



Partial anomalous pulmonary venous connection with intact atrial septum in a child with ventricular septal defect: a case report

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Partial anomalous pulmonary vein connection (PAPVC) is a rare congenital abnormal cardiac defect involving the pulmonary veins draining into the right atrium (RA) directly or indirectly by venous connection. Ninety percent of PAPVCs are accompanied by atrial septal defect (ASD). To our knowledge, there is no previous report of PAPVC with ventricular septal defect (VSD) without ASD in Korea, and in this paper, we report the first such case. A 2-day-old girl was admitted into the Chonnam National University Hospital for evaluation of a cardiac murmur. An echocardiogram revealed perimembranous VSD without ASD. She underwent patch closure of the VSD at 5 months of age. Although the VSD was completely closed, she had persistent cardiomegaly with right ventricular volume overload, as revealed by echocardiography. Three years later, cardiac catheterization and chest computed tomography revealed a PAPVC, with the right upper pulmonary vein draining into the right SVC. Therefore, correction of the PAPVC was surgically performed at 3 years of age. We conclude that it is important to suspect PAPVC in patients with right ventricular volume overload, but without ASD.

Key words: Partial anomalous pulmonary venous connection, Ventricular heart septal defect, Atrial heart septal defect

Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital abnormal cardiac defect in which the pulmonary veins drain into the right atrium (RA) directly, or indirectly by venous connection¹⁾. Drainage of the right pulmonary vein into the RA or the superior vena cava (SVC) is the most common type of PAPVC. Since ninety percent of PAPVC are accompanied with

atrial septal defect (ASD)²⁾, the echocardiologist should try to find the PAPVC whenever ASD is detected. On the other hand, PAPVC with ventricular septal defect (VSD) is rarely reported³⁾, therefore, when the VSD is initially detected, the echocardiogram may fail to find the PAPVC. We report the first case of a baby who had VSD accompanied by PAPVC, with intact atrial septum in Korea.

Case report

A 2 day-old girl was examined at the Chonnam National University Hospital for the evaluation of a cardiac murmur. She was born by Caesarean section at the local hospital, weighted 3.4 kg at birth, and was suspected to have VSD on antenatal ultrasonography. On physical examination, there were no significant findings apart from a grade 1 systolic murmur on the left lower sternal boarder. Transthoracic echocardiography revealed perimembranous VSD (Fig. 1A-C),

and persistent left superior vena cava draining into the RA via the coronary sinus, without bridging the left innominate vein.

At 5 months old, she was admitted for the symptoms of cough, cold sweating, and tachypnea. On admission day, blood pressure was 110/70 mmHg, body temperature was 36°C, pulse rate was 120 beats per minute and respiration rate was 50 per minute. The chest X-ray showed cardiomegaly with pulmonary congestion. Electrocardiography revealed RA enlargement, and left ventricular hypertrophy. Follow-up echocardiography showed large perimem-

