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Case Report

A rare case of Abmormal Connection of Right Inferior Pulmonary Vein to Right atrium with Intact Intera trial Septum

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Abstract: Isolated Partial Anomalous Pulmonary Venous Connection is a rare congenital defect that may not clinically manifest until adulthood. It can be misdiagnosed as a secundum type atrial septal defect by an echo cardiographer too. We present a 9 year-old girl who presented to us with complaints of occasional exertional fatigue. An ejection systolic murmur was heard in the 2rd inter-costal space at left parasternal area. The cardiothoracic ratio was normal in routine chest X-ray with an anomalous vessel adjacent to the left pulmonary hilum. Echocardiography revealed an ostium secondum type of atrial septal defect of 18x10 mm in size with absence of inferior limb. She was taken up for surgery for closure of atrial septal defect. Intraoperatively, it was found that the atrial septum was intact but the right inferior pulmonary vein was draining to the right atrium. Surgically the defect was corrected by resecting part of atrial septum and tunneling the right inferior pulmonary vein to left atrium with a Dacron patch. The postoperative outcome was uneventful.
Keywords: Atrial Septal Defects, pulmonary vein, Echocardiography

INTRODUCTION

Partial anomalous pulmonary venous connection (PAPVC) is an uncommon congenital anomaly and it is extremely rare in the absence of atrial septal defect (ASD) [1]. Embryo logically, the persistence of the venous drainage of the lung into the systemic cardinal and umbilical vitelline systems results in to a variety of pulmonary venous returns anomalies [2]. PAPVC is found in around 0.4–0.7% of autopsies [3]. Anomalous pulmonary venous connection occurs in approximately 10–15% of ostium secundum ASD and 85 % of patients with sinus venosus ASD [3].

THE CASE

A 9 year-old girl presented to us with complaints of occasional exertional dyspnoes. Her vital parameters were normal at routine physical examination except an ejection systolic murmur was heard in the 2rd inter-costal space at left parasternal area. A routine chest X-ray showed a normal cardiac silhouette with mild loss of pulmonary bay (Figure 1). Thansthoracic echocardiography was reported as an osteum secondum type of atrial septal defect of 18x10 mm in size with absence of inferior limb. She was taken up for surgery for closure of atrial septal defect. Aortobicaval cannulation was performed. Cardiopulmonary by-pass was established. Aortic cross clamp applied followed by aortic root plegia was started and heart got arrested immediately. Right atrium was opened and it was found that the atrial septum was completely intact but the right inferior pulmonary vein (RIPV) was draining to the inferior aspect of right atrium near opening of the inferior venacava (Figure 2). Surgically the defect was corrected by resecting a part of secondum atrial septum and rerouting the RIPV to left atrium via a piece of Dacron patch. The patch was put in such a way that the RIPV's neoopening to left atria, coronary sinus opening and inferior venacava opening is not compromised. Right atrial opening was closed in two layers and came out of cardiopulmonary bypass in normal sinus rhythm.

The postoperative outcome was uneventful. Postoperative echocardiogram showed no turbulence at RIPV neoopening to left atria and inferior venacava opening at right atrium.

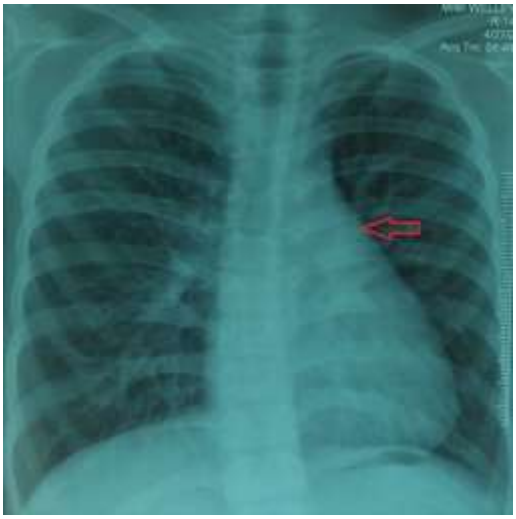

Fig 1: Posterior anterior view of chest x-ray showing normal looking lung fields and loss of pulmonary bay (Red arrow) of cardiac silhouette

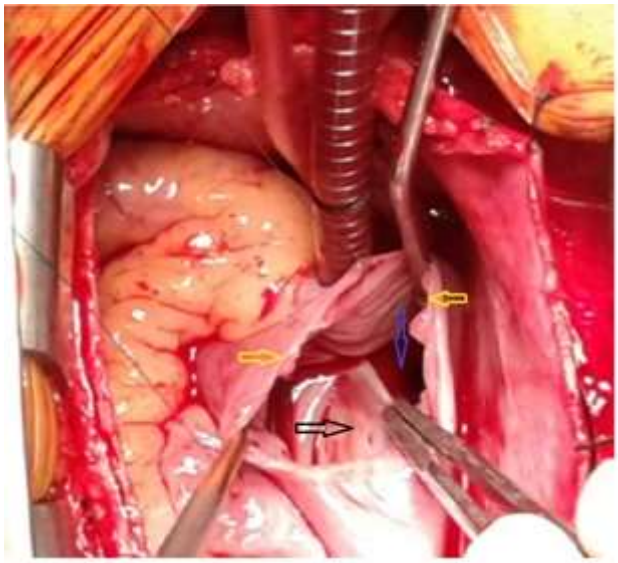

Fig 2: Intraoperative picture shows surgically slit open right atrial margins (Yellow arrow), intact interatrial septum (Black arrow) and large opening (Blue arrow) of RIPV in right atrium

