without any abnormality on echocardiography.

DISCUSSION

PAPVC occurs when some of the pulmonary veins connect to the right atrium or one of its venous tributaries.[1–4] Most anomalous pulmonary veins originate from the right lung and are connected to the superior vena cava or, less commonly, to the right atrium or inferior vena cava. In left-sided PAVC, most often a vertical vein drains the left upper lobe into the brachiocephalic vein and then to the right atrium. The diagnosis of isolated left PAPVC with intact interatrial septum is rare in children.[1–4] Alsoufi et al. reported on a series of 306 children who underwent surgery for PAPVC. In this series, only 17 (7%) patients had isolated left-sided PAPVCs and 5 (2%) had bilateral drainage. In a review in 2015, Naimo et al. reported that only 15 children underwent surgical repair for left-sided PAPVC.[3] Our search revealed no other pediatric case to date since that report. Although acceptable short and long-term outcomes of surgical correction in adults have been reported,[1] there are still limited data on the outcomes of surgical repair of left-sided PAPVC in children.[2–4]

PAPVC may remain undetected until adulthood, as it often presents with few clinical symptoms. Although most pediatric patients are asymptomatic, natural history may include progressive right ventricular failure and pulmonary disease, if a large left-to-right shunt occurs, especially in patients with septal defects. In more than 75% of reported cases, an atrial septal defect is diagnosed concomitantly and this association increases the possibility of right ventricular failure.[1–4] Therefore, the diagnosis of left-sided PAPVC and concomitant intracardiac defects in children is important to prevent cardiopulmonary morbidities at an earlier age. On diagnosis, transthoracic echocardiography is used as an initial modality to detect cardiovascular pathologies, but it may not be effective to define the detailed anatomy and course of pulmonary veins in PAPVC due to technical limitations. Transesophageal echocardiography can be used effectively in experienced hands to detect abnormalities of pulmonary venous return. Computed angiography and magnetic resonance imaging can be used effectively to detect PAPVC. Moreover, cardiac catheterization can be an alternative modality to reveal the detailed anatomy of pulmonary venous return. Its major advantage is for calculation of the Qp/Qs ratio and exclusion of dual drainage of pulmonary veins.

The aim of the surgical repair is to direct or reroute the blood flow from the left upper lobe to the left atrium. Indications for surgery in asymptomatic children with left-sided PAPVC are similar to atrial septal defect repair. Surgery is recommended if left-to-right shunt ratio (Qp/Qs ratio) is more than 1.5.[3,4] ElBardissi et al. (1) recommended that surgical correction for isolated left PAPVC can be performed when patients are asymptomatic or there is evidence of right-sided overload. Because the reported outcomes of surgery are excellent, the authors suggest early repair to prevent right ventricular failure and pulmonary disease.[1–4] Technically, repair of isolated left-sided PAPVC can be achieved without cardiopulmonary bypass using a simple thoracotomy with excellent results. The advantages of thoracotomy approach are better cosmetic results, early return to daily life, and the avoidance of cardiopulmonary bypass and sternotomy-related complications. In our patient, we used a thoracotomy incision successfully. The vertical vein was divided from its junction with the brachiocephalic vein and then anastomosed to the left atrial appendage. Postoperative course of the patient was uneventful and she was discharged five days after the operation.

Technically, the off-pump surgical technique can be a beneficial approach in the surgical repair of an isolated anomalous venous connection of the left lung. Before clamping and division of the vertical vein, systemic heparinization is performed with a bolus intravenous dose of 100 mg/kg to prevent thromboembolization. In cases with total anomalous drainage of the left lung, the left pulmonary artery should be clamped before division of the vertical vein, which may cause hemodynamic instability during the procedure. Therefore, the use of cardiopulmonary bypass is recommended in such cases. But, in partial anomalous drainage of the left lung, the hemodynamics can be better tolerated during clamping, and off-pump procedures can be performed uneventfully. In our case, the left pulmonary artery was not clamped and vital signs were stable during the procedure. Anesthetic management is also important to preserve lung functions and the levels of blood gases at normal levels. Clamping of the left pulmonary artery or vertical vein diminishes the actual volume of pulmonary perfusion. Thus, the use of hyperventilation, effective hydration, sedation, and pulmonary vasodilators can be beneficial during the operation. Moreover, the course of the

vertical vein and 3-dimensional geometry of the final anastomosis are important in the prognosis of the patients. A kink in the vertical vein or stenosis of the anastomosis may lead to pulmonary venous obstruction and may cause adverse events after surgery.

In cases with concomitant atrial septal defects, a consensus should be found between pediatric cardiology and cardiovascular surgery clinics. Defects can be repaired using a hybrid approach with percutaneous closure of the septal defect and then surgery for left-sided PAPVC, a single-staged repair with sternotomy using cardiopulmonary bypass, or staged surgical repair of both defects.

In conclusion, a left-sided PAPVC is an unusual diagnosis in childhood. Surgical repair using thoracotomy incision is a feasible and safe approach to prevent right ventricular failure and pulmonary complications in the future.

Conflict-of-interest: None declared.

****Supplementary video file associated with this article can be found in the online version of the journal.***

REFERENCES

1. El Bardissi AW, Dearani JA, Suri RM, Danielson GK. Left-sided partial anomalous pulmonary venous connections. Ann Thorac Surg 2008;85:1007–14. [CrossRef]
2. Kumar T, Patra S, Ramalingam R, Agrawal N, Agarwal A, Manjunath CN. Pulmonary hypertension due to presence of isolated partial anomalous pulmonary venous connection: A case report. J Cardiovasc Dis Res 2013;4:239–41.
3. Naimo PS, d'Udekem Y, Brizard CP, Konstantinov IE. Outcomes of repair of left partial anomalous pulmonary venous connection in children. Interact Cardiovasc Thorac Surg 2015;21:254–6. [CrossRef]
4. Alsoufi B, Cai S, Van Arsdell GS, Williams WG, Caldarone CA, Coles JG. Outcomes after surgical treatment of children with partial anomalous pulmonary venous connection. Ann Thorac Surg 2007;84:2020–6. [CrossRef]

Keywords: Childhood; partial anomalous pulmonary venous connection; surgery.

Anahtar sözcükler: Çocukluk çağı; parsiyel anormal pulmoner venöz dönüş; cerrahi.