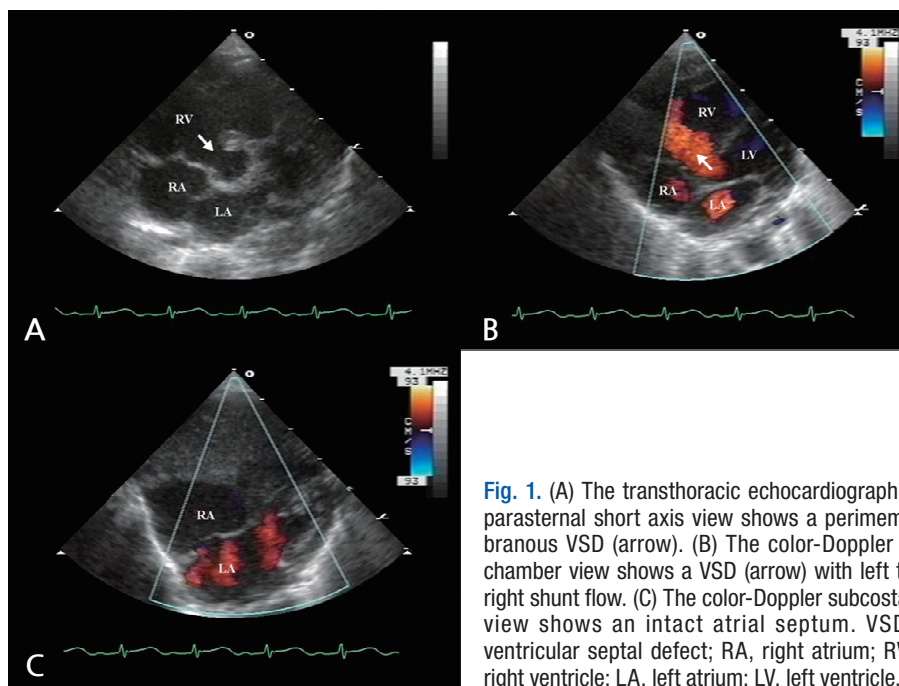


Fig. 1. (A) The transthoracic echocardiographic parasternal short axis view shows a perimembranous VSD (arrow). (B) The color-Doppler 4 chamber view shows a VSD (arrow) with left to right shunt flow. (C) The color-Doppler subcostal view shows an intact atrial septum. VSD, ventricular septal defect; RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle.

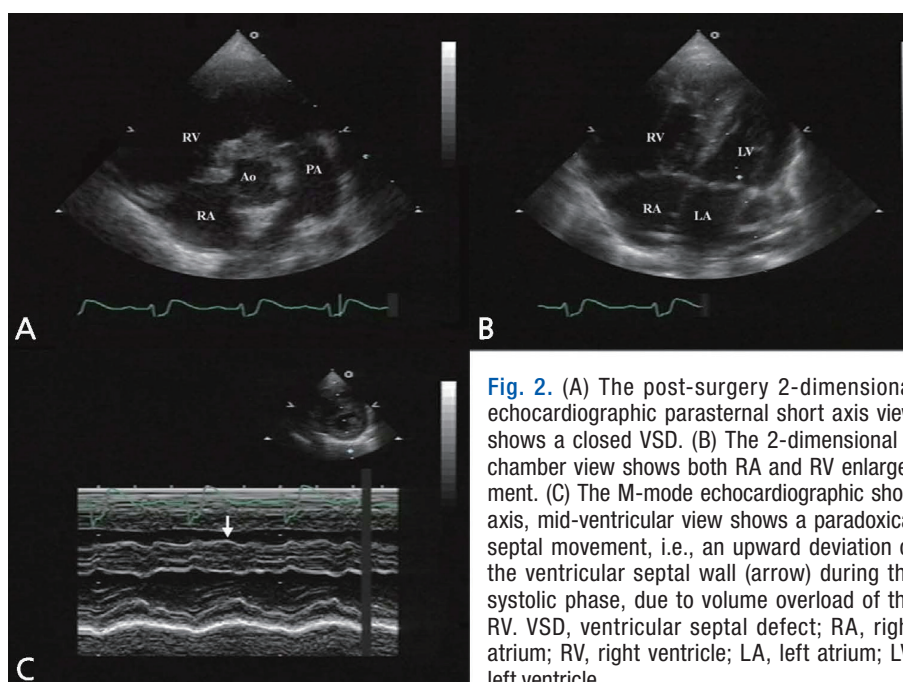


Fig. 2. (A) The post-surgery 2-dimensional echocardiographic parasternal short axis view shows a closed VSD. (B) The 2-dimensional 4 chamber view shows both RA and RV enlargement. (C) The M-mode echocardiographic short axis, mid-ventricular view shows a paradoxical septal movement, i.e., an upward deviation of the ventricular septal wall (arrow) during the systolic phase, due to volume overload of the RV. VSD, ventricular septal defect; RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle.

