Intrapulmonary Vertical Vein Associated With an Infracardiac Type of Totally Anomalous Pulmonary Venous Connection

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We encountered 2 cases of a very rare type of anomalous pulmonary venous drainage associated with infracardiac type of total anomalous pulmonary venous connection. The pulmonary vein from the entire left lung traversed the posterior mediastinum and joined the right pulmonary vein at the hilum. Furthermore, a large drainage vein coursed caudally through the pulmonary parenchyma and penetrated the diaphragm. The morphology of the pulmonary venous system could not be delineated by preoperative echocardiography. It should be noted that the vertical vein of this variant cannot be found, even if the posterior mediastinum is extensively dissected during operation, and exploration of the pleural cavity is necessary.


Intrapulmonary vertical vein is a rare anomaly of the pulmonary veins seen in total anomalous pulmonary venous connection (TAPVC) [1]. A large drainage vein running through the pulmonary parenchyma that collects the entire pulmonary venous blood from both lungs characterizes this anomaly. In all previously reported cases, the drainage site was in the supracardiac region such as the innominate vein or superior vena cava. In this report, we describe 2 extremely rare cases of intrapulmonary vertical vein associated with infracardiac type of TAPVC.

Case Reports

Patient 1

A 4-day-old male neonate with a 2.1 kg body weight and without heterotaxy was referred to our department for cyanosis and progressive acidosis. Preoperative echocardiography showed a horizontal common pulmonary vein posterior to the left atrium that connected with 2 branches from the left lung. The inferior vena cava (IVC) was dilated, and mosaic flow was observed. The preoperative diagnosis was infracardiac type TAPVC, although the descending vertical vein could not be identified. Emergency surgery was performed. Cardiopulmonary bypass was established with an ascending aortic cannula and bicaval venous cannulae. The posterior pericardium was explored and the left superior and inferior pulmonary venous branches were found to form a common pulmonary vein that ran horizontally. However, a vertical drainage vein, which usually has a tree-shaped configuration in typical cases of infracardiac TAPVC [2], could not be found despite extensive exploration of the posterior mediastinum. The pleural cavity was not entered. Anastomosis was carried out between the horizontal common pulmonary vein and the left atrium, and the atrial septal defect was closed. The postoperative course was uneventful and the angiography performed after the operation delineated the original configuration of the pulmonary venous system. There was pulmonary venous flow from the entire left lung that traversed the mediastinum and joined the right pulmonary vein, which ran caudally through the pulmonary parenchyma draining into the IVC close to the hepatic vein orifice (Fig 1A:B). The lowest segment of the drainage vein was stenotic, which could have been the cause of the patient’s debilitated preoperative condition. The newly created anastomosis between the pulmonary vein and the left atrium was widely open.

Patient 2

A 5-day-old male neonate weighing 3.5 kg without heterotaxy was brought to our unit for tachypnea and cyanosis. Preoperative echocardiography showed a common chamber posterior to the left atrium, with distension of the IVC and the hepatic vein. An infracardiac type TAPVC was suspected even though the descending vertical vein could not be identified. The posterior pericardium was explored after cardiopulmonary bypass was established. The left superior and inferior pulmonary veins formed a horizontal common pulmonary vein; however, a descending vertical vein could not be found. The right pleural cavity was entered. We assured that the pulmonary vein from the left lung connected to the right pulmonary vein at the hilum, and a large drainage vein ran down from the bottom of the right lung and penetrated the central portion of the right hemidiaphragm (Fig 2). This drainage vein was an isolated vessel from the mediastinum and surgical tape was placed around it without the need for dissection. Anastomosis was created between the horizontal common pulmonary vein and the left atrium. The atrial septal defect was closed with an autologous pericardial patch and the drainage vein in the right pleural cavity was ligated. The postoperative course was uneventful with postoperative angiography showing all of the pulmonary venous flow draining into the left atrium through the unobstructed anastomosis.

Comment

The commonly used Darling classification for TAPVC is based only on the level of the anomalous pulmonary vein
drainage. However, many variants can exist when the course of the drainage vein is taken into account. For instance, Everhart and colleagues [1] reported 2 cases with a large drainage vein running through the pulmonary parenchyma that collects the entire pulmonary venous blood from both lungs and drains into the superior vena cava; therefore, their Darling classification is supracardiac type. Since then, several cases with similar pulmonary vein have been reported [3–7] and all of them had a supracardiac drainage site. To the best of our knowledge, our report is the first to describe intrapulmonary vertical vein with infracardiac TAPVC.

Embryonic development of TAPVC can be explained by the common pulmonary vein atresia, which appears early, when the communication between pulmonary and splanchnic plexus is still preserved [8]. In typical infracardiac type TAPVC collateral channels between splanchnic plexus and the umbilicovitelline system persist and function as drainage to systemic vein. This drainage vein, known by vertical vein after birth, descends immediately anterior to the esophagus and penetrates the diaphragm through the esophageal hiatus, and most often joins the portal vein at the confluence of the splenic and superior mesenteric veins. The course of the pulmonary drainage vein in our patients is quite different from that in the common infracardiac TAPVC and it is noteworthy to mention that the pulmonary vein from 1 lung crosses the mediastinum and enters the contralateral lung. Details of the developmental process are not known, but for pulmonary veins to cross the mediastinum the anomalous process must be established during a very early stage, while the primordia of the 2 lungs are still in vascular continuity and before or during the time of canalization of the common pulmonary vein [1, 7].

Recently, most operations for TAPVC have been performed solely on the basis of preoperative echocardiography, in which some of the individual pulmonary veins or vertical veins are not visualized. When atypical morphology of the pulmonary venous system is suspected, contrast multidetector computed tomography is recommended. Additionally, intraoperative bilateral exploration of the pleural cavity should be reserved for the cases suspicious of intrapulmonary vertical vein.

References

Fig 1. Postoperative right ventriculogram in patient 1 (levophase). (A) The shape of the pulmonary venous system is clearly seen. (B) The vertical drainage vein is patent, although it is stenotic (black arrow). The broken line indicates the anastomosis between the pulmonary vein and the left atrium. (Ao = aorta; LA = left atrium; LV = left ventricle.)

Fig 2. Intraoperative photograph in patient 2. The vertical vein (*) is seen as an isolated vessel from the mediastinum in the right pleural cavity. It originates from the bottom of the right lung and penetrates the right hemidiaphragm. The white arrow shows the retracted right side of the pericardium and parietal pleura. (D = diaphragm; I = inferior vena cava cannula; L = lung; S = superior vena cava cannula.)