DISCUSSION

The physiological change of PAPVC is similar to that of ASDs and the symptoms depend on the amount of shunt of blood into the right side of heart [4]. Normally each pulmonary vein contributes an average 25% of the total pulmonary blood flow. However, in anomalous pulmonary vein return, the shunt flow can be higher since the circulation is preferentially directed to the right side due to lower pressure in the right atrium and superior vena cava than in the left atrium. This effect becomes more pronounced in conditions that tend to increase left atrial pressure such as systemic hypertension or left heart disease. So some patients with PAPVC eventually develop right-sided volume overload that leads right heart failure and pulmonary arterial hypertension lately [5]. Children with PAPVC usually remain asymptomatic and are referred to specialist based on an incidentally noted cardiac murmur. Dyspnoea and atrial arrhythmias are the usual symptoms for increased pulmonary venous flow though rarely may present as haemoptysis due to bronchitis or the development of pulmonary vascular disease [1, 4].

PAPVC is usually diagnosed by transthoracic echocardiography, Tran esophageal echocardiography, or catheter study. Agitated saline is used to aid the diagnosis by echocardiography but a PAPVC may get overlooked. A PAPVC may be mistakenly diagnosed as

ASDs if the observer is not conscious about the fact that a PAPVC may be present with an intact atrial septum, and in these cases the intervention is not required if the patient is not symptomatic [3].

There are a number of operative procedures for the correction of right inferior PAPVC is described. When the atrial septum is intact, repair can often be accomplished by making a longitudinal incision in it next to the atrial wall posteriorly and resuturing it to the lateral right atrial wall in front of the right pulmonary vein orifices [6]. Alternatively, and particularly when geometry in the right atrium does not lend itself to this simple repair, the fossa ovalis and posterior limbic tissue may be excised and a pericardial, PTFE or knitted polyester patch used for baffle reconstruction. In our case the fossa ovalis and posterior limbic tissue was excised and a Dacron patch was used to baffle the abnormal connection. The postoperative complications include kinking, stenosis of the corrected pulmonary vein, obstruction, and arrhythmias [7].

CONCLUSION

An isolated PAPVC is a rare congenital anomaly. It may be an incidental finding or can present clinically with features of increased right to left shunt at atrial level. An isolated asymptomatic PAPVC does not require any intervention. An echo cardiographer should always be careful at diagnosis of an atrial septal defect and must always rule out the presence of an isolated PAPVC.

ABBREVIATIONS

- PAPVC - Partial Anomalous Pulmonary Venous Connection
- ASD - Atrial Septal Defect
- RIPV - Right Inferior Pulmonary Vein

REFERENCES

1. Masiello P, Panza A, Morena E, Marotta A, Bellieni G, Di Benedetto G; Total anomalous left pulmonary venous connection with intact atrial septum: surgical treatment of a rare case. Eur J Cardiothorac Surg. 1995; 9(2):102–3.
2. Edwards JE; Pathologic and developmental considerations in anomalous pulmonary venous connection. Proc Staff Meet Mayo Clin. 1953; 28:441.
3. Sandeep Sahay, Richard A. Krasuski, Adriano R; Tonelli. Partial Anomalous Pulmonary Venous Connection and Pulmonary Arterial Hypertension. Respirology. 2012; 17(6): 957–963.
4. Basalus M.W.Z, Said S.A.M,StassenC.M, Fast J.H;Clinical and diagnostic features of partially anomalous pulmonary venousconnection in an adult female patient: a case report and review of the literature. Neth Heart J. 2011; 19(5): 256–258.
5. Babb JD, McGlynn TJ, Pierce WS, Kirkman P.M; Isolated partial anomalous venous connection: a congenital defect with late and serious complications. Ann Thorac Surg. 1981; 31(6):540–541.
6. Christos Tourmousoglou, Christina Kalogeropoulou, Efstratios Koletsis, Nikolaos Charoulis, Christos Prokakis, Panagiotis Alexopoulos; Right Upper Lobe Partial Anomalous Pulmonary Venous Connection. Case Reports in Vascular Medicine. 2014; 2014: 1-4
7. Van Meter C. Jr., LeBlanc J.G, Culpepper W.S.III, Ochsner J.L; "Partial anomalous pulmonary venous return," Circulation. 1990; 82:195-198.