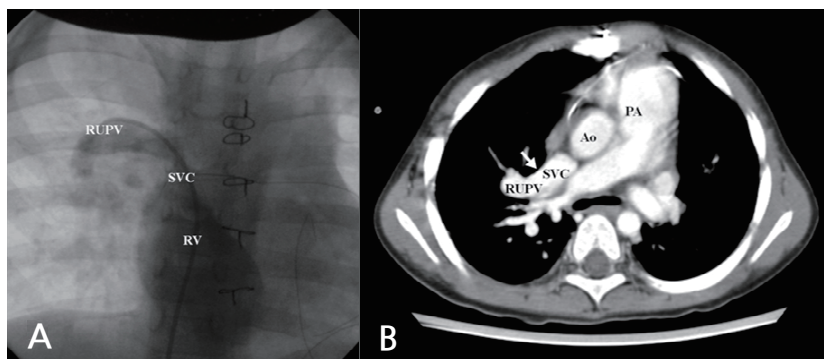


Fig. 3. (A) Angiography of the RUPV shows a partial anomalous pulmonary venous connection, consisting of the RUPV, which drains into the RA through the SVC. (B) Chest computer tomography shows the RUPV (arrow), which drains into the SVC. RUPV, right upper pulmonary vein; RA, right atrium; SVC, superior vena cava; Ao, Aorta; PA, pulmonary artery.

branous VSD, pulmonary hypertension, and moderate tricuspid regurgitation (TR). However, we could not find either ASD or patent foramen ovale. She had conservative treatment for the symptoms of congestive heart failure, including diuretics and inotropics for 2 weeks, and then she underwent a surgical patch closure of the perimembranous VSD. After the operation, despite of complete closure of the VSD (Fig. 2A), she had suffered from recurrent cough, fever, and pneumonia for 3 years. The chest X-ray showed cardiomegaly and increased pulmonary vascular markings. In addition, the electrocardiogram showed right ventricular hypertrophy, right atrial enlargement, and right axis deviation. The follow-up echocardiography showed both right atrial (RA) and right ventricular (RV) enlargement and right ventricular volume overload (Fig. 2B, C). Then, we performed cardiac catheterization and chest computed tomography at 3 years and 2-months of age. It revealed the right upper pulmonary vein draining into the right SVC, without any evidence of ASD (Fig. 3A, B). At 3 years and 3 months of age, she underwent the modified Warden technique for correction of the PAPVC consisting of the right pulmonary vein connected to the left pulmonary vein. After PAPVC correction, the echocardiography showed disappearance of the right ventricular volume overload (Fig. 4). The follow-up chest X-ray showed improvement of the pulmonary congestion. Since then, she has visited our outpatient department regularly, and her growth and development is according to her age.

Discussion

In the normal heart, there are four pulmonary veins connected to the left atrium which are the upper left, upper right, lower left and lower right pulmonary veins⁴. Thus, in patients diagnosed with PAPVC, the blood flow from the pulmonary veins returns to the RA instead of the left atrium. The majority of PAPVC has one anomalous pulmonary vein and more than one anomalous vein are

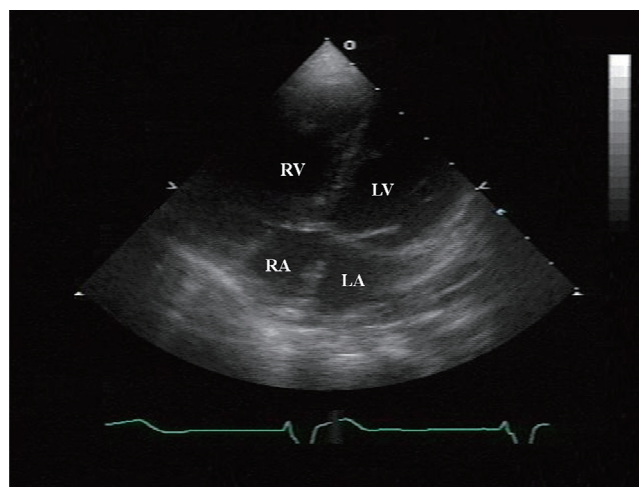


Fig. 4. After partial anomalous pulmonary venous connection correction, the 2-dimensional echocardiographic 4 chamber view shows the improvement of both RA and RV enlargement. RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle.

also rarely reported. The diagnosis can be missed easily, in up to 25 % of patients, the accurate determination of either the number or sites of anomalous connecting pulmonary veins is problematic or impossible⁵. Therefore, further knowledge of the variation patterns of the pulmonary venous drainage is necessary in order to diagnose PAPVC.

PAPVC is discovered by 0.4 to 0.7% of cases in autopsy series^{2,6}, but this incidence seemed an overestimate because many of these cases were asymptomatic. Thus the true incidence of symptomatic patients is expected to be lower. Moreover, at least one anomalous pulmonary venous connection is present in about three-quarters of the patients diagnosed with asplenia syndrome⁷. Drainage of the right pulmonary vein into either the RA or the SVC is a frequent type². Similarly, in the case presented here, the right upper pulmonary vein drained into the right SVC. Up to 90% of the PAPVC cases are accompanied by ASD². PAPVC from the right pulmonary veins occurs ten times more frequently than the connection initiated from the left pulmonary veins⁸. Additionally, PAPVC from left pulmonary vein with intact

atrial septum is extremely rare⁸).

Children with PAPVC usually remain asymptomatic and are referred to specialist based on an incidentally noted cardiac murmur⁵. Symptoms may also occur in older patients and may be secondary to right-sided volume overload or pulmonary vascular obstructive disease. The appearance of the clinical symptoms from PAPVC depends on how many pulmonary veins abnormally return to the RA, and whether the right to left shunt occurs or not^{9,10}. Cardiac arrhythmia detected on a routine clinic visit, an occasionally detected cardiac murmur as well as a widely split second heart sound, can be the first signs of PAPVC⁵. Some patients complain of tachypnea, subcostal retraction⁶, repeated pulmonary infections and failure to thrive^{11,12} or limitation of physical exercise¹². On physical examination, marked stridor can be auscultated¹¹. Additionally, in serious cases, fully developed pulmonary edema, hypoxemia, cyanosis and growth failure may appear⁷. Our case had recurrent pulmonary infection, i.e., pneumonia, due to the right heart volume overload condition.

Imaging examinations to diagnose PAPVC include chest radiography, echocardiography, cardiac magnetic resonance imaging (MRI), cardiac computed tomography, and/or cardiac catheterization. Chest radiographic findings are, as follows: cardiomegaly with prominent RA and right ventricle, dilated pulmonary vessels and pulmonary infiltrated shadow^{6,12}. Echocardiography with color flow mapping reveals pulmonary veins connected either to the RA or the SVC, right heart volume overload, turbulent flow in the SVC, and sometimes TR^{5,12}. Structural MRI is rapidly becoming the procedure of choice for further investigation of the PAPVC^{5,13}.

Even in when echocardiography findings suggest the presence of PAPVC, not all the pulmonary veins may be identified. Therefore cardiac catheterization may be necessary for both the precise anatomic diagnosis and the hemodynamic evaluations. In the clinical case presented here, we confirmed the PAPVC by cardiac catheterization.

The medical treatment of PAPVC is not a routine choice for asymptomatic patients. Patients with heart failure can be managed with diuretics, cardiac glycosides, afterload reduction, and beta blockers. Arrhythmias should be appropriately treated⁸. Percutaneous transcatheter occlusion of the anomalous pulmonary venous connection using coil was also reported¹⁴. The definitive treatment for PAPVC is the surgical repair^{12,14}. The optimal time for intervention is the preschool age¹². The operative technique depends on the number and the site of the anomalous vein or veins. Among several techniques, the Warden method or modified Warden method are usually used¹⁵.

Although PAPVC can be isolated, most PAPVC is accompanied with ASD². Very rarely, PAPVC can be accompanied with VSD, in addition to ASD. Iwasa et al.⁶ reported a case in which the PAPVC was accompanied by VSD(16), ASD, and pulmonary venous stenosis.

Kim et al.³ also reported that PAPVC was accompanied by VSD, ASD, and by coarctation of the aorta. Oh et al.¹⁷ reported an isolated case of PAPVC, without either ASD or VSD. To the best of our knowledge, the case presented here, i.e., of a PAPVC with VSD but without ASD, is the first case in Korea. When the VSD was initially detected, the PAPVC diagnosis was missed.

In conclusion, we believe that the PAPVC diagnosis should be considered every time a patient presents with right ventricular volume overload without ASD.

References

1. Elami A, Rein AJ, Preminger TJ, Milgater E. Tetralogy of Fallot, absent pulmonary valve, partial anomalous pulmonary venous return and coarctation of the aorta. *Int J Cardiol* 1995;52:203-6.
2. Broy C, Bennett S. Partial anomalous pulmonary venous return. *Mil Med* 2008;173:523-4.
3. Kim S, Lee YS, Woo JS, Cho KJ. Surgical management of coarctation of the aorta with a ventricular septal defect and coexisting partial anomalous pulmonary venous connection: a case report. *Korean J Thorac Cardiovasc Surg* 2006;39:479-81.
4. Respondek-Liberska M, Janiak K, Moll J, Ostrowska K, Czichos E. Prenatal diagnosis of partial anomalous pulmonary venous connection by detection of dilatation of superior vena cava in hypoplastic left heart. A case report. *Fetal Diagn Ther* 2002;17:298-301.
5. Lilje C, Weiss F, Weil J. Detection of partial anomalous pulmonary venous connection by magnetic resonance imaging. *Pediatr Cardiol* 2005; 26:490-1.
6. Iwasa T, Mitani Y, Sawada H, Takabayashi S, Shimpo H, Matsubayashi N, et al. Persistent lung shadow in an infant with ventricular septal defect and partial anomalous pulmonary venous connection associated with pulmonary venous obstruction. *Pediatr Int* 2008;50:397-9.
7. Coulson JD, Bullaboy CA. Concentric placement of stents to relieve an obstructed anomalous pulmonary venous connection. *Cathet Cardiovasc Diagn* 1997;42:201-4.
8. Takamori S, Hayashi A, Nagamatsu Y, Tayama K, Kakegawa T. Left partial anomalous pulmonary venous connection found during a lobectomy for lung cancer: report of a case. *Surg Today* 1995;25:982-3.
9. Bernstein D. Partial anomalous pulmonary venous return. In: Kliegman RM, Behrman RE, Jenson HB, Stanton BF, editors. *Nelson textbook of pediatrics*. 18th ed. Philadelphia: Saunders, 2007;1886.
10. Valsangiacomo ER, Hornberger LK, Barrea C, Smallhorn JF, Yoo SJ. Partial and total anomalous pulmonary venous connection in the fetus: two-dimensional and Doppler echocardiographic findings. *Ultrasound Obstet Gynecol* 2003;22:257-63.
11. Tueche SG, Demanet H, Goldstein JP, Dessy H, Viart P, Deuvaert FE. Association of a Cor Triatriatum Sinister and a right partial anomalous pulmonary venous return. A case report. *Acta Chir Belg* 2005;105:217-8.
12. Hovels-Gurich H. Pulmonary venous return anomaly. *Orphanet encyclopedia* [Internet]. May 2003 [cited 2011 Aug 1]. Available from: <http://www.orpha.net/data/patho/GB/uk-PVRA.pdf>.
13. Hazirolan T, Ozkan E, Haliloglu M, Celiker A, Balkanci F. Complex venous anomalies: magnetic resonance imaging findings in a 5-year-old boy. *Surg*

- Radiol Anat 2006;28:534-8.
14. Lee MG, Ko JS, Yoon HJ, Kim KH, Ahn Y, Jeong MH, et al. An unusual presentation of an atrial septal defect. *J Cardiovasc Ultrasound* 2009;17:151-2.
 15. Forbess LW, O'Laughlin MP, Harrison JK. Partially anomalous pulmonary venous connection: demonstration of dual drainage allowing non-surgical correction. *Cathet Cardiovasc Diagn* 1998;44:330-5.
 16. Nakahira A, Yagihara T, Kagisaki K, Hagino I, Ishizaka T, Koh M, et al. Partial anomalous pulmonary venous connection to the superior vena cava. *Ann Thorac Surg* 2006;82:978-82.
 17. Oh IY, Chang SA, Kim SH, Shin JI, Sir JJ, Hong YS, et al. Partial anomalous pulmonary venous return into coronary sinus with intact atrial septum. *J Korean Soc Echocardiogr* 2004;12:94-6.